DONETSK NATIONAL MEDICAL UNIVERSITY OF THE NAME M.Gorky

HANDBOOK OF PAEDIATRIC SURGERY FOR STUDENTS OF THE V-ts AND VI-ts YEAR OF MEDICAL UNIVERSITY

DONETSK − 2008

УДК: 617−053.2(075.8)
ББК: 54.5+57.3я7
Т 42

HANDBOOK OF PAEDIATRIC SURGERY FOR STUDENTS OF THE V-ts AND VI-ts YEAR OF MEDICAL UNIVERSITY / Under the general release of professor V.N.Grona. − Donetsk, 2008. − 230 p.

Рекомендовано Ученым советом Донецкого национального медицинского университета им. М.Горького, протокол № 5 от 19.06.2008 г.

Рецензенты:

- заведующий кафедрой госпитальной хирургии и эндоскопии Донецкого национального медицинского университета им. М.Горького, д.мед.н., проф. **П.Г. Кондратенко;**
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В руководстве, которое подготовили сотрудники кафедры детской хирургии, анестезиологии и интенсивной терапии Донецкого национального медицинского университета им. М.Горького, представлен лекционный и теоретический материал, в котором детально излагаются вопросы этиопатогенеза, клиники, диагностики и лечения хирургических заболеваний у детей, согласно программе обучения студентов медицинских вузов по детской хирургии. Руководство предлагается англоязычным студентам для самоподготовки к семинарским и практическим занятиям.

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UMBILICAL HERNIA

Anatomy

At birth, the umbilicus is surrounded by a dense fascial ring that represents a defect in the linea alba. The umbilical opening is reinforced by strongly attached remnants of the umbilical arteries and urachus in an inferior direction and the more weakly attached umbilical vein in a superior direction. A layer of fascia (Richet's fascia), which is derived from the transversalis fascia, supports the base of the umbilicus, The peritoneum forms an intact undersurface to the umbilical ring, and skin overlies the umbilicus after the cord has separated. When the supporting fascia of the umbilical defect is weak or absent, a direct hernia will result. The umbilical hernia in children is surrounded by the dense fascia of the umbilical ring through which a peritoneally lined sac attached to the overlying skin protrudes. The umbilical ring continues to close over time and the fascia of the umbilical defect strengthens, accounting for the spontaneous resolution of this defect in most children.

An indirect umbilical hernia has also been described in which the peritoneal contents herniate from a point superior to the umbilical ring. The hernia follows the umbilical canal along the umbilical vein the linea alba in an anterior direction and a thin layer of preperitoneal fascia in a posterior direction. This form of herniation has been suggested to cause proboscoid hernias in children; in this defect, the umbilical cicatrix is displaced progressively in an inferior direction as the hernia enlarges. This defect may also be responsible for umbilical hernias in some adults.

The umbilical hernia of childhood is distinguished from a "hernia of the umbilical cord". Where there is a defect in the peritoneum as well as an open fascial defect at the umbilicus. Intestines herniate into the substance of the umbilical cord itself and are covered only by amnion. In effect, this hernia of the umbilical cord represents a small omphalocele.

Incidence and Natural History

In children, umbilical hernias in childhood occur with equal frequency in boys and girls. Numerous reports document a high incidence in African and African-American infants. The umbilical ring is open throughout most of gestation but becomes progressively smaller as gestation progresses. Most umbilical hernias in infants are recognized after

cord separation in the first few weeks of life and almost all are noted by 6 months of age. Most undergo spontaneous closure during the first 3 years of life. Umbilical defects are found in many premature infants after cord separation. Although umbilical hernias are commonly found in low-birthweight infants (75% of infants weighing less than 1500 g) most will resolve. The lack of accurate longitudinal studies of children with umbilical defects does not allow definitive conclusions to be drawn about their natural history. Umbilical hernias with a small ring diameter (< 1.0 cm) are more likely to close spontaneously and close sooner than those with large ring diameters (> 1.5 cm). The diameter of the umbilical defect is prognostically important, whereas the length of the protrusion is not. There are many umbilical hernias present at 5 years of age that will close spontaneously without an operation. The relationship between umbilical hernias that become symptomatic later in life and childhood umbilical defects is unknown. The protruding portion of the hernia generally remains unchanged while the fascial ring closes until it is too small to admit any contents into the hernia sac. The hernia thus tends to disappear abruptly. Umbilical hernias are commonly observed in patients with Down syndrome, trisomy 18, trisomy 13, mucopolysacchari doses, and congenital hypothyroidism. Umbilical defects (hernia or omphalocele) are part of the Beckwith-Wiedemann syndrome. Incarceration of intestine or omentum, strangulation, perforation, evisceration, and pain are rare events in the natural history of umbilical hernias in children. The most difficult task of the pediatric surgeon is to convince the family that observation alone will be successful in most cases and that an operation is not indicated for their child, especially in infancy. It is helpful to teach the parents to follow the size of the defect relative to the size of one of their fingers.

Surgical indications

Although repair of childhood umbilical hernias has been advocated to prevent the complication of incarceration in adults, the relationship between the two events is unclear. Rare events, such as incarceration requiring reduction, strangulation, perforation, and evisceration, are absolute indications for operation. Persistence and appearance are relative indications for operative repair. Although some researchers have advocated repair as early as 2 years of age, there is ample evidence to support the decision to postone repair until later in childhood. It is reasonable to repair

a large defect (> 1.5 cm) that persists to school age. The appearance of a hernia often drives families to insist that the hernia be repaired. If the child presents with a tender umbilical mass, the hernia may be reduced by milking the air out of the incarcerated loop of intestine and applying firm, steady pressure to the incarcerated mass. Admitting the patient for observation to rule out peritonitis and performing the operation the next day are appropriate. If incarceration resists reduction, an emergency procedure is warranted. In an infant with inguinal hernias and a concomitant umbilical hernia (unless the ring itself is > 2.0 cm in diameter) the umbilical hernia should generally be left alone, because it will probably close spontaneously. Because the inguinal hernia usually necessitates surgical correction, many children are often subjected to an operation that they do not need in order to avoid a subsequent anesthetic induction despite the fact that umbilical hernias persist in very few children.

Surgical Technique, Results, and Complications

Described procedures for the repair of umbilical hernias in children range from multiple layers of closure after opening of the peritoneum to closed techniques in which the peritoneal sac is treated like an inguinal hernia sac and suture ligated. Absorbable and nonabsorbable sutures have been advocated. The redundant skin of a large defect may be left in place and improves in appearance over time. Some investigators have advocated excision of the skin and reconstruction when a large proboscoid hernia is present. However, the fundamental technique of umbilical hernia repair has changed little since the description by Gross. A secure closure of the fascia, usually in a transverse fashion, and preservation of the appearance of the umbilicus are common to all repairs. Strapping and taping of the defects have been discredited.

Repair of an umbilical hernia is performed as an outpatient procedure with the patient under general anesthesia. An infraumbilical skin crease incision is made. A subcutaneous dissection is performed to circumscribe the sac. The sac is transected and may be dissected from the undersurface of the umbilical skin, but extensive and time-consuming dissection is unnecessary. Leaving a remnant of the peritoneal sac on the undersurface of the umbilical skin causes no complications. The sac may be trimmed to a strong fascial edge or simply folded to allow placement of interrupted absorbable sutures in a transverse orientation. To ensure accurate

placement of sutures, they are only tied after placement is complete. A second layer of closure is unnecessary. Inversion of the umbilical skin is maintained with fine absorbable dermal suture between the underside of the umbilicus and the midportion of the fascial closure. The skin is closed with intradermal absorbable sutures and dressed with a small dressing that applies adequate pressure.

Although infection predisposes the patient to recurrence of the hernia, these complications are rare. In 60 repairs of umbilical hernias at the Penn State University Children's Hospital during the past 5 years, only one child had a wound infection, which resulted in a recurrent hernia that was subsequently repaired without incident. Visceral injuries are possible but should not occur if the fascial edges are kept in view during the procedure.

PRESERVATION AND RECONSTRUCTION OF THE UMBILICUS

One of the goals of all umbilical surgical procedures is to restore a normal appearance. Omphalocele, gastroschisis, omphalomesenteric remnants, urachal remnants, bladder exstrophy, prune-belly syndrome, and umbilical hernia repair all involve surgical manipulation the umbilicus. The structures of the umbilical cord must either be retained and incorporated into a reconstruction of the umbilicus or a neoumbilicus must be constructed. Under unusual circumstances, the umbilicus must be reconstructed de novo when it is entirely absent as a result of previous surgical removal. The normal location for the umbilicus is at the level of the iliac crests, which overlie the third or fourth lumbar vertebrae.

In gastroschisis and omphalocele, fascial repair may often be performed through the circular skin defect that remains after the umbilical structures have been excised. This is true for both primary and staged closures. The circular skin defect may then be closed using an intradermal pursesrting suture that is incorporared into the middle of the fascial closure. Even if the fascial defect is enlarged after incision for the application of a Silastic chimney, the lower portion of the defect can be closed in a similar circular fashion, creating the apperance of an umbilicus. Others have advocated preservation of the umbilicus in the repair of abdominal wall defects and leaving the umbilical remnants in place in continuity with the skin closure. The umbilicus is abnormally located in all children with bladder exstrophy and is often associated with a small omphalocele defect.

It may be transposed more cephalad at the time of bladder closure to create a more normal appearance. If the umbilicus has been removed in exstrophy repair, it may be reconstructed. In children with prune-belly syndrome, the umbilicus may be preserved on a vascularized pedicle and located appropriately after removal of excess skin.

Many techniques have been proposed to reconstruct the absent umbilicus. The most straightforward is the Marconi technique, which we have adapted. An oval incision, 1.5 cm high by 2.0 cm wide, is created in the midline on the intercrestal line. The disk of skin is partially defatted, with a stalk of vascularized subcutaneous tissue being retained to maintain viability. A pursestring suture is placed intradermally around the defect (which expands considerably). The center of the skin disk is securely tacked to the midline fascia, which simulates the umbilical depression, and the pursestring suture is tightened until the defect is 1 to 1.5 cm in size. The skin is sutured circumferentially. Long-term absorbable suture is used throughout. A dressing that exerts adequate pressure is applied.

INGUINAL HERNIA AND HYDROCELE

History

In 176 A.D. Galen wrote, “The duct descending to the testicle is a small offshoot (processus vaginalis peritonei) of the great peritoneal sac in the lower abdomen”, an observation that established the pathogenesis of indirect inguinal hernia. As early as 1552 B.C., the Egyptians described inguinal hernias treated by external pressure. The earliest record of surgical therapy for hernia was by Susruta in the fifth century A.D.

During the early part of the nineteenth century, the anatomy of the inguinal canal was accurately described by Camper, Cooper, Hesselbach, and Scarpa. Lister’s antisepsis made the deliberate reconstruction of the inguinal structures feasible, and Bassini in 1887 and Halsted in 1889 reported the successful use of the now basic techniques of inguinal herniorrhaphy. Although Banks recommended in 1884 that hernias be definitively treated by well-fitting trusses, he did operate on a few patients when the truss failed. He described complete removal of the hernia sac through the external ring. In 1899, Ferguson described high ligation of the sac and reconstruction without altering the relation of the cord structures to the anatomic layers of the inguinal canal. He advocated exposure of canal by incision of the external oblique aponeurosis to facilitate the dissection. MacLennan in 1914 emphasized the desirability of elective operation as a definitive cure for inguinal hernia and influenced the transition from the use of trusses to definitive operation. He was also a protagonist of early discharge of the pediatric patient. Potts, Riker, and Lewis supported the Ferguson principle of exposure, simple high ligation, and removal of the hernia sac for routine hernia repair in children, and this is now the basis of current surgical management. Under ideal circumstances, the recommended age for repairing an inguinal hernia is at the time of diagnosis. The generally accepted procedure is high suture ligation of the sac at the internal ring. With certain exceptions, elective operation is now done in the outpatient surgical unit.

Embryogenesis and Patogenesis

Continued patency of the processus vaginalis is the principal factor in the development of congenital hernia and hydrocele. The patent processus vaginalis is a potential hernia; only when it contains some part of the

abdominal viscera does it become an actual hernia. The difference between a congenital hernia and a congenital hydrocele is the caliber of the processus vaginalis and the content of the sac; with a hernia, the processus is wider and contains an intraabdominal structure, whereas in the case of a hydrocele, the processus is narrow and contains only peritoneal fluid.

The processus vaginalis develops during the third month of gestation as an outpouching of the peritoneal cavity through the internal inguinal ring. At this time, the developing testis lies within the abdominal cavity. Descent of the testis commences after the seventh month of intrauterine life and is associated with extension of the processus vaginalis into the scrotum. The processes vaginalis appears to play an integral role in testicular descent, probably by providing the necessary hydraulic force that drives the testis into the scrotum.

The processus vaginalis obliterates spontaneously from the internal inguinal ring to the testis after testicular descent has been completed. The controlling mechanism is not known, but hormonal factors related to testicular descent may be in volved. Using an organ culture model, Hutson et a1 have shown a possible role for calcitonin gene-related peptide in the fusion of the processus vaginalis. The distal processus persists as the tunica vaginalis. Incomplete obliteration of the processus predisposes to various patterns of fluid accumulation (hydrocele) and hernia. The precise time of normal postnatal closure of the processus vaginalis is not known. Some claim that closure occurs immediately after birth; others state that a high percentage remain open for several years. Snyder and Greaney reviewed six published autopsy studies concerning infants and concluded that, at birth, the processus vaginalis remains in communication with the penitoneal cavity in 80% to 94% of examined bodies. These data are difficult to interpret because they are derived from studies done between 1785 and 1885; the definition of an open processus vaginalis is not clear; and in some series, whole bodies are referred to and in others, body halves. Snyder and Greaney quoted Sachs as reporting that the processus vaginalis was completely open in 57% of the bodies of infants between the ages of 4 months and 1 year. There is more recent information on the postnatal obliteration of the processus vaginalis, but this is derived from contralateral groin exploration in patients with an apparent unilateral hernia; it may be misleading to draw conclusions from these data because these patients all

had a hernia, demonstrating an inherent abnormality of the processus vaginalis. In boys with congenital hydrocele, most resolve within the first 6 months of life. Autopsy studies in adults have provided further information on the natural history of the patent processus vaginalis. A postmortem study of adults dying without clinically apparent inguinal hernias found a patent processus vaginalis in 20% of groins examined, and in other studies the incidence of patent processus vaginalis varied from 15% to 37%.

Although the above-mentioned studies provide no firm figures as to the frequency of a patient's having a processus vaginaIls at any age, they do indicate a relatively high incidence of patency in normal infants at birth. It appears that in apparently normal individuals the processus vaginalis remain patent for several months alter birth, and in 20%, patency persists undetected throughout life. From the aforementioned data, it is clear that a hernia is not inevitable when the processus vaginalis remains patent, and factors other than simple patency are involved in the development of a clinically apparent inguinal hernia. A number of such factors have been identified: urogenital (undescended testis, exstrophy of bladder), increased peritoneal fluid (ascites, ventriculoperitoneal shunt, peritoneal dialysis), increased intraabdominal pressure (repair of exomphalos/gastroschisis, severe ascites, meconium peritonitis), chronic respiratory disease (cystic fibrosis), connective tissue disorders (Ehlers-Danlos syndrome, Hunter-Hurler syndrome, Marfan syndrome, mucopolysaccharidosis).

Incidence

Inguinal hernia repair is one of the most common general surgical operations performed by pediatric surgeons. The incidence of inguinal hernia in children ranges from 0.8% to 4.4% and is higher in infants, commensurate with the higher rate of patent processus vaginalis.

Age and Maturity

The incidence of inguinal hernia is highest during the first year of life, with a peak during the first months. Approximately one third of children with hernia are less than 6 months of age at operation. The incidence is highest in premature infants, with reports ranging from 16% to 25%.

Sex

Boys are affected approximately six times more often than girls reported sex ratios range from 3:1 to 10:1 in favor of males. The relative

risk for inguinal hernia is higher in premature infants, but there is no clear evidence of a difference between genders in these infants. Although Rajput, Cauderer, and Hack reported that in very low birth weight infants 26% of buys developed an inguinal hernia compared to 7% of girls, others have not found a significant difference.

Side

The predominance of right-sided hernias is well established. Rowe and Clatworthy reported that in boys 60% of hernias occur on the right side, 30% on the left, and 10% bilaterally. The incidence was similar in girls (right side, 60%; left side, 32%; bilateral, 8%). In this report, a hernia was designated bilateral only if the diagnosis was made preoperatively; a simple patent processus vaginalis found on contralareral exploration was not considered a hernia.

Family History

An increased incidence of congenital inguinal hernia has been documented in twins and in individual families of patients with inguinal hernia. There is a history of another inguinal hernia in the family in 11.5% of patients.

Clinical features

An inguinal hernia appears as a bulge in the inguinal region that extends toward, and often into, the scrotum with crying and straining. It may be present at birth or may not appear until weeks, months, or years later. The clinical signs of an inguinal hernia are a smooth, firm mass that emerges through the external inguinal ring lateral to the pubic tubercle and enlarges with increased intraabdominal pressure. When the patient relaxes, the hernia either reduces spontaneously or can be reduced by gentle pressure first posteriorly to free it from the external ring and the upward toward the peritoneal cavity. The position of the testis must be determined before attempting to reduce the hernia because a retractile testis lying outside the external inguinal ring looks like an inguinal swilling and may easily be confused with an inguinal hernia. The distinction is made by manipulating the retracted testes into the scrotum and then examining the groin. An undescended testis may coexist with a hernia, in which case orchidopexy is required in addition to hernia repair. In girls, the ovary may herniate and has a 27% risk of torsion and infarction.

Typically the patient is referred to a surgeon after a hernia has been seen by the parents or a physician. Although the history may be characteristic, it is necessary to confirm the presence of the hernia by identifying the inguinal swelling. A quiet infant can be made to strain the abdominal muscles by stretching him or her out supine on the bed with legs extended and arms held straight above the head. Most infants struggle to get free, thus increasing the intraabdominal pressure and pushing out the hernia. Older children can perform the Valsalva maneuver by blowing up balloon or coughing, preferably while standing.

Detecting thickening and silkiness on palpating the spermatic cord as it crosses the pubic tubercle (silk glove sign) may be a useful diagnostic aid in patients in whom an inguinal hernia is suspected clinically but is not evident. When the hernia can not be demonstrated but has previously been identified by a physician, most surgeons (65%) accept the diagnosis and operate. If there is doubt or the hernia has not been seen by a physician, we reevaluate the patient at a second visit, and if the hernia is still not confirmed, we refer the patient back to the referring physician for follow-up.

Management

An inguinal hernia does not resolve spontaneously and must be repaired because of the high risk of incarceration, particularly during the first months of life. Sixty-nine percent of incarcerated hernias occur before the age of 1 year, and in these younger patients, the hernia tends to be irreducible. Rowe and Clatworthy found that 71% of incarcerated hernias requiring operative reduction occurred in infants younger than 11 months of age.

In most patients, elective inguinal hernia repair is safely done in the outpatient surgical unit. Exceptions are high-risk newborn infants and older children with cardiac, respiratory, or other disorders that increase the risk of anesthesia. These patients may be admitted either the night before or on the morning of operation and are kept in the hospital overnight for observation and monitoring.

In infants, endotracheal anesthesia, with its greater control over ventilation, is preferred. For otherwise healthy older children, facial or laryngeal mask anesthesia is adequate. In all patients venous access is obtained for administration of fluids and drugs. Region anesthesia, by

ilioinguinal and iliohypogastric nerve block or by sacral epidural block, is recommended to minimize postoperative discomfort and is used routinely in our practice. This is supplemented with a rectally administered nonsteroidal antiinflammatory analgesic. If necessary, subsequent analgesia is adequately provided by oral acetaminophen (Tylenol).

Surgical technique

The principal objective in correcting a pediatric inguinal hernia is high ligation of the sac at the internal inguinal ring. A useful landmark is the pubic tubercle, immediately lateral to which lies the external inguinal ring. A transverse skin crease incision is made with the medial end of the incision above and lateral to the pubic tuberele. The precise details of the incision depend on the age of the patient. The older child requires a more laterally placed incision to gain optimal exposure to the internal ring. The incision should be long enough to provide adequate exposure of the cord structures; a difficult and traumatic dissection can usually be attributed to inadequate exposure. The viability of the testis and the safe handling of the spermatic vasculature and vas deferens should never be jeopardized for the sake of a short incision.

The subcutaneous fat is separated to expose Scarpa’s fascia, which is well developed in children and easily confused with the external oblique aponeurosis. The latter has a characteristic appearance with obliquely running fibers. Scarpa’s fascia is picked up with fine hemostats and incised in the line of the skin incision, revealing a further layer of fatty tissue. This is cleared from the underlying external oblique aponeurosis. The external inguinal ring is identified lateral to the pubic tubercle, and the inguinal ligament and the fibers of the external oblique aponeurosis overlying the inguinal canal are delineated.

In the newborn, the internal ring lies almost directly beneath the external ring and can be comfortably approached through the external ring either with or without incising the external oblique aponeurosis. With increasing age, the two inguinal rings become progressively farther apart as the inguinal canal lengthens, and the internal ring is approached by incising the overlying external oblique aponeurosis. A short incision is made in the external oblique in the line of its fibers, lateral to the external ring. It is not necessary to incise the external ring itself. The incised edges of the external oblique fascia are elevated, taking care to identify and protect the

underlying ilioinguinal nerve. The opening may be extended into the external ring, but this is not necessary for an uncomplicated hernia. Dissection under the inferior flap of the external oblique aponeurosis exposes the spermatic cord and the inguinal ligament.

It is neither necessary nor desirable to dissect around the spermatic cord and lift it into the wound because this may damage the underlying transversalis fascia and destroy the integrity of the posterior wall of the inguinal canal. Using a hemostat, the fibers of the cremasteric fascia are separated to expose the cord structures and the hernia sac. With a pair of nontoothed forceps, the sac is lifted and the fine layer of connective tissue overlying the sac is dissected from it, revealing golden fat overlying the testicular vessels and the vas deferens, which is usually situated below the testicular vessels and adherent to the sac. The vessels and the vas must never be held with forceps to avoid injuring them. These structures can be safely dissected from the sac by pushing with forceps or by grasping the areolar tissue immediately adjacent to them and stripping this tissue off the sac. Should the sac tear, a pair of hemostats is used to oppose the edges of the tear to prevent it from extending through the internal inguinal ring into the peritoneal cavity.

After the cord structures have been separated from the sac at the point of dissection, the sac is inspected to insure that no abdominal viscera are trapped within it and then divided between hemostats. The proximal end of the sac is lifted up, and the cord structures are dissected away from it as far as the internal inguinal ring, where the preperitoneal fat is identified. After confirming that the sac is empty, it is twisted three or four times and transfixed and ligated at the level of the internal ring. It is helpful to use a retractor or slotted teaspoon keep the adjacent cord structures from being caught up in the suture. Twisting the sac strengthens it, and this maneuver also narrows the internal ring. A dilated internal ring may still be found when a large hernia is present and occasionally needs to be narrowed by placing one or two sutures in the edges of the transversalis fascia at the inferomedial aspect of the ring below the cord structures.

It is not necessary to remove the distal part of the hernia sac, which is simply left open. Splitting or everting the sac is not necessary. Attempts to remove a large adherent sac usually result in bleeding and hematoma formation and may injure the vas. Gentle traction on the testis returns the

testis to the scrotum and the cord structures to the inguinal canal. The external oblique aponeurosis and Scarpa's fascia are closed with interrupted sutures and the skin with fine subcuticular absorbable sutures. The skin closure is reinforced with adhesive strips or a layer of collodion. At the end of the operation, the testis is again pulled well down into the scrotum to prevent it from becoming adherent in a retracted position.

The surgical approach to the inguinal canal in girls is identical to that in boys. The hernia sac is identified and dissected up to the internal inguinal ring. In 21% of cases, the fallopian tuba, occasionally with the ovary or uterus, lies in the wall of the hernia sac and does not reduce into the peritoneal cavity. For this reason, we routinely open the sac and look for the fallopian tube. If the tube is not easily visible, traction on the round ligament brings it into view so that its exact position can be determined. The sac is ligated distal to the tube and excised. The base of the sac is allowed to retract through the internal ring. Some surgeons attach the base of the sac, including the round ligament, to the undersurface of the conjoint tendon to retain the normal support of the uterus (Bastionelli procedure).

If the fallopian tube is in the wall of the sac as a sliding hernia, no attempt should be made to dissect it free because this may damage the tube. The hernia sac is ligated distal to the fallopian tube and divided; the proximal sac is then invaginated into the peritoneal cavity through the internal ring, which may be closed with sutures placed through the transversalis fascia. We do not use the more complicated pursestring suture closure procedure described by Goldstein and Potts.

Contralateral exploration

In 1951, Duckett drew attention to the need for diligent examination of the opposite side when a single hernia is present and suggested contralateral exploration when there is the slightest evidence of a hernia. Since the report in 1955 by Rothenberg and Barnett of bilateral inguinal hernias in 100% of infants under 1 year of age and in 65.8% of children over 1 year, routine contralateral exploration has been a source of debate. Clearly, what Rothenberg and Barnett were describing was a patent processus vaginalis and not a true hernia. To make a reasoned decision regarding contralateral exploration, it is necessary to know the natural history of the processus vaginalis and the incidence of true metachronous contralateral inguinal hernias.

In children with a unilateral inguinal hernia, the reported incidence of contralateral patent processus vaginalis is 46% to 60%. Rowe, Copelson, and Clatworthy, using a strict definition of patent processus vaginalis of at least 2 cm in length, with patency demonstrated by a lumen confirmed by passage of a probe or injection of fluid or air, reviewed 1965 patients with a unilateral inguinal hernia and found a mean incidence of 48%, ranging from 63% in the first 2 months of life to 41% after the age of 2 years.

From their analysis of the published data, Rowe, Copelson, and Clatworthy concluded that in infants with a unilateral inguinal hernia, the contralateral processus vaginalis obliterates in the perinatal period or during the first few months of life in about 40% of patients, leaving 60% still patent during infancy. By 2 years of age, the patency rate has decreased to 40%, and after this age obliteration is unlikely to occur; half of these children (20% of infants with a unilateral hernia) develop a clinically apparent inguinal hernia sometime in their lives, but the remaining 20% live out their lives with a patent processus vaginalis of which they remain unaware. This evidence of persistent patency of the processus vaginalis is used to justify routine contralateral groin exploration, especially in infants, although the data indicate that the exploration is negative in two out of five infants and in three out of five older children.

More important than the patency of the processus vaginalis is the risk of an inguinal hernia developing. In 1962, Sparkman reviewed seven published reports on a total of 1944 children with unilateral hernia who were followed for periods of 18 months to 37 years. The average incidence of contralateral hernia was 15.8%, with a range of 4% to 34%. Bock and Sobye followed 174 patients with unilateral inguinal hernia for 27 to 36 years, and 14.9% developed a hernia on the opposite side. McGregor, Halverson, and McVay found a 22% contralateral hernia rate in 148 children followed for 10 to 20 years. From these data, the risk of a contralateral hernia is about 15%; more recent reports suggest a lower incidence, but the length of follow-up has been shorter. Given and Ruben reviewed 847 unilateral inguinal hernia repairs followed for up to 3 years of patients, 5.6% developed a contralateral hernia; this occurred within 1 year in 40% and within 2 years in 65%. Tepas and Stafford found only two contralateral hernias in 179 boys (1%) aged 6 to 24 months at operation

and followed for 3 to 6 years. The risk is similar in infants. Surana and Puri found 12 (10%) metachronous contralateral hernias in 116 full-term infants who had unilateral hernia repairs before the age of 6 months and were followed for 5 to 17 years, and Misra et al found a contralateral hernia in 20 (10.5%) of 190 infants who had unilateral hernia repairs before the age of 6 months and were followed for up to 5 years. Of 45 very-low-birth-weight infants followed for 20 months by Rajput, Gauderer, and Hack, 6 (13%) developed a contralareral hernia during the first 20 months of follow-up. Overall the risk of a contralateral hernia developing seems to be 10% to 15%, with reports ranging from 1% to 34%. This risk is lower than the 20% estimated by Rowe, Copelson, and Clatworthy based on patency of the processus vaginalis.

Routine Contralateral Exploration

The major advantage of contralateral exploration is that it allows the discovery and closure of a patent processus vaginalis, thereby preventing the future development of an indirect inguinal hernia. Although there are risks to the testicular blood supply and the vas deferens associated with exploring the spermatic cord for the presence of a hernia, evidence from unilateral explorations suggests that these risk are low. The mortality and the common postoperative complications do not appear to be increased by extending the operation to include contralateral exploration. A major objection to contralateral exploration is that in the majority of cases it is simply unnecessary. Although with a unilateral inguinal hernia the contralareral processus vaginalis is patent in 60% of infants under 2 years of age and in 40% of older children, the risk of contralateral hernia developing is 10% to 15%. Therefore, only one in six (and possibly only one in ten) children with a unilateral inguinal hernia develops a contralateral hernia. Conversely, of those undergoing routine contralateral exploration, at least five out of six children have an unnecessary operation.

Selected Contralateral Exploration

To reduce the number of unnecessary explorations, with the potential risk of operative injury, factors such as age, sex, side of the ipsilateral inguinal hernia, and patency of the contralateral processus vaginalis have been used as criteria for contralateral exploration.

Age

Kiesewetter and Parenzan found that 42% of contralateral hernias occurred in infants who had unilateral repair during the first year of life and 35% in those operated on between 1 and 2 years of age. These data and the higher incidence of a contralateral patent processus vaginalis during the first 2 years of life have led many surgeons to limit contralateral exploration to infants. In a review by the American Academy of Pediatrics, 40% of surgeons limited contralateral exploration to infants under 1 year of age and a further 39% to infants under 2 years of age. Others, however, have not a found a relationship between the risk of contralateral hernia and age at operation. As discussed earlier, the reported incidence of contralateral hernia in infants under 2 years of age is less than 15% and may be as low as 1%, so at least five out of six contralateral explorations in this age group are unnecessary.

Side of the Hernia

The side of the primary hernia is often used as a criterion for contralateral exploration in the belief that a contralateral hernia is more likely when the initial hernia is on the left side, presumably based on the fact that primary hernias are more common on the right. A review of five series showed only a small difference between the incidence of contralateral patent processus vaginalis on the two sides: 62% in which a left inguinal hernia was present and 56% in which the hernia was on the right. Similarly, when the data from the five series were combined, the risk of a contralateral hernia developing was the same whether the presenting inguinal hernia was on the left or the right side.

Sex

Many surgeons believe that a contralateral exploration should always be done when a girl has a unilateral inguinal hernia. This is based on a belief that there is a high incidence of patent processus vaginalis in girls and that an inguinal exploration in girls is safe. The reported incidence of positive contralateral explorations in girls ranges from 40% to 57%. The incidence of contralateral hernia developing, however, is much lower and is similar to that in boys. An exception may be girls under 2 years of age with a left-sided hernia who, according to Demicran et al, have a 24% risk of developing a contralateral hernia. On the evidence available, a selective approach based on the sex of the child does not appear to be justified. Furthermore, the risk to the fallopian tube during hernia repair should not

be under estimated. It is essential that steps be taken to identify the tube if inadvertent operative damage is to be avoided. The likelihood of encountering other reproductive structures that might be damaged at operation is low.

Patency of the Processus Vaginalis

For many surgeons, the criterion for contralateral exploration is the presence of a patent processus vaginalis, and various methods have been devised for identifying the patent processus. In the past, these have included preoperative herniography, intraoperative probing of the opposite side using a choledochal dilator, and intraoperative pneumoperitoneum to detect crepitus in the contralateral canal. Ultrasonography and laparoscopy have been used for this purpose.

Erez et al in 1992 reported 200 ultrasonographic examinations in children for identifying a contralateral hernia or patent processus vaginalis. Of 168 clinically apparent unilateral hernias, 156 were correctly diagnosed by ultrasound, and 12 were diagnosed as patent processus vaginalis; a further 29 of 32 bilateral hernias were correctly identified. The accuracy of diagnosis was 92%.

With the advent of laparoscopy, there has been increasing interest in this technique for direct examination of the contralateral internal ring at the time of hernia repair. In 1995, Chin, Liu, and Wei reported 333 children examined by laparoscopy at the time of unilateral inguinal hernia repair by inserting the laparoscope through the opened ipsilateral hernia sac. Of these patients, 107 (32%) had a contralateral patent processus vaginalis, the incidence of which decreased with age, ranging from 41% in infants less than 1 year of age to 37% during the second year of life and 19% in children over 10 years of age. There was no difference between boys (31%) and girls (34%). This is lower than the incidence based on operative exploration. In some instances, it was difficult to visualize adequately the opposite internal ring using either a 30-degree or 70-degree telescope. Because of this, some insert the laparoscope through an infraumbilical incision, which allows direct visualization of the internal ring, although this has the disadvantage that an additional incision is required. Holcomb, Brock, and Morgan reviewed 195 patients under 10 years of age with a clinical unilateral inguinal hernia who underwent laparoscopy. Of these, 91 (47%) were found to have a contralateral patent processus vaginalis, which

was confirmed at operation in all patients (patency was defined as an open length of 2 cm of more). In contrast, Fuenfer et al reported their experience with 77 children under 2 years of age who presented with a unilateral inguinal hernia and underwent laparoscopy. A patent processus longer than 1.5 cm was identified in 23% of these patients, an even lower incidence than that reported by Chin, Liu, and Wei.

These data suggest that if a patent processus is regarded as an indication for exploration, by using laparoscopy, 50% to 75% of children are spared an unnecessary contralateral operation. A patent processus does not inevitably lead to hernia formation. Because the risk of a contralateral hernia developing is about 10% to 15% and may be as low as 1%, even when contralateral exploration is restricted to patients with a patent ptocessus vaginalis, at least five out of six explorations are unnecessary.

Ipsilateral Inguinal Hernia Repair Without Contralateral Exploration

Because the risk of developing a contralateral hernia is approximately 10% to 15%, restricting the operation to the side with the hernia avoids unnecessary contralateral exploration in at least 85% of patients and thus avoids the potential risk of operative trauma to the spermatic cord structures. The major disadvantage of unilateral repair is the necessity of a second operation in children who develop a metachronous hernia. Although in the past a second operation has been held to be physically and psychologically traumatic, the use of outpatient surgery and developments in modern anesthesia and pain control programs have greatly reduced the risks and stresses associated with anesthesia and surgery.

A common argument used to justify contralateral exploration is that by eliminating the risk of an inguinal hernia, the danger of a possible incarcerated hernia is also eliminated. In practice, the risk of such an incarcerated hernia developing appears to be extremely low, and we are aware of only one patient in which this has occurred. This low incidence is partly because the parents, having already had experience with the initial hernia, promptly recognize the second hernia and seek treatment. Also, several authors have noted the short time interval between the initial hernia repair and the development of the contralateral hernia. Bock and Sobye found 27% of patients developed a contralateral hernia within 1 year and 50% within 3 years, and MacGregor, Halverson, and McVay and Given and Rubin found that of hernias developing after unilateral repair, 40% to

48% developed during the first year, and 65% had occurred by the end of the second year. Most contralateral hernias therefore develop within a few years of the first operation while the child is still relatively young and is under routine medical follow-up.

Comment

The risk of a contralateral hernia developing after repair of a clinically evident unilateral inguinal hernia is low, and despite the higher incidence of a contralateral patent processus in infants, the contralareral hernia rate is no higher than in older children. We therefore do not routinely explore the opposite inguinal canal in children of any age with a unilateral inguinal hernia. There are exceptional circumstances in which prophylactic contralateral exploration should be considered, for example, when a second anesthetic is considered a high risk (as in some premature infants with lung disease) or when it would be difficult for the patient to travel back to the hospital should a second operation be required. A further possible exception might be girls under the age of 2 years with a left-sided hernia, who have been reported to have a 24% risk of developing a second hernia, but this has yet to be confirmed by others.

Those surgeons who believe that there is an advantage to the patient in ligating a contralateral patent processus vaginalis should restrict this to patients in whom patency has been demonstrated. At present, laparoscopy appears to offer the most reliable method of providing this information, but has the potential disadvantages of a second incision, a longer operative time, and increased costs.

Irreducible hernia (incarceration and strangulation)

An incarcerated hernia is one in which the contents of the sac cannot easily be reduced into the abdominal cavity. A strangulated hernia is one that is tightly constricted in its passage through the inguinal canal and has become or is likely to be come gangrenous. Although incarceration may be tolerated in adults for years, most nonreducible inguinal hernias in children, unless treated, rapidly progress to strangulation with infarction of the hernia contents. Initially, there is pressure on the herniated viscera as they pass through the internal ring, inguinal canal, and external ring, leading to impaired lymphatic and venous drainage. This pressure, in turn, results in swelling of the herniated viscera, which further increases the compression in the inguinal canal, ultimately resulting in total occlusion of

the arterial supply. Progressive ischemic changes take place, culminating in gangrene and perforation of the herniated intestine. The testis is also at risk of ischemia because of compression of the spermatic cord structures by the strangulated hernia. In girls, the ovary may herniate and become strangulated or undergo torsion.

The incidence of incarcerated hernia was reported by Rowe and Clatworthy as 12% of 2764 patients and by Stephens et al as 17% of 228 patients aged under 10 years. In the series of Rowe and Clatworthy, the incidence was similar for boys (12%) and girls (17%); 82% were on the right side, and 83% occurred in boys. Two thirds of incarcerated hernias occur during the first year of life. The greatest risk is in infancy, with reported incidences of 28% to 31% for infants under the age of 2 to 3 months and 24% for infants aged less than 6 months. The incidence of incarceration apparently decreases in premature infants. Misra et a1 reported an incidence of 13% incarcerated hernias in 89 premature infants with an inguinal hernia, and Krieger et a1 reported a 17% rate of incarceration in 52 premature infants. Harper, Garcia, and Sia reported two (18%) incarcerated hernias in 11 infants weighing less than 1000 g. We speculate that this lower incidence in premature infants is because the internal ring is often wide in these infants, they often are nursed in a neonatal intensive care unit, where they are under the constant supervision of medical and nursing staff.

Diagnosis

The symptoms of an incarcerated hernia are irritability, pain in the groin and abdomen, and vomiting. A somewhat tense, nonfluctuant mass is present in the inguinal region and may extend down into the scrotum. The mass is well defined, may be tender, and does not reduce. Occasionally, it transilluminates and must then be distinguished from a tense hydrocele. Richter’s hernia is rare in childhood and is difficult to diagnose because there may be no local abnormalities or evidence of intestinal obstruction until the effects of intestinal ischemia are advanced.

With the onset of ischemic changes, the pain intensifies, and the vomiting becomes bilious or feculent. Blood may be noted in the stools. The mass becomes tender, and often there is edema and reddening of the overlying skin, with fever and evidence of intestinal obstruction. The testes are usually normal, but in some patients with prolonged strangulation, the

testis may be swollen and hard on the affected side because of venous congestion resulting from compression of the spermatic veins and lymph channels at the inguinal ring by the tightly strangulated hernia mass. Abdominal radiographs demonstrate features of partial or complete intestinal obstruction, and gas within the incarcerated bowel segments may be seen in the scrotum. Ultrasonograph may help to distinguish between a hernia and a hydrocele.

Differential Diagnosis

Torsion of the testicle.

Torsion of the testicle may occur acutely without previous history of hernia, but there may be a history of undescended testis. Typically, there is acute, severe pain with nausea and vomiting. The testis may retract upward, and local examination reveals the smooth, exquisitely tender testicular mass in the inguinoscrotal region. The swelling does not usually extend through the external ring into the inguinal canal.

Inguinal or femoral lymphadenitis.

There is usually evidence of recent infection in the area of lymphatic drainage to these lymph nodes, which manifest as tender, fixed, firm masses in the groin, lateral to the external inguinal ring. The inguinal canal and spermatic cord are usually normal.

Torsion of the appendix testis.

Presentations of torsion of the appendix testis is with acute pain in the scrotum. On examination, there is no evidence of an inguinal hernia in the inguinal canal or scrotum. Once the patient’s apprehension has been overcome, it is usually possible to determine that the testis itself is not tender and that there is an extremely tender nodule at the upper pole of the testis (blue dot).

Hydrocele of the cord or of the canal of nuck.

There is usually a previous history of swelling in the region of the inguinal canal. There are no associated symptoms, and the swelling is smooth, well-defined, movable, translucent, and usually nontender. The external ring can be felt, and there is a definite upper limit to the swelling. Occasionally an acute hydrocele of the cord cannot be differentiated clinically from an incarcerated hernia, and an operation is required.

Management

Nonoperative management.

An incarcerated, irreducible inguinal hernia, without evidence of strangulation in a clinically stable patient, should initially be managed nonoperatively. Reduction by gentle compression of the hernia may be attempted. The attempt should not be continued if the infant is crying and resisting the pressure on the hernia. If the infant is agitated, some form of sedation or analgesia is required before reduction to enable the infant to relax; this reduces intraabdominal pressure and relieves the pressure on the neck of the hernia sac at the inguinal ring. Care must be taken to avoid respiratory depression, especially in premature infants. Placing the infant in the Trendelenburg position may aid reduction by allowing the abdominal viscera to move cephalad, creating mild traction on the hernia contents, but we have not found this to be helpful.

The sedated infant is observed for a up to 1 hour. If, by this time, spontaneous reduction has not been achieved, gentle manual reduction may be attempted. Seventy percent to 84% of incarcerated hernias can be reduced in this fashion. Elective repair is performed 48 hours later, by which time there is less edema, handling of the sac is easier, and the risk of complications is reduced. Under no circumstances should reduction be attempted under general anesthesia because of the risk of injury to the herniated intestine.

Operative management.

When the hernia cannot be reduced or is strangulated, immediate operation is indicated to prevent further damage to the intestine and testis. If there is intestinal obstruction or strangulation, initial management includes nasogastric intubation, intravenous fluids, and broad-spectrum antimicrobial therapy. When fluid and electrolyte imbalance has been corrected and the child’s condition is satisfactory, exploration is undertaken. Should an apparently irreducible hernia spontaneously reduce after the child has been anesthetized but before the incision is made, the operation should proceed.

Inguinal approach

The skin incision is somewhat longer than for elective hernia repair, and the external oblique aponeurosis must be opened through the external inguinal ring to relieve the obstruction. The hernia sac is exposed and

opened. If the bowel is found to be viable, it is reduced, and the hernia sac is repaired by high ligation and division. If there is difficulty in reducing the bowel, the internal ring is enlarged by an incision made superiorly into the arching fibers of the conjoint tendon. This must subsequently be repaired. The testis should be inspected for viability.

If the incarcerated intestine reduces before the sac is opened, the sac should be opened and inspected. If there is no evidence of intestinal ischemia (i.e., no blood-stained or foul-smelling liquid) most surgeons (82%) proceed with hernia repair without attempting to visualize the previously incarcerated bowel. When there is evidence of intestinal ischemia, however, an attempt should be made to locate the previously incarcerated bowel through the sac to ascertain its viability. Should this not be successful, we recommend exploration through an abdominal incision in the right lower quadrant. If nonviable bowel is found, it is resected.

If the hernia does not reduce spontaneously and the viability of the incarcerated intestine is in doubt, the hernia sac is opened and the incarcerated bowel is lifted out for inspection, taking care not to allow it to slip back into the peritoneal cavity. It may be necessary to enlarge the internal ring to allow the bowel to be withdrawn into the wound until normal intestine is seen proximally and distally. The ischemic intestine is covered with a sponge moistened with warm saline for not less than 5 minutes and is then again inspected for viability, notably color, mesenteric arterial pulsation, and peristalsis. If it is judged to be viable, it is reduced, and the hernia is repaired. If the intestine appears to be nonviable, resection and end-to-end anastomosis are performed, either through the hernia incision or through a separate abdominal incision if more exposure is needed.

Operation for incarcerated hernia in the infant is particularly difficult because the sac is usually edematous and readily torn, and the testicular vessels and the vas are vulnerable to injury. Where possible, the sac is repaired by transfixion and ligation; however, for an edematous, friable sac, continuous or interrupted sutures may be more secure. The remainder of the incision is closed in the usual way.

Preperitoneal approach

In 1921, Cheatle described this approach for repair of inguinal hernias in adults, and subsequently Shandling and Thomson and Boley and

Kleinhaus advocated this approach for selected children with inguinal hernia. Others have used a transperitoneal exposure. Turnock, Jones, and Lloyd used the preperitoneal approach for 10 infants with irreducible hernia and found advantages over the inguinal approach: it was easier to identify the sac, reduce the contents, and dissect the cord structures; the sac and peritoneum could be opened if necessary to inspect the hernia contents or to resect bowel; and repair of a friable sac was facilitated.

A skin-fold incision is made at the level of the anterosuperior iliac spine, and a standard grid-iron incision is used to reach the preperitoneal plane. The internal inguinal ring and hernia sac are identified lateral to the inferior epigastric vessels. At the base of the sac, the peritoneum is opened, the contents are reduced and inspected, and any abnormality is dealt with as discussed previously. If reduction is prevented by a tight external ring, this can be approached through the same incision by dissection superficial to the external oblique aponeurosis and enlarged. After dissecting the sac from the testicular vessels and vas, it is divided and the peritoneum is closed. The incision is closed in the standard way. On the right side, the incision resembles an appendectomy scar, so the parents should be warned of this.

Postoperative complications

Scrotal Swelling

After hernia repair, fluid may accumulate in the distal sac, forming a hydrocele. Usually this resolves spontaneously; rarely, as piration may be necessary. Scrotal hematoma may follow excision of the distal sac.

Iatrogenic Undescended Testis (Ascending or Trapped Testicle)

Iatrogenic undescended testis is an uncommon and possibly underreported complication because in several large series the condition is not mentioned. Kiesewetter reported two patients with this abnormality after inguinal hernia repair in a series of 248 patients, and Hecker and Ring-Mrozik reported five patients with trapped testis in a series of 1957 patients, an incidence of 0.2%. Except in patients with congenital untdescended testis, the abnormality occurs either because the testis was not replaced in the scrotum at the conclusion of the hernia repair or because it subsequently retracted. Orchidopexy is required to correct this problem.

Recurrence

It is difficult to determine a precise incidence of recurrence after repair of an indirect inguinal hernia because factors such as sex and incarceration are not always clearly defined in reported series. In general, the reported recurrence rate for uncomplicated hernias is 0% to 0.8%; this rises to about 20% after operation for incarcerated hernia. In many series, patients were not contacted for long-term review; therefore, the true incidence of recurrence is not known and is probably higher than stated. Reports on patients with incarcerated inguinal hernias do not state whether the initial management was operative or nonoperative.

Various factors may predispose to recurrence; many of these are associated with the development of primary hernias. Grosfeld and Cooney, in a series of 25 patients with ventriculoperitoneal shunts, identified three recurrent inguinal hernias (12%). Incarceration is an important risk factor for recurrence. Steinau et al found that in 24% of 29 patients (25 boys, 4 girls) with a recurrent indirect inguinal hernia, the primary hernia had been incarcerated compared to a 7.6% incidence of incarcerated hernias in 2754 patients without recurrence. Other risk factors in their study were postoperative complications (9.4% recurrence rate) and concomitant diseases and abnormalities (31.2%). Contrary to Harvey, Johnstone, and Fossard found that the level of experience of the surgeon was not a factor, although technical inadequacies contribute to recurrence. Most recurrent hernias are indirect and probably result from tearing a friable sac, failure to dissect the complete sac, a slipped ligature at the neck of the sac, or failure to ligate the sac high at the internal ring.

Less frequency, a recurrence presents as a direct hernia or a femoral hernia. Of 34 recurrent hernias reported by Steinau et a1, four were direct and one femoral. In the Fonkalsrud, deLorimier, and Clatworthy series of 14 direct inguinal hernias, 4 followed previous repair of an indirect hernia (31%). A direct hernia occurring after an indirect repair is either a concomitant hernia that was not recognized at the original operation or new pathology caused by damage to the posterior wall of the inguinal canal during the initial dissection. A recurrent hernia in the femoral area is also likely to have been a missed hernia rather than a true recurrence.

Injury to the Vas Deferens

Accidental operative injury to the vas deferens is unlikely to be recognized until adulthood and possibly only then if the injury is bilateral. Sparkman reported an incidence of proven injury to the vas deferens of 1.6%, based on finding “segments of the vas deferens” in 5 of 313 hernia sacs from children who had undergone hernia repair. Details of the five cases were not published, however, and no histologic or clinical information is available. Walker and Mills found small glandular inclusions in approximately 6% of hernia sacs from prepubertal boys, which they believe to be mullerian duct remnants and not segments of the vas deferens. They emphasized that these structures were of no clinical importance. It is likely that similar structures accounted for sonic of the findings reported by Sparkman.

The vulnerability of the vas during hernia repair was demonstrated by Shandling and Janick. In their experiments, the vas deferens of rats were exposed and grasped with fingers, non-toothed forceps, bulldog vascular clamps, or mosquito hemostats. Serial studies of the vas were done over 6 months, and damage to the vas was found with all manipulations except digital handling.

The relationship between male fertility and previous inguinal hernia repair is also not well reported. Hommonnai et al reported findings in 131 men referred to a fertility clinic who had undergone inguinal hernia repair between the ages of 2 and 35 years. Although 14% of these men had testicular atrophy or abnormal sperm findings that could be related to the hernia operation, clinical details, such as the incidence of incarceration and experience of the surgeon, were not reported. Operative injury to the vas deferens during inguinal hernia repair may result in obstruction of the vas with diversion of spermatozoa to the testicular lymphatics, and this breach of the blood-testis barrier produces an antigenic challenge with formation of spermatic autoagglutinating antibodies. In a review by Friberg and Fritjofsson of 76 infertile men with spermatic autoagglutinating antibodies, 12 have had a unilateral inguinal herniotomy during childhood. In 10 of these men, the site of the previous hernia repair was explored, and in 5 of them, an obstruction of the vas deferens was discovered. The authors concluded that accidental transection and ligation of the vas deferens can and does occur during inguinal hernia repair in a child and may be a reason

for infertility in men after prepubertal hernia repair. Similar findings were reported by Parkhouse and Hendry. Thus, although these reports do not indicate the incidence of infertility of men after hernia repair, they do confirm that an association does exist.

Testicular Atrophy

The testicular vessels are vulnerable to operative injury, particularly in small infants, but reports of testicular atrophy after routine hernia repair are rare. Fischer and Mumenthaler and Fahlstrom, Holmberg, and Johansson each reported an incidence of testicular atrophy of 1%. In these studies, the operative methods used varied, and the number of incarcerated hernias was not reported; therefore, this may nor indicate the true incidence of testicular atrophy when hernia repair is performed by an experienced surgeon using a simple high ligation to obliterate the open sac.

With incarcerated hernia, the blood supply to the testis may be impaired by compression of the testicular vessels by the incarcerated viscus. The incidence of testicular compromise in association with incarcerated inguinal hernia ranges from 2.6% to 5%. The finding of a cyanotic testicle at emergency operation is common, approximately 11% to 29%. The actual incidence of testicular atrophy as indicated by histologic examination or diminished size at follow-up is much lower, however, varying from 0% to a maximum of 19%. Unfortunately, reported series of patients treated by emergency operation consist of small numbers of patients, and the length of follow-up and the criteria for evaluation of the testis vary considerably. Puri, Guiney, and O’Donnell, in an analysis of 87 boys with incarcerated hernia treated by nonoperative reduction, found unilateral testicular atrophy in two patients. From the available data, we conclude that vascular compromise of the testis is common, but the risk of actual infarction is low. Unless the testis is frankly necrotic, it should not be removed. The herniated ovary and fallopian tubes are also susceptible to vascular compromise and infarction.

Intestinal Injury

With incarcerated hernias, the incidence of intestinal infarction is remarkably low. Between 1960 and 1965, the incidence of intestinal resection in Rowe and Clatworthy’s report in 351 patients with incarcerated hernia was 1.4%. A review of three series published since 1978 indicates no resections in 221 patients with incarcerated hernia.

Mortality

Mortality associated with inguinal hernia is related to complications of the hernia or to coexisting risk factors, such as prematurity and cardiac disease. In 1939, Thorndike and Ferguson reported an overall mortality of 2.8% for incarcerated hernias treated below 1927 and 1936. In 1954, Clatworthy and Thompson reported one death in 135 patients treated for incarcerated hernia (0.9%), and in a report from the same institution in 1970 of 351 patients treated with incarcerated hernia, there were no deaths. Since then, deaths from incarcerated hernia have become a rarity. The risk is higher when the hernia is strangulated. In the United Kingdom in 1989, five deaths in infants with strangulated inguinal hernia were reported. The risk factors identified included age less than 6 months and lack of experience in pediatric surgery on the part of the surgeon and the anesthesiologist.

Special considerations

Premature infants

There is an increased incidence of inguinal hernia in premature infants, and a bilateral presentation is more common. Walsh, in a review of 82 infants weighing less than 2000 g, found a 13% incidence of inguinal hernia; of 28 infants weighing less than 1500 g, seven (25%) had an inguinal hernia compared to four (7%) infants weighing more than 1500 g who had an inguinal hernia. Rescorla and Grosfeld reviewed 100 infants under 2 months of age who required inguinal hernia repair; 30% of these infants were prematures, and 44% had bilateral hernias. Of 1391 VLBW infants (weight < 1500 g) reported by Rajput, Gauderer, and Hack, 222 (16%) developed an inguinal hernia between 28 days and 20 months of (corrected) age. Peevy, Speed, and Hoff studied 397 newborn infants and found a 9% incidence of inguinal hernias in infants weighing 1000 to 1500 g and 30% in those weighing 500 to 1000 g. In a small series reported by Harper, Garcia, and Sia of 37 premature infants weighing less than 1000 g, 11 (30%) developed an inguinal hernia; 2 of these 11 were incarcerated hernias (18%). Although the incidence of incarceration is in creased in infancy and may be as high as 28%, it appears to be lower in premature infants, with reported incidences of 13% to 18%, compared to mature infants.

There is strong evidence of an increased risk of postoperative life-threatening apnea in premature infants after repair of an inguinal hernia. In 1982, Steward reviewed 721 infants who underwent operation for inguinal hernia; 1 of 38 full-term infants, developed respiratory complications compared to 13 of 33 prematures (39%), including six who had apnea and required manual stimulation or mask ventilation. Liu et al reviewed 41 premature infants anesthetized for a variety of reasons; seven who had not required ventilation before operation developed apnea and required postoperative ventilatory support. They found that a history of apnea in infants less than 41 weeks of conceptional age was associated with a 50% chance of requiring postoperative ventilation, but apnea is not confined to patients with a positive history of respiratory distress syndrome. In Steward's series, only two of six patients who developed apnea had a history of apnea, and nine other infants with a positive history of apnea did not develop postoperative respiratory difficulties.

The cause of postoperative apnea is not known and may result from a combination of several factors. Immaturity of the diaphragm and intercostal muscles resulting in an increased tendency to fatigue has been documented, probably because of the small number of fatigue-resistant muscle fibers in the ventilatory muscles of premature infants. Further, infants with apnea have alveolar hyperventilation during sleep, and abnormal responses to hypoxia and hypercapnia have been demonstrated. Anesthetic agents, particularly halogenated hydrocarbons, depress the brain stem ventilatory control mechanisms and decrease the peripheral chemoreceptor response to hypoxia and may impair the power and endurance of the respiratory muscles.

Admission and careful monitoring of the these high-risk infants for 12 to 23 hours after operation is recommended. Rescorla and Grosfeld offer the following guidelines for preterm and seriously ill newborn infants: (1) infants with a reducible inguinal hernia who are already in the hospital (neonatal intensive care unit) are closely observed for irreducibility. The hernia is repaired before discharge, the precise timing of operation depending on any associated disease and the child's general status. (2) For ex-premature infants who are discharged from the neonatal intensive care unit and develop a reducible inguinal hernia while at home, early operations (after 44 weeks gestational age) is recommended because

of the high risk of incarceration. After operation, the patient is admitted to the hospital for a 23-hour period of extended observation.

Ventricular Peritoneal Shunts and Peritoneal Dialysis

A significant factor in the development of an inguinal hernia is excess fluid in the peritoneal cavity, and in patients with patency of the processus vaginalis, procedures that introduce fluid into the peritoneal cavity may precipitate a hydrocele or hernia. Whether hernia occurrence is due to the physical presence of the fluid or is secondary to increased intraabdominal pressure is not known. Abnormal neuromuscular function may also be a factor. Moazam et al reviewed 134 patients who had ventriculoperitoneal shunt procedures; 19.5% of children with myelomeningocele developed hernias and 47% of those with intraventricular hemorrhage, but all of the latter were premature. Grosfeld and Cooney found a 14% incidence of inguinal hernia after insertion of ventriculoperitoncal shunts; 20% developed an incarceration, and the hernia recurred in 16%. Based on this study, the authors recommended that (1) after ventriculoperitoneal shunt procedures, infants should be closely observed for the development of a clinical inguinal hernia, (2) operation should be done promptly after diagnosis of a hernia because of the increased risk of incarceration, (3) in these patients, the contralareral side should be explored in the case of a clinical unilateral inguinal hernia.

There is a well-established risk of inguinal hernia developing in patients on long-term ambulatory peritoneal dialysis, ranging from 7% to 15%. In such cases, a patent processus is likely to develop into a frank hernia. Intraoperative herniography is recommended at the time the peritoneal dialysis catheter is inserted; water-soluble contrast material is infused through the catheter, and the patient is placed in a head-up position for 15 minutes. If a patent processus vaginalis is demonstrated, repair is in order.

Sliding Indirect Hernia

The fallopian tube is frequently found in the wall of the hernia sac in girls and is at risk of injury. The operative management has already been discussed.

The appendix may also be found in the wall of a sliding hernia sac. Appendectomy, if it can be done safely, permits high ligation of the sac in the usual way. Alternatively the sac is ligated distal to the appendix, and

the proximal sac, with the appendix, is reduced into the abdominal cavity, with or without pursestring suture closure as for a sliding hernia in a girl. The internal ring may also be tightened around the cord to reduce the risk of recurrence.

In the infant, the bladder may lie beneath the internal inguinal ring and may be pulled down with the hernia sac during dissection. If this is not recognized, high ligation of the hernia sac may include the bladder wall, leading to hematuria, possible necrosis of the bladder wall, and extravasation of urine. This situation can be avoided by careful inspection of the neck of the sac at the time of transfixion. When there is any question about this possibility the sac should be opened and the contents inspected. Occasionally the bladder may extend down on the medial wall of the sac as a true sliding hernia. Shaw and Santulli recommend a flap operation, as in the Goldstein-Potts repair in the female, but we simply ligate and divide the sac distal to the bladder, invert the stump, and narrow the internal ring.

Direct Inguinal Hernia

A direct inguinal hernia is rarely encountered in children and usually presents as a recurrent hernia after repair of a congenital indirect hernia. In this situation, it may be a missed hernia or the result of damage to the posterior wall of the inguinal canal at the time of the primary operation. Wright encountered only 19 direct hernias in more than 1600 inguinal hernia operations. The diagnosis should be suspected if, when operating on an indirect hernia, a typical sac cannot be found; and a fascial defect is found medial to the deep epigastric vessels. Management is by repair of the transversalis fascia repair or Cooper’s ligament repair when sufficiently developed.

Femoral Hernia

Femoral hernias are also rare in children and are more often missed clinically than direct defects on examination or at the time of indirect hernia repair. Fonkalsrud, deLorimier, and Clatworthy reviewed 5452 patients with inguinal hernias, and Burke reviewed 4567 patients, a total of 10019 infants and children; there were 21 patients with femoral hernia (0.2%) whose age ranged between 6 weeks and 13 years, and there were 18 girls and 10 boys, a ratio of almost 2:1. The correct preoperative diagnosis was made in 8 of the 21 patients (38%). Four patients had bilateral femoral hernias, and in five patients the hernias were incarcerated. Wright reported

16 patients with femoral hernia and recommended repair through a femoral (infrainguinal) approach, suturing the inguinal ligament to the pectineal ligament and pectineal fascia.

Inherited Disorders of Connective Tissue

Patients with Hunter-Hurler, Ehlers-Danlos and Marfan syndrome frequently have inguinal hernias and are prone to recurrence unless the floor (posterior wall) of the inguinal canal is repaired in addition to the usual high ligation of the sac. Coran and Eraklis found that 36% of 50 patients followed with Hunter-Hurler syndrome developed inguinal hernias. The recurrence rate following high ligation alone was 56%, and formal herniorrhaphy was recommended.

Cystic Fibrosis

The incidence of inguinal hernia in cystic fibrosis is increased to between 6% and 15%. The incidence of absent vas deferens in the general population is 0.5% to 1%, based on vasectomy studies. In cystic fibrosis, abnormalities of the vas deferens ranging from obstruction to complete absence are invariably present and are usually bilateral. Failure to identify the vas deferens at operation should, therefore, lead to investigation for cystic fibrosis. Agenesis of the vas deferens is usually found in association with renal dysgenesis in patients who do not have cystic fibrosis, so evaluation of the upper urinary tracts is recommended under these circumstances.

Intersex

Rarely a phenotypic female with a palpable gonad in the labia may actually be a male with androgen insensitivity syndrome, or a true hermaphrodite. If an ovary is encountered in the hernia sac of a female patient, it should be carefully examined for evidence of testicular tissue. Males with androgen insensitivity syndrome do not have fallopian tubes and a uterus but have a small testis. Hermaphrodites may have a fallopian tube in the hernia sac, and examination of the gonad reveals an asymmetric ovotestis. In both situations, if an abnormal gonad is encountered, it should not be removed. Small wedge biopsy specimens are taken from each pole, the gonad is replaced, and the hernia is repaired.

Splenogonadal Fusion

Splenic tissue may be fused to an otherwise normal testis (splenotesticular fusion). Presentation is with a scrotal mass, and the usual

preoperative diagnosis is a testicular tumor. Orchidectomy is not necessary; intraoperative frozen section provides the diagnosis and allows preservation of the testis. We have encountered splenotesticular fusion is a high intraabdominal testis, which was removed. Splenoovarian fusion may also occur.

Adrenal Rest

Ecotopic adrenal tissue appearing as a small mass of yellowish tissue in the apex of the hernia sac is not rare and was found in 10 of 385 operations for inguinal hernia (2.6%), an incidental finding in each case. The adrenal tissue at this site is likely the result of attachment of developing adrenal cells to the testis before its descent from the retroperitoneum to the scrotum during fetal life.

Congenital hydrocele

Hydrocele occurs most frequently in early infancy, is usually noted to have been present since birth, and is often bilateral. Less commonly, a hydrocele does not become apparent until the child is several years of age; in this case, a typical presentation is rapid development of scrotal swelling, which, if it is tense, may be associated with pain, in infants, a typical hydrocele is a soft, bluish, cystic swelling surrounding the testis. It fluctuates in size, becoming smaller at night when the child is relaxed. Occasionally the child presents with a roundish, tense but painless mass, situated in the upper scrotum of inguinal canal, clearly separate from the testis; this is a hydrocele of the cord. A rare form of hydrocele is the abdominoscrotal hydrocele, a large, tense mass extending from the scrotum to the abdominal cavity and sometimes bilaterally.

Hydrocele is the main consideration in the differential diagnosis of an inguinal hernia that extends to the scrotum. Usually a hydrocele can be differentiated from an inguinal hernia by clinical examination. Careful palpation determines that the neck of the hydrocele narrows at the external inguinal ring and the hydrocele does not extend into the inguinal canal. In newborns and small infants, this can be difficult to determine, especially if the hydrocele is large and tense. Attempts to reduce a tense hydrocele can push it into the external ring, giving the impression of an incarcerated hernia, but the hydrocele is more mobile than a hernia and not tender. The hallmark of a hydrocele is its brilliant transillumination when tested in a

relatively dark room. A suitable light source available in most physicians of fices is an otoscope. Occasionally an incarcerated hernia containing gas-filled bowel transilluminates so diagnostic aspiration should never be attempted.

In most children with congenital hydrocele, the processus vaginalis closes, and the hydrocele resolves during the first 24 months of life. There is no evidence that a hydrocele will be come a hernia, although in theory a hydrocele is a potential hernia. The recommended management of a hydrocele, therefore, is to avoid operation during the first 2 years of life unless a hernia cannot be excluded with certainty. An exception is a large, tense hydrocele associated with discomfort and history of a hydrocele that frequently changes in size (communicating hydrocele), documenting a significant exchange of fluid between the abdominal cavity and the hydrocele sac.

After the age of 2 years, we operate if the hydrocele shows no signs of resolution or if it arises de novo because in older children a hydrocele is not likely to resolve. Sometimes a persisting hydrocele contains a strip of omentum that has slipped into narrow processus vaginalis. In most children, if not all, hydroceles communicate with the peritoneal cavity. Therefore, the operation for hydrocele as for hernia is high ligation of the processus vaginalis. The distal hydrocele sac is opened, and the fluid collection is emptied. The open sac should be left in place. Suturing the hydrocele sac edges open as used in adult practice is rarely required in children. Reaccumulation of fluid in the sac is rare and usually resolves spontaneously.

UNDESCENDED TESTIS

History

The importance of a descended testis has been known since ancient times, but the mechanism of descent remained obscure until Hunter (1762, 1786) dissected the human fetus and found the intraabdominal testis connected to the inguinoabdominal wall by a ligament, called the gubernaculum testis because it appeared to guide the testis to the scrotum.

Embryology

Testicular descent into the low-temperature environment of the scrotum in mammals is a complex multistage process. Up to the time of sexual differentiation in the human fetus at 7 to 8 weeks gestation, the fetal testis and ovary occupy similar positions. The gonadal positions then diverge; the testes remains close to the future inguinal canal, and the ovary moves away from the groin. The gonad is held by the cranial suspensory ligament (upper pole) and the gubernaculum (lower pole).

The cranial suspensory ligament, which persists in females, regresses in boys while the gubernaculum enlarges, especially at its distal end where it is embedded in the inguinal abdominal wall. The inguinal canal forms by condensation of mesenchyme around the gubernaculum to form the inguinal musculature. The mesenchyme of the gubernaculum persists to form a solid cord, which later becomes hollowed out by a diverticulum of peritoneum, the processus vaginalis. The proximal gubernaculum, which is initially attached to the gonad, becomes expanded by growth of the caudal epididymis. The processus vaginalis grows caudally into the gubernacular mesenchyme, partly hollowing out the gubernaculum. The caudal end of the gubernaculum remains solid, but the proximal part is divided into a central column attached to the epididymis and an annular parietal layer within which the cremaster muscle develops. At the start of the third trimester, the caudal end of the gubernaculum bulges beyond the inguinal abdominal wall and migrates across the pubic region to the scrotum. The processus vaginalis elongates proportionally inside the gubernaculum so that the testis can leave the peritoneal cavity within it. Scrotal migration of the gubernaculum and the testes is complete by 35 weeks. During migration, the gubernaculum is loose within the inguinoscrotal mesenchyme, suggesting enzymatic digestion of the adjacent tissues. After

migration is complete, the processus vaginalis becomes secondarily attached to the bottom of the scrotum.

The different phases of testicular descent are probably hormonally regulated. The hormones controlling descent and their mechanism of action remain controversial. The early phase of abdominal testicular descent is regulated separately from the migratory inguinoscrotal phase. Androgen controls regression of the cranial suspensory ligament of the testis, but regression of this ligament is not essential for testicular descent. Enlargement of the gubernaculum testes is controlled by a nonandrogenic hormone, perhaps mullerian inhibiting substance (MIS). Migration of the testis and gubernaculum from the inguinal region of the scrotum is under androgenic control. In instances of complete androgen resistance or gonadotropin deficiency, inguinoscrotal migration is absent.

The mechanism of androgen control of gubernacular migration is unknown, but there is mounting evidence that implicates the genitofemoral nerve. It has been postulated, although as yet uncertain, that the genitofemoral nerve releases calcitonin generelated pepride (CGRP), which may then indirectly control gubernacular migration. The physical force for migration of the testis is probably provided by intraabdominal pressure acting through the patent processus vaginalis.

Any anomaly disrupting normal testicular descent leads to cryptorchidism. The complexity of the normal process of descent suggests that causative factors for nondescent are multifactorial. Because most undescended testes are located outside the inguinal canal, the migratory inguinoscrotal phase of testicular descent is probably deranged more commonly. In my experience, intraabdominal testes are relatively uncommon, occurring in 5% to 10% of cryptorchid boys. In most cases, the undescended testis is located near the neck of the scrotum, just outside or a little lateral to the external inguinal ring, in the superficial inguinal pouch. Abnormalities of gubernacular migration may be related to defects in the migratory mechanism itself or failure of genitofemoral nerve function. Defects in the nerve may be caused by deficiency of androgen secretion during the second and third trimester as a result of deficiency of gonadotropin production by the pituitary or the placenta. Recognizable endocrine disorders, such as MIS deficiency or decreased testosterone

synthesis or receptor function, also cause failure of testicular descent but are relatively rare.

Undescended testes lying well outside the normal line of descent, such as in the perineum or femoral region, are rare, and their cause is unknown. Hutson et al suggested that this may be the result of an abnormal location of the genitofemoral nerve with consequent abnormal migration of the gubernaculum to the wrong site. The cause of transverse testicular ectopia also is unknown, but in animal models, transverse ectopia can be induced readily by cutting the gubernacular attachment to the testes so that the gonad is no longer required to exit the abdominal cavity through the ipsilateral inguinal canal. Increased gonadal mobility may permit accidental descent through the contralateral inguinal canal.

A number of inherited syndromes are associated with undescended testes. The underlying cause is not known, although many are associated with microcephaly, suggesting the possibility of pituitary hormone or gonadotropin deficiency. Some multiple malformation syndromes are also associated with neurogenic and mechanical anomalies, for example, arthrogryposis multiplex congenita. These disorders may cause cryptorchidism either by external compression of the deformed fetus or by intrinsic neurologic anomalies. Experimental inguinoscrotal compression during testicular descent is associated with undescended testes. Intraabdominal testes are characteristic of the prunebelly syndrome. The cause of the cryptorchidism is controversial, with thoughts ranging from a mesodermal defect to transient prenatal urinary obstruction. The absence of a processus vaginalis within the inguinal canal and the position of the testes on the posterior surface of the bladder are consistent with an obstructive cause. Ten percent of infants with posterior urethral valve also have cryptorchidism.

Cryptorchidism is common in infants with abdominal wall defects, such as gastroschisis, exomphalos (omphalocele), and exstrophy of the bladder. Undescended testes occur in more than 15% of infants with gastroschisis and a least a third of children with exomphalos or omphalocele. Whether this is caused by decreased abdominal pressure or other mechanical effects is not certain, although a role for abdominal pressure has been determined in experimental animals.

Neural tube defects have a high incidence of undescended testes. When there is a myelomeningocele affecting the upper lumbar spinal cord, the incidence of undescended testes is greater than one third. This could be caused either by abdominal wall paralysis and lower-than-normal abdominal pressure or by dysplasia of the genitofemoral nerve motor nucleus at the site of the myelomeningocele.

Separation of the body of the epididymis from the undescended testis is frequently observed. This is more common in intraabdominal and high inguinal cryptorchid testis. Whether this is the cause of the cryptorchidism or secondary to decreased androgen production in utero occurring simultaneously is not known. Experimental evidence in rodents treated with antiandrogens suggests that in utero androgen deficiency causes epididymal deficiency.

Abnormalities of the vas deferens occur commonly in boys with cryptorchid testes. The impalpable intracanalicular testes may have a vas deferens forming a loop, which protrudes distal through the external inguinal ring. Based on examination of the blood supply of such a long-loop-vas, Fowler and Stephens proposed transection of the main testicular vessels to the high undescended testis to permit orchidopexy with testis viability maintained by the redundant vas deferens with its collateral blood supply. Although this operation is not commonly performed as a one-stage procedure because of the high incidence of atrophy, it is now commonly performed as a two-stage procedure with the first-stage performed by laparoscopy.

Classification of Undescended Testes

Classification of gonadal position in undescended testes is complicated by the fact that the testis is mobile and not a fixed structure in the tunica vaginalis. Undescended testis is best defined as a testis that cannot be manipulated to the bottom of the scrotum without undue tension on the spermatic cord. A normally descended testis resides spontaneously in the lower scrotum even if it was retracted when the patient was first examined. The positions of undescended testes can be divided into those arrested in the line of normal descent and those in truly ectopic positions. The intraabdominal testis is usually located within a few centimeters of the internal inguinal ring, with the vas deferens and the testicular vessels traveling extraperitoneally and then entering the testis through a short

mesorchium. Such intraabdominal testes are often difficult to find through extraperitoneal exploration through the inguinal canal and are now relatively easy to identify at laparoscopy.

A canalicular testis is one that lies within the inguinal canal but may be difficult or impossible to palpate because of the over lying musculature. Such gonads may be squeezed out of the inguinal canal and palpated at the external inguinal ring, so-called emergent testes.

Undescended testes beyond the external ring may lie near the neck of the scrotum or may be lateral and a little above the external inguinal ring in the superficial inguinal pouch, originally described by Browne. In my experience, the latter location is rarely an indication of aberrant gubernacular migration because at surgery the gubernacular attachment is nearly always at or near the neck of the scrotum. Essentially, the superficial inguinal pouch is the space created by the tunica vaginalis in the groin and is limited superficially by Scarpa's fascia and its deep attachment to the fascia lata just caudal to the inguinal ligament. Whether testes in the superficial inguinal pouch should be labeled as ectopic is controversial, but because the gubernaculum is attached at or near the neck of the scrotum, they seem to be better classified as testes arrested in the line of normal descent. Truly ectopic testes may be located in the perineum, femoral region, pubopenile region, or contralateral hemiscrotum secondary to transverse ectopia.

Retractile Testes

Transient retraction of the testis out of the scrotum is a normal reflex caused by contraction of the cremaster muscle. This muscle functions to regulate the temperature of the testis and to protect it from extrinsic trauma. Retraction occurs as a result of low temperature or stimulation of the cutaneous branch of the genitofemoral nerve (inner thigh).

The normal retractile reflex is weak or absent at birth, and the scrotum is often pendulous. Later in childhood, when androgen levels are low, cremasteric contractility is significantly increased and the cremasteric reflex more pronounced. After 10 years of age, the reflex becomes less pronounced as androgen levels rise with the onset of puberty. The cremasteric reflex and normal retractile testis have been studied by Farrington, who found a high incidence of retractility in the middle of childhood.

At present, there is no consensus about what constitutes a retractile testis. Most clinicians agree that a retractile testis is a descended testis, although careful follow-up is required because it does not always remain descended. The retractile testis probably reflects a normal physiologic response to contraction of the cremaster muscle related to age. Goh and Hutson suggest that so-called retractile testes are, in fact, testes with acquired maldescent. As the distance between the external inguinal ring and the bottom of the scrotum increases with age, it is necessary for the spermatic cord to lengthen for the testis to remain located in the scrotum. Retractile testes may represent acquired maldescent secondary to failure of the spermatic cord to elongate with age, which may be a sequela of excessive contractility of the cremaster muscle in some boys.

Ascending Testes

A newly described variant of the retractile testis is the ascending testis. In many of these children, long-term follow-up studies have demonstrated that subsequent ascent out of the scrotum later in childhood is often related to delayed descent into the scrotum within the first 3 months after birth. Ascending testes are now being documented by a number of authors. The difference between ascending and retractile testes is otherwise not clear, and it may be that they are different names for a similar problem.

Studies suggest that not all undescended testes are present from birth. Many children with cryptorchidism present later in childhood despite attempts at screening in infancy. In addition, on careful questioning of such families, there is often no history of an anomaly at birth or in early childhood.

Acquired cryptorchidism is likely to be secondary to failure of the spermatic cord to elongate in proportion to body growth. Such testes appear to ascend out of the scrotum with increasing age, but measurements of the cord length suggest that this ascent is more apparent than real. This is certainly true in patients with cerebral palsy, in whom acquired cryptorchidism approaches 50% in postpubertal boys with severe spastic diplegia.

Atwell has suggested that ascending or retractile testes may be caused by persistence of the processus vaginalis, which is likely to inhibit elongation of the adjacent vas deferens and testicular vessels. In patients

with cerebral palsy in whom the cremaster muscle has proven spasticity, the cause is clearly related to abnormal muscle contraction. In cases in which the testis migrated to the scrotum prenatally and was present within the scrotum in infancy but the position is too high later in childhood, orchidopexy is often successful through a scrotal approach as described by Bianchi and Squire.

Incidence of Undescended Testes

In a landmark study in 1960, Scorer found the incidence of undescended testes was 4.3% in infants. By 1 year of age, the incidence had fallen to 0.96%. In 1986, the incidence of cryptorchidism at 1 year of age was 1.58% in British children. The John Radcliffe Hospital Cryptorchidism Study Group found that spontaneous descent occurred postnatally in the first 3 months; beyond that time, it was rare. The rate for orchidopexy in England and Wales has effectively doubled in the last two decades. Although this difference between the incidence of cryptorchidism and frequency of orchidopexy suggests that some orchidopexies are unnecessary, it may also reflect a true increase in the incidence of undescended testes. It has been suggested that one explanation of the apparent doubling of orchidopexy rates may be related to retractile testes. Because the recommended age for surgery for congenital undescended testes has decreased to age one or below, those children with acquired undescended testes are now more readily distinguishable from those children with congenital failure of gubernacular migration.

The frequency of undescended testes is significantly increased in premature infants. When birthweight is less than 1500 g, the incidence of cryptorchidism reaches 60% to 70%. The presumptive cause of this high frequency of cryptorchidism is that normal descent is not completed generally until about 35 weeks gestation. Most undescended testes in premature infants continue to descend postnatally, so that if such children are examined at 12 weeks beyond their expected normal delivery date, the incidence of cryptorchidism has fallen to more normal levels.

Complications of Cryptorchidism

Controversy persists about whether the testis is primarily abnormal leading to maldescent or alternatively is undescended and is secondarily abnormal. Evidence now suggests that abnormalities seen postnatally in undescended testes are secondary. Occasional primary abnormalities in the

hypothalamic-pituitary-gonadal axis, however, lead to inadequate hormone secretion, maldescent, and primary testicular abnormalities.

Species differences have made investigative studies regarding the effects of undescended testes difficult to evaluate. Many studies concerning cryptorchidism have been carried out on rodents, in which the important developmental aspects of gubernacular migration are complete by the tenth day after birth. The testis, however, does not descend into the scrotum until 2 to 3 weeks in mice or 3 to 4 weeks in rats, at the time of pubertal sexual maturation. Human gubernacular migration and testicular descent occur simultaneously and are normally complete before birth. The effects of undescended testes in the rat, therefore, do not become evident until after puberty.

Temperature effects

The scrotal testis resides in a specialized low-temperature environment with the pampiniform plexus, scrotal pigmentation, absence of subcutaneous fat, and regulation by temperature-sensitive muscles, such as the cremaster and dartos muscle, all ensuring decreased temperature of the epididymis and gonad. The scrotal testis in the human is maintained at 33°C compared with 34° to 35°C noted in the inguinal region and 37°C intraabdominally. The physiology of the testis is well adapted to this lower temperature, and therefore in the undescended testis where the ambient temperature is increased, the testis undergoes progressive alteration.

Endocrine effects

Steroid pathways in rat testes made cryptorchid by surgical fixation before puberty show no gross abnormalities, indicating that Leydig cells are still functional with cryptorchidism in this model. Measurement of testicular testosterone content in rats made cryptorchid at birth shows no abnormality up to 2 to 3 weeks of age but decreased testosterone production compared with controls after puberty. Gonadotropic regulation of both Leydig and Sertoli cells is abnormal after puberty. The number of Sertoli and Leydig cells, however, remains relatively normal. Functional derangements in Sertoli cells with cryptorchidism have been well documented by de Kretser and Risbridger.

Plasma gonadotropin and testosterone levels have been measured in infants with undescended testes, and the normal postnatal rise in plasma luteinizing hormone (LH) levels and testosterone were found to be

significantly lower than normal. It is difficult to determine conclusively whether this postnatal androgen deficiency is a primary abnormality or secondary to non-descent. Steroidogenesis in both normal and cryptorchid boys has been studied in great detail by Jockenhovel and Swerdloff.

In a study of premature infants born with a mean gestational age of 30 weeks, there was a persistently high incidence (19%) of undescended testes at 18 months, despite some testes descending postnatally. The normal rise in testosterone seen in the second and third month postnatally failed to develop in these premature infants with cryptorchidism. The authors concluded that inadequate stimulation of testosterone by human chorionic gonadotropin (hCG) in utero may contribute to the pathogenesis of undescended testes in this special group. Both plasma testosterone and LH levels were decreased in cryptorchid infants between 1 and 4 months of age. Although androgen levels may be deficient, androgen receptor levels in scrotal skin fibroblasts and testicular biopsy specimens taken at orchidopexy are normal in infants with bilateral cryptorchidism. Serum levels of MIS are normally elevated between 4 and 12 months of age, but in children with cryptorchidism this postnatal rise in MIS levels was inhibited.

Germ cell development

Germ cell deficiency in cryptorchidism was previously regarded as congenital. It has been observed, however, that the histology of the testes is initially normal and becomes progressively abnormal with age. After 5 years, the testis is macroscopically smaller and softer. More subtle abnormalities can be observed much earlier after birth. Mengle et al, in a histologic study of 752 testicular biopsy specimens in children between 2 months and 15 years of age, found a fall in the number of spermatogonia per tubule and a lack in the normal enlargement of the seminiferous tubule by 3 years of age.

Using electron microscopy, Hadziselimovic, Herzog, and Seguchi were able to identify histologic abnormalities in the Leydig cells in the second year of life. More detailed studies showed impaired Leydig cell development in undescended testes in the first 2 to 6 months, whereas the Sertoli and germ cells appeared normal. By the end of the second year of life, nearly 40% of undescended testes completely lost their germ cells. They concluded that germ cell deficiency in undescended testes was a

secondary abnormality because only 1 infant out of nearly 100 younger than 1 year of age lacked all germ cells, and 70% of biopsy specimens in such small infants had normal numbers of germ cells. More recently, Huff et al studied the transformation of gonocytes to spermatogonia, an early postnatal step in development of the germ cells, and documented that this early step in germ cell development is deficient in infants with cryptorchidism.

Some authors have proposed that these early stages in germ cell development are controlled by androgens and are hence deficient because of postnatal androgen deficiency. There is some contrary evidence to suggest, however, that MIS may be controlling early postnatal germ cell development.

Fertility

Fertility is lower in men with a past history of cryptorchidism. In previous generations, it was believed that the undescended testis suffered no adverse changes until after puberty and surgical intervention was not necessary until 12 to 15 years of age. The evidence that germ cell maturation is already abnormal within the first year of life has led clinicians to appreciate that not only is postnatal degeneration an important issue but that early intervention may prevent it. In animal studies, it is relatively easy to demonstrate that surgically induced or congenital cryptorchidism causes decreased fertility because of germ cell deficiency after puberty. Paternity rates are not deficient in unilateral cryptorchidism in both animals and humans, but with bilateral cryptorchidism, fertility is significantly impaired. Data attempting to correlate fertility rates with timing of surgery are not yet available because there are no long-term studies of children undergoing orchiopexy in the first year of life. Fertility in men with a history of retractile testes remains quite controversial, with some authors describing abnormalities on sperm counts that are not reflected in paternity rates.

Malignancy

The risk of a testis tumor occurring in men with a past history of cryptorchidism was at one time believed to be 35 to 50 times greater than normal. By using different methods of calculating the relative risk, Woodhouse suggests the actual risk is 5 to 10 times greater than the normal population. When looking at all men with testis tumors, a relative risk for

those with a history of unilateral cryptorchidism is fifteenfold or thirty-threefold for bilateral undescended testes with the risk of cancer being highest with intraabdominal testes.

The progressive degeneration of germ cells and dysplasia seen in cryptorchid testes is thought to be related to the increased risk of malignancy. Testis tumors are not common in childhood, and they usually occur at the same age as testis tumors in normally descended testes (i.e., 20 to 40 years). Giwercman, Muller, and Skakkabaek have speculated that testis tumors may be caused by an intrinsic abnormality in the testis rather than secondary dysplasia. They suggest that carcinoma in situ germ cells are the forerunner of invasive tumors and are, in fact, malignant gonocytes. Such germ cells displaying histologic characteristics of carcinoma in situ can be identified in neonates with dysgenetic testes and ambiguous genitalia. Skakkabaek et a1 have described the histologic features of carcinoma in situ and provided strong evidence that these abnormal cells are a prerequisite to invasive testis tumors. They recommend that young men with a past history of cryptorchidism should be offered testicular biopsy to exclude this condition before malignancy occurs.

Inguinal hernia

The processus vaginalis normally obliterates after descent of the testis in the perinatal period. Failure of the testis to migrate to the scrotum is associated with a higher incidence of inguinal hernia, in many cases leading to early surgical intervention because of the risk of incarcerated hernia. A clinically evident hernia present in association with a cryptorchid testis is an indication for immediate intervention. Most surgeons elect to perform a herniotomy and orchidopexy simultaneously.

Torsion of a cryptorchid testis

Previous surgical textbooks quote a high incidence of up to 20% for torsion in undescended testes. In the last 20 years, however, the trend to earlier surgical intervention has meant that most pediatric surgeons now rarely see torsion in an undescended testis. The mobility of a testis within the superficial inguinal pouch may predispose to torsion, but the exact frequency is now difficult to determine.

Trauma

Inguinal testes are thought to be at a slightly increased risk of direct trauma, although as with testicular torsion, early surgical intervention has

made this a less common problem. The most common clinical cause of trauma in an undescended testis I have seen is in children with cerebral palsy requiring wheelchair restraint. In these children, an inguinal testis may be compressed by the straps of the wheelchair.

Psychological factors

Cryptorchidism is a major psychological problem because the obvious physical abnormality of the genitalia promotes parental anxiety about subsequent fertility.

Testicular-epididymal fusion abnormality

Abnormal connection between the testis and the epididymis is common in cryptorchidism. The risk of abnormal fusion is greater with testes inside the canal or the abdomen than in inguinal testes or those lying at the neck of the scrotum. These abnormalities may be related to underlying androgen deficiency in utero, and in a percentage of these, the abnormality may be sufficient to interfere with fertility.

Diagnosis

The aim of the clinical examination is to identify the presence or absence of a palpable gonad and to determine the lowest position that it will sit comfortably without undue tension. The lowest limit of testicular position without tension probably corresponds to the caudal limit of the processus vaginalis. Examination should be conducted in warm surroundings and with the child relaxed. With the child recumbent on the examination table, the genitalia should be inspected for the appearance of the scrotum and any inguinal swelling suggesting a high testis or an associated hernia. Cranial traction on the suprapubic skin to expose the scrotum often makes testes that are retracted to the upper part of the scrotum conspicuous. The appearance of the scrotum varies dramatically with age, with the neonatal scrotum being thin, pendulous, and flabby compared with the middle of childhood, when the scrotum is small and puckered. If the testis is lying within the scrotum, it is usually visible through the thin scrotal skin. Hypoplasia of the hemiscrotum suggests that the testis has never been within it. A hemiscrotum of normal size is more likely if the testis is retractile or ascending.

The key to locating a suprascrotal testis is to remember that the testis is contained within the tunica vaginalis and is therefore mobile. In addition, the bony landmarks of the inguinal ligament should be identified. To locate

a testis in the superficial inguinal pouch, light palpation with the flat of the hand is most effective. If palpation is too hard, this often displaces the testis from under the fingers, so it may be missed. At least 80% to 90% of testes are palpable in the inguinal region or can be squeezed out of the inguinal canal and felt at the external ring by pressing firmly on the abdominal wall laterally near the anterosuperior iliac spine and pressing downward and medially toward the scrotum. Intraabdominal or intracanalicular testes that cannot be delivered outside the external ring are uncommon. Once the mobile testis has been identified in the groin, one hand of the examiner attempts to push the testis toward the scrotum while the other hand attempts to grasp it through the thin scrotal skin. The aim of this maneuver is to determine the lowest level to which the testis can be manipulated without undue tension. A normally retractile testis should be able to be brought right to the bottom of the scrotum and remain there. The position of the testis at physical examination can be documented by measurement from the pubic tubercle as described by Scorer, although this degree of documentation is usually unnecessary.

The most useful clinical observation is whether or not the testis can reside in the scrotum spontaneously. Examination of the scrotum in the newborn is easy because the testes are readily visible and palpable when in the scrotum. If the testes can be felt above the scrotum, the child should be reexamined at 3 months of age to see whether there has been delayed descent. If the testis remains out of the scrotum at age 3 months, a confident diagnosis of congenital undescended testis can be made. If the testis has descended within the first 12 weeks, there is a risk that it may reascend out of the scrotum later in childhood, and such children are best kept under close observation.

Determining the exact testicular position may be difficult if there is an associated incarcerated inguinal hernia. Once the hernia has been reduced by manual compression, the position of the testis can usually be identified.

The clinical distinction between a normally retractile testis and an undescended testis caused by a congenital or acquired abnormality can be difficult. Useful criteria for distinguishing normally retractile testes are as follows:

1. The testis can be brought fully to the bottom of the scrotum without difficulty.

2. The testis remains in the scrotum after manipulation without immediate retraction.

3. The testis is normal in size.

4. There is a history that the testis resides spontaneously in the scrotum some of the time.

If the testis cannot be palpated in the usual position in the groin near the external inguinal ring, the sites for an ectopic testis should be examined, such as the femoral region and perineum. Truly impalpable testes are relatively uncommon, being variously reported in 5% to 28% of boys with undescended testes. If the testis cannot be palpated, this implies that it is either intraabdominal (45%) or within the inguinal canal (up to 25%), or alternatively it may be absent (45%). The latter situation is known as the vanishing testis and is likely the result of intrauterine torsion of the spermatic cord during migration of the gubernaculum to the scrotum. This leads to secondary atrophy of the testis, and the contralateral testis is commonly enlarged, which is a useful physical sign.

Blind inguinal exploration for the impalpable undescended testis is unlikely to be successful. Numerous imaging techniques have been recommended to identify the position of such a testis. These include abdominal and inguinal ultrasonography, computed tomography (CT) scan, magnetic resonance imaging (MRI), and spermatic venography and arteriography. Laparoscopy has been used to identify the position of an intraabdominal testis and to exclude the possibility of secondary atrophy. Laparoscopy offers the additional benefit of ligation of the testicular vessels should a two-stage Fowler-Stephens operation be contemplated.

Treatment

Hormone treatment.

Hormone therapy is based on the premise that undescended testis is caused by deficiency of the hypothalamic-pituitary gonadal axis. Therapy has been tried with testosterone itself, hCG, and luteinizing hormone-releasing hormone (LHRH). Direct androgen therapy was abandoned many years ago because excessive doses caused precocious puberty. In the last 10 to 20 years, hCG has been in common use in Europe but less commonly used elsewhere. More recently, LHRH has been tried.

The results of hormone therapy have been mixed and depend on a number of factors. Success rates for treatment range from 10% to 50%. Children older than 4 years and those with bilateral undescended testes near the scrotal entrance or retractile testes respond most favorably to hCG. Testes in the superficial inguinal pouch, the most common variant of the anomaly, have a low success rate. It has been suggested that the successful cases are due to the fact that most were retractile testes. Christiansen et al reported a 25% success rate with hCG with complete descent, and in a further 25% the position of the testis was improved. On review of their article, however, it is difficult to exclude acquired variants of undescended testis such as retractile or ascending testes. In unilateral undescended testis, which is statistically much more likely to be of congenital origin, only 14% of boys have successful hormonal therapy.

In a randomized, double-blind study comparing hCG and LHRH, Rajfer et al found 6% of boys treated with hCG responded with testicular descent compared with a 19% success rate for LHRH. The authors concluded that neither hCG nor LHRH was effective in promoting descent of truly undescended testes. A double-blind, placebo-controlled study of LHRH nasal spray in boys with cryptorchid testis showed a 9% success rate with LHRH compared to 8% with placebo treatment. A second course of LHRH therapy increased the descent rate to 18%. Young children had the lowest response rate, and LHRH was not useful for impalpable testes.

Some clinicians suggest that hormone therapy diagnoses a retractile testis (which has a high success rate) and thereby avoids surgery. Acquired undescended testes with severe retraction or secondary ascent may respond to hCG treatment at levels of 100 IU/kg intramuscularly twice a week for 3 to 4 weeks. Alternatively, LHRH can be given as a nasal spray at 100 mg in each nostril six times a day for 3 to 4 weeks. In my surgical department, hormonal therapy is used rarely, and nearly all children with congenital or acquired cryptorchidism are offered orchidopexy.

Surgical treatment.

Treatment of cryptorchidism is based on the assumption that early intervention prevents secondary degeneration of the testes caused by high temperature. The scrotal testis is 3° to 4°C cooler than the intraabdominal core temperature, which is essential for normal postnatal testicular development. The timing of surgery remains controversial, with some

studies suggesting that delayed orchidopexy late in childhood is associated with good results, whereas others show poor results. Studies showing early degeneration of the germ cells in the first 6 to 12 months, ultrastructural electron microscopic changes in the second year of life, light microscopic changes in the third and fourth years of life, and macroscopic atrophy in school-age children all suggest that undescended testes undergo progressive degeneration after birth. Although the evidence that early surgery prevents this degeneration sequence is not yet available in human subjects, it is shown in nearly all animal studies.

For some years, orchidopexy has been recommended in the second or third year of life, but it is now my current practice to recommend orchidopexy between 6 and 24 months. This is because the first signs of damage to the testes are identified at about 6 months of age. Orchidopexy in such young children, however, can be challenging. In pediatric surgical centers, it is quite reasonable to perform orchidopexy in the second 6 months of life; however, in centers with less experience in small children, surgery between 18 months and 2 years may be safer. When orchidopexy is done in a pediatric surgical center, a younger age does not increase the risk of complications.

In my clinic, I recommend routine examination of all boys at birth with repeat examination at 3 months in those children in whom one or both testes was not descended at birth. If the testis remains undescended at 3 months, the child is best referred for orchidopexy in the second 6 months of life or the second year. When the testis has descended spontaneously in the first 12 weeks, such children are best observed every few years to ensure that they do not develop acquired undescended testes later in childhood. Children presenting with a concomitant inguinal hernia should have immediate orchidopexy done at the same time that the inguinal herniotomy is performed. This is much safer than delaying the orchidopexy after the herniotomy because reexploration of the inguinal canal in this circumstance has a higher risk of damage to the vas and vessels. In older boys presenting with what is presumed to be acquired maldescent, I recommend that surgery should be offered to these children (or alternatively hormone therapy) once the testis no longer resides spontaneously in the scrotum. This is a controversial recommendation to some but represents my current practice.

Orchidopexy is performed as an ambulatory procedure with the child entering the hospital or clinic an hour or so before operation and discharged a few hours later. Topical anesthetic cream is applied to the back of the hand so induction of anesthesia by injection is not painful. Under general anesthesia, a regional or local anesthetic block is performed to provide pain relief for the first few hours postoperatively. For inguinal undescended testes, a skin crease incision is made over the external ring and extending a little laterally.

A widely patent processus vaginalis is common (up to 70%) and needs to be separated from the vas deferens and the testicular vessels. The hernia sac is wrapped around the vessels anteriorly with the vas deferens posteromedial and the vessels posterolateral. In high undescended testes, particularly those +found in the inguinal canal, the hernia sac may completely envelop the testis so that the vas and vessels are inside the sac within a mesorchium. The method of separation of the hernia sac is that employed during routine herniotomy: the sac is stretched over the index finger while round-ended, non-toothed dissecting forceps gently sweep off the other cord structures, taking care not to damage the testicular vessels and vas deferens. En masse separation of the vas deferens and vessels is easier if the sac remains intact. If an opening is made inadvertently, the edges of the peritoneum should be picked up with forceps to maintain extensile exposure. The vas deferens is adherent to the back of the hernia sac, so it must be positively identified before the sac is divided.

Once the cord structures have been separated from the sac, safely identified, and protected, the sac is divided. Dissection is continued proximally up to the internal ring, where external peritoneal fat and divergence of the testicular vessels laterally from the vas medially indicates the retroperitoneum. The processus vaginalis is then transfixed and ligated at the internal inguinal ring.

Further mobilization of the testicular vessels in the retroperitoneal space may be achieved by dividing small fibrous bands laterally that hold the testicular vessels and prevent them being gently stretched to allow the testis to reach the scrotum. In older children, straightening the curved path taken by the testicular vessels may effectively lengthen the spermatic cord, but this advantage is much less evident in small children. Since the advent of laparoscopy, it can be seen that the testicular vessels actually take a

straight path from the abdominal aorta toward the internal inguinal ring and that retroperitoneal dissection is more likely to gain length by allowing greater traction and stretching of the testicular artery rather than by straightening the path taken.

The vas deferens usually has sufficient length to reach the scrotum without any special maneuvers. In difficult cases, however, the inferior epigastric vessels can be divided or the posterior wall of the inguinal canal can be opened medial to the inferior epigastric vessels and the testis taken medially to them. This may give an extra centimeter or so of length to the vas deferens.

An alternative operation, which is particularly suitable for boys with acquired maldescent, is the transscrotal operation described by Bianchi and Squire. A transverse incision is made at the neck of scrotum, and the tunica vaginalis is exposed, delivered through the wound and placed under tension. Loose connective tissue attachments to the spermatic cord are divided to expose the spermatic cord itself. Commonly, there is a residual fibrous strand of the processus vaginalis that is not fully obliterated. Once this fibrous strand has been divided, the vas and vessels stretch out to reach the bottom of the scrotum without difficulty. The testis can be anchored by closing the neck of the scrotum or by suture of the testis to the scrotal septum.

When the testis is located within the inguinal canal or the abdomen, the spermatic cord may have insufficient length to reach the scrotum despite the maneuvers described previously. In this circumstance, the surgeon has a number of choices available. If there is necessary expertise and back-up support, microvascular anastomosis can be performed, with transection of the testicular vessels and reanastomosis to the inferior epigastric artery and vein. This technique requires a high level of experience and skill with the operating microscope so it is not often used. More commonly, if the testis has been dissected but does not reach the scrotum, it can be sutured in the groin at the lowermost point where it reaches comfortably as a first-stage procedure, and second attempt is made 6 to 12 months later. Success rates for this two-stage orchidopexy have been quoted to be 70% to 90%.

An alternative approach is the Fowler-Stephens procedure, in which the testicular vessels are ligated intraabdominally and the testis swung

down on a long-loop vas supplied by collateral circulation from the artery to the vas and some cremasteric vessels. Radical dissection of the inguinal canal before making the decision to perform a Fowler-Stephens operation may jeopardize its success by damaging the collateral blood supply. This complication can be overcome by performing the Fowler-Stephens operation as a two-stage procedure with initial ligation of the testicular vessels without disturbance to the collateral blood supply; the subsequent second-stage operation then allows the testis to be mobilized on the enlarged collateral vessels from the vas deferens, which then usually reach the scrotum. Follow-up studies with this two-stage procedure report 70% to 90% scrotal position without atrophy.

Many surgeons are now adopting the use of laparoscopy for impalpable testes in the inguinal canal and abdomen. A staged Fowler-Stephens operation is then relatively straightforward, and early results are promising. In the first stage, laparoscopic localization of the testis allows a decision to be made about orchidopexy versus orchidectomy, which should be considered if the testis is small or dysgenetic. If orchidopexy is thought warranted, simple ligation of the testicular vessels above the testes can be performed with an endosurgical tie or a clip. Six months later, the testis can then be mobilized on a flap of peritoneum containing the collateral blood supply and swung down through the medial edge of inguinal canal to reach the scrotum. At present, I perform the second procedure by open inguinal surgery, although reports are appearing of this procedure also being performed laparoscopically.

Complications of surgery.

In the hands of experienced surgeons the risk of complications after orchidopexy is less than 5%. Damage to the testicular vessels leading to atrophy is the most feared complication, but this is relatively rare. More subtle damage to the vas deferens leading to occlusion of its lumen and subsequent interference with fertility is a theoretic problem, but its exact frequency is difficult to determine. Hemorrhage in the wound secondary to poor hemostasis occurs occasionally.

The most common complication after orchidopexy, particularly now that many are done in infants, is wound infection. Both the inguinal and scrotal incisions are at risk for infection at any age, but, in my experience, scrotal infection appears more common in infants. Usually this is of no

serious consequence and responds to simple drainage. Secondary ascent of the testis after orchidopexy is an uncommon but important complication caused by inadequate mobilization of the cord or inadequate fixation of the testis within the scrotal pouch. Postoperative lymphedema and vascular congestion of the testis after orchidopexy is a common finding that resolves spontaneously over the first month or two.

After orchidopexy, the child is usually reexamined 1 to 2 weeks later to remove the dressing and assess the short-term outcome. A further follow-up examination is performed 6 to 12 months later to determine whether there has been any significant atrophy or secondary malposition of the testis. The end result and appearance are satisfactory in the majority of instances.

Success rates are higher for those testes that have passed through the external inguinal ring, whereas intracanalicular or abdominal testes have a higher incidence of persisting abnormality after orchidopexy. The intraabdominal testis may fail to reach the scrotum, at least after a single-stage procedure, or it may be an inadequate gonad subsequently and atrophy. Total infarction of the testis is rare and is reported in 3% of patients with an impalpable testis. A further 15% to 20% have some atrophy after orchidopexy. The risk of atrophy is probably increased if a second operation is required to bring the testis to the scrotum. Exact figures in this subgroup however, are not available.

The risk of atrophy after orchidopexy is increased if a simultaneous inguinal hernia is performed for incarcerated or strangulated hernia. It is difficult to determine whether the increased rate of atrophy is secondary to compromise of the testicular vessels caused by compression or to the greater dissection required with a large hernia sac. One of the reasons given in the past for not performing surgery in early infancy was the belief that this led to a higher incidence of surgical complications. The timing for orchidopexy has therefore been a compromise between the potentially increasing risk of testicular dysplasia with age compared to the potentially increased risk of postoperative atrophy in younger children. Wilson-Storey, McGenity, and Dickson compared 100 infants younger than 2 years with 100 toddlers or older children undergoing orchidopexy after 2 years. They found an incidence of testicular atrophy of 5% in both groups, suggesting that the risk of postoperative atrophy is not directly related to age.

Fertility

A significant number of studies have evaluated fertility in men after orchidopexy. Testes initially located beyond the inguinal canal have a good prognosis for fertility. Interpretation of results is difficult, however, because most children currently studied had operations between 6 and 13 years suggesting that this group includes many acquired variants such as ascending and retractile testes. These latter patients are far less likely to have abnormal fertility because early germ cell maturation would have occurred normally when the testis was in the scrotum during infancy.

An extensive review of the literature has failed to demonstrate any significant improvement in fertility with early operation within the range of 4 to 14 years. Although 27 papers were reviewed, only four reports were recently published and we can no longer extrapolate data from operations done on adolescent patients before the 1950s and 1960s. With advances in knowledge and changes in clinical management in the last 25 years (i.e., earlier surgical intervention), it is inappropriate to compare these older historical studies with the results of current treatment. Previous studies confirm that fertility is poor when operation is performed late in childhood or early adolescence. Whether orchiopexy in infancy ultimately achieves a significantly improved rate of fertility remains to be seen.

Malignancy

At present, there are no accurate data available as to whether orchiopexy in early infancy reduces the risk of subsequent testicular cancer. There will be a lag time between the current trend of orchiopexy in infancy and convincing evidence that this change in the management alters outcome. At this time, all clinicians can do is define those features that appear to affect prognosis. Good prognostic signs include the testis near the neck of the scrotum, ascending or retractile testes, and possibly operation in early infancy. Poor prognosis is associated with primary dysplasia of the testis or epididymis, intraabdominal or intracanalicular testes, an associated strangulated inguinal hernia, and possibly operation delayed until late childhood or adolescence.

VARICOCELE

Dilation of the testicular veins in the pampiniform venous plexus causes a varicocele. The countercurrent heat exchange mechanism in the spermatic cord vessels is disrupted, which leads to an increased temperature of the testis and scrotum. The abnormally high temperature can be detected by thermography and causes progressive dysfunction of the testis and epididymis. This may lead to subsequent testicular atrophy and infertility, as first proposed by Tulloch. Testicular atrophy may be significant in adult life but may become evident quite early in adolescence. The incidence of varicocele is 15% among men in general and rises to 20% to 40% in men presenting to an infertility clinic. In children younger than 10, varicocele is rare, but by the end of adolescence, the incidence has risen to that seen in the adult population. Varicocele may occur in small children with a Wilms' tumor, neuroblastoma, or hydronephrotic kidney that causes obstruction of venous return from the testis. Although most cases of varicocele occur on the left, a right-side varicocele should raise the index of suspicion of a retroperitoneal tumor.

Clinical Presentation

Varicocele presents as a soft, distensible mass in the upper part of the scrotum. Approximately 80% to 90% of cases are on the left side, with bilateral lesions reported as occurring between 2% and 20% and right-sided lesions between 1% and 7%. In the supine position, a redundant left hemiscrotum and horizontal lie of the left testis may be noted. On standing, the varicocele fills with blood to produce the typical bag of worms appearance. The lesion is not usually painful; however, the boy may complain of a dragging sensation.

Varicocele may be classified by size into grades I to III, or small, medium, and large. Small varicocele (grade I) may be evident only during Valsalva maneuver. Medium-size varicocele (grade II) is palpable without Valsalva maneuver, and large varicocele is visible as a scrotal space-occupying lesion.

Etiology

Lack of valves in the left testicular vein is one of the primary factors in the etiology of varicocele. In postmortem examinations, the left testicular vein contains no valves in 40% of specimens compared with

absence of valves in only 23% of right spermatic veins. Cromie postulated that the right-angle entry of the left spermatic vein into the high-pressure venous system of the left renal vein predisposes to varicocele. When upright, the long pressure column generated in the papminiform plexus results in poor venous return and varicose distention of the veins. Skandalakis et a1 list a number of other etiologic theories, including disruption of the venous pump created by the coverings of the spermatic cord, compression of the left renal vein between the superior mesenteric artery and the aorta, extrinsic pressure on the left testicular vein by a full sigmoid colon, and vascular spasm at the origin of the left testicular vein caused by adrenaline coming from the left adrenal gland. In a series of 659 patients undergoing spermatic venography, absence of the valves, in the left testicular veins was documented in 484. In addition, a further 172 patients with varicocele had valves intact but had reflux of blood into the testicular vein through collaterals draining the left kidney. Renal vein stenosis was identified in 103 patients. The external spermatic vein (cremasteric vein) also has been implicated, as it may be dilated in up to 50% of varicoceles.

As the testis is supplied by three separate arteries, so is the venous drainage formed by more than one set of veins. Blood reaches the testis via the testicular or internal spermatic artery from the abdominal aorta, the differential artery supplying the vas deferens and the cremasteric, or the external spermatic artery arising from the external iliac artery and inferior epigastric vessels. These three vessels form an anastomosis around the chorda epididymis. Blood drains from the testis and epididymis into the pampiniform plexus accompanying the testicular artery. Above the internal inguinal ring, the number of venous channels decreases to one or two and finally coalesces into a single testicular vein entering either the inferior vena cava on the right or the left renal vein; the latter join at a right angle. Retrograde flow in the veins is prevented by the presence of valves. Anastomoses with subsidiary veins occur along the vas deferens to the base of the bladder through the cremasteric and scrotal veins to the saphenous vein.

Effects of Varicocele

Varicocele leads to testicular atrophy and subsequent infertility in adult life, probably secondary to abnormally high temperatures. How the

excessive temperature actually produces testicular dysfunction, however, is not so clear. A number of abnormalities have been documented in hormonal function and other physiologic parameters of the testis, but whether these are primary or secondary abnormalities is uncertain. This is particularly true for a proposed defect in the hypothalamic-pituitary-gonadal axis. Serum testosterone levels are usually normal, although a subclinical defect in the androgen axis is possible. Leydig cell hyperplasia with high serum follicle-stimulating hormone (FSH) levels has been reported. Abnormal hormone levels are not commonly diagnosed in adolescence but are more readily demonstrable in adults with established testicular atrophy. Inhibited testicular development during puberty is seen in association with histologic changes that are similar to those seen in adults with infertility caused by varicocele. Where testicular atrophy is recognizable, an abnormal production of pituitary hormones occurs in response to a gonadotropin-releasing hormone stimulation test.

Indications for Treatment

The criteria for treatment are controversial, with common indications including symptoms such as chronic pain or discomfort, demonstrable atrophy of the testis in adolescence, and subfertility in adults. When there is greater than a 10% difference in gonadal volume on orchidometry, Parrott and Hewatt advocate operation. Nagar and Levran have recommended screening of school boys by physical examination to identify varicocele at any early stage of development and found varicocele was present but asymptomatic in 10% of nearly 800 boys examined. They propose that screening enabled the diagnosis to be made at an earlier age and a lower stage of disease. They speculated that this should produce improved future fertility in adults, but this will not be proven for sometime. Doppler ultrasound is currently a popular test used in adult infertility clinics to identify subclinical varicocele. In early adolescence, however, when the testicular volume and blood flow is much less than in adults, the rule of Doppler ultrasound is less reliable. Diagnostic or therapeutic testicular venography is currently popular for men with varicocele. In young adolescents, general anesthesia is usually required for such a procedure although a study using therapeutic testicular venography with insertion of spring coils has been described using only local anesthesia.

Operation

The multiple theories of etiology of varicocele have led to a wide range of surgical options. Inguinal exploration has been a standard procedure for many years, with careful ligation of all the venous channels as described by Ivanissevich and Sayfan, Soffer, and Orda. Historically, this technique has been associated with a high incidence of secondary hydrocele, accidental ligation of the testicular artery leading to testicular atrophy, and recurrence of the varicocele. Poor results in some hands have led to a search for alternative approaches, including microsurgical dissection of the testicular veins, preserving the testicular artery and lymphatics within the spermatic cord. Some authors have suggested identifying the artery or veins intraoperatively so that the artery can be preserved. For this purpose, both Doppler ultrasonograph and venography have been used, but the role of these intraoperative investigations remains uncertain.

Shafik, Chalil, and Saleh have suggested plication of the external spermatic fascia to cause external compression of the pampiniform vessels. This is a simple procedure but does not address the persisting problem of retrograde flow of blood within the testicular veins. One might predict a high frequency of recurrence, although this is not currently known.

Laparoscopic ligation of the testicular vein proximal to the internal ring has gained popularity. Some authors have recommended selective ligation of the venous channels preserving the artery, whereas others have recommended mass ligation of the artery and the veins. Many publications attest to the feasibility of laparoscopic ligation, but long-term follow-up is not available.

Palomo first proposed mass ligation of the testicular vessels, including both artery and veins in the retroperitoneum above the internal inguinal ring. The technique has a high success rate with a surprisingly low risk of testicular atrophy as long as the collateral vessels have been preserved. The operation has been available for a long time but has gained only recent popularity. Fear of devascularizing the testis has made many surgeons cautious. As stated in Palomo’s publication, however, the blood supply of the testis should be maintained if any two of the three vessels are preserved. Because the cremasteric and deferential vessels are preserved, the collateral arterial supply of the testis should be intact, and no venous

channels are accidentally excluded by being mistaken for the testicular artery itself.

Retroperitoneal mass ligation as described by Palomo is my personal choice. I offer the family the option of having the procedure done laparoscopically or by a small open retroperitoneal approach. The laparoscopic method is similar to that described elsewhere in this book, with a 5-mm telescope port through the umbilicus and two 4-mm ports, one in the left abdomen level with the umbilicus and the other in the hypogastrium. With the surgeon standing on the right side of the operating table and the assistant on the left, the video screen is placed near the left foot. The colon can be displaced readily by tilting the patient. The peritoneum is opened several centimeters away from the internal inguinal ring, and the entire vascular pedicle including artery and all venous channels is ligated in continuity with a 3-0 absorbable suture.

The open procedure is also straightforward and may be done on an ambulatory basis. Under a short general anesthetic, an incision is made in the left iliac fossa medial and just a little below the left anterosuperior iliac spine. This is usually several centimeters away from the internal inguinal ring. The oblique muscles are divided along the line of their fibers just lateral to the rectus abdominis muscle. Once the peritoneum is visualized, it is mobilized medially by pledget and blunt dissection. Using this site of access, the peritoneal cavity is shallow, and medial displacement of the peritoneum immediately exposes the testicular pedicle well below the level of the ureter entering the pelvis. The vascular pedicle is isolated from the overlying peritoneum with a right-angle forcep, and two 3-0 absorbable ligatures are placed in continuity around the pedicle. It is not necessary to divide the testicular pedicle, although this is an option. On removal of the retractors, the peritoneum immediately resumes its normal position, and a few tacking sutures can be placed in the muscle layers and the skin closed with subcuticular sutures in the normal manner. Because this approach does not disturb all the lymphatic drainage of the inguinoscrotal region and leaves collaterals to the cremasteric and deferential vessels intact, the postoperative risk of an acute hydrocele is lower than with other procedures.

Results

The risk of recurrence or persistence of the varicocele is quoted as 5% to 45%. Risk of reactive hydrocele varies between 7% and 39%. The incidence of testicular atrophy, which is one of the most important outcome measures related to this surgery, is rarely reported, so accurate figures are unknown.

Ilioinguinal nerve damage has been reported after inguinal approaches and genitofemoral nerve injuries have been reported after laparoscopy as well as injury to the vas ligation of the veins within the inguinal canal using a modified Ivanissevich procedure has 16% recurrence, compared with high retroperitoneal selective vein ligation, which has a reported recurrence rate of 11%. They also compared 32 boys treated with mass ligation through the Palomo procedure in which they found no failures and no testicular atrophy after any of the operations. I concur from my own experience that the Palomo operation is the preferred technique. Gaur, Agarwal, and Purohit have reported a laparoscopic retroperitoneal Palomo procedure. The surgical management of varicocele continues to evolve, but at present the Palomo operation done either by open or laparoscopic approaches holds reasonable promise as having higher success and lower complication rates than previous procedures.

PYO-INFLAMMATORY DISEASES

Acute and Chronic Surgical Jnfection

Pyogenic surgical infection is among the most important problems of childhood surgery. Approximately 50 per cent of surgical beds are occupied by patients with purulent diseases and in 80 per cent of cases the fatal outcome is associated with purulent complications.

The problem of pyogenic infection has passed several stages in its development: (1) the pre-antibiotic period characterized by high lethality; (2) the period of the appearance of antibiotics, when the incidence of severe and persisting forms of purulent diseases and the number of complications sharply reduced; (3) the period of the appearance of antibiotic-resistant micro-organisms, this is the current period characterized by an aggravated course of purulent diseases, a tendency towards the development of toxic, septicopyaemic and atypical forms, and by an increase in the percentage of chronic forms.

Pathogenesis

Pathogenesis of acute pyogenic surgical infection. The form of a purulent inflammatory process is determined by the virulence and pathogenicity of the causative agent and the response of the child's organism.

The skin is often the infection atrium. This is associated with its fine structure and the property of being easily injured. For instance, the horny layer in the newborn is formed of two or three layers weakly connected one with the other and is therefore easily desquamated; the epidermis is separated from the dermis, the skin is injured very easily and tends to macerate and develop intertrigo, which especially applies to immature children.

Pyogenic infection is most frequently an "endogenous autoinfection of a sensitized organism" (I.V. Davydovsky). Foci of latent and chronic infection are found in the nasolarynx, upper respiratory tract, ears, lungs, and other organs of such patients as a rule. For the inflammatory process to originate, the infectious stimulation must exceed the macro-organism's sensitivity threshold which is determined to a great measure by the species and the amount (no less than 10 microbial bodies/g tissue) of the micro-organisms and the previous condition of the child. The state of immunity

and metabolism as well as the degree of sensitization have an effect on the level of this threshold. Micro-organisms are capable of sensitizing the microorganism and in this way reducing the sensitivity threshold and increasing the probability of the development of the disease. The most frequently encountered causative agents of septico-purulent diseases are as follows (in order of diminishing incidence): staphylococcus, Pseudomonas aeruginosa, Proteus, Escherichia coli. The formation of dangerous associations (up to 60 per cent of growths) and increased role of Gram-negative and conditionally pathogenic bacteria are characteristic phenomena. Processes of pathological symbiosis (S.Ya.', Doletsky) develop in the organism in pyogenic infection. This concept denotes a single process including the response of the whole organism (immunological and neurohumoral shifts, disorders of acid-base, water-electrolyte, and energy balance) and the local process.

Some specific features of the immunological responses of a child's organism. Complement titre and phagocytic reactions are rapidly exhausted when a pathological condition develops in previously healthy newborns despite the high values of some of the factors of non-specific immunity (e.g. C-reactive protein, lysozyme). In immature babies and in infants with intrauterine disorders and birth injury, these factors are particularly inhibited and continue reducing rapidly by the end of the third or fourth week. This determines the rapid exhaustion of the protective functions of the skin, mucous membranes, and blood on meeting an infectious agent.

The property of demarcating an inflammatory process is diminished in very young children because of the peculiarities of the blood coagulation system. The lack of the principal precoagulants, reduction of the prothrombin index by half, the fibrinogen level to 1750 g/l (175 mg/100 ml), and proconvertin to 30-50 per cent in the presence of a tendency to develop anticoagulant states (increased heparin, antithrombin, and fibrinolytic activity) considerably limits fibrin production and plasma coagulation in the focus of affection. The role of this factor in demarcation is reduced as a result and conditions are created for the spreading of the inflammatory process.

The haemostasis system is especially unstable in immature infants and in those with birth injury. "Spontaneous fibrinolysis" may occur even

in mild stress situations. All this makes immature children and children with a grave premorbid condition particularly susceptible to pyogenic infection and its severe course.

Specific humoral immunity at early age also has specific features. The titre of antibodies to the most commonly encountered antigens (Esherichia coli, causative agents of paratyphoid fever, staphylococcus**)** may be sufficiently high in the newborn mainly due to passive transmission of immunoglobulins G from the immune mother to the foetus by way of the placenta. Moreover, the antibody titre may grow in the newborn under the effect of the infectious stimulus. This may be due to the production of the child's own antibodies and also may be determined by the donor maternal immunocompetent cells and some of the nucleic compounds.

From the age of 2-3 to 4-6 months the titre of antibodies to the most commonly encountered antigens decreases. The phase of "physiological immunodeficiency" sets in. This is associated with the disappearance of the maternal antibodies (catabolism, reaction to heterogeneous protein) and slow synthesis of the child's own antibodies. Functional maturation of the immunological system begins from the age of about 6 to 8 months and reaches development by the 5th to 10th year of life.

Insufficiency of the immnnity humoral phase explains in many respects the frequency of toxic and septicopyaemic forms of infection in children and the tendency to its generalization. The titre of antibodies transferred from the mother is much lower in immatnre infants and in children with a severe premorbid condition than in healthy newborns.

Reaction of neurohumoral mechanisms in pyogenic infection. Stimulation processes occur rapidly from the effect of staphylotoxin on the central nervous system and change to superexcitation. This factor is very important for the child's organism because the nervous system is still functionally underdeveloped and highly sensitive to harmful effects and marked processes of irradiation occur. This partly explains the rapid development of neurotoxicosis in acute staphylococcal infection.

Massive, generalized stimulation of the central and vegetative nervous system alters reactivity, which is conducive to the development of hyperergic reactions. Hyperthermia develops very often, especially in very young children; it is due to heat emission disorders and accumulation of heat energy and is also of central genesis.

Haemodynamic shifts are particularly pronounced in pyogenic infection and are induced by the effect of microbial exo- and endotoxin as well as by histamine and histamine-like substances. Besides, proteolytic enzymes are released in tissue destruction, which also promote the lysis of protein substances and the increase in the amount of histamine and substances which have an effect on the vessels. Their action results in paresis of the vascular wall. Haemodynamic disorders, in some cases very pronounced, occur with the collection of blood in the paretically dilated vascular system. In an effort to maintain haemodynamics, particularly in the “central” organs (the brain), the organism sets some compensatory factors into action. Stimulation of the adenohypophysis, and possibly the direct effect of the toxin, intensifies the secretion of catecholamines (adrenaline, noradrenaline, etc.) which leads to an increase in the minute volume and peripheral resistance (due to spasm of the vessels). Catecholamines also cause blood shunting, i.e. blood flows from the arterial into the venous system, by-passing the capillaries and metarterioles. The blood volume necessary for circulation in the "central" organs is thus maintained ("centralization" reaction) though tissue metabolism on the periphery is disturbed (the periphery "gives up" some of its blood flow).

The "centralization" reactions play a positive role to a certain moment, but complications may develop in a prolonged and progressive process. The renal function is disturbed because "centralization" is quite pronounced here and shunting occurs in the juxtamedullary zone. The flow of blood through the glomeruli diminishes sharply, filtration is consequently reduced, and the excretion of waste products from the body is limited.

The large amount of adrenaline released into the blood raises the need for energy resources, cell hunger develops and is aggravated by increased catabolism resulting from intensified discharge of glucocorticoids. The increased requirements in energy resources lead to disintegration of endogenous protein and fat. Many incompletely oxidized products form in this case, to which tissue hypoxia consequent upon circulatory disorders contributes.

The disorders of peripheral haemodynamics and metabolism, energy hunger and the increased production of incompletely oxidized substances

change the acid-base equilibrium, usually to metabolic acidosis. Acidosis develops particularly frequently in children with purulent processes because metabolism at this age is very intensive. Under such conditions, the respiratory system works much harder as a compensatory apparatus (excretion of carbon dioxide and other acid products, supply of oxygen).

Severe microcirculatory disorders, hypoxia, and acidosis also lead to increased vascular permeability, extravasations and even to haemorrhages. In response to this, blood coagulation in the terminal parts of the vascular system sharply increases. As a result microcirculation is still more disturbed, hypoxia of the vascular wall becomes worse and its permeability increases. Lack of procoagulants occurs later due to their intensive expenditure and bleeding increases.

Thus, the following main syndromes can be distinguished among the variety of clinical manifestations in pyogenic infection in children: disorders of haemodynamics, acid-phase equilibrium, and water-electrolyte metabolism, the "neurotoxic syndrome", the syndrome of respiratory disorders, and the thrombohaemorrhagic sindrome.

The course of the local process in pyogenic infection in children is characterized by rapid development of necrosis and oedema, inhibition of the local phagocytic reaction, tendency of the process to spread, and massive resorption of the products of inflammation from the local focus into the blood and lymph. The oedema is particularly considerable in young children. Destruction of the stroma with the accumulation of water plays an important part in the pathogenesis of the swelling. This factor acquires significance in young children because increased hydrophilism of tissues is a condition normal for them. Increased vascular permeability induced by hypoxia, histamine, histamine-like substances and endotoxins facilitates the development of considerable swelling.

Early and massive activation of proteolytic enzymes in the zone of the lesion is a specific factor of young age and plays a particular role. The proteolytic enzymes promote the release of vasokinins, which causes extensive paresis of the local vascular system and increased porosity of the vascular wall.

The local barrier functions are markedly diminished in oedema and accumulation of fluid, especially in very young children, in whom humoral immunity is still insufficient whereas the perifocal vessels are compressed

by the oedematous tissue. These factors facilitate the sharp disorders of vascular trophics and the consequent rapid spreading of the process to the healthy areas. The phagocytic reactions are inhibited by the action of the micro-organism's "defence factor". Leucocytotoxin, coagulases, and other substances secreted by some bacteria, staphylococcus in particular, cause destruction of the leucocytes and deposition of fibrin around the microbes. The last factor sharply aggravates phagocytosis. The insufficiently functioning reticuloendothelial system is inhibited by the exotoxin and immunity is markedly diminished.

The poor barrier function of the regional lymph nodes and the well developed network of blood and lymphatic vessels are conducive to generalization of the infection and resorption of the products of inllammation. The infection tends to spread also because local fibrin production is delayed due to the peculiarities of the child's haemostasis and the deficiency of the reactions of macrophageal elements and phagocytosis which often ceases in the second phase (incomplete seizure).

As a consequence, degradation of the microbial capsule is inadequate and information about its antigenic structure is poor. This explains partly the insufficient increase in the titre of the antibodies and the absence of their strict specificity in young children.

Thus, the local process in pyogenic infection, particularly in young children, tends to develop rapidly. This is promoted by the marked swelling, the extensive vascular reaction, and the insufficiently pronounced demarcation reactions. The effect of some of the microbe's "defence and aggression factors" aggravates the process.

Fulgurant (toxico-septic), septicopyaemic, hypoergic, and local forms of pyogenic surgical infection are clinically distinguished. The first three forms should be related to the manifestations of sepsis, severe toxicosis of the organism as a response to infection under conditions of altered reactivity. "Septic shock" is considered a complication of pyogenic surgical infection.

The toxico-septic form sets in and progresses violently and is manifested by extremely severe toxicosis and fever. The diagnosis of the local focus is difficult. This form develops in sensitization (hyperergic phase) and lack of humoral antibodies. Gram-negative bacteria,

streptococcus or associations with staphylococcus are frequently encountered causative agents.

In contrast to the toxico-septic form, "septic shock" may develop in any form and in any phase of pyogenic surgical infection. The reaction is explained by the entry into the blood of large amounts of bacterial endotoxins, proteases, and histamine-like substances and activation of the kinin system. This is linked with the appearance of concomitant Gram-negative bacteria, impaired demarcation of the local focus, and weakened antitoxic immunity. This form is characterized by the following signs: in the presence of the main process (often when it is stabilized) phenomena of marked collapse rapidly appear after a rise in body temperature; psychic disorders and allergic reactions are frequent occurrences.

The septicopyaemic form is marked by prevalence of the inflammatory reaction over toxicosis, though the latter is also considerable. The local foci are manifested early. The response is normergic. The bacteria are usually Gram-negative and associations are encountered. In staphylococcal infection the pyaemic foci are for the most part large and mainly found in the lungs, bone marrow, kidneys, and the soft tissues. The appearance of *Pseudomonas aeruginosa* in attendance may intensify toxicosis and lead to the development of small pyaemic foci under the epicardium, pleura, and renal capsule.*Escherichia coli* and *Proteus* sharply intensify toxicosis.

The hypoergic form develops characteristically in immature babies, in children with congenital or acquired immunodeficiency, and in those exposed to harmful effects in the intrauterine period. The form follows a typically sluggish course without high fever, with progressive anaemia, hypoproteinaemia, and cachexia. Pyaemic foci are frequent.

The local form is marked by the prevalence of a local reaction and moderate toxicosis. The response is normergic.

Chronic stage. The shifts in the organism described above are much less pronounced in the chronic stage which may be characterized as the condition of dynamic equilibrium between the microorganism and the purulent focus. The micro-organism has become adapted to the effect of the macro-organism and, at the same time, the different functions of the organs and systems and the macroorganism's biochemical processes have also changed under the new conditions. This equilibrium, however, is

rather unstable and can always be disturbed in the direction of exacerbation of the infection.

At least two factors facilitate the change of the inflammatory process to the chronic stage, namely, the deficiency of the humoral phase of immunity and the massive character of the destruction of the macro-organism's own tissues.

The deficiency of the humoral stage of immunity is determined not only by the age factor discussed above, but also by inhibition of the immunocompetent systems by tosins or as a consequence of a grave premorbid condition; it can also be congenital. .4nother possible factor is the macro-organism's tolerance to the antigen if it has already met with it in the intrauterine period. In this case the immunocompetent organs "regard" this antigen as their own (autologous) and the immunological response is therefore weak.

Antibodies to the destroyed macro-organism's tissues appear as a rule in chronic pyogenic infection. This circumstance explains the poor efficacy of antibiotic therapy. the tendency to develop exacerbations now and then and to develop amyloidosis.

Products of incomplete tissue destruction form as the result of recurrent exacerbations, which may also possess antigenic properties. Fixation of these products in organs rich in reticuloendothelial tissue and in the kidneys occurs. The fixation is linked with the antigen-antibody type reaction and with the large size of the tissue fragments exceeding the renal filtration threshold.

The use of steroid hormones is conducive to the development of the process into the chronic stage. Immunogenesis is inhibited and the "auto-antigen" prevails over the "auto-antibody". As a consequence, any immunological conflict, whether superinfection, concomitant development of some other infection, disease or injury, is extremely undesirable in a chronic infection. Exacerbation of the main process in such cases may be caused by the response of the already present antibodies to the formed antigens due to superinfection or diminished threshold of immnnological sensitivity to the former antigen (weakening of the macro-organism as the result of intercurrent infection, disease or injury). Exacerbation may also occur as a consequence of tissue destruction and the reaction of the already existing antibodies to these tissues.

All that is stated above is not an eshaustive description of the vast variety of disorders occurring in the patient's body in the chronic stage of a purulent process. The difference in the pathogenic mechanisms of the acute and chronic stages, however, makes necessary a dizerentiated approach to the treatment under some conditions or other.

Principles of management of pyogenic surgical infection in children.

The treatment of pyogenic surgical infections in children is complex and includes three main components (T.P. Krasnobaev): (1) effect exerted on the macro-organism; (2) effect exerted on the microorganism; (3) treatment of the local process.

***Effect on the macro-organism*.** The maintenance and stimulation of the body's immunological properties is one of the main objects of the treatment of a purulent infectious process. In view of this, measures improving antibody production and cell immunity mast be applied.

Passive immunization is preferable in very young children with congenital and acquired immunodeficiency, toxic and septicopyaemic forms, and in the first 5 to 7 days of the disease. It is produced by injecting specific hyperimmune gamma globulin (3 ml at intervals of 1 or 2 days) and blood plasma (6-10 ml/kg). Passive immunization is conducted because active immunotherapy has a poor effect since the body's ability to produce antibodies independently is diminished. Direct transfusions of blood are also advisable in view of its high bactericidal properties, phagocytic activity, and the presence of antibodies in it. Preliminary single immunization of the donors with the corresponding toxoid is recommended in such cases because it increases considerably the antibody titre.

Active immunization (administration of autovaccines and toxoids) is resorted to only in children over the age of 6 months. and when the child recovers from an extremely severe condition and after the 5th to 7th day of the disease.

The staphylococcus toxoid is recently gaining wider use in children's surgical clinics for the management of purulent diseases. It possesses the property of inducing antibody production and at the same time it does not inhibit the body's reticuloendothelial system. The rate of the immunological response, however, depends on the condition of the macro-organism.

Pentoxyl (8 to 15 ml for each year of the child's life) is recommended for stimulating the reticliloendothelial system and improving the phagocytic reactions. The prescription is repeated, in severe cases no less than once in 3 to 5 days.

The next task consists in desensitization of the macro-organism, which is especially important in cases with staphylococcal infection. The effect of sensitization can be reduced with antihistamines and steroid hormones given in the age doses.

Hormonal therapy in pyogenic surgical infection is expedient in the following cases: (a) in children with congenital adrenal deficiency, and also when there is a history of a marked inadequate hyperthermic reaction in some mild inflammatory diseases; (b) in children with a severe pyogenic infection who received hormonal
therapy shortly before the principal disease; in such cases latent diminution of adrenal function should be apprehended, which may develop into marked dysfunction under the effect of stress, i.e. the pyogenic infection; (c) when it is necessary to prescribe large doses of antibiotics if there is danger of intensification of allergization; (d) in a marked allergic component; (e) when the disease follows a violent course and there is danger of adrenal cortical function exhaustion.

Prednisolone or hydrocortisone is usually given in the age doses with the exception of cases with adrenal insufficiency when medication with both preparations in increasing doses is expedient. It is best to administer them in the morning, except for patients with pronounced adrenal insufficiency.

To prevent possible complications of hormonal therapy, preparations or food containing potassium (5-10 ml of 10 per cent potassium chloride orally 3 or 4 times a day) are given in addition and anabolic hormones are prescribed. Rational combination of antibiotics in sufficiently high doses and continuous maintenance of their optimum concentration in the blood are very important. Passive immunization is advisable, particularly in massive and prolonged hormonal therapy. Hormones should be given for no longer than **7** to 10 days in pyogenic surgical infection, unless there are specific indications. Glucocorticoids are gradually discontinued, first reducing the dose or cancelling the last injection.

In view of the considerable tension of metabolism, the organism needs a large amount of vitamins, particularly those of the B complex (B1up to 30 mg, B2 up to 5 mg, B6 up to 8 mg) and vitamin C (up to 300 mg daily). Vitamin therapy is prescribed from the first hours of a pyogenic infection. Cocarboxylase (activated thiamine) is indicated; it is applied in daily doses of 0.03 to 0.1 g for 10 to 15 days to improve carbohydrate oxidation, especially in the nervous tissue.

Sedatives are prescribed because the staphylococcus toxin and the products of tissue disintegration have a stimulating effect on the central nervous system.

Marked shifts in haemodynamics, metabolism, and respiration occur in gravely ill patients with pyogenic surgical infection (peritonitis, septicopyaemia, etc.). The haemodynamic disorders are corrected by replacement of the blood volume, injection of pressor amines, namely, adrenaline (0.1 per cent solution, from 0.1 to 0.5 ml subcutaneously), noradrenaline, etc. (in the absence of blood volume deficiency). Steroid hormones, in doses one and a half to two times the age dose, are prescribed in pronounced symptoms of adrenal cortex exhaustion, inefficacy of the measures listed above, and in decompensated shifts in the acid-base equilibrium.

Energy preparations (solutions of carbohydrates with insulin, 1 U per 4 g of dry glucose, and emulsion of fat, up to 1.5-2 g of fat per 1 kg of body weight) are given in disturbed acid-base equilibrium. Oxygen therapy is conducted if indicated. Metabolic acidosis is corrected by the administration of alkaline or buffer solutions (sodium hydrocarbonate) and trisamine, alkalosis is corrected with potassium chloride and diuretics. The discharge and inactivation of toxic substances are successfully accomplished by forced diuresis, abdominal and intestinal dialysis, haemo- and lymphosorption, and the administration of "intravenous sorbents" (moderate- and macromolecular blood substitutes) and protease inhibitors.

Effect on the micro-organism. This is accomplished by antibiotic therapy in which a number of principles must be adhered to.

1. Rational combination of antibiotics. The bacteria must not be resistant to the drugs prescribed. When a bacteriostatic antibiotic is used, still another one, a bacteriolytic antibiotic, is prescribed. Combination of antibiotics inducing similar side effects is inadvisable.

2. Prescription of sufficiently large doses of antibiotics. In pyogenic infection following a severe course, the doses may be increased by one and a half to four times. The concentration of antibiotics in the blood must be two to four times its minimal inhibiting concentration.

3. Continuous maintenance of optimum concentration of antibiotics in the blood and local focus. In view of this the intervals between the injections should not be too long.

4. Conduction of antibiotic therapy for at least 5 to **7** days with full age doses.

5. Regular control over the antibioticogram (at least once in 5 to **7** days). With the appearance of resistance, the antibiotic is replaced by another one or its dose is increased. Before the antibioticogram is recorded, agents acting on both Gram-positive and Gram-negative bacteria are applied. The following combinations are most rational: gentamicin and ceporin or carbenicillin; oxacillin with ampicillin; penicillin with gentamicin and oxacillin. Combination of antibiotics with dioxidine has a good effect. Further treatment is conducted under the control of the antibioticogram.

In treatment with antibiotics, particularly for a long duration and in large doses, various complications, side reactions and the toxic effect of these drugs may occur. Hypo- and avitaminosis (usually of the B complex and vitamin C) are often encountered. **A** frequent occurrence is allergization of the organism, associated with the drug itself and with the products of microbial disintegration.

Dysbacteriosis may develop in prolonged or large-dose medication with antibiotics, particularly in weak children. This is due to suppression of the saprophytic bacteria and the prevalence and activation of resistant microbes (often Escherichia coli and fungi). Marked disorders of digestion, the formation of ulcers on the mucous membrane of the gastro-intestinal tract, and sepsis may be encountered in dysbacteriosis.

The gnotobiological method (both local and general) has been recently used with success in the treatment of pyoseptic diseases. It consists in isolation of the patient (or the region of the lesion) in special isolation compartments in which pathogenic bacteria are suppressed and replaced by non-pathogenic species competitive with the disease producing microbes.

Effect on the local process comprises a complex of measures. The following principles must be followed.

1. Sparing character of surgical manipulations on the purulent focus and the surrounding tissues in view of the danger of generalization of the process due to the poor lymphatic barrier and the extensive swelling.

2. Care is taken to cause minimum blood loss during the manipulations because children are very sensitive to it.

3. Maximum drainage of the focus and removal of non-viable tissues are provided.

4. Continuous maintenance of maximum concentration of antibiotics and antiseptics in the focus.

5. Administration of specific antibodies directly into the zone of the lesion.

6. Immobilization of the affected organ in the acute stage of the disease.

The purulent focus is cleansed as well as possible from the beginning of treatment by surgical, physiotherapeutic, and other methods. Manipulations in the perifocal areas are needed in some cases to diminish the possibility of rapid spread of the process. Anticoagulants are administered locally to reduce the production of fibrin
and its destruction around the focus of affection.

Continuous irrigation of the purulent focus with solutions of antiseptics and antibiotics is applied widely. This has an effect on the bacteria in the focus and removes the products of inflammation.

Thermal procedures (hot compress, local warm bath, paraffin and mud applications) are prescribed in the infiltrative phase of the inflammation, when there is still no pus. By increasing active hyperaemia, thermal procedures facilitate resorption of the infiltrate, improve tissue trophics, relieve the vascular spasm, and reduce the content of acid products in the focus of inflammation. As a result pain is considerably relieved.

Depending on the severity of the process, thermal procedures lead either to resorption of the infiltrate or to rapid purulent melting of the tissues.

Compresses with Vishnevsky's ointment and other irritants should not be applied in the case of very young children because their skin is highly

sensitive to chemical irritating agents. Vaselin and peachkernel oil are preferable.

Quartz lamp radiation has a bactericidal effect and is therefore used in surface inflammatory processes (erysipelas, some forms of pyoderma). Ultraviolet radiation causes a stimulating effect and promotes active surface hyperaemia. Quartz-lamp irradiation is therefore advisable for inducing rapid epithelization, stimulating granulation, as well as in aseptic phlebitis (after venepuncture or venesection). Ultrahigh-frequency therapy is applied in deeply located foci (osteomyelitis, infiltrates in the abdominal cavity, etc.).

Electrophoresis provides for local saturation of the focus with antibiotics, anticoagulants, and other drugs.

The common principles of treating pyogenic surgical infection are applicable both in the acute and the chronic stages, but the priority of the measures differs. The main objects of treatment in the acute stage are exertion of an effect on the microorganism, prevention of marked destruction, and normalization of homeostasis.

In applying treatment in the chronic stage, care is taken to remove completely the focus of infection, produce an effect on cellular and humoral immunity, apply measures for desensitization, and prevent superinfection and intercurrent diseases. It is also important to exert an effect on the micro-organisms by applying cyclic antibiotic therapy for preventive purposes and in exacerbation of the process.

Prevention of pyogenic infection is conducted from the neonate period. The staff of the accouchement unit and the department for the newborn are examined regularly for carriage of bacilli. Any pyogenic disease found is treated in time.

An extremely important measure is health education of the population, which is aimed at explaining the importance of observing hygienic measures in a family with a child. It is conducted at women's and children's consultation clinics and by visiting accoucheurs and nurses. "Risk groups" must be identified; these are formed of immature infants, children with multiple developmental anomalies, those who were exposed to intrauterine detrimental factors. Infants of these groups are kept under regular medical surveillance.

The problem of intrahospital infection, hospitalism, is recently raised more and more urgently. Its occurrence in surgical departments is particularly dangerous. Intrahospital infection is caused by pathogenic microbes present in the department; the microbes are antibiotic-resistant as a rule. The considerable number of purulent complications and the difficulty of liquidating intrahospital infection determines the necessity for strict observation of the sanitary and hygienic norms in the departments. The causative agent of pyogenic infection may enter the sick child's body from inadequately treated hands of the attending personnel or from the respiratory passages of a bacillus carrier. Bacillus carriers are found rather often among the personnel, especially those of purulent surgical departments. Physicians and nurses have frequent contact with antibiotics and therefore harbour mitigated bacterial strains which on entering the child's body may cause a disease under favourable conditions.

Infection can be transmitted from one sick child to another by the air route, on direct contact, as well as through bed clothes, instruments, and the hands of the medical personnel. The patient himself may be the source of the infection in some cases, particularly after operation on the gastro-intestinal tract or in dysbacteriosis.

The prevention of hospitalism is a very difficult matter. It comprises timely diagnosis and treatment of pyoderma and other inflammatory diseases, especially those of the upper airways and nasopharynx, and treatment of any skin injuries of the personnel of surgical departments. The hands, especially of the staff of departments and dressing rooms for patients with pyogenic diseases, must be thoroughly washed and treated. Special footwear and clothes are worn in the operating room. Clothes of wool and fur and fur footwear must not be worn in the surgical department. High demands are made of the medical staff with regard to personal hygiene.

Children with a pyogenic disease or postoperative suppuration must be isolated from "clean" patients. Overcrowding in one ward of a surgical department must be avoided. Care must be taken to apply timely preventive treatment of various intercurrent inflammatory diseases (otitis, diseases of the nasopharynx, oral cavity, respiratory tract, skin, etc.) before the operation. The walls and floor of the rooms in the surgical department (the wards, dressing rooms, bathrooms, and toilet) must be covered with

smooth water-proof material. The rooms are cleaned and ventilated as often as possible, especially if cases with intrahospita1infection were recorded. The mattresses and bedclothes arechanged often. The pillows, mattresses, beds, and night-tables used by patients with pyogenic infection are thoroughly disinfected.

Adequate blowing-exhaust ventilation with 400-600-fold air exchange must be provided in the surgical department as well as sterilization (quartz-lamp irradiation) of the wards and corridors.

Pyo-inflammatory Diseases of the Soft Tissues

Omphalitis

Omphalitis is the name given to an inflammatory process in the umbilicus and the surrounding tissues. Infection from the umbilical wound often spreads to the umbilical vessels as a consequence of which thromboarteritis with purulent melting of the thrombus develops and the arterial wall and surrounding tissues may be involved in the process subsequently.

Simple, phlegmonous, and necrotic forms of omphalitis are distinguished (A.I. Lenyushkin).

The simple form is characterized by delayed healing of the umbilical wound. **A** scant seropurulent discharge from the wound forms a crust. The child's general condition does not suffer.

In the phlegmonous form the fossa of the umbilicus turns into an ulcer covered with fibrinous deposits and surrounded by a firm thickened skin ridge. Hardened umbilical vessels are palpated in some patients. Pus is discharged from the umbilical wound when the periumbilical region is compressed. The child's general condition deteriorates little at first. There is a mild rise of body temperature. Children in whom omphalitis is complicated by phlegmon of the abdominal wall are restless, they suck poorly; toxicosis intensifies in such cases and body temperature increases to 39°C and more.

The necrotic form is the next stage of omphalitis and is encountered in debilitated children. The process spreads not only laterally, as is the case in phlegmon of the anterior abdominal wall, but into the tissues lying deeper. Necrosis of the skin develops and it is separated from the

underlying tissues. The process may invade the full thickness of the anterior abdominal wall and cause eventration of the intestinal loops.

Omphalitis may be complicated by sepsis.

The ***diagnosis*** of omphalitis is easily established in most patients. In cases poorly responsive to treatment, the calciferous origin of omphalitis has to be excluded in which the concrements formed along the distance of the vessels maintain the inflammatory process. Lateral-view radiography of the anterior abdominal wall helps in making the correct diagnosis in such cases; the radiograph demonstrates clearly the shadow of the concrement.

Omphalitis has to be differentiated from congenital umbilical fistula, its necrotic form from phlegmon of the newborn.

Treatment. The simple form of omphalitis is mainly managed by local measures (toilet of the weeping umbilicus with hydrogen peroxide solution followed by coagulation of the umbilical wound with a solution of potassium permanganate or brilliant green, and ultraviolet irradiation).

Complex treatment is applied in the phlegmonous form: antibiotics, stimulating therapy (transfusion of plasma every three or four days, gamma globulin injections), vitamins, desensitization therapy. In the early stages, antibiotics are injected around the umbilicus (the daily dose is diluted in 20-30 ml of 0.25 per cent procaine solution and the tissues adjoining the umbilicus are infiltrated with the solution from 2 or 3 points). If a purulent discharge appears, dressings with hypertonic (10 per cent) solution of sodium chloride and nitrofurazone (1: 5000) are applied and ultraviolet irradiation and UHF therapy prescribed. In calcinosis of the umbilicus the cavity is curettaged with a sharp spoon and then irrigated with a solution of antibiotics.

A solution of antibiotics is injected around the involved area in the necrotic form of omphalitis and then many cuts are made in the skin on the entire involved surface and at the junction with the healthy tissues. In addition to local treatment, a complex of general measures must be applied (transfusion of blood and plasma, injection of gamma globulin, transfusion of hyperimmune antistaphylococcus plasma, physiotherapy, vitamin therapy, symptomatic agents. Omphalitis may be a source of peritonitis, liver abscess, haematogenic osteomyelitis and other pyo-septic diseases. The outcomes of phlegmonous and necrotic forms are prognosticated very carefully, especially when omphalitis is complicated by sepsis.

Phlegmona Neonatorurnm

A peculiar form of purulent lesion of the skin and subcutaneous fat, characterized by rapid spreading of the process, is encountered in the newborn. The development of the phlegmon and its rapid spread is facilitated by the following specific features of the newborn's skin and subcutaneous fat: easy vulnerability of the epidermis, poor development of the basal membrane, relatively high content of intercellular fluid, richly developed vascular network, and deficiently developed connective-tissue septa in the fatty tissue. Injury to the skin of a child given improper hygienic care promotes the entry of infection through it.

The causative agents are the staphylococcus and Gram-negative or mixed flora. The sacrococcygeal and scapular regions and the anterior and lateral surfaces of the chest are the favoured sites of the lesion.

Pathomorphological examination of the focus of affection reveals evident predominance of necrotic processes. The inflammation begins for the most part around the sweat glands. Changes are sharpest in the deep layers of the subcutaneous fat. The perifocal blood vessels are thrombosed in a short time. Signs of sclerotic endo- and periarteritis are usually pronounced in their walls. The thrombosis of the vessels and the oedema of the subcutaneous fat, spreading with lightning speed, lead to sharp disorders of nutrition of the subcutaneous fat and skin and to their subsequent necrosis.

The inflammatory process is not demarcated because the immunological reactions are still deficient and the structure of the skin and subcutaneous fat possesses specific features, so that a large surface may be involved in the process within a few hours.

The ***clinical picture*** of phlegmon of the newborn is rather typical. The disease often sets in with general symptoms. The child is listless, restless, sleeps badly, and refuses to take the breast. Body temperature increases to 38-39°C. **A** red spot appears on the involved area and grows larger rapidly in a few hours. The skin is at first reddish purple but later turns cyanotic. Infiltration and swelling of the soft tissues are noted. With the development of the process, fluctuation appears in the centre by the second day. Toxicosis intensifies and the local process may spread for a

great distance. In severe cases the skin separates and undergoes necrosis and large defects form in the soft tissues. When the phlegmon is cut open, a cloudy serous fluid or thin pus flows out, usually together with bits of subcutaneous fat of a grey colour. Sometimes the subcutaneous fat tears away from large areas.

Treatment must be complex and applied in time. The prescription of antibiotics is guided by the severity of the child's condition and the developmental stage of the local process. In the absence of pronounced general signs of the development of sepsis, treatment can be started with intramuscular injection of penicillin in large doses (200 000-300 000 U/kg/24 hours). In graver cases agents with a broad spectrum of activity are administered by the intravenous route. Immunotherapy (antistaphylococcus gamma globulin, hyperimmune antistaphylococcus plasma, direct blood transfusion), detoxication therapy, and other forms of syndrome-correcting therapy are applied.

Local treatment consists in making numerous cuts (superficial) not only in the zone of the lesion but also at its junction with the healthy areas, without fail, and for a distance of 1.5-2 cm of the healthy surface. This reduces the swelling in the borderline zone and is a preventive measure aimed at restricting the spread of the process. The dressing is changed 6 to 8 hours after the operation. If the focus continues spreading, many small cuts are immediately made again, also on the healthy skin areas. **A** moist dressing with antiseptic or antibiotic solutions is then applied. The cuts are made under general anaesthesia or after preliminary local infiltration with an 0.25 per cent procaine solution and antibiotics.

If necrosis develops, necrectomy is performed. The wound surface which remains in such cases is gradually covered with granulations if the disease follows a favourable course, and then undergoes epithelization. Physiotherapy, exposure to laser radiation, hyperbaric oxygenation therapy, and general stimulating therapy promote the development of this process.

Mastitis Neonatorurn

Purulent mastitis develops when pyogenic infection penetrates injured skin and the lactiferous ducts. The predominant causative agent is staphylococcus. The development of mastitis is often preceded by physiological hardening of the mammary glands, which is encountered in

both girls and boys and is explained by the entry of maternal oestrogens into the child's blood through the placenta or in the breast milk. The mammary glands become larger in this case and fluid resembling colostrum appears in them. With the entry of infection, hyperaemia and later fluctuation form on the hardened and enlarged breast. Body temperature rises and the child's general condition suffers. Treatment aimed at resorption of the infiltrate is indicated in the stage of infiltration: antibacterial therapy, compresses with alcohol diluted by half or with Vishnevsky's ointment, and UHF therapy.

Radical incisions are made in the stage of abscess formation. Spread of the process on the surrounding subcutaneous fat may lead to the development of phlegmon of the chest. Cuts are made on the borderline with the healthy skin in such cases. The prognosis is usually favourable, but in extensive liquefaction of the tissue of the mammary gland in girls, further development and function of the breast are impaired.

Furuncle, Carbuncle

Furuncle is inflammation of the hair follicle, i.e. ostiofolliculitis, but as distinct from the last named not only the follicle itself is rapidly involved in the necrotic process, but also the deep layers of subcutaneous fat around it.

The most common causative agent is staphylococcus. The focus is located in places subjected constantly to microinjury, i.e. the neck, back, and the buttocks. Inadequate hygienic habits, deficient nutrition, avitaminosis, gastro-intestinal disorders, chronic infection, and diabetes mellitus are predisposing factors.

Clinical picture. The disease usually causes no marked disorders in the child's general condition, but is attended by subfebrile temperature. There are swelling, congestive hyperaemia, and tenderness of the affected area, in the centre of which a hair follicle is usually seen.

Treatment depends on the stage of the inflammatory process In prevalence of infiltration and oedema without suppuration, non operative treatment is applied: antibiotics and sulphonamides, vitamins of the B complex, compresses with ointments and alcohol diluted by half, UHF therapy, and local baths with potassium permanganate. Local procaine blocade with antibiotics has a favourable effect. If pus forms, the necrotic

core is removed. Attempts to express the furuncle are futile because of the deep involvement of the snbcutaneous fat. In addition to removing the core, linear or crossshaped incisions are made. **A** dressingwith hypertonic sodium chloride solution is applied. Surgery is also resorted to when there is still no pus but the inflammation is marked by sharp tenderness, oedema, lymphangitis and lymphadenitis, and high body temperature.A furuncle follows a particularly persistent course in patients with diabetes mellitus. The development of a furuncle on the face is a dangerous condition because in this case the process may spread to the orbit and into the cranial cavity. Children with a furuncle on the face must he hospitalized immediately for comples treatment.

Carbuncle is inflammation with simultaneous involvement of many hair follicles, the subcutaneous fat, and deeper layers of the soft tissues. **A** carbuncle forms from one or from the fusion of several furuncles.

The pathomorphological picture is that of pronounced inflammation of the follicles and the adjoining tissues with proliferation of the process to the deeper lying tissues, sometimes with involvement of the fascia. Necrotic changes predominate.

Clinical picture. The manifestations of a carbuncle are more impetuous than those of a furuncle. The local sings are a pronounced and extensive infiltration and oedema, sharp tenderness, and congestive hyperaemia. Purulent plugs are seen here, with a pyo-sanguineous fluid discharged from under them. Lymphangitis and lymphadenitis are present as a rule. The child's general condition deteriorates with a chill, increased body temperature, headache, and toxicosis even to a state of impaired consciousness and delirium.

Treatment. Surgery is the only measure: the abscess is widely opened to the distance of the healthy tissues; removal of the necrotic tissues and drainage are obligatory procedures. Dressings with hypertonic sodium chloride solution are applied. General treatment and antibiotic therapy follow the rules of treatment of acute and chronic infection. It is important to take actions against the principal disease. Carbuncle takes an especially grave and persistent course in patients with diabetes mellitus and pathological obesity.

Lymphadenitis

Inflammation of the lymph glands (lymphadenitis) is a common occurrence in children, particularly at a very young age. This is linked with the functional and morphological immaturity of the child's lymphatic apparatus (wide sinuses, a thin fine capsule of the lymph glands, increased susceptibility to infection, deficiency of the barrier function).

In children, particularly in those of 1 to 3 years of age, lymphadenitis usually develops in the mandibulo-faciale reqion (the submandibular, submental, and cervical lymph glands). The axillary, popliteal, inguinal, and cubital lymph glands are affected less frequently.

Preliminary sensitization of the organism as a result of infectious and pyogenic diseases suffered in the past plays an important role in the pathogenesis of the disease. The causes of lymphadenitis are diverse. Lymphadenitis of the mandibulo-facial region is very rarely a primary disease in children. It is usually a response of the lymph glands to some inflammatory foci. Proper identification of these causes ensures the success of further treatment.

Odontogenic lymphadenitis in children is encountered less frequently than nonodontogenic lymphadenitis. The decidnons. Less often the permanent teeth are the source of odontogenic infection. Catarrh of the upper respiratory passages, influenza. sore throat, chronic tonsillitis, otitis, exudative diaihesis, pyoderma, and injury to the skin and mucous membranes are the causes on non-odontogenic lymphadenitis mostly in very young children.

The **clinical picture** is characterized by general symptoms such as indisposition, chills, increased body temperature of 38-39°C, rapid pulse, loss of appetite, headache and disturbed sleep.

The involved lymph gland (glands) is hard, enlarged, and sharply tender to palpation. Later the disease abates under the effect of timely applied treatment or acute serous lymphadenitis develops into an acute purulent process with liquefaction of the lymph gland. The softening and accumulation of pus can 11e determined from
fluctuation felt on palpation.

Diagnostic difficulties may arise because antibiotics alter the clinical picture of the disease: body temperature drops, acute oedema and

tenderness disappear, but the lymph gland remains enlarged. The disease then follows a languid course and an abscess often develops in the gland.

The **differenfial diagnosis** has to he made wit11 specific processes in the lymph glands, systemic diseases of the blood (leukaemia, lymphogranulomatosis) and tumours. Lymphadenoactinomycosis and tuberculosis of the lymph glands are the specific forms of lymphadenitis encountered. The onset of tuberculous lymphadenitis is usually not acute and the disease follows a protracted course without high body temperature. Besides, involvement of a group of glands, sometimes in the form of a packet, is encountered more often in tuberculosis. Proper evaluation of the clinical signs, the medical history which helps in identifying the infection atrium and the acute onset of the disease allow the correct diagnosis to be established.

Affection of the inguinal glands in very young children is often mistaken for strangulated inguinal hernia. The absence of the stool, the occurrence of vomiting and general sign distinguish hernia from lymphadenitis.

Epiphyseal osteomyelitis of the femur has to be distinguished sometimes from inflammation of the deep pelvic lymph glands (high temperature, pain, flexion-adduction contracturc of the thigh).

On examination of the child a tender infiltration is felt in the pelvis above the inguinal liqament, niovements at the joint are made, though limited in range. If the clinical picture is still not clear, traction is applied to correct the contracture and a radiograph is made which shows that the hone is not affected.

Treatment. Particular care is taken to remove the primary focus of infection. Non-operative treatment includes the prescription of antibiotics, desensitizing and general invigorating therapy, and proteolytic enzymes together with physical procedures.

Purulent lymphadenitis is treated by incision. In severe lymphadenitis with toxicosis, especially in young children, the incisions are made even when the inflammatory infiltrate has still not softened. A cut 2-3 cm long is sufficient for draining the purulent cavity.

Larger incisions are indicated only in extensive adenophlegmon.

Panaris

Panaris is purulent inflammation of the fingers. According to the depth of the lesion, subcutaneous, paraungual (paronychia), thecal (whitlow), bony, and articular forms of panaris are distinguished. The causative agent is usually the staphylococcus, sometimes the streptococcus.

The inflammatory process is located for the most part on the palmer surface, but the swelling is most pronounced on the dorsal surface of the finger. This is esplained by the thicltness of the skin on the palmar surface and the presence of connective-tissue intersections stretching to the depth from the surface layers. As a consequence, the transudate spreads from the palmar surface along the lymphatic slits to the deeper lying tissues and then to the dorsal surface. The swelling usually spreads over a large part of the finger.

The inflammatory process, especially in thecal whitlow, may spread along the tendon sheath. It may extend to the forearm, into "Pirogoff's space" in a lesion: of the thumb and the little finger.

Clinical picture. Panaris is characterized by tenderness and marked swelling of the finger. Pulsed pain is often experienced, particularly with the development of a purulent process. The activity sometimes not only of the finger but also of the hand and forearm is impaired. The closer the process to the surface, the more pronounced is the hyperaemia. It is sometimes difficult to find the point of the sharpest tenderness because of the considerable swelling. G.P. Zaitsev recommends searching for this point with a bulbousend probe, carefully touching different areas of the finger with it.

Treatment depends on the stage of the process. UHF therapy, antibiotic-procaine blockade, and compresses are recommended in the infiltration stage. With the appearance of suppuration, the focus is cut open, a drain is left in it, and antibiotics are prescribed. The ungual phalanx is cut in the frontal plane, in which case injury to the tactile surface of the finger is avoided and the slits between the connective-tissue intersections are cut across. This is very important because conditions for good drainage of the slits are provided. If the process is located on the other phalanges, longitudinal incisions are made on both sides of the finger without fail and drains are introduced into them.

In thecal whitlow, in which the process acquires the character of tendovaginitis, drainage with irrigation of the tendon sheath is advisable for rapid arrest of the inflammation. Bony panaris is considered as osteomyelitis of the finger phalanx.

Haematogenic osteomyelitis

Osteomyelitis is acute purulent inflammation of the bone marrow, though practically the process spreads to all the morphological structures of the bone, i.e. panostitis develops. The surrounding soft tissues are also involved in the process quite frequently.

The origin of haematogenic osteomyelitis is associated with penetration of the microbes into the marrow by way of the blood channels; local inflammation is therefore preceded by bacteraemia. If the immunological properties of the macro-organism are disturbed, the local process may he the source of sepsis and septicopyaemia.

Acute haematogenic osteomyelitis occurs predominantly in children.

According to T.P. Krasnobaev, children account for 75 per cent of cases. It is encountered mostly in children over 5 years of age. The incidence among boys is two to three times that among girls. Long tubular bones active in growth are the favoured site of osteomyelitis (in more than 70 per cent of cases).

Acute and chronic stages of osteomyelitis and atypical forms are distinguished.

Acute Haematogenic osteomyelitis

Haematogenic osteomyelitis is caused by pyogenic bacteria, but the main causative agent is the staphylococcus (up to 90 per cent of cases) or associations of staphylococcus with *Escherichia coli, Proteus,* and *Pseudomonas aeruginosa.*

The body is infected and the bacteria invade the blood through injured skin and mucous membranes, and the lymphoid throat ring. Pyoderma of the skin, inflammation of the nasopharynx, and latent infections are of definite importance.

The umbilical wound is a frequent infection atrium in infants. In some cases osteomyelitis results from the spread of a purulent process to the bone from the adjoining soft tissues or other organs (odontogenic osteomyelitis associated with dental caries, osteomyelitis of the rib

consequent upon pyothorax, osteomyelitis of the fingers after panaris, etc.).

The anatomical age features of the structure and blood supply of the bones play a significant role in the development of osteomyelitis in children: the richly developed network of blood vessels; the autonomous supply of blood to the epiphysis, metaphysis, and diaphysis; the presence of a great number of small vascular branchings stretching radially through the epiphyseal cartilage to the ossification nucleus. The epiphyseal system of blood supply prevails in children under the age of 2 years, the metaphyseal system begins developing after this age. The epiphyseal and metaphyseal systems are isolated but there are anastomoses between them. The common vascular network forms only after ossification of the epiphysis.

Affection of the epiphyseal zone is characteristic of children under the age of 2-3 years. With age, when the system of blood supply to the metaphysis begins developing intensively, it is the metaphysis that predominantly becomes affected.

Pathogenesis

The pathogenesis of acute haematogenic osteomyelitis is unclear to date. An important specific feature of this inflammation is that it is closed in by the rigid walls of the bone tube, as a result of which first the veins and then the arteries are compressed. Pain, which is a consequence of hypertension in the marrow cavity, is indirect proof of this interpretation of the circulatory disorders in the bone. Intraosseous pressure in acute osteomyelitis reaches 300-500 mm water (normal value in healthy children, 60-100 mm water).

If the osteomyelitic process is not recognized when it is still in the stage of inflammation within the boundaries of the bone-marrow cavity, then beginning from the 4th or 5th day of the disease the pus spreads along the bony haversian and Volkmann's canals under the periosteum and gradually separates it. Later (the 8th to 10th day and later) pus and the products of disintegration continue separating the periosteum, then the pus breaks through into the soft tissues and forms intermuscular and subcutaneous phlegmons. This applies to neglected cases of osteomyelitis with delayed diagnosis, the management of which is very difficult. In

spontaneous rupture of a subperiosteal abscess into the surrounding tissues pain is relieved as a rule because pressure in the bony tube drops.

Clinical picture and diagnosis

The clinical manifestations and severity of acute osteogenic osteomyelitis in children are very diverse and are determined by many factors: body reactivity, microbial virulence, the patient's age, the site of the affection, the term of the disease, and the preceding treatment. The degree of the sensitization processes is very important. A violent general reaction of the body, rather similar to anaphylactic shock, comes to the forefront if osteomyelitis develops in the maximum hyperergic phase of immunogenesis in a sensitized body. In other cases the general manifestations are less pronounced.

In accordance with the circumstances mentioned, three main forms of acute haematogenic osteomyelitis are distinguished: toxic (adynamic), septicopyaemic, and localized.

The ***toxic (adynamic) form*** follows an extremely violent course with signs of endotoxic shock. **A** state of collapse is observed as a rule, with loss of consciousness, delirium, high body temperature (up to 40-41°C*)*, and sometimes with convulsions and vomiting. Dyspnoea is found but without any clear clinical picture of pneumonia. The cardiovascular abnormalities include disorders of central and peripheral circulation, reduced arterial pressure, with the development within a short time of cardiac insufficiency and signs of myocarditis. Punctate extravazations are often seen on the skin. The tongue is dry and with a brownish coating. The abdomen is usually distended and tender in the upper parts. The liver is enlarged.

In view of the presence of general clinical symptoms of severe toxicosis, it may he difficult to idenlify the local manifestations of the disease, and the more so the exact location of the primary lesion of the bone. The local focus can be detected only some time later, when the general condition improves. Moderate swelling of the affected region, pain contracture of the adjacent joint, increased local temperature and, sometimes, an abnormally pronounced pattern of the subcutaneous veins are found in such cases. The discovery of such changes indicates the need for diagnostic, puncture of the suspected focus of affection. Increased intraosseous pressure can be recorded in acute haematogenic osteomyelitis,

although perforation of the bone almost always fails to show pus in the bone-marrow cavity.

Despite the application of massive infusion therapy, including broad-spectrum antibiotics and immunopreparations, and even the performance of operation on the focus of affection, fatal outcomes in this form of acute osteomyelitis are often encountered to date.

The ***septicopyaemic form*** of acute haematogenic osteomyelitis is marked by general septic phenomena which are also quite pronounced. The bone lesion, however, can be detected much earlier in this group of patients. The onset of the disease is also acute: body temperature rises to a high level (39-40°C), signs of toxicosis increase, and the activity of vital organs and systems is disturbed. Confused consciousness, delirium, and euphoria are sometimes encountered. Pain is experienced in the affected limb from the first days of the disease and becomes very intense due to the development of intraosseous hypertension. Septic complications caused by the spread of the purulent foci to various organs (the lungs, heart, and kidneys, as well as to the other bones) often occur.

The ***localized form*** of acute haematogenic osteomyelitis is characterized by the predominance of local signs of purulent inflammation over the general clinical manifestations of the disease. Atypical forms of osteomyelitis should also be related to this group.

The onset of the disease in typical cases is quite acute. Sharp pain appears in the limb when the general condition seems to be normal. Older children usually indicate the most tender place exactly. The child tries to keep the involved limb in a definite position because any movement increases the pain. The ligaments and the periarticular tissues are involved in the process if the focus is close to the joint. This leads to marked and stable contracture of the joint. Body temperature increases from the onset of the disease and remains high later (within the range of 38-39°C)**.** The child's general condition grows worse rapidly, he loses appetite, thirst increases, which is eviclence of the development of toxicosis.

The first signs of inflammation are seen on examination of the involved limb; swelling in the region of the lesion, complete massive infiltration of the tissues, and pronounced venous pattern of the skin. The main constant local signs of osteomyelitis are as follows: sharp local tenderness to palpation and particularly to percussion over the site of the

lesion. Oedema and tenderness extend also to the adjoining areas. Such signs as hyperaemia of the skin and, in particular, fluctuation in the region of the lesion are very late signs and are evidence of neglected osteomyelitis.

Considerable diagnostic difficulties arise in osteomyelitis of bones forming the hip joint. The local signs are indistinct on the first days of the disease due to the powerful muscular casing in this region. On careful inspection it can be seen that the lower limb is slightly flexed at the hip joint; abduction and mild external rotation are also noted. Movements at the hip joint are painful. The joint itself and the overlying skin are moderately oedematous.

Osteomyelitis of the iliac bone and vertebrae takes a very severe course. Toxicosis and high fever are present from the onset of the disease. Oedema is seen on inspection, and palpation and percussion reveal the sharpest tenderness at the site of the focus of affection.

Diagnostic puncture of the bone with subsequent cytological examination of the aspirated material should be carried out more extensively in questionable cases. Measurement of intraosseous pressure is very important in establishing the early diagnosis of acute haematogenic osteomyelitis. The discovery of intraosseous hypertension confirms the diagnosis even in the absence of pus under the periosteum and in the marrow cavity.

Radioisotope bone scanning and subsequent computer treatment of the findings are lately used for earlier and more exact determination of the location and extent of the inflammatory process. Short-lived isotopes possessing bone tropism (technetium) are used for the purpose.

Blood tests show leykocytosis (up to 30 000-40 000 per mm3) with a shift of the differential count o the left and toxic neutrophil granulation. The ESR is markedly increased (up to 60 mm/hour) and remains high for a long time.

Marked changes are found in the blood serum protein spectrum. These are dysproteinaemia, an increase in the globulin fractions, and the development of hypoalbuminaemia. Anaemia caused by bone marrow inhibition by the prolonged effect of toxins develops in a persisting and severe disease.

Disorders of the blood coagulation system are also found (the fibrinogen concentration and the fibrinolytic activity increase, the recalcification time and the coagulation time become shorter, the prothrombin index increases).

The X-ray signs of acute haematogenic osteomyelitis are manifested no earlier than on the 14th-21st day of the disease. The earliest signs of osteomyelitis can be detected on a good structural radiograph. Reduced density of the bone shadow and blurring of its contours are usually found, osteoporosis in the region corresponding to the zone of the inflammation can also be detected. The spongy substance of the bone produces a macromacular pattern due to resorption of the bony trabeculae and merging of the intertrabecular spaces as the result of intensified resorption. Later, the destructive cavities become wider and the contours of the cortex become loosened, indistinet, and uneven. Linear periostitis is the most authentic sign. The periosteal reaction is usually extensive and is seen as a thin band, sometimes as a veil-like shadow, stretching next to the cortex. Its degree is determined by the location of the focus. The periosteal reaction is most pronounced in a diaphyseal lesion, less so in a metaphyseal affection, and still less pronounced in a lesion of the epiphysis.

With further development of the inflammatory process, the bone undergoes necrosis and lysis and is replaced by pus and granulations. These changes first appear in the metaphysis, as a rule, and the process gradually spreads to the diaphysis

The **differential diagnosis** usually has to be made between acute haematogenic osteornyelitis and the articular form of rheumatism, phlegmon, tuberculosis of the bones, and injury.

Rheumatism is characterized by shifting pains in the joints and typical changes in the heart confirmed by electrocardiography. Careful inspection and palpation of the involved region reveals that in rheumatism, in contrast to osteomyelitis, tenderness and swelling are mainly localized over the joint and not over the bone. Improvement of the local process with the prescription of salicylates is animportant factor. A phlegmon can also producc a clillical picture resembling that of osteomyelitis. Hyperaemia ant1 superficial fluctuation appear much earlier in a phlegmon than in osteomyelitis. A contracture may develop if the phlegmon occurs close to a

joint. In distinction from that in osteomyelitis it is less slable and is usually corrected by careful passive movements. The final diagnosis can he established only after an incision in some cases.

The differential diagnosis with typical forms of tuberculosis of the bones is easy. Tuberculosis of the bones is now a rare occurrence and is characterized by a gradual onset. Though experiencing pain in the limb, the child still uses it. Alexandrov's sign (thickening of the skin fold on the involved limb) and muscle atrophy are found. The radiograph demonstrates osteoporosis (the "melting sugar" symptom) and an indistinct periosteal reaction. This reaction, however, may be clearly pronounced in mixed infection and in accompanying ordinary flora. The so-called acute forms of osteoarticular tuberculosis are actually cases of delayed diagnosis made when pus has already penetrated the joint. In addition to the X-ray picture, identification of the specific causative agent in material aspirated from the joint helps in establishing the correct diagnosis.

Acute haematogenic osteomyelitis sometimes has to be differentiated from bone injury. A carefully taken medical history, the absence of septic manifestations, and the findings of X-ray examination are important in such cases. Subperiosteal fractures present difficulties at times. A radiograph retaken in 6 to 8 days, however, demonstrates a fine bone callus on a restricted area.

Treatment

Complex management of osteomyelitis, substantiated in the past by T.P. Krasnobaev, is now extensively applied. It is based on three main principles: (1) producing an effect on the macroorganism; (2) producing a direct effect on the causative agent; (3) timely and adequate treatment of the local focus.

1. The effect exerted on the macroorganism is aimed at relieving completely the severe toxicosis and correcting the disturbed homeostasis.

Active detoxication therapy comprises administration of a 10 per cent glucose solution with insulin, haemodes, polyglucine, aminophylline, and native plasma. Calcium preparations, diphenylhydramine hydrochloride, chloropyramine or pipolphen are prescribed for desensitization of the organism and normalization of vascular and tissue permeability. To improve specific immunity, passive immunization of the child's organism is conducted in the acute period of osteomyelitis by hyperimmune

staplylococcus plasma and antistaphylococcus gamma globulin. With the abatement of acute phenomena, active immunization (stapliylococcus toxoid) is prescribed to induce the production of the patient's own specific antibodies.

Electrolyte metabolism, acid-base equilibrium, and the function of the urinary system must be controlled during intensive therapy. Measures regulating protein and carbohydrate metabolism are prescribed. Treatment also includes stimulation of the organism's defence forces (transfusion of blood no less than 4 or ***5*** times, no more than 100 ml each time).

Adrenal cortical activity is inhibited in severe forms of the disease. Hormonal preparations (hydrocortisone or prednisolone) are given in short cycles (up to **7** days).

2. A direct effect is produced on the causative agent with broadspectrum antibiotics. Combination of intravenous and intraosseous administration is the most effective means. The efficacy of antibacterial therapy considerably increases when it is combined with proteolytic enzymes. For the next course of treatment antibiotics possessing bone affinity (lincomycin) are prescribed in age doses for 2 or 3 weeks. Antibiotics are discontinued when body temperature returns to normal, the inflammatory reaction in the focus disappears, and the blood test values show a tendency to normalization.

3. Timely and adequate treatment of the local focus. In view of the fact that most severe forms of osteomyelitis are consequent upon intraosseous hypertension, early surgical intervention, osteoperforation, acquires primary importance. An incision, no less than 10-15 cm in length, is made in the soft tissues overlying the lesion and the periosteum is cut longitudinally. Two or three perforating openings 3-5 mm in diameter are made at the junction with the healthy bone. Pus is usually discharged under pressure in such cases, while in a disease of a long duration the contents of the marrow cavity may be seropurulent for two or three days. Pus may also be detected in the subperiosteal space (subperiosteal abscess) when patients are

admitted to the hospital at later terms (5th-6th day).

The marrow cavity is irrigated with 1:5000 nitrofurazone solution and antibiotics through the perforation in the bone.

Bone dialysis is conducted for the first 2 or 3 postoperative days in severe cases of osteomyelitis by continuous interosseous drip of solutions containing antiseptics and antibiotics (kanamycin, monomycin).

The pain syndrome is relieved considerably or completely after osteoperforation. For the time that the child is kept in bed there is no need to immobilize the involved limb in such cases. On the contrary, early movements in bed contribute to improvement of circulation and complete functional restoration of activity of joints adjoining the inflammatory focus.

The expediency of immobilization is determined from the dynamics of X-ray changes in the diseased bone. A deep plaster splint is applied if obvious signs of bone destruction are demonstrated. The need for early complex treatment of haematogenic osteomyelitis in the acute stage must be emphasized especially. Only such measures can prevent the change of an acute process to a chronic condition.

Mataepiphyseal Osteomyelitis

Metaepiphyseal osteomyelitis is mostly encountered among infants, predominantly among the newborn. By the haematogenic route the infection (usually staphylococcus) enters the bone metaphysis and the inflammatory process develops here. Due to the peculiar blood supply of the metaepiphyseal junction in very young children, however, the inflammation spreads to the growth zone and epiphysis
located in the joint. As a result, the main clinical symptoms are caused by the developing acute arthritis.

Clinical course

Metaepiphyseal osteomyelitis sets in acutely as a rule with a rise of body temperature, debility, refusal of food, reluctance to move the involved limb which the child holds in a forced position.

Examination reveals swelling over the zone of affection, deformity of the adjoining joint, increase of local temperature. Hyperaemia appears later. Palpation and passive movements of the limb cause sharp pain. The "pseudoparesis" symptom is encountered (the hand or foot of the involved limb hangs and movements in it are sharply limited). The local form of osteomyelitis may be complicated by phlegmon of the soft tissues around

the joint. In some cases, osteomyelitis is one of the septicopyaemic foci in sepsis and can be multiple.

In infants with osteomyelitis. Morphological examination shows predominance of necrotic processes i n the bone which are attended by death and resorption of bone tissue in the metaphysis and damage to the growth zone and articular surfaces. However, reparative processes are well pronounced at this age as a result of which this form of osteomyelitis does not develop into a chronic process. The late functional disorders may be considerable and are explained by impaired growth of the limb and deformities of joints.

The X-ray signs are demonstrated earlier in metaepiphyseal osteomyelitis than in the other forms. Some characteristic signs can be detected as early as the 8th-10th day: thickening of soft tissues on the affected side, widening of the X-ray joint space, a fine periosteal reaction. Foci of destruction in the metaphysis are demonstrated on the radiographs only on the 3rd week after the onset of the disease, whereas the degree of destruction of the bone epiphysis and growth zone can be judged authentically only after the appearance of the ossification nucleus. The distal and proximal ends of the femur and the proximal ends of the humerus and tibia are affected most frequently.

Treatment

The treatment of metaepiphyseal osteomyelitis is marked by specific features. General treatment is applied according to the principles of management of an acute pyogenic surgical infection with due consideration for the child's age. Broad-spectrum antibiotics possessing affinity to bone tissue (lincomicin), detoxication therapy and immunotherapy are prescribed.

Immobilization plays an important role: Schede's traction is applied to the lower limb and Desault's bandage to the upper limb. Neither surgical manipulations in the region of the growth zone nor plaster splints are applied in the newborn. Cases with an obvious clinical picture of arthritis are managed by puncture of the joint, removal of the exudate, and injection of antibiotics into the joint.

In location of the process in the proximal femoral epiphysis, spreader-bandages are applied after the acute inflammation abates to prevent pathological dislocation of the hip. After recovery from acute

haematogenic osteomyelitis the child must be kept under regular observation of an orthopaedist or surgeon.

Chronic Osteomyelitis

With abatement of the acute phenomena after the purulent focus has been opened and the appropriate treatment applied, the inflammatory process in the bone may change to a subacute stage characterized by the beginning of reparation. The X-ray picture in this stage has the same signs as that in the acute stage but the periostitis is grosser and contains elements of sclerosis, laminated in some cases. The spots of diminished density and the shadows of the separating bone fragments are demonstrated more clearly. The beginning of sclerosis of the bone itself is seen. The subacute stage terminates in recovery or develops into the chronic stage.

If the process fails to abate completely in 4 to 6 months, regular exacerbations occur, fistulae remain, and the discharge of pus continues, then it is considered that osteomyelitis has taken the chronic stage. This outcome depends on the severity and rate of the occurring alternative changes in the bone tissue and how early and properly is the treatment applied. A change to the chronic stage may be encountred in 10 to 30 per cent of cases.

Chronic osteomyelitis is marked by a prolonged course with remissions and periods of deterioration. Typical forms are characterized pathomorphologically by pieces of necrotic bone (sequestra), a sequestral cavity, and sequestral capsule (involucrum). Granulations and pus are usually present between the involucrum and the sequestrum. The sequestra vary greatly in size, from very large ones when almost the whole bone is necrotized, to small pieces a few millimetres in length, and may be solitary or multiple. Cortical sequestra formed due to necrosis of only the compact layer, and central sequestra arising from the depth of the bone also exist. The involucrum is formed by the peri- and endosteum and thickened bone tissue with dense trabeculae arranged at random. In some cases the involucrum may be larger in diameter than the bone when healthy. The formation of the involucrum and demarcation of the sequestrum may be represented as follows. Granulations begin growing from the periosteum and bone marrow around the lesion. They stimulate bone formation and impair the union between the necrotic areas and the healthy tissue. A

sequestrum forms. The nearer to the focus of necrosis, the more is the number of granulations mixed with pus. The thickened periosteum and the endosteum are later ossified and form the lateral walls of the hard capsule around the dead bone area. The perifocal sclerosis and thickening of the bone tissue usually leads to demarcation of the necrotic focus.

After the sequestrum forms the inflammatory process continues. Pus collecting in the focus is discharged through the fistulae from time to time. Small sequestra are sometimes discharged, especially in a disease of a long duration. In such cases large sequestra may break into small ones. Sharp eburnation of bone (sclerosis and hardening) occurs around the focus of chronic inflammation. The soft tissues are also sclerosed, nutrition is disturbed, and the muscles atrophied. In a severe and extensive process the periosteum may be destroyed. Bone regeneration is greatly delayed in such cases and the involucrum fails to form or is deficient as a result of which pathological fracture or pseudoarthrosis often forms.

Clinical picture

Chronic osteomyelitis is characterized by a protracted course with remissions and exacerbations. The fistulae may close during a remission. In exacerbation, body temperature increases, tenderness and toxicosis intensify. Pus is again discharged from the fistulae, sometimes in abundance.

Examination of the patient reveals oedema of the soft tissues and sometimes a swelling of the limb on the level of the lesion. Fistulae and scars in places of previously existing fistulae are typical of chronic osteomyelitis. Palpation of the limb usually causes only mild tenderness and often reveals atrophy of the soft tissues and thickening of the bone. Pallor of the skin and signs of malnutrition are also found. Body temperature is subfebrile, particularly in the evening, but sometimes reaches high levels during exacerbation.

The X-ray diagnosis in typical cases with chronic osteomyelitis is quite easy. Radiographs show areas of osteoporosis and those of pronounced osteosclerosis. The involucrum containing sequestra, usually clearly outlined, is seen.

Chronic osteomyelitis has to be differentiated from other diseases in some cases, namely, from tuberculosis and sarcoma. In contrast to osteomyelitis, tuberculosis sets in gradually, with no high temperature.

Atrophy and contracture of the joint occur early. The fistulae are usually connected with the joint and have flacid and glass-like granulations. Processes of osteoporosis prevail on the radiograph and there are neither large sequestra (the sequestra seen usually resemble melting sugar) nor pronounced periostitis. Restoration of bone trabeculae (which at first are tangled) imperceptibly continuous with the normal tissue and diminution of osteoporosis are seen in the stage of reparation.

Ewing's tumour (sarcoma) follows a wave-like course. Body temperature rises and pain increases during an attack. The diaphyses of the long tubular bones are involved in the process most often. The X-ray picture of this tumour is characterized by a bulbous contour on a localized area of the diaphysis, scattered macular osteoporosis, cortical osteolysis without sequestration, and narrowing of the marrow cavity. Osteogenic sarcoma is marked by the absence of a zone of sclerosis around the focus, by separation of the cortex and periosteum in the form of a peak, and by "spicles" (spicular periostitis).

It is often very difficult to differentiate osteoid osteoma from osteomyelitis. This tumour is characterized by a clearly demonstrated band of perifocal thickening of trabeculae around the focus of diminished density and extensive periosteal deposits in the absence of marked destruction. Severe night pain in the involved bone is typical of osteoid osteoma. In some cases the diagnosis is established only with the aid of biopsy.

Treatment

Treatment in chronic osteomyelitis comprises trephination of the bone, removal of the sequestrum (sequestrectomy), and curettage of the purulent granulations. A wide incision is made in the skin and superficial fascia overlying the lesion with excision of the fistulae. The muscles are usually divided by the blunt techniques. The periosteum is cut and separated from the bone with a raspatory. The sequestral cavity is opened by removing part of its bony wall with a chisel or ultrasonic knife. The sequestrum and pus are removed and the granulations scraped away with a sharp spoon. The remaining cavity is then treated with iodine and alcohol and filled with antibiotics, and the wound is sutured layer by layer.

The "antibiotic-blood" filling is recommended for the remaining cavity. It is prepared by mixing a sufficient volume of the patient's blood

with antibiotics and allowing time for the clot to retract. The filling is prepared extempore during the operation. The cavity is then filled with the clot. With this technique the discharge from the operative wound in the postoperative period is less. Recurrences are possible after this operation due to a number of causes: non-radical operation (pus and granulations left in the cavity, necrosis and sequestration of the walls of the remaining cavity as a consequence of their deficient nutrition) and accumulation of wound detritus, clots, and thin blood in the remaining cavity. All these remnants may become infected.

“Trough” resection of the bone is therefore advisable in an extensive lesion. With this type of resection the possibility of sequestration of the overhanging bone edges is less, whereas the soft tissues adjoining closely the surface of the bone improve its nutrition. Besides, the sequestral cavity is removed. "Trough" resection provides for good inspection of the bone and for performing the operation within the limits of healthy tissues. Recurrences are much less frequent after this operation. In large bone defects forming as the result of sequestrectomy, continuous drip irrigation of the cavity with solutions of antiseptics or antibiotics (1 : 5000 nitrofurazone solution, up to 300 ml, with antibiotics-aminoglycosides) for 7-10 days after the operation is expedient.

Atypical Forms of Osteomyelitis

Primary-chronic osteomyelitis occurs when the organism's immunobiological properties are highly developed as a consequence of which the focus is rapidly delimited. It should be emphasized, however, that the name "primary-chronic" osteomyelitis is at fault because the acute stage of these forms exists but usually remains unrecognized due to the poor clinical manifestations.

The atypical forms of osteomyelitis include Brodie's abscess, Garres sclerosing osteomyelitis, Ollier's albuminous osteomyelitis, and osteomyelitis antibiotica.

Brodie's abscess is marked by a protracted course, mild aching pains in the region of the lesion, and moderate increase of temperature. The proximal tibial, distal femoral, and proximal humeral metaphyses are the favolired sites. It can be seen on examination that the limb is moderately swollen and mildly tender to intense palpation. X-ray shows a round zone

of destruction with pronounced perifocal sclerosis. Sequestra and fistulae do not usually form. A band of diminished density, a "strip" connecting the focus with the growth zone, can often be seen.

Treatment is surgical, consisting in trephination of the bone, removal of the purulent granulations by curettage, and filling tightly the cavity of the bone abscess with antibiotics.

The clinical picture of the ***sclerosing forms*** is similar to that of Brodie's abscess. Radiologically they are manifested by marked bone eburnation and sclerosis of the marrow cavity. Sometimes small foci of diminished density are seen against the sclerotic background; these are cavities with pus, granulations or small sequestra.

Treatment consists in intense antibiotic therapy and physiotherapy (UHF therapy). Foci with sequestra or purulent granulations are curettaged with the performance of marginal bone resection.

Ollier's albuminous osteomyelitis. This is a very rare disease. The clinical manifestations are similar to those of other forms of atypical osteomyelitis though in some cases they are more pronounced. The bone is sclerosed and the marrow canal, which contains white or yellow fluid, is narrowed.

Treatment consists is trephination of the bone with removal of albuminous fluid and tight filling of the cavity with antibiotics.

The so-called **antibiotic osteomyelitis** (osteomyelitis antibiotica) may be encountered in children previously treated with antibiotics and in cases of early but insufficiently effective antibiotic therapy. The clinical picture and X-ray siqns of this form of osteomyelitis differ from the typical patterns. A number of specific features of antibiotic osteomyelitis can be distinguished:

(1) negligible exudation and destruction, the inflammatory process is delimited, only localized necrosis develops;

(2) mild clinical picture: absence of marked temperature and leukocytic, reaction, a tendency of the process to develop into a chronic stage of the type of "primary-chronic" osteomyelitis (though fistulae may form) is noted; in epiphyseal localization, the number of cases with the development of purulent arthritis decreases;

(3) the X-ray picture shows a macular pattern of the bone, the formation of small cavities, a mild periosteal reaction (in some cases it is absent),

pronounced early sclerosis of the bone and a tendency towards resorption of small sequestra.

The abortive course and the unclear X-ray picture often make it very difficult to differentiate antibiotic osteomyelitis from osteoid osteoma, osteolytic sarcoma, eosinophilic granuloma, and other diseases. Despite the obliterated clinical manifestations, antibiotic osteomyelitis must also be treated by intense and complex measures.

Consequences of chronic haematogenic osteomyelitis. Preamyloid states and amyloidosis of the internal organs must always be borne in mind in chronic osteomyelitis, particularly if it is of a long duration. Immunoelectrophoresis is conducted in diminislied concentration activity of the kidneys, proteinuria, anaemia,
stable subleukocytosis, rod-nuclear shift, and lymphopenia. This makes it possible to diagnose a preamyloid state in time and begin the appropriate treatment.

Among the local complications encountered are pathological fractures and dislocations, false joints, deformities, and disturbed bone growth.

Principles of regular medical observation and completing the cure of children with haematogenic osteomyelitis. Haematogenic osteomyelitis requires prolongetl, persistent treatment applied stage by stage and at regular intervals. Disability among children, caused by osteomyelitis, can be reduced and severe sequelae avoided only if these principles are insistently put into practice. Several successive stages in the treatment of osteomyelitis are distinguished: (1) in the acute stage; (2) in the subacute stage; **(3)** in the chronic stage; (4) in the stage of residual phenomena.

After recovery from the acute period and discharge from the hospital, the child is registered at the local out-patient clinic and examined clinically and by X-ray at least once in 2 months for 6 months. The following measures must be applied in the subacute stage: (1) second immunization by the staphylococcus toxoid; (2) UHF therapy (up to 15 sessions); (3) desensitization therapy (14 days); (4) antibiotic therapy (14days); (5) treatment with anabolic hormones (21 days); (6) protein diet; (7) careful exercising of active and passive movements at the joints.

If the subacute stage does not develop into the chronic stage, the therapeutic effect is reinforced by conducting these courses of treatment for 12 months. Treatment at a spa is recommended (in the USSR in the Crimea, Northern Caucasus, and Central Asia).

If the process changes to the chronic stage, the above described treatment is continued and should an operation become necessary the patient is hospitalized. The main object of treatment at this stage is to improve the immunobiological resistance of the patient's organism and remove radically and completely the inflammatory process and the complications induced by it.

After discharge from the hospital particular care is taken to restore the activity of the limb (therapeutic exercises and thermal procedures under cover of antibiotic therapy). Spa treatment is also indicated in this stage.

Paraproctitis

Paraproctitis, inflammation of the connective tissue around the rectum and anus, may be acute or chronic. In childhood it is mostly encountered in the neonatal period and in the first months of life. Pus cultures usually produce association of *Escherichia coli* with staphylococcus or streptococcus. The infection in most cases gains entrance through the rectal mucosa. Evidence of this are persistent fistulae formed after the abscess is opened and the frequently found openings in the anal crypts communicating with the pararectal fat.

The predisposing factors in children are microtrauma of the rectal mucosa and diseases of the skin in the region of the perineum and anus (maceration, fissures, and the presence of congenital pararectal fistulae and long sacciform crypts).

Microtrauma of the rectal mucosa is often encountered in constipation, diarrhoea, and some disorders of digestion. Faecal particles and pieces of undigested food are retained in the anal crypts and injure the mucous membrane. In diarrhoea, particularly if attended by frequent tenesmus, harder particles of faeces also cause microtrauma of the anal crypts. Finally, considerable distention of the rectum by the faecal material may also cause microcracks. An aggravating factor is the increased tonus of the sphincter ani, when favourable conditions are created for prolonged retention of firm intestinal contents and increase of intrarectal pressure.

The mucosa may also be damaged by the tip of the enema or by foreign bodies, and also as the result of injury to the perineum, though such cases are rare among children.

In some cases, paraproctitis develops from a congenital pararectal fistula when secretions accumulate in the fistula which suppurates with involvement of the surrounding subcutaneous fat. Recurrences are characteristic of congenital fistulae.

The incidence of paraproctitis is higher among boys. The lower incidence among girls can be explained by the greater elasticity and resilience of the pelvic floor as a result of which pressure in the rectum is less.

Clinical picture

The clinical picture of acute paraproctitis is determined by the location of the focus. The deeper is the abscess located, the more severe are the general disorders and the disturbed functions of the pelvic organs. Deep acute paraproctitis is a very rare occurrence in children. The abscess is usually subcutaneous or submucous; ischiorectal paraproctitis sometimes occurs. The disease has an acute onset with a rise in body temperature to 38-39°C sometimes with a chill, and severe throbbing pain in the anal region. Defaecation is painful, particularly when the process develops under the mucous coat. The younger the patient, the more frequently retention of stool and micturition due to the pain reflex are encountered.

Swelling, congestive hyperaemia, and sharp tenderness to palpation are the local signs. Fluctuation appears in suppuration.

When the focus is located in the pelvirectal and ischiorectal fat the disease begins with a feeling of heaviness and dull and not very intensive pain in the region of the pelvis or in the depth of the perineum. These signs intensify progressively. Reflex pain and disturbed functioning of the pelvic organs, particularly in a case with a pelvirectal abscess (pain experienced during micturition, ischuria paradoxa, tenesmus) are usually pronounced. Sharp disorders of the patient's general condition are characteristic. Toxicosis increases rapidly and fever and chill appear. It is sometimes difficult to make the diagnosis of a deeply located abscess. The discovery of oedema of the buttocks at the site of an ischiorectal abscess sometimes helps in recognizing the lesion. The clinical picture of pelvirectal paraproctitis may resemble that of acute appendicitis.

Rectal examination plays an important role in the diagnosis of deep pelvic abscesses. When the process develops in the ischiorectal space, digital examination reveals tenderness and thickening **(**sometimes fluctuation and swelling) of the rectal wall. A pelvirectal abscess is marked by a high level of the infiltration, fluctuation, and tenderness.

Deeply located paraproctitis is differentiated from complicated dermoid cyst, epithelial coccygeal fistulae, and osteomyelitis of the pelvic bones.

Treatment

The abscess is opened and examined without fail so as to remove pus from the pockets and leaks, and a drain is introduced. Physiotherapeutic procedures (UHF therapy), warm half baths with potassium permanganate, and antibiotic therapy are prescribed. Intensive treatment according to the principles of the management of acute surgical infection is indicated in deeply located paraproctitis.

Chronic paraproctitis. Pararectal fistulae. Chronic paraproctitis is characterized by a persistent and protracted course and may result from the development of an acute process into a chronic one or may arise from congenital pararectal fistulae. Fistulae can be complete or incomplete, depending on whether they communicate with the rectum and skin or open only into the rectum or on the skin. The persistent course is explained by the tortuous character of the fistula which gives off branches and by its communication with the rectal lumen. The fistula is usually lined with mucous epithelium which secretes fluid. This is the cause of the exacerbations occurring from time to time.

Congenital pararectal fistullae, which are evidently remnants of the tail gut, are of special interest in pediatric practice. Congenital fistulae are short as a rule, usually incomplete and mildly tortuous. The fistula is sometimes detected as a firm cord in the pararectal fat. Fistulae can be intra-, trans-, or extrasphincteral according to their relation to the sphincter ani (which is very important in operative management).

The **clinical picture** of chronic paraproctitis depends to a great measure on the character of the fistula. An incomplete fistula communicating with the skin is closed with a film. Mucous content accumulating in the fistula breaks through to the outside. Quite often this fluid becomes infected and signs of inflammation with involvement of the

pararectal fat appear. Incomplete fistulae communicating with the rectal lumen may be asymptomatic. Such fistulae are rapidly infected, however, and signs similar to those of acute paraproctitis occur.

Complete fistulae are manifested early, often on the first weeks and months of the child's life. Depending on the size of the lumen of the fistula, only mucus and pus or pus and gases are discharged from it; faeces may be discharged from a fistula with a large diameter.

Probing of the fistula with simultaneous digital control through the rectum helps in determining the character of the fistula. If the lumen of the fistula is narrow. it is advisable to perform radiofistulography or the colour test in which the appearance in the rectum of a dye administered through the skin opening of the fistula is watched.

Treatment in pararectal fistulae may be non-operative and surgical.

Non-operative treatment comprises the prescription of a diet poor in waste products, half baths with potassium permanganate and other antiseptics, and control of the stool. Curettage of incomplete fistulae communicating with the skin is effective in some cases. Sclerosing therapy is carried out: 0.4-0.5 ml of a 10 per cent silver nitrate solution or 3 per cent iodine tincture is injected into the fistula. The injection is repeated once a week for 5-6 weeks. X-ray therapy applied in some adult patients with chronic paraproctitis is not used in children.

Surgery is undertaken when non-operative treatment is ineffective. The character of the intervention is determined by the relation of the fistula to the sphincter ani. Intrasphincteral fistulae are cut and then curettaged. Gabriel's operation in which cutting of the fistula with triangular resection of the skin can be performed. The abnormal tissues around the fistula are removed as a result; the edges of the wounds do not stick so that the percentage of recurrences is less. Napkins with Vishnevsky's ointment are applied after the fistula is cut and Gabriel's operation conducted; the napkins are changed regularly. Warm antiseptic half baths or washing of the perineal region are prescribed from the third postoperative day.

In an incomplete skin extrasphincteral fistula an incision is made in the skin around its opening into which a probe is advanced, and the fistula is then excised. A congenital fistula may have no lumen in its upper part, in which case a firm cord is palpated which must be resected. Broad-spectrum antibiotics are administered into the wound and sutures are applied. A

tampon with Vishnevsky's ointment is introduced into the wound canal if the tissues around the fistula were grossly changed or the operation mas rather injurious.

Complete extrasphincteral fistulae are excised, the edges of the rectal mucosa freshened and sutures applied to them. A sanious discharge from the wound canal is usually encountered in the postoperative period. To avoid infection of the pararectal fat and a recurrence, insertion of a tampon with Wishnevsky' ointment into the wound canal is advisable, as it was suggested by A.N. Ryzhikh.

The operative intervention in trans-sphincteral fistula is more complicated, especially in the case of a complete fistula. The fistula is exposed and excised except for the part passing through the fibres of the sphincter. The intrasphincteral part is scraped with a sharp spoon. If the fistlila is located on the anterior or posterior wall the sphincter may be cut. In addition to the radical operation, cutting of the sphincter in these places creates its immobilization in the postoperative period, which is very important for the healing of the mucosal wound. The sphincter must never be cut in other places because this may impair its function.

The treatment of chronic paraproctitis is a very difficult task and sometimes fails to produce complete cure. To reduce the possibility of recurrences the approach to the choice of the operation is differentiated.

THE ACUTE APPENDICITIS

Acute purulent peritonitis - one of serious complications of various diseases and damages of organs of abdominal cavity and retroperitoneal space at children. Now achieve significant progress in common surgical and medicamental treatment, methods of an anesthesia and extracorporal detoxication, but the problem of treatment of a peritonitis and his complications remains very actual. The tendency to augmentation of densities of inflammatory processes is marked, that is connected to drop of efficacy of antibacterial drugs as a result of rising to them a resistance of a modern microflora. The most often cause of development of an acute purulent peritonitis at children is the acute appendicitis, which results to 75% of emergency surgical interventions. Other inflammatory processes of an abdominal cavity which can become complicated by a peritonitis (a diverticulitis, a cholecystitis, a pancreatitis, etc.), meet at children extremely rarely and compound no more than 0,5% an emergency surgical pathology. The perforate appendicitis at children till 5 years meets in 50%, and at children till 3 years - in 85% of observations. In general, frequency of development of an appendicular peritonitis, according to various authors, achieves from 7,5% up to 52,8%. A plenty of postoperative complications at a peritonitis always aroused anxiety of surgeons. Most often of them are the early adhesive intestinal obstruction and a progressing peritonitis. Frequency of a progressing peritonitis can achieve 23%. The statistics of various clinics indicates, that infiltrates and abscesses of an abdominal cavity arise in 1,82%-19%, and intestinal fistulas - in 0,12%-0,84% of cases. The lethality at a peritonitis of an appendicular genesis, according to various clinics, changes in significant limits - from 0,7% up to 22,9%.

Etiology and pathogenesis

The acute appendicitis represents nonspecific inflammatory process in a worm-shaped shoot. The polymicrobial flora usually participates in his development, characteristic for contents of an healthy intestine. The infection contamination is implanted in a wall of a worm-shaped shoot mainly by the enterogenous way, that is from his lumen, gets in the hematogenous or lymphogenous way less often. For originating inflammatory process in a wall of a worm-shaped shoot influence of some

promoting factors is necessary. To them carry character of a food, stagnation of contents, change of a reactivity of an organism. The apparent role in originating an acute appendicitis is played the conditions of a food. The abundant meal with the significant contents of meat and adipose nutrition usually results in constipations and an atony of an intestine and augmentation of a case rate an acute appendicitis. It proves to be true also the known fact of drop of number falling ill an acute appendicitis during social disasters, for example - within war.

Other promoting factors are bends, flexures of a worm-shaped shoot, hit in a lumen of a process feces stones, foreign bodies. The fixed role in originating an acute appendicitis is played also with change of a reactivity of an organism. Quite often acute appendicitis arises after the transferred diseases: more often anginas, children's contagions, respiratory and intestinal virus infection contaminations.

For an explanation of mechanisms of development of inflammatory process in a worm-shaped shoot series of theories from which the basic are contagious, mechanical (the theory of "stagnation") and neurovascular are assumed. It agrees neurovascular theories in the beginning there are metastasises ad nervous of a circulation (a spastic stricture, an ischemia) which result in trophic distresses in a wall of a worm-shaped shoot, down to a necrosis of separate sectors. There is a significant amount of scientific works in which the leading part is taken to the allergic factor. The certificate of it is presence of crystals of Sharkow-Leiden and a significant amount of mucilage in a lumen of the inflamed worm-shaped shoot.

At children, in comparison with adults, the acute appendicitis has series of distinctive features which can be explained by anatomic-physiological features of a growing organism. The most typical features of an acute appendicitis at children are the following.

1) At baby the appendicitis is observed extremely rarely, frequency of a case rate accrues with the years. A rarity of disease by an appendicitis of baby explain character of a food (mainly liquid milk nutrition) and small number of follicles of a mucosa of a worm-shaped shoot that frames a unfavorable background for development of an infection contamination. With the years the number of follicles is enlarged, and the case rate is in parallel enlarged by an appendicitis.

2) At children inflammatory changes in a worm-shaped shoot educe much faster, than at adults, and the child is more younger, the this feature is more brightly expressed. Speed of development of inflammatory process in an appendix at the child explain age features of a constitution of nervous system. It fixed, that at any children's age, it is especially in early, poor maturity the intramural nervous device is marked. In ganglions of a worm-shaped shoot there is a plenty of small cells such as neuroblasts. It is reflected in development of pathological process, as in tissues where the nervous system wears embryonic character, pathological process runs unusually.

Classification

One of the most wide-spread and convenient from the practical point of view is the classification suggested by Kolesov V.I. (1972). According to this classification distinguish the such forms of an acute appendicitis.

1. A feebly marked appendicitis (a so-called appendicular colic – "colica appendicularis"). Now many surgeons criticize this classification unit.
2. Simple or catarrhal, a superficial appendicitis ("appendicitis simplex").
3. The destructive appendicitis ("appendicitis destructives"): a) phlegmonous; б) gangrenous; в) perforated ("perforate").
4. The complicated appendicitis ("appendicitis complicata"): a) an appendicular infiltrate (it is good circumscribed, progressing); b) an appendicular abscess; c) an appendicular peritonitis; d) other complications of an acute appendicitis (an omentitis, a pylephlebitis, a sepsis, etc.).

Topographical and pathological anatomy

The anatomical localization of a worm-shaped shoot playing a role in clinical exhibitings, can be various. Distinguish typical and atypical locatings of a worm-shaped shoot. There are typical positions of a shoot more often: medial, lateral, ascending, descending and retrocecal. Most frequently the shoot settles down from top to bottom from a caecum. The length of a worm-shaped shoot at children of different age compounds 5-10 sm more often. To atypical positions of an appendix

concern left-hand, retroperitoneal, intercecal, intermesenterial, invagination.

Microscopic changes in a worm-shaped shoot can be various and in many cases do not depend on time, past disease from the beginning.

Simple or catarrhal an appendicitis is characterized by a moderate vasodilatation, sometimes - a hyperemia of a serous layer. The mucosa bloodshots, with plural hemorrhages. Microscopically the leukocytic infiltration of a wall of a worm-shaped shoot is marked. On a mucosa it is sometimes found defects, covered with a fibrin and leucocytes (initial defect of Ashoff).

At a phlegmonous appendicitis the surface of a serous layer is coated with a fibrinous raid, a shoot considerably intense, dwarfed and hydropic. Walls of him it is sharply infiltrated by the leucocytes. The lumen of an appendix contains the pus, sometimes only covering a mucosa, in other cases filling all lumen of the organ.

The gangrenous appendicitis is characterized by a necrosis of all worm-shaped shoot or a mucosa. At a gangrene of a shoot the wall of him is made flabby, green-black colour with purulent applyings. The lumen of a shoot contains pus with a mephitic smell. Microscopically the pattern of a serious purulent inflammation with a necrosis of a wall of the organ is determined.

At perforated appendicitis in a shoot there is various diameter of the foramen. The lumen of a shoot contains pus, feces stones. A mucosa of a shoot in part necrosis. Microscopically determine an inflammatory infiltration of all layers of a wall of the shoot.

Clinical picture

Children of all age-grades, but mainly in 5-14 years fall ill with an acute appendicitis. Girls on statistics are sick approximately in 2 times more often than boys.

The clinical pattern of an acute appendicitis is determined by age of the child, a somatotype and a resistance of an organism, presence a morbid background, and also a degree of morphological changes in a worm-shaped shoot and his locating in an abdominal cavity. Considerably alter a clinical pattern the arisen complications. For an acute appendicitis the following general attributes are characteristic: an abdominal pain, a dysphagia (a nausea or a vomiting), a dyspepsia (a diarrhea or a constipation),

infringements of an intestinal peristalsis, rise in temperature, an frequency of pulse, change of the general state. It is necessary to remember, that exists two basic clinical symptoms of an acute appendicitis - a *pain and a passive resistance* of muscles of a front abdominal wall (defans) in dextral ileal area**.**

The abdominal pain is the most constant attribute. Character, intensity and localization of a pain are different. At the majority of children the pain appears suddenly and is localized in dextral ileal area. At other patients it is localized in the beginning actually in epigastric or paraumbilical area and, later 2-3 hours, migrates in dextral ileal area. Its character children determine as a whining, constant pain. Gradually it strengthens. As a rule, the pain at an acute appendicitis does not irradiate.

The nausea and vomiting are typical symptoms of an acute appendicitis and arise almost always after a pain. Usually first there is a reflex vomiting to alimentary contents, then it becomes less abundant. Frequency of a vomiting can serve as a parameter of the form of an acute appendicitis - at the complicated process it accepts character "exhaustion". At lines of children in an initial stage of disease the delay of a feces and gases is observed, in other cases - there is a diarrhea. The body temperature at the majority of patients is leveled up subfebrile digits, can achieve 38°C. Pulse becomes frequent collaterally to rise of temperature. At the destructive forms of an appendicitis and the expressed intoxication the frequency of pulse does not meet to change of a body temperature.

Objective inspection begin with survey of a stomach, determine his participation in respiration. The dextral half of stomach usually lags behind in the act of respiration. For definition of localization of a pain to the child of oldest age suggest to cough, then it at once marks a place of the greatest morbidity. At a palpation in area of a locating of a worm-shaped shoot, more often in dextral ileal area, morbidity that is one of characteristic attributes of an acute appendicitis is determined. The important symptom of an acute appendicitis is the strain of muscles of a front abdominal wall in the dextral ileal area, arising reflexly at an inflammation parietal of the parietal peritoneum. Rate of strain of muscles happens various, from insignificantly expressed up to a stomach "as a board". For revealing a mild strain of muscles it is necessary to carry out a

cautious palpation, to start with able-bodied sectors of a stomach and gradually to come nearer to dextral ileal area. It is expediently to carry out also a simultaneous palpation of an abdominal wall in both ileal areas. In some cases, it is especially at children with a concomitant neurological pathology, the strain of muscles happens feebly marked or the explorer can not determine him at all.

For diagnostics of an acute appendicitis it is offered more than 100 symptoms based on calling by that or a different way of a pain reaction in the location of a worm-shaped shoot. Usually for diagnostics use some signs, from them the most important is *symptom of the Shetkin-Bloomberg*. It is produced by slow pressing by dactyls on an abdominal wall and take away hand. During this moment there is an intensifying a pain. The symptom is based on a irritation of the inflamed peritoneum and is one of early an attribute of a peritonitis, including at an acute appendicitis. Symptom Shetkin-Bloomberg happens positive in 90% of cases of an acute appendicitis.

And observed with the same frequency *the symptom of "sliding" of the Voskresenskiy* is not less characteristic. It is produced as follows. Having pulled the left hand a shirt of the child during an expiration, by tips of dactyls of a dextral hand do slipping movement downwards of the left hypochondrium to dextral ileal area and detain here an a hand. During this moment the patient marks intensifying a pain.

Symptom of the Rovsing, consists in jerky pressing on an abdominal wall in the location of a descending colon. Thus there is an intensifying a pain in dextral ileal area. It is caused by transfer of pressure through an abdominal wall and loops of an intestine on the inflamed worm-shaped shoot.

At an inflammation of a peritoneum the hypersensibility of an abdominal wall to each push by a hand is observed. *The symptom of the percussion morbidity of Razdolskiy* is based on it - intensifying of a pain in dextral ileal area at carrying out of a percussion of a front abdominal wall on a course of a framework of a colon in a direction "counter-clockwise". Approximately at half of children intensifying a pain in dextral ileal area in a position on the left side is marked *(symptom of the Sitkovskiy)* and at a palpation in this position *(symptom of the Bartomje-Mihelson).*

At a descending locating of the worm-shaped shoot or at drawing in process of a pelvic peritoneum research through a rectum finds out morbidity of its front wall. The vaginalis research at girls is not carried out.

For diagnostics of a retrocecal appendicitis use *symptom of the Jaure-Rozanov* (morbidity at pressing by a dactyl in area of triangle Petit) and *symptom of the Habay* (a pain during the moment a pressing dactyl take away in area of triangle Petit).

Symptom of the Gendrinskiy use for differential diagnostics of an acute appendicitis from an acute salpingitis (basically at girls). This symptom consists that in a position on a back at the patient press a stomach in the Kummel's point (on 2 sm below and more to the right from a navel) and, not taking away a dactyl, appeal for the patient to sit. Intensifying of a pain testifies to an acute appendicitis; decrease - to an acute salpingo-oophoritis.

At the majority of children at an acute appendicitis already at the beginning of disease rising quantity of leucocytes in a peripheric blood which progresses with development of inflammatory process is marked, can achieve 10 G/L-25 G/L. At the destructive appendicitis the leukocytosis achieves higher digits. Besides, the deviation of the leukocytic formula to the left is observed. For diagnostics of the destructive forms of acute surgical diseases including an acute appendicitis, Simonjan K.S. (1971) has suggested to study the ferment formula of leucocytes, which includes definition of activity of an alkaline phosphatase and peroxidase of neutrophils, suxcinatdehydrogenasa and glycerophosphasdehydrogenasa lymphocytes and phospholipids in neutrophils.

Original clinical tendency the appendicitis differs at a retroperitoneal position of an appendix. Disease begins with appearance of a moderate pain in dextral ileal or lumbar areas which sometimes irradiates in a dextral femur. The state of the child a long time remains satisfactory, slightly often pulse, in a blood there is a moderate leukocytosis. With transferring inflammatory process to fat which surround a worm-shaped shoot, the state of the patient worsens, the temperature increases up to 39°-40°C, the pain strengthens. At objective research morbidity in lumbar area is determined, where also the strain of muscles sometimes is determined. Positive

symptoms of the Jaure-Rozanov, Habay, Pusternutskiy. In a blood the leukocytosis accrues, shift to the left is determined. In urine there are fresh and alkaline erythrocytes.

At a locating of an appendix in a small basin the pain usually arises in dextral inguinal area, and is long there is no strain of an abdominal wall. At a close locating of the inflamed worm-shaped shoot to urinary bladder there are dysuric phenomena, at recruitment in process of a rectum - often desires on the act of a defecation. At increase of the inflammatory phenomena there is the strain of muscles, spread from below upwards. Early diagnostics is helped by the rectal research determining morbidity of a front wall of a rectum.

It is necessary to remember also an opportunity of a left-hand locating of a caecum with a worm-shaped shoot. In such cases all characteristic symptoms of an appendicitis are determined in the left half of stomach.

Features of a clinical pattern and diagnostics of an acute appendicitis at children of first three years of life

Recognition of an acute appendicitis at children till 3 years represents the big difficulties in view of an entangled anamnesis, features of a clinical pattern and complexity of inspection of the small child. An important role that circumstance, that is an acute appendicitis at the most small children - disease infrequent. It meets at 7-10 time less often, than at senior children. In result, practical doctors are poorly familiar with its exhibitings and first of all at address of the patient think of intercurrent disease.

Owing to age features the beginning of disease at small children frequently is looked through by parents. The baby cannot precisely explain and localize initial pains, and the mild malaise thus is interpreted sometimes by parents as a whim. Small children in general is more tolerant into to a state of discomfort. And only when pains strengthen, the child shows anxiety, cries. Thus, passes a fixed interval of time before parents will notice, that the child is sick and spares a stomach. In this connection the impression is framed, that disease arises suddenly, roughly. On an inflammatory process in the worm-shaped shoot the organism of the small child answers development by original reaction: at the beginning the general phenomenon's prevail above local and take leading place in a pattern of illness. At children of a younger age-grade, as against children of

advanced age, disease is accompanied by a multiple vomiting, high temperature, a liquid feces more often. However the specified phenomena are not always expressed in the same way, and the degree of their exhibiting depends on features of an individual resistance. Usually the vomiting occurs{comes up} in 12-16 hours from the beginning of disease. The febrile temperature is frequently fixed. The diarrhea arises not less than in 25 % of cases. In connection with that a unstable feces, high temperature and a vomiting at children till 3 years - the phenomena routine at any disease, the given circumstance confuses doctors and serves as a causing for a diagnostic mistake.

The clinical pattern of an acute appendicitis at children of younger age is quite often shaded by that localization of a pain in the beginning can be acritical. Usually small children specify area of a navel. More often a pain intensive enough. As well as grown-ups, babies frequently choose a position on the right to a side with legs clasped for a trunk. Having chosen this position, the child lays quietly and does not complain. At attentive observation it is possible to note a suffering, alert look. Local morbidity in dextral ileal area is determined at 70% of patients, at the others morbidity diffusive on all stomach. The big value has skill to survey a stomach with the purpose of revealing local morbidity and character of pains. In some cases it is expedient to begin a palpation when the child is on arms at mother. Then he is not so restless and is more trustful. At inspection of a stomach not each patient can precisely specify the increased intensity of a pain, therefore it is important to watch of a look of the child and character of crying. When the arm of the doctor passes from the left half to dextral ileal area, at presence of an acute appendicitis on the face of the child there is a grimace of a pain, and crying becomes louder. At children who difficultly contact and resist to survey, the symptom of muscle protection is better for determining during dream. For this purpose it is necessary to give the child to calm on arms at mother, to fall asleep. The muscle strain and morbidity at a palpation of a stomach at the sleeping child are survived. At absence of an acute appendicitis it is possible to palpate loosely a stomach in all areas, it remains soft, painless, but at presence of an appendicitis muscle protection on the right is marked and the child wakes up of a pain. At recognition of an acute appendicitis at the most small children

symptoms Shetkin-Bloomberg, Rovsing, Razdolskiy, Voskresenskiy, etc. frequently appear not informative.

In diagnostics of an acute appendicitis decisive importance has correct analysis of the found out symptoms. It is considered, that arithmetic calculation of symptoms pro и contra at establishment of the diagnosis is an error. Errors of diagnostics of an acute appendicitis result, on the one hand, in a significant amount of "the missed cases", and with another - not less frequently patients transfer explorative appendectomy, when as the cause of abdominal pains other disease serves. All this especially falls into to children of a younger age-grade. For today the majority of explorers is considered, that the basic diagnostic criterion – this is presence of morbidity and a passive strain of muscles of a stomach in dextral ileal area. The degree of resistance of muscles happens unequal: from small (weakly perceptible) resistances up to a sharp rigidity (a stomach ""as a board"). It is the extremely difficult to determine this symptom at weakened children and patients with neurological diseases. Besides, children being afraid of inspection, meaningly strain a stomach that results in an awake strain of a prelum abdominal. To carry out differential diagnostics between passive and awake defans *the symptom Moskalenko-Veseliy* helps. The nature of this method of diagnostics of an acute appendicitis at children will consist in the following. A palpation of a stomach carry out in a position of the patient on a spin. Gradually carry out maximal "immersing" the dactyls locating in dextral ileal area, and for some time (5-10 seconds) detain a hand in such position. For this purpose children of advanced age appeal for to make a deep expiration and to relax, and at children of younger age (till 3-5 years), with negative reaction to survey, gradual progression of a hand deep into carry out during the moment apnoe at cry or in an interval between respiratory movements. At absence of a pulsing of a dextral common ileal arteria confirm the diagnosis of an acute appendicitis, and at definition of a pulsing the diagnosis "an acute appendicitis" consider non confirm.

In some cases such pattern is observed. Later some hours or for other day from the beginning of an attack the abdominal pain at the child remit, the temperature is reduced to normal or subfebrile digits, and there comes the period of imaginary well-being. However in 1-2 days from the moment of disease pains strengthen, the temperature increases, there are attributes

of a boring of a peritoneum. The important objective attribute of gravity of disease in such cases appears a sign of divergence of pulse and temperature.

At children of a younger age-grade inflammatory process at an acute appendicitis extremely rarely accepts local character. Therefore at them disease is complicated a diffusive peritonitis more often with all consequences following from here.

At neonatal and baby the acute appendicitis is extremely infrequent disease and it is turn out more often casually on the operation undertaken concerning "other disease" (a peritonitis, for example), or on section. Disease begins with anxiety of the child, abandoning of the feedings a breast. The temperature increases up to 38°-39°C, there is a vomiting which in due course progresses. At the child features are pointed, there is a suffering expression an eyes, progress endotoxicosis and a deaquation. The stomach is inflated; the strain of belly muscles is expressed evenly in all areas. Local morbidity at a palpation to determine it is not possible. In 1-2 days there is a puffiness of an abdominal wall, the skin shines. Puffiness more all is expressed in the inferior areas, spreads on genitals. On an abdominal wall the venous drawing is distinctly seen. Symptoms of a boring of a peritoneum and divergence of curves of pulse and temperature have no importance by the value of diagnosis.

Treatment

Children with an acute appendicitis are subject operative (to surgical) treatment. Operative treatment should be carried out within 1-2 hours from the moment of an establishment of the diagnosis. In case of the complicated tendency of disease, when carrying out of preoperative preparation is necessary, prolongation of operative treatment till 4 o'clock is allowable. At children carry out appendectomy by the ligaturing or immersing method. The modern approach to treatment of an acute appendicitis at children's age provides an use of the laparoscopic methods.

PERITONITIS

Peritonitis's represent the group of inflammatory diseases of a peritoneum which differ on the causes of the originating, to mechanisms of development and features of clinical tendency.

Etiology and pathogenesis

Numerous microbiologic researches of last years have convincingly proved, that the peritonitis represents a polymicrobial aerobic-anaerobic infection more often. The major importance thus is attached to microbial contamination and to infringement of a parity of aerobes and anaerobes at morbid conditions. Now there is an opinion, that anaerobes and aerobes play a unequal role in originating, tendency and an outcome of an acute peritonitis. Anaerobes responsible for development of late stages of a peritonitis while aerobes are sowed in incipient stages more often. In a genesis endotoxicosis at a peritonitis at children the big value is attached an paresis of intestine, as to one of exhibitings serious function distresses of a gastrointestinal path. At infringement of the reduction activity of an intestinal wall it is sharply broken digestion by wall, in an intestinal wall processes of an adsorption are broken, that conjugate to entering in a blood-groove of the products of hydrolysis of proteins. At this time toxins and bacteria are capable to penetrate through an intestinal wall and to enter in an abdominal cavity. In result toxicity of inflammatory exudate is considerably enlarged. The peritoneum has the expressed ability to resorption of the exudate, that enhances intoxication of an organism.

Separate group specific peritonitises compound; to which concern a tubercular and gonococcal peritonitis, an actinomycosis and a lues of a peritoneum.

In a pathogeny of an acute purulent inflammation of a peritoneum the leading part is played the following factors, which inextricably related among themselves: 1) the pathogenic microorganism, 2) intoxication, 3) an hypovolemia, 4) expressed disbolism.

Bacterial infestation of a peritoneum is a leading pathogenetic part in development of a peritonitis. The first reaction from the side of a peritoneum at entering of the pathogenic microorganism or pus - is a dilating a vascular network and originating of a inflammatory

hyperemia. Alongside with it there is an augmentation of a permeability of vessels and is formed peritoneal exudate. In early stages exudate usually it happens serous, without strings of a fibrin. Later the contents of a fibrin in exudate is considerably enlarged, and it is postponed on serous surfaces as strings or flakes.

The nature of intoxication at a peritonitis finally is not found out. Intoxication bind mainly to formation and an adsorption from a abdominal cavity of endotoxins and exotoxins, indole and skatole, phenol and cresol. A principal cause of intoxication - are deep infringements of an interstitial exchange, in particular proteinous and water-salt. In late stages of a peritonitis intoxication is caused by toxicants which circulate in a blood.

From the very beginning of inflammatory process, at a measure of development of a peritonitis, there are serious infringements of motor function of an intestine. In result of the paresis and the subsequent paralytic obstruction of an intestine there is a stagnation of intestinal contents with its decomposing, formation of a plenty of gases and a distention of intestinal loops. Infringements of processes of an adsorption, alongside with rising of a permeability of vessels, conduct to amplified allocation and a clump of plenties of liquid in a lumen of an intestine and to a deaquation of an organism. At height of development of an acute peritonitis there are significant losses of liquid, first of all - from exocellular sector. The transudation of plenties of liquid and abjection of digestive juices in a lumen of an intestine are simultaneously accompanied by loss of plenties of protein and electrolytes. The state is aggravated with the joining vomiting, which results to the even greater loss of water, electrolytes and protein. Loss of plasma at height of development of a paralytic obstruction can achieve initial quantity of plasma. All this together with loss of liquid, protein and salts at exudation in a loose abdominal cavity results to falloff of volume of a circulating blood, a hypoxia, infringement of the microcirculation inadequate perfusion of tissues, that is a state of a shock.

At an acute purulent peritonitis it is possible to observe 2 types of a shock - hypovolemic and toxic. The hypovolemic shock is characterized by development of the circulation failure. It is produced by significant decrease of volume of a circulating blood due to the expressed decrease of

plasmatic volume, loss of exocellular liquid, a deaquation of an organism which results from accumulation in a abdominal cavity exudate with the high contents of protein, and also as a result of a vomiting. The circulatory inefficiency at a hypovolemic shock is characterized by change of minute volume of heart, falling of an arterial and central venous pressure, decrease of rate of a blood-groove, significant drop of the reducing functions of a myocardium, development of energy-dynamic failure of a myocardium.

In a pathogeny of a hypovolemic shock, cardiovascular failure at a peritonitis play a role kininums, freed as a result of action of enzymes and endotoxins. Kininums produce dilating shallow vessels and augmentation of a vascular permeability, enhance peritoneal exudation and aggravate a hypovolemic shock.

Less often the circulation failure educes develops owing to endotoxemia (endotoxic shock). Thus circulation failure grows out complex influence of bacterial toxins on heart, vessels and direct influence on a cellular metabolism. For this form of a shock the hypotension, a tachycardia, a hectic temperature curve, paleness of skin are characteristic. Hemodynamic alterations are shown depressing of cardiac emission, augmentation of peripheric vascular tone and elongation of time of circulation. The augmentation of a peripheric resistance testifies that the hypotension is not caused by a vascular collapse. Thus there are no attributes of an oligemia (the expressed changes of a hematocrit, the central venous pressure, quantity of urine). Close to normal, parameters of the central venous pressure speak about absence of a heart failure. All this confirms presence of a toxic shock.

At an acute purulent peritonitis infringements of microcirculation and a hypoxia develops. It results in infringement of a metabolism, in particular, to augmentation of an anaerobic metabolism and a metabolic acidosis. At especially big losses of a potassium the metabolic alkalosis develops. There is an expressed feed-back between value of pH (acidity) and the contents of a potassium in a blood.

The important parts in a pathogeny of an acute purulent peritonitis are infringements of function of kidneys and a liver. There is an oligemia, in result the renal blood-groove decreases, a glomerular filtration rate and a minute diuresis decreases also. It results in an oliguria, augmentation of the

contents of a filtrate nitrogen, a urea and kreatinine in a blood and in peritoneal exudates.

Changes of a function state of a bark of parakidneys at a purulent peritonitis are characterized by augmentation of a secretion of glucocorticoids. Infringements of neurohumoral regulations, intoxication result in oppression at a peritonitis of immunologic mechanisms (cytophagous activity of leucocytes, antibody formation). Decreases of quantity γ-globulins at an acute purulent peritonitis also testifies to oppression of an immune reactivity.

Pathological anatomy

The pathological pattern of an acute purulent peritonitis depends on a source of its originating, character of microbial flora, prescription of disease, prevalence of a lesion, the general state of the patient, a reactivity of an organism. The most expressed changes are observed in the peritoneum and organs of a gastrointestinal path.

In incipient states the hyperemia of a peritoneum is observed, is especial its visceral layer, in a place of a source of a peritonitis, in an omentum. The surface of a peritoneum loses the gloss, becomes dim, rough. On a peritoneum there are thin, filmy, easily taken out applyings of a fibrin. In a peritoneum shallow nodules are determined, which represent the round-cellular infiltrates. Usually, except for a so-called immediately proceeding peritoneal sepsis, in a abdominal cavity it is formed exudate. In the beginning it yellowish, without an impurity of a fibrin, contains a small amount of leucocytes. The increased contents of a fibrin in serous exudate testifies to a serous-fibrinous peritonitis. At augmentation of the contents of leucocytes in exudate it becomes in serous-purulent or purulent. At perforation of an intestine, especially a colon, or a gangrene of an intestine exudate can be putrefactive with a characteristic smell, sometimes with an impurity a fragments of the feces. At infringements of a circulation of an intestine, clottage and embolism mesenteric vessels, strangulation obstruction in a abdominal cavity there is a hemorrhagic exudate. Exudate in a abdominal cavity usually collects in a small basin, subphrenic space, between loops of intestines.

Histological research in a peritoneum determines various changes, which are characterized by the big defects of an epithelial layer, by a

hyperemia of a subject layer, a necrobiosis and a necrosis of it, and also a leukocytic infiltration. Significant changes are observed from the side of vegetative nervous system. At progressing tendency of a peritonitis the increasing destruction of ganglionic cells of a solar plexus is marked.

Changes from the side of a gastrointestinal path are expressed. The stomach extends, becomes flabby, very thin, frequently overflown with bile, intestinal contents. In a mucosa the phenomena of a hemorrhagic gastritis are determined. Motor activity in such extended stomach completely is absent. Loops of an intestine sharply extended, hydropic, overflown with liquid intestinal contents, which at pressing flows over from a loop in a loop. In all layers of an intestinal wall inflammatory infiltrates and hemorrhages are found out. The liver enlarged in the sizes, plethoric, the phenomena parenchymatous and fatty dystrophies are expressed. Alongside with it significant depletion of a liver by glycogen is marked.

In a lien the septic hyperplasia is marked. In kidneys the phenomena of a parenchymatous fatty dystrophy are marked, at patients in a serious state the pattern of an acute nephritis educes, and sometimes and the expressed hemorrhagic nephritis educes also. The sharp changes are observed in parakidneys, it are characterized by discomplixation, and disintegration of cells of a bark layer.

At serious peritonitis's changes from the side of heart and lungs are observed. The cardiac muscle becomes flabby; in it sectors fatty degenerations, a proteinous dystrophy, and in some cases and the phenomena of a myocarditis are observed. In lungs developments of stagnation, sectors of hypostases, sometimes extensive enough are observed. Some patients as a result of diffusion of inflammatory process from a peritoneum on lymphatic ways have empyema of a pleura.

Classification

K.S.Simonjan's classification (1971) according to which gravity of clinical tendency of a peritonitis is determined by dynamics of development of process in time (a reactive stage - the first 24 hours, a toxic stage - 24-72 hours and an end-stage - over 72 hours). This classification circumscribed it is applicable in children's surgery in connection with features of a resistance of a children's organism. Dynamics of a peritonitis

at children is rational to survey according to P.L.Seltsovskogo's classification (1963), modified by Fyodorov V.D. (1974). In a basis of classification the degree of prevalence of a peritonitis and morphologic-function alterations lays.

In dependence on a degree of prevalence of process distinguish: 1) an local peritonitis (circumscribed and susceptible to diffusion); 2) a diffusive peritonitis; 3) a gross diffusive peritonitis; 4) the general, or total, a peritonitis.

At an local peritonitis inflammatory process is localized no more than in two anatomical areas in limits of one floor of a abdominal cavity. The diffusive peritonitis is characterized by diffusion more than in two anatomical areas in limits of one floor of an abdominal cavity. At a gross diffusive peritonitis the peritoneum of two floors of an abdominal cavity is struck. At the general, or total, peritonitis all peritoneum is struck.

In dependence on character of an exudate distinguish: 1) serous; 2) serous -purulent; 3) fibrinous; 4) purulent; 5) a septic peritonitis. The septic peritonitis represents the special very infrequent form of a peritonitis, which is characterized by fulminant tendency, absence of an exudate in a abdominal cavity, a sharp hyperemia of a peritoneum.

Clinical picture

The clinical pattern of an acute purulent peritonitis is rather diverse and depends on an etiology, a phase, prevalence of process, character of a microflora. In one cases the peritonitis can begin suddenly (perforation of a ulcer, a rupture of an intestine). In other cases the peritonitis arises as consequence of inflammatory process in one of organs of abdominal cavity (an acute appendicitis).

The child with a peritonitis occupies the compelled position on a spin or half-sitting with hips clasped for a stomach. The consciousness clear, is conserved frequently up to the death. At an aggravation of condition there is an euphoria, exaltation. The person is pointed, the skin acyanotic, is coated cool sweat, extremities cool to the touch, cyanotic. Tongue dry. Strong thirst is marked. Arterial pressure is usually reduced and falls on a measure of progressing of inflammatory process. Pulse often, up to 120-140 imp/min, small filling. Respiration often, superficial. Educing paresis an intestine results in high standing a diaphragm and the

even greater infringement of the act of respiration. The temperature at a peritonitis is usually raised up to 38°C and more, but sometimes remains subfebrile or normal (it is the extremely rarely). The divergence between considerable increase of temperature and a significant frequency of pulse is usually found out. The diuresis at a peritonitis is reduced. On a measure of progressing of a peritonitis the anemia educes, in a peripheric blood the leukocytosis with a deviation to the left.

Local symptom of a peritonitis is the abdominal pain which in the beginning is localized in the location of a source of a peritonitis, and then is distributed on all stomach. On a measure of progressing of intoxication the pain weakens. At a palpation diffusive morbidity on all stomach is determined, the most expressed in area of the initial locus. Almost at all patients symptom Shetkin-Bloomberg is determined. One of the most typical symptoms is the strain of muscles of an abdominal wall. The expressiveness of a strain happens various, from slight, hardly revealed, up to a stomach "as a board". Revealing slight degrees of a muscle strain from children needs a gentle, superficial palpation. In late stages, on a measure of increase of an inflation of an intestine and accumulation exudate, the strain of muscles of an abdominal wall considerably decreases. For late stages of a peritonitis characteristic there is an abdominal distention which is caused by a paralytic obstruction of an intestine and an atonic gastrectasia. At this time a constant symptom becomes a vomiting, in the beginning diaphanous liquid, then bile and, at last, dark intestinal contents with a mephitic smell (fecal vomit). At research through a rectum at a gross diffusive purulent peritonitis sharp morbidity and sagging a front wall of a rectum is marked.

Treatment

One of the main moments of an operative measure at the purulent peritonitis, directly influencing a final result of treatment, is the sanitation of a abdominal cavity. The sanitation of a abdominal cavity at a peritonitis usually comes to the end with a drainage. A choice of a drainage, access from which its installation is effected, methods of a drainage are individual and depend on personal experience of the surgeon. At the same time in children's surgery the tendency to an economical drainage was planned. Alternative to a wide drainage of a abdominal cavity is prognosticated relaparotomy and a repeated sanitation of a abdominal cavity.

Antibacterial therapy of a peritonitis is not always successful, is especial at use of routine combinations of antibiotics. Therefore in last years in therapy by the patient with a peritonitis include, besides antibiotics of a wide action spectrum, drugs of Metronidazolum, Metroinidazolum, Nitazolum.

Methods of struggle with paresis an intestine at a peritonitis numerous. These are programs of a medicamental stimulation, the electrical stimulation of an intestine, of a prolonged perydural analgesia, a hyperbaric oxygenation, a reflexotherapy, various methods extrarenal depuration. One of efficient methods of controlling with paresis and a dynamic intestinal obstruction is the prolonged intubation of an intestine special probes.

THE ACQUIRE INTESTINAL OBSTRUCTION

Acute processes of a stomach at children appear unexpectedly, suddenly and sharply. They frequently amaze the person among complete health, and a condition already before developed disease of a abdominal cavity worsen, aggravate. Them divide on inflammatory (in which basis inflammatory process lays), mechanical (which basis is the occlusion of a lumen of a digestive tube or a strangulation of a mesentery of an intestine), and on traumatic. Despite of the some reserve of such division (many diseases combine elements of an inflammation, an obstruction and a trauma), during the present moment it satisfies the majority of practical doctors.

Among acute surgical diseases of organs of an abdominal cavity the acquire obstruction of an intestine on frequency takes the second place. It concedes only to an acute appendicitis. At the same time the number of lethal outcomes at it is more, than at other acute surgical diseases of organs of the abdominal cavity which have been taken together. Frequency of an obstruction of an intestine in relation to acute surgical diseases of a abdominal cavity can achieve 9,4%. Most frequently children have invagination of an intestine and an adhesive intestinal obstruction, much less often - an obstruction on ground of the Meckell's diverticulum, bends and nodulations of thin intestine and a colon, strangulated internal hernias. The general statistics of a lethality at patients with a mechanical intestinal obstruction, according to Petrov V.P. and Erjuhin I.A. (1989), make 18-20%. Tomashuk I.P. et all (1991) on the basis of the unique statistical analysis ascertain, that the lethality from an early adhesive obstruction of an intestine changes from 16,0% up to 95,2%. The same authors result own observations: from of total lethal outcomes in 21,4% the reason was an obturation obstruction, in 19,5% - was an strangulation obstruction and in 59,0% - was the mixed form of a complete intestinal obstruction.

The obstruction of an intestine represents a syndrome (a set of symptoms) which arises at various pathological processes, is shown by infringements of a peristalsis and evacuation functions of an intestine, characterized by various clinical tendency and morphological changes of the struck part of an intestine. More simple definition says, that the intestinal obstruction represents infringement of migration of the

alimentary lump on a gastrointestinal tract. *Gradation of the alimentary lump* - the grinded nutrition (it is in the mouth, and in the esophagus), primarily enzymatic alimentary lump (it is in the stomach), secondary enzymatic alimentary lump (it is in the duodenum), a chyme (it is in the jejunum and ileal intestine); not generated feces, the generated feces (it is in the colon) and the made out feces (it is in the rectum).

In first half of last century have distinguished 2 kinds of an obstruction of an intestine - mechanical and dynamic. Wahl (1889) has shared a mechanical intestinal obstruction on strangulation and obturation. In a basis of this division there is a dependence on a degree of infringement of a blood circulation of an intestine. Among set of classifications which are based on the Wahl's principle, the most convenient for practical application proved the classification of the Chuhrienko D.P. (1958). This classification shares all kinds of an obstruction as follows. By origin - *congenital and acquire.* On mechanisms of occurrence - *mechanical and dynamic*. On presence or absence of a blood circulatory disturbance in an intestine - *obturation, strangulation and combined forms (an invagination of an intestine)*. The obturation intestinal obstruction can proceed on type *actually obturation, restriction, constriction and angulation*. On clinical tendency - *complete and partial*. The partial intestinal obstruction shares on *acute, subacute, chronic and relapsing*. Besides depending on a level of an obstacle, an obstruction of an intestine distinguish on *high and low*. Border between a high and low intestinal obstruction is the initial part of a jejunum.

Dynamic intestinal obstruction on classification of the Altshool A.S. is sectioned on *spastic* and *paralytic*. This division is based on character of functional infringements of motor function of an intestinal musculation. Still half-centuries ago was considered, that a spastic and paralytic intestinal obstruction represent essentially different pathophysiological excesses. Now the opinion was ratified, that it - different phases of one pathological process.

Etiology and pathogenesis

The reasons of an intestinal obstruction can be sectioned on contributing (the anatomic-physiological features of an organism of the child belong to them) and making (precedents). According to the modern data, in a basis of a pathogeny of an acute mechanical obstruction of an

intestine the phenomena of a shock lay. Entering and a clump of plenties of a liquid and electrolytes in a lumen of an intestine is higher than a level of an obstruction, with simultaneous sharp oppression of a return adsorption, results to a dilatation of an intestinal wall and to an strengthened secretion it liquids. The stasis of intestinal contents which develops simultaneously with augmentation of a secretion and decrease of the reabsorbtion, favours to growth of microorganisms and a meteorism. The repeated vomiting, which develops at an intestinal obstruction, results to exicosis, hypoelectrolytemias. The drop of the contents of a sodium causes hyperproduction of aldosteronum, that results in a delay of a sodium and chlorine in an organism and to simultaneous augmentation of branch of a potassium with urine. In result in the subsequent the hypokaliemia develops. In later stages of a mechanical obstruction there are deeper infringements of the water-electrolytes balance and a metabolism. Disintegration of cellular mass is accompanied by release of a plenty of a potassium. Thereof there is also oligurias and hyperkaliemia. The organism of the patient significant amounts both extracellular, and cellular protein loses, its qualitative structure changes. At strangulation obstruction except for losses of protein there are also losses and exception from circulation of erythrocytes. Dederer J.M. (1971) and Welch (1958) have established, that at extensive strangulations such losses can exceed 50% of total of erythrocytes. Arising as a result of losses of a liquid the oligemia and a progressing hypoxia of tissues result in switching of the metabolism on an anaerobic glycolysis. It results to a metabolic acidosis. The hypoxia and ischemia of an intestine, ascending of proteolytic activity of serum of a blood result in formation and penetration in a current of a blood of potent vasoactive polypeptides, lizosomal enzymes. As a result of it there is a falling arterial pressure, decrease of minute volume of heart, drop of the coronary perfusion.

Clinical picture

The clinical picture of an acute mechanical intestinal obstruction depends on a lot of factors. The level of an obstruction, a kind and a degree of an obstruction, terms of disease concern to them, and also the reasons which have caused it. In practice of the children's surgeon and the pediatrist the high acquire intestinal obstruction meets extremely seldom, therefore it is meaningful to consider the basic clinical manifestations of a

low intestinal obstruction. *Acute wavy (periodic) abdominal pain, change of a configuration of a stomach, dyspepsia and dysphagia signs* concern to the basic clinical symptoms of an intestinal obstruction.

The abdominal pain is the earliest and constant attribute of a mechanical intestinal obstruction. This pain extremely intensive, inclined to the increase, more often non-local, has wavy (periodic) character. At strangulation obstruction a pain especially excruciating. There is enough frequently children groan or cry ("ileal cry"). In intervals between pain attacks patients can be abirritated a little. Frequently at children with a mechanical intestinal obstruction, it is especially at a strangulation, the pain irradiates in external genitals, an internal surface of a hip, a loin, under a scapula. Migration of a pain at this category of patients, in connection with absence of precise localization, is not characteristic. On a measure of progressing of the phenomena of an intestinal obstruction and increase endotoxicosis there is a shock, then the pain can weaken. Therefore weakening of an abdominal pain on a background of deterioration of the general condition is considered a bad prognostic attribute.

At the first time from the moment of development of an acute low mechanical intestinal obstruction the configuration of a stomach is not changed. Later 3-5 hours and later an abdominal distention is enlarged. The stomach becomes asymmetric (azygomorphous) because of the conturation loops of a small bowel (much less often - a colon) on a forward abdominal wall. At early stages of development of disease a stomach usually soft in all areas, i.e. passive defans of the muscles of a forward abdominal wall are not present. Local morbidity sometimes is marked - in case of a palpation of the invaginate or an adhesive infiltrate, for example. Frequently, basically at a strangulation, morbidity is estimated as a wide-spread. For patients with a low intestinal obstruction a lot of clinical signs is characteristic. *The symptom of the Wahl* - conturation loops of an intestine on a forward abdominal wall with a sound of a high thympanitis above it. *The symptom of the Sklyarov* (capotement) - at carrying out of a jerky palpation of a stomach is audible a capotement of a liquid which moves in a lumen of an intestine. ***The symptom of the Willms*** (hum of a falling drop) - at change of a position of the patient the hum of the falling drops on a liquid contour in a lumen of the stretched loop of an intestine is audible. *The symptom of the Kiwull* is informative enough - at a simultaneous

percussion and auscultation above a loop of an intestine which conturate on a forward abdominal wall, is audible a sound with a high metal shade. In a terminal phase of a mechanical intestinal obstruction *the symptom of the Lotheissen* (at auscultation of the stomach respiratory and cardiac hums are auscultated), *the symptom of the Obuhovsky hospital* (empty and a gaping ampoule of a rectum) are shown. In case of development of complications, such as the necrosis of an intestine and perforation, appear the symptoms of a peritonitis - a resistance of muscles of a forward abdominal wall, positive signs of the irritation of a peritoneum.

All children with an acute mechanical intestinal obstruction have nausea and a vomiting which in the beginning have reflex character. Further there is a stagnant vomiting by the contents of a small bowel (chyme). Vomitive masses of green color of various shades, rich, sticky, contain albesent and yellow-brown inclusions as lumps and strings, with an alkaline smell. In case of late diagnostics and occurrence of an end-stage of a low intestinal obstruction the vomitive secret is homogenized, gets brown color and a bad (hydrosulphuric) smell. Such vomiting gets the name "fecalis".

At children with the acquire mechanical intestinal obstruction at initial stages of illness gases and a feces in scanty quantity can go away from rectum, however on a measure of progressing of pathological process the feces and gases do not go away at all.

The basic special method of the research which confirms presence at the child of the acquire intestinal obstruction, is survey radiography of the organs of an abdominal cavity. The basic and classical radiological attribute of a complete intestinal obstruction - presence of gas bubbles with a horizontal level of a liquid (*bowl of the Kloiber*). The second characteristic attribute of an obstruction is the expansion of the separate loops, forming a light arch, which is directed to camber up - *a symptom of an arch*. It is necessary to remember, that radiological attributes of a syndrome of a complete mechanical obstruction appear already in far late cases. Besides as special methods of research it is possible to use ultrasonography, a computer tomography, a magnetic-resonant tomography, a laparoscopy, etc.

Adhesive disease, adhesive intestinal obstruction

The adhesive disease is an illness which develops as a result of occurrence and progressing of adhesive process in a abdominal cavity. Recently this pathology spare the big attention that is caused by augmentation of number of patients, and in particular augmentation of densities of an adhesive obstruction of an intestine. Adhesive process is the reason of an obstruction at 80% of patients, also causes at 76% the most dangerous a strangulation kind of an obstruction. For the first time Vezaliy and Harvey have described adnations in the abdominal cavity, which caused an obstruction of an intestine. Hunter (1793) has described the adhesions, which have arisen after a gunshot wound of a stomach. In occurrence of adhesions plays a role as influence of external factors (the operation, a trauma, inflammatory processes, etc.), so and properties of an organism. On data Perry et all. (1955), at 79% of patients the adhesion are consequence of the operative manipulations, at 18% of patients - are consequence of the inflammatory processes, and at 3% of patients they have by congenital character. One of principal causes of formation of adhesions is the trauma of a peritoneum (mechanical, chemical, thermal). Here it is necessary to relate an operative manipulation, application during its hot normal saline solution, alcohol, Iodum, other antiseptics, dry antibiotics, concentrated solutions of antibiotics, electrocoagulations, etc. All these factors cause damage of a mesothelial layer of a peritoneum. The some role play the seams, ligatures, a desiccation of intestinal loops, get in a abdominal cavity of foreign organs, in particular Talcum from gloves of the surgeon. Matters as well the closed trauma of a stomach, which results in formation of intraabdominal hemorrhages. Among various operative manipulations after which pathological adhesions are formed, at children meets appendectomy (about 70%) more often. A little bit less often the adhesive disease meets after operations concerning an obstruction of an intestine (about 15%), traumas of an abdominal cavity with damage of internal organs (up to 10%). On a share of other operations it is necessary about 5% of cases of an adhesive disease.

Finally the question on a role of inflammatory processes in development of adhesions is not found out. Experience shows, what not any inflammatory process conducts to development of an adhesive disease.

Surgeons repeatedly observed absence or insignificant quantity of adhesions in an abdominal cavity at repeated operations at patients who have transferred in the past a gross diffusive peritonitis.

In a basis of formation of adhesions lays the ability of a peritoneum to develop sticking exudate in reply to damage. Thus on the damaged surface strings of a fibrin, a cell of the exudates are postponed. Of them the network of the elastic, collagenic fibers starts to be formed, than it becomes covered by a layer of mesothelial cells. Thus, later 2-3 hours, occur restoration of defect of a serous layer of a peritoneum. If the processes of restoration of the mesothelium and restoration of a peristalsis of intestines on deseroside surfaces are broken, the fibrin does not resolve. Between strings of a fibrin appear collagenic, and later - elastic fibers, which form gentle filmy plane adhesions. These adhesions settle down between the nearest deseroside sites. Since 7-10 days in these adhesions sprout blood vessels. Later from organs, which have grown together, sprout nerves. Under influence of a peristalsis of an intestine the adhesions start to be stretched, settle down in depending on tension of intestinal loops. Then there is an inspissation of adhesions, formation of the dense strings, cords.

The separate group is made the congenital adhesions. To them concern described by Lane (1909) adhesions in the area of the ileocecal angle and a terminal site of an ileal intestine, and also a membrane or a strangulation which settle down in a transversal direction on an ascending colonic intestine. There are some points of view on a parentage of these formations. One authors regarded them as the additional fixative device for ileal, blind and ascending intestines. Other researchers regard congenital adhesions as an auxiliary source of a blood supply. Densities of congenital adhesions insignificant.

The big variety of intraabdominal adhesions by origin, to the form, a structure and localization was the reason of creation of numerous classifications. Simonjan K.S. (1966) on clinical tendency distinguishes 3 forms of an adhesive disease**:** ***acute, intermittent and chronic***. The chronic form has 3 variants of clinical tendency**:** ***specific*** (tubercular, for example), ***perivisceral*** (it is caused various periprocesses), ***abnormal*** (the Lane's adnations, the Jackson's membranes). The intermittent form is characterized by short attacks and relatively long remissions.

The acute adhesive obstruction can be shown on type strangulation and obturation obstructions. On time of occurrence it section into an early obstruction (the first 10 day from the moment of performance of operation), the early deferred obstruction (11-30 day after performance of operation) and a late obstruction (after a month and later after an operative manipulation). The strangulation adhesive obstruction results from infringement or bend loops of an intestine around of mesenteric cord adhesion. The clinically picture quickly progressing strangulation obstructions with the expressed constant pain on all stomach is shown. The obturation adhesive obstruction arises in result to an hyperflexion or a prelum of an intestine adhesions without involving in process of a mesentery and is characterized by a periodic colicky pain, a vomiting. In general, clinical manifestations of an acute adhesive intestinal obstruction nonspecific and also fully comply with the general symptoms of an acute mechanical intestinal obstruction. The presence of the compromating postoperative cicatrix on a forward abdominal wall serves as a distinctive attribute.

The chronic adhesive disease is shown as a perivisceritis more often. It is characterized by presence of adnations, periprocesses which cover various organs and cause infringements of their function. Clinically disease is characterized by an abdominal pain of various intensity, the dyspeptic phenomena, periodic inflations, a delay of a feces and gases. The aggravation of general condition usually arises after errors in a meal (plentiful rasping nutrition). During objective research it is possible to find out signs of a perivisceritis (a chronic adhesive intestinal obstruction). *The symptom of the Rozengame* - occurrence of an abdominal pain at procrastination of the left costal arch from top to bottom. *The symptom of the Karno* - intensifying of an abdominal pain at a sharp extension of a trunk. *The symptom of the Leott* - intensifying of an abdominal pain at procrastination and shift of the dermal pleat of stomach. *The symptom of the Bondarenko* - occurrence of a pain at shift of palpated organ perpendicularly its axis.

Nepokojchitskij E.O. (1974) suggests to distinguish 4 degrees of diffusion of intraabdominal adhesions. I degree - separate cord or circumscribed process amazes no more than 50 sm of a small bowel, more often her lower part. The adnations of a small bowel with a parietal

peritoneum of a forward abdominal wall and the next organs (a colon, a bladder, a uterus with appendages) are possible. II degree - is involved in adhesive process of the small bowel on an distance up to 1 meter. Adnations with operational cicatrix parietal peritoneum of a front and lateral walls of a stomach, a colon and other next organs are showed. III degree - intraabdominal adhesions amaze a small bowel on an distance from 1 meter up to 3 meters. There are extensive adnations with an omentum, an operational cicatrix, a parietal peritoneum, an organs of an abdominal cavity. Free from adhesions there is only a top part of a small bowel. IV degree - total adhesive process amazes all serous layer of an abdominal cavity.

Prophylaxis and treatment

Prophylaxis of formation of adhesions has the important practical value. Methods of the prevention of formation of adhesions can be divided into 2 groups. Actions which are directed on decrease of the trauma of the peritoneum during operation concern to the first group. To them concern: sparing surgical technics, care with tissues, a careful hemostasis and peritonization of the deseroside sites, exact indications to application of tampons and drainages, refusal of application of dry antibiotics. The second group includes methods of the prevention of formation of adhesions with the help of introduction of various solutions into a abdominal cavity. From a plenty of substances which enter intraabdominal, the attention draws to application of anticoagulants, fermental preparations and corticosteroids. Application of anticoagulant preparations (Heparin, Fibrinolysin, a polyvinylpyrolidone), and also fermental preparations (Streptokinasa, Hyaluronidasa) is based on aspiration to reduce quantity of a fibrin in an abdominal cavity. Use of steroid preparations (Cortison, Hydrocortison) is connected to their ability to oppress proliferative activity of a mesenchymal tissue, to detain formation of the granulational and a cicatrical tissue, that conducts also to inhibition of formation of intraabdominal adnations. Besides in the postoperative period for the prevention of formation of adhesions apply physiotherapeutic methods of treatment (a diathermy, electrical foresis with Lydasa, ultrasound with Hidrocortizon and unguent "Kontractubex").

Effective methods of conservative treatment of an adhesive disease, in particular during time between attacks, does not exist. At occurrence of

a pain attack and the phenomena of an obstruction of an intestine, the patients will hospitalize in surgical departments where the complex of conservative actions is carried out. At absence of effect within 2-3 hours, the children is necessary to operate concerning an acute adhesive obstruction of an intestine. The operative manipulation at an acute adhesive obstruction should be minimal and directed on elimination of an obstruction of an intestine. In case of a necrosis of an intestine carry out a resection with applying an anastomosis or shape an intestinal fistula.

CLINICAL MANIFESTATIONS, DIAGNOSTICS AND TREATMENT OF THE INVAGINATION OF THE INTESTINE AT CHILDREN

An invagination name inculcation of any piece of an intestine in a lumen of the next site of an intestine. This disease is known enough for a long time, the first have described invagination Realdus Columbus, Fabricius Goldanus and Riolan at the end of XVI century, has in more detail described Pairy in 1677.

The invagination is the most often form of an intestinal obstruction at children. At men the invagination happens in 1,5-2 times more often, than at women. Intestinal invaginate usually consist from 3 cylinders: external (perceiving) and actually invaginate. Actually invaginate consist, in turn, from middle and internal cylinders. The place of transition of the external cylinder in middle cylinder names to neck of invaginate. Transition of the middle cylinder in the internal cylinder names to as the head or an apex of the invaginate. Between internal and middle cylinders squeeze a mesentery of an intestine. Except for simple invaginations meet invaginates, consisting from 5, 7 and even the greater number of cylinders.

Depending from localization, directions of movement and a structure of the head distinguish the following kinds of invaginations. 1) A small bowel invagination - when the head of invaginate is any site of a small bowel and all invaginate is formed only from a small bowel. 2) The colonic invagination - the head of invaginate is the colon and all invaginate is formed only from a colon. 3) The blind-colonic invagination - the head of invaginate is the caecum, and worm-shaped shoot and an ileal intestine are involved together with a caecum in a lumen of a colonic intestine, pass through a Bauhinia's valve. 4) The ileal-colonic invagination - the head invaginate is formed by an ileal intestine which inculcate into a colon through a Bauhinia's valve without involving a caecum and a worm-shaped shoot. 5) The jejunum-gastric invagination - the jejunum inculcate into a stomach through a duodenal intestine or a gastrointestinal anastomosis. 6) The diverticulum-intestinal invagination – the Meckel's diverticulum invaginated in an ileal intestine. 7) The appendices-blind invagination – the worm-shaped shoot inculcate into a caecum. 8) The complex invagination - invaginate consist for 5, 7 and more cylinders. 9) The plural invagination

- inculcate occurs in several places of a gastrointestinal tract. There is more often a ileal-colonic invagination (45-60%), a blind-colonic invagination (20-25%), a colonic invagination (12-16%), an small bowel invagination (10-16%).

The invagination of an intestine usually occurs for the direction of a peristalsis (a descending invagination), less often inculcate arises in an antiperistaltic direction (retrograde or an ascending invagination). In the late diagnosed cases invaginate can reach an ampoule of a rectum and fall through an anal aperture. There are described as well as variants of a blind-colonic invagination lateral or partial inculcates of one wall of a caecum; of its dome (the Blowell's form); an invagination of the pleats (gaustraes) of a caecum.

In a parentage of an invagination play a role anatomic-physiological features of an intestine, i.e. the contributing reasons. To them concern an obtuse ileocecal angle; mobile, with the big ampoule, a caecum; a long mesentery; the anomalies of the development of Bauhinia's valve; the infringements of a parity of a lumen of small bowel and a colon. However the basic role in occurrence of this disease is played the making reasons. Inflammatory processes in a wall of an intestine, a tumour, a hematomaes, a helminthic invasion, foreign bodies, a wrong meal concern to them. In a parentage of an invagination at children the certain role is played with an adenoviral infection which causes infringements of a rhythm of a peristalsis. There are some theories which explain the mechanism of formation of the invaginate. The mechanical theory explains inculcate by presence of a tumour, of a hematoma in an overlying piece of an intestine, owing to what it invaginated in below posed part of an intestine. According to the paralytic theory, it is active peristalsis piece of an intestine inculcates in expanded (in result paresis) below laying part of an intestine. The spastic theory the conducting position spares to infringement of coordination of reductions of a circular and longitudinal musculation of an intestine. In the beginning there is a spastic stricture of a circular musculation of an intestine, then on the spastic site approaches below laying piece with spastic and the reduced longitudinal musculation. In the subsequent a circular spastic stricture of the cylinder, which inculcate, distributed to below laying sites of an intestine that promotes the further inculcate. The clinical picture of an invagination of an intestine depends from many

reasons and first of all from a degree of a prelum of a mesentery by the invaginated loop of an intestine. At the expressed prelum of a mesentery, with infringement of a circulation in it, disease proceeds as a strangulation. At an insignificant prelum of a mesentery the invagination proceeds as an obturation. Besides it is the importance the sizes of defeat, localization, age of the patient. In overwhelming majority of cases the invagination meets at baby (4-12 months). In this group disease proceeds most hardly.

On clinical tendency distinguish 3 forms of an invagination. 1) *The superacute form* with development of a serious shock and a quick necrosis of the invaginate, early occurrence of a peritonitis. If the well-timed help is not rendered, the patient perishes within day. 2) *The acute form* with less rough tendency. The phenomena of a strangulation are expressed less sharply, thus the patient can live 3-7 day, the peritonitis develops later. 3) *The subacute form* proceeds is most good-quality, mainly with the phenomena of an obturation of an intestine. Such patients without operation can live 1-2 weeks.

Disease begins with occurrence of a strong colic pain in a stomach which at the baby is shown as attacks of anxiety. In the beginning there is a reflex vomiting which renews in the late stages of disease. Than invaginate is higher, that the vomiting is observed more often. The delay of a feces and gases also takes place at an invagination. In some cases this attribute masks by allocation of a venous hemolyzed blood from an anus which appears later 4-6 hours from the beginning of disease (*the symptom of the Cruveilhier*). To last attribute is attaches great importance in diagnostics of an invagination, and prof. Mondor called persistently find out this sign, figuratively comparing blooding allocation with "raspberry jelly". Sometimes blooding allocation are determined at rectal digital research, in other cases appear after a clyster as blooding lumps of slime. At a colic invagination are observed the spastic strictures. The stomach is moderately inflated, soft at a palpation, the strain of muscles of an abdominal wall is absent (*the symptom of the Alapy*). At many patients in a abdominal cavity at a palpation it is possible to probe the tumorous formation of the cylindrical form. It elastic, circumscribedly mobile. The palpation invaginate is one of the basic signs of an invagination. The described symptoms (*a colic pain in a stomach, allocation of a blood from a rectum and find of a tumour in a abdominal cavity*) make a classical triad of signs

of an invagination which meets not at all patients. A characteristic attribute is the intensifying of the pain and spastic strictures at a palpation of invaginate (*the symptom of the Roosh*). At an ileal-colonic invagination and at a blind-colonic invagination the empty right ileal fossa is determined (*the symptom of the Shimon-Dancer*). At dynamic observation moving invaginate can be marked. The rectal research allows to determine the gape or an incontinence of sphincter of an anus, presence blooding allocations, to probe dropped out in a rectum the invaginate.

Auxiliary methods of research have the important value. To them carry ultrasonography and a pneumoirrigoscopy (pneumoirrigography). The pneumoirrigoscopy provides a contrasting of colon by air at the following technique. The empty catheter bridge with the Richardson's cylinder and enter in a rectum. The insufflation of air in a colon carry out under the manometric control. It is recommended to frame a pressure no more than 10-11 кPa. Free passage of air through the Bauhinia's valve and contrasting of the terminal site of an ileal intestine allows to exclude an ileal-colonic and colonic invagination. At presence of the invaginate in a colon the defect of filling, which having the form of a cocarde or a trident, is determined.

Methods of treatment which are applied now at treatment of an invagination part on the conservative and operative. Conservative treatment is applied in early cases of ileal-colonic and descending colonic invaginations. For straighten such invaginates apply inculcate of barium or air through a rectum under the radiological control. Circumscribed opportunities of use, the certain risk (threat of rupture of an intestine, a straighten an unvital intestine) allow to apply conservative treatment only in early terms of disease, namely - only at the first 12-16 hours. In later terms of disease the basic method of treatment is surgical. The operation is consist in a laparotomy, revision of an abdominal cavity and elimination of an invagination. With this purpose apply the manual desinvagination. The resection of the bowel at an invagination is necessary at unsuccessful desinvagination or an unvital intestine. In well equipped clinics preference give for the laparoscopic techniques.

AN INJURY THE ABDOMEN AND RETROPERITONEAL SPACE

Blunt trauma of the abdomen accounts for 3 per cent of all injuries among children and results from a heavy blow on the abdomen or back, a car accident, fall from a height, compression, etc. Any injury to the internal organs is fraught with the danger of a grave disaster which threatens the child's life almost in all cases and calls for emergency measures. In view of this children with closed abdominal trauma must be kept under close supervision and treatment in a surgical hospital. Injury to the parenchymal organs (liver, spleen) is marked by interabdominal haemorrhage with growing anaemia and haemodynamyc disorders, damage to hollow organ leads to peritonitis. All this shows the extreme importance of timely diagnosis and the choice of a rational treatment.

The condition of the abdominal organs at the time of the injury is very important factor determining the degree and severity of the damage. Active tension of the muscles of the anterior abdominal wall protects the internal organs from injury. Mild rupture of a filled hollow organ may occur with the consequent escape of its contents into the free abdominal cavity. Pathological changes of an organ predispose it to being easily injured even if the trauma is mild.

According to character and depth, the following injuries to the internal organs are distinguished: subcapsular rupture, interorganic haematoma, breaks and ruptures with disturbed intactness of the capsule, crushing, and avulsion of a part or the whale organ. Isolated ruptures of organs are encountered most frequently, multiple (simultaneous injury to more than one abdominal organ) or combined (simultaneous injury to abdominal organs and other anatomical parts of the body) are rarer. Depending on the injuries mentioned, one or another clinical picture is produced and the therapeutic tactics are chosen. When taking the medical history attention should be focused on the circumstances of the accident and the force and location of the violence. The child and his parents however are sometimes incorrectly orientated with regard to the circumstances of the injury.

The general method for an abdominal injury

Blunt injuries are particularly difficult because:

1. A patient may give no clear history that h has had a abdominal

injury, especially if he is a frightened child. His injury may be so mild that you have to question him carefully, and he may even mark into hospital.

2. His other more obvious injuries, such a fractured femur, may distract your attention.

3. He may be unconscious from a head injury and unable to tell you bus symptoms. If you anaestheze him to treat his other injures, he cannot complain of increasing abdominal pain.

4. For the first few hours after a blunt injury his abdomen may be deceptively normal. Although a haemoperitoneum usually causes pain, tenderness, guarding, and absent bowel sounds, it occasionally causes none of these things, especially in children.

5. Distinguishing between muscle pain and peritoneal irritation can be very difficult.

6. Some injuries may not show themselves for several days, especially a subcapsular haematoma of the spleen, or a retroperitoneal injury of the pancreas or duodenum.

For all these reasons, abdominal injuries need particular judgment, care and skill. So, be vigilant and suspicions. You will need a watchful age, a light touch, and sympatheticlar. Don’t let a patient go home if there is even a slight possibility that he might have injured his abdomen. If you are in any doust, observe him carefully and use the special methods.

History

Most abdominal injuries are the results of car accidents, but some follow falls from a height, especially in children.

- What object struck the patient’s abdomen?

- Where did it strike him? (For example, an injury to his spleen is much more likely after a blow to left lower chest).

- How much force was used?

Pain after an abdominal injury is always important. It is usually present, but a patient may not complain of it if he has even more painful injuries elsewhere.

- Where is the pain?

- What kind of pain is it?

- Is it getting better or worse? If pain is getting worse after abdominal injury, it probably means continued bleeding, or baking gut.

- Has the patient got pain at the tips of either of his shoulders?

Shoulder tip pain is caused by irritation of his diaphragm, usually by blood. It is a particularly useful sign of injury to the liver (right shoulder) or the spleen (left shoulder), especially if tilting the patient's head down makes it worse.

Caution!

Almost all patients with abdominal lesions after a blunt injury have persistent pain, and vomit. So the are very important signs. To begin with may be almost the only onse.

The examination of an abdominal injury

It the patient is bleeding, he is likely to be pale, anxious, and still, with cold extremities. Completely uncover his chest and abdomen and sit beside him.

1. Haw is he breathing? Shallow, irregular, or grunting respiration is typical of an abdominal injury.

2. Look for bruc'ses and abrasions. They will show you where he was hit.

3. Full for tenderness. This is less marked with a haemoperitoneum than it is with septic peritonitis. Its position may guide you as he sohich organ has been injured. Increasing tenderness usually requires a laparotomy.

Rebound tenderness is unreliable and is easily confused with muscle bruising. Pain on coughing and on percussion with your finger tips is much more reliable.

4. Fell for guarding and rigidity. Guarding progressing to rigidity is a reliable sign of peritonitis. Percuss the patient's flanks for the dullnese that may indicate a haemoperitoneum. Test for shifting dullness.

Caution. Even minimal tenderness and guarding are significant.

5. Listen for bowel sounds for 2 minutes. If you hear them, they mean nothing. When you first examine a patient, his abdomen will probably not have time to become silent. However, an abdomen which is silent, or becomes silent later, is a useful sign of peritonitis.

6. Has a patient any signs of fractured ribs? If inn's bowel left ribs are fractured, suspect a recaptured spleen. Thoraco-abdominal injures are common. Cyanosis is dangerous sign.

7. Examine limp rectally. Look also for blood on your glove. Fullness or tenderness in the recto-vaginal pouch in a girl or the recto-vesical pouch

in a boy may indicate a haemoperitoneum. Look for wounds of the perineum or buttocks at the same time.

Caution. The rectum is completely out of sight of laparotomy. To begin with its injuries may cause no symptoms. If necessary pass a sigmoidoscope.

7. Aspirate the patient's stomach and empty his bladder. If you aspirate blood, his stomach may have been injured. Leave the nasogastric tube down. You wall waste it later when he goes to the theatre.

Special methods for abdominal injuries.

These are for doubtful or difficult causes only. Where there are signs that indicate the need for a laparotomy, these methods are quite unnecessary. A position result in any of them is an indication for an abdominal injury.

1) Fest for orthostatic hypotension. This may be useful if a patient has no other obvious cause of blood boss.

Take his pulse and blood pressure while he is legging flat. Then take it again when he is sitting up. While he is lying flat, his circulation may sum to be compensated. But sitting him up may up produce a sharp fall in blood pressure, and an increase in his pulse rate. This shows that his blood volume is depleted.

2) Fest for increasing girth. Mate any initial distension and measure the patient's abdomen with a tape measure at his umbilicus. An increase in his girth will be a useful sign of the paralytic ileus that follows peritonitis or haemoperitoneum. An increase of onlug 2-3 cm indicates a large amount of abdominal fluid or gas.

3) Diagnostic paracentesis ("Four quadrant tap"). This is a useful rapid test.

Take a syringe and a 1,4 mm needle. Under local anaesthesia, or no anaesthesia at all, and using an aseptic no-touch technique, tap all four quadrants of the patient's abdomen. Push the medle through his abdominal wall until the sudden give shows that you are just inside his peritoneal cavity, then aspirate.

If aspiration is negative, take the medle out, roll him towards the side of the suspected injury, and repeat the test. If aspiration is still negative, repeat it in an hour or two, or try lavage.

4) Peritoneal lavage. Lavage is useful if you are in doubt whether a

laparotomy is necessary or not. Use lignocaine to infiltrate an area in the midline 2,5 cm below his umbilicus down to his peritoneum. Use a scalpel to wave a small nick down to his peritoneum. Using turaing movements, push a trocar and cannula into his abdominal wall. Ideally, push a peritoneal dialysis catheter through cannula and then withdraw the cannula.

If blood flows up through the tube, you have confirmed a haemoperitoneum.

A negative result does not exclude an abdominal injury.

5) Urine. Examine this for blood from a bruised kidney or a ruptured bladder.

6) X-rays. Take erect films of a patient's chest and abdomen. If he cannot sit up, take a lateral film while he is lying on his side. Another good X-ray is to turn the patient on his left side and take an AP view of his liver area.

An intravenous pyelogram is useful, so take one. The only contraindication to it is a low blood pressure which allow insufficient excretion to give you a useful film. Do a pyelogram as soon as you can. Gove him a double dose of contrast medium. Take a control film, followed by films at 5 minutes and 15 minutes; take another film at 30 minutes if you do not see the normal kidney well at 5 and 15 minutes. Look for: a functioning kidney on the other side, delayed or absent function on the injured side, and blood clots in the calyces.

7) Focused abdominal sonography for trauma (FAST). Clinician-performed sonography for the early evaluation of the injured child is currently being evaluated to determine its optimal use. This bedside examination may be useful as a rapid screening study, particularly in those patients too unstable to undergo an abdominal CT scan. The presence of peritoneal fluid should lead to the suspicion that bleeding from organ injury is present, however, solid organ injury frequently occurs without the presence of peritoneal fluid. The study may need be repeated dependent on clinical correlation. Ultrasonography shows a crush injury a patient's spleen, liver or kidney, gas under his diaphragm, peritoneal effusions or blood, haemotome of the retroperitoneal space.

8) Computed tomography (CT).

CT has become the imaging study of choice for the evaluation of injured children. CT is readily accessible in most health care facilities, is

noninvasive, is very accurate method of identifying and extent of abdominal injury, and has reduced the incidence of non-therapeutic exploratory laparatomy.

Abdominal CT imdentiopying major spleen and liver hemoperitonium. Not all children with potencial abdominal injuries are candidates for CT evaluation. Obvious penetrating injury often necessitates immediate operative intervention. The hemodynamically unstable child should not be taken out of an appropriate resuscitation room for a CT. These children may benefit from an alternative diagnostic study, such as a diagnostic peritoneal lavage, focused abdominal sonogram, or urgent operative intervention. The greatest limitation of abdominal CT scanning in trauma is the lack of ability to reliably identify intestinal rupture. Findings suggestive but not diagnostic of intestinal perforation are pneumoperitomeum, bowel wall thickening free intraperitoneal fluid, bowel wall enhancement, and dilated bowel. A high index of suspicion should exist for the presence of a bowel injury in the child with intraperitoneal fluid and no identifiable solid organ injury on CT scanning.

Presently, CT is used in children more commonly than diagnostic lavage.

9) Diagnostic and therapeutic laparoscopy.

Laparoscopy of the injured child may have its place in the evaluation of the haemodynamically stable patient. The sensitivity is comparable to diagnostic peritoneal lavage, but the specificity is higher, as one would expect by actually visualizing the injury. Studies have also shown that not only may the traumatic injury be identified with laparoscopy, but also the definitive repair may frequently be performed.

10) Angiography.

Angiography is rarely required today in the evaluation of the acutely injured children.

Caution: if you are in doubt, admit and observe patient, and examine him repeatedly. If there is any suspicion of an intra-abdominal injury, operate. If you suspect that a patient might have an abdominal injury, don’t be afraid to do laparatomy, and don’t delay. An occasional negative laparatomy is better that always waiting for some obvious indication of an abdominal injury. He will not die from a big incision, but he will die if you overlook a serious injury. If necessary, watch him carefully for at last 24

hours. The commonest of a haemoperitoneum are injures to a patient's spleen, liver, and mesentery. So search for them in that order. Even if you find no free blood or intestinal contents, he may still have a small perforation, which is temporarily sealed off. So search his abdominal organs carefully,

Spleen injury

The spleen and liver are the organs most commonly injured in dlunt abdominal trauma, with each accounting for one-third of the injures.

The spleen commonly injury in children. Diagnosis of splenic injury is suggested and confirmed by the history, physical examinatio, presence of hemoperitoneum on peritoneal lavage, radiographic assesment, or any combination of these techniques. Splenic injury usually follows blunt abdominal trauma from direct blow. Tenderness, abrasion, or ecchymosis over the left upper quadrant all suggest the possibility of injury the organ. Because the spleenlies in contact with diaphragm and chest wall, children may complain of referred left shoulder pain (Kehr's sign). Fractures of the overlying ribs are rare because of the elasticity of the child's chest wall. Microscopic hematuria suggests the possibility of splenic injury because such injury in conjunction with left renal trauma is common. Tachicardia, pallor, delayed cappilary refill time, and low hemoglobin level all suggest blood loss. If the magaitude of blood loss does not demand laparotomy for hemostasis, imaging techniques can delineate the extent of splenic injury. Children who require immediate surgery for associated extraabdominal injury can simultaneously undergo selective peritoneal lavage that will document hemoperitoneum; in children, free blood in the peritoneal cavity usually indicates injury to the spleen or liver.

Nonoperative management of a child with splenic injury is appropriate if the patient maintains stable vital signs, requires replacement of less than half of blood volume, and is free of significant concomitant intraabdominal injury. Children treated without surgery must be constantly monitored to guarantee appropriate blood volume replacement and assess adequacy of perfusion.

Liver injury

The liver is injured with same frequency as the spleen, but rupture of

the the liver with its rich blood suplay can lead to more severe hemorrhage from the intrahepatic vasculature of the vena cava. Hepatic injury is more freuquently a cause of exsanguinating hemorrhage and and may require urgent surgical intervention.

Because the liver is large, penetrating injury tj the upper abdomen often results in parenchymal trauma. Contusion and abrasion of the skin associated with tenderness in the right upper quadrant suggest liver injury. Concern about right intrathoracic injury may distract attention from a more significant infradiaphragmatic liver injury. However, the most common mechanism of injury to the liver is a blow to the right upper quadrant or right side of the chest. Blunt injury to these areas may result in varying degrees of injury, ranging from a simple capsular tear or superficial laceration to a large parenchymal fracture or extensive stellate disruption of one or both lobes of the liver. Significant hypovolemia and hypotension and obvious abdominal distention that persists despite nasogastric decompression suggest major injury to the liver.

Specific treatment of liver depends on the extent of parenchymal disruption, because most injures do not bleed actively during laparotomy. A localized and nonexpanding subcapsular hematoma with an intact capsule usually resolves spontaneously. A large, expanding subcapsular hematoma with a capsular tear requires evacuation of the hematoma, debridement of devitalized tissue and precise ligation of bleeding vessels. Diffuse parenchymal capillary bleeding often slows after cauterization or application of a hemostatic agent and manual pressure. Holding the injured parenchyma against the abdominal wall also assists in staunching blood flow.

Nonoperative management of hepatic injury is an alternative if the patient is physiologically stable and has normal systolic blood pressure, pulse pressure greater than 20 mm Hg, heart rate less than 120 beats/min., and a transfusion requirement of less than half of estimated blood volume. The goal of nonoperative treatment of liver injury is to avoid operative mortality and morbidity. Most injures to the liver stop bleeding spontaneously. Nonoperative management of stable patients is a viable option, although complications do occur with this method. The rates of successful nonoperative treatment of isolated blunt splenic and hepatic injury now exceed 90 % in most pediatric trauma centers.

Small intestine

Perforation of the small intestine may result from mild, blunt abdominal trauma. Forceful compression of distended intestine typically results in rupture along the antimesenteric border.

The child may complain of discomfort after seemingly mild trauma. If the injury is not noticed, peritonitis develops within 24 hours and is occasionally mistaken for appendicitis. Peritoneal lavage identifies organ injury if the effluent contains bacteria, bill, or stool or reveals an elevated amylase level.

Plain radiographic examination of the abdomen is of little value if the injury is located in the distal small intestine, but proximal injury results in varying degress of pneumoperitoneum evident on the radiograph.

Pancreas and duodenum

The pancreas is located in the retroperitoneum at the base of the intestinal mesentery; the right lateral and inferior margin of the pancreas is formed by the duodenum. Because of their relatively protected location, the pancreas and duodenum are infrequently subject to trauma. Blunt trauma from automobile injury, bicycle handlebar, or deliberate blows have all been implicated as cause of injury to the pancreas and duodenum.

If the diagnosis of pancreatic-duodenal injury remains in question in a child with persistent or progressive signs of peritonitis, exploratory laparatomy is the only method for accurate assessment of the pancreatic-duodenal complex.

Renal trauma

The kidney is the most commonly injured organ in the urogenital system, and children appear to be more susceptible to major renal trauma than adults.

Preexisting or congenital renal abnormalities, such as fetal lobulations, hydronephrosis, tumors, or abnormal position, may predispose the kidney to trauma despite relatively mild traumatic forces. Congenital abnormalities in injured kidneys have incidence rates of 1% to 5%.

Following any blunt trauma, the presence of hematuria (microscopic or gross), a palpable flank mass, or a flank hematoma is an indication for

urologic evaluation. Most major blunt renal injuries occur in association with other major injuries of the head, chest, and abdomen. Urologic investigation should be undertaken when trauma to the lower chest is associated with rib, thoracic, or lumbar spine fractures.

Gross hematuria is the most reliable indicator for serious urological injury. The need for imaging in the patient with blunt trauma and microscopic hematuria is not as clear cut. The degree of hematuria does not always correlate with the degree of injury. All children with any degree of microscopic hematuria after blunt trauma have typically undergone renal imaging.

Intravenous urography (IVP) has been the radiographic imaging study of choice in suspected renal trauma, and sensitivily can be as high as 90% in diagnosing renal injuries. Unfortunately IVP misses other intraabdominal injuries and may miss or understage renal injury in children by 50% compared with CT scans. CT scans are now used almost exclusively as the imaging study of choice for suspected renal trauma in hemodynamically stable adults and children. Ultrasonography has also been used to assessrenal trauma. However, its sensivity in demonstrating renal injury in comparison to CT is only 25% to 70%.

The majority of blunt renal injuries are contusions and lacerations that are minor in nature. Even in the presence of gross hematuria, most blunt renal injuries will not require exploration and will have excellent long-term outcomes. When conservative management is chosen, supportive care with bed rest, hydration, antibiotics, and serial hemoglobin and blood pressure monitoring is required. Absolute indications for renal exploration include persistent life-threatening bleeding, an expanding, pulsatile, or uncontained retroperitoneal hematoma, or suspected renal pedicle avulsion.

Rupture of the bladder

You can usually tell quite easily if a patient’s bladder has ruptured inside or outside his peritoneum from:

1. The history of the injury – a blow to his abdomen suggests rupture inside the peritoneum, whereas a fractured pelvis suggests rupture outside it.
2. The distribution of the tenderness – in extraperitoneal rupture this is narrowly localised suprapubically, in intraperitoneal rupture it is

more diffuse over his lower abdomen and ends in abvious peritonitis.

If you are in doubt, there are two investigations that may confirm that his bladder has ruptured, and show you where it has ruptured, but they are usually not necessary:

1. You can do a retrograde cystogram. Unfortunately, this requires the use of a catheter, and with it the risk of infection.
2. You can do an intravenous pyelogram, which is safer but less reliable.

If you diagnose any kind of rupture of the bladder, you will have to refer the patient urgently, or operate.

Indications for laparotomy.

The indications for laparotomy vary depending on the mechanism of injury.

I. *Blunt Trauma*.

1. Continued unstable vital signs despite adequate fluid resuscitation.
2. Transfusion requirement of half of the patient's blood volume.
3. Pneumoperitoneum.
4. Peritoneal lavage effluent containing one are more of the following:
    a) Erythrocytes in excess of 100.000 mm^3 in conjunction with normal results of computed tomography.
    b) Bile, bacteria, stool.
    c) Amylase level above 175 dl.
    d) Leukocyte count above 500/$mm^{3.}$
5. Peritoneal signs on physical examination.
6. Massive abdominal distention associated with hypotension (systolic blood, pressure less than 80 mm Hg).
7. Bladder rupture.
8. Renovascular injury.
9. Ureteral transection.
10. Pancreatic-duodenal injury.
11. Rectal laceration.

II. *Penetrating Trauma*.

1. Stab wounds:
    a) Unstable vital signs in conjunction with hypotension (systolic blood pressure less than 80 mm Hg).
    b) Signs of peritonitis.
    c) Unexplained blood loss.
    d) Peritoneal penetration.
    e) Peritoneal lavage effluent containing erythrocytes in excess of 50.000/mm^3, bile, bacteria, or stool.
    f) Evisceration.
2. Gunshot wounds: Penetration of the peritoneum.

Children with abdominal trauma who fail to respond to fluid replacement with lactated Ringer's solution and packed red blood cells require laparotomy to assess magnitude of hemorrhage and provide appropriate hemostasis. After rapid volume infusion, in the patient shows an elevation of systolic blood pressure (>80mm Hg), slowing of the heart rate (<130 beats/min), increase in the pulse pressure (<20 mm Hg) rapid capillary refill (<2 seconds), warming of the extremities, elimination of skin pallor, and clearing of the sensorium, surgery may be safely delayed whill additional essential clinical information is obtained.

ACUTE AND CHRONIC PURULENT DEFEAT OF LUNGS AND PLEURA

The lung with their combined surface area are directly open to the external environment. Thus structural, functional or microbiological changes within the lungs can be closely related to epidemiological, environmental, occupational, personal and social factors. Primary respiratory diseases are responsible for a major burden of morbidity and untimely deaths, and the lungs are after affected in multisystem diseases.

Each day our lungs are directly exposed to more than 5000-7000 litters of air which contain varying amount of inorganic and organic particles as wool as potentially lethal bacteria and viruses. In general terms, physical mechanisms including cough are particularly important in defense of the upper airways, whereas the lower airways are protected by complex mucociliary mechanisms, by antimicrobial properties of surfactant and the lung-living fluids, and by resident alveolar macrophages.

Lung defenses against infection are mechanical cellular, and humoral. Mechanisms that remove inhaled particles, including pathogenic microorganisms, consist of the anatomic design of tortuous nasal passages, the multiple divisions of the tracheobronchial tree, and the mucociliary transport system.

Cellular defenses are provided by the phagocytes – alveolar macrophages and neutrophils – and by lymphocytes.

Humoral defenses include complement, lysozyme, fibronectin, antiproteases such as α1-antitrypsin, and IgA as antivirus defense.

Protective agents in the lung-lining fluid.

- surfactant proteins – bacterial opsonisation;
- immunoglobulins (Ig A, Ig G, Ig M) – bacterial opsonisation;
- complement – bacterial opsonisation, generation of the inflammatory response;
- bacterial proteins – bacterial killing;
- proteinase inhibitors – protection of host tissues during the inflammatory response.

Infections of upper and lower respiratory tract continue to be a major cause of morbidity and mortality throughout the world and main way of infecting is by air.

Viruses are the most frequent cause of upper respiratory illnesses and than 3-5 days bacteria connected to viruses and being responsible for the majority of community – and hospital-acquired pneumonia.

The most weakness place in lower respiratory tract are bronchioles and bacteria defeats it in the first range. This viruses-bacteria damaging may be different.

Classification of pneumonia by the world pulmonary communicate:

1. Community-acquired pneumonia.
2. Hospital-acquired (nosocomial) pneumonia.
3. Suppurative and aspirational pneumonia.
4. Pneumonia in the immunocompromised patient.

Symptoms of respiratory disease.

The symptoms which point specially to the respiratory system as the seat of disease are this, breathlessness (dyspnoea) and cyanosis (but more after this is absorved by the examiner).

Cough – Perhaps the most pathognomic of all respiratory symptoms, cough is usually an expression of disease in the upper respiratory passages, the bronchi, or lung. Cough may be either a voluntary act or reflex. It consist of a forceful expiratory effect with the glottis closed, followed by sudden explosive release of the pent-up air along with spateun or other irritant matter.

Dyspnoea. The term “dyspnoea” is derived from the Greek roots *dys-* (difficult, painful) and *pnoia* (breathing).

Dyspnoea or breathlessness, may be defined as an undue awareness of respiratory effect has been related to the force used to ventilate the lungs. This force is increased when the thoracic cage or pleura is abnormally rigid, the pleural cavity filled with fluid or air, the airways obstructed, or the lungs less distensible than normal. Dyspnoea may also leave the form of tachipnoea, bradipnoea and disfarbing rhythm of breasting.

Cyanosis, a blue coloration of the skin or mucosae. It due to deficient oxygenation of the blood in the lungs resulting from inadequate ventilation of perfused areas of lung (e.g.pneumonia, emphysema); from a reduction in the total amount of air ventilating the lungs as a whole, or from a barrier to oxygen diffusion across the alveolar-capillary membrane.

Physical signs: examination of the respiratory system.

The order of examination by inspection, palpation, percussion, and auscultation is particularly suitable for respiratory system.

Inspection shows the configuration of the chest, the degree of movement and any inequalities on the two sides, the type and rate of respiration, and any special variety of breathing which may be present.

Palpation confirms the impressions of inspection, especially the movement of the chest and any abnormal prominence or recession in the chest wall. It also detects abnormal pulsation and areas of tenderness, and a special sign called vocal fremitus.

Percussion is used to demonstrate changes in the character of the lung tissue, its contained bronchi, and the surroundings pleura, and shows whether these tissues contain less or more air than normal, or fluid.

Auscultation enables the observed to suspect changes in the caliber of the bronchi by variation in the intensity of the breath-sounds, and modification of the lung susbstance by changes in the character of the breath-sounds. It also reveals the presence of abnormal exudates in the bronchi and lung alveoli which cause special adventitious sounds. Finally, it may give evidence of interference in the normal movements of the layers of the pleura on one another.

Pneumonia

Pneumonia is defined as an acute respiratory illness associated with recently developed radiological pulmonary shadowing which is either segmental of affecting more than one lobe.

Clinical features

Patients present with a short illness of cough, fever and malaise, often associated with plevritic clust pain which is occasionally referred to the shoulder or anterior abdominal wall. The sudden onset of a high fever can result in children vomiting or febrile convulsion. Appetite is usually lost and headache is a frequent accompanying symptom.

Physical examination

Inspection shows decreased movement. The entering air cannot expend the already filled or obliterated alveoli.

Palpation confirms this deficient movement and reveals an increase in vocal fremitus. Faryngeal vibrations are transmitted better though solid lung.

Percussion gives a dull note, as over solid viscera, but not quite so flat, because air is still present in the bronchi and in unaffected portions of the lung.

Auscultation generally demonstrate bronchial or tubular breathing. Adventitions sounds may or may not be present according to the amount and character of exudate in the bronchi and alveoli. In pneumonia fine inspiratory crepitations are heard in the early stages of the disease.

Empyema

This term describes the presence of the pus in the pleural space. The pus may be as thin as serous fluid or so thick that wide-bore needle. Empyema to infection in neighboring structure, usually the lung. The principal infections liable to produce empyema are bacterial pneumonia's and tuberculosis.

The pathologic manifestations of empyema are divided info three phases.

1. Exudative: this is immediate response with outpouring of thin fluid that has a low cellular content.
2. Fibrinopurulent: this stage is characterized by large quantities of pus, posteriorly and laterally, with many polymorphonuclear leukocytes and fibrin. As the fluid thickens, loculation begins and the lung is progressively less expandable.
3. Organizing: Fibroblasts grow into the exudate on both the visceral and parietal pleural surfaces, producing a membranous "peel". With increasing fibrosis. The process becomes chronic and the lung more firmly fixed.

Diagnosis

The signs and symptoms of empyema in a child are usually those of worsening pneumonia, with fever; tachypnea, shortness of breath, and, soundness, cough and cyanosis.

Breath sounds are decreased, and dullness to percussion is found.

Diagnosis is confirmed by the appearance of pleural fluid on radiography. In the early exudation phase, the fluid flows along lateral

chest wall on decubitus views. In advanced empyema, the exudate becomes a solid mass of fibrin that does not move with changes in position. In the intermediate fibrinopurulent stage, loculations are characteristic.

Ultrasonography is valuable n locoting pleural fluid for thoracentesis and in estimating the volume of fluid, but it cannot help distinguish between the density of an early exudate and that of solid fibrin. Diagnostic thoracentesis should be performed, and the fluid should be sent for Gram stain. Whenover ideal conditions, thoracentesis is done before antibiotics therapy is started.

Pneumothorax

The main cause of pneumothorax is repture of a subpleural emphysematous bulla (pneumatocelex). There are three types of pneumothorax:

1. Closed type.
2. Open type.
3. Valvular (tension) type.

Clinical features.

The onset is usually sudden, with pain or a fubing of tightuess on the afected side of the clust. The patient becomes intersingly breathlese and in severe cases cyanosed. Physical signs in the chest are of air in the pleural soace, but when the pneumothorax is small and localised there may be abnormal signs it may be revealed only by a radigraph.

Investigations.

The clust radiograph usually shows a sharply defined edge of the deflated lung. There is complete trancluceng between this and the chest wall, with no lung varking. Radiograph also show the degree of mediastinal displacement and give information regarding the presence or absence of pleural fluid and underlying pulmonary disease.

Lung abscess

A lung abscess is an area of suppuration in rohich central necrosis and cavition develop. The infection is usually polymicrobial, containing aerobic and anaerobic bacteia. The infections process develops in the pulmonary parenchyma, although congenital bronchogenic or pulmonary

cysts may became secondarily infected and be indistinguishable from lung abscess on plain clust radiographs.

Lung abscess may follow bacterial pneumonia and localaris as a rule, subpleuralea.

Symptoms of abscess are cough, productive sputum, chest pain, hemoptysis, meight loss, fever, chills and night sweats. Older children with lung abscess may produce purulent sputum, but children younger than 5 years of age do not produce sputum and may swallon the purulent material.

On physical examination, the affected area of te chest may show dulness to percussion, decreased breath sounds, and egophony. Leukocytosis is common.

The diagnosis is established by radiography, including both frontal and lateral views.

Ultrasonography and CT of the chest and a great deal to the diagnosis and management, especially for the rare central abscess abscured by infailtrates or pleural thickening.

Lung abscess in an infant suggests secondary infection in an underlying congenital anomaly (e.g. bronchogenic cyst), none of the imaging techniques (radiography, ultrasosnography, or CT) can assist in the differentiation of a secondary form a primary abscess. Such an infected cyst will not resolve and requires resection.

Management.

Whenever possible, treatment should be based on an established acbiological diagnosis. Antibacterial therapy should be modified according to the results of microbiological examination of the sputum or pus.

In practice however, the cause of the pneumonia is frequently not know in when treatment bas to the started.

Hence, broad-spectrum antibiotic therapy is pequired (e.g. a third-generation cephalosporin, or a quinolone plus an antistaphylococcal antibiotic, or an antipseudomonal penicillin plus an aminoglycoside) and this treatment is thereafler tailored according to the results of investigations and the clinical response. Prolonged treatment for 4-weeks may be reqired in some patients with lung abscess and empyema.

Physiotherapy is of particular importance in the immobile and adequate oxygen therapy, fluid support and monitoring are essential.

Diagnostic bronchoscopy, with direct aspiration of purulent material from the parent bronchus; should be performed exept in older children who can cough up a goed sputum sample.

Aspiration of fluid form the pleural cavity.

Pus and other fluids can be aspirated from pleural cavity nader local anaesthesia.

A chest X-ray, posterior-anterior and lateral, will demonstrate the site of the fluid which is usually in the most dependent part of the chest posteriory. Percuss the chest to determine the upper level of dullness before starting.

Equipment reqired.

Have at hand, on a storile tray, a wide-bore needle 7,5-10 sm long with a 50 ml syringe and three way tap as well as kidney, dish or jug. Have ready any antibiotic which is to be instilled into pleural cavity.

Position of the patient.

The patient sits erect and paces his arms on a table or bench in front of him.

Site of aspiration.

This is usually in the eighth or ninth intercostal space in the posterior axillary line.

Procedure.

Use aseptic thechnique, scrub, don mask, goon, and surgical gloves, clean the patients skin with antiseptic and use sterile drapes. Infiltrate the skin with 0,5 percent - 1 percent, xulocaine (10-20 ml) down to pleura intercostally. As the pleura is infiltreted the accuracy of the siting can be checked by aspirating a small amonat of the fluid. Gently insert the wide-bore needle, apply the three-way tap so that fluid may be aspirated into the syringe, then close the tap so that fluid may be flushed into receptacle. As the fluid reduces in amount, gently with drow the needle and cover the wound with a sterile dressing. Carry out a check chest X-ray afterwards. Keep specimens of fluid in universal containers for bacteriology, histology or cytology if necessary. Remember it is best of contact the laboratory first to ascertain the volume of fluid they require.

Warning.

Aspiration of more than 600-700 ml at a time may be dangerous. Sudden mediastial shiff may result, leading to cardiac arrhythmias.

Insertion of an intercostal drain

This technique is indicated in pneumothorax, pyopneumothorax and haemopneumothorax.

Equipment required.

The correct side should first be identified by clust X-ray, but there may not be time for this in an emergency.

Have an Argyle drain with trochar owailable, antiseptic solution, swabs needles and syriuge, local anaesthetic, gauge 1/0 silk, Prolene, or nylon, scalpel, and dressing pack. In addition an underwater seal system, dreinage tubing, and clamp should be aveilable. Use an aseptic technique. Wear gloves and gown.

Sites of insertion.

- Outside the mid-clavicular line in the second costal space.
- Posterior axillary line via eighth or ninth intercostal space.

Identification of site.

The manubrium sterni is at the level of the second costal cartilage. Count the spaces form this level to identofy the site.

Procedure.

For pneumothorax a singh drain inserted through the second interspace is usually all that is required. When there is an accompanying pyothorax drains are often inserted at both sites – the lower to drain the pus.

Local anaesthetic is infiltrate into the skin over the second interspace. Then deeper infiltration of the muscles and parietal pleura can be achiened by inserfing the needle into the space over the rib (not underit) to prevent neurovascnlaris damage. Use the scalpel to cut the skin transversely, enough to permit indertion of the drain. Then insert the trochar and cannula (Argyle drain). Considerable pressure may be required and it is wise to hold the trochar and cannula with both hands, the right pushing and the left acting as a stop when the trochar penetrates the pleura. The left hand should be about two inches (5 sm) from the tip. When the pleura has been penetrated there is considerable “give”. Withdraw the trochar whilst threading the drain into the pleural cavity to the mark of the drain. Apply the champ across the drain and suture the skin using a purse-string suture. Leave the ends untied so that the suture can be tied on removal of the drain.

Now connect the underwater real drainage system, eusuring that it is belon the level of the patient (otherwise water may run into the pleural cavity). Secure the chest drain to the skin with suture. Apply the dressing and release the champ. Are there bubbles in the drainage system? Get a check chest X-ray to determine the position of the drain.

Removal of the drain.

In uncomplicated cases the lung usually re-expands in 24-48 hours, but many take longer. Take daily erect chest X-ray to check expansion. When complete clamp the drain and remove it 24 hours later, with a furtlur chest X-ray. Seal the wound with the purse-string and a dry dressing.

Bronchiectasis

Bronchiectasis, the term used to describe abnormal dilatation of the bronchi, may be produced in different waus. It may be acquired or, less commonly congenital.

Causes of bronchiectasis:

1. Congenital:
- Ciliary dysfunction syndromes:
  = primary ciliarly dyskinesia:
  = kartagenesis syndrome (+sinusitis+situs inversus),
  = joung's syndrome (Williams-Campbell syndrome – cogenital weakness of bronchial cartilaginous rings, leads to universal bronchiectasis)
- cystic fibrosis
- primary hypogammaglobulinaemia.

2. Acquired
- Pneumonia (complicating whooping cough or measles)
- primary tuberculosis
- foreigh body.

Symptoms of bronchiectasis.

1) *Due to accumulation of ps in dilated bronchi*.

- chronic productive cough usually wors in mornings and often brought on by changes of posture. Sputum often capious and persistently purulent in advenced disease.

2) *Due to inflammatory changes in lung and pleura surrounding dilated bronchi*.

- fever, malaise and increased cough and sputum volume when stread of infection causes pneumonia, which is frequently associated with pleurisy. Recurrent pleurisy in the same site often occurs in bronchiectasis,

3) *haemoptysis.*

- can be slight or massive and is often recurrent. Usually associated with purulent sputum or an increase in sputum purulence. Can, however, be the only symptom in so-called “dry bronchiectasis”.

4) *general health.*

When disease is extensive and sputum persistenly purulent a decline in general health occurs with meight loss, anorexia, lossitude, sleep swealing, and failure to thrive in children. In the patinets digital clubbing is common.

Chest radiographs are not usually diagnostic but may show increased bronchovascular marking in the affected lodes. Bronchiectasis was once diagnosis by bronchography, but CT is now the mainstay for diagnosis. Both tests show cylindric or saccular dilatations of the bronchi, with poling and poor clearance of contrast material form thise area.

Acute infections may be differentiated from bronchiectasis.

The condition most commonly occurs in the basilar segments at the lower lobes. The entire lower lobe, middle lobe, or lingula are sometimes involved,

The preffered treatment for bronchiectasis is medical, consisting of antibiotics, postural drainage, bronchoscopy with washing of bronchi from mucos and pus.

Schuster and Schwartz outlined the criteria for patient selection for surgery in bronchiectasis with cyctic fibrosis:

1. a radiographically localized area of bronchiectasis with disease advanced beyond that of the rest of the lung.
2. symptoms from the lung infection that interfere with patients well-being.
3. evidence of irreversibility
4. and ability of the patient to wilhstand thoracotomy and pulmonary resection.

Complication had been rare, consisting primarily of atelectasis that required postoperative bronchoscopy.

Whenever possible, a specific bacterologic diagnosis should be made before treatment.

Diagnostic bronchoscopy, with direct aspiration of purulent material from the parent bronchus, should be performed, except in older children who can cough up a good sputum sample.

In pneumonic, the chest radiograph shows a homogenous opacity localised to the effected lobe or segment.

Radiological examination is also particularly helpful if a complication such as pleural effusion, intrapulmonary abscess formation or empyema is suspected.

In mortality from pneumonia is high (approximately to 30%).

1) Lammer of breathing – abdominal respiration, thoracic respiration
2) Rate and Depth of respiration
3) Abnormal types of breathing.

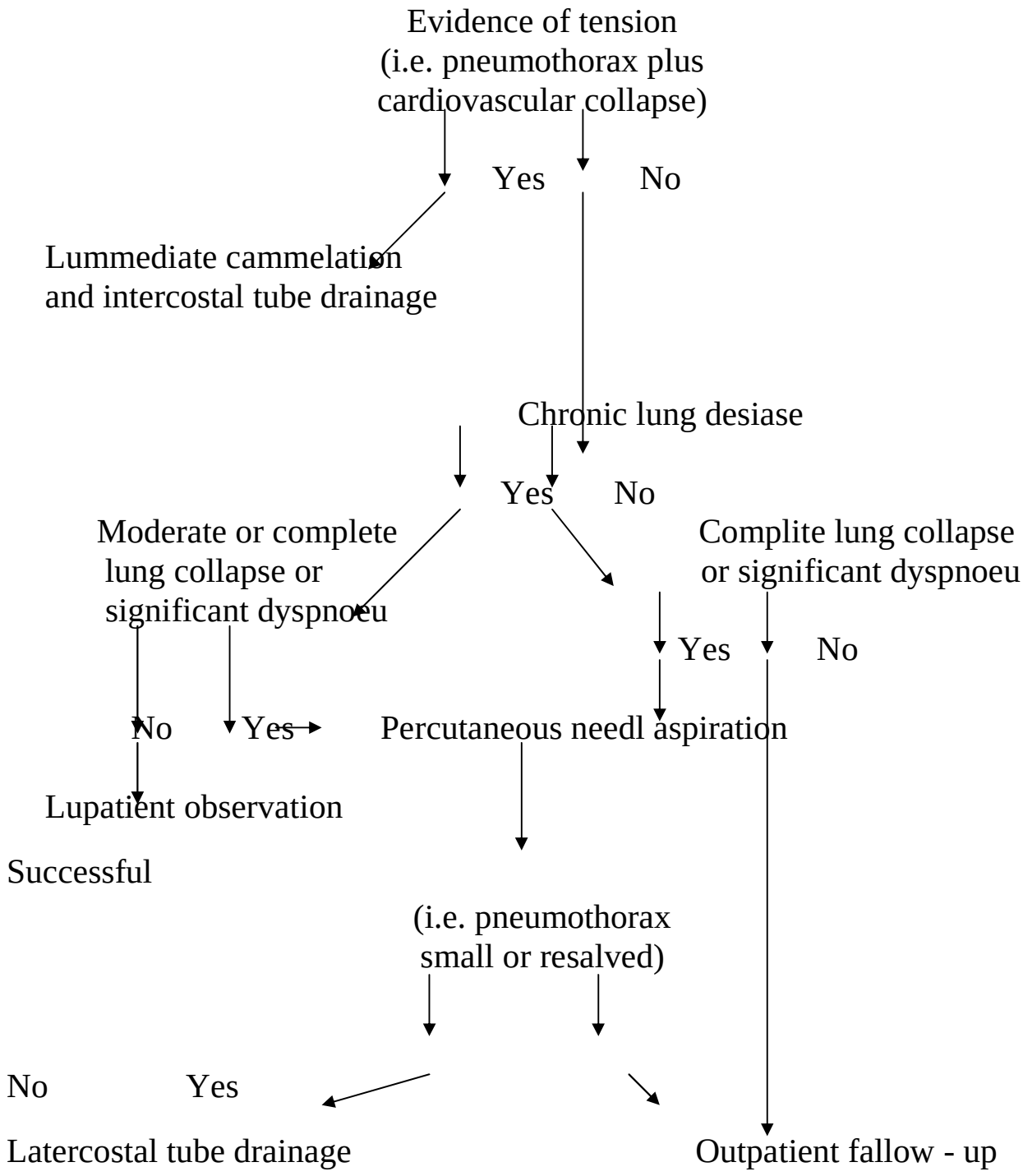
Saturation of the oxygen is in normal – 97%.

The partial pressure of O2 and CO2 in arterial blood:

Tension (pO_2) = 100 nm Hg

Tension (CO_2) = 0 nm Hg

Management of pneumothorax



CONGENITAL DISEASES OF THE KIDNEYS AND THE URETERS

Three kidney systems are distinguished onto – and phylogenetically: 1 - the fore kidneys or head kidney, the pronephros; 2 - the primary kidney or Wolffian body, the mesonephros; 3 - the permanent of final kidney, the metanephros.

The pronephros is a paired organ consisting of 6 to 10 paired excretory tubules (protonephrons) and is situated nearer to the anterior end of the body. By means of the renal funnels the protonephrons communicate with the secondary body cavity. Their opposite ends are connected with the primary excretory canal draining into the coelomic cavity.

The mesonephros appears on the third week when the pronephros has still not reduced. It is caudal to the pronephros and formed of segmented tubules also communicating with the excretory canal (Wolffian duct). Besides the Wolffian (mesonephric) duct a second paired, Muellerian (paramrsonephric) duct develops. The cephalic ends of the paramesonephric ducts communicate with the abdominal cavity, the caudal ends drain into the urogenital sinus. In males the Muellerian ducts reduce, while in females they give rise to the uterus, tubes and vagina. The Wolffian duct reduces in females but gives rise to efferent ducts of the tesus in males. The mesonephros reduces completely by the 12 th to 14 th weeks.

The metanephros develops in the caudal part of the embryo in the period of atrophy of the mesonephros from two buds, the metanephrogenic blastema and a diverticulum of the Wolffian duct. Any deviations of forming the kidneys give rise different malformations of the kidneys and the ureters.

Anomalies and malformations of the urogenital system are foremost among congenital diseases and account for over 40 per cent of them, i.e, approximately every one of fine newborn babies has a malformation of the urogenital system.

The danger of the development of severe complications which often threaten the patient’s life dictates the need to recognize these anomalies in the early period. Pyelonephritis is most common among these

complications; it usually occurs in babies with malformations of the urinary system (obstructive uropathy).

Test the urine of all patients. If indicated, read its volume in relation to time by means of a fluid chart.

Urine speciment

Obtain urine when the patient has a full bladder, usually first thing in the morning (exept for urine cytology). Take a midstream specimen, allowing about 150-200 ml to be voided before the sterile container is placed in the uninterrupted stream.

Retract the prepuce in the male, and wash the glans with saline. In women a tampon should be inserted, the labia parted and the periurethral area cleaned with sterile saline. Catheter specimens are much more liable to contamination and should be performed only if the patient is unable to cooperate.

Symptoms of urinary tract disease

The symptom suggestive of urinary tract disease include:

1. Disturbances in the act of micturition, including frequency, retention, incontinence, and disuria.
2. Alteration in the amount of urine.
3. Alteration in the appearance of the urine.
4. Pain: renal, ureteric, vesical, or urethral.
5. General symptoms of abnormal renal function.

1. **Disturbances in the act of micturition**

FREQUENCY (without increas in the amount of urine) results from irritation of the bladder.

NYCTURIA - the patient’s sleep is disturbed by the need to micturate.

RETENTION occurs in obstructive lesions of the urethra, in diseases of the spinal cord or sacral nerve-roots, and in coma due to various causes.

HESITANCY – delay in starting micturition.

INCONTINENCE of urine is common when the mental faculties are impaired. In spinal cord diseases, retention of urine is often followed by incontinence due either to overflow or to reflex evacuations of the bladder.

PRECIPITANCY – the sudden onset of micturition without warning, is another cause of occasional incotinence especially in neurological disorders.

DYSURIA means difficulty in micturation and may thus include some of the symptoms already discussed. More commonly, however, the term is used to describe pain or discomfort during the act of micturation and usually results from diseases of the bladder, prostate, or urethra.

2. **Alteration in the amount of urine**

The urine may be uncreased (polyuria), or diminished (aliguria), or absent (anuria).

POLYURIA – Large quantities of urine.

- as result of the ingestion of large quantities of liquid;
- may also result from nervousness;
- in chronic renal disease;
- in diabetic insipidus;
- dispersal of oedema

*OLYGURI*A and *ANURIA* (suppression of urine).

These symptoms are grades of the same condition, namely decrease in urinary secretion. Olyguria may be said to exist if the urinary output falls below 1/2 liter daily.

Care must be taken to prove – by catheterization, if necessary. That the cause of the dimished quantity is suppression, not retention of urine.

Suppression results from many conditions which impair the function of renal tubules or glomeruli. Examples are found in *PRERENAL* conditions causing a reduced blood-flow to the kidney (e.g. shock, haemorrhage, dehydration, and cardiac failure), primary RENAL disease such as acute glomerulonephritis, and in the late stages of urinary tract obstruction – *POSTRENAL*.

3. **Alteration in the appearance of urina**

It is only necessery here to point out that the patient may first suspect urinary disease by noticing alteration in the colour and general apperance of the urine, e.g. red in haemoturia, or cloudy in pyuria. Various drugs may colour the urine, causing alarm or sometimes delight if the patient thinks it reflects the efficacy of treatment.

INSPECTION - Inspection shows the colour of the urine and any deposit.

SPECIFIC GRAVITY – The normal specific gravity of urine is generelly about 1.015-to 1.025 (conveniently styled 1015-1025).

REACTION – The urine may be acid or slightly alkaline in health.

SMEU – A strong ammoniacal smell is frequently found in infanis.

QUANTITY - The amount of urine passed in twenty-four hours should be recorded. In health it averages 60-70 ml per hour.

The common abnormal constituents indicating the presence of urinary disease are protein, blood, and pus.

PROTEINURIA. – Protein excreted into the uriny by the kidneys otherwise fall into two classec. 1. Physiological proteinuria; 2. Pathological proteinuria. The distinction between these two is not always easy.

PHYSIOLOGICAL PROTEINURIA – This generally occurs in children or young adults, and the amount of protein is usually small, though occasionally considerable.

- arthostatic or postural proteinuria
- occasionally a few hyaline casts may occur

PATHOLOGICAL PROTEINURIA - This is generally larger in amount, though in some types of chronic renal disease it is common only to find on inconstant cloud of protein in the urine.

HAEMATURIA - Blood in the urine may vary in amount from large quantites visible to the naked eye to a few red corpuscles detectable only by microscopical examination.

Haemoturia – inicially (urethra)
- totally (kidneys, ureters, bladder)
- terminally (neck of bladder)

PYURIA - Larger quantities of pus usually indicate an inflammatory lesion of the urinary tract, and in most cases organisms are to be found on microscopical examination.

PNEUMATURIA - Gas is rarely expelled with the urine, but pneumaturia may occasionally prowe of considerable diagnostic value, susgesting a communication between the urinary tract (usually the bladder) and the alimentary tract (the colon).

Special investigations of value in the elucidation of urinary tract disease include radiography, cystoscopy, ultrasonography, chemical examination of blood and urine, tests of renal function, and renal biopsy.

Radiography

A direct radiogram of the abdomen will usually reveal the size and contour of the kidneys. The urinary passages, including the renal pelvis, the ureters, and the bladder, are best outlined by the intravenous injection of a radio-opegue dye which is excreted in the urine (intravenous pyelography).

Angiography

Abnormalities of the renal arteries or of the kidney itself can be demonstrated by the injection of dye into a catheter threaded thtough the femoral artery to the appropriate level in the aorta (renal angiography).

Cystoscopy

This is mainly of value in the diagnosis of bladder disease by direct vision and by biopsy.

Blood and urine chemistry

Disordered renal function is commonly associated with altered levels of urea and of electrolytes (sodium, putassium, chloride, and bicarbonate) and these can be measured in bath blood and urine. The commonest abnormality is an elevated blood-urea, but more important in the causation of symptoms are the disturbances of acid-base and electrolyte balance.

Test of renal function

Three main aspects of renal function can be studied: glomerular filtration, tubular reabsorption, and the differential function of the two kidneys.

The differential function of the two kidneys is assessed by comparing by scanning of the two kidneys with an external counter following the intravenous injection of radio-isotope which is excreted in the urine (radioactive renography).

Renal biopsy

The histological diagnosis of diffuse renal disease can often be estasblished by needle biopsy. This method is of particular value in determining the cause of a nephrotic syndrome.

Ultrasonography – may to reveal the general signs of inflamattery and congenital diseases of the kidneys, the ureters and bladder.

PYELONEPHRITIS

This is infection of the renal pelvis, calyces, and parenchyma which can originate from haematogenous or ascending routes.

ASSOCIATIONS: - Ureteric reflux (35 per cent of patients)
- Urinary tract abnormalities.
- Chieldhood, preguancy.

CAUSAL ORGANIZM: - Coliforms, usually streptococcus faecalis, Pseudomonas pyocyanea; Proteus, and Klibsiella.

Clinical features

ACUTE. Prodromal symptoms include headache and malaise. The onset is often sudden with rigors, vomiting, pain in the hypochondria or renal angles, and painful micturation (often described as „scalding" or „burning"). Uraemia can develop in cases associated with obstruction. Severe cases develop abdominal rigidity and hyperpyrexia (temperature greater than 40˚C).

DIAGNOSIS. – Is by clinical features but may not always be obvious, intravenous urogram may show limited excretion on the affected side with occasional mild extravasation.

- Radioisotope renography may show scattered uptake on the affected side.
- Midstream urine specimen (+microbiology+sensitivity).

TREATMENT: - Bed rest and increased oral fluid.
- Alkalinization of acid urine.
- Analgesia
- Broad-spectrum antibiotics until sensitivities are available.

CHRONIC. This may develop „ab initio" or after multiple acute attacks, and is a cause of death from uraemia.

PATHOLOGY: Females: males 3:1 The lesions are asymmetrical and involve predominantly the renal tubules which become progressively atrophic.

CLINICAL FEATURES. Intermittent pyrexia, progressive anemia, hypertension (40per cent), lumbar pain, frequent painful micturation, general malaise.

DIAGNOSIS. Midstream urine specimen: Pyuria is common. White blood cells predominate (the count varies). Bacteriology usually reveals E. Coli. Strep.faecalis, Proteus or Pseudomenas.

- *Intravenous urogram*: Poor definition and rimming (narrowing) of the renal cortex are important features.
- *Cysto-urethroscopy:* Features include chronic urethritis, trigonitis, stricture, prostatic hypertrophy, posterior urethral valves.
- *Renal function tests:* Plasma creatinine, urea, and electrolytes.

AIMS OF TREATMENT:

- Eradicate predisposing factors, e.g. correct urinary tract abnormalities.
- Eradicate local infection.
- Eradicate distant foci of infection as above.
- When unilateral and causing hypertension, nephrectomy may be curative provided the remaining kidney is functioning.
- Patients with advanced bilateral disease may need dialysis or renal transplantation.

Classification of anomalies of the kidneys and ureters

I. Anomalies of the kidneys

1. Anomalies in the number of kidneys
   - agenesis of kidneys
   - singular kidney
   - supernumerary (plural) kidneys
2. Anomalies in the size and structure of the kidneys.
   - hypoplasia of the kidney with or without dysplasia.
   - solitary cyst of the kidney
   - polycystic of the kidney
   - multicystic of the kidney.
3. Anomalies in the interrelation of the kidneys.
   - symmetric forms (horseshoe kidney, cake kidney)
   - asymmetric forms (S- and L- shaped kidneys).
4. Anomalies in the position of the kidneys (dystopia of the kidneys)

- thoracic dystopia
- lumbar dystopia
- iliac dystopia
- pelvic dystopia
- crossed dystopia

II. Anomalies of the ureters.

1. Duplication of the ureters.
   - complete duplication (uni – or bilateral)
   - incomplete duplication (uni – or bilateral)
   - ectopia of the ureteral orifice in patient with complete duplication of the ureters (uni – or bilateral).
2. Anomalies in the position of the ureters
   - retrocaval ureter
3. Anomalies in the structure of the ureter
   - neuromuscular dysplasia (megaureter)
4. Anomalies in the start of the ureters (hydronephrosis).
   - stenosis of the pelvioureteral segment
   - aberrant kidney's arteria which squeezes superior part of the ureter.
   - highly going out ureter from the pelvic.
   - fibrosis of the retroperitonial tissue which squeezes superior part of ureter.
   - fixed ureteral kink
   - valve of ureter.
5. Anomalies in the end of the ureters.
   - stenosis of the ureteral orifice (ureterocele, hydroureter)
   - insufficienty the intramural part of the ureter (vesicoureteral or vesicorenal reflux).

Agenesis. The absence of the germ of the organ is encountered in one of 1000 newborn babies. Bilateral renal agenesis is less frequent than unilateral (a ratio 0f 1:4) and is mostly encountered in male faetuses (a ratio 3:1). Babies with agenesis of both kidneys (arenia) incapable of living and are usually born dead. The solitary kidneys is suspected when an enlarged and non-tender organ is palpated. It is usually hypertrophied and

fully accomplishes the excretory function. The anomaly is asymptomatic in such cases.

Supernumerary kidney. This is a very rare anomaly. A supernumerary kidney must be differentiated from supperior segment of a duplicated kidney. A supernumerary kidney is important clinically only when the uteral orifice as ectopic (permanent urine incotinence) or in involvement in an inflammatory, neoplastic or some other pathological process.

Hypoplasia of the kidney, congenital diminution in size, is associated mainly with disturbed development of the metanephrogenic blastoma as the result of deficient supply of blood. The simple form of hypoplasia is marked only by a reduced number of calyces and nephrons. In the second form, the decrease in the number of glomeruli is combined with their increased diameter, fibrosis of interstitial tissue, and dilated tubules. Hypoplasia with dysplasia is manifested by the development of connective tissue or muscular cuffs around the primary tubules. Unilateral renal hypoplasia may produce no signs whatsoever throughout life, but it has been noted that a hypoplastic kidney is not infrequently affected by pyelonephritis and is often the source of nephrogenic hypertension. Unilateral hypoplasia complicated by pyelonephritis and hypertension is usually managed by nephrectomy. Bilateral nephrectomy with subsequent kidney transplantation is only life-saving measure in bilateral kidney hypoplasia complicated by severe renal insufficiency.

Solitary cyst. This is a solitary round or oval cystic structure arising from the renal parenchyma and protruding above its surface. Dull pain in the region of the kidney, a palpable tumour, and transitory haematuria are the most characteristic signs of a solitary cyst and are indication for surgical treatment. Pain increases and body temperature rises in suppuration of the cyst.

Polycystic disease of the kidneys (polycystic degeneration, polycystic kidneys) is a hereditary anomaly with involvement of both kidneys. The manifestations of polycyctic kidneys vary with the age group. The earlier the signs appear, the more malignant is the condition. In very young children the disease is often complicated by severe pyelonephritis leading to anuria and uraemia. In polycystic kidneys in older children the condition is marked by dull lumbar pain, periods of haematuria and arterial

hypertension (in 70 per cent). Polyuria, hypoisosthenuria, and nocturia are observed. Enlarged and tuberous kidneys are palpated. Any surgical intervention considerably aggravates the condition of a patient with polycystic kidneys. It becomes necessary in profuse renal haemorrhage, obturation by a stone, pyonephrosis or the development of a malignant tumour of the kidney. Chronic haemodialysis and kidney transplantation are resorted to in the terminal stage or renal failure.

Sponge kidney (medullary sponge kidney, kidney with spongious pyramids, Cacchi-Ricci disease) is a rather rare congenital hereditary anomaly marked by cystic dilation of the collecting tubules of the pyramids. The anomaly usually produces no clinical signs if complicatons in the form of nephrocalcinosis, calculosis or pyelonephritis do not occur. In the absence of complications a sponge kidney needs no treatment.

Multicystic dysplasia. This is an anomaly in which one or much less frequently, both kidneys are replaced by cystic cavities and are completely devoid of parenchyma. A bilateral anomaly is incompatible with life. In a unilateral lesion, complaints occur only when the cysts grow and exert pressure on the surrounding organs, in which case nephrectomy has to be performed.

Fusion of the kidneys accounts for about 13 per cent of all renal anomalies. Symmetric and asymmetric forms of fusion are distinguished. Horseshoe and cake kidneys are related to the first form, S-shaped, or sigmoid kidney, and L-shaped, or angulated kidney, to the second form. Horseshoe anomaly is marked by fusion of the upper or lower poles of the two kidneys. The renal parenchyma in such cases is shaped like horseshoe. The lower poles fuse more frequently (in 90 per cent of cases) than the upper poles (in 10 per cent). Rovsing’s sign (appearance of pain in extention of the trunk) is the main clinical manifestation of a horseshoe kidney. The pain is caused by compression of the vessels and aortic plexus by the isthmas of the kidney. The pain is often vague and attended by dyspeptic phenomena. Operation is usually performed for a horseshoe kidney only if complications (hydronephrosis, stones, tumor. etc.) develop.

A cake kidney is a flat oval organ situated on a level with the promontorium or lower. The anomaly is encountered at a rate of 1:26.000.

Asymmetric fusion accounts for 4 per cent of all renal anomalies. Fused ectopic kidneys may compress the adjoining organs and large vessels and cause ischaemia intermittens and pain. Quite often the anomalous kidney is congenitally maldeveloped or involved in a cystic process. Lithiasis and pyelonephritis may develop in a fused kidney due to the abnormal arrangement of the vessels, pelves, and ureters.

Dystopia (ectopia) of the kidneys. This term designates a malposition of the kidneys due to disturbed ascent during embryogenesis. The incidence of the anomaly is 1:800 on the average. Dystopia of the kidneys is encountered more frequently in males. In thoracic dystopia the kidney is usually a companent of the diaphragmatic hernia. Its ureter is elongated and drains into the bladder, the artery arises from thoracic aorta. In lumbar dystopia the renal pelvis is rolated slightly to the front and situated on a level with the fourth lumbar vertebra. Mobility of the kidney is limited.

Iliac dystopia is characterized by more pronounced forward rotation of the pelvis and its location on a level between the fifth lumbar and the first sacral vertebral. The kidney hardly moves with a change in the body position.

A pelvic kidney is situated on the midline under the aortic bifurcation, to the back of and slightly above the urinary bladder. Its shape may be most quaint. It is hypoplastic to some degree as a rule.

Crossed dystopia is characterized by contralateral displacement of the kidney. Both kidneys fuse in this case as a rule to form an S- or L- shaped kidney. The ureter draining the dystopic kidney opens into the bladder in the normal place. The incidence of crossed renal dystopia is 1:10.000 – 1:12.000.

Duplication of the kidneys and ureters are characterized by the presence of two pelves in a single mass of renal parenchyma and two ureters. The ureters arising from the pelves of a duplicated kidney stretch one next to the other, often in a common fascial sheath, and open into the bladder either separately or fuse on some level to form a single trunk. In complete duplication of the ureter, the main trunk arising from the inferior segment of a duplicated kidney opens in the corner of the Lientaud triangle, while the second trunk opens next to or distal to it (Weigert-Meyer’s law). The orifice of a duplicated ureter is often narrowed, which leads to the formation of a cystic cavity protruding into the lumen of the

bladder – ureterucele. A person with a duplicated kidney and ureter may live a long life without any complaints or clinical manifestations if no hidrance to the passage of urine from kidney.

In **ectopia of the ureter** its orifice opens distal to the corner of the trigone of the bladder or drains into the adjacent organs. Ectopia is mostly encountered in complete duplication of the pelvis and ureter, the ectopic ureter is usually the one which drains into superior pelvis. According to statistical data in 10 per cent of cases with duplicated ureter the orifice is ectopic, in girls 4 times more frequently than in boys. In girls the displacement may occur into the derivatives of urogenital sinus (neck of the bladder, urethra, vestibulum of the vagina) and the Miillerian ducts (vagina, uterus); in boys the orifice is displaced into the derivatives of the Wolffian ducts (posterior urethra, seminal vesicles, ductus deferens, epididymis). The patient's complaints and clinical picture are determined by the type of ectopia. In cervical and urethral ectopia the main complaint is incontinence of urine but the urge to urinate is maintained and micturition is normal. In contrast to girls boys with an ectopic ureter usually do not suffer from incontinence of urine. Ectopy of the ureteral orifice is managed surgically.

Megaureter is a congenital disease marked by considerable dilatation and elongation of the ureter. The disease may be caused by: neuromuscular dysplasia of the ureteral wall for the entire length or on a limited area; 2. vesicoureteric reflux; 3. obstruction of the distal segment of the ureter. Neuromuscular dysplasia is characterized by deficiently developed muscle fibres and lack or absence of nerve ganglia. As a cause of megaureter vesicoureteric reflux may result from embryonic underdevelopment of the distal part of the ureter and the presence of infravesical obstruction (sclerosis of the bladder neck, hypertrophy of the urethral crest or urethral valve). Cicatricial stenosis, ureterocele or a valve are responsible for obstruction of the distal part of the ureter. The clinical picture is mainly represented by signs of pyelonephritis developing as a complication in megaureter; it does not respond to drug therapy. Treatment is a diffucult task. The congenital deficiency or gross anatomical and physiological disorders of the ureter which are consequent upon severe pyelonephritis and ureteritis, make it hardly fit for plastic surgery. Modelling and implantation of the ureter into the bladder by the antireflux technique is the

plastic operation performed most widely in megaureter. Continuons and prolonged medical observation over the children with megaureter, urinalysis control, and regular radiourological examinaton are necessary.

Hydronephrosis is progressive dilatation of the renal pelvis and calyces as a consequence of obstruction to the passage of urine from the kidney. The disease in children is congenital as a rule. Unilateral hydronephrosis prevails, a bilateral process is encountered in only 6 to 9 per cent of cases. The disease occurs more often in girls (1,5:1). The causes of congenital hydronephrosis are: 1. stenosis of pelvioureteral segment; 2. accessory (aberrant) vessel; 3. fixed ureteral kink; 4. high origin of ureter; 5. embryonic adhesions; 6. valve of the ureter. In clinical picture the pain, the palpable tumour sign and abnormalities in the urine are the main manifestations of hydronephrosis. Treatment in hydronephrosis is surgical. In view of the considerable compensatory and regenerative capacities of the child's organism a minimally functioning organ should be preserved and one that has died completely is removed.

The orifice of a ureter is narrowed, which leads to the formation of a cystic cavity protruding into the lumen of the bladder – ureterocele and hydroureter. Treatment in these cases are surgical.

Vesicoureteric reflux (vesicoureteric regurgitation of urine, vesicorenal reflux) is found in 35 per cent of children suffering from pyelonephritis. Primary and secondary forms are distinguihed. Primary vesicoureteric reflux results from immaturily of anatomical structures in the region of the ureteral orifice which normally prevent regurgitation of urine. The secondary form is associated with infravesical obstruction and a neurogenous urinary bladder. Vesicoureteric reflux is marked by signs of chronic pyelonephritis. Some older children may experience pain in the side or small of the back during urination, which is explained by overstretching of the renal pelvis by urine regurgitated under pressure. The diagnosis is based on the finding of micturating cystography. Five degrees of the reflux are distinguished according to its severity. In I degree urine returns only to the distal parts of the ureter. In II degree the contrast medium fills the kidney pyelocalyceal system in which X-ray shows no abnormalities. Reflux of III degree is characterized dy a moderately distended ureter and rounded calyceal fornices, the pelvis is slightly stretched. In IV degree the cavities of the kidney are markedly dilated and

the ureter is distended and tortnous. In V degree the reflux is manifested by a typical picture of megaureter: a sharply distorted irregularly distended and considerably elongated ureter and ectatic calyces surrounded by a band of thin renal parenchyma are found. The removal of the infravesical obstruction is an indispensable condition. In IV-V degrees reflux one-stage operation for resection of the terminal part of the ureter and reimplantation into the bladder after the Palitano-Leadbetter antireflux method is performed. In case of III degree reflux Gregoir's operation (extravesical implantation of the ureter under detrusor) is conducted. Children with I-II degrees reflux are treated by non-operative measures.

All these congenital diseases of kidneys are recognized during excretory urography, scanning of the kidneys and micturating cyctography.

MEGACOLON. HIRSCHSPRUNG'S DISEASE

Megacolon is abnormal dilatation and hypertrophy of a greater or smaller part, or of the whole length of the large intestine. This is a collective term for diseases differing in origin and pathoanatomica1 essence whose main sign is persistent constipation. Megacolon can be caused by deficient development of the intramural nervous system of the large intestine, developmental anomalies of the anorectal region (fistular forms of rectal atresia), lesions of the central nervous system, endocrine disorders, avitaminosis, psychogenic factors responsible for disturbance of normal defaecation, etc. In view of all said above, the following rational classification of megacolon (Yu. F. Isakov and **A.** I. Lenyushkin) acquires considerable importance.

Causes	Congenital	Acquired
Of functional character: without affection of intramural ganglia	Hirschsprung's disease	Hypovitaminosis B1**.** Chagas' disease. Infectious diseases
with affection of intramural ganglia or with their secondary involvement	Idiopathic megacolon	Psychogenic constipation, endocrine disorders. Effect of drugs. Anal fissures
Of mechanical character	Anorectal developmental anomalies	Cicatricial stenosis of rectal region after burns, injury, inflammatory processes, etc.

Pathogenesis

In his classic textbook titled *Pediatric Surgery,* Dr. Orvar Swenson—who is eponymously associated with one of the classic surgical treatments for Hirschsprung's disease—described this condition as follows: ". . .

congenital megacolon is caused by a malformation in the pelvic parasympathetic system which results in the absence of ganglion cells in Auerbach's plexus of a segment of distal colon. Not only is there an absence of ganglion cells, but the nerve fibers are large and excessive in number, indicating that the anomaly may be more extensive than the absence of ganglion cells." This description of Hirschsprung's disease is as accurate today as it was nearly 50 years ago, and summarizes the essential pathologic features of this disease: absence of ganglion cells in Auerbach's plexus and hypertrophy of associated nerve trunks. The cause of Hirschsprung's disease remains incompletely understood, although current thinking suggests that the disease results from a defect in the migration of neural crest cells, which are the embryonic precursors of the intestinal ganglion cell. Under normal conditions, the neural crest cells migrate into the intestine from cephalad to caudad. The process is completed by the twelfth week of gestation, but the migration from midtransverse colon to anus takes 4 weeks. During this latter period, the fetus is most vulnerable to defects in migration of neural crest cells. This may explain why most cases of aganglionosis involve the rectum and rectosigmoid. The length of the aganglionic segment of bowel is therefore determined by the most distal region that the migrating neural crest cells reach. In rare instances, total colonic aganglionosis may occur.

The first description of a child with Hirschsprung's disease appeared in the eighteenth century by Domenico Battini, whose description of a typical child with congenital megacolon was published after Battini's death in 1800. In 1886, Harald Hirschsprung, a Danish pathologist, described several cases of the condition that ultimately bore his name. During subsequent years, surgeons were taught to resect the grossly abnormal colon and perform an anastomosis to the rectum (which rarely worked) or a colostomy. In his textbook on pediatric surgery published in 1926, Fraser said: This is an obscure disease of the large intestine, in which the essential features are an inability of the colon to part with its contents. The only curative treatment which the pathology and clinical course of the affection show to be applicable is that of excision of the affected bowel with the union of unaffected gut above to the upper end of the rectum below.

Between the turn of the nineteenth century and the 1940s, a number of papers were published that observed abnormalities in the innervation of the colon, but the absence of ganglion cells that we now consider to be the sine quo non of Hirschsprung's disease was not widely recognized until 1948, when Whitehouse and Kernohan summarized the literature and presented a series of cases of their own. Shortly thereafter, Swenson confirmed that aganglionosis was the cause of obstruction in these children and recommended rectosigmoidectomy as the optimal treatment of this disease. Although initially this operation was performed without decompressing colostomy in most children, technical difficulties in small infants and the debilitated and malnourished state in which many children presented caused most surgeons to adopt a multistaged approach with colostomy as the initial step. In more recent years, advances in surgical technique and earlier diagnosis have resulted in an evolution toward one-stage and minimal access procedures for the treatment of this disease.

Recent studies have shed light on the molecular basis for Hirschsprung's disease. Patients with Hirschsprung's disease have an increased frequency of mutations in several genes, including *GDNF*, its receptor *Ret*, or its coreceptor *Gfra-1*. Moreover, mutations in these genes also lead to aganglionic megacolon in mice, which provides the opportunity to study the function of the encoded proteins. Initial investigations indicate that *GDNF* promotes the survival, proliferation, and migration of mixed populations of neural crest cells in culture. Other studies have revealed that *GDNF* is expressed in the gut in advance of migrating neural crest cells, and is chemoattractive for neural crest cells in culture. These findings raise the possibility that mutations in the *GDNF* or *Ret* genes could lead to impaired neural crest migration in utero, and the development of Hirschsprung's disease

Megacolon consequent upon congenital agangliosis of the large intestine is of most clinical significance. It is very important in practice and the term Hirschsprung's disease is often used as a synonym of megacolon.

It was believed for a long time that Hirschsprung's disease was extremely rare (1 : 20 000-1 : 30 000). In recent years it is found more frequently, which is evidently due to earlier and exact diagnostics. Its incidence among boys is 4 to 5 times that among girls. The hereditary

(genetic) nature of this severe malformation has been proved at the current stage.

It was believed for many decades that Hirschsprung's disease was caused by congenital malformation of the muscular components of the large intestine, the presence of mucosal folds in its distal part, kinking of the elongated sigmoid colon and its congenital atonia, altered sympathetic tone, etc.

Changes of the histostructure of the intramural nerve apparatus in a definite segment of the large intestine are the principal element in the pathogenesis of Hirschsprung's disease (0. Swenson; Yu. Isakov). Morphological studies showed considerable changes of ganglia not only in Auerbach's (intermuscular) but in Meissner's (submucous) plexus in the constricted zone. The results of these studies allow this form of megacolon to be characterized as congenital agangliosis of
an area of the large intestine, a condition in which peristalsis cannot be produced in intestinal areas devoid of Auerbach's plexus or in those with its deficiency.

Severe changes and even death of the muscular layers of the aganglionic zone aggravate the peristaltic disorders and make them permanent. In accordance with these disorders, the zone is aganglionic morphologically and aperistaltic clinically.

Absence of peristalsis in this intestinal segment results in stasis of the faecal material above the site of the affection, which in turn causes dilatation and hypertrophy of the proximal parts of the large intestine. Hypertrophy develops as a consequence of intensive peristalsis of the proximal segments to propel the contents through the aperistaltic aganglionic area. The diameter of the dilated intestine may be very large. The aganglionic segment, in contrast, is narrowed.

Study of the pathological anatomy of the large intestine in congenital agangliosis showed prevalent involvement of the rectosigmoid segment (70 per cent) and the perineal and ampullar parts of the rectum (20 per cent). The occurrence of aganglionic zones in two places and in the proximal parts of the large intestine is much rarer. Total affection of the large intestine with agangliosis is known.

Clinical picture and diagnosis

Failure to pass stool (chronic constipation) is the principal sign of Hirschsprung's disease. The disturbed activity of the gastro-intestinal tract is manifested by constipation from birth or the first months of life as a result of which
chronic faecal toxicosis gradually develops. Meteorism occurring, like constipation, in the first days and weeks of life is a constant sign of the disease.

With the gradual chronic retention of faeces and gases, the sigmoid colon and later the proximal segments of the colon become dilated as a consequence of which the abdomen becomes larger. The high position of the diaphragm leads to the formation of a barrel chest.

In prolonged and persistent retention of faeces and gases, the abdomen grows still larger and acquires the form of a "frog" abdomen**.** The abdominal wall is thin. The distended intestinal loops are outlined and their intensified peristalsis can be seen now and again. The characteristic "clay" sign is found by abdominal palpation in most cases (clearly defined depressions felt through the skin of the abdominal wall form from pressure with the fingers on the intestine overfilled with faeces). It must be pointed out that the child's general condition also suffers in such a case, he is retarded physically and mentally, and anaemia and cachexia develop. These signs are more pronounced in older children. Inflammation of the intestinal mucosa, manifested by diarrhoea, occurs sometimes as the result of dysbacteriosis.

The initial manifestations of constipation and their subsequent character and persistence are determined to a great measure by the length of the aganglionic segment, the character of feeding, and the compensatory capacities of the intestine.

Severe neglected forms of Hirschsprung's disease are rarely encountered
nowadays. Elaboration of methods for its early diagnosis even in the neonatal period was conducive to this. Three forms of the clinical course of Hirschsprung's disease are distinguished: severe (acute), moderately severe (subacute), and mild (chronic).

The *severe,* or *acute,* form of congenital agangliosis is manifested from the first days of life as low intestinal obstruction. Meconium is

retained or passed in very small amounts, gases are not passed. Abdominal distension increases progressively, intestinal peristalsis is visible, and copious vomiting occurs. Percussion produces tympany due to the sharp meteorism. Enemas given regularly prove ineffective and the passage of gas is poor. Children are often admitted to the
hospital for suspected high intestinal obstruction.

The discrepancy between the clinical signs characteristic of high intestinal obstruction (vomiting of bile) and the X-ray evidence of impaired evacuation of the large intestine (distension of the loops and the presence of air in them) gives rise to the suspicion that the child is suffering from Hirschsprung's disease and prompts the physician to conduct thorough clinical and X-ray examination.

Hirschsprung's disease in the newborn is managed by non-operative methods as a rille, and only in rare cases when they prove ineffective and the patient's
condition grows worse rapidly and toxicosis increases a faecal fistula of the ascending colon is established.

The *moderately severe,* or *subacute form* is usually an intermediate stage between the severe and mild forms and develops when the aganglionic zone is smaller in length and forms no kinks. The general condition deteriorates slowly but continuously. Constipation becomes more and more stubborn. Non-operative measures produce a temporary effect. Siphon enemas are resorted to more often to evacuate the intestine. The child's condition changes depending on the degree
of stool retention. Most children are physically retarded, lose weight, and signs of toxicosis and anaemia develop.

The *mild, or chronic form*. Patients with this form hardly differ from healthy babies in the first days and sometimes weeks of life. The stool may occasionally be retained and mild abdominal distension and vomiting occur in attendance, but the general picture causes no alarm, the more so since stool is passed after a small enema or introduction of a colonic tube. But the child's condition grows worse
when artificial feeding is given, and stool is passed only after a cleansing enema. Faecal stones form in prolonged coprostasis due to inadequate care. They are so large in neglected cases that are mistaken for an abdominal tumour. The general condition gradually deteriorates, which is linked with

chronic faecal toxicosis. Anaemia and hypotrophy are, however, mild. The abdomen is usually distended,
enlarged transversely, and spread out. Peristalsis of the dilated loops of the large intestine is seen in occasional cases. Digital rectal examination shows increased tonus of the sphincter muscle.

A chronic course of Hirschsprung's disease is due to a short aganglionic zone in which the functional disorders are mild. Forms with an aganglionic zone of a greater length (from the descending colon to the rectal ampulla) are, however, also encountered and are marked by gradual development of the disease. The final diagnosis is made only from the results of X-ray examination. Much importance must be attached to functional methods of examination of the rectum (measurement of intraintestinal pressure, study of the condition of the sphincter ani externus and internus muscles, etc.).

X-ray examination is undertaken after the intestine is completely evacuated of faecal material. Scout radiography of the abdominal cavity usually reveals dilated and distended loops of the large intestine and a high position of the dome of the diaphragm. Radiocontrast study by means of an enema (irrigography) is the only method demonstrating the most characteristic signs. A barium suspension in a 1 per cent common salt solution is used as the contrast medium; 30 to 80 ml is a sufficient amount for the newborn and infants, older children are given up to 500 ml by enema. A narrowed zone in the large intestine with a suprastenotic dilatation proximal to it is an authentic X-ray sign of Hirschsprung's disease.

The most characteristic changes are demonstrated on lateral view radiographs showing not only the localization of the narrowed aganglionic zone but the degree and length of the narrowing. Evacuation of the intestine is delayed after X-ray rectal examination. This is one of the main diagnostic signs of Hirschsprung's disease in the newborn babies and infants, in whom the difference in the diameter of the stenosed and dilated parts of the intestine is still negligible.

Faecal stones can sometimes be demonstrated by X-ray. In contrast to Hirschsprung's disease, in other types of megacolon a constricted aganglionic area is not found while the large intestine is distended and in some cases also elongated.

Since Hirschsprung's disease is attended by other malformations, particularly often by diseases of the urinary system, urological examination along with irrigography must be conducted in all cases.

Differential diagnosis

The differential diagnosis of Hirschsprung's disease is made with developmental defects and some diseases attended by constipation.

Congenital elongation, dilatation or atonia of the large intestine differs from Hirschsprung's disease in having a later onset (from the age of 2-3 years) and less pronounced signs. With a change in the diet, stool is passed spontaneously at regular intervals, which never happens in children over 12 months of age suffering from Hirschsprung's disease. The abdomen is usually normal in shape and size. The general condition is never disturbed like in Hirschsprung's disease. **A** wide rectal ampulla filled with faeces is found on digital examination of the rectum.

Secondary megacolon consequent upon congenital or acquired cicatricial stenosis of the rectum is revealed by examination of the anal region and digital rectal examination. Sigmoidoscopy is performed in some cases.

Common constipation caused by anal fissures, haemorrhoids, and other diseases are diagnosed from careful analysis of the medical history and the findings of examination of the anal region. Such constipation is managed by removing the cause.

The practical importance of differential diagnosis consists in early recognition and treatment of Hirschsprung's disease since it is the most severe sickness. Biopsy of the rectum is resorted to in questionable cases; it will reveal an absence of nerve ganglia in Hirschsprung's disease.

Anorectal manometry aids in the diagnosis of Hirschsprung's disease through identification of the rectoanal inhibitory reflex, which is present in normal individuals but absent in the vast majority of children with Hirschsprung's disease. Although anorectal manometry is possible in newborns, it is not widely available for this age group and may be unreliable. In older children, the test is technically easier, but false-positive results may occur due to masking of the relaxation response by contraction of the external sphincter.

Definitive diagnosis of Hirschsprung's disease is based on histologic evaluation of a rectal biopsy, looking for the presence or absence of

ganglion cells and the finding of hypertrophied nerve trunks. The biopsy is usually taken 1-2 cm above the dentate line; going too distally may result in a false-positive diagnosis of Hirschsprung's disease because ganglion cells may normally be absent in this area. The most common technique for rectal biopsy is a suction device that samples mucosa and underlying submucosa. Many pathologists believe that a suction rectal biopsy lacking ganglion cells is consistent with (but is not diagnostic) of Hirschsprung's disease. Evaluation of suction biopsies may be enhanced by staining for acetylcholinesterase, which has a characteristic staining pattern in the submucosa and mucosa. Other stains, such as glial fibrillary acidic protein, have also been described for the diagnosis of Hirschsprung's disease, but have not been widely adopted. Punch biopsies or full-thickness biopsies, which may provide more tissue and deeper levels, may be needed if the suction biopsy sample is inadequate. Some surgeons prefer these techniques as the first choice

Treatment

There is full agreement today that Hirschsprung's disease must be treated surgically. Abdomino-perineal resection of the narrowed segment and part of the distended large intestine proximal to it is the radical and pathogenetically substantiated operation. The best time for its performance is between the age of 2 and 3 years.

Much significance is attached to *non-operative mamgement* in the period preceding the radical operation. It is usually conducted at home and its object is regular evacuation of the intestine.

An aperient diet is prescribed according to the child's age, in which food activating intestinal peristalsis is included (oatmeal arid buckwheat porridge, prunes, beetroot, carrots, apples, honey, etc.). Sour-milk products (sour clotted milk, sour fermented milk, fresh yoghurt) have a beneficial effect. Massage of the abdomen and a complex of exercises for strengthening the abdominal muscles are necessary. The parents must be instructed in these simple manipulations and carry them out daily for 10 to 15 minutes before each feeding.

Various types of enemas (a common cleansing enema, hypertonic, vaseline, siphon, etc.) are the principal measures in non-operative treatment of Hirschsprung's disease. **A** 1 per cent common salt solution of room temperature is used for a siphon enema. Warmed water must not be used

because in improper evacuation of the intestine the broken-up faecal material is rapidly absorbed and toxicosis and oedema of the brain develop which may cause sharp deterioration of the condition and even death. The volume of the siphon enema is determined by the child's age (from 0.5 to 2-3 litres for infants and 3 to 10 litres for older children). After the enema, a colonic tube is advanced through the narrowed zone into the dilated part of the large intestine and left in place for 1-2 hours. Laxatives are not advisable, oral vegetable oils (peachkernel, sunflower, olive oil) are preferable (1 teaspoonful for children of nursery age, 1 dessertspoonful for those of preschool age, and 1 tablespoonful **3** times daily for schoolchildren).

Intensive non-operative measures, even if applied in an in-patient clinic, fail to produce the needed effect in some cases and the child's condition grows worse progressively. The formation of a preternatural anus is indicated in such patients.

Stretching the anus with the fingers under anaesthesia improves the passage of the intestinal contents in considerably increased tonus of the sphincter ani internus muscle. The manipulation is repeated at intervals of 10-14 days.

Surgical options

The goals of surgical management for Hirschsprung's disease are to remove the aganglionic bowel and reconstruct the intestinal tract by bringing the normally innervated bowel down to the anus, while preserving normal sphincter function. There have been many operations devised to accomplish these goals, but the most commonly performed at the present time are the Swenson, Duhamel, and Soave procedures. There are no prospective controlled series comparing surgical treatments of Hirschsprung's disease. It is therefore difficult to determine if there are any significant advantages to one over the others. It is probably true that surgeons will get the best results doing the operation they have been trained to do and do with some frequency.

The Swenson procedure is essentially a low anterior resection of the rectum with an end-to-end anastomosis performed by prolapsing the rectum and pulled-through bowel outside the anus. A number of publications have documented excellent results from this approach,

including a more recent long-term follow-up of a large group of patients, including some of Swenson's original patients.

Duhamel described a technique in which the native rectum is left in situ and the normally innervated colon is brought behind the rectum in the presacral space. An end-to-side anastomosis is then performed, and the two lumens are joined. Originally, this was accomplished by placing several clamps and cutting between them, but in more recent years most surgeons use a linear stapler for this. The Duhamel procedure has also been widely used around the world, and excellent long-term results have been published.

The Soave procedure (or endorectal pull-through) was designed to avoid injury to pelvic vessels and nerves, which are theoretically at risk with the aforementioned procedures, particularly the Swenson procedure. The operation consists of a mucosal proctectomy with preservation of the rectal muscular cuff, and the normally innervated colon is pulled through the muscular cuff and anastomosed just above the dentate line. In the original description, the pulled-through bowel was left hanging out for several weeks, and was then amputated and the anastomosis was completed. Boley's modification, in which the anastomosis is performed primarily, is the technique usually employed today.

Other approaches that are performed more frequently outside North America include the Rehbein procedure and the use of long myectomy without resection. The Rehbien operation involves a somewhat higher anastomosis than the previously mentioned operations, although long-term follow-up suggests very good results in experienced hands. For children with short-segment Hirschsprung's disease, some surgeons have employed a simple myectomy, going proximally up to 5 or 6 cm. This can be performed transanally or through a posterior approach. Although good results have been reported in some series, long-term outcomes have not been reported and this approach has not been widely adopted in North America.

Since the earliest descriptions of surgery for Hirschsprung's disease, most authors have advocated a preliminary colostomy. This allows for definitive pathology and colonic decompression followed by a period of growth and a subsequent reconstructive operation. There are a number of options for the approach to creation of the stoma. Some surgeons prefer to

create a transverse loop colostomy as the first stage, followed by a laparotomy with biopsies to determine the transition zone and a pull-through at the same time. The colostomy is then closed in a third stage. This approach is problematic if the transition zone is near the splenic flexure because there may not be enough length to perform the pull-through. More commonly, surgeons do a laparotomy with biopsies as the first stage, and bring the colostomy out just above the transition zone. The second stage usually involves using the colostomy to pull through, and only using a more proximal colostomy in cases where the anastomosis is difficult or tenuous. An ileostomy may occasionally be required for cases in which cecal perforation has occurred or in cases of total colonic disease.

In the early days of surgery for Hirschsprung's disease, most children presented with malnutrition, severe enterocolitis, or an extremely dilated colon, and a colostomy was performed as a lifesaving procedure. In addition, there was a sense that performing a reconstructive operation in a small neonate was technically difficult and that results could be improved by waiting until the child was bigger. The standard approach was therefore to routinely perform a colostomy as the first step, and then do a definitive pull-through around 1 year of age. Over the years, as surgical techniques and magnification techniques improved, many surgeons began to do the definitive operation at earlier ages. However, the use of a routine stoma remained the practice of most pediatric surgeons, except for some children who presented at an older age and did not have extremely dilated proximal colons.

In the 1980s, a number of surgeons began to report one-stage pull-through procedures without the use of a defunctioning colostomy, even in smaller children. During the early to mid-1990s, one-stage operations became increasingly popular, and many reports documented the safety of this approach. Some studies suggested that there were advantages to avoiding the known morbidity of stomas in infants and that a one-stage approach was more cost effective. Despite the move toward one-stage surgery for Hirschsprung's disease, it is still universally accepted that a stoma may be indicated for children with severe enterocolitis, perforation, malnutrition, or massively dilated proximal bowel, and in situations where there is inadequate pathology support to reliably identify the transition zone on frozen section.

Minimal Access Surgery

With the popularity of laparoscopic surgery in the early 1990s, many pediatric surgeons began to include these techniques in their practices. Georgeson was the first to describe a laparoscopic approach to surgery for Hirschsprung's disease, which is a modification of the Soave procedure. His operation involves laparoscopic biopsy to identify the transition zone, laparoscopic mobilization of the rectum below the peritoneal reflection, and a short mucosal dissection from below. The rectum is then prolapsed through the anus and the anastomosis is performed from below. This procedure is associated with a shorter time in hospital, and the early results appear to be equivalent to those reported for the open procedures. Laparoscopic approaches have been also described for the Duhamel and Swenson operations, with excellent short-term results reported.

The transanal Soave procedure uses the same mucosal dissection from below as the Georgeson operation, but without the intraabdominal mobilization of the rectum. The rectal muscle is incised circumferentially several centimeters above the dentate line, and the dissection is continued on the rectal wall, dividing the vessels right on the rectum. The entire rectum and part of the sigmoid colon can be delivered through the anus. The transition zone is identified, and the anastomosis is performed from below. This operation has been shown to be safe and associated with a short hospital stay, early feeding, and minimal analgesia requirements, particularly when compared with the open Soave operation. The transanal approach can be used in a patient with a preexisting colostomy and has the advantage that it can be performed by any pediatric surgeon, including those without advanced laparoscopic skills.

There is controversy surrounding the need to determine the level of the transition zone prior to beginning the anal dissection, both for the laparoscopic and the transanal pull-through. Options for accessing the proximal bowel in order to do the biopsy include laparoscopy or a small umbilical incision. These approaches can also be used to mobilize the splenic flexure in children with higher transition zones. Proponents of biopsy point to the inaccuracy of the contrast enema in predicting the level of aganglionosis. This is particularly important if the predicted transition zone is in the distal colon and the true transition zone is in the proximal colon because many surgeons prefer a different surgical approach to long-

segment disease. A more recent study suggests that 8% of children with a rectosigmoid transition zone on contrast study have a more proximal pathological transition zone, data that support this approach. However, there is no particular benefit to surgeons who prefer to use a straight Soave pull-through for long-segment disease. One large multicenter review of patients undergoing the transanal pull-through demonstrated no difference in terms of postoperative pain, feeding, and hospital stay between those who had a preliminary biopsy to determine the pathological transition zone and those who did not.

Intraoperative and early postoperative complications

The complications of surgery for Hirschsprung's disease include the general group of complications of any abdominal surgery, including bleeding, infection, injury to adjacent organs, and the risks of anesthesia. Those children who undergo a staged procedure with a preliminary stoma may experience stoma-specific complications such as stricture, retraction, prolapse, and skin breakdown.

Anastomotic complications, although uncommon, can be seen after any of the standard pull-through procedures. Anastomotic leak occurs infrequently, and can be avoided by close attention to adequate blood supply of the pulled-through bowel and to minimizing tension on the anastomosis. Although it has never been studied in a prospective fashion, the incidence of anastomotic leak in series of laparoscopic and transanal pull-throughs appears to be lower than that reported in the older literature of open pull-throughs. Strictures and retraction of the pull-through may also occur as a result of poor blood supply and tension, and appear to be less common with the Duhamel procedure.

Late complicaitions

The long-term problems in children with Hirschsprung's disease include ongoing obstructive symptoms, incontinence, and enterocolitis. Quite often, an individual child may have a combination of problems. The incidence of these problems varies in the literature, but ranges up to 50% in some series. More recent publications report higher numbers, likely due to increasing recognition of these problems.

GASTROINTESTINAL BLEEDING

Gastrointestinal (GI) bleeding in infants and children routinely provokes parental anxiety. Even small amounts of blood appear large when spread out or mixed with stool or vomitus. Fortunately, GI bleeding in childhood, although common, is usually limited; thus, time is available for diagnosis with careful history and physical examination aided by standard endoscopic and radiographic procedures. Because GI bleeding occurs in children across all age and size groups and results from several dozen different diagnoses (Table 1), it is most appropriate to divide the patients into four diagnostic age groups. Within each group, evaluation is directed at establishing the most common diagnoses, always keeping in mind the strange or remote causes of bleeding as alternatives.

Table 1. Common Causes of Gastrointestinal Tract Hemorrhage

DISORDER	PATIENTS LESS THAN 1 YEAR OF AGE (N=263)	PATIENTS GREATER THAN 1 YEAR OF AGE (N= 169)
Anal fissure	78	5
Intussusception	50	21
Gangrenous bowel	14	0
Duodenal ulcer	10	8
Gastric ulcer	8	9
Meckel's diverticulum	6	3
Ileal hematoma	1	0
Duplication-colon	1	0
Colonic polyp	0	94
Esophageal varices	0	11
Ulcerative colitis	0	6
Regional enteritis	0	1
Hemorrhoids	0	1
Swallowed maternal blood	12	0
Hemorrhagic disease	19	0
Prolapse	15	0
Unexplained neonatal bleeding	49	0

COMMON CAUSES OF BLEEDING IN NEONATES

Upper Gastrointestinal Tract:

Hemorrhagic disease
Swallowed maternal blood
Gastritis
Coagulopathy

Lower Gastrointestinal Tract:

Anal fissure
Necrotizing enterocolitis
Malrotation with volvulus

The most common presentation of GI bleeding in the neonate occurs in children born prematurely or with some form of neonatal distress. These babies, who have stressful birth histories, periods of resuscitation, or low Apgar scores, frequently have "coffee-ground" gastric aspirate or melena. Even if this bleeding is minor, it is sufficient reason to check that the newborn received vitamin K shortly after birth and has not developed hemorrhagic disease of the newborn. Screening studies for a coagulopathy are in order. If the amount of this initial aspiration is unusually large and blood-tinged, an Apt test should be done to confirm swallowed maternal blood. Otherwise, the source of bleeding is probably stress gastritis. Nasogastric suction, saline lavage, and antacids in small amounts are usually sufficient treatment and obviate further evaluation unless the bleeding persists or worsens.

Rarely, continued or massive hematemesis heralds peptic ulcer disease. Endoscopy is now possible in neonates and infants, quickly establishes the diagnosis, and allows immediate therapy with lavage, antacids, and histamine-2 antagonists. If treatment is unsuccessful, surgery appropriate to the lesion (i.e., gastric resection; vagotomy and pyloroplasty; or, rarely, antrectomy and vagotomy) is necessary. In reality, almost none of these is necessary today because of success with supportive treatment.

When blood is noted in the stool, the three most likely diagnoses are fissure, necrotizing enterocolitis, and malrotation with midgut volvulus. Fissures produce bright-red blood that streaks the stool or occurs in small spots on the diaper, whereas necrotizing enterocolitis or malrotation produce darker, often maroon or purple stools with varying amounts of mucus.

Simple anal examination, sometimes performed with a nasal speculum, confirms the presence of an anal fissure, which is the most common lesion producing GI bleeding in infants. Further tests are unnecessary, and successful treatment in almost all cases includes stool softeners and rectal dilatation.

Necrotizing enterocolitis in most neonates is suggested by the history. Radiography confirms the diagnosis so that aggressive medical resuscitation and therapy can be instituted. Seventy percent to 80% of these patients recover with supportive treatment, including antibiotics, bowel rest, and total parenteral nutrition. The rest require laparotomy, drainage, resection, or decompression. Recurrent bleeding after apparent recovery can herald a second occurrence of the disease or can accompany a postnecrotizing enterocolitis stricture.

The sudden onset of melena with bilious vomiting in an apparently healthy baby who often does not have abdominal distention suggests malrotation and midgut volvulus. The early physical examination generally yields negative results, although the abdomen progressively distends and tenderness ensues. An immediate upper intestinal contrast study with or without a concomitant colon contrast study confirms the malrotation. Immediate laparotomy reveals the anomaly and the presence of volvulus. A Ladd procedure is done in conjunction with derotation of the bowel.

Despite the current ability to aggressively investigate neonates, the causes of neonatal GI bleeding remain unexplained in at least 50% of cases. However, Sherman and Clatworthy reported that none of their 94 neonates with hematemesis (50%), hematochezia (35%), and melena (17%) required urgent surgical intervention.

COMMON CAUSES OF BLEEDING IN INFANTS 1 MONTH TO 1 YEAR OF AGE

Upper Gastrointestinal Tract:
Esophagitis
Gastritis

Lower Gastrointestinal Tract:
Anal fissure
Intussusception
Gangrenous bowel

As children pass beyond the neonatal period, the common sites of GI bleeding shift. In children older than 1 month of age, the two most common causes in the upper GI tract are esophagitis, usually secondary to reflux, and peptic ulcer disease.

An extensive literature review on gastroesophageal reflux documents the frequency of this problem. In addition to a barium swallow that documents reflux in about one half of the children with the problem, esophagoscopy, intraluminal pH monitoring, esophageal manometrics, and technetium milk scanning can be used to confirm the diagnosis.

Bleeding associated with reflux esophagitis is usually controlled with antacids, thickened feedings, upright positioning, and prokinetic agents. An antireflux procedure is rarely performed solely to control bleeding, but concern over unresponsive esophagitis, stricture formation and the often associated failure to thrive may mandate surgery. Because gastroduodenoscopy frequently reveals concomitant gastric and duodenal erosions, medical therapy for the esophagitis may serve dual purposes.

In this 1-month to 1-year-old age group, hypertrophic pyloric stenosis occasionally presents with blood-tinged vomitus from gastritis. Diagnosis is easily made on the basis of age, sex, presence of nonbilious vomiting, and the characteristic palpable "olive." Pyloromyotomy corrects the problem and stops the bleeding.

From 6 to 18 months of age, the probable cause of lower GI bleeding is intussusception. If the baby has episodic abdominal pain and a palpable sausage-shaped mass, the diagnosis is easily made; many of these children, however, are atypical and have minor and irregular pain and no abdominal findings. Venous hypertension in the intussusception results in loss of blood and mucus that produces the classic currant-jelly stools; however, bloody stools from bright red to black are seen almost as frequently. Suspicion based on the infant's age and presentation warrants screening ultrasonography. If a "pseudokidney sign" is identified, a barium or pneumatic enema is administered for definitive diagnosis and reduction. Enema is successful in 60% to 80% of cases with or without sedation or glucagon; if not, surgical reduction must be done.

The second most common cause of rectal blood loss in this age group is gangrenous bowel. The cause of the gangrene varies but is usually

attributed to some form of volvulus - either malrotation with volvulus; segmental small-bowel volvulus; omphalomesenteric remnant with volvulus; internal hernia; or, very remotely, sigmoid volvulus. The rectal bleeding varies from melena to hematochezia. When first seen, many of these children are profoundly ill with dehydration, abdominal distention, masses, and even free perforation. If the child is seen early in the course of the illness, combined upper or lower contrast studies (or both) plus proctoscopy, if indicated, confirm the diagnosis of malrotation with volvulus; when coupled with proctoscopy, these studies may be therapeutic for sigmoid volvulus. Segmental volvulus related to twisting around an omphalomesenteric duct remnant is generally found at the time of laparotomy performed for an acute surgical abdomen or persistent small-bowel obstruction.

COMMON CAUSES OF BLEEDING IN INFANTS 1 YEAR TO 2 YEARS OF AGE

Upper Gastrointestinal Tract:
Peptic ulcer disease
Lower Gastrointestinal Tract:
Polyps
Meckel's diverticulum

After the age of 1 year, peptic ulcer disease is the most common cause of hematemesis. Most of these ulcers, gastric or duodenal, are acute and occur in children who have other problems: burns (Curling's ulcer), head trauma (Cushing's ulcer), malignant disease, or sepsis. Significant hematemesis warrants immediate endoscopy under sedation and topical anesthesia. This procedure has much higher diagnostic accuracy than radiography and thus is now the preferred approach. Therapy for acid-peptic disease in children is similar to that in adults and relies heavily on antacids, histamine-2 antagonists and control of the underlying condition. Obstruction or persistent bleeding indicates the need for surgical therapy.

Common sources of rectal blood in this age group are polyps and Meckel's diverticulum. Polyps in this age group are overwhelmingly of the juvenile type located throughout the colon and, in rare instances in the

small intestine. Presentation is painless defecation streaked or mixed with fresh blood. Although an air-contrast barium enema can show polyps, colonoscopy is now preferred because it permits complete examination of the colon and potential removal of the bleeding polyps. In addition, biopsy of multiple lesions identifies the occasional rare case of adeno-matous polyps (about 3% of children with polypoid lesions) associated with familial polyposis. Heterotopic gastric mucosa in a Meckel's diverticulum promotes the development of ileal ulcers adjacent to the aberrant mucosa or at the ileal junction with the diverticulum. Erosion into a small arteriole leads to painless, brisk, red rectal bleeding. Technetium scanning to identify the aberrant gastric mucosa makes the diagnosis with 90% accuracy. If scanning fails to document the presence of a Meckel's diverticulum, the only other diagnostic test that might help would be a tagged red cell scanning.

Large-volume, continuing, fresh blood loss in the proper clinical setting indicates the need for laparotomy, even without a definitive diagnosis. Resection of the diverticulum and ulcer is curative.

COMMON CAUSES OF BLEEDING IN CHILDREN OLDER THAN 2 YEARS OF AGE

Upper Gastrointestinal Tract:

Esophageal and gastric varices

Lower Gastrointestinal Tract:

Polyps

Inflammatory bowel disease

Trauma

Miscellaneous lesions (Fig.1)

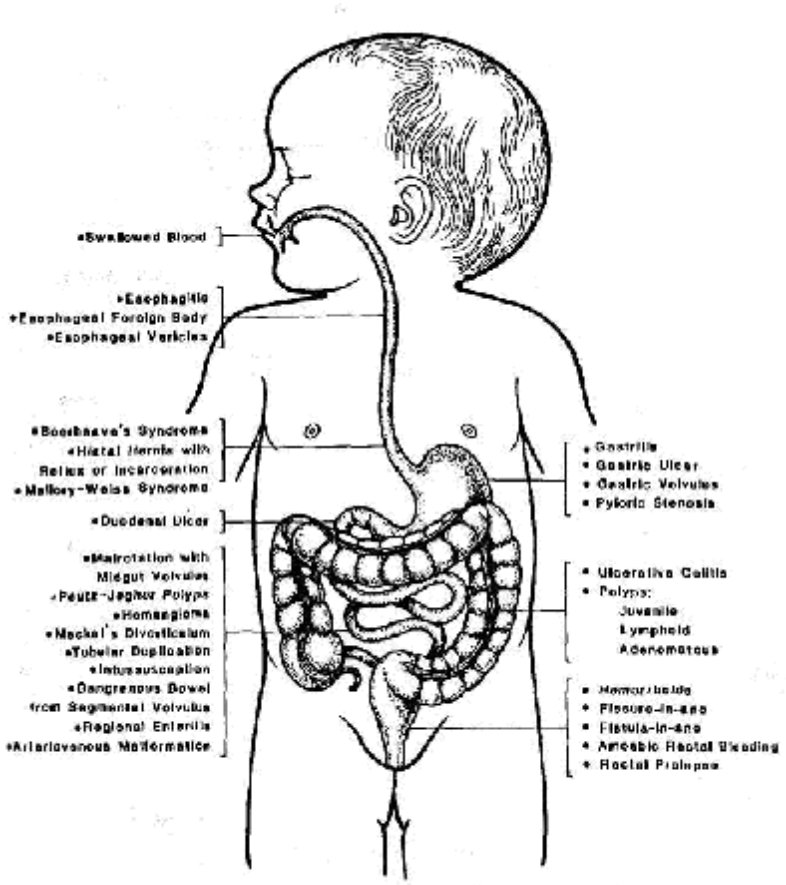


Figure 1 Anatomic sites of more common causes of gastrointestinal bleeding in children.

Varices secondary to portal-vein thrombosis become apparent as children approach 2 to 3 years of age. Massive hematemesis is often the first symptom. On more careful examination, these children are found to have splenomegaly; prominent abdominal veins; and, occasionally, hemorrhoids. Because liver function is preserved and clotting function is normal in most of these children, the bleeding, although heavy, is controllable. Esophagoscopy confirms the diagnosis and concomitant and repetitive sclerotherapy usually controls the bleeding until varices are spontaneously decompressed by portal venous transformation or the child has grown to a size at which portosystemic shunting can be achieved with reasonable expectations of long-term patency.

Portal hypertension related to cirrhosis may be associated with bleeding any time after 6 months of age but is most prominent in children older than 2 years of age. Bleeding is managed in a manner similar to that associated with extrahepatic portal hypertensin.

The most common cause of rectal bleeding in children older than 2 years of age continues to be juvenile polyps. Not until the teenage years do

polyps cease to be a major cause of bleeding. Infectious colitis and inflammatory bowel disease may also be associated with bleeding, sometimes impressive in degree, but the diagnosis and treatment are usually straightforward.

This brief survey of GI bleeding in children older than 2 years of age omits a vast variety of rare and unusual diagnoses for which the precise evaluation is tailored to the history and physical examination. Aggressive use of gastroduodenoscopy and colonoscopy identify most causes of bleeding. However, undiagnosed bleeding requires special studies such as small-bowel enema arteriography or tagged red cell scanning. At times, celiotomy and careful exploration is required. Unlike patients with chronic abdominal pain, in whom laparotomy produces great frustration and few findings, at least half of the children with persistent gastrointestinal bleeding have a demonstrable lesion detected at surgery; lesions include arteriovenous malformations, hemangiomas, duplications, and foreign bodies.

PORTAL HYPERTENSION

In 1877, Eck successfully created a portosystemic shunt in a canine model. Band (1898) described a syndrome of splenomegaly and gastrointestinal bleeding that became known as the portal hypertensive state. No further research occurred for almost half a century until the 1940s and 1950s, when protein intolerance in the cirrhotic patient was studied by Sherlock. The first clinical application of portal decompression, splenorenal shunting, was performed by Allen Whipple in 1945. Alternate techniques for diversion of portal blood for the management of bleeding esophageal varices were developed soon after. Studies suggesting that encephalopathy occurring after the liver has been deprived of portal blood prompted Warren, Zeppa, and Foman and others to emphasize the importance of maintaining prograde hepatic portal inflow, from which evolved selective portosystemic shunting.

Technical obstacles imposed by the size of vessels for shunting in children have been overcome, and both selective and non-selective portosystemic shunts have been successful in small patients. Emphasis has

recently been placed on pharmacologic intervention for treatment of variceal bleeding. Such options as variceal obliteration by sclerosis or endoscopic ligation have attained therapeutic prominence. Refinements in techniques of portal diversion, including the use of slender interposition grafts and transjugular intrahepatic portosystemic shunting (TIPS) have emerged. For selected patients with portal hypertension and end-stage liver disease, replacement of the liver by transplantation is now an option.

Physiology

Portal pressure {P) is a function of splanchnic blood flow (Q) and resistance to flow (R) —P = Q X R (Ohm's law). Thus, portal hypertension reflects the product of both increased flow and increased outflow resistance. The primary effector of portal resistance is the radius of the outflow vessel. Small changes in vessel size result in large changes in resistance. When resistance is low, increases in flow can be accommodated; when vascular resistance is high, small changes in flow result in a marked increase in pressure. Total venous resistance initiates portal hypertension, and increased splanchnic flow sustains it.

The net effect of increased flow and resistance is the development of alternate channels to decompress the portal circuit. These include diaphragmatic; periumbilical; lumbar; splenore-nal; gonadal; perirectal; and, of clinical significance, gastro-esophageal connections. Esophageal varices develop when the portal pressure gradient (the difference between the portal pressure and the pressure in the inferior vena cava) increases. The threshold gradient for clinically significant portal hypertension is greater than 10 mm Hg. Hemorrhage from gastroesophageal varices imperils the life of the patient with portal hypertension.

The consequences of obstruction to portal flow may be subtle and readily compensated or alarming and life-threatening. The prognosis for the patient with portal hypertension is influenced by many factors, particularly the status of hepatic function. In patients with extrahepatic portal vein obstruction, the liver parenchyma is not subject to increased pressure and liver function is preserved. Therefore, these patients tolerate bleeding better than do patients in whom portal flow is obstructed within the liver secondary to hepatic fibrosis. The latter patients are often nutritionally and physiologically compromised. Hemorrhage is poorly

tolerated in these patients, and resuscitation is more difficult than in patients with intact liver function.

Management of the child with portal hypertension is largely determined by the cause of obstruction to portal venous flow. The cause may be prehepatic, hepatic presinusoidal (nonparenchymal - e.g., congenital hepatic fibrosis and schistosomiasis) or hepatic postsinusoidal (parenchymal - e.g., cirrhosis). Much less common is suprahepatic obstruction (Budd-Chiari syndrome).

Extrahepatic obstruction

Portal vein thrombosis secondary to intraabdominal sepsis or perinatal omphalitis with recanalization and the development of collateral venous channels, referred to as cavernomatous transformation of the portal vein, previously accounted for most cases of portal hypertension in children. Now, however, liver disease accounts for most cases. Biliary atresia, palliated but not cured by the portoenterostomy procedure, is the most frequent cause of liver disease resulting in portal hypertension. Additional causes include congenital hepatic fibrosis, a disease of obscure cause that occurs in two forms: familial, with an associated renal tubular anomaly, and sporadic, in which kidney involvement is unusual. The incidence of significant portal hypertension with consequent gastrointestinal hemorrhage is high. Because the fibrosis is presinusoidal, the liver parenchyma is spared and liver function remains relatively preserved. The outlook is generally favorable. Approximately 10% to 15% of patients with cystic fibrosis develop focal biliary cirrhosis and portal hypertension. Cirrhosis resulting from the metabolic abnormality α_1-antitrypsin deficiency or from a complication of chronic active viral hepatitis is an additional etiologic consideration in children.

Suprahepatic obstruction: Budd-Chari syndrome

Hepatic vein or suprahepatic vena caval obstruction, not uncommon in Central and South America, is a rare cause of portal hypertension in North America and Europe. In Asian populations, suprahepatic caval obstruction frequently results from obstructing intraluminal webs or diaphragms. The association between hepatic vein thrombosis and the use of oral contraceptives is recognized. In most cases, however, a specific etiologic factor is not identified. The onset of the condition is insidious, and its course indolent. Often months or years lapse between the first

symptoms and recognition of the syndrome. The hepatic parenchyma is subject to extensive destruction because of high-grade outflow block. Attempts to relieve thrombosis by systemic anticoagulation or the administration of fibrinolytic agents have had only limited success. Partially diverting portosystemic shunts, including side-to-side portacaval, splenorenal, and mesocaval shunts, have been successful in alleviating symptoms and extending survival. Cameron and Maddrey described a mesoatrial shunt through which portal flow is directed into the systemic circulation by a graft from the superior mesenteric vein to the right atrium for portal decompression in patients with caval narrowing. Surgical intervention by portal decompression or, in extreme circumstances, liver replacement, is appropriate for patients in whom inexorable deterioration leading to death is certain.

Gastrointestinal hemorrhage from esophageal varices

Whether the portal pressure is elevated secondary to portal vein thrombosis or by intrahepatic disease, collateral venous channels inevitably develop. The life-threatening complication of portal hypertension is hemorrhage from the variceal collaterals around the esophagus and stomach. Gastrointestinal hemorrhage presenting in a previously healthy child, particularly a child with a history of abdominal infection or umbilical vein cannulation (perinatal), suggests portal vein thrombosis as the cause. Stigmata of cirrhosis (jaundice, ascites, spider angiomata, and hepatosplenomegaly) indicate that the portal hypertension is more likely to be the result of liver disease. The essentials of initial management of the patient with gastrointestinal hemorrhage are the same regardless of the cause. Central venous access is obtained, and a tube is placed in the stomach for gastric lavage with iced normal saline. To avoid water intoxication, particularly in small children, the volume of solutions used for irrigation must be monitored and the stomach must be aspirated completely after each flush with irrigant.

Pharmacologic treatment of acute variceal bleeding

The intravenous administration of vasopressin causes splanchnic vasoconstriction, which in turn reduces portal blood flow and pressure (Fig.2). The recommended rate of intravenous infusion is between 0.2 and 0.6 U/min. However, the systemic effects of vasopressin, including systemic vasoconstriction, water retention, and hyponatremia, have

stimulated the search for alternate agents. Glypressin is a synthetic analogue that, like vasopressin, causes splanchnic vasoconstriction without inciting unfavorable side effects. Glypressin has been used effectively in combination with nitroglycerin. The vasodilator effect of nitroglycerin lessens systemic vasoconstriction and reduces resistance to outflow in the portal system, a primary component to sustaining elevated portal pressure.

Somatostatin, a naturally occurring gastrointestinal inhibitor has also been shown to effectively control variceal bleeding. Somatostatin and particularly its synthetic analogue octreotide (which has a half-life 100 times longer than somatostatin) decrease portal blood flow. These agents have few side effects and are as effective as or superior to vasopressin when administered intravenously at a dose of 25 μg/hr.

Pharmacologic treatment of portal hypertension

β-blockers (propranolol or nadolol plus isosorbide mononitrate cause splanchnic vasoconstriction and reduction of cardiac output, the net effect of which lowers portal pressure. Several studies have confirmed the effectiveness of β-blockade in reducing the mortality rate in patients at risk for hemorrhage from esophageal varices. Side effects are minimal, and there seem to be few contraindications to recommending propranole for patients who have bled or are at serious risk for variceal bleeding.

Balloon tamponade

Balloon tamponade is hazardous in infants and small children because of the risk for aspiration of secretions that accumulate after blockage of the esophagus by the balloon. Further, the accidental dislodgement of the tube in an agitated child can resuit in catastrophic airway obstruction. When balloon tamponade is used, the margin for safety is increased by protecting the airway by endotracheal intubation. Nonetheless, direct compression of the varices by balloon tamponade is effective in most patients. Because bleeding often recurs after deflation of the balloon, this procedure should be regarded as a temporary intervention for control of life-threatening variceal bleeding while the patient awaits more definitive treatment, including pharmacologic therapy and emergency sclerotherapy.

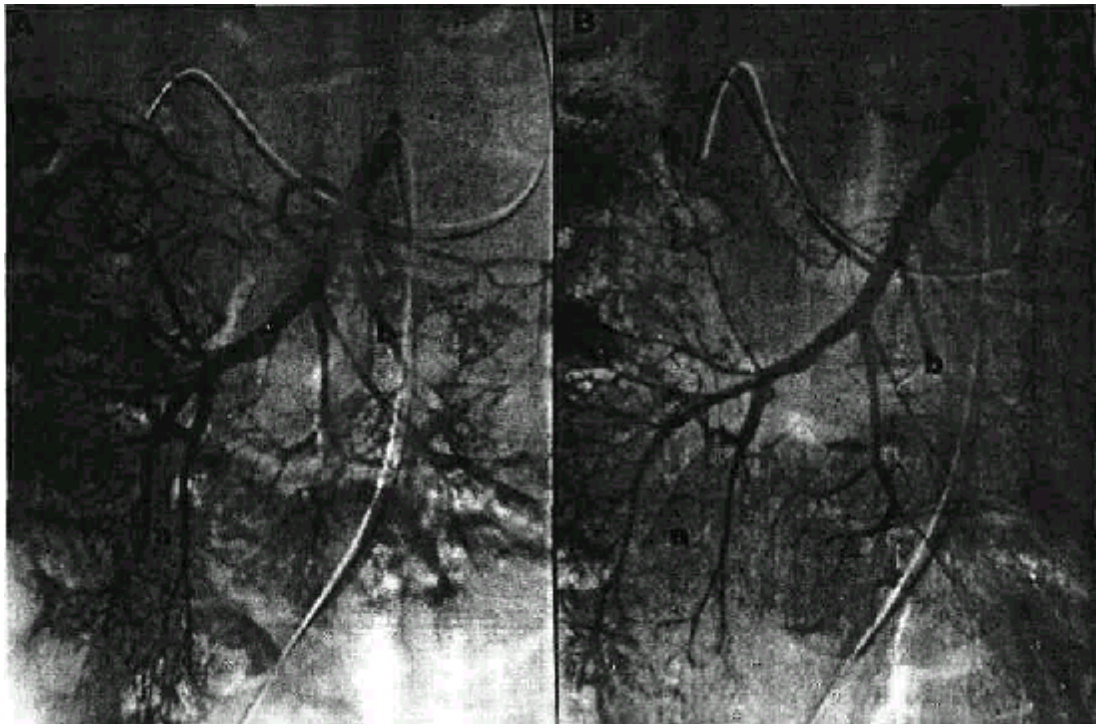


Figure 2. Effect of vasopressin infusion in superior mesenteric artery. A, Selective superior mesenteric arteriogram before and after B, a 20-minute infusion of vasopressin, 0.1 U/rnl per minute. Angiograms were obtained at the same flow rate of contrast and exposed 4 seconds after the start of injection. Note marked decrease in caliber of ileal *(a)* and jejunal *(b)* arteries and decreased capillary stain, reflecting markedly reduced visceral blood flow after vasopressin infusion. This effect also can be obtained by peripheral intravenous infusion of vasopressin or a somatostatin analogue (octreotide).

Endoscopy

Direct visualization of the esophagus, stomach, and duodenum through the flexible endoscope is the most accurate and reliable technique for assessing the patient with upper gastrointestinal hemorrhage. Endoscopic observation of the esophagus is both diagnostic and prognostic. Variceal size correlates with risk for bleeding. Larger varices have a greater tendency to bleed than small ones. Interval endoscopic evaluation in patients with varices might identify those at a greater risk for bleeding. Because endoscopic sclerosis or banding of hemorrhaging varices is recommended for initial intervention, diagnosis and therapy are initiated simultaneously.

Direct obliteration of esophageal varices by endoscopic sclerotherapy was first described by Crafoord and Frenckner in 1939. Because the portal pressure was unmodified, new collaterals developed and bleeding usually recurred. For many years, injection procedures were out of favor. The resurgence of interest in sclerotherapy was stimulated by Terblanche et al, and the procedure is now preferred as the first line of interventional therapy. Direct injection of sclerosant concomitant with pharmacologic measures is recommended for the emergency management of patients with acute hemorrhage and those who have been resuscitated and have had the bleeding controlled.

Variceal injection can be done successfully by using rigid endoscopic equipment or, depending on the experience and preference of the operator, the flexible endoscope. By using a modified slotted rigid endoscope, the varix is trapped and 0.5 to 2.5 ml of sclerosant is injected directly. Paravariceal injection is also effective. The esophagoscope is rotated so that the solid side can perform tamponation at the injection site. Several clusters of varices are similarly treated. Visualization is easier through the flexible endoscope, but postinjection tamponade is not possible. Flexible endoscopy also permits limited injection of gastric varices. Both techniques are effective. Serial injections are usually required to obliterate the varices but are well tolerated. Relatively infrequent but serious complications of the procedure include hemorrhage, mucosal ulceration, dysmotility, esophageal stricture, and perforation. The procedure is relatively safe in experienced hands and is recommended in children with portal hypertension either secondary to portal vein thrombosis or to cirrhosis.

Endoscopic banding of esophageal varices

This technique, advocated by Steigmann, can be applied to acute bleeding or the short-term and long-term management of esophageal varices. To expedite the repeated intubations of the esophagus required by the banding procedure, an endoscopic overtube or sheath is placed in the esophageal lumen, thereby allowing expeditious passage of the endoscope. A ligating device is adapted to the endoscope, which is advanced to the target varix and establishes full circumferential contact. Suction draws the varix into the banding chamber, and a latex banding ring is released around the base of the varix (Fig. 3). The procedure can be done during hemorrhage or after bleeding has been controlled by initial pharmacologic

intervention. Subsequent ligations are performed until all distal esophageal varices are ligated. Interval inspections are performed to ensure that the varices are completely eradicated. Endoscopic ligation has proven to be as effective as sclerotherapy for control of active leeding and has a lower incidence of adverse sequelae. The procedure is operator-dependant and requires a skilled endoscopist for successful outcome.

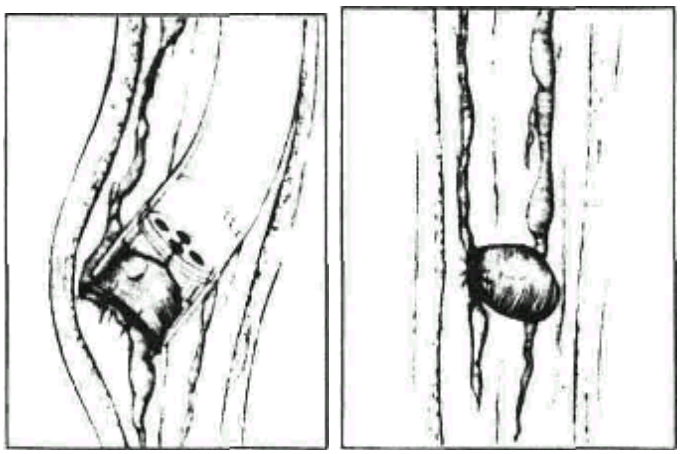


Figure 3. Endoscopic variceal banding. A, Suction through the endoscope draws the target varix into the banding chamber. B, A trip wire releases the "O" ring, which ensnares and strangulates the varix.).

OTHER DIAGNOSTIC TECHNIQUES

Splenoportography

Percutaneous splenoportography can be done in children. Portal pressure is measured, and injection of contrast directly into the spleen accurately represents the patency and caliber of the splenic and portal veins. This may have particular application for patients considered for surgical shunt procedures or TIPS (see Fig.8, B).

Angiography

Mesenteric artery cannulation is feasible in infants and children and sequential radiographs obtained after injection usually demonstrates the venous anatomy, including collateral channels and esophageal varices. The administration of vasoactive agents into the mesenteric artery controls variceal bleeding, but vasopressin or somatostatin infused through a

peripheral vein also reduces splanchnic blood flow. The peripheral vein is the preferred route for administration.

Percutaneous transhepatic occlusion of the coronary vein

Occlusion of the coronary vein with obliteration of bleeding collateral esophageal varices can be an effective temporizing measure. Portal vein patency is a requisite. The portal vein is accessed by direct percutaneous transhepatic puncture, and coils are delivered into the coronary vein tributary, thereby interrupting flow to bleeding varices. This procedure is usually used in combination with pharmacologic manipulation when endoscopic techniques have been unsuccessful for the management of life-threatening variceal bleeding.

Shunt therapy

Decisions regarding the advisability of portosystemic shunts as opposed to nonshunt options are often difficult. Important factors include the clinical status of the patient, the cause of portal hypertension, and the experience and resources of the institution. The therapeutic dilemma is compounded by the inability to foretell which patients will rebleed despite medical treatment and which will develop liver failure or encephalopathy with or without a portosystemic shunt. Child's criteria for assessing hepatic reserve have been useful in predicting the patient's tolerance to portal diversion and the results to be expected from shunting portal flow. In Fig.4, the clinical status of patients with extrahepatic portal hypertension is compared with that of a cohort with elevated portal pressure and gastrointestinal hemorrhage secondary to cirrhosis. Most patients with portal vein obstruction have essentially normal liver function and are in child's A category, whereas patients with cirrhosis have impaired liver function and are in the less favorable B or C categories. Clatworthy has aptly contrasted extrahepatic and intrahepatic portal hypertension, referring to the former as "good liver, bad veins" and the latter as "bad liver, good veins." Effective and reliable portal decompression in small patients has taxed the surgeons' ingenuity. The concept of shunting blood from the high-pressure portal to the low-pressure systemic circulation was pioneered by Whipple and refined by Clatworthy, Warren, and Sarfeh. Through their efforts, the end-to-side and the side-to-side portacaval and splenorenal shunts were proven effective for decompressing the portal system and controlling hemorrhage from esophageal varices. Clatworthy and Boles

developed the central splenorenal shunt for specific application in children. Clatworthy and Marion independently described die direct mesocaval shunt for patients in whom neither the portal nor splenic veins were patent. The interposition mesocaval shunt was popularized by Drapanas in adults and applied in children by Altman and Nay and Fitzpatrick. Orloff, Orloff, and Rambotti citing the high mortality rate from uncontrolled or recurrent hemorrhage, advocated emergency intervention by portosystemic shunt as initial therapy for children with variceal bleeding. He reports remarkable survival with a minimal incidence of late adverse sequelae. Excellent results with portal decompression by shunt procedures in infants and children have been achieved in many centers. Thus, neither age nor size should be considered limiting factors.

Shunt Therapy for Extrahepatic Portal Obstruction

Patients with extrahepatic portal obstruction usually present between 2 and 4 years of age, and gastrointestinal hemorrhage may be the first indication of the underlying disorder. Because the obstruction to portal flow is extrahepatic, liver function is unimpaired and bleeding is well tolerated. The feasibility of shunting in children has been demonstrated by Maksoud and Mies, Bismuth, Franco, and Alagille, and Orloff, Orloff, and Rambotti. Mitra et al reported an extensive experience with side-to-side splenorenal shunt without splenectomy in children with noncirrhotic portal hypertension with 87% patency. Their reports contrast with the traditional recommendation that collateral channels will eventually develop, decompress the portal system, and obviate die need for surgery in all but a few patients**.**

Childs Criteria for Assessement of Hepatic Reserve

Criterion	(A) Good Risl	(B) Moderate Risk	(C) Poor Risk
Serum bilirubin (mg/100ml)	<2.0	2.0-3.0	>3.0
Serum albumin (gm/100ml)	>3.5	3.0 - 3.5	<3.0
Ascites	None	Easily controled	Poorly controled
Encephalopathy	None	Minimal	Advanced
Nutrition	Excellent	Good	Poor

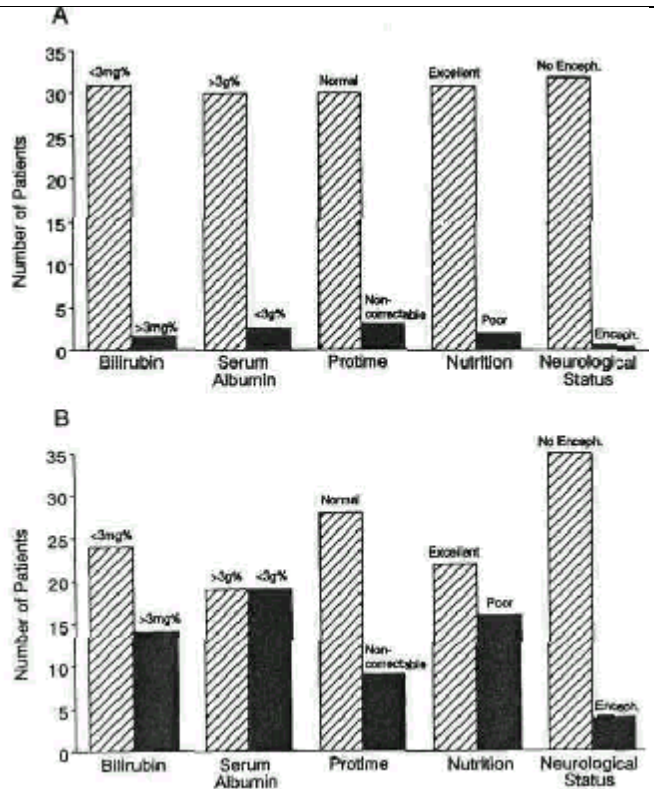


Figure 4. Child's criteria for assessment of hepatic reserve. Clinical and laboratory status of patients who bled from esophageal varices based on cause of bleeding. A, Extrahepatic portal vein obstruction. B, Cirrhosis. The patient in the extrahepatic portal hypertension group is in the favorable Child's A category, whereas those with liver disease are compromised clinically and nutritionally.

Shunt Therapy for Intrahepatic Obstruction in Cirrhosis

As cirrhosis progresses, the likelihood of bleeding increases. In patients with compromised liver function, recurrent bleeding is ominous. Here too, the overall health status of the child influences die choice of therapy. For example, children with cystic fibrosis and severe pulmonary disease are probably not eligible for surgical portal decompression,

whereas those with adequate lung reserve may be good candidates. When hemorrhage represents the principal threat to life in patients with biliary atresia, who demonstrate relief of jaundice and stable hepatic histologic findings, portosystemic shunting may be life saving. The mesocaval H-graft is applicable in patients who have previously had surgery in the porta hepatis (e.g., Kasai portoenterostomy) or in patients in whom a portocaval or splenorenal shunt has failed. Autogenous jugular vein has been used successfully for creation of the shunt (Fig.5).

The ideal portosystemic connection preserves liver function while shunting the high-pressure portal flow to the systemic circulation. Although bleeding is routinely controlled, serious concerns remain regarding undesirable side effects after conventional shunt therapy. The hemodynamic flow pattern and physiologic consequences to the patient vary with the type of shunt constructed. The end-to-side portocaval shunt directs al portal flow around the liver (Fig.5). The side-to-side portocaval and all splenorenal and mesocaval shunts theoretically direct only a portion of the splanchnic flow to the systemic circulation (see Fig.6). However, reversal of blood flow in the portal vein, which allows the vein to act as an outflow tract, is a potential consequence of any of these shunts. This phenomenon is implicated in the development of disabling post shunt hepatic encephalopathy. Vorhees et al described serious neuropsychiatric complications in adolescents and adult previously subjected to portosystemic shunting. These complications raise serious questions about the advisability of shunting portal blood flow in children.

Stimulated by concerns about the high incidence of encephalopathy after standard shunting, Warren, Zeppa, and others proposed the selective distal splenorenal shunt (DSRS) (see Fig. 6, B). By this technique, portal circulation is partitioned into two components: (1) antegrade portal flow to the liver and (2) selective, transplenic flow from the esophageal varices through the short gastric veins through the splenic vein into the renal vein and systemic circulation. Thus, portal inflow to the liver is maintained while the collateral esophageal varices are selectively decompressed. This surgery has been used successfully in children, and extended patency after selective distal splenorenal shunt has been confirmed.

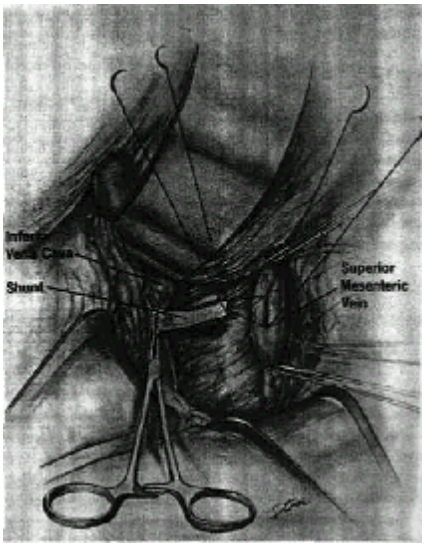


Figure 5. Interposition mesocaval shunt operation in a 14-month-old child. After portoenterostomy (Kasai's procedure) was performed at age 8 weeks, the serum bilirubin level was normal and the child's growth was in the 50th percentile. Three episodes of life-threatening hemorrhage from esophageal varices led to interposition mesocaval shunting that used the autogenous internal jugular vein. The relation between the superior mesenteric vein and inferior vena cava is seen. The duodenum is retracted superiorly, and the biliary conduit is retracted toward the left. The caval anastomosis is completed first. The caval and mesenteric venotomies are twice the diameter of the vein graft, which will distend after flow is restored. Shunt patency has been confirmed by transfemoral venography.

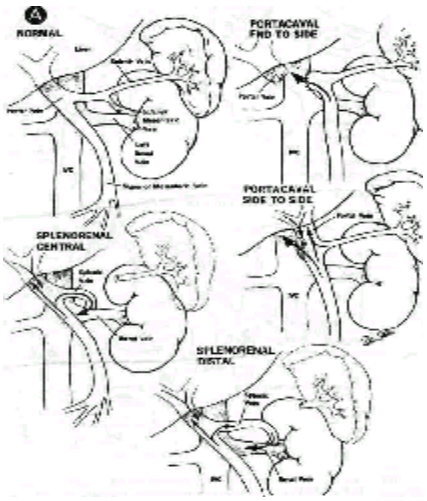


Figure 6. The normal relationship between the portal and systemic circulation is seen, and the several options for establishing portosystemic diversion are shown. A, In the end-to-side portacaval shunt, all portal flow is diverted to systemic circulation. In the side-to-side portacaval shunt, the major direction of portal flow is to the systemic circulation; however, the hepatic portal perfusion is retained, depending on the resistance within the liver. Flow dynamics are such that there is a potential for reversal of flow in the portal vein (hepatofugal), shown by the *broken arrow*. In a central splenorenal shunt, the principal direction of portal flow is to the systemic circulation. Central placement of the shunt minimizes angulation of the splenic vein (spleen removed). In the distal splenorenal shunt, the major direction of portal flow is to the systemic circulation. Perfusion of the liver with portal blood and potential hepatofugal flow is shown (spleen removed).

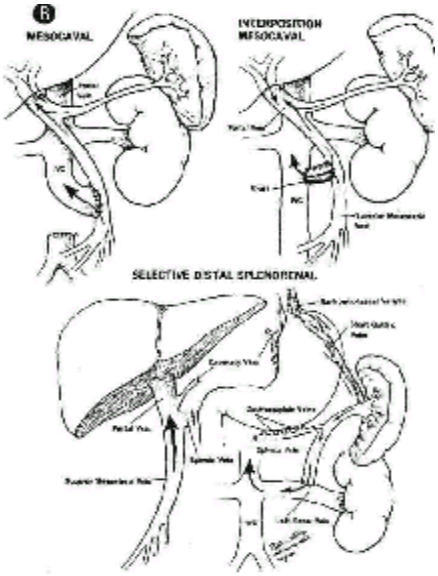


Figure 6, Cont'd. B, Mesocaval shunt. Vena cava is transected and proximal cava is anastomosed to the side of the superior mesenteric vein. Major direction of visceral flow is toward the vena cava. Hepatopetal flow and the potential for hepatorugal flow are depicted. An interposition mesocaval shunt is hemodynamically similar to a mesocaval shunt. Autogenous vein graft is preferred for creation of this shunt in infants and children. In selective distal splenorenal shunt, the portal flow is partitioned. Portal inflow to the liver is preserved, while gastroesophageal varices are simultaneously decompressed through short gastric veins and then to systemic circulation by a distal splenorenal shunt.

Selective Shunting

Notwithstanding the encouraging reports from many centers, shunt procedures in children may be complicated by the small size of the veins available and the increased risk for thrombosis. The long-term consequences of diversion of portal blood away from the liver into the systemic circulation in a young patient are not completely known. Further, rearrangement of extrahepatic veins may compromise the chances of a patient who subsequently competes for scarce liver grafts should transplantation be required.

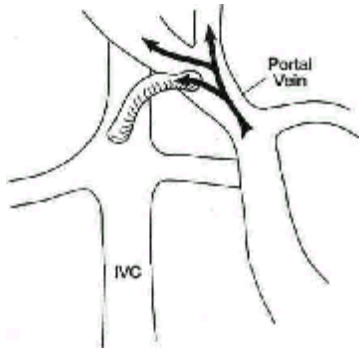


Fig. 7. Slender interposition portocaval graft effectively decompressed the portal system while preserving prograde portal flow.

Portocaval H-Graft Shunt

Sarfeh applied a slender graft using polytetrafluoroethylene (PTFE) interposed between the portal vein and inferior vena cava to reduce portal pressure sufficiently to control variceal bleeding while preserving prograde portal hepatic perfusion (Fig. 7). Collateral venous channels and the coronary vein are ligated. Thus, portal perfusion of the liver is maintained while partial portal diversion reduces portal flow and pressure. Although experience with small-diameter portocaval H-grafts in children has been limited, the procedure is attractive because studies have demonstrated that the standard mesocaval graft, even when the autogenous vein is used, can result in total portal diversion from the liver. An additional advantage of Sarfeh's partial shunt is that these grafts are readily managed in case of subsequent transplantation. They do not distort the venous anatomy and require only simple ligation to obliterate the shunt and redirect all splanchnic flow through the portal vein.

Transjugular Intrahepatic Portosystemic Shunting

Percutaneous TIPS eliminates the need for laparotomy and overcomes the disadvantage of altering the extrahepatic venous anatomy. By this technique, an intrahepatic communication is established between the hepatic venous outflow and the portal venous inflow (Fig. 8). The portosystemic communication is accomplished under fluoroscopic guidance. It is dilated and the intrahepatic tract secured by insertion of an expandable stent. These procedures are routinely successful in decreasing the portal pressure and eliminating the dangerous gradient between the portal and systemic venous systems (Table 2).

Ascites is regularly resolved with TIPS. The TIPS shunt is subject to stenosis and occlusion, and vigilant surveillance by interval ultrasonographic evaluation is essential. Stenotic shunts can be successfully dilated by a percutaneous approach. The procedure is particularly applicable for patients in whom future liver transplantation may be anticipated. The advantages of the TIPS procedure over surgical shunts, including the elimination of surgical mortality and preservation of intact extrahepatic veins, warrant its inclusion as a therapeutic option for children.

Table 2.

Hemodynamics at application of TIPS

Variable	Before TIPS	After TIPS
Splenic pulp pressure, mm Hg	**38**	**-**
Portal vein pressure, mm Hg	**38**	**26**
Right atrial pressure, mm Hg	**11**	**15**
Corrected sinusoidal pressure, mm Hg	**27**	**11**
Cardiac output, L/min	**3,3**	**3,5**

TIPS – Transjugular intrahepatic portosystemic shunting

The TIPS procedure has been proven effective for controlling bleeding esophageal and gastric varices even when other measures have failed. As with all therapies for portal hypertension, however, TIPS may be associated with immediate and long-term complications. In an attempt to define the role of TIPS, the National Digestive Diseases Advisory Board made the following recommendations on the basis of the safety, efficacy, and indications for TIPS procedures.

Accepted indications for TIPS include acute variceal bleeding or recurrent bleeding refractory to pharmacologic measures for which sclerotherapy (banding) has failed. Less clear is the application of TIPS for treatment of refractory ascites although the results (including results seen through personal experience) have been encouraging. The Advisory Board cautions about the use of TIPS for the initial therapy for variceal hemorrhage as well as for prophylactic use in patients who have never bled.

Contraindications to the use of TIPS include right-sided heart failure (elevated central venous pressure) and severe hepatic failure. Patients with polycystic liver disease have additional risks (in the patient shown in Fig. 8, however, congenital hepatic fibrosis caused portal hypertension).

Transjugular intrahepatic portosystemic shunting represents a potentially important therapeutic alternative, but additional studies, particularly in children, are needed to better define and understand the indications and limitations of the procedure.

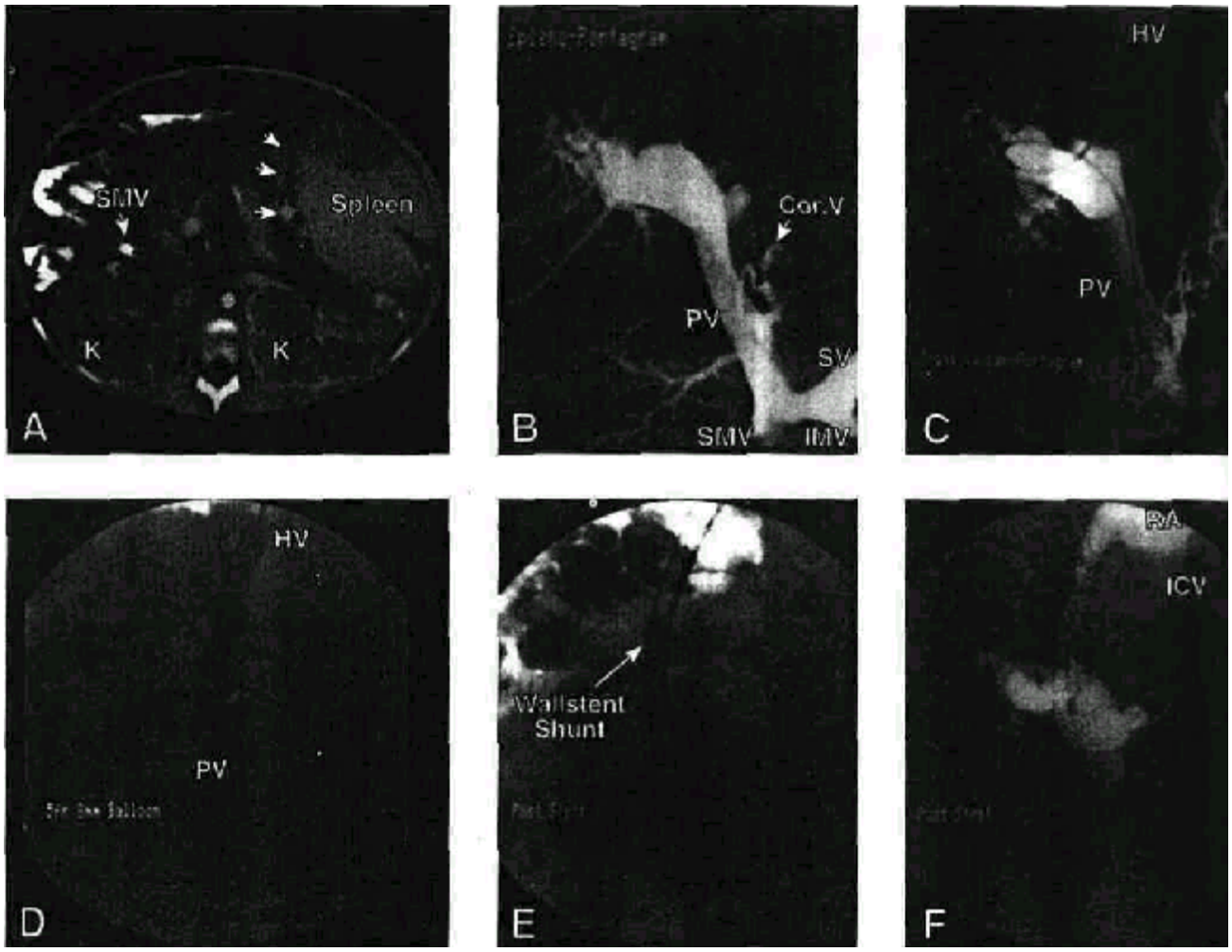


Figure 8. The patient is a 7-year-old girl with portal hypertension secondary to congenital hepatic fibrosis. A, Computed tomographic scan shows a massively enlarged spleen with serpiginous variceal collaterals apparent in the splenic hilum *(arrows).* The large kidneys with characteristic low attenuation, signal are also seen. The large superior mesenteric vein *(SMV)* is patent. Images obtained closer to the head demonstrate a patent portal vein. B, Splenoportogram shows the extrahepatic venous anatomy. The splenic vein *(SV)* is confluent with the superior mesenteric vein *{SMV).* The portal vein *(PV)* is patent, and flow is hepatopetal. The inferior mesenteric vein *(IMV)* is seen, and the coronary vein (COR V) is shown coursing toward the head. C, Through a transjugular approach, a 10-French catheter has been placed in the hepatic vein *(HV).* The portal vein is identified, and a 5-French catheter is inserted into its lumen. D, A balloon has been placed and inflated in the liver between the hepatic and portal veins. The inflated balloon establishes and dilates the connection between the systemic and portal circulations. E, The tract is secured by placement of an 8-mm Wallstent

shunt. F, The postshunt portovenogram confirms wide patency with contrast seen entering the right atrium *(RA)* and minimal visualization of the inferior vena cava *(IVC).*

Surgical, nonshunt alternatives

Surgical alternatives to portal decompression have limited application in children. Transthoracic ligation of varices or direct ligation by an abdominal approach provides only short-term palliation. Esophageal transection, esophagogastrectomy, short-segment colon or small-bowel interposition, and transplantation and translocation of the spleen and omentum have all been used as temporizing procedures in young patients. These operations have largely been abandoned and are now of only historic interest.

Devascularization Procedures

Sugiura and Futagawa advocated extensive esophagogastric devascularization combined with esophageal transection and reanastomosis. Ligation procedures or interruption of varices by use of direct suture or staples combined with devascularization also have been recommended. Although these procedures can be expedited by the use of staples, they tend to be long and tedious. Devascularization procedures are effective in controlling and preventing future bleeding, but theyhave not attained great popularity in children.

Liver Transplantation

In theory, liver transplantation would be optimal for the treatment of patients with life-threatening variceal hemorrhage from portal hypertension caused by liver disease. Replacing the diseased liver eliminates resistance to portal flow, thereby restoring normal hemodynamics. However, liver transplantation should not be considered primary treatment for portal hypertension. As emphasized by Shaw, liver transplantation is the treatment of end-stage liver disease, which may include portal hypertension and its complications. Thus, liver replacement is appropriately reserved for patients with liver failure, and portal hypertension is managed by any of the applicable therapeutic alternatives singly or in combination. These methods cannot be discarded in favor of liver replacement for many reasons, not the least of which is the scarcity of available organs the risks for immunosuppression. However, ineffective measures with little likelihood of success are not in the best interest of the patient. For a

selected group, transplantation of the liver may be the most feasible intervention.

Ascites

A particularly troublesome consequence of obstructed portal blood flow is the development of ascites. The mechanism for formation of ascites is incompletely understood. It has been proposed that expansion of the total extracellular fluid compartment causes decreased plasma volume, resulting in retention of sodium and water in the kidney. Pharmacologic and dietary control is possible only in patients with relatively intact hepatic and renal function. In some patients, ascites is refractory to pharmacologic management. Patients with cirrhosis and limited hepatic function or inadequate renal function may require more aggressive therapy.

For some children, the peritoneovenous (LaVeen or Denver) shunt has been successful in controlling refractory ascites. In small patients, the shunt is seated in the iliac fossa, which is approached extraperitoneally (Fig. 9). From this position, the peritoneum is easily reached for placement of the collecting tubing, while the venous canula is tunneled laterally to the neck for placement in the superior vena cava through the internal jugular vein.

Portal hypertension and variceal hemorrhage are often a daunting clinical problem, but many effective therapeutic interventions are available (Fig. 10). Interval endoscopic surveillance to identify patients at risk and the long-term administration of β-blockers to selected patients is prudent. For most bleeding patients, initial pharmacologic management with vasopressin or, preferably, the somatostatin analogue octreotide with concurrent endoscopic sclerosis or variceal banding will succeed. For patients in whom these techniques are unsuccessful, other alternatives are available. The surgical options include both standard and selective shunt procedures. Despite the theoretical objections, the central splenorenal shunt or mesocaval graft can be life saving. Alternatives to standard shunts, including the slender portocaval graft and TIPSs procedures, are feasible in children and warrant additional clinical application and investigation. Liver transplantation is reserved for patients with end-stage disease that is further complicated by portal hypertension.

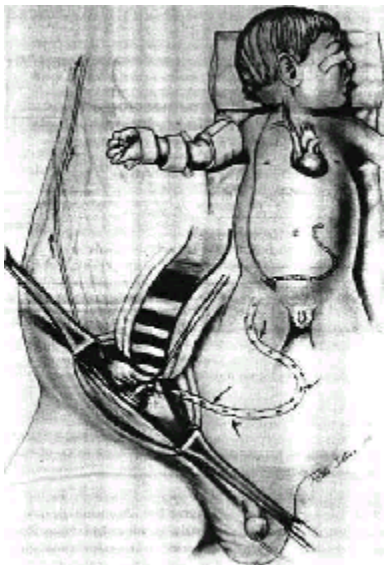


Figure 9. Illustration of 1-year-old infant with cirrhosis, portal hypertension, and intractable ascites. A portoenterostomy was performed at 4 months of age, but liver disease progressed despite good flow of bile. The LeVeen valve was easily accommodated in the right iliac fossa, which was approached extraperitoneally (at age 10 months). The venous tubing is tunneled laterally to enter the superior vena cava by way of a jugular cutdown. Paracentesis was performed just before the procedure to minimize leakage of ascitic fluid and to avoid sudden circulatory overload.

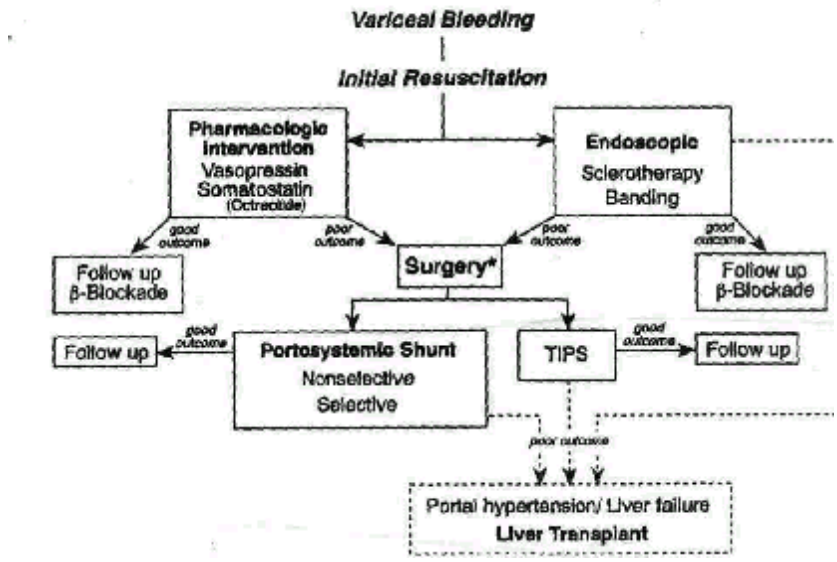


Figure 10. Suggested alogarithm for managing the patient with variceal bleeding, Human resuscitation is standard. Pharmacologic measures, followed by or are administered concomitandy with diagnostic and therapeutic endoscopy, are the first interventions. In most patients, these measures control the bleeding. β-blockade and follow-up study at appropriate intervals is recommended. If bleeding is refractory to these measures or if it recurs despite β-blockade and endoscopic therapy, several surgical options are available, primarily portosystemic shunt and TIPS. (Devascularization procedures are infrequently used in children.) Nonselective shunts, including the slender interposition portocaval graft, are favored, but selective shunting is feasible in pediatric patients. For patients with portal hypertension and variceal bleeding that further complicates end-stage liver disease, liver replacement by transplantation is a rescue option. Other surgical alternatives, i.e. devascularization.

TUMORS IN CHILDREN

The history of pediatric oncology is considerably important. First, it illustrates why pediatric oncology is practiced in a multidisciplinary fashion. This type of approach was responsible for important early successes in the treatment of solid tumors in children, and it is also important for future progress. Second, the history of childhood cancer has demonstrated that treatment concepts and biologic advances developed in children are often applicable to a more general understanding of cancer.

The discipline of pediatric oncology really began in 1948 when Farber et al reported temporary remission in children with acute lymphoblastic leukemia when the folic acid antagonist aminopterin was administered. This report was the earliest evidence that chemotherapy could be effective treatment for malignant conditions of children. Another major step forward occurred in 1956 with the use of methotrexate to cure a solid tumor, choriocarcinoma. The combination of several chemo-therapeutic agents (combination chemotherapy) to cure Hodgkin's disease and acute lymphoblastic leukemia during the 1960s led to use of this principle in virtually all types of treatable pediatric cancers.

The successful use of multidisciplinary treatment in oncology was first demonstrated in Wilms' tumor. Multidisciplinary treatment refers to the combined use of surgery, radiation therapy, and chemotherapy to effect a cure. This principle, which is now used throughout the field of oncology, was first developed by the collaborative efforts of pediatric surgeons, radiation therapists, and pediatric oncologists in the 1950s and 1960s. Other advances in pediatric oncology have included the development of interdisciplinary national cooperative clinical research groups to scientifically test new therapies, the use of adjuvant chemotheray in such solid tumors as osteosarcoma, the use of dose-intensive chemotherapy programs to improve the outcome of advanced-stage solid tumors, and the understanding of cell growth through the identification of tumor-suppressor genes (retinoblastoma). The development and application of these principles and advances has led to profound improvements in the survival and quality of life of children with cancer.

Cancer in children is uncommon and represents only about 2% of all persons with cancer. Nevertheless, after trauma it is the second most

common cause of death in children older than 1 year of age. Approximately 130 new cases of cancers are identified each year per million children younger than 15 years of age. Approximately 9000 children are diagnosed with cancer in the United States annually. From 1973 to 1987, there was a 4% increase in the incidence of childhood cancer in the United States. Leukemia is the most common form a cancer in children, and brain tumors are the most common solid tumor. Lymphomas are next most common followed by neuroblastoma, soft tissue sarcoma, Wilms' tumor, and osteosarcoma; each type of cancer makes 5% to 8% of the total.

Hemangiomas are the most common tumors of infancy and early childhood. They are usually not present at birth but appear in the neonatal period, usually in the first 2 weeks of life. Approximately 30% to 40% are seen at birth as a premonitory cutaneous mark: a barely visible "anemic nevus," a telangiectatic or macular red stain, or an ecchymotic spot mimicking a bruise. Rarely, the tumor is present, fully grown, at birth; such tumors are designated "congenital hemangioma."

A nevus is a focal area of hyperpigmentation resulting from a benign increase in melanocyte cellularity. Most nevi are a risk factor for melanoma in adulthood, and giant congenital melanoma and familial atypical mole or melanoma also are associated with childhood melanomas.

These nevi develop after birth and continue to increase in number and size into early adulthood. They are small (<5 mm in diameter), benign neoplasms of melanocytes that have well-defined borders and usually occur in sun-exposed areas.

They are categorized according to their location in the dermis: junctional, compound, or dermal. Increased number of acquired nevi is a risk factor for malignant melanoma.

Malignant melanoma is a disease of both children and adults. The infrequency of this malignancy in children leads to ignorance of the fact that it does occur, delay in diagnosis, and subsequent later presentation with a worse prognosis in 60% of cases. A thorough understanding of risk factors and nevi with malignant potential is critical for early detection and intervention because treatment of metastatic disease is often not effective.

Malignant melanoma occurring before age 20 years accounts for 1% to 3% of childhood cancers and 1% to 4% of all melanomas. There are three presentations of malignant melanoma in childhood. First, malignant

melanoma can be congenital, either de novo or by transplacental spread from the mother. Congenital melanoma is clinically unpredictable, with complete regressions described in the literature. Second, melanoma can develop in congenital nevi, particularly in giant (>20-cm) congenital melanocytic nevi. Third, melanomas can develop in areas away from nevi. Puberty seems to play a critical role in the development of malignant melanoma; thus, many larger series include melanomas after the onset of puberty with those occurring in adult population.

Prognosis of childhood melanoma is determined by depth of invasion: either anatomic involvement (Clark's classification) or measured depth of invasion (Breslow's classification). The explanation for the association of prognosis and depth of melanoma invasion was suggested by Folkman and later confirmed by Doppler ultrasonography of melanomas. Melanomas that invade deeper tissues stimulate local growth of new blood vessels and angiogenesis, with subsequent increase in tumor size and metastatic potential.

Before description of the Spitz nevus, the prognosis in children was thought to be more favorable than that in adults. Spitz nevus has a benign natural history and was often confused with melanoma. Eighty percent of children present with stage I melanoma. The 5-year survival rate with metastatic disease is 33%. Childhood melanomas are now thought to carry a prognosis similar to that of to adult melanomas, with a combined mortality rate of 40%.

The term tumors of germ cell origin invites some confusion be cause it refers to a broad group of tumors with various histologic types involving many different anatomic locations.

Although the term suggests tumors of the gonads, many gonadal tumors are not of germ cell origin, but rather from epithelial or stromal precursor cells. Additionally, tumors of germ cell origin can be found in paraaxial anatomic locations remote from the gonads, such as the brain, neck mediastinum, retroperitoneum, and coccyx.

Further confusion is created by use of specific histopadiologic terms (such as embryonal carcinoma or endodermal sinus tumor) for tumors located in both gonadal and extragonadal tissues. The common denominator is tumor derivation from the same embryologic precursor cells. The classification system proposed by Dehner demonstrates the tumor multiplicity and the presumed embryologic relationship. The first

description of a teratoma has been attributed to a seventeenth century French obstetrician who was called to assist with the delivery of a baby with a large sacrococcygeal mass. Virchow described a sacrococcygeal tumor in 1869 that contained many different types of tissues apparently originating from remote regions of the body. He used the term teratoma, derived from the Greek, teraton, meaning "monster." The term became commonly used but poorly defined.

Teratomas were soon described in other regions of the body. More recently, teratomas have been defined as "neoplasms that originate in pluripotent cells, composed of a wide diversity of tissues foreign to the organ or anatomic site in which they arise. At times, pathologists have argued that tissues from each of the germ layers must also be present in a tumor to be classified as a teratoma. Although many well-differentiated teratomas may include derivatives from the endoderm, ectoderm, and mesoderm, this restrictive definition eliminates many tumors that clinicians and pathologists would universally agree are teratomas, based on their location and gross appearance.

The multiplicity of tissues discovered in teratomas and their numerous anatomic locations have fostered many different theories about the histogenesis of these tumors. These theories purport that the teratoma cell of origin is (1) primordial germ cells; (2) nongerminal, or embryonic, cells; (3) extraembryonic cells; (4) stem cells; (5) conjoined or maldeveloped twins; or (6) from multiple different cell types, depending on anatomic site. Of these many theories, the most widely accepted concept is the germ cell dieory.

The primitive germ cells are first identifiable in the yolk sac endoderm. These cells migrate to the embryo by the nineteenth day of gestation and are incorporated in the hindgut. These cells continue to migrate along the hindgut mesentery to die gonadal ridge and eventually become the gonads. Primitive germ cells possess all the deoxyribonucleic acid (DNA) necessary for future development of any type of cell; that is, they are totipotential cells. This suggests that these cells retain the ability to form the many different types of tissue seen in teratomas.

Wilms' tumor continues to be the subject of intensive investigations that, with the aid of four cooperative protocols by the National Wilms' Tumor Study (NWTS), have resulted in marked improvement in survival.

Furthermore, the NWTS serves as the model for cooperative group studies in children and is an example of the successful blending of surgical excision, radiation therapy, and chemotherapy for treatment of malignant neoplasms. In the recently completed NWTS-4, the emphasis was on finding the minimum effective dose for cure while reducing unwanted side effects of treatment. The current NWTS-5 is a nonrandomized therapeutic trial with treatment based on age at diagnosis, stage, tumor weight, and histologic findings.

However, even with the most successfully treated types of cancer in children, there continues to be a problem with anaplastic tumors that are resistant to therapy. A continuing quest for a basic understanding of these types of tumors may help in earlier diagnosis and improved outcome by the use of focused treatments.

New developments in molecular biology and pathology are highlighted, and the goals of NWTS-5 are included. We envision an exciting future for the treatment of these tumors.

A brief look at the early reports of Wilms' tumor serves as a prelude to our clinical presentation. Classic pathologic descriptions of the tumor now known as nephroblastoma were written as early as 1872 and described by Osier in 1879. Osier realized that physicians were reporting renal tumors in children and giving them different names, whereas he recognized that they were all describing the same type of solid tumor. However, it was not until 1899, when Wilms thoroughly reviewed the literature and added seven cases of his own, that the definitive paper was produced. His presentation of the clinical picture was so clear that the eponym "Wilms' tumor" continues to be preferred over "nephroblastoma." Surgical excision was the only therapeutic option until 1915 when radiation therapy was added by Friedlander. Ladd and Ladd and White gradually improved the surgical technique and increased the survival rate to 20%. Chemotherapy with actinomycin-D was begun in 1954, and vincristine was added in 1963. In 1956, Farber, using a combination of surgical excision, postoperative irradiation, and chemotherapy, ushered in the modern era with a report of 2-year survival of 81%.

Neuroblastoma is one of the most common solid tumors in infancy and childhood. This neoplasm, of neural crest origin, may arise along the adrenal medulla and the sympathetic ganglion chain from the

neck to the pelvis. The clinical course varies as this highly malignant tumor demonstrates unusual behavior. Although instances of spontaneous regression and tumor maturation from malignant to a benign histologic form have been observed. In many cases, the disease is progressive. Although survival in children with other malignancies, such as Wilms' tumor, rhabdomyosarcoma, leukemia, Hodgkin's disease, and non-Hodgkin's lymphoma, has been significantly improved by the aggressive use of combined modalities of cancer treatment, the outlook for many children with advanced neuroblastoma remains dismal.

Neuroblastoma in situ in the adrenal gland is seen in 1 of every 260 neonates dying of congenital heart disease and in as many as 1 in 39 infants in the first 3 months of life who die from other causes. The clinical incidence of the tumor is approximately 1 in 8,000 to 10,000 children. The cause remains unknown. Ninety percent of cases occur within the first 7 to 8 years of life. More than half the patients are under 2 years of age at the time of diagnosis. Neuroblastoma is slightly more common in boys than girls, with a ratio of 1.2:1.0. It is the most common intraabdominal malignancy in the newborn. The embryonal nature of this tumor has been documented by identification of neuroblastoma on prenatal ultrasound, and the tumor has been known to invade the placenta during the antenatal period.

In infants born with congenital neuroblastoma, flushing and hypertension have been found in women during pregnancy. The neoplasm has been described in twins on many occasions, and familial occurrences in both mother and child and father and son have been reported. Neuroblastoma has been reported in infants with Beckwith-Wiedemann syndrome (BWS), Hirschsprung's disease, fetal alcohol syndrome, and in offspring of mothers taking phenytoin (fetal hydantoin syndrome) for seizure disorders.

Neuroblastoma may occur at any site where neural crest tissues are found in the embryo. The neuroblast is derived from primordial neural crest cells that migrate from the mantle layer of the developing spinal cord. Tumors may arise in the neck, posterior mediastinum, retroperitoneal (paraspinal) ganglia, adrenal medulla, and pelvic organ of Zuckerkandl. In 75% of the cases, the tumor is located in the retroperitoneum either in the adrenal medulla (50%) or in the

paraspinal ganglia (25%). In 20% of cases, the tumor is in the posterior mediastinum. Fewer than 5% of the tumors occur either in the neck or the pelvis.

List of recommended books.

1. Yu. F. Isakov. − Paediatric surgery, 1986.
2. Oldham, Keith T.; Colombani, Paul M.; Foglia, Robert P.; Skinner, Michael A. − Principles and Practice of Pediatric Surgery, 2005
3. Mazyar Kanani,Martin Elliott – Applied Surgical Physiology Vivas, 2004.
4. McLatchie G.R. – Oxford handbook of clinical surgery, 1991.
5. Chabner D. E. − The language of medicine. 1981.
6. Noble Chamberlain E. and Ogilvie C. M. – Symptoms and signs in clinical medicine, 1967.
7. Doherty, Gerard M.; Way, Lawrence W. − Current Surgical Diagnosis & Treatment, 2006.
8. Bruce G. Wolff, James W. Fleshman, David E. Beck, John H. Pemberton, Steven D. Wexner. − The ASCRS Textbook of Colon and Rectal Surgery, 2007