Chapter 3
Pediatric Abdominal Surgery

Andrew Ford, John Hutson, and Brendon J. Coventry

General Perspective and Overview

The major difference between counseling in pediatric surgery and any other surgical specialty is that counseling is done through a third party, the parents or other significant carers/guardians. As a result, no matter what decision is made, neonates and children will be excluded from a decision that may well affect them for the rest of their lives. This is especially true when correcting major congenital anomalies in the newborn. In diaphragmatic hernia, for example, we are able to determine which fetuses may well survive, and those that are likely to die at birth, giving the parents the option of perhaps terminating the pregnancy if the lesion is discovered early enough, or even embarking on prenatal therapy. Any such counseling has to be done with caution and with many caveats, as ultrasound is not totally accurate for subtle differences from the norm. Furthermore, there may well be implications for intellectual delay in many of the fetuses with an anatomical abnormality.

The section is divided for practical purposes into (i) newborn/infant and (ii) older child. Neonates and older children often have different patterns of surgical complications. *In the newborn*, the complications of surgery are often directly related to the anatomical abnormality that has to be corrected and any associated anomalies that impact on recovery. *In the older child*, the patterns of complications are more likely to follow those seen after surgery in the younger adult, but the child is far more able

A. Ford, MB, ChB, ChM, FRCSEd, FRCS, FRACS (✉)
Division of Paediatric Surgery (Chief of Surgery), Women's and Children's Hospital, Adelaide, Australia
e-mail: andrew.ford@me.com

J. Hutson, BS, MD, FRACS, DSc, FRAP (hon)
Department of Paediatrics, Royal Children's Hospital, Melbourne, Australia

B.J. Coventry, BMBS, PhD, FRACS, FACS, FRSM
Discipline of Surgery, Royal Adelaide Hospital, University of Adelaide, L5 Eleanor Harrald Building, North Terrace, 5000 Adelaide, SA, Australia
e-mail: brendon.coventry@adelaide.edu.au

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to tolerate and recover from surgery than an adult – both physically and psychologically. This division is, of course, not absolute, and surgery usually performed in the older child may need to be performed in the newborn or infant on occasions. Likewise, surgery performed in the newborn or infant may need to be performed, or perhaps even repeated later, in the older child in special circumstances.

The *neonate* is physically primed to tolerate the massive physiological changes that occur at birth, and as a result, neonates appear to tolerate the physiological changes associated with surgery that may prove lethal to most adults (Rowe 1998). Nevertheless, their immune system is immature (Grant et al. 1997), they may have hereditary congenital diseases, and they may have anatomical anomalies that impact dramatically on the success or failure of any surgery.

Neonates with congenital anatomical anomalies often have more than one. Take, for example, esophageal atresia (an incomplete esophagus). This anatomical anomaly can be associated with anorectal abnormalities, vertebral anomalies, urinary tract anomalies, upper limb deformities, and most importantly major cardiac defects that may prove lethal (Harmon et al. 1998).

Neonates may also have underlying hereditary problems that not only predispose them to surgical problems but may also predispose them to a prolonged and complicated recovery. For example, babies with cystic fibrosis have a 14 % chance of having a small bowel obstruction at birth (meconium ileus), which will often require a laparotomy (Efrati et al. 2010). These babies may also have perforated their intestine before birth causing meconium peritonitis and massive dense adhesions (Rescorla et al. 1998) or may also have undergone an antenatal segmental volvulus. After any surgery, all babies with cystic fibrosis are at risk of progressive lung damage, which is accelerated by their requirement for a longer postoperative ventilation period than other babies.

Preterm delivery adds to the problems of any surgery and induces surgical problems of its own (Rowe 1998). Anatomical anomalies can also induce preterm labor and delivery. For example, esophageal atresia blocks swallowing and induces polyhydramnios, which in turn can trigger preterm labor. Thus, the birth weight of babies with esophageal atresia is frequently below 2.0 Kg and is often close to 1.0 Kg. Therefore, after surgery, ventilation is required which adds to the preoperative problems of gastric contents soiling the lungs from below (coming up the commonly present fistula from the stomach to the trachea) and saliva entering the lungs from above, as it cannot go down the blocked esophagus (Also see section on "Esophageal Atresia" later in this Volume).

Premature babies have diminished immune function and a very labile vascular system. When stressed a premature baby may well shut down the blood supply to the intestine and produce malfunction of the mucosal barrier in the small bowel. As a result, bacteria within the gut lumen can get into the intestinal wall and set up an active and proliferating infection. This infection is frequently with gas-producing organisms, so "gas gangrene" of the intestinal wall is the result – "necrotizing enterocolitis" (NEC) (Albanese et al. 1998). While this would appear to be a terrible situation where a tiny premature baby (some as small as 500 g) has gas gangrene of the intestinal wall, these babies respond well to antibiotics, IV feeding, and gut rest

and may even survive a laparotomy with massive gut resection. This physiological insult does, however, enhance the likelihood of the extremely small premature baby having "intellectual delay" (Chacko et al. 1999) (see section on "NEC").

So, there are distinct advantages and disadvantages to carrying out surgery in the newborn period. On the whole, however, the full-term neonate responds well to surgery and survives that surgery surprisingly well; the premature may survive, but may be compromised. This improved tolerance to surgery extends to some extent into older children, so that the otherwise well child is a far better candidate for routine surgery than an adult, in general terms. The healing process is rapid, and there are rarely the psychological issues that appear to slow recovery in some adults.

But again, there are chronic diseases in the child that predispose to a poor outcome and predispose to the requirement for surgery in the first place. One common example would be cerebral palsy. These children often suffer severe intellectual delay and lie all day in contorted positions because of spasticity. They are prone to severe gastroesophageal reflux and feeding difficulties. As a result they often come to fundoplication with a gastrostomy or either one alone. But, cerebral palsy is associated with a poor outcome from that form of surgery, and there is a 30–50 % failure rate for fundoplication in this group (Boix-Ochoa et al. 1998).

Pediatric patients and surgical operations are often different from many procedures that are performed in adults, although many similarities, of course, exist. Diagnosis is frequently and increasingly within the antenatal period for a range of congenital conditions and anomalies. Even surgery is able to be performed in utero for some conditions, but many are corrected after birth. Disorders that require immediate surgery may be true emergencies or are virtually semi-elective in nature, where rapid patient optimization, or even a period of stabilization or growth, is preferable before surgery. As mentioned before, the relative *lack of ability to communicate* directly with the patient is another fundamental difference, necessitating the absolute reliance on the parent, guardian or State to make decisions on the child's part, with the advice of the surgeon, medical practitioners and other advisors.

Especially with many types of neonatal surgery, the options are extremely limited should surgery not be possible or performed, and often the natural consequence of not performing surgery is *death*. Examples of these situations are severe forms of esophageal atresia, biliary atresia, liver tumors, neuroblastoma, large congenital diaphragmatic hernia, exomphalos (also termed omphalocele), and pyloric stenosis. Despite surgery, death may still ensue.

Parent's expectations may not be realistic, and their individual understanding of the situation that their child is in may not be complete or even adequate to make rational and informed decisions. The situations are often emotionally charged, and the clinician(s) is always dependent on the capacity and willingness for understanding, especially in situations where the options with and without surgery are dire and carry high risks of morbidity and mortality. Realization of this must be kept in mind when medicolegal considerations are made.

The situation and available facilities are important too, as these are not equivalent for all patients in all countries throughout the world.

With these factors and facts in mind, the information given in these chapters must be appropriately and discernibly interpreted and used.

The **use of specialized units with standardized preoperative assessment, multidisciplinary input, and high-quality postoperative care** is essential to the success of complex pediatric surgery overall and can significantly reduce risk of complications or aid early detection, prompt intervention, and cost.

Many of the procedures explained here are of a more "straightforward" nature and are elective, so that the time and ability to explain to the parents and even the patient (for older children) is often more realistic and practical. In emergency settings, the ability to do this and assimilate information may be different.

Important Note
It should be emphasized that the risks and frequencies that are given here *represent derived figures*. These *figures are best estimates of relative frequencies across most institutions*, not merely the highest-performing ones, and as such are often representative of a number of studies, which include different patients with differing comorbidities and different surgeons. In addition, the risks of complications in lower- or higher-risk patients may lie outside these estimated ranges, and individual clinical judgment is required as to the expected risks communicated to the patient and staff or for other purposes. The range of risks is also derived from experience and the literature; while risks outside this range may exist, certain risks may be reduced or absent due to variations of procedures or surgical approaches. It is recognized that different patients, practitioners, institutions, regions, and countries may vary in their requirements and recommendations.

Important Caveat
The section contains a description of the more common diseases and procedures carried out in the pediatric age group, but cannot be exhaustive. There are diseases and procedures that are carried out in both children and adults, but for those that are more common in young adults, readers are referred to that section.

Pediatric Surgery in the Newborn and Infant

Introduction

Abdominal wall defects in the newborn and infant can vary from a small congenital umbilical hernia due to failure of the umbilical cicatrix to close completely by birth to major failure of the abdominal wall to develop with complete herniation of the exposed abdominal contents. The former usually closes by 2 years of age, while the

larger defects may not be compatible with life, with or without surgery, especially with coexistence of other congenital anomalies. Small abdominal wall herniae are covered under the section "Pediatric Surgery in the Older Child," but may apply to surgery in the newborn and infant in certain situations where early repair is advisable. The major defects of the abdominal wall are essentially all surgically repaired early in life and are included here in this section of the chapter, although further surgical procedures may be required into childhood and adulthood. Inguinal herniae can occur at any age in the child or adult, but are especially liable to strangulate in children under 1 year of age, so are typically repaired promptly after diagnosis, so are described under this section, but may apply for the older child too.

General abdominal surgical procedures in the newborn and infant include a variety of procedures principally related to obstruction of the tracts of the gut or biliary systems arising from, for example, atresia, intussusception, hypertrophy, or malrotation; infections; or internal herniation. Although these are principally surgical procedures performed in the newborn and infant, they may be performed or revised in the older child also. Some of the surgical procedures described in section "Pediatric Surgery in the Older Child" may apply also to surgery in the newborn and infant in certain situations, such as gastrostomy or appendectomy.

Pediatric Surgery in the Newborn and Infant

Abdominal Wall Defect Surgery

Open Herniotomy (Inguinal Hernia "Repair")

Description

Open inguinal hernia correction in children is performed under general anesthesia. Inguinal herniae occur through the anterior abdominal wall, via the deep inguinal ring, when a patent processus vaginalis persists as it follows the testicle down to the scrotum. Herniotomy is a relatively simple procedure that ligates the peritoneal sac at its origin at the deep inguinal ring and does not aim to repair the defect itself, as opposed to the aim in adult inguinal hernia repair which repairs the defective muscle wall. Because inguinal herniae in the first year of life are especially at risk of strangulation, prompt repair is advisable. In older children, unless symptomatic, elective repair within a month or two is advisable.

Anatomical Points

In children under one year of age, the deep and superficial inguinal rings overlie each other, and the peritoneal sac protrudes almost directly outward beneath the skin. In older children, the deep inguinal ring moves further and further laterally so

that it is approximately deep to the midpoint of the inguinal ligament, so the inguinal canal is formed. This contains the spermatic cord in the male or the vestigial processus vaginalis in the female (some refer to this as the gubernaculum), and herniae arising at the deep ring transit indirectly from the deep toward the superficial opening.

It is rare for the defect to be big in the child, so that a simple herniotomy is effective. No strengthening procedure such as mesh is required. The size of the defect may be bigger in the ventilated premature; otherwise, the defect is usually small. In the male, the hernia contains bowel or omentum. In males, inguinal herniae are the most common surgical condition, usually within the first year (particularly the first 3 months) of life, with about 60 % right sided, 25 % left, and 15 % bilateral. About 12 % of all indirect inguinal herniae occur in females, where the hernia usually contains the ipsilateral ovary and the fallopian tube. Rarely the appendix (nearly always the ovary in the female) may insinuate into the sac. The testis in the neonatal male may not have descended properly, making it necessary to do an orchidopexy at the same time, so as to avoid scar tissue holding the testicle up and making subsequent orchidopexy very difficult.

It is important to realize that in the child, the gonad is at risk of infarction. The testis in the male due to incarcerated bowel presses on the testicular veins, and in the female, the ovary and tube can tort on their long axis. It is also important to realize that the younger the child, the smaller the ring(s), the more likely it is that the bowel will incarcerate and therefore put the gonad at risk. Therefore, the younger the child presents with a hernia, the sooner the surgery should be done. In the newborn, this should be within days. There is no place for the "wait until the child is older and bigger for the surgery" approach.

Perspective

See Table 3.1. Herniotomy is a much simpler procedure than open inguinal hernia repair in adults. Complications from herniotomy are infrequent and usually minor in nature. However, precise recording of these is limited by under-reporting. Spermatic cord injury occurs in at least 1 in 200 children (as the histopathology of the resected sac does not take into account internal blockage of the vas from handling). Therefore, the practice of exploring both sides in males under 2–4 years of age is disappearing. Bowel injury is rare, but when it does occur in the compromised ventilated neonate, it can trigger necrotizing enterocolitis and major ongoing problems. Infection and bleeding are other complications, but are usually minor. True recurrence rates are not known, as the follow-up must extend into later adult life to accurately analyze these, and most patients are lost to late follow-up. But, within childhood, the recurrence rate for healthy children could be as low as 0.1 % for open surgery and 3–5 % for laparoscopic closure of the deep ring, thereby implying that laparoscopic closure has a greatly increased recurrence rate in children. In the male, an asynchronous hernia occurs in approximately 8 % on the right if the index (initial) hernia repair was on the left and 4 % on the left if the previous

Table 3.1 Open herniotomy (inguinal hernia "repair") estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Bleeding or hematoma formation[a]	1–5 %
Hernia recurrence[a] (10 year)	
For open surgery	0.1–1 %
For laparoscopic surgery	1–5 %
Testicular ischemia, testicular atrophy	
Incarcerated, but forcibly reducible	1–5 %
Incarcerated, but forcibly not reducible	5–20 %
Ovarian torsion/loss (ovarian loss in the newborn has been reported to be up to 14 %)	5–20 %
Small bowel obstruction	
In the newborn	1–5 %
In > newborns	0.1–1 %
Laparotomy[a] (bowel injury or adhesion-related strangulation or ischemia as for the newborn)	1–5 %
Rare significant/serious problems	
Infection[a]	0.1–1 %
Neural injury[a]	
Ilioinguinal nerve	0.1–1 %
Iliohypogastric nerve	0.1–1 %
Vascular injury – artery or vein	0.1–1 %
Spermatic cord injury[a] (parts of vas have been found in 0.5 % of operative specimens of the sac)	0.1–1 %
Less serious complications	
Pain/discomfort/tenderness(<2 months; usually only days)[a]	5–20 %
Pain/discomfort/tenderness(>2 months)	0.1–1 %
Scrotal/labial swelling	5–20 %
Urinary retention	0.1–1 %
Dehiscence[a] (rare, may occur after testicular infarction)	<0.1 %
Dimpling/deformity of the skin[a]	0.1–1 %
Wound scarring (all)	0.1–1 %
Drain tube(s)[a]	0.1–1 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

index repair was on the right. Gonadal infarction and injury is usually a consequence of the problem, rather than a surgical complication per se, but is important to discuss preoperatively where practicable.

Major Complications

The **loss of the gonad** from venous infarction or torsion is a serious consequence. **Bowel injury** is rare in children, but can be significant especially after bowel incarceration, obstruction, or infarction with perforation. In the neonate, an appendix in

the hernia can cause problems with deep-seated and systemic infection (and the rare T-antigen exposure) if the appendix has infarcted in the hernia, which cannot be predicted preoperatively. **Infection** also increases the risk of **dehiscence**, especially if nonabsorbable sutures are used.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Urinary obstruction*
- Risk of other abdominal organ injury*
- Gonad loss
- Hernia recurrence*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Exomphalos (Omphalocele)

Description

Exomphalos (also termed omphalocele) is an abdominal wall defect that is nearly always picked up by antenatal ultrasound, if one has been done. Exomphalos occurs in approximately 1: 5,000–6,000 live births. There is a field defect in the abdominal wall, which may be small – "exomphalos minor" – or may be large and remain within the abdominal area – "exomphalos major" (which is large enough to contain liver). One subgroup, the "giant exomphalos," has a lesion bigger than the head, the lungs are hypoplastic, and the mortality is high. An even more massive defect is where the lesion extends up into the chest – the "thoracoabdominal cleft." As the exomphalos with its field defect extends further and further, so more and more structures are involved, and the mortality rises. With thoracoabdominal cleft, for example, when picked up antenatally, most fetuses will die before birth, and those that are live-born will have a high mortality.

In a large exomphalos, there is a broad-based midline defect that can extend extensively up and down the abdomen, thorax, or pelvis, with defects in the organs within that field. The lesion is covered by a membrane that is probably the remnant of the ectoderm and the endoderm, where the muscle somites did not grow between these two layers. Severe versions in the lower abdomen are often associated with exstrophy of the urinary bladder; severe lesions in the upper abdomen and thorax are associated with exstrophy of the heart, heart defects, midline chest wall defects, and underdeveloped lungs. There may be associated lethal chromosome defects, but these are surprisingly more often associated with the smaller exomphalos minor.

The closure of a major exomphalos (one containing most of the liver) or giant exomphalos (where the lesion is bigger than the baby's head) can be very difficult. Where the exomphalos is large, a staged procedure is often used to gradually force the viscera back into the abnormally small abdomen over a period of 7–10 days. The covering membrane is usually removed at birth to be replaced with a tough Silastic sheet or polyvinyl bag (the silo), which in turn is used to force the viscera back into the abdomen as fast as can be tolerated. Usually a muscle or fascial sheath (using flaps of rectus sheath turned medially) can be achieved, but for giant exomphalos, the procedure may have to be staged over years after initial primary skin closure alone. The mortality can be high, especially with large, repeated, or complicated procedures.

Anatomical Points

For omphalocele (exomphalos) additional defects are present in up to 70 % of patients (higher when detected in utero, as the most complex cases die in utero). The size of the defect determines the anatomical disturbance necessitating surgical correction. The extent of herniation and amount of abdominal wall available for reconstruction can vary considerably with the degree of the field defect involved. Accordingly, the surgery has to be individualized. For thoracoabdominal clefts, the heart defects, the pericardial defect, the diaphragmatic defects, and pulmonary hypoplasia increase risks and may make surgery almost impossible.

Perspective

See Table 3.2. Exomphalos, in its various forms, has complications proportional to the amount of anatomical disturbance and associated organ defects. The early complications of abdominal wall repair are mainly related to respiratory insufficiency, either due to the force used to reduce the viscera or due to primary pulmonary hypoplasia or both. Infection can be problematic when the defect cannot be closed quickly, especially where the closure is not ideal. Silastic may be used as a device to force the contents in, as a temporary device to get the abdomen closed placed beneath the skin, but it commonly becomes infected. Staged repairs are used and collagen matrix "Surgi-sis®" is getting some usage to leave a stronger scar where the muscles cannot be brought together for exomphalos. Absorbable materials are preferred for closure where possible, and nonabsorbable patch materials are usually avoided. For patients in multi-organ failure, skin closure with later definitive surgery is often used. Nonabsorbable patches nearly always become infected and nonabsorbable sutures may cause suture erosion where there has been difficult closure. Even "PDS®" (polydioxanone suture) may last too long and cause problems. There are often small incisional herniae especially where there has been a flapped fascial repair. Where the muscles cannot be brought together at the first session, then a large ventral hernia has to be dealt with as a later staged repair. Even if the muscles can be brought together, they often drift apart again leaving a low-profile dome of scar tissue with the centrally placed liver lying directly underneath this. Inability to close the abdominal wall with tearing of the Silastic sac from the abdominal wall

Table 3.2 Surgery for exomphalos (omphalocele) estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection (higher with more severe defects)[a]	20–50 %
Bleeding or hematoma formation[a]	5–20 %
Numbness/altered sensation	1–5 %
Hernia recurrence[a] (10 year)	1–5 %
Cardiorespiratory failure (especially high with congenital heart disease)[a]	5–20 %
Multisystem organ failure (especially high with congenital heart disease)[a]	5–20 %
Prolonged ventilation[a]	>80 %
TPN (total parenteral nutrition)[a]	50–80 %
Suture abscess +/− suture sinus[a]	1–5 %
Small bowel obstruction (later)	20–50 %
Dehiscence[a]	20–50 %
Death[a]	20–50 %
Less serious complications	
Pain/discomfort/tenderness(<2 months)	20–50 %
Pain/discomfort/tenderness(>2 months)	0.1–1 %
Seroma formation	5–20 %
Scarring/dimpling/deformity of the skin[a]	>80 %
Drain tube(s)[a]	50–80 %

[a]Dependent on underlying pathology, disease extent, anatomy, surgical technique, and preferences

muscles can occur, causing subsequent increased difficulty in later surgery, with sepsis and multi-organ failure. Most babies have to be ventilated for a prolonged period postoperatively and especially where the viscera are being forced back in. Total parenteral nutrition (TPN) is often required, with its associated metabolic and septic complications. Death occurs in about 30 % of live-born babies with a giant exomphalos, and in utero mortality occurs in about 50 % of complex patients detected on antenatal ultrasound, that is, death occurs before or at birth.

Major Complications

Death, early and late **bowel obstruction, failure to close the defect, respiratory insufficiency, and ureteric obstruction** can arise from the pressure being used**. Infection** may be a problem, with skin organisms predominating, unless bowel injury has occurred. The presence of a major foreign body (the silo) may cause infection where it is sutured to the muscle and can contaminate the bowel in the silo. Nonabsorbable sutures can increase bacterial colonization and the risk of infection. **Bleeding** is seldom severe unless an omental or organ injury occurs. **Portosystemic anastomoses** around the umbilicus can produce annoying bleeding, but is rarely severe. **Hernia recurrence** rates for these herniae are rather high and further increased in the presence of extensive abdominal wall deficiencies and infection.

When an exomphalos major is being forced back, then **abdominal compartment syndromes** can arise from high pressures. If the liver is being forced more than the rest of the viscera, then **liver infarction** can occur, especially in those with anatomical anomalies of the blood supply. Where the pressure is more severe below the liver, the **ureteric obstruction** can arise causing postrenal failure. Obviously the pressure being utilized has to be tailored to achieve reduction, while avoiding these major issues. **Death** is a serious risk without surgery, but postoperative **multisystem organ failure** and **sepsis** remain major determinants of mortality.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Urinary obstruction*
- Risk of organ ischemia/infarction
- Risk of other abdominal organ injury*
- Possible further surgery*
- Hernia recurrence
- Risks without surgery*
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Gastroschisis

Description

Gastroschisis is a condition that is a mechanical accident where the midgut loop ruptures out through the side of an apparently normal umbilical cord with a normal abdominal wall. This leads to protrusion of small bowel and occasionally other viscera into the amniotic cavity, from the base of the umbilical cord. The defect is becoming much more common and appears to be a vascular accident, where the frequency has been shown to increase in young mothers taking vasoactive drugs, such as cocaine. The incidence of gastroschisis in the general female reproductive age range is about 1 per 5,000 live births, but 1 per 200 live births in pregnant women under 18 years of age.

Because the intestine is protruding, irritated by amniotic fluid, it is thick walled, edematous, and shortened. As a result, there is a prolonged period of bowel dysmotility after birth that can last for life. Sections of the gut can lie over the sharp edge of the small defect and can infarct in utero. Short bowel syndrome may then occur so that 5–10 % may die or require a small bowel transplant. Up to 70 % of

gastroschisis patients can have the herniated viscera returned to the abdomen primarily without the use of polyvinyl bag "silos" as the abdominal wall has its normal potential.

Anatomical Points

The size of the defect can vary, as can the site, degree and length of bowel affected. Anatomical variants causing vascular or bowel obstruction may complicate the surgical anatomy and dictate resection of small bowel or more complex repairs, with attendant associated complications.

Perspective

See Table 3.3. Gastroschisis is not usually associated with pulmonary hypoplasia, so lung problems are less frequent overall. The abdominal cavity is fully developed, but tends to be small because the gut has not been present inside to expand it, as the body grows. Even staged closure is relatively easy in comparison to exomphalos. Gastroschisis is a problem especially for young mothers. They require a great deal of care to get the child growing properly, and nearly all have problems with constipation because of the motility problems. Those with short bowel are often hospitalized frequently and for months, with or without transplantation. The abdominal wall repair can be associated with respiratory insufficiency, primarily due to the tightness when the viscera are reduced. Infection can occur from cutaneous or bowel organisms, especially when bowel injury occurs in utero or at surgery. Abdominal prosthetic patches are rarely required, and absorbable sutures are typically preferred.

Major Complications

Infection may be a problem, with skin organisms predominating unless bowel injury has occurred. Since foreign material is seldom used, infection rates are generally lower than for exomphalos repairs. Nonabsorbable sutures can increase bacterial colonization and the risk of infection. **Bleeding** is seldom severe unless an omental, bowel, or organ injury occurs. **Hernia recurrence** rates for these herniae are moderate and further increased in the presence of infection, poor healing, multisystem organ failure, or nutritional deficiency. Lifelong immunosuppressants following SB transplantation or home **chronic TPN** may be necessary if short bowel syndrome occurs. **Liver failure** may ensue. Overall, **multisystem organ failure** and **death** are not common. **Nutritional deficiency**, **short bowel syndrome,** and **bowel dysmotility** can be significant chronic problems. Repeated or **prolonged hospitalization** can be a significant consequence.

Table 3.3 Surgery for gastroschisis estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection (higher with more severe defects)	20–50 %
Bleeding or hematoma formation[a]	5–20 %
Numbness/altered sensation	1–5 %
Hernia recurrence[a] (10 year)	1–5 %
Cardiorespiratory failure (especially high with congenital heart disease)[a]	5–20 %
Multisystem organ failure (especially high with congenital heart disease)[a]	5–20 %
Prolonged ventilation[a]	50–80 %
TPN (total parenteral nutrition)[a]	50–80 %
Bowel dysmotility[a]	>80 %
Malabsorption and failure to thrive[a]	5–20 %
Suture abscess +/− suture sinus[a]	1–5 %
Small bowel obstruction (later) or pseudo-obstruction for life[a]	20–50 %
Multiple hospital admissions (for all complications)[a]	20–50 %
Liver failure (often from short bowel syndrome and TPN)[a]	5–20 %
Small bowel transplant[a]	5–20 %
Dehiscence[a]	20–50 %
Death[a]	5–20 %
Less serious complications	
Pain/discomfort/tenderness(<2 months)	20–50 %
Pain/discomfort/tenderness(>2 months)	0.1–1 %
Seroma formation	5–20 %
Scarring/dimpling/deformity of the skin[a]	1–5 %
Drain tube(s)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Nutritional deficiency
- Risk of other abdominal organ injury*
- Hernia recurrence
- Possible further surgery*
- Risks without surgery*
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

General Abdominal Surgery

Open Pyloromyotomy for Pyloric Stenosis

Description

General anesthesia is used. Pyloric stenosis is hypertrophy of the pyloric muscle and occurs in approximately 1:200 male and about 1:1,000 female live births (i.e., 85 % are male). It typically develops in the first 4–6 weeks of life in the first-born male. The aim is to divide the constricting hypertrophic muscle that encircles the pyloric region, using a longitudinal (Ramstedt) pyloromyotomy, exposing the underlying intact mucosa. The pyloromyotomy allows drainage by incising longitudinally, approximately 1–2 cm, through the muscle wall of the pyloric canal from immediately proximal to the duodenal bulb and then back to the antrum. This longitudinal incision deliberately leaves the muscle wall gaping, down to the intact mucosa, which then balloons into the muscle defect allowing passive drainage of gastric contents.

Anatomical Points

The anatomy of the pylorus is relatively constant; however, the extent of the pyloric hypertrophy may vary, proximally up the antral wall. The hypertrophy always ends abruptly at the junction of the pylorus and duodenum, with no duodenal extension, and there is a normal duodenal bulb. The degree of preoperative obstruction, projectile vomiting, and consequent acid-base and electrolyte imbalance will vary considerably and has to be corrected prior to surgery.

Perspective

See Table 3.4. Most of the complications are of a minor nature, and children usually recover rapidly from surgery. Major complications are related to perforation of the mucosa, wound infection, recurrence of pyloric obstruction, and rarely later complications of small bowel obstruction. Correction of preoperative electrolyte disturbances and ensuring adequate nutrition are important considerations.

Major Complications

Inadvertent **perforation** and **leakage** may be serious and occasionally not recognized at operation, leading to **intra-abdominal infection** and **abscess** formation. If the perforation is recognized and closed during operation, the risk of infection is approximately doubled, but if not recognized, then intra-abdominal sepsis is almost inevitable. **Infection and multisystem failure** may be catastrophic, although **death**

Table 3.4 Open pyloromyotomy for pyloric stenosis estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	
Subcutaneous/wound	5–20 %
Intra-abdominal	0.1–1 %
Systemic	0.1–1 %
Bleeding or hematoma formation[a]	1–5 %
Wound breakdown and evisceration	5–20 %
Perforation of the mucosa or duodenum	5–20 %
Reflux esophagitis	5–20 %
Delayed gastric emptying[a]	20–50 %
Recurrence/persistence of pyloric obstruction[a]	1–5 %
Mortality[a]	0.1–1 %
Mortality without surgery	20–50 %
Rare significant/serious problems	
Liver injury[a]	0.1–1 %
Small bowel obstruction (early or late; lifetime)[a] [Adhesion formation]	0.1–1 %
Subphrenic abscess	<0.1 %
Multisystem failure (renal, pulmonary, cardiac failure)	<0.1 %
Less serious complications	
Paralytic ileus	1–5 %
Slow recovery requiring prolonged feeding with small frequent feeds	5–20 %
Wound scarring (poor cosmesis/wound deformity)	1–5 %
Incisional hernia	5–20 %
Prolonged use of nasogastric tube[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

is very rare. **Bleeding** is rare and is usually controlled at surgery. **Wound infection** is common and occurs especially in malnourished infants with severe prolonged vomiting prior to presentation and diagnosis. **Wound dehiscence** and consequent **hernia formation** may result. **Persistent pyloric stenosis** can result from inadequate division of the hypertrophic muscle, but true **recurrent pyloric stenosis** is very rare. **Small bowel obstruction** is rare but may occur from postoperative adhesions, even many years later.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*

- Risk of other abdominal organ injury*
- Perforation*
- Recurrence of stenosis
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Laparoscopic Pyloromyotomy for Pyloric Stenosis

Description

General anesthesia is used. Pyloric stenosis is hypertrophy of the pyloric muscle and occurs in approximately 1:200 male and about 1:1,000 female live births (i.e., 85 % are male). It typically develops in the first 4–6 weeks of life in the first-born male. The aim is to divide the constricting hypertrophic muscle that encircles the pyloric region, using a longitudinal (Ramstedt) pyloromyotomy, exposing the underlying imperforated mucosa. The pyloromyotomy allows drainage by incising longitudinally, approximately 1–2 cm, through the muscle wall of the pyloric canal from immediately proximal to the duodenal bulb and then back to the antrum. This longitudinal incision deliberately leaves the muscle wall gaping, down to the intact mucosa, which then balloons into the muscle defect allowing passive drainage of gastric contents. Approximately half of all pyloromyotomies are now carried out laparoscopically even in premature babies. This frequency will continue to increase, as it is a good training procedure for younger surgeons starting laparoscopy in pediatric patients. In all babies (down to 1.0 Kg), there is no need for access ports other than for the 3 mm telescope. The other 3 mm instruments are placed directly through small stab wounds so that the cosmetic results are superior.

Anatomical Points

The anatomy of the pylorus is relatively constant; however, the extent of the pyloric hypertrophy may vary, proximally up the antral wall. The hypertrophy always ends abruptly at the junction of the pylorus and duodenum, with no duodenal extension, and there is a normal duodenal bulb. The degree of preoperative obstruction, projectile vomiting, and consequent acid-base and electrolyte imbalance will vary considerably and has to be corrected prior to surgery. Adhesions from previous surgery or an enlarged liver may make laparoscopic access more difficult.

Perspective

See Table 3.5. Most of the complications are of a minor nature, and children usually recover rapidly from surgery. With laparoscopic approaches, the risk of wound

Table 3.5 Laparoscopic pyloromyotomy for pyloric stenosis estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	
Subcutaneous/wound	5–20 %
Intra-abdominal	0.1–1 %
Systemic	0.1–1 %
Bleeding or hematoma formation[a]	1–5 %
Wound breakdown and evisceration	5–20 %
Perforation of the mucosa or duodenum	5–20 %
Reflux esophagitis	5–20 %
Delayed gastric emptying	20–50 %
Conversion to open operation	1–5 %
Recurrence/persistence of pyloric obstruction	1–5 %
Mortality without surgery	20–50 %
Rare significant/serious problems	
Pneumothorax	0.1–1 %
Gas embolus	0.1–1 %
Liver injury	0.1–1 %
Small bowel obstruction (early or late; lifetime)[a] [Adhesion formation]	0.1–1 %
Subphrenic abscess	<0.1 %
Multisystem failure (renal, pulmonary, cardiac failure)	<0.1 %
Mortality	0.1–1 %
Less serious complications	
Paralytic ileus	1–5 %
Slow recovery requiring prolonged feeding with small frequent feeds	5–20 %
Surgical emphysema	1–5 %
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Port-site herniae	5–20 %
Wound scarring (poor cosmesis/wound deformity)	1–5 %
Incisional hernia	5–20 %
Prolonged use of nasogastric tube[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

infection, wound dehiscence, and incisional hernia is reduced. Major complications are related to perforation of the mucosa, wound infection, recurrence of pyloric obstruction, and rarely later complications of small bowel obstruction. Correction of preoperative electrolyte disturbances and ensuring adequate nutrition are important considerations. With laparoscopy, there is an increased risk of mucosal perforation in the pyloric canal and of tears to the duodenal cap as the grasper holds the duodenum during division and spreading of the very mobile, hypertrophied pylorus muscle. Laparoscopic entry is associated with the complication of herniation of the omentum, or rarely bowel, through any of the port-site wounds, of about 5–10 %.

Major Complications

Inadvertent **perforation** and **leakage** may be serious and occasionally not recognized at operation, leading to **intra-abdominal infection** and **abscess** formation. If the perforation is recognized and closed during operation, the risk of infection is approximately doubled, but if not recognized, then intra-abdominal sepsis is almost inevitable. **Infection and multisystem failure** may be catastrophic, although **death** is very rare. **Bleeding** is rare and is usually controlled at surgery. **Wound infection** is common and occurs especially in malnourished infants with severe prolonged vomiting prior to presentation and diagnosis. **Wound dehiscence** and consequent **hernia formation** may result. **Persistent pyloric stenosis** can result from inadequate division of the hypertrophic muscle, but true **recurrent pyloric stenosis** is very rare. **Small bowel obstruction** is rare but may occur from postoperative adhesions, even many years later. **Gas embolus** and **major vascular injury** are additional serious, although very rare, complications of the laparoscopic approach. **Conversion to open operation** is a small but significant risk rather than a complication per se.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Perforation*
- Recurrence of stenosis
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Congenital Diaphragmatic Hernia Repair

Description

General anesthesia is used. Congenital diaphragmatic hernia is a defect in the diaphragm that occurs before birth, leaving a free communication between the chest and abdominal cavities. The most common form is a posterolateral defect through the foramen of Bochdalek and occurs on the left side in over ¾ of cases. Abdominal contents in the chest often inhibit lung development and growth. After birth, this lack of lung volume and maturation makes gas exchange difficult or impossible.

Even if the baby survives the immediate period after birth, higher than normal pulmonary vascular resistance promotes persistence of the right to left ductal and cardiac shunting of blood through a ductus arteriosus and patent foramen ovale (persistence of fetal circulation). The size of the defect and degree of herniation of contents are directly associated with the risk of complications and mortality. Larger-sized defects are less likely to close. The larger the defect, the more medially it extends until the liver herniates upward through a large hole with medial extension. If the liver has rotated up into the chest, then the baby is more likely to die than if the liver is in the abdomen. CDH is often associated with many other abnormalities: cardiac and chromosomal which often prove to be lethal. The mortality is probably 30–50 % for those detected antenatally (as long as there is no other lethal defect – the isolated CDH), but for those seen to have liver in the chest on antenatal ultrasound, the mortality is greater than 90 %.

Emergency surgery is no longer the treatment of choice. The patient must be stabilized prior to surgery, and this may take days or even weeks. When there is sufficient respiratory reserve for the patient to be able to tolerate the surgery, then a transverse upper abdominal incision is used. In those that have a good prognosis, closure of the defect is usually straightforward as the defect will not be big, and there will usually be good muscle shelves that can be unfolded. If the defect is large or the diaphragm is completely absent, then partial closure can be obtained as above, and the rest (or the whole diaphragm) can be closed using an abdominal wall flap or Gerota's fascia (the perirenal fascia is dissected up from the lower pole of the kidney to the top, leaving the attachment to the upper pole of the kidney as a hinge from the kidney which has become more popular recently). Using a muscle flap leaves a weakness in the abdominal wall that produces a permanent bulge and possibly scoliosis later in life as the child grows. Use of Gerota's fascia has only been introduced recently, but appears to be an effective closure without such side effects. Closing the defect with synthetic material has been used in the past, but this material does not grow with the child so that there is a very high rate of recurrence as the attachments give way. Suturing the anterior ribs to the posterior ribs at the lateral end of the defect can be used, but may lead to severe chest wall deformity and a small volume stomach later, as there is no space between the ribs for the stomach to act as a reservoir.

Anatomical Points

The diaphragm is formed from the pleuroperitoneal membranes, the septum transversum, the dorsal mesentery of the esophagus, and the body wall. The closure of the pleuroperitoneal canals joining the thorax and abdomen starts to occur by about the 6th week of gestation and should be complete by approximately 10 weeks of gestation. Failure to close fully leads to persistence of the posterolateral pleuroperitoneal canal – the foramen of Bochdalek – which occurs in approximately 1:2,200 conceptions and 1:4,000 live births, as many die before birth of multiple anomalies. Some 80 % occur on the left side. Herniation through the retrosternal area – foramen of Morgagni – is a rare defect.

Perspective

See Table 3.6. Death occurs in approximately 50 % of cases when CDH is discovered antenatally and higher in preterm infants. The time at which the defect stops closing determines the outcome. If the defect stops closing early in gestation, the liver rotates into the chest, and being more solid than intestine causes more damage. The abdominal viscera (liver and intestine) push the left lung up and the heart over to the right and compress the right lung as well. So both lungs are compressed. As a result both lungs are small but also undergo a maturation arrest. The larger the defect, the more immature and small the lungs and the more complex the surgery to close it effectively. The presence of additional anomalies also contributes to the mortality and complications. Delayed lung maturation leads to a lung histology that is similar to the lung development of a very preterm baby. In addition, the lungs are smaller, so the clinician has to support a full-term baby with lungs that are far too small, and have arrested development (in the pseudoglandular phase) so that ventilator-induced chronic lung disease (of the newborn) frequently occurs, but usually resolves by a year of age – in survivors. This may be responsible for late deaths weeks and months after birth. Viral infections during this period may well tip the baby into respiratory failure and death. Coexistence of cardiac anomalies increases the mortality. The neonate is typically completely unaware of this as he/she will be paralyzed, ventilated, and sedated. When CDH is detected later (up to 20 years after birth), then pain and awareness are often more of an issue.

Major Complications

Death occurs in up to 50 % of those born alive, and death has already occurred in another 20 % before birth or immediately after birth due to the basic inability to ventilate these babies. If the liver was seen to be in the chest before birth, and if the lung to head ratio is less than 0.8 on antenatal ultrasound, death before, during, or after birth is likely in more than 90 % of fetuses. In those that survive the immediate period after birth, the next major hurdle is the **high pulmonary vascular resistance**, which maintains a "persistent fetal circulation". There is often a suprasystemic pulmonary blood pressure, so that blood shunts away from the lungs at the level of the ductus arteriosus and the foramen ovale. Because of the high pressure, the right side of the heart also fails. Support therefore becomes complex, often involving pulmonary vasodilators such as inhaled nitric oxide, longer-term sildenafil therapy, and right ventricular support. Long before this occurs, the baby is often placed on oscillating **ventilation** to try to reduce lung damage, but long-term data has not shown any improvement in survival for those that are oscillated. This situation of hypoxia, shunting, and cardiac failure (leading to multi-organ failure) has a high mortality and is exceptionally difficult to treat. Secondary **lung infection** is not uncommon and may affect either lung. Viral pneumonitis is common months after the surgery, and in those that have only just survived the initial problems, the infection may well tip the baby into terminal respiratory failure. If the fluid in the almost empty left hemithorax becomes infected, this rapidly leads to generalized sepsis and multi-organ failure. Overventilation leading to **pneumothorax** or **persistent air leak** creates a situation that cannot be resolved on conventional ventilation.

Table 3.6 Surgery for congenital diaphragmatic hernia estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection	
Subcutaneous/wound	1–5 %
Intrathoracic (pneumonia; pleural)	1–5 %
Mediastinitis	0.1–1 %
Systemic	0.1–1 %
Prolonged assisted ventilation[a,b]	80 %
Gastroesophageal reflux	50–80 %
Chronic lung disease; pulmonary failure[a]	50–80 %
Diaphragmatic injury/dysfunction/paresis	5–20 %
Small bowel obstruction	5–20 %
Scoliosis and chest wall deformities (postoperative)[a] (especially where prosthetic patches or muscle flaps have been used)	5–20 %
Volvulus	1–5 %
Multisystem organ failure (renal, pulmonary, cardiac failure)[a] (ultimate cause of death in approximately 50 % of neonates)	20–50 %
Mortality	
Term infants[a]	20–50 %
Preterm infants[a] (includes those detected antenatally – usual today)	50–80 %
Mortality without surgery (almost 100 % for severe early defects)[a]	>80 %
Rare significant/serious problems	
Bleeding/hematoma formation	
Wound	0.1–1 %
Hemothorax	0.1–1 %
Pulmonary contusion	0.1–1 %
Surgical emphysema	0.1–1 %
Persistent air leak[a]	0.1–1 %
Deep venous thrombosis	0.1–1 %
Osteomyelitis of ribs[a]	<0.1 %
Less serious complications	
Pain/tenderness[a,b]	
Acute (<4weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Wound scarring	5–20 %
Deformity of rib/chest or skin (poor cosmesis)	1–5 %

[a]Note: Dependent on the extent and underlying disease/pathology, location of disease, and/or surgical preference which will alter the relative risks

[b]The neonate will usually be completely unaware being paralyzed, ventilated, and sedated; for repairs in later life, it can be a problem

Oscillation or ECMO may have to be used to retrieve this situation. ECMO as a primary treatment for CDH to get the neonate through the first few days or weeks of life has not achieved universal acceptance and is rarely used in Australia. ECMO as a retrieval from an air leak can be effective.

There is an emerging group of neonates that now only just survive, as we get gradually better at retrieving the hypoxic shunting baby. These babies are now

managed with permissive hypercapnea and apparently less than ideal hypoventilation (which drastically reduces lung damage and mimics the levels of tissue oxygenation that was present before birth). These babies have previously rarely seen problems.

Moderate to severe **tracheobronchomalacia,** where major airway collapse can occur from the soft walls of the airways, from the larynx to the major bronchi can occur. To take these patients off a ventilator, a **tracheostomy** may be required for months until the trachea and bronchi become more rigid and are able to support themselves. This will take months or up to a year. Another problem is **persisting pulmonary hypertension** that may go on for months. In the past this was an acute problem only, as the patient either recovered or died. Now, the patients are surviving through the acute phase (weeks) only for the problem to persist for months, if not a year or so.

Multisystem organ failure is extremely serious and is the usual mode of death in those that survive long enough to be ventilated. This is usually the result of the inability to oxygenate the patient or generalized sepsis. Cardiac anomalies will contribute to multi-organ failure, so that the ultimate cause of death in these patients is often a combination of problems leading to this scenario. Once established, there is a high mortality. **Mortality** is greater for infants with large herniae, those with a large volume of liver in the chest, cardiac disease, bilateral herniae, and in very preterm/low-birth-weight infants (<1,500 g). Those with chromosomal anomalies, e.g., trisomy 13 or 18, rarely survive birth and often die in utero. Even if they survive birth, they soon die, and treatment would not be a viable option.

For repeat surgery: In the neonate, **bowel perforation** is rare, but may occur in repeat surgery especially where a prosthetic patch has been used, has torn off its attachments as the baby grows, and then has to be dissected off the colon. Then the subsequent leakage and infection may be devastating. Pleural space infection (**empyema**) may follow leakage from the bowel, but is virtually unheard of in the first operation.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Nutritional deficiency
- Risk of other abdominal organ injury*
- Hernia recurrence
- Persistent pneumothorax
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Biliary Atresia and Choledochal Cyst Surgery: Biliary Bypass Drainage: Roux-en-Y Hepaticojejunostomy (The Kasai Procedure)

Description

The aim is to restore bile flow from the liver to the intestine by resecting, bypassing, or relieving the obstruction.

Biliary atresia occurs in about 1 in 10,000 live births (and may be higher in Asian populations), resulting in fibrous obliteration of the biliary tree (may occur early in life probably as a form of neonatal sclerosing cholangitis).

Choledochal cysts are at least fivefold (1:50,000) less common (but also relatively higher in Asians and females) and comprise a group of congenital dilatations of the bile ducts. The most common in the neonate is a "fusiform" dilation of the common bile duct and common hepatic duct up to the confluence of the left and right hepatic ducts. In the neonate, choledochal cysts may show some of the features of biliary atresia, and the two can coexist. For example, it is not uncommon to see a choledochal cyst in the CBD, accompanied with classical histological biliary atresia in the more distal CBD.

Long-term cholangiocarcinomas may develop in unresected choledochal cysts, a few before age 25. But, only 57 % of the cholangiocarcinomas occur in the area of the cyst. Therefore, nearly 50 % will occur in the residual biliary system after the cyst is removed and bypassed. This implies that the whole of the biliary tree is at risk. Surgery depends on the type of abnormality and extent of obstruction. For the more common choledochal cysts in childhood, the choice is limited to (i) a Roux-en-Y jejunostomy anastomosed to the common hepatic duct or (difficult in a neonate) to the right and left hepatic ducts after "fish-mouthing" the inferior borders of those ducts or (ii) a cholecocho-duodenostomy, where the common hepatic duct is joined to the upper border of the duodenum. The latter can be done quite effectively laparoscopically in the child. Close follow-up is usually required to detect late stenosis, which may produce multiple stones within the biliary tree above the stenosis; or in the case of choledochal cysts, cholangiocarcinoma of the remaining bile duct(s) can occur. Occasionally, resection of the short, affected portion of extrahepatic bile duct can be performed with primary end-end anastomosis.

In *biliary atresia*, the Kasai portoenterostomy joins the porta hepatis to a Roux-en-Y jejunal loop, after removing the scar tissue at the hepatic plate, leaving only the inner layers of the plate, so as to avoid entering the liver parenchyma. The dissection is carried out as far laterally as possible, and the lymphatics are preserved (not coagulated) as they may be the source of future bile drainage. In biliary atresia, one third of patients will never drain bile and go into liver failure within months of birth. Of the two thirds that drain bile, only half of them (one third or those operated on) will continue to drain bile beyond 5 years of age and may reach 20–30 years of age before they require transplant.

Anatomical Points

Knowledge of the common and uncommon biliary and vascular anatomical variants is essential for reducing mishap during biliary surgery. For choledochal cyst, the

cyst is dissected out by peeling it away from its surrounding tissues, bearing in mind the anatomical variants that can occur. For biliary atresia, scar tissue is present, so the fibrous band that represents the GB is dissected down to the "fibrous pyramid" that represents the CHD and the right and left hepatic ducts, and then the vessels are identified one by one around this pyramid, taking care to identify the small arteries in the neonate (and up to 3 months of age). Once that is done, the fibrous pyramid is removed down to the last inner layer of the hepatic plate. Correct identification of the common bile duct, rather than the common hepatic duct, should be confirmed to ensure that a choledochotomy is not performed in a bile duct that is too small (no luxury of choice may exist as the cyst may well go up into the CHD and even the confluence of the hepatic ducts). Even CT cholangiograms or MRCP will not usually clarify the anatomy particularly well in the very young. Preoperatively in the neonate, the choledochal cyst can occupy the whole of the abdomen, and the whole system is distorted. For the common form of choledochal cysts, the CBD, the GB and part of the CHD are involved. For intrahepatic cysts, the cyst would probably be left until the child is older, or if causing cholangitis, a liver resection (if practical with multiple cysts) may be preferable.

Perspective

See Table 3.7. Early major complications are related to failure to drain bile, bleeding, bile leakage, and infection, and later complications include biliary stenosis and bile duct malignancy (for cysts) and recurrent cholangitis and liver (cirrhosis) fibrosis (for those with biliary atresia (BA)). Minor complications are common and usually resolve without sequelae. Coexisting congenital disorders or anomalies, e.g., cardiac, especially left atrial isomerism, may predispose to higher risks of complications and failures. When these patterns of disease coexist with biliary atresia, they are known as syndromal forms of BA. When these syndromal forms occur, they have a far worse prognosis than the idiopathic form. Longer-term survival is dependent on disease progression, recurrent infections, cholangiocarcinoma (in choledochal cysts), and the inevitable ongoing fibrosis of the liver in biliary atresia. This leads eventually to liver failure in every patient with BA. *Liver transplantation* will typically be required for biliary atresia in the long term. So, all patients would die without transplant at some stage, 2/3 before 5 years of age. The best units internationally can obtain 40–45 % of patients out to 5 years without transplant, but if not transplanted, all eventually succumb to liver failure. Transplant can be carried out down to 5 Kg body weight and possibly less, depending on the patient, disease, and unit. Without transplant all of these patients will go into liver failure at some age and die.

For choledochal cysts, there is a high rate of stenosis and cholelithiasis usually occurring 10–20 years after the initial surgery. This may even require liver transplantation. For the Kasai procedure, death will be inevitable without liver transplant at varying times (however, the success for the Kasai procedure is only about 5 years or so of adequate drainage).

Table 3.7 Surgery for biliary atresia and choledochal cyst estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Death in biliary atresia as above	
Late stenosis, cholelithiasis, and liver failure in choledochal cysts	
Infection[a] overall	1–5 %
Subcutaneous/wound	1–5 %
Intra-abdominal/liver bed/pelvic	0.1–1 %
Liver(hepatitis; abscess)	0.1–1 %
Cholangitis	20–50 %
Systemic	0.1–1 %
Bleeding/hematoma formation	
Wound	1–5 %
Anastomotic; raw liver surface	1–5 %
Gastrointestinal (incl. variceal) hemorrhage	20–50 %
Liver failure long term (cirrhosis)[a]	20–50 %
Portal hypertension	20–50 %
Bile leak/collection	20–50 %
Biliary fistula[a]	5–20 %
Biliary ischemia/stenosis/obstruction/cholelithiasis[a]	20–50 %
Small bowel fistulae[a]	1–5 %
Later cholangiocarcinoma[a]	1–5 %
Mortality (10 year)[a]	20–50 %
Mortality without surgery[a]	>80 %
Rare significant/serious problems	
Injury to the bowel or blood vessels	0.1–1 %
Gastric/duodenal/small bowel/colonic	
Bile/hepatic duct injury	0.1–1 %
Liver injury	0.1–1 %
Biliary ascites	0.1–1 %
Failure to detect/remove calculi	0.1–1 %
Jejunal fistula	0.1–1 %
Small bowel ischemia	0.1–1 %
Multisystem organ failure (renal, pulmonary, cardiac failure)[a]	0.1–1 %
Small bowel obstruction (early or late)[a] [Anastomotic stenosis/ischemic stenosis/adhesion formation]	0.1–1 %
Operative cholangiogram	
Dye reaction/cholangitis/pancreatitis/radiation exposure	<0.1 %
Possibility of colostomy/ileostomy (very rare)[a]	0.1–1 %
Pancreatitis/pancreatic injury/pancreatic cyst/pancreatic fistula[a]	0.1–1 %
Aspiration pneumonitis	0.1–1 %
Wound dehiscence	0.1–1 %
Incisional hernia formation (delayed heavy lifting/straining)	0.1–1 %
T-tube-related complications (if used)	
T-tube cholangiogram	
Dye reaction/cholangitis/pancreatitis/radiation exposure	<0.1 %

(continued)

Table 3.7 (continued)

Complications, risks, and consequences	Estimated frequency
Blockage of T-tube	0.1–1 %
Dislodgment of T-tube	1–5 %
Persistent biliary fistula (after removal; cholangio-cutaneous)	0.1–1 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Seroma/lymphocele formation	0.1–1 %
Muscle weakness (atrophy due to denervation esp subcostal incision)	1–5 %
Paralytic ileus[a]	50–80 %
Nasogastric tube[a]	1–5 %
Blood transfusion	<0.1 %
Wound drain tube(s)[a]	1–5 %
Wound scarring (poor cosmesis/wound deformity)	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

Major Complications

For **biliary atresia**, one of the main complications is **failure of adequate biliary drainage** and **persistent jaundice**, with continued liver fibrosis/cirrhosis no matter whether they get drainage or not. Only the relative rates change. Fibrosis is faster in those that are not drained, so that synthetic function fails earlier, with subsequent **liver cirrhosis**, also due to progressive biliary fibrosis. Paradoxically, in those that drain well, there is a higher incidence of subsequent bacterial **cholangitis**, which can be very difficult to treat, accompanied by cessation of bile drainage, and may become chronic leading to permanent loss of drainage and **liver failure**. If there is no anatomical path for drainage (e.g., failed Kasai), then there is no path for bacteria, and cholangitis does not occur. **Bile leakage** may occur and can lead to **bile ascites** or **fistula** formation, surprisingly easily controlled and usually stops within a week or two. Venous **bleeding** can be catastrophic during the procedure and difficult to control and maintain flow. **Portal hypertension** with **variceal bleeding** often occurs after a Kasai procedure and often presents a significant risk to the patient. **Cirrhosis** with fibrosis of the liver and biliary system with excretory failure is a serious problem. **Bleeding** may be severe, arising from either arterial or venous injury (more likely in the adult patient as, in the child, the arterial vessels are very small). However, bleeding usually stops, but the liver parenchyma can be devascularized, the extent of which depends on the level of injury.

For **choledochal cyst**, **biliary stricture formation** may occur at any stage postoperatively and necessitate **further surgery**. Recurrent attacks of hyperamylasemia (raised lipase also) and **pancreatitis** are not infrequent after choledochal cyst surgery and are usually mild and can be managed nonsurgically in the majority of cases. Severe pancreatitis is relatively rare. **Cholangiocarcinoma** in the remaining choledochal cyst is not uncommon, often many years later.

Wound infection, **peritonitis,** or **abscess formation** may lead to **multisystem organ failure** and **death,** but the incidence of these complications is surprisingly small for the first operation. A second exploration, after sudden cessation of bile drainage, is now rarely used and multiple operations virtually never. With repeat operations, the complication rates increase, and the complications of subsequent liver transplantation are also increased, so that repeat surgery is employed less and less. These procedures carry a significant short- and longer-term risk of further complications and **mortality**.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Nutritional deficiency
- Risk of other abdominal organ injury*
- Recurrent biliary stricture
- Liver failure and transplantation
- Later malignancy
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Congenital Duodenal Obstruction

Description

General anesthesia is used. Congenital duodenal obstruction is due to either duodenal atresia (75 %) or stenosis (25 %) resulting in third-trimester polyhydramnios, duodenal dilatation, and early vomiting. Duodenal atresia or stenoses due to malrotation (with Ladd's bands and/or volvulus), annular pancreas, and duodenal web represent the main underlying causes.

Duodenal atresia is often diagnosed antenatally. The diagnosis usually occurs late in the pregnancy so that termination is no longer possible. Approximately 30 % of those diagnosed antenatally will have a chromosomal abnormality (nearly all trisomy 21), and many duodenal atresia patients will have other abnormalities (even if they do not have a chromosomal abnormality). As a result, prenatal counseling can be complex. Duodenal atresias can present surprisingly late after birth, often

after several days. The over-distended stomach empties itself immediately after birth, and the baby can then apparently tolerate small feeds for a considerable time while the stomach refills. Atresia is corrected by a duodenoduodenostomy, where the distal duodenum is rotated and flipped over the area of the obstruction to be joined to the proximal duodenum. This avoids dissecting into the area where the bile and pancreatic ducts enter the duodenum (nearly always close to the level of the obstruction).

Duodenal stenosis can vary, and the degree of obstruction determines the timing and severity of onset of symptoms, after birth. Occasionally, a patient with duodenal stenosis may not present for several years. The aim of surgery is either to open the obstructed area or to bypass it. A duodenal web is usually corrected by a longitudinal duodenotomy with incision or fulguration of the antimesenteric portion of the web (again to avoid damage to the bile ducts which can run either on surface of the web or usually closer to the mesenteric border). Once a web has been divided, the duodenum is closed transversely.

Anatomical Points

The anatomy of the duodenum is determined by the type of congenital defect present and whether malrotation is also a factor. The ampulla of Vater may be injured in the surgery, largely depending on the proximity to the site of obstruction and surgery, as the bile ducts open above or below the level of obstruction, or on the actual web when present. Concurrent cardiac anomalies may cause cardiac failure postoperatively and increase mortality. Other anomalies are present frequently in association with atresia or stenosis including Down's syndrome (trisomy 21).

Perspective

See Table 3.8. Overall mortality is often more directly related to severe associated anomalies (chromosomal and congenital) rather than the duodenal obstruction itself. Those with lethal chromosomal abnormalities (e.g., trisomy 18) may be so severe as to preclude surgery. The major surgical complication is a leak at the anastomosis. Gastric emptying will seldom be normal, and the stomach takes weeks before it is emptying adequately. This may necessitate a naso-enteric tube. Duodenal obstruction is often seen within days of birth. Intravenous feeding may be required if the stomach will not empty postoperatively. Many complications are of a minor nature, and babies recover rapidly from surgery. Major complications are related to infection, perforation, recurrence of duodenal obstruction, leakage, and later complications of small bowel obstruction. Major infections, multisystem failure, and mortality are higher in those with significant congenital anomalies, often cardiac.

Table 3.8 Surgery for congenital duodenal obstruction estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	
Subcutaneous/wound	1–5 %
Intra-abdominal	5–20 %
Mediastinitis	<0.1 %
Systemic (especially in those with chromosomal abnormalities +/– heart disease)	5–20 %
Bleeding and hematoma formation[a]	1–5 %
Pancreatic injury/leakage/damage to the bile ducts	5–20 %
Green bilious aspirates (universal for days or weeks after the surgery)	>80 %
Duodenal leak at the anastomosis	5–20 %
Recurrence/persistence of duodenal obstruction (some degree of duodenal dysmotility will persist for life, so that GOR is very common)	1–5 %
Reflux esophagitis (GOR)	20–50 %
Multisystem failure (renal, pulmonary, cardiac failure) – overall (especially high in those with chromosomal abnormalities +/– heart disease)[a]	1–5 %
Small bowel obstruction (early or late)[a] [Anastomotic stenosis/adhesion formation]	1–5 %
Mortality – overall (depends on severity of other anomalies, rather than the duodenal obstruction itself)	1–5 %
Mortality without surgery (may be up to 100 %) (lethal chromosomal abnormalities (e.g., trisomy 18) may preclude surgery)[a]	>80 %
Rare significant/serious problems	
Liver injury	0.1–1 %
Subphrenic abscess	<0.1 %
Complete anastomotic (duodenotomy) breakdown	0.1–1 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Intolerance of large meals (necessity for small frequent meals)	20–50 %
Delayed gastric emptying (Universal for up to several weeks and probably universal to some extent for life)	>80 %
Wound scarring (poor cosmesis/wound deformity)	1–5 %
Nasogastric tube[a] (sometimes for weeks)	50–80 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
Important Note: Many of these complications are individually closely determined by the exact nature of the problem and surgery

Major Complications

Anastomotic leakage is serious and occasionally not recognized early, leading to **intra-abdominal infection** and **abscess** formation. **Systemic infection and**

multisystem failure may then ensue. Concurrent cardiac anomalies may cause cardiac failure and increase mortality. **Bleeding** is rare and is usually controlled at surgery. **Wound infection** may occur and may lead to an incisional hernia. Wound **dehiscence** is rare. **Persistent duodenal obstruction** can result from inadequate division of a duodenal web or from anastomotic stenosis or functionally from severe dysmotility. **Severe gastroesophageal reflux** can be prolonged and may require fundoplication. **Small bowel obstruction** may occur from postoperative adhesions, even many years later. **Intravenous feeding** may be required if the stomach will not empty postoperatively.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Reflux problems
- Feeding problems
- Duodenal leakage
- Risk of other abdominal organ injury*
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Midgut Obstructions and Inflammation: Malrotation, Volvulus, Jejunoileal Obstruction, and Meconium Ileus

Description

In any neonate with an intestinal obstruction, the objective of the operation is to define the problem and alleviate the obstruction. Resection of bowel may be required depending on the pathology and the degree of obstruction, ischemia, or necrosis present. For children, and especially neonates, a transverse muscle-cutting incision is typically used. There are few anatomical variations that affect the small bowel except for malrotation with Ladd's bands and a Meckel's diverticulum and other derivatives of the vitello intestinal duct. Malrotation is where the midgut loop fails to return to the abdominal cavity, with its normal rotation, between 10 and 12 weeks' gestation. A Meckel's diverticulum is a remnant of the vitellointestinal duct (the duct that runs from the yolk sac through the umbilicus to the apex of the midgut

loop – where a Meckel's is situated), which can give rise to other problems as well as the well-known Meckel's diverticulum (see section "Surgery for Meckel's Diverticulum and Vitellointestinal Remnants"). In addition, small intestine atresias and meconium ileus are causes of small bowel obstruction in the newborn.

Malrotation is any departure from the usual final adult positioning of the gut within the abdomen. If a patient presents with an intestinal obstruction and hypovolemic shock as a newborn, the degree of malrotation is usually complete, with the second, third, and fourth parts of the duodenum running vertically down just to the right side of the midline, with the cecum and ascending colon running up just next to it, but to the left of the midline. These, together with all of the intervening small bowel, make up the midgut loop of the fetus. Where there is a complete malrotation, the midgut loop is closely applied to a central thick cordlike "universal mesentery" that hangs free in the abdomen. This contains the blood supply to all of the midgut loop, with the proximal small bowel vessels going to the right and the distal small bowel and large bowel vessels going to the left. As a result, this pendulum-like arrangement is very liable to twist on itself at the upper point of attachment, as the midgut fills with food for the first time. The baby presents with pain (screaming), hypovolemic shock, green bile vomiting (with or without blood), and the passage of blood rectally. Venous gangrene of the whole of the midgut loop – middle of the first part of the duodenum to just short of the splenic flexure of the colon – may occur. In the older child (or even adult), the degree of malrotation is often not as marked causing intermittent attacks of obstruction, with central abdominal pain and green bile vomiting. There is often a history of several such attacks, which appear to be able to resolve, presumably by the midgut untwisting, until the diagnosis is eventually made. Nevertheless, surgery is often as an emergency, because of a midgut volvulus. This can occur at any age, and patients have presented well into their 40s and older. The treatment is essentially the same as in babies (see Ladd's procedure below).

Anatomical Points

Malrotation

There are few anatomical variations that affect the small bowel except for Meckel's diverticulum and malrotation with Ladd's bands. A Meckel's diverticulum is a remnant of the vitellointestinal duct, which can give rise to other lesions as well as the well-known Meckel's diverticulum.

Malrotation can vary from complete failure to rotate to minor forms of maldescent of the cecum. In the fetus, the midgut loop is attached to the back wall of the abdomen at the middle of the second part of the duodenum and again just short of the splenic flexure. Between these two points, it is outside the abdomen in the umbilical cord. In the normal process of the midgut returning to the abdomen during the 10–12th week of gestation, the cecum and transverse colon rotate anterior to (over) the base of the small bowel mesentery with the cecum moving down to the RIF, as the duodenum rotates deep to it up into the LUQ. In that way, the root of the mesentery is attached diagonally across the whole of the abdomen on the longest

Table 3.9 Surgery for midgut obstructions and inflammation estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	5–20 %
Subcutaneous	5–20 %
Intra-abdominal/pelvic (peritonitis; abscess) (especially in the ELBW; premature)	5–20 %
Systemic sepsis (especially in the ELBW; premature)	2–5 %
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Small bowel obstruction (postoperative early or late)[a] [Adhesion formation]	1–5 %
Repeated further surgery[a]	1–5 %
Rare significant/serious problems	
Perforation (spontaneous preoperative)[a]	0.1–1 %
Anastomotic leakage[a]	0.1–1 %
Fecal/enterocutaneous fistula[a] (very rare)	<0.1 %
Ureteric injury (very rare)[a]	<0.1 %
Multisystem organ failure (renal, pulmonary, cardiac failure)	0.1–1 %
Mortality[a]	0.1–1 %
Mortality without surgery (should surgery be refused)[a]	>80 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Nerve parasthesia	0.1–1 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %
Nasogastric tube[a]	1–5 %
Wound drain tube(s)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
Note: ELBW extremely low-birth-weight newborn, <1,000 g
Important Note: Many of these complications are individually closely determined by the exact nature of the problem and surgery. The extent and underlying disease will alter the relative risks

attachment possible. If, as the intestine returns to the abdomen, it fails to rotate, then it simply hangs in the abdomen attached as it was in the fetus. Ladd's bands represent the course the colon should have taken and therefore stretch across the duodenum.

Mirror image malrotation, although thought to be rare, is now more frequently detected antenatally in association with left atrial isomerism, with situs inversus abdominus. The gut is the mirror image of normal, but is also frequently malrotated. A barium meal after birth confirms the situs inversus with the malrotation, and the patient then undergoes a mirror image Ladd's procedure – often laparoscopically.

Malrotation: Complications of the Disease

Volvulus, usually of the entire midgut, occurs clockwise when viewed from in front. The vascular supply will be compromised to a varying degree and may cause venous then arterial infarction. In addition, the malrotation may be associated with duodenal obstruction from congenital bands running from the ascending colon across the front of the duodenum (Ladd's bands). Volvulus occurs in the baby, while partial obstructions or volvulus can occur in the older child. If left too long, the patient will die of hypovolemic shock and/or venous gangrene of the whole of the midgut loop – middle of the second part of the duodenum to just short of the splenic flexure of the colon. The most significant complication is ischemic infarction of the midgut loop with subsequent death or years of parenteral nutrition followed by a gut transplant. With an established volvulus, death is inevitable without surgery.

In the premature it is very difficult to differentiate between necrotizing enterocolitis (see 46.10) and a midgut volvulus. Both have the same presentation, viz., hypovolemic shock and bowel obstruction. But, NEC is treated expectantly, initially, which would be disastrous where there is volvulus of the midgut loop. An ultrasound of the upper abdominal vasculature and the first few loops of bowel may well differentiate, whereas a plain X-ray may not. But, if there is any doubt, an emergency laparotomy should be performed without delay.

Malrotation: The Surgery and Its Complications

Ladd's Procedure: The surgery for malrotation with or without volvulus is done as soon as possible to try to minimize gut loss. As for all obstructions to the small bowel in the neonate, an upper transverse muscle-cutting approach is used. Any volvulus is untwisted anticlockwise as you look at it. Ladd's bands (fibrous bands running from the ascending colon across the duodenum) are divided, allowing the colon to be moved further to the left, exposing the root of the mesentery and the duodenum. The thick rope-like "universal mesentery" is splayed/teased open, so that the vessels to the duodenum and jejunum can be moved to the right, and the vessels to the terminal ileum, ascending colon, and transverse colon can be moved to the left. In this way the corresponding intestine can be moved with the vessels as far away from the midline as possible. Normal anatomy cannot be established as the ileocecal vessels that normally run down to the RIF now run a very short distance to the ascending colon, which is in the high midline. The most mobile parts of the rest of the small intestine are then placed back over the raw area created by splitting the mesentery, so that they will form adhesions to that area, hopefully preventing further twisting. The cecum is placed in the LIF and an appendectomy is usually carried out.

Laparoscopic Ladd's Procedure: The procedure can now be performed laparoscopically, even in the presence of volvulus, and is being performed more and more commonly with time. Obtaining an adequate window to obtain an adequate view is often difficult when the gut is distended, so conversion to an open procedure is not uncommon when volvulus has occurred.

Complications of the Disease: If there is gangrenous bowel, then it has to be resected, hopefully, without losing the entire midgut loop, as this will be essentially lethal, without prolonged TPN, +/− SB transplantation. If surgery is delayed, then the patient may be irretrievable.

Complications of the Surgery: Short-term problems are survival of the child and the intestine and then leaking anastomoses. The dissection in the mesentery often injures lacteals, which will produce a temporary chylous ascites. A short-to medium-chain fat dietary substitute or parenteral nutrition for 4–6 weeks may be useful if prolonged ileus occurs.

Longer term, the deliberate placing of the mobile part of the small bowel into the raw dissected mesentery will lead to extensive adhesions. While this will diminish the tendency to a recurrence of any volvulus, it does increase the incidence of adhesive obstruction, so that there is a 20–30 % lifetime risk of an adhesive bowel obstruction (Murphy et al. 2006).

Jejunoileal Obstruction in the Newborn

Jejunoileal obstruction in the newborn is predominantly associated with an atresia probably caused by a complete interruption to the blood flow to the intestine in the fetus. This leads to loss of the intestine. Because the fetal bowel contains no organisms, the dead bowel disappears, and the two blind ends seal themselves. Intestinal atresias take various forms, from loss of the lumen only to a significant gap, or multiple atresias with small remaining segments – "a string of sausages" (Federici et al. 2003). Incomplete obstruction associated with stenosis is rare (but does occur in the duodenum). Jejunoileal obstructions are rarely associated with other congenital anomalies. Unless the atresia is associated with a significant or lethal loss of bowel, the long-term outlook is usually good. In those with a very proximal jejunal atresia, the degree of dilatation of the proximal bowel may be grotesque so that disparity between it and the distal bowel is a major issue.

Complications of the Disease

Jejunal Atresia

Proximal to a jejunal atresia, the obstructed intestine undergoes massive dilation and becomes inert (thought to be due to ischemic changes in the antimesenteric wall plus the back-pressure effect). These dramatic changes are easily detected on antenatal ultrasound. At birth, an X-ray will show relatively few grossly distended loops of bowel. As with all small bowel obstructions in the newborn, if left untreated, the proximal intestine will progressively distend and will eventually perforate or infarct with subsequent peritonitis and death.

The Surgery: In all small bowel atresias, the proximal intestine becomes distended, and the distal intestine becomes shrunken and unused. The more proximal the atresia, the more marked the upstream distension, giving rise to a gross disparity

in size especially in proximal jejunal atresias, so that anastomoses are difficult. Where possible the most distended part of the intestine (the distal end of the proximal part) is resected, thereby presenting a less distended part to be anastomosed. Jejunal atresias are frequently close to the DJ flexure, limiting the surgeon's ability to resect all of the distended bowel. Nevertheless, the most grossly dilated bowel is resected, where possible. The rest of the distended proximal intestine is tailored to a narrower tube (by discarding the floppy antimesenteric part) and is anastomosed with its end anastomosed to a longitudinal incision made into the antimesenteric border of the distal (contracted and unused) small bowel – the so-called end-to-back anastomosis used in all anastomosis of the small and large intestine where there has been a congenital small or large bowel obstruction. The end-to-back anastomosis allows a larger than normal proximal intestine to be anastomosed to a smaller than normal distal intestine. As for all intestinal anastomoses in the newborn, a single-layer interrupted absorbable suturing technique is usually used. In North America, however, stapling is often used especially for the tailoring procedure in jejunal atresia. Where there are multiple atresias, resection and multiple anastomoses may be required. Occasionally, a temporary proximal stoma may be used to protect multiple distal anastomoses while they seal and heal.

Complications of the Surgery: There are two common postoperative issues. The first is that the long tailoring suture line may leak, and/or stricture, and secondly the proximal intestine will take a long time to recover its motility after the prenatal ischemic damage (that caused the atresia in the first place) and the long-tailored anastomosis. Therefore, these babies often require IV nutrition for weeks.

Ileal Atresia

This is less common, but has its own unique problems. In one specific circumstance where there is a distal ileal atresia, the blood supply to the distal ileum is derived from an ascending branch of the ileocolic artery running back up the small bowel from the ileocecal junction. So, if there is a gap in the mesentery anywhere in the distal ileum supplied by this vessel, then the intestine is free to twist around the artery, and if it does, it creates the appearance of an "apple-peel atresia." As with jejunal atresia, the proximal intestine will dilate, but not as markedly as for jejunal atresias.

The Surgery: Uncomplicated atresias can occur anywhere in between these two sites (the grossly distended proximal jejunal atresia and the distal ileum's apple peel). Then, the surgery is far more straightforward; the most distended part of the proximal intestine is resected and an end-to-end anastomosis carried out. Recovery times are far shorter, but still take a few days. The apple-peel atresia's tenuous blood supply makes it difficult to straighten out the intestine and reanastomose the ends, as the part that has been spiraled around the artery may lose its blood supply as it is straightened. Therefore, partial resection of this intestine is frequently necessary to ensure a good blood supply to both ends.

Long-term complications for jejunal and ileal atresias are few. Adhesive obstruction occurs in approximately 5 % and usually within 2 years of the

surgery. Where there has been an abdominal wall defect, the adhesive obstruction rate is higher (van Eijck et al. 2008). Rarely the loss of intestine is critical. If it is, then long-term parenteral nutrition is required, while enteral feeding is gradually established. Hopefully, the residual gut gradually adapts to enable full enteral feeding. In the newborn, the gut is still growing and appears to be able to compensate for some of the loss of length, as well as being able to undergo mucosal adaptation.

If, however, the residual gut is not long enough to support life, then there are gut lengthening procedures (where the gut is split longitudinally and then reanastomosed end-to-end) or interposition procedures (that slow the transit time) that have been used. They have met with mixed success. The "multiple step" procedure is now gaining popularity (proceedings of the World Congress of Surgery, Adelaide, 2008). This is a procedure where there are multiple incisions made into the side of the gut for just over half of its circumference, and then those incisions are closed longitudinally. The gut is then lengthened rather like a paper chain. This procedure appears to increase transit time, and as the residual narrowed gut dilates, it increases the absorptive surface area as well. Initial results would suggest that this is more successful than the older procedures.

If the terminal ileum has to be resected, then vitamin B_{12} absorption will be poor. If the resection is carried out in the neonate, however, then passive absorption appears to compensate (Ooi et al. 1992) to a far greater extent than in the adult, so that serum levels of B_{12} are usually normal. The Schilling test will be abnormal. If the serum levels are normal, supplements are not necessary in childhood, but levels should be checked. When such an affected female becomes pregnant, however, the fetus requires folate and B_{12} at levels that cannot then be supplied by a normal diet for this mother, so that females who have lost the terminal ileum need to be aware of the extra requirement during pregnancy.

Meconium ileus

Meconium ileus is a neonatal intestinal obstruction that occurs in 14 % of newborns with cystic fibrosis (Efrati et al. 2010) (but obstruction may occur in older cystic fibrosis patients when it is known as "meconium ileus equivalent"). It is due to viscid intestinal mucus causing obstruction of the small bowel in the distal ileum, with proximal intestinal dilatation. On antenatal ultrasound, this dilatation can be detected, and it is often accompanied by echogenic bowel at that time.

Some 10 % of cystic fibrosis patients will present at birth with this form of bowel obstruction, approximately 1:20,000 live births (Rescorla et al. 1998). The obstruction in meconium ileus may be a simple intraluminal small bowel obstruction (SBO) from inspissation of mucus, which becomes a concrete-like series of plugs in the ileum. Or, the back-pressure from this obstruction may cause proximal segmental volvulus with loss of intestine in the fetus. Or, the obstruction may cause a proximal perforation and leak of meconium before birth. In the latter, the baby is born with meconium peritonitis, where the abdominal viscera are fixed in highly inflamed adhesions from the sterile peritonitis (Rescorla et al. 1998).

Conservative Treatment

With meconium ileus, as above, ischemia, perforation, sepsis, and respiratory difficulties from abdominal distension may supervene at birth. If there is an uncomplicated meconium obstruction, repeated contrast enemas with gastrografin (which contains a detergent) may clear the obstruction, but laparotomy is often required because the obstruction may not clear fast enough to alleviate abdominal distension and respiratory distress.

Surgical Correction

At laparotomy for the intestinal obstruction, resection of the most distended part of the intestine is performed if there is a simple distal luminal obstruction. The residual distal ileum is then flushed through with normal saline until the colon is seen to clear. If this is ineffective, then a temporary stoma of the type that allows the distal intestine to be flushed is used. If there has been a proximal volvulus, then the ischemic gut is resected if it is still present. If it has already disappeared, the resultant atresia is dealt with as above. If cystic fibrosis has been complicated by meconium peritonitis before birth, the adhesions are usually dense and inflamed. Then a temporary stoma is often used. A follow-up laparotomy is carried out 4–6 weeks later when the inflammation has subsided. The areas of adhesive obstruction can be corrected and the stoma closed. Another cause of segmental volvulus can occur around a persistent vitellointestinal band connecting the umbilicus with the small bowel at the site of a Meckel's diverticulum (see section 46.12).

Perspective

See Table 3.9. The complications of operations for small bowel obstruction depend on the initial pathology for which the procedure was performed. Death from hypovolemic shock and/or midgut necrosis and gangrene is well recorded if the surgery for midgut volvulus is delayed. In the very premature that have a volvulus, the diagnosis can be missed as these babies are thought to have necrotizing enterocolitis (NEC), which is much more common and presents with a similar initial picture. The two diseases may be combined, as the midgut loop develops gangrene.

A serious complication of any gut resection is an anastomotic leakage, the risk of which is increased by more distal obstruction of any cause or perforation with peritonitis. So, it is vital to alleviate any significant obstruction distal to any anastomosis. Therefore, in any baby with an atresia, it is mandatory to follow the distal bowel to the pelvic floor, preferably flushing it with warm saline at the same time to ensure its patency, as there may well be multiple atresias. The consequence of an anastomotic leakage is contamination of the peritoneal cavity leading to generalized peritonitis or intra-abdominal abscess formation, anywhere and everywhere in the neonate. Surprisingly, however, the neonate tolerates this well, and if a drain is placed through the wound after a leak is suspected, then the leak will often

spontaneously close as peristalsis distal to the anastomosis becomes established. The risks of an anastomotic leakage are reduced by ensuring good blood supply to the bowel ends, minimal tension, and no factors contraindicating an anastomosis.

The risks of wound infection, small bowel obstruction, enterocutaneous fistula and short bowel syndrome are significant, but fortunately uncommon complications, and are seen more often after necrotizing enterocolitis (see section 46.10). After a Ladd's procedure, the lifetime risk of an adhesive SBO may be as high as 20–30 % (Murphy et al. 2006). After a tailored jejunal atresia correction, SBO may be as high as 20 %, with reoperation in the first few weeks of life. The risk of short-gut syndrome depends on the type of lesion and the amount of small bowel resected or lost before birth, as does the risk of nutritional deficiency.

Ileus is common, but usually short lived (except in jejunal and duodenal atresias), being dependent on the amount of gut manipulation and inflammation. Incisional herniae are usually minor and often close themselves when surgery is performed on the newborn. Older age groups carry a higher risk of non-closure.

Major Complications

The type of complication is dependent on the surgery required and the extent of the disease entity present. Detorsion of a volvulus may be associated with **reperfusion-type injury** with release of ischemic products with **shock, hypotension,** and **sepsis**. **Perforation of the bowel** is associated with **intraperitoneal sepsis**, including **abscess formation** and **wound infection.** Where used, **anastomotic leakage** may occur. **Enterocutaneous or internal fistula** may arise. Adhesion formation and **small bowel obstruction** may occur which can be recurrent. **Systemic sepsis** may occur and may lead to **multisystem organ failure** and **death.**

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Feeding problems
- Risk of other abdominal organ injury*
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*
- ***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Necrotizing Enterocolitis

Necrotizing enterocolitis (NEC) is a life-threatening condition of uncertain etiology, which mainly affects the very premature baby (Albanese et al. 1998). It is associated with ischemic damage to any part of the bowel, from the DJ flexure to the rectum. The most common sites of disease are the terminal ileum and the sigmoid colon. It acts as a gas gangrene of the bowel wall with partial- or full-thickness necrosis of the intestine for short segments, or all of the bowel, or anything in between. Gas collects in between the layers of the intestinal wall and shows up on X-ray as pneumatosis before perforation. When there has been full-thickness necrosis, perforation will be demonstrated by a pneumoperitoneum and further sepsis.

The initial treatment of NEC is broad-spectrum antibiotics, IV feeding, and gut rest. Once the bowel is necrotic, subsequent bacterial leakage, infection, and perforation will occur, possibly causing death. The disease can be remarkably rapid in its progress. The indications for surgery are obvious perforation, increasing difficulty in ventilation because the abdomen is so tense, increasing systemic sepsis, thrombocytopenia, and inability to control the unstable patient. But, before any of these occur, attempts are made to treat the patient with broad-spectrum antibiotics, IV feeding, and gut rest.

Anatomical Points

The extent of gut affected largely determines the amount and type of resection, but based on the relatively constant anatomical blood supply. However, in situations where malrotation or other congenital anomalies, such as situs inversus, exist, the blood supply and anatomy can be considerably distorted.

Complications of the Disease

NEC affects approximately 5 % of the premature, which in turn accounts for 1 % of newborns. Of those >1,000 g birth weight, conservative management with IV antibiotics and IV feeding leads to a 70 % recovery without surgery. For those <1,000 g, the extremely low birth weight (ELBW), the disease is remarkably rapid and affects large segments, if not all of the small bowel.

In the ELBW neonates, the occurrence of generalized peritonitis occurs early so that the patient is often not fit for major surgery. In that scenario, placement of peritoneal drains will buy time in which to try to stabilize the baby before a laparotomy. Occasionally, no further surgery is necessary as the perforation(s) close themselves. Nevertheless, late surgery for fistula, stricture, and obstruction is the rule.

Many of the complications listed under complications of the surgery are really complications of the disease, e.g., strictures, malabsorption, and recurrent obstruction.

Table 3.10 Surgery for necrotizing enterocolitis estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	5–20 %
Subcutaneous	5–20 %
Intra-abdominal/pelvic (peritonitis; abscess) (especially in the ELBW; premature)	5–20 %
Systemic sepsis (especially in the ELBW; premature)	2–5 %
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Small bowel obstruction (postoperative early or late; stricture formation)[a] [Adhesion formation]	1–5 %
Repeated further surgery[a]	1–5 %
Intellectual impairment[a] (especially ELBW)	50–80 %
Rare significant/serious problems	
Perforation (spontaneous preoperative)[a]	0.1–1 %
Anastomotic leakage[a]	0.1–1 %
Temporary stoma(s)[a]	0.1–1 %
Fecal/enterocutaneous fistula(e)[a] (very rare)	<0.1 %
Ureteric injury (very rare)[a]	<0.1 %
Liver failure from prolonged cholestasis[a]	0.1–1 %
Nutritional deficiency B_{12} malabsorption[a]	0.1–1 %
Wound breakdown[a]	0.1–1 %
Multisystem organ failure (renal, pulmonary, cardiac failure)	0.1–1 %
Mortality[a] (especially ELBW)	50–80 %
Mortality[a] (term infants)	5–20 %
Mortality <u>without</u> surgery (should surgery be refused)[a]	>80 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Nerve parasthesia	0.1–1 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %
Nasogastric tube[a]	1–5 %
Wound drain tube(s)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
Note: ELBW extremely low-birth-weight newborn, <1,000 g
Important Note: Many of these complications are individually closely determined by the exact nature of the problem and surgery. The extent and underlying disease will alter the relative risks

The Surgery

Open Laparotomy for Perforation, Sepsis, and Ischemic Gangrene

Laparotomy and gut resection are reserved for where there is failure of conservative treatment or for obvious perforation with sepsis, for prolonged and frank bowel

obstruction, or for where the patient is deteriorating but could still tolerate a laparotomy. In these circumstances, there is nearly always more disease than expected, often with obvious segments of gangrenous intestine with or without multiple perforations.

For Those Unfit for Laparotomy

In the ELBW neonate, the disease often progresses so rapidly that there is systemic collapse. In that situation there is still controversy as to whether or not an immediate laparotomy should be carried out or drains should be inserted in the ICU with analgesia. At the moment, the consensus is that drains should be used to gain time followed by a laparotomy when the neonate is fitter. Others would stop at the drainage alone and wait and see if the intestine will seal, and gut continuity will be restored. The literature claims that this may occur in up to 30 % of patients treated in this manner, but local experience has met with virtually no success.

Spontaneous Perforation of the Small Bowel

There is a group of growth restricted (too small for gestational age at birth) neonates that develop a so-called spontaneous perforation of the terminal ileum (Messina et al. 2009). This appears to be the only affected area of the gut. Often there is a distal microcolon where the colon is small, shut down, and dysmotile. This appears to cause a localized back-pressure perforation in the last part of the small bowel and small areas of muscle loss in the intestine elsewhere. Clinically, the abdomen is soft, but distended, abdominal X-ray demonstrates a huge pneumoperitoneum, and initially the neonate is stable with no systemic effects. This perforation occurs in the first few days of life, whereas NEC usually only occurs several days if not weeks after birth. This local perforation is treated very differently to those with gross and frank widespread NEC. This localized lesion can be treated through a minilaparotomy in the RIF, creating a small local stoma to allow the small bowel to work and to allow the neonate to be fed enterally while waiting for the colon to start to work.

Complications of the Surgery

Neonates >1,000 g tolerate laparotomies and major gut resections well, even in the presence of gross intra-abdominal sepsis. For neonates >1,000 g who undergo surgery (30 % of those that develop the disease), there is a 5–10 % perioperative mortality, with the potential for intellectual morbidity in those that survive.

But, in those <1,000 g (the extremely low-birth-weight group [ELBW], usually 24–26 weeks' gestation), there is no hard figure for how many will get NEC and how many will survive without surgery (on conservative Rx, e.g., drainage only – see above), but for those that go to surgery, approximately 60 % will die soon after the surgery because there is so much dead bowel that nothing can be done, and the abdomen is just closed again. For those that survive surgery, 70 % will have moderate to severe intellectual handicap(s) (Chacko et al. 1999). The brain damage is thought to arise from gross pre- and postoperative vascular instability with large

shifts in blood pressure, especially in the central and intracranial venous systems, accompanied by large shifts in blood gases. After counseling before surgery, parents nearly always insist on the laparotomy despite this poor prognosis, which may leave them with a severely handicapped child.

Intra- and Postoperative Hemorrhage

In the ELBW group, there is little supporting stroma for blood vessels to the intestine and especially blood vessels in the liver. The liver is also very soft and is disproportionately large, often well down in the RIF, overlying the terminal ileum, one of the most frequently diseased areas in NEC. Therefore, the liver can be easily lacerated, even by simple retraction, and will not stop bleeding. Therefore, in the ELBW patient who requires a laparotomy, avoid entering the abdomen high on the right.

Liver Failure from Prolonged Cholestasis

Liver failure from prolonged cholestasis is a major problem after major gut resection in premature neonates. Contributing factors are prematurity, multiple abdominal procedures, a gut that will not tolerate enteral feeding because of prolonged ileus, prolonged parenteral nutrition, and intra-abdominal sepsis with or without systemic sepsis.

Nutritional Deficiency B_{12} Malabsorption

Although the terminal ileum is frequently removed, the incidence of a low serum B_{12} is actually low. The Schilling test is abnormal. So, it appears that if the terminal ileum is removed as a premature baby, then passive absorption takes over to a degree that maintains serum levels (Ooi et al. 1992). But, be careful to warn the mother and the female child who has lost the terminal ileum (when older) that although the patient's B_{12} and folate may be normal, absorption is such that it will decompensate during a pregnancy. Therefore, the fetus of a mother who has lost her terminal ileum as a baby is at risk of intrauterine brain damage, unless aggressive supplementation is carried out in that mother.

Wound Breakdown

Wound breakdown can occur especially in the very premature. Because of the poor resistance to infection in this age group and the major difficulties in maintaining adequate nutrition (where there is intra-abdominal sepsis), wound infection is very common. Superficial breakdown of the wound is also very common. Surprisingly wound dehiscence is rare, and the wounds gradually close.

Stricture (Early and Late) and Obstruction

Strictures occur because of the disease itself. They are not apparent at the emergency laparotomy as the process is one of gangrene at that time. These diseased

areas that are left behind then scar and stricture down. Presentation is bimodal: there is an acute presentation due to inflammation, necrosis, and swelling causing an obstruction, which may settle down with conservative treatment, but often goes on to a true stricture within days or weeks of the laparotomy, and/or there is a later presentation (up to years afterward but usually within a year) where a mature fibrous stricture forms. This will not relent.

Fistula(e)

Fistulae are seen often after surgery. They are more common after primary anastomoses. Surprisingly most settle on conservative management: nil by mouth and IV feeding. This does not need a laparotomy straightaway and does not mandate that drains should be placed at the time of the initial surgery, as this appears to increase the fistula rate.

Perspective

See Table 3.10. Death and brain damage are likely outcomes in the extremely low-birth-weight (ELBW) group. Anastomotic leak and anastomotic stricture are common (diseased ends to the bowel anastomosed, when every effort is being made to preserve bowel length). These problems are difficult to avoid, as the whole of the gut has poor perfusion, which is the presumed trigger for the disease. Temporary stomas are therefore frequently used to avoid an anastomosis in a patient who is in multi-organ failure, who has a tense abdomen and has to be ventilated at high pressures. Possibly simple drainage should be used, but that decision is made on a case-by-case basis. Short bowel syndrome may be a long-term issue. The extent of preoperative obstruction, ischemia, or perforation and contamination is a significant factor in the severity of postoperative complications and consequences. NEC is associated with relatively more severe complications overall, including nonsurgically related delayed neural development, as the disease occurs predominantly in the premature, which itself leads to a higher risk of intellectual impairment. The presence of other anomalies, and chronic lung disease, associated with prematurity may greatly influence the incidence of multisystem organ failure and mortality.

Major Complications

Anastomotic leakage, **spontaneous perforation**, **abscess formation,** and **abdominal sepsis** may lead to **systemic sepsis**, **multisystem organ failure,** and **death**. **Repeated further surgery**, with or without **small bowel obstruction**, may occur. **Mental and intellectual disabilities**, especially in the very low-birth-weight groups, are serious longer-term problems that may develop. **Bleeding** intra- and postoperatively can occur. **Liver failure** may supervene. **Nutritional disturbances**, including malnutrition, may result if large lengths of bowel are resected.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Feeding problems
- Nutritional deficiency
- Risk of other abdominal organ injury*
- Later liver problems
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Hirschsprung's Disease

Description

The failure of complete migration of neural crest cells to form the submucous and myenteric plexuses at the distal end of the bowel leads to the aganglionosis of Hirschsprung's disease. As there is failure of these cells to reach the end of the hindgut, Hirschsprung's disease is the absence of ganglion cells from just above the dentate line back up the intestine for a variable distance, to the point where the neural crest cell migration ceased. In 70 % of patients, the aganglionosis will involve part of the rectum, the whole of the rectum, or the rectum and sigmoid; the remaining 30 % will have longer segments. The disease occurs in approximately 1 in 5,000 live births.

The final diagnosis is a tissue diagnosis, not an imaging diagnosis, so biopsies have to be taken. Bowel obstruction may be significant, and perforation can occur, especially if enterocolitis supervenes. The aim of surgery is to resect the aganglionic segment of colon and anastomose ganglionic bowel to the anus, just above the dentate line. Occasionally, the resected segment may involve the entire colon and very rarely may involve large parts of the small bowel to the extent that survival is not possible. The disease is more common in trisomy 21 (Ieiri et al. 2009) and other rarer chromosomal abnormalities, e.g., a 13q deletion (Khong et al. 1994); there are familial cases and known gene defects strongly suggesting that there is a hereditary predisposition to the disease.

Surgery

The surgery for Hirschsprung's disease has traditionally been a staged procedure with a temporary colostomy, later the definitive procedure, and then closure of the

colostomy. But, total correction at birth as a one-stage procedure is becoming more popular. Many would do the definitive procedure as a combined laparotomy and transanal approach, but a purely transanal neonatal approach without a colostomy or laparotomy is now also gaining favor. When a laparotomy is combined with a perineal procedure, there are three ways of doing the operation: the "Soave," the "Duhamel," or the "Swenson." All have their proponents, the basic aim remains: to remove the aganglionic section and partly destroy the internal sphincter action and bring ganglionic bowel down to just above the dentate line.

Emergency Colostomy

Where the neonate is too ill to undergo a definitive procedure (e.g., where there is a life-threatening Hirschsprung's colitis), an initial colostomy is used but must be in ganglionic bowel (monitored by frozen section).

Routine Colostomy

With a staged approach to the disease, plain X-ray and contrast studies may help to determine where the transition from aganglionic to ganglionic bowel occurs. Often, these are not accurate. If a colostomy is to be used as a preliminary to an interval definitive procedure, then frozen section biopsies are taken to ensure that the colon is placed in ganglionic bowel. Seromuscular biopsies are taken from the peritoneal reflection (to prove the diagnosis) and then another at the apparent transition zone, where there is a change in caliber from dilated proximal to narrowed distal intestine. If the transition zone is confirmed, then a separated double-ended stoma, 5–10 cm proximal to that transition zone, is formed, as that transition zone will not be fully ganglionic. This will usually be in the LIF (as 70 % of patients will have disease up to the rectosigmoid) so the surgery is initiated through a muscle-splitting incision. If there are no ganglion cells, then more and more proximal biopsies are needed. Extending the transverse incision across the abdomen will suffice, as the whole of the abdomen can be reached from a low transverse incision in the neonate. It is advisable to avoid a colostomy in the transverse colon as they frequently prolapse, sometimes within months.

Definitive Staged Procedure

Again the choice is operator dependent, using one of the three forms of pull-through procedure. The "Swenson" is a low anterior resection, eviscerating the rectal stump to complete the anastomosis outside the anus. The posterior end of the excision goes down to and includes the posterior part of the internal sphincter. The "Duhamel" is a procedure that brings normally innervated intestine down behind a rectal stump converting them both into one large chamber, again to a level that destroys the back of the internal sphincter. The "Soave" is an endorectal pull-through procedure removing the mucosa but leaving the external muscle layers of the anorectum intact as a sleeve (but cut down the back to avoid stricture), again down to the dentate line. All three of these have been used as a one-stage procedure in the neonate.

Neonatal One-Stage Transanal Approach

The dissection starts through the anus, elevating the mucosa off the underlying smooth muscle from just above the dentate line and then converting to full thickness at 5–6 cm (the level of the pelvic floor and above the ureters). Full-thickness frozen sections are taken as the bowel is eviscerated through the anus, until such time as ganglionic bowel is reached. If the length of the intestine to be eviscerated through the anal canal does not get into the ganglionic bowel (the limit of simple evisceration through the anus is approximately 30 cm), then laparoscopy with 3 mm instruments can be employed to divide the mesocolon to a point where ganglionic bowel can then be eviscerated through the anus.

Full-thickness frozen sections are taken at points at the predicted transition zone, or at any change in caliber, and continue to be taken until the pathologist is sure that there are ganglion cells. The ganglionic bowel is then anastomosed to the dentate line outside the anus. This is done as a one-stage procedure, and the baby is often feeding fully the next day! If the surgeon plans to do a transanal "Soave" (second stage) after doing an emergency colostomy in a sick neonate with Hirschsprung's enterocolitis (first stage), then a colostomy in the LIF is too low, as it will interfere with the mobilization of the colon from below. Then, for the emergency colostomy, its placement should be far more proximal (e.g., at the splenic flexure), so that the Soave can be done later.

A neonatal "Duhamel" can be done as a combined perineal laparoscopic procedure using a stapler from below.

Anatomical Points

The extent of the aganglionic segment(s) is variable in different individuals, and this needs to be determined using biopsy during surgery. Other anomalies or genetic defects may alter the anatomy.

Complications of the Disease

Early

The worst complication is Hirschsprung's enterocolitis. This can be the presenting condition for these babies, and in cases where the diagnosis has been missed, this can be the presentation in the older child. If this occurs as the primary presentation, then the disease must be treated with broad-spectrum antibiotics and a colostomy with frozen section control. Once under control, the definitive surgery can then take the normal pathway(s). Enterocolitis (Teitelbaum et al. 1998) is not limited to those who have yet to undergo surgery, as a few will continue to have bouts of enterocolitis for years after the surgery. Hold up in the intestine is blamed for these recurring attacks, whether this is due to inherent sluggishness in the residual disease or the result of inadequate surgery is unclear.

Table 3.11 Surgery for Hirschsprung's disease estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	5–20 %
Subcutaneous	5–20 %
Intra-abdominal/pelvic (peritonitis; abscess)	5–20 %
Systemic sepsis (especially in the ELBW; premature)	2–5 %
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Small bowel obstruction (postoperative early or late; stricture formation)[a] [Adhesion formation]	1–5 %
Repeated further surgery[a]	1–5 %
Stomal problems[a] (prolapse; intussusception)	5–20 %
Stricture formation[a] (anorectal)	5–20 %
Fecal retention (peculiar to the Duhamel)[a]	50–80 %
Fecal incontinence and fecalomas retention[a]	50–80 %
Hirschsprung's enterocolitis[a]	5–20 %
Peri-anal excoriation[a]	50–80 %
Intellectual impairment[a] (especially ELBW)	50–80 %
Rare significant/serious problems	
Perforation (spontaneous preoperative)[a]	0.1–1 %
Anastomotic leakage[a]	0.1–1 %
Temporary stoma(s)[a]	0.1–1 %
Bowel injury (operative serious)[a]	0.1–1 %
Ureteric injury (very rare)[a]	<0.1 %
Fecal/enterocutaneous fistula(e)[a] (very rare)	<0.1 %
Liver failure from prolonged cholestasis[a]	0.1–1 %
Nutritional deficiency B_{12} malabsorption[a]	0.1–1 %
Wound breakdown[a]	0.1–1 %
Multisystem organ failure (renal, pulmonary, cardiac failure)	0.1–1 %
Mortality[a] (especially ELBW)	50–80 %
Mortality[a] (term infants)	5–20 %
Mortality without surgery (should surgery be refused)[a]	>80 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Nerve parasthesia	0.1–1 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %
Nasogastric tube[a]	1–5 %
Wound drain tube(s)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
Note: ELBW extremely low-birth-weight newborn, <1,000 g
Important Note: Many of these complications are individually closely determined by the exact nature of the problem and surgery. The extent and underlying disease will alter the relative risks

Complications of the Surgery

See Table 3.11. If a staged procedure is used, then there are complications of the stoma, of the later definitive procedure, and the closure of a stoma. If a primary neonatal procedure is used, then the stomal complications do not occur, but stricture rates for the neonatal transanal approach are higher (but respond well to dilatation). Ischemia of the stoma occurs in 1–5 % leading to 'die-back' of the stoma and refashioning. Intussusception of the stoma is uncommon if placed in the sigmoid or low descending colon, but frequently occurs if the stoma is placed in the transverse colon. Postoperative obstruction and stricture formation rates depend on the procedure employed. A stricture rate of up to 5–20 % (more likely in the Swenson, and especially the transanal Soave) can occur, but these strictures can be reversed by almost routine anal canal dilatations, giving a good long-term result.

Later problems include incontinence and fecalomas, with overflow incontinence. Fecal retention occurs in all forms of surgery for Hirschsprung's disease if an aggressive stance is not taken to make sure that the bowel is not regularly emptied. Therefore, the first few years of life have to be supervised closely to make sure that all the necessary medication is given to stop the neorectum from becoming overfull, distended, and ectatic. While this problem is common to all forms of surgery for Hirschsprung's disease, fecal retention in the anterior pouch of a Duhamel procedure can be marked and a very long-term problem with late recurrences.

Fecal incontinence is also a long-term problem after surgery for Hirschsprung's disease. The incontinence may well precipitated by the lack of internal sphincter tone which is a deliberate part of the definitive procedure. If the sphincter is left intact, then enterocolitis and inability to empty in the first years of life are more likely, but if the sphincter is destroyed or partly destroyed by the surgery, then incontinence is more likely as a teenager.

In addition, where the fecal stream has been diverted before the surgery with a stoma, the peri-anal skin appears to be particularly sensitive to local excoriation after bowel continuity is restored. This can be so severe that defecation is very painful, and the surrounding skin can become badly scarred. This excoriation is common to both Hirschsprung's disease and to anorectal malformations where there has been a diverting colostomy prior to surgery.

Major Complications

Serious complications include **anastomotic leakage**, **spontaneous perforation**, **abscess formation,** and **abdominal sepsis**, which may lead to **systemic sepsis**, **multisystem organ failure** and even **death**. **Stricture formation** is not uncommon and may require repeated dilatations. **Repeated further surgery**, with or without stricture, may occur. **Bleeding** intra- and postoperatively can occur. **Fistula formation** may arise. **Fecal incontinence** and/or **fecal impaction** may occur, as can **anal skin excoriation** and the need for a **diverting stoma**. Where used **stomal problems** may arise and be considerable, even requiring further surgery or reversal.

Surgery for Imperforate Anus and Anorectal Malformations

Description

The anal canal is formed from fusion of the skin ectoderm and hindgut endoderm, where they have been separated by the perineal membrane. Failure to do this properly produces the various forms of imperforate anus, perhaps better described as anorectal malformation(s). Developmental fistulae to the perineum, bladder, and urethra occur in the male and to the vagina in the female. In an extreme form in the female, there is a common channel draining the urinary bladder and the hindgut – the cloaca – which can be 1–10 cm in length. Significant anorectal malformations occur in approximately 1:5,000 live births. Minor forms are more frequent but not well documented. They vary considerably from a stenosed opening to apparent absence of the anus. Essentially the anomalies are divided into *high* and *low* lesions. Some surgeons include an *intermediate* category also.

In *high* lesions, the hindgut ends above the sphincter complex and in the male is usually accompanied by a high fistula leading into the urethra or bladder. In the female with a high lesion, any fistula leads into the introitus of the vagina. There are also very complex anomalies that can produce a single channel, the cloaca, with or without duplication of the urogenital tract, and exstrophy of part or all of the urogenital and hindgut systems. A true rectal atresia is rare and appears to be a vascular accident in the already formed rectum. Rectal agenesis is relatively rare and usually occurs in patients with trisomy 21, where the bowel ends in the middle of the sphincters with no fistula. This is now included as an anorectal malformation.

In the *low* lesions, the bowel ends below the sphincter complex, and the end may be covered by skin in the male or comes to the surface as a subcutaneous fistula that travels forward to a varying degree. In the female, the low lesion is represented by an anterior ectopic anus, which is often stenosed, or a small opening in the introitus to the vagina.

Important associated anomalies include genitourinary defects, which occur in approximately 50 % of all patients with anorectal malformations. Associations with other anomalies or genetic problems include VACTERL (vertebral anomalies, anal atresia, cardiac malformations, tracheoesophageal fistula, renal anomalies, and limb anomalies), MURCS (Mullerian duct aplasia, renal aplasia, and cervicothoracic somite dysplasia), OEIS (omphalocele, exstrophy, imperforate anus, and spinal defects), trisomy 21, trisomy 13, trisomy 18, or Hirschsprung's disease.

In males, some 90 % patients require a posterior sagittal approach alone, while in 10 % an abdominal component (with laparotomy or laparoscopically) is necessary to mobilize a very high rectum. However, in females, in some 30 % of cloacas, the rectum or vagina is so high that an abdominal approach is needed (Khong et al. 1994).

Anatomical Points

The anal canal is formed from fusion of the skin ectoderm and hindgut endoderm, and failure produces the various forms of imperforate anus and anorectal agenesis.

Table 3.12 Surgery for imperforate anus/rectal agenesis estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	5–20 %
Subcutaneous	5–20 %
Intra-abdominal/pelvic (peritonitis; abscess) (especially in the ELBW; premature)	5–20 %
Systemic sepsis (especially in the ELBW; premature)	2–5 %
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Constipation[a]	50–80 %
Fecal incontinence and fecalomas retention[a,b]	20–50 %
Urinary incontinence[a,b]	20–50 %
Recurrent fistula(e)[a,b]	5–20 %
Stricture formation (anorectal)[a,b]	5–20 %
Peri-anal excoriation[a,b]	20–50 %
If an abdominal component of surgery is used	
Small bowel obstruction (postoperative early or late)[a] [Adhesion formation]	1–5 %
Stomal problems[a] (prolapse; intussusception)	5–20 %
Incisional hernia[a]	5–20 %
Rare significant/serious problems	
Anastomotic leakage[a]	0.1–1 %
Bowel injury (operative serious)[a]	0.1–1 %
Ureteric injury (very rare)[a]	<0.1 %
Wound breakdown[a]	0.1–1 %
Mortality without surgery (should surgery be refused)[a]	50–80 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Nerve parasthesia	0.1–1 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
[b]Higher in patients with a common cloaca >3 cm
Note: ELBW extremely low-birth-weight newborn, <1,000 g
Important Note: Many of these complications are individually closely determined by the exact nature of the problem and surgery. The extent and underlying disease will alter the relative risks

Developmental fistulae are often present if minimal or no opening exists, and these will often complicate the surgery. True rectal atresia occurs in about 1 % of all cases of anorectal malformations.

Perspective

See Table 3.12.

Complications of the Anomaly

Anorectal malformations are often accompanied by other anomalies as noted, as it forms one of the anomalies that make up the VATER (or VACTERL) association. This includes congenital heart disease, tracheoesophageal fistulae, and renal disease as well as the anorectal malformation. Therefore, as the other lesions could be lethal, they greatly influence the outcome. At birth, intestinal obstruction will supervene in males with high and low lesions (where the lower part of the canal is a small caliber fistula, or the anus is covered with skin). In the female, intestinal obstruction is less common as even the high lesions have a fistula that ends in the introitus or vagina and allows for decompression.

Complications of the Surgery

The level at which the bowel ends (with or without a continuing fistula) dictates the type of surgery. A *low anomaly* where the bowel passes through the sphincters before becoming a fistula requires a local procedure only: the anoplasty. A high lesion where the bowel ends above the sphincters before becoming a fistula requires more complex surgery, which can be done as a primary procedure in the neonate or after a temporary colostomy. The procedure is an anorectoplasty, a type of rectal pull-through reconstruction. As with Hirschsprung's disease, the surgical procedures and especially their timing are changing dramatically. Classical teaching is that for a low lesion, a cutback in either the male or female will suffice. While that may be true in the male, in the female this leaves a scarred perineum with the anterior lip of the anal canal still in the introitus. So, in the female, an anterior sagittal anorectoplasty is used either as a primary procedure after dilating the fistula up to a good size or after a temporary colostomy. In the high lesions in the male, a colostomy can be used while other investigations are carried out, but a primary procedure without a colostomy is now gaining popularity.

For *high lesions*, the classical treatment was a colostomy succeeded by investigations (to determine the actual level, connections to the urinary tract, etc.). Contrast studies through the stoma (if used) and up through the vagina in the female and through the urethra in both genders also help to define the anatomy before definitive surgery. Further investigations, e.g., an MRI to determine as accurately as possible the muscle anatomy (often grossly deficient in high lesions), will help with dissection and prognosis. In addition, the MRI may identify intraspinal lesions that may be the cause of the field defect in the first place. Furthermore, these spinal cord lesions may well contribute to poor sensation in the pelvis, in turn contributing to fecal and urinary incontinence and dysfunction.

Major Complications

Serious complications are less common with *low* anomalies, but include **infection**, **bleeding**, **abscess formation**, **anal stricture formation**, **fistula formation**, **wound breakdown, fecal impaction, constipation, and fecal incontinence** and **anal skin excoriation**. In *higher* lesions, these also occur, but additionally, where abdominal surgery is required, **anastomotic leakage**, **spontaneous perforation**, **abscess formation,** and **abdominal sepsis** may rarely lead to **systemic sepsis**, **multisystem organ failure,** and even **death**. **Repeated further surgery**, with or without stricture, may occur. **Incisional hernia** is reported. The need for a **diverting stoma**, where used may give rise to **stomal problems**, even requiring further surgery, or later reversal.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Feeding problems
- Nutritional deficiency
- Risk of other abdominal organ injury*
- Possible stoma
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Meckel's Diverticulum and Vitellointestinal Remnants

Description

General anesthesia is used. The patient is usually positioned supine. The objective of the operation is to define the Meckel's diverticulum on the antimesenteric border and resect either the diverticulum itself or a section of the small bowel. If perforation has occurred, then drainage of purulent material and/or an abscess and peritoneal washout is necessary. The site of pain may "migrate" as the small bowel moves around in the abdominal cavity, making diagnosis more difficult. When an inverted Meckel's acts as the lead point for an intussusception, resection is necessary with primary anastomosis, but a stoma may be required if there is doubtful viability

+/− peritonitis. A good arterial blood supply in both bowel ends is essential before attempting an anastomosis. A single-layer interrupted technique using absorbable suture material is usually used by pediatric surgeons. In children, stapling techniques are rarely used.

The inspissated mucus of cystic fibrosis may act as a lead point, and the history of this disease should be sought. The disease increases the risks of complications (e.g., inspissated mucous obstruction and paralytic ileus after the procedure) including mortality risk. A patch of mucosa at the umbilicus requires simple excision; otherwise, it will persist, weeping mucus or bleeding. At excision (under a GA), care must be taken to look for a continuation deep into the abdominal cavity. A completely patent vitellointestinal duct is resected down to the true small bowel, through the umbilicus itself, or by minimal enlargement of the cicatrix. If there is a volvulus, it is reduced, and resection may be necessary.

Anatomical Points

The communication between the small bowel and the yolk sac is usually completely obliterated and disappears before birth; however, in approximately 2–5 % of individuals, the communication may persist. A Meckel's diverticulum is one of the remnants of this vitellointestinal duct that connected the apex of the embryological midgut to the yolk sac. The duct can rarely be complete with an intestinal lumen from the ileum to the umbilicus, can be a complete band, can be part of a band with or without cysts in it, can be a Meckel's diverticulum, or can be any combinations of these. A band can serve as a point for volvulus of the small bowel. Rarely, the patent vitellointestinal duct may be associated with umbilical discharge. A Meckel's diverticulum may become inflamed, contain ectopic gastric or pancreatic mucosa, and bleed, obstruct, or ulcerate and perforate (in descending order of frequency). An island of pancreatic mucosa may be found in the intestinal wall without an accompanying diverticulum. The most common remnant, however, is a patch of mucosa in the umbilicus that persists after birth as a "felt-like" granuloma.

The diverticulum is often defined by the "rule of twos," within 2 ft (0.6 m) of the ileocecal valve (but can occur up to the jejunum), 2 in. (5 cm) long, 2 % population, and 2 % become complicated.

Complications of the Anomaly

If the communication is completely patent as the vitellointestinal duct between the bowel and umbilicus, it will then discharge feces soon after birth. A Meckel's can act as the lead point for an intussusception. In this case the lead point can be a simple diverticulum, or the lead point can be a solid island of pancreatic mucosa (where there is no obvious diverticulum). There can be peptic ulceration and hemorrhage at the neck of the Meckel's when gastric mucosa is present. The hemorrhage may be sufficient to produce hypovolemia with marked bleeding PR. A

Table 3.13 Surgery for Meckel's diverticulum and vitellointestinal remnants estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	1–5 %
Subcutaneous	1–5 %
Intra-abdominal/pelvic (peritonitis; abscess) (especially in the ELBW; premature)	1–5 %
Systemic sepsis (especially in the ELBW; premature)	1–5 %
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Small bowel obstruction (postoperative early or late)[a] [Adhesion formation]	1–5 %
Rare significant/serious problems	
Anastomotic leakage[a]	0.1–1 %
Bowel injury (operative serious)[a]	0.1–1 %
Mortality	<0.1 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
Note: ELBW extremely low-birth-weight newborn, <1,000 g
Important Note: Many of these complications are individually closely determined by the exact nature of the problem and surgery. The extent and underlying disease will alter the relative risks

simple Meckel's itself can give rise to local inflammation and peritonitis similar to appendicitis, especially where the Meckel's is narrow necked. Cysts in the remnant can be in the abdominal wall, where they can give rise to repeated infections and discharge through the umbilicus. Where there is a band from the abdominal wall to point of attachment to the intestine, segmental volvulus can occur around this band.

Perspective

See Table 3.13. The complications of any of these procedures will depend on the initial pathology encountered. Where there is bowel obstruction, preoperative perforation and sepsis increase the risk of postoperative complications, the most serious complication being anastomotic leakage, the risk of which is increased by distal obstruction of any cause. It is therefore vital to alleviate any significant obstruction distal to level of anastomosis (but this is highly unlikely with a Meckel's). The consequence of an anastomotic leakage is contamination of the peritoneal cavity leading to generalized peritonitis or intra-abdominal abscess formation, typically in the paracolic gutters, pelvis, or the subphrenic spaces. The incidence of anastomotic

leakages is reduced by ensuring good blood supply to the bowel ends, minimal tension, and no factors contraindicating an anastomosis. Wound infection, small bowel obstruction, and enterocutaneous fistula are significant, but fortunately uncommon complications.

Major Complications

Where there is bowel obstruction, preoperative perforation and sepsis increase the risk of postoperative complications. The most serious complication is **anastomotic leakage**. But, this is uncommon in the young patient. **Wound infection**, **small bowel obstruction,** and **enterocutaneous fistula** are significant, but fortunately uncommon complications. **Systemic sepsis** is uncommon, but is increased when delayed diagnosis occurs, and is rarely followed by multisystem organ failure and even death.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Possible stoma
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Intussusception

Description

General anesthesia is used. The patient is positioned supine. The objective of the operation is to define the problem and alleviate the obstruction. Resection of bowel may be required depending on the pathology and degree of fixity, obstruction, ischemia, or necrosis present. For children, a transverse muscle-cutting incision is typically used.

Intussusception is an invagination (telescoping) of proximal bowel into distal bowel, eventually causing obstruction. This is not a common disease in the

newborn, but is a common disease in infants from 6 to 10 months of age, and after that, the disease gradually becomes less common again. It is temporally related to the time of weaning, the introduction of non-sterile foods, and the seasons when viral infection (and possibly localized swelling of Peyer's patches) is more common (spring and autumn). The usual form is an invagination of the terminal ileum and ileocecal junction into the ascending colon. It is the most frequent cause of intestinal obstruction in the first 2 years of life. Some 75–90 % of cases can be reduced by using a form of hydrostatic enema (barium, air, or saline), under imaging control.

Over the years, that has progressed from a barium enema to an air enema, to ultrasound (U/S) diagnosis (far more accurate than a plain X-ray) with ultrasound-controlled reduction using a saline enema. The latter avoids all radiation (some 70 % of barium enemas ordered for suspected intussusception showed that there was none) and avoids risk of perforation and barium peritonitis. Surgery may be required if hydrostatic pressure reduction is unsuccessful or, for multiple recurrences, as either an open or a laparoscopic procedure, with the occasional patient requiring bowel resection for ischemia, or more rarely because there is a small bowel tumor (6 % chance in the older age groups, and usually a lymphoma).

Anatomical Points

There are few real anatomical aspects that are pertinent to intussusception, except for the presence of a Meckel's diverticulum, swollen Peyer's patches, or bowel tumor mass which can all act as a firm leading bolus which can then be moved down the gut with peristaltic contractions.

Complications of the Lesion

Intussusception will progressively involve more and more of the bowel as it telescopes. This leads to a bowel obstruction and to progressive loss of blood supply. Eventually there will be a venous gangrene of the intussusceptum. Once that has become established, bowel resection is inevitable. If the intussusception still has not been treated, then bacteremia and endotoxic shock can ensue.

Complications of Nonsurgical Intervention (Radiology)

Hydrostatic or pneumostatic reduction using an enema (with imaging) can lead to a perforation of the weakened bowel as pressure is applied. The combination of barium and feces in the peritoneal cavity is potentially lethal, so that other modalities have been sought. The first was to use air with an image intensifier, then saline reduction using U/S. The last has the advantages of being able to diagnose the lesion (before the enema) and to effect a treatment, both without radiation.

Table 3.14 Surgery for intussusception estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	1–5 %
Subcutaneous	1–5 %
Intra-abdominal/pelvic (peritonitis; abscess) (especially in the ELBW; premature)	1–5 %
Systemic sepsis (especially in the ELBW; premature)	1–5 %
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Small bowel obstruction (postoperative early or late)[a] [Adhesion formation]	1–5 %
Recurrent intussusception[a]	5–20 %
Rare significant/serious problems	
Perforation[b] (spontaneous preoperative)	0.1–1 %
Anastomotic leakage[a] (when performed)	0.1–1 %
Bowel injury (operative serious)[a]	0.1–1 %
Mortality	<0.1 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
[b]Spontaneous perforation may occur, as can perforation in association with hydraulic reduction techniques
Note: ELBW extremely low-birth-weight newborn, <1,000 g
Important Note: Many of these complications are individually closely determined by the exact nature of the problem and surgery. The extent and underlying disease will alter the relative risks

Complications of Surgery

Failure to reduce the intussusception will lead to a bowel resection. But, in the more common ileo-ileocolic intussusception, reduction can often be achieved even after failure of radiological intervention. If a resection is still required, then there is the potential for stricturing at the site of the anastomosis if the ends of the intestine were still ischemic at the points of resection. More likely to cause a stricture, however, is where partially devitalized intestine is put back after reduction without resection.

Recurrences

Recurrences after radiological or surgical reduction do occur, approximately 10 % after radiological reduction and approximately 5 % after surgical reduction. The first recurrence is treated in the same way as the first presentation with similar results. Repeated

recurrences occur and are treated in a similar fashion. Individual surgeons have different thresholds for surgery to try to prevent repeated recurrences, and the surgery for the recurrences is not always successful. Carrying out a fixation of the cecum is now rarely used. A simple appendectomy diminishes the recurrence rate but not entirely; a right hemicolectomy is radical but is nearly always successful. But, just waiting through the multiple recurrences that can occur is as successful, but distressing to the family.

Major Complications

See Table 3.14. Where there is bowel obstruction, preoperative perforation and sepsis increase the risk of postoperative complications. The most serious complication is **anastomotic leakage**. But, this is uncommon in the young patient. **Wound infection**, **small bowel obstruction**, and **enterocutaneous fistula** are significant, but fortunately uncommon complications. **Recurrent intussusception** can occur. **Small bowel obstruction** due to adhesions may occur at any stage. **Systemic sepsis** is uncommon, but is increased when delayed diagnosis occurs, and is rarely followed by multisystem organ failure and even death.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Possible stoma
- Possible further surgery*
- Risks without surgery*
- Multisystem organ failure
- Death*

***Dependent on pathology, comorbidities, and surgery performed**

Pediatric Surgery in the Older Child

Introduction

Abdominal wall defects in the older child are typically small umbilical herniae due to failure of a congenital defect to close completely after birth and on

occasions may be larger and strangulate or obstruct. If these herniae fail to close by 2 years of age or are symptomatic, surgical repair is advisable. Surgery for inguinal herniae is performed in the newborn and infant in most situations, where early repair is advisable, but may also present in the older child and require surgical repair.

General abdominal surgical procedures in the older child include a variety of procedures principally related to inflammation, reflux, feeding access, or tumor.

Some of the surgical procedures described in section "Pediatric Surgery in the Newborn and Infant" may apply to surgery in the older child in certain situations, such as when diagnostic features are more subtle, for example, from incomplete lower-grade gut obstruction, which may present later.

Abdominal Wall Defect Surgery

Surgery for Umbilical/Supraumbilical/Epigastric Herniae

Description

General anesthesia is used. Umbilical herniae (UH) are present in up to 85 % of premature newborns and approximately 20 % of full-term newborns. Over 90 % will close by the age of 2–5 years in Caucasians, but much later (teenagers) in Africans. Therefore, surgery should not be performed for simple umbilical herniae until after these ages, respectively, unless symptomatic. Supraumbilical herniae virtually never close on their own, so surgery can be carried out at the most suitable age for the child and the institution concerned (outside children's hospitals, many anesthetists are uneasy about GA use for children <3 years of age). Umbilical herniae are through the embryological physiological defect at the umbilical cord where the expanding midgut herniates, but then returns to the abdominal cavity by 10 weeks of gestation. Supraumbilical herniae are also congenital defects in the abdominal wall cicatrix just above the center of the umbilicus, then turning inferiorly. Paraumbilical hernial defects laterally to the umbilicus are extremely rare in children. In those over 2 years of age with central umbilical hernia and in those with a supraumbilical hernia, the surgery is essentially the same. A peritoneal sac is usually present containing omentum, or less commonly bowel, particularly in larger herniae. In obese children, the diagnosis may be especially difficult. Symptoms from an umbilical hernia are extremely rare in children; therefore, surgery is essentially for cosmetic reasons alone. Incarceration and bowel obstruction are almost unheard of in children but would require emergency intervention. A transverse skin incision is made below the umbilicus. The sac is defined and excised at the edge of the linea defect and from the overlying skin of the umbilicus. The contents are reduced, and the defect is closed as an overlapping two-layer closure (Mayo) with heavy absorbable sutures or as a two-layer turned-in (Keel) repair. Mesh is sometimes used.

Table 3.15 Surgery for umbilical/supraumbilical/epigastric herniae in the older child estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection	1–5 %
Hernia recurrence[a] (10 year)	0.1–1 %
Suture abscess +/− suture sinus[a]	0.1–1 %
Bleeding (including intra-abdominal)	<0.1 %
Less serious complications	
Pain/discomfort/tenderness	
(<2 months; days only in children)	20–50 %
Pain/discomfort/tenderness	
(>2 months)	0.1–1 %
Bruising or hematoma formation[a]	5–20 %
Dimpling/deformity of the skin[a]	1–5 %
Scarring	1–5 %

[a]Dependent on underlying pathology, anatomy, hernia type, surgical technique, and preferences

Epigastric herniae are midline defects in the linea alba anywhere between the umbilicus and the xiphoid. Because the falciform ligament and its fat are deep to these defects, they never contain viscera. They rarely cause symptoms, but the free edge of the defect may nip the herniated fat to cause very localized pain. If that happens, then it is worthwhile closing the defect with absorbable sutures; otherwise, it is again a cosmetic issue. These can be seen in the child, but effectively disappear in the adult as the subcutaneous fat becomes thicker.

Anatomical Points

The main variance is in the site of the hernia as described above, and extreme versions of each, which are exceedingly rare. Multiple defects are also extremely rare.

Perspective

See Table 3.15. The complications related to UH, SUH, and epigastric hernia repair are generally minimal. Local bruising and superficial wound infection may occur.

Major Complications

Infection from skin, and occasionally bowel organisms, may occur. **Bleeding** is seldom severe, often from omental or mesenteric vessels, and may increase risk of infection. Established infection may rarely necessitate **removal of mesh or sutures**.

Hernia recurrence is uncommon and further increased with obesity.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Hernia recurrence
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

General Abdominal Surgery

Open Gastrostomy

Description

General anesthesia is used. The aim is to establish a portal to the stomach from the exterior so as to be able to feed the child. A small RUQ transverse incision is made over the stomach at a position where the gastrostomy will not impinge on the rib cage (as the child grows, the skin migrates upward). At open surgery, a button device (a short, valved feeding tube that has a small flat external flange which sits flush on the skin) is usually inserted directly into the stomach and held in place with two purse-string sutures. The stomach is also sutured to the anterior abdominal wall, with at least three sutures. Occasionally an 18 G Foley catheter may be used rather than a button. The abdomen is then closed around the gastrostomy. Some surgeons will bring the gastrostomy out through a separate stab wound.

Anatomical Points

The colon, small bowel, liver, and omentum may overlie the stomach and make access difficult. Although these organs are at risk, generally these can be displaced easily in children to enable the procedure to be performed. Patients with cerebral palsy often have curved spines that will occasionally prevent the stomach being brought to the anterior abdominal wall. Then, an alternative procedure will have to be found (e.g., a feeding jejunostomy).

Table 3.16 Open gastrostomy estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection	
Wound granulation around the button/catheter	5–20 %
Subcutaneous cellulitis; abscess	1–5 %
Intraperitoneal leak	1–5 %
Systemic[a]	0.1–1 %
Bleeding/hematoma formation	1–5 %
Gastric leakage[a]	5–10 %
Gastrocutaneous fistula (persistent after removal)	1–5 %
Aspiration pneumonitis	
Overall	1–5 %
In those with CNS defects	5–20 %
Less serious complications	
Discharging abscess sinus	1–5 %
Incisional hernia formation	0.1–1 %
Tube dislodgment (internalization or extraction)	5–20 %
Gastroesophageal reflux[a]	5–20 %
Paralytic ileus	1–5 %
Scarring/wound deformity	5–20 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

Perspective

See Table 3.16. Pediatric gastrostomy is used for feeding and very rarely for drainage. Gastrostomy feeding is especially important for patients who are unable to feed or spill into their lungs on feeding. The most common type of patient will have brain damage, e.g., patients with cerebral palsy. The procedure is often combined with a fundoplication, as a gastrostomy will usually increase any reflux that is present or may cause reflux de novo. Some 5–20 % of patients will get reflux for the first time, and others will experience deterioration of any reflux already present. Where possible a percutaneous endoscopic gastrostomy (PEG) is used, so that an open procedure is avoided. Recently laparoscopic gastrostomy has been introduced using increasing sizes of dilator introduced under laparoscopic control and over a guide wire. This is especially useful in those who have undergone a fundoplication as part of the same procedure.

Major Complications

If a Foley catheter is used as the first access device (rather than a button), the balloon of the Foley catheter may occasionally migrate distally and can lead to **stomach outlet obstruction**. Therefore, buttons are now more commonly used as the initial device after open fundoplication in children, as being so short they cannot migrate. Separation of the stomach from the anterior abdominal wall may result in **intraperitoneal**

leakage of stomach contents and peritonitis, with or without abscess formation or generalized sepsis. This is most likely to occur at the first change of a button or catheter. If the new button or catheter is pushed down the track blind, the stomach can be pushed off the abdominal wall. Therefore, in pediatric practice, the first change is often covered by an upper GI endoscopy to make sure that the new device is in place. **Pressure necrosis** of the stomach against the catheter balloon and free **perforation** is rare. The most frequent complication, however, occurs around the exit of the catheter where **minor infection** and exuberant **granulation tissue** form**. Gastric acid leaks around the tube** may cause **excoriation.** Associated **abscess formation** is uncommon. **Systemic sepsis** is rare, but may be severe, often related to the underlying condition(s), and can lead to death. Persistent **gastrocutaneous fistula** after removal of the device is not uncommon after prolonged use of a gastrostomy and infrequently can be persistent, but most close. **Aspiration pneumonitis** may occur in any patient, but risk in those with cerebral palsy and other CNS lesions may be as high as 20 %.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Intraperitoneal leakage
- Leakage around tube
- Skin excoriation
- Migration and pyloric obstruction
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Percutaneous Endoscopic Gastrostomy

Description

In children, general anesthesia is used for percutaneous endoscopic gastrostomy (PEG). The aim is to establish a portal to the stomach from the exterior. The peroral endoscope is turned anteriorly inside the stomach so that the scope light is visible through the anterior abdominal wall. The surgeon placing the PEG indents the stomach at the best point to place a gastrostomy. A small stab incision is made through the skin only, and a trocar and cannula (in the PEG set) is inserted

into the stomach under endoscopic control. A flexible looped wire is inserted through this into the stomach and grasped by the endoscopist and pulled out with the endoscope through the mouth. The PEG catheter (usually 18FG has a pointed tip with a guide wire and a flange at the rear end) is then tied to the wire and pulled back point first through the stomach and abdominal wall. The flange at the back end of the catheter pulls the stomach wall up against the abdominal wall. There are no stitches so tension has to be maintained by using a tightly fitting external flange. This catheter will be replaced by a button 6 weeks to 3 months later, under endoscopic control. The stomach wall is usually adherent to the abdominal wall by then, thereby avoiding leakage of feeds into the peritoneal cavity, although this can occur at any time, should the stomach and abdominal wall separate.

Anatomical Points

The colon, small bowel, liver, and omentum may overlie the stomach, and as the stomach is inflated by the gastroenterologist to get visualization, the greater curvature of the stomach tends to rotate upward pulling the colon and small bowel in front. As a result, the trocar and cannula may then go through one of those before getting into the stomach. This can lead to the PEG being gradually pulled through the stomach wall over time and dislodging into that organ (colon or small bowel). Once that has occurred, it will produce marked diarrhea. Obesity or curvature of the spine may also make the procedure more challenging.

Perspective

See Table 3.17. Gastrostomy is used for feeding in children. Percutaneous endoscopic gastrostomy (PEG) is almost exclusively used for gastric access, where an endoscopy can be performed. Complications are few, but misplacement of the catheter through another viscus can occur, and skin infection and irritation are common. Change from the PEG catheter to a button has a sufficiently high risk of pushing the stomach away from the abdominal wall that endoscopy is now routinely used at this first change to make sure that the new device has travelled down the new track correctly and is now in the stomach. The risk of intraperitoneal feed leakage can be as high as 10 % if endoscopy is not used at the first changeover. The balloon and valved buttons frequently fail and have to be replaced. Endoscopy is not usually required after the first change, unless difficulties are encountered. Inability to perform the procedure can be as high as 10 % especially in the very young, those with a badly deformed spine and oro-esophageal problems, after prior upper GI surgery, and if the procedure is accompanied by a laparoscopic fundoplication. Aspiration pneumonitis may be as high as 20–30 % in those with CNS defects. Perforation of the colon or small bowel by the catheter during PEG insertion can be as high as 5–10 %, but can be reduced with proper illumination

Table 3.17 Percutaneous endoscopic gastrostomy estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection	
Wound granulations	5–20 %
Subcutaneous cellulitis; abscess	1–5 %
Intraperitoneal leak	1–5 %
Systemic	0.1–1 %
Bleeding/hematoma formation[a]	1–5 %
Gastric leakage into the peritoneal cavity[a] (higher risk at first changeover)	1–5 %
Persistent gastric fistula (following removal)	1–5 %
Discharging abscess sinus	1–5 %
Free esophageal/gastric perforation	1–5 %
Failure to perform endoscopically[a]	5–20 %
Tube dislodgment (internalization or extraction) and duodenal obstruction	5–20 %
Aspiration pneumonitis	
Overall	1–5 %
In those with CNS defects	5–20 %
Catheter going through colon or small bowel[a]	1–5 %
Conversion to open laparotomy/laparoscopy[a] (early or late)	0.1–1 %
Less serious complications	
Paralytic ileus	1–5 %
Pneumoperitoneum	5–20 %
Gas bloating (transient)	5–20 %
Gastroesophageal reflux	5–20 %
Injury to mouth, teeth, pharynx, or larynx	1–5 %
Hernia formation (incisional)	0.1–1 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

and extra care. Laparoscopic gastrostomy can now be employed which avoids the potential for an esophageal injury and also reduce risk of colonic or small bowel injury.

Major Complications

Occasionally, the flange of the initial PEG catheter may migrate distally and can lead to **stomach outlet obstruction**. Separation of the stomach from the anterior abdominal wall will result in **intraperitoneal leakage** of stomach contents and **peritonitis**, with or without **abscess formation** or generalized **sepsis**. **Pressure necrosis** of the stomach against the catheter balloon and free **perforation** is very rare in children. The most frequent complication, however, occurs around the exit of the catheter where exuberant **granulation tissue** and **excoriation** from acid leakage are common. Associated **abscess formation** is not uncommon. **Systemic sepsis**

is infrequent, but may be severe, often related to the underlying condition(s), and can lead to **death.** Although PEG approach is usually easy and safe, additional risks of **esophageal perforation** or **teeth injury** can occur, but are very infrequent. Persistent **gastrocutaneous fistula** is not uncommon after prolonged use of a gastrostomy and infrequently can be persistent, but most close.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Intraperitoneal leakage
- Leakage around tube
- Skin excoriation
- Migration and pyloric obstruction
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Open Gastroesophageal (Nissen) Fundoplication

Description

General anesthesia is used. The aim is to form a ring of stomach around the lower part of the esophagus, as a complete sleeve, by wrapping the upper fundus of the stomach from the left around behind the lower esophagus to bring it in front of the esophagus, to be sutured in front to the non-wrapped part of the fundus. In that way, it acts as a higher-pressure zone, allowing food and drink in, but not allowing either to reflux. There are many variations to this operation; however, the basic variations are:

1. Taking the anterolateral wall of the fundus of the stomach and sliding it up and around behind the esophagus and then sewing it to itself in front of the esophagus. This operation does not require division of the short gastric vessels.
2. Division of the short gastric vessels may be required if the spleen is pulled into the gap behind the stomach as the wrap is achieved. In pediatric patients, short gastric division is rarely required if the bands from the diaphragm to the esophagus and stomach are adequately divided – especially on the left side.

Most surgeons use a large bougie (30–36 FrG) within the esophageal lumen, while the wrap is being undertaken to try to prevent making the wrap too tight, but with experience, the bougie is often no longer required.

Anatomical Points

Very infrequently the anterior wall of the stomach is not generous enough to be taken around behind the esophagus without division of the short gastric vessels. The short gastric vessels may be very high on the greater curvature of the stomach or tightly applied to the spleen. The abdominal esophagus may be very short. Adhesions to the spleen, diaphragm, or colon may exist. Liver or splenic enlargement may reduce access to the stomach. Rare anomalies of the vascular supply to the stomach may render the stomach fundus susceptible to ischemia upon division of the short gastric vessels, especially with tensioning of the stomach. There may be a hiatus hernia, which will have to be dealt with, and the extra care taken in the closure of the hiatus. Occasionally the stomach cannot be brought to the abdominal wall because of spinal curvature blocking the access (not uncommon in cerebral palsy patients).

Perspective

See Table 3.18. Many types of fundoplication exist, and within the Nissen-type alone, controversy persists as to whether the short gastrics should or should not be divided in total fundoplications. The "physiological fundoplication" of Boix Ochoa uses a procedure that increases the angle of His and does an anterior wrap only suturing it to the right crus and then a hitch of the fundus to the left diaphragm. Partial fundoplications are variously described with the wrap only encircling the posterior 180°–270° of the esophagus, and similarly anterior partial fundoplications, encircling part of the anterior part of the esophagus. In those who are neurologically impaired, a partial fundoplication rarely works. They may, however, be appropriate for a second procedure in neurologically intact children, where the first fundoplication produced unrelenting dysphagia that has to be relieved surgically. Even then, they frequently fail. As the child grows, the wraps become looser, so that partial wraps are more likely to fail with time, and tight wraps will slacken, so that dysphagia relents. Open fundoplications have been largely replaced by laparoscopic approaches. Exceptions occur. The most common would be where there have been multiple abdominal procedures and adhesions; the most extreme would be where neither an open abdominal nor a laparoscopic procedure can be achieved. An example would be after an exomphalos major repair at birth, where the liver now overlies the stomach and is attached to the spleen with adhesions, making access to the upper stomach impossible through the abdomen alone, where a thoracoabdominal approach is often necessary. Dysphagia is frequent early but declines to 5–10 % by 6 months. Similarly, gas bloat syndrome is higher early on but improves over the

Table 3.18 Open gastroesophageal (Nissen) fundoplication estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	
Subcutaneous/wound	1–5 %
Intra-abdominal	0.1–1 %
Mediastinitis	<0.1 %
Intrathoracic (pneumonia; pleural)	1–5 %
Subphrenic abscess	0.1–1 %
Systemic	0.1–1 %
Late – post-splenectomy sepsis (vaccination)	<0.1 %
Bleeding and hematoma formation[a]	1–5 %
Dysphagia (>6 months postoperatively)	5–20 %
Persistent or recurrent gastroesophageal reflux (lifetime)	5–20 %
Gas bloat syndrome	
(<6 months postoperatively)	5–20 %
(>6 months postoperatively)	1–5 %
Inability to vomit or belch[a]	5–20 %
Diaphragmatic injury	1–5 %
Diaphragmatic (paraesophageal or wrap) herniation[a]	5–20 %
Breakdown of fundoplication[a]	
Overall	1–5 %
In the neurologically impaired	20–50 %
Delayed gastric emptying[a]	1–5 %
Bilious vomiting	1–5 %
Dumping syndrome[a]	
(>6 months postoperatively)	1–5 %
(<6 months postoperatively)	5–20 %
Diarrhea	5–20 %
Small bowel obstruction (early or late; lifetime risk)[a] [Adhesion formation]	1–5 %
Rare significant/serious problems	
Liver injury[a]	0.1–1 %
Splenic injury[a] (rarely splenectomy)	0.1–1 %
Pancreatic injury/pancreatitis/pancreatic cyst/leakage/pancreatic fistula	0.1–1 %
Bowel injury (duodenum, small bowel, colon)[a]	0.1–1 %
Pneumothorax	0.1–1 %
Death[a]	<0.1 %
Less serious complications	
Paralytic ileus	1–5 %
Intolerance of large meals (necessity for small frequent meals)	50–80 %
Excessive flatus[a]	50–80 %
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Wound scarring (poor cosmesis/wound deformity)	1–5 %
Incisional hernia	0.1–1 %
Nasogastric tube[a]	50–80 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

years. Inability to vomit or belch is much higher in the pediatric population, but is diminishing as the wraps are now shorter and looser. Dysphagia, gas bloat syndrome, and inability to vomit or belch reflect a too tight wrap. Persistent gastroesophageal reflux in pediatrics, which reflects failure or excessive looseness of the wrap, is ~5–10 % for otherwise normal children, but as high as 30–50 % in neurologically impaired children.

The incidence of hiatus hernia, including or excluding the wrap, is high in children where (in days gone by) the hiatus was not closed behind the esophagus. There is probably a 15 % increased lifetime risk of hiatus hernia, and this could be as high as 20 % in the first 5 years in those who are neurologically impaired, but again closing the hiatus has reduced this substantially. Breakdown of the fundoplication occurs in ~1–5 % of cases overall, but up to 50 % in neurologically impaired patients. Similarly, delayed gastric emptying occurs in ~1–5 % of cases, but increases to ~20 % of neurologically impaired patients.

Major Complications

The most common major complication is **aphagia**, or very severe **dysphagia**, which can occasionally require very **early reoperation** (within days). **Persistent or recurrent gastroesophageal reflux** can occur after failure of the surgery either initially or later, with loosening of the wrap. Damage to the spleen that requires **splenectomy** is very rare in children. Full-thickness **esophageal or gastric** damage is a major complication, but this occurs very infrequently indeed; in primary anti-reflux surgery, it is usually seen immediately and repaired immediately. **Infection and multisystem failure** are very rare, but a late perforation or undetected leak can occur to cause this. **Bleeding** is rare and is usually controlled at surgery.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Intraperitoneal leakage
- Dysphagia
- Recurrence of reflux
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Laparoscopic Gastroesophageal (Nissen) Fundoplication

Description

General anesthesia is used. The aim of the procedure is the production of a one-way valve between the esophagus and stomach and is identical to that for open fundoplication. Most surgeons use five ports in this procedure, although the epigastric port used for retraction is sometimes replaced with the Nathanson hook retractor. In pediatric patients, division of the short gastric vessels is rarely required. The mobilization of the anterior wall of the stomach and the esophagus is similar to that carried out in open fundoplication with the exception that dissection behind the esophagus and exposure of the pillars of the hiatus are more frequently carried out during laparoscopic fundoplication. This is because it is necessary to create a clear window behind the esophagus to safely and more easily draw the stomach through behind the esophagus. For this reason almost all surgeons today practice narrowing of the hiatus posteriorly with one, two, or more sutures.

Anatomical Points

The chief variation which has practical importance in laparoscopic fundoplication relates to the size of the left lobe of the liver. When this is large and bulky, it can obscure vision of the hiatus. Previous surgery can be problematic, altering anatomy. Very rarely, abnormal blood supply of the stomach fundus may cause ischemia, especially after short gastric vessel division.

Table 3.19 Laparoscopic gastroesophageal (Nissen) fundoplication estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	
Subcutaneous/wound	1–5 %
Intra-abdominal	0.1–1 %
Mediastinitis	<0.1 %
Intrathoracic (pneumonia; pleural)	1–5 %
Subphrenic abscess	0.1–1 %
Systemic	0.1–1 %
Late – post-splenectomy sepsis (vaccination)	<0.1 %
Bleeding or hematoma formation[a]	1–5 %
Dysphagia (>6 months postoperatively)	5–20 %
Persistent or recurrent gastroesophageal reflux (lifetime)	5–20 %
Gas bloat syndrome	
(<6 months postoperatively)	5–20 %
(>6 months postoperatively)	1–5 %

Table 3.19 (continued)

Complications, risks, and consequences	Estimated frequency
Inability to vomit or belch[a]	5–20 %
Conversion to open operation	1–5 %
Diaphragmatic injury	1–5 %
Diaphragmatic (paraesophageal or wrap) herniation[a]	5–20 %
Breakdown of fundoplication[a]	
Overall	1–5 %
In the neurologically impaired	20–50 %
Delayed gastric emptying	1–5 %
Bilious vomiting	1–5 %
Dumping syndrome	
(>6 months postoperatively)	1–5 %
(<6 months postoperatively)	5–20 %
Diarrhea	5–20 %
Small bowel obstruction (early or late; lifetime risk)[a] [Adhesion formation]	1–5 %
Rare significant/serious problems	
Splenic injury[a] (rarely splenectomy)	0.1–1 %
Pancreatic injury/pancreatitis/pancreatic cyst/leakage/pancreatic fistula	0.1–1 %
Bowel injury (duodenum, small bowel, colon)	0.1–1 %
Liver injury	0.1–1 %
Pneumothorax	0.1–1 %
Gas embolus	0.1–1 %
Less serious complications	
Paralytic ileus	1–5 %
Intolerance of large meals (necessity for small frequent meals)	50–80 %
Excessive flatus[a]	50–80 %
Surgical emphysema	1–5 %
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Wound scarring (poor cosmesis/wound deformity)	1–5 %
Port-site herniae	0.1–1 %
Incisional hernia	0.1–1 %
Nasogastric tube[a]	50–80 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

Perspective

See Table 3.19. Many types of fundoplication exist, and within the Nissen-type alone, controversy persists as to whether the short gastrics should or should not be divided in total fundoplications, but in pediatric surgery, short gastric division is rarely used. The "physiological fundoplication" of Boix Ochoa uses a procedure that increases the angle of His and does an anterior wrap only suturing it to the right crus and then a hitch of the fundus to the left diaphragm. Partial

fundoplications are variously described with the wrap only encircling the posterior 180°–270° of the esophagus, and similarly anterior partial fundoplications, encircling part of the anterior part of the esophagus. In those who are neurologically impaired, a partial fundoplication rarely works. They may, however, be appropriate for a second procedure in neurologically intact children, where the first fundoplication produced unrelenting dysphagia that has to be relieved surgically. Even then, they frequently fail. As the child grows, the wraps become looser, so that partial wraps are more likely to fail with time, and tight wraps will slacken, so that dysphagia relents. Laparoscopic fundoplications have largely replaced open approaches. Exceptions occur. The most common would be where there have been multiple abdominal procedures and adhesions; the most extreme would be where neither an open abdominal nor a laparoscopic procedure can be achieved. An example would be after an exomphalos major repair at birth, where the liver now overlies the stomach and is attached to the spleen with adhesions, making access to the upper stomach impossible through the abdomen alone, where a thoracoabdominal approach is often necessary. Dysphagia is frequent early but declines to 5–10 % by 6 months. Similarly, gas bloat syndrome is higher early on but improves over the years. Inability to vomit or belch is much higher in the pediatric population, but is diminishing as the wraps are now shorter and looser. Dysphagia, gas bloat syndrome, and inability to vomit or belch reflect a too tight wrap. Persistent gastroesophageal reflux in pediatrics, which reflects failure or excessive looseness of the wrap, is about 5–10 % for otherwise normal children, but as high as 30–50 % in neurologically impaired children. The incidence of hiatus hernia, including or excluding the wrap, is high in children where (in days gone by) the hiatus was not closed behind the esophagus. There is probably a 15 % increased lifetime risk of hiatus hernia, and this could be as high as 20 % in the first 5 years in those who are neurologically impaired, but again closing the hiatus has reduced this substantially. Breakdown of the fundoplication occurs in about 1–5 % of cases overall, but up to 50 % in neurologically impaired patients. Similarly, delayed gastric emptying occurs in ~1–5 % of cases, but increases to around 20 % of neurologically impaired patients.

Major Complications

The most common major complication is **aphagia**, or very severe **dysphagia**, which can occasionally require very **early reoperation** (within days). **Persistent or recurrent gastroesophageal reflux** can occur after failure of the surgery either initially or later, with loosening of the wrap. Damage to the spleen that requires **splenectomy** is very rare in children, and splenic injury occurs far less frequently in laparoscopic surgery than in open surgery. Full-thickness **esophageal or gastric** damage is a major complication, but this occurs very infrequently indeed; in primary antireflux surgery, it is usually seen immediately and repaired immediately. **Infection and multisystem failure** are very rare, but a late perforation or undetected leak can occur to cause this. **Bleeding** is rare and is usually controlled at surgery. **Gas embolus** and **major vascular injury** are additional serious, although very rare,

complications of the laparoscopic approach. **Conversion to open operation** is a small but significant risk rather than a complication per se.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Intraperitoneal leakage
- Gas embolism
- Conversion to open surgery
- Dysphagia
- Recurrence of reflux
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Open Appendectomy

Description

General anesthesia is used. The patient is positioned in the supine position and is best examined when anesthetized to assess whether there is a mass. This may influence the decision to carry out an open (more likely with a mass) procedure or laparoscopic procedure and to determine the best site for the incision, if there is a mass. Rectal examination under anesthesia may be useful to assess the presence of any pelvic mass. Nevertheless, an ultrasound may have been performed. The objective of the operation is to remove the appendix. If, at laparotomy, the appendix does not appear to be inflamed, then the next objective is to determine if there is other pathology by examining the pelvis for pelvic pathology, particularly in the female, and the terminal ileum for the presence of a Meckel's diverticulum or other pathology causing local peritonitis. If there is a large mass, then open appendectomy should be carried out in children, as interval appendectomy does not appear to be warranted. In children, it is rare to be unable to gain access to other pathology from an extended RIF incision. Under most circumstances the appendix can be removed using a small transverse (Lanz) skin incision and a muscle-splitting incision of the internal oblique muscle. A small incision is often made to obtain a good cosmetic result. Surgeons should never hesitate to increase the length of the skin incision and either divide the

internal oblique muscle or cut the anterior rectus fascia, retracting the rectus to the midline and opening the posterior rectus sheath after dealing with the inferior epigastric vessels, or both, to provide better access to the peritoneal cavity. Under these circumstances, the cecum should be mobilized by dividing the congenital adhesions to bring the cecum well into the wound to display the full length of the appendix, particularly its junction with the cecum.

Anatomical Points

The appendix origin lies at the confluence of the taenia coli; however, its tip can vary enormously in position, lying retrocecally (~75 % cases), pelvic (20 %), or retro-ileal/pre-ileal (5 %). The length of the appendix varies also and can reach the upper ascending colon posteriorly. Rarely the appendix (very rare) and cecum may enter a large inguinal hernial sac. Irritation of the bladder or colon can cause urinary urgency and/or diarrhea. An inflamed appendix, if retrocecal or pelvic in location, may irritate the ureter, so that hematuria or dysuria may occur. Irritation of the psoas muscle by an inflamed retrocecal appendix or abscess may cause hip discomfort on movement. Maldescent of the appendix is rare due to malrotation of the cecum, which remains high in the hepatic region. Agenesis, duplication, and situs inversus (left-sided appendix) are exceedingly rare, but can occur. There are, however, a series of patients with left atrial isomerism (picked up on antenatal scans) who are known to have situs inversus.

Perspective

See Table 3.20. Infective complications are the most common following appendectomy, wound infection being the most frequent. This is often avoided by adequate exposure, preoperative prophylactic antibiotics, and copious lavage of the abdominal cavity and the wound with large volumes of warm saline (usually with an antibiotic in the solution in children). In grossly contaminated (dirty) wounds, the use of drainage of the pelvis and wound, delayed primary skin closure, or the use of gauze pledgets impregnated with antiseptic may be used in an effort to reduce the risk of infection, but this is very rarely needed in children. Abscess formation can occur in the pelvis, right paracolic gutter, between loops of small bowel, or occasionally subphrenic space, usually from preexisting peritonitis. In children, approximately 1/3 of patients will have perforated within 24 h of the onset of symptoms. Damage to anatomical structures in the region rarely occurs in children, but the ilioinguinal or iliohypogastric nerves as they traverse close to the incision or the inferior epigastric vessels may be damaged. Damage to the ovary and right fallopian tube, however, is a well-recognized complication in the young female with peritonitis, either from sepsis or iatrogenically, and infertility may result. Right inguinal hernia and right femoral hernia are more common after appendectomy. Different techniques of dealing with the appendix stump can avoid complications associated

Table 3.20 Open appendectomy estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	5–20 %
Subcutaneous	5–20 %
Intra-abdominal/pelvic (peritonitis; abscess) (especially in the very young)	5–20 %
Systemic sepsis	2–5 %
Especially in the young	
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Small bowel obstruction (early or late)[a] [Adhesion formation]	1–5 %
Rare significant/serious problems	
Nerve parasthesia	0.1–1 %
Iliohypogastric/ilioinguinal nerve	
Inguinal hernia (right side)	0.1–1 %
Fallopian tube obstruction[a] (right; left very rare) – overall	0.1–1 %
After pelvic sepsis[b]	1–5 %
Female infertility[a] – overall[c]	<0.1 %
After pelvic sepsis[c, b]	0.1–1 %
Fecal fistula[a] (very rare)	<0.1 %
Ureteric injury (very rare)[a]	<0.1 %
Multisystem organ failure (renal, pulmonary, cardiac failure)	0.1–1 %
Mortality	<0.1 %
Mortality without surgery (should surgery be refused)	>80 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %
Nasogastric tube[a]	1–5 %
Wound drain tube(s)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences
[b]Fallopian tubal obstruction unilaterally may be as high as 8 %; lower bilaterally
[c]The rate of female infertility is related to the extent of perforation, abscess formation, and pelvic sepsis

with the stump including intraperitoneal abscess, "recurrent" appendicitis, and fecal fistula from breakdown of the wound closure of the cecum (stump abscess). Moreover, long-term complications of small bowel obstructions with adhesions, either to the appendix base or to the aperture of the appendix mesentery can occur. Inversion of the stump has been associated with increased risk of small bowel obstruction. Firm suture transfixion/ligation of the appendix base against the cecum usually avoids appendix stump complications.

Major Complications

Abscess formation, fistula or sinus formation, and **systemic sepsis** are serious complications that may rarely lead to **multisystem organ failure** and even mortality. Early surgery and preoperative antibiotics have reduced these complications significantly. Preexisting comorbidities include established generalized peritonitis and immunosuppression, which can increase risk of infection greatly. **Short-term failure to feed** may indicate **ongoing sepsis. Severe bleeding** is rare and transfusion uncommon. Concealed postoperative bleeding is rare. Persistent **wound sinuses** and a **fecal fistula** are very rare and require prolonged hospitalization and dressings, but most close within 2 months. **Prolonged ileus** and later (even decades later) **small bowel adhesive obstruction** can occur, but are surprisingly uncommon even with extensive adhesions. **Ureteric injury** and **iliac arterial injury** are exceedingly rare, but can be catastrophic. Scarring/adhesions from infection and inflammation can be associated with **fallopian tube obstruction**, tubal non-patency, ectopic or tubal pregnancy, ovarian or tubal torsion, infertility, and adhesional bowel obstruction, depending on the pathology and extent of sepsis.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (wound/abscess/systemic)*
- Risk of other abdominal organ injury*
- Intraperitoneal leakage
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Laparoscopic Appendectomy

Description

For those with reasonable experience in laparoscopy, appendicitis can be dealt with laparoscopically. General anesthesia is used. The patient is positioned in the supine position and is best examined when anesthetized to assess whether there is a mass to determine whether or not to proceed with the laparoscopy or whether to carry out an open procedure. Initial experience with laparoscopic appendectomy in the presence of an inflammatory mass was associated with a higher incidence of

postoperative ileus and intra-abdominal sepsis. That difference in outcome seems to be reducing as experience increases. Rectal examination under anesthesia may be useful to assess the presence of any pelvic mass. The objective of the operation is to remove the appendix. If the appendix is not inflamed, then the pelvis needs examination for pelvic pathology, particularly in the female, and the terminal ileum for the presence of a Meckel's diverticulum or other pathology causing local peritonitis. Occasionally, the inflammatory process, phlegmon, or abscess is so extensive the appendix cannot be removed, and it is judicious to convert to an open procedure. Drainage alone is very rarely used in children. If other pathology is encountered, determining whether to continue laparoscopically, will depend on the pathology. For example, Crohn's disease affecting the terminal ileum and cecum may necessitate open laparotomy.

Anatomical Points

The appendix origin lies at the confluence of the taenia coli; however, its tip can vary enormously in position, lying retrocecally (~75 % cases), pelvic (20 %), or retro-ileal/pre-ileal (5 %). The length of the appendix varies also and can reach the upper ascending colon posteriorly. Rarely the appendix (very rare) and cecum may enter a large inguinal hernial sac. Irritation of the bladder or colon can cause urinary urgency and/or diarrhea. An inflamed appendix, if retrocecal or pelvic in location, may irritate the ureter, so that hematuria or dysuria may occur. Irritation of the psoas muscle by an inflamed retrocecal appendix or abscess may cause hip discomfort on movement. Maldescent of the appendix is rare due to malrotation of the cecum, which remains high in the hepatic region. Agenesis, duplication, and situs inversus (left-sided appendix) are exceedingly rare, but can occur. There are, however, a series of patients with left atrial isomerism (picked up on antenatal scans) who are known to have situs inversus.

Perspective

See Table 3.21. Infective complications are the most common following appendectomy, wound infection being the most frequent. This is often avoided by adequate exposure, preoperative prophylactic antibiotics, and copious lavage of the abdominal cavity and the wound with large volumes of warm saline (usually with an antibiotic in the solution in children). In grossly contaminated (dirty) wounds, the use of drainage of the pelvis and wound, delayed primary skin closure, or the use of gauze pledgets impregnated with antiseptic may be used in an effort to reduce the risk of infection, but this is very rarely needed in children. Abscess formation can occur in the pelvis, right paracolic gutter, between loops of small bowel, or occasionally subphrenic space, usually from preexisting peritonitis. In children, approximately 1/3 of patients will have perforated within 24 h of the onset of symptoms. Damage to anatomical structures in the region rarely occurs in children, but the ilioinguinal or iliohypogastric nerves as they traverse close to the incision or the

Table 3.21 Laparoscopic appendectomy estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	5–20 %
Subcutaneous	5–20 %
Intra-abdominal/pelvic (peritonitis; abscess) (especially in the very young)	5–20 %
Systemic sepsis (especially in the young)	2–5 %
Hepatic portal sepsis (rare)	<0.1 %
Bleeding/hematoma formation[a]	
Wound	1–5 %
Intra-abdominal	0.1–1 %
Small bowel obstruction (early or late)[a] [Adhesion formation]	1–5 %
Conversion to open operation[a]	1–5 %
Extension of wound for access/safety (for improving exposure)[a]	1–5 %
Mortality	<0.1 %
Mortality without surgery (should surgery be refused)	>80 %
Rare significant/serious problems	
Nerve parasthesia	0.1–1 %
Iliohypogastric/ilioinguinal nerve	
Gas embolism[a]	<0.1 %
Ureteric injury[a]	<0.1 %
Vascular injury[a]	<0.1 %
Fecal fistula[a]	<0.1 %
Inguinal hernia (right side)	0.1–1 %
Multisystem failure (renal, pulmonary, cardiac failure)	0.1–1 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	1–5 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus (peritonitis from preoperative perforation)	20–30 %
Wound scarring (poor cosmesis/wound deformity)[a]	1–5 %
Nasogastric tube[a]	1–5 %
Wound drain tube(s)[a]	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

inferior epigastric vessels may be damaged. Damage to the ovary and right fallopian tube, however, is a well-recognized complication in the young female with peritonitis. Right inguinal hernia and right femoral hernia are more common after appendectomy. Different techniques of dealing with the appendix stump can avoid complications associated with the stump including intraperitoneal abscess, "recurrent" appendicitis, and fecal fistula from breakdown of the wound closure of the cecum (stump abscess). Moreover, long-term complications of small bowel obstructions with adhesions, either to the appendix base or to the aperture of the appendix mesentery can occur. Inversion of the stump has been associated with increased

risk of small bowel obstruction. Firm (double) loop ligation of the appendix base against the cecum usually avoids appendix stump complications. Conversion to open laparotomy may be required for a complicated appendix or other pathology. Complications of laparoscopy are relatively very rare, but include gas embolism, vascular or bowel trauma, surgical emphysema, and pneumothorax. Gas embolism is associated with Veress needle insertion, which can virtually be eliminated by open cutdown methods. Pneumothorax is a rare, idiosyncratic complication, probably from diaphragmatic leakage of gas.

Major Complications

Abscess formation, fistula or sinus formation, and **systemic sepsis** are serious complications that may rarely lead to **multisystem organ failure** and even mortality. Early surgery and preoperative antibiotics have reduced these complications significantly. Preexisting comorbidities include established generalized peritonitis and immunosuppression, which can increase the risk of infection greatly. **Short-term failure to feed** may indicate **ongoing sepsis. Severe bleeding** is rare and transfusion uncommon. Concealed postoperative bleeding is rare. Persistent **wound sinuses** and a **fecal fistula** are very rare and require prolonged hospitalization and dressings, but most close within 2 months. **Prolonged ileus** and later (even decades later) **small bowel adhesive obstruction** can occur, but are surprisingly uncommon even with extensive adhesions. **Gas embolism** is a very rare but catastrophic complication. **Ureteric injury** and **iliac arterial injury** are exceedingly rare but can be catastrophic. **Conversion to open operation** is a small but significant risk rather than a complication per se.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (wound/abscess/systemic)*
- Risk of other abdominal organ injury*
- Intraperitoneal leakage
- Gas embolism
- Conversion to open surgery
- Possible further surgery*
- Risks without surgery*

***Dependent on pathology, comorbidities, and surgery performed**

Surgery for Liver Tumors and Limited Liver Resection (Segmentectomy, Sectorectomy, and Sector Resection)

Description

General anesthesia is used. The aim of performing a segmental resection is typically to remove a solitary benign or malignant liver tumor, although several lesions may be amenable to segmental resection. Hepatoblastoma (45 %) and hepatocellular carcinoma (25 %) account for most primary malignant liver tumors. Hemangioendothelioma is the most common benign tumor. Although metastatic liver tumors are relatively rare in children, they do occur with Wilms' tumors of the kidney. They can be amenable to resection.

For any resection of a liver tumor, the goal is to achieve clear margins around the lesion(s), as well as excising any liver parenchyma, devascularized from occlusion of segmental portal inflow. Segmental resections can be combined with a contralateral major hepatectomy for complete resection of bilateral disease. Hemihepatectomy or lobectomy may be more appropriate than segmentectomy for livers in small children.

Anatomical Points

The anatomical variance in the performance of segmentectomy or sectorectomy is primarily dictated by the possible variant inflow that can occur with the right lobe of the liver. The right hepatic inflow that supplies segments 5, 6, 7, and 8 arises from the junction of the right and left portal vein. In a majority of cases, there is a common right portal vein that branches into the right anterior sectorial and right posterior sectorial branches. However, the main right portal vein leading to the right anterior and right posterior sectorial branches may be absent, instead originating at the same junction as the left portal vein. Another main portal anatomic variance can occur with the early take off of the right posterior sectorial vein, with the bifurcation then occurring at the left portal vein and the right anterior sectorial vein. Inflow within the right hepatic lobe can also vary with segment-6 inflow branches originating from the anterior sectorial branches and creating an isolated segment-7 branch. This anatomical variant is important to ensure that only a single segment of inflow is occluded instead of the entire right lobe or the anterior or posterior segment, respectively.

Perspective

See Table 3.22. Potential major complications of hepatic resection in children are **bleeding, hepatic failure from devascularized liver, and biliary leakage**. **Bleeding** during segmentectomy and sectorectomy primarily occurs from the outflow hepatic veins. These are thin-walled veins that tear easily and can develop

Table 3.22 Surgery for liver tumors estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection	
Wound	1–5 %
Intra-abdominal(including liver/liver bed/subphrenic abscess)	1–5 %
Intrathoracic (pneumonia; pleural)	5–20 %
Mediastinitis (if vena cava isolation used)	0.1–1 %
Systemic	1–5 %
Bleeding/hematoma formation overall	5–20 %
Arterial, venous (caval, renal, portal, hepatic, or lobar vessels)	1–5 %
Raw liver surface	1–5 %
Extrahepatic	1–5 %
Subcapsular hematoma[a] (major)	1–5 %
Serous ascitic collection	1–5 %
Bile duct ischemia	1–5 %
Bile duct stenosis	1–5 %
Biliary obstruction	5–20 %
Bile leak/collection	5–20 %
Biliary ascites	1–5 %
Biliary fistula	1–5 %
Hyperbilirubinemia	50–80 %
Jaundice	1–5 %
Common/extrahepatic/intrahepatic bile duct injury	1–5 %
Unresectability of malignancy or tumor/involved resection margins[a]	Individual
Recurrence of malignancy[a]	Individual
Bowel injury (stomach, duodenum, small bowel, colon)	1–5 %
Thrombosis	
Arterial (hepatic)	1–5 %
Venous (hepatic)	1–5 %
Liver failure (ischemia; toxicity; acute hepatic necrosis) early or late	5–20 %
Liver injury (to remaining liver)	1–5 %
Surgical emphysema[a] (major)	1–5 %
Gastrointestinal erosion, ulceration, perforation, hemorrhage	1–5 %
Small bowel obstruction (early or late)[a] [Ischemic stenosis/adhesion formation]	1–5 %
Reflux esophagitis/pharyngitis/pneumonitis	1–5 %
Coagulopathy	1–5 %
Disseminated intravascular coagulopathy	
[a]Consumption transfusion (large bleed)	
Pericardial effusion	1–5 %
Muscle weakness (atrophy due to denervation esp subcostal incision)	1–5 %
Nutritional deficiency – anemia, B12 malabsorption[a]	5–20 %
Multisystem failure (renal, pulmonary, cardiac failure)	1–5 %
Mortality[a]	1–5 %
Mortality without surgery[a] (for hepatoblastoma virtually 100 %)	>80 %

(continued)

Table 3.22 (continued)

Complications, risks, and consequences	Estimated frequency
Rare significant/serious problems	
Aspiration pneumonitis	0.1–1 %
Portal venous thrombosis[a]	0.1–1 %
Deep venous thrombosis	0.1–1 %
Air embolus (major)	0.1–1 %
Renal/adrenal injury renal vein	0.1–1 %
Diaphragmatic injury paresis	0.1–1 %
Diaphragmatic hernia	0.1–1 %
Pancreatic injury/pancreatitis/pancreatic cyst/pancreatic fistula	0.1–1 %
Thoracic duct injury (chylous leak, fistula)[a]	0.1–1 %
Budd-Chiari (acute)	0.1–1 %
Splenic injury (conservation (consequent limitation to activity; late rupture) or splenectomy)	0.1–1 %
Hepatitis (drug, CMV, recurrent)[a]	0.1–1 %
Renal failure (hepatorenal syndrome)[a]	0.1–1 %
Hyperglycemia	0.1–1 %
Hypoglycemia	0.1–1 %
Wound dehiscence	0.1–1 %
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Incisional hernia formation (delayed heavy lifting)	1–5 %
Paralytic ileus	20–50 %
Nasogastric tube[a]	1–5 %
Blood transfusion[a]	5–20 %
Wound drain tube(s)[a]	Individual
Wound scarring (poor cosmesis/wound deformity)	1–5 %

[a]Dependent on underlying pathology, anatomy, surgical technique, and preferences

lateral tears, which can extend up to the main venous branches or to the inferior vena cava underneath intact hepatic parenchyma. Thus, any form of segmentectomy or sectorial resection must identify all of the major hepatic venous outflow structures to ensure adequate hemostasis and to minimize blood loss. **Biliary leakage** is primarily a problem in patients who are undergoing some form of bile duct resection and require biliary reconstruction. Biliary leakage is less common when performing a segmentectomy or sectorectomy and primarily will occur because of the inadvertent transection of a (small) bile duct without adequate closure. The performance of a segmentectomy should not be automatically assumed to be a lesser operative procedure compared to hepatic lobectomy or some form of extended hepatic lobectomy. There have been recent evaluations showing that intraoperative blood loss is significantly greater when a segmentectomy is performed, compared to an anatomic hepatic lobectomy. The reason for this is principally difficulty with small venous outflow control that may lead to increased blood loss during the

resection phase. All segmental resections are not of similar difficulty. A segment-3 resection is technically far easier, compared to a segment-8 resection. The anatomical variation, the depth of the liver parenchyma, and the patient body habitus can make various types of segmental resections more difficult in certain patients.

Major Complications

Intraoperative bleeding during a segmentectomy can be the most serious complication because of injury without ligation of the outflow hepatic veins. If the central venous pressure (CVP) is not low, the risk of intraoperative hemorrhage is increased. Hepatic venous outflow hemorrhage from inadvertent transection of the hepatic veins primarily occurs because of the inability to identify anatomic variations intraoperatively. **Intraoperative air embolus** can be a severe and life-threatening complication due to inadvertent laceration of the hepatic veins during hepatic parenchymal transection, with aspiration of air into the vena cava. This complication can be related to low CVP while parenchymal transection is performed. Hence, optimal controlled hypotension with a CVP of 0–1 cm H_2O has been proven to be the most effective anesthetic management in patients who undergo any form of hepatic resection. The acute management of a patient who has sustained an air embolus is immediate steep Trendelenburg (head-down) position, with occlusion of the parenchymal transection site with a wet laparotomy gauze pack and aggressive support measures by the anesthetist. This complication can be effectively prevented by ensuring identification and proper ligation of all hepatic venous branches prior to transection. **Postoperative bile leakage** after a segmentectomy or sectorectomy can also lead to significant morbidity, depending on the extent of injury. A recent prospective randomized controlled trial has shown that intraoperative drains placed in patients undergoing hepatic resections do not lead to decreased perioperative morbidity or lessen the need for subsequent postoperative drainage. Thus, meticulous intraoperative hemostasis, as well as identification and ligation of all bile ducts during hepatic transection cannot be overemphasized. Omentoplasty has been utilized to prevent bile leakage after resection; however, in a recent prospective randomized controlled trial, this technique was not found to significantly reduce bile leakage. Further review of this report also showed that omentoplasty did not adversely affect the patient either; thus, the utilization should be surgeon determined. The vast majority of patients who sustain a postoperative bile leak either resolve spontaneously or can be managed with a percutaneous drainage and bedside supportive care.

Devascularization: In the small child with a hepatoblastoma, the vessels are small so that small segmental arteries may go into vasospasm, or they can be inadvertently injured in the porta hepatis, even using bipolar diathermy. This can even be seen to the residual arterial supply after major hepatic resection, and especially so with the Kasai procedure, when the patient is only weeks old, where extensive dissection in the porta hepatis is paramount to get an extended resection of the hepatic plate. So, all of the arterial supply is at risk.

Tumor recurrence following resection of a hepatic malignancy is a significant problem and is integrally related to the width of resection margin of normal liver around the lesions(s). Tumor rupture may occur preoperatively or intraoperatively especially with large, necrotic tumors. Tumor leakage may disseminate tumor and worsen the prognosis. Development of further tumor metastases after metastasectomy arising from previously subclinical micrometastases is another limitation to successful surgical treatment. Although these are not strictly complications per se, they are contingent on effective preoperative evaluation and surgical technique. **Infection** of the wound or peritoneal cavity (**peritonitis**, **abscess**) can lead to sepsis and **multisystem organ failure** and significant **mortality**. Respiratory and cardiac complications are moderately common and are increased if anomalies are present. **Liver failure** can result from a long operative ischemic time. **Recurrence of tumor** depends heavily on the pathology and further metastasis. If a hepatoblastoma is not fully resected, then death is almost inevitable despite aggressive chemotherapy. In those circumstances, if the tumor is thought to be unresectable, it may be better to plan a liver transplant, as metastases do not appear to occur early when the child is having chemotherapy. At the initial presentation, hepatoblastomas often look unresectable, but with chemotherapy, they may become resectable with conventional techniques.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Pain/discomfort
- Bleeding/hematoma*
- Infection (local/systemic)*
- Risk of other abdominal organ injury*
- Intraperitoneal leakage
- Air embolism
- Possible further surgery*
- Recurrence of tumor
- Risks without surgery*
- Multisystem organ failure
- Death

***Dependent on pathology, comorbidities, and surgery performed**

Open Splenectomy

Description

General anesthesia is used. The aim is to electively remove the spleen, including any small remnant splenunculi, which may be separate from the main splenic mass.

The degree of difficulty and relative risk of complications is proportional to the size of the spleen and underlying disease process. A midline, left subcostal, or left transverse upper abdominal incision is usual. The spleen is mobilized on its pedicle, freeing any adhesions with the abdominal wall or organs. The short gastric vessels often require division specifically. Occasionally, the splenic flexure of the colon overlies the spleen and needs to be "taken down" to permit better exposure. In general, the spleen is freed from attachments to surrounding structures and is then lifted and rotated anteromedially toward the right side to expose the splenic hilum from behind. A clamp(s) is then placed across the splenic pedicle, divided then ligated, taking care not to injure the pancreatic tail. Individual vessels can be identified easily in children, so each artery can be ligated, followed by the veins. The spleen can then be delivered through the abdominal incision and removed. The pedicle and splenic bed are checked and hemostasis is achieved. A drain may be used, more for possible pancreatic leakage than bleeding. Accessory splenic tissue is sought (splenunculi), particularly in patients with idiopathic thrombocytopenic purpura (ITP), and removed as each can grow to a degree that allows disease recurrence. Removal of the spleen for severe trauma or surgical rupture carries a different spectrum of risk, should conservative management not be possible or appropriate. In children a standard layered closure with absorbable sutures is usually used.

Anatomical Points

The spleen may be lobulated as a normal variant, as embryologically, it forms from fusion of individual lobules. A fissure may even occur with two or more separate lobes or separate spleens. Small, usually rounded, deposits of splenic tissue may exist as splenunculi, often around the splenic hilum, vessels within the lesser sac, or omentum. The short gastric vessels may be closely applied to the spleen, making division difficult and the risk of injury greater especially at the upper pole. Adhesions to the anterior, lateral, or posterior abdominal wall, diaphragm, or bowel may occur. The splenic flexure of the colon may be tethered to the spleen or above to the lateral abdominal wall, reducing access. The tail of pancreas may overlie the splenic hilum where it is at risk of injury and may also impede access. The splenic vessels may be multiple and widely separated, requiring several individual ligations. The kidney is usually easily separated from the spleen, but can be adherent on occasions, particularly with malignant involvement or severe inflammatory processes. An enlarged spleen can migrate toward the right iliac fossa and render access to the hilum and delivery difficult.

Perspective

See Table 3.23. Splenectomy can be an elective procedure (eg. small spleen in ITP, or a moderate-sized to massive spleen for hematological disorders or metastatic malignancy) or an acute procedure (eg. hemorrhage from splenic trauma in a shocked patient). Note that conservative management of splenic trauma is the norm for children, and laparotomy is rarely used for isolated splenic trauma.

Table 3.23 Open splenectomy estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection[a]	
Subcutaneous	1–5 %
Wound	1–5 %
Intra-abdominal	0.1–1 %
Chest infection	1–5 %
Subphrenic abscess[a]	0.1–1 %
Late – overwhelming post-splenectomy sepsis (with vaccination) lifetime risk	5–10 %
Mortality from post-splenectomy sepsis	1–5 %
Bleeding/hematoma formation[b]	0.1–1 %
Small bowel obstruction (early or late)[a]	1–5 %
Excessive NG losses (usually lasting no more than 5 days)	5–20 %
Rare significant/serious problems	
Pancreatic injury/pancreatitis/pancreatic cyst/leakage/pancreatic fistula	0.1–1 %
Bowel injury (stomach, duodenum, small bowel, colon)[b]	0.1–1 %
Renal/adrenal injury[a]	0.1–1 %
Diaphragmatic injury[a]	0.1–1 %
Accessory spleen formation[b] (Mainly ITP; trauma)	0.1–1 %
Mortality without surgery[c]	Variable
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus[b]	5–20 %
Incisional hernia (avoid lifting/straining for 8/52)	0.1–1 %
Wound scarring (deformity/dimpling of wound scar/poor cosmesis)	1–5 %
Drain tube(s)[a]	5–20 %

[a]Dependent on underlying pathology, surgical technique preferences, and location on the body
[b]Incidence may be higher for moderate and massive splenomegaly
[c]Depending on the underlying disease; much higher for malignancy. NB: The spectrum of risk following trauma is different

The degree of difficulty can vary markedly between these situations as can the risks and complications. Spill of splenic tissue (e.g., with rupture) can lead to recurrent ITP or malignancy, depending on the initial pathology and situation. Infection is more common in malignant conditions, in immunocompromised individuals, and after multi-trauma, especially if preexisting or concurrent lung trauma or infection is present. Infection of a hematoma in the splenic bed may result in a subphrenic abscess. Inadvertent injury to the bowel or pancreas may result in infection or a fistula, which can be chronic and debilitating, with slow closure. Overwhelming post-splenectomy pneumonia or sepsis carries a lifetime risk of up to 2–10 % of those that have a hematological reason for the

splenectomy and 1 % of those after trauma. Vaccination does not give adequate protection for all strains of pathogens, especially pneumococcus, but may be effective for hemophilus, so that "overwhelming post-splenectomy infection (OPSI)" still occurs. So, parent/patient must be educated to seek medical advice immediately the baby/child/adult appears to be mildly infected. Often, an early megadose of penicillin is very effective. Paralytic ileus is common but usually resolves spontaneously within a week. Injury to the adrenal and kidney is very rare. Excessive nasogastric loses frequently occur for several days after surgery, possibly as a result of division of the short gastric vessels.

Major Complications

Respiratory infection is perhaps the most common complication and may lead to lobar **pneumonia** and **severe systemic sepsis**. **Bleeding** and ongoing **oozing** can be significant, especially in patients with coagulopathies; however, hemostasis at surgery usually controls this adequately. **Recurrent ITP** can be a problem, if ITP was the reason for splenectomy, and may require intraoperative nuclear scans and further surgery. **Recurrent malignancy** can also occur, if tumor or blood spill occurs during splenectomy for malignancy. The use of a plastic bag around the spleen before splenic ligation may reduce the risk of both forms of recurrence. **Wound infection, peritonitis,** and **intra-abdominal abscess formation** may predispose to **wound dehiscence** and even "burst" abdomen. **Pancreatic leakage**, **pseudocyst formation,** and **fistula formation** are relatively uncommon, but can be very debilitating. **Small bowel obstruction** is not uncommon even years after surgery. **Overwhelming post-splenectomy sepsis** is a rare, potentially serious later complication.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Bleeding/hematoma
- Infection (local/systemic)
- Pain/discomfort
- Possible tumor recurrence*
- Other abdominal organ injury
- Respiratory complications
- Venous thromboembolism
- Possible blood transfusion
- Risks without surgery

***Dependent on pathology and type of surgery performed**

Laparoscopic Splenectomy

Description

General anesthesia is used. The aim is to remove the spleen, using laparoscopic techniques, including any small remnant splenunculi, which may be separate from the main splenic mass. The degree of difficulty and relative risk of complications is proportional to the size of the spleen and underlying disease process. A range of patient positions have been used, including the right lateral decubitus, but the usual to have the left sided elevated to 30°–45°. Ports are then placed and the procedure performed. A small midline, left subcostal, or left transverse upper abdominal incision may be used in conjunction for larger or difficult spleens. The spleen is mobilized on its pedicle, freeing any adhesions with the abdominal wall or organs. The short gastric vessels will often require division specifically. Occasionally, the splenic flexure of the colon overlies the spleen and needs to be "taken down" to permit better exposure. In general, the spleen is freed from attachments to surrounding structures and is then lifted laterally to the left side to expose the splenic hilum from in front. A vascular stapling device(s) is then placed across the splenic pedicle, which ligated and divided, taking care not to injure the pancreatic tail. The spleen can then be placed in a plastic bag, delivered through an abdominal incision, or morcellated (minced) and removed. The pedicle and splenic bed are checked and hemostasis is achieved. A drain may be used, more for possible pancreatic leakage than bleeding. Accessory splenic tissue is sought, particularly in idiopathic thrombocytopenic purpura (ITP) cases, and removed. Port-site and abdominal closure is usually performed.

Anatomical Points

The spleen may be lobulated as a normal variant, as embryologically, it forms from fusion of individual lobules. A fissure may even occur with two or more separate lobes, or separate spleens. Small, usually rounded, deposits of splenic tissue may exist as splenunculi, often around the splenic hilum, vessels within the lesser sac, or omentum. The short gastric vessels may be closely applied to the spleen, making division difficult and the risk of injury greater. Adhesions to the anterior, lateral, or posterior abdominal wall, diaphragm, or bowel may occur. The splenic flexure of the colon may be tethered to the spleen or above to the lateral abdominal wall, reducing access. The tail of pancreas may overlie the splenic hilum where it is at risk of injury and may also impede access. The splenic vessels may be multiple and widely separated, requiring several individual ligations. The kidney is usually easily separated from the spleen, but can be adherent on occasions, particularly with malignant involvement or severe inflammatory processes. An enlarged spleen can migrate toward the right iliac fossa and render access to the hilum and delivery difficult.

Table 3.24 Laparoscopic splenectomy estimated frequency of complications, risks, and consequences

Complications, risks, and consequences	Estimated frequency
Most significant/serious complications	
Infection	
Subcutaneous	1–5 %
Wound	1–5 %
Intra-abdominal	0.1–1 %
Late – post-splenectomy sepsis (with vaccination)	0.1–1 %
Chest infection	1–5 %
Subphrenic abscess[a]	0.1–1 %
Late – overwhelming post-splenectomy sepsis (with vaccination) lifetime risk	5–10 %
Mortality from post-splenectomy sepsis	1–5 %
Bleeding/hematoma formation[b]	0.1–1 %
Small bowel obstruction (early or late)[a]	1–5 %
Excessive NG losses (usually lasting no more than 5 days)	5–20 %
Conversion to open surgical procedure[b]	1–5 %
Rare significant/serious problems	
Gas embolus	0.1–1 %
Pancreatic injury/pancreatitis/pancreatic cyst/leakage/pancreatic fistula	0.1–1 %
Bowel injury (stomach, duodenum, small bowel, colon)	0.1–1 %
Renal/adrenal injury	0.1–1 %
Diaphragmatic injury	0.1–1 %
Small bowel obstruction (early or late)	0.1–1 %
Subphrenic abscess	0.1–1 %
Accessory spleen formation[b]	0.1–1 %
(mainly ITP trauma)	
Mortality without surgery[c]	Variable
Less serious complications	
Pain/tenderness	
Acute (<4 weeks)	5–20 %
Chronic (>4 weeks)	<0.1 %
Paralytic ileus[b]	5–20 %
Port-site hernia (s) (avoid lifting/straining)	0.1–1 %
Wound scarring (deformity/dimpling of wound scar/poor cosmesis)	1–5 %
Drain tube(s)[a]	5–20 %

[a]Dependent on underlying pathology, surgical technique preferences, and location on the body
[b]Incidence may be higher for moderate and massive splenomegaly
[c]Depending on the underlying disease; much higher for malignancy

Perspective

See Table 3.24. Splenectomy can range from an elective procedure with a small spleen in ITP, for a moderate-sized spleen, or massive spleen for hematological disorders or metastatic malignancy to an acute procedure for hemorrhage from

splenic trauma in a shocked patient. But, open surgery is more likely to be used for massive splenic trauma, where there may be liver damage as well. Note that conservative management of splenic trauma is the norm for children, and laparotomy is rarely used for isolated splenic trauma. The degree of difficulty can vary markedly between these situations as can the risks and complications. Spill of splenic tissue (e.g., with rupture) can lead to recurrent ITP or malignancy, depending on the initial pathology and situation. Infection is more common in malignant conditions, in immunocompromised individuals, and after multitrauma, especially if preexisting or concurrent lung trauma or infection is present. Infection of a hematoma in the splenic bed may result in a subphrenic abscess. Inadvertent injury to the bowel or pancreas may result in infection or a fistula, which can be chronic and debilitating, with slow closure. Overwhelming post-splenectomy pneumonia or sepsis carries a lifetime risk of up to 2 % of those that have a hematological reason for the splenectomy and 1 % of those after trauma. Vaccination does not give adequate protection for all strains of pathogens, especially pneumococcus, but may be effective for hemophilus, so that "overwhelming post-splenectomy infection (OPSI)" still occurs. So, parent/patient must be educated to seek medical advice immediately the baby/child/adult appears to be mildly infected. Often, an early megadose of penicillin is very effective. Paralytic ileus is common but usually resolves spontaneously within a week. Injury to the adrenal and kidney is very rare. Excessive nasogastric loses frequently occur for several days after surgery, possibly as a result of division of the short gastric vessels.

Major Complications

Respiratory infection is perhaps the most common complication and may lead to lobar **pneumonia** and **severe systemic sepsis**. **Bleeding** and ongoing **oozing** can be significant, especially in patients with coagulopathies; however, hemostasis at surgery usually controls this adequately. **Recurrent ITP** can be a problem, if ITP was the reason for splenectomy, and may require intraoperative nuclear scans and further surgery. **Recurrent malignancy** can also occur, if tumor or blood spill occurs during splenectomy for malignancy. The use of a plastic bag around the spleen before splenic ligation may reduce the risk of both forms of recurrence. **Wound infection, peritonitis,** and **intra-abdominal abscess formation** may predispose to **wound dehiscence** and even "burst" abdomen. **Pancreatic leakage**, **pseudocyst formation,** and **fistula formation** are relatively uncommon, but can be very debilitating. **Small bowel obstruction** is not uncommon even years after surgery. **Overwhelming post-splenectomy sepsis** is a rare, potentially serious later complication. Laparoscopic **injury to bowel or blood vessels** is uncommon, although greater than for open splenectomy, and **gas embolism** although potentially catastrophic is very rare.

Consent and Risk Reduction

Main Points to Explain

- GA risk
- Bleeding/hematoma
- Infection (local/systemic)
- Pain/discomfort
- Possible tumor recurrence*
- Other abdominal organ injury
- Respiratory complications
- Venous thromboembolism
- Possible blood transfusion
- Gas embolism
- Possible open operation
- Risks without surgery

***Dependent on pathology and type of surgery performed**

Further Reading, References, and Resources

General Reading[1] [2]

Ellis H. Clinical anatomy. 11th ed. Blackwell; Oxford, 2006.
Jamieson GG, editor. The anatomy of general surgical operations. Edinburgh: Churchill-Livingstone Elsevier; 2006. ISBN 9780443100079.
Moore KL. The developing human: clinically oriented embryology. 2nd ed. Philadelphia: WB Saunders; 1977. ISBN 0-7216-6471-7.
Scott-Conner CEH, Dawson DL. Operative anatomy. 3rd ed. Philadelphia: Lippincott Williams & Wilkins; 2008. ISBN 97807817653.
Skandalakis LJ, Skandalakis JE, Skandalakis 3rd PN, editors. Surgical anatomy and technique: a pocket manual. 3rd ed. New York: Springer; 2009. ISBN 9780387095141.
Ashcraft KW, Holcomb GW, Murphy JP, editors. Ashcraft's pediatric surgery. 5th ed. Philadelphia: Saunders/Elsevier; 2009. ISBN 9781416061274.
Caty MG. Complications in pediatric surgery. New York: Informa Healthcare; 2008. ISBN 9780824728366.
Grant HW, Chuturgoon AA, Kenoyer DG, Doorasamy T. The adaptive immune response to major surgery in the neonate. Pediatr Surg Int. 1997;12(7):490–3.

[1] Embryology and Operative Anatomy

[2] Operative Surgery

Grosfeld JL, editor. Pediatric surgery. 6th ed. Philadelphia: Elsevier Science; 2006. ISBN 9780323028424.
Harmon CM, Coran AG. Congenital anomalies of the esophagus, associated anomalies, Ch 62. In: O'Neill Jr JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG, editors. Pediatric surgery. 5th ed. St Louis: Mosby Press; 1998. p. 945–6.
Hutson JM, O'Brien M, Woodward AA, Beasley SW. Clinical paediatric surgery: diagnosis & management. 6th ed. Melbourne: Wiley-Blackwell; 2008. ISBN 978-1405162678.
O'Neill Jr JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG, editors. Pediatric surgery. 5th ed. St Louis: Mosby Press; 1998.
Spitz L, Coran AG, editors. Operative pediatric surgery. 6th ed. London: Hodder-Arnold; 2006.
Puri P, editor. Newborn surgery. 2nd ed. London: Arnold; 2003. ISBN 9780340761441.
Rowe MI. The newborn as a surgical patient, Ch 4. In: O'Neill Jr JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG, editors. Pediatric surgery. 5th ed. St Louis: Mosby Press; 1998. p. 43–70.
Stringer MD, Oldham KT, Moriguard PDE. Pediatric surgery and urology. 2nd ed. Cambridge: Cambridge University Press; 2006a. ISBN 9780521839020.
Stringer MD, Oldham KT, Mouriquand PDE. Pediatric surgery and urology: long-term outcomes. Cambridge: Cambridge University Press; 2006b.
Welch KJ, Randolph JG, Ravitch MM, et al., editors. Pediatric surgery. 5th ed. Chicago: Year Book Medical; 1998.

More Specific Reading [3] [4] [5] [6] [7] [8] [9] [10]

van Eijck FC, Wijnen RM, van Goor HJ. The incidence and morbidity of adhesions after treatment of neonates with gastroschisis and omphalocele: a 30-year review. Pediatr Surg. 2008;43(3):479–83.
Albanese CT, Rowe MI. Necrotizing enterocolitis, Ch 87. In: O'Neill Jr JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG, editors. Pediatric surgery. 5th ed. St Louis: Mosby Press; 1998. p. 1297–320.
Boix-Ochoa J, Rowe MI. Gastroesophageal reflux, Ch 66. In: O'Neill Jr JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG, editors. Pediatric surgery. 5th ed. St Louis: Mosby Press; 1998. p. 1007–17.
Chacko J, Ford WDA, Haslam R. Neurological developmental outcome in very low birth weight infants after laparotomy. Pediatr Surg Int. 1999;15:496–9.
Cole C, Freitas A, Clifton MS, Durham MM. Hereditary multiple intestinal atresias: 2 new cases and review of the literature. J Pediatr Surg. 2010;45(4):E21–4.
Dalla Vecchia LK, Grosfeld JL, West KW, Rescorla FJ, Scherer LR, Engum SA. Intestinal atresia and stenosis: a 25-year experience with 277 cases. Arch Surg. 1998;133(5):490–6; discussion 496–7. Review.

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[3] Gastroschisis and Omphalocele

[4] Atresia and Gut Obstructions

[5] Congenital Diaphragmatic Hernia

[6] Hirschsprung's and Anorectal Malformations

[7] Fundoplications

[8] Hernia Repair

[9] Gastrotomy

[10] Splenectomy

Efrati O, Nir J, Fraser D, Cohen-Cymberknoh M, Shoseyov D, Vilozni D, Modan-Moses D, Levy R, Szeinberg A, Kerem E, Rivlin J. Meconium ileus in patients with cystic fibrosis is not a risk factor for clinical deterioration and survival: the Israeli multicenter study. J Pediatr Gastroenterol Nutr. 2010;50(2):173–8.

Federici S, Domenichelli V, Antonellini C, Dòmini R. Multiple intestinal atresia with apple peel syndrome: successful treatment by five end-to-end anastomoses, jejunostomy, and transanastomotic silicone stent. J Pediatr Surg. 2003;38(8):1250–2.

Khong TY, Ford WDA, Haan EA. Umbilical cord ulceration in association with intestinal atresia in a child with deletion 13q and Hirschsprung's disease. Arch Dis Child. 1994;71:F212–3.

Messina M, Molinaro F, Ferrara F, Messina G, Di Maggio G. Idiopathic spontaneous intestinal perforation: a distinct pathological entity in the preterm infant. Minerva Pediatr. 2009;61(4): 355–60.

Murphy FL, Sparnon AL. Long-term complications following intestinal malrotation and the Ladd's procedure: a 15 year review. Pediatr Surg Int. 2006;22(4):326–9.

Ooi BC, Barnes GL, Tauro GP. Normalization of vitamin B12 absorption after ileal resection in children. J Paediatr Child Health. 1992;28(2):168–71.

Pueyo C, Maldonado J, Royo Y, Skrabski R, Di Crosta I, Raventós A. Intrauterine intussusception: a rare cause of intestinal atresia. J Pediatr Surg. 2009;44(10):2028–30.

Rausin L, Khamis J, Paquot JP, Langhendries JP. Obstruction of unusual origin in a small preterm baby-girl. J Belge Radiol. 1992;75(5):402–3.

Rescorla FJ. Meconium ileus, Ch 75. In: O'Neill Jr JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG, editors. Pediatric surgery. 5th ed. St Louis: Mosby Press; 1998. p. 1159–72.

Stringer MD, Oldham KT, Moriguard PDE. Pediatric surgery and urology. 2nd ed. Cambridge: Cambridge University Press; 2006. ISBN 9780521839020.

Shah SR, Gittes GK, Barsness KA, Kane TD. Multimedia article. Thoracoscopic patch repair of a right-sided congenital diaphragmatic hernia in a neonate. Surg Endosc. 2009;23(1):215.

Zwaveling S, van der Zee DC. Laparoscopic repair of an isolated congenital bilateral lumbar hernia in an infant. Eur J Pediatr Surg. 2012;22(4):321–3.

Holschneider AM, Hutson JM. Anorectal malformations in children: embryology, diagnosis, surgical treatment, follow-up. Berlin: Springer; 2006. ISBN 9783540317500.

Holschneider AM, Puri P. Hirschsprung's disease and allied disorders. Dordrecht: Springer; 2007. ISBN 9783540339342.

Ieiri S, Higashi M, Teshiba R, Saeki I, Esumi G, Akiyoshi J, Nakatsuji T, Taguchi T. Clinical features of Hirschsprung's disease associated with Down syndrome: a 30-year retrospective nationwide survey in Japan. J Pediatr Surg. 2009;44(12):2347–51.

Levitt MA, Peña A. Operative management of anomalies in female. In: Holschneider AM, Hutson J, editors. Anorectal malformations in children. Heidelberg: Springer; 2006a. p. 303–6.

Levitt MA, Peña A. Treatment of cloacas. In: Holschneider AM, Hutson J, editors. Anorectal malformations in children. Heidelberg: Springer; 2006b. p. 307–14.

Levitt MA, Pena A. Anorectal malformations. Rev Orphanet J Rare Dis. 2007; 2:33. doi:10.1186/1750-1172-2-33.

Nievelstein RA, Vos A, Valk J. MR imaging of anorectal malformations and associated anomalies. Eur Radiol. 1998;8(4):573–81.

Pena A, Levitt MA, Hong A, Midulla P. Surgical management of cloacal malformations: a review of 339 patients. J Pediatr Surg. 2004;39(3):470–9.

Pena A, Migotto-Krieger M, Levitt MA. Colostomy in anorectal malformations: a procedure with serious but preventable complications. J Pediatr Surg. 2006;41(4):748–56. Discussion 748-56.

Tei E, Yamataka A, Segawa O, Kobayashi H, Lane GJ, Tobayama S, Kameoka S, Miyano T. Laparoscopically assisted anorectovaginoplasty for selected types of female anorectal malformations. J Pediatr Surg. 2003;38(12):1770–4.

Teitelbaum DH, Coran AG, Weitzman JJ, Ziegler MM, Kane T. Hirschsprung's disease and related neuromuscular disorders of the intestine, Ch 94. In: O'Neill Jr JA, Rowe MI, Grosfeld JL,

Fonkalsrud EW, Coran AG, editors. Pediatric surgery. 5th ed. St Louis: Mosby Press; 1998. p. 1391–3.

Barsness KA, St Peter SD, Holcomb 3rd GW, Ostlie DJ, Kane TD. Laparoscopic fundoplication after previous open abdominal operations in infants and children. J Laparoendosc Adv Surg Tech A. 2009;19 Suppl 1:S47–9. 2009 Apr.

Lopez M, Kalfa N, Forgues D, Guibal MP, Galifer RB, Allal H. Laparoscopic redo fundoplication in children: failure causes and feasibility. J Pediatr Surg. 2008;43(10):1885–90.

Mauritz FA, van Herwaarden-Lindeboom MY, Stomp W, Zwaveling S, Fischer K, Houwen RH, Siersema PD, van der Zee DC. The effects and efficacy of antireflux surgery in children with gastroesophageal reflux disease: a systematic review. J Gastrointest Surg. 2011;15(10): 1872–8.

Rothenberg SS, Chin A. Laparoscopic Collis-Nissen for recurrent severe reflux in pediatric patients with esophageal atresia and recurrent hiatal hernia. J Laparoendosc Adv Surg Tech. 2010;20:787–90.

Tovar JA, Luis AL, Encinas JL, Burgos L, Pederiva F, Martinez L, Olivares P. Pediatric surgeons and gastroesophageal reflux. J Pediatr Surg. 2007;42(2):277–83.

Treef W, Schier F. Characteristics of laparoscopic inguinal hernia recurrences. Pediatr Surg Int. 2009;25(2):149–52.

Fraser JD, Ponsky TA, Aguayo P, Boulanger S, Parry R, Nixdorf N, DiLuciano J, Smith P, Sharp SW, Holcomb GW, Ostlie DJ, St Peter SD. Short-term natural history of the standard approaches for gastrostomy tube placement in the pediatric patient. J Laparoendosc Adv Surg Tech A. 2009;19(4):567–9.

Naiditch JA, Lautz T, Barsness KA. Postoperative complications in children undergoing gastrostomy tube placement. J Laparoendosc Adv Surg Tech. 2010;20:781–5.

Virnig DJ, Frech EJ, Delegge MH, Fang JC. Direct percutaneous endoscopic jejunostomy: a case series in pediatric patients. Gastrointest Endosc. 2008;67(6):984–7.

Coventry BJ, Watson DI, Tucker K, Chatterton B, Suppiah R. Intraoperative scintigraphic localization and laparoscopic excision of accessory splenic tissue. Surg Endosc. 1998;12(2):159–61.

Kollias J, Watson DI, Coventry BJ, Malycha P. Laparoscopic splenectomy using the lateral position: an improved technique. Aust N Z J Surg. 1995;65(10):746–8.

Watson DI, Coventry BJ, Chin T, Gill PG, Malycha P. Laparoscopic versus open splenectomy for immune thrombocytopenic purpura. Surgery. 1997;121(1):18–22.