Essential Pediatric Surgery

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Dan Poenaru, Emmanuel A. Ameh, Arlene Muzira, and Doruk Ozgediz

Principles of Children's Surgery in Low-Resource Settings

The Essentials

- Burden of pediatric surgical disease in LMICs is large, the spectrum is wide, and presentation is often late.
- Appropriate size instruments and equipment and support services, including intensive and critical care, are limited.
- Prior planning in conjunction with the host surgeon is critical to success.

Burden of Pediatric Surgical Disease

Pediatric surgical conditions account for a significant portion of the burden of surgical disease in low- and middle-income countries (LMICs). This is partially due to regional demographics as more than half of the population is < 18 years

D. Poenaru, MD, MHPE, PhD (🖂)

Department of Surgery, Montreal Children's Hospital, 1001 Decarie Boulevard, Room B04.2022, Montreal, QC h4A3J1, Canada e-mail: dpoenaru@gmail.com

E.A. Ameh, MBBS, FWACS Division of Paediatric Surgery, Department of Surgery, National Hospital, Central District, Federal Capital Territory, Abuja 900001, Nigeria e-mail: eaameh@yahoo.co.uk

A. Muzira, MBChB, MMed (Surg) Department of Surgery, Mulago Hospital, Mulago Hospital Complex, Kampala, Uganda e-mail: arlenemzr@gmail.com

D. Ozgediz, MD, MSc Department of Surgery, Yale New Haven Hospital, 333 Cedar Street, New Haven, CT 06520, USA e-mail: doruk.ozgediz@yale.edu old in many countries, and fertility rates remain as high as seven per family, even in countries with limited life expectancies [1]. Prospective studies suggest that up to 85% of children in these settings will require surgical intervention by age 15 and that 40% of surgical unmet need at the population level is in children [2, 3].

Features of Pediatric Surgery in LMICs

The surgeon who comes to assist in a LMIC quickly notices that the pediatric surgical practice in this setting is radically different from what he/she had been exposed to in high-income settings. Pediatric surgery in LMICs is first characterized by a very wide spectrum of illness comprising all the pediatric subspecialties. The children frequently present late for care, sometimes in the teenage years or even adulthood, and the disease processes, especially in the case of tumors or colorectal disease, are very advanced. Many "classical" index conditions in Western pediatric surgery, such as congenital diaphragmatic hernia or necrotizing enterocolitis, are rarely encountered or grossly underreported. On the other side, nonfatal congenital anomalies such as imperforate anus, hypospadias, cleft lip and palate, or club foot are quite prevalent, reflecting a large backlog of chronic surgical disability. The pediatric surgeon in LMICs rarely inserts central lines (due to the absence of total parenteral nutrition, chemotherapy, and/or central venous catheters), and gastrostomy buttons are also rarely available. He/she deals instead with significant trauma, surgical infections, and surgical emergencies in older children (who, unlike the neonates, have a better chance to survive).

In the North American environment, a significant portion (40% or more) of pediatric operations will be for emergency conditions, and studies suggest this is comparable in resource-constrained settings [4]. A snapshot of selected inpatients on day 1 of the pediatric general surgery unit at Mulago Hospital (a national referral center) in Kampala, Uganda, reflects the broad spectrum of conditions treated there, ranging from trauma to congenital anomalies, oncology, and infectious diseases, and their attendant complications (Table 20.1).

Ancillary Services: NICU, PICU, Radiology, and Pathology

Ancillary services are often limited and less functional than one would be accustomed to in high-income settings. Neonatal ICUs often have no functioning neonatal ventilators, hemodynamic monitoring, or parenteral nutrition, consisting of oxygen by nasal prongs and peripheral hydration [5].

Radiology uniformly includes plain films and ultrasound. Contrast studies and computer tomography are inconsistent and often unaffordable. Fluoroscopy is rarely available. Pediatric radiologists are extremely rare.

Histopathological services are usually available only in large referral hospitals and in private settings. Pediatric radiologists are rarely found, and frozen section pathology is not available, to our knowledge, anywhere in East Africa [5, 6].

Anorectal malformations	Trauma
2-year-old with repaired vestibular fistula with re-fistula	8-year-old with splenectomy post blunt trauma
Neonate with cloaca	10-year-old with bilateral chest tubes
Neonate with imperforate anus	s/p motorcycle crash
3-year-old with fecal incontinence post-PSARP	
Hirschsprung's disease	Infection
6 months old s/p Swenson with colostomy	9-year-old boy with abdominal sepsis without source at laparotomy
3-year-old with enterocutaneous fistula post- stoma takedown	5-year-old with post-op enterocutaneous fistula after typhoid perforation
2-year-old post-Swenson operation with incontinence	10-year-old s/p appendectomy
Infant with constipation and non-definitive pathology for Hirschsprung's disease	2-year-old with extremity gangrene after febrile illness
	5-year-old with snakebite
	1-year-old with wound tetanus
Oncology	Miscellaneous
2-year-old with testicular rhabdomyosarcoma	2-month-old with jaundice and?
4-month-old with sacrococcygeal teratoma	biliary atresia
2-year-old with Wilms' tumor	

Table 20.1 Selected conditions present on pediatric surgery unit 1 day at Mulago Hospital 2010

Equipment and Supplies

General adult surgical instruments are generally available, but small and specialized instruments are often missing, making sometimes for a frustrating operative experience. Visiting surgeons are encouraged to bring their own small set of specialized instruments.

Catheters and tubes, such as thoracostomy tubes, Foley catheters, Replogle tubes, feeding tubes, and drainage tubes, are inconsistently available and typically not in small sizes. Again, the surgeon is encouraged to bring a small sampling of these from home.

Sutures are another major area of concern, with typical hospitals having only silk, nylon and one braided absorbable material, and nothing smaller than 3–0.

In all these areas, we recommend the visiting surgeon to contact his/her host in advance and request a list of needed equipment and supplies worth bringing over for the specific types of procedures likely to be performed. There are also several charitable organizations distributing selected donated supplies and equipment at low cost.

Support and Referral Contacts

Work in a foreign setting, often in a foreign language and using different drug names and test units, can be very challenging. Recruiting the help of a local surgeon is essential for safe and effective care, as well as professional comfort and satisfaction. There are several helpful resources addressing pediatric surgery in LMICs including textbooks (geared to the district and tertiary hospitals in Africa), and papers highlighting pediatric surgical challenges in Africa, including tips for short-term pediatric general surgical missions [7–10].

Pediatric Acute Care, Anesthesia, and Resuscitation

The Essentials

- Understanding of the basic physiology and fluid and electrolyte management in children is essential.
- Local anesthesia resources and expertise may be limited.
- Knowledge of fundamentals of pediatric anesthesia and airway management are important to minimize morbidity and mortality.

Frequently, the greatest perioperative morbidity and mortality in the care of pediatric surgical conditions is related to safe anesthesia and perioperative care. Knowledge of the fundamentals of pediatric anesthesia and airway management and common pitfalls are essential before practicing in a resource-constrained environment.

An affiliated visiting anesthetic team may accompany the surgeon but, more importantly, should review availability of local anesthetic resources (human resources, equipment, drugs) and expertise. For example, ketamine is likely to be frequently used for shorter cases, compared to the high-income setting, and the visiting surgeon should be familiar with its basic properties and risks. In many settings, nonphysician clinicians may be the primary local anesthesia providers, and knowledge of the local practice is useful. There may be limited local practice to formally recover patients postoperatively. Particularly for elective cases, it may be useful to know the general volume of pediatric surgical cases (especially in neonates and infants). In some regions, especially outside of larger national referral hospitals, there may be no pediatric surgery performed other than by visiting teams [11].

The general surgeon with primarily adult training must have a sense of his/ her comfort level with a particular case if outside the scope of practice in their home environment. In uncertain cases, the surgeon may wish to consult with colleagues or local experts to determine the safety of specific elective cases. If there is a low volume of pediatric operations performed under general anesthesia, especially for neonates and infants, deferring the operation to an older age, perhaps greater than 6 months old, may be considered, as the morbidity of general anesthesia is reduced. Sometimes, in the absence of anesthetic expertise, in neonates with an abdominal emergency, it may be safer to perform the operation using abdominal field block with local anesthetic, if referral to a larger center is not feasible.

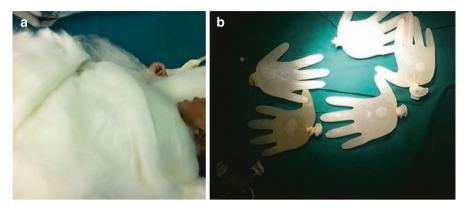


Fig. 20.1 Keeping babies warm in the absence of warming devices. (a) Using cotton wool and (b) using gloves with warm water

Vascular Access

Sometimes the greatest challenge may be intravenous access. The surgeon should be ready to establish alternate IV access if routine peripheral sites fail for infants and small children. IV access is often exacerbated by delay in presentation, failure to thrive for chronic conditions, and acute dehydration with need for resuscitation. Vascular access may be performed by intraosseous route, cutdown (saphenous, femoral, external jugular, umbilical in neonates), or percutaneous (external jugular vein or suitable scalp vein). Tunneled lines and fluoroscopy will likely be unavailable.

Heat Loss

Neonates are at risk of rapid heat loss even in tropical environments, and in the absence of formal warming devices, neonates and infants should be warmed intraoperatively with cotton wool rolls, gloves with warm water, or by other means (Fig. 20.1a, b). Warming intravenous fluids and blood before transfusion is helpful. Prolonged anesthetic time may compromise outcome in such children, and this may affect the surgical approach. Many hospitals may not have a neonatal intensive care unit or capacity to ventilate pediatric patients. This should of course be considered for any elective case that may require postoperative respiratory support.

Fluid and Electrolytes

Knowledge of the basic maintenance fluid requirements for infants and children is essential. For neonates, due to fluid overload in the first several days of life,

Table 20.2 Maintenancefluid requirements forchildren	Weight	Fluid requirement
	<10 kg (first 10 kg)	4 cc/kg/h
	10–20 kg (second 10 kg)	+2 cc/kg/h
	>20 kg	+1 cc/kg/h
Table 20.3 Estimated	Age	Volume
total blood volumes in	Age Preterm	Volume 90–100 cc/kg
	6	
total blood volumes in	Preterm	90–100 cc/kg

maintenance fluids are limited to 60–80 cc/kg/day, and D10 water is provided, with a gradual transition to 1/4 normal saline solution if possible over the first week. During this time, healthy neonates gradually lose this excess sodium through a physiologic diuresis. Neonatal resuscitation skills and resources may not be available in the austere setting; however, a major program (Helping Babies Breathe) is currently attempting to reduce neonatal deaths by teaching neonatal resuscitation skills in resource-limited settings [12]. Generally, the "4/2/1" rule for maintenance IV fluids can be used for children (Table 20.2). In other words, a 22 kg patient would require $(4 \times 10 = 40) + (2 \times 10 = 20) + (1 \times 2 = 2) = 62 \text{ cc/kg/h of IV fluid.}$ Normal saline and Ringer's lactate are the most available fluids, and solutions will require mixing to meet the appropriate requirement. Pumps will not be available, and providers must work closely with nursing staff to give four to six hourly fluid infusions to compose the 24 h requirement.

Potassium depletion is common, particularly in those with abdominal emergencies presenting late (e.g., intestinal obstruction, typhoid perforation). This should be carefully corrected before surgery. Potassium replacement should also be considered postoperatively in patients who have had abdominal surgery and are NPO for more than 48 h.

Local blood bank capacity should be evaluated, especially for elective cases where blood may be necessary. Blood volume is highest in the neonatal period and decreases gradually over time (Table 20.3).

Nutrition

Caloric requirements are highest in the neonatal period and first year of life (100–150 kcal/kg/day) and gradually decrease to 60–80 kcal/kg in adulthood. Patients in many limited resource settings may not have access to parenteral nutrition or the diversity of enteral formulas available in a more resource-rich environment. Even if available, enteral formulas or specific components of parenteral nutrition may be available only in private pharmacies at an exorbitant cost to families. These factors

should all be considered in operative planning and in any operation that may render a patient NPO for a prolonged period. Generally, parenteral feeding is indicated in children expected to be NPO for over 5 days. Enteral nutrition is always preferred if at all possible given the above factors. A higher proportion of children (20–30%) in limited resource settings will be undernourished at presentation, contributing to higher rates of postoperative complications such as superficial and deep wound infections [13]. Though NPO guidelines vary by setting, in general, a clear liquid diet can be taken up to 3 h before surgery, breast milk for 4 h.

Pain Management

The Essentials

- Unrelieved pain in children can lead to increased morbidity and mortality.
- Adequate and appropriate pain control should always be provided.
- Pain control should be multimodal, and two or more analgesics should be used to ensure effectiveness and minimize side effects.

Children, including neonates and preterm, do feel pain and require adequate and appropriate pain control when needed. Pain treatment is a recognized fundamental human right, and every effort should be made to alleviate it [14]. Despite this fundamental right, the management of pain in children in LMICs is often inadequate and suboptimal, due to several barriers: limited availability of appropriate drugs, lack of skills and training in pain management, and poor attitude toward pain care. Untreated or poorly treated pain in children has important consequences, including [15]:

- · Apnea and syncope in neonates and infants
- Decreased mobilization with attendant risks of atelectasis and deep venous thrombosis
- Long-term consequences such as psychological and behavioral changes, late pain-related behavior and perception, and decreased cooperation due to fear of pain

Most pain in surgical practice is related to surgical and bedside procedures, trauma, infective conditions, and cancer. In any of these situations, the pain may be severe, moderate, or mild, and the management would depend significantly on the severity.

Management Approach

The perception and expression of pain by children is influenced by age, cultural background, and previous pain experience in addition to the cause of the pain.

Access to pain care should be an integral part of perioperative care; adequate planning should be made for every child undergoing any surgical and painful procedure [16].

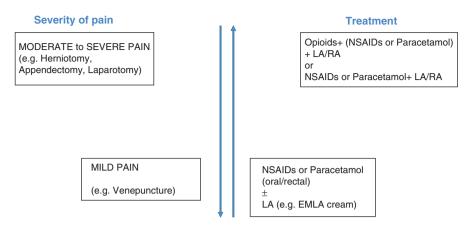
Treatment of pain requires the consideration of the following issues:

- Careful assessment of the cause and severity of pain. Pain assessment in children could be problematic as response depends on several factors. Assessment should be as objective as possible, using available pain assessment tools for children [17].
- 2. Frequent reevaluation and reassessment of the adequacy and effectiveness of treatment.
- 3. Availability and experience with commonly used drugs in the local setting. Knowing what drugs are available for pain treatment in the local setting is helpful. The safety profile and cost of available drugs should be taken into consideration, as the spectrum of available drugs may be limited and high cost may make preferred drugs unaffordable. Availability of nursing and anesthetic personnel with experience in pain management is crucial in the planning of appropriate multidisciplinary care.
- 4. Appropriate health personnel attitudes to modern pediatric pain treatment guided by appropriate education and advocacy.

The perception of pain is multidimensional, with several important components, all of which need to be addressed for optimal pain relief [18]. The components include:

- 1. Emotional (affective) component.
- 2. Behavioral component: behavioral response to pain.
- 3. Cognitive components: beliefs and cultural attitude to pain and pain control.
- 4. Sensory (neurological) component: the experience and response to pain. This component is the focus of most pharmacological (drug) treatment.

Treatment of pain should preferably be multimodal by combining appropriate pharmacological and non-pharmacological methods, whenever possible, to enhance the effectiveness of treatment. In choosing the drugs to use, combining two or more drugs helps to reduce dosage requirements and minimize the side effects of individual drugs. A practical approach, following the World Health Organization's step ladder treatment, is detailed in Fig. 20.2. The WHO recently proposed a two-step strategy for persistent pain from medical illness [18–20]. This is adaptable for treatment of surgically related pain, considering that differentiating between moderate and severe pain in children may be difficult, especially in younger children. Moreover, less potent opioids than morphine are of limited use in children. An exhaustive discussion of the treatment modalities is beyond the scope of this chapter.



LA/RA are used for postoperative analgesia where applicable

Fig. 20.2 Practical treatment of pain adapted for children. LA Local anesthesia by infiltration with bupivacaine, ropivacaine, or lignocaine, RA regional anesthesia (e.g., caudal block, nerve blocks, epidural block), NSAIDs nonsteroidal anti-inflammatory drugs (ibuprofen, ketorolac is a potent injectable form), EMLA eutectic mixture of local anesthetics. Notes: (1) Combine each modality with appropriate non-pharmacological treatment such as making the child comfortable, having parent/caregiver present to comfort the child, relaxation, and the use of appropriate images; (2) combining non-opioids (NSAIDs or paracetamol) with an opioid analgesic for moderate to severe pain is noted to produce opioid-sparing effect and reduces opioid requirements as well as the side effects [19]; (3) treatment should be tailored to the individual child, with dosing at regular intervals; (4) de-escalate or to lower levels as postoperative days progress and pain reduces or escalates to higher levels if severity of pain increases as identified on frequent reassessment; (5) avoid sedation in acute pain as it may mask response and severity [20, 21]; (6) morphine should be used in small doses and with caution, especially in neonates and infants and children with respiratory disease. Less potent opioids such as tramadol and pentazocine are of limited use in children as their safety remains inconclusive. However, the latter is often used in LMICs due to nonavailability of morphine; (7) below the age of 3 months, paracetamol should be used rather than NSAIDs; (8) the analgesics should be given intravenously for moderate and severe pain; (9) other modalities such as surgical options may be required in chronic pain

Pediatric Trauma

The Essentials

- Significant internal organ injury may occur without skull or rib fracture due to pliable skeleton.
- Traumatic brain injury is common and produces most mortalities.
- Therapeutic decision is often based on clinical findings as advanced imaging may not be available.

Abdominal Trauma

Introduction

Worldwide, but more so in LMICs, mortality due to trauma in children is largely attributable to head injuries. Abdominal and retroperitoneal injuries can lead to significant morbidity and mortality, especially following diagnostic delays due to shortage of affordable and readily available, up to date investigative modalities. This is unfortunately frequent in low-income countries.

Compared to adults, children are more susceptible to injuries from a low-velocity mechanism (such as falls) because their protective skeleton isn't yet fully developed; they have less fat to absorb and distribute mechanical forces, and their organs are in close proximity with each other. Motor vehicle crashes (MVCs) constitute the majority of pediatric injuries, and are more likely to have injury patterns in keeping with absent or poorly fitted restraints, or pedestrians hit by motor vehicles. However physiologically, children are more adept at compensation to shock, with minimal changes in vitals. One needs to be aware of the variability of vital signs based on age, with tachycardia an indicator of compensating hemodynamics.

Presentation

Abdominal organ injury is suspected on history and physical examination, and presentation may be delayed. Tachycardia in children is a sign of possible shock. Abdominal exam may reveal contusions, abrasions, tenderness, or distension, and these should prompt further evaluation. "Seat belt sign" and "handle bar" sign, or focal bruising in keeping with history of blunt force being exerted to the abdomen, should make one suspect abdominal injuries.

Investigation

Blood workup Baseline blood cell counts, blood group, and cross matching should be obtained during the initial trauma assessment.

Sonography Focused abdominal sonography for trauma (FAST)—done by the provider, where available—can be used during primary assessment as a screening tool for free fluid in the abdomen. An ultrasound is more likely to be available than CT scan. A formal abdominal ultrasound (performed by a technician) may also have similar benefits [22].

X-ray Though not usually helpful in the diagnosis of solid organ injuries, it may be useful in diagnosis of hollow viscus injury and pelvic injuries as well as skeletal trauma patterns that may prompt further evaluation for solid organ injury. Pneumoperitoneum is associated with intestinal perforations; pelvic fractures with bladder, urethral, and rectal injuries; and posterior rib fractures; thoracic and lumbar vertebral fractures may be associated with renal and pancreatic-duodenal injuries, respectively.

CT CT is highly accurate in diagnosis and staging of liver, spleen, and renal injuries. With contrast, duodenal, pancreatic, and vascular injuries can be diagnosed.

CT availability and affordability in LMICs are variable, and one should be in a position to make decisions on the available information, without awaiting a CT scan in an unstable child.

Explorative laparotomy Given that laparoscopy is not readily available in most LMICs, laparotomy is the next logical option in cases of diagnostic dilemmas such as free fluid with no solid organ injury, pancreatic injury, biliary tree injury, mesenteric hematomas, and diaphragmatic tears that may be missed in the initial evaluation and investigations.

Management

Liver and Spleen

Ninety to 95% of liver and splenic injuries in the pediatric population can be managed nonoperatively. This decision depends on accurate diagnosis and grading of the injury, hemodynamic stability, and the availability of close monitoring of the child, for any signs of deterioration. Signs of hemodynamic instability, peritonitis, or persistent fluid accumulation should prompt operative management. Nonoperative management involves hospital admission, observation, and possible repeat ultrasound scan. Higher grades of injury may require intensive care where possible or monitoring in a higher dependence unit. Children require restriction in activity up to 6 weeks after injury for higher-grade injuries that heal with nonoperative management.

Some children will present with tachycardia and evidence of intra-abdominal fluid, and initial resuscitation may fail to stabilize them. These children require emergency operative management because options such as massive transfusion protocols may not apply due to scarcity of blood and blood products in most LMICs, and the use of blood prior to achieving hemostasis is not advisable. However if available, 20 ml/kg of whole blood should be given to maintain hemodynamic stability till operative management ensues. Otherwise crystalloids should be used to sustain intravascular volume.

In the case of operative management for liver and splenic injuries, the patient is prepared and draped to provide surgical access to the chest, abdomen, pelvis, and femoral vessels. IV access should be initiated above the diaphragm. Packing of all four quadrants upon gaining access to the abdomen is done to achieve tamponade. Splenectomy is indicated in an unstable child with a high-grade injury. Postsplenectomy sepsis from encapsulated organisms is rare (0.2%), and pentavalent vaccine can be given especially to children <2 years who have splenectomy done to achieve hemostasis. Immunization against meningococcal infection and prophylaxis for malaria will also be required. The benefit of a splenic implantation into the omentum is uncertain, and partial splenectomy and splenorrhaphy, if appropriate, can be considered depending on the patient's stability and the surgeon's experience.

Operative hepatic injuries are more difficult to manage in resource-poor settings due to complexity of injuries and scarcity of general surgeons exposed to hepatobiliary surgery. In deep lacerations of the liver, in an unstable child, "damage control surgery" is a primary strategy. A Pringle maneuver, with intermittent occlusion of the porta, can help differentiate hepatic arterial and venous bleeding and can help to achieve hemostasis as resuscitation ensues. Adequate exposure requiring mobilization of hepatic ligaments is required, and this is followed by temporary maneuvers like packing and temporary abdominal closure to prevent abdominal compartment syndrome. Multiple reexplorations, change of packings, and repair of lacerations with deep figure-of-eight "liver sutures" may suffice for some injuries. Large hepatic fractures are best treated with anatomical liver resections. Packing and temporary abdominal closures can be employed until transfer to a facility with liver resection competence, but mortality in such cases remains high due to lack of critical care and other support measures.

Abdominal Compartment Syndrome

Intra-abdominal hypertension, associated with organ dysfunction, can occur in abdominal injuries resulting in decreased intra-abdominal organ perfusion and cardiopulmonary compromise. Manometry is scarce in LMICs, and one has to have a high index of suspicion, proper documentation of physical findings, and staged abdominal closure after damage control surgery. Signs of abdominal compartment syndrome include abdominal distension and firmness, hypotension, low urine output, and poor ventilation. Alternatively, this may develop in a patient managed nonoperatively and require laparotomy.

Biliary Injuries

These can be difficult to diagnose at initial presentation and may present at a delay with features of peritonitis or a biloma that are characterized by fevers, abdominal pain, altered bowel habits, and elevated liver enzymes. In LMICs, as interventional radiology is unavailable, management may involve placement of drains in the liver bed to drain the bile and transfer to a facility with ERCP with stenting. If this expertise is unavailable, operative exploration may be needed.

Pancreatic Injuries

These are difficult to diagnose without cross-sectional imaging or MRCP-ERCP. In the absence of peritonitis, allowing for formation of a pancreatic pseudocyst and managing the cyst later may be the safest solution in LMICs. If distal pancreas is involved, then distal pancreatectomy can be done.

Diaphragmatic Injury

When present, this is indicative of severe trauma. This can be diagnosed on two-view X-ray but may present at a delay, as well reported even in high-income settings. This may occur with pneumothorax or bowel herniation may cause hemodynamic instability. Primary repair with long-term absorbable suture is often possible.

Hollow Viscus Injury

Focal or localized blunt trauma can result in injury to hollow viscus. Seat belt injuries and acceleration-deceleration injuries also cause viscus injuries at points of

mesenteric fixation. Traumatic hollow viscus injury usually manifests as peritoneal irritation secondary to peritoneal contamination. Emergency laparotomy is indicated. Resuscitation, debridement, and primary closure may be attempted in the stable patient and depending on the location of the injury; diversion of stool may be needed in the unstable patient with multiple injuries, rectal injuries, or questionable bowel perfusion in mesenteric hematomas or bowel wall hematomas. For injuries to the stomach, primary repair is sufficient, and the gastroesophageal junction as well the posterior wall of the stomach must be inspected. Duodenal hematomas generally resolve with nonoperative management but may require a period of nutritional support that may not be possible in the resource-poor setting, and hematoma evacuation may thus be required. Isolated rectal injuries and perineal injuries also require a workup for possible sexual abuse.

Renal Injuries

Blunt trauma accounts for 80–90% of renal injuries, most commonly due to MVCs, and patients present with hematuria and flank pain. Organ preservation is the goal, and similar to other abdominal solid organs, nonoperative management is successful in the vast majority of cases. For the hemodynamically stable child, bed rest, serial exams, and interval ultrasounds (as needed) to monitor injury progress, as well as urinalysis until hematuria, have resolved. High-grade renal injuries with associated hemodynamic instability warrant nephrectomy, as do large urinomas.

Urinary Bladder Injuries

These may occur together with pelvic injuries. High-grade injuries with laceration and intra- or extraperitoneal extravasation of urine require operative management. Two-layered closure with absorbable suture (2/0) and bladder decompression through a suprapubic or urethral catheter are required to facilitate healing. Peritoneal lavage and suctioning of all intra-abdominal urine is important in preventing post-operative fevers and peritonitis.

Thoracic Trauma

The majority of thoracic trauma in children, again, is due to blunt force as a result of MVCs and may be part of a more complex picture in a multiply injured patient. A cardinal feature of pediatric trauma that also applies in the abdomen applies in the chest: the great pliability of the pediatric skeleton means that high-force injuries would cause broken bones/ribs in an adult, in a child may not lead to fractures but rather to underlying solid organ injury or contusion. In the chest this generally means that even with a significant degree of force, a child's ribs may not be broken, but the underlying lung may be contused.

Chest X-ray will be the primary investigation available as a CT scan is not likely to be routinely available. Lung contusion may be most apparent on a chest X-ray showing opacification of the affected area but no visible bone fracture. Pulmonary contusions generally will resolve with nonoperative intervention though X-ray resolution may lag. Lower rib fractures can be associated with abdominal visceral injuries, and these should be evaluated during the initial assessment.

Pneumothoraces, unless very small, require tube thoracostomy done in a similar technique to placement in an adult, but with a much smaller tube. Tension pneumothorax would ideally be identified in the primary survey and may mandate needle thoracostomy prior to placement of a larger chest tube. A large air leak suggestive of a major airway disruption may require thoracotomy and repair. Injuries to the great vessels (i.e., aortic tears) are not as common as they are in adults. Hemothoraces draining more than approximately 15–20 cc/kg or 2–3 cc/kg/h for 3 h or more may require thoracotomy for hemorrhage control. Penetrating thoracic trauma will require operative intervention in the majority of cases.

Head Injury

Traumatic brain injury (TBI) is different in children than adults. The relatively large head compared to the rest of the body makes the center of gravity higher and the head more likely to be injured. Unfused sutures allow some expansion and limited protection for the brain. Intracranial injuries frequently occur without skull fractures as the skull bones are thin and pliable and do not provide much protection.

TBI is common in children and accounts for up to 32% of all injuries in children in some studies. In LMIC reports of TBI combining children and adults, children account for about 4–13% [23, 24]. The majority of TBI in children occur from motor vehicle accidents (MVA) (often as pedestrians), falls from height, and assaults in the older child. In conflict areas, injury from missiles may become more prevalent [25, 26].

The patient may present with TBI in isolation or as part of multiple injuries. Due to lack of prehospital care in many LMIC settings, most of the patients are brought into the emergency room unresuscitated and without any prior first aid and may arrive hypoxic and hypotensive. Thorough examination of all systems is necessary (after resuscitation) to identify other injuries, which may take priority over TBI. Careful neurological examination should ascertain the severity of head injury using the Glasgow Coma Scale appropriate for children (Table 20.4) [27]. A tense anterior fontanelle (when the fontanelle is patent) suggests rising intracranial pressure. Lateralizing signs, including pupillary changes and limb weakness/paralysis, should be ascertained but are late signs especially in younger children with unfused sutures. Bleeding from any open head wounds should be controlled by gentle firm pressure and covered by warm, moist, and sterile gauze.

While evaluating the patient, those with moderate to severe head injury should have their C-spine protected using appropriate-sized cervical collar or collar improvised from available material such as cardboard. Alternatively, small sand bags and tightly rolled clothes can be placed on either side of the neck. Adequate resuscitation should be achieved as much as possible before moving the child out of the emergency room for any imaging studies.

Assessed function	Infants	Children	Score
Eye opening	Open spontaneously	Open spontaneously	E4
	Open in response to verbal stimuli	Open in response to verbal stimuli	E3
	Open in response to pain only	Open in response to pain only	E2
	No response	No response	E1
Verbal response	Alert, coos, and babbles	Oriented, appropriate	V5
	Spontaneous irritable cry	Confused	V4
	Cries in response to pain	Inappropriate words	V3
	Moans in response to pain	Incomprehensible words/ sounds	V2
	No response to pain	No response	V1
Motor responses	Moves spontaneously and purposefully	Obeys commands	M6
	Withdraws to touch	Localizes painful stimulus	M5
	Withdraws in response to pain	Withdraws in response to pain	M4
	Response to pain with decorticate posturing (abnormal flexion)	Abnormal flexion to pain	M3
	Response to pain with decerebrate posturing (abnormal extension)	Abnormal extension to pain	M2
	No response to pain	No response to pain	M1
Grimace component	Spontaneous normal facial/or motor activity (e.g., sucks tube, coughs)		G5
	Less than usual spontaneous ability or only responds to touch		G4
	Vigorous grimace to pain		G3
	Mild grimace or some change in facial expression to pain		G2
	No response to pain		G1

Table 20.4 Glasgow Coma Scale for children

Adapted from Abantanga et al. [27]

Severity in head injury using EVM components: GCS 14-15 = minor head injury, GCS 9-13 = moderate head injury, and GCS $\leq 8 =$ severe head injury

Plain radiographs (anteroposterior and lateral films) may be useful in the initial assessment as the presence of fractures may suggest possibility of intracranial injury (depressed fractures may be identified and the extent of depression of the inner plate could be helpful), and pneumocephalus may be seen [28]. However, intracranial injuries often occur without skull fractures, and radiographs are of limited value in therapeutic decision-making but may be the only imaging modality available. Anteroposterior and lateral C-spine radiographs should be obtained in moderate to severe injuries; however, children may have spinal cord injury without showing obvious radiological abnormalities (SCIWORA). CT scan is the desired imaging modality to identify and characterize brain injuries. If available, CT scan should

always be obtained in moderate to severe TBI. MRI is rarely available except in a few large tertiary hospitals. In the absence of CT scan, transfontanelle ultrasonography (if the fontanelle is patent) may be helpful in identifying intracranial hematoma.

Resuscitation is crucial to the outcome of TBI. Respiratory and cardiovascular stability should be achieved. In moderate to severe injury, oxygen should be administered by available methods (face mask, nasal catheter, nasal prongs). This may mean endotracheal intubation and mechanical ventilation in severe head injury. Hypotension should be corrected.

Restraining for restlessness should be avoided and sedation may mask neurological signs and making it difficult to identify deterioration. Rather, the cause of restlessness (hypoxia, hypotension, increasing intracranial pressure, electrolyte derangements, full bladder) should be identified and addressed. Seizures can be controlled with phenobarbitone or phenytoin, which are less likely to interfere with neurological assessment.

Due to limited resources, the approach to treatment of TBI in children in LMICs may need to be modified (Fig. 20.3). The majority of TBI in children can be managed nonoperatively; only about 3–21% require surgical intervention for evacuation of intracranial hematoma or elevation of significantly depressed skull fracture [23, 24, 29]. The decision to operate is guided by clinical assessment due to lack of CT scan in many settings. Rehabilitation may be necessary for those with residual neurological deficits.

The outcome of TBI in children is good, as the majority has mild injury. One recent systematic review of children with mild TBI concluded that most achieve functional physical and psychological recovery [30]. Post-traumatic seizures have been reported in about 18% of children with moderate to severe injury, more so in those <10 years old [31]. Mortalities of 3–15% have been reported from TBI in LMICs, mostly from severe TBI [23, 24, 29].

Burns and Burn Care

Please see Chap. 21 Plastic Surgery for the Non-Plastic Surgeon in the Low Resource Setting.

Fractures

See Chap. 16 Essential Orthopedics for Global Surgery as well. Musculoskeletal injuries are common in children. The management of these injuries in children differs from the adult due to the fact that [32]:

- The periosteum is thicker and provides greater fracture stability.
- The periosteum is more active and has greater potential for fracture healing by traditional bone formation.
- Fracture healing is faster.

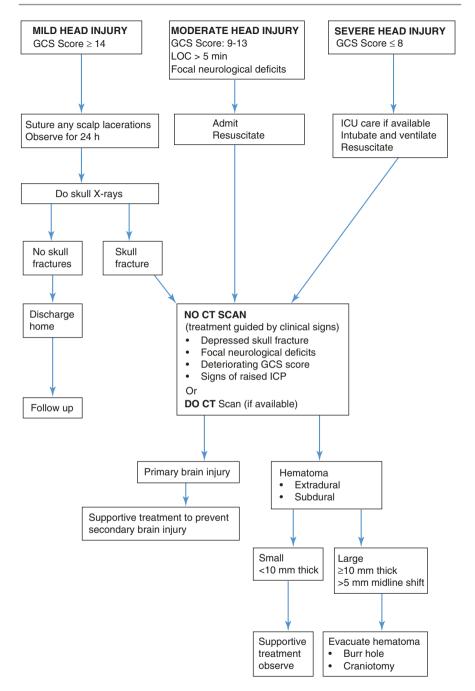


Fig. 20.3 Suggested algorithm for management of head injury in children in LMICs. *GCS* Glasgow Coma Scale, *LOC* loss of consciousness, *ICU* intensive care unit, *CT* computed tomography (Modified and adapted from algorithm provided by Dr. MR Mahmud, Consultant Neurosurgeon, Division of Neurosurgery, National Hospital, Abuja, Nigeria)

Type I	Clean wound <1 cm diameter with simple fracture pattern, no skin crushing
Type II	Laceration >1 cm $-$ <10 cm without significant soft tissue crushing. The wound bed may appear moderately contaminated
Type III	Open segmental fracture or a single fracture, >10 cm laceration with extensive soft tissue injury
А	Adequate soft tissue cover, despite extensive laceration and skin flaps
В	Moderate soft tissue cover, periosteal stripping
С	Vascular injury

Table 20.5 Gustilo-Anderson's classification of open fractures

Although the management of these injuries can be straightforward, lifelong deformity and disability may occur with inappropriate management. The most common sites for fractures are:

- The upper limbs
- The femur
- The tibia
- The fibula

The injury may be isolated but in about 14%, the fracture occurs as part of multiple injuries [33]. Most children will present with pain, swelling, or deformity of the limb and inability to bear weight on the affected limb. An inconsistent history and fractures at different stages of healing should raise the suspicion of child abuse, especially below the age of 5 years [34]. Tenderness and deformity at the fracture site is often present. If the fracture involves only one cortex of the bone (greenstick fracture), deformity may be absent, and inability to use the limb may be the only finding. Any break in the skin close to the facture site should raise the suspicion of open fracture, which if present should be categorized according to the Gustilo-Anderson's classification. Although attempts have been made to modify this classification for pediatric size, the original classification detailed above remains useful (Table 20.5) [35]:

Neurovascular injury should always be excluded by careful evaluation. Neurovascular injuries may be due to entrapment between the fracture ends, contusion, or transection and are particularly at risk in supracondylar fracture of the humerus, distal femoral fracture, and posterior dislocation of the knee. Paresthesia and pallor distal to the site of fracture are features of vascular injury and compartment syndrome. They are often late signs, and in children, differentiating between the pain of compartment syndrome and fracture pain may be difficult. It's important to always consider the possibility of these complications.

The affected limb should be splinted using available appropriate material (cardboard, plastic, purpose-made splints, strapping to opposite limb in femoral fracture) to reduce pain and minimize further soft tissue injury. Plain radiographs are often all that is needed for decision-making. At least two views (anteroposterior and lateral) should be obtained to identify the site of fracture, the presence and extent of displacement and angulation, and joint involvement. A greenstick fracture is common in younger children and should be carefully looked for. The presence of gas within the soft tissues should raise the suspicion of clostridial (and other anaerobic) infection, which can occur in those presenting late and following traditional bonesetter intervention. In the child with multiple injuries, if available, the Lodox® (low-dose whole-body X-ray) can be helpful in quick identification of skeletal injuries.

Resuscitation and management of life-threatening injuries (cardiothoracic, abdominal injuries, and intracranial hematomas) take priority over definitive treatment of fractures. Compartment hypertension requires fasciotomy. During resuscitation and care of life-threatening injuries, fractures should be splinted. Appropriate analgesia (preferably intravenous) is provided to control pain and make the child comfortable.

Closed Fractures

Most closed fractures in children are amenable to nonoperative treatment (closed manipulation and reduction). This is best done under image intensifier or fluoroscopy (if available) and under general anesthetic. In the absence of general anesthesia, the fracture site can be anesthetized by nerve blocks or infiltration of local anesthetic (bupivacaine or lignocaine/lidocaine) into the fracture site hematoma, but care is taken to avoid intravascular injection of the local anesthetic. Fracture reduction is then achieved by gentle manipulation, including correction of displacements and rotational deformities. After reduction, adequacy of distal pulses should be confirmed to avoid vascular compromise. If reduction was not image guided, adequacy of fracture reduction should be confirmed by post-reduction radiographs. Slight overlap of the fracture ends is considered acceptable as it would usually correct over time by modeling. In HICs, operative reduction and internal fixation is done for these fractures in adolescents, but this option may be limited in many LMIC hospitals due to lack of appropriate resources and cost. However, operative treatment has the advantage of precision, earlier mobilization, and quicker return to activities. Type I open fractures can be safely treated in the same manner after cleaning of the wound [36].

Open Fractures

Type II and III fractures require careful and meticulous attention to the wound and accompanying soft tissue injuries. Cross-matched compatible blood should be available for transfusion. Wound debridement should be done preferably under general anesthetic and with tourniquet in place (tourniquet time should not exceed 30 min at a time). Debridement consists of adequate exposure (this requires extending the wound by incision), generous irrigation with large volumes of warm saline for removal of all foreign material, and excision of devitalized and necrotic tissue. (A large syringe, e.g., 50 ml or free-flowing saline from an infusion giving set can be used) [32, 34]. Bleeding or contracting muscle after tourniquet removal indicates the tissue is viable. If the viability of soft tissue is doubtful, it may be safer to defer excising such tissue until a second look. Both fracture ends should be visualized and cleansed as foreign material

or soft tissue may be lodged between them. Unlike in adults, devitalized bone should not be removed but left in place as the periosteum will incorporate it during healing. Any exposed bone should be covered by local tissue. Before wound closure, the compartments should be palpated to exclude compartment tension. Following adequate debridement, the extended incisions can be closed primarily and the main trauma wound left open for delayed primary closure. Alternatively, the main trauma wound can be closed over a drain. Debridement may need to be repeated after 48 h in type III fractures. The timing of wound irrigation and debridement has been controversial, with the previous belief that infection rates are high if the wound is treated after 6 h from time of injury. In one report including adults and children, [37] and another report of 536 children with 554 open fractures, [38] there was no significant difference in infection rates across all types of open fractures, if treatment is done within 6 h compared to treatment after 6 h. Although there are no randomized controlled trials in children, it's now considered safe to treat open fractures within 24 h without increasing infection rates if appropriate antibiotics are given at admission [32].

In addition to these measures, parenteral broad-spectrum antibiotics (commenced on admission) should be given to prevent infection. A 3–23% overall infection rate have been reported in open fractures in HICs, but this rate may be higher in LMICs [37, 38]. The choice of antibiotics should be guided by prevailing local sensitivity profile, but where this is not available, a cephalosporin and metronidazole (or Co-Amoxiclav + aminoglycoside + metronidazole) should be given. Tetanus prophylaxis using tetanus toxoid should be given in unimmunized patients and patients with unknown immunization status to prevent tetanus. Fractures occurring in a particularly dirty environment (e.g., farm) require, in addition, passive tetanus immunization using human immune globulin or anti-tetanus serum as available.

Following nonoperative reduction, closed fractures can be immobilized by any available appropriate method including plaster of Paris cast (or Scotch cast) and traction (skin traction for younger children). When casts are used, the proximal and distal joints should be included in the immobilization. Immobilization of femoral fractures is particularly problematic as immobilizing the hip joint with a hip spica may be ineffective (the plaster cast may become wet and soften). Before application of a full cast, half plaster cast ("back slab") should be used first to allow edema to subside, to avoid creating a compartment syndrome. In clavicular fractures, the figure-of-eight splint may be all that is required. If operative reduction was done, appropriate internal fixation using available implants is done. It's recommended that implants be removed as soon as consolidation occurs, to avoid difficulties at removal.

Following operative care for open fractures, immobilization should be provided in such a way that allows access to the wound to facilitate wound care and identify any infection early. This can be provided by external fixators (if available) or plaster cast. If a cast is used, a "window" can be cut over the wound site after the cast has set.

Early mobilization, once there's consolidation to allow some weight bearing, should be encouraged to aid healing and minimize joint stiffness. Appropriate rehabilitation is important to ensure quick and full recovery to activities and function.

When appropriately and adequately treated, the outcome for closed fractures and type I open fractures are good. Nonunion is not common and acceptable overlaps

would usually correct by remodeling. Osteomyelitis can be a problem following open fractures and occurs in up to 6% of patients in HICs [37] but in LMICs, the risk if osteomyelitis can be significantly higher. This should be minimized by early administration of appropriate antibiotics. Complications arising from traditional bonesetting are common in LMICs. Limb gangrene from these complications results in need for amputation to control progressing and avoid overwhelming infection.

Surgical Infection in Children

Typhoid Fever

The Essentials

- Severe surgical complications can occur if typhoid fever is not adequately treated.
- Intestinal perforation is the commonest severe surgical complication.
- Adequate preoperative resuscitation and appropriate antibiotics are crucial to survival.
- Simple closure and segmental resection are effective treatments for intestinal perforation.

Typhoid fever is a multisystem infection caused by *Salmonella*. The disease is transmitted by feco-oral route and is endemic in many LMICs, largely due to improper sewage disposal systems, inadequate supply of clean water, and unhygienic environment. Twenty-one million cases occur annually, with children aged 5–15 primarily affected, though it does also occur in younger children [39]. Untreated, several surgical complications can occur (Table 20.6).

Complications of typhoid fever frequently present late, commonly after attempting treatment with over-the-counter antibiotics or local medications. Nearly 10% of children with typhoid fever develop intestinal perforation while on medical treatment, which tends to mask the features [40]. Symptoms can include fever and headache; abdominal pain sets in frequently after 1 week of onset of fever, and sudden increase in abdominal pain suggests intestinal perforation or other intra-abdominal complication. Abdominal distension follows in patients with intestinal perforation but may also be present in those without perforation. Diarrhea or constipation may

Table 20.6 Surgical complications of typhoid	Common Intestinal perforation	Less common Abscesses (hepatic, splenic, other)
fever	Intestinal hemorrhage	Pancreatitis
	Cholecystitis	Orchitis
	Osteomyelitis	Pleural effusion

be present and may be bloody in those with intestinal hemorrhage. Jaundice suggests the development of cholecystitis or overwhelming infection. Chest pain and pain in the limbs are suggestive of complication in those areas.

The typical child with typhoid is critically ill, particularly in cases of perforation. The diagnosis of intestinal perforation is mainly clinical, but laboratory tests and imaging may be necessary to guide treatment and also to exclude other conditions [41]. Serum electrolytes and creatinine should be drawn: in some patients, the electrolytes may be normal at presentation, but a repeat analysis after resuscitation commonly reveals depletion. For this reason, the decision to operate should not be made until the electrolytes are analyzed after resuscitation. However, lack of electrolytes and creatinine result should not unduly delay surgical intervention once adequate resuscitation has been achieved. Hypokalemia and metabolic acidosis are common. Complete blood count may identify anemia. Leukocytosis and neutrophilia are common in those with intestinal perforation. Blood should be grouped and crossmatched for pre-, intra-, and postoperative transfusion. Radiograph of the chest and upper abdomen may show pneumoperitoneum, which when large may need to be vented to improve respiration and reduce hypoxia. Abdominal ultrasonography can help to identify cholecystitis and also intraperitoneal abscesses.

Adequate preoperative resuscitations, including correction of fluid and electrolyte depletion and, if necessary, blood transfusion and nutritional support, are crucial to outcome of treatment. Intravenous broad-spectrum antibiotic combinations (to include an anti-salmonella antibiotic) should commence before surgery (e.g., amoxicillin or ampicillin + gentamicin + metronidazole, third-generation cephalosporin + metronidazole or ciprofloxacin + metronidazole).

The definitive treatment for intestinal perforation is surgical, to evacuate fecal contamination and prevent further contamination. In patients who are very ill and considered poor anesthetic risks, the use of ketamine is a safe and effective alternative. In addition to thorough peritoneal lavage, effective surgical options include simple closure of perforations, segmental resection, or damage control enterostomy, and decision should be determined by intraoperative findings.

Given the severity of infection and often delayed presentation in patients with typhoid intestinal perforation, complications following surgical treatment occur in 53–79% of patients, and mortality is variable with reported rates of 4.8–41% [42].

HIV and Tuberculosis

The Essentials

- HIV disease may present in children with surgical conditions such as soft tissue infections, cancers, and acquired rectovaginal fistula.
- Coinfection with tuberculosis may be manifested in thoracic or abdominal complications that may be difficult to differentiate from other pathologies.
- Antitubercular therapy should be attempted for the patient with chronic abdominal tuberculosis, but surgical exploration may be required.

In some regions, the prevalence of HIV in the pediatric population remains very high. Most children with HIV are infected through vertical transmission. As HIV+ patients may initially present to the medical system with a surgical condition, surgeons should be familiar with the general criteria for diagnosis of HIV infection. An infant or child may present with adenopathy suggestive of HIV disease, of HIV-associated tuberculosis, or with a soft tissue infection secondary to immune compromise. HIV-infected children also have a higher incidence of lymphoma. Several HIV-defining surgical pathologies bear mention such as spontaneous rectovaginal fistula or neonatal CMV enteritis. The symptomatic patient with HIV with an elective surgical condition should have surgery deferred until the medical status is optimized.

Tuberculosis is more prevalent in high HIV prevalence countries, with an overall reported incidence of 0.7–2 per 1,000 children, and accounts for about 15% of bowel obstruction in children [43]. General workup may include skin test and sputum for AFB. For chronic abdominal symptoms, contrast studies (from above and below) may show thickened bowel wall or strictures, and in case of ascites, a paracentesis may be performed. A more chronic presentation may have other causes of bowel obstruction in the differential, such as a delayed presentation of Hirschsprung's disease. Rectal examination may detect fissures, fistula, or stenosis. Ultrasound may show bowel thickening or peritoneal nodules. Nodular disease may also suggest malignancy; ascites may also occur from other medical problems such as liver failure or undernutrition.

Tuberculosis of the abdomen may present as acute or chronic peritonitis or bowel obstruction. For chronic symptoms, antitubercular therapy may be attempted for several weeks prior to surgery. Laparotomy may reveal thickened omentum, mesentery, or bowel wall. Copious ascites or adhesions may be present. Large or small bowel obstruction may occur due to thickening or strictures; presentation with frank bowel perforation is less common. High-grade strictures may require stricturoplasty in Heineke-Mikulicz fashion, and in cases of perforation, ostomy may be required due to peritoneal contamination. As tuberculosis cannot be "cleared" with surgery alone, the goals of operation are to treat the acute problem and to continue antitubercular chemotherapy postoperatively. Perforation is generally preferentially treated with resection and anastomosis rather than oversewing of the perforation alone, due to concern about tissue integrity around the site of perforation. Partial intestinal obstruction may be relieved with medical therapy over a period of months and may be attempted in selected circumstances.

Omphalitis: Surgical Complications

The Essentials

- Omphalitis is more common in LMICs and often responds to medical therapy.
- More aggressive infections can occur and require debridement and abdominal exploration.

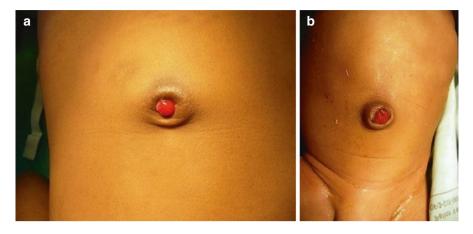


Fig. 20.4 Other umbilical pathologies to be differentiated from omphalitis. (**a**) Umbilical granuloma. (**b**) Patent vitelline duct (omphalomesenteric fistula)

Omphalitis is an infection of the umbilicus and umbilical stump. It is predominantly a disease of newborns but can also affect infants. Although omphalitis is mainly a medical disease, surgical complications may develop. The incidence of this condition in LMICs has been reported as 2–7 per 100 live births [44, 45]. In addition, the umbilicus is generally the portal of entry for cases of neonatal tetanus [46].

It is commonly caused by aerobic bacteria, including *Staphylococcus aureus* (most common pathogen), Group A streptococcus, *Escherichia coli, Klebsiella*, and *Proteus* species. In one-third of patients, anaerobic bacteria (*Bacteroides fragilis, Peptostreptococcus, Clostridium perfringens, Clostridium tetani*) are involved.

The disease is usually noticed at age of 3–5 days in preterm infants and 5–9 days in term infants. The local signs of omphalitis include purulent or foul-smelling discharge from the umbilicus or umbilical stump, periumbilical erythema, edema, and tenderness. Pyrexia, hypothermia, and jaundice may be present. Other features will depend largely on the nature of presenting surgical complication. Omphalitis should be clinically differentiated from other conditions such as umbilical granuloma, vitelline duct anomalies, and urachal anomalies (Fig. 20.4a, b)—which typically present without an infectious picture.

A microbiological swab of the umbilicus and blood should be sent for aerobic and anaerobic culture, and antibiotic sensitivity profile should be obtained to guide treatment. A blood count with differential for white cell counts may show a neutrophilia (or occasionally a neutropenia). Other investigations including plain abdominal radiography and ultrasonography may become necessary depending on complications suspected and to exclude other diagnoses.

Prompt antibiotic administration along with appropriate cord care normally controls uncomplicated omphalitis. In the absence of culture results, empiric antibiotic treatment should be started with Ampiclox + gentamicin (or a cephalosporin) + metronidazole. Tetanus prophylaxis is necessary in in most infants. The treatment of surgical complications is detailed in Table 20.7.

Time scale	Complication	Clinical notes	Treatment
Early	Necrotizing	Most common surgical complication	Antibiotics
	fasciitis	Starts initially as periumbilical cellulitis	Excision of all devitalized tissue
		The scrotum and abdominal wall commonly	Local wound dressing until infection controlled
		affected	Cover defect by direct suturing, skin grafting, or flaps as appropriate
	Intestinal	Usually the small intestine, occasionally the large	Cover the intestine with moist gauze and place in transparent plastic bag
	CVISCCIALIOII	Eviscerated intestine may be strangulated	Umbilical defect may need extension
)	Cleanse the intestine and return to peritoneal cavity
			Nonviable intestine should be resected
			If peritonitis present, do formal laparotomy and cleanse peritoneal cavity
	Peritonitis	Could occur without abscess	If no abscess, antibiotics alone may suffice
		Ultrasonography needed to exclude abscess	If abscess present, do laparotomy and drain
	Distant abscesses	May be retroperitoneal, hepatic, or elsewhere on the body	Abscess should be drained by:
		Abscess needs to be localized by appropriate imaging	(a) Percutaneous aspiration using wide-bore needle under imaging. May need to be repeated
			(b) Open drainage
Late	Portal vein	Portal hypertension is the major consequence	
	thrombosis	Although early complication, the major consequence is late	Portosystemic shunt required if portal hypertension develops ¹⁰
		A cavernoma ma produce biliary obstruction	Biliary obstruction should be treated appropriately
	Umbilical hernia	A common problem	Most would close spontaneously or significantly reduce in size by age of 2-4 years
		Usually asymptomatic but complications may develop	If not closed by 4 years, or complication develops, surgical repair required
	Peritoneal	A result of subclinical or treated peritonitis	Laparotomy and excision of adhesions required
	adhesions	Adhesions cause intestinal obstruction, which is usually not responsive to nonoperative measures	Any gangrenous intestine to be resected

20 Essential Pediatric Surgery

Uncomplicated omphalitis usually resolves if treated promptly. Most patients with surgical complications should recover but delayed presentation may result in a high mortality rate.

Pyomyositis

This is a bacterial soft tissue infection often distinguishable from the common simple soft tissue abscess by its presence within muscle tissue. This can be distinguished from infection in these areas due to adjacent osteomyelitis but may initially be treated in a similar fashion. It is most common in the extremities (lower limb > upper limb) and may present with a warm, swollen, fluctuant extremity. In other cases it may just present as pain with difficulty in walking. Ultrasound may be helpful but unnecessary given typical clinical presentation. Treatment consists of incision and drainage, with packing as necessary. An alternative depending on size and location may be incision and drainage through a modest incision and then a counterincision with the use of a Penrose drain across the wound to obviate the need for packing.

While the etiology of pyomyositis is unclear, workup for immunodeficiency may need to be considered. In addition, distinguishing the swollen possibly infected extremity from soft tissue sarcoma with surrounding inflammation can be difficult, and this should always be considered. If in doubt, biopsy of the surrounding tissue is critical. This can be especially challenging in the buttock in patients with a history of medicine injections into the soft tissue with swelling and scarring of the tissue. Familiarity with availability and effectiveness of local pathology services is also critical.

Empyema

The Essentials

- Pleural effusions can initially be treated by thoracentesis but may require tube thoracostomy if persistent.
- Mini-thoracotomy may be required for empyema, but thoracotomy and full decortication should be avoided if possible.

Pleural effusions occurring in the presence of underlying pneumonia (parapneumonic effusion, PPE) and empyema thoracis (ET) are common in LMICs and are estimated to complicate community-acquired pneumonia in about 20–53% of cases in children [47, 48]. It has been estimated that there are 151 million new episodes of community-acquired pneumonia in children below 5 years in developing countries (about 0.29 episodes/child-year) compared to 4 million new episodes (or 0.05

episodes/child-year) in high-income countries and remains a leading cause of death in under-five worldwide in 2015 [48–51]. Although the incidence of pneumonia is thought to have reduced over the decades, ET remains an important complication, and the incidence appears to be rising. The introduction of pneumonia conjugate vaccine has been effective in reducing the incidence of postpneumonic ET in some settings, with a 50% reduction in the incidence reported from South Africa [52].

A large majority of ET occur as a complication of pneumonia. However, in a small number of patients, it may complicate pulmonary tuberculosis and trauma, following thoracic surgery or needle aspiration of pleural effusion from other causes. The complication may also be part of the manifestation of systemic disease such as typhoid fever.

The clinical presentation of ET is frequently that of cough, fever, and respiratory distress, in a child being treated for pneumonia, but in a few patients, ET may be the first presentation. In large collections, there would be trachea deviation and lung collapse. Small collections may present simply as lack of improvement in a child being treated for pneumonia. Anemia and varying degrees of malnutrition are present in some patients, both of which could have impact on recovery and outcome.

The diagnostic evaluation should include an initial chest radiograph which is helpful in establishing the presence of collection (Fig. 20.5) and presence of lung parenchyma disease. In large collections, fluid levels may be present and loculations may be identified. However, it may be difficult to differentiate

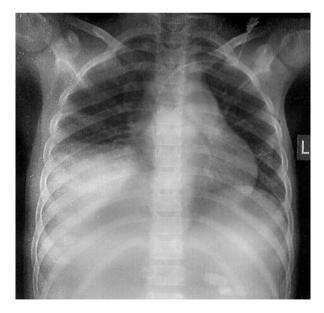


Fig. 20.5 Right-sided empyema thoracis in a 10-year-old girl

between collections and consolidation, and small collections may be missed. Thoracic ultrasound is good at quantifying the volume of collection and determining the presence of pleural peel (thickness) and loculations, as well as therapeutic decision-making, and should always be done if the facility is available.

Thoracentesis using a wide-bore needle (18–21G) helps to confirm the presence and nature of collection and should preferably be done under ultrasound guidance. In the absence of ultrasound, the site of thoracentesis should be guided by clinical and radiographic findings or done in the most dependent area of the chest. Repeated thoracentesis should be avoided. The aspirate should be cultured: Staphylococcus aureus is the isolate in >75%, mostly methicillin-sensitive Staphylococcus aureus, but methicillin-resistant Staphylococcus aureus (MRSA) is beginning to appear with increasing frequency in some LMIC settings and is important in HIV-associated ET [50, 53]. Streptococcus pneumoniae is cultured to a lesser extent, especially in older children. However, one recent report from South Africa, in which both the pleural fluid and blood were cultured, indicated that S. pneumoniae was the commonest cause of ET complicating community-acquired pneumonia: S. pneumoniae was cultured in 48% compared to S. aureus in 17%, with the former being more frequently cultured from blood [52]. In some reports, cultures are sterile in >50% of patients, possibly a result of prior treatment with over-the-counter antibiotics before arrival in hospital [50, 51]. Where possible, the aspirate should be analyzed for white cell count, lactate dehydrogenase (LDH), pH, and glucose, to help in categorization, but analysis for these parameters should not be routine. Lymphocytosis in the aspirate should prompt evaluation for pulmonary tuberculosis and malignancy [54].

Although computed tomography (CT) scan is effective at defining and characterizing ET, and assessing the state of the lung parenchyma, it should not be done routinely. Moreover, CT scan is not available in many LMIC hospitals (except large tertiary hospitals) and is costly. CT scan is best reserved for the planning of definitive surgical intervention when that becomes necessary. Sputum should be cultured and Ziehl-Neelsen stain done to exclude pulmonary tuberculosis, as appropriate. Complete blood count may show leukocytosis.

An understanding of the pathophysiology of PPE and ET has allowed the classification of the condition into various stages and grades. However, pleural infection is a continuum as detailed in the three stages detailed below [54]:

- Stage I (exudative): the pleural fluid/collection is clear.
- Stage II (fibrinopurulent): there's deposition of fibrin, increase in white cells, and eventual pus formation. Loculations may form.
- Stage III (organizational): there's infiltration of the cavity with fibroblasts and formation of thick inelastic pleural membrane (or peel). This stage may result in spontaneous healing or become chronic with lung entrapment and restriction.

It has been suggested that PPE is complicated if pH of the aspirate is <7.2, glucose <40 mg/dl, lactate dehydrogenase (LDH) \geq 1,000 IU/L, size of the collection is

>1 cm, loculations are present, and bacteria culture is positive [55]. However, these biochemical parameters are not routinely necessary in clinical practice [54].

Clinical parameters and appropriate imaging should guide treatment. Early non-purulent collections may be treated by thoracentesis, but repeated thoracentesis should be avoided; all but very small collections require more definite drainage. Tube thoracostomy drainage is effective in 75–85% of cases [48]. As purpose-made chest tubes and drainage receptacles may not be available, other available materials can be used (e.g., Malecot's catheter, Nelaton's catheter, suction tubes, large nasogastric tubes). An algorithm for the treatment of ET, adapted to the LMIC setting, is detailed in Fig. 20.6. Most patients will require tube thoracostomy drainage using an appropriate-sized tube, and the presence of thick pus requires larger tube to ensure adequate drainage. The chest drain should be removed when drainage reduces to <10–15 ml/day, there's clinical improvement, and lung re-expansion is adequate as determined by repeat chest radiograph.

In HICs, placement of small catheters <14 F (e.g., pigtail catheter) along with fibrinolysis (using tissue thromboplastin activator or urokinase), even if there are loculations, and video-assisted thoracoscopic surgery (VATS) are considered first-line treatments and are associated with quicker recovery rates [47]. Although thoracostomy and fibrinolysis are used in some LMICs, these options are of limited use in the typical LMIC setting due to limited resources and cost. In the few patients, in whom tube thoracostomy tube drainage fails or is not appropriate, a mini-thoracotomy (using a small incision of about 3 cm in length) or rib resection should be done to break down loculations and a tube drain placed. This is effective in most patients, and decortication should be reserved for patients with lung restriction from peel, as this surgery may be associated with significant morbidity and long hospital stay. In some patients with chronic ET, an open chest drain could be used in some patients, but this is associated with prolonged morbidity. Lung parenchyma involvement such as lung abscess, lung necrosis, or bronchiectasis usually does not require surgical treatment.

Before availability of culture results, empirical broad-spectrum antibiotics should be started, including those effective against *Staphylococcus aureus* and *Streptococcus pneumoniae*. Once culture results are available, the antibiotic regimen should be guided by sensitivity profile, especially if the patient is not improving. Initially, antibiotics should be given parenterally for 5–7 days and then changed to oral route once pyrexia has subsided and continued for 1–4 weeks depending on the spectrum and sensitivity of bacteria involved and the response of the patient. Patients with underlying tuberculosis require administration of appropriate antituberculous drugs for 6–9 months. Other primary pulmonary pathologies should be treated as indicated [54].

Blood transfusion and nutritional rehabilitation should be done as necessary. Physiotherapy is considered unhelpful and should not be routinely done. However, in older children, early mobilization and exercise is helpful in aiding recovery [54].

With the administration of potent and appropriate antibiotics, and appropriate drainage and treatment, most patients with ET should recover. However, in LMICs,

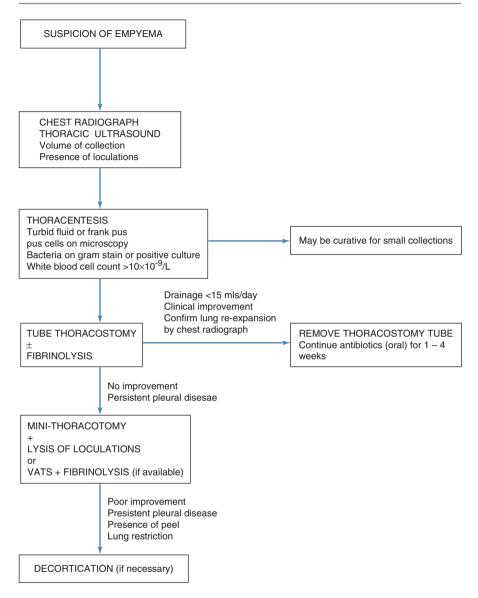


Fig. 20.6 Algorithm for treatment of empyema thoracis in LMICs. *VATS* Video-assisted thoracoscopic surgery. Fibrinolysis: use tissue thromboplastic activator or urokinase

mortalities of 4–16% have been reported, often from delayed presentation and underlying systemic disease [50, 52]. In tuberculous ET, bronchopleural fistula and malnutrition are common, pleural drainage is longer, and residual pleural fibrosis may persist [56].

Parasitic Infestations

The Essentials

- Ascaris intestinal infections can first be treated medically in stable patients.
- Exploration is required for failed medical management or peritonitis and may require resection, ostomy, or decompression of worms through enterotomy.
- Amoeboma and liver abscess should be first be treated medically with metronidazole, but failure may require percutaneous drainage or operative exploration.
- Hepatic hydatid disease should first be treated medically, but larger cysts may require operative exploration and excision with obliteration of the cyst cavity.

Parasitic infestations are endemic in many LMIC communities, and children often tend to bear the brunt of the disease. The World Health Organization (WHO) estimates that 882.5 million children required preventive chemotherapy for soil-transmitted helminths in 2009, with 74.3% of them located in Africa and Southeast Asia [57].

The common infestations are from helminths, protozoa, and ectoparasites and usually occur as a result of contact with or ingestion of contaminated food, water, or the primary host. Although parasitic infestations produce mostly medical illness, complications may arise that require surgical intervention [58, 59]. Some of the more common complications requiring surgical intervention are detailed in Table 20.8.

Gastrointestinal Complications

The gastrointestinal tract (GIT) is by far the leading site for surgical complications of parasites, ranging from acute to chronic manifestations and diagnostic confusion.

Intestinal Obstruction

Presentation with acute abdominal pain, arising from intestinal obstruction, intestinal volvulus, and intestinal perforation, is common manifestation with *Ascaris lumbricoides*, affecting about 41% of children with ascariasis [59–61]. In some settings, ascariasis is so endemic that it ranks as the leading cause of intestinal obstruction in children, reaching 60% in one report [58]. Intestinal obstruction is usually the result of heavy load of adult worms in the intestinal lumen, with the worms entangling and forming a "ball." Intestinal volvulus occurs as a result of the loaded loop of the intestine becoming redundant and twisting, intestinal gangrene may occur from the volvulus or from pressure necrosis caused by the ball of worms, and perforation occurs from ischemia of overlying intestinal wall. Occasionally, a worm may migrate through an anastomotic suture line leading to anastomotic leakage. Other

Site	Surgical complication	Parasites
Gastrointestinal tract	Intestinal obstruction	B. Ascaris lumbricoides
		Taenia saginata (solium)
	Intussusception	C. Ascaris lumbricoides
		Entamoeba histolytica
	Rectal prolapse	Enterobius vermicularis
		D. Trichuris trichiuria
		E. Entamoeba histolytica
	Recurrent abdominal pain	F. Ascaris lumbricoides
		Taenia saginata (solium)
		G. Enterobius vermicularis
		Trichuris trichuria
		H. Entamoeba histolytica
	Amoeboma (right iliac fossa	I. Entamoeba histolytica
	mass)	
	Intestinal hemorrhage	J. Entamoeba histolytica
	Toxic megacolon	K. Entamoeba histolytica
Biliary tract	Biliary obstruction	L. Ascaris lumbricoides
	Cholangitis	Clonorchis sinensis
Urinary bladder	Granuloma	Schistosoma hematobium
	Carcinoma (squamous cell)	
Liver and lungs	Abscess	M. Entamoeba histolytica
	Cysts	Echinococcus granulosus
		(multilocularis)
Soft tissue	Dracontiasis (dracunculiasis)	N. Dracunculus medinensis
	Myiasis	Larvae of fly

Table 20.8 Surgical complications of parasitic infestation in children

worms such as *Taenia saginata* have been known to produce intestinal complications.

There are no clear distinguishing features from other causes of intestinal obstruction, but there may be a history of passage of worms in stool or vomitus. Plain abdominal radiographs may show the "whirlpool" pattern in addition to features of intestinal obstruction (intestinal dilatation and multiple fluid levels). Abdominal ultrasonography is helpful and may show outlines of floating worms within free fluid in the intestine [60]. Stool microscopy may show the ova of the causative parasite.

The initial treatment approach is medical, in addition to resuscitation, nasogastric drainage, and monitoring. Medical treatment is effective 50–78% of children with intestinal obstruction from ascariasis and consists of administration (through the nasogastric tube or oral if feasible) of albendazole, mebendazole, or levamisole [60, 61]. Gastrografin given by nasogastric tube may hasten the expulsion of the parasites. The progress of medical treatment can be monitored by ultrasonography.

In about 22–50% of children with ascaris obstruction, surgical intervention is necessary, due to worm impaction, peritonitis from intestinal gangrene, volvulus, or intestinal perforation. Impacted worms can be milked into the colon (to be passed out in the stool) [60, 61]. If this fails, the impacted worms can be evacuated through an enterotomy in a healthy looking adjacent segment of the intestine (not directly over the impacted worms as that segment may be compromised). Intestinal perforation and gangrene/necrosis require resection of the affected segment. Those treated surgically initially should receive appropriate anthelminthic once it's safe to administer enteral medications. Mortality of 1-19% from ascaris intestinal obstruction have been reported, but this should be unusual if presentation is early and treatment prompt.

Recurrent Abdominal Pain

A wide spectrum of GIT parasites (Table 20.8) produce abdominal pains and sometimes vomiting, the character of which is nonspecific and may be present for several weeks to months. Therefore, any child presenting with recurrent abdominal pain should have parasitic infestation excluded, usually by stool microscopy and abdominal ultrasonography. Even when evaluation reveals a surgical pathology, any identified parasitic infestation should be treated with anthelminthic before treating the surgical condition, to avoid postoperative morbidity from the parasitic infestation.

Other GIT Complications

Gastrointestinal parasites may also cause other GIT complications. Intussusception should be treated on its merit along with administration of appropriate anthelmintic drugs. Rectal prolapse may occur as a result of tenesmus and rectal irritation caused by the parasites. Stool microscopy (using fresh stool sample for amoebiasis) should show ova of the parasites or trophozoites of *Entamoeba histolytica*. Uncomplicated rectal prolapse is best treated nonoperatively by manual reduction and appropriate anthelmintic, including metronidazole for amoebiasis.

Entamoeba histolytica may produce a granulomatous reaction in the ileocecal region, resulting in the formation of a mass (amoeboma). This mass may be clinically indistinguishable from other causes of a mass in that region. Diagnosis may be confirmed from stool microscopy showing trophozoites of *E. histolytica*. Serologic tests using enzyme-linked immunosorbent assay (ELISA) or other immunological methods may be helpful. Sometimes, the diagnosis is only suspected when a mass is encountered in the ileocecal region at laparotomy. If a preoperative diagnosis is made, treatment should be medical by administration of metronidazole or tinidazole for 7-10 days. If the mass persists or intraoperative diagnosis is suspected, surgical excision should be performed and histopathological examination of the specimen done.

Biliary Complications

Jaundice from biliary obstruction and cholangitis may complicate infestation by parasites such as *Ascaris lumbricoides* and *Clonorchis sinensis* (liver fluke). Ultrasonography may show the outline of a worm within the biliary tree. Sometimes, *Ascaris* can migrate into the biliary tree following administration of anthelmintic for intestinal infestation. Medical treatment with appropriate anthelmintic is usually effective, and the response to treatment can be monitored by ultrasound if the parasite was identifiable at initial ultrasonography.

Liver and Lungs

Amoebic Liver Abscess

Hepatic abscess may occur as a complication/progression of intestinal amoebiasis. The patients usually present with right hypochondrial pain, fever, and tender mass in the liver with or without intercostal tenderness over the hepatic area. Sometimes, the abscess, which is usually in the right lobe, ruptures into the pleura or lung with the latter producing coughing of chocolate-colored material. Rupture into the peritoneal cavity produces amoebic peritonitis. Chest radiograph including the upper abdomen showing "tenting" or elevation of the right hemidiaphragm is suggestive of the diagnosis. Ultrasonography that would show the diagnosis of collection in the liver and needle aspiration (ultrasound guided if possible) of chocolate-colored fluid (anchovy sauce) confirms the diagnosis. Identification of amoebic trophozoites in the aspirate is unlikely, as it contains mostly necrotic liver tissue. The aspirate should be cultured to identify superimposed bacterial infection. The serological tests could be helpful if available and are positive in >90% of patients. Microscopic examination of fresh stool sample may identify the trophozoites of *Entamoeba histolytica* in patients with invasive disease.

The treatment of amoebic liver abscess is medical. Metronidazole or tinidazole should be given, initially intravenous, but should be changed to oral route once fever and tenderness have subsided. The medication should be continued for 10–14 days. Other anti-amoebic drugs including chloroquine and dehydroemetine are available but rarely used due to the fact that metronidazole is effective and associated with less side effects. Other appropriate antibiotics should be given if superimposed bacterial infection was identified at culture. Response to treatment should be monitored clinically and by serial ultrasonography. Surgical drainage is rarely required. Nonresponding abscesses (after 72 h of amoebicide) and abscess in the left lobe can be aspirated under ultrasound guide. In peritonitis, the amoebic pus should be evacuated percutaneously under ultrasound guide. However, if percutaneous drainage is not possible, the pus should be evacuated at laparotomy and the peritoneal cavity thoroughly lavaged with warm saline while protecting the wound edges to prevent cutaneous involvement.

Hydatid Disease

Hydatid disease is caused by *Echinococcus granulosus* and *multilocularis* (less commonly *Echinococcus vogeli*) and is endemic in North Africa and the Mediterranean region. The liver and lungs are most commonly affected and children aged 10 years and above predominate [62, 63]. The disease presents as single or multiple cysts in the liver and/or lungs. Diagnosis is established with the use of ultrasonography which is capable of identifying daughter cysts and hydatid sand and is able to differentiate it from amoebic liver abscess and pyogenic liver abscess. CT scan has a high sensitivity.

Medical treatment is recommended for cysts <5 cm in diameter and multiple cysts [63]. For cysts \geq 10 cm and failure of medical treatment, surgical treatment (along with medical treatment) offers the best hope for cure. Surgery usually

consists of removal of the parasites, sterilization of the cyst cavity with a scolicidal agent (e.g., formalin, chlorhexidine, cetrimide, povidone-iodine, ethanol, hypertonic saline), and protection of surrounding structures. The most effective surgical options include cystectomy or pericystectomy and capitonage (obliteration of the resulting cavity by multiple sutures). The effective anthelmintic for this infection includes albendazole and mebendazole, given over several weeks.

Surgical Emergencies in Neonates

Abdominal Wall Defects

The Essentials

- Distinguishing features of gastroschisis and omphalocele should be clear to a visiting clinician.
- While the bowel can be serially reduced and the defect closed in gastroschisis, the absence of TPN adversely impacts outcomes.
- Though associated anomalies are more common in omphalocele, outcomes are improved as nonoperative management is often sufficient in the neonatal period for larger defects, with subsequent repair of abdominal wall hernia at an older age.

Surgeons are frequently involved in the management of abdominal wall defects in the neonatal period. Gastroschisis involves a full-thickness defect in the abdominal wall to the right of midline, with bowel outside the abdominal cavity. In omphalocele, which is slightly more common, the bowel is outside the abdominal cavity, but the peritoneal lining is preserved (except when the coverings are ruptured).

In gastroschisis, immediate priority is resuscitation of the baby from third spacing and prevention of heat loss. Over the last decade, there has been a trend to placement of a temporary silo, serial reductions, and delayed closure of the abdominal wall under general anesthesia. If primary reduction is not possible at initial evaluation in a resource-limited setting, a temporary abdominal wall covering should be devised; in the absence of a spring-loaded silo, a basic dressing using a plastic bag may able to be used that covers the abdomen and the lower extremities to retain heat and minimize fluid losses. Alternatively, a silo can be constructed by using or sewing together several pieces of urine bags (generally softer than IV fluid bags) or female condoms and then performing gradual reductions once or twice a day [64] (Fig. 20.7). A Replogle tube should be placed for bowel decompression. In resourcerich settings, most babies have a 4-6-week hospital stay and a prolonged dependence on TPN due to inflammation of the bowel (from amniotic fluid exposure prenatally) and associated ileus that persists even after abdominal wall closure. This presents great difficulty in the absence of TPN. Even in the presence of a NICU and TPN, a recent South African series reported mortality of 43%, primarily due to



Fig. 20.7 Improvised silo (surgical silo) for omphalocele using urine bag

sepsis [65]. Volvulus and bowel ischemia may complicate gastroschisis, especially with late presentation (Fig. 20.8).

If general anesthesia is unavailable for closure and if the bowel can be gently reduced into the abdominal cavity without excessive increase in intra-abdominal pressure, the defect can be temporarily closed with either the umbilicus or "Wharton's jelly" folded over, a temporary piece of mesh, or suitable low-cost local alternative. This may result in a delayed formation of a hernia at this site, but this may be acceptable in this setting.

With omphalocele, associated defects are more common, and these are more often the cause of mortality than the omphalocele itself. Blood glucose should be obtained, and echocardiogram should be performed if available in the setting of an audible murmur and an abdominal ultrasound obtained. For smaller defects (<5 cm), primary closure is indicated; for larger defects, an escharizing agent such as 1% silver sulfadiazine (or native honey if the sac is infected) may be used to thicken the peritoneum and the defect and associated hernia repaired when the child is older (Figs. 20.9 and 20.10). If the fascia cannot be closed, then skin only can be closed, leaving a hernia that can be repaired at a later date. With an intact peritoneal covering, there is usually no delay in gastrointestinal function, and the baby can be started on feeds. Repairing the hernia at a later date may require the use of a mesh. If the sac is intact, serial gradual compression bandaging over a few weeks may help to facilitate earlier repair of the defect. In the case of ruptured omphalocele that cannot be primarily repaired, a temporary silo can be devised in a similar manner described above, with serial reductions subsequently performed. In the infant with omphalocele, tetanus prophylaxis should be given if the mother did not receive this during antenatal period or if the status is unknown.

Neonatal Bowel Obstruction

In the neonatal period, anorectal malformations (discussed below) are the most common source of bowel obstruction, followed by intestinal atresia-stenosis,



Fig. 20.8 Late presenting gastroschisis with bowel ischemia



Fig. 20.9 Omphalocele treated nonoperatively-newborn and after nonoperative treatment

Hirschsprung's disease, and malrotation. Perhaps the greatest difference between acute abdomen in the neonatal period in higher- and lower-income countries is that necrotizing enterocolitis, primarily a disease of prematurity, is rare in settings without a neonatal intensive care service and the capacity to care for the medical problems of prematurity. It will therefore not be discussed.





Malrotation and Midgut Volvulus

The Essentials

- Bilious vomiting mandates evaluation for malrotation, best assessed by upper GI imaging.
- Peak incidence is in the first several months of life.
- Abdominal exploration may be required based on history and physical exam alone in some cases, and correction requires a Ladd's procedure.

Malrotation generally presents (80%) with bilious vomiting in the term infant 1 month of age or younger. The infant with bilious vomiting should be presumed to have malrotation until proven otherwise. The disease is due to abnormal fixation of



Fig. 20.11 Intestinal malrotation on upper GI study

the bowel prenatally, leading to a shortened distance between the ligament of Treitz and the ileocecal junction and a narrow mesentery, resulting in heightened risk of volvulus (Fig. 20.11). In the presence of abdominal distension and peritoneal signs, the patient may proceed to laparotomy without imaging, after establishing IV access and resuscitation. A nasogastric tube should be placed to decompress the bowel.

Plain abdominal films may show a range of findings, from near normal-appearing bowel gas pattern to "white out" due to bowel edema and ascites or pneumoperitoneum with perforated volvulus. A Doppler ultrasound may show swirling of the mesenteric vessels at the base of the mesentery with complete volvulus, though this may require an experienced pediatric radiologist. Though there is some controversy, the gold standard is an upper gastrointestinal study, to identify the duodenojejunal junction (DJ) and rule out other causes of proximal obstruction. The DJ should be located to the left of midline and at the level of the duodenal bulb, at approximately the L1 pedicle. This study, however, may not be available in the limited resource setting, and laparotomy may be indicated based on clinical suspicion alone [66].

A transverse laparotomy incision should be performed. Chylous ascites may be encountered in the presence of partial or intermittent obstruction. The bowel should



Fig. 20.12 Chronic midgut volvulus from intestinal malrotation

be fully eviscerated. In the setting of midgut volvulus (Fig. 20.12), the bowel is derotated in the counterclockwise direction and examined for improvement in perfusion. A pulse should be sought at the root of the superior mesenteric artery and can be evaluated by Doppler if available. The bowel can be covered with warm sponges, and a period of waiting 10–15 min may be appropriate. In cases of questionable viability and demarcation, a second-look laparotomy may be planned for 12–24 h if intensive care is available, and a temporary abdominal dressing may be devised. If the bowel is well perfused, a Ladd's procedure should be performed: (1) the mesentery should be broadened at the base, which involves carefully freeing peritoneal adhesions in this area; (2) the ligament of Treitz should be taken down to straighten the duodenum and upper jejunum; (3) the small bowel should be placed on the right and the cecum in the left hypochondrium; (4) an appendectomy should be performed. At the end of a Ladd's procedure, the bowel is left in a position of "nonrotation." After closure, feeds are commenced after evidence of return of bowel function.

Intestinal Atresia

The Essentials

- Intestinal atresias present in the neonatal period with bowel obstruction and diagnosis can be established with basic imaging.
- Repair requires establishment of intestinal continuity, often requiring tapering procedures.
- Outcomes are adversely impacted by availability of anesthesia and perioperative care.



Fig. 20.13 Presentation of intestinal atresia on abdominal radiograph

Jejunoileal atresia and stenosis may have varied epidemiology, with an incidence ranging from 1 in 1,000 births in the African setting to 1 in 3,000 births in the United States. Duodenal atresia may also be encountered but is thought to be less common [67].

Neonates with atresia present in the early postnatal period with abdominal distension, vomiting, intolerance of feeds, and failure to pass meconium, while intestinal stenosis may not present until older age. Plain abdominal X-rays may show dilated intestinal loops (Fig. 20.13). Enema may show a microcolon or may confirm meconium plugs or ileus, and for these conditions, the enema may be therapeutic, and the patient may not require surgical intervention. Demonstration of colon continuity preoperatively also obviates the need to do this at laparotomy. Nasogastric decompression and IV resuscitation are necessary prior to laparotomy. Generally, nasogastric aspirate of > 20-30 cc in the newborn suggests obstruction.

Treatment at laparotomy depends on the type of atresia encountered (Fig. 20.14). Generally, the ends of the atresia must be resected before restoration of bowel continuity. At this time, the rest of the bowel can be flushed with

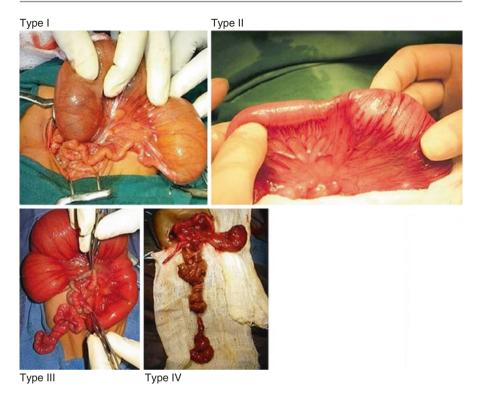


Fig. 20.14 Four types of intestinal atresia

normal saline to evaluate for the presence of multiple atresias (20%). There may be a significant size discrepancy between dilated proximal bowel and decompressed bowel distal to the atresia. The approach may be dictated by the anatomy present. Sometimes resection of the bulbous end of dilated proximal bowel may facilitate anastomosis; an antimesenteric "slit" may also enlarge the distal bowel diameter. Tapering enteroplasty or imbrication may also be performed on dilated proximal bowel to address the size discrepancy. A continuous suture may need to be used to minimize anesthetic time for a neonate. Feeding is commenced after return of bowel function, and if possible, total parenteral nutrition is maintained during this time. For more proximal atresias, a transanastomotic feeding tube and a gastrostomy tube may be particularly helpful in an environment without parenteral nutrition. While survival in high-income countries is >90%, it may be as low as 40–50% in sub-Saharan Africa due to late presentation and the lack of parenteral nutrition [68].

Hirschsprung's Disease

The Essentials

- Hirschsprung's disease involved congenital aganglionosis of the intestinal tract, most commonly involving the rectosigmoid colon.
- Delayed presentation is the rule in resource-poor areas, with abdominal distension, chronic constipation, and malnutrition.
- While pathologic confirmation is the gold standard in HICs, exploratory laparotomy may be required in LMICs based on history, physical, and basic investigations.
- Full correction requires resection of the aganglionic segment and a low rectal anastomosis.
- A staged approach may be necessary in advanced presentation, with a temporary stoma, followed by a pull-through procedure, and then stoma takedown.

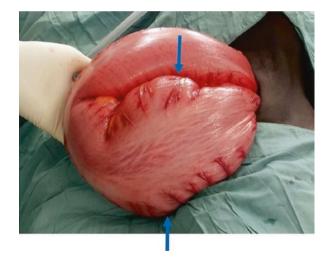
Hirschsprung's disease is less common overall than anorectal malformations, at approximately 1 in 5,000 births. It is marked by congenital aganglionosis of the intestinal tract and generally limited to rectosigmoid region (75–80%). In limited resource settings, a delayed presentation of constipation, abdominal distension, and failure to thrive is the general rule for rectosigmoid disease [69–71] (Fig 20.15a, b). Patients with longer segment disease generally present in the neonatal period. Older patients have generally been treated for medical causes of distension such as parasitic infestations or tuberculosis. Historically, the disease was treated in three stages, with initial decompressing colostomy, a subsequent rectosigmoid resection and pull-through, and, finally, colostomy takedown. In most well-resourced centers, a one-stage procedure is now performed, but frequently this is not possible when a patient presents with long-standing constipation and neglected disease as is the general rule in the resource-limited environment.

At initial evaluation, the patient should be examined for signs of peritonitis and enterocolitis. Enterocolitis is the primary cause of death in Hirschsprung's disease. Any patient presumed to have enterocolitis should be treated with broad-spectrum antibiotics, rectal decompression, and irrigations, as well as bowel rest. A nasogastric tube may be necessary for temporary decompression; laparotomy and fecal diversion may be necessary for inadequate decompression. At laparotomy, there may be a gross transition zone from proximal dilated ganglionic to distal aganglionic bowel (Fig. 20.16), and the colostomy should ideally be created just proximal to this gross transition zone. An ostomy created with aganglionated bowel is unlikely to function. Often the decision will need to be made without intraoperative pathology availability.

In the newborn period, the patient may present with signs of bowel obstruction and failure to pass meconium within 24 h of birth or may present with cecal



Fig. 20.15 Delayed presentation of Hirschsprung's disease: (a) marked abdominal distension and (b) radiograph showing colonic dilation and heavy fecal loading





perforation in the absence of atresia. A rectal examination may stimulate the passage of meconium. Plain X-rays are generally consistent with distal bowel obstruction, and a contrast enema may show a transition zone with dilated bowel proximal to a decompressed distal aganglionic segment. Definitive diagnosis is provided by full-thickness rectal biopsy, usually done under general anesthesia or caudal block. While suction rectal biopsy is favored in many better-resourced centers, this is generally unavailable in a limited resource setting. The biopsy should be taken approximately 2-2.5 cm proximal to the dentate line and in the posterior midline. Biopsies taken too high risk intraperitoneal perforation and too low risk the biopsy of the normal zone of hypoganglionated bowel just proximal to the dentate line. An anterior biopsy risks urethral (male) or vaginal (female) injury and should not be performed. Hallmarks of Hirschsprung's disease are the absence of ganglion cells and hypertrophied nerve fibers in the submucosal and myenteric plexuses. One of the greatest challenges in the limited resource environment might be the absence of reliable pathology services to interpret biopsy results. In these cases the surgeon will have to use best clinical judgment. Ideally, definitive pull-through should not be embarked upon without histologic diagnosis but can be safely performed in areas remote from pathology support services if surgical support is adequate. Intraoperatively, a gross transition zone can be a guide. For the neonate with cecal perforation, the site may be oversewn and a transverse colostomy created.

The goal of definitive surgery is the removal of aganglionic bowel and the restoration of bowel continuity. The three common operations described are the Soave, the Swenson, and the Duhamel. A detailed description of each of these is beyond the scope of this chapter. No one procedure has proven superior to others; thus, the surgeon should perform the procedure with which he or she is most comfortable or safest given the local environment. In the absence of safe neonatal anesthesia, the creation of a colostomy allowing the child to grow to an older age before definitive repair may be an acceptable alternative. For the patient presenting with massive fecal loading (Fig. 20.15b), on-table fecal washout will be necessary at the time of exploration and colostomy creation, and a mucous fistula should ideally be created at this operation to facilitate continued postoperative washouts. If the proximal colon is markedly dilated and loaded with much feces, it may be safer to resect most of this segment to reach the colon with manageable caliber. This will facilitate postoperative care and subsequent pull-through.

Patients will likely be encountered who have had a stoma created for "suspected Hirschsprung's" due to a clinical history of constipation, failure of meconium passage as a neonate, and functional bowel obstruction. In the interim many patients may develop complications such as colostomy stenosis (Fig. 20.17). This occurs as many colostomies may be created simply with a small antimesenteric incision (sewn to the skin) that are not fully defunctioning, leading to distension and bowel obstruction due to spillover. Massive prolapse (Fig. 20.18) may also be encountered and need to be addressed prior to or during a definitive procedure.

Even after a definitive pull-through procedure, patients with Hirschsprung's disease are at risk of enterocolitis, and this should be stressed to caregivers. A regimen of dilations should be prescribed as described above for anorectal malformations. Most patients are followed at least to age 5, but many will go on to require bowel management even after this period.



Fig. 20.17 Stoma stenosis in Hirschsprung's disease

Anorectal Malformations

The Essentials

- Anorectal malformations can present in a broad spectrum and often require a divided colostomy as an initial procedure.
- A three-stage procedure is often required, with anoplasty, followed by stoma takedown.
- Previous experience and familiarity with anoplasty if to be undertaken by the visiting surgeon.
- Associated VACTERL anomalies should be evaluated.



Fig. 20.18 Massive stoma prolapse in Hirschsprung's disease

Anorectal malformations are one of the most common congenital anomalies encountered in neonates and children in limited resource settings, with an estimated incidence of 1 in 3,500 births. There is a wide spectrum of disease, ranging from limited perineal anomalies to complex cloacal anomalies requiring more complex reconstruction. Familiarity with the variations in presentation is essential [72] (Fig. 20.19). Most anomalies other than perineal fistula can be approached through operative repair in stages, with colostomy, followed by definitive repair (posterior sagittal anorectoplasty) and then colostomy takedown after several months.

Anomalies are generally classified as high or low. In male patients, the most common anomaly is imperforate anus with rectourethral fistula, while in female patients, it is a vestibular fistula. Only approximately 5% of male infants will have imperforate anus without fistula. Most babies with no anal opening at all will be referred for surgical evaluation within 24–48 h of birth. Even if not diagnosed by trained birth personnel in a rural area, abdominal distension and failure to pass meconium will generally prompt parents to seek medical care. However, in LMICs,

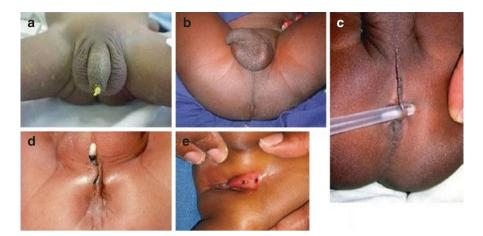


Fig. 20.19 Spectrum of presentation of anorectal malformations

delay in presentation is common. The construction of a colostomy beyond this time period can be very challenging due to progressive abdominal distension, and perforation can occur in cases of further surgical delay. Other patients with an abnormally positioned anal opening who are able to stool with some difficulty may have a delayed presentation to later infancy or childhood. Sometimes these patients are treated unsuccessfully for chronic constipation due to presumed medical causes and suffer great morbidity. Girls with an uncorrected anteriorly displaced anal opening or rectovestibular fistula may live their whole lives with this anomaly. Some practitioners have raised concerns about future vaginal delivery in these patients; however, studies with sufficient long-term follow-up are lacking.

Delay in presentation and complications of anorectal malformations are the rule, either pre- or postoperatively [70]. A common scenario might be one in which the surgeon is asked to "close a colostomy" that was created in the neonatal period, before the anoplasty has actually been done. In the acute neonatal setting, in the absence of an anal opening, a full physical examination should be performed. Anorectal malformations are part of the VACTERL syndrome, which includes vertebral anomalies, cardiac defects, renal anomalies, and limb anomalies. Cardiac auscultation may reveal a murmur; if available, an abdominal ultrasound should evaluate for renal structural anomalies. In the absence of hemodynamic instability, a formal echocardiogram may not be indicated. A careful perineal exam should be performed, noting the nature of the perineum, the integrity of muscle formation, and palpable sacral defects. Generally, a "flat" bottom with poor muscle formation and sacral defects is associated with a high malformation, while a well-formed perineum is more suggestive of a low malformation. For an abnormally located anal opening in the perineum (i.e., perineal fistula), the size should be noted with the knowledge that most normal neonatal anal orifices accommodate a size 10–12 Hegar dilator. In girls, all orifices should be examined to ensure there is not a single orifice (cloacal anomaly, the rarest malformation).

An abdominal X-ray is likely to show distally dilated intestinal loops; a crosstable lateral X-ray (an invertogram should not be done as vomiting and respiratory complications may occur) may be used at 8–24 h postnatally to estimate the distance from the rectum (should be visible with a column of air) to the perineal skin (marked with a radiopaque marker). If this distance is less than 1 cm, an experienced surgeon may choose to perform an anoplasty rather than a colostomy. If referred in the first day of life, one should wait 24 h before the construction of a colostomy in case meconium passes in this time period to reveal a low malformation that can be primarily treated with a perineal procedure. The presence of a "buckethandle" anomaly on physical examination (Fig. 20.19c) suggests a low malformation that would be amenable to anoplasty, and sometimes a small previously undetected opening is revealed by gently using a small probe.

In cases of a large rectourethral fistula (as sometimes apparent by meconium staining of the urine), the baby may pass meconium in the urine, and the bowel may rarely be decompressed with a Foley catheter passed through the urethral opening.

The creation of a colostomy for the newborn with acute abdominal distension with no anal opening may be a lifesaving procedure. As recommended by Pena et al., this is done with a left lower quadrant incision [72]. The sigmoid colon is generally extremely distended on entry to the peritoneal cavity, and it may need to be decompressed prior to exteriorization. This can be done by placing a purse-string suture and decompressing with a 25 gauge needle. The proximal and distal ends of the sigmoid colon should be clearly identified, and the bowel should be divided. A divided colostomy is favored over a loop due to concern about partial diversion and spillover of stool. In addition, the colostomy should be constructed in the first mobile portion of the sigmoid colon (just after the descending colon) to allow maximum bowel length for the later pull-through procedure. The distal colon-rectum should be washed out at the time of colostomy creation. The baby can then start feeding when the colostomy is functional and the abdomen decompressed. Most families in austere settings will not have access to stoma supplies, and providers and patients usually improvise to care for the stoma with reusable materials such as napkins.

Prior to the definitive surgery, a distal colostogram should be done to identify the level of the malformation as well as the location of any fistula. Typically, no fistula may be identified as it might be plugged. In the absence of fluoroscopy, a series of X-ray images can be obtained using water-soluble contrast flushed through the distal colon. If barium is used, it should be flushed out to prevent impaction in the distal colon.

Approach to the definitive pull-through procedure depends on the type of malformation. In males, high malformations (bladder neck fistula) will need an abdominal approach to ligate the fistula (if present) and mobilize the rectum, while low malformations can be approached through the perineum only through the standard approach of Pena (Posterior Sagital AnoRectoPlasty or PSARP) [72] (Fig. 20.20). The goal of the operation is to fully mobilize the rectum, divide the fistula (if present) without urethral injury, and replace the rectum within the borders of the sphincter complex. Muscle stimulators to define the borders of the sphincter complex are



Fig. 20.20 Posterior sagittal anorectoplasty

expensive and may not be available locally; therefore, the visiting surgeon should either bring one or assemble a low-cost but effective alternative [10, 70]. The use of diathermy set at low voltage may also be helpful in identifying the borders of the sphincter complex. Anesthesiologists in some settings may also have a nerve stimulator that can be used.

In girls, for the most common malformation, a vestibular fistula (Fig. 20.19e), the surgeon must decide whether to perform a colostomy initially, with the repair (to "protect the repair"), or primary repair without diversion. There is also lack of consensus on the appropriate timing for surgery, whether in the neonatal period or delayed until the patient is several months old. This is dependent on the comfort of the surgeon in performing this procedure in the neonatal period and the condition of the patient. However, if there are any anesthetic limitations, it may be safer to do the PSARP after several months, when anesthesia is safer. A newborn who breaks down her repair may need diversion and possible redo surgery. Deep infection at the level of the repair also may result in incontinence—the best opportunity to achieve good results is at the first operation.

Postoperatively, rectal dilations should commence 10–14 days after surgery and can be continued at home by the child's caregivers. As Hegar dilators are generally



Fig. 20.21 Mohan's valvotome for treatment of posterior urethral valves

unavailable, a smooth appropriately sized candle covered by a glove with lube is a cheap suitable alternative for parents to continue dilations at home, as are other soft locally available supplies such as rounded pen end and the caregiver's gloved little finger. For children beyond several months of age, dilations are poorly tolerated, and most will put up quite a fight. As the anastomosis can scar down, follow-up is critical and this must be stressed to families. Caregivers can also be taught digital dilation. These patients need to be followed through infancy and childhood as solid food is commenced and also through the period of toilet training. Children with low malformations have a tendency to constipation, and those with higher malformations tend more toward incontinence. Continued follow-up over these periods is critical as dietary modification and the introduction of laxatives or constipating agents may be necessary. Anal stenosis may also produce these symptoms, so stenosis should always be excluded before taking a decision about treatment that has significant social and psychological consequences. In excluding stenosis, it should be noted that it may not always be at the anal verge and may be located slightly above this level.

Genitourinary Anomalies

The Essentials

• In settings where treatment equipment is not available, posterior urethral valves may need to be treated with vesicostomy.

These include posterior urethral valves (PUV) and bladder exstrophy. PUV present with obstructed bladder outlet and manifested by poor stream, repeated urinary infections, and even renal failure. The diagnosis is suspected by ultrasound which typically shows a thick, dilated, trabeculated urinary bladder, sometimes with associated upper tract dilatation. Confirmation is obtained by voiding cystourethrogram (VCUG) showing the posterior (proximal) urethral obstruction with dilated proximal urethra and bladder neck. Definitive treatment relies on endoscopic valvulotomy, but treatment of the newborn often lacks the appropriate instrument sizes and thus needs to be temporized until the baby grows or can be referred to a facility able to treat him/her. Temporization is best done with a formal vesicostomy approximating the bladder mucosa to the skin, thus alleviating the need for an indwelling catheter. The Mohan's valvotome (Fig. 20.21) is cheap and safe and can be used in the

neonates. The oft-quoted practice of blind valve ablation using a Foley catheter balloon is dangerous and cannot be recommended.

Bladder exstrophy is a rare condition in which the anterior wall of the bladder and the abdominal wall overlying it are absent. The defect is obvious at birth and is surprisingly tolerated quite well by the children, with minimal risk of infections but tremendous social disability and stigma. The repair of the condition is not urgent but requires complex procedures. In the neonatal period, primary closure is indicated, often accompanied by iliac osteotomies with external fixation. After a couple of years of age, restoring continence becomes extremely unlikely even after multiple procedures, and the choice in limited resource settings is of a urinary diversion procedure, typically in the form of a continent ureterosigmoidostomy such as the Mainz pouch II procedure [73]. A visiting surgeon without appropriate expertise is best advised to direct these patients to a regional institution able to handle them (often in another country).

Thoracic Neonatal Conditions

The Essentials

- Esophageal atresia most commonly presents with excessive salivation and inability to pass an NG tube, with a distal tracheoesophageal fistula the most common type.
- Thoracotomy with fistula ligation and repair is the standard of care, but outcomes in the absence of critical care support are generally poor in LMICs.

The most common thoracic condition requiring intervention in the neonatal period is esophageal atresia, with the most common form being proximal atresia (EA) with distal tracheoesophageal fistula (TEF). While this anomaly is identified in the first day of life in most high-income countries, often diagnosis is delayed in LMICs and presents with no visible anomaly, but the baby has excessive salivation and inability to feed. Attempts to pass a nasogastric tube often reveal obstruction, with a chest X-ray showing the tube in the upper thorax. The presence of distal air in the gastro-intestinal tract confirms the presence of a TEF. While multiple other types may be found, this is the most common type.

Unfortunately, mortality for this condition remains very high in low- and middleincome countries, often due to delayed presentation, anesthetic limitations, poor nutrition, and limited capacity for intensive and perioperative care. The most acute problem is typically the tracheoesophageal fistula that can cause abdominal compartment syndrome and also the accumulation of saliva in the proximal esophageal pouch that can lead to aspiration. Repair of these anomalies is best performed at a tertiary care center with intensive care capacity. Stabilization includes a small bore (8 French) orogastric tube on continuous suction in the proximal pouch and IV fluid resuscitation. In high-income countries, the most common congenital neonatal condition encountered is a prenatally diagnosed cystic lung lesion, i.e., cystic pulmonary airway malformation (CPAM). Postnatal management in asymptomatic patients remains controversial though excision is generally performed in the first year of life due to risk of infection and (rare) risk of malignancy. These conditions are much more uncommon in resource-poor areas due to lack of antenatal imaging and due to the asymptomatic nature of the lesion.

Surgical Emergencies in Older Children

Appendicitis

The Essentials

- History and exam may mandate exploration in the absence of diagnostic tests.
- Perforation may be more common in resource-poor areas and requires exploration through right lower abdominal incision or laparotomy.
- A stable patient with a possible phlegmon may be treated with antibiotics with plans for interval appendectomy.

Presentation of appendicitis in the child is quite similar to adults. In North America and Europe, the lifetime incidence of appendicitis is estimated to be 7–8%; in Africa, it is estimated to be lower, closer to 1–2%, perhaps due to variation in diet and the immune system. Peak incidence is in the 12–18 age group and rare in neonates and infants [74].

In most austere settings, the decision to operate for appendicitis will be based on history and physical examination alone, without the aid of any ancillary tests. However, imaging can be helpful. Plain films may show a fecalith in the right lower quadrant or an obstructive pattern. Ultrasound may be highly sensitive and specific, though this is mostly observer dependent. In resource-rich environments, perforated appendicitis with abscess is generally diagnosed on CT scan, and imaging-assisted drainage may be performed with a plan for interval appendectomy several months later. This approach for perforated appendicitis is not possible in most resourceconstrained settings without interventional radiology capacity, and nearly all such patients will require surgical intervention. Moreover, most patients with perforated appendicitis in these settings will have general peritonitis at presentation, making initial nonsurgical approach inappropriate.

In the child <4 years old, presentation with perforation is the rule, due to both the difficulty for young children to describe mild symptoms and the fact that the omentum in young children is thinner and less able to contain infection. The patient with symptom duration >3 days who presents with a palpable right lower quadrant mass, but is pain-free and tolerating a diet, may be treated with antibiotics for 7–10 days and presumed to have appendicitis with contained perforation. This may be then followed up with interval examination and then appendectomy as indicated. A rectal examination should be performed in patients presumed to have perforation and abscess, and if initial nonoperative therapy is considered, transrectal drainage of a pelvic abscess can be performed under sedation.

An additional challenge is that fever, abdominal pain, and other symptoms similar to those of appendicitis are common in malaria-endemic regions due to improper use of antimalarials. These medicines are readily available at pharmacies without a prescription, and parents may not seek medical attention until their child has not responded to a full course of therapy. Alternatively, they may first see a traditional healer. These factors contribute to the frequent advanced presentation of diseases for acute abdominal conditions. In addition, the broader differential diagnosis for acute abdominal pain in children in the tropics must be considered, including, for example, intestinal perforation from infectious causes such as typhoid fever, infectious complications of undernutrition or immune suppression, as well as spontaneous bacterial peritonitis.

In the patient with diffuse peritonitis, a midline approach may be chosen to allow for adequate exploration and washout. In children the appendix may be ligated with a simple or purse-string suture and dissected either antegrade or retrograde depending on the ease of dissection. Drains may be useful in localized abscess cavities. In the absence of formal Jackson-Pratt drains, other available tubing or a corrugated rubber sheet may be used.

In extreme cases where the appendiceal stump is liquefied and the surgeon is concerned about the development of a cecal leak and subsequent colocutaneous fistula, a diverting loop ileostomy may be constructed, though this can have significant morbidity in an austere setting. A tube cecostomy, perhaps with a Foley catheter, may be more helpful and easier to manage in such a case.

Intussusception

The Essentials

- Intussusception is most common in the 3-month to 3-year age group and is frequently mistaken for a medical gastroenteritis.
- Crampy intermittent abdominal pain is typical and ultrasound the gold standard for diagnosis, though a suspicious history and physical requires exploration.
- Prior to laparotomy adequate resuscitation is needed including IV fluids, nasogastric tube decompression, and antibiotics.
- While enema fluoroscopic reduction is common in HICs, laparotomy with reduction and resection is often required in LMICs, with a temporizing stoma often required due to patient status.

Intussusception occurs most frequently in the 3-month- to 3-year-old age group and is most commonly idiopathic in origin, frequently due to a preexisting viral



Fig. 20.22 Intussusception protruding from the anus

illness that may have produced intra-abdominal lymphadenopathy. These mesenteric lymph nodes, usually in the terminal ileum, are the most common lead point of the intussusception. In infants with gastroenteritis, vigorous peristalsis may also precipitate an intussusception. The intussusception is generally ileocolic (approximately 80–90%) but can extend as far as the rectum or colon. In a minority of cases, the intussusception may be colocolic or limited to the small bowel. It is one of the most common causes of acute abdomen and bowel obstruction in children in poor countries [75, 76].

Classically, children present with fits of intermittent crampy abdominal pain, (where the child draws their knees up to the chest), with intervening periods of normal behavior. It may or may not be possible to elicit a prior history of viral illness. There may be bilious or non-bilious vomiting, or "currant-jelly stools," (bloody mucoid stools) an indicator of possible mucosal compromise. On physical examination, the abdomen is generally distended. The presence of peritoneal signs should raise concern for possible bowel compromise and mandates operative exploration. The child may have visible signs of dehydration and sepsis. Other causes of bowel obstruction should be excluded on physical examination. With early presentation, an abdominal mass

may be present. A rectal examination should be performed as well to evaluate for blood in the stool, and in the most extreme cases, the intussusception may protrude through the anus (Fig. 20.22), initially perhaps raising the question whether this may be rectal prolapse. Vital signs may reflect dehydration or septic response.

Plain abdominal films may show distended bowel loops consistent with intestinal obstruction or, in early cases, may be more normal appearing. Ultrasound may be confirmatory if the characteristic features of target and pseudo-kidney signs are present. With late presentation, pneumoperitoneum may be present. A contrast enema, in the hands of an experienced radiologist, may be therapeutic in up to 80% of the time; however, this modality is frequently unavailable in a resource-limited setting. Moreover, contrast enema is contraindicated if there are features of intestinal strangulation, which is common in these settings. The medium of the enema may be pneumatic or hydrostatic and is generally performed with the aid of fluoroscopy or ultrasound. If an enema is available in the local environment and the first attempt does not fully reduce the intussusception, it may be attempted again if some progress was made on the first attempt, in the absence of peritoneal signs or hemodynamic instability. In resource-limited settings, surgeon-performed pneumatic reduction enema may also be performed safely in the operating room under general anesthesia [77]. Rarely, reduction by enema can result in perforation and in extreme cases with abdominal compartment syndrome requiring emergency decompression with an 18 gauge needle at the bedside, prior to transport for laparotomy. It should be stressed that the presence of peritoneal signs and clinical signs of sepsis mandates laparotomy. Aggressive resuscitation and close monitoring of urine output are important.

In this age group, a primarily right-sided transverse or midline laparotomy may be performed. The bowel is delivered and the intussusception identified. The intussusception should be reduced by milking the intussuscepted bowel from the distal end proximally rather than trying to "pull it out" at the site of intussusception, as this may result in tearing of the bowel. Once reduced, the bowel should be evaluated for viability. Nonviable bowel should be resected and an anastomosis performed between ends of well-perfused bowel. In cases of systemic sepsis or poor perfusion, creation of a temporary ostomy may be the safest option after bowel resection rather than risking anastomotic breakdown. In addition, patients must be closely observed postoperatively in cases of manual reduction without resection, as reduced bowel that appears viable intraoperatively still has risk of necrosis postoperatively, especially in the poorly resuscitated patient. In such cases, postoperative decompensation mandates exploration to rule out bowel necrosis and perforation. Intestinal malrotation may coexist and should be excluded intraoperatively.

In cases where the intussusception is reduced nonoperatively, there is an approximately 10% risk of recurrence, and this should be stressed to the parents. In some cases, also, the intussusception may reduce on its own, without any intervention. Intussusception in the older child raises the question of a pathologic lead point, and this generally requires laparotomy for definitive diagnosis. The most common pathologic lead points are Meckel's diverticulum or polyps of the small and large bowel but may also be malignant conditions such as small bowel lymphoma. As with other surgical conditions in the resource-limited setting, patients often present with high-grade bowel obstruction and severe dehydration, and this compromises outcomes, with mortality rates of 8–50% reported in these settings, compared to negligible mortality rate in high-income countries [78].

Typhoid Perforation

See Chap. 15 Tropical Infectious Disease Medicine for Surgeons – A Primer section on Typhoid fever.

Acute Scrotum

The Essentials

- Testicular torsion requires prompt recognition and surgical treatment to avoid testicular loss.
- Exploration may either be through a midline raphe incision or an ipsilateral scrotal incision.
- Detorsion and orchiopexy should be performed or orchiectomy in cases where the testicle is necrotic.
- Contralateral orchiopexy should be performed.

"Acute scrotum" is a term used to define sudden onset of scrotal pain, which may or may not be associated with scrotal swelling and or erythema. The most crucial step in management of any acute scrotum is the timely recognition of testicular torsion. Any degree of ischemia is associated with impaired testicular function and morphology. There is a 4–8 h window before significant damage to the testis occurs, but this depends on the degree of twist. Testicular torsion is primarily a disease of neonates and adolescents though it may occur at any age.

Intravaginal torsion (IVT) describes the torsion that occurs in the "bell clapper" anomaly, whereby the tunica vaginalis has a high attachment onto the spermatic cord resulting in a long mesorchium and highly pendulous and mobile testis that is susceptible to torsion. This is present in 12% of males. In extravaginal torsion (EVT), the entire testis complex, which hasn't yet fused to the scrotal bed, twists en bloc. In addition, vestigial appendices may occur anywhere along the testis, and these may undergo torsion, independent of the testis (including appendix testes or appendix epididymis). Most neonatal cases are an extravaginal torsion.

A cremasteric spasm associated with trauma, vigorous exercise, cold weather, or nocturnal erections is the most common trigger. Direction and extent of torsion affect degree of ischemia.

At birth, a hard swollen, non-tender edematous, and dusky hemiscrotum that doesn't transilluminate may be noted, indicative of prenatal torsion. Meanwhile, a child with a painful, edematous, and erythematous hemiscrotum with history of a normal scrotum

at birth suggests postnatal torsion. A child who initially had a palpable testis but with time the testis is non-palpable ("vanishing" testis), is usually due to atrophy of a testis that underwent torsion in late gestation. Fifty percent of patients with acute torsion have experienced previous episodes of testicular pain that resolved spontaneously, after a short time, suggesting that "intermittent" torsion is also possible.

In cases of "acute scrotum" in which testicular torsion is highly suspected, time is of the essence! History taking and physical examination are sufficient to make an accurate diagnosis in 60–90% of cases. The following features are most suggestive:

- 1. Pain, rapid and severe, can be associated with nausea and or vomiting with inability to stand straight. This pain however may begin to dissipate after 6 h.
- 2. Pain can radiate to the medial thigh and lower abdomen.
- 3. Previous episodes of self-limiting scrotal pain ("prophetic pain") are suggestive of intermittent torsion.
- 4. Loss of the cremasteric reflex on the affected side (negative predictive value of 96% for testicular torsion).
- 5. A high-riding testis, close to the external inguinal ring due to shortening of the cord, is present in 26–80% of patients.
- 6. Angell's sign is present in 25–90% of patients—this refers to the "horizontal lie" or positioning of the contralateral testis when patient is examined in the upright position.
- 7. Prehn's sign refers to elevation of affected testis in comparison to the contralateral testis.
- 8. Fever is an ominous sign indicative of infarction.

Investigations should not delay intervention, as time is of the essence and may delay testicular salvage surgery. Investigations can be used if diagnosis is equivocal, and symptoms are of a longer duration than hours. Ultrasound is suggestive but is operator dependent, and detection of blood flow doesn't necessarily rule out torsion.

Key principles of management are as follows:

- 1. Manual derotation (if early presentation); two-thirds of patients have internal rotation, and manual derotation in the external rotation direction (opening a book) can be attempted. If this fails, derotation in the opposite side can then be attempted. Sedation and spermatic cord blocks are discouraged as one cannot determine the success of the maneuver if the patient cannot report "relief" of the symptoms.
- 2. Surgical exploration of the scrotum; a single incision through the median raphe allows exploration of both scrotal compartments. Alternatively, mid- to upper scrotal incisions can be used.
- If the testis is obviously necrotic, orchiectomy/orchidectomy should be done, and lavage of scrotal pouch is recommended. A future silastic prosthesis can be considered if available.

- If the testes fail to regain pink/red color after derotation/detorsion, and only dark/ black blood oozes from a nip on the tunica albuginea, then orchiectomy/orchidectomy is indicated.
- 5. After untwisting, if the testis regains some mottled color after wrapping in warm saline for 5–10 min, with red bleeding from the tunica albuginea, the testis should be preserved.
- 6. Once found salvageable, the affected and the contralateral testes should be fixed in a sub-dartos pouch using absorbable sutures at three points. Avoid silk, as it may lead to abscess formation. Eversion of the tunica vaginalis combined with a dartos pouch fixation is associated with excellent fixation results.

For torsion of appendices, if the diagnosis is certain, management includes bed rest, scrotal elevation, and analgesia (especially NSAIDS), and inflammation will resolve in 2–7 days. A minority of these patients may require surgery if pain and inflammation persist beyond 2 days. If found, on scrotal exploration, the twisted appendix is simply excised. Failure to fix the contralateral testes may result in torsion in the other testes.

Pediatric Solid Tumors

Wilms' Tumor

The Essentials

- Wilms' tumor more commonly presents with advanced disease in LMICs than HICs.
- Initial diagnosis and staging requires biopsy and ultrasound.
- Advanced stage of presentation often requires neoadjuvant chemotherapy although resectable tumors can be excised initially after staging.
- After neoadjuvant chemotherapy, resection through a transverse incision can be performed.

This is one of the most common solid childhood tumors. In LMICs, it represents about 3–20% of all malignant tumors in children [79, 80]. Patients may present with an incidental abdominal mass or with microscopic hematuria, hypertension, malaise, weight loss, or anemia. Pain with fever and a rapidly enlarging mass may indicate acute tumor rupture. In developing countries, due to late presentation, a large abdominal mass is obvious at presentation (Fig. 20.23). Anemia and malnutrition are present in more than 50% of the patients at presentation [81]. A minority of patients with Wilms' tumors have an associated named syndrome, and those associated features may be present.

Abdominal ultrasound can identify the renal origin of the tumor and can assess the contralateral kidney, and ipsilateral renal vein and inferior vena cava for tumor extension. Intravenous urography may show a distortion of the pelvicaliceal system of the affected kidney, and sometimes there may be non-excretion of contrast. CT scan is most accurate in characterizing the extent of disease, but this is often unavailable or too expensive in LMICs. As a result, IV urography may be used more frequently. A TruCut® needle biopsy of the tumor to confirm histological diagnosis can be done under ultrasound guidance (preferably using a Doppler ultrasound). Biopsy may be required also to exclude Burkitt's lymphoma. Open biopsy risks severe hemorrhage and tumor dissemination and should be avoided. A plain chest film should evaluate for pulmonary metastasis. The tumor should be staged, and the National Wilms Tumor Study (NWTS) or Société Internationale d'Oncologie Pédiatrique (SIOP) staging system can be used. In sub-Saharan Africa, >50% of patients present with stage III and IV disease.

The treatment of Wilms' tumor involves surgery, chemotherapy, and radiotherapy. The NWTS and SIOP protocols can be used. The NWTS protocol advocates that resectable tumors are excised primarily. However, the SIOP protocol may be better suited for the LMIC setting [82]. This protocol consists of administration of preoperative chemotherapy for all patients, using vincristine and actinomycin D (given for 4 weeks and surgery in the fifth week) without histological diagnosis: this is aimed at reducing the tumor size to minimize the risk of intraoperative tumor rupture/spillage and increase the chance of complete resection. Following surgery, chemotherapy is continued based on risk stratification according to tumor histology (favorable or unfavorable) for a total of 18–24 weeks. Goals of surgery are abdominal exploration, complete tumor resection without spillage (in low-stage tumors), nephroureterectomy, lymph node biopsy at the renal hilum, and removal of tumor from the renal vein or inferior vena cava in cases of tumor extension to these locations.

Wilms' tumor is presently considered a curable disease, and long-term survival can be expected for >80% of patients in developed countries. In LMICs, however, the 5-year survival is below 50%, largely due to the late stage at diagnosis, comorbidities, and problems with completion of chemotherapy.



Fig. 20.23 Late presentation of Wilms' tumor

Lymphadenopathy and Lymphoma

The Essentials

- Lymphadenopathy is common and may require incisional or excision biopsy.
- The differential diagnosis is broad and medical workup is needed.
- Larger nodes than 2 cm that are persistent may require biopsy.

The general surgeon is frequently called to evaluate "lumps and bumps" in children, often to establish the malignant potential of these lesions and to decide whether a biopsy is indicated. Most of the time, if there is a reasonable level of suspicion, a biopsy should be performed as these are low-risk procedures that can give high-yield diagnostic information.

Lymphomas are generally classified as Hodgkin's (85% globally) and non-Hodgkin's (15%) type [83]. In sub-Saharan Africa, Burkitt's-type lymphoma is endemic and the most common type of lymphoma in children, often presenting with a mass about the jaw (Fig. 20.24). The endemicity may be partly due to an



Fig. 20.24 Burkitt's lymphoma

association and interaction between EBV and malaria which may also be affected by influence of HIV. In older children, the other forms are more common [84]. Burkitt's lymphoma is one of the "small round blue cell" tumors of childhood, and one of the most rapidly dividing human tumors, with a doubling time of 24–48 h. It is therefore also very chemosensitive, and this lends urgency to diagnosis and initiation of therapy. Generally, the incidence of B-cell lymphomas has increased in high HIV incidence areas, and HIV-related Burkitt's lymphoma has been shown to be less chemosensitive.

Initially, a careful history should be performed asking specifically about size of mass, other possible lymph nodes, and constitutional symptoms such as fever, weight loss, and lethargy. Risk factors for immune compromise should be evaluated, and an HIV test should be obtained. Up to 18% of patients with HIV disease may present with adenopathy [85].

Careful lymph node examination should be performed of all regions assessing whether nodes are fixed or mobile, matted, single, or multiple and whether there is associated abdominal visceromegaly. Nodes greater than 2 cm or those with rapid growth may be considered more suspicious for malignancy. Any node in the supraclavicular region is considered abnormal and suspicious and should be biopsied. In the cervical region, nodes in the posterior triangle are more suspicious for malignancy than those in the anterior neck. Cervical or axillary (the most common locations) adenopathy should ideally be investigated by chest X-ray (to rule out tuberculosis or a mediastinal mass).

With suspicion for infected nodes, a short course of empiric antibiotics may be administered. Without response or if nodes have persisted beyond 4–6 weeks, a biopsy should be obtained. Blood tests are unlikely to alter the need for a biopsy. A single fluctuant tender node is more suggestive of acute suppurative lymphadenitis and can be treated with incision and drainage or aspiration with culture.

Tuberculous lymphadenitis is particularly common in high HIV prevalence areas; even in the absence of TB, lymphadenopathy may be the presenting symptom of HIV disease. This is in distinct contrast to the microbiology in North America, where infection with atypical mycobacteria is more common. BCG vaccination itself can also cause lymphadenitis in 36/1,000 vaccinations.

FNA usually cannot be done in children without adequate sedation, and the aspirate may not provide adequate tissue for flow cytometry required to diagnose lymphoma. An open incisional or excisional biopsy technique is preferred in most instances. In some cases, diagnosis may be obtained by bone marrow biopsy or by aspiration of pleural fluid.

A wide spectrum of infectious and non-infectious diseases can also cause lymphadenopathy and the reader is referred to other sources for a more complete discussion.

Though lymphadenopathy may be the most common presentation of lymphoma in children, and the characteristic jaw mass of Burkitt's lymphoma may be well recognized, other presentations include intussusception and or bowel obstruction, abdominal mass, mediastinal mass, pleural effusion, or splenomegaly [86]. In the rare cases of isolated bowel tumors with Burkitt's lymphoma, a complete resection, if possible, will be essentially curative, though postoperative chemotherapy will still be needed as the disease is multifocal. Most abdominal masses, however, will be unresectable (only a biopsy needs to be taken in most cases), and chemotherapy will be the mainstay of therapy. Staging laparotomy, once part of the management of lymphoma, is no longer indicated. Prognosis will depend on the local availability of chemotherapy programs as well as compliance with therapy [87].

Sacrococcygeal Teratoma (SCT)

The Essentials

- Sacrococcygeal teratomas most commonly present with an exophytic tailbone mass, but some subtypes also have a large pelvic component.
- Full excision including coccygeal resection is required as recurrence and malignant degeneration is possible.

These congenital germ cell tumors are the most common tumor of the newborn period (Fig. 20.25). The classic form consists of a growth over the tailbone that can be very large. The basic classification includes four types: Type I that is predominantly external; Type II that is mixed external and pelvic, but predominantly external; Type III that is visible externally but has a predominant pelvic component; and Type IV that is purely pelvic. As with other teratomas, they are comprised of tissue from all three germ cell layers. Commonly they have a large cystic component. A pelvic ultrasound is recommended to identify a pelvic component if one exists, as



Fig. 20.25 Giant sacrococcygeal teratoma

this may change the operative approach. Most tumors can be excised in a prone position, and must include a coccygectomy, otherwise these tumors can recur. Control of the middle sacral artery is a critical step of the procedure, and bleeding from this vessel can be life-threatening. Excision in the newborn period is recommended as these tumors are benign but have a propensity for malignant degeneration if untreated or if they are incompletely excised. Unfortunately, in resource-poor areas, in addition to delayed presentation, poor identification of the disease, treatment such as incision and drainage of a presumed sacral abscess, or incomplete excision due to lack of a coccygectomy can lead to significant complications.

Elective Conditions in Children

Hernias and Hydroceles: Pediatric Aspects

The Essentials

- Pediatric hernias do not self-resolve and should be repaired by high ligation through a groin incision.
- Incarcerated hernias require reduction and subsequent repair.
- Non-communicating hydroceles require repair after age 1 and can be approached in a similar fashion to inguinal hernias.

Hernias and hydroceles are very common surgical conditions in children, regardless of context, affecting up to 1-5% of term children and a higher percentage of premature babies. Hernias are more common on the right side, more commonly affect males, and 99% are indirect hernias due to a patent processus vaginalis. Hernias in children are more commonly associated with some conditions such as abdominal wall defects and in children with ascites. Incarceration is more common in the neonatal period and possesses risk not only to the bowel but to the ipsilateral testicle (atrophy due to ischemia or frank necrosis).

Generally, parents will report that the baby has had an intermittent bulge on either side. A careful physical exam should be performed. Both testicles should be examined to ensure they are descended, the genitalia should be examined, and the placement and size of the anal opening should be evaluated. If no hernia is initially palpated or seen, the examiner can induce increased abdominal pressure by gently holding the baby's arms and/or legs down or putting gentle, downward pressure in the suprapubic region. Transillumination may help to detect a hydrocele; if this is felt to be a noncommucating hydrocele (no history of change in size), then operation can be deferred to age one, as some of these may self-resolve (Fig. 20.26). In addition, a "silk glove sign" may be present with a thickened hernia sac; this is detected by rolling the cord structures over the ipsilateral pubic tubercle.

Timing of repair is important: ideally, pediatric hernias are repaired close to the time of diagnosis to minimize chance of incarceration; however, in a resource-poor setting, the availability and safety of neonatal general anesthesia should be considered



Fig. 20.26 Congenital hydrocele

as general anesthesia is preferred. Ketamine may also be used. Controversy remains over routine contralateral exploration, but most large retrospective series suggest that only a minority (7%) of children will develop a contralateral hernia.

A standard high ligation at the internal ring should be performed; this is the most important part of the procedure. Given the high preponderance of indirect hernias, a floor repair (i.e., Bassini or other) is generally unnecessary. In infant girls in particular, approximately 20% may contain the fallopian tube or ovary as part of a sliding hernia, and the surgeon should be prepared to invert the sac around a purse-string suture at the internal ring if necessary. In infants, the external ring and internal ring are nearly superimposed, and it may be unnecessary to open the aponeurosis of the external oblique muscle upon entry. For large hernia sacs that extend into the scrotum, scrotal extent of the sac does not need to be excised. This may cause excessive bleeding and traumatize the cord structures. At the conclusion of the operation, if the testicle has been lifted out of the scrotum, it should be returned to its normal location to minimize the chance of iatrogenic cryptorchidism, a rare complication. In the case of transection of the vas deferens, it should be repaired with fine absorbable sutures. These operations are more challenging in infants and young children due to the small size of the vas and vessels and, often, a delicate hernia sac. The bladder in an infant is an intra-abdominal organ and at risk for injury, especially with an incision that strays medially.

If an incarcerated hernia is detected clinically, attempts should be made at reduction, ideally with sedation and analgesia. Gentle pressure is usually able to reduce the hernia, and the patient should be observed at least for 4–8 h to ensure that feeds are tolerated and that peritonitis does not develop. Ideally, the hernia repair should be performed in the next 2–3 days, allowing for some of the tissue edema in the cord to abate. Even several days later, the tissues and sac are likely to be extremely friable. In cases of suspected strangulation, and possible bowel compromise, reduction should not be attempted, and as in adults, a groin exploration should be performed to evaluate the bowel. If this cannot be adequately performed through the groin, a laparotomy may be necessary.

Undescended Testes

The Essentials

- Intervention for undescended testicles is delayed until at least a year old to allow for possible descent.
- Role of ultrasound is controversial for non-palpable testicles.
- For inguinal testicles, a two-incision groin approach is used, with mobilization through the upper incision and orchiopexy through the lower incision.
- Abdominal testicles require exploration and orchiopexy or orchiectomy if nonviable.

Cryptochidism (undescended (UDT) or maldescended testis) is one of the commonest urogenital malformations in the male pediatric population worldwide. Its etiology is multifactorial and still quite obscure. If "undescended" the testis may be regarded as palpable or not palpable, retractile or "peeping," ascending, and ectopic or acquired UDT which may be iatrogenic (e.g., postinguinal hernia repair). Occasionally, the child may have anorchia. This variability in nomenclature poses a diagnostic dilemma, which may require maneuvers like squatting, increasing abdominal pressure, and sitting cross-legged as the physicians run their fingers from the iliac crest to the scrotum in order to classify the condition. If both testes are not palpable, hormonal testing to stimulate evidence of the presence of adequate testosterone levels may be required, prior to attempting exploration; however, this test may not be available in most resource-poor areas.

The incidence of UDT is higher in premature or low birth weight infants, but these testes are usually expected to descend by 1 year of age, beyond which, spontaneous descent is deemed unlikely, and orchiopexy is warranted. The coexistence of UDT with other congenital anomalies is fairly common as is an associated inguinal hernia, and these must be addressed in the same sitting [88].

Cryptorchidism is one of the most common visible urogenital malformation among African newborn males and school boys and sometimes in resource-poor areas, even in adolescents. While many patients present at age >5, occasionally the patient may even present in their early twenties. If there is a coexisting hernia or if the testis is trapped in the inguinal canal, the mother may report an incidental, nontender, inguinal swelling. Rarely, cryptorchidism is an incidental finding by the pediatrician or general practitioner [89].

The use of ultrasound in localizing testis in children with UDT in LMIC is controversial. Ultrasound is not necessary in cases where the testis is palpable. It may be requested in cases of a non-palpable testicle, but its sensitivity has been questioned in some studies [90].

Once a diagnosis of UDT is made, orchiopexy is the logical next step in children >1 to ensure fixation of the testes in the scrotum for optimum function. Other indications (besides possible fertility benefits) for orchiopexy include (1) prevention of future torsion, (2) detection for cancer screening, and (3) less likelihood of traumatic injury.

The inguinal approach is most common for the palpable UDT, with takedown of the gubernaculum and a herniotomy to obtain sufficient cord length to plant the testis in the scrotum and its fixation in a sub-dartos pouch. A hernia-type incision is made to identify and mobilize the testicle, and subsequently, an upper or lower scrotal incision is made to create the pouch. Usually absorbable sutures are used. A Bassini-type approach to reinforce the floor of the inguinal canal in cases where there is a large associated direct hernia may be done.

If there is difficulty obtaining adequate length, the proximal cord may be further mobilized, and potential ligation of the epigastric vessel may be helpful. If the testes cannot reach the base of the scrotum, it should be left as low in the inguinal canal as possible to facilitate future physical examination to monitor for malignancy. If there is a very small testicle, or a "nubbin," this should be removed. In cases of nonpalpable testicle, an exam under anesthesia should be performed. A minilaparotomy may be needed in the absence of laparoscopy, and the testicular vessel may need to be ligated for adequate length to bring it to an extra-abdominal location (in the presence of a normal contralateral descended testicle). The family should be counseled that this testicle may or may not be viable, and the blood supply will depend on adequate collateral supply from the vas deferens.

Immediate post-operative complications may include scrotal hematoma, and early recurrent inguinal hernia. Ideally, children after orchiopexy should be followed for at least a year to be sure that the testicle remains descended.

Head and Neck Conditions

The Essentials

- Lymphatic malformations are common and require intervention due to risk of infection and hemorrhage, though intervention is nonemergent and may wait to 6 months to 1-year-old.
- Distinguishing between macrocystic lesions and microcystic lesions can be helpful, as sclerotherapy may be an effective first-line treatment for macrocystic lesions.
- Resection should not compromise critical neurovascular structures.
- Branchial cleft remnants are present at birth and may or may not be symptomatic.
- Resection is nonurgent and can be deferred to 6 months to 1-year-old.
- Resection often involves following the tract to the oropharynx to minimize chances of recurrence.

The most common pediatric neck masses are enlarged lymph nodes, and lymphoma would be the most concerning entity in the differential diagnosis. In the neonatal period, lymphatic malformations (formerly known as "cystic hygromas") may be encountered. The type and timing of intervention depends mostly on the severity of clinical symptoms. Lymphatic malformations may range from small cystic masses



Fig. 20.27 Large cervical lymphatic malformation treated by sclerotherapy with bleomycin. At presentation at age 7 days, after 10 months, and after 20 months

to large disfiguring lesions that can compress and even involve the airway. They are generally classified as being either "macrocystic" or "microcystic" depending on the type of cysts involved—with large unilocular cysts considered "macrocystic." This affects management in that macrocystic lesions may be temporized by aspiration (with nearly 100% chance of recurrence), while microcystic lesions often behave as solid tumors and do not respond to aspiration due to many septations between small cysts. Ultrasound is a reasonable first imaging study and can often confirm the cystic nature of the lesion and identify septations and size of spaces to determine whether the cyst is microcystic or macrocystic. This can also confirm (using Doppler) that there is no significant vascular component of the lesion, as sometimes lymphatic malformations can have a mixed arterial or venous component. In resource-poor areas, optional further definition of the lesion with CT or MR imaging may not be available.

As with other neonatal conditions, in settings with limited resources, deferring invasive surgery to an older age when anesthesia is safer is preferred. Urgency of intervention would be determined primarily by the severity of symptoms, especially of airway compression, and feeding. The general indications for intervention for these lesions are (1) infection, (2) bleeding, (3) risk of growth and impact on adjacent structures, and (4) cosmesis. Lymphatic malformations are not believed to be a risk factor for malignant degeneration, although lymphangiosarcoma is considered a different clinical entity.

Surgery is the preferred treatment option for microcystic lesions. Goals of surgical excision in the most aggressive cases are to grossly excise the lesion without sacrificing any critical neurovascular structures. Closure may be a challenge; often there is extra skin (with the lesion acting almost as a "tissue expander"), and the lesion may come up to and sometimes even involve the skin—microcystic malformations in particular have a propensity to "weep" through the skin. Often a large space is left behind and a drain should be considered.

Sclerotherapy should be the first line of treatment for macrocystic lesions, using bleomycin or doxycycline-based solution [91] (Fig. 20.27). OK-432, which is the preferred sclerosant, is rarely available in LMICs. Even if the lesion does not completely disappear, the residual mass would be smaller and easier to remove at surgery.

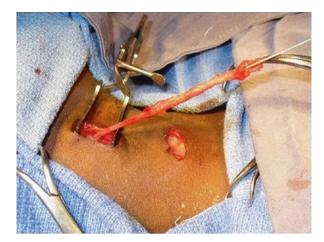


Fig. 20.28 Branchial cleft sinus excision

For malformations that present with infection, incision and drainage may be necessary in addition to antibiotics. Definitive excision of the lesion should not be undertaken in the setting of active infection due to surrounding inflammation.

Cystic masses in the neck may also include branchial cleft remnants. Their presentation and the spectrum of disease also vary greatly, from a large cyst to a small punctum draining saliva. It would be unusual to have to intervene for remnants in the neonatal period as they are generally asymptomatic. The most common type is the type II remnant, reliably located in the midsternomastoid. For a cystic mass, ultrasound may be helpful in identifying cystic or solid nature but likely not diagnostic. Remnants may be composed of true cystic masses but also may be long sinus tracts with origin in the hypopharynx. If excision is undertaken, a probe in the sinus tract may be helpful to guide the dissection. The tract may course between the carotid/internal jugular vein and also near other critical structures in the neck. In some circumstances they may also course through the thyroid gland. If the punctum is low in the neck with a long sinus tract, a "stepladder"-type incision above the original incision may be necessary for adequate exposure (Fig. 20.28). The most proximal aspect of the sinus tract should be ligated; nonetheless recurrence is possible. As with lymphatic malformations, definitive excision should not be undertaken in the context of active infection. Indications for excision are similar to lymphatic malformations. These cysts if left untreated are not thought to be malignant precursor lesions, but in the setting of chronic infection for years, carcinoma has been reported. Recurrence rates are much higher if the lesion is excised after a history of prior infection, and families should be counseled about this accordingly.

Thyroglossal duct remnants are also a common congenital neck mass (Fig. 20.29). These are midline lesions related to the development and descent of the thyroid from the base of the tongue. They can be found anywhere from the base of the tongue in the midline superiorly to the sternal notch inferiorly. They are generally cystic mobile masses; they may move with tongue protrusion. Incision and drainage



Fig. 20.29 Thyroglossal duct cyst

may be needed if there's an abscess, but definitive excision is recommended subsequently. The most critical aspect of definitive excision is not only of the cyst but the tract through the hyoid bone; the central part of the bone should be excised with the cyst (Sistrunk procedure) and a tract above the bone followed up to the base of the tongue. This tract above the bone is generally a thick band of tissue that one may ligate en masse. Often the proximal tract has multiple branches, and a wide excision will help to avoid inadequate incision. Again, in cases that have been previously infected, risk of recurrence is significantly higher.

In high HIV incidence LMICs, some masses may more likely represent infection—such as tuberculosis and bacterial infection—and these should be considered in the differential.

Management of Common Subspecialty Conditions

Hydrocephalus and Spina Bifida

Hydrocephalus

Hydrocephalus, or the abnormal collection of cerebrospinal fluid (CSF) in the ventricles, is a relatively common pediatric neurosurgical condition. In Western countries it is most likely due either to congenital occlusion of the aqueduct of Sylvius, acquired blockage by clot in premature babies, or secondary to brain tumors and in babies with spina bifida. In low-resource settings, half the cases appear to be infectious in etiology—due to untreated or poorly treated neonatal encephalitis or meningococcal meningitis in the older child. Congenital, neoplastic, and neural tube-related cases are also encountered.

The clinical presentation of hydrocephalus reflects the increase in intracerebral pressure, causing bulging and failure of closure of the fontanels, prominent scalp



Fig. 20.30 Neonatal hydrocephalus

veins, and sunset eyes (Fig. 20.30). Obstruction of CSF flow can also result in CSF infection, and the common denominator is progressive brain damage.

Treatment, aimed at reducing the intracranial pressure, is therefore urgently needed. In extremis a simple ventricular tap (with a 20 gauge needle) can be used to temporarily reduce the pressure and also examine the CSF for bacteria and white cells. The definitive treatment remains internal bypass of the obstruction, either using a shunt catheter or endoscopic ventriculostomy. The commonest shunt type is a ventriculoperitoneal shunt (VPS), with other drainage sites (pleural cavity, pericardium, gallbladder) reserved for unique situations. An endoscopic third ventriculostomy (ETV) is a fenestration performed in the floor of the third ventricle, thus allowing CSF drainage into the basal cisterns.

Both procedures are very meticulous and require special expertise and are therefore to be avoided by any surgeon without the needed expertise. Complications, both infectious and mechanical, eventually occur in at least half the shunt placements, requiring frequent shunt exteriorizations and/or revisions. Shunts are well known to block, to migrate, to cause bowel obstruction, and to erode through the bowel, vagina, or skin. Each complication typically worsens the neurological status of the child, often irreversibly.

Spina Bifida

There are several neural tube defects, including spina bifida, myelomeningocele, myeloschisis, and tethered cord. Spina bifida (Fig. 20.31) is mostly of the aperta (open) type, clearly visible at birth along the spinal canal. It is a major neurological



Fig. 20.31 Spina bifida. Myelomeningocele; lipomeningocele

defect associated with multiple other significant abnormalities. Affected children often develop hydrocephalus, may have severe kyphosis and uncal herniation (as in the Chiari II malformation), may be paraplegic, and often have bowel and urine incontinence, club feet, and other rarer manifestations (like lumbar hernias and uter-ine prolapse).

Investigations are seldom needed beyond the ultrasound for ventriculomegaly (encountered in two-thirds of patients) and hydronephrosis. Treatment includes early closure of the spinal defect to prevent its infection, followed by ongoing treatment of all the other manifestations as they appear: shunting or ETV for hydrocephalus, casting for the club feet, bowel and urine continence measures, and eventually surgery for kyphosis and other skeletal deformities. The commonest cause of death in these children is renal failure, and daily clean intermittent catheterization (CIC) is the treatment of choice to prevent urinary infections and vesicoure-teric reflex (VUR). In low-resource settings, most children are not offered multiple continence procedures such as Mitrofanoff conduits and antegrade colonic enemas (ACE). Instead, daily digital disimpaction and CIC with the possible addition of oxybutynin appear to be quite efficient in providing social continence and renal preservation.

As in other specialty areas, the visiting non-neurosurgeon is discouraged from performing the often challenging spinal defect closure and to work in close collaboration with existing urologists and orthopedic surgeons in managing the other longterm complications of this condition.

Cleft Lip and Palate

These are discussed in Chap. 21 Plastic Surgery for the Non-Plastic Surgeon in the Low Resource Setting.

Club Foot

This and other pediatric orthopedic conditions are discussed in Chap. 16 Essential Orthopedics for Global Surgery.

Hypospadias

Hypospadias, or the abnormal opening of the urethra proximal to its normal glandular location, is a relatively common condition. It is typically divided into distal types (glandular, coronal, distal shaft) and proximal (proximal shaft, penoscrotal, and perineal). The proximal forms are typically associated with chordee or curvature of the shaft and may merge into ambiguous genitalia (disorders of sexual differentiation, DSD).

Hypospadias is readily identifiable at birth but remains largely asymptomatic. It is typically isolated but may coexist in syndromes. The presence of palpable gonads typically differentiates it from DSD. No extra investigations are required, except for an abdominal ultrasound.

Repair of hypospadias is typically done before 1 year of age, though patients in resource-poor settings often come much later, even in their teens. The repair is the same regardless of age and is dictated primarily by the location of the abnormal meatus. Distal defects are repaired by primary urethroplasty, most commonly through a tubularized incised plate (TIP) repair as described by Warren Snodgrass [92, 93]. Glandular forms require even simpler localized repairs, and in fact the need for repairing them is questionable.

Proximal forms typically need both a urethroplasty and an orthoplasty (for chordee or curvature). Mild forms of curvature often correct once the penis is degloved but, if not, can be corrected by posterior plication. Severe forms require division of the shorter urethral plate. This can be done as one step with the urethroplasty but often requires staging (chordee release and creation of new urethral plate, followed by urethroplasty). Most repairs require a dripping stent for infants and special indwelling catheter for older children, for 5–10 days postoperatively.

There is a multitude of procedures for hypospadias, testimony to the fact that none are perfect. Complications occur in up to 30% of hypospadias repairs, especially in the proximal types. They include meatal stenosis, fistula, and repair breakdown. Iatrogenic urethrocutaneous fistulas require fistula repair 6 months later. Repeated failures, not uncommon in low-resource settings, result in "hypospadias cripples" whose penis has lost most skin coverage. These require complex repairs with replacement of the mucosa with the buccal mucosa or dermis.

As with other elective procedures outside the Western pediatric surgical spectrum, repair of hypospadias is best left for urologists or plastic surgeons with appropriate expertise.

Group of	5	
drugs	Drug names	Dosages
Analgesics	s and anesthetics	
	Morphine	Oral: 0.1–0.5 mg/kg/dose 4–6 hourly
		IV: 0.05–0.2 mg/kg/dose 6–8 hourly (do not exceed 0.1 mg in neonates)
	Paracetamol (acetaminophen)	IV: 10–15 mg/kg/dose 4–6 hourly
		oral: 15 mg/kg 4–6 hourly
		Rectal: 20 mg/kg 4–6 hourly (do not exceed 75 mg/kg/ day)
	Ketorolac	0.5 mg/kg/dose, 6 hourly
		Not to exceed 5 days
		Not advisable <2 months
	Ibuprofen	4–10 mg/kg/dose, 6 hourly
	Diclofenac	75–100 mg po q day in 3 divided doses if child <12
	Ketamine	Anesthesia induction: 4–5 mg/kg IM x 1 or IV 1–2 mg/ kg
		Sedation: 6–10 mg/kg po 30 mins before procedure; 5–20 mcg/kg/min
	Bupivacaine	>12 years old max dose 175 mg; 225 mg (with
		epinephrine); can repeat in 3 h
	Lignocaine (lidocaine)	Max 4.4 mg/kg
Antibiotics	5	
	Amoxicillin	50 mg/ kg po tid
	Ampicillin	50 mg/kg po q 6 h
	Ampiclox	250 mg po q 6 h age <2; 500 po q 6 h >2
	Amoxicillin +	30 mg/kg/day divided bid age < 3 months
	clavulanic acid	25 mg/kg/day divided bid age > 3 months
	Cloxcillin	50–100 mg/kg/day divided q6
	Chloramphenicol	25–50 mg/kg/day
	Ciprofloxacin	10–20 mg/kg q day po bid
	Ceftriaxone	50 mg/kg/day IV q day
	Cefuroxime	30 mg/kg/day po if age >3 months
	Gentamicin	7.5 mg/kg/day divided q3 h
		5 mg/kg/q day div. bid
	Metronidazole	10 mg/mg/dose IV tid

Appendix: Commonly Used Drugs in Children in LMICs

Group of drugs	Drug names	Dosages
Antiparasi		Dosuges
r nup ur us	Albendazole	>6 months: 400 mg single dose
		>6 months, <10 kg: 200 mg single dose
		Hydatid disease: 10–15 mg/kg/day in 2 divided doses. Give continuously without break
	Mebendazole	>6 months: 100 mg BD for 3 days
		>6 months, <10 kg: 50 mg BD for 3 days
		Hydatid disease: 40-50 mg/kg/day in 3 divided doses
	Metronidazole	For Amoebic infestation:
		35-50 mg/kg/day in 3 divided doses
		Give for 5 days for colitis and 10 days for liver abscess
	Tinidazole	For amoebic infestation:
		50 mg/kg QD for 3–5 days
Sclerother	apy of lymphangioma	
	Doxycycline	10-20 mg/ml concentration in normal saline
	Bleomycin	0.5–1 mg/ml aqueous solution. Maximum cumulative dose is 5 mg/kg
Seizure co	ntrol in head injury	
	Phenytoin	Loading: 20 mg/kg iv
		Maintenance: 5-10 mg/kg/day, 12 hourly IV or oral
	Phenobarbitone (phenobarbital)	Loading: 20 mg/kg IV
		Maintenance: 5–10 mg/kg/day, 12 hourly IV or oral (to start 12 h after loading dose)

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