CHAPTER 119
NEURODISABILITY

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Introduction

Neurodisability is probably the most significant contributor to chronic surgical disability in African children. It involves primarily hydrocephalus (HC) and spina bifida (SB), which can occur in isolation or together. Other causes of neurodisability include encephaloceles, which are part of the spectrum of neural tube defects (NTDs). True cerebral palsy is primarily nonsurgical, and therefore not included in this chapter.

Hydrocephalus is the excessive accumulation of cerebrospinal fluid (CSF) within the cranial vault. Its management in developing nations is hindered by significant economic constraints and delays in treatment—most patients, in fact, do not present for several months after the onset of clinical symptoms. The management of hydrocephalus and the complications associated with its treatment require considerable surgical judgment and a lifelong approach to patient follow-up.

Spina bifida is the term used for a spectrum of congenital NTDs. Other terms used for these anomalies are spinal dysraphisms or myelodysplasias. NTDs are complex medical problems that challenge surgeons and paediatricians alike, both in their initial management and in their lifelong complications. Even though SB may become a vanishing disease in developed countries, it remains a very significant cause of morbidity and disability in the developing world.

Demographics

The prevalence and incidence of HC in developed nations is estimated at 0.9–1.2 per 1,000 and 0.2–0.6 per 1,000, respectively.[1] No reliable estimate is available in the African literature, but its incidence in Africa is likely higher due to untreated or poorly treated neonatal meningitis and nutritional deficiencies.

The key epidemiologic features of SB are wide regional and ethnic differences in prevalence, a worldwide decline in prevalence over the past three decades, and female preponderance.[2] The reasons behind the decline are unclear and most likely multifactorial, although folic acid supplementation and fortification and selective termination of pregnancies are probably key factors.[3] The range in prevalence in Western nations is roughly 0.1–1 per 1,000 live births; a few non-Western studies quote higher rates, widely spread.[4–6]

Aetiology/Pathophysiology

CSF is produced predominantly by the choroid plexus of the four cerebral ventricles, at a rate of 20 ml/hour. It flows via the foramina of Luschka and Magendie into the subarachnoid space, and it is absorbed by the arachnoid villi into the venous system via the superior saggital sinus.

HC has been categorised as communicating or noncommunicating. The former is due to the failure of CSF absorption by the arachnoid villi, whereas the latter involves obstruction of CSF flow into the subarachnoid space. A small minority of cases exhibit excessive production of CSF—most commonly secondary to a choroid plexus papilloma.

In developed nations, HC has historically been most commonly due to myelomeningocele, with the posthaemorrhagic hydrocephalus of prematurity becoming at least as common in recent years.[2] Some reports have suggested that the most common causes of hydrocephalus in central Africa are NTDs and congenital aqueductal stenosis. Similarly, in Zambia, the ratio of congenital to "postmeningitic" HC has been reported to be 2:1.[7] In contrast, well-documented prospective series in East Africa have shown the aetiology of HC to be 57% postinfectious, 29% non-postinfectious, and 13% myelomeningocele.[8] Thus, neonatal meningitis or ventriculitis is likely the most common cause of hydrocephalus in East Africa.[8]

SB may result either from failure of closure of the neural tube or from secondary reopening of a closed tube, although most of the evidence favours the former theory.[9] The aetiology of SB is multifactorial.[2] A genetic component is evidenced by the familial risk, which appears to be 20–50 per 1,000 if one child is affected, 100 per 1,000 if two children are affected, and 30 per 1,000 if the mother is older than 35 years of age.[3] Environmental factors include low socioeconomic factors, maternal hyperthermia, and medications–primarily carbamazepine, valproic acid, and folate. Mothers taking carbamazepine and valproic acid have a 1% risk of having infants with SB. Folic acid, conversely, has been conclusively shown to both prevent the first occurrence of SB defects in pregnant women and to cause a 70% reduction in recurrent SB in mothers who already had pregnancies with NTDs.[10]

Clinical Presentation

The clinical presentation of HC is characterised by signs and symptoms of increased intracranial pressure (ICP). Symptoms may include headache, vomiting, failing vision, drowsiness, fatigue, deteriorating mental function, and enlarged head circumference. Signs include wide tense fontanelle, papilloedema, reduced visual acuity, failure of upward gaze (the sunsetting sign), general clumsiness, dyspraxic gait, and increasing head circumference (Figure 119.1). Older children will not present with increased head circumference; they often complain of the classic triad: headache, vomiting, and lethargy.

The obvious (apparent) spinal defects include myelomeningoceles, meningoceles (together referred to as *spina bifida cystica*), and

Figure 119.1: Severe congenital hydrocephalus with sunset eyes.

lipomeningoceles. Occult lesions include diastematomyelia (split cord), tight filum terminale, dorsal dermal sinus, and spinal lipoma. The term spina bifida occulta should be reserved for spinal bone fusion defects only.

The appearance of the spinal defect reveals its identity. Myelomeningoceles usually have a central "open" defect without normal skin, often with a visible placode (the open spinal cord). They may appear flat at birth, then often fill up with CSF. Older unoperated children will often have significant scarring, and the skin may indeed completely close the defect (Figure 119.2). Meningoceles and lipomeningoceles are fully skin-covered from birth, with the former typically cystic (Figures 119.3 and 119.4) and the latter fatty in consistency (Figures 119.5 and 119.6).

The distribution of the levels of SB depends on referral patterns and access to care, but usually about 40% are lumbosacral, 30% lumbar, and 30% thoracic or thoracolumbar.[11] The accurate assessment of the spinal cord function is critical. It must be kept in mind that the skin level of the defect may not accurately reflect the spinal level, that children may exhibit both upper and lower motor neuron lesions, and that the level may be asymmetrical. Areas of changed pigmentation, hairy patches, haemangiomas, lipomas, and deep skin pits in the thoracolumbar and sacral midline may belie an underlying NTD.

HC occurs in 80–90% of infants with spina bifida[1] (Figure 119.7), but may not be apparent until the spinal defect is closed. HC is less frequently seen in children with sacral defects. The authors' experience, as well as that reported elsewhere, points to a possible lower incidence of HC in developing countries.[11]

The associated Chiari II malformation may also cause specific hindbrain herniation symptoms in about 20% of children with SB. These symptoms include apnoea, a high-pitch cry, and swallowing difficulties.[3,9]

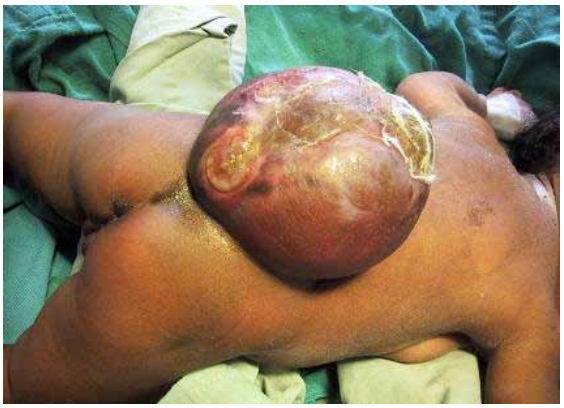

Figure 119.2: Late presentation of large myelomeningoceles with superinfection and partial scarring.

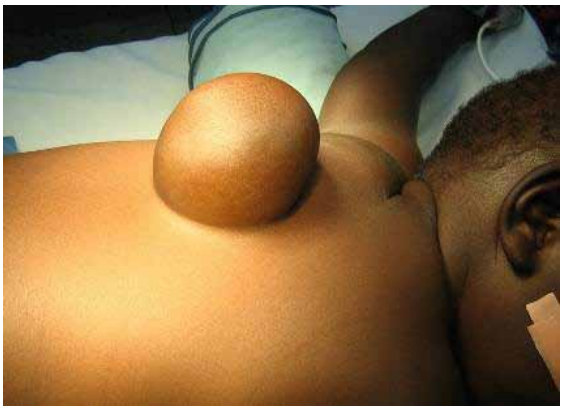

Figure 119.3: Thoracic meningocele with no neurological deficit.

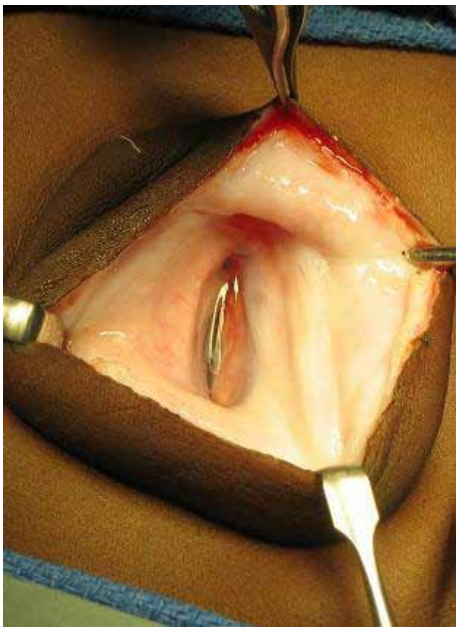

Figure 119.4: Intraoperative appearance of meningocele.

Figure 119.6: Intraoperative appearance of lipomeningocele, with adipose tissue invading dura and surrounding nerve fibres.

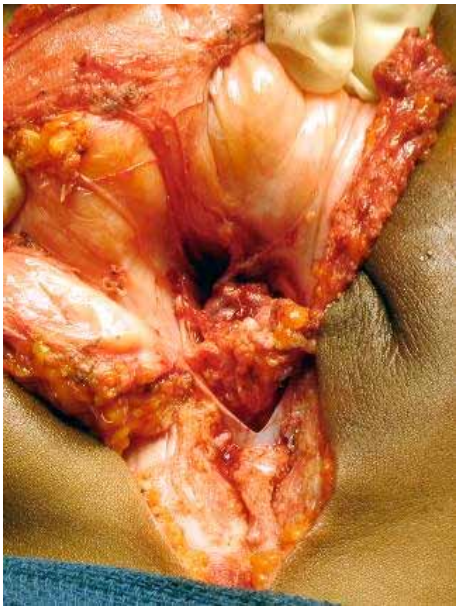

Figure 119.5: Large lipomeningocele.

Figure 119.7: Severely malnourished child with spina bifida and hydrocephalus.

Investigations

The clinical exam is the most readily available investigation for the diagnosis of increased ICP. Cranial ultrasonography (US) is an essential diagnostic tool in developing countries; it can readily assess ventricular size with minimal training, and it is relatively inexpensive. Depending on operator skill, the size of the fourth ventricle can be assessed on US as a proxy indication of the patency of the aqueduct. This may be particularly relevant in stratifying patients for treatment with prosthetic shunts versus endoscopic third ventriculostomy (ETV).[8] Serial US imaging may be appropriate in patients with an equivocal presentation of ICP prior to subjecting them to shunt revision. All children with shunts should be followed up regularly, including a baseline US within 3 months of surgery. Although acute changes from baseline may help in the subsequent diagnosis of shunt failure, up to a third of patients will not exhibit any evidence of ventriculomegaly.[12]

Both computed tomography (CT) and magnetic resonance imaging (MRI) are excellent modalities, but their routine use is prohibitively costly in developing nations. Nevertheless, CT may be necessary in assessing the ventricular size in older children with closed fontanelles. Evidence of increased ICP in children with closed fontanelles can also be obtained through direct measurement of CSF pressure by lumbar puncture: the CSF column height is measured in a piece of IV tubing connected to the spinal needle via a three-way stopcock.

No immediate investigations are required in a regular case of SB. Spinal x-rays may reveal other occult dysraphisms in 10% of patients,[6] although this will likely not affect the management. High-resolution US of the spinal cord is as effective as MRI up to the age of 6 weeks, identifying diastematomyelia and tethered cord, and screening for dilatation of the urinary tract. MRI of the spine is frequently performed in developed nations, although rarely necessary routinely. US of the head for HC is useful, although the ventriculomegaly may not be evident until the CSF leak through the spinal defect is closed. Several investigations for the genitourinary system are discussed under the subheading "Urological Problems" later in this chapter.

Management

The definitive management of HC at present remains surgical. The diuretic acetazolamide has been shown to decrease CSF production in animal and human studies,[14] but it is of temporary benefit and should be used only in the palliative setting or in equivocal cases until a definitive diagnosis can be made. It has also been used in posthaemorrhagic HC of the newborn as a temporising measure to avoid shunting.[15]

The most common surgical intervention to treat HC is the insertion of a shunt through the skull and cortical mantle into the ventricle, with the distal catheter placed into a physiologic drainage basin, typically the peritoneal space (ventriculoperitoneal (VP) shunt). Other sites for CSF diversion include the right atrium and the pleural space. The advantage of a CSF shunt is that it is beneficial in nearly all types of HC, regardless of aetiology.

CSF Shunts

CSF shunts usually contain three parts: a ventricular catheter, a valve, and a distal catheter. Most valves are designed to allow for sampling via needle puncture. The so-called "differential pressure valves" use the gradient between the ventricle and the tip of the distal catheter to effect flow. Medium pressure valves are those that drain CSF if the pressure gradient is >10 mm Hg, and are used most commonly. Although many different valve designs exist, including siphon-limiting, flow-limiting, and programmable valves (whose settings can be changed by using an external magnet), there is limited evidence for their benefit. A large randomised trial has demonstrated no difference in time to first shunt failure with a standard differential pressure valve compared to two other higher-generation valves in the treatment of children with newly diagnosed HC.[15] Similarly, the use of an adjustable shunt was not shown to be of any benefit in terms of overall survival.[16] Finally, and of most relevance to the developing world, there is good evidence from a prospective randomised controlled trial demonstrating that the Chhabra® shunt (made by Surgiwear in India) is equivalent to its common Western counterpart in incidence of shunt complications, but sells for only about one-twentieth the cost.[17] The Chhabra shunts are available free to qualifying centres through the International Federation of Spina Bifida and Hydrocephalus (www.ifglobal.org) In extreme situations, a piece of IV tubing or a sterile Silastic® feeding catheter can be used as a VP shunt, but these nonvalved alternatives are associated with frequent complications and are not recommended.

Pleural Shunts

Pleural shunts are rarely required, but can be placed via the 4th to 6th intercostal space at the anterior axillary line into the pleural cavity, with care to avoid placement into lung parenchyma or the chest wall. The associated CSF effusion and iatrogenic pneumothorax resolve conservatively in most patients.

Ventriculoatrial Shunts

Ventriculoatrial shunts are rarely performed because of complications of cor pulmonale, shunt nephritis, and catheter embolisation. They require intraoperative US or fluoroscopy to document catheter placement into the atrium via the internal jugular vein. Their use is not recommended.

Endoscopic Third Ventriculostomy

The morbidity of the life-long shunt has created interest in the use of endoscopic third ventriculostomy (ETV), a procedure that can effectively treat HC without insertion of any foreign body. The principles of placement of an ETV include frontal access, ventricular cannulation, and insertion of a rigid or flexible neuroendoscope into the 3rd ventricle via the lateral ventricle and the foramen of Monroe. A fenestration is made in the base of the third ventricle between the infundibular recess and the mammillary bodies. This is commonly performed by using a combination of electrocautery and a balloon dilator, thus creating a cerebrospinal fluid fistula between the subarachnoid space and the 3rd ventricle.[18] More recently, the concurrent performance of cauterisation of the choroid plexus (CPC) has been added as a means of increasing shunt avoidance.

Classically, ETV was used for older children or adults with congenital aqueductal stenosis. Working in Uganda, Dr. Benjamin Warf has convincingly demonstrated, however, that ETV is a reasonable option for all children >1 year of age (irrespective of HC aetiology), with a shunt avoidance rate of 80%.[17] In addition, with the use of cranial US and direct endoscopic visualisation of the aqueduct, he stratified younger children based on aqueductal patency, and has demonstrated 70% success

in those <1 year of age who have a post-infectious obstruction of the aqueduct. In addition, the use of ETV in combination with choroid plexus cauterisation has increased shunt avoidance from 35% to 76% in children with myelomeningocele, and from 20% to 71% in children <1 year of age with a non-postinfectious HC and an open aqueduct.

Performance of ETV is beyond the scope of this review, and requires adequate mentoring by an experienced surgical team and significant technical support for maintaining the endoscopic system. Although the avoidance of a shunt (and the accompanying morbidity and mortality of shunt failure) in the developing world is a reasonable goal, the additional safety of ETV has not yet been proven in the long term, and most authors counsel that the same life-long follow-up is required in either case.[1]

Spina Bifida Surgery

The management of a child with SB is lengthy and complex.[3,19] The closure of the spinal defect is the most obvious step, although it is by far not the most challenging one. In contrast, some older asymptomatic children presenting with relatively small defects that are fully skin-covered and mostly scarred may not need to have their defect "closed", especially as the surgery in those instances can be very difficult and dangerous. Such children will, however, need to be carefully followed up for the appearance of tethered cord symptoms and signs.

In the newborn with SB, the spinal defect should be closed ideally within the first 2 days, although delays within the first week of life while the child is on antibiotics do not seem to adversely affect the outcome.[2] In developing countries, children typically present after the first week of life,[11,20] and the defect is often grossly infected (see Figure 119.2). While preoperative intravenous antibiotics are the rule in all settings, there is little advantage to lengthy preoperative courses of antibiotics and dressings.

The standard repair of open SB includes the following steps:[3,9]

Step 1	The sac or the skin surrounding the placode is incised; the placode is isolated circumferentially with removal of all keratinized areas as well as the superficial granulation tissue.
Step 2	The placode is tubularised with a running monofilament fine suture to reduce the raw surface and re-create a tubular cord.
Step 3	All adhesions to the cord are lysed both proximally and distally.
Step 4	The dura is dissected circumferentially with overlying fatty tissue, down to the central region of the defect.
Step 5	A watertight dural closure is achieved with one layer of running fine monofilament suture.
Step 6	The skin and subcutaneous tissues is undermined widely laterally.
Step 7	The fascia is closed with interrupted absorbable sutures at the level of the original junction of the skin, dura, and defect covering. The skin is closed with no tension.

Variations to the above steps may include:

- no tubularization of the placode if it is technically difficult;
- division of a thickened filum terminale and correction of diastematomyelia, if found;
- resection of placode and transection of the cord in cases with full paraplegia, especially if infection is present and the cord is severely atrophic/dysplastic (Figure 119.8);
- irrigation and intrathecal injection of antibiotic (usually gentamycin); and
- transverse dural closure proximally only if the cord has been transacted.

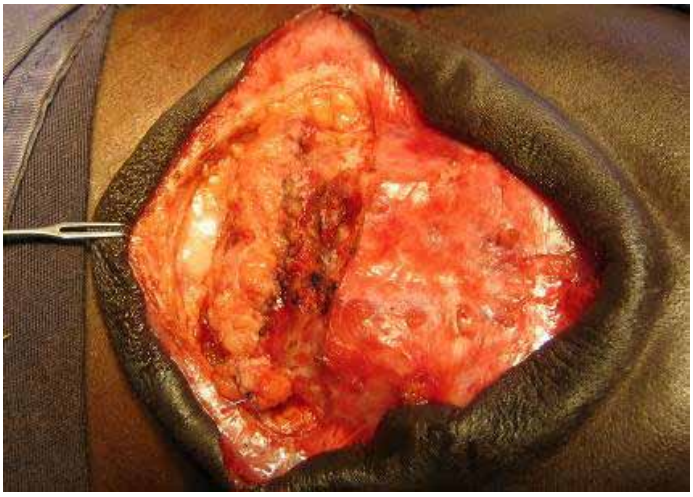

Figure 119.8: Proximal dural closure following cordotomy in thoracic-level myelomeningocele with paralytic lower extremities.

Postoperative Complications

Postoperative complications of shunt insertion can generally be classified as mechanical or infectious.

Mechanical Shunt Failure

Mechanical shunt failure can occur through proximal obstruction; distal obstruction; component separation, fracture, or migration; or excessive CSF drainage.

Proximal or distal obstruction generally presents with signs and symptoms of increased ICP, whereas infectious failure presents with fever, redness, or swelling at the surgical site, drainage of pus or CSF from the wound; nuchal rigidity; abdominal pain; or peritonitis. Shunt obstruction in older patients often presents with headache and is not associated with pain along the shunt tract. Although the signs and symptoms of shunt failure have been examined empirically, there is no ideal diagnostic test.

Revision of a noninfected but obstructed shunt should generally be approached from the cranial incision because obstruction is more common at the ventricular limb. Both ventricular and abdominal limbs can then be tested in sequence from this location after they have been disconnected from the valve. Nonfunctioning ventricular catheters that are adherent can be removed by gentle traction under most circumstances, but we do not advocate more aggressive measures; rather, a new shunt should be placed at an alternative site. Passage of a new peritoneal shunt down an established fibrous tract is occasionally possible as long as shunt infection has been ruled out preoperatively. Routine CSF culture in the absence of infectious signs or symptoms is not recommended because positive cultures in this setting are not predictive of subsequent infection.[21]

Shunt Infection

Shunt infection is widely believed to be caused by contamination at the time of surgery, with occasional infections caused by later wound breakdown either due to CSF fistulisation or skin breakdown over hardware (Figure 119.9). Most shunt infections usually present within 3 months of shunt insertion, and almost all within 6 months. The rate of shunt infection in North America is approximately 8–10%,[22] with some series reporting less than 1%,[23] but it is likely higher in Africa.[24] The most common organisms grown are consistently *Staphylococcus epidermidis* (~40%), followed by *S. aureus* (~20%).[25]

Risk factors for shunt infection include prematurity,[26,27] elaborate shunt configurations, multiple separate shunts, previous shunt infection, surgical inexperience, myelomeningocele, postoperative CSF leak, and longer duration of surgery.[28]

Common techniques to avoid shunt infection include the use of generous skin preparation, meticulous and consistent surgical

technique, and preoperative prophylactic antibiotics.[29] Although there is significant worldwide heterogeneity with regard to choice of antibiotics,[30] we recommend ceftriaxone or cefazolin upon induction of anaesthesia. A Cochrane review in 2006 has supported the use of antibiotic prophylaxis in reducing shunt infection, with no evidence of benefit beyond 24 hours.[31]

As advocated by Faillace and colleagues, the avoidance of shunt-to-skin contact resulted in a three-fold decrease in shunt infection rates, from 9.1% to 2.9%.[32] As mentioned earlier, valve design does not appear to have any effect on shunt infection. The surgeon should develop a consistent routine by using the same equipment, and a meticulous approach; some authors have reported very low infection rates using similar techniques.[33] Finally, shunts should not be relegated to the most inexperienced member of the team. We agree with the author who suggested: "Although less glamorous than other neurosurgical cases, shunting procedures deserve no less attention to detail."[34]

Because conventional antimicrobial techniques are not effective in treating the bacterial biofilms that affect most neurosurgical-related device infections, all infected shunts should be removed.[35] This is to be combined with the insertion of an external ventricular drain with ongoing systemic and optional intrathecal antibiotics, with replacement of the shunt 10–14 days later with preoperative confirmation of a sterile CSF. Treatment with antibiotics alone has a high failure rate and probably is relevant only in the high-risk surgical patient. Vancomycin has poor CSF penetration by IV route, and thus preservative-free ventricular vancomycin has been commonly used to treat shunt infections, despite the incomplete understanding of its side effects and toxicity.[36]

Shuntalgia Syndrome

Shuntalgia syndrome is an unusual shunt complication that presents with focal discomfort around the shunt site *without* swelling, fluctuance or redness. There may be tenderness along the shunt itself, and there is usually a hard fibrotic sheath of scar tissue around the shunt. Shuntalgia syndrome is common in adolescents and should generally be treated with conservative measures, although narcotic analgesics have been required in some cases.

Shunt Separation, Fracture, or Migration

Shunts rarely may separate, fracture, and/or migrate within the first few months of surgery. Clinical examination and, if necessary, a shunt series of plain radiographs along the entire shunt tract are sufficient to confirm the diagnosis (Figure 119.10). The phenomenon of arrested HC (whereby a child develops true shunt independence) is infrequent: 80% of children who have stable ventricles despite a disconnected shunt have a raised ICP.[37] Therefore, most nonfunctioning shunts should be revised.

Hollow Viscus Perforation

Perforation of virtually every hollow viscus by the peritoneal catheter has been described, but is usually diagnosed by observation of the catheter protruding from the anus (Figure 119.11). The risk of hollow viscus perforation has been estimated at 1 per 1,000 shunt years.[38] Remarkably, peritonitis is *rarely* a presenting feature, likely due to the gradual erosion of the shunt through bowel. Treatment is similar to other cases of shunt infection and involves removal of the shunt in its entirety via a single valve-site incision in the cranium, external drainage for 10–14 days with intrathecal or intravenous (IV) antibiotics, followed by shunt replacement. Laparotomy is reserved for the rare case of peritonitis, but is not routinely required to remove the shunt.

Spina Bifida Problems

In spina bifida, wound problems are frequent, including infection, dehiscence, and necrosis.[11,39,40] They almost always can be managed conservatively with dressings, debridement, and sitz baths. CSF leaks may occur,[11] and although most will resolve with time and control of the CSF pressure (with shunting or acetazolamide), persistent leaks may require re-exploration.

Figure 119.9: Ventriculoperitoneal shunt erosion.

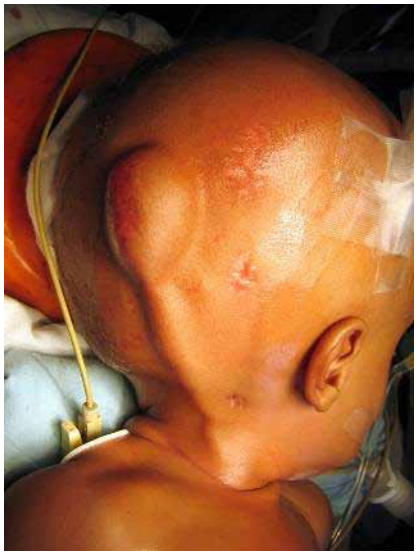

Figure 119.10: Perishunt CSF collection.

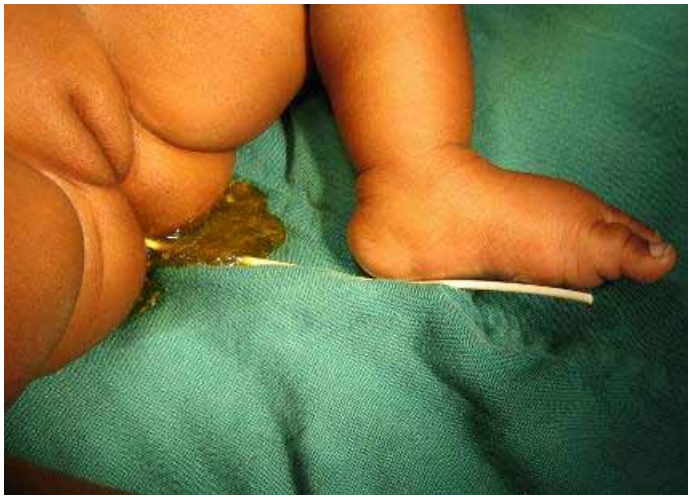

Figure 119.11: Shunt extrusion per rectum following chronic intestinal erosion.

Retethering of the cord at the repair site occurs in 15–20% of long-term cases[9] and requires prompt surgical untethering where there are progressive symptoms.

Chiari II Problems

Concomitant shunting for HC at the time of the neonatal spinal defect closure is standard in developed nations, but the approach must differ in the developing world. In the African experience, early shunting leads to frequent shunt infection and ventriculitis.[41] The authors' evidence-based practice is therefore to wait 5 days after the spinal closure for shunting, or longer if there is any evidence of wound infection. Mild, stable, HC with a cortical mantle of at least 3.5 cm can be observed safely for several months without deleterious effects.[3]

Other Chiari II symptoms and signs (e.g., apnoea, stridor, poor swallowing) are initially managed by decreasing the ICP through shunting.[9] Persistent symptoms may require a posterior fossa decompression,[3,9] although this procedure is challenging and should be referred to specialised centres.

Musculoskeletal Problems

Scoliosis and/or kyphosis are the most common orthopaedic associations of SB. They develop in up to 60% of children with SB,[11] especially in children with thoracic defects.[3] Seating appliances can help, but braces are of questionable value, and surgical management (spinal fusion) is very challenging.

Talipes equinovarus (TEV, or clubfoot) is the next most common orthopaedic problem.[11] Ideally, it is treated conservatively through casting in the neonatal period; later on, a posteromedial release may be required.[42] Other lower-extremity problems include high-arch foot deformity, leg-length discrepancy, flat foot, foot valgus, and congenital dislocation of the hip.[11]

Depending on the motor level, patients with SB may require a variety of orthotic devices to allow partial or full ambulation. These include above- and below-knee braces, crutches, walkers, and wheelchairs.[3] Traditional teaching states that independent ambulation is possible if the quadriceps are strong (L3–4), but long-term studies have shown that the mobility of children with SB decreases with age despite stable neurological status.[3,9]

Gastrointestinal Problems

Nutritional problems are frequent in the SB population. Whereas obesity from limited activity is common in developed nations, many children with SB in the developing world suffer from nutritional deficiencies.

Defecation problems are the main challenge in this population.[43,44] Constipation occurs predominantly in children with high lesions due to slow colonic transit, and in children with sacral lesions due to deficient rectal sensation.[45] Constipation may be managed through dietary manipulation combined with regular finger stimulation or manual evacuation.

Faecal incontinence is much more of a challenge, with at least half of children with SB being affected.[11,45] Children with lumbosacral lesions often have pellet-like stools from slow left colonic transit, evacuated without voluntary control despite fair sphincter function. They are best managed with intentional constipating foods and daily manual evacuation. School-age children who fail this regimen should first be tried on retrograde washouts every 1 to 3 days.[45,46] An enema device including a plastic "cone" used with an enema tube is an effective, easily produced, daily wash-out tool. The next step is the Malone antegrade continence enema (MACE) procedure.[46,47] In this procedure, the child's colon is cleaned daily with up to 500 ml water or saline administered through a cutaneous continent appendicostomy. In developed countries, this procedure has been modified to use a small cecostomy "button" device inserted under radiographic guidance.[48] The standard cutaneous appendicostomy, however, is quite effective and well-suited for the developing world.

Integumentary Problems

Decubitus ulcers occur frequently in patients with SB, especially those beyond the age of 5 years.[11] These ulcers represent therapeutic challenges. Similar to management of ulcers in other patients with neurological deficits, a conservative approach with saline dressings and avoidance of pressure areas is always warranted. Refractory ulcers can benefit from plastic surgical procedures, although recurrences are frequent.

Other ulcers in these patients are found in the perineal area and are caused by urinary and/or stool incontinence. These ulcers must be managed by attempting to address the underlying incontinence problems.

Latex Allergy

Latex allergy is an immunoglobulin E (IgE)-mediated problem leading to the spectrum of urticaria, bronchospasm, and anaphylaxis. Although latex allergy is a frequent (20-30%) complication in children with SB in developed countries,[49,50] it is rarely reported in the developing world. Some evidence from South Africa suggests a lower overall incidence compared to Western nations.[51]

Urological Problems

Spina bifida is the most common cause of neurogenic bladder dysfunction in Africa, although sacral agenesis, anorectal malformations, and sacrococcygeal teratoma also contribute. Neuropathic bladder leads to renal failure[52,53] in 50% of untreated patients by the age of 5 years, and this constitutes the main cause of mortality in this population.[54] Children with SB should be monitored from birth whenever possible—otherwise, late presentations may include palpable bladder, chronic renal failure, urinary tract infection (UTI), or urinary incontinence. Micturating cystourography (MCUG) and US may reveal a thick-walled bladder with hydronephrosis and vesicoureteric reflux (VUR).

The normal function of the bladder is to store urine at a safe pressure by coordinated relaxation of the detrusor muscle with tonic sphincter contraction, and to empty completely at low pressure on command by detrusor contraction with reflex sphincter relaxation. Failure of the sphincter tone leads to a low leak point pressure (LPP) and a wet child, but essentially a "safe" bladder. This may be assessed by examination of the anocutaneous reflex. If the anus is closed and contracts (winks) when the adjacent skin is scratched, this suggests an intact reflex arc and is usually matched by similar urinary sphincter competence. In contrast, the flaccid anus with no wink is usually associated with a low bladder outlet resistance.

An intact anocutaneous reflex, however, does not ensure coordinated detrusor sphincter activity, and lack of reflex sphincter relaxation during detrusor contraction (sphincter dyssynergia, or DSD) causes bladder outlet obstruction, often leading to detrusor hypertrophy and fibrosis. This, in turn, results in a poorly compliant bladder in which the pressure rises during filling until the leak pressure is overcome and the child is wet. If this pressure is high enough, reflux and renal damage ensue. Detrusor instability occurs as a result of primary nerve injury or secondary to bladder damage caused by the DSD. In some cases, however, the detrusor is of low tonicity, and the bladder will be large and low-pressure even with a low fixed-resistance sphincter. Whenever there is poor detrusor tone or DSD, voiding will not be complete and there will be a significant post-void residual (PVR); this reduces the functional capacity of the bladder and increases the infection risk.

In summary, there is an interplay between the bladder and sphincter muscles, which may be considered in three groups: synergic (both muscles acting in unison (19%); dyssynergic with or without detrusor hypertonicity (DSD) (45%), and denervated (36%). The basic urological work-up of patients with SB includes renal US with PVR measurement, serum creatinine, and volume urodynamics. This may be performed with disposable pressure sensors attached to a dedicated computer-based system, or, more appropriate to the African setting, using a simple burette and three-way stopcock apparatus.[55] Pressure measurement at 50% of estimated bladder capacity (8 ml/kg for infants

or 30 ml/year of age), PVR and LPP are the most important guides. More sophisticated electromyography (EMG) urodynamics is rarely contributory to outcome or management.

The mainstay in the treatment of the neurogenic bladder of children with SB is clean intermittent catheterisation (CIC). This procedure is simple to perform and to teach in all patient groups; in addition, it is inexpensive, safe,[56] and very efficient. CIC has revolutionised urological care in SB and is well suited for resource-poor settings. It is most efficacious when started in infancy, although it can be started at any point in time. It is performed by the main caregiver of the child until the age of 6–7 years, after which self-catheterisation can be taught.[56]

Most European centres perform urodynamics early and at intervals of up to 5 yearly depending on individual assessment and management needs. Alternatively, a nonselective approach can be used, as performed at BethanyKids, volume urodynamics are performed 3 days after the closure of the spinal defect, and caregivers are then taught to perform CIC if the volumetric criteria are met (LPP ≥ 30 cm or a PVR ≥ 10 cc).[55] Other criteria used include laboratory evidence of renal dysfunction (abnormal renal US, creatinine, or urinalysis), recurrent UTIs, and the need to promote social continence in older children. A pressure rise of greater than 20 cm H_2O at 50% estimated bladder capacity has been associated with a poor renal outcome, and in developed countries is a relative indication for a bladder augmentation or diversion.

Antimuscarinics such as oxybutinin (administered either orally or intravesically) complement CIC by reducing detrusor overactivity; combined with CIC, these have been shown to significantly reduce the development of renal damage. CIC with or without medications will prevent renal deterioration in 90% of children with SB, and achieve social continence in about 85%.[3] Only a minority of patients should require urological procedures such as bladder augmentation, bladder neck reconstruction, or urinary diversion.[53,57,58] Augmentation cystoplasty and bladder neck repair have high revision and complication rates and should be performed only where ongoing surveillance and care can be guaranteed.

Long-term renal follow-up is essential in all SB patients, however. The authors advise twice yearly renal US initially, with void pattern and urodynamics at intervals depending on the assessment of risk and evidence of changing urological status. In developed countries, renal transplantation is ultimately the treatment of choice for renal failure,[54] but this is rarely an option in developing countries, underlining the importance of prevention of renal damage by good, early, assessment and management.

Prognosis and Outcomes

The mortality associated with shunt placement is about 0.1%, but shunt failure is more lethal—1–4%—especially in the African setting with its frequent difficult and delayed access to health care. The operative risk of ETV is still being defined, but perioperative mortality may approach 1%.[1] Shunts fail due to mechanical or infectious causes at a rate of 30–40% within the first year after placement,[22] 15% within the second year, and 1–7% per year thereafter. Shunt infection is associated with reduced intelligence quotient (IQ) and poor school performance, as well as a higher risk of future shunt infection.[59] Although the risk of ventriculitis is reduced after ETV, it is not zero.

The mortality in patients with SB appears to be 25–50% into adulthood,[3,50] and naturally higher in developing countries. Renal failure as well as sepsis and shunt complications are common causes of mortality.[60] According to a long-term Western study, more than 85% of SB patients have VP shunts, and most have undergone at least one revision.[50] A third of the patients have undergone a tethered cord release, and half have scoliosis. The same long-term Western study showed, however, that 85% of patients are attending or have graduated from high school and/or college, and more than 80% have social bladder continence.[50] A British study found a 50% mortality after 30 years; among the survivors, 70% had an IQ of 80 or more, 37% lived independently in the community, 39% drove a car, 30% could walk more than 50 metres, and 26% were in formal employment.[61]

Results from developing countries are rather scanty. A Nigerian study found that 40% of children with myelomeningoceles were "functionally disabled" and could not be adequately rehabilitated because of limited resources.[39] The authors of that study therefore advocated selective management. A South African study revealed a 70% ambulation rate, 45% urinary continence, and a mean IQ of 80.[20] As expected, results were better in urban areas and in the higher socioeconomic groups. The authors' own experience in Kenya has shown that the quality of life of children with SB was quite acceptable, and was not related to the degree of the spinal defect.[62] Interestingly, quality of life in SB appears strongly influenced not only by neurological characteristics, but also by "soft" factors such as parental hope.[62]

Prevention

In East Africa, the single most common cause of hydrocephalus is infection of the CNS, usually via neonatal meningitis or ventriculitis. Neonatal sepsis is common and is exacerbated by the lack of skilled perinatal care for the majority of births in Africa.[8] Newborns presenting with febrile illness should ideally receive appropriate diagnostic tests and directed antibiotic therapy, and not just empiric therapy for presumed malaria. Efforts at improving perinatal care in developing countries will undoubtedly help to reduce the incidence of post-infectious HC.

Based on the overwhelming evidence for the importance of folic acid in preventing NTDs, folate supplementation of at least 400 μg daily has been uniformly recommended for all women of childbearing age. However, the difficulty in reaching this wide at-risk group makes food fortification, adopted in most developed countries, a much better method.[10] This policy, if implemented fully, is expected to result in a 50% reduction in NTDs. Unfortunately, only 10% of African countries have been able to implement this policy.[63]

Ethical Issues

There is a paucity of literature concerning the ethics of nontreatment of HC and the issues surrounding resource allocation in the treatment of this disease. Informed consent of the patient and family should address the need for lifelong ongoing surveillance of any child treated for HC, and the risks of shunt infection, shunt failure, and death. Families should be educated regarding signs of infection or shunt failure at the first admission.

Although we are not aware of any guidelines regarding assessment of medical futility in the setting of HC, we recommend extreme caution in shunting children with head circumferences >60 cm, or those who have nonhealing pressure sores on the skull. For many of these children, aggressive surgical intervention is futile, and family resources are challenged by lengthy or repeated admissions.

Similarly SB treatment is a long-term commitment, and the decision to operate must be carefully discussed with the family. Major associated congenital anomalies and prenatal large HC may lead to a palliative approach without surgery, but the level of the defect should not affect that decision. Although in the past some have advocated a selective approach to SB,[39,64] there is good evidence that a nonselective approach yields equally good results, while giving a chance for life to many more children.[3,62,65] In fact, the overall mortality and the IQs of the unselected groups compare favourably with those of the "best" infants from the selected group.[2] Looked at differently, 60% of the children from the selected group, who were allowed to die, would have been "competitive" if allowed to survive.[3]

Evidence-Based Research

Table 119.1 presents a prospective study of 195 Ugandan children that compares the Chhabra and Codman-Hakim Micro Precision shunt systems. Table 119.2 presents a trial study to evaluate the effect of folate supplementation periconceptionally in reducing foetal neural tube defects.

Table 119.1: Evidence-based research.

Title	Comparison of 1-year outcomes for the Chhabra and Codman-Hakim Micro Precision shunt systems in Uganda: a prospective study in 195 children
Authors	Warf BC
Institution	CURE Children's Hospital of Uganda, Mbale, Uganda
Reference	J Neurosurg (Pediatrics 4) 2005; 102:358–362
Problem	The high cost of commercial ventriculoperitoneal (VP) shunt systems is prohibitive in Africa.
Intervention	Insertion of VP shunts in hydrocephalic children.
Comparison/ control (quality of evidence)	Randomisation of 195 children between the Chhabra shunt system (cost: US$35) and Codman-Hakim Micro Precision Valve system (cost: US$650).
Outcome/ effect	No statistical difference was found in any shunt complication or overall outcome between the two shunt systems.
Historical significance/ comments	Classic, robust paper documenting the efficacy of appropriate simple technology for the African setting.

Table 119.2: Evidence-based research.

Title	Periconceptional supplementation with folate and/or multivitamins for preventing neural tube defects
Authors	Lumley J, Watson L, Watson M, Bower C
Institution	Cochrane Reviews
Reference	Cochrane Database Syst Rev 2001, Issue 3
Problem	The need to reduce the incidence of neural tube defects.
Intervention	Periconceptual folate supplementation.
Comparison/ control (quality of evidence)	Four trials including 6,425 women, all randomised between folate supplementation and control.
Outcome/ effect	Periconceptional folate supplementation reduced the incidence of neural tube defects (relative risk 0.28, 95% confidence interval 0.13–0.58). Folate supplementation did not significantly increase miscarriage, ectopic pregnancy, or stillbirth.
Historical significance/ comments	Critical Cochrane review documenting the clear positive effect of folate in reducing neural tube defects without any deleterious effects.

Key Summary Points

Hydrocephalus

1. Early treatment of HC with ventriculoperitoneal shunts remains the best method in Africa of preventing lifelong disability from increased intracranial pressure.
2. Clinical symptoms and signs and cranial ultrasound are sufficient for the diagnosis and management of children with HC.
3. Simple valved shunts, such as the Chhabra shunt, are as effective as more sophisticated devices and save significant resources.
4. Shunt placement must be a thoroughly sterile procedure performed by skilled, experienced surgeons.
5. Both mechanical and infectious shunt complications can be significantly reduced through meticulous technique and experience.
6. Children shunted for HC must be followed up for life and must have rapid access to health care facilities in case of complications.
7. Endoscopic third ventriculostomy shows significant promise for avoiding shunt morbidity in children with HC, but remains limited by technology and skill.
8. Besides early diagnosis, prevention of HC can be accomplished through efforts directed at the correct management of neonatal infections and folic acid supplementation for all women of child-bearing age.

Spina Bifida

1. Folic acid can prevent half of the cases of SB, and therefore efforts should be made to supplement folic acid in the diets of all women of child-bearing age. As this is very difficult in developing nations, policies for folic acid fortification of common foods should be actively pursued.
2. All children with SB should be thoroughly examined for associated conditions, including HC, musculoskeletal, genitourinary, gastrointestinal, and skin problems.
3. Initial investigations need include only head ultrasound and postoperative renal evaluation.
4. A nonselective approach to the treatment of SB leads to satisfactory results in developing nations and can be adopted if the resources are available.
5. Most SB defects in Africa present beyond 48 hours of life, and must therefore be considered infected.
6. Surgical repair may include transection of the cord, excision of the placode, and proximal dural closure in severe paraplegic cases.
7. Dural replacements and skin flaps must be avoided due to the risk of infection.
8. Shunting for HC, if needed, should be delayed by at least 5 days after the closure of the SB defect, due to the risk of infection.
9. The long-term management of children with SB is complex and requires multidisciplinary resources; therefore, these children should, as much as possible, be treated or referred to centres able to provide the necessary care.
10. The key impact on the long-term survival of children with SB is proper urological management, including urodynamic evaluation, clean intermittent catheterisation, and detrusor overactivity relaxants. Clean intermittent catheterisation is effective, inexpensive, and feasible in developing nations.

References

In addition to the following references, the reader is directed to the website for the International Federation for Spina Bifida and Hydrocephalus, www.ifglobal.org.

1. Garton H, Piatt J. Hydrocephalus. J Pediatr Clin NA 2004; 51:305–325.
2. Park TS. Myelomeningocele. In: Albright L, Pollack I, Adelson D, eds. Principles and Practice of Pediatric Neurosurgery. Thieme Medical Publishers, 1999, Pp 291–320.
3. Dias MS. Neurosurgical management of myelomeningocele (spina bifida). Pediatr Rev 2005; 26(2):50–60.
4. Airede KI. Neural tube defects in the middle belt of Nigeria. J Tropic Pediatr 1992; 38(1):27–30.
5. Kulkarni ML, Mathew MA, Reddy V. The range of neural tube defects in southern India. Arch Dis Child 1989; 64(2):201–204.
6. Msamati BC, Igbigbi PS, Chisi JE. The incidence of cleft lip, cleft palate, hydrocephalus and spina bifida at Queen Elizabeth Central Hospital, Blantyre, Malawi. Cent Afr J Med 2000; 46(11):292–296.
7. Adeloye A: Management of infantile hydrocephalus in Central Africa. Trop Doct 2001; 31:67–70.
8. Warf BC. Hydrocephalus in Uganda: the predominance of infectious origin and primary management with endoscopic third ventriculostomy. J Neurosurg (Pediatrics) 2005; 102:1–15.
9. Kaufman BA. Neural tube defects. Pediatr Clin N Amer 2004; 51(2):389–419.
10. Lumley J. Preconceptional supplementation with folate and/or multivitamins for preventing neural tube defects. Cochrane Database Syst Rev 2006;1.
11. Kumar R, Singh SN. Spinal dysraphism: trends in northern India. Pediatr Neurosurg 2003; 38(3):133–145.
12. Iantosca MR, Drake JM. Cerebrospinal fluid shunts. In: Albright AL, Pollack IF, Adelson PD, eds. Operative Techniques in Pediatric Neurosurgery. Thieme Medical Publishers, 2000, Pp 3–14.
13. Carrion E, Hertzog JH, Medlock MD, Hauser GJ, Dalton HJ. Use of acetazolamide to decrease cerebrospinal fluid production in chronically ventilated patients with ventriculopleural shunts. Arch Dis Child 2001; 84(1):68–71.
14. Libenson MH, Kaye EM, Rosman NP, Gilmore HE. Acetazolamide and furosemide for posthemorrhagic hydrocephalus of the newborn. Pediatr Neurol 1999; 20(3):185–191.
15. Kestle J, Drake J, Milner R, et al. Long-term follow-up data from the shunt design trial. Pediatr Neurosurg 2000; 33(5):230–236.
16. Pollack IF, Albright AL, Adelson PD. A randomized, controlled study of a programmable shunt valve versus a conventional valve for patients with hydrocephalus. Neurosurg 1999; 45:1399–1411.
17. Warf BC. Comparison of One-Year Outcomes for the Chhabra and Codman-Hakim Micro Precision shunt systems in Uganda: a prospective study in 195 children. J Neurosurg (Pediatrics 4) 2005; 102:358–362.
18. Pople, IK. Hydrocephalus. Surgery (Oxford) 2004: 22(3):60–63.
19. Dias L. Orthopaedic care in spina bifida: past, present, and future. Develop Med Child Neurol 2004; 46(9):579.
20. Buccimazza S, Molteno C, Dunne T. Pre-school follow-up of a cohort of children with myelomeningocele in Cape Town, South Africa. Ann Tropic Paediatr 1999; 19(3):245–252.
21. Steinbok P, Cochrane D, Kestle JR. The significance of bacteriologically positive ventriculoperitoneal shunt components in the absence of other signs of shunt infection. J Neurosurg 1996; 85(5):985–986.
22. Drake JM, Kestle JR, Milner R, et al. Randomized trial of cerebrospinal fluid shunt valve design in pediatric hydrocephalus. Neurosurg 1998; 43:294–305.
23. Choux M, et al. Shunt implantation: reducing the incidence of shunt infection. J Neurosurg 1992; 77:875–880.
24. Mwang'ombe NJ, Omulo T. Ventriculoperitoneal shunt surgery and shunt infections in children with non-tumour hydrocephalus at the Kenyatta National Hospital, Nairobi. East Afr Med J 2000; 77(7):386–390.
25. Turgut M, et al. Cerebrospinal fluid shunt infections in children. Pediatr Neurosurg 2005; 41:131–136.
26. Bruinsma N, et al. Subcutaneous ventricular catheter reservoir and ventriculoperitoneal drain-related infections in preterm infants and young children. Clin Microbiol Infect 2000; 6(4):202–206.
27. Kulkarni AV, et al. Cerebrospinal fluid shunt infection: a prospective study of risk factors. J Neurosurg 2001; 94:195–201.
28. Kontny U, Höfling B, Gutjahr P, et al. CSF shunt infections in children. Infection 1993; 21(2):89–92.
29. Haines SJ, Walters BC. Antibiotic prophylaxis for cerebrospinal fluid shunts: a metanalysis. Neurosurg 1994; 34(1):89–92.
30. Biyani N, Grisaru-Soen G, Steinbok P, et al. Prophylactic antibiotics in pediatric shunt surgery. Child Nerv Syst 2006; 22(11):1465–1471.
31. Ratilal B, Costa J, Sampaio C. Antibiotic prophylaxis for surgical introduction of intracranial ventricular shunts. Cochrane Database Syst Rev 2006; 3:CD005365.
32. Faillace WJ. A no-touch technique protocol to diminish cerebrospinal fluid shunt infection. Surg Neurol 1995; 43:344–350.
33. Choksey MS, Malik IA. Zero tolerance to shunt infections: can it be achieved? J Neurol Neurosurg Psychiatry 2004; 75(1):87–91.
34. Kanev PM, Sheehan JM. Reflections on shunt infection. Pediatr Neurosurg 2003; 39:285–290.
35. Braxton EE, et al. Role of biofilims in neurosurgal device-related infections. Neurosurg Rev 2005; 28(4):249–255.
36. Bafeltowska JJ et al. Therapeutic vancomycin monitoring in children with hydrocephalus during treatment of shunt infections. Surg Neurol 2004; 62(2):142–150.
37. Whittle IR, Johnston IH, Besser M. Intracranial pressure changes in arrested hydrocephalus. J Neurosurg 1985; 62:77–82.
38. Vinchon et al. Bowel perforation caused by peritoneal shunt catheters: diagnosis and treatment. Neurosurg 2006; 58(1 Suppl):ONS76-86.
39. Shehu BB, Ameh EA, Ismail NJ. Spina bifida cystica: selective management in Zaria, Nigeria. Ann Tropic Paediatr 2000; 20(3):239–242.
40. McLone DG, Dias MS. Complications of myelomeningocele closure. Pediatr Neurosurg 1991; 17(5):267–273.
41. Margaron F, Poenaru D, Bransford R, Albright L. Timing of ventriculoperitoneal shunt insertion following spina bifida closure in Kenya. Childs Nerv Sys (in print).
42. Flynn JM, Herrera-Soto JA, Ramirez NF, Fernandez-Feliberti R, Vilella F, Guzman J, et al. Clubfoot release in myelodysplasia. J Pediatr Orthopaed, Part B 2004; 13(4):259–262.
43. Doolin E. Bowel management for patients with myelodysplasia. Surg Clin N Am 2006; 86:505–514.
44. Lemelle JL. A multicentre study of the management of disorders of defecation in patients with spina bifida. Neurogastroenterol Motil 2006; 18:123–128.
45. Rintala RJ. Fecal incontinence in anorectal malformations, neuropathy, and miscellaneous conditions. Sem Pediatr Surg 2002; 11(2):75–82.
46. Christensen P, Kvitzau B, Krogh K, Buntzen S, Laurberg S, Christensen P, et al. Neurogenic colorectal dysfunction—use of new antegrade and retrograde colonic wash-out methods. Spinal Cord 2000; 38(4):255–261.

47. Ekmark E, Adams RC. The antegrade continence enema (ACE) surgical procedure: patient selection, outcomes, long-term patient management. Euro Jour Pediatr Surg 2000; 10(Suppl 1):49–51.
48. Chait PG, Shandling B, Richards HF. The cecostomy button. J Pediatr Surg 1997; 32(6):849–851.
49. Obojski A, Chodorski J, Barg W, Medrala W, Fal AM, Malolepszy J, et al. Latex allergy and sensitization in children with spina bifida. Pediatr Neurosurg 2002; 37(5):262–266.
50. Bowman RM, McLone DG, Grant JA, Tomita T, Ito JA. Spina bifida outcome: a 25-year prospective. Pediatr Neurosurg 2001; 34(3):114–120.
51. Johar A, Lim DL, Arif SA, Hawarden D, Toit GD, Weinberg EG, et al. Low prevalence of latex sensitivity in South African spina bifida children in Cape Town. Pediatr Allerg Immunol 2005; 16(2):165–170.
52. Carr MC. Bladder management for patients with myelodysplasia. Surg Clin N Am 2006; 86:515–523.
53. Lemelle JL, Guillemin F, Aubert D, Guys JM, Lottmann H, Lortat-Jacob S, et al. A multicenter evaluation of urinary incontinence management and outcome in spina bifida. J Urol 2006; 175(1):208–212.
54. Muller T, Arbeiter K, Aufricht C. Renal function in meningomyelocele: risk factors, chronic renal failure, renal replacement therapy and transplantation. Curr Opin Urol 2002; 12(6):479–484.
55. Jeruto A, Poenaru D, Bransford R. Clean intermittent catheterization: overview of results in 194 patients with spina bifida. Afr J Paed Surg 2004; 1(1):20–23.
56. Campbell JB, Moore KN, Voaklander DC, Mix LW. Complications associated with clean intermittent catheterization in children with spina bifida. J Urol 2004; 171(6, Pt 1):2420–2422.
57. Aslan AR, Kogan BA. Conservative management in neurogenic bladder dysfunction. Curr Opin Urol 2002; 12(6):473–477.
58. Gonzalez R, Schimke CM. Strategies in urological reconstruction in myelomeningocele. Curr Opin Urol 2002; 12(6):485–490.
59. Kanev PM, Park TS. The treatment of hydrocephalus. Neurosurg Clin N Am 1993; 4(4):611–619.
60. McDonnell GV, McCann JP. Why do adults with spina bifida and hydrocephalus die? A clinic-based study. Euro J Pediatr Surg 2000; 10(Suppl 1):31–32.
61. Oakeshott P, Hunt GM. Long-term outcome in open spina bifida. Brit J Gen Prac 2003; 53(493):632–636.
62. Cornege-Blokland E, Jansen HE, de Jong-de Vos van Steenwijk CCE, Poenaru D. Quality of life of children with spina bifida in Kenya is not related to the degree of the spinal defects. Tropical Med Int Health (in print). Trop Med Int Health 2011; 16 (1):30–36
63. UNICEF: The Micronutrient Initiative. Vitamin and mineral deficiency—a global damage assessment report. UNICEF, 2004.
64. Lorber J. Results of treatment of myelomeningocele. An analysis of 524 unselected cases, with special reference to possible selection for treatment. Develop Med Child Neurol 1971; 13(3):279–303.
65. Hunt GM. Open spina bifida: outcome for a complete cohort treated unselectively and followed into adulthood. Develop Med Child Neurol 1990; 32(2):108–118.