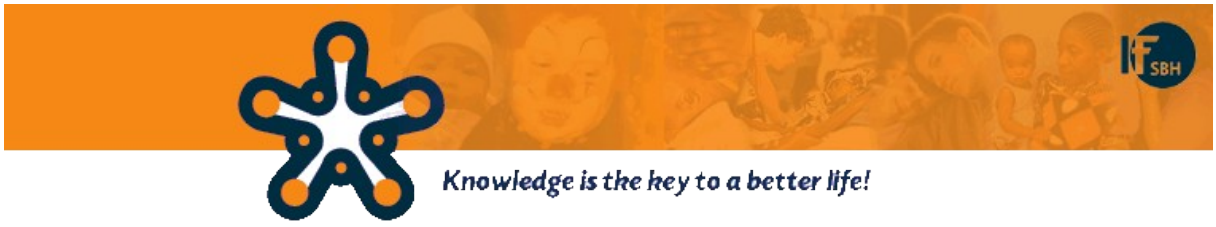

Myelomeningocele and Hydrocephalus Management Protocol for Developing Countries

Introduction

Children with myelomeningocele present various and complex management problems. However, the first two most pressing issues are: 1) the risk of meningitis and/or ventriculitis secondary to the exposed neural tissue and CSF; and, 2) hydrocephalus. The most urgent task, when the child first presents to the clinic or hospital, is that of achieving a water-tight closure of the spina bifida defect to prevent infection. Subsequently, if it is not already present, hydrocephalus is likely to develop. On a more elective basis, issues such as orthopedic deformities and neurogenic bladder dysfunction need to be addressed, as well as education of the family and establishment of an overall plan of rehabilitation to optimize the child's potential for development.

This protocol addresses only those primary issues of spina bifida closure and initial management of hydrocephalus. This suggested protocol assumes conditions typical of a developing country.

Management of newborns with myelomeningocele

Newborns should be placed on intravenous ceftriaxone (or cefotaxime) and gentamicin* immediately and the spina bifida defect covered with sterile saline – soaked gauze, which should be changed as needed to keep it moist and clean.

Regardless of whether there is CSF leak, the infant should be scheduled for urgent surgery to close the defect (ideally within 24 hours of admission), if it is medically stable.

The infant should remain on the same empiric intravenous antibiotic coverage for a total of one week. This would require modification in the event of meningitis or ventriculitis.

Anterior fontanel tension and head circumference should be monitored to assess for development of hydrocephalus. If hydrocephalus is already present at birth, the spina bifida defect should be closed first (rather than treating the hydrocephalus) in order to avoid ventriculitis. The only reason to treat hydrocephalus urgently is if the infant is symptomatic (poor feeding, vomiting, lethargy) or is manifesting symptoms of the Chiari malformation in the face of untreated hydrocephalus (stridor, frequent apnea spells, nasal regurgitation). In such cases, urgent management can be achieved with percutaneous ventricular puncture for drainage of CSF. Repeated taps are not advised, and should only be performed as needed until a shunt or tapping reservoir can be placed.

If the child develops hydrocephalus following spina bifida closure, it is ill-advised to place a VP shunt within 2 weeks of closure because of the increased risk of shunt infection. In the presence of CSF infection or the need for hydrocephalus management soon after spina bifida closure, a ventricular catheter attached to a subcutaneous tapping reservoir (or a blind-ended shunt valve tied off at its distal end) should be placed for temporary management until shunt placement is safe. External ventricular drains are more dangerous to manage on the hospital ward. In the event of CSF infection, the CSF should be sterile (as demonstrated by serial cultures) for one week prior to shunt placement.

If no hydrocephalus has developed prior to discharge from the hospital, the child needs to be monitored monthly as an outpatient at 1, 2, 3, 6 and 12 months post-operatively. Hydrocephalus can become manifest late, but typically not after 6 months. Other aspects of hydrocephalus management will be considered below.

Late presentation of myelomeningocele

Infants may present for management of myelomeningocele relatively late (weeks or even months after birth). These may present with: 1) gross infection of the sac and neural elements; 2) CSF leak or poorly epithelialized sac with no clinically apparent infection; 3) CSF leak or poorly epithelialized sac with evidence of meningitis/ventriculitis; 4) complete epithelialization of the sac with no evidence of infection or CSF leak. Except for the latter, these should all be placed on ceftriaxone (or cefotaxime) and gentamicin and scheduled for urgent surgery.

In the case of gross purulent infection of the sac and neural elements, thorough debridement of pus and inflammatory material should be performed. If the child is septic, surgery should be delayed until the condition is stabilized with medical management. If the child is paraplegic, sections of grossly infected placode or lower nerve roots hopelessly encased in pus may be resected. A culture swab should be obtained at the time of surgery prior to irrigation with antibiotic solution. After debridement, the wound and the canal proximal to the spina bifida defect should be copiously irrigated with antibiotic irrigation. The arachnoid and dura are often thick with inflammation, but primary dural closure can virtually always be achieved, and skin closure can proceed as usual. The child should continue antibiotic therapy for 14 days. The antibiotics used may require adjustment according to culture and sensitivity results.

Children presenting with CSF leak (with or without clinical infection) or a poorly epithelialized sac should be managed as described above for the newborn. In cases of meningitis and sepsis, surgery should be delayed until the child has had time to respond sufficiently to medical management and is stable for surgery. If meningitis/ventriculitis is being treated, antibiotics should be continued for 14 days. The antibiotics used may require adjustment according to culture and sensitivity results.

Children (usually weeks or months of age) with well-epithelialized sacs can undergo elective repair, and do not require a course of antibiotics, except for a single perioperative dose for staph coverage (e.g. Cloxacillin) at the time of anesthesia induction. This should not be undertaken by a surgeon with little experience, since the situation is not urgent and there is a risk of creating new neurologic deficits if any lower extremity function is present. The reasons to pursue elective repair of these lesions are: 1) untethering of the spinal cord; and, 2) removal of the sac and reconstruction of the tissues.

Symptomatic Chiari malformation

In around 5% of infants with myelomeningocele, the Chiari malformation may cause vocal cord weakness (secondary to lower cranial nerve dysfunction) leading to life-threatening airway obstruction. Early warning signs of this condition include nasal regurgitation of milk and stridor with crying. If airway obstruction becomes significant, the child may require intubation to maintain the airway until surgical management can be undertaken.

Untreated hydrocephalus should be addressed first. Decompression of hydrocephalus may relieve the problem. However, if stridor persists in the absence of untreated hydrocephalus, upper cervical

laminectomies to decompress the brainstem may be necessary. Further discussion is beyond the scope of this protocol.

Management of hydrocephalus

Management of hydrocephalus in developing countries is more difficult for many reasons. The cost of shunts may be prohibitive. The incidence of shunt infection in developed countries is around 10%, and higher for newborns. Shunt malfunction requiring surgery occurred in around 40% of patients within 2 years of initial placement in a recent prospective multicenter trial in North America. In a developing country, it can be difficult for patients to access qualified care in the case of shunt complications. Thus, shunts-dependence is more dangerous in this context.

It has been shown that endoscopic third ventriculostomy (ETV) is effective in treating hydrocephalus without the need for a shunt in most children over the age of 1 year, and in the majority of children under one year with post-infectious aqueductal obstruction (as determined by the clinical history and a small 4th ventricle on
the ultrasound or CT scan)[1]. It has subsequently been demonstrated that the addition of choroids plexus cauterization to the ETV procedure significantly increases the of treatment in infants less than I year of age that shunt dependency can be avoided in most children with hydrocephalus. [2]

Unfortunately, this technique is not available in most centers in the developing world, and shunts are the only recourse. If ETV is available in the region, the child should be referred to that center for evaluation because of the desirability of avoiding life-long shunt dependency.

Shunts produced in North American and Europe are prohibitively expensive. Some centers have been reluctant to use inexpensive alternatives. A recent prospective study has demonstrated no statistically significant difference at one year in any outcome parameter between an expensive Western-made shunt, costing $650 USD each, and a relatively inexpensive Indian-made shunt (the ChhabraTM shunt, manufactured by Surgiwear)[3]. These shunts cost about $35 USD each. The International Federation for Spina Bifida and Hydrocephalus supplies the Chhabra shunt to its project partners in developing countries.

When placing a Chhabra shunt, it is important to test the valve by first flushing it with sterile saline solution and then measuring its closing pressure by running a column of saline down a vertically-suspended length of IV tubing (or a manometer) and through the valve. The valve should close (i.e., the fluid should stop flowing) at a fluid column height of between 4 and 12 centimeters for a medium pressure valve. If the valve doesn't seem to be working properly at first (sluggish flow at high pressure or closing at a too high of a pressure), flush it again forcefully with saline and measure the closing pressure again. An occasional valve will be defective (<10% in our experience) and should be discarded. Most patients should receive a medium pressure valve.

Dr. Ben Warf

[1] Hydroceaphalus in Uganda
[2] ETV/CPC
[3] Cchabbra/codman