

Neurosurgery and Spina Bifida

How does Neurosurgery treat Spina Bifida?

As pediatric neurosurgeons, we see parents before delivery for prenatal counseling and to meet parents before the big day. On the day of birth, we evaluate the infant, get an ultrasound or CT scan to evaluate for hydrocephalus, and close the malformation on the back within 1-3 days of birth. We also assess the infant for hydrocephalus (buildup of fluid in the brain) and insert a shunt for treatment. We monitor the child for evidence of symptoms from the Chiari malformation, shunt malfunction, and spinal cord tethering, and treat these conditions if the child is symptomatic from them.

How often will the patient see Neurosurgery?

We typically see infants about once every 3-4 months during the first year, every 6 months during the next two years, and once a year thereafter for the rest of their childhood.

What test might Neurosurgery order? What does that test tell Neurosurgery?

We order tests for evaluation of shunt problems, Chiari malformations, or tethering generally only if the child is symptomatic (having problems) from these things. Remember that all children with spina bifida have abnormalities on their CT or MRI scans - spinal cord tethering is present on MRI in essentially all; Chiari malformations will be present in 98%; syringomyelia (or syrinx), a collection of fluid within the spinal cord, will be present in about 60-70%; and some hydrocephalus will be present in about 85-90%. However, the mere presence of abnormalities on imaging doesn't mean that they require treatment - pediatric neurosurgeons only treat those patients who are symptomatic - having clinical problems - related to these abnormalities. For example, only about 15% of children will have problems that require surgery for the Chiari malformation, and only about a third will need a tethered cord release (untethering or detethering). For this reason, we don't generally get radiographic studies such as CT or MRI scans unless patients are having troubles. We don't get annual CT scans 'just to check', as it has been demonstrated that routine annual CT scans are not helpful and also are expensive and result in a lot of radiation exposure for the child. We monitor the child clinically and, if they develop problems, we would get CT or MRI scans as necessary. We also generally get CT scans after any shunt revision, to ensure we have an up to date scan with the currently functioning shunt.

We may also order swallowing studies, examination of the vocal cords, strength testing by physical or occupational therapists (manual muscle testing), evaluations of urinary function (urodynamics), or scoliosis X-rays if we think there is a problem with either the Chiari or spinal cord tethering, depending upon the clinical circumstances.

Therapy - what types of therapy does Neurosurgery offer? How often might a patient need to have therapy?

Neurosurgical operations are needed for all children with spina bifida (myelomeningocele). All children with spina bifida (myelomeningocele) will have their spinal cord on the back closed at birth. About three-quarters will need to have a shunt for hydrocephalus. Shunts unfortunately are fickle and malfunction with some frequency. Statistically, 40% of shunts placed at birth will need to be changed or replaced for blockage or infection within one year of placement, 60% will need to be changed within 2 years, and 85% within 10 years of placement.

Surgery for the Chiari malformation is uncommon, with only 15% of children requiring this operation. Ninety percent will have it during the first year of life; Chiari surgery for a child with spina bifida is very uncommon after the first birthday. If Chiari symptoms develop, the first thing to check is the shunt to make absolutely certain it is working properly.

Tethered cord release is necessary in about one-third of children at some point, usually for back or leg pain, decline in muscle strength or increasing numbness or loss of sensation, urinary or bowel worsening, orthopedic deformities or scoliosis. Again, assuring that the shunt is working properly is vitally important before doing a tethered cord release, as shunt malfunction can present with any of these symptoms or signs.

Drainage of syringomyelia is rarely required in these patients, usually performed if they are declining in strength in arms or legs, or have increasing scoliosis. Again, the shunt should be checked carefully before draining a syrinx, as many of the syrinx cavities arise or enlarge because of shunt malfunction.

Mark Dias, MD