Guest Editorial

Reoperation in Spinal Dysraphism

Since the entity was first comprehensibly described in 1976, the tethered cord remains one of the most controversial pediatric neurosurgical entities with differing etiological concepts and management options. It has been well elicited by radiological and autopsy studies that the conus reaches the adult level above L1–L2 disc space at birth or at best by 3 months. A cord, which is lying lower than this level, can be considered as a low lying and most likely tethered. Although a low-ending spinal cord most often is considered as "tethered," several reports have described tethering in a normally positioned conus with demonstrated clinical improvement following detethering.

A tethered cord being chronically stretched predisposes to progressive and persistent spinal cord injury by deranged blood flow and altered metabolism. The growth and gain in height during childhood and puberty worsen the spinal traction. Experimental studies have revealed that with repetitive flexion and extension of the spine, subtle traction causes ischemic changes to the spinal cord most at the site of the attachment. The lumbar region of the spinal cord suffers the maximal brunt with lesser physical damage in the thoracic and cervical part of the spinal cord.

In a tethered cord, there are three "anchors" which tether a cord: the short filum terminale, the attachment of the cord to the adjacent structures (postsurgical adhesions in myelomeningoceles and fatty lipoma in lipomyelomeningocele) and the short nerve roots. In a low-lying cord without any previous surgery, it is thick filum which is predominantly responsible for the tethering.

Neurological deterioration is evident in most patients who are symptomatic for tethering and do not undergo surgery. A previous study found that at 12 years of follow-up, 89% of patients needed end-organ surgical repair who initially did not undergo surgery for detethering.[1] On the other hand, detethering in secondary tethered cord syndrome has been demonstrated to lead to clinical benefits. Although the rate of improvement varies among clinical studies, most studies have demonstrated either improvement or stabilization of progressively worsening symptoms or deficits. Herman *et al*. documented significant improvement in neurological deficits (63% motor and 35% bladder) at 4-years follow-up after detethering.[2] Stabilization of the progressive neurological deficits was reported in about 25% patients, and in 71%, the deficits improved.[3] However, in another report, 84% patients, the deficits stabilized and in 26% it improved.[4]

Development of tethered cord following initial surgery for myelomeningocele or lipomyelomeningocele due to scarring is often termed as secondary tethered cord syndrome. Although a low-lying spinal cord is evident anatomically and radiologically in most patients with previous myelomeningocele or lipomyelomeningocele surgery, secondary tethered cord syndrome is evident in only 15%–25% of patients. The diagnosis of secondary tethering is purely clinical as in postoperative magnetic resonance imaging, the tip of the conus has not been shown to ascend after a detethering procedure. The clinical symptoms of either cord are only seen with a small percentage of patients and these patients should be considered for detethering. Recent occurrence of scar site pain or tenderness, frequent urinary tract infections or gradually worsening neurological deficits, or development of new deficits are the common clinical picture. Development of a terminal syrinx in the lumbar region of the cord is often seen with tethering.

The most common age of presentation is between 6 and 13 years which correlates with the rapid gain in height and growth period. Children who present at younger ages with symptoms of tethering have a higher risk of tethering during their growth years requiring multiple release procedures.[5]

The role of surgery in tethered cord is somewhat controversial. It is important to differentiate between the symptomatic and asymptomatic patients with tethered cord and those who are likely to deteriorate over time. In children, it is sometimes difficult to determine if the child has subtle symptoms or is really asymptomatic. Asymptomatic children with tethered cord often are missed as it is often difficult to recognize bladder dysfunction in children. A cystometrogram should always be included. Symptomatic secondary tethered cords should be recognized early as a delayed diagnosis often is associated with permanent neurologic deficits.

The extent of detethering has been considered to correlate with the neurological outcome.[6] Although a better outcome has been reported with a complete circumferential untethering, incomplete untethering has also been demonstrated to have benefits.

The greatest challenge lies in the identification of functional neural tissue and its preservation during untethering procedure. Electrophysiological monitoring including compound action potential monitoring and

electromyographic studies during surgery have been found to be useful during untethering procedure.[7] Nonneural tissue such as scar tissue and thickened filum can be easily distinguished from functional neural tissue as they have a higher threshold for stimulation. Neurophysiologic monitoring has been reported to be effective in minimizing iatrogenic neural injury and altering the plan during the surgical procedure. Usage of CO_2 laser and FM wand has also been considered to be useful in dissecting the scar tissue in close proximity to the neural tissue. The usage of FM wand does not interfere with electromagnetic monitoring studies.

Prevention of future retethering after initial detethering has not been very successful. Some of the techniques used include construction of a capacious dural space around the untethered cord and placement of artificial grafts which prevent scarring (Gore-Tex). A capacious cerebrospinal fluid (CSF)-filled dural space has been attempted by (a) placing dural retention sutures, (b) placement of dural substitutes effective a generous duroplasty, and (c) primary pial closure in lipomyelomeningocele/myelomeningocele to minimize exposure of the raw surface. Transection of the autonomous placode (placode which is completely disconnected from the functioning spinal cord superiorly but has an intact efferent pathway to prevent retethering has also been suggested.[7]

In the present communication, the authors have reported nine patients who were operated for tethered cord with the initial surgery performed for myelomeningocele, lipomyelomeningocele, tethered cord, and dermal sinus tract.[8] The authors indicate that majority of patients were operated by nonneurosurgeons at birth or a few days later and hence possibly a complete detethering was not performed. After a repeat surgery with detethering, all patients improved to certain extent in their preoperative neurological deficits with bladder and bowel functions being improved most. The authors indicate that possibly an adequate release of the tethered cord at the initial surgery would have prevented recurrence in most of these patients.

This study raises several important issues for discussion specifically in developing countries like India where pediatric neurosurgical care is still at best localized to certain urban locations. The neural tube defects in infants with open myelomeningoceles have to be operated within 24–48 h of birth to prevent central nervous system infection. Transportation of these children is often not possible, especially if they need other supportive measures. In remote places without a pediatric neurosurgeon or a neurosurgeon, the general surgeon usually closes the defects in the first few days of life. About 15 years ago, during my tenure in a busy tertiary specialized neurosurgical center in Bangalore, India, it was very uncommon to see a newborn with a myelomeningocele or a lipomyelomeningocele in the initial 24 h after birth. However, we had several infants who were seen a month of two after birth whose defects were closed at a nearby medical facility by a nonneurosurgeon. Although it was somewhat concerning how the defect was closed, we were thankful to the surgeon who took the efforts to close the open defect within the initial 24–48 h and prevented meningitis. In my opinion, the operating surgeon went outside his usual scope of practice and saved lives for many of these unfortunate newborns.

Unfortunately, the same logic cannot be extended to children with spina bifida occulta and other closed defects such as lipomyelomeningocele and dermal sinus tracts with an intact skin cover. At the initial surgery, every attempt should be made to close the spinal cord after decompression of the lipoma to prevent a detethering. These surgeries are considered elective and always should be performed under microscope by a trained pediatric neurosurgeon or a neurosurgeon with adequate pediatric neurosurgical exposure. As mentioned before, prevention of a secondary tethering is of utmost importance in these children and every effort should be made to attain a pial closure, facilitate a patulous dural closure and duraplasty, and achieve a watertight closure to prevent a CSF leak.

The second concern is about the adequate follow-up of these children with a specific effort to identify clinical tethering and development of delayed hydrocephalus. As demonstrated in the 1980s and 1990s, adequate and timely management of hydrocephalus often is associated with near-normal or normal cognitive development leading to living as a productive adult in the society. Regarding the secondary retethering which occurs in about 15%–15% of children; identification of this group is essential as an early retethering can halt the progression or improve the neurological deficits which if untreated can become permanent.

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