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RISK FACTORS ASSOCIATED WITH CORRECTIVE SURGERY IN CONGENITAL SCOLIOSIS WITH TETHERED CORD*

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Severe neglected congenital scoliosis with tethered cord presents major difficulties in management. The primary objective of this study was to identify the risk factors associated with corrective scoliosis surgery in neglected cases presenting with severe deformity.

Six patients who presented with such problems draw attention to the importance of a staged anterior and posterior approach in order to obtain satisfactory functional results. However, corrective surgery is associated with major neurologic and systemic complications, and every effort should be made to intervene with the progression of the deformity before corrective measures becomes necessary. *Key words: tethered cord, diastematomyelia, congenital scoliosis, surgery.*

The close relationship between neural and vertebral development in the early embryological period leads to a high incidence of neurologic malformations co-existing with congenital scoliosis. Scoliosis associated with tethered cord may be due to diastometamyelia or other occult intraspinal anomalies and tight filum terminale, hence the tethering of the cord, or to congenital anomalies of the vertebral column, or to paralysis associated with spinal cord tethering[1-3]. The goal of treatment for these patients is to obtain a stable spine in reasonably good alignment over a level pelvis with preservation of normal sagittal alignment, and ideally should be performed before any neurologic problem occurs. Treatment in such cases consists of early untethering of the cord in a child with a significant growth potential, and early recognition and fusion of the spinal segments that are likely to be involved in a significant spinal deformity that may in turn lead to spinal column inbalance in the frontal or sagittal planes.

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Treatment after skeletal maturity remains controversial. Untethering of the spinal cord seldom leads to improvement in neurologic symptoms and is usually unnecessary[1, 3, 4]. Surgical correction of a spinal deformity is often very difficult even in the most experienced hands and may be associated with severe complications.

The objective of this study was to analyze the results and complications associated with surgical correction of a severe imbalance of the spinal column due to a neglected spinal deformity in a group of patients with tethered spinal cord.

Material and Methods

Nine cases of scoliosis associated with tethered cord that were operated on between 1992 and 1995 were prospectively analyzed. Another case who had corrective surgery in 1985 was added to this population. Four of these cases were skeletally immature patients who had staged untethering as well as anterior and posterior fusion operations without correction, aiming to arrest the progression of the severe deformity and neurologic involvement. The follow-up period ranged from one to 10 years for the five patients that had corrective surgery, while the sixth patient died in the third postoperative month because of respiratory problems.

The study group consisted of six patients who had corrective surgery with instrumentation for congenital scoliosis associated with tethered cord. The average age of patients was 13.8 (9-17) years. The cause of tethering was identified as diastematomyelia in four and tight filum terminale in two patients. Five patients (three with diastematomyelias and two with tight filum terminales) had untethering operations prior to corrective surgery. One patient with diastematomyelia did not have surgery for excision of the bony spur.

The average preoperative Cobb measurement was 74.8 (50-100) degrees. Fixed pelvic obliguity was present in five cases, averaging 9.3 (5-15) degrees. T1 tilt to one side averaged 23 degrees in four patients. All of the patients had frontal imbalance measured as the distance from the center of S1 to the gravity line passing from T1, and averaged 23 (12-33) mms. One patient had fixed hyperlordosis in the thoraco-lumbar area with a shift of the gravity line 27 mm posterior of the center of S1.

Two patients had only posterior surgeries, while four had both anterior and posterior. One Harrington and one Texas Scottish Rite Hospital (TSRH) instrumentation was used for the cases with only posterior surgeries with extension of the instrumentation down to the pelvis in the latter patient. ISOLA™ instrumentation was used for the other four cases with anterior and posterior multiple osteotomies, and none had to be extended down to the pelvis. The osteotomies were performed as closing wedges of the convex side of the

deformity so as not to elongate the spinal column and stretch the spinal cord, and did not coincide with the level of the diastematomyelia in any of the cases. Use of sublaminar wires was avoided in most cases unless the spinal canal could be demonstrated to be normal in diameter during surgery (Fig. 1).

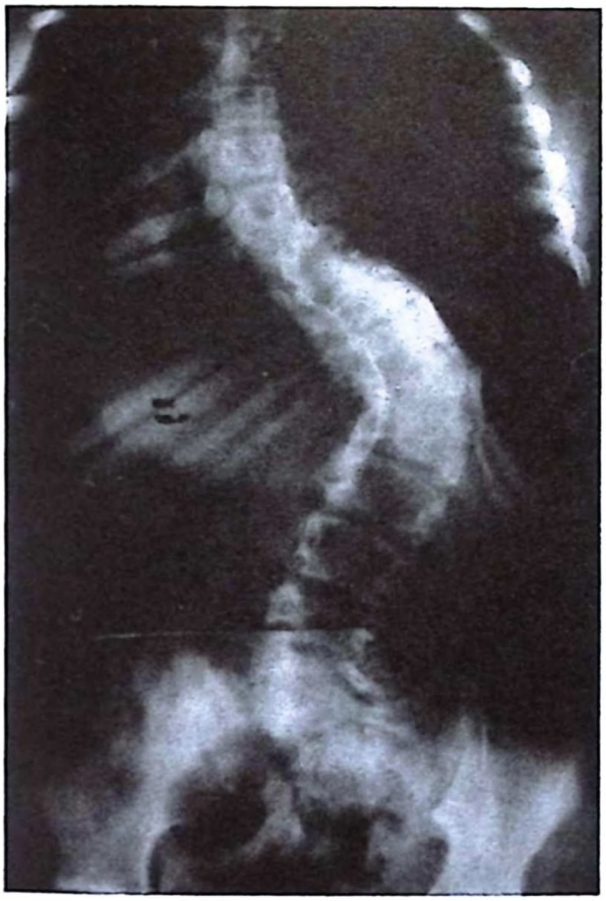

Fig. 1a: A/P radiographs of a patient who had a severe thoracolumbar curve (Th9-L3; 86 degrees).

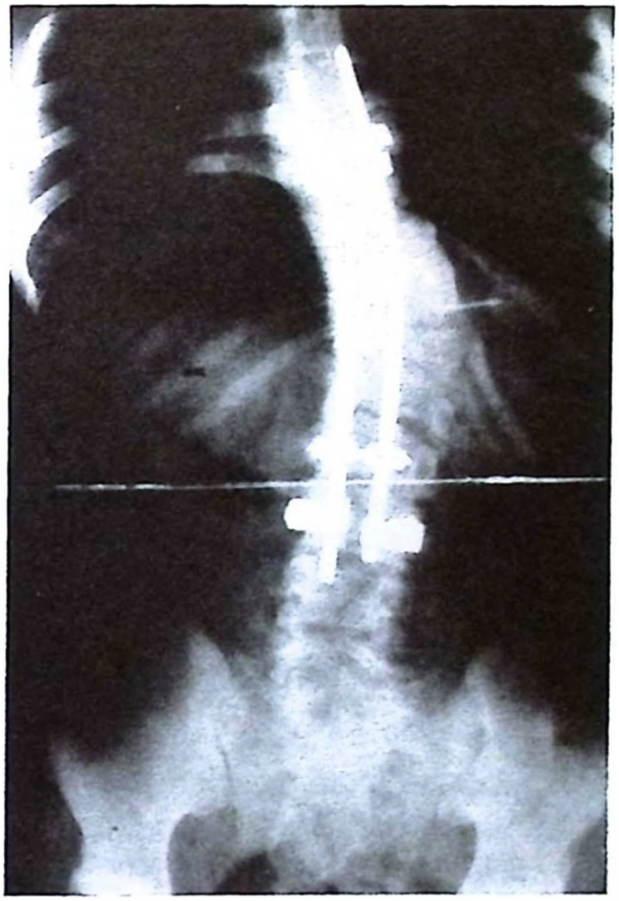

Fig. 1b: The postoperative result. Curve was corrected to 38 degrees. The spinal canal configuration in this particular case allowed for sublaminar wire application.

Results

The average postoperative Cobb angle was 40.5 degrees, with an average correction of 45.9 percent Fixed pelvic obliguity was corrected to 5.7 (0-10) degrees. T1 tilt was measured to be 23 degrees on average for the entire study group and could be corrected to 8.6 degrees in those patients with preoperative tilt, with an average correction of 63 percent. Frontal imbalance could be corrected to an average of 4.4 (0-12) mm. The sagittal balance of the patient with hyperlordosis improved to 7 mms from the plumb line.

Complications were seen in two patients. One patient had late deep infection of the instrumentation that necessitated hardware removal with mild loss of

correction (from 45 degrees to 57 degrees). Another patient wo had the most severe deformity in this group presented with a curve of 100 degrees and a forced vital lung capacity of 850 ccs. She had undergone sequential anterior-posterior surgery and demonstrated monoparesis of the left lower extremity after this intervention. Despite prompt removal of the instrumentation allowing collapse of the spinal column, her neurologic status gradually deteriorated to complete paraplegia in one week. This complication could not be attributed to specific pathology except very poor oxygenation of the cord because of respiratory problems following surgery that necessitated continuous mechanical ventilation via a tracheostomy for one week; she was weaned of the respirator in six weeks. This patient died three months after her discharge because of an acute upper respiratory tract infection.

Discussion

Diastematomyelia is the most common cause of dysraphism among the occult intraspinal anomalies coexisting with congenital scoliosis, and it is the leading cause of all spinal cord tethering[1,2]. It has been estimated that five to 16 percent of patients with congenital scoliosis have diastematomyelia. Other less common anomalies presenting with tethered cord include: neuroenteric, epidermoid and dermoid cysts; teratoma; neurofibroma; lipofibroma; fibrous bands; and a tight filum terminale.

The goal of the treatment of scoliosis associated with tethered cord, with or without neurologic problems, should be prevention of the progression of any deformity[1,3-5]. Severe congenital spinal deformities with spinal cord tethering are clinically associated with impairment of pulmonary function, compression of the abdominal viscera, and poor sitting and standing posture. Pain, increasing spasticity, and decreasing motor function of the lower extremities are indications for operative release of the tethered cord as well as spinal arthrodesis[5]. While there is ongoing controversy regarding the treatment of tethered cord without neurologic problems, it is generally accepted that symptomatic spinal cord tethering presenting with severe scoliotic deformity should be released before any attempt is made to correct the scoliosis[1,5-8]. As the presence of intraspinal abnormalities limits the ability of the spinal cord to move within the spinal canal, which is likely to occur during correction of the deformity, distraction may cause serious neurologic complications.

Formation or segmentation anomalies of the vertebrae are prone to produce an imbalance in the longitudinal growth of the spine and result in progressive congenital scoliosis. All patients with congenital curves should be monitored closely for progression of the existing deformity. Accurate assessment of neurological status should be made by serial examinations. Progression of the deformity should never be permitted and should be prevented by early fusions,

Winter et al.[2,3] suggested that in-situ arthrodesis of the spine is satisfactory for most patients presenting with congenital curves. However, for neglected cases of congenital scoliosis presenting with severe deformity and imbalance, an attempt to correct the deformity surgically may be considered. Anterior and posterior releases realised by sequential osteotomies and instrumentation may allow for major correction of the spinal deformity as well as stabilization of pulmonary function[5]. Patients with severe neglected congenital deformities present a very difficult problem surgically as well as ethically.

When dealing with the severe curves associated with congenital vertebral or neurologic anomalies, aiming to shorten the convexity of a curve, as was done in the majority of cases in this series, appears to be safer than aiming to lengthen the concavity. The only patient with harrington distraction instrumentation appeared to have been operated on before the commencement of modern imaging techniques and only luckily escaped severe neurologic complications, probably because of the modest amount of correction that could be obtained. Correction of a severely deformed spinal column is a very demanding procedure and should be performed by experienced surgeons. These operations have been associated with a high incidence of complications such as neurologic deterioration, loss of correction due to pseudoarthrosis, sepsis, and cardiopulmonary or urinary system problems. Routine intraoperative monitoring of cord function during corrective surgery is essential to avoid neural complications[3-5].

The severe complication that occurred in one patient in our series needs to be emphasized. The occurrence of neurologic involvement despite the routine wake-up test during surgery and its progression to complete paraplegia during the postoperative period while the patient was in severe respiratory distress suggest the posibility of poor spinal cord oxygenation coupled with surgical trauma as the cause. This patient, as stated above, was the most recently operated case in this series and eventually died due to respiratory problems. Our experience with this patient suggests that the general respiratory status of the patient may be the most important prognostic factor, and any severe restriction of pulmonary function before corrective surgery for congenital scoliosis should be evaluated very carefully; moreover, both the patient's and the medical center's ability to withstand such major surgery should be taken into consideration.

In conclusion, for severe cases of congenital spinal deformity associated with tethering of the spinal cord, corrective surgery remains a procedure associated with a high complication rate. Six patients who presented with such problems drew attention to the importance of a staged anterior and posterior approach in order to obtain satisfactory results. The general circulatory and respiratory condition of the patient may be major factors affecting the neurologic and overall prognosis of the patient and should be taken into consideration before surgery.

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