UNUSUAL ASSOCIATION OF CONGENITAL KYPHOSIS AND CONUS
LIPOMA PRESENTING AS A DOUBLE SPINAL CORD TETHER

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ABSTRACT
The case of a four-year-old child is described who presented to our institution with cervicothoracic deformity and a two-year history of progressive paraparesis. His past medical history was significant for meningocele which was closed at age two months. Imaging studies revealed severe congenital kyphosis with a hypoplastic T3 vertebra, as well as a tethered filum terminale with a conus lipoma. The spinal cord was found to be severely compressed at the apex of the kyphotic deformity. Discussion is focused on the diagnosis of tethered cord syndrome, and treatment options. In particular, this case required careful thought on the order of events, which followed initial tethered cord release and removal of the conus lipoma, and subsequent kyphectomy and fusion of the upper thoracic spine. A favorable clinical outcome was obtained with complete reversal of the paraparesis.

INTRODUCTION
Type I congenital kyphoses—in particular, upper thoracic—are the most deforming and have the highest neurologic risk. Rigid deformities with neurological impairment are indicated for spinal cord decompression followed by anterior and posterior fusion. Congenital malformations of the spinal axis may be associated also with occult forms of dysraphism and potentially may be a second source of spinal cord tether. The object of this paper is to report an unusual presentation of a double spinal cord tether caused by congenital kyphosis and a conus lipoma.

CASE REPORT
A four-year, ten-month-old Hispanic male child presented to us with a two-year history of unstable gait, frequent falls and diminished strength in both lower extremities, accompanied by progressive kyphotic deformity in the cervicothoracic junction. At two months of age he had been subject to surgical excision of a lumbar meningocele. His parents were unaware of any bowel or bladder dysfunction.

On physical examination the child had a midline scar in the lumbar region and a prominent cervicothoracic kyphosis. He presented with a wide-based gait and poor balance, with bilateral atrophy of the gluteal, thigh and calf muscles. Radiographs showed a thoracic kyphosis of 90° with T3 apex, thoracolumbar scoliosis and widened lumbar pedicles (Figure 1).

During the following six weeks, he became unable to stand without assistance, and developed a positive Babinski sign. Urologic evaluation revealed a hypotonic bladder with incomplete voiding. CT and MRI further demonstrated severe kyphoscoliosis of 138° with cord compression at T3 and abnormal cord signal (Figure 2), and a conus lipoma at T11-T12 with cord tethering (Figure 3).

A two-stage procedure was indicated with: (1) Tethered cord release and resection of the conus lipoma, which was found to be anchored to the dorsal dural sac (Figure 4); (2) Cord decompression at T2-T4 two weeks later, with kyphectomy and fusion of the cervicothoracic junction. The spine was exposed through a T-shaped incision and costo-transversectomy of the second through fourth ribs (Figure 5). Following subperiosteal exposure

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of T2-T5, the T2-T4 pedicles were removed, and the intervening nerve roots were dissected and preserved. Three hemivertebrae were excised with rongeurs and curetted back to the posterior cortex. The spinal canal was entered at the T2-T3 disc space and bone was removed from convex to concave, toward the apex. An anterior rib strut was placed between T1-T5. Posterior in situ fusion from T1-T5 was then carried out through the same incision, with rib autograft and morselized allograft. The patient was placed in a Minerva brace for six months until fusion was certain (Figure 6).

At the two-month follow-up visit neurological recovery was already evident, and at six months his neurological assessment was normal. At his last visit three-and-a-half years after surgery, he continued to be neurologically intact and fully active in school, including soccer and other sports.

**DISCUSSION**

Congenital kyphosis secondary to formation and/or segmentation defects progresses, especially during the adolescent growth spurt, and may result in severe deformity and spinal cord compression.\(^1\)\(^-\)\(^6\) McMaster\(^2\) described that 7 of 68 patients developed spontaneous neurological deterioration. Kyphosis was the most prevalent deformity (42 of 43 patients) causing neurological deficit in the review of Lonstein et al.\(^2\)\(^2\) Congenital kyphosis is classified based on anatomical characteristics: Type I—Anterior failure of vertebral-body formation; Type II—Anterior failure of vertebral-body segmentation, or; Type III—A combination of both. Type I deformities— in particular, upper thoracic deformities like the present case—are the most deforming and have higher neurologic risks.\(^2\)^\(^3\)
Successful management depends on recognizing poor prognosis at an early stage (age <5 years), and on balancing spinal growth by means of a posterior arthrodesis with a kyphosis of < 45°. Anterior and posterior fusions are indicated in Type I deformities when kyphosis exceeds 50° in children >3 years old. Early treatment of the deformity is relatively straightforward and provides excellent results, whereas late treatment is difficult and the results are usually less than ideal. Once neurologic symptoms develop, treatment depends on the onset of symptoms and flexibility of the curve. If the onset of symptoms is <3 years of age and the apex is flexible, the deformity may be progressively improved with distraction followed by fusion if neurological symptoms improve satisfactorily. In rigid deformities and/or if immobilization and rest do not result in neurological recovery, then decompression of the spinal cord, followed by anterior and posterior fusions, is indicated. Decompression of the spinal cord in kyphotic deformities is risky because of the potential for anterior migration of the cord, which is significantly increased in the presence of a distal tether. For this reason, tethered cord release was chosen as the first procedure.

Congenital malformations of the spinal axis may also be associated with occult dysraphism and malformations of other systems or organs. Myelodysplasia is often associated with tethered cord syndrome (TCS), a broadly used term for progressive neurological deterioration localized to lower spinal abnormalities (fibrous bands, adhesions, thickened filum terminale, diastematomyelia, or intradural lipomas) resulting in traction of the conus medullaris.

Symptoms of TCS develop gradually as a product of disproportionate longitudinal growth between the vertebrae and the tethered cord, resulting in stretching of the conus medullaris and nerve roots. Clinical manifestations include back pain, sensory disturbance, gait deterioration, contractures of lower extremity muscles and increasing foot deformities (pes cavus, pes adduc-
tus). Progressive scoliosis, neurogenic bowel and/or bladder, and frequent urinary tract infections (impaired bladder compliance) are also common manifestations. Hyperreflexia associated with motor dysfunction or the Babinski sign can be found in 10-17% of patients with TCS.\textsuperscript{7,8} The initial evaluation includes a thorough neurological examination and appropriate imaging. Spina bifida occulta is found in 90% of patients with tethered cord syndrome. Alternatively, few children with incidental radiologic findings of spina bifida occulta have cord tethering. An elongated spinal cord caudal to L2 and thickened filum terminale (>2 mm) are the most common findings.\textsuperscript{7,8} TCS occurs in 3-15% of patients with history of a repaired meningocele.\textsuperscript{10,11} Management of patients with TCS is controversial,\textsuperscript{7,15} and there are two different approaches: (1) Prophylactic release of the tethered cord,\textsuperscript{14,15} or; (2) Observation until neurological signs develop. Reversal of upper motor neuron symptoms may be poor once neurological signs and/or orthopedic deformities are detected, and prophylactic surgery has shown better results.\textsuperscript{15} On the other hand, not all patients with tethered cord develop clinical symptoms, and low-lying conus does not necessarily translate into TCS. For this reason, some authors advocate close surveillance to determine the need and timing for surgical untethering.\textsuperscript{19} Prolonged or accentuated neuronal dysfunction may lead to structural damage to the neuronal perikarya and axons. Untethering improves the oxidative metabolism of the cord,\textsuperscript{20,21} and gait has a greater chance for improvement than bladder function.\textsuperscript{11} In the present case, bladder function normalized, as did the gait disturbance.

REFERENCES


