

## **Progressive paraparesis in 14 - year-old male with kyphoscoliosis: A case report**

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### **ABSTRACT**

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A 14-year-old male presented with congenital kyphoscoliosis along with progressive paraparesis. Radiographs confirmed kyphoscoliosis and magnetic resonance imaging revealed a stretched and flattened spinal cord over the kyphotic deformity and a T6 hemivertebra. Before the surgical treatment the patient had clinical signs and symptoms of paraparesis. A gradual deterioration in

the neurologic status was observed and patient became paraplegic after the surgery. Currently, the patient moves in a wheelchair, has a pyramidal syndrome of the lower limbs and neurogenic bladder.

**Key words:** congenital kyphoscoliosis, spastic paraparesis

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## INTRODUCTION

The first description of congenital kyphosis was by Von Rokitsansky in 1844; however, it was not until 1932 that Van Schrick differentiated a failure of vertebral-body formation from a failure of vertebral-body segmentation as a cause of congenital kyphosis. In 1955, James described twenty-one patients who had a congenital kyphoscoliosis; five became paraplegic [1].

Congenital scoliosis is the abnormal development of the spine resulting in combination of missing portion, partial formation, or lack of separation of the vertebrae [2]. It is caused by anomalous vertebral development in the embryo. Congenital scoliosis accounts 10% of all scoliosis and require for the corrective surgery [3]. The frequency of congenital scoliosis is approximately 1 in 1,000 births [4]. Congenital kyphosis and kyphoscoliosis are much less common than congenital scoliosis. They are potentially more serious because compression of the spinal cord and paraplegia sometimes develop. It has been postulated that congenital kyphosis or kyphoscoliosis needs to be addressed surgically before the age of 5 years and before the kyphosis exceeds 50°. The vertebral malformations spanned the length of the entire spine and were classified as butterfly vertebrae, segmentation defect, hypoplasia and hemivertebrae. Butterfly vertebrae were defined by the presence of sagittal cleft. These deformities have a tendency to progress with age [5]. The natural history depends on the type of anomaly and the location of anomaly [6]. Patient evaluation focuses on the history and physical examination, followed by appropriate imaging modalities [7]. The hallmark of surgical treatment is early intervention before the development of large curvatures. Early identification and management of concomitant defects can improve the patient's quality of life [8].

In this case report, we present a 14 - year-old male child having with congenital kyphoscoliosis with paraparesis underwent posterior correction of curve with instrumentation.

## CASE PRESENTATION

A 14 -year- old male was born by vaginal route after at a 37-week gestation with weight of 2400 g. The Apgar score was 10. Neonatal period was complicated by sepsis and septic arthritis of the left hip. He fed soya milk until 3 years due to sensitive to cow's milk. Milk-restricted diet was continued up to 6 years of age. Parameters of physical development were abnormal: weight and height were below the 3 rd percentile. At the age of

2 years of age due to congenital kyphoscoliosis was under care of Orthopedic Clinic.

Since the age of seven he was treated with Milwaukee brace and leveling pad due to limb-shortening about 1, 5 cm. He had a lower physical development. The skeletal age was delayed to chronological age by a mean of 2 years. The densitometry results were below the normal age (- 2.55 SD in the lumbar spine). At the age of 12 years locomotor deficits increased and shortening of the left leg 4cm was observed. The radiological investigation confirmed the presence of a double thoracic kyphoscoliosis. In November 2010, a magnetic resonance imaging scan revealed kyphoscoliosis at Th3-Th6. apex of the kyphosis). In December 2010, a child neurologist recognized spastic paraparesis. In March 2011, preoperative MRI evidenced cord compression at the level of T5-T6 and a flattened cord with signal changes over the apex of kyphotic deformity 110°. (Figure 1). His paraparesis had worsened and he had difficulty in walking.



**Figure 1.** Preoperative T2 weighted magnetic resonance imaging shows an overstretched flattened cord with signal changes over kyphotic deformity at Th5-Th6 in sagittal.

There was no history of trauma. A surgical correction and instrumented posterior arthrodesis was decided based on the severity of the neurological deficit. Postoperatively the patient developed paralysis of the lower limbs. The patient was treated several times in our institution.

Currently, the patient moves in a wheelchair and has spastic paraparesis below Th8. The superficial sensation is diminished below the Th8 dermatome. Knee and ankle reflexes are brisk bilaterally, clonus is present in both ankles as well. He is reporting no sensation of bladder filling and

catheterization is performing 5 times a day. The urodynamic test confirmed overactive bladder of neurogenic origin. Somatosensory evoked potentials revealed prolonged latency cortical component of the cortical and central conduction time suggesting impairment of somatosensory stimuli conduction between the level of L1 and the sensory cortex.

## **DISCUSSION**

Rapid onset of paraplegia after kyphoscoliosis correction is a well-known fact, but in the present report, we described a case wherein gradual deterioration in the neurologic status was observed before and post deformity correction surgery [8-10].

Direct mechanical trauma to the cord during the surgery could have triggered off the neuronal apoptotic changes. It is suggested that severity of anterior angulation of spine is proportional to the pressure within the canal, which in turn may cause vascular insufficiency to the cord as it increases, thereby leading to an altered neurology [11].

Another experimental study suggests that kyphotic angle leads to flattening of the cord over the deformity thereby causing demyelination of anterior funiculus, neuronal loss and atrophy of anterior horn and decreased vascularity at the ventral side of compressed spinal cord [12].

According to Shenouda et al. [13] a kyphotic spine should be decompressed anteriorly so that the tense flattened cord translates into the vertebral body thereby achieving decompression.

There is no accurate method for predicting an unfavorable outcome such as postoperative deteriorated neurologic status after deformity correction surgery and despite all the precautions and preventive measures, unexplained deterioration of neurology can occur and the spine surgeons should be aware of this phenomenon.

No one operative procedure can be applied to all types and sizes of deformity. The method of surgical treatment depends on the age of the patient, the type and size of the deformity, and the presence or absence of spinal cord compression causing a neurologic deficit [14]

All patients with congenital kyphosis or kyphoscoliosis should be treated by a posterior arthrodesis before the age of 5 years and before the kyphosis exceeds 50 degrees. A kyphosis that does not reduce to less than 50 degrees as measured on the lateral spine radiograph made with the patient supine requires an anterior release and arthrodesis with strut grafting followed by posterior arthrodesis with instrumentation (if possible) [15].

In our patient kyphoscoliosis was caused by defects of segmentation and vertebral wedge

deformity of Th4-Th6. Family history has been identified as normal.

A family history of congenital spinal deformity is rare. Winter and colleagues found that only 13 of 1250 patients had a positive family history of such deformity. Studies of twins usually have shown that if one twin has an anomaly, the other does not, even if the twins are identical. However, several investigators have described hereditary congenital scoliosis [16].

In addition to spinal segmentation defects, segmentation defects often occur in the ribs, leading to the development of a small, stiff thorax and, frequently, to pulmonary compromise. Almost 20% of patients with congenital scoliosis develop the neurological deficits [6,17].

Clinically significant sagittal deformities can also lead to severe pain, problems with swallowing, gastrointestinal and cardiopulmonary complications. When the kyphotic deformity exceeds a certain point and rehabilitation therapy is no longer sufficient surgical intervention is indicated [14,15].

This case is interesting because of the severity of the symptoms. The patient had a progressive bilateral spastic paraparesis and finally a spastic paraplegia.

## **CONCLUSIONS**

An early diagnosis of congenital kyphosis or kyphoscoliosis is very important. They can progress rapidly, resulting in severe deformity and possible neurological deficits. In-depth knowledge of the natural history of the disease is necessary to plan the treatment and prevention of complications. In progressive congenital kyphosis, surgical treatment is mandatory for a successful result. Rehabilitation should be carried out as an individual diagnostic and therapeutic program.

### **Conflict of interest**

The authors declare that they have no conflict of interest related to the publication of this manuscript.

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