# Late Onset of Thoracic Myelopathy with Type 2 Congenital Deformity: A Case Report

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**Abstract:** *Background*: Although there are several reports of paralysis developing after aggravation of the deformities of congenital kyphosis or kyphoscoliosis, the onset of myelopathy in middle-aged patients is rare. *Objective*: To describe a case of late onset thoracic myelopathy in a patient with anterolateral fused vertebrae. The progression of kyphoscoliosis in this middle-aged patient preceded the onset of myelopathy. *Methods*: A case of thoracic myelopathy in a 51-year-old female suffering from kyphoscoliosis resulting from anterolateral fused vertebrae between T10 and T12. The patient was treated surgically by costotransversectomy to allow spinal cord decompression and T7–L2 posterior stabilization. *Results*: The patient was successfully surgically treated using costotransversectomy for spinal cord decompression and extensive posterior stabilization. *Conclusion*: Type 2 congenital kyphosis may lead to thoracic myelopathy in a middle-aged patient.

**Keywords:** Kyphoscoliosis, paraplegia, spinal cord compression, congenital scoliosis.

## INTRODUCTION

Although there have been several reports that paralysis developed from aggravation of the deformity of congenital kyphosis or kyphoscoliosis, the onset of myelopathy in middle-aged patients is rare [1]. We report a case of onset of thoracic myelopathy in a 51-year-old female suffering from kyphoscoliosis due to an unsegmented anterolateral bar.

## CASE REPORT

A 51-year-old woman presented at our hospital with lower leg numbness that had persisted for two months. Her medical history stated that she was born by normal delivery and had no abnormalities at birth. Scoliosis was found when she was a junior high school student, but was not treated. An examination revealed that she had a limp and lower limbs paresthesia with slight paralysis and exaggerated patellar and Achilles tendon reflexes without bladder or bowel disturbance. Standing roentgenograms showed 46 degree scoliosis and 60 degree kyphosis in T9-L1. Because her neurological signs improved with the use of a molded plastic thoraco-lumbo-sacral barace, she refused surgery at that time.

Eight years after the initial visit, at age 59, she returned because of difficulty in ambulation and urinary incontinence. Her ambulation was severely impaired by spasticity, ataxia, and slight muscle weakness in both legs. Her Japanese Orthopaedic Association (JOA) score for thoracic spine function was 2.5. The JOA system, an 11-point scale measuring lower-extremity motor function, lower-extremity and trunk sensory function, and bladder function is listed in Table **1** [2].

**Table 1. Summary of the Modified JOA Scoring System for Assessment of Thoracic Myelopathy**

| Neurological Status Score | |
|---|---|
| Lower-limb motor dysfunction | |
| Unable to walk | 0 |
| Able to walk on flat floor w/walking aid | 1 |
| Able to walk up/downstairs w/handrail | 2 |
| Lack of stability & smooth reciprocation of gait | 3 |
| No dysfunction | 4 |
| Lower-limb sensory deficit | |
| Severe sensory loss or pain | 0 |
| Mild sensory deficit | 1 |
| No deficit | 2 |
| Trunk sensory deficit | |
| Severe sensory loss or pain | 0 |
| Mild sensory deficit | 1 |
| No deficit | 2 |
| Sphincter dysfunction | |
| Unable to void | 0 |
| Marked difficulty in micturition | 1 |
| Minor difficulty in micturition | 2 |
| No dysfunction | 3 |

JOA: Japanese orthopedic association.

Standing roentgenograms revealed an increase in her kyphoscoliosis at T9-L1 to 52 degrees scoliosis and 72 degrees kyphosis. Computed tomography (CT) and three-

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dimensional computed tomography demonstrated (i) an anterolateral unsegmented bar from T10 to T12; (ii) aplasia of the T10 and T11 lamina, T10 and T11 pedicles on the right side, the T11 spinous process, and the right 10th and 11 ribs; and (iii) hypoplasia of T10 and 12 spinous process and the right 12th rib (Figs. **1, 2**). Disc space narrowing and spur formation were seen at T9/10 and lateral translation was also seen at T12/L1. CT after myelography revealed a highly atrophic spinal cord at the T10/11 level (Fig. **3**, left).

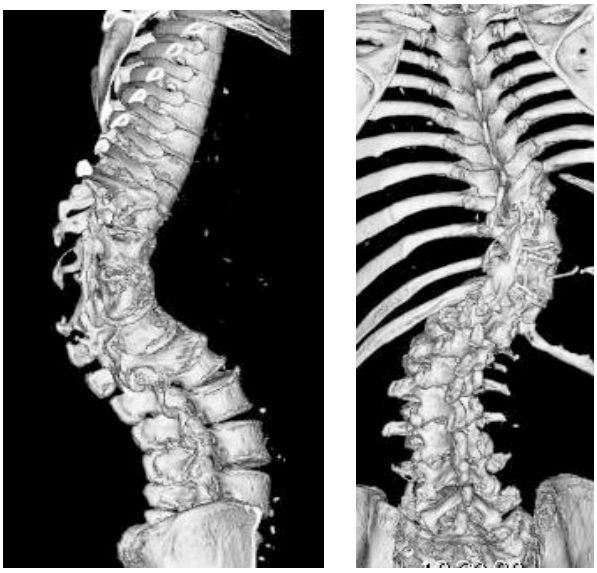

**Fig. (1). Three-dimensional computed tomograms of the thoracolumbar spine.** A lateral image viewed from the right side shows kyphotic deformity with laminar and spinous process defects (left). In a posterior view, right convex scoliosis with aplasia or hypoplasia of the right 10th through 12th rib is illustrated (right).

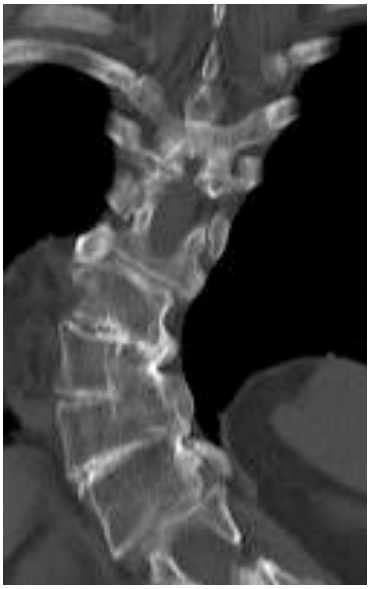

**Fig. (2). A coronal reconstructive computed tomogram of the thoracic spine.** An anterolateral unsegmented bar from T10 to T12, spondylotic changes at the upper adjacent segment (straight arrow) and lateral translation at the lower adjacent segment (dotted arrow) are illustrated.

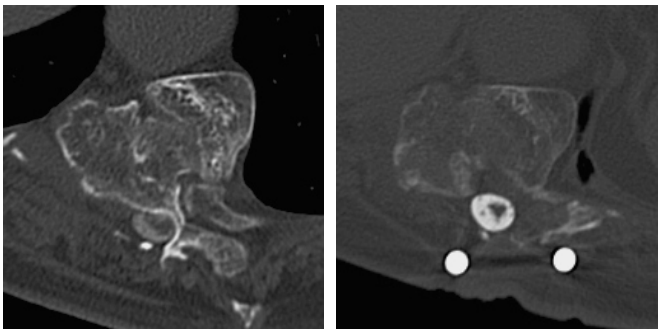

**Fig. (3). Pre- and post-surgical computed tomograms after myelography at the T11 level.** The pre-surgical image shows a laminar defect and a highly atrophic spinal cord (left). The spinal cord was completely decompressed by resection of the posterior part of vertebra by use of costotransversectomy (arrows, right).

Posterior surgery was performed to treat the diagnosis of myelopathy with congenital kyphoscoliosis. After laminectomy and costotransversectomy of T10 and T11, the spinal cord was decompressed by resection of the posterior vertebra at T10 and 11. Posterior in situ fusion and instrumentation from T7 to L2 were performed. Immediately after surgery, the patient's leg symptoms improved and she could walk without difficulty. Fourteen months after the surgery, her JOA score was 11 and CT after myelography confirmed adequate decompression at the T10/11 level (Fig. **3**, right). Postoperative roentgenograms showed a slight reduction of the kyphoscoliosis to 39 degrees scoliosis and 63 degrees kyphosis (Fig. **4**). Post surgical courses have been uneventful during the 14 months after surgery.

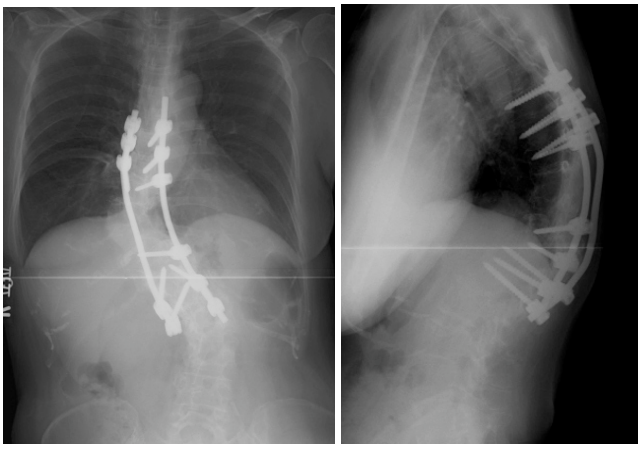

**Fig. (4).** Antero-posterior (left) and lateral (right) standing roentgenograms taken 14 months after the instrumentation surgery. The Cobb measurement shows the 39 degree scoliosis and 63 degrees kyphosis in T9-L1.

## DISCUSSION

Myelopathy rarely develops in patients who only have scoliosis. When kyphosis coexists, myelopathy frequently develops during the growth phase, which aggravates the kyphosis and surgery is required [1]. In kyphosis or kyphoscoliosis, the spinal cord may be stretched at the top of the curvature and is compressed by the vertebral body in

front and the dura and/or thickened soft tissues from the back [1]. Ischemia in the spinal cord may contribute to the development of myelopathy [3, 4]. Based on his experience with forty-three young patients showing rapid progression of kyphosis, Lonstein reported that neurological deficits occurred at an average of 95 degrees kyphosis (range: 50-180 degrees) [1].

McMaster classified congenital spinal deformity into three groups; failure of formation (Type 1), failure of segmentation (Type 2), and a mixture of failure of formation and segmentation (Type 3) [5]. Type 2 consists of an anterior unsegmented bar and an anterolateral unsegmented bar. Compared to the failure of formation found in Type 1, failure of segmentation (Type 2) is less likely to show progression [5]. There is only one previous report of a 17-year-old male who developed myelopathy because of an unsegmented bar [4].

There have been several reports examining the progression of congenital kyphoscoliosis in middle-aged patients [6-8]. McMaster stated that the curvature resulting from an unsegmented bar is not aggravated after the growth phase [6]. To our knowledge, there is only two case reports of the late onset of myelopathy, which were in a 41-year-old and a 48-year-old [1, 9]. In our case, progressive degeneration occurred at adjacent segments above and below the unsegmented bar. Postmenopausal osteoporosis and muscle weakness are probably responsible for the aggravation of the patient's kyphosis [10, 11].

The usual treatment for myelopathy of congenital kyphoscoliosis in youths involves correction of alignment of kyphoscoliosis with fusion [1, 12, 13]. In our case, anterolateral decompression of the spinal cord was performed under direct vision by use of costotransversectomy [14]. Because of the coexistent rotation of the unsegmented bar, anterior decompression through costotransversectomy was not technically difficult. To avoid further progression of the deformity and to preclude future neurological aggravation, extensive fusion was performed using instrumentation. Considering the rarity of myelopathy and successful surgical results even in the case of severe neuropathy, early surgery may not be necessary for Type 2 congenital kyphoscoliosis.

## DISCLAIMER

Reprints will not be available from the authors.

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## REFERENCES

[1] Lonstein JE, Winter RB, Moe JH, *et al*. Neurological deficits secondary to spinal deformity. A review of the literature and report of 43 cases. Spine 1980; 5: 331-55.

[2] Japanese Orthopaedic Association: [Scoring system for cervical myelopathy]. Jpn J Orthop Assoc 1994; 68: 490-503.

[3] Dommisse GF. The blood supply of the spinal cord. A critical vasculature in spinal surgery. J Bone Joint Surg Br 1974; 56(2): 225-35.

[4] Khanna N, Molinari R, Lenke L. Exertional myelopathy in Type 2 congenital kyphosis. Spine 2002; 27: E488-E492.

[5] McMaster MJ, Singh H. Natural history of congenital kyphosis and kyphoscoliosis. A study of one hundred and twelve patients. J Bone Joint Surg Am 1999; 81: 1367-83.

[6] McMaster MJ. Spinal growth and congenital deformity of the spine. Spine 2006; 31: 2284-87.

[7] Winter RB, Turek-Shay LA. Twenty-eight-year follow-up of anterior and posterior fusion for congenital kyphosis. A case report. Spine 1997; 22: 2183-7.

[8] Winter RB, Lonstein JE. Congenital thoracic scoliosis with unilateral unsegmented bar and concave fused ribs: rib osteotomy and posterior fusion at 1 year old, anterior and posterior fusion at 5 years old with a 36-year follow-up. Spine 2007; 32: 841-4.

[9] Sato T, Kokubun S, Tanaka Y, Aizawa T. Paraparesis associated with mild congenital kyphoscoliosis in an adult. Tohoku J Exp Med 1997; 183: 303-8.

[10] Cutler WB, Friedmann E, Genovese-Stone E. Prevalence of kyphosis in a healthy sample of pre- and postmenopausal women. Am J Phys Med Rehabil 1993; 72: 219-25.

[11] Fon GT, Pitt MJ, Thies AC Jr. Thoracic kyphosis: range in normal subjects. Am J Roentgenol 1980; 134: 979-83.

[12] Kim YJ, Otsuka NY, Flynn JM, Hall JE, Emans JB, Hresko MT. Surgical treatment of congenital kyphosis. Spine 2001; 26: 2251-7.

[13] McMaster MJ, Singh H. The surgical management of congenital kyphosis and kyphoscoliosis. Spine 2001; 26: 2146-55.

[14] Smith JT, Gollogly S, Dunn HK. Simultaneous anterior-posterior approach through a costotransversectomy for the treatment of congenital kyphosis and acquired kyphoscoliosis deformities. J Bone Joint Surg Am 2005; 87: 2281-9.

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