

CASE REPORT

Rare Sagittal Compensatory Balance in Neuromuscular Scoliosis Complicated by Hydrocephalus: A Case Report

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ABSTRACT

Objective • This case report aims to present a rare case of thoracic lordosis and lumbar kyphosis and describe the posterior instrumented scoliosis correction performed.

Case presentation • A 59-year-old female presented with low back pain. She had undergone ventriculoperitoneal shunt placement 8 years ago. I scored 76 on the Wechsler Adult Intelligence Scale. MRI of the lumbar spine showed spinal canal stenosis at L3/4, L4/5, and L5/S1. Full spine X-ray revealed thoracic lordosis and lumbar kyphoscoliosis, the coronal imbalance, and the sagittal compensatory balance. In order to avoid the risk of brain swelling and paraplegia, pedicle subtraction osteotomies

(PSO) in the L2 and lumbar posterior instrumented scoliosis correction were performed under electroencephalogram and neuro electrophysiological monitoring. Shoulder imbalance was observed 1 year after surgery, but there was no loss of lumbar correction.

Conclusion • In future cases of complex spinal deformity, it is important to observe whether there is cerebral ventricular dilatation on MRI before the operation. If severe thoracic lordosis is combined with lumbar scoliosis, over-correcting the lumbar scoliosis should be avoided to prevent shoulder imbalance. (*Altern Ther Health Med.* [E-pub ahead of print.])

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INTRODUCTION

During the embryonic period, the human spine acquires a specific morphology and stability. As the infant learns to walk upright, the spine gradually assumes four physiological curvatures in the sagittal plane, which serve to distribute the weight of the body and reduce pressure on the spine. As D. Polly stated that, "Life is a kyphosing event", Sebaaly et al.¹ described 11 degenerative spinal morphologies based on the Roussouly classification of the sagittal spinal morphology. Spinal reverse arch refers to the physiological curvature that bends in the opposite direction. Compared with cervical kyphosis and lumbar kyphosis, the incidence of pure thoracic lordosis is lower, and it is more common in neuromuscular scoliosis and adolescent idiopathic scoliosis. At present, the definition of thoracic lordosis is still unclear; some scholars have defined the thoracic kyphosis angle <0° as severe thoracic lordosis, while the T5-T12 kyphosis angle between 0° and

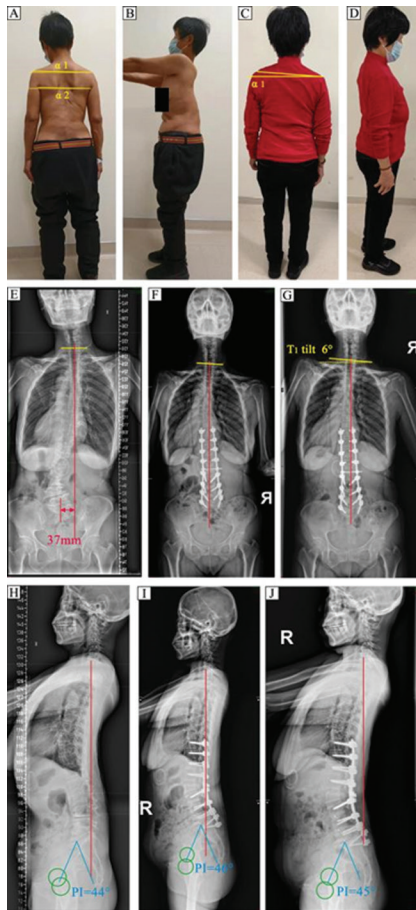
10° is defined as thoracic reduced kyphosis (i.e., invisible thoracic lordosis).² Cerebrospinal fluid (CSF) is a colorless, aqueous, and nutrient-rich liquid, which contains molecules that regulate embryonic development and is essential for maintaining the homeostasis and metabolism of the central nervous system. Hydrocephalus alters the CSF hydrodynamics, and spinal developmental malformation caused by pressure differences within the intracranial and intraspinal is the generally accepted etiology hypothesis for the development and progression of scoliosis by scholars at home and abroad.³ Qi et al.⁴ first proposed long-standing overt ventriculomegaly in adults (LOVA) in 1996. Clinical symptoms such as increased intracranial pressure and mental retardation did not appear until adulthood. It is inferred that aqueductal stenosis may be the potential cause of LOVA.⁵

We report a case of neuromuscular scoliosis complicated by following ventriculoperitoneal shunt placement for hydrocephalus. A quite satisfactory clinical outcome of correcting lumbar kyphosis deformity was achieved by posterior instrumented scoliosis correction. This case provides evidence for the correlation between CSF circulation disorder and spinal deformity.

CASE PRESENTATION

A 59-year-old woman, thoracic lordosis was found since childhood with growth and development lag behind her peers,

Figure 1. Appearance and radiographic assessment. Preoperative appearance, α_1 is shoulder angle, α_2 is axilla angle(A, B), postoperative appearance(C, D), x-rays in coronal plane on pre-op (E), post-op 3 month (F) and post-op 1 year (G), x-rays in sagittal on pre-op (H), post-op 3 month (I) and post-op 1 year (J)



low back pain started at about 40 years of age. A diagnosis of hydrocephalus was made at the age of 51 years with binocular blurry vision and underwent ventriculoperitoneal shunt placement. Previous history of hyperlipidemia and asymptomatic cerebral infarction. She denied any history of spinal deformity or hydrocephalus in her parents and three children. Specialist examination: claudication, height 153 cm in standing position, weight 53 kg, the unequal height of the bilateral shoulder, the thoracic lordosis, left lumbar kyphoscoliosis (Figure 1), the spinous process and paravertebral tenderness of the lumbosacral were obvious, the lumbar movement was limited, and the bilateral Babinski sign was negative. The patient's intelligence status is within the normal range, slightly below the average level, with a score of 76 on the Wechsler Adult Intelligence Scale The full spine X-ray show the coronal imbalance and concave coronal malalignment (type 1),⁶ and the sagittal compensatory balance (Figure 1). A lumbar spine MRI displays L3/4, L4/5, and L5/S1 spinal canal stenosis. Digital 3D reconstruction of the whole spine did not reveal spina bifida (Figure 2). The whole spinal cord MRI did not reveal any developmental malformation of the spinal cord.

Figure 2. 3D models by MIMICS software. Preoperative 3D model on the front, back, left and right (A, B, C, D), 3D models of PSO in L2 on pre-op and pre-op 10 day (E, F), postoperative 3D model on the front and left (G, H)

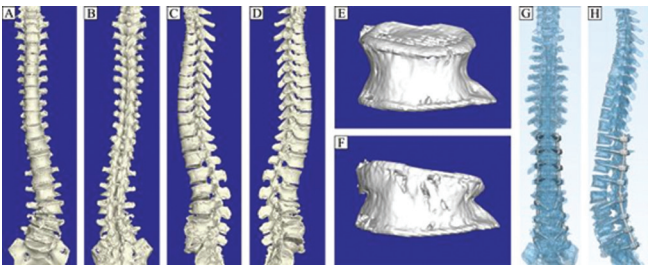


Table 1. Radiographic assessment

	Pre-op	Post-op 3 month	Post-op 1 year
thoracic scoliosis (°)	-25	9	10
lumbar scoliosis (°)	35	-1	-2
thoracic kyphosis, TK(°)	-42	-13	-16
lumbar lordosis, LL (°)	-1	33	30
sacral slope, SS(°)	22	23	22
elvic tilt, PT(°)	22	23	23
PI-LL	45	14	15

Note: The "-" represents the direction, specifying the scoliosis to the right and reverse physiological curvature as "-".

Table 2. ASIA, VAS, ODI and JOA of patient before and after surgery

	Pre-op	Post-op 3 month	Post-op 1 year
ASIA	C	D	D
VAS	7	2	1
ODI (%)	52	22	12
JOA	12	21	23

ECG: heart rate 53 bpm, sinus bradycardia, atropine test was negative. Echocardiography: LVEF 57%. Pulmonary function: VC 2.34L, FEV1 115.4%, FEV1/FVC 90.36%.

After successful general anesthesia, the patient is placed prone on the surgical bed with a soft cushion placed on the chest. The pedicle screws were placed from T11 to S1 by using the anatomic freehand technique under electroencephalogram and neuroelectrophysiological monitoring. L3/4, L4/5, L5/S1 were decompressed, and the PSO osteotomy was performed on L2 by using the eggshell osteotomy technique with an ultrasonic bone scalpel to correct the lumbar kyphoscoliosis (Figure 2). After satisfactory orthopedic treatment, a bone graft bed was ground between the spinous process and the lamina of the thoracolumbar vertebra. One cage was placed in each of the L3/4, L4/5, and L5/S1 intervertebral spaces, and autologous fragmented bone was mixed with allogeneic bone and implanted. Three silicone drainage tubes were placed behind the lamina at the deep part of the incision and Layer-by-layer suture. The operation lasted for 345 minutes, and the blood loss was 2500 ml, with ambulation in postoperative 2 weeks.

Review 3 months postoperatively, the internal fixation and fusion were good. Follow-up after one year revealed the shoulder imbalance without pseudoarthrosis, loss of correction, and neurological complications (Figure 1) (Table 1, 2).

DISCUSSION

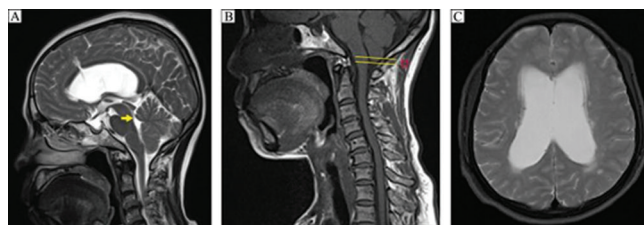
As we know, this may be the third formally reported case in which two or more consecutive physiological curvatures with reverse arch following Safdarian et al.⁷ reported a case of arthrogryposis multiplex congenital with scoliosis and Wang et al.⁸ reported a case of adult degenerative scoliosis. The difference from the previous two case reports is that our case global sagittal sequence was balanced and combined with CSF circulation disease (Figure 3). This poses a great challenge for our orthotic therapy.⁹

The preoperative cranial MRI revealed a slight aqueductal stenosis (Figure 3) and a score of 76 on the Wechsler Adult Intelligence Scale, giving us reason to think that the patient was suffering from LOVA. Xie et al.¹⁰ found that the coiled-coil domain containing 57 (ccdc57) mutation impaired the formation and motor function of ependymal cilia and then affected urotensin signal pathway by interfering with the transmission of adrenaline, thus regulating the development of zebrafish body axis. Epinephrine deficiency can affect the circulatory system, slowing down the heart rate. This is sufficient to explain the ECG manifestation of sinus bradycardia at the time of our patient's admission.

Prior to the onset of low back pain, there was no discomfort due to thoracic lordosis, echocardiography and pulmonary function at the time of admission showed no obvious cardiorespiratory abnormalities. Considering that our patient has been undergoing ventriculoperitoneal shunt for 8 years, Kirksey et al.¹¹ reported the fatal cerebral swelling associated with scoliosis surgery in two patients with neuromuscular scoliosis and congenital hydrocephalus. The drainage tube has poor mobility due to scar hyperplasia and severe surrounding inter-tissue adhesion.¹² If thoracic kyphosis is corrected, the drainage tube may cause shunt malfunction due to thoracic shape changes, resulting in brain swelling or paraplegia.¹³ Therefore, we adopted the treatment principle of degenerative scoliosis to improve her lower back pain, restore the normal physiological curvature of the lumbar, and improve her quality of life. Therefore PSO was used to restore lumbar lordosis because we decided to recover to $10^{\circ} \leq \text{PI-LL} \leq 20^{\circ}$ after operation, considering the patient is older.¹⁴ There may be a risk of spinal dural tear during lumbosacral decompression. In order to avoid the occurrence of cerebellar tonsillar hernia, considering the abnormal position of her cerebellar tonsillar, It is immediately sutured or a dural patch is used to maintain a certain intradural pressure when the tear occurs.

There are some limitations in the diagnosis and treatment of this case. This patient experienced shock due to massive intraoperative bleeding, which was compounded by abnormal CSF flow that interfered with adrenaline transmission. A research report indicates that a reduction in thoracic kyphosis is associated with increased blood loss, offering a possible explanation for the excessive bleeding encountered during the operation.¹⁵ Unfortunately, this patient and family refused to perform genetic testing for economic reasons. Because only the physiological curvature of the lumbar spine has been

Figure 3. The preoperative MRI of brain and cervical spine. Aqueductal stenosis (A), cerebellar tonsil position is critical abnormality, D=3.9 mm (B), lateral ventricular dilatation on axial T2WI-FS (C)



restored, it was found that the bilateral shoulder height was unequal in three months after surgery, and it was found that the shoulder imbalance was aggravated and the left shoulder was significantly increased in one year after surgery (Figure 1).¹⁶ In addition to the fact that myogenic diseases can cause muscle tension imbalance in the muscles around the shoulders, our analysis may be the overcorrection of lumbar scoliosis, and lower instrumented vertebra (LIV) was selected in S1 to cause the lumbar spine to lose the compensatory ability. However, our patient had osteoporosis and L5/S1 spinal canal stenosis, Bai et al.¹⁷ reported long segment fusion can relieve low back pain better and improve walking ability when PI-LL is mismatched, so prolonged internal fixation to S1 was inevitable. At the same time, the post-operation compensatory ability of the thoracic spine in the coronal plane is overestimated. The reasons mentioned above caused the angle mismatch between the thoracic curve and the lumbar curve cobb angle in one year after the operation, in which the thoracic curve was significantly greater than the lumbar curve, resulting in shoulder imbalance.

CONCLUSION

In the future, we should carefully observe for any signs of cerebral ventricular dilatation on MRI prior to complex spinal deformity surgery. When severe thoracic lordosis is present along with lumbar scoliosis, excessive correction of the lumbar scoliosis should be avoided to prevent shoulder imbalance.

CONFLICT OF INTEREST

The authors have no potential conflicts of interest to report relevant to this article.

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This study did not receive any funding in any form.

AUTHOR CONTRIBUTIONS

JW and SY designed the study, JW and ZZ collected the data, SY and ZZ analyzed the data, JW and SY prepared the manuscript. All authors read and approved the final manuscript.

ETHICAL COMPLIANCE

The ethics committee of Affiliated Hospital of Jining Medical University approved this study. Signed written informed consent were obtained from the patients and/or guardians.

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