Treatment of Severe Congenital Scoliosis with Type II Respiratory Failure

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ABSTRACT

We report a case of a 14-year girl with severe congenital scoliosis (CS) and type II respiratory failure (RF), who underwent preoperative halo-gravity traction in combination with intraoperative orthopaedic surgery on the spine based on the results of physical examination, pulmonary function tests (PFTs), computed tomography (CT), and blood gas analysis. The patient's coronal and sagittal Cobb's angles changed from 100° to 45° and 40°, respectively, and RF changed from type II to type I after treatment. Scoliosis was corrected well, and RF was improved. During follow-up for nearly 3 years, no loss of deformity correction and no serious complications occurred, and the patient showed a good clinical outcome and balanced spine.

Key Words: *Congenital scoliosis, Halo-gravity traction, Respiratory failure.*

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INTRODUCTION

Scoliosis is a three-dimensional structural deformity of the spine with sagittal and coronal deformities and vertebral rotation. The Scoliosis Research Society (SRS) defines scoliosis as curvature of the spine with Cobb's angle ≥10°.[1] Congenital scoliosis (CS) is caused by a structural spinal defect and occurs in about 1 in 1000 live births; the incidence rate is second only to idiopathic scoliosis (IS).[2]

Previous studies indicated that 30–60% of CS cases have accompanying congenital deformities that may involve the respiratory, cardiovascular, nervous, musculoskeletal, urogenital, and other systems, especially the respiratory, urogenital, and cardiovascular systems, which have a common embryological origin in the mesoderm.[3] Thus, respiratory damage, including respiratory failure (RF), is a well-recognised complication of severe congenital spinal deformity.[4] RF is divided into types I and II, the former only showing a reduction of partial pressure of arterial oxygen (PaO_2), while the latter is characterised by PaO_2 <60 mmHg and $PaCO_2$ >50 mmHg. Untreated patients can suffer from acute RF due to the gradual reduction of respiratory volume, and the incidence and intraoperative and postoperative mortality rates are high.[5]

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Although previous studies showed that preoperative halo--gravity traction is effective in treating severe CS,[6] treatment of patients with CS and type II RF was reported rarely. Standard management plans are not applicable in such patients, and multi-stage procedures may be required to minimise risks and optimise results.

Herein, we report an adolescent CS patient with severe spinal curvature and RF. The patient was treated successfully with preoperative halo-gravity traction and orthopaedic surgery on the spine. We present the results of almost 3 years of clinical, radiographic, and pulmonary function follow-up.

CASE REPORT

A 14-year child with dyspnea was admitted to the hospital on August 19, 2016. The patient had a known history of untreated scoliosis, progressive worsening of her pulmonary failure with acute dyspnea and palpitations exacerbated by a recent upper respiratory tract infection.

She was diagnosed with CS, pulmonary infection, type II RF, extremely severe restrictive pulmonary ventilation dysfunction, and respiratory acidosis. The results of pulmonary function tests (PFTs) indicated extremely serious restrictive pulmonary ventilation dysfunction (FVC 18.8%, FEV_1 17.7%) and pulmonary reserve dysfunction with maximal voluntary ventilation (MVV) of 51.3%. Blood gas analysis revealed type II RF (PaO_2 57 mmHg, $PaCO_2$ 108 mmHg) and acidosis (pH 7.21). On physical examination, she weighed 31 kg and was 126 cm tall. She had a slightly high right shoulder, a significant right thoracic rib hump, mild pelvic obliquity, normal limb development (Figure 1A), and an obvious rib hump deformity with an angle of 35° (Figure 1B).

Radiographically, she had a Cobb's angle of 100° in both the coronal and sagittal planes (Figure 1C). Three-dimensional CT of the spine (Figure 1D) suggested severe scoliosis, obvious compression of the lung leading to thoracic insufficiency syndrome (TIS) (Figure 1E), and pulmonary infection (Figure 1F). The upper thoracic segment was bent to the left with thoracic (T) 1 and T2 as the centre, and the lower thoracic segment was bent to the right with T9-T10 as the centre. The patient's growth and development were slow, *e.g.*, Risser 2, closure of trigonal cartilage, and menarche were delayed by half a year.

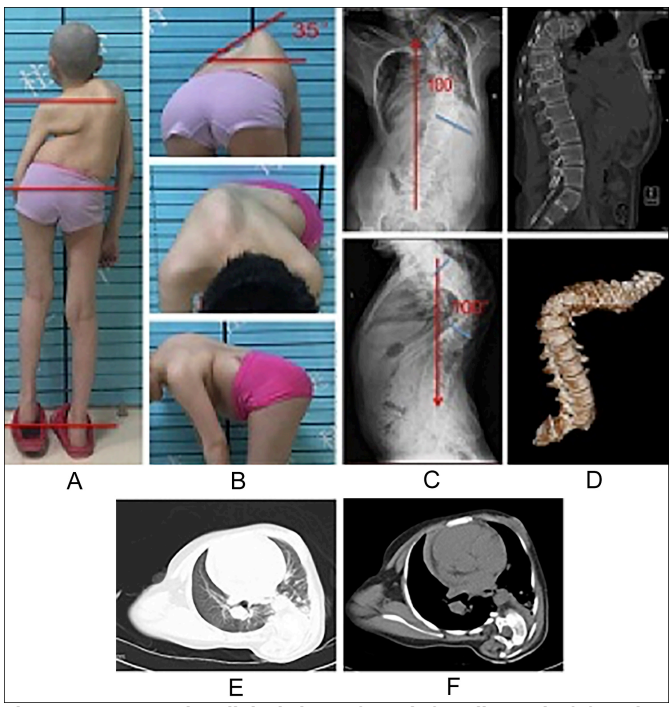

Figure 1: Preoperative clinical photos (A and B), radiographs (C), and CT photos (D, E, and F).

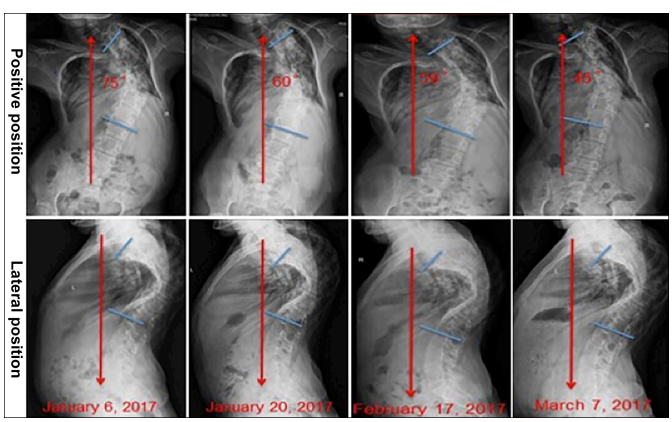

Figure 2: CT images using halo-gravity traction. During the traction using the halo-gravity traction, the coronal (above) and sagittal (below) Cobb's angles changed from 100° to 45° and 40°, respectively. The time was from January 6, 2017 to March 7, 2017.

The patient was first placed in halo-skull–pelvis traction for treatment because of her severe spinal deformity and dyspnea. After 3 months of halo-gravity traction, the symptoms of dyspnea were slightly relieved, with PaO_2 112 mmHg, $PaCO_2$ 41 mmHg, and pH 7.39 on blood gas analysis. In addition, the coronal and sagittal Cobb's angles changed from 100° to 45° and 40°, respectively (Figure 2), and her condition was improving. The results of PFTs, *i.e.*, normal pulmonary reserve function (MVV: 88.2%), slightly decreased diffusion function, and normal airway resistance showed that the patient was gradually recovering. However, lung volume measurement indicated that the total lung volume had decreased and the residual volume increased, with a residual-to-total ratio of 58.17%. Meanwhile, the patient showed extremely serious restrictive pulmonary ventilation dysfunction (FVC 32.8%, FEV_1 31.8%). Her weight was 40 kg, and her height was 138 cm, approximately 12 cm taller than at her initial measurement.

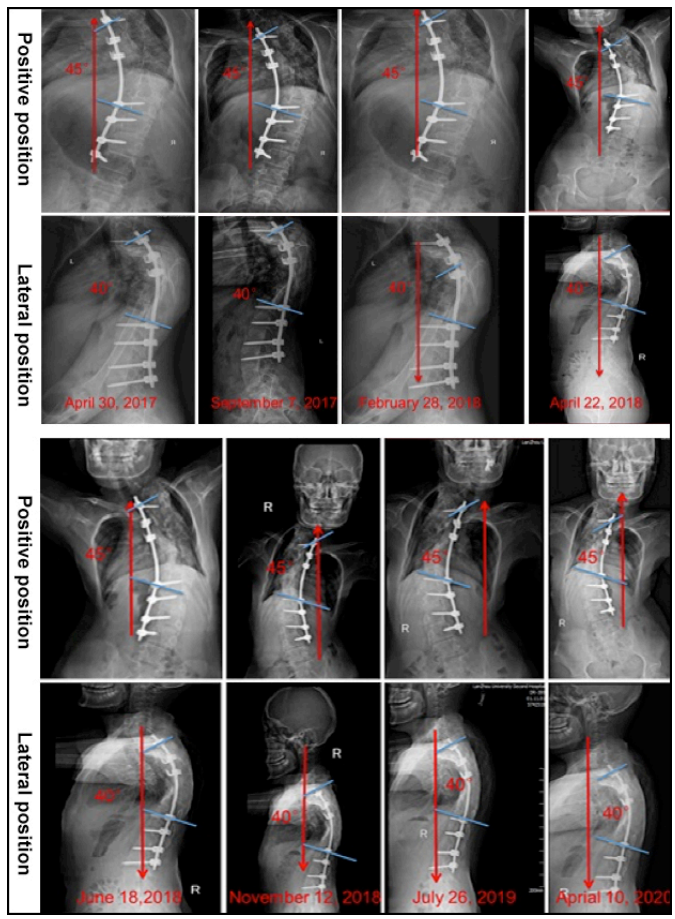

Figure 3: CT images of the spinal coronal (above) and sagittal (below) plane were followed up after the operation using posterior scoliosis correction from April 30, 2017 to April 10, 2020. The Cobb's angle of coronal curvature was 45° and the sagittal plane was 40°.

The patient underwent spinal orthopaedic surgery on April 25, 2017. First, an incision was made along the midline from the centre of T2 to L3 spinous process after general anaesthesia with the patient prone. On the concave side of the vertebrae, five pedicle screws were placed in T2, T12, L1, L2, and L3, and two lamina hooks were placed in T3 and T4, pre-bending and placing the connecting rod, and completing spinal orthopaedic surgery. After anaesthesia and resuscitation, she was transferred to ICU for further observation. Finally, surgical restraint protection was provided after the operation. In the latest follow-

up on April 10, 2020, blood gas analysis showed PaO_2 of 67.3 mmHg, $PaCO_2$ of 44.1 mmHg, and pH of 7.36. PFTs showed that respiratory function had gradually declined compared to before surgery due to a lack of professional home care, with reduced pulmonary reserve function (MVV: 36.0%) and extremely serious restrictive pulmonary ventilation dysfunction (FVC 33.4%, FEV_1 34.0%). In the latest follow-up on April 10, 2020, blood gas analysis showed that PaO_2 was 67.3 mmHg, $PaCO_2$ was 44.1 mmHg, and pH was 7.36. CT indicated that the spinal orthopaedic operation was successful. After nearly 3 years of follow-up, no deformity correction loss and no serious complications were observed. She demonstrated a good clinical outcome and a balanced spine (Figure 3). Professional nurses tended to her and took care of her daily activities during the traction stage and postoperatively.

DISCUSSION

Severe CS results in unilateral or total thorax volume decrease and developmental obstruction, giving rise to TIS. It is very important to the diagnosis and prognosis of TIS that the limitations of the chest and lungs are accurately evaluated. For example, the congenital short-rib syndrome often leads to a fatal lateral growth disorder of the chest (such as Jeune asphyxiating thoracic dystrophy), and congenital shortening of the thoracic spine can lead to overall shortening of the chest (such as Jarcho–Levin syndrome), with a mortality rate of 33%.[7] In early life, a well-developed deformed thorax may not support the size of lungs needed in adulthood (chest "dysplasia"). Therefore, it is necessary to perform a physical examination, CT, PFTs, and other tests to evaluate thoracic development in such patients. In this case, the patient had breathing difficulty at the age of 14 years. After further examination, she was found to have severe CS, TIS, type II RF, and extremely severe restrictive pulmonary ventilation dysfunction. A comprehensive analysis of the patient's status was required, with a formulation of a suitable treatment plan involving preoperative traction in combination with spinal orthopaedic internal fixation.

Congenital spinal deformities sometimes present as rigid, severe curves, which may be impossible to correct using standard instrumentation techniques. The safety of traction in the treatment of congenital spinal deformities, especially in cases involving preexisting neurological deficits, has been questioned by MacEwen *et al*.,[8] who discussed the risk of traction-induced congenital paraplegia. In recent years, traction has been widely used in severe deformities, including congenital deformities.[6,9] Sink *et al*.[9] reported the application of halo-gravity traction, which was then elaborated by Rinella *et al*.[6] Neither of these studies included cases of permanent neurological deficits. Halo-gravity traction allows the patient to gradually increase the weight of the ring. This technique allows partial curve correction and improves respiratory function before surgery. Mehlman *et al*.[10] used halo-femoral traction in 24 patients, with X-ray images showing an average preoperative Cobb's angle of 95° and post-traction angle of 44°. The final curve correction rate was 71%, and there were no permanent neurological deficits. In the present case, the patient underwent halo-skull-pelvis traction and halo-gravity traction, which was instrumental in improving lung function and correcting the curve deformity. RF changed from type II to type I, the coronal Cobb's angle changed from 100° to 45°, and the curve correction was 55%.

The challenges, in this case, included severe and rigid CS with poorly managed type II RF. During the course of treatment, several factors played important roles in the correction of CS. First, minimally invasive surgery, a short surgical time, and administration of anaesthesia were beneficial to the compromised pulmonary profile in this case. Preoperative halo-gravity traction improved not only the flexibility of the spine and chest but also the lung function and allowed continuous monitoring of the patient's nervous system function. In addition, the required degree of correction was obtained at the end of the traction procedure, which was followed by *in-situ* fusion and total pedicle screw fixation. Finally, professional nurses provided guidance and took care of daily activities for the patient during the stage of traction and postoperatively.

PATIENT'S CONSENT:
Written consent was taken from the patient.

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
YM, YW, XK: Halo-skull–pelvis traction and orthopaedic surgery.
XD, FY: Data collection and follow-up.
YM, ZL, XK: Literature search and final draft of the case report.
All the authors have approved the final version of the manuscript to be published.

REFERENCES

1. Wang Y, Yang F, Wang D, Zhao H, Zhanjun Ma, Peifen Ma, *et al*. Correlation analysis between the pulmonary function test and the radiological parameters of the main right thoracic curve in adolescent idiopathic scoliosis. *J Orthop Surg Res* 2019;**14(1)**:443. doi: 10.1186/s13018-019-1451-z.
2. Hensinger RN. Congenital scoliosis: Etiology and associations. *Spine* 2009; **34(17)**:1745-50. doi: 10.1097/BRS.0b013e3181abf69e.
3. Dos SJA, de Miranda DM, Simoes ESA. Congenital anomalies of the kidney and urinary tract: An embryogenetic review. *Birth Defects Res C Embryo Today* 2014; **102(4)**:374-81. doi: 10.1002/bdrc.21084.
4. Gao Z, Wang Z, Liu J, Niu B, Yang W, Wang Y, *et al*. Evaluation of renal function in children with congenital scoliosis and congenital anomalies of the kidney and urinary tract. *Med Sci Monit* 2018; **24**:4667-78. doi: 10.12659/MSM.908839.
5. Kanagaraju V, Chhabra HS, Srivastava A, Mahajan R, Kaul R, Bhatia P, *et al*. A case of severe and rigid congenital thoracolumbar lordoscoliosis with diastematomyelia presenting with type 2 respiratory failure: Managed by staged correction with controlled axial traction. *Eur Spine J* 2016; **25(10)**:3034-41. doi: 10.1007/s00586-014-3624-0.

6. Zhang HQ, Deng A, Tang MX, Liu SH, Wang YX, Gao QL. Posterior-only surgical correction with heavy halo-femoral traction for the treatment of rigid congenital scoliosis associated with split cord malformation. *BMC Musculoskeletal Disorders* 2020; **21(1)**:98. doi: 10.1186/s12891-020-3124-9.
7. Ramerez N, Santiago-Cornier A, Arroyo S, Acevedo J. The natural history of spondylothoracic dysplasia. Read at the annual meeting of the pediatric orthopaedic society of North America. 2001; May 2-6, Cancun, Mexico.
8. MacEwen GD, Bunnell WP, Sriram K. Acute neurological complications in the treatment of scoliosis. A report of the scoliosis research society. *J Bone Joint Surg Am* 1975; **57(3)**:404-8.
9. Sink EL, Karol LA, Sanders J, Birch JG, Johnston CE, Herring JA. Efficacy of perioperative halo-gravity traction in the treatment of severe scoliosis in children. *J Pediatr Orthop* 2001; **21(4)**:519-24.
10. Mehlman CT, Al-Sayyad MJ, Crawford AH. Effectiveness of spinal release and halo-femoral traction in the management of severe spinal deformity. *J Pediatr Orthop* 2004; **24(6)**:667-73. doi: 10.1097/00004694-200411000-00014.