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GRAND ROUNDS

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

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Abstract

Introduction Congenital lordoscoliosis is an uncommon pathology and its management poses formidable challenge especially in the presence of type 2 respiratory failure and intraspinal anomalies. In such patients standard management protocols are not applicable and may require multistage procedure to minimize risk and optimize results.
Case description A 15-year-old girl presented in our hospital emergency services with severe breathing difficulty. She had a severe and rapidly progressing deformity in her back, noted since 6 years of age, associated with severe respiratory distress requiring oxygen and BiPAP support. She was diagnosed to have a severe and rigid congenital right thoracolumbar lordoscoliosis (coronal Cobb's angle: 105° and thoracic lordosis −10°) with type 1 split cord malformation with bony septum extending from T11 to L3. This leads to presentation of restrictive lung disease with type 2 respiratory failure. As her lung condition did not allow for any major procedure, we did a staged procedure rather than executing in a single stage. Controlled axial traction by halogravity was applied initially followed by halo-femoral traction. Four weeks later, this was replaced by halo-pelvic distraction device after a posterior release procedure with asymmetric pedicle subtraction osteotomies at T7 and T10. Halo-pelvic distraction continued for 4 more weeks to optimize and correct the deformity. Subsequently definitive posterior stabilization and fusion was done. The detrimental effect of diastematomyelia resection in such cases is clearly evident from literature, so it was left unresected. A good scoliotic correction with improved respiratory function was achieved. Three years follow-up showed no loss of deformity correction, no evidence of pseudarthrosis and a good clinical outcome with reasonably balanced spine.
Conclusion The management of severe and rigid congenital lordoscoliotic deformities with intraspinal anomalies is challenging. Progressive reduction in respiratory volume in untreated cases can lead to acute respiratory failure. Such patients have a high rate of intraoperative and postoperative morbidity and mortality. Hence a staged procedure is recommended. Initially a less invasive procedure like halo traction helps to improve their respiratory function with simultaneous correction of the deformity, while allowing for monitoring of neurological deficit.

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Subsequently spinal osteotomies and combined halo traction helps further improve the correction, following which definitive instrumented fusion can be done.

Keywords Lordoscoliosis · Diastematomyelia · Split cord malformation · Halo traction · Pseudarthrosis · Controlled axial traction

Case presentation

A 15-year-old girl reported in the emergency department with respiratory distress on BiPAP support on a wheel chair associated with severe scoliotic deformity. The deformity was noted at 6 years of age, which gradually progressed over the years in the absence of any treatment. However, in the last 2 years there was a rapid growth noted in the curve, which coincided with her growth spurt. This rapid growth of thoracic curve led to severe reduction in lung volume. Consequently she complained of severe breathing difficulty, which became worse 3 months before presentation. This started initially with exertional dyspnoea after walking 100 m and at time of presentation had progressed to dyspnea even at rest. Two weeks earlier she had an episode of an acute respiratory distress and was admitted at a small peripheral center where she was started on BiPAP. She was discharged home a week later with BiPAP and was counseled about the dismal prognosis and progressive nature of the condition. She was sent home for palliative care. At the time of presentation in our hospital, she was on BiPAP support and functionally restricted to bed. On examination her right shoulder and left anterior superior iliac spine were at higher levels compared to contralateral side, with a prominent rib hump on right side. She had significant coronal and sagittal imbalances (coronal imbalance of 5 cm and a negative sagittal balance of 7 cm and a fixed pelvic obliquity of 40°). A tuft of hair was noted on her mid back region suggesting an underlying congenital cord anomaly. Her neurological assessment was intact. She was diagnosed to have a congenital right thoracolumbar lordoscoliosis (McMaster type III—mixed or unclassifiable vertebral anomalies) with curve extending from T1 to L4 with apex at T8, coronal Cobb's angle measuring 105°, thoracic lordosis −10° with diastematomyelia (type 1 split cord malformation, SCM1) extending from T11 to L3 without any evidence of cord tethering and skeletal maturity at Risser 4 (Figs. 1, 2). The curve was also rigid as the flexibility was less than 10 % on bending films (Fig. 1). X-ray and chest CT depicted reduced pulmonary space on both sides with severe reduction on right side (Fig. 2). Her oxygen saturation was 70 % on room air and 88 % with 5 l of oxygen. Her pulmonary function test and arterial blood gas analysis (FVC 23 %, FEV_1 22 %, VC 26 %, $PaCO_2$ 52 mmHg, RR 37/min, O_2 saturation 88 %) were indicative of an underlying severe restrictive lung disease (FVC, forced vital capacity; FEV_1, forced expiratory volume in 1 s; VC, vital capacity; RR, respiratory rate). Also her nutritional status was suboptimal with body mass index of 16 %. The challenge was to correct the deformity safely to preserve and improve lung function and survival.

Historical review of the condition, epidemiology, diagnosis, pathology

Respiratory compromise is a well-recognized complication of any congenital spinal deformity. Congenital scoliosis has its origin very early in embryonic life and may be associated with pulmonary bronchial and alveolar hypoplasia with rib anomalies that contribute to the reduction of VC [1]. The decompensated lung function in congenital scoliosis is further exacerbated by progressive thoracic scoliosis and in particular lordoscoliosis [2]. Nash and Nevins [2] demonstrated that patients with scoliosis tended to have a decreased ratio of anteroposterior to lateral thoracic diameter and this decreased ratio correlated with pulmonary dysfunction. Winter et al. [3] subsequently observed lordoscoliosis to cause more respiratory compromise than kyphoscoliosis. Furthermore, he correlated the improvement in pulmonary function after surgery directly to the amount of lordosis corrected.

A thoracic kyphosis of less than 20° is considered evidence of a relative lordosis by the Scoliosis Research Society [4]. The natural history of congenital lordosis, especially in the thoracic spine, is not encouraging, since progressive deterioration in pulmonary function is inevitable and early death can result from respiratory insufficiency and cor pulmonale [4]. This makes an early surgical intervention mandatory [5]. Even difficulty in swallowing of solid food can be a problem due to esophageal compression [4].

Significant pulmonary compromise adds challenge to already daunting surgical procedures. The initial series of patients with thoracic lordosis and respiratory failure as managed by Winter et al. [6] and Bradford et al. [5] were disappointing and suggested for better surgical techniques. Payo et al. [7] reported 58 % overall complication rate of single stage surgical correction in pediatric spinal deformities with forced vital capacity (FVC) <40 %.

Rationale for treatment and evidence-based literature

The aim of surgery in these patients is to halt the progress of a disabling deformity, to diminish the size of the curve and to restore trunk balance while improving the patient's

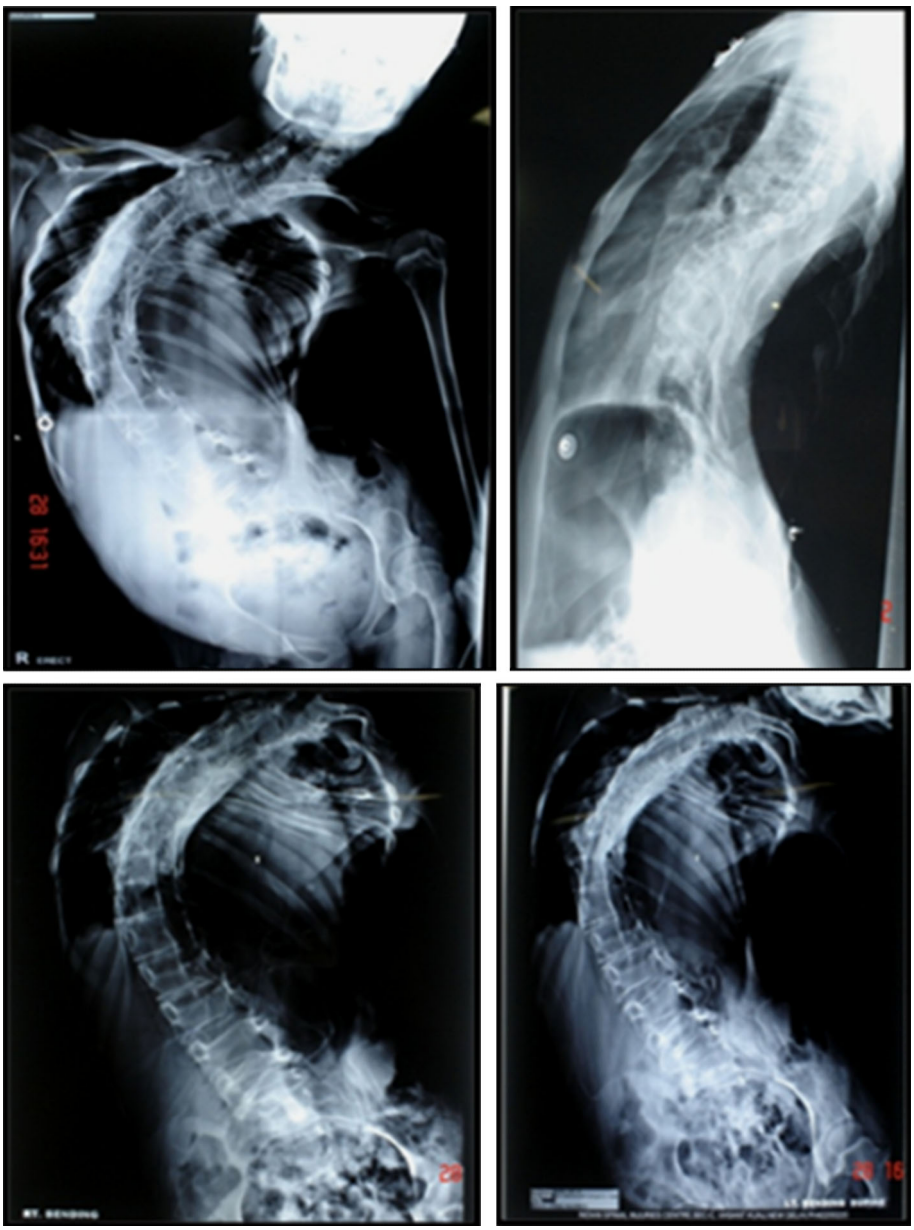

Fig. 1 AP and lateral standing X-rays showing coronal Cobb's angle of 105°, sagittal thoracic Cobb's angle of −10° and bending films showing flexibility <10 %

quality of life, providing cardiopulmonary stability and avoiding long-term sequelae from restrictive lung disease, painful degeneration and curve decompensation [8, 9].

Severe and rigid spinal deformities are made supple with anterior and or posterior release with or without osteotomies and easily maneuvered intraoperatively by pedicle screw based instrumentation and with the advent of newer techniques with neurological safety under intraoperative monitoring [10]. Though the technology has advanced, not all cases be managed in a single stage and management has to be tailored to individual cases based on the etiology, curve character and associated anomalies and complications.

The spinal cord has poor tolerance to acute traction in severe and rigid scoliosis especially when associated with intraspinal anomalies. Also there is a high risk of injury to the spinal cord due to forcible intraoperative maneuvering during single stage correction. It would be difficult to manage such cases satisfactorily by a single stage surgical procedure in consideration of the neurological safety and respiratory compromise [11]. Dutoit [12] has reported 5.3 % incidence of permanent neurological injury and 46 % transient neurological deficit in single stage deformity surgeries and this has further increased with associated intraspinal anomalies. Staged surgery has been used in the treatment of severe, rigid scoliosis to prevent neurological compromise [13–15]. Also a staged, less invasive procedure like halo axial traction helps to improve the lung function, simultaneously aids in correction of deformity to some extent. Since

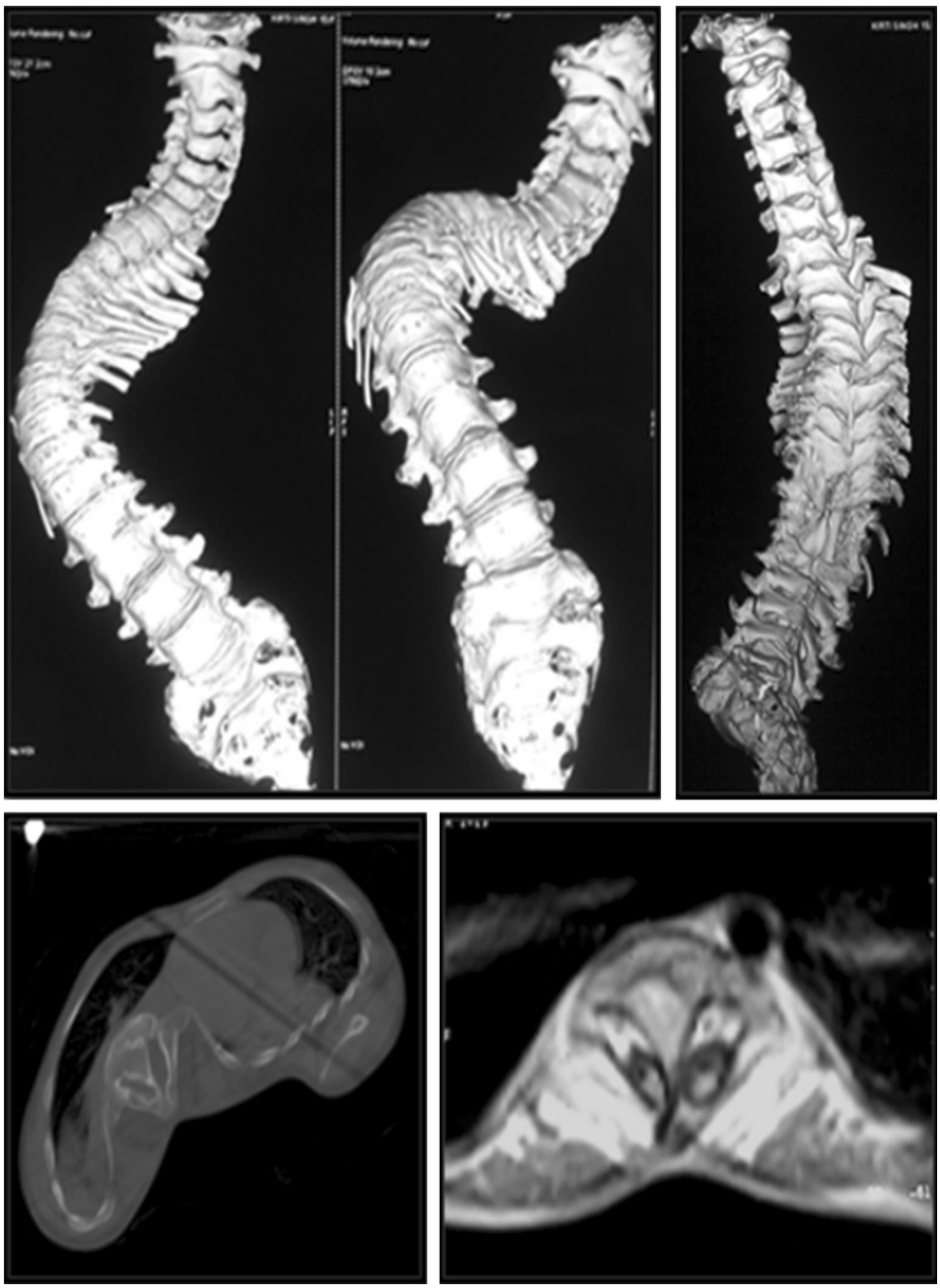

Fig. 2 CT and MRI showing the severity and the type of deformity and also the intraspinal anomaly (SCM1). Further, the AP diameter of the chest is reduced compared to the transverse diameter on axial CT

the traction is done awake, neurology can be closely monitored [13]. This patient also had diastematomyelia (SCM1) extending from T11 to L3 without any evidence of cord tethering (Fig. 2). There is a controversy over the management of spinal deformity associated with diastematomyelia among spine surgeons over years. The concept of deformity correction without excision of the spur is not new and has also been supported by literature [16, 17]. Wang et al. [18] also support this view especially in type 1 SCM. Moreover, significant neurological damage has been recorded during the prophylactic removal of bony spurs in setting of scoliosis without neurological deficit [17]. The impact of traction on pulmonary function deserves further comment. In severe scoliosis and kyphoscoliosis, poor pulmonary function is in part due to the restrictive spinal deformity. With the preoperative application of halo-gravity traction, especially in the setting of severe and rigid scoliosis, pulmonary function has been demonstrated to improve significantly [11]. The safety and efficacy of halo-gravity traction in this setting has been described in various reports. They have also reported positive impact of traction on sagittal and coronal profile [19–21].

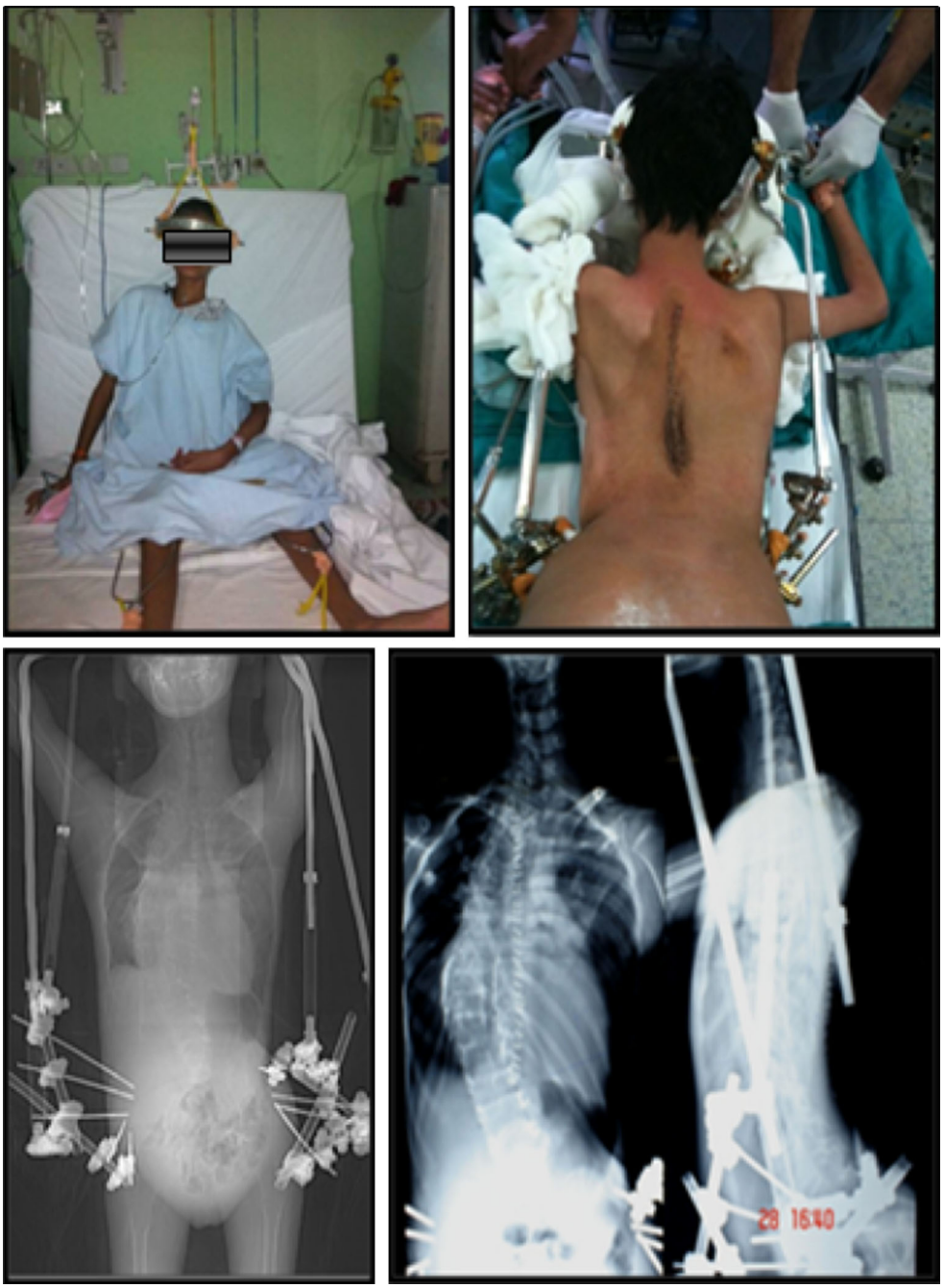

Fig. 3 Halo-gravity and halo-femoral traction followed by posterior release and periapical osteotomies with halo-pelvic distraction. Note the amount of correction at the end of 4 weeks of halo-pelvic distraction

Procedure

Though single stage surgery was successful in many of our cases with severe scoliosis, we have encountered neurological complication in two cases (1 with neurofibromatosis and the other with congenital kyphoscoliosis) during intraoperative maneuvering. Also considering her progressing respiratory failure with poor pulmonary reserve and intraspinal anomaly, we have planned to stage the surgery and first focus on improving her respiratory status by halo traction. After informing consent, halo-gravity traction with 5 kg weight was started and gradually increased at the rate of 1 kg per day. At the end of 2 weeks, we added a halo-femoral traction to facilitate further correction and continued for 2 more weeks (Fig. 3). Her respiratory function improved clinically as she was comfortable with minimal oxygen support without BiPAP, and the curve measured 85° at the end of

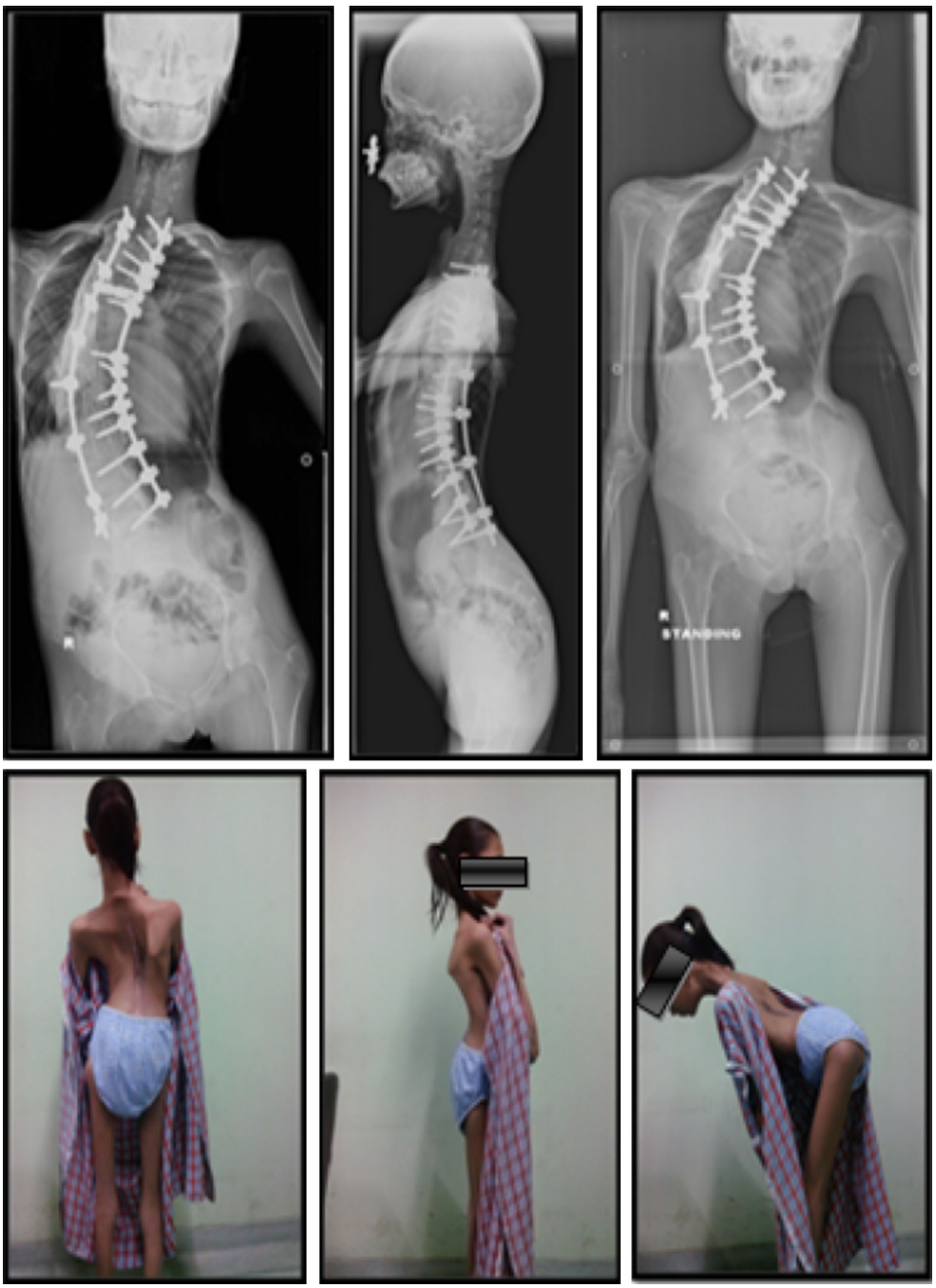

Fig. 4 Postoperative correction of coronal Cobb's angle to 60° and sagittal thoracic Cobb's angle to neutral (0°) and maintained at final follow-up at 3 years with improved clinical outcome and quality of life

4 weeks. Meanwhile these 4 weeks were also helpful to optimize her general condition. As her respiratory function improved clinically she was taken to operating room wherein halo-pelvic apparatus was applied and multiple posterior releases by facetectomies and asymmetric pedicle subtraction osteotomies at T7 and T10 were done (Fig. 3). The bony spur of diastematomyelia was left unresected. We did not contemplate any procedure by anterior approach to avoid further respiratory embarrassment to her already decompensated lung. Postoperatively awake, axial distraction on halo-pelvic apparatus was started at the rate of 1 turn per day and was continued for a week in hospital. This was uneventful and consequently she was discharged home on the advice to continue the distraction at home for 3 more weeks. She was warned about the red flags and asked to report urgently to our

hospital in case of any neurological deterioration. At the end of 4 weeks she reported with an intact neurology and curve corrected to 60°. We accepted the correction obtained and did a posterior pedicle screw instrumentation from T1 to L4 with an in situ fusion with bone grafts and rhBMP-2 (Fig. 4). Intraoperative somatosensory evoked potential (SSEP) monitoring combined with wake-up test was done during both the operative procedures. Postoperative periods were uneventful. She was discharged 1 week later and reported at 3 months, 6 months, 1 year, 2 years, and recently at 3 years (Fig. 4).

The challenges in this case include severe and rigid congenital scoliosis on the background of poorly managed type II respiratory failure, which is further complicated by the presence of thoracic lordosis and diastematomyelia. We would attribute the success behind the reasonable correction of the deformity without intraoperative and postoperative morbidity or mortality to several factors like staging and less invasive procedures, short surgical time and administration of anesthesia favorable to her compromised pulmonary profile. Preoperative controlled, awake axial traction with halo-gravity and halo-femoral traction under local anesthesia followed by posterior release with periapical osteotomies and halo-pelvic distraction, under closed neurological supervision, averted any significant neurological deficits in the presence of diastematomyelia. The desired degree of correction was obtained at the end of the traction program and was followed by an in situ fusion and all pedicle screw instrumented stabilization. Prophylactic excision of diastematomyelia especially in asymptomatic patients is debatable and fraught with neurological deterioration as pointed out by various authors [16–18]. The authors felt that simultaneous excision of diastematomyelia would have added significant morbidity, duration, blood loss and increased the chances of neurological deficit in the procedure. This would have complicated the matters further in the presence of compromised pulmonary function, so they were left unresected.

Outcome and follow-up

Her pulmonary function at 3 years postoperative follow-up has improved moderately (FVC 48 % predicted and FEV_1 45 % predicted). Also her physical health and the quality of life were improved, as demonstrated by the absence of respiratory distress on walking and moderate running. A reasonably good curve correction (coronal 105°–60° and sagittal −10°–0°) with coronal and sagittal balance (imbalance less than 1 cm) was achieved without any neurological deficit. At 3 years follow-up she was happy with the correction achieved and with her respiratory functional improvement. Her clinical and radiological coronal and sagittal balances were reasonably good with no signs of pseudarthrosis or implant failure (Fig. 4).

Conflict of interest The authors have no financial disclosures.

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