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GRAND ROUNDS

Selective hemivertebrae resection in a congenital scoliosis patient with multiple hemivertebrae deformities

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Abstract

Purpose To report the surgical experience of selective hemivertebrae resection for a case of congenital scoliosis with multiple hemivertebrae deformities.
Methods A 14-year-old male presented with progressive rib hump, tilted torso and spine deformity was admitted in our department. No abnormalities were detected in neurological examination and the comprehensive imaging study demonstrated congenital scoliosis of multiple hemivertebrae in T5, T10, L1 and L3. Treatment of the patient commenced with a 10-day skin traction therapy prior to the surgery. Selective resection of hemivertebrae in T5 and L1 was performed with segmental fusion from T3 to L2.
Results After surgical procedure, the patient achieved a good coronal and sagittal balance along with a good correction of the curve. 18-month postoperative follow-up showed no evidence of significant loss of correction. No device-related complication such as implant loosening or failure or neurologic complication occurred during the follow-up. Besides, patient's shoulder balance was further improved and coronal balance was maintained in a normal range.
Conclusion Many factors have to be considered in the clinical decision-making of congenital scoliosis with multiple hemivertebrae deformities patients. Much emphasis in this regard is laid on the type and location of the hemivertebrae as well as the patient's age. Selective hemivertebrae resection may be more suitable for such patients.

Keywords Congenital scoliosis · Hemivertebrae · Pedicle screw fixation · Spinal fusion · Hemivertebrae resection

Case presentation

A 14-year-old male presented with progressive rib hump, tilted torso and spine deformity was admitted to the orthopedic department of our hospital. The malformations were found 3 years before and progressed since then, which was confirmed by the radiography of whole spine and unsuccessfully controlled by conservative treatment. The patient denied other congenital abnormalities in the past medical history. Physical examination shows shoulder imbalance, rib hump and tilted torso. There was no abnormality detected in neurological examination.

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Diagnostic imaging

Patient's erect radiography of whole spine demonstrated a thoracic curve with a Cobb angle measurement of 42° and 3.9 cm of coronal imbalance to the left. The left shoulder was 3 cm higher than the right shoulder. Lateral X-ray showed a thoracic kyphosis of 42° and a 2.4 cm positive sagittal balance. The Risser sign was grade I and hemivertebrae deformities were observed in L1 and L3 (Fig. 1). The bending X-ray of spine, computerized tomography (CT) including three-dimensional reconstruction and magnetic resonance imaging (MRI) scans of whole spine were taken. The bending X-ray showed a rigid thoracic curve and no abnormality was observed in spinal cord on MRI. CT demonstrated segmentation failure in C5–6, butterfly vertebrae in T6 and hemivertebrae deformities in T5, T10, L1 and L3 (Fig. 2).

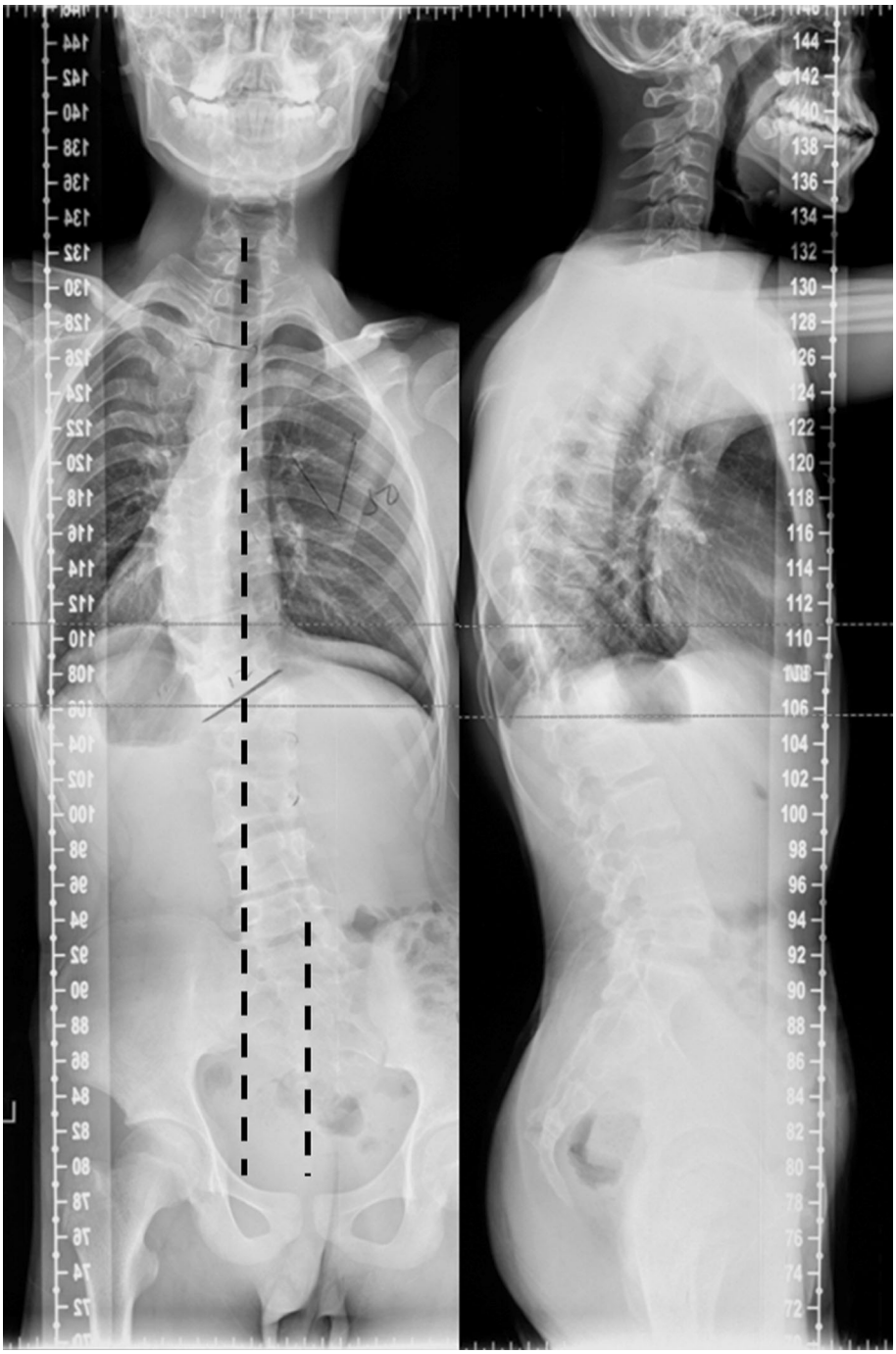

Fig. 1 Preoperative erect radiography of whole spine demonstrated a thoracic curve and hemivertebrae deformities in L1 and L3

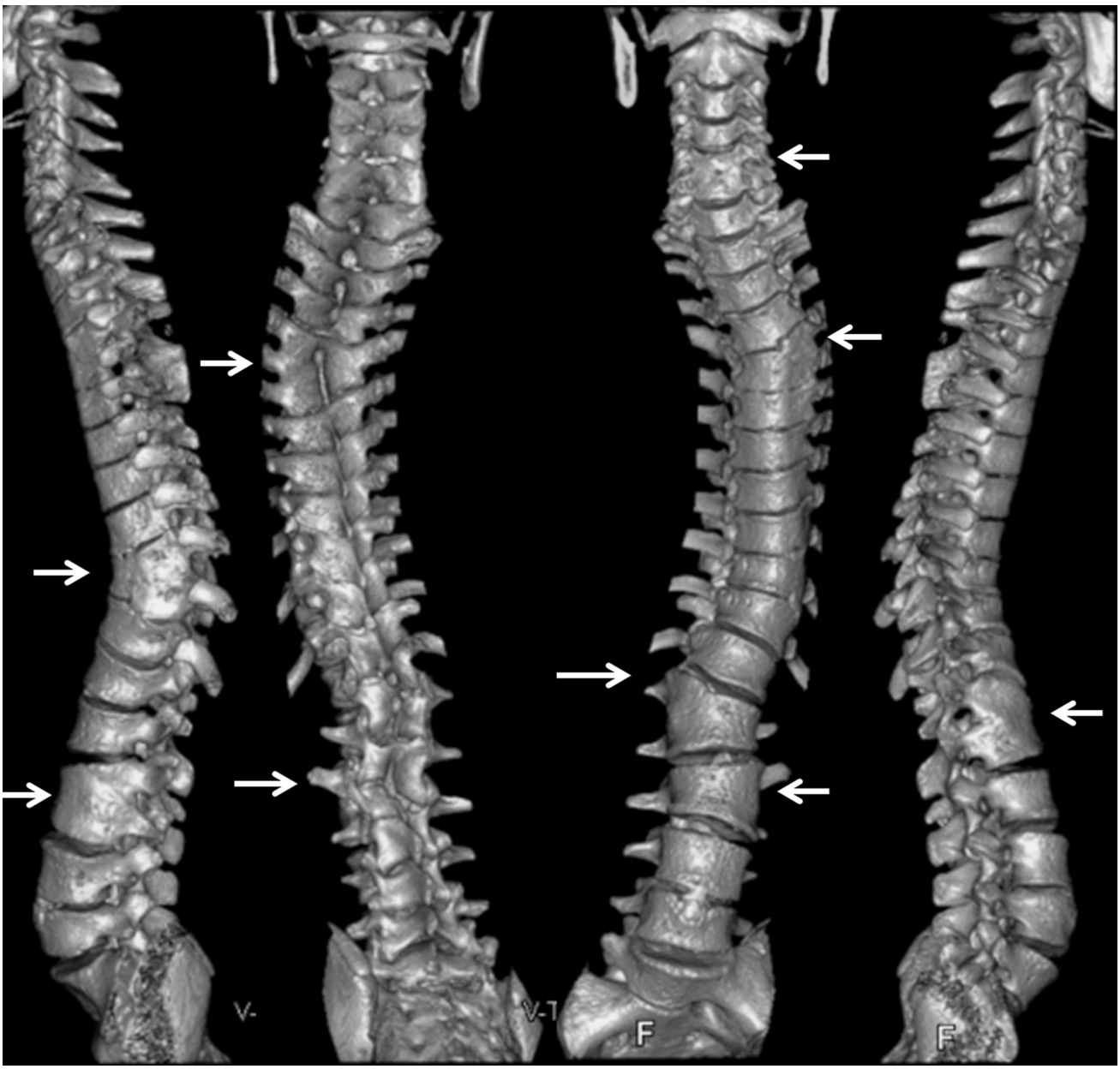

Fig. 2 Preoperative CT scan of whole spine demonstrated segmentation failure and multiple hemivertebrae deformities

Historical review

Congenital scoliosis is one of the common etiologies of scoliosis. It is due to failure of vertebral formation or segmentation causing the asymmetric growth of spine which eventually leads to spinal deformity [1]. Hemivertebrae deformity is the most common cause accounting for about 46% of congenital scoliosis [2]. There have been plenty of studies showing good correction outcome for congenital scoliosis patients treated with one-stage hemivertebrae resection and pedicle screws instrumentation via posterior approach. And the outcomes were maintained during the long-term follow-up [3, 4]. However, unlike patients with single-level hemivertebrae, patients with multiple hemivertebrae deformities need unique surgical strategy to choose which hemivertebrae should be excised or preserved. To the best of our knowledge, little literature was published to describe the treatment of the multiple hemivertebrae deformities in detail. In this case, we report a case of congenital scoliosis caused by multiple hemivertebrae deformities and describe the surgery strategy we performed.

Rationale for treatment and evidence-based literature

In most patients with congenital scoliosis with single-level hemivertebrae, early single hemivertebra resection and short fusion could be performed for a mild flexible curve without structural compensatory curve to preserve more growth potential of spine [3–7]. However, the surgical strategies are totally different when it comes to multiple hemivertebrae. Combination of different distribution and types of hemivertebrae makes the treatment extremely complex. Therefore, it is important to make a rational choice of hemivertebrae resection.

McMaster et al. [8] suggested that the progression of congenital scoliosis depends on four factors: the type of hemivertebrae, location, number and their relationship with each other as well as the age of the patient. Similar to those of progression, the selection of hemivertebrae resection is also based on the above factors. Except for non-segmented or incarcerated type, most of hemivertebrae including semi-segmented and fully segmented have normal growth plates which will more likely lead to progression of deformity. Yet the decision to make also depends on the location of hemivertebrae. Upper thoracic (T1–4) have a great impact on shoulder balance; the lower thoracic and thoracolumbar (T5–L1) hemivertebrae need early intervention since it is easy to cause progression of scoliosis; the lumbosacral segment (L1–S1) may play a role in pelvic balance and coronal balance and need to be removed if necessary [8, 9]. Besides, when the hemivertebra is located in the apical vertebrae or end vertebrae of the curve, it should be resected since the asymmetric stress will cause or deteriorate the scoliosis. Ruf et al. [7, 10] suggested that hemivertebrae should be treated early before the deformity becomes rigid and structural changes occur in

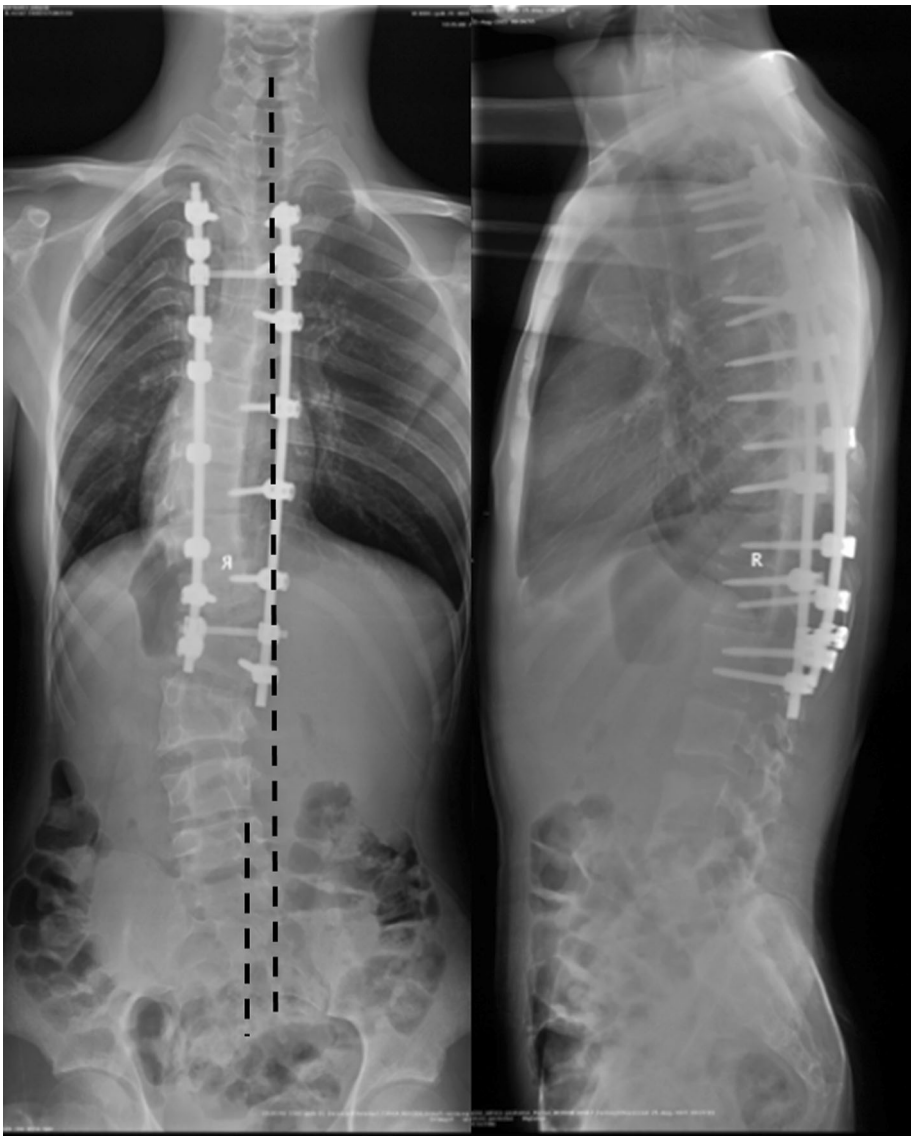

Fig. 3 Postoperative erect radiography of whole spine revealed satisfactory coronal and sagittal balance as well as a well correction of curve

Fig. 4 The photographs taken before discharge showed that torso and shoulder balance improved significantly

the adjacent segments. Early surgery could allow the normal growth in the rest spine via preventing the progression of local deformity and secondary structural curves.

In this case, considering the progression of spine deformity and malalignment of spine as well as patient's requirement of appearance, our main goal was to eliminate

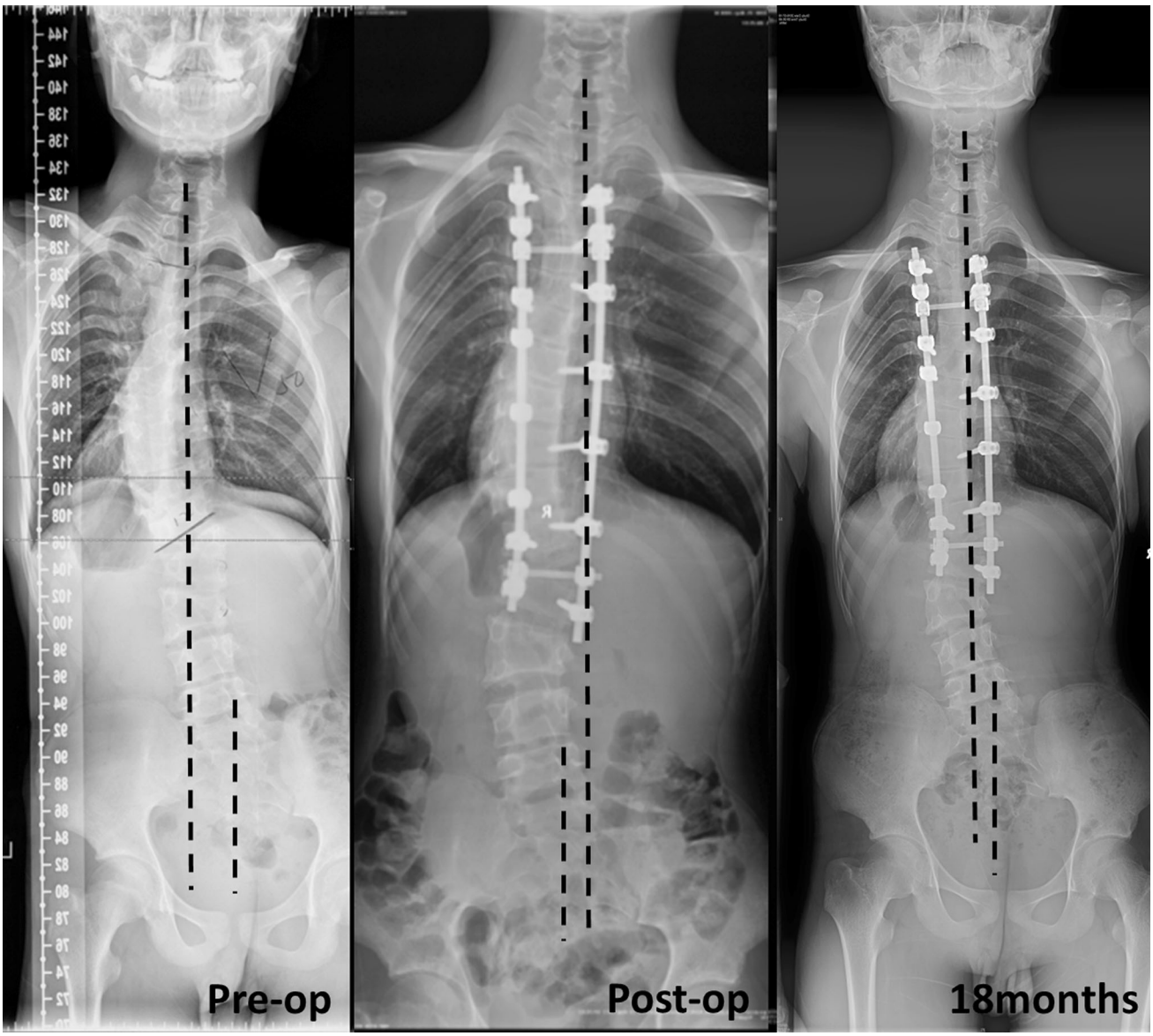

Fig. 5 Erect radiography of whole spine taken 18 months after the operation which showed no significant progression in both thoracic and lumbar curve and coronal and sagittal balance was maintained in a normal range. Meanwhile, patient's shoulder balance was further improved

the main cause of progression, restore spine alignment and correct the scoliosis to some extent. The semi-segmented hemivertebrae in L1 should be first removed since it was located in the end vertebrae of thoracic curve, resulting in an asymmetric stress on T12 which may deteriorate the deformity. This hemivertebrae resection greatly improved the patient's coronal balance with a little benefit to the correction of curve. The location of hemivertebrae in T5 was the apical vertebrae of thoracic curve and its excision decreased the magnitude of scoliosis and also improved shoulder balance to a certain extent. Meanwhile, due to the poor flexibility of thoracic curve and patient's age, we decide to perform long fusion instead of local fusion. Besides, given the age of the patient and the characteristics of the hemivertebrae in T10 and L3, we decided to preserve both of the segments. It was demonstrated on radiography that the direction of lumbar curve (L3–5) was same to the thoracic curve, which means the lumbar curve was not secondary to the thoracic curve. Meanwhile, the poor flexibility of lumbar curve (L3–5) makes it a structural curve which could be a relatively stable base to support the thoracic spine. If we try to excise the hemivertebrae in L3, this balanced condition would be destroyed. Therefore, we preserve the hemivertebrae in L3 and the motion segments from L3 to L5. Also, the location and type of hemivertebrae in T10 determines that it has very little influence on the curve and coronal balance and we leave it intact. The surgical outcome achieved a good coronal balance and a

satisfied correction of the thoracic curve eventually. The erect radiography of whole spine taken at 18-month follow-up post-operation verified our assumption that the lumbar curve provided a stable base and there was no significant progression occurred.

Surgical procedure

The treatment of patient commenced with a 10-day skin traction therapy prior to the surgery. To address the multiple hemivertebrae and progressive spine deformity, selective resection of hemivertebrae in T5 and L1 was performed with pedicle screws fixation from T3 to L2. First, sub-periosteal exposure of the posterior elements was performed from T3 to L2 through a posterior middle-line incision. Pedicle screws were then discontinuously implanted both in convex and concave sides. Then the wedge-shaped resection was performed in hemivertebrae of T5 and L1. After complete resection of the hemivertebrae, two precontoured rods were placed and the residual gaps after resection were slowly closed by posterior compression. The somatosensory evoked potentials (SSEP) monitoring was used during resection and correction and it did not reveal any changes. Meanwhile, wake-up test was used and showed no abnormal after the correction. Decortication of the posterior elements and fusion was then performed from T3 to L2.

Outcome and follow-up

Postoperative erect radiography of whole spine revealed satisfactory coronal and sagittal balance along with a good correction of the curve. The thoracic curve was improved to 30°, while the thoracic kyphosis to 33°. The coronal imbalance was improved to 1.2 cm and sagittal balance was improved to 2.1 cm. Meanwhile, the shoulder imbalance was reduced to 1.5 cm (Fig. 3). Before discharge, the photographs of patient were taken 2 weeks after surgery, which showed that torso balance and shoulder balance improved significantly (Fig. 4). Follow-up at 18-month post-operation did not show a significant loss of correction, with the thoracic curve being further reduced to 25° and thoracic kyphosis increased to 34°. The coronal and sagittal balance was maintained in a normal range of 1.5 and 2.3 cm. Patient's shoulder balance was further improved to 1 cm (Fig. 5). No device-related complication such as implant loosening or failure or neurologic complication occurred during the follow-up.

Conclusion

Many factors have to be considered in the clinical decision-making of congenital scoliosis with multiple hemivertebrae deformities patients. Much emphasis in this regard is laid on the type and location of the hemivertebrae as well as the patient's age. Selective hemivertebrae resection may be more suitable for such patients.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest. No funds were received in support of this work. No relevant financial activities outside the submitted work.

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