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CASE REPORT

Long-term management of congenital lordoscoliosis of the thoracic spine

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Abstract
Purpose The objective of this study is to report the progression of congenital hyperlordoscoliosis in a 2-year-old patient and the use of multiple surgical interventions in the treatment of hyperlordoscoliosis of the thoracic spine.
Methods A 2-year-old patient with thoracic hyperlordosis underwent observation for 1 year. To halt the progression of hyperlordosis, a posterior laminectomy was carried out to remove all the fused segments. Despite surgery, lordosis progressed via spontaneous autofusion with development of scoliosis with unilateral unsegmented bar. At the age of 9 years, the patient underwent posterior osteotomy at the fused segments, which was unsuccessful in the correction of hyperlordosis, but was successful in the correction of scoliosis. At the age of 12 years, the patient complained of mild breathing difficulties resulting from hyperlordosis of the thoracic spine, and underwent posterior multilevel vertebral osteotomy (PMVO) again to correct lordoscoliosis.
Results Follow-up in the 3 years after PMVO showed that correction of the deformity was well maintained, with a good clinical outcome and a well-balanced spine.
Conclusions PMVO is a potential intervention to manage rigid and severe congenital lordoscoliosis of the thoracic spine.

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Keywords Lordoscoliosis · Progression · Spontaneous autofusion · Posterior multilevel vertebral osteotomy

Introduction

Congenital lordoscoliosis is a very rare spinal deformity that is associated with vertebral, rib and intraspinal anomalies, and known to be difficult to manage [1–4]. Weiss and Moramarco [5] recently reported that surgery is unnecessary in treating congenital scoliosis with the application of advanced bracing technology even in face of the challenge of predicting the final outcome for patients with congenital scoliosis. However, correction of lordosis in a patient who has previously had a fused spine poses a surgical challenge. Posterior multilevel vertebral osteotomy (PMVO) has advantages such as, posterior-only procedure, which avoids risk to pulmonary complications and gives satisfactory correction [6]. We report on the management of congenital lordoscoliosis of the thoracic spine using PMVO to correct the deformity in a patient who had undergone previous surgical procedures.

Case report

A 2-year-old boy had undergone several surgical procedures to correct the multiple-joint deformity since the age of 1 year. At the age of 2 years, he visited the spine center for hyperlordosis of the thoracic spine (Fig. 1a). Neurological examination showed no definite abnormalities. Plain radiography and computed tomography (CT) showed hyperlordosis and congenital fusion of all of the thoracic segments (Fig. 2a–c). Because the thoracic lordosis had progressed, all fused segments were removed (Fig. 3) by

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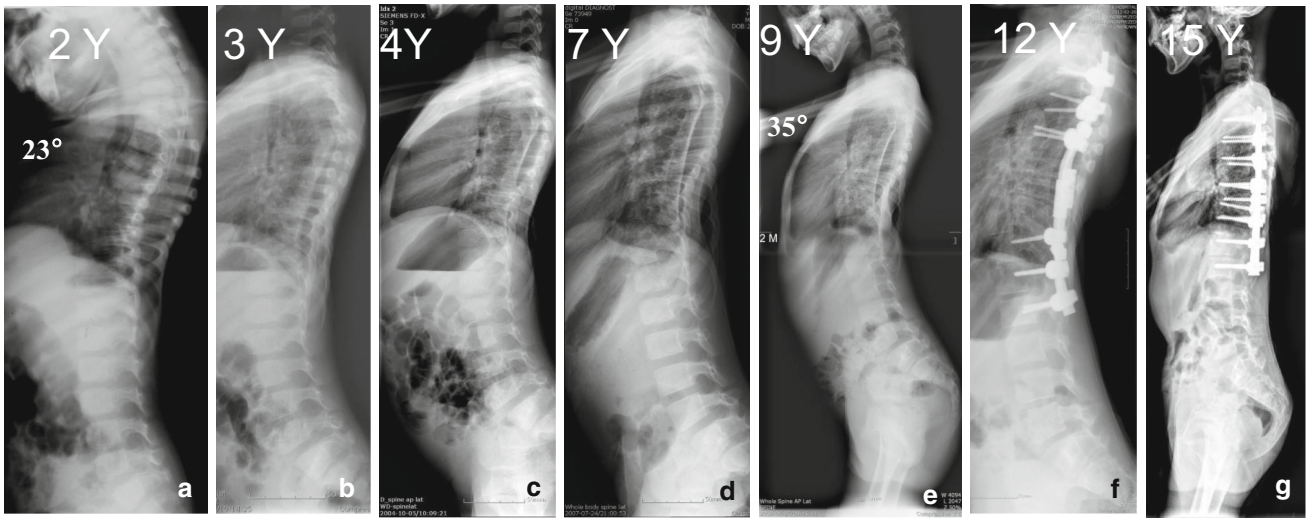

Fig. 1 Disease course of thoracic lordosis in the patient showing progression **a–e** (after 6 years of removal of fused lamina). **f** 3 years after correction of scoliosis. **g** 3 years after posterior crack osteotomy: 35° of thoracic lordosis was corrected with 5° of kyphosis

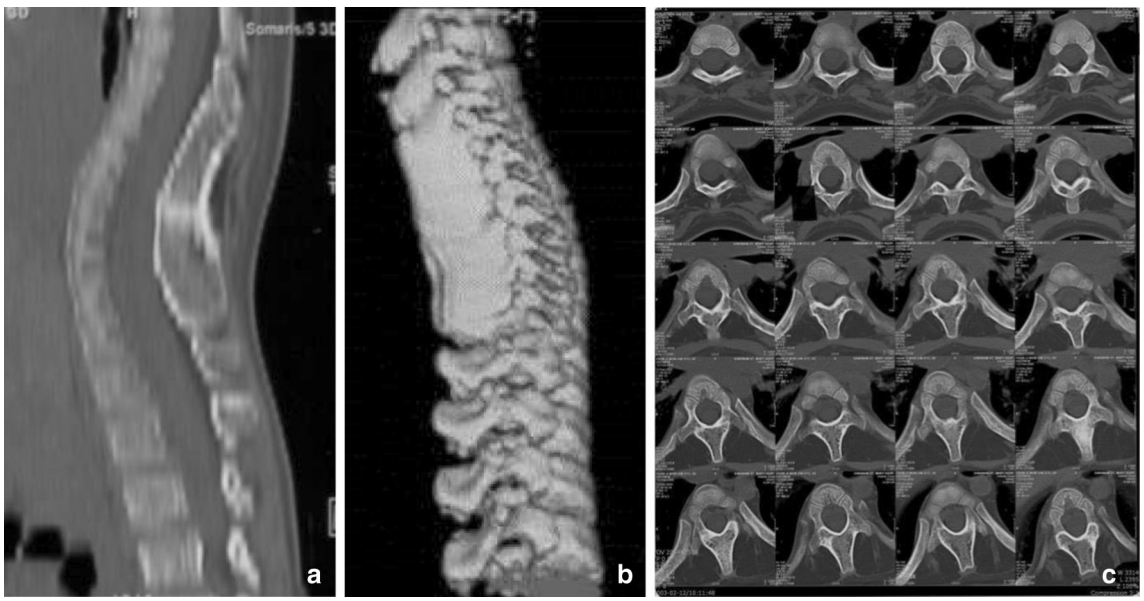

Fig. 2 Computed tomography **a**, **b** congenital fusion from T4 to T11 at age of 3 years. **c** Axial views show normal neurosynchondrosis

posterior laminectomy to prevent further progression of lordosis.

Lordosis progressed during the following 7 years (Fig. 1b–e), and CT showed spontaneous autofusion (Fig. 4a, b). Plain radiography showed the appearance of a unilateral unsegmented bar (Fig. 5a–e). Revision surgery and correction of scoliosis were undertaken alongside the correction of hyperlordosis by osteotomy of the fused segments, combined with anterior fusion. Correction of the hyperlordosis was unsuccessful, but correction of the scoliosis was successful. Three years after this procedure (Figs. 1f, 5f at the age of 12 years), the lordosis had progressed, and the patient was experiencing a harder time breathing while walking (Fig. 6a–e) with forced vital capacity (FVC) was 0.98L and forced expiratory volume in one second (FEV1) 0.74L. Surgery was carried out to remove the implants and to perform PMVO of the fusion mass and correction of the deformity. After screw fixation from T1 to L1 was performed, PMVO was carried out at two sites: between T8 and T9 and between T11 and T12 (Fig. 7a). Osteotomy for anterior one-third of the vertebral body was completed by shaking of an osteotome in cephalad and caudal direction with an osteotome being in situ mobility of body was confirmed [6]. However,

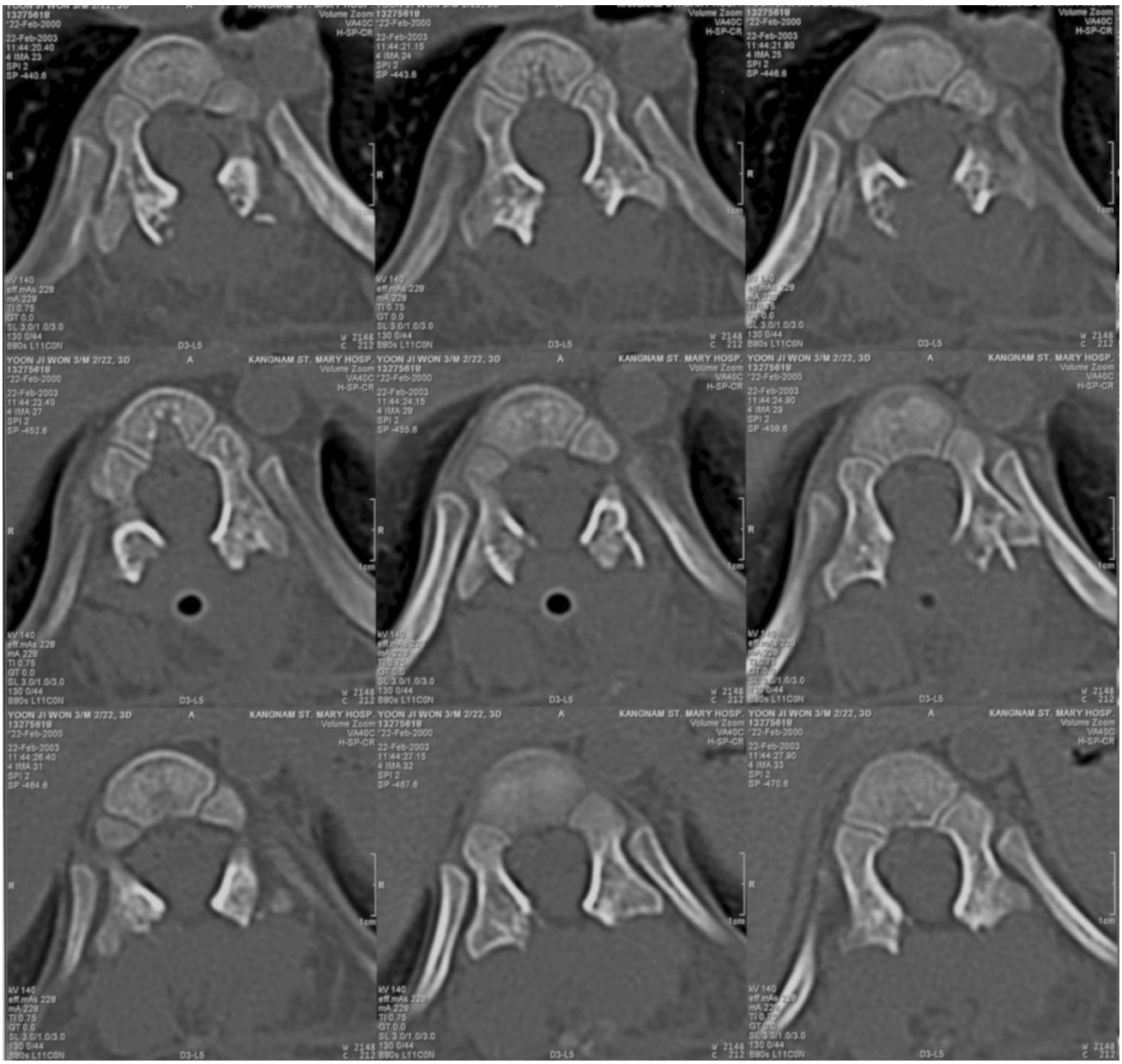

Fig. 3 Computed tomography after removal of fused lamina

during the operation, transient changes were observed in the motor evoked potentials (MEPs), along with changes in oxygen saturation. The procedure was stopped, and temporary rod fixation on one side was applied and MEPs returned to normal. Bilateral pleural effusion and paralytic ileus developed after surgery; the former condition was managed with a chest drain and the paralytic ileus resolved on its own. The patient was weaned off the ventilator gradually over a period of 4 days until he was comfortable. The surgical procedure was attempted again 1 week later. In the second procedure, two rods were slowly manipulated under MEP monitoring to achieve correction of thoracic kyphosis by 5°. The intraoperative course was uneventful. The patient was discharged after 1 week and has been in follow-up for the last 3 years (Figs. 1g, 5g, 7b). At last follow-up, FVC was 1.00L and FEV1 0.94L. Spine length on sagittal CT scan had been 202 mm from T1 to L2 preoperatively and measured 210 mm after 2.5 years of surgical correction (Fig. 8).

Discussion

Scoliosis associated with thoracic lordosis is linked with a significant degree of respiratory compromise and rib and intraspinal anomalies [4, 7–11]. Bartlett et al. [12]

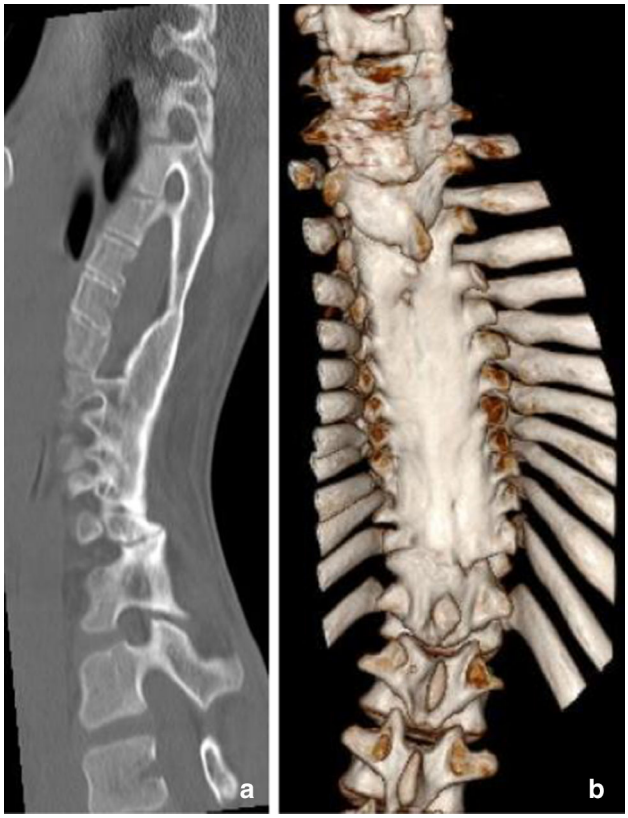

Fig. 4 7 years after removal of the fused lamina, spontaneous autofusion developed from T3 to T11. **a** Sagittal view. **b** 3-D computed tomography

recommended early correction to prevent irreversible changes to the lung function for thoracic lordoscoliosis associated with focal bronchial obstruction.

Correction of thoracic lordoscoliosis using a staged and combined anterior and posterior approach has been proposed [1, 2]. In this case, the initial surgery was performed to obtain mobile segments by removing the fused lamina, with expected spontaneous correction of lordosis from anterior longitudinal growth resulting from the primary ossification center, which was visible from CT. The neurocentral synchondrosis is situated anterior to the pedicles and at a distance that enables formation of the true vertebral body not only at the center, but also at an adjacent portion of the neural arch. Therefore, we expected anterior longitudinal growth through removal of the tethering effect of congenital fused lamina, led to hyperlordosis. At that time, however, we did not realize a unilateral unsegmented bar which was not shown on radiographs. Lordoscoliosis progressed with appearance of unilateral unsegmented bar. When the patient was 9-year-old, CT showed that spontaneous autofusion had arisen after previous removal of the fused lamina, which could have prevented anterior longitudinal growth. Spontaneous autofusion has been known as one of the complications after spinal surgery for young children's spinal deformity. This study also showed unexpected autofusion after simple removal of the fused mass. Although lordosis remained uncorrected, scoliosis had been corrected in a satisfactory manner. Three years later, thoracic lordosis had progressed to causing mild breathing difficulties. At the time of this procedure, PMVO was a new technique of a 3-column osteotomy, which had developed to correct rigid scoliosis [6, 13, 14]. The PMVO can enhance flexibility in a rigid spine with a fusion mass, thereby allowing the degree of correction to be distributed at the osteotomized area. Therefore, this surgical procedure

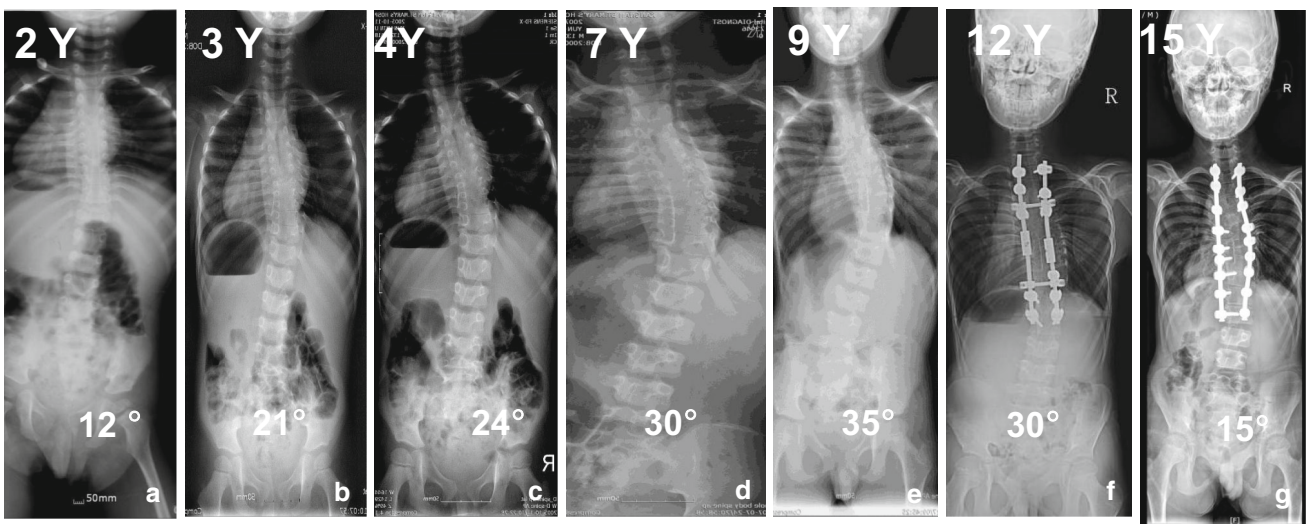

Fig. 5 Radiographs showing progression of scoliosis. **a** Scoliosis curve of 12° at age of 2 years. **b** Unilateral unsegmented bar appears after laminectomy. **c–e** Progression of scoliosis. **f** 3 years after correction of scoliosis. **g** 3 years after posterior crack osteotomy; scoliosis was corrected from 30° to 15°

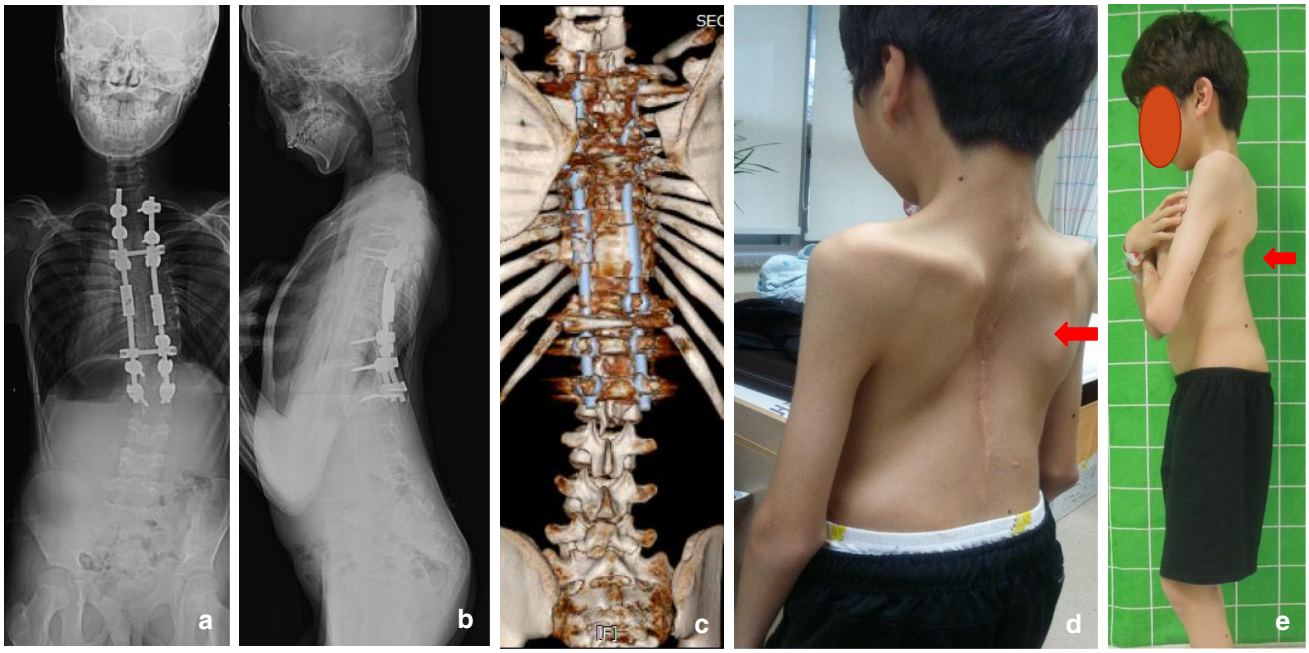

Fig. 6 Preoperative **a**, **b** Radiographs. **c** Computed tomography. **d**, **e** Clinical photographs; *arrows* indicate hyperlordosis of the thoracic spine

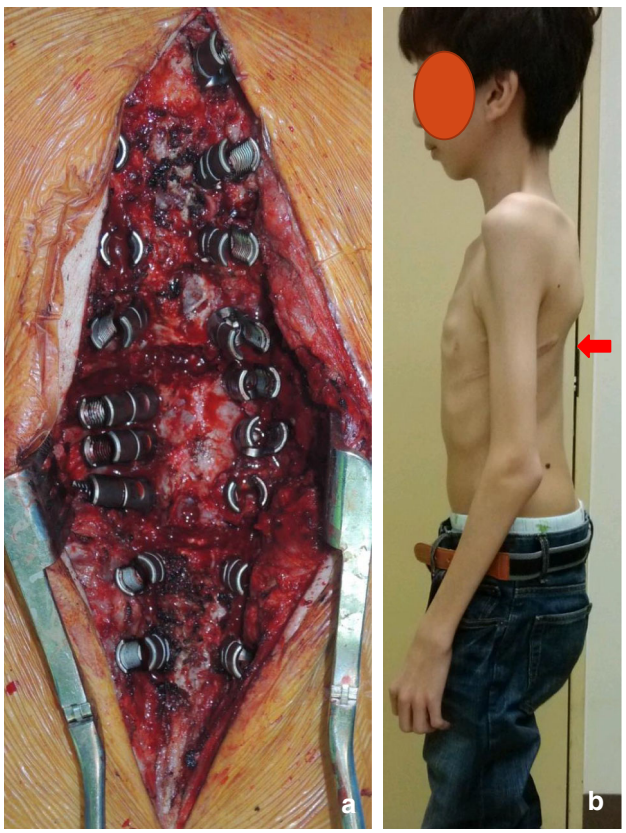

Fig. 7 **a** PMVO was performed between T8 and T9 and between T11 and T12. **b** Clinical photos show a well-balanced spine with significant improvement of lordosis (*arrow*) of the thoracic spine

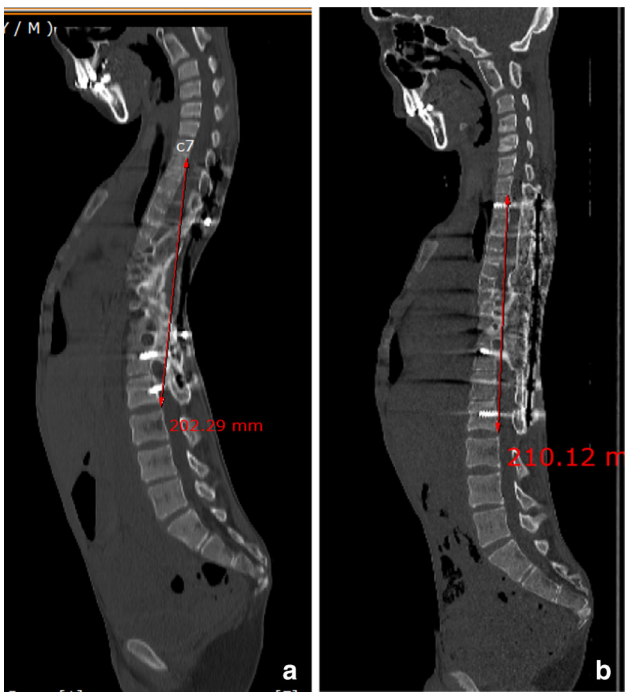

Fig. 8 Spine length was 202 mm from T1 to L2 preoperatively (**a**) and 210 mm after 2.5 years of the surgical correction (**b**)

can improve the coronal and sagittal balance. Follow-up at 3 years after the PMVO showed that correction of the deformity had been well maintained, and showed a good clinical outcome with a well-balanced spine. Therefore,

PMVO could be a potential option for managing rigid and severe congenital lordoscoliosis under MEP surveillance.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

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