# Two-Staged Surgery for Kyphoscoliosis in Larsen Syndrome with A 30-Year Follow-Up: A Case Report

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Larsen syndrome, which is a rare genetic disorder affecting connective tissue, is characterized by multiple joint dislocations, distinct facial features known as "dish face," cervical kyphosis or spinal scoliosis, and vertebral segmentation anomalies[1)]. Among these, thoracolumbar scoliosis can directly affect respiratory function and might require surgical treatment[2)]. However, long-term outcomes of surgeries for scoliosis in Larsen syndrome have been rarely reported[3)]. Thus, we present a surgically treated pediatric scoliosis case with Larsen syndrome comprehensively followed up for as long as 30 years since the initial visit.

A 5-year-old girl was referred to our hospital due to scoliosis with Larsen syndrome. Initial consultation revealed severe thoracic kyphoscoliosis, with a Cobb angle measuring 130 degrees of scoliosis and 65 degrees of kyphosis (Fig. 1-a). Therefore, a growth-friendly surgery was initially considered but not pursued due to possible complications associated with the procedure and her mental immaturity due to young age. She had undergone 9-year orthosis treatment until 14 years of age (Fig. 1-b, 1-c, 1-d), when she was deemed fit for correctional surgery. At the time of surgery, she exhibited skeletal maturity and severe kyphoscoliosis with a Cobb angle of 178 degrees of scoliosis and 95 degrees of kyphosis (Fig. 1-e). Two-staged surgery was performed: posterior release and external fixation for gradual correction (Fig. 2-a, 2-b), followed by posterior fusion[4)]. As she had severe respiratory dysfunction, which was unmeasurable by spirometry, anterior release was not performed. The surgery successfully improved thoracic scoliosis to 63 degrees and kyphosis to 60 degrees (Fig. 2-c).

Furthermore, her respiratory function drastically improved postoperatively; the vital capacity measured 580 ml at postoperative 1 year and increased to 780 ml at 2 years. Her physical condition gradually improved and she started to work at a facility for people with disabilities at the age of 18. However, she started to experience dyspnea when she was around 29-years-old along with weight gain. Her weight increased from 25 kg at the age of 14 to 46 kg at 29. She required home oxygen therapy at 29 and continuous positive airway pressure during night at the age of 35.

During the postoperative 20-year follow-up, no other complications nor correction loss were observed (Fig. 2-d). The clinical evaluation using SRS-30 showed a favorable outcome at the latest follow-up. The total average score was 3.9, and subdomain scores include the following: Function: 3.5; Pain: 5; Self-Image: 3.6; Mental Health: 4; and Satisfaction: 5.

While there have been surgical reports for cervical kyphosis in Larsen syndrome[5)], scoliosis treatments and their long-term outcomes remain unclear[6,7)]. Severe thoracic scoliosis can lead to respiratory dysfunction. It is crucial to prevent thoracic deformity until alveoli are formed around 8 years old and to recognize the necessity of early surgical intervention[8,9)]. In the current case, the delayed timing of surgery may have hindered optimal lung development. Since the surgery was performed after lung growth halted, improvement of her respiratory function had been limited, resulting in decompensation in 20-kg weight gain. If we knew the delayed surgery resulted in poor lung development, proactive weight-control measures could have been implemented. Higher

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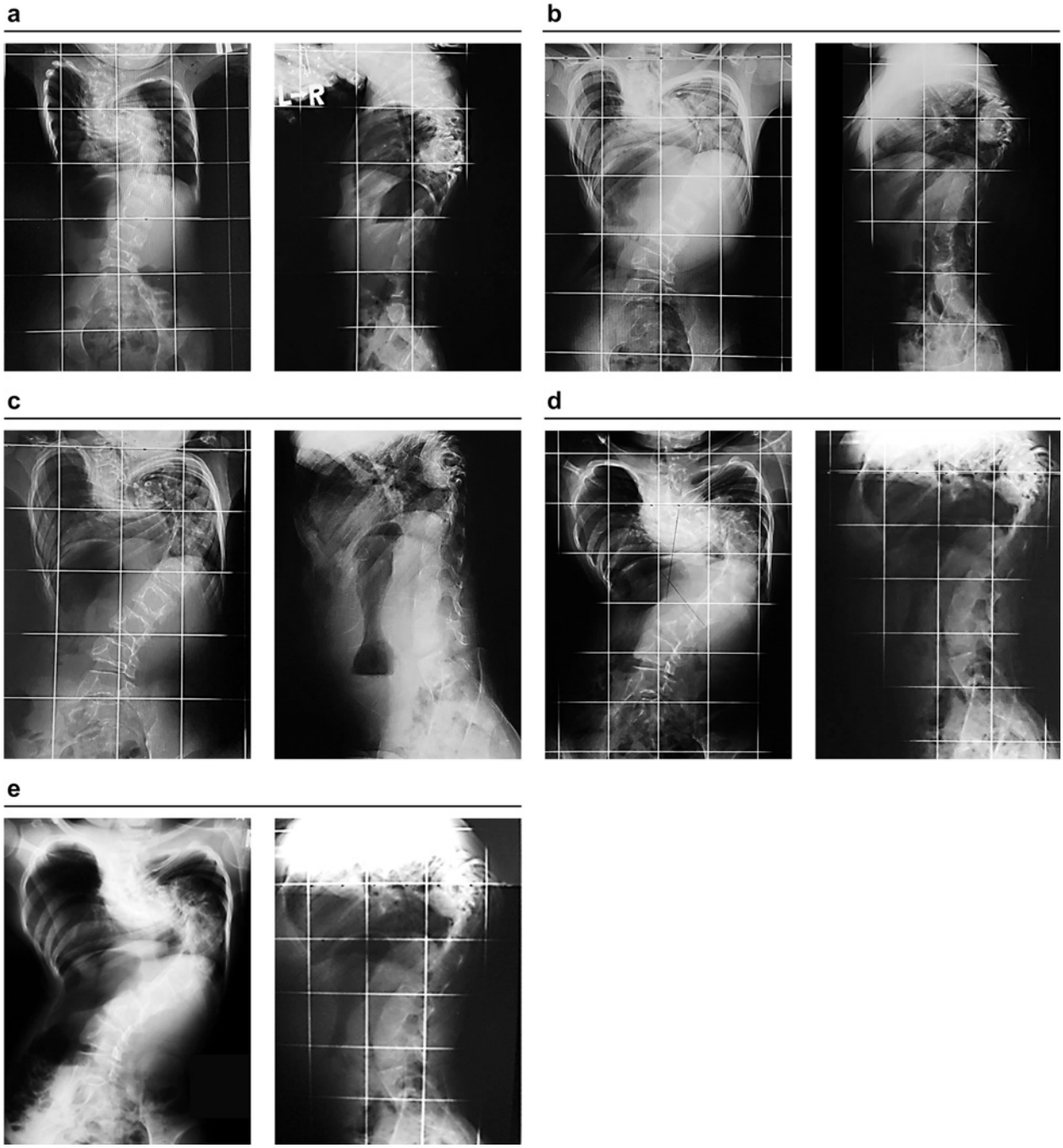


**Figure 1.** Preoperative radiography.
(a) At the age of 5, she already presented with 130 degrees of scoliosis and 65 degrees of kyphosis at the initial hospital visit. (b) At the age of 7 with 150 degrees of scoliosis and 70 degrees of kyphosis were found and (c) at the age of 9 with 160 degrees of scoliosis and 75 degrees of kyphosis were found. (d) At the age of 13, the deformity deteriorated to 165 degrees of scoliosis and 85 degrees of kyphosis, and (e) the preoperative Cobb angle was 178 degrees of scoliosis and 95 degrees of kyphosis.

body mass index (BMI) leads to reduced respiratory function, and weight loss could improve the dysfunction[10]. As for our patient, the height could not be accurately measured because of contractures in her hip and knee joints. Although minute changes in BMI were unclear, her BMI was 26.4 (132 cm, 45 kg) at the age of 29, when dyspnea appeared. Moreover, earlier surgical intervention and weight-control education might have improved her respiratory function.

Thus, we presented a case of severe scoliosis in Larsen syndrome, which highlights the significance of early surgical intervention and careful monitoring of scoliosis progression. Timely surgery can lead to improved long-term outcomes and potentially better respiratory function.

**Conflicts of Interest:** YT and KK: Surgical Spine Inc. (Endowed course). The other authors declare no competing

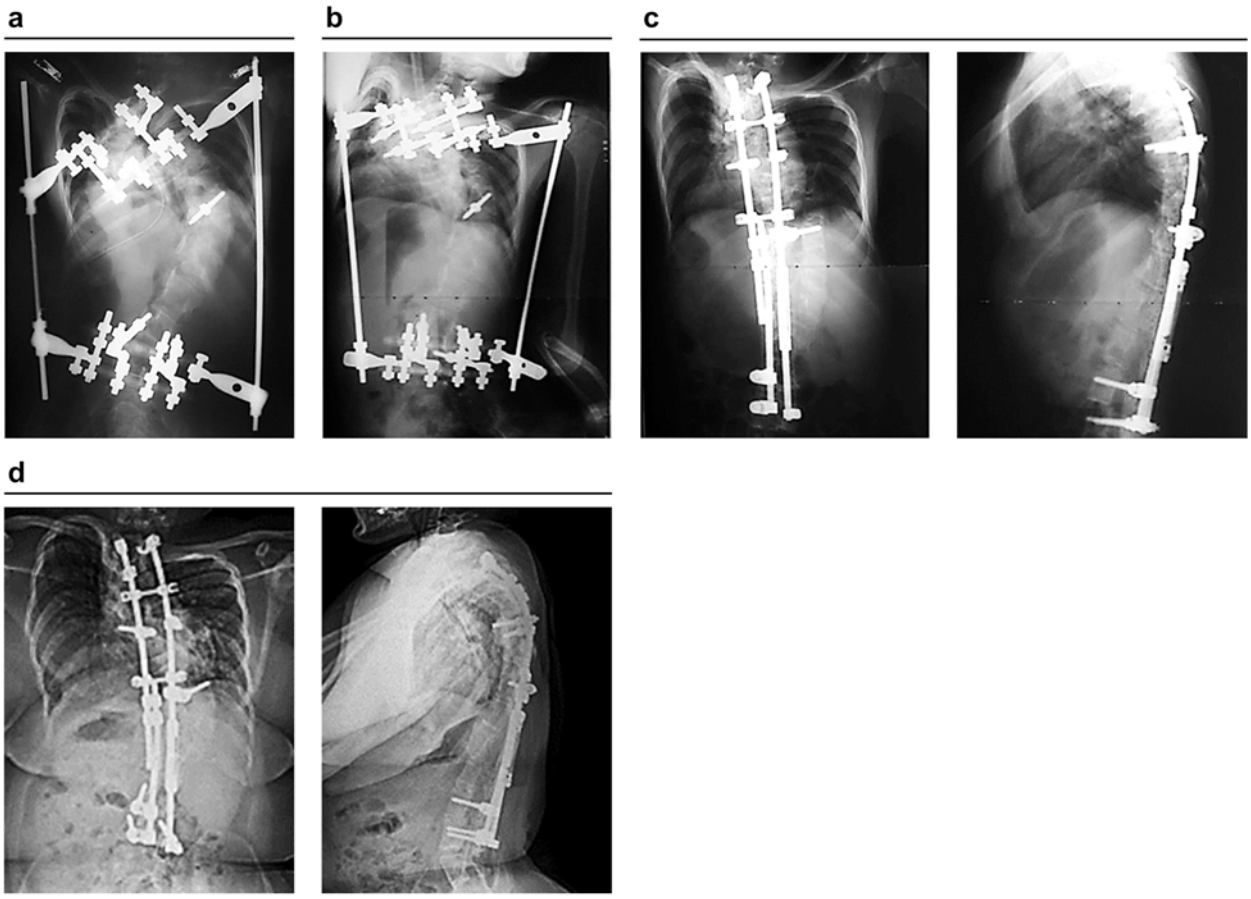


**Figure 2.** Postoperative radiography.
(a) The posterior release and external fixation was performed. Scoliosis was corrected from 178 degrees to 148 degrees and kyphosis was corrected from 95 degrees to 80 degrees. (b) The deformity was corrected gradually, and the scoliosis was corrected to 78 degrees after the one-month gradual correction. (c) One month later, the posterior definitive fusion was performed and the scoliosis was corrected to 60 degrees. (d) She had a favorable course for 20 years without any correction loss.

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**Ethical Approval:** This study was approved by the institutional review board (IRB) of Kobe University Hospital/Kobe University Graduate School of Medicine (approval code: B190002).

**Informed Consent:** Written informed consent was obtained from the patient and family. Furthermore, they were informed that data from the case would be submitted for publication and gave their consent.

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