Early-Onset Scoliosis Associated with Shprintzen-Goldberg Syndrome Treated with Growing Rods and Required Multiple Unplanned Surgeries: A Case Report

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Shprintzen-Goldberg syndrome (SGS) is a systemic connective tissue disease reported by Shprintzen et al. in 1982 and causes skeletal malformations similar to Marfan syndrome¹). Robinson reported that 23 of 37 patients with SGS presented with scoliosis²). However, there are few reports on the surgical treatment for early-onset scoliosis with SGS that was followed until bone maturity³).

Previously, Watanabe reported four patients with SGS whose scoliosis was treated surgically and revealed a high incidence of complications⁴. We followed one patient in the previous report until bone maturity.

She was born weighing 2600 g on the 39th week and 5th day. At infancy, she was pointed out of the hypertelorism, scaphocephaly, arachnodactyly, and thorax deformity and was diagnosed with SGS from these clinical features. She was referred to our department because of scoliosis progression at 3 years and 6 months of age. Standing radiographs showed scoliosis of 53° at T1-T7, 103° at T7-L1, and 60° at L1-L5 and kyphosis of 61° at T9-L3 (Fig. 1).

Considering severe deformity, growing rod surgery was initiated at 4 years of age, placing the proximal anchor at T 2 and T3 and distal anchor at L3 and L4 with the dual growing rods (Fig. 2). She underwent rod lengthening five times at 6 years of age, but she had dislodgement of the proximal anchor four times during that period. She had deep wound infection at 10 years of age. Although surgical debridement was performed several times, wound infection could not be controlled. We finally removed the spinal implant at 10 years of age. Afterward, thoracic kyphosis and

scoliosis deteriorated progressively to 66° and 117° , respectively, at 11 years of age. Although the bone was immature with Risser 0 grade and triradiate cartilage was open, we decided to perform posterior correction and fusion from T2 to L4. A few weeks postoperatively, the hooks placed at the T2 were dislodged and caused evident prominence at the base of the neck, which finally caused skin inflammation and infection. We removed the implants at the uppermost instrumented vertebra twice. Finally, we removed all spinal implants because we could not control deep wound infection at 13 years and 6 months of age. Although the infection subsided, kyphosis at the upper thoracic area progressed to 168° at 14 years of age (Fig. 3). She had aortic valve surgery for aortic regurgitation at 12 years of age.

We planned correction and fusion surgery from C3 to T 12, but we could not approach the middle and lower cervical areas because of severe kyphosis at the upper thoracic area. Since the lumbar curve (T12-L4) has already fused by the previous fusion surgery with acceptable balance in both coronal and sagittal planes, we did not include the lumbar curve in the surgical area. Thus, we performed T2-T12 posterior correction and fusion surgery with T4 posterior vertebral column resection. Postoperatively, kyphosis was corrected to 81° (Fig. 4). In the second surgery, we extended fixation up to C3, and kyphosis was finally corrected to 50° (Fig. 4). At 6 months postoperatively, slight spondylolisthesis at C2/C3 was noted on radiographs, which was stable until 4 years postoperatively at 20 years of age.

In SGS, the risk of wound infection and implant dislodge-

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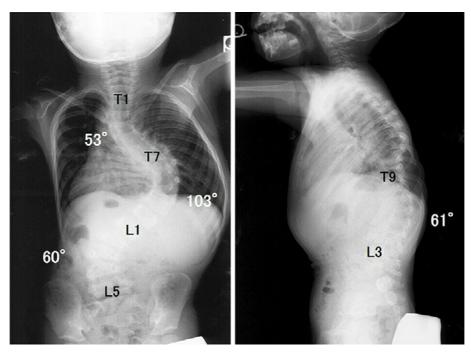


Figure 1. Radiographs before growing rod treatment. Standing whole spine radiographs showed scoliosis of 53° at T1–T7, 103° at T7–L1, and 60° at L1–L5 and kyphosis of 61° at T9–L3.

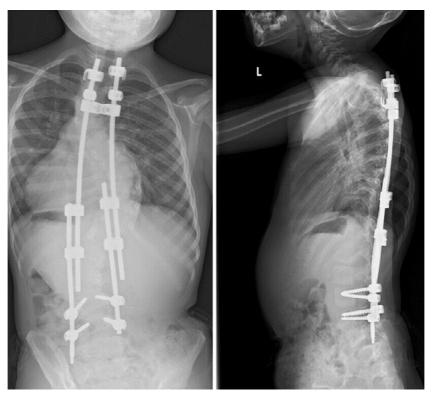


Figure 2. Radiographs after growing rod placement. At 4 years of age, growing rod surgery was initiated, placing the proximal anchor at T2 and T3 and distal anchor at L3 and L4, with the dual growing rods connected using side-to-side connectors.

ment was higher because of leptosome and fragility of the soft tissues and bone. Furthermore, the high complication rate related to implants of growing rod surgery has been reported^{3,5,6)}. To prevent complications in the treatment of this disease, we should consider a two-term operation plan that precedes the installation of an anchor, reduction of damage

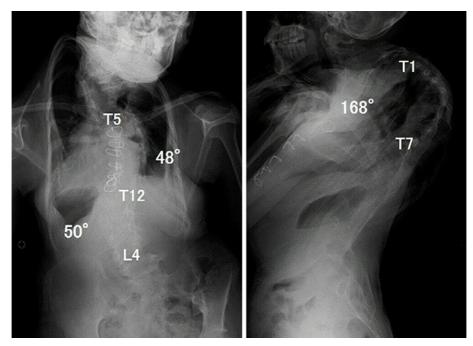


Figure 3. Radiographs before final correction and fusion surgery. Kyphosis at the upper thoracic area significantly progressed, after all spinal implants were removed because of wound infection. At 14 years of age, kyphosis reached 168°.

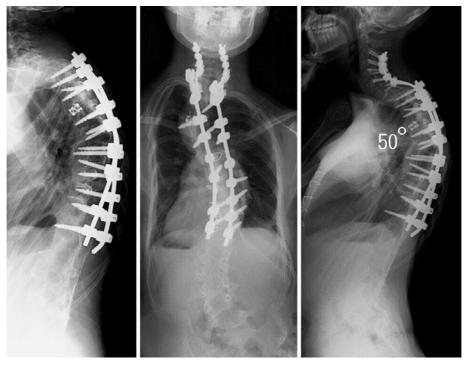


Figure 4. Radiographs after final correction and fusion surgery with T4 VCR. After T2–T12 posterior surgery with T4 posterior vertebral column resection, kyphosis was corrected to 81° that enabled access to the cervical spine (left). In the second surgery, we extended fixation up to C3, and kyphosis angle was finally corrected to 50° (middle and right).

of soft tissue, and a posterior element such as the spinous interspinous ligament and use of more flexible rods and flap to cover the implants.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Ethical Approval: None

Informed Consent: The patient was informed that data concerning the case would be submitted for publication, and she provided consent.

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