

*Case Report*

## Computer Navigation-Assisted Spinal Fusion with Segmental Pedicle Screw Instrumentation for Scoliosis with Rett Syndrome: A Case Report

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Scoliosis is a common clinical manifestation of Rett syndrome, a neurodevelopmental disorder that almost exclusively affects females. The spinal curve in patients with Rett syndrome is typically a long C curve of a neuromuscular type. As the onset of the scoliosis is very early and shows rapid progression, early surgical intervention has been recommended to prevent a life-threatening collapsing spine syndrome. However, there are high perioperative risks in Rett syndrome patients who undergo spinal surgery, such as neurological compromise and respiratory dysfunction due to rigid spinal curve. We herein report the surgical result of treating severe rapid progressive thoracic scoliosis in a 16-year-old girl with Rett syndrome. Posterior segmental pedicle screw fixation was performed from T1 to L3 using a computer-assisted technique. Post-operative radiography demonstrated a good correction of the curve in both the sagittal and coronal alignment. There were no postoperative complications such as neurological compromise. The patient had maintained satisfactory spinal balance as of the 3-year follow-up examination.

**Key words:** Rett syndrome, scoliosis, computer navigation-assisted surgery, segmental pedicle screw fixation

**R**ett syndrome (RS) is a neurodevelopmental disorder, seen almost exclusively in females (1 : 10,000) in all racial and ethnic groups worldwide [1]. After a 6-to 18-month period of normal development, RS manifests such symptoms as hypotonia, autism-like behavior, aphasia, loss of voluntary control of the upper limbs with the appearance of typical involuntary movements, and unstable wide-based gait with equinus deformity [2]. The cause of RS is a genetic mutation (MECP2) on the X chromosome

(Xq28) [3]. The diagnosis of RS is most often made in young girls at an age of 2-5 years, and the patients usually survive into adulthood, barring illness or complications. In reports on RS, the loss of ability or regression is often addressed [4]. These include communication, fine motor ability, and gross motor ability.

Scoliosis is a common clinical manifestation of RS, occurring in two-thirds of RS patients [5]. The curve in RS is a neuromuscular type with a very early onset, as well as rapid progression. Although there are a few reports of surgery in RS patients with scoliosis [6-10], surgical outcomes are not satisfactory due to the rigid spinal curve [9]. To achieve a good correc-

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tion of the curve and maintain the correction, segmental pedicle screw fixation has been recommended [11–13]. A new navigation system that allows relatively safe performance of this procedure became available recently [14, 15]. However, to date there is only one report on navigation-guided spinal fusion with segmental pedicle screw for the treatment of scoliosis with RS [10]. We present a second report of this procedure.

## Case Report

**Patient's history.** A 16-year-old female with a 12-year history of scoliosis in RS visited our hospital in 2004 due to scoliosis and spinal imbalance. She showed an early period of apparently normal development until she reached 2 years old. A period of temporary regression followed during which she lost communication skills and purposeful use of her hands. Thereafter, stereotypic involuntary hand movements and gait disturbances appeared. These stereotypic hand movements included hand-washing and hand-clasping motions. Soon thereafter she could not move her body smoothly and also lost her sense of coordination. Seizures tended to lessen in their intensity in later adolescence. She was diagnosed with RS at a local hospital in 1993. When she first came to our hospital in 2004, she could not walk because of the spinal imbalance. Two years later, her scoliosis had gradually progressed, and her spine had developed a severe curve.

**Examination.** The patient was not able to walk without supports, and she was confined to a wheelchair. She was 138 cm tall and weighed 31 kg. Her mental retardation prohibited her ability to communicate. She always moved her fingers like she was washing her hands. There was hyperreflexia of her legs, but no apparent muscle weakness. There was a severe rib hump and spinal imbalance (Fig. 1).

**Imaging.** Plain radiographs demonstrated severe thoracic scoliosis from T5 to L2. The curve pattern was a long C, and its Cobb angle was 88 degrees. The patient underwent magnetic resonance imaging (MRI), but no specific findings were identified. In the radiographs under traction, the Cobb angle was reduced to 55 degrees, and some flexibility was confirmed (Fig. 2). Three-D computerized tomography was taken for operation planning and the com-

puter-assisted procedure (Fig. 3).

**Operation.** Posterior segmental pedicle screw fixation was performed from T1 to L3. The Stealth Station navigation system<sup>R</sup> (Sofamor Danek, Memphis, TN, USA) was used during insertion of pedicle screws. The accuracy of the navigation was 0.4 mm. During surgery, recording of spinal cord evoked potential after spinal cord stimulation was performed, and there was no abnormality of the waves. The surgery time was 5 h 30 min, and estimated blood loss was 1,600 ml. A transfusion of 740 ml of autologous blood was enough to control the post-operative anemia. Post-operative radiography demonstrated a good correction of the curve both in the sagittal and coronal alignment. The post-operative Cobb angle was 23 degrees, and the correction rate was 67% (Fig. 4).

**Postoperative course.** There were no postoperative complications, such as neurological compromise. Two weeks post-operatively, the patient was able to walk with a little support, and she was thereafter discharged from the hospital without any brace. She had good spinal balance at the 3-year follow-up examination (Fig. 5).

## Discussion

Scoliosis in RS is very common [5]. Its onset is very early, and the curve patterns are usually long thoracic or thoracolumbar curves with frequent con-

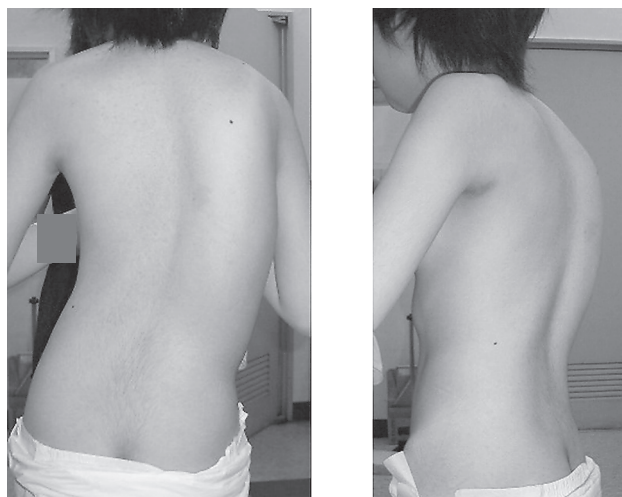


Fig. 1 Preoperative appearance of scoliosis in a patient with Rett syndrome.

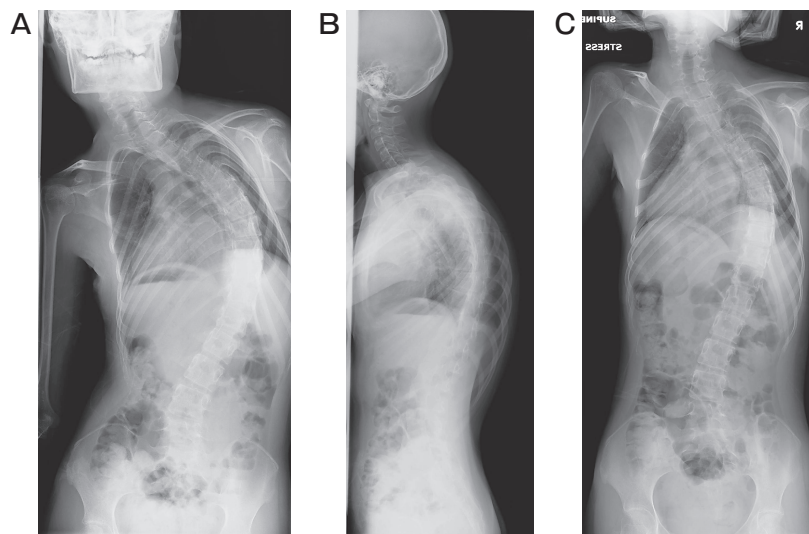


Fig. 2 Preoperative A-P (A), lateral (B), and traction (C) radiography. The Cobb angle was 88 degrees from T5 to L2, and the spinal valance was not acceptable.

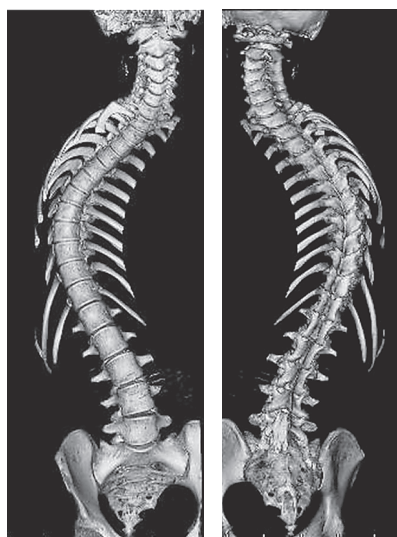
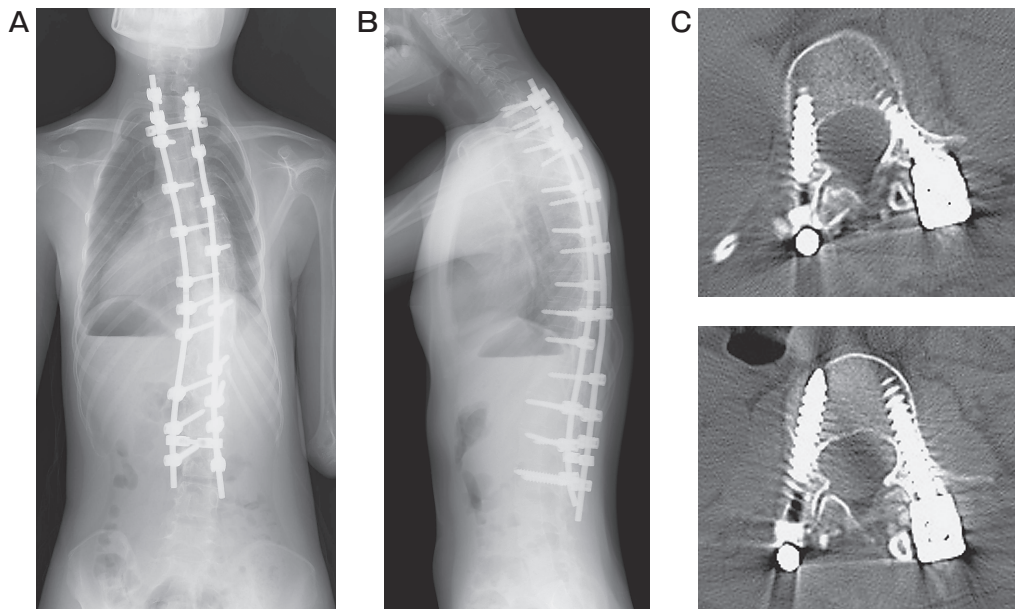


Fig. 3 Preoperative 3-D computerized tomography. There is no obvious bony anomaly.

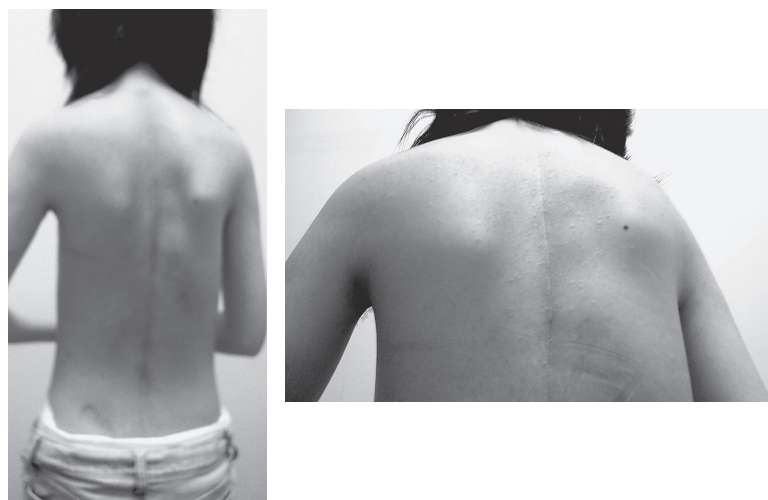
comitant marked kyphosis [19]. As scoliosis with RS is classified as a neuromuscular scoliosis, the spinal curve will continue to progress after skeletal maturity. The deformity progresses rapidly and often requires surgical treatment. There are few reports of surgery in scoliosis with RS [5–10], and the evaluation of its results has been controversial. However, based on the fact that severe scoliosis will shorten the patient's life expectancy due to cardiopulmonary complications, we recommend early surgery if the patient has a curve

over 50 degrees or severe pelvic obliquity.

A surgical correction was reported in 91 classic cases of scoliosis with RS, and the authors recommended planning for surgery when the curve surpasses 40 degrees [8]. Soultanis *et al.* reported a combined surgery (anterior and posterior instrumentation and fusion) in a two-stage procedure for a 15-year-old female with RS, who had a rigid right thoracic curve of 120° with marked kyphosis [9]. Due to the severe rigidity of the curve, a limited correction was achieved. Currently, segmental pedicle screw fixation is a reliable surgical treatment method for the correction and maintenance against rigid scoliosis. To achieve an excellent correction in our patient, we performed segmental pedicle screw fixation. Kuklo *et al.* reported the surgical findings of 1,428 patients with adult idiopathic scoliosis [12]. They found that all pedicle screw and anteroposterior constructs had a lower surgical revision rate in comparison to hook and hybrid constructs [12]. Suk *et al.* reported 35 scoliosis patients who had severe scoliotic curves over 70° treated by segmental pedicle screw fixation [13]. After a minimum follow-up of 2 years, the results were a correction of 66% (53 degrees) and a loss of correction of 3.0% (3.7 degrees) [13]. They thus concluded that posterior segmental pedicle screw fixation without anterior release in severe scoliosis had a satisfactory deformity correction without a significant loss of curve correction.



**Fig. 4** Postoperative A-P (A), lateral radiography (B), and CT (C). The Cobb angle was corrected from 88 degrees to 43 degrees (A). The sagittal alignment became normal (B). The upper right CT is at the T9 level (apex), and the lower right CT is at the L1 level. All pedicle screws were correctly inserted.



**Fig. 5** Postoperative appearance. Spinal balance became normal.

Scoliosis patients with Rett syndrome are relatively young, so the diameters of pedicles are smaller than those of adolescent idiopathic scoliosis patients. The computer-assisted technique is thus helpful to attain a safe and correct insertion of the pedicle screws in patients with RS. Previous reports suggest

that the navigation system is a beneficial tool to perform the segmental pedicle screw fixation safely [14, 15]. Kotani *et al.* reported 45 patients who received posterior correction surgeries, and a perforation was observed in 11% of the control group (25 patients) and in 1.8% of the navigation group (20 patients) [14].



They concluded the use of surgical navigation system successfully reduced the perforation rate and insertion angle errors, demonstrating the clear advantage in safe and accurate pedicle screw placement during scoliosis surgery. Rajasekaran *et al.* also reported using Iso-C based navigation in pedicle screw fixation in 27 patients with scoliosis [15]. There were 54 (23%) pedicle breaches in the non-navigation group in comparison to only 5 (2%) in the navigation group. Iso-C navigation increases accuracy, while also reducing surgical time and radiation in thoracic deformity correction surgeries. Only one case of a segmental pedicle screw fixation for scoliosis in a patient with RS using a navigation system has been reported in the literature. The authors reported a 10-year-old female with RS, who had a thoracolumbar curve of 115 degrees with a reduced respiratory volume [10]. A two-stage surgical procedure (anterior and posterior) using a computer-assisted technique was performed, resulting in excellent reduction in the deformity (post-operative 24-degree Cobb angle). The results in the present case also suggest the usefulness of navigation-guided segmental pedicle screw fixation for the safe and satisfactory correction and maintenance against rigid spinal scoliosis in patients with RS.

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