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Surgical Experience with Scoliosis Associated with Adolescent Turner Syndrome: Two Case Reports and a Literature Review

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Abstract

Background: Turner syndrome (TS) affects approximately one in 2500 live births in females. Scoliosis is one of the skeletal manifestations of TS, but most cases only require observation or conservative treatment. We experienced two adolescent TS cases in which progression of scoliosis required surgical intervention, which is very rare in TS. **Case Presentation:** Case 1: An 11-year-old female with TS had a single thoracic curve that rapidly progressed to a triple major curve with a 76˚ main thoracic curve at age 13.5 years. Case 2: A 14-year-old female with TS had a 59˚ single thoracic curve. In both cases, growth hormone and estrogen replacement therapy were administered preoperatively and planned postoperatively. Posterior correction and instrumented fusion using simultaneous translation on two rods technique and direct vertebral rotation with the use of multiple rod introducers were successfully performed in both cases. No crankshaft phenomenon or distal adding on were observed during those postoperative courses. **Conclusions:** Although curve pattern of the deformity is similar to adolescent idiopathic scoliosis (AIS), bone quality in patient with TS is lower. In the context of surgical interventions for scoliosis associated with TS, it is imperative to employ surgical techniques that take into account the suboptimal bone quality. If continuation of hormone replacement therapy is planned after corrective surgery for scoliosis in TS patients, it is essential to follow the patient closely postoperatively until bone maturation is complete.

Keywords

Turner Syndrome, Scoliosis, Hormone Replacement Therapy, Simultaneous Translation on Two Rods, Rod Introducer

1. Introduction

TS is characterized by a complete or partial absence of an X chromosome. The incidence of TS in females is estimated to be 1/2500 to 1/3000 [1] [2]. Skeletal deformities including short stature, spinal deformity, delayed skeletal maturation, and osteoporosis have been associated with TS, and it is important for orthopedic surgeons to be familiar with these skeletal manifestations because TS may be the primary diagnostic consideration for a patient who presents with short stature, scoliosis, or slipped capital femoral epiphysis. The reported prevalence of scoliosis in patients with TS is higher than that in the general population [3] [4]. However, there is a paucity of research on surgical interventions for scoliosis in these patients, with the reported frequency of surgical correction in TS scoliosis ranging from 0% to 11% [5]. Here, we describe two rare cases of scoliosis associated with TS that were successfully treated surgically, along with a review of the literature.

2. Case Presentation

2.1. Case 1

An 11-year-old female was diagnosed with TS (karyotype: 45, X) at six years old by a pediatrician at another healthcare facility. Three years later, at age nine, she started growth hormone replacement therapy (GHRT) for short stature. At age 10, a plain chest X-ray incidentally revealed scoliosis, and during subsequent regular follow-up, the patient was referred to our hospital due to the progressive nature of the deformity. At the initial consultation with our hospital at age 11 years 6 months, the right thoracic Cobb angle was 41˚ (**Figure 1**). We proposed introduction of orthotic brace therapy to her parents, but they hesitated and withheld consent. Six months later, at age 12, the Cobb angle had progressed to 47˚, and a year later to 58˚. At age 13, the patient started low-dose estrogen replacement therapy (ERT), in addition to GHRT. Since she was scheduled to continue hormone replacement therapy (HRT), we expected further progression. At age 13 years 6 months, the main curve had progressed to 76˚ and was classified as Lenke type 4C(N) [6]. Thus, corrective surgery was planned (**Figure 2**).

A preoperative physical examination showed that the patient had a height of 135 cm and a short stature (−2.9 SD). There were no preexisting cardiovascular or psycho-neurological morbidities, and the anesthesiologist reported no complications during scoliosis surgery under general anesthesia. To avoid venous thromboembolism (VTE), ERT was suspended one month prior to surgery. Preoperative bone mineral density (BMD) tests revealed low values for the whole body (0.757 g/cm^2) and femoral neck (0.455 g/cm^2), so bone fragility had to be considered during the corrective surgery.

At age 14, posterior correction surgery extending from T2 to L3 was performed. Considering the poor bone quality, a posterior approach and a multilevel segmental anchoring technique using all uniplanar pedicle screws in conjunction with an O-arm navigation system (Medtronic Sofamor Danek, Memphis, TN, USA) were used. Ponte osteotomy [7] was added to the proximal thoracic and main thoracic

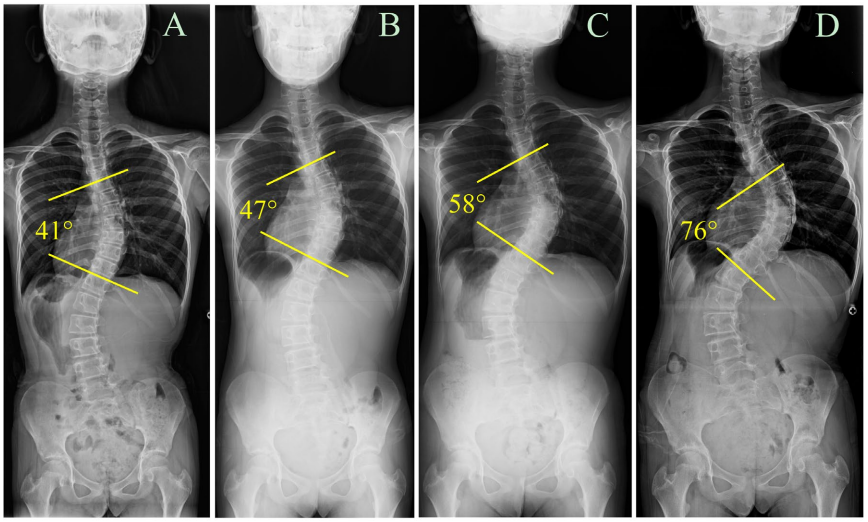

Figure 1. Time course of full-length standing spine radiographs in case 1. (A) 11 years 6 months; (B) 12 years; (C) 13 years; (D) 13 years 6 months.

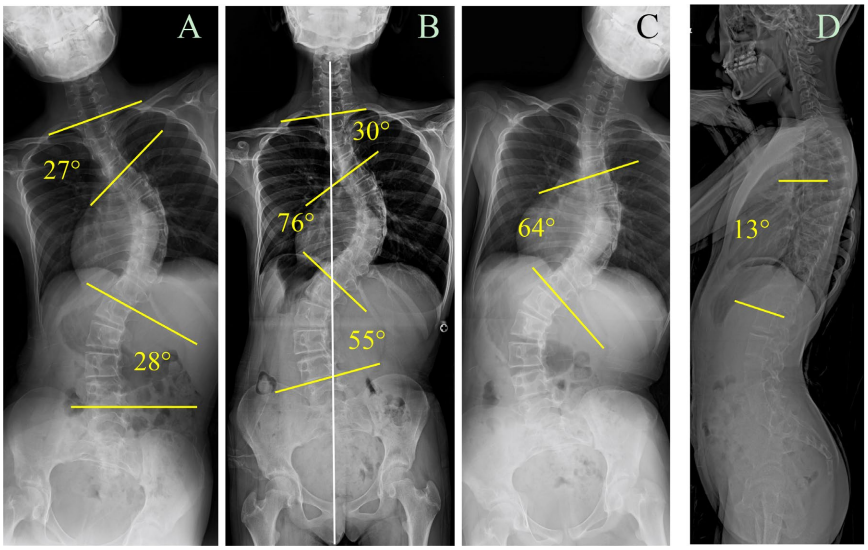

Figure 2. Preoperative evaluation of curve flexibility in case 1. (A) (C) Supine position lateral bending test; (B) Standing postero-anterior radiograph; (D) Standing lateral view radiograph. The white line in (B) indicates the center sacral vertical line.

curves. Correction of the coronal plane was mainly carried out using the simultaneous translation on two rods (ST2R) technique with use of multiple rod introducers (or rod reducers) bilaterally. Two 6.0-mm cobalt-chrome alloy rods were differently contoured according to the sagittal and axial plane correction. After rod introducers were attached to all screw heads, the two contoured rods were inserted into the introducers and gently passed through all segments simultaneously. Then, translational forces were gently and slowly added by gradual tightening of the rod introducers. A bilateral direct vertebral rotation maneuver (DVR) was added to correct the axial rotational deformity. The corrective maneuvers were executed with particular caution and slowness to avoid loosening or pulling

out of the screws. The surgical time was 5 hours 40 minutes and estimated blood loss was 640 ml, which was covered by autologous blood transfusion only.

Immediately after surgery, the main thoracic curve was corrected to 12˚ and the thoracic kyphosis to 28˚. GHRT was administered until the age of 18, at which point skeletal maturation was achieved. At seven years after surgery, while the patient is still receiving ERT, the corrected position remains stable and the correction rate is high, at 83% (Figure 3) without a crankshaft or adding-on phenomenon. The total SRS-22 score increased from 76% preoperatively to 88% six years postoperatively, with notable improvements in self-image and satisfaction (Table 1).

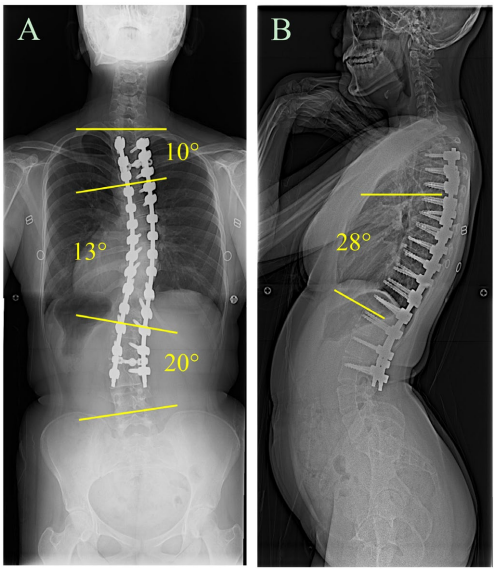

Figure 3. Full-length standing postero-anterior radiograph (A) and lateral radiograph (B) of the spine 7 years after surgery in case 1. The corrected alignment was well maintained. The correction rate was 83% without instrumentation failure and a crankshaft/adding-on phenomenon.

Table 1. Scoliosis Research Society (SRS)-22 scores.

		Before surgery	Latest follow-up
Case 1	Function	4.2/5	4.2/5
	Pain	4.4/5	4.6/5
	Self-image	3.4/5	4.6/5
	Mental health	3.6/5	4.4/5
	Satisfaction	3.0/5	4.0/5
	Total	3.8/5 (76.0%)	4.4/5 (88.0%)
Case 2	Function	4.8/5	4.8/5
	Pain	4.0/5	5.0/5
	Self-image	2.2/5	4.4/5
	Mental health	3.8/5	4.8/5
	Satisfaction	5.0/5	5.0/5
	Total	3.8/5 (76.0%)	4.8/5 (96.0%)

2.2. Case 2

A 14-year-old female was diagnosed with TS (karyotype: 45, X/46, XX mosaic) at age 10 by a pediatrician at our hospital, and GHRT was initiated for treatment of short stature. Subsequently, low-dose ERT was started at age 13. Scoliosis was detected on a chest X-ray at age 10 and has since deteriorated, leading to referral of the patient to our orthopedic department. At her initial visit, at 14 years 9 months old, her height was 148 cm (−1.3 SD). BMD in the lumbar spine was relatively low (0.740 g/cm^2) in a preoperative test. There were no preexisting cardiovascular or psycho-neurological morbidities. The Cobb angle of the main thoracic curve was 59˚ and was classified as Lenke type 2A(N). Consequently, surgical intervention was scheduled.

Under O-arm navigation guidance, posterior correction and fusion were gently performed from T3 to L2 using segmental uniplanar pedicle screws (except for right T11 because the pedicle was too narrow to insert the screw) and a ST2R maneuver with use of multiple rod introducers bilaterally. Ponte osteotomy was added for the proximal thoracic and main thoracic curves. DVR was gently added to correct the axial rotational deformity. The basic surgical techniques in this case were the same as those in case 1 above. The surgical time was 5 hours 10 minutes and estimated blood loss was 410 ml. Only autologous blood transfusion was performed. The main thoracic curve was corrected to 17˚ and the thoracic kyphosis to 14˚ after surgery. Three years later, at the time of writing of this case report, the patient continues to receive both GHRT and low-dose ERT. Although bone age remains below the level of maturity, the alignment has been maintained, and the correction rate of the main thoracic curve is 71% (Figure 4). The total SRS-22 score increased from 76% preoperatively to 96%, with notable improvements in self-image and mental health (Table 1).

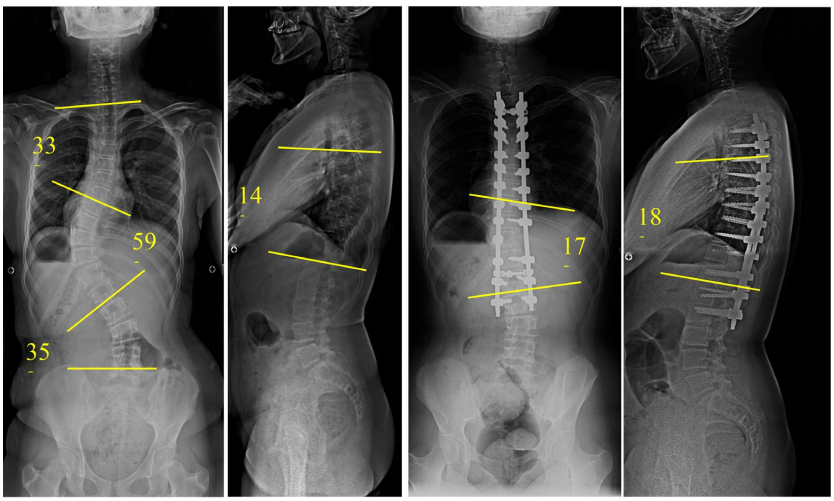

Figure 4. Full-length standing postero-anterior and lateral spine radiographs in case 2 before surgery (A) (B) and 3 years after surgery (C) (D). Good correction was achieved with a correction rate of 71%.

3. Discussion

TS is the most common female chromosomal anomaly, and results in physical features that include underdeveloped genitalia, cardiac and urinary anomalies, and skeletal manifestations such as short stature, Madelung deformity, cubitus valgus, and spinal deformities [3] [8] [9]. The incidence of scoliosis associated with TS ranges from 10% to 59%, which is significantly higher than the prevalence of AIS in the general population [3] [4] [10]-[12]. However, most cases of scoliosis in TS are mild, with a main Cobb angle of less than 20˚. To our knowledge, surgical intervention has rarely been reported in cases of TS [13] [14]. Acosta *et al*. [9] proposed that patients should be followed closely until age 20 because the onset age of scoliosis in TS is relatively older than that in AIS. Their proposed treatment guidelines include a surgical indication based on curve severity and are almost identical to those for AIS because the characteristics of the deformity are similar. This similarity suggests that the etiology of scoliosis in TS may be related to that of AIS, but the actual causation and progression of scoliosis in TS remains unknown.

The relationship between scoliosis and GHRT in TS is controversial [3] [10] [11]. GHRT has been suggested to be an accelerant for scoliosis [9] [10], and Ricotti *et al*. found that patients with TS have a higher risk of developing scoliosis due to an indirect effect of GHRT [3]. Also, Acosta *et al*. suggested that GHRT may promote progression of pre-existing scoliosis in patients with TS [9]. Furthermore, in a report of four surgical cases, Ozone *et al*. noted that GHRT was continued after scoliosis surgery in two of those cases and a crankshaft phenomenon was observed postoperatively [13]. This phenomenon is characterized by a loss of three-dimensional correction of the scoliosis curve, which occurs after posterior fusion in children who have not yet completed skeletal growth due to rotational deformity of the spine created by continued anterior spinal growth [14]. The crankshaft phenomenon is mainly observed in infantile and juvenile idiopathic scoliosis cases, although it has also been documented in adolescents who are immature at the time of spinal fusion [15]. In the present cases, HRT was continued after surgery, and fortunately, the crankshaft phenomenon did not occur during the postoperative period.

ERT compensates for reduction in estrogen hormones in females. Its primary use is managing symptoms and maintaining bone quality in postmenopausal women. Involvement of female hormones, particularly estrogen, in the etiology of spinal deformities is uncertain, but current evidence suggests that estrogen may play an important role in development of AIS, possibly by influencing the growth and maturation of spinal structures [16]. However, the exact mechanism through which estrogen exerts its effects on the spine remains unclear, and no direct causal relationship of ERT with scoliosis in patients with TS has been proven. One of the most important effects of ERT in adolescent TS is improvement of low BMD and poor bone quality. Faienza *et al*. suggested that patients with TS have lower cortical bone density than age-matched healthy controls at a young age [17]. In addition, Gravholt *et al*. found that patients with TS have a 10.12 times relative risk of

osteoporosis and a 2.44 times relative risk of osteoporotic spine fractures compared to age-matched controls [18]. ERT is typically pursued until the patient reaches natural menopause age of about 50 years old. This prolonged treatment is necessary because patients with TS frequently experience hormonal imbalances due to ovarian dysfunction. In the current cases, we had an intraoperative sense of poor bone quality during pedicle screw placement.

Pagadala *et al*. [5] reported on two cases of adolescent TS in a retrospective study of 306 patients with TS. The age at which surgery was performed, the correction method, and the technical details were not provided, but the correction rates of deformity were 22% and 59%, which cannot be considered sufficient. There are many modern surgical techniques that can be used to treat scoliosis, including rod rotation, simultaneous double rod rotation, vertebral translation, DVR, cantilever rod bending, in situ bending, and distraction-compression. We decided that translational force had the least risk of screw loosening and pull-out for scoliosis correction due to the low preoperative BMD in our cases, and thus, we chose the ST2R procedure [19] [20] with use of multiple rod introducers bilaterally. The principle of this surgical technique is predicated on correction by translation force and is based on the Isola system [21]. The advantages of this procedure are 1) the rod introducer can apply the correction force even before the rod is inserted into the screw head, 2) correction can be performed simultaneously and gently over the entire fusion level, and 3) since there is no need to bend the rod precisely to match each screw head position, the procedure is effective for correction of long segments and complex curvature. Furthermore, two recent studies reported that ST2R with rod introducers can restore thoracic hypokyphosis in patients with AIS [22] [23]. We were able to use this correction maneuver for syndromic scoliosis with poor bone quality, and the correction rate was 83% in case 1 and 71% in case 2. Our successful outcomes provide further evidence of the advantages of this technique.

To date, there has been no deterioration of the deformity or adjacent segment problems such as a distal adding-on phenomenon in either of our cases. Case 1 has received ERT for 7 years postoperatively and has a most recent femoral neck BMD of 0.662 g/cm^2, which is a significant improvement from the preoperative BMD of 0.445 g/cm^2. The postoperative SRS-22 scores demonstrate excellent improvement, especially in the three domains of self-image, mental health, and satisfaction. In fact, both patients started preoperative HRT for short stature, and both are very happy with their height gains postoperatively.

A limitation of the present study is the insufficient sample size, which precludes the possibility of conducting a statistical analysis. Thus, there is a need for accumulation and evaluation of more surgically treated TS cases in future studies.

4. Conclusion

If surgery is required to treat scoliosis in adolescents with TS, we recommend that the method of deformity correction should be chosen carefully, taking into account

the poor bone quality. Treatment of TS requires long-term HRT, and it is essential for orthopedic surgeons to collaborate with pediatricians and gynecologists to provide long-term follow-up for spinal deformities associated with TS using a multidisciplinary approach.

Consent to Patient

The patients and their parents were informed that data from these cases would be submitted for publication and gave their consent.

Conflicts of Interest

The authors declare that there is nothing to disclose regarding the content of this paper.

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