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CASE REPORT

Surgical Treatment of Scoliosis in Patients with Osteogenesis Imperfecta: A Case Report

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ABSTRACT

Patients with osteogenesis imperfecta (OI) face many challenges, including skeletal deformities and repeated extremity fractures. Spinal deformities in patients with OI are especially challenging for spine surgeons because of poor bone stock and rigid curves, often necessitating surgical treatment.

This case report presents two OI patients with scoliosis managed in our institution. Patient A was a woman in her 20s, while patient B was in her 30s. Both had progression of curves past skeletal maturity and underwent pre-operative bisphosphonate therapy before deformity correction and instrumentation. Due to acceptable bone density pre-operatively, non-fenestrated pedicle screws without cement augmentation were utilized for both patients. We did not encounter implant failure, junctional kyphosis, or pseudoarthrosis. Despite a loss of correction of 3% (mid-thoracic) and 9% (lumbar) for patient A and 2% (thoracic) and 6% (thoraco-lumbar) for patient B, both patients did not return to pre-operative measurements after 1 year of follow-up. They were also ambulatory and returned to employment postoperatively.

Keywords. scoliosis, osteogenesis imperfecta, pedicle screws, bisphosphonates

INTRODUCTION

Osteogenesis imperfecta (OI) is a rare disorder associated with collagen-related syndromes that may present in many ways. Patients with OI have fragile bones and are susceptible to extremity fractures, skeletal deformity, and typically have short stature. Historically, OI was classified by Sillence and Danks into 4 types,[1] but the classification was later expanded by Cole et al. to 11 types.[2] The types are classified according to four salient features: skeletal dysplasia, dentinogenesis imperfecta, bluish sclerae, and otosclerosis. Type III OI usually presents with early onset scoliosis, triangular facies, frontal bossing, basilar invagination, and extremely short stature. These types usually warrant pinning of both femurs prophylactically or as treatment for pathologic fractures. Type XI, on the other hand, presents with bone dysplasia, ligamentous laxity, and scoliosis. Patients typically have normal stature and no frontal bossing.[2]

Patients with OI may present with scoliosis in 20 to 40% of cases. Bracing may play a role depending on the curve severity and age at diagnosis.[3] Surgery is indicated if the curve is greater than 40 to 50 degrees in skeletally mature patients or if the curve progresses significantly after skeletal maturity. In addition, iliopelvic fixation is warranted if there is more

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than 10 degrees of pelvic obliquity.[4] If left untreated, patients may face balance issues and restrictive lung disease.[5]

The bone quality of these patients is expected to be poor, making surgery and fixation difficult. Pre-operative bisphosphonates have been used to prevent implant failure and future fracture risk.[6] But bisphosphonates must be discontinued at least 6 months before spinal fusion surgery to allow the spinal fusion mass to mature. Bisphosphonates prevent de novo bone formation and remodeling and may interfere with the postoperative course.[6]

Since bone stock is poor, implants have poor purchase and can pull out when using regular non-fenestrated pedicle screws. While bone cement has been used to augmented fenestrated screws, cement extravasation and cement emboli are still dreaded complications, which occur 12.7% and 5.4% of the time respectively.[7] Cement augmentation at the tips of non-fenestrated screws may provide subjective improvements in pain, but does not significantly improve postoperative activity levels.[8]

Given the absence of higher-level studies on managing scoliosis in patients with OI, best practices for preoperative treatment and implant fixation are still unclear.

This study aimed to evaluate the surgical management and outcomes of scoliosis in patients with OI using posterior spinal fusion with non-fenestrated pedicle screws: the achievable amount of correction, subjective improvements, and complications (if any).

CASE

Two patients with scoliosis associated with OI were treated in a tertiary government hospital. Patient A was a woman in her 20s diagnosed with OI type III because of the following features: (-) dentinogenesis imperfecta, (+) short stature, thoracolumbar kyphosis and scoliosis with pelvic obliquity >10 degrees, and frontal bossing (Figure 1). Birth and maternal, family, and social histories were all unremarkable. She first consulted at eight years old for extremity fractures and thoracolumbar spine asymmetry. She started bracing and bisphosphonate treatment (Pamidronate) with monthly infusions for the first six months, every other month for the next six months, then twice a month for the next two years.

Extremity fractures were initially immobilized conservatively but were eventually treated surgically because of fracture recurrences. She underwent pin fixation of the left femur in 2008, pin fixation of the right femur in 2010, and revision of the latter applied implant in the same year (Figures 2 and 3). After she had reached skeletal maturity, scoliotic curves progressed to greater than 40 degrees and was ambulatory only with a cane. Surgery was indicated to correct her spine deformity. Preoperative dual-energy x-ray absorptiometry (DEXA) revealed osteopenia (Table 1). She was referred to the endocrinology service, and she started Alendronate 70 mg/tab once a week for three months, with no repeat DEXA warranted. The surgery was postponed to approximately six months after completion of Alendronate treatment. The latest curve measurements before surgery were a 30-degree proximal thoracic curve, a 58-degree thoracic curve, and a 38-degree thoracolumbar/lumbar curve (Figure 4).

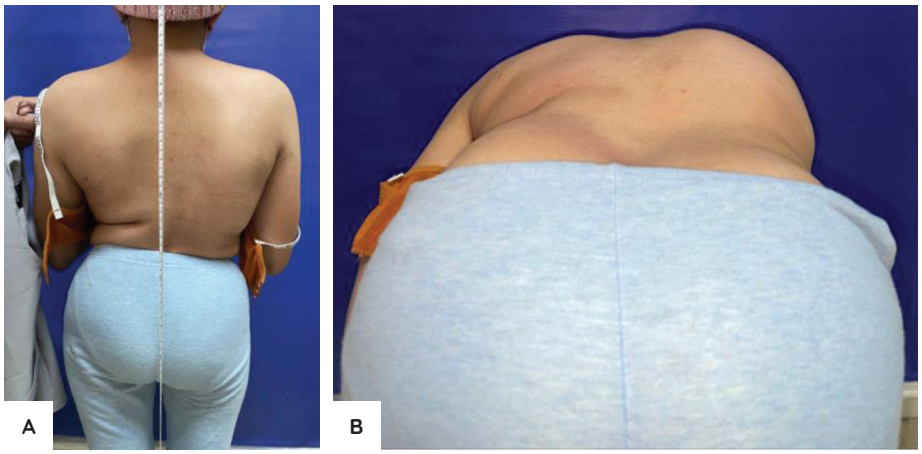

Figure 1. ***Patient A clinical pictures***. Standing gross pictures of patient with pelvic obliquity and grossly scoliotic curve more focused on the lumbar spine **(A)**. Forward bending test outlining direction of curves on both thoracic and lumbar sections of the spine **(B)**.

Table 1. Dual energy x-ray absorptiometry readings post treatment (prior to surgery)

	Patient A			Patient B		
	BMD (g/cm²)	T- score	Z- score	BMD (g/cm²)	T- score	Z- score
Lumbar spine (L1–L2)	0.877	-1.6	-1.6	1.164	1.2	1.2
Lumbar spine (L2–L4)	0.857	-1.9	-1.9	1.276	2.0	2.0
Right femoral neck	0.764	-1.4	-1.4	1.481	4.5	4.6
Left femoral neck	0.620	-2.6	-2.6	1.452	4.2	4.3

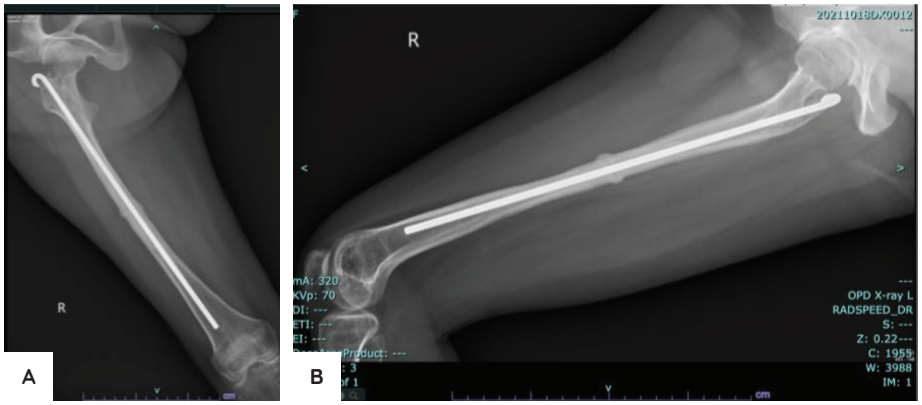

Figure 2. ***Right femur xrays (with fixation)***. Anteroposterior x-ray of the right femur showing healed pathologic fracture and adequately inserted implant **(A)**. Lateral x-ray of the right femur showing healed pathologic fracture and adequately inserted implant **(B)**.

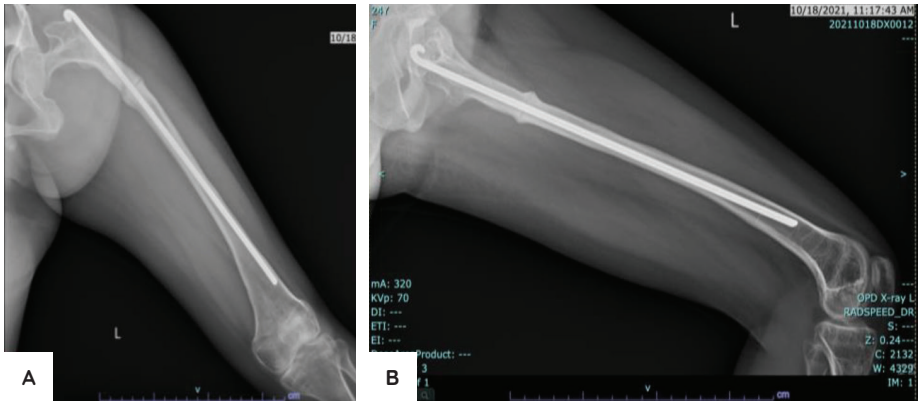

Figure 3. ***Left femur xrays (with fixation)***. Anteroposterior x-ray of the left femur showing healed pathologic fracture and adequately inserted implant **(A)**. Lateral x-ray of the left femur showing healed pathologic fracture and adequately inserted implant **(B)**.

Patient B was a woman in her early 30s diagnosed with OI type XI because of the following features: (+) scoliosis, (+) ligamentous laxity, pelvic obliquity <10 degrees, normal stature, (-) frontal bossing (Figure 5). She first consulted at 13 years of age and was advised bracing. She had recurrent lower extremity fractures, which were all managed conservatively with immobilization. There were no problems encountered with fracture healing. Pamidronate infusion was also started, infusing 1 mg/kg monthly for the first six months, then every other month for the next six months, then twice a month until three years were completed. Her curves also progressed after reaching skeletal maturity, prompting operative management.

Preoperative dual-energy x-ray absorptiometry (DEXA) showed normal findings (Table 1). The latest curve measurements before operation were: a 54-degree thoracic curve and a 64-degree thoracolumbar/lumbar curve (Figure 6).

For patient A, instrumentation was done from T6 to L5 with non-fenestrated pedicle screws and bilateral iliac screw fixation due to the noted pelvic obliquity. No cement augmentation was done. We used 15 monoaxial reduction-type and 2 polyaxial screws; the latter were chosen to facilitate rod application on the bilateral iliac screws. Screw sizes ranged from 5.0 to 5.5 mm in diameter and 35 to 40 mm in length. Brainlab navigation was used to verify screw trajectory before insertion. Full navigation was utilized in all steps for pedicle screw insertion in the T6 vertebra. We performed Smith-Petersen osteotomies on the apices of both curves, namely at T9 and L3. The curves were corrected via unilateral rod application and derotation. The contralateral rods were then applied, and the constructs tightened. To further improve correction, on the apices, the convex sides were compressed and the concave sides were distracted. Decortication was done, cross-links applied for rotational stability, and a bone graft was applied before closure. Total operative time was six hours, and total blood loss was estimated to be 1200 mL with three units of packed red blood cells (pRBC) transfused. Intraoperatively, the screws did not pull out. Postoperatively, there were no neurologic changes. Once discharged, the patient and her family decided to go to their hometown for recovery. We could not obtain immediate follow-up standing radiographs for two months, six months, the one year postoperatively (Figure 7, Table 2). Despite the loss of correction on subsequent follow-ups (2.1% loss of thoracic correction and 10.6% loss of lumbar correction), the curves did not revert to pre-operative measurements, and no other surgical complications were encountered, such as surgical site infection, implant failure, junctional kyphosis, or pseudoarthrosis. The patient became ambulatory without

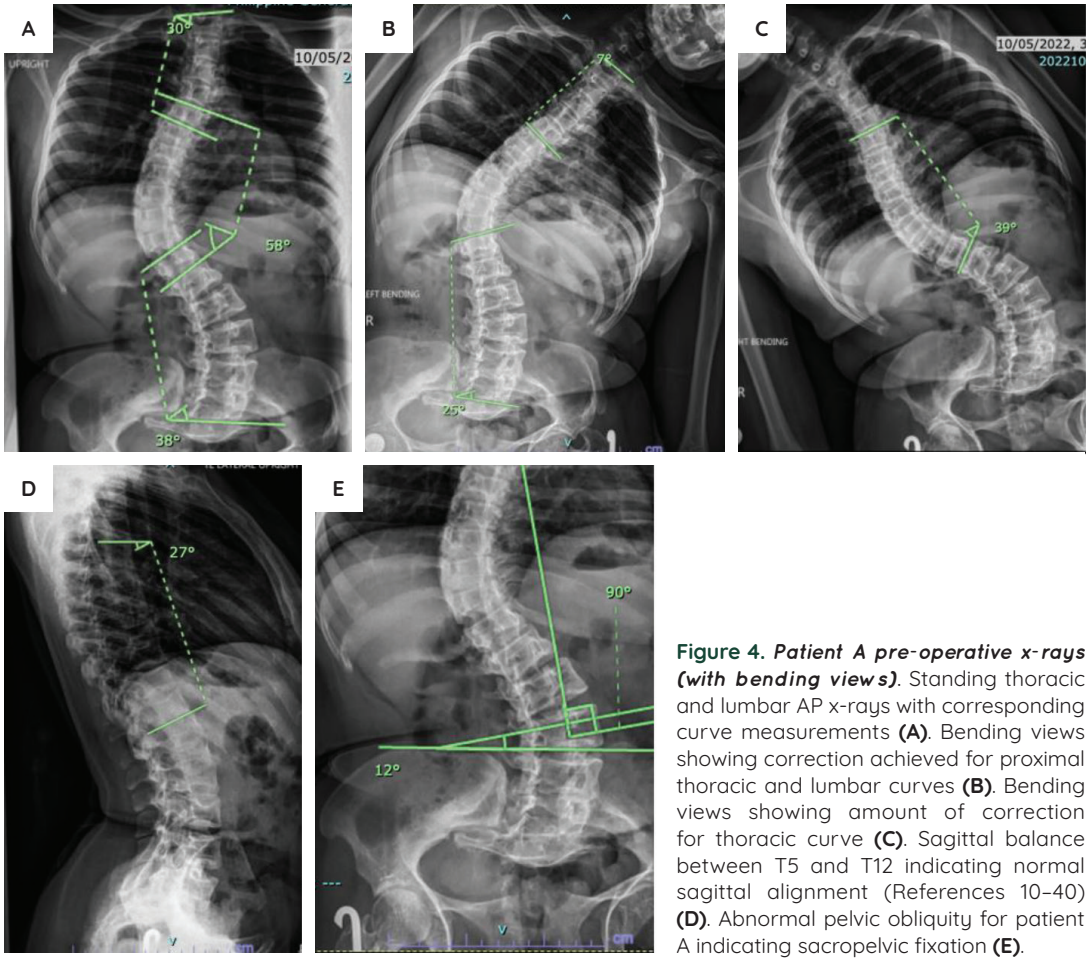

Figure 4. ***Patient A pre-operative x-rays (with bending views)***. Standing thoracic and lumbar AP x-rays with corresponding curve measurements **(A)**. Bending views showing correction achieved for proximal thoracic and lumbar curves **(B)**. Bending views showing amount of correction for thoracic curve **(C)**. Sagittal balance between T5 and T12 indicating normal sagittal alignment (References 10–40) **(D)**. Abnormal pelvic obliquity for patient A indicating sacropelvic fixation **(E)**.

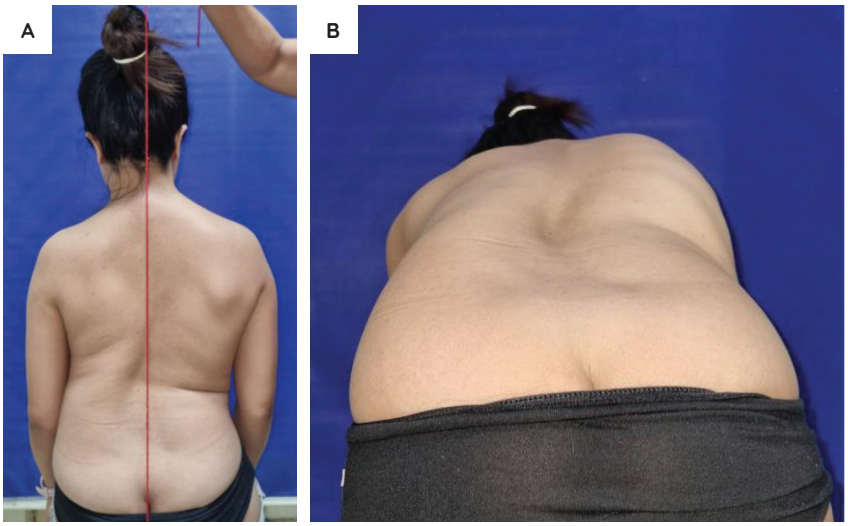

Figure 5. ***Patient B clinical pictures***. Standing clinical picture for patient B showing the central sacral vertical line in relation to the clinical curve **(A)**. Forward bending test outlining direction of curves on both thoracic and lumbar sections of the spine **(B)**.

any assistive device. The final corrections achieved were 31.9% and 54.5% for thoracic and lumbar curves, respectively. Visual analog scale scores were 9/10 immediately postoperatively, 5/10 at six months, and 1/10 at one year. No repeat DEXA scans were warranted. She was able to resume baking and selling pastries.

Patient B underwent instrumentation from T5 to S1 using non-fenestrated pedicle screws without cement augmentation. We used 16 screws, all 5.5 mm in diameter and 40 mm in length, six of which were polyaxial. Pedicle screw tracts were created freehand, and Brainlab navigation was used to verify screw trajectory before insertion. We performed a Smith-

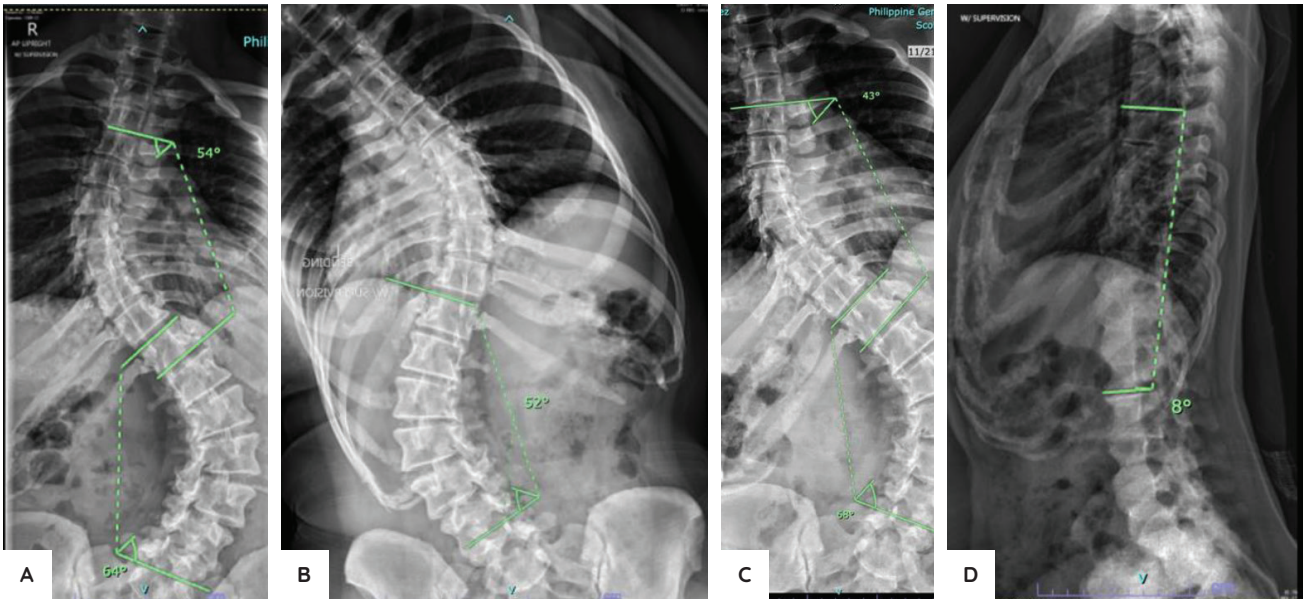

Figure 6. ***Patient B pre-operative x-rays (with bending views)***. Standing thoracic and lumbar AP x-rays showing curve measurements for thoracic and lumbar curves **(A)**. Bending views showing amount of correction achieved for the lumbar curve **(B)**. Bending views showing amount of correction achieved for the thoracic curve **(C)**. Lateral x-ray showing below-normal sagittal alignment (Reference: 10–40 degrees) indicating a hypokyphotic spine **(D)**.

Petersen osteotomy on L3 (apex), followed by derotation. The contralateral rod was inserted and fixed. To further improve correction, at the apex, the convex sides were compressed and the concave sides were distracted. Decortication was done, cross-links applied for rotational stability, and bone graft was applied before closure. The surgical time was 5 hours and 30 minutes, with blood loss estimated at 1150 ml. Intraoperatively, the screws did not pull out. Postoperatively, there were no neurologic changes. Serial radiographs showed a loss of correction of 2.6% for the thoracic and 6.3 % for the lumbar (Figure 8, Table 3). There were no noted complications such as surgical site infection, implant failure, junctional kyphosis, or pseudoarthrosis. Final corrections achieved were 33.3% and 37.5% for the thoracic and lumbar curves, respectively. Pain score (numerical rating scale) was 6/10 at two months, 5/10 at three months, 3/10 at six months, and 1/10 on the last follow-up.

As described, both patients had unremarkable post-operative courses and resumed normal activities beyond six months postoperatively. This was evident even before the patients' one-year follow-ups. Hardware failure or junctional kyphosis did not occur despite loss of correction. Both patients were also ambulatory without any assistive devices. Both patients signed journal-provided consent forms before manuscript and protocol writing.

DISCUSSION

Osteogenesis imperfecta (OI) patients with frequent fractures, episodes of pain, and poor bone mineral density are indicated to undergo bisphosphonate, whether intravenously (e.g., Pamidronate) or orally (e.g., Alendronate).[8]

Intravenous bisphosphonates can be given for years, beginning once monthly, followed by twice monthly.[1] Oral bisphosphonates may also be given to adult patients with OI when indicated. There are no significant differences when comparing oral or intravenous bisphosphonate treatments in terms of fracture frequency and bone mineral density.[9]

With the progression of the main curves of both patients beyond 40 degrees, surgical management was indicated.[10,11] Upper instrumented vertebrae of both patients were selected based on the recommendation to fuse one vertebra above the endpoints of the Cobb measurement (end vertebra). It was T6 for Patient A, and T5 for patient B. Lower instrumentation included the pelvis for patient A due to pelvic obliquity (Figure 4), while the lower instrumented vertebra was S1 for patient B, since it was the vertebra nearest where the central sacral vertical line (CSVL) intersected or was medial to the pedicle outline.[11]

Given the patients' poor bone quality, cement augmentation was a point of contention. While cement augmentation provides higher pull-out strength, cement extravasation and cement emboli can still occur.[6] Adding cement to fixation with non-fenestrated screws yields subjective improvements in pain, but no significant differences in terms of curve correction and fusion rates in OI patients.[7]

With our choice of non-fenestrated screws without cement, we achieved 31.9% thoracic and 54.6% lumbar correction for patient A, and 33.3% thoracic and 37.5% lumbar correction for patient B (Tables 2 and 3). Possible complications when treating scoliosis in OI patients are hardware failure, surgical site infection, pseudoarthrosis, and proximal junctional

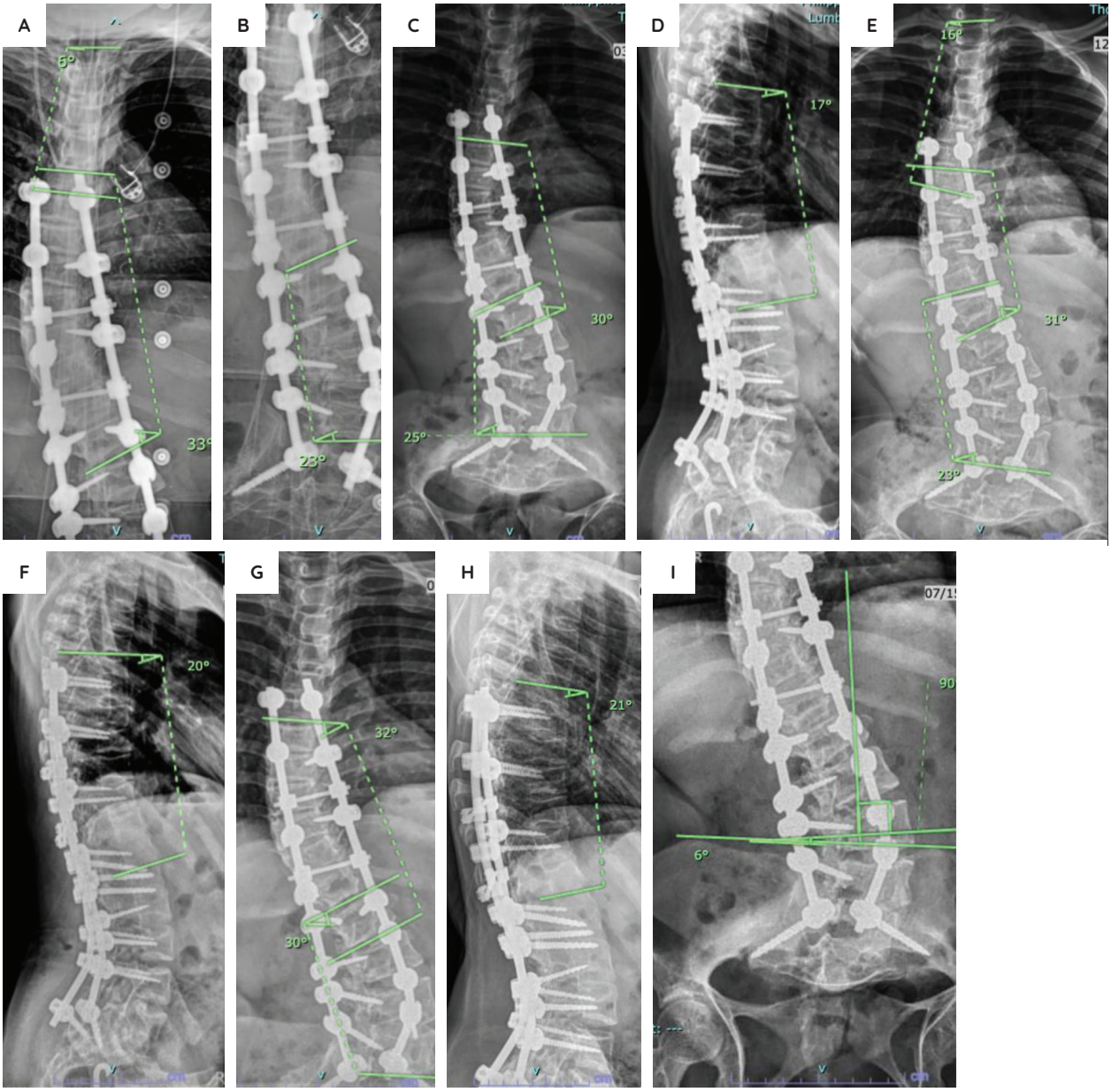

Figure 7. ***Patient A postoperative radiographs***. Postoperative AP xrays of patient A **(A and B)**. Standing AP and lateral xrays of Patient A two months postoperatively, with intact fixation and no screw pullouts **(C and D)**. Standing AP and lateral xrays of Patient A six months postoperatively showing loss of correction of the thoracic curve of 1 degree and kyphosis by 3 degrees but the latter still within normal limits (10–40 degrees) **(E and F)**. Standing AP and lateral xrays of Patient A one year post-operatively showing total loss of correction of the thoracic curve of 2 degrees and kyphosis by 1 degree but the latter still within normal limits **(G and H)**. Signs of fusion are also seen. Measurement of pelvic obliquity one year postoperatively within normal limits (<10 degrees) **(I)**.

Table 2. Pre-operative and postoperative curve measurements for patient A

	Standing	Bending	Predicted	Post-op standing				Correction
				2 weeks	2 months	6 months	12 months	
Thoracic (degrees)	47	7	85.1%	31 (34%)	30	31	32	31.9%
Thoracolumbar / lumbar (degrees)	66	25	62.1%	23 (65.1%)	25	23	30	54.5%
Sagittal alignment (T5–T12) (degrees)	17			N/A (Prone Xray)	17	20	21	

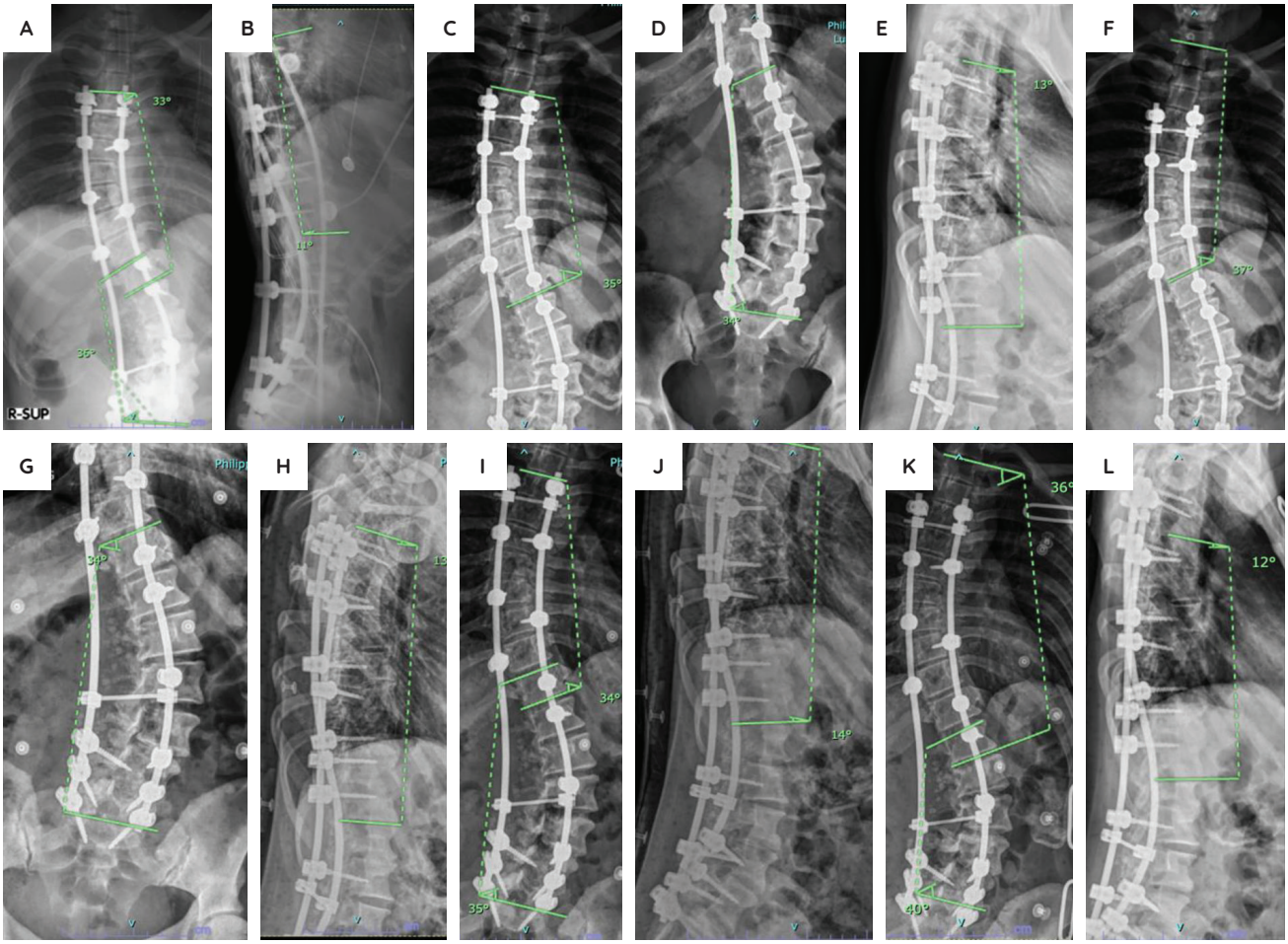

Figure 8. ***Patient B postoperative radiographs***. Standing AP and lateral xrays of Patient B two weeks postoperatively, with intact fixation and no screw pullouts **(A and B)**. Standing AP and lateral xraysof Patient B one month postoperatively showing loss of correction of the thoracic curve of proximal thoracic curve by 2 degrees, maintained thoracolumbar/lumbar curve, and an increase of kyphosis by 2 degrees, but still within normal limits (10–40 degrees) **(C, D, E)**. Standing AP and lateral xrays of Patient B three months postoperatively showing loss of correction of the thoracic curve of proximal thoracic curve by 4 degrees, maintained thoracolumbar/lumbar curve, and kyphosis within normal limits **(F, G, H)**. Standing AP and lateral xrays of Patient B six months postoperatively maintained correction of the proximal thoracic curve, thoracolumbar/lumbar curve, and kyphosis **(I and J)**. Standing AP and lateral xrays of Patient B 12 months postoperatively showing loss of correction of the proximal thoracic curve by 3 degrees and thoracolumbar/lumbar curve by 4 degrees, and maintained kyphosis **(K and L)**.

Table 3. Pre-operative and postoperative curve measurements for patient B

	Standing	Bending	Predicted	Post-op (2 weeks)	Post-op standing				Correction
					1 month	3 months	6 months	12 months	
Thoracic (degrees)	54	43	20.3%	33 (38.9%)	35	37	34	36	33.3%
Thoracolumbar / lumbar (degrees)	64	52	18.8%	36 (43.8%)	34	34	35	40	37.5%
Sagittal alignment (T5–T12) (degrees)	8			11	13	13	14	12	

kyphosis.[11,12] Fortunately, these didn't occur in our patients through serial radiographic monitoring at 2 weeks, 1 month, 3 months, 6 months, 9 months, 1 year, and 2 years postoperatively. Despite the initial loss of correction, fusion was achieved for both cases. None of the curves returned to the pre-operative measurements and sagittal balance, as evidenced by kyphotic measurements.

Mobilization was initiated on the second postoperative day. Return to work was restricted for both patients until the 6th month postoperatively.[12]

With no noted complications at their one year follow-up, both patients resumed their normal activities. Patient A resumed her part-time job as a baker while Patient B resumed work as a digital media manager.

CONCLUSION

We successfully treated two patients with osteogenesis imperfecta with associated scoliosis using non-fenestrated pedicle screws without cement augmentation. Instrumented levels were chosen based on the end vertebrae, the major and structural curves, and the presence of pelvic obliquity. However, our data cannot justify any conclusion regarding the use of fenestrated screws or cement augmentation. Loss of correction and screw pull-outs should subsequently be monitored. Further studies on different fixations need to be conducted for patients with osteogenesis imperfecta and scoliosis.

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ETHICAL CONSIDERATION

Patient consent forms were obtained before manuscript submission.

STATEMENT OF AUTHORSHIP

All authors certified fulfillment of ICMJE authorship criteria.

CREDIT AUTHOR STATEMENT

BLAC: Methodology, Formal analysis, Investigation, Resources, Data Curation, Writing – original draft preparation, Writing – review and editing, Visualization; **AGH:** Investigation, Project administration; **SAG:** Conceptualization, Writing – review and editing, Supervision, **AKGA:** Validation, Writing – review and editing, Supervision

AUTHOR DISCLOSURE

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

No datasets were generated or analyzed for this research.

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None.

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