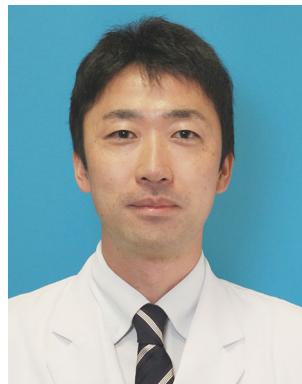


# Scoliosis correction surgery for patients with McCune–Albright syndrome using pedicle screws: a report of two cases with different characteristics and a review of the literature

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## Abstract



**Purpose** Scoliosis can occur secondary to McCune–Albright syndrome (MAS); it can be progressive and sometimes requires surgical treatment. It is still unclear if pedicle screw (PS) fixation in these patients with poor bone quality can be considered an effective treatment for scoliosis. The purpose of this study is to report two MAS patients with spinal fibrous dysplasia (FD) who underwent scoliosis surgeries with the PS system.

**Methods** Case 1: a 12-year-old girl. Standing posteroanterior radiographs revealed a 58° right curve from T7 to L2. Computed tomography (CT) showed small areas of FD

throughout the spine. A posterior spinal arthrodesis from T4 to L3 using PS fixation was performed with a CT-based navigation system. Case 2: a 26-year-old woman. Radiographs in the standing position revealed a right 87° curve from T8 to L2 and a 55° kyphosis from T8 to T12. CT images showed multiple areas of severe spinal FD causing angular deformity and collapse of vertebral bodies. The patient underwent posterior spinal arthrodesis from T8 to her pelvis using a CT-based navigation system for PS fixation.

**Results** Superior scoliosis corrections were obtained using PS instrumentation, attaining complete bony union in both cases without major complications. However, Case 2 had some technical difficulties in treating due to the multiple large FD lesions.

**Conclusions** PS fixation can be considered an effective treatment for correcting scoliosis and maintaining the correction at follow-up in MAS patients with poor bone quality. However, great care must be taken when performing correction and follow-up.

**Keywords** McCune–Albright syndrome · Polyostotic fibrous dysplasia of bone · Spinal fibrous dysplasia · Scoliosis correction surgery · Pedicle screw fixation

## Case presentation and diagnostic imaging section

### Case 1

A 12-year-old girl was diagnosed with McCune–Albright syndrome (MAS). Her history included precocious puberty, pathological fracture of the right femur, and hyperthyroidism. She was the only MAS patient in her family. One year prior to presentation, she developed a rib hump and

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her Cobb angle was  $20^\circ$  from T7 to L2. She could not wear a brace due to mental retardation. She was referred to our institute because of progressive scoliosis. Although her scoliosis caused her to have poor balance while sitting, her neurological examination at her first visit was normal. Standing posteroanterior radiographs revealed a  $58^\circ$  right curve from T7 to L2, with Risser grade 4. Computed tomography (CT) showed small areas of fibrous dysplasia of bone (FD) throughout the spine (Fig. 1).

#### Case 2

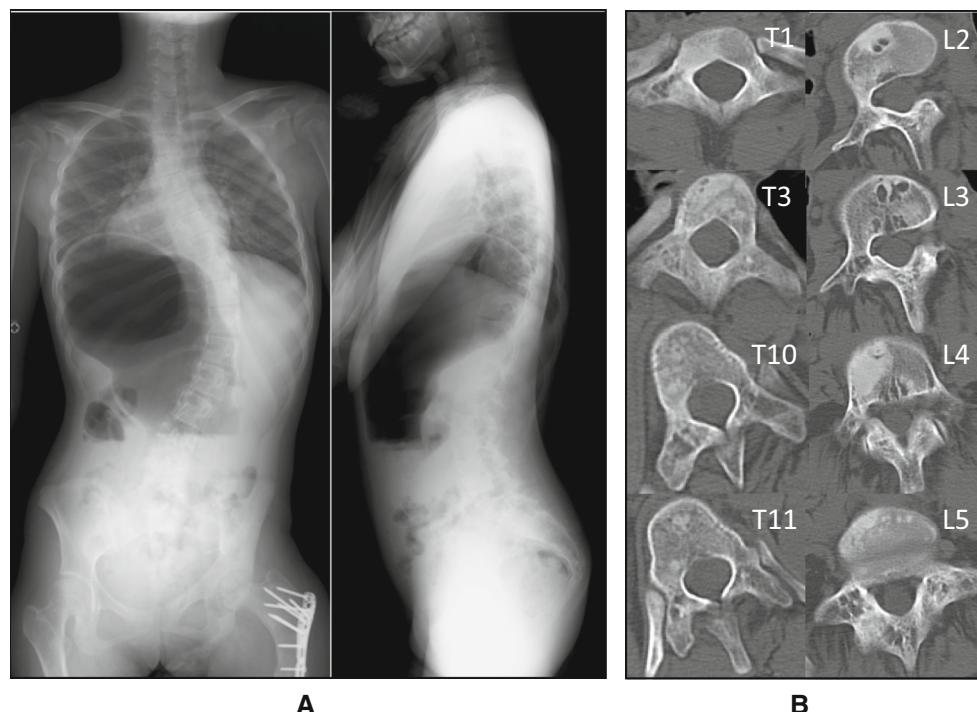
A 26-year-old woman was diagnosed with MAS. She had a history of hyperthyroidism and bilateral pathological fractures of her femurs. She was the only patient with MAS in her family. She was diagnosed with the disease at 18 years of age and had worn a brace for 2 years. Radiographs at the age of 23 years showed a  $59^\circ$  scoliosis from T8 to L2. She was referred to our institute because of progression of the abnormal Cobb angle. She complained of slight lower back pain and a negative body image. Neurological examination was intact. Radiographs in the standing position revealed a right  $87^\circ$  curve from T8 to L2 and a  $55^\circ$  kyphosis from T8 to T12. CT images showed multiple areas of severe spinal FD causing angular deformity and collapse of vertebral bodies, as well as a wide area of iliac FD (Fig. 2).

#### Historical review of the condition and diagnosis

MAS is defined by the clinical triad of polyostotic FD, café-au-lait skin spots, and various endocrinopathies [1–3]. Scoliosis can occur secondary to MAS; it can be progressive and sometimes requires surgical treatment [4]. There are nine case reports in the literature on surgery for scoliosis in polyostotic FD with or without MAS [4–8]. We performed corrective surgeries for two MAS patients with different characteristics using the pedicle screw (PS) system. One patient was an adolescent with a small spinal FD lesion and an unstructural thoracic curve. The other patient was in her mid-20s and had severe spinal FD lesions and a structural kyphoscoliosis.

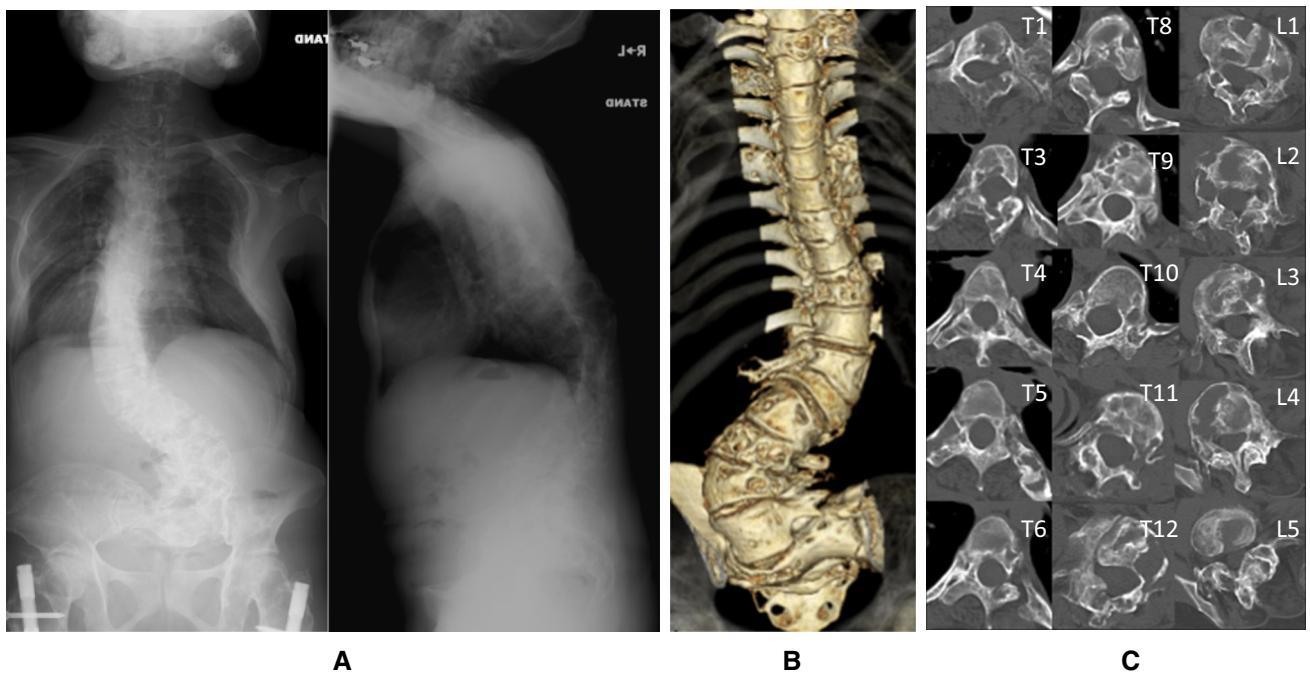
#### Rationale for treatment and evidence-based literature

Patients with polyostotic FD have poor bone quality, but they have normal bone healing [9]. Several authors have shown satisfactory rates of bony union in scoliosis patients with spinal FD who were treated by surgery [4–8]. However, the correction rate of these patients was insufficient (Table 1). Guille described a case report of MAS stating that severe spinal FD lesions were too soft to use some types of instrumentation (e.g., Harrington rod and wiring), and he performed in situ posterior spinal arthrodesis without instrumentation [5].



**Fig. 1** **a** Preoperative radiographs showing a  $58^\circ$  major curve from T7 to L2. The Risser sign is  $4^\circ$ . Sagittal spinal parameters in the natural standing posture: SVA 4.6 cm, TK  $24^\circ$ , LL  $57^\circ$ . **b** Axial CT

showing FD lesions characterized by ground-glass opacities, a variable pattern of sclerosis and medullary expansion in multiple vertebrae



**Fig. 2** **a** Preoperative radiographs showing an 87° major curve from T8 to L2 and kyphosis from T8 to T12 measuring 55°. In the natural standing posture: SVA 9.7 cm, TK 56°, LL 59°. **b** 3D CT showing

It is still unclear whether a PS system is truly effective for correcting scoliosis and maintaining the correction at follow-up in spinal FD patients. The first report of scoliosis surgery for a spinal FD patient was described by Shikata in 1992. He used Luque SSI and sublaminar wiring [8]. In recent years, thoracic PS systems have become common in scoliosis surgeries [10, 11]. However, there are few reports on corrective surgery with PS instrumentation for polyostotic FD or MAS patients.

### Procedure and procedure imaging section

#### Case 1

A posterior spinal arthrodesis from T4 to L3 using PS fixation was performed with a CT-based navigation system. A large amount of corticocancellous iliac bone graft and a small amount of beta-tricalcium phosphate particles (b-TCP) were applied for arthrodesis. The scoliosis could be adequately corrected because the PS holding force was satisfactory, and the curve was unstructural. After surgery, the Cobb angle was 0° from T7 to L2. There were no surgical complications such as PS perforations or nerve injury. A brace was not applied postoperatively. At 3-year follow-up, there was no correction loss and crankshaft phenomenon. Radiographs showed a solid fusion mass (Fig. 3).

varying degrees of angular deformity and collapse of vertebral bodies. **c** Axial CT showing FD lesions that are spreading over a wider range compared to Case 1

#### Case 2

Surgery was performed to correct the kyphoscoliosis and prevent progression. The patient underwent posterior spinal arthrodesis from T8 to her pelvis using a CT-based navigation system for PS fixation. A large amount of b-TCP was placed for arthrodesis because the majority of corticocancellous iliac bone was replaced with FD. PSs could not be inserted into some vertebrae due to narrow pedicles, and there were many loose screws during surgery because of the spinal FD lesions. It was not possible to adequately reduce the kyphosis for fear of PS failure. After surgery, the scoliosis was corrected to 28° from T8 to L2, but kyphosis was 52° from T8 to T12, nearly equal to the preoperative amount. Postoperatively, the patient wore a hard brace for 6 months. Three years after surgery, scoliosis and kyphosis were 30° and 54°, respectively, with no correction loss. There was no obvious PS loosening, and there was complete bony union (Fig. 4). Although mild proximal junctional kyphosis (PJK) appeared, and the sagittal vertical axis (SVA) increased at the latest follow-up, the patient was doing well.

#### Outcome and follow-up

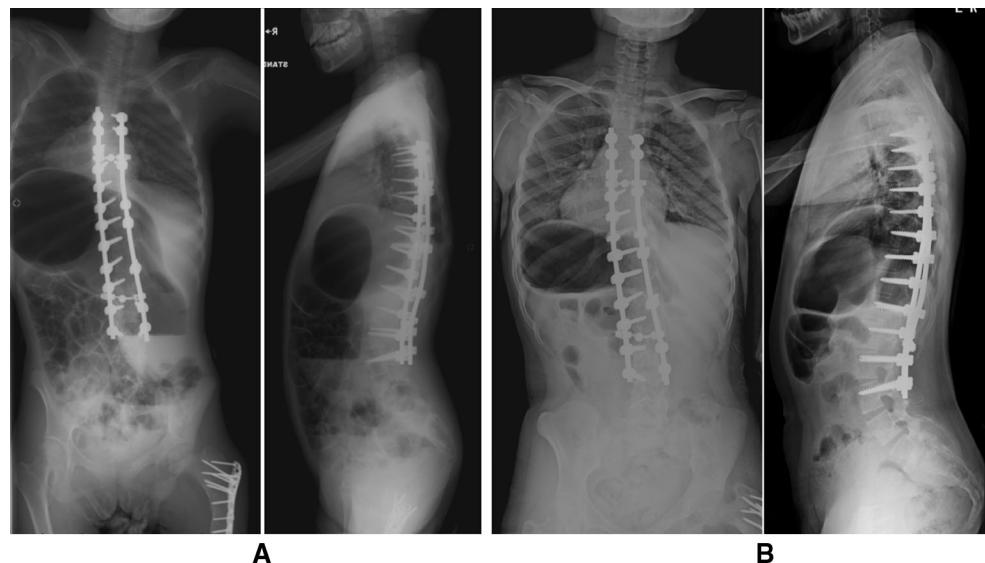
Corrective surgery was successfully performed using PS instrumentation in the present two MAS patients with different characteristics. The first case was an adolescent

**Table 1** Data from published reports and this study concerning surgery of scoliosis with fibrous dysplasia

References	Age/gender	Diagnosis	SFD	Scoliosis	Pre-op Angle (°)	Post-op Angle (°)	Correction rate (%)	Operative procedure	Complications	Bone union	Follow-up period (months)
Shikata et al. [9]	34/F	PFD	+	T6-L2	75	38	49	Luque SSI + BG	None	+	12
Guille et al. [4]	19/M	PFD	+	T2-T9	80	80	0	No instrumentation + BG	Excessive bleeding	+	60
5/M	MAS	ND	T5-T12	45	45	0	No instrumentation + BG	None	+	33	
Janus et al. [5]	12/M	PFD	+	T4-T10	26	13	50	CD instrumentation + BG	Pseudoarthrosis	48	
Leet et al. [6]*								instrumentation + BG			
Mancini et al. [7]	11/F	MAS	-	T2-T6	40	ND	ND	ND	None	+	48
Present report (2015)	12/F	MAS	+	T7-L2	58	0	100	PS instrumentation + BG	None	+	36
	26/F	MAS	+	T8-L2	87	28	68	PS instrumentation + BG	None	+	36

*SFD* spinal fibrous dysplasia, *PFD* polyostotic fibrous dysplasia, *SSI* segmental spinal instrumentation, *ND* no description, *BG* bone graft, *MAS* McCune-Albright syndrome, *CD* Cotrel-Dubousset

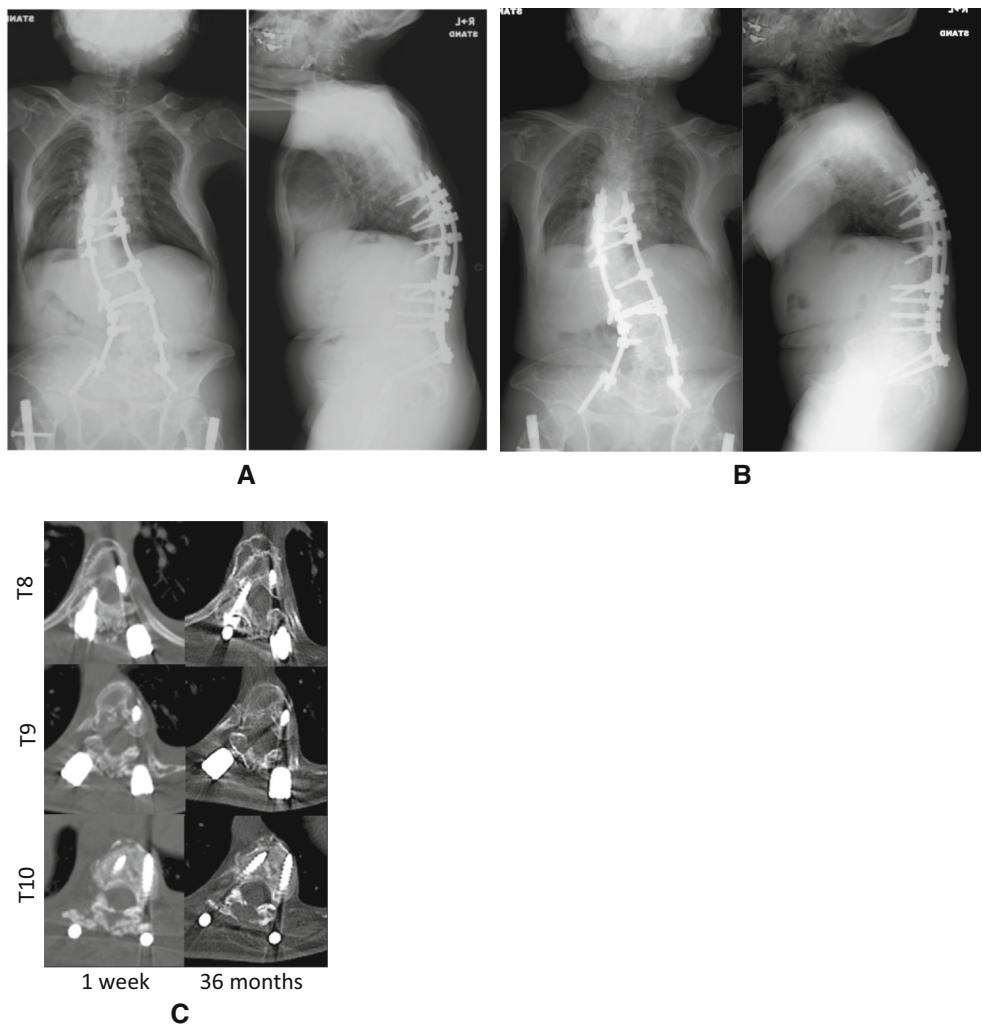
\* Four cases with spinal FD had posterior arthrodesis with devices, but there were no detailed descriptions



**Fig. 3** **a** Immediate postoperative radiographs. The major curve from T7 to L2 measures 0°. In the natural standing posture: SVA 4.0 cm, TK 19°, LL 52°. **b** Radiographs at 35 months after surgery showing

no correction loss and a solid fusion mass. Sagittal spinal parameters remain unchanged from the immediate postoperative radiographs

**Fig. 4** **a** Immediate postoperative radiographs. The major curve from T7 to L2 measures 28°, and the kyphosis from T8 to T12 measures 52°. In the natural standing posture: SVA 8.9 cm, TK 48°, LL 42°. **b** Radiographs at 36 months after surgery showing a 30° curve from T8 to L2 and 54° kyphosis from T8 to T12. Proximal junctional kyphosis (PJK) has appeared. In the natural standing posture: SVA 10.9 cm, TK 50°, LL 42°. **c** Axial CT showing pedicle screws inserted through FD lesions. There is no pedicle screw loosening



who had small spinal FD lesions. The surgery was not difficult, because the deformity was plastic. Strong PS fixation with adequate correction was established. In Case 2, there were some technical difficulties in treating due to the multiple large spinal FD lesions. The patient had a severe deformity, and the curve was strictly structural. Thoracic and lumbar PS insertion was difficult due to narrow pedicles and loose due to poor bone quality. Thoracic kyphosis was reduced in the prone position. The scoliosis correction rate was 68 %. Powerful correction was difficult due to poor bone quality. Some additional devices might be useful to prevent implant failure, such as polymethylmethacrylate screw augmentation and sublaminar wiring [12].

The postoperative courses of the two cases were satisfactory, as were the other types of surgeries previously described (Table 1) [4–8]. Superior scoliosis corrections were obtained using PS instrumentation, attaining complete bony union in both cases without major complications. Huge spinal FD lesions can affect the PS stability and cause implant failure or correction loss. In Case 2, a hard corset was effective for achieving bony union without further PS loosening despite the disadvantage of using autogenous bone graft. With respect to sagittal alignment, it is difficult to establish a strong correction relying only on spinal instrumentation due to bone fragility. An osteotomy and release could have been expected to correct sagittal alignment in Case 2. At the latest follow-up, SVA increased under the influence of PJK. In the long-term course, it is important to observe for sagittal imbalance.

**Conflict of interest** The authors declare that they have no conflict interests.

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