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Surgical Treatment for Scoliosis in Marfan Syndrome

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Study Design. Review of results of patients with Marfan syndrome treated with instrumented posterior fusion alone for scoliosis.

Objective. To analyze the results of surgical treatment for scoliosis in Marfan syndrome.

Summary of Background Data. Few studies have been reported in the literature on surgical treatment for scoliosis in Marfan syndrome, analyzing long-term results of posterior instrumented fusion.

Methods. Twenty-three patients with Marfan syndrome with a mean age of 17 years (range, 11–31 years) were treated surgically from 1982 to 1995 for scoliosis, using a posterior instrumented fusion alone (Harrington rod with sublaminar wires in the first 16 cases, and a more recent hybrid instrumentation in the remaining 7 cases). All of the patients received a long posterior instrumented fusion, including 12.3 levels on average (range, 9–17), extending the fusion area to vertebrae that were neutral and stable in both coronal and sagittal planes before surgery. Patients were analyzed as two different groups (Group 1 and Group 2) according to the different posterior instrumentations employed: Group 1 included 16 patients treated by the Harrington distraction rod technique with sublaminar wires, while Group 2 included 7 patients treated using more recent hybrid instrumentations. Presentation features, complications, and results were analyzed.

Results. At a minimum follow-up of 7 years (maximum, 18 years), all 23 patients were reviewed. The mean age was 26.8 years (range, 20–38 years). The average preoperative scoliosis value of 69.91° was initially corrected to 38.17°, averaged 40.89° 1 year after surgery, and was finally equal to 44.09° at the last follow-up. Differences in terms of scoliosis correction achieved with different instrumentations (Groups 1 and 2) did not reach statistical significance. In Group 2 patients, the percentage of postoperative correction was slightly lower (44.23%) than that of Group 1 (46.55%) but remained more stable at the last follow-up (40.97% *vs.* 36.38% of Group 1). There were 11 complications in 10 of the 23 patients (43.4%); two complications occurred in 1 patient. Intraoperatively, dural tears occurred in 2 cases (8.6%). Pseudarthrosis with instrumentation failure in 2 cases (8.6%) required revision surgery. Five (21.7%) distal hook dislodgements with moderate loss of scoliosis correction, 1 (4.3%) mild loss of correction without instrumentation failure, and 1 asymptomatic cervicothoracic junctional kyphosis. did not require surgery. All complications occurred among the 16 Group 1 patients, treated using the Harrington rod instrumentation with sublaminar wires.

Conclusions. These results seemed to demonstrate that a satisfactory stabilization of scoliosis can be achieved by posterior instrumentation alone in patients with Marfan syndrome. Instrumented posterior fusion should be extended to include vertebrae that are neutral and stable in both coronal and sagittal planes before surgery, in order to ensure stabilization of the deformity and reduce the risks of decompensation of the spine.

Key words: Marfan syndrome, scoliosis, surgical treatment. **Spine 2005;30:E597–E604**

Marfan syndrome (MFS) is one of the most common connective tissue disorders. A prevalence of about 0.01% has been reported in the general population.[1] This syndrome has been shown to be the result of an autosomal dominant mutation of the fibrillin-1 gene located on chromosome 15.[2,3] The fibrillin abnormality causes associated manifestations in different systems, such as the skeleton (scoliosis, tall stature, arachnodactyly, sternal deformities), the cardiovascular system (mitral valve prolapse, dilatation of the ascending aorta, dissecting aortic aneurysm), the ocular system (ectopia lentis, myopia, retinal detachment), the lung (spontaneous pneumothorax), the skin (striae and hernia), and the central nervous system (dural ectasia).[4,5] MFS can be misdiagnosed as Ehlers-Danlos syndrome,[6] homocystinuria,[7] congenital contractural arachnodactyly,[8–10] and other different conditions.[11,12] Intelligence is normal in patients with MFS, and in case of mental retardation other diseases must be suspected,[13] such as homocystinuria. Careful examination can avoid any error in diagnosis, according to the more recent Ghent criteria[4], and can permit the exclusion of patients with "forme-fruste" manifestations of MFS.[1,14–16]

Spinal deformities in MFS include progressive scoliosis[5,17] and thoracic lordosis with concomitant loss of lumbar lordosis,[17,18] and more rarely thoracolumbar kyphosis,[19] severe spondylolisthesis,[17,20,21] and cervical problems.[22–24] The most common disorder affecting the spine is scoliosis,[5,16,17,25] even though in 1896, when Marfan[26] first described the syndrome, he noticed that scoliosis might have been an associated finding. The scoliosis prevalence in MFS ranges from 30% to 100%, according to different authors.[5,15,16,25,27,28] The curve pattern resembles that of idiopathic scoliosis, although

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there is a higher prevalence of double thoracic and triple major curves in patients with MFS.[5] There is a higher propensity for curves to progress in MFS with an average rate of 10.2° per year,[29] presenting with an increasing rigidity.[13,16,17,30] Curves exceeding 40° to 50° were found to have a high likelihood to progression also after maturity.[31]

Bracing was often ineffective as a definitive method of treatment for scoliosis in MFS,[15–17,32,33] being successful in only 17% of patients.[34] Brace treatment represented the initial management in children with mild scoliosis,[16,17] while the definitive treatment was arthrodesis for most progressive curves of more than 40°.[17,27,56] Posterior fusion with instrumentation has been widely used,[16–18,25,27,32,35–40] but a high rate of major complications has been reported,[16–18,25,36,40] such as failure of instrumentation, pseudarthrosis, and postoperative progression of scoliosis, using Harrington rods alone or with sublaminar wires.

The purpose of this study was to verify the results of posterior instrumented fusion alone at a long-term follow-up in a group of 23 patients with MFS affected by scoliosis.

Materials and Methods

Patients presenting with MFS at our Department in the last 20 years for surgical treatment of scoliosis were considered for inclusion in this study. Inclusion criteria were: 1) confirmed diagnosis of MFS (according to the most recent Ghent criteria[4]); 2) scoliosis without concomitant thoracic hyperkyphosis; 3) surgical treatment with posterior instrumented fusion alone; and 4) minimum follow-up of 7 years.

Twenty-three patients (8 males and 15 females) with a mean age of 17 years (range, 11–31 years), treated from 1982 to 1995 (Table 1), were included in the study.

Medical records provided: age at surgery, preoperative treatment, surgical procedures performed, including blood loss, intraoperative and postoperative complications, and type and length of postoperative immobilization for all patients. Preoperative, postoperative, 1-year follow-up, and latest follow-up radiographs were studied to measure sagittal and coronal Cobb angles and calculate correction percentages as a percentage change from the preoperative measurements.

All 23 patients presented on radiographs vertebral bodies typically elongated vertically with the aspect of biconcave vertebrae. Before surgery, CT scan or MRI of the lumbar spine was performed on 15 of the most recently treated patients (65.2%). In all 15, vertebral scalloping was found in the lumbar region, as a form of erosion through the central aspect of the vertebral body, resulting from dural ectasia.

All of the patients had failed to respond to treatment with a brace or cast, which was applied at a mean of 7 years old and worn until the time of surgery, or until the end of growing age in the 5 patients older than 17 years. Of the 23 patients, 8 had a Type 1 scoliosis, three a Type 5, ten a Type 3 curve, and two a Type 4 curve pattern, according to the Lenke *et al*[41] classification for idiopathic scoliosis. For each patient with a double curve or a triple curve pattern, the two or three curves were considered separately; therefore, the total number of curves was 37. The mean curve magnitude was 69.91° (range, 30°–110°).

With regards to the sagittal profile, none of the 23 patients presented with concomitant thoracic hyperkyphosis. Eighteen patients had coexisting thoracic hypokyphosis with a mean angular value of 12° (range, 7°–14°). A thoracolumbar (T10–L2) kyphosis was present in 5 cases, with a mean average value of 29° (range, 21°–35°).

Risser stage at time of surgery was 0 in 1 case, 3 in 5 cases, 4 in 10 cases, and 5 in the remaining 7 patients. The mean vital capacity was 2,144 (range, 690–4,400), corresponding to 43% of the theoretical value.

All of the patients received a long posterior instrumented fusion, including 12.3 levels on average (range, 9–17 levels), extending the fusion area to vertebrae that was neutral and stable in both coronal and sagittal planes before surgery. The fusion was from stable zone to stable zone. Even in 8 patients with Type 1 curve, the fusion area included part of upper thoracic and entire thoracolumbar nonstructural curves, including distally L2 (1 case), L3 (6 cases), and L5 (1 case) (Table 1).

Mean time of surgery was 330 minutes (range, 180–450 minutes). Mean blood loss was 1,850 mL (range, 950–2,850 mL). In the last 7 patients, preoperative autologous blood donation and intraoperative red cell recovery were used successfully.

Patients were analyzed as two different groups, Group 1 and Group 2, according to the different posterior instrumentations employed.

Group 1. This group included 16 patients treated between 1982 and 1990 by the Harrington distraction rod technique with sublaminar wires,[39,42] applied at each level (10 on average) (Figures 1, 2). In 5 of these patients, a compression Harrington rod was also used, with 5 hooks on average. The posterior fusion instrumentation was extended to include 12.5 levels on average (range, 11–17 levels), cranially from T1 (1 case), T2 (2 cases), T3 (6 cases), T4 (6 cases), T5 (1 case), including, distally L2 (1 case), L3 (6 cases), L4 (6 cases), and L5 (3 cases). In all cases, the bone grafts were autologous, harvested from the right iliac crest. In all of these patients, a prolonged postoperative cast was used (worn for 8 months on average), which enabled walking about 7 days after surgery.

Group 2. This group included 7 patients treated between 1991 to 1995, using more recent hybrid instrumentations. In 2 patients, Cotrel-Dubousset instrumentation was used, and in the remaining 5 the Colorado[43] system, using hooks only in 1 case, pedicle screws only (13 altogether) in another patient, and hybrid construct with proximal hooks and distal pedicle screws (6–11) in the remaining 5 cases (Figure 3). The posterior fusion instrumentation was extended to include 12.0 levels on average (range, 9–14 levels), cranially, from T2 (1 case), T3 (3 cases), T4 (2 cases), T5 (1 case), including, distally, L3 (4 cases) and L4 (3 cases). The bone grafts used were homologous (femoral epiphysis harvested from our Bone Bank) in all of these cases. In the postoperative period, the patients resumed walking about 6 days after surgery, with a TLSO brace worn for 5 months on average.

Results

At a minimum follow-up of 7 years (maximum, 18 years; mean, 9.8 years), all 23 patients were personally reviewed with radiographs by an independent physician

Table 1. Summary of Patients

Case No.	Age (yr)	Scoliosis Curve	Fused Levels	Instrumentation	Scoliosis (°) Preop	Postop	1 Yr	Follow-up	Follow-up (yr)	Complications
1	17	T4–T11	T1–L5	Harrington + sublaminar wires	110	80	82	95	16	Hooks dislodgement
		T12–L5		Compression rod	65	45	50	56		Loss correction >10°
2	16	T6–T12	T4–L3	Harrington + sublaminar wires	60	40	40	40	8	Intraoperative dural tear
3	18	T11–L4	T4–L4	Harrington + sublaminar wires	70	33	35	40	12	Hook dislodgement
				Compression rod						Loss correction <10°
4	16	T4–T10	T3–L3	Harrington + sublaminar wires	75	45	46	50	8	No
		T11–L3			75	46	48	51		
5	16	T5–T12	T3–L3	Harrington + sublaminar wires	88	40	40	40	10	Intraoperative dural tear
6	17	T5–T11	T3–L3	Harrington + sublaminar wires	75	37	40	40	18	No
		T12–L3			55	39	40	40		
7	31	T4–T11	T2–L4	Harrington + sublaminar wires	95	70	70	80	7	Hook dislodgement
		T12–L4		Compression rod	95	66	76	80		Loss correction >10°
8	16	T5–T10	T4–L2	Harrington + sublaminar wires	65	30	35	35	7	No
9	17	T5–T11	T3–L5	Harrington + sublaminar wires	80	50	50	53	7	No
10	14	T5–T12	T4–L4	Harrington + sublaminar wires	90	41	50	50	11	Loss correction <10°
		L1–L4			74	45	45	45		Kyphosis C7–T4
11	19	T5–L1	T4–L3	Harrington + sublaminar wires	65	17	20	22	15	No
12	13	T6–T11	T5–L5	Harrington + sublaminar wires	50	22	30	32	15	Pseudoarthrosis (rod fracture)
		T12–L4		Compression rod	70	30	40	40		Revision surgery (5 yrs later)
13	16	T5–T9	T3–L4	Harrington + sublaminar wires	42	15	15	17	8	No
		T10–L3		Compression rod	58	34	37	39		
14	11	T5–L1	T3–L3	Harrington + sublaminar wires	70	15	20	32	9	Hook dislodgement
										Loss correction >10°
15	16	T5–T11	T4–L4	Harrington + sublaminar wires	80	35	40	50	8	Hook dislodgement
		T12–L4			70	30	38	42		Loss correction >10°
16	16	T5–T8	T2–L4	Harrington + sublaminar wires	30	20	22	20	11	Pseudoarthrosis (rod fracture)
		T9–T12			30	20	21	22		Revision surgery (8 yrs later)
		L1–L4			60	32	34	42		
17	22	T2–T6	T2–L4	CD (hooks)	30	21	21	22	10	No
		T7–T11			55	21	21	22		
		T12–L4			40	20	20	21		
18	16	T7–L3	T3–L4	Colorado hybrid (10 screws)	105	60	60	62	10	No
19	14	T4–T9	T4–L3	CD hybrid (8 screws)	65	39	39	40	8	No
		T10–L3			65	40	40	42		
20	14	T8–L2	T4–L3	Colorado hybrid (6 screws)	80	38	38	40	8	No
21	16	T5–T11	T5–L4	Colorado (screws)	90	45	45	45	7	No
		T12–L4			60	24	24	26		
22	25	T3–T11	T3–L3	Colorado hybrid (8 screws)	60	48	48	50	7	No
23	16	T4–T12	T3–L3	Colorado hybrid (11 screws)	60	44	45	46	7	No

not involved in the study. The mean age was 26.8 years (range, 20–38 years). One patient (4.3%) had been treated recently by cardio-surgeons. All 23 patients lead a practically normal life: 6 of them have sedentary jobs, and 5 are university students.

The average preoperative scoliosis value of 69.91° (range, 30°–110°) was initially corrected to 38.17° (range, 15°–80°), which averaged 40.89° (range, 15°–82°) 1 year after surgery, and was finally equal to 44.09° (range, 17°–95°) at the last follow-up (Table 2). The postoperative curve correction was 45.88% on average (range, 20%–78.57%), which fell to 41.9% (range, 20%–71.43%) 1 year after surgery, and was 37.69% on average at the last follow-up (range, 11.58%–66.15%).

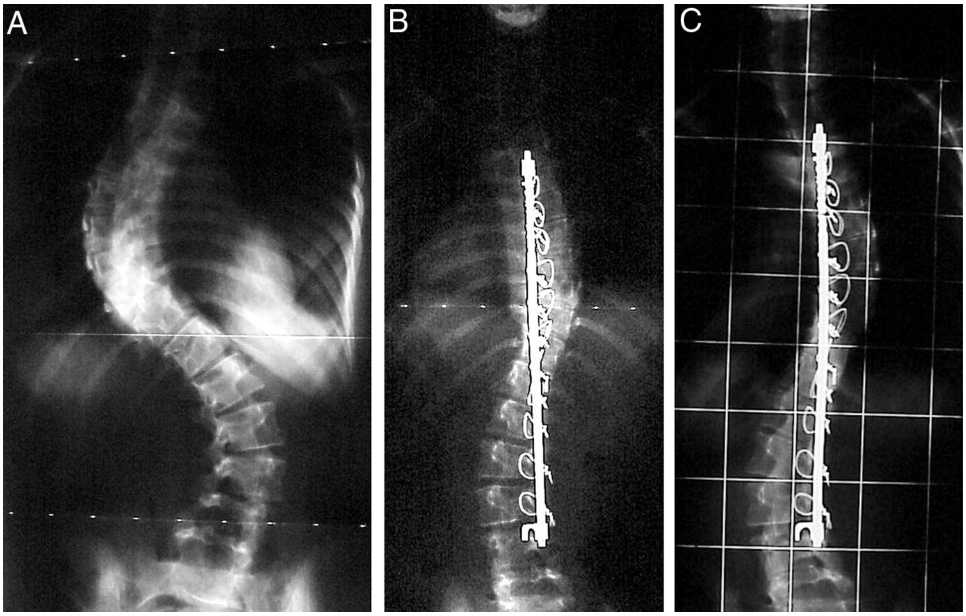

Figure 1. Case 15: female, 16 years old (**A**), treated with T4–L4 posterior instrumented fusion (Harrington rod with sublaminar wires), as evident in the postoperative radiographs (**B**). Eight years later, distal hook dislodgement with loss of correction more than 10° (**C**), which did not require surgery.

In Type 1 curves (8 patients), the average preoperative thoracic structural curve value of 68.5° was initially corrected to 35.5° and averaged 39.75° (40.9% of correction) at the last follow-up. In Type 3 curves (10 patients), the average preoperative thoracic and thoracolumbar/lumbar structural curve value of 72.95° was initially corrected to 34.6° and averaged 48.0° (32.3% of correction) at the last follow-up. In Type 4 curves (2 patients), the average preoperative structural curve value of 27.33° was initially corrected to 22.33° and averaged 24.83° at the last follow-up. In Type 5 curves (3 patients), the average preoperative structural curve value of 85.0° was initially corrected to 43.66° and averaged 47.33° at the last follow-up.

In Group 1 (Harrington with sublaminar wires), the average preoperative scoliosis value of 70.68° was initially corrected to 38.28°, equal to 46.55% of correction, averaged 41.72° (41.53% of correction) 1 year after surgery, and was equal to 45.72° (36.38% of correction) at the last follow-up (Table 3).

In Group 2 (segmental instrumentations), the average preoperative scoliosis value of 68° was initially corrected to 39.7°, equal to 44.23% of correction, averaged 40.7° (42.83% of correction) 1 year after surgery, and was equal to 41.9° (40.97% of correction) at the last follow-up (Table 4).

A two-tailed Student's *t* test was used to determine statistical significance in the differences between postop-

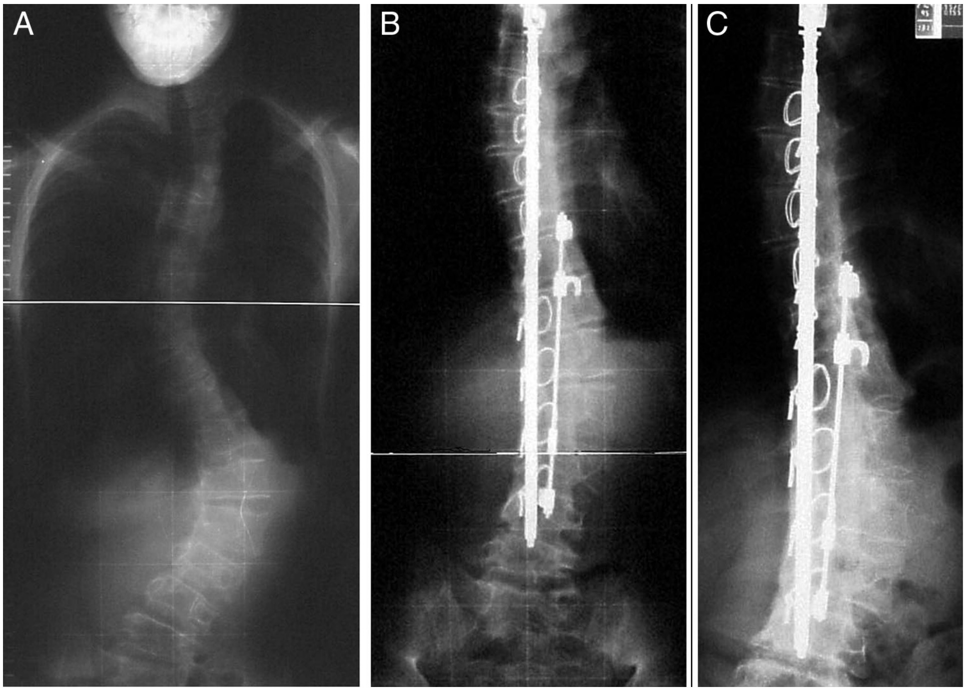

Figure 2. Case 3: male, 18 years old (**A**), treated with T4–L4 posterior instrumented fusion (Harrington compression-distractor rods with sublaminar wires), as evident in the postoperative radiograph (**B**). Twelve years later, distal hook dislodgement with loss of correction less than 10° (**C**), which did not require surgery.

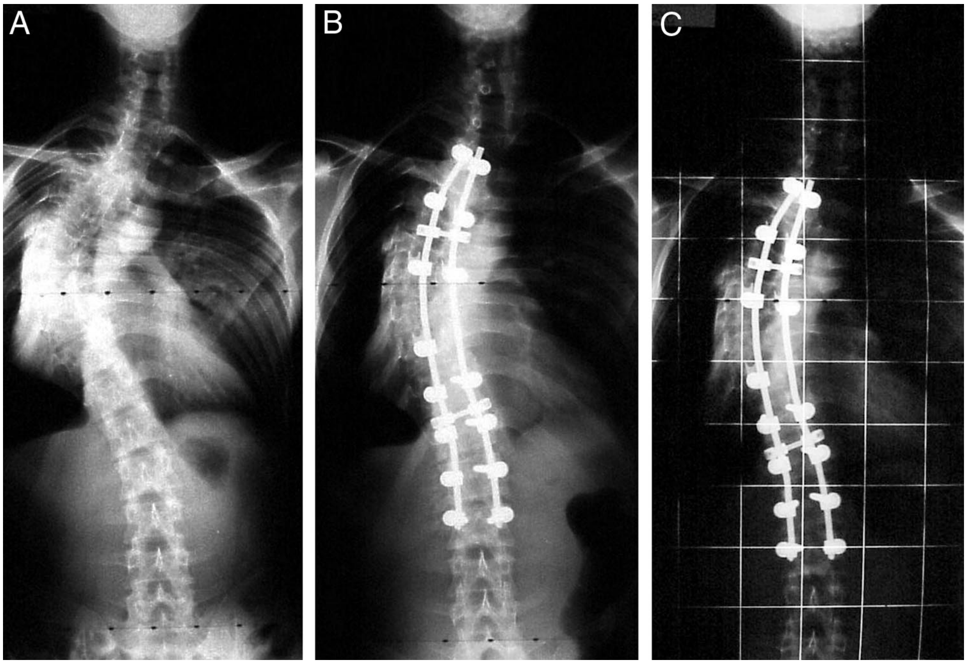

Figure 3. Case 22: male, 25 years old (**A**), treated with T3–L3 posterior instrumented fusion (more recent hybrid instrumentation) (**B**). No complications in the 7 years radiograph follow-up (**C**).

erative, 1 year, and the last follow-up scoliosis correction of patients in Groups 1 and 2. Differences in terms of scoliosis correction achieved with different instrumentations (Groups 1 and 2) did not reach statistical significance. In Group 2 patients, the percentage of postoperative correction was slightly lower (44.23%) than that of Group 1 (46.55%) but remained more stable at the last follow-up (40.97% *vs.* 36.38% of Group 1). However, it should be pointed out that Group 1 follow-up (10.6 years) was longer compared with that of Group 2 (8.1 year).

At the last follow-up, 21 curves of the 37 (59.4%) were stable, belonging to 15 patients (65.2%) of the overall 23: they were considered stable because the angular variation at the last follow-up was always <5°, and equal to 5° in only 2 cases. In the other 16 curves, belonging to the 8 remaining patients (34.7%), a loss of correction of more than 5° and more severe complications occurred.

Complications

No cases of coronal or sagittal decompensation were observed. There were no neurologic complications. Altogether, there were 11 complications, 2 intraoperative and 9 late ones, in 10 patients (2 complications in 1 patient).

Intraoperatively, dural tears occurred in 2 cases (8.6%) and were repaired without late consequences. Late complications were observed in 9 patients (39.1%). In 2 patients (8.6%), surgery had to be repeated, 5 and 8 years after the previous posterior fusion, respectively (Cases 12 and 16): there was a pseudarthrosis with Harrington rod fracture and broken wires; the Harrington rod and sublaminar wires were removed, and revision of the arthrodesis and new hybrid instrumentation were performed; at the last follow-up, 10 and 3 years, respectively, after revision surgery, there was no further loss of correction or new instrumentation failure.

Five patients (21.7%) (Cases 1, 3, 7, 14, and 15) presented distal hook dislodgements with loss of scoliosis correction at follow-up within 10° in 1 case (7°) and more than 10° (range, 14°–17°) in the other 4 cases that did not require surgery (Figure 1). Loss of scoliosis correction within 10° was also observed in another patient (4.3%) (Case 10), without instrumentation failure, that did not require surgery; this patient presented also another complication, a cervicothoracic junctional kyphosis 11 years after posterior instrumented fusion.

Therefore, no complications occurred in the 7 Group 2 patients (segmental instrumentations). All complications occurred among the 16 Group 1 patients, treated using the Harrington rod instrumentation with sublaminar wires.

Discussion

In the past, respiratory insufficiency and cardiovascular diseases had a negative influence on spinal deformity

Table 2. Results of Scoliosis Correction Considering All 23 Patients

	Preop (°)	Postop (°)	1 Yr (°)	Follow-up (°)	% Correction Postop	% Correction 1 Yr	% Correction Follow-up
Min	30	15	15	17	20	20	11.58
Max	110	80	82	95	78.57	71.43	66.15
Mean	69.91	38.17	40.89	44.09	45.88	41.9	37.69
SD	17.57	15.13	15.25	17.08	13.95	13.76	14.12

Table 3. Results of Scoliosis Correction in Patients of Group 1 (Harrington With Sublaminar Wires)

	Preop (°)	Postop (°)	1 Yr (°)	Follow-up (°)	% Correction Postop	% Correction 1 Yr	% Correction Follow-up
Min	30	15	15	17	26.32	20	11.58
Max	110	80	82	95	78.57	71.43	66.15
Mean	70.68	38.28	41.72	45.72	46.55	41.53	36.38
SD	17.42	16.2	16.15	18.36	14.34	13.61	13.97

surgery in MFS, by exposing the patient to the risk of severe complications.[15,17,37,44] The pessimism that shrouded the surgical treatment of these patients is no longer justifiable.[13] The improvement in cardiovascular surgery has extended the lifespan of these patients. In the 1960s, the average life expectancy of patients with MFS was 32 years.[45] It is now 40 years for male patients and 45 for female ones,[13] approaching, according to some,[46] even that of the general population. Careful preoperative patient assessment and more recent anesthesia techniques have helped to limit the risks. Presurgical assessment should take into consideration the cardiovascular abnormalities (mitral valve prolapse and dilatation of ascending aorta) present in MFS.[47] Pulmonary function must also be studied carefully because of the possibility of pulmonary blebs or emphysema problems and the risk of intraoperative spontaneous pneumothorax.[48] Anesthesia techniques,[49] such as autologous blood donation and intraoperative blood loss recovery, can be used successfully in these patients to improve the control of bleeding, as in our 7 Group 2 patients. Indeed, bleeding is greater than that of posterior fusion in idiopathic scoliosis,[50] 1,850 mL on average in our 23 patients and 2,150 mL in those of Jones *et al*.[38]

There are some technical difficulties in spinal deformities surgery in MFS, especially concerning vertebral dysplasia with bone weakness.[17] The Marfan lumbosacral spine presents bony attenuation at one or more levels.[51] This bone weakness may result from abnormal vertebral growth in MFS and, more likely, dural ectasia. Dural ectasia (widening of the dural sac) can be asymptomatic, sometimes responsible for low back pain, and much more rarely cause neurologic problems.[31,38] Vertebral "scalloping" was well described on standard radiographs even in the pre-CT period,[17,24] and is generally due to ectasia. The rate of dural ectasia in the lumbosacral region is now as high as 92% in patients with MFS.[38,52,53] It was observed in all 15 patients of our series that underwent preoperative CT or MRI of the lumbosacral region but was not severe. This finding is frequently reported in other disorders besides MFS,[51,54,55] such as neurofibromatosis,[56,57] Ehlers-Danlos syndrome,[58] or ankylosing spondylitis.[59]

Dural ectasia causes bony erosion of the lumbosacral vertebrae and thinning of the laminae.[24,31,51,53] Preoperative CT enables the most reliable levels of lumbar anchorage of the posterior instrumentation to be chosen. Indeed, dural ectasia causes a compromised lumbar fixation of posterior instrumentation, and loosening of the distal hooks and loss of correction is common. This has been reported in the literature with "historical" instrumentation[16,17,25,29,40] and also found in our Group 1 patients, who were treated by the Harrington rod with sublaminar wires. The use of lumbar pedicle screws obviated these complications in our Group 2 patients.

Although some authors have reported thinning of the pedicle[53,60] and its fracture,[51] this did not occur in our patients. Dural ectasia may also be responsible for the increased rate of dural tears during surgery, which occurred in 2 patients of our series (8.6%) and 3 (8%) of those in another series.[38] Once these lesions were sutured intraoperatively, there were no consequences. It is advisable to perform the operation in the anti-Trendelenburg position to decrease ballooning of the distal dura.[38]

Posterior fusion with instrumentation has been widely adopted for the surgical treatment of scoliosis in patients with MFS.[16–18,25,27,32,36,38,39,40,61] The introduction of new posterior instrumentations has reduced complications, such as instrumentation failure, pseudarthrosis, and postoperative progression of scoliosis, as reported in the literature when using Harrington rods alone or combined with sublaminar wires,[16,17,25,27,29,36,40] and has markedly improved posterior surgery results. A prolonged postoperative immobilization is, however, still recommended, by using braces and not plaster casts.

It is not clear whether there is statistically significant difference in the arthrodesis rate of MFS patients when using autologous iliac bone graft or allograft, as already reported in the literature for posterior fusion in idiopathic scoliosis.[62] It is not fully clear, either, if postoperative infections are more common in MFS than in patients with idiopathic scoliosis: this complication did not

Table 4. Results of Scoliosis Correction in Patients of Group 2 (New Segmental Instrumentations)

	Preop (°)	Postop (°)	1 Yr (°)	Follow-up (°)	% Correction Postop	% Correction 1 Yr	% Correction Follow-up
Min	30	20	20	21	20	20	16.67
Max	105	60	60	62	61.82	61.82	60
Mean	68	39.7	40.7	41.9	44.23	42.83	40.97
SD	18.74	14.2	14.6	14.59	13.51	14.82	14.69

occur in any of our patients or in those of Lipton *et al*,[61] but some cases were reported by Jones *et al*.[38]

Posterior fusion must be extended to include many levels in patients with MFS. In agreement with other authors,[61] we think that the criteria proposed by King *et al*[63] for choosing fusion levels in idiopathic scoliosis should not be applied to these patients. On the contrary, posterior fusion should be extended to include vertebrae that are neutral and stable in both coronal and sagittal planes before surgery. By following this criterion, there were no cases of decompensation in our series, and the loss of correction was limited at follow-up.

Conversely, Lipton *et al*[61] reported a mean loss of scoliosis correction of 20° at a mean follow-up of about 5 years in patients treated with selective posterior fusion (primary curve fused and partial fusion of the secondary curve), whereas correction was kept stable in those patients that had been treated by long posterior fusion (including primary and secondary curves).

Jones *et al*[38] reported high rate of decompensation in MFS after posterior fusion both on the sagittal plane (21%) and coronal one (8%) in a series of patients treated by short fusion. To avert this complication, the same authors[38] recommended not performing short fusion, and including any coronal curve more than 30° in the fusion. The weaker connective tissue in MFS might be a contributing factor to decompensation.[38] Overcorrection of the deformity has also been indicted as playing a role in decompensation[38]; thus, more limited correction has been advocated.

We agree with this attitude of caution when correcting scoliosis in MFS. Surgical correction should not exceed that obtained on preoperative side bending radiographs.[36] Scoliosis correction is, however, much less in patients with MFS than it is in patients with adolescent idiopathic scoliosis. There is no good explanation why these curves are more rigid.[13,17,30] It is rather curious, considering there is a collagen disorder that should be associated with increased flexibility. By posterior instrumented fusion alone, it is more realistic to be content with stabilizing the scoliosis in MFS, and our mean percentage of correction at the longest follow-up of 18 years is less than 40%.

The percentage of correction was more or less similar in both of our groups, even slightly higher in Group 1 patients treated by Harrington instrumentation with sublaminar wires. More recent instrumentations (Group 2) enabled correction to be maintained in the long-term, thus avoiding mechanical complications, nonunion, and loss of correction, which were all observed in Group 1 patients. Scoliosis correction percentages similar to ours were reported in other series, published in the 1970s and 1980s, therefore using "historical" instrumentations, *i.e.*, 38.2%,[29] 41%,[1] 43%,[17] and 44%,[15] but at the expense of severe complications. More recently, Jones *et al*,[38] reported a better correction percentage, which was a mean of 47.4%, and in some cases between 72% and 100%, albeit using different surgical techniques, not only posterior fusion, but also, in some patients, combined anterior and posterior surgery. However, complications were more common than in our series. It is much less risky to perform more modest corrections, on account of the bone weakness typical of this disease.

An anterior release and discectomy, before the posterior instrumentation, may improve correction in severe scoliosis in MFS.[30] However, posterior instrumented fusion alone may provide equally satisfactory results[61] and avoid further risks from an anterior approach. Also, when thoracic hyperkyphosis is concomitant with scoliosis, an anterior stage has been combined with posterior fusion in MFS patients.[25,61,64] Thoracic hyperkyphosis is not very common in MFS and, according to some authors, even very rare.[65] It was not present in any of our patients. The most common sagittal alteration in MFS concomitant with scoliosis is, indeed, thoracic lordosis, which can be adequately corrected by posterior surgery, using sublaminar wires in the past[39] and nowadays the new posterior instrumentations.

■ Conclusion

Despite the pessimism in the literature, our long-term results demonstrate the possibility to obtain a satisfactory stabilization of scoliosis, without concomitant pathologic hyperkyphosis, by posterior instrumentation alone in patients with Marfan's syndrome. Furthermore, our findings suggest that instrumented posterior fusion should be extended to many levels, including vertebrae that are neutral and stable in both coronal and sagittal planes before surgery, in order to ensure a stable correction over time and to avoid the decompensation problems reported in literature.

■ Key Points

- A group of 23 patients with Marfan syndrome were treated surgically with posterior instrumented fusion alone for a scoliosis.
- At a minimum follow-up of 7 years, the results seemed to demonstrate that a satisfactory stabilization of scoliosis can be achieved by posterior instrumentation alone.
- Posterior fusion should be extended to include vertebrae that are neutral and stable in both coronal and sagittal planes before surgery, in order to ensure stabilization of the deformity and reduce the risks of decompression of the spine.

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