Original Article

Sublaminar fixation versus hooks and pedicle screws in scoliosis surgery for Marfan syndrome

ABSTRACT

Background: In patients with Marfan syndrome (MFS), surgical correction of spinal deformities with hooks and/or pedicle screws involves a higher rate of complications than in patients with adolescent idiopathic scoliosis. Therefore, sublaminar instrumentation is often a last resort option. This study wants to assess the ability of sublaminar fixation to achieve three-dimensional scoliosis correction and spine stabilization compared with hook and/or pedicle screw systems.

Methods: Twenty-one MFS patients who underwent posterior spinal fusion at a highly specialized medical center in 1995–2017 were divided into two different groups retrospectively evaluated at a minimum follow-up of 2 years. Group 1 (8 patients) was composed by hooks and screws instrumentation, while Group 2 (13 patients) was composed by hook or pedicle screw system associated to sublaminar wires/bands. Radiological (correction and long-term stability) and general endpoints (mean blood loss, surgery time, and complications) were compared between the groups.

Results: The degree of correction compared with the preoperative status was satisfactory with both approaches, although the difference between them was not significant. No significant differences were found for general endpoints between groups.

Conclusion: Our data suggest that scoliosis correction with sublaminar fixation is not inferior to treatment with hooks and/or pedicle screws.

Level of Evidence: III.

Keywords: Arthrodesis, fixation, hybrid constructs, scoliosis, screws

INTRODUCTION

Marfan syndrome (MFS) is a rare connective tissue disorder caused by a mutation in the fibrillin-1 gene[1] which determines severe connective tissue changes involving various organs and systems.[2-6] The involvement of the musculoskeletal apparatus manifests as ligament laxity, abnormally long upper limbs, arachnodactyly, and spinal deformities like scoliosis.[7-9] Conservative treatment is often uneffective,[10,11] and surgical management is inevitable in a large number of patients.[8,9,12] The aim of this study was to assess the ability of sublaminar fixation to achieve three-dimensional scoliosis correction and spine stabilization compared with hook and/or pedicle screw systems.

METHODS

The records of the patients who underwent posterior spinal fusion (PSF) from 1995 to 2017 at Hesperia

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Hospital (Modena, Italy), which specializes in the treatment of spinal disorders and deformities, were retrieved and analyzed retrospectively. The inclusion criteria were a diagnosis of MFS according to the revised Ghent nosology,[13] spinal deformity treated by PSF, follow-up of at least 2 years, and complete clinical records. Patients who had undergone earlier surgical treatment elsewhere were excluded. The patients meeting these criteria were divided into those treated with hooks or pedicle screws (Group 1) and those treated with a hook or pedicle screw system and sublaminar wires/bands (Group 2). Their records were mined for data on age, blood loss, surgery time, complications, and length of hospitalization.

The preoperative assessment included blood tests, an anesthesiological assessment, and surgical examination. Diagnostic imaging for diastematomyelia,[14] syringomyelia, dural ectasia, dysplastic pedicles, and/or vertebral scalloping was performed by magnetic resonance imaging and/or computed tomography, as appropriate. Severe pedicle dysplasia and/or vertebral scalloping, which hamper hook and screw fixation, was the only criterion for using sublaminar fixation.

All patients underwent PSF with autologous and synthetic bone grafts under continuous monitoring of motor and sensory evoked potentials. Intraoperative blood salvage enabled assessment of blood loss in real time, and reinfusion reduced the requirement for transfusion in the next few days. The postoperative protocol envisaged bed rest for the first 2 days, then gradual mobilization and resumption of ambulation.

During hospitalization, psychological support was offered to patients as well as families to ensure their understanding of the patient's condition and the importance of compliance with the discharge instructions.[15] Patients underwent clinical examination and standing X-rays in two views before discharge, and then clinical and X-ray evaluation at 3, 6, 12, and 18 months and at 2, 3, 5, and 10 years. The preoperative (T0), immediately postoperative (T1), and 2-year (T2) scans were analyzed independently by two operators for scoliosis correction and spine stability in the three spatial planes.

Statistical analysis

Data are reported as mean ± standard deviation. Differences between pre- and post-operative Cobb angles (frontal and sagittal curves, major curve, minor curve, thoracic lordosis, and lumbar kyphosis) were analyzed by Student's *t*-test. Statistical significance was set at $P < 0.05$. Data analysis was performed using STATA 13 software (StataCorp. LLC, TX, USA).

RESULTS

Of the 26 MFS patients, who met the inclusion criteria, 3 (11.54%) were excluded because they had previously been treated elsewhere; one (3.85%) because of loss to follow-up, and one (3.85%) due to insufficient follow-up duration, leaving 21 patients (9 males and 12 females) whose age at the time of surgery was 12–29 (mean 16 ± 4) years. Of these, 8 (38.10%) were Group 1 and 13 (61.90%) were Group 2. Group 1 comprised 6 male and 2 female patients whose mean age was 15.87 (±3.27) years; Group 2 consisted of 3 male and 10 female patients, whose mean age was 16.53 (±4.46) years. The two groups were similar in terms of age ($P > 0.05$).

Imaging findings

All patient data are reported in Table 1.

The analysis of the preoperative radiographs demonstrated that the mean values of the major and minor curve were respectively 67° ± 13.56° and 58.5° ± 17.57° in Group 1 and 69.31° ± 14.07° and 61° ± 13.02° in Group 2, without significant differences between the two groups. Inversion of the sagittal thoracic curve was found in 16 patients, 6 (37.50%) of Group 1 and 10 (62.50%) of Group 2, without significant differences in their mean values. Eight patients, one (12.5%) of Group 1 and 7 (87.5%) of Group 2, had an inversion of the sagittal lumbar curve. This distribution prevented the analysis of these data with Student's *t*-test. Comparison of T1 and T2 radiographs demonstrated that both surgical approaches achieved good correction of the spinal deformities. The measurement of the Cobb angle demonstrated that the mean values of the major curve at T1 and T2 were, respectively,

Table 1: Detailed mean radiological results obtained in Group 1 and Group 2

	Group 1	Group 2
Major curve T0 mean Cobb angle (°)	67±13.56	69.31±14.07
Minor curve T0 mean Cobb angle (°)	58.5±17.57	61±13.02
Thoracic lordosis T0 mean Cobb angle (°)	14.77±10.97	14±11.65-
Lumbar kyphosis T0 mean Cobb angle (°)	20±0	22.57±12.20
Major curve T1 mean Cobb angle (°)	26.32±10.65	25.54±9.45
Minor curve T1 mean Cobb angle (°)	27.67±16.34	28.43±15.90
Thoracic lordosis T1 mean Cobb angle (°)	13.1±7.65	22±8.97
Lumbar kyphosis T1 mean Cobb angle (°)	0±0	7.8±10.10
Major curve T2 mean Cobb angle (°)	28.12±13.87	27.85±11.02
Minor curve T2 mean Cobb angle (°)	29.41±12.74	31.86±14.17
Thoracic lordosis T2 mean Cobb angle (°)	11.66±7.33	18.55±8.06
Lumbar kyphosis T2 mean Cobb angle (°)	3±0	8±9.41
Mean correction of the major curve T0-T2 (%)	58.02±13.04	60.70±12.27
Mean correction of the minor curve T0-T2 (%)	49.72±18.82	48.89±15.27
Correction of thoracic lordosis T0-T2 mean Cobb angle (°)	29.16±17.32	32±9.46
Correction of lumbar kyphosis T0-T2 mean Cobb angle (°)	17±0	17.86±6.51

26.32° ± 10.65° and 28.12° ± 13.87° in Group 1 and 25.54° ± 9.45° and 27.85° ± 11.02° in Group 2. The mean values of the minor curve at T1 and T2 were respectively 27.67° ± 16.34° and 29.41° ± 12.74° in Group 1 and 28.43° ± 15.90° and 31.86° ± 14.17° in Group 2. The improvement was significant in both groups ($P < 0.05$). The T0–T2 percentage correction was 58.02% ±13.04% (major curve) and 49.72% ± 18.82% (minor curve) in Group 1 and 60.70% ± 12.27% and 48.89% ± 15.27%, respectively, in Group 2, without significant differences between the groups. A very satisfactory correction of thoracic lordosis and lumbar kyphosis was also obtained in the sagittal plane, with mean T2 values of 29.16° ± 17.32° and 17° ± 0° in Group 1 and 32° ± 9.46° and 17.86° ± 6.51° in Group 2, respectively (both $P < 0.05$) Figures 1 and 2.

General results

Mean blood loss, calculated on the basis of intraoperative salvage, was 827.69 ± 274.57 ml in Group 1 and 975 ± 272.55 ml in Group 2, without significant differences.

Surgery time was 258.46 ± 56.36 min in Group 1 and 259.25 ± 50.58 min in Group 2; hospital stay was 7.62 ± 1.06 days in Group 1 and 8.30 ± 1.65 days in Group 2. Differences were not significant for either parameter.

Four patients experienced complications. In Group 1, the correction was lost in one patient, who did not, however, require revision. In Group 2, a dural lesion without arachnoid tear was managed with a dural seal and supine decubitus for 2 days; two patients with mesenteric artery syndrome were managed with a nasogastric tube feeding for 3 days, followed by a semi-liquid diet for 7 days.

DISCUSSION

The spinal deformities seen in MFS patients are similar to those seen in androgen insensitivity syndrome (AIS), but are more variable in terms of severity, tendency to progression and response to treatment, either conservative or surgical.[10,16-19] Spinal stiffness, dysplastic pedicles, and vertebral scalloping made the surgical correction high demanding and involves a higher risk of curve decompensation than in AIS patients; for these reasons, the literature recommends longer instrumentation when treating MFS scoliosis.[3,17] However, now that the Harrington rod system is no longer the only available option, the surgeon can choose among a number of tools including hooks, screws, and sublaminar wires and bands. As a result, the rate of correction that can be achieved with surgery has increased from 40% with Harrington's instrumentation to about 50% with hooks alone and 65%–70% using pedicle screws.[18] Where feasible, screw fixation is currently the most effective option, but it involves a higher risk of vascular and dural tears as well as a rate of screw malposition of approximately 10%, due to their manual insertion.[16,17] The sublaminar instrumentation, based on Luque's technique,[18,19] has greatly improved scoliosis correction, also in MFS patients, who suffer from bone brittleness and pedicle dysplasia which is sometimes associated with dural ectasia.[2,20] In these patients, stress shielding at the hook-bone interface can result in laminar fracture. The scarce bone stock also hampers treatment with pedicle screws; in selected patients, poor screw-bone grip may be managed by augmentation with polymethylmethacrylate cement.[21-27]

Sublaminar fixation provides a critically useful option in patients where pedicle fixation would be unsuccessful. We

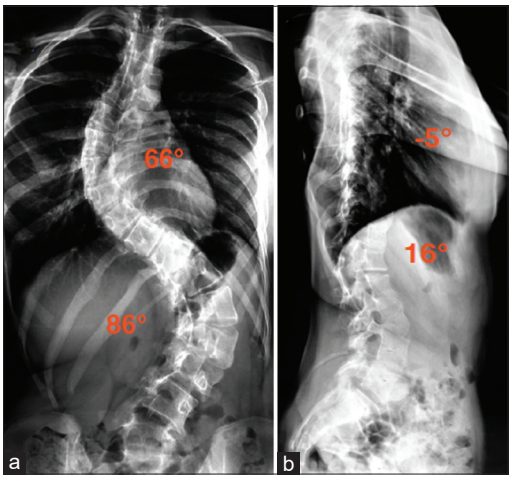

Figure 1: Fourteen years and 3-month-old affected by severe spinal deformity related to Marfan syndrome. Radiological imaging before surgery. Frontal view (a) and lateral view (b)

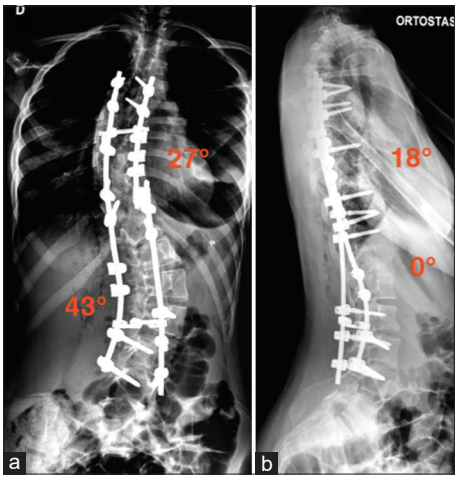

Figure 2: Same patient of picture 1. Sixteen years and 8-month-old girl. Radiological imaging after surgery. The patient was treated with screws and sublaminar bands with good results. Frontal view (a) and lateral view (b)

report the results obtained in two groups of patients, who were treated with hooks and/or pedicle screws with and without sublaminar fixation. The two approaches provided a similar degree of correction, approximately 60% and 49% for the major and the minor curve, respectively. The literature describes widely variable outcomes. Di Silvestre *et al*.[3] reported a correction of about 45% in a 10-year period using several different approaches (Harrington rods, hooks, screws, and sublaminar instrumentation). Qiao *et al*.[16] assessed the effectiveness of hybrid instrumentation (hooks and wires) with or without anterior release and achieved a correction of about 58% with both approaches. These outcomes are similar to those we have obtained in our patients. However, curve correction is not the sole objective, since these procedures involve a high risk of intraoperative, perioperative, and postoperative complications. The most common complications are dural tears and blood loss (early) and pseudarthrosis and curve decompensation (late). Blood loss is the main acute complication since these complex approaches are inherently invasive and MFS patients often suffer from comorbid cardiovascular and respiratory conditions that may impair reaction to abundant blood loss. The literature data are contrasting; for instance, Jones *et al*.[17] described a mean loss of 2148 ml in six patients, whereas Zenner *et al*.[28] reported a mean loss of 1748 ml in 11 patients. This indicates that scoliosis correction surgery involves a greater blood loss in MFS than AIS patients. Similar data have been described by Di Silvestre *et al*.,[3] whereas other researchers have reported comparable blood loss in the two patient groups.[29] Our patients lost about 1000 ml (Group 1, 827.69 ± 274.57 ml and Group 2, 975 ± 272.55 ml).

Dural tears have been reported in 63%–93% of procedures.[30] This high rate probably reflects the considerable incidence of dural, especially lumbosacral ectasia in MFS patients, since dural ballooning is not only a possible cause of spinal pain, but also a risk factor for iatrogenic dural injury and cerebrospinal fluid leakage.[2,30]

Pseudarthrosis is the most severe late complication, shared by most spinal deformities related to a genetic syndrome such as Ehlers–Danlos or neurofibromatosis.[31,32] Its incidence in procedures involving MFS patients is approximately 6%.[3,30-32]

Curve progression and loss of correction is a late complication involving both the coronal and the sagittal plane. Betz *et al*.[32] have described a rate of curve decompensation of 8% and 21% for the coronal and the sagittal curve, respectively, in line with other reports.[28-34] The prevention of curve progression or loss of correction requires a thorough examination, classification, and understanding of each patient's spinal deformities. Jones *et al*.[17] recommend including curves >30° with a stable sagittal profile in the arthrodesis, to perform a careful tissue dissection, and to avoid overcorrection. We adopted these recommendations, as demonstrated by the correction achieved in our patients: about 58% in Group 1 and about 60% in Group 2. Mild progression not requiring surgical revision occurred in a single patient.

This study suffers from a number of limitations, chiefly the small sample size and the wide follow-up range. The data we have collected show that scoliosis correction in MFS patients is a highly demanding procedure that is burdened by a greater risk than surgery in AIS patients. Advances in the instrumentation now enable the correction of severe spinal deformities. Our results demonstrate that sublaminar fixation is a valuable alternative to pedicle fixation, especially in patients with severe scoliosis and severe pedicle dysplasia and vertebral rotation.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Judge DP, Dietz HC. Marfan's syndrome. Lancet 2005;366:1965-76.
2. De Maio F, Fichera A, De Luna V, Mancini F, Caterini R. Orthopaedic aspects of Marfan syndrome: The experience of a referral center for diagnosis of rare diseases. Adv Orthop 2016;2016:8275391. doi: 10.1155/2016/8275391.
3. Di Silvestre M, Greggi T, Giacomini S, Cioni A, Bakaloudis G, Lolli F, *et al*. Surgical treatment for scoliosis in marfan syndrome. Spine (Phila Pa 1976) 2005;30:E597-604.
4. Chun JY, Dillon WP, Berger MS. Symptomatic enlarged cervical anterior epidural venous plexus in a patient with Marfan syndrome. AJNR Am J Neuroradiol 2002;23:622-4.
5. Gargiulo G, Girardo M, Rava A, Coniglio A, Cinnella P, Massè A, *et al*. Clinical comparison between simple laminectomy and laminectomy plus posterior instrumentation in surgical treatment of cervical myelopathy. Eur J Orthop Surg Traumatol 2019;29:975-82.
6. Girardo M, Zenga F, Bruno LL, Rava A, Massè A, Maule M, *et al*. Treatment of aggressive vertebral hemangiomas with poly vinyl alcohol (PVA) microparticles embolization, PMMA, and short segment stabilization: preliminary results with at least 5 years of follow-up. World Neurosurg 2019;128:e283-8.
7. Taylor LJ. Severe spondylolisthesis and scoliosis in association with Marfan's syndrome. Case report and review of the literature. Clin Orthop Relat Res 1987;(221):207-11.
8. Sponseller PD, Hobbs W, Riley LH 3rd, Pyeritz RE. The thoracolumbar spine in Marfan syndrome. J Bone Joint Surg Am 1995;77:867-76.
9. Cinnella P, Rava A, Mahagna AA, Fusini F, Masse A, Girardo M. Over 70° thoracic idiopathic scoliosis: Results with screws or hybrid constructs. J Craniovert Jun Spine 2019;10:108-13.
10. Rava A, Dema E, Palmisani M, Palmisani R, Girardo M, Cervellati S. Effectiveness and reliability of brace treatment in scoliosis related to Marfan syndrome. Minerva Ortop Traumatol 2019;70:188-92.
11. Sponseller PD, Bhimani M, Solacoff D, Dormans JP. Results of brace

treatment of scoliosis in Marfan syndrome. Spine (Phila Pa 1976) 2000;25:2350-4.
12. Savini R, Cervellati S, Beroaldo E. Spinal deformities in Marfan's syndrome. Ital J Orthop Traumatol 1980;6:19-40.
13. Loeys BL, Dietz HC, Braverman AC, Callewaert BL, De Backer J, Devereux RB, *et al*. The revised Ghent nosology for the Marfan syndrome. J Med Genet 2010;47:476-85.
14. Girardo M, Rava A, Fusini F, Lea S, Massè A, Cinnella P. Dysraphism in scoliosis: a case report of diastematomyelia in severe right thoracolumbar congenital kyphoscoliosis. Minerva Ortop e Traumatol; 70. Epub ahead of print April 2019.
15. Giaj Levra N, Cuniberti FA, Rava A, Vietti G, Sciascia S. Health literacy and discharge instruction adherence. J Gen Intern Med 2012;27:273.
16. Qiao J, Xu L, Liu Z, Zhu F, Qian B, Sun X, *et al*. Surgical treatment of scoliosis in Marfan syndrome: Outcomes and complications. Eur Spine J 2016;25:3288-93.
17. Jones KB, Erkula G, Sponseller PD, Dormans JP. Spine deformity correction in Marfan syndrome. Spine (Phila Pa 1976) 2002;27:2003-12.
18. Palmisani M, Dema E, Rava A, Palmisani R, Girardo M, Cervellati S. Surgical treatment of spinal deformities in Marfan syndrome: Long-term follow-up results using different instrumentations. J Craniovert Jun Spine 2019;10:172-8.
19. Ouellet J. Surgical technique: Modern luqué trolley, a self-growing rod technique. Clin Orthop Relat Res 2011;469:1356-67.
20. Altman A, Uliel L, Caspi L. Dural ectasia as presenting symptom of Marfan syndrome. Isr Med Assoc J 2008;10:194-5.
21. Palmisani M, Dema E, Cervellati S, Palmisani R. Hybrid constructs pedicle screw with apical sublaminar bands versus pedicle screws only for surgical correction of adolescent idiopathic scoliosis. Eur Spine J 2018;27:150-6.
22. Rava A, Fusini F, Cinnella P, Massè A, Girardo M. Is cast an option in the treatment of thoracolumbar vertebral fractures? J Craniovertebr Junct Spine 2019;10:51-6.
23. Qiao J, Zhu F, Xu L, Liu Z, Sun X, Qian B, *et al*. Accuracy of pedicle screw placement in patients with Marfan syndrome. BMC Musculoskelet Disord 2017;18:123.
24. Girardo M, Rava A, Gargiulo G, Coniglio A, Artiaco S, Massè A, *et al*. Clinical and radiological union rate evaluation of type 2 odontoid fractures: A comparison between anterior screw fixation and halo vest in elderly patients. J Craniovertebr Junct Spine 2018;9:254-9.
25. Girardo M, Rava A, Fusini F, Gargiulo G, Coniglio A, Cinnella P. Different pedicle osteosynthesis for thoracolumbar vertebral fractures in elderly patients. Eur Spine J 2018;27:198-205.
26. Girardo M, Cinnella P, Gargiulo G, Viglierchio P, Rava A, Aleotti S. Surgical treatment of osteoporotic thoraco-lumbar compressive fractures: The use of pedicle screw with augmentation PMMA. Eur Spine J 2017;26:546-51.
27. Girardo M, Rava A, Coniglio A, Cinnella P, Aprato A, Massè A, *et al*. Importance of polymethylmethacrylate augmentation in the treatment of thoracolumbar osteoporotic vertebral fractures. Minerva Ortop Traumatol 2019;70:65-9. DOI: 10.23736/S0394-3410.19.03920-1.
28. Zenner J, Hitzl W, Meier O, Auffarth A, Koller H. Surgical outcomes of scoliosis surgery in Marfan syndrome. J Spinal Disord Tech 2014;27:48-58.
29. Gjolaj JP, Sponseller PD, Shah SA, Newton PO, Flynn JM, Neubauer PR, *et al*. Spinal deformity correction in Marfan syndrome versus adolescent idiopathic scoliosis: Learning from the differences. Spine (Phila Pa 1976) 2012;37:1558-65.
30. Levy BJ, Schulz JF, Fornari ED, Wollowick AL. Complications associated with surgical repair of syndromic scoliosis. Scoliosis 2015;10:14. doi: 10.1186/s13013-015-0035-x. eCollection 2015.
31. Cappella M, Bettini N, Dema E, Girardo M, Cervellati S. Late post-operative paraparesis after rib penetration of the spinal canal in a patient with neurofibromatous scoliosis. J Orthop Traumatol 2008;9:163-6. doi: 10.1007/s10195-008-0010-x.
32. Betz RR, Iorio R, Lombardi AV, Clancy M, Steel HH. Scoliosis surgery in neurofibromatosis. Clin Orthop Relat Res 1989;(245):53-6.
33. Birch JG, Herring JA. Spinal deformity in Marfan syndrome. J Pediatr Orthop 1987;7:546-52.
34. Robins PR, Moe JH, Winter RB. Scoliosis in Marfan's syndrome. Its characteristics and results of treatment in thirty-five patients. J Bone Joint Surg Am 1975;57:358-68.