Original Article

Surgical treatment of spinal deformities in Marfan syndrome: Long-term follow-up results using different instrumentations

ABSTRACT

Background: Scoliosis is the most frequent spinal deformity related to Marfan syndrome (MFS). Treatment with a brace is often ineffective, and surgical treatment is very challenging; many instrumentations were used along the years. Our retrospective study has the purpose of identifying the reliability of different devices in three-dimensional correction of the spine deformities in MFS.

Materials and Methods: We reviewed retrospectively the records of patients surgically treated, in a single institution between 1999 and 2016, for spinal deformities in MFS. X-rays were reviewed for analyzing the magnitude of the curves in preoperative time (T0), the amount of correction in the immediate after surgery period (T1), and it's stability at follow-up (FU) (T2). The clinical outcomes were also evaluated with the Scoliosis Research Society 24.

Results: A total of 21 patients with a mean age at surgery of 16 years met inclusion and exclusion criteria. Four different construct types were identified: hooks with sublaminar wires (G1), hooks and pedicle screws (G2), pedicle screws (G3), and pedicle screws with sublaminar wires (G4). The mean FU time was 8 years. The average major scoliosis curve had a mean value of 63.48 at T0 and was corrected to 28.81 at T2. Furthermore, minor curve, thoracic lordosis, and lumbar kyphosis (when associated to scoliosis) were also corrected. Student *t*-test showed significative differences ($P < 0.05$) for all curves between T0–T1 and T0–T2 while between T1 and T2, no differences were found. We also evaluated separately the results of each instrumentation, and G3 obtained the best performances.

Conclusions: Our results shows that screws may guarantee a better correction of the deformities.

Level of Evidence: III

Keywords: Hooks, Marfan, scoliosis, screws, sublaminar wires, surgical treatment

INTRODUCTION

Marfan syndrome (MFS) is an autosomal dominant pathology that occurred in around 2–3 in 10,000 people.[1] Scoliosis over 10° can be found in over 63% of MFS patients, and it is often associated with thoracic lordosis and lumbar kyphosis, severe spondylolisthesis, dural ectasia, and pedicle dystrophy.[2-5] Conservative treatment with brace described in the literature is often ineffective.[3,6] Arthrodesis is suggested for over 40° Cobb curves, but a lot of complications are reported in literature.[3,7-9] Our study aims to identify the reliability of different devices in three-dimensional correction of the spine deformities related to MFS.

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MATERIALS AND METHODS

We reviewed retrospectively the records of patients surgically treated, in a single institution between 1999 and 2016, for spinal deformities in MFS. Inclusion criteria were patients aged from 10 to 30 years of age, diagnosis of MFS by Ghent criteria,[10] scoliosis treated with posterior spinal fusion, 2 years of follow-up (FU) at least, and complete medical data. Patients previously treated for other spine defects such as spondylolisthesis or treated in other institutions were not included in the study. All medical data were reviewed for patient's age, surgical procedure, construct type, estimated blood loss (EBL), surgical time, and complications. Magnetic resonance imaging and computed tomography scan were performed before surgery to evaluate the presence of diastematomyelia,[11] syringomyelia, dural ectasia, or other spinal cord malformations. All patients underwent posterior segmental spinal arthrodesis; hooks with sublaminar wires, hooks with screws, screws, and screws with sublaminar wires were used through the years. Intraoperative motor and somatosensory evoked potentials and blood cell salvage technique were adopted for all patients. Intensive care unit recovery was indicated by anesthesiologist in selected cases.

After surgery, all patients remained in bed for the first 2 days; from early after surgery days, walking with physiotherapist was granted. Psychological support service followed, during all hospital stay, patients and their families for improving the adherence to discharge instructions.[12]

Radiographic examinations were conducted before the discharge; clinical and radiographic examinations were conducted at 3, 6, and 12 months after surgery. Yearly examinations were done for the first 3 years and then every 5 years during the FU [Figures 1-3]. The X-ray examinations were double blindly reviewed for analyzing the magnitude of the curves in preoperative time (T0). Major and minor curve angles in frontal view and eventual thoracic lordosis or lumbar kyphosis in the sagittal plane were evaluated using the Cobb method; the mean value of the two measurements was taken as correct for every item for reducing bias. Immediately, postoperative (T1) X-ray examinations and latest FU (T2) ones were also examined evaluating the three-dimensional correction: major and minor curve percentage correction and thoracic lordosis and lumbar kyphosis correction. The clinical outcomes were evaluated with Scoliosis Research Society 24 (SRS24) patient outcome questionnaire,[13] investigating problems related to the intervention such as pain, the recovery of motor skills, the esthetic results, and the eventual affliction of the patient's social life. Score of each domain ranges from 1 (worst) to 5 (best).

Figure 1: Patient 1 (male, 20 year old). Clinical aspect before and after surgery (a and b); posterior view (c and d) sagittal view

Figure 2: Patient 1 (male, 20 year old) treated with pedicle screws' instrumentation. X-ray examinations: (a) frontal view before surgery, (b) frontal view at immediate postoperative time, (c) frontal view at follow-up, (d) lateral view before surgery, (e) lateral view at immediate postoperative time, and (f) lateral view at follow-up

Statistical analysis

Collected data are expressed as mean ± standard deviation. Student's *t*-test was performed to show differences in angular values of frontal and sagittal curves (major curve, minor curve, thoracic lordosis, and lumbar kyphosis). Level of significance was set at $P < 0.05$. Statistical analysis was performed using STATA 13 Software (StataCorp. LLC, USA).

RESULTS

A total of 21 patients (9 males, 12 females) aged between 12 and 29 years of age at surgery (mean: 16 ± 4 years) met inclusion and exclusion criteria and were included in the study. Construct types were hooks with sublaminar wires (G1) in nine cases (9/21; 42.87%), hooks with pedicle screws (G2) in three cases (3/21; 14.29%), pedicle screws (G3) in five cases (5/21; 23.80%), and pedicle screws with sublaminar wires (G4) in four cases (4/21; 19.04%). The mean FU time was 8.04 ± 4.90 years (range: 2–17).

Major scoliosis curve had a mean value of 63.48° ± 17.98° (range: 17°–88°) at T0, corrected to 26.47° ± 15.24° (range: 0°–68°) at T1, and finally achieved the mean of 28.81° ± 15.81° (range: 2°–69°) at T2. Major curve percentage of correction stated at 56.44% ± 17.54% at the latest FU.

In ten cases (10/21; 47.62%), a minor curve was associated to the major one, its mean value was 58.60° ± 14.02° (range: 33°–76°) at T0 that was initially corrected to 27.1° ± 13.99° (range: 10°–60°) at T1, which was 30.30° ± 12.47° (range: 17°–60°) at T2. Minor curve percentage of correction was stated at 47.92% ± 16.22% at the latest FU.

Thoracic lordosis was associated to scoliosis in 16 patients (16/21; 76.19%), and in six of these (6/16; 37.5%), it was associated to lumbar kyphosis that in two cases was associated with scoliosis (2/21; 9.52%). Mean thoracic lordosis was −15.31° ± −11.63° (ranged from −43° to −10°) at T0, 15.80° ± 6.88° (ranged from −16° to 30°) at T1, and 13.60° ± 7.05° (ranged from −12° to 29°) at T2. A mean correction of 28.19°°± 12.93° (range: 5°–56°) was achieved for thoracic lordosis at latest FU.

Mean lumbar kyphosis was stated at 22.25° ± 11.33° (range: 8°–40°) at T0, 6.25° ± 9.53° (range 0°–22°) at T1, and 7.37° ± 8.78° (range: 0°–21°) at T2. At latest FU, the mean lumbar kyphosis correction was 14.87° ± 4.97° (range: 6°–20°). All details are available in Tables 1-4.

Student *t*-test showed significative differences ($P < 0.05$) for all curves between T0–T1 and T0–T2 while between T1 and

Table 1: Complete information about G1

	Hooks with sublaminar wires (G1)								
	Patient number								
	1	2	3	4	5	6	7	8	9
Major curve T0 (cobb°)	52	78	74	68	84	62	70	58	45
Minor curve T0 (cobb°)	50		66		72				40
Thoracic lordosis T0° (cobb°)			12−	10−	43−	5−	5−		15−
Lumbar kyphosis T0 (cobb°)	18	8	38		40	20			
Major curve T1 (cobb°)	22	50	42	40	68	32	27	12	12
Minor curve T1 (cobb°)	18		32		60				10
Thoracic lordosis T1 (cobb°)			10+	20+	16−	30+	22+		15+
Lumbar kyphosis T1 (cobb°)	8	0	22		20	0			
Major curve T2 (cobb°)	24	51	48	47	69	34	29	13	13
Minor curve T2 (cobb°)	23		35		60				17
Correction major curve T0-T2 (%)	53.84	34.61	35.13	30.88	20.23	45.16	58.57	77.59	71.11
Correction Minor curve T0-T2 (%)	54		46.97		16.67				57.5
Thoracic lordosis T2 (cobb°)			8+	16+	13−	29+	20+		11+
Lumbar kyphosis T2 (cobb°)	8	2	21		21	0			
Correction thoracic lordosis T0-T2 (cobb°)			20	26	56	24	25		26
Correction lumbar kyphosis T0-T2 (cobb°)	10	6	17		19	20			
Complications			Dural tear			Anemia that required blood transfusion			Mesenteric artery syndrome
Follow-up (years)	17	15	13	15	11	11	9	7	6
UIV	T7	T4	T3	T3	T4	T4	T4	T4	T2
LIV	L4	L3	L5	L3	L4	L5	L1	L2	T12
EBL (ml)	1000	840	750	650	600	1600	850	870	450
Operating time (min)	240	300	280	260	300	280	250	230	185

All items about three-dimensional correction and its stability; years of follow-up; UIV and LIV; EBL and time duration of surgery. UIV - Upper instrumented vertebra; LIV - Lower instrumented vertebra; EBL - Estimated blood loss

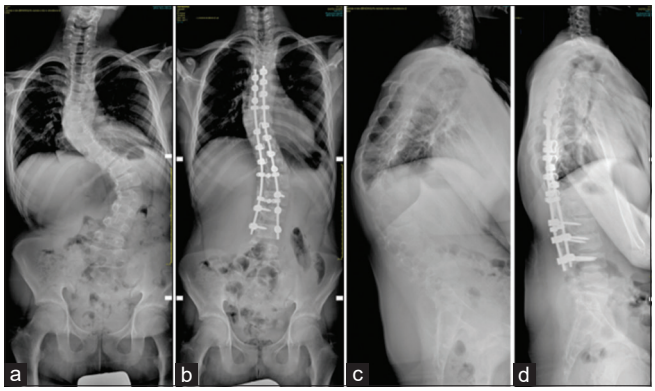

Figure 3: Patient 2 (male, 15 year old) treated with hooks and pedicle screws' instrumentation. X-ray examinations: (a) frontal view before surgery, (b) frontal view at follow up, (c) lateral view before surgery, and (d) lateral view at follow-up

T2, no differences were found, demonstrating so the efficacy and reliability of surgery. We have also compared the results of each construct as reported in Table 5; inferential statistic was not applicable, but evaluating descriptive statistics only, G3 reaches the best performances.

Revising the surgical reports, average EBL and operating time were 883.81 ml ± 276.72 ml (range: 450–1600 ml) and 258.76 min ± 52.93 min (range: 185–400 min), respectively. Complication occurred in four patients (4/21; 19.04%): two patients (2/4; 50%) suffered from mesenteric artery syndrome that resolved thanks to nasogastric intubation, one patient (1/4; 25%) was subjected to red cells concentrated bags transfusion for posthemorrhagic anemia, and one patient (1/4; 25%) suffered from dural tear resolved with the use of spinal sealant system. No cases of pseudoarthrosis, rod breakage, or screw's misplacement occurred.

Upper instrumented vertebra (UIV) and lower instrumented vertebra (LIV) were also recorded. UIV was T2 in one case (1/21; 4.76%), T3 in five cases (5/21; 23.81%), T4 in 13 cases (13/21; 61.91%), T5 in one case (1/21; 4.76%), and T7 in one case (1/21; 4.76%). LIV was T12 in one case (1/21; 4.76%), L1 in three cases (3/21; 14.28%), L2 in two cases (2/21; 9.53%), L3 in seven cases (7/21; 33.33%), L4 in six cases (6/21; 28.57%), and L5 in two cases (2/21; 9.53%). A mean of 12.09 ± 1.48 vertebrae was instrumented for each patient.

Clinical results of SRS24 questionnaire at FU are reported in Table 6.

DISCUSSION

Spine deformities affect many MFS patients; their severity may range from minor curves that do not require any intervention to severe surgical deformities.[2,14] The most representative is

Table 2: Complete information about G2

Hooks with pedicle screws (G2)			
	Patient number		
	1	2	3
Major curve T0 (cobb°)	70	17	37
Minor curve T0 (Cobb°)	60		33
Thoracic lordosisT0° (cobb°)		24−	33−
Lumbar kyphosis T0 (cobb°)			
Major curve T1 (cobb°)	30	0	21
Minor curve T1 (cobb°)	35		22
Thoracic lordosis T1 (cobb°)		20+	17+
Lumbar kyphosis T1 (cobb°)			
Major curve T2 (cobb°)	30	2	25
Minor curve T2 (cobb°)	36		24
Correction major curve T0-T2 (%)	57.14	82.35	32.43
Correction minor curve T0-T2 (%)	40		27.27
Thoracic lordosis T2 (cobb°)		20+	13+
Lumbar kyphosis T2 (cobb°)			
Correction thoracic lordosis T0-T2 (cobb°)		44	46
Correction lumbar kyphosis T0-T2 (cobb°)			
Complications			
Follow-up (years)	2	2	2
UIV	T4	T3	T3
LIV	L4	L3	L3
EBL (ml)	650	900	1400
Operating time (min)	280	360	264

All items about three-dimensional correction and its stability; years of follow-up; UIV and LIV; EBL and time duration of surgery. UIV - Upper instrumented vertebra; LIV - Lower instrumented vertebra; EBL - Estimated blood loss

scoliosis, but sagittal alterations (hyper- and hypo-kyphosis) are usually associated; several times, the inversion of sagittal curves can be appreciated. These skeletal deformities may be associated with severe respiratory deficiency and congenital cardiovascular disease that conditioning the surgical treatment.[3,15] Despite these considerations, the improvement in the quality of life and survival in patients with MFS made surgical treatment of spinal deformities often indicated.

Surgery is more challenging in MFS than in adolescent idiopathic scoliosis (AIS). Spine surgeon must consider several factors; first of all, the type of instrumentation. For a long time, the Harrington rod instrumentation was the only feasible choice[3,7,16] despite a poor rod sagittal modeling. The evolution of the instrumentation including hooks, sublaminar wires/bands, and screws were performed in posterior arthrodesis for these syndromic deformities. Our study reports results of different instrumentations that evolved along the years. In our initial study cases, we adopted hooks with sublaminar wires arthrodesis that provide deformity stabilization with limited correction; moreover, the shearing forces at the bone–hook interface may lead to lamina fracture caused by osteopenia present in Marfan patients.[17] A weak

Table 3: Complete information about G3

Pedicle screws (G3)					
	Patient number				
	1	2	3	4	5
Major curve T0 (cobb°)	60	84	48	71	45
Minor curve T0 (cobb°)				66	
Thoracic lordosis T0° (cobb°)	10−	28−	5−		
Lumbar kyphosis T0 (cobb°)			20		5−
Major curve T1 (cobb°)	18	32	14	26	11
Minor curve T1 (cobb°)				15	
Thoracic lordosis T1 (cobb°)	20+	12+	10+		
Lumbar kyphosis T1 (cobb°)			0		0
Major curve T2 (cobb°)	19	34	19	27	10
Minor curve T2 (cobb°)				20	
Correction Major curve T0-T2 (%)	68.33	59.52	60.42	61.98	77.7
Correction minor curve T0-T2 (%)				69.7	
Thoracic lordosis T2 (cobb°)	18+	12+	7+		0
Lumbar kyphosis T2 (cobb°)			3		
Correction Thoracic lordosis T0-T2 (cobb°)	28	40	12		5
Correction Lumbar kyphosis T0-T2 (cobb°)			17		
Complications					
Follow-up (years)	5	5	5	2	3
UIV	T4	T4	T4	T4	T3
LIV	L1	L1	L3	L4	L3
EBL (ml)	650	850	950	1250	1150
Operating time (min)	250	230	200	280	210

All items about three-dimensional correction and its stability; years of follow-up; UIV and LIV; EBL and time duration of surgery. UIV - Upper instrumented vertebra; LIV - Lower instrumented vertebra; EBL - Estimated blood loss

Table 4: Complete information about G4

Pedicle screws with sublaminar wires (G4)				
	Patient number			
	1	2	3	4
Major curve T0 (cobb°)	86	88	72	64
Minor curve T0 (cobb°)		76	68	55
Thoracic lordosis T0° (cobb°)	22−	16-	5−	7−
Lumbar kyphosis T0 (cobb°)		22	12	
Major curve T1 (cobb°)	24	36	24	15
Minor curve T1 (cobb°)		28	30	21
Thoracic lordosis T1 (cobb°)	12+	14+	15+	20+
Lumbar kyphosis T1 (cobb°)		0	0	
Major curve T2 (cobb°)	22	43	26	20
Minor curve T2 (cobb°)		29	36	23
Correction Major curve T0-T2 (%)	74.41	51.14	63.89	68.75
Correction minor curve T0-T2 (%)		61.84	47.06	58.18
Thoracic lordosis T2 (cobb°)	9+	10+	11+	20+
Lumbar kyphosis T2 (cobb°)		4	0	
Correction thoracic lordosis T0-T2 (cobb°)	31	26	19	23
Correction Lumbar kyphosis T0-T2 (cobb°)		18	12	
Complications		Mesenteric artery syndrome		
Follow-up (years)	6	7	12	14
UIV	T5	T4	T4	T4
LIV	L2	L4	L3	L4
EBL (ml)	750	850	900	650
Operating time (min)	400	230	200	205

All items about three-dimensional correction and its stability; years of follow-up; UIV and LIV; EBL and time duration of surgery. UIV - Upper instrumented vertebra; LIV - Lower instrumented vertebra; EBL - Estimated blood loss

bone quality and the risk of deformity progression in MFS patients are well described in literature; segmental fixation needed better with pedicle screws or sublaminar bands. In cases with a very poor bone quality, cement augmentation may be used as in osteoporotic fractures.[18-23]

A well inserted, till the vertebral body, pedicle screw has a greater moment arm that permits to apply greater corrective forces with respect to hooks.[17,24] These results may be appreciated in our series; in fact, the correction of major curve improved from a mean of 47.46 ± 19.27 in G1 to 57.30 ± 24.96 in G2, when hybrid constructs were used, but better results were achieved with only screw intrumentations.

The most frequent complications reported in literature are dural lesions, pseudoarthrosis, blood loss, and progression/decompensation of all curves.

The first one was reported in approximately 63%–93% of cases.[25] In MFS, a widening of a dural sac called dural ectasia is very common, especially in the lumbosacral region; so, the risks of dural lesions during insertion of screws or positioning of hooks are increased. Dural ectasia may affect the anatomy of the surrounding structures, especially pedicle results to be thinner or dysplastic. Like other spinal deformities associated to congenital syndrome such as neurofibromatosis and[26] MFS, spinal deformities are burdened by this risk of pseudoarthrosis that is stated at almost 10%.

EBL is reported as increased in MFS patients respect the AIS ones. Many studies report an average blood loss of 2000 ml in Marfan patients compared with 1000 ml in AIS.[18,27] These results have probably many causes such as rigid deformities, more challenging surgery, combined anterior and posterior approach, and many others. Despite these literature results in our series, we had EBL comparable to which reported in AIS; we can speculate that these results may be achieved, thanks to posterior approach only, superlative anesthesiologic support, and careful hemostasis.

Appropriate classification and the study of deformity are recommended for preventing progression or decompensation of curves. Jones *et al*. suggested three laws: coronal

Table 5: Comparison of results obtained with different type of instrumentation

Instrumentation	*n*	Correction Major curve T0-T2 (mean %)	Correction minor curve T0-T2 (mean %)	Correction thoracic lordosis T0-T2 (mean Cobb°)	Correction lumbar kyphosis T0-T2 (mean Cobb°)
Hooks with sublaminar wires (G1)	9	47.46±19.27	43.78±18.60	29.50±13.17	14.40±6.10
Hooks with pedicle screws (G2)	3	57.30±24.96	33.63±9	45±1.41	-
Pedicle screws (G3)	5	65.59±7.59	69.70±0	21.25±15.78	17±0
Pedicle screws with sublaminar wires (G4)	4	64.54±9.92	55.69±7.68	24.75±5.06	15±24

Mean percentage of correction for major and minor curve and mean correction in Cobb° for thoracic lordosis and lumbar kyphosis obtained by different type of instrumentation analyzed in our study

Table 6: Clinical results

	Mean	Range
SRS pain	4.62±0.59	3-5
SRS general self-image	4.80±0.51	3-5
SRS self-image after surgery	4.86±0.48	3-5
SRS function after surgery	4.24±0.54	3-5
SRS general function	4.66±0.48	4-5
SRS function activity	4.28±0.64	3-5
SRS satisfaction with surgery	4.95±0.21	4-5

The clinical results obtained with the use of SRS-24. SRS - Scoliosis Research Society

curves >30° and sagittal stable zone must be included in the arthrodesis (selective thoracic fusion is inadequate for double curves), dissection must be limited as much as possible, and correction should be limited to 50%–60% avoiding correction of 80%–100% of technically possible.[18] We agree with these three laws that we follow every day like reported by our results; mean major curve correction ranged from 47.46% to 65.59%.

We are aware that our study has no control group and that the subgroups (G1, G2, G3, and G4) are not enough to permit to conduct inferential statistic examination.

CONCLUSIONS

Considering our results and comparing them to the literature, we can assert that spine deformity surgery in patients affected by MFS is challenging for every surgeon and should be treated in specialized centers cause the possible complications such as dural lesions, high volume blood loss, pseudoarthrosis, and curve decompensation. Evaluating the different results obtained with different instrumentations, screws seem to guarantee better correction of the deformities.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Judge DP, Dietz HC. Marfan's syndrome. Lancet 2005;366:1965-76.
2. Sponseller PD, Hobbs W, Riley LH 3rd, Pyeritz RE. The thoracolumbar spine in Marfan syndrome. J Bone Joint Surg Am 1995;77:867-76.
3. Savini R, Cervellati S, Beroaldo E. Spinal deformities in Marfan's syndrome. Ital J Orthop Traumatol 1980;6:19-40.
4. 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.
5. 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.
6. 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.
7. Birch JG, Herring JA. Spinal deformity in Marfan syndrome. J Pediatr Orthop 1987;7:546-52.
8. Winter RB, Anderson MB. Spinal arthrodesis for spinal deformity using posterior instrumentation and sublaminar wiring. A preliminary report of 100 consecutive cases. Int Orthop 1985;9:239-45.
9. 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.
10. 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.
11. 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 Traumatol 2019;70:107-11. [doi: 10.23736/S0394-3410.19.03917-1].
12. Giaj Levra N, Cuniberti FA, Rava A, Vietti G, Sciascia S. Health literacy and discharge instruction adherence. J Gen Intern Med 2012;27:273.
13. Weigert KP, Nygaard LM, Christensen FB, Hansen ES, Bünger C. Outcome in adolescent idiopathic scoliosis after brace treatment and surgery assessed by means of the scoliosis research society instrument 24. Eur Spine J 2006;15:1108-17.

14. Sponseller PD, Sethi N, Cameron DE, Pyeritz RE. Infantile scoliosis in Marfan syndrome. Spine (Phila Pa 1976) 1997;22:509-16.
15. 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.
16. 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.
17. 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.
18. Jones KB, Erkula G, Sponseller PD, Dormans JP. Spine deformity correction in Marfan syndrome. Spine (Phila Pa 1976) 2002;27:2003-12.
19. 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.
20. 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.
21. 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.
22. Rava A, Girardo M, Legrenzi S, Massè A, Fusini F. Short segment fixation of thoracolumbar fractures with pedicle fixation at the level of fracture. Eur Mediterr Biomed J 2019;14:60-1.
23. 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 Junction Spine 2018;9:254-9.
24. 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.
25. Altman A, Uliel L, Caspi L. Dural ectasia as presenting symptom of Marfan syndrome. Isr Med Assoc J 2008;10:194-5.
26. 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.
27. Shapiro F, Sethna N. Blood loss in pediatric spine surgery. Eur Spine J 2004;13 Suppl 1:S6-17.