

Clinical Study

Trends in spinal deformity surgery in Marfan syndrome

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Abstract

STUDY DESIGN: Analysis of a national database.

OBJECTIVE: To analyze trends in fusion surgery for spinal deformity in Marfan syndrome (MFS) patients, compare patients with and without Marfan, and evaluate differences in surgical approaches.

SUMMARY OF BACKGROUND DATA: National trends of fusion surgery for spinal deformities in MFS patients are not known. Given the rarity of MFS and the nuanced differences in the spinal deformity it causes, it is important to explore differences in fusion surgery between spinal deformity patients with and without MFS.

METHODS: We identified 314 patients (1,410 weighted) with a diagnosis of MFS and spinal deformity who underwent spinal fusion between the years 2003 and 2014. Our primary outcome was national trends in the use of posterior (PSF), anterior-posterior (APSF), and anterior (ASF) spinal fusions. We also compared perioperative complications, mortality rate, length of stay, and hospital charges in a propensity score matched sample of spinal fusion patients with and without a diagnosis of MFS.

RESULTS: The proportion of PSF surgeries increased significantly ($p < .01$) from 66.7% in 2003 to 92.0% in 2014. MFS patients were more likely to have higher neurologic (2.4% vs. 0.79%, $p = .01$) complications. There was a significant association between age and approach ($p < .01$). PSF had a mean age of 20.2, whereas APSF and ASF had mean ages of 27.1 and 35.2, respectively. Approximately 62% of cervical fusions used ASF.

CONCLUSIONS: Our study provides findings from the largest sample analyzed to date and is the only thus far that investigates national trends. Our results are largely consistent with those of other works in that MFS patients undergoing spinal fusion surgery have higher neurologic complications. We also report that surgical treatment has shifted toward a posterior approach. Our findings can give surgeons a better understanding of the postoperative complications and changing national trends in spinal fusion surgery for patients with MFS. © 2019 Elsevier Inc. All rights reserved.

Keywords:

Cervical spine; Costs; Kyphosis; Marfan syndrome; National database; Scoliosis; Spinal fusion; Spinal deformity; Surgical approach; Trends

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Introduction

Marfan syndrome (MFS) is an autosomal dominant disorder caused by a deficiency in the fibrillin-1 gene, resulting in abnormal connective tissue that affects multiple organ systems. Spinal deformity, most commonly scoliosis, is one of the musculoskeletal manifestations of MFS [1]. Despite non-operative measures such as early bracing, up to one quarter of MFS patients with scoliosis may need surgical correction [2]. Although there may be some similarities between scoliosis in MFS and in adolescent idiopathic scoliosis, it has been shown that MFS curves are more likely to progress secondary to connective tissue deficiency and altered transmission of force through the ribs to the spine [2,3].

The available literature on spinal deformity in MFS has mostly focused on treatments and clinical outcomes on a limited number of patients from individual institutions [2,4–8]. A recent retrospective case series demonstrated that the surgical treatment of scoliosis in MFS is associated with an increased risk of complications compared to AIS [8]. In this study, our goal was to analyze and compare national trends and complication rates for fusion surgery in patients with and without MFS, in adult and pediatric MFS patients, and in MFS patients by surgical approach.

Materials and methods

Data source

The Nationwide Inpatient Sample (NIS), the largest publicly available database in the United States, was used for data collection. It contains an approximate 20% sample of hospital inpatient admissions to adult and pediatric hospitals in the United States. Each hospitalization in the NIS includes diagnoses and procedures, patient demographics, hospital characteristics, lengths of stay, charges, and outcomes [9]. Sampling weights, also supplied by the NIS, were used for calculation of national estimates. Our study was exempt from institutional review board approval because the NIS database is publicly available and the data are de-identified.

Patient population

International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnostic and procedure codes were used to identify our patient population. For each year from 2003 to 2014, the NIS database was queried for the following ICD-9-CM codes: adolescent postural kyphosis (737.0), acquired postural kyphosis (737.10), kyphosis due to radiation (737.11), postlaminectomy kyphosis (737.12), kyphosis not elsewhere classified (737.19), acquired postural lordosis (737.20), postlaminectomy lordosis (737.21), other postsurgical lordosis (737.22), other acquired lordosis (737.29), idiopathic scoliosis and kyphoscoliosis (737.30), resolving infantile idiopathic scoliosis (737.31), progressive infantile idiopathic scoliosis (737.32), scoliosis due to

radiation (737.33), thoracogenic scoliosis (737.34), scoliosis not elsewhere classified (737.39), unspecified curvature of the spine (737.40), kyphosis from secondary causes (737.41), lordosis from secondary causes (737.42), scoliosis associated with other conditions (737.43), other curvatures of the spine (737.8), and unspecified curvature of the spine (737.9). The records of these patients were then queried for cervical (81.02, 81.03), thoracic (81.04, 81.05), and lumbar fusions (81.06, 81.07, 81.08).

After all patients were identified with the inclusion criteria above, those with a diagnosis of MFS (759.82) were placed in the index group and removed from the control group. The database was then used to query for patient demographics, institutional characteristics, length of stay, complications, and total hospitalization charges.

Outcome measures

Our primary outcome was national trends in fusion surgeries in MFS patients from 2003 to 2014. Operative characteristics that were evaluated included the surgical approaches posterior spinal fusion (PSF), anterior-posterior spinal fusion (APSF), and anterior spinal fusion (ASF). We also compared major in-hospital complication rates, mortality rate, blood transfusions, use of bone graft, bone morphogenetic protein (BMP), thoracoplasties, osteotomies, length of stay, and hospital charges between spinal fusion patients with and without a diagnosis of MFS.

ICD-9-CM codes were used to query for major perioperative complications using previously described methodology [10]. We collected data on the following complications: neurologic, cervical-spine related, pulmonary, cardiac, thromboembolic, renal, infectious, implant-related, incidental durotomy, and urinary.

Statistical methods

To minimize confounding caused by the uneven distribution of patient characteristics in our analysis between the index group (ie, MFS patients) and the control group (ie, non-MFS patients), we used a 1:5 propensity score matching (PSM). The propensity score was calculated by a logistic model using individual-level covariates including age, gender, race, insurance status, and 23 comorbidities defined by the Elixhauser algorithm [11] (the other 8 Elixhauser comorbidities were not used because very few cases had these diagnoses). Sampling weights were included as a “pseudo-control” variable in this propensity score model because survey weights may capture relevant confounding factors such as geographic and demographic variations. After the propensity score modeling, we performed covariate imbalance testing and confirmed that the standardized bias of covariates was substantially reduced, and the covariates were well-balanced between the index group and the matched control group.

The associations with respect to patient characteristics, hospital characteristics, complications, and comorbidities

were analyzed using weighted chi-square tests. Weighted *t*-tests were used to compare continuous variables (eg, age, length of stay, hospital charges) between the study and matched control groups. Similar statistical methods were employed in other analyses of adult and pediatric MFS patients and in the analysis of MFS patients by surgical approach (PSF, APSF, and ASF). Analysis of variance (ANOVA) model was used to compare continuous variables in the analysis of MFS patients with respect to the three surgical approaches. Significance was set at $p < .05$.

Results

We identified a total of 1,253 patients with MFS and a spinal deformity between the years 2003 and 2014. Within this patient population, 314 received fusion surgery for their deformity. Using the NIS discharge weights, a total of 1,410 fusions were estimated. The percentage of MFS patients receiving fusion decreased significantly from 24.3% in 2003 to 17.7% in 2014 ($p = .01$). During the study period, the proportion of PSF surgeries increased significantly ($p < .01$) from 66.7% in 2003 to 92.0% in 2014 (Fig. 1). APSF use decreased significantly from 22.2% in 2003 to 4% in 2014 ($p < .01$) and ASF use decreased from 11.1% in 2003 to 4.0% in 2014 ($p = .62$). Fusions of the thoracic spine were more commonly done compared to cervical and lumbar fusions; 63.7% of the surgeries involved the thoracic spine, whereas 8.4% and 27.9% involved the cervical and lumbar spines, respectively.

MFS group vs. control group

Table 1 presents patient and hospital characteristics with and without sampling weights, and before and after matching. There were 314 patients with a diagnosis of MFS and 83,148 without (unweighted numbers) who underwent spinal fusion in the sample. These correspond to weighted numbers of 1,410 patients with a diagnosis of MFS and 351,151 patients without. Those with MFS were significantly younger (23 vs. 50, $p < .01$) and more likely to be men (45% vs. 33%, $p < .01$). Those in the MFS group were more often treated at teaching hospitals (84% vs. 65%, $p < .01$) and had significantly more

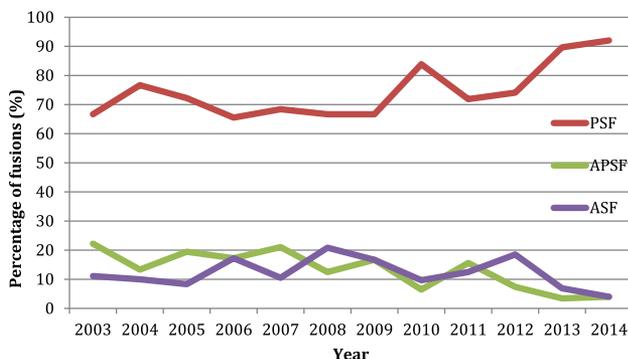


Fig. 1. National trends in surgical fusion. PSF, posterior spinal fusion; APSF, anterior-posterior spinal fusion; ASF, anterior spinal fusion.

operations covered by Medicaid (24% vs. 10%, $p < .01$) and private insurance (61% vs. 47%, $p < .01$).

PSM matching was performed to balance these covariates between MFS and non-MFS patients before comparing their outcomes (Table 1). The matching was achieved for a total of 6,913 patients (weighted number), among whom 1,392 had a diagnosis of MFS (there were 18 patients for whom a matched control could not be found) and 5,521 did not have a diagnosis of MFS (ie, controls; most MFS patients had 5 controls identified, but other MFS patients had only 3 or 4 controls that could be identified from the sample). After the matching, we found that most of the patient and hospital characteristics were successfully balanced (Table 1 and Fig. 2). Figure 2 summarizes the balanced covariates across the MFS and the matched group compared to the covariate distribution before matching.

Compared to the matched non-MFS patients, MFS patients were more likely to have neurologic complications (2.4% vs. 0.79%, $p = .01$; Table 2). There was no significant difference with respect to total complications (20.0% vs. 19.2%, $p = .76$) between the matched MFS and non-MFS patients. There was also no significant difference between the two matched groups with respect to pulmonary, cardiac, thromboembolic, renal, infectious, implant, and UTI complications, or the use of blood transfusions, bone graft, BMP, thoracoplasty, and osteotomy. Finally, there was no significant difference between the two matched groups with respect to hospital length of stay and total hospital charges (Table 2).

MFS patients by age (≤ 18 years and > 18 years)

Overall, there were fewer adult MFS patients receiving spinal fusions with 66% of MFS fusion patients being pediatric patients (≤ 18 years). The mean age of adult patients was 41.8 (range 19–69), whereas the mean age of pediatric patients was 13.3 (range 3–18). There was a significant association between race and age group with adult patients being more likely to be white (88.5% vs. 66.3%, $p < .01$). Pediatric patients were more likely to receive surgeries at teaching hospitals (94.2% vs. 64.3%, $p < .01$) and have their surgeries covered by Medicaid (33.5% vs. 10.28%, $p < .01$). There was no significant difference between length of stay or total hospital charges. Pediatric patients were also more likely to have higher APR-DRG severity of illness scores ($p < .01$) [12].

Adult MFS patients were more likely to have the following comorbidities: depression (8.4% vs. 1.5%, $p < .01$), hypertension (23.4% vs. 6.9%, $p < .01$), and fluid and electrolyte disorders (21.5% vs. 8.8%, $p < .01$). They were also more likely to have higher thromboembolic complications (2.8% vs. 0%, $p = .04$), osteoporosis (4.7% vs. 0%, $p < .01$), urinary tract infections (3.7% vs. 0.48%, $p = .048$), and total complications (28% vs. 16.9%, $p = .02$). Pediatric patients had higher rates of blood transfusions (24.2% vs. 14%) and lower rates of BMP use (5.8% vs. 27.1%).

Table 1
Comparison of patient and hospital characteristics before and after matching

Characteristics	Unweighted sample			Weighted before matching			Weighted after matching		
	Marfan	Control	p Value	Marfan	Control	p Value	Marfan	Control	p Value
Total number of patients	314	83,148		1,410	351,151		1,392	5,521	
Age, mean	23.0	49.9	<.01*	23.2	49.5	<.01*	23.2	25.6	.05
Race (%)			<.01*			.01*			.30
White	57.6	68.5		57.8	67.5		57.8	60.3	
Black	8.6	5.9		8.6	5.7		8.6	7.5	
Hispanic	7.6	4.8		6.8	4.7		6.9	9.0	
Other	4.8	4.0		4.7	4.0		4.8	2.6	
Missing	21.4	16.8		22.1	18.1		21.9	20.6	
Gender (%)			<.01*			<.01*			.87
Male	44.7	33.2		44.8	32.9		44.7	45.2	
Female	55.3	66.8		55.2	67.1		55.3	54.8	
Teaching hospital (%)			<.01*			<.01*			.35
Yes	83.6	64.9		83.8	64.8		83.6	80.8	
No	16.4	35.1		16.2	35.2		16.4	19.3	
Hospital bedsize (%)			<.01*			<.01*			.10
Large	57.3	62.4		56.3	63.5		56.0	58.4	
Medium	18.8	22.3		19.7	22.4		19.8	23.1	
Small	23.9	15.3		24.0	14.1		24.2	18.5	
Hospital location (%)			.06			.06			.36
Urban	98.7	96.6		98.7	96.7		98.7	97.8	
Rural	1.3	3.4		1.3	3.3		1.3	2.2	
Hospital region (%)			.08			.11			.81
Northeast	18.8	14.4		19.8	14.4		19.9	17.8	
Midwest	24.2	23.0		25.8	23.3		26.0	25.0	
South	37.3	39.1		34.4	38.7		34.4	36.3	
West	19.7	23.5		20.0	23.6		19.7	10.9	
Insurance status (%)			<.01*			<.01*			.97
Medicare	6.7	37.4		6.9	36.8		7.0	7.7	
Medicaid	25.6	9.9		24.1	9.8		24.2	23.9	
Private	59.4	46.4		60.7	47.1		60.6	59.7	
Self-pay, no charge, other	8.3	6.3		8.2	6.4		8.3	8.7	

* Significance at p<.05.

MFS patients by surgical approach (PSF, APSF, and ASF)

There were a total of 234 PSF, 42 APSF, and 38 ASF cases (1,126, 200, and 200 weighted, respectively). There was a significant association between mean age and

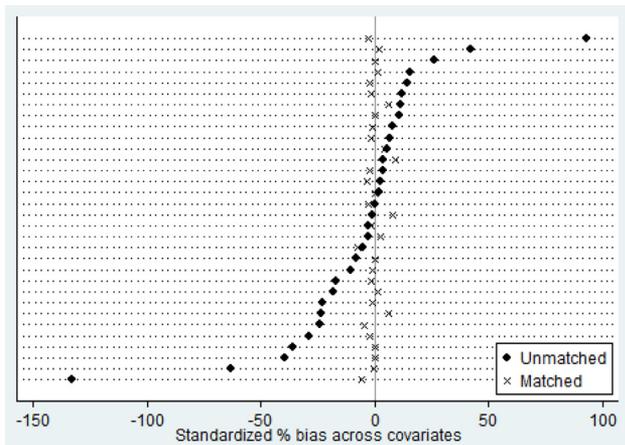


Fig. 2. Standardized bias of covariates before and after propensity score matching.

Table 2
Outcomes of PSM-matched patients

	Marfan	Control	p Value
Complications (%)			
Neurologic	2.4	0.79	.01*
Cervical spine-related	0	0.85	.11
Pulmonary	8.2	6.0	.19
Cardiac	1.7	2.8	.37
Thromboembolic	1.1	0.33	.11
Renal	0.35	0.53	.69
Infectious	0.69	1.4	.35
Implant-related	6.5	5.0	.36
Incidental durotomy	0.71	1.6	.25
UTI	1.6	2.8	.23
Any complication	20.0	19.2	.76
Died (%)	0.35	0	.3
Blood transfusion (%)	19	20.5	.62
Bone graft (%)	41	41.4	.9
BMP (%)	13.1	12.6	.8
Thoracoplasty (%)	1.4	2.0	.5
Osteotomy (%)	5.8	5.0	.61
Total charges, dollars (mean)	143,401	140,414	.66
Length of stay, d (mean)	7.2	6.2	.2

PSM, propensity-score matching.

* Significance at p<.05.

approach ($p < .01$): those who underwent PSF had a mean age of 20.2, whereas those who underwent APSF and ASF had mean ages of 27.1 and 35.2, respectively. ASF surgeries were less likely to be performed at teaching hospitals (65.6% vs. 86.9% and 84.2 for PSF and APSF, respectively; $p = .01$). There was a significant association between surgical approach and total mean charges ($p < .01$): \$150,247 for PSF, \$191,284 for APSF, and \$96,170 for ASF surgery. There was also a significant difference in the mean length of stay by approach ($p < .01$): 5.9 days for PSF, 9.6 days for APSF, and 10.9 days for ASF. Those undergoing PSF were more likely to be covered by Medicaid (28.3% vs. 16.7% and 18.4% for APSF and ASF, respectively; $p < .01$). APSF patients were more likely to have higher APR-DRG severity of illness scores ($p < .01$) [12].

There was no significant difference between approach and individual or total complications ($p = .62$). PSF and APSF patients were more likely to receive transfusions (23.1% and 21.4% vs. 5.3%; $p = .03$) and bone grafts (46.2% and 45.2% vs. 23.7%; $p = .03$).

MFS patients by fusion location

There were a total of 188 thoracic, 66 lumbar, and 28 cervical cases (914, 312, and 137 weighted, respectively). There were also multiregional fusions with 30 involving the thoracic and lumbar spines and 1 involving the thoracic and cervical spines (144 and 5 weighted, respectively). We compared cervical fusions to thoracolumbar fusions. Patients undergoing cervical fusion were significantly older with a mean age of 42.1 compared to 21.1 for the thoracolumbar group ($p < .01$). Thoracolumbar fusions were more likely to be performed at academic institutions (86.2% vs. 57.1%, $p < .01$). There was also a significant difference in the total mean charges ($p < .01$): \$156,193 for thoracolumbar and \$80,894 for cervical fusions. There was no significant difference in length of stay.

Discussion

In the current study, we have demonstrated important national trends in the use of spinal fusion surgery for MFS patients with spinal deformity. We sought to compare spinal deformity patients with and without MFS. Additionally, we compared MFS patients by age and surgical approach. We discovered that the rate of spine surgery in MFS patients with spinal deformities has slightly decreased from 2003 to 2014. Previous studies have reported the rate of surgery for scoliosis in MFS to range from 14% to 21% [3,13]. Our results are consistent with these reports. We found that 24.3% of MFS patients with spinal deformities underwent fusion in 2003, whereas that number was 17.7% in 2014. Though the surgical approach is largely dependent on the location and severity of the deformity, we have found a significant increase in the use of PSF. Although PSF was used in 66.7% of the surgeries in 2003, it was the principal approach in 2014 with 89.7% of the surgeries being PSF.

Although the reason for this is likely multifactorial (eg, increased use of pedicle screw instrumentation for the thoracic spine, lower complications), we also discovered that PSF was associated with a significantly lower length of hospital stay (5.9 days) compared to APSF and ASF (9.6 and 10.9 days, respectively).

In terms of comparing MFS patients to the control group, we found that MFS patients have higher rates of neurologic complications and no differences in other or total complications. This finding is most likely due to MFS's association with dural ectasia, which can result in thinning of the pedicles rendering pedicle screw placement challenging [14]. In a recent study of 119 pediatric patients that investigated the frequency of dural ectasia in MFS patients, Veldhoen et al. found that the prevalence of dural ectasia in their study population was 90.3% [15]. Other studies similarly report numbers as high as 92% [16]. Dural ectasia results in weakening of the dura, making it more susceptible to tears and neurologic injury. Due to this high prevalence, a preoperative MRI is recommended before spinal surgery in all MFS patients [17].

Many authors in the literature have commented on increased blood loss during spinal deformity correction surgery for MFS [4,6,17,18]. However, there are some conflicting findings in the literature. In a 2012 retrospective case control study, Gjolaj et al. found no significant difference in blood loss when comparing 34 MFS patients and 68 AIS patients undergoing spinal deformity surgery [5]. In our analysis of MFS patients by age, we discovered that pediatric patients had significantly higher rates of blood loss. We suspect that this younger patient population has the more severe deformities, which are addressed at a pediatric age. Moreover, spine deformity in MFS is usually seen in these younger patient populations. Indeed, out of all MFS patients in our analysis, 84.6% of those who had MFS coded as their primary diagnosis were 18 or younger. Our findings support the conclusion that MFS patients undergoing deformity correction are at increased risk for blood loss.

In our analysis of MFS patients by fusion location, we discovered that 62.1% of cervical fusions used ASF. Though prior studies have advocated for the use of ASF in cervical kyphosis due to lower complication rates [19], there is a lack of literature on its use on the cervical spine in MFS. Due to the abnormal bone morphology and ligamentous laxity in MFS [3,20], one would expect that the cervical spine may be just as vulnerable as the rest of the spine. In a 1997 prospective radiographic and clinical analysis of 104 patients, Hobbs et al. found that cervical spine abnormalities, such as focal kyphosis and increased atlantoaxial instability, are more common in patients with Marfan than in the general population [21].

In a retrospective case series, Herzka et al. reported on the treatment of three MFS patients ages 9, 13, and 20 who presented with atlantoaxial rotatory subluxation [20]. Two of the patients received PSF whereas the other was treated nonoperatively. Our study found that in MFS patients who

received cervical fusions, 62.1% received ASF whereas 27.6% and 10.3% received PSF and APSF, respectively. This finding sheds light on the national trends in the treatment of cervical spine involvement in MFS, as ASF might be the preferred approach to prevent further subluxation in these patients [22].

Overall, however, we found that PSF was the most widely used approach, comprising 80.4% of thoracic and 71.9% of lumbar fusions. In a 2006 study, Qiao et al. compared PSF and APSF use in Marfan patients with scoliosis; they treated 30 patients with APSF and 66 patients with PSF [23]. Though the APSF group yielded slightly larger correction rates for thoracic (62.5% vs. 56.2%) and lumbar (68.3% vs. 62.7%) scoliosis, the authors concluded that PSF can yield comparable outcomes with lower rates of blood loss and operation time. In a 2011 retrospective study of 12 patients, Li et al. similarly concluded that PSF alone is safe and effective for the treatment of MFS patients with scoliosis [24]. These recommendations are consistent with the trends we identified in our study, in that APSF use decreased significantly from 2003 (22.2%) to 2014 (4.0%). The APSF surgeries that are still carried out are likely performed in the most severe cases, which necessitate more reliable corrections.

Our study has several limitations. First, the NIS is an administrative database that relies on accurate and consistent data entry; incorrect and inconsistent coding are the potential sources of error that affect the data and our results. The NIS also does not have the level of detail to comment on the severity of deformities nor does it provide radiographic data, which make it difficult to investigate why certain treatment modalities were used. Last, it would be of clinical interest to see how many patients underwent instrumentation to the pelvis, but the database is not granular enough to provide this information. Despite these limitations, the NIS has been useful in evaluating spinal deformity treatment trends in syndromic and genetic conditions [25–28].

Conclusions

Our study provides findings from the largest sample analyzed to date and is the only thus far that investigates national trends. Our results are largely consistent with those of other works in that MFS patients undergoing spinal fusion surgery have higher neurologic complications and greater blood loss.

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