

Surgical outcome of scoliosis in Marfan syndrome: a case series report

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ABSTRACT

Scoliosis in marfan syndrome (MFS) manifests on 60% patients. Moreover, the scoliosis noticeable in earlier age is more progressive, refracted, and rigid compared to idiopathic adult scoliosis. The surgical correction provides notorious higher perioperative risk, whereas conservative treatment using brace is not effective to prevent progressivity of the scoliosis. In this a case report, we reported the surgical outcome of MFS scoliosis patients with MFS who operated using posterior fusion instrumentation by mean of the quality of life SF-36 questioner. This was a retrospective case series involving five MFS scoliosis patients who underwent posterior fusion instrumentation with initial Cobb angle of $87.4 \pm 17.57^\circ$ and initial kyphotic angle of $32.8 \pm 14.52^\circ$. Clinical, radiological and quality of life of the patients based on SF-36 questionnaire were evaluated within 6-36 months follow up. Post-operative showed the Cobb angle become $46.2 \pm 16.3^\circ$ and the kyphotic angle become $21.6 \pm 9.94^\circ$. No intraoperative or post-operative complications were observed. After 6-36 months follow up, the Cobb angle became $45.2 \pm 17.48^\circ$ and the kyphotic angle became $21.6 \pm 9.94^\circ$. In addition, all patients had physical and mental health scored similar to 2 years post-surgery scoliosis scoring according to SF-36 orthopedic scoring guidelines. In conclusion, the surgical outcome of posterior fusion instrumentation in MFS scoliosis showed good correction of Cobb angle and Kyphotic angle. The quality of life of the patients based on physical and mental health questionnaire is satisfactory.

ABSTRAK

Skoliosis diderita pada 60% penderita sindrom Marfan (SM). Selain itu, skoliosis yang diderita sejak usia muda lebih progresif, bias dan kaku dibandingkan dengan skoliosis dewasa idiopatik. Penatalaksanaan melalui tindakan bedah memberikan risiko perioperative lebih tinggi, sedangkan penatalaksanaan konservatif dengan penjepitan tidak efektif untuk mencegah progresivitasnya. Dalam laporan kasus ini disampaikan luaran tindakan bedah pasien SM dengan skoliosis dan kualitas hidupnya berdasarkan pertanyaan dalam SF-36 setelah dilakukan tindakan dengan peralatan fusi posterior. Laporan kasus serial retrospektif ini melibatkan lima penderita skoliosis dengan SM yang menjalani instrumentasi fusi posterior sudut Cobb awal $87,4 \pm 17,57^\circ$ dan sudut kifotik awal $32,8 \pm 14,52^\circ$. Kondisi klinik, hasil pemeriksaan radiologi dan kualitas hidup berdasarkan kuisisionair SF-36 dievaluasi selama pengamatan 6-36 bulan. Pasca operasi menunjukkan sudut Cobb menjadi $46,2 \pm 16,30^\circ$ dan sudut kifotik menjadi $21,6 \pm 9,94^\circ$. Tidak dijumpai komplikasi intraoperasi dan pasca operasi selama pengamatan. Setelah dilakukan pengamatan selama 6-36 bulan, sudut Cobb menjadi $45,2 \pm 17,48^\circ$ dan sudut kifotik menjadi $21,6 \pm 9,94^\circ$. Semua pasien mempunyai skor kesehatan fisik dan mental sama dengan skor skolastis setelah 2

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tahun pasca operasi menurut petunjuk penilaian oropedi SF-36. Dapa disimpulkan luaran bedah instrumentasi fusi posterior pada pasien scoliosis dengan SM menunjukkan koreksi yang baik terhadap sudut Cobb dan kifotik. Kualitas hidup pasien berdasarkan kuisionair kesehatan fisik dan mental memuaskan.

Keywords: scoliosis - Marfan syndrome – quality of life - questionnaire SF-36 – Cobb angle

INTRODUCTION

Marfan syndrome (MFS) is one of multi systemically disorder caused by generalized collagen abnormality (FBN1; fibrillin-1) that is inherited in autosomal dominant.^{1,2} Other than the excessive longitudinal growth on growth plate cartilage (hyperchondroplasia), thin and long extremity seen as *spider-like finger (arachnodactyly)*; the tangible sign are the facial features (dolichocephalic, enophthalmos, down slanting palpebral fissures, malar hypoplasia, retrognathia), and chest asymmetry (pectus excavatum/carinatum).^{3,4} The condition is somehow stigmatism, hamper the life insurance opportunity, as well as the psychosocial burden². The diagnosis criteria have been revised for the purposes not to over diagnosed or under diagnosed it.^{2,3} Genetic evaluation is not the only diagnosis tool, yet other ancillary technique is not always available and feasible for our community.

The undetected dilatation of aortic base such as aneurysm of ascending aorta might complicate the scoliosis surgery or the other way the abnormal thoracic cage would complicate the cardiac and pulmonary condition which could happened later in young adult age.^{1,2,3,4} The main management for Marfan syndrome depends on which chief organ system involved.^{3,4,8,9} Scoliosis prevalence in this syndrome is around 60 %. Scoliosis in MFS usually occur and being noticeable at younger age, more progressive,

refractory, and rigid. It is also the main complaint of back pain in the later age.^{5,6,7}

In common community, scoliosis is the disease, not just the symptom. Very seldom the underlying cause was investigated. In our former study in screening the junior high school student in Surabaya, 2.7% students indicate scoliosis and 3 neglected cases were found.¹⁰ Most of the cases are adolescent idiopathic scoliosis (AIS); which surgery is performed when the cobb angel is more than 40°. But in MFS the early onset and progressive scoliosis is the problem to be solved.^{3,4,5} The use of brace in conservative treatment is not effective for preventing the progressivity of the scoliosis.⁵ Surgery by posterior fusion instrumentation is one of the technique for scoliosis correction nevertheless the collagen abnormality and anatomical deformity in MFS might complicate the perioperative risk and the correction result.^{7,11,12} The purpose of this study is to evaluate the surgical outcome not only the correction by clinical and radiology but also by using SF-36 questionnaires to value the physical and mental health of the MFS patient.

CASE REPORT

This is a descriptive retrospective study of case series. The research protocol was approved by institutional ethics and review board in both 2 hospitals, Dr Soetomo General Hospital Surabaya and Surabaya Orthopedic & Traumatology Hospital. Five MFS patients with scoliosis were underwent corrective

surgery using posterior fusion instrumentation according to standardized department protocol during 2014 to 2016. All patients were managed by a single orthopedic surgeon, same instrument, and same management protocol. The MFS was diagnosed based on Revised Ghent criteria.² The pre-operative management was incorporating cardiac evaluation, ophthalmology evaluation, and MRI of the spine to evaluate the dural ectasia. None of the patients had aortic root aneurysm, ectopia lentis, or family history. But 4 out of 5 had thick myopia, the facial features, and >7/20 systemic features involvement.² Database of initial patient condition were recorded from medical record, including the demographic data, clinical and radio imaging data. Pre and post Cobb angle and Kyphotic angle were compared to the follow-up measurement. The SF-36 questionnaire was performed by interviewing the patient on the follow up visit. Follow up was 6-36 months.

RESULTS

The 5 MFS patients were all female with the age range of 11–17 years (:13.6 years) at

surgery; 13–18 years (:15.6 years) at the follow up, and all were without cardiac or respiratory abnormalities. All of them were came when the Cobb angle were > 70°; 2 of them were > 100° and 2 patients of double curves (RT-LL). The mean Cobb angle before surgery was (87.4±17.5)°. The initial kyphotic angle was (32.8±14.5)°. The degree of correction directly post-surgery was (41.2±1.78)° and (16.8±9.83)° respectively. The number of the segment fusion were 15(Th2-L4) – 17(Th1-L5) segments. After surgery all patients were immobilized with brace for 3-6 months.

The length of the surgery averaged (322±38.3) minutes (range:270-370 minutes). The average blood loss was (495±44.7) cc (range: 450–550 cc). There was absent of surgical complication during and after the surgery. On Follow-up after 6 – 36 months, there were almost no scoliosis nor kyphotic progression in all patients. The physical health status and mental health status from SF-36 questionnaire were good with the average of PHS 48.4 and MHS 49.

TABLE 1. Clinical, radiology, correction, and quality of life (SF-36) in Scoliosis Marfan syndrome patients

Patient	Sex	Age (years)	Curve pattern	Fusion Level	Cobb angle (°)			Kyphotic angle (°)			Op time (min)	Blood Loss (cc)	SF-36 score	
					Pre-op	Post op	FU	Pre-op	Post op	FU			PHS	MHS
1	F	18	RTL	T2-L5	102	58	58	45	16	16	340	500	45	55
2	F	16	RT	T2-L4	70	30	25	38	32	32	270	450	55	46
3	F	18	RT-LL	T1-L4	80/68	40/32	40/32	8	22	22	330	550	42	48
4	F	13	RT	T1-L4	110	68	68	33	8	8	300	450	47	50
5	F	13	RT-LL	T1-L5	75/55	35/20	35/20	40	30	30	370	525	53	46

RTL: right thoraco-lumbar; RT: right thoracic; RT-LL: right thoracic-left lumbar; Pre-op: preoperative; Post op: post-operative; Op time: operative time; FU: follow up (6-36 months); PHS: Physical Health Status; MHS: Mental Health Status



FIGURE 1. Clinical appearance 18 years female with RT-LL curve treated with posterior fusion instrumentation, (a) initial pre-operative, (b) follow up post-operative after 1 year

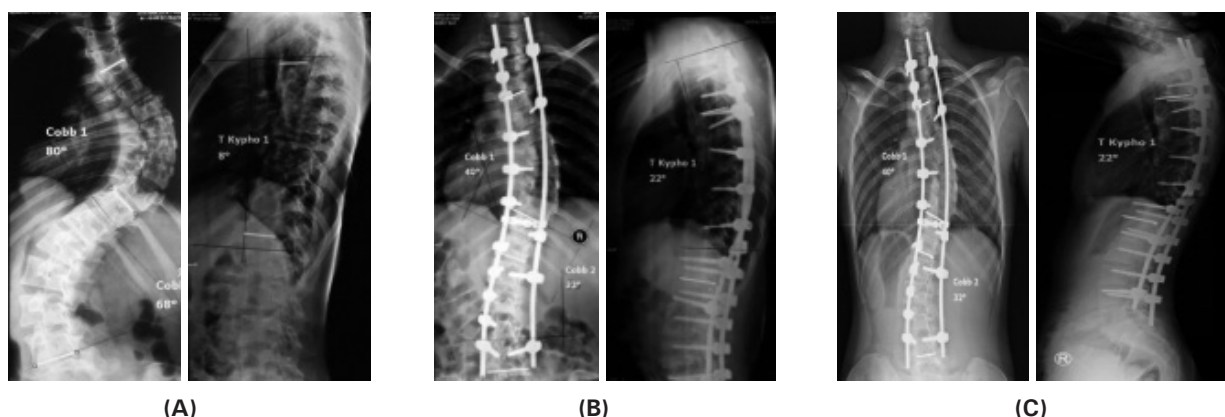


FIGURE 2. Radiological imaging of 18 years female with RT-LL curve treated with posterior fusion instrumentation, (a) pre-operative, (b) post-operative, (c) follow up after 1 year

DISCUSSION

In this study the surgical outcome is not only about the physical outcome (cobb angel and kyphotic angel) but also about the quality of life. Since the main complaint from the patient's site is the body contour, the self-esteem confident is also considered as one of the surgical outcome. The SF-36 do not have the cut-off point for good or bad result.¹³ The SF-36 forms have been used often in examining orthopaedic patient populations. The brief guide from Various Orthopaedic Procedures and Conditions comparing SF-36 pre and post-surgery reported similar result

of scoliosis patient, PH 42.2 to 46.4 and MH 48.9 to 50.6.¹⁴

Surgical outcome of areconstruction surgery is usually concern for degree of correction. The degree of correction of the cobb angel (40–44)° and kyphotic angel (6-29) ° were comparable to study reported by Zenner et.al when using posterior spinal fusion only (44°) yet less than when using combine PSF and Anterior (57°).¹¹ Nonetheless blood loss and time of surgery were better in posterior spinal fusion only.^{6,7,11,12} When comparing with surgery of AIS, the correction, blood loss, and time of surgery were similar and not significant.^{6,15}

Surgical therapy is an effective choice of therapy for scoliosis in Marfan syndrome, bracing has been proven only success in 17% cases but eventually those cases need surgery as well, owing to the progressing curve.^{5,7,11,12} The use of growing rod is a good choice, anterior release is not necessary if surgery was not postponed; when the curve has progressed rapidly which is the notorious problem in MFS.^{6,7,11} Hook is not advisable due to the underlying desmogenic disorder. Before surgery MRI should be assessed for possible dural ectasia, pedicle thinning, and dysplastic lamina.¹⁵

The surgery must cover all the major curve including pelvis fusion involvement when necessary to avoid the re-surgery.^{11,12,15} We fused 15-17 level and after up to 3 years follow up the progression was none to minimal (5°) in one case. The invention of pedicle screws gives important progression in scoliosis correction. Pedicle screws, using the strongest part of vertebral body as an anchor, provide the spine surgeon with an enhanced three-dimensional deformity correction. Pedicle screws that placed in the vertebral body have 30 % greater moment arm for applying corrective forces than posterior hooks. Posterior segmental instrumentation with a powerful pedicle screw anchor offers satisfactory correction without significant loss of curve correction even in severe deformity cases.¹² Posterior fusion with instrumentation has been widely used for the surgical treatment of scoliosis in the Marfan syndrome, particularly in the curves ranging beyond 40°–50° that tend to progress more after skeletal maturation.^{7,12}

This study need to be continued with larger samples and multicenter to give orthopedic surgeons precise and merit planning in surgical management of scoliosis in MFS. Scoliosis in MFS would not be too complicated if planned in the current knowledge of underlying the multi systemically disorder as MFS.

CONCLUSION

The surgical outcome of posterior fusion instrumentation in MFS scoliosis shows good Cobb angle and Kyphotic angle correction. The blood loss, time of surgery, and surgical complication is all satisfying and comparable to other study with larger samples. The quality of life of the patients based on physical and mental health questionnaire (SF-36) is similar to other various orthopedic procedure and condition.

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