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Mitral valve replacement after Bentall de Bono procedure in a Patient with Marfan Syndrome – A case report

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ABSTRACT

Marfan syndrome is a connective tissue disorder which is caused by a mutation in the FBN1 gene, located on chromosome 15q21. This mutation results in abnormal fibrillin synthesis. Impaired structure of fibrillin protein results in the development of complications within the cardiovascular system, the skeleton and organ of sight. The most common cause of death among patients with Marfan syndrome is aortic disease, but mitral valve dysfunction is also possible. We present the case of a 30-year-old patient with Marfan syndrome who underwent a Bentall de Bono procedure at the age of 18 due to aneurysmal dilation of the aortic bulb. After several years, the patient was qualified again for surgery, because he developed symptomatic severe mitral regurgitation with calcifications present on the mitral annulus and also rupture of the chordae tendineae - which was all confirmed by echocardiography. Due to advanced anatomical changes and the inability to perform plastic surgery on the diseased valve, the patient was implanted with a Sorin Bicarbon 33 mechanical prosthesis. The postoperative course was uneventful. The patient in good general condition was discharged home and was given some recommendations to follow up. The case described emphasizes the importance of long-term cardiological care for patients with Marfan syndrome after aortic surgery. Progressive degenerative changes resulting from defective connective tissue structure may also contribute to the development of pathology in other heart valves. To improve the prognosis in this group of patients, regular echocardiographic monitoring of all heart structures and individualization of treatment strategies are necessary.

Keywords: Marfan syndrome, Bentall de Bono, cardiac surgery, mitral valve

1. INTRODUCTION

Marfan syndrome (MFS; OMIM #154700) is a genetic connective tissue disorder. The pathogenic FBN1 gene is located on chromosome 15q21 and is inherited in an autosomal dominant manner with variable penetrance (Zeigler et al., 2021). The

disease prevalence is 6.5/100,000 (Andersen et al., 2022). The FBN1 gene, located on chromosome 15q21, encodes a protein – fibrillin - which is one of the main components of the extracellular matrix (in the form of microfibrils), which in turn is an essential supporting element of connective tissue. It plays a special role in the arteries, the cartilage, and the lens of the eye (Milewicz et al., 2021). For this reason, MFS is characterised by pleiotropic clinical expression, with a particular predilection for the cardiovascular, skeletal, and visual systems (Milewicz et al., 2021; Loeys et al., 2010). Over 3,000 mutations have been identified in the FBN1 gene, most of which are unique to a given patient or family. Up to 25% of pathogenic variants arise de novo (Milewicz et al., 2021; Groth et al., 2015).

The main complications of MFS affect the cardiovascular system and are the most frequent cause of morbidity and mortality in these patients (Judge et al., 2005). Initially, asymptomatic enlargement of the thoracic aortic root is most common, which tends to progress and may lead to aortic dissection in the future, which is a direct threat to life (Milewicz et al., 2021; Judge et al., 2005). Among the observed changes, we also distinguish mitral valve prolapse (MVP) that can co-occur with or without mitral regurgitation - which is one of the clinical criteria for the diagnosis of the disease (Judge and Dietz, 2005). Aortic ring dilation (and secondarily, aortic valve regurgitation), peripheral vascular dilation, as well as cardiac arrhythmias, are observed slightly less frequently (De Backer, 2009). It has been shown that men with MFS are being diagnosed with development of aortic diseases at a younger age comparing to the women with MFS (Vanem et al., 2018). Aortic aneurysm rupture is the most frequent cause of death in patients with MFS (Carbone et al., 2023). Other common complications observed in MFS affect the musculoskeletal system – excessive growth, joint hypermobility, scoliosis, chest deformities and hip joint protrusions (Andersen et al., 2022; Pollock et al., 2021). Ophthalmic complications most commonly include lens displacement (ectopia) that is often accompanied by iridodonesis and also advanced myopia. These conditions usually require surgical treatment (Akram et al., 2021).

The Bentall de Bono procedure involves simultaneous replacement of the aortic bulb, ascending aorta and aortic valve, with reimplantation of the coronary vessels (Wagner et al., 2019). Although this operation is a recommended and effective method of treating ascending aorta pathology and often coexisting aortic valve regurgitation, it should be emphasized that in patients with MFS, the connective tissue defect associated with the FBN1 gene mutation predisposes to the progression of pathological changes throughout the cardiovascular system, including the mitral valve (Loeys et al., 2010; Wagner et al., 2019). However, progressive mitral valve dysfunction requiring surgical intervention after Bentall de Bono surgery is rarely reported in the literature, and most reports focus on the long-term results of aortic treatment (Loeys et al., 2010).

This article describes a case of a patient with MFS who underwent MV replacement after a previous Bentall de Bono procedure, a clinically rare procedure. The decision to replace the MV, especially after a last aortic surgery, reflects the advanced stage of the disease and the need to individualize the cardiac surgery treatment strategy.

2. CASE PRESENTATION

A 30-year-old man, 205 cm tall, weighing 77 kg, was admitted to the Cardiac Surgery Clinic with confirmed Marfan syndrome based on the Modified Ghent Criteria. Genetic testing was unsuccessful both nationally and internationally (tests were performed in Poland and the Netherlands). The patient had no addictions and complained of increasing exertional dyspnoea (NYHA II). Severe MV regurgitation was confirmed by echocardiography. Medical history: Bentall de Bono surgery (2013) at the age of 18 due to aneurysmal enlargement of the aortic bulb. The procedure itself was performed preventively. The patient remained under constant cardiological supervision.

Table 1. Results of echocardiography before the surgery.

RV		RVIT		LA area	52 mm2		4CH
IVS	9 mm	Ao root		RA area	35 mm2	EDV	461 ml
LV	82/76 mm	Ao asc	29 mm	LA Vol	350 ml	ESV	225 ml
PW	7 mm	LA	68 mm			EF	52%
IVS - Interventricular Septum; LV- Left Ventricle; PW - Posterior Wall; LA - Left Atrium; EDV – End Diastolic Volume; ESV- End Systolic Volume; EF – Ejection Fraction							

Transthoracic echocardiography (TTE) revealed severe MV regurgitation (ERO 72 mm², VC 8 mm, PISA r 13 mm, MR vol 109 ml, RF 50%, according to Carpentier II, I; Diastole: V max E: 1.3 m/s, Edec 187 ms, A: 0.45 m/s), with predominant anterior leaflet pathology (prolapse of segment A2/3), rupture of the chordae tendineae, enlargement (55 mm) and calcification of the mitral annulus (MAC) and

features of volume overload of the left ventricle (LVEDd 75 mm, LVEF 52%) and left atrium. Enlargement of the tricuspid annulus was demonstrated (59 mm; VC 3 mm, PISA r 6 mm, V max 2.87 m/s, P max 33 mmHg, RVSP 38 mmHg). The parameters measured in TTE before surgery are presented in Table 1.

After assessment by the Heart Team, due to advanced valve degeneration and unfavorable anatomy, a decision was made to replace the MV. The patient was assessed on the EuroSCORE II scale at 6.51%.

The course of the surgical procedure

Right femoral artery and vein were cannulated. Due to reoperation and increased risk, extracorporeal circulation was used before sternotomy (hypothermia -32^{0}C). Median sternotomy was performed, and the heart was dissected from adhesions. The aorta was clamped. Del Nido cardioplegia was used. The left atrium (LA) was opened. A fibrotic, enormous valve with a completely calcified mitral annulus was visualized, with no possibility of valve implantation within the annulus. A Sorin Bicarbon 33 valve was implanted, with felt strips on the left atrial side and the leaflets secured with Ethibond 2.0 patches. The left atrium was closed with a Prolence 3.0 suture. After venting the heart, the aorta was unclamped. Next, the right atrium (RA) was opened and a Carpentier Edwards MC3 36 ring was implanted, after which the RA was closed. The exit from extracorporeal circulation proceeded without complications. Decannulation was performed, hemostasis was achieved, and the chest was closed in layers. Three drains (one in the mediastinum, two in the right and left pleura) and electrodes for temporary cardiac pacing were left in place. The cardiac surgery lasted 320 minutes. Figure 1 shows intraoperative image of mitral valve in patient with MFS (A) versus in patient without MFS (B).

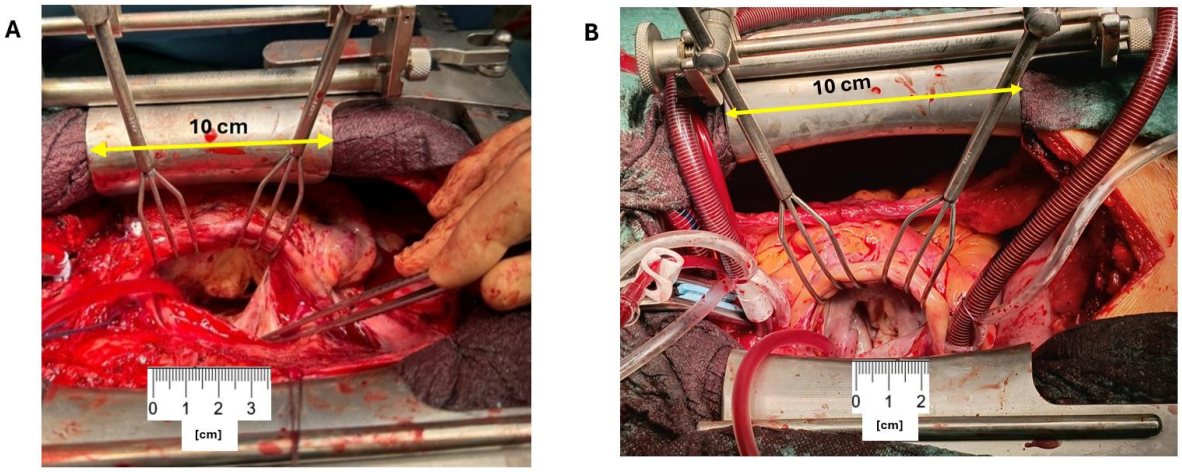

Fig. 1. Intraoperative MV image in the described case of Marfan syndrome (A), valve in a patient without a diagnosis of Marfan syndrome (B)

The intraoperative course was uneventful. The patient was admitted to the postoperative ward — haemodynamically stable, with moderate drainage and spontaneous diuresis preserved. Extubation took place 445 minutes after admission to the post-operative ward. After extubation, passive oxygen therapy was administered via nasal prongs, and no respiratory complications were reported.

Table 2. Results of echocardiography after the surgery.

RV	31 mm	RVIT	50 mm		4 CH
IVS	13 mm	Ao root	28 mm	EDV	390 ml
LV	77/72 mm	Ao asc	29 mm	ESV	302 ml
PW	11 mm			EF	23%
IVS - Interventricular Septum; LV- Left Ventricle; PW - Posterior Wall; LA - Left Atrium; EDV – End Diastolic Volume; ESV- End Systolic Volume; EF – Ejection Fraction					

On the sixth day after surgery, TTE was performed, which revealed akinesis of segments 3, 4, 9, 10, 14, 15, and hypokinesis of segments 2', 8', 13'; no signs of pericardial effusion. Mitral valve (mechanical prosthesis; V max E 1.6 m/s, P max 10 mmHg, Pmean 5

mmHg). Tricuspid valve (Pmean 1 mmHg; VC 2mm, V max 2.3 m/s, P max 20 mmHg, RVSP 35 mmHg). Parameters measured in TTE after surgery are presented in Table 2.

On the 11th day, the patient was discharged home in good general condition, with an INR of 2.49 and the following recommendations:

1. Systematic use of medication as prescribed. (INR min. 2.5)
2. Regular check-ups at the Cardiology Clinic.
3. Check-up at the Cardiac Surgery Clinic on the agreed date.
4. Intensive respiratory rehabilitation.
5. The patient was instructed on further care.
6. It is recommended to limit alcohol consumption and refrain from using tobacco products.
7. Diet adapted to patients after MV replacement surgery.

In addition to the above recommendations, the patient was to report to the cardiology clinic for a post-operative check-up.

3. DISCUSSION

The case presented above of a 30-year-old patient suffering from Marfan syndrome, who developed severe mitral valve regurgitation requiring replacement after previous Bentall de Bono surgery, is a case study in contemporary medical literature and poses a challenge for clinicians (Helder et al., 2014; Kunkala et al., 2013). Older patients with some additional diseases most often require reoperation rather than young patients, such as our Patient (David et al., 2025). Significant elongation and prolapse of the mitral valve leaflets, chordal rupture, enlargement and calcification of the mitral annulus which were observed on TTE and confirmed during the surgery are characteristic for myxomatous mitral valve disease (MMVD) that is documented cardiovascular manifestation of Marfan syndrome. Progressive connective tissue degeneration in the mitral valve apparatus, characteristic of MMVD, was probably the factor leading to severe mitral regurgitation in our patient, despite previous uncomplicated ascending aorta replacement surgery (Helder et al., 2014). MMVD is common in patients with MFS, but few patients treated with the Bentall procedure have mitral valve regurgitation severe enough to require surgical intervention (Burgstaller et al., 2018).

More information on the treatment of primary MV regurgitation in MFS can be found in the available literature, where valve repair is the preferred treatment, provided that anatomical conditions allow it (Wagner et al., 2019; David et al., 2025). Current calcification of the mitral annulus makes repair of such a valve more difficult, and in some cases even impossible (David et al., 2025). The case we describe involved progressive and advanced MV dysfunction developing after aortic surgery, which most likely prevented effective and permanent valve repair and ultimately necessitated the implantation of an artificial valve in the patient.

The decision to use a mechanical MV prosthesis in a 30-year-old patient required careful consideration of both the long-term benefits and risks. In younger patients, the prosthesis of choice is a mechanical valve, which is preferred due to its extended durability compared to bio prostheses, thus minimizing the risk of further reoperation in the future, which was particularly beneficial in the case of our patient (David et al., 2023; Vahanian et al., 2022). Disadvantage of using a mechanical prosthesis is fact that it requires chronic anticoagulant therapy and INR monitoring, and it is also associated with an increased risk of bleeding (Soria Jimenéz et al., 2023).

Available literature on long-term outcomes of Bentall de Bono operations in patients with MFS focuses on aortic outcomes with poor consideration to other valves, including tricuspid valve and the valve of pulmonary trunk – which without the doubt affects patient's overall hemodynamics (Burgstaller et al., 2018; David et al., 2023). There is insufficient number of studies that assess risk factors and the incidence of progressive mitral dysfunction in patients suffering from Marfan syndrome (Sama et al., 2024).

4. CONCLUSION

The case draws attention to the necessity of systematic monitoring patients with MFS after performed Bentall de Bono surgery, since they can develop a progressive mitral dysfunction at any time after the operation. It should be the standard to provide patients with MFS a long-term cardiological care and echocardiographic monitoring, especially with detailed assessment of all heart valves. Future studies should focus on identifying and describing risk factors for the development of severe MV regurgitation, such as specific FBN1 gene mutations or baseline left ventricular geometry. Prospective, multicenter registries of MFS patients undergoing Bentall surgery would be an extremely valuable source of knowledge, enabling the determination of the true incidence of this complication and the optimization of patient treatment strategies.

Abbreviations:
MFS – Marfan Syndrome
FBN1 – Fibrillin-1 gene
OMIM – Online Mendelian Inheritance in Man
MVP – Mitral Valve Prolapse
MAC – Mitral Annular Calcification
MMVD – Myxomatous Mitral Valve Disease
MV – Mitral Valve
MV regurgitation / MR – Mitral Regurgiation
LV – Left Ventricle
LA – Left Atrium
RV – Right Ventricle
RA – Right Atrium
RVIT – Right Ventricular Inflow Tract
EF – Ejection Fraction
EDV – End-Diastolic Volume
ESV – End-Systolic Volume
IVS – Intraventricular Septum
PW – Posterior Wall
TTE – Transthoracic Echocardiography
LA Vol – Left Atrial Volume
VC – Vena Contracta
PISA – Proximal Isovelocity Surface Area
ERO – Effective Regurgitant Orifice
RF – Regurgitant Fraction
LVEDd – Left Ventricular End-Diastolic Diameter
LVEF – Left Ventricular Ejection Fraction
RVSP – Right Ventricular Systolic Pressure
NYHA – New York Heart Association classification
ESC/EACTS – European Society of Cardiology/European Association for Cardio-Thoracic Surgery
INR – International Normalized Ratio
EuroSCOREII – European System for Cardiac Operative Risk Evaluation II
RA area/LA area – Right/Left Atrium Area
4CH – Four-Chamber View
Ao root – Aortic Root
Ao asc – Ascending Aorta
E/A ratio – Early-to-Atrial filling velocity ratio
Pmax/Pmean – Maximum/Mean Pressure Gradient
LVEDd/LVESd – Left Ventricular End-Diastolic/Systolic Diameter
RA area/RV diameter – Right Atrium area/Right Ventricle diameter

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All authors have read and agreed with the final published version of the manuscript.

Informed consent
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Conflict of interest
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Data and materials availability
All data associated with this study will be available based on the reasonable request to corresponding author.

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