

One-Stage Surgical Repair of Heart Disease and Pectus Excavatum in Marfan Syndrome Strategy about the First Case Performed in the UAE

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Abstract

The association of pectus excavatum and cardiac anomalies in patients with Marfan syndrome is not exceptional. The surgical treatment is always challenging. The first case performed in the UAE of a complex surgery in a one-stage procedure with simultaneous complex cardiac valve repair (valve-sparing aortic root replacement associated with mitral valve repair) and pectus deformity correction is reported. An update of the surgical strategy is proposed.

Keywords: Aorta, Marfan syndrome, mitral valve regurgitation, pectus excavatum

INTRODUCTION

In Marfan syndrome (MFS), the association of pectus excavatum and cardiac anomalies is not exceptional.^[1] Surgical strategy has been controversial regarding the repair of chest deformity and heart disease.^[2] The first case performed in the UAE of such a complex repair is reported, and an update of the surgical strategy is proposed.

CASE REPORT

A 29-year-old male with a known history of MFS was referred to our institute for surgical assessment. The patient complained about shortness of breath, palpitations as well as psychological impact owing to chest wall deformity with a severe symmetric pectus excavatum [Figure 1]. Auscultation detected severe systolic mitral murmur. Echocardiography revealed a severe mitral valve regurgitation due to posterior mitral valve prolapse and a 5-cm aortic root dilation with a normal aortic valve. Pre-operative computed tomography (CT) scan confirmed the severity of the chest deformity and a critical dilation of the aortic root [Figures 2 and 3]. The patient was proposed for a one-stage combined surgical repair of pectus excavatum and valve lesions through a sternotomy approach.

First, a sub-perichondrial resection of the depressed costal cartilages, following the modified Ravitch technique,^[3] was performed to remove all abnormal costal cartilages bilaterally from the fourth to the seventh ribs. Then, the cardiac lesions were approached through a median sternotomy. The mitral valve was exposed first with a trans-septal approach, and a mitral valve repair was done with a simple plication on P2 and an annuloplasty using a 38-mm complete ring. Then, a valve-sparing aortic root replacement according to David Procedure was carried out using a 30-mm Gelweave Valsalva™ Conduit Graft. Finally, after the closure of the sternum, a temporary retrosternal titanium plate was placed at the level of the sixth intercostal space and fixed bilaterally to the ribs providing chest stability and keeping the sternum up. The post-operative outcome was event free.

Two months after the surgery, CT scan showed an evident improvement of the chest deformity and a remarkable decompression of the right cardiac chambers [Figure 4]. Six

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Figure 1: Pre-operative image of the patient showing a severe and symmetric pectus excavatum



Figure 2: Pre-operative angiographic computed tomographic scans. Transverse cross section. The heart is displaced in the left hemithorax. Right cardiac cavities are compressed. Haller index is 7.8. Aortic root is dilated

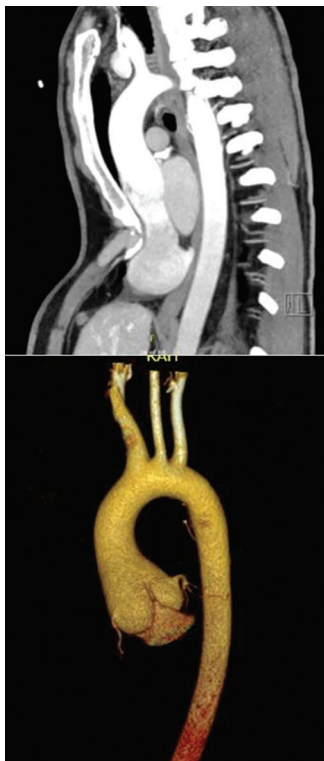


Figure 3: Pre-operative angiographic computed tomographic scan. Sagittal cross section. Severe pectus excavatum with compression of the aortic root. Three-dimensional reconstruction of the aorta showing an aortic root aneurysm >50 mm

months after the surgery, echocardiography demonstrated a good cardiac function without residual aortic or mitral regurgitation. Then, the retrosternal plate was removed under general anaesthesia from a right side skin incision with a stable and satisfactory cosmetic result [Figure 5].

DISCUSSION

MFS is a severe, chronic, life-threatening disease with multiorgan involvement including the skeletal and cardiovascular systems.

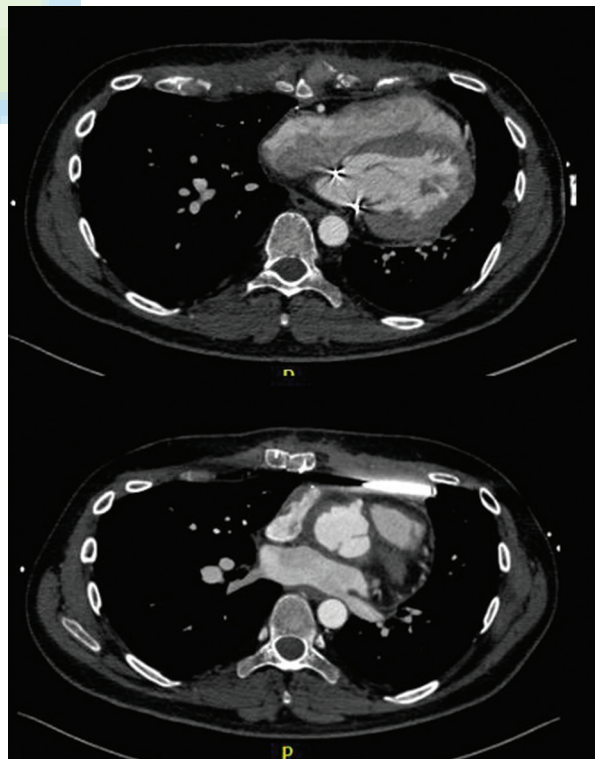


Figure 4: Post-operative angiographic computed tomographic scans. Transverse cross section showing the corrected thoracic deformity (Haller index of 3.2) without any residual compression of the heart. The aortic root has a normal diameter according to the graft implanted



Figure 5: Post-operative image of the patient showing the corrected thoracic deformity with a satisfactory cosmetic result

It is well known that poor life expectancy in MFS is mainly triggered by cardiovascular complications. In most cases, aortic root dilation is the predominant aortic manifestation. To avoid acute aortic syndromes, prophylactic surgery of the aortic root is recommended in patients with MFS who have a maximum ascending aortic diameter of ≥ 50 mm or ≥ 45 mm in case of risk factors.^[4] Although composite graft replacement with an aortic valve prosthesis as Bentall procedure remains the standard, most studies have shown excellent results using valve-sparing root replacement,^[5,6] and nowadays, valve-sparing procedure is recommended in young patients.^[4] The David re-implantation technique is more appropriate in MFS because the aortic annulus is more stabilised with less long-term aortic insufficiency.^[7,8]

Mitral valve dysfunction is common in MFS and mainly related to valve prolapse.^[9] Interestingly, survival and risk of reoperation after mitral valve repair are similar in MFS and degenerative disease, and in MFS, survival is better after mitral valve repair than replacement.^[10] The cardiac strategy developed in the present case, in respect of the guidelines, with concomitant David procedure and mitral valve repair has confirmed the good results of valve preservation techniques in MFS.^[11]

Pectus excavatum is often associated with a shift of cardiac structures in the left thorax and/or their mechanical compression by the depressed chest wall.^[12] During cardiac surgery, the chest deformity involves difficulty in dividing the sternum in the midline, in retraction symmetrically and in cardiac exposure;^[13] moreover, when the pectus deformity is not addressed, the compression of the heart by pectus deformity may contribute to post-operative hemodynamic instability. One-stage procedure with simultaneous cardiac and pectus deformity repair has been performed successfully.^[14,15] In children, cardiac surgery with left thoracic approach and combined with a Nuss procedure was reported.^[16] In adults, chest wall is less flexible and sternotomy approach is more appropriate, especially to reach the mitral valve; however, the

risk of post-operative sternal dehiscence is a matter of concern when a Nuss procedure is combined with.^[14] Consequently, in the present case, a modified Ravitch procedure was done first to facilitate the sternotomy and the exposure of the cardiac structures. The temporary titanium plate was useful to provide firmness to the chest wall according to the Wurtz technique.^[17]

CONCLUSION

In patients with MFS, when cardiac anomalies are associated with pectus excavatum and their correction is mandatory, single-stage repair of both lesions is currently the method of choice. As an alternative to Nuss procedure, modified Ravitch procedure and median sternotomy provide a good exposure of the cardiac lesions and make their repair easy in respect to the current guidelines. Modified Ravitch procedure also allows to correct the chest wall deformity successfully without compromising the post-operative stability of the sternum, as it has been demonstrated in the reported case.

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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Conflicts of interest

There are no conflicts of interest.

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