

## Case Report

# Surgical approach of bentall procedure in a patient of pectus excavatum

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### ABSTRACT

A middle-aged man diagnosed case of Marfan syndrome associated with pectus excavatum presented with chest pain and dyspnea. Chest X-ray, transthoracic echocardiography and Computed tomography (CT) of heart and aorta revealed severe Aortic regurgitation with dilated aortic root, sinotubular junction and ascending aorta with normal size arch and descending aorta. Patient was taken for surgery. Pectus excavatum creates difficulties for heart exposure and cannulation for cardiopulmonary bypass. We planned for femoro-femoral bypass to carry out ahesiolysis and Bentall procedure without much difficulties. Postoperative stay of the patient was uneventful and followed up in regular interval.

**Keywords:** Ascending aorta, Bentall procedure, Cardiopulmonary bypass, Marfan syndrome, Pectus excavatum

### INTRODUCTION

Marfan syndrome is a genetic disorder that affects the body's connective tissue. This disease is autosomal dominant and it is caused by a mutation in the FBN-1 gene.<sup>1</sup>

FBN-1 gene encodes an extracellular matrix glycoprotein, named fibrillin 1, which is essential for the formation of the elastic fibers and microfibrils, that provide strength and flexibility to the connective tissue. This mutation results in production of abnormal elastic fibers.<sup>2</sup>

The most complications in patients with Marfan syndrome are due to defect in the tunica media of the blood vessels, leading to the formation of aortic aneurysms.<sup>3</sup> The wall of the aorta becomes progressively weaker as the aneurysm enlarges. If the aneurysm is left untreated, it may rupture and cause death from cardiac tamponade.<sup>4</sup> In many patients with Marfan syndrome osteoarticular manifestations occur, such as pectus excavatum or pectus carinatum. Marfan syndrome can

also seriously affect the eyes and vision.<sup>5</sup> Aortic root replacement combined with pectus repair is uncommon.<sup>6</sup>

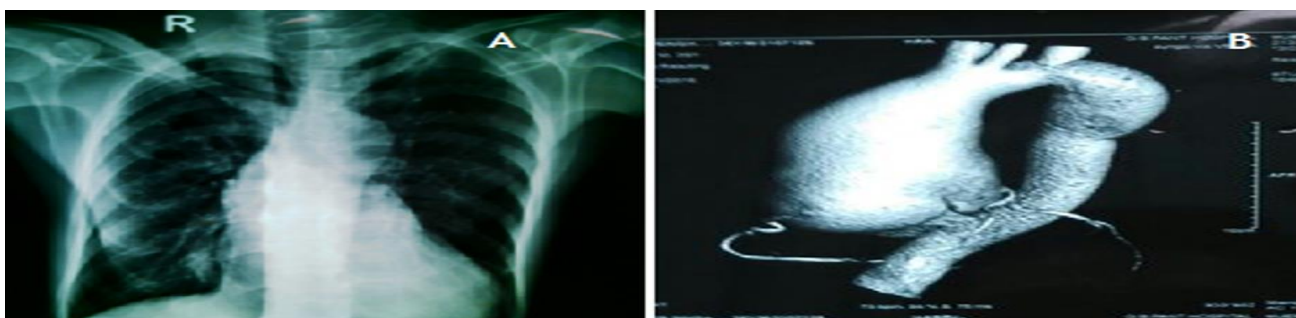
Cardiac surgical correction requires adequate exposure, which is particularly difficult with coexisting pectus excavatum. We report a case of different surgical approach of Bentall procedure in pectus excavatum deformity.

### CASE REPORT

A 38-year-old male with Marfanoid habitus and pectus excavatum (Figure 1A, B, C, D, E) presented with left side chest pain and NYHA class III Dyspnea. There was presence of all peripheral signs of Aortic regurgitation. Chest X-ray revealed cardiomegaly with dilated ascending aorta (Figure 2A). Transthoracic echocardiography and Computed tomography (CT) of heart and aorta revealed severe Aortic regurgitation with dilated aortic root (8.1cm) sinotubular junction (6.2cm) and mid ascending aorta (5cm) with normal size arch and descending aorta (Figure 2B).



**Figure 1: A) Pectus excavatum, (B, C, D): arachnodactyly, (E): high arched palate.**



**Figure 2: A) Chest X ray showing cardiomegaly with dilated aortic root and ascending aorta, B) CT angiography showing dilated aortic root and ascending aorta.**

Patient was taken for surgery. Midline sternotomy was done in a curvilinear fashion according to the shape of sternum. There was a large size aorta occupying most of the pericardial cavity, displacing the heart leftward and downward giving very little exposure of heart to the surgeon inside the pericardial cavity. In addition to this, the 11-pericardial adhesion and space constraints in pericardium due to pectus excavatum denied easy access for cannulation. In such situation it was prudent to institute femoro - femoral bypass to carry out atherosclerosis and Bentall procedure without much difficulties.

The Bentall procedure was performed using a mechanical prosthesis (31 mm). The distal part of the 1 conduit was anastomosed with the ascending aorta using a continuous 3/0-prolene suture reinforced with Teflon felt strips. Gradually weaning from cardiopulmonary bypass. Immediate post-operative period was uneventful. Patient was extubated on day 1. Patient was discharged from hospital on day 9 in a stable condition and he was followed up in a regular interval.

## DISCUSSION

The patients with Marfan syndrome, pectus excavatum, and associated aortic or cardiac surgery poses a major clinical challenge. There is a definite association of cardiovascular anomalies with thoracic skeletal

deformities in patients with Marfan syndrome. The main issue in such cases is how to approach surgically in pectus excavatum and any cardiac disease because of the difficulties that may result from the cardiac displacement into the left thoracic cavity. In patients with Marfan syndrome commonly affect the structure of the aorta and the function of the heart valves. Reports of combined operations in patients with Marfan syndrome are rare. Many studies have shown that these patients are often enrolled into staged procedures in order to manage their conditions.<sup>7,8</sup>

In our case, we decided to go cardiopulmonary bypass through femora-femoral bypass and then sternotomy done. Adequate cardiac exposure was achieved and Bentall procedure done without much difficulties.

## CONCLUSION

Marfan syndrome is associated with many disorders that affect different systems of the body, these patients always require an individual approach. Femoro – femoral bypass prior to main surgical procedure makes surgery in easy way.

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