Aortic and mitral valve surgery through a superior ministernotomy in pectus excavatum associated with Marfan’s syndrome

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Received 7 July 2002; received in revised form 10 December 2002; accepted 29 December 2002

Abstract

The combination of chest wall abnormalities such as pectus excavatum and cardiac disease requiring operative correction represents a clinical challenge to the surgeon. We report two cases of patients with Marfan’s syndrome and severe pectus excavatum in whom cardiac operations were successfully performed using a superior ‘T inverted’ ministernotomy. This approach allowed safe isolation of the target cardiac structures, provided excellent operative exposure and enhanced chest wall stability by preserving the integrity of the lower sternum.

Keywords: Valve surgery; Pectus excavatum; Ministernotomy

1. Introduction

The combination of pectus excavatum and cardiac disease requiring operative correction represents a clinical challenge to the surgeon. In this setting, full midline sternotomy as well as sternal reconstruction is technically cumbersome. Furthermore, patients with severe chest deformity may develop postoperative respiratory problems and a better preservation of the integrity of the thoracic cage is highly desirable under these circumstances. Finally, in redo cases, opening of the inferior part of the chest can lead to inadvertent cardiac structure injuries due to the contact of the heart and great vessels with the lower portion of the sternum. We report two cases of patients with Marfan’s syndrome and severe pectus excavatum in whom cardiac operations were successfully performed using a superior ‘T inverted’ ministernotomy, which allowed excellent exposure of the target cardiac structures.

2. Case report 1

A 16-year-old male with Marfan’s syndrome and severe pectus excavatum was admitted because of heart failure. Serial echocardiography had demonstrated an ascending aorta aneurysm that rapidly increased from 5.5 to 7.4 cm in diameter over the last 13 months. Severe aortic and mitral insufficiency was present as well. A computed chest tomography (CT) showed marked deviation of the heart into the left chest and severe posterior displacement of the sternum which was in direct contact with the cardiac structures and great vessels (Fig. 1). His chest deformity was responsible for a moderate obstructive respiratory dysfunction. He was referred for urgent surgery. At operation, a ‘T inverted’ upper ministernotomy was carried out to the fourth intercostal space. The femoral artery and vein and the superior vena cava were cannulated and hypothermic cardiopulmonary bypass was instituted. The distal ascending aorta was clamped, the aneurysm was opened and cold blood cardioplegia was delivered directly into the coronary ostia. The mitral valve was approached through a conventional left atriotomy after dissection of the interatrial groove. Because of the severe left atrial enlargement, the exposure obtained was satisfactory and the mitral valve was replaced with a 33 mm SJM mechanical prosthesis. After atrial closure, the aortic root was replaced with a composite graft consisting of a 25 mm SJM valve and a 26 mm Hemashield graft. Both the left and right coronary arteries were reimplanted in the aortic conduit. The sternum was approximated with figure of eight interrupted heavy
stainless steel wires (Fig. 2). The postoperative course was uneventful and the patient was discharged on the 12th postoperative day with preserved thoracic stability and uncomplicated wound healing. At 6 months he is symptom-free and echocardiography is unremarkable.

3. Case report 2

A 41-year-old female with Marfan’s syndrome and severe pectus excavatum was referred to our institution for urgent mitral valve repair/replacement after an episode of heart failure due to severe mitral regurgitation. Transesophageal echocardiography confirmed the presence of severe mitral insufficiency in the context of typical Barlow’s disease. Left ventricular end diastolic diameter was 74 mm and ejection fraction 36%. CT scan demonstrated complete deviation of the heart into the left chest and severe displacement of the sternum just above the underlying heart vessels and chambers. Previously performed lung test function showed moderate obstructive respiratory dysfunction. The patient, in chronic atrial fibrillation, was progressively developing pulmonary edema despite intravenous therapy with furosemide, dopamine and nitrates. Therefore, she had to be operated on an urgent basis. At operation, the femoral vessels were exposed for cannulation. The thorax was opened through a ‘T inverted’ ministernotomy extended to the fourth intercostal space. The superior vena cava was cannulated, standard cardiopulmonary bypass instituted and the myxomatous mitral valve successfully repaired with the ‘edge-to-edge’ technique. The postoperative course was uneventful and the patient was discharged 5 days after surgery. At 6 months, her clinical conditions are very satisfactory and the mitral valve is functioning well.

4. Discussion

Cardiac valve operations in the presence of chest wall deformities such as pectus excavatum remain challenging. The posterior displacement of the sternum, the asymmetric angulation that it produces and the marked deviation of the heart into the left chest limit operative exposure. Moreover, such a funnel-shaped depression of the sternum is often lying just above the heart vessels and chambers. This makes a full median sternotomy technically difficult and, in redo cases, potentially dangerous because of the risk of cardiac structure injuries. Although definitive repair of pectus excavatum should ideally precede a cardiac procedure for optimal functional and cosmetic results, several authors have described techniques for concomitant repair of pectus deformities at the time of cardiac operation [1–3]. However, in the cases here reported, we preferred to correct the cardiac disease first. Indeed, the first patient was scheduled for a Bentall procedure associated with a mitral valve repair. To reduce as much as possible the duration of the operation and the risk of postoperative bleeding, we preferred to avoid the simultaneous repair of the pectus excavatum and of the cardiac disease. In the second case, because of the rapid decompensation of the underlying cardiac abnormality, mitral valve repair had to be performed on an urgent basis and there was no time to plan a concomitant correction of the chest deformity together with the thoracic surgeon, as we usually do. From a surgical perspective, a ‘T inverted’ ministernotomy down to the
fourth intercostal space [4] was used. To the best of our knowledge, this alternative approach has never been reported in this setting, although, in our opinion, it can be most beneficial. Besides the obvious advantages in terms of patient comfort and cosmetic results, it provides a satisfactory operative exposure and enhances chest wall stability. Preservation of the integrity of the lower sternum may improve postoperative pulmonary function promoting earlier extubation [5] and a smooth postoperative course. Moreover, in redo cases, it allows safe isolation of the target cardiac structures preventing the risk of cardiac injury and intraoperative hemorrhage. This superior ministernotomy incision can be adopted in patients with Marfan’s syndrome and severe pectus excavatum requiring cardiac surgery as a safer and more effective approach than standard median sternotomy whenever the cardiac operation needs to be performed first.

References