Management of severe asymmetric pectus excavatum complicating aortic repair in a patient with Marfan’s syndrome

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Abstract

We describe the case of a 28-year old man with Marfan’s syndrome and severe pectus excavatum who required an aortic root replacement for an ascending aortic aneurysm. There was a near-vertical angulation of the sternum that presented challenges with opening and exposure of the heart during aortic surgery. Furthermore, removal of the sternal retractor after aortic repair resulted in sudden loss of cardiac output. A Ravitch procedure was then performed to successfully close the chest without further cardiovascular compromise. We propose that patients with a severe pectus excavatum and mediastinal displacement seen on preoperative CT scanning should be considered for simultaneous, elective repair.

Keywords: Marfan • Pectus excavatum • Aortic • Aortic root

CASE PRESENTATION

A 28-year old patient with Marfan’s syndrome was found to have progressive aortic root enlargement to 5 cm. Elective repair was recommended. His preoperative CT scan demonstrated a severe asymmetric pectus excavatum (Haller index of 11.8). His sternum was angled almost vertically due to an abnormal curvature of the anterior arc ribs on the right, with a maximum tilt of ≏75°. This resulted in significant cardiac luxation (Fig. 1).

The near-vertical sternal tilt resulted in technical challenges during initial sternotomy. This was managed by manually reducing the deformity with a handheld retractor, allowing the sternal saw to be used. Due to the extreme tilt in the right hemisternum, two sternal retractors were required. A smaller retractor was used for the manubrium, and a deeper thoracotomy retractor was used for the angulated lower sternum. Afterwards, a standard valve sparing aortic root replacement with a reimplantation technique was performed. Post-repair transoesophageal echocardiography demonstrated normal ventricular function with no aortic insufficiency.

However, following removal of the sternal retractor, there was sudden loss of cardiac output, resulting in near arrest. Pressure largely from the right hemisternum caused direct compression of the right heart, and subsequent reduction in preload. Cardiac output promptly recovered when the sternal pressure was relieved. A Ravitch procedure was therefore performed with constant upward traction to relieve right ventricular compression. In brief, bilateral pectoralis muscle flaps were raised, and the costal cartilages from rib 3 to 8 were resected, taking care to preserve the perichondrial sheaths. Once the sternal depression was corrected, haemodynamic instability was no longer observed. The sternotomy was closed with steel wires, and reduction was maintained with an 8” substernal bar (BioMet Pectus Bar, Part 01-3808, Jacksonville, Fl, USA). No significant bleeding occurred either intraoperatively or postoperatively. His postoperative course was uneventful; subpectoral drains were removed on Day 3, and he was discharged on Day 6. A follow-up CT scan demonstrated satisfactory reduction in the pectus deformity with a stable aortic repair (Fig. 2).

COMMENT

Simultaneous pectus excavatum and aortic pathology is common, and the sequence of repair remains controversial in the literature. Initial pectus surgery is theoretically problematic since a substernal bar could complicate emergency exposure for dissection/rupture [1]. Alternatively, pectus repair after aortic surgery may be potentially dangerous due to postoperative mediastinal adhesions. Indeed, we would consider only the Ravitch procedure for this strategy; adhesions would be prohibitively risky for the Nuss procedure as described by Craner et al. [2].

It is becoming apparent that simultaneous repair is gaining traction in the literature. Prior concerns regarding blood loss, increased operative time and complications have not been observed in recent reports [3, 4]. While we chose to perform a Ravitch repair following separation from cardiopulmonary bypass (CPB), other groups have advocated alternate strategies of repair. Rousse et al. [3] have proposed performing first a subperichondrial resection of...
this phenomenon was due to cardiac luxation into the left chest by the pectus excavatum deformity (Fig. 1). We hypothesize that the mediastinum was allowed to return to the midline after sternotomy and exposure for aortic repair. Therefore, when the retractor was subsequently removed, the more deformed right hemisternum directly compressed the right ventricle. This compression was relieved by the correction of the excavatum (Fig. 2). This phenomenon was also observed by Stephens et al. [5], and in their report, an unplanned, emergency Ravitch procedure was performed.

It is presently unclear as to what are the specific risk factors for haemodynamic collapse following sternotomy in a patient with severe pectus deformity. It appears that the common features between the two cases include (i) significant mediastinal shift resulting in cardiac luxation and (ii) Haller index of >10. It is possible that the presence of either of these risk factors may predict for haemodynamic compromise upon sternal closure. We suggest that patients with severe pectus excavatum undergoing cardiovascular surgery should be considered for simultaneous repair with a planned collaboration of both thoracic and cardiac surgeons.

Conflict of interest: none declared.

REFERENCES


eComment. Simultaneous repair of pectus deformities and cardiac surgery under cardiopulmonary bypass

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Yeung et al. report the management of severe pectus excavatum (PEx) at the time of an aortic repair in a patient with Marfan’s syndrome and the difficulties encountered during the procedure [1]. As underlined by the authors, the Ravitch-type repair is the more reliable technique in the setting of concomitant pectus deformity correction and cardiopulmonary bypass surgery [2-4]. In the case of PEx, we usually carry out a staged repair of the pectus deformity by first performing the subperiosteal resection of the enlarged/deformed costal cartilages (third to sixth and extremities of seven). Once these paired cartilages are resected, the sternum can be easily elevated, thus facilitating sternotomy and placement of a standard sternal-retractor allowing satisfactory operative exposure. Cardiac procedure with CPB is then performed. Last, after heparin reversal and sternal closure as usual, the PEx repair is achieved by performing additional resection of the second cartilages, if needed, transverse wedge osteotomy across the anterior table of the upper sternum and placement of a straight metal support bar positioned anterior to the ribs laterally and secured to sternum with an absorbable suture [3]. Finally, in severe asymmetric PEx, such as that reported