

Aortic Surgery in a Patient with Marfan Syndrome and Pectus Excavatum

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Severe cardiovascular disorders, including aortic dissection and aneurismal dilation of the aorta, are the main life-threatening complications of the Marfan syndrome¹. Approximately two thirds of individuals who have this syndrome have chest wall deformities such as pectus excavatum or pectus carinatum². When a patient with pectus excavatum needs aortic surgery, the surgeon may face a major clinical challenge in choosing the optimal surgical approach³. We present a case of the Marfan syndrome with severe pectus excavatum who underwent aortic surgery (*Iranian Heart Journal* 2012; 13 (1):55 -58).

Case presentation

A 33-year-old man was admitted because of new-onset severe chest and back pain from about 24 hours earlier. The pain radiated to the abdomen and the left lower limb. General physical examination revealed features of the Marfan syndrome (tall stature, increased arm span, arachnodactily, and flat feet).

Severe deformity of the chest wall in the form of pectus excavatum (Fig.1), bounding pulses of the right upper limb, and diminished pulses of the lower limbs were observed.

No abnormality in the other organ systems was found.

Transthoracic and transesophageal echocardiographic examinations revealed severe left ventricular enlargement with moderate to severe dysfunction (left ventricular ejection fraction =30-35%) and a tricuspid aortic valve with severe regurgitation. Also, there was an ascending aortic aneurysm (6.9 cm in diameter), which terminated before the origin of the innominate artery, and the dissection of the descending thoracic aorta with the intimal flap originated just after the left subclavian artery.

Multi-slice CT scanning confirmed the diagnosis of descending thoracic aortic



dissection, extending to the aortic bifurcation.
Fig. 1. Severe pectus excavatum with posterior deviation of sternal body

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It demonstrated the extreme displacement of the mediastinal structures into the left hemithorax and also showed that the sternal body almost touched the vertebral column.

The simultaneous existence of two life-threatening complications of the Marfan syndrome and also the severe deformity of the chest wall prompted a debate over the best timing for surgery and the most optimal approach to the patient's management. The patient refused emergent surgical treatment; consequently, medical management was commenced based on the control of blood pressure and monitoring of the function of the vital organs. The patient was discharged after one week and was scheduled to undergo open heart surgery four weeks afterward.

On second admission, repeat multi-slice CT scanning was performed. Fortunately, the dissection of the thoracic aorta had not extended and the size of the aorta had started to decrease. The evaluation of the respiratory system revealed restrictive lung disease and it did not require any preoperative treatment.

A thoracic surgeon was consulted and he stated that combined thoracic wall reconstruction and heart operation would increase the risk of morbidity and mortality remarkably and, thus, recommended performing the heart operation first.

Opening of the thorax was undertaken by an anterior left-sided trap-door (second through sixth intercostal spaces) thoracotomy incision (Figs. 2, 3).

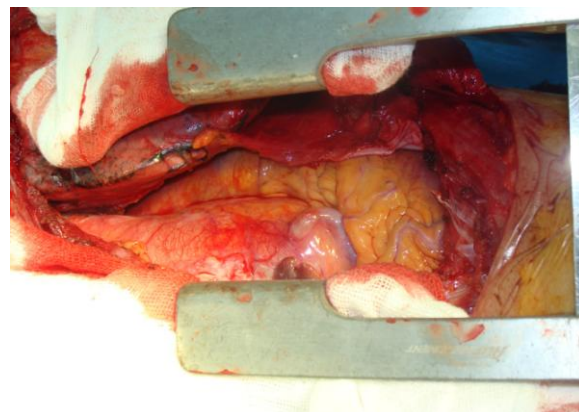


Fig. 3. The anterolateral left thoracotomy provides excellent exposure of the heart, aortic root and the entire ascending aorta (surgeon's view).

The patient was extubated after seven hours. His recovery course was uneventful and he was discharged from the hospital on the seventh postoperative. We recommended pectus excavatum repair after six months, but the patient did not wish to undergo a second operation.

The cannulation of the distal ascending aorta and the right atrium and establishment of cardiopulmonary bypass (CPB) with moderate (28°C) hypothermia was established. Replacement of the aortic root and ascending aortic aneurysm (Bentall Operation) with a valved conduit was accomplished in a standard fashion. Weaning from CPB was achieved with acceptable hemodynamic parameters. Intraoperative transesophageal echocardiography demonstrated good valve function without any paravalvular leakage or abnormal gradient, and the other echocardiographic indices were within the normal range. Total pump time was 176 minutes and the aortic cross-clamp time was 127 minutes. Eighteen months after surgery, his clinical condition was satisfactory and he exhibited no symptoms. The results of cardiac and pulmonary tests were acceptable.

Fig. 2. The incision line is demarcated. Cross sign (X) denotes the level of the aortic root.

Patients with severe chest wall deformity who need cardiac surgery are challenging. Deviation and posterior displacement of the sternum, as well as extreme shift of the heart and great vessels to the left hemithorax, renders the standard median sternotomy a very difficult and almost useless approach.

There are some reports in the literature regarding different techniques for cardiac surgery in patients with pectus excavatum. Westaby and colleagues⁴ presented the feasibility of partial upper sternotomy for aortic root replacement in patients with the Marfan syndrome, but they did not recommend it as a routine practice. Molina et al.⁵ described a combined sternotomy and anterolateral thoracotomy in a "trap door" configuration. Nisanoglu et al.⁶ addressed a partial upper sternotomy extended to the left hemithorax. Both cases were performed for the correction of the ascending aortic aneurysm without simultaneous repair of the pectus excavatum.

In our case, we chose an anterolateral thoracotomy, starting in the second left intercostal space. The thoracotomy was thereafter extended vertically through the division of the ribs, third through sixth, near their costochondral junctions, and ended in the sixth intercostal space. This incision provided excellent exposure of the heart and the ascending aorta. The sternum was left intact so as not to complicate a later chest wall reconstructive surgical operation.

Another important issue is whether to combine cardiac surgery with sternal deformity correction or choose the staged repair. Javangula and colleagues⁷ reported a one-stage Bentall operation and correction of the pectus excavatum in a patient with the Marfan syndrome. Schwill et al.⁸ presented an alternative surgical approach for the combined surgery of the pectus excavatum and acute aortic dissection type-A in the

Marfan syndrome. These reports show the feasibility of a combined operation; however, they may underestimate the relevant risk imposed on the patient. It is clear that concomitant repair of such a deformity, as was the case in our patient, would prolong the operation significantly. It invariably increases the risk of bleeding and the potential of infection and pulmonary complications. Therefore, we deemed it prudent to postpone the repair of the chest wall deformity. We conclude that a surgical approach through a left anterolateral "trap door" thoracotomy not only provides an excellent exposure but can also be performed with minimal complications in the aortic surgery of patients with the Marfan syndrome and pectus excavatum. Deferring chest wall repair will not adversely influence the postoperative course of cardiac surgery.

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