

Simultaneous Repair of Pectus Excavatum and Tetralogy of Fallot: Report of a Case

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We report an 18-month-old boy with the association of pectus excavatum and tetralogy of Fallot (TOF). We successfully performed simultaneous pectus repair using sternal elevation without any prosthetic support and total correction of TOF after a prior modified Blalock-Taussig shunt. Retracting a divided costo-sternal complex with a rectus abdominal flap away from the operative field before the cardiac operation provided excellent surgical exposure. The modified Blalock-Taussig shunt prior to the combined repair prevented life-threatening hypoxic spells during dissection of the deformed sternum and costochondral cartilages before institution of cardiopulmonary bypass. (Ann Thorac Cardiovasc Surg 2005; 11: 320–3)

Key words: pectus excavatum, tetralogy of Fallot

Introduction

The association of pectus excavatum with congenital heart disease is uncommon¹⁾ and there are only two reports describing pectus excavatum associated with tetralogy of Fallot (TOF).^{1,2)} Most reports on repair of intracardiac lesions associated with pectus excavatum relate to adults with Marfan's syndrome.^{3,4)} In children, the literature is sparse concerning the management of congenital heart disease associated with pectus excavatum. Traditionally, a combined repair of pectus deformities and coexisting heart defects has been avoided because of fear of technical difficulties of intracardiac repair or other major complications including bleeding.⁵⁾ Recently, there have been several English-language publications documenting successful simultaneous repair of pectus deformities and coexisting heart defects.^{2,6,7)} Here, we report an association of pectus excavatum and symptomatic TOF in an infant who underwent successful simultaneous pectus repair and intracardiac repair.

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Case Report

A male infant was born at 38 weeks' gestation and weighed 3,162 g. Shortly after birth he was noted to be cyanosed and to have pectus excavatum and was referred to another hospital. Echocardiography demonstrated TOF. He was given beta blockade and followed in the hospital outpatient department. At 11 months of age he was referred to our institution because of severe hypoxic spells. Since the spells were intractable and could not be controlled medically, an emergency left modified Blalock-Taussig shunt using a 4 mm expanded polytetrafluoroethylene graft through a left posterolateral thoracotomy was performed. At 18 months of age, he was readmitted for simultaneous definitive intracardiac repair and pectus repair. Physical examination showed pectus excavatum (Fig. 1) and mild cyanosis. A chest computed tomography (CT) scan revealed a depressed sternum and displacement of the heart in the left hemithorax (Fig. 2). The operation was performed through a midline skin incision over the sternum. The pectoralis major muscle insertions were elevated off exposing the sternum and the deformed cartilages. The normal sternum was divided transversely in the third intercostal space, at a point above the origin of any posterior angulation deformity. The perichondrium was bilaterally incised and peeled off from the 4th to the 7th costal cartilages. Bilaterally, all deformed cartilages were transected a few millimeters laterally from



Fig. 1. Preoperative view of the chest.

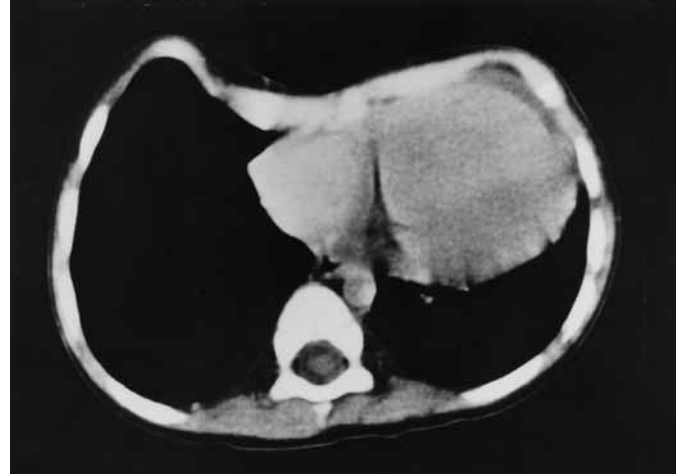


Fig. 2. Preoperative chest CT scan.

the sternochondral junction. The costo-sternal complex with a rectus abdominis muscle flap was then retracted away from the operative field caudally toward the abdomen to gain maximal exposure of the heart for cardiopulmonary bypass. The costo-sternal complex was maintained in that position covered by moistened sponges and wrapped with a nylon bag. The manubrium was longitudinally divided and widely spread with a retractor (Fig. 3). Exposure of the heart and great vessels was excellent, despite displacement of the heart into the left hemithorax by the chest wall deformity. A rectangular segment of autologous pericardium was harvested for use in the right ventricular outflow tract reconstruction. Cardiopulmonary bypass was started using an aortic cannula and bicaval venous drainage cannulae. The left modified Blalock-Taussig shunt was divided immediately after beginning cardiopulmonary bypass. The ascending aorta was cross-clamped and crystalloid cardioplegia was infused into the aortic root. Pulmonary valvotomy and infundibular resection was performed through a transatrial-transpulmonary approach. The ventricular septal defect was closed through a right atriotomy. The pulmonary arteriotomy was widened with a patch of the previously harvested autologous pericardium. After removal of the aortic cross-clamp, spontaneous sinus rhythm resumed and cardiopulmonary bypass was discontinued. After administration of protamine sulfate and cautious hemostasis, the fragments of the manubrium were wired together.

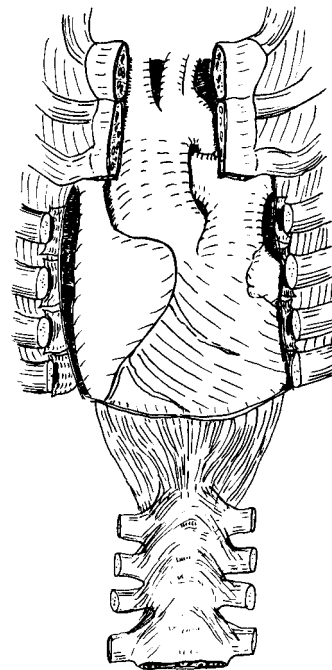


Fig. 3. Diagram showing exposure of the heart and great vessels after division of the sternum and caudal retraction of the costo-sternal complex.

The bilateral overgrown and deformed portions of the 4th through 7th costal cartilages were resected. The sternum was reapproximated to the manubrium with sternal wires and then elevated following anterior wedge osteotomy of the sternum at a level lower than the most deformed site. After the necessary shortening and tailoring, the sternal costal cartilages were sutured to the lateral costal cartilages with 2-0 polyester sutures to achieve chest wall stability without prosthetic materials. Drains were inserted into the thoracic cavity and retrosternal space. The pectoral muscles, subcutaneous tissues, and skin were approximated anterior to the sternum. He was extubated the



Fig. 4. Chest appearance one month after combined repair of pectus and tetralogy of Fallot.

following morning and discharged 14 days later in good condition with a stable anterior chest wall. Postoperative echocardiography showed a good result of the intracardiac repair without residual right ventricular outflow tract obstruction, residual ventricular septal defect or pulmonary insufficiency. The cosmetic appearance has been excellent at 1 month (Fig. 4) and 5 years postoperatively. A postoperative chest CT scan 3 years after operation demonstrated no recurrence of pectus excavatum (Fig. 5).

Discussion

The combination of complex congenital heart disease such as TOF requiring operative correction and pectus excavatum represents a major challenge to the surgeon. Pectus excavatum may complicate a planned cardiac procedure by making midline sternotomy technically more difficult, by limiting the operative exposure, and by the possibility of exacerbated postoperative cardiac and pulmonary dysfunction if the chest wall deformity remains uncorrected.^{5,9)} The traditional approach to patients with combined cardiac surgical disease and pectus deformity was to treat the conditions through separate surgical procedures.^{5,9)} Recently, there have been several English-language published reports documenting successful simultaneous repair of a pectus deformity and coexisting congenital heart defect.^{2,6,7)} However there is only a single

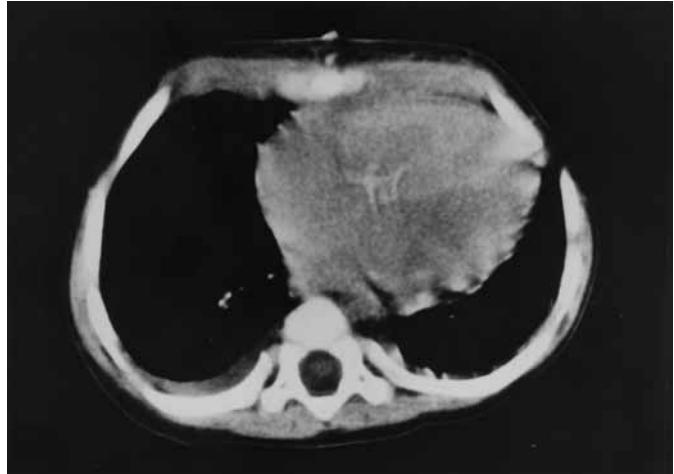


Fig. 5. Chest CT scan 3 years postoperatively. No recurrence of pectus excavatum was noted.

report describing simultaneous pectus repair and total correction of TOF.²⁾ In simultaneous pectus repair and total correction of TOF, possible life-threatening hypoxic spells during the time-consuming dissection of the deformed sternum and costochondral cartilages before institution of cardiopulmonary bypass are an important issue. Therefore, we performed a Blalock-Taussig shunt prior to the combined repair which enabled us to perform all procedures during the definitive combined repair without risk of hypoxic spells.

Our procedure involves three steps. First, the normal sternum is divided transversely and the sternal plate (costo-sternal complex) with rectus muscle pedicle is retracted caudally away from the operative field to gain maximum exposure of the heart for intracardiac repair; second, subperichondrial resection of the deformed costal cartilages; and third, elevation of the anterior chest wall with reapproximation of the sternum and sternocostal suture fixation without any prosthetic support. Our technique offers several advantages when employed in conjunction with open cardiac procedures. Except for the manubrium, the sternum is completely mobilized with its attached costal cartilages and retraction of the sternal plate obviates the need to divide the deformed sternum in the midline. It also offers excellent operative exposure with access to the heart and great vessels, even with severe cardiac displacement. Preservation of blood supply to the sternum via the rectus muscle pedicle should maintain viability of the sternum during operation and postoperatively. In spite of avoiding the use of prosthetic support, postoperative chest wall stability is sufficient. More-

over, our technique obviates the need for a subsequent operation to remove an internal prosthetic support.

Our patient underwent pectus repair in very early childhood at 18 months of age. Haller et al.⁸⁾ described that repair of pectus excavatum in very early childhood can interfere with chest wall growth and recommended that repair of pectus excavatum should be performed after 6 to 8 years of age. Because of the association with symptomatic TOF, our patient had to undergo pectus repair at a younger age. However, he is now 5 years postoperative and has shown excellent results both cosmetically and functionally.

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