



# Concomitant Tirone David and Ravitch Procedure in Marfan Syndrome: A Case Report

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## Abstract

Pectus excavatum is a known manifestation of Marfan disease and, when co-existing with aortic diseases imposing surgical intervention, its treatment is performed in simultaneous surgery. We describe a 29-year old man affected by Marfan syndrome, who underwent a Tirone David intervention for Valsalva sinuses dilatation and in concomitance a Ravitch procedure was needed. The patient had already undergone an esthetical intervention for the thoracic deformity, but the preoperative CT found a severe form of pectus, which imposed the aortic and thoracic correction in one single time. Surgery and postoperative outcome were satisfactory.

## Subject Areas

Surgery & Surgical Specialties

## Keywords

Ravitch, Pectusexcavatum

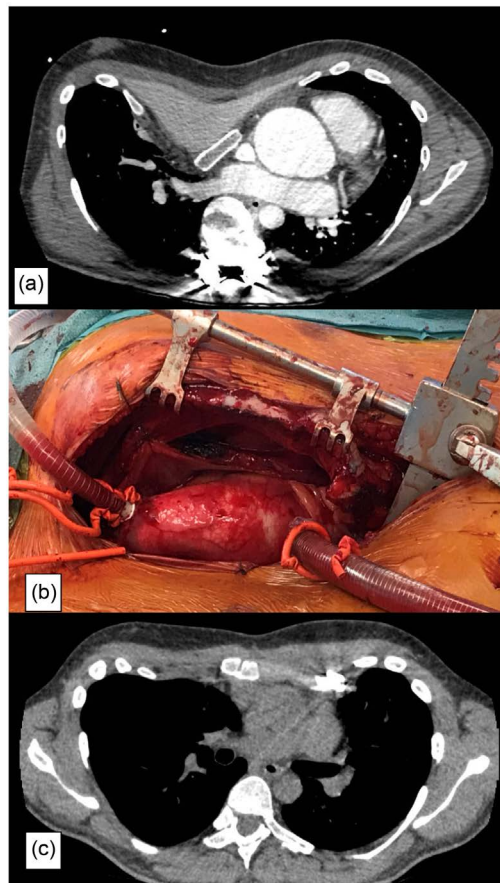
## 1. Introduction

The surgical treatment of pectus excavatum in concomitance with cardiac surgery is nowadays the preferred approach: it prolongs surgical time and potentially the bleeding risks as well, but it allows a correct resynthesis of the sternal edges, without compression of the right ventricle that is appealing for surgical and anesthesiological management.

## 2. Case Report

A 29-year old man with a dilatation of the Valsalva sinuses whose diameter was

measured 54 mm and a minimal insufficiency in a tricuspid aortic valve was addressed to our division for a valve sparing intervention. The patient was affected by Marfan syndrome with FBN 1 mutation, with bone, eyes and vascular involvement. In particular, the diagnosis was established after the examination of an important degree of scoliosis during childhood, which implied a spinal fusion and the assessment of a thoracic deformity as pectus excavatum. He was asymptomatic for pectus, but at the age of 22 the patient underwent the implantation of a silicon-prosthesis in sternal position, as esthetical correction. Preoperative CT-scan showed the proximity of the sternal bone to the dorsal vertebrae and a complete deviation of heart and mediastinal space towards the left thoracic side (**Figure 1(a)**). We hypothesized that the previous surgical correction for scoliosis may have decreased the distance between spine and sternum, so as to exacerbate mediastinal deviation. So after multidisciplinary discussion by cardiac and thoracic surgeons, the Tirone David procedure of valve sparing and the Ravitch intervention for pectus treatment were programmed as a single-time surgery.



**Figure 1.** (a) Preoperative CT: evident proximity of sternal bone to heart structures and dorsal vertebrae and complete mediastinal left-deviation. (b) Mammary retractor used for surgical exposition. (c) 5-month CT control, showing good result of the procedure.

After skin incision as usual, median sternotomy was performed with a sternotome, in order to lift up the sternal bone and spare the tissues behind. Because of the remarkable tilting and depth of the sternum towards the right of the patient, an asymmetric retractor (IMA Couëttil Retractor, Delacroix-Chevalier, Paris) was used to retract the sternal edges (**Figure 1(b)**). After obtaining an adequate exposure, cardiopulmonary bypass (CBP) was established through cannulation of the ascending aorta and right atrium. The aortic valve was reimplanted into a Valsalva graft (Gelweave Valsalva, Vascutek Terumo, Glasgow Ltd, UK), according to Tirone David technique: CBP time and cross clamping time were respectively 129 and 112 minutes. After CBP weaning and accurate haemostasis, the Ravitch procedure was initiated. The sternum and costal cartilages were exposed. After perichondral incision, the 5<sup>th</sup>, 6<sup>th</sup> and 7<sup>th</sup> costal cartilages on the right side were removed, followed by the 5<sup>th</sup> and 6<sup>th</sup> costal cartilages on the left side. As the right hemisternum was more challenging to mobilise, a partial transverse sternotomy of the inferior third was necessary to allow a complete mobilization of the sternal body and a complete correction of the sternal malformation. Result was secured with an osteosynthesis using a titanium bar (Thorib®, NeuroFrance, France) inserted under the sternum through the 5<sup>th</sup> intercostal space, and fixed with titanium staples on both sides. Osteosynthesis material is planned to be removed 6 months after surgery. Median sternotomy was then closed with usual metallic wires. Postoperative hemodynamic was supported by noradrenaline, gradually downshifted and the hospitalization was regular, without transfusions or biochemical alterations. We report a pericardial effusion drained 10 days after surgery, without complications. The patient was discharged on postoperative day 19, with a satisfactory functional and cosmetic result. A 5-month CT control showed a slight residual excavation in the lower part of the sternum, not clinically certifiable.

### 3. Discussion

Aortic and pectus excavatum surgery in Marfan patients have been carried out with many surgical techniques and approaches [1]. The Ravitch and the Nuss procedures are the most common techniques for the treatment of pectus excavatum, they are not simply esthetical and both have advantages and disadvantages: Ravitch technique implies cartilage excision with a greater risk of infection, but, in case of thoracic revision, the metallic bar excision is feasible; Nuss technique is performed usually with shorter times, but emergent resuscitation and bar excision may be more challenging. We chose the former one, as more manageable in case of urgent reaccess to the operative room. Our patient didn't need any emergent procedure, but a surgical drain had to be put in place, because of a pericardial effusion and it has been managed through a small incision beneath the xiphoid, without any complication. The concomitant treatment of aortic disease and pectus excavatum has become popular in literature, both in pediatric [2] and adult patients [3]. Most authors propose the single-time approach as the

best, as it allows an early cosmetic result and satisfaction. Besides, at time of sternal closure, a non-repaired pectus may compress the right ventricle with hemodynamic deterioration immediately after surgery [4]. The simultaneous correction allows a better cardiac and pulmonary function after surgery and allows a better surgical exposition, as well [5]. On the other hand it prolongs the surgical time and may increase the risk of bleeding and infection. In order to minimize surgical bleeding the thoracic procedure is started after protamin administration; besides, the perichondrium left in place avoids thoracic vessels to be damaged. In our case, mammary arteries were spared, so as to allow a better healing of bone and wound. In our experience Ravitch after aortic surgery extends the surgical time, but it presents low risks of acute bleeding or acute early morbidity. The fact of resynthesizing the sternal edges avoids an immediate heart compression and it is an appealing opportunity to the surgeon and to the operative team.

### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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