

SCIENTIFIC LETTER

AORTIC ROOT DILATION SURGERY AFTER CORRECTIVE REPAIR OF TETRALOGY OF FALLOT

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ABSTRACT

Two patients who underwent corrective repair of tetralogy of Fallot later developed aortic root aneurysms that required surgery 19 and 26 years after their index surgeries. Both patients were found to have the aneurysm wall intimately attached to the internal table of the sternum. Therefore, re sternotomy was performed using deep hypothermic circulatory arrest. This facilitated the use of a mediastinal approach without damage to the aneurysm. Surgery consisted of a classic Cabrol's procedure.

Keywords: *Tetralogy of Fallot; aneurysm; aorta; heart defects; congenital*

INTRODUCTION

Aortic root dilation in patients treated with corrective repair of tetralogy of Fallot (TOF) has been discarded for quite some time now after isolated cases of patients eventually requiring aortic valve replacement.

Several theories have tried to explain the origin of such dilation: aortic hyperflow due to congenital malformations has been proposed as one of the predisposing factors. Also, the coexistence of a bicuspid aortic valve or trauma sustained during corrective repair⁽¹⁾.

Similarly, the anatomical association between such dilation and the sternum is nothing but surprising, a situation probably triggered by the persistence of open parietal pericardium. This adds risk to the whole procedure due to the possibility of massive bleeding following sternal reopening for aortic surgery^(2,3).

We present the cases of 2 patients treated with corrective repair of TOF during their childhood who had to go under surgery again 19 and 27 years later

due to the presence of two 11-cm and 8-cm aortic root aneurysms, respectively.

CASE #1

We present the case of a 22-year-old male patient treated with TOF at 3 years old who underwent the closure of the interventricular communication (IVC), pulmonary valvuloplasty, and bicuspid aortic valve repair. The patient remained asymptomatic during his childhood and adolescence until, 3 months prior to this consultation, he started showing signs of dry cough, and bronchospasm, both refractory to treatment. After performing the corresponding imaging modalities, the coronary computed tomography angiography confirmed the presence of an 11-cm aneurysm in both the aortic root and the ascending aorta compressing both primary bronchi (*figure 1 and figure 2*). Due to the presence of laryngeal stridor the patient was referred for emergency treatment.



FIGURE 1. Coronary computed tomography angiography with 3D reconstruction showing a giant aneurysm of root and ascending aorta. The marks left by the metal stitches left by the closure of the sternum on the aneurysm wall become evident.

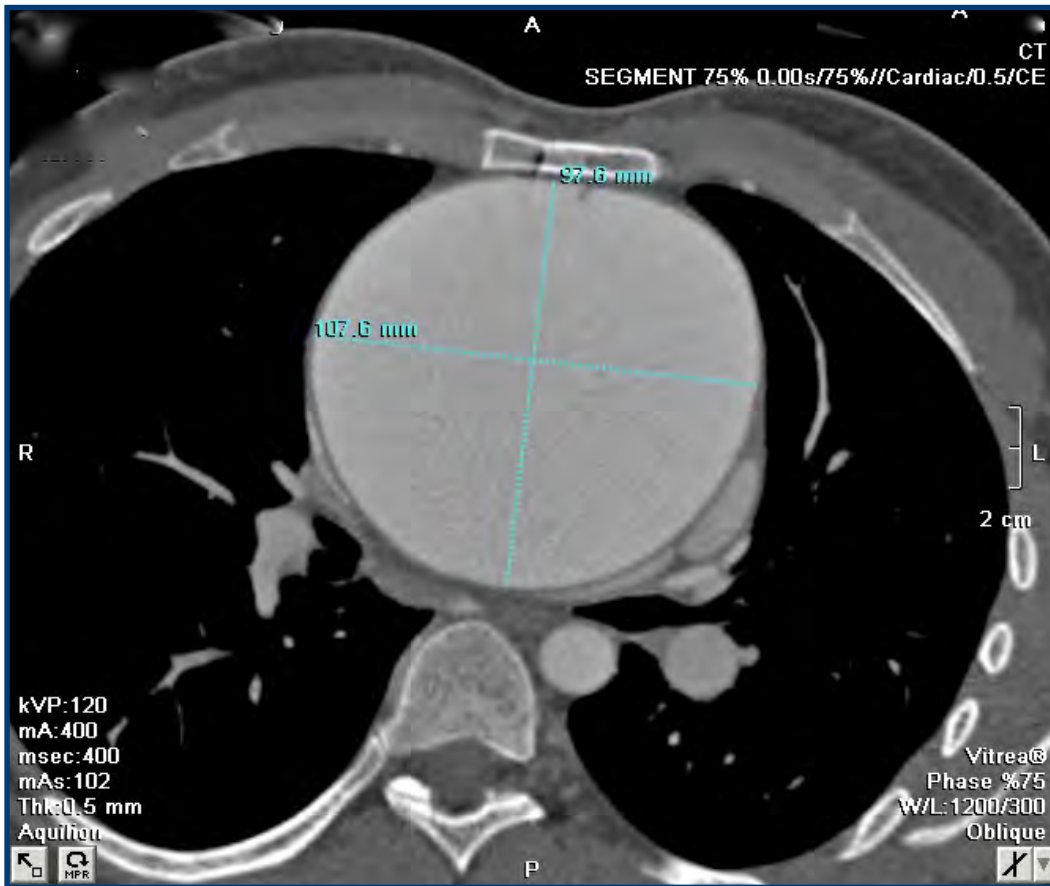


FIGURE 2. Axial projection showing the intimal correlation between the aneurysm wall and the internal table of the sternum.

Given the anatomical proximity of the aneurysm wall to the sternum, femoro-femoral cannulation, and extracorporeal circulation (EC) were performed followed by induced hypothermia down to 64.4°F. Hypothermic circulatory arrest, and median sternotomy followed with the depressurized aneurysm without any damage to its wall. The procedure was performed longitudinally up to 1 cm from the brachiocephalic trunk emergency where distal anastomosis was performed with a separate piece from a 28-mm Dacron graft. Eighteen minutes after circulatory arrest, EC was restarted with graft clamping and temperature was raised to 89.6°F. Myocardial protection was performed with antegrade cold blood cardioplegia through the coronary ostia, and retrograde cold blood cardioplegia through the coronary sinus intermittently. The next stage consisted of aortic valve resection, that was bicuspid, followed by Carbomedics 25/28 aortic valved conduit implantation. Given the significant distance between the coronary ostia and the conduit and the existing fibrosis due to previous surgery, a decision was made to perform bypass surgery between both ostia and the conduit using a 10 mm intervascular Dacron graft using the Cabrol technique to avoid excessive tension in both anastomoses. Finally, prostheto-prosthetic anastomosis was performed from the valved conduit

towards the anastomosed prosthesis and the distal ascending aorta.

The histopathological finding of the aortic wall was cystic necrosis of the tunica media.

The patient progressed without complications and his respiratory symptoms disappeared immediately. The patient was discharged from the hospital 8 days after surgery and remained asymptomatic at the 12-year follow-up. Figure 3 shows the coronary computed tomography angiography performed 1 year after surgery.

CASE #2

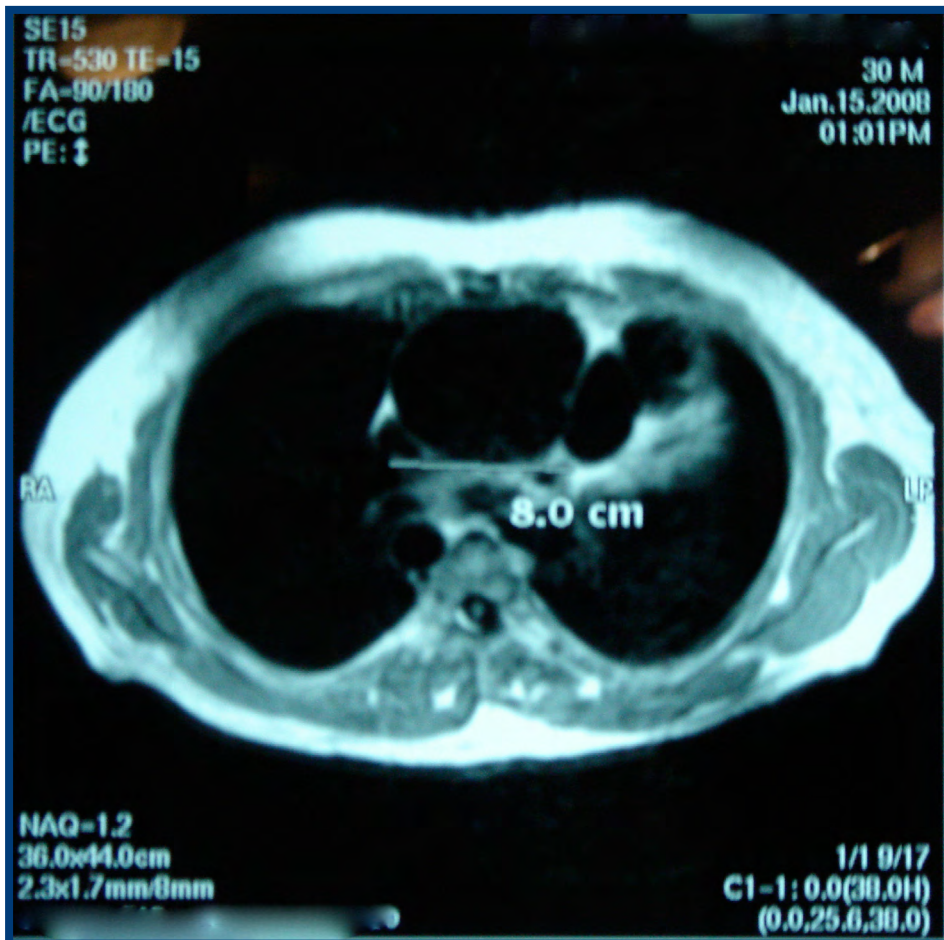
This is the case of a 30-year-old male patient with a past medical history of corrective repair of TOF at 4 years of age. It included the closure of the IVC followed by pulmonary valvuloplasty.

The patient remained asymptomatic during his growth and development, which were both normal. A routine x-ray check-up revealed the presence of mediastinal widening. A transthoracic echocardiogram was performed in the first place, and it revealed the presence of a 6-cm aortic root and an 8-cm ascending aorta with mild aortic regurgitation. These findings were confirmed on the thoracic coronary computed tomography angiography performed (figure 4).



FIGURE 3. Coronary computed tomography angiography with 3D reconstruction 1 year after surgery showing the aortic valved conduit with the prosthesis between both coronary ostia and the conduit.

FIGURE 4. Axial projection showing the intimal correlation between the aneurysm wall and the internal table of the sternum.



With this diagnosis a decision was made to remove the aneurysm that, as in the former case, was also attached to the internal table of the sternum.

Femoro-femoral cannulation and EC were performed followed by induced hypothermia down to 64.4°F. Hypothermic circulatory arrest was followed by median sternotomy with the depressurized aneurysm without damage to its wall. EC was restarted after a 3-minute pause. Afterwards, the distal ascending aorta was clamped immediately proximal to the brachiocephalic trunk where there was a 30 mm neck. The aortic valve, root, and ascending aorta were replaced by a Carbomedics 27/30 aortic valved conduit with interposition of a 10-mm intervascular Dacron graft between both coronary ostia with latero-lateral anastomosis to the conduit using the Cabrol technique.

The immediate postoperative was satisfactory and the patient was discharged on day 9.

The histopathological finding was focal hyalinosis with discrete calcification of the valve and the aortic wall.

The patient remained asymptomatic at the 11-year follow-up. The coronary computed tomography angiography performed is shown on figure 5.

DISCUSSION

Very few studies have been published on the association between the appearance of aortic dilation and corrective repair of TOF.

Back in 2001, Niwa found that 32 patients out of a series of 218 patients treated during their childhood developed this condition later in life. This happened more frequently in male patients with pulmonary atresia and presence of a right-sided aortic arch⁽¹⁾.

Also, a high incidence rate of extracellular matrix disorders like cystic medial necrosis associated with Marfan syndrome and/or bicuspid aortic valve has been reported.

Other authors have published isolated cases with the same condition^(4,5).

Tan y col. conducted a bibliographic search to try to understand the mechanisms involved in this phenomenon. They also studied their own series of 17

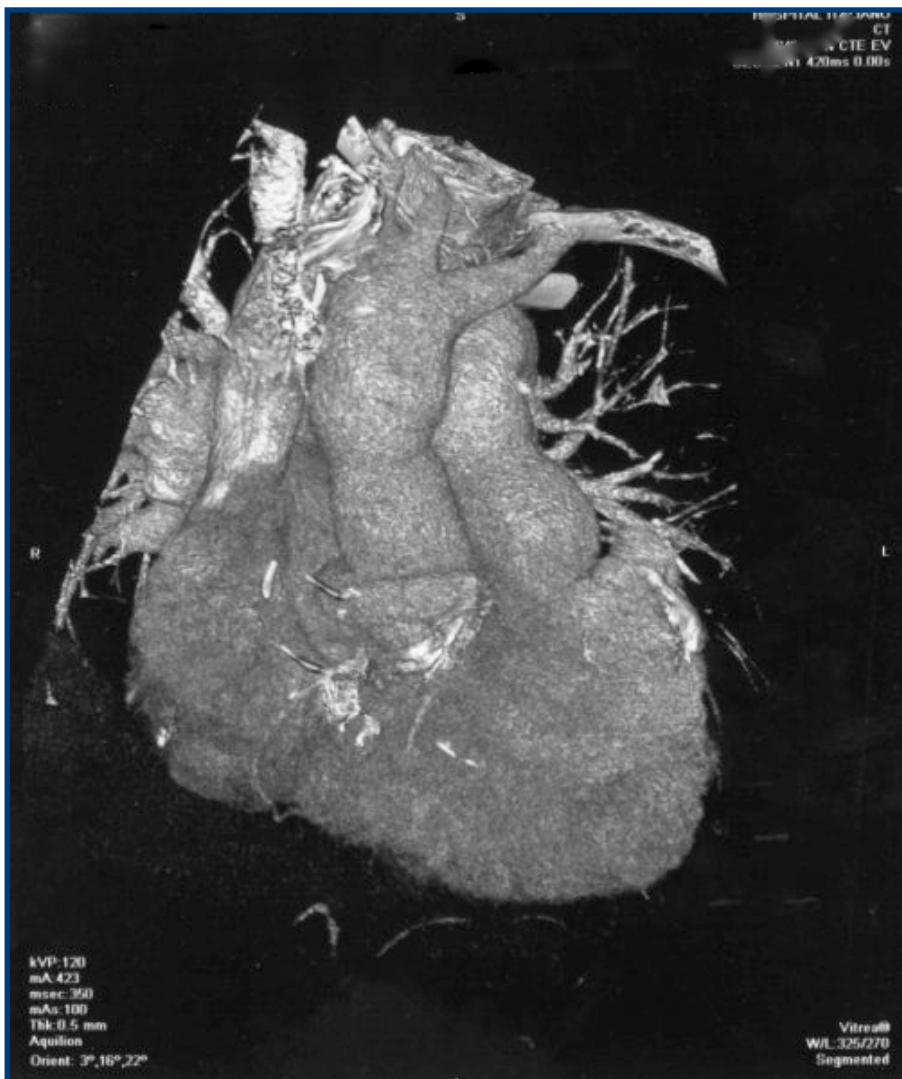


FIGURE 5. Coronary computed tomography angiography with 3D reconstruction 1 year after surgery showing the aortic valved conduit with the prosthesis between both coronary ostia and the conduit.

postmortem patients to eventually find a correlation between this complication and the index between the measurement of the left ventricular outflow tract and the sinotubular junction of the aorta⁽⁶⁾.

On the other hand, the significant diameter reached by the aorta in its growth creates an anatomical correlation with the chest wall that turns open surgery into a true surgical challenge. Therefore, to this date, it is essential to use a postoperative coronary computed tomography angiography with preoperative contrast to plan the surgical strategy in general and proceed to open up the sternum in particular⁽⁷⁾.

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