# A Third-time Emergent Operation For Aortic Prosthesis Subtotal Dehiscence: Diagnostic Iconography

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### Abstract

A 31 years old female was operated on in Our Institution for an emergent subtotal dehiscence of aortic mechanical valve. She had a diagnosis of Marfan syndrome and underwent two previous cardiac operations in another Institution, both for infective endocarditis. The aortic prosthesis was kept in its position only by 2-3 surgical stitches and its impressive systo-diastolic movement is presented in the diagnostic iconography (TE echo and angiography). A 23 Freestyle root bioprosthesis was used to replace the previous valve. A complete AV block required a definitive pacemaker. The patient was discharged after 16 days and she is in healthy condition at a six month follow-up.

# CASE REPORT

A 31 years old female was admitted in our Institution in October 2000 after an acute onset episode of aphasia and syncope.

## HISTORY

The patient had a previously made diagnosis of Marfan syndrome and underwent two previous cardiac operations, both in another Institution:

In January 2000 she underwent a therapeutic abortion and in August of the same year she had some episodes of atrial flutter and fever with negative blood cultures.

# **DIAGNOSTIC STEPS**

At the admission, she was conscious, anxious and restless, with no evident neurological deficits and no chest pain.

ECG showed a tachycardic sinus rhythm with complete left bundle branch block and ST elevation in the first two precordial leads. Total CPK was 588 UI/l, with an MB value of 153 UI/l and troponin I value of 5.43 (g/dl.

CT scan showed no aortic dissection and no cerebral haemorrhage. During this exam the patient had to be intubated for acute dyspnea.

TEE showed an extensive dehiscence of the aortic prosthesis, with a moderate-severe aortic insufficiency. Note the diastolic limitation to the opening of the anterior leaflet of the mitral valve caused by the prosthesis falling down in the outflow tract.

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Angiography was performed to study the patency of the venous graft, the coronary arteries and the whole aorta. It showed a very impressive displacement of the aortic prosthesis which was pulled aside by the antegrade flow and then fell down during diastole in its nearly physiologic position, obtaining in some way a diastolic occlusion of the LV outflow tract. The prosthesis behaved like a mono-leaflet valve hinged on the few surgical points still attached.

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## SURGICAL PROCEDURE

Operation started five hours after the onset of symptoms. CPB was instituted through femoral artery and bicaval cannulation after reopening of median sternotomy, lysis of the adhesions and positioning of LV vent. Only the proximal portion of venous graft was isolated. A complete transverse aortotomy was used to access the aortic valve. The prosthesis was still attached only in 2-3 stitches in the noncoronary annulus and the aortic annulus was completely frayed. The prosthesis was replaced with a Freestyle size 23 root bioprosthesis. A large amount of desaturated blood was noted during the procedure flowing from the left side of the heart and an atrial septal defect (fossa ovalis) was identified in caval occlusion and closed with a running suture.

### **POSTOPERATIVE COURSE**

The postoperative course was complicated by:

A complete AV block which required a permanent bipolar endocavitary pacemaker, implanted in POD 15; Some neurological deficits (right facial paresis, right arm hypostenia, dysarthria) fully recovered in 1 week.

A cerebral CT scan showed an old thalamic ischemic lesion and luxury perfusion on the distribution of mean cerebral artery due to an hypoxic state. No cultures were positive. Histology of the aortic wall confirmed a cystic medionecrosis. The patient was discharged in POD 16 and she is in healthy condition at a follow-up check she had in March 2001.

## DISCUSSION

Aortic valve endocarditis is not the most common presentation of children with Marfan syndrome [ $_1$ ]. We have no data about the cultural exams performed in the previous operations, but the anatomic description at the time of surgery showed some characteristics of infective disease. Some Authors [ $_2$ ] described a very acceptable mortality rate in reoperation on the aortic root. The case described was particular because of the extension of the dehiscence, which made highly probable the complete embolization of the prosthesis, and because of the occluding function that the prosthesis still had, which kept the haemodynamic status in a life-compatible range.

According to the variability of presentation and the speed of recovery, it can be assumed that neurological deficits were due to arrhythmic complications and to the low output syndrome and not to tissue mobilization from the aortic annulus. On the other hand, old thalamic lesion could be due to the misdiagnosed AS defect. The suspicion of endocarditis as pathological cause was based on the patients history, on the acute onset, and on the macroscopic aspect of the annulus tissue.

The use of Freestyle bioprosthesis as a rtic root  $[_{3,4}]$  in the reported case has various positive aspects:

A strict follow-up is required for these patients, in order to prevent complications, to support them during life and to avoid, whenever possible, performing of reoperation in emergency condition.

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