Aorto-Right Ventricular Tunnel in Transposition of the Great Arteries

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Aorto-ventricular tunnel is an extremely rare congenital heart defect, consisting of failure of attachment of an aortic leaflet along the semilunar hinge. In all published reports the leaflet involved was either the right coronary leaflet, most frequently, or the left coronary leaflet, in most of the cases opening toward the left ventricle, with only one-eighth of the reported cases communicating with the right ventricle. Treatment of the aorto-ventricular tunnel has been anecdotally reported by interventional closure with a device and more frequently with surgical approach, either as an isolated malformation or as associated lesions. To the best of our knowledge, the presence of an aorto-ventricular tunnel of the non-adjacent aortic leaflet in transposition of the great arteries has never been reported.

We have observed an aorto-ventricular tunnel involving the non-adjacent leaflet of the aortic root, which after arterial switch became the pulmonary root. The patient presented 18 years after the arterial switch with progressive dilatation of the right ventricle due to severe degree of pulmonary valve regurgitation, confirmed by echocardiography and cardiac MRI. Indication for surgery was given with the plan for a pulmonary valve implantation. Because of the intra-operative finding of disconnection of the anterior leaflet of the pulmonary valve (former aortic valve) along the semilunar hinge, the surgical plan was modified and the anterior leaflet was attached to the valve annulus, with subsequent plasty in correspondence with the right and left commissurae to reduce the size of the dilated annulus to normal diameter. The post-operative course was uneventful, with extubation after few hours and discharge 4 days after surgery, with echocardiography showing trivial degree of pulmonary valve regurgitation. The patient remains in good conditions 6 months after surgery.

Keywords: annular hinge, aorto-ventricular tunnel, arterial switch, new pulmonary valve regurgitation, semilunar valves

INTRODUCTION

Aorto-ventricular tunnel is an extremely rare congenital heart defect (1–25), consisting of failure of attachment of an aortic leaflet along the semilunar hinge (24, 25). In all published reports, the leaflet involved was either the right coronary leaflet, most frequently, or the left coronary leaflet (1–25), in most of the cases opening toward the left ventricle, with only one-eighth of the reported cases communicating with the right ventricle (24, 25). Treatment of the aorto-ventricular tunnel has been anecdotally reported by interventional closure with a device (26) and more frequently with surgical approach, either as an isolated malformation (27–62) or with associated lesions (63–70).
To the best of our knowledge, the presence of an aorto-ventricular tunnel of the non-adjacent aortic leaflet in transposition of the great arteries has never been reported. We have observed an aorto-ventricular tunnel involving the non-adjacent leaflet of the aortic root, which after arterial switch became the pulmonary root.

**BACKGROUND**

Because of severe cyanosis in the presence of transposition of the great arteries and ventricular septal defect, a neonate underwent emergency balloon atrio-septostomy procedure at the age of 2 weeks. After few days, arterial switch and closure of ventricular septal defect were performed with uneventful post-operative course. The child was then followed with regular follow-up visits; at the age of 3 years, a follow-up cardiac catheterization showed normal reimplanted coronary arteries. From the age of 1 year, the echocardiography reports noted the presence of moderate degree of pulmonary valve regurgitation, increasing to severe degree around the age of 9 years, with the patient remaining asymptomatic. Since after, the echocardiographic investigations showed a progressive dilatation of the right ventricle, confirmed by MRI at the age of 17 years, showing an indexed end-diastolic right ventricular volume = 211 ml/m².

At the 18 years of age, 64 kg of body weight, he presented with mild right ventricular impulse on the chest on examination, and systo-diastolic murmur compatible with free pulmonary valve regurgitation. Electrocardiography showed sinus rhythm with 53’ heart rate, partial right bundle branch block with QRS duration = 110 ms. Echocardiography showed severe pulmonary valve regurgitation, with Doppler velocity = 2.7 m/s through the pulmonary valve.

Cardiac MRI confirmed the presence of severe pulmonary valve regurgitation with fraction of regurgitation = 57%, dilated pulmonary valve annulus (23 mm, with 18.5 mm normal for the body surface area) (**Figures 1A,B**), severe dilatation of the right ventricle (**Figure 1C**), with ejection fraction = 50%, indexed end-diastolic right ventricular volume = 226 ml/m², indexed end-systolic right ventricular volume = 113 ml/m², normal size and function of the left ventricle (**Figure 1C**). CT scan confirmed the findings, showing mildly dilated ascending neo-aorta and normal coronary arteries.

Cardiopulmonary stress test showed peak VO2 = 39 ml/kg/min (91% of normal), and peak heart rate 157’ (78% of normal). The test was terminated before reaching the maximum heart rate because of ST depression in V3 and V4, without chest pain.

At the multi-disciplinary meeting, the decision was agreed to proceed with pulmonary valve implantation, because of the severe degree of pulmonary valve regurgitation and right ventricular dilatation. The patient was informed and consent was given for the planned surgical procedure.

On general anesthesia, transesophageal echocardiography confirmed the presence of severe regurgitation of the pulmonary valve. The cardiopulmonary bypass, established between the left femoral artery and the right atrium, was maintained through the entire procedure on normothermia with beating heart. Through a longitudinal incision of the main pulmonary artery, in anterior position after the previous Lecompte maneuver, the presence of three leaflets pulmonary valve (native aortic valve) was confirmed, but while the two posterior leaflets appeared absolutely normal, the anterior non-adjacent leaflet was completely disconnected from the annulus and floating inside the pulmonary artery (**Figure 2A**). The disconnected anterior non-adjacent leaflet was reattached to the annulus with double running suture of 5/0 monofilament (**Figure 2B**). At this point, the pulmonary valve annulus, dilated to 24 mm diameter, was reduced to 19 mm diameter (normal for the age = 18.5 mm) with plasty in correspondence of the right and left commissurae with 5/0 monofilament suture pledged with a small patch of autologous pericardium. After closure of the incision of the main pulmonary artery with double running suture of 5/0 monofilament, the patient was easily weaned from cardiopulmonary bypass after 35 min, without inotropic support.

**Figure 1** | **(A)** Cardiac MRI with sagittal view of the right ventricular outflow tract showing the anterior leaflet of the pulmonary valve without attachment to the annular hinge. **(B)** Cardiac MRI with axial view of the pulmonary valve showing morphologic abnormality of the anterior leaflet. **(C)** Cardiac MRI with four-chamber views showing the dilatation of the right ventricle. Abbreviation: LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.
Intra-operative transesophageal echocardiography confirmed the good result with trivial regurgitation of the pulmonary valve (Figure 3A). The rest of the hospitalization was uneventful, with extubation after few hours, discharge from ICU next morning and discharge home on the fourth post-operative day. Echocardiography before discharge confirmed the presence of well functioning pulmonary valve and reduced the size of the right ventricle (Figures 3B, C). The patient remains asymptomatic 6 months after surgery.

DISCUSSION

In all published reports of aorto-ventricular tunnel, extremely rare congenital heart defect, the leaflet involved was either the right coronary leaflet, most frequently, or the left coronary leaflet, but never the anterior non-adjacent leaflet (1–25).

We are not aware of any report, an aorto-ventricular tunnel of the non-adjacent aortic leaflet in transposition of the great arteries, with, therefore, aorto-right ventricular tunnel.

The patient we report underwent arterial switch in the neonatal period, but at that time no one observed the presence of native aortic valve regurgitation in the right ventricle, while regular
CONCLUDING REMARKS

A very unusual combination of aorto-ventricular tunnel with transposition of the great arteries came to observation 18 years after the arterial switch performed in the neonatal period, due to progressive regurgitation of the native aortic valve in pulmonary position. Surgical repair of the lesion with attachment of the non-adjacent leaflet to the annulus avoided the planned valve implantation.

ETHICS STATEMENT

Written and informed consent was obtained from the patient for publication of this case report.

AUTHOR CONTRIBUTIONS

AFC proposed the report of this malformation never observed before, prepared the entire manuscript, and the revised edition. SD contributed to the preparation of the manuscript and the choice of the illustrations. RA revised and edited the manuscript after contributing to the literature review.

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