IMAGES **in PAEDIATRIC CARDIOLOGY**

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Abstract

Aorto-left ventricular tunnel is a rare congenital cardiac anomaly, consisting of a short abnormal pathway, usually from a sinus of Valsalva into the left ventricular cavity. It is usually diagnosed with echocardiography. We report a case of a newborn presenting with heart murmur and rapid progression to heart failure and left ventricular enlargement due to an aorto-left ventricular tunnel. Despite successful closure of the tunnel, the patient required a Ross procedure due to progressive aortic disease.

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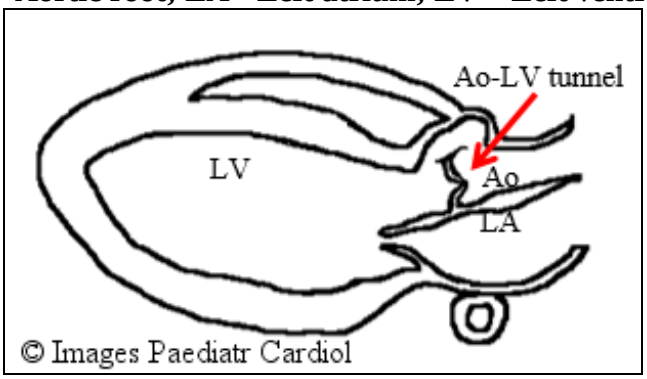
Aorto-left ventricular tunnel is a rare congenital cardiac anomaly, consisting of a short abnormal pathway, usually from a sinus of Valsalva into the left ventricular cavity. It is usually diagnosed with echocardiography. We report a case of a newborn presenting with heart murmur and rapid progression to heart failure and left ventricular enlargement due to an aorto-left ventricular tunnel. Despite successful closure of the tunnel, the patient required a Ross procedure due to progressive aortic disease.

Introduction

Aorto-left ventricular tunnel is a rare congenital cardiac anomaly.[1] first described in 1963.[2] It consists of a short abnormal pathway, usually from the right sinus of Valsalva, through the upper end of the ventricular septum, into the LV cavity[3] (Figure 1).

Figure 1. Representation of aorto-left ventricular tunnel, as seen on transthoracic echocardiogram, parasternal long-axis (PLAX) view.
Ao – Aortic root; LA - Left atrium; LV – Left ventricle.



Its embryological basis remains unknown[4] and the presentation ranges from in-utero fetal death[5] to asymptomatic adulthood.[6] Some patients present as neonates with cardiomegaly and severe heart failure, thrill, and harsh to and fro murmur.[3,4,7] When symptoms are present in infancy and surgical repair is not accomplished, death usually occurs within a few months.[3] Diagnosis can usually be made with two-dimensional echocardiography and Doppler color flow imaging[3] and many patients also have aortic valve abnormalities, as in this case presented hereunder.[4]

Case Report

A newborn with a heart murmur was referred for cardiac evaluation on the first day of life. He was born after a twin pregnancy, at 36 weeks. He had a birth weight of 2100g and an Apgar score of 3, 9, and 10 at 1, 5, and 10 minutes, respectively. At his first observation, there was a systolic ejection click and a harsh to-and-fro heart murmur was heard along the left sternal border; there were no signs of heart failure and no other remarkable findings. The chest x-ray demonstrated enlarged cardiac silhouette (cardiothoracic ratio: 65%); the electrocardiogram revealed left ventricular hypertrophy and the echocardiography showed moderate-to-severe aortic valve stenosis (peak and mean gradient 70 and 40 mmHg, respectively), a patent ductus arteriosus (PDA) and a severe aortic diastolic flow not related with aortic regurgitation, due to a tunnel between the left ventricle and the aortic root, above the right coronary ostium (figures 2 and 3).

Figure 2 (a and b). Transthoracic echocardiogram, parasternal long-axis (PLAX) view, showing non valvular aortic regurgitation with color Döppler (B), due to a tunnel* between the aortic root (Ao) and the left ventricle (LV), above the right coronary ostium. Ao – Aortic root; LA - Left atrium; LV – Left ventricle; * - tunnel

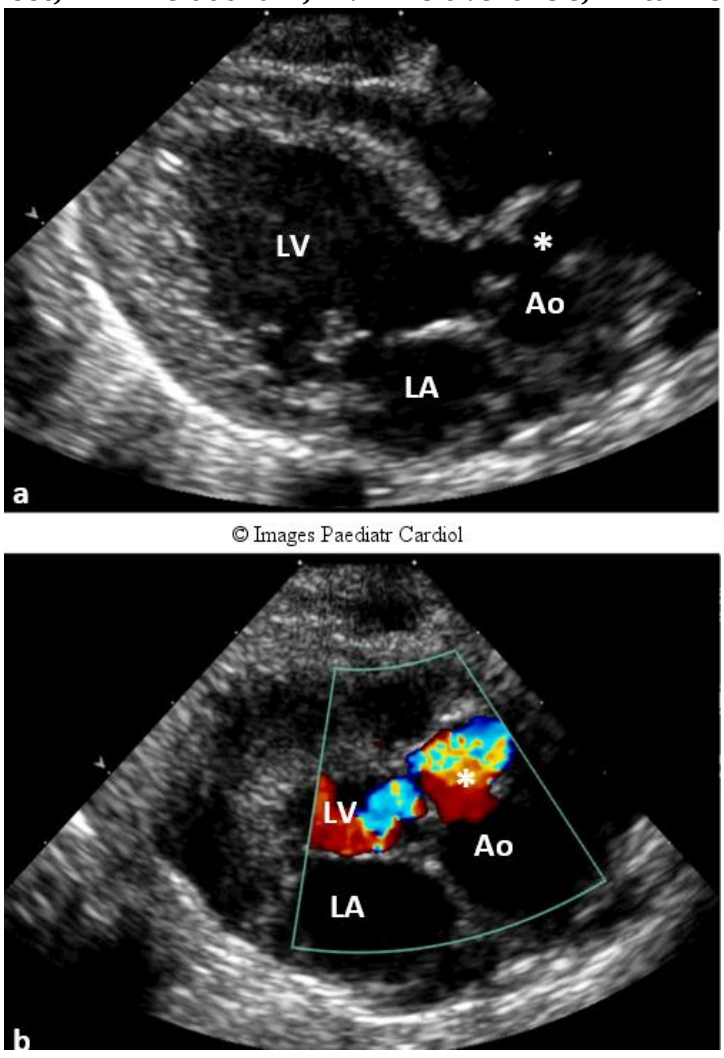
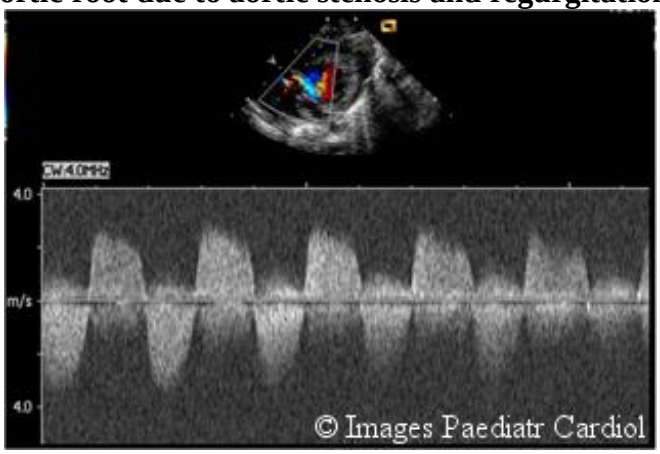
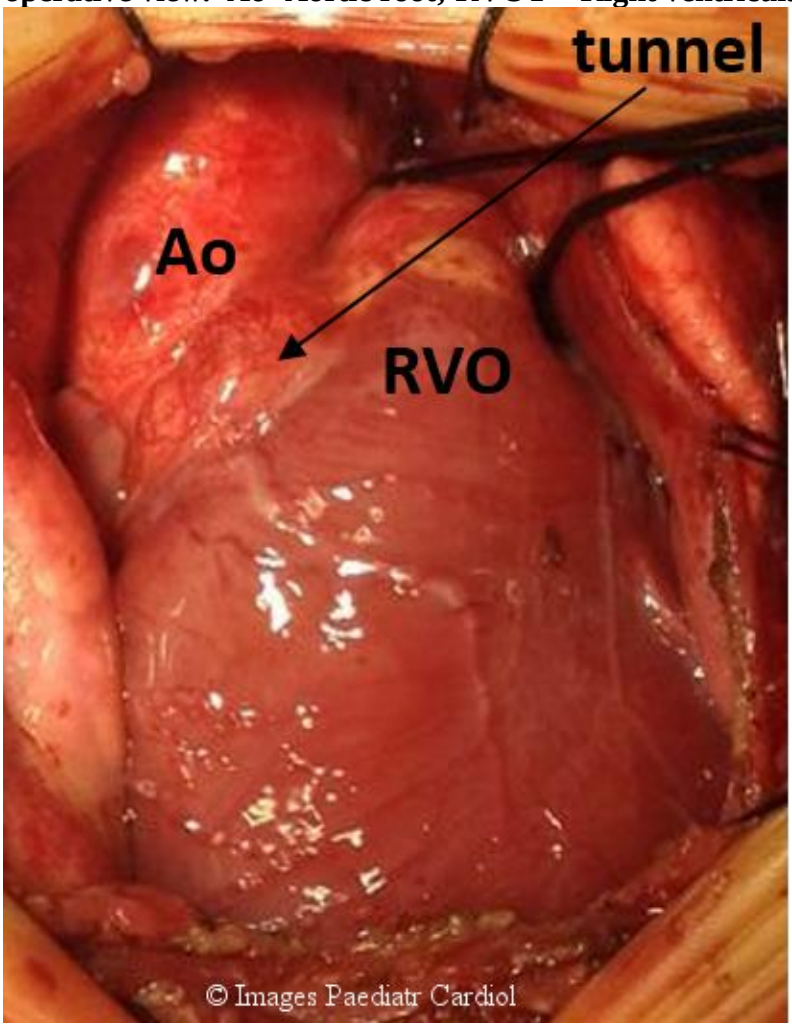


Figure 3. Transthoracic echocardiogram, subcostal view, systo-diastolic wave Döppler of the aortic root due to aortic stenosis and regurgitation.



He gradually developed signs of heart failure and progressive enlargement of the left ventricle. The patient was submitted to surgical closure of the ventricular opening using a Gore-Tex® patch, aortic valvuloplasty and PDA closure on the 13th day of life (Figure 4). The post-operative period was uneventful and the patient was discharged home on the 22th day of life on furosemide and captopril, for residual aortic regurgitation.

Figure 4. Intra-operative view. Ao- Aortic root; RVOT – Right ventricular outflow tract



Even though there was no residual tunnel, the continuous progression of the aortic valve disease resulted in severe aortic stenosis (peak and mean gradient 90 and 50 mmHg, respectively) and regurgitation, left ventricular hypertrophy and dilation of the ascending aorta. Therefore, in the 6th month of life, the patient was submitted to Ross surgery, but unfortunately did not survive as an untreatable ventricular fibrillation occurred 14 hours after surgery, despite an otherwise favorable surgical result.

Discussion

This case is an example of this extremely rare cause of heart failure in the newborn and its progression despite surgical correction, due to an associated defect. Despite presenting as an isolated heart murmur, the size of the tunnel resulted in rapid progression to left ventricular dilation and heart failure within the neonatal period, as described in the literature.[3,6,7] Even though the tunnel could be successfully closed, there was rapid progression of the associated aortic valve disease, resulting in the need of a new intervention in the 6th month of life. In patients with significant aortic valve disease, ventricular arrhythmias are common both before and after intervention.[8] In the case reported, in spite of an otherwise favorable surgical result, the patient di not survive a postoperative arrhythmia which is a recognized and possible cause of the death after the Ross procedure.[8,9]

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