Critical aortic stenosis in combination with an aorto-left ventricular tunnel: a rare congenital malformation

Christian Schreiber¹*, Jürgen Hörrer¹, Andreas Kühn², and Manfred Vogt²

¹Clinic of Cardiovascular Surgery, German Heart Center Munich at the Technical University, Lazarettstrasse 36, 80636 Munich, Germany and ²Clinic of Pediatric Cardiology and Congenital Heart Disease, German Heart Center Munich at the Technical University, Lazarettstrasse 36, 80636 Munich, Germany
* Corresponding author. Tel: +49 89 12184111, Fax: +49 89 12184113, Email: schreiber@dhm.mhn.de

On the first day of life, a female newborn with a three out of six systolic murmur on the right sternal border showed signs of heart failure: dyspnoea, tachypnoe, low blood pressure. On echocardiography, the diagnosis of a critical aortic stenosis with a pin whole ostium was established and a prostaglandin infusion was started.

The aortic valve appeared tricommissural with an opening from the right sinus of valsalva to a lateral and anterior extra-aortic tunnel (Panel A), the right coronary artery only few millimetre separate. In subcostal echo views, the tunnel could be demonstrated in a more longitudinal axis from its beginning lateral of the aorta (Ao) to its end in the left ventricular outflow tract (Panel B). The flow through this tunnel was antegrade from the left ventricle (LV) to the aorta in systole with a backflow Ao→LV in diastole. The maximum systolic instantaneous gradient on CW Doppler as measured through the stenotic aortic valve was 95 mmHg, mean Doppler gradient 42 mmHg.

On the fourth day of live, a balloon valvuloplasty was performed and the pressure gradient dropped to maximum 48 and mean 28 mmHg on Doppler with a mild aortic regurgitation through the valve. Seven months later aortic stenosis became again clinically relevant (101/46 mmHg maximum and mean gradients).

At operation, the enlarged aortic root bulging towards the pulmonary trunk was found (Panel C). The dysplastic aortic valve was excised (see Supplementary material online, Movie S1). The aortic annulus was incised towards the mouth of the tunnel (Panel D). Thus, the aortic annulus was enlarged to accommodate the smallest available mechanical heart valve prosthesis. The aortic root was augmented with a patch. The post-operative course was uneventful.

An aorto-left ventricular tunnel is a rare congenital malformation where a channel connects the ascending Ao above the sinotubular junction to the cavity of the LV. Associated lesions of the aortic valve occur in ~20%, ranging from bicuspid valves without obstruction to dysplasia and even atresia. Usually, after adequate tunnel closure, the aortic valvar mechanism is not a clinical problem during follow-up. However, in some patients, aortic insufficiency may increase. In our described case, a severe dysplasia required early valve replacement.

Supplementary material is available at European Heart Journal online.

Panel A Transthoracic echocardiography, subcostal view: demonstrating the connection of the LV with the Ao via the stenotic aortic valve and the left ventricular aortic tunnel (asterisk). Colour Doppler shows the regurgitation from the Ao to the LV through the left ventricular aortic tunnel (arrow).

Panel B Transthoracic echocardiography, parasternal short axis: shows the connection of the left ventricular aortic tunnel (asterisk) with the Ao in the region of the right aortic coronary cusp. Colour Doppler proves blood flow across this connection (arrow). RCA indicates the right coronary artery.

Panel C At operation, a prominent area of the ascending aorta just above the tunnel was found.

Panel D Surgeons’ view from the aorta. Depiction of the tunnel (probe through the tunnel).