Case report: A 3 day old neonate was transferred to our unit because of a heart murmur detected on routine examination before discharge from nursery. Apart from a systolic-diastolic murmur grade 4/6 the child was completely asymptomatic with normal vital signs, blood gases and without elevated lactate.

Echocardiography revealed a thickened, dysplastic and severely stenotic aortic valve with a Doppler derived peak gradient of 70 mmHg. The Ao-LV tunnel consisted of an aneurysmatic extracardiac component measuring 1,3 x 1,8 cm. The aortic orifice of the tunnel was 3 mm in diameter entering the Aorta just above the right coronary sinus. The ascending aorta was significantly dilated. The left ventricle was mildly dilated with increased apical trabeculations but without endocardial fibroelastosis.

Based on the echocardiographic findings the child underwent successful Ross operation on the 9th day of life and was discharged 14 days thereafter. At that time echocardiographic findings showed normal LV function, no aortic stenosis with trace regurgitation.

Discussion: Ao-LV tunnel is a rare anomaly with an unknown etiology. In a substantial number it is associated with aortic valve anomalies including severe aortic stenosis. The few caess described in the literature presented with critical hemodynamic status calling for immediate action. Our case however was hemodynamically stable with preserved LV function due to the Ao-LV tunnel that served as a natural bypass. The aortic orifice of the tunnel was big enough to maintain cardiac output, but restrictive enough to limit regurgitation. Thus we could postpone surgery to the ninth day. To the best of our knowledge this is the first case of Ao-LV tunnel and critical aortic stenosis that did undergo neonatal Ross procedure.