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A rare pathology in a newborn with cardiomegaly: Aortic left ventricular tunnel

Short title: Aortic Left Ventricular Tunnel

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A female infant born at 38 + 5 weeks *via* C/S, weighed 3300 g with an APGAR score of 1/5. The patient, who was evaluated as sinus valsalva aneurysm in the echocardiographic evaluation performed due to respiratory distress, was referred to us for surgery on the 2nd postnatal day. Physical examination revealed a 3/6 systolodiastolic murmur at the right sternal border and a hyperdynamic precordium. TELE showed a cardiothoracic index increase to 0.7 and an electrocardiography indicated findings consistent with left ventricular hypertrophy. Echocardiography showed a large aneurysm connecting to the left ventricle and aorta and causing right ventricle outflow obstruction in parasternal long axis (Figure 1A) and 5-chamber view (Figure 1B) (Supplementary material, *Videos S1–2*). Computed tomography (CT) angiography showed a tunnel extending from the aorta to the left ventricle, which was tortuous and associated with an aneurysm (Figure 1C).

Surgery was performed on the 5th postnatal day, and the diagnosis of aortic-left ventricular tunnel (ALVT) was confirmed during surgery. The ventricular and aortic entrances of the tunnel were closed with an autologous pericardial patch. Postoperative echocardiography showed no residual defect and mild aortic valve regurgitation (Figure 1D). After surgery, the patient monitored in the pediatric cardiac intensive care unit (ICU). The patient was discharged on postnatal day 28 after uncomplicated ICU follow-up.

Aortic-left ventricular tunnel is a rare congenital heart defect with an incidence of 0.001%, characterised an extracardiac connection between the left ventricle and aorta [1]. Although the etiology remains unclear, it's thought to be related to underdeveloped extracardiac tissues separating the aortic and pulmonary sinuses and their leaflets [2].

Typically, patients may present signs of rapidly developing heart failure in the neonatal period. Untreated cases usually result in death in the first year of life. A very small proportion of patients may reach adulthood as asymptomatic [3].

Although transthoracic echocardiography is sufficient for diagnosis, additional imaging modalities such as CT/magnetic resonance (MR) angiography may be required to support the diagnosis [3]. Differential diagnoses involve sinus of valsalva aneurysm rupture, corono-cameral fistulas, and aortico-ventricular fistulas. Sinus of valsalva aneurysms are differentiated from ALVT because they originate below the sinotubular junction and usually open into the right ventricle, whereas corono-cameral fistulas are distinguished by their intramural course [3].

There are different anatomical variants of ALVTs. According to the most commonly used classification by Hovaguimian et al. [4], ALVTs are divided into 4 types [4]. We classified our patient as type 4 because the aneurysm was both extracardiac and intracardiac and caused RVOT obstruction.

Treatment of ALVT is surgical. Surgical closure may be a combination of suture and patch techniques depending on the location and size of the defect. Although it is difficult to perform surgery especially in type 4 cases, two-patch technique is preferred in infants [5]. Operative mortality is between 3% and 8.3%. The causes of death after surgery were congenital anomalies in the coronary arteries, severe aortic stenosis despite simultaneous valvuloplasty, poor left ventricular function, and rupture of the infected suture line. Aortic valve regurgitation is one of the most significant short and long-term complications after surgery and may require valve replacement. Residual aorto-ventricular tunnel is another concern during long-term follow-up and can be successfully closed *via* transcatheter using an Amplatzer duct occluder [5].

ALVT should be considered in early-onset and rapidly progressing neonatal heart failure. The presence of an extracardiac tunnel connecting the aorta and left ventricle on echocardiography is diagnostic for ALVT, but supplementary imaging like CT/MR angiography may be necessary in uncertain cases. Treatment for ALVT is surgical and the results of early surgery are successful.

Article information

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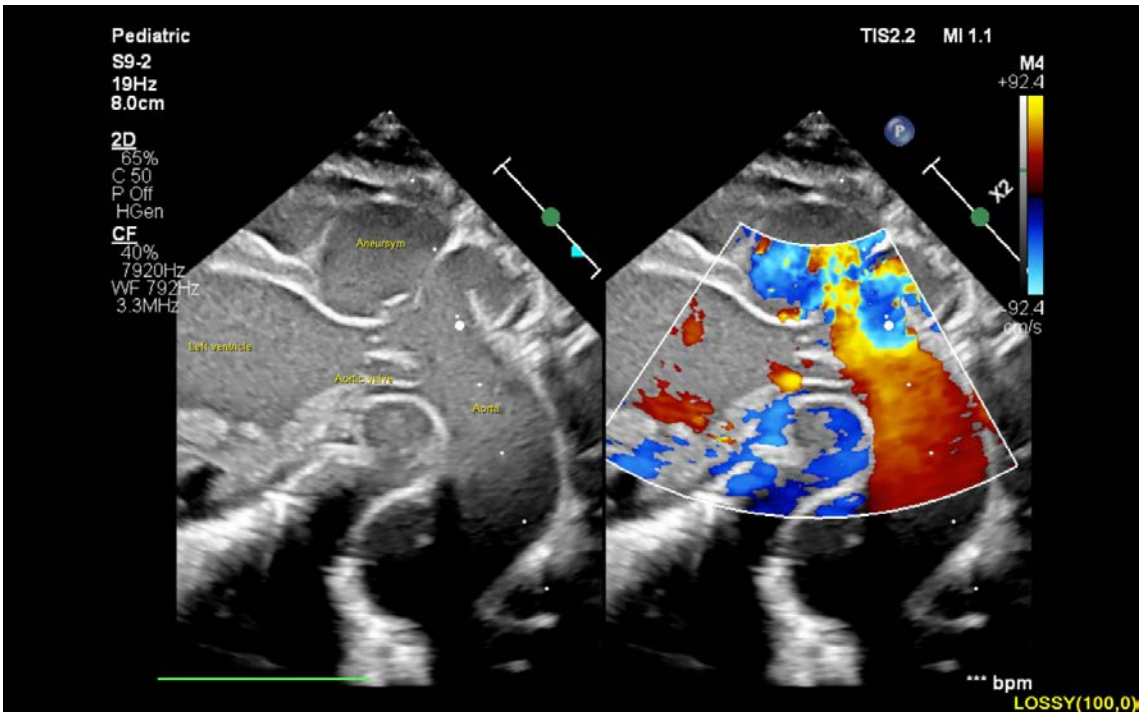
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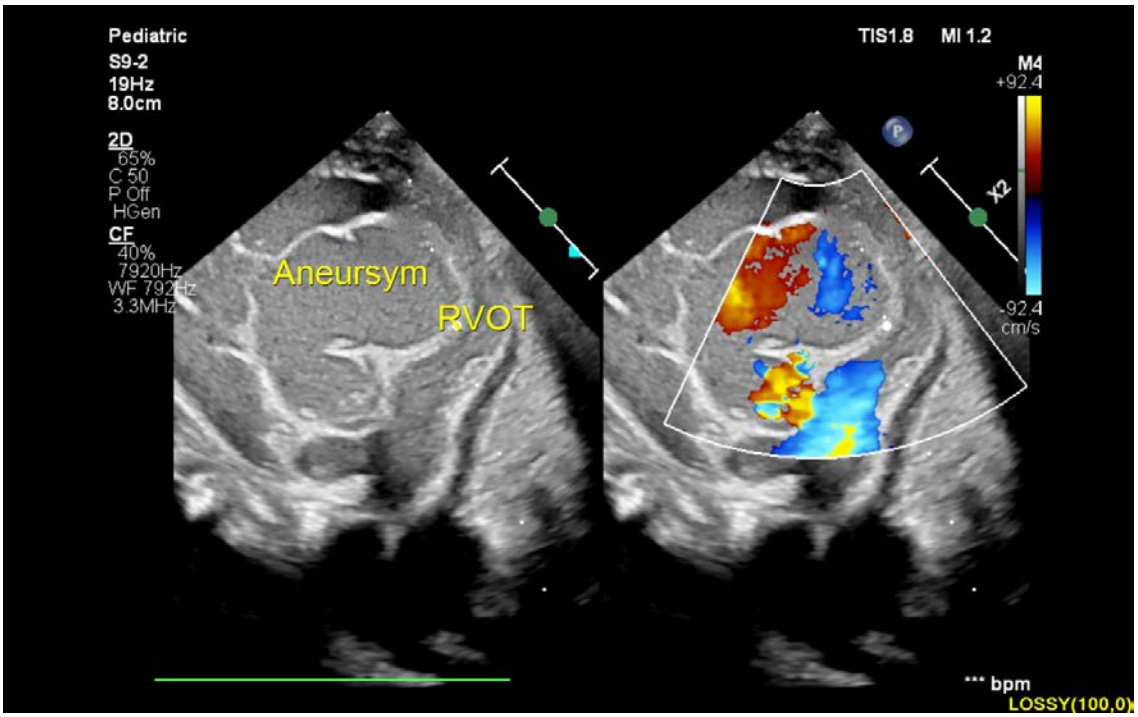
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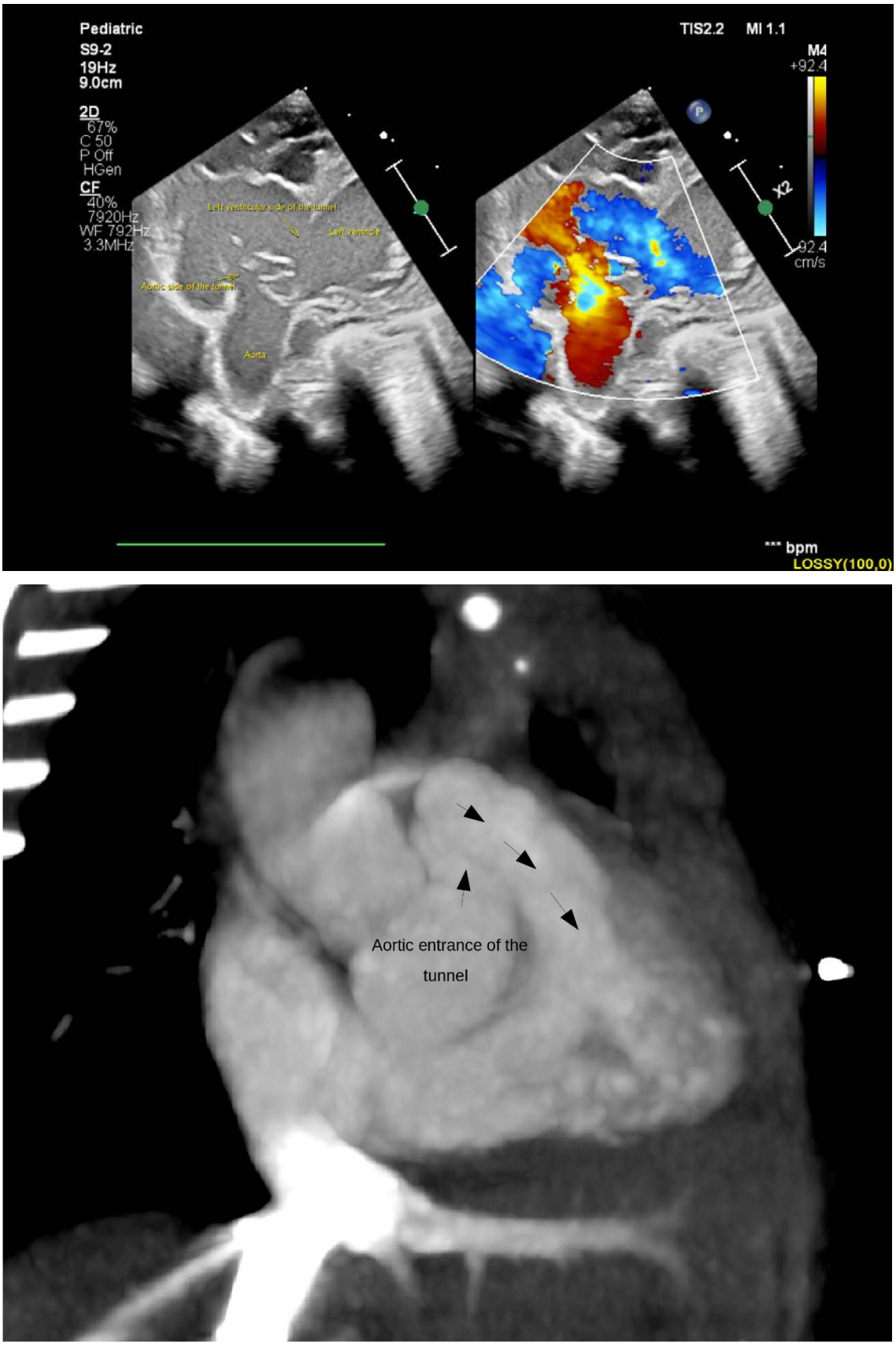

Figure 1. A. Parasternal long axis view. The aortic entrance and aneurysmatic structure of the tunnel can be seen at the sinotubular junction level. **B.** The modified subcostal image shows the aneurysm caused by the tunnel and stenosis in the right ventricular outflow tract. **C.** 5-chamber view shows the

aortic entrance of the tunnel, its course, aneurysmatic appearance and entry into the ventricle. **D.** Computerized tomography angiography shows the aortic entrance of the tunnel, its course and entry into the ventricle