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CLINICAL VIGNETTE

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Large left heart ventricle — difficulties in recognizing the Aorto-left ventricular tunnel (ALVT)

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INTRODUCTION

Aorto-left ventricular tunnel (ALVT) is an extremely rare congenital cardiac malformation, accounting for 0.03% to 0.46% of all congenital heart diseases. Since its first description as a congenital lesion in 1963, fewer than 300 cases of this heart defect have been reported in the English literature. Aorto-left ventricular tunnel creates an abnormal connection between the lumen of the left ventricle and the aorta, although less frequently, it may involve the aorta and the right ventricle. The incidence of ALVT is twice as high in boys compared to girls. Despite ongoing research, the embryological pathomechanism underlying ALVT remains elusive. However, it appears to be associated with abnormal development of the endocardial processes responsible for dividing the common trunk into the aorta and pulmonary trunk [1, 2].

CLINICAL CASE

A 26-year-old primigravida woman with no comorbidities was referred for fetal echocardiography due to suspected ventricular septal defect and tricuspid valve regurgitation detected during a second-trimester prenatal examination. At the 30th week of gestation, fetal echocardiography at a reference center revealed a disproportion between the heart chambers and a slightly enlarged heart size (heart area to chest area (HA/CA) ratio of 0.36). The left ventricle was hypertrophic and enlarged, with hypokinesis (fractional shortening of the left ventricle — 23%) and a hyperechoic endocardium. The Aortic Valve (AoV) had a Z-Score of 1.2, with normal valve leaflets and flow. The 3-vessel view (3VV) showed an enlarged main pulmonary artery and transverse aortic arch, with Z-Scores of 3.2 and 3.1, respectively. An abnormal connection between the Aorta and the Left Ventricle, characteristic of an Aorto-left ventricular tunnel (ALVT) with retrograde flow approximating 2 m/s, was also identified. The fetal middle cerebral arterial (MCA) peak systolic velocity (PSV) was 75 cm/s, likely secondary to the heart defect. Ultrasound findings are presented in Figures 1A through 1E.

The fetus exhibited supraventricular extrasystoles that did not necessitate pharmacological intervention. Despite the cardiac abnormalities, the fetus maintained efficient circulation, scoring 7 points on the cardiovascular score, with deductions for heart size and function. Standard ultrasound care was provided. At the 39th week of gestation, the patient underwent a caesarean section for obstetric reasons and delivered a son, who received an Apgar score of 8/8. On the first day of life, the newborn was transferred to the Department of Cardiac Surgery, Heart Transplantation, and Mechanical Circulation Support in Children in Zabrze, Poland. The defect was confirmed postnatally, and cardiac surgery was performed on the second day of life (Fig. 1F). The tunnel was closed in two layers, and the fistula's wall was secured with a pull-on suture and a continuous suture, preserving a connection between the aorta and the right coronary artery. The tunnel wall and aortotomy were sutured. Postoperative ultrasound confirmed complete closure of the tunnel. Echocardiography revealed slight regurgitation through the aortic valve after the procedure. The patient was started on standard treatment to improve left ventricular function, including captopril, carvedilol, and spironolactone. Currently, the boy is under outpatient follow-up, demonstrating normal development and no cardiovascular symptoms.

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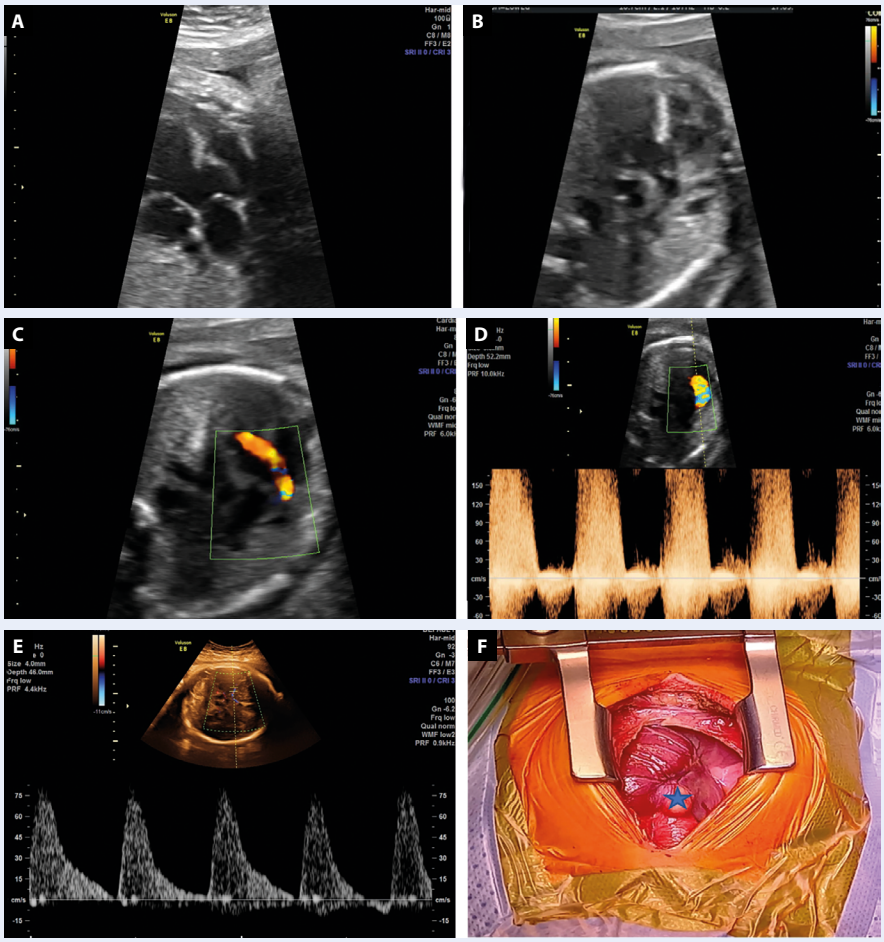

Figure 1. A. Aorto-left ventricular tunnel (ALVT) in gray scale; **B.** AVLT in gray scale; **C.** AVLT in color doppler; **D.** Retrograde flow in AVLT; **E.** Middle cerebral arterial (MCA) peak systolic velocity (PSV); **F.** Visualization AVLT during operation

DISCUSSION

The embryological pathomechanism underlying ALVT remains elusive, but it is thought to be associated with abnormal development of the endocardial processes involved in the division of the common trunk into the aorta and pulmonary trunk. According to the literature, ALVT can be diagnosed after the 18th week of gestation. A dilated left ventricle with increased wall thickness may raise suspicion of ALVT. Approximately 45% of ALVT cases are associated with other heart defects, often coexisting with aortic valve abnormalities [1, 3, 4]. The 'cockade sign', indicative of a tunnel adjacent to the aortic annulus, can aid in making a correct diagnosis [5]. An elevated peak systolic velocity in the fetal middle cerebral artery may also assist in diagnosis. Differential diagnoses include aortic regurgitation, Valsalva fistula, common arterial trunk (CAT), aortopulmonary window, and coronary ventricular fistula. The standard cardiac repair procedure involves closing the tunnel with a patch inserted through an aortotomy [6]. Perioperative mortality is low, and prompt intervention is recommended. Surgical correction is indicated not only to prevent heart failure but also to halt the progression of aortic valve damage. Patients who undergo surgery within the first six months of life have the best-documented normalization of left ventricular size and function [2, 7–9].

Article information and declarations

Ethics statement

None.

Conflict of interest

The authors declare no conflict of interest.

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