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RESEARCH ARTICLE

ACUTE RESPIRATORY DISTRESS REVEALING A COMPLETE ATRIOVENTRICULAR CANAL IN A 22-MONTH INFANT

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Abstract

The atrioventricular canal (AVC) is a complex and rare congenital cardiac anomaly. It is potentially severe due to the complexity of its lesions and associated fixed pulmonary arterial hypertension. We report the case of a 22-month-old infant girl with Trisomy 21 syndrome, admitted for acute respiratory distress. TTE showed a complete AVC with pulmonary arterial hypertension. Prognosis depends on the promptness of diagnosis and therapeutic management.

Introduction:-

The atrioventricular canal (AVC) is a complex heart condition, accounting for less than 4% of congenital heart diseases (CHD) [1]. It is most commonly discovered during genetic anomalies. Various anatomical variations of AVC are distinguished, including partial, complete, or intermediate forms.

We report the case of a 22-month-old girl infant with Trisomy 21, admitted for dyspnea with signs of respiratory distress (SRD) associated with anemia. Transthoracic echocardiography (TTE) revealed a complete AVC with pulmonary arterial hypertension (PAH).

Case report

A 22-month-old girl infant, born to non-consanguineous parents, the youngest of a sibling of 9, from a rural background, was admitted for respiratory distress syndrome (severe dyspnea andthoracoabdominal rocking) associated with anemia at 5g/dL. Maternal history revealed a well-followed pregnancy, carried to term, with a normal vaginal delivery and no neonatal complications. Personal history included two hospitalizations for viral bronchiolitis at the age of 5 months and a Trisomy 21.

On admission, the general examination found a conscious infant with hemoglobin saturation at 79% in ambient air, increasing to 90% after oxygen administration. No cyanosis was observed, heart rate was normal at 125 beats per minute, respiratory rate was 34 cycles per minute, afebrile at 36.7 degrees Celsius, and no signs of peripheral hypoperfusion.

Cardiovascular examination revealed a systolic murmur, 3/6, maximum at the apex, and a split of the second heart sound. Pulmonary examination did not reveal any rales on auscultation.

Chest X-ray showed cardiomegaly, a cardiothoracic index of 0.65, and mild pulmonary vascularity (Figure 1).

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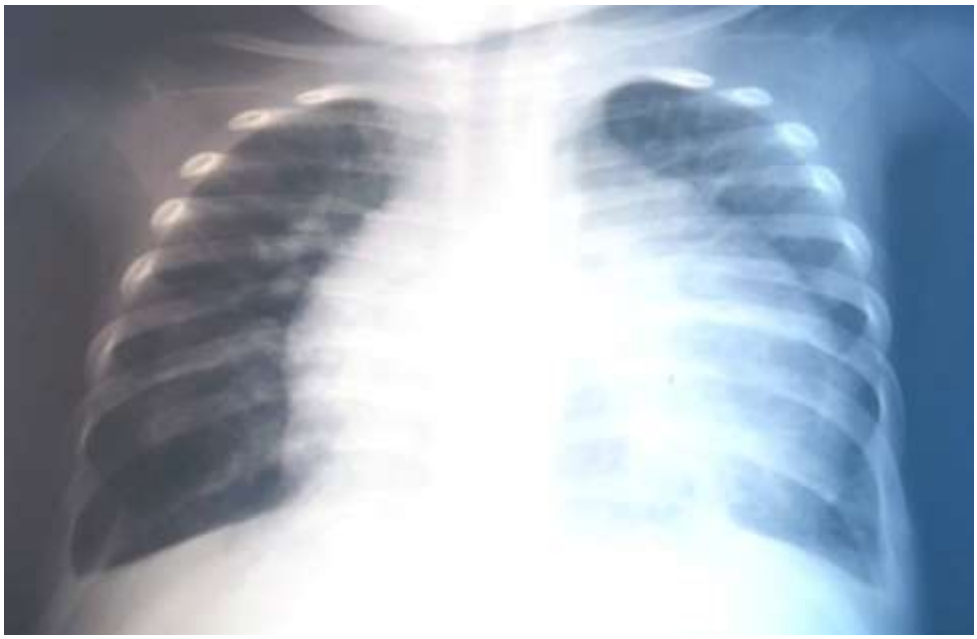

Figure 1: Frontal chest X-ray showing cardiomegalymild pulmonary vascularity.

Transthoracic echocardiography (TTE) revealed a significant 4 cm ventricular septal defect (VSD) with a common atrioventricular valve and severe regurgitation. There was also a peak gradient across the right ventricle and right atria measuring 88 mmHg, along with a non-dilated compressible inferior vena cava. Additionally, moderate pericardial effusion was observed around the right ventricle without signs of tamponade. Although right ventricular hypertrophy was present, there was no dilation, and the left ventricular contractility was good. Furthermore, there was no evidence of pulmonic valvulopathies or aortic coarctation.

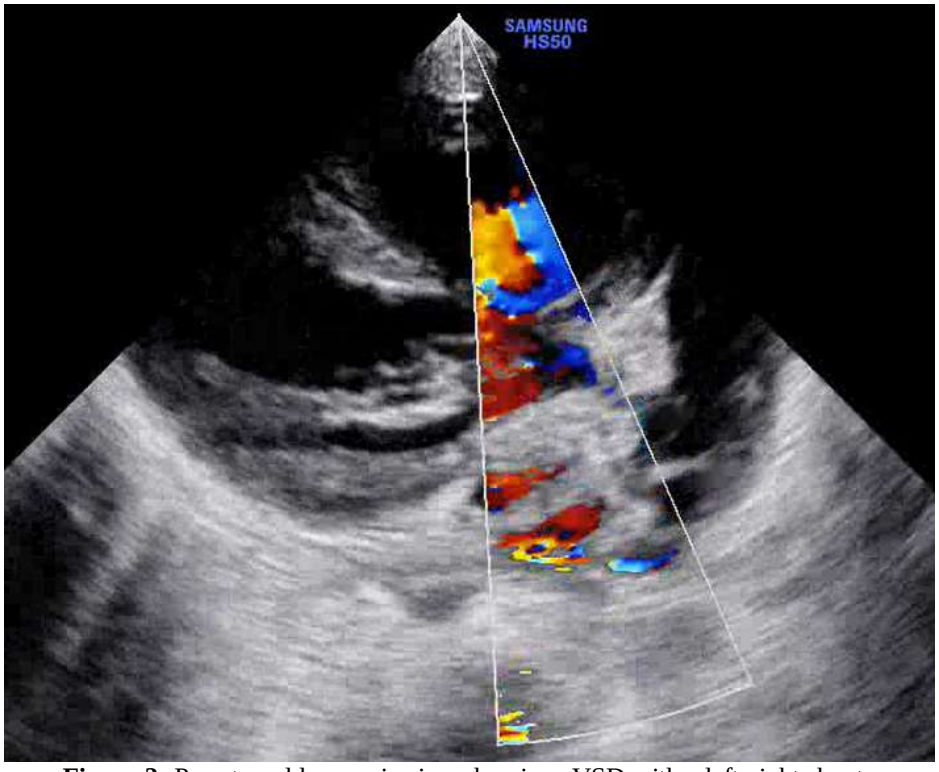

Figure 2: Parasternal long-axis view showing aVSD with a left-right shunt.

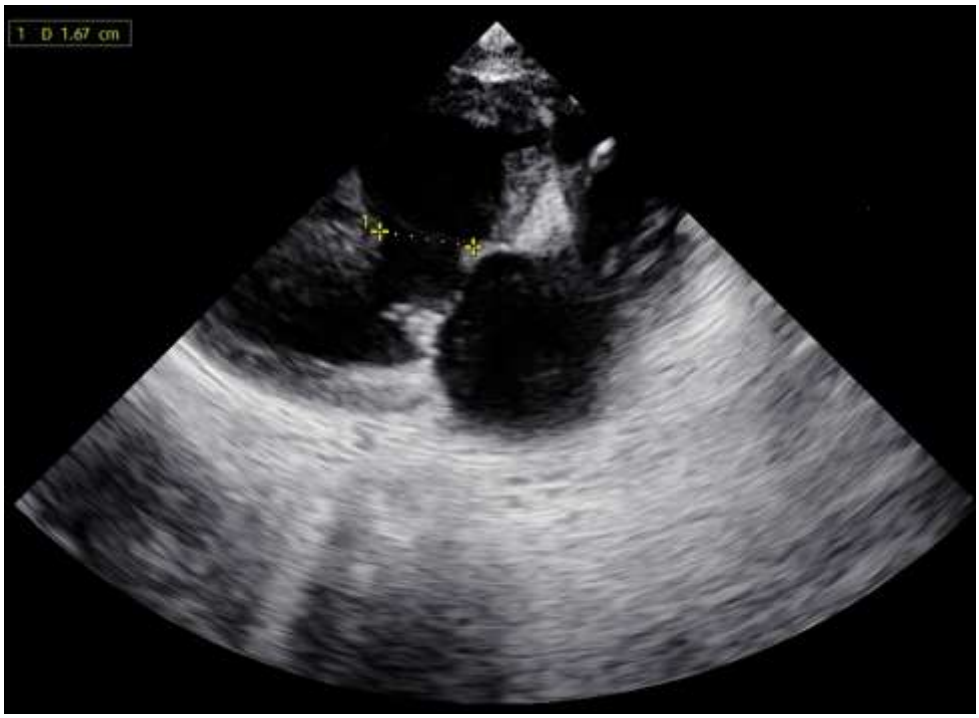

Figure 3: Modified parasternal long-axis view showing the 1.67cm VSD.

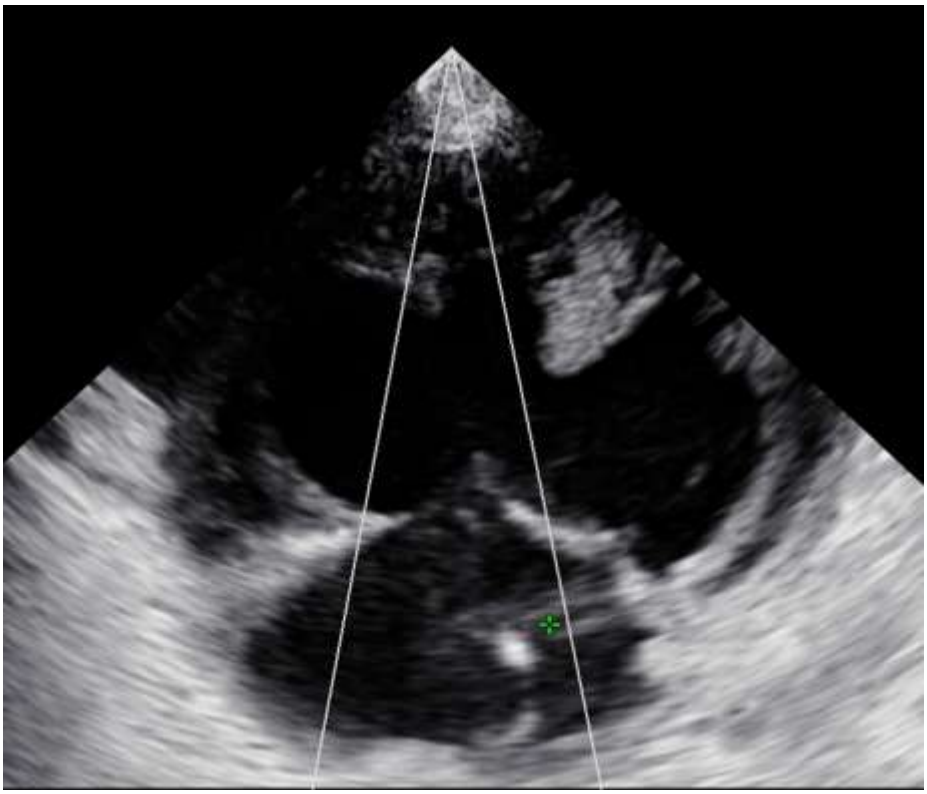

Figure 4: Apical Four chamber view showing the common atrioventricular valve, and the ASD.

Discussion:

Complete AVC is a congenital heart defect observed in about 20% of patients with Trisomy 21 [1]. Discovery of this heart defect in adulthood is exceptional; half of the patients die within the first 24 months of life [2].

Echocardiography performed early, around the 18th week of pregnancy easily diagnoses complete AVC. Its search is systematic in any newborn carrying a genetic anomality such as Trisomy 21 [3]. TTE is a non-invasive and an easy exam that allows diagnosis and specifies the anatomical form and associated lesions while assessing its hemodynamic impact [4].

The partial form of AVC is much more common and less severe than the complete form [1]. The complete form is characterized by the absence of atrioventricular septal structures and the presence of a single atrioventricular valve, with the primum ostium and the admission interventricular communication in continuity. The partial form presents a low primum-type interatrial communication with a mitral cleft [5]. These forms result from a developmental anomaly of the buds during embryonic life [6].

The diagnosis of our 22-month-old infant is a complete AVC, confirmed by TTE with significant PAH at 80 mmHg. As described in the literature, two-dimensional echocardiography allows visualization of 04 chambers in apical section, continuity between the primum ostium with a defect of the atrioventricular septum, and the admission interventricular communication with fusion of the atrioventricular valves [7]. Color Doppler allows the study of regurgitant flows through the atrioventricular valves and the visualization of shunts. It also assesses systemic pulmonary arterial pressure (PAPS) and its impact [1]. TTE in our patient revealed an interatrial communication, an admission interventricular communication, a single atrioventricular valve, and a PAPS of 80 mmHg.

The treatment of complete AVC relies on surgery. Recommendations advise performing it between the third and sixth month of life before the establishment of elevated systemic vascular resistances [8]. The repair involves closing the septa under extracorporeal circulation using synthetic patches. It is done through a right atriotomy. The risk of mortality remains high (around 8.7%) [9]. Postoperative prognosis depends on the promptness of management and residual mitral regurgitation, which may require later reintervention [10].

Our patient was referred for cardiovascular surgery.

Conclusion:

In the presence of Trisomy 21, the systematic search for congenital heart disease is essential. Complete AVC is a rare but complex manifestation. The diagnosis is easily made by transthoracic echocardiography. The speed of surgical intervention (before the first 6 months of life) is an important prognostic factor before the onset of fixed pulmonary arterial hypertension.

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