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Holms Heart in a Fetus with Maternal Anticonvulsant Drug Exposure: A Case Report

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ABSTRACT

Double inlet left ventricle (DILV) is a rare congenital cardiac malformation that is defined as an anomaly with univentricular atrioventricular (AV) connection, and single ventricular morphology. This variation can be associated with the inlet and outlet cardiac abnormalities. In the current report, we present a case of Holms heart, as a rare variant of double-inlet left ventricle.

Introduction

Double-inlet left ventricle (DILV) is a rare congenital cardiac anomaly with an incidence of 0.05 to 0.1 per 1000 live births. This anomaly is a one of the most common forms of univentricular atrioventricular connection or single ventricular morphology, and includes about 60–70% of all forms [1, 2, 3]. In the fetus, double inlet ventricle exists when the most part of both atrioventricular (AV) valves receive blood by a single ventricular chamber (in DILV with the chamber of left ventricle morphology). This anomaly contains various groups of cardiac defects that can involve atrioventricular valves, ventriculoarterial connection and systemic or pulmonary outflow tract [1, 2]. Holms heart, a variant of DILV, is a rare sub type, first described in 1824 by Andrew F. Holms. In this anomaly, the aorta originates from left ventricle (LV) and a fibrous

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continuity exists between aorta, mitral valve and tricuspid valve, indicating that the great arteries are normally related. A small bulbo-ventricular foramen (BVF) is located between large size LV and the right ventricle, which again confirms that the great arteries are normally related [4]. Morphologically, the right ventricle (RV) sinus is absent, but the LV sinus is present [4].

Case presentation

A 43-year-old G1-P0-L0 pregnant woman, with history of 15 years primary infertility and long-term usage of anti-convulsant drugs (Carbamazepine), referred to our clinic at 27 weeks gestation. In fetal echocardiography evaluation, cardiac orientation and position were normal (levocardia and levoposition). Apical four-chamber view showed hypoplastic, small and rudimentary right ventricle morphology and also large ventricular septal defect (VSD) (Fig. 1). In the short axis view of great vessels, normal relations between aorta (AO) and pulmonary artery (PA) and ventriculoarterial (VA) concordance (PA connection to RV) were seen (Fig. 2). Large inlet type VSD was also shown in another view of all cavities. (Fig. 4). Color Doppler placement through AV valves did not show significant regurgitation. A girl baby was born at 38th of gestational age without respiratory signs

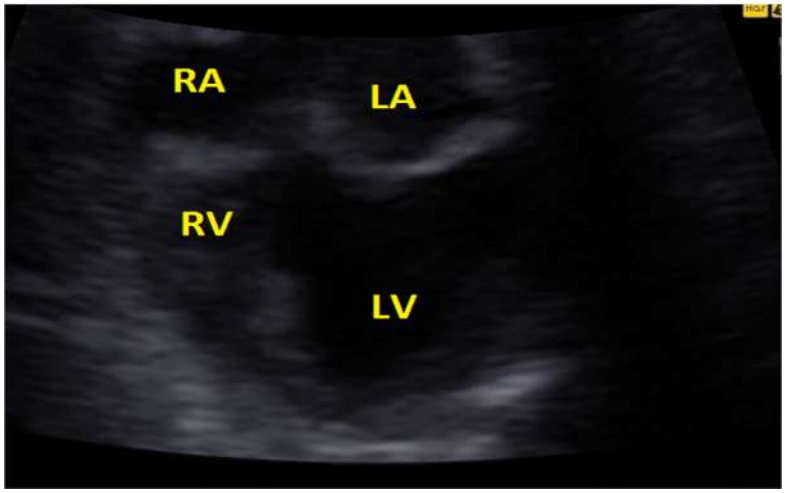

Fig. 1. Apical 4 chamber view shows single ventricle LV morphology, right atrioventricular valve overriding (due to double inlet left ventricle), small hypoplastic rudimentary (outlet) right ventricle.

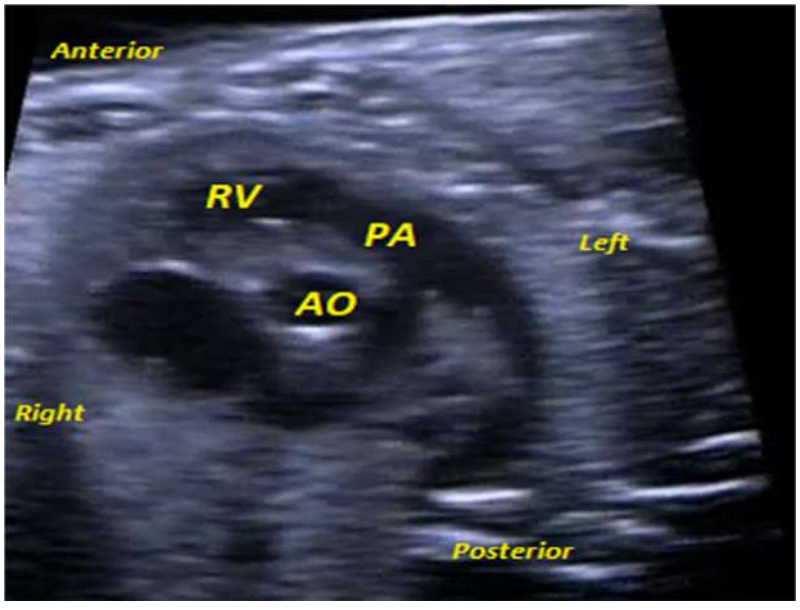

Fig. 2. A short axis view of great vessles shows normaly related great arteries (left and anterior PA, right and posterior AO).

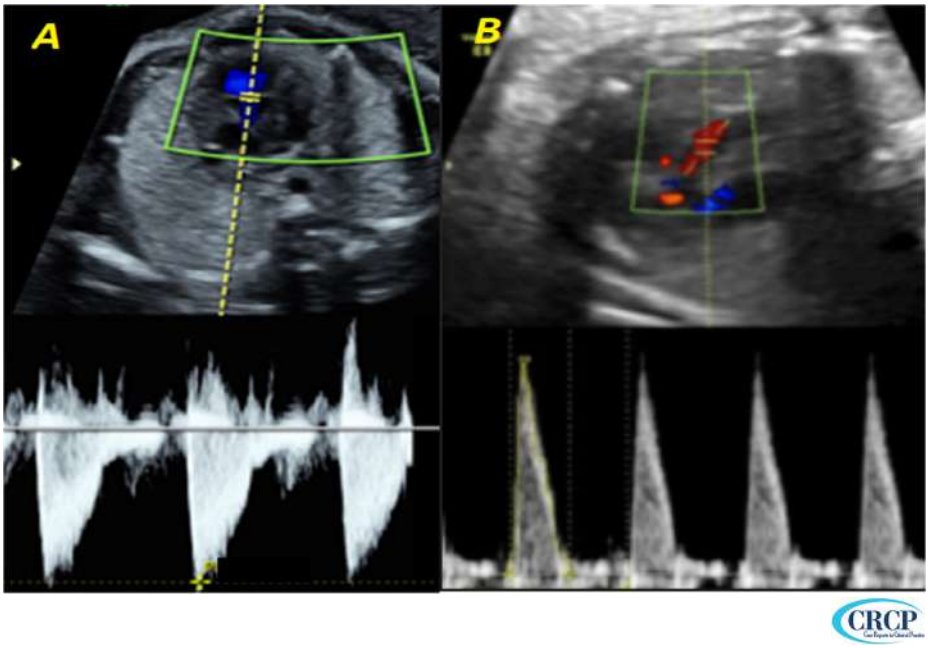

Fig. 3. Doppler spectral of PA (A) and Aortic root (B) shows normal pattern (rapid early systolic spike and slow deceleration) and normal velocity (less than 80ms in PA and about 90ms in AO).

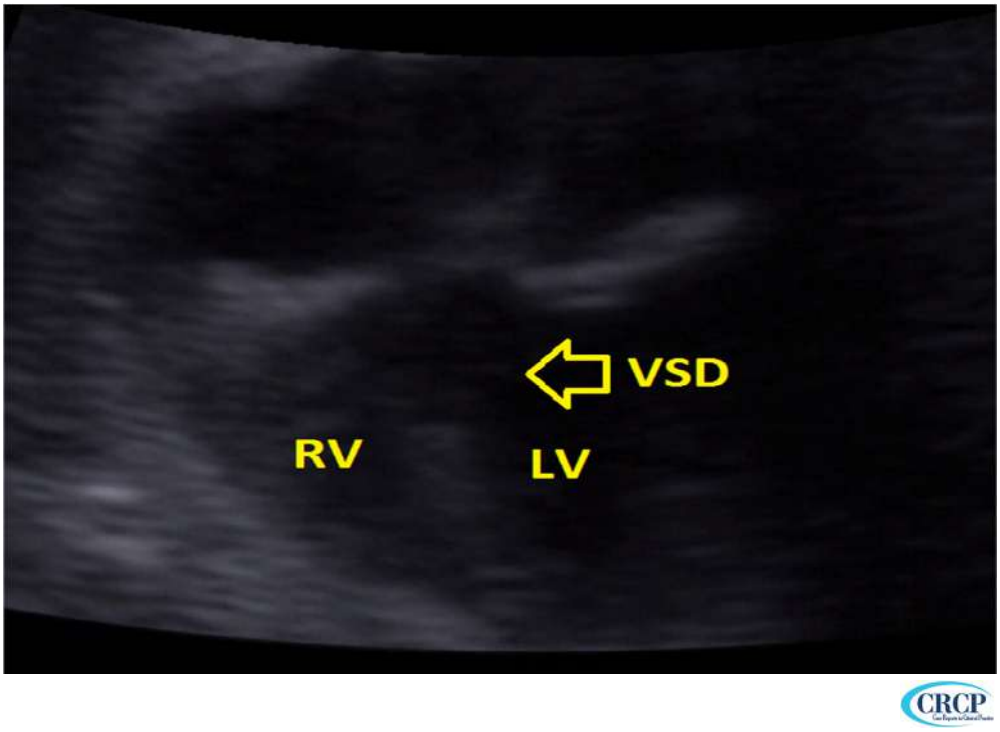

Fig. 4. A 4 chamber view shows large inlet type ventricular septal defect.

and trans-thorasic echocardiography was performed for her. Para sternal long axis view showed single ventricle with left ventricular morphology, large inlet type VSD, right AV valve overriding and hypoplastic RV (Fig. 5, 8). Apical four-chamber view accentuated univentricular AV connection (Fig. 6). In short axis view of great arteries, normal relation between aorta and pulmonary was confirmed (Fig. 7).

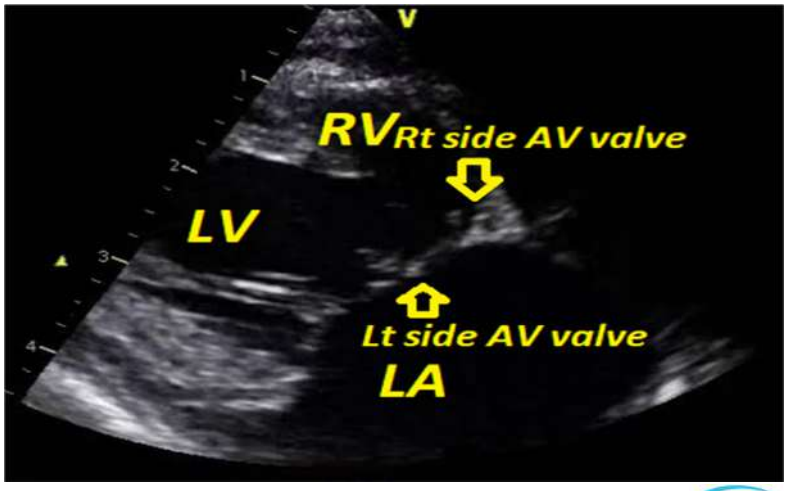

Fig. 5. Echocardiography after birth. Para sternal long axis view shows LV dominance, small RV. Right (Rt) and left (Lt) AV valve dominantly connected to LV and Rt AV valve (overriding).

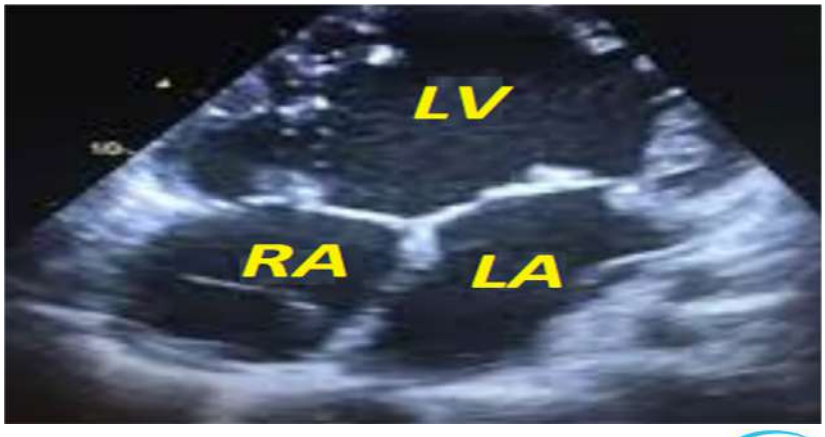

Fig. 6. Apical 4 chamber view shows single LV morphology and dominat connection of both atriums to LV.

Fig. 7. Para sternal short axis view shows normal relation between AO and PA (great arteries) and PA connection to small RV.

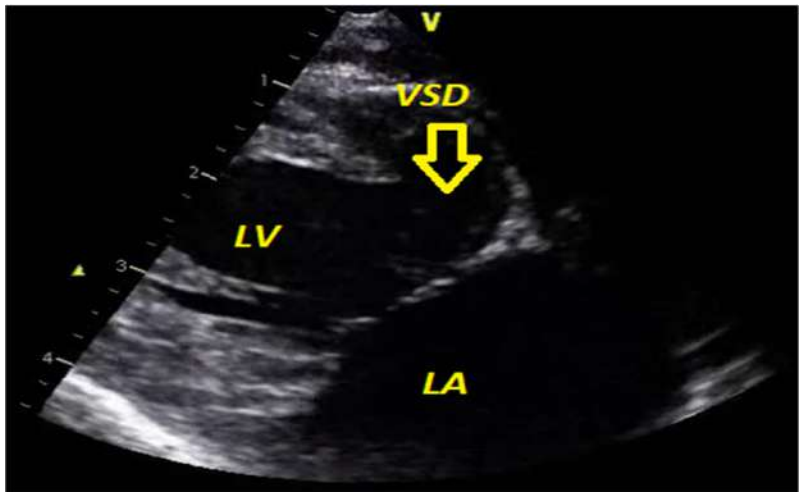

Fig. 8. Para sternal long axis view shows a large inlet type VSD.

Discussion

Holms heart is a rare and complex congenital heart disease. Prenatal diagnosis of this anomaly has increased over the last decades. This abnormaity and associated anomalies can be diagnosed prenatally due to an abnormal four-chamber and outflow tract views with fetal echocardiography and ultrasound [1,4]. However, the diagnosis of single ventricle case is not very difficult on fetal echocardiography, but it could quite challenging when there exists an abnormal four-chamber, two atria joined to the same ventricle and without interventricular septum. Solitary ventricles are extremely rare and in most cases an undeveloped chamber neighboring the main ventricle can be seen. Holmes heart consists of normally related great vessels, but in the most types of DILV, a transposition of great arteries occurs. The clinical outcome and surgical plan for the neonates with DILV depend on the obstruction of outflow tract and the connection of the great arteries. In Holmes heart, ductus arteriosus can be the only source of pulmonary blood flow, however, high pulmonary blood flow can exist and lead to congestive heart failure [3]. Prenatal diagnosis allows delivery to be planned at a tertiary care facility with good postnatal management, and also helps to discuss and counsel with parents about this serious congenital heart anomaly and prepare them to make decision for pregnancy termination or continiuation [1,4].

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

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