Atrioventricular Discordance with Double-Outlet Right Ventricle in Mirror Imagery and Levocardia: A Very Rare Case Report

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Abstract

A newborn without prenatal diagnosis, with bronchial and abdominal situs inversus in levocardia, was referred to our hospital for accurate evaluation; echocardiography showed venoatrial connections in mirror-image arrangement, atrioventricular (AV) discordance, and double-outlet right ventricle (DORV). Additional cardiac malformations were double upper caval district, atrial communication, subpulmonary interventricular communication, and moderate subvalvular and valvular pulmonary stenosis. Few days after birth, the patient presented low oxygen saturation and the heart team decided for a palliative surgery. We describe a very rare case in a newborn with bronchial-abdominal mirror imagery, AV discordance, and DORV in levocardia.

Keywords: Atrioventricular discordance, double-outlet right ventricle, levocardia, mirror-imagery

Introduction

The discordant atrio-ventricular (AV) connection is a rare cardiac malformation in which the ventricles are connected inappropriately to the atrial chambers: the morphologically right atrium, receiving systemic venous blood, is connected to the morphologically left ventricle, and the morphologically left atrium, receiving pulmonary venous blood is connected to the morphologically right ventricle[1]. We report a very rare case of a complex combination of cardiac malformations with bronchial and abdominal situs inversus, levocardia, AV discordance, double-outlet right ventricle (DORV), subpulmonary interventricular communication, subvalvular and valvular pulmonary stenosis and right aortic arch. Multi-modality imaging has been effective to integrate cardiac and extracardiac anatomical details and to allow an accurate diagnosis before surgical treatment.

Case Report

A 1-day-old male patient, born at 41 weeks’ gestation after a normal pregnancy, was referred to our hospital for low oxygen saturation. His body weight was 3.4 kg, at birth Apgar Index was 9–10, arterial blood gas showed metabolic acidosis, and peripheral oxygen saturation was 94%; on physical examination, a Grade 3/6 parasternal systolic murmur was noted on auscultation. The newborn did not present extracardiac malformations associated. A chest radiograph showed levocardia, bronchial situs inversus, left-sided liver, and right-sided stomach. A sequential segmental echocardiographic analysis confirmed abdominal arrangement [Figure 1c] and levocardia. Based on the appendage shape, right-sided appendage seemed to have left morphology and left-sided appendage seemed to have right morphology [Figures 2a, 3c and 4d, Video 1]. Venoatrial connections were in mirror-image arrangement: pulmonary veins drained in the right-superior atrial roof, left inferior and left dominant superior caval veins drained in the atrial chamber placed on the left-sided. A minor right superior caval vein flowed in the atrial chamber placed on the right side [Video 1]. Coronary sinus was absent. An interatrial communication allowed a complete

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blood flow mixing in both atrial chambers [Figure 2b and Video 2]. Moreover, four-chamber view [Figure 2d] showed a left intra-atrial membrane parallel to AV floor, not causing inflow obstruction. D-loop ventricular arrangement. The morphologically right ventricle was connected to the atrial chamber sited on the right through trileaflet tricuspid valve and the morphologically left ventricle was connected to the atrial chamber sited on the left through bivalvelet mitral valve as shown in short-axis subcostal view [Figure 3 and Video 4]. Biventricular systolic function was preserved. There was a subpulmonary interventricular communication. Both great arteries arose from the right ventricle with the aorta anterior, rightward, and parallel to the pulmonary trunk [Figure 3 and Video 4]. Color-flow imaging revealed a moderate subvalvular and valvular pulmonary stenosis, with bicuspid and dysplastic valve. Aortic arch and patent ductus arteriosus were both on the right side [Figure 1]. This is a very rare case of AV discordance, double-outlet right ventricle (DORV) in levocardia, and bronchial-abdominal mirror imagery [Figures 1-4].

Contrast-enhanced computed tomography (CT) confirmed the diagnosis. Both great arteries arose from the right ventricular outflow tract. The aortic valve was located anteriorly and to the right of the pulmonary valve. The left-hand sinus gave rise to the left main coronary artery, and the right-hand sinus gave rise to the right coronary artery [Figures 3b and 4e].

One week after birth, pulmonary resistance decreased and oxygen saturation reduced to 70%, therefore the heart team decided to treat the patient through a systemic-pulmonary shunt. During open-heart surgery, an usual pattern of coronary arteries and confirmed D-loop ventricular morphology were described.

**DISCUSSION**

The arrangement of thoracoabdominal organs in mirror-image variant, also described as situs inversus, is a lateralization pattern other than normal and represents a horizontal flip in the orientation of bodily organs. According to the Anderson’s morphological method,[2] the analysis of cardiac arrangement must begin identifying structures on the basis of their most constant part, then for the atrial segment with the morphology of the atrial appendages. The distinguishing feature of atrial appendages is the extent of pectinate muscles to the AV junctions; this helps to identify the isomeric variants from those having mirror imagery. Only the atrial appendages are truly isomeric in the heart.

The International Nomenclature Committee excluded mirror imagery when defining “heterotaxy,” restricting this one to the lesions characterized by the presence of isomerism of the atrial appendages. Right atrial appendage isomerism is characterized by the extension of pectinate muscles bilaterally to the crux, while in the left appendage isomerism, atrial vestibules are smooth.[3]

Differentiation between morphologically right and left atrial appendages remains hard to judge by echocardiography. CT angiography allows an accurate diagnosis of abnormal cardiac arrangements.[4] In our case, echocardiography images suggested atrial appendage morphology as mirror image; CT angiography confirmed appendage arrangement on the basis of pectinate muscle extension toward the AV junction. Moreover, as it is known, the arrangement of the atrial appendages is usually well correlated with the bronchial morphology,[1] as in our case.

Discordant AV connection in association with DORV is an extremely rare malformation which can be easily misdiagnosed.
with congenital corrected transposition of great arteries and ventricular septal defect (VSD), as both malformations are morphologically similar. Physiologically, blood from the morphologically right atrium goes across the left ventricle and passes preferentially to the pulmonary arteries, as the pulmonary trunk is usually close to the interventricular communication.\[^1\]

In literature,\[^6,7\] similar cardiac malformations have previously been reported. In 1994, Imai \textit{et al.} described 18 cases with AV discordance, but the combination described in our case of visceral situs inversus, levocardia, AV discordance, interatrial communication, left intra-atrial membrane, DORV, interventricular communication, subvalvular and valvular pulmonary stenosis, and right aortic arch, has not been reported until now. Aherrera \textit{et al.} reported a 19-year-old female referred
for exertional dyspnea with a final diagnosis of atrial and visceral situs inversus with malposition of the great vessels, DORV, complete AV septal defect with a common AV valve, bilateral caval veins, and pulmonary stenosis. Similar case in adulthood has been reported by Goyal et al., describing a 16-year-old female with situs inversus and congenitally corrected transposition of the great arteries, dextrocardia, VSD, and pulmonary stenosis. Chang et al. described a rare similar case of a triply reversed heart in a 57-year-old female referred for dyspnea and situs inversus with congenitally corrected transposition of the great arteries.

Usually, thoraco-abdominal mirror imagery occurs in dextrocardia; isolated levocardia is a rare condition in this arrangement. The reported incidence is 1/22,000 in the general population and from 0.4% to 1.2% in all patients with congenital heart diseases. Data about isolated levocardia suggested the association with other complex congenital cardiac defects and extracardiac malformations, especially volvulus or bowel obstruction, which were absent in our case.[8]

In summary, our case is a very rare arrangement of bronchial and abdominal situs inversus, levocardia, AV discordance, interatrial communication, intra-atrial membrane, double superior caval district, DORV, subpulmonary interventricular communication, subvalvular and valvular pulmonary stenosis, and right aortic arch. The peculiar feature of this case is the complex combination of numerous cardiac malformations which have been never reported in a newborn until now; echocardiographic diagnosis integrated with CT scan allowed to exactly define the cardiac and extracardiac anatomy and to achieve an optimal surgical result.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

References
1. Wilkinson JL, Anderson RH. Anatomy of discordant atrioventricular connections. World J Pediatr Congenit Heart Surg 2011;2:43-53.
2. Freedom RM, Jaeggi ET, Lim JS, Anderson RH. Hearts with isomerism of the right atrial appendages - one of the worst forms of disease in 2005. Cardiol Young 2005;15:554-67.
3. Anderson RH, Spicer DE, Loomba R. Is an Appreciation of Isomerism the Key to Unlocking the Mysteries of the Cardiac Findings in Heterotaxy? J Cardiovasc Dev Dis 2018;5(1):11.
4. Wolla CD, Hlavacek AM, Schoepf UJ, Bucher AM, Chowdhury S. Cardiovascular manifestations of heterotaxy and related situs abnormalities assessed with CT angiography. J Cardiovasc Comput Tomogr 2013;7:408-16.
5. Loomba RS, Pelech AN, Shah PH, Anderson RH. Determining bronchial morphology for the purposes of segregating so-called heterotaxy. Cardiol Young 2016;26:725-37.
6. Aherrera JA, Magno JD, Uy CC, Abrahan LL 4th, Maria HF, Buitizon RR, et al. The triply twisted heart: Cyanosis in an adult with situs inversus, levocardia, double outlet right ventricle, and malposition of the great arteries. Cardiol Res 2015;6:362-6.
7. Imai Y, Sawatari K, Hoshino S, Ishihara K, Nakazawa M, Momma K. Ventricular function after anatomic repair in patients with atrioventricular discordance. J Thorac Cardiovasc Surg 1994;107:1272-83.
8. Gindes L, Hegesh J, Barkai G, Jacobson JM, Achiron R. Isolated levocardia: Prenatal diagnosis, clinical importance, and literature review. J Ultrasound Med 2007;26:361-5.