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Atrioventricular Discordance with Pulmonary Atresia, Double-outlet Right Ventricle, and Isolated Levocardia: A Case Report of a Rare Congenital Heart Disease

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Abstract: Almost all cases of isolated levocardia or situs inversus with levocardia are associated with congenital heart disease. We report a case of situs inversus with levocardia, double-outlet right ventricle, atrioventricular discordance, pulmonary atresia, patent ductus arteriosus, atrial septal defect, and ventricular septal defect in a neonate. An overview of anatomy and relevant definitions with radiographic correlation will be presented along with a synopsis of current medical literature.

Keywords: *congenital heart disease, situs inversus, isolated levocardia, double-outlet right ventricle, atrioventricular discordance*

Case Presentation

The patient is a monoamniotic-monochorionic twin who in utero had an atrioventricular canal defect found on fetal ultrasound. The patient was delivered via cesarean section at the 34th week of gestation. Transthoracic echocardiogram confirmed complex ductal-dependent congenital heart disease. With stable oxygen saturation in the high 70% to 80% range, the patient was admitted to the intensive care unit for management of prematurity-related disorders and administration of prostaglandin to maintain the opening of the patent ductus arteriosus (PDA). The patient underwent cardiac magnetic resonance (MR) imaging with ferumoxytol for further delineation of cardiovascular anatomy. The imaging revealed situs inversus and levocardia (Figures 1, 2). A computed tomography coronary arteriogram showed a trilobed left lung and

Key Points

- Situs inversus with levocardia is rare and typically associated with congenital heart disease.
- Radiologist's familiarity with a variety of possible defects, including double-outlet right ventricle and levo-transposition of the great arteries, can help in image-based preoperative planning.

Abbreviations

PDA: patent ductus arteriosus
MR: magnetic resonance
SVC: superior vena cava
IVC: inferior vena cava
DORV: double-outlet right ventricle
L-TGA: levo-transposition of the great arteries
CHD: congenital heart disease

Figure 1. Computed Tomogram, Sagittal View, of the Left Lung in a Neonate with Situs Inversus and Isolated Levocardia

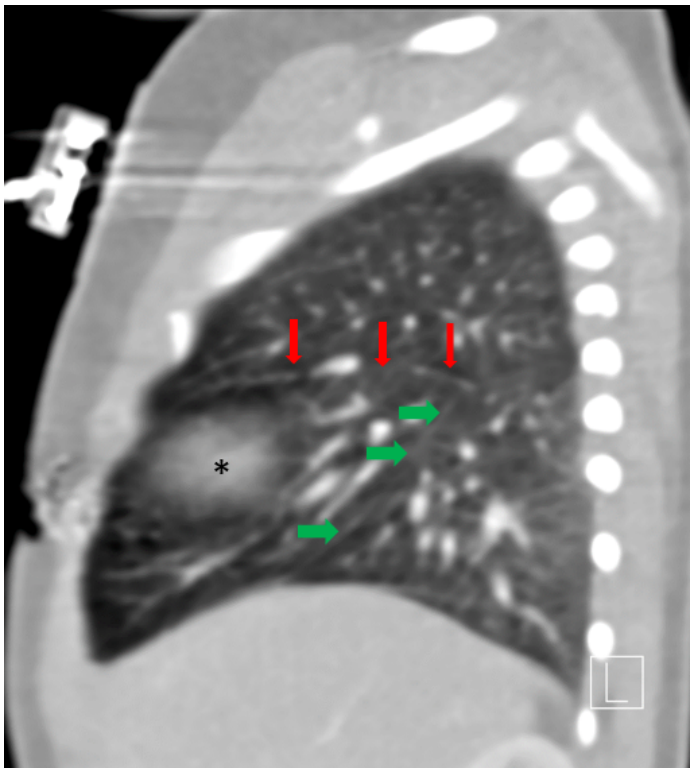
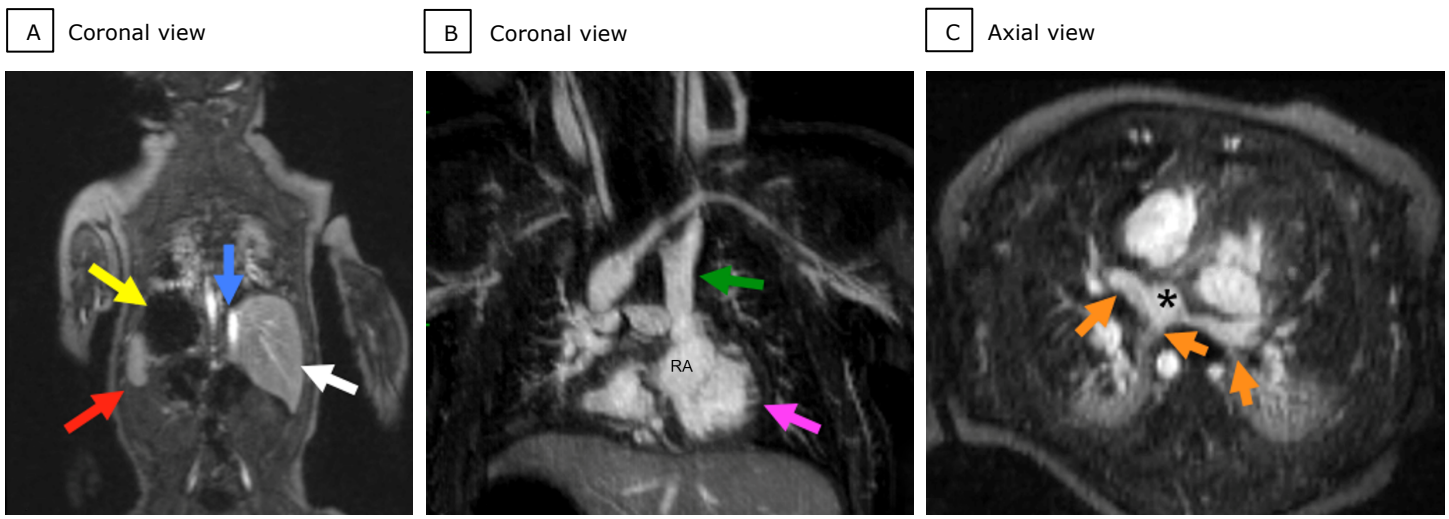


Image shows a trilobed left lung with minor fissure (red arrows) and a major fissure (green arrows). The cardiac apex (asterisk) is seen, proving the position of the left lung.

inversed branches of a right-sided aortic arch. On MR angiography, the superior vena cava (SVC) was on the left side, and the inferior vena cava (IVC) was coursing uninterrupted beyond a left-sided liver and continuing with conventional branching in the left paraspinal region (Figure 2). Within the abdomen, in addition to a left-sided liver, imaging showed right gastric bubble and a right-sided spleen (Figure 2).

Further analysis of the patient's cardiac anatomy on MR angiography showed evidence of pulmonic stenosis, right ventricular hypertrophy, double-outlet right ventricle (DORV), atrial septal defect, and ventricular septal defect (Figure 3). Moreover, levo-transposition of the great arteries (L-TGA)—atrial inversion with a left-sided morphologic right atrium receiving systemic venous return via a left-sided SVC and the IVC and a right-sided morphologic left atrium receiving pulmonary venous return—was observed (Figure 3). In addition to the right ventricle being connected to the aorta, there was obstruction of the right ventricular outflow tract secondary to pulmonary atresia (Figure 3). These right ventricular anatomic findings are hallmarks of DORV. The atrial and the ventricular septal defects provided admixture of flow. Because of the severe pulmonic stenosis, the patient's survival depended on an

Figure 2. Contrast-enhanced Cardiac Magnetic Resonance Imaging of Situs Inversus with Levocardia in a Neonate



Coronal view (A) shows inversion of abdominal viscera with a left-sided liver (A, white arrow), a right-sided stomach (A, yellow arrow), and a right-sided spleen (A, red arrow). A left-sided inferior vena cava (A, blue arrow) is seen traversing through the liver. Coronal view (B) shows a left-sided superior vena cava (B, green arrow) draining into the morphologic right atrium (RA) connected to the morphologic left ventricle (B, pink arrow). Axial view (C) shows the pulmonary veins (C, orange arrows) draining into a right-sided morphologic left atrium (C, black asterisk).

extremely large and tortuous PDA, which shunted blood from the aorta into the pulmonary arterial circulation distal to the stenosis.

After the MR examination, the patient underwent stenting of the PDA and the left pulmonary artery. The procedure intended to preserve the patient's ability to produce oxygenated blood via the PDA shunting. The patient's oxygen saturation was between 80% and 90%. Following the procedure, the patient was discharged from the hospital at 8 weeks of age. The patient was recommended to gain weight prior to the next stage of surgical intervention.

Discussion

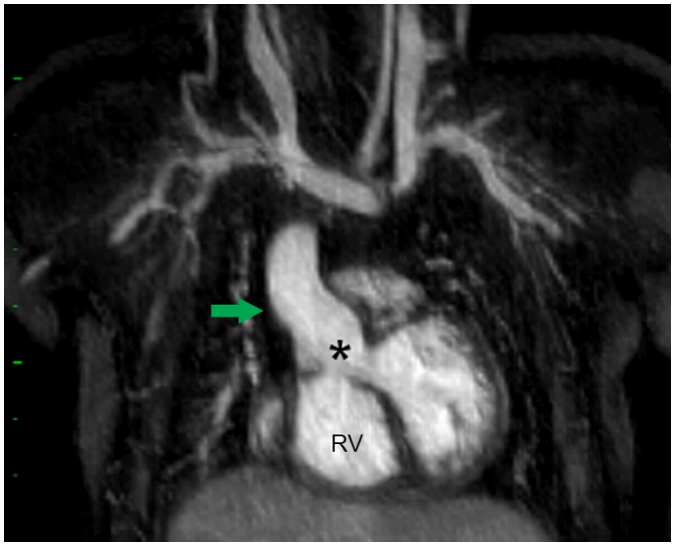
Situs inversus is a congenital anomaly in which organs are transposed laterally from their normal positions.[1,2] Most patients with situs inversus totalis, situs inversus with dextrocardia, do not present with congenital heart disease (CHD).[1,3] However, when situs inversus occurs with the heart positioned within the left hemithorax, isolated levocardia, CHD is consistently seen in nearly all patients.[1,3] Situs inversus with levocardia is relatively uncommon,[4] with an incidence of 1:22 000. It should be noted that the terms levocardia and dextrocardia refer to the position of the cardiac apex only and do not characterize any other part of the heart or other organs.[1]

Double-outlet right ventricle is a form of congenital heart disease where both of the great arteries connect (in whole or in part) to the right ventricle.[5] In our case, where there is pulmonary stenosis, the flow physiology is similar to that of Tetralogy of Fallot defined by four congenital cardiac defects: pulmonary stenosis, ventricular septal defect, right ventricular hypertrophy, and an overriding aorta, which allows blood to enter the aorta from both ventricles.[6]

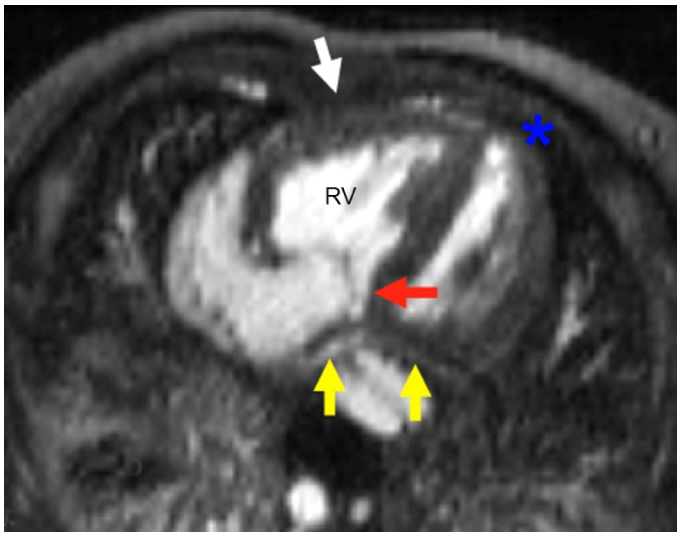
The case reported here is rare in that there is also a concomitant levo-transposition of the great arteries (L-TGA). A reported incidence of L-TGA is 1 in 3 500-5 000 births.[7] It manifests as atrioventricular and ventriculo-arterial discordance and accounts for 0.5% of all congenital heart defects.[8] When uncomplicated with situs inversus, L-TGA occurs because of an embryologic looping abnormality of the developing heart, causing the morphologic left ventricle to be

Figure 3. Contrast-enhanced Cardiac MRI in a Neonate with Situs Inversus, Isolated Levocardia, and Atrioventricular and Ventriculo-arterial Discordance

A Coronal view



B Axial view

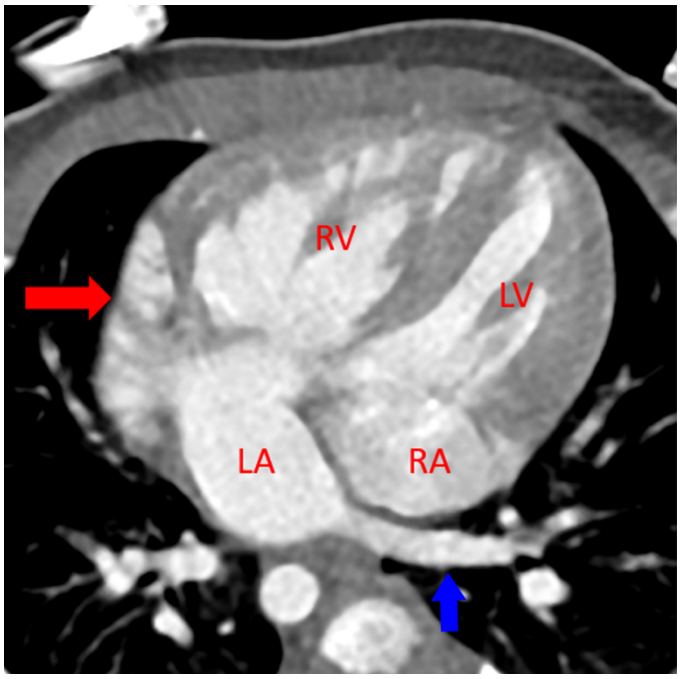


Coronal view (A) shows a membranous ventricular septal defect with the aorta (A, black asterisk) connecting to the morphologic right ventricle (RV) that remains on the right despite overall situs inversus. Axial view (B) shows levocardia (B, blue asterisk) with a narrowed pulmonary artery outflow component of the double-outlet right ventricle (B, red arrow). The pulmonic valve is imperceptible, likely atretic, along with diminutive pulmonary arteries (B, yellow arrows). Right ventricle hypertrophy (B, white arrow) is evident. There is a right sided-aortic arch (A, green arrow).

positioned to the right of the morphologic right ventricle.[9]

Patients with L-TGA have a predisposition toward the development of right ventricular hypertrophy and eventual heart failure,[8] as the less muscular morphologic right ventricle functions as a systemic ventricle, under chronic pressure and volume overload. In our patient, with underlying situs inversus and the morphologic right atrium on the left, the ventricles were positioned correctly, with the morphologic right ventricle remaining on the right. Nevertheless, the physiologic ramifications of having the right ventricle functioning as a systemic one remains. Patients with isolated L-TGA are likely not to experience the symptoms of heart disease throughout infancy and childhood,[8] but patients, such as the one described in this case report, with additional associated abnormalities of the heart, are likely to be seriously ill at birth and have a complicated presentation and course of their condition.

Figure 4. Contrast-enhanced Cardiac Computed Tomogram, Axial View, of Atrioventricular Discordance in a Neonate with Situs Inversus and Isolated Levocardia



The morphologic left atrium (LA) with a narrow, long appendage (red arrow) connects to the morphologic right ventricle (RV). The pulmonary veins (blue arrow) are seen draining into the morphologic left atrium (LA), which is seen on the right. The morphologic right atrium (RA) connects to the thick-walled morphologic left ventricle (LV).

Conclusion

The presentation of situs inversus with levocardia, double-outlet right ventricle, and atrioventricular discordance is an extremely rare clinical scenario. The patients can be exceptionally ill and, with the interplay between many different defects, diagnosis and surgical treatment can be challenging. Familiarity with the anatomic appearance of each of the defects along with an in-depth understanding of the functional consequences of these defects is critical for radiologists who hope to detect them in atypical combinations. Proper utilization of diagnostic imaging tools, such as fetal ultrasound, echocardiography, and cardiac MRI, are important for establishing diagnosis and planning of surgical treatment.

Author Contributions

Conceptualization, J.T.L. and B.L.Z.; Acquisition, analysis, interpretation of data, and writing – original draft preparation, B.L.Z.; Review and editing, J.T.L.; Supervision, J.T.L. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Disclosures

None to report.

References

1. Abdullah NL, Quek SC, Seto KY, Teo LL. Clinics in diagnostic imaging (160). Levocardia with abdominal situs inversus. *Singapore Med J.* 2015;56(4):198-202. DOI: 10.11622/smedj.2015059
2. Aaron Ong HJ, Toh SMY, Muhammad TJ, Norazrulrizal MN, Sia KK. Complex congenital heart disease: A rare case of situs inversus with isolated levocardia. *AHJ* 2019;26(4):4-6. Published 2019 Nov 21. https://doi.org/10.31762/AHJ1926.0402
3. Lin AE, Krikov S, Riehle-Colarusso T, et al. Laterality defects in the national birth defects prevention study (1998–2007): birth prevalence and descriptive epidemiology. *Am J Med Genet A.* 2014;164A(10):2581-2591. DOI: 10.1002/ajmg.a.36695
4. Vijayakumar V, Brandt T. Prolonged survival with isolated levocardia and situs inversus. *Cleve Clin J Med*. 1991;58(3):243–247. DOI: 10.3949/ccjm.58.3.243

5. Obler D, Juraszek AL, Smoot LB, Natowicz MR. Double outlet right ventricle: aetiologies and associations. *J Med Genet*. 2008;45(8):481–497. DOI: 10.1136/jmg.2008.057984
6. Apitz C, Webb GD, Redington AN. Tetralogy of Fallot. *Lancet*. 2009;374(9699);1462–1471. DOI: 10.1016/S0140-6736(09)60657-7
7. Martins P, Castela E. Transposition of the great arteries. *Orphanet J Rare Dis.* 2008;3:27 Published 2008 Oct 13. https://doi.org/10.1186/1750-1172-3-27
8. Hornung TS, Calder L. Congenitally corrected transposition of the great arteries. *Heart*. 2010;96(14):1154-1161. DOI: 10.1136/hrt.2008.150532
9. Lane DA. L-transposition of the great arteries: anatomy, physiology, clinical features, and diagnosis. In: Post TW, ed. *UpToDate.* UpToDate;2021. Accessed March 19, 2022. https://www.uptodate.com/contents/l-transposition-of-the-great-arteries-anatomy-physiology-clinical-features-and-diagnosis#H3660715995