

IMAGES in PAEDIATRIC CARDIOLOGY

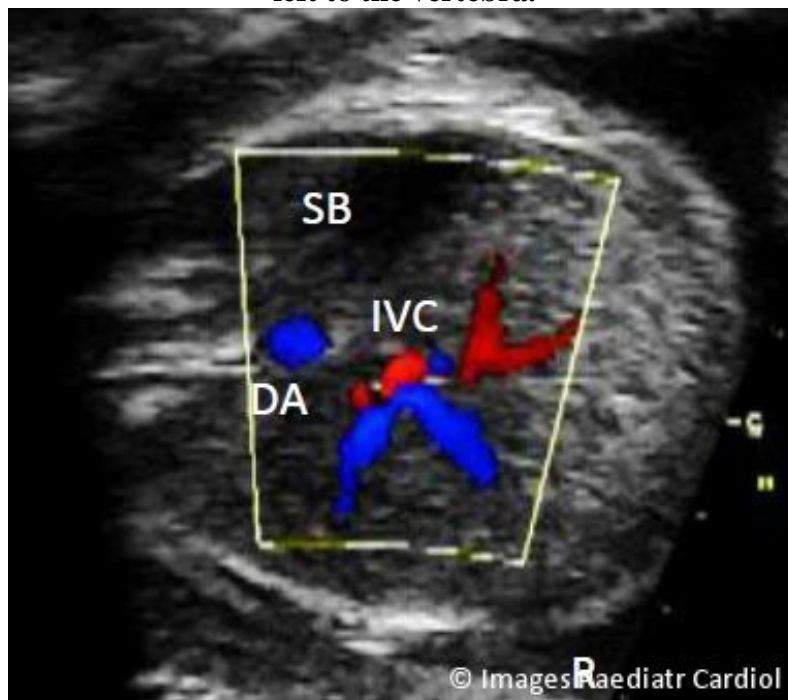
Kanagawa N, Inamura N, Narita J, Kawazu Y, Kayatani F. Prenatal diagnosis of isolated atrioventricular discordance using fetal echocardiography. *Images Paediatr Cardiol* 2014;16(1):5-10. Department of Pediatric Cardiology, Osaka Medical Center and Research Institute for Maternal and Child Health, Osaka, Japan

MeSH

Fetal echocardiography, isolated atrioventricular discordance, complete heart block

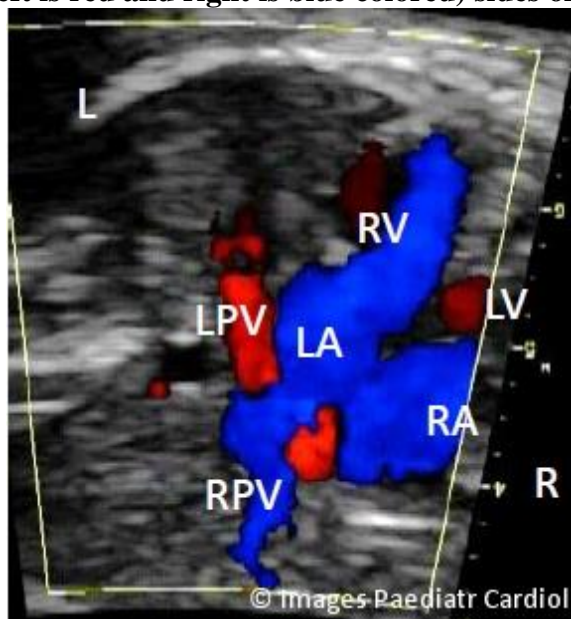
A 28-year-old female was referred to our institute at 25 weeks of gestation due to fetal bradycardia. Fetal echocardiography showed the stomach bubble was on the left side with the heart axis pointing to the left. The right sided inferior vena cava connected to the right atrium (RA) (Figure 1), and both pulmonary veins connected to the left atrium (LA) (Figure 2).

Figure 1: SB positioned left side. IVC (red color) drains to RA. DA (blue color) is positioned left to the vertebra.



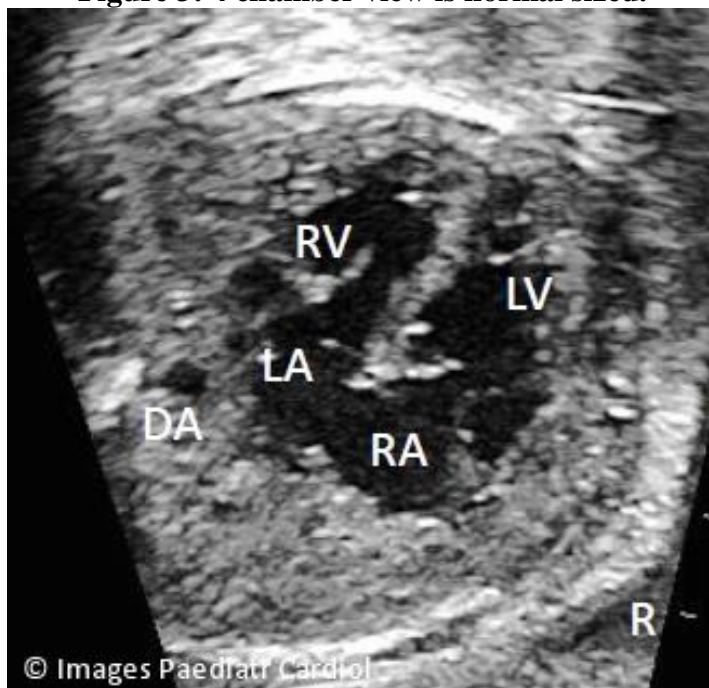
SB: stomach bubble, DA: descending aorta, IVC: inferior vena cava, R: right, L: left, RPV: right pulmonary vein, LPV: left pulmonary vein, RA: right atrium, LA: left atrium, RV: right ventricle, LV: left ventricle, AO: aorta, PA: pulmonary artery, SVC: superior vena cava, A: atrial systole, V: ventricular systole.

Figure 2: Both (left is red and right is blue colored) sides of PVs drain to LA.



The visceratrial situs was solitus. The four chamber view demonstrated that, each ventricle was normal size, and the left atrioventricular valve (morphologically tricuspid valve) inserted slightly more apically than the right sided valve (morphologically mitral valve). Furthermore, we diagnosed the left sided ventricle right ventricle (RV), because it had a moderator band (Figure 3). Thus, the atrioventricular alignment was discordant.

Figure 3: 4 chamber view is normal sized.



In addition, a great vessel arising from the left ventricle (LV) did not diverge, it was the aorta (Figure 4, 5).

Figure 4: AO arises from LV.

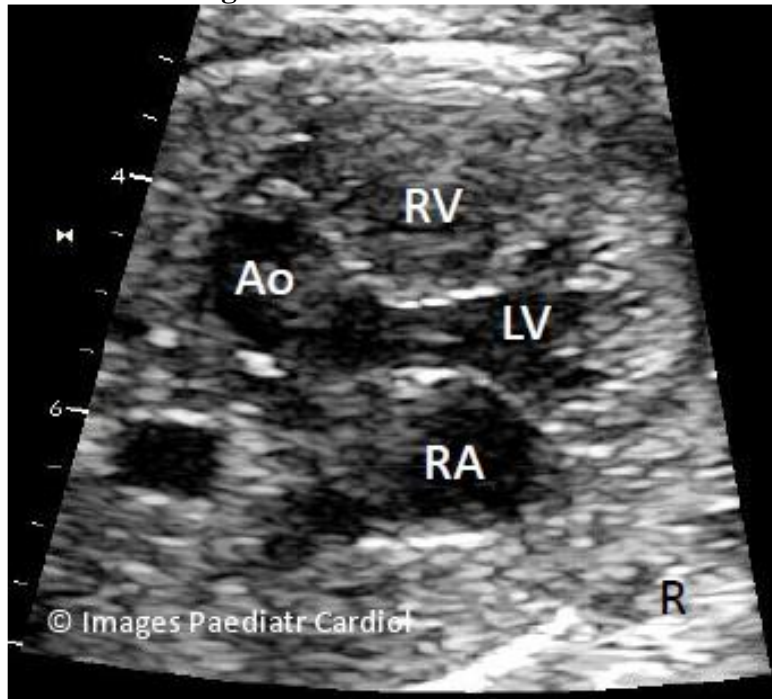
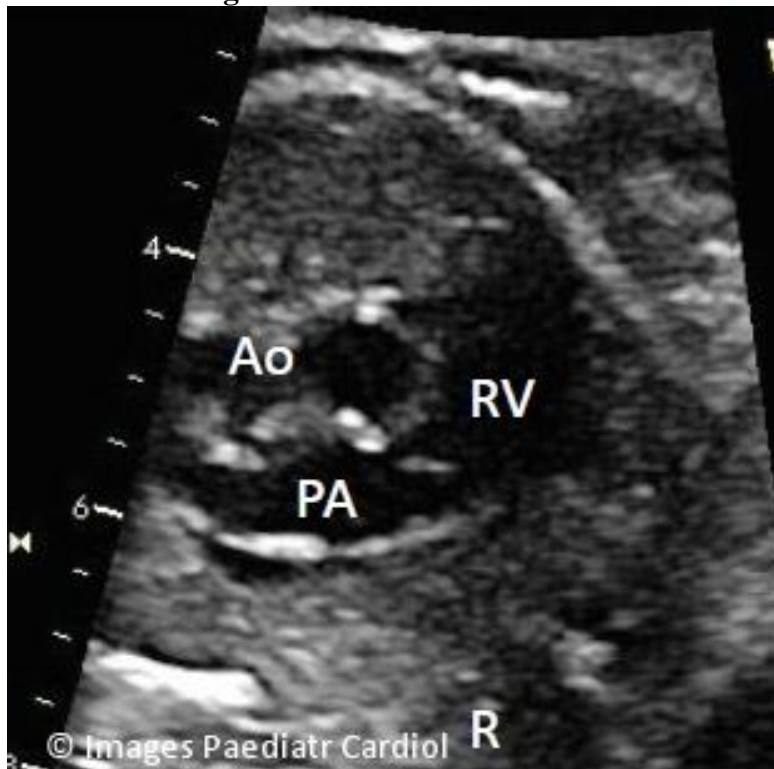


Figure 5: AO arises from LV.



In contrast, another great vessel arising from the RV diverged; this was the pulmonary artery (Figure 6, 7).

Figure 6: PA arises from RV

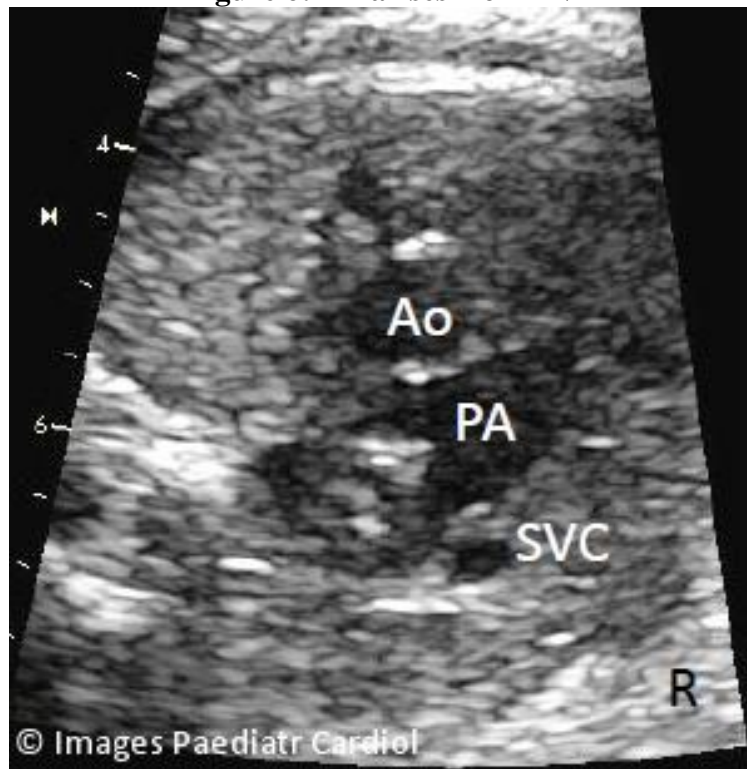
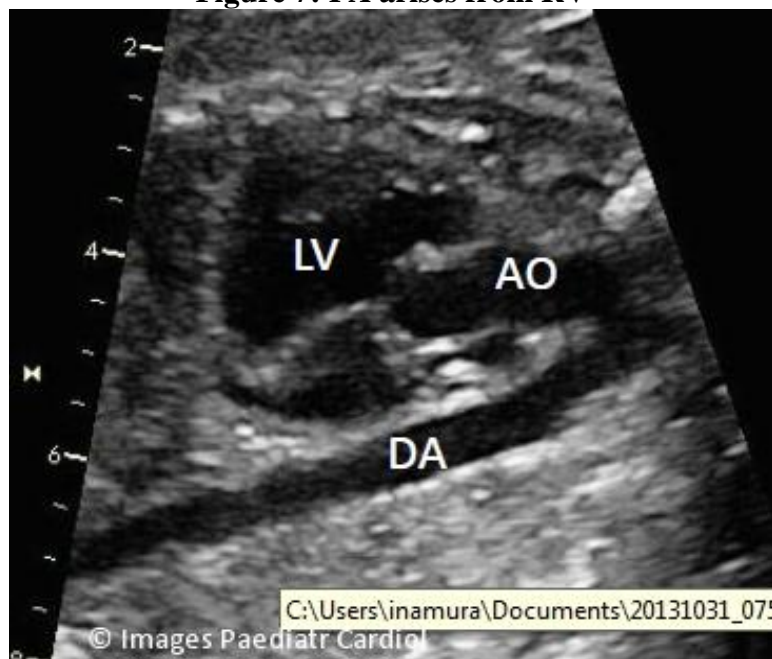


Figure 7: PA arises from RV

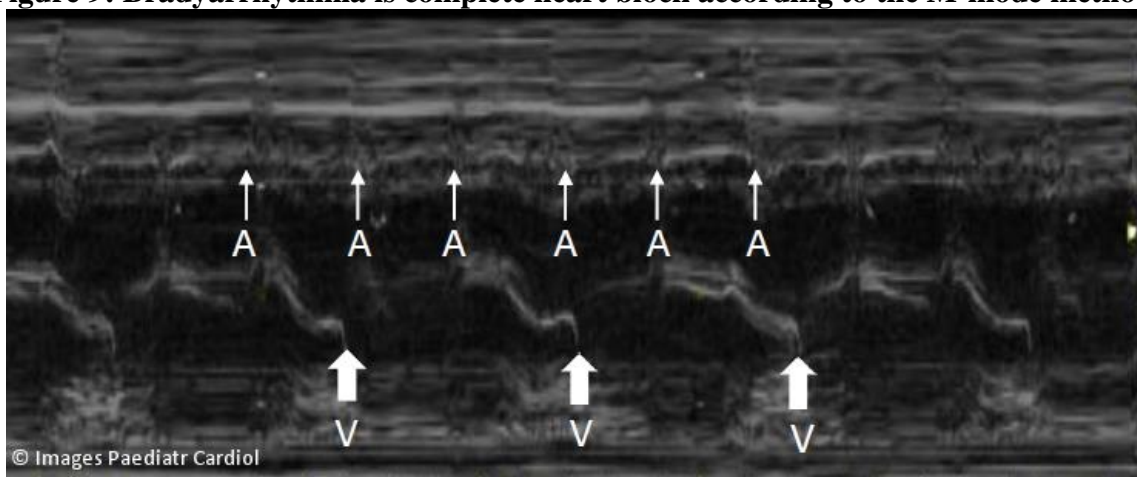


The ventriculoarterial alignment was concordant. A further evaluation using the three vessel view revealed a reversed placement of the great vessels (Figure 8). Bradyarrhythmia was diagnosed the complete heart block using the M mode examination (Figure 9).

Figure 8: In three vessels view, AO and PA are transposed.



Figure 9: Bradyarrhythmia is complete heart block according to the M-mode method.



Discussion

Isolated atrioventricular discordance is a rare cardiac malformation. The main complications of isolated atrioventricular discordance have been reported to be ventricular septal defect, aortic stenosis, coarctation of the aorta.¹⁻⁴ Complete heart block is a rare complication, but it is important as one of the postoperative complications. For such patients, the atrial switch operation provides a good prognosis because it maintains the left ventricle in the systemic circulation.⁵

There have so far been few reports describing the prenatal diagnosis of isolated atrioventricular discordance. The initial fetal echocardiography suggested this patient to have atrioventricular and ventriculoarterial discordance (corrected transposition of the great arteries: corrected TGA). In our center, when TGA is suspected, we make it a rule to find the 'I-shaped' sign in the three vessel and trachea view.⁶ However, in this case, we could not identify the 'I-shaped' sign. This situation led us to examine fetal echocardiogram more detail. We were therefore able to make a definitive diagnosis based on morphological examinations.

References

1. Hazan E, Baillot F, Rey C, Dupuis C. Isolated ventricular discordance and complete atrioventricular canal in situs inversus. Report of successful surgical repair. *Am J Cardiol.* 1977;40:463-6.
2. Konstantinov IE1, Lai L, Colan SD, Williams WG, Li J, Jonas RA, Van Praagh R. Atrioventricular discordance with ventriculoarterial concordance: A remaining indication for the atrial switch operation. *J Thorac Cardiovasc Surg.* 2004;128:944-5.
3. Sharma R, Marwah A, Shah S, Maheshwari S. Isolated atrioventricular discordance: surgical experience. *Ann Thorac Surg.* 2008;85:1403-6.
4. Shimpo H, Tani K, Hioki I, Miyake Y, Onoda K, Hayakawa H, Yada I. Isolated atrioventricular discordance with solitus viscera, inverted atria, D-loop ventricles, and solitus normally related great arteries: report of a rare case with successful surgical management. *J Thorac Cardiovasc Surg.* 1999;117:393-4.
5. Ostermeyer J, Bircks W, Krian A, Sievers G, Hilgenberg F. Isolated atrioventricular discordance. Report of two surgical cases with isolated ventricular inversion. *J Thorac Cardiovasc Surg.* 1983;86:926-9.
6. Ishii Y, Inamura N, Kawazu Y, Kayatani F, Arakawa H. 'I-shaped' sign in the upper mediastinum: a novel potential marker for antenatal diagnosis of d-transposition of the great arteries. *Ultrasound Obstet Gynecol.* 2013;41:667-71.

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