Criss-Cross Heart with Atrioventricular Discordance, Ventriculoarterial Concordance, and Hypoplastic Right Ventricle

Monther Obeidat[1], Rania Sanouri[1], Ahmad Othman[2*] and Ayat Alwan[3]

[1]Department of Pediatrics, Queen Rania Hospital for Children, Amman, Jordan

[2]Faculty of Medicine, Yarmouk University, Irbid, Jordan

[3]Department of Radiology, Queen Rania Hospital for Children, Amman, Jordan

Received: 12 February 2026

Accepted: 5 August 2026

*Corresponding Author: hmadabuhalawah@gmail.com

DOI 10.5001/omj.2030.17

Abstract

We report a rare and complex case of criss-cross heart with atrioventricular discordance and ventriculoarterial concordance in a 28-day-old neonate who presented with profound cyanosis. Multimodality imaging, including transthoracic echocardiography, computed tomography angiography, and cardiac catheterization, demonstrated criss-cross atrioventricular connections, a hypoplastic right ventricle, moderate subvalvular pulmonary stenosis, a large secundum atrial septal defect, and a small ventricular septal defect. The resulting anatomy produced a transposition-like physiology with markedly reduced pulmonary blood flow. The patient underwent a modified Blalock–Taussig shunt with initial improvement in oxygen saturation but subsequently developed multisystem organ failure and died six days postoperatively. This case highlights an exceptionally rare anatomical combination, underscores the importance of early recognition using multimodality imaging, and illustrates the challenges of surgical decision-making in neonates with complex cyanotic congenital heart disease.

Keywords: Criss-cross heart; atrioventricular discordance; ventriculoarterial concordance; congenital heart disease

Introduction

Congenital heart disease (CHD) is the most common congenital anomaly in newborns and is defined as a structurally significant abnormality of the heart or intrathoracic great vessels that impairs normal cardiac function.[1,2] Criss-cross heart (CCH) is an exceptionally rare CHD, accounting for <0.1% of congenital cardiac malformations, and is characterized by crossing of the ventricular inflow streams due to abnormal rotation of the ventricular mass along its long axis.[3]

CCH is frequently associated with other cardiac anomalies, particularly ventricular septal defects (VSDs) and ventricular outflow tract abnormalities, including transposition of the great arteries, double-outlet right ventricle, and pulmonary stenosis.[3] Surgical management ranges from palliative procedures to definitive anatomical correction depending on the associated defects.[4]

Case Report

A 28-day-old male infant was referred for evaluation of complex congenital heart disease with cyanosis present since birth. He was born at 35 weeks' gestation to a 42-year-old gravida 11, para 9 mother by urgent cesarean section for non-reassuring cardiotocography in the setting of poorly controlled insulin-treated type 2 diabetes mellitus. The mother had obesity (pre-pregnancy BMI 33.1 kg/m^2), persistent hyperglycemia (250–300 mg/dL), and severe polyhydramnios. She denied smoking, alcohol, illicit drug use, teratogen exposure, congenital heart disease, autoimmune disease, or significant infections during pregnancy. Pregnancy medications included insulin and folic acid. Previous maternal cardiac evaluations showed normal sinus rhythm and trace mitral regurgitation.

The parents were consanguineous, although there was no family history of congenital heart disease, genetic syndromes, or inherited disorders. Routine antenatal care was received with monthly follow-up; despite persistent maternal hyperglycemia, no fetal structural abnormalities were detected, and no invasive prenatal genetic testing was performed.

The infant was delivered in cephalic presentation without difficulty, weighed 3.4 kg (large for gestational age), had Apgar scores of 8 and 8 at 1 and 5 minutes, respectively, and required no resuscitation. Cyanosis shortly after birth prompted neonatal intensive care unit admission. Initial laboratory investigations and chest radiography were unremarkable. Continuous cardiorespiratory monitoring, supplemental oxygen, broad-spectrum antibiotics, and supportive care were initiated pending surgical intervention.

On admission, the infant had central cyanosis without signs of congestive heart failure or dysmorphic features. Cardiovascular examination revealed normal first and second heart sounds with a faint systolic murmur. Oxygen saturation was 55% on room air. Prostaglandin E1 infusion, supplemental oxygen, and broad-spectrum antibiotics were started.

Transthoracic echocardiography, cardiac computed tomography angiography, and cardiac catheterization demonstrated situs solitus with levocardia and criss-cross heart with atrioventricular (AV) discordance and ventriculoarterial (VA) concordance. The AV alignments were discordant (**Figures 1 and 2**), while the ventriculoarterial connections were concordant. The left atrium drained into a hypoplastic right ventricle through an elongated AV tunnel (**Figures 1 and 2**). The right ventricle was positioned anteriorly and to the right of the left ventricle. The aorta arose from the left ventricle and was mildly dilated, whereas the pulmonary artery originated from the right ventricle (**Figure 3**).

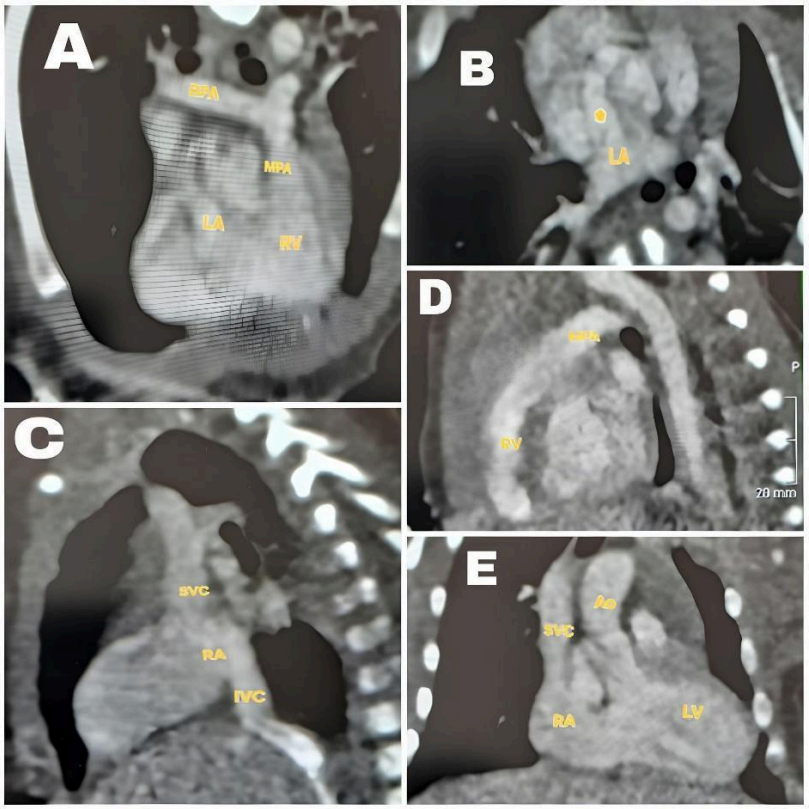

Figure 1: CTA images demonstrating atrioventricular and ventriculoarterial anatomy. (a) oblique axial view. LA drains into RV which gives rise to MPA. (b) oblique axial view. pulmonary veins draining into small LA that continue as a tunnel-like structure (star) to a hypoplastic RV. (c) sagittal view. SVC and IVC draining into RA. (d) sagittal view. Small anterior RV gives rise to MPA. (e) coronal view. SVC draining into RA, which empties into LV that gives rise to PA. Aorta arising from LV.

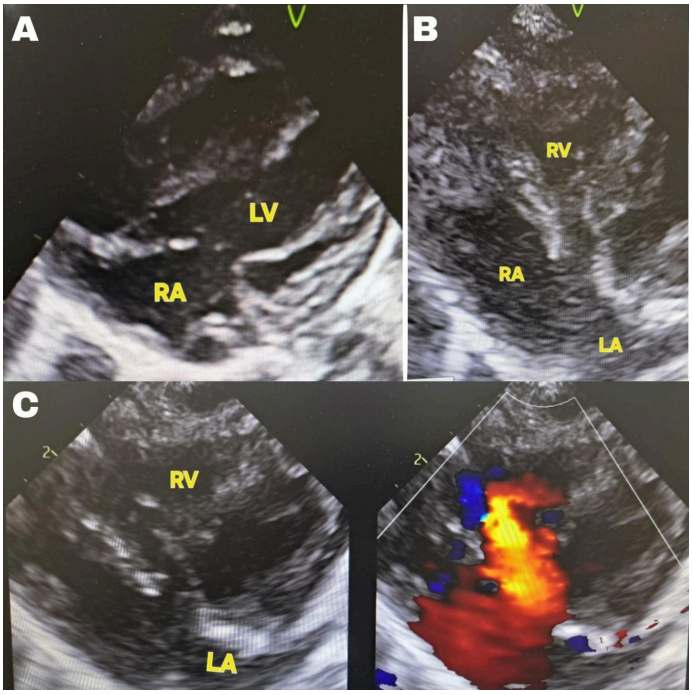

Figure 2: 2D images of the same case. (**a**) A small LA drains through a tunnel-like structure into a small RV with a large ASD. **(b)** with colour doppler. **(c)** RA empties in the left ventricle.

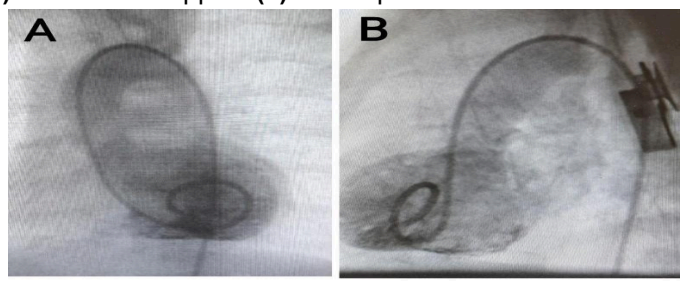

Figure 3: LV angiographic images showing smooth surface, posterior ventricle (LV) emptying into the aorta in AP (**a**) and lateral (**b**) views.

Because of critically reduced pulmonary blood flow, a 3.5-mm modified Blalock-Taussig shunt was placed, resulting in immediate improvement in oxygen saturation. The postoperative course was complicated by acute kidney injury, lactic acidosis, coagulopathy, and peripheral hypoperfusion requiring high-dose inotropic support and peritoneal dialysis. Despite maximal intensive care, the patient died six days after surgery.

Discussion

Criss-cross heart results from abnormal rotation of the ventricular mass during cardiac development while the atrial segment remains relatively fixed.[5] Anderson et al. proposed that clockwise or counterclockwise post-septational rotation produces this anomaly; clockwise rotation usually results in AV concordance, whereas AV discordance, as in the present case, is less common.[6] Additional apical tilting may produce a superior-inferior ventricular arrangement.[7]

A systematic review by Ramadan et al. identified 101 patients reported between 1974 and 2021. AV concordance was present in 75.2% of cases, whereas AV discordance was less frequent.[8] Cyanosis was the most common presenting feature, followed by dyspnea and tachycardia. VSD was the most common associated anomaly, followed by pulmonary stenosis, double-outlet right ventricle, and transposition of the great arteries.[8]

Transthoracic echocardiography is the primary diagnostic modality and should raise suspicion when a standard four-chamber view cannot be obtained or crossing AV inflow tracts are visualized.[9,10] Cardiac catheterization provides hemodynamic assessment and identifies associated defects, while cardiac magnetic resonance imaging offers detailed three-dimensional anatomical reconstruction.[11] Surgical treatment targets associated hemodynamically significant lesions rather than ventricular rotation itself.[4] Without intervention, prognosis is poor, with approximately 64% of deaths occurring during childhood and nearly half during the neonatal period.[4]

In this patient, the complex cyanotic anatomy, hypoplastic right ventricle, and pulmonary outflow obstruction necessitated palliative modified Blalock-Taussig shunt placement, a PTFE graft connecting a systemic artery to a pulmonary artery to improve pulmonary blood flow and reduce cyanosis.[12] Postoperative acute kidney injury is a common complication after neonatal cardiac surgery, affecting 30%–60% of patients, and is associated with increased morbidity and mortality due to immature renal physiology, low cardiac output, contrast exposure, and perioperative hemodynamic instability.[13,14]

Although the embryological mechanism of CCH remains uncertain, optimization of maternal health may reduce the overall risk of CHD.[15] In the present case, the patient underwent a modified Blalock–Taussig shunt at Queen Alia Military Hospital for Children, a national tertiary pediatric cardiac referral center with dedicated pediatric cardiac surgery and intensive care services. Despite initial improvement, fatal postoperative complications reflected the severity of the underlying cardiac malformation rather than limitations in surgical expertise. Vertical HIV transmission has been associated with cardiomyopathy, ventricular dilatation, myocardial wall thickening, and myocarditis,[16] but no association with CCH has been reported. Both the mother and infant were HIV-negative, excluding perinatal HIV exposure as a contributing factor.

This case is notable for the exceptionally rare combination of CCH with AV discordance and VA concordance producing transposition physiology. Sheikh et al. reported four cases of CCH, only one of which had the same morphology but with associated coarctation,[10] Only a few comparable cases have been described.[17-19]

Conclusions

Criss-cross heart with atrioventricular discordance and ventriculoarterial concordance is an exceptionally rare congenital anomaly associated with substantial neonatal mortality despite advances in surgical and perioperative care. Early recognition using multimodality imaging is essential for accurate diagnosis and appropriate surgical planning.

Disclosure

Father’s consent was obtained to report this case.

References

1. Sun R, Liu M, Lu L, Zheng Y, Zhang P. Congenital Heart Disease: Causes, Diagnosis, Symptoms, and Treatments. Cell Biochem Biophys 2015 Jul;72(3):857-860. .

2. van der Bom T, Zomer AC, Zwinderman AH, Meijboom FJ, Bouma BJ, Mulder BJ. The changing epidemiology of congenital heart disease. Nat Rev Cardiol 2011 Jan;8(1):50-60. .

3. Yang Q, Zhang Y, Zhao Y, Liu X, Sun L, Gu X, et al. Prenatal diagnosis of criss-cross heart - case series and review of the literature. J Matern Fetal Neonatal Med 2024 Dec;37(1):2362333. .

4. Oliveira ÍM, Aiello VD, Mindêllo MM, Martins YdeO, Pinto VC Jr. Criss-cross heart: report of two cases, anatomic and surgical description and literature review. Rev Bras Cir Cardiovasc 2013 Mar;28(1):93-102. .

5. Rahajoe AU, Roebiono PS, Fahmi MB, Harimurti GM. CRISS - CROSS HEART (Report of four cases). Indonesian Journal of Cardiology. 2012;33(1):34-40. doi:10.30701/IJC.V33I5.74.

6. Anderson RH, Baker EJ, Penny DJ, Redington AN, Rigby ML, Wernovsky G. Paediatric cardiology. Paediatric Cardiology. 2009 Sep 18;1–1327.

7. Salih M, Dzhelepova N, Elnoury R, Eltayeb Y. An Intriguing Case of a Twisted and Tilted Heart. SVOA Paediatrics. 2025;(3):61-69. .

8. Ramadan A, Roshdy MR, Hamad AA, Refaey N, Mclaren BA, Care H, et al. Epidemiological, Clinical and imaging features, and outcome of patients with Crisscross heart disease: A Systematic review of 46 published case reports [Internet]. 2023 Oct 12.

9. Yang YL, Wang XF, Cheng TO, Xie MX, Lü Q, He L, et al. Echocardiographic characteristics of the criss-cross heart. Int J Cardiol 2010 Apr;140(2):133-137. .

10. Sheikh AM, Kanwal A, Sattar H. Criss cross heart: A challenging diagnosis. J Pak Med Assoc 2022 Jul;72(7):1429-1431. .

11. Ming Z, Yumin Z. Magnetic Resonance Evaluation of Criss-Cross Heart. Pediatric Cardiology 2007 29:2. 2007 Oct 5;29(2):359–65. PubMed PMID: 17917768.

12. Alahmadi MH, Sharma S, Bishop MA. Modified Blalock-Taussig-Thomas Shunt. StatPearls. 2024 Oct 22. PubMed PMID: 37983362.

13. Khuong JN, Wilson TG, Iyengar AJ, d’Udekem Y. Acute and Chronic Kidney Disease Following Congenital Heart Surgery: A Review. Ann Thorac Surg 2021 Nov;112(5):1698-1706. .

14. Chesney RW, Kaplan BS, Freedom RM, Haller JA, Drummond KN. Acute renal failure: an important complication of cardiac surgery in infants. J Pediatr 1975 Sep;87(3):381-388. .

15. Liu S, Joseph KS, Lisonkova S, Rouleau J, Van den Hof M, Sauve R, et al; Canadian Perinatal Surveillance System (Public Health Agency of Canada). Association between maternal chronic conditions and congenital heart defects: a population-based cohort study. Circulation 2013 Aug;128(6):583-589. .

16. Arriola-Montenegro L, Sanchez MV, Chen G, Brown C, Rasmussen M, Salter C, et al. Cardiovascular disease associated with congenital infections. Prog Pediatr Cardiol 2025 Mar;76:101780. .

17. Attie F, Muñoz-Castellanos L, Ovseyevitz J, Flores-Delgado I, Testelli MR, Buendia A, et al. Crossed atrioventricular connections. Am Heart J 1980 Feb;99(2):163-172. .

18. Asokan DrA. G DrA, Kanakaraju DrK, Rajenthrakumar DrH. Criss Cross Heart with Normally Related Great Arteries. Scholars Journal of Medical Case Reports. 2021 Nov;9(11):1090-1092. .

19. Geva T, Sanders SP, Ayres NA, O’Laughlin MP, Parness IA. Two-dimensional echocardiographic anatomy of atrioventricular alignment discordance with situs concordance. Am Heart J 1993 Feb;125(2 Pt 1):459-464. .