ESC European Society of Cardiology

European Heart Journal - Case Reports (2025) **9**, ytaf223
https://doi.org/10.1093/ehjcr/ytaf223

CASE REPORT
Cardiovascular imaging

Multimodality cardiac imaging for pre-operative evaluation in a paediatric criss-cross heart patient: a case report

Kwannapas Saengsin [1]*, Surin Woragidpoonpol[2], Tanop Srisuwan [3], Rekwan Sittiwangkul[1], and Krit Makonkawkeyoon[1]

[1]Division of Cardiology, Department of Pediatrics, Faculty of Medicine, Chiang Mai University, 110 Inthawarorot Road, Sri Phum Subdistrict, Mueang Chiang Mai District, Chiang Mai 50200, Thailand; [2]Division of Cardiovascular Thoracic Surgery, Department of Surgery, Faculty of Medicine, Chiang Mai University, 110 Inthawarorot Road, Sri Phum Subdistrict, Mueang Chiang Mai District, Chiang Mai 50200, Thailand; and [3]Department of Radiology, Faculty of Medicine, Chiang Mai University, 110 Inthawarorot Road, Sri Phum Subdistrict, Mueang Chiang Mai District, Chiang Mai 50200, Thailand

Received 8 September 2024; revised 23 December 2024; accepted 1 May 2025; online publish-ahead-of-print 15 May 2025

Background Criss-cross heart (CCH) is considered as one of the most complex, challenging, and exceptionally rare congenital heart diseases (CHDs). Criss-cross heart is defined by the twisted atrioventricular connection and the crossing of inflow streams to both ventricles, caused by the rotation of the ventricular mass along its longitudinal axis. The treatment depends on the anatomical complexity. Diagnosis of CCH is difficult, and interpretation of imaging results may be challenging.

Case summary A one-year-old girl presented with cyanosis and abnormal heart sounds. We report a patient with complex congenital CCH who underwent successful biventricular repair using multimodality cardiac imaging, including echocardiography, cardiac computerized tomography, 3D printing, and 4D flow-CMR (cardiac magnetic resonance).

Discussion Criss-cross heart is one of the most complex and rare CHD. Multimodality cardiac imaging is essential for finding the answers and providing better care for complex CHD patients.

Keywords Congenital heart disease • Criss-cross heart • Case report • Cardiac magnetic resonance • Four-dimensional cardiovascular magnetic resonance flow imaging (4D flow-CMR) • 3D printing in congenital heart disease • Advanced imaging techniques

ESC curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 2.4 Cardiac computed tomography

Learning points

- Criss-cross heart is an exceptionally rare cardiac anomaly. It is commonly associated with diverse cardiac defects.
- The surgical approach depends on the morphology of the primary defect.
- Multimodality cardiac imaging greatly contributed to understanding the cardiac anatomy and haemodynamics for pre-operative evaluation, resulting in improved patient management.

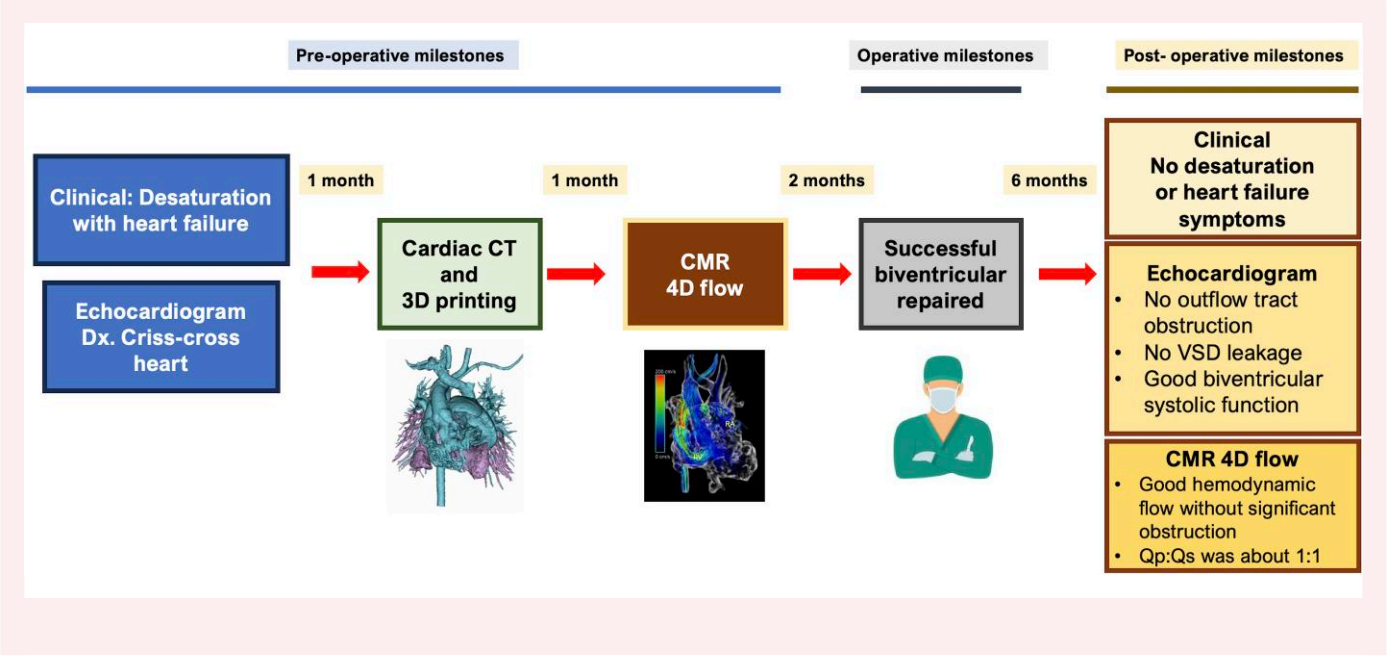
* Corresponding author. Tel: +(66)95-7813706, Email: kwannapas_09@hotmail.com
Handling Editor: Christoph Jensen
Peer-reviewers: Sharipah Intan Syed Abas; Andi Rroku
Compliance Editor: Sheetal Vasundara Mathai

Introduction

Criss-cross heart (CCH) is recognized as one of the most intricate and extraordinarily rare congenital heart diseases (CHDs). It is estimated to occur in about eight cases per 1 000 000 live births, accounting for <0.1% of all CHD instances.[1] The characteristic that defines CCH is the twisted atrioventricular connection and crossing of inflow streams to both ventricles, resulting from the rotation of the ventricular mass along its longitudinal axis.[2] It can appear with various atrioventricular and ventriculo-arterial arrangements and is commonly associated with a superior–inferior orientation of the ventricular mass.[3,4] The treatment depends on the anatomical complexity. In most cases, CCH patients are not candidates for a two-ventricle repair and are directed towards a Fontan-type surgery.[3,5] Diagnosis of CCH is difficult, and interpretation of imaging results may be challenging. Using multimodality cardiac imaging, we report a complex congenital CCH patient who underwent successful biventricular repair.

The institutional review board of the faculty of medicine approved this study.

Summary figure



Case presentation

A one-year-old girl presented with cyanosis and abnormal heart sounds. She developed clinical heart failure, dyspnoea, and failure to thrive. Cyanosis with room air oxygen saturation of 85% was noted on examination. On auscultation, there was a loud systolic ejection murmur grade III/VI at the left upper parasternal broader. Chest X-ray showed levocardia and cardiomegaly (cardiothoracic ratio of 60%) with mildly increased pulmonary blood flow (*Figure 1*). Two-dimensional (2D) echocardiography showed {I,L,L} abdominal situs inversus, atrial situs inversus with superior vena cava and inferior vena cava draining to the left-sided morphologic right atrium, and crossing of inflow streams of the right-sided right ventricles due to an apparent twisting of the heart (criss-cross heart physiology). The imaging showed L-loop ventricle in which the atrioventricular connections are concordant and the topology of the right ventricle is of the 'left hand' type. Furthermore, echocardiography showed a double outlet right ventricle (DORV) with L-malposition of great arteries, conoventricular ventricular septal defect (VSD), infundibular pulmonary stenosis (PS), right aortic arch, and good biventricular systolic function (*Figure 2*). Cardiac computerized tomography (CCT) for preoperative planning showed cardiac segment anatomy and confirmed diagnosis with CCH. Additional information from the CCT showed a superior–inferior ventricle and right-sided aortic arch with an aberrant left subclavian artery (*Figure 3*). Cardiac 3D printing was performed for pre-operative planning (*Figure 4*). Four-dimensional cardiovascular magnetic resonance flow imaging (4D flow-CMR) without contrast definitively demonstrated the crossing of inflow streams of the two ventricles and accelerated flow from below the pulmonic valve. Estimated pulmonary blood flow (Qp) to systemic blood flow (Qs) was 2.46:1 under FiO_2 100% (*Figure 5*). The scan time was about 15 min. The patient underwent successful biventricular repair and was discharged after 10 days. An echocardiogram before discharge showed good left ventricular function and unobstructed right and left ventricular outflow tracts. After total correction, 4D flow-CMR showed good haemodynamic flow without significant obstruction and the Qp:Qs was about 1.

Four-dimensional cardiovascular magnetic resonance flow imaging protocols

All MRIs were taken using a 1.5 T clinical MRI system (Signa HDxt 1.5T; GE Healthcare, Milwaukee, WI, USA.) utilizing a clinical 12-Channel HD Body Array Coil. 4D flow (four-dimensional flow) uses 3D phase contrast (PC-MRI) with time-resolved acquisition. Sequences typically include gradient-echo or steady-state free precession. A bipolar gradient is applied to encode velocity information based on phase shifts in the MRI signal, and velocity encoding gradients are used in three orthogonal directions (x, y, z) to capture flow in all directions. The VENC (velocity encoding) is set according to the expected flow speeds in the region of interest. The 'fourth dimension' refers to the temporal aspect, where a series of images is acquired over time, typically during a single cardiac cycle. The total scan time was 5–6 min depending on cardiac triggering rate. PC velocity encoding was 200 cm/s, and was applied along RL, AP, and FH flow directions.

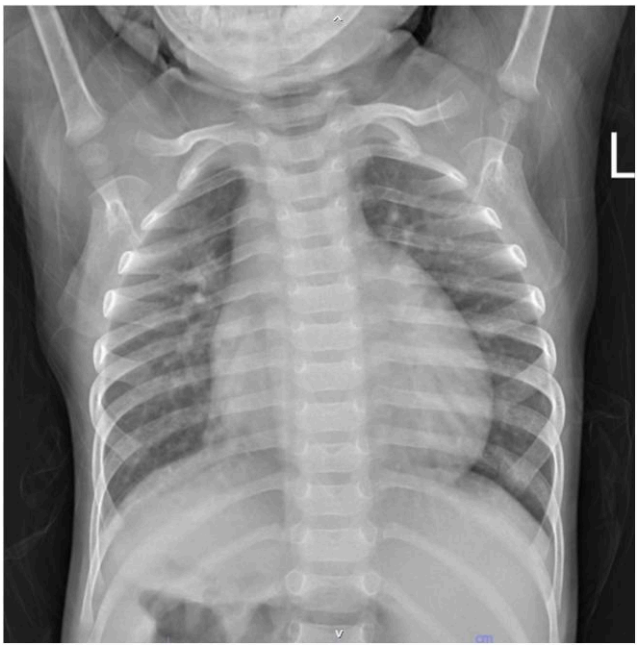


Figure 1 Chest X-ray AP supine demonstrates levocardia and cardiomegaly with increased pulmonary blood flow.

Discussion

Criss-cross heart is an exceptionally rare CHD characterized by the crossing of ventricular entry axes, which are typically parallel in a normal heart.[6] In CCH, there is a displacement of the ventricular mass, resulting in each ventricle seeming to be located on the opposite side of its corresponding atrium. Although the base of the heart stays in place, the ventricles appear to have twisted along their longitudinal axis. This shift in blood flow dynamics results in the crossing of flows through the atrioventricular (AV) valves, creating the illusion that each atrium is delivering blood to the opposite ventricle.[7]

The presentation of CCH can include solitus or situs inversus and concordant or discordant

AV or ventriculo-arterial (VA) connections.[2] Most cases that present with situs solitus are with atrial situs solitus.[8] In our case, the patient presented with situs solitus but with atrial situs inversus. The VA connection should be described separately and may be concordant or discordant.[9,10] The CCH is always invariably associated with other cardiac defects such as VSD, atrial septal defect, PS, DORV, or transposition of the great arteries.[8] Our case showed L-loop ventricle, DORV with L-malposition of great arteries, conoventricular VSD, infundibular PS, and right aortic arch. Due to the complexity of cardiac anatomy, most CCH patients are directed towards a Fontan-type surgery.[3,5] We report a successful biventricular repair of a complex congenital CCH, achieved with using multimodality cardiac imaging.

The 2D-transthoracic echocardiography is the primary imaging method used to assess the configuration and structural arrangement of all cardiac chambers and AV valves, as well as their connections to the great arteries.[11,8]

Cardiac computerized tomography is a crucial tool for diagnosis of complex CHDs. Advances in CCT technology now allow for the elimination of motion artefacts without the need for anaesthesia and a substantial reduction in radiation exposure.[12] Using advanced multiplanar imaging combined with 3D volume reconstruction, we can effectively visualize and clarify anatomical structures, making their complexities easier to understand and accurately depicting the size of both ventricles. Furthermore, in this case, the CCT imaging added important details, such as the presence of a superior–inferior ventricle and a right-sided aortic arch with an aberrant left subclavian artery.

The use of 3D-printing models has enhanced our understanding of complex cardiac anatomy. The models have demonstrated their usefulness by helping surgical teams prepare more effectively for correcting major abnormalities, including the closure of ventricular septal defects. By clarifying three-dimensional anatomy, 3D-printed models improve decision-making, planning, and the safety of complex congenital heart surgeries.[13] Alifu A and colleagues reported the effectiveness of using a 3D-printed model to replicate the detailed anatomy of a CCH with situs solitus and DORV in a 13-year-old female patient. Using the 3D-printed model enhanced the comprehension of the patient's unique anatomy and enabled more accurate surgical planning, resulting in a successful biventricular repair.[14]

In assessing challenging anatomical abnormalities such as CCH, 4D flow-CMR proves invaluable. The 4D flow-CMR showed the flow steaming of the two ventricles to cross each other due to the heart

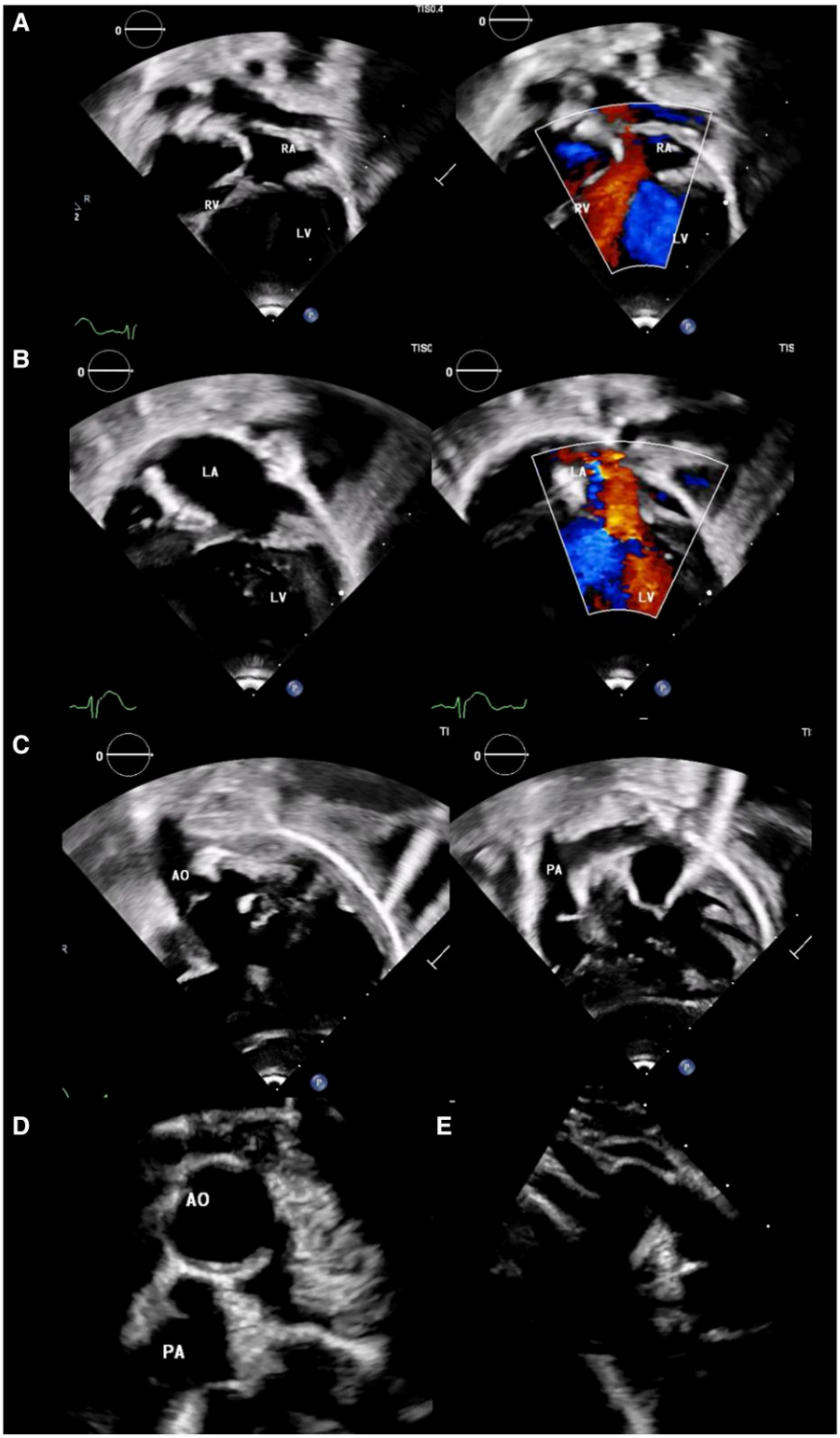


Figure 2 Echocardiography images demonstrate. (*A*) Left-sided right atrium connected to right-sided right ventricle; (*B*) right-sided left atrium connected to left-sided left ventricle; (*C*) double outlet right ventricle with two great arteries arising from right ventricle; (*D*) malposition of great artery with aorta left anterior to pulmonary artery; (*E*) right-sided aortic arch.

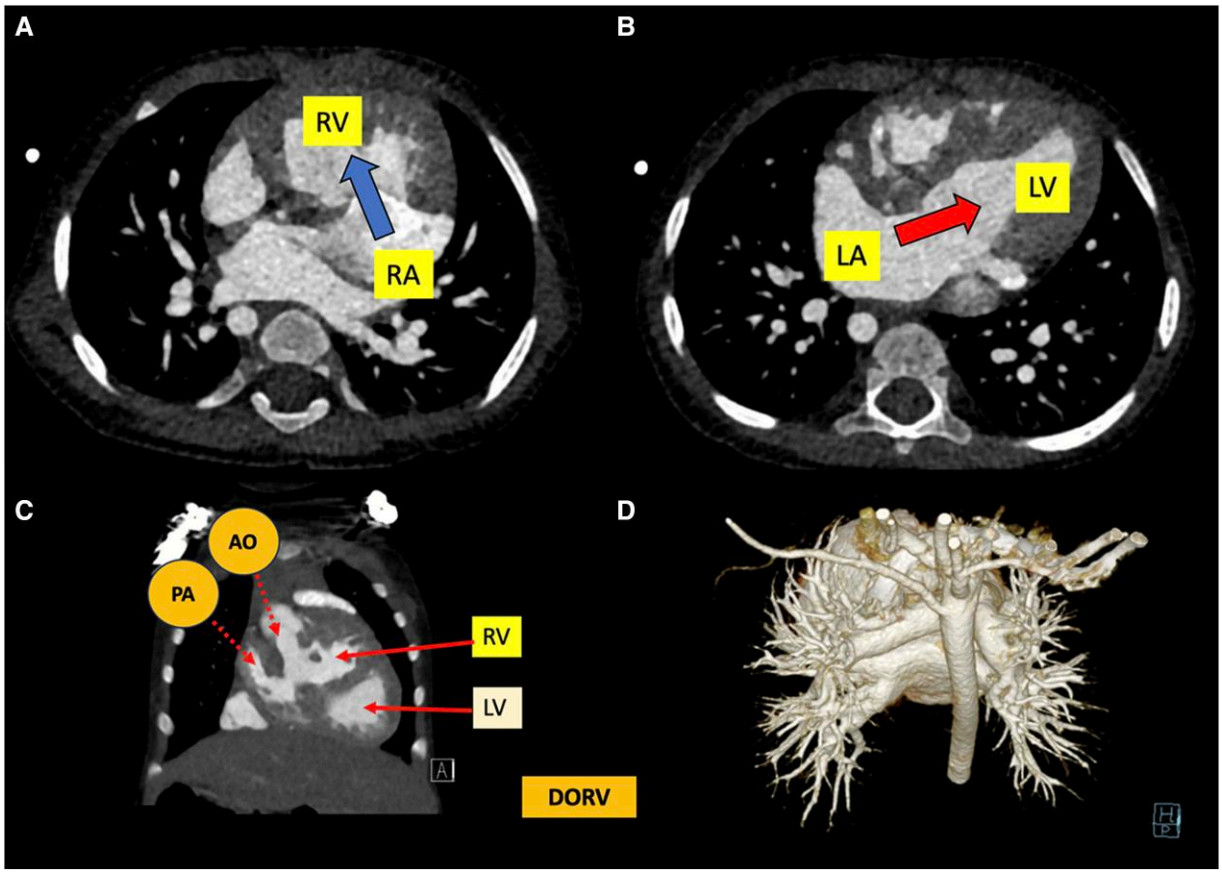


Figure 3 Cardiac computerized tomography demonstrated. (*A*) Axial image showed left-sided right atrium connected to right-sided right ventricle; (*B*) the axial image showed right-sided left atrium connected to left-sided left ventricle; (*C*) coronal image showed superior–inferior relationship of the two ventricles and double outlet right ventricle; (*D*) volume-rendered 3D reconstruction showed right-sided aortic arch with an aberrant left subclavian artery.

twisting around its long axis. As a comprehensive tool for flow analysis, 4D flow-CMR enables accurate visualization of blood flow patterns, along with the study of complex haemodynamic parameters.[15] Additionally, 4D flow-CMR provides significant value in analysing the haemodynamics of intracardiac shunts. It offers a reliable approach for measuring the Qp/Qs ratio and directly quantifying shunts, demonstrating high reproducibility.[16] To the best of our knowledge, this case report may be the first to utilize 4D flow-CMR to enhance visualization and evaluate haemodynamics pre- and post-operatively in a paediatric patient with CCH.

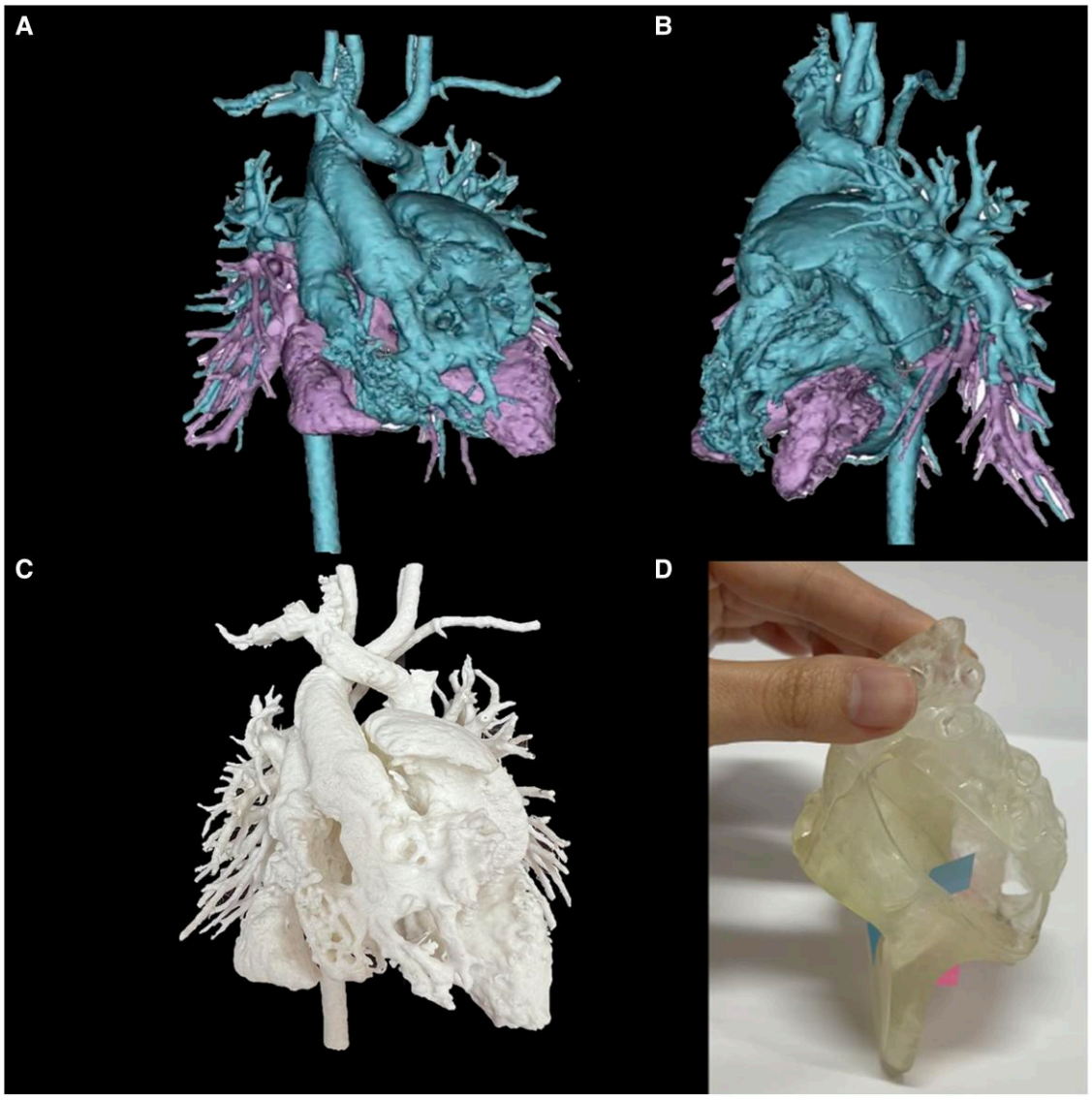


Figure 4 (*A*) Volume-rendered 3D reconstruction showed criss-cross heart with double outlet right ventricle; (*B*) criss-cross heart with double outlet right ventricle with superior–inferior ventricle; (*C*) 3D printing with blood pool reconstruction; (*D*) 3D printing with myocardial reconstruction.

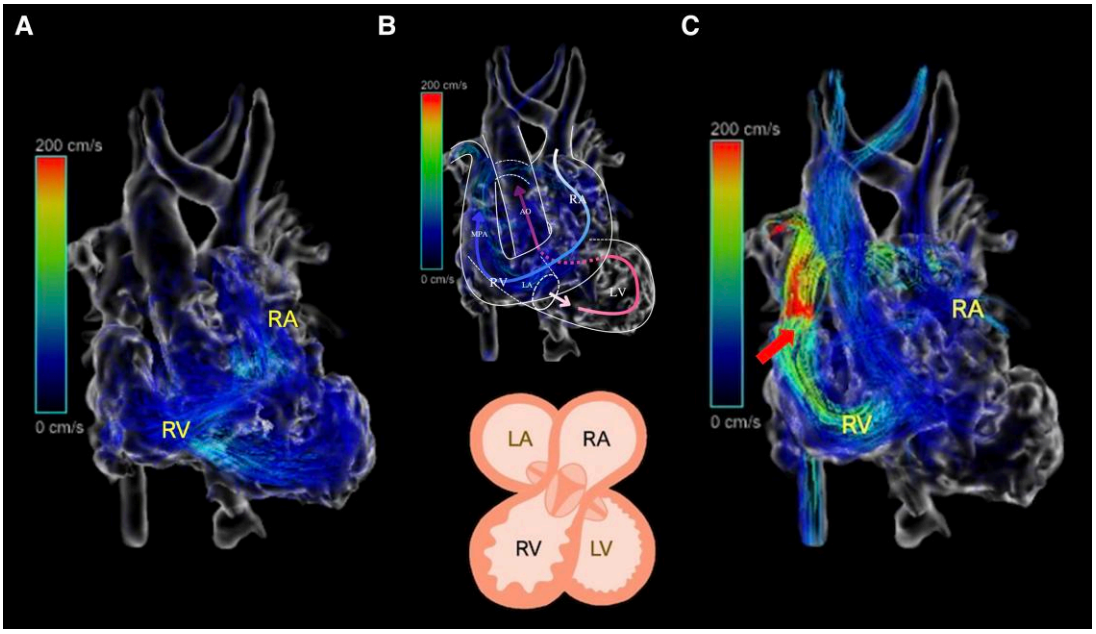


Figure 5 Four-dimensional cardiovascular magnetic resonance flow imaging. (*A* and *B*) The crossing of inflow streams of the two ventricles; (*C*) the accelerated flow from below the pulmonic valve.

Lead author biography

Assistant professor, Kwannapas Saengsin, MD, PhD, is a paediatric cardiologist at the Department of Pediatrics, Faculty of Medicine, Chiang Mai University, Chiang Mai, Thailand.

Consent: The authors confirm that the patient gave written consent for the submission and publication of this case, in accordance with COPE guidance.

Conflict of interest. None declared.

Funding: None declared.

References

1. Oliveira ÍM, Aiello VD, Mindêllo MM, Martins YO, Pinto VC Jr. Criss-cross heart: report of two cases, anatomic and surgical description and literature review. *Rev Bras Cir Cardiovasc* 2013;**28**:93–102.
2. Anderson RH. Criss-cross hearts revisited. *Pediatr Cardiol* 1982;**3**:305–313.
3. Cantinotti M, Bell A, Hegde S, Razavi R. A segmental approach to criss-cross heart by cardiac MRI. *Int J Cardiol* 2007;**118**:e103–e105.
4. Carminati M, Valsecchi O, Borghi A, Balduzzi A, Bande A, Crupi G, et al. Cross-sectional echocardiographic study of criss-cross hearts and superoinferior ventricles. *Am J Cardiol* 1987;**59**:114–118.
5. Podzolkov VP, Ivanitsky AV, Makhachev OA, Alekian BG, Chiaureli MR, Ragimov FR. Fontan-type operation for correcting complex congenital defects in a criss-cross heart. *Pediatr Cardiol* 1990;**11**:105–110.
6. Sidhu HS, Guleria M. Cardiac CT in criss-cross heart. *BJR Case Rep* 2021;**7**:20200096.
7. Anderson RH, Shinebourne EA, Gerlis LM. Criss-cross atrioventricular relationships producing paradoxical atrioventricular concordance or discordance. Their significance to nomenclature of congenital heart disease. *Circulation* 1974;**50**:176–180.
8. Ming Z, Yumin Z. Magnetic resonance evaluation of criss-cross heart. *Pediatr Cardiol* 2008;**29**:359–365.
9. Morelli PJ, Kimball TR, Witt SA, Meyer RA. Echocardiographic considerations in demonstrating complex anatomy of criss-cross atrioventricular valves and discordant atrioventricular and ventriculoarterial relations. *J Am Soc Echocardiogr* 1996;**9**:727–729.
10. Nagatsu M, Harada Y, Takeuchi T, Goto H, Ota Y. Can concordant criss-cross heart be ameliorated by hemodynamic changes? *Ann Thorac Surg* 1995;**60**:699–701.
11. Sato K, Ohara S, Tsukaguchi I, Yasui K, Nakada T, Tamai M, et al. A criss-cross heart with concordant atrioventriculo-arterial connections. Report of a case. *Circulation* 1978;**57**:396–400.
12. Saengsin K, Pickard SS, Prakash A. Utility of cardiac CT in infants with congenital heart disease: diagnostic performance and impact on management. *J Cardiovasc Comput Tomogr* 2022;**16**:345–349.
13. Kappanayil M, Koneti NR, Kannan RR, Kottayil BP, Kumar K. Three-dimensional-printed cardiac prototypes aid surgical decision-making and preoperative planning in selected cases of complex congenital heart diseases: early experience and proof of concept in a resource-limited environment. *Ann Pediatr Cardiol* 2017;**10**:117–125.
14. Alifu A, Wang H, Su Y, Chen R. Case report: use of three-dimensional technology in criss-cross heart with double outlet right ventricle. *Front Cardiovasc Med* 2023;**10**:1172104.
15. Rizk J. 4D flow MRI applications in congenital heart disease. *Eur Radiol* 2021;**31**:1160–1174.
16. Urmeneta Ulloa J, Álvarez Vázquez A, Martínez de Vega V, Cabrera J. Evaluation of cardiac shunts with 4D flow cardiac magnetic resonance: intra- and interobserver variability. *J Magn Reson Imaging* 2020;**52**:1055–1063.