Cardiology in the Young

cambridge.org/cty

Case Report

Cite this article: Larsson M, Synnergren M, and Rodrigo T (2025) Case report: morphological challenges to surgical repair in complete atrioventricular septal defect with isolated ventricular component. *Cardiology in the Young* **35**: 1085–1087. doi: 10.1017/S1047951125001441

Received: 7 February 2025
Revised: 20 February 2025
Accepted: 21 February 2025
First published online: 2 April 2025

Keywords:
atrioventricular septal defect; cardiac morphology; cardiothoracic surgery; trisomy 21; CHD

Corresponding author:
Thushara Rodrigo;
Email: thushara.rodrigo@gosh.nhs.uk

© The Author(s), 2025. Published by Cambridge University Press.

Case report: morphological challenges to surgical repair in complete atrioventricular septal defect with isolated ventricular component

Martina Larsson[1], Mats Synnergren[2] and Thushara Rodrigo[1]

[1]Department of Paediatric Cardiology, Great Ormond Street Hospital, London, UK and [2]Department of Cardiothoracic Surgery, Great Ormond Street Hospital, London, UK

Abstract

We describe a trisomy 21 patient with postnatal diagnosis of atrioventricular septal defect with isolated ventricular component who had a complicated post-surgical course following complete repair. Clinical outcomes included moderate-severe residual atrioventricular valve regurgitation needing re-operation, complete heart block leading to pacemaker insertion, seizures secondary to subdural haemorrhages, and chylothorax. We describe the surgical considerations specific to this cardiac morphology.

Case report

This patient presented in the newborn period with respiratory distress. They received a postnatal diagnosis of complete atrioventricular septal defect and trisomy 21 despite having had normal antenatal scans. The cardiac morphology on initial echocardiogram indicated a common partitioned atrioventricular valve with two well-developed papillary muscles in the left ventricle. There was a secundum atrial septal defect with no primum component, but a large inlet ventricular septal defect, and small patent ductus arteriosus (Figure 1). The left atrioventricular valves showed moderate regurgitation through the zone of opposition without stenosis. The left atrioventricular valve measured 13 mm. In the context of mitral valve Z-scores, it was 1.4.[1] The right atrioventricular valve had attachments to the septal crest with mild regurgitation and no clear straddling attachments.

At two months of age, the patient was initiated on diuretics along with feeding alteration due to heart failure. The full surgical repair was undertaken at four months of age at a weight of 4.4 kg, following multidisciplinary team discussion. Intraoperative assessment revealed a complete atrioventricular septal defect—Rastelli type B—with a small left atrioventricular valve, a small size cleft, a stiff and small superior bridging leaflet attached to the primary septum, and prolapsing inferior bridging leaflet. In addition, there were straddling chords retracting through the ventricular septal defect, which had not been identified pre-operatively.

The ventricular septal defect was closed using bovine pericardial patch. Around the prolapsing inferior leaflet, the straddling chords were partly divided and reattached, with one of the chords remaining intact in the ventricular septal defect and attaching to the right side of the atrioventricular valve. The cleft was then closed. Intraoperative transoesophageal and epicardial echocardiograms following initial repair demonstrated moderate right atrioventricular valve regurgitation and severe left atrioventricular valve regurgitation. This resulted in recommencing cardiopulmonary bypass to attempt repair of the left atrioventricular cleft. Due to the stiff and prolapsing superior bridging leaflet with minimal valve tissue, mattress sutures were applied to the cleft further out; with moderate regurgitation following reintervention with a plan to allow the child to grow and monitor the atrioventricular valve regurgitation. The total bypass time was 224 minutes. The patient was in heart block, hence came out of the theatre with DDD pacing.

Following admission to ICU, complications included arrhythmia, seizures, chylothorax, and evolving severe left atrioventricular valve regurgitation. The arrhythmia initially manifested as atrial flutter with variable block requiring amiodarone infusion, attempted overdrive pacing with epicardial leads, and two episodes of synchronised cardioversion. This later progressed to complete heart block requiring biventricular pacemaker insertion on post-operative day 21. Clinical seizures occurred on post-operative day three, which were secondary to subdural haemorrhages identified on neuroimaging. This was medically managed with Levetiracetam. The chylothorax was managed with dietary modification and drainage for two days.

The most important complication related to her cardiac substrate was severe residual atrioventricular valve regurgitation due to fixed superior bridging leaflet, appearance of an open

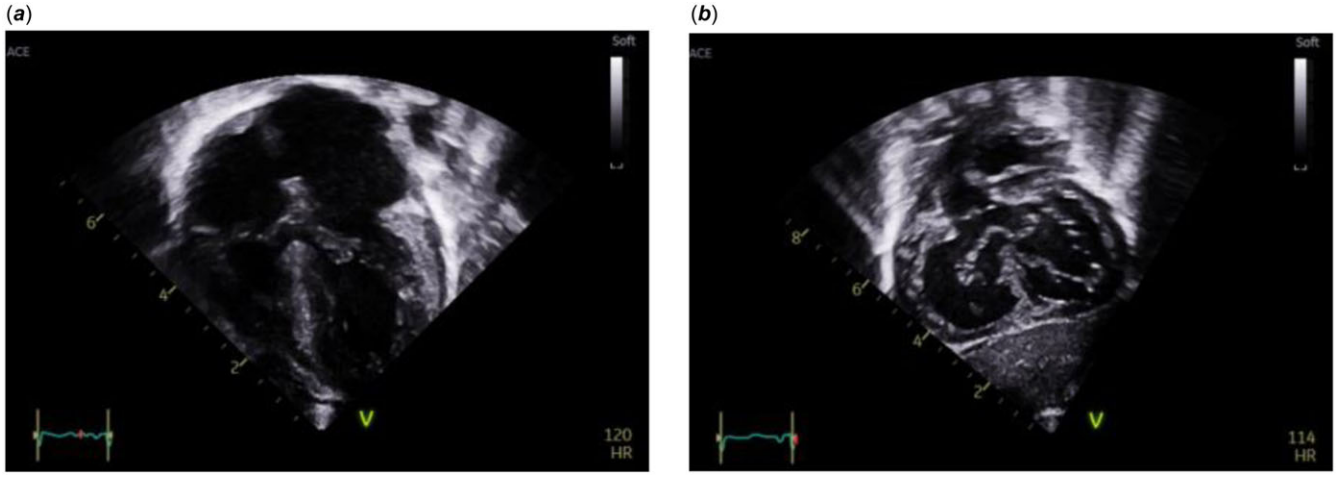


Figure 1. (a) Apical 4 chamber view showing isolated inlet ventricular defect; (***b***) Subcostal view showing common atrioventricular value.

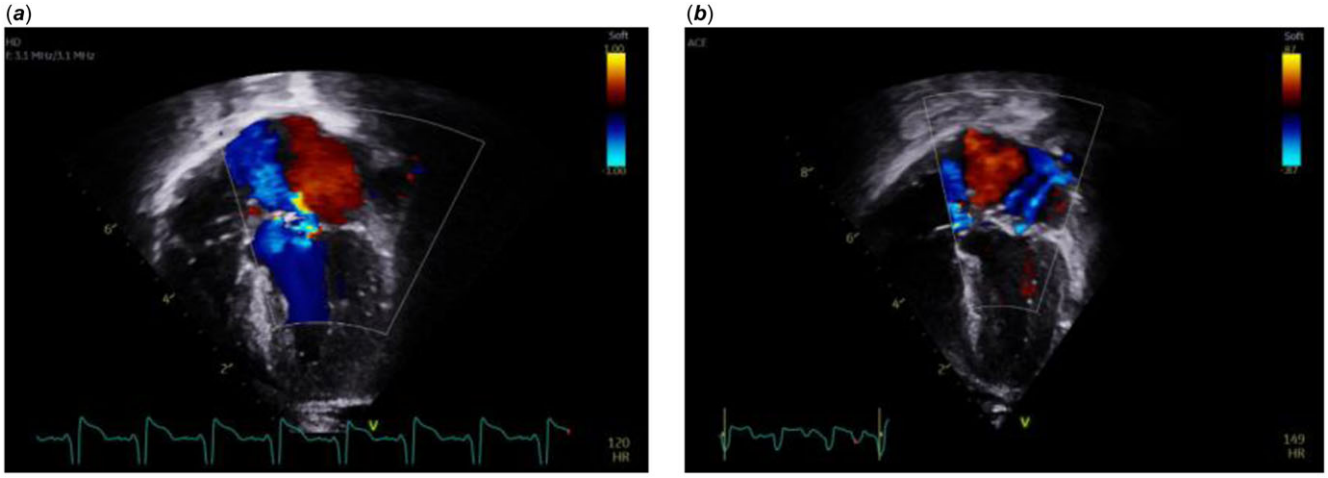


Figure 2. (***a***) Servere left atrioventricular valve regurgitation (AVVR) after first repair; (***b***) Echocardiogram on discharge showing improved left AVVR.

cleft, and prolapse of the inferior bridging leaflet, with preserved ventricular function. Medical management failed and the decision regarding reoperation was complicated by the subdural haemorrhage.

Following multidisciplinary team discussion, on post-operative day 35, the patient underwent a redo surgery with left atrioventricular valve repair with patch and commissuroplasty and permanent dual-chamber pacemaker insertion. The intraoperative findings were an intact previously repaired cleft with a tear in the superior bridging leaflet and prolapse of the inferior bridging leaflet. The superior bridging leaflet was repaired after mobilising it at the level of the annulus with an equine decellularised patch to the basal part of the leaflet and resuturing the tear in the superior bridging leaflet. After a saline injection, it was also decided to complete the repair with commissuroplasty to improve the coaptation between the leaflets. The right atrioventricular valve appeared to be competent with no major regurgitation jet during this time. Intraoperative transoesophageal and epicardial echocardiograms identified preserved systolic function with residual moderate left atrioventricular valve regurgitation, improved from severe regurgitation (Supplementary Figure). Considering that the patient was too small for a mechanical valve and not appropriate for Melody™ transcatheter pulmonary valve without further discussion, the residual regurgitation was accepted.

The patient made satisfactory clinical progress with a total admission duration of 66 days, including 44 days in the ICU and 19 cumulative days on ventilation. Transthoracic echocardiogram prior to discharge showed preserved biventricular function with moderate residual right atrioventricular valve regurgitation via multiple jets and moderate residual left atrioventricular valve regurgitation via multiple jets without residual shunts (Figure 2). The patient was discharged home on Captopril and Furosemide.

Discussion

Atrioventricular septal defects are one of the most common presentations of CHD in patients with trisomy 21.[2] There are minimal reports of complete atrioventricular septal defect with isolated ventricular component.[3,4] This case describes post-operative clinical outcomes in a unique cardiac morphology of

https://doi.org/10.1017/S1047951125001441 Published online by Cambridge University Press

atrioventricular septal defect with isolated ventricular component. The resulting comorbidities complicated the decision-making process.

There were numerous surgical challenges considering the cardiac morphology, particularly in addressing the residual atrioventricular valve regurgitation. In the index case, it was felt that increased tension after the cleft repair due to the described morphology led to severe residual left atrioventricular valve regurgitation. Hence, preoperative assessment should be thorough, and the surgeon should be aware of the technical difficulties of the cleft repair.

A retrospective review at one institution identified 16 patients with "absent or diminutive primum atrial components" of a cohort of 183 patients presenting for complete atrioventricular septal defect repair. They report a 31% rate of reoperation for left atrioventricular valve regurgitation in this cohort, of which two developed cleft dehiscence. A pre-operative risk factor for reoperation was identified as "leftward malposition of the atrial septum on the common atrioventricular valve."[5] These findings are corroborated by our case experience.

A review of cardiac specimens found that 6% of specimens ($n = 5$ of 78) with atrioventricular septal defect had an isolated ventricular component.[6] They also identified that valve leaflets are more prone to prolapse in this specific morphology, and this arrangement may play a role in the development of valve insufficiency. They hypothesised it is "highly likely" that there is "some degree of nodal displacement" in this specific morphology. In the initial intervention for this patient, a transseptal approach was used to approach the left atrioventricular valve. Studies have not identified an increase in post-operative arrhythmia with a transseptal approach compared to left atriotomy.[7,8] In our scenario, repair of the large ventricular septal defect and prolonged cardiopulmonary bypass time with two bypass runs imposed additional risks for developing complete heart block; though the prolonged cardiopulmonary bypass time likely reflects the challenges of repair. Additionally, children with trisomy 21 may have increased risk of postoperative chylothorax secondary to lymphatic dysplasia.[9]

This case highlights the complicated post-operative course in a patient with atrioventricular septal defect and isolated ventricular component. Thorough pre-operative assessment will help in surgical planning and particularly in counselling parents. Additionally, this case elaborates on the rational decision-making required in a complex post-operative scenario.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951125001441.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Competing interests. None.

Ethical standards. Not applicable.

References

1. Lopez L, Colan S, Stylianou M, et al. Relationship of echocardiographic Z scores adjusted for body surface area to age, sex, race, and ethnicity. Circ Cardiovasc Imaging 2017; 10: doi: 10.1161/CIRCIMAGING.117.006979.
2. Dimopoulos K, Constantine A, Clift P, et al. Cardiovascular Complications of Down Syndrome: Scoping Review and Expert Consensus. Circulation 2023; 147: 425–441. doi: 10.1161/CIRCULATIONAHA.122.059706.
3. Kaski JP, Wolfenden J, Josen M, et al. Can atrioventricular septal defects exist with intact septal structures? Heart 2005; 92: 832–835. doi: 10.1136/hrt.2005.069278.
4. Suzuki M, Kobayashi M, Fukae K. Atrioventricular septal defect with an absent or tiny ostium primum defect: a case series of three surgical cases. In J Thorac Cardiovasc Surg 2022; 38: 666–669. doi: 10.1007/s12055-022-01404-4.
5. Kwon MH, Schultz AH, Lee M, et al. Complete atrioventricular septal defect with absent or diminutive primum component: incidence, anatomic characteristics, and outcomes. J Thorac Cardiovasc Surg 2022; 163: 1156–1162. doi: 10.1016/j.jtcvs.2021.06.041.
6. Adachi I, Uemura H, McCarthy KP, et al. Morphologic features of atrioventricular septal defect with only ventricular component: further observations pertinent to surgical repair. J Thorac Cardiovasc Surg 2009; 137: 132–138.e2. doi: 10.1016/j.jtcvs.2008.03.032.
7. Khawaja ZR, Cambronero GE, Aboutabl YM, et al. Superior transseptal versus left atriotomy approaches in isolated mitral valve surgery. JTCVS Open 2024; 22: 208–213. doi: 10.1016/j.xjon.2024.08.019.
8. Guiraudon GM, Ofiesh JG, Kaushik R. Extended vertical transatrial septal approach to the mitral valve. Ann Thorac Surg 1991; 52: 1058–1062. doi: 10.1016/0003-4975(91)91281-Y.
9. Kan C-D, Wang J-N, Wu J-M, et al. Isolated chylopericardium after intrapericardial procedures: possible role of inadvertent right efferent lymphatic trunk injury. Tex Heart Inst J 2007; 34: 82–87. http://www.ncbi.nlm.nih.gov/pubmed/17420799.