

# Late recovery of sinus rhythm following perioperative complete atrioventricular block in a child with congenitally corrected transposition

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## ABSTRACT

We report a patient with l-transposition of the great arteries who developed perioperative complete atrioventricular block at 5 years of age in conjunction with a modified double-switch operation, but had unexpected late recovery of normal sinus rhythm months later. This case highlights that even for patients with l-transposition, which is particularly vulnerable to developing both spontaneous and perioperative complete atrioventricular block, late recovery of atrioventricular node conduction may still be possible after surgery.

**Keywords:** Complete atrioventricular block, congenitally corrected transposition, l-transposition of the great arteries, surgical atrioventricular block

## INTRODUCTION

Congenitally corrected transposition, or l-transposition of the great arteries, accounts for 0.05% of congenital cardiac malformations with a reported incidence of 1/33,000 live births.<sup>[1]</sup> In atrial situs solitus, l-transposition is levomalposition of the aorta; the morphologic left atrium connects across the tricuspid valve to the right ventricle, which gives rise to the aorta. A ventricular septal defect is the most common coexisting anomaly and is present in about 80% of patients.<sup>[2]</sup> The natural history of l-transposition includes very high risk for abnormal conduction including complete atrioventricular block, likely stemming from abnormal positioning of the atrioventricular node that is often located anteriorly in the right atrium at the lateral junction of the pulmonary and mitral valve.<sup>[3]</sup> At birth, 5%–10% of infants will have complete atrioventricular block; by adulthood, the prevalence of complete atrioventricular block increases to about 30% with a lifetime cumulative risk that continues to rise.<sup>[4]</sup> Patients

with l-transposition are at particularly increased risk of perioperative atrioventricular block, occurring in more than 20% of these patients at cardiac surgery.<sup>[5]</sup> We report a patient with l-transposition who developed perioperative complete atrioventricular block at 5 years of age in conjunction with a modified double-switch operation, but had unexpected late recovery of normal sinus rhythm several months later.

## CASE REPORT

A Caucasian female was born with l-transposition of the great arteries along with a large ventricular septal defect. At 2 months of age, she underwent a pulmonary artery band procedure to limit pulmonary overcirculation [Figure 1a]. She remained clinically asymptomatic in normal sinus rhythm with preserved biventricular function. To prevent late complications from a systemic right ventricle, she had a modified

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double-switch operation performed at 5 years of age. Along with band removal and ventricular septal defect closure, she underwent an arterial switch operation and hemi-Mustard (inferior vena cava to the right ventricle baffle) atrial switch with bidirectional Glenn cavopulmonary anastomosis [Figure 1b]. This technique avoids superior vena cava suture lines within the atrium and may prevent later complications of baffle leak and sinus node dysfunction when compared to the traditional double switch.<sup>[5]</sup>

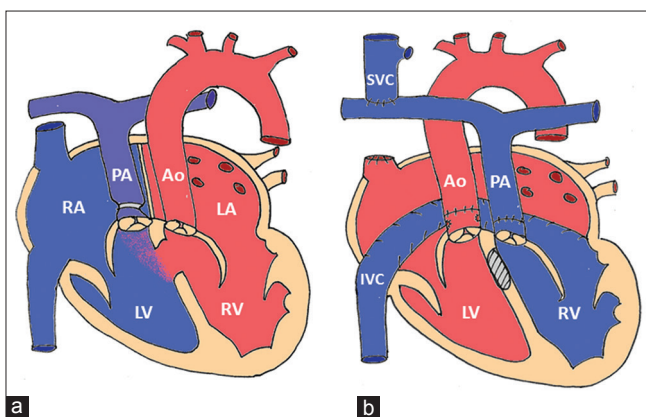
At the time of surgery, she developed complete atrioventricular block with a narrow complex escape rhythm, so a dual-chamber epicardial pacemaker system (Medtronic ADDRL1 Adapta DR, Minneapolis, MN, USA) was implanted 1 week later. On postoperative day 31, device interrogation confirmed ongoing atrioventricular block with 100% ventricular pacing requirement [Figure 2a]. On postoperative day 53, device interrogation showed >99% ventricular pacing requirement with one episode of breakthrough sinus tachycardia at 183 beats/min during a period of exercise, so her device was changed to Managed Ventricular Pacing (AAIR<->DDIR). On postoperative day 84, interrogation revealed <0.3% ventricular pacing with a surface electrocardiogram that showed normal sinus rhythm with a PR interval of 144 ms and a rate of 114 beats/min; her P-wave morphology and PR interval matched her preoperative electrocardiograms. She remained in sinus rhythm with intact atrioventricular node conduction on multiple subsequent electrocardiograms, and device interrogations have shown no additional ventricular pacing. During an in-office pacemaker check to provocatively test her atrioventricular node more than

2 years later, atrial pacing at 180 beats/min showed 1:1 conduction, confirming normal atrioventricular node function [Figure 2b].

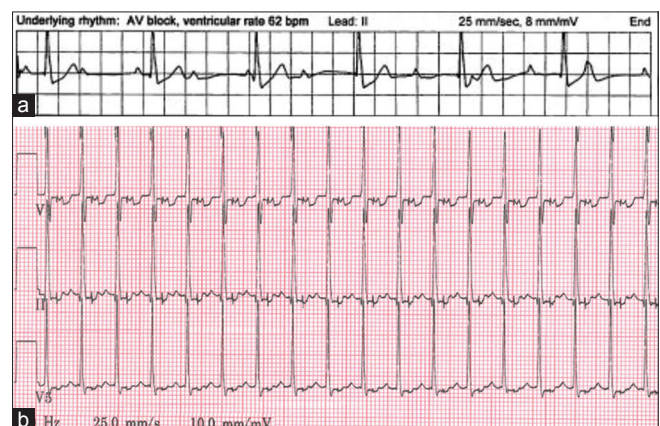
## DISCUSSION

Complete atrioventricular block is a complication that can occur in 1%–3% of palliative surgeries for all congenital heart disease.<sup>[6]</sup> While atrioventricular block may only be transient following surgery, patients with block that persists beyond postoperative days 7–14 are much less likely to recover atrioventricular node conduction. Weindling *et al.* reported that >95% of patients who regain atrioventricular node function do so by postoperative day 9.<sup>[7]</sup> The current American Heart Association guidelines make a Class I recommendation for permanent pacemaker placement for complete atrioventricular block that is not expected to resolve or that persists at least 1 week following surgery for congenital heart disease.<sup>[8]</sup>

A small subset of patients may have late return of normal atrioventricular conduction following surgery for congenital heart disease. One institution reported that 7 of 72 patients had late return of normal atrioventricular node function at a median of 41 days (range 18–113 days).<sup>[9]</sup> Another reported that 7 of 58 patients with pacemaker placement recovered conduction between 3 weeks and 7 years following surgery.<sup>[10]</sup> Two of the patients cited in these reports had discordant atrioventricular connections, though the timing of their late recovery is unclear. Despite requiring pacemaker implantation, our patient’s recovery of normal sinus rhythm more than 2 months after surgery highlights that, even for those patients particularly vulnerable to developing both spontaneous and perioperative complete atrioventricular block, late recovery of atrioventricular node conduction may still be possible.



**Figure 1:** (a) Diagram showing I-transposition of the great arteries with a ventricular septal defect status-post a pulmonary artery band procedure. (b) Diagram showing I-transposition of the great arteries status-post band removal, ventricular septal defect closure, arterial switch operation, and hemi-Mustard (inferior vena cava to right ventricle baffle) atrial switch with bidirectional Glenn cavopulmonary anastomosis. RA: Right atrium, LA: Left atrium, RV: Right ventricle, LV: Left ventricle, PA: Pulmonary artery, Ao: Aorta



**Figure 2:** (a) Device interrogation on postoperative day 31 showing persistent complete atrioventricular block. (b) Device interrogation nearly 2 years postoperatively to provocatively test the patient’s atrioventricular node. Atrial pacing at 180 beats/min showed 1:1 conduction, confirming normal atrioventricular node function

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

### Disclaimer

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