

Uhl's anomaly: A one and a half ventricular repair in a patient presenting with cardiac arrest



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Uhl's anomaly, first reported in 1952, is an extremely rare congenital cardiac defect characterized by partial or complete loss of the right ventricular myocardium and unknown etiology. Fewer than 100 cases have been described. The response to medical management is poor and there is no known ideal surgical approach or timing for treatment. We report the case of a previously active adolescent male presenting with cardiac arrest, who underwent successful bidirectional cavopulmonary anastomosis ("Glenn" anastomosis) with right atrial reduction and right ventricular free wall plication.

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Introduction

Uhl's anomaly, first reported in 1952, is an extremely rare congenital cardiac defect characterized by partial or complete loss of the right ventricular myocardium and unknown etiology. Fewer than 100 cases have been described [1]. Histopathologically, the myocardial layer is replaced by nonfunctional fibroelastic tissue

resulting in the right ventricular free wall having a parchment-like appearance. These changes lead to progressive right heart failure and/or significant arrhythmias, including ventricular arrhythmias, bilateral bundle branch block, and atrioventricular (AV) block [1–6]. Uhl's anomaly typically presents in infancy [2], which was the case in our patient. However, our patient was stable and asymptomatic until presenting in adolescence with cardiac arrest.

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Case report

Our patient is 12-year-old male with a history of Uhl's anomaly and regular follow-up who was playing at home when he became cyanotic and lost consciousness. Cardiopulmonary resuscitation was performed by his aunt and 911 was called. On arrival, paramedics found the patient to be in ventricular fibrillation. Defibrillation was performed twice with return of spontaneous circulation, and he was urgently transported to a nearby emergency department. After initial stabilization in the outlying facility, he was transported by air to our facility, and upon arrival he was noted to be in normal sinus rhythm. During continued resuscitation, he experienced a generalized tonic-clonic seizure attributed to low cardiac output, and high-dose vasoactive medications were necessary to support the circulation.

Shortly after arrival to the intensive care unit, echocardiography revealed severe right atrial and right ventricular dilation with a thin-walled right ventricle and severely depressed right ventricular function (Fig. 1). The severity of the right ventricular dilation contributed to impaired left ventricular filling and severe tricuspid valve insufficiency. His electrocardiogram was consistent with right atrial and right ventricular enlargement. Cardiac magnetic resonance imaging demonstrated a severely thinned and noncontractile right ventricular parietal wall that lacked fat tissue, consistent with the diagnosis of Uhl's anomaly.

Six days after admission, and after significant clinical recovery, he underwent an electrophysiology study which revealed an unusual form of AV nodal reentrant tachycardia (narrow QRS

tachycardia) and fascicular-ventricular reentrant circuit (wide complex tachycardia) [3]. Given the gravity of his presentation and the unusual nature and severity of his anatomic derangements, a number of options were considered, including heart transplantation, but the family was not amenable to this option. After considerable discussion of other alternative surgical options, he underwent surgical right atrial reduction, right ventricular plication, and a bidirectional cavopulmonary anastomosis ("Glenn" procedure). Intraoperatively, severe enlargement of the right ventricular cavity, a regurgitant tricuspid valve with severe annular dilation, and a very small atrial septal defect were confirmed. The right ventricular appearance was similar to what was described on preoperative imaging. A one and a half ventricular approach was felt to be the optimal approach to achieve good long term functional outcome. The mechanism of tricuspid valve insufficiency was believed to be secondary to the enlarged right ventricle. Volume unloading with a bidirectional Glenn procedure led to better tricuspid valve competence. To optimize hemodynamics, the chest was left open with projected return to the operating room in 24 hours for implantation of an epicardial ICD with delayed sternal closure. The patient tolerated both procedures well and was extubated quickly following the second procedure.

On postoperative Day 2, milrinone and dopamine infusions were weaned and discontinued. The patient experienced no postoperative arrhythmias. He required supplementary oxygen by nasal cannula for several days due to expected cyanosis as he began to ambulate and his body adjusted to the new single-ventricle physiology.

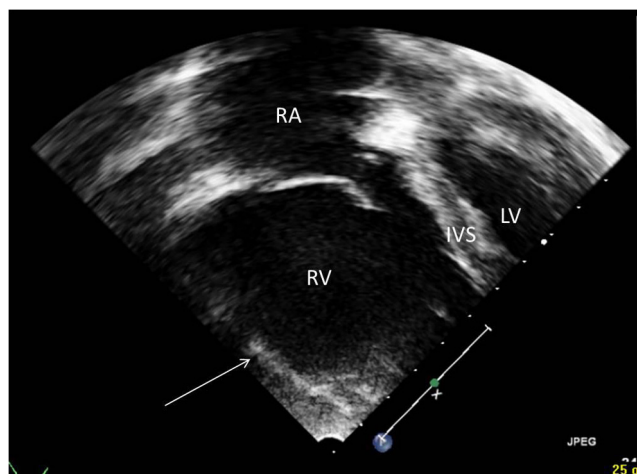


Figure 1. Apical four-chambered view of the heart showing dilated right atrium (RA) and right ventricle (RV). The right ventricular wall (arrow) is thin. The interventricular septum (IVS) is shifted towards the left ventricle (LV).

This was weaned off gradually and discontinued on postoperative Day 8. Prior to discharge, pulse oximetry was consistently in the high 70 s to low 80 s on room air. As anticipated, echocardiography demonstrated improvement in dilation of the right ventricle, but given the underlying anomaly, there was essentially no ventricular contraction (unchanged from preoperative studies). Left ventricle systolic function was normal; mild tricuspid regurgitation and significant improvement in the degree of tricuspid valve prolapse were also noted.

At follow-up 5 months later, he was symptom-free with no arrhythmias and no ICD shocks, with oxygen saturations in the high 80 s to low 90 s on room air.

Discussion

Uhl's anomaly is an extremely rare cardiac malformation that typically presents in infancy or early childhood. Few adult cases of Uhl's anomaly have been reported, occasionally involving partial Uhl's anomaly (partial absence of the right ventricular musculature) [2]. Furthermore, the response to medical management is poor and there is no known ideal surgical approach or timing for treatment. Reported surgical interventions described during infancy or early childhood include a single ventricle strategy (bidirectional cavopulmonary anastomosis, atrial septectomy, and disconnection of the main pulmonary artery) [1], cardiac transplant [7], one and a half ventricle repair with partial right ventriculectomy [8], and a Fontan procedure [9].

Yoshii and associates [8] documented the use of the one and a half ventricular repair in Uhl's anomaly; however, their operation was performed when the patient was 9 months old. We report the case of a previously active adolescent male presenting with cardiac arrest who underwent successful bidirectional cavopulmonary anastomosis ("Glenn" anastomosis) with right ventricular free wall plication. A one and a half ventricular repair is employed when there is a normally functioning left ventricle (normal size and function) along with

a dysfunctional right ventricle, inadequate to handle the entire cardiac output, but considered of sufficient size to handle one half to two thirds of the cardiac output [10]. Given the patient's cardiac imaging, we determined that a one and a half ventricular repair approach, coupled with ICD placement to address arrhythmia risk, would be the best option for palliation of this exceedingly rare defect. In conclusion, we are satisfied with the procedure, as the patient has had a good outcome with resolution of clinical symptoms and no arrhythmias during our short-term follow-up.

Conflicts of interest

None to disclose for all authors.

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