

# One and a Half Ventricular Repair for Uhl's Anomaly with One Year Follow Up

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Uhl's anomaly is characterized by complete or partial absence of the myocardium of the right ventricle, with apposition of the endocardium and epicardium. We report our experience with surgical treatment of this anomaly.

**Keywords:** Myocardium, Uhl's anomaly

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## Introduction

Among the embryogenic defects in right ventricular development there is a very unusual form which affects the muscular tissue.<sup>1</sup> Uhl's anomaly which was first described by Uhl in 1952, consists of almost total absence of right ventricular (RV) myocardium.<sup>2</sup> In 1905, Osler described a case in which the right ventricular wall was thin like paper.<sup>3</sup> Uhl's anomaly as recognized is extremely rare, and by 1979, fewer than 20 cases had been reported, each individually as a case report.<sup>4</sup>

Primary non-development of myocytes or selective apoptosis are supposed to be the cause of this anomaly.<sup>5</sup>

Diagnosis of Uhl's anomaly was previously made at autopsy, but now advanced imaging methods<sup>3,6</sup> Few reports have been published on surgical treatment.<sup>7</sup> We report a case of Uhl's anomaly undergoing the bidirectional Glenn shunt, namely, one and a half ventricle repair, combined with partial right ventriculectomy and tricuspid annuloplasty.

## Case Report

A 9-year-old girl, weighing 25 kg was admitted to our hospital on account of dyspnea and mild cyanosis. On physical examination, she was slightly tachypneic, mildly cyanotic with symmetrical puls-

es. New York Heart Association (NYHA) functional class was 3. On cardiac auscultation the intensity of the first heart sound was decreased and a II/VI pansystolic murmur was heard in the left lower sternal border. She also had a mild hepatomegaly. On chest X-ray marked cardiomegaly with right atrial enlargement could be seen. Electrocardiography showed tall P waves and diminished QRS amplitudes in right precordial leads. Echocardiography demonstrated marked dilation of the right cardiac chambers. Tricuspid valve leaflets were seen arising appropriately from the annulus and there was a severe low pressure tricuspid regurgitation. RV function was severely decreased.

On cardiac catheterization, marked dilatation of the right atrium, right ventricle and significant tricuspid insufficiency were confirmed. Manometry showed a mean pressure of 21 mmHg in the right atrium and 26/0-17 mmHg in right ventricle, main pulmonary artery pressure was 26/17 mmHg and its mean pressure was 13 mmHg. (Fig. 1)

Operation was performed with the aid of transesophageal echocardiography (intraoperative transesophageal echocardiography or IOTEE) monitoring. IO TEE confirmed transthoracic echocardiographic findings as well as decreased thickness of RV free wall to 2 mm. Tricuspid annulus diameter was 4.5 cm ( Z score was more than 2) (Fig. 2).

On close examination, both the right atrium and ventricle were massively enlarged and tricuspid valve annulus was severely dilated. RV had a thin

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**Figure 1.** Angiographic anteroposterior and lateral views of right ventricle showing severely enlarged right ventricle with thin wall.

and fibrotic wall. Therefore the RV free wall was partially resected and the tricuspid valve was repaired with No 32 Carpentier ring. Superior vena cava was then divided and with interposition of a Gore-Tex graft No 18 between superior vena cava and right pulmonary artery a bidirectional Glenn anastomosis was constructed. IOTEE post-cardiopulmonary bypass revealed decreased RV size, reduced tricuspid valve annulus to 3 cm and decreased tricuspid regurgitation with moderate RV dysfunction (Fig. 3). The patient had an uneventful postoperative course.

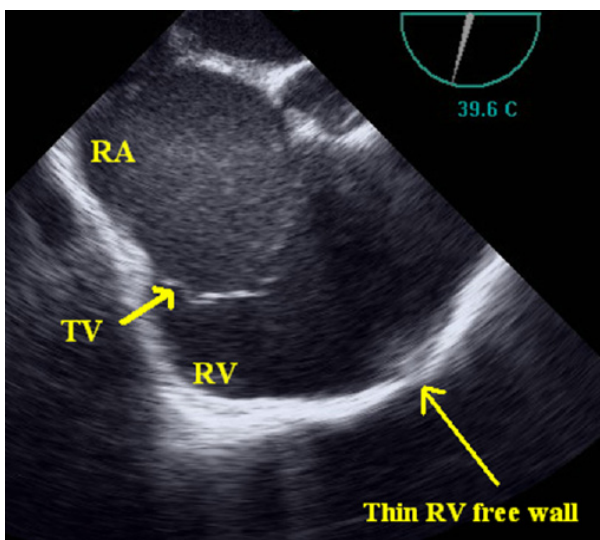
Pathologic specimens confirmed the Uhl diag-

nosis (Figs. 4 and Fig. 5)

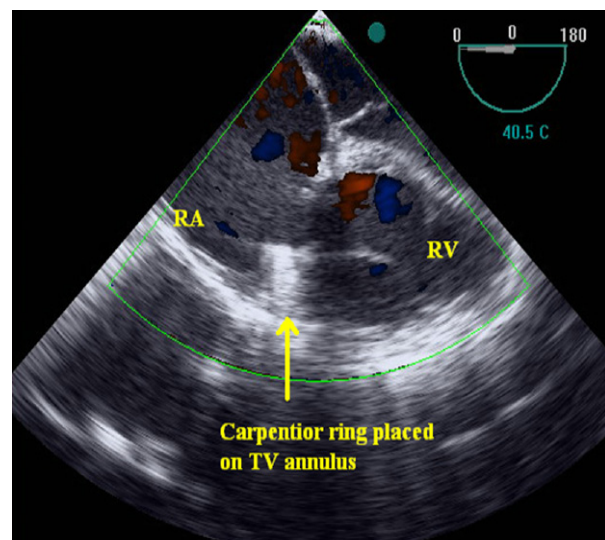
One year follow-up with serial transthoracic echocardiography showed moderate RV dysfunction and moderate tricuspid regurgitation. On transthoracic echocardiography tricuspid valve annulus was 3 cm (Z score=2) and RV was moderately enlarged. Patient's general condition was better with New York Heart Association (NYHA) functional class 2. (Fig.6)

### Discussion

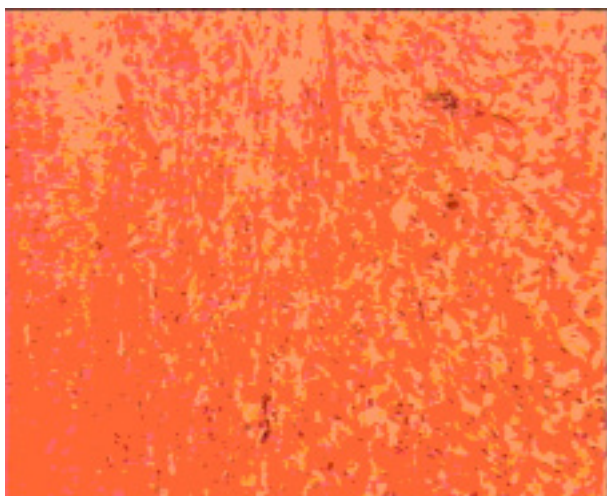
Advances in echocardiography methods have made it possible to diagnose the Uhl's anomaly ac-



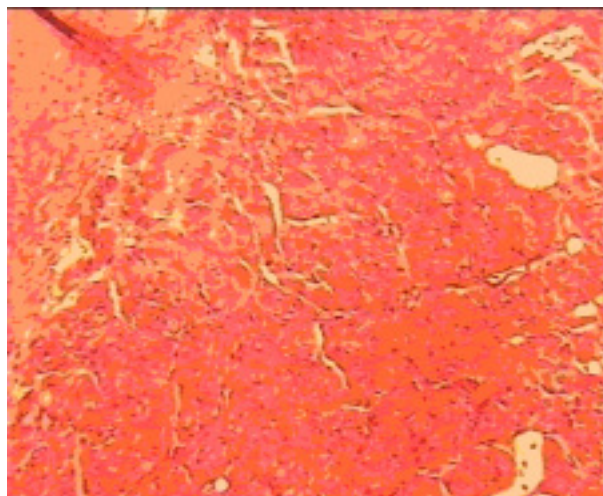
**Figure 2.** IOTEE pre procedure showing dilation of RV and tricuspid annulus and thin RV free wall



**Figure 3.** IOTEE post- procedure showing reduced RV size and tricuspid annuloplasty



**Figure 4.** Specimen from RV biopsy showing a fibromyxoid and fatty tissue with congestion and few entrapped bundles of myocyte



**Figure 5.** Specimen from right atrial biopsy showing bundles of moderately hypertrophic cardiac myocytes with some foci of myocytolysis, interstitial fibrosis and fresh hemorrhage

curately. Echocardiography can reliably estimate the right ventricular diastolic wall thickness (mean  $4.0 \pm 1.62$  mm).<sup>8</sup> IOTEE can help the surgeon plan the surgery especially in congenital heart diseases by identifying the more detailed anatomy and hemodynamic evaluation.

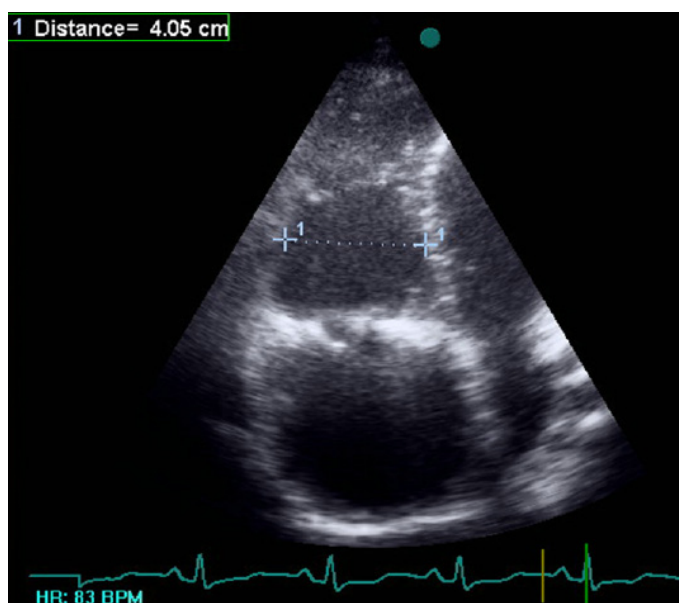
There are some published cases of successful surgical treatment for Uhl's anomaly by applying one and a half ventricle repair combined with right ventriculectomy. They underwent subsequent total cavopulmonary connection.<sup>9</sup> We carried out one and a half ventricle repair in order to reduce

RV volume and believe that reducing the RV size with partial right ventriculectomy will help adequate filling and contraction of left ventricle.

This procedure has produced satisfactory results after one-year follow-up and will be evaluated in the long term.

#### Acknowledgements

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**Figure 6.** Transthoracic echocardiography after one year showing 4 cm RV diameter

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