

Uhl's anomaly- a rare cause of congestive heart failure

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INTRODUCTION Uhl's anomaly is characterized by absence of the right ventricular myocardial layer. Although it appears to be of congenital origin, etiology is still uncertain. Patients usually present with congestive heart failure. Diagnosis is generally suggested by echocardiography. Medication treatment is only palliative, but surgical methods offer etiological cure.

CASE REPORT A 10-year old boy presented in 2014 with malaise, pallor, headache and dizziness followed by sudden collapse. After admission to hospital his condition progressed to cardiogenic shock and he was successfully reanimated. Diagnosis of right ventricular dilated cardiomyopathy was set by echocardiography and NMR. Uhl's anomaly and arrhythmogenic right ventricular dysplasia (ARVD) couldn't be differentiated because the parents refused myocardial biopsy. Implantation of cardioverter defibrillator was also recommended, which the parents also refused. Heart failure medications were prescribed and the patient was discharged. In the following years the patient presented several times with heart failure symptoms and was conservatively treated. In August 2018 he was put on the transplant waiting list and was hospitalized from early October because his heart function worsened with right ventricular ejection fraction of 13%. On January 4, 2019 modified bicaval cardiac transplantation was performed. Postoperatively the patient was hemodynamically stable, and three weeks later was released from the hospital.

CONCLUSION The overall incidence of heart failure in children is low, but the associated morbidity and mortality are high. Uhl's anomaly is exceedingly rare and usually presents with congestive cardiac failure. Early diagnosis and treatment are important in order to prevent life threatening complications.