Combined Heart and Liver Transplantation for Uhl's Anomaly: A Case Report

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ABSTRACT

Background. Uhl's anomaly is an extremely rare congenital heart defect characterized by absence of the right ventricle myocardium and preserved left ventricular myocardium. Although the disease has a poor prognosis and is generally fatal in the perinatal period, some patients may reach adulthood.

Methods. We describe a case of Uhl's anomaly complicated with heart failure and decompensated cardiac cirrhosis in a 42-year-old man treated by combined heart-liver transplant.

Results. The patient underwent heart transplant using the bicaval technique followed by subsequent liver transplant with the piggyback technique without venovenous bypass. Total ischemia time was 108 minutes for the heart and 360 and 25 minutes of cold and warm ischemia, respectively, for the liver. No intraoperative complications occurred. The patient was discharged without severe complications on postoperative day 22. Pathologic examination of the organs reported advanced cirrhosis of the liver and severe dilated myocardiopathy of right ventricle with absence of myocardium and a normal left ventricle. Twenty-seven months after the transplant the patient has been free from hospital admissions, with normal function of both transplanted organs.

Conclusions. We report the first successful combined heart-liver transplant for Uhl's anomaly indication in an adult patient. Despite of the insufficient knowledge of natural history of this exceptional disease, we successfully apply the management principles of other end-stage right heart disorders complicated with liver failure.

UHL 's anomaly (UA) is an extremely rare congenital heart defect of unknown origin characterized by complete or partial absence of the right ventricle (RV) myocardium [1,2]. Although UA has a poor prognosis and is generally fatal in the perinatal period in complete forms, patients may reach adulthood [3-5]. We report here a case of UA heart failure in an adult patient complicated with decompensated cardiac cirrhosis successfully treated by combined heart-liver transplant (CHLT).

CASE PRESENTATION

A 42-year-old man was referred for impaired liver function. The unexpected finding of cardiomegaly led to the diagnosis of UA

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0041-1345/20
https://doi.org/10.1016/j.transproceed.2021.08.036

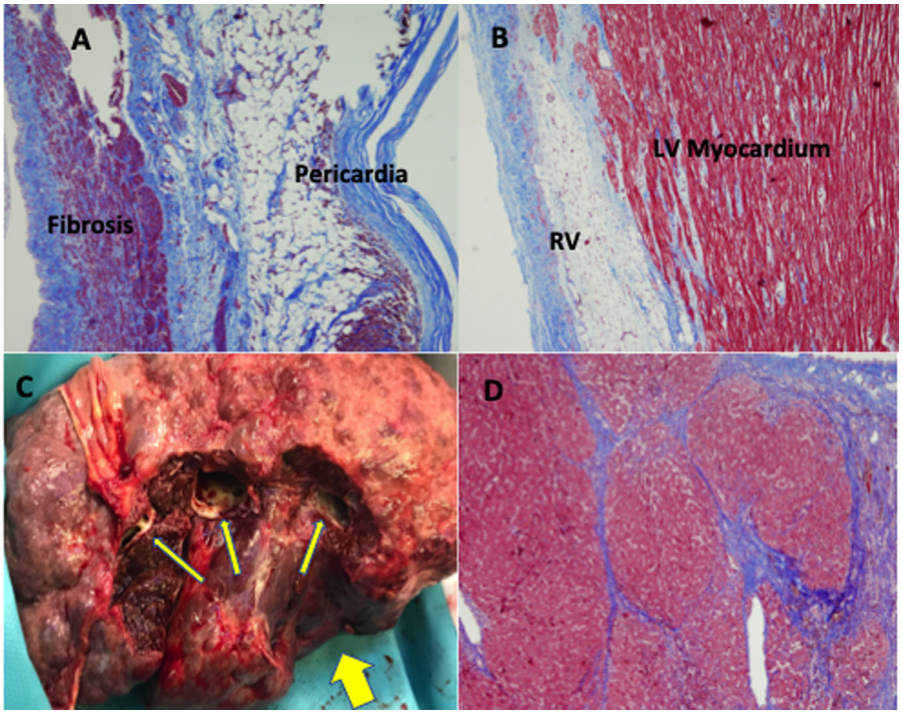

Fig 1. Pathology examination: **(A)** Microphotography of right ventricular (RV) wall shows a normal endothelium with subendothelial fibrosis and atrophy of the myocardial layer, next to normal pericardia. Trichromic stain, 40×. **(B)** Septum image shows a normally developed left ventricular (LV) myocardia and completely absence of RV myocardia, replaced by fibrosis. Trichromic stain, 40×. **(C)** Macroscopic image of explanted liver shows severe cirrhosis with dilated inferior vena cava (big arrow) and dilated hepatic veins (small arrows). **(D)** The histology of the liver shows and advanced liver disease in cirrhosis stage. Trichromic stain, 40×.

in childhood. He remained asymptomatic until age 26 when he started with paroxysms of supraventricular tachycardia. Right heart failure and liver cirrhosis with portal hypertension appeared thereafter. The indication of CHLT was set on the following criteria: (1) decompensated cirrhosis with a model for end-stage liver disease score of 12 with sarcopenia; (2) heart failure (RV ejection fraction 14%, RV volume 221 mL/m^2) and maximum oxygen consumption 11mL/kg/min. A 33-year-old brain-dead donor was offered. The heart transplant was performed using the bicaval technique. The thoracic surgery was followed by liver transplant with the piggyback technique. The patient was discharged on postoperative day 22 without complications. Pathologic examination of the liver showed advanced liver disease in cirrhosis stage; the heart presented a severe dilated myocardiopathy of right ventricle, with replacement of myocardial cells by fibrosis and a normal left ventricle (Fig 1). Twenty-seven months after the transplant, in June 2021, the patient is asymptomatic with normal function of both transplanted organs.

DISCUSSION

The UA is one of the differential diagnoses in RV disorders, which include RV arrhythmogenic dysplasia, Ebstein's anomaly, absence of the pericardium, and pulmonary atresia, just to name a few. In 1993 Gerlis et al [1] reported 84 cases of UA since the beginning of the 20th century. Although less than 15 cases of UA have been reported to the date in adult living patients, the prevalence of partial UA may be underestimated because patients can be asymptomatic for decades [3]. The CHLT is a complex procedure and represents about 0.3% of all heart transplants. Less than 250 cases are described to the date in the United States, and there are no published cohorts in Europe. Congenital heart diseases are indication of heart transplant in only 3% of adult recipients, whereas 22% of adult recipient receive a CHLT for congenital heart disease. In adults with congenital cardiopathy and confirmed liver cirrhosis, isolated heart transplant is associated with mortality rates up to 50%, making CHLT a necessary option. Moreover, a possible immunoprotective effect of CHLT over isolated heart transplant has been suggested by significant reduction of alloimmune injury in the heart allograft. The CHLT is a technically demanding procedure with mortality up to 15%.

CONCLUSIONS

In summary, we report the first successful CHLT for UA indication in an adult patient. Despite the lack of strong selection criteria for CHLT candidates, we believe that a priority for dual transplant should be proposed to avoid underestimation of the severity of both end-stage heart and liver failure and decrease waitlist mortality.

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