



A Rare Case of Tricuspid Regurgitation in the Setting of Orthotopic Heart Transplantation

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Abstract

A 13-year-old female was admitted to our hospital with terminal heart failure. After 50 days of extracorporeal life support, she underwent orthotopic heart transplantation (OHT). The donor had no cardiac history. During the following days after OHT, the patient presented right ventricular failure with severe tricuspid regurgitation (TR). The decision to perform a tricuspid valve repair was taken after right heart catheterization and multiple echocardiographies. The diagnosis of Ebstein's-like anomaly was made based on intraoperative echocardiographic features and anatomical observance during surgery. This case illustrates that congenital cardiac defects must be considered when facing an important TR after OHT.

Introduction

Tricuspid regurgitation (TR) is the most common valve abnormality after an orthotopic heart transplant (OHT) [1]. In the first few hours or days and in the majority of the situations, its cause is functional. However, an anatomic cause must be considered. While rare, an Ebstein's anomaly is among the congenital heart lesions that can present into adulthood without any or only few symptoms [2]. We present a case of Ebstein's-like anomaly diagnosed on the recipient after OHT from a donor with unknown history of cardiac disease.

Case Presentation

A 13-year-old female, with a respective weight and height of 38.1 kg and 159.5 cm, was followed-up in our institution after a previous mitral valve repair due to a congenital mitral valvulopathy that was associated with left ventricular dysfunction. At the last follow-up visit, very poor biventricular function was observed with severe mitral and tricuspid regurgitation and elevation of the right heart pressures. In fact, the patient had become more symptomatic during the last 3 previous months but did not consult. In face of extremely severe heart failure symptoms, it was decided to introduce an extracorporeal life support (ECLS) as bridging therapy until OHT. After 53 days of ECLS therapy, the transplantation team accepted a heart despite the advanced age of the donor. This decision was taken because of the prolonged duration of the ECLS therapy and the difficulty to find a matching organ.

The donor was a 65 year-old-female, with a respective weight and height of 60 kg and 165 cm, with known hypercholesterolemia, high arterial blood pressure, treated rheumatoid arthritis but no history of cardiac disease. The cause of death was cerebral hemorrhage. The transthoracic echocardiography (TTE) performed in order to propose for heart donation did not reveal any cardiac abnormalities either functional or anatomic. A bicaval OHT was accomplished with no difficulties although the patient needed two hours of assistance and high doses of inotropic support before weaning from the cardiopulmonary bypass (CPB). The ischemic time was 249 min. The intraoperative transesophageal echocardiography (TEE) showed a mild TR after improvement of the right ventricular (RV) function with the inotropic agents. The electrocardiogram performed postoperatively showed a right bundle branch block. The evolution during the following days was slow. Weaning from the ventilator was achieved after 6 days but renal failure and signs of RV

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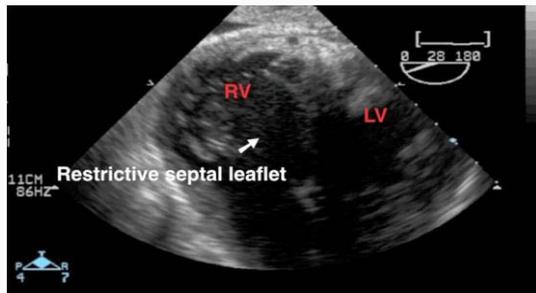
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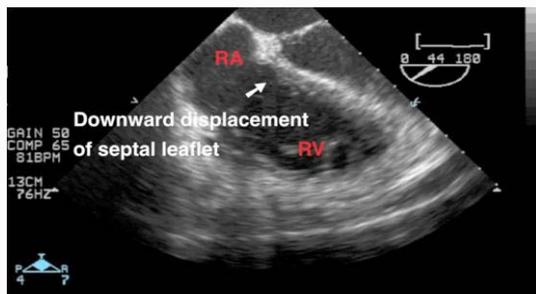
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Video 1: Transplanted heart characterized by a restrictive motion.



Video 2: Downward displacement of the septal leaflet.

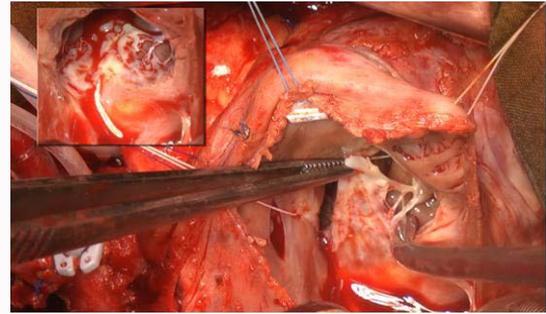


Figure 1: Anatomical observations during the tricuspid valve repair confirmed the diagnosis.

Functional TR

The common causes of functional TR after OHT include:

- RV dysfunction/failure: As RV dysfunction may lead to TR it must be considered as one of the main causes of TR. There are different reasons responsible for RV dysfunction such as prolonged ischemic time of the donor heart, donor-recipient size mismatch, and acute rejection, mechanical obstruction at the pulmonary anastomosis, pre-existing pulmonary arterial hypertension and pulmonary arterial hypertension induced by the protamine administration [2,3].

- Surgical techniques: biatrial versus bicaval. Many studies have concluded that the bicaval technique, also used by our surgical team, reduces the risk of TR after OHT among other benefits [4].

Anatomic TR

The donor’s heart is usually considered to be free of defects because an initial functional and structural evaluation is systematically performed on a heart considered for donation. Nevertheless, anatomical defects should be considered while facing a TR after OHT.

There are two forms of anatomic TR:

- Acquired: chest trauma is probably the most likely to be encountered in the context of a heart transplant. Other causes of acquired valvulopathy can be considered but are unlikely to be observed in this context: endocarditis, rheumatic valve disease, etc. [5].
- Congenital: In Belgium, congenital cardiac defects account for 6.6 per 1000 births including life births, late fetal deaths and termination of pregnancy for fetal anomaly [6].
- Ebstein’s anomaly is one of the congenital diseases of the tricuspid valve (TV). Its incidence is one to five per 200.000 births accounting for less than 1% of all congenital heart diseases. The main feature of this anomaly is an incomplete delamination of the septal and sometimes posterior leaflets of the TV during embryological development with an apical displacement of the functional annulus of the TV. This can lead to different abnormalities:

1. Atrialization of the RV resulting in a hypoplastic functional ventricle.
2. Variable TR with a central regurgitant orifice. A stenotic TV can be exceptionally found.
3. Enlargement of the right atrium.
4. Variable RV and LV dysfunction.
5. Variable RV inflow and/or outflow tract obstruction.

dysfunction persisted. A TTE was performed regularly during the following days after OHT. From day 12 after OHT and further on, TR progressively aggravated and became severe with a leaflet coaptation defect of 1 cm. The cause of this TR was unclear. An evaluation of the right-sided heart pressures was performed under light sedation. The central venous pressure was 23 mmHg - 24 mmHg, RV systolic and diastolic pressures were respectively 34 mmHg and 6 mmHg and the pulmonary artery systolic, diastolic and occlusion pressures were respectively 33 mmHg, 12 mmHg and 20 mmHg. The decision to proceed to a new surgical intervention was thoughtfully discussed between all parties as the patient already had undergone multiple heart surgeries and the OHT was still recent. Based on the TR aggravation and cardiac catheterization results, the decision to repair the tricuspid valve was taken.

The intraoperative TEE that we show here revealed an Ebstein’s-like anomaly on the transplanted heart characterized by a restrictive motion (Video 1) and a downward displacement of the septal leaflet (Video 2).

The anatomical observations during the tricuspid valve repair confirmed the diagnosis (Figure 1). One year after OHT, the patient is clinically doing very well. The most recent TTE showed a good ventricular function associated to a mild to moderate TR.

Discussion

Comment

TR after OHT is not uncommon, with a reported incidence of up to 84%. Its incidence varies strongly based on the definition used, on the surgical techniques performed and on the timing of the diagnosis. Here we will exclusively focus on the immediate TR after OHT. The TR etiology can be divided into two main mechanisms: functional and anatomic [1]. Functional TR is generally accepted as the most frequent cause of TR after OHT.

The echocardiographic features of an Ebstein's anomaly are traditionally defined as a variable degree of tethering of the RV leaflets (septal and posterior) towards the right ventricular free wall and the ventricular septum resulting in a "bubble-like" appearance. These abnormalities of the tricuspid leaflets result in a reduction in the size of the functioning RV and the so called "atrialization" of the RV. It has been suggested that the displacement of the annular hinge of the septal leaflet as compared to the annular hinge of the anterior mitral leaflet as seen in the apical 4-chamber view can be diagnostic. Other echocardiographic features of the Ebstein's anomaly are dilated tricuspid valve annulus, redundant and/or fenestrated anterior leaflet, shortened cordal support, absence of the septal or mural leaflets and dilated RV. An atrial septal defect or a patent foramen ovale is often observed [7].

The conduction system is often abnormal because it is prolonged in the enlarged right atrium. Right bundle branch block is extremely common and this was also the case with our patient. The accessory atrioventricular pathways in Ebstein's anomaly commonly result in supraventricular and ventricular tachyarrhythmias. The timing of the occurrence and the presentation of the symptoms will be dictated by the severity of the disease. The most severe forms will present with heart failure in the neonatal period. Patients with a moderate form may present the first symptoms in adulthood as dyspnea, fatigue and arrhythmias. In the immediate postoperative period of an OHT, a functional TR is clearly the most common mechanism of tricuspid pathology. In this case, the association of an anatomical defect as an Ebstein's-like anomaly to a functional stress caused intraoperatively, such as prolonged ischemic time and donor-recipient size-mismatch, triggered a TR which most likely was not present at the time of donor proposal. As shown in this case, an Ebstein's-like anomaly can possibly be asymptomatic in adulthood and as such undiagnosed in suitable donor hearts. While most of the cardiac abnormalities will be detected during the initial evaluation for donor organ proposal, it is acceptable to consider that an experienced adult cardiologist

would miss a rare and minor form of a congenital heart disease. This case illustrates that, while rare, congenital cardiac defects must be considered when facing an important TR after OHT. Therefore, a thorough intraoperative TEE examination of the TV at the moment of OHT should exclude any acquired and congenital tricuspid malformations. Moreover, a reliable preoperative echocardiography examination should be part of the work-up leading to a transplant free of anatomical malformations.

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