Surgical Management of Isolated Congenital Tricuspid Valve Regurgitation in an Adult

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Abstract

Isolated tricuspid valve regurgitation can be caused by either primary valvular abnormalities or secondary to annular dilation. Congenital tricuspid valve regurgitation in the adult is rare and encompasses a heterogenous group of anatomical variants that is often associated with or is a sequela of other congenital heart diseases that poses a unique challenge to the surgical management of these patients. We present a case of primary isolated congenital tricuspid valve regurgitation and successful tricuspid valve repair in an adult.

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Running Title: Isolated Congenital Tricuspid Regurgitation

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Abstract (79/150)

Isolated tricuspid valve regurgitation can be caused by either primary valvular abnormalities or secondary to annular dilation. Congenital tricuspid valve regurgitation in the adult is rare and encompasses a heterogenous group of anatomical variants that is often associated with or is a sequela of other congenital heart diseases that poses a unique challenge to the surgical management of these patients. We present a case of primary isolated congenital tricuspid valve regurgitation and successful tricuspid valve repair in an adult.

Keywords: congenital heart disease, tricuspid valve disease, tricuspid regurgitation

Introduction

Congenital tricuspid regurgitation (TR) is rare and represents a heterogeneous group of pathologic lesions[1,2]. The clinical presentation of patients with congenital TR varies based on etiology, which can occur in isolation such as Ebstein anomaly, tricuspid valve (TV) dysplasia, and TV cleft, or in association with other

congenital heart defects such as atrioventricular septal defect, tetralogy of Fallot, and pulmonary stenosis with ventricular septal defect[1]. Even rarer, is primary isolated congenital (non-Ebstein) TR. Here, we report successful surgical tricuspid valve repair (TVr) in an asymptomatic adult with severe isolated congenital TR.

Case Presentation

A 43-year-old man with medical history significant for TR was referred to our center for surgical management of his TV following annual transthoracic echocardiogram (TTE) that revealed severe TR with moderately enlarged right atrium and right ventricle (RV). The patient was born in a foreign country and was told by his mother that he was diagnosed with a heart murmur and congenital TR as a child prior to immigrating to the United States. He denied chest trauma, intravenous drug use, history of rheumatic fever, or connective tissue disorder. The patient was an avid hiker and reported occasional palpitations, but was otherwise asymptomatic. In order to further assess TV morphology, the patient underwent 2- and 3-dimensional transesophageal echocardiogram (TEE) and cardiac magnetic resonance imaging (MRI). TEE confirmed severe TR with evidence of a flail anterior TV leaflet and a dilated tricuspid annulus measuring 5.4cm (Figures 1 and 2). The regurgitant jet was eccentric and directed toward the septal wall of the atrium (Figure 3). Cardiac MRI demonstrated a severely dilated RV with normal systolic function and normal left ventricular size with mildly decreased systolic function. Although the patient was asymptomatic, to prevent further RV dilatation and development of right heart failure, the patient was taken to the operating room for elective TVr. Institutional Review Board approval and informed patient consent was waived given the case report nature of the manuscript.

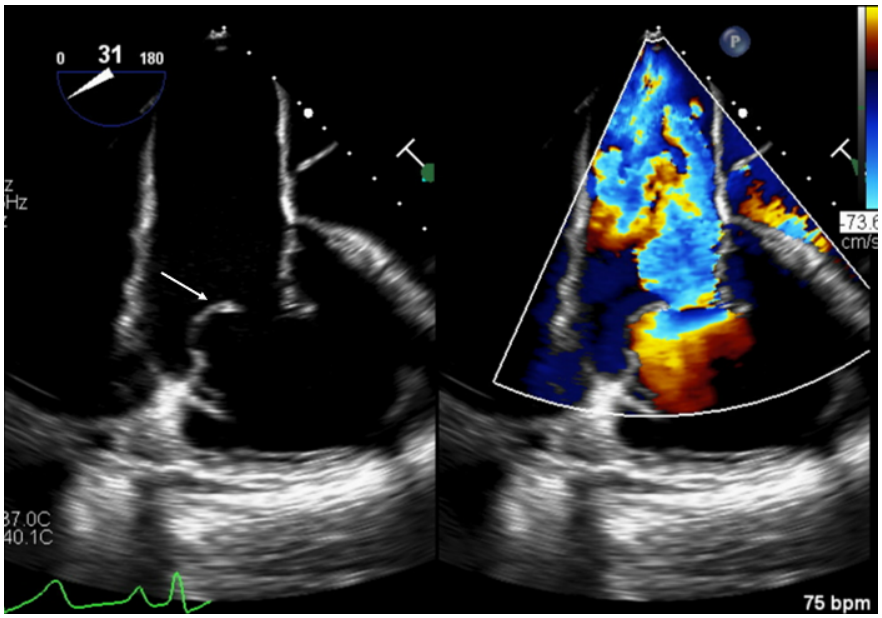
In the operating room, evaluation of the TV revealed a severely dilated annulus. The septal and posterior leaflets were normal in appearance, while the anterior leaflet was redundant with thickened edges. The chordae tendineae of the anterior leaflet appeared to be elongated without evidence of chordal rupture. The papillary muscles appeared to be normal. An Alfieri clover repair was performed. A running 5-0 polypropylene suture was placed between the edges of the anterior, septal, and posterior leaflets to effectively reduce the height of the redundant leaflets. A 32mm Contour 3D (Medtronic, Minneapolis, MN) tricuspid annuloplasty band was added to complete and stabilize the repair. The repair was evaluated by saline filling of the right ventricle, which demonstrated no residual TR. TEE assessment of the valve after weaning from cardiopulmonary bypass confirmed an acceptable repair with mild TR and a mean pressure gradient of 1 mmHg across the valve (Figure 4). The patient had an uneventful postoperative course and was discharged home on postoperative day 3. On follow-up, the patient was doing well with no signs or symptoms of right heart failure.

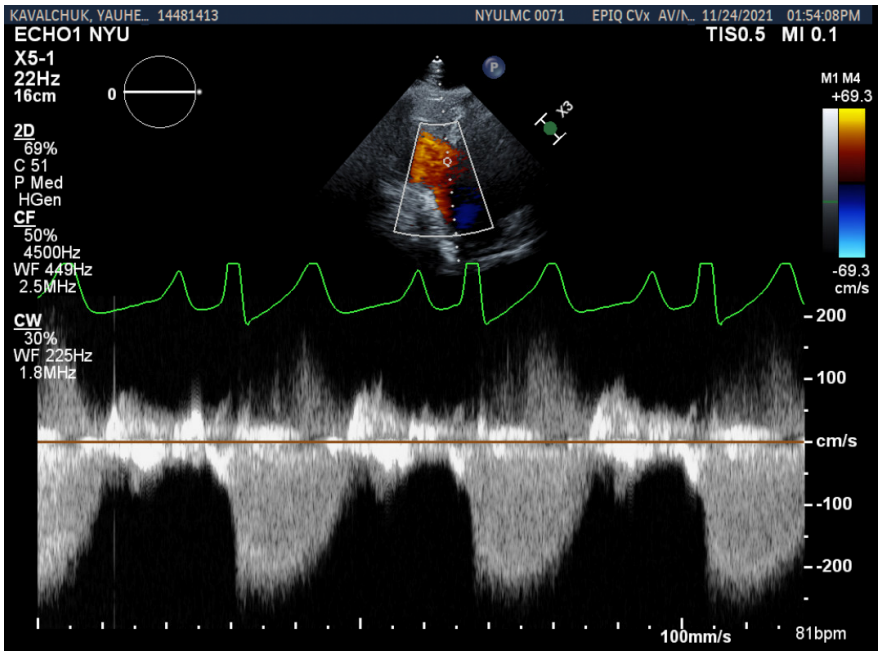
Discussion

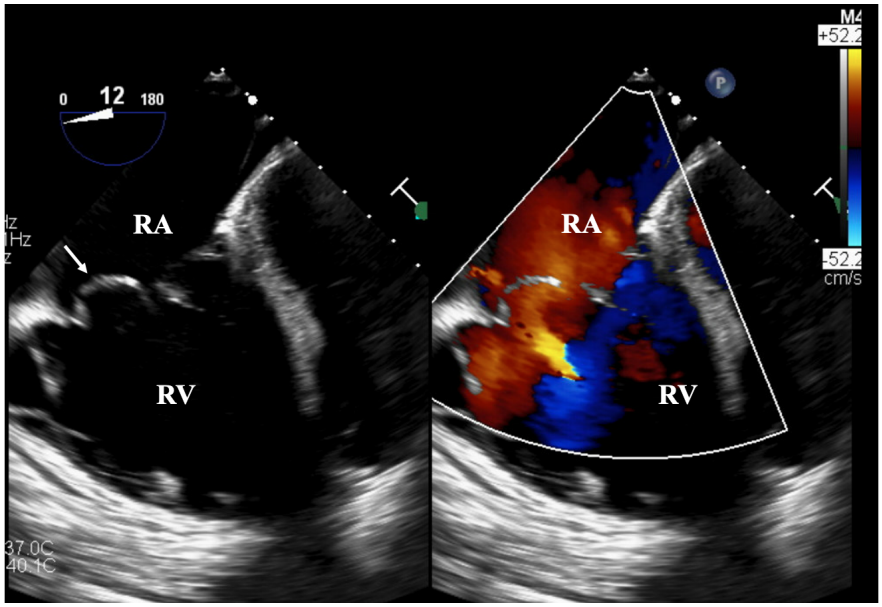
Congenital TV abnormalities resulting in isolated TR are uncommon and often become clinically significant in the setting of other associated congenital heart diseases[3]. TVr is the preferred surgical treatment for TR, particularly in congenital TV disease[4]. Severe TR can precipitate arrythmias and induce RV dilatation, dysfunction, and ultimately right heart failure. Studies have demonstrated that isolated moderate to severe TR is an independent risk factor for morbidity and mortality[5,6]. However, despite this, there is a paucity of outcome studies in this patient population, thus the timing and indications for surgical intervention remain an area of ongoing discussion. Currently, the American College of Cardiology and American Heart Association guidelines recommend TV surgery to be considered in patients with severe primary TR and progressive RV dilation, even in those who are asymptomatic[7,8]. Therefore, while the surgical approach to TVr is challenging and unique to each patient's TV morphology, the overall treatment of TR irrespective of valve pathology has shifted towards an emphasis on early surgical intervention to preserve RV function.

In our case, the patient did not have any additional associated congenital heart diseases and chordal elongation was believed to be responsible for prolapse of the anterior TV leaflet. TVr for congenital TR secondary to chordal abnormalities is challenging. We elected to perform the repair using the Alfieri edge-to-edge method given the patient's valve pathology and safety and feasibility of the technique. In a recently published retrospective study of 237 adult patients with moderate-to-severe TR who underwent TVr using the

edge-to-edge technique, Lee et al. demonstrated favorable long-term outcomes, emphasizing three important advantages of this technique: ease and feasibility, flexibility to accommodate additional approximation stitches if necessary, and ability to be performed without an annuloplasty[9]. One major concern of the edge-to-edge repair is the risk for tricuspid stenosis. In order to mitigate this, we performed a stepwise evaluation of the repair and only when acceptable coaptation was achieved and confirmed with a saline test was the suture tied. Additionally, at the completion of the repair, intraoperative TEE demonstrated a satisfactory mean pressure gradient across the valve of 1 mmHg. In conclusion, isolated congenital TR is extremely rare and highly variable and we advocate for early referral for surgical evaluation and TVr in isolated congenital TR prior to the development of severe RV dysfunction.







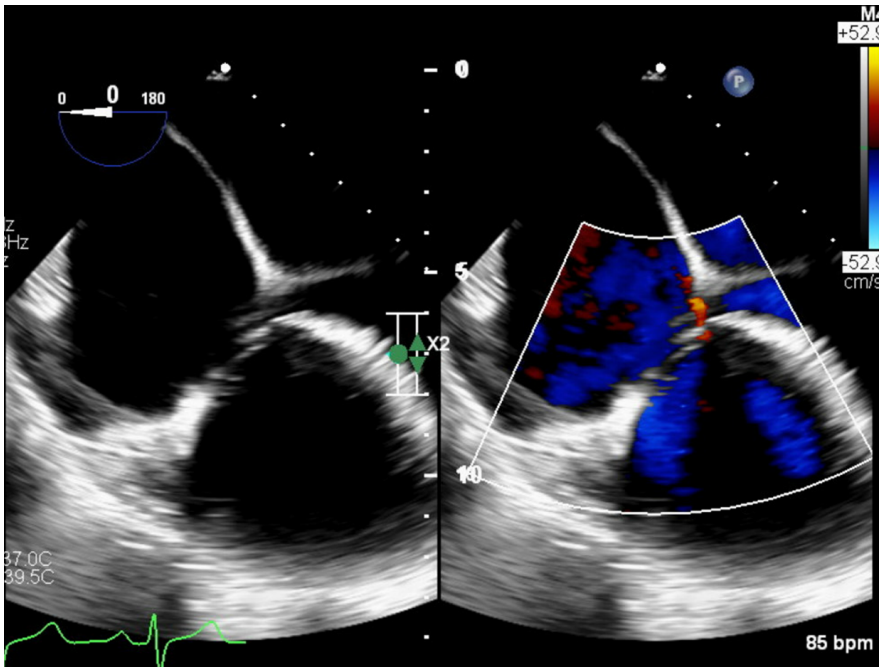


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Figure 1. Transesophageal echocardiogram depicting severe regurgitation and flail anterior leaflet of the tricuspid valve (arrow).

Figure 2. Continuous wave color flow Doppler through the tricuspid valve illustrating the classic triangular “dagger-shaped” Doppler pattern in severe tricuspid regurgitation.

Figure 3. Transesophageal echocardiographic evaluation of severe tricuspid regurgitation demonstrating right atrial and ventricular enlargement with septally directed eccentric regurgitant jet and prolapse of anterior leaflet of the tricuspid valve (arrow). RA, right atrium. RV, right ventricle.

Figure 4. Intraoperative transesophageal echocardiogram following completion of tricuspid valve repair and weaning from cardiopulmonary bypass, depicting mild tricuspid regurgitation.