The Tricuspid Valve in Adult Congenital Heart Disease

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KEYWORDS

• Tricuspid valve • ACHD • Functional tricuspid regurgitation • Annuloplasty • Ebstein anomaly • ASD • VSD

KEY POINTS

- Tricuspid valve disease is a common finding primarily and secondarily in congenital heart disease and tends to be underestimated in its clinical significance.
- The clinical assessment of tricuspid valve disease is subtle but critically important to management.
- Echocardiographic and magnetic resonance imaging assessment of tricuspid valve disease is essential to assess cause, severity, and management options.
- Surgical management of tricuspid valve disease is complex and evolving and can involve multiple repair techniques, or valve replacement.
- More attention in terms of research needs to be focused on the management of tricuspid valve disease in adult congenital heart disease.

Videos of valve repair accompany this article at http://www.heartfailure.theclinics.com/

INTRODUCTION

Tricuspid valve disease is a frequent primary and secondary issue in adult congenital heart disease (ACHD), although its incidence has not been well defined.[1] In the past, tricuspid valve disease has been underappreciated in its importance, although this seems to be changing.[2–5] It has received less attention in terms of research or guidelines. Guidelines for surgical management of tricuspid disease are less aggressive and more subjective than those of other valves.[6] Indications for surgical intervention, and methods of approach and repair, are not uniform across institutions.

These issues also apply to acquired tricuspid disease. However, over the past decade significant progress has been made in establishing the importance of tricuspid valve disease, predominantly tricuspid regurgitation (TR), to patient outcomes in acquired heart disease.[7] In addition, rational approaches to methods of repair have been formed and relative agreement on modes of replacement has been developed. Nevertheless, tricuspid valve repair remains underused in this population.

Rigorous understanding of the contribution of this important lesion to symptoms, morbidity, and mortality is not well established in the ACHD population. Much understanding in this area comes from acquired heart disease. Imaging of tricuspid valve disease is complex and multimodality imaging is often required.[8] It is up to the ACHD community to perform more rigorous and extensive studies to define the extent of the problem and its contribution to disease in this population. Clinical assessment, pathologic entities,

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Heart Failure Clin 10 (2014) 131–153
http://dx.doi.org/10.1016/j.hfc.2013.09.019
1551-7136/14/$ – see front matter

investigations, management (both nonsurgical and surgical) are addressed in this article.

EMBRYOLOGY, ANATOMY, AND RELATIONSHIPS OF THE TRICUSPID VALVE

Embryology

The tricuspid valve forms after the division of the primitive atrioventricular (AV) canal (the connection between the primitive atrium and primitive ventricle) into 2 parts with the ingrowth and fusion of the anterior and posterior endocardial cushions (**Box 1**). As the ventricle enlarges, the tricuspid leaflet tissue delaminates from the wall of the ventricle, along with chordal and papillary muscle apparatus. The endocardial cushions contribute to the formation of the valve, particularly the septal leaflet. In various congenital heart conditions this process occurs abnormally. For example, in the endocardial cushion defects, there is abnormal or no fusion of the anterior and posterior cushions, giving rise to atrial and ventricular septal defects (VSDs) but also abnormal formation of the AV valve tissue. In Ebstein anomaly, delamination of the tricuspid valve from the right ventricle (RV) is abnormal, giving rise to the classic findings of septal and posterior leaflet fusion to the ventricular wall and abnormal excessive chordal attachments.

Anatomy

The tricuspid valve has several distinctive anatomic features (**Box 2**).[9,10] It has 3 leaflets: anterior, posterior, and septal. The anterior leaflet is the largest of these three and usually occupies about 40% of the circumference of the annulus (**Figs. 1** and **2**). The tricuspid valve annulus is displaced more toward the apex than in the mitral valve. The annulus is saddle shaped (highest anteriorly and posteriorly) and slightly ovoid. There are 2 major papillary muscles (anterior and posterior) providing attachment to the anterior and posterior leaflets and the posterior and septal leaflets respectively. There are also septal chordal connections via the muscle of Lancisi (unlike the mitral valve), which provide additional support to the anterior and septal leaflets in the region of the anteroseptal commissure and are vulnerable to injury during surgery that involves the septum, such as patch closure of VSD.

Box 1
Embryology of the tricuspid valve

- Tricuspid valve forms when the primitive common AV valve is divided by anterior and posterior endocardial cushions
- Delaminates from the endomyocardium of the right ventricle (RV)
- Failure of the endocardial cushions to fuse causes AV canal defects
- Failure of delamination from the RV causes Ebstein anomaly

Box 2
Anatomy and relationships of the tricuspid valve

- Tricuspid valve annulus more apically displaced than mitral annulus
- Tricuspid valve has 3 leaflets: anterior (the largest), posterior, and septal
- Two major papillary muscles (anterior and posterior) that arise from the RV free wall, but commonly there are septal chordal attachments (unlike the mitral valve)
- Annulus is saddle shaped (highest points are anterior and posterior)
- The triangle of Koch is immediately above the septal leaflet, with the AV node at its apex, vulnerable to surgery in this area

Relationships

Several important anatomic structures are closely related to the tricuspid valve. The right atrium (RA) in patients with normal situs and D-looping of the ventricles is immediately above the tricuspid valve. The triangle of Koch sits superiorly to the septal leaflet. This anatomic triangle is made up of the coronary sinus ostium at its base, the tendon of Todaro (which is the anterior continuation of the eustachian valve), and the septal leaflet of the tricuspid valve. At its apex lies the AV node, which is susceptible to damage during surgery (**Fig. 3**). Anterior to this structure lies the central fibrous body, penetrated by the His bundle, and further anterior lies the membranous septum. The mitral annulus is leftward of the tricuspid annulus. The aortic valve lies in the space between the two AV valves anteriorly.

CLINICAL ASSESSMENT

History

History in the patient with tricuspid valve disease can be vague (**Box 3**). It is uncommon for patients with significant tricuspid valve disease to complain of shortness of breath. Much more commonly patients with tricuspid disease describe lack of energy and poor exercise capacity as fatigue and