Management of Tricuspid Regurgitation in Patients With Hypoplastic Left Heart Syndrome

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Tricuspid valve (TV) performance is critical for palliation of hypoplastic left heart syndrome. We will review current TV repair techniques, outcomes, and novel approaches.

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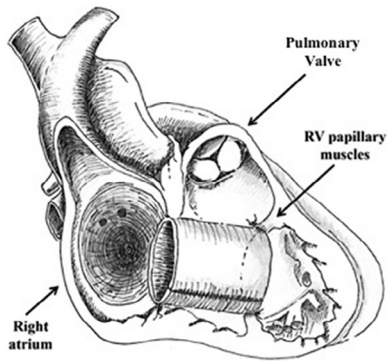
Keywords: HLHS, tricuspid valve regurgitation, tricuspid valve repair, single ventricle tricuspid valve disease



Options for surgical treatment of tricuspid regurgitation in Hypoplastic Left Heart Syndrome.

Central Message

Tricuspid valve performance is critical for palliation of hypoplastic left heart syndrome.

INTRODUCTION

Tricuspid valve regurgitation (TR) requiring surgical treatment occurs in up to 25% of patients with hypoplastic left heart syndrome within 10 years, and has been identified as a risk factor for death.[1,2] Elmi et al have reported that risk factors for eventual tricuspid valve (TV) surgery in hypoplastic left heart syndrome (HLHS) include mitral atresia and longer myocardial ischemia time during the Norwood operation. Shunt type, shunt size, and age at Glenn procedure were not found to be predictive.[1] Takahashi et al, using 2- and 3-dimensional echo techniques, found that moderate TR was associated with increasing patient age, geometric changes in the annulus, leaflet prolapse, and anterior papillary muscle displacement.[3] Because of the importance of all anatomical components contributing to TV function, it is helpful to evaluate the anatomy and function at each level of the TV: leaflet, annular, and subannular.

LEAFLET PATHOLOGY

Stamm et al reviewed the morphology of the TV in 82 patients with HLHS.[4] They demonstrated that TV morphology in these patients differs substantially from normal hearts, and for unclear reasons, this dysplasia was more frequently associated with patency of the mitral valve. Moderate to severe dysplasia of the TV leaflets was identified in 29 patients (35%) of patients. Included were bileaflet TV (10), quadricuspid TV (2), and valves with an accessory TV orifice (2). Leaflet clefts and commissural separation are also common features in regurgitant valves.[5,6]

ANNULAR PATHOLOGY

Pathology at the annular level has been nicely demonstrated by Takahashi et al.[3] As evaluated by 3-dimensional echo, not only are tricuspid annular dimensions significantly increased in patients with HLHS, but other measures of annular geometry, such as annular bending angle and the vertical geometry of the annulus, are also abnormal. Nii et al reported dilated annuli with abnormal TV annular dynamics and attributed this to the lack of interaction of the interventricular septum with a left ventricle.[7]

SUBANNULAR OR PAPILLARY MUSCLE PATHOLOGY

It is increasingly recognized that TV valve function is impacted by ventricular morphology and function. In the analyses by Stamm et al, papillary muscle support of the TV, especially the septal leaflet, was greatly influenced by the degree of development of left heart structures[4] (Fig. 1). Using echocardiography, Takahashi et al identified patients with lateral displacement of the anterior papillary muscle that resulted in the inability of the anterior leaflet to coapt with other leaflet components (so-called tethering), and identified this as an important contributor to TV regurgitation in these patients (Fig. 2).[3]

VALVE ASSESSMENT

It is important to appreciate the contributions of each component: leaflet, annulus, and subannular levels. An appreciation of the functional consequences of valve dysfunction at each level is important when formulating a repair strategy. This process begins preoperatively, and it is important for the surgeon and echocardiographer to participate in a careful echo assessment of the TV, evaluating the anatomy and function of the valve at all 3 levels (annulus, leaflet, papillary muscle) when planning the repair.

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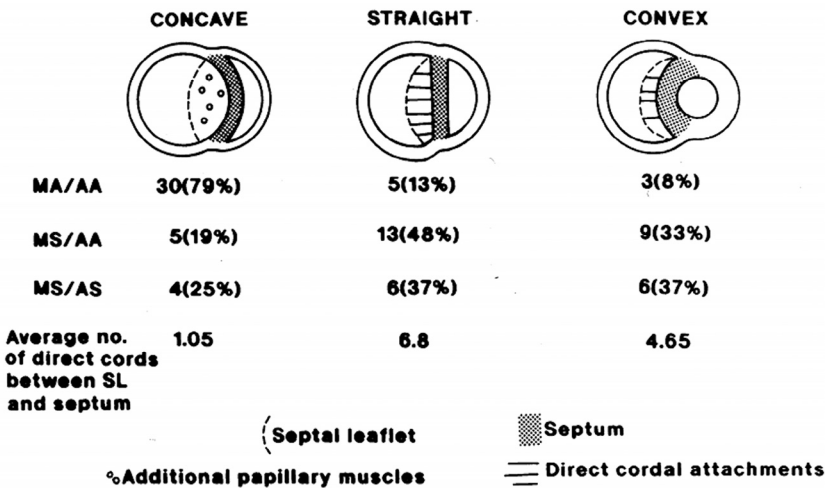

Figure 1. Shape of the ventricular septum in relation to the subtype of HLHS and average number of direct tendinous cords between septal leaflet and septum.

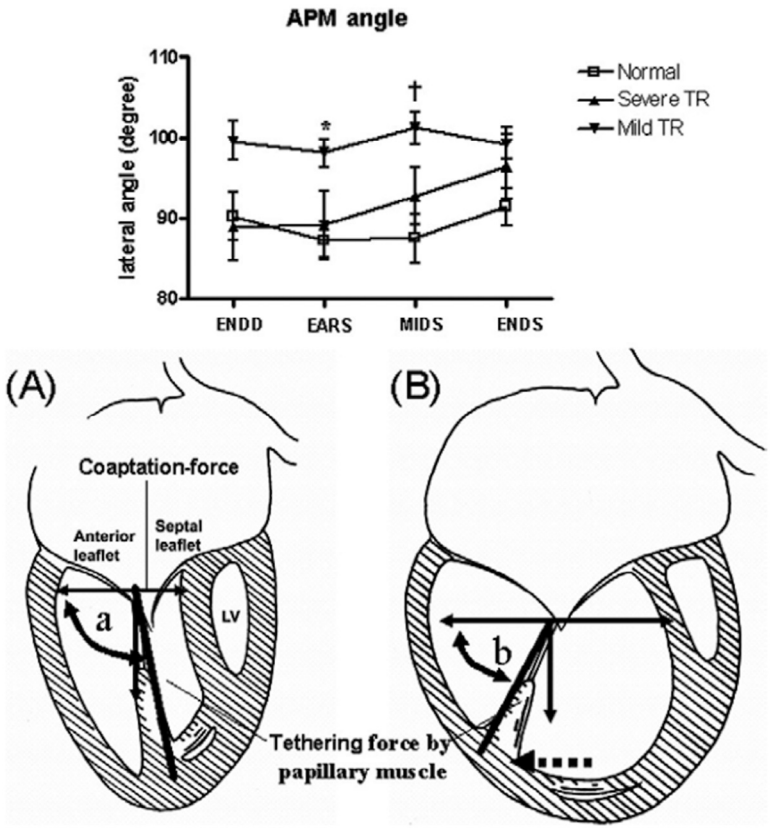

Figure 2. (Top) Anterior papillary muscle (APM) lateral tethering angle at end diastole (ENDD), early systole (EARS), midsystole (MIDS), and end systole (ENDS). Note consistently large lateral angle in patients with mild tricuspid regurgitation (TR). Larger lateral angle tethering force (A) of APM works effectively to provide better coaptation between anterior and posterior and septal leaflet (angle between solid line through papillary muscle and horizontal line with arrows). Conversely, with smaller lateral angle, tethering force works to pull leaflets apart (B), as seen in those cases with significant tricuspid regurgitation and right ventricular dilatation. LV, left ventricle. *EARS: $P < 0.05$, mild TR vs normal. †MIDS: $P < 0.01$, mild vs normal.