Results of surgery for Ebstein anomaly: A multicenter study from the European Congenital Heart Surgeons Association

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Objective: Since most centers' experience with Ebstein anomaly is limited, we sought to analyze the collective experience of participating institutions of the European Congenital Heart Surgeons Association with surgery for this rare malformation.

Methods: The records of all 150 patients (median age 6.4 years) who underwent surgery for Ebstein anomaly in the 13 participating Association centers between January 1992 and January 2005 were reviewed retrospectively. Patients with congenitally corrected transposition were excluded.

Results: Most patients (81%) had Ebstein disease type B or C and significant functional impairment (61% in New York Heart Association class III or IV) and 16% had prior operations. Surgical procedures (n = 179) included valve replacement (n = 60, 33.5%), valve repair (n = 49, 27.3%), 1½ ventricle repair (n = 46, 25.6%), palliative shunt (n = 13, 7.26%), and other complex procedures (n = 11, 6.14%). There were 20 hospital deaths (operative mortality 13.3%) after valve replacement in 5 patients, valve repair in 3, 1½ ventricle repair in 7, palliative procedures in 3, and miscellaneous procedures in 2. Younger age and palliative

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procedures were univariate risk factors for operative death, but only age was an independent predictor on multivariable analysis.

Conclusions: Most patients coming to surgery presented in childhood and were significantly symptomatic. More than half underwent valve replacement or repair, but a considerable proportion had severe disease necessitating 1½ ventricle repair or palliative procedures. Operative mortality did not differ significantly among repair, replacement, and 1½ ventricle repair but was associated with palliative procedures for severe disease early in life, young age being the only independent predictor of operative death.

Abbr eviations and Acronyms
ASD = atrial septal defect
ECHSA = European Congenital Heart Surgeons Association
RV = right ventricle/ventricular
TV = tricuspid valve

Ebstein anomaly, named after the German pathologist who first described it[1] in 1866, is a rare complex congenital defect of the tricuspid valve (TV) and the right ventricle (RV).[2,3] It is characterized by a spectrum of several features, the main ones being (1) apical displacement of the functional tricuspid annulus, that is, of the attachments primarily of the septal and posterior leaflets, with consequent reduction of the functional RV size, (2) dilation of the "atrialized" portion of the RV with variable degrees of wall thinning, (3) adherence of the tricuspid leaflets to the underlying myocardium (failure of delamination) with redundancy, fenestrations, and tethering of the anterior leaflet, and (4) dilation of the true tricuspid annulus (atrioventricular junction). These abnormalities cause tricuspid regurgitation resulting in right atrial and RV dilatation, substrates for the development of atrial and ventricular arrhythmias. An atrial septal defect (ASD) is usually present, allowing right-to-left shunting, resulting in systemic arterial desaturation and cyanosis. In approximately 10% to 15% of patients, accessory conduction pathways with Wolff-Parkinson-White syndrome are encountered.[4] Carpentier and associates[5] have proposed a 4-grade classification of the severity of this anomaly: In type A, the volume of the true RV is adequate. In type B, there is a large atrialized component of the RV, but the anterior leaflet moves freely. In type C, the anterior leaflet is severely restricted in its motion and may also cause significant obstruction of the RV outflow tract. Type D is characterized by almost complete atrialization of the RV with the exception of a small infundibular component. A complementary classification system for Ebstein anomaly has been published by Dearani and Danielson[2] as part of the International Congenital Heart Surgery Nomenclature and Database Project, which was adopted by The Society of Thoracic Surgeons, The European Association for Cardio-Thoracic Surgery, and The European Congenital Heart Surgeons Association (ECHSA). This nomenclature system also describes four types of Ebstein anomaly based on the morphology of the anterosuperior leaflet of the TV and then further subclassifies each of these four types based on the morphology of the leading edge of the anterior leaflet.

The wide anatomic spectrum of Ebstein anomaly is reflected in an extremely variable clinical presentation and natural history ranging from very high mortality despite treatment in symptomatic neonates to long-term survival in some adults with and even without treatment.[4] Accordingly, a large variety of surgical approaches for this anomaly have been used, ranging from valve replacement to various types of valve repair, 1½ ventricle "repair," univentricular palliation, and transplantation. Initially, after the first report of prosthetic valve replacement for Ebstein anomaly in 1963,[6] valve replacement was the mainstay of surgical therapy for this condition, but results were not satisfactory.[7] In 1964, Hardy and colleagues[8] introduced a technique of valve repair which, based on the concept of Hunter and Lillehei,[9] emphasized exclusion of the atrialized portion of the RV by transverse plication, transposing the displaced leaflets to the true annulus and reducing the size of the dilated annulus. As the focus of surgery shifted away from valve replacement, numerous types of repair were devised and refined, and indeed excellent outcomes have been reported from various centers. Danielson and colleagues'[10] modification of the Hardy repair (to which posterior TV annuloplasty and reduction right atrioplasty were added) was reproducibly effective and became a very popular, "standard" repair technique. Carpentier,[5] Chauvaud,[11] and their associates introduced a different repair approach involving temporary detachment of the anterior TV leaflet to achieve its complete mobilization, longitudinal plication of the atrialized RV, clockwise advancement of the anterior TV leaflet (which may be augmented with pericardium),[12] and posterior tricuspid annuloplasty (with prosthetic ring placement), resulting in a monocusp valve. Quaegebeur,[13] Chen,[14] and their colleagues also developed a similar technique, without prosthetic ring annuloplasty. Vargas and coworkers[15] introduced the concept of "annuloplasty" at the level of the displaced leaflets, plicating the atrialized RV above the level of the reconstructed valve. Others (Augustin,[16] Schmidt-Habelmann,[17] Hetzer,[18] Friesen,[19] and their associates) have challenged the value of plication of the atrialized RV and have simplified the repair by performing only a posterior plication of