A valve-in-valve approach for Ebstein’s anomaly

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Introduction: Ebstein’s anomaly

Ebstein’s anomaly is a malformation of the right ventricle and its atrioventricular valve. It is a rather rare congenital defect with an incidence of 1 per 200,000 live births [1]. The anatomical hallmark of this malformation is a downward displacement of the hinge point of the septal and posterior tricuspid leaflets below the annulus due to incomplete delamination of these tricuspid valve leaflets from the underlying myocardium during embryogenesis. Some affected individuals die in utero because of right ventricular dysfunction, others need surgery in early childhood. In a less severe form, Ebstein’s anomaly results in various degrees of tricuspid regurgitation or, rarely, stenosis. Additional interatrial communications are present in 80% to 94% of patients with Ebstein’s anomaly [2] and accessory pathways in approximately 6% to 36% of patients [1]. Medical therapy is used to relieve symptoms of right heart failure or to control arrhythmias, but the majority of patients will require tricuspid valve repair or replacement during their lifetime. The timing of surgical intervention depends on symptoms, exercise performance, heart size, the occurrence of arrhythmias or the presence and consequences of cyanosis due to interatrial right-to-left shunting, eventually complicated by paradoxical embolism [3]. Reconstruction of the tricuspid valve is nowadays the preferred repair technique if feasible. Different surgical techniques are currently used, such as pericardial septal leaflets, cone repairs and leaflet augmentation, in addition to conventional annular plication and sliding leaflet plasties. There are many variants in an Ebsteinoid tricuspid valve, making it important to have more than one repair strategy in a surgical armamentarium [4]. Bioprosthetic valve replacement as opposed to mechanical valve replacement is usually performed in adults not amenable to valvular reconstruction, owing to the perceived lower risk of prosthetic thrombosis and the lack of need for anticoagulation in the absence of atrial arrhythmias. However, bioprosthetic valves are susceptible to leaflet degeneration, necessitating re-do surgery at a given time.

Case report

A mother of twins was diagnosed with Ebstein’s anomaly at the age of 34 years. After an uneventful pregnancy and delivery, she complained about persistent shortness of breath on exercise and undue fatigue. Her symptoms were initially attributed to exhausting family demands. Three years after delivery, she was found to have Ebstein’s anomaly with severe tricuspid regurgitation – the calculated Celemayer index was 0.55 indicating a moderate degree of disease severity. She underwent cardiac surgery two years later as a result of persistent exercise intolerance. She was in New York Heart Association (NYHA) functional class II–III. Reconstruction of the dysplastic tricuspid valve could not be accomplished. A 33-mm stented Perimount bioprosthesis was implanted at the level of the hypothetical tricuspid annulus. The
mean gradient after surgery was 3 mm Hg. Exercise capacity improved substantially after surgery to NYHA I–II. After a period of five years without cardiac complaints, the patient noticed a decline in her exercise capacity. At physical examination, an elevated jugular venous pressure was observed. On cardiac auscultation, however, no systolic or diastolic murmur was heard. Transthoracic echocardiography showed a fusion and severely restricted motion of two of the three bioprosthesis leaflets causing a stenosis with a mean gradient of 10 mm Hg at a heart rate of 67 bpm. There was only mild tricuspid regurgitation. Cardiopulmonary exercise testing revealed a decrease in peak oxygen consumption compared with the last examination one year before. The oxygen pulse curve was suggestive of a cardiac limitation as primary mechanism for the impaired exercise capacity.

She refused to undergo re-do surgery at this time. The option of a percutaneous valve-in-valve procedure was discussed. On transoesophageal three-dimensional echocardiography, the true internal diameter measured 27 mm in diameter (fig. 1). These dimensions were further confirmed with a cardiac computed tomography (CT) scan, suggesting that a percutaneously deployed 29 mm Edwards SAPIEN bioprosthesis could be anchored within the rigid bioprosthetic annulus.

With the aim to delay re-do surgery, the patient decided to opt for a percutaneous tricuspid valve replacement. The procedure was done in a hybrid catheterisation suite with the cardiac surgeon at standby in case the deployed valve embolised into the right ventricle. An Edwards SAPIEN 29 mm bioprosthesis was successfully implanted (fig. 2) via the transfemoral approach and with an Amplatz extra stiff guidewire (placed into the pulmonary artery). The valve was placed with 30%–50% of the stent frame below the tricuspid bioprosthetic valve in order to have an optimal sealing effect. Periprocedural transoesophageal echocardiography showed a reduction of the transvalvular mean gradient from 7 to 1 mm Hg (fig. 3). There was no peri- or transvalvular regurgitation seen after the valve-in-valve implantation. At follow-up three month after intervention, the patient had improved clinically and we measured a mean gradient of 4 mm Hg at a heart rate of 63/min.

**Discussion**

The last 50 years have witnessed dramatic changes in survival of patients with congenital heart disease. Despite all medical advances, many adults with con-
genital heart defects face still multiple re-operations in their lifetime and each operation has inherently an increasingly higher surgical risk [5]. In a survey of patients with congenital heart disease from one of the largest centers worldwide, perioperative mortality constituted 18% of the total mortality burden seen in these patients [6].

In recent years, more and more experience has been gained with transcatheter valve replacement for aortic and pulmonary valves, and in the meantime valve-in-valve procedures have also been established. The Melody transcatheter valve for replacement of degenerated valves in right ventricle-to-pulmonary artery conduits or homografts was the first commercially available percutaneous valve [7]. Currently no established percutaneous options are available to replace a right-sided atrophicventricular valve or native pulmonary valve. The absence of stable anchoring structures at the native atrioventricular level makes primary percutaneous tricuspid valve replacement challenging. However, in a patient already having a bioprosthesis in the tricuspid valve position, experience with limited percutaneous valve replacement has recently been gained. The first procedures were carried out using a Melody transcatheter pulmonary valve [8]. An important limitation of the Melody valve, which is sourced from bovine jugular venous valves, is its maximal diameter. Melody valve competence has been tested and recommended only up to a diameter of 22 mm, even though the Melody valve diameter can be expanded to 24 mm. Its advantage is that the delivery system and usual mounting of the valves for pulmonic interventions is designed for an antegrade delivery of the valve, as opposed to the retrograde access routinely used for percutaneous aortic valve replacement.

Nevertheless, considering the available sizes of the Edwards SAPIEN valve (up to 29 mm), valves initially developed for an aortic position may become the preferred prostheses for valve-in-valve implantation in the tricuspid position also [9]. The Edwards XT valve represents a suitable device to use for this indication, although the reversed crimping on the balloon needs to be done with care in order to avoid leaflet damage. The correct sizing is performed using the internal diameter dimension of the labelled implanted surgical valve plus CT scan measurements. The size of the SAPIEN valve should include an oversizing of 10%–20% for safe anchoring. Beyond doubt, percutaneous tricuspid valve-in-valve implantation is still a rare procedure. To our knowledge, this is only the third case report of a percutaneous aortic valve implanted in a tricuspid bioprosthesis in adults with repaired Ebstein’s anomaly [9, 10].

Conclusions

In summary, percutaneous valve-in-valve procedures with large aortic valves have the potential to reduce the surgical burden in adults with Ebstein’s anomaly and degenerated bioprosthetic tricuspid valves. With this in mind, the limitations of early degeneration of biological valves in the tricuspid position in a young patient population can potentially be reduced. However, this new therapeutic option is still an emerging technique and long-term follow-up is unknown.

Disclosures

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References