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EDITORIAL COMMENT

Branch Pulmonary Valves

Lessons Learned*

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Remarkable progress in the surgical management of complex congenital heart conditions has translated into markedly improved survival, with over 85% of patients now reaching adulthood. In fact, with growth steady at 5% per year, the prevalence of congenital heart disease in the adult population has surpassed that of the pediatric population. Despite this remarkable success, recurrent or residual right ventricular outflow tract (RVOT) dysfunction remains a major problem for this growing population. With the report of a percutaneous pulmonary valve implantation (PPVI) into the RVOT in the year 2000, a nonsurgical treatment option was pioneered (1). This concept developed by Dr. Philipp Bonhoeffer marked the beginning of a new era in cardiovascular medicine, with transcatheter valve repair or replacement techniques becoming a standard of care for some pulmonic, aortic, and mitral valve disease and potentially soon for tricuspid regurgitation. Interestingly, the majority of the surgically treated, remodeled and dysfunctional RVOTs (either native or more so patch- augmented RVOTs) are deemed unsuitable for PPVI with the existing valves due to the size limitation of the Melody (~24 mm) (Medtronic, Minneapolis, Minnesota) and SAPIEN valves (up to 29 mm) (Edwards Lifesciences, Irvine, California). Although this limitation was recognized early on and different percutaneous approaches to large RVOT were developed, 18 years down the line there is still no transcatheter treatment option for these patients to receive a new orthotopic valve outside early clinical feasibility trials. As a consequence, additional, alternative approaches such as branch pulmonary artery (PA) valve implantation are becoming adopted for those with unfavorable RVOT.

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In this issue of *JACC: Cardiovascular Interventions*, Qureshi et al. (2) report the largest multicenter experience of percutaneous, heterotopic pulmonary valve implantation. The authors are to be congratulated for collecting this largest dataset on the topic, which adds to the ongoing discussion on how such patients, unsuitable for PPVI, can be treated nonsurgically. The authors report intermediate-term outcomes of branch pulmonary valves implanted from January 2007 to May 2016 at 13 centers and followed for a median of 2 years.

Fifty of the attempted 52 valves were successfully implanted in 32 patients (mostly adults with comorbid conditions), and 6 procedural complications occurred. There were 5 late deaths noted among the sickest patients, reported to be New York Heart Association functional class III or IV. There was symptomatic improvement with favorable RV remodeling evidenced by reduced RV volumes. The reported outcomes are quite impressive in the context of the advanced symptoms and comorbidities predating the procedure. These results argue that percutaneous branch PA valve implantation is a reasonable option in carefully selected patients, and high absolute risk does not inherently constitute a contraindication to intervention.

In addition to the retrospective nature of the study, there are some limitations that merit attention. The authors propose that the apparently nonuniform reduction in RV volumes indicate remodeling due to reduction in pulmonary

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regurgitation and thereby reduced RV volume overload. However, it should be noted that in nearly 30% of patients, coexisting branch PA stenosis was treated simultaneously with valve implantation. From a pathophysiological point of view, this introduces a confounding factor. With worsening RV function, afterload mismatch may be subtle but significant and not quite readily apparent. The dependence of RV volumes and function on RV afterload rather than preload reduction has been demonstrated repeatedly and in different experimental settings (3,4). With branch PA valvulation, the RV-pulmonary circulation coupling and uncoupling very likely gets metamorphosed to an immensely complex level. The attributable benefit of reducing afterload versus relief from volume overload will remain an open question for patients with heterotopic valves. Regardless of the presence or absence of tricuspid valve (TV) regurgitation, it might have been useful to have accurate PR fraction quantification on cardiac magnetic resonance imaging, both before and after valve implantation, to potentially dissect out the volume-overload and pressure-overload contributions. Due to the small patient numbers, it is unlikely there will be a randomized trial in the future to address this specific question and to understand the impact of affecting volume versus pressure on survival. It admittedly stands to reason that the risks and benefits should be very carefully discussed for patients with higher New York Heart Association functional class in light of the results of this study.

From a technical standpoint, with more than the usual amount of hardware crossing the TV, similar and even stringent precautions as for PPVI, such as flossing the TV with an inflated balloon for initial and subsequent wire positioning, need to be followed to prevent inadvertent TV injury. Branch PA pulmonary valve treatment may reduce, but likely not eliminate the risk of ascending aortic or coronary compression. With such wide variations in the reconstructed RVOT anatomies described, caution should be exercised when dealing with branch PAs arising from a short RVOT. In 1 patient, an Edwards SAPIEN 29-mm valve, which was initially deemed to be unsuitable for orthotopic implantation, was successfully deployed in the RVOT when it migrated down from the branch PA. Planned overexpansion and post-dilatation of Edwards SAPIEN valve up to +5 ml with significant gain over the defined nominal area and successful deployment in the aortic position has been described (5). This may be an option for orthotopic valve implantation, even for large RVOTs up to 31 mm in diameter in some patients. Three-dimensional printing and discussion at a local structural heart team meeting could be very useful.

The durability of the branch valves needs to be assessed over time as it is subjected to nearly one-half the closing volumes of an orthotopically placed valve. The factors that govern branch pulmonary valve closure may include differential lung perfusion, elastic recoil of RV, and the relative degree of loss of stroke work from the dysfunction RVOT that may act like an energy sink. Each branch valve will likely have different closing volumes, pressure, and velocities. Pre- and post-procedural nuclear lung scans or cardiac magnetic resonance may be useful to understand this relationship. With the valves in the branch PAs, it is now expected that RV inflow, dilated body, oversized RVOT, main PA, and the main PA branches proximal to the valve should fill in the given diastolic time frame, leaving some physiological dead space and slow flow. Atrial kick to allow diastolic forward flow into the main PA may play an important role in filling. The relatively thick Edwards SAPIEN valve leaflet designed to close at higher aortic pressures may freeze as noted occasionally when placed in the aortic position (6).

The value of continued longitudinal follow-up of these patients to assess for ongoing RV remodeling is immense. In the future it may be interesting to assess the RV pressure-volume loops and understand the differences in the RV behavior with various loading conditions between orthotopic and heterotopic pulmonary valves. Simple noninvasive assessment of load-independent indices of RV function may be useful to assess periodically. The present study answers several questions and at the same time leaves some questions unanswered regarding pathophysiological consequence of heterotopic valve implantation, patient selection, costs, valve durability with individual lung volumes, and the effect of physiological dead space in the long term. Nevertheless, it gives an additional option to offer while we wait for new-generation valves to be approved for clinical use.

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