JACC: CASE REPORTS VOL. 3, NO. 2, 2021

EDITORIAL COMMENT

Björk-Fontan and the Restoration of Right-Sided Conduits*

Robbert J. de Winter, MD, PhD,[a] Dave R. Koolbergen, MD, PhD[b]

With the development of elaborate surgical techniques for the alleviation of complex congenital heart conditions in neonates and subsequent advances in neonatal, pediatric, and adult cardiology, the prevalence of congenital heart disease nowadays is approximately 1 in 150 in adulthood (1). The right ventricular outflow tract (RVOT) is affected in approximately 20% of newborns with a congenital cardiac abnormality such as tetralogy of Fallot or truncus arteriosus, or pulmonary stenosis, which will require reconstruction techniques with trans-annular patches, conduits, or bioprosthetic valves. More complex conditions like pulmonary atresia with intact ventricular septum or tricuspid atresia used to be palliated with a single chamber circulation with one of the classical Fontan operations, in which extra-anatomic (valved) conduits were used to connect the right atrium to the pulmonary artery or residual right ventricle. Many of these patients will need reintervention later in life, either with surgical or transcatheter procedures, as the durability of bioprosthetic valves and conduits are limited (2). To schedule sequential surgical and transcatheter interventions is now an integral part of lifetime planning in the care of patients with congenital heart disease and requires adaptation of techniques to optimize the options for the next intervention (i.e., develop surgical conduits and bioprosthetic valve implantation techniques that accommodate transcatheter valves and clips that in turn best allow for the next transcatheter or surgical intervention). During the past 3 decades, the operative technique for this population has changed to staged total cavopulmonary connection, with an extracardiac conduit from inferior caval vein to the right pulmonary artery. Until now, these conduits are associated with fewer problems than the classical Fontan, although we must wait until larger groups have reached their middle-aged years.

The options for transcatheter procedures in patients with adult congenital heart disease will greatly increase due to new techniques developed in the structural heart disease space, including new valves, clips, plugs, and sutures (3). Interventional operators treating patients with congenital heart disease may need training in transcatheter aortic valve implantation (TAVI) and MitraClip procedures or at least familiarize themselves with these techniques to gain maximal benefit from currently available transcatheter options. Vice versa, many case reports have reported the use of closure devices and vascular plugs, routinely used in the pediatric and congenital space, as an add-on to MitraClip placement or as an option to treat paravalvular leak or shunts after TAVI. Ideally, programs in congenital and structural heart disease interventions are developed in synchrony together with surgeons and interventionalists in tertiary referral centers to optimize care and drive future developments.

To deliver such care, we must train the workforce that is needed in the near future. In the recently published Society for Cardiovascular Angiography and Interventions position paper on adult congenital cardiac interventional training, competencies and organizational recommendations, the need for both interventional and conceptual expertise is emphasized: applicants are required to complete an interventional cardiology fellowship and an adult

*Editorials published in *JACC: Case Reports* reflect the views of the authors and do not necessarily represent the views of *JACC: Case Reports* or the American College of Cardiology.

From the [a]Department of Cardiology, Amsterdam University Medical Centers, Amsterdam, the Netherlands; and the [b]Department of Cardiothoracic Surgery, Amsterdam University Medical Centers, Amsterdam, the Netherlands.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2020.12.018

structural fellowship that will provide the necessary skillset, but in addition need ample exposure to complex adult congenital heart disease conditions and interventions (4).

In this issue of *JACC: Case Reports*, Schamroth Pravda et al. (5) describe a very interesting case that superbly illustrates what is possible nowadays when imaging specialists, interventional radiologists, and congenital and interventional cardiologists work together to devise a strategy to treat complex anatomy with transcatheter techniques. Many pediatric cardiac surgical procedures from the past, such as atrial switch, according to Mustard for transposition of the great arteries or Björk-Fontan (5)for hypoplastic right ventricles are no longer considered optimal. Yet, patients who underwent these procedures reach adulthood with failing "legacy cardiac anatomy" that require creative solutions.

The authors used 3-dimensional printing techniques to assess the anatomy of a failing Björk-Fontan conduit (connecting the right atrium to the pulmonary artery) (6) in a patient considered very high risk for surgery. A custom-made stent graft was implanted, followed by 2 more endografts to construct a smooth neo-conduit from the inferior vena cava to the pulmonary artery. The Edwards Alterra device was implanted to reduce the dimensions of the conduit to such a degree that it could accommodate the placement of an Edwards Sapien transcatheter valve. There was no obstruction of the hepatic veins just caudal to the neo-conduit, and the Alterra provided an unobstructed entrance to the valve, which was competent, delivering pulsatile flow to the pulmonary circulation. The authors are to be commended with designing, preparing, and skillfully executing such a technical masterpiece.

One question that remains is whether the cardiac venous return from the coronary sinus will remain patent. The authors checked patency with transesophageal echocardiogram at the time of stent implantation, but long-term patency remains to be established.

Just recently, the experience with the first 15 patients treated with the Alterra device has been reported in *JACC: Cardiovascular Interventions* (7), demonstrating safety and feasibility, 100% procedural success, and promising 6-month outcomes. Patients weighed at least 20 kg, had dysfunctional RVOT/pulmonary valve (PV), and RVOT/PV dimension 27 to 38 mm and at least 35 mm of length that could accommodate the Alterra device as a landing zone for a 29-mm Edwards Sapien 3 transcatheter heart valve. Careful patient selection and preparation included constructing 3-dimensional physical models of the RVOT at peak systole and diastole for each patient.

Of course, long-term durability of complex stent-spacer-valve constructs, such as beautifully illustrated in this case, is uncertain. Therefore, such "experimental" procedures should be reserved for patients with very high risk of (repeat) cardiac surgery. In addition, this patient is likely to survive to a point in which she requires a valve-in-valve procedure or repeat surgical procedure when the newly implanted bioprosthesis starts to fail. Moreover, there may be an unknown but sizeable risk of infective endocarditis as with all bioprostheses in the pulmonary position.

We are entering an exciting time of increased possibilities with new technologies that provide minimally invasive and transcatheter options for patients with congenital heart disease. Transcatheter heart valve replacement is now feasible in all 4 positions and new-generation valves, conduits, and clips will expand our armamentarium to the benefit of our patients. The total volume of the market in the adult structural heart disease will provide the financial impetus for manufacturers to continue the development of new products that ultimately become available for the benefit of our patients.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Prof. Dr. Robbert J. de Winter, Department of Cardiology, B2-137, Amsterdam University Medical Centers, Meibergdreef 9, 1105AZ Amsterdam, the Netherlands. E-mail: r.j.dewinter@amsterdamumc.nl.

REFERENCES

1. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation 2007;115:163–72.

2. Verheugt CL, Uiterwaal CS, van der Velde ET, et al. Mortality in adult congenital heart disease. Eur Heart J 2010;31:1220–9.

3. Ghobrial J, Aboulhosn J. Transcatheter valve replacement in congenital heart disease: the present and the future. Heart 2018;104:1629–36.

4. Daniels CJ, Ghobrial J, Horlick E, et al. SCAI position statement on adult congenital cardiac interventional training, competencies and organizational recommendations. Catheter Cardiovasc Interv 2020;96:643–50.

5. Pravda NS, Kornowski R, Assa HV, et al. Complex catheter-based structural heart reconstruction in a patient with tricuspid atresia and Björk palliative conduit. J Am Coll Cardiol Case Rep 2021;3:212–6.

6. Björk VO, Olin CL, Bjarke BB, Thorén CA. Right atrial-right ventricular anastomosis for correction of tricuspid atresia. J Thorac Cardiovasc Surg 1979;77:452–8.

7. Shahanavaz S, Balzer D, Babaliaros V, et al. Alterra adaptive prestent and SAPIEN 3 THV for congenital pulmonic valve dysfunction: an early feasibility study. J Am Coll Cardiol Intv 2020;13:2510–24.

KEY WORDS congenital heart defect, imaging, stents, valve repair