Editorial

The Fontan extracardiac conduit: one size does not fit all

Frank Cetta[1], Harold M. Burkhart[2]

[1]Division of Pediatric Cardiology, Department of Cardiovascular Diseases, Mayo Clinic, Rochester, MN, USA; [2]Division of Thoracic and Cardiovascular Surgery, University of Oklahoma Health Sciences Center, Oklahoma City, OK, USA

Correspondence to: Harold M. Burkhart, MD. Division of Thoracic and Cardiovascular Surgery, University of Oklahoma Health Sciences Center, 800 Stanton L. Young Blvd, Oklahoma City, OK 73104, USA. Email: harold-burkhart@ouhsc.edu.

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The size of all components of the Fontan circuit need to be adequate in order to produce sustainable palliation for patients with functional single ventricle physiology. It is well established that patients with undersized pulmonary arteries (low Nakata index) usually do not fare well in the long-term with Fontan physiology. Assessment of flow hemodynamics with MRI substantiate this concept (1). Similarly, stenoses of the Fontan conduit may become hemodynamically important even when there is only a 1–2 mmHg gradient. With these observations in mind, one reads with great interest the recent article by Cho *et al.* in the *European Journal of Cardiothoracic Surgery* (2) in which they describe the outcomes for young patients who received 16 mm polytetrafluoroethylene (PTFE) tubes for extracardiac conduit (ECC) Fontan procedures.

The authors evaluated 66 patients who received a 16 mm PTFE ECC. They compared this patient cohort to patients with similar body weight and age who received larger conduits. Interestingly, they were not matched according to body height. This was a cohort of patients who had Fontan operation at a relatively young age (mean age in both groups was only 2.9 and 3.1 years respectively). Patient outcomes for the following ten years were evaluated. Conduit-related events were significantly lower in the smaller conduit group versus the larger conduit group. Similarly, Dabal *et al.* (3) have reported excellent results and a freedom from reoperation of greater than 90% when utilizing a 16 mm ECC.

The author's cite that the typical inferior vena cava in an adult is 18–20 mm. This makes the rationale for using a small tube somewhat controversial. They postulate that by using a 16 mm tube, the risk of thrombus due to sluggish Fontan follow is reduced since an 18–20 mm tube in a 3-year-old would be quite large. They noted gradients of 1–2 mmHg in the 16 mm tubes during the follow-up period. The authors have previously shown that during a 3-year follow-up period ECC cross-sectional area decreases by 14% (4), a similar decrease in ECC cross-sectional area over time was noted by other authors (5). Thrombosis of the Fontan conduit is an important but fortunately rare event. Admittedly, it may be under-recognized if the only imaging modality utilized is transthoracic echocardiography.

While the smaller 16 mm ECC makes sense if one assumes that the patient will develop into a smaller adult, it is difficult to know if this strategy should be applied to all patients at a young age. Furthermore, this study did not evaluate the very important teenage growth spurt when patients with small a ECC may develop Fontan stenosis. In addition, if one waited until patients were at least 4 years old before performing a Fontan operation, one might make the argument that the ECC could be larger (18–22 mm).

However, use of the "bigger is better" philosophy may not be accurate either. Itatani *et al.* showed that flow dynamics in patients may better with 16 and 18 mm ECC than in patients with larger Fontan tubes (6). They demonstrated increased lateral conduit stagnation in larger conduits that could increase the chance of thrombus formation. But Fontan flow is dependent on several factors including: adequate respiratory mechanics, low ventricular

filling pressure and relatively low resistance to flow through the lung bed. It would be interesting to test the authors' theories regarding optimal flow properties in smaller ECC with state of the art MRI flow propagation mapping. In addition, they point out that the flow characteristics of a larger conduit in adult size patients may be optimal and have yet to be evaluated.

An interesting factor that the authors did not evaluate was the size of the parents of these patients. If children usually grow to the average size of their parents, perhaps that could be used to help differentiate those patients who at age 3 years should receive smaller versus larger ECC? It may be patient length rather than body weight that determines adequacy of the 16 mm ECC. A smaller ECC would intuitively seem to need increased intervention. Percutaneous stenting of the PTFE tube, if it is undersized along its entire length, will usually not yield satisfactory results.

Since Fontan associated liver disease (FALD) has become an increasingly recognized problem in these patients (7), it would seem that the teenager and young adult with a 16 mm Fontan tube may be at higher risk of developing elevated hepatic pressure and passive congestion. While the authors may speculate that the risk of thrombosis will be lower with smaller tubes, the long-term negative impact of a small ECC may be more detrimental for patients if this strategy is generalized to the entire Fontan population.

The surgical philosophy that the "right patient needs to have the right surgery at the right time" should also include, and the "right size ECC" for patients needing Fontan operation. One worries that although *in situ* Fontan conduit thrombosis may be diminished with smaller ECC tubes, will FALD be worse? A good scientific study usually raises as many questions as it solves. Is a smaller conduit sufficient through adulthood? Do we need to delay the Fontan, if possible, to allow for a bigger conduit? Or are there patients that are best treated with an appropriate smaller, efficient conduit initially and reassessed for a larger conduit when they grow to a certain size? The authors are commended for their thought provoking study.

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Footnote

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