Authors have nothing to disclose with regard to commercial support.

REPLY: THERE IS NO “ONE-SIZE-FITS-ALL” IN FONTAN SURGERY!

Reply to the Editor:

We thank the authors for their kind review of our work. The authors present an important point related to patients with abnormal ventricular positioning. Our data suggest that early fenestration closure leads to inferior clinical outcome in patients who undergo the Fontan procedure. Furthermore, we found a significantly higher rate of early fenestration closures in patients undergoing extracardiac conduit (ECC) compared with the lateral tunnel (LT) Fontan technique. The question still remains if the early fenestration closure occurred because of a less favorable hemodynamic situation starting off with or because of inferior postoperative hemodynamics in the ECC group. Looking at the baseline characteristics of our Fontan cohort, it revealed that actually the ECC group showed more favorable preoperative hemodynamics with lower pulmonary artery pressure and pulmonary vascular resistance and a higher rate of left dominant ventricular morphology supporting the systemic circulation. However, the ECC group also showed a higher rate of heterotaxy compared with the LT group (16.6% vs 8.3%).¹ Patients with single ventricular morphology requiring a Fontan circulation have diverse, often complex anatomic diagnoses. By directing the systemic venous blood flow passively to the pulmonary circulation, the Fontan circulation is highly dependent on favorable flow hemodynamics and minimal energy loss. Malpositioning of the heart with apicocaval juxtaposition and heterotaxy syndrome with abnormalities of systemic or pulmonary venous connection are known to complicate a Fontan procedure and attribute to a higher early mortality in this group.² However, the “ideal” Fontan pathway is still controversial. Considerations include risk of flow maldistribution (right-left) related to orientation of the tunnel, risk of pulmonary vein compression from the graft, and available volume of atrial cavity for LT. We believe that computational techniques to predict anatomic lie and

3-dimensional hydrodynamic flow studies to predict flow distribution and energy loss will be critical in making the choice of Fontan type in this population.³ For the same reason, many modifications of the ECC have been suggested to increase the offset distance between the inferior and superior vena cava to avoid head collision of blood flows.⁴ Flow simulations in patients with complex anatomy such as apicocaval juxtaposition revealed that a straighter and shorter conduit on the ventricular apical side provides better laminar blood flow with less energy loss.⁵ All of this is still speculation, and to answer the question what Fontan technique is more suitable for patients with complex anatomy will require further, ideally prospective studies. Clearly, Fontan surgery in patients with abnormal ventricular anatomy remains challenging and requires individualized planning.

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<https://doi.org/10.1016/j.jtcvs.2020.02.098>