Reviewer's report

Title: Extracardiac Fontan with T-shape conduit in non-confluent pulmonary arteries

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Reviewer: Davis c Drinkwater, Jr.

Reviewer's report:

The authors report on a completed fenestrated extracardiac Fontan in a 3 year old patient with single ventricle non confluent pulmonary arteries using PTFE graft to replace intrepericardial PAs and simultaneously connect IVC to the PA. At the first operation at 4 weeks of age, the decision made by the authors was to use PTFE 6 mm replacement and 4 mm BT shunt. This first operation set in motion a unique approach, using PTFE graft with subsequent upsizing at each procedure, rather than unifocalizing the true PAs. With dissection and mobilization of the branch PAs, while this is generally possible and likely the better longterm option, the authors felt this was not the case. Their use of increasing sizes of PTFE graft, including a composite T graft creation prior to cardiopulmonary bypass is creative and apparently succesful on the part of the authors. The first question has to be what precluded a full bilateral mobilization and unifocalization in combination with a central PA shunt at the first stage? Certainly later in life this mobilization is more difficult and it is understandable that subsequent procedures would involve further PTFE interposition graft. As selection of prosthetic material, was ringed PTFE graft considered to prevent extrinsic compression by a presumably large ascending aorta? At 32 months, it is not clear following a cath demonstrating 11 mm Hg mean gradient at the Glenn anastomosis, that a repair or attempted catheter-based intervention was carried out in addition to collaterals being embolized. Would the authors please clarify by describing their thoughts in particular if nothing was carried out in the face of SVC stenosis and apparent preferential flow to the left lung on angiogram? To protect against clot formation in the PTFE graft, was antiplatelet therapy only used or was coumadin employed at any time before completion-Fontan? The outcome of the patient seems excellent, however a chronic concern that central PAs may need subsequent replacement surgeries points out the potential benefits of native unifocation and subsequent long term growth potential. In point of fact, a 12 mm PTFE graft if remaining fully patent will likely suffice long term. The experience with central pulmonary artery reconstruction using graft material in biventricular repairs such as PA VSD patients would suggest that 12 mm is adequate in size. However, the laminar flow requirements in Fontan (venous-driven) physiology may warrant the author’s concern about sizing particularly if there is any intimal or clot build up over time. Would the authors add a brief comment on this aspect in their discussion section? Finally, the pericardial cuff-fenestration is interesting. Presumably a circumferential anastomosis is made around an atriotomy site. One potential concern is the skirt-anastomosis might create a separate cavity.
which could be an area for clot development. Would the authors kindly decribe
the actual penetration completion and whether coumadin is considered
mandatory with this technique?

The authors will need to address grammatical issues to clarify what is otherwise
a very interesting case report. Major Compulsory Revisions (that the author must
respond to before a decision on publication can be reached)

Minor Essential Revisions (such as missing labels on figures, or the wrong use of
a term, which the author can be trusted to correct)

Discretionary Revisions (which the author can choose to ignore)

What next?: Unable to decide on acceptance or rejection until the authors have
responded to the major compulsory revisions

Level of interest: An article whose findings are important to those with closely
related research interests

Quality of written English: Needs some language corrections before being
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Statistical review: No, the manuscript does not need to be seen by a
statistician.