Fontan Fenestration and the Role of the Covered Stent

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Abstract

Key Points : * Interventional therapies directed at fenestration closure in the Fontan patient must rely on good hemodynamic data * The Large Optimus-CVSTM stent is an additional armamentarium for fenestration closure however, longer term follow up is needed * Multi institutional studies defining the long-term benefits of fenestration closure and outlining fenestration management guidelines may help improve the long-term morbidity and mortality in this group of patients.

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Key Points :

- Interventional therapies directed at fenestration closure in the Fontan patient must rely on good hemodynamic data
- The Large Optimus-CVS^{TM} stent is an additional armamentarium for fenestration closure however, longer term follow up is needed

- Multi institutional studies defining the long-term benefits of fenestration closure and outlining fenestration management guidelines may help improve the long-term morbidity and mortality in this group of patients.

Fontan and Baudet (1) in 1971 first reported the repair of tricuspid atresia with a surgical procedure now known as "Fontan repair". The Fontan repair has undergone multiple modifications (2-8) to optimize the physiologic and anatomic factors that play a vital role in the Fontan circulation. In 1988 Laks et al (7) described the creation of an adjustable fenestration which in the post-operative period allowed controlled right to left shunting allowing for improved cardiac output and decreasing the incidence of post-operative pleural effusions. The benefit of the fenestration comes at the cost of a lower systemic arterial saturation and the increased risk of stroke. Closure of the fenestration spontaneously or intentionally improves the arterial saturation and may reduce the risk of thromboembolic events (9).

Haddad et al present a case series of three patients with total cavo-pulmonary connection with an extracardiac interposition Gore-Tex graft (W.L. Gore & Associates, Inc., Flagstaff, AZ, USA) of 16 or 18mm with a 5-mm punch-hole-type fenestration. After hemodynamic evaluation the patients underwent fenestration closure with a 43mm/Large Optimus-CVS^{TM} stent crimped over a Altosa-XL^{TM}PTA balloon catheter (AndraTec GmbH, Koblenz, Germany) and implanted through a 10 Fr delivery sheath. The stent was placed across the fenestration in the extra cardiac Gor-tex graft. At follow up immediately and at one month there was no residual shunt across the fenestration. The Optimus-CVS^{TM} is a balloon-expandable stent with a HD-Cobalt-Chromium-MP35N alloy frame that is flexible because of the hybrid cell design and has excellent radial strength. The PTFE-cover on the stent has a unique end-free sealing design which minimizes the risk of overhanging. The Altosa-XL^{TM} PTA balloon catheter is a high pressure low profile balloon catheter which may allow for a smaller sheath size to deliver the stent making it particularly attractive in children.

There is currently no single ideal device for fenestration closure and to some extent the selection of the device is dictated by the size and location of the fenestration and operator preference. Several devices have been used in fenestration closure including occluder devices and covered stents (10-12). The occluder devices and the stents have their merits and drawbacks. Covered stents are effective in closing fenestrations with good mid-term results (11). The immediate results for fenestration closure with the Large Optimus-CVS^{TM}stent is promising. Long term follow-up in a larger cohort of patients is needed to better assess the efficacy and safety of this stent in closing Fontan fenestrations. Customized covered biodegradable stents may prove a good alternative to the traditional stents.

The overwhelming unknown factor remains the selection of patients who would benefit long term from fenestration closure. Hijazi et al (13) in an elegant study demonstrated that immediately following balloon occlusion of the fenestration there was an immediate increase in aortic saturation with decreased cardiac output. The increase in aortic saturation did not fully compensate for the decrease in cardiac output resulting in a decrease in systemic oxygen delivery and an increase in oxygen extraction in the vast majority of patients. In addition, short term follow-up studies following fenestration closure (14-15) demonstrate an increase in the arterial oxygen saturation but not greater event free survival. Multi institutional studies defining the long-term benefits of fenestration closure and outlining fenestration management guidelines may help improve the long-term morbidity and mortality in this group of patients.

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