

An expanded view of congenital heart disease

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Congenital heart disease is generally perceived as being characterized by a single or collection of anomalies of the heart and great vessels. This view leads to the impression that the complications will be local or regional. The reviews by Dr. Yoo and Dr. Slesnick challenge this orthodoxy for patients with complex congenital heart disease. Dr. Yoo notes that children live in a constant state of systemic hypertension following Fontan palliation that adversely affects multiple organ systems. The liver is particularly susceptible to the complications of systemic venous hypertension since the pressure gradient between the portal vein and hepatic veins is so much smaller than systemic arterial to systemic vein pressure gradients. Other factors further complicate gastrointestinal and hepatic flow. Hepatic damage is universal and in severe cases is life threatening. Cirrhosis and hepatocellular carcinoma can follow Fontan palliation. The etiologically more poorly understood protein losing enteropathy carries a high mortality as well. Cardiovascular imagers must expand their perceptions of congenital heart disease and actively search for evolving gastrointestinal complications of Fontan procedures. Dr. Yoo has pointed out specifically the important role magnetic resonance imaging can play in this regard.

Dr. Slesnick has pointed out how new technology can improve the energy efficiency of the ever evolving Fontan

palliation. More energy efficient Fontan connections improve flow and ameliorate or at least delay the complications of systemic pulmonary venous hypertension noted above. The evolution of Fontan configurations from atrio-pulmonary to total cavopulmonary connection with anastomotic offsets and caval flaring has improved flow dynamics, but optimization requires individualized modeling. Computational flow dynamics allows for modeling different potential Fontan conduits and configurations prior to surgery. Modeling techniques combine morphologic and flow data that is moving from a slice orientation to a volume approach. The current fashion in medicine is patient centered medicine. The development of Fontan conduits specifically designed for individual patients to reduce energy losses and optimize the distribution of hepatic venous flow may be a high water mark in patient centered care. Perhaps 3D printers in the future will manufacture individualized Fontan conduits.

Over time, cross-sectional imaging has evolved from a slice approach to a more organic volume approach. Our approach towards determining the optimal configuration of Fontan conduits will follow along the same path. Similarly, we must expand our view of congenital heart disease away from being a localized condition to a more systemic illness.

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