

**Patient/Family Lay Summary:** FON Case Review Conference September 2022

**Coordinated by:** Cincinnati Children's Hospital Medical Center

**Topic:** Fontan Associated Liver Disease

**The Problem:** We focus on the Fontan liver as a ‘diseased organ’. Liver fibrosis is ubiquitous, but decompensated cirrhosis is rare. There is little correlation between the severity of heart disease and liver fibrosis. And some ‘liver issues’ (e.g., ascites – free fluid in the abdomen) may or may not be related to liver fibrosis. Liver complications can be devastating (e.g., liver cancer) or otherwise important (e.g., vasodilation, lung arteriovenous malformations).

 **Clinical Course of the Cases Presented:**

* **Case 1**: Our patient is an 18-year-old man with hypoplastic left heart syndrome. He underwent the Norwood procedure at 2 days of age; bidirectional Glenn at 6 months and extracardiac fenestrated Fontan (20 mm conduit) at three years. His only medication is a baby aspirin. He is asymptomatic (though he doesn’t do very much!). He has had no prior liver evaluation.
* Case 1 Findings and Course: Liver elastography identified a very stiff liver, leading to further evaluation showing a narrowed Fontan pathway. Stiffness was improved with dilation and stenting of the narrowing.
* Case 1 Lessons: Liver stiffness is a function of liver histology (e.g., fibrosis) + congestion (fluid). A finding of elevated stiffness raised suspicion for Fontan pathway obstruction, in the context of limited cardiovascular imaging. Alleviating Fontan pathway impedance to flow may facilitate hepatic decongestion, and thereby delay progression of liver disease.
* **Case 2**:Our patient is a 13-year-old girl with double inlet left ventricle + left AVV atresia, s/p 20 mm extracardiac fenestrated Fontan. She has a history of protein losing enteropathy. This is resolved after surgical enlargement of a restrictive ASD. She has normal ventricular function and excellent hemodynamics. She has her first liver imaging at age 13 years.
* Case 2 Findings and Summary: Liver imaging showed liver masses. Longitudinal imaging showed growth and with biopsy liver cancer was diagnosed.
* Case 2 Lessons: Serial imaging, preferably with MRI, is essential to evaluate changes in liver lesions and determine timing for biopsy. Long transplant wait times emphasize the importance of early detection and temporizing treatment. Combined liver transplant is the only curative option for some individuals, performed in conjunction with heart transplant.

 **Important Points, Lessons Learned, and Potential Solutions:**

* Liver assessment provides not only insight into liver disease, but also a window into Fontan hemodynamic status.
* Liver cancer screening is an integral part of Fontan care. There is little evidence that other liver evaluation, such as surveillance liver biopsy provides a benefit, however.
* The path from HCC diagnosis to transplant may not be straightforward, and early diagnosis is critical.