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Intrahepatic Transcriptomics Discriminate Advanced Fibrosis in Patients with the Fontan Circulation

The most complex forms of congenital heart disease include having a single chamber to pump blood to the rest of the body. These patients are at risk for dying in infancy, but having a Fontan surgery can help them survive into adulthood. In this surgery, the venous drainage is re-routed away from the heart, so blood goes directly to the lungs. This circuit, though lifesaving, has consequences on many organs and almost all patients with this problem will develop stiffening of the liver. The degree of stiffening could vary, and, in some cases, it could be so severe that a liver transplant is required. Unfortunately, current imaging and laboratory techniques to assess the stiffening do not correlate with what is seen after taking a liver sample. Thus, in this proposal, we aim to characterize the molecular pathways of advanced liver stiffening and adverse outcomes in patients with the Fontan procedure. This may help design future noninvasive markers and therapeutic strategies in this population.