Title of Project: Characterization of Hepatic Dysfunction in Patients following the Fontan Operation

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The Fontan operation, widely used in infants and children with univentricular congenital heart disease, circumvents the requirement for the right ventricle by providing blood flow directly and passively into the pulmonary circulation [1]. Survival has drastically improved over the years with recent studies showing 90% survival at 10 years post surgery [2,3]. As survival continues to improve, new complications are emerging due to the abnormal physiology of the Fontan circulation [4]. One of these complications is progressive hepatic congestion leading to hepatic fibrosis, eventual cirrhosis, and in some cases hepatic malignancy [5,6]. This unrecognized liver dysfunction places the patient at risk for bleeding, ascites, compromised immune status and can limit cardiac treatment options, including heart transplant if needed. This process begins in childhood, but it is not known at what point these changes develop [7]. Liver biopsy, the standard for diagnosis, has many risks in this population, and is not routinely used to screen for the development of liver disease. As the disease mechanism has not been characterized, neither therapies nor non-invasive screening protocols have been established.

The purpose of this pilot study is to characterize the degree of hepatic fibrosis in pediatric patients following the Fontan procedure by magnetic resonance imaging (MRI). MRI has been used to characterize fibrosis in other hepatic diseases. In addition, we will assess whether decreased cardiac output and alteration in hepatic biomarkers correlate with the degree of hepatic disease.