

Adam M. Lubert, MD

Assistant Professor

Adult Congenital Heart Disease

Cincinnati Children's Hospital Heart Institute

Department of Pediatrics

University of Cincinnati College of Medicine

Cincinnati, OH



Renal Functional Reserve in Adults with a Fontan Circulation

Kidney disease is common in single ventricle congenital heart disease patients who have had a Fontan procedure. Multiple factors contribute to kidney dysfunction in these patients including the abnormal blood flow patterns of the Fontan circulation and prior kidney injuries, specifically at the time of heart surgeries. Nonetheless, most adults with a Fontan circulation have normal screening kidney laboratory values. This is due to the kidney's remarkable ability to increase its function to accommodate for prior injuries. In fact, screening kidney function laboratory values can remain normal even when as much as 50% of the kidney is non-functional. However, in those with prior kidney injuries, there may be no reserve to further increase renal function in the setting of a physiologic or pathologic stimulus. For example, eating a high protein meal causes a reproducible increase in renal function, but this normal response may be absent in patients with prior kidney injuries. We can measure the renal functional reserve (RFR) by evaluating the change in kidney function after an oral protein load. Patients with healthy kidneys typically have a high RFR with an increase in kidney function by about 50%. However, those with little or no change in kidney function after a protein load (people without much RFR) are at higher risk of developing clinically significant chronic kidney disease. This study will measure kidney function before and after the ingestion of a high protein shake in adults with a Fontan circulation. The RFR will be calculated by the difference between kidney function measured before and after ingestion of the protein shake. The study will then assess how RFR in adults with a Fontan circulation compares to healthy adult controls and evaluate the association between RFR, history of kidney injuries, and other clinical factors. If we can demonstrate that RFR can be easily measured with this protocol, further studies may prove RFR to be a useful clinical tool to predict which patients are at the highest risk of kidney failure and aid in medication dose adjustment or monitoring.