Supplementary Methods

Databases: R.F. has access to the Cerner Health Facts database through a nonhuman subject designation approved by the Institutional Review Board at Children's Mercy Hospital. J.E. has access to the Scientific Registry of Transplant Recipients (SRTR, Minneapolis, MN) and Vizient Database (Irvine, TX) through protocols approved by the Institutional Review Board at the University of Southern California.

Cerner Health Facts- Investigation began by searching patient records (all ages) from years 2009 to 2017 inclusive. Single ventricle classification was achieved by interrogating records across outpatient, surgery and inpatient locations for the specific ICD-9 and ICD-10 codes of 745.3 (common ventricle), 746.7 (hypoplastic left heart syndrome), Q20.4 (double inlet ventricle), Q22.9 (congenital malformation of tricuspid valve, unspecified), and Q23.4 (hypoplastic left heart syndrome). Additional identification was performed using CPT-4 codes for repair of complex cardiac anomalies by simple Fontan (33615) and repair of complex cardiac anomalies by modified Fontan (33617). These codes were then referenced against multiple liver disease ICD-9 and ICD-10 codes and related procedure codes (**Table S2**).

Scientific Registry of Transplant Recipients- The SRTR data system includes data on all donor, waitlisted candidates, and transplant recipients in the U.S., submitted by the members of the Organ Procurement and Transplantation Network (OPTN). The Health Resources and Services Administration (HRSA), U.S. Department of Health and Human Services provides oversight to the activities of the OPTN and SRTR contractors. Data were reviewed for all patients in the SRTR enrolled between January 1, 1998 through December 31, 2019. The data reported here have been supplied by the Hennepin Healthcare Research Institute (HHRI) as the contractor for the SRTR. The interpretation and reporting of these data are the responsibility of the authors and in no way should be seen as an official policy of or interpretation by the SRTR or the U.S. Government.

Identification of potential patients with FALD in SRTR: When a patient is registered within the United Network for Organ Sharing (UNOS) and SRTR databases, programs may enter an organ-specific primary and secondary diagnosis. For example, a cardiac diagnosis such as “congenital heart disease” may not be selected for a patient being listed for a liver transplant, and vice versa, a patient being listed for heart transplant cannot have a liver-specific diagnostic code selected. For all organs, there is an option for a ‘free text’ diagnosis. There are no diagnostic codes for history of Fontan procedure or FALD in the UNOS or SRTR database. For patients listed for or who have received a heart transplant, there is a diagnostic code “1207: congenital heart defect with prior surgery” that has been used in prior publications to potentially capture combined heart-liver transplant patients with a history of Fontan[1]. For the purposes of the present study, potential Fontan patients were identified using the diagnostic code 1207 and also for free text diagnosis containing the word “Fontan” in the heart transplant dataset. Patients who have received a liver transplant were searched for a free text diagnosis containing the word “Fontan”. The unique patient identifier “PERS_ID” was used to link patients who received heart transplants to potential patients in the liver transplant waiting list and transplant datasets.

References for Supplemental Methods

1. Bradley EA, Pinyoluksana KO, Moore-Clingenpeel M, et al. Isolated heart transplant and combined heart-liver transplant in adult congenital heart disease patients: insights from the united network of organ sharing. *Int J Cardiol*. 2017;228:790–795. doi:10.1016/j.ijcard.2016.11.121
2. Taber DJ, Chavin KD, Bratton CF, et al. Effective systems of care improve outcomes in liver transplantation, regardless of patient-level risk factors. *Hepatology*. 2015;62(1 Suppl):837A–838A. doi:10.1002/hep.28163
3. Macomber CW, Shaw JJ, Santry H, et al. Centre volume and resource consumption in liver transplantation. *HPB (Oxford)*. 2012;14(8):554–559. doi:10.1111/j.1477-2574.2012.00503.x
4. Godown J, Thurm C, Dodd DA, et al. A unique linkage of administrative and clinical registry databases to expand analytic possibilities in pediatric heart transplantation research. *Am Heart J*. 2017;194:9–15. doi:10.1016/j.ahj.2017.08.014

DATABASE	POPULATION	DATA EXTRACTION	STRENGTHS	PITFALLS RELATED TO FONTAN PATIENTS
VIZIENT (UNIVERSITY HEATH SYSTEM CONSORTIUM)	107 academic medical centers and their affiliated hospitals Data from approximately 90% of the U.S. academic medical centers	CPT, ICD codes	Captures pediatric and adult population Inpatient and outpatient encounters Linkage to SRTR is possible [2,3]	CPT codes for Fontan procedure do not follow patients after index admission for surgery No specific ICD code to identify Fontan patients or FALD
CERNER HEALTH FACTS	Aggregate, deidentified, longitudinal EHR data from participating institutions ~70 million patients, ~500 million encounters	CPT, ICD codes	Captures pediatric and adult population Inpatient and outpatient encounters	CPT codes for Fontan procedure do not follow patients after index admission for surgery No specific ICD code to identify Fontan patients or FALD
STS CONGENITAL HEART SURGERY DATABASE	516 074 congenital heart operations in North America	Procedures by name/code	Gold standard congenital cardiac surgery database Externally audited data and accuracy rate of more than 97% Fontan data are clearly identifiable	Events are captured as individual episodes of care, not patients, eg, 1 patient with Glenn and Fontan during 2 different hospitalizations will be 2 separate events that cannot be linked Logistic challenges for linkage to external datasets Lack of longitudinal follow-up and outcomes
SCIENTIFIC REGISTRY OF TRANSPLANT RECIPIENTS (SRTR)	Transplant registry data for all U.S. Programs including live and deceased donors, transplant candidates, and transplant recipients since 1987	SRTR-specific diagnostic codes	Comprehensive dataset of solid organ transplant waiting list and transplants, specifically heart and liver	No diagnostic code for Fontan (congenital heart disease-with surgery is closest) Cardiac SRTR codes cannot be applied to noncardiac organs ie, liver, and vice versa No information related to patients evaluated and declined prior to listing
PEDIATRIC HEALTH INFORMATION SYSTEMS (PHIS)	49 children's hospitals in the U.S.	CPT, ICD codes	Large pediatric dataset Multiple institutions for rare conditions Linkage to the SRTR is possible [4]	CPT codes for Fontan procedure do not follow patients after index admission for surgery No specific ICD code to identify Fontan patients or FALD No adult patient data

Table S1: Summary of selected administrative data sources available for FALD research.

Liver Disease Diagnostic Codes	
International Classification of Diseases, Tenth Revision, Clinical Modification (ICD-10-CM)	
C22.0	Liver cell carcinoma
D13.4	Benign neoplasm of liver
I85.01	Esophageal varices with bleeding
K65.2	Spontaneous bacterial peritonitis
K76.6	Portal hypertension
T86.4	Complications of liver transplant
T86.40	Unspecified complication of liver transplant
T86.41	Liver transplant rejection
T86.42	Liver transplant failure
T86.43	Liver transplant infection
T86.49	Other complication of liver transplant
Z48.23	Encounter for aftercare following liver transplant
Z94.4	Liver transplant status
International Classification of Diseases, Ninth Revision (ICD-9)	
155	Malignant neoplasm of liver, primary
155.2	Malignant neoplasm of liver, not specified as primary or secondary
211.5	Benign neoplasmof liver and biliary passages
456	Esophageal varices with bleeding
567.2	Other suppurative peritonitis
572.3	Portal hypertension
996.82	Complications of transplanted liver
V42.7	Liver replaced by transplant
Liver Disease Procedure Codes	
Current Procedural Terminology, 4th Edition (CPT-4)	
47000	Biopsy of liver, needle; percutaneous
47100	Biopsy of liver, wedge
47135	Liver allotransplantation, orthotopic, partial or whole, from cadaver or living donor, any age
International Classification of Diseases, Tenth Revision, Procedure Coding System (ICD-10-PCS)	
0FB03ZX	Excision of liver, percutaneous approach, diagnostic
0FB03ZZ	Excision of liver, percutaneous approach
0FB13ZX	Excision of right lobe of liver, percutaneous approach, diagnostic
0FB23ZX	Excision of left lobe of liver, percutaneous approach, diagnostic
0FY00Z0	Transplantation of liver, allogeneic, open approach
International Classification of Diseases, Ninth Revision (ICD-9)	
50.11	Closed (percutaneous) [needle] biopsy of liver
50.12	Open biopsy of liver

Table S2: ICD/CPT Codes used to identify cases of liver disease.

DATABASE	POPULATION	ENROLLMENT	DATA COLLECTED	DATA AVAILABILITY	LONGITUDINAL DATA
Pediatric Heart Network Fontan Cross sectional study	546 deidentified patients age 6-18 years old from 2003-2004	Enrolled study	Functional health status questionnaire, echocardiogram, exercise test, EKG, cardiac MRI, BNP	Publicly available as a SAS dataset. Must register to use data.	No. Study measurements obtained in a 3-month window following enrollment
Australian & New Zealand Fontan Registry ***(International Fontan Interest Group)***	1528 participants as of July 2018. All procedures from Australia & New Zealand starting from 1975 Avg age is 18, 57% male, avg time of Fontan at 5.7 years age	Created in 2009. Population-based repository of Fontan patients. Mostly consented patients, but some either waivered or nonconsented patients in registry	Ages (current and operation), primary diagnosis, procedure type, medication, survival, risk factors affecting survival	Unclear	Yes, until patient drops out of registry, is transplanted, has Fontan taken down, or dies
ACC/NCDR Impact Registry	Pediatric and adult congenital heart disease patients undergoing diagnostic catheterizations and catheter-based interventions	Reported by institutions performing cardiac catheterization who elect to be in the registry	Demographics, medical history and risk factors, detailed procedural information, hemodynamic data, and information related to adverse events	Must be registered IMPACT institution	Unclear
Alliance for Adult Research in Congenital Cardiology: Fontan liver health study	Six adult congenital heart disease centers in US. Sept 2009-2012	Multi-institutional, cross-sectional observational study	Hepatic CT or MRI, Cardiac CT and MRI, liver biopsies, patient characteristic data and medical history, labs	Core institution: Boston Children's Hospital/Brigham. Additional institutions: Emory, Children's Wisconsin, UCLA, OHSU, GWU	No. Testing done within a 6-month period of liver biopsy
Cardiac Networks United	Consortium of 5 U.S. based registries including Pediatric Cardiac Critical Care Consortium (PC4), Pediatric Acute Care Cardiology Collaborative (PAC3), National Pediatric Quality Improvement Collaborative (NPC-QIC), ACTION Heart Failure Network, and Cardiac Neurodevelopment Collaborative (CNOC)	“Network of networks” consisting of PAC3, PC4, Action, NPC-QIC, and CNOC	Multiple datasets collected from clinical registries, research and quality improvement networks, and electronic health record.	Available to member institutions. Core institutions are University of Michigan (data care) and Cincinnati Children’s Hospital (improvement core)	No, however, currently a pilot program of the network, with patient reported outcomes from 2 institutions.

Table S3: Summary of selected Fontan Registries available for FALD research.