

SUPPLEMENTAL MATERIAL

Table S1. BC Children’s Hospital liver health surveillance pathway for Fontan patients.

Annual Investigations
Physical examination
Laboratory studies: ALT, AST, GGT, Alkaline phosphatase, bilirubin, albumin, CBC, INR, Hepatitis serology, APRI
Abdominal ultrasound
Transient elastography (Fibroscan®)
10 years post-Fontan [Earlier if clinically indicated]
Cardiac catheterization
Transjugular liver biopsy

Table S2. System used to stage liver fibrosis and grade portal inflammation in all transjugular liver biopsy specimens.

Parameter	Score	Degree of severity	Description
Scheuer portal fibrosis score (0-4)	0	Mild	No Fibrosis
	1		Enlarged, fibrotic portal tracts
	2	Moderate	Periportal or portal-portal septa but intact architecture
	3	Severe	Fibrosis with distorted architecture but no obvious cirrhosis
	4		Cirrhosis, probable or definite
Sinusoidal fibrosis score (0-4)	0	Mild	No fibrosis
	1		Fibrosis present in <1/3 of sinusoids
	2	Moderate	Fibrosis present in 1/3-2/3 of sinusoids
	3	Severe	Fibrosis present in >2/3 of sinusoids
	4		Cardiac cirrhosis
Portal inflammation (0-3)	0	Mild	No inflammation
	1		Mild inflammation in some or all portal tracts
	2	Moderate	Moderate inflammation in some or all portal tracts
	3	Severe	Marked inflammation in some or all portal tracts
Sinusoidal dilation (0-3)	0	Mild	No dilatation
	1		Dilatation present in <1/3 of sinusoids
	2	Moderate	Dilatation in 1/3-2/3 of sinusoids
	3	Severe	Dilatation present in >2/3 of sinusoids

Table S3. Association between the median liver stiffness and the type of systemic ventricle.

	Type of systemic ventricle			
	Right (n=35)	Left (n=33)	Indeterminate (n=8)	p
Median (IQR) liver stiffness	17.1 (14.5-21.3)	15.0 (13.3-20.0)	16.4 (11.7-21.4)	0.3

Statistical analysis performed using the Kruskal-Wallis test.

Table S4. Histological scores and demographics for each patient who underwent liver biopsy. (*Not enough portal tracts sampled. May not be representative).

Patient number	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17*
Scheuer portal fibrosis score (0-4)	2	0	1	1	1	1	1	2	2	1	0	0	1	0	1	2	1
Sinusoidal fibrosis score (0-4)	1	1	1	1	1	1	1	1	2	1	1	1	1	1	1	1	1
Portal inflammation (0-3)	1	0	1	2	1	1	1	1	0	1	1	0	1	1	1	1	0
Sinusoidal dilatation (0-3)	1	2	1	1	1	1	2	2	1	2	2	1	1	2	2	1	0
Congestive hepatic fibrosis (0-3)	0	1	0	2A	0	2A	2A	2A	2B	2B	1	0	2A	0	2B	2B	2A
Physiologic ventricle	L	R	L	R	L	R	R	R/L	R	R	L	R	L	L	L	R	L
Liver stiffness (kPa)	48.6	37.4	35.3	26.4	21.3	21.3	21	19.8	17.6	17.5	17.3	17.3	17.1	N/A	14.5	N/A	N/A
Time from Fontan	10	10	8	11	13	10	12	13	11	11	12	11	8	12	13	17	11
Age at biopsy (yr)	14	13	14	18	14	17	13	16	17	15	14	14	11	16	18	22	14

Table S5. Association between the median liver stiffness and the severity of liver pathology.

Degree of severity	N	Median (IQR) liver stiffness (kPa)	p
Scheuer portal fibrosis			
Mild (0-1)	9	21.3 (17.5-26.4)	0.5
Moderate (2)	2	33.1 (17.6-48.6)	
Severe (3-4)	0	-	
Sinusoidal dilation			
Mild (0-1)	8	21.3 (17.4-30.8)	1.0
Moderate (2)	3	21.0 (17.5-37.4)	
Severe (3-4)	0	-	
Sinusoidal fibrosis			
Mild (0-1)	10	21.3 (17.5-35.3)	0.5
Moderate (2)	1	17.6 (17.6-17.6)	
Severe (3-4)	0	-	
Portal inflammation			
Mild (0-1)	10	21.1 (17.5-35.3)	0.5
Moderate (2)	1	26.4 (26.4-26.4)	
Severe (3-4)	0	-	

Statistical analysis performed using the Wilcoxon rank-sum test.

Table S6. Association between the median APRI and the degree of severity of liver pathology.

Degree of severity	N	Median (IQR) APRI	p
Scheuer portal fibrosis			
Mild (0-1)	9	0.5 (0.4-0.5)	0.5
Moderate (2)	4	0.6 (0.3-1.0)	
Severe (3-4)	0	-	
Sinusoidal dilation			
Mild (0-1)	7	0.5 (0.5-0.7)	0.5
Moderate (2)	6	0.4 (0.3-0.5)	
Severe (3-4)	0	-	
Sinusoidal fibrosis			
Mild (0-1)	12	0.5 (0.4-0.6)	0.3
Moderate (2)	1	0.7 (0.7-0.7)	
Severe (3-4)	0	-	
Portal inflammation			
Mild (0-1)	12	0.5 (0.4-0.7)	0.7
Moderate (2)	1	0.5 (0.5-0.5)	
Severe (3-4)	0	-	

Statistical analysis performed using the Wilcoxon rank-sum test.

Table S7. Association between the median PVR and the severity of liver pathology.

Degree of severity	N	Median (IQR) PVR (mmHg.min/L)	p
Scheuer portal fibrosis			
Mild (0-1)	13	1.6 (0.9-1.9)	0.5
Moderate (2)	4	1.3 (1.0-1.6)	
Severe (3-4)	0	-	
Sinusoidal dilation			
Mild (0-1)	10	1.4 (0.9-1.8)	0.8
Moderate (2)	7	1.6 (1.0-1.9)	
Severe (3-4)	0	-	
Sinusoidal fibrosis			
Mild (0-1)	16	1.5 (0.9-1.8)	0.9
Moderate (2)	1	1.4 (1.4-1.4)	
Severe (3-4)	0	-	
Portal inflammation			
Mild (0-1)	16	1.5 (0.9-1.8)	0.4
Moderate (2)	1	0.9 (0.9-0.9)	
Severe (3-4)	0	-	

Statistical analysis performed using the Wilcoxon rank-sum test.