SUPPLEMENTARY DATA

All patients with FUH who underwent cardiac catheterization procedures prior to PCPC and TCPC, between January 2004 and January 2024, were eligible for inclusion. The study was approved by the local ethics committee of our institution, which waived the need for patient consent. Retrospective collection of demographic, clinical, and surgical data was conducted through a comprehensive review of electronic medical records. For pulmonary artery growth data, all cardiac catheterizations were reviewed, and the size of the pulmonary artery branches was re-measured. The diameter of the right and left PA branches was measured on angiograms proximal to the first lobar division. To mitigate potential measurement biases, three measurements were taken for each pulmonary branch, and the mean value was used to represent the size of the pulmonary artery branches. We calculated the cross-sectional area of the PA branches by assuming that the PA was cylindrical. The Nakata index was also calculated using the following formula: (right PA cross-sectional area + left PA cross-sectional area)/body surface area. Absolute growth of PA branches was evaluated by calculating the difference between the section area before PCPC and before TCPC. Demographic, anatomic, and hemodynamic characteristics were collected at the time of catheterization preceding PCPC and TCPC. Surgical techniques employed and postoperative courses were also recorded for each surgery. Demographic data included prenatal diagnosis, sex, prematurity (<37 weeks gestation), birth weight, low birth weight (<2500 g), small for gestational age (<the 10th percentile for gestational age), underweight (weight-for-age < −2 SD from the median of the World Health Organization Child Growth Standards), CHD subtypes, genetic abnormalities, and associated extracardiac comorbidities. Hemodynamic and anatomic parameters included pulmonary pressures, transpulmonary gradient, and the presence of PA branch stenosis, among others. Finally, we collected data on mortality or transplant, postoperative morbidity characteristics (eg, ECMO postsurgery), and mid- and long-term morbidity indicators (eg, failing Fontan status, hemodynamic contraindications to fenestration closure when measured…). Failing Fontan was defined as the presence of at least 1 of the following: Fontan pressure > 15 mmHg, protein-losing enteropathy,

plastic bronchitis, refractory arrhythmia, heart failure requiring hospitalization, listing for heart transplantation. Hemodynamic contraindication to fenestration closure was defined as a Fontan pressure > 15 mmHg or evidence of elevated systemic venous pressure associated with desaturation. Additionally, we ensured to document all patients who died between PCPC and TCPC, aiming to evaluate the presence of any potential selection bias.

Statistical methods

descriptive data are presented as proportions for categorical variables, and continuous variables as mean value ± standard deviation when variables were normally distributed, and median [interquartile range] when nonnormally distributed. To assess the growth of PA branches, measurements before the PCPC anastomosis and before the TCPC were first compared using a Wilcoxon signed-rank test. Then, we determined PA section area growth as our first outcome as follows (ΔSAp = (PA section area pre-TCPC - PA section area pre-PCPC)). The best-fitting model for PA branch growth over time was determined using the multivariable fractional polynomial method. Since no superiority of a degree 2 model over the linear model was observed (P = 0.2), linear growth of PA over time was assumed. Then, a generalized linear regression model was used to study the covariables associated with PA growth. In particular, we used multiple linear regression to compare the growth of PA branches between groups with and without APBF. Covariates identified in univariate analysis as associated with PA growth, along with previously identified potential confounding variables, were included in the multiple regression. Baseline PA size and inter-stage duration were included as forced covariates in all multivariable models. Robust standard errors were used to account for potential heteroscedasticity and intra-patient variability. Regression model estimates were reported with regression coefficients (ß) and 95% confidence intervals (CI). To not only assess statistical differences but also provide clinical relevance, effect size was calculated (evaluated by the correlation coefficient z reported to the square root of the sample) between pre-PCPC and pre-TCPC measures. Effect sizes of 0.1, 0.3, and 0.5 were considered small, medium, and

large, respectively. Finally, morbidity and mortality between groups with and without APBF were compared. Time to death or transplant was described using Kaplan–Meier curves. Survival rate estimates were calculated with the use of the Kaplan–Meier method and compared using the log-rank test. A sensitivity analysis was performed by removing the subgroup of children with HLHS considered to be at higher risk for death or transplantation. All tests were bilateral; a *P*-value less than 0.05 was considered statistically significant. All analyses were performed using Stata version 15.

Figure S1. Transplant-free survival according to patients with and without additional pulmonary blood flow (APBF)

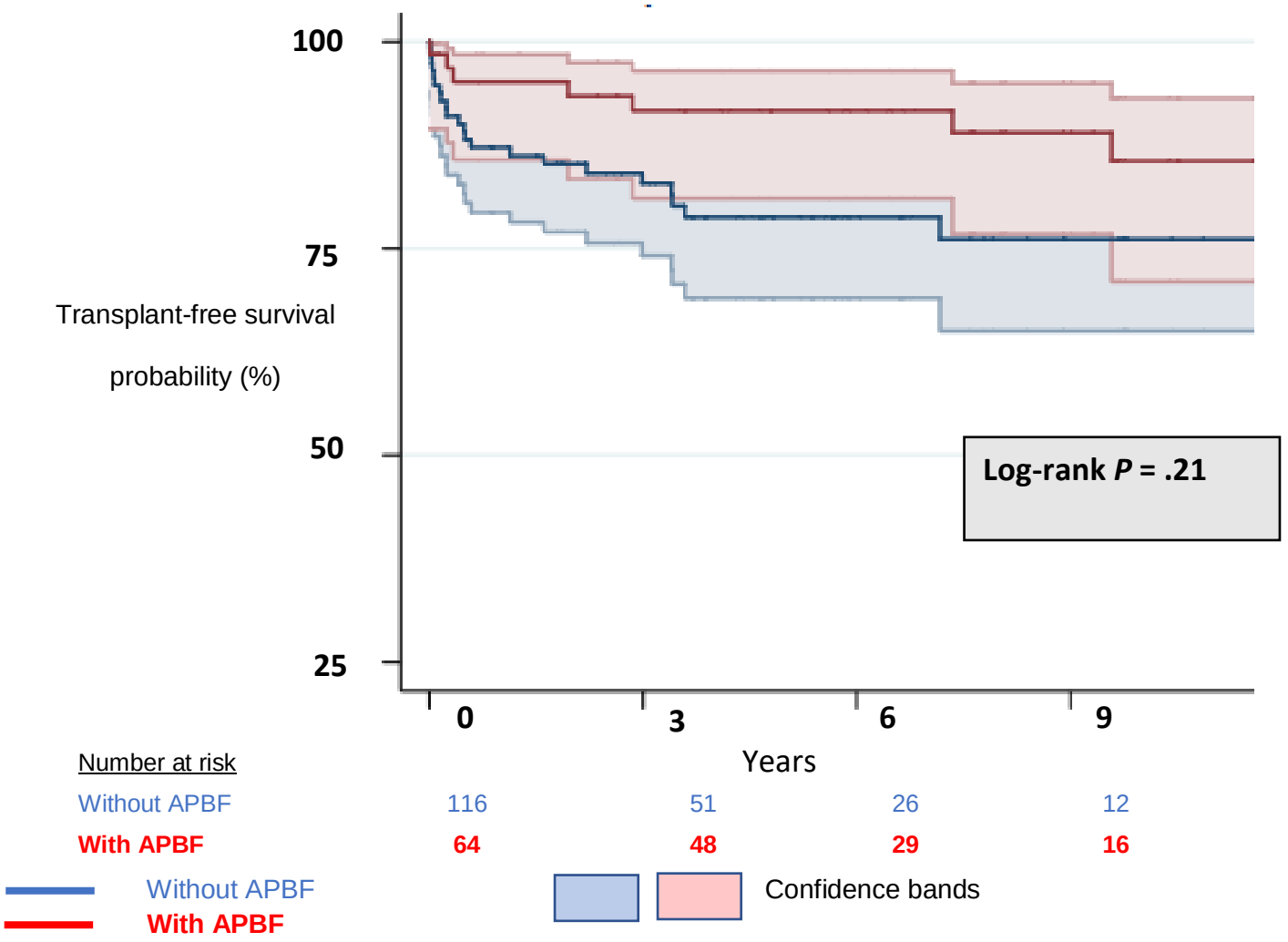

Table S1. Patient characteristics comparing patients with and without additional pulmonary blood flow (APBF)

Variables	All FUH N=180	Without APBF n = 116	With APBF n = 64	*P*
Demographic characteristics				
Sex, male	61.1	62.9	57.8	*.50*
Prenatal diagnosis	83.6	84.8	81.3	*.55*
Prematurity	5.5	1.9	0.0	*.07*
Median gestational age	39 [38-39]	39 [38-39]	39 [38-40.5]	*.87*
Small for gestational age	15.5	12.5	22.9	*.15*
Birth weight < 2500 g	8.8	10.4	5.6	*.3*
Extracardiac malformation	18.8	20.2	16.1	*.52*
Comorbidity	30.8	32.1	28.3	*.62*
Syndromic or or genetic mutation identified	*18.1*	*19.2*	*16.1*	*.52*
Median birth weight	3.1 [0.5]	3.1 [0.54]	3.0 [0.51]	*.57*
Characteristics of congenital heart defect				
CHD subtypes				

Atrioventricular valve atresia	27.2	25.9	29.7	*.58*
Double inlet ventricule	20.0	21.6	17.2	*.48*
HLHS and left ventricular hypoplasia	16.7	24.1	3.1	*< .01*
Pulmonary atresia with intact septum	11.1	12.9	7.8	*.30*
Large VSD	7.2	2.6	15.6	*< .01*
AVSD	7.2	3.5	14.1	*<.01*
Other	10.6	9.5	12.5	*.92*
UV-PA obstacle	43.6	34.8	59.4	*<.01*
UV-aorta obstacle	35.8	48.7	12.5	*< .01*
FUH morphology				*.01*
Left ventricle	43.9	42.2	46.9	
Right ventricle	41.7	48.3	29.7	
Biventricular	9.5	9.5	23.4	
Atrioventricular valve	53.6	62.6	37.5	*< .01*
Initial AVV insufficiency > or = to grade II	5.6	2.9	12.2	*.05*
Global characteristics of interventions				
Median total number of interventions	6 (2)	6 (1)	6 (2)	*.64*

Surgery	*3(0)*	*3(0)*	*3(1)*	*.16*
Interventional catheterization	*1(1)*	*1(1)*	*1(2)*	*.45*
Number of surgical procedures before PCPC	149 (82.7)	101 (87.1)	48 (75)	*.1*
Blalock shunt	30.2	29.9	30.9	*.58*
Pulmonary artery banding	40.3	36.5	50	*.1*
Norwood stage 1	24.4	30.8	9.5	*.02*
Cardiac catheterization before PCPC shunt				
Median age, mo	6.5 [9.5]	5.9 [6.6]	9.6 [32.1]	*< .01*
Weight retardation	35.8	37.9	31.8	*.41*
Median Weight (IQR)	6.6 [3.1]	6.3 [2.4]	7.7 [5.5]	*.05*
Median pulmonary pressure	15 [5]	15 [5]	14 [4]	*.09*
Median Transpulmonary gradient	6 [6]	6[5]	5 [5]	.2
Left PA branch stenosis	19.2	26.3	6.4	*< .01*
Right PA branch stenosis	24.1	31.3	14.3	*.11*
AVV insufficiency > or = to grade II	8.3	7.8	9.4	*.71*
Median diameter of the right PA branch, mm	8.6 [4.7]	7.9 [3.6]	10.4 [4.9]	*< .01*
Median diameter of the left PA branch, mm	8.2 [4.2]	7.4 [3.5]	10.3 [6.3]	*<.01*
Median Area of right PA branch, mm²	58.5 [68.1]	49.8 [46.2]	85.9 [80.8]	*< .01*
Median area of left PA branch, mm²	53.0 [56.6]	43.1 [42.5]	83.3 [103.5]	*<.01*
Nakata index mm²/m²	299.3 [250.2]	260.3 [199.6]	361.6 [300.2]	*<.01*

PCPC anastomosis				
Median time between catheterization and PCPC shunt, d	28 [59]	24.5 [52]	36.5 [63]	*.44*
Median age, mo	7.2 [10.4]	6.5 [7.1]	11.3 [31.0]	*< .01*
Anastomosis to the right PA branch	85.3	83.5	87.8	*.49*
Bilateral anastomosis	13.3	13.8	12.5	*.81*
PA bifurcation surgery	23.9	34.5	4.7	*< .01*
Right PA branch surgery	7.2	7.8	6.3	*.71*
Left PA branch surgery	8.3	12.0	1.6	*.01*
Median oxygen saturation before PCPC	80 [8]	81 [9]	80 [8.5]	*.9*
Median oxygen saturation after PCPC	86 [8]	84 [8]	88 [4.5]	*< .01*

APBF, antegrade pulmonary blood flow; CHD, congenital heart defect; FUH, functional univentricular heart; HLHS, hypoplastic left heart syndrome; PA, pulmonary artery; PCPC, partial cavopulmonary connection.

The data are expressed as percentage or median [interquartile range].

Table S2. Univariate analysis for growth of PA

	LPA			RPA			PAs branches		
	β	95%CI	*P*	β	95%CI	*P*	β	95%CI	*P*
Prematurity	3.6	−13.9 to 21.2	0.69	.9	−19.9 to 21.7	.93	4.5	−28.2to– 37.2	.78
SGA	1.1	−6.5 to 26.8	.23	−11.2	−3.8 to 8.4	.26	−1.03	−31.0 to 28.9	.94
LBW (< 2500 g)	−5.1	−24.1 to 13.8	.59	−1.9	−26.6 to 22.7	.87	−7.1	−43.8 to 29.6	.70
Chromosomal abnormality	18.6	−18.4 to 51.4	.31	−.5	−31.0 to 3.1	.98	18.1	−36.9 to 73.2	.51
Extracardiac malformation	−7.1	−3.1 to 15.9	.54	−4.4	−36.7 to 28.0	.789	−9.38	−51.1 to 32.3	.65
Comorbidity	−7.6	−26.5 to 11.3	.43	−4.6	−31.2 to 22.1	.734	−9.54	−43.6 to 24.6	.58
Initial physiology									
UV-PA obstruction	16.2	4.7 to 27.6	**.01**	−1.3	−15.5 to 12.9	.85	14.9	−6.7 to 36.0	.17
UV-aorta obstruction	−2.3	−14.4 to 9.7	.71	−3.1	−17.9 to 11.5	.67	−5.5	−27.4 to 16.7	.63
Increase of pulmonary blood flow	−17.1	−28.4 to -−5.8	**.01**	3.2	−1.8 to 17.4	.65	−13.8	−35.2 to 7.6	.20
Initial AVV insufficiency	−1.6	−27.4 to 6.2	.21	−4.2	−19.4 to 11.1	.59	−14.6	−37 to 8.4	.3
PCPC anastomosis									
Weight retardation at PCPC shunt	−1.5	−21.8 to 18.7	.88	−18.0	−32.5 to −3.6	.02	−13.9	−36.2 to 8.4	.21
Bilateral PCPC anastomosis	−.7	−17.6 to 16.2	.93	−25.1	−45.4 to −4.7	.02	−25.7	−57.3 5.4	.1
PA branch surgery at PCPC shunt	6.3	−14.5 to 27.1	.55	−11.5	−38.1 to 15.1	.42	−9.5	−31.2 to 15.1	.49
PA bifurcation surgery	.9	−12.5 to 14.5	.88	18.2	2.0 to 34.5	.03	19.2	−5.6 to 44.2	.56
APBF	18.5	6.8 to 3.1	< .01	17.3	2.8 ; 31.7	.02	36.6	14.9 to 58.3	< .01
Superior caval syndrome post PCPC shunt	−.3	−15.7 to 14.3	.97	2.4	−14.7 ;19.6	.78	2.2	−13.8 ; 19.1	.42
Collateral embolization between PCPC shunt and TCPC	−8.3	−27.4 to 1.8	.39	−2.1	−29.4 ; 25.1	.88	−5.1	−29.1 ; 17.8	.78
TCPC anastomosis									

Collateral embolization between BDCP shunt and TCPC	−8.3	−27.4 to 1.8	.390	2.4	−13.6 ;18.6	.76			
Time groups *TCPC < 3 y after PCPC* *TCPC between 3 and 6 y post PCPC* *TCPC 6 y post PCPC*	 Ref 18.9 27.9	 Ref 6.9 to 3.8 8.5 to 47.3	 Ref < .01 .01	 Ref 16.4 26.2	 Ref 1.5- 31.2 −7.2 -5.4	 Ref .03 .06	 Ref 35.2 51.1	 Ref 13.1-54.4 18.1 – 9.2	 Ref <.01 .01
Underweight at TCPC	4.4	−17.7 to 26.4	.69	−19.2	−39.3; 1.01	.06	−8.9	−33.2 ; 15.4	.54