

## Appendix 2

Table A1. Diagnosis IPCCCs that indicate the diagnosis is likely to be hypoplastic left heart related malformation or another condition rather than HLHS

03.01.05	Left isomerism ('polysplenia')
03.01.04	Right isomerism ('asplenia')
01.03.09	Atrioventricular and-or ventriculo-arterial connections abnormal
01.01.14	Double inlet atrioventricular connection (double inlet ventricle)
01.04.03	Double inlet right ventricle
01.04.04	Double inlet left ventricle
06.01.01	Tricuspid atresia
01.05.01	Discordant ventriculo-arterial connections (TGA)
01.01.02	Transposition of great arteries (TGA) (concordant atrioventricular & discordant ventriculo-arterial connections) & intact ventricular septum
01.01.03	Congenitally corrected transposition of great arteries (discordant atrioventricular & ventriculo-arterial connections)
01.01.04	Double outlet right ventricle
01.01.17	Double outlet right ventricle: Fallot type (subaortic or doubly committed ventricular septal defect & pulmonary stenosis)
01.01.40	Double outlet right ventricle: subaortic or doubly committed ventricular septal defect without pulmonary stenosis ('VSD type')
01.01.18	Double outlet right ventricle: transposition type (subpulmonary ventricular septal defect)
01.01.19	Double outlet right ventricle: with non-committed ventricular septal defect
01.01.24	Double outlet right ventricle: with intact ventricular septum
01.05.03	Double outlet left ventricle
09.01.01	Common arterial trunk (truncus arteriosus)
09.05.11	Pulmonary atresia
09.05.12	Pulmonary atresia: imperforate valve
01.01.07	Pulmonary atresia + intact ventricular septum
01.01.06	Pulmonary atresia + ventricular septal defect (including Fallot type)
01.01.25	Pulmonary atresia + ventricular septal defect + systemic-to-pulmonary collateral artery(ies) (MAPCA(s))
06.02.09	Straddling mitral valve
06.06.00	Atrioventricular septal defect (AVSD)

06.06.01	Atrioventricular septal defect: isolated atrial component (primum ASD) (partial AVSD)
06.06.08	Atrioventricular septal defect: isolated ventricular component
06.06.10	Atrioventricular septal defect: atrial & (restrictive) ventricular components + separate atrioventricular valve orifices ('intermediate')
06.06.09	Atrioventricular septal defect: atrial & ventricular components with common atrioventricular orifice (complete)
01.01.20	Atrioventricular septal defect and tetralogy of Fallot
06.07.26	Atrioventricular septal defect with ventricular imbalance
06.05.01	Atrioventricular septal defect atrioventricular valvar abnormality
06.05.06	Atrioventricular septal defect atrioventricular valvar regurgitation
07.08.41	Ventricular imbalance: dominant left ventricle + hypoplastic right ventricle
07.02.00	Right ventricular hypoplasia
10.03.01	Heart tumour

Table A2. Frequency of off pathway interventions of different types over the stages of HLHS treatment. Note each off pathway procedure can have multiple intervention types.

	Intervention type	Post Primary Procedure (%)	Post Stage 2 (%)	Post Stage 3 (%)	Post Heart Transplant (%)
Surgical procedures	Revision or replacement of arterial shunt or RV-PA conduit (Sano)	63 (19.7)	10 (3.9)	3 (1.8)	0
	Pulmonary arterioplasty procedure(s)	20 (6.3)	10 (3.9)	0	0
	Re-repair/reconstruction of the aortic arch	16 (5.0)	4 (1.6)	0	0
	Tricuspid valve repair or replacement	10 (3.1)	20 (7.8)	1 (0.6)	0
	HLHS Stage 1 (Norwood) redo procedure	9 (2.8)	0	0	0
	Creation/enlargement of interatrial communication	7 (2.2)	9 (3.5)	7 (4.1)	0
	Relief of right ventricular outflow tract obstruction	4 (1.3)	0	0	0
	Bilateral pulmonary arterial bands	4 (1.3)	0	0	0
	Pacemaker placement	2 (0.6)	5 (2.0)	11 (6.5)	0
	Debanding of pulmonary arteries	1 (0.3)	0	0	0
	Repair of anomalous pulmonary vein(s)	1 (0.3)	4 (1.6)	0	0
	Take down procedure	1 (0.3)	3 (1.2)	0	0
	Systemic vein procedure	1 (0.3)	0	0	1 (11.1)
	HLHS stage 2 (Glenn) redo procedure	0	11 (4.3)	0	0
	Occlusion procedure of systemic-to-pulmonary collateral artery(ies)	0	2 (0.8)	0	0
	Fenestration formation	0	0	5 (2.9)	0
	HLHS stage 3 (TCPC) redo procedure	0	0	3 (1.8)	0
	Neo-aortic valvoplasty	0	0	1 (0.6)	0
Not completely described , e.g. 'Cardiac procedure'	11 (3.4)	4 (1.6)	2 (1.2)	1 (11.1)	
Catheter Procedures	Stenting or dilation of residual coarctation or recoarctation	113 (35.4)	47 (18.4)	2 (1.2)	1 (11.1)
	Stenting or dilation of pulmonary artery(s)	45 (14.1)	71 (27.8)	44 (25.9)	0
	Creation/enlargement of interatrial communication	32 (10.0)	7 (2.7)	1 (0.6)	0
	Balloon or stent procedure to right ventricular outflow tract, RV-PA conduit or BT shunt	5 (1.6)	1 (0.4)	0	0
	Balloon dilation or stent of systemic vein	3 (0.9)	24 (9.4)	5 (2.9)	6 (66.7)
	Arterial duct related procedure	3 (0.9)	0	0	0
	Occlusion procedure of systemic-to-pulmonary collateral artery(ies) or venovenous collateral with device	2 (0.6)	43 (16.9)	15 (8.8)	0
	Aortic valve balloon dilation	2 (0.6)	0	0	0
	Pulmonary vein stenosis procedure	1 (0.3)	3 (1.2)	0	0

Pacemaker placement	0	1 (0.4)	1 (0.6)	0
Fenestration closure	0	0	36 (21.2)	0
Fenestration creation	0	0	33 (19.4)	0
Not completely described, e.g, 'Balloon dilation'	18 (5.6)	25 (9.8)	36 (21.2)	1 (11.1)
Total	319	255	170	9

**Abbreviations:** BT, Blalock-Taussig; HLHS, hypoplastic left heart syndrome; PA, pulmonary arterial; RV, right ventricle; TCPC, total cavopulmonary connection.