

Pediatric Heart Network
Code List A: Single Ventricle Anatomic Diagnoses

Text in red denotes change made since Version B of Code List dated 12/31/02

Level 1	Level 2	Level 3	Level 4	Level 5		
A1					Single Ventricle	
	01					Double inlet left ventricle
		01				{S,L,L} or {S,L,D} or {S,L,I} Outlet chamber (bulboventricular foramen)
			01			Non-restrictive bulboventricular foramen
				01	No pulmonary stenosis or atresia	
				02	Pulmonary stenosis	
				03	Pulmonary atresia	
			02			Restrictive bulboventricular foramen
				01	No pulmonary stenosis or atresia	
				02	Pulmonary stenosis	
				03	Pulmonary atresia	
		02				{S,D,D} or {S,D,L}, or {S,D,I} Outlet chamber (bulboventricular foramen)
			01			Non-restrictive bulboventricular foramen
				01	No pulmonary stenosis or atresia	
				02	Pulmonary stenosis	
				03	Pulmonary atresia	
			02			Restrictive bulboventricular foramen
				01	No pulmonary stenosis or atresia	
				02	Pulmonary stenosis	
				03	Pulmonary atresia	
		03				{S,D,N} Holmes heart
		04				Double outlet left ventricle with one ventricle
		05				Double outlet right ventricle with one ventricle
		06				{I,D,D} or {I,D,L}, or {I,D,I} Outlet chamber (bulboventricular foramen)
			01			Non-restrictive bulboventricular foramen
				01	No pulmonary stenosis or atresia	
				02	Pulmonary stenosis	
				03	Pulmonary atresia	
			02			Restrictive bulboventricular foramen
				01	No pulmonary stenosis or atresia	
				02	Pulmonary stenosis	
				03	Pulmonary atresia	
		07				{I,L,L} or {I,L,D}, or {I,L,I} Outlet chamber (bulboventricular foramen)
			01			Non-restrictive bulboventricular foramen
				01	No pulmonary stenosis or atresia	
				02	Pulmonary stenosis	
				03	Pulmonary atresia	

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A1					Single Ventricle (continued)			
	01					Double inlet left ventricle (continued)		
		07					{I,L,L} or {I,L,D}, or {I,L,I} Outlet chamber (bulboventricular foramen) (continued)	
			02					Restrictive bulboventricular foramen
				01				
		02						Pulmonary stenosis
		03					Pulmonary atresia	
		02					Double inlet right ventricle	
	01						Double outlet right ventricle	
			01					No pulmonary stenosis or atresia
			02					Pulmonary stenosis
			03					Pulmonary atresia
	02						Outlet chamber (bulboventricular foramen)	
			01					Non-restrictive bulboventricular foramen
			02					Restrictive bulboventricular foramen
	03						Other	
			01					No pulmonary stenosis or atresia
			02					Pulmonary stenosis
			03					Pulmonary atresia
	03						Mitral atresia	
			01					Double outlet right ventricle
			02					{S,D,N}
			03					{S,L,L} or {S,L,D} or {S,L,I} (corrected transposition)
				01				
		02						Pulmonary atresia
		03					Pulmonary stenosis	
		04					{S,D,D} or {S,D,L} or {S,D,I} (transposition)	
			01					No pulmonary stenosis or atresia
			02					Pulmonary atresia
		03					Pulmonary stenosis	
		05					{I,D,D} or {I,D,L} or {I,D,I} (corrected transposition)	
			01					No pulmonary stenosis or atresia
			02					Pulmonary atresia
			03					Pulmonary stenosis
		06					{I,L,L} or {I,L,D} or {I,L,I} (corrected transposition)	
			01					No pulmonary stenosis or atresia
			02					Pulmonary atresia
			03					Pulmonary stenosis

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A1					Single ventricle (continued)		
	04				Tricuspid atresia		
	01				D-loop, No transposition of the great arteries (Type 1)		
		01				Pulmonary atresia (Type 1a)	
		02				Pulmonary hypoplasia, small ventricular septal defect (Type 1b)	
	03				No pulmonary hypoplasia, large ventricular septal defect (Type 1c)		
	02				L-loop, No transposition of the great arteries		
		01				Pulmonary atresia	
		02				Pulmonary hypoplasia, small ventricular septal defect	
	03				No pulmonary hypoplasia, large ventricular septal defect		
	03				D-loop transposition of the great arteries (Type 2)		
		01				Pulmonary atresia (Type 2a)	
		02				Pulmonary or subpulmonary stenosis (Type 2b)	
	03				Large pulmonary artery (Type 2c)		
	04				L-loop transposition of the great arteries (Type 3)		
		01				Pulmonary or subpulmonary stenosis (Type 3a)	
		02				Subaortic stenosis (Type 3b)	
	03				Pulmonary atresia		
	05				Unbalanced atrioventricular canal defect		
		01				Right dominant	
			01				No pulmonary stenosis or atresia
			02				Pulmonary stenosis
		03				Pulmonary atresia	
		02				Left dominant	
			01				No pulmonary stenosis or atresia
	02					Pulmonary stenosis	
	03				Pulmonary atresia		
	06				Heterotaxia syndrome		
		01				Double outlet right ventricle with atrioventricular canal defect	
			01				Asplenia
		02				Polysplenia	
		02				Single left ventricle	
	03				Other		
	07				Other single ventricle		
		01				Mostly left ventricle	
		02				Mostly right ventricle	
03					Indeterminate		

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Level 1	Level 2	Level 3	Level 4	Level 5				
A2					Hypoplastic left heart syndrome			
	01				Aortic atresia and mitral atresia			
	02					Aortic atresia and mitral stenosis		
		01				Intact ventricular septum		
			02				Ventricular septal defect	
				01				Nonrestrictive ventricular septal defect
	02				Restrictive ventricular septal defect			
	03					Aortic atresia and ventricular septal defect (well-developed MV and left ventricle)		
		01				Nonrestrictive ventricular septal defect		
		02				Restrictive ventricular septal defect		
	04					Aortic stenosis and mitral stenosis		
		01				Intact ventricular septum		
			02				Ventricular septal defect	
				01				Nonrestrictive ventricular septal defect
				02				Restrictive ventricular septal defect
	05					Aortic stenosis and MV hypoplasia		
		01				Intact ventricular septum		
			02				Ventricular septal defect	
				01				Nonrestrictive ventricular septal defect
				02				Restrictive ventricular septal defect
	06					Hypoplastic aortic valve (AV) and mitral valve (MV) and left ventricle		
		01				Intact ventricular septum		
			02				Ventricular septal defect	
				01				Nonrestrictive ventricular septal defect
				02				Restrictive ventricular septal defect
	07					Aortic stenosis and mitral atresia		
		01				Nonrestrictive ventricular septal defect		
		02				Restrictive ventricular septal defect		
A3					Other functional single ventricle not fitting any other categories			
	01				Pulmonary atresia with intact ventricular septum			
	02					Ventricular septal defect(s)		
		01				No aortic or pulmonary stenosis		
		02				Aortic or subaortic stenosis		
		03				Pulmonary stenosis or atresia		
	03					Tricuspid valve anomaly		
		01				Straddling		
		02				Stenosis		
		03				Ebstein's		

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Level 1	Level 2	Level 3	Level 4	Level 5				
A3					Other functional single ventricle not fitting any other categories (continued)			
	04					D-loop double outlet right ventricle with two ventricles		
		01					Two atrioventricular valves	
			01					No aortic or pulmonary stenosis
			02					Aortic or subaortic stenosis
		03					Pulmonary stenosis or atresia	
		02					Common atrioventricular valve	
			01					No aortic or pulmonary stenosis
			02					Aortic or subaortic stenosis
		03					Pulmonary stenosis or atresia	
		05					Double outlet left ventricle with two ventricles	
	01						No aortic or pulmonary stenosis	
	02						Aortic or subaortic stenosis	
	03						Pulmonary stenosis or atresia	
	06					D-loop transposition of the great arteries with two ventricles		
		01					Solitus atria {S,D,D},{S,D,L},{S,D,A}	
			01					No aortic or pulmonary stenosis
			02					Aortic or subaortic stenosis
		03					Pulmonary stenosis or atresia	
		02					Inverse atria {I,D,D},{I,D,L}, {I,D,A}	
			01					No aortic or pulmonary stenosis
			02					Aortic or subaortic stenosis
		03					Pulmonary stenosis or atresia	
		07					L-loop transposition of the great arteries or L-loop double outlet right ventricle with two ventricles	
	01						Solitus atria {S,L,L},{S,L,D},{S,L,A}	
			01					No aortic or pulmonary stenosis
			02					Aortic or subaortic stenosis
	03						Pulmonary stenosis or atresia	
	02						Inverse atria {I,L,L},{I,L,D}, {I,L,A}	
			01					No aortic or pulmonary stenosis
			02					Aortic or subaortic stenosis
	03						Pulmonary stenosis or atresia	
	08						Mitral valve anomaly	
01						Straddling		
02						Stenosis		
09					Hypoplastic left ventricle with ventricular septal defect(s)			
	01					Tetraology of Fallot		
10					Hypoplastic right ventricle with ventricular septal defect(s)			
	01					Straddling tricuspid valve		
	02					Tricuspid valve stenosis		
A4					Unclassified (unable to classify diagnosis into any of above A1 - A3 categories)			