

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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| Level 1 | Level 2 | Level 3 | Level 4 | Level 5 | | | | |
|-----------|---------|---------|---------|---------|-------------------------------------|---|---|---|
| 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 | | | | |
| | 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) | | | |