**Inclusion/Exclusion criteria and pathway for HLHS patients**

# Identify HLHS group

## Step 1: Include patients who had evidence of HLHS

Diagnostic code evidence for HLHS that meet at least one of the following criteria:

### Patients who have a diagnosis code for HLHS

#### Table A Diagnosis codes for HLHS

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| 010109: HLHS |
| 091503: Aortic atresia |
| 060201: Mitral atresia |
| 091506. Aortic valvar atresia. |
| 060202. Mitral atresia with imperforate mitral valve. |
| 060226. Mitral atresia with absent valvar annulus (connection-junction). |

### Patients who have a code that provides some evidence for the diagnosis of HLHS

#### Table B Diagnosis codes suggestive of HLHS

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| 070842: Ventricular imbalance: dominant right ventricle and HLV |
| 070700: Left ventricular hypoplasia |

### Procedure based evidence of HLHS

#### Table C Procedure codes linked to HLHS

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| Codes in isolation:  |
| 121000: Norwood type procedure |
| 120643: Right ventricle to pulmonary artery valveless conduit construction (Japanese modification: 'Sano') as part of Norwood procedure |
| 121004: Application of bilateral pulmonary arterial bands & transcatheter placement of stent in arterial duct |
| 122020: Hypoplastic left heart syndrome hybrid approach (transcatheter & surgery): stage 1 |
| 122021: Hypoplastic left heart syndrome hybrid strategy (transcatheter & surgery) |
| 122022: Hypoplastic left heart syndrome hybrid approach (transcatheter & surgery) 'stage 2': aortopulmonary amalgamation + superior cavopulmonary anastomosis(es) + debanding of pulmonary arteries |
| 122023: Hypoplastic left heart syndrome hybrid approach (transcatheter & surgery) 'stage 2': aortopulmonary amalgamation + superior cavopulmonary anastomosis(es) + debanding of pulmonary arteries + arch repair |
| Combination codes |
| Code sets I: one code of |
| 120903: Damus-Kaye-Stansel type procedure: pulmonary trunk to aorta end/side anastomosis |
| Code sets I: and at least one of these code: |
| 123103: Modified R Blalock shunt  |
| 123104: Modified L Blalock shunt  |
| 123106: Central systemic-PA interposition shunt  |
| 123146: Modified Blalock shunt  |
| 123130: Systemic-to-pulmonary arterial shunt procedure  |
| Code sets II: one code of  |
| 121419: Application of right & left pulmonary arterial bands |
| Code sets II: and the following code within 4 weeks |
| 121014: stent placement in arterial duct |

## Step 2: Exclusions

### Exclude patients with any of the following that indicate another SV diagnosis

#### Table D Codes that mean the condition is HLHS related malformation or another type of SV disease

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| Codes in HLHS related malformation or another type of SV disease |
| 030105: Left isomerism ('polysplenia')  |
| 030104: Right isomerism ('asplenia') |
| 010309: Atrioventricular and-or ventriculo-arterial connections abnormal |
| 010114: Double inlet atrioventricular connection (double inlet ventricle) |
| 010403: Double inlet right ventricle |
| 010404: Double inlet left ventricle |
| 060101: Tricuspid atresia |
| 010501: Discordant ventriculo-arterial connections (TGA) |
| 010102: Transposition of great arteries (TGA) (concordant atrioventricular & discordant ventriculo-arterial connections) & intact ventricular septum |
| 010103: Congenitally corrected transposition of great arteries (discordant atrioventricular & ventriculo-arterial connections) |
| 010104: Double outlet right ventricle |
| 010117: Double outlet right ventricle: Fallot type (subaortic or doubly committed ventricular septal defect & pulmonary stenosis) |
| 010140: Double outlet right ventricle: subaortic or doubly committed ventricular septal defect without pulmonary stenosis ('VSD type') |
| 010118: Double outlet right ventricle: transposition type (subpulmonary ventricular septal defect) |
| 010119: Double outlet right ventricle: with non-committed ventricular septal defect |
| 010124: Double outlet right ventricle: with intact ventricular septum |
| 010503: Double outlet left ventricle |
| 090101: Common arterial trunk (truncus arteriosus) |
| 090511: Pulmonary atresia |
| 090512: Pulmonary atresia: imperforate valve |
| 010107: Pulmonary atresia + intact ventricular septum |
| 010106: Pulmonary atresia + ventricular septal defect (including Fallot type) |
| 010125: Pulmonary atresia + ventricular septal defect + systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) |
| 060209: Straddling mitral valve |
| 060600: Atrioventricular septal defect (AVSD) |
| 060601: Atrioventricular septal defect: isolated atrial component (primum ASD) (partial AVSD) |
| 060608: Atrioventricular septal defect: isolated ventricular component |
| 060610: Atrioventricular septal defect: atrial & (restrictive) ventricular components + separate atrioventricular valve orifices ('intermediate') |
| 060609: Atrioventricular septal defect: atrial & ventricular components with common atrioventricular orifice (complete) |
| 010120: Atrioventricular septal defect and tetralogy of Fallot |
| 060726: Atrioventricular septal defect with ventricular imbalance |
| 060501: Atrioventricular septal defect atrioventricular valvar abnormality |
| 060506: Atrioventricular septal defect atrioventricular valvar regurgitation |
| 070841: Ventricular imbalance: dominant left ventricle + hypoplastic right ventricle |
| 070200: Right ventricular hypoplasia |
| 071000. VSD |
| 071001. Perimembranous VSD |
| 071012. VSD + malaligned outlet septum |
| 071101. Muscular VSD |
| 071200. Subarterial VSD |
| 071201. Doubly committed subarterial VSD |
| 071402. Communication between left ventricle + right atrium (Gerbode defect) |
| 071405. Inlet VSD |
| 071504. Multiple VSDs |
| 071505. Single VSD |
| 010405.Double inlet to solitary ventricle of indeterminate morphology. |
| 020303.Crisscross heart (twisted atrioventricular connections). |
| 060311.Congenital anomaly of right-sided atrioventricular valve in double inlet ventricle. |
| 060411.Congenital anomaly of left-sided atrioventricular valve in double inlet ventricle. |
| 010110.Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect. |
| 090908. Pulmonary artery from ascending aorta (hemitruncus) |
| 091010. Discontinuous (non-confluent) pulmonary arteries |
| 010126.Tetralogy of Fallot with pulmonary atresia. |
| 010157.Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral artery(ies) (MAPCA(s)). |
| 090516.Congenital pulmonary atresia. |
| 090705. Absent or atretic pulmonary trunk (main pulmonary artery). |
| 060598. Deficient mural-lateral leaflet of left ventricular component of common atrioventricular valve (left atrioventricular vale) |
| 050603.Common atrium with common atrioventricular junction. |
| 060514.Atypical common atrioventricular valve. |
| 060525. Double orifice of left ventricular component of common atrioventricular valve (left atrioventricular valve). |
| 060560.Common atrioventricular valvar regurgitation. |
| 060571.Atypical right ventricular component of common atrioventricular valve (right atrioventricular valve). |
| 060572.Atypical left ventricular component of common atrioventricular valve (left atrioventricular valve). |
| 060705. Atrioventricular septal defect (AVSD) with ventricular imbalance with dominant right ventricle and hypoplastic left ventricle. |
| 060706. Atrioventricular septal defect (AVSD) with ventricular imbalance with dominant left ventricle and hypoplastic right ventricle. |
| 060727. Atrioventricular septal defect (AVSD) with balanced ventricles. |
| 060728.Common atrioventricular junction with spontaneous fibrous closure of atrioventricular septal defect (AVSD). |
| 060736. Common atrioventricular valve with unbalanced commitment of valve to ventricles. |
| 060737.Common atrioventricular valve with unbalanced commitment of valve to right ventricle. |
| 060738.Common atrioventricular valve with unbalanced commitment of valve to left ventricle. |
| 071002.Inlet perimembranous ventricular septal defect (VSD) without atrioventricular malalignment without a common atrioventricular junction. |
| 071004. Outlet perimembranous ventricular septal defect (VSD) with anteriorly malaligned of outlet septum. |
| 071017.Outlet ventricular septal defect (VSD) with anteriorly malaligned outlet septum. |
| 071018.Outlet ventricular septal defect (VSD) with posteriorly malaligned outlet septum. |
| 071019. Outlet perimembranous ventricular septal defect (VSD) with posteriorly malaligned outlet septum. |
| 071102.Inlet muscular ventricular septal defect (VSD). |
| 071103.Trabecular muscular ventricular septal defect (VSD) apical. |
| 071104.Trabecular muscular ventricular septal defect (VSD) midseptal. |
| 071105.Trabecular muscular ventricular septal defect (VSD)s multiple (Swiss cheese septum). |
| 071106. Outlet muscular ventricular septal defect (VSD) without malalignment. |
| 071107.Trabecular muscular ventricular septal defect (VSD) anterosuperior. |
| 071112.Trabecular muscular ventricular septal defect (VSD) postero-inferior. |
| 071115. Outlet muscular ventricular septal defect (VSD) with anteriorly malaligned outlet septum. |
| 071116. Outlet muscular ventricular septal defect (VSD) with posteriorly malaligned outlet septum. |
| 071202. Doubly committed juxta-arterial ventricular septal defect (VSD) without malalignment and with muscular postero-inferior rim. |
| 071203. Doubly committed juxta-arterial ventricular septal defect (VSD) without malalignment and with perimembranous extension. |
| 071205. Doubly committed juxta-arterial ventricular septal defect (VSD) with anteriorly malaligned fibrous outlet septum and perimembranous extension. |
| 071206. Doubly committed juxta-arterial ventricular septal defect (VSD) with posteriorly malaligned fibrous outlet septum and perimembranous extension. |
| 071207. Doubly committed juxta-arterial ventricular septal defect (VSD) with anteriorly malaligned fibrous outlet septum and muscular postero-inferior rim. |
| 071208. Doubly committed juxta-arterial ventricular septal defect (VSD) with posteriorly malaligned outlet septum and muscular postero-inferior rim. |
| 071209.Outlet ventricular septal defect (VSD) without malalignment. |
| 071212.Doubly committed juxta-arterial ventricular septal defect (VSD) with anteriorly malaligned fibrous outlet septum. |
| 071213.Doubly committed juxta-arterial ventricular septal defect (VSD) with posteriorly malaligned fibrous outlet septum. |
| 071406.Inlet perimembranous ventricular septal defect (VSD) with atrioventricular septal malalignment and without common atrioventricular junction. |

### Exclude patients who had a procedure that can never occur in f-SV patients

Remove patients if had a procedure that can never occurred in SV patients Table E.

#### Table E List of procedure codes that should not occur in HLHS or in any SV patients

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| 120017: Scimitar syndrome (partially anomalous pulmonary venous connection) repair |
| 120029: Systemic venous pathway procedure (post Senning-Mustard) |
| 120107: Patent foramen ovale (PFO) closure with transluminal device |
| 120198: Interatrial communication closure with transluminal device |
| 120401: Atrioventricular septal defect (AVSD): partial (primum ASD) repair |
| 120409: Atrioventricular septal defect (AVSD): partial with isolated ventricular component (VSD) repair |
| 120501: Atrioventricular septal defect (AVSD): complete (common valve orifice) repair |
| 120510: Atrioventricular septal defect (AVSD): 'intermediate' repair |
| 120571.Atrioventricular septal defect (AVSD) repair with direct ventricular component closure & patch to atrial component (Nunn/Wilson). |
| 120635: Double chambered right ventricle repair |
| 120719: Left ventricular outflow tract obstruction relief by transcatheter coronary chemical ablation |
| 120738: Partial left ventriculectomy-volume reduction (Batista) |
| 120801: Ventricular septal defect (VSD) closure |
| 120802: Ventricular septal defect (VSD) closure by direct suture |
| 120803: Ventricular septal defect (VSD) closure using patch |
| 120807: Ventricular septal defect (VSD) closure with transluminal device |
| 120816: Closure of multiple ventricular septal defect (VSD)s |
| 120819: Open fenestration of ventricular septal defect (VSD) patch |
| 120820: Transluminal fenestration of ventricular septal defect (VSD) patch |
| 120828: Intraoperative ventricular septal defect (VSD) closure with transluminal device (hybrid approach) |
| 121100: Common arterial trunk (truncus) repair |
| 121201: Aortopulmonary window closure  |
| 121321: Pulmonary valvar replacement (not conduit) |
| 121322: Pulmonary valvar replacement using homograft |
| 121351: Transluminal pulmonary valvar insertion with stent mounted valve |
| 121355: Pulmonary valve repair converted to pulmonary valvar replacement |
| 121381: Transluminal aortic valvar insertion with stent mounted valve |
| 121384: Transapical aortic valve implantation (hybrid approach) |
| 121385: Transluminal pulmonary valvar insertion with stent mounted valve including prestenting |
| 121386: Transluminal pulmonary valvar prestenting procedure in preparation for valve replacement |
| 121430: Pulmonary artery origin from ascending aorta (hemitruncus) repair |
| 121630: Ross procedure: aortic valve or root replacement with pulmonary autograft & pulmonary valvar replacement |
| 121662: Ross-Konno procedure |
| 121681: Aortic sinus of Valsalva distal fistula closure |
| 121685: Aortic sinus of Valsalva aneurysm repair |
| 121690: Aorto-left ventricular tunnel closure |
| 121732: Pulmonary arterial sling repair |
| 121799: Aortic root replacement of implanted pulmonary autograft & pulmonary valve re-replacement & pulmonary valve re-replacement |
| 122300: Anomalous coronary artery (eg ALCAPA) repair |
| 122342: Transluminal chemical occlusion of coronary artery |
| 122380: Anomalous aortic origin of coronary artery repair |
| 122601: Tetralogy of Fallot repair  |
| 122613: Tetralogy of Fallot repair with transannular patch |
| 122620: Tetralogy of Fallot repair without transannular patch |
| 122621: Absent pulmonary valve syndrome (Fallot type) repair |
| 122701: Double outlet right ventricle with subaortic or doubly committed ventricular septal defect (VSD) & pulmonary stenosis (Fallot-type) repair |
| 122702: Double outlet right ventricle repair with intraventricular tunnel |
| 122745: REV procedure: intraventricular left ventricle to aorta tunnel with infundibular septum resection & direct right ventricle to pulmonary trunk anastomosis |
| 122750: Double outlet left ventricle repair |
| 122778: Aortic root translocation to over left ventricle (including Nikaidoh) |
| 122801: Pulmonary atresia & ventricular septal defect (VSD) (including Fallot-type) repair |
| 122811: Pulmonary atresia, ventricular septal defect (VSD) & systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) repair |
| 122901: Senning procedure (atrial inversion) |
| 122902: Mustard procedure (atrial inversion) |
| 122920: Double outlet right ventricle repair |
| 122921: Arterial switch procedure  |
| 122925: Arterial switch & atrial inversion procedures ('double switch') |
| 122926: Atrial inversion and Rastelli procedures |
| 122952: Pulmonary venous pathway procedure (post Senning-Mustard) |
| 122979: Atrial inversion procedure (Mustard or Senning) revision |
| 123760: Lung(s) transplant |
| 123825: Transluminal left atrial appendage occlusion with device |
| 121005. Hypoplastic left heart biventricular repair |
| 120511: Atrioventricular septal defect (AVSD) & Tetralogy of Fallot repair |
| 122911: Rastelli procedure: intraventricular left ventricle to aorta tunnel & right ventricle to pulmonary artery conduit |
| 122940: Complex transposition of great arteries repair |
| 123210: Heart tumour resection  |

### Exclusion step c), d) and e) should be done after the pathway analysis.

### Exclusion of patients with small left heart who go down a biventricular pathway

If there is a stage one Type B (Table F), AND also there is NO stage one type A nor stage two nor stage three then exclude. (stage one type B can occur as an off pathway in patients who do have these procedures - stage one type A or stage two or stage three)

#### Table F: Coarctation/interrupted arch repair codes

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| **Stage one Type B: Coarctation/interrupted arch repair** |
| 121800: Coarctation-hypoplasia of aorta repair |
| 121801: Aortic coarctation-hypoplasia repair by resection & end to end anastomosis |
| 121802: Aortic coarctation-hypoplasia repair by patch aortoplasty |
| 121803: Aortic coarctation-hypoplasia repair by subclavian flap aortoplasty |
| 121804: Balloon dilation of native aortic coarctation-hypoplasia |
| 121810: Aortic coarctation-hypoplasia repair by resection & extended end to end anastomosis |
| 121815: Aortic coarctation-hypoplasia repair by resection & insertion of tube graft |
| 121827: Aortic coarctation transluminal obstruction relief |
| 121830: Aortic arch repair |
| 122100: Interrupted aortic arch repair.  |

### Exclusion of patients who are selected by diagnosis code 070700 Left ventricular hypoplasia only and have no procedures in HLHS pathway

### Exclusion of patients who had pre pathway only and an ASD closure code

#### Table G: ASD closure code

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| 120102. Atrial septal defect (ASD) secundum closure with direct suture, |
| 120103. Atrial septal defect (ASD) secundum closure with patch, |
| 120108. Interatrial communication closure: partial, |
| 120189.Patent foramen ovale (PFO) closure using transluminal suture. |

# Step 3 Identify HLHS Pathway

Consider patients selected in [step 1](#_Step_1:_Diagnostic) and [step 2](#_Step_2:_Procedure) and then assign pathway type to each procedure.

HLHS patients can be only managed in SV pathway.

The first occurrence of a pathway procedure identifies the occurrence of that pathway. Hence for any of the 3 defined pathway procedures, i.e., palliative first stage procedure, stage two and stage three, only the first occurrence will be the pathway of that type. Any subsequent occurrence of a procedure in the same pathway group will be identified as a re-do or additional or off pathway procedure. Note if palliative first stage procedure appeared after stage two/three, it will be off pathway procedure. Patients can have up 3 pathway procedures in whole history.

Patients who have a procedure before the first pathway procedure are considered to have had a ‘pre-pathway procedure’. These will be described.

Patients who have no pathway procedures will overlap in the types of procedures they are having with pre-pathway procedures, these will be described.

Then patients who have a pathway procedure and then have additional (post pathway), additional or off pathway procedures which will be described divided by the stage at which they occur.

Assign pathway type to each procedure:

## Palliative first stage procedure/stage one for HLHS, type A and C

#### Table H: Palliative first stage procedure for HLHS (in hierarchy)

|  |
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| **Type A: Norwood type (including Sano and Damus)** |
| 121000: Norwood type procedure  |
| 120643: Right ventricle to pulmonary artery valveless conduit construction (Japanese modification: 'Sano') as part of Norwood procedure |
| 120903: Damus-Kaye-Stansel type procedure: pulmonary trunk to aorta end/side anastomosis. Should be with one of (may not always be coded):* 123103: Modified R Blalock interposition shunt
* 123104: Modified L Blalock interposition shunt
* 123106: Central systemic-PA interposition shunt
* 123146: Modified Blalock interposition shunt
* 123130: Systemic-to-pulmonary arterial shunt procedure
 |
| **Type C Hybrid procedure used in HLHS** |
| Codes in isolation |
| 121004. Application of bilateral pulmonary arterial bands & transcatheter placement of stent in arterial duct, |
| 122020. Hypoplastic left heart syndrome hybrid approach (transcatheter & surgery): stage 1, |
| 122021. Hypoplastic left heart syndrome hybrid strategy (transcatheter & surgery) |
| 124130: Hybrid strategy (combined surgical & transluminal).  |
| Combination codes: One of these |
| 121014: Stent placement in arterial duct (PDA stand) |
| 124511: Stent placement |
| And one the these within 4 weeks: |
| 121419: Application of right & left pulmonary arterial bands  |
| 121402: Pulmonary trunk band (PA band). |

## Stage two: Glenn or comprehensive stage two

#### Table I: SV stage two code

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| **Glenn:**  |
| 123111: Bidirectional superior cavopulmonary (Glenn) anastomosis |
| 123115: Hemi-Fontan procedure |
| 123144: Bilateral bidirectional superior cavopulmonary (Glenn) anastomoses |
| 123145: Unidirectional superior cavopulmonary (Glenn) anastomosis |
| 123172: Superior caval vein to pulmonary artery anastomosis. |
| **Comprehensive stage two:**  |
| A Glenn code in combination with a • 121000: Norwood type procedure• 120903: Damus-Kaye-Stansel type procedure: pulmonary trunk to aorta end/side anastomosis |
| Or Codes in isolation |
| 122022: Hypoplastic left heart syndrome hybrid approach (transcatheter & surgery) 'stage 2': aortopulmonary amalgamation + superior cavopulmonary anastomosis(es) + debanding of pulmonary arteries |
| 122023: Hypoplastic left heart syndrome hybrid approach (transcatheter & surgery) 'stage 2': aortopulmonary amalgamation + superior cavopulmonary anastomosis(es) + debanding of pulmonary arteries + arch repair |

## Stage three: Fontan

#### Table G: Fontan code

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| 123001: Fontan type procedure |
| 123005: Total cavopulmonary connection (TCPC) using extracardiac inferior caval vein (IVC)-pulmonary artery conduit with fenestration |
| 123006: Total cavopulmonary connection (TCPC) with fenestrated lateral atrial tunnel |
| 123013: Fontan procedure with atrioventricular connection |
| 123028: Fontan-type connection without fenestration |
| 123032: Fontan procedure with direct atriopulmonary anastomosis |
| 123050: Total cavopulmonary connection (TCPC) |
| 123051: Total cavopulmonary connection (TCPC) with lateral atrial tunnel |
| 123054: Total cavopulmonary connection (TCPC) using extracardiac inferior caval vein (IVC)-pulmonary artery conduit |
| 123060: Completion of total cavopulmonary connection (TCPC) using transcatheter inferior to superior caval vein covered stent |
| 123092.Total cavopulmonary connection (TCPC) using intra-extracardiac conduit: fenestrated. |
| 123093.Total cavopulmonary connection (TCPC) using intra-extracardiac conduit: nonfenestrated. |

Stage three refinement:

* if a patient has no stage three but it has the code “123027: Fenestration of Fontan type connection”, then this indicates that a stage three occurred.
* If a patient has two stage threes without stage two, the first stage three under 1 year old will be stage two.

# Step 5: Identify diagnostic subgroups

HLHS is a single and there is no subgroups.

# Step 6: Remove patients according to the violation rules.

## Generic rule: Exclude patients if

* had only non-contributory procedure records via activity algorithm.

## HLHS specific rule: Exclude patients if

Note in future routine monitoring these could be raised with centre and re included if corrected and deemed as genuine HLHS patients but in Champion we treat as violation rules.

In this rule the following are counted as a stage one – Norwood, hybrid for HLHS, one of the hybrid codes in isolation these two 121419 application of bilateral bands OR 121014 stent of duct stage one type B Table F.

* Patients who had no stage one for HLHS by age three months and are surviving at the time point of three months of age do not have HLHS – exclude
* Any patients who had a regular stage two without stage one occurring by the age of three months (skipped stage one), does not have HLHS– exclude

# Step 7: Flagging rules to centre

Include these in the cohort. In future routine monitoring all such patients will be flagged with the treating centres for correction.

## Patients with suspected miscoded/missing data

 These patients will be excluded from pathway analysis and reintervention monitoring.

### Generic rule (for patients under SV pathway:)

* Patients recorded as having stage two at less than one month old
* Patients recorded as having stage three at less than six months old

### HLHS specific rule:

* Patients who had a stage one for HLHS (Norwood or hybrid) and no stage two by 18 months old whilst surviving, whatever happens to them after they reach the age of 18 months.
* Patients who had a stage one (Norwood or hybrid) for HLHS and stage three, but no stage two by any age.
* Patients who had a stage one (Norwood or hybrid) for HLHS and multiple stage two but no stage three by age 8 years whilst surviving whatever happens to them after age 8
* Patients who had a possibly incompletely coded hybrid (only allow the more specific hybrid codes here)– one of these codes 121419 application of bilateral bands OR 121014 stent of duct, before age three months, followed by stages two and or three depending on age of follow up.
* Patients who had a stage one as type B table F followed by stages two and or three depending on age of follow up.

## Minor data errors or patients with unusual records

### Generic flagging rule:

* Flag patients if there is a cardiopulmonary bypass surgery as a pre-pathway procedure - please check this patient’s diagnostic and procedure coding is correct