**Inclusion/Exclusion criteria and pathway**

**for Pulmonary Atresia patients**

# Identify pulmonary atresia (PA) group

## Step 1: Include patients who had evidence of pulmonary atresia

### Diagnostic code evidence of primary diagnosis pulmonary atresia including those with intact septum and those with ventricular septal defect

Diagnostic code evidence for pulmonary atresia including those with intact septum (PA-IVS) and those with ventricular septal defect (PA-VSD) that meet at least one of the following criteria:

#### Patients who have a diagnosis code for pulmonary atresia with intact septum (PA-IVS) or pulmonary atresia with ventricular septal defect (PA-VSD)

##### Table A Diagnosis codes for PA-IVS or PA-VSD as listed for Top rank

|  |
| --- |
| **Diagnosis codes for Pulmonary Atresia as listed for Top rank** |
|  010107. Pulmonary atresia + intact ventricular septum |
|  010106. Pulmonary atresia + VSD (including Fallot type) |
|  010125. Pulmonary atresia + VSD + systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) |
| 090908. Pulmonary artery from ascending aorta (hemitruncus) |
| 010126.Tetralogy of Fallot with pulmonary atresia. |
| 010157.Tetralogy of Fallot with pulmonary atresia and systemic-to-pulmonary collateral artery(ies) (MAPCA(s)). |

#### Patients who have a code that provides some evidence or indirect evidence for the diagnosis of one of the pulmonary atresia primary types

##### Table B Diagnosis codes suggestive of pulmonary atresia primary types

|  |
| --- |
| 090511. Pulmonary atresia |
| 090512. Pulmonary atresia: imperforate valve |
| 090726. Solitary arterial trunk (absent intrapericardial pulmonary arteries) |
| 090908 Pulmonary artery from descending aorta  |
| 091010. Discontinuous (non-confluent) pulmonary arteries |
| 090516.Congenital pulmonary atresia. |
| 090705. Absent or atretic pulmonary trunk (main pulmonary artery). |
| 090902.Right pulmonary artery from arterial duct. |
| 090903.Right pulmonary artery from ascending aorta. |
| 090904.Left pulmonary artery from arterial duct. |
| 090905.Left pulmonary artery from ascending aorta. |
| 090911. Pulmonary artery from arterial duct. |
| 091030.Congenitally discontinuous. non-confluent right and left pulmonary arteries. |
| 091075.Absent or atretic right pulmonary artery. |
| 091077.Absent or atretic left pulmonary artery. |

### Procedure code evidence of primary diagnosis pulmonary atresia

#### Identify patients based on procedure evidence

Any patients who are not picked up by diagnosis but who have procedure evidence of Pulmonary Atresia be identified as follows Table C:

##### Table C Procedure codes linked to repair in Pulmonary atresia diagnosis

|  |
| --- |
| **Definitive Pulmonary atresia repair codes**  |
| 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, |
| **Procedures very strongly linked to Pulmonary atresia diagnosis**  |
| 121309. Pulmonary valvar transluminal perforation & dilation |
| 122500 MAPCA Uni-focalisation  |
| 121430 Pulmonary artery from aorta (hemitrunc) repair |

## Step 2: Exclude patients who had more complex CHD

### Exclude patients who have a code listed in Table D, as well as those identified as having HLHS, FUH, or TGA according to the CHANPION rules, which cannot be adequately captured by Table D alone

### Table D: Pulmonary atresia diagnosis Exclusion Codes (codes that are indicative of more complex CHD condition)

As this is a diagnosis-based analysis, we mainly use diagnostic exclusion codes with a small number of key procedure exclusion codes. This is because the procedures undertaken in the patients are very variable and we aim to be inclusive to variations in management pathway.

|  |
| --- |
| **Diagnostic codes** |
| 010109. Hypoplastic left heart syndrome |
| 060201. Mitral atresia |
| 091503. Aortic atresia |
| 010114. Double inlet AV connection (double inlet ventricle) |
| 010122. Functionally univentricular heart |
| 010124. Double outlet right ventricle: with intact ventricular septum |
| 010403. Double inlet RV |
| 010404. Double inlet LV |
| 020305. Solitary ventricle of indeterminate morphology |
| 070841. Functionally univentricular heart |
| 070842. Functionally univentricular heart |
| 090101. Common arterial trunk (truncus arteriosus) |
| 090200. Truncal valvar abnormality |
| 090203. Truncal valvar regurgitation |
| 092931. Interrupted aortic arch |
| 010103. Congenitally corrected transposition of great arteries (discordant AV & VA connections) |
| 010116. Partially anomalous pulmonary venous connections: Scimitar syndrome |
| 010133. Shone's syndrome: left heart obstruction at multiple sites, |
| 010503. Double outlet left ventricle |
| 050201. Cor triatriatum (divided left atrium) |
| 090401. Aortopulmonary window |
| 091600. Supravalvar aortic stenosis |
| 094101. Anomalous origin of coronary artery from pulmonary artery |
| 060134. Ebstein's malformation of tricuspid valve |
| 050601. Common atrium (virtual absence of atrial septum) |
| 060501. AVSD AV valvar abnormality |
| 060506. AVSD AV valvar regurgitation |
| 060600. Atrioventricular septal defect |
| 060601. AVSD: isolated atrial component (primum ASD)(partial) |
| 060608. AVSD: isolated ventricular component |
| 060609. AVSD: atrial & ventricular components with common AV orifice (complete) |
| 060610. AVSD: atrial & (restrictive) ventricular components + separate AV valves ('intermediate') |
| 010120. AV septal defect and Tetralogy of Fallot |
| 060726. AVSD with ventricular imbalance |
| 010117. Double outlet right ventricle: Fallot type (subaortic or doubly committed VSD & pulmonary stenosis) |
| 091501. Aortic valvar stenosis: congenital |
| 091513. Aortic valvar stenosis |
| 091592. Aortic stenosis |
| 050202. Supravalvar mitral ring |
| 060207. Mitral valvar stenosis: congenital |
| 060213. Mitral subvalvar stenosis |
| 060256. Parachute malformation of mitral valve |
| 060292. Mitral stenosis |
| 060293. Mitral valve stenosis |
| 040600. Totally anomalous pulmonary venous connection: supracardiac |
| 040805. Totally anomalous pulmonary venous connection |
| 040806. Obstructed pulmonary venous connection(s) |
| 040810. Totally anomalous pulmonary venous connection: intracardiac |
| 040820. Totally anomalous pulmonary venous connection: infracardiac |
| 040830. Totally anomalous pulmonary venous connection: mixed |
| 030104. Right isomerism ('asplenia')  |
| 030105. Left isomerism ('polysplenia') |
| 010118. Double outlet right ventricle: transposition type (subpulmonary VSD) |
| 010501. Discordant VA connections (TGA) |
| 010102. Transposition of great arteries (concordant AV & discordant VA connections) & IVS |
| 070700. Left ventricular hypoplasia  |
| 010309 AV and or/VA connections abnormal  |
| 090525 Absent pulmonary valve syndrome  |
| 091506. Aortic valvar atresia. |
| 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. |
| 090111. Common arterial trunk (truncus arteriosus) with aortic dominance and one pulmonary artery absent from trunk. isolated pulmonary artery. |
| 090112. Common arterial trunk (truncus arteriosus) with pulmonary dominance and aortic arch obstruction. |
| 090114. Common arterial trunk (truncus arteriosus) with aortic dominance and both pulmonary arteries arising from trunk. |
| 090115. Common arterial trunk (truncus arteriosus) with aortic dominance (no aortic arch obstruction). |
| 090118. Common arterial trunk (truncus arteriosus) with pulmonary dominance and interrupted aortic arch. |
| 090119. Common arterial trunk (truncus arteriosus) with pulmonary dominance and aortic coarctation. |
| 090201.Dysplasia of truncal valve. |
| 090218.Congenital truncal valvar stenosis. |
| 090219.Congenital truncal valvar regurgitation. |
| 010110.Transposition of the great arteries with concordant atrioventricular connections and ventricular septal defect. |
| 092932. Interrupted aortic arch distal to subclavian artery. type A. |
| 092933. Interrupted aortic arch between subclavian & common carotid arteries. type B. |
| 092934. Interrupted aortic arch between carotid arteries. type C. |
| 150503. Pulmonary vein obstruction |
| 020101.Extrathoracic heart (ectopia cordis). |
| 040802.Congenital atresia of pulmonary vein(s). |
| 040804.Congenital anomaly of pulmonary vein(s). |
| 040831.Congenital pulmonary venous stenosis and-or hypoplasia. |
| 090407.Congenital aortopulmonary window. |
| 091618. Congenital supravalvar aortic stenosis. |
| 093134.Vascular ring of left aortic arch and right arterial duct or ligament. |
| 093135.Vascular ring of right aortic arch and left arterial duct or ligament. |
| 094103. Anomalous origin of left coronary artery from pulmonary artery (ALCAPA). |
| 094221.Anomalous aortic origin of coronary artery with ventriculo-arterial concordance. |
| 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. |
| 060202. Mitral atresia with imperforate mitral valve. |
| 060203.Dysplasia of mitral valve. |
| 060204. Mitral annular hypoplasia. |
| 060205. Overriding mitral valve. |
| 060211.Congenital anomaly of mitral valve. |
| 060221.Congenital anomaly of mitral subvalvar apparatus. |
| 060222. Congenital mitral subvalvar stenosis. |
| 060226. Mitral atresia with absent valvar annulus (connection-junction). |
| 060272. Congenital mitral valvar prolapse. |
| 040807. Anomalous pulmonary venous connection. |
| 091517. Aortic 'annular' hypoplasia. |
| 092944.Descending thoracic or abdominal aortic coarctation. |
| **Procedure exclusion codes (that have a special link to this diagnosis and therefore can lead to patients being misallocated)**  |
| 122911 Rastelli |
| 122621 Absent pulmonary valve syndrome repair |
| 122745 REV procedure  |
| 121100 Common arterial trunk repair |
| 121201 aortopulmonary window repair |
| 121402 Pulmonary trunk band  |
| 121312 Pulmonary valvectomy  |

# Step 3 Identify Pulmonary atresia diagnosis - Pathway

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

Note that assignment of procedures on a palliative pathway or a biventricular Pulmonary atresia diagnosis reparative pathway are in parallel.

The individual types of pathway procedure are ‘Stage one palliative procedures’ these can occur in any patient, ‘stage two (glenn)’ and ‘stage three (fontan)’, which are palliative procedures for single ventricle pathway, and ‘reparative procedures for Pulmonary atresia diagnosis’ which are of various types and are biventricular procedures.

In our data management, once a glenn or fontan occurs the patient goes on ‘single ventricle pathway sequence’. Patients who do not have a glenn or fontan are potential patients for the reparative pathway sequence, (assumption in Pulmonary atresia diagnosis group is that these are biventricular patients).

If multiple types of pathway procedure happened in the same record, then we mark the individual procedure as the most complex one, i.e., the hierarchy/complexity order is palliative procedures in these groups first stage three, then stage two including in this case stage 1.5 ventricle repairs, then reparative procedure for Pulmonary atresia diagnosis, then stage one (procedures as listed).

The first occurrence of a pathway procedure Identifies the occurrence of this type of pathway procedure. For any of the defined pathway procedures, i.e., palliative procedures (stages one two or three) and reparative procedure for Pulmonary atresia diagnosis, only the first occurrence will be the pathway procedure of that type. Other subsequent occurrence of these individual procedures will be identified as re-do and off pathway. Patients can have up to 4 pathway procedures in whole history.  Of note, additional reparative or stage one procedure(s) after final expected reparative procedure will be identified as additional or off pathway. Stage two and three can arise at any time in journey, and when they occur then these procedures identify the overall management pathway of the patient as a single ventricle sequence of procedures.

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 be described.

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

Pathway and off pathway procedures will later be described by TGA subtypes see Step 5.

## Pathway applicable to Pulmonary atresia diagnosis

Assign pathway type to each procedure:

### Palliative procedures applicable in Pulmonary atresia diagnosis

#### Palliative first stage procedure

Look for staged palliative procedures, i.e., palliative first stage procedure

##### Table E: Palliative stage one type procedures that may occur in Pulmonary atresia diagnosis

|  |
| --- |
| **Palliative procs in Pulmonary atresia diagnosis – Type D Securing pulmonary blood flow** |
| 121014: Stent placement in arterial duct (PDA)  |
| 123103: Modified R Blalock interposition shunt |
| 123104: Modified L Blalock interposition shunt |
| 123106: Central systemic-Pulmonary Atresia interposition shunt |
| 123130: Systemic-to-pulmonary arterial shunt procedure |
| 123146: Modified Blalock interposition shunt |
| **Also for pulmonary atresia this is a palliative procedure that also needs to flag to centre**  |
| 120618 stent placement in RVOT |

Stages two and three - If the patient has either or both of these procedures (Glenn and or Fontan), then the patient has undertaken a ‘single ventricle pathway’

In many clinical situations the stage three is diagnostic of single ventricle pathway and the stage two remans ambiguous as to whether or not single ventricle pathway has been undertaken. However in this analysis we will describe patients who have either or both of these stages (Glenn and Fontan) as having taken single ventricle pathway.

#### Stage two (3 types):

##### Glenn with reparative proc

Glenn codes occur with a reparative procedure for pulmonary atresia

##### Glenn in isolation

In presence of code:

* 123111: Bidirectional superior cavopulmonary (Glenn) anastomosis
* 123115: Hemi-Fontan procedure
* 123144: Bilateral bidirectional superior cavopulmonary (Glenn) anastomoses
* 123145: Unidirectional superior cavopulmonary (Glenn) anastomosis
* 123144: Bilateral bidirectional superior cavopulmonary (Glenn) anastomoses
* 123145: Unidirectional superior cavopulmonary (Glenn) anastomosis
* 123172: Superior caval vein to pulmonary artery anastomosis.

##### 1.5 V repair

In Pulmonary atresia diagnosis we also have this possible procedure which can sit within the single ventricle pathway procedures for this analysis

* 120619. 1.5 ventricle repair: superior cavopulmonary (Glenn) anastomosis + right ventricular outflow tract reconstruction.

#### Stage three: Fontan

In presence of code:

|  |
| --- |
| * 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 occasionally occurs with a procedure in Table J. This is allowed.

### Pulmonary atresia diagnosis pathway: reparative procedure for Pulmonary atresia diagnosis

To label the Pulmonary Atresia repair pathway, the pathway is determined by the first pathway procedure that occurs for the patient. Then if two or more Fallot repair types (meaning from i-iv) occur in the same record at the same time. then there is a hierarchy to assign which it is assigned to i to v in this order

#### Pulmonary atresia diagnosis repair pathway surgery

##### Table F: Definitive Reparative procedures for Pulmonary atresia diagnosis (surgery)

|  |
| --- |
| **Single code for repair**  |
| 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, |
| 122821. Pulmonary atresia. ventricular septal defect (VSD) & systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) repair: status post complete unifocalisation with all usable collaterals incorporated. |
| 122822. Pulmonary atresia. ventricular septal defect (VSD) & systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) repair: status post incomplete unifocalisation with not all usable collaterals incorporated. |

#### Pulmonary atresia diagnosis repair pathway catheter

 Single code for Reparative procedures for Pulmonary atresia diagnosis (catheter):

* 121309. Pulmonary valvar transluminal perforation & dilation

#### Other reparative procedures in pulmonary atresia

These codes can arise even as first procedures in patients with PA, therefore if arising as a first reparative we consider these to be the ‘pathway procedure’. These procedures may also appear as re operations later in the history if they happen after a reparative pathway procedure then they are counted as additional or off pathway.

##### Table G: Reparative types of procedure

|  |
| --- |
| **Code in isolation**  |
| 122500. Systemic-to-pulmonary collateral artery (ies) (MAPCA(s)) unifocalisation procedure, |
| 121430. Pulmonary artery origin from ascending aorta (hemitruncus) repair, |
| 122920 Double outlet right ventricle repair  |
| 122567. Unilateral Systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) unifocalisation procedure. |
| 122587. Bilateral systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) unifocalisation procedure: complete with all usable collaterals incorporated. |
| 122588. Bilateral systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) unifocalisation procedure: incomplete with not all usable collaterals incorporated. |

#### Reparative procedures only involving the pulmonary valve and or pulmonary arteries (excluding 122500 Systemic-to-pulmonary collateral artery (ies) (MAPCA(s)) unifocalisation procedure)

These may happen with or without a VSD / ASD repair code.

##### Table H Pulmonary Atresia repair type codes only involving the pulmonary valve or pulmonary arteries, report the catheter and surgery types separately

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| --- |
| **One of these (allow in isolation or combo with VSD closure codes) Surgery codes**  |
| 121300. Pulmonary valvar procedure, |
| 121302. Pulmonary valvotomy: open, |
| 121321. Pulmonary valvar replacement (not conduit), |
| 121322. Pulmonary valvar replacement using homograft,  |
| 123601. Right ventricle to pulmonary arterial tree conduit construction, |
| 120641 Right ventricular outflow tract obstruction relief  |
| 120600 Right ventricular outflow tract procedure  |
| 120643 RVPulmonary Atresia Sano type  |
| 121420 Pulmonary arterioplasty/ reconstruction, |
| 121401 Pulmonary trunk arterioplasty |
| 121421 Pulmonary arterioplasty / reconstruction  |
| 121422 Pulmonary arterioplasty peripheral  |
| 120612.Right ventricle to pulmonary artery valveless conduit construction. |
| 120647.Right ventricular outflow tract reconstruction: transannular patch & constructed monocuspid valve. |
| **Catheter procedures**  |
| 121305 Balloon dilatation of the pulmonary valve |
| 120605 Balloon dilation of the RVOT |

#### Reparative procedures with incomplete coding

##### Table I Reparative procedures with incomplete coding – Patients who have these flag to centre

|  |
| --- |
| **Catheter procedures**  |
| 123600 Conduit construction  |
| 123640 Procedure involving a conduit |

#### Fallot type reparative procedures

##### Table J Procedure codes indicating a Fallot type repair – Patients who have these flag to centre

|  |
| --- |
| **Definitive Fallot repair codes**  |
| 122601. Tetralogy of Fallot repair |
| 122613. Tetralogy of Fallot repair with transannular patch |
| 122620. Tetralogy of Fallot repair without transannular patch |
| 122701. Double outlet right ventricle with subaortic or doubly committed VSD & pulmonary stenosis (Fallot-type) repair |

#### VSD/ASD repair codes

These suggest a repair happened, if they are in isolation then the coding is incomplete so flag to centre

##### Table K VSD / ASD closure codes– Patients who have these flag to centre

|  |
| --- |
| **VSD repair codes**  |
| 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, |
| 120828. Intraoperative ventricular septal defect (VSD) closure with transluminal device (hybrid approach), |
| 120101. Atrial septal defect (ASD) secundum closure |
| 120102. Atrial septal defect (ASD) secundum closure with direct suture |
| 120103. Atrial septal defect (ASD) secundum closure with patch |
| 120106. Atrial septal defect (ASD) secundum closure with transluminal device |
| 120110. Sinus venosus defect (ASD) closure |
| 120122. Atrial septation procedure |

# Step 4: Identify diagnostic subgroups

## Pulmonary Atresia & VSD including DORV and Fallot MAPCA types

If one of these codes in Table L appears then group as Pulmonary Atresia VSD including DORV and Fallot MAPCA types .

#### Table L Pulmonary Atresia VSD Codes

|  |
| --- |
| 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, |
| 122500. Systemic-to-pulmonary collateral artery (ies) (MAPCA(s)) unifocalisation procedure, |
| 121430. Pulmonary artery origin from ascending aorta (hemitruncus) repair, |
| 010101. Tetralogy of Fallot |
| 010117. Double outlet right ventricle: Fallot type (subaortic or doubly committed VSD & pulmonary stenosis) |
|  010106. Pulmonary atresia + VSD (including Fallot type) |
|  010125. Pulmonary atresia + VSD + systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) |
| 120801. VSD closure |
| 120802. VSD closure by direct suture |
| 120803. VSD closure using patch |
| 120828. VSD closure with prosthesis - intraoperatively |
| 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 |
| 071501. Tiny VSD (Maladie de Roger) |
| 071504. Multiple VSDs |
| 071505. Single VSD |
| 010119 DORV with uncommitted VSD |
| 010104 Double outlet right ventricle  |
| 122601. Tetralogy of Fallot repair,  |
| 122613. Tetralogy of Fallot repair with transannular patch,  |
| 122620. Tetralogy of Fallot repair without transannular patch,  |
| 122518. Systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) occlusion, |
| 122519. Transluminal procedure to systemic-to-pulmonary collateral artery (MAPCA(s)), |
| 122562. Stent placement in systemic-to-pulmonary collateral artery (MAPCA(s)), |
| 122565. Transluminal occlusion of systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) with coil-device, |
| 122572. Balloon dilation of systemic-to-pulmonary collateral artery(ies) (MAPCA(s)), |
| 090726. Solitary arterial trunk (absent intrapericardial pulmonary arteries) |
| 090801. Major systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) |
| 092025. Systemic-to-pulmonary collateral arter(ies) (MAPCA(s)) stenosis(es) |
| 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. |
| 122567. Unilateral Systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) unifocalisation procedure. |
| 122587. Bilateral systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) unifocalisation procedure: complete with all usable collaterals incorporated. |
| 122588. Bilateral systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) unifocalisation procedure: incomplete with not all usable collaterals incorporated. |
| 122821. Pulmonary atresia. ventricular septal defect (VSD) & systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) repair: status post complete unifocalisation with all usable collaterals incorporated. |
| 122822. Pulmonary atresia. ventricular septal defect (VSD) & systemic-to-pulmonary collateral artery(ies) (MAPCA(s)) repair: status post incomplete unifocalisation with not all usable collaterals incorporated. |

## Pulmonary Atresia with intact ventricular septum (IVS)

Patients who have one of the codes in Table M, group to Pulmonary Atresia IVS

#### Table M Codes Pulmonary Atresia IVS codes

|  |
| --- |
|  |
| 010107. Pulmonary atresia + intact ventricular septum |
| 060101. Tricuspid atresia |
| 072100 intact ventricular septum  |

Pulmonary Atresia and IVS group is all patients with a code in Table M as definitive Pulmonary Atresia IVS evidence AND

Patients who have none of the codes for Pulmonary Atresia with VSD that are listed in Table L, are to go in group Pulmonary Atresia IVS. These are included patients who need to be flagged to centre for checking in however these can all be analysed and reported as Pulmonary Atresia with IVS.

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

## Generic rule: Exclude patients if

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

## Pulmonary Atresia specific rule:

None

# 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.

## If single ventricle pathway then

## Patients recorded as having stage two at less than one month old

## Patients recorded as having stage three at less than six months old

## If reparative pathway then (currently none)

## Minor data errors or patients with unusual records

Patients can be included in analysis with these as long as they didn’t have major data missing.

Generic flagging rule –

* if there is a cardiopulmonary bypass surgery as a pre pathway procedure then flag to centre as please check this patient’s diagnostic and procedure coding is correct

Pulmonary Atresia specific flagging rule –

* Patients who have no reparative procedure and are alive at the age of 3 years. Flag to centre rather than remove. Many might have data error but are plausible.
* Patients who had 120618 stent placement in RVOT as palliative first stage operation.
* Patients in Pulmonary Atresia IVS group who do not have any code in Table M – these patients have some minor coding errors / ambiguous coding which might be improved by centre.
* Patients who have a reparative procedure with subtypes Reparative procedures with potentially incomplete coding, fallot type repair and VSD/ASD repair.