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

# Identify Fallot group

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

## Diagnostic code evidence of Fallot

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

### Patients who have a diagnosis code for Fallot

#### Table A Diagnosis codes for Fallot as listed for Top rank

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| **Diagnosis codes for Fallot as listed for Top rank** |
| 010101. Tetralogy of Fallot |
| 010117. Double outlet right ventricle: Fallot type (subaortic or doubly committed VSD & pulmonary stenosis) |
| 090525. Absent pulmonary valve syndrome: Fallot-type |

### Patients who have a code that provides some evidence or indirect evidence for the diagnosis of Fallot

#### Table B Diagnosis codes suggestive of Fallot

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| None |

### 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 Fallot be identified as follows Table C:

##### Table C Procedure codes linked to Fallot / Fallot repair

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| **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 |
| 122621. Absent pulmonary valve syndrome (Fallot type) repair |
| 122701. Double outlet right ventricle with subaortic or doubly committed VSD & pulmonary stenosis (Fallot-type) repair |

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

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.

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

#### Table D Fallot Exclusions Codes (codes that are indicative of more complex CHD condition)

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| **Diagnostic codes (since this is a diagnosis based analysis we mainly rely on diagnosis codes here)** |
| 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 |
| 060101. Tricuspid atresia |
| 070841. Functionally univentricular heart |
| 070842. Functionally univentricular heart |
| 090101. Common arterial trunk (truncus arteriosus) |
| 090200. Truncal valvar abnormality |
| 090203. Truncal valvar regurgitation |
| 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 |
| 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)) |
| 090511. Pulmonary atresia |
| 090512. Pulmonary atresia: imperforate valve |
| 090726. Solitary arterial trunk (absent intrapericardial pulmonary arteries) |
| 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 |
| 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 |
| 050202. Supravalvar mitral ring |
| 060207. Mitral valvar stenosis: congenital |
| 060213. Mitral subvalvar stenosis |
| 060225. Mitral regurgitation: congenital |
| 060236. True cleft of mitral leaflet (without AVSD) |
| 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 |
| 092901. Aortic coarctation |
| 092911. Aortic arch hypoplasia (tubular) |
| 030104. Right isomerism ('asplenia') |
| 030105. Left isomerism ('polysplenia') |
| 092931. Interrupted aortic arch |
| 091501. Aortic valvar stenosis: congenital |
| 091512. Eccentric opening of tricuspid aortic valve |
| 091513. Aortic valvar stenosis |
| 091592. Aortic stenosis |
| 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. |
| 091506. Aortic valvar atresia. |
| 091618. Congenital supravalvar aortic stenosis. |
| 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 codes that have a special link to this diagnosis and therefore can lead to patients being misallocated** |
| 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 |
| 120511. Atrioventricular septal defect (AVSD) & Tetralogy of Fallot repair |
| 070200 right ventricular hypoplasia |
| 070700 left ventricular hypoplasia |

# Step 3 Identify Fallot 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 the SV pathway and Fallot pathway are in parallel, so patients can under both pathways.

If multiple types of pathway happened in the same record, then mark the procedure as the most complex one, i.e., the hierarchy/complexity order is reparative procedure for Fallot, stage two and palliative procedure.

First occurrence of a pathway procedure identifies the pathway. For any of the 3 defined pathway procedures, i.e., palliative procedure, stage two and reparative procedure for Fallot, only the first occurrence will be the pathway of that type. Others will be re-do and off pathway. Patients can have up to 3 pathway procedures in whole history.  Of note, additional procedure(s) after final expected reparative procedure will be either a reparative procedure redo (eg residual VSD, residual RV outflow tract obstruction or PA stenosis); or off repair pathway, but part of longer term expected pathway i.e., they are related to replacement of the pulmonary valve which is often needed in children with tetralogy, usually in teenage years or early adulthood, due to post-procedural pulmonary regurgitation, particularly if original repair involved a transannular patch. Others may have had an RV-PA valved conduit at the original definitive repair, which fails (stenosis =/- regurgitation) or becomes relatively too small for the growing child, and needed to be replaced.

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 non pathway procedures are having additional or off pathway procedures which will be described divided by the stage at which they occur.

## Pathway applicable to

Assign pathway type to each procedure:

### Palliative procedures applicable in Fallots

Look for staged palliative procedures, i.e., palliative first stage procedure and stage two. Note for the palliative codes, if some of these appear with a VSD closure code (see Table J) then they will be classified as a repair because of the hierarchical allocation.

### Table E: Palliative procedure (stage one type D): Securing pulmonary blood flow, e.g. systemic-to-pulmonary arterial shunt

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| **Palliative procs in Tetralogy of Fallot** |
| 121014: Stent placement in arterial duct (PDA) |
| 123103: Modified R Blalock interposition shunt |
| 123104: Modified L Blalock interposition shunt |
| 123106: Central systemic-PA interposition shunt |
| 123130: Systemic-to-pulmonary arterial shunt procedure |
| 123146: Modified Blalock interposition shunt |
| 120618: Stent placement in right ventricular outflow tract |
| 123105: Waterston (ascending aorta-right pulmonary artery) anastomosis |
| 121305: Balloon dilation of pulmonary valve |
| 121309: Pulmonary valvar transluminal perforation & dilation |
| 121302: Pulmonary valvotomy: open |
| 120641: Right ventricular outflow tract obstruction relief |
| 120600. Right ventricular outflow tract procedure, |
| 120821: Subpulmonary obstruction relief |

### Stage two: Glenn operation

See code list in SV stage two, Generic pathway document. (does not include comp stage two), if any glenn codes appears with a repair code this will be classed as a Fallot repair plus.

### Table F: stage two codes

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| **Glenn codes** |
| 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 |
| 123111. Bidirectional superior cavopulmonary (Glenn) anastomosis |
| **Glenn code with 1.5 ventricle repair** |
| 120619. 1.5 ventricle repair: superior cavopulmonary (Glenn) anastomosis + right ventricular outflow tract reconstruction, |

### Fallot pathway (reparative procedure for Fallot)

To label the Fallot 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

#### Fallot and absent pulmonary valve repair pathway

##### Table G: Fallot and absent pulmonary valve repair Code (from Table D)

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| **As a single code tetralogy and absent PV repair code** |
| 122621. Absent pulmonary valve syndrome (Fallot type) repair |

#### Tetralogy repair pathway that may occur in any TOF absent pulmonary valve, tetralogy or tetralogy DORV patient

Tetralogy repair pathway procedures – this is a subset of Table D

##### Table H : Tetralogy Repair Codes

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| **Single codes that mean tetralogy type repair** |
| 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 |

#### Other reparative procedure linked to Fallot diagnosis (may with atypical or incomplete coding)

These codes are not typical repair pathway codes however they are noted to arise even as first procedures in patients with Fallot, therefore if arising first we consider these to be the ‘Fallot pathway procedure’. These procedures may also appear as re operations later in the history. Note in flagging rules we will ask centres to check data if the patients repair is coded as per Table J. We will use the data if coded like this unless changed by centre in later routine use.

##### Table J: Codes for Fallot repair pathway with atypical or incomplete coding

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| **A single code – these are full repair type codes** |
| 120635. Double chambered right ventricle repair |
| 122920. Double outlet right ventricle repair |
| 122702. Double outlet right ventricle repair with intraventricular tunnel |
| **The following codes usually appear with a VSD closure code but allow in isolation as a reparative code in tetralogy as first occurrence (in data checking these codes do appear in isolation as repair codes)** |
| 123601. Right ventricle to pulmonary arterial tree conduit construction |
| 121321. Pulmonary valvar replacement (not conduit), |
| 121322. Pulmonary valvar replacement using homograft |
| **The following codes are reparative when in combination with VSD codes (these codes appear reasonably often in isolation as palliative procedures)**  **– one of these (with)** |
| 120641: Right ventricular outflow tract obstruction relief |
| 120600. Right ventricular outflow tract procedure, |
| 120626. Right ventricular procedure, |
| 121302: Pulmonary valvotomy: open |
| 120821: Subpulmonary obstruction relief |
| **One of these VSD codes at same procedure** |
| 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 |
| 120828. Intraoperative ventricular septal defect (VSD) closure with transluminal device (hybrid approach) |
| 120816 Closure of multiple VSDs |

#### VSD type repairs in tetralogy patients

Occasionally the first reparative procedure is listed as either VSD repair. As such this is allowed as a pathway type. Note if these occur at the same time as Fallot repair types i, ii, iii the pathway type is labelled as the applicable type i-iii given the hierarchy in assignment.

##### Table K: VSD repair code

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| **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 |
| 120828. Intraoperative ventricular septal defect (VSD) closure with transluminal device (hybrid approach) |
| 120816 Closure of multiple VSDs |

**Plus Codes that may occur in Fallot Pathway**

For the reparative procedure for Fallot, the occurrence of one of the additional list of plus codes listed in Table L means it is a more complex repair.

##### Table L: Plus Codes

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| **Plus codes in tetralogy A stage two code or PA reconstruction code** |
| 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 |
| 121420. Pulmonary arterioplasty/ reconstruction |
| 121421. Pulmonary arterioplasty/ reconstruction: central (proximal to hilar bifurcation) |
| 121422. Pulmonary arterioplasty/ reconstruction: peripheral (at-beyond hilar bifurcation) |

# Step 4: Identify Fallot diagnostic subgroups

## Tetralogy absent pulmonary valve

Patients identified with any of these codes:

* 122621. Absent pulmonary valve syndrome (Fallot type) repair
* 090525. Absent pulmonary valve syndrome: Fallot-type

## Tetralogy with any evidence of double outlet right ventricle (DORV)

Patients with any of these codes:

* 010117. Double outlet right ventricle: Fallot type (subaortic or doubly committed VSD & pulmonary stenosis)
* 122701. Double outlet right ventricle with subaortic or doubly committed VSD & pulmonary stenosis (Fallot-type) repair
* 122702. Double outlet right ventricle repair with intraventricular tunnel

## Standard tetralogy

Everyone else

# Step 5: Remove patients according to the violation rules.

## Generic rule: Exclude patients if

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

# Step 6: 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 missing or miscoded data

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

* Patients who were under SV pathway (had stage two or stage three) but had stage two/three older than five years old
* Standard tetralogy patients who were under SV pathway (had stage two or stage three)
* Patients who had at least one procedure then no stage two, stage three or reparative procedures by age two years whilst surviving (note patients who only have a reparative procedure at older age can be included)

## Patients with minor data errors/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

Tetralogy flagging rule –

* if the reparative procedure is identified by Table J flag or Table K VSD type to centre to consider whether the procedure coding is complete or do they need to correct / add a code
* if patients had a Fontan codes.

|  |
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| **Fontan codes:** |
| 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. |