Tetralogy of Fallot (TOF)

(Core Condition)

Description	The simultaneous presence of a ventricular septal defect (VSD), pulmonic and subpulmonic stenosis, a malpositioned aorta that overrides the ventricular septum, and right ventricular hypertrophy. 
Inclusions	Pentalogy of Fallot – Tetralogy of Fallot with an associated inter-atrial communication, either a patent foramen ovale (PFO) or an atrial septal defect (ASD). Tetralogy of Fallot (TOF) Tet Pulmonary atresia with VSD (see 'Additional information')
Exclusions	Simultaneous occurrence of a VSD and pulmonary stenosis that has TOF physiology but has not been diagnosed as Tetralogy of Fallot. Also, some coding systems may also include Trilogy of Fallot, or Fallot's Triad – the simultaneous presence of an atrial septal defect, pulmonic stenosis, and right ventricular hypertrophy. This is not to be included as TOF.
ICD-9-CM Codes	745.2
ICD-10-CM Codes	Q21.3
CDC/BPA Codes	745.20 – 745.21, 747.31 (Note: code 746.84 (trilogy of Fallot) has been removed)
Diagnostic Methods	While Tetralogy of Fallot may be suspected by clinical presentation, it may be conclusively diagnosed only through direct visualization of the heart by cardiac echo (echocardiography), catheterization, surgery, or autopsy.
Prenatal Diagnoses Not Confirmed Postnatally	These conditions may be included as cases when only diagnosed prenatally. However, if it is possible to ascertain the degree of certainty of the prenatal diagnosis, this should factor into the decision as to whether or not to include an individual case in the surveillance data. Live-born children who survive should always have confirmation of the defect postnatally.

Additional Information:

Children with Tetralogy of Fallot may experience episodes of cyanosis or hypoxia that result from shunting of unoxygenated blood across the VSD from the right to the left ventricle. Children who have a coexisting VSD and pulmonary stenosis, but do not have Tetralogy of Fallot, may experience similar episodes. Thus, the occurrence of cyanosis or hypoxia does not necessarily mean a child has been diagnosed with Tetralogy of Fallot.

Tetralogy of Fallot is one of several abnormalities of the outflow tract of the heart known as conotruncal defects. Some infants (approximately 1 in 7) with these defects have a deletion on the short arm of