

Introduction Classic tetralogy of Fallot (TOF) is a congenital heart defect (CHD) that is comprised of 4 anatomical alterations: a large, anteriorly malaligned ventricular septal defect (VSD), an overriding aorta which results in infundibular (ie, sub-pulmonary) right ventricular outflow tract obstruction (RVOTO), and consequent right ventricular hypertrophy secondary to chronic systemic pressures. These may include atrial septal defects, patent ductus arteriosus, supraaortic pulmonary stenosis, branch pulmonary artery stenoses, hypoplastic branch pulmonary arteries, pulmonary valve atresia which may develop during fetal life as the subpulmonary infundibular narrowing progresses, a disconnected left pulmonary artery that originates from the ascending aorta formerly known as hemitruncus, a left pulmonary artery arising from the ductus arteriosus, absent left pulmonary artery, absent pulmonary valve, anomalous coronary arteries, anomalous pulmonary venous return, aortic incompetence, aortopulmonary window, and atrioventricular septal defect (AVSD).[3] Patients with TOF and pulmonary atresia may have a remnant of a pulmonary artery trunk with different calibers of the central pulmonary arteries and variable pulmonary tree anatomy. The VSD is most frequently located in the perimembranous septum; however, the defect can extend to the muscular septum, and infrequently, there might be additional muscular VSDs.[1] A right aortic arch is observed in 20% to 25% of TOF.[2] The clinical presentation will also depend on the associated cardiovascular anomalies in roughly 40% of patients with TOF. In the other 50%, pulmonary flow is from multiple collateral vessels, usually from the descending thoracic aorta, and is not ductal-dependent. Occasionally, collateral arteries might arise from the head and neck, abdominal aorta, or coronary arteries. The pulmonary valve annulus is often hypoplastic, with a pulmonary valve that is dysplastic and stenotic. In approximately 50% of these patients, the right and left pulmonary arteries are confluent, and blood flow is ductal-dependent.