*Case Reports*

# Ductal Stent Implantation in Tetralogy of Fallot with Aortic Arch Abnormality

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*Stenting of patent ductus arteriosus is an alternative to palliative cardiac surgery in newborns with duct-dependent or decreased pulmonary circulation; however, the use of this technique in patients with an aortic arch abnormality presents a challenge. Tetralogy of Fallot is a congenital heart defect that is frequently associated with anomalies of the aortic arch and its branches. The association is even more common in patients with chromosome 22q11 deletion.*

*We present the case of an 18-day-old male infant who had cyanosis and a heart murmur. After an initial echocardiographic evaluation, the patient was diagnosed with tetralogy of Fallot and right-sided aortic arch. The pulmonary annulus and the main pulmonary artery and its branches were slightly hypoplastic; the ductus arteriosus was small. Conventional and computed tomographic angiograms revealed a double aortic arch and an aberrant left subclavian artery. The right aortic arch branched into the subclavian arteries and continued into the descending aorta, whereas the left aortic arch branched into the common carotid arteries and ended with the patent ductus arteriosus. After evaluation of the ductal anatomy, we implanted a 3.5 × 15-mm coronary stent in the duct. Follow-up injections showed augmented pulmonary flow and an increase in oxygen saturation from 65% to 94%. The patient was also found to have chromosome 22q11 deletion.* ***(Tex Heart Inst J 2015;42(3):281-4)***

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Tetralogy of Fallot (TOF) is one of the most common forms of cyanotic congenital heart disease. It occurs in 3 of every 10,000 live births and constitutes 7% to 10% of all congenital heart defects.[1] Approximately 6% to 21% of all TOF patients and 20% of patients with aortic arch abnormality have chromosome 22q11 deletion syndrome. However, the combination of TOF and double aortic arch is rarely seen in cases of 22q11 deletion.[2-4]

The initial presentation of the patient with TOF depends on the degree of right ventricular outflow tract (RVOT) obstruction. Most often, cyanosis is mild at birth and gradually worsens with age, as hypertrophy of the right ventricular (RV) infundibulum progressively obstructs the RVOT. In some cases, severe cyanosis is seen at birth or shortly thereafter.[5] Diagnosis of TOF in the neonate before closure of the ductus arteriosus presents the challenging clinical necessity of predicting the degree of pulmonary blood flow once the ductus has undergone spontaneous closure. Neonates with critically restricted antegrade flow and duct-dependent circulation must be started on prostaglandin $E_1$ and considered for aortopulmonary-shunt adjustment or total repair.[6]

Despite advances in surgical repair techniques of cyanotic congenital heart disease and a trend toward early primary repair, first-stage palliation via the surgical creation of an aortopulmonary shunt remains, in developing countries, an important option for the treatment of duct-dependent cyanotic congenital heart disease. Yet shunt surgery during the neonatal period is an important cause of morbidity and death.

In neonatal patients, percutaneous stenting of the patent ductus arteriosus (PDA) is an alternative to surgical palliation,[7-10] but its use in cases of aortic arch abnormality presents a major technical challenge. Herein, we describe a ductal-stent implantation procedure in a newborn with TOF, aortic arch abnormality, and 22q11 deletion syndrome.

## Case Report

An 18-day-old male infant was referred to our hospital with cyanosis and heart murmur. The patient had dysmorphic facial features, a length of 50 cm, a weight of 3.2 kg,

a heart rate of 140 beats/min, and a 65% oxygen saturation rate on room air as measured by pulse oximetry. A harsh grade 3/6 systolic ejection murmur was detected in the 3rd right intercostal space. An electrocardiogram revealed right-axis deviation, and a chest radiograph showed oligemic lung fields with no cardiomegaly. After initial transthoracic 2-dimensional and color-flow Doppler echocardiographic evaluation, the patient was diagnosed with TOF (Figs. 1A and B). The pulmonary annulus and the main pulmonary artery and its branches were slightly hypoplastic; the ductus arteriosus was small. Prostaglandin $E_1$ infusion was started immediately, and catheterization was performed for diagnostic purposes. Angiography after injection of contrast medium into the RV showed right-to-left shunting across the ventricular septal defect (VSD) into the overriding aorta, RV hypertrophy, and subpulmonic and valvular pulmonary stenosis (Figs. 2A and B). In addition, injections into the aorta passing through the VSD revealed a double aortic arch and aberrant left subclavian artery (SCA). The right aortic arch branched into the right and left SCAs and continued into the descending aorta, whereas the left aortic arch branched into the right and left common carotid arteries and ended with the PDA.

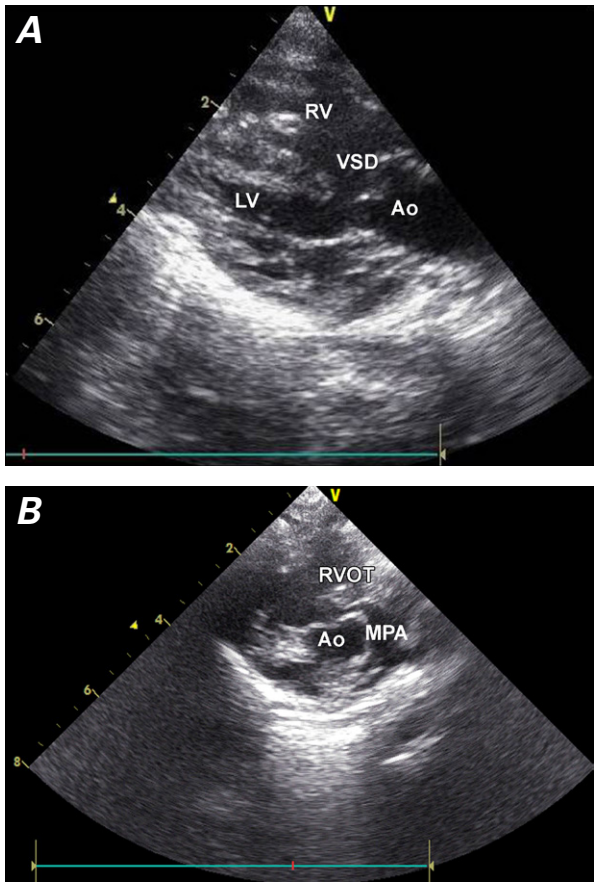


***Fig. 1*** *Two-dimensional echocardiograms in **A**) parasternal long-axis view and **B**) short-axis view show tetralogy of Fallot with subpulmonic and valvular stenosis.*

*Ao = aorta; LV = left ventricle; MPA = main pulmonary artery; RV = right ventricle; RVOT = right ventricular outflow tract; VSD = ventricular septal defect*

Because of these aortic arch anomalies, the patient was scheduled for ductal stent implantation via the antegrade approach. The ascending aorta was reached via the "right atrium through RV through VSD" pathway. A 5F right Judkins catheter was fed through the duct over a 0.035-in hydrophilic guidewire, and a 0.014-in floppy coronary guidewire was inserted through the PDA into the right pulmonary artery. After evaluating the ductal anatomy, we implanted a 3.5 × 15-mm pre-mounted coronary stent (Abbott Vascular, part of Abbott Laboratories; Abbott Park, Ill) in the duct (Fig. 2C). Post-implantation angiography revealed an increase in pulmonary blood flow, and pulse oximetry revealed an increase in oxygen saturation (94%). No complications occurred during the procedure or follow-up. After the procedure, heparin was administered for 24 hours, and aspirin thereafter. After a one-night follow-up, the patient was discharged from the intensive care unit.

In order to reevaluate the aortic arch anatomy and the position of the stent, 256-slice multidetector computed tomographic angiography (CTA) was performed. The CTA results revealed a double aortic arch anomaly, with the right and left SCAs originating from the right arch (Fig. 3A) and the right and left common carotid arteries from the left arch (Fig. 3B). The stented PDA originated 13 mm distal to the left common carotid artery outlet and ended at the main pulmonary artery bifurcation. Cytogenetic analysis revealed microdeletion of the long arm of chromosome 22.

## Discussion

Tetralogy of Fallot is often associated with chromosome 22q11 deletion. The frequency of the association has been reported to be between 6% and 21%. It has also been well documented that TOF often occurs together with anomalies of the aortic arch and its branches, including right aortic arch, and (less frequently) elongation and high positioning of the aortic arch, aberrant origin or isolation of the SCA, isolation of the left pulmonary artery, absence of the ductus arteriosus, and the presence of major aortopulmonary collateral arteries.[11] Momma and colleagues[12] reported a higher incidence of anomalies of the aortic arch and its branches in patients with TOF and 22q11 deletion than in TOF patients who lacked the chromosome deletion: specifically, right aortic arch, high aortic arch reaching as far as the 3rd rib posteriorly and the clavicle anteriorly on frontal angiograms, aberrant origin of the SCA, isolation of the SCA, absence of the ductus arteriosus, and presence of the major aortopulmonary collateral arteries.

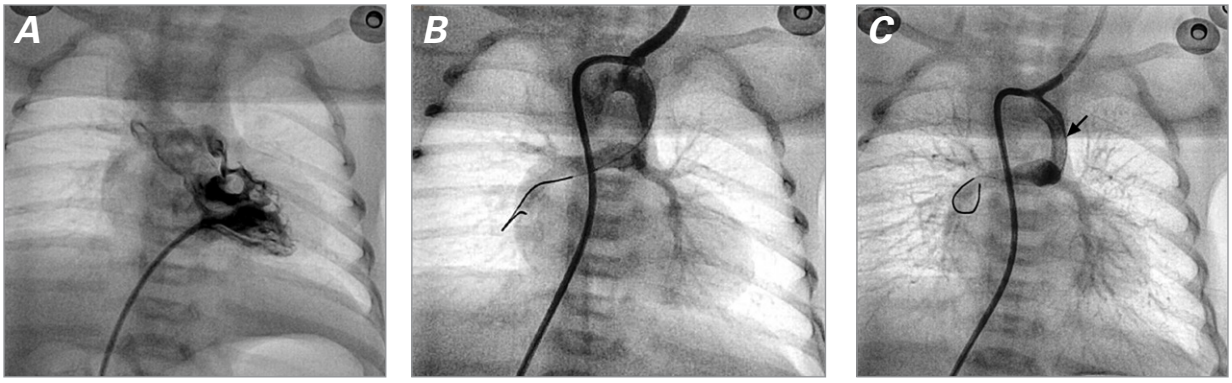


***Fig. 2*** ***A*** *and* ***B****) Before stent implantation, angiograms show patent ductus arteriosus in a neonate with severe tetralogy of Fallot and aortic arch abnormality.* ***C****) The ductal stent (arrow) after implantation.*

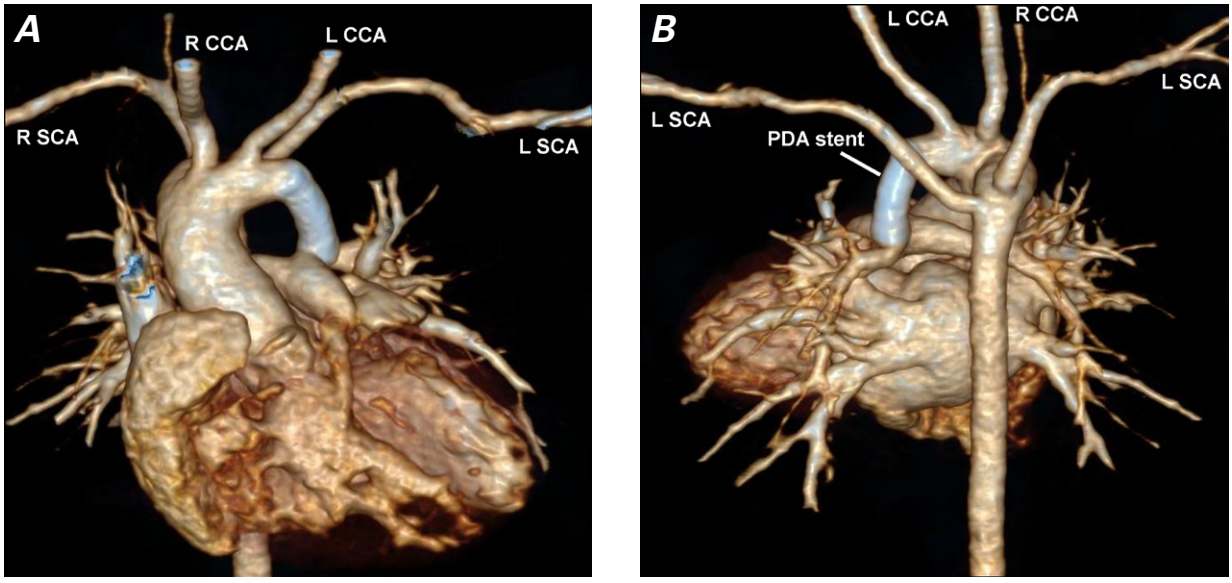


***Fig. 3*** ***A****) Multidetector computed tomographic angiogram shows the aortic arch abnormality before stent implantation. The left aortic arch branches into the common carotid arteries and ends with the patent ductus arteriosus.* ***B****) After stent implantation, the right aortic arch branches into the subclavian arteries and continues into the descending aorta.*

*CCA = common carotid artery; L = left; PDA = patent ductus arteriosus; R = right; SCA = subclavian artery*

In our patient, 256-slice multidetector CTA showed that the right aortic arch branched into the SCAs and continued into the descending aorta, whereas the left aortic arch branched into the left and right common carotid arteries and ended at the PDA. In our judgment, this case is an example of the very rare association of TOF with aortic arch abnormality in chromosome 22q11 deletion. There is no information on how often TOF is seen together with double aortic arch, except for a very few case reports. One of these[13] describes the case of a 3-month-old symptomatic female infant with 22q11 deletion, who was found to have TOF and double aortic arch during preoperative echocardiography and angiography; and another paper[14] reports the case of a 27-year-old symptomatic woman who was diagnosed with TOF and double aortic arch after echocardiography and conventional angiography.

These days, surgery is the definitive treatment for TOF. Although symptomatic patients need surgery, there is some controversy regarding the optimal timing. Some centers continue to offer surgical palliation for neonates and small infants, by constructing an aortopulmonary shunt. In recent years, there has been a resurgence in transcatheter palliation, including balloon dilation and stenting of the RVOT and PDA.[1,6] Ductal stenting to achieve adequate pulmonary artery blood supply in newborns with severe TOF (critical RVOT obstruction or pulmonary atresia) is used as an alternative to a surgical aortopulmonary shunt.[7-10,15] Even today, shunt surgery has notably high morbidity and mortality rates

and can lead to such sequelae as phrenic or vagal nerve paralysis, chylothorax, growth of differential pulmonary artery branches, distortion of pulmonary arteries, pulmonary hypertension, surgical adhesions, shunt occlusion, and stenosis.[16,17] Dirks and associates[18] evaluated 32 patients who had received a modified Blalock-Taussig shunt (median age, 10.5 d) and found acute shunt thrombosis in 3 patients (9%) and chylothorax, phrenic nerve paralysis, necrotizing enterocolitis, and abdominal hemorrhage of unknown origin in one patient each (3%). Noting that 3 of the patients died (9%), the authors pointed out that implantation of the modified Blalock-Taussig shunt has high morbidity and mortality rates, despite its appearance as a safe procedure.

The objective of PDA stenting is to delay or completely avoid aortopulmonary-shunt surgery in neonates and small infants. This option can also reduce shunt-related sequelae like pulmonary artery distortion and differential branch growth, which can increase the rates of morbidity and death in subsequent corrective surgery.[6,11] The specific experience of the center that performs the shunt surgery or PDA stenting is very important. Performed during the neonatal period, these procedures can have very high mortality and morbidity rates in developing countries such as ours. Although PDA stenting might be a desirable alternative to surgery in selected cases, one must consider that stenting provides less durable palliation than does shunt construction and that patients with a ductal stent, therefore, need closer clinical follow-up. The definitive surgical intervention must be performed within 6 to 18 months after PDA stenting.[7]

Ductal stenting via the retrograde femoral approach is technically feasible in cases of tricuspid atresia, pulmonary atresia with intact ventricular septum, and critical pulmonary stenosis with PDA arising from the proximal descending aorta.[7,9,19] However, gaining access to the ampulla is much more difficult if the PDA arises proximally to the aortic arch ("vertical" ductus). In such cases, the catheter can be advanced antegradely, after introduction via femoral vein access.[7] We preferred the antegrade approach because the aortic arch anomaly rendered the PDA nearly inaccessible via retrograde access. Stenting was performed by reaching the base of the aorta through the VSD.

In conclusion, our case shows that antegrade PDA stenting can be an alternative to aortopulmonary-shunt surgery in severely cyanotic newborns with TOF, aortic arch abnormality, and duct-dependent or decreased pulmonary circulation.

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