Tetralogy of Fallot with Absent Pulmonary Valve – A Case Report

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Abstract
A 25 years male patient of Tetralogy of Fallot (TOF) with congenital absent of pulmonary valve (APV) presented with symptoms of palpitation and exertional respiratory distress without congestive heart failure. He underwent successful repair of intracardiac defects. The procedures consisted of patch closure of ventricular septal defect and right ventricular outflow tract reconstruction with a monocusp transannular patch. Resection or plication of dilated pulmonary artery was not required. The patient is doing well without any symptoms.

Key words: Tetralogy of Fallot, Absent pulmonary valve

Introduction
Tetralogy of Fallot (TOF) associated with absent pulmonary valve (APV) is a rare variant that comprises 2-6 % of all patients with Tetralogy of Fallot.1,2 In addition to the typical intracardiac abnormalities associated with Tetralogy of Fallot (Malalignment ventricular septal defect (VSD), stenosis of right ventricular outflow tract (RVOT), right ventricular hypertrophy (RVH) and overriding of aorta, patient with TOF / APV have rudimentary or absent pulmonary valve leaflets with severe pulmonary regurgitation (PR). Most importantly, neonates and infants presenting with TOF / APV commonly have characterstic aneurysmal dilatation of the pulmonary artery and its branches which can cause compression of the tracheobronchial tree expressing as a syndrome called absent pulmonary valve syndrome (APVS).3 Consequently TOF/ APV patients frequently require preoperative ventilatory support and often need early neonatal surgical repair4-6. Surgical treatment of neonates and infants with TOF/ APV has been associated with increased post operative respiratory complications and hospital mortality.7-10

Several modifications of surgical technique have emphasized reduction of dilated pulmonary arteries, establishment of pulmonary valve competency using valve conduits and relief of obstruction of the tracheobronchial tree by means of suspension of the compressing pulmonary arteries, translocation of the pulmonary artery or even segmental resection of the lung10-13. The surgical modification noted above and recent advance in post operative care and ventilatory management of neonates and infants have all contributed to improve surgical outcomes in patients with. We report a successful repair of this rare anomaly in a young adult.

Case Report
A 25 years old Bangladeshi youngman was admitted at Ibrahim Cardiac Hospital & Research Institute for elective surgery for TOF with APV having the past history of exertional respiratory distress and palpitation without heart failure. Auscultation revealed a systolic murmur associated with an early diastolic murmur alone the mid-left sternal border. The electro-cardiogram showed right atrial hypertrophy (RAE), right ventricular hypertrophy (RVH) and complete right bundle branch block (RBBB). The chest roentgenogram demonstrated cardiomegaly with a cardiothoracic ratio of 60 % and pulmonary vascular marking were slightly increased with normal lung fields. Two dimensional echocardiography demonstrated valvular pulmonary stenosis with severe pulmonary regurgitation, large inlet septal VSD with mild aortic regurgitation (AR);

Fig.1.2-D echocardiogram.Parasternal

long-axis view demonstrating a large aortic root overriding a ventricular septal defect with an enlarged right pulmonary artery.
a large aorta overriding the VSD (Fig.-1), cardiac catheteriza-
tion and angiography confirmed the diagnosis of TOF with
absent pulmonary valve and right aortic arch. 90 mm of Hg
systolic pressure gradient across the pulmonary valve annu-
lus noted with pulmonary artery pressure of 26/14 mm of
Hg recorded.

A pulmonary arteriogram demonstrated dilated Rt.
Pulmonary artery (20 mm), a distally displaced stenotic pul-
monary valve annulus (14mm) without pulmonary valve
structure. A right ventriculogram demonstrated dilatation of
right pulmonary artery (RPA) 20 mm (Fig.-2).

In June 2009, Corrective surgery through a median sterno-
tomy was performed. The main pulmonary artery (MPA) was
almost normal size but shorter in length and the pulmonary
valve annulus was very narrow. Cardiopulmonary bypass
was established with bicaval cannulation and the heart was
arrested with antegrade cold blood cardioplegia. Right atri-
otomy (RA-tomy)was done. The pulmonary artery was lon-
gitudinally incised and the incision was extended to the
right ventricular outflow tract. Vestigial remnants of the
leaflets of the pulmonary valve were present in a small ven-
triculo-arterial junction.

Excision of septal and parietal bands were done at in-
fundibulum. The VSD was closed using Dacon patch and
right ventricular obstruction was relieved with insertion of
pericardial monocusp transannular patch. The peak systolic
pressure gradient across the right ventricular outflow tract
(RVOT) came down to 32 mmHg at the end of operation.
The post operative course was uneventful.

Follow up after 6 months by echocardiography revealed –
no residual left to right shunting at ventricular level, the
peak systolic pressure gradient across the right ventricular
outflow tract was 28mmhg, moderate pulmonary regurgita-
tion with mild tricuspid regurgitation. He is well without
any symptoms following 6 months of repair.

Discussion

Congenital absence of pulmonary valve is rare isolatedly
and is more often associated with various heart defects usu-
ally tetralogy of Fallot.14 A combination of TOF with APV is
also rare. Symptoms of absent pulmonary valve vary from
life threatening respiratory obstruction by aneurysmally dilat-
ed pulmonary arteries or cardiac failure in a neonate to
absence of symptoms in patients who may live unrestricted
lives for many years.

The markedly symptomatic patients especially neonate or
infant with pulmonary atelectasis and right ventricular fail-
ure needs aggressive medical treatment.3 Usually the surgi-
cal results in the newborn is poor.4 But if the medical treat-
ment fails, should undergo a corrective operation involving
insertion of a valve or valved conduit in the RVOT and par-
tial resection, plication or both of the aneurysmal pulmonary
arteries.3,15 In contrast , older infants, children and younger
patients having minimal symptoms or asymptomatic, such
in our young adult one, should undergo a corrective opera-
tion electively at the time of diagnosis.3 That is why we
relieved the right ventricular outflow tract (RVOT) obstruc-
tion and pulmonary regurgitation with a pericardial mono-
cusp transannular patch in absence of pulmonary hypertension
in our patient. The insertion of a pulmonary valve pros-
thesis, and resection or plication of the dilated right pul-
monary artery is not essential.

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