

Spectrum of pulmonary valve morphology and its relationship to pulmonary trunk in tetralogy of Fallot

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Abstract

Background: Tetralogy of Fallot (TOF) is a complex congenital heart disease with anatomic variations. Although the pulmonary valve in TOF is abnormal, it has not been studied well, especially on newer imaging modalities such as multidetector computed tomography (CT), which gives excellent anatomic detail. **Aims:** The aim of this study was to assess the morphology of pulmonary valve in TOF on CT and evaluate its association with the degree of hypoplasia of infundibulum and pulmonary trunk. **Materials and Methods:** The cardiac CT scans of 30 patients with TOF were reviewed to evaluate the morphology of the pulmonary valve, infundibulum, and pulmonary arteries. Fisher's exact test was performed to examine the association between pulmonary valve morphology and degree of hypoplasia of the infundibulum and pulmonary trunk. **Results:** 16.7% of patients with TOF had pulmonary atresia. The prevalence of tricuspid, bicuspid, and absent valves were 10%, 53.3% and 6.7%, respectively. In another 13.3% of patients, although valve tissue was present, exact morphology could not be determined on CT. The commissures of 62.5% of the bicuspid valves were at 12 o'clock and 6 o'clock or slightly off the midline. There was statistically significant association between valve morphology and degree of infundibular hypoplasia ($P < 0.001$) and calibre of pulmonary trunk ($P < 0.001$). **Conclusion:** Morphological abnormality of the pulmonary valve is common in TOF. The most common type of pulmonary valve in TOF patients is bicuspid valve with commissures at 12 o'clock and 6 o'clock or slightly off the midline. Fewer cusps of the pulmonary valve are associated with a more severe degree of pulmonary artery hypoplasia.

Key words: Artery; hypoplasia; pulmonary; tetralogy of Fallot; valve

Introduction

Tetralogy of Fallot (TOF) is the most common congenital heart disease with an incidence of 3.01 per 10000 live births,^[1] and is characterized by a large ventricular septal defect (VSD), overriding aorta, obstruction of the right ventricular outflow tract, and right ventricular hypertrophy. The right ventricular outflow obstruction could be subvalvular, valvular, supra-valvular, or at the

level of the pulmonary arterial branches.^[2] The pulmonary valve morphology in patients with TOF is commonly abnormal, and most data in literature has been obtained from the study of surgical heart specimens during autopsy, which could be biased due to different survival rates of patients with various anatomic variations. Because of the current trends towards valve sparing techniques and native valve cusp augmentation techniques to reduce the

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incidence of pulmonary regurgitation and its long-term complications following corrective TOF repair,^[3,4] preoperative awareness of pulmonary valve morphology could contribute towards better surgical planning. Multidetector cardiac computed tomography (CT) gives good anatomic detail of the cardia, its vascular structures, and valves. The purpose of this study was to evaluate the pulmonary valve morphology and to assess its association with the degree of hypoplasia of infundibulum and pulmonary trunk in TOF.

Materials and Methods

This was a retrospective study of 30 patients with combined clinical and echocardiographic diagnosis of TOF who were referred for cardiac CT between November 2014 and August 2016.

All the CT scans were performed in a 64 slice CT scanner (General Electric Medical Systems; Discovery CT 750HD) with retrospective electrocardiographic (ECG) gating. Nonionic contrast agent, calculated according to the weight of patient, with a maximum dose of 2 ml/kg was injected at a rate of 2.5 ml/s into a peripheral vein. The scan was triggered at contrast density of 200 HU in the pulmonary trunk. The exposure factors were optimized for the pediatric age. The scanning parameters were gantry rotation time of 350 ms, pitch of 0.24, tube potential of 80–100 kVp, tube current of 300 mAs, and field of view of 25 cm. The acquired images were reconstructed at 10% interval of R-R and then transferred to a separate workstation (AW Server 2.0, General Electric Medical Systems) for multiplanar reconstruction.

The CT images were reviewed for the morphology of pulmonary valve, infundibulum, pulmonary trunk, right pulmonary artery, and left pulmonary artery as described below.

Pulmonary valve: The size of the pulmonary annulus, the number of cusps, the number and location of commissures, and the number of raphe were noted. Each pulmonary valve was categorized as quadricuspid, tricuspid, bicuspid, unicuspid, absent, atretic, or indeterminate. The pulmonary valve was considered atretic if there was tissue interposed between the infundibulum and the main pulmonary artery. The pulmonary valve morphology was considered indeterminate if there was definite valve tissue present, which could not be categorized into typical quadricuspid, tricuspid, bicuspid, or unicuspid morphology on CT.

Infundibulum: Infundibular stenosis was categorized as none, mild-to-moderate, severe, or atretic based on the expected size of the infundibulum for that patient, taking into account the narrowest portion of the infundibulum.^[5]

Pulmonary arteries: The calibre of pulmonary trunk was categorized into normal, mild-to-moderate hypoplasia, severe hypoplasia, atretic, or dilated based on the comparison with the calibre of adjacent ascending aorta.^[5] The pulmonary trunk was considered normal if the calibre was more than 80% of the aorta. The pulmonary trunk was considered mild-to-moderately hypoplastic if the calibre was between 50 and 80% of that of aorta. The pulmonary trunk was considered severely hypoplastic if the calibre was less than 50% of the aortic calibre. The pulmonary trunk was termed atretic if there was no lumen. The pulmonary trunk was termed dilated if the calibre was more than that of aorta. The right and left pulmonary arteries were also assessed for any coexisting stenosis.

Statistical analysis

Fisher's exact test was performed to examine the association of pulmonary valve morphology and the degree of hypoplasia of the infundibulum and pulmonary trunk in these patients.

Results

A total of 30 patients with TOF underwent cardiac CT between November 2014 and August 2016. All the CT studies were technically adequate to assess the morphology of the infundibulum, pulmonary valve, and pulmonary arteries. There were 24 boys (80%) and 6 girls (20%) with a median age of 4.7 years.

Pulmonary valve: The most common pulmonary valve morphology in TOF patients were bicuspid valves (53.3%). There was pulmonary atresia [Figure 1A and B] in 16.7% of the TOF patients. In 6.7% of the patients, the pulmonary valves were absent, whereas 10% had tricuspid valves [Figure 2]; but none had unicuspid or quadricuspid valves. 13.3% had indeterminate valve morphology. Of the 16 patients with bicuspid valves, 6 (37.5%) had commissures located at 3 o'clock and 9 o'clock or slightly off the midline [Figure 3A], and 10 (62.5%) had commissures located at 12 o'clock and 6 o'clock or slightly off the midline [Figure 3B and C]. 81% of bicuspid valves did not have a median raphe and 19% had one median raphe [Figure 3D].



Figure 1 (A and B): Reconstructed oblique sagittal view (A) and oblique axial view (B) of cardiac CT showing atresia of the pulmonary trunk (short arrows) in TOF

It was found that 56% (14 patients) of TOF patients without pulmonary atresia had mild-to-moderate pulmonary annular stenosis and 40% (10 patients) had severe pulmonary annular stenosis. One patient had normal pulmonary annulus. Of these patients with severe annular stenosis, 6 had bicuspid valves, 1 had tricuspid valve, and in 3 patients, the valve morphology was indeterminate.

Infundibulum: Of the patients in this study, 18 (60%) had severe infundibular stenosis and 3 (10%) had mild-to-moderate infundibular stenosis. Five patients (16.7%) had atresia of the infundibulum, and 4 (13.3%) were found to have a normal infundibulum. All the cases had anterior displacement of the outlet septum. A total of 73.3% had both anterior infundibular wall hypertrophy and hypertrophy of trabecula septomarginalis, 10% had isolated anterior infundibular wall hypertrophy, 6.7% had isolated hypertrophy of trabecula septomarginalis, and 10% had isolated displacement of outlet septum.

Table 1 provides information regarding the type of pulmonary valve and the degree of infundibular hypoplasia. There was significant association ($P < 0.001$) between pulmonary valve morphology and the degree of infundibular stenosis using Fisher's exact test.

Table 1: Relationship between the type of pulmonary valve and degree of infundibular stenosis

Infundibulum pulmonary valves	Normal	Mild to moderate stenosis	Severe stenosis	Atresia	Total
Tricuspid	1	0	2	0	3
Bicuspid	1	3	12	0	16
Indeterminate	0	0	4	0	4
Unicuspid and quadricuspid	0	0	0	0	0
Absent	2	0	0	0	2
Atretic	0	0	0	5	5
Total	4	3	18	5	30

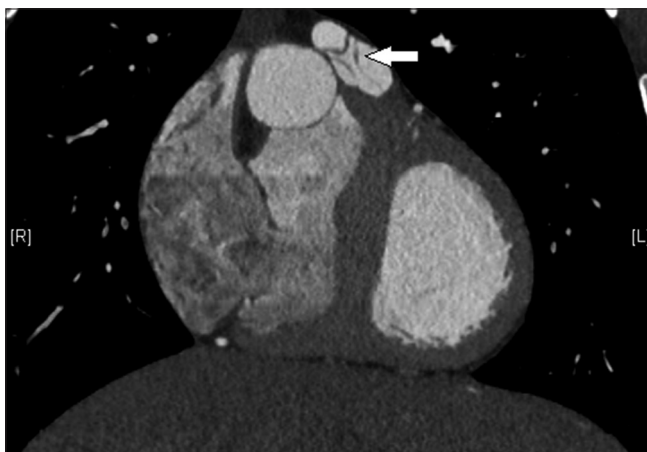


Figure 2: Reconstructed oblique coronal view of cardiac CT showing three cusps (arrow) of the pulmonary valve in cross section

Infundibular and pulmonary valve atresia were found to coexist, as expected. The 2 patients with absent pulmonary valves did not have any infundibular stenosis. Majority of the TOF patients without atresia had severe infundibular stenosis.

Pulmonary artery: Twelve patients had severe pulmonary trunk hypoplasia (40%), 10 patients had mild-to-moderate pulmonary trunk hypoplasia (33.3%), 5 had pulmonary trunk atresia (16.7%), 2 had aneurysmal dilation of pulmonary arteries (6.7%), and 1 had normal pulmonary trunk (3.3%).

Table 2 provides information regarding the type of pulmonary valve and the degree of pulmonary trunk hypoplasia. There was statistically significant association between pulmonary valve morphology and degree of hypoplasia of the pulmonary trunk ($P < 0.001$). Of the 16 patients who had bicuspid valves, 56.3% had severely hypoplastic pulmonary trunk and 43.8% had mild-to-moderately hypoplastic pulmonary trunk. Of the 4 patients with indeterminate morphology, 3 had severe hypoplasia and one had mild-to-moderate hypoplasia. Patients with tricuspid valve had normal or mild-to-moderate hypoplasia of the pulmonary trunk.

Hypoplasia of the pulmonary trunk was also associated with hypoplasia of the central right and left pulmonary artery branches. Two patients with absent pulmonary valves had mild-to-moderate hypoplasia of the pulmonary annulus and aneurysmal dilatation of pulmonary trunk and its first order branches [Figure 4].

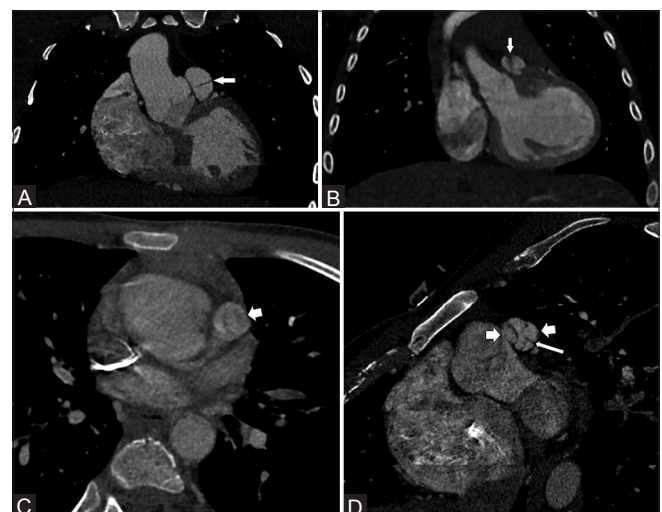


Figure 3 (A-D): (A) Reconstructed coronal view of cardiac CT showing bicuspid pulmonary valve with commissures at 3 o'clock and 9 o'clock or slightly off the midline (arrow). (B) Reconstructed coronal view of cardiac CT in another patient showing bicuspid pulmonary valve with commissures at 12 o'clock and 6 o'clock or slightly off the midline (arrow). (C) Reconstructed oblique axial CT view showing open bicuspid valve (arrow). (D) Open bicuspid pulmonary valve (short arrows) with one of the cusps showing median raphe (long arrow)

Table 2: Relationship between the type of pulmonary valve and calibre of pulmonary trunk

Pulmonary trunk pulmonary valves	Normal	Mild-to-Moderately hypoplastic	Severely hypoplastic	Atresia	Dilation	Total
Tricuspid	1	2	0	0	0	3
Bicuspid	0	7	9	0	0	16
Indeterminate	0	1	3	0	0	4
Unicuspid and quadricuspid	0	0	0	0	0	0
Absent	0	0	0	0	2	2
Atretic	0	0	0	5	0	5
Total	1	10	12	5	2	30

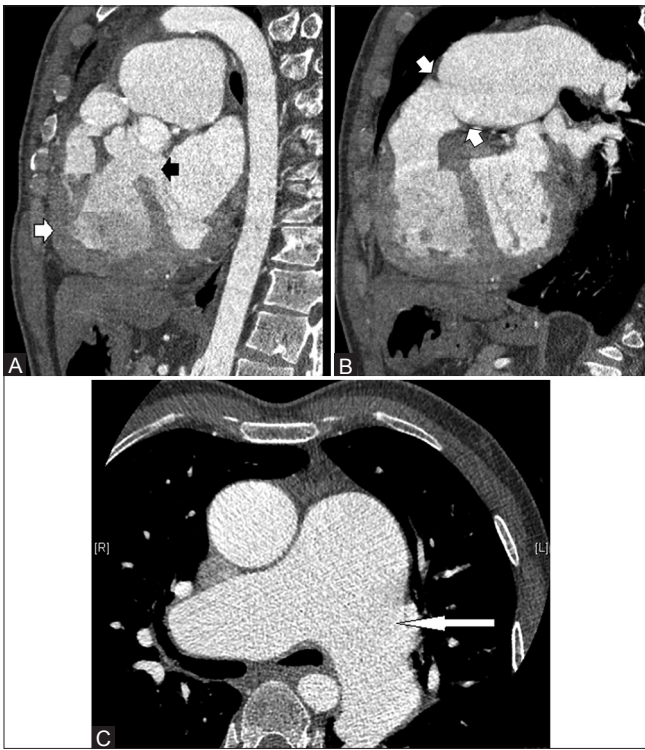


Figure 4 (A-C): Cardiac CT scan images of a patient with TOF and absent pulmonary valve. (A) Subaortic ventricular septal defect (short black arrow), overriding of aorta and right ventricular hypertrophy (short white arrow) shown in reconstructed oblique sagittal view. (B) Ring like constriction suggestive of stenosis (arrow) seen at the level of pulmonary annulus and absent pulmonary valve seen in another section of reconstructed oblique sagittal view. (C) The pulmonary trunk and its first order branches are aneurysmal (long white arrow)

Discussion

Congenital abnormality of the pulmonary valves are uncommon. Congenital pulmonary valve abnormalities are usually associated with congenital cardiac abnormalities such as TOF. On transthoracic echocardiography, assessment of the pulmonary valve anatomy is suboptimal as short axis views of the pulmonary artery are not clearly obtained.^[6] Hence, most studies of pulmonary valve anatomy have been performed on surgical heart specimens from autopsies. With the advent of cardiac CT with its multiphase images, it is possible to perform a detailed assessment of the cardiovascular structures including the pulmonary valve. To our knowledge, this is the first study

assessing pulmonary valve morphology in TOF patients using cardiac CT. We also assessed the relationship of pulmonary valve morphology and degree of hypoplasia of the infundibulum and pulmonary trunk in these patients.

Knowing the conventional and newer surgical repair techniques performed in TOF helps to understand the clinical relevance of the anatomy of the pulmonary valve. The traditional two-stage repair of TOF was composed of an early temporary arteriopulmonary shunt to increase the pulmonary blood flow, which was followed by a corrective repair. Because there is significant mortality associated with preoperative shunting, the current surgical trend is towards a corrective repair early in life. Conventional repair includes correction of the ventricular septal defect through right ventriculotomy, relief of right ventricular outflow tract obstruction using a pericardial patch, and if required, commissurotomy, valvotomy, and or transannular patch. However, right ventriculotomy and transannular patch leads to complications of pulmonary insufficiency, right ventricular functional deterioration, and arrhythmias, which could even lead to sudden death after repair. Hence, the current focus is the transatrial approach for VSD repair to reduce arrhythmias and pulmonary valve sparing and pulmonary valve augmentation techniques to reduce the incidence of pulmonary insufficiency. Pulmonary valve sparing technique is used if the diameter of the pulmonary annulus is within two standard deviations of the normal predicted for the patient's body surface area. If not, transannular patch and pulmonary valve cusp augmentation is performed using autologous pericardium to reduce the incidence of pulmonary insufficiency after repair. For augmentation of the pulmonary cusp, an incision on the valve is made depending on the location of the commissures. If the valve is bicuspid with commissures at 3 o'clock and 9 o'clock, the incision is made in the midline of the anterior cusp. If the valve is bicuspid with commissures at 12 o'clock and 6 o'clock or slightly off the midline, the valve is divided at or near the commissures to preserve as much as valvular tissue as possible.^[7]

In our study, bicuspid valve was found to be the most common, as was also reported by Altrichter *et al.* in their set of 61 patients with TOF.^[8] We also found that the commissures were located at 12 o'clock and 6 o'clock or

slightly off the midline in a majority (64%) of the patients with bicuspid valve, unlike Anagnostopoulos *et al.* who observed intraoperatively that most of the TOF patients had commissures at 3 o'clock and 9 o'clock.^[7]

Our study showed statistically significant association between the morphology of pulmonary valve and the degree of infundibulum and pulmonary trunk. Lesser number of cusps in the pulmonary valve showed a trend towards severe pulmonary artery hypoplasia. Other than patients with absent pulmonary valves who did not have infundibular stenosis, most others had severe infundibular stenosis or atresia irrespective of the valve morphology. These findings are similar to a study of 85 surgical heart specimens of TOF patients by Satyanarayana *et al.*^[5] Anterior displacement of outlet septum is the primary cause for infundibular stenosis in TOF. Infundibular stenosis is also accentuated by hypertrophy of anterior wall of infundibulum and hypertrophy of trabecula septomarginalis.^[9] All our cases had anterior displacement of the outlet septum. Majority of our patients also had both hypertrophy of anterior wall of infundibulum and hypertrophy of trabecula septomarginalis.

Absence of the pulmonary valve is rare in TOF. Unlike others, TOF patients with an absent pulmonary valve show aneurysmal dilatation of the pulmonary trunk and branch pulmonary arteries,^[10] with mild stenosis at the level of the pulmonary annulus. This was seen in two of the patients in this study. Both these patients had normal-sized infundibulum. The aneurysmal pulmonary artery is known to cause tracheobronchial compression^[10] and severe obstructive airway disease in these cases.

Our study is limited by a relatively small number of patients. In 4 of the 30 patients, although pulmonary valve tissue was seen on cardiac CT, it could not be categorized into any typically described valve morphology types and were termed indeterminate.

Conclusion

Morphological abnormality of the pulmonary valve is common in TOF. The most common type of valve in TOF

patients is bicuspid valve with commissures at 12 o'clock and 6 o'clock or slightly off the midline. The pulmonary valve morphology is significantly associated with the degree of hypoplasia of infundibulum and pulmonary trunk. Fewer cusps of the pulmonary valve is associated with a more severe degree of pulmonary artery hypoplasia.

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Conflicts of interest

There are no conflicts of interest.

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