

INTRAOPERATIVE MORPHOLOGY OF PULMONARY VALVE IN TETRALOGY OF FALLOT PATIENT

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ABSTRACT

Background: TOF is the most common cyanotic congenital heart disease. Intracardiac morphology differs in each patient. Morphology of pulmonary valve is usually abnormal, but it is not studied in detail. The aim of this study was to assess the pulmonary valve morphology intraoperatively in TOF repair patients. **Materials and Methods:** This is a retrospective study which included 198 surgically repaired TOF patients from August 2017 to August 25 in a tertiary care hospital. In all patients the pulmonary valve leaflet morphology was assessed in view of tricuspid valve, bicuspid valve with anteroposterior commissure, bicuspid valve with lateral commissure, and unicuspid valve anatomy. **Results:** A total of 198 patients were operated in the study period. Tricuspid pulmonary valve leaflet was found in 36 (18%) patients, bicuspid pulmonary valve leaflet with 6-12 O'clock commissure was seen in 52 (26%) patients, bicuspid pulmonary valve leaflet with 3-9 O'clock commissure in 107 (53%) Patients, and unicuspid pulmonary valve was seen in 3 (1.51%) patients. **Conclusion:** Abnormal pulmonary valve morphology is common in TOF congenital heart disease. The most common type is bicuspid pulmonary valve with 3 – 9 O'clock Commissure.

INTRODUCTION

TOF is the most common cyanotic congenital heart disease,^[1,2] in children with incidence of 3.01 per 10,000 live birth.^[3] It is caused by anterior, leftward, and cephalic deviation of infundibular septum during the embryological development of the heart as proposed by Van Praagh resulting into characteristic features of TOF which include a large mal-aligned subaortic VSD, aortic overriding, right ventricular outflow tract obstruction, and right ventricular hypertrophy. Right ventricular outflow tract obstruction can occur at any level starting from infundibulum, pulmonary annulus, pulmonary valve, main pulmonary artery, branch pulmonary artery, and peripheral pulmonary artery branch level.^[4] The intracardiac anatomy in TOF patients is not similar in every patient and pulmonary valve morphology usually differs in patient to patient.

The morphology of pulmonary valve decides the type of repair in pulmonary valve stenosis like commissurotomy, valve sparing surgery, or trans-annular patch repair technique.

MATERIALS AND METHODS

This study was conducted in retrospective manner and included 198 patients operated between August 2017 to August 2025 in a single center i.e. SMS Hospital Jaipur, Rajasthan, India, and by single surgeon.

Medical records consisting of history, physical examination, chest x ray, ECG, 2D echocardiography, intraoperative notes, perfusion notes, ICU and ward postoperative data of all patients were thoroughly studied and all required data were collected.

A total of 198 patients were included in this study. Patients were followed up to 1 year after surgery and outcome was assessed.

Data consisted of: Age at surgery, weight at surgery, gender ratio, echocardiographic and intraoperative findings regarding size of pulmonary annulus, size of main pulmonary artery and branch pulmonary arteries, status of infundibulum, status of pulmonary valve leaflets and commissures, commissurotomy, type of repair, post repair pressure ratio of right ventricle to left ventricle, extubation time, inotrope requirement were recorded. Z – Score was used to assess adequate size of pulmonary valve annulus,

main pulmonary artery and branch pulmonary arteries.

Total stay in ICU after surgery, rhythm disturbances, bleeding, re exploration for bleeding or incomplete repair, high gradient across the right ventricular outflow tract (> 40 mm Hg) or right ventricular dysfunction, heart block, requirement of temporary and permanent pacing, residual VSD, postoperative ventilation, sternal wound infection and dehiscence, chylothorax, seizure, neurological deficit, renal failure, status of diaphragm, and total stay in hospital after surgery were noted.

Each pulmonary valve was categorized as Tricuspid, bicuspid with 3-9 O'clock commissure, bicuspid with 6-12 O'clock commissure, unicuspid, and dysplastic valve leaflet.

Surgical technique: The technique described here was used in all patients with no deviations. Experience of single surgeon was included in this study.

The median sternotomy approach was used in all patients. Patient was taken on cardiopulmonary bypass with aortic and selective bicaval cannulation after ACT crossing 480 seconds. After cross clamping the aorta antegrade root Del Nido cardioplegia was given and patient was cooled to 30 degrees Celsius with hematocrit maintained at 30%. CUF was used during bypass and MUF was done after coming off bypass in patients less than 10 kg weight. PDA was ligated in all patients. Right atrium was opened after snaring SVC and IVC. Main pulmonary artery was opened vertically along with right atrium in all patients. Left ventricle was vented through existing PFO or ASD or created PFO. After opening the pulmonary artery required hegar dilator was used to assess size of right and left pulmonary artery. If required, the pericardial patch was extended to enlarge the size of hypoplastic branch pulmonary artery. Pulmonary valve was inspected carefully regarding number of leaflets, type of commissures, commissural stenosis, and size of pulmonary annulus. Pulmonary valve commissurotomy along with removal of commissural tethering was done where stenosis was present. Deviated infundibular septum along with hypertrophic parietal bands in right ventricular outflow tract were excised. Adequate size hegar dilator according to z-score of individual patients was passed across the pulmonary annulus. If actual annulus is smaller than required annulus, then pulmonary arteriotomy was extended across the annulus for approximately 5 mm proximal to annulus. In case of the tricuspid pulmonary valve and bicuspid pulmonary valve with lateral commissure the incision was extended vertically downward through the anterior sinus. In bicuspid pulmonary valve with anteroposterior commissure, the incision was extended through the anterior commissure to enlarge the pulmonary annulus. Pulmonary artery and annulus were enlarged with untreated pericardial patch with continuous prolene suture. Only one patient required right ventriculotomy.

VSD was closed with savage patch with prolene suture in continuous manner. Additional small muscular VSDs were closed with interrupted pledgeted prolene suture whenever required. SAM was excised when it was present. PFO was closed with direct repair with continuous prolene suture and ASD was closed with untreated pericardial patch with prolene suture in continuous fashion. Right atrium was closed in single layer with continuous prolene suture after checking for tricuspid regurgitation with saline insufflation test.

Aortic cross clamp was removed after proper de-airing of heart. All patients came off bypass in sinus rhythm with minimal to moderate inotropes. After coming off bypass pressure in left ventricle, right ventricle, right ventricular outflow tract and main pulmonary artery were assessed.

3 patients required temporary pacing, for first degree heart block in one patient and 2:1 block in 2 patients. 2 patients developed junctional ectopic tachycardia which responded to medical management. Sinus rhythm was achieved within 48 hours after surgery. Patients were shifted to ICU with stable hemodynamics.

Postoperative 2D echocardiography was done at the time of discharge from hospital, at 6 month and 1 year follow up.

RESULTS

Most common age of surgery at this center was between 1 – 5 years (n=95, 48%) followed by age group of 5-10 years (n=52, 26%) (Table 1).

In this series it was found that males are nearly 2.5 times more commonly affected than females (72% vs 28%). (Table 2)

In majority of cases (n=151) transatrial-transpulmonary artery approach was used (76%) and pulmonary valve was preserved while in 24% cases (n=47) transannular repair was done (Table 3). In all cases, hypoplastic pulmonary artery was enlarged with untreated pericardial patch to match the Z score. Patch was extended to left pulmonary artery in 3 cases.

In this series the most common pulmonary valve morphology appeared to be bicuspid pulmonary valve with 3-9 O'clock commissure (n=107, 53%) followed by bicuspid valve with 6-12 O'clock commissure (n=52, 26%). Tricuspid valve anatomy was seen in 36 cases (18%) and unicuspid valve was seen in 3 cases (Table 4). So, overall, bicuspid morphology is more common than tricuspid valve morphology. Out of 159 cases of bicuspid valve morphology, only 2 patients had no valvar stenosis while in 36 cases of tricuspid valve anatomy, 17 patients had no valvar stenosis. Therefore, it appears that bicuspid morphology is associated more with valvar stenosis.

92.92% (n=184) were extubated within 3 to 4 hours after surgery and 10 patients (5.05%) had longer extubation time of 7 to 8 hours because of slight delay

in gaining consciousness and muscle power. 3 patients (1.51%) required re exploration for bleeding and extubated 5-6 hours after exploration. One patient required re exploration for right ventricular dysfunction and PFO was created.

93 patients (46.96%) were shifted out of ICU on 4rd postoperative day and 82 patients (41.41%) on 5th day. 9 patients (5.7%) required 6 to 7 days in ICU because of increased drain output (n=4), collapsed lung (n=2), and fever (n=3).

Only 2 patients required temporary pacing (1.01%) for 2: 1 heart block (Table5). All these patients regained sinus rhythm within 48 hours of surgery. None of the patients required permanent pacemaker insertion. Junctional ectopic tachycardia was seen in 3 patients (1.51%) which responded well to amiodarone therapy and sinus rhythm was achieved before discharge.

5 patients (2.52%) had residual VSD which are less than 3 mm and none required surgical re-exploration for altered hemodynamics. During follow-up it was found that all these VSDs were closed by itself.

No patient was re-explored for sternal wound infection or sternal dehiscence. Although some patients developed hypertrophic scar and keloid which were treated with intradermal triamcinolone injection and responded well.

Prolonged hospital stay of more than 7 days was seen in 8 patients (4.04%) while 67% (n=132) were discharged on 5th day after surgery and 29.29% (n=58) patients on 6th day of surgery.

Mortality was seen in 12 patients (6%), three patients had excessive endotracheal bleeding for unknown reason, 2 patients had hematuria with renal failure, 4 patients succumbed to septicemia, and 3 patients developed low cardiac output syndrome and multiorgan failure.

Table 1: Age wise distribution

S.No.	Age in years	Number	Percentage (%)
1	Less than 1 year	4	2
2	1 – 5	95	48
3	5 – 10	52	26
4	10 – 15	27	14
5	15 – 20	4	2
6	>20	16	8
Total		198	100

Table 2: Gender wise distribution

S.No	Gender	Number	Percentage (%)
1	Male	142	72
2	Female	56	28
total		198	100

Table 3: Type of repair

S.No.	Type of repair	Number	Percentage (%)
1	Transatrial valve preserving	151	76
2	Transannular patch repair	47	24
Total		198	100

Table 4: Morphology of pulmonary valve

S.No.	Type of pulmonary valve	Number	Percentage (%)
1	Unicuspid valve	3	1.5
2	Bicuspid valve with anteroposterior commissure	52	26.26
3	Bicuspid valve with lateral commissure	107	54.04
4	Tricuspid valve	36	18.18
		198	100

Table 5: Postoperative complications

S.No.	Complication	Number	Percentage (%)
1	Reintubation	1	0.5
2	Re exploration for bleeding	2	1.01
3	Residual VSD	5	2.52
4	Re exploration for significant residual VSD	0	0
5	Re exploration for right ventricular dysfunction	1	0.5
6	Transient heart block	2	1.01
7	Permanent pacemaker	0	0
8	Arrhythmia	1	0.5
9	Sternal wound infection	0	0
10	Sternal dehiscence	0	0
11	Chylothorax	0	0
12	Diaphragmatic paralysis	0	0
13	Renal failure	0	0
14	Neurological deficit	0	0
15	Mortality	12	6

Table 6: Relation of morphology of pulmonary valve with type of repair

S.No.	Type of pulmonary valve	Transartrial Valve preserving surgery (%)	Transannular patch repair (%)
1	Unicuspid valve	2 (1.01)	1 (0.5)
2	Bicuspid valve with anteroposterior commissure	41 (20.70)	12 (6.06)
3	Bicuspid valve with lateral commissure	83 (41.91)	23 (11.61)
4	Tricuspid valve	24 (12.12)	12 (6.06)
Total		150	48

DISCUSSION

Congenital anomaly of the pulmonary valve is not uncommon in TOF patients. Anatomy of pulmonary valve leaflets and annulus affect the complete repair of TOF. In this series, all patients were operated with trans right atrial – pulmonary artery approach. None of the patients required a separate incision on right ventricular outflow tract.

Whenever anteroposterior commissure was present, the commissurotomy was done in 12 O' clock and 6 O' clock position as done by Anagnostopoulos et al,^[5] and extended across the annulus through the 6 O' clock commissure if required for transannular repair.

In case of lateral commissure, if transannular patch is required, then anterior leaflet is incised in midline and extended across the annulus till the required annulus diameter is achieved.

Pulmonary valve sparing technique was used if annulus is of adequate size i.e. within 2 standard deviation of normal predicted for the patient's body surface area. If annulus was inadequate, then trans annular patch repair was done with incision extending into right ventricular outflow tract approximately 5 mm upstream the annulus.

In this study, bicuspid valve was the most common type of pulmonary valve anatomy. The similar results were also reported by Altrichter et.al,^[6] Vida et al,^[7] and Jiang X et al.^[8]

In a series by Su Jin Choi et al,^[9] 77% patients had overall bicuspid valve morphology, 16% had unicuspid valve and only 6.4 % had tricuspid valve morphology while in our study incidence of bicuspid valve was nearly similar (79%) but tricuspid valve constituted 18% and unicuspid valve 1.5% only.

The 3-9 O'clock Type of commissure with bicuspid valve was more common than 6-12 O'clock commissure with bicuspid valve in this study (53% vs 26%) as also reported in other studies,^[5,10] but study by Binita R Chacko et al found that bicuspid valve with 12-6 O' clock commissure was more common than 3-9 O' clock commissure.^[11]

Satyanarayana et al,^[12] found that morphology of pulmonary valve and degree of infundibulum and pulmonary trunk hypoplasia are interrelated. They showed that lesser number of cusps in pulmonary valve were associated with more pulmonary artery hypoplasia i.e. number of cusp is inversely related to degree of pulmonary artery hypoplasia. In this study this relationship was not studied but it was found that pulmonary artery was universally hypoplastic in all cases and needed enlargement.

Robert D Stewart et al,^[13] showed that presence of tricuspid pulmonary valve was an indicator of good outcome, significantly larger pulmonary annuli, lower Pressure ratio of right ventricle to left ventricle, and less incidence of pulmonary regurgitation. They could preserve pulmonary valve in all of the tricuspid valve morphology in their series as compared to valve preservation in 74% cases of bicuspid valve morphology. In this study, we could also preserve the pulmonary valve in approximately 78% cases of bicuspid valve but in tricuspid valve cases 67% patients had valve preserving surgery not 100% (Table 6). Our study also showed that out of 36 cases 17 patients (47%) of tricuspid valve morphology had no stenosis of pulmonary valve while in bicuspid valve morphology with lateral commissure only 2 patients had no stenosis rest 105 had valvar stenosis and all 52 cases of bicuspid valve with anteroposterior commissure required commissurotomy.

CONCLUSION

Anatomic variations in the pulmonary valve in tetralogy of Fallot are not unusual and bicuspid morphology is more common than tricuspid valve morphology. Bicuspid valve with 3-9 O' clock commissure is the most common variant and bicuspid valve morphology itself is associated with stenosis at valvar level than tricuspid valve.

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