Echocardiographic Characteristics of Children under 16 Years Old with Tetralogy of Fallot Operated at Cho Ray Hospital

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ABSTRACT:

Objective: Describe the echocardiographic characteristics of patients under 16 years old with tetralogy of Fallot before total repair surgery at Cho Ray Hospital.

Subjects and methods: Describe retrospective case series of echocardiographic characteristics of children under 16 years old with tetralogy of Fallot before total repair surgery, from February 2023 to February 2024.

Results: 23 patients were included in the study from February 2023 to February 2024. The median age of patients was 10.5 months (5 – 34.5). Female is 65.2%. Median weight is 7.8 kg (6 – 9.5). Median SpO_2 was 80.0% (70.0 – 88.0). The small left ventricle accounted for a high proportion of 56.5%. Right ventricular dilatation at the base, mid, and long diameters accounted for 30.4%, 39.1% and 30.4%, respectively. 69.6%, 56.5%, 21.7%, 8.7% of patients had pulmonary valve ring, pulmonary trunk, right pulmonary artery, left pulmonary artery small, respectively. Medium and large membranous ventricular septal defect accounted for 65.2% and 34.8%, respectively. The median aortic overriding percentage of patients was 30.0% (30.0-45.0). 100.0% of patients had right ventricular hypertrophy. Most patients had stenosis at all three locations below the valve, at the valve, and above the pulmonary valve, accounting for 52.2%. The most common associated defect was patent ductus arteriosus (69.6%). Echocardiography was quite accurate in diagnosing patient characteristics when compared with surgical reports. 8.7% of patients had inaccurate assessment of pulmonary valve leaflet characteristics.

Conclusion: Echocardiography is non-invasive, safe, has good correlation with intraoperative characteristics, and can be thoroughly applied at all times before, during, and after surgery. Improved echocardiographic methods are needed to overcome the limitations in assessing pulmonary valve characteristics in tetralogy of Fallot.

***Keywords*:** Tetralogy of Fallot, Echocardiography, Children under 16 years old, Before total repair surgery.

1. Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect, accounting for 5.0-10.0% of congenital heart disease cases(1). The disease consists of four main lesions: right ventricular outflow tract stenosis (RVOT), ventricular septal defect (VSD), aorta (AOR) riding on the interventricular septum, right ventricular hypertrophy (RV). TOF

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is a serious disease, if left untreated, about 70.0% of children with TOF die before the age of 10, if completely repaired surgically, children can live like normal people(2,3). Many anatomical features on echocardiography such as left ventricular diameter (LVD), right ventricular diameter (RVD), valve ring, body and two branches of the pulmonary artery (PA)... play an important role in indicating temporary or complete repair of TOF. Domestic and international reports describing the echocardiographic characteristics of TOF patients before total repair surgery are not yet complete and detailed, focusing on the characteristics during surgery as well as the postoperative results. Author Phan HV(4) 2005, mentioned the changes in the shape of the two ventricles, RVOT before and after total repair surgery for TOF, but it is not clear whether the above values were within normal limits before surgery. Author Nguyen TTL(5) 2009, did not mention the value of LVD, RVD, the valve ring, the body and the two branches of PA. Recent studies on TOF also do not describe these values in detail, such as the studies of Nguyen TLA(6) 2017, Choi SJ(7) 2020, Luong TNH(8) 2021, Cao DK(9) 2021. The challenges are obvious: whether the abnormal TOF characteristics that the surgeon obtains through preoperative echocardiography are consistent with the results during surgery. Because choosing the wrong time for complete repair surgery can have a negative impact on the patient(10). If the patient does not receive surgery early, the mortality rate will increase over time (25.0% in the first year, 40.0% at 3 years old, 70.0% at 10 years old, 95.0% at 40 years old), increasingly severe cyanosis, heart failure, progressive malnutrition, polycythemia, stroke and brain abscess(2,3). When there is a connection between the preoperative echocardiography features of total TOF repair and the features detected during surgery, the surgeon will have a plan for preoperative and postoperative evaluation, treatment, and follow-up.

2. Subjects and Methods

Research subjects:

All patients with Tetralogy of Fallot under 16 years old undergoing complete repair surgery at Cho Ray Hospital from February 2023 to February 2024.

Selection criteria:

Patients with tetralogy of Fallot undergoing complete surgical repair include those with previous palliative surgery or intervention.

Exclusion criteria:

Cases with no pulmonary valve or other complex congenital heart lesions: atrioventricular canal, Ebstein's anomaly, double outlet right ventricle, transposition of the great arteries, single ventricle,... or cases with insufficient medical records.

Research methods:

Retrospective, descriptive case series study.

Data collection and processing methods:

Data were collected from medical records and processed with SPSS software. Echocardiography results were performed by two experienced echocardiographers.

Research ethics:

Patient information is completely confidential and is only used for diagnosis, treatment and scientific research purposes.

3. Results

During the study period from February 2023 to February 2024, a total of 23 patients met the inclusion criteria, of whom 3 patients (13.0%) had previous palliative intervention.

Table 1. Preoperative clinical characteristics of the study group (n= 23)

Patient characteristics	**Value**
Age (months)	10.5 (5 – 34.5)
0 - < 5 Age (years)	21 (91.3)
0 - < 1	13 (56.5)
1 - < 2	2 (8.7)
2 - < 3	3 (13.0)
3 - < 4	2 (8.7)
4 - < 5	1 (4.3)
5 - < 10 Age (years)	2 (8.7)
5 - < 6	1 (4.3)
6 - < 7	0 (0.0)
7 - < 8	0 (0.0)
8 - < 9	1 (4.3)
9 - < 10	0 (0.0)
10 - <16 Age (years)	0 (0.0)
Female	15 (65.2%)
Weight (kg)	7.8 (6 – 9.5)
SpO_2 (%)	80.0 (70.0 – 88.0)
No cyanosis (spO_2 > 90.0%)	5 (21.7)
Mild cyanosis (spO_2 85.0 – 90.0%)	6 (26.1)
Moderate cyanosis (spO_2 75.0 – 85.0%)	3 (13.0)
Severe cyanosis (spO_2 65.0 – 75.0%)	7 (30.4)
Critical cyanosis (spO_2 < 65.0%)	2(8.7)
Number of cyanosis episodes	
0	14 (60.9)
1	8 (34.8)
2	1 (4.3)

Value: median (interquartile range), number of patients (%)

Table 2. Characteristics of the ventricles at end-diastole (n= 23)

	Value	Value range
Z LVIDd	-2.5 (-2.9 – (-0.2))	-5.1 – 1.9
Small LV	13 (56.5%)	
Normal LV	10 (43.5%)	
Dilated LV	0 (0.0%)	
Z basal RV diameter	1.3 (0.4 – 2.2)	-1.1 – 3.3
Small RV	0 (0.0%)	
Normal RV	16 (69.6%)	
Dilated RV	7 (30.4%)	
Z mid-cavity RV diameter	1.5 (1.3 – 2.5)	-1.1 – 3.5
Small RV	0 (0.0%)	
Normal RV	14 (60.9%)	
Dilated RV	9 (39.1%)	
Z longitudinal RV diameter	1 (0.3 – 2.1)	-1.5 – 3.4
Small RV	0 (0.0%)	
Normal RV	16 (69.6%)	
Dilated RV	7 (30.4%)	
Z right ventricular thickness	6.2 (5.6 – 7)	4.9 – 13.1

Value: median (interquartile range), number of patients (%)

Table 3. Characteristics of ventricular septal defect (n= 23)

Characteristic		**Number of patients**	**Ratio (%)**
VSD type	Perimembranous	23	100.0
	Other	0	0.0
VSD and AoR valve ring size ratio	< 1/3	0	0.0
	1/3 – 2/3	15	65.2
	> 2/3	8	34.8

Table 4. Aortic characteristics (n= 23)

Characteristic	Value	Value range
Z AoR root	6.3 (5.5 – 7.8)	5 – 10.5
Overriding Percentage of AoR (%) < 30 = 30 > 30	30.0 (30.0 – 45.0) 0 (0.0%) 13 (56.5%) 10 (43.5%)	30.0 – 50.0

Value: median (interquartile range), number of patients (%)

Table 5. Characteristics of right ventricular outflow tract stenosis (n= 23)

		Number of patients	Ratio (%)
Subvalvular pulmonary stenosis		6	26.1
Subvalvular and valvular pulmonary stenosis		4	17.4
Subvalvular and supravalvular pulmonary stenosis		1	4.3
Subvalvular, valvular, supravalvular pulmonary stenosis		12	52.2
Pressure Gradient (mmHg)	< 36	0	0.0
	36 – 64	5	21.7
	> 64	18	78.3

Table 6. Right ventricular outflow tract characteristics (n= 23)

	Value	Value range
Z PA valve ring Small PA valve ring Normal PA valve ring Dilated PA valve ring	-2.9 (-4.3 – (-1.8)) 16 (69.6%) 7 (30.4%) 0 (0.0%)	-5.6 – 0.9
Z PA Small PA Normal PA Dilated PA	-2.1 (-4.6 – (-1.1)) 13 (56.5%) 10 (43.5%) 0 (0.0%)	-8.8 – 1.6

Z right PA	-0.9 (-1.7 - 0)	-3.6 – 3.1
Small right PA	5 (21.7%)	
Normal right PA	16 (69.6%)	
Dilated right PA	2 (8.7%)	
Z left PA	0.1 (-1.2 – 1.4)	-2.6 – 3.2
Small left PA	2 (8.7%)	
Normal left PA	18 (78.3%)	
Dilated left PA	3 (13.0%)	
Z Proximal RVOT	0.5 (0 – 1.6)	-0.7 – 3.2
Small proximal RVOT	0 (0.0%)	
Normal proximal RVOT	19 (82.6%)	
Dilated proximal RVOT	4 (17.4%)	
Z Distal RVOT	-3.4 (-3.8 – (-2.6))	-6.4 – (-2.1)
Small distal RVOT	23 (100.0%)	
Normal distal RVOT	0 (0.0%)	
Dilated distal RVOT	0 (0.0%)	
Pressure Gradient of RVOT (mmHg)	85 (70 – 97)	41 – 110

Value: median (interquartile range), number of patients (%)

Table 7. Cardiac indices (n= 23)

	Value	**Value range**
Mc Goon index	1.7 (1.5 – 2)	1.1 – 2.6
> 1.5	16 (69.6%)	
0.5 – 1.5	7 (30.4%)	
< 0.5	0 (0.0%)	
EF (%)	70.0 (66.0 – 74.0)	53.0 – 79.0
Z score TAPSE	0 (-0.8 – 1)	-5 – 2.6
< -2	2 (8.7%)	
> -2	21 (91.3%)	

Value: median (interquartile range), number of patients (%)

Table 8. Associated malformations in tetralogy of Fallot (n= 23)

	Number of patients	Ratio (%)
Patent ductus arteriosus	16	69.6
Patent foramen ovale	15	65.2
Atrial septal defect	9	39.1
Right aortic arch	5	21.7
Persistent left superior vena cava	4	17.4
Coronary artery abnormalities	1	4.3

VSD lesions, AoR overriding on the interventricular septum, RV thickening detected during surgery were similar to the preoperative echocardiography diagnosis. The characteristics of stenosis of RVOT detected during surgery were quite similar to the preoperative echocardiography diagnosis. Most patients had subvalvular, valvular, and supravalvular stenosis (52.1%). Echocardiography detected 15 patients with bicuspid aortic valves, but at surgery, the number of patients with bicuspid aortic valves was 17 ($p > 0.05$). The characteristics of combined malformations in TOF were similar between echocardiography and surgical reports..

4. Discussion

In our study, the median age of patients was recorded as 10.5 months (5 – 34.5). The majority of patients were operated under 12 months, accounting for 56.5%. The special feature of our Center is that we only operate on patients under 16 years old with tetralogy of Fallot. This retrospective study aims to summarize the echocardiographic characteristics of the patient group in a specific period and we do not go into a detailed analysis on a specific age group. Nowadays, most advanced cardiac surgery centers perform complete repair for children under 1 year old. Typically, in the study of author Luong TNH(8), TOF surgery can be successfully performed at a low age of 4.8 ± 1.1 months (3 - 6). The benefit of early surgery is to limit many complications: brain abscess, heart failure, death... and at the same time help children develop physically and mentally better because they do not have to endure prolonged hypoxia. Different studies have recorded different preoperative male/female sex ratios. However, there is no document proving a clear relationship between gender and TOF(9,11). In this study, the median weight was 7.8 kg (6 – 9.5), similar to the studies of Luong TNH(8), Cao DK(9) and lower than the previous study of Nguyen TLA(6) with an average weight of 11.5 ± 8.4 kg. Our SpO_2 results were similar to previous studies. Most children had moderate to severe cyanosis. Based on classification of cyanosis according to SpO2 by author Phan TL(12), the proportion of patients with SpO2 below 85.0% accounted for 52.2%. Cyanosis and syncope represent severe hypoxia when the narrowed right ventricular funnel suddenly contracts, causing a sudden reduction in blood flow to the lungs. 56.5% of patients

had a small left ventricle. The cause of a small left ventricle is due to a narrowing of the right ventricular outflow tract, leading to a decrease in blood flow to the left heart. Kirklin JW(13) noted a high risk of surgery for patients with a small left ventricle, which is even more important for surgical centersn with limited resuscitation and elderly patients. Our group of patients with small left ventricles, most of whom had a left ventricular end-diastolic diameter Z-score from -3 to -2, along with an optimal surgical strategy to widen the right ventricular outflow tract, with preservation of the pulmonary valve ring, had good postoperative results. The right ventricular end-diastolic diameter was larger than normal, indicating a prolonged overload in the right ventricular chamber leading to right ventricular remodeling.

In the study, medium-sized ventricular septal defects accounted for the highest proportion of 65.2%, and large-hole ventricular septal defects were 34.8%. This is consistent with tetralogy of Fallot, which is usually a medium and large ventricular septal defect.

All patients had aortic root dilatation. According to author Hoang QT(2), the degree of cyanosis and aortic dilatation is proportional to the degree of pulmonary artery stenosis.

Our pulmonary valve ring Z score is similar to that of author Nguyen SH(11), the pulmonary valve ring Z score is – 2.5 ± 1.9. Our pulmonary artery Z is much lower than the previous study of Luong TNH(8)-1.46 ± 1.73. Because the narrow pulmonary valve ring does not affect the indication for complete surgery, up to 69.6% of cases have a pulmonary valve ring Z value < -2.

According to Kirklin, one of the main risk factors for early postoperative death is the small size of the valve ring and the pulmonary artery trunk(14). Our right and left pulmonary artery Z is similar to previous studies of Nguyen SH(11), Luong TNH(8). The majority of patients have a normal proximal RVOT, accounting for 82.6%. All patients have a small distal RVOT. This is consistent with the lesions in TOF, mainly distal RVOT stenosis, rarely proximal stenosis, stenosis in the right ventricular funnel and this condition will progress more severely(2). Right ventricular outflow tract stenosis is the most important lesion in TOF, leading to right ventricular hypertrophy, right ventricular dilatation and dysfunction in older children, reducing pulmonary blood flow, reducing arterial oxygen saturation, adversely affecting postoperative outcomes as well as long-term outcomes, so early complete surgical repair in young children aims to limit these consequences(15). In our study, patients with right ventricular outflow tract stenosis at all three locations: below the PA valve, at the PA valve and above the PA valve accounted for the highest rate of 52.2%, all patients had below the pulmonary valve stenosis. The study results of Phan HV(4) also showed similar results: below the valve, at the valve and PA stenosis accounted for the highest rate of 76.0%. The majority of patients had Mc Goon > 1.5, accounting for 69.6% and there were no cases with Mc Goon < 0.5. This result is similar to the study of Phan HV(4), Mc Goon was 1.7 ± 0.2. If the Mc Goon index is < 1.5, the pulmonary artery is small and the prognosis is poor; when this index is < 0.5, there is no indication for complete repair surgery(14). Most of the patients who underwent surgery had normal left and right ventricular function. The results are similar to other studies. The rate of patients with patent ductus arteriosus is

quite high at 69.6%. The accompanying lesions in TOF are similar to those of author Cao DK(9), the most common accompanying lesion is the patent ductus arteriosus with 38.8%.

Through comparison with the surgical report, we found that echocardiography is a very good means to examine the degree of right ventricular outflow tract stenosis in TOF before surgery, helping to indicate the surgical method as well as monitor the results after complete repair surgery. There were two cases (8.7%) where echocardiography misdiagnosed a tricuspid pulmonary valve, but at the time of surgery it was a bicuspid valve.

5. Conclusion

Echocardiography in TOF is non-invasive, safe, has good correlation with cardiac characteristics assessed during surgery, and can be thoroughly applied at all times before, during, and after surgery. Therefore, echocardiography helps cardiologists plan pre- and post-operative evaluation and monitoring. Therefore, we recommend that improved echocardiographic methods are needed to overcome limitations in evaluating pulmonary valve charateristics in TOF.

Abbreviation: AOR= aorta; LV= left ventricle; LVD= left ventricular diameter; LVIDd= left ventricular internal diameter end diastole; PA= pulmonary artery; RV= right ventricle; RVD= right ventricular diameter; RVOT= right ventricular outflow tract; TOF= tetralogy of Fallot; VSD= ventricular septal defect; Z= z score.

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