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Evaluation Of Short and Long-Term Outcomes Of Children With Tetralogy Of Fallot

Fallot Tetralojisi Tanısı İle Takip Edilen Hastaların Kısa ve Orta Dönem Sonuçlarının Değerlendirilmesi

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Abstract

Objective: In children with tetralogy of Fallot (TOF), correction surgery may be performed in the presence of certain findings. We aimed to evaluate the surgical procedures and short- and intermediate-term outcomes of our patients with TOF retrospectively.

Material-Method: This study included patients with the diagnosis of TOF who were followed-up in the pediatric cardiology unit of our hospital between August 2017 and June 2020. Surgical procedures performed and data at pre- and postoperative follow-up were noted from the patients' records.

Results: Among the 27 cases with the diagnosis of TOF, 16 were male and 11 were female. Nineteen patients who fulfilled the required levels of McGoon ratio, Nakata index and pulmonary artery z-score underwent at least one operation. Among them, 13 were operated once, whereas 6 were operated twice. In 14 patients, complete repair was undertaken at the first surgery (median age: 10 months). Postoperative follow-up duration ranged between 1 and 143 months. In 2 patients, pulmonary valve replacement was carried out after complete repair (at the ages of 91 months and 148 months). None of the patients developed severe arrhythmia.

Conclusion: The results of this study suggest that in infants with TOF, complete repair by assessing the McGoon ratio, Nakata index and pulmonary artery z-score may be associated with favourable short- and intermediate-term outcomes.

Keywords: Tetralogy of Fallot, Follow-up, Surgery, Outcome.

Özet

Amaç: Fallot tetralojisi tanısı alan çocuklarda, belirli bulguların varlığında, düzeltme ameliyatları yapılmaktadır. Bu çalışmada Fallot tetralojisi tanılı hastalarımızda uygulanan cerrahi tedaviler ile olguların kısa ve orta dönem sonuçlarını geriye dönük olarak değerlendirmeyi amaçladık.

Materyal-Metot: Çalışmaya Ağustos 2017-Haziran 2020 tarihleri arasında hastanemizin pediatrik kardiyoloji bölümünce izlenen Fallot tetralojisi tanılı hastalar dahil edildi. Olgulara uygulanan cerrahi işlemler ile ameliyat öncesi ve sonrası izlemlerdeki veriler, hasta dosyaları incelenerek kaydedildi

Bulgular: Çalışmaya alınan 27 hastadan 16'sı erkek, 11'i kız idi. McGoon oranı, Nakata indeksi ve pulmoner arter z-skoru, ameliyat edilme şartlarını karşılayan 19 hastaya cerrahi uygulanmıştı. Tek operasyon geçiren hasta sayısı 13 iken, iki operasyon geçiren hasta sayısı 6 idi. İlk operasyonda 14 hastaya (ortanca yaş: 10 ay) tam düzeltme operasyonu uygulanmıştı. İlk ameliyattan sonra izlem süreleri 1-143 ay arasında değişmekteydi. Tam düzeltme ameliyatından sonra 2 hastaya (91 aylık ve 148 aylık iken) ağır pulmoner yetmezlik gelişmesi ve sağ ventrikül yetmezliğiyle sonuçlanması sebebiyle pulmoner kapak replasmanı yapılmıştı. Hastaların hiçbirinde ciddi aritmi gelişmediği kaydedildi.

Sonuç: Çalışmamızın bulguları, bir yaş altı Fallot tetralojisi tanılı çocuklarda, McGoon oranı, Nakata indeksi ve pulmoner arter z-skoru değerlendirilerek tam düzeltme ameliyatı yapılmasının olumlu sonuçlar ile ilişkili olduğunu düşündürmektedir. Daha geniş serili çalışmalarla Fallot tetralojisinde ameliyat sonuçları daha da yüz güldürücü olacaktır.

Anahtar kelimeler: Fallot Tetralojisi, İzlem, Cerrahi, Sonuç.

Introduction

Tetralogy of Fallot (TOF) is the most common congenital cyanotic heart disease. It constitutes 5-7% of all congenital cardiac disorders, and about 10% of congenital cyanotic heart diseases (1).

Some formulas have been developed to help in reducing the preoperative mortality of children with TOF. Previously, only the proportion of end-diastolic diameter of left ventricle to aortic diameter was used. However, in recent years, the McGoon ratio, pulmonary valve z-score and Nakata index have also been included in the assessment of the risk. A palliative surgery will be carried out in cases with a McGoon ratio lower than 1.7 and a Nakata index lower than 200 mm2/m2 (2). The McGoon ratio, pulmonary valve z-score and Nakata index are

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important in planning complete repair (2-8). In this study, we aimed to evaluate the short- and intermediate-term outcomes of our patients with the diagnosis of TOF.

Material and Methods

Our study was approved by the Clinical Research Ethics Committee of Suleyman Demirel University (23.07.2019/ Approval no:177). This study included 27 patients with the diagnosis of TOF who admitted to our pediatric cardiology unit between August 2017 and June 2020. Surgical procedures were carried out in other clinics and operated cases were follow up pre – and postoperatively in the pediatric cardiology department of our hospital. Details of the operative procedures and the age of the cases at the time of the operation were collected from the hospital records. Pre- and postoperative follow-up durations were noted. Findings of echocardiographic examinations were recorded, and the McGoon ratio, Nakata index and pulmonary valve z-score were calculated in all the cases.

Echocardiographic Examination

Transthoracic two-dimensional and Doppler echocardiographic examinations were performed by using the 5.0 and 8.0 MHz transducers of the Philips Affiniti 70C Ultrasound Machine (Philips Healthcare, Andover, USA). M-mode echocardiographic measurements were carried out from the level of the posterior mitral valve according to the guidelines of the American Society of Echocardiography (9).

Angiography

From the angiography records, the McGoon ratio, Nakata index and pulmonary valve z-score were calculated. Following formulas were used for the calculation of each variable.

McGoon ratio: the sum of the diameter of the pulmonary arteries divided by the diameter of the descending aorta.

Nakata index: the sum of the cross-sectional area of the pulmonary arteries divided by the body surface area.

Pulmonary valve z-score: the difference between the measured pulmonary artery diameter and the mean value of the normal pulmonary artery diameter divided by the standard deviation of the mean value of the normal pulmonary artery diameter.

Statistical Analysis

Data were analysed using the SPSS for Windows 22 package program (SPSS; Chicago, Illinois, USA).

Results

Demographic data and operative characteristics of the cases are presented in Table 1. Among the 27 patients included into the study, 16 (59.3%) were male, whereas 11 (40.7%) were female. Left and right arcus aorta were present in 22 (81.4%) and 5 (18.6%) cases, respectively. Age at the time of diagnosis ranged between 0 and 197 months 13 (48%) patients underwent one operation, whereas 6 (22 %) patients underwent two operations. Age at the time of the first operation varied from 0 to 122 months, whereas age at the time of the second operation ranged between 17 and 148 months.

Table 1. Demographic data and operative characteristics of the cases

	N (%)	Median	Mean±SD	Minimum	Maximum
Female	11 (40.7)				
Male	16 (59.3)				
Body weight (kg)	27	15		5	50
Length (cm)	27	110		56	165
Age (months)	27	63		2	201
Age at diagnosis (months)	27	0		0	197
SpO_2 at first operation (%)	19	76		68	81
Age at first operation (months)	19	10		0	122
Age at second operation (months)	6	34		17	148
Body weight at first operation (kg)	19	7.75		3	43
Body weight at second operation (kg)	6	10		8.7	40
McGoon ratio at first operation	19	2	1.93±0.29	1.3	2.40
Nakata index at first operation (mm^2/m^2)	19	224	220.62±26.74	170	270
Pulmonary valve z-score at first operation	19	-3.1	-3.29±0.686	-4.6	-2.3
McGoon ratio at second operation	4	2	2.00±0.081	1.9	2.1
Nakata index at second operation (mm^2/m^2)	4	235	237.75±27.32	210	270
Pulmonary valve z-score at second operation	4	-3	-3.02±0.28	-2.7	-3.4
Not operated	8				
Left arcus aorta	22(%81.4)				
Right arcus aorta	5 (%18.6)				
Preoperative pulmonary gradient (mmHg)	19	80		60	110
Postoperative follow-up duration (months)	19	25		1	143

Table 2. Accompanying cardiac and non-cardiac anomalies of the cases

	Non-cardiac anomalies					Cardiac anomalies					
	Down syndrome	DiGeorge syndrome	Noonan syndrome	Anal atresia	Congenital hypothyroidism	Polydactyly	PFO	PDA	AI	Secundum ASD	Muscular VSD
N	2	1	1	1	1	1	5	2	2	4	1

Table 3. Types of the operations and age of the cases at the time of surgery

	N	Age (months) (median)	Age (months) (min)	Age (months) (max)
Complete repair at first operation	14	14	8	122
Central shunt at first operation	2		8 days	1
Right Blalock-Taussig shunt at first operation	2		15 days	1
Balloon valvuloplasty and RVOT stenting at first operation	1		1	1
Complete repair at second operation	4	21	17	48
Pulmonary valve replacement at second operation	2		91	148

Table 4. Postoperative echocardiographic findings of the cases who underwent complete repair (n=19)

	Tricuspid insufficiency	Pulmonary insufficiency	Residual VSD	Residual pulmonary stenosis
None	5	4	16	1
First degree or mild*	9	8	3**	12
Second degree or moderate*	4	3	0	5
Third degree or severe*	1	4	0	1

*Valvular insufficiency is classified as first, second or third degree, whereas valvular stenosis is classified as mild, moderate or severe. **Three patients had a small (<3 mm) VSD.

Accompanying cardiac or non-cardiac anomalies of the cases are presented in Table 2. Two patients had the diagnosis of Down syndrome. The most common accompanying cardiac anomaly was patent foramen ovale.

Table 3 summarizes types of the operations and the age of the cases at the time of surgeries. A total of 14 patients who fulfilled the suggested McGoon ratios, Nakata indices and pulmonary valve z-scores underwent complete repair at the first operation. In 4 patients, palliative surgeries including Blalock-Taussig shunt, central shunt or right ventricular outflow tract stenting were carried out due to the presence of severe pulmonary hypoplasia firstly, and complete repair was performed thereafter in their follow-up. In two patients with severe pulmonary insufficiency, pulmonary valve replacement with injectable self-expanding valves was performed. In another patient with severe pulmonary insufficiency, pulmonary valve replacement is planned currently.

Table 4 shows the echocardiographic findings of 19 cases after complete repair.

Discussion

In patients with TOF, the most important factor affecting the morbidity and mortality of the surgery is the degree of pulmonary hypoplasia. Pulmonary artery hypoplasia should be assessed using the McGoon ratio, Nakata index and pulmonary valve z-score. Although one operation for complete repair is the desired approach in TOF, this may sometimes be not possible due to the presence of severe pulmonary hypoplasia. In such cases, two-stage operations may be necessary.

In the presence of severe pulmonary hypoplasia, complete repair by closing the ventricular septal defect (VSD) may result in a significant increase of right ventricular pressure, a decrease in cardiac output and the development of right ventricular failure (5, 10-12). Among our patients, 4 underwent a palliative procedure (Blalock-Taussig shunt, central shunt or right ventricular outflow tract stenting). In these patients, complete repair was carried out when they fulfilled the required levels of McGoon ratio, Nakata index abd pulmonary valve z-score. They are still being followed-up in good clinical status.

In recent years, it has been demonstrated that operating patients with TOF at an early age before the development of right ventricular failure has a favourable impact of morbidity and mortality (13, 14). A complete repair performed at an early age diminishes right ventricular hypertrophy and slows down the development of fibrosis in the right ventricle (15). A lower mass of muscle will be excised in these cases and a smaller graft will be placed into the outflow of the right ventricle which enables the obtainment of better right ventricular functions and prevents development of aneurysms. This decreases the probability of arrhythmia development (15). In our patients, complete repair was done at an early age, and none developed severe arrhythmia.

Mild pulmonary valve insufficiency which may occur following the complete repair of TOF is usually well tolerated (16). Among our 14 patients who underwent complete repair at their first operation, 10 developed mild pulmonary insufficiency, whereas 4 developed 3rd degree pulmonary insufficiency at their follow-up. In 2 of the patients with 3rd degree pulmonary insufficiency, pulmonary valve replacement was performed later (at the ages of 91 months and 148 months) due to the enlarged right ventricles. The other 2 patients with significant pulmonary insufficiency are still being followed-up.

Şaşmazel et al. have reported a McGoon ratio over 2.0 which indicates an adequate pulmonary arterial maturation to be

the most significant factor for deciding on a complete repair in patients with TOF (17). The authors also observed that a shunt operation or a palliative procedure was more suited in infants younger than 6 months (17). In our study, one-stage complete repair was carried out in cases with a McGoon ratio over 2.0, a Nakata index over 200 mm2/m2 and a pulmonary artery z-score over -3, the median age at the time of surgery being 10 months. Patients with a McGoon ratio lower than 1.7 underwent two-stage operations (a shunt operation or a similar surgery, and complete repair at an older age).

As right ventriculotomy was applied in our patients for the purpose of complete repair, the percentages of residual pulmonary stenosis and residual VSD were found quite low. It has been reported that the transatrial closure of VSD or making a small ventricular incision reduces the risks of sudden death and arrhythmia by preventing the development of right ventricular dilatation postoperatively (18). However, right ventriculotomy is preferred more commonly as it makes both the closure of the VSD and the resection procedure easier by enabling an accurate assessment of the infundibular muscle bundles (19).

Right arcus aorta is seen more frequently in cases with TOF than in the general population (20, 21). We found quite a high percentage (18.6%) of our cases to have arcus aorta which is consistent with the literature.

Conclusion

In conclusion, deciding on a complete repair by taking the McGoon ratio, Nakata index and pulmonary artery z-score into account seems to be associated with favourable short- and intermediate-term outcomes in infants with TOF who are within their first year of life. The results of our study are in accordance with those of recent studies in the literature. However, the low number of the cases included and the retrospective nature of the research are the main limitations of this study. Large-scale studies are needed to make more precise and comprehensive conclusions on the outcomes of children affected by TOF.

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