

TETRALOGY OF FALLOT: REPORT OF TWO CASES IN OUTPATIENT PRACTICE

Gordana Đorđević*, Gordana Grujić-Ilić

Department of Pediatrics, Health Center Niš, Niš, Serbia

Abstract. *Tetralogy of Fallot or Tetras Fallot is a congenital heart disease with cyanosis, which involves four associated anomalies of the heart. As physicians in primary health care of children, we treat and monitor the growth and development of children at risk with special attention. Among them are also the children with tetralogy of Fallot. The aim of this report was to present and compare postnatal status of two children with tetralogy of Fallot. We presented the girl and a boy, approximately the same age, operated at the same surgical clinic. Both patients were without hereditary load. The girl had a milder clinical course without complications. The boy had associated anomalies, cerebrovascular insult, and a reoperation. After complete surgical correction of congenital heart disease, both children are progressing and developing within the normal range for age. ECG of both children showed the signs of right bundle branch block. Our mission is to continue to closely monitor these children and react in case of complications or worsening of recent findings.*

Key words: *Tetralogy of Fallot, postnatal course, pediatric patient, outpatient care*

Introduction

Tetralogy of Fallot (TOF) or Tetras Fallot is cyanogen, congenital heart disease with the right-to-left shunt. TOF includes combination of four associated heart anomalies: right ventricular outflow tract obstruction (stenosis of pulmonary trunk), ventricular septal defect (VSD), right ventricular hypertrophy and “overriding” aorta [1–7].

Embryologically, these anomalies occur as a result of disorders in morphogenesis of the right ventricle infundibulum [1]. In this congenital heart disease, infundibular septum is moved anterosuperior [1, 8], resulting in a large defect of the membranous part of the septum, immediately below the aortic valve, and movement of the aortic confluence to the right, and consequently, narrowing of the pulmonary tree. Aorta is dilated [4, 9, 10] and communicates with both ventricles (“overriding” aorta). Aortic arch was oriented to the right in 15–25% of patients [2, 4, 7, 11], without hemodynamic significance [2].

Ventricular septal development is a complex process that includes different septal structures of various origins and different positions. Ventricular septum is formed from the 5th to the end of the 7th week of embryonic development, by merging muscular septum with ventricular outflow tract and aortopulmonary septum [12].

The incidence of TOF is 0.4/1000 of live births [1, 13]. Compared to all congenital heart diseases, presented

by various authors, the incidence of TOF ranged from 3–5% [3, 4, 8, 14], to 7–10% [1, 2, 9]. TOF representation in boys and girls was almost in the same range [3, 16], with a slightly higher percentage among boys (1.56:1) [15].

Etiology of TOF is multifactorial, and it is considered that the genetic factors, in combination with environmental conditions are responsible for developing this heart anomaly [2, 17, 18]. Recent studies have indicated that the TOF is often associated with deletion on chromosome 22—22q11 deletion, as in DiGeorge [2, 3, 7, 19] and Shprintzen (velo-cardial-facial) syndromes [3, 7, 19]. It was also indicated that there is a relationship between the congenital heart disease and the level of the vascular endothelial growth factor, considering that its concentration was elevated in children with cyanotic heart disease [20].

Diagnosis is based on anamnestic data, and clinical examination (central type of cyanosis, heart murmurs, fatigue, dyspnea and occurrence of paroxysmal cyanotic crises, development of clubbing fingers, slow growth, chest deformity), chest rentgenography, electrocardiogram (ECG), echocardiography, laboratory analysis, cardiac catheterization (in candidates for surgery), computer tomography of the heart, aortography and coronary arteriography [1–4, 7], and recently, magnetic resonance imaging [4].

Anomalies associated with TOF have been described and these include the ASD [1, 2, 7, 10, 11, 21], or multiple VSD [7], or congenital absence of pulmonary valve [7, 16, 21]. In the older literature [1, 10], TOF associated with ASD has been labeled as Pentalogy of Fallot.

The aim of this study was to present subjective and objective status of two children with TOF operated at the same surgical clinic and to compare their preoperative and

Correspondence to: Gordana Đorđević, MD, specialist of pediatrics Health Center Niš, 15 Vojvode Tankosića St, 18000 Niš, Serbia
Phone: +381 18 503503 Fax +381 18 237990
E-mail: dr.goca@yahoo.com

postoperative course with the cases of (inter)national practice.

Material and Methods

Presented data were based on anamnesis, physical examination, ultrasound findings, data from medical records and discharge papers of the two children of male and female gender and similar age.

Ethical code has been respected; the author obtained the written consent from the parents (mothers) of both children for scientific presentation of children's medical records.

Case Reports

Case 1 (8.5 years old girl). It is the second child from the (second) normal pregnancy. Both parents and older sister are in good health, and without congenital heart disease. The child was born at term in a natural way. At birth she was blue, started crying later, body weight (BW) was 3800 g and the Apgar score 9. TOF was diagnosed at birth.

In the first year of life, the girl did not develop properly. Mild cyanosis was noticed in the third month of life, auscultation revealed systolic murmur intensity IV/6. The mother noticed that the girl was tired while feeding. Laboratory findings of erythrocytes, hematocrit and hemoglobin were within the normal range. The girl had upper respiratory infections several times and one hospitalization due to lower respiratory infection. The girl was not indicated for cardiac medicamentous therapy until the term for surgery.

Complete surgical reparation was performed at the age of one year. Diagnostic methods — echocardiography and heart catheterization, performed preoperatively, indicated the following abnormalities: non-restrictive perimembranous defect was positioned on the interventricular septum, over which the overriding aorta was about 50%, the right ventricle was hypertrophic, and right ventricular outflow tract was narrowed in the infundibular level with thick muscular wall. Aortic arch was described as “left”, without anomaly.

A large perimembranous VSD was determined intraoperatively; it was 12–14 mm in diameter, with overriding aorta over 50%, fibro-muscular obstruction of the infundibulum, and severe hypertrophy of the right ventricle.

The postoperative period was normal. The girl progressed well in weight and height; echocardiography showed normal findings, regurgitation was registered at the root of the pulmonary artery, but it was hemodynamically insignificant. Auscultation sounds were clear, with normal intensity, without murmurs, and the ECG findings pointed to a right bundle branch block with sinus rhythm.

The author of this work regularly monitored the girl since 2011. During this time the girl had six upper respiratory infections, inflammation of the middle ear three times, and flu (influenza) but without complications.

During follow-up by the author, she did not have inflammation of lower respiratory tract; according to data from medical record, one year after surgery, she had bronchitis, which was complicated with pneumonia. Systematic examination for school enrollment indicated that her body weight corresponded to the average values for age, while her body height was below average, but still within normal values. Blood elements were within the normal range, as well as blood pressure.

Her recent (July 2014) ECG is presented here (Fig. 1).

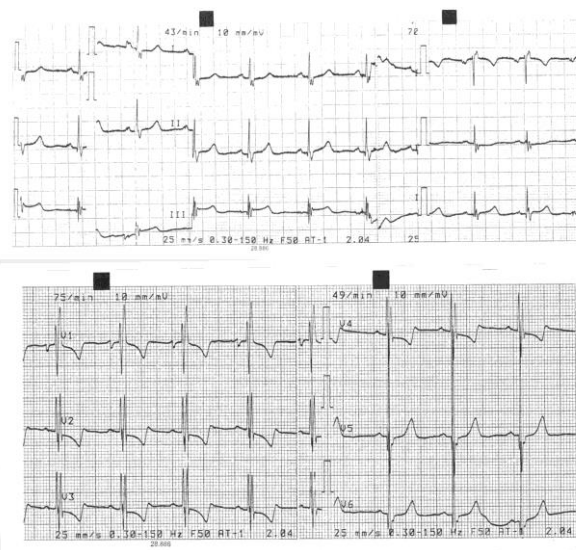


Fig. 1. Sections of ECG of the girl. The girl's ECG: normal axis; signs of the incomplete right bundle branch block in leads V_1 – V_4 (presence of 2 R waves - RSR^1 ; QRS 0.08 s); negative T waves in leads V_1 – V_4 .

Case 2 (8 years old boy). He is the firstborn, bigger child (in body weight) from a twin pregnancy. Birth was premature — in the 36th week, however the Apgar score was 9. Diagnosis — Pentalogy of Fallot associated with bilateral inguinal hernia was determined at birth.

In the first month of life there were mild cyanosis, signs of anemia and systolic-diastolic murmur intensity IV/6. Therapy included iron containing medication. One month later, blood elements were within the normal range, but peripheral cyanosis and the same heart murmur were still present. In the third month, during the regular check-up, the mother stated that the child had severe cyanosis after defecation and during intensive crying. In the fourth month of life the crisis of cyanosis and vomiting occurred, and that was the reason why the child was admitted to hospital; the drug Propranolol has been introduced and the child was prepared for surgery. Preoperative examination confirmed larger right side of the heart than the left, large perimembranous outlet VSD and overriding aorta with left-oriented arch, infundibular and valvular stenosis of the pulmonary artery, and large ASD secundum.

During the cardiac catheterization the boy fell into hypotension, hypoxemia and consequently developed

cerebrovascular insult (CVI). Several days later, a neurological deficit has developed, presented with paresis of the left half of the body; which was partially withdrawn for two weeks. In the process of another preparation for surgery, the drug Propranolol was discontinued. However, two days later, at first mild, and then severe crises of cyanosis and loss of consciousness appeared. Propranolol was reintroduced, while the Medical Consilium decided to immediately perform a complete surgical correction.

Intraoperative diagnosis determined a large infracaval ASD (missed the entire bottom edge of the interatrial septum), a large perimembranous VSD with about 12 mm in diameter, overriding aorta (> 60%), while the infundibulum of the right ventricle was very narrow, almost like atresia, however, the process of fibrosis was not being expressed.

The postoperative course was normal until the 6th day, when the crises of cyanosis occurred again, accompanied by extreme hypertension (more than 200 mm Hg). Echocardiography pointed to residual VSD and significant tricuspid insufficiency. It was followed by reintervention and after that, the boy's condition stabilized.

In the further course, the boy was followed by a cardiologist and a neurologist. Auscultation revealed systolic murmur intensity III/6 along the left edge of the sternum. Neurological examination registered the phenomenon of "sunset", the occasional eye deviation to the left, hypotonia of the axis of the body and upper extremities and that boy could not sit spontaneously and his reflexes were enhanced. Over time, the boy progressed in body weight and height and gradually recovered neurologically.

The boy had the first infection of the airways (pneumonia) at the age of eight months. By the age of 8 years he had bronchitis nine times and pneumonia three times.

Inguinal hernias, diagnosed at birth, were operated at the age of 2.5 years.

At the systematic examinations, starting from the third month of life until the age of seven, the boy was shorter in height, with lower body weight compared to his twin brother. There was considerable discrepancy in the growth and development from the age of three months to one year, but even after that period, the boy was in a certain disadvantage compared to his twin brother, who had a lower birth weight.

The author of this study followed and treated the boy since 2009. During this time the boy had six lower respiratory tract infections, without pneumonia. The systematic examination upon school enrollment showed that the boy's weight and height corresponded to the average values for age, auscultation of the heart indicated a systolic murmur intensity IV/6, blood elements were in the normal range, as well as the blood pressure.

His recent (September 2014) ECG is presented here (Fig. 2).

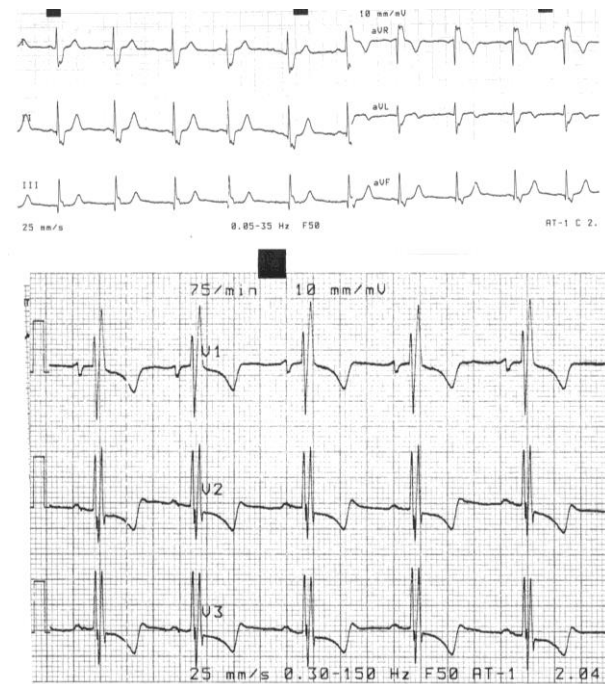


Fig. 2. Sections of ECG of the boy. The boy's ECG: normal axis, signs of the complete right bundle branch block in leads V_1 - V_3 (presence of 2 R waves - RSR' ; QRS 0.132 s); there are no other rhythm disorders.

Discussion

We presented two children of similar age with TOF — one girl (from the second pregnancy) and one boy (from twin pregnancy). Both children were without hereditary load. However, the boy, according to the literature, had the risk of "twin process" [19]. The literature describes cases of twin pregnancies in which only one of the twins had a TOF [2, 10, 14, 19]. In addition to possible genetic causes, this is also explained by the previously mentioned "twin process", where blood flow is greater in one twin, while the other has less blood flow, which can cause the occurrence of congenital heart defects. Such a twin is smaller [5, 19], with lower body weight (BW) and body height, which was not the case with our little boy, who was the heavier twin at birth.

Our boy also had associated anomalies — ASD and bilateral inguinal hernia. According to the literature, TOF associated with ASD was found in 2% of those patients. Only in the older literature [1, 10], TOF associated with ASD has been labeled as Pentalogy of Fallot.

In these patients, symptoms and treatments are the same as those with TOF, but surgical correction and postoperative course are more complex [2], which was also the case with our patient. Inguinal hernia, as an extracardiac anomaly, associated with TOF is also described in the literature [5, 14].

In the first three months of life gradual appearance of cyanosis in both children was observed, which was objectively confirmed in other children as well [2, 7, 8].

Also, the crises of cyanosis, which typically begin to appear a few months after the birth during the child's agitation (crying) and decreased hydration [2], were observed in our boy in the fourth month of life. Temporary introduction of Propranolol in the treatment of cyanotic crises, according to the literature, is justified [2, 3, 7].

For the girl who had no associated anomalies and in whom the symptoms were mild, surgical correction of TOF was planned and performed at the age of one year. With the boy, surgical correction was performed at the age of 6 months, because the attacks of cyanotic crises became stronger. Age at which both corrections were done and the fact that complete surgical corrections of heart defects were performed, without prior palliative interventions, is in line with the attitude and experience of authors of recent (inter) national literature [2–4, 8, 11, 13, 22, 23]. In preparation for surgery, the boy had CVI, which briefly delayed the operation. In the available literature, there is evidence that 1.5% of patients had CVI before surgery TOF [15].

The postoperative course for the girl was without complications, while the boy had a complicated postoperative course because of reoperation and the previous stroke. After surgery, both children progressed

well, to some degree compensating the backlog of BW and height and continue to have development within the normal range for their age. Such favorable postoperative course and favorable prognosis for life are indicated by many authors [2, 11, 13, 16, 24, 26].

The right bundle branch block, presented in ECG, is a common finding in patients after TOF surgery [11]. According to available data, incomplete right bundle branch block after surgery was present in about 20.2% of patients [25], while complete right bundle branch block, according to various authors was present in 65% [25] or 88% [15], or even 94% [26].

Conclusion

We presented the postnatal course of TOF in 8-years-old girl and boy. After a complete surgical correction, both children developed within the normal range for age.

Our mission in outpatient practice is to continue to closely monitor these children, to control regularly their health status, ECG and blood pressure and react promptly in case of complications or worsening of recent findings.

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