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Arrhythmias and conduction abnormalities in children after repair of tetralogy of Fallot

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Aim. *To find out types and frequency of cardiac arrhythmias and conduction abnormalities in the group of children who underwent surgery for tetralogy of Fallot (TOF).* ***Methods****. Forty-six pedicatric patients who underwent a complete repair of TOF at the age of 1 to 13 (mean 2.89 ± 2.36) were studied. Thirty-eight (82.60%) had total correction and 8 (17.40%) had palliative operation first, and total correction afterwards. Twenty-four-hour Holter ECG monitoring was performed in all 46 pediatric patients aged from 1 to 16 yrs (mean 6.48 ± 4.04) after surgery as follows: in 1 patient (2.17%) after a year, in 20 patients (43.47%) after 2 to 5 years and in 25 patients (54.34%) after 5 years. Mean age of patients on Holter monitoring was 9.25 ± 4.39 (range 4–19). Twenty of them (43.47%) were girls and 28 (56.53%) were boys. All the patients were evaluated by standard methods (clinical signs, clinical findings, ECG before surgery, ECG before Holter monitoring and 2D Doppler echocardiography.* ***Results.*** *Types of heart rhythm found out by Holter monitoring were: sinus nodus dysfunction in 1 child (2.17%), significant premature atrial contraction (PAC) in 8 (17.39%), supraventricular paroxysmal tachycardia in 3 (6.53%), transient nodal rhythm in 2 (4.34%), premature ventricular contraction (PVC) Lown grade I-III in 9 (19.56%) and Lown grade IV in 2 (4.34), atrioventricular (AV) block grade I in 2, right bundle branch block (RBBB) in all 46 (100%) and RBBB + left anterior hemiblock (LAH) in 4 (8.96%). There was no presence of atrial flutter, ventricular tachycardia or complete AV block. None of them experienced sudden death. Using cross procedure statistical methods, it was found that all the patients with PVC had right ventricular dilatation. There was no relation of other types of arrhythmia found on Holter monitoring to the other parameters from echocardiography, neither to the other standard methods. Children did not need the pace-maker, but 36.95% of the them required antiarrhythmic drugs.* ***Conclusion.*** *Twenty-four hour Holter ECG is a noninvasive and very sensitive method for discovering heart rhythm disturbances in children after the repair of tetralogy of Fallot, especially in asymptomatic patients. The patients after the repair of this congenital heart disease needed a long-term follow-up for early recognition of serious heart rhythm disturbances and their treatment.*

K e y w o r d s : **tetralogy of Fallot; cardiac surgical procedures; arrhythmia; heart conduction system; electrocardiography, ambulatory.**

Introduction

All the patients who undergo cardiac surgery due to congenital heart defects are at risk to develop postoperative arrhythmias (1). Those may appear in the early postoperative period, but their appearance significantly increases late after surgery. Many of these arrhythmias are sudden, unexpected, and life threatening (2, 3).

The repair of specific cardiac defects predisposes pediatric patients to develop specific arrhythmias. The highest incidence of these arrhythmias appears after intraatrial repair of d-transposition of great arteries, Fontan operation of single ventricle and repair of tetralogy of Fallot (TOF). Factors associated to postoperative arrhythmias include: natural history of lesion, preexisting arrhythmias or hemodynamic abnormalities, specificities of surgical procedure, and the presence of residual hemodynamic abnormalities. Factors that affect myocardial preservation during surgery, such as cardio-pulmonary bypass time, hypothermic arrest and techniques of cardioplegia could

also be expected to influence postoperative events. Early repair may prevent the development of ventricular fibrosis or atrial dilatation – a provider substrate for life threatening arrhythmias (4).

The discovery of arrhythmias, their follow-up and the need for treatment with medicaments, heart stimulation-pacemaker or defibrillator, or directing to radiofrequent ablation and the permanent follow-up of effects of applied therapy is the main purpose of monitoring of heart rhythm disturbance in pediatric patients operated on for congenital heart disease using 24-hour Holter electrocardiography monitoring (5–7).

Tetralogy of Fallot is a congenital heart defect, which includes: large ventricular septal defect, dextroposition of overriding aorta, hypertrophy of the right ventricle and the stenosis of pulmonary artery. Repair of TOF includes patch closure of ventricular septal defect with infundibular subpulmonic resection, with or without trannsanular patch across pulmonic valve (procedure uses a cardio-pulmonary-bypass). The specificity of this surgical procedure and timing influenced the outcome in those children. Technical specificities of the procedure may damage specific areas of the conduction system and affect developing heart rhythm disturbances (8, 9).

The aim of this study was to find out types and frequency of cardiac arrhythmias and conduction abnormalities in a group of children in whom tetralogy of Fallot was repaired.

Methods

Forty-six children who underwent repair of TOF were studied. This study included children who had total correction of this congenital heart disease, and also children who had palliative operation first, and then total correction.

The group of children with TOF was a subgroup of children operated on for different congenital heart diseases. In the period of 10 years (1991–2000) in our country, the number of children operated on for TOF (all in foreign cardiosurgical centers) was 56. We included 46 patients who had regular periodical check-up. The other 10 were not followed up (probably moved to another institution, died or did not come for the check-up).

The main method used in the study was 24-hour Holter ECG monitoring (type two-channel recorder) which was designed for out-patient examination. ECG activity of patients was being recorded for 24 hours, and analyzed later by Space Labs Medical Holter analysis system.

We performed Holter ECG monitoring at least a year after the surgery (in most of our pts it was performed many years after the surgery) because we wanted to defect late arrhythmias and conduction abnormalities.

The following Holter parameters were observed: minimal, maximal and mean heart rate; presence of various types of arrhythmias and conduction abnormalities such as: sinus node dysfunction (SND), premature atrial contraction (PAC), atrial flutter, supraventricular paroxysmal tachycardia (SVT), nodal rhythm, premature ventricular contraction (PVC), ventricular tachycardia (VT), various degrees of atrioventricular (AV) blocks, right bundle branch block (RBBB) and bifascicular block (RBBB + LAH).

In all the patients standard methods were used, too: 12-channel ECG before surgery with the presence or absence of sinus bradycardia data, PAC, PVC, AV blocks, RBBB, LAH; clinical signs (tachycardia, arrhythmia, chest pain, dizziness, collapse) and clinical findings (tachycardia, bradycardia, arrhythmia, exstrasistolies) in heart auscultation (immediately before Holter application); 12-channels ECG immediately before Holter ECG; 2D Doppler echocardiography with the presence or absence of 5 parameters: right ventricular dilatation, tricuspid regurgitation, residual left-right (L-D) shunt across ventricular septal defect (VSD), pulmonary incompetence and right ventricular outflow obstruction.

All the data were statistically evaluated using SPSS 10.0 (Statistical program for Windows) which included descriptive statistics: a) Frequency tables b) Descriptive procedures c) Cross tabs procedures cross tabulation, Chi-squared and Fisher's exact test.

a) Frequency tables were used to display data on the presence or absence of variables (excluding numeric variables), and their percentage.

b) Descriptive procedures were used to calculate the numeric variables (age at surgery, duration of Holter ECG monitoring, time interval between surgery and Holter monitoring) and to calculate the data for heart rate (minimum, maximum and mean heart beat /min).

c) Cross-tabulations were used for crossing the parameters found by Holter ECG with parameters received by standard methods (data from clinical signs and findings, data from ECG before operation and before Holter monitoring and echocardiography data).

- Chi-squared test was used for showing the probability of association of two variables when null hypothesis can be assumed or rejected
- Fisher's exact test was applied for the comparison of the frequency of observation when numbers in fourply table were too small for the Chi-squared test.

Results

Forty-six children who underwent repair at the age of 1 to13 (mean 2.89 ± 2.36) were studied. Thirty-eight patients (82.60%) had total correction and 8 pts (17.40%) had palliative operation first, and total correction afterwards.

Twenty-four hour Holter ECG monitoring was performed in all 46 pts from 1–16 (mean 6.48 ± 4.04) after surgery. After 1 year Holter was performed in 1 patient (2,17%), between 2–5 years in 20 pts (43.47%) and after 5 years in 25 pts (54.34%).

Mean age of patients on Holter monitoring was 9.25 ± 4.39 (range 4–19). Twenty of them (43.47%) were girls and 28 (56.53%) were boys.

Three pts (6.52%) had symptoms of heart beating before Holter monitoring, 1 (2.17%) collapsed, and one had dizziness. During clinical examination (heart auscultation): 2 (4.34%) pts had tachycardia, 1 (2.17%) bradycardia, and one exstrasystolies.

One patient had PAC on 12-channel ECG before surgery. Before Holter monitoring all patients were examined by 12-channel ECG and the data showed: sinus bradycardia in 1 (2.17%) pt, AV block gr I in 2 (4.34), and RBBB was found in all our pts [(43 (93.47%) pts had complete, and 3 (6.53%) had incomplete RBBB)].

Five parameters were evaluated on 2D Doppler echocardiography: right ventricular dilatation was found in 31 (67.39%) children; tricuspid regurgitation was found in 6 (13.04%); L-R shunt across the VSD in 4 (8.7%) children; significant right ventricular outflow obstruction (Pg > 60 mm Hg) in 1 child (2.17%), and mild obstruction in 39 (84.78%); significant pulmonary incompetence (regurgitation) was found in 5 pts (10.86%), and other 38 (82.6%) had mild pulmonary regurgitation.

Heart rhythm disturbances in children after the repair of TOF found out by 24 h Holter ECG monitoring are shown in Table 1.

The analysis of heart rate by 24-hour Holter monitoring showed:

- minimal heart rate: mean 55.76 ± 7.64 (range 38–71) beat per minute.
- maximal heart rate: mean 156.06 ± 20.01 (121–201)
- mean heart rate: mean 85.97 ± 10.26 (65–107)

Each type of heart rhythm disturbance found out by 24 h Holter monitoring was crossed with the following parameters: ECG before surgery, clinical signs, clinical findings, ECG before Holter, and 2D Doppler echocardiography.

Cross-tabulations showed that all our pts with PVC, namely 9 with PVC Lown gr I-III and 2 with PVC gr IV had right ventricular dilatation on echocardiography. Other cross-tabulations, Chi-squared test and Fisher's exact test for other variables showed that there wasn't a significant relation between the observed variables.

Discussion

It is known that postoperative arrhythmias and conduction abnormalities after TOF appear from the earliest total repair (10).

Comparing the results of arrhythmia type found on Holter ECG in our study (Table 1) to data from literature (Wetter VL) showed in Table 2, it was concluded that the

Table 1

Heart rhythm disturbances after repair of tetralogy Fallot found out by Holter ECG

Type of heart rhythm disturbance	No of pts	%
Sinus nodus dysfunction	1	2.17
Premature atrial contraction – non-significant	22	47.42
Premature atrial contraction significant	8	17.39
Supraventricular paroxysmal tachycardia	3	6.53
Nodal rhythm - transient	2	4.34
Atrial flutter	0	0
Premature ventricular contraction Lown gr. I-III	9	19.56
Premature ventricular contraction Lown gr. IV	2	4.34
Atrioventricular block gr. I	2	4.34
Atrioventricular block gr. III	0	0
Right bundle branch block - complete	43	93.47
Right bundle branch block - incomplete	3	6.53
Right bundle branch block + left anterior hemiblock	4	8.69
Sudden death	0	0

Table 2

Tetralogy of Fallot and postoperative arrhythmias (Wetter VL):

Arrhythmia	Incidence (%)
Right bundle branch block	59.0 –100.0
Right bundle branch block + left anterior hemiblock	7.0 – 25.0
Atrioventricular block gr III	1.0 – 2.0
Supraventricular paroxysmal tachycardia/Atrial flutter	10.0 – 15.0
Premature ventricular contraction	30.0 – 67.0
Ventricular tachycardia	10.0 – 15.0
Sudden death	1.4 – 6.0

appearances of RBBB, RBBB + LAH and PVC in our study were within the range of the results shown in Table 2. We did not have pts with atrial flutter, VT, sudden death, and complete AV block. It was, perhaps, due to the high percentage of early repair in our group, namely 13 of 46 patients were operated on by the end of the first year of life, and 15 of 46 were operated on in the second year (total percentage of early repair was 60.86%). The remaining 18 of 46 repaired at the older age. All our pts are presently alive.

In her study of 89 pts with early repair Alexiou et al. (11) concluded that early repair of TOF was associated with an acceptable operative risk and the low incidence of significant arrhyhmias, and that it provided a long-term survival similar to that observed in general population.

Performing echocardiography, we showed right ventricular dilatation in 31 (67.39%) pediatric patients; tricuspid regurgitation in 6 (13.04%); L-R shunt across the VSD in 4 (8.7%), significant right ventricular outflow obstruction (Pg > 60mm Hg) in 1 child (2.17%) and significant pulmonary regurgitation in 5 (10.86%) pts.

After the repair of TOF postoperative ventricular arrhythmias and tachycardia (defect with a long standing right-ventricular pressure or volume overload with resulting fibrosis and dysfunction), or atrioventricular block occurred most frequently. Ventricular arrhythmias occurred most frequently in children in whom repair was done at an older age, and had poor hemodynamic repair with ventricular dysfunction. Residual hemodynamic problems may affect especially significant residual pulmonary insufficiency; residual VSD, ventricular outflow obstruction and tricuspid regurgitation (12–14).

Holter ECG analysis showed ventricular arrhythmias of the Lown type grade I-III in 9/46 and of grade IV in 2/46 pts (summary - total percentage of PVC was 23.90 %). All of the pediatric patients were asymptomatic, PVC was found neither in clinical examination nor in 12-channel ECG before Holter monitoring. One child with PVC (Lown IV) on Holter had also AV block grade I. None of our pts had ventricular tachycardia. None of them experienced sudden death. Nine of 11 pts with PVC required treatment with antiarrhythmic drugs.

Using statistical methods (cross-tabulations), we found that all 9 pts who had PVC (Lown grade I -III) and one patient who had PVC (Lown grade IV) on Holter ECG had right ventricular dilatation on echocardiogram. Other echo hemodinamyc parameters (tricuspid regurgitation, residual pulmonary insufficiency, residual VSD, and ventricular outflow obstruction) were not associated with the appearance of PVC.

Jonsson et al. (15) referred to a high percentage of PVC among his pts on Holter recording, namely 83% had PVC, with 27% PVC of Lown grade ≥ II. Joffe et al. (16) evaluating ventricular arrhythmia found out that 14% of pts had PVC at early follow-up, but 28% had PVC at late follow-up (similar to our results). In his study, late ventricular arrhythmia was not correlated with the estimated right ventricular dilatation, nor with right ventricular outflow obstruction, or with the degree of pulmonary incompetence. Dalliento et al. (14) in the study of 66 pts referred to 28.7% without ventricular arrhythmia, but 51.5% with minor PVC, 10.6% with ventricular tachycardia and 9% with SVT or ventricular fibrillation.

The incidence of PVC on routine ECG varied from 5 to 18%. Twenty-four-hour Holter ECG monitoring revealed ventricular arrhythmia incidence of 35–70% (19).

In our study, other types of arrhythmias and conduction abnormalities were also present. The child with SND had sinus atrial block and a significant number of PAC (both well tolerated). Eight children (17.39%) had significant PAC, requiring antiarrhythmic drugs. Two (4.34%) had transient nodal rhythm without complaints. SVP tachycardia was discovered in 3 (6.52%) with max heart rate of 201/min. Attacks were short during Holter monitoring without complaints, and did not need antiarrhytmic drugs. There were no pts with atrial flutter.

Gunal et al. (17) in the study of 31 pts operated on for TOF reported 39 % with supraventricular arrhythmias, three pts with SVPT, and 7 pts with rare PAC.

Conduction abnormalities of AV block type grade I was found in two pts. Those pts had AV block on Holter monitoring and on 12-channels EKG after surgery. The pts also had complete RBBB. Both were well tolerated. Complete AV block was not found in our pts.

Postoperative TOF patients were also noticed to have various supraventricular arrhythmias including sinus bradycardia, wandering atrial pacemaker, ectopic atrial rhythm, premature atrial contraction, supraventricular tachyarrhythmias, including atrial flutter, and junctional rhythm (18).

All our pts had RBBB (93.47% complete and 6.53% incomplete). Four of them (8.69%) had LAH, too. We had no patient with trifascicular damage (RBBB + LAH + AV block III).

Our results are similar to those in Jonsson's paper (15), who referred to 79/80 pts with RBBB, and 7/80 with RBBB + LAH.

The etiology of RBBB after repair of TOF attributed to the injury of the proximal right bundle branch as in courses along the rim of VSD, produced by repair of the VSD, or by delay or block in the peripheral right ventricular Purkinje fibers. Most cases of right bundle branch block with LAH occurred at the time of surgery and persisted on (19). Early reports in literature associated sudden death with conduction abnormalities, especially with the development of right bundle branch block, left anterior hemiblock, with presumed progression to the complete heart block (20, 21).

None of the pts in our study experienced sudden death.

A number of patients operated on for TOF experienced sudden death (22). Up to 30% of patients with ventricular arrhythmias after TOF experienced sudden death In recent studies, the presence of ventricular arrhythmias was noted as a factor associated with sudden death (23). Hokanson et al. (24) reported that late sudden death was

strongly associated with transient complete heart block that persisted beyond the third postoperative day.

Mean heart rate was in the range of frequencies for that age. Only one child (11.5 year) had minimal heart rate of 38/min during sleep (child with SND). Three children had maximal heart rate higher than 160/min (children with SVPT on Holter ECG); other 13 children had higher rate than 160/min during exercise at the time of the recording.

Conclusion

Twenty-four-hour Holter ECG monitoring is a noninvasive and very sensitive method for discovering heart rhythm disturbances in pediatric patients after the repair of tetralogy of Fallot, especially in asymptomatic patients. We found the presence of serious heart rhythm disturbances: ventricular arrhythmias in 23.91%; supraventricular arrhythmias in 28.26%; sinus node dysfunction in 2.17%; conduction abnormalities of the type AV block grade I in 4.34 %; and RBBB in 100% of the patients. All the 11 pts with premature ventricular contraction on Holter ECG had right ventricular dilatation on echocardiography. There was no relation of other types of arrhythmia, found on Holter, with other parameters from echocardiography, nor with the other standard methods. No child needed the application of pace-maker, but 36.95% children required antiarrhythmic drugs. No patients experienced sudden death.

Patients after the repair of this congenital heart disease needed a long-term follow-up for early recognition of serious heart rhythm disturbances and their treatment.

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A p s t r a k t

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ARITMIJE I POREMEĆAJI SPROVOĐENJA KOD DECE POSLE KOREKCIJE TETRALOGIJE FALO

Cilj. Otkrivanje vrsta i učestalost i aritmija i sprovodnih anomalija srca u grupi dece operisane od tetralogije Falo (TOF) u detinjstvu. **Metode.** Analizovano je 46 bolesnika koji su imali kompletnu reparaciju TOF u uzrastu od 1 do 13 god. (srednje 2,89 ± 2,36). Totalnu korekciju imalo je 38 (82,60%) bolesnika, a 8 (17,40%) prvo palijativnu pa zatim totalnu korekciju. Kod svih 46 bolesnika od 1 do 16 godina (6,48 ± 4,04) nakon operacije primenjeno je 24-časovno praćenje EKG-a po Holteru i to kod jednog deteta (2,17%) posle jedne godine, kod 20 (43,47%) posle 2 do 5 godina i kod 25 (54,34%) bolesnika posle 5 godina. Metoda je primenjivana u srednjem uzrastu 9,25 ± 4,39 (4–19). Svi bolesnici su bili pregledani standardnim metodama (klinički urađen je EKG pre operacije, EKG pre aplikacije primene aparata po Holter-u i 2D Dopler ehokardiografija). **Rezultati.** Putem Holter EKG-a otkriveni su sledeći tipovi poremećaja srčanog ritma: disfunkcija sinusnog čvora kod deteta 1 (2,17%), značajne pretkomorne ekstrasistole kod 8 (17,39%), supraventrikulska tahikardija kod 3 (6,53%), ventrikulske ekstrasistole (PVC) Lown gr I-III kod 9 (19,56%) i Lown IV kod 2 (4,34%), atrioventrikulski (AV) blok gr I kod 2, blok desne grane kod svih 46 (100%) i bifascikularni blok kod četvoro (8,96%) dece. Nije bilo bolesnika sa lepršanjem pretkomora, ventrikulskom tahikardijom i kompletnim AV blokom. Nije bilo iznenadne smrti ni kod jednog bolesnika. Svi bolesnici sa PVC imali su dilataciju desne komore na ehokardiogramu. Nisu pronađeni odnosi između ostalih tipova aritmija otkrivenih po Holteru i ostalih ehokardiografskih parametara, kao ni drugih standardnih metoda. **Zaključak.** Dvadesetčetvoročasovno praćenje po Holter EKG je neinvazivna veoma senzitivna metoda za otkrivanje poremećaja srčanog ritma kod dece operisane zbog TOF, posebno kod asimptomatskih bolesnika. Posle operacije ove srčane mane neophodna je dugoročna kontrola bolesnika zbog ranog otkrivanja ozbiljnih poremećaja srčanog ritma i njihovog lečenja.

K lj u č n e r e č i : **tetralogija Falo; hirurgija, kardijalna, procedure; aritmija; srce, provodni sistem; elektrokardiografija, Holter.**

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