Adult patient after correction of tetralogy of Fallot – diagnostic and therapeutic issues

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Abstract: Over the last 40 years, we have witnessed an impressive revolution in the management of congenital heart disease (CHD). The major factors contributing to this advance include: enhanced diagnostic abilities, improved surgical procedures, sophisticated intensive care and finally better understanding of natural history of CHD. Recent epidemiological studies have documented that although CHD still remains the principal cause of death among infants, a significant increase in the number of patients with heart defects who survive into the adulthood have been observed. Nowadays, for the first time, the number of adults with CHD equals the number of children with this defect. Tetralogy of Fallot (TOF) is one of the most frequent congenital cyanotic heart diseases treated with either corrective or palliative surgical techniques. Successful operation followed by specialized care enable patients to have almost normal life expectancy. In the current paper, we have provided the update on the management, diagnostic approach and treatment of adult patients after the TOF correction. Figures present typical problems related to the echocardiographic evaluation.

Key words: congenital cyanotic heart disease, survival of patients after TOF correction, tetralogy of Fallot, total or palliative correction

INTRODUCTION

The great progress achieved in the last decades in cardiology, cardiosurgery and anesthesiology has considerably improved the survival rate and prognosis for infants with congenital heart disease (CHD). Abnormalities in heart structure are recognized in 1 of 100–150 new-born children with the slightly higher prevelance of male sex [1]. Considering the above rate has not changed over the years, it has been estimated that during the last 25 years the CHD mortality has decreased by about 40%. Despite the fact that the highest mortality reduction concerns patients under the age of 5, the group is still characterized by the highest mortality rate. In the 1950s only every fourth infant with the CHD survived the first year of life, while nowadays 85% of the CHD patients achieve adulthood. For the first time the number of adults with the CHD is close to and in a short time will outnumber the CHD children population [2]. It has been estimated that there are 0.8–1 million adults with the CHD in the USA, about 100 thousand in Great Britain and both populations are increased by 25 and 3 thousand of patients coming of age every year, respectively [1,3]. It is supposed that the population of adult patients with the CHD will be a new challenge for contemporary cardiology. Patients with the CHD after surgical correction are characterized by a good overal life expectancy. However, dynamic, progressive hemodynamic disturbances cause that a considerable percentage requires further cardiosurgery procedures during the lifetime [3]. Certainly, the medical follow-up in special, experienced centers is also required. The surveys show that only 26% of the CHD adults are liable to a regular cardiology control [4]. These data need to be improved – patients after the CHD correction should be aware of the importance attributed to cardiology surveillance. The general practitioners, internists and cardiologists in turn, should know the specificity of this disease.

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Tetralogy of Fallot

Tetralogy of Fallot (TOF) consists of the following anatomical heart defects: the right ventricular outflow tract (RVOT) obstruction, ventricular septal defect (VSD), the over VSD aortal reposition and right ventricular hypertrophy [5]. It was described in 1888 by a French scientist Ethienne Fallot. Moreover, the presence of the atrial septal defect co-forms the penthalogy of Fallot. At the base of this pathology lies the incorrect evolution of ventricular bunch. The RVOT obstruction may be infundibular, valvular or usually the combination of both [5].

Even if the genetic origin has not been identified yet, in some groups of patients the association of the TOF and deletion in the 22 chromosome or the DiGeorge and Goldenhar

syndromes have been observed [6].

The TOF remains the most frequent cyanotic heart defect over the age of 1 and represents about 10% of congenital heart diseases. It is characterized by a marked variability of morphological, pathophysiological and symptomatic features and the progression depends on the RVOT obstruction. The consequence of higher right ventricular pressure in association with a large VSD is the compensation of ventricular pressure. Higher right ventricular pressure causes the right-left blood flow and the decrease of oxygenized blood level, and finally central cyanosis.

The abnormalities during the physical examination, the ECG or the chest X-ray suggest the CHD. Nevertheless, the diagnostic method that allows the correct diagnosis, preoperative evaluation and a long-term post-intervention follow-up is echocardiography.

The prognosis for patients who did not undergo the correct treatment is poor and the most common causes of death are: pulmonary haemorrhagia, brain abscess and thromboembolic complications. Some TOF infants require an early (4–6 month of life) surgical correction. In the first year of life every fourth of no operated infants dies. By the age of 10 the percentage of children who die is 70% and by 40 every twentieth is alive [5].

The surgical treatment of the TOF consists in a complete correction or a palliative operation. Total treatment is primarily based on the VSD closure and the RVOT narrowing correction. Moreover, the closure of foramen ovale, the VSD, the ASD or aorto-pulmonary shunts is necessary.

However, the palliative procedures involve many different aorto-pulmonary connections being able to increase the pulmonary blood flow and thus, to decrease the tissue hypoxia. From many types of these connections the following should be mentioned: the Blalock-Taussig shunt (subclavic artery-pulmonary artery), the Waterston-Cooley shunt (ascending artery-right pulmonary artery), the Pott's procedure (descending artery-left pulmonary artery) and its variations. Palliative procedures also consist of the Brock procedure (infundibular resection), pulmonary valvulotomy and the shunt between right ventricle and pulmonary trunk without closing the VSD [5].

Patients after palliative interventions need to undergo the total correction after some time because of secondary pulmonary hypertension.

Patient after correction of tetralogy of Fallot

The largest number of the TOF patients underwent the defect correction in childhood. Adults with an untreated defect represent a negligible percentage and about 3% of them reach the age of 40 [5]. Patients after the defect correction during childhood can not be considered healthy and quite often cause significant problems for a leading physician. The main cause is the performed earlier complex cardiovascular intervention and its consequences. Depending on a type of intervention, there are varying late complications. While assessing the patient after the TOF surgical treatment, we should consider the possibility of:

1) pulmonary hypertension
2) pulmonary valve insufficiency
3) tricuspid value insufficiency
4) dilation and right ventricular failure
5) ascending aortic aneurysm
6) aortic value insufficiency
7) secondary left ventricular failure.

In contrast to serious complications mentioned above, adults are characterized by a long-lasting lack of overt clinical symptoms [1,3]. Thus, patients seek for help of the physician with delay and their main symptoms are: exercise dyspnoea or limited exercise tolerance and periodically occurring palpitations. One of the symptoms forcing patients to visit the physician may also be syncope that exists as the most common manifestation of complicated ventricular heart rhythm abnormalities. It is necessary to mention that the ventricular heart rhythm abnormalities seriously increase the risk of sudden heart death and shows the symptoms of fainting only in a group of 10–15% of the examined [3,5,8].

The physical examination of the TOF patients in most cases reveals some heart murmurs such as: the pulmonary regurgitation murmur, sometimes coexisting with the aortic regurgitation murmur and also the holosystolic murmur in the case of the coexisting residual VSD. In consequence of proceeded vessel junctions the permanent systolic-diastolic murmur may occur. Incidentally, in patients with the progressing RVOT narrowing, the occlusion of pulmonary conduits or in the case of pulmonary hypertension, the cyanosis may occur [5].

The ECG at rest and the Holter recordings constitute the fundamental elements of follow-up examinations. All of them are used to detect early ventricular and supraventricular beats, atrial flutters and fibrillation incidents and nonsustained, rarely sustained, ventricular tachycardia. The signs of right and eventually left ventricular overload may also occur.

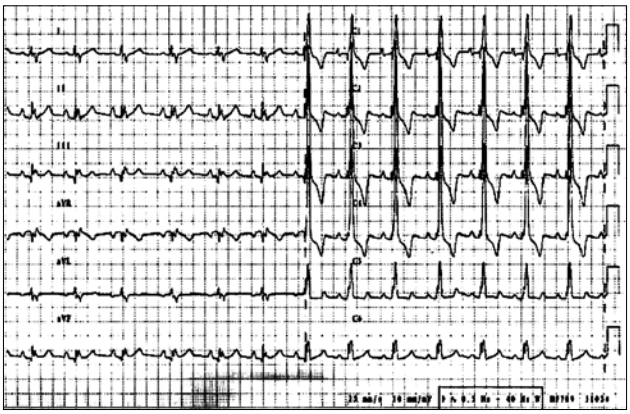

Fig. 1. 12-lead ECG in a patient after correction of tetralogy of Fallot

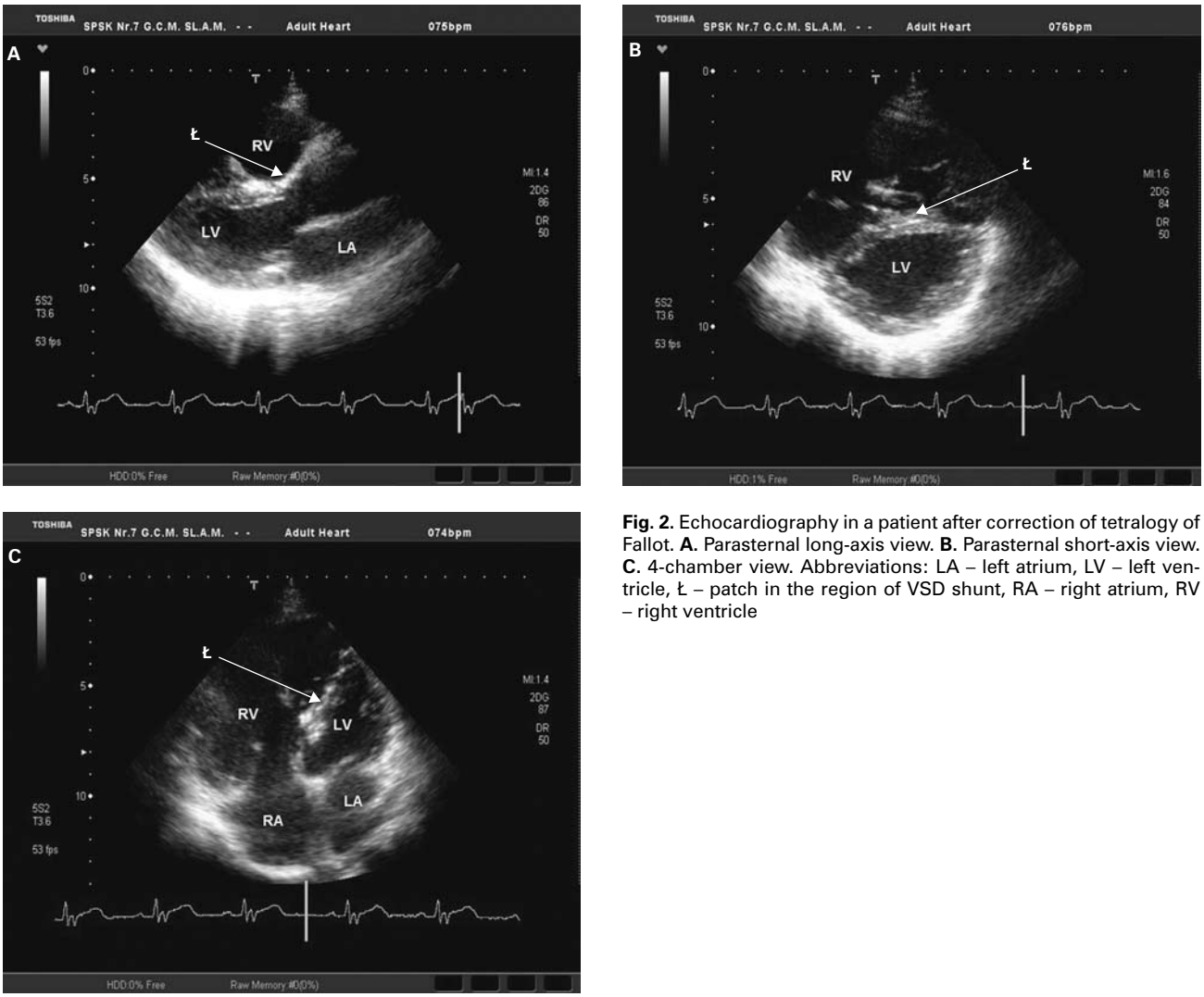

Fig. 2. Echocardiography in a patient after correction of tetralogy of Fallot. **A.** Parasternal long-axis view. **B.** Parasternal short-axis view. **C.** 4-chamber view. Abbreviations: LA – left atrium, LV – left ventricle, Ł – patch in the region of VSD shunt, RA – right atrium, RV – right ventricle

Conduction disturbances are very common, especially in the form of the right bundle branch block (fig. 1). The assessment of the widened QRS complex is indicated. The above parameter is considered as an independent risk factor of sudden heart death. The period of the QRS complex lasting over 180 ms is particularly unfavorable with regard to prognosis. The increased frequency of complex ventricular arrhythmias are then being observed [8-12].

The episodes of syncope and pharmacological treatment resistance, quite often observed, show a great possible role of the electrophysiological examination and ablation procedures. However, that kind of procedures is rather rarely performed, partially because of technical difficulties resulting from surgically changed anatomy.

A standard radiological diagnosis in the case of adult patients after the TOF correction may only be useful in same particular cases. In patients after palliative procedures, increased pulmonary flow (Blalock-Taussig shunt) or pulmonary hypertension features (Waterston-Colley or Pott's shunt) as well as the heart enlargement in the case of correction performed in the past, resulting from right ventricle dilation or/and ascending aortal extension, may appear [5].

The basic diagnostic tool revealing the essence of existing disorders is still echocardiography. There are some parameters that should be paid attention to (tab.1, fig. 2,3). The evaluation of the residual RVOT obstruction, pulmonary and tricuspid valve insufficiency or the residual VSD are absolute indications. The functional assessment of the right and left ventricles and coexisting aortal insufficiency is also indicated. In patients after the palliative correction the possibility of turbulent constant blood flow through aorto-pulmonary junctions imaging (with the suprasternal view using Doppler and colour Doppler imaging) should be taken into account [5]. Echocardiography in patients after the congenital heart defect correction is not an easy task. Firstly, it is necessary to deter-

Table 1. Heart structures of special interest in echocardiography assessment

Heart structures	Parametr/pathology
right ventricle	diameters/dilatation and dysfunction systolic function right ventricular outflow tract/stenosis
interventricular septum	patch in defect region/residual shunt
pulmonic valve	systolic flow/degree of stenosis diastolic flow/insufficiency
tricuspid valve	systolic flow/insufficiency
aortic valve	diastolic flow/insufficiency
ascending aorta	diameter/aneurysm

mine the type of procedure and what needs to be evaluated. The secondary pulmonary valve insufficiency remains often undetected despite being large in some cases (fig. 3).

In some circumstances during the follow-up examination of adults after the TOF correction, the invasive angiography is necessary to perform. It is indispensable in situations when the procedure of pulmonary valve replacement or dilation of pulmonary artery stenosis is planned. To evaluate the shunt patency, the invasive diagnostic method is also necessary in patients after the palliative treatment with increasing number of complaints [1,5].

Very promising new diagnostic techniques in the case of the CHD are multislice computed tomography and cardiac magnetic resonance that allow a detail imaging of heart structures and great vessel topography [13]. They seem to be of great importance in the CHD, also after a surgical correction. Especially magnetic resonance is optimal to assess both heart structure and heart valve functions [13,14]. There are some data in literature on pulmonary valve insufficiency assessment by cardiac magnetic resonance in patients after the TOF correction [15].

Adults after the TOF correction treatment need a special attention. Both in the case of a reconstructive operation and palliative procedures the prolonged improvement is obtained. However, unnoticeable haemodynamic disturbances occur. Patients that underwent the surgical systemic-pulmonary shunt in childhood, depending on its type, in adulthood may reveal the pulmonary hypertension, distal (one or both) pulmonary artery stenosis, left heart volume overload and heart failure, artery-venous and veno-venous collaterals evolution and aneurismal junction dilation [1,5]. The preferential flow from the aorta to pulmonary circulation through the junction may also lead to the RVOT narrowing progression [1]. On the other hand, in the case of reparative procedures performed in the past, late consequences might be the serious pulmonary valve insufficiency, secondary right heart overload with its dilation and secondary tricuspid insufficiency. Both groups of patients need to be operated on for the second time after few years from the first correction. In the case of vascular junctions cre-

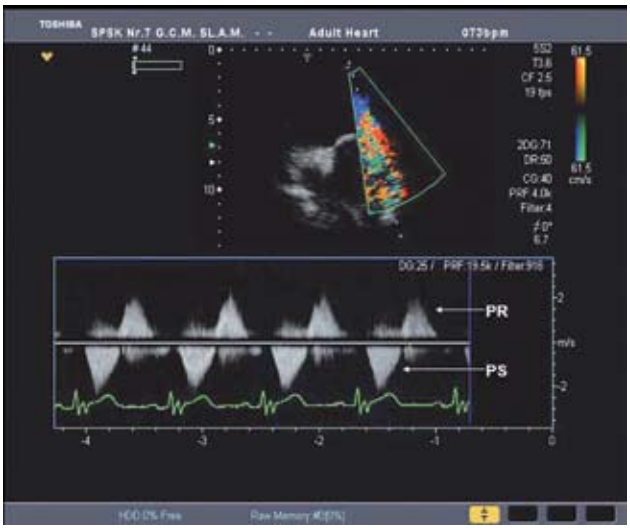

Fig. 3. Echocardiography in a patient after correction of tetralogy of Fallot with pulmonary valve insufficiency – Doppler assessment of pulmonic valve flow. Abbreviations: PS – pulmonic valve stenosis, PR – pulmonic valve regurgitation

ated earlier a complete reparative treatment is enough, and in the other case of serious pulmonary insufficiency – pulmonary valve replacement using homograft might be needed. Considering the last example it becomes very important to find a precise moment to perform the procedure, just before the right ventricular dysfunction occurs. Choosing an adequate time for valve replacement is also important from the point of a homograft survival (about 10 years) in the face of future operations. Unfortunately, the research findings prove that the procedure, with only a 1–2% surgical risk, is in many scientific centres performed too late, and is also aggravated by the lack of common indications which help in choosing the proper time for performing this procedure [16-18].

Another difficult issue is the therapy of arrhythmias. Despite the fact that the substrate for this pathology can be found in the right heart anatomical changes and haemodynamic disorders, and the causal surgical treatment is indicated, persistent arrhythmias, in spite of the performed correction, may occur. Atrial arrhythmias are diagnosed quite frequently (in about 1/3 of patients) and complicated ventricular arrhythmias (especially persistent ventricular tachycardia) become a problem. Considering the risk of sudden heart death, the patient's identification using electrophysiological examinations becomes prognostically significant. Unfortunately, even aggressive stimulation protocols very often show negative results [7,19]. Moreover, even in the case of severe ventricular arrhythmias and the administration of pharmacological treatment, there is no proof that the lifetime in this group could be prolonged [7,20]. It seems that ablation or defibrillator implantation might be useful. In the first case the short-term treatment effect is suspected, and in the next one – difficulties with the evaluation of the proper time for the defibrillator

implantation because of the absence of precise implantation indications [7,20,21].

Heart failure treatment in patients after the TOF correction should be performed according to the general standards. However, it is necessary to highlight the absence of multi-centre studies related to the heart failure and arrythmia treatment in patients after the congenital defect correction. For this reason, standard procedures are transferred to the mentioned group of patients and this is why a physician's experience is of vital importance in effectiveness of therapy.

While discussing the treatment, it is necessary to note that patients after the TOF correction require infective endocarditis prophylaxis throughout the lifetime [1].

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