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An adult patient with tetralogy of Fallot and anomalous left anterior descending artery after conduit type of repair (RCD code: IV-2A.1)

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Abstract

Tetralogy of Fallot is one of the most common cyanotic congenital cardiac disease concerning 10% newborn with congenital heart failures. About 3% of those patients have anomalous left anterior descending coronary artery. We report a case of 21 – year – old patient with tetralogy of Fallot and with coronary artery abnormality after staged cardiac surgeries. Due to an anomalous coronary artery, crossing the right ventricule outflow tract, the patient needed to conduit type of repair. JRCD 2014; 1 (8): 19–26

Key words: tetralogy of Fallot, valveless conduit, coronary artery anomaly

The present patient had been a subject of a report published elsewhere [1]. A 21-year-old male patient with congenital heart disease – tetralogy of Fallot with coronary artery anomaly was admitted to our hospital. At 13-mounths of age, boy underwent left Blalock-Taussig shunt. At 4-years of age total corrective surgery including ventricular septal defect closure and implanting valveless conduit between right ventricle and main pulmonary artery was performed. At 5-years of age, angioplasty of pulmonary artery with homograft implanting due to its aneurysm and dilation of conduit simultaneously, was performed. Furthermore patient suffered from the mental regurgitation, bilateral cryptorchismus, chronic gastroesophageal reflux disease and oesophagitis. For the next 14 years the boy was under care of the Institute of Pediatrics and subsequently, when he reached 18 years of age, he became the patient of Grow-up Congenital Heart Outpatient Clinic of The John Paul II Hospital in Cracow. Up to date, he was treated by metildigoxin (0,1 mg per day) and omeprazol (40 mg per day).

Patient was admitted to the hospital due to chronic fatigue and reduced exercise tolerance. According to his mother's report he was in NYHA class II/III for a few months.

On examination, heart rate was 70/min, blood pressure 105/70 mm Hg, systolic murmur in left second intercostal space was heard – 3/6 according to Levine's scale. Respiratory rate was 17/min and vesicular sound was heard on lungs auscultation. Peripheral swellings were absent and no cyanosis was present.

ECG revealed sinus rhythm 70 bpm, right axis deviation, Q wave in III, aVF, inverted T waves in I, aVL and signs of RV enlargement (Figure 1). 24-hours Holter monitoring showed sinus rhythm, HR: average 79/min, max.115/min, min.56/min, PQ 200 ms, episodes of wandering peacemaker and episode of supraventricular rhythm (Figure 2).

The cardiopulmonary exercise (CPET) test was performed and it was finished after 12 minutes 13 seconds due to fatigue and dyspnea and showed reduced exercise tolerance with maximal oxygen consumption VO2 max 19,0 ml/kg/min (Table 1). The first CPET was conduced 6 months earlier – it was finished after 3 minutes and 20 second due to fatigue and dyspnea and showed reduced exercise tolerance with maximal oxygen consumption VO2 max 12,4 ml/kg/min.

The echocardiography showed:

- normal size and function of the left ventricle with residual ventricular septal defect
- enlargement of right atrium (22 cm^2) and right ventricle (34 mm), with septomarginal trabecule
- preserved systolic function of right ventricle
- right ventricle outflow tract – 25 mm with turbulent flow
- hypoplastic pulmonic valve with calcification, residual flow and with regurgitation

Conflict of interest: none declared.

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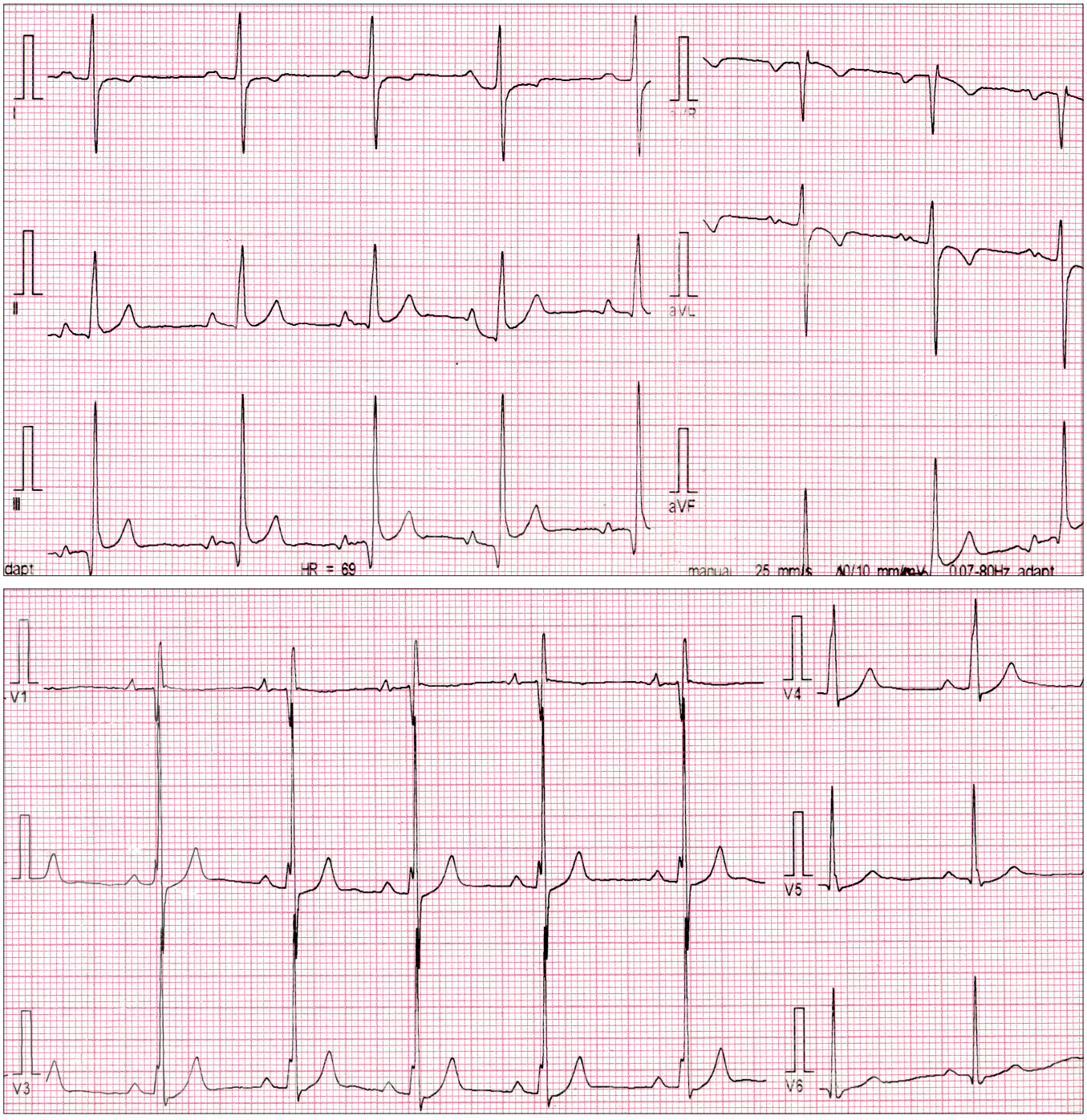

Figure 1. Electrocardiogram. Sinus rhythm 70 bpm, right axis deviation, Q wave in III, aVF, inverted waves in I, aVL and signs of right ventricular enlargement

– pressure gradient measured in the right ventricular outflow tract 32/19 mmHg which stayed stable during 2 years of observation (Table 2)
– diameter of conduit between right ventricle and main pulmonary artery – 15 mm with turbulent flow (Figure 3)
– mild tricuspid regurgitation, moderate aortic regurgitation.

Diagnostic was extended by computed tomography of the heart which revealed:

– enlargement of right atrium (27 cm^2) and right ventricle (34 cm^2),
– subvalvular stenosis (the narrowest diameter was 1,7 × 0,6 cms)
– hipoplastic pulmonic valve (diameter 2,0 × 0,7cms),
– main pulmonary artery 25 × 34 mm, right (15 × 9 mm) and left (16 × 10) pulmonic arteries,
– right ventricle and main pulmonary artery connected by conduit, with calcification in the middle part of conduit reducing its lumen to 1,9 × 1,5 cms (Figure 4)
– wide, permeable, connection of the conduit with right ventricle and pulmonary artery

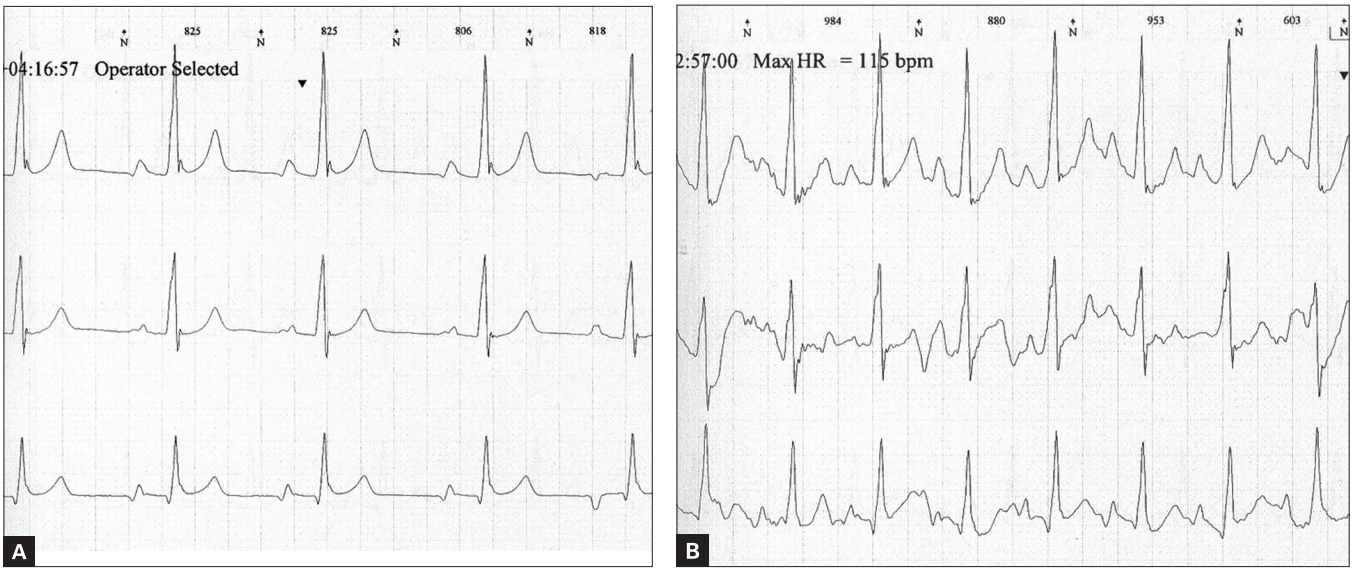

Figure 2. 24-hour Holter electrocardiogram. **A.** Episode of wandering peacemaker. **B.** Episode of supraventricular rhytm – 115 bpm lasting 30 seconds

Table 1. Cardiopulmonary exercise test

Parameter	Unit	Rest	MAX	MAX% PRED	PRED.	STATUS
Time	sec	0:00–2:04	12:31–13:03	-	-	-
Speed	km/h	0.0	4.8	-	-	-
Load	W	0	122	62	196	Low
HR	beats/min	86	122	61	193	Low
HRR	beats/min	106	76	-	<10	High
SBP	mmHg	120	140	68	180–230	Low
DBP	mmHg	80	60	75	70–90	Low
VO2	l/min	0.4	1.17	45	2.59	Low
VO2/kg	Ml/(kg*min)	6.4	19.0	44	42.7	Low
VCO2	l/min	0.39	1.16	35	3.35	Low
RER	-	0.97	0.99	85	1.07–1.25	Low
VE/VO2	l/l	21.0	22.9	62	36.7	Low
VE/CO2	l/l	21.6	23.0	81	28.5	Low
VE	l/min	11	31	29	109	Low
BR	l/min	109.9	89.4	-	>15.0	Normal
Bf	1/min	18	32	64	50	Low

Pred – predicted, HR – heart rate, SBP – systolic blood pressure, DBP - diastolic blood pressure, VO2 – oxygen uptake, VCO2 – carbon dioxide output, VE/VO2 – ventilation/oxygen output ratio, VE/VCO2 – ventilation/carbon dioxide output ratio, VE – minute ventilation, BR – breathing rate, Bf – breathing frequency

Furthermore, examination confirmed anomalous coronary arteries (Figure 5) – single artery originates from left sinus of Valsavia, coursing in front of the aortic valve, giving the right coronary artery, then courses in the right atrioventricular groove, encircle aortic valve and bifurcates to the anterior descending coronary artery and circumflex artery.

CMR was performed and it confirmed: the enlargement of the right atrium (31,5 cm^2), right ventricle (37,5 cm^2, EDV 192 ml), subvalvular stenosis (the diameter 1,7 × 0,6 mm) and normal sys-

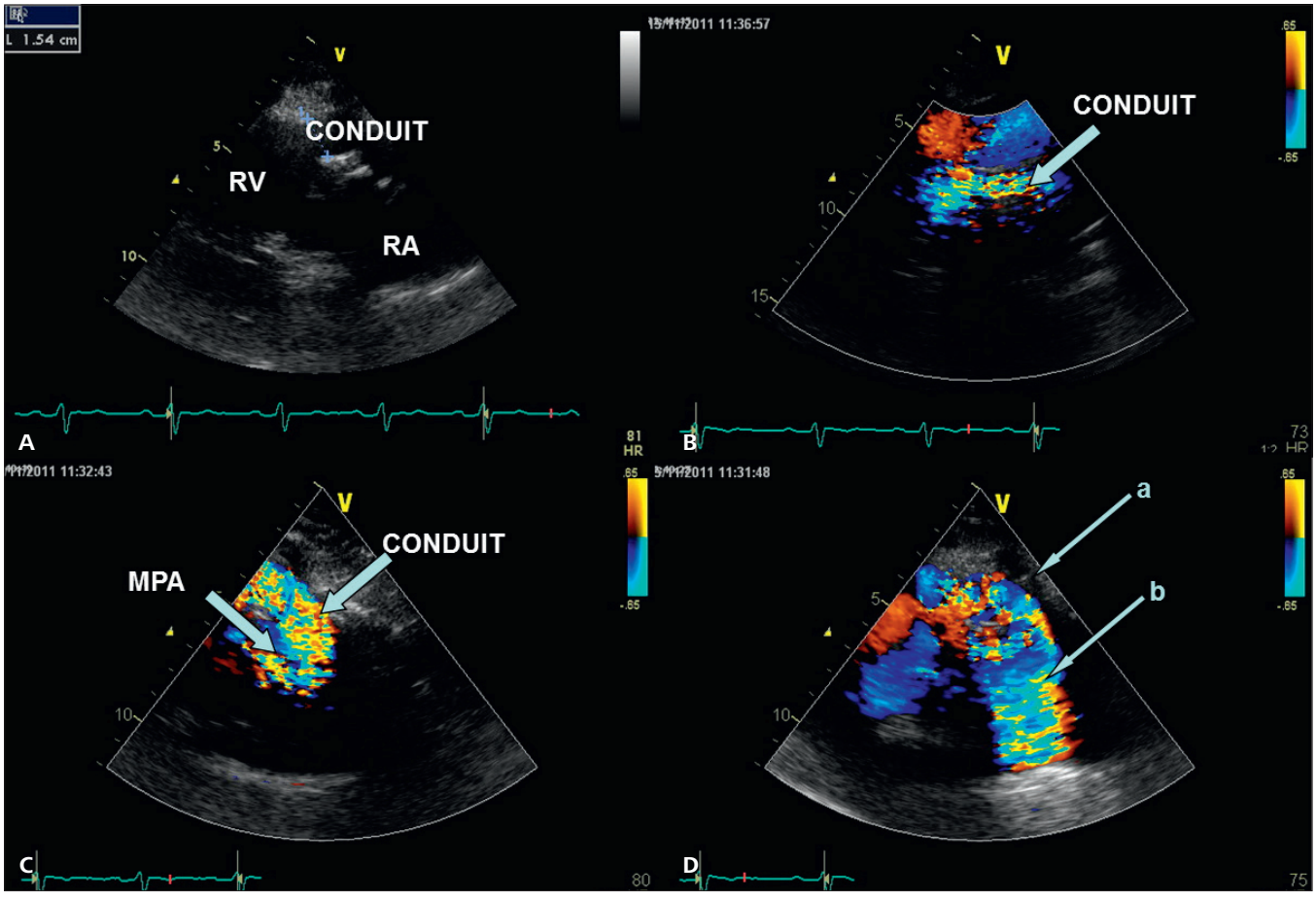

Figure 3. Transthoracic echocardiography. Conduit between right ventricle (RV) and main pulmonary artery (MPA). **A.** Connection of the conduit with right ventricular outflow tract (RVOT), dimeter 15 mm. **B.** Conduit with turbulent flow. **C.** Connection of the conduit with MPA. **D.** Turbulent flow in the conduit (a) and MPA (b). RV – right ventricle, RA – right atrium, MPA – main pulmonary artery, RVOT – right ventricular outflow tract

Table 2. Pressure gradient measured in the right ventricular outflow tract

	January, 2011	July, 2011	November, 2011	May, 2012
RVOT (mm Hg)	28.5/17	55/32	30/15	31/19

RVOT – right ventricular outflow tract

tolic function of right ventricle (EF 62%). The examination revealed also wide permeable connection of the conduit between right ventricle and main pulmonary artery, the narrowing of right and left pulmonary artery and overloading of right ventricle (late gadolinum enhancement in the connection of the right ventricular posterior wall and interventricular septum).

Due to the differences between symptoms observed by the patient's mother and the obtained results of the examinations performed in the clinic, the case was presented on the Experts Committee for the discussion to decide the best way of treatment. As a result the patient was selected for observation and catheterization.

Discussion

Tetralogy of Fallot is one of the most common cyanotic congenital cardiac disease. This heart malfunction consists of ventricular septal defect, obstruction of the right ventricular outflow tract which can be valvular, subvalvular or composite of both, overriding aorta and right ventricular hypertrophy [2]. About 3% of patients have anomalous coronary artery. An anomalous coronary artery, crossing the right ventricule outflow tract, makes the reconstruction of RVOT by using a trans – annular patch impossible and it may necessitate a conduit type of repair. Conduits connect right ventricle and main pulmonary artery and they have enabled to repair previously uncorrectable congenital heart failures. They can be nonvalved or valved – xenograft, bioprosthetic valves, bovine jugular vein (Contegra) pericardial or pulmonary/ aortic homograft.

The first conduit (nonvalved) was used and placed in 1964 by Rastelli and co-workers in 6-years old child with pulmonary atresia [3]. After two years Ross and Sommerville used an aortic valve homograft for the correction of pulmonary atresia [4]. Durability of the conduits is unfortunately limited by lack of growth, aneurysmal dilatation or degeneration and calcification. Conduit can become

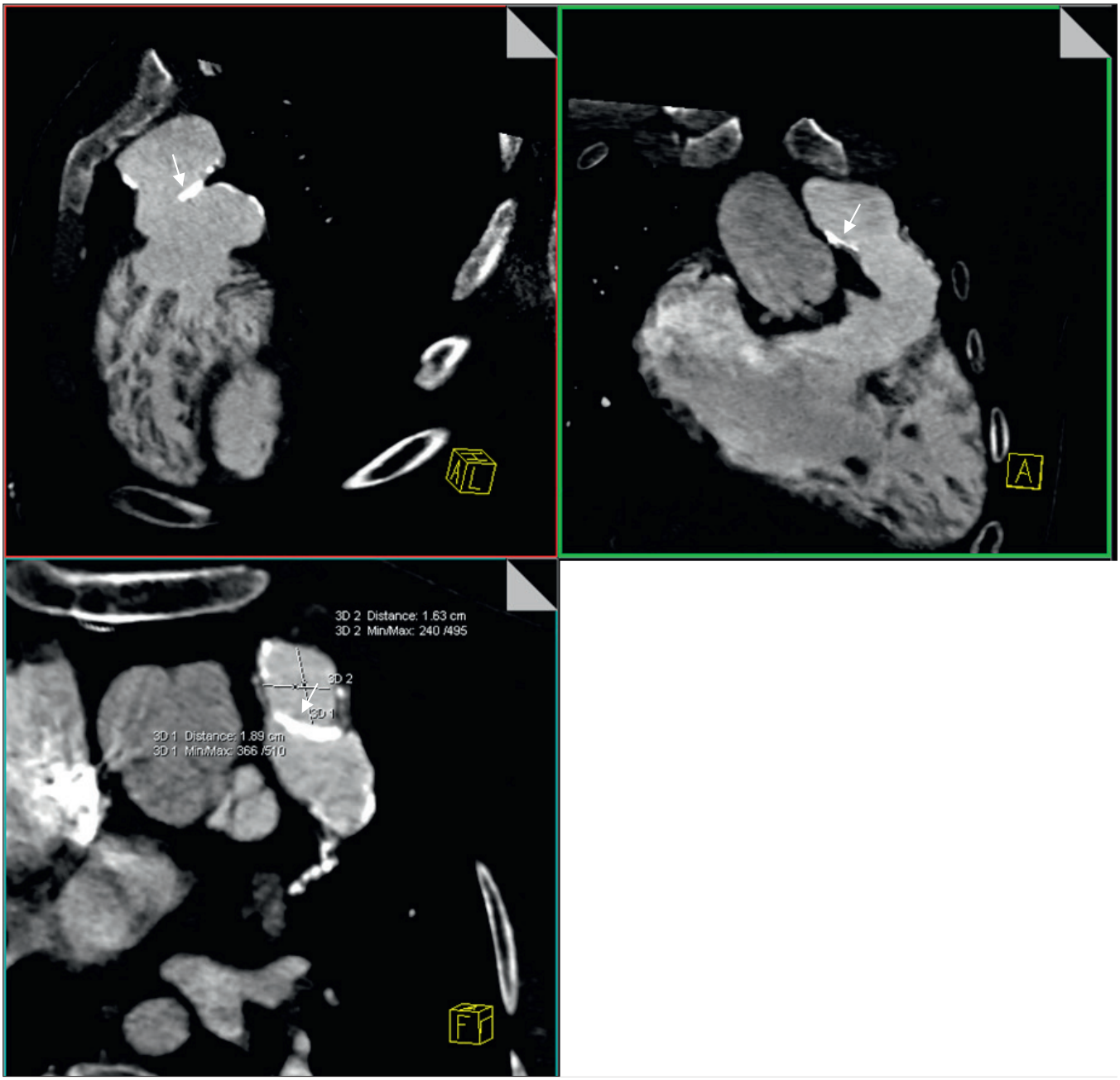

Figure 4. Computed tomography. Conduit connecting right ventricle (RV) with main pulmoanry artery (MPA) with calcifications (arrow)

dysfunctional and it may undergo obstruction and/or regurgitation. It is associated with such effects as right ventricular dilatation and its systolic and diastolic dysfunction, extension of tricuspid annulus and in consequence tricuspid regurgitation. In the long term it may cause arrhythmias, heart failure and sudden death [5]. About 40% of the patients with conduit does not require reoperation within upcoming 20 years whereas rest of the patients are affected by the hemodynamic consequences of conduit failure and need surgical procedure due to conduit failure [6]. Noteworthy, in some studies the lifespan of conduits is reported to be less than10 years [7–9]. Thus, the majority of the patients with conduits from the RV to PA undergo multiple surgical operations during their lifetime.

The most common symptoms reported by patient with conduit failure are: dyspnea, palpitations, syncope. The patients with the comparable degree of conduit failure usually experience the symptoms subjectively and their tolerance varies between the individuals. The subjects with mental disorder constitute a substantial group patients with congenital heart failure. What is more, they represent a special diagnostic difficulty, due to the lack of proper cooperation with a physician. That is why the objectivation of the symptoms, yet indispensable, is usually difficult. The clinical examination, such as cardiopulmonary exercise testing, including such parameters as time of exercise, maximum oxygen uptake, ventilation efficiency (VE/VCO2 slope), chronotropic and blood pressure response, exercise-induced arrhythmias, which usually enables

Table 3. Indication for intervention in patients after repair of tetralogy of Fallot [12]

Indications	Class	Level
Aortic valve replacement should be performed in patients with severe AR with symptoms or signs of LV dysfunction	I	C
Reoperations should be performed in symptomatic patients with severe pulmonary regurgitation and/or stenosis (RV systolic pressure >60 mmHg, TR velocity >3.5m/s)	I	C
Reoperations should be considered in asymptomatic patients with severe pulmonary regurgitation and/or pulmonary stenosis when at least one of the following criteria is present: ■ Decrease in objective exercise capacity ■ Progressive right ventricular dilation ■ Progressive right ventricular systolic dysfunction ■ Progressive tricuspid regurgitation (at least moderate) ■ RVOTO with RV systolic pressure >80 mmHg (TR velocity >4.3 m/s) ■ Sustained atrial/ventricular arrhythmias	IIa	C
VSD closure should be considered in patients with residual VSD and significant LV volume overload or if the patient is undergoing pulmonary valve resurgery.	IIa	C

AR – aortic regurgitation, LV – left ventricle, RV – right ventricle, RVOTO – right ventricular outflow tract obstruction, TR – tricuspid regurgitation, VSD – ventricular septum defect

Table 4. Indication for intervention in patients with right ventricular to pulmonary artery conduits [12]

Indications	Class	Level
Symptomatic patients with RV systolic pressure >60 mm Hg (TR velocity >3,5 m/s; may be lower in case of reduce flow) and/or moderate/ severe PR should undergo surgery.	I	C
Asymptomatic patients with severe RVOTO and/or severe PR should be considered for surgery when at least one of the following criteria is present: ■ Decrease in exercise capacity (CPET) ■ Progressive RV dilatation ■ Progressive RV systolic dysfunction ■ Progressive TR (at least moderate) ■ RV systolic pressure >80 mm Hg (TR velocity >4,3 m/s) ■ Sustained atrial/ventricular arrhythmias	IIa	C

RV – right ventricle, TR – tricuspid regurgitation, PR – pulmonary regurgitation, RVOTO – right ventricular outflow tract obstruction, CPET – cardiopulmonary exercise test

for the assessment of fitness of the patients and can correlate with the mortality as well as morbidity of the subjects with congenital heart disease [10] is in this case insufficient. In such circumstances it is necessary to perform additional, supplementary examination. Among available techniques, magnetic resonance imaging (MRI) offers the valuable ability to investigate the heart both morphologically and functionally. MRI is considered as a more valuable technique than transthorasic echocardiography in assessing the right ventricle volume, ejection fraction and its outflow tract, as well as in the examining of the condition of the right ventricle – pulmonary artery conduit and the measurements of regurgitation of pulmonic valve.

For the patients with conduit failure with indications for intervention, catheter procedure (like balloon dilatation, stent implantation or PPVI) should be considered. In order to reduce the number of cardiac re-sugeries in the group of patients with progressive obstruction or regurgitation of the conduit, endovascular treatment is possible in some cases. Moreover taking into consideration, that those patients have already underwent a number of surgical procedures, and will probably suffer from more interventions in the future, a non-invasive approach is always preferable [11]. Surgery is preferred when additional interventions (like tricuspid annulus plastic) are concerned.

Up to date, the data about endovascular treatment in patients with tetralogy of Fallot and nonvalved conduit are limited.

One of the transcatheter treatment option of the stenosis of conduit is stent implantation. There are some studies which demonstrate that stenting the obstructed RV-PA conduits may reduce pressure gradient across conduit and RV systolic pressure [9]. According to the study, the reduction of the gradient was more significant in patients with higher base pressure and in those, who did not underwent the interventions on branches of pulmonary artery before [12]. The study included 241 patients, whose median basic gradient RV pressure and conduit gradient were 89 mmHg and 58 mm Hg, respectively. According to the study, conduit stenting enabled to delay the cardiac re-surgery for 3,9 years in the group of children older than 5 years, and for 2,7 years overall. The intervention significantly reduced RV systolic pressure (89±18 to 65±20 mm Hg) and peak RV-PA gradient (59±19 to 27±14 mm Hg). Unfortunately, the complication occurring frequently in patients who underwent the follow-up, was the fracture of the stent without any hemodynamic consequences, diagnosed during catheterization in 40% of patients. However, this intervention has a significant drawback, due

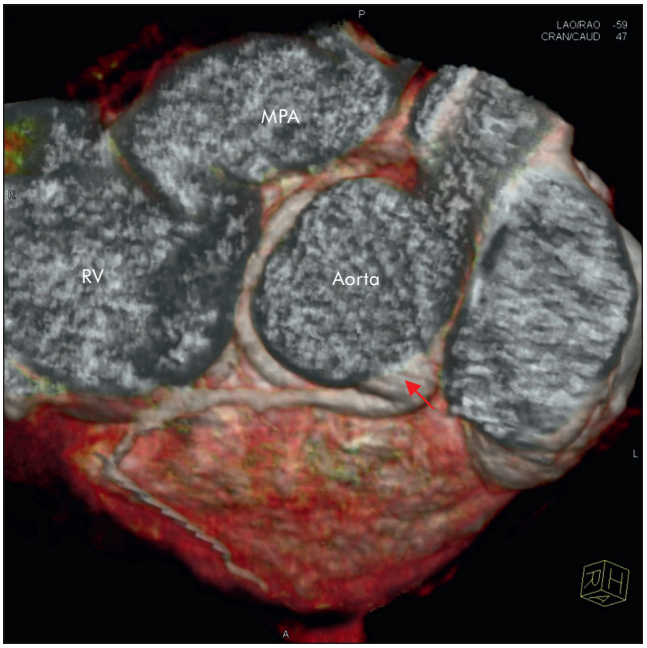

Figure 5. Computed tomography angiography. Single artery origin from left sinus of Valsavia (red arrow). RV – right ventricle, MPA – main pulmonary artery

to the fact that the relief of stenosis leads to the creation of free pulmonary regurgitation. During the stent implantion, the important part of the procedure is predilatation of the stenotic area. It allows for the determination of the location of the stenosis, eventual compression of the coronary arteries, with simultaneously coronary angiography, and it helps to limit the risk of stent malpostion.

The latest innovation, percutaneous pulmonary valve implantation (PPVI), is used with patients with dysfunctional conduit or homograft connecting right ventricle with pulmonary artery. It was implanted for the first time in year 2000 and since then more than thousand procedures have been performed worldwide. Currrently, the contraindications for PPVI are: the occlusions of central veins, active infection, outflow tract of native tissue and of unfavorable morphology (>22 mm diameter), or conduit <16 mm, unfavorable coronary anatomy (compression by the extended implant) [12].

The first results of the studies summarizing the effectiveness of percutaneous pulmonary valve implantation were recently published. The study showed the results of PPVI in 102 patients, which belonged to one of the three groups: pulmonic valve regurgitation (18 patients), pulmonic stenosis (36 patients) or combined dysfunction of conduit/pulmonary homograft [14]. After 352 days of observation 9% of patients needed re-catheterization due to PG> 50 mm Hg, in 5% of patients the fracture of the stent was observed, 1 patient was affected by endocarditis and 1 patient died. For the rest of the patients the reduction of pulmonary pressure gradient, pulmonary regurgitation and the improvement of right ventricle function were reported. Maximum oxygen uptake before intervention was 22,4 ml/kg/min and it has been stable during observation.

Eicken et al [13] also showed a significant reduction of the pressure gradient in the RVOT and indicated improvement of the right ventricle parameters within 1 to 12 months of the follow up after PPVI. A total of 65 patients were divided in two groups: first one included the patients with stenosis, whereas the dominance of regurgitation was grouped in the second one. The study showed also that systolic function and maximum oxygen uptake has been improved significantly only in the group of patients with stenosis.

In 2011 a 6- months prospective follow up was published. The study included 10 patients with RVOTO, who underwent Melody valve implantation and prestenting with BMS. [14]. After six months of observation after PPVI with prestenting with BMS, the reduction of RVOTO was associated with the significant decrease in right ventricular (RV) end-diastolic and end-systolic volumes, improvement in RV ejection fraction alongside with the decrease in New York Heart Association class assessment. Importantly, no stent fractures were observed.

In 2009, the study summing up the comparison of hemodynamic effects between PPVI and treatment of right ventricular outflow tract with a bare metal stent implanting (BMS) was published [15]. Fourteen patients, with congenital heart diseases (transposition of the great arteries, tetralogy of Fallot, double-outlet right ventricle) with significant right ventricular outflow tract obstruction, after various surgical interventions (Rastelli procedure, Ross procedure) underwent BMS followed by PPVI. The effects (ventricular volumes and function, great vessels flow and hemodynamic – invasive pressure measurements) of the procedures were assessed by using an an x-ray/magnetic resonance hybrid laboratory. The measurements were performed before BMS, after BMS, and after PPVI. According to the study: BMS significantly reduced the ratio of right ventricular to systemic pressure with no further change after PPVI. However, BMS resulted in free pulmonary regurgitation, which was eliminated after PPVI. Effective right ventricular stroke volume (right ventricular stroke volume minus pulmonary regurgitant volume) after BMS remained unchanged but was significantly increased after revalvulation with PPVI. The improvements after PPVI were accompanied by the significant reduction in the heart rate at maintained cardiac output. The study demonstrated the superior acute hemodynamic effects of PPVI over BMS in patients with right ventricular outflow tract obstruction.

During all kind of the above procedures, it is necessary to exclude potential possibility of the coronary artery damage during stent implantation what could be the cause of sudden death. Anomaly of the left anterior descending coronary artery arising from the right sinus of Valsalvia is frequent with tetralogy of Fallot, affecting 3% of patients. The right coronary artery originating from the left anterior descending coronary artery, which passes through the aortic root and the MPA, can be compressed by the stent. For that reason a proper estimation of coronary arteries tracts is necessary. In some cases the balloon test should be performed, because it can mimic the situation after stenting. The balloon inside the homograft should be firstly decompress to the diameter similar to the size of the valve or stent after the implantation to exclude the coronary artery compression by the placed implant. Selective coronarography should be simultaneously performed.

Patient management and follow-up

According to the ESC guidelines from 2010 year, for the management with adult patients with congenital heart disease our patient has periodic cardiac follow – up in Grow-up Congenital Heart Outpatient Clinic of The John Paul II Hospital in Cracow. Up until now, the patients is stable and he has no indications for intervation (Table 3, 4). Medical treatment was modified – metildigoxin was excluded and replaced by B-blocker (metoprolol CR 25 mg a day). If the symptoms be stable, he is planned to visit the clinic, every year.

Echocardiography and cardiac MR will be performed every year to look for the complications of the diseases such as: right ventricle outflow tract obstruction, pulmonary regurgiation, right ventricle dilatation and dysfunction, residual ventricular septal defect, aortic root dilatation with aortic regurgiatation, left ventricular dysfunction, endocarditis. In the case of progressive symptoms CT will be performed to image the conduit, pulmonary artery and coronary artery anatomy.

More over cardiopulmonary test will be performed to monitor such parameters as time of exercise, maximum oxygen uptake, ventilation efficiency (VE/VCO2 slope), chronotropic and blood pressure response, exercise-induced arrhythmias.

To indicate supraventricular or ventricular arrhythmias, which could be the cause of sudden cardiac death, ECG and holter monitoring, will be performed every visit. In case of new symptoms: syncope, presyncope, fatigue, reduce exercise tolerance it will be more frequent and EP testing will be considered.

Nowadays there is no contraindications for regular physical activity. To avoid infective endocarditis, an antibiotical prophylaxis is recommended.

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