Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

EXPERT: Professor Janusz Skalski, pediatric cardiac surgeon

Affiliation: *Department of Pediatric Cardiac Surgery, University Children's Hospital of Krakow, Poland*

CASE SUMMARY

The opinion focuses on a 25-year old female patient with a severe complex heart defect. After birth, she was diagnosed with transposition of the great arteries with pulmonary stenosis and ventricular septal and atrial septal defect. At the same time, she was found to present with signs of right ventricular hypoplasia, what was an indication for the bi-directional Glenn anastomosis (1993); what is puzzling is the delay in performing the said anastomosis (superior vena cava-right pulmonary artery anastomosis). At the time, the therapeutic team did not qualify the patient for the subsequent stage of treatment - the Fontan procedure - precisely due to her presenting with signs of moderate rather than complete right ventricular hypoplasia (borderline right ventricle), with the cardiac surgical intervention consisting solely in the Glenn anastomosis. For several years, the condition of the patient was satisfactory, with gradually decreasing arterial blood saturation and deteriorating exercise tolerance. None of the cardiac surgical centers in the country was willing to attempt to complete the multi-stage treatment consisting in the Fontan procedure. In 2009, a decision was made to perform the Rastelli procedure (directing the outflow from the left ventricle under a directional patch to the aorta, anastomosing the right ventricle with the pulmonary artery by an external valve homograft[1]), leaving, however, the Glenn anastomosis in situ. In subsequent years, the condition of the patient was good and she tolerated exercise well (NYHA I^{o}). No pharmacotherapy was indicated. Recently, the Holter monitoring demonstrated nocturnal episodes of bradycardia (33 bpm), and in ECG, she presented with atrioventricular dissociation and RBBB. Pharmacotherapy was unsuccessful. Echocardiography demonstrated hemodynamically insignificant tricuspid valve regurgitation, an insignificant RV-PA gradient with moderate homograft calcifications, insignificant residual VSD, insignificant LVOT stenosis, EF – 60%. A myocardial perfusion scintiscan and angio-CT were performed and demonstrated partial compression of the initial segment of the

John Paul II Hospital in Kraków
Jagiellonian University, Institute of Cardiology
80 Prądnicka Str., 31-202 Kraków;
tel. +48 (12) 614 33 99; 614 34 88; fax. +48 (12) 614 34 88
e-mail: rarediseases@szpitaljp2.krakow.pl
www.crcd.eu

left coronary artery by the conduit joining the right ventricle and the pulmonary artery. The scintiscan showed loss of radiotracer uptake in 18% of the myocardium (anterio-lateral, posterio-lateral segments and partially the inferior wall of the left ventricle). In spite of the detected deviations from the norm, the patient shows good exercise tolerance and does not suffer any pain.

DISCUSSION

It is difficult to comment on the treatment the patient received in her early childhood, since we do not have at our disposal complete information on the premises and assumptions underlying the then planned management strategy. We cannot find the answer to the question why no cardiac-surgical treatment was initiated at the time, and the first palliative surgery was performed only when the patient was five years old (1993). One may surmise that the observed since birth right ventricular hypoplasia (albeit moderate) seemed to be an excessive strain, dissuading the cardiac surgeons from performing the highly risky - as it was believed at the time - Rastelli procedure. A palliative procedure, i.e. the bi-directional Glenn anastomosis, was performed, resulting in achieving satisfactory arterial blood saturation and a relative improvement of the patient's general condition. The good adaptation of the patient to the employed cardiac-surgical solution does not justify the therapeutic team leaving the patient without any continuation of operative treatment for many years and their failure to reach any decision. It is the more baffling that the long-term **decreasing the load** on the right heart did not exert any beneficial effect on the possible further development of the right ventricle. Only when the patient did reach maturity, the gradual deterioration of her condition and physical capacity necessitated the search of a radical solution for her health problems.

The employed in 2009, when the patient was 21 years old, concept of surgical treatment consisting in partial correction of the defect by the Rastelli method combined with decreasing the load of the right heart by leaving in situ the functioning Glenn anastomosis was, as it seems, an optimal solution[2]. Such a method of combining palliation with a partial repair of the defect is employed in various variants of modern cardiac-surgical management of the most complex congenital heart defects[3]. Nevertheless, there are no unambiguous recommendations referring to management of an adult patient with a complex heart defect involving transposition of the great arteries, ventricular septal defect, significant pulmonary stenosis and concomitant partial left ventricular hypoplasia[4].

Considering that a complication in the form of ischemia associated with compression of the coronary artery is an absolutely exceptional phenomenon, in similar situations, determinants of the management strategy should be established individually. To date, no complications consisting in compression exerted by a grafted conduit on the coronary vessels system following the Rastelli procedure have been described. One might, however, anticipate that in some percentage of cases, the grafted homogenous conduit will develop highly advanced calcifications, which will in time lead to compression of the coronary artery that extends just underneath. As long as the conduit remains elastic and susceptible to mutual vascular interactions, one may surmise that the high-pressure flow in the coronary artery is not at risk of compression by the low-pressure RV-PA conduit - assuming the physiological flow and physiological pulmonary resistance – just as it happened in the analyzed case. Typical complications following the Rastelli procedure in an adult include an increasing gradient in the right ventricular outflow tract as well as arrhythmias with bradycardia and

RBBB (due to the past ventriculotomy)[5]. In case of a RV-PA conduit obstruction, the only rational treatment strategy is conduit replacement and such a management method is highly probable in the analyzed patient later in time – then, the compression of the coronary artery would be automatically relieved, if indeed it is present at all. At present, the gradient in the right ventricular outflow tract is insignificant and there are no firm indications for graft replacement.

EXPERT'S OPINION

In the present case, the compression of the coronary artery by the conduit is well tolerated by the patient, what is supported by her good exercise tolerance, general well-being, absence of pain and high left ventricular ejection fraction, while the only manifestation of the effects of the said compression is the scintiscan result, which, irrespectively of clinical symptoms, cannot be the only indication for surgical intervention. It cannot be ruled out that the symptoms of ischemia, the presence of which is confirmed by the scintiscan only, may be associated with consequences of long-term hypoxemia and secondary myocardial ischemia. We are not sure whether the condition was apparent previously (preexisting the last operation), since no scintigraphy imaging was performed.

It may be thus postulated that the performed surgical treatment ensures an incomparably better physical comfort than the planned years ago and considered Fontan procedure, which as a rule is the only radical therapeutic method in a single-ventricle heart. In this case, a successful attempt has been made at making the use of and loading the moderately hypoplastic right ventricle.

SUMMARY

In the discussed case, the result of the performed surgical treatment of the severe complex heart defect should be considered good. Satisfactory physical capacity of the patient has been achieved, as well as good exercise tolerance and high quality of life. Residual defects (trivial tricuspid valve regurgitation, trivial stenosis of the right ventricular outflow tract, moderate homograft calcifications, a small residual VSD, mild subaortic stenosis), the impact of which is insignificant for the circulatory function, do not affect the general assessment of the good therapeutic result.

In her present clinical condition, the patient should be subjected to elective coronarography and its result should be the only basis for deciding on possible interventional management. Episodes of bradycardia require further verification and initiation of radical treatment, including a possible implantation of an artificial pacemaker, in keeping with the obligatory standards.

REFERENCES

[1] Recommendations of ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Clinical Features and Evaluation: Dextro-Transposition of the Great Arteries After Rastelli Operation, JACC, 2008;52(23): 143-263 (**11.6. e224**).
[2] Elizari A, Somerville J.Cardiol Young. Experience with the Glenn anastomosis in the adult with cyanotic congenital heart disease.1999;9(3):257-65.
[3] DiBardino DJ, Heinle JS, Fraser CD Jr. The hemi-Mustard, bi-directional Glenn, and Rastelli operations used for correction of congenitally corrected transposition, achieving a "ventricle and a half" repair. Cardiol Young 2004;14(3):330–2.
[4] Warnes C.A. Congenital Heart Disease for the Adult Cardiologist. Transposition of the Great Arteries. Circulation, 2006; 114: 2699-2709.
[5] Chamaidi A., Griselli M., Triposkiadis F. Long-term outcome after Rastelli repair [in:] Gatzoulis MA, Webb GD, Broberg CS, Hideki U.: Cases in adult congenital heart disease. Churchill Livingstone, Elsevier, Philadelphia, 2009, p. 299-301.

John Paul II Hospital in Kraków
Jagiellonian University, Institute of Cardiology
80 Prądnicka Str., 31-202 Kraków;
tel. +48 (12) 614 33 99; 614 34 88; fax. +48 (12) 614 34 88
e-mail: rarediseases@szpitaljp2.krakow.pl
www.crcd.eu