Medical Expertise

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

EXPERT: Dr Jacek Kołcz, pediatric cardiac surgeon

Affiliation: *Department of Pediatric Cardiac Surgery, Jagiellonian University Medical College, Krakow, Poland*

CASE SUMMARY

This is a case of 46 year-old woman with double inlet left ventricle with the most frequent settings of atrio-ventricular and ventriculo-arterial connection, rudimentary right ventricle and rare physiology of well-balanced pulmonary and systemic circulation (1). The morphologically left systemic ventricle and transposition of the great arteries with subvalvular and valvular stenosis of the pulmonary outflow tract cause relatively good function of circulatory system and balance of the systemic and pulmonary blood flow (2). Although the complex defect was diagnosed in early infancy no surgical treatment was applied. With the mild pharmacologic support there are no symptoms of circulatory decompensation (NYHA II) assuring good quality of life (the woman gave a birth to three healthy children).

DISCUSSION

The natural history of the double inlet left ventricle with well-balanced systemic and pulmonary circulations and obstruction of the blood flow to the pulmonary circulation is favorable (3,1). Some patients survive without treatment for many years. Hager et al. ([4]) collected 10 patients over 50 years old from the literature; the oldest was 66 years old, reported by Ammash and Warnes ([5]). Seven of those 10 were alive at the time when they were reported. Since that publication, one patient who died aged 59 ([6]) and two survivors aged 57 years ([7]) and 71 years ([8]) have been reported. These patients are almost always those with enough pulmonary stenosis to give balanced circulations. Death is usually due to severe hypoxemia, congestive heart failure, infective endocarditis or brain abscess. Staged treatment of the defect is complex and for obtaining the best possible results must be initiated in early infancy and finished in early childhood. Unfortunately there are no studies comparing strategy of staged construction of Fontan circulation in the group of selected patients with DILV and discussed above favorable physiology versus strategy of doing nothing. There is also no

sufficient scientific evidence of successful late surgical procedures versus natural history of the defect. Taking into account both early and late complications after the Fontan surgery in this patient strict follow-up and pharmacological treatment will be the best option.

EXPERT'S OPINION

This is very rare case of single ventricle defect by means of double inlet left ventricle (DILV) with well-balanced pulmonary and systemic circulations with long-term survival without any surgical intervention. The natural history of this particular variant is favorable and there is no theoretical advantage of converting circulation into Fontan physiology at this stage. The strict follow-up and pharmacological treatment will be the best option for the patient. The elective cardiac catheterization and assessment of the pulmonary circulation should be taken into consideration. The prevention of infective endocarditis and antiplatelet or anti-coagulation therapy is indicated.

CONCLUSION

There is no recomendation for surgical treatment at this stage. Patient should be folowwed-up very strickly and treated pharmacologically. The elective cardiac catheterization and assessment of the pulmonary circulation should be taken into consideration. The prevention of infective endocarditis and antiplatelet or anti-coagulation therapy is indicated.

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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