

## Medical Expertise

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

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#### **CASE SUMMARY**

31 year old patient with tricuspid atresia type IIb, after staged surgical creation of TCPC (1985r.- Blalock- Tausig, 1988r.- classic Fontan procedure (atrio – pulmonary anastomosis), 1999r., conversion to the TCPC with hepatic vein exclusion, 2002r.- hepatic vein inclusion). The patient was many times hospitalised because of rhythm disturbances, cyanosis, heart failure, upper respiratory tract infections, and episodes of bleeding from gastro – intestinal tract. He also underwent ablation of accessory conduction pathways, he has hyperthyroidisms, (amiodarone induced), thrombocytopenia, poliglobuly, and underwent cleft palate surgery (1983r.), inguinal hernia surgery. He underwent virus hepatitis B, and has varices of the lower extremities. In physical examination: RR 115/80 mmHg, heart rate 90/min irregular rhythm, systolic murmur in the precordial area 3/6 in Levine’s scale, cyanosis, club fingers, scoliosis and pectus excavatus. In echocardiography apart from typical features characteristic for tricuspid atresia after Fontan surgery, the severe atrioventricular (mitral) valve insufficiency was noted, the left atrium was enlarged. In 24-hour ECG (Holter) examination – the atrial fibrillation and interventricular conduction disturbances were noted. Also ventricular tachycardia, accessory ventricular contractions (1164), and bradycardia episodes (37/min) during the night. After the ablation, the paroxysmal ventricular arrhythmia was eliminated, but there was no improvement in terms of atrial fibrillation (it was also confirmed in Holter 12.2013r.). In whole bodyplethysmography the restrictive ventilation impairment of mild degree was noted. Spiroergometry revealed 63% of maximal exercise capacity, the peak oxygen consumption was 18,7ml/kg/min, that’s 47% of normal value; VE/VCO<sub>2</sub> – was 38; also during the test desaturation was noted in pulsoxymetry (70 do 68%). Ultrasound investigation revealed symptoms of hepatic cirrhosis, hepatic veins congestion, and small amount of fluid in the peritoneal cavity |

## DISCUSSION

The Fontan procedure originally was introduced in treatment of tricuspid atresia<sup>(1)</sup> and than after numerous modifications it was applied to all forms of single ventricle congenital heart defects. At present the survival rate after 20 years of Fontan physiology approaches 85%<sup>(2)</sup>. During the last two decades, patients who were treated using original modification of Fontan surgery in tricuspid atresia (atrio – pulmonary connection) have reached the adulthood and developed hemodynamic and different type of organ complications of Fontan physiology. The atrio-pulmonary connection was abandoned as a method of treatment and majority of patients who developed congestive heart failure and major organ complications, underwent conversion surgery (to the total cavo – pulmonary connection or extracardiac conduit) with intraoperative antiarrhythmic therapy and / or heart resynchronisation<sup>(3)</sup>. In the presented case we see typical symptoms of Fontan circulation failure. This can be observed during the years and variety of therapeutic options available is scarce. Patients reveal growth retardation, physical activity restriction, systemic ventricle dysfunction, systemic atrio – ventricular valve insufficiency, arrhythmias, thrombo – embolic complications, effusions protein enteropathy and plastic bronchitis<sup>(4)</sup>. Congestion of the hepatic veins of some degree is present in almost all patients after Fontan surgery and increased central venous pressure after years can contribute to hepatic cirrhosis development<sup>(5)</sup>.

## EXPERT'S OPINION

In the present case the symptoms of Fontan circulation failure after conversion of atrioventricular connection to TCPC and hepatic inclusion are described. Organ symptoms of circulatory failure are noted despite of pharmacologic treatment. The therapeutic options which should be taken into consideration are mitral valve insufficiency (which is the main cause of circulatory failure) surgery and antiarrhythmic surgery (both intraoperative or direct videoscopic after surgery). The chronic pulmonary vasodilator therapy (sildenafil) should be introduced. Another option is heart transplantation.

## CONCLUSION

In patient after classic Fontan surgery and conversion to the TCPC with hepatic vein inclusion the symptoms of circulatory failure exist despite of pharmacological treatment. Apart from severe mitral valve insufficiency, the main cause of the circulatory failure are rhythm disturbances (persistent atrial fibrillation). The mitral valve plasty should be proposed to the patient with antiarrhythmic surgery and chronic pulmonary vasodilatory therapy should be introduced (sildenafil). The ultimate option is heart transplantation.

## REFERENCES

<sup>1</sup> Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971;26:240 e 8.

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<sup>3</sup> d'Udekem Y, Iyengar AJ, Cochrane AD, et al. The Fontan procedure: contemporary techniques have improved



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<sup>5</sup> Baek JS, Bae EJ, Ko JS, et al. Late hepatic complications after Fontan operation; non-invasive markers of hepatic fibrosis and risk factors. *Heart* 2010;96:1750e5.