

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

The presented patient is a 31-year old female with univentricular heart, in whom no procedures that are routinely performed in case of such a defect have been employed due to the lack of consent. In addition, 9 years ago, the patient had endocarditis, which resulted in moderate/significant tricuspid valve incompetence. The patient presents with central cyanosis with polyglobulia and peripheral lesions characteristic of cyanosis. Since the time her tricuspid valve incompetence increased, her physical ability has significantly deteriorated and at present, the patient is in NYHA class III. Presently, the possibilities of treatment are limited and oscillate between choosing the lesser of two evils. The patient may be proposed the following options: the high risk Fontan operation; the temporary hemi-Fontan procedure, which is associated with a lower risk, but does not result in significant improvement in blood oxygenation; the systemic-pulmonary anastomosis, which will initially improve blood oxygenation, but - due to an increased volume overload of the single ventricle - will intensify atrioventricular valve incompetence; and the heart transplant associated with an increased risk. We do not know to which treatment modality the patient will give her consent. From my viewpoint, the best solution for the patient would be the hemi-Fontan/Glenn procedure, after a time followed by the Fontan procedure or a heart transplant.

CONCLUSION

In such a complex situation the best solution for the patient would be the hemi-Fontan/Glenn procedure, after a time followed by the Fontan procedure or a heart transplant.

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