Medical Expertise*

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

EXPERT: Prof. Piotr Podolec, cardiologist

Affiliation: *Department of Cardiac and Vascular Diseases, John Paul II Hospital, Krakow, Poland*

CASE SUMMARY

A 30-year-old women with an congenitaly corrected transposition of great arteries . At the age of 10 years the heart malformation was diagnosed. The patient had not been systematically carried by the cardiologist. The patient has manifested symptoms of heart failure NYHA II. Cardiopulmonary exercise test show very good exercise tolerance with peak oxygen consumption 24 ml/kg/min. MR showed an increased volume of systemic ventricle with decreased systolic function. On admission the patient's general condition was good. Physical examination revealed no enlargement of the liver no odema. Laboratory studies have found: E – 4600000, Hg- 13,7 g/dl, Ht- 39,6%, PLT- 170 000, INR- 0,89, ASPAT- 21 U/L, ALAT- 14 U/L, chol 3,91 mmol/l, LDL 2,16 mmol/l, pBNP – 180pg/ml, glucose 4,22 mmol/l. Holter monitoring revealed sinus rhythm with single ventricular and supraventricular extrasystolies (atrial fibrilation). Echocardiography showed: situs solitus, dextrocardia, impaired function of systemic (anathomical rigt) ventricle, minor systemic atrio-ventricular valve regurgitation. In echocardiography the structure in the left atrial appendage was seen, which could be a thrombus. The LMH was administered, which function is controlled by anty-Xa activity.

DISCUSSION

An congenitally corrected transposition of the great arteries (CCTGA), the heart twists abnormally during fetal development, and the ventricles are reversed. CCTGA is a rare heart defect. Only 0.5 to 1 percent of all people with heart defects have CCTGA. This means there are about 5,000 to 10,000 people in the United States with this condition.The anatomic left ventricle continues to act as the pulmonary pump and the anatomic right ventricle acts as the systemic pump.

Acording to ESC Guidelined the patient was classify to III WHO Class.

- For women in WHO class III, there is a high risk of complications, and frequent (monthly or bimonthly) cardiology and obstetric review during pregnancy is recommended.

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

- Significantly increased risk of maternal mortality or severe morbidity. Expert counselling required. If pregnancy is decided upon, intensive specialist cardiac and obstetric monitoring needed throughout pregnancy, childbirth, and the puerperium.
- Pre-pregnancy assessment including medical history, echocardiography, and exercise testing is indicated in all patients, with other diagnostic tests indicated on an individual patient basis.
- Functional status before pregnancy and history of previous cardiac events are of particular prognostic value
- An exercise test before pregnancy achieving ,70% of expected workload, showing a drop in arterial pressure or a drop in oxygen saturation may identify women at risk of developing symptoms or complications during pregnancy.
- In asymptomatic patients with moderate or good ventricular function, vaginal delivery is advised. If ventricular function deteriorates, an early caesarean delivery should be planned to avoid the development or worsening of heart failure.

EXPERT'S OPINION

Patient with this type of congenital heart disease appears to be a very difficult problem for both the cardiologists and obstetricians. Physiological increase of blood volume and heart rate cause heart overload during pregnancy. Patient presents good systolic function of left ventricle, however the supraventricular arrhythmias occur. The structure in the left atrial appendage seen in echocardiography might be a thrombus. When the atrial appendage diameter is so increased, the thrombus isn't very unusual finding. The well-being of mother and child should be our primal care, labor shouldn't be accelerated. Premature childbirth cause many complications for premature newborn. Caesarean section needs to be prolonged in time. Patient needs special attention, especially hypervolemia is a threat. Close follow-up and regular assessment of her clinical status including BNP changes monitoring is recommended. Low-molecular-weight heparin (LMWH) should be given.

CONCLUSION

The patient should remain in close cardiological and gynecological follow-up. LMWH treatment is advised.

REFERENCES

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e-mail: rarediseases@szpitaljp2.krakow.pl
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