Medical Expertise*

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

EXPERT: Prof. Krzysztof Rytlewski, gynecologist

Affiliation: *Department of Obstetrics and Perinatology, Jagiellonian University Medical College, Krakow, Poland*

CASE SUMMARY

At the age of 10 congenitally corrected transposition of great arteries was diagnosed. The case was discussed at the 26 week of her first pregnancy. She was in the good general condition, assessed as NYHA II. In this case an during cardiac Echo examinations thrombus in left atrium was found and anticoagulant treatment has been implemented (Enoxaparinum 2x80 mg), and because of ventricular and supraventricular extra systoles (during Holter monitoring) started adequate treatment (Metoprolol 2x50 mg). She was admitted to the Ob-Gyn Unit (Department of Obstetrics and Perinatology, Jagiellonian University Medical College, Krakow, Poland) at the 28 week of pregnancy due to steroid therapy to prepare the child for possible early labor. Studies have shown normal intrauterine development of the child.
An increase in blood volume, which is the basis of adaptive changes in pregnancy leads to an increase in the load of the right ventricle, which is exactly the system chamber.
According to guidelines - 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.
Systemic right ventricle WHO Class III: 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. 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. 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.
Delivery:
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.

LITERATURE REVIEW

Connolly HM, Grogan M, Warnes CA. Pregnancy among women with congenitally corrected transposition of great arteries. J Am Coll Cardiol. 1999;33:1692–5.
Therrien J, Barnes I, Somerville J. Outcome of pregnancy in patients with congenitally corrected transposition of the great arteries. Am J Cardiol. 1999;84:820–4.
Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease. Circulation. 2008;118:e714-e833; originally published online November 7, 2008;
Please fill in the literature review

EXPERT'S OPINION

Satisfactory condition of the pregnant allows for a continuation pregnancy to term of natural fetal maturity (over 39.0 weeks of gestation) or earlier if there will be a spontaneous birth contractions of the uterus. But because of the additional lesions (abnormal heart rhythm and blood clot in the right atrium) should be seriously considered a solution cesarean section during of pregnancy - as above. By the occurrence of the features of intrauterine fetal distress - treatment as in usual obstetric cases. In the case of worsening of the mother's circulatory - a immediately solution should be considered.
Advisable controls at the clinic of Pathology of pregnancy every 2 weeks to assess of intrauterine fetal condition.
In any case, congenital heart disease is indicated echocardiography of the fetal heart in the 18 -22 weeks of pregnancy.
Please fill in your opinion

CONCLUSION

The increase of the blood volume , which is a fundamental change adaptation in pregnancy leads to an increase in the workload of the right ventricle , which is actually a chamber system . Therefore, before making a decision about the pregnancy should be comprehensive clinical , ECG, chest X-ray and to assess the hemodynamic efficiency , state of the valves and ejection fraction .
Pregnancy in these cases gives a chance for live birth in 60-80 % of cases. In these cases , there was no congenital heart defects in children. One patient experienced heart failure in the third trimester .
Overall, left ventricular ejection fraction (LVEF) less than 40 % combined even with a small aortic

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

valve insufficiency of the systemic compartment (SAVV) gives a very small chance of success in pregnancy.
With the correct operation of the systemic chamber (right) and good clinical condition of the patients (NYHA I and II) it is possible to conduct a vaginal delivery, but because of the hemodynamic changes associated with childbirth should seriously be considered the solution by cesarean section. With the existence of exponents of deterioration of the systemic (right) chamber - solution cesarean section.

Please fill in the conclusion

REFERENCES

Connolly HM, Grogan M, Warnes CA. Pregnancy among women with congenitally corrected transposition of great arteries. J Am Coll Cardiol. 1999;33:1692–5.
Therrien J, Barnes I, Somerville J. Outcome of pregnancy in patients with congenitally corrected transposition of the great arteries. Am J Cardiol. 1999;84:820–4.
Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease. Circulation. 2008;118:e714-e833; originally published online November 7, 2008;

Please fill in the references

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