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

This is the case of 27 year-old patient with tetralogy of Fallot after palliative procedure in early childhood (systemic – pulmonary shunt or modified Blalock – Taussig anastomosis). He presented with symptoms of fatigue, resting and exercise chest burning, dizziness and palpitations. He is cyanotic and had episode of syncope during exercise. Hematological tests showed increased hematocrit and hemoglobin concentration. The basic biochemical profile was within normal limits. In the CPX study the exercise restriction was shown. Echocardiography revealed a typical features of tetralogy of Fallot and patent shunt between the left subclavian and left pulmonary artery. The restrictive physiology of the right ventricle and anatomical details were confirmed by CT scans and MRI study. After all clinical tests patient was discharged from the hospital at his own request with relevant medication. He is a heavy smoker and alcohol abused with very poor compliance with the team.

DISCUSSION

Life expectancy for unrepaired TOF is not encouraging. More than 95% are dead by the fourth decade, whereas after successful repair in childhood, 30-year survival has exceeded 90%. Palliated patients may develop pulmonary hypertension as a result of the arterial shunt procedure. For patients with palliative shunts the pressure overload of the RV and often volume overload of the LV as well as distortion of the pulmonary arteries may lead to biventricular dysfunction and failure, exercise intolerance, and arrhythmias. Some of these symptoms are already present in this particular patient (arrhythmias, exercise intolerance, desaturation). Additionally there are some symptoms suggesting myocardial ischemia both because of cyanosis or development of coronary artery disease. The important accessory risk factors for myocardial dysfunction in this patient are smoking and alcohol dependency. This

patient seems to be a good candidate for total correction of the defect but for above discussed reasons the assessment of PA pressure and coronary circulation should be undertaken before surgery. Although there have been done imaging studies (angio-CT, MRI and echocardiography) that did not reveal coronary artery origin and course anomalies but careful consideration should be given also to coronary angiography to rule out acquired coronary artery disease. Unfortunately there is evidence that late repair of tetralogy of Fallot may not necessarily decrease the risk of ventricular and supraventricular arrhythmias but should stop devastating changes in cardiovascular system and improve quality of life of the patient ([1]). Without definitive repair, the average life expectancy in patients with tetralogy of Fallot is 12 years, and fewer than 5% survive to their 40s. Older survival is exceptional. In the few reported cases of long survival without repair, there have been hypoplastic pulmonary arteries, ventricular hypertrophy and extra-cardiac shunts (previous palliative anastomoses, patent arterial duct or MAPCAs ([2]).

Early surgical outcomes after total correction in adults have improved significantly during the last decade and recent series report perioperative mortality rates of 16% ([3]) to 3% ([4]). Nevertheless, the patient must be informed that there are late complications after repair ([5]) such as arrhythmia, heart failure, risk of sudden death, and that close, long-term follow-up and perhaps additional therapies will be necessary ([6], [7], [8]).

EXPERT'S OPINION

The patient is good candidate for repair and there are indications for such surgery but there are some factors that should be considered beforehand. Before surgery cardiac catheterization should be undertaken and careful assessment of both pulmonary and coronary circulation should be carried out. The patient has to fully understand the risks and benefits of the operation. Also factors that may affect the patient's decision (inability to comprehend the situation, alcohol abuse, nicotine abuse) should be considered. He must be informed that there are late complications after repair such as arrhythmia, heart failure, risk of sudden death, and that close, long-term follow-up and perhaps additional therapy will be necessary.

CONCLUSION

The total correction procedure is feasible and indicated after informed conset of the patient. Before surgery cardiac catheterization should be undertaken and careful assessment of both pulmonary and coronary circulation should be done. Risk related to long-term period after surgery (arrhythmia, heart failure, risk of sudden death) should be minimized by close follow-up and accessory therapies.

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