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

There was a 23-year-old male patient with a history of congenital heart disease and cirrhosis of the liver, epilepsy, HCV infection, right hemiparesis, ascites, hypoalbuminaemia, protein – losing enteropathy and anaemia microcytic At birth he was diagnosed with transposition of great arteries, hypoplasia of right ventricle, left atrioventricular valve atresia and ventricular septal defect. In the years 1989 – 1998 he underwent three stages of sugical corection: banding of pulmonary artery and atrium septostomy, hemiFontan operation and Fontan operation

In 2013 he was hospitalized due to progression of heart failure to class NYHA III and presents symptoms of easy fatigue and intolerance of physical exercise. Physical examination was significant for mild cardiac insufficiency with abdominal swelling and peripheral oedema Echocardiography revealed ejection fraction of systemic ventricle about 45-50% and mild atrioventricular valve regurgitation with 1,9/1,2 mmHg gradient through atrioventricular valve. Also the echocardiography showed extras echo in the tunnel – suspicious of thrombus and liquid in cavitas pleuralis 20-30 mm.

The ECG showed sinus rhythm with heart rate 90 beats per minute, atrioventricular block I' (PQ 220ms). In addition, episodes of nodal rhythm 60 bpm with advanced II block were noted.

Holter monitoring was performed, showing sinus rhythm with AV block I – PQ 240 ms, episodes of nodal rhythm QRS – 100 ms and advanced II block - type Mobitz and II block type 2:1

The patient was treated with enoxaparine 40 mg, furosemide spironolacotne 50 mg, kalium, prednisone 2,5 mg walproinian acid 500, wigabatrine 500 mg, ferrum, phospholipids. After hepatology consultation to treatment during hospitalization was added: albumins furosemide 20 mg i.v. and aldactone 200 mg i.v. He did not cooperated during hospitalization. After heparin treatment reduction of additional echo in the tunnel was observed.

DISCUSSION

Recent progress in cardiac surgery and pediatric cardiology has resulted in large numbers of adult patients who have surgically corrected complex congenital heart defects. The Fontan operation has become the treatment of choice in patients with the univentricular heart (single ventricle - SV); the anomaly accounts for about 8% of all congenital heart defects. Despite the improvements in surgical techniques that reduce perioperative mortality, late deterioration in functional status can be observed with longer duration of follow-up (1). The most common late complication are: arrhythmia, thrombosis, cyanosis, heart failure and protein losing entheropathy (1, 2). The incidence of exudative enteropathy ranges from 3% to 15% in patients after the Fontan procedure (3). The symptoms related to protein deficiency (fluid retention, thromboembolic complications, immunosuppression, hypocalcemia) are associated with a poor prognosis: 30% of patients die within 2 years, and 50% within 5 years after the diagnosis (5,6). Causes of exudative enteropathy are not clear. The complication is believed to result from increased systemic venous pressure, intestinal ischemia, enterocyte heparan sulfate deficiency and inflammatory factors.

EXPERT'S OPINION

There are no effective treatments modalities, either (4). Heparin and/or steroids are not always of benefit, and they cause numerous side effects. High protein diet, diuretics, iron deficiency supplementation are recommended. Exercise training and psychological support also should be considered.

CONCLUSION

Patient should be qualify for conservative treatment in centre specialized in adults with congenital heart diseases.

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