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Potts' shunt in a child with idiopathic pulmonary arterial hypertension – one-and-a-half year observation

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Abstract

This paper presents the case of a young girl with idiopathic pulmonary hypertension, who developed signs of severe heart failure within a short period of time. Pharmacotherapy with sildenafil and bosentan (among other drugs) was ineffective. Heart catheterization revealed suprasystemic pressure in the pulmonary artery. At the age of 7.5 years, the patient underwent a surgical Potts shunt (namely, a direct side-by-side anastomosis from the left pulmonary artery to the descending aorta). The procedure resulted in a significant improvement of the clinical, echocardiographic, and biochemical parameters, which persists after one and a half years of follow-up. After the surgery, pharmacotherapy with bosentan was gradually discontinued.

Key words: idiopathic pulmonary hypertension, Potts' procedure.

Streszczenie

W pracy opisano przypadek dziewczynki, u której w krótkim czasie rozwinął się obraz ciężkiej postępującej niewydolności krążenia spowodowanej pierwotnym nadciśnieniem płucnym. Farmakoterapia, m.in. sildenafilem i bozentanem, była nieefektywna. W przeprowadzonym cewnikowaniu serca stwierdzono suprasystemowe ciśnienie w tętnicy płucnej. Gdy dziecko miało 7,5 roku, wykonano chirurgiczne zespolenie Pottsa (połączenie pomiędzy lewą tętnicą płucną a aortą zstępującą), uzyskując poprawę parametrów klinicznych, echokardiograficznych oraz biochemicznych, która utrzymuje się półtora roku po zabiegu. Po zabiegu stopniowo wycofano się z farmakoterapii bozentanem.

Słowa kluczowe: pierwotne nadciśnienie płucne, operacja Pottsa.

Introduction

Idiopathic pulmonary hypertension (IPH) is a very rare disease with poor prognosis, especially in children. Its effects include the development of suprasystemic pressure in the pulmonary artery (PA), a very significant dilatation of the right ventricle (RV), as well as a number of consequent physiopathological changes, including RV compression on the left ventricle (LV), causing recurrent spells of unconsciousness and sudden death. According to one American register, the time of survival from the moment of diagnosis in children with IPH was 10 months and in adults 2.8 years [1]. The prevalence of IPH has been estimated at approximately 2 cases per million [2]. Recently, a number of new agents have been introduced to IPH therapy, but they offer only short-term improvement [3].

On the other hand, it has been known for a long time that the prognosis for Eisenmenger's syndrome caused by pulmonary hypertension with right-to-left shunt (e.g., in non-operated heart defects such as patent ductus arteriosus or ventricular septal defect) is much better, and the time of survival is estimated at 40-60 years [4]. In 2004, Blanc *et al.* proposed a novel strategy for the treatment of IPH in children by performing the long-known Potts shunt (i.e., anastomosing the left pulmonary artery with the descending aorta) [5]. There are few publications on the subject [5, 6]. The present report describes the case of a child with severe IPH, in whom a successful Potts shunt was performed; the good outcome of the surgery was confirmed by 1.5 years of follow-up.

Case study

The girl (G1, T1, natural birth in the 34th week of gestation, birth weight 2690 g, Apgar score 8/9) was admitted to the clinic at the age of 6 years due to quickly increasing exercise intolerance, chest pain, and a single episode of loss of consciousness; her medical history included frequent re-

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spiratory tract infections before the age of 3 years, which later subsided. The child could not participate in school activities and was recommended to take part in an individual education program. Initially, based on non-invasive examinations, the cardiac clinic suspected primary pulmonary hypertension; the patient was administered sildenafil, l-arginine, and captopril (the parents did not consent to cardiac catheterization at the time). Physical examination performed at admission to the clinic established the patient's weight as 22 kg (35^{th}-65^{th} percentile) and height as 120 cm (35^{th}-65^{th} percentile). The child was in a severe general condition (she exhibited fatigue even during light exercises – NYHA III/IV), showing signs of heart failure, clear dyspnea, as well as tachypnea (36 breaths per minute) and tachycardia (130-140 bpm). O_2 saturation was 94%. The result of the patient's 6-minute walk test (6MWT) was 215 m (Borg scale result – 5). Moreover, a systolic murmur (1-2/6 at the base of the 2nd left intercostal space) and a diastolic murmur (1/4 in the 4^{th} left intercostal space) were present. The liver was enlarged by 3 cm. Electrocardiography and echocardiography revealed: enlargement of the right atrium and ventricle, presence of fluid in the pericardial sac, signs of right ventricular compression on the left ventricle, and signs of pulmonary hypertension (PulmAT increased to 48 ms). Doppler echocardiography also demonstrated significant pulmonary regurgitation (+++) and tricuspid regurgitation (++/+++). Posterior-anterior chest X-ray examination revealed wide, vascular hila and significantly enlarged cardiac silhouette with full waist; the cardiothoracic ratio was 0.71. Ventilation-perfusion scintigraphy excluded pulmonary embolization, and NMR excluded left ventricular noncompaction and arrhythmogenic right ventricular dysplasia (ARVD), revealing great enlargement of the right ventricle, significant widening of the trunk of the pulmonary artery and its two branches, as well as displacement of the left ventricle compressed by the right ventricle. Cardiac catheterization revealed suprasystemic pressure in the pulmonary artery (invasive measurement of pulmonary artery pressure: 125/70/91 mmHg, noninvasive cuff measurement of aortic pressure: 81/47/64 mmHg). The attempt to place the catheter in the peripheral branch of the pulmonary artery ended in a pulmonary hypertensive crisis with bradycardia and a drop of O_2 saturation to 30%. The circulation was only stabilized after a period of continuous intravenous infusion of amrinone. The serum concentration of NT-proBNP during this time was 13,433 pg/ml. Bosentan was added to the therapy at the initial dose of 2 × 31 mg (later increased to 2 × 62 mg). Despite the employed treatment, the patient's condition continued to deteriorate. On December 12, 2012, during a cardiac surgery consultation with Prof. M. Zembala, the child was qualified for a Potts procedure. Several days later, the patient suffered from a severe respiratory tract infection which was managed with antibiotic therapy. On January 1, 2013, the planned surgery was performed at the Clinic of Pediatric Cardiac Surgery (headed by Prof. J. Skalski) of the University Children's Hospital in Kraków. Pre- and post-operatively, the patient received intravenous infusion of Flolan (prostaglandin meant to reduce the pressure in the pulmonary artery). The postoperative course was complicated by an incident of circulatory insufficiency with the presence of fluid in the left pleural cavity, requiring a period of underwater drainage. At two weeks after the procedure, the child was referred from Krakow to our center. Oxygen saturation measured with pulse oximetry was 78% (lower extremities) and 96% (upper extremities). The patient still suffers from thrombocytopenia and remains under the supervision of an outpatient pediatric hematology clinic in Zabrze (Prof. Z Szczepański). At present (1.5 years after the procedure), the girl feels well (NYHA class I/II); she has returned to school; her liver is no longer palpable under the costal margin. No further episodes of syncope have been observed. According to her parents, the girl's exercise tolerance has increased; her height has increased by 8 cm and her weight by 5 kg (body mass = 27 kg, 35^{th}-65^{th} percentile; height = 128 cm, 15^{th} percentile). In comparison to preoperative examinations, the patient's echocardiogram indicates improved proportions of the ventricular cavities (LV dimensions in the lower range of normal; the right ventricle is smaller, but still larger than LV in apical 4-chamber view) as well as reduced tricuspid regurgitation (+/++) and pulmonary regurgitation (++) without fluid in the pericardial sac. The diameter of the Potts shunt was established at 6 mm by the latest echocardiographic examination; the maximal gradient through the anastomosis was established at 14 mmHg. The new result of 6MWT was 472 m, and the result of NT-proBNP was 1000 pg/ml. At approximately 1 year after the procedure, the treatment with bosentan was gradually discontinued; the patient currently receives only sildenafil and acenocoumarol.

Discussion

The treatment options for IPH are limited. Although pharmacotherapy with state-of-the-art pulmonary vasodilators – endothelin-1 receptor antagonists (bosentan), phosphodiesterase type 5 inhibitors (sildenafil), and intravenous prostacyclins (epoprostenol) constitutes an interesting treatment option resulting in improved prognosis [7], the long-term efficacy of these agents in the treatment of IPH is relatively low [3]. These observations are in accord with the case study presented above, as our patient's condition continued to deteriorate despite the intensification of pharmacological therapy (combined therapy with sildenafil and bosentan). Idiopathic pulmonary hypertension can also be treated with balloon atrioseptostomy and pulmonary transplantation, but both these methods are known to have significant limitations.

The idea to use Potts' shunt in the treatment of IPH [5] is based on the observations of the physiopathology of Eisenmenger's syndrome and patent ductus arteriosus. Right-to-left shunt between the left pulmonary artery and the descending aorta (as is the case with the Potts procedure) prevents the desaturation of the CNS and coronary circulation and the occurrence of paradoxical embolisms in the CNS. Concurrently, right ventricular afterload is im-

proved, and the suprasystemic pressure in the pulmonary artery becomes equal to that in the aorta. Left ventricular function is unburdened, and LV ejection fraction improves. Recently, Baruteau *et al.* [6] presented a series of 8 children with IPH from 6 cardiology centers in France (median age: 8 years) who were treated with Potts' procedure. All the children were in NYHA class IV, and 6 suffered from episodes of loss of consciousness. The mean time of postoperative follow-up was approximately 5 years. Two patients, in whom pharmacological treatment with state-of-the-art pulmonary vasodilators was discontinued in the early postoperative period, died on the 11th and 13th day after the surgery due to hypertensive crises (postoperative mortality: 25%). No long-term mortality was noted in this group. These observations indicate that the aforementioned agents should be discontinued gradually at a later time after the surgery, as was the case with our patient.

Potts' shunt can also be created using interventional catheterization [8]. Four patients were treated in this manner. A retrograde puncture of the left pulmonary artery from the descending aorta was made; after the creation of a veno-arterial loop, a covered stent was introduced into the puncture site. The procedure was successful in 3 patients, two of whom survived. One of the patients died during the procedure due to massive bleeding into the chest; another died shortly after the procedure due to multiple organ dysfunction syndrome. In view of these results, the application of this method appears limited and reserved for carefully selected cases.

In conclusion, palliative Potts' shunt constitutes a new and interesting therapeutic alternative to lung transplantation. The method offers a chance to prolong the life of children with idiopathic pulmonary hypertension and to improve its quality.

Dislocure

Authors report no conflict of interest.

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