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Pseudo-thrombosis of the superior vena cava as a manifestation of pulmonary hypertension crisis after Glenn procedure

A.A. Morozov, A.K. Latypov, A.V. Vasilets, A.A. Lyapunova, E.V. Grekhov

Federal North-West Medical Research Centre, St Petersburg, Russia

Corresponding author:
Alexandr A. Morozov, Federal North-West Medical Research Centre, 2 Akkuratov street, St Petersburg, Russia, 197341.
E-mail: morozov_1981@mail.ru

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Abstract

Objective. Pulmonary hypertension is one of the typical complications of the congenital heart defects with pulmonary overload circulation. Hemodynamic status of the pulmonary circulation is critically important for the patients with univentricular heart anatomy and determines opportunities and results of the surgical treatment. **Case report.** We present a case report of superior vena cava syndrome feigning a thrombosis of the anastomosis in a patient after Glenn procedure. Clear clinical picture, hemodynamic instability of the patient and inability to exclude anastomosis dysfunction were an indication for re-operation. There were no signs of anastomosis dysfunction or thrombosis. This clinical situation was assessed as a manifestation of pulmonary hypertension (PH) feigning a thrombosis of the Glenn anastomosis. Applying of pulmonary vasodilation agents gave an opportunity to avoid a PH crisis and promoted stable postoperative period. **Conclusions.** In consideration of our clinical experience we believe that inclusion of computer tomography scan or magnetic resonance imaging into examination protocol is appropriate for the patients with high risk of PH crisis.

Key words: pulmonary hypertension, Glenn anastomosis, superior vena cava syndrome

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Псевдотромбоз верхней полой вены как проявление криза легочной гипертензии после создания верхнего двунаправленного каво-пульмонального анастомоза

А. А. Морозов, А. К. Латыпов, А. В. Василец, А. А. Ляпунова, Е. В. Грехов

Федеральное государственное бюджетное учреждение «Северо-Западный федеральный медицинский исследовательский центр» Министерства здравоохранения Российской Федерации, Санкт-Петербург, Россия

Контактная информация:
Морозов Александр Александрович, ФГБУ «СЗФМИЦ» Минздрава России, ул. Аккуратова, д. 2, Санкт-Петербург, Россия, 197341.
E-mail: morozov_1981@mail.ru

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Резюме

Одним из типичных осложнений течения врожденных пороков сердца с гиперволемией малого круга кровообращения является формирование легочной гипертензии (ЛГ). Состояние гемодинамики малого круга кровообращения является критически важным для пациентов с единственным желудочком сердца и во многом определяет возможность проведения и результат хирургической коррекции порока. В настоящем сообщении описывается случай развития синдрома верхней полой вены у ребенка после операции Гленна с клиникой тромбоза верхнего двунаправленного каво-пульмонального анастомоза. Развернутая клиническая картина, гемодинамическая нестабильность пациента, а также невозможность исключения дисфункции анастомоза потребовали повторного оперативного вмешательства. В результате проведения интраоперационной ревизии признаков тромбоза, дисфункции анастомоза не выявлено. Данная клиническая ситуация была расценена как кризовое течение ЛГ, симулирующее клинику тромбоза верхнего двунаправленного каво-пульмонального анастомоза. Использование легочных вазодилататоров в послеоперационном периоде позволило избежать рецидива кризов ЛГ и способствовало стабильному течению послеоперационного периода. С учетом данного клинического опыта включение мультиспиральной компьютерной томографии с контрастированием или магнитно-резонансной томографии в сосудистом режиме в протокол обследования является целесообразным для пациентов, входящих в группу риска по развитию ЛГ при возникновении подобных осложнений.

Ключевые слова: легочная гипертензия, анастомоз Гленна, синдром верхней полой вены

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Introduction

Pulmonary hypertension (PH) is a common complication of several congenital heart diseases (CHD) with excessive pulmonary blood flow. They are divided into simple CHD with left-to-right shunt (septum defects, patent ductus arteriosus and others) and complex (complete atrioventricular canal, common arterial trunk and others) [1]. Pulmonary hemodynamics is particularly important for patients with an univentricular heart (UVH). Due

to the anatomical features radical intervention in these patients is impossible, so the hemodynamic defect correction is the main treatment option. This surgical approach includes several stages, such as the application of top bidirectional cavopulmonary anastomosis (BCPA, Glenn procedure) [2, 3], followed by Fontaine operation (Fontan procedure) [4, 5]. Diagnostic criteria for applicability of these techniques include a sufficient degree of development and no deformation of the pulmonary arteries, as well as the pulmonary artery pressure level and pulmonary vascular resistance. Thus, the pulmonary circulation characteristics are crucial for the subsequent surgical treatment. Among patients with UVH there is a subgroup of patients with nonobstructive pulmonary blood flow. In these patients the first step is limiting of the pulmonary blood flow to prevent pulmonary hypertension development, and this is achieved by pulmonary artery banding (PAB) [6]. Later in patients fulfilling diagnostic criteria, the above mentioned steps of hemodynamic correction described =are performed.

We present a case report of BCPA in a patient with UVH, complicated by the development of the superior vena cava (SVC) thrombosis.

The clinical case description

A 4-month girl A. was admitted to hospital with double inlet single ventricle with the correct location of the main vessels, and secondary atrial septal defect (ASD). The child underwent pulmonary artery band at the age of 1.5 months. At admission acrocyanosis despite high saturation (95 %) according to pulse oximetry data was noted, suggesting an excessive pulmonary blood flow. According to transthoracic echocardiography the following defects were confirmed: a double inlet single ventricle with atrioventricular valve failure up to I degree, satisfying single ventricle contractility without local dyskinesia, small diameter atrial septal defect, the pressure gradient on the band about 85–90 mm Hg. Multislice computed tomography (MSCT) showed a normal inflowing of cava vena -and pulmonary veins, and an absence of patent ductus arteriosus (PDA). Angiocardiography and a direct manometry were conducted in order to define further treatment approach. It showed no signs of stenosis or deformation of the pulmonary artery branches, the pulmonary artery pressure distal to band was 2/11 (average — 14), proximal — 85/5 (average — 39), systolic pressure gradient — 64 mm Hg, pulmonary vascular resistance — 2 Wood units, calculated pulmonary arterial index — 198.2 mm/m2.

Thus, it was decided to perform the next stage of the surgical correction. The upper bidirectional cavo-pulmonary anastomosis was created, pulmonary trunk was ligated, and an open atrioseptostomy was performed. According to intraoperative monitoring the pressure in cavo-pulmonary tract was 11 mm Hg, and saturation was 86–89 %. During the first days after surgery there was a progressive decline of oxygen saturation to 60 %, it was dependent on blood pressure and required prolongation of mechanical ventilation with high inspiration oxygen, nitric oxide administration, sedation, and paralyzation of patient with a positive effect. Transthoracic echocardiography showed neither any signs of thrombosis, nor cavo-pulmonary anastomosis dysfunction. Based on the clinical data the insufficiency of the blood flow volume in anastomosis was suggested; it was decided to create an additional source for pulmonary blood flow. During repeated surgery the ligature from pulmonary trunk was removed and it was opened. Blood flow in the pulmonary trunk was limited by band, oxygen saturation was 85–87 %, and the pressure in the cavo-pulmonary tract was 14–15 mm Hg. Seven hours after the reoperation, the child was extubated, stable hemodynamics without inotropic support and saturation about 90–92 % were maintained.

Repeated echocardiography showed laminar flow of cavo-pulmonary anastomosis with the speed of 0.6 m/s, the narrowing of the pulmonary trunk to 3 mm, and the pressure gradient on the band (peak/average) of 116/70 mm Hg.

The early postoperative period was stable, there was a moderate swelling of the upper body with positive dynamics. However, on the 5th day after the initial intervention there was an acute desaturation to 62 % with tachypnea to 65 per minute. At physical examination breathing was auscultated on both sides above all lung fields. There was sinus rhythm with the heart rate about 120–130 per minute, blood pressure was 79/38 mm Hg with a tendency to hypertension. According to echocardiography

there was no blood flow in the SVC and in the anastomosis with right pulmonary artery (PA), and a systolic blood flow in pulmonary trunk and PA bifurcation was visualized. In addition, there was a clear cyanosis of the upper body at physical examination. Therefore, considering the obvious clinical picture and hemodynamic instability acute thrombosis of SVC was suspected, and an urgent anastomosis without any additional diagnostic procedures revision was performed. Under extracorporeal circulation anterior semicircle of cavo-pulmonary anastomosis was opened, the blood flow in SVC was visualized, there were no signs of thrombosis either of SVC or of pulmonary arteries. An anterior anastomosis semicircular grafting by xenopericardium was performed to prevent stenosis of the cavo-pulmonary anastomosis, and the surgery was completed as usual. Taking into account clinical and instrumental data, intraoperative revision results (there were no signs of thrombosis), the case was considered as a manifestation of PH feigning a thrombosis of the Glenn anastomosis including echocardiography. In this regard, NO donator (nitric oxide) was added to therapy, after extubation sildenafil was prescribed. Later, postoperative period was stable, and oxygen saturation maintained above 80%.

Conclusion

PH is one of the severe complications of CHD with excessive pulmonary blood flow, and PH crises are one of the causes of unfavorable outcomes in the postoperative period. An abrupt pressure increase in the PA is accompanied by a saturation drop, cyanosis, hypotension, and other clinical manifestations. However, after the creation of upper bidirectional cavo-pulmonary anastomosis PH crisis may manifest as SVC syndrome. This fact should be taken into account in the differential diagnosis. A MSCT scan with contrast or magnetic resonance imaging with angiography are appropriate in patients at high risk of PH development in case of such complications.

Conflict of interest

Authors declare no conflict of interest.

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Author information

Alexandr A. Morozov, MD, Pediatric Heart Surgeon, Department of Pediatric Cardiac Surgery, Federal North-West Medical Research Centre;

Alexandr K. Latypov, MD, PhD, Head, Department of Pediatric Cardiac Surgery, Federal North-West Medical Research Centre;

Anastasiya V. Vasilets, MD, Pediatric Heart Surgeon, Department of Pediatric Cardiac Surgery, Federal North-West Medical Research Centre;

Anna A. Lyapunova, MD, Pediatric Cardiologist, Department of Pediatric Cardiac Surgery, Federal North-West Medical Research Centre;

Evgeny V. Grekhov, MD, PhD, Leading Researcher, Head, Research Group of Pediatric Cardiac Surgery, Federal North-West Medical Research Centre.