

Treatment of elevated pulmonary artery pressure in a child after Glenn procedure: transcatheter closure of pulmonary artery banding with subsequent sildenafil therapy

Leczenie podwyższonego ciśnienia płucnego u dziecka po operacji Glenna: przezcewnikowe zamknięcie miejsca bandingu tętnicy płucnej, a następnie zastosowanie terapii sildenafilem

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Abstract

An additional source of pulmonary blood flow in a patient with bidirectional Glenn procedure (BGD) may cause elevation of mean pulmonary artery pressure (MPAP), precluding safe completion of the Fontan operation. We present a case of single ventricle physiology after pulmonary artery banding (PAB) and Glenn procedure. At the age of six years, cardiac catheterisation revealed in the patient elevated MPAP (22 mm Hg). The PAB was closed through the right internal jugular vein with an Amplatzer Atrial Septal Occluder. After the procedure, MPAP remained at a similar level. Sildenafil oral therapy was applied for six months. Subsequent heart catheterisation confirmed complete closure of PAB and decrease of MPAP to 10 mm Hg. The abovementioned complex treatment of elevated MPAP pressure in a child after Glenn therapy allowed safe completion of the Fontan operation.

Key words: single ventricle heart, pulmonary artery banding, transcatheter closure, sildenafil

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INTRODUCTION

Children with single ventricle (SV) physiology often undergo staged palliative surgical procedures to regulate their systemic and pulmonary blood flow. This includes pulmonary artery banding (PAB), connection of superior vena cava with pulmonary artery (bidirectional cavopulmonary Glenn anastomosis — BDG) and others. The final palliation in a case of SV is the Fontan operation (inclusion of inferior vena cava to pulmonary arteries). The key to a successful Fontan type operation is low pulmonary artery pressure (PAP) and low pulmonary resistance. According to the classical indications for the Fontan operation, mean PAP (MPAP) should be below

15 mm Hg. Recent publications have presented effective treatment of elevated PAP in congenital heart diseases [1], also after a Fontan operation [2] with oral sildenafil therapy.

We report our experience in a child with elevated MPAP after a Glenn procedure with additional systemic to pulmonary flow (through the PAB), treated successfully with transcatheter closure of this communication, and subsequent sildenafil therapy.

CASE REPORT

The patient was born with mitral atresia and hypoplastic left ventricle. He had SV with right ventricle morphology. Mo-

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reover, L-malposition of the great vessels and restrictive atrial septum defect were diagnosed. At the age of two months, PAB and Blalock-Hanlon operation (surgical atrioseptectomy) were performed. At the age of one year, the patient underwent BDG. The postoperative course and clinical follow-up were unremarkable. At the age of six years, cardiac catheterisation was performed to assess suitability for a Fontan operation. Right femoral artery and right internal jugular and left femoral veins were cannulated. Ventriculography demonstrated open accessory flow from SV through pulmonary banding (Fig. 1A). The MPAP was 22 mm Hg, with a Qp/Qs 3:1. Saturation of arterial blood was 77%. A decision was taken for transcatheter closure of PAB. A 4 F Judkins left catheter and a 0.035 × 260 cm guidewire were inserted from the jugular vein through the pulmonary banding to the SV. Thereafter, a 4 F vascular sheath was exchanged for a 6 F delivery system (AGA Medical Comp) and a 5 mm Amplatzer Atrial Septal Occluder (ASO) (AGA Medical Comp) was opened in distal and proximal parts of the banding. Pulmonary arteriography showed well developed pulmonary branches, and good position of the device closing PAB (Fig. 1B). Postprocedural MPAP was 22 mm Hg, and fluoroscopy time was 21 min. In further hospital observation, thrombus formation was detected in the pulmonary trunk below the closed banding. This was confirmed by angio-computed tomography. Treatment with warfarin was introduced. Taking into consideration the elevated MPAP observed after PAB closure, sildenafil therapy at 1 mg/kg/day (in two doses) was started. The child was discharged home after six weeks of hospitalisation. After six months, another cardiac catheterisation was performed, and good position of

the device and complete closure of banding were confirmed. The patient's MPAP decreased to 10 mm Hg, pulmonary arteries were of good diameter (left 10.4 mm, right 11.2 mm and McGoon index: 2.5). His arterial blood saturation was 65–70%. He was qualified and scheduled for completion of Fontan circulation.

DISCUSSION

Pulmonary artery banding is a palliative surgical procedure used to treat functionally single ventricular hearts without pulmonary artery stenosis. The PAB remains the preferred palliation to delay definitive repair until age and body weight are suitable for available techniques. The primary goal of performing PAB is to reduce excessive pulmonary blood flow and protect the pulmonary vasculature from hypertrophy and irreversible pulmonary hypertension [3, 4]. In the setting of single ventricle physiology, control of pulmonary blood flow is important in order to avoid congestive heart failure and to prevent development of pulmonary vascular disease that might preclude future surgery. In order to minimise the degree of cyanosis, patients are occasionally left with accessory antegrade flow from the ventricle into the pulmonary arteries at the time of BDG, as was the case here.

A few cases of transcatheter closure of accessory ventriculopulmonary connections [5, 6] have been reported in the literature. For such purposes, Amplatzer Duct Occluders have mainly been used, and, occasionally, vascular plugs and ASO. We decided to use an ASO device taking into consideration the retrograde method of implantation. Unfortunately, unlike the case described by Ebeid et al. [5], we observed no decrease of MPAP immediately after the procedure.

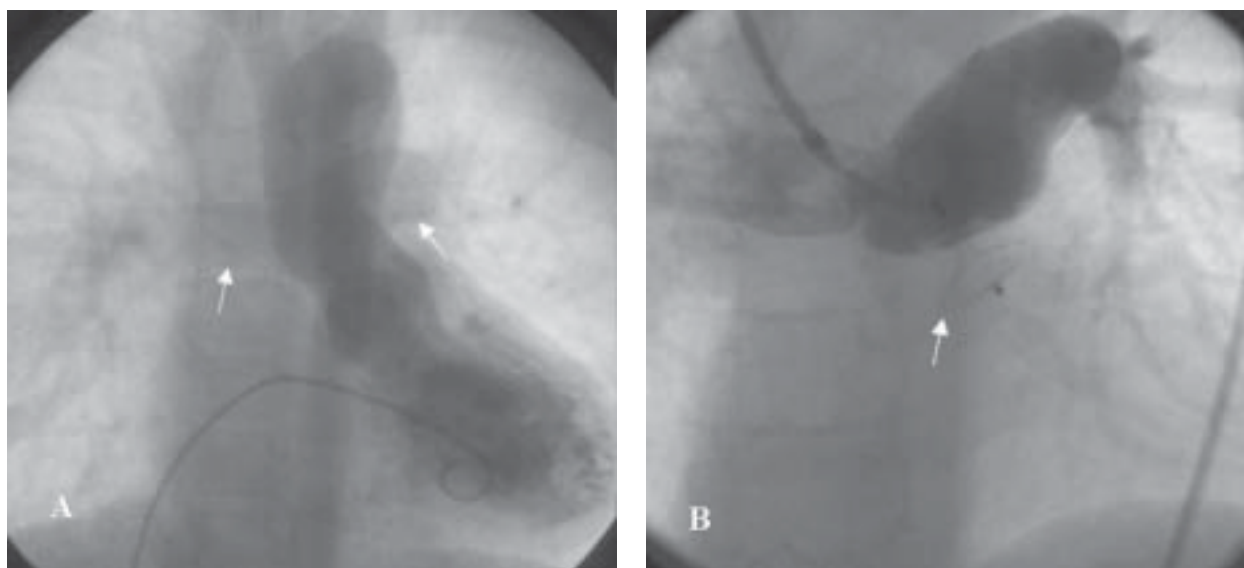


Figure 1. A. Angiography in single ventricle. Simultaneous appearance of aorta and pulmonary arteries (white arrows); B. Angiography of pulmonary arteries (access from right jugular vein). Amplatzer Atrial Septal Occluder (white arrow) is closing the pulmonary artery banding

According to our previous experience in treating a patient with failing Fontan with sildenafil [2], we decided to introduce this drug to the therapy. The administration of pulmonary vasodilators such as sildenafil has been shown to reduce elevated PAP. It can decrease pulmonary vascular resistance and MPAP, as well as improve cardiac output and cardiac index [1, 7, 8]. These observations confirmed the results achieved in our patient.

CONCLUSIONS

Elevated PAP in a child after Glenn shunt, as well as the presence of accessory ventriculopulmonary connection (persistent PAB) can be effectively treated with therapy combining percutaneous closure of PAB with subsequent sildenafil treatment.

Conflict of interest: none declared

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