

Proximal ductus arteriosus stenosis after the hybrid stage I procedure in a newborn with hypoplastic left heart syndrome

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Abstract

We present a case of a severely ill newborn with hypoplastic left heart syndrome after the hybrid stage I procedure, with right systemic ventricle insufficiency due to proximal ductus arteriosus stenosis. The child was successfully treated with percutaneous second PDA stent implantation.

Key words: hypoplastic left heart syndrome, hybrid procedures in HLHS, stage I of HLHS treatment, newborn.

Introduction

The first stage of palliative hybrid treatment of children with congenital heart disease (HLHS hybrid stage I) in the form of hypoplastic left heart syndrome (HLHS) is an alternative to the Norwood procedure [1, 2]. It includes right and left pulmonary artery (RPA and LPA) banding with concomitant persistent arterial duct (PDA) stenting. One of the complications of this form of treatment in children is PDA stenosis [3].

We present a case of a female patient with HLHS after the first stage of hybrid treatment, who presented with deterioration of the systemic right ventricle contractility caused by stenosis in the proximal part of the PDA, which was not covered with the stent.

Case report

A 2-day neonate with prenatally diagnosed congenital heart disease in the form of hypoplastic left heart syndrome underwent RPA and LPA banding and stent implantation to the PDA as the first stage of the hybrid treatment (HLHS hybrid stage I). Alprostadil (Prostin®) was discontinued. This was followed by balloon atrioseptostomy. The child was extubated on the second day after the procedure without any complications. A control echocardiographic examination (ECHO) performed routinely after the

procedure confirmed the presence of normal right ventricular function, normal tricuspid and pulmonary valve function, free flow through the stent into the descending aorta with an effective backward flow to the ascending aorta and broad inter-atrial communication. From the 7th day after the procedure ECHO studies showed progressive deterioration of the right ventricular function with increasing tricuspid regurgitation and symptoms of stenosis in the proximal part of the PDA, which was confirmed by the diagnostic catheterization (Figure 1). For that reason a second stent was immediately implanted to the stenosed proximal PDA segment using a telescopic technique (Figure 2). Free flow from the pulmonary artery to the aorta was achieved (Figure 3). Control examinations after the procedure demonstrated normalization of the right ventricular and tricuspid valve function, which permitted the child to be discharged from the hospital on the 3rd day after the intervention. A second stage of treatment was performed in the 5th month of the child's life (HLHS comprehensive hybrid stage II, Figure 4). Currently the patient is under ambulatory follow-up and remains in good clinical condition with an observation period of 8 months.

Discussion

Irrespectively of the treatment modality used ("Norwood" type operation or hybrid treatment) the result of

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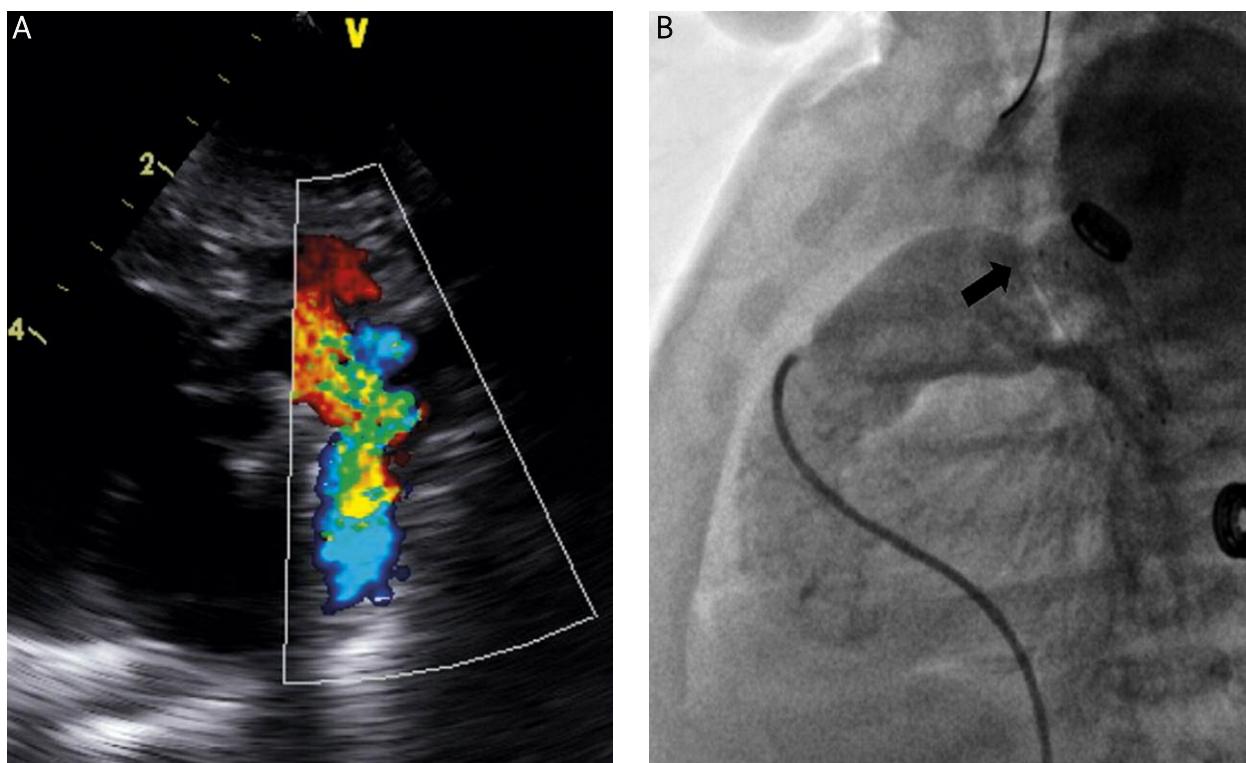


Fig. 1. Proximal ductus arteriosus stenosis after the hybrid stage I procedure in a newborn with hypoplastic left heart syndrome. **A** – Echocardiography – restrictive blood flow through ductus arteriosus. **B** – Ventriculography – proximal stenosis of unstenated part of the ductus (arrow)

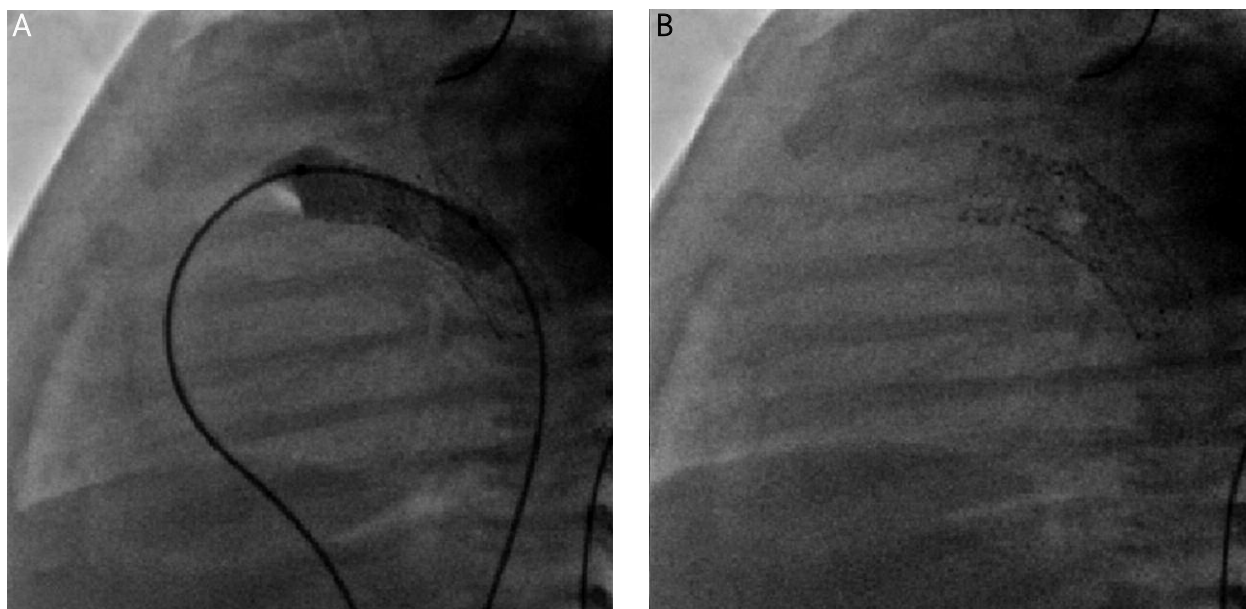


Fig. 2. Cardiac catheterization – lateral view. **A** – Second stent implantation into ductus arteriosus. **B** – Post-intervention view

treatment of neonates with HLHS depends on the right ventricular function and on optimal blood distribution to the systemic and pulmonary circulation [4]. Any deterioration of the ventricular function may be catastrophic and should be immediately diagnosed and, if possible,

a causative treatment should be performed. Prompt diagnosis and appropriate treatment in the presented case led to normalization of the systemic flow and improvement of the right ventricular function. This led to stabilization of the child's general condition and permitted adequate

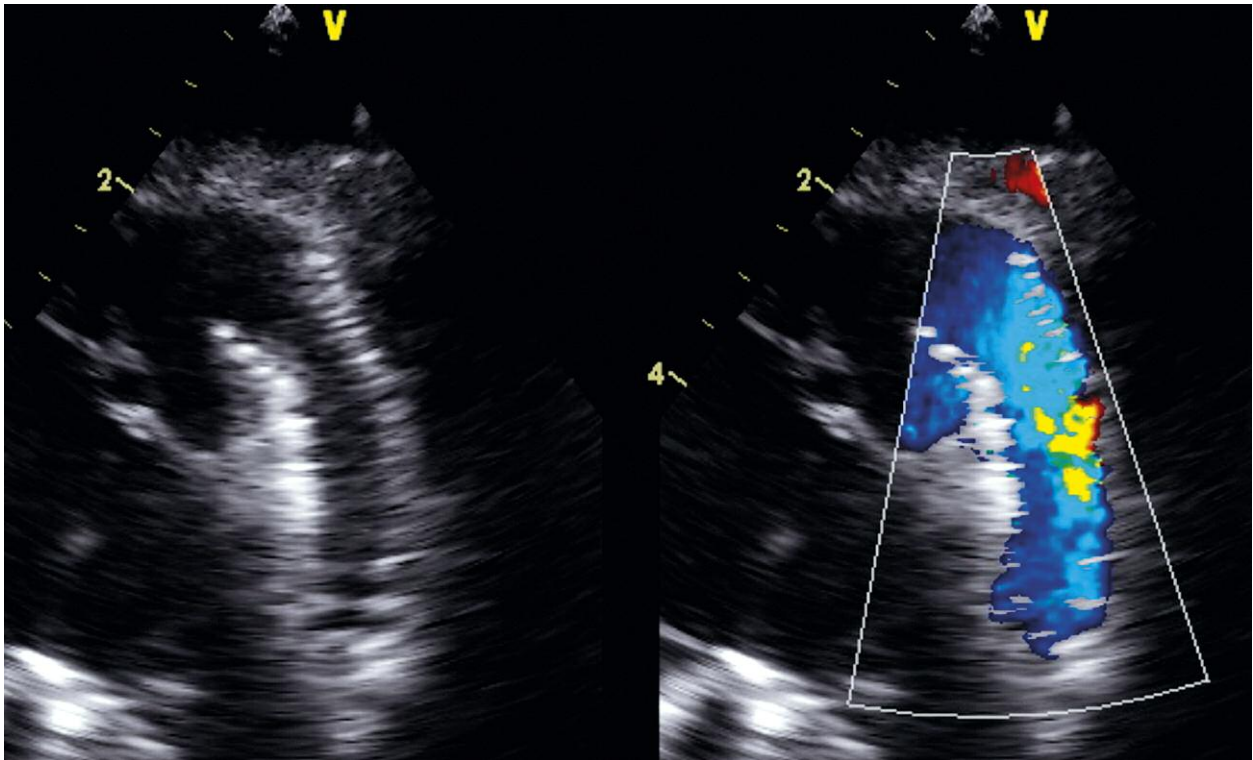


Fig. 3. Control echocardiographic examination after second stent into ductus arteriosus implantation – non-restrictive blood flow from pulmonary artery into aorta

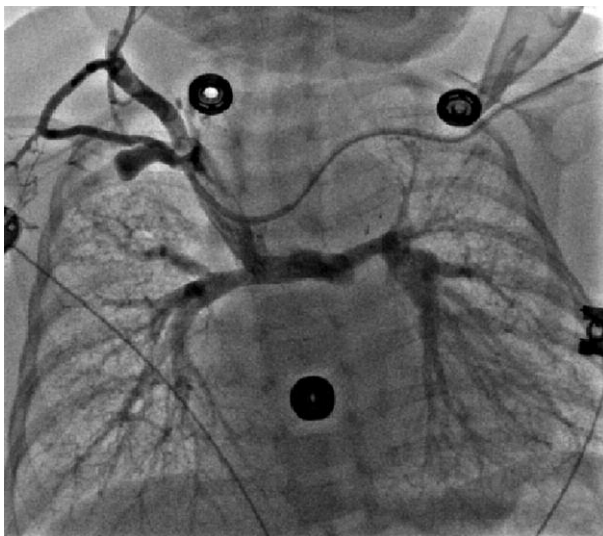


Fig. 4. Phlebography – status after HLHS comprehensive hybrid stage II. Antero-posterior view; contrast infusion through vena subclavia sinistra; opacification of both pulmonary arteries through Glenn anastomosis

preparation of the child for the second stage of HLHS treatment – the HLHS comprehensive hybrid stage II. Hybrid treatment consisting of classic cardio-surgical methods and interventional cardiology techniques is a modern, alternative form of treatment due to reduction of potential side

effects of both methods used individually, particularly in patients at the highest risk [5-7].

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