Pediatrics and Neonatology (2016) **57**, 145—148

Available online at www.sciencedirect.com

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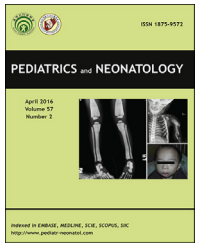

CASE REPORT

Transjugular Balloon Pulmonary Valvuloplasty Through a Bidirectional Glenn Shunt for Dysplastic Pulmonary Valve Stenosis in an 8.7-Year-Old Boy with Inaccessible Femoral Veins

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Received Jan 31, 2013; received in revised form Jun 10, 2013; accepted Jun 18, 2013
Available online 6 October 2013

Key Words
cyanosis;
dysplastic pulmonary valve stenosis;
dyspnea;
Glenn shunt;
transjugular balloon pulmonary valvuloplasty

An 8.7-year-old boy was affected by exertional dyspnea with cyanosis of the lip at 6 years old. Oxygen saturation (SpO_2) was 66%. A bidirectional Glenn shunt (BGS) was constructed to successfully elevate SpO_2 to 88%. Unfortunately, he again experienced exertional dyspnea with flagrant cyanosis of the lip at 8.5 years old. SpO_2 decreased to 65%. Echocardiography revealed a dysplastic pulmonary valve with severe stenosis. Considering the potential growth of the right ventricle and the branch pulmonary arteries, transjugular balloon pulmonary valvuloplasty (BPV) through a BGS was performed as a palliative treatment for cyanosis in this boy because of inaccessible femoral veins. After gradational BPV, the opening of the pulmonary valve was dilated from 2.59 mm to 6.65 mm, the pressure gradient decreased from 60 mmHg to 25 mmHg, and the SpO_2 increased to 85%. He became physically active and was free of exertional dyspnea at the 12-month follow-up. BGS is irrefutably an alternative vascular access through which transjugular BPV could be performed to ameliorate cyanosis due to dysplastic pulmonary valve stenosis in patients with inaccessible femoral vessels.

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http://dx.doi.org/10.1016/j.pedneo.2013.06.007

1. Introduction

Since the pioneer report describing a technique of using the radial forces of balloon inflation of a balloon catheter positioned across the pulmonary valve, balloon pulmonary valvuloplasty (BPV) has become the treatment of choice for isolated valvular pulmonary stenosis because of its excellent long-term results and minimal complications.[1] This paper reports the case of an 8.7-year-old boy presenting with cyanosis due to severe dysplastic pulmonary valve stenosis, which was treated successfully by transjugular BPV through a bidirectional Glenn shunt (BGS). The clinical dilemma encountered at performing interventional cardiac catheterization through alternative vascular access in some patients with congenital heart diseases will be briefly discussed.

2. Case Report

An 8.7-year-old boy, who had been treated by radiofrequency guidewire pulmonary valvotomy (RFPV) for pulmonary atresia with intact ventricular septum (PAIVS; Z-value was − 4.5 and was − 1.5 for the pulmonary valve and the tricuspid valve, respectively) since birth, presented with exertional dyspnea with cyanosis of the lip at 6 years old. A BGS was constructed at that time to treat the cyanosis. Oxygen saturation (SpO_2) was elevated significantly from 66% to 88% after BGS. Unfortunately, exertional dyspnea with cyanosis of the lip became especially bothersome at 8.5 years old. At the outpatient clinic, SpO_2 decreased to 65%. Two-dimensional echocardiography showed a small opening (2.59 mm) in the dysplastic pulmonary valve. The pulmonary valve and tricuspid valve measured 6.5 mm and 11.2 mm in diameter, respectively. Intra-atrial right-to-left shunting through incompetent patent foramen ovale (4 mm) was present. However, transthoracic echocardiography could not accurately estimate the transvalvular pressure gradient or assess the severity of valvular pulmonary stenosis. The patient had mild pulmonary regurgitation. He was admitted for cardiac catheterization to assess the severity of pulmonary valve stenosis. At admission, he was 8.7 years old, 17.6 kg in weight, and 113 cm in height. His heart rate was 100/min, respiratory rate 20/min, and blood pressure 100/80 mmHg. A plain chest film showed cardiomegaly (cardiothoracic ratio of 70%) and decreased pulmonary vascularity. The patient's hemoglobin level was 17.5 g/dL and hematocrit 52%. A blood coagulation study, liver function test, and renal function test were within normal limits. We did not prescribe a prophylactic antibiotic. At cardiac catheterization, both femoral veins were found to be inaccessible by a routine Seldinger maneuver. Instead, we used a transjugular approach. Prior to transjugular BPV, the peak systolic pressure of the right ventricle (RV) was 70 mmHg and that of the main pulmonary artery (MPA) was 10 mmHg. There was no significant pressure gradient between the superior vena cava (SVC), BGS, MPA, and branch pulmonary arteries. The ratio of systolic RV pressure to systolic aortic pressure (sRV/sAo) was 70%. Countercurrent angiography of the MPA showed a scanty jetting of dye from the MPA through a tiny opening in the dysplastic pulmonary valve to a hypoplastic bipartite RV (Figure 1A). A 0.035-inch Amplatz Super Stiff (Medi-tech, Boston, MA, USA) was housed in a 4-Fr Multipurpose Catheter (Cordis, Johnson-Johnson, LJ Roden, The Netherlands) and advanced as a system from the right internal jugular vein, SVC, BGS, right pulmonary artery, MPA, pulmonary valve, RV, and right atrium, and was finally left anchored deep into the inferior vena cava. A balloon catheter was railed along the guidewire to cross over and override the dysplastic pulmonary valve. Two sets of Wanda Balloon Catheters (8.0 mm × 20 mm; 12.0 mm × 20 mm; Wanda, Galway, Ireland) were used to dilate the pulmonary valve gradationally. BPV was considered complete only when the narrow waist of the inflated balloon disappeared (Figures 1B and 1C). After transjugular BPV, the systolic pressure of the RV decreased to 40 mmHg and that of the MPA was increased to 15 mmHg. The transvalvular pressure gradient decreased from 60 mmHg to 25 mmHg, and sRV/sAo ratio decreased from 70% to 40%. RV angiogram showed an increased diameter of the opening of the dysplastic pulmonary valve (Figure 1D). SpO_2 increased remarkably to 85%. The patient was free of exertional dyspnea and cyanosis of the lip at the 12-month follow-up. In summary, transjugular BPV through a BGS can be performed to relieve the patient from the bothersome cyanosis caused by dysplastic pulmonary valve stenosis.

Discussion

RFPV has been advocated as a treatment of choice for patients with PAIVS, provided that patent infundibulum is present and RV-dependent coronary circulation (RVDCC) is absent.[2] RFPV was considered a definite treatment for PAIVS, if the Z-value of the tricuspid valve was −0.1 or higher and that of the pulmonary valve was −4.1 or higher;[3] however, the cutoff Z-values of the pulmonary valve and the tricuspid valve as prerequisites for RFPV remain to be determined. RV steal phenomenon is potentially associated with ventriculocoronary circulation in patients with PAIVS; however, the presence of ventriculocoronary circulation alone, but without RVDCC, did not preclude RFPV in patients with PAIVS.

Although the femoral vein is the most preferred route for BPV, alternative routes through the jugular vein,[4] axillary vein,[5] and hepatic vein[6] have been applied successfully in patients with inaccessible femoral veins. Indications of transjugular cardiac catheterization included thrombosis of the inferior vena cava, myocardial biopsy after cardiac transplant, evaluation of patency of the total cavopulmonary connection (TCPC), SVC obstruction after the Mustard operation, failed transfemoral venous approach, and absence of the hepatic segment of the inferior vena cava.[4] The hepatic vein is another safe and effective route for interventional procedures in patients with limited venous access due to conditions such as bilateral obstructed femoral veins, and obstructed femoral veins and SVC, etc.[6]

The circumstances under which alternative vascular access should be used to perform interventional catheterization merit discussion. First, femoral veins might be sacrificed by puncture or cutdown for repeated BPV in small patients with residual pulmonary valve restenosis following RFPV for PAIVS. Second, superior cavopulmonary connection, initially constructed in patients with univentricular