Acta Cardiol Sin 2018;34:87–91
Case Report
doi: 10.6515/ACS.201801_34(1).20170731B

Balloon Aortic Valvuloplasty in a Premature Neonate with Critical Aortic Valve Stenosis Weighing 1493 g

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The use of balloon aortic valvuloplasty for congenital aortic valve stenosis was well established in literatures. However, balloon aortic valvuloplasty performed in low body weight neonates had been infrequently reported. Here we report a 5-day-old premature neonate diagnosed critical aortic valve stenosis. Balloon aortic valvuloplasty was performed as first-line therapy while the patient weighed only 1493 g. Balloon aortic valvuloplasty went successfully with transvalvular pressure gradient decreased from 80 mmHg to 44 mmHg. Aortic regurgitation after balloon aortic valvuloplasty was mild. The patient's clinical condition stabilized after balloon aortic valvuloplasty and was able to gain weight to 2665 g. Our report demonstrates that balloon aortic valvuloplasty is possible, safe and efficient as first-line approach for critical aortic valve stenosis in neonates with low body weight.

Key Words: Congenital heart disease • Infant • Percutaneous intervention

INTRODUCTION

Over the years, balloon aortic valvuloplasty (BAV) remains the first-line treatment for congenital aortic valve stenosis (AS) in many centers.[1-3] The management of critical AS is especially difficult in premature neonates due to their low body weight and more complicated clinical conditions. BAV as the first therapeutic intervention in critical AS in low body weight neonate is rarely reported in Taiwan. We report a premature neonate weighing 1493 gm undergoing BAV as the first therapeutic intervention for critical AS, which to our knowledge, is the lowest body weight reported to have BAV in Taiwan.

Received: January 29, 2017 Accepted: July 31, 2017
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CASE REPORT

A male infant born at 33 + 3 weeks' gestation was noted to have respiratory distress and heart murmur after birth. His birth weight was 1600 g and was delivered by a gravid 2, para 2 mother via Caesarean section. The mother had preeclampsia and had her blood pressure controlled at around 150/110 mmHg. Prenatal care results were otherwise normal. C-section was carried out because of hypertension. Systolic blood pressure of the mother rose to 180 mmHg accompanied with blurred vision. Only one dose of dexamethasone was given prior to the delivery. Apgar score at one and five minutes were 8 and 10 respectively. The patient had respiratory distress and a grade 2 systolic heart murmur at birth. Hepatomegaly was noted on physical examination. The patient's heart rate was 120 beats per minute and blood pressure was 63/36 mmHg. Echocardiography revealed severe AS with a pressure gradient measured 48 mmHg, relatively small left ventricle (LV) with thick interventricular septum (IVS) and posterior wall, poor LV ejection fraction (EF) (35.5%), and paradoxical septal movement. Aortic valve annulus was 0.38 cm and valve opening was

0.078-0.1 cm on long axis view (Figure 1), apical five chamber view and subcostal long axis view. Mild mitral regurgitation (MR) was noted on apical four chamber view. IVS was intact. The patient also had patent ductus arteriosus (PDA) 0.38-0.444 cm and a secundum type atrial septal defect 0.383 cm. Bidirectional flow through PDA, moderate tricuspid regurgitation, dilated right ventricle and main pulmonary artery and pulmonary arterial pressure was estimated 50 mmHg. Prostaglandin E1 (PGE1) was given to keep PDA open. Dopamine infusion was used for poor LV contractility.

The patient was admitted to neonatal intensive care unit with continuous positive airway pressure (CPAP) support. After a discussion in share decision making (SDM) meeting with nurses, pharmacist, neonatologist, pediatric cardiologist, cardiovascular surgeon, anesthesiologist, social worker, and the patient's parents, BAV was arranged on the 5th day of life. The patient weighed 1493 gm on the day when cardiac catheterization was performed. Percutaneous access to the right femoral artery was gained by direct puncture using a 21G percutaneous entry thin wall needle (5 cm; Cook, Bloomington, IN, USA) and a 4 French radio focus introducer sheath (7 cm; Terumo, Tokyo, Japan). By using 4 French Berman angiographic balloon catheter (2-lumen, 50 cm; Arrow, Chelmsford, MA, USA), we first reached main pulmonary artery (MPA) and right ventricle (RV) through PDA. MPA pressure was 54/29 mmHg (mean 40 mmHg), indicating pulmonary hypertension. RV pressure was 56/7 mmHg. There was no pressure gradient between RV and MPA.

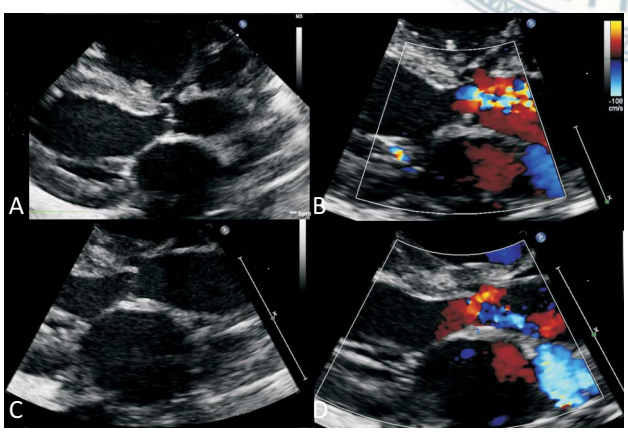

Figure 1. *Echocardiogram showed thick aortic valve with a small valve opening on long axis view (A) and color Doppler (B). After balloon aortic valvuloplasty, improvement of aortic valve opening (C) and color Doppler through aortic valve (D) was seen.*

Since it was difficult to pass the Berman catheter through aortic valve, we changed the catheter to a 4 French radio focus Optitorque Judkins Right 4.0 cm (JR4; Terumo). With the assistance of coronary Runthrough NS guidewire (floppy tip, 0.014 inch, 180 cm; Terumo), we passed JR4 catheter to LV. LV pressure was 136/9 mmHg. Ascending aorta (AsAo) pressure was 56/31 mmHg (mean 41 mmHg). Pull-back pressure gradient between LV and AsAo was 80 mmHg. LV angiography in true PA and lateral view revealed narrow aortic valve opening (Figure 2). Aortic valve annulus was measured around 4 mm by LV angiography and echocardiography. We first used Sapphirell coronary dilatation catheter (3.5 mm/10 mm; OrbusNeich, The Netherlands) for BAV by distending the balloon to 3.5 mm, 6 atm for 6 times, each time lasting 4-8 seconds. However, we were not able to see a waist while the balloon was inflated even if we distended the balloon to 3.8 mm, 10-12 atm. LV pressure (128/13 mmHg) was still high, suggesting an unsuccessful BAV result. Therefore we re-evaluated the aortic valve annulus size by repeating LV angiography. Aortic valve annulus was measured 5.35 mm on true PA view and 5.19 mm on true lateral view. Tyshak II 5 mm/20 mm percutaneous transluminal valvuloplasty catheter (NuMED Inc., Hopkinton, NY, USA) was chosen to perform BAV again. The balloon was fully distended by 6 atm for 4 times, each time lasting 4-6 seconds. Pressure gradient between LV and AsAo reduced to 44 mmHg (LV pressure 98/5 mmHg, AsAo pressure 62/26/41 mmHg) after successful BAV. We did not repeat LV angiography because we tried to reduce radiation doses. We also wanted to reduce contrast doses because renal insufficiency is a common problem in premature neonates. Since LV pressure and pull-back transvalvular pressure had decreased

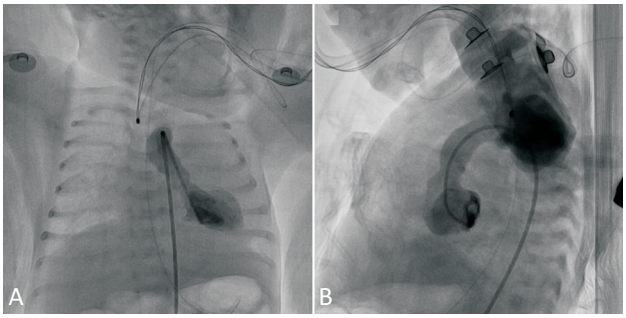

Figure 2. *Left ventriculogram (contrast injection speed 6 ml/s, contrast amount 3 ml) in posteroanterior view (A) and true lateral view (B) showed narrow aortic valve annulus and small LV.*

significantly, we believed repeating LV angiography was not necessary. We did not give anticoagulants to the patient. The intervention was performed under general anesthesia with the patient intubated and supported by a ventilator. We did not perform RV pacing.

Echocardiography after BAV showed improvement of AS (pressure gradient 32 mmHg), mild aortic regurgitation (AR) and trivial MR. Aortic valve annulus was measured 0.43 cm and valve opening was 0.342 cm after the procedure (Figure 1). We kept PGE1 and dopamine infusion before LV systolic function improved. Pulses of right dorsalis pedis and femoral artery were well palpable after the intervention.

DISCUSSION

Balloon aortic valvuloplasty has been long adopted as the first-line treatment for congenital aortic valve stenosis in many centers worldwide.[1-3] In the past, we had had experience of performing BAV on a 3-day-old newborn weighing 1556 g with multiple anomalies in 2005.[4] Many years later, we encountered a premature neonate with even lower body weight who had critical AS. In such cases, neither BAV nor surgical intervention is easy due to the patient's low body weight and complicated clinical conditions. A thorough discussion between neonatologist, pediatric cardiologist, cardiovascular surgeon, anesthesiologist, social worker, and the patient's parents was carried out before a clinical decision was made.

The safety and efficacy of BAV for critical AS are well established in literatures.[1-3,5-7] In 2015, Torres et al. assessed the outcomes of BAV through a multi-center prospective study and found high procedural success rate.[2] No death was procedure-related in the study. Twenty percent of the patients had adverse effects. The study also found out that the only factor associated with high severity adverse effect was age less than one month old when having BAV. Aside from the age of the patient, other factors more frequently reported in literatures which are associated with poor outcome after initial BAV can be categorized as the patient's clinical condition and the condition of left heart structures and function. The former includes the use of mechanical ventilation prior to the procedure and unstable hemodynamic status. The later includes smaller aortic valve annulus, smaller aortic diameter at the sinotubular junction, smaller subaortic area, lower baseline LV shortening fraction, and the presence of endocardial fibroelastosis. Some also include the morphology of aortic valve and the presence of AR.[1-3,5,7] Our case was a 5-day-old premature neonate with low body weight who required CPAP use, inotropic agent and PGE1 support. Small aortic valve annulus, small LV size and poor LVEF all complicated the situation and made any intervention challenging. Concerning the weight and age of the patient, we decided to perform BAV as the firs-line therapy to release left heart pressure and to postpone surgical intervention, if one is necessary later in life, till the patient gains weight. Fortunately, BAV was successful and only mild AR was noted after the procedure. The patient's hemodynamic condition stabilized afterwards and was able to gain weight to 2665 gm in one month.

In 2001, McCrindle et al. published a single institutional prospective study comparing the outcomes of BAV and surgical valvuloplasty (SAV) as treatment for critical AS in neonates.[5] The two treatments resulted in similar mortality and re-intervention rate. The re-intervention rate after BAV, with most follow-up time exceeds 10 years, was reported from 35% to 60%.[1-3,5-7] However, as surgical interventions advanced over time, there is a trend showing that the outcome of SAV is superior to BAV. Siddiqui et al. in 2013 compared SAV to BAV.[7] The decrease in the transvalvular pressure gradient was similar in both groups. However, in terms of re-intervention rate, SAV was superior to BAV. Even though the study concluded SAV as the best approach for congenital AS, the authors also stated that the benefit of both approaches should not only be assessed by re-intervention rate. Patients may take advantages from the delay of the second intervention.

In neonates, cardiac catheterization using femoral artery carries a risk of fermoral artery occlusion. Some centers therefore use antegrade approach, taking advantage of the existence of patent foramen ovale in neonates, to avoid femoral artery damages. Evidence showed less vascular complications with antegrade approach.[8,9] Even though certain techniques can avoid damaging the mitral valve apparatus,[8,9] it is still a concern especially in premature low body weight neonates. The balloon is relatively long considering the size of the heart in this patient group. Considering the risk of damaging the mitral

valve apparatus, we chose retrograde approach. We did not use echo-guide technique to assist femoral artery puncture because we were familiar enough with the technique of direct puncture. If the operator is familiar with echo-guide technique, it will definite add guarantee to the successfulness of the procedure. One of the worries about retrograde approach is aortic valve perforation.[9] In our case, the aortic valve was a lot thicker than the usual valve morphology. Along with the use of a floppy tip guide wire, we believed we had made the possibility of valve perforation low enough. Follow-up echocardiography showed no evidence of aortic valve perforation. AR remained mild after BAV. There was no sign of poor right foot perfusion. Pulses of right femoral artery and dorsalis pedis were strong after the intervention.

To secure the balloon during BAV, many techniques have been used. One of the most popular one is rapid RV pacing. By increasing the patient's heart rate, cardiac output is lowered to achieve better balloon stability. However, rapid RV pacing has several down sides including longer procedure time and the drops in blood pressure and cardiac output.[10] Low body weight neonates are more susceptible to blood pressure and cardiac output changes. Also, to maintain the body temperature of a preterm neonate is always an issue. Lengthening the procedure time will make it even more challenging. When performing BAV in such patients, the balloon is relatively long because the size of the heart is small. Balloon movement therefore has less impact on the result of the procedure. In our case, we used a balloon that is 20 mm long. Without rapid RV pacing, BAV was performed effectively without difficulty.

In our case, transvalvular pressure gradient was low on echocardiography comparing to the measurement on catheterization. Sonography is an indirect method and we recommend giving priority to the results of catheterization. The aortic valve annulus was 0.38 cm measured by echocardiography and 4 mm on the first LV angiography. However, when we used a 3.5 mm balloon, a waist was not seen when the balloon was fully inflated. On the repeated LV angiogram, the annulus was 5.35 mm. We do not know whether it is the different aspects that echocardiograph and angiography takes on examining the heart or the impact of the first BAV attempt which causes the discrepancy on the measurement. However, this experience suggests that echocardiography and angiography can be incorrect on measuring the annulus size. To prevent complications of BAV, we recommend operators to be conservative at first but re-evaluate and adjust balloon size if the first attempt fails.

CONCLUSIONS

Our case demonstrats that BAV is possible and safe in treating critical AS in neonates with low body weight. For such cases, BAV is a reasonable first-line management either as therapeutic approach or an adjunct to surgical intervention.

ACKNOWLEDGMENT

None.

CONFLICT OF INTEREST

All the authors declare no conflict of interest.

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