Case Report

Combined Aortic and Pulmonic Valvular Stenosis: Report of 2 Cases

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Objective: *To report 2 cases of severe combined aortic and pulmonic valvular stenosis.*
Material and Method: *To find patients with a diagnosis of severe combined aortic and pulmonic valvular stenosis, the authors searched in the patient profile at the cardiology unit, Queen Sirikit National Institute of Child Health (QSNICH) from 1995 to 2010.*
Results: *There are 2 cases of severe combined aortic and pulmonic valvular stenosis from 19,416 case records of pediatric cardiac patients. The first one is a 9-year-old girl and the other is a male neonate. The interval between the presentations of these two cases was 10 years. The older patient, who had associated moderate aortic regurgitation, underwent aortic root replacement and pulmonic valvulotomy in the year 2000. The neonate, who presented in 2010 had undergone bilateral percutaneous balloon aortic and pulmonic valvuloplasty and required repeated balloon aortic valvuloplasty at the age of two months. Both patients responded well to treatment and were asymptomatic at the last follow-up of 10 years and 6 months respectively after treatment.*
Conclusion: *Combined aortic and pulmonic valvular stenosis is a very rare congenital heart disease. The prevalence is 0.01% of congenital heart disease at QSNICH. Bilateral balloon valvuloplasty of aortic and pulmonic valve is effectively performed in this rare congenital heart disease and can be done safely in a sick neonate.*

Keywords: *Aortic valve stenosis (AVS), Pulmonic valve stenosis (PVS), Congenital heart disease (CHD), Aortic valvuloplasty, Pulmonic valvuloplasty*

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Aortic valve stenosis (AVS) is not uncommon in Western countries but relatively rare in the Eastern countries. The prevalence of AVS in infants from the literature is 3-8% of congenital heart disease (CHD), compared to 1.38% of the non-published data from Queen Sirikit National Institute of Child Health (QSNICH). Pulmonic valve stenosis (PVS) is more common than AVS. The prevalence of PVS from the literature is about 8-10% of CHD, which is higher than QSNICH data of 5.5%. Combined AVS and PVS disease is hardly ever seen, the first report was published in 1957 by Horlick[1]. As far as the authors knew, there were 11 more papers about this type of CHD[2-9], only 3 papers reported in the last 3 decades[7-9]. Some reported cases were associated with VSD, ASD or PDA and some had Noonan syndrome. Severe outflow tract obstruction, either on the left or right side of the heart, in early neonate can be treated with PGE-1 to open the ductus arteriosus. However, the patient with severe hemodynamic obstruction of both sides of the heart could not survive by medical treatment only. Management, either by surgical or catheter intervention in severe combined AVS and PVS, is difficult especially in the neonatal period. Inadequate surgical relief of either aortic valve or pulmonic valve obstruction may end up with fatal outcome. Bilateral balloon aortic and pulmonic valvuloplasty is an option of treatment[7,9] and the procedure can be repeated.

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Material and Method

QSNICH is a super-tertiary care hospital with more than 1,300 new cases of congenital heart disease per year. In the past fifteen years, the authors have

seen two cases of severe combined aortic and pulmonic valvular stenosis.

Case 1

A 9-year-old girl was presented in April 2000. She was a known case of congenital heart disease due to mild cyanosis and cardiac murmur since birth. Her parents refused surgery in the past. She had mild symptoms on exertion but could walk to school. Physical examination showed that she was thin, 14 kg body weight, mild cyanosis, normal peripheral pulses, systolic thrill at LUSB, holosystolic murmur grade 4/6 with systolic ejection click at LUSB, diastolic blowing murmur grade 2/4 at LUSB and normal S2. Her chest X-ray showed mild cardiomegaly with normal pulmonary vasculature and her electrocardiogram showed normal sinus rhythm, QRS axis (+) 80 degree with combined ventricular hypertrophy. Her echocardiogram showed severe PVS with peak pulmonic valve gradient of 130-150 mmHg and moderate TR (TR jet 150 mmHg). She also had severe AVS with peak aortic valve gradient of 88 mmHg and moderate AR. The AV annulus was 17-18 mm in diameter, mild dilate ascending aorta and main pulmonary artery. Both ventricles showed hypertrophy with normal size and good LV systolic function.

She had undergone aortic root replacement with 18 mm homograft and pulmonic valvulotomy performed in May 2000. Post operative echocardiogram showed good ventricular systolic function, mild sub-aortic narrowing without significant gradient across the LV outflow tract and no aortic regurgitation. No significant pulmonary stenosis but moderate pulmonic valve regurgitation was detected.

She has been well without any symptoms after the operation. The recent echocardiogram performed in May 2010, at 10 years post operative showed calcified AV, mild AVS with peak gradient 34 mmHg and mild AR, no residual PS, moderate PR, mild TR (TR jet 29 mmHg), mild right ventricular enlargement, normal size and good LV systolic function with mild MR.

Case 2

An 18-day-old boy was presented at QSNICH in April 2010 with difficulty breathing. He was born with normal, uneventful delivery, 2.5 kg at birth with Apgar score of 9, 10 at 1 and 5 minutes respectively. His cardiac murmur has been noted since birth. Physical examination showed that his weight was 2.9 kg. He was afebrile with heart rate of 150/min, Respiratory rate 50/min, BP at right arm 88/63 mmHg, mild dyspnea, no cyanosis, and poor peripheral pulses of all extremities. His cardiac examination showed slightly active precordium, left ventricular (LV) heaving; systolic ejection murmur grade 3 at left upper sternal border, normal S2. Both lungs were clear. Liver was palpated 2 cm below right costal margin and spleen was just palpable. His chest x-ray showed moderate cardiomegaly with mild pulmonary congestion (Fig. 1) and his electrocardiogram showed sinus tachycardia, low voltage of both limb and chest leads with right atrial enlargement and ST-T wave change. His echocardiogram showed that there was severe AVS, dome shape of aortic valve with peak gradient of 61 mmHg and mean gradient 38 mmHg (Fig. 2). The aortic valve (AV) annulus was 8 mm in diameter. There was severe PVS, dome shape of pulmonic valve with peak gradient of 68 mmHg (Fig. 3). The PV annulus was 7.6 mm in diameter. He also had mild tricuspid regurgitation (TR) with small perimembranous ventricular septal

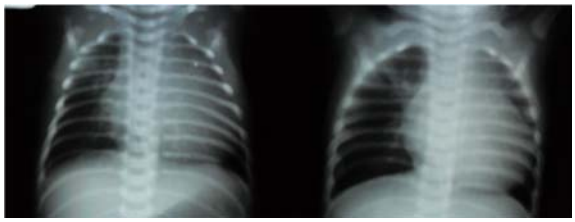

Fig. 1 Chest x-ray of the newborn patient (Case 2) shows cardiomegaly and pulmonary congestion before (on the left) and less prominent heart size (on the right) after bilateral balloon valvuloplasty

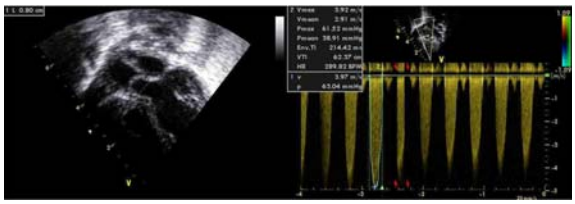

Fig. 2 Five chamber view echocardiogram shows doming of aortic valve and high Doppler flow gradient across the aortic valve (Case 2)

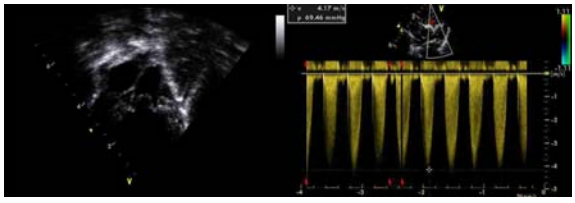

Fig. 3 Mid parasternal short axis view echocardiogram shows doming of pulmonic valve and high Doppler flow gradient across the pulmonic valve (Case 2)

defect (VSD) 3.5 mm in diameter and small patent ductus arteriosus (PDA) 2.5 mm in diameter. Color flow Doppler showed small left to right shunt at the patent foramen ovale (PFO), VSD and PDA. The LV function was poor with left ventricular ejection fraction (LVEF) was 38%. Both ventricles showed hypertrophy.

Left and right heart catheterization was performed on the next day. His hemodynamic was very unstable. Very poor left ventricular function was observed from fluoroscopy during the catheterization and also from ascending aortogram. The catheter could not pass across the aortic valve retrogradely due to severe stenotic of aortic valve. Femoral arterial-venous loop across the aortic valve was performed by using 0.025 floppy guide wire across the aortic valve antegradely and snare in the aorta. Balloon aortic valvuloplasty was performed by using 8*20 mm balloon catheter (Smash) over the wire via right femoral vein. (Fig. 4). The patient developed a short period of ventricular tachycardia and ventricular fibrillation during balloon inflation and needed cardiac massage, DC cardio-version and intravenous amiodarone. After restoration of the vital signs and his ECG returned to normal sinus rhythm, balloon pulmonic valvuloplasty was performed by using 10*20 mm balloon catheter (Smash) (Fig. 5). His echocardiogram on the subsequent day showed improved LV systolic function with mild residual AVS (45 mmHg gradient) and mild residual PVS (45 mmHg gradient). He was quite well and hemodynamically stable during the recovery period with continuation of intravenous inotropic support for a few days post valvuloplasty. He was discharged from the hospital after a three-week course of antibiotic treatment for clinical sepsis.

One month after bilateral balloon valvuloplasty, he still had mild dyspnea during feeding. Echocardiogram showed improved LV systolic function to normal with increased gradient across the aortic valve to 61 mmHg but further decrease in gradient across pulmonic valve to 27 mmHg, tiny perimembranous VSD (46 mmHg gradient across the VSD) and tiny PDA. His peripheral pulses were still poor. At the age of 2 months, repeated balloon aortic valvuloplasty was performed via right femoral artery approach by using 10*20 mm EverCross EV3 balloon catheter could pass across the aortic valve retrogradely over 0.035 exchange wire. Transient sinus bradycardia occurred during balloon inflation which needed a bolus dose of intravenous adrenaline. The gradient across AV was decreased from 54 to 20 mmHg with good peripheral pulses after the second procedure.

Echocardiogram at one day after the second balloon aortic valvuloplasty showed mild residual AVS with peak gradient of 29 mmHg and mild aortic regurgitation, mild residual PVS with gradient of 27 mmHg and mild pulmonary regurgitation (PR), spontaneously closed VSD and PDA, nearly normal LV systolic function (LVEF 52%).

He was quite well, asymptomatic, normal growth and development on the last follow-up at the age of 7 months. He has been on digoxin, furosemide

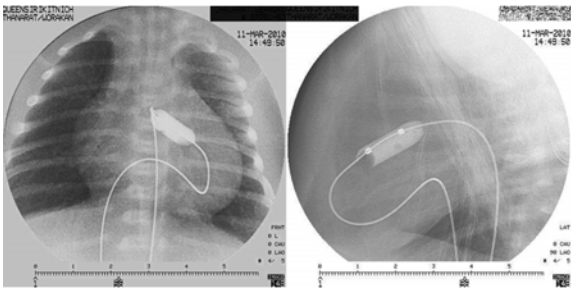

Fig. 4 Balloon aortic valvuloplasty: AP and lateral views. The balloon catheter passes across the aortic valve antegradely via femoral vein by using femoral arterial-venous loop

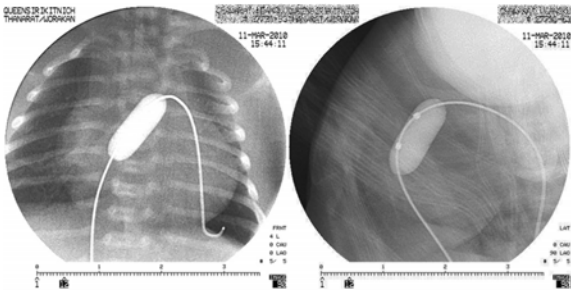

Fig. 5 Balloon pulmonic valvuloplasty: AP and lateral views

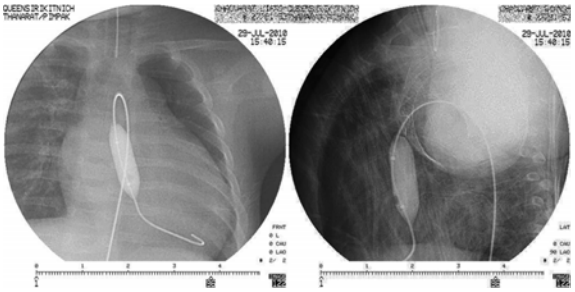

Fig. 6 Repeated balloon aortic valvuloplasty at two months of age, the balloon catheter passes across the aortic valve retrogradely: AP and lateral views

and enalapril since then.

Discussion

Combined AVS and PVS is a rare cardiac anomaly. So far, less than 15 cases have been reported in the literatures, including these two reported cases. It is possible that there may be more mild cases, which have not been reported. The prevalence of AVS is about 1.38% of CHD compared to 5.5% of PVS and 0.01% of severe combined AVS and PVS from non-published data at QSNICH. The severe obstruction of both ventricular outflow tracts of the heart is not compatible with life, even in the fetal circulation. However, bilateral hemodynamic obstruction of the heart may occur in association with many cardiac anomalies. Supravalvular aortic stenosis and peripheral pulmonary artery branches stenosis can be seen in William syndrome. Sub-aortic and sub-pulmonic conus with some obstruction may be seen in double outlet right ventricle or hypertrophic cardiomyopathy. Sub-aortic membrane may be associated with doubly committed VSD. This lesion is associated with high prevalence of aortic valve prolapsed and it can protrude into the right ventricular outflow tract causing infundibular obstruction. Treatment of combined AVS and PVS is difficult in severe cases due to unstable hemodynamics from pressure load of both ventricles conjunction with low myocardial perfusion and low cardiac output. Myocardial ischemia may be a problem during cardiac catheterization or perioperative period. Surgical correction has been the gold standard treatment for CHD for a long time. The first report of a survived case from surgical treatment in combined AVS and PVS was in 1979[6]. After the successful nonsurgical treatment of valvular stenosis by using percutaneous balloon valvuloplasty, bilateral balloon aortic and pulmonic valvuloplasty had been an optional treatment for combined AVS and PVS. There were only two success reports which used this procedure for treatment of combined AVS and PVS in an adult patient in 1994[7] and a neonate patient in 2009[9]. Morbidity and mortality can occur in either form of treatment, especially in neonates with poor pre-operative condition. Ventricular arrhythmia during the catheter manipulation or balloon occlusion of the outflow tract in patients with unstable hemodynamic is expected. The increment in pressure gradient across the aortic valve after an immediate decrease post balloon aortic valvuloplasty in this neonate case could be from the improvement in cardiac function with significant residual obstruction, which is an indication for redo valvuloplasty.

Conclusion

The authors report 2 cases of severe, combined aortic and pulmonic valvular stenosis. These two cases were presented at a period of 10 years interval. The first child who presented in childhood with severe, combined aortic and pulmonic valvular stenosis with moderate aortic valve regurgitation had undergone aortic root replacement and pulmonic valvulotomy. The second one, who presented during the neonatal period with poor ventricular function and poor cardiac output, needed urgent balloon valvuloplasty of both aortic and pulmonic valves. This infant needed aortic valvuloplasty redo at two months of age. Both patients responded well to treatment and were stable in functional class I for 10 years and 6 months respectively after treatment.

Potential conflicts of interest

None.

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โรคลิ้นหัวใจเอออร์ติกร่วมกับลิ้นหัวใจพัลโมนิกตีบแต่กำเนิด: รายงานผู้ป่วย 2 ราย

ธนะรัตน์ ลยางกูร, วรการ พรหมพันธุ์, ธวัชชัย กิระวิทยา, ชัยสิทธิ์ แสงทวีสิน, พีระพัฒน์ มกรพงษ์

วัตถุประสงค์: *เพื่อรายงานผู้ป่วยเด็กที่มีลิ้นหัวใจเอออร์ติกร่วมกับลิ้นหัวใจพัลโมนิกตีบที่รุนแรงแต่กำเนิด*
วัสดุและวิธีการ: *ผู้นิพนธ์ได้ค้นหาผู้ป่วยเด็กโรคหัวใจที่ได้รับการวินิจฉัยว่าเป็นลิ้นหัวใจเอออร์ติก ร่วมกับลิ้นหัวใจพัลโมนิกตีบที่รุนแรงแต่กำเนิด โดยค้นหาจากฐานข้อมูลของหน่วยโรคหัวใจสถาบันสุขภาพเด็กแห่งชาติ มหาราชินี ตั้งแต่ปี พ.ศ. 2538 ถึง 2553 รวม 15 ปี*
ผลการศึกษา: *พบว่ามีผู้ป่วยที่ได้รับการวินิจฉัยว่าเป็นโรคลิ้นหัวใจเอออร์ติกร่วมกับลิ้นหัวใจพัลโมนิกตีบแต่กำเนิดที่รุนแรงจำนวน 2 ราย จากผู้ป่วยเด็กโรคหัวใจทั้งหมด 19,416 ราย รายแรกเป็นเด็กหญิงอายุ 9 ปี มาตรวจรักษาตั้งแต่ปี พ.ศ. 2543 มีลิ้นหัวใจเอออร์ติกรั่วปานกลางร่วมด้วย ได้รับการผ่าตัดทำการเปลี่ยนฐานรากลิ้นหัวใจเอออร์ติกและผ่าขยายลิ้นหัวใจพัลโมนิก รายที่สองเป็นเด็กทารกแรกเกิดอายุ 18 วันได้รับการวินิจฉัยในปี พ.ศ. 2553 ได้รับการรักษาด้วยการสวนหัวใจขยายลิ้นหัวใจทั้งสองในคราวเดียวกัน เด็กรายนี้จำเป็นต้องได้รับการขยายลิ้นหัวใจเอออร์ติก ซ้ำอีกครั้งเมื่ออายุ 2 เดือน จากการติดตามอาการครั้งหลังสุดที่ 10 ปี และ 6 เดือนหลังการรักษาตามลำดับ พบว่าผลการรักษาของเด็กทั้งสองราย เป็นที่น่าพอใจไม่มีอาการผิดปกติ อยู่ใน functional class I.*
สรุป: *ลิ้นหัวใจเอออร์ติกร่วมกับลิ้นหัวใจพัลโมนิกตีบที่รุนแรงแต่กำเนิดมีรายงานน้อยมาก อุบัติการณ์ของโรคนี้ที่สถาบันสุขภาพเด็กแห่งชาติมหาราชินีพบประมาณร้อยละ 0.01 ของเด็กที่เป็นโรคหัวใจการรักษาด้วยวิธีการสวนหัวใจเพื่อขยายลิ้นหัวใจ ทั้งสองในคราวเดียวกัน สามารถทำได้ด้วยความปลอดภัยแม้ในทารกแรกเกิดที่มีอาการรุนแรงและเป็นการรักษาทดแทนการผ่าตัดได้*