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NEONATAL AORTIC STENOSIS WITH SHOCK - A TALE OF DRAMATIC RECOVERY

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Abstract :
We report a case of a 14 day old newborn baby who presented with congestive heart failure and cardiogenic shock to our hospital. After evaluation the baby was diagnosed to have bicuspid aortic valve with severe stenosis and left ventricular dysfunction on two dimensional echocardography. Emergency percutaneous balloon aortic valvotomy was performed. Post-procedure the child showed immediate relief of aortic stenosis with marked improvement in clinical status and left ventricular ejection fraction. These improvements were maintained at follow up.
Keyword : Aortic valve stenosis, Bicuspid aortic valve, Balloon Valvotomy

INTRODUCTION:
Congenital aortic stenosis is a spectrum of abnormalities ranging from normally functioning but abnor mal bicuspid or unicuspid aortic valves, to the life threatening severe or critical aortic stenosis manifesting in the severest form as hypoplastic left heart syndrome. Among these stenosis due to congenit al bicuspid aortic valve (BCAV) is the most common, which commonly is asymptomatic upto adulthoo d except in minority of the patients who present with signs and symptoms of congestive heart failure i n infancy (1). These patients if deemed to possess adequate anatomy to support two ventricle circulat ion should undergo emergency balloon valvulotomy (BAV) or surgical valvotomy although the former has now become the treatment of choice for such cases (1,2). We report a case of emergency BAV p erformed in a 14 day neonate presenting with cardiogenic shock and circulatory collapse diagnosed w ith congenital aortic stenosis. There was a dramatic improvement in left ventricular (LV) function in th e immediate post procedure hospital stay with the child doing well at a four month follow up.

PROCEDURE :
A 14 day neonate presented to paediatric casualty at our hospital in respiratory distress

(respiratory rate 87/min), with cold clammy peripheries and prolonged capillary refill time. The systolic blood pressure was recorded as 68mm of Hg. On examination a grade III/VI harsh crescendo-drescendo ejection systolic murmur was heard at the base of the heart. A two dimensional echocardiogram (2D ECHO) done showed restricted aortic valve opening , a bicuspid aortic valve with a peak and mean gradient of 59 mm Hg and 36mm Hg respectively across the BCAV (Figure 1A, 1B).There was severe left ventricular systolic dysfunction with a LV ejection fraction of 30%. Left atrium and left ventricle were dilated.There was no patent ductus arteriosus, thoracic or abdominal coarctation and no interruption of aorta noted on 2D ECHO.

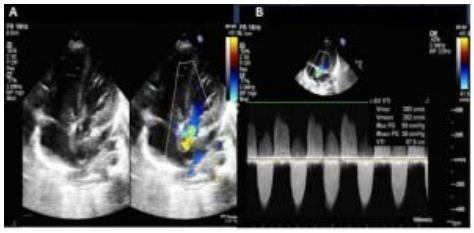

Figure 1. A) Transthoracic 2D ECHO with Apical 5 chamber showing restricted aortic valve opening and turbulence across the aortic valve. B) Continuous wave Doppler showing a mean gradient of 36mm Hg across the aortic valve

The patient was taken up for an emergency balloon aortic valvotomy (Figure 2). The child was administered general anaesthesia by paediatric cardiac anaesthetist. The right femoral artery and vein were percutaneously cannulated with modified Seldinger technique. The aortic valve was crossed with a 5 French (F) Judkins right (JR) diagnostic catheter and 0.025 inch Terumo glide wire (260 cm).

The pressure tracings of the LV and ascending aorta were recorded with a LV-Aorta (Ao) peak to peak gradient noted as 31 mm Hg (Figure 3). The catheter was changed to Pig tail catheter and the glide wire was replaced with 0.025" extra-stiff Amplatz super stiff wire. The aortic valve annulus measured was 10 mm and thus dilation was carried out with 10mm x 30mm Tyshak II (NuMed. Canada Inc.) balloon dilation cathter (Fig 2B). Post dilatation gradient decreased significantly with a gradient of 10 mm of Hg measured on catheter pull back across the aortic valve (Figure 3), there was moderate aortic regurgitation visualized angiographically (Figure 2C) immediately post procedure.

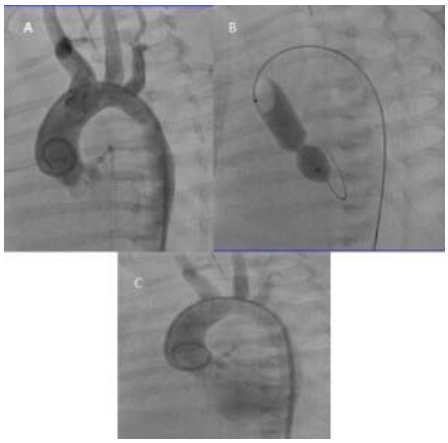

Figure 2 : A) Aortic angiogram showed severe aortic valvar stenosis. B) Balloon dilation across the aortic valve with a waist seen. C) Post BAV aortic root angiogram showed good opening of aortic valve leaflets with moderate aortic regurgitation

The immediate post procedure 2D ECHO showed good opening of the aortic valve with significant improvement in the left ventricular systolic function; also moderate aortic regurgitation (AR) was noted .

There were no local or systemic complications noted. The child was monitored in the PICU and discharged the next day with oral decongestive therapy. At four months on follow up examination the child showed had significant clinical improvement with 2D ECHO showing unrestricted opening of the aortic valve with mean gradient of 10 mm Hg (Fig 4A), normalized left ventricular ejection fraction (LVEF 50%) and only mild aortic regurgitation was noted (Figure 4B).

DISCUSSION :

Congenital aortic stenosis occurs in different forms and is classified according to the level of obstruction in relation to the aortic valve i.e valvar, subvalvar or supravalvar. Valvar aortic stenosis comprises 60- 75 % of cases of congenital aortic stenosis and generally is not apparent in infancy and early childhood (3). Stenosis of the valves occurs due to cusp fusion, thickening and rigidity of valve leaflets with the most common malformation being the bicuspid aortic valve (1). Congenital BCAV is one of the most common congenital abnormality noted with autopsy studies having reported a prevalence of 1.3 % in the population (4). Males predominate with a reported male to female ratio ranging from 3:1 to 5:1 (1,3). The most common associated anomalies are ventricular septal defect, PDA and coarctation of aorta (1).

Bicuspid aortic valve results from partial or complete fusion of two of the aortic valve cusps with or without a raphe present at the site of fusion. The most commonly conjoined cusps are the right and left followed by the right and the noncoronary cusps with patients with the latter showing more rapid progression of fusion, calcification and stenosis (1,5). In addition to the morphological abnormality of the valve leaflets other factors like myxomatous degeneration, inflammatory changes, fibrosis, lipid accumulation, calcification, annular dilation and acquired fibrotic fusion of true commissures play a role in valve dysfunction. Only 10% of the cases of congenital aortic valve stenosis present with features of congestive heart failure in infancy (1). The presence of symptoms is dependent on the functional response of the left ventricle to pressure overload which in turn depends on the severity of stenosis. If the left ventricular size and function are adequate to handle the cardiac output, even patients with mild to moderate aortic stenosis may have only mild or no symptoms even after closure of the PDA ; but in the severest forms the LV may not

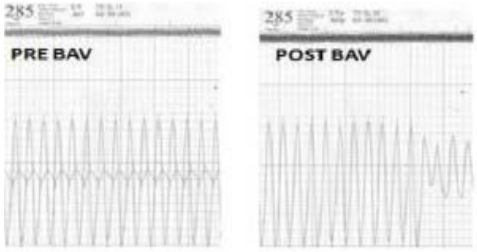

Figure.3 Pre- BAV simultaneous LV and ascending aorta pressure trace showing peak to peak LV- Ao gradient of 31 mm Hg. On the right is a post BAV pullback pressure tracing across the aortic valve showing mild residual gradient of 10mm Hg

compensate for the increased demands after birth and the neonate presents with shock and circulatory collapse (1). Initial management of neonatal critical aortic stenosis consists of maintaining ductal patency with prostaglandin E1 infusion. The neonates with severe or critical aortic valve stenosis with adequate two ventricle function should be offered balloon aortic valvuloplasty or surgical valvotomy with the former now being the

treatment strategy of choice (1,2). More recently, temporary palliation has been achieved with the stage I hybrid procedure involving bilateral pulmonary artery banding and PDA stenting.This tides over the acute crisis, provides an oppurtunity for better assessment of LV adequacy and increases the outcome success of definitive surgery performed outside the neonatal period (1). Percutaneous balloon valvuloplasty for critical aortic valve stenosis in infancy was first described in 1985 and since then has undergone vast improvements in techniques, safety, ease and eventual successful outcomes. It has now replaced surgery as the treatment of choice for palliation in infancy (2). Techniques for balloon aortic valvotomy include the 1.**retrograde approach** via the femoral (most common), umblical and the carotid artery**, 2. the antegrade approach** (2)**.**The retrograde approach via the femoral arteries is the most commonly used technique world over. The balloon dilation catheter used is 0.8-0.9 times the aortic annulus size with the smaller size being used if there is discrepancy between the annulus measurements between echocardiography and on cardiac catherization (2). A true waist may not be always be visible in these patients compared to those in the older age groups. One or two low pressure dilations across the valve are usually adequate and care must be taken not to overinflate or rupture the cusps

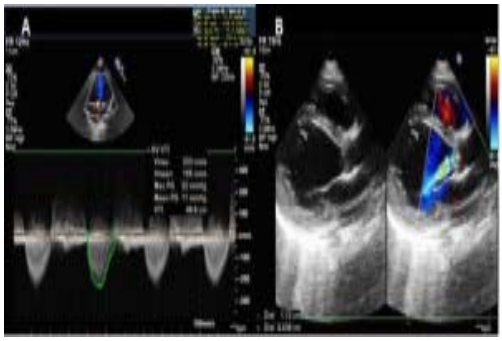

Figure 4. Follow up 2D ECHO at four months showing A) Continous wave doppler across AV with mean gradient of 11mm Hg. B) Parasternal long axis view showing unrestricted opening of aortic valve leaflets with mild AR.

which may result in increased aortic insufficiency. Pass et al have suggested a consensus criteria for success of valvuloplasty which are :1) a fall in peak systolic ejection gradient 50%; 2) a fall in LVEDP; and 3) in patients on prostaglandins with an open ductus, a improved lower extremity oxygen saturations (2).The most common complication is femoral or iliac arterial injury with pulse loss noted in 39% of patients undergoing access via femoral arteries. In the current era with use of smaller sheaths, there has been a marked decrease in incidence of these complications (2). Balloon dilation of the cusp may result in aortic insufficiency (15%) (6) , the only patient-related or procedural factor associated with decreased freedom from moderate or severe AR being larger balloon-annulus ratio. Prolonged umblical artery manipulation was associated with sepsis and vessel disruption whereas the transcarotid approach carries the risk of residual restenosis of the the right carotid artery (2). Overall BAV for AS during the first 60 days of life results in short-term relief of AS in the majority of patients. Among early survivors, initially small left heart structures may be associated with worse subacute outcomes but typically normalize within 1 year. Reintervention for residual/ recurrent AS or iatrogenic AR is relatively common, particularly during the first year after balloon valvuloplasty, but aortic valve replacement during early childhood is seldom necessary (6).

In conclusion BAV is a life saving intervention in severe aortic stenosis in infancy and is a safe palliative measure to tide over this life threatening crisis in patients presenting with shock and circulatory collapse.

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