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Pre-natal Echocardiographic Diagnosis and Neonatal Balloon Dilatation of Severe Valvar Pulmonic Stenosis

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Abstract. A case is presented below where pre-natal echocardiographic diagnosis of critical pulmonic valvar stenosis was made at 36 weeks of gestation. In view of the severe heart failure, successful balloon valvotomy was performed on day 4 of life. The child was asymptomatic at one month follow-up. **(Indian J Pediatr 1999; 66 : 799-803)**

***Key words** : Fetal echocardiography; Pulmonic stenosis; Neonatal intervention.*

Fetal echocardiography is a simple method to diagnose congenital heart disease(CHD) in utero[1,2]; outcome and management of neonate can be planned accordingly. Among the CHD, valvar pulmonary stenosis occurs in 8-10% of patients[3]. Critical neonatal pulmonary stenosis can be treated by percutaneous transluminal balloon valvuloplasty[4,5]. We report a case of severe pulmonic valvar stenosis with right ventricular failure, diagnosed in-utero by fetal echocardiography and then successfully balloon dilated on the 4th day of life.

CASE REPORT

A 36-year-old primiparous lady underwent a routine antenatal ultrasonography scan at 36 weeks of gestation. The fetus was suspected to have a cardiac tumour. She was referred to the Department of Cardiology, K.E.M. Hospital, Mumbai for a fetal echocardiogram.The study was done on a Hewlett-Packard Sonos 1000 machine using a 2.5 MHz transducer. The echocardiogram revealed aneurysmal dilatation of the right atrium (RA) and the right ventricle (RV) (Fig. 1) There was severe valvar pulmonic stenosis(PS). The pulmonic valve was thick and doming with an annulus of 7.3 mm. The peak systolic gradient across the pulmonary valve was 25 mmHg. There was moderate functional tricuspid regurgitation. The interventricular septum was intact. The RV contractility was impaired. No other abnormality was detected. A diagnosis of severe valvar PS with RV dysfunction was made. The parents were counselled and an in-hospital delivery was planned, so that the baby could be taken care of immediately after birth.

The lady delivered a 2.5 kg. male child after a few days. The neonate showed signs of right heart failure. There were no extracardiac anomalies. An echocardiogram confirmed the prenatal diagnosis. The gradient across pulmonary valve was 88 mm Hg (Fig. 2) and pulmonary annulus was 8 mm (Fig. 3) The ductus arteriosus was patent and measured 2.5 mm in diameter. A

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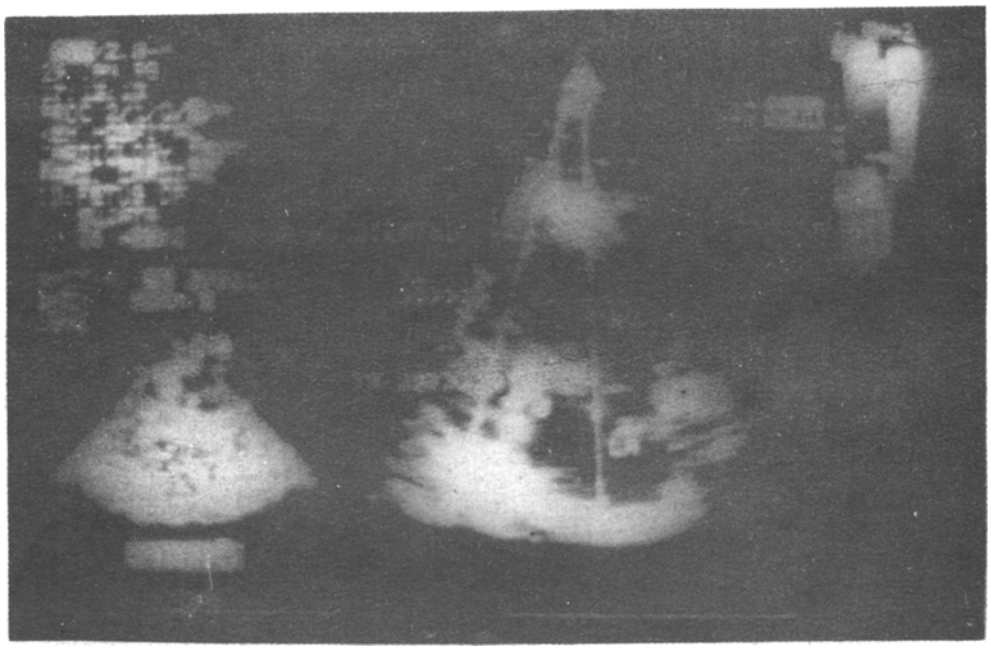

Fig 1. Four chamber view of fetal echocardiography showing dilated right atrium [RA], right ventricle [RV] and tricuspid regurgitation [TR jet].

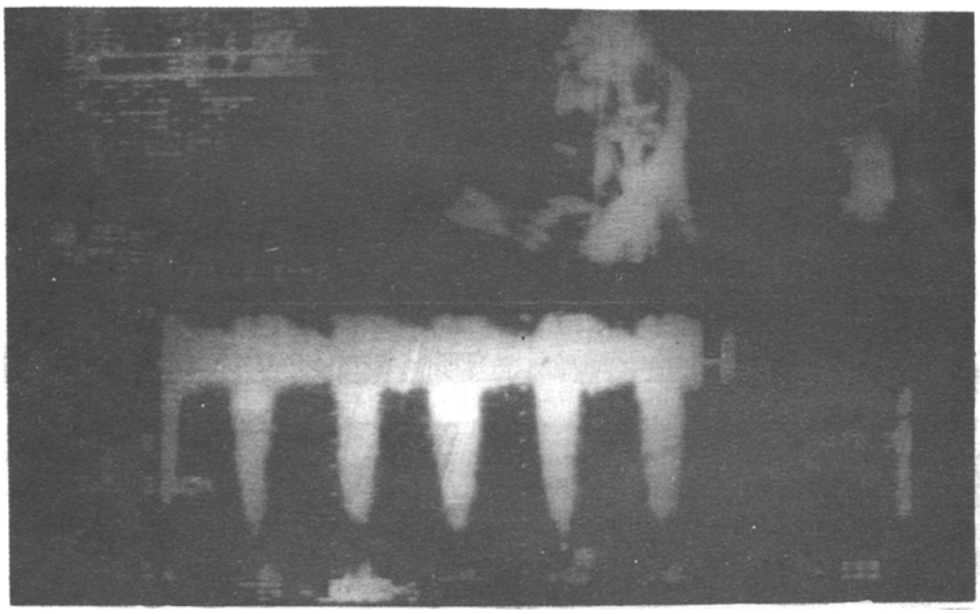

Fig 2. Short axis view of transthoracic echocardiography showing pulmonary valve [PV] and gradient across right ventricular outflow tract [ROVT] by Doppler velocity

Fig 3. Short axis view of trans-thoracic echocardiography showing doming pulmonary valve[PV] and pulmonary valve annulus.

transcatheter balloon dilatation of pulmonary valve was done on the fourth day of life, under general anesthesia. Vascular access was obtained via percutaneous puncture of the femoral vessels. A 3 French(F) valved introducer sheath in the left femoral artery was used to monitor the blood pressure and to obtain samples for blood gas analysis. A 5 F valved introducer sheath was used for venous access through the right femoral vein. Intravenous heparin was administered in a bolus dose of 100 units per kilogram body weight. A 5 F multipurpose catheter was passed into RV. The RV systolic pressure was 88 mmHg. and the end-diastolic pressure was 12 mmHg. A 0.025' hydrophilic straight tipped glide wire (Terumo, Inc, Tokyo, Japan.) was passed through the multipurpose catheter and manipulated to cross the pulmonary valve. The pulmonary artery pressure was 24/18/20 mmHg. A 0.025' exchange length wire was used to position a balloon dilatation catheter (Tyshak II Pediatric dilataion catheter. Numed Inc. Ontario, Canada) across the PV. The balloon had an outer diameter of 10 mm and was 3 mm long. The shaft size of the balloon catheter was 5 F. The pulmonary valve was dilated with a single inflation (Fig 4). Post dilatation PA pressure recorded using a multipurpose catheter was 50/34/38 mmHg. A continuous pull-back gradient from the PA to the RV showed a residual peak systolic gradient of 4 mmHg. The RV systolic pressure had dropped to 54 mmHg. and the end-diastolic pressure also decreased to 4 mmHg. Since the patent ductus arteriosus was not large, it was decided not to tackle it in the same sitting. The total procedure time was

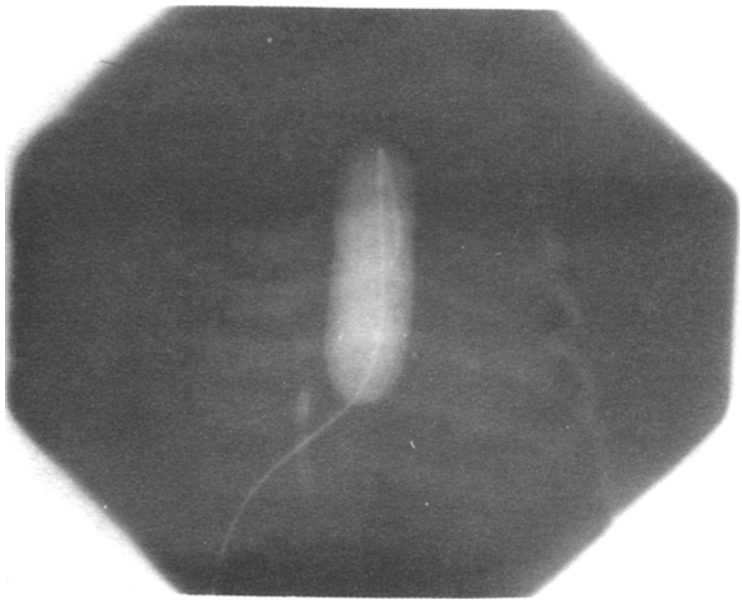

Fig 4. Inflated balloon across pulmonary valve showing total disappearance of the waist of the balloon.

60 minutes with a fluoroscopic time of 8 minutes. There was no procedural complication.

Follow-up echocardiograms on days 2 and 10 after the procedure showed good relief of PS, improvement in the RV contractility and a reduction in the size of the RA and RV. The peak systolic gradient across the PV was 15 mmHg.The size of the PDA remained the same and the LV did not show any volume-overload. The child was discharged from the hospital on day 10. At one month follow-up, the child was asymptomatic. The echocardiogram showed further reduction in the size of the RA and RV. There was no LV dilatation and both the RV and LV showed good contractility. It was planned to close the PDA by percutaneous transcatheter coil occlusion at a later date.

DISCUSSION

Severe valvar pulmonic stenosis presents as neonatal RV failure and has a poor prognosis unless it is rapidly diagnosed and corrected[6]. Surgical pulmonic valvotomy is possible but has a high procedural and intra-hospital mortality[7]. Transcatheter balloon dilatation has been described[4,5] and promises better results compared to surgery. The mortality rate is 6% as compared to 22% in surgery[7]. Fetal echocardiography can diagnose this critical condition prenatally and helps the obstetricians, neonatologists and cardiologists in the management of the neonate with severe PS. The interventional procedure was well planned and the necessary hardware arranged before the procedure. The PDA was not closed at the same sitting because it did

not cause any significant haemodynamic compromise. A transcatheter coil occlusion of the PDA was planned at a later date. One case of neonatal pulmonary valve dilatation not day 2 has been reported in twin-twin transfusion syndrome (TTTS) by Zosmer *et al*[8]. To the best of the authors' knowledge, this is the first case of pre-natal diagnosis of a congenital heart disease followed by successful neonatal intervention from India.

REFERENCES

1. Martin GR, Ruckman RN Fetal echocardiography : a large clinical experience and follow-up. *J Am Soc Echocardiogr.* 1990; 3 (1) : 4-8.
2. Castor S, Fouron JC, Teyssier G, Sonnesson SE, Chartrand C. Skoll A. Assessment of fetal pulmonic stenosis by ultrasonography. *J Am Soc Echocardiogr.* 1996; 9 (6) : 805-813.
3. Heart disease in infants, children, and adolescents including the fetus and young adult (5 edn). Vol II : Moss and Adams; Williams and Wilkins, Baltimore, Maryland 21202 USA.
4. Gildein HP, Kleinert S, Goh TH, Wilkinson JL. Treatment of critical pulmonary valve stenosis by balloon dilatation in the neonate. *Am Heart J* 1996; 131 : 1007-1011.
5. P. Syamasundar Rao. Balloon valvuloplasty in the neonate with critical pulmonary stenosis. *J Am Coll Cardiol* 1996; 27 : 479-480.
6. Coles JG, Freedom RM, Olley PM, Coceani F, Williams WG, Trusler GA. Surgical management of critical pulmonary stenosis in the neonate. *Ann Thorac Surg* 1984; 38 : 458-465.
7. Hanley FL, Sade RM, Freedom RM, Balckstone EH, Kirklin JW. Outcome in critically ill neonates with pulmonary stenosis and intact IVS : A multi-institutional study. *J Am Coll Cardioll* 1993; 22 : 183-192.
8. Nurit Zosmer, Rekha Bajoria, Ehud Weiner *et al.* Clinical and echographic features of in utero cardiac dysfunction in the recipient twin in twin-twin transfusion syndrome. *Br Heart J* 1994; 72 : 74-79.