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Pulmonary Atresia with Intact Ventricular Septum and Large Right ventricle as a Cause of Neonatal Heart Failure

R.P. Singh
A. Narang
C.K. Banerjee
P.S. Bidwai

Pulmonary atresia with intact ventricular septum is an uncommon but serious cardiac malformation and constitutes upto 2.5% of all structural cardiac defects(1). It is usually associated with a hypoplastic right ventricular cavity, however cases with large right ventricular cavity have been reported in literature and these cases have been classified into Type I and Type II depending upon the right ventricular cavity size(2-4). Type II cases with a normal or dilated right ventricular cavity constitute about 15-20% cases of pulmonary atresia with intact ventricular septum(5-9) and may be associated with Ebstein like anomaly of tricuspid valve(6) or even absent tricuspid valve leaflets(7).

We report a rare case of pulmonary atresia with large right ventricular cavity causing heart failure in the neonatal period.

From the Departments of Pediatrics, Pathology and Cardiology, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

Reprint requests: Dr. A. Narang, Additional Professor, Department of Pediatrics, PGIMER, Chandigarh 160 012.

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Case Report

A 1840 g, 36 weeks gestation baby was born to 3rd gravida with uneventful pregnancy. At 12 hours age, the baby was found to have acrocyanosis, and a soft blowing pansystolic murmur in the parasternal area with single second heart sound. Although there were no signs of congestive heart failure at that time, at 21 hours of age, the child developed frank signs of hypoxemia and heart failure with central cyanosis, tachypnea, hepatomegaly and prominent systolic murmur suggestive of tricuspid regurgitation.

Investigations revealed a massive cardiomegaly (with CT ratio of 0.9) on chest skiagram and the ECG revealed a terminal qRs vector pointing superiorly with a counter clockwise loop. Right sided chest leads were bizzare with terminal portion of qRs widely separated from the initial portion: the P waves were normal. There was no suggestion of left ventricular septum, very large right ventricular cavity with dilated tricuspid ring. The pulmonary valve was atretic with small pulmonary artery. The left ventricular cavity, aortic valve and mitral valve were normal.

The baby was given medical management for heart failure but showed progressive worsening and died at the age of 48 hours. On autopsy, the heart was found to occupy whole of the thoracic cage. The anterior surface of the heart was formed mainly by the right ventricle. The right ventricle was markedly dilated with papery thin walls and dilation of right atrium and tricuspid ring. Interventricular septum was intact with a large interatrial communication. Pulmonary valve was atretic with hypoplastic pulmonary arteries. The left side was normal and there was venous congestion of the liver.

Discussion

Pulmonary atresia with intact ventricular septum generally presents with cyanosis and heart failure in the first week of life(4). Cyanosis occurs soon after birth due to poor pulmonary blood flow and progressive worsening with ductal closure(1-4). Presence of massive cardiomegaly, tricuspid regurgitation murmur and ECG showing right axis deviation with right atrial enlargement and right ventricualr hypertrophy suggests Type II pulmonary atresia whereas patients with Type I abnormality have mild cardiac enlargement and ECG shows left ventricular dominance and right atrial enlargement. Although, our patient had classical presentation, ECG was not characteristic possibly due to very early onset of symptomatology. However, echocardiography confirmed the diagnosis antemortem.

The mortality in pulmonary atresia with intact ventricular septum without surgical intervention is 100% and a majority of deaths occur in the neonatal period(8). The optimal surgical approach is pulmonary valvotomy with or followed by systemic to pulmonary artery shunt and atrial septostomy. With improving technology, survival rates following surgery have been reported around 50%(8,9). Our patient though diagnosed in time, could not be offered any surgery.

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Antenatal Diagnosis of Acrania

K. Balakumar

Sonography with its capability to diagnoses lethal fetal anomalies at an early antenatal age, has radically changed the management policies in pregnancy with fetal abnormality. A case of acrania diagnosed by ultrasound at thirteenth week of gestation, confirmed subsequently by follow up scan and the delivered fetus is reported.

Case Report

A 23-year-old seventh gravida (consanguineous marriage) was referred for sonographic examination at thirteenth week of gestation. She gave a history of six consecutive abortions with no detectable cause; all occurring in the late third trimester except for one which aborted in the fifth month and was a case of anencephaly.

Sonogram revealed a near normal sac volume and the fetus was active. The crown-rump length was less (48 mm) due to the small cephalic region representing the brain. Neither the midline structures nor the ventricles were visualized at this time, probably because of the distortion produced by the absence of the calvaria. The usually echoic skull contour was absent (*Fig. 1*).

Though the diagnosis of acrania was made, she was reassessed at 16 weeks of gestation for confirmation. At this time (*Fig. 2*), the brain volume was corresponding to the gestational age but without skull bones. There was a thin membrane over it. The bones forming the base of skull and the face were visualized and the other definable bones were also within the normal limits. No associated significant gross abnormality was detected on repeated attempts.

Therapeutic abortion was performed which confirmed the diagnosis of acrania

From the P.V.S. Cancer Research Centre, Calicut 2, Kerala.

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