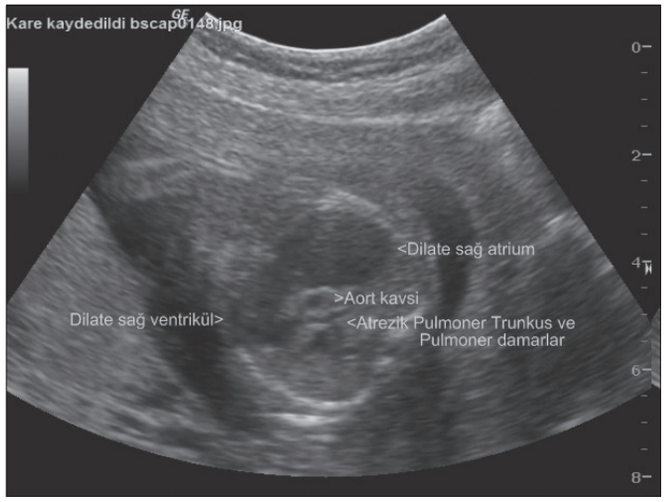
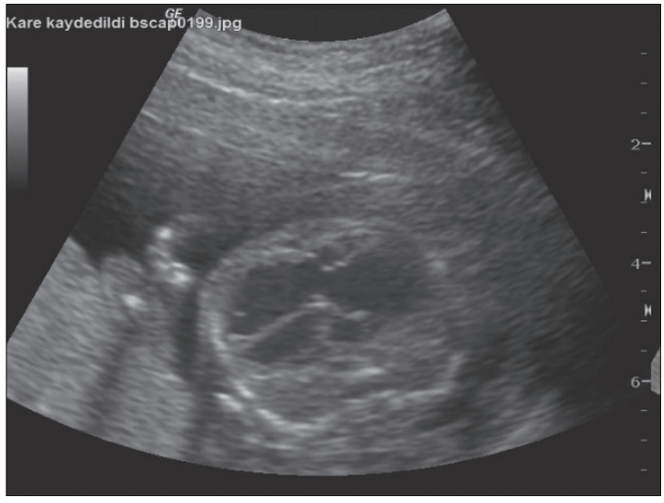
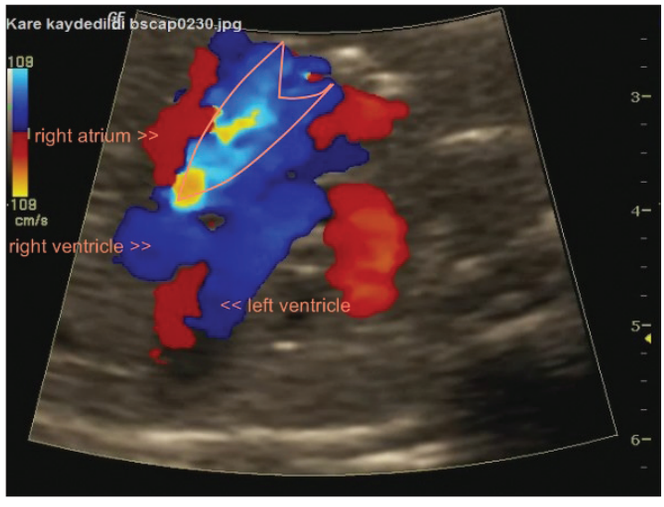
What is your diagnosis ?







Address for Correspondence / Yazışma Adresi: *Özlem Özdeğirmenci, M.D., Etlik Zubeyde Hanim Women's Health Teaching and Research Hospital, Etlik, Ankara, Turkey*
Phone: +90 312 221 38 99 Mobile: +90 532 721 62 41 e.mail: ozlemerman@gmail.com
doi:10.5152/jtgga.2010.016

Answer

Pulmonary atresia (PA) is a congenital malformation of the pulmonary valve in which the valve fails to develop. The incidence of PA with IVS is 4.5 per 100.000 live births (1). It is associated with a broad spectrum of anatomic variability. PA can occur either with an intact ventricular septum (IVS) or with ventricular septal defect.

Two types of pulmonary atresia with an intact ventricular septum have been described (2). In type I disease, a combination of pulmonary valvular atresia, competence of the tricuspid valve, and an intact ventricular septum result in massive right ventricular hypertrophy and chamber obliteration with suprasystemic pressures. In type II disease, proximal pulmonary arterial atresia and an intact ventricular septum are present, but tricuspid incompetence allows retrograde flow of blood from the right ventricle into the right atrium and across an atrial septal defect. Thus, in type II disease, the right ventricle is either normal or dilated.

Demonstration of reversal of blood flow through the ductus is virtually always present in pulmonary atresia with an intact ventricular septum. The tricuspid valve orifice is usually proportional to the size of the right ventricular cavity. The valve leaflets are often dysplastic, which results in varying degrees of insufficiency or stenosis. A dilated right atrium is another finding commonly associated with this disorder. The degree of dilation is dependent on the amount of tricuspid regurgitation present or the inability of blood to enter the right ventricle.

The outcome for infants born with pulmonary atresia and an intact ventricular septum is poor. They are dependent on a patent ductus arteriosus for survival in the neonatal period. Administration of prostaglandin E1 at birth is essential to maintain patency until surgery can be performed (3, 4).

In the presented ultrasound image of a fetus at 18 weeks of gestation, pulmonary atresia with intact ventricular septum is visualized. Imaging demonstrated marked dilatations of the right atrium and ventricle. A small pulmonary artery was seen with an atretic pulmonary valve. The tricuspid valve was present and tricuspid insufficiency was confirmed by color Doppler indicated by an arrow in the figure.

Özlem Özdeğirmenci, Metin Kaplan
Etlik Zübeyde Hanım Women's Health Teaching and Research Hospital, Ankara, Turkey

References

1. Daubeney PE, Sharland GK, Cook AC, Keeton BR, Anderson RH, Webber SA. Pulmonary atresia with intact ventricular septum: impact of fetal echocardiography on incidence at birth and postnatal outcome. UK and Eire collaborative study of pulmonary atresia with intact ventricular septum. Circulation 1998; 98: 562-6.
2. Davignon AL, Greenwold WE, Dushane JW, Edwards JE. Congenital pulmonary atresia with intact ventricular septum: Clinicopathologic correlation of two anatomic types. Am Heart J 1961; 62: 591-602.
3. Kelekci S, Kılavuz O, Yilmaz B, Cankaya A. Hypoplastic right ventricle with intact ventricular septum: A case report. Türkiye Klinikleri J Gynecol Obst 2005; 15: 325-7.
4. Barrett HS. Hypoplasia of the right ventricle. In: Drose JA, ed. Fetal Echocardiography. Saunders 2010. p.145-56.