J Obstet Gynecol India Vol. 56, No. 1 : January/February 2006 Pg 76-78

CASE REPORT

The Journal of Obstetrics and Gynecology of India

Hypoplastic left heart syndrome

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Key words : fetal echocardiography, hypoplastic left heart syndrome, prenatal diagnosis

Introduction

The most common major congenital anomalies are cardiovascular ones, which occur in approximately 8 of 1000 births. Congenital cardiovascular malformations account for approximately 20% of neonatal deaths and 50% of infant deaths, and are seen four to five times more frequently in stillbirths than in live-born babies [1]. After a three-year study that included 875 fetal echocardiograms, Macedo et al [2] found the rate of congenital heart disease as 4.2% and the prevalence of hypoplastic left heart syndrome as 13.5% in this affected group. The etiology of hypoplastic left heart syndrome is not precisely known. Autosomal recessive, autosomal dominant and polygenic inheritances have been suggested. However multifactorial is the more likely form of transmission [3]. Because of high perinatal mortality, prenatal diagnosis with good counseling is mandatory. We present a hypoplastic left heart syndrome case, which we diagnosed in labor room because of fetal tachy-arrythmia.

Case report

A 34 year old, G3P1, pregnant woman with a history of previous cesarean delivery for breech presentation was admitted to labor ward in active labor. The baby was living and healthy. She had a stillbirth at 21 weeks in her second pregnancy. During this pregnancy cardiotocographic abnormalities were diagnosed (Figure 1) and ultrasound examination revealed dilated right ventricle and atrium, mitral atresia, hypoplasia of aorta, dilated main pulmonary artery (Figures 2, 3, 4), and reverse excursion of foramen ovale flap. Hypoplastic left heart syndrome was diagnosed by doppler ultrasound. After counseling the parents opted for vaginal delivery which was uneventful. A female baby weighing 2820 g was born with apgar scores of 6 and 8 at 1 and 5 minutes respectively. Prenatal diagnosis was confirmed by neonatal echocardiography. We did not have a neonatal cardiovascular surgery center. Seven days after birth, the baby was lost due to cardiopulmonary failure. The parents refused autopsy on the fetus.

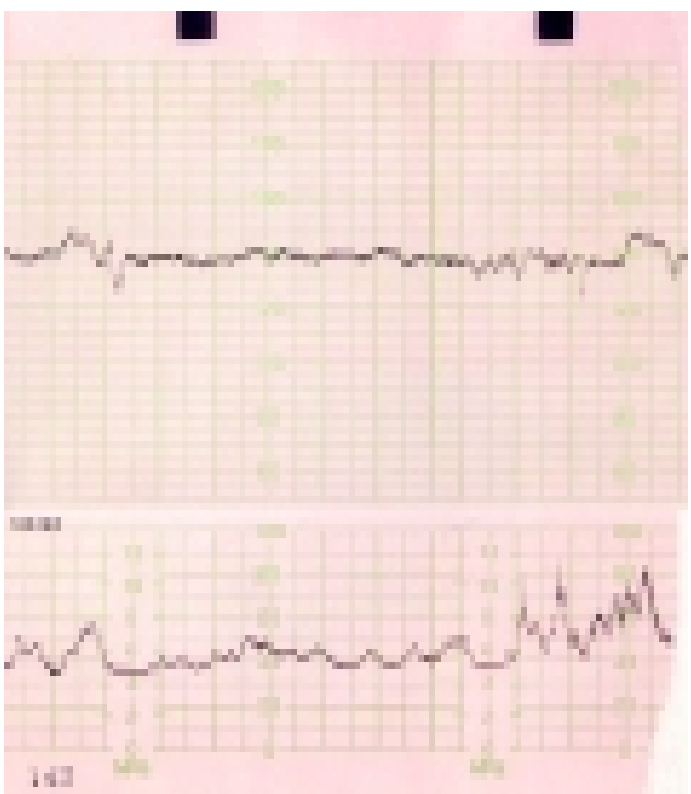

Figure 1. Abnormal carditocography.

Paper received on 30/06/2004 ; accepted on 09/09/2004

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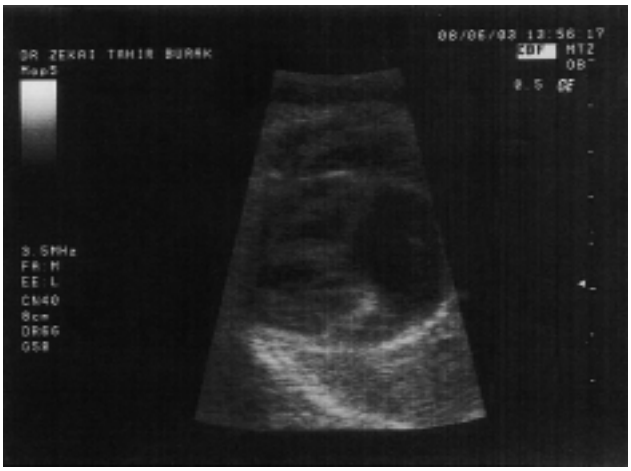

Figure 2. A small, thick-walled and hyperechoic left ventricle and enlarged right ventricle and atria.

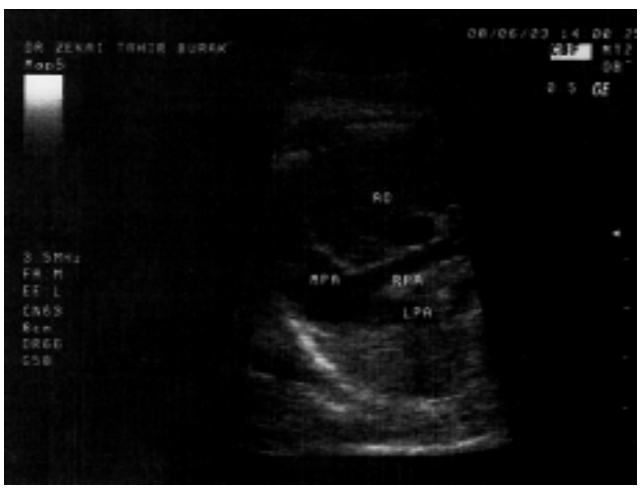

Figure 3. Dilated main pulmonary artery trunk and its branches, and the hypoplastic aortic root.

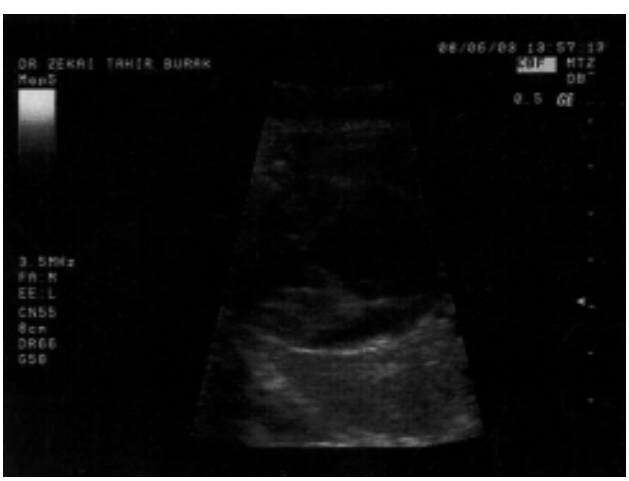

Figure 4. Right ventricle - is hypertrophied and dilated, and its infundibular and septal architecture are exaggerated by the hypertrophy

Discussion

Hypoplastic left heart syndrome can be detected by fetal ultrasonography with four-chamber view. The sensitivity of sonographic detection for isolated left heart syndrome is reported as 61.9% in a study where in cardiac defects affecting the size of the ventricles had the highest detection rate [4]. In another study, sensitivity rates for prenatal sonographic diagnosis is reported to be 36.6% to 37% [5]. The prenatal diagnostic approach is important for appropriate counseling, gives time to consider treatment options, and is associated with fewer adverse perioperative neurologic events. It is also important for preventing ductal shock (by giving prostaglandin E_1 on time) and keeping the patients' preoperative condition good [6]. However, the mortality rate does not differ between those who have been diagnosed prenatally and those diagnosed postnatally [7].

Aside from the cardiac anomalies, extra-cardiac defects are frequently seen associated with hypoplastic left heart, and the most common are two-vessel umbilical cord, and craniofacial, gastrointestinal, genitourinary and central nervous system abnormalities [3]. The risk of aneuploidy associated with fetal cardiac anomalies is much greater (ranging from 13 to 32%) than that associated with advanced maternal age [1]. A karyotype analysis should be offered to pregnant women having a baby of hypoplastic left heart syndrome and a detailed ultrasonographic scan should be done to rule out extra-cardiac anomalies.

The hypoplastic left heart syndrome usually presents during the first week of life with signs of low systemic perfusion secondary to constriction of the ductus arteriosus due to the decrease in pulmonary vascular resistance. These babies usually tolerate their defect for a few days while the ductus remains widely open. When the ductus constricts arterial pressure decreases and a severe metabolic acidemia develops [8]. Hypoplastic left heart syndrome is responsible for 25% of cardiac deaths in the first week of life. Almost all of the affected infants die within 6 weeks if they are not treated. Several palliative procedures including atrial septectomy, banding of the pulmonary artery, and creation of aortopulmonary shunt have been used hoping for a better prognosis [9,10].

In the management of hypoplastic left heart syndrome, apart from determining the karyotype and investigating of associated anomalies, no obstetrical interventions are needed during pregnancy [11]. Prenatal diagnosis is important for pregnancy counseling and for planning the delivery, which is of particular relevance in fetuses with hypoplastic left heart syndrome, due to the severity of this condition and the specialized surgical treatment that is required.

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