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Antenatal Diagnosis of Congenital High Airway Obstruction Syndrome.

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ABSTRACT

Congenital high airway obstruction syndrome is a rare but near fatal condition of multifactorial inheritance and so need antenatal diagnosis. The fetus show typical findings such as a dilated, fluid filled trachea, enlarged hyperinflated echogenic lungs, inverted or flattened diaphragm, squeezed heart and ascites. It is the result of congenital obstruction of the airway secondary to laryngeal atresia, tracheal atresia or a laryngeal cyst.
Keywords: CHAOS- Congenital high airway obstruction syndrome, CCAM- congenital cystic adenomatoid malformation.

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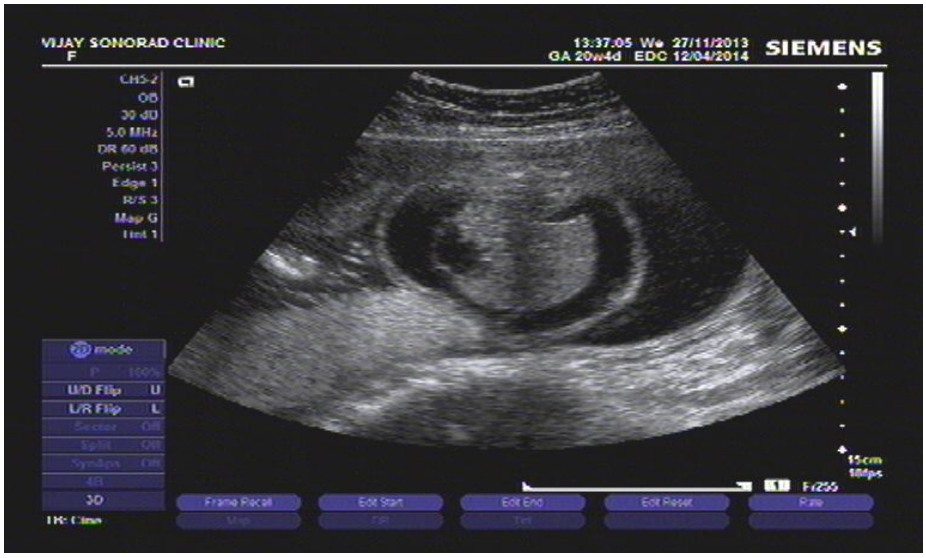
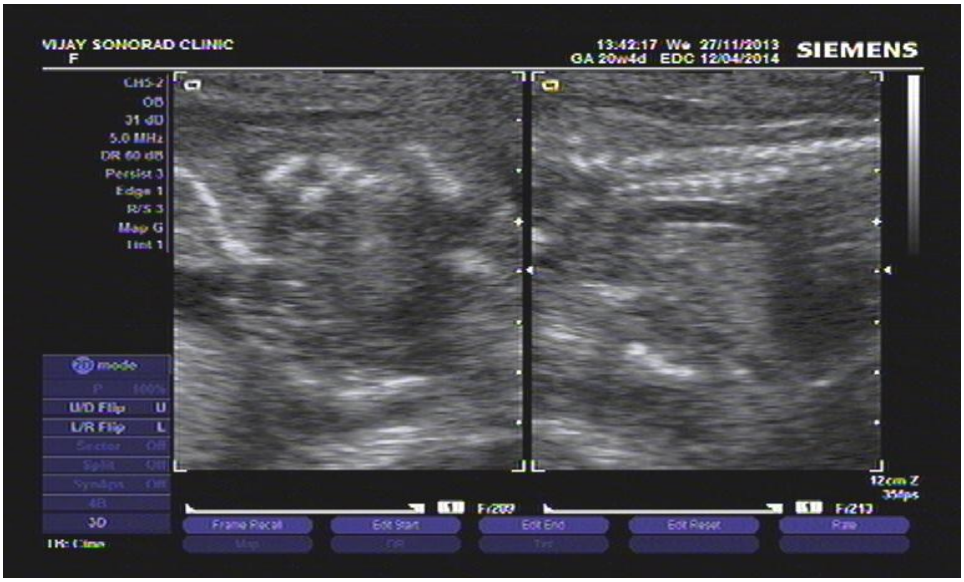
CASE REPORT

21 years old primigravida came for anomaly scan at 20 wks. Her previous sonography was done at 9 wks which was unremarkable. There was no history of consanguinity. On examination, lateral ventricles were dilated (width-17 mm). There was no spinal, facial or limb anomaly. Lungs were echogenic and over expanded with inverted domes of diaphragm and tubular compressed heart. Trachea and bronchi were filled with fluid. Evidence of ascites with cystic structures was seen in it. No other organs were identifiable except liver. Amniotic fluid was excess. Diagnosis of Congenital high airway obstruction syndrome was made.



Transverse image through abdomen showing ascites



Transverse and sagital view through chest showing dilated and fluid filled trachea

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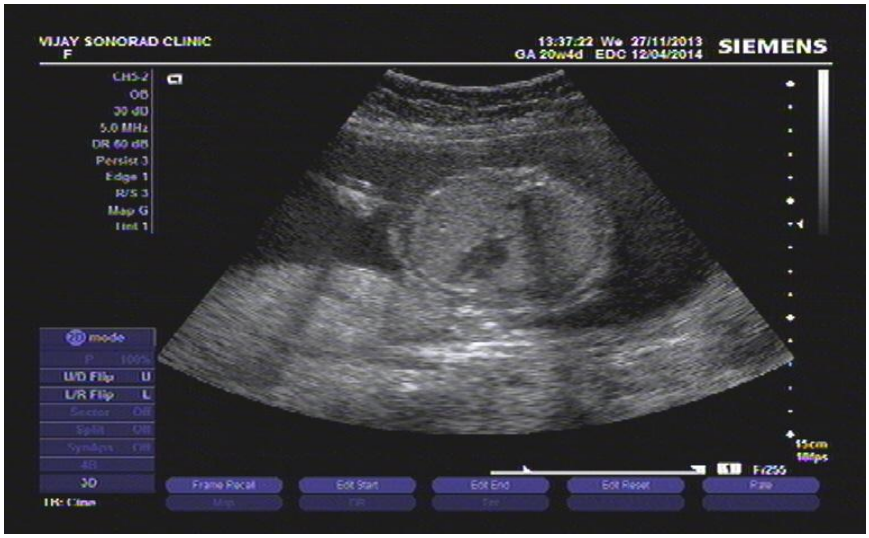
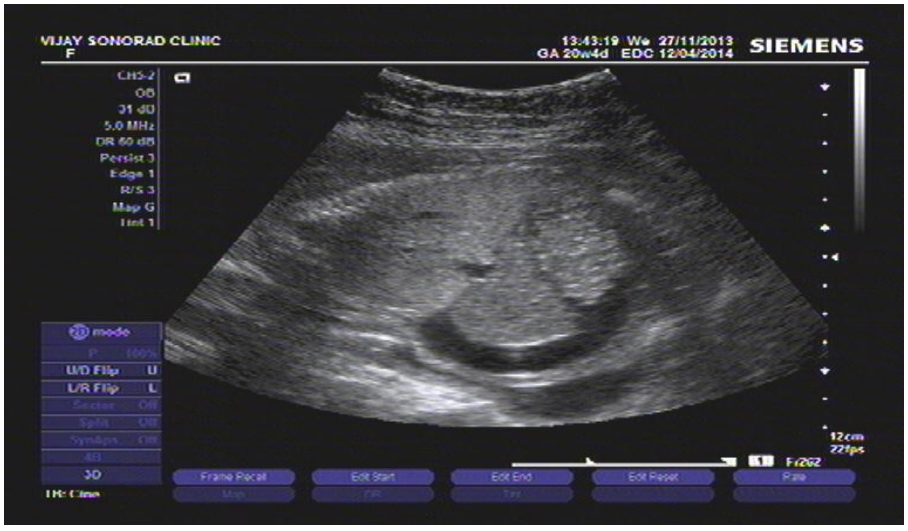
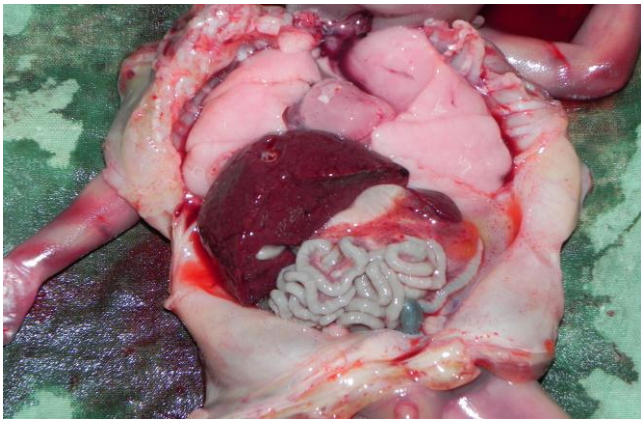


Transverse image through chest showing echogenic lungs with compressed heart



Sagital view through lower chest and upper abdomen show over expanded lung with inverted dome of diaphragm



Gross appearance of thorax and abdomen show enlarged both lungs with costal impression on the external surface.

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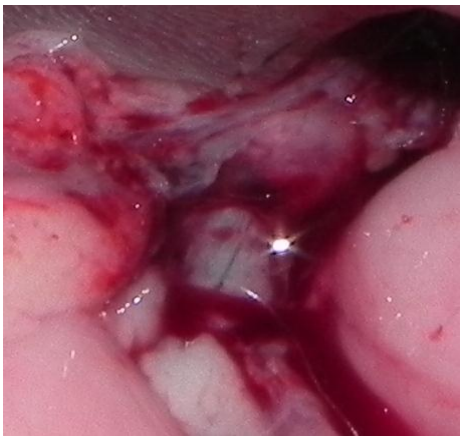

Hypertrophied cricoids

After explanation of unfavorable outcome of pregnancy, the parents opted for termination of the pregnancy. The aborted fetus showed distended thorax and abdomen. The abdominal cavity showed a large amount of clear yellow fluid. An intrathoracic examination showed the enlargement of the right and the left lungs. The larynx showed complete obstruction at the infraglottic level caused by the overgrowth of the cricoid cartilage. Autopsy findings were compatible with laryngeal atresia type II described by Smith and Bain in 1965

Due to non-affordability of parents, karyotyping was not done. And due to reluctance of relatives block dissection was not possible.

DISCUSSION

Laryngeal atresia is a rare congenital malformation and is usually fatal. Prenatal diagnosis of CHAOS was possible as early as 15 weeks of gestation [1]. The malformation is caused by non development of the 6^{th} branchial arch during normal embryological development [2]. Smith and Bain have classified laryngeal atresia into three types: type 1, in which there is complete atresia of the larynx with midline fusion of the arytenoid cartilages and intrinsic muscles; type 2, in which there is infraglottic obstruction that is characterized by a dome-shaped cricoid cartilage obstructing the lumen; and type 3, in which there is occlusion of the anterior fibrous membrane and fusion of the arytenoid cartilages at the level of the vocal processes [2]. Many etiologies were proposed including laryngeal or tracheal webs, laryngeal cysts, tracheal atresia, subglottic stenosis or atresia, laryngeal or tracheal agenesis. Most common cause is laryngeal atresia [3].

This malformation is generally fatal; however, there are reports of a few cases that have been successfully treated with neonatal interventions such as Ex Utero Intrapartum Treatment (EXIT procedure) and fetoscopic laser decompression, while the fetus is still connected to the placenta [4]. The EXIT procedure is a technique for safely managing airway obstruction at birth, in which placental support is maintained until the airway is evaluated and secured. Several successful cases have been reported [5, 6].

The sonographic findings of CHAOS include increased in lung size and echogenicity; fluid-filled, dilated trachea, fetal hydrops, and polyhydramnios. Antenatal USG shows enlarged hyperechoic lungs, a dilated tracheobronchial tree, ascites, and an inverted or flattened diaphragm [7]. In laryngeal atresia, the trachea is dilated because of nonclearance of fluid (which is normally secreted by the lungs). In high airway obstruction, the nonclearance of fluid from the lungs results in parenchymal hyperplasia, which is apparent on USG as enlarged hyperechoic lungs. An enlarged lung causes compression of the great veins and the right atrium, and this leads to ascites [9, 10].

The findings were difficult to differentiate with congenital cystic adenomatoid malformation (CCAM) type III. However, bilateral CCAM was very rare; therefore, the diagnosis of CHAOS was considered. Laryngeal atresia

may be associated with other structural and genetic abnormalities such as left persistent superior vena cava, single umbilical artery, abnormal fingers and toes, esophageal atresia, or renal agenesis, partial trisomy 9 and 16, chromosome 5p [11, 12]. The most common associated genetic disorder with CHAOS is Fraser's syndrome which is inherited by autosomal recessive form and characterized by urogenital defects, laryngeal atresia, syndactyly, and cryptophthalmos [8].

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