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Prenatal sonographic diagnosis of upper airway obstruction: a challenge that can be achieved

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Abstract
Congenital upper airway obstruction presents a life-threatening situation when encountered in the delivery room. We present a case in which *in utero* diagnosis of this malformation was suspected, but unfortunately the neonate did not survive due to the inability to provide adequate ventilation. The prenatal sonographic features of this rare malformation are outlined, and the literature reviewed.

Key words: Ultrasonography, prenatal; Airway obstruction

Introduction

The birth of a baby with upper-airway obstruction is one of the most stressful events a paediatrician faces in the delivery room, since it requires immediate, and usually surgical, opening of the airway. The true incidence of this malformation is unknown, but is considered very rare.

Prenatal ultrasonography has provided the possibility of *in utero* diagnosis of this rare malformation. The first prenatal diagnosis of laryngeal atresia was reported in the Japanese literature by Arizawa *et al.* in 1989. Since then, only a few cases have been added where a prenatal diagnosis was made and confirmed by autopsy (Watson *et al.*, 1990; Furness *et al.*, 1991; Choong *et al.*, 1992; Dolkart *et al.*, 1992; Richards *et al.*, 1992; Weston *et al.*, 1992; Meagher *et al.*, 1993; de Hullus *et al.*, 1995). Although prenatal diagnosis is feasible, the majority of the foetuses die shortly after birth.

We present a case in which upper airway obstruction wsa suspected *in utero* after bilateral echogenic lung and ascites were diagnosed at 22 weeks' gestation. Our goal is to draw attention to the complexity of this situation and the crucial need for a multidisciplinary setting for delivery of the foetus.

Case report

A 29-year-old gravida 4 para 2 was referred to our high-risk pregnancy unit in the 23rd week of gestation for evaluation of foetal ascites detected by routine ultrasound examination. The patient had two healthy children delivered by caesarean sections because of foetal distress. Her present pregnancy was uneventful until the 22nd week, when severe foetal ascites was detected. Blood type was A positive, maternal serum antibody titre for cytomegalovirus, toxoplasma, herpes smplex, and syphilis were negative. The foetus had a normal 46XY karyotype.

Ultrasound examination in our unit showed a severely hydropic fetus with enlarged 'white' (Figure 1) lungs compressing what appeared to be a structurally normal heart, and polyhydramnios. The trachea ended in a blind loop, and communication between the upper airway tract (larynx) and the trachea could not be demonstrated (Figure 2a and b).

In the 26th gestational week spontaneous rupture of the membranes necessitated an emergency caesarean section. The neonate was severely hydropic with bradycardia, and and several attempts to intubate him failed due to inability to bypass the vocal folds. The baby died shortly after birth. In accordance with the family's request, autopsy was not performed.

Discussion

Upper airway obstruction (UAO) consists of two major entities: laryngeal and tracheal atresia, both requiring prompt neonatal treatment to ensure survival. This malformation can occur as a single, or a part of a more complex genetic disorder (Schauer *et al.*, 1990; Moerman *et*

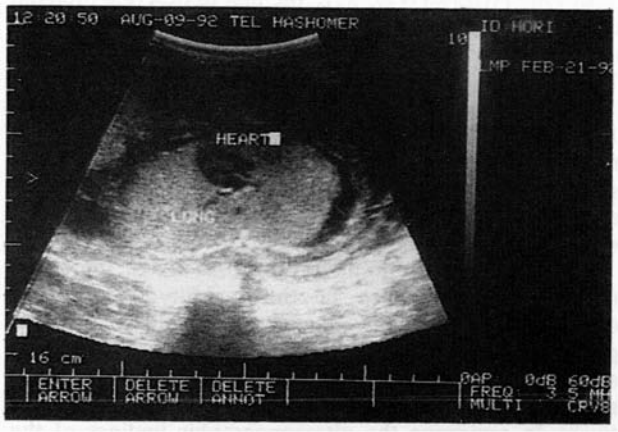

FIG. 1
Transverse section of the foetal chest shows massive lung congestion causing compression of the heart.

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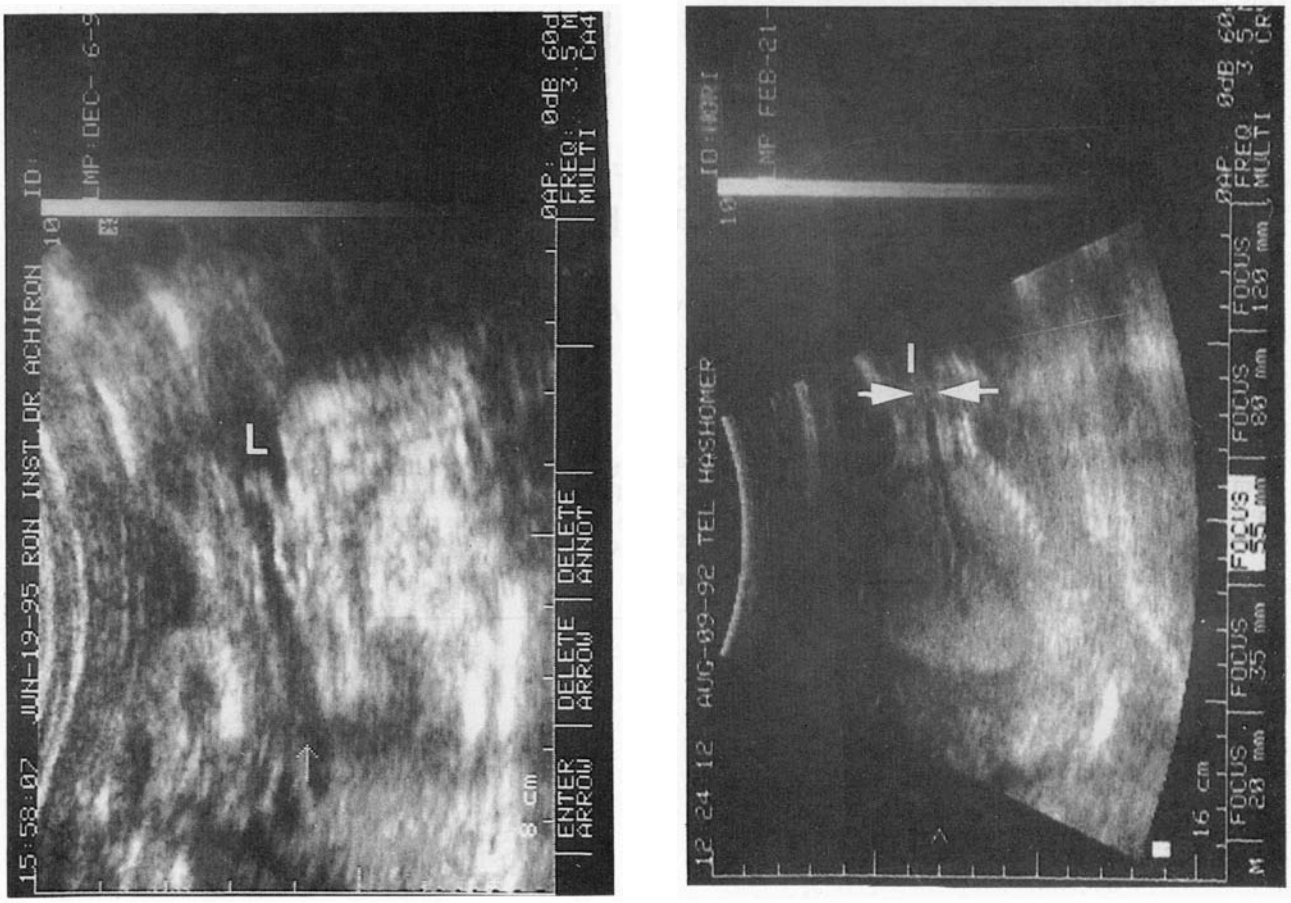

(A) (B)

FIG. 2

Coronal ultrasonographic sections of foetal upper airway. A) Normal appearance of the larynx (L) communicating through the epiglottis with the trachea (arrow head). B) Direct visualization of upper airway obstruction manifested by absence of communication (arrow heads) between the larynx (l) and trachea.

al., 1992). The primary insult to the upper airway is thought to be either a vascular one occurring in the fourth to sixth week of pregnancy, or a defect in the recanalization process in the ninth week (Holinger, 1976). The majority of reported cases are from the neonatal or paediatric literature and in most, the diagnosis was made only after birth (Cotton and Richardson, 1981).

In 1989 Arizawa *et al.* were the first to describe prenatal diagnosis of UAO. Since that time, nine new additional cases of *in utero* diagnosis were reported (de Hullu *et al.*, 1995).

Prenatal diagnosis of UAO has been made in a total of 10 cases. In all cases the pregnant women were referred for further ultrasound evaluation due to the appearance of second-trimester foetal ascites, polyhydramnios, hydrops and echogenic lungs. Three cases were terminated, and seven were followed-up.

However, despite prenatal diagnosis, six of the seven neonates died because of the inability to pass an endotracheal tube. Only one foetus benefited from the prenatal diagnosis in early neonatal setting with immediate surgical intervention. Therefore, it seems that in most reported cases the diagnosis was performed retrospectively. In reviewing the ultrasonographic findings of the above cases, a common marker has been presented in all reported cases. The bilateral echogenic or 'white lungs' seems to be a pathognomonic finding of this condition. This peculiar ultrasonographic appearance of foetal lungs is attributed to the overdistention of the alveoli filled with retained fluid and bronchial secretions (Choong *et al.*, 1992). The differential diagnosis of echogenic lungs is congenital cystic adenomatoid malformation (CCAM) type III. However, this is only rarely a bilateral disease and does not usually compress the heart, as was shown in most cases with airway obstruction. The prenatal distinction between UAO and CCAM is of great importance, since the first requires a special neonatal setting to perform immediate tracheotomy, while in the latter even spontaneous resolution *in utero* has been described (Meagher *et al.*, 1993). Therefore, every effort should be made to establish a correct *in utero* diagnosis. Detailed evaluation of the upper airways is now feasible and even nomograms have been established (Richards and Farah, 1994).

Moreover, colour Doppler technology may enhance demonstration of flow movement through the upper airways (Fox *et al.*, 1993; Isaacson and Birnholz, 1991). In our cases it was shown that a direct demonstration of the level of tracheal interruption was possible *in utero*. However, we are aware of the lack of histological correlation in the present case. Furthermore, we are also aware that diagnosis was only retrospectively established. However, it is understandable, because at the time of ultrasound examination in our case only two previous cases had been published in the world literature, and only one was in English. With expansion of our knowledge and current experience in *in utero* visualization of upper airways, we hope that more infants might be salvaged following prenatal diagnosis of UAO.

In summary, the visualization of enlarged, diffusely echogenic foetal lungs is a clue to detailed evaluation of

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the upper airways. Demonstration of laryngeal or tracheal obstruction is feasible and should promote an appropriate personnel setting around delivery that includes a neonatalogist and otolaryngologist.

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