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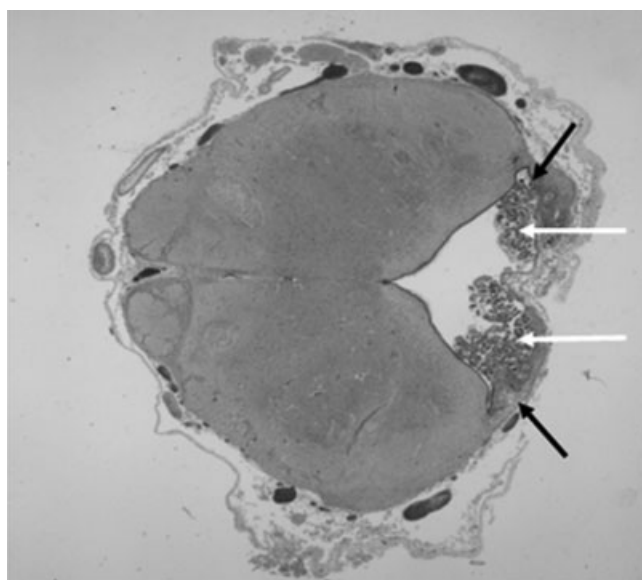
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**Figure 3** Axial pathological section of the lower part of the mesencephalon. Pathological examination confirmed the sonographic data, also showing remnants of the rhombic lips (black arrows) at the root of the choroid plexus (white arrows).

case of an affected 17-year-old boy attending a normal school, to which he cycles in spite of moderate ataxia, has been reported recently<sup>6</sup>. Because cerebellar agenesis occurs early during embryogenesis, the less severe clinical presentation may be explained by a cerebral compensation phenomenon leading to moderate manifestations despite a quasi constant impairment in motor development.

According to the algorithm proposed by Guibaud and des Portes for the anatomical classification of prenatal posterior fossa abnormalities, 'complete cerebellar agenesis' should be included in the section 'abnormal cerebellar anatomy' as an exceptional form of agenesis, which is limited to the vermis in the vast majority of cases<sup>7</sup>. Regarding the retrocerebellar fluid-filled space, postnatal imaging has shown an increased volume in a few cases<sup>1,6</sup>, but it can also be decreased as in our case, in which the anatomical space of the posterior fossa was occupied by the ptosis of the occipital lobe related to absence of the cerebelli tentorium.

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DOI: 10.1002/uog.6402

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## Early sonographic diagnosis of congenital high-airway obstruction syndrome

A 35-year-old primigravida underwent an initial routine anomaly scan at 15 weeks' gestation. She had no significant personal or family medical history. The pregnancy was dated by early sonography, and fetal biometry was consistent with the date of the last menstrual period. Chorionic villus sampling (at the woman's request) at 11 weeks revealed a normal male karyotype. Serology for cytomegalovirus was negative.

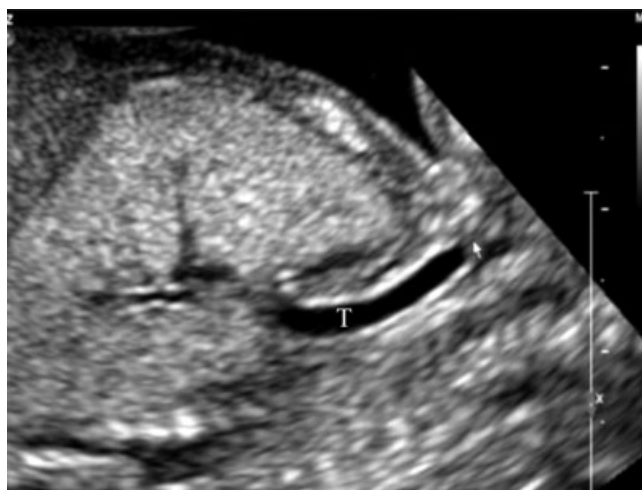
High-resolution transvaginal sonography (Philips IU, Bothell, WA, USA) revealed three major findings: bilateral enlarged hyperechogenic lungs, inverted diaphragm, and a greatly dilated tracheobronchial tree filled with hypoechogenic fluid. A sharp border was noted between the completely occluded upper larynxes and the dilated trachea (Figure 1). Together, the three signs provided direct evidence of the presence of congenital high-airway obstruction syndrome (CHAOS). This conclusion was supported by the curved orientation of the great vessels (aorta and inferior vena cava), as opposed to their normal linear anatomical position, across the thorax and abdominal cavity, an indirect sign of CHAOS. There were no other gross abnormalities. The couple was referred to a genetics specialist, and they opted to terminate the pregnancy owing to the severity of the malformation.

CHAOS is a rare clinical life-threatening condition caused by complete or near-complete obstruction of the fetal airway. Owing to advances in imaging techniques, the syndrome can readily be recognized prenatally. Since the initial report of its prenatal diagnosis in 1989 by Arizawa *et al.*<sup>1</sup>, at least 35 additional cases have been reported<sup>2</sup>. Our group also previously described one case of CHAOS diagnosed at 22 weeks' gestation<sup>3</sup>. A search of the Medline database from 1965 to August 2007 revealed that the present case is the earliest diagnosed to date. We attribute this finding to our routine use of high-resolution

**Table 1** Differential diagnosis of congenital high-airway obstruction syndrome (CHAOS)

| Type of pathology       | Time of diagnosis | Laterality                            | Resolution of abnormal sonographic findings | Associated early hydrops |
|-------------------------|-------------------|---------------------------------------|---------------------------------------------|--------------------------|
| CHAOS                   | Early             | Bilateral                             | Rarely                                      | Always                   |
| Type III CCAM           | Late/early        | Unilateral more common than bilateral | Rarely                                      | Rare                     |
| Pulmonary sequestration | Late/early        | Unilateral more common than bilateral | Rarely                                      | Rare                     |
| Diaphragmatic hernia    | Early/late        | Unilateral more common than bilateral | Never                                       | Rare                     |
| Mediastinal teratoma    | Late/early        | Unilateral more common than bilateral | Rarely                                      | Rare                     |

CCAM, congenital cystic adenomatoid malformation.



**Figure 1** Very large and highly distinct trachea (T) with a sharp border (arrow) between it and the completely occluded upper larynx (four parallel white lines).

transvaginal sonography during the first-trimester fetal anomaly scan.

The upper airway obstruction in CHAOS blocks the normal clearance of the fluids produced by the lungs through the larynx and leads to a number of structural changes that can be visualized sonographically in early pregnancy. The first is enlargement of the lungs, indicated by their abnormally high echogenicity relative to the kidneys. This finding is confirmed by the bilateral compression and flattening of the diaphragm and dilatation of the tracheobronchial tree which result directly from the lung expansion. All reported cases of CHAOS in the literature have also been associated with ascites<sup>4</sup>, which appears to develop very early<sup>5</sup>. The mechanism underlying the development of ascites probably involves kinking of the aorta and inferior vena cava, as noted in the present case.

The tracheal enlargement is specific to CHAOS and a key factor in differentiating the disorder from Type III bilateral congenital cystic adenomatoid malformation of the lungs, pulmonary sequestration, diaphragmatic hernia and mediastinal teratoma (Table 1).

In the present case, using high-resolution transvaginal sonography we were able to pinpoint the level of obstruction at the larynx. Others have described this

using the breathing movements of the fetus as an aid to locating the obstruction<sup>6</sup>, but this was precluded in our case by the early gestational age. Together with a normal tracheal size in early pregnancy, a finding of fluid flow across the trachea during fetal breathing later in pregnancy indicates a diagnosis other than CHAOS.

It is also important that clinicians examine fetuses with suspected CHAOS for Fraser syndrome, a multiorgan autosomal recessive disease. The major signs include renal agenesis, syndactyly, cryptophthalmos and ambiguous genitalia. In these cases, a finding of oligohydramnios is attributable to the renal agenesis. By contrast, in isolated CHAOS polyhydramnios is expected later in pregnancy when the enlarged lung mass obstructs the esophageal passage of amniotic fluid or the ascetic fluid compresses the stomach and intestine<sup>2,7</sup>.

Pulmonary enlargement detected in early pregnancy may spontaneously resolve later during gestation. However, this does not necessarily indicate resolution of the underlying pathology, although it may suggest only minor pharyngotracheal or laryngotracheal communications, which are associated with a less severe natural history. In these cases, a multidisciplinary approach is needed for delivery of the infant. The resolution of the sonographic signs in a fetus with CHAOS may be due to the development of a fistula that decompresses the airway<sup>3,8</sup>.

The early recognition of this rare malformation is important because it allows the option of termination of pregnancy to be provided earlier, with less risk of physical and psychological complications. The present report shows that with the transvaginal approach CHAOS can be diagnosed as early as 15 weeks' gestation, before the development of ascites.

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DOI: 10.1002/uog.6408

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### Conservative management of spontaneous heterotopic cervical pregnancy using an aspiration cannula and pediatric Foley catheter

Heterotopic cervical pregnancy is extremely rare, with most reported cases being associated with assisted reproductive technology<sup>1</sup>. To our knowledge, only two cases of spontaneous heterotopic cervical pregnancy have been reported in the English language literature<sup>2,3</sup>. We report the case of a 30-year-old woman referred at 8 weeks of gestation in her first pregnancy, which had been conceived spontaneously. Initial ultrasound examination revealed a heterotopic pregnancy with two gestational sacs, one intrauterine and the other in the upper part of the cervix, 21 mm from the external cervical os (Figure 1). A 2.0-cm fetal pole (8 weeks of gestation) with visible cardiac activity was observed in both gestational sacs. The woman and her partner were counseled on the potential complications of heterotopic pregnancy, and they decided to preserve her intrauterine pregnancy.

Under transabdominal ultrasound guidance (Accuvix XQ, Medison, Seoul, Korea), the cervical gestational sac was aspirated using a Karman cannula (4 mm in diameter) and a single-valve aspirator (Ipas, Chapel Hill, NC, USA) without cervical dilatation. Complete removal of the gestational sac was confirmed by sonographic demonstration of a smoothly echogenic lining of the cervical canal. However, active cervical bleeding was then observed. To minimize the risk of cervical injury, a 6-Fr pediatric Foley catheter was



**Figure 1** Transvaginal ultrasound image at 8 weeks of gestation showing a heterotopic pregnancy, consisting of one gestational sac located in the uterine fundus and the other within the uterine cervix.

inserted at the level of the implantation site, and 3 mL of saline was introduced to achieve hemostasis by compression. Bleeding stopped after compression and the catheter was removed the following day. On the second day after aspiration, ultrasound examination revealed an intracervical hematoma, measuring 3.0 cm × 3.0 cm (Figure 2).

Serial ultrasound examinations revealed that the intrauterine fetus showed appropriate growth, and that the intracervical hematoma decreased gradually and disappeared by 20 weeks of gestation. The remaining antenatal course was uneventful, and an elective Cesarean section was performed at 37 weeks of gestation because of breech presentation. The patient's postpartum course was uneventful.

Early diagnosis of heterotopic cervical pregnancy using ultrasonography can shift the paradigm of management toward conservative methods. The goals of these methods include the minimization of blood loss, the preservation of fertility and the protection of a coexisting intrauterine pregnancy. Several modalities of conservative management for heterotopic cervical pregnancy have been developed, including pharmaceutical management, cervical suture or cerclage operation, hysteroscopic ablation, and angiographic embolization.

Although several cases of heterotopic cervical pregnancy have been managed by injection of potassium



**Figure 2** Transvaginal ultrasound image showing an intracervical hematoma, with a diameter of 3 cm, 1 week after the procedure to remove the cervical gestational sac.