More or less CHAOS: case report and literature review suggesting the existence of a distinct subtype of congenital high airway obstruction syndrome

A. C. VIDAEFF*, P. SZMUK†, J. M. MASTROBATTISTA*, T. F. ROWE‡ and O. GHELBER§

*Division of Maternal-Fetal Medicine, Department of Obstetrics, Gynecology and Reproductive Sciences and §Department of Anesthesiology, University of Texas Health Science Center at Houston and †Maternal-Fetal Medicine Associates of South Texas, TX and ‡Department of Anesthesiology, University of Texas Southwestern and Children’s Medical Center, Dallas, TX and Outcome Research Institute, Louisville, KY, USA

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ABSTRACT

Congenital obstruction of the upper airway (CHAOS) is a rare, usually lethal abnormality. A literature review of 36 prenatally diagnosed cases of CHAOS and the analysis of our own case suggest the existence of a distinct subtype of CHAOS, raising important implications for diagnosis and management. Serial fetal ultrasound examinations at 17–23 weeks’ gestation showed hyperechoic and enlarged lungs, mediastinal shift, flattened diaphragm, polyhydramnios and apparently fluid-filled esophagus, findings interpreted as bilateral cystic adenomatoid malformation Type III. Ultrasound findings normalized around 32 weeks. The diagnosis of CHAOS was made after birth at term by direct laryngoscopy prompted by ventilatory difficulties and failed attempts at intubation. A pinhole opening posterior to the cricoid cartilage allowed the passage of an endotracheal tube. Based on observations in our case and those of five similar cases in the literature, we describe for the first time a subtype of CHAOS that is characterized by minor pharyngotracheal or laryngotracheal communications and associated with a less severe natural history and even resolution of ultrasound findings. In spite of this, a high index of awareness should be maintained because resolution of ultrasound findings does not necessarily indicate resolution of underlying pathology. Copyright © 2007 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORT

A 27-year-old woman, gravida 2, para 1, was referred to our institution at 31 weeks of gestation. Previous ultrasound findings at 17, 20 and 23 weeks were interpreted as suggestive of bilateral congenital cystic adenomatoid malformation (CAM) of the lung, Type III. Both lungs were hyperechoic, with the mediastinum shifted to the midline, diaphragm flattened, and the heart appearing compressed (Figures 1 and 2). In addition, an elongated hypoechoic structure in the posterior mediastinum was identified and thought to represent the fluid-filled esophagus (Figure 3). Polyhydramnios and a single umbilical artery were also detected. The patient underwent genetic counseling and amniocentesis, which revealed a normal male karyotype. Ultrafast fetal magnetic resonance imaging performed at 24 weeks showed a grossly normal fetal chest (sequences after balanced fast field-echo localizers consisted primarily of T2-weighted single-shot imaging with additional fast field-echo images).

The initial ultrasound scan at our facility at 31 weeks revealed normal echogenicity of the lungs, and the previously described elongated hypoechoic structure in the posterior mediastinum was no longer detectable. Amniotic fluid volume was normal by 32 weeks of gestation, when pectus excavatum and right ventricular enlargement were also noted. Fetal echocardiography confirmed right ventricular enlargement and suspected pulmonary hypertension. A repeat ultrasound scan at 35 weeks yielded no additional relevant information. Biometry indicated appropriate fetal growth, with an estimated fetal weight at the 39th percentile for dates.

The mother had no significant personal or family history. The current pregnancy was complicated by diet-controlled gestational diabetes and late-onset gestational hypertension. She was admitted for labor induction at 38 weeks of gestation after spontaneous rupture of membranes. Ten hours after admission, a non-reassuring fetal heart rate pattern necessitated a primary low-transverse Cesarean delivery under lumbar epidural anesthesia. A liveborn male infant weighing 2655 g
Figure 1 Transverse view through the fetal chest, demonstrating the large hyperechoic lungs causing heart compression and midline displacement. The amount of amniotic fluid is increased.

Figure 2 Longitudinal view of the fetal chest and abdomen, demonstrating the enlarged hyperechoic lungs and the downward displacement of the diaphragm.

Figure 3 Longitudinal sonogram of the fetal chest. The elongated anechoic structure anterior to the upper spine represents the dilated, fluid-filled trachea, initially thought to represent the esophagus.

was delivered, with Apgar scores of 3 and 7 at 1 and 5 min. Upon delivery, the neonate appeared morphologically normal; however, he did not cry and manifested decreased breathing efforts and cyanosis. Two attempts at intubation with a 2.5 endotracheal tube (ET) failed, and the team resorted to mask ventilation. Half an hour later, under mask ventilation, the O₂ saturation was above 90% and a blood gas analysis revealed a pH of 6.95, pCO₂ > 115 mmHg and pO₂ of 73 mmHg. A chest X-ray demonstrated white lungs bilaterally, with air bronchograms and flattened diaphragm. One additional attempt to intubate the child failed and 2.5 h after birth the child was taken to the operating room. A telescopic laryngoscopy revealed normal vocal cords with subglottic atresia at the level of the cricoid cartilage. A pinhole opening was present posteriorly.

A 2.0 ET with a stylet was passed with significant difficulty into the trachea through the posterior opening. Despite good breath sounds bilaterally, the O₂ saturation remained low (50%) and the ET CO₂ was also low (3–5 mmHg). A presumptive diagnosis of pulmonary hypertension was made. Sodium bicarbonate (3 mEq) was administered, and the ET CO₂ and O₂ saturation increased rapidly to 50–60 mmHg and 100%, respectively. A tracheostomy was performed at the level of the third tracheal cartilage. During the procedure, the trachea was noted to be filled with pink foamy fluid. The hemodynamic parameters were stable and blood gas analysis confirmed normalization of gas exchange for the latter part of the surgical procedure. A postoperative echocardiogram showed moderate patent ductus arteriosus, patent foramen ovale, and pulmonary hypertension with right ventricular hypertrophy. The infant is scheduled to undergo an anterior cricoid resection with crico-tracheal anastomosis.

DISCUSSION

Congenital high airway obstruction syndrome (CHAOS) is a rare abnormality, usually with a lethal outcome. Laryngeal atresia is the most frequent cause but other etiologies include laryngeal or tracheal webs, laryngeal cysts, tracheal atresia, subglottic stenosis or atresia, and laryngeal or tracheal agenesis. When prenatal diagnosis of possible upper airway obstruction is made, the specific type of obstruction can only rarely be determined, making the term CHAOS, introduced by Hedrick et al.¹ in 1994, more appropriate. We conducted a literature search between 1965 and January 2006 using the MEDLINE bibliographic database and employing a combination of keywords, including ‘laryngeal atresia’, ‘laryngeal obstruction’, ‘prenatal diagnosis’, ‘fetus’ and ‘CHAOS’. All references in the retrieved articles were screened for further citations. The search yielded 36 prenatally diagnosed cases of upper airway obstruction¹⁻²⁸.

CHAOS is characterized by a number of sonographic structural secondary changes, some detectable as early as 16 weeks²⁸. Fetal lung size is increased, and the lungs are hyperechoic bilaterally. The increase in lung volume, up to 15 times the expected normal weight, is consistent with the stimulation in lung growth and hyperplasia that can be induced by upper airway obstruction. The hyperplastic lungs are edematous but otherwise histologically normal.
The increased number of tissue–fluid interfaces produces the hyperechoic appearance of the lungs on ultrasound. The fetal heart is not significantly displaced in CHAOS, although it usually assumes a more central position, and the diaphragm is flattened or everted.

In the absence of tracheal atresia, an enlarged fluid-filled trachea can be demonstrated, and nomograms for fetal tracheal diameter in relation to gestational age have been published. Using color Doppler, one can demonstrate absence of flow in the trachea during fetal breathing, and in some cases even localize the level of obstruction. The enlarged lungs may compress the superior vena cava, thoracic duct and heart, impeding venous return and causing ascites, non-immune fetal hydrops and placentomegaly. Oligohydramnios may be present early in pregnancy, due to the absence of lung liquid contribution to the total volume of amniotic fluid. Oligohydramnios is also present in cases of Fraser syndrome, in which renal or ureteral agenesis may be associated with the characteristic findings of cryptophthalmos, syndactyly and laryngeal atresia. Hydramnios may also develop, usually later in pregnancy, from compression on the esophagus and decreased fetal swallowing.

The differential diagnosis of CHAOS should take into consideration the possibility of bilateral CAM Type III, a condition that may be associated with polyhydramnios and hydrops fetalis. However, bilaterality in CAM III is very rare and, in contrast to CHAOS, fluid flow in the trachea can be demonstrated using color Doppler during fetal breathing. The diagnosis is more challenging in cases of CHAOS coexisting with a tracheo-esophageal fistula. This communication, by neutralizing the pressure effect, leads to less enlarged or normal-sized lungs. At the same time, the beneficial pressure effect is lost and cases of hypoplastic lungs have been reported. Such observations make the potential benefit of fetal surgery with in utero fetal tracheal decompression in cases of CHAOS uncertain.

We report here a case of a fetus with laryngeal atresia of the subglottic type whose prenatal ultrasound findings were initially considered to represent CAM Type III. Initial ultrasound abnormalities resolved by 31 weeks of gestation, and the underlying pathology was diagnosed only in the first hours of postnatal life by direct laryngoscopy, prompted by ventilatory difficulties and failed attempts at intubation. A pinpoint opening was noted posterior to the cricoid cartilage. We consider that the existence of this small laryngotracheal communication was responsible for the resolution of the secondary ultrasound changes in the third trimester and for the survival of the newborn in spite of the unanticipated diagnosis. Similar observations have been made by others, suggesting a particular natural history for CHAOS when minimal pharyngotracheal or laryngotracheal communications are present. In a case reported by Lim et al., the existence of a pinpoint posterior laryngotracheal fistula also allowed abatement of elevated intrathoracic pressure, leading to a decrease in lung volumes, reversal of diaphragmatic eversion, and resolution of ascites and polyhydramnios between 22 and 32 weeks of gestation. Another case reported by the same group similarly had a tiny posterior laryngotracheal fistula that explained the relatively stable lung volumes between 22 and 32 weeks of gestation. In the case of laryngeal atresia reported by Bui et al., gradual regression of ascites, lung hyperechogenicity and diaphragmatic eversion were observed between 25 and 30 weeks of gestation. A minimal communication measuring < 1 mm was found in the posterior cricoidal area after birth. It was presumed that the patent pharyngoglottic duct explained the resolution of secondary ultrasound changes. Still another case of CHAOS with in utero resolution of secondary ultrasound changes was reported by Richards et al. as early as 1992. By 30 weeks of gestation, ascites and polyhydramnios had resolved and the lungs appeared less enlarged. Spontaneous vaginal delivery was allowed at term and a tracheostomy was performed immediately after delivery, while the umbilical cord was left intact for continuous placental oxygen supply. The neonate was noted to have subglottic stenosis and a 1-mm posterior opening. This pinpoint communication allowed in utero release of the lung fluid with improvement in ultrasound abnormalities, as well as minimal air passage at birth, explaining the fortunate outcome even with such unique management. Minior et al. in 2004 presented a case of laryngeal atresia with a posterior pharyngotracheal duct having a more benign antepartum course, without fetal ascites or hydrops, and with less restriction on cardiac output. Obviously unaware of similarities with previously reported cases, the authors commented: ‘The sonographic appearance of our case of laryngeal atresia is unlike others in the literature’.

Based on these five previously reported cases and the sixth case reported here (17% of all the prenatally diagnosed cases of CHAOS known to us), we propose the existence of a subtype of CHAOS. All of these cases have in common the presence of a minor pharyngotracheal or laryngotracheal fistula responsible for partial decompression of the respiratory tract. This is in contradistinction to the complete decompression seen with larger tracheoesophageal fistulas, which lead to a totally different and frequently unfavorable outcome. It is also in contradistinction to the sustained occlusion characterizing the majority of CHAOS cases without any tract or outlet for decompression. Minor posterior fistulous communications have been described in the past in laryngeal atresia and have been associated with less severe clinical presentations. From the perspective of prenatal diagnosis, this subtype of CHAOS may be less severe, even with resolution of sonographic findings. It is important to reiterate that ultrasonographic resolution of fetal hyperechoic lungs does not necessarily indicate resolution of the underlying pathology, as suggested in 1993 by Meagher et al., referring to both CHAOS and CAM cases. Maintaining awareness and a high index of suspicion even after ultrasound findings resolve is important.

De Cou et al. reported in 1998 the successful use of ex utero intraartum treatment (EXIT) procedure for CHAOS owing to laryngeal atresia. The concept...
of securing the fetal airway, in cases of congenital upper airway anomalies, while the fetus is still on placental support was first advanced in 1954. Planned, anticipatory, direct laryngoscopy and tracheostomy can be salutary if prenatal diagnostic awareness exists, except for cases of tracheal agenesis or atresia, which cannot be salvaged. After 1998, our literature search identified eight additional cases of CHAOS in which an adequate airway was established while fetoplacental circulation was maintained, with favorable outcomes. In cases of CHAOS, including the CHAOS subtype described by us, the importance of predelivery planning cannot be overemphasized. Involving a multidisciplinary team including obstetricians, neonatologists, anesthesiologists and pediatric surgeons is crucial. Neonatal survival is improved if a well-planned EXIT procedure is performed at the time of a controlled near-term Cesarean delivery.

REFERENCES


