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Review

Airway compromise in the fetus and neonate: Prenatal assessment and perinatal management

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Congenital high airway obstruction syndrome (CHAOS)
Micrognathia
Pierre Robin sequence

SUMMARY

The fetus with a potentially obstructed airway can be identified on routine antenatal imaging. These cases should be referred to fetal care centers, which have the necessary expertise to fully evaluate and manage these fetuses and neonates appropriately. Complete airway obstruction may result in fetal hydrops and intrauterine demise. If a newborn infant has a compromised airway at delivery, the inability to secure its airway quickly may result in a hypoxic cerebral insult or death. In the most severely affected cases, prenatal, perinatal, or postnatal surgical intervention may be necessary. The timing of such an intervention will depend on the exact cause of the airway obstruction, other associated findings and the anticipated difficulty in establishing an airway at delivery. Fetal ultrasound and magnetic resonance imaging can differentiate between intrinsic and extrinsic airway obstruction, which allows for the optimal planning and management of the delivery and neonatal resuscitation.

1. Introduction

Fetal airway obstruction may be categorized into two groups: those that are extrinsic or intrinsic to the airway (Box 1).

2. Extrinsic airway obstruction

Any cause of extrinsic airway obstruction, from the lips down to the main-stem bronchi, may result in problems during fetal development, and difficulties in establishing an airway at delivery. Extrinsic airway obstructions may be in an oral, cervical, or thoracic location and the most frequent causes are cervical teratomas or epignathus, cervical lymphangiomas, and micrognathia.

2.1. Oral

2.1.1. Oropharyngeal teratoma: epignathus

Epignathus is a rare oropharyngeal teratoma arising from the sphenoid bone, palate or pharynx, with an estimated incidence of 1:35,000 to 1:200,000 [1]. It usually presents as a large mass protruding through the mouth and obstructing the upper airway. Immediate removal is often necessary at the time of delivery to allow an airway to be established by orotracheal intubation. A large epignathus tumor may present in the second trimester with polyhydramnios and airway obstruction [2]. If pharyngeal obstruction is complete, the ensuing polyhydramnios may require serial amnioreductions to minimize the risks of premature labor and delivery [3]. Imaging with ultrasound and ultrafast magnetic resonance imaging (MRI) can provide important information about the mass (Fig. 1).

Management of the fetus with epignathus is dictated by the degree of airway obstruction, gestational age at presentation and any other physiologic impact that the mass may have on the fetus. Some masses are small and do not completely obstruct the fetal airway and therefore can be managed by conventional intubation. By contrast, an epignathus which grows very rapidly to large exophytic proportions may completely obstruct the fetal airway and may even cause fetal hydrops from the increased blood flow through the tumor, resulting in a high output failure state. All such cases warrant urgent consultation at a fetal care center. Treatment involves resection of the mass, which, despite its large size, usually arises from a narrow pedicle on the fetal palate. Depending on gestation and severity, this might theoretically warrant

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Box 1
Potential causes of fetal airway obstruction.

Extrinsic airway obstruction

Oral
- Epignathus
- Epulis
- Micrognathia
- Parotid tumor

Cervical
- Teratoma
- Lymphangioma
- Congenital goiter
- Solid thyroid tumor
- Cystic anomaly of thyroid
- Branchial cleft cyst
- Hamartoma
- Hemangioma
- Lipoma
- Anterior neural tube defects

Thoracic
- Bronchogenic cyst
- Mediastinal teratoma
- Pericardial teratoma
- Congenital pulmonary airway malformation (CPAM)?

Intrinsic airway obstruction

Laryngeal atresia
Laryngeal cyst
Laryngeal web
Laryngeal stenosis
Tracheal atresia
Tracheal stenosis
Bronchial atresia
Bronchial web

consideration of open fetal surgery to resect the mass or, if there were a single systemic arterial feeding vessel, interstitial ablation of that feeding vessel. To date, neither of these approaches has been reported.

After 28 weeks of gestation, ex-utero intrapartum treatment (EXIT) to resect the mass and secure the airway prior to delivery should be considered [3]. EXIT will be discussed in detail later in this chapter (Fig. 2, Box 2).

2.1.2. Congenital epulis (Neumann's tumor)

Epulis, also known as a gingival granular cell tumor (GGCT) of the newborn [4,5], is an oral mass (or masses), which protrudes through the mouth and may obstruct the airway. Embryologically, the tumor arises from the gingival mucosa. Congenital epulis has a female preponderance of 8:1 and, although most are very small and incidental clinically, tumors vary in size from a few millimeters to as large as 7.5 cm (Fig. 3).

Epulis has often been reported to regress spontaneously after birth, suggesting that its growth may depend somewhat on maternal hormonal stimulation during pregnancy [6]. If an epulis is large, the diagnosis can often be made prenatally, allowing the planning of delivery in collaboration with the ear, nose and throat (ENT) or pediatric surgery airway teams in case an emergency resection or tracheostomy in the newborn is necessary.

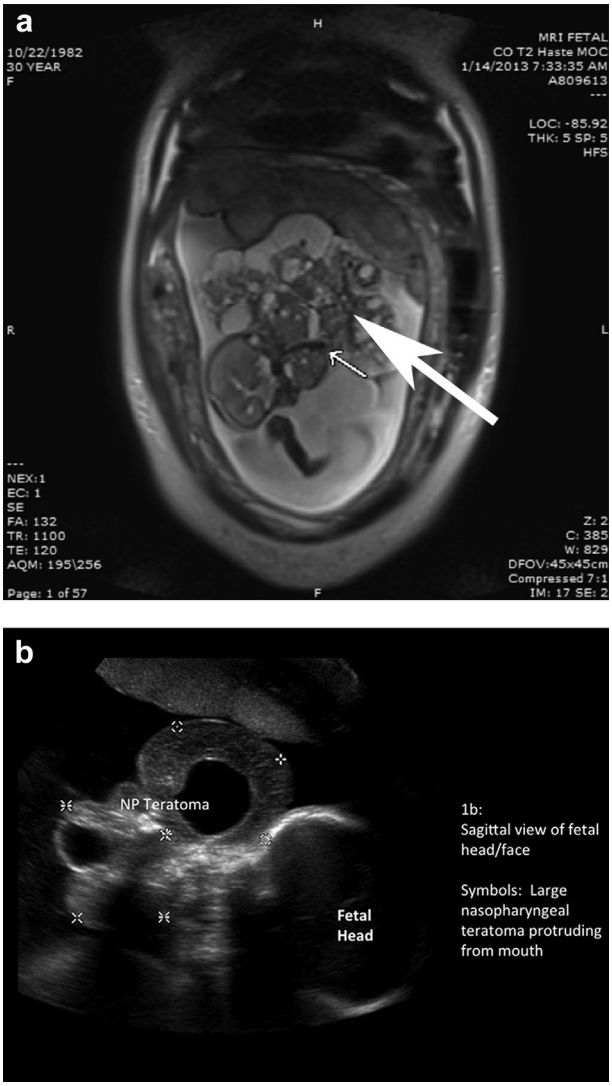

Fig. 1. (a) Fetal magnetic resonance image of a 25-week fetus with large exophytic epignathus mass larger than fetus. Arrow indicates a large vessel supplying the epignathus. (b) Fetal ultrasound showing a large nasopharyngeal teratoma protruding from the mouth.

At birth, intervention may be necessary, depending on the size and likelihood of airway obstruction. Epulis often originates from a pedicle and can often be easily resected before establishing an airway in the controlled environment of an EXIT-to-airway delivery [3].

2.1.3. Mandibular hypoplasia: micrognathia/retrognathia

Micrognathia or retrognathia may be isolated or syndromal and, in severe cases, may lead to upper airway obstruction. Pierre Robin sequence (PRS) may cause upper airway obstruction, and is associated with micro-retrognathia and glossoptosis. PRS occurs in 1:8500 to 14,000 births, being characterized by a small mandible accompanied by a U-shaped cleft palate [7]. Approximately 40% of PRS cases occur in isolation and 60% present as part of a syndrome. Other conditions associated with micrognathia include Stickler or velocardiofacial syndromes [8] (Fig. 4).

Severe micrognathia can also be part of a group of rare conditions known as arthrogryposis multiplex congenita, characterized by multiple non-progressive joint contractures, glossoptosis,