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Congenital and acquired developmental problems of the upper airway in newborns and infants ☆

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ABSTRACT

Aim: To review the current knowledge on congenital and acquired developmental problems of the upper airway in newborns and infants.
Data synthesis: Causes of airway obstruction include problems with the nasal airway (choanal atresia), craniofacial syndromes (Apert syndrome, Crouzon syndrome), problems with facial/tongue anatomy (Pierre-Robin syndrome), the tongue (Down syndrome), or the larynx (laryngomalacia, vocal cord palsy, subglottic stenosis, subglottic hemangioma), along with lower developmental problems (tracheo/bronchomalacia). After establishing a safe airway, a detailed assessment and appropriate management are necessary. Treatment may involve simple observation, conservative management, chest physiotherapy, CPAP ventilation, and surgery, urgently or in a second phase.
Conclusion: Upper airway diseases in neonates and infants may be life threatening, or challenging regarding diagnosis and management. There should be a very low threshold for referring these children, after establishing a safe airway, for a specialist opinion and care in a tertiary unit, if local facilities are limited or unavailable.

1. Introduction

The upper airway extends from the nasal aperture to the trachea and can be the site of multiple types of congenital or acquired diseases leading to anatomical or functional obstruction [1]. Neonatal airway obstruction may occur in any setting (home, delivery unit, hospital) and in various forms (life threatening, acute, sub-acute, and chronic). Therefore, all physicians involved should be familiar with the clinical presentations and endoscopic findings of the respective diseases, so that appropriate measures can be initiated at an early stage, thus avoiding significant complications.

The aim of the present paper is to review the current knowledge on the development of the airway, give an overview of causes of neonatal airway obstruction, and outline the latest management recommendations.

2. Development and characteristics of the neonatal airway

2.1. Development of the airway

The airway begins to develop as an outgrowth of the ventral part of the foregut at the fourth week of embryonic life. Thus the internal membrane of the respiratory system is of endodermal origin. The muscles and cartilages of the larynx are derived from the branchial arches. Initially the respiratory diverticulum communicates freely with the foregut, but as the diverticulum expands caudally, it becomes separated from the foregut by the oesophago-tracheal ridges, which fuse to form the oesophago-tracheal septum. At that time the internal epithelium also proliferates and occludes the lumen of the airway. This then recanalises so that the definitive airway is usually formed by the seventh week of gestation.

Developmental problems usually arise due to incomplete or non-canalisation (leading to laryngeal webs or atresia, respectively), or difficulties with the separation of the oesophagus and trachea (leading to tracheo-oesophageal fistulas).

The nasal cavity is also important especially in the neonate, and this begins to develop with the development of the nasal placode (a thickening of surface ectoderm) at the end of the fourth week of gestation. During the fifth week the placode is surrounded by the lateral and medial nasal swellings. This leads to the formation of a nasal pit. During the sixth week these pits deepen. The nasal cavity is separated at this point from the oral cavity by the oronasal membrane. This breaks down by the seventh week to form the primitive choanae. Later, once the palate has developed, the definitive choanae are located at the junction of the nasal cavity and the pharynx.

2.2. Characteristics of the neonatal airway

The neonatal airway is both absolutely and relatively smaller than the adult airway. As resistance is inversely proportional to the fourth

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power of the radius, very small decreases in the radius of the airway will lead to a large increase in resistance. The speed of the airflow has to increase to overcome the resistance, leading to turbulent airflow and increased noise, which manifests as stridor-a harsh sound (stridor is derived from the Latin term of "to creak").

The cartilages of the neonatal airway are also softer than that of a child or adult, and therefore more susceptible to extrinsic and intrinsic compressions, thus reducing the radius of the airway.

Finally, the administration of β-agonists tends to make the symptom of the neonatal airway worse, as it annuls the splinting effect of the bronchial muscles.

3. History and examination of the neonate suggesting airway problems

With the advent of detailed high resolution ultrasound scanning, prenatal diagnosis of potential airway difficulties has become more common. Ultrasound can detect tumours of the head and neck that could cause potential airway compromise, and can also detect congenital high airway obstruction (CHAOS). The features of CHAOS on ultrasound are polyhydramnios, increased echogenicity of the lungs, a dilated trachea (if there is no tracheooesophageal fistula to allow lung fluid to escape) and flattened or inverted diaphragm [2].

More recently ultrafast MRI scanning can be used to assess any masses in more detail, and allow planning of the ex utero intrapartum treatment (EXIT) procedure [3]. The EXIT procedure is a procedure in which the baby's head and shoulders are delivered by caesarean incision. The foeto-maternal circulation is maintained and this allows about 60 min of surgical time for the multidisciplinary team to establish a safe airway by intubation or tracheostomy. This has become more successful as the multidisciplinary team approach has been developed.

Airway obstruction cannot always be diagnosed or predicted prenatally. The main symptom after birth is stridor. This can be inspiratory, expiratory or biphasic. Inspiratory stridor arises from the supraglottic airway (nose to glottis). Airway obstruction more distally may be biphasic or expiratory. Features which should be noted about stridor are its onset after birth, whether it is intermittent, or positional, whether the voice is normal, whether there are any blue spells, and how feeding is affected.

On examination, any dysmorphic features need to be noted, particularly the size of the jaw, any cleft lips or palate, or facial deformities. The rate of respiration and work of breathing also need to be recorded. Severe stridor or a very quiet baby, tracheal tug and subcostal recession are worrying signs, as they imply impending airway disaster. In contrast, cyanosis is a late sign.

Causes of airway obstruction include problems with the nasal airway, craniofacial syndromes, problems with facial or tongue anatomy, problems with the tongue, and problems with the larynx. Although located just below the upper airway, developmental problems of the trachea or bronchi present with similar symptomatology, and will also be discussed in brief.

4. Developmental problems of the neonatal airway

4.1. Nasal problems

4.1.1. Choanal atresia

Choanal atresia is the most common cause of nasal airway obstruction (Fig. 1). It has been attributed to the persistence of the bucco-pharyngeal membrane. It occurs in 1 in 10,000 births, twice as often in girls, and is more often unilateral (unilateral:bilateral 2:1). The problem with choanal atresia stems from the fact that neonates are obligate nasal breathers. When the nose is blocked the child makes an effort to breathe, and when the mouth is closed the tongue is drawn up to the palate. The baby then becomes cyanotic until the mouth is open to cry. The child then pinks up quickly until the mouth is closed and the cycle begins again.

Bilateral choanal atresia is a neonatal emergency. The baby may need to be resuscitated at birth. The aforementioned history will give rise to the suspicion of choanal atresia. Other signs include a failure to pass a fine catheter through the nose, and absence of misting when a cold spatula is placed under the nostrils. The differential diagnosis includes septal deviation, septal dislocation, nasal masses, and encephalocele. Endoscopes may be very useful in the differential diagnosis.

The first priority is to establish an airway, and this can be done by intubation, or the insertion of a McGovern nipple (an oropharyngeal airway fashioned by cutting off the end of a large feeding nipple, placing it in the baby's mouth and attaching it using tapes around the baby's head) with a port for feeding. This will break the mouth seal and prevent airway obstruction. The baby must then be transferred to a specialist centre for definitive management.

If the newborn suffers from bilateral atresia, full examination must be undertaken to ascertain whether there are features of the CHARGE association (coloboma, heart defects, atresia of the choanae, growth retardation, genitourinary abnormalities and ear deformities-about 50% of children with bilateral atresia will have some of the other abnormalities). Further diagnosis of the condition requires imaging with a CT scan. It is vital to decongest and suction the nose prior to scanning in order to delineate the anatomy accurately. Atresia can be bony, membranous, or mixed [4].

Regarding the surgical management, blind puncture of the choanae is no longer used, due to recurrence and the dangers of CSF leaks. The main routes of repair are endo-nasally with the aid of 120° telescope, or trans-palatally. Maintenance of the surgical outcome is a matter of debate as different centres recommend stents to keep the newly formed airway patent or repeated dilations.

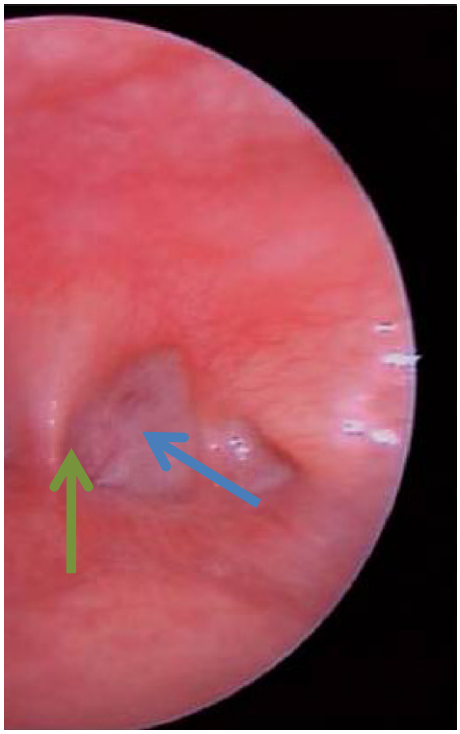

Fig. 1. Right choanal atresia (endoscopic view through the mouth. Green arrow vomer, blue arrow membranous atresia).