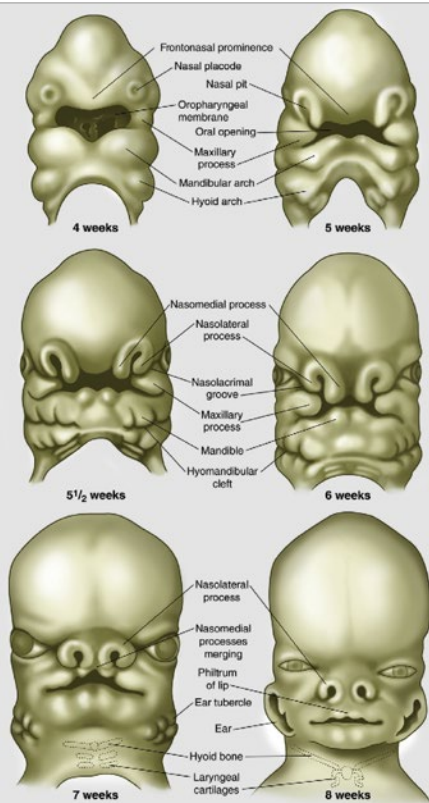


If you cannot bag mask
ventilate a neonate:
consider nasal obstruction



Durand S, Kämpfen S, Stöcklin B, Gürtler N, Department of Neonatology (DS, KS, SB), University Children's Hospital of Basel, Department of ENT Surgery (GN), University Hospital of Basel, Switzerland

Title figure:

Embryology of facial development

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INTRODUCTION

Choanal atresia (CA) is a rare congenital malformation with dramatic consequences if not recognized early. We report on a female newborn who suffered from postnatal asphyxia due to unrecognized bilateral CA.

CASE REPORT

This female infant was born to a 30-year-old G2/P2 at 41 0/7 weeks' gestation with a birth weight of 4100 g (P75), a length of 54 cm (P85) and a head circumference of 37 cm (P75). The pregnancy was unremarkable, without any maternal medication. The first child of the family was healthy. The infant was delivered by elective repeat Cesarean section in a peripheral hospital. The Apgar scores were 8, 1 and 1 at 1, 5 and 10 minutes, respectively. Arterial umbilical cord pH value was 7.30.

After an initial strong cry, she was given shortly to the mother for bonding in the operating theater. At two minutes of life, she showed marked respiratory distress followed by apnea and cyanosis. She was taken directly to the resuscitation table where she was noted to be unresponsive, bradycardic without respiratory effort and cyanosed. Cardiopulmonary resuscitation was initiated immediately without improvement. When the neonatal transport team arrived at 30 minutes of life, the child was still bradycardic and required intermittent cardiopulmonary resuscitation with positive pressure ventilation and chest compressions.

First attempts to intubate nasally were not successful though both nostrils; the baby girl was eventually intubated orally. Bradycardia and cyanosis resolved after a second dose of adrenaline. When blood pressure remained low despite multiple fluid boli, a continuous infusion with dopamine was started.

The first blood gas after intubation showed profound mixed acidosis with a pH of 6.81, pCO₂ 12.9 kPa (97 mmHg), and a BE -18.9 mmol/l. After initial stabilization, the baby girl was transferred to the tertiary NICU for therapeutic hypothermia due to hypoxic ischemic encephalopathy (HIE) with a Sarnat score of two.

The neurological symptoms resolved completely within the first few days, without the occurrence of any seizures. An MRI showed a small infarction in the right caput of the nucleus caudatus, but no typical signs of HIE.

Choanal atresia (CA) was suspected to be the likely underlying cause for failed nasal intubation attempts. This was confirmed by nasal endoscopy and a CT scan, which revealed complete bilateral mixed CA (Fig. 1). Apart from a small atrial septal defect and patent ductus arteriosus no associated defects were detected.

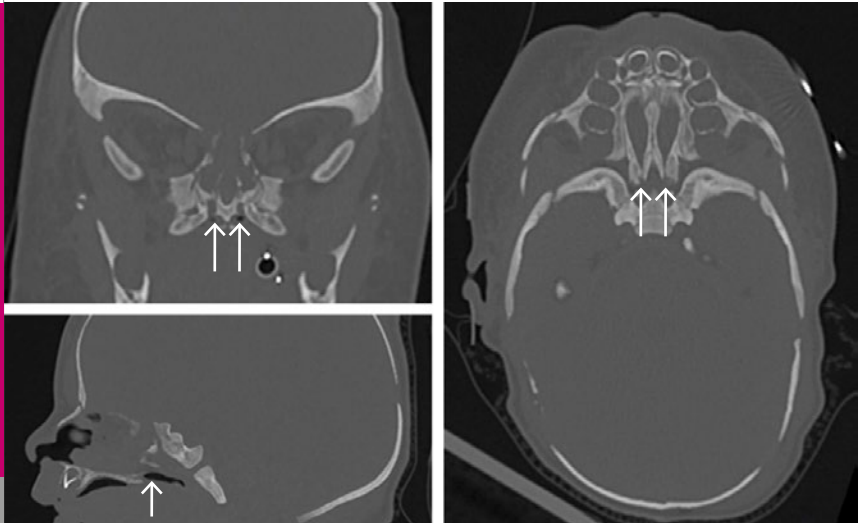


Fig. 1

CT scan demonstrating bilateral choanal atresia (top left: coronal view, bottom left: sagittal view, right: transversal view).

Endoscopic surgery was performed on day of life (DOL) 7 by resecting the atretic plate with the posterior part of the vomer and placement of nasal stents (size 2.5). The baby was extubated one day later, but required reintubation on DOL 9 because of respiratory failure due to obstruction of the nasal stents.

On DOL 11, the stents were replaced by bigger nasal stents (size 3.5). Topical corticosteroids and antibiotics, as well as an oral proton pump inhibitor were administered to avoid re-stenosis. The patient was successfully extubated to CPAP on DOL 12, and the stents were removed on DOL 20. Four days later, she was discharged home.

She was followed by the ENT surgeons. Due to secondary stenosis, an additional surgical intervention was performed at the age of 8 months.

At neurological follow-up, there was normal neurodevelopment.

DISCUSSION

Choanal atresia (CA) is a rare cause of upper airway obstruction that occurs in 1:5'000–7'000 live births with a female to male ratio of 2:1. Two thirds of patients have unilateral CA. In addition, 30 % of CA have a bony component only, whereas the remaining cases have also a membranous component (i.e., mixed CA) (1, 2).

The embryology and pathogenesis of CA is not completely understood, but several hypotheses have been proposed over the past years. During the first 4 weeks of gestation, face formation is driven by proliferation and migration of neural crest cells from the ectoderm to form nasal pits, followed by nasal sacs. This occurs by invagination of the ectoderm into the mesoderm. The wall of this sac consists of the oronasal membrane (of Hochstetter), that normally quickly ruptures to build the posterior primitive choane. CA is thought to occur if this membrane does not rupture (Fig. 2).

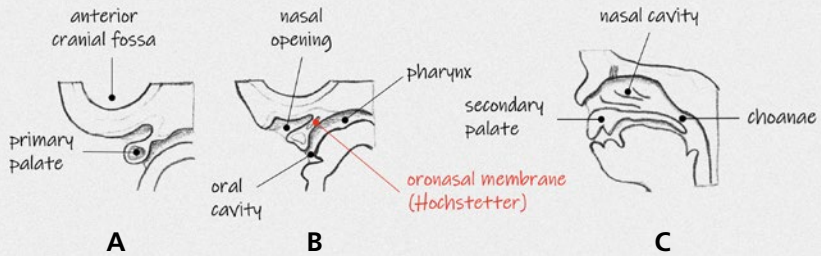


Fig. 2

Embryology of the face, nose and choanal formation: Proliferation and migration of neural crest cells form nasal sacs by invagination of ectoderm into mesoderm (A). The wall of this sac consists of the oronasal membrane (of Hochstetter) that normally quickly ruptures to build the posterior primitive choanae (B, C). CA is thought to occur, if that membrane does not rupture (redrawn after Götte et al.).

Other possible pathophysiological mechanisms include the diversion of neural crest cell migration, persistence of the buccopharyngeal membrane of the foregut and persistence of mesodermal adhesions in the nasochoanal region. Finally, medications such as methimazole, an antithyroid drug, administered in the first trimester, or the use of retinoids have been proposed as an underlying cause. Retinoids are thought to disturb mesenchymal cell migration and division. However, a causal relation between these two drugs and CA has never been proven (1). Our patient's mother had not taken any medications during pregnancy.

CA can be isolated or associated with several syndromes. In about 25 % of cases, CA is associated with the CHARGE syndrome (coloboma, heart defect, atresia of choanae, retarded growth and development, genitourinary abnormalities and ear defects). CA has also been described in patients with midface malformations or Treacher Collins syndrome (1, 3).

The diagnosis of CA, especially in patients with bilateral CA, can be suspected at birth based on its impressive clinical presentation. Affected patients suffer from severe respiratory distress with subsequent bradycardia and cardiac arrest because neonates are preferential nose breathers. In infants with such clinical presentations, unsuccessful nasal insertion of a feeding or suctioning tube beyond 3 to 3.5 cm

makes the diagnosis highly likely. After oral intubation and clinical stabilization of the infant, the diagnosis is confirmed by flexible nasal endoscopy. A CT scan of the skull allows differentiation between bony, membranous or mixed CA, and will help guide further management.

Diagnosis of unilateral CA is usually made later, as newborns can often breathe sufficiently through one nostril. There are several case reports in the literature where diagnosis was made in late childhood or even adulthood. In children with facial anomalies, CA should be explicitly excluded.

The initial management of bilateral CA is to maintain upper airway patency via orotracheal intubation. Newborn infants with bilateral CA often have a good initial cry at birth and collapse afterwards when nose breathing is not possible. Positive pressure ventilation is often difficult, and CA should be suspected when bag and mask ventilation is impossible. Insertion of a Guedel tube or laryngeal mask airway (LMA) can be life-saving until a skilled physician can proceed with oral intubation (Fig. 3) (4).

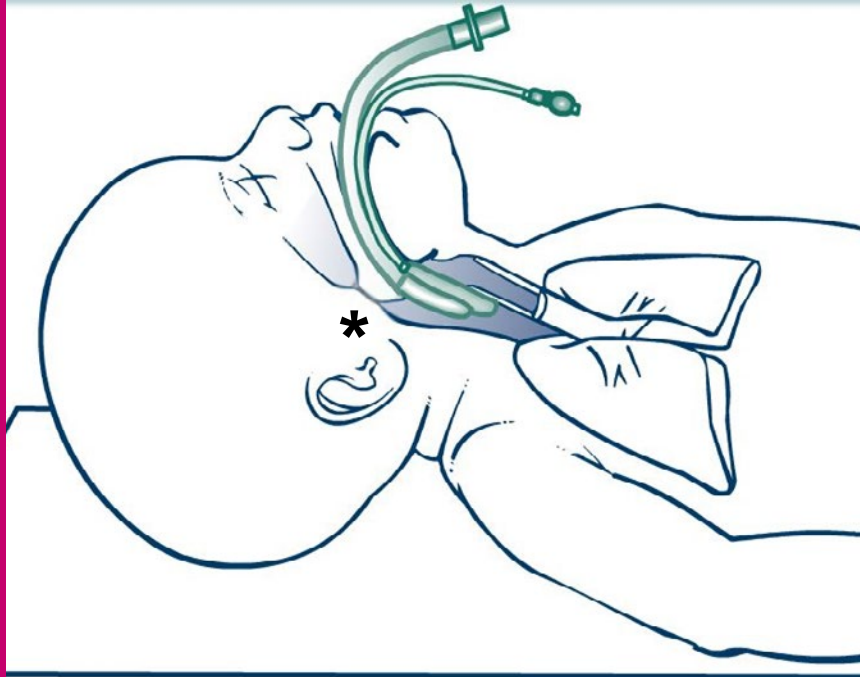


Fig. 3

The use of a laryngeal mask airway (LMA) – until a team skilled in neonatal intubation arrives – can be life-saving in neonates with bilateral CA (asterisk).

Additional malformations should be excluded by ultrasound screening (abdomen, head, and heart). Hearing tests (otoacoustic emissions) should be performed to detect any hearing impairment early.

The surgical management depends on the nature of the CA (membranous/bony). Historically, CA resection was performed by transnasal puncture, but was often complicated by relapse of stenosis requiring dilatation or subsequent surgeries. A transpalatine (open) approach was also proposed in the past, but had also several complications including fistulae and orthodontic issues (like cross-bite).

For the past 20 years, the transnasal endoscopic resection has been the most commonly used technique, with or without stenting to maintain choanal patency (Fig. 4). Interestingly, stenting is discussed controversially in the literature (1, 5, 6). Some of the complications of this technique could be related to the stent, like granulation disorders or lesions caused by the stent itself leading to restenosis. Strychowsky et al. (5) reported that surgery with placement of a stent was not superior compared to surgery without a stent.



Fig. 4

Nasal stents after atresia repair (not our patient).

The most recently proposed approach is the trans-septal endoscopic technique, which so far has shown good results with very low re-stenosis rates (1, 7, 8). This technique spares the mucosa and includes resection of the vomer. Apparently, resection of the vomer creates enough space to avoid restenosis. It can be challenging though when there is not enough space for the surgical instruments.

The postoperative management has the goal to keep the neochoanae open and support the healing process. Topic application of glucocorticoids and antibiotics (like fusidic acid) during the first two weeks after surgery has been described to reduce re-stenosis rates. Afterwards, treatment with a corticoid nasal spray for 6 to 8 weeks has been suggested. Some small trials suggested that perioperative local application of mitomycin C reduces granulation formation, re-stenosis and the need for repeated surgery (9). However, the effectiveness of such treatment has yet to be confirmed in larger clinical trials.

CONCLUSION

CA is a rare congenital malformation, with potentially dramatic consequences if not recognized early. In particular, bilateral CA is a life-threatening condition. When bag mask ventilation is not possible in a depressed infant after birth, CA should be strongly considered and airway patency should be maintained by oral intubation. A Guedel tube or a LMA can be used temporarily until a team with the necessary intubation skills arrives. When perinatal asphyxia can be prevented, prognosis of isolated CA is generally good.

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CONTACT

Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch