

CHOANAL ATRESIA

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Condition and Description

Choanal atresia is a term that describes congenital obstruction of the nasal passage due to presence of a membrane or boney tissue obstructing airflow through the nostrils. This obstruction fails to rupture when it typically does on about the 38th day of fetal development. For an unknown reason, this complex developmental process in the midline of the face is disrupted, interrupting the normal process of formation of the nasal passages.

Babies are “obligate nose breathers”, meaning that they can only breathe through their noses; the only time newborns breathe through their mouths is when they are crying. Babies with bilateral choanal atresia (both nostrils affected) will have severe respiratory distress (trouble breathing) in the newborn period with cyclical cyanosis (periods of their skin turning grey/blue due to lack of oxygen). This is because with crying they will be able to exchange air by mouth-breathing. However, when they close their mouths to suck they will not be able to breath. If only one nasal passage is obstructed this condition might not be detected for months or years.

To test a baby for the presence of choanal atresia a thin plastic tube will be gently inserted into the nasal passage. In normal babies it will pass through readily into the back of the throat.

Prevalence

Choanal atresia is the most common nasal abnormality and occurs in about 1 of every 7000-10,000 births. When only one nostril is affected (unilateral atresia) it will typically be

the right nostril. Twice as many babies are born with only one nostril affected (unilateral) as with both nostrils obstructed (bilateral choanal atresia). There is a slightly increased risk in twins. It occurs in all races and slightly more often in females.

Common Associated Conditions

Chromosomal anomalies are found in 6% of infants with choanal atresia and 5% of babies have syndromes with multiple defects. Nearly half of all babies will have associated palate and nasal wall malformations. An example is a syndrome called CHARGE syndrome that typically involves eye conditions (i.e. coloboma of the iris, choroid, and/or microphthalmia), a heart defect (atrial septal defect, ASD or endocardial cushion defect), choanal atresia, delayed growth and development, genital conditions (such as cryptorchidism, microphallus, and/or hydronephrosis), and ear defects.

Short-term Treatment and Outcomes

CT (computerized tomography) scanning will be done to evaluate the extent of the choanal atresia once a feeding tube fails to pass through the nasal passage. A small plastic tube will be surgically inserted in the nostril to create an airway for the baby until more extensive surgery can be done. The baby will need very close observation and careful suctioning of mucous to be sure that breathing is maintained. Some babies do learn to mouth-breathe so surgery can be delayed. However, feeding is very difficult, so an airway tube through the mouth or a tracheostomy (breathing tube in the neck) may be necessary.



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Long-term Treatment and Outcomes

Surgery will be necessary, perhaps involving both a nose-and-throat specialist (otolaryngologist) as well as a plastic surgeon to create a functioning airway and to obtain a good cosmetic appearance of the nose. Surgery may be delayed a few months so that the baby's facial structures can grow and repair will be more successful if the child's airway and feeding can be maintained.

Sometimes further surgical correction is necessary. The ultimate outcome for the child should be very good if there is no problem with resuscitating the baby at birth or with tolerating the surgical repairs. Babies who have complex syndromes and other conditions will have variable outcomes depending on those other problems. Their choanal atresia, once detected and repaired, should not pose long-term problems for those babies with complex conditions.

Common Complications

The nasal passageways may narrow as time goes by (stenosis) and need to be dilated.

Implications for Children's Development

If the baby is resuscitated successfully at birth and the condition is detected before significant problems occur (i.e. brain damage from lack of oxygen during the cyanotic spells), there should not be any long-term implications for the baby's development. However, nearly half of the children will have some other defect of the nose or palate, so there may be a number of surgeries that may cause discomfort and affect the child's appearance.

Choanal atresia ranges from very mild to severe. It may be impossible to detect once the nasal passage has been surgically repaired, or in other cases, it may significantly involve the appearance of the face, causing the child to be self-conscious. Building the child's self-confidence in light of the problem will be a very important task for parents and others who interact with the child at home, at school and in medical settings. Dental abnormalities might also be associated with the condition, making it important that the child be cared for in a pediatric multi-disciplinary cranial-facial clinic that brings together all the professionals who care for the child, including genetics, speech therapy, nursing, nutrition, plastic surgery, and ear-nose-throat specialties.

Monitoring the child's growth and nutritional status will be important, and social workers might also be helpful to the family coping with raising a child with complex, long-term health care needs.

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