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# Imaging of Nasopharyngeal Atresia

J. Keith Smith, M. Castillo, S. Mukherji, J. Buenting, and A. Drake

**Summary: CT and MR revealed a case of nasopharyngeal atresia, a malformation in which the soft palate is not formed, and the hard palate extends posteriorly to fuse with the anterior surface of the clivus, resulting in complete isolation of the nasal and oral cavities and the absence of a nasopharynx. We believe this rare anomaly results from abnormal persistence of the embryologic bucconasal plate and/or anomalous migration of the nasoseptal elements.**

**Index terms: Nasopharynx, abnormalities and anomalies; Infants, newborn**

## Case Report

A 3300-g girl was born at 39 weeks' gestational age via spontaneous vaginal delivery. The pregnancy was complicated by polyhydramnios. Prenatal ultrasound had not shown any abnormalities. There was no family history of birth defects or genetic abnormalities. At birth the patient was noted to have a bilaterally cleft lip, without cleft palate. Physical examination also revealed multiple skin tags anterior to the left tragus and deformity of the left pinna. There was mild facial asymmetry with left-sided microsomia. The palpebral fissures were bilaterally shortened and horizontal with a slightly decreased intercanthal distance. Chromosomal analysis revealed a normal female karyotype (46,XX). These abnormalities did not conform to any known syndrome.

Shortly after birth the patient had respiratory difficulty, with pronounced mouth breathing. At this time passage of a 6F catheter for nasal airway was attempted through both nostrils without success. An oral airway was established.

Computed tomography using axial 2-mm contiguous images with a high-resolution edge-enhancement reconstruction algorithm revealed fusion of the posterior hard palate and vomer with the anterior surface of the sphenoid bone (Fig 1A and B). The nasal cavity was filled with secretions, and the nasopharynx was absent. The malar eminences were small bilaterally.

Magnetic resonance imaging of the face and brain confirmed the absence of the nasopharynx and soft palate with the hard palate and vomer fused with the clivus (Fig 1C and D). The brain was normal.

Rigid nasal endoscopy was performed with a pediatric nasal endoscope. This revealed a progressive distal narrowing of the nasal cavity with complete agenesis of the nasopharynx. The palate was fused to the posterior pharyngeal wall without communication to the nasal cavity. A tracheostomy was performed to secure an adequate airway for the patient. The patient is being followed and is expected to learn oral breathing.

## Discussion

During the fourth week of intrauterine life, the nasal placodes develop. These thickened skin folds are located lateral to the frontonasal prominence and superior to the oral opening. Two small depressions, the nasal pits, form in the center of the nasal placode. The nasal pits become deeper and are separated from the ectoderm overlying the stomodeum (cephalic aspect of gut) by the *bucconasal plate* or *membrane* (1). Eventual resorption of this membrane establishes communication between anterior (primary) choanae and stomodeum (1). The external openings of the nasal pits are the nostrils. The posterior openings of the nasal pits are the primitive posterior choanae and form later (2). Once the primitive choanae have opened, the nasal cavity communicates opening with the oral cavity. By the eighth week of life, the nasal cavity is separated in halves vertically by the developing nasal septum and separated inferiorly from the buccal cavity by the palate, formed as the palatine shelves converge from the lateral walls. Together, these structures are referred to as the *nasoseptal elements* (1). Establishment of the soft palate completes the development of the nasal cavities. Elongation and migration of the lateral and superior nasoseptal elements, the hard palate, and the nasal septum add further tubulation to the primitive nasal and oral cavities forming the second-

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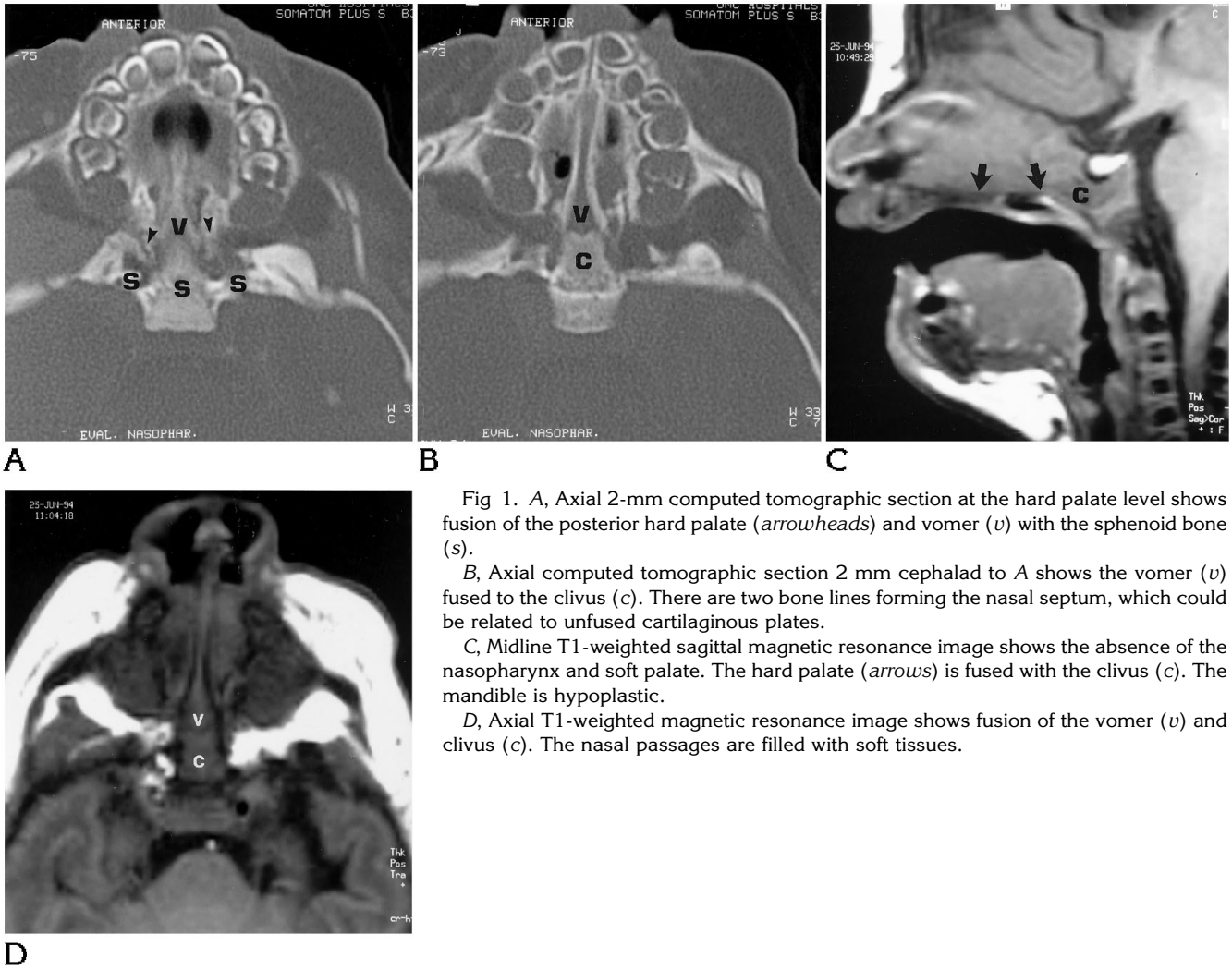


Fig 1. *A*, Axial 2-mm computed tomographic section at the hard palate level shows fusion of the posterior hard palate (*arrowheads*) and vomer (*v*) with the sphenoid bone (*s*).

*B*, Axial computed tomographic section 2 mm cephalad to *A* shows the vomer (*v*) fused to the clivus (*c*). There are two bone lines forming the nasal septum, which could be related to unfused cartilaginous plates.

*C*, Midline T1-weighted sagittal magnetic resonance image shows the absence of the nasopharynx and soft palate. The hard palate (*arrows*) is fused with the clivus (*c*). The mandible is hypoplastic.

*D*, Axial T1-weighted magnetic resonance image shows fusion of the vomer (*v*) and clivus (*c*). The nasal passages are filled with soft tissues.

ary (permanent) posterior choanae. The mechanism responsible for the production of choanal atresia is unclear. It is commonly accepted that persistence of the bucconasal plate is responsible (3). We believe that this view is not completely correct, because this plate forms adjacent to the anterior choanae, whereas atresia involves the posterior choanae. Overgrowth of the posterior nasoseptal elements induces or results from lack of formation of the permanent posterior choanae. This overgrowth may result in stenosis or bony posterior choanal atresia. It is also possible that incomplete recanalization of the temporary epithelial nasal plugs leads to membranous atresia. It has been postulated that misdirection of mesodermal (nasoseptal elements) flow produces the thickening of the posterior inferior vomer, which characterizes bony choanal atresia. We believe that persistence of the bucconasal plate possibly in combination with abnormal migration of the nasoseptal elements results in the abnormality presented here, nasopharyngeal atresia. In this unusual malformation there is complete isolation of the nasal and oral cavities. The soft palate is not formed, and the hard palate extends posteriorly to fuse with the anterior surface of the clivus. The posteroinferior vomer is large, both in length and width, and merges with the anteroinferior clivus. The nasopharynx is not formed. This constellation of findings results in severe respiratory distress in newborns, who are obligate nasal breathers for the first several months of life. Absence of the choanae and agenesis of the nasopharynx precludes endoscopic repair of the choanal atresia. Similarly, a standard transpalatal approach to the nasopharynx is not feasible because of the close proximity of the skull base to the hard palate and the resultant risk of intracranial injury. Al-

though creation of an oronasal fistula is surgically possible, it would be located fairly far anteriorly in the oral cavity because of progressive distal narrowing of the nasal cavity. The tongue would occlude a surgical fistula in the midpalate, and thus this approach would not provide an adequate nasal airway. To ensure an airway through the newborn period, a tracheostomy is necessary, despite the risks associated with this procedure in neonates.

In summary, nasopharyngeal atresia is a very rare malformation, which is clinically indistinguishable from the more common posterior choanal atresia. Arrested or misdirected migration of nasoseptal elements leads to the absence of posterior choanae, the absence of the nasopharynx, and a hard palate that is fused with the clivus. Imaging studies confirm these findings and are indispensable in guiding treatment.

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