NeoReviews™

NeoQuest March 2023: Term Newborn with Stertor

March 9, 2023

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Article type: Resources

A nondysmorphic term infant born via uncomplicated vaginal delivery develops respiratory distress with grunting, retractions, stertor, and cyanosis shortly after birth. When crying, the infant's cyanosis resolves. A 5-French catheter is passed through both nares. A computed tomography scan is obtained (Figure 1).

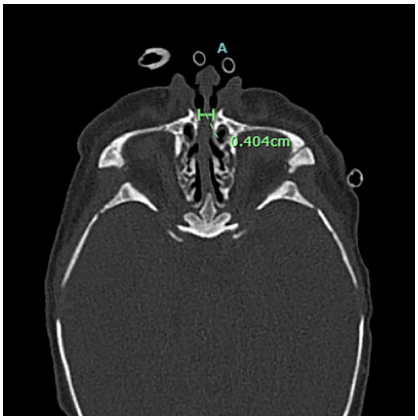

Figure 1: An axial maxillofacial computed tomography image of a neonate with respiratory distress and stertor notes a narrow pyriform aperture measuring 4mm (green measurement) in the anterior nasopharynx. Image from: Burch JL, Whitesel E, Manzi B, Adil E. A neonate with obstructed nasal breathing. *Neoreviews*. 2023;24(3)e195.10.1542/neo.24-3-e195 [1]

Which of the following clinical features is most likely associated with this condition?

A. Craniosynostosis
B. External ear anomaly
C. Meningitis
D. Solitary maxillary central incisor

Explanation:

Answer: D. Solitary maxillary central incisor

The infant in this case has clinical and imaging features consistent with congenital nasal pyriform aperture stenosis (CNPAS). The pyriform aperture is the narrowest and most anterior portion of the nasal airway.[2] Anatomic narrowing of this bony structure will significantly increase airway resistance, resulting in a congenital nasal obstruction. Depending on the degree of obstruction, the clinical presentation of CNPAS can vary from feeding difficulties to mild-to-severe respiratory distress with grunting, nasal flaring, and retractions.[2] Some individuals with severe CNPAS may also develop apnea and cyclical paradoxical cyanosis that resolve with crying.[3] The diagnosis of CNPAS requires clinical suspicion as well as

confirmation of pyriform aperture narrowing within the anterior nasal airway on maxillofacial computed tomography (CT) imaging (Figure 1).[3] In a full-term infant, the aperture is considered stenotic when the distance between the medial aspects of the maxillae is less than 11 mm, or when the maximum transverse diameter of each aperture is less than 3 mm.[3] This condition can be an isolated malformation or be associated with midline defects such as a cleft lip and palate, holoprosencephaly, and solitary maxillary central incisor (**Option D**) (Figure 2).[4]

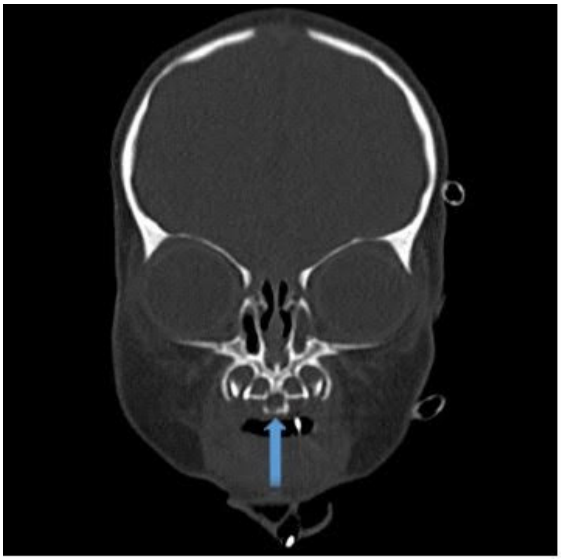

Figure 2: A coronal maxillofacial CT scan of a newborn with CNPAS that reveals a solitary maxillary central incisor (blue arrow). Image from: Burch JL, Whitesel E, Manzi B, Adil E. A neonate with obstructed nasal breathing. *Neoreviews.* 2023;24(3)e195.10.1542/neo.24-3-e195

Nasal dermoid cysts are congenital midline nasal masses that may contain derivatives of ectoderm and mesoderm (eg, hair follicles, sebaceous glands, or eccrine glands) and can present similarly with symptoms of nasal obstruction.[3] Most cases are sporadic and are not associated with syndromes. However, other craniofacial abnormalities such as cleft lip and palate, craniosynostosis (**Option A**), and hypertelorism have been described in children with nasal dermoids.[3,5] Hair protruding from a sinus pit or ostium on a midline nasal mass is pathognomonic for nasal dermoid cyst.[3]

Choanal atresia (CA) exhibits similar clinical features and should be differentiated from CNPAS. CA is a congenital obstruction of the posterior nasopharynx that develops due to a failure of the bucconasal membrane to perforate between the fifth and sixth weeks of gestation.[3] CA can be bilateral or unilateral, membranous or bony.[2] Similar to CNPAS, affected infants with CA classically present with respiratory distress at birth that is relieved by crying. The diagnosis of CA can be suspected by failure to pass a 5-French flexible tube through both nares.[5] Flexible nasal endoscopy and the presence of uni- or bilateral posterior nasal narrowing visualized on maxillofacial CT (Figure 3) can confirm the diagnosis and help differentiate CA from CNPAS.[5] CA is highly associated with other congenital anomalies and/or syndromes, including CHARGE (*c*oloboma, *h*eart defects, *a*tresia of the choanae, *r*etardation of growth and/or development, *g*enital/urinary abnormalities, and *e*ar anomalies/deafness) (**Option B**), Crouzon, Pfeiffer, Treacher Collins, and Antley-Bixler syndromes.[3] Of these syndromes, CHARGE is most commonly associated with CA.[5]

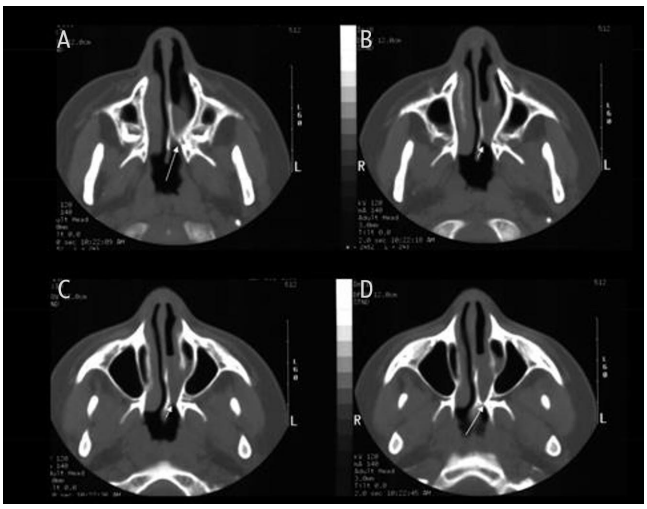

Figure 3: Axial maxillofacial CT demonstrating narrowing of the left posterior nasal airway due to bony (long arrows, panels A and D) and membranous soft tissue (short arrows, panels B and C) components, confirming left choanal atresia. Image from: Goldstein NA, Gitman L. Neonatal nasal deformities. *Neoreviews.* 2015;16(1):e36–e46 [3]

Nasal encephaloceles are a type of neural tube defect in which meninges and/or brain tissue herniates into the nasal cavity (Figure 4).[2,6-9] Along with respiratory distress, infants with a nasal encephalocele may present with a visible nasal or orbital mass, broadened nasal bridge, and hypertelorism, depending on the subtype and/or location of the herniation (Figure 5).[6-7] A hallmark feature of a nasal encephalocele is an acute enlargement of the pulsatile mass while crying, known as Furstenberg's sign.[2,10] Individuals with this type of meningeal herniation are at risk for cerebrospinal fluid leak and meningitis (**Option C**).[6,10]

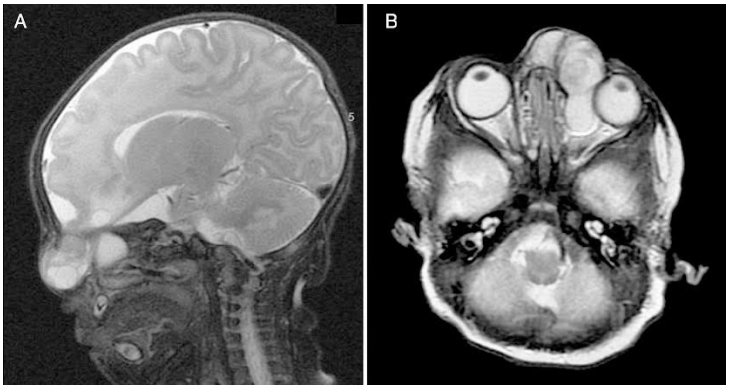

Figure 4: Sagittal (A) and axial (B) magnetic resonance imaging of a nasal encephalocele in a neonate. Image from: Van Heukelom JG. Case 3: an abnormal nose mass. *Neoreviews*. 2019;20(3):e158–e160 [6]

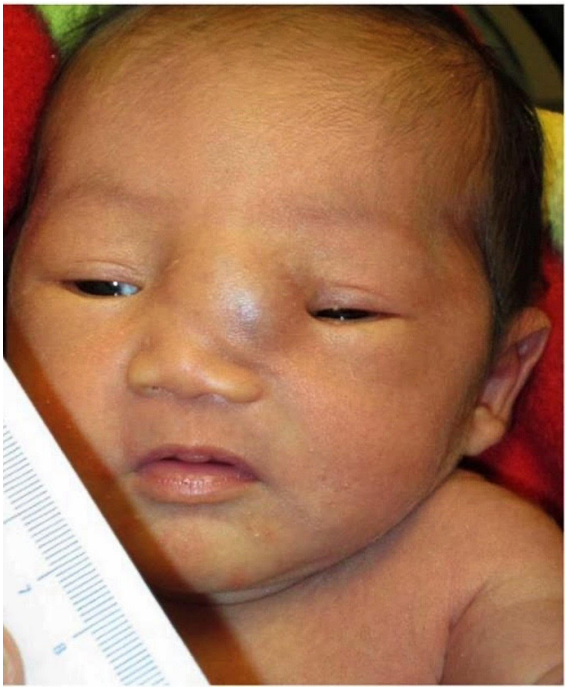

Figure 5: Photo of a left-sided nasal encephalocele in a neonate. Image from: Van Heukelom J. Case 3: an abnormal nose mass. *Neoreviews*. 2019;20(3):e158–e160 [6]

Did you know?

- Because CNPAS can be associated with chromosomal abnormalities and midline defects (eg, holoprosencephaly, solitary maxillary central incisor, and pituitary deficiencies), it is imperative that a thorough examination and workup, including brain magnetic resonance imaging and endocrinologic and genetic evaluations, are performed.[11]

What is unique about a neonate's anatomy and physiology that result in preferential nasal breathing in infants compared to adults?

To find the answer, please read the following articles:

- Goldstein NA, Gitman L. Neonatal nasal deformities. 2015;16(1):e36-e46[3]
- Bergeson PS, Shaw JC. Are infants really obligatory nasal breathers? *Clin Pediatr (Phila)*. 2001;40(10):567-569 [12]

March NeoQuest Authors

Allison N. J. Lyle, MD, MA, FAAP, University of Washington

Neena Jube-Desai, MD, MBA, FAAP, University of Maryland

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