Congenital Nasal Pyriform Aperture Stenosis

Priya P. Dmello BSN, RNC-NIC
Lehigh Valley Health Network, Priya_P.Dmello@lvhn.org

Jane M. Nemeth MSN, RNC-NIC
Lehigh Valley Health Network, Jane_M.Nemeth@lvhn.org

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Congenital Nasal Pyriform Aperture Stenosis

Priya Dmello BSN, RNC-NIC and Jane Nemeth, MSN, RNC-NIC
Neonatal Intensive Care Unit, Lehigh Valley Reilly Children's Hospital, Allentown, Pa.

This offering details the presentation, differential diagnosis and evidence-based interprofessional management of a neonate afflicted with Congenital Nasal Pyriform Aperture Stenosis (CNPAS) illustrated by a case study in a level IV neonatal intensive care unit (NICU).

CASE PRESENTATION

- Term female infant delivered via spontaneous vaginal delivery
  - Transferred to mother-baby unit
- Nasal congestion and increased work of breathing (WOB) unrelieved by suctioning at 2–3 hours of life (HOL); transferred to level II NICU
- Difficulty passing NGT on left nares
- Clinical presentation – Noisy breathing, respiratory distress, worsened with feeding/ improved with crying
  - Neonates are obligate nose breathers
  - In CNPAS – overgrowth of anterior bony opening of nose in facial skeleton causes airway obstruction
- ABG and CXR within normal limits (wnl)
- Required 3L HFNC; weaned to oxyhood with cool mist
- Day of life (DOL) 2 started on Neosynephrine nasal drops q12 hours
- DOL 3 – able to pass NGT bilaterally; however, continued increased WOB transferred to level IV NICU for higher level of care and ENT evaluation

DIAGNOSTIC FINDINGS

- ENT unable to pass 3.5mm flexible scope past anterior right nares
- CT scan face- confirmed CNPAS
- Discovered single central maxillary incisor (may be associated with pituitary abnormalities)
- Brain MRI – normal pituitary, no holoprosencephaly
- Pediatric Endocrine consult- often high association with growth hormone deficiency
  - Glucoses wnl x 24hrs
  - IGF-1 wnl
- ECHO in light of other midline anomalies – small VSD
  - Microarray wnl

INCIDENCE
1/50,000 live births

PATHOPHYSIOLOGY
Etiology unclear –
linked to chromosomal abnormalities

DIAGNOSTIC FINDINGS

- 65% of infants possess single central maxillary incisor
- May have other possible congenital anomalies

Increased incidence of:

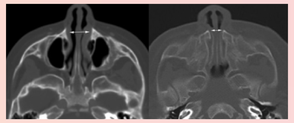
- Choanal atresia, pituitary insufficiency, growth hormone (GH) deficiency, microcephaly, intellectual disability, congenital heart disease, cleft lip and palate, hypopituitarism, ambiguous genitalia, short stature and holoprosencephaly

DIFFERENTIAL DIAGNOSIS

- Choanal atresia/stenosis
- Nasolacrimal mucocele
- Arhinia
- Bone dysplasia
- Neoplasm
- Encephalocele

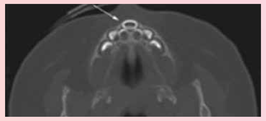
UNCOVERING THE DIAGNOSIS

- Inability/difficulty passing NGT or bedside scope
- CT scan:
  - Small nasal opening



Left - normal infant

Right - infant with CNPAS- narrowed opening of nasal aperture



Axial view showing single central maxillary incisor

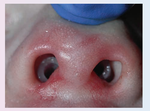
Yang, Sara et al. "Congenital nasal pyriform aperture stenosis in association with solitary median maxillary central incisor: Unique radiologic features." *Radiology Case Reports* vol. 11,3 178-81. 1 Aug. 2016, doi:10.1016/j.radcr.2016.06.004

STANDARD TREATMENT

- Recommend endocrine consult and baseline endocrine labs for pituitary evaluation
  - IGF-1, cortisol, FT4 and TSH
- Management – ENT consult
- Conservative therapy – suctioning, positioning, parenteral/ enteral support
  - More severe forms usually require surgical correction (Dilation and/or drilling, nasal stenting)
- Decongestants – often sufficient for patients with mild CNPAS
  - Nasal saline drops (gold standard)
  - Dexamethasone 0.1% ophthalmic solution intranasally (2nd line)
  - Phenylephrine 0.125% nasal drops (3rd line)

HOSPITAL COURSE

- Started on intranasal dexamethasone drops
- Airway positional requiring side-lying placement; improved by DOL 4, able to maintain O2 saturation levels supine
- D/c to home on DOL 5 to complete 10-day dexamethasone nasal drops course
- Continued to have nasal congestion and intermittent increased WOB
- 3 weeks of life (WOL) – underwent pyriform aperture stenosis drill out with dilation and stent placement
- Nasal stents removed on post-op day (POD) #2 and D/c to home with 10-day course of dexamethasone nasal drops
- 7 WOL – underwent nasal endoscopy with dilation and stent placement
- Stents removed on POD #2 – D/c home with dexamethasone drops and ofloxacin drops



Rajan, R., and Tunkel, D. (2018). Choanal atresia and other neonatal nasal anomalies. *Clinics in Perinatology*, Volume 45(4), 751 – 767.

NURSING PEARLS

- Watch for hypoglycemia, hypotension, conjugated/unconjugated hyperbilirubinemia
- Interprofessional collaboration is essential (speech, PT, OT, medicine-endocrine/ENT/Neonatology)
- Family support is crucial- need assistance with feeding, positioning, coordination of care and psychosocial support

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