Congenital Nasal Pyriform Aperture Stenosis

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CONGENITAL NASAL PYRIFORM APERTURE STENOSIS

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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, Difficulty passing NGT on left nares

• 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)

• Inability/difficulty passing NGT or bedside scope

• More severe forms usually require surgical correction (Dilation and/or drilling, nasal stenting)

• ABG and CXR within normal limits (wnl)

• Growth hormone deficiency

• Increased incidence of:
  - Choanal atresia, pituitary insufficiency, growth hormone (GH) deficiency
  - Congenital nasal pyriform aperture stenosis

• Bone dysplasia

• Nasal atresia

• Encephalocele

• Neoplasm

• Congenital anomalies

• Interprofessional collaboration is essential (speech, PT, OT, management)

• Family support is crucial – need assistance with feeding, positioning, coordination of care and psychosocial support

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

NURSING PEARLS

• Watch for hypoglycemia, hypotension, conjugated/unconjugated hyperbilirubinemia

• Interprofessional collaboration is essential (speech, PT, OT, medicine-endocrine/ENT/Nephrology)

REFERENCES

