

AMSER Case of the Month

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A 1-day-old female with increase work of breathing

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Patient Presentation

- **HPI:** Patient is a 1-day-old female with increased work breathing requiring intubation. Patient was referred to NICU with concerns of bilateral choanal atresia. History is limited as parents were not at the bedside.
- **Birth History:** Born at 37 weeks by C-section due to poor fetal heart tracing. The pregnancy was complicated by gestational hypertension. Following birth, patient had episodes of respiratory distress and was intubated at 1st hour of life.
- **Exam:**
 - **Nose:** Following intubation, a nasal endoscopy was performed but was unable to pass due to swelling and bleeding. A 6 mm nasogastric tube was passed through the left naris, but unable to be passed through the right naris

What Imaging Should We Order?

ACR Appropriateness Criteria

Variant 2:

Child. Persistent sinusitis (worsening course or severe presentation, or not responding to treatment), or recurrent sinusitis, or chronic sinusitis, or define paranasal sinus anatomy before functional endoscopic sinus surgery. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
CT paranasal sinuses without IV contrast	Usually Appropriate	⦿⦿⦿
CT paranasal sinuses with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT paranasal sinuses without and with IV contrast	Usually Not Appropriate	⦿⦿⦿⦿
MRI paranasal sinuses without and with IV contrast	Usually Not Appropriate	○
MRI paranasal sinuses without IV contrast	Usually Not Appropriate	○
Radiography paranasal sinuses	Usually Not Appropriate	⦿

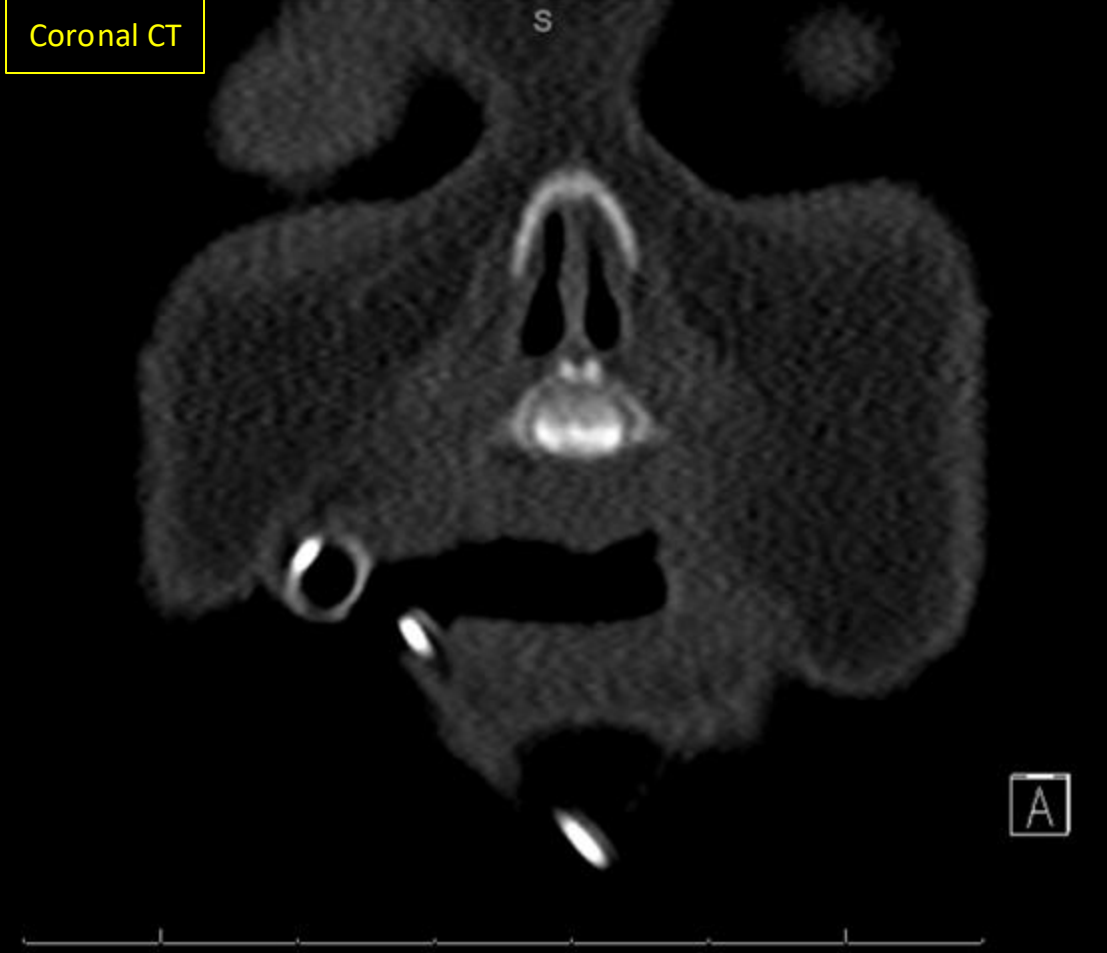
This imaging modality was ordered by the ENT physician

Findings (unlabeled)

Axial CT

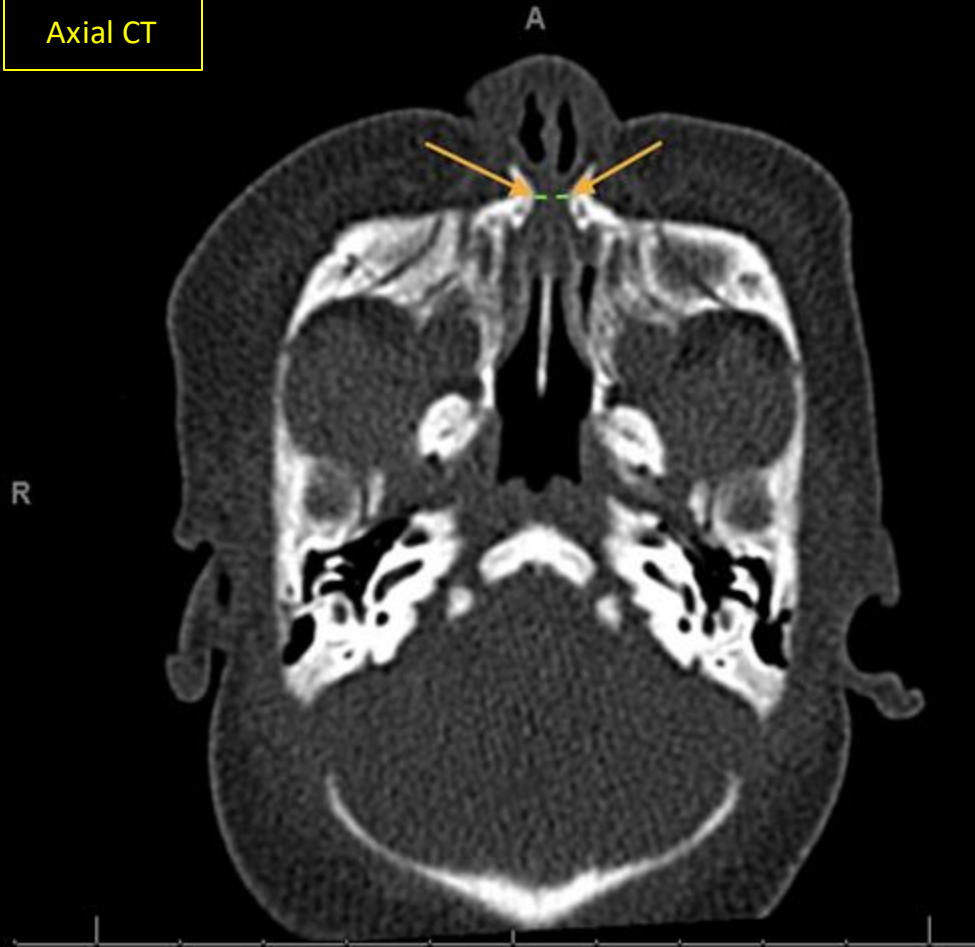


Coronal CT



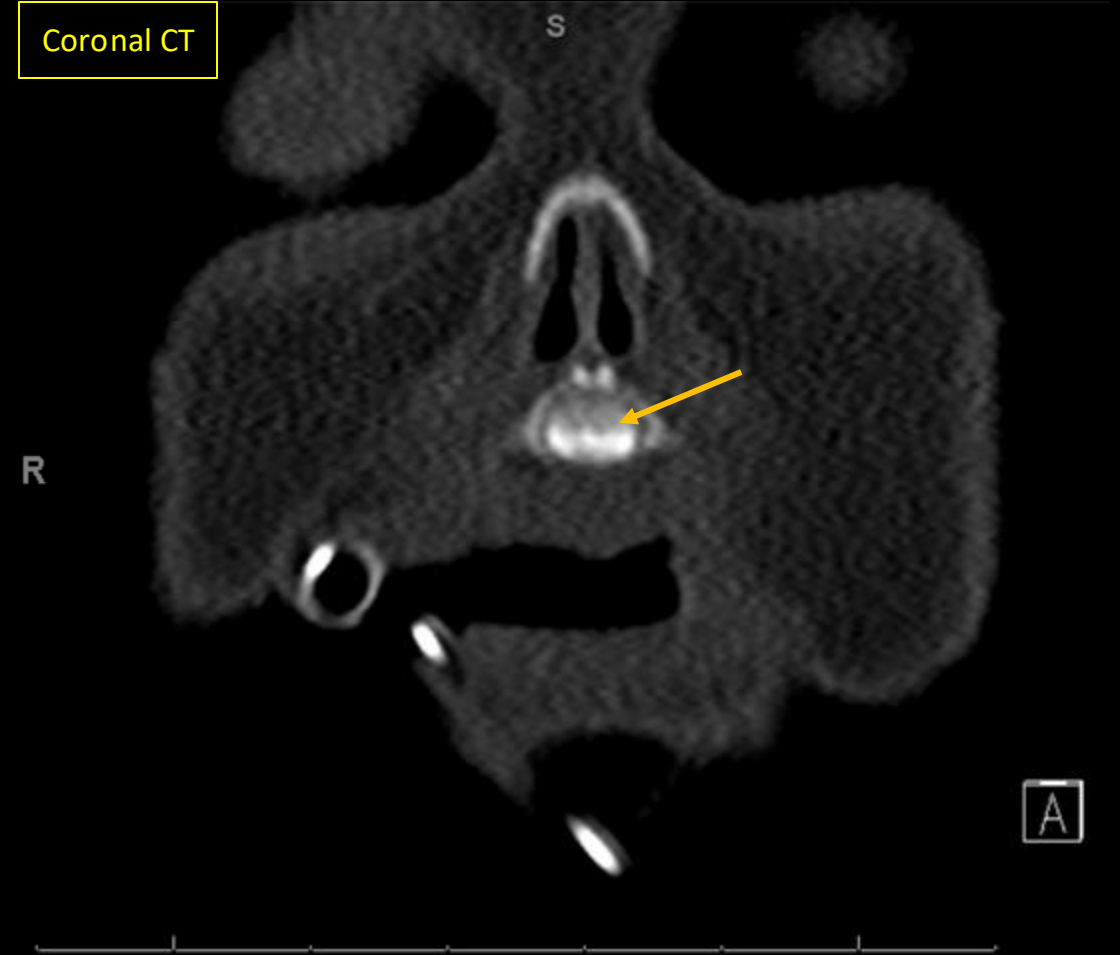
Findings (labeled)

Axial CT



Inward bowing and thickening of nasal processes of maxilla (**arrows**) resulting in pyriform sinus aperture stenosis measuring **5 mm**

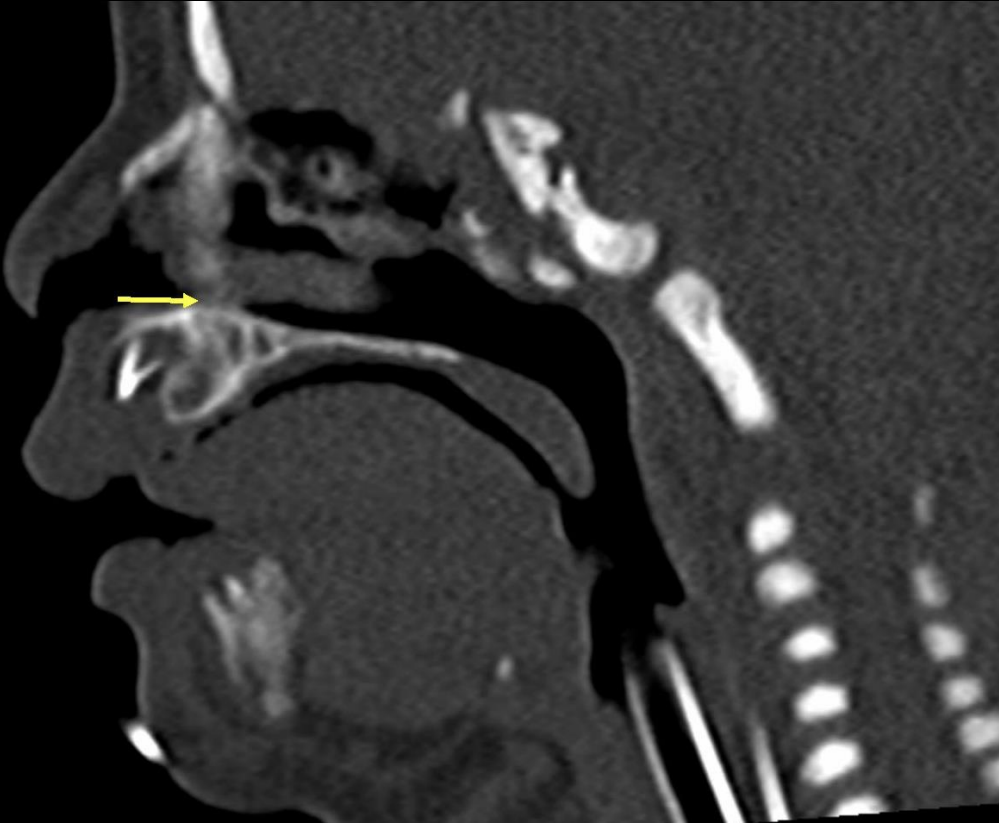
Coronal CT



Central megaincisor

Additional Views

Sagittal CT



3D Model (Anterior)



Pyriform aperture stenosis from sagittal view (left) and 3D maxillofacial configuration CT (Right)

Final Dx:

Pyriform Aperture Stenosis

Case Discussion

Etiology:

- Pyriform Aperture Stenosis (PAS) is caused by an overgrowth of the nasal process of the maxilla resulting in a narrowed nasal aperture.¹

Epidemiology:

- It is a rare, congenital condition often associated with other congenital anomalies. Although it's exact incidence is not well established, it may be approximated as 1 in 25,000 live births.² Maternal diabetes mellitus is a potential risk factor.³

Clinical Features:

- Neonates present with signs of upper airway obstruction, including cyclical cyanosis, feeding difficulties, and respiratory distress. A common diagnostic clue is the inability to pass a nasogastric tube through nasal passages.⁴

Case Discussion

Imaging Findings:

- PAS is diagnosed using thin-section CT imaging, which reveals a narrowed pyriform aperture—defined as less than 11 mm in a term neonate—due to medial overgrowth and inward bowing of the nasal processes of the maxilla. Associated findings may include a single central maxillary incisor, a midline palatal ridge, and a triangular-shaped hard palate. If a solitary central incisor is present, MRI of the brain is recommended to assess for midline anomalies such as holoprosencephaly.^{7,8}

Management:

- For mild stenosis, nasal stenting may improve symptoms. Severe stenosis requires surgical enlargement of the pyriform aperture once child is large enough to be able to tolerate the surgery. After surgery, the relatively small aperture may increase likelihood of developing obstruction with upper respiratory infections.^{5,6}

References:

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