

MR CASE STUDY: MR's Ability to Diagnose Clinically Significant Congenital Nasal Passage Airway Obstruction in Fetuses Referred for Seemingly Unrelated Intracranial Observations

The current cases correlate postnatal CT and MR findings with retrospectively established fetal MR observations.

A PubMed literature review using the search terms, fetal MRI, fetal nasal passage, choanal atresia and pyriform aperture stenosis identified no publications.

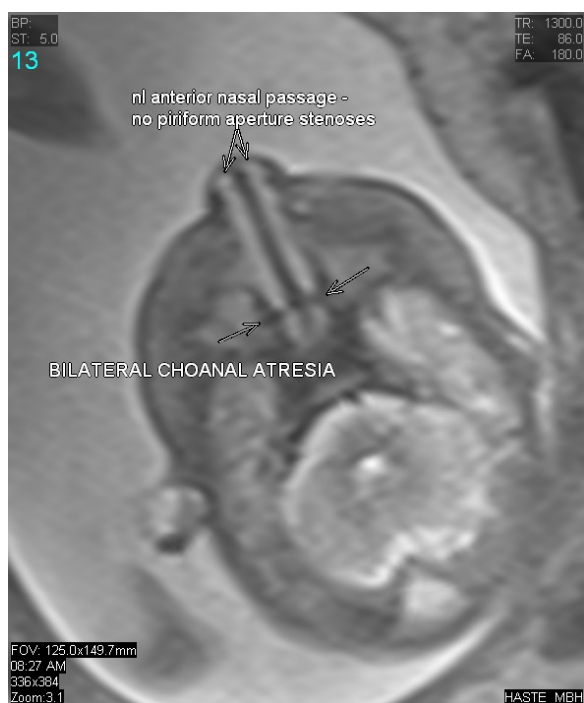
CASE 1: Choanal atresia with CHARGE association. This pregnancy was referred to us at 29 weeks for outside ultrasound studies that demonstrated mild ventriculomegaly in a patient that had declined karyotyping. Negative maternal serum TORCH titers.

MR demonstrated mild ventriculomegaly and non-normalization of the cavum septum. Neonatal nasal congestion and difficulty feeding prompted ENT consultation and imaging. Postnatal CT and MR identified bilateral choanal atresia and a hypoplastic posterior corpus callosum. Neuroophthalmology exam demonstrated right optic nerve and retinal coloboma and left optic nerve atrophy. The constellation of these findings was felt to be compatible with CHARGE association. Choanal atresia necessitated neonatal surgical correction prior to discharge.

(CHARGE association: **C**oloboma of iris, **H**heart defect, **C**hoanal **a**tresia, **I**nter uterine growth **r**estriction, **G**enital anomalies and **E**ar anomalies.)

Retrospective review of the fetal MR demonstrates findings consistent with bilateral choanal atresia.

Arrows in image **1A** point to the fetal bilateral bony (dark) narrowing of the parallel fluid-filled (white) nasal passage. Arrows in image **1B** point to the corresponding postnatal CT findings.

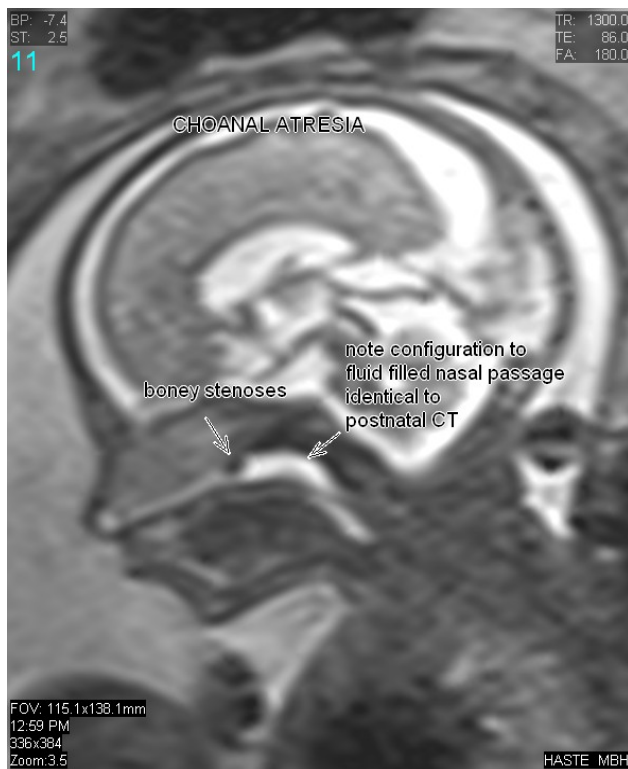


1 A



1 B

Sagittal fetal MR image **2A** and postnatal sagittal CAT scan image **2B** corroborate this imaging diagnosis.



2 A



2 B

CASE 2: Final Diagnosis – Nasal passage obstruction due to pyriform aperture stenosis and basal frontal encephalocele with dermoid requiring intubation and tracheostomy. .

HISTORY: 30 week twin gestation referred for Twin B mild ventriculomegaly with non-normalization of the corpus callosum on fetal ultrasound. The pregnancy was the result of IVF with normal twin karyotype by CVS.

Fetal MR confirmed complete agenesis of the corpus callosum and demonstrated no other identifiable intracranial abnormalities.

The twin with agenesis of the corpus callosum (ACC) had difficulty breathing and feeding, requiring intubation and then tracheostomy. Postnatal MR and CT for the evaluation of upper airway issues identified two sources of nasal passage obstruction – pyriform aperture stenosis (PAS) and a small 6 x 4 mm anterior nasal encephalocele with an accompanying fatty dermoid component. Clinical exam and imaging demonstrated a large single midline maxillary incisor – a frequent accompaniment to PAS.

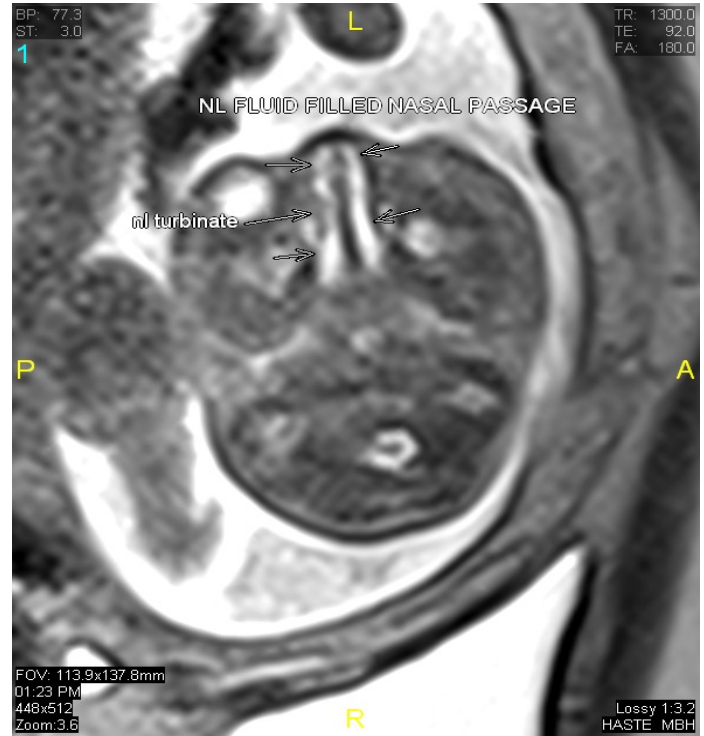
Pyriform aperture stenosis (PAS) creates an obstruction to the anterior aspect of the nasal passage (in comparison to choanal atresia which is obstruction to the posterior aspect of the nasal passage).

Retrospective review of fetal MR findings demonstrates evidence of the postnatally detected clinical and imaging abnormalities.

Arrows in image **3A** demonstrate the abnormal fetal mid to anterior nasal passage – abnormal due to the absence of the parallel white lines of fluid that are usually present, extending from anterior to posterior, as seen on image **3B**, a normal fetal nasal passage, for comparison.



3 A



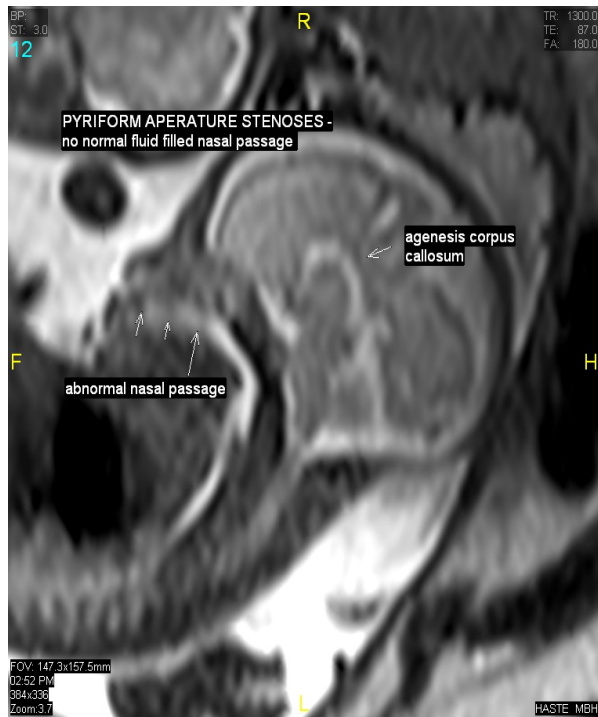
3 B

Image **3C** is the corresponding abnormal postnatal CT of this patient, demonstrating marked crowding and effacement of air column within the anterior nasal passage.



3 C

Image **3D** is the abnormal fetal nasal passage in the sagittal plane of section – note the absence of a normal white fluid-filled nasal passage in comparison to a normal sagittal fetal nasal passage as demonstrated on image **3E**.

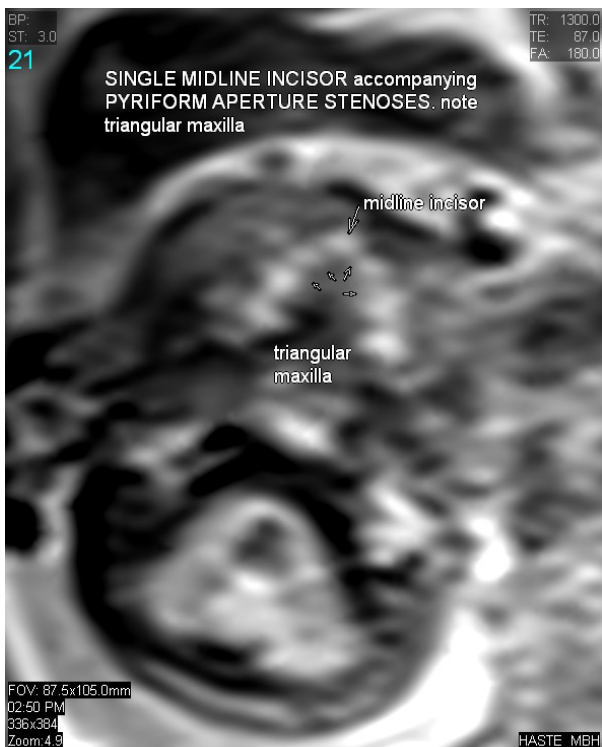


3 D

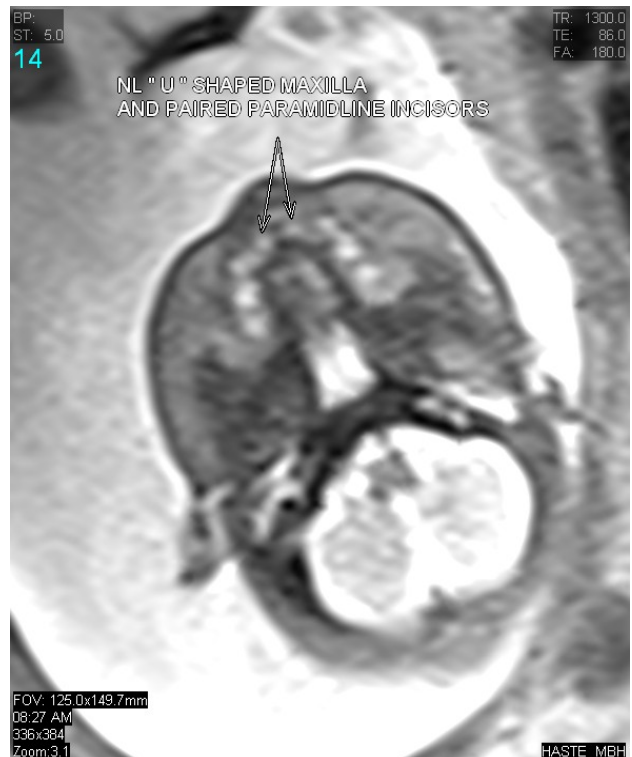


3 E

Image **4A** is the axial fetal MR view of the maxilla, demonstrating the single midline maxillary incisor and the typical triangular shape of the maxilla that accompanies pyramidal aperture stenosis. Image **4B** is the normal fetal MR appearance of the maxilla for comparison – note the “u-shaped” maxilla and the normal paired para-midline maxillary incisors.

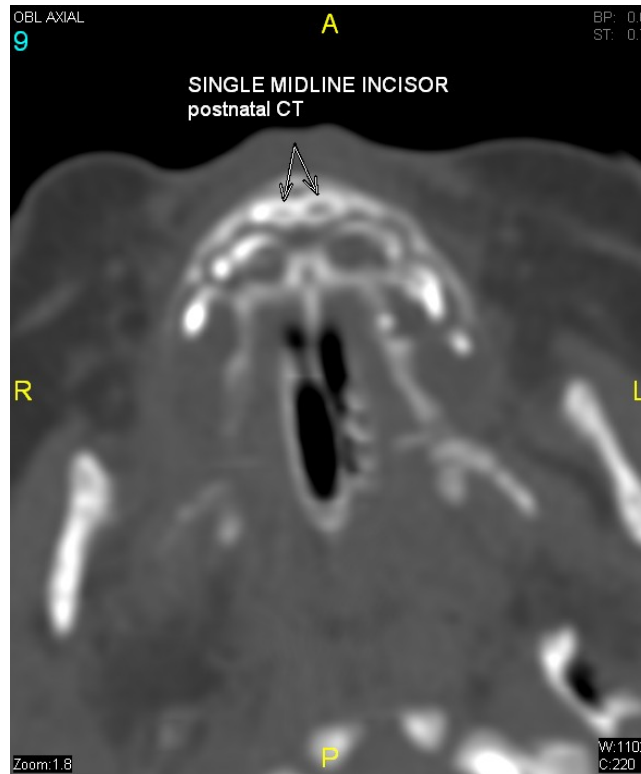


4 A



4B

Image 4C is the postnatal CT showing the large single maxillary incisor.



4 C

The second component of nasal passage compromise was the encephalocele and accompanying dermoid. Image 5A is the abnormal sagittal T2 fetal MR image. The soft tissue between the arrows denotes the downward herniation of the encephalocele. Note the focal obscuration of the white CSF and the black bone forming the floor of the anterior cranial fossa. Image 5B is an example of the normal fetal MR appearance to the floor of the anterior cranial fossa.

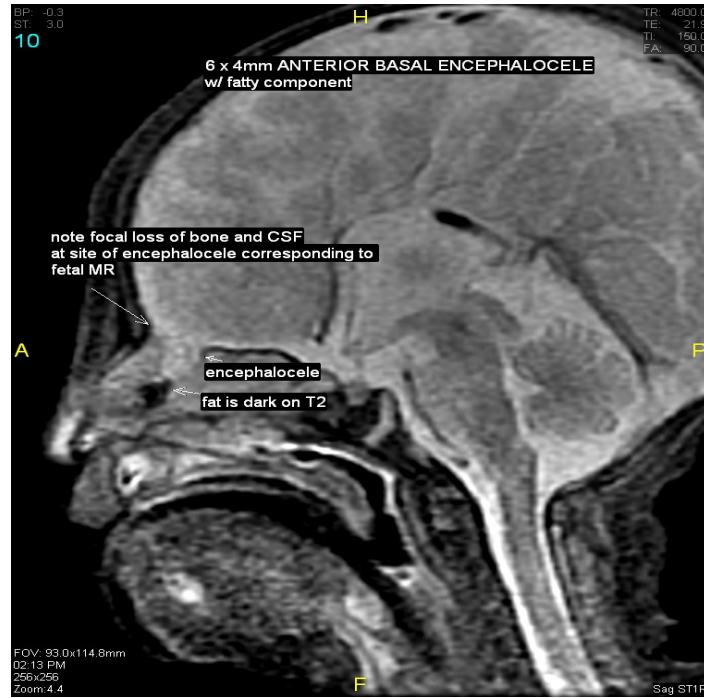


5 A



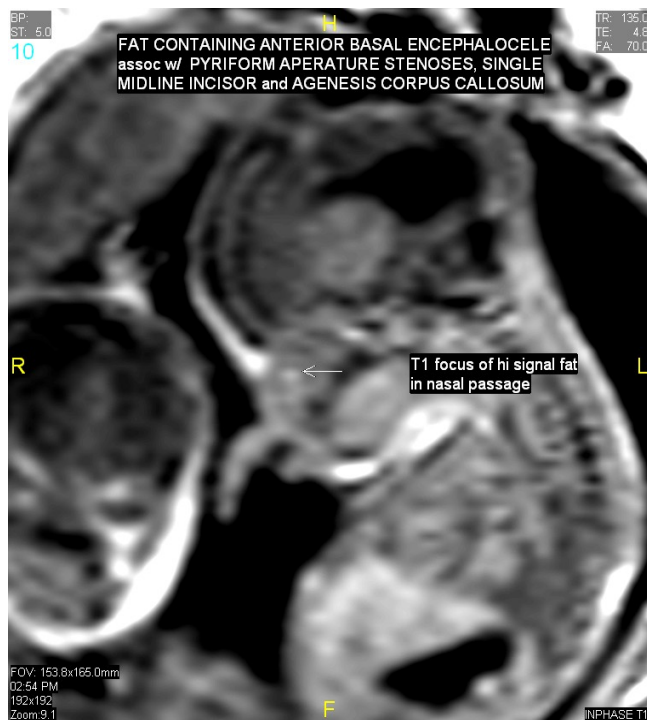
5 B

Image **5C** is the postnasal T2 MR sequence showing the encephalocele and the herniated dark focus of dermoid fat indicated by arrows.

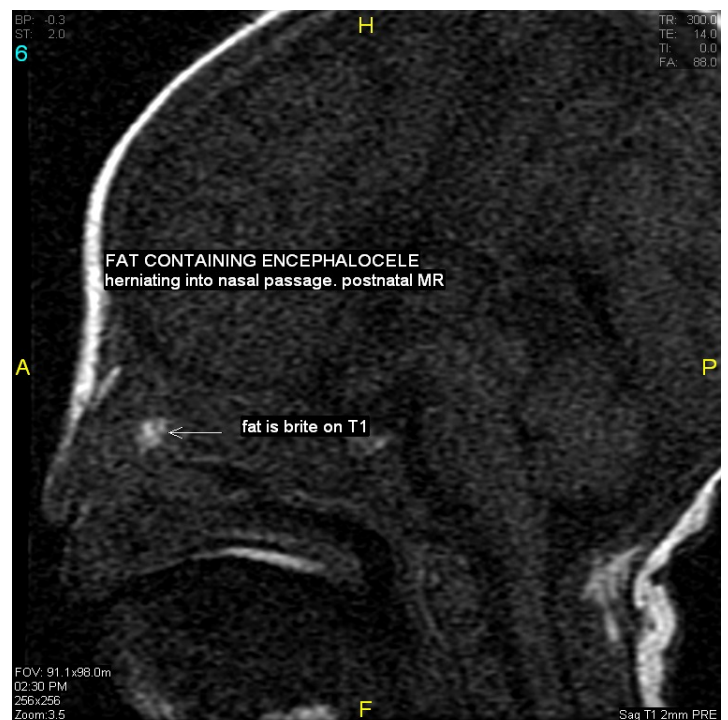


5 C

Image **5D** is the corresponding fetal sagittal T1 sequence demonstrating abnormal hypersignal intense (bright white) focus of dermoid fat in the nasal passage (fat is bright on T1 and dark on T2). Image **5 E** is the corresponding postnatal sagittal T1 MR identifies the identical focus of dermoid fat as a bright white mass in the nasal passage.



5 D



5 E

SUMMARY:

1. The presented material demonstrates MR's ability to identify congenital stenosis of the nasal passage – unsuspected on antenatal ultrasound – in patients who are referred for intracranial ultrasound observations.
2. The identification of nasal passage pathology can be an important clue to syndromic conditions such as CHARGE association. Other pathologies associated with CHARGE syndrome that can be uniquely detected on MR included coloboma, absence of olfactory sulci (whose MR detection would be gestational age dependent), and abnormalities of the inner ear.

A partial list of other syndromes in which choanal atresia can be seen include Cerebro costo mandibular syndrome, Treacher Collins syndrome; Ectrodactyly – Ectodermal Dysplasia – Clefting, (EEC) syndrome, Craniosynostosis syndromes and Coumadin exposure.

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