

# Rare Combination of Frontonasal and Bilateral Naso-orbital Encephaloceles

Alan A. Alexander<sup>1\*</sup>, Megan R. Saettele<sup>2</sup>, Daniel L'Heureux<sup>1</sup>, Paras A. Shah<sup>3</sup>,  
Kristin A. Fickenscher<sup>4</sup>

1. Department of Radiology, Georgetown University Hospital, Washington, USA

2. Department of Radiology, University of Missouri- Kansas City, Kansas City, USA

3. Department of Radiology, Barrow Neurological Institute, Phoenix, USA

4. Department of Radiology, Children's Mercy Hospital, Kansas City, USA

\* **Correspondence:** Dr. Alan A. Alexander, Georgetown University Hospital, Department of Radiology, 3800 Reservoir Road, Washington, D.C. 20007, USA  
(✉ [aaz.alexander@yahoo.com](mailto:aaz.alexander@yahoo.com))

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## ABSTRACT

Encephaloceles, while a common entity affecting 1:4000 live births, typically occur in the occipital region. Encephaloceles involving the frontal region comprise only 15% of all cases. Naso-orbital encephaloceles are rarely seen. Our case profiles a child born at term with an atrial septal defect (ASD), micrognathia, cleft lip, and frontonasal as well as bilateral naso-orbital encephaloceles. At birth the encephaloceles were undetected. During the cleft palate pre-operative preparation, the bilateral naso-orbital encephaloceles were diagnosed as dacryocystoceles for which the child underwent surgical repair. Misdiagnosis and loss to follow up lead to delayed surgical treatment until the child was almost two years of age; the right eye was near complete closure due to the increasing size of the encephalocele. This case highlights the importance of meticulous radiologic interpretation of midline nasal masses, as a correct diagnosis impacts clinical management and directs surgical repair.

## CASE REPORT

### CASE REPORT

A four-day-old full term male infant initially presented to University Hospital with wide complete bilateral cleft lip and palate, and a protuberant premaxilla and prolabium. At the time of presentation, an atrial septal defect was also found by echocardiogram. Between three months and one year of age, he underwent multiple surgeries for cleft lip and palate repair including the placement of a Latham device and cleft lip adhesion. He was misdiagnosed with having bilateral dacryocystoceles which were repaired with Crawford tubes. Cytogenetic analysis was performed and was negative for any chromosomal abnormalities. The patient also had recurrent episodes of rash on his arm and a short hospitalization for

pneumonia. By one year of age the patient's ASD had closed spontaneously.

A few months later, during clinical evaluation, he was noted to have trigonocephaly (Figure 1), hypertelorism, epiphora, and soft tissue masses at the medial aspect of the orbits. Computed tomography scanning of the head demonstrated the presence of a large bony defect across the nasal bridge, as well as bilateral medial orbital fossa mixed density masses (2.5 cm on the right and 2.3 cm on the left) with possible communication with the gyrus rectus with bilateral encephaloceles extending into the medial orbital wall. Additional bony abnormalities included a low lying cribriform plate with metopic craniosynostosis. MRI was then

performed, which confirmed the presence of herniated gray matter through these bilateral bony defects as well as a midline frontonasal encephalocele. Abnormal T2 signal was noted in the inferior aspect of the frontal lobes (Figure 2).

The patient was scheduled to undergo surgical repair of frontonasal and bilateral orbital encephaloceles, as well as reconstruction of his metopic craniosynostosis. Over the course of approximately one year there were multiple instances in which the patient did not show up for scheduled appointments, however, he was eventually cleared for surgery by both consult services. At nearly two years of age, the patient's right eye was almost completely swollen shut which was notably worse after the patient had been lying down. There was prominent swelling of the nasal bridge from one medial canthus to the other (Figure 3). A repeat CT scan and MRI were performed. The CT scan re-identified the bilateral bony defect in the medial orbital walls which had increased, measuring 4.9 mm on the right and 4.6 mm on the left, in which brain was noted to exit through into the orbit (Figure 4). The abnormal brain tissue running posteriorly in the orbit exerted a mild to moderate mass effect on the medial rectus muscle and the globe (Figure 5), additionally the anterior frontal lobe extended through the low lying cribriform plate. MRI demonstrated the bilateral defect in the medial orbital walls, as well as the 5x8 mm frontonasal encephalocele (Figure 6).

At 23 months of age, surgical repair was performed by a facial plastic and neurosurgical team in an extensive operation that involved resection of the patient's bilateral orbital encephaloceles, reconstruction of the anterior cranial fossa with a split-thickness calvarial bone graft, fronto-orbital reconstruction, nasal reconstruction with a split-thick thickness calvarial bone graft, resection of bilateral nasal processes with maxillary and medial orbital bone contouring, bilateral dacryocystorhinostomy, medial canthoplasties, a pericranial flap, and a forte del cantho procedure. The patient tolerated the procedure well and had an uneventful post-operative period. Post operative MR scan demonstrated complete repair of the anterior encephaloceles (Figure 7). At 10 weeks following surgery, the patient was doing well.

## DISCUSSION

This case demonstrates the challenges a young child with a midline frontonasal mass poses to clinicians. Misdiagnosis and delays in follow up can be common, and also detrimental to the child's health. This case also highlights radiologic features that help to define anterior encephaloceles, as correct imaging assessment is crucial to the diagnosis, clinical management, and treatment of these patients. Our case profiles a child born at term with ASD, micrognathia, cleft lip, and frontonasal as well as bilateral naso-orbital encephaloceles.

While common congenital lesions occur in 1:4000 live births, encephaloceles typically occur in the occipital region [1]. The term encephalocele refers to the herniation of brain

tissue through a skull defect, and is classified as fronto-ethmoidal (sincipital), posterior (occipital), basal, or parietal, depending on where the skull defect is located. The anterior or fronto-ethmoidal encephalocele accounts for only 15% of all encephalocele cases and can be further subdivided into fronto-nasal (40-60%), naso-ethmoidal (30%), and naso-orbital. A combination of both frontonasal and naso-orbital encephaloceles is rare.

In this case, we presented a two-year-old male who underwent multiple imaging studies and surgical repair of both frontonasal and bilateral naso-orbital encephaloceles. Anterior (fronto-ethmoidal) encephaloceles can occur with aberrant development of the frontonasal region or the anterior neuropore [1]. In addition to encephaloceles, nasal dermal sinus or glioma can also result. Making the correct diagnosis and determining the depth of intracranial extension is important to guide clinical decisions.

Development of a sincipital (anterior) encephalocele begins around the fourth week of gestation [1]. If a disturbance in the separation of neural and surface ectoderm occurs at the closure site of the rostral neuropore during neurulation then this will lead to a midline mesodermal defect. Brain tissue and epidermis may then herniate through the defect. In the case of a frontoethmoidal encephalocele the brain tissue is able to herniate through the foramen cecum and into the prenasal space.

An anterior encephalocele is suspected at birth when an infant presents with a visible nasal or orbital mass [2]. Broadening of the nasal root and hypertelorism are often present as well. The soft, sometimes pulsatile mass characteristically enlarges with crying, the Valsalva maneuver, and with jugular compression (positive Furstenberg sign). The differential diagnosis of a midline nasal mass includes encephalocele, nasal glioma, dermoid cyst, epidermoid cyst, hemangioma, lymphangioma, dacryocystocele, and dacryocystitis. Imaging is often essential to differentiate between each of the entities [3]. For instance, a dacryocystocele, which was the misdiagnosis first given to the patient, has features that include nasolacrimal duct dilatation and a well-defined, thin-walled mass, with fluid attenuation involving the medial canthus or nasal cavity. Imaging features of nasal encephaloceles, on the other hand, display a soft-tissue mass in connection with the subarachnoid space via an enlarged foramen cecum [3]. If there is no mass present, the diagnosis of an encephalocele becomes even more difficult and may not be detected clinically at birth. Symptoms such as persistent nasal congestion and rhinorrhea may lead a clinician to have imaging performed where the defect is then found.

In addition to initial imaging necessary for diagnosis, pre-operative imaging is essential to determine the type and extent of the lesion. Various imaging modalities can help to give details about the size of the bony defect and also to look for the major vascular supplies to that area of the brain. It is especially important to locate the torcula and superior sagittal sinus in cases with posterior midline encephaloceles, and equally important to look for other associated brain anomalies that may alter a child's prognosis [3]. Anomalies associated

with encephaloceles include intracranial cysts, callosal agenesis, interhemispheric lipomas, facial clefts, and schizencephaly.

Computed tomography (CT) imaging is important for defining the bony anatomy and determining the size of the defect, but caution must be used when interpreting newborn, infant, or young children's images as the unossified nasal process of the frontal, nasal, and ethmoid bones all have CT attenuation similar to brain and nasal cartilage in the first 6-8 months of life [1]. This can lead to the false interpretation of a bony defect where one is not actually present. Magnetic Resonance (MR) imaging then is the test of choice when one suspects an anterior encephalocele. The herniated tissue in question should demonstrate continuity with the brain. T1-weighted imaging will show hypointense CSF and tissue that is isointense to the brain's grey matter. T2-weighted images will show changes of gliosis within the tissue and bright CSF signal. MR imaging is also superior because it allows for the detection of any associated anomalies.

Correct imaging interpretation and diagnosis will guide the necessary reconstructive surgery. Surgical repair is important to prevent further enlargement of the skull defect and a subsequent increase in brain herniation that can occur with age [4]. Surgery reduces herniated intracranial contents back to their normal location within the calvarium, repairs the abnormal dura, corrects any bony defects, restores aesthetic facial appearance, and can also prevent CSF leakage or meningitis that may occur as a result of an enlarging encephalocele. If treated, normal development occurs in approximately 60% of cases. The prognosis is influenced by anatomical site, volume of neural contents, and presence of coexisting malformations [5].

It is important to keep encephaloceles in the differential diagnosis when a child presents with a nasal or orbital mass. A skilled otolaryngologist and radiologist are helpful in assisting to making the correct diagnosis. Misdiagnosis and delays in follow up can unnecessarily prolong corrective surgery and may lead to further complications. CT and MR imaging are the modalities of choice for pre-operative determination of lesion extent.

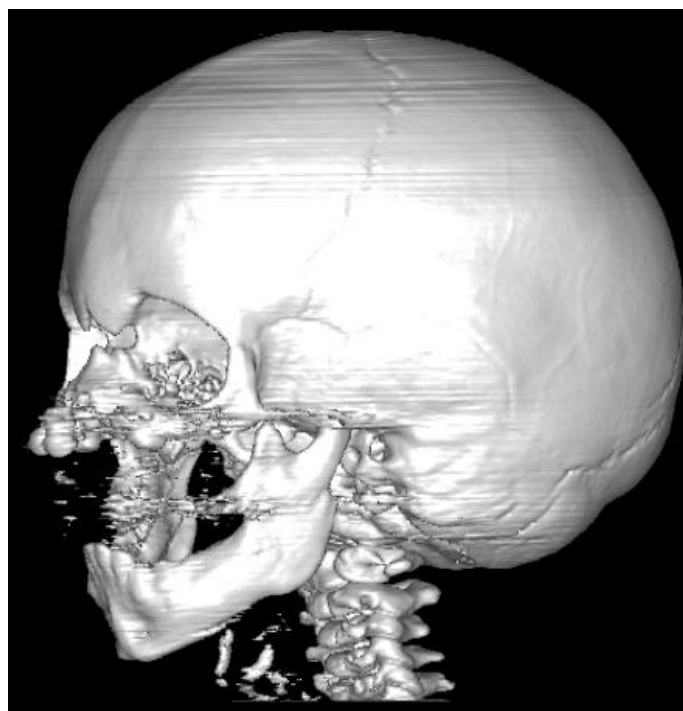
TEACHING POINT

It is important to keep encephaloceles in the differential diagnosis when a child presents with a nasal or orbital mass. Prompt surgical correction is necessary and can improve a child's prognosis.

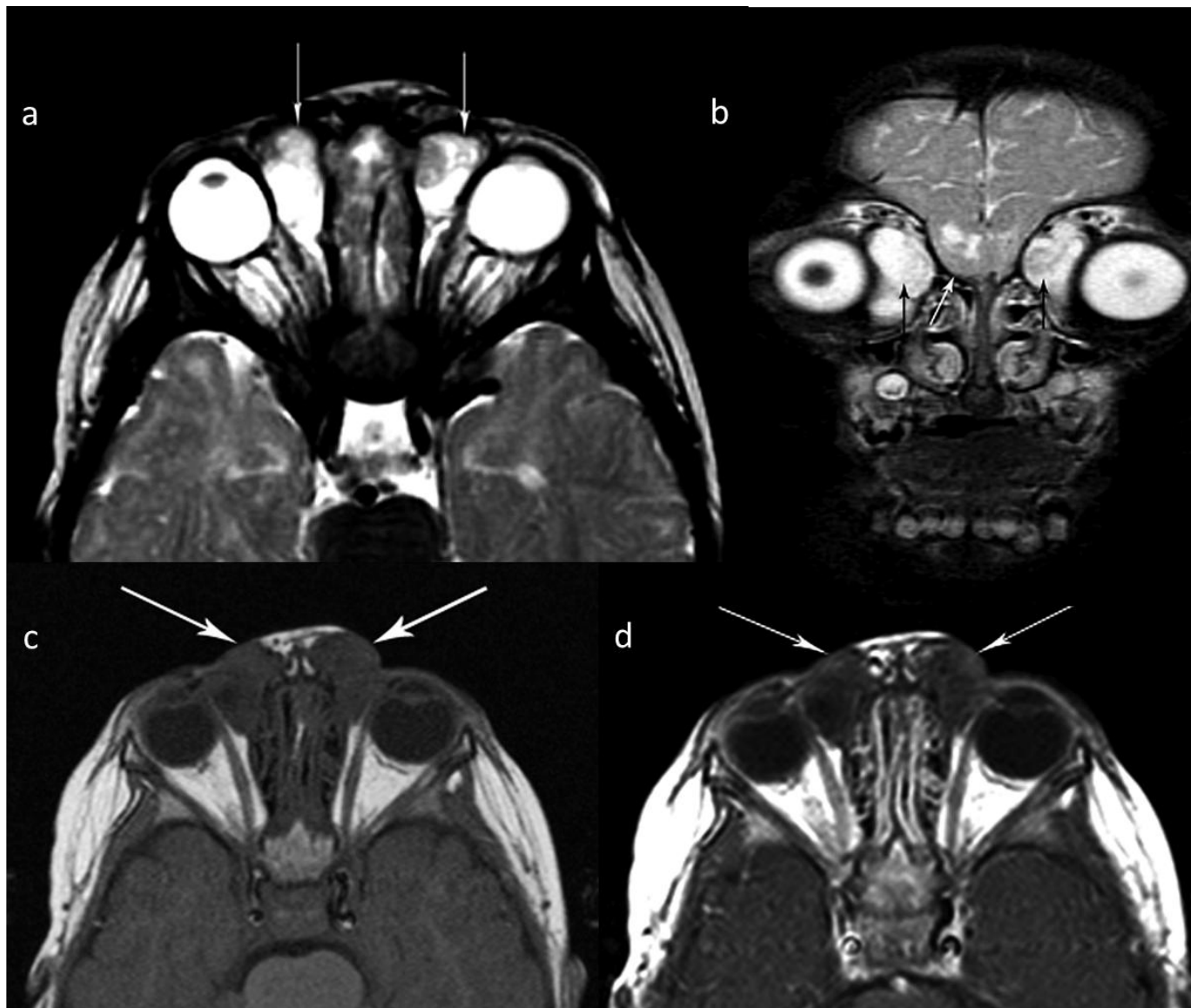
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FIGURES



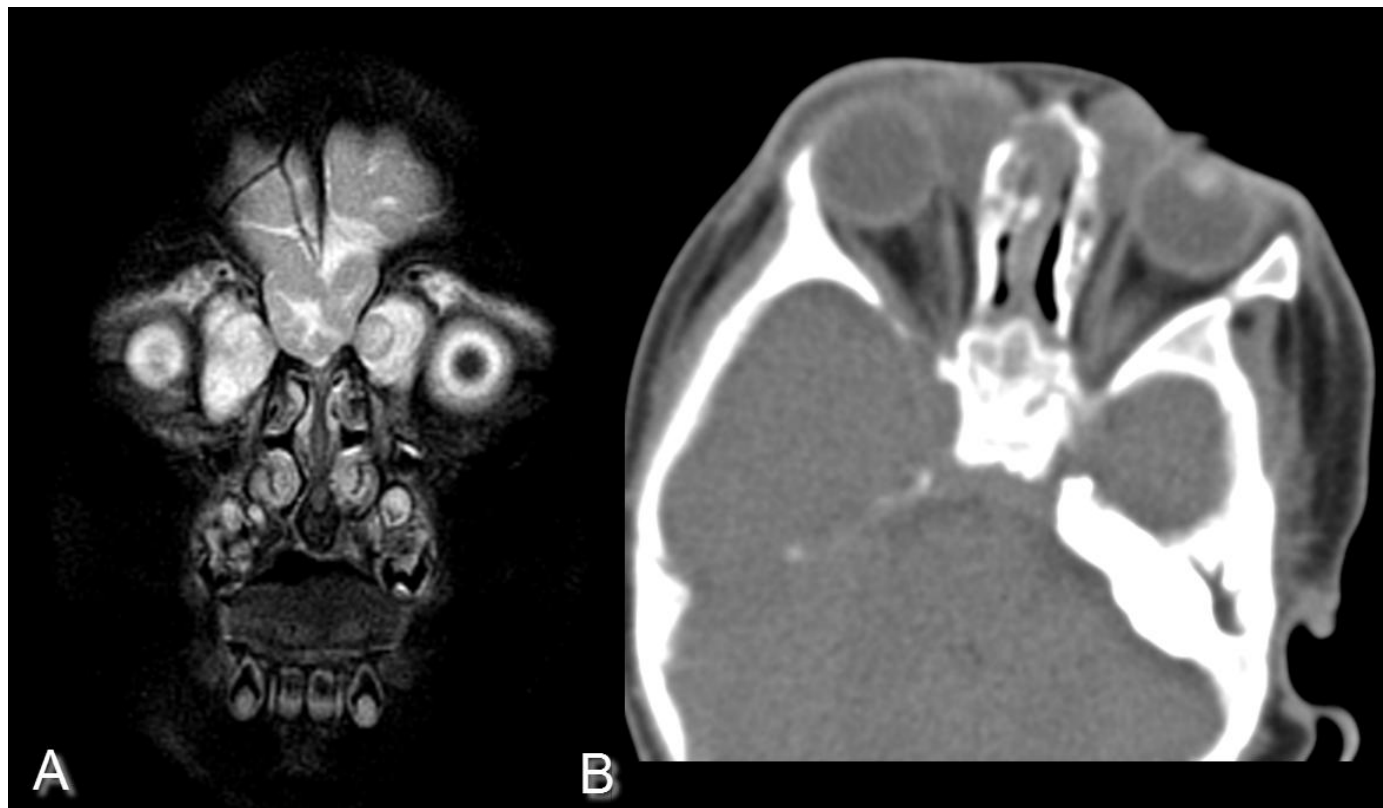
**Figure 1:** Approximately 1 year old male with bilateral naso-orbital encephaloceles and fronto-nasal encephalocele. 3D surface reconstruction from CT (kvp 120, mAs 380) shows fusion of the metopic suture with trigonocephaly and beaking of the frontal bone along the midline.



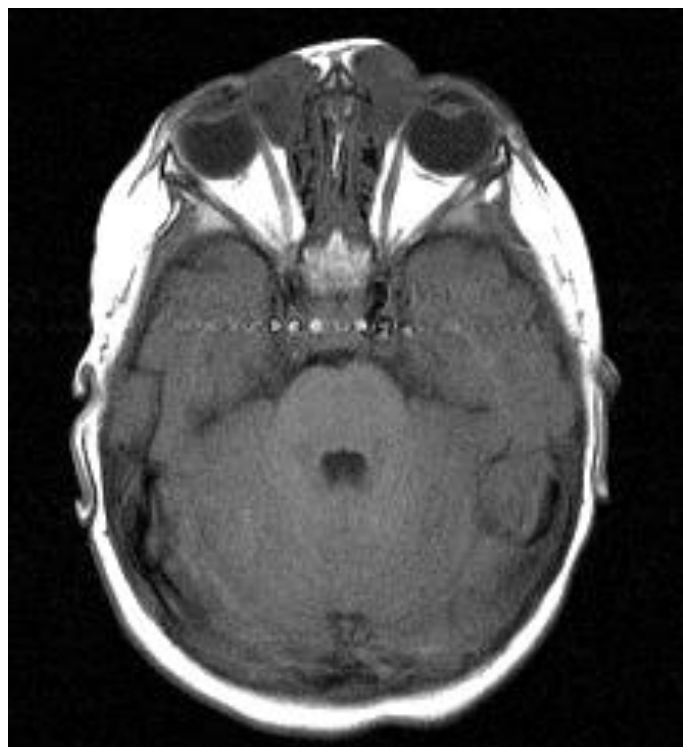
**Figure 2:** Approximately 1 year old male with bilateral naso-orbital encephaloceles and fronto-nasal encephalocele. **a:** Axial T2 MRI (TR 4250, TE 81.39) of the brain at the level of the orbits depicts the bilateral naso-orbital encephaloceles (solid arrows). **b:** Coronal T2 MRI (TR 4250, TE 81.39) of the brain at the level of the orbits depicts the bilateral naso-orbital encephaloceles (black arrows) as well as abnormal T2 signal noted in the inferior aspect of the right frontal lobe (white arrow). **c:** Axial T1 (TR 700, TE 17) pre contrast non fat-saturated MRI of the brain at the level of the orbits depicts the bilateral naso-orbital encephaloceles (white arrows). **d:** Axial T1 (TR 700, TE 17) post contrast non-fat saturated MRI of the brain at the level of the orbits depicts the bilateral naso-orbital encephaloceles (white arrows).



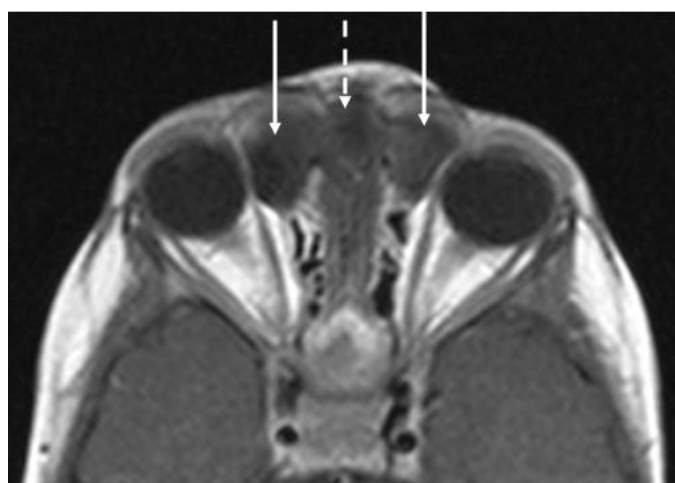
**Figure 3 (left):** Photograph of approximately 2 year old male with bilateral naso-orbital encephaloceles and frontal encephalocele. Photograph demonstrates prominent swelling of the nasal bridge with compression and swelling medial to the right epicanthal fold, obtained prior to surgery.



**Figure 4:** Approximately 2 year old male with bilateral naso-orbital encephaloceles and fronto-nasal encephalocele. a) Coronal T2 weighted MR (TR 4400 TE 89.41) demonstrates bilateral defects of the medial orbital wall with associated bilateral encephaloceles. b) Axial CT (kvp 120, mAs 380) in soft tissue windows demonstrates bilateral masses within the medial orbital canal which are consistent with the patient's known diagnosis of bilateral encephaloceles.



**Figure 5:** Approximately 2 year old male with bilateral naso-orbital encephaloceles and fronto-nasal encephalocele. Axial T1 weighted MR (TR 700 TE 17) without contrast demonstrates moderate mass effect (right greater than left) of the encephaloceles on the medial rectus muscle and the globe.



**Figure 6:** Approximately 2 year old male with bilateral naso-orbital encephaloceles and fronto-nasal encephalocele. Axial T1 MRI (TR 700 TE 17) of the brain at the level of the orbits depicts the bilateral naso-orbital encephaloceles (solid arrows) as well as the midline fronto-nasal encephalocele (dashed arrow).



**Figure 7 (left):** Approximately 2 year old male with bilateral naso-orbital encephaloceles and fronto-nasal encephalocele. Post-operative Axial T1 MRI (TR 700 TE 17) of the brain demonstrating repair.

<b>Etiology</b>	Congenital- herniation of brain tissue
<b>Incidence</b>	0.8-4:10,000 live births
<b>Gender Ratio</b>	1:1
<b>Age Predilection</b>	Birth
<b>Risk Factors</b>	n/a
<b>Treatment</b>	Surgical resection
<b>Prognosis</b>	Normal development in approximately 60% of cases
<b>Findings of imaging</b>	Hypointense on T1, Hyperintense on T2

**Table 1:** Summary table for encephalocele

	<b>Encephalocele</b>	<b>Dermoid</b>	<b>Epidermoid cyst</b>	<b>Hemangioma</b>	<b>Lymphangioma</b>	<b>Dacryocystocele</b>	<b>Dacryocystitis</b>	<b>Glioma</b>
<b>X-Ray</b>	Soft tissue density, possible sinus opacification	Soft tissue density, possible sinus opacification	Soft tissue density, possible sinus opacification	Soft tissue density, possible sinus opacification	Soft tissue density, possible sinus opacification	Soft tissue density, possible sinus opacification	Soft tissue density, possible sinus opacification	Soft tissue density, possible sinus opacification
<b>US</b>	Fluid filled mass	Increased echogenicity	Fluid filled mass	Hypoechoogenic with heterogenous echotexture	Hypoechoogenic	Fluid filled mass	Enlargement of lacrimal sac	Solid appearing structure
<b>CT</b>	Hypodense mass	Hypodense, less dense than fluid.	Hypodense	Hyperintense	Hypodense	Homogenous, well-defined, fluid attenuated	Hypodense	Varies; isodense or hypodense to surrounding tissue
<b>MRI-T1</b>	Hypointense	Classically Hyperintense	Hypointense	Hypointense	Heterogenous	Hypointense	Variable	Variable
<b>MRI-T2</b>	Hyperintense	Classically Hypointense	Hyperintense	Hyperintense	Heterogenous	Hyperintense	Hyperintense	Variable
<b>MRI-DWI</b>	Not diffusion restricting	Not diffusion restricting	Not diffusion restricting	Not diffusion restricting	Not diffusion restricting	Not diffusion restricting	Diffusion restricting	Not diffusion restricting
<b>Pattern of contrast enhancement</b>	Rim of enhancement to homogeneous enhancement	Rim of enhancement	Rim of enhancement	Variable enhancement depending on vascularity	Does not enhance post-contrast	Slight rim of enhancement	Variable enhancement depending on vascularity	Avid to heterogeneous
<b>Scintigraphy</b>	Variable	Variable	Variable	Increased activity with technetium labeled RBCs	Variable	Variable	Increased activity with technetium labeled WBCs	Variable
<b>PET</b>	Same as brain metabolic activity	No metabolic activity	No metabolic activity	Increased metabolic activity	No metabolic activity	No metabolic activity	Increased metabolic activity	Increased metabolic activity

**Table 2:** Differential diagnosis table for encephalocele

**ABBREVIATIONS**

CSF = Cerebral Spinal Fluid  
 MR = Magnetic Resonance  
 CT = Computed Tomography

**KEYWORDS**

Encephalocele; Neuroradiology; Pediatric radiology; frontonasal; naso-orbital

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