PEERING INTO P.E.I.R.
(Persisting Embryonal Infundibular Recess)

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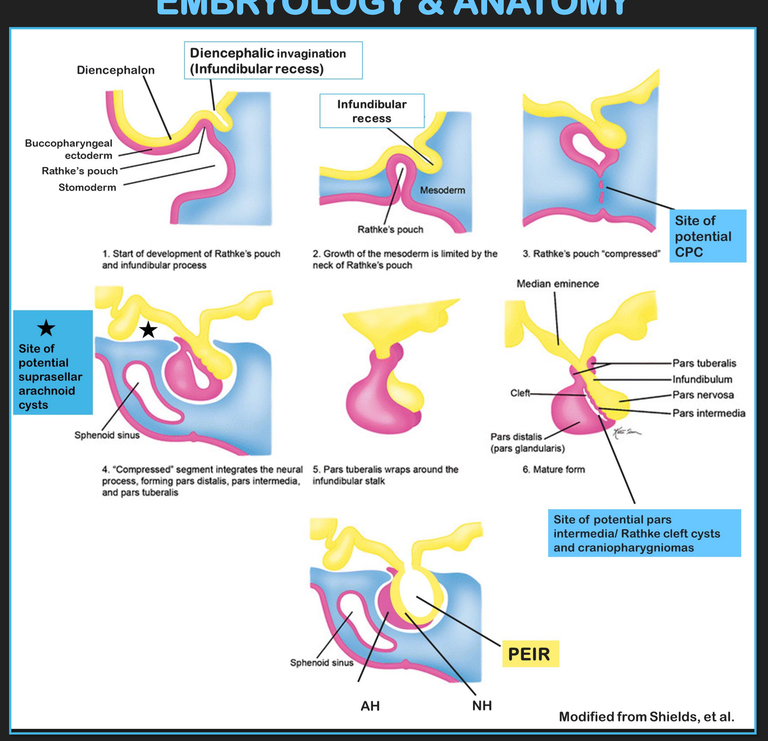
OBJECTIVES & LEARNING POINTS

1. Describe the entity of PEIR and review the appearance on MR imaging.
2. Describe the embryology and key anatomy to understand the basis of PEIR and anatomic relationship of PEIR with surrounding structures.
3. Distinguish and contrast PIER from other congenital anomalies and pathology of the sellar region using a cased based approach.
4. Reference syndromes associated with PEIR.

INTRODUCTION

- The **persisting embryonal infundibular recess (PEIR)** is a rare anomaly of the neurohyphophysis resulting in a tubular shaped extension of the infundibular recess of third ventricle into the pituitary stem and sella.
- This is hypothesized to be a result of dysembryogenesis from failure of the embryonal infundibular recess (diencephalic invagination)to undergo obliteration by cellular proliferation. Over the last 45 years only approximately 10 cases have been described in the literature with the term PEIR recently coined by Steno, et al. To our knowledge only 2 pediatric cases have been reported in the literature.
- We present the largest cases series of PEIR to date, two which were associated with a morning glory disc anomaly and moya-moya disease, illustrating the embryology, anatomy, and imaging findings of 3 of the 5 pediatric cases of PEIR as well as referencing associated syndromes (the second case associated with morning glory syndrome and an incidental case will not be shown).
- We speculate that PEIR is likely underrecognized and in this presentation, we describe the characteristic imaging findings of PEIR—highlighting some of the key differences imaging findings and embryologic basis for other common cystic lesions of the pituitary.

EMBRYOLOGY & ANATOMY

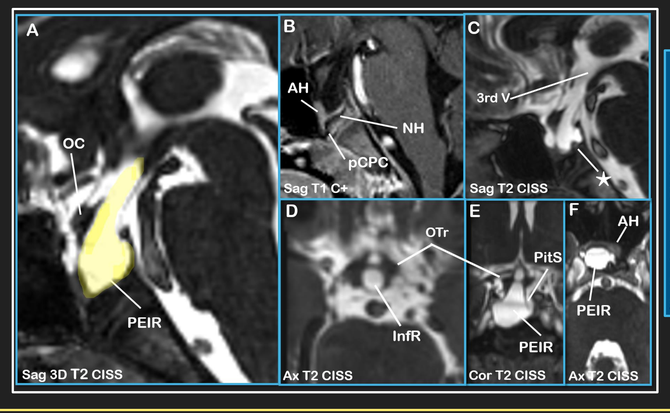


ABBREVIATION KEY:

3rd V = 3rd Ventricle	**MGDA** = morning glory disc anomaly
AH = Adenohypophysis	**NH** = Neurohypophysis
CPC = craniopharyngeal canal	**PEIR** = Persisting embryonal infundibular recess.
InR = Infundibular recess of the 3rd ventricle	**PitS** = Pituitary Stalk (infundibulum)
OC = Optic chiasm	**OTr** = Optic Tracts

CASE 1
Delayed Puberty & Low Growth Hormone

14-year-old female with delayed puberty found to have growth hormone deficiency and hypogonadotropic hypogonadism.

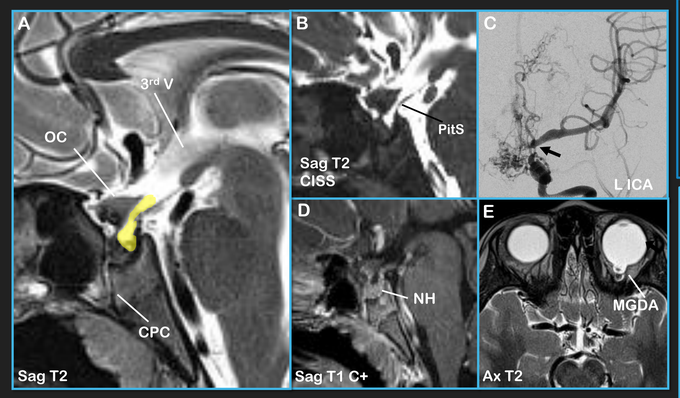


Clinical Pearl:
Concurrent pituitary anomalies and pathology may exist
Although the neurohypophysis and hypophysis have different embryologic derivatives, the inductive tissue interactions between the developing neurohypophysis and hypophysis set off a complex cascade of neuroectodermal signaling on the primordial pituitary gland.

Figure 1: (A,C,E,F) 3D high resolution T2WI of a patient with **PEIR** demonstrates a tubular CSF filled extension of the third ventricle into the pituitary stalk (infundibulum) and sella (highlighted in yellow in A). (B) Post contrast T1WI demonstrate a thinned neurohypophysis displaced by the PEIR. (A,B). The patient was also noted to have a small persistent craniopharyngeal canal with a small amount of the adenohypophysis noted to be herniated through the persistent craniopharyngeal canal (C) The sella is expanded, thinned and partially eroded (white star), and likely related to CSF pulsations. (D) The infundibular recess of the 3rd ventricle is more dilated than typical in a patient without hydrocephalus.

CASE 2
Morning Glory Syndrome

- 7-year-old female with chronic intermittent headaches with history of congenital nystagmus, coloboma, and equatorial staphyloma, and left eye esotropia. Post imaging ophthalmologic examination demonstrated a left globe morning glory disc anomaly (MGDA)



Findings:
- PEIR
- CPC
- MGDA
- Moya Moya
- Persistent trigeminal artery (not shown)
- Optic chiasm thickening

Clinical Pearl: The diagnosis of Morning Glory Syndrome was suggested by the radiologist, prompting re-assessment by neuro-ophthalmology, confirming the left MGDA. Although differentiating a coloboma from a MGDA can be challenging both by exam and imaging, recognizing PEIR as part of syndrome may aid in proper diagnosis.

Figure 2: (A,B) Sagittal T2WI and high resolution T2 demonstrate a PEIR with CSF filled pituitary stalk forming a channel between the third ventricle and the sella (highlighted in yellow in A). There is thickening of the optic chiasm. (A,B,C) A small craniopharyngeal canal is noted, which was confirmed on CT (not shown). (C) Diagnostic cerebral angiogram shows stenosis of the left ICA terminus with extensive basal and lentriculostriate collateral arterial vessels compatible with moya-moya disease (black arrowhead). The right ICA angiogram (not shown) demonstrated a tortuous dilated right intracranial internal cerebral artery with narrowing of the ICA terminus as well as a large persistent right trigeminal artery. (D) Post contrast sagittal T1WI demonstrates a CSF cleft within the posterior pituitary and heterogonous enhancement of the gland due to numerous collaterals within the sella and suprasellar cistern. On the non-contrast T1WI, the posterior pituitary T1 bright spot was preserved (not shown). (E) Axial T2WI demonstrated a left morning glory disc anomaly with extensive basilar arterial collaterals. A coloboma was present on the right (not shown).

CASE 3
Hydrocephalus & Aqueductal Stenosis

Previously healthy 17-year-old female who presented with headaches, ataxia and facial droop found to have chronic hydrocephalus and aqueductal stenosis.

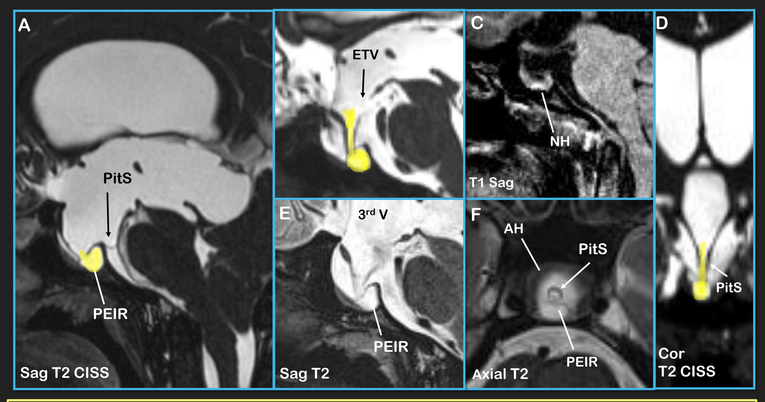


Figure 3: (A) MRI prior to endoscopic third ventriculostomy (ETV). Sag T2 CISS demonstrates aqueduct stenosis, severe ventriculomegaly with downward displacement of the floor of the third ventricle, and pituitary stalk. There is dilation of the infundibular recess, extending as a cystic dilatation into the sella, compatible with a **PEIR.** There is also tonsillar herniation/ Chiari 1 anomaly with horizontal orientation of a foreshortened clivus. Follow-up MRI was obtained 2-days later after ETV. (B,D,F) After decompression of the ventricles, high resolution T2WI demonstrates the persisting embryologic infundibular recess connecting the third ventricle and the sella more clearly. Note the expanded appearance of the sella with osseous remodeling on the sagittal images. (C) Sagittal T1WI shows the posterior pituitary T1 bright spot surrounding the recess of CSF within the sella, delineating a PEIR from pars intermedia cyst.

CONCLUSION

- **PEIR** is developmental anomaly, most likely resulting from failure to obliterate the primary embryonal diencephalic evagination.
- Key imaging features include:
  - Continuation of the infundibular recess of the third ventricle as a tubular shaped CSF cavity into the sella.
  - +/- T1 neurohypophysis bright spot may be present and sometimes can be delineated as tissue surrounding PEIR and is often anterior to the area of cyst-like dilatation. This is a key feature in delineating PEIR from cystic anomalies of the anterior pituitary lobe.
  - +/- Sellar enlargement with osseous remodeling is often present (likely due to CSF pulsation)
  - +/- Optic chiasm thickening may occasionally be present (as in cases with a MGDA)
  - +/- May co-exist with abnormalities of the adenohypophysis, such as a persistent craniopharyngeal canal.
- When hydrocephalus is present it is likely incidental and merely uncovers the existing anomaly.
- PEIR may be isolated as an incidental finding but can be associated with syndromes involving midline structures such as morning glory syndrome and moya-moya disease.
- +/- Normal pituitary function. We hypothesize that PEIR is a marker of pituitary dysfunction rather than a cause. Recommendation for endocrine evaluation should be considered.

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