

Pituitary Dysfunction in Children with Ectopic Posterior Pituitary

Ghada Najji^{1,4}, Erica Poletto^{2,4}, Kristin Brown³, Rita Ann Kubicky^{1,4}

¹Section of Endocrinology and Diabetes, St. Christopher's Hospital for Children;
²Section of Radiology; ³Motion Analysis Lab, St. Christopher's Hospital for Children;
⁴Department of Pediatrics, Drexel University College of Medicine, Philadelphia, PA.

BACKGROUND

Imaging studies aid in the detection of structural abnormalities that may be associated with pituitary dysfunction, such as ectopic posterior pituitary (EPP). The location of the ectopic lobe can vary, but it is most commonly located along the median eminence in the floor of the third ventricle. An EPP could result from complete or partial defective neural migration during embryogenesis, which could explain the different loci of EPP. Affected individuals have either severe isolated growth hormone deficiency (IGHD) or combined pituitary hormone deficiencies (CPHD); diabetes insipidus is not a feature, indicating that the EPP is functioning normally.

AIM

To detect the prevalence of IGHD or CPHD in children with EPP. In addition, to evaluate the association between the location of EPP and pituitary dysfunction.

METHODS

A retrospective chart review of MRI reports at St. Christopher's Hospital for Children (SCHC) from 2006-2018 that were found to have EPP. Pituitary hormone function was evaluated in the majority of the patient population.

RESULTS

Of the 26 patients with EPP, [16 males and 10 females], mean chronologic age was 5.98±5.18 yrs, only 20 patients had an endocrine evaluation at SCHC. Of the 20 children, 14 had CPHD and 4 had IGHD. Patients were classified into 3 groups (upper, middle & lower) according to EPP location along the pituitary stalk. Of the 21 patients with upper EPP, 17 had pituitary dysfunction (14 with CPHD, 3 with IGHD). Of the 4 children with middle EPP, only 1 had pituitary dysfunction which was IGHD. One patient had lower EPP with hyperprolactinemia. Diabetes insipidus was not identified in any of the children.

Table 1: Demographics Data

	Age mean (std)	Height SDS mean (std)	Gender –male n(%)
Patient Population n=26	5.98 (5.18)	-3.06 (4.77)	16 (61.5)

Table 2: Demographic Data Based on EPP Location

Group 1 - Upper: at or above optic chiasm
Group 2 - Mid: below chiasm, above the insertion of stalk on adenohypophysis
Group 3 - Lower : At insertion of stalk on adenohypophysis

	Group 1 - Upper (n=21)	Group 2 - Mid (n=4)	Group 3 - Lower (n=1)	Total (n=26)
Age (yrs) mean (std)	5.25 (4.74)	7.25 (5.62)	16.4 (0)	5.98 (5.18)
Gender (male) n (%)	13 (62)	3 (75)	0 (0)	16(62)
Height SDS mean (std)	-3.37 (5.09)	-1.87 (0.52)	0.183(0)	-3.06 (4.77)

Figure 1: Images

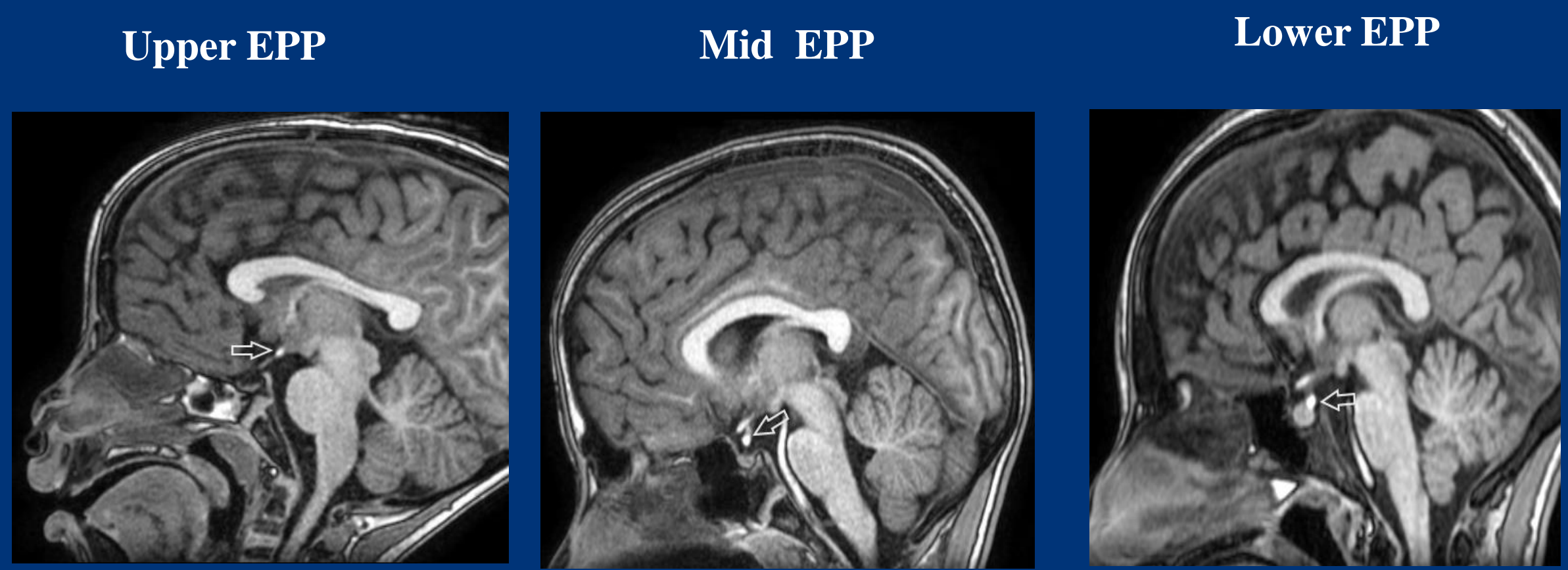


Table 3: Frequency of Diagnostic Categories for Groups 1, 2, 3

Diagnosis	Group 1 - Upper (n=21) n (%)	Group 2 - Mid (n=4) n (%)	Group 3 - Lower (n=1) n (%)	Total (n=26) n (%)
Combined Pituitary Hormone Deficiencies (CPHD)	14 (67)	0 (0)	0 (0)	14(54)
Isolated GHD (IGHD)	3 (14)	1 (25)	0 (0)	4 (15)
DI	0(0)	0(0)	0(0)	0(0)
Hyperprolactinemia (Microcephaly)	0 (0)	0 (0)	1(100)	1(4)
No Endocrine Evaluation	4 (19)	2 (50)	0 (0)	6(23)

Table 4: Mean Age and Height SDS for Patients with Isolated GHD in Mid and Upper Group

	Upper n= 3 mean (std)	Mid n=1 Mean (std)
Age (yrs)	4.5 (3.122)	13 (0)
Height (SDS)	-3.678 (2.225)	-2.245 (0)

CONCLUSIONS

Our study supports previous reports that CPHD and IGHD are frequent in patients with EPP. Similarly, our data further demonstrate that no cases of diabetes insipidus have been reported in children with EPP. This study also elucidates that patients with IGHD with upper EPP were shorter and younger than those with middle EPP. In our study, EPP is most commonly located along the upper third of the pituitary stalk at the median eminence level, with a higher prevalence of CPHD and IGHD, a finding similar to prior studies. No CPHD was reported in middle/lower but IGHD was found in the middle EPP group.

REFERENCES

- Mitchell LA, Thomas PQ, Zacharin MR, Scheffer IE. Ectopic posterior pituitary lobe and periventricular heterotopia: Cerebral malformations with the same underlying mechanism? AJNR Am J Neuroradiol 2002;23(9):1475-1481.
- Saleem SN, Said AH, Lee DH. Lesions of the hypothalamus: MR imaging diagnostic features. Radiographics 2007;27(4):1087-1108.
- Van der Linden AS, Van Es HW. Case 112: Pituitary stalk transection syndrome with ectopic posterior pituitary gland. Radiology 2007;243(2):594-597.