Primary Empty Sella Syndrome and Clinical Endocrine Polymorphisms in children.

P2-P322

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Background: Primary empty sella syndrome (PES) is rare in children. Some reports relating its association with various endocrine (GHD, precocious puberty, IHH, etc.) and non-endocrine manifestations (ophthalmologic symptoms, benign cranial hypertension, etc.) in children have been published. Asymptomatic patients have also been reported. Its precise incidence remains unknown, although estimates of 1.2% (children without symptoms) and 68% (in children with associated endocrinopathies) have been reported (1, 2).
PES results from the herniation of subarachnoid space into sella turcica, pushing the pituitary gland to either the bottom or to one side of the sella.

Objective: To analyze causal relationships between primary ES syndrome and endocrine manifestations in 15 pediatric cases seen in our clinical settings, and suggest a follow-up of both symptomatic and asymptomatic patients.

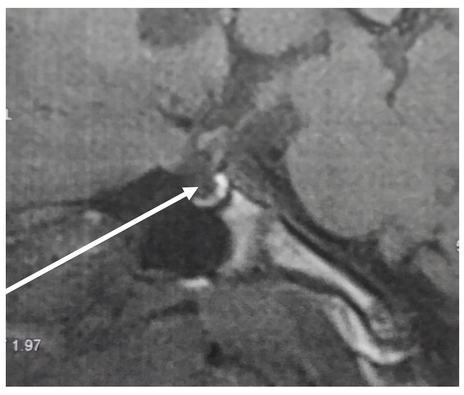
Population & Methods: Retrospective review of 15 children diagnosed with ES in our pediatric endocrinology clinics. Anthropometric measures, mode of presentation, clinical manifestations and associated endocrine abnormalities were extracted from each medical record. Results of clinical examinations, endocrine work-up and cranial MRI were analyzed.

Results: We gathered 15 cases of PES syndrome, 11 were boys and 4 girls, with mean age of 11 years (range 3yrs$^{5/12}$ to 17yrs$^{8/12}$), 4 patients were obese (2 first degree, 2 second degree obesity). Cranial MRI was performed to investigate endocrine abnormalities in 10 cases, 1 patient had Noonan syndrome. MRI was performed for other reasons in 3 cases (1 chronic migraine, 1 macrocephaly, and 1 suspected benign intracranial hypertension), in 2 other cases ES diagnosis was incidental. 11 patients had endocrine manifestations: 7 GHD, 2 HH, 3 pubertal delay (2 associated with GHD, 1 isolated), 1 boy had CPP.
PES syndrome was total in 9 cases and partial in 6 (Pituitary height 3 mm: 5 cases; 2 – 2.5 mm: 7 cases; < 2mm: 1 case) (Table 1). There seemed to be no correlation between pituitary height and the type of endocrine manifestations. On follow-up over a 10 years period (10 cases) the arachnoidocele persisted in all cases without additional symptoms or endocrine abnormalities. GHD occurred secondarily at age 11 years in 1 patient (PES diagnosed at 3 years 3/12), endocrine testing remains unremarkable in another case diagnosed at 6 years 5/12.

Table I: Patients Characteristics, Diagnostic Circumstances, Pituitary Height on MRI.

S. No.	Gender	Age (years)	Pituitary height (MRI) in mm	Endocrine abnormalities
1	M	10.4	2.3	CPP
2	M	17.8	< 2.0	HH
3	M	16.10	2.0	HH
4	M	15.9	2.0	Partial GHD
5	M	16.2	2.0	GHD
6	M	15.4	3.0	CDP
7	M	3.5	< 2.0	GHD
8	M	7.4	2.0	GHD
9	M	4.4	3.0	Partial GHD
10	M	10.1	2.0	No endocrine abnormality
11	M	9.3	3.0	Partial GHD
12	F	11.0	3.0	Partial GHD
13	F	12.1	2.0	No endocrine abnormality
14	F	10.4	2.0	No endocrine abnormality
15	F	6.5	2.0	No endocrine abnormality

CPP: Central Precocious Puberty; **GHD**: Growth Hormone Deficiency; **HH**: Hypogonadotrop Hypogonadism; **CDP**: Constitutional Delay of Puberty.



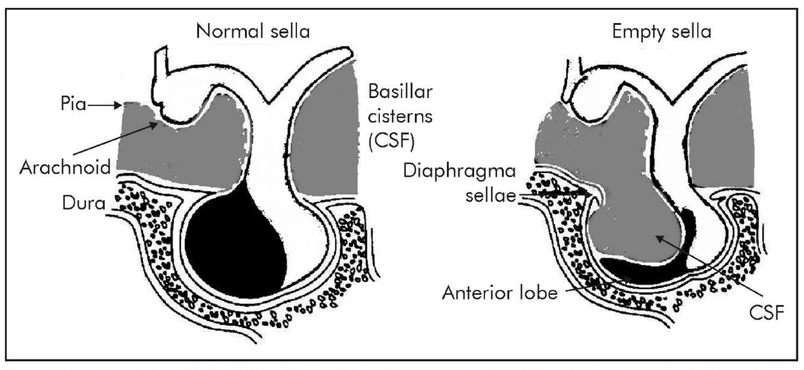
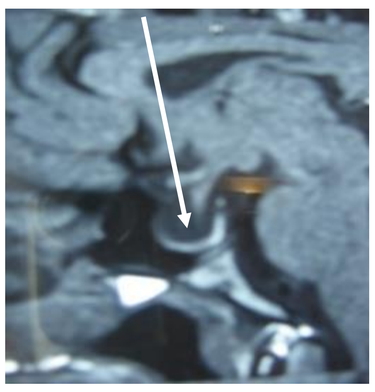


Fig. 1 : Showing normal anatomic relationships of sella (left) and arachnoid herniation through an incompetent diaphragma sellae, with ballooning of the sella (right). J Indian Academy of Clinical Medicine 2001; 2 (3):198-202



COMMENTS:

- The etiologies of PES syndrome remains speculative, and its mechanisms unclear,
- The physiopathological mechanisms explaining the association between PES and various pituitary pathologies are still unknown, as PES is also found in asymptomatic patients. Its congenital origin is likely in some cases.
- GHD is the commonest endocrine manifestation in patients with PES, it may occur over time (1- 4).
- As reported earlier by others (1, 5, 6), there was a male gender predominance in our case series in opposition to female predominance in adult cases (7).
- There seem to exist no correlation between pituitary gland height on MRI on one side, and the associated endocrinopathies on the other,
- Variability in clinical manifestations associated with PES raises the question as to whether PES is the cause of endocrine dysfunctions or just an incidental finding.
- We suggest annual follow-up (growth, ophthalmological, neurological and pubertal screenings) with baseline endocrine testing in asymptomatic children at diagnosis (1-4, 6-8). Control MRI whether indicated or not remains questionable.

CONCLUSION: PES is rare in children, but endocrine surveillance of diagnosed children is mandatory. Long-term follow-up is indicated as pituitary hormone deficiencies may occur later in childhood.

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