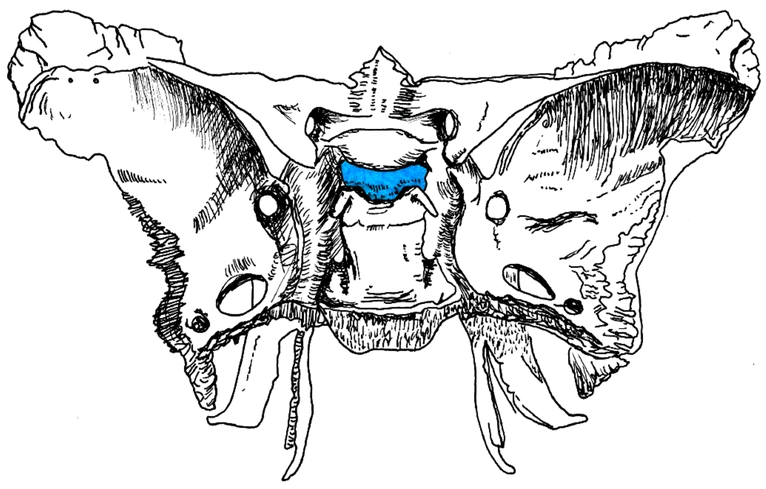

Endocrine and Radiological Abnormalities in Empty Sella Syndrome

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Introduction

Empty Sella Syndrome (ESS) is diagnosed when on imaging of the brain, the pituitary fossa appears empty. It is classified as primary or secondary depending on the aetiology.

Primary ESS is the herniation of the meninges through a congenitally incompetent diaphragma sellae into the **sella turcica**. Hydrostatic CSF pressure may enlarge the herniation and compress the gland against the wall or floor of the fossa. This gives the radiological appearance of an empty sella although the term is a misnomer as it is not empty but rather filled with CSF.

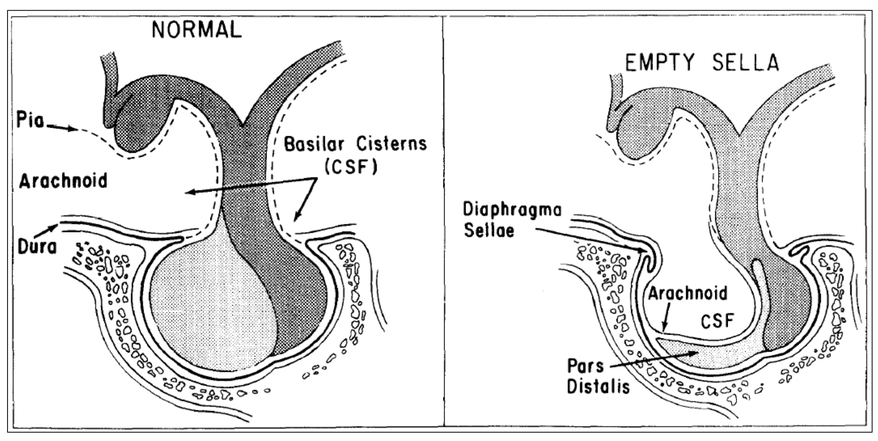


Figure 1: The Anatomy of Primary ESS. The arachnoid mater herniates through an incompetent diaphragma sellae. Taken from Jordan, Kendall and Kerber (1977).

Secondary ESS is caused by damage to pituitary tissue which results in an empty sella turcica.

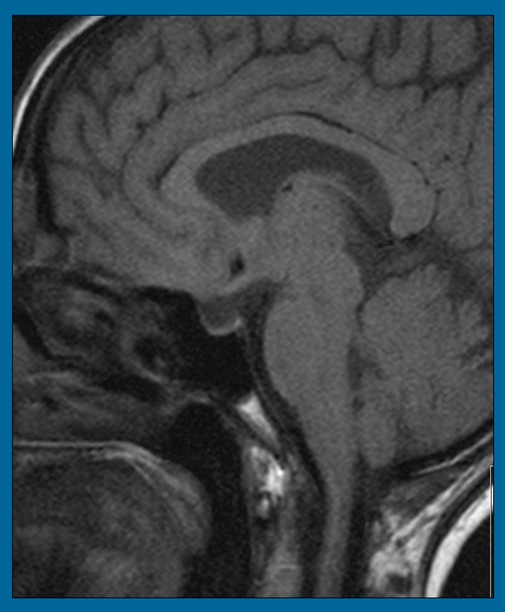


Figure 2: The typical appearance of primary ESS on Magnetic Resonance Imaging.

The posterior pituitary bright spot is visible and there is a thin layer of pituitary tissue lining the sella floor. The stalk is normally positioned and there are no gross brain abnormalities.

Context

There is significant lack of agreement in the literature regarding the number of patients with empty sella syndrome (ESS) who suffer from pituitary dysfunction; some papers report that pituitary function is usually preserved while others state that it is not.

Aims

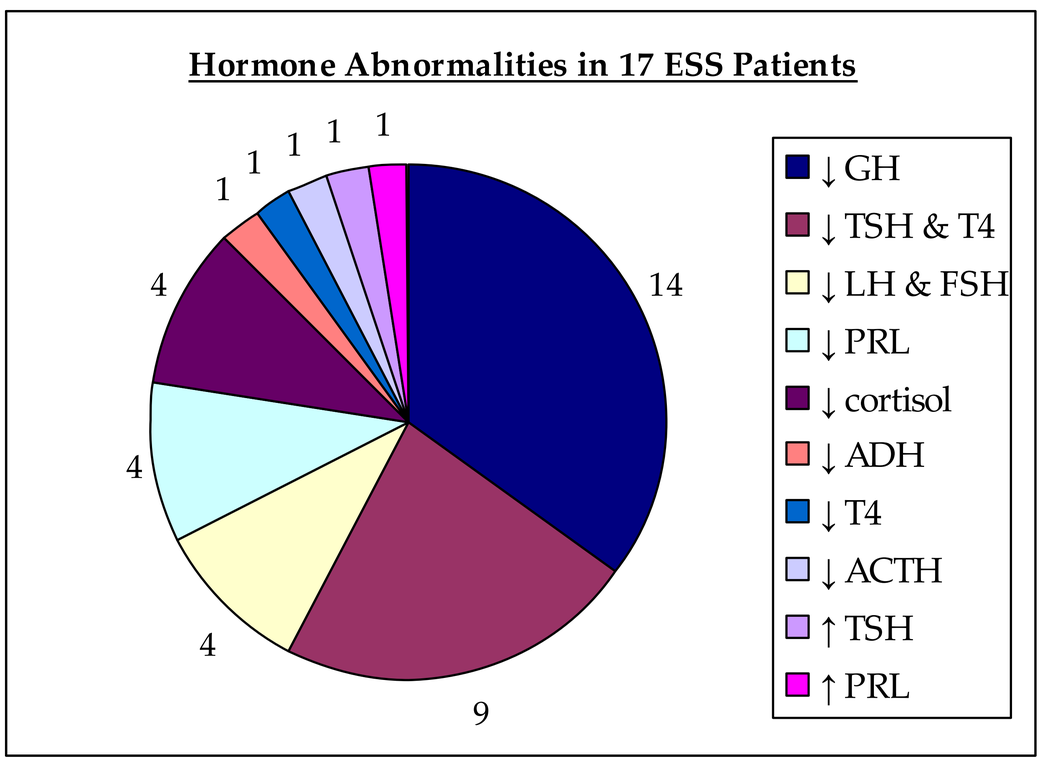
The aim of this study was to determine the proportion of patients at St. George's Hospital with ESS who had endocrine abnormalities on presentation and to define radiological features to differentiate between primary and secondary ESS.

Materials and methods

Records of all patients diagnosed with an 'empty sella turcica' at St. George's Hospital since 1990 were retrospectively reviewed. 26 patients satisfied these criteria and their MRI scans were re-evaluated by a 'blinded' neuroradiologist to confirm a diagnosis of primary or secondary ESS. Other patient information such as demographics, presenting complaint and, most importantly, results of baseline endocrine testing were also collected.

Results

- Type of ESS diagnosed from MRI: partial: 4, primary: 18, secondary: 4.
- Gender ratio: 2.7 F : 1.0 M.
- **65% (n=17) of patients diagnosed with ESS showed evidence of pituitary dysfunction.**
- The most common endocrine abnormality was **GH deficiency** which affected 54% (n=14), followed by **secondary hypothyroidism** in 9 patients with low levels of TSH and T_4.



- Headache (39%) and fatigue (27%) were the most common presenting complaints.
- Three patients were asymptomatic.

Discussion

The existing literature on this subject comprises ten studies reviewing a total of 562 ESS patients between 1972 and 2006. They stated that an average of 42.8% (range 18.8 - 66.7%) of ESS patients had a degree of hormone dysfunction.

GH secretion is thought to be most commonly affected hormone due to the lateral location of the somatotroph cells, which comprise almost 50% of the anterior pituitary.

Females are more commonly affected due to the affect of pregnancy on the pituitary gland. It enlarges and then regresses postpartum which could cause herniation of the subarachnoid space.

Conclusion

ESS is a heterogeneous condition with varied, nonspecific symptoms and high rate of endocrine dysfunction. Endocrine function should be assessed in all patients with this diagnosis. To our knowledge this was the only study ever performed in the UK which evaluated the impact of ESS on pituitary function.

References

Jordan, R. M., Kendall, J. W., Kerber, C. W. (1977) The primary empty sella syndrome: analysis of the clinical characteristics, radiographic features, pituitary function and cerebrospinal fluid adenohypophysial hormone concentrations. *Am J Med.* 62 (4), pp. 569 – 580

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