KARNATAKA RADIOLOGY EDUCATION PROGRAM

# CASE PRESENTATION - CASE 2

**CASE OF POSTERIOR PITUITARY STALK INTERRUPTION SYNDROME**

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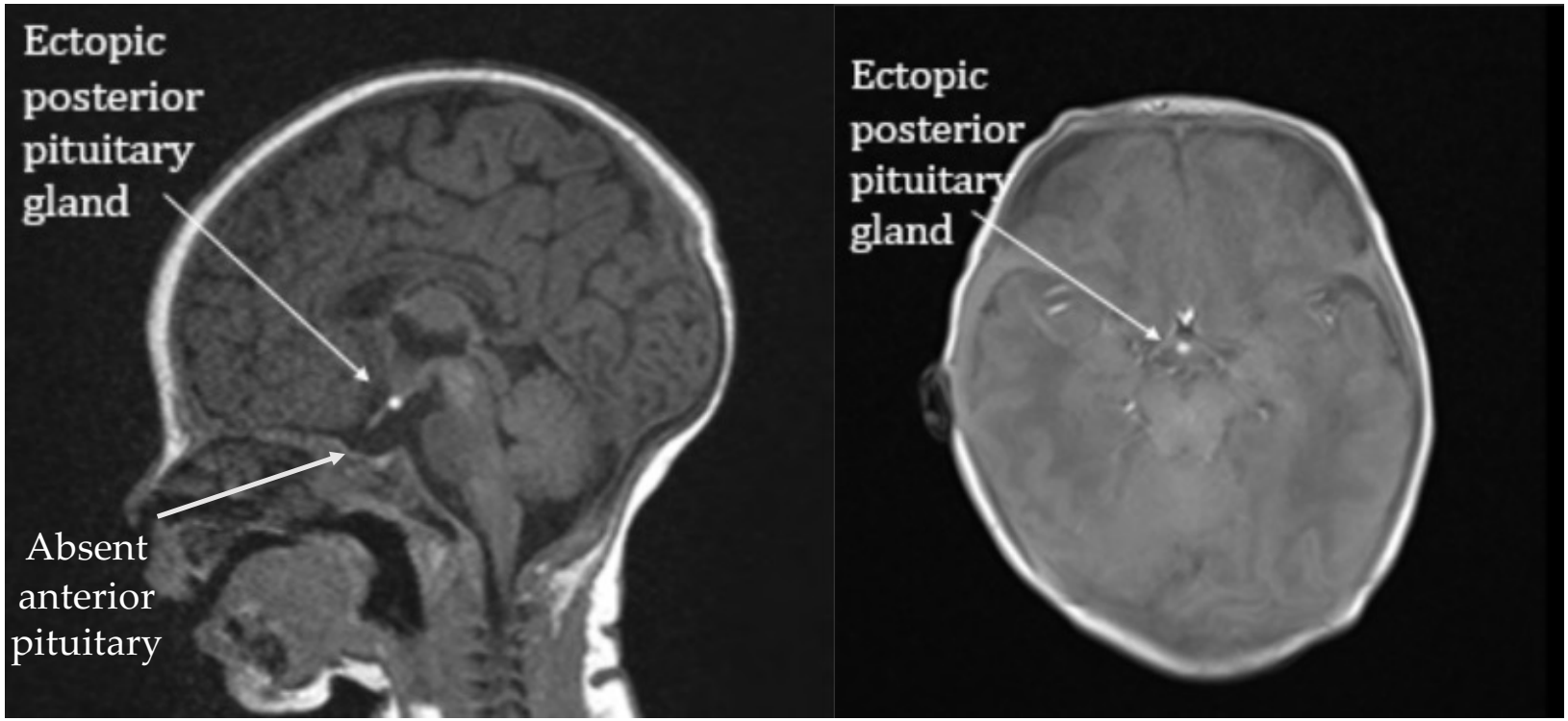
# CASE 2

- A 17-day old male neonate presented with persistent hypoglycemia and neonatal jaundice.
- The neonate was a full term with breech presentation and LSCS delivery, he cried after 1 cycle of bag and mask ventilation.
- Initial hormonal panel showed low levels of serum cortisol (0.82), TSH (0.51) and free T4 (0.7).

# CASE 2

MRI Brain plain:




MRI of the brain showed an absent anterior pituitary gland and absent pituitary stalk with an ectopic posterior pituitary gland, consistent with Pituitary Stalk Interruption Syndrome (PSIS).

# Differentials

1. Empty sella
2. Septo-optic dysplasia
3. Kallmann syndrome

# MRI Findings in Posterior pituitary stalk interruption syndrome :

- Absence or Hypoplasia of the Pituitary Stalk:

  Seen as a thin, interrupted, or completely absent stalk, with no visible connection between the hypothalamus and the pituitary gland.
- Abnormal Pituitary Gland:

  Hypoplastic/ small anterior pituitary lobe

  May be absent or displaced into an ectopic location.

  The posterior pituitary lobe might be located more caudally than usual, with an abnormal configuration.
- Upward displacement of the Hypothalamus
- Enlarged third ventricle due to the displacement of brain structures.
- Arachnoid cysts can occasionally be seen near the pituitary stalk

# Clinical symptoms:

- PSIS typically manifests in early childhood with endocrine, developmental, and neurological symptoms.
- Endocrine issues include growth hormone deficiency causing short stature, multiple pituitary hormone deficiencies that may lead to panhypopituitarism, hypothyroidism, hyperprolactinemia, and adrenal insufficiency.
- Developmental challenges include delayed puberty, learning difficulties, seizures, and hypoglycemia.
- Birth complications such as breech presentation, neonatal hypoglycemia, and jaundice are common.

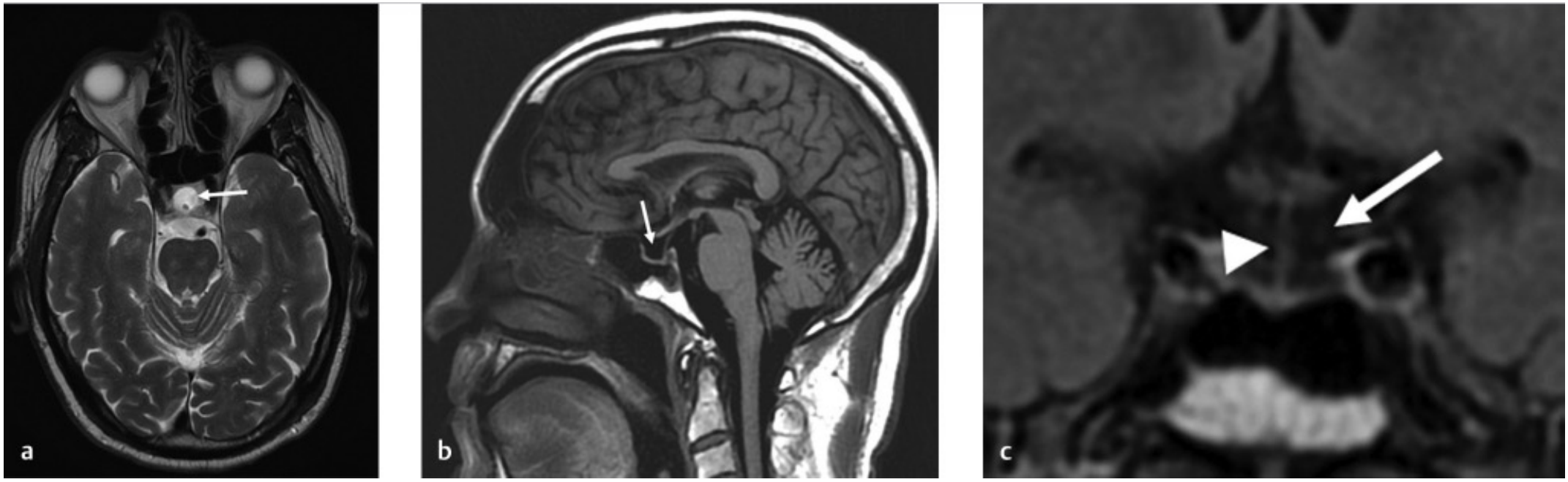
# Treatment:

- After the complete work up the patient was started on hydrocortisone and resulted in reversal of the hypoglycemia and neonatal jaundice.

# Empty sella

| POINTS IN FAVOUR | POINTS AGAINST |
|---|---|
| Empty sella | Ectopic posterior pituitary |
| | Thin or absent pituitary stalk |
| | Clinical features - panhypopituitarism |

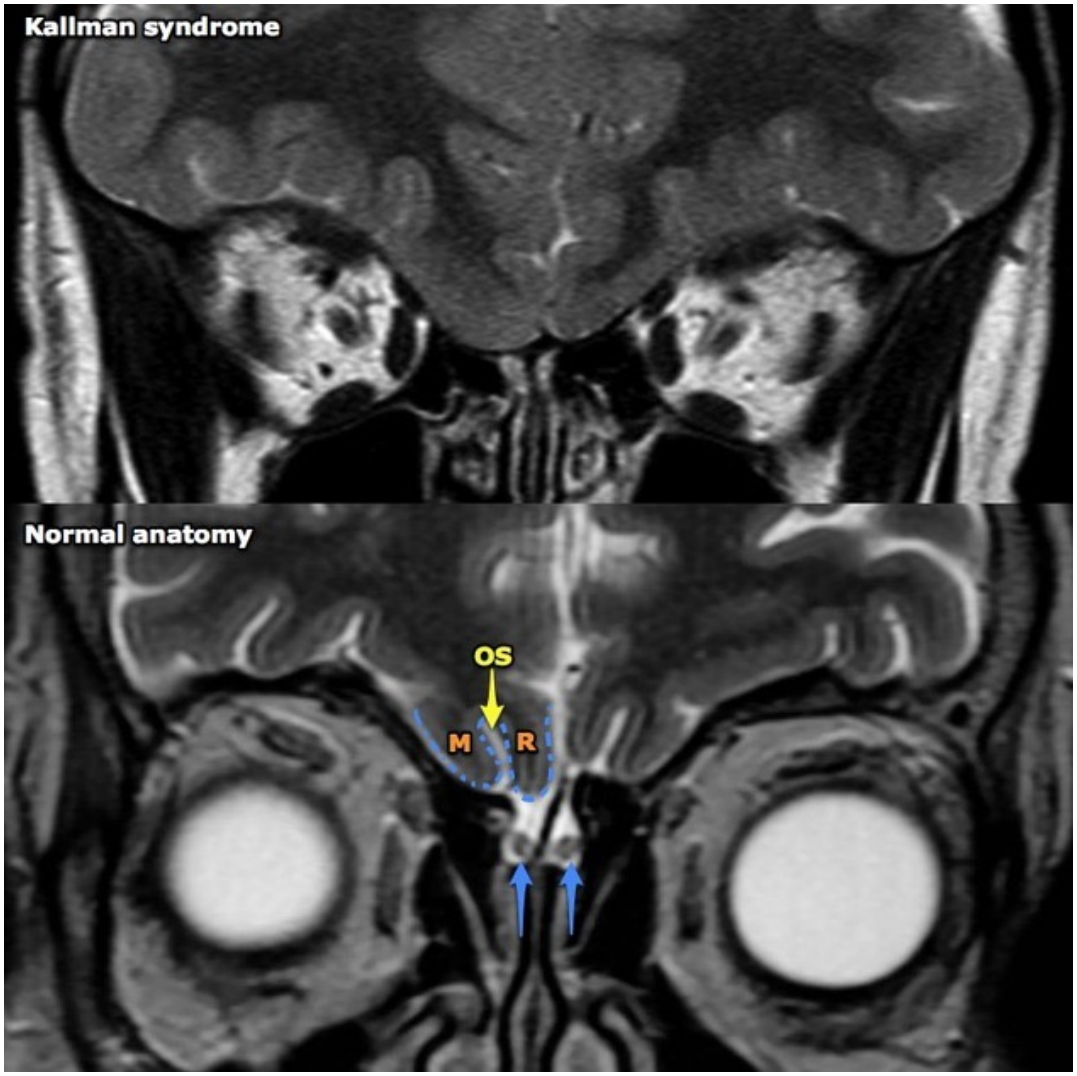
# Empty sella



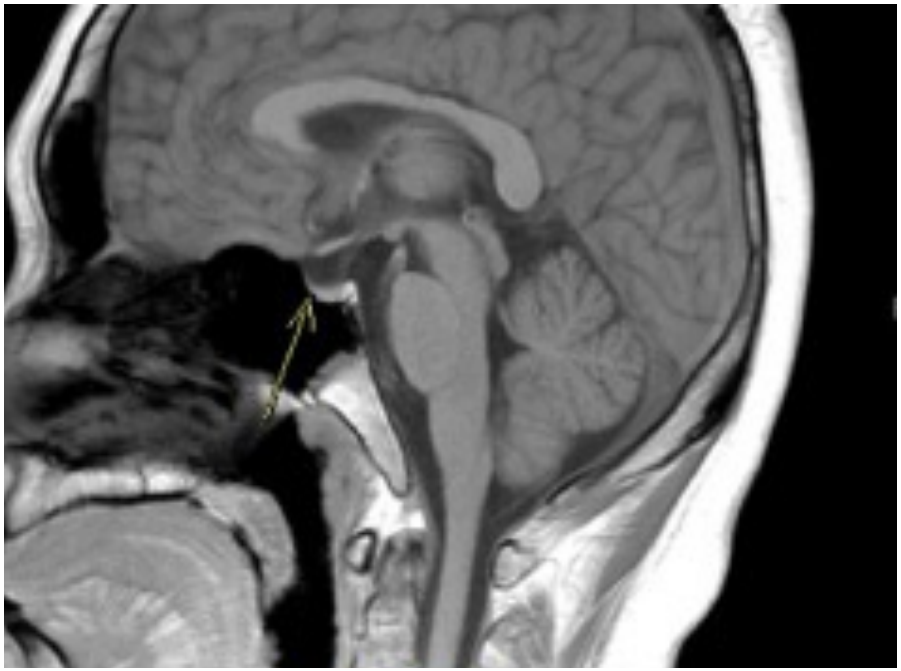
# Kallmann syndrome

| POINTS IN FAVOUR | POINTS AGAINST |
|---|---|
| Hypoplastic pituitary stalk | Small anterior lobe of the pituitary gland |
| | Hypoplastic or absent olfactory bulbs |
| | Genetic association - like PROKR2, FGFR1, and HESX1 |
| | Isolated gonadotropin deficiency – in case of Kallman syndrome |
| | Clinical features : Anosmia or hyposmia |

*Absent olfactory bulbs*



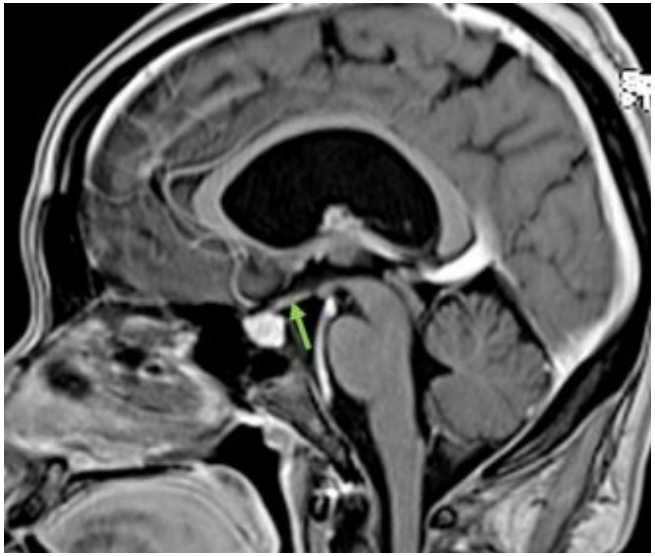

*Pituitary gland hypoplasia*



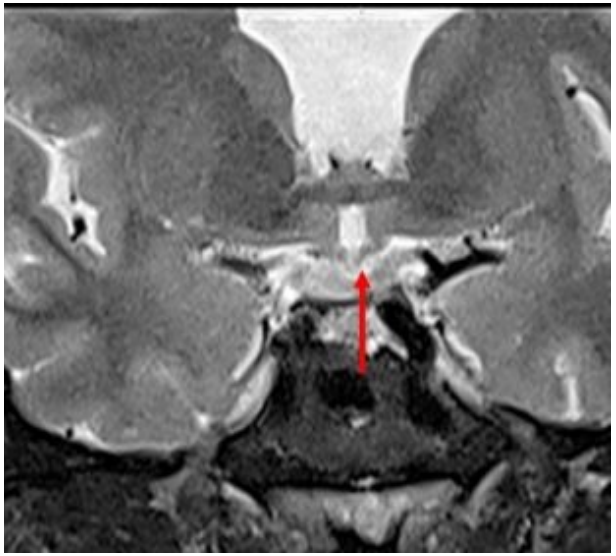
Hilman, S., Dewi, D. K., & Kartika, E. (2023). A rare disease of Kallmann syndrome: A case report. *Radiology Case Reports*, *18*(3), 1232–1238. https://doi.org/10.1016/j.radcr.2022.12.036

# Septo-optic dysplasia

| POINTS IN FAVOUR | POINTS AGAINST |
| --- | --- |
| Small pituitary gland with hypoplastic or absent infundibulum | Hypoplastic optic chiasm/optic nerves and globes,<br>Absent septum pellucidum,<br>Schizencephaly |
| Clinical features:<br>• Endocrine anomalies - hypothalamic-pituitary dysfunction | Clinical features:<br>• Visual impairment (due to optic nerve hypoplasia)<br>• Seizures or developmental delay |

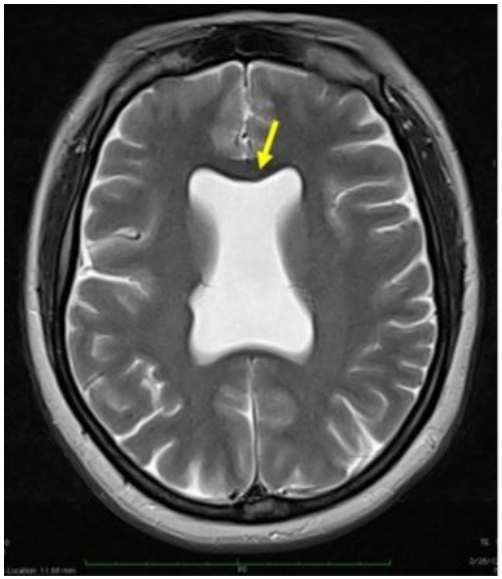


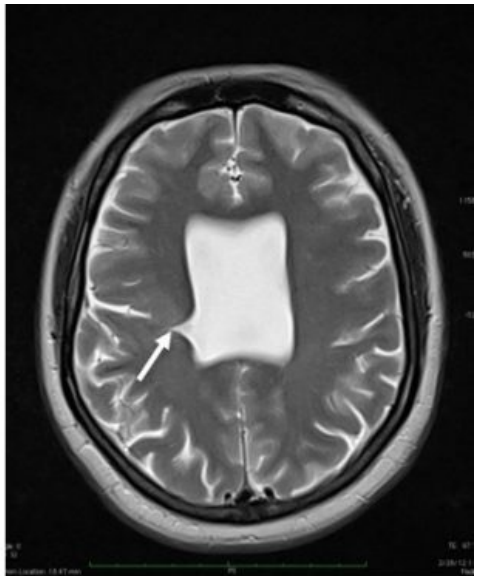
Normal pituitary gland, however ***pituitary stalk*** appears small (green arrow).



Small ***optic nerves*** and optic chiasm (red arrow).

***Absent septum pellucidum*** with "box-like" configuration of the frontal horns of the lateral ventricles (yellow arrows).





***Schizencephaly*** - Incomplete cleft (lined by grey matter) from the lateral wall of the body of the right lateral ventricle which traverses across almost to join the sulci (white arrows).

THANK YOU.