**Introduction**

- Hypopituitarism leads to one or more pituitary hormones deficiency.
- Hypopituitarism can be congenital or acquired.
- The incidence of congenital hypopituitarism is between 1 in 4000 and 1 in 10,000 live births.
- Adamantinomatous type craniopharyngioma is a relatively rare childhood tumor with the prevalence of 6%–10% of all intracranial tumours.
- Congenital craniopharyngioma is a very rare suprasella brain tumour which constitute around 0.5–1.5% of all congenital tumours.
- We report the first case of congenital craniopharyngioma in Sri Lanka presenting with hypopituitarism.

**Case report**

- Term baby presented with micro phallus with bilateral hypoplastic scrotum and atrophic testis.
- Managed as hypopituitarism and planned MRI brain at one year.
- Baby presented at the age of eleven months with bulging fontanel suggestive of increase intracranial hypertension.
- MRI brain at that time revealed solid and cystic mass involving the sella and suprasella region with obstructive hydrocephalus compatible with adamantinomatous type craniopharyngioma.

**Investigations**

<table>
<thead>
<tr>
<th></th>
<th>Initial Evaluation</th>
<th>Follow up</th>
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<tbody>
<tr>
<td>LH</td>
<td>&lt;0.07IU/L</td>
<td></td>
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<tr>
<td>FSH</td>
<td>0.07IU/L</td>
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<tr>
<td>Basal cortisol</td>
<td>&lt;22nmol/L</td>
<td></td>
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<tr>
<td>SST 0min</td>
<td>&lt;22</td>
<td></td>
</tr>
<tr>
<td></td>
<td>30min</td>
<td>78.7</td>
</tr>
<tr>
<td>Free T4 (9-25)</td>
<td>10.10pmol/L</td>
<td>7.2</td>
</tr>
<tr>
<td>TSH (1.82-8.21)</td>
<td>3.22mIU/L</td>
<td>1.98</td>
</tr>
</tbody>
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**Management**

- He underwent excision of the craniopharyngioma.
- Post op needed multiple pituitary hormones replacement (hydrocortisone, thyroxine and desmopressin).
- Post operatively he had developmental deterioration and visual impairment.
- Radiotherapy was not given due small age.

**Conclusion**

- This case highlights the very rare congenital cause of hypopituitarism.
- Antenatal and early postnatal imaging with the ultra sound brain would help in the early diagnosis.
- Total resection is the treatment modality. Even with improving neurosurgical technology, prognosis is poor due to the age, size and location of the tumour.