CRANIOPHARYNGIOMAS – 35 YEARS OF EXPERIENCE IN A CENTRAL HOSPITAL’S ENDOCRINOLOGY DEPARTMENT
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INTRODUCTION
- Craniopharyngiomas: rare epithelial tumors of the sellar and parasellar region, with high survival rates (~90%) but with frequent tumor recurrence or persistence and significant morbidity.
- Annual incidence: 0.5-2.0 cases/million/year.
- Bimodal age distribution: peak incidence rates at 5–14 years and at 50–74 years.
- Clinical manifestations related to mass effect and hypothalamic/pituitary deficiencies.

METHODS
- Information collection from clinical records
- Review of the epidemiology, diagnosis, treatment and follow-up of patients with diagnosis of craniopharyngioma followed in an Endocrinology Department between 1980 and 2015
- Statistical analysis using SPSS v. 22.0

RESULTS
- Sample size: 40 patients
- 50% male, 50% female
- Median age by diagnosis: 36 years (minimum 6, maximum 70)
- Clinical manifestations at diagnosis:
  - visual alterations 77.5% (n=31)
  - headache 72.5% (n=29)
- 77.5% with sellar + suprasellar involvement
- Cystic component detected in 89.7% and calcifications in 47.5%
- Initial surgical approach: transcranial in 74.4%, transsphenoidal in 25.6%
- Neuropathology results (n=29):
  - adamantinomatous 79.3% (n=23)
  - papillary 20.7% (n=6)
- Average number of surgical interventions per patient: 2 (minimum 1, maximum 7)
- Eight patients (20.0%) treated with radiotherapy
- 47.5% of patients with residual tumor in the last neuroimaging evaluation
- Panhypopituitarism detected in 6/30 patients preoperatively and in 23/39 in follow-up
- All isolated anterior pituitary deficits also more frequent in follow-up

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<thead>
<tr>
<th>At diagnosis</th>
<th>Last follow-up</th>
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<tbody>
<tr>
<td>Panhypopituitarism</td>
<td>20.0% (n=6/30)</td>
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<tr>
<td>GH deficit</td>
<td>67.7% (n=21/31)</td>
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<td>Secondary hypogonadism</td>
<td>81.3% (n=26/32)</td>
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<td>Secondary hypothyroidism</td>
<td>40.0% (n=12/30)</td>
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<td>Secondary adrenal insufficiency</td>
<td>38.7% (n=12/31)</td>
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- Weight gain seen in 83.9% of patients (average ↑ 20.5 Kg)
- Prevalence of diabetes mellitus, arterial hypertension and dyslipidemia also increased

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<td>Overweight/obesity</td>
<td>56.2% (n=18/33)</td>
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<tr>
<td>Dyslipidemia</td>
<td>36.8% (n=14/38)</td>
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<tr>
<td>Arterial hypertension</td>
<td>13.2% (n=5/38)</td>
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<tr>
<td>Diabetes mellitus</td>
<td>5.3% (n=2/40)</td>
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<tr>
<td>Central diabetes insipidus</td>
<td>16.2% (n=6/37)</td>
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- Average follow-up time: 15.5 (± 10) years
- Six patients died (4 diagnosed between 6 and 14 years of age)
- Neurological sequelae: 32.4% (n=11/34)
- Visual deficits: improvement in 51.4% (n=19/37), unchanged in 45.9% (n=17/37)

CONCLUSION
- Despite the evolution in neurosurgical technique and the possibility of radiotherapy, craniopharyngiomas are still associated with high rates of tumor persistence and substantial morbidity.
- Hypothalamic alterations, pathological or treatment-induced, have a major impact on prognosis mainly due to hypothalamic obesity. Treatment decisions for primary and recurrent disease need to consider long-term tumor control as well as treatment-related morbidity.
- Craniopharyngioma often must be managed as a chronic disease by experienced multidisciplinary teams.
- Optimization of the treatment of any endocrine and metabolic sequelae is essential.