Endocrine-metabolic characterization of pediatric patients with craniopharyngioma. A single-centre cohort study.

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Introduction:

Craniopharyngioma is a rare, embryonic malformation of the sellar/parasellar region with low histological grade.

In childhood-onset craniopharyngioma (CoCR), endocrine dysfunctions, severe obesity and metabolic syndrome (MetS), neurological impairment and reduced quality of life have been described as consequences of both localization and treatment.

Objectives:

To characterize a population with CoCR and to correlate endocrine/metabolic sequelae with different surgery approach.

Methods:

We performed a retrospective longitudinal study on a single-centre cohort of 66 children (36 males) with CoCR, followed from 1990 to 2017. Clinical evaluations were performed yearly.

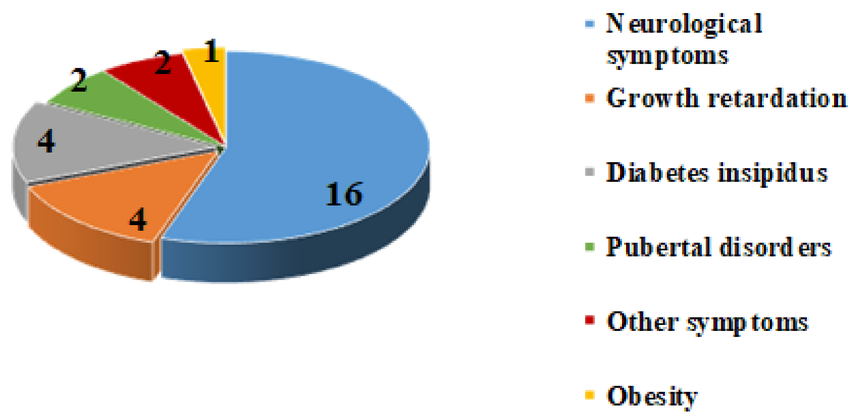
Results:

Age at diagnosis (7.24±4.92 yrs), established in 34/66 patients, was **not statistical different between sex** (males 6.19±3.86 vs females 8.18±5.64)

Clinical presentation was heterogeneous:

- neurological symptoms (55.2%)
- growth retardation (13.8%)
- diabetes insipidus (DI) (13.8%)
- pubertal disorders (6.9%)
- other symptoms (10.3%)
- in 1 case severe obesity was the unique presentation symptom.

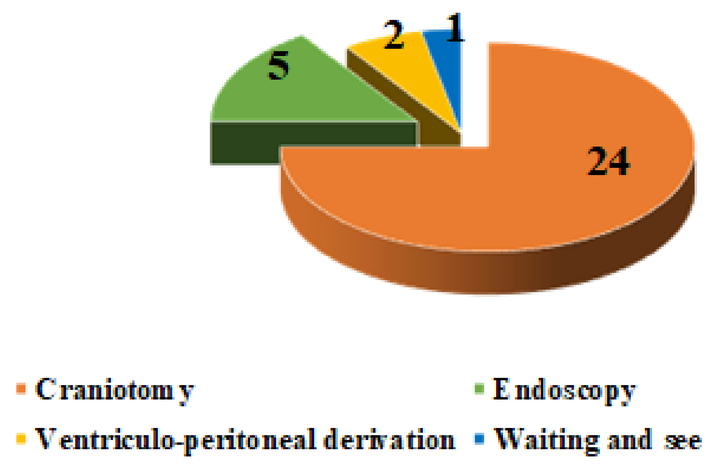
Only patients with **pubertal disorders** were **older** than others.



Surgery technique was defined for 32 patients:

- in 90.6% tumour was removed (by craniotomy in 82.8% and by transsphenoidal/transventricular approach in 17.2%);
- partial resection was found in 85.7%.

Patients who underwent craniotomy were significant younger (5.9±4.7 vs 11.8±3.1 ys; p=0.009), probably reflecting the easier feasibility of this technique.

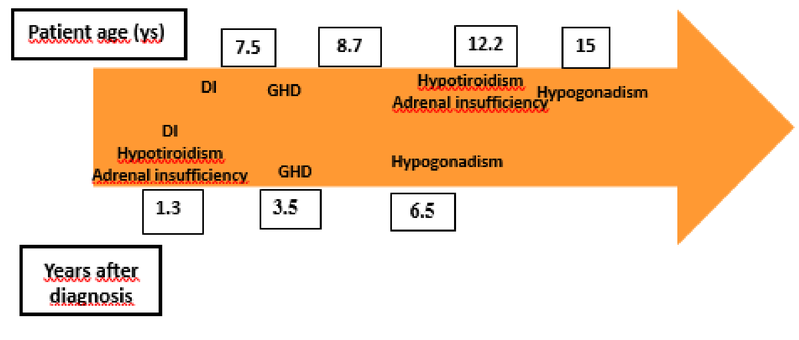


Pituitary deficiencies developed in majority of cases after diagnosis:

- hypothyroidism in 92.9%
- central adrenal insufficiency (cAI) in 91%
- DI in 78.6%,
- growth hormone deficiency in 74.5%
- hypogonadism in 73.1%.

The first disorders that appeared were hypothyroidism, cAI and DI.

Only DI was correlated with surgery approach, with a significant prevalence in craniotomy technique (p=0.001), even in case of partial resection.



Obesity at the end of follow-up showed no correlation with surgery approach/extension.

For 8 patients (4 males) we had data about body mass index (BMI) both at diagnosis and at the end of follow-up:

- all patients had normal BMI at diagnosis, but 2 patients (1 male) were obese at the last visit;
- BMI SDS at last control was significant correlated with BMI SDS at diagnosis;
- no significant differences were found in age at diagnosis, duration of follow-up, surgical approach/extension.

Conclusions:

In our cohort of patients with a long follow-up **cAI was present in >90%**, indicating the need of frequent assessment of adrenal axis function to avoid adrenal crisis.

We also found that **BMI during follow-up was related to BMI at diagnosis**.

We suggest that in every patient, especially in those with initial overweight/obesity, **criteria of MetS** (waist circumference, blood pressure, glucose and lipid profile) **should be strictly evaluated**.