

The Children's Memorial Health Institute Department of Endocrinology and Diabetology

Craniopharyngioma – symptoms, treatment and follow up – an analysis of 100 cases

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Craniopharyngioma is a rare embryogenic malformation of the sellar area with low-grade histological malignancy.

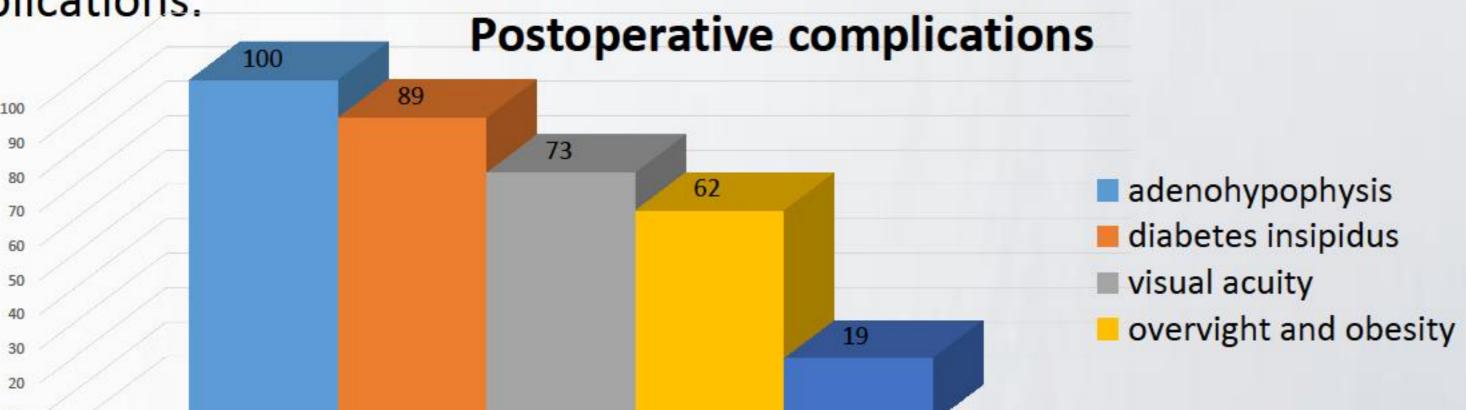
Its incidence is 0,5-2,0 cases per milion people per year, 30-50% of all cases become apparent in children.

The aim of the study was to analyze the clinical symptoms and treatment efficacy in children with diagnosis of craniopharyngioma.

A retrospective analysis included 100 children with craniopharyngioma treated in Institute in years 1999 - 2011.

Results:

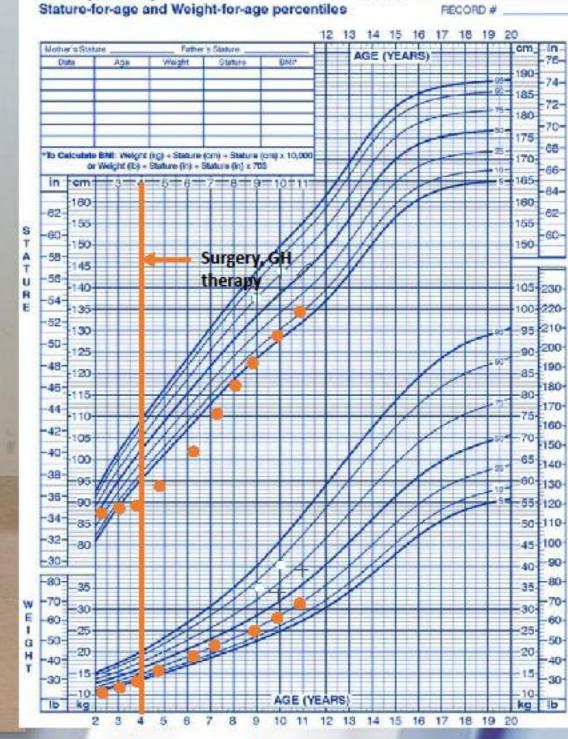
- The sex ratio was 1:1 and the median age at primary diagnosis was 8.5 years (2 weeks -18 years old).
- The symptoms observed included:
 - o endocrine deficits (78%)
 - a growth failure and decrease of growth rate (50%)
 - delayed puberty (22%)
 - secondary hypothyroidism (14%)
 - diabetes insipidus (8%)
 - adrenal insufficiency (7%)
 - cranial hypertension (57%)
 - o visual impairment (54%)
- Complete resection of tumor was achieved in 90% of patients, partial resection in 10%. 19 % of patients underwent radiotherapy
- Overall 23 patients underwent a second surgery due to a local recurrence with a median recurrence time of 2 years (7 months to 8 years old) and 4 due to tumor progression after incomplete neurosurgery.
- A second recurrence was seen in 2 patients with a median time of 5 years. This
 patients needed decompression of a cystic part of tumor.
- Most of surgical interventions were made by the same neurosurgeon (89%).
- The 5-year survival in our series is 90%, 89% at 10 years. However, 7 patients died between 0,5 5 years after surgery (median 2 years). Five of them died due to severe hypothalamo-hypophyseal disturbances, two because of neurosurgical complications.



at the age of 2 Suprasellar tumor Suprasellar tum







Conclusions:

- Craniopharyngioma is a tumor, which could cause many different symptoms depending on localization, size of the tumor and time in which the diagnosis was made.
- Surgical treatment is effective in most cases, especially if they are operated by an experienced neurosurgeon.
- Long term survival is high, however complications have serious implications on quality of life.





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