Introduction:
Cranioopharyngioma 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 (7.2±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 transphenoidal/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 yrs; 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.