

Management and treatment outcome of craniopharyngiomas in young children before 4 years of age in Italy: multicentre collection of 16 cases



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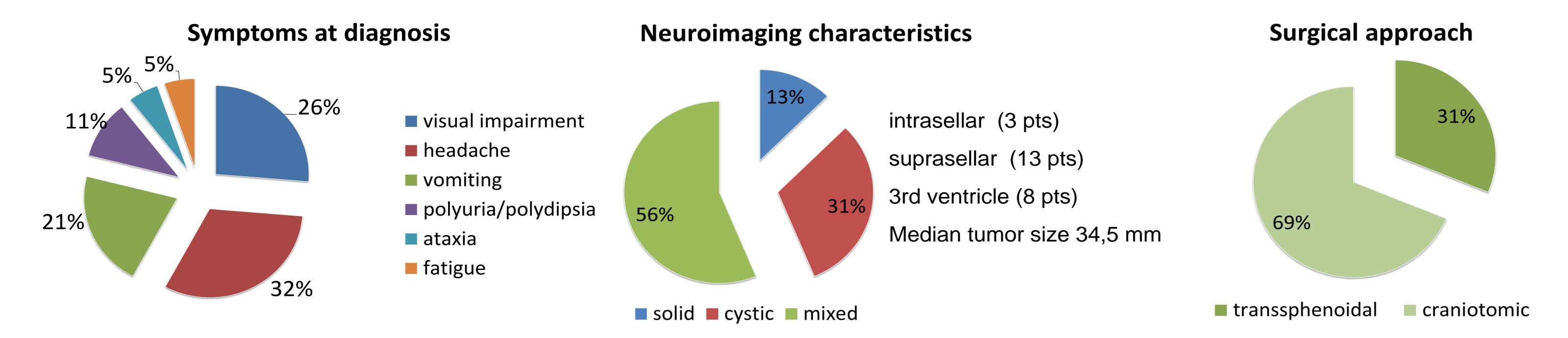
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Disclosure Statement: The authors have nothing to disclose

Background: Craniopharyngiomas (CP) are rare pediatric intracranial tumors (1.2-4%) with a peak of incidence between 5-14 years.

Objective: We retrospectively reviewed data of 16 cases (M/F 4/12) diagnosed before 4 years of age (median follow up 7.2 years) from a cohort of 117 patients (pts) (M/F 56/41) diagnosed after 01/01/2000, followed-up in 14 Italian centres of pediatric endocrinology belonging to the Italian Society for Pediatric Endocrinology and Diabetology.

Results: The pts were treated in 8 different centers across Italy. Median age at diagnosis is 2.7 years (yrs) (range 0.1-3.7) with a median delay of 6 months (0-2.3 yrs) from the symptoms onset, one pt was diagnosed prenatally



Radical tumor removal was obatined in 10/16 pts (62%).

Multiple pituitary hormone deficiencies (12/16 pts)

early after surgery started

I-thyroxin, hydrocortisone and DDAVP

hGH treatment (11/16 pts)

Median hGH dose: 0.07-0.2 mg/kg/wk

after 1.28 (0.5-2.6) years after diagnosis of CP Start:

Indication: GH deficiency and growth impairment

Overweight (4/16 pts)

3 CT/1 TS: BMI SDS 1.8-2.6

3/4 pts: hypothalamic syndrome (HS).

Conclusion:

CP in our children showed an aggressive behavior, with severe symptoms at diagnosis, relapsed in 69% of cases even in those with radical removal. In all cases at least 3 pituitary hormone deficiency arose after treatment, hGH seems not to increase the incidence of recurrences. HS is a frequent complication irrespective of type of surgery.

Recurrences

11/16 pts (69%), 1 in 4 pts, 2 in 6, 3 in 1.

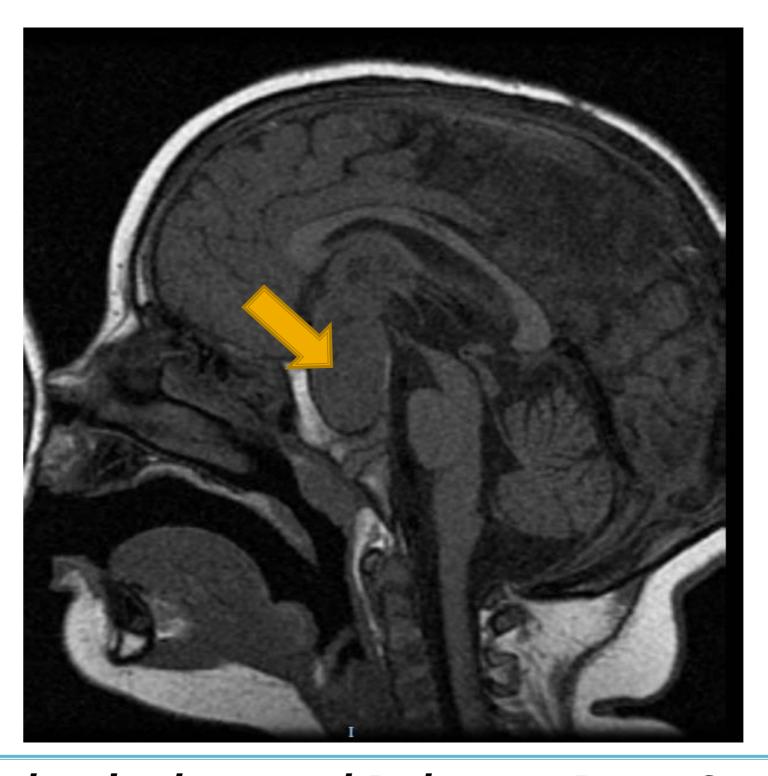
first relapse 6 months (range 0.4-4 yrs) after surgery 50% of patients relapsed despite radical tumor removal

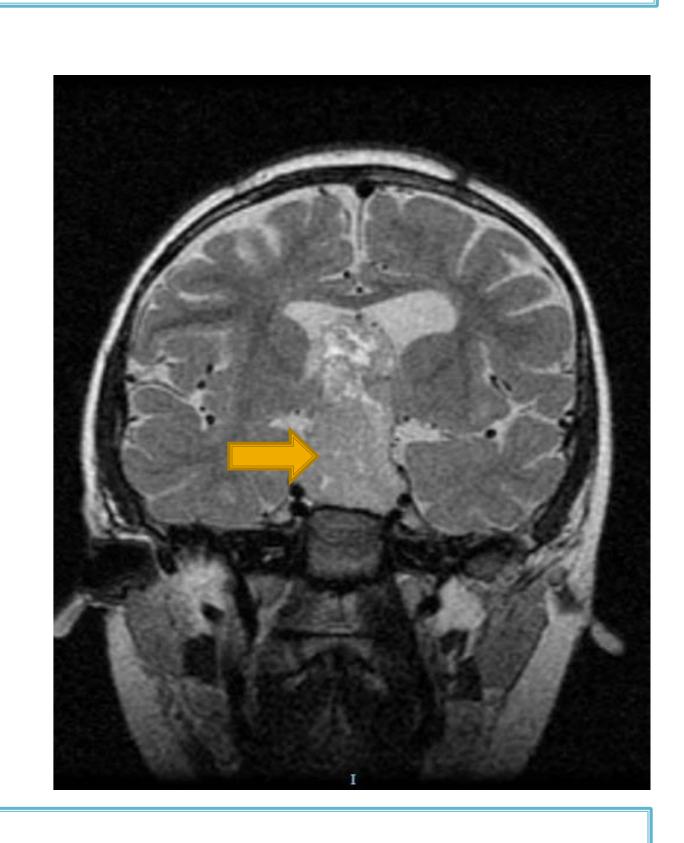
3 /11 pts relapsed during GH treatment (27%).

3/5 pts relapsed among those never GH treated (60%)

Recurrences treatment: surgical reintervention,

associated with radiotherapy in 5 cases (1 γ-knife, 2 proton, 2 conventional).





ESPE 2019- Wien - Session: Pituitary, Neuroendocrinology and Puberty - P1-106



Poster presented at:





