Management and treatment outcome of childhood-onset craniopharyngioma (CP) in Italy: multicentre collection of 117 cases

S Zucchini[1], J Fantini[1], D Mazzatenta[2], G Pozzobon[3], C Partenope[3], S Pedicelli[4], G Ubertini[4], M Parpagnoli[5], L Genitori[6], D Driul[7], R Menardi[7], P Matarazzo[8], G Tuli[8], C Guzzetti[9], L Iughetti[10], T Aversa[11], R Di Mase[12], I Rutigliano[13], ML Iezzi[14], V Cherubini[15], A Grandone[16], A Cassio[1]

[1]Pediatric Endocrinology Unit, Dept of Woman Child and Urologic Diseases AOU S.Orsola H, Bologna, Italy, [2] Dept of Neurosurgery, IRCSS Bellaria H. Bologna, [3]Dept of Pediatrics, San Raffaele Scientific Institute, Milan, [4]Endo Unit Bambino Gesù Children's H., IRCCS, Rome, [5]Auxend Ped Ginecology, Meyer Children's H., Florence, [6]Neuro-Oncology Unit, Dept of Pediatric Oncology, Meyer Children's H., Florence, [7]Division of Ped Endocrinology, Dept of Pediatrics, University of Udine, [8]SSD Endocrinology-O.I.R.M.-Città della Salute, Torino, [9]Microcitemico H, Cagliari, [10]Modena and Reggio Emilia University Modena, [11]Dept of Human Pathology in Adulthood and Childhood, University of Messina , [12]Maternal – infantile Dept AOU Federico II Naples, [13]Dept of Pediatrics IRCCS Casa Sollievo della Sofferenza, San Giovanni Rotondo, [14] Ped Clinic, Hospital District 'San Salvatore', L'Aquila, [15]Dept of Women's and Children's Health, AOU Ospedali Riuniti, G. Salesi H Ancona, [16]Dept of Woman, Child and of General and Specialized Surgery, University of Campania Luigi Vanvitelli, Naples

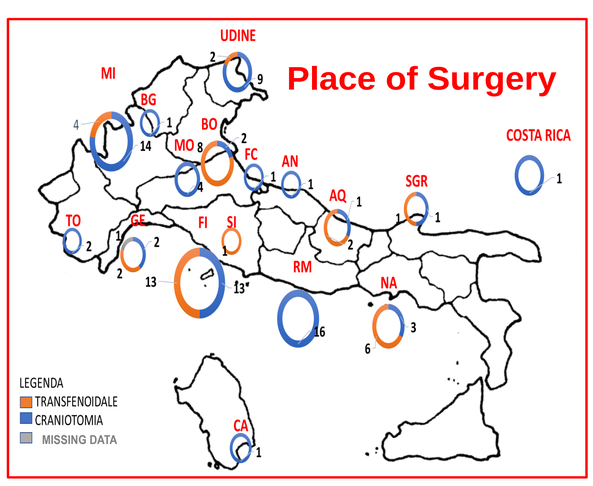
Disclosure Statement: The authors have nothing to disclose

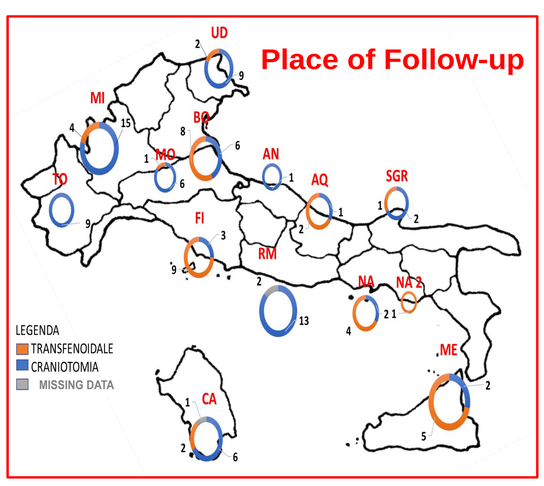
Background: craniopharyngiomas (CP) is rare pediatric intracranial tumor (1.2-4%) that is some countries is centrally managed. In Italy, treatment of the lesion differs throughout the country.

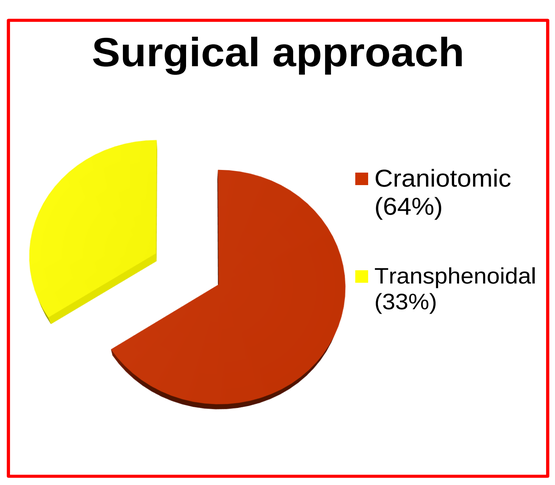
Objectives: to evaluate in Italy the management and surgical approach of children and adolescents affected by CF and followed-up in centres of pediatric endocrinology. Furthermore to evaluate complications and recurrence, including hypothalamic syndrome, according to surgical approach.

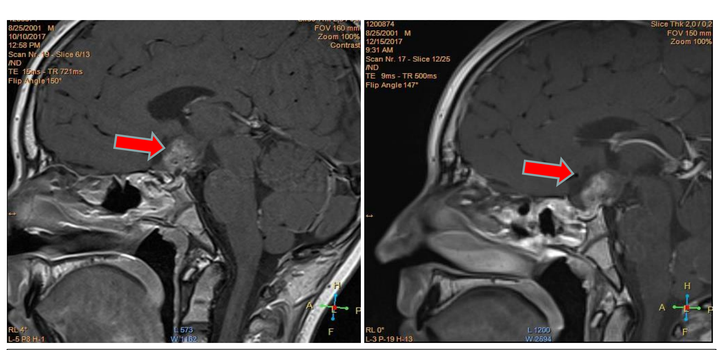
Methods: we retrospectively collected data of 117 patients (M/F 56/61) with CP and diagnosed after 01/01/2000, followed-up in 14 centres of paediatric endocrinology belonging to the Italian Society of Paediatric Endocrinology and Diabetology. Five centres provided data on more than 10 patients (range 12-19) (see upper left picture centres in red), while the remaining on 1-9. 46 pts were diagnosed between 2000-2010 and 71 afterwards. Follow-up was 7.5±4.1 years.

Results: Histology was adamantinomatous in 107 patients (91.4%), papillary in 6 patients (5.1%) and Rathke cyst in 4 (3.5%). The lesion was suprasellar in 92 patients (79%), involved the 3rd ventricle in 72 patients (61%). Age at intervention was 8.5±4 years (1 mo-18.6 yrs), with 16 cases operated before age 4 years (see poster 904 by Baronio et al.)

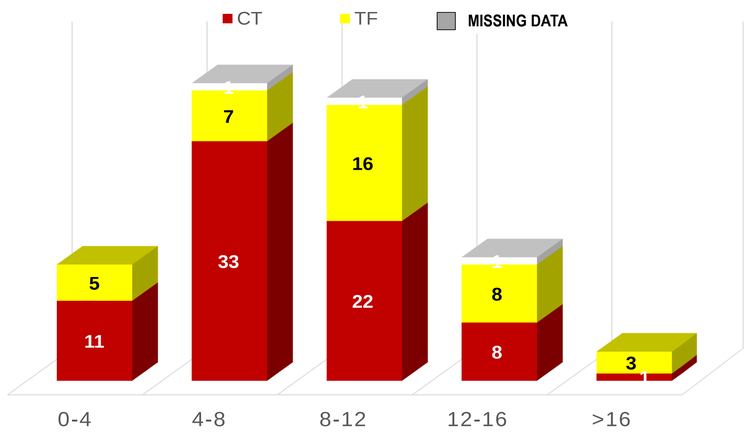




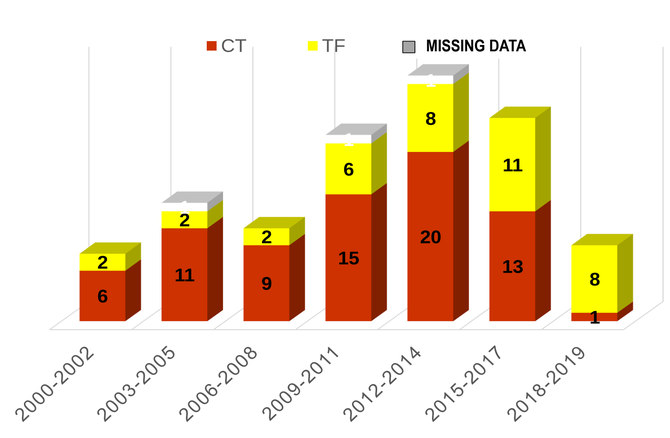




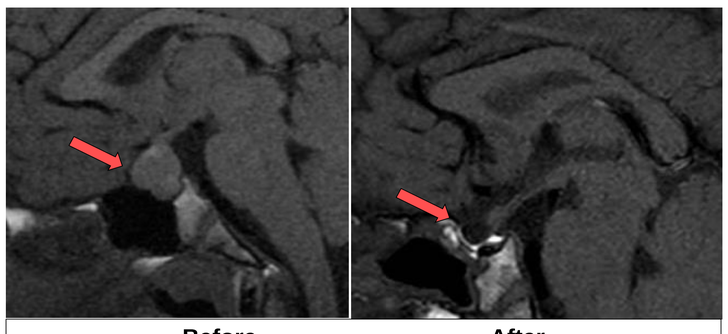
Before	After
Large suprasellar lesion: partial removal in 2017. Subsequently treated with proton therapy	



Type of surgery according to patient age (yrs)



Type of surgery according to year of intervention



Before	After
Quite homogeneous sellar/suprasellar lesion: radica removal Histologica diagnosis: Rathke cyst	

Type of Surgery changed over time (2000-2010 vs 2011-2018) with craniotomic approach declining, not significantly, from 76% (35/46) to 58% (56/71) with transsphenoidal surgery increasing from 20% (9/46) to 43% (30/71) (p=0.01).

Radical removal remained stable, from 64% (29/46) to 55% (39/71) between the 2 periods.

Post-surgery complications occurred in 58 patients (50%) (missing in 8) with Na electrolytes disorders in 19 patients followed by SAH in 12 pts and liquoral fistula (7 cases). Post-surgery complications were similar between those operated cranially (37/75; 49%) vs transsphenoidally (20/39, 51%).

Recurrences occurred in 49 patients (42%): 1 in 30, 2 in 15, 3 in 2, 4 in 1 and 6 in 1.

Overweight : in 98 patients hypothalamic syndrome was evaluated during follow-up and it occurred in 27 (27.5%). 9 patients also developed severe obesity (BMI > 2 SDS) during follow-up: this group (27+9 pts) had either craniotomic (26/64, 41%) or transsphenoidal surgery (10/34, 29%) (p=ns).

Conclusion: Italian children followed-up for CP were diagnosed and treated at all ages in several centres around the country. The proportion of patients undergoing craniotomic vs transsphenoidal surgery varied significantly among the various centres. We confirm that recurrence occurred in about 40% of patients and hypothalamic disturbances in more than 1/3 of patients irrespective of type of surgery.