Survival, Hypothalamic Obesity, and Neuropsychological / Psychosocial Status after Childhood-onset Craniopharyngioma: Newly reported Long-term Outcomes

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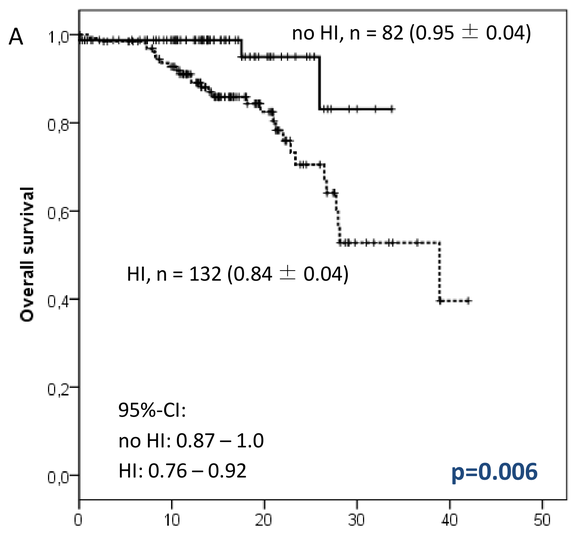
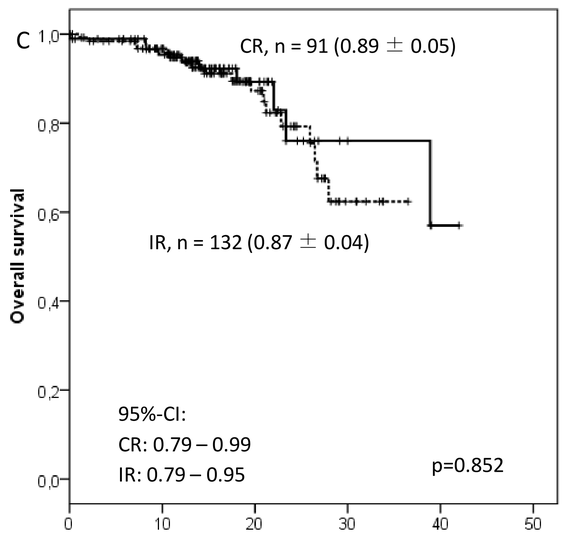
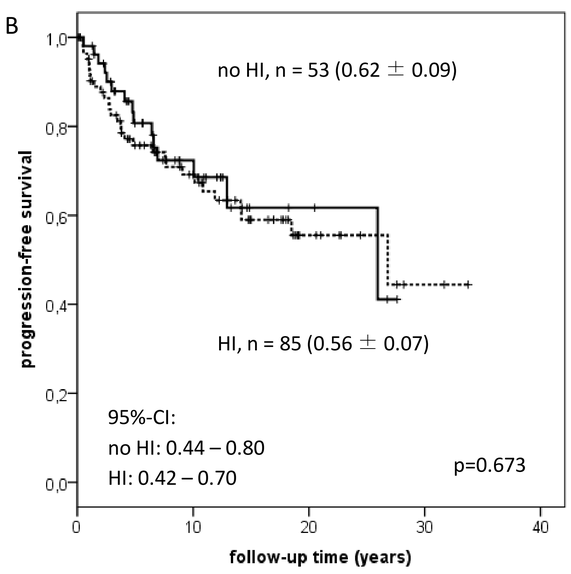
Introduction

Quality of life and long-term prognosis are often severely impaired in craniopharyngioma (CP). Knowledge of risk factors for long-term outcome is important for optimization of treatment. We analyzed long-term survival in 261 pts. with CP diagnosed before 2000.







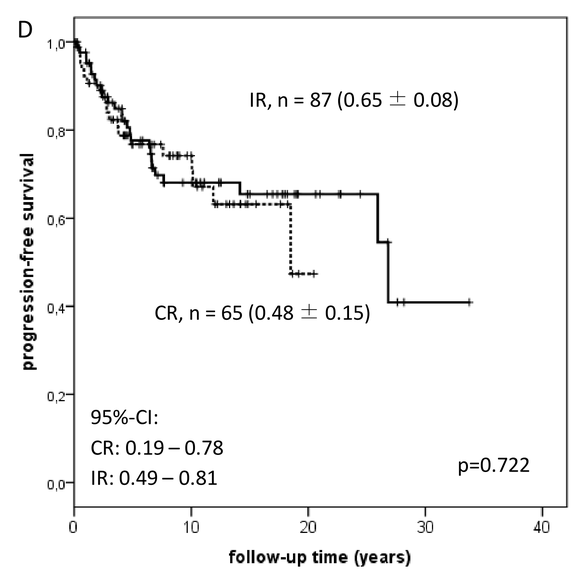


Figure 1: 20-yr overall (OS) and 20-yr progression-free survival (PFS) of CP pats. OS (Fig.1A) and PFS (Fig.1B) related to hypothalamic involvement (HI). OS (Fig.1C) and PFS (Fig.1D) related to complete (CR) or incomplete (IR) resection.

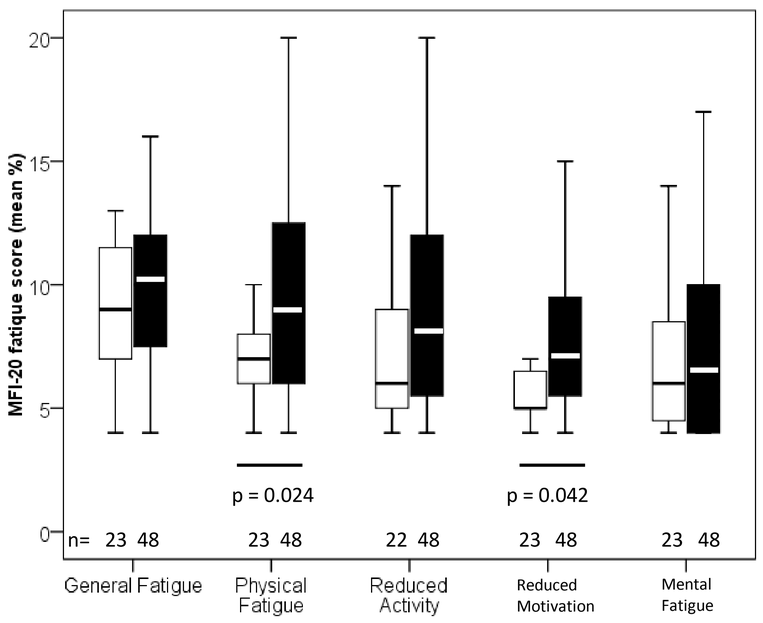
Methods

In addition to survival rates body mass index (BMI), neuropsychological status (EORTCQLQ-C30, MFI-20), and psychosocial status were analyzed in 108 of 261 patients with childhood-onset CP diagnosed before 2000 and longitudinally observed in HIT-Endo.

Patient cohort for QoL assessment

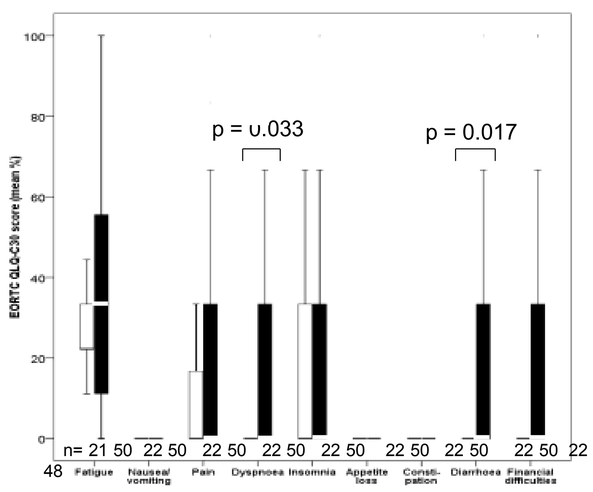
Characteristics	all	proven HI	without HI
Number of patients, n (%)	108	52 (48)	25 (23)
Age at diagnosis, (yrs)	8.1 (0.05 – 18.8)	7.6 (0.05 – 18.8)	10.1 (4.1 – 15.9)
Age at evaluation, (yrs)	24.8 (14.8–42.7)	25.4 (15.1–42.7)	25.4 (15.3–42.5)
Follow-up time, (yrs)	16.3 (9.8 – 36.4)	16.5 (10.1–36.4)	15.3 (9.8 – 29.1)
Gender (male / female)	50 / 58	27 / 25	10 / 15
Tumor location, n (%)			
intrasellar	1 (1)	0	0
extrasellar	10 (9)	8 (15)	0
intra- and extrasellar	40 (37)	28 (54)	12 (48)
n.a.	58 (53)	16 (31)	13 (52)
Tumor size (cm^2)	8.0 (1.5 – 98.8)	**12.0 (1.5–98.8)[a]**	**6.13 (1.5 – 9.0)[a]**
Degree of resection, n (%)			
total resection	44 (41)	21 (40)	13 (52)
subtotal resection	54 (50)	29 (56)	10 (40)
n.a.	10 (9)	2 (4)	2 (8)
Hypothalamic involvement			
proven HI, n (%)	52 (48)	52 (48)	
no HI	25 (23)		25 (23)
not specified	31 (29)		
Radiotherapy, n (%)	36 (33)	20 (38)	7 (28)
n.a., n (%)	13 (14)	2 (1)	1 (0.25)
Repeated surgery n (%)	23 (21), 39 n.a.	17 (33), 17 n.a.	3 (12), 5 n.a.
20 yrs Overall survival	0.98 ± 0.24	0.96 ± 0.04	1.0
20 yrs Event-free survival	0.63 ± 0.91	0.75 ± 0.08	0.63 ± 0.17
BMI-SDS at diagnosis	+0.73 (-2.7+7.0)	+0.9 (-2.6–+7.0)	-0.1 (-2.7–+4.3)

Figure 2: MFI-20 fatigue domains in CP according to hypothalamic involvement. White boxes: patients without hypothala-mic involve-ment. Black boxes: patients with hypothalamic involvement.



Used instruments / questionnaires

EORTC QLQ-C30, MFI-20, and a newly designed questionnaire created for the current study was used to assess the psychosocial status of the adult (age >20 years) long-term CP survivors of our cohort.



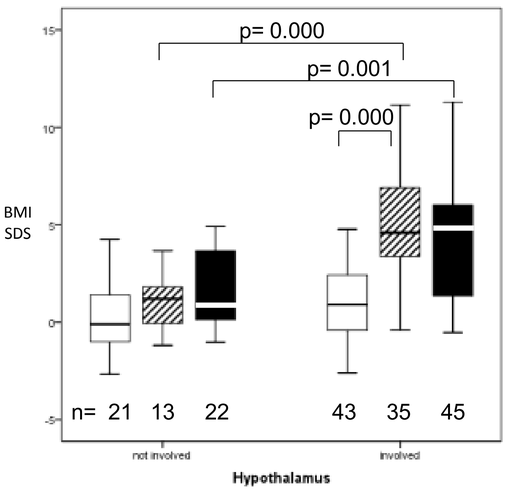


Figure 3: EORTC QLQ-C30 symptom scales in childhood-onset CP patients according to hypothalamic involve-ment. White boxes: patients without hypothalamic involvement. Black boxes: patients with hypothalamic involvement.

Figure 4: BMI development in CP pts related to hypothalamic involvement. BMI SDS is shown at time of dgx and at two intervals after dgx (8–12 yrs and >12 yr). White boxes: BMI at dgx; hatched boxes: 8–12 yrs follow-up; black boxes: >12 yrs follow-up.

Results

20-yr OS was lower (p=0.006) in CP with hypothalamic involvement (HI) (n=132, 0.84±0.04) when compared to CP without HI (n=82, 0.95±0.04). OS was not related to degree of resection, gender, or diagnosis age or year (before/after 1990). PFS was not associated with HI, degree of resection, nor gender. HI led to severe weight gain during the first 8–12 yr of follow-up (median BMI increase: +4.59SD) compared to no HI (median increase: +1.20SD) (p=0.00). During >12 yr follow-up, patients with HI presented no further BMI increase. QoL in pats with HI was impaired by obesity, physical fatigue, reduced motivation, dyspnea, diarrhea, and worse psychosocial development.

Conclusions

OS and QoL are impaired by HI in long-term survivors of CP. HI is associated with severe obesity, plateauing after 12 years. OS/PFS are not related to degree of resection, but gross-total resection should be avoided in cases of HI to prevent further hypothalamic damage, exacerbating sequelae.

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Pituitary

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