

Hydrocephalus and Hypothalamic Involvement in Pediatric Patients with Craniopharyngioma or Cysts of Rathke's Pouch: Impact on Long-term Prognosis



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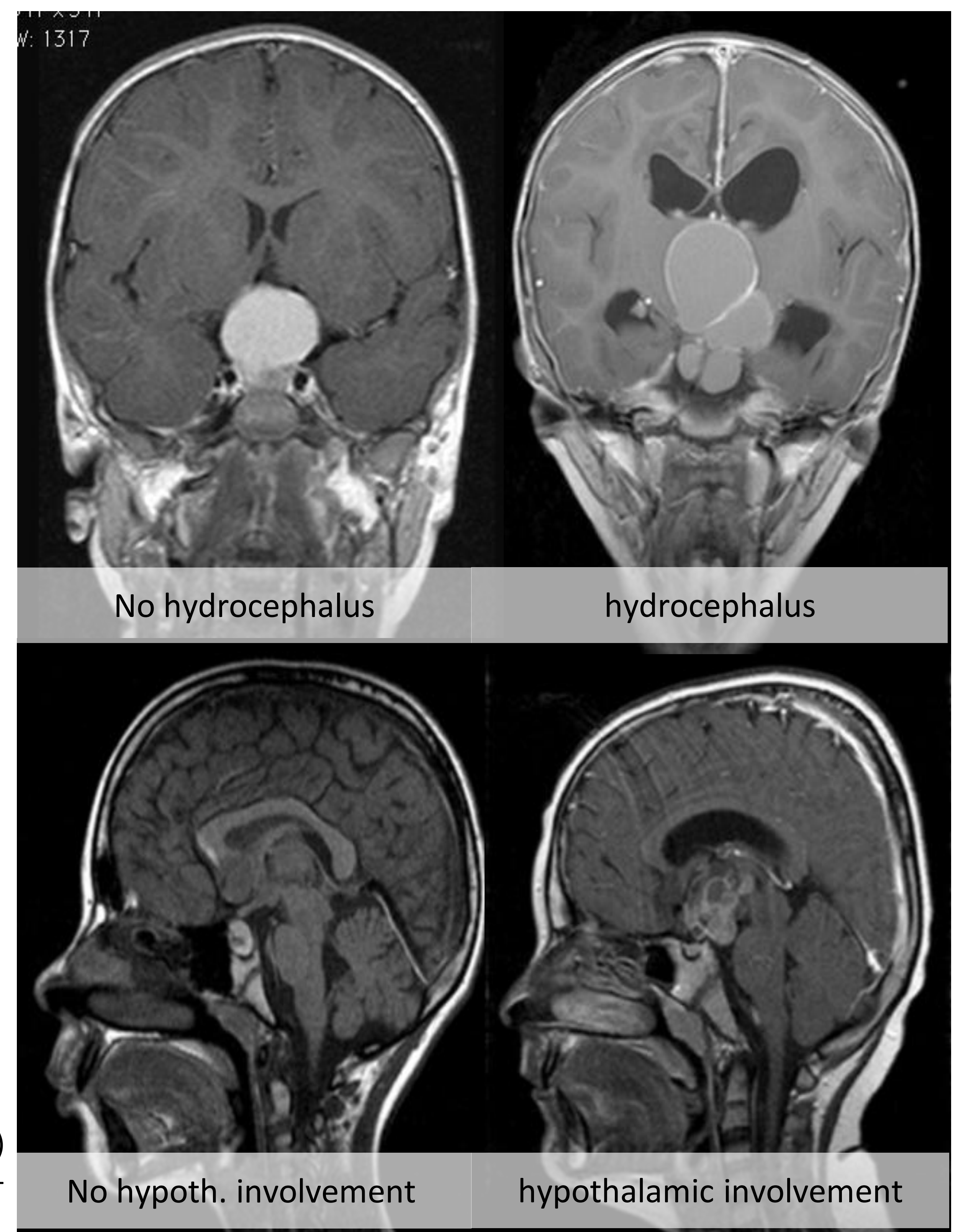
Introduction

Pediatric patients with sellar masses such as craniopharyngioma (CP) or cyst of Rathke's pouch (CRP) frequently suffer disease- and treatment-related sequelae. We analyzed the prognostic relevance of initial hydrocephalus (HY) and hypothalamic involvement (HI) on long-term survival and functional capacity (FC) in pediatric CP or CRP.

Subjects and methods

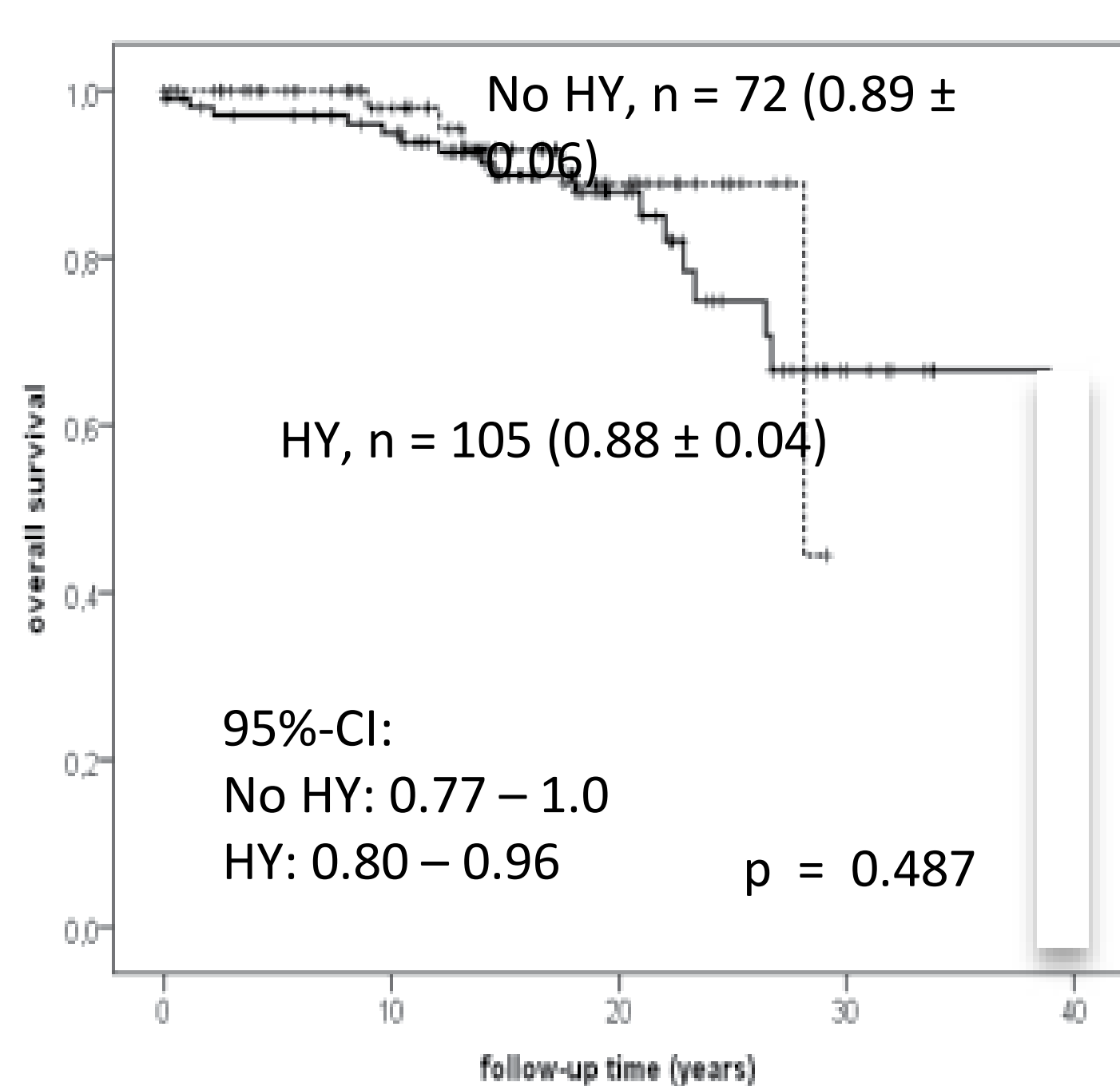
Using retrospective analysis of patient records, presence of initial HY or HI was assessed in 177 pediatric patients (163 CP, 14 CRP). Twenty-year overall survival (OS) and progression-free survival (PFS), functional capacity, and body mass index (BMI) were analyzed with regard to initial HY, degree of resection, or HI.

	Total cohort	Proven initial hydrocephalus (HY)	No initial hydrocephalus (HY)	Proven initial hypothal. Involv. (HI)	No initial hypothal. Involv. (HI)
Patients; n	177	105	72	96	69
Initial HY; n (%)	105 (59.3)	CP:103; CRP:2	CP:60; CRP:12	–	–
Initial HI; n (%)	96 (58.2)	–	–	CP:94; CRP:2	CP:57; CRP:12
Age at dx; yr, median (range)	8.8 (0.1–25.2)	7.2 ^c (1.5–25.2)	10.5 ^c (0.1–20.9)	8.9 (0.1–25.2)	9.8 (1.2–20.9)
Age at last contact; yr, median (range)	24.2 (1.5–44.8)	24.5 (1.5–44.8)	23.0 (6.3–42.7)	26.7 ^b (4.6–44.8)	20.8 ^b (1.5–43.4)
Gender; n, m/f	92 / 85	52 / 53	40 / 32	51 / 45	36 / 33
Follow-up interval; yr, median (range)	14.7 (0.0–38.9)	16.1 ^a (0.0–38.9)	13.3 ^a (0.0–29.1)	17.5 ^c (0.0–38.9)	13.6 ^c (0.0–33.7)
FMH; n, median (range)	48.5 (5–97)	47 (5–97)	50 (7–95)	47 ^b (5–97)	50 ^b (6–95)
BMI at dx; SDS, median (range)	0.44 (-3.7–7.6)	1.1 ^c (-2.2–7.6)	0.0 ^c (-3.7–7.6)	1.08 ^c (-2.2–7.6)	-0.28 ^c (-3.7–5.3)
BMI last visit; SDS, median (range)	2.3 (-1.9–13.6)	3.4 ^c (-1.9–13.6)	0.7 ^c (-1.5–9.0)	4.0 ^c (-1.9–13.6)	0.83 ^c (-1.6–9.2)
Tumor size; cm ² median (range)	8.75 (1.5–98.8)	12.0 ^c (2.0–98.8)	5.0 ^c (1.5–25.0)	12.0 ^c (2.0–98.8)	4.0 ^c (3.5–25.9)
Compl. resection; n (%)	65 (36.7)	32 (30.5) ^a	33 (45.8) ^a	30 (31.3)	30 (43.5)
Radiotherapy; n (%)	61 (34.5)	46 (43.8) ^b	15 (20.8) ^b	42 (43.8) ^b	13 (18.8) ^b
Surg. approach; n (%)					
- transcranial	109 (61.6)	73 (69.5) ^b	36 (50.0) ^b	67 (69.8) ^b	34 (49.3) ^b
- transsphenoidal	10 (5.6)	1 (1.0) ^c	9 (12.5) ^c	1 (1.0) ^b	9 (13.0) ^b

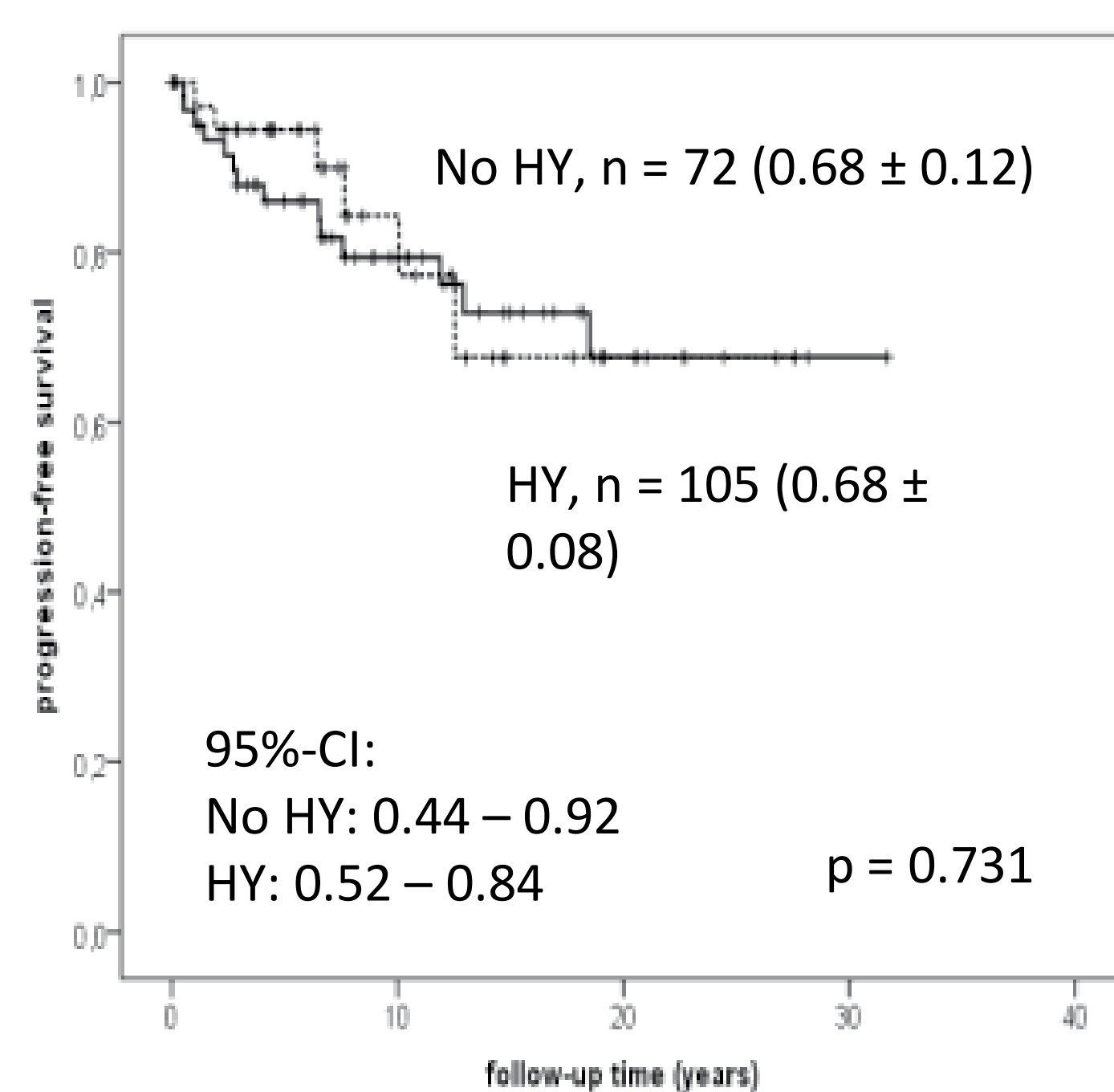


Characteristics of 177 pediatric patients with sellar masses (163 craniopharyngioma, 14 cysts of Rathke's pouch) diagnosed 1966-2001 and recruited in HIT Endo. HY hydrocephalus, HI hypothalamic involvement, VP ventriculo-peritoneal, FMH ability scale, BMI body mass index. ^a p < 0.05, ^b p < 0.01 ^c p < 0.001.

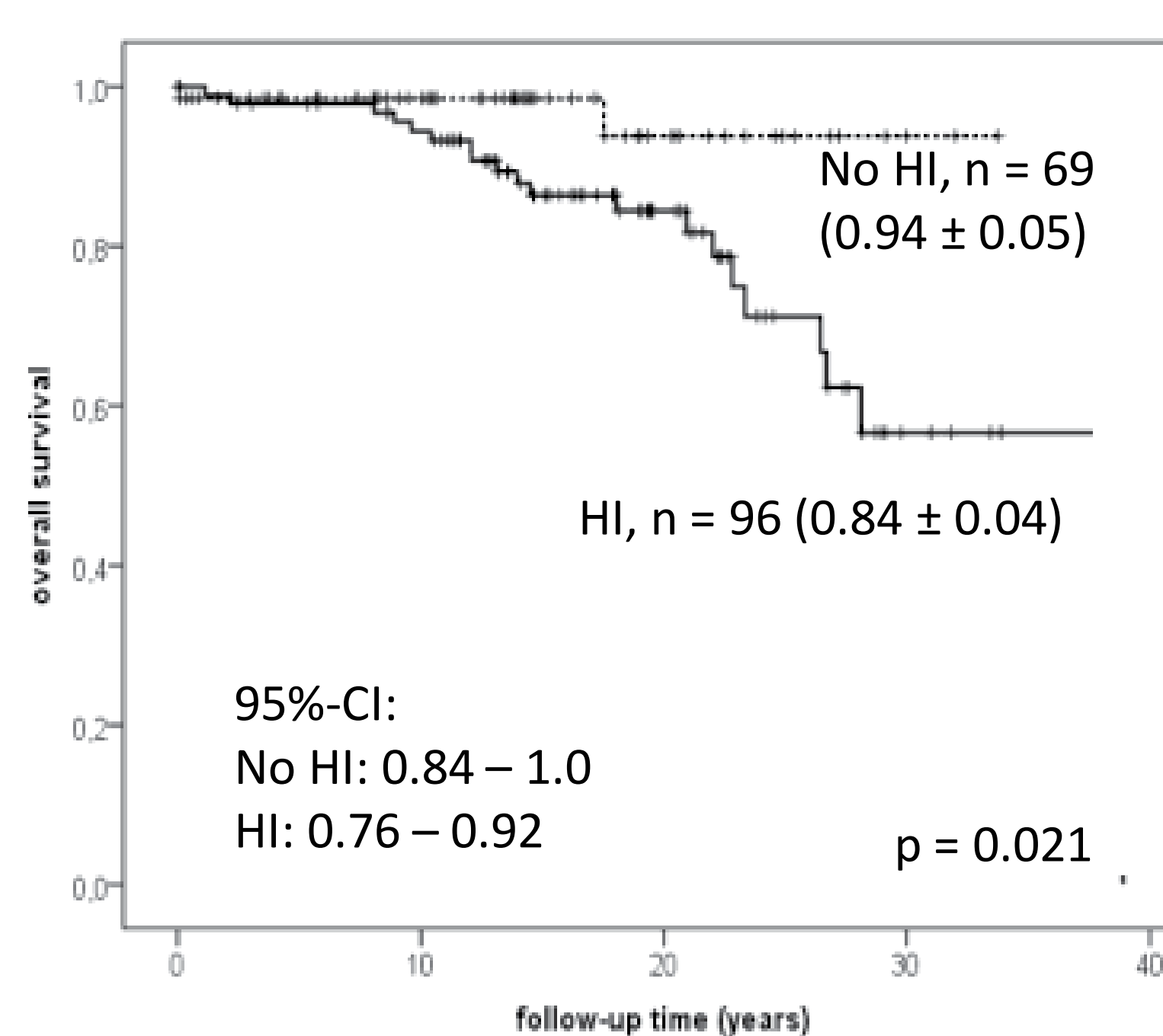
Overall survival and hydrocephalus



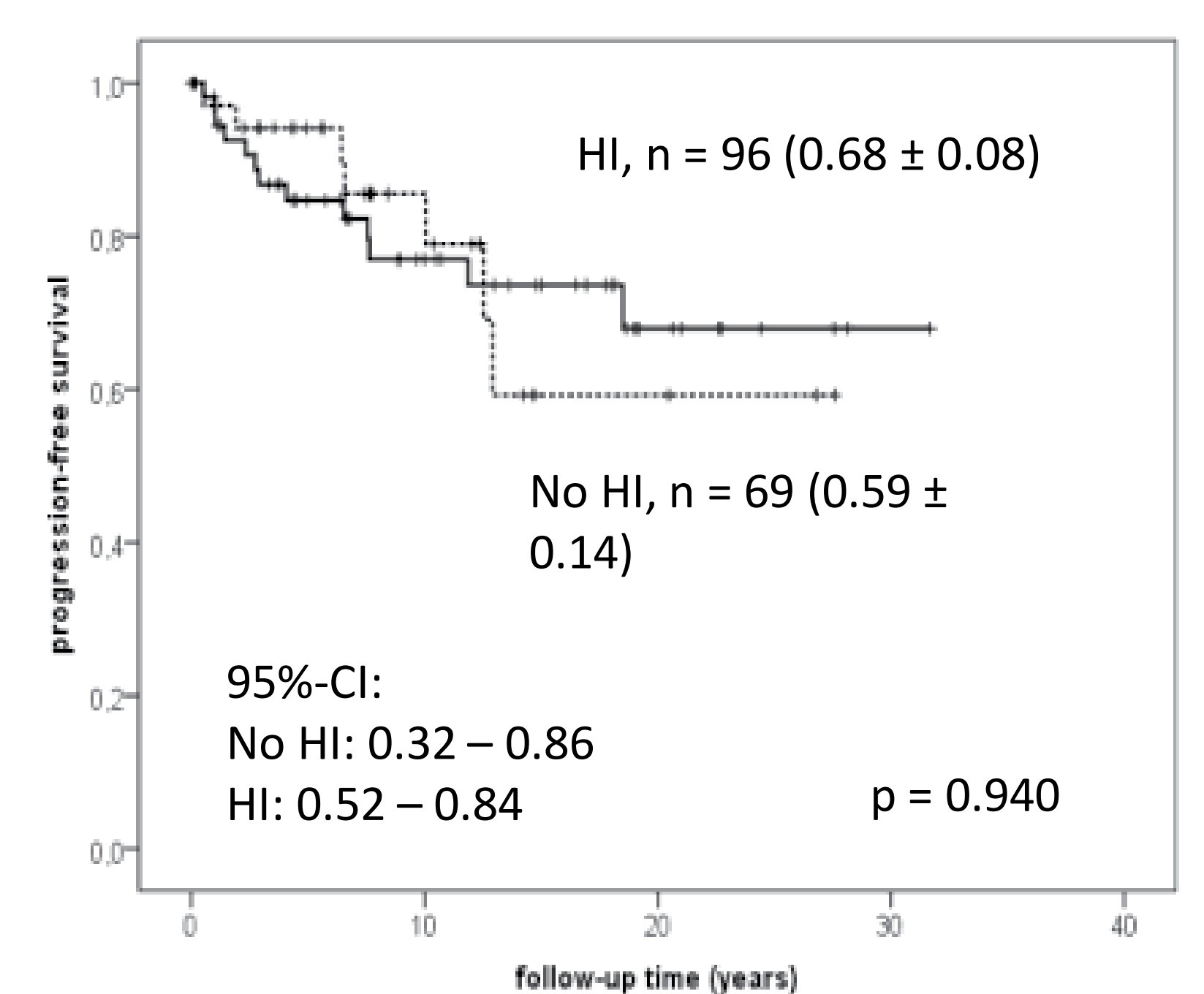
Progress. free survival and hydrocephalus



Overall survival and hypoth. involv.



Progress. free survival and hypoth. involv.



20-yr overall survival and progression-free survival rates of 177 patients (163 CR, 14 CRP) diagnosed between 1966 and 2001 related to MRI findings of hydrocephalus (HY) at the time of primary diagnosis.

20-yr overall survival and progression-free survival rates of 177 patients (163 CR, 14 CRP) diagnosed between 1966 and 2001 related to MRI findings of hypothalamic involvement (HI) at the time of primary diagnosis.

Results

105 patients (103/163 CP, 2/14 CRP) presented with initial HY and 96 presented with HI. OS, PFS, and FC were not affected by HY at initial diagnosis. Initial HI (96/177) had major impact on long-term prognosis. HI was associated with lower OS (0.84±0.04), lower FC (p=0.003), and higher BMI at dxg and last follow-up (p=0.000) when compared to pts. without HI (OS: 0.94±0.05). PFS was not affected by HI or degree of resection.

Conclusion

Initial HY has no impact on outcome in patients with sellar masses. OS and FC are impaired in survivors presenting with initial HI. PFS is not affected by HY, HI, or degree of resection. Accordingly, gross-total resection is not recommended in sellar masses with initial HI to prevent further hypothalamic damage.

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