33 year old male with a history of resected craniopharyngioma (12 years ago) presents after a seizure

Jess Hwang
9/27/12

Craniopharyngioma history

- In 2000, at age 22, he presented with headache and blurry vision was found to have a 3 x 3.2 x 3.4 cm suprasellar mass

Labs 2000 on presentation

UA: SG 1.015

Prolactin 30 (0-15)

IGF-1 144 (182-780)

FSH 2.1

LH 2.1

Total Testosterone 339 (RR 312-1240)

8AM Cortisol 19.9→27→33

8AM ACTH 45

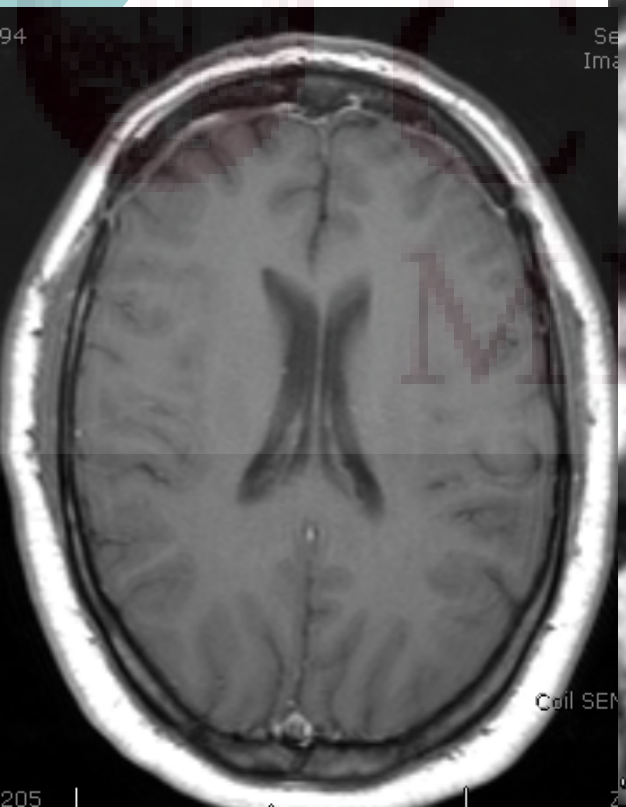
MRI 2000

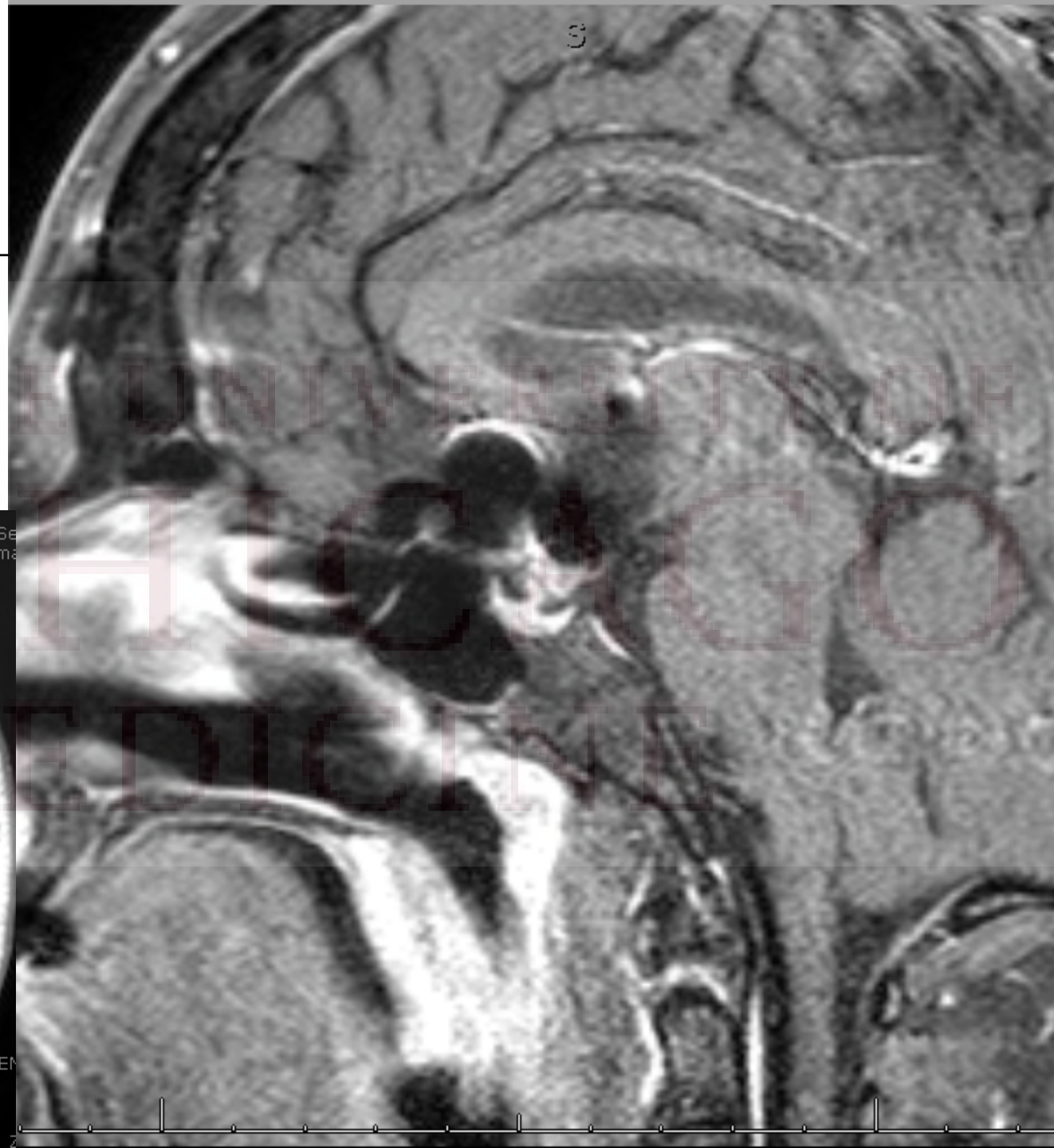
- Large cystic/solid suprasellar mass exerting significant mass effect on the basal ganglia and thalamus. The mass extends into the 3rd ventricle and herniates into the prepontine cistern.

Treatment Course 2000

- 9/13/2000 Bifrontal craniotomy for resection of mass and ventriculostomy. Pathology- craniopharyngioma.
- Completed adjuvant EBRT w/fractionated stereotactic RT in 2001
- Hormone replacement: hydrocortisone, LT4, desmopressin, testosterone

MRI 2008





HPI 9/2012

- Seizure morning of admission. First episode ever.
- Denies headache, visual changes, fevers/chills, nausea/vomiting. No focal neurologic deficits. No altered mental status per wife.
- His energy level is fair. He drinks to thirst and does not endorse polyuria or polydypsia.
- Takes hydrocortisone 10mg q9AM/10mg q9PM and synthroid 175mcg daily.

PMH

- Craniopharyngioma
- Pan-hypotuitarism
- Diet-controlled DM

FHx

- No cancer

SHx

- Lives with wife + son

Meds

- Hydrocortisone
  - 10mg 9AM
  - 10mg 9PM
- LT4 175 mcg daily

Physical Exam

Vitals: 5’4” 250lb BMI 43 133/79 88 95%

HEENT: bicoronal scars, PERLLA, EOMI, visual fields grossly full, hearing intact

Neck: no neck pain, +acanthosis nigricans

CV: RRR, no murmurs

Resp: CTAB

GI: obese, soft, non-tender

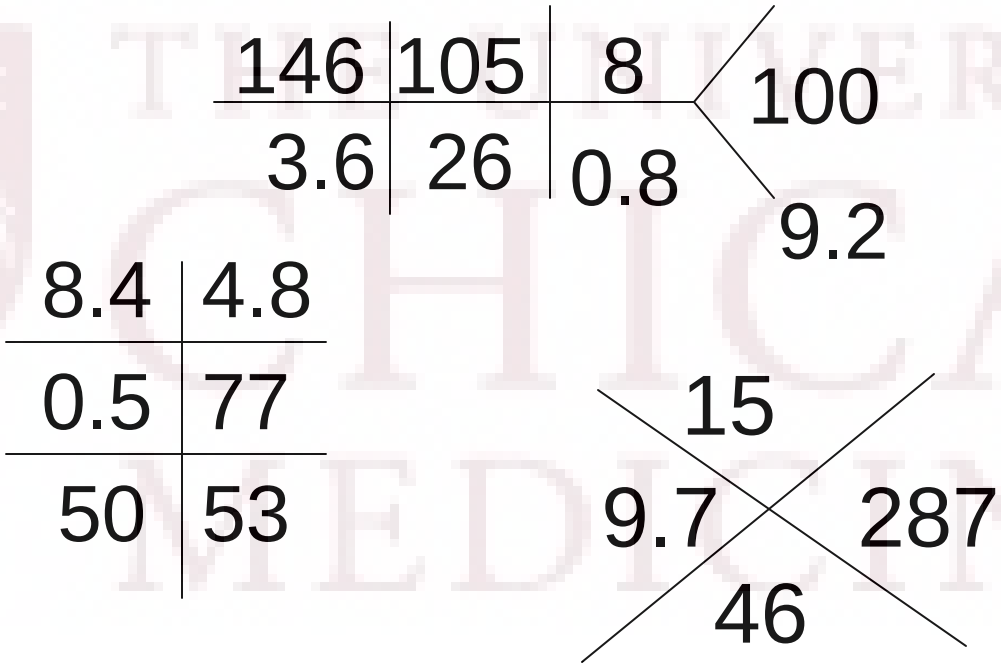
Neuro: A+Ox3, reflexes 2+, strength 5/5 BUE/BLE, CN II-XII grossly intact, sensation intact

Psych: normal mood, affect

Differential Diagnosis New Seizure

- Structural
  - Mass (recurrent CP, new mass)
  - Hemorrhage
  - Ischemic infarct
- Metabolic cause
  - Hypoglycemia
  - Electrolyte imbalance (low Na, low Ca)
- Infection (meningitis, encephalitis)
- Drugs (EtOH withdrawal)

Admission Labs 2012

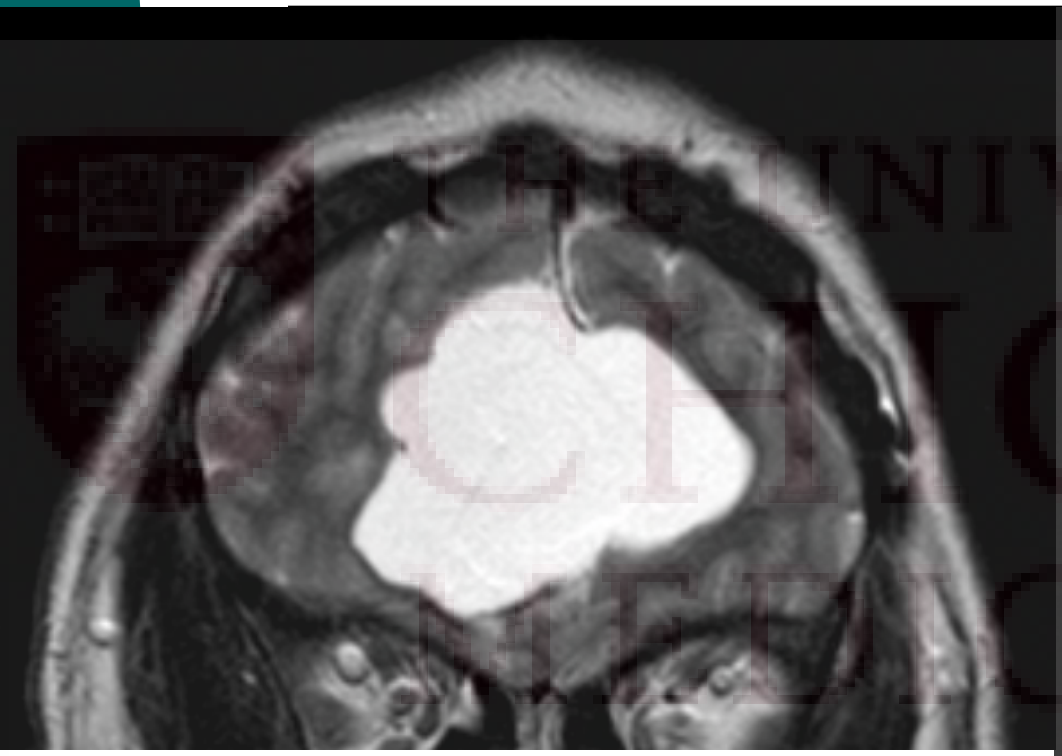


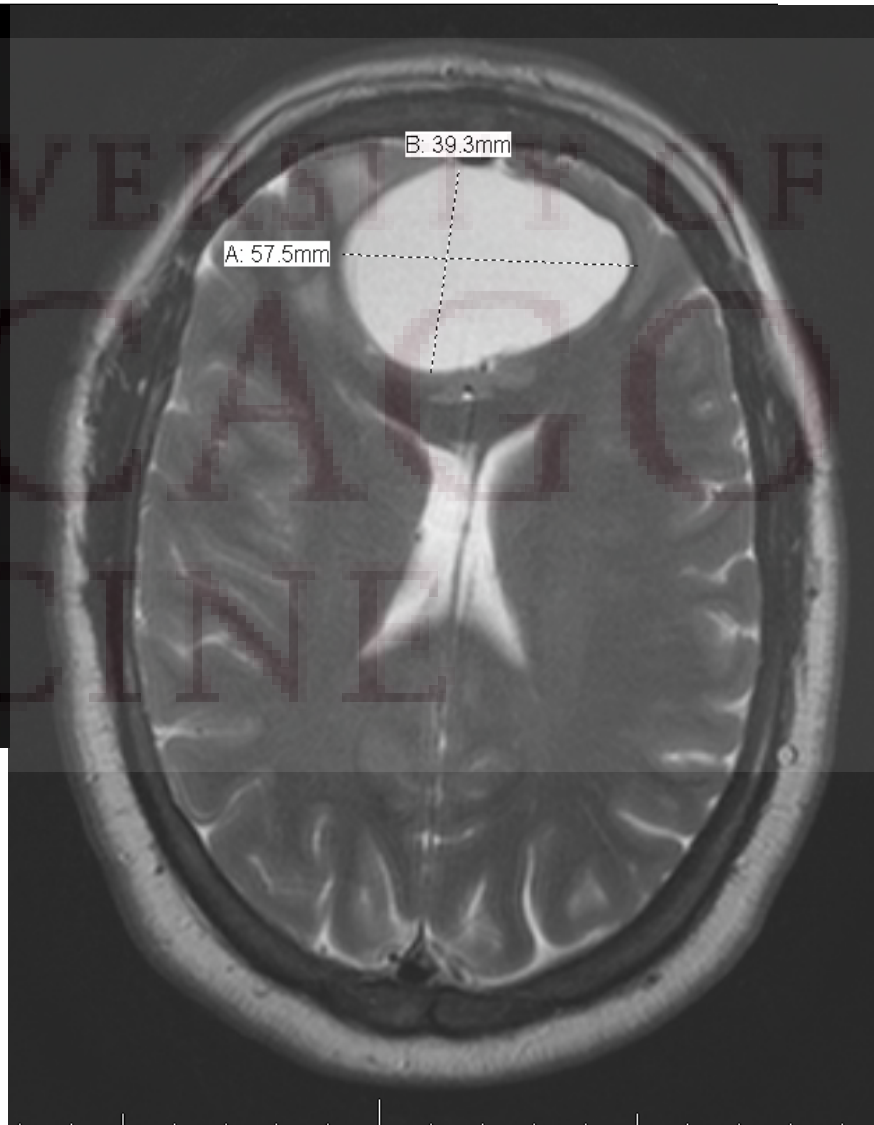
Admission Labs 2012

4PM Cortisol 2.2, ACTH 13.8
LH < 0.01
FSH < 0.01
Free Testosterone 234
Prolactin 13.53 (10-80)
TSH 0.45 (0.3-4.0)
FT4 0.75 (0.9-1.7)
HbA1c 6.9

These labs were on Hydrocortisone and Levothyroxine

MRI Brain 2012





56 mm x 39 mm x 15 mm frontal mass with mass effect, overall appearance likely represents recurrence of craniopharyngioma

- 9/14/12 Bicoronal craniotomy for resection of a craniopharyngioma.
- Pathology- recurrent craniopharyngioma

Craniopharyngioma

- 6-8% pediatric, 1-4% adult brain tumors
- Peak age in adults is usually 50-60 yo
- Second most frequent tumor in hypothalamic-pituitary axis in any age group
- Histologically benign, epithelial neoplasm but can have malignant course
- Recurrence reported in 25-70% patients

Clinical Questions

- Outcomes in childhood onset vs adult onset craniopharyngioma?
- Hormone deficits pre vs post-op?
- Morbidity: Transcranial vs Transsphenoidal approach?
- Incidence of ectopic recurrence?

Eur J Endo 2005 CO vs AO

Table 3 Data relating to GH and other hormone deficiencies (mean±S.D.).

	CO	AO	*P*
Height (cm)	165.5±10.7	168.9±9.6	0.0018
Height SDS	−0.54±1.4	−0.05±1.2	0.0001
IGF-I SDS	−4.42±2.03	−2.05±1.83	0.0001
LH/FSH deficit %	95.3	94.2	ns
ACTH deficit %	86.8	89.6	ns
TSH deficit %	94.1	92.2	ns
AVP deficit %	60.5	63.5	ns
Panhypopituitary %	57.9	59.8	ns

European Journal of Endocrinology 2005 152 557-567

Hormone Deficiency: Pre-op

TABLE 1: Clinical characteristics of 112 patients who underwent operations for craniopharyngioma according to age at surgery*

Variable	Children	Adults	Total
no. of patients	34	78	112
mean age (yrs) at surgery (± SEM)	13.2 ± 0.6	42.0 ± 1.7	33.3 ± 1.8
female sex	15 (44.1)	40 (51.3)	55 (49.1)
adrenal function deficit	16 (47.1)	40 (51.3)	56 (50)
thyroid function deficit	16 (47.1)	37 (48.7)†	53 (48.2)†
gonadal function deficit	NA	59 (75.6)	59 (75.6)
growth deficit	28 (82.3)	NA	28 (82.3)
hyperprolactinemia	7 (20.6)	24 (30.8)	31 (27.7)
diabetes insipidus	12 (35.3)	30 (38.5)	42 (37.5)
obesity	9 (26.5)	12 (15.4)	21 (18.7)
visual deficit	22 (64.7)	63 (80.8)	85 (75.9)
previous surgery	5 (14.7)	14 (17.9)	19 (17.0)
transsphenoidal approach	10 (29.4)	26 (33.3)	36 (32.1)

* Values given as number of patients (%) unless otherwise indicated. Childhood is defined as < 18 years old, adulthood as ≥ 18 years old.

J Neurosurg 2011; 114:1350-1359

Hormone Deficiency: Post-op

TABLE 4: New cases of pituitary deficiency according to the type of surgical approach*

Pituitary Hormone	Transsphenoidal	Transcranial	Total	p Value
GH	5/5 (100)	9/12 (75)	14/17 (82.3)	NS
gonadotropin	4/7 (57.1)	8/11 (72.7)	12/18 (66.7)	NS
thyrotropin	9/18 (50)	31/37 (83.8)	40/55 (72.7)	<0.03
adrenocorticotropin	10/17 (58.8)	31/37 (83.8)	41/54 (75.9)	<0.09
antidiuretic hormone	7/25 (28.0)	41/44 (93.2)	48/69 (69.6)	<0.01

J Neurosurg 2011; 114:1350-1359

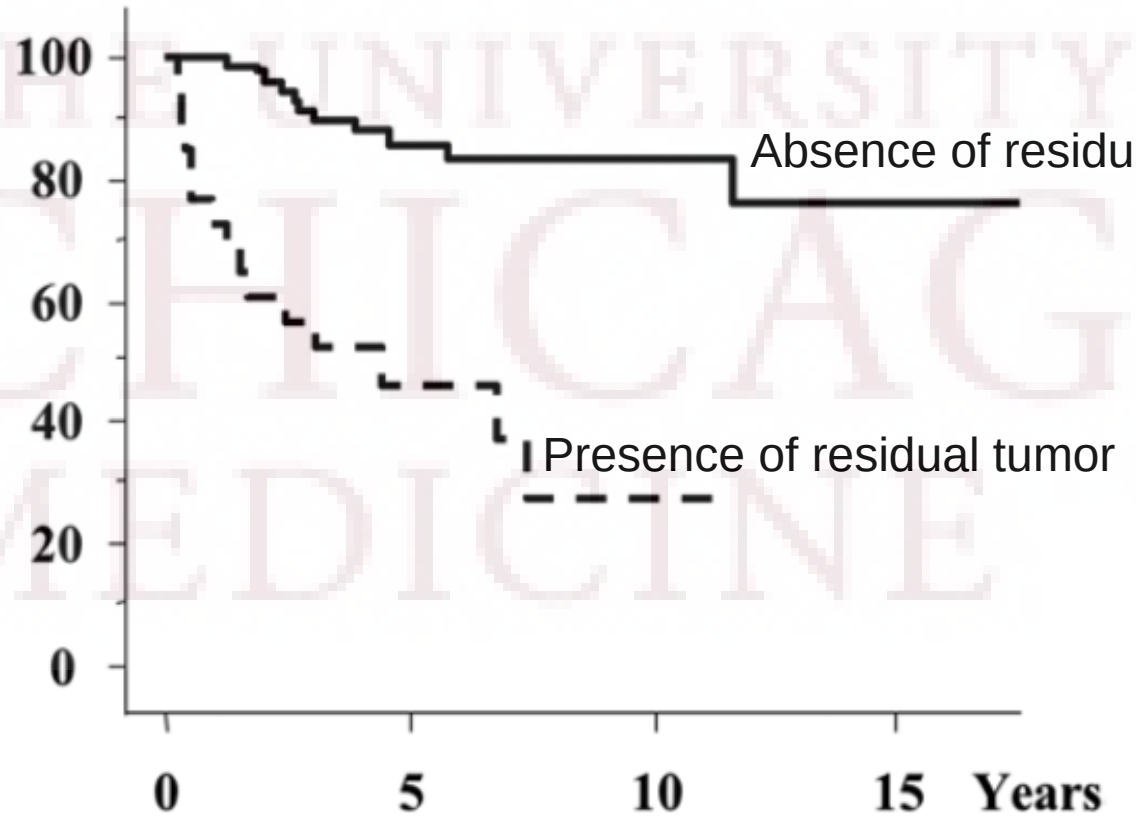
Transcranial vs Transsphenoidal

TABLE 7. Comparison of Baseline Characteristics and Outcomes Following Resection of Pediatric Craniopharyngiomas via Transcranial and Transsphenoidal Approaches[a]

Variable	Transcranial Series, % (no.)	Transsphenoidal Series, % (no.)	*P* value
Preoperative vision deficits	53.5 (1051 of 1966)	68.5 (124 of 181)	<.001
Preoperative hydrocephalus	41.7 (678 of 1625)	5.1 (5 of 99)	<.001
Preoperative increased ICP	42.6 (729 of 1713)	0 (0 of 138)	<.001
GTR	60.9 (1693 of 2780)	72.1 (199 of 276)	.0003
Recurrence after GTR	17.6 (261 of 1518)	8.0 (16 of 201)	.001
Operative mortality	2.6 (68 of 2622)	1.3 (5 of 373)	.21
Neurological morbidity	9.4 (200 of 2140)	3.1 (10 of 325)	<.001
Diabetes insipidus	69.1 (1437 of 2076)	23.9 (76 of 318)	<.001
Vision improvement	47.7 (454 of 1051)	85.5 (106 of 124)	<.001
Vision deterioration	13 (263 of 2029)	2.3 (8 of 352)	<.001
Obesity or hyperphagia	32.2 (439 of 1363)	32.1 (35 of 109)	1.00
Overall survival	90.3 (2039 of 2258)	93.9 (216 of 230)	.075

Neurosurgery 2011 Sept 69(3):630-643

Recurrence-free survival



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Ectopic Recurrence

- Remote recurrence is rare
- 2 to 26 years post-surgical resection
- 2 patterns of spread
  - Along the surgical tract
  - Dissemination through CSF
- Retrospective analysis of 86 cases showed 4 patients (4.7%) of ectopic recurrence.

Post-op Hypernatremia

	9/14	9/14	9/15	9/15	9/16	9/16	9/17	9/17	9/18
Na	151	153	154	148	153	147	155	147	149
P_{osm}					322	316			
U_{osm}				217					
ddAVP (mg PO)							0.05		
IVF	0.9NS								
	2900		700		750				
UOP	4600		5490		8800		3050		
PO	NPO		4620		6540		2200		

Hospital course

- Post-op hypernatremia
- Discharge meds
  - Lantus 18U qday, Novolog 6U qAC
  - Levothyroxine 175mcg daily
  - Decadron 4mg BID taper
  - 0.05 mg PO ddAVP prn

Take home points

- Hormone deficiencies common both pre- and post-op
- Possible complications from TC vs TS approach
- Ectopic recurrence is rare but does happen so long-term follow-up of at least a decade or more of serial imaging is important to identify this early

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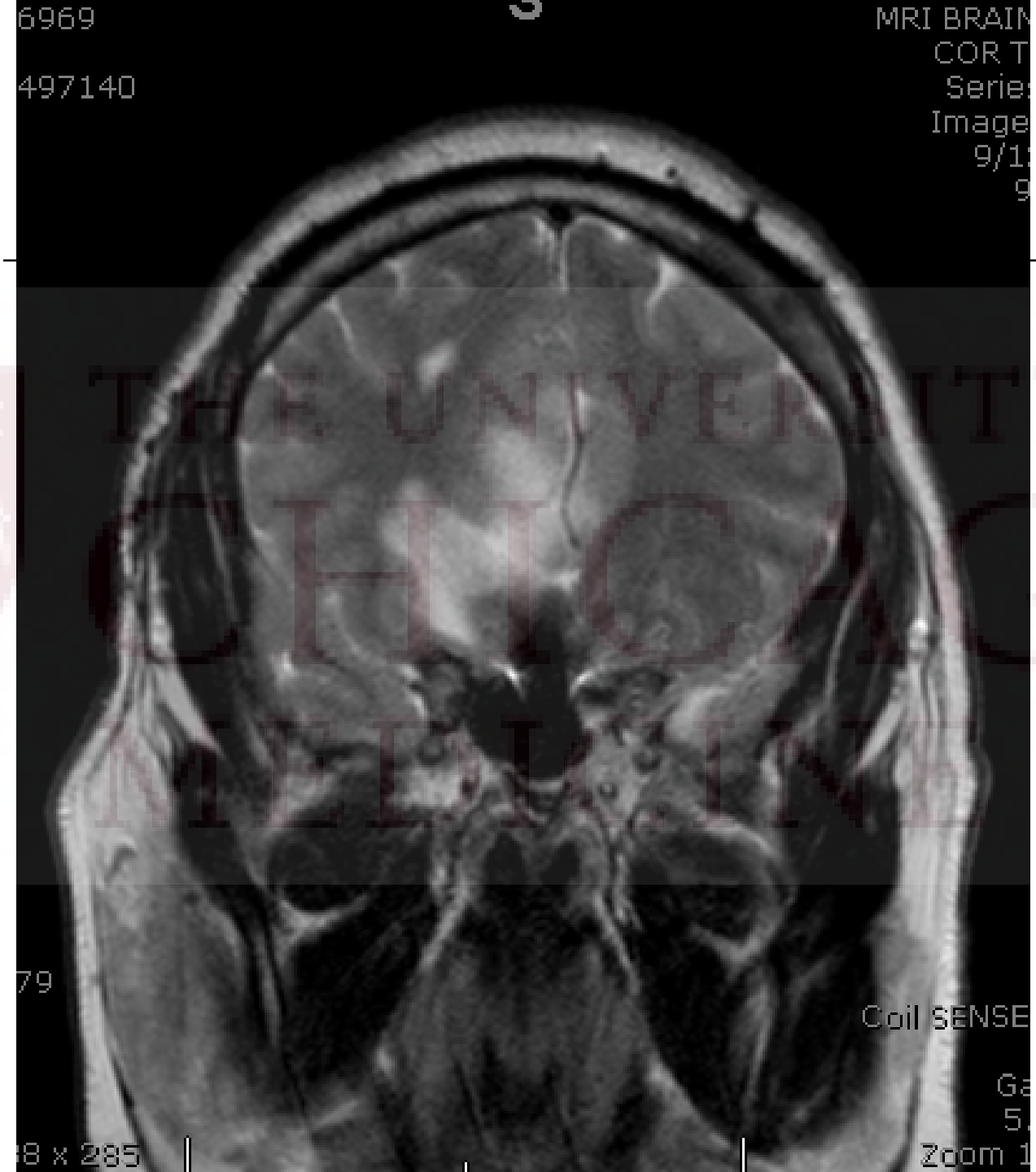
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