

# PINEAL REGION TUMORS

MODERATORS :  
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# Arterial supply

- PCA
  - P1
    - Quadrigeminal a.— superior colliculi
  - P2
    - M P.ch
      - Pineal body, corpora quadrigemina, tela choroidea ventriculi tertii, thalamus
    - L P.ch
      - Choroid plexus of lat. Ventricle, LGB, thalamus
- SCA
  - Inferior colliculi

## Venous drainage

- Internal cerebral vein
- Basal Rosenthal vein
- Vein of Galen and its tributaries.

# SURGICAL ANATOMY

- Most tumors arise from or are attached to undersurface of velum interpositum
- Tumors rarely extend above velum
- Blood supply comes mainly from M P.ch & L P.ch with anastomoses to pericallosal & quadrigeminal a.
- Most tumors are centered at pineal gland, some extend to For. Monro.

# SURGICAL ANATOMY

- Mostly, ICV, Galen , Rosenthal & precentral cerebellar veins surround or cap the periphery of these tumors.
- Rarely, ICV are ventral to tumor.
- Most tumors are not highly vascular except
  - Pineoblastomas
  - Hemangioblastomas
  - Hemangiopericytomas (Angioplasmic meningioma)

# Pathology

New York Neurological institute data  
(1981-2001)

Germ cell tumors	-32%
Pineal cell tumors	-25%
Glial cell tumors	-27%
Miscellaneous	-15%

Data from Youmans neurological surgery ,4th edition

# Differential diagnosis

- GERM CELL TUMORS
  1. Pure germinoma
  2. Teratoma
  3. Embryonal cell carcinoma
  4. Endodermal sinus tumor
  5. Choriocarcinoma
  6. Mixed germ cell tumor
- PINEAL PARENCHYMAL TUMORS
  1. Pineoblastoma
  2. Pineocytoma
  3. Pineal parenchymal tumors of intermediate differentiation

# Differential diagnosis

- TUMORS OF SUPPORTIVE TISSUES & ADJ. STR.
  1. Astrocytoma
  2. Ependymoma
  3. Meningioma
  4. Hemangiopericytoma
  5. Ganglioglioma
  6. Ganglioneuroma
  7. Chemodectoma
  8. Quadrigeminal cistern lipoma
- Metastatic tumors (very rare)
  - Lung, breast, stomach, kidney
- Nonneoplastic lesions
  - Pineal cysts
  - Arachnoid cysts
  - Cysticercous cysts
  - Vascular lesions
    - Aneurysmal dilatation of vein of Galen

# Epidemiology

- Tumor in this location are more common in childhood( 3-8 % of childhood brain tumors) than adult (1 %).
- Germ cell tumors: pediatric population, more in boys.
- Pineal cell tumors : young adults, M=F
- Glial cell tumors and meningioma: more in adults.
- Heritability: rare : E.g. Trilateral retinoblastoma [ Pinealoblastoma + b/l retinoblastoma, gene deletions ]



# Clinical features

- Symptoms-
  1. Raised ICP [Hydrocephalus]
  2. Direct brainstem compression-
    - A) PARINAUD SYNDROME
      - Paralysis of up gaze
      - Convergence/ Retractory nystagmus
      - Light-near pupillary dissociation
    - B) SYLVIAN AQUEDUCT SYNDROME
      - Paralysis of downgaze/ horizontal gaze
    - C) 4<sup>th</sup> CN PALSY
      - Diplopia

# Clinical features

## D) Inferior colliculi compression

- Hearing disturbances

## E) Inv. Of superior cerebellar peduncle

- Ataxia

## 3. Endocrine dysfunction (hypothalamic invasion/HCP)

- Diabetes Insipidus
- Precocious puberty[boys] : b- h CG secretion in choriocarcinoma/Germinoma

## 4. Pineal apoplexy

- In vascular tumors : Pineal cell tumors/Chorioca.

# MRI

- MRI with gadolinium contrast is gold standard for diagnosis and planning surgery
  - Degree of HCP
  - Size of tumor: lateral & superior extent
  - Vascularity
  - Homogenous/heterogeneous
  - Irregularities of margination & probability of invasion
  - Anatomical relationship of tumor

	Germinoma	Teratoma	Pinealoblastoma	Pinealocytoma	Glioma	Meningioma
Age	Child	Child	Child	Adult	Young	Adult
Sex	male	male	-	-	-	-
Location	pineal	pineal	pineal	pineal	parapineal	parapineal
Signal int.	Homoge. (+- h'rge)	Strikingly heteroge.	Homoge.	Homoge.	Homoge.	Homoge.
H'rge	common	typical	common	common	rare	rare
Calcification	rare	typical	common	common	uncommon	common
Edema	common	variable	common	Uncomm.		Occ.
Mets	yes	variable	yes	No	variable	No
Enhancement	Dense	variable	Dense	Variable	variable	Dense.

# CSF Levels of tumor markers in germ cell tumors

	Alpha fetoprotein(<5 ng/ml)	Beta-HC G(<5 IU/ml)	PLAP	Melatonin
Germinoma	-	+ ( < 770 )	++	-
Mature teratoma	-	-	-	-
Immature teratoma	+/- ( <1000)	-		
Choriocarcinoma		+++ ( >2000)		-
Yolk sack tumor	+++		+/-	-
Embryonal ca.	++ (1000)	+ ( < 770 )	+	-
pinealocytoma	-	-	-	+/-

# MANAGEMENT OF HYDROCEPHALUS

- Symptomatic pt's:-

ETV is best

1. Allow gradual reduction of intracranial pressure.
2. Eliminate complications ass. With VP shunt.
3. Biopsy can be taken

- Mildly symptomatic pt's:-

If gross total resection is anticipated –

Put EVD at time of surgery and remove/ convert to vp shunt as management dictate.

# Management strategy

- Test dose radiation- concept is controversial.
- Histological diagnosis is necessary to optimize the treatment as diverse pathology occurs and individual tumor influences-
  - adjuvant therapy
  - Metastatic workup
  - Prognosis & long term follow up.
- CSF cytology and radiology not sufficiently sensitive
- Only time when biopsy is unnecessary is when malignant germ cell markers are available.

# Tissue diagnosis

- Options are-

Stereotactic or open biopsy or endoscopic

- OPEN BIOPSY

- More extensive tissue sampling in view of heterogeneity & mixed cell population
- Debulking: favorable response to adjuvant therapy
- 1/3 rd tumors are benign and tumor resection are complete and curative
- Shunting can be avoided in the pt. with mild HCP



# STEREOTACTIC BIOPSY

- STEREOTACTIC BIOPSY

Indication-

Suited for patients with primary systemic tumors , multiple lesions, clinical conditions that contraindicate open surgery/general anesthesia, radiologically invasion of brainstem.

Advantage –

- Relative ease of performance and reduced complication
- Resection may not be necessary depending on the histopathology
- Biopsy may guide effective non-surgical therapies

Disadvantage-

- Small biopsy volume : Difficulty in diagnosis
- Experienced neuropathology team required
- Associated with risk of hemorrhage

# STEREOTACTIC BIOPSY

- **ANTEROLATEROSUPERIOR APPROACH:**
  - Low precoronal entry point just behind the hairline & just above the superior temporal line.
  - Needle trajectory: Through the frontal lobe , underneath the lateral ventricle & lateral & inferior to ICV.
- **POSTEROLATEROSUPERIOR APPROACH:**
  - Entry point near PO junction, for tumors extending laterally or superiorly.

Multiple specimens to be obtained.

Side cutting cannula preferred over cup forceps

Hemorrhage: Continuous suction & irrigation x 15 min.

# Endoscopic biopsy

- Through 3<sup>rd</sup> ventricle.
- Generally combined with ETV
- Drawbacks - difficult procedure
  - sampling error.
  - difficult to control bleeding
- CACE  
( computer assisted cisternal endoscopic approach)

# CACE

( computer assisted cisternal  
endoscopic approach)

- Novel technique.
- Use frameless stereotactic principle and endoscope through post fossa keyhole approach.
- 4 quadrant biopsy is taken.
- Till awaits clinical use.

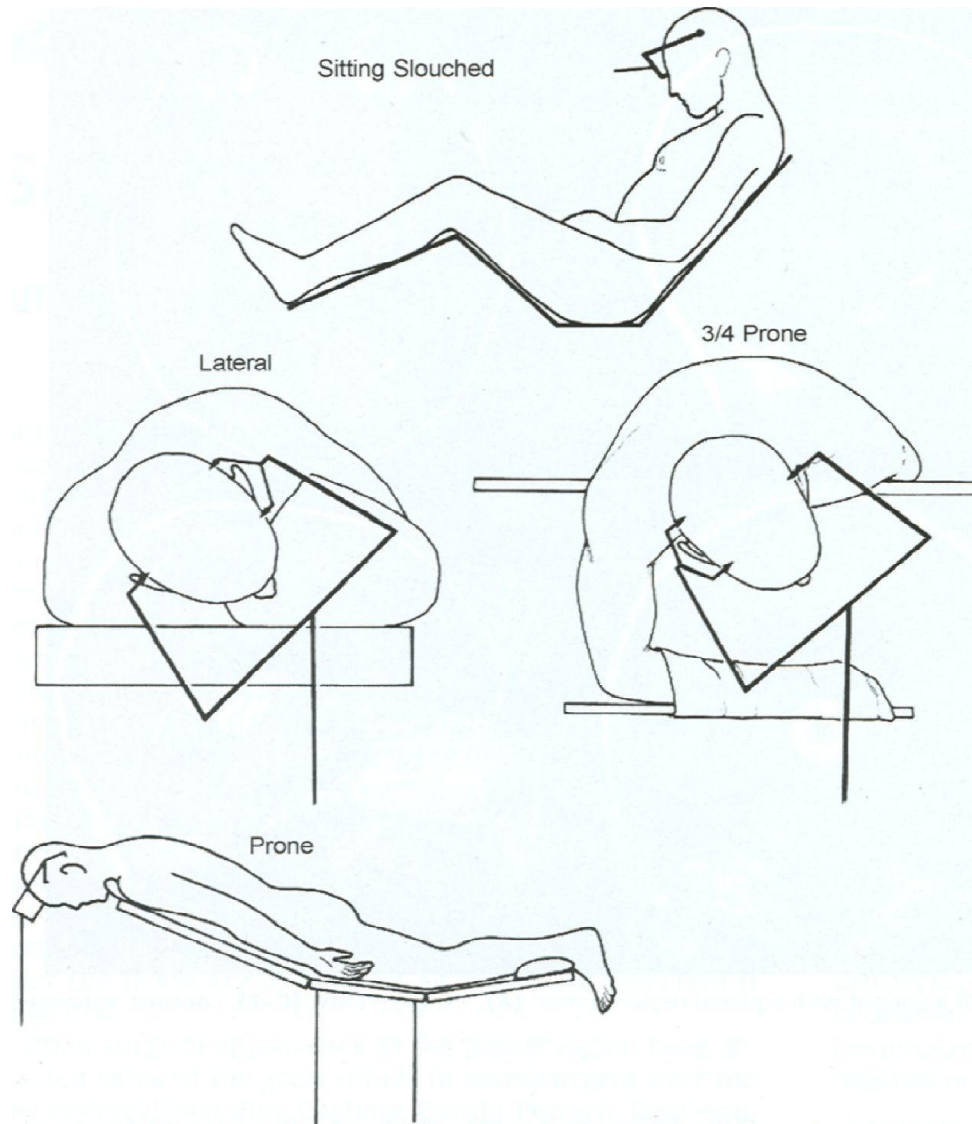
[Youssef AS](#), [Keller JT](#), [van Loveren HR](#).

## Source

Department of Neurosurgery, University of South Florida, Tampa, Florida,  
USA. [editor@mayfieldclinic.com](mailto:editor@mayfieldclinic.com)

[Acta Neurochir \(Wien\)](#). 2007;149(4):399-406. Epub 2007 Feb 26.

# Patient positioning



# Operative approaches

- **INFRATENTORIAL-**

1. Infratentorial-supracerebellar
2. Paramedian Infratentorial-supracerebellar

- **SUPRATENTORIAL-**

1. Occipital transtentorial
2. Post./ parietal transcallosal interhemispheric
3. Ant. Transcallosal transventricular transvelum interpositum
4. Post. Transcortical transventricular
5. Transcallosal interforncial
6. Others

- **COMBINED INFRA AND SUPRATENTORIAL APPROACH**

# Operative approaches

- HISTORICAL PERSPECTIVE

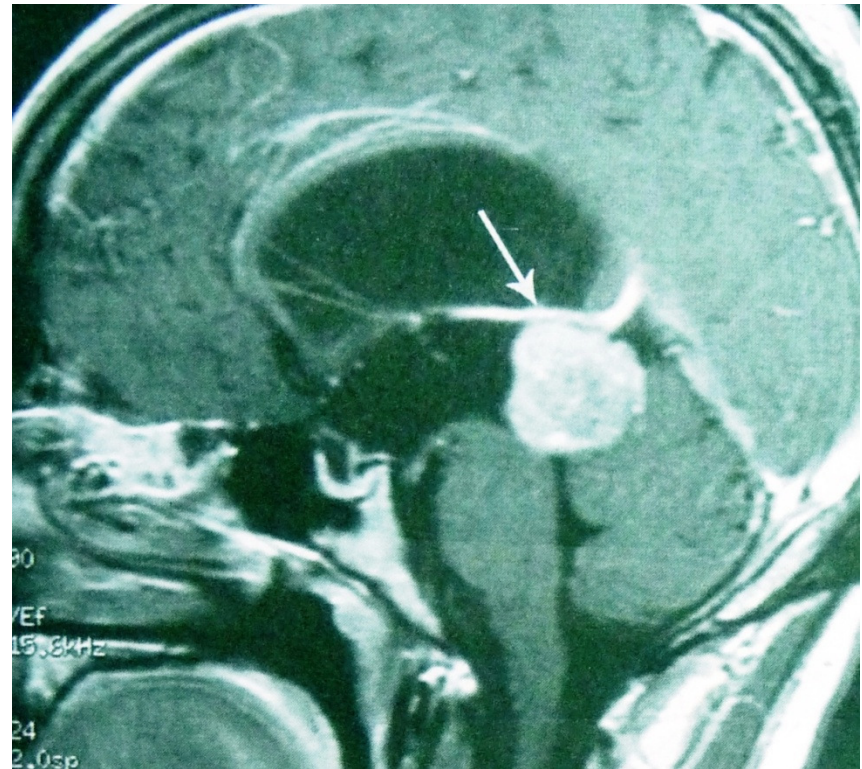
- Infratentorial supracerebellar [Horsley, Krause (1913), Stein (1971)]
- Parietal transcallosal [ Dandy (1921), Kunicki ]
- Posterior transventricular [ Van Weegen, 1931 ]
- Parieto-occipital with splitting of tentorium/splenium [Heppner, Poppen and Marino, Glasauer, Jamieson, Lazar & Clark]

- HISTORICAL PERSPECTIVE
  - Transvelum interpositum [Sano]
  - Lateral paramedian infratentorial [ Van den Bergh, 1990]
  - Other approaches [ TRANSCALLOSAL :  
transforaminal, interfornicial,  
subchoroidal, transchoroidal] – primarily for  
ant./mid 3<sup>rd</sup> ventricle tumors.



# INFRATENTORIAL SUPRACEREBELLAR

- INDICATIONS
  - Tumor with major bulk in midline
  - Tumor ventral to velum interpositum & deep venous system
- ADVANTAGES
  - Minimal risk to deep veins
  - No normal neural tissue violated en route
  - Exposure comparable with that of other routes

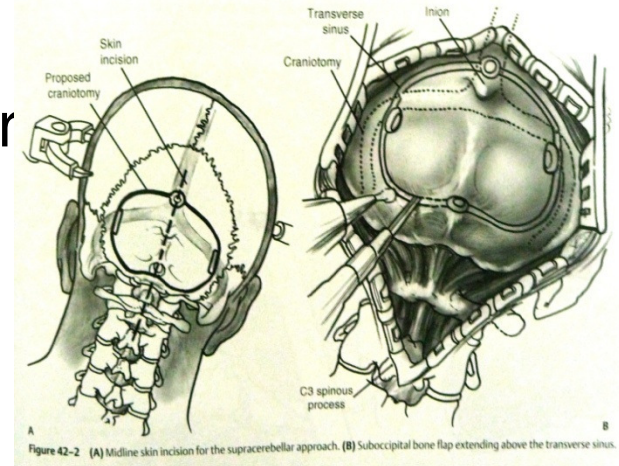


# INFRATENTORIAL SUPRACEREBELLAR

- COMPLICATIONS
  - Risks of sitting position
  - Limited upgaze & convergence
  - Ataxia
  - Cognitive impairment
- More frequent in patients having preop deficits, prev. radiation or invasive tumors.

# INFRATENTORIAL SUPRACEREBELLAR

- Position: Sitting ,  $\frac{3}{4}$  prone, concorde
- Head flexed: tentorium parallel to floor
- Midline incision : inion- C4
- Burr holes:
  - At sagittal sinus above torcula
  - At lateral aspect of transverse sinus b/l
  - Suboccipital Midline above for. magnum
- CSF can be drained if required from the trigone via EVD through lambdoid suture at midpupillary plane. Or later via opened cisterna magna.
- Craniotomy preferred over craniectomy as it reduce post operative aseptic meningitis, fluid collection, pain and discomfort.



# INFRATENTORIAL SUPRACEREBELLAR

- Durotomy: gentle curving incision
- Microscope with variable objective
- Bridging & precentral cerebellar veins cauterized & divided.
- Until arachnoid is opened & cerebellum freed from brainstem, the trajectory is to aimed at Vein of Galen to avoid injury to ICV/Rosenthal V. and then downward.
- Internal debulking/capsule dissection
- Most difficult & dangerous part : inferior portion of tumor adherent to dorsal midbrain.
- Copious irrigation to remove all clots and limited use of haemostatic agents as they can block aqueduct.

# LATERAL PARAMEDIAN INFRATENTORIAL

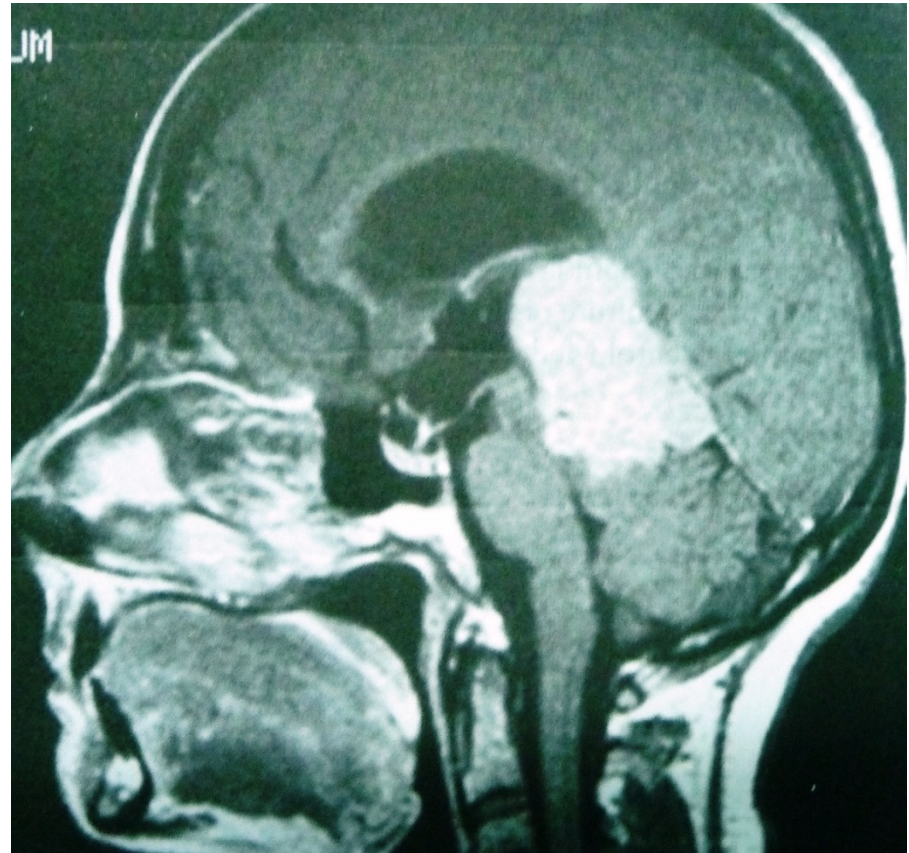
- **INDICATIONS**
  - Biopsy
  - Small quadrigeminal area tumor
- **ADVANTAGE**
  - Minimal damage to neural tissues
  - Useful in steep tentorium
  - Reduced risk of air embolism
- **DISADVANTAGES**
  - Narrow space
  - Difficult to reach tumor portion extending to inferoposterior part of 3<sup>rd</sup> ventricle

# LATERAL PARAMEDIAN INFRATENTORIAL

- POSITION
  - On the side: usually right side down
  - Upper part of trunk raised 30°
  - Head flexed with neck stretched & rotated 45° face down
- SURGICAL TECHNIQUE
  - S-shaped incision behind mastoid
  - Oval craniectomy close to sigmoid sinus laterally & transverse sinus superiorly
  - Durotomy : cruciate
  - Bridging veins divided, petrosal & precentral cerebellar veins preserved.
  - Tentorial incisura reached, preserving SCA.

# SUPRATENTORIAL APPROACH

- INDICATIONS
  1. Tumors extending superiorly
  2. Tumors involving or destroying the posterior aspect of corpus callosum
  3. Tumors deflecting the deep venous system dorsolaterally
  4. Tumors extending laterally to region of trigone
  5. Tumors displacing deep veins in ventral direction ( e.g.. Meningiomas)



# OCCIPITAL TRANSTENTORIAL

- Originally described by Horrax and later modified by Poppen
- **INDICATIONS**
  - Tumors straddling or lying above the tentorial notch
  - Vascular lesions : varices of vein of Galen, AVM, P3/4 PCA aneurysms.
- **ADVANTAGES**
  - Excellent view both above & below the notch
- **DISADVANTAGES**
  - Damage to occ. Lobe: visual field defects.
  - Damage to splenium
  - Difficult to access subtentorial C/L portion of tumor



# OCCIPITAL TRANSTENTORIAL

- Surgical technique
  - Position:  $\frac{3}{4}$  th prone position is preferred, with nondominant side down
  - Incision: U-shape rt. occipital flap that reflected inferiorly.
  - Craniotomy : 6 burr holes : 2 on left, 2 on right of sag. Sinus ,1 just rostral to trans. Sinus & 1 parietal.
  - Durotomy: T- shape & reflected along sinuses
  - Retractor on inferior surface of occipital lobe

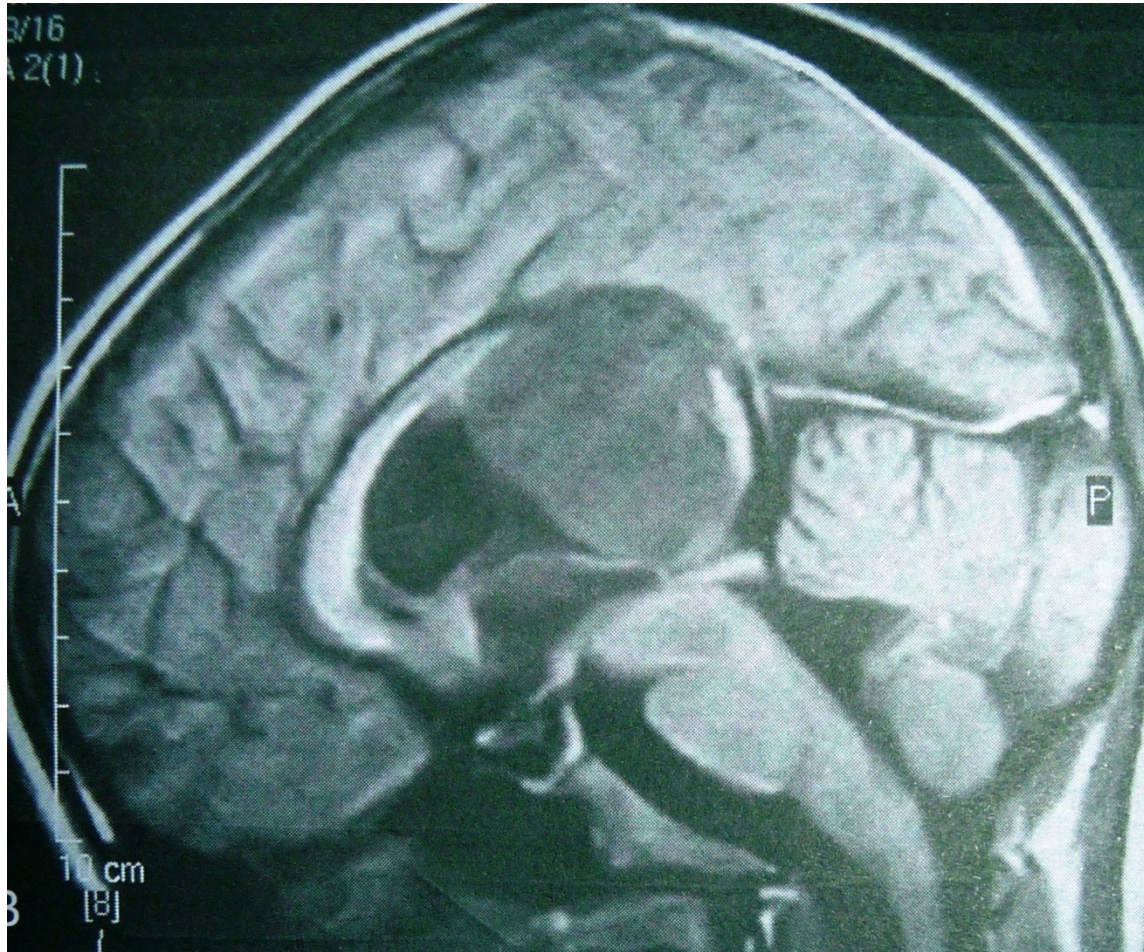
# OCCIPITAL TRANSTENTORIAL

- Falx retracted medially
- Tentorium cut 1-1.5 cm from & parallel to straight sinus.
- Quadrigeminal cistern opened , CSF drained
- Veins visualized : Galen vein – right Rosenthal—ICV—precentral cerebellar .

# OCCIPITAL TRANSTENTORIAL

- Cleavage plane found in small tumor
- Debulking in large tumor
- For hypervascular tumor: feeding arteries identified & coagulated prior to debulking .
- To avoid venous injury, total resection is not necessary & should not be attempted.
- Immaculate hemostasis, water-tight dura closure.

# Transcallosal inter-hemispheric



# Parietal transcaldosal inter-hemispheric

- 1<sup>st</sup> described by Dandy in 1921.
- Prone or sitting position
- Craniotomy centered over the vertex.
- U shaped flap across midline
- 2 burr hole ant and post over sigmoidal sinus
- Bone flap should extend 1-2 cm left of sss.
- Dura opened in U shape manner and reflected medially
- Bridging veins identified.
- Corpus callosum-white structure
- Approx. 2 cm divided.
- Tentorium/falx can be divided for exposure.
-

# Transcortical transventricular

- 1<sup>st</sup> developed by Van Wagenen
- Rarely used as exposures is limited
- Indication- for tumor that enter in the lateral ventricle

# TRANS -VELUM INTERPOSITUM

- **INDICATIONS**

- Huge tumors in pineal region/posterior 3<sup>rd</sup> ventricle
- Tumors extending anterior to massa intermedia

- **ADVANTAGES**

- Tumors extending into lateral ventricular can also be managed

- **DISADVANTAGES**

- Damage to anterior corpus callosum
- Damage to fornix

# TRANS -VELUM INTERPOSITUM

- Surgical technique
  - Position : Supine with head elevation 20` in pin
  - Coronal/Quadrangular skin flap on nondominant side
  - Quadrangular bone flap , extending to midline & anterior to coronal suture
  - Right frontal lobe retracted, corpus callosum exposed , split 3-4 cm to enter pars centralis



# TRANS -VELUM INTERPOSITUM

- Velum interpositum ( choroid plexus + tela choroidea + ICV ) cut just lateral to tenia fornices & medial to choroid plexus of lateral ventricle
- B/L fornices & ICV retracted medially to explore tumor b/w these structures & right thalamus.

# COMBINED SUPRA-INFRA-TENT. TRANS-SINUS

- Indications
  - Large tumors > 4.5 cm
  - Tumor arising from tentorium or extending above & below
  - Tumor well below plane of cerebellar retraction ( 2 cm below sup. Surface of cerebellum)
  - Very vascular tumors
  - Tumors encasing imp. Venous structures.

# Postoperative care

- High dose steroid should be maintained in first few days
- Lethargy and mild cognitive impairment is common( can also be due to extensive subdural air)
- Shunt malfunction is frequent immediate problem ( air, blood)- can be worrisome
- If EVD drain placed at at time of surgery should be removed at 72 hrs or convert to shunt.
- Post op MRI with contrast should be performed within 72 hrs.

# Complications of surgery

- Postoperative Hemorrhage
- Pupillary abnormality, accommodation abnormality, ocular palsies, upward gaze paresis, ataxia, impaired consciousness, shunt malfunction, ETV blockage.
- Cognitive impairment or akinetic mutism due to brainstem manipulation
- Sitting position: air embolism, hypotension, ventricular collapse, subdural hygroma / hematoma
- Supratentorial approach-: hemiplegia (retraction or compromise of bridging vein) / seizures
- Parietal lobe retraction: sensory/ stereognostic deficits.
- Occipital retraction : Visual field defects

# Outcome

- Overall mortality range from 0-8% and permanent morbidity from 0-12 %
- Impact of surgery depends on histology and response to adjuvant therapy
- Result of pineal region tumor at New York Neurological institute (1990-2008)()

Total procedures	128
Benign	55 (43%)
Malignant	73 (57%)
Diagnosis	127(99%)
Surgical morbidity	
Death	2 (2%)
Major morbidity Perm.	1 (1%)
Major morbidity temp.	7 (5%)

Data from Youmans neurological surgery ,4<sup>th</sup> edition

# Post op w/u

- Contrast MRI within 72 hr.
- Tumor markers
- Spine MRI
- CSF cytology
- Prophylactic spinal irradiation is not recommended

# Adjuvant therapy

## RADIOTHERAPY

- For malignant germ cell/pineal cell tumors:
  - 4000cGy : Ventricular system
  - 1500cGy : Tumor bed
- Total 550 Gy in 180 cGy daily fractions
- 3500 cGy to spine if spinal seeding radiographically documented.
- RT maybe withheld: for histologically benign pineocytoma /ependymoma completely resected.

# Adjuvant therapy

- Germinoma is most radiosensitive with 5 yr survival > 75 % and 10 yr survival of 69 % with RT dose 5000 Gy reported
- Germinoma with raised b- hCG has less favorable prognosis.
- Prophylactic spinal irradiation is avoided
- Side effects of RT
  - Cognitive deficits.
  - Hypothalamic/Endocrine dysfunction.
  - Cerebral necrosis.
  - De novo tumor formation.



# CHEMOTHERAPY

- Indications
  - Non germinomatous malignant germ cell tumors
  - Germinoma with syncytiotrophoblastic giant cells
  - Recurrent /disseminated pineal cell tumors
- Cisplatin/ carboplatin + Etoposide
- Others: vincristine/ lomustine/ cyclophosphamide

Delayed surgery after radiotherapy and chemotherapy is indicated for the patients with residual tumors whose tumor markers are normalized.

# Adjuvant therapy

## RADIOSURGERY-

- For tumors less than 3 cm
- For tumors that recur locally.
- Risk of recurrence : pineal cell/germ cell tx.
- No therapeutic coverage to ventricular system.
- MRI for target definition, SPGR (spoiled gradient recalled acquisition) sequences 1 mm slices

Thank you