PINEAL REGION TUMORS DIAGNOSIS & SURGICAL APPROACHES

ARTERIAL SUPPLY

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

ARTERIAL SUPPLY

- P3/P4
  - Medial occipital artery
    - Calcarine artery – calcarine sulcus
    - Parieto-occipital artery – parieto-occipital sulcus
  - Posterior pericallosal artery

- SCA
  - Inferior colliculi

VEINS OF PINEAL REGION

- Great cerebral vein of Galen
  - Pineal veins(sup & inf) -- pineal body, trigonum habenulae
  - Quadrigeminal veins(sup&inf)—corpora quadrigemina
  - Superior vermian vein – superior vermis
  - Precentral cerebellar vein– cerebellum, superior cerebellar peduncle
  - Posterior pericallosal vein
  - Internal occipital vein

VEINS OF PINEAL REGION

- Internal cerebral veins
  - Septal veins(ant&post) –septum pellluvidum
  - Anterior caudate vein – caudate nucleus
  - Thalamostriate(terminal) vein
  - Choroidal vein –choroid plexus lat. Ventricle
  - Medial atrial (trigonal)vein – Trigone
  - Direct lateral veins

VEINS OF PINEAL REGION

- Basal vein of Rosenthal
  - Vena cerebri media profunda
  - Venae centrales (striate) inferiores
  - Vena cerebri anterior
  - Vena apicis cornua temporalis (hippocampal vein)
  - Vena atrii lateralis (lateral atrial vein)
  - Vena cornus temporalis (atriotemporal vein)

DIFFERENTIAL DIAGNOSIS

- GERM CELL TUMORS (account for 50%)
  - Pure germinoma
  - Embryonal cell carcinoma
  - Endodermal sinus tumor
  - Teratoma
  - Mixed germ cell tumor
  - Choriocarcinoma

DIFFERENTIAL DIAGNOSIS

- PINEAL PARENCHYMAL TUMORS
  - Pineoblastoma
  - Pineocytoma
- TUMORS OF SUPPORTIVE TISSUES & ADJ. STR.
  - Astrocytoma
  - Ependymoma
  - Meningioma
  - Hemangiopericytoma

DIFFERENTIAL DIAGNOSIS

  - Ganglioglioma
  - Ganglioneuroma
  - Chemodectoma
  - Quadrigeminal cistern lipoma
- Metastatic tumors (very rare)
  - Lung, breast, stomach, kidney
- Nonneoplastic lesions
  - Pineal cysts

DIFFERENTIAL DIAGNOSIS

- Arachnoid cysts
- Cysticercous cysts
- Vascular lesions
  - Aneurysmal dilatation of vein of Galen

COMMON PATHOLOGIES

- Germ cell tumors
  - Germinoma
  - Teratoma
- Pineal cell tumors
  - Pineoblastoma
  - Pineocytoma
- Other cell tumors & tumor like masses
  - Pineal cysts

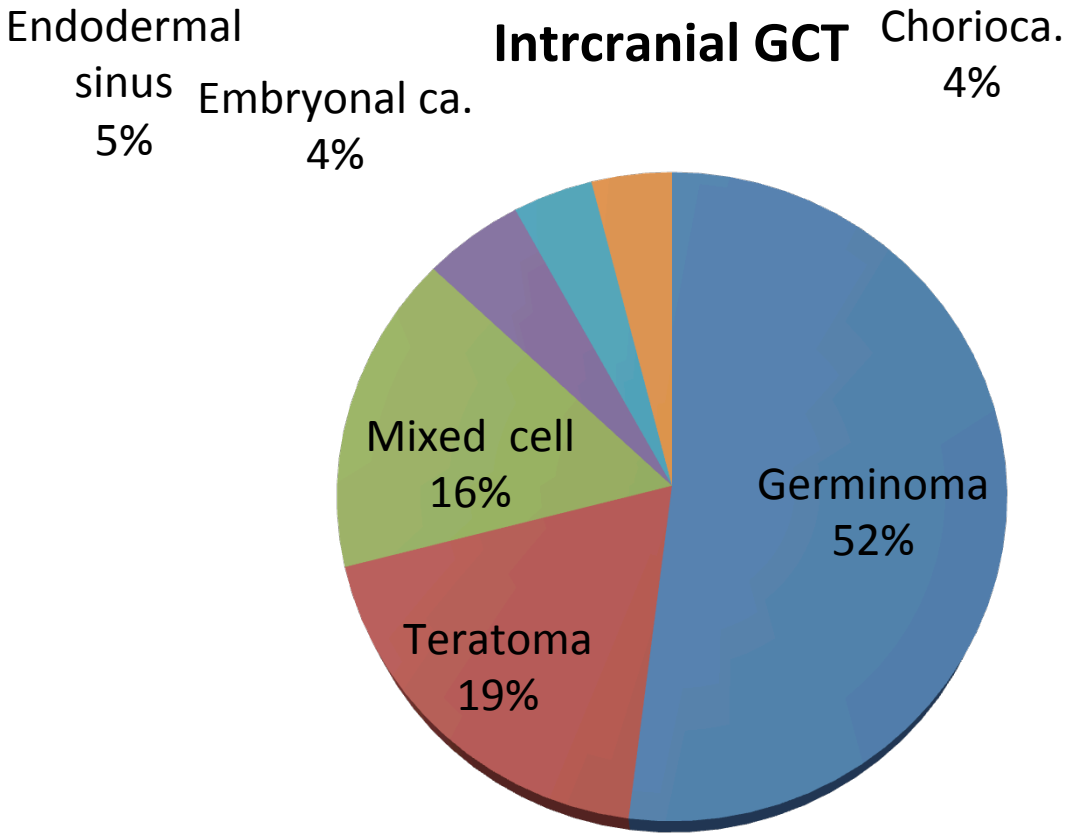
COMMON PATHOLOGIES

  - Astrocytoma (thalamus, midbrain, tectum, corpus callosum)
  - Meningioma
  - Metastases
  - Vascular malformation (Vein of Galen)
- Misc.
  - Lipoma
  - Epidermoid
  - Arachnoid cyst

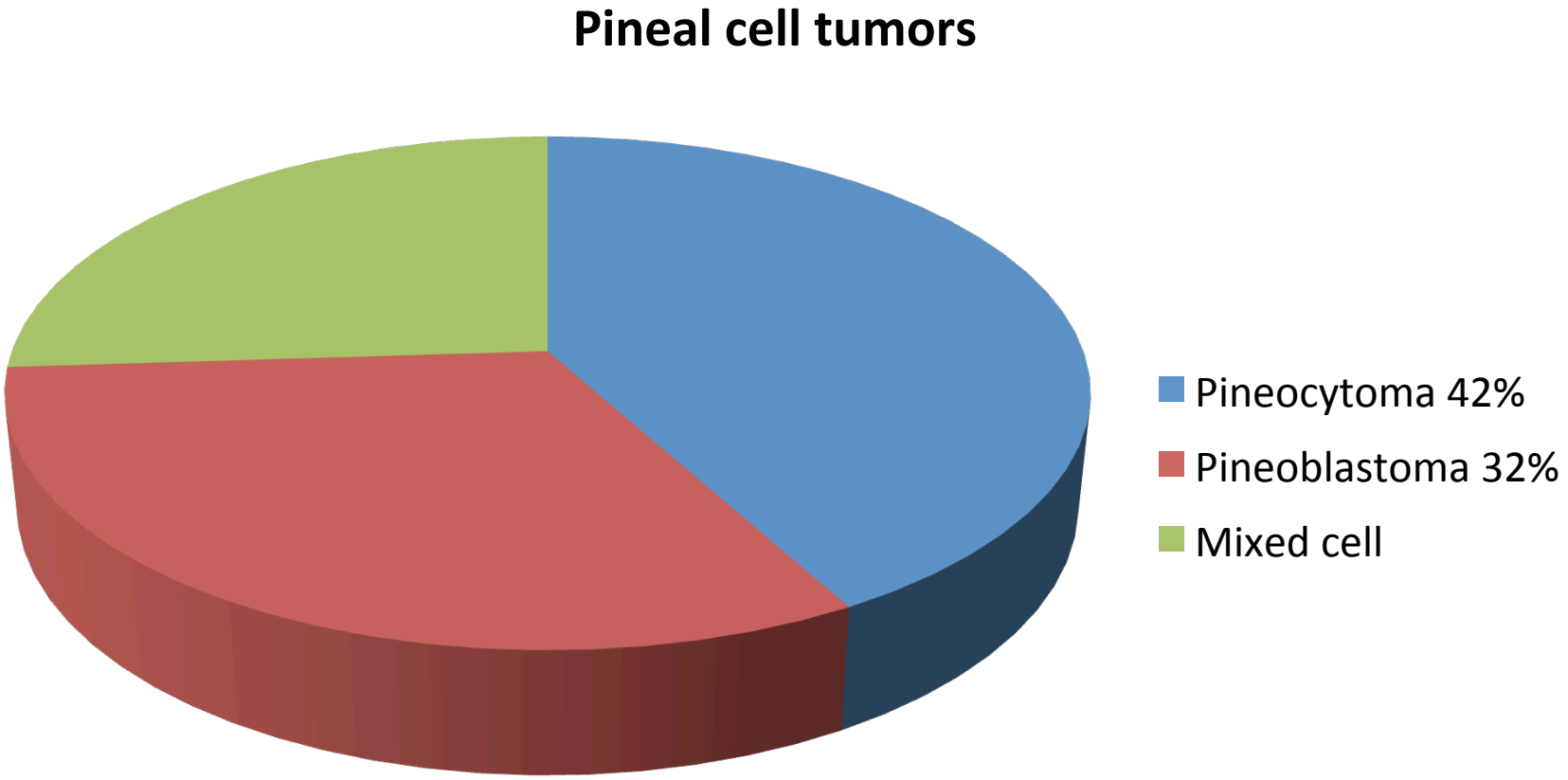
EPIDEMIOLOGY

- Incidence : <1% of CNS tumors
- Germ cell tumors: more in pediatric pop., M>F
- Pineal cell tumors : more in young adults,M=F
- Heritability: rare : E.g. Trilateral retinoblastoma[Pinealoblastoma + b/l retinoblastoma, gene deletions]

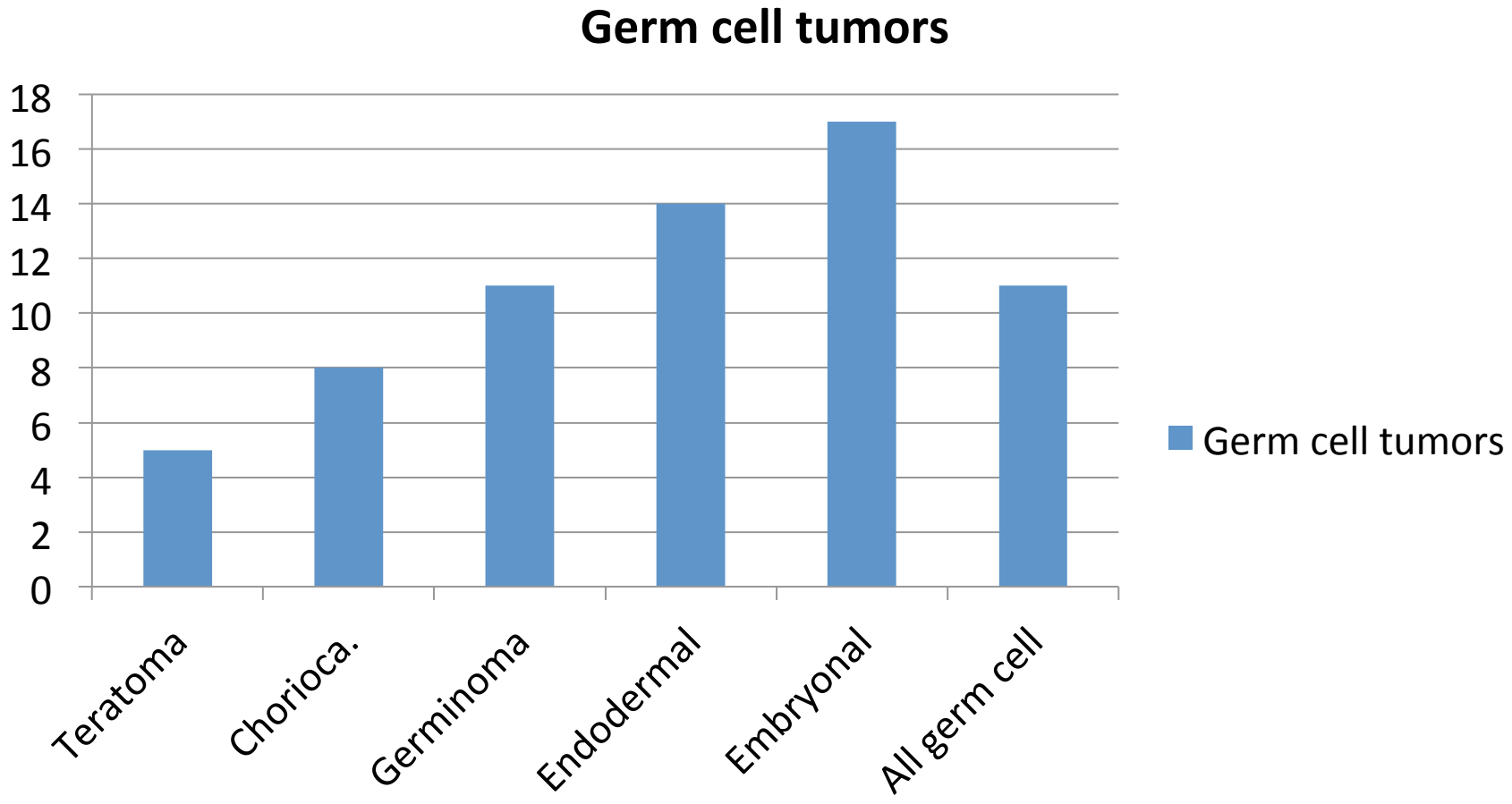
Relative % of histological subtypes of I/C GCT based on 399 tumors reviewed by Jennings et al



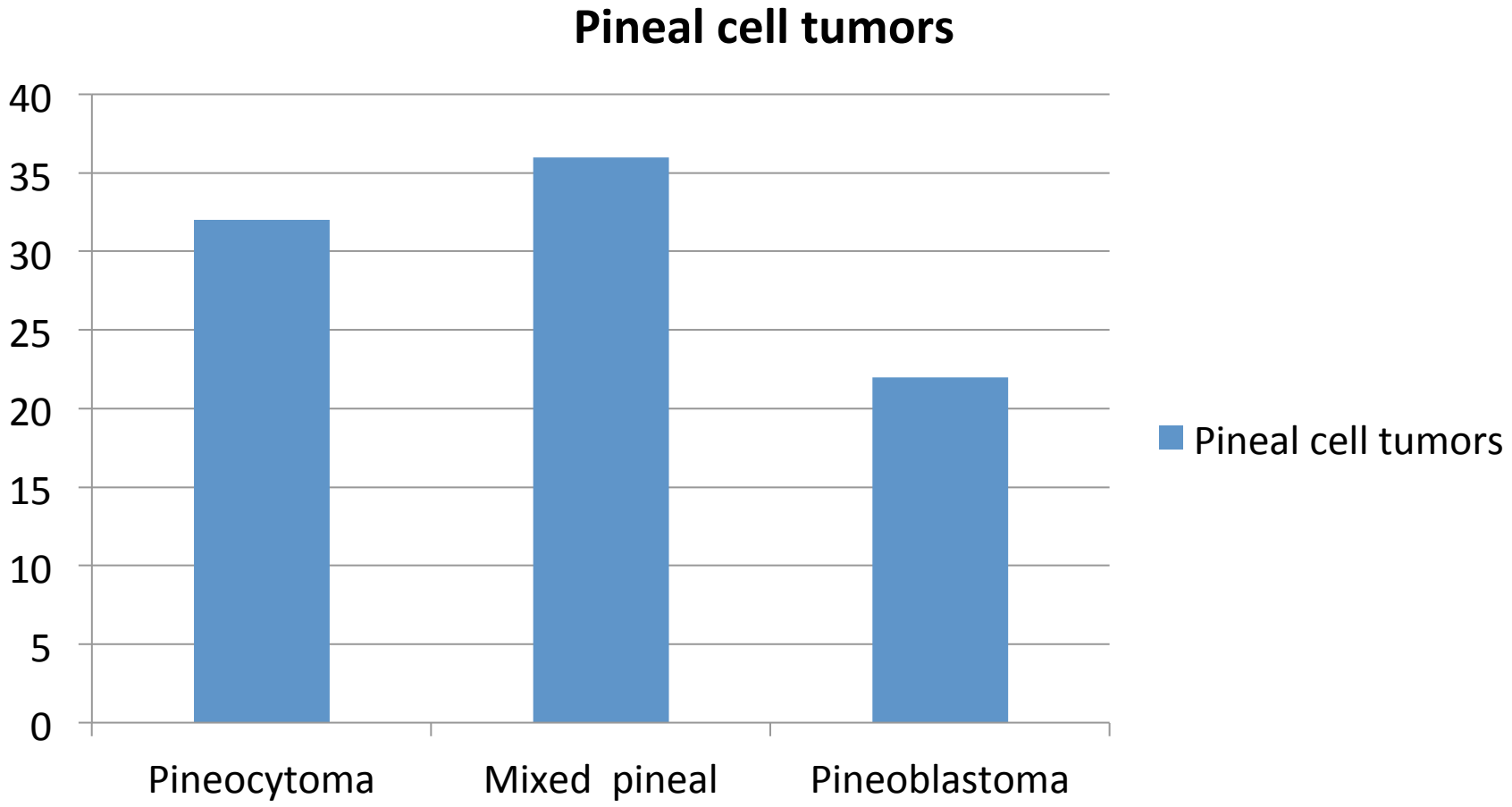
Relative % of histological subtypes of pineal cell tumors based on 90 tumors reviewed by Herrick & Rubinstein et al



Most common age of onset of I/C GCT reviewed by Jennings et al



Average age of onset for pineal cell tumors based on 90 tumors reviewed by Herrick & Rubinstein et al



CLINICAL FEATURES

- Mechanism
  - Raised ICP [Hydrocephalus]
  - Direct cerebellar/brainstem compression
  - Endocrine dysfunction
- Presentation
  - HEADACHE (most common) – Aqueductal obstruction – HCP– N/V, Papilledema, Obtundation, Cognitive deficits.

CLINICAL FEATURES

- EXTRAOCULAR PALSIES
  - PERINAUD SYNDROME
    - Paralysis of up gaze/convergence
    - Retractory nystagmus
    - Light-near pupillary dissociation
  - 4^{th} CN PALSY
    - Diplopia

CLINICAL FEATURES

- Compression/infiltration of dorsal midbrain
  - Paralysis of down gaze
  - Ptosis
- Inv. Of superior cerebellar peduncle
  - Ataxia/dysmetria
- Inferior colliculi compression
  - Hearing disturbances

CLINICAL FEATURES

- Endocrine dysfunction: hypothalamic invasion/HCP
  - Diabetes Insipidus
  - Precocious puberty[boys] : b- h CG secrn. In chorioca./Germinoma with STGC – androgen secretion by Leydig cells
- Pineal apoplexy
  - In vascular tumors : Pineal cell tumors/Chorioca.

CSF Levels of tumor markers in germ cell tumors

	Germinoma	Immature teratoma	Embryonal cell ca	Mature teratoma	Chorioca.	Endodermal sinus tumor
Alpha fetoprotein	--	--/+	+	--	--	+++
Beta-HCG	--/+	--	++	--	+++	--
PLAP	++	--	--	--	--	--

Other markers

- C-kit proto-oncogene
- CEA
- LDH
- B5 monoclonal antibody
- S antigen, melatonin

IMAGING

- CT/MRI/ANGIOGRAPHY
  - Size of tumor: lateral & superior extent
  - Vascularity
  - Homogenous/heterogeneous
  - Irregularities of margination & probability of invasion

IMAGING

- ANATOMICAL relationships
  - Inv of 3rd ventr/ position within 3rd ventr.
  - Extension into/above corpus callosum
  - Superolateral extension into ventricular trigone
  - Inv/compression of quadrigeminal cistern/aqueduct
  - Relation to ant. Cerebellar vermis
  - Location of deep venous system

ADJUVANT THERAPY

- RADIOTHERAPY
  - For malignant germ cell/pineal cell tumors: 4000cGy : Ventricular system

    1500cGy : Tumor bed

- 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 with raised b- hCG has less favorable prognosis.
- Side effects of RT
  - Cognitive deficits.
  - Hypothalamic/Endocrine dysfunction.
  - Cerebral necrosis.
  - De novo tumor formation.

ADJUVANT THERAPY

- 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

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

RADIOSURGERY

- Most patients tolerate 15 Gy to brainstem surface at the edge of the lesion.
- Dose limitation not required because of proximity of deep veins/ corpus callosum.
- Follow up radiology : Germinoma (1 month), for benign histologies (3-6 months).

SURGICAL ANATOMY

- Most tumors arise from or attached to the undersurface of velum interpositum.
- Tumors rarely extend above velum.
- Blood supply comes from within velum mainly from M P.ch & L P.ch with anastomoses to pericallosal & quadrigeminal artery.
- 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 (Angioblastic meningioma)

MANAGEMENT OF HYDROCEPHALUS

- If complete tumor removal is anticipated: EVD at surgery: removal on POD:2/3
- Conversion to VP shunt
- ETV preferred :
  - Gradual reduction of ICP
  - Eliminates potential complications
    - Infection
    - Overshunting
    - Peritoneal seeding by malignant cells

OPERATIVE: BIOPSY

- For adjuvant therapy
- Metastatic workup
- Prognosis & long term follow up.
- OPEN BIOPSY
  - More extensive tissue sampling in view of heterogeneity & mixed cell population
  - Debulking: favorable response to adjuvant therapy
  - Risk of postop. bleeding in incompletely resected tumor bed.

OPERATIVE: BIOPSY

- STEREOTACTIC BIOPSY
  - Suited for patients with multiple lesions, clinical conditions that contraindicate open surgery/ general anaesthesia.
  - Multiple specimens to be obtained.
  - Side cutting cannula preferred over cup forceps
  - Hemorrhage: Continuous suction & irrigation x 15 min.

STEREOTACTIC APPROACHES

- INDICATIONS
  - For biopsy to achieve diagnosis
  - Aspiration of cystic masses
  - Radiosurgery for treatment : meningioma, pineocytoma, AVM
  - As adjuvant management with chemotherapy for germ cell neoplasms & pinealoblastomas.

STEREOTACTIC APPROACHES

- ADVANTAGES
  - Resection may not be necessary depending on the histopathology
  - Biopsy may guide effective non-surgical therapies
  - Radiosurgery for small volume pathologies

- DISADVANTAGES
  - Small biopsy volume : Difficulty in diagnosis
  - Experienced neuropathology team required
  - Risk of radiation related damage & deficits

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.
- POSEROLATEROSUPERIOR APPROACH:
  - Entry point near PO junction, for tumors extending laterally or superiorly.

ENDOSCOPY

- BIOPSY : In conjunction with ETV
- FLEXIBLE ENDOSCOPE : Limited trajectory to tumor through foramen magnum
- RIGID ENDOSCOPE : Low frontal burr hole
- LIMITATIONS
    - Limited tissue sampling
    - Difficulty in hemostasis

SURGICAL APPROACHES

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

SURGICAL APPROACHES

- HISTORICAL PERSPECTIVE
  - Transvelum interpositum [Sano]
  - Lateral paramedian infratentorial [Van den Bergh, 1990]
  - Other approaches [TRANSCALLOSAL : transforaminal, interfornicial,

    subchoroidal, transchoroidal] – primarily for ant./ mid 3rd ventricle tumors.

POSITIONING

- SITTING
- LATERAL : Non dominant hemisphere down for supratentorial approaches
  - Standard lateral : Nose rotated 30` towards floor
  - ¾ Prone : As above + head extended + rotated 45` towards floor.
- PRONE: Steep angle of tentorium makes it impractical for infratentorial approaches.
  - Concorde : Head rotated 15` away to facilitate occipital lobe retraction(desirable for patients < 3 yrs with excessively large ventr. with increased risk of collapse).

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 enroute
  - Exposure comparable with that of other routes

INFRATENTORIAL SUPRACEREBELLAR

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

SURGICAL TECHNIQUE

- Position: Sitting , ¾ prone, concorde
- Head flexed: tentorium parallel to floor
- Midline incision : inion- C3
- Burr holes:
  - At sagittal sinus above torcula
  - At lateral aspect of transverse sinus b/l
  - Suboccipital Midline above for. magnum

SURGICAL TECHNIQUE

- Durotomy: gentle curving incision
- Microscope with variable objective/275mm
- 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.

SURGICAL TECHNIQUE

- Internal debulking/capsule dissection
- Most difficult & dangerous part : inferior portion of tumor adherent to dorsal midbrain.
- Copious irrigation to remove all clots that 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 3rd 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

LATERAL PARAMEDIAN INFRATENT

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

OCCIPITAL TRANSTENTORIAL

- 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: semiprone with nondominant side down
  - Incision: U-shape
  - 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
- Ventricular drain placed in occipital horn
- 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.

TRANS -VELUM INTERPOSITUM

- INDICATIONS
  - Huge tumors in pineal region/posterior 3rd ventricle
  - Tumors extending anterior to adhesio interthalamica
- 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 fornicis & medial to choroid plexus of lateral ventricle
- B/L fornices & IJV retracted medially to explore tumor b/w these structures & right thalamus.

COMBINED SUPRA-INFRATENT. TRANSSINUS

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

COMBINED SUPRA-INFRATENT. TRANSSINUS

- Cerebral angiogram is mandatory to look for venous anatomy, size & communication b/w transverse sinus & deep venous system.
- VP Shunt 2-4 weeks prior to planned surgery
- Somatosensory evoked potentials from UL & LL B/L & BAER monitoring during surgery
- Position : Semiprone with proposed trans. Sinus section placed inferiorly.

COMBINED SUPRA-INFRATENT. TRANSSINUS

- U-shape incision
- Burr holes inferior to trans. Sinus (4) & just above for. Magnum (2) – suboccipital craniotomy
- Trans. Sinus separated & occipital craniotomy performed on one side followed by other.
- Suboccipital dura opened in transverse fashion inferior to trans. Sinus

COMBINED SUPRA-INFRATENT. TRANSSINUS

- Occipital dura is then opened parallel to sinuses
- 20 G butterfly needle inserted in trans. Sinus just lateral to torcula & medial to temporary clip placed for test occlusion x 5 min.
- Nondominant trans. Sinus can be safely sectioned if : venous pressure > 5 mm Hg, no brain swelling, no change in evoked potentials.
- Tentorium is then cut parallel to straight sinus.

COMBINED SUPRA-INFRATENT. TRANSSINUS

- Deep vein injury : repair with 7-0 prolene/8-0 nylon
- Trans. Sinus can be reconstructed with a short vein graft interposed with 6-0 prolene.
- Not necessary to suture tentorium
- Dural graft to allow watertight closure & expansion of posterior fossa.
- If brain swelling + : suboccipital bone kept out.

COMBINED SUPRA-INFRATENT. TRANSSINUS

- Postoperative Care
  - Look for respiratory abn in tumors compressing brainstem
  - CT on POD1 to l/f pneumocephalus/clots
  - Nystagmus/ataxia/oscillopsia
  - Ventricular drain x 3-4 days – 20 cm above head x 24 hrs– test clamping x 24 hrs with ICP monitoring —CT to l/f ventricle size.
  - Vision abnormalities

COMPLICATIONS OF SURGERY

- Postop. Hemorrhage/apoplexy
- Pupillary abn., accomodation abn., ocular palsies, upward gaze paresis, ataxia, impaired consciousness, shunt malfunction, ETV blockage.
- Sitting position: air embolism, hypotension, cortical collapse, subdural hygroma
- Parietal approaches: sensory/stereognostic deficits.
- Occipital : Visual field defects

THANK YOU !!