Children's Hospital Boston

Endoscopy for pineal tumors

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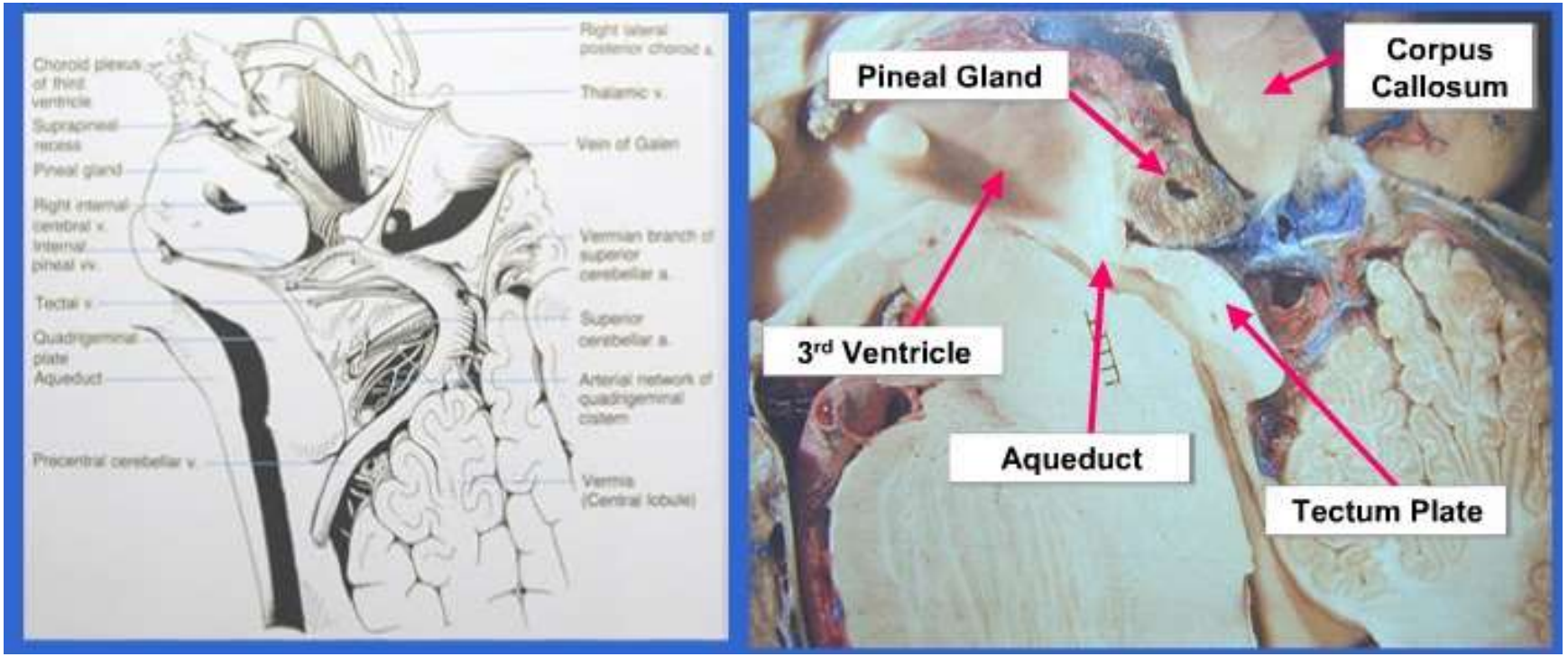
Advances in the Management of Pineal Area Tumors

- **Surgical**
  - **endoscopy**
- **Biology**
  - **markers**
- **Adjuvant therapy**

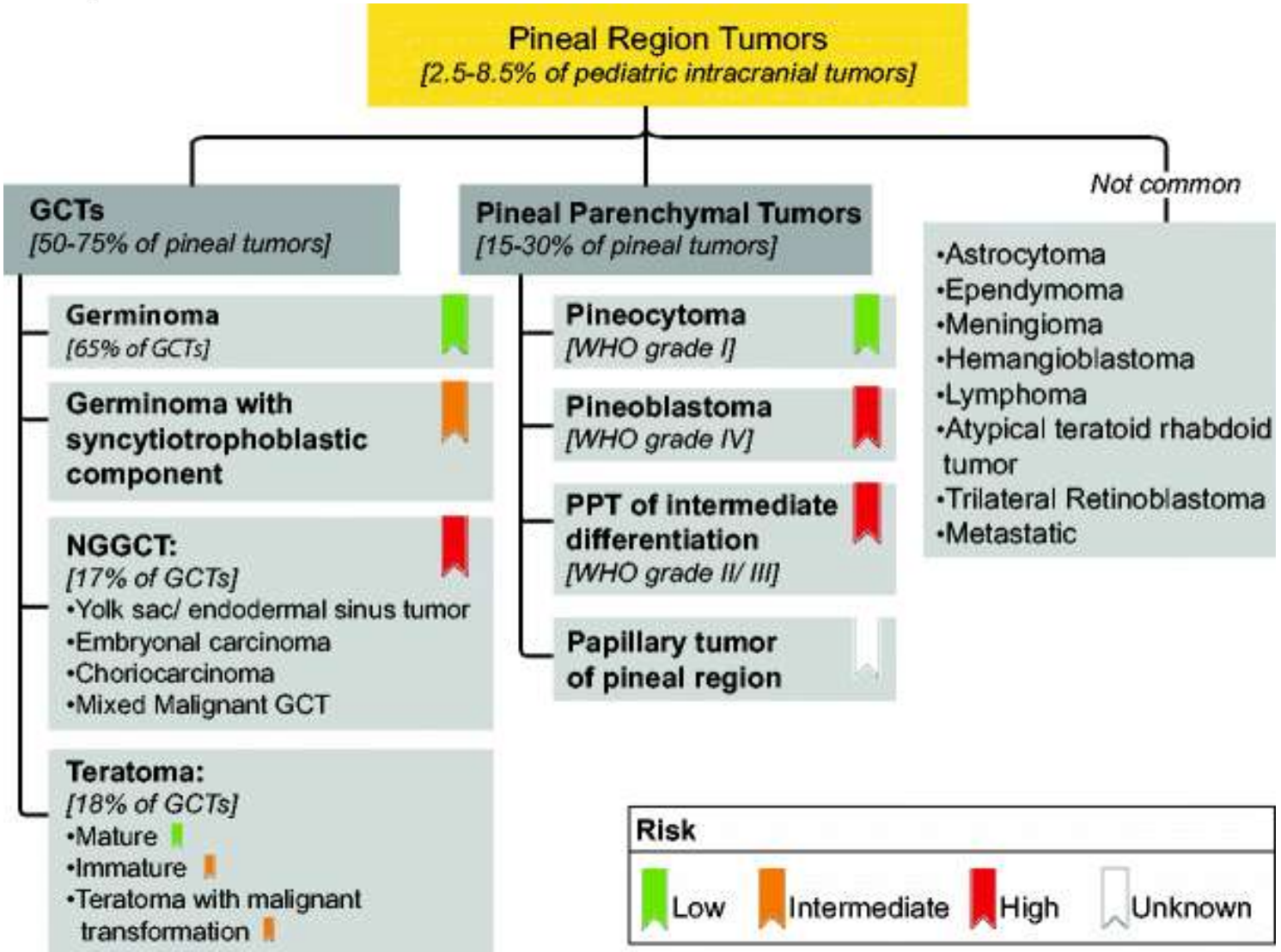
Pineal tumors - anatomy



Pineal tumors - pathology

- **Adult Population**
  - Pineal Parenchymal Tumors
    - **Pineocytoma**
    - Pineoblastoma
    - Intermediate
  - Germ Cell Tumors
    - GGCT (Germinoma)
    - NGGCT (Mixed Germ Cell Tumor)
  - Glial Cell Tumors
    - **Glioblastoma multiforme**
    - **Ependymoma**
  - Cysts
    - III ventricular
    - Pineal gland
  - Vascular
    - AVM
    - Cavernous malformation
    - Vein of Galen
  - Miscellaneous
    - **Meningioma**
    - Hemangioblastoma
    - Metastatic
    - Lymphoma

- **Pediatric Population**
  - **Germ Cell Tumors**
    - **GGCT (Germinoma)**
    - **NGGCT (Mixed Germ Cell Tumor)**
    - **Mature teratoma**
  - Pineal Parenchymal Tumors
    - Pineocytoma
    - **Pineoblastoma**
    - **Intermediate**
  - Glial Cell Tumors
    - **Astrocytoma (fibrillary, pilocytic)**
    - Glioblastoma multiforme
    - **Ependymoma**
  - Cysts
    - III ventricular
    - Pineal gland
  - Vascular
    - AVM
    - Cavernous malformation
    - **Vein of Galen**
  - Miscellaneous
    - Meningioma
    - Hemangioblastoma
    - Metastatic
    - Lymphoma
    - **Choroid Plexus Tumor**
    - **Triretinal retinoblastoma**


Pineal Region Tumors
[2.5-8.5% of pediatric intracranial tumors]
GCTs
[50-75% of pineal tumors]
Germinoma
[65% of GCTs]
Germinoma with syncytiotrophoblastic component
NGGCT:
[17% of GCTs]
•Yolk sac/ endodermal sinus tumor
•Embryonal carcinoma
•Choriocarcinoma
•Mixed Malignant GCT
Teratoma:
[18% of GCTs]
•Mature
•Immature
•Teratoma with malignant transformation
Pineal Parenchymal Tumors
[15-30% of pineal tumors]
Pineocytoma
[WHO grade I]
Pineoblastoma
[WHO grade IV]
PPT of intermediate differentiation
[WHO grade II/ III]
Papillary tumor of pineal region
Not common
•Astrocytoma
•Ependymoma
•Meningioma
•Hemangioblastoma
•Lymphoma
•Atypical teratoid rhabdoid tumor
•Trilateral Retinoblastoma
•Metastatic
Risk
Low
Intermediate
High
Unknown

Pineal Tumors - Clinical

- 0.4%-1% of all intracranial tumors
- 3-8% in the pediatric population (Japan 4-6%)
- Germ cell tumors 0.4-3.4% of intracranial tumors (0.1 per 100 000 persons per year)
  - Germinomas 35-65% with 4:1 male:female ratio
  - Most within the first 3 decades
  - NGGCT males in the first 2 decades
- **Hydrocephalus**
  - Headache, nausea, vomiting
  - Double vision
  - Papilledema
- Parinaud's syndrome (upward gaze palsy with impaired convergence)
  - Poor response to light/accommodation
- DI, endocrinopathy and visual disturbance
  - Synchronous germinomas in the pineal and suprasellar region
  - Precocious puberty

Goals of Surgery for Pineal Area Tumors

- Establish Pathologic Diagnosis
- Manage hydrocephalus
- Resection of tumor

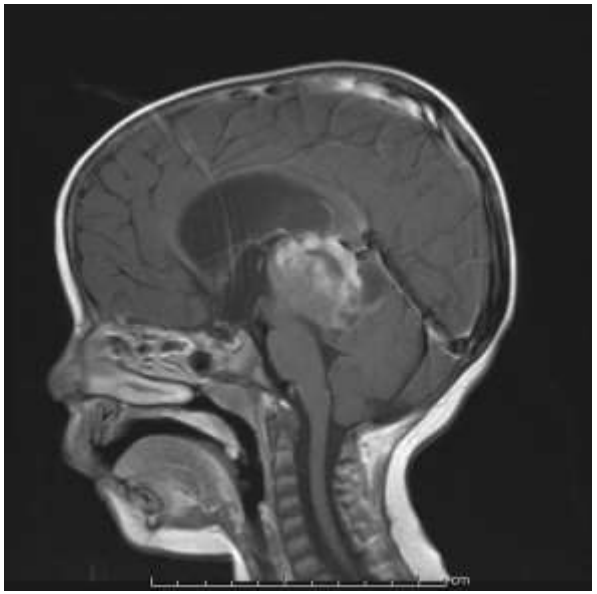
- Is a tissue sample always needed?
  - **No**
- VP shunt insertion vs. ETV
  - **ETV**
- Is resection always indicated?
  - **No**
- Morbidity of above procedures

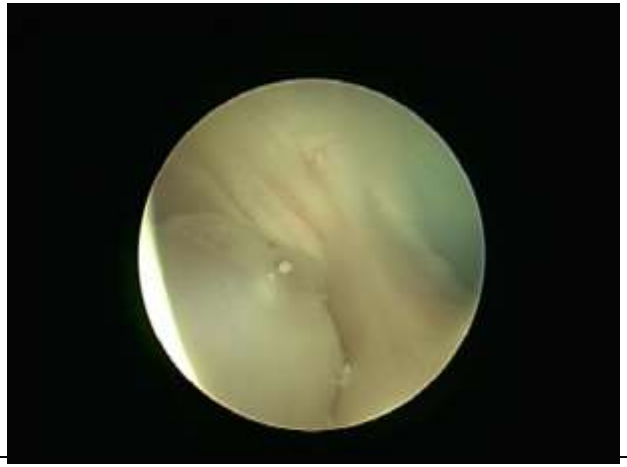
Pineal area germ cell tumors

- Serum and CSF markers can distinguish between germinomatous and NGGCT tumors
- Chemotherapy can be initiated after biologic diagnosis and obviates the need for tumor surgery at time of presentation
- Markers are used postoperatively to monitor response to chemotherapy and relapse of tumor/initiation of subsequent therapy

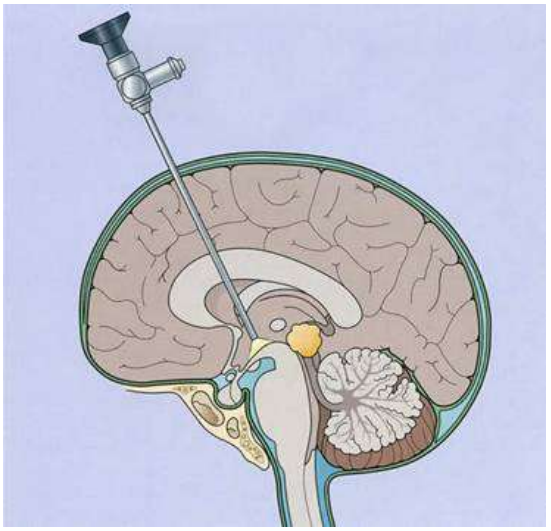
Pineal Tumors

- Diagnosis
  - MRI Brain and spine
  - Serum and CSF Markers
- Endoscopy
  - Treat hydrocephalus
  - Endoscopic biopsy
  - Endoscopic resection
- Surgery





Pineal Tumors – Initial Evaluation and Treatment



- History/physical examination
- Imaging MRI Brain and Spine
- BrainLab MRI
- Serum and CSF Markers
- Treatment of Hydrocephalus
  - EVD +/- Role
  - Shunt (No, dissemination)
  - Endoscopy
    - ETV Alone
    - ETV and Biopsy of Tumor

Pineal tumors – endoscopic biopsy

- Anatomical considerations
  - Location of burr holes
  - Location of tumor
  - Size of lateral ventricles
  - Size of third ventricle
  - Size of foramen of Monro
  - Relationship of tumor to anatomic structures
  - Ependymal lining/tumor interface

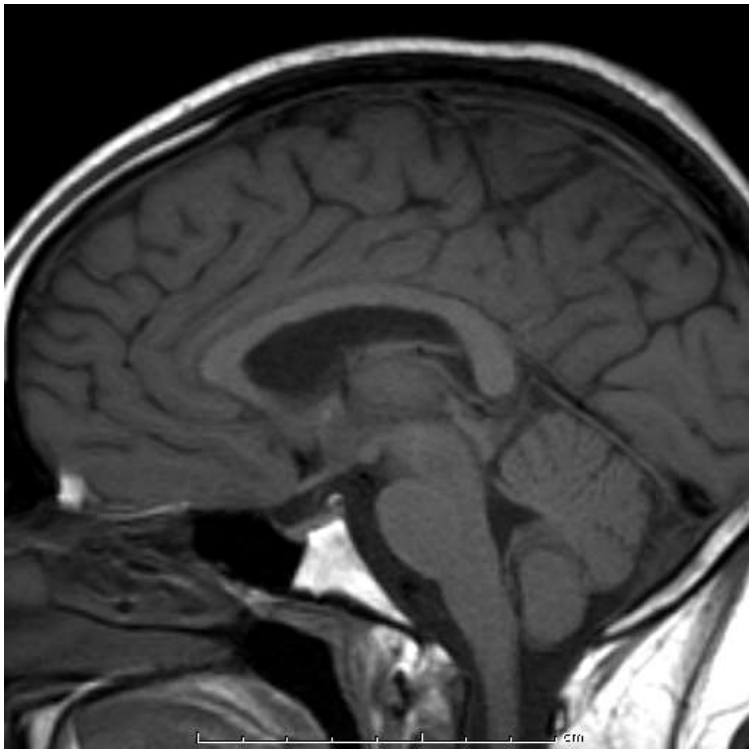
Pineal Tumors: Endoscopic biopsy

- Endoscope systems
  - **Rigid**: Gaab / Storz, Aesculap
  - **Flexible**: Storz or Codman
  - 0 and 30 degree angled scope
  - Instruments
    - Biopsy forceps
    - Cautery
    - Holding device
- Position of burr hole
- Pathology

19 year old male with headaches, blurry vision and vomiting

ETV, Endoscopic biopsy - germinoma

RT, normal, college graduate, no disease

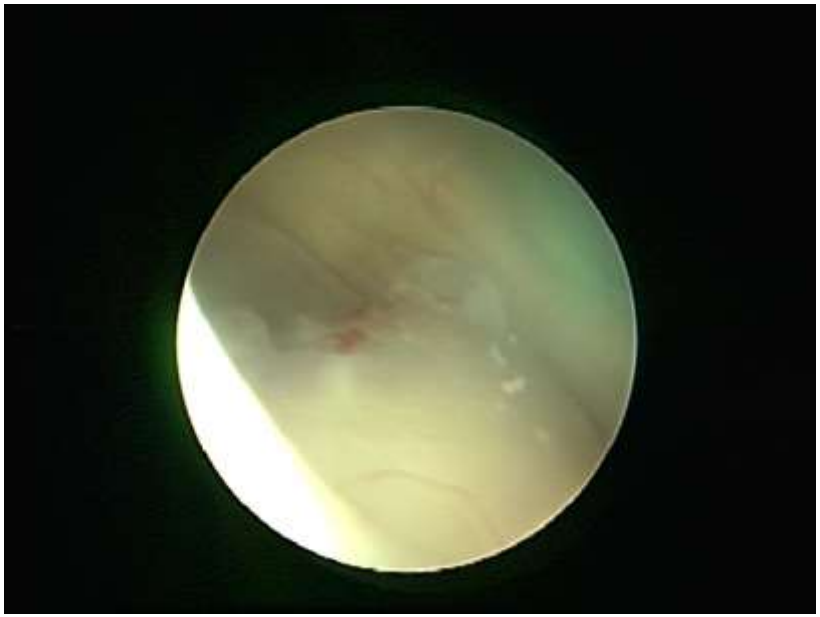


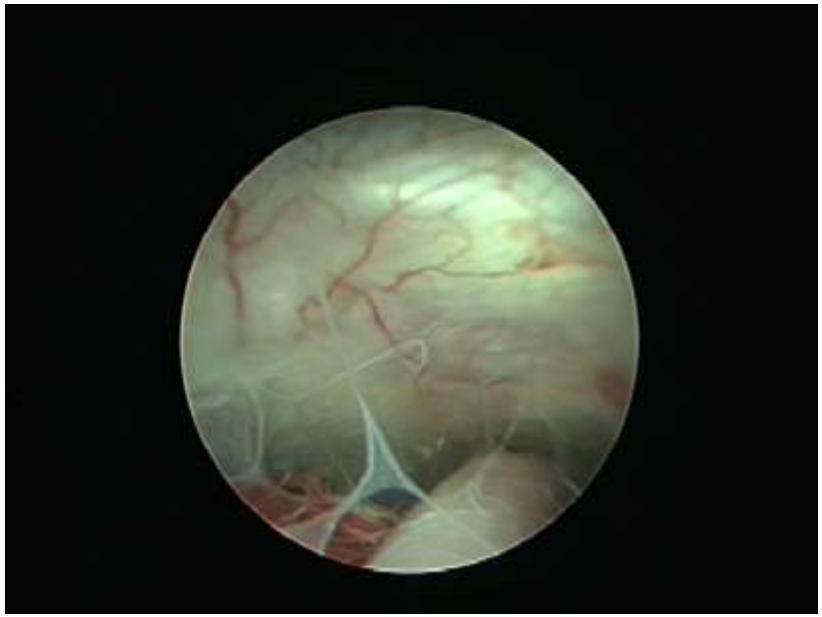
Endoscopy in diagnosis

- 18 year old male presents with headaches
- Serum markers non-contributory
- Papilledema

Endoscopy reveals irregularity of floor of III ventricle

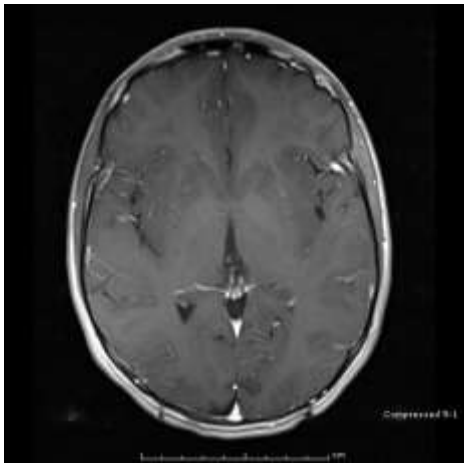
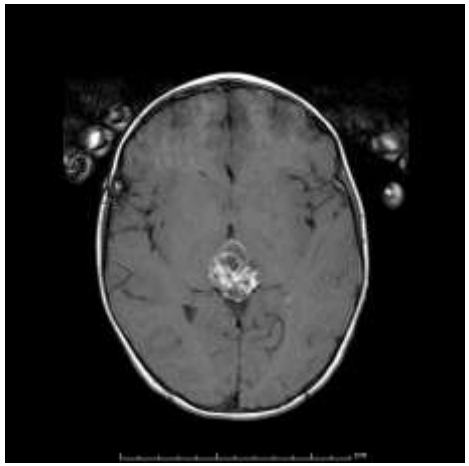
10 year old with hydrocephalus
ETV and biopsy (LGG)

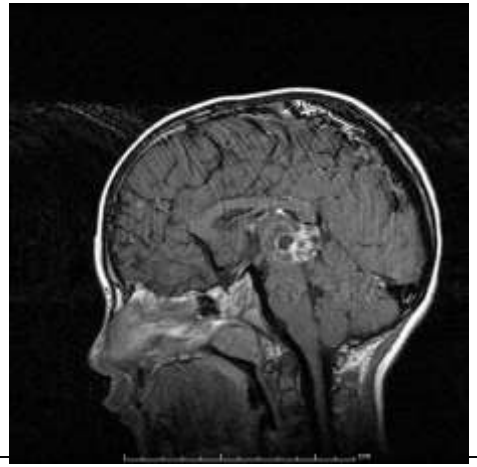




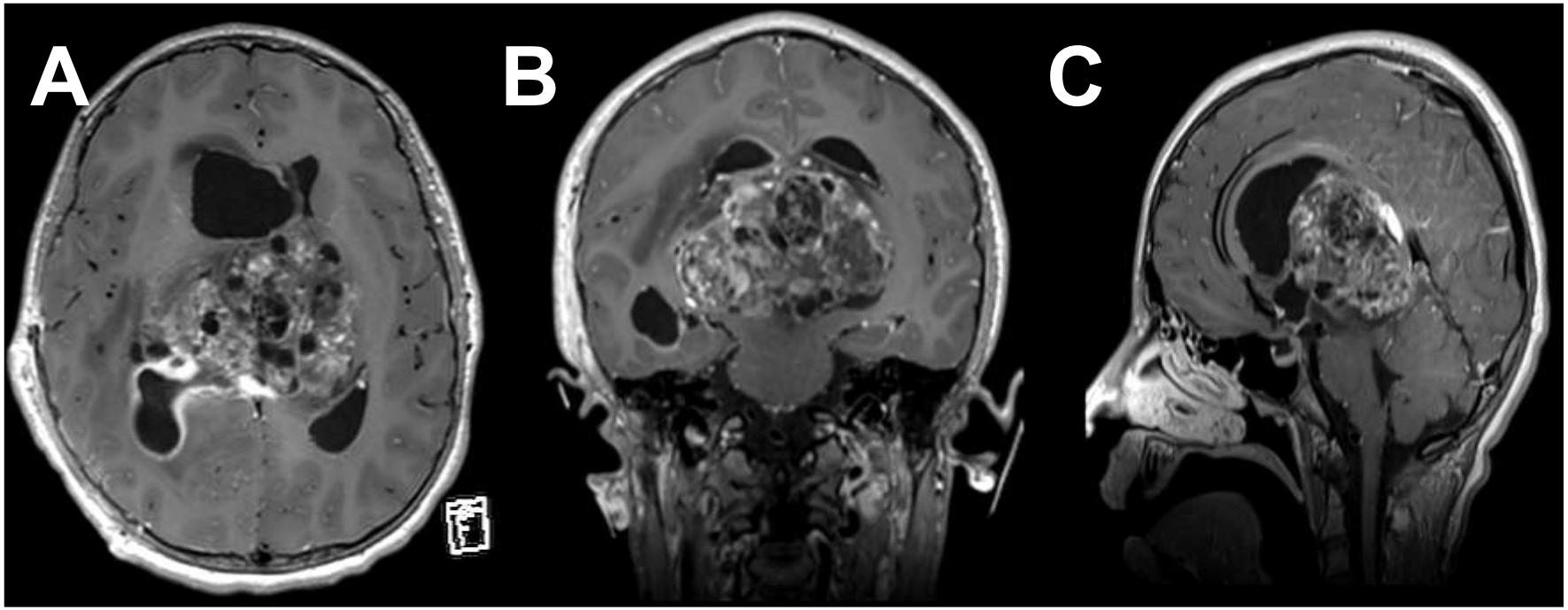
7 year old with hydrocephalus
Dermoid/mature teratoma with CSF dissemination

16 year old male who presented at age 2 with hydrocephalus, ETV, 6 months later presented with teratoma, completely resected.
Recurrence in 2 years, complete resection followed by focal RT.

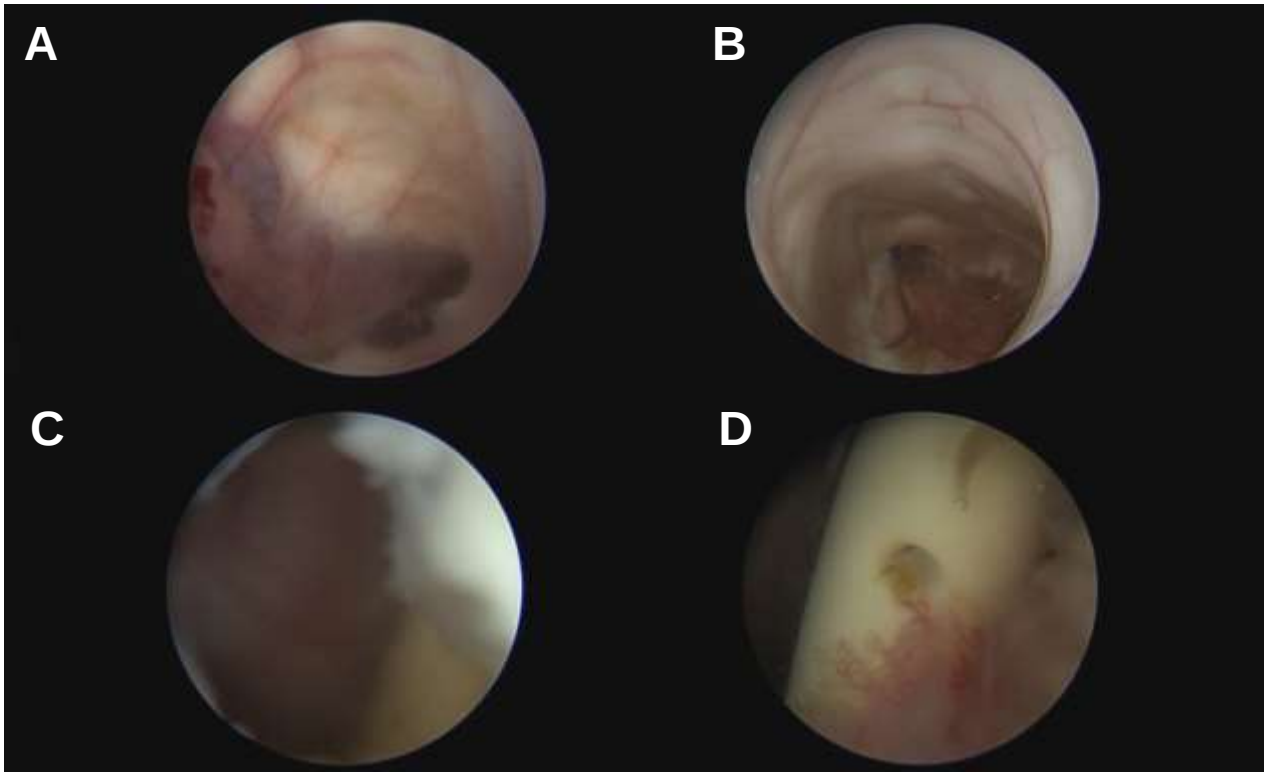





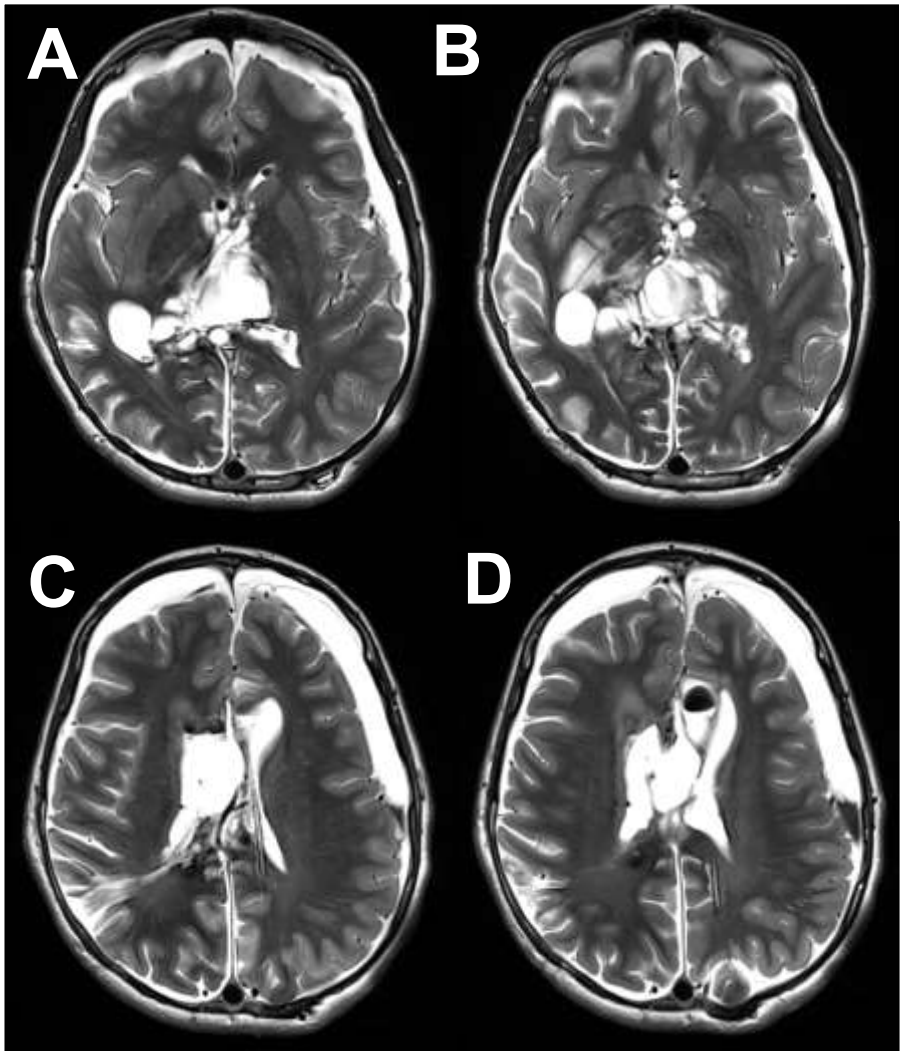
9 year old male with NGGCT, biopsy at outside institution showed teratoma with some immature elements, growing on chemotherapy

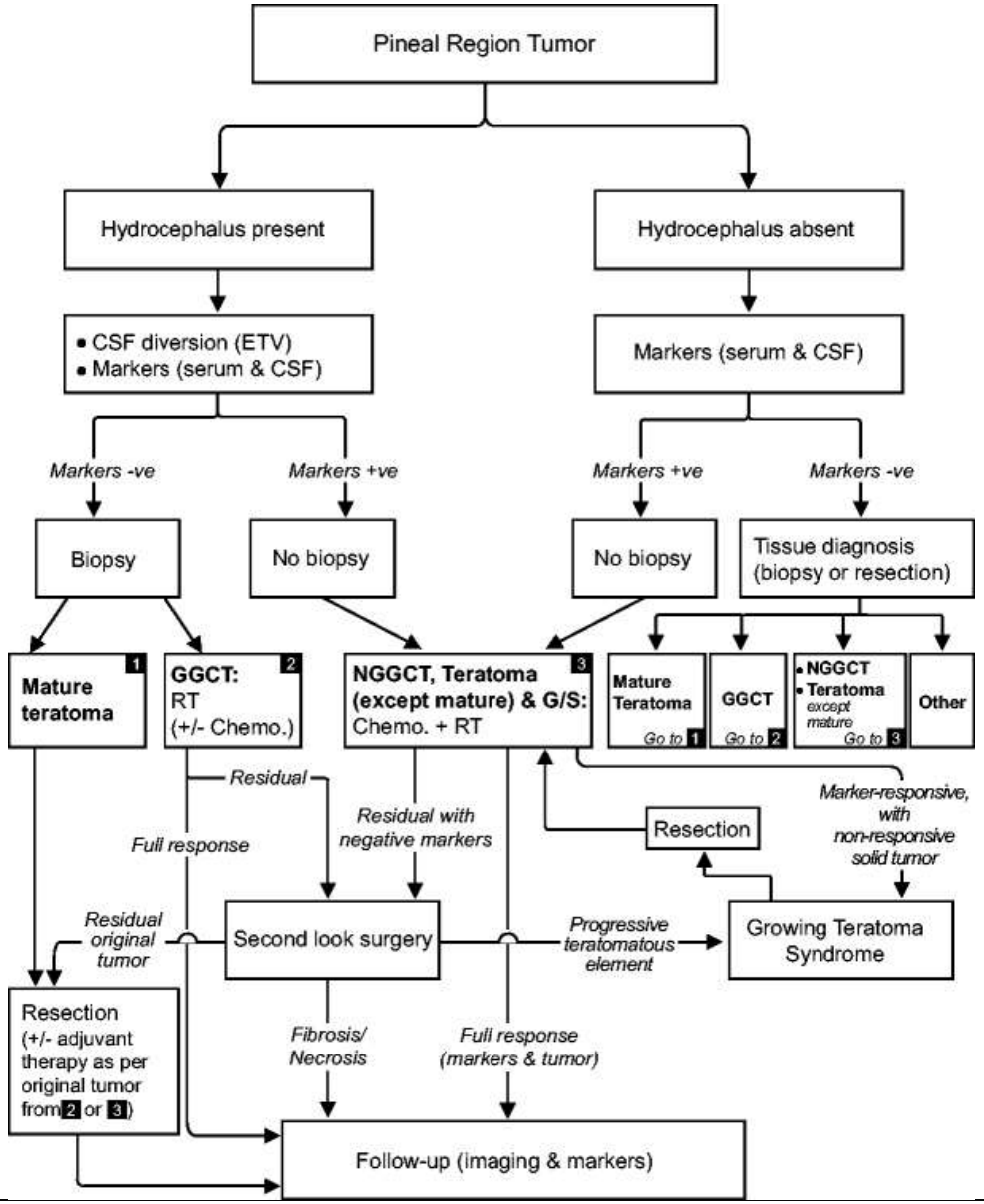


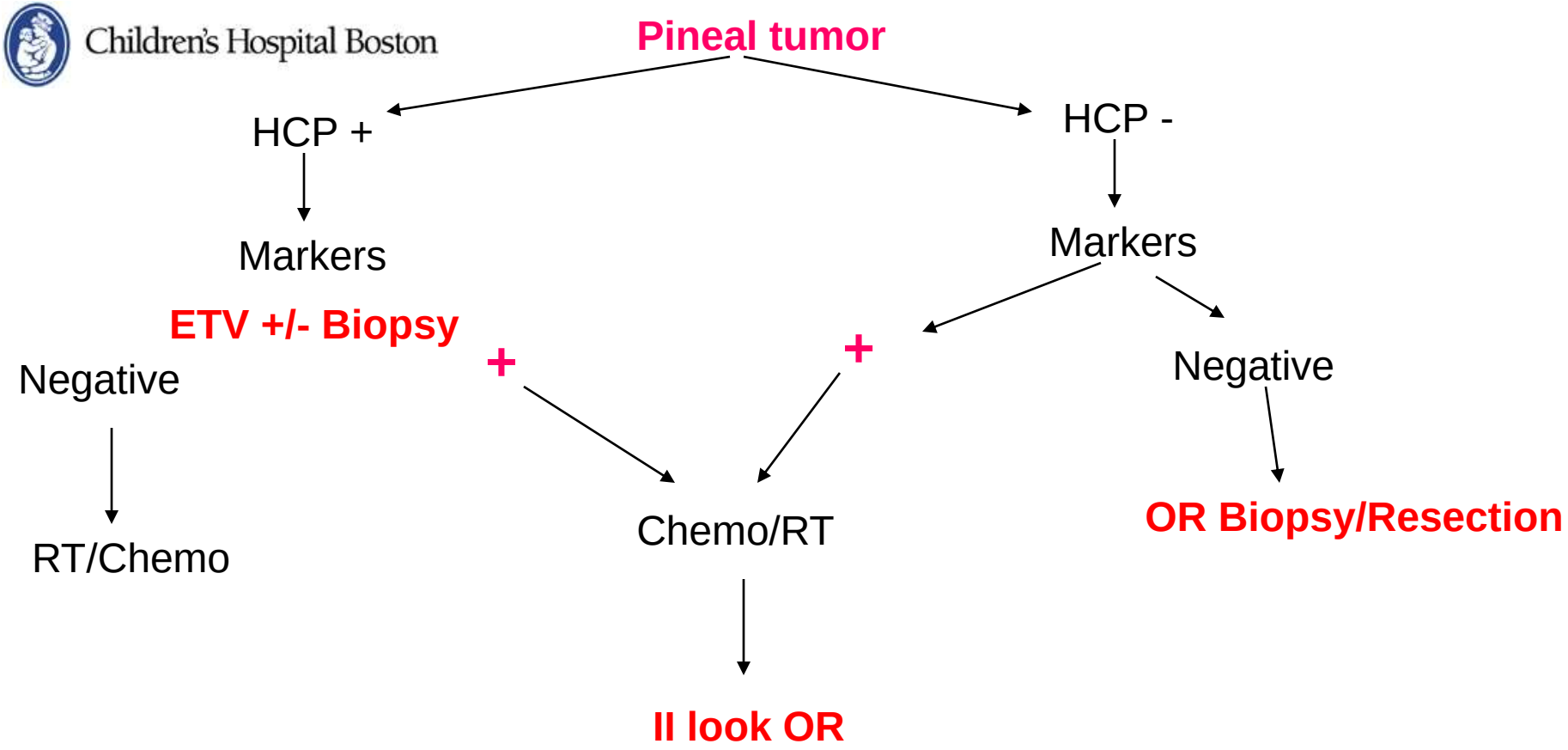
Rigid and flexible
Multiple burr holes (2)
Neuronavigation



- Disease free 4 yrs
- Recovering from initial neurologic deficits
- Single shunt system
- Normal markers




Pineal Region Tumor
Hydrocephalus present
Hydrocephalus absent
• CSF diversion (ETV)
• Markers (serum & CSF)
Markers (serum & CSF)
Markers -ve
Markers +ve
Markers +ve
Markers -ve
Biopsy
No biopsy
No biopsy
Tissue diagnosis (biopsy or resection)
1 Mature teratoma
2 GGCT: RT (+/- Chemo.)
3 NGGCT, Teratoma (except mature) & G/S: Chemo. + RT
Mature Teratoma Go to 1
GGCT Go to 2
• NGGCT • Teratoma except mature Go to 3
Other
Residual
Full response
Residual with negative markers
Resection
Marker-responsive, with non-responsive solid tumor
Residual original tumor
Second look surgery
Progressive teratomatous element
Growing Teratoma Syndrome
Resection (+/- adjuvant therapy as per original tumor from 2 or 3)
Fibrosis/ Necrosis
Full response (markers & tumor)
Follow-up (imaging & markers)


Pineal tumor
HCP +
HCP -
Markers
Markers
ETV +/- Biopsy
Negative
+
+
Negative
RT/Chemo
Chemo/RT
OR Biopsy/Resection
II look OR

Germ Cell Tumor/Pineal Tumor Management

- Hydrocephalus Yes
- CSF diversion procedure
  - ETV +/- biopsy
- Markers (serum and CSF)
  - Positive markers – proceed with chemotherapy
  - Non-contributory markers – tissue diagnosis
  - Endoscopic
  - Craniotomy

- Hydrocephalus No
- Markers (serum and CSF)
  - Positive markers – proceed with chemotherapy
  - Non-contributory markers – tissue diagnosis
  - Craniotomy

Combined Role of Endoscopy and Surgery

- ETV to treat HCP
- Biopsy in select cases
- Surgery for diagnosis
- Surgery at recurrence
- Surgery for growing teratoma syndrome
- II look surgery

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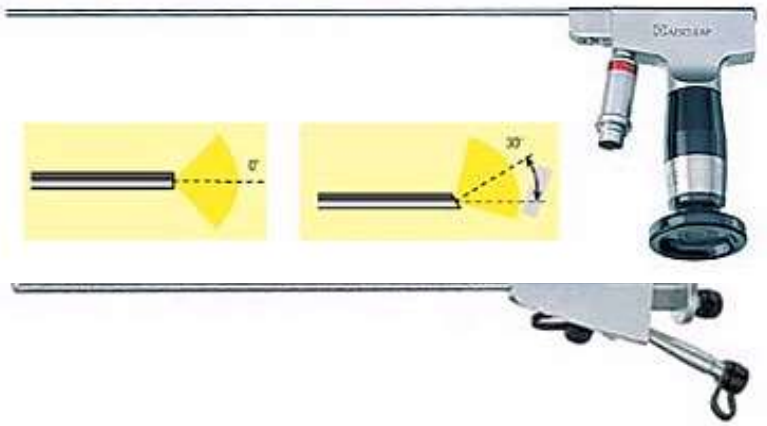
Thank you

Endoscopy



- Endoscopic procedures
  - bypass obstruction so that CSF can be absorbed via natural mechanisms and pathways
  - simplify shunt systems

Endoscopy - Equipment

- Rigid endoscopes
  - Rod lens telescope (Hopkins)
  - Introducer (sizes vary)
  - Working channels (0-4)
  - Holders (pneumatic, rigid)
  - Instruments (forceps, cautery, scissors, irrigation/suction)
  - Straight or angled (30º, 70º)
  - Camera
  - Light source
  - Recording equipment
- Flexible endoscopes
  - Fiberoptic (# of cables vary)
  - Introducer (different sizes)
  - Working channels (1-2)
  - Instruments
  - All are 0º
  - Provide 90º to 150º of movement
  - Holders (rigid)