Surgical approaches to pineal region tumors

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Dr P Sarat Chandra
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History

- Pinealis – pine cone
- Vedas – one of the 7 centers of vital energy
- Herophilus first described the pineal gland
- Descartes – seat of the human soul
- Studnicka 1905 – glandular function
- Lerner 1958 – melatonin discovered
- Derlincort 1717 – first pineal tumor
History of surgery of pineal tumors

• Horsley – 1910  
  first attempted resection

• Krause – 1913  
  first successful surgery

• Dandy – 1921  
  parieto occipital transcallosal approach

• Van Wagenen – 1931  
  transcortical transventricular approach

• Poppen – 1960  
  occipital transtentorial approach

• Stein – 1971  
  popularized infratentorial supracerebellar approach
Introduction

• Deep seated
• Difficult to access
• Diverse pathologies
• Surrounded by important structures
• The depth to the pineal region is the same from all approaches

“Personally, I have never succeeded in exposing pineal region tumor sufficiently well to justify an attempt to remove it “

*Cushing (1932)*

*Pineal tumors are perhaps the most dangerous of all intracranial tumors to attack surgically.*

—*Walter E. Dandy*
Arterial supply

- **P1**
  - Quadrigeminal artery
    - superior colliculus

- **P2**
  - Medial posterior choroidal artery
    - Pineal body, corpora quadrigemina, tela choroidea, thalamus
  - Lateral posterior choroidal artery
    - Choroid plexus lat ventricle, LGB, Thalamus

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

- **SCA**
  - Inferior colliculus
• Pineal parenchymal tumor
  » Pinealocytoma
  » Pinealoblastoma
  » Pineal parrenchymal tumor of intermediate differentiation
  » Papillary tumor of pineal region

• Germ cell tumors
  » Germinoma
  » Non germinomatous germ cell tumor
    • Embryonal carcinoma
    • Yolk sac tumor (endodermal sinus tumor)
    • Choriocarcinoma
    • Teratoma (mature, immature, malignant)
    • Mixed germ cell tumor

• Glial cell tumors
  » Astrocytoma
  » Oligodendroglioma
  » Ependymoma
  » Choroid plexus papilloma
  » Anaplastic astrocytoma/ GBM

• Mesenchymal cell tumors
  » Meningioma
  » Cavernoma/ haemangioblastoma

• Other tumors
  » Epidermoid
  » Craniopharyngioma
  » Ganglioglioma
  » Lipoma

• Metastasis
• Lymphoma
• Non neoplastic mass
  » Pineal cyst
  » Arachnoid cyst
  » Cysticercosis
  » Tuberculoma
  » Sarcoidosis
  » Aneurysm of vein of Gallen
Pineal tumors WHO 2007

<table>
<thead>
<tr>
<th>GERM CELL TUMOURS</th>
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<tbody>
<tr>
<td>Germinoma</td>
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<tr>
<td>Embryonal carcinoma</td>
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<tr>
<td>Yolk sac tumour</td>
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<tr>
<td>Choriocarcinoma</td>
</tr>
<tr>
<td>Teratoma</td>
</tr>
<tr>
<td>Mature</td>
</tr>
<tr>
<td>Immature</td>
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<tr>
<td>Teratoma with malignant transformation</td>
</tr>
<tr>
<td>Mixed germ cell tumour</td>
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</tbody>
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<table>
<thead>
<tr>
<th>grade</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
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## Pineal mass with age

<table>
<thead>
<tr>
<th>Age group</th>
<th>Most common</th>
<th>Less common</th>
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<tbody>
<tr>
<td>Infants</td>
<td>Pinealoblastoma</td>
<td>Arachnoid cyst Vein of Galen malformation</td>
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<tr>
<td>Childhood</td>
<td>Germinoma Glioma Tuberculoma</td>
<td>Pinealoblastoma Pineal cyst</td>
</tr>
<tr>
<td>Young adults</td>
<td>NGGCT Glioma</td>
<td>Pinealocytoma Pineal cyst</td>
</tr>
<tr>
<td>Older adults</td>
<td>Pinealocytoma Glioma</td>
<td>Meningioma Epidermoid Metastasis</td>
</tr>
</tbody>
</table>
Presentation

• Hydrocephalus
• Brainstem compression
  • Parinaud’s syndrome
  • Downgaze palsy
  • Dorsal midbrain compression/ infiltration – Lid retraction/ ptosis
  • Rarely IV palsy
  • Inferior colliculus compression – hearing disturbance
• Cerebellar signs
  • Superior peduncle – ataxia, dysmetria
• Endocrine disturbance
  – Diabetes Insipidus
  – Precocious puberty: β-HCG secretion. In chorio ca./Germinoma with NSGCT – androgen secretion by Leydig cells
• Pineal apoplexy
  – In vascular tumors: Pineal cell tumors/Choriocarcinoma
Imaging

- **X ray**
  - Calcification below 10 years is abnormal
- **CE MRI**
  - Spine should be imaged in all
    - Size and extent
    - Relation to surrounding structures
    - MRV
- **CT**
  - Rarely required
  - Augments information from MRI
    » Calcification
    » BBB breakdown
    » Vascularity
Germ cell tumor

- CT - Hyperdense
  - Sharp borders
  - Intrinsic calcification

- T1 - Hypo
  - germinoma

- T2 - Hypo
  - Uniform intense enhancement

Young
M>F
Choriocarcinoma- haemorrhage
Teratoma - calcification

Mixed germ cell tumor
Pinealoblastoma

Homogenous hyperintense on CT
Exploded (peripheral) calcification
Isointense on T1
Iso – hypointense on T2
Slightly non uniform enhancement
Areas of haemorrhage
## Imaging

<table>
<thead>
<tr>
<th>Tumor</th>
<th>CT</th>
<th>T1</th>
<th>T2</th>
<th>CMRI</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pineal cyst</td>
<td>Hypodense</td>
<td>Hypo</td>
<td>Hyper</td>
<td>Peripheral enhancement</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Rim calcification</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Germ cell tumors</td>
<td>Hyperdense</td>
<td>Hypo</td>
<td>Hypo</td>
<td>Uniform intense enhancement</td>
<td>Young M&gt;F</td>
</tr>
<tr>
<td></td>
<td>Sharp borders</td>
<td></td>
<td></td>
<td></td>
<td>Choriocarcinoma-haemorrhage</td>
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<tr>
<td></td>
<td>Intrinsic calcification</td>
<td></td>
<td></td>
<td></td>
<td>Teratoma - calcification</td>
</tr>
<tr>
<td>Pineal parrenchymal tumors</td>
<td>Hyperdense</td>
<td>Iso- hypo</td>
<td>Blastoma – iso/hypo</td>
<td>blastoma – slightly non uniform</td>
<td>Haemorrhage Non uniform borders</td>
</tr>
<tr>
<td></td>
<td>Blastoma-homogenous</td>
<td></td>
<td>Cytoma hyper</td>
<td>Blastoma – non uniform</td>
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<tr>
<td></td>
<td>Cytoma non homogenous</td>
<td></td>
<td></td>
<td>Cytoma – non uniform</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Exploded calcification</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Papillary tumor of pineal region</td>
<td>Variable</td>
<td></td>
<td>Marked hyper</td>
<td>Variable non homogenous</td>
<td>Cystic areas</td>
</tr>
<tr>
<td>Glioma</td>
<td>Hypodense</td>
<td>Iso/ hypo</td>
<td>hyper</td>
<td>Variable non homogenous</td>
<td>Adults</td>
</tr>
<tr>
<td></td>
<td>Calcification rare</td>
<td></td>
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</tbody>
</table>
Imaging

– ANATOMICAL relationships
  • Involvement of 3\textsuperscript{rd} ventricle/ position within 3\textsuperscript{rd} ventricle
  • Superolateral extension into ventricular trigone
  • Location of deep venous system and its relation to the tumor
  • Supratentorial spread of lesion
Tumor markers

- Presence indicates malignant germ cell tumor, converse not true
- More significance in follow up/ recurrence
- Help avoid unnecessary surgery
## Tumor markers

<table>
<thead>
<tr>
<th>Tumor</th>
<th>β-HCG</th>
<th>AFP</th>
<th>PLAP</th>
</tr>
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<tbody>
<tr>
<td>Germinoma</td>
<td>+(CSF)</td>
<td>-</td>
<td>+(CSF)</td>
</tr>
<tr>
<td>Chorionic carcinoma</td>
<td>++</td>
<td>-</td>
<td>+/-</td>
</tr>
<tr>
<td>Yolk sac tumor</td>
<td>-</td>
<td>++</td>
<td>-</td>
</tr>
<tr>
<td>Embryonal Ca</td>
<td></td>
<td></td>
<td>variable</td>
</tr>
<tr>
<td>Mature teratoma</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Immature teratoma</td>
<td>+/-</td>
<td>+/-</td>
<td>-</td>
</tr>
<tr>
<td>Mixed GCT</td>
<td></td>
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</tbody>
</table>
CSF analysis

• Cytology for cells

• Tumor markers
  • Non secreting tumors – CSF level increased, not detectable in blood
  • Germinoma β HCG and PLAP only in CSF
Management

• Hydrocephalus
  • EVD
  • Shunt
  • ETV (+/- biopsy)

• Tissue diagnosis
  • ETV + biopsy
  • Stereotactic biopsy
  • Open surgery

• Tumor control
  • Radiotherapy
  • Surgery
Hydrocephalus

• Present in almost all cases
• Must be addressed prior to tumor surgery
• Stable patient, complete resection likely, temporary EVD at time of surgery
• Symptomatic raised ICP
  • ETV +/- biopsy
    » Gradual reduction of ICP
    » Avoids peritoneal seeding
    » Avoids shunt related complications
  • VP shunt
SURGICAL ANATOMY

• Most tumors arise from or attached to 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 Foramen of Monroe
SURGICAL ANATOMY

• Mostly, ICV, Galen, Rosenthal & precentral cerebellar veins surround or cap the periphery of these tumors.

• Rarely, ICV are ventral to tumor.

• Highly vascular tumors
  – Pineoblastomas
  – Hemangioblastomas
  – Hemangiopericytomas (Angioplastic meningioma)
Surgery common approaches

- **Infratentorial supracerbellar**
  - Approach to centre of tumor
  - Minimizes risk to veins
  - Good exposure
  - No violation of normal tissue

- **Occipital transtentorial / Transcallosal interhemispheric**
  - Tumors extending superiorly
  - Extending laterally
  - Displaces veins ventrally
  - Large tumors
  - Greater exposure
Choice of approach

• Location of tumor (tentorial incisura)
• Tumor morphology (lateral extent)
• Displacement of great veins
• Probable diagnosis on imaging
• Angle of tentorium/ posterior fossa size
• Surgeons preference
Infratentorial supracerebellar approach

• Position
  • Sitting preferred
    – Can also be done in Concorde position
  • Large ventricle/ <3 years – 3 quarter prone
  • Table should be able to go low
  • Head flexed to keep tentorium parallel to floor
  • Patient tilted forward
Infratentorial supracerbellar approach

• Exposure
  • Incision – inion to C4, spinous process of C2 exposed
  • Burrhole – above torcula, lateral aspect of transverse sinus
  • Craniotomy – above transverse sinus and torcula
  • Bone edges waxed
  • If dura tense release CSF (ventricular tap)
  • Dural incision – curved between lateral most aspect of transverse sinus
  • Dura retracted avoid excess retraction – sinus occlusion
Infratentorial supracerebellar approach

• Surgical technique
  – Cauterize and divide adhesions and veins between cerebellum and tentorium
  – Retract vermis postero – inferiorly
  – Open arachnoid over the tumor (opaque white), midline precentral cerebellar vein may be divided
  – Small branches of choroidal and SCA over tumor divided
  – Trajectory of dissection changed towards the tumor
  – **Internal debulking of tumor**
  – **Lateral** walls dissected, vessels on it are choroidal and may be sacrificed
  – Dissection of **inferior** tumor from brainstem – most dangerous part, assistant retracts capsule upwards
  – Final dissection – **superior** along velum interpositum, great veins at risk
Infratentorial supracerbellar approach

• Mortality 3-4%

• Complications
  • Transient ocular dysfunction
  • Ataxia
  • Cognitive impairment, akinetic mutism – brainstem handling
  • Bleed in incompletely resected tumor
<table>
<thead>
<tr>
<th>Advantage</th>
<th>Disadvantage</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Gravity aided drainage of blood/ CSF</td>
<td>• Air embolism</td>
</tr>
<tr>
<td>• Gravity aided cerebellar retraction</td>
<td>• Surgeon fatigue</td>
</tr>
<tr>
<td>• Midline – orientation easy</td>
<td>• Difficult in very young and old</td>
</tr>
<tr>
<td>• No neural structures en route</td>
<td>• Quadriplegia from excessive flexion in elderly</td>
</tr>
<tr>
<td></td>
<td>• Hypotension</td>
</tr>
</tbody>
</table>
Lateral paramedian infratentorial

• INDICATIONS
  • Biopsy
  • Small quadrigeminal area tumor

• ADVANTAGE
  • Minimal damage to neural tissues
  • Useful in steep tentorium
  • Reduced risk of air embolism (lateral position)

• DISADVANTAGES
  • Narrow space
  • Difficult to reach tumor portion extending to infero posterior 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

• 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.
Occipital transtentorial approach

• Commonest supratentorial approach

• Indications
  – Predominantly supratentorial
  – Corpus callosum extension
  – Lateral extension into cerebral hemisphere
  – Thalamic extension
  – Predominantly third ventricular mass

• Advantage
  – Extensive tumor view
  – Managing bleeding is easier
  – Working distance is smaller
  – Access to pineal, third ventricle, midbrain, superior vermis

• Disadvantages
  – View obstructed by Galenic venous system
  – Restricted view of opposite side
Occipital transtentorial approach

• Position
  – Lateral decubitus with
    • rt side down
    • Midsagittal plane 30’ above horizontal
  – Three quarter prone
  – Prone
  – Sitting

• Craniotomy
  – 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 approach

• Surgical steps
  – Occipital retraction to be kept minimum
  – CSF release (from posterior callosal/ dorsal mesencephalic cisterns)
  – Opening of arachnoid (venous system lies in it)
  – Yasargil – positively identify vein of Rosenthal – Galen junction (Vein of Rosenthal may be mistaken for darkly colored dorsal mesencephalic cistern)
  – Tentorium incised 5 – 10 mm from the midline, medial flap sutured to falx
  – Identify and preserve IV nerve when manipulating tent
  – Precentral cerebellar vein may be sacrificed
Occipital transtentorial approach

- 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 haemostasis, water-tight dura closure.
Transcallosal interhemispheric

• Indications
  • Predominantly supratentorial tumor

• Position
  • Sitting/prone preferred
  • Lateral/3 quarter prone

• Surgery
  – U shapes skin flap across the midline
  – Bone flap across the midline
  – Position of bone flap depending on centering of the tumor
  – Wide craniotomy for alternate corridors to avoid bridging veins
  – Avoid sacrifice of more than 1 bridging vein
  – Pericallosal retracted
  – Callosotomy <2 cm centered over the tumor bulge
  – Identify deep veins early
Transcortical transventricular

• **Indication**
  – Tumor extending into lateral ventricle

• **Disadvantage**
  • Limited exposure
  • Cortical incision required
  • Stereotactic guidance may be required
Combined supra – infratentorial trans sinus
Complications of supratentorial approach

• Hemiparesis
  – Brain retraction
  – Sacrifice of bridging veins

• Sensory stereognostic deficits
  – Parietal lobe retraction injury

• Visual field defects
  – Occipital lobe retraction injury

• Disconnection syndrome
  – Corpus callosum section

• Memory defects
  – Fornix injury

• Bleed in residual tumor
• Venous infarction
Stereotactic biopsy

• Indications
  – Invasive disseminated tumor at diagnosis
  – Multiple medical problems
  – Selected cases with very large tumors
  – Neonate with large tumor (highly malignant, poor prognosis)
  – Presentation suggestive of infectious/ metastatic disease with diffuse systemic disease

• Target selection
  – Avidly enhancing tumor, preferably from the centre
  – Multiple sites
Trajectories

• **Orthogonal lateral (orange)**
  • Traverses the temporalis muscle
  • Technically difficult using a stereotactic frame

• **Oblique anterolateral (green)**
  • Most preferred
  • Low frontal trajectory below the plane of the internal cerebral veins

• **Posterolateral (pink)**
  • Lesions with significant lateral extension
Radiotherapy

• Primary
  – Germinoma

• Adjuvant
  – Pinealoblastoma (55 Gy to bed, 35 Gy to spinal axis)
  – Pinealocytoma (NO EFFECT on survival in incompletely excised tumors)
  – All malignant germ cell/ pineal cell neoplasm
  – CAN BE withheld for HISTOLOGICALLY benign COMPLETELY resected pinealocytoma, ependymoma
GKRS

- Histologically confirmed
- Maximum experience with pinealocytoma
- Used as an adjuvant therapy
- Possibly primary therapy for pinealocytoma
- Indications still evolving
- Current possible indications
  - Pineal paranchymal tumors
  - Germinoma
  - NGGCT
  - Astrocytoma
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
<table>
<thead>
<tr>
<th>Approach</th>
<th>Advantage</th>
<th>Disadvantage</th>
<th>Indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Midline innfratentorial</td>
<td>Midline – orientation</td>
<td>Air embolism</td>
<td>Midline masses No extension laterally/above incisura Tumor &lt; 3 cm</td>
</tr>
<tr>
<td>supracerebellar (Krause)</td>
<td>Tumor below major veins Gravity assists retraction</td>
<td>Narrow corridor Sacrifice of veins – infarction Difficult to reach above incisura</td>
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</tr>
<tr>
<td>Lateral paramedian</td>
<td>No sacrifice of veins</td>
<td>Only for small tumors SCA and branches at risk Cannot see posterior 3rd ventricle</td>
<td>Small tumors below tentorial notch with unilateral lateral extension</td>
</tr>
<tr>
<td>infratentorial (Van Wagenen)</td>
<td>Possible with steep slope of tent Less air embolism</td>
<td></td>
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<tr>
<td>Occipital transtentorial</td>
<td>Good view of structures above and below the tent</td>
<td>Retraction damage to occipital lobe Damage to splenium Cannot see posterior third ventricle</td>
<td>Tumors extending above and below tentorial incisura Tumors with unilateral lateral extension</td>
</tr>
<tr>
<td>(Poppen)</td>
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<tr>
<td>Posterior transcallosal</td>
<td>Lesion above tentorial notch with extension into 3rd ventricle</td>
<td>ICV in approach Callosotomy – disconnection syndrome Parietal lobe retraction damage</td>
<td>Posterior 3rd ventricular mass Mass between splenium and venous system</td>
</tr>
<tr>
<td>(Dandy)</td>
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<td>---------------------------------------------------------------------------</td>
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<td>-----------------------------------------------------------------</td>
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<tr>
<td>Posterior transventricular (Van Wagenen)</td>
<td>Exposes atrium and posterior body of lateral ventricle</td>
<td>Fornix section – memory deficit Seizures</td>
<td>Tumor extending into posterior lateral ventricle</td>
</tr>
<tr>
<td>Anterior transcallosal, transventricular transvellum interpositum (Sano)</td>
<td>Wider room No fornix section Supine – low air embolism</td>
<td>Increased depth of approach Callosotomy defecits Fornix damage by retraction</td>
<td>Large tumors extending anteriorly in 3rd ventricle</td>
</tr>
<tr>
<td>Combined supra – infra tentorial trans sinus (Ziyal and Sekhar)</td>
<td>Access tumor above and below tent Amole room Sinus may be resutured</td>
<td>Division of transverse sinus – venous infarcts / delayed raised ICP</td>
<td>Large meningioma, epidermoid, teratoma</td>
</tr>
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Results of Pineal Region Surgery at the New York Neurological Institute (1990-2008)

<table>
<thead>
<tr>
<th>Total Procedures</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>Total Procedures</td>
<td>128</td>
</tr>
<tr>
<td>Benign pathology</td>
<td>55 (43%)</td>
</tr>
<tr>
<td>Malignant pathology</td>
<td>73 (57%)</td>
</tr>
<tr>
<td>Diagnosis established</td>
<td>127 (99%)</td>
</tr>
</tbody>
</table>

Surgical Morbidity

<p>| | |</p>
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<thead>
<tr>
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<tbody>
<tr>
<td>Death (pulmonary embolism/cerebellar infarct)</td>
<td>2 (2%)</td>
</tr>
<tr>
<td>Permanent major morbidity</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Transient major morbidity (with recovery)</td>
<td>7 (5%)</td>
</tr>
</tbody>
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