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# Surgical outcome of pineal region lesions in paediatric population

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Keywords
paediatric population, pineal region lesions, surgical outcome

## ABSTRACT

**Introduction.** Pineal region pathologies are heterogenous, spectrum ranging from benign, infective to malignant in nature such as parenchymal, germ cell, glial, melanomas, metastatic, tubercular, etc. These lesions are commoner in the pediatric age group and have a variable outcome. The goal of this study is to present our experience regarding the surgical outcomes of pineal region lesions in the paediatric population.

**Objective.** The objective of this study is to access the surgical outcome of pineal region lesions in the paediatric population

**Methods and Material.** Eighty cases of pineal region lesions in the paediatric population (up to 15 years of age) operated in the neurosurgery department, G.B Pant Hospital New Delhi via either Occipital transtentorial or Supracerebellar Infratentorial approaches were retrospectively analysed.

**Results.** There were four main groups of lesions with pineal parenchymal tumours (35 cases) being the commonest. Glial tumours were the next most common contributing 29 cases. Germ cell tumours comprise 7 and miscellaneous 9 cases.

The outcome showed graded improvement with the extent of tumour removal and 57 patients (71.25%) improved following surgery. The clinical status of 20 patients (25%) remained unchanged and 3(3.75%) deteriorated from their pre-operative status. 3 patients (3.75%) died in the immediate post-operative period and 11(13.75%) died due to recurrent disease.

**Conclusion.** A graded increase in survival was noted with increasing the degree of resection and postoperative adjuvant therapy in malignant pineal tumours whereas benign pineal lesions can be managed with surgery. Pure germinomas are the only tumour for which the survival rate was unrelated to extent of tumour resection.

## INTRODUCTION

The pineal region lesions are heterogenous in nature such as germ cell tumours, glial tumours, pineal parenchymal tumors, primary melanoma, metastasis,cysts, infective, etc.[1,2,3]

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These tumours are common in children comprising of more than 10% of all pediatric tumours with common presentation of raised intracranial pressure secondary to obstructive hydrocephalous due to close relation with posterior third ventricle.[4,5,6]

With better microsurgical skills, the surgical outcome is associated with less morbidity and mortality, however the long term surgical and functional outcome depends on various factors such as age, extent of resection of tumour, pathological grade, distant seedling at the time of presentation, neurological condition, visual status, possibility of giving adjuvant therapy .[8,9,10, 11,12,13,14,15,16].

In the present study, we retrospectively analyzed the surgical outcome of pineal region lesion in children treated over last 15 years.

## Material and methods

Eighty cases of pineal region lesions in children operated via two approaches i.e occipital transtentorial and supracerebellar infratentorial (Fig1A,B) in the Department of Neurosurgery, GIPMER, New Delhi were retrospectively analyzed. The techniques for both the approaches were as standards described in the literature. Postoperatively clinical and radiological outcome was analyzed. Postoperative tumour excision was graded as gross total, near total, subtotal or biopsy when >99%, >90%, 51-90% or <50% tumour was resected respectively based on MRI study.

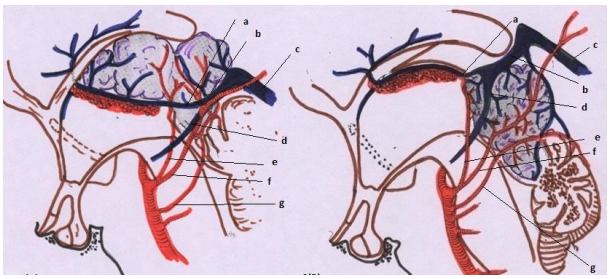


**Figure 1**. **A.** Occipital transtentorial; **B.** Supracerebellar Infratentorial.
a-inferior saggital sinus,b-great cerebral vein of Galen, c-transverse sinus, d-internal cerebral vein, e-posterior cerebral artery, f-superior cerebellar artery, g-anterior inferior cerebellar artery.

## Results

Eighty children were followed up for 1-5 years depending upon the pathology. Twenty children (25%) were in the age group of 4-6 years and 16 cases (15%) were 8-10 years old; the youngest patient being 2 years and oldest was14 years old with a mean age of 7.5 years. There were 39 males (48.75%) and 41females(51.25%) with the M:F ratio of 0.95:1(Table 1)

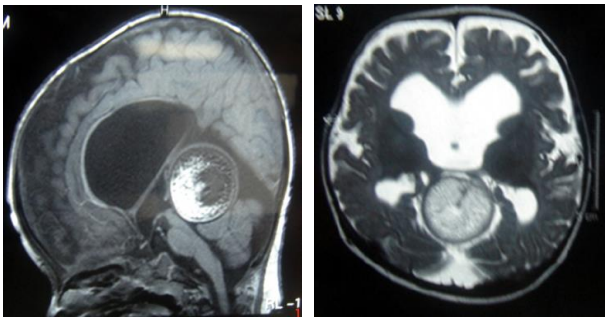

**Figure 2A.** Pre-operativeT1 saggital & T2 axial images of pineal region teratoma

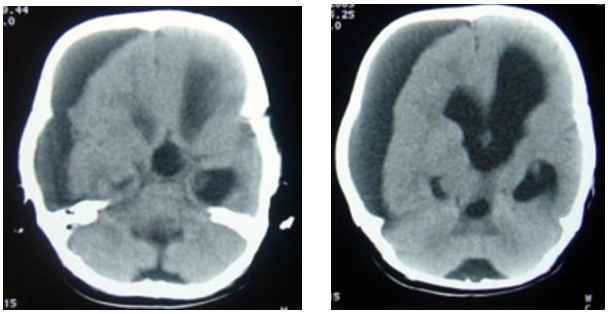

**Figure 2B.** Post-operative CT showing complete removal of Teratoma

Histopathologically pineal parenchymal tumors was constituting 43.75%, out of which pinealoblastoma(21.25%) constituted the major portion followed by glial tumours (36.25%), germ cell tumours(8.75%). Remaining lesions like meningioma, choroid plexus papilloma, epidermoid cyst and tubercular abscess and cysticercosis were uncommon. (Table 2).

Headache was the most common presenting symptom observed in all cases followed by vomiting in 41.25%, visual disturbance in 41.25%, diplopia in 31.25%, papilloedema 46.25%. Parinauds syndrome was present in 38.75%, weakness in upper or lower extremity was common in glial tumors infiltrating surrounding structures, 15% had cerebellar signs, 6.25% hormonal imbalance and 3.75% were in altered sensorium at the time of presentation.

Eighteen cases underwent HCG or AFP level assessment in CSF and 3 cases showed elevated levels in germinoma & teratoma.

Out of 80 cases,57 (71.25%) improved, 20 (25%) remained unchanged and 3(3.75%) patients deteriorated in postoperative period. Three patients (3.75%) died in immediate post-operative period and 11(13.75%) died in the follow up due to recurrent disease.

Surgical outcome based on extent of resection is given in table 4.Based on extent of resection, total removal led to improvement in 75-100 % cases irrespective of pathology. Improvement in pilocytic astrocytomas was 90%, pineocytoma 57%, pineoblastoma 47%, intermediate pineal tumor was 45% and 60% in germinoma cases. Improvement was almost 100% in benign lesions such as meningioma, epidermoid cyst, oligodendroglioma, choroid plexus papilloma, infective pathologies (Figure 3,5) Near total removal showed 47% improvement in pineoblastoma whereas patients with pathologies such as fibrillary astrocytoma, anaplastic astrocytomas, germinomas showed 100% improvement even after near total excision.

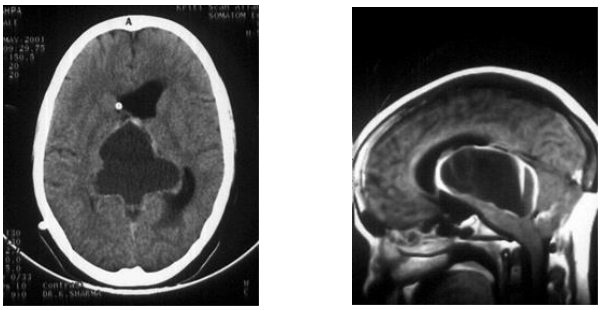

**Figure 3A & 3B**. Pre- operative axial & saggital images of pineal region epidermoid.

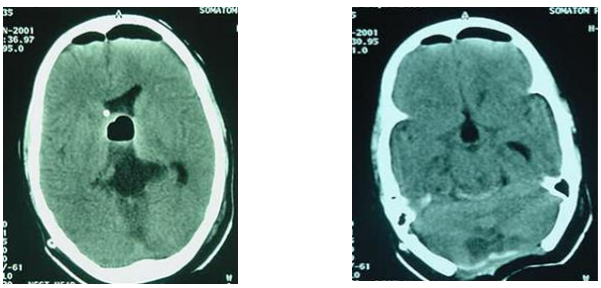

**Figure 3C & 3D.** Post-operative axial CT showing excision of pineal region epidermoid.

Except for germinoma subtotal removal of tumours showed no improvement in postoperative period. In the biopsy group out of 2 patients,50% remained unchanged and 50% deteriorated with one immediate and one delayed mortality. All cases of ependymoma improved after gross and near total resection. Out of 5 patients of germinoma, 3 patients improved after gross total removal, near total and subtotal resection and radiotherapy (Figure 4), 2 patients underwent biopsy out of which one remained unchanged and one deteriorated and died in post operative period due to disseminated disease. Out of 2 cases of immature teratoma (Figure 2A,B), 1 patient improved and one remained unchanged after near total excision.(Table 3 & 4). A CSF diversion procedure as ventriculoperitoneal shunt was needed in 31 patients(38.75%) of pineal tumour patients .

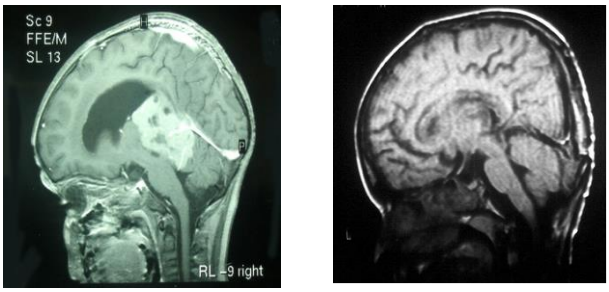


**Figure 4A & 4B.** Pre-operative saggital MR image in T1 contrast of pineal region germinoma & post operative saggital T1 MR after excision and radiotherapy.

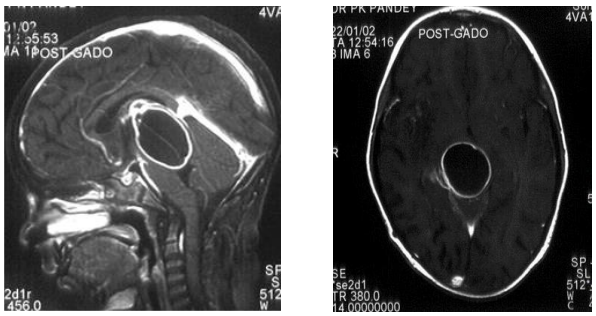


**Figure 5A & B.** Pre-operative saggital & axial T1 contrast MR images of tubercular abscess.

**Figure 5C & D.** Post-operative axial CT of tubercular abscess after excision of tubercular abscess.

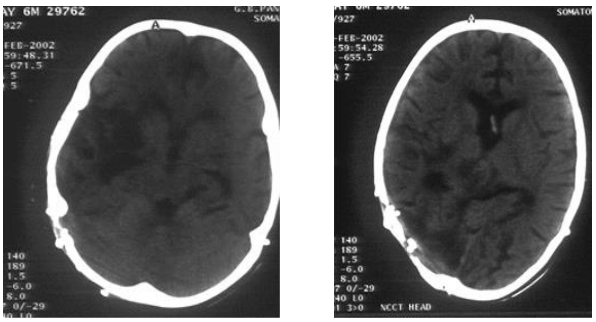

**Table 1.** Age and sex distribution

| Sr. No. | Age Group | No. of Females | No. of Males | Total No. of cases |
|---|---|---|---|---|
| 1 | 0-2 | 1 | 0 | 1 |
| 2 | 2-4 | 6 | 7 | 13 |
| 3 | 4-6 | 9 | 11 | 20 |
| 4 | 6-8 | 9 | 5 | 14 |
| 5 | 8-10 | 7 | 9 | 16 |
| 6 | 10-12 | 5 | 3 | 8 |
| 7 | 12-14 | 4 | 4 | 8 |
| 8 | 14-15 | 0 | 0 | 0 |
| | Total | 41 | 39 | 80 |
| | | 51.25% | 48.75% | |

**Table 2.** Histopathology of 80 cases of pineal region tumours

| Diagnosis | No. of Cases |
|---|---|
| • Pineal parenchymal tumors<br>• Pineocytoma<br>• Pineal parenchymal tumor of intermediate differentiation<br>• Pineoblastoma | 35 (43.75%)<br>7 (8.75%)<br>11 (13.75%)<br><br>17 (21.25%) |
| Gliomas<br>• Pilocytic astrocytoma<br>• Fibrillary astrocytoma<br>• Anaplastic astrocytoma<br>• Glioblastoma<br>• Oligodendroglioma<br>• Ependymoma<br>• Subependymal giant cell astrocytoma | 29 (36.25%)<br>10 (12.50%)<br>4 (5%)<br>3 (3.75%)<br>1 (1.25%)<br>3 (3.75%)<br>6 (7.50%)<br>2 (2.5%) |
| Germ cell tumors<br>• Germinoma<br>• Immature teratoma | 7 (8.75%)<br>5 (6.25%)<br>2 (2.5%) |
| Miscellaneous<br>• Meningioma<br>• Choroid plexus papilloma<br>• Epidermoid cyst<br>• Tubercular abscess<br>• Cysticercosis | 9 (11.25)<br>2 (2.25%)<br>3 (3.75%)<br>2 (2.25%)<br>1 (1.25%)<br>1 (1.25%) |
| **Total** | **80 (100%)** |

**Table 3.** Surgical outcome according to histopathology

| Diagnosis | No. of cases (n) | Outcome | | | Mortality (%) |
|---|---|---|---|---|---|
| | | Improved (%) | Unchanged (%) | Deteriorated (%) | |
| Pineocytoma | 7 | 4(57.14%) | 3(42.85%) | 0 | 0 |
| Pineal parenchymal tumor of intermediate differentiation | 11 | 5(45.45%) | 6(54.54%) | 0 | 4(36.36%) |
| Pineoblastoma | 17 | 8(47.05%) | 7(41.17%) | 2(11.76%) | 8(47.05%) |
| Pilocytic astrocytoma | 10 | 9(90%) | 1(10%) | 0 | 0 |

| | | | | | |
|---|---|---|---|---|---|
| Fibrillary astrocytoma | 4 | 4 | 0 | 0 | 0 |
| Anaplastic astrocytoma | 3 | 3 | 0 | 0 | 0 |
| Glioblastoma | 1 | 0 | 1 | 0 | 1 |
| Oligodendroglioma | 3 | 3 | 0 | 0 | 0 |
| Ependymoma | 6 | 6 | 0 | 0 | 0 |
| SEGA | 2 | 2 | 0 | 0 | 0 |
| Germinoma | 5 | 3(60%) | 1 | 1(20%) | 1(20%) |
| Immature teratoma | 2 | 1 | 1 | 0 | 0 |
| Meningioma | 2 | 2 | 0 | 0 | 0 |
| Choroid plexus papilloma | 3 | 3 | 0 | 0 | 0 |
| Epidermoid cyst | 2 | 2 | 0 | 0 | 0 |
| Tubercular abscess | 1 | 1 | 0 | 0 | 0 |
| Cysticercosis | 1 | 1 | 0 | 0 | 0 |
| **Total (%)** | **80 (100%)** | **57 (71.25%)** | **20 (25.00%)** | **3 (3.75%)** | **14 (17.5%)** |

**Table 4.** Surgical outcome based on extent of tumour removal

| **Diagnosis** | **No. of cases** | **Extent of tumour removal** | **No of cases** | **Outcome** | | | **Mortality** |
|---|---|---|---|---|---|---|---|
| | | | | **Improved** | **Unchanged** | **Deteriorated** | |
| Pineocytoma | 7 | GT | 4 | 3(75%) | 1(25%) | 0 | 0 |
| | | NT | 1 | 1 | 0 | 0 | 0 |
| | | ST | 2 | 0 | 2 | 0 | 0 |
| Pineal parenchymal tumor of intermediate differentiation | 11 | GT | 6 | 5(83.33%) | 1(16.67%) | 0 | 0 |
| | | NT | 2 | 0 | 2(100%) | 0 | 1 |
| | | ST | 2 | 0 | 2(100%) | 0 | 2 |
| | | Biopsy | 1 | 0 | 1(100%) | 0 | 1 |
| Pineoblastoma | 17 | GT | 5 | 4(80%) | 1(20%) | 0 | 0 |
| | | NT | 7 | 4(57.14%) | 3(42.85%) | 0 | 3 |
| | | ST | 3 | 0 | 2(66.66%) | 1(33.33%) | 3 |
| | | Biopsy | 2 | 0 | 1(50%) | 1(50%) | 2 |
| Pilocytic astrocytoma | 10 | Excision | 2 | 2(100%) | 0 | 0 | 0 |
| | | GT | 6 | 6(100%) | 0 | 0 | 0 |
| | | NT | 1 | 1(100%) | 0 | 0 | 0 |
| | | ST | 1 | 0 | 1(100%) | 0 | 0 |
| Fibrillary astrocytoma | 4 | GT | 3 | 3(100%) | 0 | 0 | 0 |
| | | NT | 1 | 1(100%) | 0 | 0 | 0 |
| Anaplastic astrocytoma | 3 | GT | 2 | 2(100%) | 0 | 0 | 0 |
| | | NT | 1 | 1(100%) | 0 | 0 | 0 |
| Glioblastoma | 1 | ST | 1 | 0 | 1(100%) | 0 | 1 |
| Oligodendroglioma | 3 | GT | 3 | 3(100%) | 0 | 0 | 0 |
| Ependymoma | 6 | GT | 5 | 5(100%) | 0 | 0 | 0 |
| | | NT | 1 | 1(100%) | 0 | 0 | 0 |
| SEGA | 2 | Excision | 1 | 1(100%) | 0 | 0 | 0 |
| | | GT | 1 | 1(100%) | 0 | 0 | 0 |
| Germinoma | 5 | GT | 1 | 1 | 0 | 0 | 0 |
| | | NT | 1 | 1 | 0 | 0 | 0 |
| | | ST | 1 | 1 | 0 | 0 | 0 |
| | | Biopsy | 2 | | 1(50%) | 1(50%) | 1 |
| Immature teratoma | 2 | NT | 2 | 1(50%) | 1(50%) | | 0 |
| Meningioma | 2 | Simpson's Grade III Excision | 2 | 2(100%) | 0 | 0 | 0 |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| Choroid plexus papilloma | 3 | GT | 3 | 3(100%) | 0 | 0 | 0 |
| Epidermoid cyst | 2 | Excision | 2 | 2(100%) | 0 | 0 | 0 |
| Tubercular abscess | 1 | Excision | 1 | 1(100%) | 0 | 0 | 0 |
| Cysticercosis | 1 | Excision | 1 | 1(100%) | 0 | 0 | 0 |

GT, gross total resection; NT, near total resection; SEGA, subependymal giant cell astrocytoma; ST, subtotal resection

Upgaze palsy was present in 3 which persisted in post-operative period. Subdural effusion was present in 4 cases & was self limiting. Most severe complication was postoperative haemorrhage in 2 cases due to incomplete resection of tumor. In Infratentorial supra-cerebellar approach venous air embolism & hypotension was noted in 8 cases. Venous air embolism was treated with rapidly packing the operative field with wet sponges,waxing bony edges, air aspiration from right atrium via CVP catheter, ventilation of patients with 100% O2, use of vasopressor & volume expanders.Postoperative hemianopia was the most common complication following suboccipital transtentorial approach and was encountered in 7 cases due to prolonged retraction of occipital lobe. Meningoencephalitis was noticed in one case.

Suitable candidates were referred for adjuvant treatment.

Five-year survival rate of the patients is shown in Table 5.

**Table 5.** Survival rates for patients with pineal tumours

| | 5 Year Survival |
|---|---|
| Pineocytoma (PC) | 71% |
| Pineal parenchymal tumour of intermediate differentiation (PID) | 63% |
| Pineoblastoma(PB) | 46% |
| High grade glial tumours | 80% |
| Germ cell tumours (GCTs) | 90% |

## Discussion

Transcallosal approach for pineal lesions was first described by Dandy18 in 1921, however all patients died initially and first successful excision was performed in 1931. The operative mortality of pineal region lesions was as high as 90% as per reported by Russel and Sachs 19 in 1943. Thereafter with more refined surgical skills and knowledge of anatomy, increasing sophistication of stereotactic surgery and the high morbidity and mortality rates of open operation on the pineal region, neurosurgeons advocated stereotactic biopsy for pineal region masses. However with recent advances, surgery on the pineal region has became increasingly safer & is now the treatment of choice as it allows significant debulking of lesions and adequate specimen to identify the pathology of lesion. We are also in favor of surgery to radically remove the lesion to avoid unnecessary radiation to benign lesions and radiation associated significant morbidity particularly in children. Radiation can cause arachnoid thickening, adhesions and which can be troublesome especially for recurrent tumours. Hoffman et al.20 also recommended open surgery in order to establish accurate histological diagnosis to guide the adjuvant therapy indications.

The median age of presentation in this study was 7.5years (range 1-15) and male to female ratio was 0.95:1 (Table 1).

In the present study pineal parenchymal tumours(PPT) were most common constituting 43.75% followed by glial tumors comprising 36.25% and germ cell tumours<9%.In Indian population PPT is commoner than germ cell tumours(Table 2). TatkeM et al. reported 42% of tumours to be PPT in Indian population.24 In contrast to this, Cho BK et al. reported even higher incidence of both PPT and germ cell tumours i.e. 69% and 19% respectively in Far East population22. Aal-Hussaini M et al collected data from 17 SEER registries (these data included a total of 5306606 tumors diagnosed from January 1973-December 2005) and reported germ cell tumors as most common histological type followed by pineal parenchymal tumors23. Rosenstock17 et al and reported 32% pineal parenchymal tumors and 26% glial tumours respectively in their series.

In this study headache was present in all cases followed by vomiting and diplopia in 41.25% and 38.75%respectively. Papillodema was observed in 46.25% with Parinauds syndrome in 38.75%.Various studies by Hoffman et al,20, Villa et al, 25Konovalov and Pitskhelauri3 et al , Cho B.K et al also found signs and symptoms of raised intracranial pressure in 80-

90% and Parinaud's syndrome in 25%-50% of cases.

Al-Hussaini M et al collected data from 17 SEER registries (these data included a total of 5306606 tumors diagnosed from January 1973-December 2005) and concluded that amoung malignant tumours of pineal region germ cell tumours had best outcome, which was further improved by radiotherapy use but not by total excision23.

Konovalov AN and Pitskhelauri DI3 et al reported pure germinomas having 95% and 88% 5 year and 10 year survival; high grade glial tumors as 80% and 50%, 5 year and 10 year survival rate; malignant pineal parenchymal tumours as 44% and 0% 5 year and 10 year survival rate; malignant GCTs as 20% and 0% 5 year and 10 year survival rates respectively. In our study highest mortality was noted in cases of pineoblastoma(47.05% mortality), which is the most malignant tumour of pineal region followed by pineal parenchymal tumor of intermediate differentiation(36.36%). In our study pineal parenchymal tumor of intermediate differentiation had 70% overall 5 year survival rate; pineoblastoma having 70.58% overall 5 year survival rate; germinoma having 75% overall survival rate. There is 34.28% mortality in pineal parenchymal group followed by 20% mortality in germ cell tumor group and 3.44% mortality in glial tumor group without any mortality in bening lesions of pineal region, correlating mortality with the histopathological diagnosis.

Konovalov AN and Pitskhelauri DI3 found an association between the extent of tumor removal and survival in patients with all types of malignant tumors except pure germinoma. The 5 year survival rate was 70% for patients who had a total tumor resection and 30% for patients who had a biopsy or partial resection. Pure germinomas was the only tumor for which survival rate was not associated with the extent of tumor resection. Tatke M et al reported a graded increase in survival with increasing degree of resection (5 year survival rate:84% for GTR vs 53% for STR vs 29% for debulking in case of pineoblastomas. (Table4,5).

Konovalov AN and Pitskhelauri DI3 found an association between the extent of tumor removal and survival in patients with all types of malignant tumors except pure germinoma. The 5 year survival rate was 70% who had a total tumor resection and 30% who had a biopsy or partial resection. Pure germinomas was the only tumor for which survival rate was not associated with the extent of tumor resection perhaps these are the tumours which responds best to radiotherapy. Tatke M24 et al also reported a graded increase in survival with increasing degree of resection in pineoblastomas.

Clark AJ et al suggested that aggressive surgical resection provides a survival benefit over subtotal resection for patients of pineocytomas 26. In our study, 75% of pineocytomas improved after gross total removal , 83.33% of pineal parenchymal tumors improved after gross total removal and 80% of pineoblastomas improved after gross total removal. In cases of pilocytic astrocytoma all cases of gross total and near total removal improved with one case of subtotal removal remained unchanged.

All cases of fibrillary astrocytoma, anaplastic astrocytoma, ependymomas and SEGA showed improvement after gross and near total excision. One case of glioblastoma expired after subtotal removal. All cases of germinomas showed improvement regardless of extent of removal except one case of disseminated germinoma which died in post op period and one case remained in unchanged status. All other miscellaneous lesions improved after excision, gross total or near total removal(Table3,4)

Konovalov AN and Pitskhelauri DI3 described the principles of treatment of the pineal region tumours on the basis of total 287 operations done over a period from 1976 to 1999. It included 161 males and 126 females with a mean age of 20 years at presentation. They used occipital transtentorial approach in 138(54%) and infratentorial supracerebellar approaches in 87(34%) of patients. Total removal was achieved in 58% patients, subtotal removal in 29% and partial removal in 13% patients. 58% patients had obstructive hydrocephalus that underwent craniospinal fluid shunting before radiotherapy or with tumour removal. Features of raised intracranial pressure and eye movement disorders were among the most common presentation. Present study, includes 80 patients with pineal region lesions. A CSF diversion procedure as ventriculoperitoneal shunt was needed in 31 patients(38.75%) of pineal tumour patients .

Two approaches were commonly used: Supracerebellar infratentorial approach was used in 62(77.5%) and occipital transtentorial in 18(22.5%) patients of pineal tumours. Stein BM 27 reported their experience of 6 cases of pineal lesions,

operated using Krause's Supracerebellar Infratentorial approach. They had no operative mortality. Operative morbidity was negligible and except for a varying degree of meningeal reaction managed effectively with steroids, their pateints had uneventfull postoperative courses.

Suzuki and Iwabuchi have reported the largest series of successfully treating lesions of the pineal region using the direct surgical approach. 17 of 19 cases underwent total or near total removal via a supratentorial approach.

In our study post operative complications according to surgical approach were as follows;

Infratentorial supracerebellar approach: Ocular abnormality in the form of upgaze palsy was present in 3 which lasted for few weeks in post-operative period. Subdural effusion was present in 4 cases & was self limiting. Most severe complication was postoperative haemorrhage in 2 cases due to incomplete resection of tumor.

Occipital transtentorial approach: Postoperative hemianopia was the most common complication in this approach encountered in 7 cases due to excessive retraction of occipital lobe. Gentle & gradual retraction of occipital lobe is necessary to avoid this complication. Meningoencephalitis was noticed in one case.

Clark AJ et al reported that patients receiving subtotal tumor resection (STR) versus those undergoing STR plus External beam radiotherapy (XRT), the addition of adjuvant radiation does not yield a survival benefit when compared to STR alone. In the light of the superiority of gross total resection (GTR) to STR, it appeared that pineocytomas are relatively radioresistant and are optimally treated with aggressive surgery rather than XRT.

Stoiber EM et al reported that fractionated local radiotreatment of pineocytomas seems to be safe and effective method with a local recurrence control rate of 100%, intensification of therapy in aggressive variants of intermediate pineocytomas as well as pineoblastomas seemed necessary28. The CCG-921(Childrens cancer group) report suggested that radiation therapy has a significant impact on survival with a 3-year survival of 61% in paediatric pineoblastoma population. Konovalov AN and Pitskhelauri DI et al reported that radiation therapy should be administered to all patients with pineal region tumors other than meningioma, mature teratoma and some rare bening tumors or cysts3.Germinoma is chemotherapy & radiosensitive. For intracranially arising germonimas radiation therapy is the primarily curative. Radiation therapy is best avoided in young children & many centres advocate few cycles of chemotherapy followed by radiation therapy as well. More aggressive tumors of pineal region like malignant germ cell and pineal parenchymal tumors tend to invade surrounding cerebellar structures and have a high risk of spinal dissemination and commonly cannot be totally resectable, so radiation therapy and chemotherapy play a crucial role in the treatment3.

Schild S.E et al. reported that pineoblastomas and pineal parenchymal tumor of intermediate differentiation, it is possible to obtain better results with surgical resection and relatively high doses of radiation (50 to 55 Gy)29. According to Tate M et al the addition of radiotherapy to gross total removal did not improve survival; however, the addition of radiotherapy to subtotal removal did yield a survival benefit in cases of pineoblastoma19. In our study total 30(37.5%) patients received radiotherapy and 12(15%) patients received chemotherapy. In patients with pineoblastomas, pineal parenchymal tumors of intermediate differentiation, germinomas, immature teratomas, anaplastic astrocytomas, patients who received post-operative radiotherapy have better overall and progression free survival. Chemotherapy as part of treatment for pineal tumors(cisplatin-etoposide combination) was used.

Many of our patients were in the advanced stage of their illness bearing large tumours which is an additive factor to the morbidity and mortality of our patients. In patients with bening lesions, surgical removal is curative. In malignant tumours, satisfactory decompression of tumour improve response to postoperative adjuvant therapy.

## Conclusion

Pineoblastoma is the most common histological type among pineal region tumours in children.A graded increase in survival noted with increasing the degree of resection and post-operative adjuvant therapy in malignant pineal tumors.Pure germinomas are the only tumor for which survival rate was not associated with the extent of tumor resection. Selection of the surgical approach (Supra/infra-tentorial) according to the extension of the tumor (above or below the

tentorium) may give window to the greater surgical resection and reflect the improved outcome.

## References


1. Bruce JN, Stein BM. Surgical management of pineal region tumors. Acta neurochirurgica. 1995 Sep;134(3):130-5.
2. Hernesniemi J, Romani R, Albayrak BS, Lehto H, Dashti R, Ramsey III C, Karatas A, Cardia A, Navratil O, Piippo A, Fujiki M. Microsurgical management of pineal region lesions: personal experience with 119 patients. Surgical neurology. 2008 Dec 1;70(6):576-83.
3. Konovalov AN, Pitskhelauri DI. Principles of treatment of the pineal region tumors. Surgical neurology. 2003 Apr 1;59(4):252-70.
4. Abay EO II, Laws ER, Grand GL, et al. Pineal tumors in children and adolescents. J Neurosurg1981;55:889–95.
5. Fuller BG, Kapp DS, Cox R. Radiation therapy of pineal region tumors: 25 new cases and a review of 208 previously reported cases. International Journal of Radiation Oncology* Biology* Physics. 1994 Jan 1;28(1):229-45.
6. Hart MN, Earle KM. Primitive neuroectodermal tumors of the brain in children. Cancer. 1973 Oct;32(4):890-7.
7. Bruce JN, Stein BM. Supracerebellar approaches in the pineal region. In: Apuzzo MLJ, ed. Brain surgery: complication, avoidance and management. New York: Churchill-Livingstone, 1993:511–36.
8. Jooma R, Kendall BE. Diagnosis and management of pineal tumors. Journal of neurosurgery. 1983 May 1;58(5):654-65.
9. Neuwelt EA, Glasberg M, Frenkel E, Clark WK. Malignant pineal region tumors: A clinico-pathological study. Journal of neurosurgery. 1979 Nov 1;51(5):597-607.
10. Neuwelt EA. An update on the surgical treatment of malignant pineal region tumors. In: Clinical neurosurgery: proceedings of the congress of neurological surgeons, Volume 32. Baltimore: Williams & Wilkins, 1985:397-428.
11. Cumins FM, Taveras JM, Schlesinger EB. Treatment of gliomas of the third ventricle and pinealomas. With special reference to the value of radiotherapy. Neurology 1960;10:1031–6. Neuwelt EA, Glasberg M, Frenkel E, Clark WK. Malignant pineal region tumors: A clinico-pathological study. Journal of neurosurgery. 1979 Nov 1;51(5):597-607.
12. Dandy WE. Operative experience in cases of pineal tumor. Archives of Surgery. 1936 Jul 1;33(1):19-46.
13. Davidoff LM. Some considerations in the therapy of pineal tumors. Bulletin of the New York Academy of Medicine. 1967 Jul;43(7):537.
14. Horrax G. Treatment of tumors of the pineal body: experience in a series of twenty-two cases. Archives of Neurology & Psychiatry. 1950 Aug 1;64(2):227-42.
15. Poppen JL, Marino R. Pinealomas and tumors of the posterior portion of the third ventricle. Journal of Neurosurgery. 1968 Apr 1;28(4):357-64.
16. Suzuki J, Iwabuchi T. Surgical removal of pineal tumors (pinealomas and teratomas): Experience in a series of 19 cases. Journal of Neurosurgery. 1965 Dec 1;23(6):565-71.
17. Packer RJ, Sutton LN, Rosenstock JG, Rorke LB, Bilaniuk LT, Zimmerman RA, Littman PA, Bruce DA, Schut L. Pineal region tumors of childhood. Pediatrics. 1984 Jul;74(1):97-102.
18. Dandy WE. Operative experience in cases of pineal tumor. Arch Surg 1936;33:19–46.
19. Russell OW, Sachs E. Pinealoma. A clinicopathologic study of seven cases with a review of the literature. The Journal of Nervous and Mental Disease. 1944 Jun 1;99(6):974.
20. Hoffman HJ, Otsubo H, Hendrick EB, Humphreys RP, Drake JM, Becker LE, Greenberg M, Jenkin D. Intracranial germ-cell tumors in children. Journal of neurosurgery. 1991 Apr 1;74(4):545-51.
21. Tate M, Sughrue ME, Rutkowski MJ, Kane AJ, Aranda D, McClinton L, McClinton L, Barani IJ, Parsa AT. The long-term postsurgical prognosis of patients with pineoblastoma. Cancer. 2012 Jan 1;118(1):173-9.
22. Cho BK, Wang KC, Nam DH, Kim DG, Jung HW, Kim HJ, Han DH, Choi KS. Pineal tumors: experience with 48 cases over 10 years. Child's Nervous System. 1998 Mar;14(1):53-8.
23. Al-Hussaini M, Sultan I, Abuirmileh N, Jaradat I, Qaddoumi I. Pineal gland tumors: experience from the SEER database. Journal of neuro-oncology. 2009 Sep;94(3):351-8.
24. Kumar P, Tatke M, Sharma A, Singh D. Histological analysis of lesions of the pineal region: a retrospective study of 12 years. Pathology-Research and Practice. 2006 Feb 15;202(2):85-92.
25. Villà S, Miller RC, Krengli M, Abusaris H, Baumert BG, Servagi-Vernat S, Igdem S, Lucas A, Boluda S, Mirimanoff RO. Primary pineal tumors: outcome and prognostic factors—a study from the Rare Cancer Network (RCN). Clinical and Translational Oncology. 2012 Nov;14(11):827-34.
26. Clark AJ, Sughrue ME, Ivan ME, Aranda D, Rutkowski MJ, Kane AJ, Chang S, Parsa AT. Factors influencing overall survival rates for patients with pineocytoma. Journal of neuro-oncology. 2010 Nov;100(2):255-60.
27. Stein BM. The infratentorial supracerebellar approach to pineal lesions. Journal of neurosurgery. 1971 Aug 1;35(2):197-202.
28. Stoiber EM, Schaible B, Herfarth K, Schulz-Ertner D, Huber PE, Debus J, Oertel S. Long term outcome of adolescent and adult patients with pineal parenchymal tumors treated with fractionated radiotherapy between 1982 and 2003--a single institution's experience. Radiation Oncology. 2010 Dec;5(1):1-7.
29. Schild SE, Scheithauer BW, Schomberg PJ, Hook CC, Kelly PJ, Frick L, Robinow JS, Buskirk SJ. Pineal parenchymal tumors: clinical, pathologic, and therapeutic aspects. Cancer. 1993 Aug 1;72(3):870-80.