



PREVALENCE OF HYDROCEPHALUS IN PAEDIATRIC MIDLINE POSTERIOR FOSSA TUMORS WITH ITS MANAGEMENT AND REVIEW OF LITERATURE: AN INSTITUTIONAL EXPERIENCE

Dr Harshit Agarwal	Mch Resident, Department of Neurosurgery, Gajra raja medical college, Gwalior, Madhya Pradesh (India).
Dr Anand Sharma*	Mch Neurosurgery, Assistant Professor, Department of Neurosurgery, Gajra raja medical college, Gwalior, Madhya Pradesh (India). *Corresponding Author
Dr Avinash Sharma	Professor, Department of Neurosurgery, Gajra raja medical college, Gwalior, Madhya Pradesh (India).
Dr Avdhesh Shukla	Professor, Department of Neurosurgery, Gajra raja medical college, Gwalior, Madhya Pradesh (India).
Dr SN Iyengar	Professor and Head, Department of neurosurgery, Gajra raja medical college, Gwalior, Madhya Pradesh (India).

ABSTRACT Hydrocephalus in paediatric patients with posterior fossa mass is a matter of debate, obstructive hydrocephalus is reported in 70% to 80% of children with posterior fossa tumours. Hydrocephalus is usually managed by the temporary external ventricular drain, endoscopic third ventriculostomy and permanent ventriculoperitoneal shunt. Treatment of hydrocephalus with a VP shunt before sub-occipital craniectomy was a safe procedure that significantly lowered the morbidity & mortality of subsequent tumour removal. VP shunt is still an option to treat hydrocephalus preoperatively. It can be done in those patients who are in a poor neurological state as a result of delayed diagnosis and advanced disease. This study was planned to evaluate the prevalence of hydrocephalus in paediatric midline posterior fossa tumours with its management and review of the literature.

KEYWORDS : Hydrocephalus, Midline posterior fossa tumours

INTRODUCTION:

Primary brain tumours are the most common solid tumours in the paediatric population, comprising 20% to 25% of all childhood tumours. About 60% to 70% of all pediatric brain tumours originate in the posterior fossa.^{1,4} Most common posterior fossa tumours of childhood are medulloblastomas, ependymomas, and astrocytomas. The posterior fossa is considered a critical region of the brain because it contains vital brainstem nuclei and masses in this region often cause hydrocephalus by blocking cerebrospinal (CSF) outflow pathways which results in signs and symptoms of raised intracranial pressure.⁵ Symptoms usually with headache, often worse in the morning followed by vomiting and eventually gait disturbance.

Hydrocephalus in paediatric patients with posterior fossa mass is a matter of debate, Obstructive hydrocephalus is reported in 70% to 80% of children with posterior fossa tumours and is frequently the cause of clinical deterioration at the time of diagnosis.^{6,7} CSF diversion in form of a ventriculoperitoneal shunt, temporary external ventricular shunt or endoscopic third ventriculostomy is indicated before definitive surgery. Placement of ventriculoperitoneal shunt has certain complications like infection, blockage, peritoneal seeding and rare complications such as upward herniation. 15% to 30% of patients require permanent CSF diversion following resection of the tumour.^{7,8} This study was planned to evaluate the prevalence of hydrocephalus in paediatric midline posterior fossa tumours with its management and review of the literature.

MATERIAL AND METHODS

Place And Period Of Study: This prospective study is carried out at the Department of Neurosurgery, JA group of hospitals, Gajra Raja Medical College, Gwalior.

Study Design – Prospective study

Study Population: Patients of midline posterior fossa mass up to 15 years of age, presented to neurosurgery department, JA groups of hospitals, G R Medical College, Gwalior over 24 months.

Inclusion Criteria: All children up to 15 years of age and CT/MRI brain suggestive of midline posterior fossa mass. Midline in the posterior fossa mass includes mass arising from vermis of the cerebellum, 4th ventricle, brain stem and mass arising from Cerebellar hemisphere approaching towards the midline (within 1 cm of the midline).

Study Procedure:

All patients up to the age of 15 years with midline posterior fossa mass presented to the neurosurgery department assessed demographically. Patients having hydrocephalus underwent CSF diversion procedures like VP Shunt as an emergency followed by definitive surgery. Patients not having hydrocephalus directly underwent definitive surgery as midline suboccipital craniectomy. Depending upon extension and infiltration of surrounding structures) with primary dural repair/augmentation duraplasty.

- After histopathological confirmation, adjuvant radiotherapy/ chemotherapy / both started depending upon HPE report.
- Patients with brain stem glioma directly underwent chemoradiotherapy.

RESULT:

Table 1: Age Wise Distribution Of Paediatric Midline Posterior Fossa Tumors:

Sr .No.	AGE	NUMBER	PERCENTAGE
1	0-5 Years	5	17 %
2	5-10 Years	8	28%
3	10-15Years	15	55%
4	Total	28	100

Among 28 patients 5 patients (17%) were in the age group of 0-5 years, and 8 patients (28%) were in the age group of 5-10 years. 15 patients (55%) were presented in the 10-15 years of age group (Table 1).

Table 2: Prevalence Of Hydrocephalus:

Sr No.	PRESENTATION	NUMBER	PERCENTAGE
1	Hydrocephalus	17	60%
2	No Hydrocephalus	11	40%
3	Total	28	

Among the 28 patients, 60 % of the patients had hydrocephalus at the time of admission (N=17, 60%), while 11 patients didn't have hydrocephalus (N=11, 40%) at the time of admission (Table 2).

Table 3: Age Distribution Of Patients With Hydrocephalus With Midline Posterior Fossa Tumors:

Sr.No.	AGE	NUMBER	PERCENTAGE
1	0-5 Years	5	29 %

2	5-10 Years	8	47%
3	10-15Years	4	23%
4	Total	17	100

Among the 17 patients with hydrocephalus, the most common age group was between 5-10 years (N=8, 47%) followed by 0-5years (N=5, 29%) and 10-15 years (N=4, 23%) (Table 3).

Table 4: Gender Wise Distribution Of Patients With Hydrocephalus:

Sr No.	Gender	No. of patients	Percentage
1	MALE	10	60%
2	FEMALE	7	40%
3	Total	17	100

Among 17 patients of hydrocephalus 10 patients were male while 7 patients were female with M: F ratio of 1.33:1 (Table 4).

Table 5: Clinical Presentation Of Patients With Hydrocephalus With Midline Posterior Fossa Tumors

Clinical feature	Percentage
Headache	75%
Vomiting	67%
Blurring of vision	7%
Cerebellar dysfunction	67%
Papilloedema	60%

Most of the patients (75 %) presented with Headache followed by Cerebellar dysfunction (67%), vomiting (67%). Papilloedema was present in 60% (Table 5).

Table 6: Management Of Hydrocephalus In Patients With Midline Posterior Fossa Tumors

Management	N	%
Ventriculoperitoneal shunt	17	100
Temporary EVD	0	0
Endoscopic Third ventriculostomy	0	0
Total	17	100

In the present study, all the patients with midline posterior fossa with hydrocephalus were treated with the permanent ventriculoperitoneal shunt (Table 6).

Table 7: Histology Of Paediatric Midline Posterior Fossa Tumors

Sr No.	Histopathology	No of patients	percentage
1	Medulloblastoma	15	54%
2	Ependymoma	4	15%
3	Pilocytic astrocytoma	4	15%
4	Epidermoid	1	3%
5	Dermoid	1	3%
6	Total	25	

In the present study, the most common histology was medulloblastoma (N=15, 54%) followed by Ependymoma and pilocytic astrocytoma (N=4, 15%) each (Table 7).

Table 8: Correlation Of Hydrocephalus With Histology:

Sr no.	Histopathology	N	Patient with Hydrocephalus	Percentage
1	Medulloblastoma	15	9	60%
2	Ependymoma	4	5	80%
3	Pilocytic astrocytoma	4	3	75%
4	Epidermoid	1	0	-
5	Dermoid	1	0	-
6	Total	25	17	

In the present study hydrocephalus was more common with Ependymoma (N=5,80%) followed by medulloblastoma (N=9, 60%) (Table 8).

Table 9: Shunt Related Complications:

Sr No.	Complication	No. of patients	percentage
1	Peritoneal metastasis	1	6%
2	Pseudocyst	1	6%
3	Shunt obstruction	2	12%
4	Total	4	

The most common complication with ventriculoperitoneal shunt was shunt obstruction presented in 12 % (N=2) followed by peritoneal metastasis and pseudocyst in 6% cases (Table 9).

DISCUSSION:

In the present study, the most common age group with midline posterior fossa tumour was 10-15 years, which is slightly different from previous studies. Sudha Iyengar et al⁷ (2016) reported that the most common age group is 6-8 years. Ahmed et al.¹⁰ (2007) reported most cases in the age group 5-9 years. The higher age group in this study may be due to late presentation to the health system despite having symptoms.

Prevalence of hydrocephalus: In the present study 60% of midline posterior fossa tumours develop hydrocephalus, which is slightly less than the previous study conducted by **Due-Tønnessen B.J et al 2007**¹² reported hydrocephalus in 79 % of posterior fossa masses **Culley et al (1994)**²² studied 64 children with posterior fossa mass and found that out of 64 patients,54 had hydrocephalus (84%). Due to the anatomic relationships of these tumours to cerebrospinal fluid (CSF) drainage pathways, hydrocephalus is common, usually occurring in 71–90% of children with posterior fossa tumours.¹¹

Management of hydrocephalus with midline posterior fossa tumours: Standard treatment for such type of hydrocephalus is still a matter of debate. In 1985, the American Society of Pediatric Neurosurgery commissioned a study to find out the best way to manage the dilemma of hydrocephalus secondary to posterior fossa tumours. Robert McLaurin concluded after obtaining the data from different hospitals of North America that there was no clear evidence of having any advantage to doing preoperative shunt insertion¹⁹. Hydrocephalus in these patients is usually managed by the temporary external ventricular drain, endoscopic third ventriculostomy and permanent ventriculoperitoneal shunt. The tumours in this region usually present earlier. Such tumours require prompt decision and quick action as most of the patients have associated hydrocephalus at presentation¹². Since in our setup most patients come from a rural background and presented in advanced stages & patient has been vomiting for a prolonged period, is dehydrated with electrolyte disturbance, poorly nourished, in poor general condition, blindness due to papilloedema and also obtunded, it may be necessary to correct the hydrocephalus first with controlled CSF drainage. This will render the patient in better general condition for major respective surgery.

External ventricular drain

Placement of a ventricular drain (EVD) may be hazardous in the presence of a large posterior fossa tumour, because upward coning and secondary haemorrhage in the brainstem may occur and cause further obtundation and blindness due to the compression of the posterior cerebral arteries at the tentorial edge¹³. The main issue related to EVD is CSF infection. It can be left safely, as long as needed, provided that meticulous care is taken for EVD insertion and nursing. EVD duration seems to not affect the incidence of infection¹⁴. The infection rate of 7% - 10% was noted per procedure¹⁴.

Endoscopic third ventriculostomy (ETV)

Neuroendoscopic treatment of hydrocephalus (endoscopic third ventriculostomy) is the most recent approach for management¹⁵. ETV creates communication between the ventricular system and subarachnoid spaces at the level of the floor of the third ventricle. Endoscopic third ventriculostomy in principle is efficacious, the procedure resolves the increased intracranial pressure before posterior fossa surgery in all cases, reduces the risk of postoperative hydrocephalus, eliminate the risk of upward herniation, and provides a burr hole that can be used as an emergency portal for CSF drainage.

The use of preoperative ETV is an effective, feasible and safe procedure¹⁶. It is helpful in immediate and long term control of hydrocephalus¹⁶. It eliminates the risk of CSF infection related to EVD and avoids the complications of VP shunt like infection and malfunction. In developing countries where the presentation is late, ETV is a recommendable option due to large ventricles¹⁶. However, ETV cannot always prevent postoperative hydrocephalus in all cases of posterior fossa tumours. The use of a postoperative VP shunt is the alternative¹⁶ limitation of ETV is availability and cost.

Ventriculoperitoneal Shunt

In 1963, Abraham and Chandy managed the hydrocephalus related to posterior fossa tumours with ventriculoatrial (VA) shunts¹⁷. This method

of managing hydrocephalus became popular. Many neurosurgeons adopted the policy of putting VP or VA shunts before the definitive posterior fossa tumour surgery to have a better operative field. It resulted in 100% of the patients having permanent shunt insertion or dependency¹. In 1985, the American Society of Pediatric Neurosurgery commissioned a study to find out the best way to manage the dilemma of hydrocephalus secondary to posterior fossa tumours. McLaurin concluded after obtaining the data from different hospitals of North America that there was no clear evidence of having any advantage to doing preoperative shunt insertion¹⁹. It is still a matter of discussion. VP shunt is still an option to treat hydrocephalus preoperatively. It can be done in those patients who are in a poor neurological state as a result of delayed diagnosis and advanced disease. Papilledema responds well to VP shunt. It significantly reduces the overall morbidity and mortality²⁰. Our study revealed 60 % of posterior fossa tumours had hydrocephalus which was managed by placement of ventriculo peritoneal shunt as an emergency procedure. It is not standard practice to perform a permanent CSF diversion procedure with placement of a ventriculoperitoneal shunt before tumour resection, because perhaps only one-third of patients will require permanent CSF diversion²¹

Although there are certain benefits of putting VP shunt : (1) Being able to delay resection surgery (2) Reducing the likelihood of needing external CSF diversion, which may carry the risk of infection. (3) Reducing the risk of post-resection CSF leak or pseudomeningocele²² (4) Reduced incidence of post-resection hydrocephalus. Nevertheless, several arguments have been raised against systematic pre-shunting, depending on the reported 10% rate of upward herniation in cases of posterior fossa tumours subjected to preliminary shunting²³, and risk of extraneural metastasis through the shunt tubing especially to the peritoneum²⁴ & Shunt related complications (shunt malfunction, infection, pseudocyst).

Symptoms of posterior fossa tumours usually result from either due to raised intracranial pressure (Hydrocephalus) or due to compression of vital structures. Symptoms and signs due to raised intracranial pressure were headache nausea vomiting and papilledema on fundus examination. The most common presenting symptom in our study was a headache in 75% of cases followed by vomiting and difficulty in walking 67 % each, which is almost similar to previous studies. Tabatabaei SM et al (2012) has reported Cerebellar symptoms were the most common cause of presentation (80.9%) followed by headaches (73.8%) and vomiting (38.1%). Visual problems were found in 7.14% of their patients probably due to late presentation. Lachi pavan Kumar et al (2015) found that vomiting was present in 90% of cases, followed by gait unsteadiness in (69%) followed by visual blurring in 42 % of cases.

Medulloblastoma was the most common histology presented in as 54% follow by astrocytoma and ependymoma 15 % each. Compared to the previous study Due-Tonnessen B.J et al 2007¹² showed 35% medulloblastoma, 38% had astrocytoma & 14% had ependymoma. In the present study hydrocephalus was more common with Ependymoma (N=5,80%) followed by medulloblastoma (N=9, 60%) (Table 8). The probable region is the propensity of Ependymoma to arise from the floor of the fourth ventricle and its elastic mature to get insulated into the foramen of luschka and magandie.

Shunt related complications

Shyamal C. Bir (2017) studied 48 patients with posterior fossa mass and VP shunt insertion, shunt failure was seen in 13 (27%) patients. In our study out of 28 patients, 17 patients underwent VP shunt surgery and 2 patients (11%) had shunt obstructions that were managed by shunt revision. One patient developed peritoneal metastasis which is a very rare phenomenon. Various abdominal complications of VP shunt surgery are, ascites, bowel and abdominal wall perforation and inguinal hernias. The abdominal pseudocyst is an uncommon manifestation of a VP shunt²⁵. Rainov N et al²⁵ (1994) presented a case series of 115 patients and found incidence of pseudocyst formation of after VP shunt surgery was 4.5%

In our study 1 patient (5.8%) developed pseudocyst after 9 months of follow up.

CONCLUSION:

Hydrocephalus in association with posterior cranial fossa tumours remain a matter of controversy, ether to perform CSF diversion prior to definitive surgery. Other authors refrain from ventricular decompression, as it may result in sudden decrease in ICP, and some

cases have been reported to develop intratumorally haematoma, which may have ominous complications & subsequent tumour excision difficult have raised questions of pre-operative shunting. Treatment of hydrocephalus with a VP shunt prior to sub-occipital craniectomy was a safe procedure that significantly lowered the morbidity & mortality of subsequent tumour removal. VP shunt is still an option to treat hydrocephalus preoperatively. It can be done in those patients who are in poor neurological state as a result of delayed diagnosis and advanced disease.

REFERENCES:

- Allen JC. Childhood Brain Tumors: Current Status of Clinical Trials in Newly Diagnosed and Recurrent Disease. *Ped Clin N Am.* 1985; 32:633-651
- Laurent JP, Cheek WR. Brain Tumors in Children. *J Pediatr Neurosci.* 1985; 1:15-32
- Section of Pediatric Neurosurgery of the American Association of Neurological Surgeons. *Pediatric Neurosurgery.* New York 1982
- Joona R, Hayward RD, Grant DN. Intracranial Neoplasms During the First Year of Life: Analysis of One Hundred Consecutive Cases. *Neurosurgery.* 1984; 14:31-41
- Kaderali Z, Lamberti-Pasculli M, Rutka J. The changing epidemiology of paediatric brain tumours: a review from the Hospital for Sick Children. *Childs Nerv Syst.* 2008;25:787-793.
- Meng L, Yuguang L, Shugan Z, Xingang L, Chengyuan W. Intraventricular epidermoids. *J Clin Neurosci.* 2006;13:428-30.
- Kachhara R, Bhattacharya RN, Radhakrishnan VV. Epidermoid cyst involving the brain stem. *Acta Neurochir (Wien)* 2000;142:97-100.
- Carson BS, Weingart JD, Guarnieri M, Fisher PG. Third ventricular choroid plexus papilloma with psychosis. Case report. *J Neurosurg* 1997;87:103-5.
- sudha iyenagr ,arti sharmAa Infratentorial pediatric brain tumors: An institutional Experience Volume : 6 | Issue : 12 | December : 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 79.96
- Ahmed N, Bhurgr Y, Sadiq S, et al. Pediatric brain tumours at a tertiary care hospital in Karachi. *Asian Pac J Cancer Prev* (2007), 8, 399-404.
- Sainte-Rose C, Cinalli G, Roux FE, Maixner R, Chumas PD, Mansour M, et al. Management of hydrocephalus in pediatric patients with posterior fossa tumors: The role of endoscopic third ventriculostomy. *J Neurosurg* 2001;95:791-7
- Due-Tonnessen, B.J. and Helseth, E. (2007) Management of Hydrocephalus in Children with Posterior Fossa Tumors: Role of Tumor Surgery. *Pediatric Neurosurgery*, 43, 92-96.
- Saira Alli, ... James T. Rutka, in *Principles of Neurological Surgery* (Fourth Edition), 2018
- Kulkarni, A.V., Drake, J.M. and Lamberti-Pasculli, M. (2001) Cerebrospinal Fluid Shunt Infection: A Prospective Study of Risk Factors. *Journal of Neurosurgery*, 94, 195-201.
- Rami AQ, Nidal K, Feras H, Muath A (2017) Neuroendoscopy Sway in the Treatment of Posterior Fossa Tumours associated Hydrocephalus in Children. *J Neurol Stroke* 6(3): 00203. DOI: 10.15406/jnsk.2017.06.00203
- Nagy, M.R. and El-Beltagy, M.A. (2012) Cerebrospinal Fluid Diversion Procedures in Children with Posterior Fossa Tumors: Ventriculoperitoneal Shunts versus Endoscopic Third Ventriculostomy versus External Ventricular Drainage. *Medical Journal of Cairo University*, 80, 249-254
- Abraham, J. and Chandy, J. (1963) Ventriculo-Atrial Shunt in the Management of Posterior Fossa Tumors Preliminary Report. *Journal of Neurosurgery*, 20, 252-253.
- Albright, L. and Reigel, D.H. (1977) Management of Hydrocephalus Secondary to Posterior Fossa Tumors. *Journal of Neurosurgery*, 46, 52-55.
- McLaurin, R.L. (1985) on the use of pre-operative shunting in management of Post. fossa tumours in children A Cooperative Study. In: Chapman, P.H., Ed., *Concepts in Pediatric Neurosurgery*, Vol. 6, Karger, New York, 1-5.
- Abou-Madawi, A. (2007) VP-Shunt Requirement in Patients with Posterior Fossa Tumors. *Suez Canal Univ Med J*, 10, 121-128
- Santos de Oliveira R, Barros Juca CE, Valera ET, et al. Hydrocephalus in posterior fossa tumors in children. Are there factors that determine a need for permanent cerebrospinal fluid diversion? *Childs Nerv Syst.* 2008;24:1397-1403.
- Culley, Deborah J., M.D.; Berger, Mitchell S., M.D.; Shaw, Dennis, M.D.; Geyer, Russell, M.D. March 1994, Volume 34, Number 3 402 An Analysis of Factors Determining the Need for Ventriculoperitoneal Shunts after Posterior Fossa Tumor Surgery in Children.
- Epstein F, Murali R (1978) Paediatric posterior fossa tumors: hazards of the "preoperative" shunt. *Neurosurgery* 3(3): 348-350.
- Yuh SJ, Vassilyadi M. Management of abdominal pseudocyst in shunt-dependent hydrocephalus. *Surg Neurol Int.* 2012;3:146.
- Frederick A. Boop, Jimmy Ming-Jung Chuang, in *Complications in Neurosurgery*, 2019