DOI: 10.21088/ijaa.2349.8471.12325.12

Indian Journal of Anesthesia and Analgesia
Volume 12 Number 3 July - September 2025

CASE REPORT

Cysto-peritoneal Shunt Placement in an Infant with Dandy Walker Syndrome: A Case Report

Vikram Singh[1], Kunal Kumar Sharma[2]

HOW TO CITE THIS ARTICLE:

Vikram Singh, Kunal Kumar Sharma. Cysto-peritoneal Shunt Placement in an Infant with Dandy Walker Syndrome: A Case Report. Ind J Anesth Analg. 2025; 12(3): 246-249.

ABSTRACT

Background: Dandy-Walker malformation (DWM) is a rare congenital anomaly of the posterior fossa characterized by hypoplasia or agenesis of the cerebellar vermis, cystic dilatation of the fourth ventricle, and enlargement of the posterior fossa. It is frequently associated with hydrocephalus and various neurological deficits. Early diagnosis and appropriate intervention are essential for optimizing outcomes.

Case Presentation: We present the case of a 1 year old female child who presented with progressive macrocephaly, irritability, vomiting, and delayed developmental milestones. Neuroimaging revealed classical features of Dandy-Walker malformation, including partial agenesis of the cerebellar vermis, a large retrocerebellar cyst communicating with the fourth ventricle, and dilated lateral and third ventricles indicative of associated hydrocephalus.

Management and Outcome: The patient underwent successful cystoperitoneal (CP) shunt placement to divert cerebrospinal fluid from the posterior fossa cyst to the peritoneal cavity. Postoperative recovery was uneventful, with gradual reduction in head circumference and improvement in feeding and irritability. Follow-up imaging demonstrated decompression of the cyst and reduced ventricular size. The patient continues to receive multidisciplinary follow-up, including neurodevelopmental monitoring and rehabilitation.

Conclusion: This case underscores the importance of timely neuroimaging in infants with progressive macrocephaly and developmental delay. Cystoperitoneal shunting remains an effective surgical option in managing hydrocephalus associated with Dandy-Walker malformation. Early intervention may contribute to favorable developmental outcomes, although long-term prognosis largely depends on the extent of associated anomalies and overall cerebral development

KEYWORDS

• Infant • Hydrocephalus • Shunt • Dandy Walker syndrome

AUTHOR'S AFFILIATION:

[1] Resident, Departemnt of Neurosurgery, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India.
[2] Assistant Professor, Departemnt of Neurosurgery, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India.

CORRESPONDING AUTHOR:

Kunal Kumar Sharma, Assistant Professor, Departemnt of Neurosurgery, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India.

E-mail: kunaal_kumar@yahoo.com

➢ **Received:** 08-05-2025 ➢ **Accepted:** 17-06-2025

INTRODUCTION

Dandy-Walker malformation (DWM) is a rare congenital brain malformation involving the cerebellum and the fluid-filled spaces around it (figure 1). It primarily affects the development of the cerebellar vermis, the fourth ventricle, and often leads to hydrocephalus. Key features associated with are: (i) Partial or complete absence of the cerebellar vermis; (ii) cystic enlargement of the fourth ventricle; and (iii) enlarged posterior fossa.[1] This pathology is often sporadic, but it can be associated with chromosomal abnormalities (trisomy 13, 18, 21) and antenatal factors during pregnancy.[2] The symptoms vary by severity with infants presenting with enlarged head, developmental delays, poor muscle tone and coordination, irritability, vomiting and seizures.[3] Undiagnosed adult patients may present with headaches, balance issues, or can be simply asymptomatic.

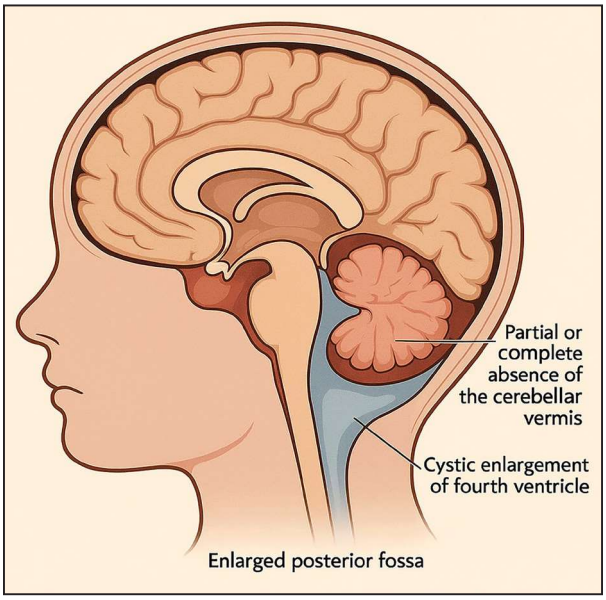

Figure 1: A schematic diagram of the intracranial pathology of Dandy Walker syndrome

Diagnosis is based upon prenatal sonography or MRI of brain. The treatment is symptom based. Cysto-peritoneal shunt placement has been associated with significant success in relieving the clinical picture. Supportive measures likephysical therapy, occupational therapy, special education is required by these children in early childhood and adoloscence.[4]

Case details

We describe our successful experience in managing such infant at our resource limited tertiary care centre. This case report adheres to the care (Case Report) guidelines of the Equator network. The patient was a 1 year old female infant presenting with hydrocephalus due to Dandy Walker Malformation. Based on the neuroimaging, placement of a Cysto-peritoneal shunt was decided by the neurosurgeons. The child did not have any venous acess, therefore she was planned for inhalational induction with sevoflurane. After priming the paediatric breathing circuit with the anesthetic gas mixture, the child was brought inside the operation theatre and facemask connected to the patient. The sevoflurane concentration was tapered down from 6 % to 2 % so that peripheral venous access could be taken in the limb. Once the access was taken. Once established, we gave preservative free lidocaine , fentanyl, propofol and atracurium for successful placement of endotracheal tube. The ETT size was calculated as per Cole's formula and after laryngoscopy using a Miller's blade, it was fixed at 12 cm mark with confirmation of bilateral equal air entry. The infant was maintained on pressure controlled ventilation using the O_2: Air: Sevoflurane anesthetic admixture. Intermittent bolus of fentanyl was given for intraoperative analgesia every hour. The intraoperative fluid management was done using a mixture of balanced salt solution (90 ml) with dextrose 5% (10 ml) as per the modified Holliday Segar formula.

The neurosurgeons sterilized and draped the head, neck and abdomen. For CP shunt, there are two incision sites-one cranial (occipital or suboccipital) and one abdominal (typically right lower quadrant). To expose the cranium a burr hole was made near the posterior fossa. Thereafter, dura was opened carefully. A catheter was introduced into the cyst cavity under direct visualization. CSF backflow confirmed the correct placement. Thereafter, a long subcutaneous tunnel was created from the cranial site to the abdomen using a tunneling device or long forceps. The distal end of the cyst catheter was tunneled through this path. Thereafter a small transverse incision was made in the right lower quadrant of abdomen. The peritoneum was opened. The distal end of the cyst catheter was connected to the peritoneal catheter which was inserted into the peritoneal cavity. The system (cyst catheter ± distal tubing) was connected securely and the connections were placed in a small subcutaneous pocket to avoid kinking or disconnection. Now the dura was closed,

followed by closure of scalp and abdominal wounds in layers.

After completion of the surgery and application of the skin sutures, the minimum alveolar concentration (MAC) of sevoflurane was washed off by increasing the net oxygen flow. Thereafter, the infant was reversed from muscle relaxation & extubated. She was crying and consolable after removal of the ETT. Her GCS was 15 with no limb deficits post-operatively.

DISCUSSION

Anesthetic induction agents do not cause significant effect on ICP or cerebral oxygenation.[5] Certain anaesthetics, like dexmedetomidine are even neuro-protective due to their anti-inflammatory action.[6] Dexmedetomidine even has better post-operative neurocognitive outcomes compared to other anaesthetics.[7] Nevertheless, anaesthetic management needs to be individualized to the patient needs as per the co-existing systemic disorders.

Cystoperitoneal (CP) shunt placement is an effective treatment for managing hydrocephalus and posterior fossa cysts associated with Dandy-Walker malformation (DWM). However, its effectiveness depends on individual factors like the size of the cyst, degree of hydrocephalus, and associated brain anomalies. CP shunts are generally effective in reducing symptoms such as increased head circumference, vomiting, and irritability by diverting excess cerebrospinal fluid (CSF) from the cystic areas to the peritoneal cavity.[8]In most patients, CP shunts alleviate raised intracranial pressure (ICP) symptoms (e.g., irritability, vomiting, enlarged head).[9]

It is believed that early intervention with a shunt can result in improved neuro developmental outcomes if placed prior to formation of hydrocephalus. CP shunts can lead to significant reduction in the size of posterior fossa cysts and associated ventricular dilation, contributing to improved neurological function.[10] Post-shunt MRI often confirms this.

However there are caveats. The first issue is thatpatients often become reliant on the shunt for CSF diversion, necessitating long-term monitoring and potential revisions. Complicationslikeshuntmalfunction,infection, over-drainage, and need for revision surgery mark the clinical course for such patients. Therefore frequent follow-up is required. While CP shunts address hydrocephalus, they do not correct the underlying cerebellar malformations, and some neurological deficits may persist. Thelong-term prognosis is closely tied to associated anomalies and the degree of cerebellar and cortical development, rather than to the success of the shunt alone.[11]

In some cases, a combination of cystoperitoneal and ventriculoperitoneal shunts may be employed to address both cystic and ventricular components of hydrocephalus.[12] However, the choice of shunting strategy should be individualized based on the patient's specific anatomical and clinical considerations.

Posterior Reversible Encephalopathy Syndrome (PRES) can sometimes co-exist with hydrocephalus. It can also be confused with each other due to overlapping symptoms like altered mental status, seizures, and visual disturbances. However, there are certain hallmark features of PRES which can delineate the differential diagnosis. In paediatric PRES there is**failure of cerebral autoregulation** and **endothelial dysfunction**, resulting in vasogenic oedema, in **posterior white matter regions** (parieto-occipital lobes), evident as hyperintense signals on T2 / FLAIR MRI.[13]

CONCLUSION

CP shunt placement can be an effective component of the surgical management of Dandy-Walker malformation, particularly in alleviating hydrocephalus-related symptoms. However, due to potential complications and the complexity of the condition, a multidisciplinary approach involving neurosurgeons andneuroanesthetists is essential for optimizing patient outcomes.

Sources of Funding: None

Conflicts of Interest: None

REFERENCES

1. Maria B.L., Bozorgmanesh A., Kimmel K.N., Theriaque D., Quisling R.G. Quantitative assessment of brainstem development in Joubert syndrome and Dandy-Walker syndrome. *J Child Neurol.* 2001; 16(10): 751-758.
2. Elsayed LEO, AlHarbi N.A., Alqarni A.M., Eltayeb H.H.E., Mostafa N.M.M., Abdulrahim

M.M., Zaid H.I.B., Alanzi L.M., Ababtain S.A., Aldulaijan K., Aloyouni S.Y., Othman MAK, Alkheilewi M.A., Binduraihem A.M., Alrukban H.A., Ahmed H.Y., AlRadini F.A., Alahdal H.M., Mushiba A.M., Alzaher O.A. Chromosome 16 p. 11. 2 microdeletion syndrome with microcephaly and Dandy-Walker malformation spectrum: expanding the known phenotype. *Hum Genomics*. 2024;18(1):95.

3. Imataka G., Yamanouchi H., Arisaka O. Dandy-Walker syndrome and chromosomal abnormalities. *CongenitAnom (Kyoto)*. 2007; 47(4): 113-118.
4. Bolduc M.E., Limperopoulos C. Neuro developmental outcomes in children with cerebellar malformations: A systematic review. *Dev Med Child Neurol*. 2009; 51(4): 256-267.
5. Sharma K.K., Surve R.M., Reddy K.R.M., Christopher R., Chakrabarti D., Pandarisamy S., Palakuzhiyil S.V., Kamath S. Impact of anesthetic induction with etomidate, thiopentone, and propofol on regional cerebral oxygenation: An observational study in patients with traumatic brain injury. *Journal of Anaesthesiology Clinical Pharmacology* 2025; 41(1): 90-97.
6. Sharma K.K., Chauhan B. Integrated dexmeditomidine-sevoflurane algorithm for anesthetic induction - A viable asset for neurosurgery. *SurgNeurol Int.* 2024; 15: 455.
7. Sharma K.K., Sharma S., Takkar V., Devi M, Krishnaswami S. Neurocognition after Electroencephalography Guided Anesthetic Induction with Dexmeditomidine in Neurosurgical Patients: A Case Series.*J. Neuroscience and Neurological Surgery.* 2025; 17(3): 1-6.
8. Bindal A.K., Storrs B.B., McLone D.G. Management of the Dandy-Walker syndrome. Pediatr Neurosurg. 1990-1991; 16(3): 163-169.
9. de Brito Henriques J.G., Henriques K.S., Filho G.P., Fonseca L.F., Cardoso F., Da Silva M.C. Bobble-head doll syndrome associated with Dandy-Walker syndrome. Case report. *J Neurosurg*. 2007; 107 (3 Suppl): 248-250.
10. Marinov M., Gabrovsky S., Undjian S. The Dandy-Walker syndrome: diagnostic and surgical considerations. Br J Neurosurg. 1991; 5(5): 475-483
11. Kalidasan V., Carroll T., Allcutt D., Fitzgerald R.J. The Dandy-Walker syndrome--a 10-year experience of its management and outcome. *Eur J PediatrSurg*. 1995; 5 (Suppl 1): 16-18.
12. Gabbita, Abhirama Chandra; Raju, Subodh. Management of Complex Hydrocephalus. *Neurology India*. 2021; 69 (Suppl 2): pp. S350-S356.
13. Surve, R.M., Sharma K.K., Sharma P. PRESenting a Challenge: Posterior Reversible Encephalopathy Syndrome in Pediatric Patients with Guillain-Barré Syndrome: A Case Series and Review of Literature. *Pediatric Neurology* 2024; 156: 162 - 169.