

Anaesthetic management for ventriculoperitoneal shunt insertion in an infant with Dandy–Walker Syndrome

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Dandy–Walker Syndrome (DWS) is a rare congenital brain anomaly affecting the cerebellum and the fourth ventricle. The chief components of the syndrome include cystic dilatation of the fourth ventricle and agenesis or hypoplasia of the cerebellar vermis. These abnormalities are typically associated with hydrocephalus. Patients often present in infancy for cerebrospinal fluid shunt procedures. Anaesthetic concerns include those related to other frequently associated congenital abnormalities. Airway management requires particular attention. The limited literature on this subject suggests that these patients require postoperative intensive care admission. This is not always possible in the resource-limited environment. This case report describes the successful anaesthetic management of an infant with Dandy–Walker Syndrome without postoperative intensive care admission.

Keywords: Dandy–Walker, neuroanaesthesia, paediatric anaesthesia

Introduction

Dandy–Walker Syndrome (DWS) is a rare congenital brain anomaly affecting the cerebellum and the fourth ventricle. It has a reported incidence of 1: 25 000–1: 30 000 with a slight female predominance.¹ The chief components of the syndrome include cystic dilatation of the fourth ventricle and agenesis or hypoplasia of the cerebellar vermis. These abnormalities are typically associated with hydrocephalus.² Patients often present in infancy for cerebrospinal fluid shunt procedures. Anaesthetic management of patients with DWS may prove challenging due to the frequent association of abnormalities in other systems. Airway management may also prove to be difficult.³ We describe a case of a nine-month-old infant presenting for elective ventriculoperitoneal shunt insertion. Written consent was obtained from the patient's mother to publish a case report and ethics approval was obtained from the Human Research Ethics Committee.

Case report

A nine-month-old female infant presented with a four-month history of progressive hydrocephalus secondary to a Dandy–Walker malformation. She was planned for an elective insertion of a ventriculoperitoneal shunt. She had no significant past medical or surgical history and was not on any chronic medication.

There was no history of seizures, irritability or vomiting. No feeding difficulties were reported and she had well-coordinated swallowing with no fatigue on feeding. There was no history of aspiration or recurrent respiratory infections; neither was there a history of apnoeas or breathing abnormalities. On examination the infant weighed 11.9 kg and was playful with good environmental interaction. She moved all limbs equally and showed no evidence of cranial nerve palsies. There were no abnormalities of eye movement or nystagmus. Tone was appropriate with good muscle bulk. The patient had delayed developmental milestones as she was unable to sit independently

due to poor head control. She was able to pull to a standing position but was unable to crawl. Her head circumference was 54 cm, which was greater than the 97th centile for age indicating macrocephaly. Both anterior and posterior fontanelles were enlarged and full, but not bulging.

No predictors of a difficult airway were noted. No facial defects were noted and the oral cavity was normal with no evidence of cleft lip or palate or micrognathia. No dysmorphic features indicating the presence of other syndromes were noted. The cardiac, respiratory and abdominal examinations were normal. Of particular note, there were no murmurs, palpable heart sounds or thrills.

The patient's preoperative haemoglobin was 13.1 g/dL. Renal function tests were normal.

After routine monitors were placed on the patient, induction of anaesthesia was achieved with sevoflurane and oxygen. The patient's position was optimised with the use of rolls of towels to elevate the body to prevent hyperflexion of the neck. After induction the airway was easy to manage and spontaneous ventilation continued without obstruction. Ventilation was supported manually to prevent hypercarbia. Several unsuccessful attempts were made to obtain venous access. Eventually the decision was made to secure the airway after topicalising it with lignocaine spray. Laryngoscopy was easy and a Cormack and Lehane grade I view of the larynx was achieved. The patient was intubated with a size 4 oral endotracheal tube. Vascular access was eventually obtained via a scalp vein opposite to the operative side. Cefazolin prophylaxis was administered prior to skin incision. A total of 30 µg of alfentanil and 20 mg of propofol were titrated to facilitate ventilation. Pressure-controlled ventilation was used with peak pressures of 14 cmH₂O at a rate of 28–32 bpm to maintain normocapnia. Anaesthesia was maintained with sevoflurane (2–3%) with supplemental boluses of alfentanil (2–5



Figure 1: Sagittal CT brain showing large posterior fossa cyst.

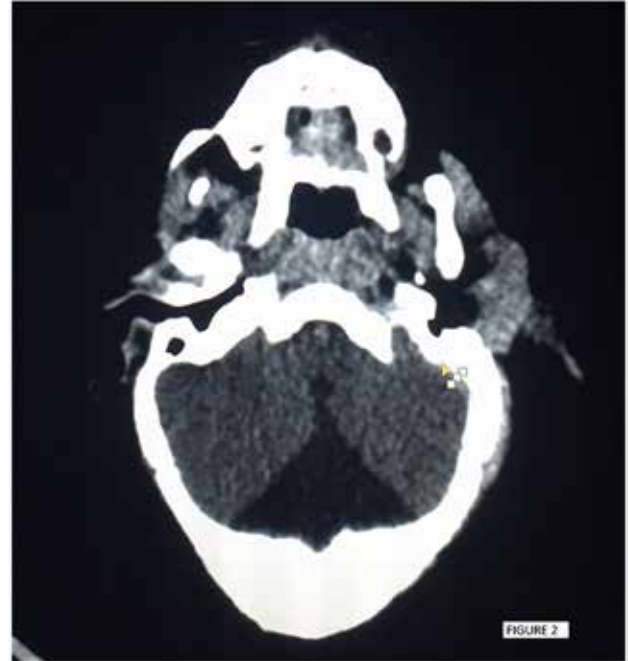


Figure 2: Axial CT brain showing large posterior fossa cyst extending into fourth ventricle producing a 'keyhole' appearance.

µg/kg) during periods of stimulation. Local anaesthetic infiltration of the skin with bupivacaine was performed by the surgeon.

The patient was haemodynamically stable throughout surgery. Despite the use of a forced air warmer and protective plastic covers, her temperature dropped to 34.4° C. This is not uncommon in cases of shunt surgery due to the large surface area that is exposed. Surgery was completed with no complications. The patient was rewarmed to normothermia. She began good respiratory efforts immediately after termination of mechanical ventilation and was extubated once fully awake.

After an uneventful stay in the recovery room she was discharged to the general ward since there are no paediatric high-care facilities at our hospital. An apnoea monitor was used overnight; however, the patient did not experience any episodes of apnoea or respiratory complications. She was discharged home on the second postoperative day.

Discussion

The term 'Dandy–Walker Syndrome' was first used by Benda in 1954 to describe this group of congenital posterior fossa abnormalities.⁴ Whilst DWS has been discussed extensively in paediatric and radiological literature, relatively little has been published about it in anaesthesia-related literature. This probably relates to its rare incidence. Despite this, anaesthetists should be aware of the syndrome and its associations since up to 80% of cases will develop hydrocephalus requiring anaesthesia and surgery.¹ Additionally, these patients may require anaesthesia for diagnostic CT or MRI scans.

The aetiology of DWS is heterogeneous and it may result from chromosomal disorders or environmental factors. In addition a single gene error has been implicated in cases where it occurs as part of other syndromes like Warburg Syndrome or Meckel–Gruber Syndrome.⁵ Dandy–Walker malformations may present as part of PHACES syndrome (posterior fossa abnormalities of the brain, haemangiomas of the face and scalp, arterial abnormalities, cardiac defects, eye anomalies and sternal abnormalities). There is also evidence that heterozygous

deletions of ZIC1 and ZIC4 genes may play a role in the development of DWS.⁶

The classic features of DWS are described by the triad of cystic dilatation of the fourth ventricle, a hypoplastic or aplastic cerebellar vermis and hydrocephalus.⁷ The majority of cases will present in the first year of life with features of hydrocephalus, often with developmental delay.⁸ A significant proportion of patients with DWS have other neurological abnormalities, the most common being agenesis of the corpus callosum. Sawaya and McLaurin suggest that associated abnormalities are suggestive of a poor prognosis.⁹ In their series, almost all cases that died were found to have associated abnormalities.

Of particular concern is the possibility of coexisting airway and cardiac abnormalities. Ecker *et al.* found that structural heart defects were the most common extra-cranial abnormality in foetuses with DWS.¹⁰ Atrial septal defects, ventricular septal defects, patent ductus arteriosus and valvular abnormalities have all been described in association with DWS.¹ Anaesthetists should therefore have a high index of suspicion for the presence of cardiac abnormalities during the preoperative assessment. If the history and clinical assessment are suggestive of a possible cardiac abnormality, a preoperative paediatric cardiology review should be sought to rule out coexisting cardiac abnormalities. In the absence of findings on the clinical assessment that are suggestive of a cardiac abnormality (i.e. no history of feeding difficulties, no recurrent chest infections, no murmur or cyanosis) we feel that the formal cardiology assessment may be deferred, especially in a resource-limited environment.

Airway difficulties experienced whilst anaesthetising these patients may be encountered due to the presence of a cleft palate, macroglossia or micrognathia.^{8,11} Laryngeal mask airways (LMAs) have been used successfully as rescue airway devices.¹² Additionally, the dolichocephalic shape of the head with a large occiput may make positioning difficult and prone to over-flexing, resulting in poor visualisation on direct laryngoscopy. A pillow or



Figure 3: Axial CT brain showing hypoplasia of the cerebellar vermis.

soft roll can be positioned under the body of the patient to elevate it in line with the head, thereby improving the view.

Sevoflurane is the agent of choice for inhalational induction in patients presenting for neurosurgical procedures.¹³ Evidence suggests that it causes less cerebrovascular vasodilatation than isoflurane, whilst CO₂ reactivity and autoregulation are preserved below 1.5 MAC.¹³

Maintenance of adequate mean arterial pressure is important in patients with hydrocephalus in order to maintain an adequate cerebral perfusion pressure. An assessment of the patient's fluid status preoperatively as well as a review of any co-morbidities (especially cardiac) should direct the anaesthetist in choosing either a fluid bolus or vasopressors/inotropes to manage hypotension.

Ventilation should be carefully controlled in order to prevent hypercarbia, which would cause a further rise in intracranial pressure. Muscle relaxants may be used to facilitate intubation; however, potent short-acting opioids like alfentanil or remifentanyl may also be used for this purpose without significantly raising intracranial pressure.¹⁴ In the case described above, we opted to avoid muscle relaxants and planned to use alfentanil to facilitate intubation and blunt the haemodynamic response to intubation. However, obtaining intravenous access proved difficult and we proceeded with intubation following sevoflurane induction and topical application of lignocaine spray to the vocal cords, which was successful.

Whilst Ewart and Oh suggest that return to spontaneous ventilation may be delayed, our patient demonstrated a brisk return to adequate spontaneous ventilation.¹⁵ In addition, they consider postoperative intensive care unit (ICU) admission to be

essential, stating a risk for apnoea. In the resource-limited environment, however, the limited supply of ICU beds limits the number of patients that may be operated on, leading to long waiting lists to the detriment of patients with time-sensitive conditions. In view of this as well as the fact that our patient had no preoperative risk factors for apnoea or any history of breathing abnormalities, we opted to manage the patient in the general ward with an apnoea monitor. In our experience we have found mothers to be the most vigilant caregivers in the postoperative period and as such they are counselled about monitoring the infant's breathing and about how to react in the event of an apnoea alarm.

Involvement of respiratory centres in the brainstem may result in irregular breathing. Krieger *et al.* reported apneustic breathing and cluster-type breathing possibly due to a pontine lesion in their case report.¹⁶ If the anaesthetist suspects that breathing might be abnormal after a general anaesthetic, especially against a background of preoperative respiratory abnormalities, then a high-care or ICU admission may be necessary postoperatively. Mayhew *et al.* described vocal cord paralysis, possibly due to vagus nerve traction, that prevented extubation of an infant postoperatively.¹⁷ In view of this the anaesthetist should be vigilant for post-extubation stridor.

In our patient, there were no indicators of respiratory center involvement preoperatively and no airway issues during or after the anaesthetic. Our anaesthetic management did not include the use of neuromuscular blockers or long-acting opioids therefore we did not feel that postoperative intensive care admission was warranted since the risk of respiratory complications was low. However, paediatric high-care admission would have been considered if our institution had these facilities. In the unlikely situation of complications arising that required re-intubation or that prevented extubation our plan would have been to ventilate the patient in theatre whilst arranging transfer to an appropriate centre. In those patients with DWS who have no other associated abnormalities and are in a good clinical condition it is our view that surgery of this type need not be delayed until a postoperative ICU bed is available. Indeed in the resource-limited environment this may lead to an unnecessarily long wait (often days to weeks) during which an otherwise well patient may deteriorate further.

In conclusion, patients with DWS may present several anaesthetic challenges when they present for diagnostic or therapeutic procedures. A thorough preoperative assessment should focus on excluding respiratory and cardiac abnormalities. The anaesthetist should be well prepared to manage airway difficulties in these patients. Postoperative disposition should be individualised based on the coexistence of other congenital abnormalities. In resource-limited settings, however, patients without additional abnormalities may be managed in a general paediatric ward with appropriate monitoring as we have shown in our case.

Conflict of interest/financial disclosure – None.

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