ANESTHETIC MANAGEMENT FOR A PATIENT
WITH DANDY-WALKER SYNDROME

by

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**Abstract**

One in 30,000 infants develops a congenital disease of the central nervous system (CNS), Dandy-Walker Syndrome.¹ This disease consists of anatomical malformations of the cerebellum, fourth ventricle, posterior fossa, and foramina which often results in hydrocephalus and upward displacement of the lateral sinuses.¹⁵ Such malformations commonly result in increase intracranial pressure (ICP) and anatomical distortions of the face and skull such as cleft lip/palate, high-arched palate, retrognathia (receded jaw), and poor dentition.¹ Therefore, the anesthetist must prepare for a difficult airway, take measures intraoperatively to control ICP, and attentively monitor the patient post operatively.¹³⁻⁵
Case Report

A 40 year-old, 157 cm, 58 kg caucasian female presented for a laparoscopic excision of abdominal lesions. Past medical history significant for Dandy-Walker syndrome with hydrocephalus and ventriculoperitoneal (VP) shunt placement at 9 months old, VP shunt revision at age 4, 21, and 35, seizures as an infant, seasonal allergies, and depression. Previous anesthesia resulted in no adverse events. The patient presented with a Mallampati class II airway, small mouth opening, recessed chin, and a thyromental distance of 4 cm, but full range of motion of the neck. The patient’s medications included singulair 10 mg once a day, bupropion 100 mg twice a day, and a multivitamin once a day. She confirmed no allergies to drugs. Additionally, the patient reported no signs of increased ICP currently.

Treatment included a 20 gauge peripheral intravenous (IV) catheter placed in the left hand and midazolam 2 mg IV in the preoperative area. Application of standard monitors took place after the patient arrived in the operating room. Patient received pre-oxygenation with 100% O2 for five minutes. Normal vital signs justified giving induction agents starting with fentanyl 150 mcg IV, lidocaine 60 mg IV, and propofol 150 mg IV. After absence of eyelash reflex confirmed, patient’s eyes taped closed, and a size 8 oral airway inserted. Easy mask ventilation allowed for rocuronium 35 mg IV administration. After confirmation of neuromuscular relaxation with no twitches by train of four monitor, insertion of a Miller 2 laryngoscope blade provided a Cormack-Lehane grade II view of the vocal cords. Patient’s trachea easily intubated with a 7.0 endotracheal tube. Positive end tidal carbon dioxide (ETCO₂) waveform on the monitor and bilateral breath sounds used to confirm placement of endotracheal tube and mechanical ventilation began.
Isofurane 1.3% inspired concentration and 2 L/min oxygen used to maintain anesthesia with fentanyl 50 mcg IV boluses for pain control titrated to heart rate and blood pressure. Neuromuscular blockade assessed by train of four monitor. Forced warm air machine applied. Normal saline used for fluid administration. Ondansetron 4 mg IV and dexamethasone 8 mg IV given for nausea and vomiting prophylaxis. ETCO₂ maintained at 30-35 mm Hg. At the end of the case, neuromuscular blockade reversal agent neostigmine 3 mg IV and anticholinergic agent glycopyrrolate 0.6 mg IV given after four of four twitches confirmed with train of four monitor. Mechanical ventilation changed to pressure support, respirations decreased to 4 breaths per minute, and patient began to initiate spontaneous breathing. Mechanical ventilation turned off, and the patient maintained adequate tidal volumes and respiratory rate with confirmatory sustained tetanus with train of four monitor. Volatile agent turned off, oral airway re-inserted, patient suctioned, and when the patient opened her eyes to command, endotracheal tube removed. Oxygen mask applied as patient continued to breath spontaneously at a regular rate with adequate tidal volumes. Simple oxygen mask applied at 6 L/min O₂ and patient taken to the postoperative recovery unit. Patient had an uneventful recovery from anesthesia and therefore discharged from the hospital later that day.

Discussion
Although cases of patients who first present with signs and symptoms of Dandy-Walker syndrome when they are adolescences or adults exist, most patients present with Dandy-Walker syndrome when they are infants.⁴⁻⁵ Anesthetists must be able to care for these patients with acutely diagnosed Dandy-Walker syndrome as well as after VP shunt placement. All Dandy-Walker syndrome patients, regardless of treatment for hydrocephalus, retain high probability for
craniofacial abnormalities such as micrognathia, macrocephalus, cleft lip/palate, high-arched palate, retrognathia, anteriorly placed larynx, and poor dentition.\textsuperscript{1-2,4} Potentially difficult airways require anesthetists prepare the proper equipment.\textsuperscript{6} In the case study presented, the patient’s small mouth opening and recessed chin signified potential for a difficult airway.\textsuperscript{6} Availability of difficult airway algorithm equipment at the bedside, such as a GlideScope (Verathon Inc., Bothell, WA) and endotracheal tube introducer (Bougie) (SunMed Healthcare, Largo, Florida), may avoid complications and therefore part of the meticulous preparation for the patient in the case study.\textsuperscript{6} Also, placement of an oral airway occurred before the attempted bag-mask ventilation in the case study. Paralytics administered only after confirmation of easy bag-mask ventilation ensured avoidance of a cannot intubate/ventilate situation in the case study.\textsuperscript{6}

An obstruction of the fourth ventricle results in hydrocephalus and increased ICP in patients with Dandy-Walker syndrome.\textsuperscript{1-5} Volatile anesthesia agents vasodilate cerebral vasculature and thus increase ICP.\textsuperscript{6} Although the patient in the case study did not present with signs of increased ICP, most likely due to a functional VP shunt, cerebral spinal fluid volume and ICP maintenance measures occurred throughout the case. Isoflurane minimally affects cerebral blood flow and facilitates cerebral spinal fluid absorption and therefore chosen as the volatile agent in the case study.\textsuperscript{6-7} A case study by Jang et al reported the use of propofol and remifentanil for induction and maintenance to reduce ICP while maintaining general anesthesia in a patient with hydrocephalus induced increased ICP.\textsuperscript{4,7} The case study presented included propofol and fentanyl in the induction sequence to maintain ICP. In addition to these measures, requirements exists for ETCO\textsubscript{2} maintenance between 30-35 mm Hg to reduce increases in ICP.\textsuperscript{4,6-7} In the case study, the patient’s ETCO\textsubscript{2} remained less than 45 mm Hg even during emergence.
In addition to the fourth ventricle and cerebellum abnormalities, partial or complete absence of the corpus callosum commonly exists in Dandy-Walker Syndrome thus increasing the likelihood of apnea and respiratory failure.\textsuperscript{1,3,5} In an article by Kusumoto and Shinozuka, successful extubation of the patient after a dental extraction occurred, but the patient later needed reintubation due to hypoxia and pneumonia.\textsuperscript{1} This article recommends monitoring Dandy-Walker syndrome patients in the intensive care unit postoperatively.\textsuperscript{1} Not only are these patients at risk for postoperative decompensation, but they are also at risk for sudden and unexpected death due to the CNS malformations.\textsuperscript{2} Precautions taken in the case study to confirm adequate ventilation and oxygenation included train of four monitoring, confirmation of adequate reversal of muscle relaxation, and obtainment of spontaneous breathing tidal volumes between 400ml-500ml by the patient before removal of endotracheal tube. Monitoring of the patient in the case study included 2 hours in the post-operative care unit and then another hour in the recovery room before discharged. Diagnosis and treatment with a VP shunt as an infant allowed for discharge that same day and removed the requirement to stay overnight for observation.

Due to the increased likelihood of craniofacial abnormalities, increased ICP, and postoperative ventilation complications associated with Dandy-Walker syndrome, requisites exists for a thorough preoperative assessment and precautious anesthesia plan of care.\textsuperscript{1,4} Regardless of the age of diagnosis, the anesthesia plan includes a thorough airway evaluation, difficult airway preparation, diligent monitoring of muscle relaxation, and attentive post operative care monitoring.\textsuperscript{4}
References


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