orphanesthesia

Anesthesia recommendations for

Dandy-Walker syndrome

Disease name: Dandy-Walker syndrome

ICD 10: Q03.1 (Atresia of foramina of Magendie and Luschka)

ORPHAcode: 166 (Dandy-Walker malformation)

Synonyms: Dandy-Walker malformation, Dandy-Walker anomaly, Dandy-Walker complex, Dandy-Walker variant

Brief disease summary: Dandy-Walker syndrome (DWS) is an uncommon congenital disorder of the posterior cranial fossa with important implications for perioperative anesthetic and neurosurgical management. The syndrome is anatomically defined by cystic dilatation of the fourth ventricle, partial or complete agenesis of the cerebellar vermis, and enlargement of the posterior fossa, typically accompanied by hydrocephalus and varying degrees of neurological impairment. The reported incidence is approximately 1 in 30,000 live births [1,2]. From an anesthesiologic perspective, patients with DWS frequently present with a constellation of difficult airway anatomy, altered levels of consciousness, elevated intracranial pressure (ICP), and an increased risk of postoperative respiratory complications.

Hydrocephalus represents the dominant neurological manifestation, affecting approximately 70-90% of patients and often necessitating early neurosurgical intervention due to progressive intracranial hypertension and its detrimental impact on neurodevelopment. Additional intracranial anomalies contribute to clinical heterogeneity; encephalocele is reported in approximately 16% of cases, while abnormalities of the corpus callosum (ranging from hypoplasia to complete agenesis) are present in nearly 30% of patients [2]. Beyond the central nervous system, DWS is frequently associated with extracranial congenital anomalies, most notably involving the cardiovascular and craniofacial systems. Non-cyanotic congenital heart defects, such as atrial and ventricular septal defects, as well as complex lesions including tetralogy of Fallot, have been described. Craniofacial dysmorphisms and musculoskeletal abnormalities (including scoliosis), and association with syringomyelia, further emphasize the systemic nature of the syndrome and underscore the need for comprehensive multidisciplinary assessment [3,4].

Clinically, DWS commonly manifests with signs of increased ICP, including progressive macrocephaly, headache, nausea, and vomiting, in combination with cerebellar and brainstem-related dysfunction. These features may include impaired motor coordination, abnormal respiratory patterns, reduced gag reflex, feeding difficulties, poor nutritional status, and global neurodevelopmental delay.

Diagnosis may be incorrect; if uncertainty exists, the diagnosis should be re-evaluated.

Every patient is unique; individual circumstances must always guide clinical care.

Medicine is in progress; new clinical knowledge may not be yet reflected in this recommendation.

Recommendations are not rules or laws; they provide a framework to support clinical decision-making. Although this recommendation has passed a structured review process, it does not meet the formal criteria of a guideline.

Translations may not always reflect the most recent updates of the English version.

Find more information on the disease, its centers of reference and patient organizations on Orphanet: www.orpha.net

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Functional difficult airway factors: Elevated ICP and brainstem involvement, reduced level of consciousness, increased risk of aspiration due to brainstem involvement reduced gag reflexes, non-cyanotic congenital heart disease, and scoliosis. Anatomical difficult airway factors: Craniofacial dysmorphism, macrocephaly, micrognathia, and spinal deformities such as scoliosis. Key anesthetic considerations: Premedication should be tailored to the patient's level of consciousness and available monitoring facilities. Measures to minimize aspiration risk are essential. A difficult airway management strategy should be anticipated and prepared in advance. Routine use of muscle relaxants and maintenance with volatile anesthetics should be avoided when possible. When neuromuscular blockade is indicated, anesthetic management should favor agents with a predictable and short duration of action, such as cisatracurium, or drugs allowing prompt pharmacological reversal, such as rocuronium with sugammadex; continuous quantitative neuromuscular monitoring using train-of-four (TOF) is essential. Total intravenous anesthesia (TIVA) is generally preferred. Careful postoperative monitoring is required due to the risk of respiratory failure. An opioid-sparing analgesic strategy is recommended. Antibiotic prophylaxis targeting Staphylococcus species, typically with cefazolin, is recommended. Vancomycin should not be used routinely as first-line prophylaxis and should be reserved for specific circumstances, such as known colonization or infection with methicillin-resistant Staphylococcus species or according to local antimicrobial resistance patterns.
B	**BLOOD PRODUCTS (COAGULATION)**	There are no specific recommendations regarding blood product transfusion or anticoagulation; decisions should be made on a case-by-case basis, guided by the patient's clinical condition and the planned surgical approach.
C	**CIRCULATION**	Age-adjusted normal pediatric hemodynamic values. Possible presence of an underlying congenital heart anomaly. In the presence of increased ICP, it should be noted that cerebral autoregulation in pediatric patients is typically reported within a mean arterial pressure range of approximately 20-60 mmHg.

D	**DRUGS**	Avoid deep premedication, volatile anesthetics, and routine use of muscle relaxants. TIVA is preferred. Employ an opioid-sparing strategy for postoperative analgesia.
E	**EQUIPMENT**	Difficult airway equipment should include age-appropriate endotracheal tubes, supraglottic airway devices (LMA and intubating LMA), a bougie, pediatric video laryngoscope, pediatric fiberoptic bronchoscope, and a pediatric surgical airway management kit. If central venous cannulation is required, femoral access may be preferred, as subclavian or internal jugular approaches can impair venous return and potentially increase ICP due to partial obstruction of venous flow.

Additional disease information

DWS is, in most cases, identified, diagnosed, and managed during the prenatal, neonatal, or pediatric periods. Nevertheless, several case reports describe diagnoses made in adulthood. In these reports, adult patients are often asymptomatic, with the condition discovered incidentally during routine examinations or following hospital admission for unrelated medical reasons or mild, nonspecific symptoms. Reported scenarios include imaging performed after motor vehicle accidents, evaluation of unexplained headaches, or routine neuroimaging following minor trauma, such as a fall while playing tennis [5-8]. Rare cases of adult DWS presenting with visual disturbances or symptoms mimicking myasthenia gravis have also been reported [9,10].

Once the diagnosis is established, neurosurgical consultation typically recommends clinical and radiological follow-up to monitor for disease progression and to determine the need for intervention. In asymptomatic patients, follow-up may therefore be appropriate depending on the individual clinical status.

Typical surgery and procedures

Symptomatic hydrocephalus remains the primary indication for surgical intervention, and ventriculoperitoneal shunt placement is the standard neurosurgical treatment. In some cases, communication between the supratentorial and infratentorial spaces via the aqueduct

is disrupted, necessitating additional neurosurgical procedures such as the placement of an additional infratentorial catheter, or endoscopic or open fenestration. Patients with DWS may also undergo other types of surgery (e.g., reconstructive and general surgery, orthopedics, cardiac surgery) under general anesthesia [11-13].

Type of anesthesia

The literature reports that general anesthesia is the predominant anesthetic approach. Only one case report describes a neonate with DWS undergoing umbilical hernia repair under caudal anesthesia [14].

Necessary additional preoperative testing (beside standard care)

The perioperative challenges associated with DWS arise from both the underlying neuroanatomical abnormalities and their systemic consequences. Anesthetic management must anticipate elevated ICP, a high likelihood of difficult airway management often exacerbated by craniofacial dysmorphism, and an increased risk of postoperative respiratory compromise related to brainstem involvement. In pediatric patients, these challenges are further compounded by age-specific physiological considerations and difficulties in establishing and maintaining reliable venous access. Accordingly, meticulous preoperative evaluation and coordinated multidisciplinary planning are essential to optimize perioperative safety and neurological outcome [15,16].

Particular preparation for airway management

Preoperative airway assessment is therefore a critical component of anesthetic planning in patients with DWS, particularly in the presence of craniofacial or neurodevelopmental abnormalities. Macrocephaly, micrognathia, macroglossia, and cervical spine anomalies may significantly increase the risk of difficult mask ventilation and tracheal intubation, necessitating readiness for advanced airway techniques [15-19].

Particular preparation for transfusion or administration of blood products

There are no established guidelines for the perioperative use of blood products in patients with DWS.

Particular preparation for anticoagulation

Perioperative anticoagulation in patients with DWS should be individualized due to the increased risk of intracranial hemorrhage from hydrocephalus, posterior fossa cysts, or ventriculoperitoneal shunts. Preoperative imaging should assess ventricular size, cystic structures, and shunt status, and anticoagulant therapy carefully reviewed, with bridging or temporary interruption as needed. During procedures, the minimal effective anticoagulant dose should be used if therapy cannot be stopped, with close coagulation monitoring. Postoperatively, anticoagulation should resume only after neurosurgical clearance, with vigilant neurologic observation. Multidisciplinary coordination among anesthesiology, neurosurgery, and hematology is essential to balance bleeding and thromboembolic risks.

Particular precautions for positioning, transportation and mobilization

Patients with DWS necessitate meticulous perioperative management regarding positioning, transportation, and mobilization due to the increased risk of intracranial hypertension, hydrocephalus, and brainstem dysfunction. Intraoperatively, the head should be maintained in a neutral, midline alignment with mild elevation to optimize cerebral venous outflow, while avoiding extremes of cervical flexion, extension, or rotation; all pressure points should be adequately padded, particularly in the presence of hypotonia or musculoskeletal deformities. During patient transfer, careful attention should be paid to preserving head and cervical alignment and maintaining hemodynamic stability, with gentle handling to prevent abrupt

accelerations or decelerations. Special precautions are warranted in patients with ventriculoperitoneal shunts to prevent kinking, traction, or external compression of the catheter. Postoperatively, mobilization should be progressive and assisted as indicated, with avoidance of rapid postural changes and continuous monitoring for neurologic signs such as headache, nausea, dizziness, or altered level of consciousness. Adherence to these anesthetic and neurosurgical principles is critical to minimizing fluctuations in ICP and reducing the risk of neurologic compromise in patients with DWS.

Interactions of chronic disease and anesthesia medications

Although the fundamental principles of pediatric neurosurgical anesthesia parallel those applied in adult practice, congenital neuroanatomical abnormalities introduce distinct complexities. Key anesthetic priorities include continuous assessment and control of ICP, secure airway management in anatomically challenging patients, reliable venous access, maintenance of age-appropriate mean arterial pressure to preserve cerebral perfusion, and strict avoidance of hypoxemia and hypercapnia, both of which may exacerbate intracranial hypertension [16].

Anesthetic procedure

Preoperative evaluation includes careful assessment of the level of consciousness, signs of raised ICP, baseline neurological status, hydration, and associated congenital conditions, particularly cardiac anomalies, to optimize perioperative safety. Premedication should be tailored to the patient's neurological status and avoided in children with impaired consciousness.

Airway management and vascular access can be challenging. Inhalational induction may facilitate venous access in uncooperative children, whereas reduced consciousness necessitates rapid sequence induction (RSI) to minimize the risk of aspiration. Central venous access is reserved for cases with difficult peripheral access or anticipated major fluid or blood requirements, with femoral access often preferred. Ultrasound guidance improves both success and safety. Meticulous airway management is essential, as avoidance of hypoxia, hypercapnia and aspiration is critical for maintaining stable ICP.

Standard monitoring is usually sufficient, with invasive monitoring and neuromonitoring added according to the patient's condition and surgical complexity. Anesthetic maintenance is preferably achieved with TIVA, avoiding volatile agents and minimizing the use of muscle relaxants to reduce the risk of prolonged neuromuscular blockade.

Emergence and extubation require careful planning, as pediatric neurosurgical patients have an increased risk of respiratory complications and delayed extubation. Postoperative respiratory failure remains a major concern. Central nervous system abnormalities, including corpus callosum agenesis, pontine hypoplasia, and distortion of the medullary respiratory centers, may impair central respiratory drive and airway protective reflexes, increasing the risk of hypoventilation or airway obstruction after extubation [16]. Laryngospasm has been reported to occur more frequently and with greater severity in patients with brainstem pathology [16,18].

To reduce respiratory risk, the use of TIVA with short-acting agents, avoidance of muscle relaxants when feasible, and TOF monitoring when neuromuscular blockers are required are strongly recommended [20]. This approach facilitates rapid emergence, timely extubation, and preservation of spontaneous ventilation.

Effective postoperative pain management is central to recovery. Opioids may be used with close monitoring, while multimodal analgesia including paracetamol, non-steroidal anti-inflammatory drugs (NSAIDs), dexmedetomidine, and regional scalp blocks, can reduce opioid requirements and associated adverse effects.

Anesthetic considerations in DWS

Perioperative phase	Key considerations
Preoperative evaluation	– Assessment of level of consciousness – Evaluation for signs of raised ICP – Baseline neurological status documentation – Identification of associated congenital conditions, particularly cardiac anomalies – Premedication individualized to neurological status
Airway management and vascular access	– Inhalational induction to facilitate venous access in uncooperative children – RSI in patients with reduced consciousness to reduce aspiration risk – Femoral venous access often preferred – Use of ultrasound guidance to improve success and safety – Strict avoidance of hypoxia, hypercapnia, and aspiration
Intraoperative monitoring and maintenance	– Standard monitoring usually sufficient – Addition of invasive monitoring based on patient condition and surgical complexity – Use of neuromonitoring when indicated – Preference for TIVA – Avoidance of volatile anesthetic agents – Minimization of muscle relaxant use to reduce prolonged neuromuscular blockade
Emergence, extubation, and postoperative care	– Careful planning of emergence and extubation – Increased risk of respiratory complications, laryngospasm and delayed extubation – Central nervous system abnormalities may impair respiratory drive and airway reflexes – TOF monitoring when neuromuscular blockers are required – Preservation of spontaneous ventilation and timely extubation

Particular or additional monitoring

Muscle relaxants require appropriate reversal and objective confirmation of neuromuscular recovery to prevent postoperative residual curarization (PORC). Quantitative TOF monitoring provides a more reliable and objective assessment of neuromuscular recovery than clinical evaluation alone and can help guide reversal, reduce residual paralysis, and minimize associated respiratory complications [21-23]. Pediatric patients may be particularly vulnerable to delayed or unpredictable recovery because of age-related differences in the pharmacokinetics and pharmacodynamics of neuromuscular blocking agents. Therefore, quantitative neuromuscular monitoring should be considered even after a single dose of a muscle relaxant, with particular attention to younger children and those at increased risk of postoperative respiratory compromise [24].

Current ESAIC/ESPA recommendations emphasize that, whenever a neuromuscular blocking agent is used in children, quantitative neuromuscular monitoring should be performed to assess the depth of blockade and exclude residual neuromuscular block before extubation. Clinical assessment alone should not be relied upon to confirm adequate recovery. The guidelines recommend confirming a TOF ratio ≥0.9 before extubation and favor sugammadex over neostigmine for reversal of aminosteroid neuromuscular blockade. These recommendations highlight the importance of objective neuromuscular monitoring and documented recovery before extubation [25].

Possible complications

Postoperative respiratory failure, laryngospasm, and postoperative residual curarization represent major perioperative complications in patients with DWS, particularly in the presence of brainstem involvement and neuromuscular blockade [24].

Postoperative care

Effective postoperative analgesia is a critical component of care in neurosurgical patients with DWS. Opioid use should be judicious due to the risk of respiratory depression and altered mental status, while nonsteroidal anti-inflammatory drugs may be limited by bleeding risk. A multimodal analgesic approach incorporating regional techniques, paracetamol, dexmedetomidine, ketamine, NSAIDs, and scalp blocks can provide effective pain control while minimizing opioid exposure, reserving opioids for inadequate analgesia [26-29].

Disease-related acute problems and effect on anesthesia and recovery

TIVA using short-acting agents such as propofol and remifentanil may be advantageous in high-risk neurosurgical patients, particularly when combined with avoidance of muscle relaxants when clinically appropriate, to facilitate rapid recovery and minimize postoperative respiratory complications [30,32]. Evidence comparing TIVA with volatile anesthesia remains heterogeneous; therefore, the choice of anesthetic technique should be individualized according to patient risk factors, neurological status, and surgical requirements [30,31].

In neurosurgical practice, TIVA may also provide favorable conditions for intraoperative brain relaxation, predictable emergence, and reduced postoperative nausea and vomiting [32].

Large observational data further suggest that TIVA is associated with lower postoperative morbidity, although mortality outcomes appear similar to those with volatile anesthesia [33].

Ambulatory anesthesia

Day-case anesthesia is typically not recommended for patients with DWS due to the potential for significant perioperative complications.

Obstetrical anesthesia

There are no reported cases of patients with DWS undergoing obstetric surgical procedures under anesthesia. Several case reports in the literature have addressed DWS during pregnancy. No differences in treatment compared with other cases have been reported, with emphasis placed on the role of the fetal neurologist [34,35]. Different studies have focused on intrauterine genotypic diagnosis and the role of prenatal phenotypic ultrasound in achieving accurate in utero diagnosis [35]. Intrauterine intervention using decompressive techniques for hydrocephalus has also been reported as a prenatal treatment strategy [36].

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