orphanesthesia

Anesthesia recommendations for

Dyke-Davidoff-Masson syndrome

Disease name: Dyke-Davidoff-Masson syndrome

ICD 10: Q04.3

ORPHAcode: -

Synonyms: Cerebral hemiatrophy, Cerebral hemi-hypoplasia

Brief disease summary: Dyke-Davidoff-Masson syndrome (DDMS) is an extremely rare neurological condition that occurs more commonly in the pediatric population, with an increasing number of cases being reported in adults as well [1]. It is characterized by cerebral hemiatrophy or hypoplasia, resulting from either congenital or acquired insults to the developing brain [2]. It was described by three clinicians in 1933, Dyke, Davidoff and Masson, who reported patients with facial asymmetry, contralateral hemiparesis, seizures and intellectual disability with characteristic pneumatoencephalographic findings on skull X-ray [2]. There is a slight male predominance [1]. Adult cases have also been reported, though infrequently [3]. The main causes of this rare syndrome are either congenital (in utero vascular occlusion) or acquired (perinatal hypoxia, intracranial hemorrhage and infections). Brain imaging with computed tomography (CT) and magnetic resonance imaging (MRI) is required for diagnosis [4]. Published literature regarding anesthetic considerations for this syndrome is limited. Differential diagnoses include Sturge-Weber syndrome, Fishman syndrome, Basal cell germinoma, Silver-Russell syndrome, Linear nevus syndrome and Rasmussen encephalitis [5].

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

Emergency information

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Potentially difficult airway due to facial and skull asymmetry, seizure predisposition and intellectual disability. Spontaneous ventilation should be maintained until the airway is secured. General anesthesia is preferred. Neuraxial anesthesia may be limited by spinal deformities and poor patient cooperation.
B	**BLOOD PRODUCTS (COAGULATION)**	No inherent coagulation disorder. Coagulation profile should be ordered for patients on sodium valproate. No special transfusion requirements.
C	**CIRCULATION**	No typical cardiac abnormalities. Hemodynamics are usually stable. Individualized assessment is recommended.
D	**DRUGS**	Antiepileptic drugs should be continued perioperatively. Enzyme induction may increase anesthetic drug requirements. Seizure-provoking agents should be avoided.
E	**EQUIPMENT**	Difficult airway cart with fiberoptic bronchoscope/video laryngoscope. Portable ultrasound for vascular access and regional blocks. Equipment for neuromuscular and depth of anesthesia monitoring. Postoperative HDU/ICU monitoring.

Additional disease information

DDMS is characterized by cerebral hemiatrophy or hypoplasia, resulting from either congenital or acquired insults to the developing brain [2]. The condition typically presents with a classic triad of contralateral hemiparesis, recurrent seizures, and intellectual disability, often accompanied by facial asymmetry [2].

Radiological features include unilateral cerebral atrophy with compensatory calvarial thickening, hyperpneumatization of paranasal sinuses, and ipsilateral ventricular dilatation [6]. The severity of neurological deficits depends on the timing and extent of the cerebral insult.

Seizures are frequently refractory and require long-term antiepileptic therapy. Motor deficits may lead to functional impairment, musculoskeletal deformities, and increased susceptibility to trauma. Behavioral disturbances and psychiatric comorbidities may also be present in some patients [5].

There is no definitive cure, and management is primarily supportive, focusing on seizure control, physiotherapy, and rehabilitation. Surgical interventions such as hemispherectomy may be considered in selected cases with intractable epilepsy [1].

Major anesthetic concerns include seizure disorder, altered pharmacodynamics due to antiepileptic drugs, potential airway challenges due to craniofacial asymmetry, and difficulties related to patient cooperation [7].

Typical surgery and procedures

Patients with DDMS may present for both elective and emergency surgical procedures, most commonly in pediatric patients [1]. Owing to recurrent seizures and hemiparesis, these patients are prone to falls and frequently sustain orthopedic injuries. Surgeries may, therefore, be required for fractures, non-unions, contractures or, sometimes, for neurosurgical interventions. Rarely, a few patients may need to undergo hemispherectomy for the treatment of resistant seizures, with a reported success rate of 85% in carefully selected patients.

Type of anesthesia

There is limited literature regarding anesthetic considerations for patients with DDMS and anesthesia for these patients can be quite challenging due to neurological impairment and associated comorbidities. In the presence of intellectual disability, patient cooperation for regional anesthesia and intravenous cannulation may be difficult. The use of ultrasound guidance for vascular access is recommended to improve success rates. Spinal deformities can further complicate neuraxial block placement and effectiveness. Although both general and regional anesthesia may be administered with appropriate precautions, the preferred technique in these patients remains general anesthesia, with adjunct use of ultrasound-guided regional nerve blocks. Patients receiving anticonvulsants may have enzyme induction, resulting in altered responses to anesthetic drugs, including increased requirements of non-depolarizing neuromuscular blocking agents and opioids [8].

Necessary additional preoperative testing (beside standard care)

Neuroimaging [4], including skull radiographs, contrast-enhanced computed tomography (CECT), MRI of the brain, and electroencephalography (EEG), is essential in all cases. These scans often show atrophy or hypoplasia of one of the cerebral hemispheres. History regarding any neurological insult in the fetal or early childhood period, or maternal thrombophilia for congenital cases, should be elicited. Other causes of intellectual disability must be ruled out. A complete neurological assessment is necessary to ascertain the level of hemiparesis (grade of motor/sensory loss). An evaluation of the underlying cause of seizures should be performed, and adequate epilepsy management should be initiated preoperatively. Serum anticonvulsant levels may be measured in selected cases to ascertain adequate antiepileptic medication levels, and to rule out their toxicity. Sodium valproate can interfere with coagulation [9] and a coagulation screening should ideally be performed before surgeries at risk of bleeding. Special attention must be given to renal function tests and serum electrolytes.

Particular preparation for airway management

Difficult airway management may be encountered due to facial asymmetry, ipsilateral skull hypertrophy, and sinus enlargement [10]. Mask ventilation may be difficult owing to inadequate mask seal. Awake intubation may not be possible due to lack of patient cooperation. Inhalation anesthesia or intravenous sedation, with preservation of spontaneous ventilation, should be used for endotracheal intubation if a difficult airway is suspected. Aspiration prophylaxis [11] may be given in patients with recurrent or intractable seizures and intellectual disability. A difficult airway cart should be readily available at all times and standard difficult airway guidelines must be followed for airway management.

Particular preparation for transfusion or administration of blood products

There are no disease-specific recommendations regarding transfusion of blood products.

Particular preparation for anticoagulation

There are no specific recommendations for anticoagulation. Caution needs to be exercised in patients taking anticonvulsants and warfarin because of enzyme induction [8].

Particular precautions for positioning, transportation and mobilization

Perioperative positioning requires special attention as these patients may have intellectual disability and associated craniofacial and skeletal abnormalities [10]. Preexisting neurological deficits should be carefully documented. All pressure points must be adequately padded to prevent neurovascular injuries. Transportation and mobilization must be gentle and well-coordinated to prevent musculoskeletal injuries.

Interactions of chronic disease and anesthesia medications

There are no known disease-specific interactions with anesthetic drugs. However, the use of succinylcholine should be avoided in view of the presence of occult myopathy, where it can lead to rhabdomyolysis and hyperkalemia [12]. Moreover, the enzyme-inducing effect of anticonvulsant medications may necessitate anesthetic dose adjustments [8].

Anesthetic procedure

Standard general anesthesia followed by administration of regional nerve blocks (ultrasound-guided), whenever appropriate, is preferred in these patients. Adequate seizure control, aspiration prophylaxis, preparedness for difficult airway, securing reliable venous access, and ensuring optimal oxygenation and ventilation are the cornerstones of a successful outcome. Pediatric patients may display exaggerated separation anxiety and agitated behavior [12], and require adequate anxiolysis preoperatively. Extubation must also be cautious and performed only after ensuring adequate spontaneous tidal ventilation. Postoperatively these patients should be nursed in high-dependency units (HDU) with appropriate monitoring.

Particular or additional monitoring

All the standard ASA monitors must be used, especially capnography, which is essential to prevent hypercarbia-induced seizures. Peripheral neuromuscular monitoring and depth of anesthesia monitoring are recommended to prevent delayed reversal or awakening from anesthesia. Depth of anesthesia (Bispectral Index/Entropy) monitoring may also be useful in identifying intraoperative seizures, which may lead to a brisk increase in BIS/Entropy values [13]. However, this would require measurement of preoperative awake values, which can be quite challenging in these patients. Invasive hemodynamic monitoring may be indicated in patients with associated cardiopulmonary abnormalities. Temperature monitoring must be meticulous to prevent the development of hypothermia in these vulnerable patients.

Possible complications

Complications can be disease-, anesthesia- or procedure-related. Hypoxia or hypercarbia may precipitate seizures [14]. Some patients may develop emergence delirium or agitation and may require a prolonged hospital stay. The incidence of PONV (postoperative nausea and vomiting) may be increased, although evidence is limited. Nonetheless, pharmacologic prophylaxis for PONV should be administered to all patients.

Postoperative care

These patients must be monitored in a HDU in the postoperative period. Anticonvulsant medications must be continued, and patients should be closely monitored for seizures. Supplemental oxygen, adequate analgesia, hypothermia prevention, and psychological support must be ensured.

Disease-related acute problems and effect on anesthesia and recovery

There are no disease-related acute problems or any specific effect on anesthesia and recovery. However, delayed recovery from general anesthesia may occur due to prolonged effects of anesthetic agents, electrolyte disturbances, or a postictal state following perioperative seizures [15]. Neuromuscular monitoring is essential to ensure adequate reversal of muscle relaxation prior to extubation.

Ambulatory anesthesia

Ambulatory anesthesia is advisable only for short, minimally painful procedures in carefully selected patients. Standard perioperative monitoring and postoperative care protocols must be followed. Discharge should be permitted only in the presence of a competent caregiver [16]. In general, it is best to avoid day care surgeries [17] due to the risk of seizures, intellectual disability, and potential maxillofacial abnormalities that may complicate airway management and recovery.

Obstetrical anesthesia

Literature is sparse regarding obstetric anesthesia in patients with DDMS. Aspiration prophylaxis, continuation of pregnancy-safe anticonvulsants (levetiracetam/lamotrigine), adequate oxygenation, avoidance of supine-hypotension syndrome, and difficult airway preparedness must be ensured [18]. Administering regional anesthesia (central neuraxial block) may be extremely challenging due to lack of cooperation and musculoskeletal deformities. Monitoring antiepileptic drug levels and folic acid supplementation is important in this population. Appropriate antenatal counseling should be undertaken to establish rapport and facilitate patient cooperation for regional anesthesia, which is generally safer than general anesthesia in obstetric patients.

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