### Worry About
- Hydrocephalus with elevated ICP and possible seizures
- Pt's ability to cooperate and follow commands
- Aspiration risk
- Ventilation challenges because of craniofacial abnormalities
- Postanesthetic respiratory depression
- Multiorgan disease resulting in cardiac and urogenital abnormalities

### Overview
- Dandy-Walker complex represents a group of related congenital disorders of brain development, including Dandy-Walker malformation, mega cisterna, and Dandy-Walker variant.
- Includes congenital brain malformation involving a hypoplastic cerebellum with variable defects in formation of the cerebellar vermis, enlargement of the fourth ventricle, and cyst formation in the posterior fossa.
- Commonly associated conditions with variable severity include hydrocephalus, defects in corpus callosum formation, developmental delay, and abnormalities of the heart, urogenital tract, and bones. There may be associated developmental syndromes including PHACIES, spina bifida, and others, which may complicate management. Careful Hx and physical exam are required to identify comorbidities.
- ICP and seizure management are primary concerns.
- Rostral brain involvement may predispose pt to apnea following anesthetic.

### Etiology
- Believed to be the result of multifactorial gene mutations. TUBA1A has been identified as a major driver, resulting from mutation of tubulin transport proteins. Inheritance is mostly sporadic, with a small familial association.

### Usual Treatment
- Depends upon disease presentation. Hydrocephalus is often treated with ventriculoperitoneal shunt, medication for seizures, physical therapy for muscular involvement, occupational therapy, and education for learning disabilities.

### Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Craniofacial abnormality, macrocephaly, micrognathia, macroglossia, occipital meningocele, nystagmus | | | |
| CV | Varied cardiac abnormalities, including Tetralogy of Fallot | SOB, poor exercise tolerance, "Tet spells" | Cyanosis, heart murmur | CXR, ECG, angiography |
| RESP | Medullary control of respiratory center | | Apnea | |
| RENAL | Urogenital malformation | Urinary tract infections | | UA |
| CNS | Intracranial pressure, developmental delay, CN palsy | N/V, seizure | Palsy, altered mentation | CT |
| MS | Abnormal vertebrae, prominent occiput, frontal bossing, cleft palate, truncal ataxia, muscle spasticity | | Ataxia | CT |

**Key References:** National Institutes of Health: Genetic and rare diseases information center. <https://rarediseases.info.nih.gov/gard/6242/dandy-walker-complex/resources/1>, 2016 (Accessed 12.04.16.); Shweta M, Rao S, Ladi SD, et al.: Dandy Walker syndrome: case report, *Innov J Med Health Sci* 4(1):309–311, 2014.

### Perioperative Implications

**Preoperative Preparation**
- Identify organ involvement, aspiration risk, and anatomic defects.

**Monitoring**
- Standard monitoring
- Arterial line if cardiac dysfunction warrants

**Airway**
- Craniofacial abnormalities may compromise ventilation and intubation.
- Macrocephalus may be managed with a shoulder bag to improve positioning.
- Rapid sequence induction if aspiration risk exists.

**Induction**
- Avoid increased ICP with smooth induction, normocapnia, and muscle relaxants.
- Preop ventriculoperitoneal shunt may be needed before other surgeries.
- Succinylcholine may need to be avoided because of renal disease or elevated ICP.
- Cognitive impairment may render pt uncooperative.
- pt may have CV disease.

**Maintenance**
- Pt may have CV instability.
- Monitor for seizure activity; maintain normocapnia.

**Extubation**
- Anticipate challenges with reintubation.
- Pt may be at risk of apnea and delayed spontaneous ventilation due to diminished respiratory drive.

**Adjuvants**
- Shoulder bag, video laryngoscope, and fiberoptic laryngoscope

**Postoperative Period**
- Monitor respiratory status closely.
- Monitor for seizure activity; avoid increased ICP.

# De Morsier Syndrome

Ashley R. Valentine | Jeffrey R. Kirsch

### Risk
- For live births: 1:10,000; equal male to female prevalence
- Associated with younger maternal age
- May not be identified until later in life

### Perioperative Risks
- Reduced cortisol stress response in undiagnosed or untreated pts. Hormone tests may be normal in nonstress conditions.
- Treatment of one hormone deficiency (e.g., hypothyroidism, or hypothyroidism and adrenal insufficiency) may unmask another or others (e.g., adrenal insufficiency, DI).

### Worry About
- Unrecognized hypothalamic/pituitary axis deficiencies
- Neurocognitive disorders causing agitation, seizures, or confusion in periop period

### Overview
- Highly phenotypically variable disorder diagnosed when at least two of three features are present: ONH, midline/CNS neuroradiographic abnormalities (may include absence of the septum pellucidum), and/or hypothalamic/pituitary abnormalities.
- ONH is third most common cause of any vision impairment in children <3 y in USA.
- ONH associated with other neuro abnormalities (e.g., developmental delay, autistic spectrum disorder, epilepsy, disrupted circadian rhythm).
- Hypothalamic/pituitary hormone abnormalities can develop at any age and may include growth hormone deficiency (most common), hypothyroidism, ACTH deficiency, and DI (least common).
- Limb abnormalities (e.g., syndactyly) and MSK abnormalities (e.g., spastic quadriparesis, hypotonia) also may be present.

### Etiology
- Majority of cases are sporadic, and less than 1% have currently identifiable genetic mutation.
- Environmental risk factors may include antenatal drug/ETOH use and low socioeconomic status.
- Genetic mutations in HESX1, SOX2, SOX3, or OTX2 may be causal.
- See also Adrenal Insufficiency, Hypopituitarism, Hypothyroidism, and Seizure.

### Usual Treatment
- Pts followed at least every 6 mo for growth and development
- At least annual vision evaluation and treatment as indicated
- Endocrine function followed for life because hypothalamic/pituitary abnormalities can develop at any age
- Supportive services tailored to individual pt's needs (e.g., occupational, speech, developmental, and/or physical therapy; neuropsychology; ophthalmology)
- Genetic counseling for families with identifiable genetic mutation
- May need surgical correction of associated strabismus or orthopedic deformities

### Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Vision deficits, nystagmus, strabismus | History of abnormal vision, blindness, vision correction surgeries | HEENT exam | Evaluation by ophthalmologist |
| ENDO | Hypothalamus/pituitary abnormalities | Growth delay, precocious or delayed puberty, excessive thirst, excessive hunger, heat or cold intolerance, fatigue, constipation, weakness, history of treatment for known abnormalities | Vitals, puffy face, weakness | Endocrine hormones (TSH, $T_3/T_4$, GH, cortisol or cortisol stim test, glucose, IGF-1, IGFBP-3, LH, FSH |
| CNS | Cognitive delay, autistic spectrum disorder, behavioral disorder, epilepsy, sleep disorder, abnormal temperature regulation, hyperphagia or hypophagia, polydipsia | Stereotypical behaviors, not meeting developmental milestones, speech delay, seizures, abnormal sleep/wake cycles, temperature instability (unexplained fevers or frequent hospitalizations to rule out sepsis), insatiable appetite or food aversion, water-seeking behavior | General appearance, neuropsych exam | MRI, CT |
| MS | Weakness, spasms, motor delay, limb deformities | Not meeting developmental milestones, frequent falls, clumsiness | MSK exam<br>NEURO exam | X-ray, CT, and/or MRI |

**Key References:** Borchert M: Reappraisal of the optic nerve hypoplasia syndrome, *J Neuroophthalmol* 32(1):58–67, 2012; Sherlock DA, McNicol LR: Anaesthesia and septo-optic dysplasia: implications of missed diagnosis in the peri-operative period, *Anaesthesia* 42(12):1302–1305, 1987.

### Perioperative Implications

**Preoperative Preparation**
- Evaluation and treatment of endocrine abnormalities (especially thyroid, ACTH, GH and deficiencies)
- Consider ONH if neonatal history of jaundice, hypoglycemia, and nystagmus/visual deficits
- Consider premedication for pts with cognitive delay.

**Monitoring**
- Standard monitors
- Neuromuscular (TOF) for recovery from muscle relaxants

**Induction**
- Consider avoiding succinylcholine in pts with severe weakness or immobility.
- Consider avoiding etomidate in pts with untreated secondary adrenal insufficiency.

**Airway**
- No special difficulty

**Intraoperative Considerations**
- Intraoperative CBG if history of hypoglycemia.
- Consider stress-dose steroids if secondary adrenal insufficiency.

**Extubation**
- No special difficulty but may need support for agitated pts

**Postoperative Period**
- Consider stress-dose steroids for secondary adrenal insufficiency.
- May have increased risk of postop delirium given vision/cognitive impairment.

### Anticipated Problems or Concerns
- Abnormal temperature regulation may be difficult to differentiate from postop infection. Multidisciplinary supportive services may be needed postop.

# Deep Vein Thrombosis

Sophia T. Cisler | Lee A. Fleisher

### Risk
- Incidence in USA: 170,000–200,000 new cases; 90,000–100,000 recurrent cases.
- VTE is the third most frequent acute cardiovascular syndrome after MI and CVA.
- Half of all episodes are associated with recent surgery or hospitalization.
- VTE is recognized as the leading cause of preventable death in hospitalized pts.

### Perioperative Risks
- Modified Caprini risk model can be used to predict risk in general surgical pts.
- Without prophylaxis, DVT develops in close to 30% of general surgical pts.
- With chemical prophylaxis, risk can be reduced to 8% for general surgical pts.
- Incidence of fatal PE: 0.1 (general surgery)–5% (total knee replacement).

### Worry About
- Pulm embolism
  - Cardiac arrest, electromechanical dissociation
  - Increased A-a gradient, increased dead space, potentially leading to respiratory acidosis
- Increased bleeding risk, safety of regional anesthesia in anticoagulated pts
- Risks and benefits of discontinuing anticoagulation for surgery

### Overview
- Classic symptoms of DVT: swelling, pain, and erythema of the involved extremity.
- GA associated with increase in tissue factor, vWF, tissue plasminogen activator, resulting in hypercoagulable/hypofibrinolytic state.
- Dx.
  - Contrast venography (gold standard); requires IV contrast exposure; 2–3% incidence of inducing thrombosis.
  - Compression/duplex ultrasonography of femoral/popliteal veins has sens/spec of 97% in symptomatic pts (less sens for more distal [calf] veins).
  - IP, also more sensitive in proximal (90%) than distal.
  - D dimer has high negative predictive value useul to rule out VTE).
- See also Pulmonary Embolism.

### Etiology
- Pt-specific risk factors: age >40 y, immobility, obesity, malignancy, smoking, history of VTE, lower limb injury, inherited hypercoagulability
- Risk increased significantly by major surgery or critical illness
- Without prophylaxis, incidence is approximately 14% in gynecologic surgery, 22% in neurosurgery, 26% in abdominal surgery, and 45–60% in hip/knee surgery
- Risk decreased with regional anesthesia versus general, especially in LE orthopedic surgery

### Usual Treatment
- Anticoagulation (UF heparin, LMW heparin, warfarin, or direct oral anticoagulants such as factor IIa or Xa inhibitors)
- Thrombolytics
- Thrombectomy, catheter or open surgical
- IVC filter for PE prevention in high-risk pts or if anticoagulation is contraindicated

### Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| CV | | | Tachycardia, RV strain | ECG, TTE/ TEE |
| RESP | PE | Chest pain, dyspnea, hemoptysis | Tachypnea, wheezing, hypoxemia | $SpO_2$, ABGs, $ETCO_2$ |
| HEME | | | | PT, APTT, Plt, Hgb, D dimer |
| DERM | | Fever | Unilateral edema, erythema, warmth | |
| MS | | Limb pain | Tenderness, Homans sign | Ultrasound, venography |

**Key References:** Streiff MB, Agnelli G, Connors JM, et al: Guidance for the treatment of deep vein thrombosis and pulmonary embolism, *J Thromb Thrombolysis* 41(1):32–67, 2016; Krishnan KN: Deep vein thrombosis and pulmonary embolism—prevention, management, and anaesthetic considerations, *Indian J Anaesth* 54(1):8–17, 2010.