Perioperative Implications

- Review "required reconsideration" of the DNR orders.
- All changes to DNR status must be communicated to all members of the periop team and documented in the pt's medical record.
- Best if discussion of DNR orders can be done preop to develop a better pt-doctor relationship, avoid production pressure influences, and to allow time to contact all appropriate parties (surrogate, surgeon, primary care physician).
- This discussion should include what procedures are essential for the anesthetic and operation (e.g., intubation paralysis); iatrogenic arrest; and if the DNR order is modified, when and if it should be reinstated.
- The document for *Informed Consent for Anesthesia Care in The Patients with An Existing Do-Not-Resuscitate Order* created by The American Society of Anesthesiologists Committee on Ethics provides three resuscitation options during the periop period:
  - Full resuscitation.
  - Limited resuscitation: Procedure-directed, documents specific procedures the pt refuses.
  - Limited resuscitation: Goal-directed, allows resuscitation if the anesthesiologist and surgeon believe the adverse events are temporary and reversible. Allows resuscitation if the anesthesiologist and surgeon believe the resuscitation efforts support specified and documented goals of the pt.
- Consider consultation with an ethics expert if there is disagreement or concern about DNR orders and the surgery is not emergent.

Anticipated Problems/Concerns

- Anesthesiologists rarely have an established relationship with the DNR pt but must discuss and clarify resuscitation wishes.
- Aspects of anesthesia care (intubation, vasopressors, IV fluid therapy, transfusion, etc.) are resuscitative therapies.
- Medications used for anesthesia may cause cardiac depression, respiratory depression, and cardiac arrest.
- Anesthesiologists may be morally conflicted with the pt's desire for limited intervention. For a nonemergent case, the anesthesiologist can decide not to perform the anesthetic as long as there is another available physician and the change is not detrimental to the pt.

Double Aortic Arch

Anthony J. Clapcich

Risk

- Vascular rings account for <1% of cardiovascular malformations that require surgical correction. Double aortic arch is the most common form of complete ring that encircles both the trachea and the esophagus.
- Race/gender predilection: None.

Perioperative Risks

- Recurrent respiratory infections often aggravate chronic airway obstruction.
- Baseline dynamic tracheal compression can progress to complete airway obstruction upon induction and muscle relaxation.
- Persistent postop airway obstruction requiring prolonged mechanical ventilation and CPAP.

Worry About

- Esophageal obstruction: Dysphagia, choking, emesis, aspiration, FTT.
- Tracheal obstruction: Chronic cough, wheezing, barky-brassy cry, inspiratory/expiratory stridor; acute episodes of severe respiratory distress, apnea, cyanosis, and ALTE.
- Associated cardiac anomalies (10–20%): VSD, ASD, interrupted aortic arch, transposition of the great arteries, tetralogy of Fallot, truncus arteriosus, and complex univentricular lesions.
- Chromosome 22q11 deletion syndrome (20%): Genetic defect associated with syndromes, such as DiGeorge, velocardiofacial, CHARGE, and VACTERL; features include endocrine abnormalities (hypocalcemia, thyroid/parathyroid dysfunction, short stature), palatal and laryngotracheal abnormalities, developmental delay/neurologic abnormalities, renal tract malformations, thrombocytopenia, T-cell deficiencies, and autoimmune disorders.

Overview

- Vascular rings can be classified as complete or incomplete. Double aortic arch is the most common form of complete ring that encircles and compresses both the trachea and esophagus.
- Symptoms usually occur at birth or within the first 3 mo of life. The degree of tracheal and esophageal compression will dictate the severity of respiratory and GI perturbation.
- Initial work-up with CXR and upper GI can reveal tracheal deviation/narrowing and proximal esophageal distention/indentation. After the diagnosis is suspected, ECHO is used to examine arch anatomy and rule out other intracardiac anomalies. Both MRI and CT are very useful in further delineating vascular, airway, and GI anatomy. Cath is now reserved for assessing complex cardiac defects that require additional hemodynamic information. Bronchoscopy is often performed at the time of repair to evaluate the location, degree, and extent of airway obstruction, which may help to identify those pts at risk for postop respiratory compromise.

Etiology

- During normal human development, six branchial arches are sequentially formed and penetrated by six paired aortic arches that arise from the aortic sac and terminate in paired DA. These primitive arches largely regress (the fourth and sixth being the most persistent) and by the eighth week, the right DA largely involutes and forms the distal part of the right subclavian artery, leaving only the left DA to form the distal aortic arch and descending aorta. Failure of the right DA to involute results in a double aortic arch, whereby the ascending aortic arch divides into two arches, passes on each side of the trachea and esophagus, and joins posteriorly to form the descending aorta. The right carotid and subclavian arteries arise from the usually dominant, posterior right arch, whereas the left carotid and subclavian arteries arise from the smaller, anterior left arch.

Usual Treatment

- Medical therapy: None.
- Surgery: The goal is to relieve tracheal and esophageal compression by dividing the vascular ring and dissecting any fibrous bands. A thoracotomy is usually performed on the side ipsilateral to the minor arch. Right (posterior) arch is dominant in >75% of cases; thus left posterolateral thoracotomy is commonly used to expose the left (anterior) arch. Video-assisted thoracoscopic repair is also an effective option. Median sternotomy with cardiopulmonary bypass is reserved for cases that require concomitant repair of associated cardiac anomalies.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Chromosome 22q11 deletion features: Facial abnormalities Palatal abnormalities Velopharyngeal incompetence Congenital laryngeal web	Previous difficulties with anesthesia or intubation FTT Nasal regurgitation of formula; delayed speech/poor articulation (childhood) Noisy breathing, abnormal cry	Low set ears, short philtrum, hypertelorism, small mouth, small chin Cleft palate Hypernasal speech (childhood) Inspiratory/expiratory stridor, aphonia/weak high-pitched cry Hoarseness (childhood)	Flexible bronchoscopy Direct laryngoscopy/ bronchoscopy
CV	Depends on presence of associated cardiac anomalies (10–20% cases); None if *only* double aortic arch present	Cyanotic spells, CHF, dyspnea, diaphoresis, FTT	Murmur, cyanosis, four-limb noninvasive BP discrepancy, grunting, rales/wheezes, hepatosplenomegaly	Pulse oximeter, ECG ECHO Cardiac MRI Cardiac cath
RESP	Airway obstruction Recurrent respiratory infection	Dyspnea, apnea, intermittent cyanosis, ALTE Coughing, wheezing	Insp/expiratory stridor (± positional), hyper-extended head, brassy-barky cry, intercostal retractions, nasal flaring	CXR Bronchoscopy MRI CT
GI	Esophageal obstruction	Dysphagia, FTT		UGI Esophagoscopy

Key References: Licari A, Manca E, Rispoli GA, et al.: Congenital vascular rings: a clinical challenge for the pediatrician, *Pediatr Pulmonol* 50(5):511–524, 2015; Backer CL, Mongé MC, Russell HM, et al.: Reoperation after vascular ring repair, *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 17(1):48–55, 2014.

Perioperative Implications

Preoperative Preparation

- Oxygen therapy if decreased arterial oxygen saturation present
- Antibiotics for bronchopneumonia

Monitoring

- Bilateral upper extremity SpO_2 and Doppler probes are useful for assessing subclavian, carotid, and temporal pulses during temporary occlusion of the arch that is to be resected. Regional tissue oxygenation of the brain can be monitored via bilateral cerebral NIRS probes, which can reveal unilateral desaturation if carotid flow is compromised during arch manipulation and the patient's circle of Willis anatomy is not adequately providing collateral circulation.
- Potential for hemodynamic and respiratory instability warrant placement of arterial cath; presence of an aberrant subclavian artery may affect appropriate cath site.
- Large-bore IV access is essential; central venous line should be considered for pts with poor vascular access and those who require extensive repair on CPB.

Airway

- Dynamic and static airway obstruction likely; significant tracheal compression may require smaller ETT size than predicted.

Induction

- Inhalation induction without neuromuscular blockade until airway maintenance is documented by mask and/or ETT is placed distal to area of obstruction.
- Bronchoscopy during spontaneous ventilation allows for direct assessment of tracheal pathology and degree of dynamic airway collapse, thus identifying pts at risk for postop respiratory compromise.

Maintenance

- Balanced technique of narcotics and volatile agent is usually well tolerated.

Extubation

- Extubation at end of case if tracheomalacia and stenosis absent

Postoperative Period

- Good pain control essential for stable hemodynamics and avoidance of respiratory complications; IV opioids, rectal acetaminophen, intercostal nerve blocks, one-shot caudal, and caudal epidural cath have all been used with success.

Anticipated Problems/Concerns

- Despite surgical correction, persistent postop airway obstruction requiring prolonged mechanical ventilation and CPAP can occur secondary to edema, mucosal friability/reactivity, and long-segment tracheomalacia.

Down Syndrome

Stephanie Black

Risk

- Trisomy 21 is the most common autosomal aneuploidy; approximately 1:1000 live births.
- 80% of children with this condition survive beyond 1 y; average life expectancy 60 y.
- Increased incidence in mothers >35 y, but most are born to younger mothers, owing to higher fertility rates.
- Incidence decreased by elective termination of pregnancy from prenatal screening: high beta-hCG, low AFP, cell-free DNA, thickened nuchal fold, abnormal ductus venosus waveforms, absent nasal bone—cell-free DNA can now increase the sensitivity and specificity of these tests.

Perioperative Risks

- Airway obstruction
- Cardiac dysfunction due to CHD
- Cervical spine instability
- Immune and endocrinologic dysfunction

Worry About

- Airway obstruction:
  - Upper airway obstruction common immediately on induction of GA due to macroglossia, midface crowding, small mandible, short neck.
  - Subglottic stenosis in 20–25%; may cause postop stridor in children.
  - Obstructive sleep apnea in 30–50%. Central apnea also common.
    - Chronic hypoxemia may contribute to pulm Htn risk and increased opioid sensitivity.
- Congenital cardiac dysfunction:
  - 40% are born with CHD.
  - Most common: Complete atrioventricular canal defect (40%), VSD (25%).
  - Cyanotic CHD in 4% (usually tetralogy of Fallot).
  - Risk for pulm Htn because of pulm overcirculation.
  - May develop R-to-L shunting with profound hypoxemia.
  - Risk for paradoxical/systemic air emboli (coronary or cerebral vessels).
  - Bradycardia with inhalational induction with sevoflurane.
- Cervical spine instability:
  - Extension during intubation can cause neurologic symptoms (neck pain, arm pain, upper extremity weakness, torticollis) from atlanto-occipital instability.
- Generalized joint laxity; TMJ may sublux with jaw thrust.
- Endocrine: hypothyroidism (4–6% in children; 15–20% in adults), hypothermia.
- Immune dysregulation causes higher rates of certain cancers (ALL and AML) and respiratory infections.
- GI: Duodenal atresia in 4%; recurrent aspiration may cause pneumonia.
- Developmental delay:
  - May have fears of the unknown; can become physically resistant to entering OR.
  - Alzheimer disease and other mental illnesses (depression, psychosis) may coexist.

Overview

- Most common autosomal aneuploidy with an increasing life expectancy because of early interventions for multiple comorbidities
- Concerns for congenital cardiac disease, hypotonia, immune dysregulation, airway obstruction, recurrent pneumonia, oncologic predisposition, and GI disorders
- Physical exam findings: Midface hypoplasia, brachycephaly, epicanthal folds, simian crease, downward medial slant of eyes, high-arched palate, glossoproptosis, and murmur
- May require surgery for tympanostomy, strabismus, CHD repair, duodenal/esophageal atresia, marrow aspiration/biopsy, cervical spine fusion

Etiology

- Genetic: Trisomy 21

Usual Treatment

- Depends on penetrance and pathophysiology; may include the use of CPAP, thyroid hormone replacement, OT/PT

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Redundant tissue Midface hypoplasia Subglottic stenosis	Sleep apnea Intubation Hx Hearing deficit	Macroglossia Glossoproptosis "Down's facies"	Audiology Polysomnography
CV	CHD in 40% CAVC most common Tetralogy of Fallot in 4% Bradycardia on induction Risk for pulm Htn	Symptoms of CHF "Tet spells" Hx of CHD repair	Cyanosis Murmur Clubbing	ECHO ECG
ENDO	Hypothyroidism Metabolic syndrome	Thyroid hormone replacement Hypothermia	Obesity	Thyroid hormone levels
IMMUNE/ONC	Oncologic predisposition Immune dysregulation	Respiratory infections AML and ALL	Cough Lymphadenopathy	Auscultation Bone marrow biopsy/aspiration
MS	Hypotonia Subluxation of C1/C2	Upper motor neuron symptoms	Joint laxity	Cervical spine radiographs (controversy over whether these should be routine)

Key References: Arumugam A, Raja K, Venugopalan M, et al.: Down syndrome—a narrative review, *Clin Anat* 29(5):568–577, 2016; Maxwell LG, Goodwin SR, Mancuso TJ, et al.: Systemic disorders: down syndrome. In Davis PJ, Cladis FP, Motoyama EK, editors: *Smith's anesthesia for infants and children,* ed 8, Philadelphia, PA, 2011, Elsevier, pp 1172–1174.