Worry About

- Bradycardia
- PAH, RV failure, and shunt reversal
- Atrial arrhythmias resulting from atrial enlargement

Overview

- AV canal is associated with atrial and ventricular septal defects manifested by a variety of abnormal communications between the left and right heart structures.
- Categorized into atrial septal defects and partial or complete AV canal defect.
- Main hemodynamic problems include AV valve dysfunction, interatrial shunting, and interventricular shunting.
- L-to-R shunting results in RV or LV dysfunction or failure, frequent resp infections, and failure to thrive.
- Chronic L-to-R shunt causes increased pulmonary vascular resistance and shunt reversal (Eisenmenger syndrome), which may preclude surgical intervention.
- Diagnosis includes chest radiograph (enlarged heart), physical exam findings (murmur), prolonged electrocardiogram PR interval, and ECHO.

Etiology

- AV canal defects arise from abnormal endocardial cushion development between 4–5 wk gestational age.
- Failure of endocardial cushion fusion results in deficiencies in the interventricular septum that can form a common AV valve, common AV valve annulus, or interatrial communication.

Usual Treatment

- Medical management (before repair) directed to improve cardiac function and overall health (digitalis, diuresis, positive inotropic drugs, afterload reduction, adequate nutrition).
- Surgical management is definitive; includes repair of the septal defects and AV valves.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Feeding difficulties	Failure to thrive	Decreased weight/height for age	Compare with ideal weight/height
CV	CHF PAH	Fatigue, dyspnea, diaphoresis, coughing Dyspnea, tachypnea	Murmur, wheezing, rales, hepatosplenomegaly Increase in CHF Sx	CXR, TEE, cardiac cath TEE, cardiac cath
RESP	CHF, pneumonia	Dyspnea, tachypnea	Wheezing, rales	CXR
RENAL	Renal insufficiency			Cr, BUN
MS	Exercise intolerance			

Key References: Wenink AC, Zevallos JC: Developmental aspects of atrioventricular septal defects, *Int J Cardiol* 18(1):65–78, 1988; Bergin ML, Warnes CA, Tajik AJ, et al.: Partial atrioventricular canal defect: long-term follow-up after initial repair in patients ≥40 years old, *J Am Coll Cardiol* 25(5):1189–1194, 1995.

Perioperative Implications

Preoperative Preparation

- Midazolam (0.05–0.1 mg/kg) to reduce anxiety and facilitate cooperation.
- Anxiolytics not recommended for children <1 y of age.
- Use caution as anxiolytics may cause hypoventilation, hypercapnia, increased PVR, and shunt reversal.

Monitoring

- Standard ASA monitors; arterial and central venous cath
- CVP monitoring in the setting of PAH
- TEE if not contraindicated

Airway

- Anticipate difficulty when trisomy 21 is present

Induction

- Meticulous exclusion of air from IV tubing to avoid paradoxical air embolism.
- Inhalation induction time is minimally affected by L-to-R shunt but may be prolonged in R-to-L shunt.
- Choice of IV anesthetic for induction based on severity of heart failure.

Maintenance

- Decrease in afterload due to IV or volatile anesthetics may worsen R-to-L shunt.
- Adjuvant opioids to allow use of lower volatile anesthetic concentrations.

Extubation

- Extubation feasible in the operating for partial AV canal defects without heart failure or PAH.
- Airway obstruction or hypoventilation after extubation may increase PVR, requiring subsequent hyperventilation, increased FIO_2, or inhaled nitric oxide, or ECMO.

Adjuvants

- Positive inotropic drugs to enhance myocardial contractility
- Inhaled nitric oxide or prostaglandin I_2 to reduce PVR

Postoperative Period

- Closely monitor and reduce pulm artery pressures in pts with preop PAH.
- Reduced cardiac output may occur as a result of RV or LV dysfunction or LV outflow tract obstruction.
- May require temporary transvenous pacing or develop postop arrhythmias.

Anticipated Problems/Concerns

- Presence of arrhythmias, reduced cardiac output, moderate or severe mitral regurgitation, and elevated PVR is associated with greater risk of mortality

Endocarditis

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Risk

- Incidence: 3–10:100,000 population.
- Rheumatic heart disease is a key risk factor in lower-middle income countries.
- Valvular and cyanotic heart disease, DM, cancer, and IV drug use are risk factors in higher income countries.

Perioperative Risks

- Septic embolization to other organs (CNS, renal, and lung) is seen in 25–50% of pts.

Worry About

- Acute heart failure (valvular regurgitation or obstruction), stroke, and metastatic infection (i.e., epidural abscess/osteomyelitis)
- AV or bundle branch blocks due to infection extension

Overview

- IE is an infection of the heart, most commonly seen in pts with structural heart disease (i.e., CHD, mitral valve prolapse).
  - Acute infections commonly caused by *Staphylococcus aureus* and *S. epidermidis*.
  - Subacute infections commonly caused by *Streptococcus viridans* or HACEK group organisms (*Haemophilus* species, *Actinobacillus actinomycetemcomitans*, *Cardiobacterium hominis*, *Eikenella corrodens*, and *Kingella* species).
  - *S. aureus* bacteremia is highly associated with IE, and ECHO should be obtained with all *S. aureus*-positive blood cultures.
- Non-IE can be seen with systemic inflammatory disorders, such as SLE (Libman-Sacks endocarditis).
- Left-sided valves (mitral, aortic) are most commonly affected, except in IV drug users, in whom right-sided valves (tricuspid > pulmonary) predominate.
- IE diagnosed by the modified Duke criteria and presentation can be variable and nonspecific.

Etiology

- Autoimmune activity or bacterial colonization leads to endothelial injury and valvular damage.
- *S. aureus* is the most common bacterial isolate in high-income countries.
- Oral viridans group is the most common bacterial isolate in lower-middle income countries.
- Group D streptococci is classically seen in pts with colonic tumors.

Usual Treatment

- Empirical antibiotics (often combination therapy with focus on Gram-positive bacteria) started after blood cultures obtained when suspicion for IE exists
- Antibiotic treatment for at minimum 4 wk
- Surgery indicated in severe valvular dysfunction, with uncontrolled infection, and to prevent stroke/other embolic process

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Roth spots (retinal hemorrhage)	Visual disturbance	Retinal exam	Dilated funduscopic exam
RESP	Pulmonary edema	Dyspnea	Crackles Wheezing	CXR
CV	New or changed murmur Left ventricular or right ventricular failure Conduction abnormality	Dyspnea Orthopnea Light headedness/ syncope	Exam for signs of CHF (left or right sided, depending on lesion)	ECHO (TEE more sensitive than TTE) ECG
GI	Mesenteric ischemia Splenomegaly	N/V Pain	Acute abdomen Hypoactive bowel sounds	Contrasted CT Lactate
CNS	Stroke	Focal neurologic deficit(s)	Detailed neurologic exam	Head CT, MRI
RENAL	Hematuria Glomerulonephritis Pyuria	Urgency Pain Urine discoloration	CVA tenderness	UA
MS	Osler nodes Janeway lesions Splinter hemorrhages	Fever Night sweats Malaise	Skin and nailbed exam	ESR/CRP Rheumatoid factor

Key References: Cahill TJ, Prendergast BD: Infective endocarditis, *Lancet* 387(10021):882–893, 2016; Methangkool E, Howard-Quijano K, Ho JK, et al.: Infective endocarditis: the importance of intraoperative transesophageal echocardiography, *Anesth Analg* 119(1):35–40, 2014.

Perioperative Implications

Preoperative Preparation

- Assess cardiac status.
- Optimize volume status.
- CT head for baseline study and to rule out embolic stroke.

Monitoring

- Large bore central venous access
- Arterial pressure monitoring
- Noninvasive and invasive cardiac monitoring as indicated

Airway

- Potential for laryngeal involvement (i.e., edema, ulceration, VC paralysis) in pts with SLE.

Induction

- Consider etomidate, ketamine, and/or opioids to avoid SVR reduction.
- Awake arterial line may be beneficial.

Extubation

- Assess and manage postop cardiac dysfunction.
- Consider extubation to NIPPV in pts with continued signs of CHF.

Adjuvants

- Prophylaxis for dental procedures and respiratory tract procedures with biopsy recommended for pts with:
  - Prosthetic heart valves.
  - Unrepaired CHD.
  - Heart transplant with valvular disease.
- Prophylaxis not recommended for bronchoscopy (without biopsy) and GI or GU procedures.

Postoperative Period

- Early postop neurologic exam to assess for CNS embolization

Anticipated Problem/Concerns

- Embolization (intracranial, coronary, mesenteric) can lead to secondary infection.
- Ischemic stroke not a contraindication to surgery, but hemorrhagic stroke requires surgical delay of at least 1 mo.

Epidermolysis Bullosa

Sumita Bhambhani

Risk

- 1:17,000, 50% dystrophic form
- Racial distribution equal

Perioperative Risks

- Difficult IV access, airway, intraop positioning, reflux, steroid dependence, intraop hemorrhage, sepsis, iatrogenic corneal abrasion, blister formation, and airway obstruction

Worry About

- Problems similar to those found in pts with severe skin burns; severely compromised pts
- Difficult intubation (23%) secondary to microstomia
- Establishing monitoring and IV access
- Dehydration and malnutrition
- Anemia, hypoalbuminemia, electrolyte imbalance, and thrombocytosis
- Septicemia
- Renal and adrenal dysfunction

Overview

- Characterized by epithelial blistering resulting from minor trauma by lateral shearing forces, not pressure, because of absence of normal intracellular bridges caused by collagen abnormality
- Four types: SEB, JEB, DEB, and Kindler syndrome
- Associated conditions: Growth retardation, pyloric stenosis, esophageal stricture, pseudosyndactyly, enamel hypoplasia, muscular dystrophy, squamous cell carcinoma, and malignant melanoma
- SEB: Most common form; intraepidermal blisters on the soles and palms only in Weber-Cockayne form, generalized in Kobner form, generalized herpetiform in Dowling-Meara form, and generalized in association with muscular dystrophy in the MD form
- JEB: Blisters formed in the intralamina lucida and in intertriginous areas in the inversa form, which are generalized with growth retardation in the Herlitz form, generalized without growth retardation in the non-Herlitz form, and generalized with pyloric atresia
- DEB: Blisters formed in the sublamina densa and in intertriginous areas in the inversa form, on ankles in the pretibial form, on arms and legs in the pruriginous form; generalized blisters in the non–Hallopeau-Siemens form and with growth retardation and severe extracutaneous involvement in the Hallopeau-Siemens form, and aggressive squamous cell carcinomas (very commonly)
- Kindler syndrome: Blisters formed at multiple levels, intralamina lucida and sublamina densa; Kindler syndrome (previously considered as poikilodermatous photosensitivity disease); skin findings including atrophic scarring and nail dystrophy; possibly associated with severe colitis, esophagitis, urethral strictures, and ectropions; squamous cell carcinoma (can develop)

Etiology

- SEB: Inherited autosomal, usually dominant, mutation producing abnormal keratin intermediate filament proteins 5 or 14, which weaken the epidermal architecture; in the MD form abnormality, plectin (cytolinker protein) is the cause.
- JEB: An inherited autosomal recessive mutation produces abnormal laminin 5, abnormal type XVII collagen, and abnormal $\alpha_6\beta_4$ integrin.
- DEB: An inherited autosomal dominant or recessive mutation produces abnormal type VII collagen.

Usual Treatment

- Treatment is supportive, similar to initial burn treatment, with silver impregnated creams and collagen allografts.
- Retinoids and growth-stimulator factors are used to induce wound-repair keratin 6, 16, and 17, which form a more normal epidermis.
- An emerging treatment uses isothiocyanate sulforaphane which induces keratin 16 and 17 and occurs naturally in broccoli sprouts.
- Pts receive steroids and supportive treatment, such as nutritional support, wound care, contracture release, esophageal dilation, oral surgery, and treatment of skin cancers.
- Future treatment is expected to involve gene therapy.