Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Aortic valve dysfunction	Dyspnea with exercise	High-pitched, early diastolic, decrescendo blowing murmur Mid-diastolic low-pitched murmur (Austin Flint) Widened arterial pulse pressure (water-hammer) To and fro bobbing of head (de Musset sign)	CXR ECHO Cardiac MRI
	LV dysfunction	Dyspnea with exercise Nocturnal dyspnea	Displaced posterior MI S_3	ECG CXR ECHO Cardiac MRI Cardiac cath
RESP	CHF	Dyspnea Nocturnal dyspnea	Rales S_3	CXR
GI	Splanchnic ischemia	Abdominal pain	Distended abdomen	

Key References: Nishimura RA, Otto CM, Bonow RO, et al: 2014 AHA/ACC guideline for the management of patients with valvular heart disease: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, *Circulation* 129(23):2440–2492, 2014; Wilson W, Taubert KA, Gewitz M, et al: Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group, *Circulation* 116(15):1736–1754, 2007.

Perioperative Implications

Preoperative Preparation

- Consider optimizing LV performance with vasodilators, inotropes, and diuretics.
- Avoid reduction in aortic diastolic pressure, and be vigilant regarding low mean arterial pressures despite apparently normal systolic pressures.
- Emergent procedures (acute aortic regurgitation); full-stomach precautions.

Monitoring

- Arterial cath.
- ECG leads II/V5 and ST-segment analysis.
- Consider pulmonary artery cath and transesophageal ECHO.

Preinduction/Induction

- Elective: Avoid hypertension, hypotension, hypoxemia, and bradycardia; nondepolarizing muscle relaxants may be preferred over succinylcholine due to their lack of bradycardic effects.
- Emergency (acute aortic regurgitation with aortic dissection): Weigh the aspiration risk against the danger of acute increases in aortic wall tension and avoidance of bradycardia and consider rapid-sequence technique.
- Decreased aortic diastolic pressure combined with elevated LV diastolic pressures can lead to decreased coronary perfusion and subendocardial ischemia.
- Bradycardia and elevations in SVR increase regurgitant fraction and decrease cardiac output.

Maintenance

- Hemodynamic goals remain to avoid bradycardia, increases in SVR and decreases in diastolic blood pressure.
- Pulmonary capillary wedge pressures may underestimate LVEDP due to premature closure of the mitral valve.
- Pulmonary capillary wedge pressures may overestimate LVEDP in pts with combined aortic regurgitation and mitral regurgitation.

Extubation

- Consider extubation for patients undergoing valve replacement in the intensive care unit after respiratory and hemodynamic criteria are met.

Postoperative Period

- Consider augmenting preload to maintain and preserve filling volume of a still-dilated LV cavity.
- Inotropic support may be required to maintain cardiac output if inadequate intraop myocardial preservation was achieved.
- Evaluation for neuro injuries secondary to embolism during valve replacement. Meticulous de-airing maneuvers will lessen gaseous microembolization.

Anticipated Problems/Concerns

- Prolonged Trendelenburg position may be poorly tolerated during central venous cath insertion.
- Intraaortic balloon counterpulsation contraindicated before valve replacement.
- Atrial fibrillation or other SVTs may be poorly tolerated and may require aggressive treatment.
- Retrograde cardioplegia (not anterograde) may be required for myocardial protection.
- Associated diseases may present difficult intubation (e.g., rheumatoid arthritis, Marfan syndrome, trauma from acute aortic dissection).
- On separation from cardiopulmonary bypass, complete ECHO exam is recommended to examine the integrity of the replacement, as well as unanticipated iatrogenic injuries to other cardiac structures.

Aortic Stenosis

Jared Feinman

Risk

- Most common valvular heart disease; prevalence only 0.2% among adults aged 50-59 y, but increases to almost 10% after age 80 y.
- Calcific aortic stenosis: Major risk factors are increasing age, LDL, diabetes mellitus, smoking, hypertension, and bicuspid valve anatomy. Less common risk factors include disorders of calcium metabolism, renal failure, and history of mediastinal radiation.
- Bicuspid aortic valve is present in 1-2% of USA population and accounts for 60% of AVRs in pts under age 70 y and 40% over 70 y.
- Rheumatic aortic stenosis: Late sequela of streptococcal infection, more common in developing countries and often involves other valves.

Perioperative Risks

- Hypovolemia and/or vasodilation from anesthetic drugs lead to hypotension due to lack of preload reserve necessary to overcome systolic pressure gradient in pts with severe AS
- Risk of myocardial ischemia is elevated due to increase in LVED pressure (reducing coronary perfusion gradient) and LVH (associated with structural coronary abnormalities)
- Bicuspid valve associated with ascending aortic aneurysm and dissection, with a lifetime risk of about 6%

Worry About

- Drop in SVR and preload leads to reduced stroke volume through stenotic valve.
- Reduced SVR and stroke volume leads to hypotension, which reduces coronary perfusion and may lead to myocardial ischemia.
- Tachycardia poorly tolerated.
- Diastolic dysfunction very common.
- Atrial fibrillation; atrial kick provides up to 40% of LVED volume in AS pts, and its loss can lead to profound hypotension.

Overview

- Normal valve area (AVA) 2.6-3.5 cm^2; AS classified as mild (AVA >1.5 cm^2), moderate (AVA 1-1.5 cm^2), and severe (AVA <1 cm^2).
- Stenosis at the aortic valve leads to development of pressure gradient from LV to aorta.
- Increase in LV systolic pressure increases wall tension, producing LV hypertrophy.
- LV hypertrophy and augmented preload are primary means of maintaining adequate stroke volume and cardiac output in severe AS.
- Hypertrophy decreases LV compliance and diastolic dysfunction may ensue, making atrial contraction critical for maintaining adequate LV filling and stroke volume.
- Preload reserve generally exhausted in severe AS, so hypovolemia and reduced SVR are poorly tolerated.
- Elevated LVED pressure and alterations in coronary microcirculation associated with LV hypertrophy reduce coronary perfusion.
- Angina, dyspnea, and syncope are common presenting symptoms.
- Diagnosis of AS is made using ECHO or in the cath lab by assessing pressure gradient and valve area.
- Mean and peak pressure gradients across the valve also are used to classify severity.

- Pressure gradients may be low despite severe AS in pts with reduced EF (low-flow/low-gradient AS) or small ventricular volumes (low-flow/low-gradient AS with preserved EF).

Etiology

- Congenital bicuspid (and rarely unicuspid) aortic valve
- Rheumatic aortic stenosis
- Calcific degenerative disease

Usual Treatment

- Early in the disease process, medical therapy is indicated, including lifestyle modification (e.g., smoking cessation, exercise), and judicious use of antihypertensives.
- In severe AS, medical therapy does not prolong life, and AVR is the only effective treatment.
- AVR is a Class I indication in pts with
  - Symptomatic severe AS.
  - Asymptomatic severe AS with a LVEF <50%.
  - Asymptomatic severe AS in pts undergoing other cardiac surgery.
- AVR is a Class IIa indication in pts with:
  - Asymptomatic severe AS and low surgical risk.
  - Asymptomatic severe AS and decreased exercise tolerance or fall in BP with exercise.
  - Symptomatic low-flow/low-gradient severe AS.
  - Moderate AS in pts undergoing other cardiac surgery.
- TAVR is now a commonly used alternative to open AVR in pts who pose a high surgical risk due to age, comorbidities, and/or previous cardiac surgery.
- Balloon valvuloplasty may be used as a bridge to definitive surgical treatment with open AVR or TAVR.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Severe aortic stenosis Myocardial ischemia Diastolic dysfunction Arrhythmias	Angina, dyspnea, syncope Angina Dyspnea Palpitations, syncope	Systolic murmur Rales, edema, wheeze Rales, edema, JV distention Irregular pulse on exam	ECHO, cardiac cath, and dobutamine stress ECHO in suspected low-gradient AS ECG, ECHO, coronary angiography CXR, ECHO ECG, Holter
CNS	Syncope	Syncope		ECG, Holter, ECHO

Key References: Rashedi N, Otto C: Aortic stenosis: changing disease concepts, *J Cardiovasc Ultrasound* 23(2):59–69, 2015; Cook D, Housmans P, Rehfeldt K: Valvular heart disease: replacement and repair. In Kaplan J, editor: *Kaplan's cardiac anesthesia: the echo era,* ed 6. St. Louis, 2011, Elsevier, pp 570–584.

Preoperative Preparation

- Adequate premedication to reduce preop tachycardia due to anxiety.
- Ensure adequate preload but beware of administering large amounts of volume rapidly due to diastolic dysfunction.
- Pts with severe symptomatic AS may benefit from postponement of elective surgery until after AVR is performed.

Monitoring

- ECG for ST segment analysis.
- Preinduction invasive arterial pressure monitoring.
- Pulm artery cath may be useful in major surgery with large fluid shifts to better assess LVED pressure and volume status.
- Transesophageal ECHO warranted when blood loss or volume shifts are anticipated and an experienced echocardiographer is available.

Airway

- None

Preinduction/Induction

- Narcotic heavy induction is beneficial due to bradycardia, maintenance of SVR, and blunting of sympathetic response to laryngoscopy.
- Alpha agonist like phenylephrine should be used to treat hypotension with induction.
- Judicious use of propofol (and co-administration of an alpha agonist) is warranted to limit drop in SVR. Etomidate may also be useful for this reason.
- Laryngoscopy only after sufficient sympathetic attenuation.

Maintenance

- Intraop fluid management should aim for maintaining already elevated left-sided filling pressures with adequate replacement of blood loss and insensible losses.
- Balanced anesthetic using narcotics, muscle relaxant, and a lower dose of volatile agent is preferred.
- Higher doses of volatile agents may depress cardiac function, increase risk of arrhythmia-induced hypotension, and drop SVR, leading to hypotension and myocardial ischemia.
- Caution with agents that decrease preload and afterload (e.g., nitroglycerin, nitroprusside), or any agent with significant histamine release.
- Caution with agents that directly or indirectly increase heart rate (e.g., pancuronium, atropine).
- Early electrical cardioversion for intraop atrial fibrillation.
- Generally, avoid neuraxial anesthesia (especially spinal) due to hypotension from sympathectomy. Epidural anesthesia may be used with extreme caution in laboring pts and other cases where its benefit is strong, but must be carefully dosed for a gradual onset of block with minimal drop in SVR, while simultaneously augmenting preload and administering vasoconstrictors when needed.

Extubation

- Minimize sympathetic stimulation and tachycardia.

Postoperative Period

- Aggressive pain control
- Maintenance fluids as appropriate to maintain adequate preload

Anticipated Problems/Concerns

- Myocardial ischemia with intraop hypotension.
- Diastolic dysfunction.
- Dysrhythmias can lead to precipitous hypotension and ischemia and should be treated aggressively until a return to sinus rhythm is achieved or hemodynamics stabilize.

Apert Syndrome (Acrocephalosyndactyly Type 1 and 2)

Andrea Johnson

Risk

- 15:1,000,000 live births
- Equal M:F ratio

Perioperative Risk

- Aspiration
- Bronchospasm
- Resp depression
- Airway obstruction

Worry About

- Difficult mask, airway, or IV access
- Elevated intracranial pressure, temperature dysregulation, and seizures
- Corneal abrasions (due to exophthalmos)
- PACU and perioperative monitoring for apnea
- Cardiac anomalies (10% of cases)
- Anatomic anomalies (regional/neuraxial anesthesia)

Overview

- Apert syndrome is a disorder identified by synostoses of the cranium, vertebral bodies, and digits. It is caused by a mutation in the FGFR-2 gene.
- Two major manifestations are bicoronal synostosis and maxillary hypoplasia. High-arched V-shaped palates and cleft palates are common.
- Strabismus, syndactyly, and conductive and neuronal hearing loss are common manifestations.
- Cognitive delay (IQ <70) see in 67% of pts.

Etiology

- Autosomal dominate disorder; however, most cases are sporadic mutations of the FGFR-2 gene.
- Sporadic mutations are associated with paternal age >40 y.

Usual Treatment

- Craniosynostosis release: Frontoorbital advancement usually around 6 to 8 mo of age
- Midface advancement: Correction of brachycephaly, orbital dystopia, or midface hypoplasia
- Correction of hypertelorism: Interorbital bone resection
- Mandibular and maxillary advancement and orthodontics: Usually conducted after cranial maturation to enhance cosmetic appearance