Ventricular Preexcitation Syndrome

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Risk

- Not all patients are symptomatic or have a prior diagnosis; 1–3% of diagnoses are made following ECG in previously asymptomatic individuals.
- WPW syndrome affects 0.1–0.3% of the population, is more prevalent in males, and is characterized by symptomatic arrhythmias and an ECG showing a short PR interval (<120 ms) with a widened QRS (>100 ms) and often, but not always, δ waves, representing fusion of early and late depolarization via the accessory pathway and AV node. Abnormal conduction occurs via a bundle-of-Kent accessory pathway between atria and ventricles. Risk of SCD is estimated to be 0.25% per annum in WPW syndrome.
- LGL syndrome affects 0.5% of adults, is more prevalent in women, and reflects abnormal conduction through the James bundle (atria to bundle of His). Characterized by a short PR interval, normal QRS, and no δ waves, it manifests through reentrant type PSVT or atrial fibrillation/flutter. No studies have shown an increased risk of SCD in LGL.
- The Mahaim type is a rarer form of preexcitation, caused by Aps in the right ventricle, called Mahaim fibers. It is characterized by a normal PR interval, long QRS, and δ waves and may trigger episodes of SVT.

Perioperative Risks

- General or regional anesthesia may unmask ventricular preexcitation syndromes.
- Asymptomatic individuals with a WPW ECG pattern may present no added risk. Symptomatic individuals with WPW syndrome are prone to PSVT (up to 75%), or less commonly to atrial fibrillation (15–30%) and flutter (5%); rapid ventricular rates may occasionally deteriorate to VT or VF.
- There is a danger of misrecognition of WPW ECG patterns for BBB (wide QRS); myocardial infarction (negative δ waves simulating pathologic Q waves); other tachyarrhythmias (including VT). All prompt inappropriate treatment.
- Drugs used to suppress AV conduction (to slow the ventricular rate in treatment of atrial fibrillation/flutter) may dangerously accelerate the rate in WPW.

Worry About

- Periop nausea, gagging, hypothermia, pneumoperitoneum, and pregnancy can all accentuate conduction via abnormal APs. Hyperadrenergic states, overstimulation (including laryngoscopy), and other interactions can also provoke or aggravate tachyarrhythmias.
- High spinal anesthesia may block sympathetic cardiac accelerator nerves and suppress normal AV conduction. Alongside relative parasympathetic predominance, this may further facilitate conduction by the APs, resulting in preexcitation and tachyarrhythmias.
- Cholinergic medications, such as suxamethonium and reversal agents, along with other AV-nodal blocking drugs (diltiazem, digoxin) may enhance conduction via APs, worsening tachyarrhythmias.
- Potential of WPW-related PSVT to deteriorate into atrial fibrillation or flutter, with danger of extremely rapid ventricular rates and ensuing VT and VF.

Overview

- The presence of a short PR interval, often with a δ wave, reflects the underlying early depolarization of ventricles and defines preexcitation syndrome.
- The extent of preexcitation may change depending upon the conduction characteristics of the abnormal AP, AV node, and autonomic tone. The majority of pathways allow dual conduction, both anterograde (i.e., atrial to ventricular) and retrograde (i.e., ventricular to atrial). Retrograde-only conduction occurs in 15% and anterograde only is rare.
- Pts with a ventricular preexcitation syndrome have an abnormal AP that bypasses the AV node to electrically connect atria and ventricle. The majority of these APs generate fast action potentials due to rapid sodium current influx, resulting in faster conduction of electrical impulses to the ventricle than the slower calcium current–dependent AV nodal route. This electrochemical distinction means that the onset of ventricular activation occurs *earlier* than if impulses had conducted through the AV node; hence preexcitation. Additionally, at faster atrial rates, progressive prolongation of AV nodal conduction is also bypassed, meaning that atrial tachycardias may deteriorate into VF.
- A range of arrhythmias may occur in preexcitation syndromes, from SVTs (more common) to life-threatening ventricular arrhythmias (less common).
- OAVRTs are due to abnormal circuits, in which anterograde conduction occurs via the AV node and retrograde conduction via the AP. Since the normal AV pathway is used for ventricular depolarization, QRS complexes are narrow and no δ waves are present. This accounts for most tachyarrhythmias in WPW syndrome (70%).
- Antidromic tachycardias derive from anterograde ventricular activation via an AP, with retrograde current reentering atria via the AV nodal route. Wide QRS and δ waves are present on the ECG.
- The first episode of PSVT in many pts appears before the age of 20 y, rarely in middle age, and infrequently after age 50. The frequency of episodes of PSVT increases with age in WPW syndrome.

Etiology

- APs are congenital in origin, and preexcitation syndromes may be hereditary.
- Associated with congenital cardiac defects (most commonly tricuspid value lesions) and acquired cardiac defects (e.g., cardiomyopathy, idiopathic hypertrophic subaortic stenosis, asymmetric septal hypertrophy).

Usual Treatment

- Long-term therapy in recurrent symptomatic tachyarrhythmias is through the delivery of electrical or radiofrequency impulses to the AP using catheter ablation. Success rate is about 90–95%.
- Pharmacologic prophylaxis is reserved for pts in whom catheter ablation has failed. Agents that prolong refractoriness in APs are recommended, such as disopyramide, procainamide, and flecainide.
- In the acute setting with a hemodynamically unstable pt, the treatment of choice is synchronized DC electrical cardioversion (50–200 J). The minimum effective energy should be used initially and energy subsequently titrated to minimize potential injury to the myocardium.
- The most common tachyarrhythmia in pts with preexcitation is a regular, narrow-complex tachycardia due to an OAVRT. These tachycardias respond well to treatments that momentarily block transmission through the AV node. In stable pts, vagal maneuvers may be attempted, followed by rapid administration of adenosine (unless contraindicated). Second-line agents include calcium channel blockers such as verapamil. An external cardioverter-defibrillator should be immediately available, as adenosine can increase risk of atrial fibrillation with preexcitation, and calcium channel blockers can precipitate reduced cardiac output in those who have structural heart disease.
- If a regular, broad complex tachyarrhythmia occurs in a pt with preexcitation, it should be considered as ventricular in origin as it is often difficult to determine the difference between ventricular origin and an OAVRT with bundle branch block, for example. Drugs that block the AV node (adenosine, calcium-channel blockers, beta blockers, digoxin) must *not* be used as they could precipitate VF and cardiac arrest. AP-blocking drugs such as procainamide and flecainide (class 1a) are recommended first-line treatment in the stable pt. Amiodarone slows AV node conduction as well as AP conduction and should be used with caution.
- If the pt presents with a tachyarrhythmia that is irregularly irregular, this is likely due to the different degrees of fusion at the ventricular level, as AV and AP conduction coexists. Preference should be to block the AP conduction (e.g., procainamide) with amiodarone as the second line.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Arrhythmia LV function	Palpitations, dizziness, syncope, angina, chest pain, sometimes asymptomatic Weakness, exercise intolerance, CHF	Monitor BP; variable S_1-pulse amplitude; fast regular, irregular, and/or weak pulse	12-lead ECG, Holter ECG, cardiac EP study, ECHO, possible further study

Key References: Blomström-Lundqvist C, Scheinman MM, Aliot EM, et al.: ACC/AHA/ESC guidelines for the management of patients with supraventricular arrhythmias—executive summary: a report of the American college of cardiology/American heart association task force on practice guidelines and the European society of cardiology committee for practice guidelines (writing committee to develop guidelines for the management of patients with supraventricular arrhythmias), *Circulation* 108(15):1871–1909, 2003; Bengali R, Wellens H, Jiang Y: Perioperative management of the Wolff-Parkinson-White syndrome, *J Cardiothorac Vasc Anesth* 28(5):1375–1386, 2014.

Perioperative Implications

Preoperative Preparation

- Take a thorough cardiac history to elucidate symptoms in keeping with existence of preexcitation, such as palpitations, syncope, dyspnea, angina, or dizziness. A 12-lead ECG can identify preexcitatory syndrome patterns, but these may not always be apparent.
- If pt is symptomatic, a cardiology consultation should be sought. This may involve exercise testing and Holter monitoring to determine the AP anterograde refractor period. If the pt is symptomatic, he or she will almost always be offered catheter ablation therapy.
- Pts on prophylactic medications must continue these and have them on the day of surgery, with minimal delay to taking them postop.
- Pts may benefit from RA where appropriate to avoid sympathetic stimulation, stress, and tachycardia.

Monitoring

- Ventricular preexcitation does not mandate use of invasive BP monitoring, central venous access, or placement of defibrillator pads. The anesthetist must consider the ability to urgently access the chest or arms while the pt is under the drapes or in the prone position.

Induction
- A smooth induction with the use of anxiolytics should be implemented in pts who may hyperventilate secondary to stress. A deep plane of anesthesia must be balanced with the effects of agents such as propofol, which can cause hypotension and compensatory tachycardia.
- Aim to obtund the effects of laryngoscopy by using a supraglottic airway device such as a laryngeal mask airway where appropriate.
- Adequate preloading should be considered to avoid the use of sympathomimetics for BP, and sympathomimetics should be used cautiously.

Maintenance
- The volatile agent halothane can precipitate conduction via APs and should be avoided.
- Avoid agents that can precipitate tachycardia, such as ketamine and pancuronium.

Extubation
- Avoid neostigmine, which induces vagal tone, causing slowing of the AV node and preference for conduction down AP. Avoid atropine and glycopyrrolate, which can induce tachycardia.

Adjuvants
- For control of postop N/V, avoid metoclopramide and cyclizine, which can induce tachycardia.

Postoperative Period
- Adequate pain control is essential and use of regional anesthesia may be beneficial.

Anticipated Problems/Concerns
- SVT or VF in those known to have preexcitation with or without symptoms throughout the periop period.
- Vigilance is required in interpreting the ECG of a tachyarrhythmia to avoid incorrect drug selection.

Ventricular Septal Defect (Congenital)

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Risk
- Incidence: About 2–6:1000 live births.
- May be isolated or part of several complex malformations such as TOF.

Perioperative Risks
- Mortality higher in older children; elevated PVR (>7 Wood units); surgery may be complicated by complete heart block.

Worry About
- Worsening of L-to-R shunt with hyperventilation and increased FIO_2
- Paradoxical embolization
- Hypothermia
- Post–CPB pulm Htn and RV failure

Overview
- Small defects asymptomatic, present with murmur, and usually close spontaneously.
- Larger unobstructed defects result in CHF symptoms, poor weight gain, and URIs beginning at 3–12 wk of age, as decreases in PVR cause increase in L-to-R shunting.
- Untreated large L-to-R shunting may result in fixed pulm Htn (Eisenmenger syndrome) in some pts.

Indications/Usual Treatment
- Some 75% of small defects close spontaneously. Small, unrepaired defects do not require antibiotic prophylaxis.
- Medical therapy for symptoms of CHF includes digoxin, ACE inhibitors, and furosemide.
- Surgery is indicated when CHF not amenable to medical treatment or if there is FTT.
- Surgical repair contraindicated if PVR >10 Woods units unless reactive to selective pulm vasodilators.
- Transcatheter closure is often used for muscular defects, which can be difficult to access surgically.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Low forward cardiac output due to L-to-R shunt Pulm Htn due to excessive flow	CHF symptoms, FTT Age of pt	Loud holosystolic murmur and thrill Cyanosis	Auscultation, ECHO, cardiac cath
RESP	Congestion/edema due to L-to-R shunt	Frequent URIs	Rhonchi	CXR
HEME	Anemia in massive L-to-R shunt; polycythemia in R-to-L shunt	Pallor or cyanosis	Paleness or plethora	Hct
MS	Chronic hypoxemia due to late reversal of shunt flow (Eisenmenger syndrome)	Cyanosis	Clubbing of digits	Pulse oximetry

Key References: Penny DJ, Vick GW 3rd: Ventricular septal defect, *Lancet* 377(9771):1103–1112, 2011; Scully BB, Morales DL, Zafar F, et al.: Current expectations for surgical repair of isolated ventricular septal defects, *Ann Thorac Surg* 89:544–549, discussion 550–551, 2010.

Perioperative Implications

Preoperative Preparation
- Digoxin and furosemide until day of surgery; ACE inhibitors controversial, but vasoplegic syndrome following CPB less common in pediatric pts.
- May not be possible to delay operation until pt is free of upper resp symptoms.

Anesthesia
- Limit FIO_2 to minimum necessary prior to CPB to restrict excessive pulm blood flow.
- Maintain normal to slightly high $PaCO_2$ to restrict excessive pulm blood flow.
- Pts typically receive inhalational anesthesia for induction; if peripheral IV in place, IV drugs can be administered alternatively.
- Avoid N_2O to prevent sequelae of paradoxical air embolization.

Monitoring
- Indwelling arterial catheter for invasive monitoring in all pts.
- Central venous access and pressure monitoring in most pts undergoing surgery with CPB.
- Standard ASA monitoring, including pulse oximetry, ECG, capnometry, multiple-site temp monitoring.
- TEE

Induction/Maintenance
- Mask induction with sevoflurane in most cases; IV drugs if peripheral IV in situ; IM induction possible for uncooperative pts.
- High-dose opioid anesthesia technique rarely used.

Surgical Stages
- Pre-CPB:
  - Low FIO_2, normal to high $Paco_2$.
  - Avoid hemodilution with large amounts of crystalloid and/or colloid prior to CPB.
- CPB:
  - After pt's Hct has been obtained in the OR, dilutional Hct including CPB prime is calculated. If calculated Hct is less than 25%, consider priming of CPB with whole blood or reconstituted whole blood (PRBC and FFP).
  - Inhalational anesthetic administration via CPB or continuous IV drug administration is recommended to allow for fast-tracking in most pts presenting for VSD repair.
- Post-CPB:
  - Rule out residual shunting by TEE.
  - Maintain Hct >25% to 30%.

Postoperative Period
- Most pts presenting for VSD repair can be extubated at end of surgery.
- Consider mechanical ventilation and sedation in the immediate postop period in pts prone to pulm hypertensive crises (e.g., Down syndrome).
- Infective endocarditis prophylaxis for 6 mo; if residual defect is present, should be continued indefinitely.

Anticipated Problems/Concerns
- Imbalance in pulm to systemic blood flow ratio:
  - Excessive pulm blood flow results in high arterial saturation but with diminished tissue perfusion and metabolic acidosis.
  - Diminished pulm blood flow results in good tissue perfusion but with cyanosis and potential injury due to hypoxia.
- Postop ventricular dysfunction more likely with ventriculotomy.
- Pulm Htn and/or right heart failure.
- Coagulopathy, particularly in very small children.