

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
NEURO	Tremor, muscular rigidity, dysarthria, apraxia	Medication history, difficulty ambulating, talking	Focused neuro exam looking for strength/rigidity	
CV	Early: LV thickening, SVTs Late: Hyperdynamic state—high CO, low SVR	SOB, chest pressure/flutter	Auscultate, pronounced LLSB	ECG; TTE if indicated by clinical symptoms
RESP	Pulm shunting 2/2 high portal pressures Hepatopulmonary syndrome in severe cases	SOB, hypoxia	Auscultate	CXR, PFTs
HEME	Anemia, thrombocytopenia	Decreased Hct	Signs of bruising, petechiae	CBC, PT/INR
GI	Esophageal varices, ascites, hepatomegaly/splenomegaly	Upper GI bleeding, paracentesis, abdominal fullness	Abdominal pain, hepatomegaly, splenomegaly	Lytes, liver enzymes
RENAL	Renal failure, can be acute or chronic	Oliguria	Decrease in UOP	Lytes; rarely need kidney biopsy

**Key References:** Baykal M, Karapolat S: Anesthetic management of a pediatric patient with Wilson's disease. *J Clin Med Res* 2(2):99–101, 2010; Vaja R, McNicol L, et al: Anaesthesia for patients with liver disease. *Contin Edu Anaesth Crit Care Pain* 10(1):15–19, 2010.

**Perioperative Implications**

**Monitoring**

- Standard ASA monitors.
- Recommend arterial line and central line in fulminant liver disease.
- Also consider TEE or PA catheter.

**Induction**

- Decreased doses of hypnotic agents 2/2 cardiac function and neurologic disease.

- Vecuronium/rocuronium have prolonged elimination.
  - Cisatracurium does not rely on hepatic metabolism.
- Maintenance**
- Isoflurane, sevoflurane, and desflurane undergo minimal hepatic metabolism.
  - Morphine metabolism can be delayed 2/2 decreased hepatic blood flow, and its active metabolite, morphine-6-glucuronide, will accumulate 2/2 renal failure.

**Postoperative Period**

- Rare concerns for respiratory failure 2/2 ascites.
- Avoid dopaminergic drugs (i.e., droperidol, metoclopramide).

**Anticipated Problems/Concerns**

- Remember: Anything you would be concerned about for ESLD can be seen in Wilson disease.
- Assess neurologic and cardiac status. Let the severity of symptoms guide your periop plan.

## Wolff-Parkinson-White Syndrome

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**Risk**

- WPW pattern (asymptomatic) prevalence: 0.15–0.25% in the general population and 0.55% in pts with a primary relative with WPW; autosomal dominant trait.
- WPW syndrome (ECG pattern and arrhythmia) prevalence is 0.005% to 0.07% in the general population and approximately 2% out of pts with WPW. It is often first presented in ages 20–40 y.

**Overview**

- Definition: WPW syndrome is a preexcitation syndrome. Ventricular depolarization occurs in part via an AP from the atrium (bundle of Kent) bypassing the AV-His Purkinje conduction system.
- The AP allows for antegrade or retrograde conduction which is faster than the AV node resulting in a shortened PR interval (<0.12 sec). The impulse then spreads through the muscle fibers until it joins the regular conduction system resulting in a slurred upstroke and widening of the QRS complex on the ECG.

- PSVT results from a reentrant circuit involving the AV node and AP. The QRS complex during PSVT matches the usual QRS morphology when conduction is antegrade through the AV system and retrograde through the AP (i.e., orthodromic). 5–10% of the time, conduction through the AP is antegrade (i.e., antidromic in the reentrant circuit), producing a wide QRS complex. This rhythm may be confused with VTach.
- AFIB and/or AFLT is more common in pts with WPW. Usually, AFIB is precipitated by an episode of PSVT. Rapid ( $\geq 300$  bpm) ventricular rates may occur in pts with APs with short refractory periods. These pts are at risk for developing Vfib and hemodynamic collapse.
- Other heart abnormalities (e.g., Ebstein's anomaly) are often commonly (7–20%) associated with WPW.

- AFIB (15–35%); increasing incidence with age. A major concern is rapid ventricular response due to antegrade conduction over AP.
- Atrial flutter (5%).
- VFIB/sudden death (0–0.4%): Out of rapid ventricular response due to antegrade conduction over AP in AFIB/AVRT.

**Usual Therapy**

- With severe hemodynamic compromise, synchronized DC cardioversion (50–100 J).
- AVRT and/or narrow complex tachycardia: Apply vagal maneuvers or IV adenosine (6–12 mg IV). A small incidence of induction of AFIB with adenosine therapy for PSVT in WPW has been described.
- AFIB: Agents that reduce the accessory bundle refractory period (digoxin, Ca<sup>2+</sup>-channel blockers, beta-blockers, and adenosine) increase the risk of causing VFIB and hemodynamic collapse in pts with WPW and AFIB and should therefore be avoided.
- Broad complex tachycardia (i.e., antidromic AVRT) should be treated with IV procainamide or amiodarone.

**Perioperative Risks**

- AVRT (80% of pts WPW syndrome): Rapid HR impairs LV filling, leading to hemodynamic instability and/or myocardial ischemia.

Assessment Points			
ECG Criteria	P Wave and PR Interval	QRS	Comments
Classic (type A)	Shortened PR interval, typically <0.12 s (left-sided bypass track)	Slurred upstroke (delta wave), widened QRS complex	The faster the AP conduction, the more prominent the delta wave and the wider the QRS
Atypical (type B)	Shortened PR interval (right-sided bypass track)	Q waves (inverted delta wave) in V1	May be confused with MI
Concertina effect	Periodically progressive shortening of the PR interval, with the P wave disappearing in QRS	The shorter the PR interval, the more pronounced is the delta wave (wider QRS)	This is the result from a periodically increased conduction via the AP
Intermittent WPW	May be mistaken for frequent ventricular premature beats, if it persists for several beats may be held for accelerated idioventricular rhythm		

**Key References:** Wheeler DW, Sayeed RA, Ritchie AJ: Unsuspected Wolff-Parkinson-White syndrome causing arrhythmias after cardiac surgery. *J Cardiothorac Vasc Anesth* 16(3):354–356, 2002; Bengali R, Wellens HJ, Jiang Y: Perioperative management of the Wolff-Parkinson-White syndrome. *J Cardiothorac Vasc Anesth* 28:1375–1386, 2014.

### Perioperative Implications

#### Preoperative Preparation

- If preexcitation on ECG or Hx of WPW, consider cardiology evaluation.
- If symptomatic, consider electrophysiologic study and catheter ablation.
- Continue all preop cardiac and anti-arrhythmic medications.

#### Monitoring

- ECG for detection of periop PSVT or AFIB.
- Consider arterial line and CVP catheter if LV dysfunction or valve disease, as these pts have a high dependence on preload and atrial kick.
- For emergency surgery, consider placement of defibrillator pads prior to induction.

#### Maintenance

- Consider RA techniques to avoid sympathetic stimulation.
- Avoid laryngoscopy and non-depolarizing muscle relaxants if possible to avoid reversal agents and

neostigmine, which can facilitate transmission via an accessory pathway. Use LMAs when indicated.

- Avoid light planes of anesthesia, anxiety, hypovolemia, hypothermia, hyperventilation, which can all increase sympathetic tone that may decrease the refractory period and therefore accelerate the conduction in the AP and AV node. This may facilitate the precipitation of AVRNT, AFIB, and/or VFIB.
- Volatile anesthetics and IV induction agents such as propofol and benzodiazepines seem to have no influence on the conduction system and are safe to use. Sevoflurane and isoflurane as well as medications that enhance vagal tone (e.g., opioids, dexmedetomidine) actually decrease conduction via the AP and are safe to use as well.
- Limit the use of vagolytic agents (e.g., glycopyrrolate, atropine) and ketamine.
- Use  $\alpha$ -1 agonists (phenylephrine) instead of ephedrine to avoid positive chronotropy and arrhythmias.

### Postoperative Period

- Implement pain management to avoid catecholamine excess.
- If the delta wave appears in periop period, rule out myocardial infarction (decreased AV conduction second to ischemia facilitating increasing AP conduction).

### Anticipated Problems/Concerns

- AV nodal blockers (digoxin,  $Ca^{2+}$ -channel blockers, adenosine, and beta-blockers) may shorten refractoriness in the AP and thereby provoke VFIB in WPW pts with AFIB.
- Hemodynamic collapse may occur when verapamil or beta-blockers are used in the treatment of antidromic (wide-complex) PSVT in pts with WPW that is mistaken for VTach.