Overview

- Most common abdominal tumor of childhood; prognosis related to staging.
- Because of the location of the tumor, blood loss can be significant.
- Tumor is also associated with other congenital abnormalities, which may affect anesthetic and/or surgical management.
- Tumor extension into IVC and heart carries increased morbidity and mortality.

Etiology

- Embryonal neoplasm
- No consistent chromosome abn, although abnormalities in chromosomes 1 and 11 are common.
- Three genes associated with Wilms:
  - 11p13 interstitial deletion associated with Wilms-aniridia-growth retardation
  - 11p15.5 deletion associated with Beckwith-Wiedemann syndrome
  - Third locus not determined to be associated with familial Wilms

Usual Treatment

- Chemotherapy (with vincristine, actinomycin D, and adriamycin).
- Radiotherapy.
- Surgical removal of tumor: If tumor bilateral, surgery has focused on nephron-sparing procedures. (Procedure including biopsy followed by chemotherapy and delayed definitive resection.)
- Open or laparoscopic procedure.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Beckwith-Wiedemann syndrome	Obstructive airway secondary to large tongue	Direct exam	Blood glucose levels
CV	Htn Tumor extension into heart	Asymptomatic	Htn	ECG CT abdomen US renal vein/IVC Cardiac ECHO
RESP	Resp compromise	Abdominal distentions Metastatic disease Tumor embolization	Increased RR Hypoxemia	Pulse oximetry CT abdomen US renal vein/IVC Possible cardiac ECHO
GI	Gastric reflux	Increased intraop pressure Hx of reflux	Abdominal distention	Review CT scan
HEME	Von Willebrand syndrome	Unusual bleeding		Bleeding time Ristocetin platelet aggregation

Key References: Whyte SD, Mark Ansermino J: Anesthetic considerations in the management of Wilms' tumor, *Paediatr Anaesth* 16(5):504–513, 2006; Green DM: The evolution of treatment for Wilms tumor, *J Pediatr Surg* 48(1):14–19, 2013.

Perioperative Implications

Preoperative Preparation

- Htn-controlled.
- R/O renal vein and/or IVC tumor involvement.
- Evaluate for bleeding disorder.
- Evaluate cardiovascular function if prior chemotherapy Rx.

Monitoring

- Arterial cath may be indicated.
- CVP cath may be needed. especially if IVC and tumor extend mid-heart.
- For preexisting hematuria, Foley cath can aid in fluid balance.
- IV catheters above diaphragm; large-bore cath preferable.
- $ETCO_2$ to rule out air and/or tumor embolus.

Airway

- May be a problem if Beckwith-Wiedemann syndrome is present.

Preinduction/Induction

- Age-appropriate use of sedation.
- Rapid-sequence if increased intraabdominal pressure.
- Regional anesthesia; epidural or paravertebral block for postop pain.
- Preexisting chemotherapy may have cardiac depressant effect.
- IV access above diaphragm.

Maintenance

- Requires a prolonged procedure.
- Avoid N_2O.
- Maintain temperature.
- Increased third space fluid requirements.
- Procedure may be associated with large blood loss.
- Pulm function may be compromised, secondary to metastasis and/or tumor embolization, abdominal distention, and/or surgical traction.

Extubation

- Expected if temp maintained and pt hemodynamically stable.

Postoperative Period

- Administer pain control:
  - Multimodal anesthetic
  - RA (epidural or paravertebral)
- Third space fluid requirements.
- Htn may still be present.

Anticipated Problems/Concerns

- Risk of tumor and/or air embolus: If tumor extends into renal vein, IVC may have to be cross-clamped, the IVC opened, and the tumor removed.
- Intraop blood loss can be extensive.
- Periop implications.

Wilson Disease

Cobin D. Soelberg

Risk

- Incidence: 1:30,000.
- Slightly more common among Eastern European Jewish populations.
- Children and young adults tend to present with nonspecific GI symptoms.
- Adults tend to present with neurologic symptoms.

Perioperative Risks

- Increased risk of liver failure, kidney failure, and cardiac complications.
- 6–12% of all pts require liver transplantation.

Overview

- Presentation can vary widely. Hepatic symptoms tend to present prior to neurologic symptoms.
- Often nonspecific symptoms such as abdominal pain, nausea, and vomiting may occur. Rarely presents with acute liver failure.
- More commonly presents with elevated transaminases, hepatomegaly or hepatosplenomegaly, or mild jaundice.
- Can be diagnosed by presence of Kayser-Fleischer rings and low serum ceruloplasmin levels. In the absence of Kayser-Fleischer rings, diagnosis is more difficult, relying on free copper and liver copper concentrations.

Etiology

- Autosomal recessive.
- In the liver, copper is not passed to ceruloplasmin and so the liver has an excess of copper. Once this excess exceeds the ability of the liver to hold it, it is released in its free form into the blood. It then accumulates in tissues and causes damage.
- Copper balance is regulated through excretion of bile.
- Copper accummulation leads to hepatic cirrhosis
- Lenticular degeneration.

Usual Treatment

- D-penicillamine, an oral chelating agent
- Trientine in pts with adverse reactions to penicillamine
- Liver transplantation in rare cases of fulminant liver failure

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

Sara K. Davis | Jeffrey R. Kirsch

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 (≥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.

Perioperative Risks

- AVRT (80% of pts WPW syndrome): Rapid HR impairs LV filling, leading to hemodynamic instability and/or myocardial ischemia.
- 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^{2+}-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.

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.