PART ONE

Co-Existing Diseases

ACHONDROPLASIA

GRETE H. PORTEOUS, MD

OVERVIEW

- Definition: Rare (4/100,000) genetic disorder characterized by abnormal endochondral bone formation.
- Achondroplasia is the major cause of dwarfism.
- Etiologies
 - > Spontaneous mutation is the most common cause (80% of cases).
 - > Autosomal dominant inheritance accounts for the remaining cases.
- Characterized by
 - ≻ Short stature
 - ➤ Shortened extremities
 - ➤ Enlarged head
 - ≻ Saddle nose
 - ➤ Trunk of relatively normal length
- In most cases there are no changes in intelligence, longevity, or reproductive ability.
- Usual Rx: None

PREOP

Issues/Evaluation

- Difficult airway. Pts w/ achondroplasia are traditionally considered to have difficult airways because of
 - ➤ Large tongue
 - ➤ Large mandible
 - ➤ Foramen magnum stenosis
 - Limited neck extension
 - ➤ Increased incidence of atlanto-axial instability
- Difficult spinal & epidural anesthetic. This is the result of several spine abnormalities:
 - ➤ Marked kyphoscoliosis & lumbar lordosis.
 - Bony landmarks are difficult to identify.
 - Prolapsed intervertebral discs, osteophytes, shortened pedicles, & a narrow epidural space increase the risk of dural puncture & decrease the chance of successful catheter placement.
 - Spinal stenosis may impair free flow of CSF, making dural puncture hard to identify.

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Achondroplasia

- Anesthetic may spread unpredictably in the epidural space, leading to "patchy" block or higher-than-expected block.
- ➤ In the past, many considered regional anesthesia to be contraindicated in this pt population, but several recent studies have reported success w/ few complications from regional.
- Pulmonary
 - May have decreased chest wall compliance & FRC because of thoracic kyphoscoliosis & rib deformities.
 - Frequent respiratory tract infections may (rarely) lead to cor pulmonale.
- Pregnancy
 - ➤ High incidence of cesarean section for delivery due to cephalopelvic disproportion.
 - Airway management can become even more difficult because of the typical changes of the parturient superimposed on a preexisting difficult airway.

What To Do

- Obtain careful surgical/anesthetic history & any available anesthetic records.
- Inform pt of potential difficulty of airway management & possibility of awake fiberoptic intubation.
- Document neurologic exam carefully if regional anesthesia planned, as preexisting neuropathies due to spinal abnormalities are not uncommon.

INTRAOP

- Airway management. Prepare for difficult intubation:
 - ➤ Multiple laryngoscope blades
 - Smaller-than-usual endotracheal tubes
 - ≻ LMAs
 - ➤ Fiberscope readily available
 - > Avoid vigorous cervical extension w/ intubation
- Regional anesthesia
 - Spinal block: There is no consensus in the literature on type & dose of anesthetic; it is probably rarely performed.
 - Epidural block: There is more reported experience w/ this technique. Epidural catheters are placed in the usual manner, maintaining high suspicion for unrecognized subarachnoid catheter placement. Recommended test dose is the usual (1.5 cc of lidocaine 1.5% + epinephrine 1:200,000). The most appropriate type

Achondroplasia

Acromegaly

& volume of epidural dose is unclear, but generally considered to be less than a usual dose. Successful examples in the literature for lumbar epidurals for C-section:

- 8 cc of 2% lidocaine + bicarb + epi (1:200,000)
- * 12 cc of 2% lidocaine + bicarb + epi (1:200,000) + fentanyl 37.5 mcg
- 9 cc of 3% 2-chloroprocaine
- 5–12 cc of 0.5% bupivacaine (at 12 cc a C5 level achieved)
- 21 cc of 0.75% bupivacaine

POSTOP

- Standard management, w/ attention to airway patency & adequacy of ventilation.
- Document neurologic exam prior to discharge from PACU.

ACROMEGALY

BETTY LEE-HOANG, MD

OVERVIEW

- Definition
 - Acromegaly is a condition caused by overproduction of growth hormone from the anterior pituitary, usually by a pituitary tumor.
 - > The condition results in overgrowth of skeletal, soft & connective tissues.
 - > Pts usually have enlarged hands, feet, jaw & tongue.
 - Major organs including the heart, lungs, liver & kidney are also increased in size.
 - Airway anatomy is altered, including enlargement of the tongue & epiglottis, mandible hypertrophy & generalized soft tissue growth, which may make airway management difficult
- Usual Rx
 - > Hypophysectomy (excision of the pituitary tumor)
 - The surgical approach usually taken is transsphenoidal; alternately, a bifrontal craniotomy approach can be taken.

PREOP

Issues/Evaluation

- A thorough history & airway exam are required.
- If pt complains of dyspnea, hoarseness or stridor or has been recently diagnosed w/ sleep apnea, indicating a risk of airway obstruction

CAMBRIDGE

Acromegaly

Acute Hepatitis 5

w/ sedation or general anesthesia, consider an awake fiberoptic intubation.

- Pts w/ acromegaly may have glottic or subglottic stenosis, nasal turbinate enlargement, vocal cord thickening, or recurrent laryngeal nerve involvement.
- Evaluate pts for hypertension, hyperglycemia, congestive heart failure, peripheral nerve or artery entrapments, skeletal muscle weakness.

INTRAOP

- Be prepared for a difficult airway w/ several laryngoscope blades, laryngeal mask airways (LMA) & a fiberoptic bronchoscope as backup.
- Pts may need treatment of hypertension intraop, especially if a transsphenoidal procedure is performed, since the nasal septum is usually prepped w/ cocaine, epinephrine, or phenylephrine.

POSTOP

- Because pts are at risk of airway obstruction & may have difficult airways, make sure pt is fully awake & following commands before extubation.
- If pt has undergone a hypophysectomy as treatment for acromegaly, pituitary insufficiency may arise postop (eg, TSH, ACTH may be low).

ACUTE HEPATITIS

JOSEPH COTTEN, MD, PHD

OVERVIEW

- Definition: inflammation of hepatocytes
 - Multiple causes, including viral infection, toxin, fatty liver infiltration of pregnancy, sepsis, congestive heart failure.
- Viral hepatitis
 - Symptoms in order of incidence: dark urine, fatigue, anorexia, nausea, fever, emesis, headache, abdominal discomfort, lightcolored stool, pruritus.
 - ≻ Hepatitis A
 - · Fecal-oral transmission; blood transmission is rare.
 - Chronic disease or carrier state does not exist.
 - ≻ Hepatitis B
 - Parenteral, oral-oral, sexual transmission.

Acute Hepatitis

- HBsAg indicates infectivity.
- HBcAg indicates high infectivity.
- HBsAg presence for >6 mo suggests a chronic carrier state (1–10% become chronic).
- · Chronic active disease often progresses to cirrhosis.
- ≻ Hepatitis C
 - · Causes most posttransfusion hepatitis.
 - Chronic liver disease develops in 80% of infected pts & 20% of these will develop cirrhosis.
 - Sexual & casual household contact w/ saliva is inefficient means of transmission.
- ➤ Hepatitis D
 - Requires the presence of hepatitis B for its expression.
 - · Hepatitis B vaccination prevents hepatitis D.
- Epstein-Barr virus (EBV): usually produces mild hepatitis associated w/ infectious mononucleosis
- Cytomegalovirus (CMV): CMV is present in most adults; liver disease is mild & nonchronic
- Drug-induced
 - > Toxins: alcohol, carbon tetrachloride, vinyl chloride
 - Therapeutic drugs most frequently implicated: isoniazid, methyldopa, rifampin, acetylsalicylic acid, nonsteroidal antiinflammatory drugs (NSAIDs)
 - ➤ Halothane produces two types of hepatotoxicity:
 - Mild self-limited postop toxicity due to changes in hepatic blood flow that affect hepatic oxygenation
 - Halothane hepatitis, a life-threatening immune-mediated response after repeat exposure to halothane via trifluoroacetyl metabolite (less likely in pediatric pts)
 - Isoflurane/enflurane/desflurane/sevoflurane, like halothane, can produce mild self-limited postop toxicity due to changes in hepatic blood flow. However, they all maintain blood flow similarly & better than halothane
 - These compounds, w/ exception of sevoflurane, may be capable of producing a more severe immune-mediated hepatitis but undergo less extensive metabolism than halothane

PREOP

Issues/Evaluation

Elective surgery should be postponed for pts w/ acute hepatocellular injury due to increased morbidity & mortality.

Acute Hepatitis

- One study found a 31% mortality rate for pts undergoing exploratory laparotomy w/ unsuspected parenchymal liver disease.
- Another study noted a 9.5% mortality rate for pts w/ acute viral hepatitis undergoing laparotomy.
- Severely jaundiced pts (>8 mg/dL) are more likely to develop postop renal failure & sepsis.
- Decreased perfusion of the liver, which occurs during all anesthetics, neuraxial & general, may be responsible for poor outcomes in pts w/ parenchymal liver disease.
- Liver perfusion is affected most greatly by procedures anatomically adjacent (eg, cholecystectomy).

What To Do

- Rule out acute liver failure (as opposed to merely acute hepatitis).
 - Findings in acute liver failure can include encephalopathy, cerebral edema, coagulopathy, renal failure, infection, hypoglycemia, etc.
 - > See also Coexisting Disease chapter "Acute Liver Failure."
- Blood glucose may be low w/ severe liver injury & should be corrected.
- Thrombocytopenia should be corrected if present.
- Evaluate LFTs & viral serology if appropriate.
 - > Prothrombin time: assesses current liver synthetic function
 - May be prolonged by vitamin K deficiency; consider vitamin K administration if this is suspected
 - Aspartate aminotransferase (AST), alanine aminotransferase (ALT):
 - · Found in large quantity in the liver.
 - Levels >500 U/L occur w/ acute hepatocellular injury.
 - Modest injury <300 U/L occurs in a variety of conditions (eg, acute or chronic hepatocellular injury, infiltrative disease, biliary obstruction).
 - ALT is generally more sensitive than AST for viral hepatitis.
 - AST is elevated twofold in excess of ALT in alcoholic liver disease.
 - If transaminase levels are >3 times normal, nonelective procedures requiring general or regional anesthetic should be postponed & a gastroenterologist consulted.

Acute Hepatitis

- Lower elevations require repeat evaluation to assess for worsening levels, stable levels, or improvement in levels as well as viral hepatitis serologies.
- Surgery may proceed if moderately elevated transaminase levels are stable or trending down & viral serologies are negative; otherwise, seek gastroenterology consult.
- \succ Albumin
 - · Synthesized by liver
 - Half-life of 14–21 days, so is not beneficial for evaluation of acute disease
- Alkaline phosphatase (AP): present in bone, intestine, liver. AP is elevated in biliary obstruction, cholestasis, space-occupying lesions, infiltrative diseases.
- ➤ Gamma-glutamyl transpeptidase (GGT)
 - Increases in GGT & AP tend to occur in similar diseases.
 - GGT is elevated in pts ingesting certain agents (eg, alcohol, barbiturates, phenytoin).
- Lactate dehydrogenase (LDH): abundant in the liver but may arise from many sources, including red blood cells, as during hemolysis
- Total bilirubin (a byproduct of heme degradation) is either conjugated (direct-acting, water-soluble, renally excreted) or unconjugated (indirect-acting, protein-bound).
 - Jaundice is apparent when levels exceed 3-4 mg/dL.
 - Indirect bilirubin increases w/ hemolysis, Gilbert's or Crigler-Najjar syndrome, heart failure, or portosystemic shunting.
 - Direct bilirubin increases w/ hepatocellular dysfunction or biliary tract obstruction.

INTRAOP

Management

- Drug disposition may be difficult to predict.
 - Isoflurane, desflurane, sevoflurane may be best for maintenance of hepatic blood flow; consider supplementation w/ nitrous oxide & IV agents (which may have delayed clearance).
 - > Pseudocholinesterase deficiency is rare such that succinylcholine & mivacurium action should not be prolonged.
 - Atracurium & cis-atracurium are cleared independent of liver function.

Acute Hepatitis

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- Vecuronium & rocuronium are unlikely to be prolonged unless large doses are used.
- To optimize liver perfusion & prevent secondary ischemic injury, minimize hypotension & hypoxia & maintain normocarbia.
- Administer fresh-frozen plasma for coagulopathy.
- Consider monitoring of blood glucose, acid-base status, coagulation profile, urine output.

POSTOP

- Be alert to worsening liver function after surgical stress.
- Severe postop jaundice may be related to hypotension, hypoxia, multiple transfusions.
- "Shock liver" is a condition caused by marked or prolonged hypotension.
 - ➤ Typical findings include
 - Tenfold increase in transaminase levels
 - Coagulopathy
 - · Possibly liver failure
 - > A milder form can be seen in pts after cardiopulmonary bypass.
- Reversible, minor abnormalities in LFTs can be detected in up to 50% of all patients postop.
- Postop jaundice is present in 20% of all patients after major surgery.
- Causes of postop jaundice (can be distinguished by LFT evaluation).
 - ➤ Increased bilirubin
 - Hemolysis
 - · Hemolysis of transfused blood
 - · Resorption of hematoma
 - ➤ Hepatic damage
 - Intrahepatic cholestasis
 - · Circulatory failure
 - Drug-induced
 - · Pre-existing disease
 - ➤ Obstructive
 - Common bile duct stone
 - Bile duct injury
 - Pancreatitis
 - \succ Other
 - Gilbert's disease (7–10% of patients) exacerbated by fasting state, cholecystitis

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Acute Liver Failure (Fulminant Hepatic Failure)

ACUTE LIVER FAILURE (FULMINANT HEPATIC FAILURE)

JOSEPH COTTEN, MD, PHD MANUEL PARDO, JR., MD

OVERVIEW

- Definition: hepatic encephalopathy & coagulopathy in the setting of acute hepatic disease. Time from onset of jaundice until onset of encephalopathy distinguishes fulminant (within 8 wks) from subfulminant (within 26 wks) liver failure.
- Pts w/ fulminant hepatic failure (FHF) are usually critically ill & do not undergo elective procedures. Urgent procedures that may involve the anesthesia provider include central line placement, dialysis line placement, tracheal intubation for airway protection, GI endoscopy, ICP monitor placement, head CT scan, or liver transplantation.

Causes

- ➤ Viral hepatitis
 - Most commonly A or B
 - Occasionally cytomegalovirus, Epstein-Barr virus, herpes viruses are implicated
- ≻ Drugs
 - Predictably toxic drugs (eg, acetaminophen; typically >12 g ingestion, less in the presence of alcohol or starvation)
 - Idiosyncratic reactions to inhaled anesthetics, sulfonamides, phenytoin, oral hypoglycemics (troglitazone), others

≻ Toxins

- · Organic solvents: trichloroethylene, tetrachlorethane
- Herbal remedies: kava kava
- Toxins from the "death cap" mushroom (Amanita phalloides)
- ≻ Vascular
 - Myocardial infarction, cardiac arrest, cardiomyopathy, pulmonary embolism, metastatic or infiltrative cancer, amyloidosis, veno-occlusive disease (Budd-Chiari, chemotherapy)
- ➤ Miscellaneous causes
 - Fatty liver of pregnancy (third trimester; frequently w/ preeclampsia)
 - Reye's syndrome
 - Wilson's disease (acute liver failure can be first presentation)
- Important: Understand the multiple organ system complications of FHE The most significant complications include