Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Inadequate	Noticeable SOB	Respiratory rate high	ABG
MS	Weakness	Exercise, fatigue	Limb tone Hypoactive muscle stretch reflexes	HyperPP: Ictal serum K^+ level (elevated in approximately 50%) EMG myotonia (in approximately 75%) HypoPP: Low ictal serum K^+ level Exclusion of secondary causes (TSH, fT4 fT3 levels) Glucose/insulin or ACTH infusion induces paralysis attack Plasma biochemistry after attack: elevated myoglobin, creatine kinase Muscle fiber conduction velocity may be slower than normal No EMG myotonia

Key References: Suetterlin K, Männikkö R, Hanna MG: Muscle channelopathies: recent advances in genetics, pathophysiology and therapy, *Curr Opin Neurol* 27(5):583–590, 2014; Bandschapp O, Iaizzo PA: Pathophysiologic and anesthetic considerations for patients with myotonia congenita or periodic paralyses, *Paediatr Anaesth* 23(9):824–833, 2013.

Perioperative Implications

Preoperative Preparation

- HyperPP: carbohydrate loading during fasting period; consider 24-h furosemide for K^+ depletion
- HypoPP: avoid large glucose and salt loads; 24-h acetazolamide if not already given; only glucose-free solutions IV; if Hx of frequent instability, prepare infusion with K^+; reduce pt's anxiety

Monitoring

- Both types: Temperature (esophageal) (keep warm); ECG (detection of dyskalemia); NM monitoring mandatory (minimize relaxant dose)

Airway

- Both types: no special difficulty, but may need support

Preinduction/Induction

- Both types: Regional techniques are appropriate; relaxation with short-acting nondepolarizing agents as indicated.
- HyperPP: Avoid ketamine; no succinylcholine (severe myotonia, hyperkalemia with resulting postop muscle weakness over days).

Maintenance

- Both types: Use warming blankets and keep normothermic; warm all IV fluid.
- HyperPP: Use glucose 5% as maintenance, avoid hypoglycemia; do not give K^+-containing solutions, maintain normokalemia (use glucose/insulin if needed).
- HypoPP: Use MH trigger-free anesthetic methods; glucose-free solutions as maintenance, avoid hyperglycemia; give solutions containing K^+, aim for K^+ 4–5 mEq/L; ventilation during anesthesia should be normocarbic to avoid K^+ shifts.

Extubation

- HyperPP: Evidence of muscle weakness should be treated with IV calcium gluconate or chloride 10% 10 mL slowly over 5 min; anticholinesterase drugs may worsen/trigger myotonic symptoms.
- HypoPP: Evidence of muscle weakness should be treated with IV potassium chloride; normal reversal as indicated clinically; maintenance by IPPV if evidence of weakness in postop phase; severe postop weakness may be aggravated by Ca^{2+}.

Adjuvants

- Both types: Anticipate usual analgesic requirements for age and surgery; regional techniques are appropriate.
- HyperPP: Some experimental evidence suggests that condition (e.g., postop weakness) may be helped by phenytoin or by salbutamol.
- HypoPP: Ca^{2+}-channel blockers do not appear to be contraindicated in pts with concomitant CV disease.

Anticipated Problems/Concerns

- Both types: Cold triggers attack.
- HyperPP: Succinylcholine may not give relaxation, and therefore intubation may be difficult; severe myotonia may create resp difficulty; hypoglycemia and K^+ can trigger hyperkalemic attack; hyperkalemia can cause cardiac arrhythmia.
- HypoPP: May have associated supraventricular or conduction defect arrhythmias; resp muscle weakness may occur postop; must maintain serum K^+ above 4.0 mEq/L.

Fanconi Syndrome

Rayhan A. Tariq | Henry Liu

Risk

- FS can be inherited, acquired, or caused by exogenous factors.
- Incidence is sporadic. Exact incidence in USA is not clear.
- Most diseases associated with FS are inherited in an autosomal recessive pattern.
- Cystinosis is the most common cause in pediatric pts.

Perioperative Risks

- Potential for hypotension secondary to hypovolemia
- Renal failure, proximal renal tubular dysfunction
- Lyte imbalance (especially hypokalemia causing tachyarrhythmias)

Worry About

- Polyuria, polydipsia, and the resulting dehydration
- Type 2 renal tubular acidosis (defect in the reabsorption of bicarbonate in the proximal tubule)
- Hypokalemia-induced ventricular arrhythmogenicity
- Hypophosphatemia and associated osteomalacia and loss of bone density
- Hypokalemia may cause muscular weakness
- May be part of a genetic syndrome, such as Lowe syndrome (oculocerebrorenal syndrome)

Overview

- Disease of the proximal convoluted tubules in which glucose, amino acids, uric acid, phosphate, and bicarbonate are passed into urine, not being reabsorbed.
- Signs and symptoms reflect the tubular abnormality, including polyuria, polydipsia, and acidosis due to bicarbonate loss.
- Pts are very likely to have renal failure at the time of surgery.
- Muscle weakness due to hypokalemia.
- Renal phosphate wasting presents as osteomalacia or rickets.
- Most pts have proteinuria, although it is often minimal.
- Severe photophobia might be present with cystinosis (cystinosis is the most common cause of inherited form of FS).
- FS is different from Fanconi anemia characterized by progressive pancytopenia.

Etiology

- May be hereditary or acquired.
- Common hereditary causes are cystinosis (most common cause in children), Wilson disease, Lowe syndrome, galactosemia, glycogen storage diseases, and hereditary fructose intolerance mitochondrial cytopathies.
- Can be acquired via exposure to heavy metals (lead) and medications such as tetracycline (particularly when outdated), cisplatin, tenofovir, adefovir, rifampin, deferasirox, and aminoglycoside antibiotics.
- Dysproteinemias, such as multiple myeloma, amyloidosis, light-chain nephropathy, and benign monoclonal gammopathy may cause FS in adults.

Usual Treatment

- Mainstay of treatment is replenishment of lytes and fluids lost in urine.
- Metabolic acidosis due to bicarbonate loss is corrected by the administration of HCO_3^-, (usually 3–10 mg/kg/d of sodium bicarbonate in divided doses).
- Addition of a thiazide diuretic (1–3 mg/kg per d) of hydrochlorothiazide may be necessary to avoid volume expansion, which amplifies the excretion of bicarbonate by lowering the renal threshold (also need K^+ supplementation to account for extra K^+ lost in the urine with the use of TZD diuretic).
- For preventing bone disease, phosphate and vitamin D supplementation are necessary in addition to correction of metabolic acidosis.
- Losses of glucose and amino acids are not usually symptomatic and do not require treatment.
- Kidney transplantation in pts with renal failure due to cystinosis.
- Liver transplantation in case of Wilson disease.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Arrhythmogenicity due to electrolyte disturbances, hypovolemia	Assess for clinically symptomatic bradycardia, heart block, CHF	Auscultation of heart sounds, ECG	Continuous ECG monitoring
RENAL	Metabolic acidosis, polyuria, K^+, Mg^{2+}, Ca^{+2}, PO_3^- loss in urine	Assess GFR and residual renal function.	Assess for dehydration	ABG, UA, BMP
CNS	Lyte imbalance, hypoglycemia may cause confusion/ disorientation and/or seizures; rarely muscle weakness	Evaluate pt compliance	Lung sounds: rales, edema of extremities	Neurologic assessment

Key References: Klootwijk ED, Reichold M, Unwin RJ, et al.: Renal Fanconi syndrome: taking a proximal look at the nephron, *Nephrol Dial Transplant* 30(9):1456–1460, 2015; Pandey R, Garg R, Chakravarty C, et al.: Lowe's syndrome with Fanconi syndrome for ocular surgery: perioperative anesthetic considerations, *J Clin Anesth* 22(8):635–637, 2010.

Perioperative Implications

Preoperative Concerns

- Existence of any other coexisting genetic/metabolic disorder should be ruled out by thorough Hx, physical exam, and special test(s).
- Preop ABGs, ECG.
- Preop electrolytes level (K^+, Mg^{2+}, Ca^{+2}, PO_3^-) and glucose in the morning of surgery.
- Correction of electrolyte imbalances (K^+, Mg^{2+}, Ca^{+2}, PO_3^-).
- Metabolic acidosis corrected by administering $NaHCO_3^-$ preop to maintain plasma bicarbonate levels at about 20 mEq/L.

Induction/Maintenance

- During laryngoscopy, special attentions to avoid overextension and pt positioning to prevent injury to the rickety bones
- Closely monitoring acid-base and fluid-electrolytes imbalance during the surgery
- Monitoring volume status by UO and CVP

Postoperative Period

- Postop labs of urine analysis, serum lytes, calcium, phosphorous, glucose, BUN, creatinine, albumin, and hematologic profile are used to guide postop care
- ECG monitoring in PACU (hypokalemia)

Anticipated Problems/Concerns

- Lyte imbalances warrant monitoring and correction periop
- Potential hypovolemia
- Other coexisting metabolic/genetic disorder(s)

Fat Embolism

Shamsuddin Akhtar

Risk

- Long bone fractures and pelvic fractures:
  - 80–100% fat embolism
  - Less than 1–30% FES
- Male-female ratio: 4:1
- Adult greatly increased over pediatric
- Multiple fractures >single fractures
- Pathologic fractures >traumatic fractures
- Total hip, total knee replacement, intramedullary nailing:
  - 27–100% fat embolism
  - Unknown incidence FES
- Unusual causes: Liposuction, fat injection, bone marrow harvest and/or transplantation, vertebroplasty, cardiopulmonary bypass, CPR, burns, pancreatitis, sickle cell disease, osteomyelitis, fatty liver, soft tissue injury

Perioperative Risks

- FES: <10% mortality
- Preexisting FES: Respiratory failure/ ARDS, RV dysfunction, shock, coagulopathy, neurologic dysfunction
- Intraop fat embolism: Shock, hypoxemia

Worry About

- Preexisting FES: Hypoxemia, reduced pulm compliance, pulm Htn, RV failure, hypotension, cardiac arrest, coagulopathy
- Intraop embolism: Hypotension, RV failure, hypoxemia, paradoxical embolization, stroke, neurologic dysfunction (delirium to coma, postop)

Overview

- Fat particles (globules of marrow fat) traveling into blood and lung.
- Must distinguish fat embolism, from FES (triad of hypoxemia, petechiae, and neurologic abnormalities). Fat embolism is more common than FES.
- FES can produce mild pulm dysfunction to severe ARDS.
- Pulm Htn and acute RV failure may occur in severe cases of FES.
- Typically the onset of signs and symptoms of FES happen 12–72 h following injury.
- Fat embolism occurs commonly during femoral reaming and cementing in hip arthroplasty.
- FES is confounded with cement reaction during arthroplasty.

Etiology

- Most frequently follows orthopedic trauma with release of marrow fat into venous circulation
- Pathology produced by mechanical obstruction by intravascular fat passing into the pulm and systemic arterial circulation and by production of endogenous inflammatory mediators

Usual Treatment

- Early fracture fixation to decrease embolization.
- Use of noncemented prosthesis or venting of femoral shaft may reduce embolization during hip arthroplasty.
- Unreamed nailing for fracture fixation to reduce embolization.
- O_2 therapy to maintain SaO_2 >90%.
- Low tidal volume ventilation strategy with PEEP as for ARDS.
- Aggressive hemodynamic support with fluid and/or inotropes for shock and/or RV failure.
- Factor replacement for coagulopathy with bleeding.
- Corticosteroids, heparin, ethanol, dextran, aspirin, and prophylactic vena caval filter are of unproven benefit.