MKSAP CLINICAL PEARLS

Clinical and Board Applicability

- Thalassemia trait is associated with mild anemia, microcytosis, hypochromia, normal RBC count and target cells on the peripheral blood smear

- α-Thalassemia trait normal hemoglobin electrophoresis results
- β-thalassemia trait a slightly increased hemoglobin A2 and some residual hemoglobin F.
- No treatment

Rachmilewitz EA, Giardina PJ. How I treat thalassemia. Blood. 2011 Sep 29;118(13):3479-88.

What to do with a pacemaker or AICD during surgery?

The Issue: electocautery makes pacer or AICD think that there is a beat or arrhythmia!

Pacer: asynchronous pacing with magnet.

AICD: shocking stopped with magnet; to put in asynchronous pacing will need reprogramming

Epstein AE, DiMarco JP, Ellenbogen KA, et al. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. Circulation. 2013 Jan 22;127(3):e283-352.

Those with unruptured intracranial aneurysms; smoking increases chance of rupture.

Wiebers DO, Whisnant JP, Huston H 3rd, et al; International Study of Unruptured Intracranial Aneurysms Investigators. Unruptured intracranial aneurysms: natural history, clinical outcome, and risks of surgical and endovascular treatment. Lancet. 2003 Jul 12;362(9378):103-10.

More Info:

- It is likely that smoking is associated with an increased risk of SAH by contributing to formation of an aneurysm and increasing its rate of growth.
- prevalence of active smoking ranging from 45 to 75% in patients with SAH .
- Those who had quit smoking had no increased risk of aneurysm growth.
- Therefore, cessation of cigarette smoking may help reduce both the risk of formation of aneurysms and the risk of rupture.

Juvela S: Natural history of unruptured intracranial aneurysms: risks for aneurysm formation, growth, and rupture. Acta Neurochir Suppl 82:2730, 2002

Juvela S: Risk of subarachnoid hemorrhage from a de novo aneurysm. Stroke 32:19331934, 2001

Repair of unruptured cerebral artery aneurysms are recommended 7mm in posterior circulation and 12mm in the anterior circulation.

MS Exacerbation: What to consider before high dose steroids?

Pseudorelapse of Multiple Sclerosis

Thrower BW. Relapse management in multiple sclerosis. Neurologist. 2009 Jan;15(1):1-5. PMID: 19131851

What is it?

- Worsening of baseline neurologic symptoms due to physiologic stressors.
- Don’t treat for MS exacerbation but treat current disease process.

The most appropriate and cost-effective means of assessing **acute monoarthritis** ?

Aspiration and analysis of the synovial fluid for leukocytes, gram stain, with culture, and crystal analysis.

Joshua F. Ultrasound applications for the practicing rheumatologist. Best Pract Res Clin Rheumatol. 2012 Dec;26(6):853-67. PMID: 23273796

Migraine Headache Diagnostic Accuracy

Eighty percent of those who come to office or ED with headache:

Migraine

Diagnosis of Migraine:

P	pulsatile
O	one day duration
U	unilateral
N	nausea
D	disabling

- 4 to 5 of the above symptoms is 92 percent.
- 3 of the 5 symptoms then the probability decreases to 64 percent.
- 0 to 2 symptoms, likelihood of a migraine is 17 percent.

- Schulman E. Refractory migraine—a review. Headache. 2013 Apr;53(4):599-613.

Issue of Asian patients and epilepsy:

Genetic anomaly: HLA-B 1502 allele

Krumholz A, Wiebe S, Gronseth GS, et al. Evidence-based guideline: Management of an unprovoked first seizure in adults: Report of the Guideline Development Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Neurology. 2015 Apr 21;84(16):1705-13.

Hypersensitivity to:

- Carbamezapine
- Lamotrigine
- Oxcarbazepine
- phenytoin.

The Problem:

- Steven Johnson's
- DRESS

Pregnancy & epilepsy:

No valproic acid!

Lamotrigine(have to adjust) & levetiracetam with folic acid are the best choices.

Krumholz A, Wiebe S, Gronseth GS, et al. Evidence-based guideline: Management of an unprovoked first seizure in adults: Report of the Guideline Development Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Neurology. 2015 Apr 21;84(16):1705-13.

Hepatitis C Screening testing:

Centers for Disease Control and Prevention (CDC). Testing for HCV infection: an update of guidance for clinicians and laboratorians. MMWR Morb Mortal Wkly Rep. 2013 May 10;62(18):362-5. PMID: 23657112

- Positive **anti-HCV** with negative **HCV RNA**

= false-positive **anti-HCV** or cleared infection.

- Positive **anti-HCV** with positive **HCV RNA**

= active infection.

- **HCV RNA** may be positive despite a negative **anti-HCV**

= Acute HCV infection or immunosuppressed patient.

Osteoporosis Treatment Monitoring:

- Follow BMD not T score in following up for worsening of OP on or off of treatment.
- Every effort should be made to have subsequent scans done on the same machine.

- A calculated change of about 4% likely represents a statistically significant change.

- Adler RA, El-Hajj Fuleihan G. Managing osteoporosis in patients on long-term bisphosphonate treatment: report of a Task Force of the American Society for Bone and Mineral Research. J Bone Miner Res. 2016 Jan;31(1):16-35.

Primary Hereditary Hemochromatosis genetics and diagnosis:

Autosomal recessive defect in the HFE gene:

- Two main defects in HFE: the C282Y and H63D mutations. Neither mutation is common in blacks or Asians with iron overload.

Testing:

- serum transferrin saturation
- positive test:
  greater than 60% in men .
  greater than 50% in women.

- Elevated serum ferritin further supports the diagnosis.

- Homozygosity for C282Y is found in 85% to 90% of phenotypically affected persons.
- Only 10% of those homozygous for C282Y develop symptoms.
- One copy of C282Y and one copy of H63D, known as compound heterozygotes.
  - at much lower risk for iron overload than those who are homozygous for C282Y.

Liver biopsy not a benign procedure!

MRI first choice for assessing cardiac and liver iron overload.

- 5% to 10% of patients with iron overload have a negative hemochromatosis gene test.
- The absence of C282Y does not eliminate the diagnosis of an iron overload disorder.
- Testing should be performed in first-degree relatives of patients with classic HFE-related hemochromatosis.

Hemoglobin A1c problems:

- falsely elevated in the setting of chronic kidney disease.
- falsely decreased, iron deficiency, blood transfusions, and increased erythropoiesis with erythropoietin use.
- blood glucose measurements do not correlate with the most recent hemoglobin A1c

Fructosamine:

- reflect mean blood glucose values over a much shorter period of time (two weeks).
- not as good with low albumin
- supportive use with serial glucose measurements like A1C.

Principle of DM2 and hypertensive management:

- Lower dosage of medication gives the post potent glucose lowering or blood pressure lowering effect .

- Higher dosage of medication doesn't help lower glucose or bp but significant increases bothersome and serious side affects.

Pheochromocytoma α blockade indications:

- a contrast-enhanced adrenal CT scan; administering iodine contrast media to could incite a hypertensive crisis.

Primary Aldosteronism (htn, low K):

- midmorning ambulatory plasma renin activity (PRA) and plasma aldosterone concentration (PAC).
- Normal volume and potassium.
- positive if PAC is frankly elevated (>15 ng/dL), PRA is suppressed, and PAC/PRA ratio is greater than 20.

Medications that get in the way of results:

- Stop spironolactone and eplerenone 4 weeks prior to testing.
- Diuretics should also be discontinued to assure euvolemia.

ACE-i/ARB's:

- if PRA is suppressed despite treatment with an ACE inhibitor or angiotensin receptor blocker, PA is likely.

- If results are difficult to interpret:
  Verapamil, hydralazine, and α-blockers can be substituted for blood pressure control if necessary.

TSH pearls:

- Treat to level of less than 2.5 in pregnancy.
- Normal up to 8 in the elderly.
- Secondary hypothyroidism: follow T4 levels.

Hyperthyroidism treatment needed but work up incomplete:

- Treat with thionamide until euthyroid.
- Stop thionamide for 1 week and get radioactive iodine uptake scan than resume treatment.

Testosterone Replacement Pearl:

- If on testosterone replacement for one year a patient's testes will not produce testosterone again.
- Placing a young man on replacement causes infertility.

Primary Hyperparathyroidism indications for parathyroidectomy:

- Impaired kidney function (defined as eGFR <60 mL/min/1.73 m2
- 24-h urine calcium >400 mg/24 h.
- the presence of nephrolithiasis or nephrocalcinosis by radiograph, ultrasound, or CT
- age younger than 50 years
- a serum calcium level greater than or equal to 1 mg/dL above upper limit of normal
- a T-score of −2.5 or worse at the lumbar spine, total hip, femoral neck, or distal radius
- those in whom medical surveillance is neither desired nor possible.

Vitamin D Replacement Pearl:

Ergocalciferol/Vitamin D2:

-50,000 U every other day to
-recommended when a patient's vitamin D level is less than 10 ng/mL.

Cholecalciferol/Vitamin D3:

-400 units to 2000 units daily
-is often used when the level is between 20 and 30 ng/mL or for maintenance.

Either:

-between 10-20 mg/ml

Distinguish between primary hyperparathyroidism and familial hypocalciuric hypercalcemia (FHH) :

- Young patients (teens to 30’s)
- Parathyroid hormone level is toward the upper end of the normal range.
- Mild hypercalcemia.
- Family history of hypercalcemia.

Diagnostic differentiation:

-24-hour urine collection for calcium and creatinine

Results:

-Total urine calcium of less than 200 mg/24 h (5 mmol/24 h) and a calcium-creatinine ratio less than 0.01 are highly suggestive of familial hypocalciuric hypercalcemia (FHH).

Specifics:

- Specific calcium-sensing receptor in the parathyroid glands and kidneys
- Making this diagnosis is crucial because it may prevent unnecessary parathyroidectomy for the patient.
- Rarely associated with hypercalcemia and therefore does not require therapy to lower serum calcium levels
- Screening other family members for the disorder is indicated.

HSV Encephalitis Pearl:

- The sensitivity and specificity of cerebrospinal fluid (CSF) PCR for the diagnosis of herpes simplex encephalitis are greater than 95%, and PCR has replaced brain biopsy as the gold standard for laboratory confirmation of that condition.

- Early in the course of infection, the PCR result may be falsely negative.