Screening for Cerebrovascular Disorders: A Guide for Primary Care

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Carotid disease

• 2 Sources:
  – 2011 Guideline on the management of patients with extra-cranial carotid and vertebral artery disease
  – 2017 Clinical practice guidelines of the European Society for Vascular Surgery
Asymptomatic Carotid disease screening

• DUS is recommended as the frontline test
  – Low cost, low risk, accessible
  – For asymptomatic patients with suspected disease
  – Is of uncertain benefit for those without
    • Clinical manifestations
    • Risk factors
Asymptomatic Carotid Screening

• Follow-up DUS *annually* is reasonable for
  – Postprocedure followup
  – Evaluate response to treatment in those with > 50% stenosis
Asymptomatic Carotid Screening
Don’t forget the basics!

• Smoking cessation
• Hypertension control
• Hyperlipidemia management
• ASA
• Diabetes- “strict glycemic control”
Which Asymptomatic patients should be referred?

• >70% stenosis by non-invasive study
• The rate of stroke/MI/Death needs to be < 3%
• There is no role for revascularization in those with
  – Chronic total occlusion
  – < 50% stenosis
  – Severe disability
Symptomatic Carotid Stenosis

- Neurologic event (stroke or TIA) within 6 months
- DUS + another modality (MRA, CTA, Angiography)
  - Confirm the degree of stenosis
  - Anatomic data to determine options
- $\geq 70\%$ noninvasive
  - 50-69\% noninvasive may benefit but NNT much higher
Symptomatic Carotid Stenosis

• Interventions should have complication < 6%
• CEA remains the gold standard
  – CAS alternative based on
    • Comorbidities eg significant CAD, CHF, COPD
    • Prior RTX to the neck
    • Anatomic factors e.g. high bifurcation, isolated hemisphere
Unruptured Aneurysms (UIA)

Prevalence:
2000-4000/100K UIA
C/W 10/100K SAH

Only a small percentage rupture

F>M

Peak age 50-60s

Risk Factors
1. Genetics
2. Family Hx
3. Modifiable

Stroke 2015;46:2368-2400
Genetic Risk Factors

- At risk disorders (<10% of UIA):
- Most common: AD PCKD (4-13%)
- EDS type IV
- Marfan
- HHT
Family Risk Factors

• Family occurrence 7-30%

• Increased risk with first degree relatives with h/o SAH
  – 4% by MRA
  – Siblings > children of affected

• Aneurysm in >=2 relatives
Modifiable Risk Factors

• Smoking
• Hypertension
• Excessive Alcohol (> 3 drinks/day)
Factors for rupture of UIAs

- Age > 60
- Female sex
- Japanese or Finnish descent
- Size > 5mm
- Posterior circulation
- “Symptomatic”
- Evidence of growth on serial imaging
Screening modalities

• CTA and MRA are useful for detection and follow up
  – MRA avoids radiation
  – MRA better for detection of aneurysm > 3mm size
  – MRA is not as sensitive for infundibulum vs aneurysm

• Angiography can be useful compared with noninvasive modalities
  – If treatment is considered
  – The most sensitive modality for previously treated aneurysms
Who to screen for UIA

• >=2 family members with UIA or SAH
  – This is particularly high yield in combo with HTN, smoking and female sex
  – Siblings > children of index SAH patient

• Certain conditions
  – AD PCKD, especially with Family H/O IA
  – Coarctation of aorta
  – Microcephalic osteoplastic primordial dwarfism
**Arteriovenous Malformations (AVM)**

Abnormal connection between arterial and venous systems
- Lack intervening capillary bed

Hemorrhage leads to significant morbidity/mortality
- Incidence is 2-4% annually
  - ARUBA trial 2.2%

Many present with seizure or headache

Noninvasive imaging leads to increased diagnosis of unruptured AVM

Angiography is gold standard for evaluation
Arteriovenous Malformations (AVM)

• ARUBA 2014 concluded the natural history of unruptured brain AVMs is better than any form of treatment

• Lots of controversy
  – Wide range of treatment modalities
  – Lack of subgroup analysis
  – Small "n"
  – Insufficient follow-up (<3 years, only funded out to 5 years)

Hong CS et al Clin Neuro Neurosurg 2016;150:133-138
Not all AVMs are the same

• This is a “young person” issue
  – The longer you live, the more likely this will become symptomatic
• Treatments are often tailored to individual and involve multiple specialities
  – Open surgery
  – Stereotactic radiosurgery
  – Endovascular surgery
• Elements that increase risk of bleeding:
  – Associated aneurysms
  – Venous stenosis
  – Infratentorial location
Cavernomas

AKA:
Cavernous angioma
Cavernous hemangioma

Prevalence up to 0.5%
Annual detection of 0.56/100K adults

Clinical Manifestations
Seizures
ICH
Neurologic deficits w/o hemorrhage
Incidental 20-50%

Akers et al Neurosurg 2017;80: 665-680
2 forms of cavernomas

- Genetic basis well established
  - Mutations in CCM1-3
- 20% familial
  - AD, but incomplete penetrance, variable presentation
  - Usually multiple
- 80% sporadic
  - Usually solitary
  - Associated with a developmental venous anomaly (DVA)
  - No germline mutations of CCM genes
  - h/o brain XRT
Recommendations for Genetic Testing

• 3 generation FHx at time of diagnosis, looking for
  – Headache
  – Seizure
  – Stroke
  – Abnormal MRI

• Consider genetic testing of CCM1-3 genes in
  – Multiple CCM without DVA
  – Without h/o brain XRT
  – Positive family hx
Imaging and f/u

- Brain MRI is the gold-standard for diagnosis and followup
  - Should include either a GRE or SWI sequence
  - Catheter angio where ddx includes AVM
- MRI repeated for new/worsened symptoms
  - Looking for new hemorrhage
  - Surveillance followup uncertain
Management overview

• Since the presentation is variable, so is the management
  – IVH and ICH are treated as per standard care
  – Asymptomatic incidental CCM – observe
  – Symptomatic easily accessible CCM - resect
  – Medically refractory seizures- resect
  – Radiosurgery - controversial