Screening for Cerebrovascular Disorders: A Guide for Primary Care

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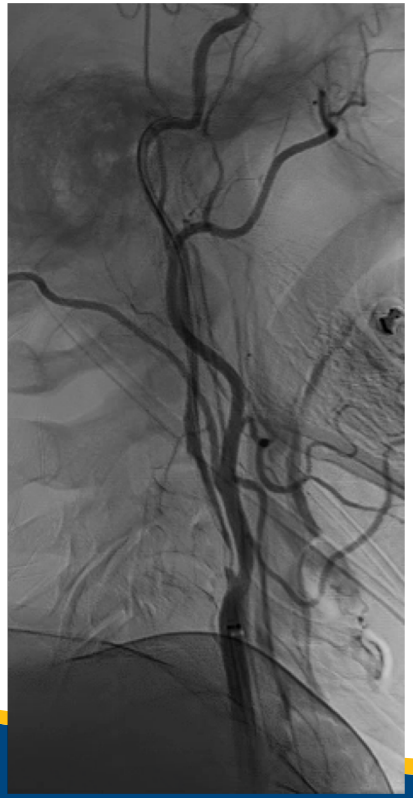
Carotid disease



- 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
- > =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

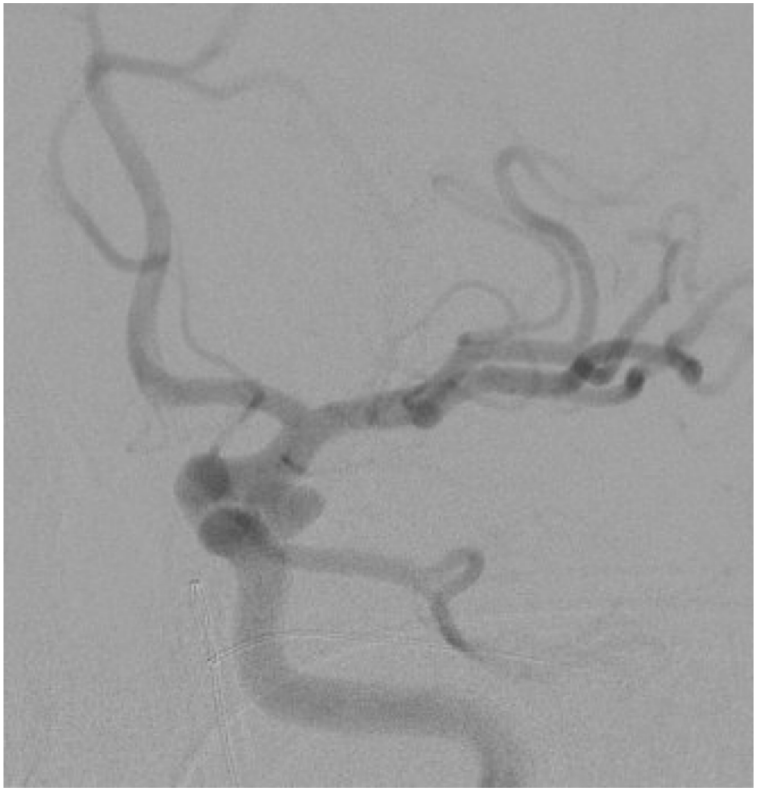
Only a small percentage rupture

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

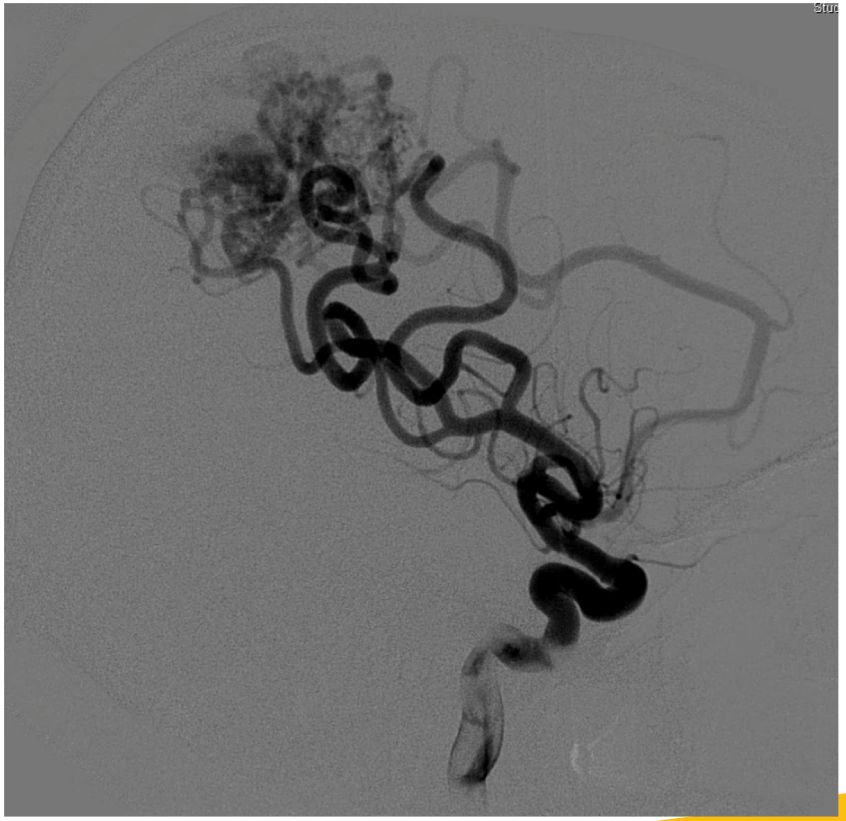
Hemorrhage leads to significant morbidity/mortality

- 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 (<3years, only funded out to 5 years)

Mohr JP et al Lancet 2014;383:614-21
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

Eur Concensus Statement Acta Neurochir 2017;159:1059-64
Ajiboye N et al. Sci World J 2014;http://dx.doi.org/10.1155/2014/649036

Cavernomas

AKA:

Cavernous angioma

Cavernous hemangioma

Prevalence up to 0.5%

Annual detection of 0.56/100K adults

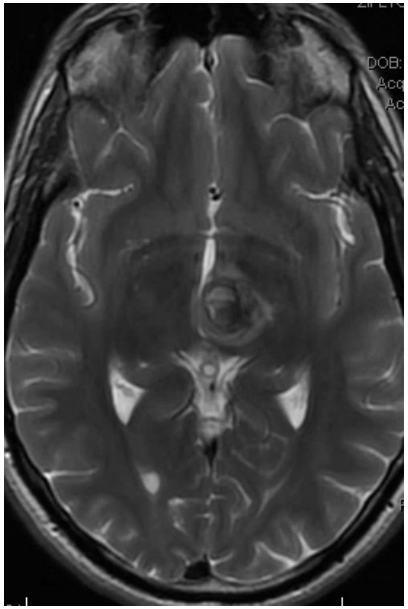
Clinical Manifestations

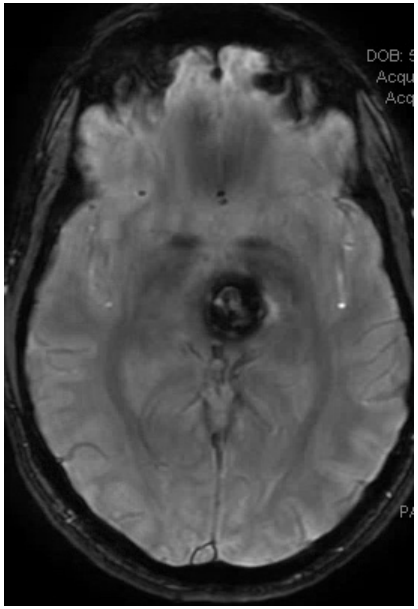
Seizures

ICH

Neurologic deficts w/o hemorrhage

Incidental 20-50%





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