Management of Cerebral Aneurysms in Polycystic Kidney Disease

Dr H Stockley
Consultant Neuroradiologist
Greater Manchester Neuroscience Centre
What is a cerebral aneurysm?

- Developmental degenerative arterial anomaly
- Very thin walled
- Predispose to SAH
Why is a Neuroradiologist talking to you about them?

- Imaging
- Treatment
1. What is the frequency of cerebral aneurysms in the general population?

2. What is the frequency of cerebral aneurysms in ADPKD?

1. What is the risk of rupture of a cerebral aneurysm?

2. What is the best way to detect cerebral aneurysms?

3. What are the risks of treatment?
   - Open neurosurgery
   - Endovascular neurosurgery

4. Should we screen ADPKD patients?
General population

- Rinkel et al 2007
- Meta-analysis
- 2-3% of general population harbours UCA
- Overall risk of rupture ≈1%/year
- 20-25% >1 aneurysm
- Risks of rupture:
  - Increasing age
  - Female gender
  - Location of aneurysm
  - Size of aneurysm
  - Symptomatic
UCAs in PKD

• Prevalence of UCA in ADPKD ≈8% (4-22%)
  • 2-3 x general population

• Prevalence of UCA is higher in ADPKD patients with family history of UCA or SAH
  • 16% vs 6%
  • vs 19% in other ‘familial’ aneurysms

• Prevalence is also higher in patients with previous SAH
  • seen in non-ADPKD patients
Why is the risk increased?

- PKD1 and PKD2 encode for polycystins
- Mouse embryos with mutations in PKD1 and PKD2 exhibit vascular leakage and blood vessel rupture
- Polycystins probably regulate interactions between cells in arterial wall

- Multifactorial
  - Other genes
  - Smoking
  - Hypertension
What is the risk of rupture of a UCA?

• International Unruptured Aneurysm Study (ISUIA) 2003
  • Overall rupture rate 0.45%
  • <7mm 0.07%

• Rinkel 2007 - more robust data
  • Overall rupture rate 1.9%
  • <10mm 0.7%
  • Depends on length of follow-up
    • <5yrs 1.2%, 5-10yrs 0.6%, >10yrs 1.3%
Summary of research

- Rupture risk from small aneurysms is low

- Rupture risk increases with
  - aneurysm size
  - anatomical locations
  - history of previous SAH
  - smoking
  - hypertension

- Not an exact science
Most UCAs are <5mm
Most are anterior circulation
Risk of rupture is small (<0.5%)

UCAs in PKD tend to rupture at a younger age than non-PKD
  • Similar to other ‘familial’ aneurysms
  • 65% occurring before 50yoa
  • BUT often not evident before 30yoa
What happens if a UCA ruptures?
What happens if a UCA ruptures?

- Prevalence of SAH from any aneurysm is 10 in 100,000
- Mortality 30-40%
- Risk of rebleed is highest in first 72hrs; most rebleed within 6hrs
- Complications include hydrocephalus, vasospasm, stroke
What is the best way to detect cerebral aneurysms?

- Catheter angiography
- CT Angiography
- MR Angiography

- Sensitivity of CTA/MRA = 90%
- Smaller aneurysms are harder to detect
- Don’t go looking in the first place
Catheter angiography

- Gold standard
- Invasive, expensive & time-consuming
- Low but defined mortality & morbidity rates
- 0.07% permanent neurological morbidity \( (\text{Cloft et al Stroke 1999}) \)
- 98-99.6% accurate \( (\text{White et al Radiology 2000}) \)
Catheter angiography
CT Angiography

- Close to the accuracy of DSA
- Lower risk
- Fast imaging
MR Angiography

- Poorer resolution
- Longer imaging times
- Flow artefact
- Reduced by contrast enhanced TOF
- Lowest risk investigation
Treatment of cerebral aneurysm

• Surgery, Endovascular or Expectant

• Mortality rates vary between ruptured and unruptured

• Overall around 1 - 2% mortality and 8 - 25% morbidity
  • Any event ≈6%
  • ↑ with size of aneurysm
Surgical clipping

- Application of titanium clip across the base of the aneurysm
- Open operation
- Risks include:
  - Stroke
  - Rupture
  - Seizures
  - Infection
  - Not allowed to drive for 6 weeks
Endovascular coiling

- Deployment of platinum coils directly into the aneurysm
- Less invasive as via the blood vessels
- Risk include:
  - Stroke
  - Rupture
  - Unable to access arterial tree/aneurysm
Risks of treating UCAs

- **Surgical clipping**
  - Mortality - 1.6%
  - Any event - 8.35%

- **Endovascular coiling**
  - Mortality - 0.57%
  - Any event - 3.7%

- **International subarachnoid aneurysm trial (ISAT) 2005**
  - Assessed clipping vs coiling in ruptured aneurysms
  - Coiling was more likely to result in independent survival at 1 year
  - Long term efficacy of coiling uncertain
Should we screen? If so, whom?

The eventual goal of screening is not to detect or to treat an aneurysm, but to increase the number of quality years of life

Rinkel 2008
• Risks and benefits of screening should be weighed up prior to imaging
• Anxiety before screening
• Reassurance of negative result
• Anxiety during surveillance
• Realisation that even screening, repeated screening and preventative treatment cannot prevent all episodes of SAH
• Other considerations - false positives, insurance, unable to drive Group II vehicles
Screening Recommendations

**DO NOT SCREEN**

- UNLESS
  - APKD with family history of aneurysms
  - Severe aneurysm phobia

- These patients should be screened only after referral to multidisciplinary vascular team
  - Counselling
  - Imaging
  - Treatment
  - Screened every 3-5 years
  - Start screening at ≈30yoa
Summary

- Cerebral aneurysms are ‘common’
- But SAH is rare
- Increase in prevalence of UCAs in ADPKD cf general population; clusters in families
- Most UCAs are small; rupture risk is very small
- Treatments are not ‘risk-free’
- Screening should be avoided but can be considered in certain groups