NON-ATHEROMATOUS CAROTID DISEASE

FIBROMUSCULAR DYSPLASIA (FMD)

Medium-sized arteries in young/middle-aged women
Unknown aetiology
Classification: intimal fibroplasia/medial dysplasia= 80%/adventitial fibroplasia
Asymptomatic: conservative (surveillance)
Symptomatic: stroke/TIA/dissection/false aneurysm → as for atheromatous disease
(resect+bypass/dilatation/angioplasty)

ARTERITIS

Takayusu's Arteritis:

Transmural, granulomatous vasculitis causing fibrotic occlusion
Female 7:1

(a)Fever, malaise, arthralgia/myalgia
(b)Neuro symptoms when occluded

Type 1 (arch branches): cerebrovascular/occular
Type 2a (asc aorta, aortic arch, arch branches): cerebrovascular/occular
Type 3 (arch vessels, abdo aorta and branches): 65%; stroke/renovascular disease/mesenteric ischaemia

Management: immunosuppression (steroid/cyclophosphamide/methotrexate)
Surgery: avoid in acute phase; long fibrotic segments so no endarterectomy/angioplasty → bypass (from ascending aorta, not subclavian)

Giant Cell Arteritis:

Commonest primary vasculitis in adults; females 4:1
1) Systemic inflammatory syndrome 2) cranial arteritis 3) large vessel arteritis
Intracranial vessels unaffected;
Malaise, headache, myalgia, jaw claudication (50%); ocular complications (blindness)
Tenderness over temporal artery (50%)
Treatment: steroids

CAROTID ANEURYSM

CAROTID DISSECTION

2% of strokes (20% in young adults)
Spontaneous (FMB), iatrogenic, extension of central dissection (Type A), trauma (ICA crushed between mastoid process and C2 TVP)
Type 1: no stenosis
Type 2: 70-99% stenosis and/or >50% dilatation
Type 3: flamed-shaped occlusion 2-3cm distal to bifurcation

Symptoms
(i)Ipilateral head/neck pain (70%)
(ii)Stroke/TIA (50-75% present thus)
(iii)Pulsatile tinnitus
(iv)Syncope
(v)Occular: amaurosis fugax, oculo-sympathetic paresis, hemianopia, ischaemia optic neuropathy, painful Horner's
(vi)Cranial nerve palsies (3,4,6,7,9,10,12) due to mural haemoatoma compressing them/stretching

Investigations: US/CTA/MRA (dissection starting 2-3cm beyond origin of ICA
Management: conservative with anticoagulation/endovascular for Type 2 if trauma/recurrent cerebral events

CAROTID BODY TUMOUR

Derived from neural crest ectoderm, tumour is nest of neoplastic epitheloid chief cells in in space between ICA and ECA; enlarging tumour splays bifurcation → neck swelling
5% bilateral; 5% malignant
Presents as neck swelling, pain, hoarseness, CN palsies, Horner's
Hormone-mediated syndrome: flushing, dizziness, arrhythmias, hypertension
Diagnosis: USS/MRA/CTA
Diff dx: glomus vagale tumour (doesn't splay bifurcation; resecting → hoarseness/dysphagia)
Management: excision

PRESENTATION OF CEREBROVASCULAR DISEASE

TIA: sudden onset focal deficit with no other apparent cause except vascular cause w/ complete recovery in 24 hrs
Causes: (i)microemboli from thrombi$_{(atherosclerosis, AF)}$ (ii)hypoperfusion $_{eg\ subclavian\ steal}$
Infarction prevented by intact autoregulation and collaterals
Tend to recur and herald CVA

CVA: Sudden onset focal deficit lasting >24 hours *(85% thromboembolic, 15% haemorrhagic)*
Causes: (i)**atheromatous** (low flow, thrombosis, embolism) (ii)**haemorrhagic** (cerebral, cerebellar, SAH)
Also: air, fat, other emboli/arteritis+vasculitis/SLE/SOL: tumour, SDH, EDH/MS plaque/venous infarct
Thromboemboli: cardiac/great vessels/carotids/verebrals
Stroke in evolution: deficit worsens due to haemorrhage, oedema, enlarging infarct

CADASIL: cerebral autosomal dominant arteriopathy w/ subcortical infarcts + leucoencephalopathy

<u>CAROTID TERRITORY</u>
Aphasia (dom hemi)/hemi,mono paresis+sensory loss/transient monocular blindness (TMB)/higher cortical dysfunc.
TMB w/o emb source=ant. isch. optic atrophy (post. cilary art. microvasc disease ->acute ischaemia of optic head)
Diff Dx: stroke mimics and AIOA

<u>VERTEBROBASILAR</u>
Homonymous hemianopia or bilateral blindness/hemi or bilateral motor and sensory impairment
Cerebellar:dysarthria/diplopia/nystagmus/ataxia/ /vertigo/dizziness
Transient global amnesia: several hours; amnesia with complete recovery

Isolated dizziness/vertigo/diplopia = something else
**VB TIA stroke risk similar to carotid esp if 50-99% stenosis (30% 90 day risk)

<u>NON-HEMISPHERIC</u>
Isolated syncope, presyncope, isolated dizziness, isolated diplopia, isolated vertigo
Must exclude cardiac or inner ear pathology

Diff Dx:
FOCAL EPILEPSY: characteristic progression/irritative phenomena
MIGRAINE FOCAL PRODROME: Headache/visual disturbance/neurology crosses territories

<u>History</u>
Pc:
HPc: identify dominant hemisphere → identify laterality of lesion
PMHx: MI/DM/HTN/AthSc/arteritis/thrombotic disorders
FHx: as for PMHx
Medx: primary/secondary prevention;
smoking/EtOH
SHx:

<u>Examination</u>
Physical Exam: full neuro exam/evidence of embolus: carotid, subclavian bruit/AF/valve disease

INVESTIGATION OF CAROTID DISEASE

1. HISTORY & EXAMINATION (neuro exam, full CVS exam including PVS)
2. BLOODS: fasting lipids/glucose
3. BEDSIDE: ECG $_{(AF)}$, ECHO, BP
4. IMAGING

DUPLEX ULTRASOUND

Assess disease extent for CEA

Good: can operate off US only (need second sonographer to corroborate)

Bad: identifying "vulnerable plaque" difficult/ only sees cervical CA/heavily calcified plaque confounds measurement

MRA

Wider view (can see aortic arch, inflow, outflow and intracranial vessels) to assess **stent suitability**

nephrogenic systemic fibrosis

CTA

For 70-99% stenosis, CTA has highest specificity (94%, above CEMRA and then US)

Good: minimally invasive, anatomical information, more accessible than MRA, well tolerated, less artefact

Bad: contrast, radiation, no dynamic info, trickle flow not reliably seen, calcification impairs estimation of severity

ARCH ANGIOGRAPHY

Assess anatomical suitability of CAS

Stroke/death risk 1.5% → not part of routine workup

INITIAL MANAGEMENT

1. BEST MEDICAL TREATMENT/SECONDARY PREVENTION

(i)**Antiplatelets** (not if AF/flutter);: **Clopidogrel 75mg** first-line aspirin 75mg OD/dipyramidole 20mg BD second-lines

(ii)**Anticoagulation** (AF/flutter/cardioembolic source): **warfarin**/thrombin inhibitor/Xa inhibitor

TIA: immediate anticoag

CVA: 14days aspirin 300mg OM then anticoag

(iii)**Lipids: atorvastatin** 20-80mg ON (aim for 40% HDL reduction)

(iv)**Blood Pressure**: ACEi/CaChB/Thiazides → SBP <130mmHg (allow 140-150 in severe bilateral disease)

(v)**Lifestyle**: diet and exercise

2. SPECIALIST REFERRAL FOR CEA (NICE 2008)

TIA w/ **ABCD^2 score 4-7** high risk → specialist assessment and investigation **within 24 hours** of symptom onset

0-3 low risk → specialist assessment and investigation **within 7 days** of symptoms

*treat crescendo TIA as high risk

*treat late presenters (>7d post symptom onset) as low risk)

*ref. decision should never be based on bruit; presence/absence of bruit does not correlate w/ degree of stenosis**

DEFINITIVE MANAGEMENT

1. CAROTID ENDARTERECTOMY (CEA)

(a)Symptomatic disease
Within 2 weeks of symptom onset (up to 3 months)
(i)TIA with 50-99% NASCET stenosis (men)/70-99% NASCET stenosis (women)
(ii)Non-disabling CVA

Urgent (<24hrs): stroke in evolution/crescendo TIA/stuttering hemiplegia

(b)Asymptomatic disease
Trials: CETC report 5 year results from VA, ACAS and ACST trials

Complications: Death 3%, CVA 3%, CN injury (VIIm, X, XII) 8% (reduce by LA, shunt, patch)
Death + VA 30d rates same for LA and GA

CEA before or after CABG?
*CABG then CEA: MI 0.9%, CVA 6.3%, death 2% *lowest MI but highest stroke**
*CEA then CABG: MI 6.5%, CVA 2.7%, death 3.9% *lowest CVA but highest MI**
*CEA + CABG: MI 3.6%, CVA 4.6%, death 4.6% *high rates of all three, avoid**

2. CAROTID ARTERY STENTING

Symptomatic disease only (no role in asymptomatics)
Useful if CEA too difficult/dangerous: prev neck surgery, DXT, short neck, high carotid bifurcation
Contraindications: absolute= occluded ICA, visible mural thrombus
relative= tortuosity
Pros: local anaesthesia so useful if comorbid; less injury risk
Cons: higher CVA rate than CEA

ASYMPTOMATIC CAROTID STENOSIS TRIALS

1. VETERAN AFFAIRS
400 men 50-99% NASCET stenosis randomised to (i)aspirin alone (ii)aspirin + CEA
Short= no difference in CVA/death at 30d
Long= Nonsignificantly lower ipsilateral stroke in surgery group (4.7% v 9.4%)

2. ACST
3000 adults 60-99% NASCET stenosis randomised to (i)immediate CEA + BMT (ii) later CEA when symptoms + BMT
Short= 30d CVA rate after CEA 3.1%
Long= halved overall CVA/death rate (6.4% vs 11.8%) (benefit not seen until 2 yrs postop)

3. ACAS
1600 adults 60-99% NASCET stenosis randomised to (i)aspirin alone (ii)aspirin + CEA
Short= lower short term CVA/death rates with CEA
Long= CEA had lower 5yr ipsilateral CVA/death rates (5% vs 11%)

WOMEN= CEA doesn't benefit asymptomatic women (ACAS, ACST meta-analysis)
TIMING: (i)max benefit within 2 weeks (ii)not beyond 3 mths (NNT125 w/ ARR 0.8% at 5 years)
BENEFIT BY GROUP (i)50-69% NASCET ARR 7.8% NNT 13 at 5yrs
(ii)70-99% NASCET ARR 15.6% NNT 6 at 5yrs

VERTEBROBASILAR REVASCULARISATION

15-25% of ischaemic strokes; no major RCT like ECST/NASCET
NE Posterior Circulation Registry: 40% embolic (60% cardiac/40%arterial), 32% haemodynamic. 28% misc (trauma/aneurysm, arteritis, compression against osteophyte etc)

Management options:
Angio +/- stent: 99% technical success/30d stroke 1.1%/1% recurrent stroke 2 yrs/restenosis 3x higher in BMS vs DES
Surgery: open recon with vein bypass/CCA transposition

SUBCLAVIAN STEAL: SCA origin stenosis/occlusion with retrograde flow in ipsilateral vertebral on upper limb exertion; forearm claudication and VB ischaemic symptoms
Treatment: angio first, surgery second

CORONARY STEAL: as for subclavian steal but affects internal mammary used to bypass coronary artery
Treatment: angio first, carotid-subclavian bypass second