ACHALASIA

Functional motility disorder due to (i)failure of LOS to relax and (ii)impaired secondary peristalsis in distal oesophagus

Epidemiology:
A: 30-50 G: ESI

Aetiology:
1° Idiopathic = progressive loss of Auerbach's myenteric plexus inhibitory ganglia (NANC)
2° Trypansoma cruzii parasitic infection (Chagas' disease)/antireflux surgery/truncal vagotomy (historic)

Pathogenesis:
EARLY: Vigorous Achalasia (inibitiory NANC ganglia lost → high amplitude non-peristaltic contractions resembling D.O.S.)
LATE: Classic Achalasia (progressive loss of excitatory cholinergic neurons → dilatation + low amplitude contractions in oes body)

Clinical features:
1. Dysphagia: solids and liquids/intermittent/chronic *(cf cancer: absolute/progressive/rapid/more marked wt loss)*
2. Regurgitation
3. Odynophagia
4. Weight loss

ECKHARDT SCORE: 0=occasionally 1=daily 2=several times daily 3=every meal; remission = score 3 or less over 6mths

Investigations:
1. X-ray: dilatation with retrocardiac fluid level
2. Barium Swallow (i)Bird's beak deformity(hold up distally + dilatation) (ii)Impaired secondary peristalsis
3. OGD: *Evaluate dysphagia + therapeutics* (i)tight LOS (ii)dilated oesophagus
4. HR manometry: *Gold standard + predicts recurrence by 3 subtypes*

MANAGEMENT

Aim: reduce LOS/GOJ pressure
Can be managed, not cured: cannot restore oesophageal motility as myenteric plexus is gone, gone, gone.

MEDICAL
1. DRUGS: *Only for frail/mild symptoms* Nitrates/CaChB's
2. INTRASPHINCTERIC BOTOX (100iU→LOS) *High risk patients not suitable for intervention* (32% symptom relief at 12mths)

ENDOSCOPIC
1. PNEUMATIC BALLOON DILATATION: 70% symptom relief at 12mths || 5% perforate
2. POEM (PER ORAL ENDOSCOPIC MYOTOMY) Submucosal tunnel 10-15cm proximal to GOJ → 3cm distal to GOJ
Pros: longer myotomy than LHM/less GOR(intact PE lig)/less vagus injury/less pain/less bleeding/faster recovery
Cons: perforation/difficult after multiple PBDs(LOS muscle layer adhesions)

SURGICAL
LAPAROSCOPIC HELLER'S MYOTOMY + PARTIAL[impaired 2ndary peristalsis] **FUNDOPLICATION**(Toupet/Dor)
5 ports; cut 6cm prox and 2 cm distal; fundo for reflux/cover leak; post-op barium for mucosal breach?
European Achalasia Trial: LHM = PBD at 5yrs with fewer needing re-dilatation
Pros: definitive treatment ||Cons: tradeoff of dysphagia vs GOR

FOLLOW UP

End-point: (i)symptom relief and (ii)50%/min emptying on TBE
GOR: 30% in POEM/LHM but mild and PPI-amenable

OTHER MOTILITY DISORDERS

DIFFUSE OESOPHAGEAL SPASM

Unknown aetiology
Severe chest pain and dysphagia
Barium= corkscrew oesophagus
HRM=**high amplitude/duration** waves but **aperistaltic** *cf nutcracker*
Treatment: nitrates/CaChBs/PDEis/botox/long myotomy

NON-SPECIFIC

Crico-pharyngeal achalasia: UOS
Nutcracker oesophagus: **high amplitude** contractions with **normal peristalsis**
Hypertensive LOS (>45mmHg): dysphagia + chest pain
Oesophageal diverticulum: most **pulsions** (motility disorder → distal obstruction); **HRM** then **excise** diverticulum
Autoimmune disease: SSc (80% involved, smooth muscle atrophy → weak peristalsis and reflux)
PM/DM (weak striated muscle in upper third → aspiration
SLE: similar to SSc
PAN
RhA: aretynoid arthritis