Οξέα Αορτικά Σύνδρομα: επιδημιολογία, ταξινόμηση και διάγνωση

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No conflict of interest to declare in relation to this talk
Acute Aortic Syndromes (AAS): the aortic catastrophe

AAS – new term for an ‘old’ disease

• 1760: 1st case report (King George II of England)
• 1955: 1st successful op repair (DeBakey)

- In harmony with modern cardiology terms
  • acute coronary syndromes / acute chest pain syndromes
  • aortic emergencies
- To include the diverse aortic pathology (> dissection)
- Update was needed because of great evolution in
  • imaging modalities (new insights in pathophysiology)
  • interventional and surgical techniques
AAS - knowledge from registries
- few guidelines / recommendations

- single-center registries until IRAD
- IRAD
  > 2,000 patients
  26 centers
  11 countries
  many publications since 1996
AAS - epidemiology

- **UNCOMMON disease**
  - 100-200 times less common vs CAD
  - 2-3 times more common than rupture of the abdominal aorta
  - Incidence LARGELY UNKNOWN - difficult to define (SCD or early death)
    - Autopsy-based: up to 1-3 %
    - USA: 2-3.5 cases/100,000 patients/year
    - 3 cases for every 1,000 patients coming to A&E with chest pain

- The most common FATAL condition in patients with chest pain
  - Mortality STILL VERY HIGH today with/out treatment
    - ≈ 40% die before arrival to hospital (vs 25% with ruptured AAA)
    - ≈ 40% of cases are ‘missed’ at the time of presentation
      50% are diagnosed after >6h in Europe and >12 h in US
    - BUT 1% mortality / hour after symptom onset (Hirst & Kime 1958)
    - ≈ 35-40% of pts arriving to hospital will die even if center is experienced
      of those operated: 5-20% die shortly after operation
AAS – pathophysiology

Heterogeneous group of patients with similar clinical profile inter-related conditions with different pathophysiological mechanisms

Classic Aortic Dissection (AoD) 80%

Intramural Hematoma (IMH) 15%

Incomplete Dissection (ID)

Penetrating Atherosclerotic Ulcer (PAU) 5%

Aortic rupture

• Mostly separate
• May progress to another type
• May co-exist
• Often difficult to differentiate
• May stabilize or rupture
**Classic AoD (class I)**

- Entrance tear (sites of high hydraulic stress)
  - right lateral wall of asc aorta
  - proximal desc
- Flap (intimo-medial)
- False lumen (blood separating medial layers)
- Re-entrance tear (absent or not seen)

**Dx with CT/TOE/MRI**

- aortic flap (flow through)
- double channel aorta
  (FL usually > TL)
IMH or intra-aortic hematoma (IAH) (class II)

5-25% of AAS
- AoD without a tear – noncommunicating type of AoD
- Hematoma caused by hemorrhage into the media (rupture of vasa vasorum)

- Probably more often DISTAL
- Older people

Dx with CT/TOE/MRI
- No flow within hematoma / No enhancement with contrast on CT
- TOE: typical image density ≈ thrombus reduced diameter of TL central displacement of intimal calcification
PAU  (class IV)
2.3 – 7.6% of AAS
- Ulceration of an aortic ATEROMATOUS lesion - penetrates the internal elastic lamina into the media
- With intramural-subadventitial hemorrhage (TL>FL)
- Usually FOCAL in DESCENDING aorta (may be >1)
- Degree of risk for rupture

Dx with CT/TOE/MRI /aortography
- Typical image
- Atheroma present
Incomplete Dissection or subtle or discrete (class III)

- Laceration of intima & part of the media
  - BUT separation of medial layers is not so large
  - after the acute phase the bare area is covered with fibrous tissue and endothelium

- Mostly ASCENDING aorta
  - Posterior wall, above L coronary ostium
  - May disturb aortic cusps → AR

- Dx is DIFFICULT - CAREFUL imaging – ?aortography
  - Subtle eccentric bulge at the tear site
  - Resembles IMH
    - no typical flap separating TL-FL
    - but aorta looks dilated (vs IMH: reduced TL)
  - Presence of AR, periaortic fluid
Traumatic aortic rupture (class V)

- 18% of road traffic accident deaths
- 2nd most frequent cause of death
- 1st cause of death in < 40 years old
- < 15% reach hospital alive (<2% survive)
AAS – classification

Very important for decisions on intervention and prognosis

De Bakey Type I  Type II  Type III
Stanford  Type A  Type B

Ascending aorta +/- Arch  Proximal AAS  Distal AAS

Descending aorta

60%  10-15%  25-30%
Proximal AAS
Ascending aorta mortality:
30% surgery vs 60% medical tx

Distal AAS
Descending aorta mortality:
10% medical tx vs 30% surgery
### AAS – risk factors / conditions predisposing

<table>
<thead>
<tr>
<th>Increased aortic wall stress</th>
<th>Hypertension (moderate/severe)</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>Smoking, dyslipidemia</td>
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<tr>
<td></td>
<td>Cocaine, crack, amphetamines</td>
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<td></td>
<td>Pheochromocytoma</td>
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<td>Weight lifting / Valsalva maneuver</td>
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<td>common a Hx of extreme exertion or emotional stress prior to onset</td>
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<table>
<thead>
<tr>
<th>Abnormalities of aortic media</th>
<th>Genetic / Hereditary syndromes</th>
</tr>
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<tbody>
<tr>
<td>IATD: most common for AAoD in young</td>
<td>Connective Tissue: Marfan (5% of AAS but 50% in &lt;40yrs)</td>
</tr>
<tr>
<td></td>
<td>Ehlers-Danlos, Loeys-Dietz, Turner</td>
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<td></td>
<td>Bicuspid AoValve (10 times higher risk), coarctation</td>
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<td>Familial thoracic aortic aneurysm (13-19% of TAA) and AoDissection</td>
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</table>

### Atherosclerosis

### Vascular inflammation

- Autoimmune (Giant-cell, Takayasu arteritis, Bechet’s, Ormond’s)
- Infection (TB, syphilis)

### Other

- Pregnancy, chronic immunosuppression, polycystic kidney disease
- Known aneurysm (16%)

### Other

- Trauma (deceleration or torsional injury): Car accident, fall, sports trauma
- Iatrogenic Intervention (Catheter, instrument) / Surgery (valve, aorta, grafts)
AAS – clinical presentation similar for all types!

A) PAIN / chest discomfort (96% IRAD)

- Aortic chest pain is usually very typical
  - can be recognised rapidly
  - differential diagnosis from usual causes of acute chest pain
- Severely intense (max intensity at onset ≠ ACS) 90%
- Acute onset 85%
- Tearing or ripping
- Pulsating and migratory
- Radiating to anterior chest, neck, jaw: proximal AAS (asc aorta) back, abdomen: distal AAS (desc aorta)
- **SILENT only in 5-10% of pts:** older / Marfan / on steroids other presentation (syncope, CVA, HF)
  higher mortality

B) CARDIAC complications

C) PERFUSION DEFICITS and end-organ ischemia
B) CARDIAC complications
mostly with type A
increased mortality and serious complications

- **Acute AR** very common in type A 40-75%
  various mechanisms: root dilatation, prolapse of flap
dissection extending in cusps

- **Myocardial ischaemia-infarction** 10-15%
dissection extending in the coro ostium or
FL compressing proximal coro artery- usually RCA

- **Pericardial effusion and tamponade**
  transudation via thin FL wall or dissection extending in pericardium

- **HF and shock**

- **Syncope** common 15-20% (with or without pain)
  multifactorial: cardiac, vascular, neurologic, volume-related
AAS – clinical presentation (III)

C) PERFUSION DEFICITS and end-organ ischemia

JACC 2010
AAS – physical examination very variable

- Hypertension is common
- Low BP is an ominous sign

- Very common in type A
  AR murmur
  Pulse deficits
  Arm BP difference >20 mmHg
- With both pulse deficits & BP difference),
  probability of AoD ≈100%
- Both associated with more complications
  and increased mortality

- Limb ischemia is associated with end-organ
  ischemia (GI, brain, heart) and increased
  mortality (45% vs 15%)
- Pulses should be checked in ALL limbs

Physical findings of 591 patients with type A AoD (IRAD)
AAS – diagnostic assessment

First important step: estimation of the risk for an AAS
class I recommendation (ACC/AHA guidelines 2010)

High-risk conditions (History)
- Marfan, vascular Ehlers-Danlos, Turner, other CTD
- FHx of TAA / AoD
- Known AoV disease or TAA
- Recent aortic manipulation

High-risk features (Symptoms)
- Pain of abrupt onset
- Pain very severe
- Pain of tearing, ripping, stabbing or sharp
- Syncope or acute neurological complaint: check again

High-risk signs (Focused examination)
- Pulse deficits
- Arms BP difference >20mmHg
- Focal neurological deficit
- New AR murmur
- Low BP or shock

Screening tests: alternate dx or suspect AAS

EXPEDITE aortic imaging and seek surgical advice

0 present risk low
1 present intermediate
≥2 present high
AAS – diagnostic assessment: aortic imaging

CT / TOE / MRI: class I in all high-risk patients

- **DIAGNOSIS** of AAS (safe and fast)
- definition of the **ANATOMY** to guide tx
  - type - extent – classification - branches involved - complications

**CT, TOE, MRI:**
acceptable
diagnostic accuracy

**Meta-analysis**
Equally reliable pooled sensitivity (98%-100%) and specificity (95%-98%)

*Arch Intern Med. 2006;166:1350-1356*
AAS – aortic imaging

Use of imaging methods in real world

- CT is the 1st commonest method used for the dx
- TOE is the 2nd most used
- 70% of patients will need >1 methods for the diagnosis (mean: 1.8)

Aortography used very rarely in some difficult cases
- Fail to dx with other techniques but high clinical suspicion
- Need experienced operator
- ? unstable pts

Moore et al (IRAD), AJC 2002
Echocardiography in aortic disease: EAE recommendations for clinical practice. Evangelista et al, EJE 2010
AAS – which imaging modality to choose?

The modality that will provide, in any individual setting, the most accurate and reliable answer and as fast as possible depending on

- **Imaging equipment**: immediate availability and quality
- **Doctor to use the imaging techniques**: skill and expertise
- **Patient**: stability, ability to get sedation, allergies, renal function

**ACC / AHA recommendation**

1. (C) the selection of method should be based upon patient variables and institutional capabilities
2. (C) if clinical suspicion is high and 1st aortic study (-), then a 2nd study should be done
Acute Aortic Syndromes – key messages

AAS: ‘Uncommon’ but FATAL condition

EARLY diagnosis is crucial

• Thorough Hx and examination (risk assessment)
  (many atypical findings, no good screening tests)

• Appropriate use of imaging (diagnosis and anatomy)

Early activation of Heart Team for best treatment

• Cardiologists (Non-invasive and Invasive)
• Radiologists
• Cardiac surgeons
• Vascular surgeons
• Anaesthetists
Σας ευχαριστώ πολύ την προσοχή σας
BACK-UP SLIDES
# AAS – aortic imaging

<table>
<thead>
<tr>
<th></th>
<th>(+) Advantages</th>
<th>(-) Disadvantages</th>
</tr>
</thead>
</table>
| **MDCT** | **Available** (widely, readily)  
**Fast** acquisition (<30s) / unstable pts  
**Sensitivity** 93-100%  
**Excellent anatomic** detail (branches)  
**Post-image processing** - 3D reconstruction  
Largely **operator-independent** | **IV contrast** (allergy-kidneys)  
? Radiation (f-up)  
**Image artifacts**  
- aortic root: ECG gating  
- overestimation of aortic size  
- may miss the entry tear  
No info on AoV, coronaries (TTE) |
| **TOE**  | **Unstable pts – bedside**  
**Available readily** – immediate info  
**No contrast** – radiation  
**Sensitivity** high  
Assess LV, cardiac complications (TTE)  
Differentiate FL-TL (Doppler) | **Operator dependent**  
**Poor delineation of arch vessels**  
Lower sensitivity for type B  
(below diaphragm)  
**Semi-invasive** – sedation, secure airway, technically difficult |
AAS –
differentiate between true and false lumen

**TL – true lumen**

1. < false lumen
2. Systolic expansion
3. Systolic antegrade flow
4. Flow early and fast
5. Rare thrombus

**FL – false lumen**

1. > True lumen
2. Systolic compression
3. No or reduced systolic antegrade flow or retrograde
4. Flow delayed and slow
5. Thrombus frequent
Classic AoD

- Evolution
  - Longitudinal progression (back in the true lumen)
  - Partial / total thrombosis of the FL (if no re-entrance)
    - especially early
  - Early rupture of the FL external wall (usually aorta → hemopericardium, tamponade, death)
  - Slow oozing → periaortic hematoma (impending rupture)
  - Acute compression of TL → severe distal ischemia
IMH or intra-aortic hematoma (IAH)

- Evolution: unpredictable
  - Aortic rupture (complete or contained) (20-50%???)
  - Dissection localised incomplete or complete or ulcer (30-50%???)
  - Localised remodeling and aneurysm
  - Growing-progression or stable
  - Spontaneous resolution without sequelae (distal) -10%
PAU (class IV)

- **Evolution: unknown**
  - Slow progression to aneurysm
  - Pseudoaneurysm (contained aortic rupture)
  - Complete rupture (more often than AoD)
  - AoD via the crater (becomes the entrance tear) – 40%
  - Peripheral emboli
AAS – clinical presentation

Peak at early morning  Increase in winter

IRAD. Mehta et al, Circulation 2002

- Hyperacute  symptom onset  <24 hours
- Acute  2-7 days
- Subacute  8-30 days
- Chronic  >30 days
AAS – diagnostic algorithm

- Age: 55-65 years
- Male gender
- HT present (70-80%)
- Other risk factors present
  - Marfan, FHx, known aneurysm, BAV
- Atherosclerosis in type B
- Also common a Hx of extreme exertion or emotional stress prior to onset

Diagnosis may be more difficult
- Young < 40 years (not expected)
- Elderly > 70 years (signs are not present)
- Women (older, less pain, fewer signs, altered consciousness)
AAS – diagnosis based on imaging criteria

Table 1  Distinguishing features of true and false lumen in classic aortic dissection

<table>
<thead>
<tr>
<th>Features</th>
<th>True lumen</th>
<th>False lumen</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total diameter</td>
<td>Small</td>
<td>Large</td>
</tr>
<tr>
<td>Diameter in systole</td>
<td>Increase</td>
<td>Decrease</td>
</tr>
<tr>
<td>Thrombosis</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Spontaneous contrast</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Aortic flow</td>
<td>Laminar, early systolic</td>
<td>Swirling, turbulent, late systolic</td>
</tr>
<tr>
<td>Beak sign</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Cobweb sign</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Three lumen sign</td>
<td>Central</td>
<td>Peripheral</td>
</tr>
<tr>
<td>External wall (thickness)</td>
<td>Normal</td>
<td>Thin</td>
</tr>
</tbody>
</table>

Table 2  Imaging criteria to distinguish classic aortic dissection, intramural aortic haematoma (IAH) and incomplete dissection

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Classic dissection</th>
<th>IAH</th>
<th>Incomplete dissection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dissection flap</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Double aortic lumen</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Entrance tear</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Aortic wall thickening</td>
<td>No</td>
<td>Yes*</td>
<td>Yes</td>
</tr>
<tr>
<td>Decreased aortic lumen</td>
<td>Yes</td>
<td>Yes†</td>
<td>No</td>
</tr>
</tbody>
</table>

*In many instances an intimomedial tear (stellate or linear discontinuity of the internal aortic layers) or a subtle aortic bulging can be seen; †in some cases, IAH may not necessarily compress or significantly diminish the aortic lumen.
Acute Aortic Syndromes (AAS) – the basics of the aortic catastrophe

Structure of the presentation

• Terminology
• Epidemiology
• Pathophysiology - Classification
• Brief overview
  • Clinical presentation
  • Diagnosis
AAS – pathophysiology

‘Old’ nomenclature

Classes of intimal tear

I. AoD
II. IMH
III. ID
IV. PAU
V. Iatrogenic / traumatic
AAS – diagnostic assessment (II): Labs

- Hb, Ht, WBC, LFTs, U&Es
- Blood type - Antibody screen
- Cardiac enzymes – Troponins

Biomarkers
- Plasma SM myosin heavy chain protein
- Calponin, serum elastin fragments
- D-Dimers

D-Dimers
- ↑↑ in AoD (???? in limited IMH or thrombosed FL)
  In other conditions (VTE, ACS, sepsis, trauma, surgery)
- Meta-analysis (11 studies): **pooled sensitivity 94% but specificity 40-100%**
- No data from prospective studies
- **Cannot be used to ‘rule-out’ the disease**

No ACC / AHA recommendation (no large prospective trials)
AAS – diagnostic assessment (III): ECG

- Normal 30%
- Non specific abnormalities common
  ST-T changes, prior MI, LVH

- But also ACS - acute STEMI
  in 5-10% of type A AoD
  involving mainly RCA

ACC / AHA recommendation: class I

If there is ST elevation suggestive of AMI but the patient is at high-risk of AAS, then aortic imaging should be done FIRST

Role of TTE in such cases
AAS – diagnostic assessment (IV): CXR

- Abnormal 60-85%
  - widened aorta-mediastinum
  - abnormal cardiac contour
  - pleural effusion
  - displaced aortic calcification

- But can be NORMAL in up to 20%
  - likely proximal asc aorta
  - cannot be used to exclude dx if clinical suspicion is very high!

ACC / AHA recommendation

I (B)  CXR depending on the patient’s risk for AAS
I (C)  CXR in all low and intermediate-risk
III  in high-risk patients, a (-) CXR should not delay definitive aortic imaging
AAS – aortic imaging - MRI

Many Advantages
• Gold standard
• 3D reconstruction
• High resolution
• Dynamic information
• No contrast – radiation

- Chronic aortic dissection / Follow-up
- If all other techniques fail

Important Disadvantages
• Limited availability
• Time-consuming
• Not suitable for unstable patients
• Accuracy for branches 82%

Litmanovich et al, AJR 2009:193:928