Aortic Pathology
Diagnosis and Management

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Learning Objectives

As a result of participating in this live learning activity, you should be able to:

1. Define thoracic, thoracoabdominal, and abdominal aneurysms
2. Risk factors for aneurysmal development
3. AHA/ACC guidelines for diagnosis, treatment, and surveillance of aortic aneurysmal disease.
4. Define aortic dissection and risk factors for development.
5. AHA/ACC guidelines for diagnosis, treatment, and surveillance of aortic dissection.
The Aorta

- **Thoracic**
  - Aortic Root
    - Aortic valve annulus
    - Aortic valve cusps
    - Sinus of valsalva
  - Ascending aorta
    - Sinotubular junction to origin of brachiocephalic (innominate) artery
- **Arch**
  - Origin of brachiocephalic
  - Includes great vessels
- **Descending Aorta**
  - Begins at isthmus between origin of left subclavian artery and ligamentum arteriosum
  - Through the diaphragm
- **Abdominal aorta**
  - Visceral
  - Juxtarenal
  - Infrarenal
  - Aortic bifurcation
What is an aneurysm?

• Definitions

  o Aneurysm- a permanent localized dilation of the artery that is 50% greater than its normal diameter.
    ▪ Involves all 3 layers of the arterial wall (intima, media, adventia)
    ▪ Caused by medial degeneration

  o Pseudoaneurysm- disruption in the arterial wall with extravasation of blood contained by periarterial connective tissue and not all layers of the arterial wall.
Types of aneurysms

- Fusiform - symmetrical bulge around the circumference of the aorta (most common)
- Saccular - asymmetrical bulge which appears on one side of the aorta
- Pseudoaneurysm - enlargement of only the outer layer. Commonly caused by prior surgery or trauma
Thoracic Aortic Aneurysm (TAA)

- Normal Aortic Diameters (aneurysm diameter)
  - Aortic root
    - Female 3.5-3.72 cm (5.2-5.5 cm)
    - Male 3.63-3.91 cm (5.4-5.8 cm)
  - Ascending
    - 2.86 cm (4.2 cm)
  - Mid Descending
    - Female 2.45-2.64 cm (3.6-3.9 cm)
    - Male 2.39-2.98 cm (3.4-4.3 cm)
  - Distal descending
    - Female 2.40-2.44 cm (3.6 cm)
    - Male 2.43 - 2.69 cm (3.6-3.9 cm)

- Risk Factors for development
  - Hypertension
  - Hypercholesterolemia
  - Smoking
  - COPD
  - Concomitant aneurysm (Cerebral, Abdominal)
  - Bicuspid aortic valve, Coarctation
  - Genetic syndromes
    - Marfan syndrome, Loeys-Dietz, Ehlers-Danlos, Turners
  - Inflammatory diseases
    - Takayasu arteritis, giant cell arteritis
  - Family history of aortic aneurysm or dissection
Clinical Presentation

- Thoracic aortic aneurysms are asymptomatic
- Symptoms can occur if surrounding structures compressed
  - Hoarseness: left recurrent laryngeal nerve stretching
  - Stridor: tracheal or bronchial compression
  - Dyspnea: lung compression
  - Dysphagia: esophageal compression
  - Plethora and edema: SVC compression
  - Aortic regurgitation and heart failure: Aortic root / Ascending dilation
- Diagnosis
  - Commonly found incidentally
  - CXR, CT scan, MRI, or Echocardiogram
Management

• AHA/ACC 2010 Guideline: Medical treatment for patients with small aneurysms not requiring surgery and patients who are not candidates for surgery (Class I Level C)
  o Stringent control of hypertension
  o Lipid profile optimization
  o Smoking cessation
  o Atherosclerosis risk-reduction

• Recommended Blood Pressure control (Class I Level B)
  o Antihypertensive therapy
  o goal BP < 140/90 (patients without diabetes)
  o < 130/80 (patients with diabetes or CKD)
Management Continued

- Beta blocker therapy (Class I Level B)
  - Administered to all patients with Marfan syndrome and aortic aneurysm to reduce rate of growth unless contraindicated

- Beta blockade and ACE-I or ARB (Class IIa Level B)
  - To control BP in setting of thoracic aneurysm to the lowest point patient can tolerate without adverse effects

- ARB and Marfan syndrome (Class IIa Level B)
  - Reasonable to reduce the rate of aortic dilatation

- Dyslipidemia (Class IIa Level A)
  - Treatment with statin to achieve LDL < 70 mg/dL
    - Noncoronary atherosclerosis, aortic aneurysm, and coexistent coronary heart disease
Surgical Indications

- Asymptomatic patients with ascending aorta or aortic sinus diameter of 5.5 cm or greater should be evaluated for surgical repair (Class I Level C)
- Patients with Marfan syndrome or genetically mediated disorders should undergo elective repair at smaller diameters 4.0-5.0 cm depending on condition and to avoid acute dissection or rupture. (Class I Level C)
- Rate of growth > 0.5 cm/y in an aorta < 5.5 cm (Class I Level C)
- Patients undergoing AV repair or replacement and have an aortic root or ascending aorta > 4.5 cm should consider concomitant repair (Class I Level C)
• Repair or replace aortic root (sinuses) or replace ascending aorta is indicated in asymptomatic patients with BAV if 5.5 cm or > (Class I Level B)

• Repair or replace aortic root (sinuses) or replace ascending aorta is reasonable in asymptomatic patients with BAV if 5.0 cm or > and additional risk factor (Class IIa Level B)
  o Family hx aortic dissection
  o Rate of growth ≥ 0.5 cm/y
  o Low risk for surgery and Surgery at an experienced aortic center

• Replacement of ascending aorta in patient with BAV undergoing AVR for severe AS or AR when the diameter is greater then 4.5 cm (Class IIa Level B)
Abdominal Aortic Aneurysm (AAA)

- Normal Aortic Diameter (Aneurysmal)
  - Supraceliac
    - Female 2.7 cm (4.0-4.1 cm)
    - Male 3.0 cm (4.5 cm)
  - Suprarenal
    - Female 2.7 cm (4.01-4.1 cm)
    - Male 2.8 cm (4.2 cm)
  - Infrarenal
    - Female 2.2 cm (3.3 cm)
    - Male 2.4 cm (3.6 cm)

- Risk Factors
  - Smoking
  - Male gender
  - Advancing age
  - Caucasian race
  - Atherosclerosis
  - Family history AAA
  - Other aneurysms (iliac, femoral, popliteal, cerebral)
  - Connective tissue disorder
  - H/o aortic dissection
  - h/o aortic surgery or instrumentation
Clinical Presentation

- Usually asymptomatic
- Pulsatile midline mass
- May have abdominal, back, or flank pain
  - Could be sign of impending rupture
- Symptoms can be similar to other diseases
  - If known AAA then should be presumed the aneurysm until proven otherwise
- Risk of rupture
  - Large initial diameter > 5.5cm
  - Current smoking
  - Hypertension
  - Rapid expansion > 0.5 cm/y
  - Female gender
Rupture AAA

- Often present without known history of AAA
- High mortality: 50% don’t survive to be treated
- Acute severe abdominal pain and back pain
- Limb ischemia
  - Embolization of thrombus or atherosclerotic plaque
  - Weak/nonpalpable pulses
- Ecchymosis
  - Flank, Umbilicus, Proximal thigh
- Hypotension
- Shock
- Refer to ED for emergent evaluation
Management

- Blood pressure and lipid monitoring and control (Class I Level C)
- Smoking cessation (Class I Level B)
- Screening High Risk Populations
  - Men 60 years or older with 1st degree relatives with AAA should have physical exam and US screening (Class I Level B)
  - Men 65-75 who have ever smoked should have physical and 1 time US screening for AAA detection (Class I Level B)
- Juxtarenal and Infrarenal AAA 5.5 cm or greater should have elective repair to eliminate risk of rupture (Class I Level B)
- Juxtarenal and infrarenal AAA 4.0-5.4 cm should be monitored with US or CT every 6-12 months to monitor expansion (Class I Level A)
Crawford Classification
Thoracoabdominal aneurysms

Type I  Type II  Type III  Type IV
Referral

• Cardiothoracic surgery
  • Aortic root
  • Ascending aorta
  • Arch
  • Descending
  • Thoracoabdominal

• Vascular surgery
  • Descending
  • Thoracoabdominal
  • Abdominal
Aortic Dissection AoD

• An acute aortic syndrome
• Defined as:
  • Disruption of the media layer of the aorta with bleeding within and along aortic wall resulting in separation of the layers of the aortic wall.
    • 90% of time there is intimal disruption resulting in tracking of blood in dissection plane
• Incidence: Difficult to determine

1. Acute AoD is quickly fatal
2. Frequently missed on initial presentation and early mortality often not classified as dissection related.
IRAD: International Registry of Acute Aortic Dissections

• 6,000-10,000 cases reported annually
• Developed to assess presentation, management, and outcomes.

IRAD Review: > 7000 cases

• Mean age 63
• Significant Male predominance (65%)

Risk Factors

• HTN 76.6% *Most Common*
• Atherosclerosis 27%
• Known Aneurysm 16%
• Previous cardiac surgery 16%
• Marfan Syndrome 5%
• Iatrogenic 4%
• Cocaine use 1.8%
Types of AoD: 2 classification systems

- **DeBakey**
  - Based on the origin of the entry tear and extent of dissection.
    - 3 main types I, II, III
    - Type I: Tear starts in the ascending aorta and extends to arch and descending
    - Type II: Tear starts in and confined to ascending aorta
    - Type III: Tear starts in the descending aorta and moves distally

- **Stanford**
  - 2 categories, those that involve the ascending aorta and those that do not.
    - Type A: Ascending aorta involved
    - Type B: Involvement of the arch and descending aorta.
Classification of AoD
Presentation AoD

• Sudden onset chest pain radiating to back
  ▪ Described as a sharp, stabbing, ripping or tearing pain
  ▪ Pain may ease or abate, providing false reassurance

• Other symptoms related to location and progression
  ▪ Stroke symptoms: Great vessel involvement
  ▪ Abdominal pain, nausea, vomiting etc: Visceral ischemia
  ▪ Limb pain or numbness: if extension into iliac and femoral arteries
Management of AoD
ACC/AHA Guidelines 2010

- Debakey Type II and II, Sanford Type A = Surgical Emergency (Class I Level B)
- Debakey Type III, Sanford Type B
  - Medical management:
    - Unless malperfusion, progression of the dissection, enlarging aneurysm, inability to control BP or symptoms (Class I Level C)
  - Goal: Decrease aortic wall stress (Class I Level C)
  - Urgent surgical evaluation
  - HTN control: Anti-impulse therapy
    - Beta Blockade to achieve HR 60 or less
    - Avoid vasodilators until rate control initiated to avoid reflex tachycardia which can increase aortic wall stress and dissection propagation and expansion. (Class III Level C)
  - LONG TERM SURVAILLANCE: Cardiothoracic and Vascular surgery
Surgical management
Ascending and Root Aneurysms

- Cardiothoracic surgeons
- Ascending tube graft
- Bentall Procedure (valve conduit with reimplantation of Coronaries)
- Valve Sparing root replacements (David vs Yaqub techniques)
Surgical management
Ascending and Root Aneurysms

Ascending Aortic Aneurysm and Surgical Repair

Before surgery

After surgery
Surgical management
Ascending and Root Aneurysms
Mechanical Valve Bentall
30 M with congenital Unicuspid AV with 5.5cm Root aneurysm
Aortic Arch Surgery

- Cardiothoracic surgeon (sternotomy, CPB, Protection of brain, Heart and spinal Cord)
- Highly specialized techniques (deep Hypothermic circulatory arrest)
- Special training
- Open surgery is gold standard
- Endovascular approaches are being tested and will find their place in appropriately selected high risk patients
Aortic Arch Surgery
Complete arch vs Hybrid Arch
Thoraflex Hybrid Graft for Arch Reconstruction
Abdominal Aortic Aneurysms

- Taken Care of by Vascular Surgeons
- Open and Endovascular Stentgraft both mainstream approaches
- Currently 6 FDA approved Stentgrafts (All with individual nuances)
- Open surgery is mostly reserved for young and healthy
- Endovascular repair for elderly and High risk
- Long term results are equivalent at 10 Yrs
- High risk for recurrent interventions with Endovascular approach
- Open surgery is curative if patient can tolerate
Open surgical repair: advantages

- Aneurysm opened, graft sewn in, aorta wrapped and closed around graft
- Established procedure (with more than 40 years of clinical experience)
- Excludes aneurysm and prevents sac growth
- Proven, long-term results
Open surgical repair: drawbacks

- Significant incision in the abdomen
- 30–90 minute cross-clamp
- Up to 4-hour procedure
- Contraindicated in some patients
- 1–2 days intensive care
- 7–14 days hospitalization
- 4–6 weeks recovery time
Endovascular AAA Repair

• Inserted through a peripheral vessel
• Surgical exposure
• Positioned and deployed using image guided catheter/guidewire techniques
Endovascular Grafts

- Unibody graft
- Modular graft
- Aortoiliac graft
Endovascular AAA Repair

Advantages
- regional anesthesia
- shorter hospital stay
- shorter recovery
- potential for lower morbidity and mortality

Disadvantages
- not all AAA’s treatable
- need for follow up
- endoleaks
- long term efficacy unknown
- grafts expensive
Perigraft Flow/Endoleak: Definitions*

Type I
Attachment leak

Type II
Branch flow

Type III
Defect in graft or modular disconnection

Type IV
Fabric porosity

* White et.al., Endoleak Classification, Journal of Endovascular Surgery, 1998;5:305-309
Thoracoabdominal Aortic Repair

- Highly specialized surgery
- Morbid operation through big thoracoabdominal incision
- Need Cardiothoracic Surgeon and Perfusionist to perfuse the organs while the aorta is cross clamped.
- Spinal cord protection is key (distal aortic perfusion, Reimplantation of Intercostals, Lumbar drain, MEP, SSEP monitoring)
- Endovascular fenestrated and branched grafts are being tested and will become mainstream in next 5-10 yrs
Thoracoabdominal Aortic Repair
Thoracoabdominal aortic aneurysm Repair
Endovascular approach

FENESTRATED STENT-GRAFT

MULTI-BRANCHED STENT-GRAFT

ILIAC BRANCH STENT-GRAFT
Aortic Dissections

Type A
- Open Heart Surgery

Type B
- Complicated
- Uncomplicated
  - TEVAR
  - Medical Therapy
Aortic Dissection

Magnetic Resonance Imaging

DeBakey II
Stanford A

DeBakey III
Stanford B
Surgery for Aortic Dissection
Aortic Dissection Pathology
Type A Dissection
Type B aortic Dissection

Combined team approach (CV and Vascular)
TEVAR is gold standard (Class Ib) for Complicated Type B
Open Surgery is not recommended (Class IIa)
Medical Management is Currently Gold standard for Uncomplicated Type B (Class IA)
TEVAR will become standard of Care in high risk uncomplicated Type B aortic dissections
Malperfusion syndrome treated with endovascular stent-graft and PETTICOAT; a) angiography of lower body malperfusion; b) reperfusion after proximal stent-graft; c) 3D CT reconstruction of acute complicated dissection with malperfusion; d) reconstructed aorta and abolished malperfusion after stent-graft and PETTICOAT.
Medical: uncomplicated Type B dissection over time
Contrast-enhanced MRA of chronic type B dissection originating from the aortic arch region in MIP (A) and as volume-rendered 3D reconstruction (B). Follow-up MRA at 7 days after stent-graft placement shows a completely sealed proximal entry to the thrombosed false lumen. The diameter of the true lumen is normalized and the descending aorta is reconstructed (C).
Hartford Hospital Proposed Criteria
Type B dissection

• Complicated
  1. Rupture
  2. Malperfusion
  3. Persistent Shock
  4. Blood contents outside aortic wall
  5. Persistent pain Despite adequate BP control
  6. Aorta >5.5cm
  • Goal TEVAR if candidate to cover Entry tear and expand TL and perfusion of visceral organs, Surgery if not a candidate for TEVAR. Left SCA coverage and revascularization (multidisciplinary approach and individualize)

• Uncomplicated
  1. Aortic Dia (<4, 4-5, >5cm)
  2. Entry Tear (<1cm, 1-2cm, >2cm)
  3. FL size (<18mm, 18-22mm, >22mm)
  4. FL thrombus (Complete, No thrombus, partial Thrombus)
  5. Site of primary Tear (Abd aorta, Distal DTA or Abd, Prox DTA)
  • Composite Score 5-15
  • <7: Outpatient f-up and imaging
  • 7-12: Close surveillance with Imaging prior to DC
  • >12: Intervene with TEVAR/Surgery if candidate with close follow up imaging
Acute Type A dissection
Proposed Protocol

Diagnosis
- Acute Type A confirmed with CT or TTE or TEE (flap or mural hematoma)

Is the patient Operative candidate?
- Yes (low to intermediate risk)
- No (palliative care consult, institute medical management)
- High risk (surgical and medical) Medical management, consider endovascular options

Presentation
- Acute Type A with Clinical malperfusion (multidisciplinary discussion prep about perfusion strategy intraop and postop to address malperfusion)
- Acute Type A without clinical malperfusion

Operative Strategy: Replacement of Ascending aorta and hemiarch using HCA Root and or Valve replacement if involved
Consider complete or partial arch if aneurysmal or unable to exclude tear or young age with collagen vascular pathology
Regionalization of Aortic Care

Referral Network Aortic Service (for dissection analogy: rAAA)

Figure 1: Streamlined care and swift management begins with rapid emergency transfer to a certified emergency care centre followed by diagnostic imaging. Diagnostic CT images may be shared with the surgical/interventional team in another hospital or directly fed into the hybrid theatre for optimal care e.g. an EVAR first strategy if anatomy allows.

Regionalized care offering all treatment options from ascending aorta to distal malperfusion...
Hartford Hospital Approach

- Multidisciplinary approach (CV, Vascular, Cardiac Anesthesia with TEE capabilities, Dedicated CVICU, Neurosurgery back up, Hybrid OR, Imaging specialists, Shock team)
- All hands on deck
- Both open and endovascular approaches employed depending upon the patient’s risk profile and anatomy
- Cover the whole gamut from AV to femoral bifurcations and beyond
- Nurse Coordinator
- Genetic counseling and screening of Family members
- Acute care and longitudinal care
- Once an Aortic Patient always an aortic patient
- Address long term morbidity and mortality concerns
References


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