Overview of Aortic Root Pathologies- Complete Radiological Spectrum

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Disclaimer: We do not have any conflict of interest or financial gain to disclose

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Introduction

• Aortic root is the anatomic segment between left ventricle & ascending aorta

• It consists of aortic annulus, cusps, sinus of Valsalva (SoV), & the sino-tubular junction functioning as a unit

• Though echocardiography is usually the initial imaging modality, it has many limitations and at times may not be sufficient

• Cardiac MDCT/MRI offers excellent non-invasive assessment in accurately diagnosing and evaluating many aortic root pathologies

• Recognizing typical imaging manifestations with adequate clinical correlation is essential for timely and accurate diagnosis as well as for guiding treatment

In this exhibit, we discuss the characteristic multimodality imaging findings and differential diagnosis of common congenital and acquired aortic root pathologies. Increased awareness of such entities will contribute to optimized care of patients
Learning objectives:

1. Study radiological anatomy of aortic root
2. Study the role of imaging in diagnosis and evaluation of various aortic root pathologies
3. Discuss pathology based pertinent imaging findings helpful in preoperative planning
4. Discuss imaging based differential diagnosis

A standardized approach to the measurement of the aorta is needed. CT and MR images should be manipulated on a dedicated 3D workstation with multiplanar reformatting. True and accurate aortic diameters are measured orthogonally to the aortic lumen so as to avoid over or undermeasurements that are often seen when measured off-axis.

Common Aortic Root Pathologies

- Aortic stenosis
- Aortic regurgitation
- Aortic dissection
- Dilated aortic root/aneurysm
- Sinus of Valsalva aneurysm
- Sinus of Valsalva rupture
- Aortic valve infection
- Sub-aortic membrane
- Post-operative complications
- Aortic valve tumours
Anatomy of Aortic Root

- Aortic root is the anatomic segment between left ventricle & ascending aorta
- Consists of aortic annulus, cusps (arrows), SoV, & the sino-tubular junction functioning as a unit
- The aortic valve is a tricuspid structure that prevents diastolic retrograde flow into the ventricle
- Annulus (blue arrow) is the ring of attachment of the three aortic cusps
- SoV are hollow spaces enclosed by the aortic valve cusps and the surrounding dilatations of the aortic root wall

Courtesy Horacio Murillo, M.D. Sacramento, Ca.
Normal Variants: Bicuspid Aortic Valve (BAV)

- Most common congenital cardiac abnormality
- Prevalence 1%–2%
- Autosomal dominant with incomplete penetrance
- Sievers classification
- Fusion of the right and left coronary cusps is most common (Type I BAV)
- Commonly associated with aortic root dilatation - Bicuspid aortopathy

Patients with bicuspid aortic valve should routinely undergo imaging to monitor aortic size. Screening of family members is important, & in affected family members, aortopathy may be present even without BAV
Aortic Stenosis

- 3rd most prevalent cardiovascular disease, after hypertension & CAD
- Occurs a decade earlier in patients with a BAV than with normal tricuspid valve
- Can be degenerative or rheumatoid arthritis
- Leads to left ventricular hypertrophy (LVH), angina, dyspnea, syncope, & CCF
- Average survival without intervention: 2–3 years

In aortic stenosis, CT & MR demonstrate thickening of aortic valve cusps, diminished valve area, LVH, & post-stenotic dilatation. Stenotic jets can be seen in cine SSFP MR. Delayed myocardial enhancement (patchy & sub-endocardial in basal segments) at MR imaging can be seen with LVH exceed 18 mm. MDCT on the other hand is the modality of choice for evaluating anatomy before transcatheter aortic valve implantation.
Aortic Regurgitation (AR)

- Abnormal back flow of blood from aorta into LV during diastole seen as signal voids on SSFP MRI (arrows)
- Affects 10% of all patients with valvular heart disease
- Rheumatic heart disease is the most common cause in the developing world
- BAV & ectasia of the aortic root are more prevalent causes in developed nations
- Acute AR may be caused by endocarditis or dissection
- Mild or moderate AR is managed conservatively
- Patients with severe symptomatic AR typically undergo aortic valve replacement surgery

Diastolic CT and MR images can demonstrate the regurgitant orifice in moderate to severe AR. MDCT can be limited in presence of mild stenosis & valvular calcification. MR allows accurate quantification of regurgitant volume, fraction, & ventricular function
Aortic Root Dissection

- Most common acute emergent condition of the aorta
- Spontaneous longitudinal separation of aortic intima & adventitia by circulating blood
- Classic history: acute-onset central chest pain radiating to the back
- Proximal dissections can be associated with AR in 40–50% of cases
- Type A dissections account for 60%–70% and require urgent surgical intervention to avoid extension into aortic root, pericardium, or coronary arteries
- Early diagnosis & treatment are essential for improving prognosis

The overall outcome is determined by the type and extent of dissection and presence of associated complications; therefore, evaluation of the entire aorta, branch vessels, and iliac and proximal femoral arteries is recommended to aid in treatment planning. Untreated, type A dissections are associated with a mortality rate of over 50% within 48 hours.
CCTA demonstrating Stanford type A dissection of aortic root (red arrows) extending into the ascending aorta (blue arrows). The patient also has aneurysm of aortic root and ascending aorta.
CT angiogram thoracic aorta axial, coronal, and sagittal images show Stanford type A dissection. The dissection flap (yellow arrow) involves aortic root (green arrow).

CCTA demonstrating Stanford type A dissection of aortic root (red arrows) extending into the ascending aorta (blue arrow). The patient also has hemopericardium (green arrows).
Dilated Aortic Root/Aneurysm

- 60% of thoracic aortic aneurysms involve aortic root, ascending aorta, or both
- Risk of rupture/dissection is 6.9% per year with maximal diameter > 6 cm
- Management depends on aortic root diameter
- Normal aortic diameters depend on age, sex, body size, & anatomic segment
- Average aortic diameter at the Sinuses of Valsalva is 3.0 cm ± 0.5 cm
- Elective surgical intervention should also be considered in patients with rapid aortic growth (>0.5–1 cm per year),

Aortic aneurysms are associated with many disorders including hypertension, atherosclerosis, BAV disease, familial thoracic aortic aneurysmal disease, vasculitis, & heritable connective tissue disorders like Marfan syndrome. Management, particularly the timing of surgical intervention, is based on size, necessitating precise, accurate, reproducible measurement with noninvasive imaging with MDCT and MRI.
34–years-old male with Ehler Danlos Type IV. Status post aortic and mitral valve surgery. CCTA demonstrating dilated aortic root (red arrows) and ascending aorta (blue arrows). The patient also had tricuspid insufficiency.

Up to 80% of patients with type IV Ehlers Danlos syndrome experience vascular complication by 40 years of age. Imaging with CT or MRI is the primary surveillance method because of high risk of arterial injury during conventional angiography. The classic appearance for Marfan syndrome is annuloaortic ectasia with dilatation of annulus & sinuses with effaced sinotubular junction. In Loeys-Dietz syndrome, aortic dissection may occur at even smaller diameters & surgical repair is often recommended with aortic diameter > 4–4.5 cm, or aortic expansion of >5 mm per year.
Mycotic Aneurysm

- True primary bacterial infection of the ascending aortic wall resulting in aneurysm formation is rare.
- Believed to occur either after an episode of bacterial endocarditis or from an aortic jet lesion causing endothelial trauma.
- Most common organisms include Staphylococcus aureus, Staphylococcus epidermidis, Salmonella & Streptococcus.

Bacterial endocarditis and acquired L-SoV aneurysm axial (a) and saggital (b) views at the level of SoV (arrows)

Aortic Pseudoaneurysm

- A false aneurysm/pseudoaneurysm involves enlargement of the aorta due to dilatation of only the outer layers of the vessel wall.
- They generally involve focal penetration of the intima and media, usually manifesting as a complication of trauma, surgery, infection, or atherosclerosis.
- The adventitia and perivascular connective tissue remain intact.

26-years-old male status post aortic valve and ascending aortic dacron graft 10 years back, now septic and presents with an perigraft fluid/abscess (red arrows) and root pseudoaneurysm (blue arrows).

Moktassi A, et al. JACR 2006;57:238
86-years-old male status post aortic valve replacement, now septic. MDCT shows perivalvular leak (red arrows) and aortic root pseudoaneurysm (blue arrows)

49-years-old male status post aortic valve replacement. MDCT shows aortic root pseudoaneurysm (blue arrows)
45-years-old IV drug abuser female and Cocaine (+). Aortic valve and mitral valve replacement (blue arrows) 10 months before for bacterial endocarditis. New onset chest pain. MDCT shows pseudoaneurysm of the aortic root involving left coronary sinus (red arrows) pushing upward left main CA.
Post-operative collection and Pseudoaneurysm

29-years-old IV drug abuser male. Aortic valve and ascending aortic dacron graft, 10 years before. New onset chest pain and fever. MDCT shows pseudoaneurysm of the aortic root (red arrows) and collection likely abscess around the ascending aorta graft (blue arrows)

Potential imaging pitfalls in the postoperative period include mimickers of pathologic processes such as felt pledgets, graft folds, and nonabsorbable hemostatic agents

Sinus of Valsalva (SoV) Aneurysm

- SoV are hollow spaces enclosed by the aortic valve (AV) cusps and the surrounding dilatations of the aortic root wall (yellow shade).
- Congenital SoV aneurysms most commonly arise from right coronary SoV (R-SoV) and the non-coronary SoV (N-SoV) because of incomplete fusion (or weakness) of 2 halves of distal bulbar septum.
- Congenital left SoV are rare.

Congenital SoVA may also be associated with VSD (50%), Aortic insufficiency (20-30%), Bicuspid aortic valve (10%), pulmonary stenosis, coarctation, & ASD.

JRadiol Case Rep. 2011; 5(8): 14–21
AJR 2010; 194:W495–W504
Pathophysiology and progression of Congenital SoV Aneurysms (arrows)

Courtesy Horacio Murillo, M.D.
Sacramento, Ca.
Clinical Presentation of SoV Aneurysms

- Unruptured SoV aneurysms are asymptomatic
- Unless large enough to cause:
  - Obstruction of the rvot
  - Tricuspid stenosis and insufficiency due to prolapse of aneurysm through tricuspid valve
  - Conduction abnormalities due to mass-effect on bundle of his or its fascicles

Ruptured SoV aneurysms present with sudden onset of severe chest pain and dyspnea
Ruptured SoVA

- Rupture can occur spontaneously, following trauma, strenuous exertion or bacterial endocarditis.
- When ruptured, produces an intracardiac fistula (L→R shunt).
- When a SoVA from the left coronary sinus ruptures, may bleed into the pericardium.
- Association with ventricular septal defect (VSD) (59%) and aortic regurgitation (25%) is common.

J Thorac Cardiov Surg 1990;99:288

56 yo female with fatal rupture of R-SoV into the RV.

13 yo female with fatal rupture of R-SoV aneurysm into RA.
Acute Presentation

- Usually due to sudden, large rupture/perforation

- Symptoms include sudden, severe chest pain and dyspnea

- Chest X-ray: enlarged cardiac silhouette, pulmonary edema, and CHF

- Since the aortic root is intracardiac, the aneurysm may not be visible in plain radiographs
Diagnostic Imaging

- Echocardiography with color Doppler is the current modality to confirm diagnosis.

Diagnostic Criteria:

- Root of aneurysm must be above the aortic annulus.
- Aneurysm is saccular.
- The size of the aorta above is normal.
Cardiac CTA, aortography, and magnetic resonance imaging may also be used.

CCTA volume rendered 3D reconstruction (a) of an unruptured SoV aneurysm (white arrow). Catheter angiography (b) demonstrating an unruptured N-SoV aneurysm. Magnetic resonance imaging (c) demonstrating an unruptured N-SoV aneurysm.
Rupture SoVA into the RV

Previously healthy, 40 y.o. athletic male with one week of severe chest pain, shortness of breath, and orthopnea

*Courtesy Horacio Murillo, M.D. Sacramento, Ca.*
Pre and Post-Surgical Repair - R-SoV ruptured aneurysm into RV

Courtesy Horacio Murillo, M.D. Sacramento, Ca.
Courtesy Horacio Murillo, M.D.
Sacramento, Ca.
Aortic Valve Infection

- Aortic Valve infection can be seen in non-drug abusers and in IV drug abusers
- Degenerative valvular disease now more common than rheumatic heart disease as a major risk factor
- 50% of all cases in the elderly (USA)
- Complications include CNS (stroke, abscess), visceral and MSK infarction/infection

CCTA demonstrate Aortic valve vegetations (arrow), in a patient with infective endocarditis
CCTA demonstrate aortic valve vegetation (red arrows) in a patient with infective endocarditis.

CCTA demonstrate aortic valve vegetation (red arrow) and left coronary sinus aneurysm (blue arrow) in a patient with infective endocarditis.
Aortic Root Injury

- Life threatening condition that requires prompt diagnosis and management
- Proposed mechanisms: rapid deceleration, shearing forces, osseous pinch, and hydrostatic forces or water-hammer phenomenon
- Most commonly occur at sites of aortic tethering; the aortic root, the isthmus and at the diaphragmatic hiatus
- Associated with a numerous additional injuries
- Direct signs: intramural hematoma, intimal flap & pseudoaneurysm
- Indirect signs: periaortic hematoma, change in aortic caliber and irregular aortic contour

Traumatic aortic injuries have a high mortality rate and a high index of suspicion and careful evaluation is needed for accurate diagnosis. TAIs can be mimicked by ductal diverticula, arterial infundibuli, and venous mediastinal hematoma. Technical factors such as the use of ECG gating and obtaining reconstructions with different slice thickness or in a different plane may be helpful.
MDCT showing aortic root injury extending into the ascending aorta (arrows) in a 32-year-old female patient. Status post motor vehicle accident
Subvalvular Aortic Stenosis

- A rare CHD in which a fibrous ring, muscular ridge or fibromuscular tunnel either alone or in combination causes LVOTO
- Discrete membranous type is the most common (90%)
- Mean age - 20 years
- Associated with other CHD like bicuspid aortic valve, VSD, Shone’s complex
- Can develop left ventricular hypertrophy, aortic insufficiency & prone for sub-acute bacterial endocarditis

Shem-Tov A et al; Circulation. 1982 Sep;66(3):509-12

Discrete subaortic stenosis (SAS) in a 47 year old female. CCTA shows subaortic stenosis from the presence of a thin membrane in the subaortic region (black arrows).
Subvalvular Aortic Stenosis

**Discrete subaortic stenosis** (SAS) in a 47 year old female. CCTA shows subaortic stenosis from the presence of a thin membrane in the subaortic region (black arrows).
Aortic valve tumor- Papillary Fibroelastoma

- Commonest neoplasm of the valves
- Composed of collagen and elastic tissue
- Symptoms if present are usually related to embolic events
- Well circumscribed small mobile (<1.5 cm) nodule
- Iso-hypodense on MDCT
- Low signal on SSFP MR sequences, often with peri-lesional flow artifact & intermediate on T1 & T2
- Differential diagnosis: vegetation or thrombus

Conclusion

• MDCT and MR imaging help in the diagnosis, surveillance, and pre- & postoperative evaluation of aortic root pathologies

• MDCT is the modality of choice for evaluating anatomy before transcatheter aortic valve implantation

• MR allows accurate quantification of regurgitant volume, regurgitant fraction, & ventricular function

• Radiologist must be aware of normal and abnormal postoperative imaging findings as normal postoperative appearances may mimic those of pathologic conditions

• Increased awareness will contribute to optimized care of patients and improve the confidence of referring cardiologists and surgeons

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