Συγγενείς ανωμαλίες των στεφανιαίων αρτηριών. Κλινική σημασία

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Sudden Cardiac Death in Athletes

- HCM (36%)
- Unexplained cardiac mass (10%)
- Aberrant coronary arteries (13%)
- Other coronary abnormalities (6%)
- Ruptured aortic aneurysm (5%)
- Tunnelled LAD (5%)
- Aortic valve stenosis (4%)
- Myocarditis (3%)
- DCM (3%)
- ARVC (3%)
- Myocardial scarring (3%)
- MVP (2%)
- Atherosclerotic CAD (2%)
- Other congenital diseases (1.5%)
- LQTS (0.5%)
- Sarcoidosis (0.5%)
- Sickle cell trait (0.5%)
- "Normal" heart (2%)
Most important to know normal coronary anatomy and normal variants.
Normal Coronary Anatomy

- Left main – arises from the superior portion of the left aortic sinus, just below the sinotubular ridge of the aorta
  - Subsequently bifurcates or trifurcates
Left Coronary Anatomy

Left Main Coronary Artery

[Diagram showing anatomy of the left coronary artery, including the left anterior descending, left main, and left circumflex arteries.]

LAO Caudal “Spider” Projection
Normal Coronary Anatomy

- LAD – courses along the anterior interventricular groove toward the cardiac apex
  - Normal variant – “twin” LAD with a branch (often intramyocardial) supplying the septals and a branch supplying diagonals
Left Coronary Anatomy

Left Anterior Descending Artery

LAO Cranial Projection
Normal coronary anatomy

- Left Circumflex – originates from the left main and courses within the posterior AV groove
  - Typically supplies 1-3 obtuse marginal branches depending on the size of the posterolateral branches
  - Sinus node branch – arise 60% from RCA and as a left atrial branch of the CX in 40%
  - Left dominance CX supplies the PLV, PDA and AV nodal artery
Left Coronary Anatomy

Left Circumflex Artery

[Image of coronary anatomy with labels: LAD, Left Main, Left Circumflex, Obtuse Marginal Branch, RAO Cranial Projection]
Normal coronary anatomy

- RCA – originates from the right anterior aortic sinus somewhat inferior to the origin of the left coronary artery. It passes along the right AV groove
  - First branch is typically the conus (supplies RVOT, AV nodal artery typically present in right dominant arteries)
  - Second branch is typically the sinoatrial branch
  - Terminates in the PDA and PLV
Normal coronary anatomy

- **Coronary Dominance**
  - Defined by whether the LCA or the RCA supplies the PDA.
  - RCA is dominant 85% of the time
  - LCA is dominant 8%
  - Balanced dominance 7% - RCA that supplies the PDA with a CX that supplies the PLV is a balanced or “codominant” circulation.
Anomalous Coronary Arteries

- Found in ~0.1%-1.3% of patients undergoing cardiac catheterization
- Can be assoc w/ congenital heart dz or be isolated anomaly
- Angio evaluation can be challenging; misdiagnosis in up to 50% of cases
- Rare but important cause of CP, arrhythmia, MI & sudden cardiac death; TREATABLE
Why Is It So Dangerous?

- Not fully understood; many variants benign
- But some variants w/ mortality rates >50%
- Depends on course of anomalous artery: retroaortic & anterior courses benign
- Dangerous: “interarterial” course b/w aorta & RVOT
- Pathophysiology unclear: compression or kinking during systole vs. abnl narrowing of ostium
Role for Noninvasive Imaging

- Often challenging to diagnose in selective coronary angiogram (e.g. difficult to see relationship to MPA)
- Limited eval of small vessels w/ echo
- CT allows eval of not just arterial caliber and lumen but also their course and relationship to adjacent structures
- Cardiac MRI/A may also be useful but cannot perform on pts c pacers/AICDs
Congenital Coronary Anomalies

- Occur infrequently <1%
- Need to determine benign from potentially serious – it is important to define the origin and course of anomalous coronary arteries
- Largest study reviewed 126,595 cases from Cleveland Clinic
# Coronary Anomalies

**Data Based On Analysis of 126,595 Angiograms**

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>Incidence (%)</th>
<th>Anomalies (%)</th>
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</thead>
<tbody>
<tr>
<td><strong>Benign anomalies (80%)</strong></td>
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<tr>
<td>Separate, adjacent LAD and LCX ostia</td>
<td>0.41</td>
<td>30.4</td>
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<td>LCX origin from RSV or RCA</td>
<td>0.37</td>
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<tr>
<td>Anomalous origin PSV</td>
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<td><strong>Anomalous origin from aorta:</strong></td>
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<tr>
<td>LMCA</td>
<td>0.01</td>
<td>1.0</td>
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<tr>
<td>RCA</td>
<td>0.12</td>
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<tr>
<td>Absent LCX</td>
<td>0.003</td>
<td>0.2</td>
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<tr>
<td>Small fistulae</td>
<td>0.12</td>
<td>9.7</td>
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<td><strong>Potentially serious (20%)</strong></td>
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<td>Origin of coronary artery from opposite aortic sinus:</td>
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<td>LMCA from RSV</td>
<td>0.02</td>
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<tr>
<td>LAD from RSV</td>
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<tr>
<td>RCA from LSV</td>
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<td><strong>Anomalous origin from pulmonary artery:</strong></td>
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<td>LMCA</td>
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<td>0.6</td>
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<tr>
<td>LAD or RCA</td>
<td>0.003</td>
<td>0.2</td>
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<tr>
<td>Single coronary artery</td>
<td>0.05</td>
<td>3.3</td>
</tr>
<tr>
<td>Multiple or large coronary fistulae</td>
<td>0.05</td>
<td>3.7</td>
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</table>
LAD and CFX Arising from Separate Ostia

BENIGN CORONARY ANOMALIES

• Incidence...1.07%, Of all anomalies...80.6%.
• a) Separate origin of left anterior descending artery and left circumflex from the left sinus Valsalva (0.41% incidence, 30.4% of all anomalies).
High Takeoff Coronary Ostia Above Sinuses of Valsalva
Anomalous Circumflex Artery

- Anomalous circumflex:
  - Either off R sinus or branches off RCA
  - ALMOST ALWAYS RETROCARDIAC → BENIGN
Retro-Aortic Left Circumflex
Retro-Aortic Left Circumflex
Benign coronary anomalies

2nd most common anomaly is **CX** from the RCA or right sinus of valsalva – 27.7%

**Suspect when LM is long with a paucity of vessels**

Injection of the RCA reveals filling of the left circumflex artery. The path is described by the arrows. It always courses posteriorly to the aorta to reach the right AV groove. In the left panel, the left anterior descending (LAD) is shown as a single vessel.
Benign Coronary Anomalies

- Injection of the right coronary artery in the left anterior oblique projection with cranial angulation shows the anomalous left circumflex coronary artery.
- Can try an ALI
Anomalous Circumflex:
Retroaortic $\rightarrow$ BENIGN

Normal Anatomy
Retroaortic Anomalous Circumflex
Potentially ischemic anomalies

Coronary fistulas mostly connect to the RV, RA PA or coronary sinus.

When large they may cause chronic volume overload or ischemic and must be closed.

Smaller fistulas can be managed medically

Prevelance is 0.1%

Left coronary angiography performed in the RAO projection shows a number of small vessels (arrows) arising from the proximal LAD and forming a plexus emptying into the proximal pulmonary artery.
Myocardial Bridges

- Prevalence by cardiac CT
  - Recent article in JACC – evaluate 118 patients
    - Prevalence was 30.5%

- Location
  - Most were located in mid left anterior descending coronary artery (LAD), 27 of 47 (57%)
  - Distal LAD, 7 of 47 (15%)
  - Diagonal, 6 of 47
  - Intermediate, 4 of 47
  - Obtuse Marginal 3 of 47
  - RCA – no bridging present in series

- The CCTA features in the LAD showed 3 patterns:
  - Superficial septal, 10 of 34 (29.4%)
  - Deep septal, 14 of 34 (41.1%)
  - Right ventricular type, 10 of 34 (29.4%).

Konen, Eli, Goitein, Orly, Sternik, Leonid, Eshet, Yael, Shemesh, Joseph, Di Segni, Elio
The Prevalence and Anatomical Patterns of Intramuscular Coronary Arteries: A Coronary Computed Tomography Angiographic Study
J Am Coll Cardiol 2007 49: 587-593
The mid LAD (arrow) shows a typical deviation and straitening and is only partially surrounded by myocardium. Of note, an atherosclerotic plaque in the proximal LAD, whereas the intramuscular segment is free of disease.
Myocardial bridge – deep type

- Intramuscular LAD, deep type, as seen on axial plane (A, B) and multiplanar reformation (C, D). The mid LAD crosses deeply into the myocardium (arrows).

- ~1/3
Intramural LAD, right ventricular type (arrow). In this variant it is frequently difficult to follow the LAD on sequential axial images (A, B) because it disappears between the right ventricular trabeculae, whereas the multiplanar reformation images easily show its intraventricular course (C, D).

~1/3
Compression of the mid segment of the LAD artery during cardiac systole.

The compression of the LAD persists during diastole, accounting for the patient’s exercise-induced ischemia.
Myocardial Bridging
Tunneled LAD

- Autopsy: ~30%, Angiographically: <5%
- Prevalent in HCM patients
- Segment proximal to bridge frequently shows atherosclerotic plaque (tunnel spared)
- Symptomatic patients may be treated with β-blocker or CCB
- Myotomy, CABG, and stenting in refractory cases
Left Main Arising from Right Coronary Sinus

Subtypes:

1. **Anterior free-wall** course
2. **Retro-aortic** course
3. **Septal** course
4. **Inter-arterial** - incidence 1:12,500
   [Accounts for 60% of anomalous left main from right coronary sinus (2.8% overall coronary anomalies). Recognized association with ischemic symptoms and sudden death >50%]
The left main trunk arises ectopically from the right sinus of Valsalva and passes epicardially across the RV outflow tract.
Left Main from Right Sinus
Anterior Course
Anomalous Left Coronaries
Retro-Aortic Circumflex, Anterior LAD

The LAD and circumflex branches arise from the right sinus of Valsalva. The LAD passes epicardially across the RVOT, and the CFX passes behind the aorta.
The left main trunk originates from the right sinus of Valsalva and passes behind the aorta before dividing into the LAD and CFX.
Retro-Aortic Course of Left Main

*Dot-sign posterior to aortic root

The left main trunk arises ectopically from the right sinus of Valsalva and passes between the aorta and pulmonary artery.
Inter-arterial Course of Left Main Arising From Right Coronary Sinus

*Dot sign anterior to aortic root

Anomalous Coronary Artery

- 15-year-old male soccer player
- Syncope one year previously (ECG normal).
- Died suddenly while running
- Acute angle take-off of LMCA from right coronary sinus
Anomalous LCA from the RSV

Left Main Coronary Artery Arising From the Right Sinus of Valsalva

Cross-sectional Representation  Representative RAO Angiographic Features

Aortic Valve  Septal Course
  Pulmonic Valve  Anterior Descending
  Septals

Aortic Valve  Anterior Course
  Pulmonic Valve  Anterior Descending
  Circumflex

Aortic Valve  Interarterial Course
  Pulmonic Valve  Anterior Descending
  Circumflex

Aortic Valve  Retroaortic Course
  Pulmonic Valve  Anterior Descending
  Circumflex
Diagram of a single coronary artery originating from the right sinus of Valsalva with the left coronary trunk coursing between the pulmonary artery and aorta.
Anomalous LCA from the RSV

- Interarterial or preaortic course of the left main coronary artery
- The course of the LM between the aorta and the pulmonary trunk is clearly delineated.
- This anomaly has been linked with sudden cardiac death
Anomalous LCA from the RSV

- Posterior or retroaortic course of the left main coronary artery
- The LM originates from the right sinus of Valsalva and passes posterior and inferior to the aortic root.
LAD FROM RCA
RCA from the LSV

- Anomalous origin of the right coronary artery. In the right panel, the left anterior oblique projection shows the right coronary artery originating from the left sinus of Valsalva. The course of the right coronary artery passes between the aorta and pulmonary artery.
Anomalous Right Coronary

- Anomalous RCA:
  - Either off L sinus or branches off single left coronary
  - Can be retroaortic but **IN VAST MAJORITY (>90%) OF CASES INTERARTERIAL** → MALIGNANT
Anomalous RCA:
Retroaortic → BENIGN

Normal Anatomy
Coronary Artery Anomalies
Magnetic Resonance Imaging
Blood flows from the RCA via collaterals to the left coronary artery, and then into the pulmonary artery.
Anomalous origins from the PA

- LCA origin from the PA – typically presents with CHF and myocardial ischemia in first months of life
- 25% of patients survive to adulthood

Left Circumflex Origin from PA
Other Anomalies

- Anomalous connection of right coronary artery to left circumflex artery. This anomaly is known as the Miller connection, as shown in this shallow left anterior oblique.
Absent Circumflex
Super-dominant RCA
Absent Right Coronary Super-dominant Circumflex
Normal Anatomy
Normal Anatomy

Anomalous LCA:
Interarterial $\rightarrow$ ISCHEMIA!!
Normal Anatomy
Normal Anatomy

Anom LCA: Retroaortic

Anom LCA: Anterior

Anom LCA: Intramural
Normal Anatomy

Anom LCA: Retroaortic

Anom LCA: Anterior

Anom LCA: Intramural

BENIGN!!
Normal Anatomy

Anomalous RCA:
Interarterial → ISCHEMIA!!
Coronary Artery Anomalies Demonstrated by Echocardiography
Συμπεράσματα

- Σπάνια αλλά υπαρκτή οντότητα
- Ιδιαίτερη προσοχή στο αιμοδυναμικό
- Στην πλειοψηφία τους καλοήθεις
- Η πορεία καθορίζει τη σημασία
- Οι νεότερες απεικονιστικές τεχνικές αποτελούν σημαντικό βήμα προόδου
R A V groove

L sinus of Valsalva

INTERARTERIAL → ISCHEMIA!!!

Anomalous RCA
Takeoff From Left Coronary Sinus

*The most common, potentially serious coronary anomaly, accounting for 8.1% of serious coronary anomalies (25% incidence of sudden cardiac death).