#### Congenital heart disease imaging



# Anomalous origin of the coronary arteries





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# Statement of Financial Interest

I currently have, or have had over the last two years, an affiliation or financial interests or interests of any order with a company or I receive compensation or fees or research grants with a commercial company:

✓ I do not have any potential disclosure to report



#### AAOCA: anomalous aortic origin of coronary arteries



## Imaging tools

- Diagnosis of AAOCA
- Relevance of AAOCA
- Myocardial ischemia
- Therapeutic management
- Follow-up after correction





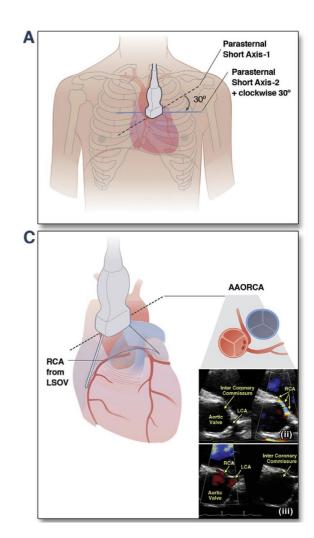
#### Echocardiography

#### Transthoracic echocardiography

- In children : most valuable diagnostic tool
- In adults: limited acoustic window

# Transthoracic echocardiography in children for pre participation screening program

- Standardized protocols
- Visualization of coronary ostia



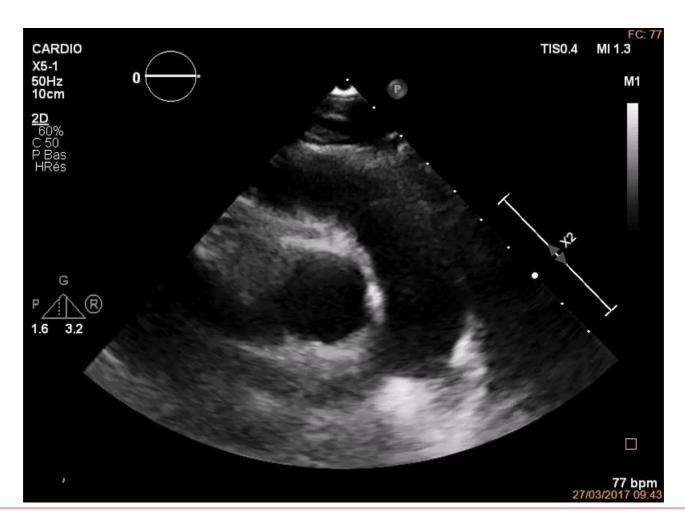
Courtesy of Xavier Iriart (Bordeaux)





#### **Echocardiography**

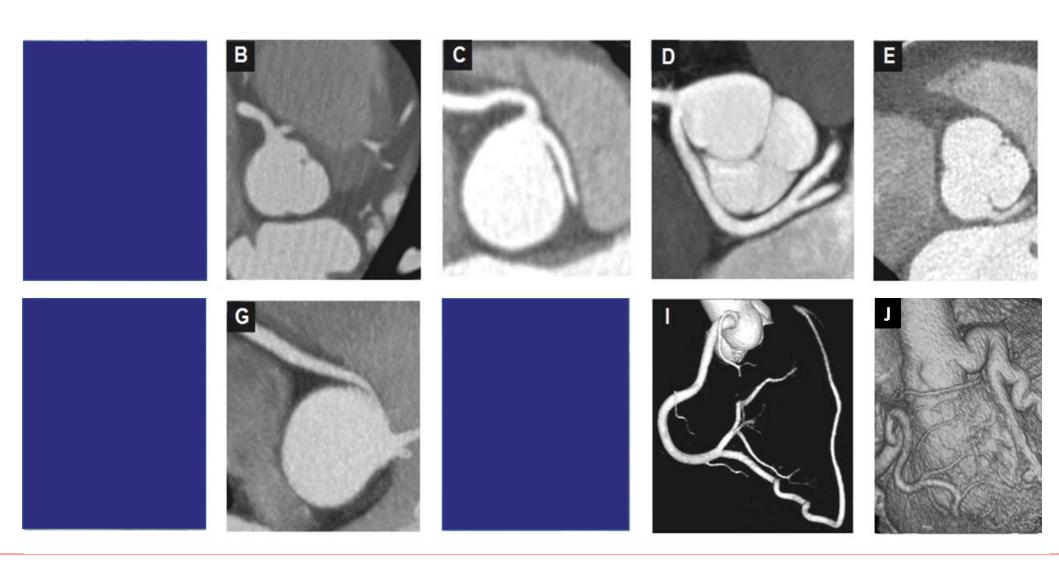
- 17-year-old boy
- Pre syncope after exertion







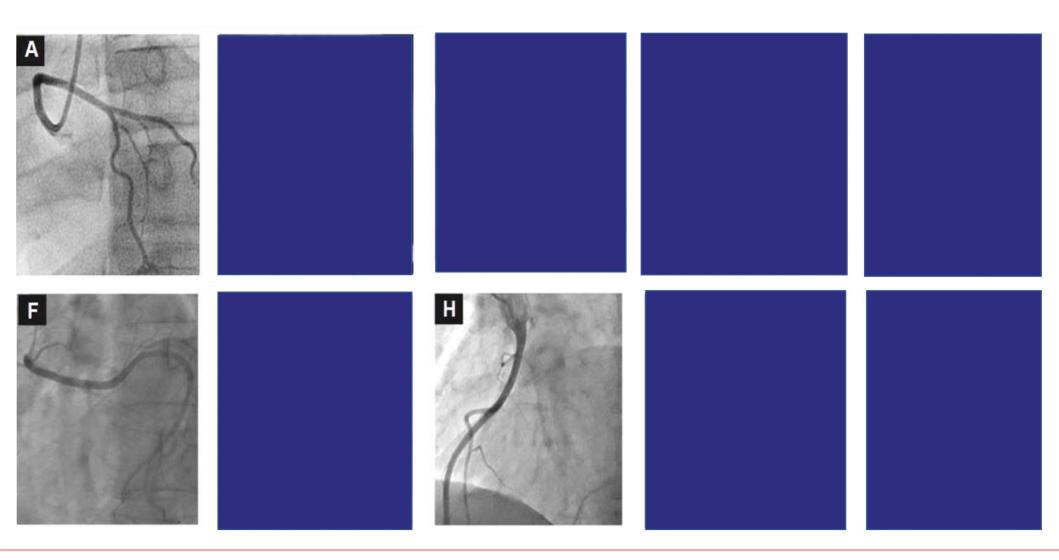
#### Coronary CT angiography







#### Coronary angiography

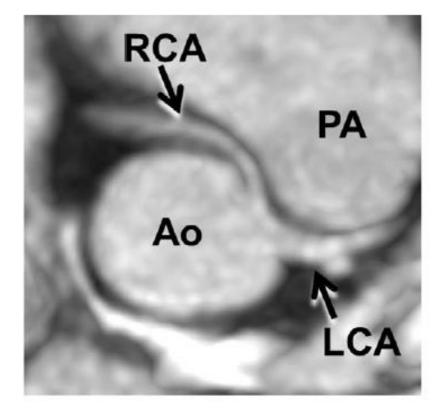






#### Cardiac MRI

- US guidelines
- CMRI recommended over coronary CTA in youngs
- Experienced centers
- Limited use in adults



Harris AH et al. Ann Thor Surg 2015





Risk of sudden cardiac death/myocardial ischemia

AAOCA without known risk AAOCA with known risk **Prepulmonic Interarterial** PA or or RS RS Retroaortic Retropulmonic PA PA or or PΙ PI RS RS

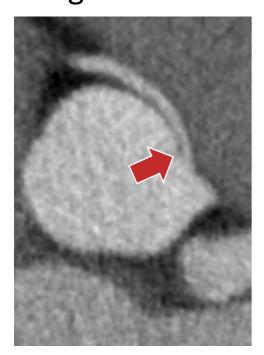




#### Coronary CT angiography

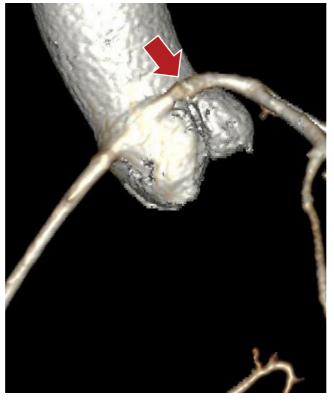
#### AAOCA with known risk

right AAOCA



interarterial course

left AAOCA



interarterial course

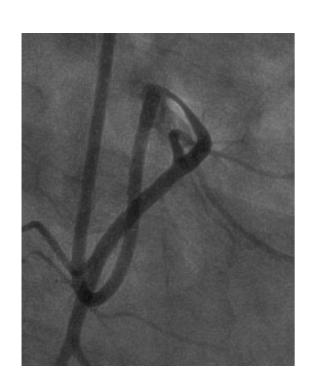
left AAOCA



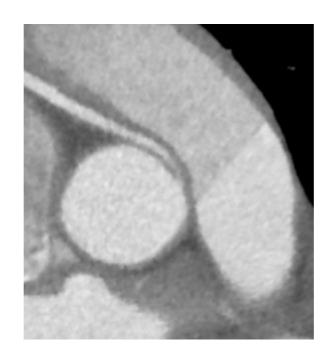
retropulmonic course



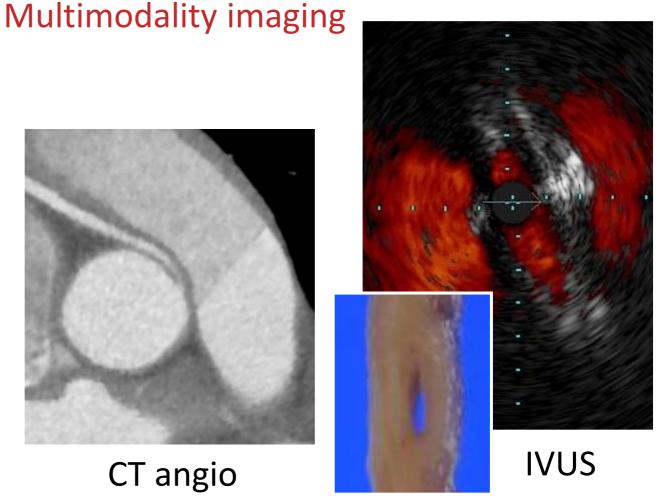
#### Identification of intramural aortic course



**Angio** 



CT angio

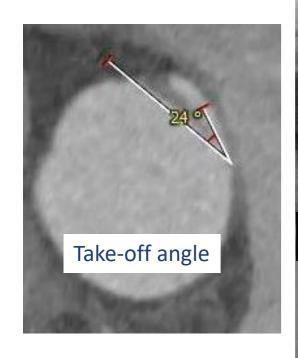


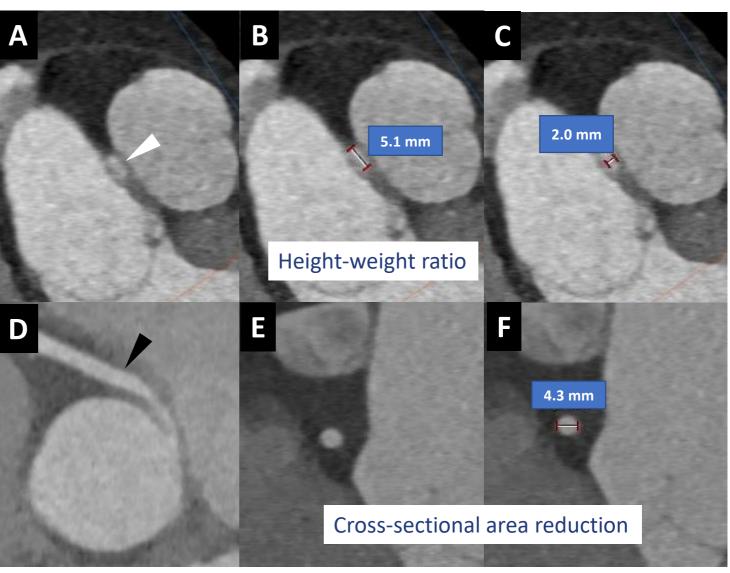
Hata Y et al. Cardiovasc Pathol 2014.





#### Identification of anatomic high-risk features









# Myocardial ischemia

Non invasive ischemia testing/Invasive ischemia testing

- TT echocardiography
- Single-photon emission CT
- Cardiac MRI
- Physiological evaluation (iFR, FFR)
- Endovascular imaging (IVUS/OCT)

- Low incidence of ischemia detection
- Limits of exercise stress protocols
- Dynamic obstruction associated



Dobutamine infusion
+ chronotropic /+ inotropic stress
Better protocol?



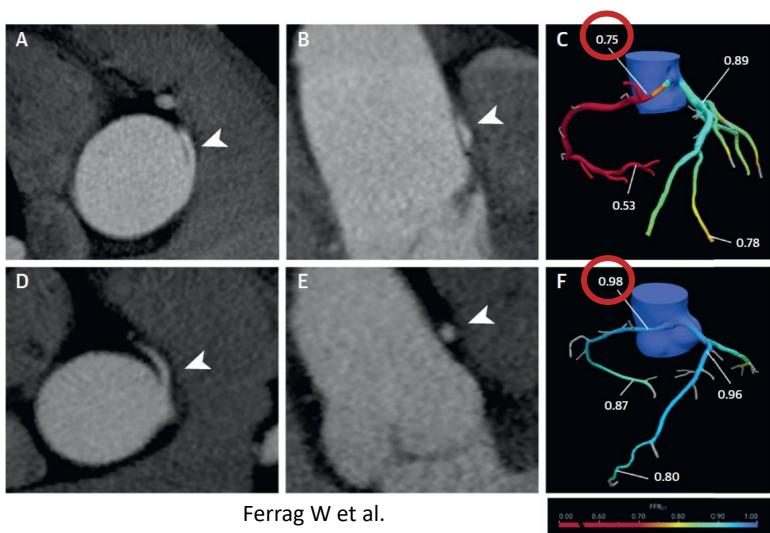


# Myocardial ischemia

#### Coronary CT angiography and FFR-CT images

Right-AAOCA with intramural pathway

Right-AAOCA without intramural pathway



JACC: CV Imaging 2020





# Therapeutic management

#### **ESC** guidelines



**ESC GUIDELINES** 

2020 ESC Guidelines for the management of adult congenital heart disease

#### Anomalous aortic origin of the coronary artery

Surgery is recommended for AAOCA in patients with typical angina symptoms who present with evidence of stress-induced myocardial ischaemia in a matching territory or high-risk anatomy.<sup>c</sup>



Surgery should be considered in <i>asympto-matic</i> patients with AAOCA (right or left) and evidence of myocardial ischaemia.	lla	С
Surgery should be considered in asymptomatic patients with AAOLCA and no evidence of myocardial ischaemia but a high-risk anatomy. <sup>c</sup>	lla	С
Surgery may be considered for symptomatic patients with AAOCA even if there is no evidence of myocardial ischaemia or highrisk anatomy. <sup>c</sup>	IIb	С
Surgery may be considered for <i>asymptomatic</i> patients with AAOLCA without myocardial ischaemia and without <u>high-risk anatomy</u> <sup>c</sup> when they present at young age (<35 years).	IIb	С
Surgery is not recommended for AAORCA in asymptomatic patients without myocardial ischaemia and without high-risk anatomy. <sup>c</sup>	Ш	С

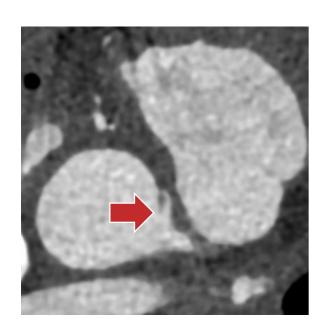


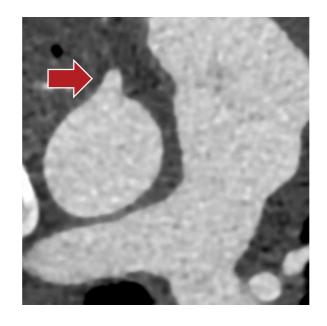


# PJESFC Follow up after correction

#### Coronary CT angiography

Surgical correction of right AAOCA with neo-ostium creation





Angioplasty of right AAOCA with stenting







### Conclusions

- CT in first line for the diagnosis of AAOCA
- Identification of AAOCA at risk by CT
- Identification of anatomic high-risk features by CT/endovascular imaging/angio
- Non invasive/invasive functional imaging (myocardial ischemia)
- Scores for risk stratification not yet available
- Therapeutic decision-making often difficult

