

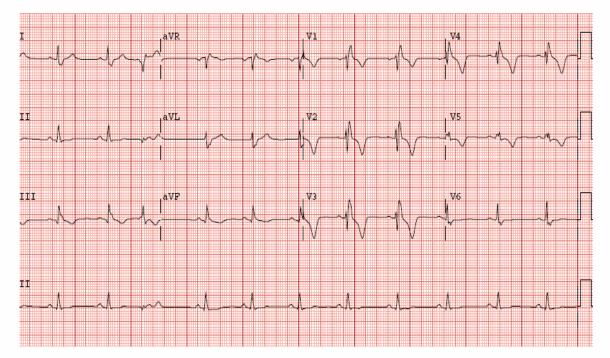
#### **Case Presentation 1**

55 yow presented to outside hospital with SOB and near-syncope. Unwitnessed event with exertion, and without prodrome. Denied prior dizziness, palpitations or syncope.

- Chest CT for PE revealed RV mass.
- PMHx: "childhood murmur", vertigo
- Meds, SHx, FHx: unremarkable.
- PE: 106/60, 72 no orthostatic changes, no murmur at rest or with maneuvers.



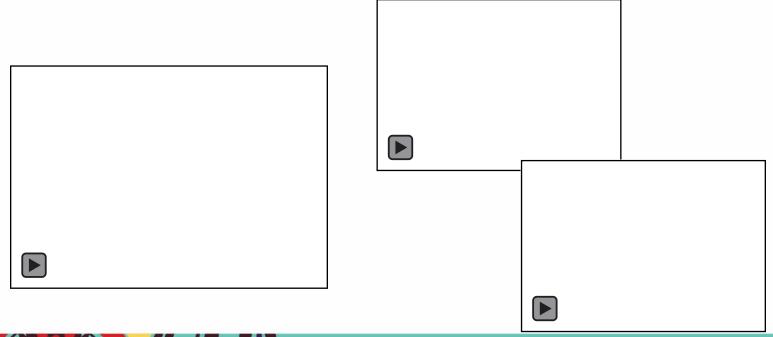
#### ECG: RAD, RVH, RBBB







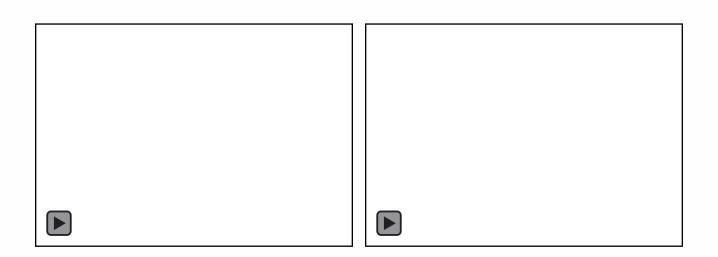
#### **ECHOCARDIOGRAM**







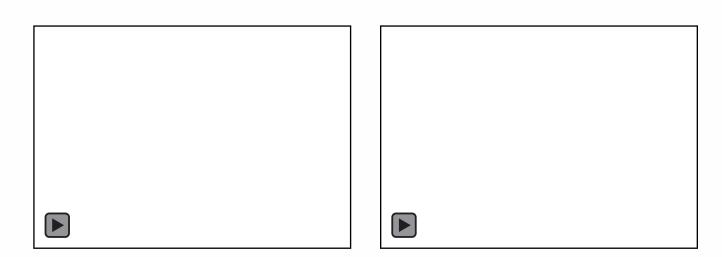
#### **ECHOCARDIOGRAM**







#### **ECHOCARDIOGRAM**



Invading RV apical myocardium and pericardium?





#### What would you do next?

- 1. More imaging:
  - Cardiac CT
  - TEE
  - MRI
  - PET
  - Coronary angiogram
- 2. Endomyocardial Biopsy?
- 3. Call a surgeon





#### **Differential Diagnosis Matters**

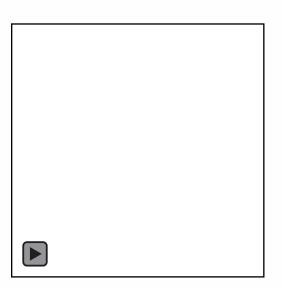
#### Cardiac Masses

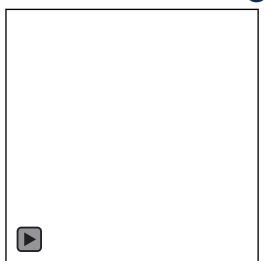
- Primary Tumors:
  - Benign:
    - Myxoma, fibroelastoma (intracardiac)
    - Lipoma, fibroma, rhabdomyomas (children TS)
  - Malignant:
    - Sarcoma (angiosarcoma), lymphoma
- Metastatic cardiac involvement
- Thrombus
- Infection
- HCM variant?
  - Is this the cause of her childhood heart murmur?





#### Cardiac MRI





Pre-gadolinium injection

Post-gadolinium injection

Avid enhancement (vascular), but uniform and no necrosis, isointense to muscle





#### **Endomyocardial biopsy**

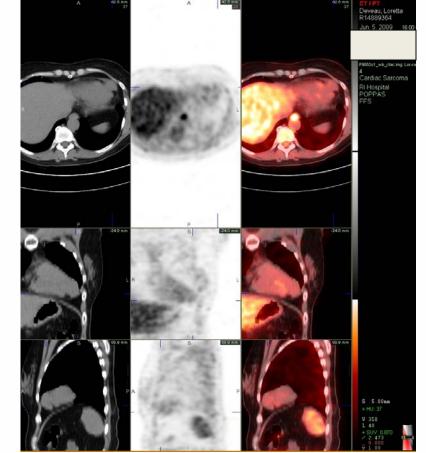
Three samples from RV septum
 Normal endocardium





PET without FDG uptake: not malignant

No evidence of metastatic









## **Preoperative Angiogram**

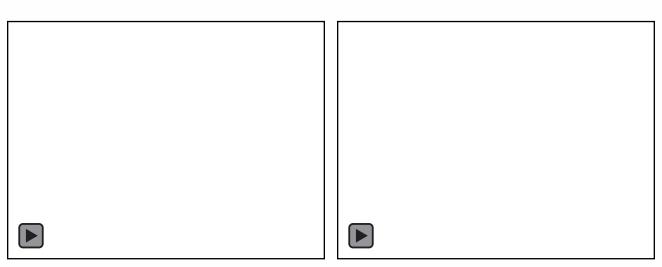
Tumor blush from LAD septal perforators







#### **Intraoperative TEE**



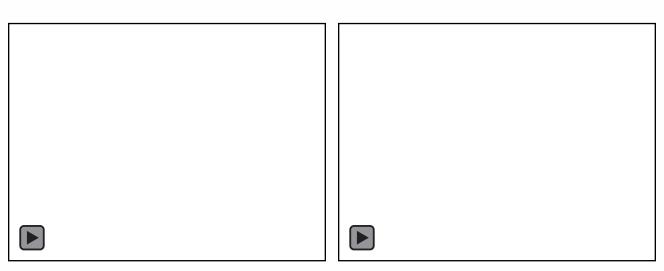
Transgastric 90 RV

transgastric 120 RV, 3 papillary muscles





## **Intraoperative TEE**



Deep Transgastric RV, nml TV

Transgastric SAX RV





#### **Postop Echoes**



Intraoperative, post CPB TEE

Postoperative day 7 TTE





#### **Intraoperative Pathology**



Diagnosis: spindle cell tumor Benign Fibroma









#### Take home messages

- 1. More imaging: To define extent and etiology
  - Cardiac CT
  - TEE
  - MRI
  - PET
  - Coronary angiogram
- 2. Endomyocardial Biopsy
- 3. Surgery: careful planning by the heart team

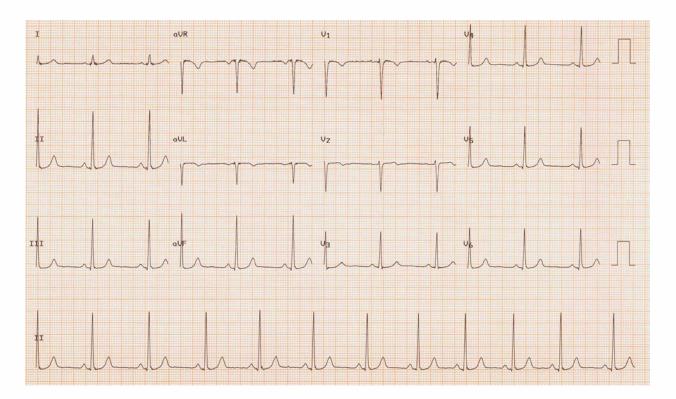
#### Case presentation II

- 18yow with exertional chest pain and syncope presents at 38 weeks gestation.
  - Age 17 ED visit for same
  - echo report "possible anomalous coronary"
- PE: BMI 35, 110/60 and equal, HR 90
  - CV: 1/6 SEM, normal S2, no S3
  - Lungs are clear
  - Abdomen gravid
  - Extremities pulses equal 2+ no edema
- ECG:





#### **ECG**







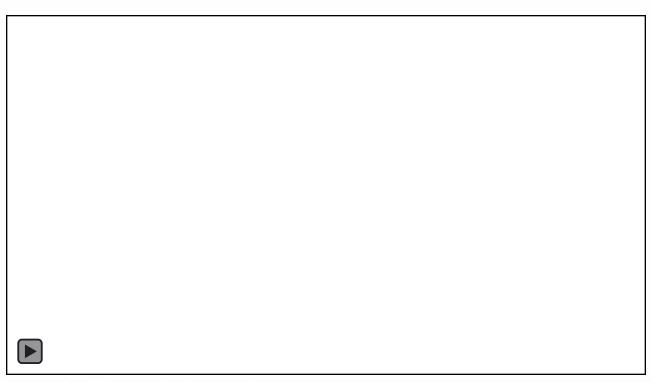
#### **Question I:**

#### What would you do next?

- 1. Repeat the echo
- 2. CT angio
- 3. Cardiac catheterization
- 4. MRI
- 5. Deliver the baby



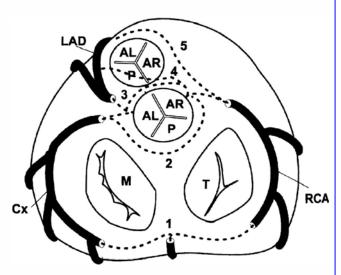








#### **Anomalous Aortic Origin of Coronary Artery: AAOCA**



Angelini Circ. 2007;115:1296-1305

- LM/LAD from right sinus
- RCA from left sinus
  - Course between AO and PA may result in compression with ischemia/SCD
  - Incidence 0.1-0.3% births
- Registry of SCD athletes
  - 2<sup>nd</sup> most common,13% of 286
- SCD in military recruits
  - Most common etiology
    - JACC 2003;41:974
    - Ann Int Med 2004;141:829.
- Low SCD if asymptomatic
  - JACC 2007;50:2083







## Estimated fetal and maternal effective doses of diagnostic and interventional radiology procedures

Procedure	Fetal exposure		Maternal exposure	
Chest radiograph (PA and lateral)	<0.01 mGy	<0.01 mSv	0.1 mGy	0.1 mSv
CT chest	0.3 mGy	0.3 mSv	7 mGy	7 mSv
Coronary angiography <sup>a</sup>	1.5 mGy	1.5 mSv	7 mGy	7 mSv
PCI or radiofrequency catheter ablation <sup>a</sup>	3 mGy	3 mSv	15 mGy	15 mSv



## **ESC Guidelines Helpful**

Echocardiography should be performed in any pregnant patient with unexplained or new cardiovascular signs or symptoms.	1	С
MRI (without gadolinium) should be considered if echocardiography is insufficient for diagnosis.	lla	С
A chest radiograph, with shielding of the fetus, may be considered if other methods are not successful in clarifying the cause of dyspnoea.	ПЬ	С
Cardiac catheterization may be considered with very strict indications, timing, and shielding of the fetus.	IIb	U
CT and electrophysiological studies, with shielding of the fetus, may be considered in selected patients for vital indications.	IIb	U
Coronary bypass surgery or valvular surgery may be considered when conservative and medical therapy has failed, in situations that threaten the mother's life and that are not amenable to percutaneous treatment.	IIb	С

Balance risk to mother of NOT doing the test





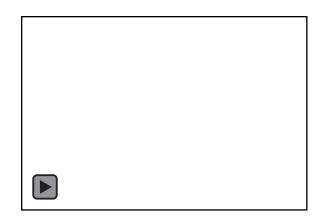
#### **Treatment Plan**

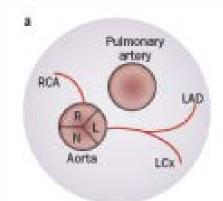
- Initiate betablocker
- Admit to hospital
- Proceed with induction of labor
- Pain management with epidural
- Early referral to congenital heart surgeon



#### **Take Home Points**

- Anomalous Coronaries:
  - Course and symptoms
  - Echo and CT both
- CT scan is safe in pregnancy
  - care for two
- Team approach with expertise
  - L&D hemodynamically labile

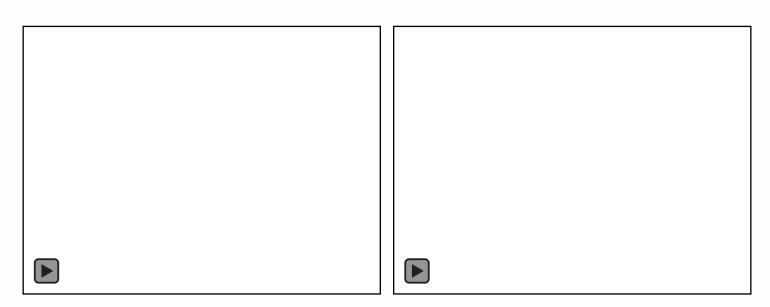








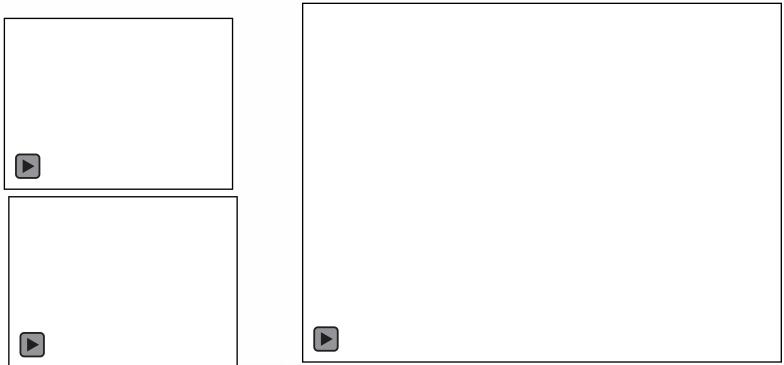
#### CASE III: What is the etiology of the AI?







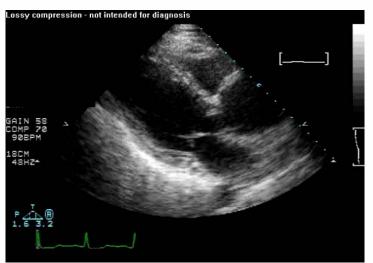
#### Trauma with avulsion of AV at annulus

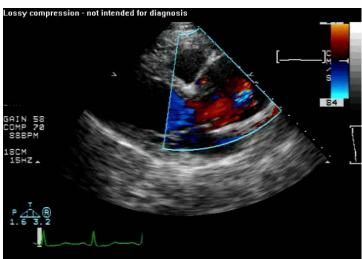






#### Aortic dissection and severe AV

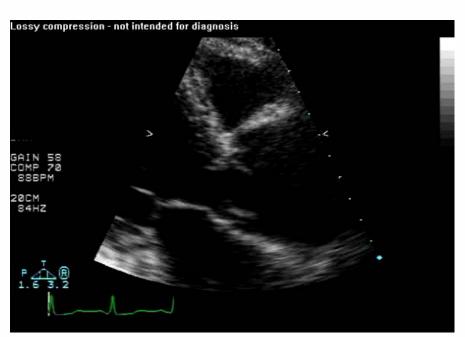


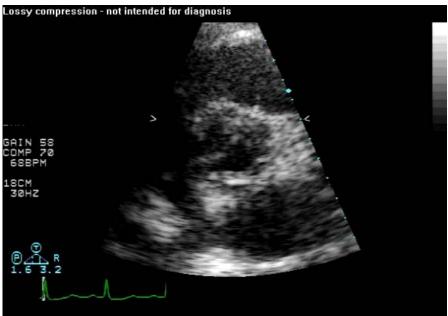






## Dissection flap prolapsing across AV





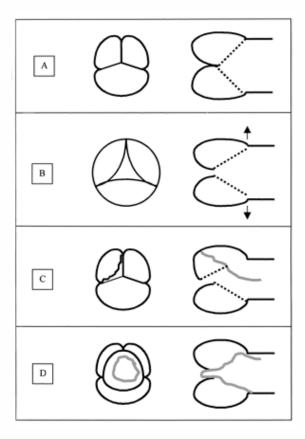




#### TEE in Dissection: Repair or Replace

- A. Normal AV
  - Leaflet tips coapt
  - STJ = annulus
- B. Incomplete closure
  - Dilated STJ
- C. Leaflet prolapse
  - Leaflet attachment disrupted
- D. Dissection Flap prolapse
  - Nml AV leaflets

Movosowitz HD et al. JACC 2000;36:884

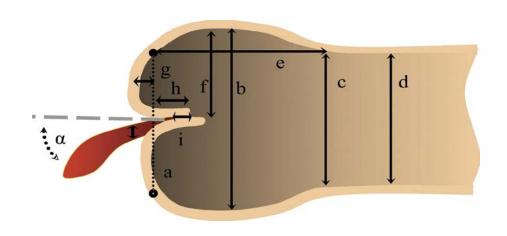






#### TEE to guide in the OR: Repair or Replace?

	With the Cox Multivariate Analysis					
Multivariate Analysis	HR	95% Confidence Interval	Cox p Value			
Coaptation length	0.82	0.63-1.00	0.05			
Tips below the level of the aortic annulus	7.9	6.52–9.28	<0.01			
Diameter of aortic annulus	1.18	1.03-2.45	0.01			
Residual AR	5.3	1.47-6.57	0.01			

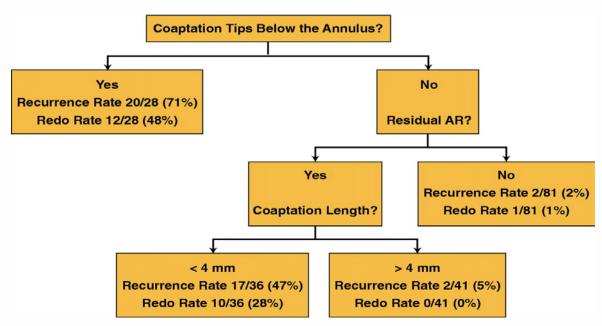


Schematic Representation of the TEE Measurements

le Polain de Waroux, J.-B. et al. J Am Coll Cardiol Img 2009;2:931-939



#### Risk of AV Repair Failure According to immediate postop TEE

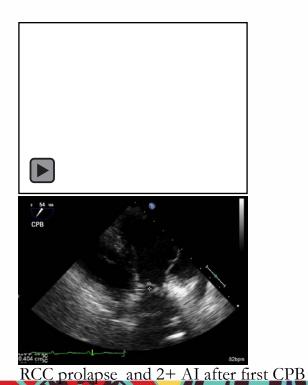


le Polain de Waroux, J.-B. et al. J Am Coll Cardiol Img 2009;2:931-939





#### Assess after root replacement/valve repair





No prolapse or AI after rerepair



#### Surgical Classification of Al

- Similar to Carpentier for MV disease
  - Functional classification to guide repair techniques...
    - » VanDyck M. Anesth Analg 2010;111:59
- TEE coorelated w/ OR inspec
  - 1. Enlarged Aorta & normal cusps
  - 2. Cusp prolapse (or fenestration): eccentric jets
  - 3. Cusp retraction (poor quality/quantity)
    - » El Khoury Circ 2007;116:1264.
- 781 chronic AI elective surgeries/5 yrs
  - 1. Central: dilation only
  - 2. Eccentric: dilation and valve
    - » Lansac E. EJCS 2008;33(5):872-878.



# Classification scheme: predicted short and long term repair success

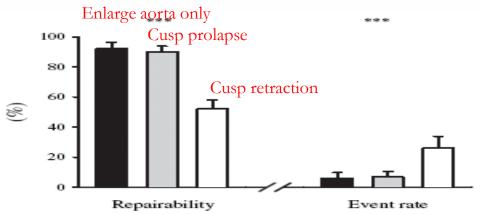


Figure 2. Incidence of valve sparing or repair surgery in the whole population (left) and event rate in patients undergoing sparing or repair surgery (right), according to anatomic classification by TEE. Closed bars, type 1; Half-tone bars, type 2; Open bars, type 3. \*\*\* $\chi^2$ <0.001 type 3 vs type 1 and 2.

El Khoury Circ 2007;116;I264

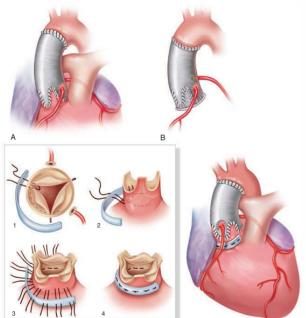






# Surgical Treatment: valve sparing aortic root replacement

A. Remodeling of root:
Replacement of sinuses



B. Reimplantation of valve: for annuloaortic ectasia

Aortic annuloplasty:
For annuloaortic ectasia

David TE: Aortic root aneurysms: Remodeling or composite replacement? Ann Thorac Surg 64:1564, 1997







#### Take home message

- Imaging in crucial:
  - preop to define etiology and valve structure suitability
  - intraop for successful repair
- Understand surgical approaches
  - communicate structure and function



#### Classification scheme: surgical techniques

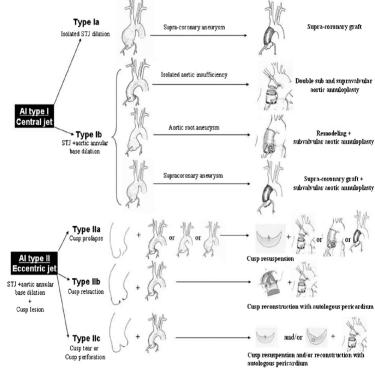


Fig. 2. Lesional classification of aortic insufficiencies (AI) and adapted surgical strategies. STJ: sino-tubular junction.

Lansac EJCS 2008;33:872-878.



