Case Presentation

Rebecca T. Hahn

Mark Lebehn

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History

- Ms XY is a 33 yo woman with a history of PVCs, PACs, mitral valve leaflet prolapse, and mild mitral regurgitation.
  - Treadmill stress ECG terminated in less than 1 minute due to frequent PVCs and NSVT (5 beats).

  Medications: metoprolol ER 100 mg daily

  PSH: None

  FH: Father- CAD; Mother- HLD

  SH: Independent for ADLs. Non smoker. Rare social drinker. Runs a Hebrew School and works in the home.

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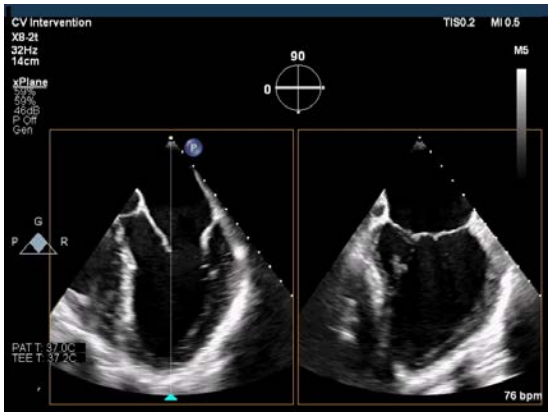
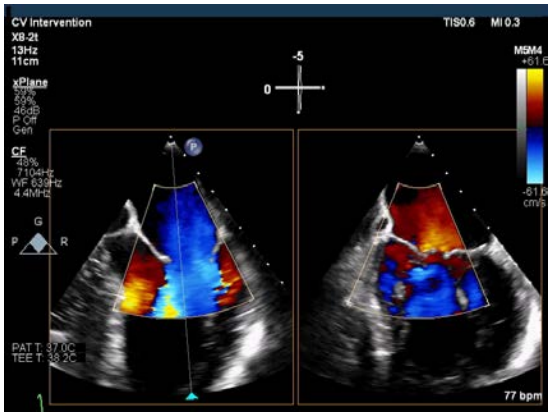
TEE to Assess Severity of MR





What is the etiology of the arrhythmia?

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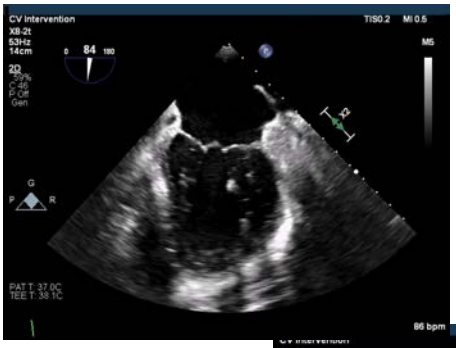

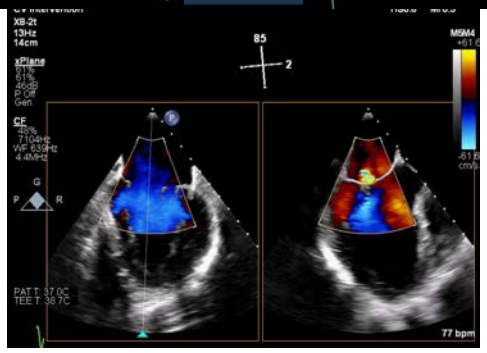

Arrhythmic Mitral Valve Prolapse Syndrome (with Mitral Annular Disjunction)

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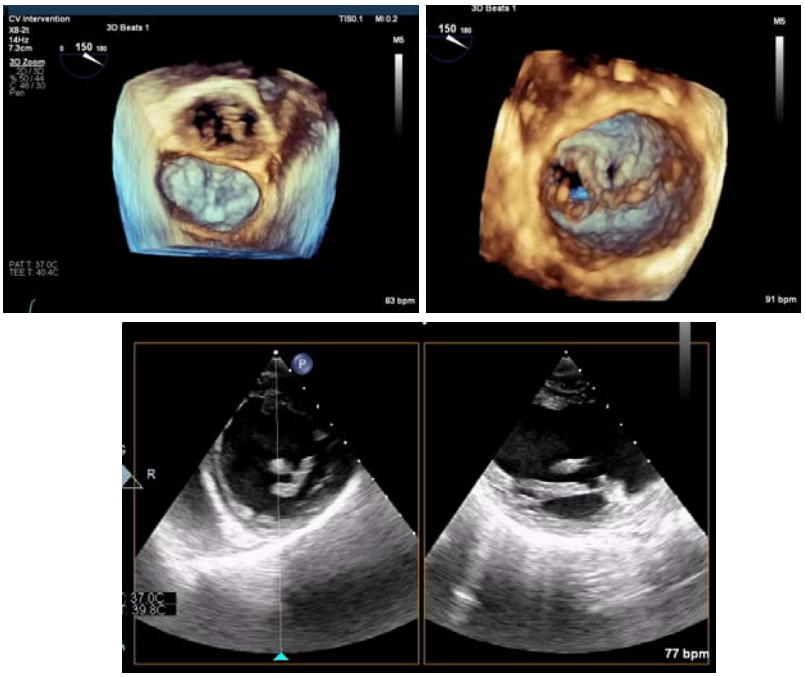
Arrhythmic Mitral Valve Prolapse Syndrome (with Mitral Annular Disjunction)



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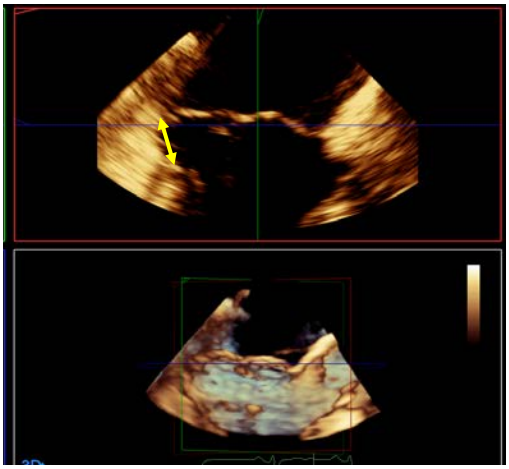

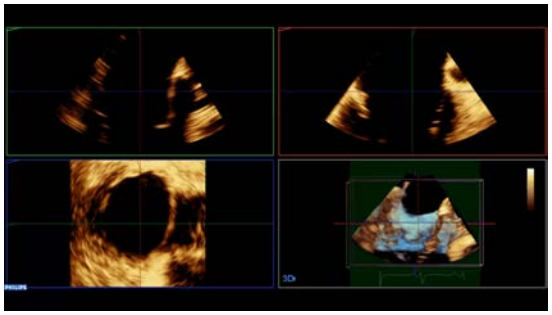

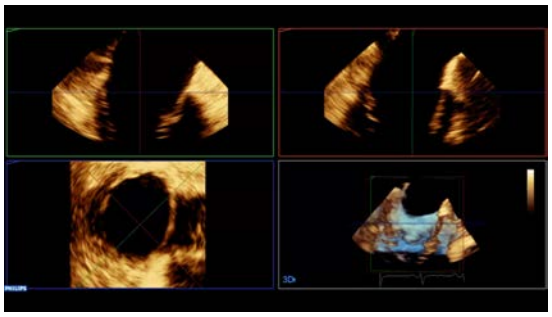

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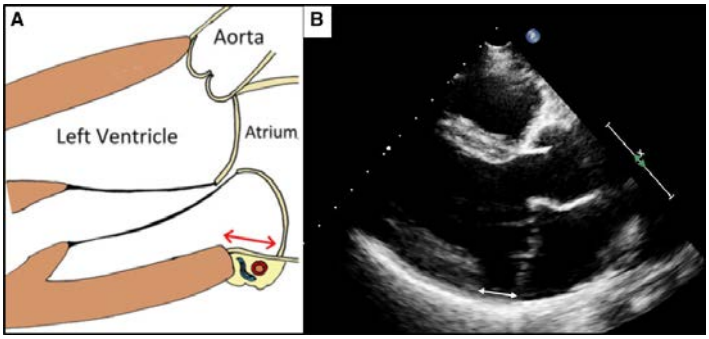
Measurement of MAD



- MAD is measured from the left atrial wall–MV posterior leaflet junction to the top of the LV posterior wall during end-systole (double-headed gray arrow

Basso et al. Circulation. 2019;140:952–964.

MAD with disjunction > 8.5 mm was associated with nonsustained ventricular tachycardia (OR 10 95% CI 1.28-78.1).

Bennett S, Thamman R, Griffiths T, Oxley C, Khan JN, Phan T, Patwala A, Heatlie G, Kwok CS. **Mitral annular disjunction: A systematic review of the literature.** Echocardiography. 2019;36:1549–1558.

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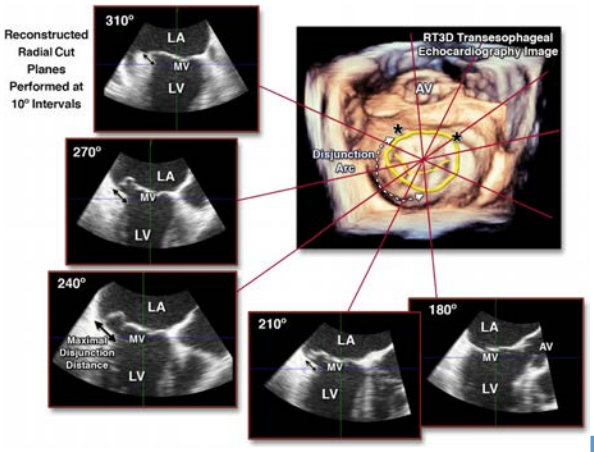
RT3DE Datasets of the Annulus



- There is annular disjunction **(doubled arrows)** spanning circumferentially from 210° to 310° (i.e., disjunction arc degree = 100°).
- The maximal disjunction distance, defined as the maximal separation between the atrial wall–MV attachment and the basal LV musculature, is 10 mm.
- The disjunction index, calculated as the product of the disjunction arc degree and the maximal disjunction distance, is 100° × 10 mm = 1,000° · mm.

Yellow line depicts the true atrial–ventricular junction.
Asterisks indicate the fibrous trigones

Most common location is P2 and P1 (less common P3).

Lee AP, et al. JACC Cardiovasc Imaging. 2017 Dec;10(12):1424-1433.

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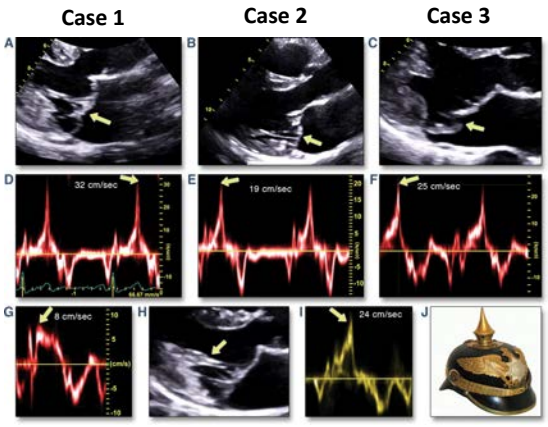
Pickelhaube Sign



- TTE demonstrating myxomatous bileaflet MVP **(arrows)**
- High-velocity mid-systolic spikes on tissue Doppler (lateral annulus) at different times in systole
- G represents a normal medial annulus tissue Doppler

The Pickelhaube Sign: Novel Echocardiographic Risk Marker for Malignant Mitral Valve Prolapse Syndrome.
Muthukumar L, Rahman F, Jan MF, Shaikh A, Kalvin L, Dhala A, Jahangir A, Tajik AJ.
JACC Cardiovasc Imaging. 2017 Sep;10(9):1078-1080. doi: 10.1016/j.jcmg.2016.09.016. Epub 2016 Dec 21.

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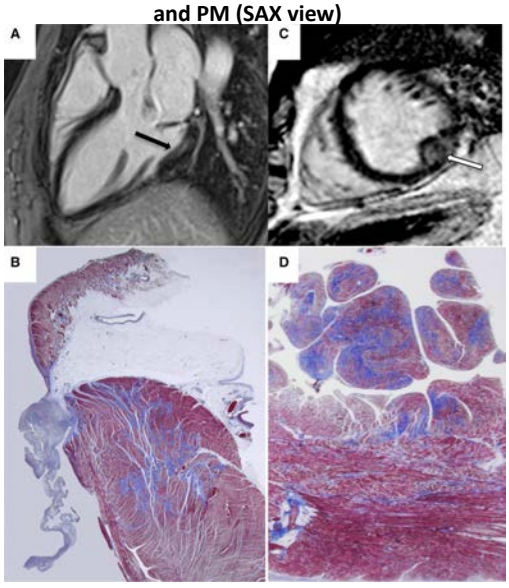
Myocardial fibrosis in arrhythmic MVP

- Basso et al. demonstrated a high prevalence (88%) of myocardial fibrosis in the inferobasal wall on autopsy of young adults with MVP who experienced sudden cardiac death.

Cristina Basso et al. Mitral Valve Prolapse, Ventricular Arrhythmias, and Sudden Death. Circulation. 2019;140:952–964.

Mid-mural LGE in the LV inferobasal region (LAX) and PM (SAX view)



Equivalent Histopathology (fibrosis = blue)

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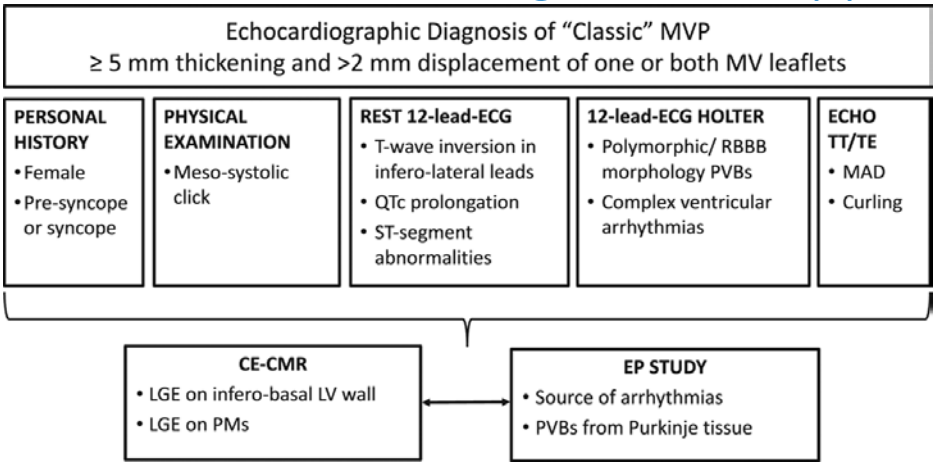
Clinical Profile and Diagnostic Tools For Risk Stratification and Targeted Therapy



Basso et al. Circulation. 2019;140:952–964.

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January 22nd, 2020

Case Presentation: Is This Severe AS or Pseudo AS (Echo and CT Imaging)

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1

Case

Clinical History	
Age	83 years
Gender	Male
Medical history	Heart Failure, HTN, Hypercholesterolemia S/P MI, MidLAD and RCA Stent, S/P MitraClip
Rx	ASA 81 mg QD Metoprolol ER 50 mg QD Furosemide 40 mg QD Spironolactone 37.5 mg QD Atorvastatin 20 mg QD

Heart Failure NYHA class III, ACC/AHA stage C

Creatinine 0.81 mg/dL

NT pro-BNP 9 952 pg/mL

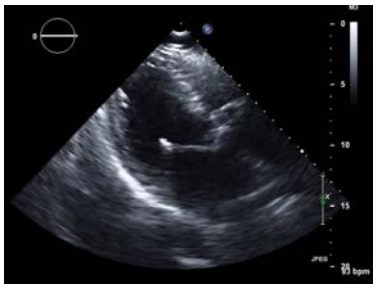
ECG: Sinus rhythm

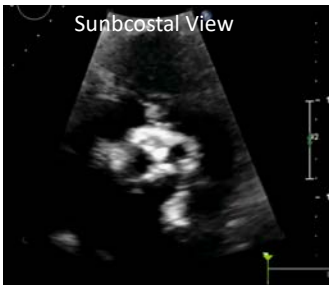
Physical Examination
Height (cm): 177, Weight (kg): 70 BSA 1.87 m^2
BP 124/70; HR 68 bpm
No jugular venous distention Soft S2 III/VI SM RUSB Clear lungs No edema

2

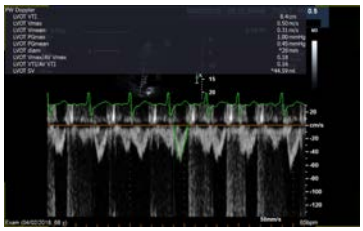
Baseline TTE

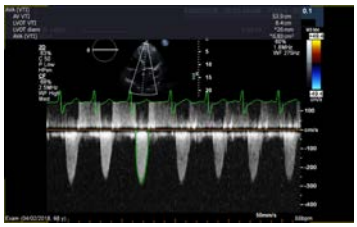
Echo Variable (TTE/TEE)	Measure
Jet Velocity (m/s)	2.98
Mean Gradient (mmHg)	21.3
Calculated AVA (cm^2)	0.83
Calculated AVA index (cm2/m2) **1.87 m2** **BMI = 22.3 kg/m^2**	0.46 (severe)
DVI	0.16
TTE LVOT TTE annulus diameter	2.55 cm 2.62 cm
Ejection Fraction (%)	35%
LV Stroke Volume (ml)	44.6 ml 24 ml/m^2
Severity of AR	1-2+
Severity of MR	1-2+
RV Pressure (mmHg)	38 mmHg





Very severe calcification of the aortic valve





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Baseline EF 35%

S/P RCA and LAD stents

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Stages of Valvular AS

Stage	Definition	Valve Anatomy	Valve Hemodynamics
D1	Symptomatic severe high-gradient AS	Severe leaflet calcification or congenital stenosis with severely reduced leaflet opening	• Aortic Vmax ≥4 m/s or mean ΔP ≥40 mm Hg • AVA typically is ≤1.0 cm2 (or AVAi ≤0.6 cm2/m2) but may be larger with mixed AS/AR
D2	Symptomatic severe low-flow/low-gradient AS with reduced LV EF	Severe leaflet calcification with severely reduced leaflet opening	• AVA ≤1.0 cm2 with Aortic Vmax <4 m/s or mean ΔP <40 mm Hg • Dobutamine stress echocardiography shows AVA ≤1.0 cm2 with Vmax ≥4 m/s at any flow rate
D3	Symptomatic severe low-gradient AS with normal LVEF or paradoxical low-flow severe AS	Severe leaflet calcification with severely reduced leaflet opening	• AVA ≤1.0 cm2 with Aortic Vmax <4 m/s or mean ΔP <40 mm Hg • AVAi ≤0.6 cm2/m2 and • Stroke volume index <35 mL/m2 • Measured when patient is normotensive (systolic BP <140 mm Hg)

Nishimura, RA et al JAmCollCardiol 2014 Jun 10;63(22):2438-88

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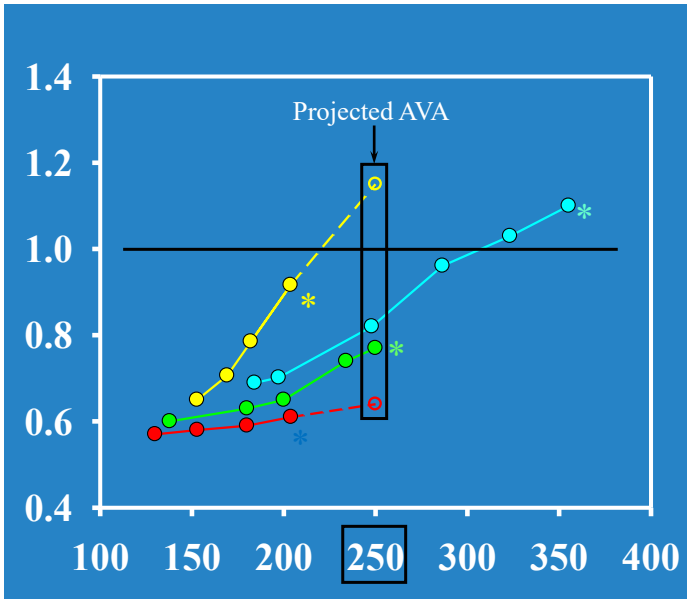
Concept of the Projected AVA (250 mL/s)

Simplified method:
Slope=ΔAVA/ ΔQ

AVAproj=
AVA_{rest}+slope(250-Q_{rest})



Slide courtesy of Philippe Pibarot

* Peak AVA during DSE

Blais et al, Circulation 2006;113:711-721

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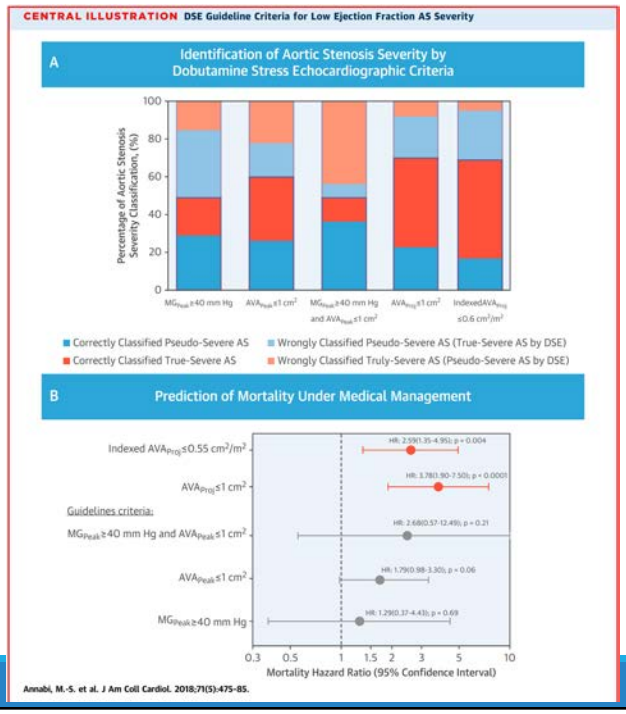
TOPAS Registry

Up to 50% of patients remain with discordant grading at DSE

Projected AVA (250 ml/s) ≤1.0cm² was more accurate than DSE peak AVA or peak ΔP_{mean} for distinguishing "True-Severe" vs. "Pseudo-Severe" AS



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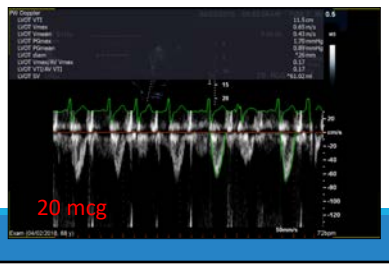
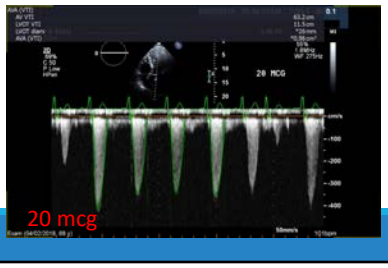
Dobutamine Stress Case 2

	LVOT VTI (cm)	SV (ml)	ET (msec)	Flow (ml/s)	AV Pk Vel (m/s)	AV Mn Grad (mmHg)	AV VTI (cm)	DVI	AVA (cm^2)
Rest	8.4	44.6	264	169	2.98	21.3	53.9	0.16	0.83
5 mcg	9.7	51.5	264	195	3.08	20.2	53	0.18	0.96
10 mcg	9.9	52.6	235	223	3.07	21.0	51	0.19	1.00
20mcg	11.04	56.4	232	**253**	3.52	23	55.4	0.20	1.02



31% increase in stroke volume



AVAi = 0.545

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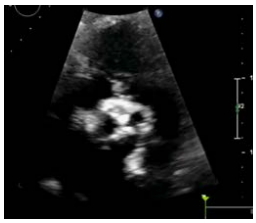
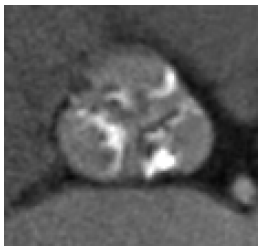
D2: Low Flow, Low EF

Baseline	
Peak gradient	35.5 mmHg
Mean gradient	21.3 mmHg
AVA	**0.83 cm^2 (0.44 cm^2/m^2)**
DVI	0.16
SV	44.6 ml (24 ml/m^2)
LVEF	25%

Peak dose (20 mcg/kg/min)	
Peak gradient	49 mmHg
Mean gradient	23 mmHg
AVA	**1.02 cm^2 (0.55 cm^2/m^2)**
DVI	0.20
SV	58.6 ml (31.3 ml/m^2)
LVEF	30%



Subcostal SAX View



Calcium Score = 2750 AU

31% increase in stroke volume with dobutamine
Severe aortic stenosis by indexed AVA

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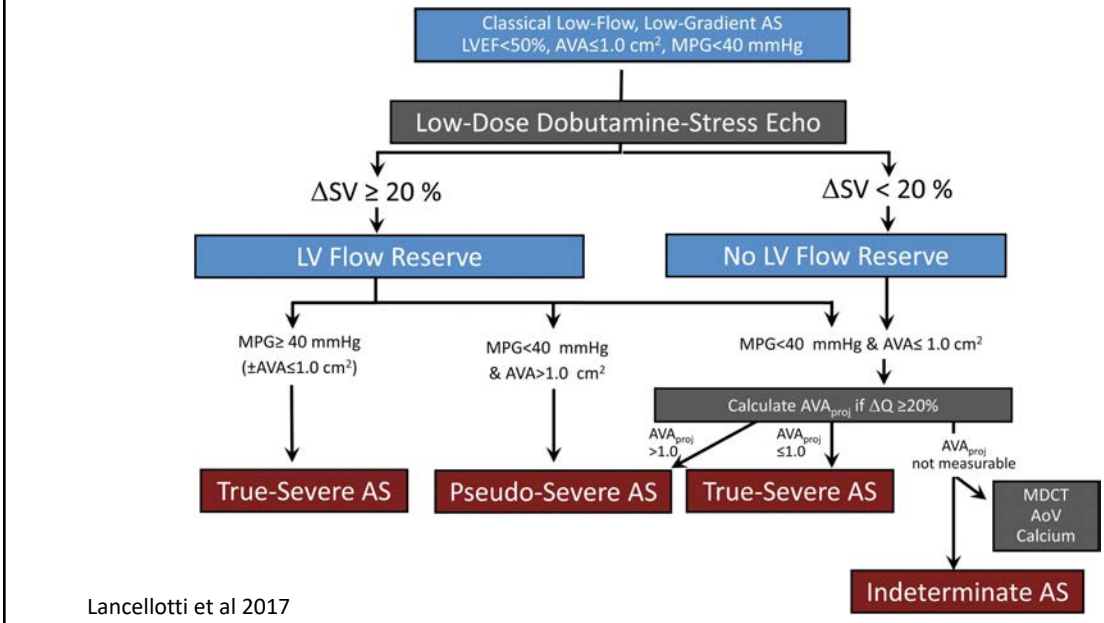

Lancellotti et al 2017

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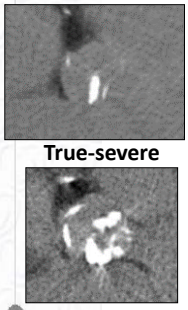
Criteria that increase the likelihood of severe AS in pts. with AVA < 1.0 cm², mean gradient < 40 mmHg and preserved EF

(Baumgartner et al)

Criteria *(continued)*	
Quantitative imaging data *(continued)*	• Low flow (SVi <35 mL/m²) confirmed by techniques other than standard Doppler technique (LVOT measurement by 3D TOE or MSCT; CMR, invasive data).
	• Calcium score by MSCT: – Severe aortic stenosis very likely: men ≥3000; women ≥1600, – Severe aortic stenosis likely: men ≥2000; women ≥1200, – Severe aortic stenosis unlikely: men <1600; women<800.



www.escardio.org/guidelines

2017 ESC/EACTS Guidelines for the Management of Valvular Heart Disease (European Heart Journal 2017 - doi:10.1093/eurheartj/ehx391)

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Guidelines Indications for AVR in Classical Low-Flow, Low-Gradient AS

Stage D2 Definition: AVA≤1.0 cm², Mean gradient<40 mmHg, LVEF<50%

Guidelines	Recommendation for AVR	Class
ACC-AHA 2014/2017	AVR is reasonable in symptomatic patients with low LVEF, low-flow/low-gradient severe AS **with a DSE that shows a mean gradient ≥40 mm Hg with an AVA ≤1.0 cm²** at any dobutamine dose	IIa
ESC-EACTS 2017	AVR should be considered in symptomatic patients with low LVEF, low-flow/low-gradient **severe AS (mean gradient ≥40 mmHg) with flow reserve on DSE**	I
ESC-EACTS 2017	AVR may be considered in symptomatic patients with low LVEF, low-flow/low-gradient **severe AS without flow reserve on DSE, particularly when CT calcium scoring confirms severe AS**	IIa

Vahanian et al. EHJ 2012 *Nishimura, Otto et al. JACC 2014*

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