1

A Not-So-Common Case Of Cardiomyopathy

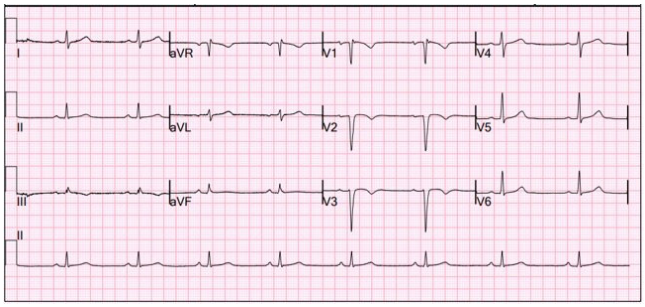
Tareq Al Saadi

12/18/2023

2

Initial Presentation

- 41 yo, PMHx of HIV [normal CD4 count and non-detected viral load], smoker, works as a truck driver
- No family history of SCD or cardiomyopathy.
- cc: syncope
- hsTroponin peaked at 600, TWI on EKG
- ESR/CRP normal
- CTPE negative for PE.
- CTA aorta negative for dissection.
- CCTA: no CAD, calcium score of 0



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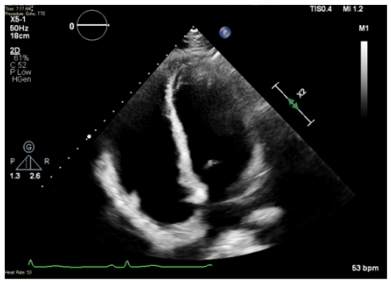
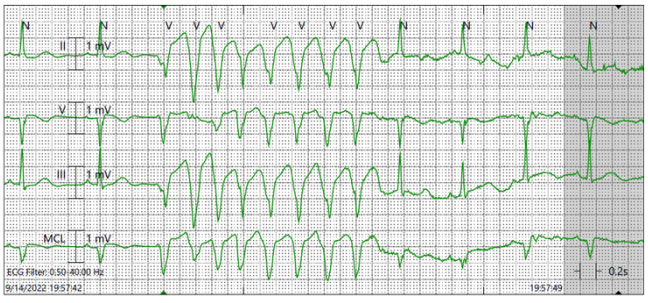
Echocardiogram



- LVEF 57%
- No regional wall motion abnormalities.
- Borderline RV enlargement
- No significant valve abnormalities.

Telemetry



- 2 symptomatic NSVT episodes

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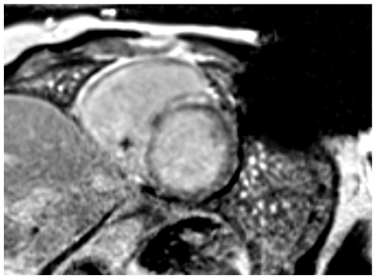
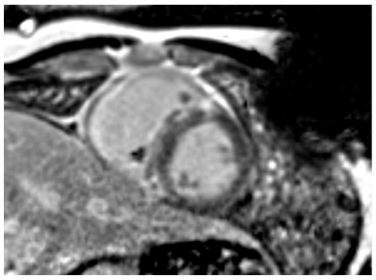
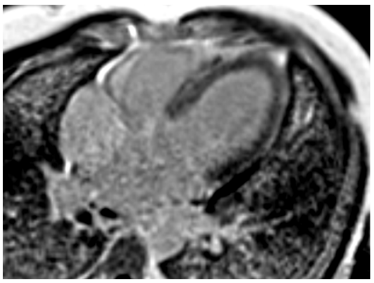
CMR

- Non-ischemic mid myocardial delayed enhancement involving the basal to mid interventricular septum, especially on the RV side of the septum. RV insertion sites delayed enhancement also present.
- Mild RV enlargement, mild RV systolic dysfunction.

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2010 revised criteria for diagnosis of ARVC

I. Global or regional dysfunction and structural alterations	
Major	By 2D echo: ● Regional RV akinesia, dyskinesia, or aneurysm ● and 1 of the following (end diastole): — PLAX RVOT ≥ 32 mm (corrected for body size PLAX/BSA ≥ 19 mm/m2) — PSAX RVOT ≥ 36 mm (corrected for body size PSAX/BSA ≥ 21 mm/m2) — or fractional area change ≤ 33% By MRI: ● Regional RV akinesia or dyskinesia or dyssynchronous RV contraction ● and 1 of the following: — Ratio of RV end-diastolic volume to BSA ≥ 110 mL/m2 (male) or ≥ 100 mL/m2 (female) — or RV ejection fraction ≤ 40% By RV angiogra ● Regional RV a
Minor	By 2D echo: ● Regional RV a ● and 1 of the f — PLAX RV — PSAX RV — or fracti By MRI: ● Regional RV akinesia or dyskinesia or dyssynchronous RV contraction ● and 1 of the following: — Ratio of RV end-diastolic volume to BSA ≥ 100 to < 110 mL/m2 (male) or ≥ 90 to < 100 mL/m2 (female) — or RV ejection fraction > 40% to ≤ 45%
II. Tissue characterization of wall	
Major	Residual myocytes < 60% by morphometric analysis (or 50% if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy
Minor	Residual myocytes 60% to 75% by morphometric analysis (or 50% to 65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

Diagnostic terminology:

- Definite diagnosis: 2 major or 1 major and 2 minor criteria or 4 minor from different categories.
- Borderline: 1 major and 1 minor or 3 minor criteria from different categories.
- Possible: 1 major or 2 minor criteria from different categories.

III. Repolarization abnormalities	
Major	Inverted T waves in right precordial leads (V1, V2, and V3) or beyond in individuals > 14 years of age (in the absence of complete right bundle-branch block QRS ≥ 120 ms)
Minor	● Inverted T waves in leads V1 and V2 in individuals > 14 years of age (in the absence of complete right bundle-branch block) or in V4, V5, or V6 ● Inverted T waves in leads V1, V2, V3, and V4 in individuals > 14 years of age in the presence of complete right bundle-branch block
IV. Depolarization/conduction abnormalities	
Major	Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V1 to V3)
	ard ECG
	including R, in V1, V2,
	ative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)
Minor	● Nonsustained or sustained ventricular tachycardia of RV outflow configuration, left bundle-branch block morphology with inferior axis (positive QRS in leads II, III, and aVF and negative in lead aVL) or of unknown axis ● > 500 ventricular extrasystoles per 24 hours (Holter)
VI. Family history	
Major	● ARVC/D confirmed in a first-degree relative who meets current Task Force criteria ● ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative ● Identification of a pathogenic mutation† categorized as associated or probably associated with ARVC/D in the patient under evaluation
Minor	● History of ARVC/D in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current Task Force criteria ● Premature sudden death (< 35 years of age) due to suspected ARVC/D in a first-degree relative ● ARVC/D confirmed pathologically or by current Task Force Criteria in second-degree relative

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Marcus, F. I., McKenna, W. J., Sherrill, D., Basso, C., Bauce, B., Bluemke, D. A., ... & Zareba, W. (2010). Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the task force criteria. *Circulation*, *121*(13), 1533-1541.

6

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Minor	By 2D echo: ● Regional RV akinesia or dyskinesia ● and 1 of the following (end diastole): — PLAX RVOT ≥ 29 to < 32 mm (corrected for body size PLAX/BSA ≥ 16 to < 19 mm/m2) — PSAX RVOT ≥ 32 to < 36 mm (corrected for body size PSAX/BSA ≥ 18 to < 21 mm/m2) — or fractional area change > 33% to ≤ 40% By MRI: ● Regional RV akinesia or dyskinesia or dyssynchronous RV contraction ● and 1 of the following: — Ratio of RV end-diastolic volume to BSA ≥ 100 to < 110 mL/m2 (male) or ≥ 90 to < 100 mL/m2 (female) — or RV ejection fraction > 40% to ≤ 45%
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IV. Depolarization/conduction abnormalities	
Major	Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V1 to V3)
Minor	● Late potentials by SAECG in ≥ 1 of 3 parameters in the absence of a QRS duration of ≥ 110 ms on the standard ECG ● Filtered QRS duration (fQRS) ≥ 114 ms ● Duration of terminal QRS < 40 V (low-amplitude signal duration) ≥ 38 ms ● Root-mean-square voltage of terminal 40 ms ≤ 20 V ● Terminal activation duration of QRS ≥ 55 ms measured from the nadir of the S wave to the end of the QRS, including R, in V1, V2, or V3, in the absence of complete right bundle-branch block
V. Arrhythmia	
Major	Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)
Minor	● Nonsustained or sustained ventricular tachycardia of RV outflow configuration, left bundle-branch block morphology with inferior axis (positive QRS in leads II, III, and aVF and negative in lead aVL) or of unknown axis ● > 500 ventricular extrasystoles per 24 hours (Holter)
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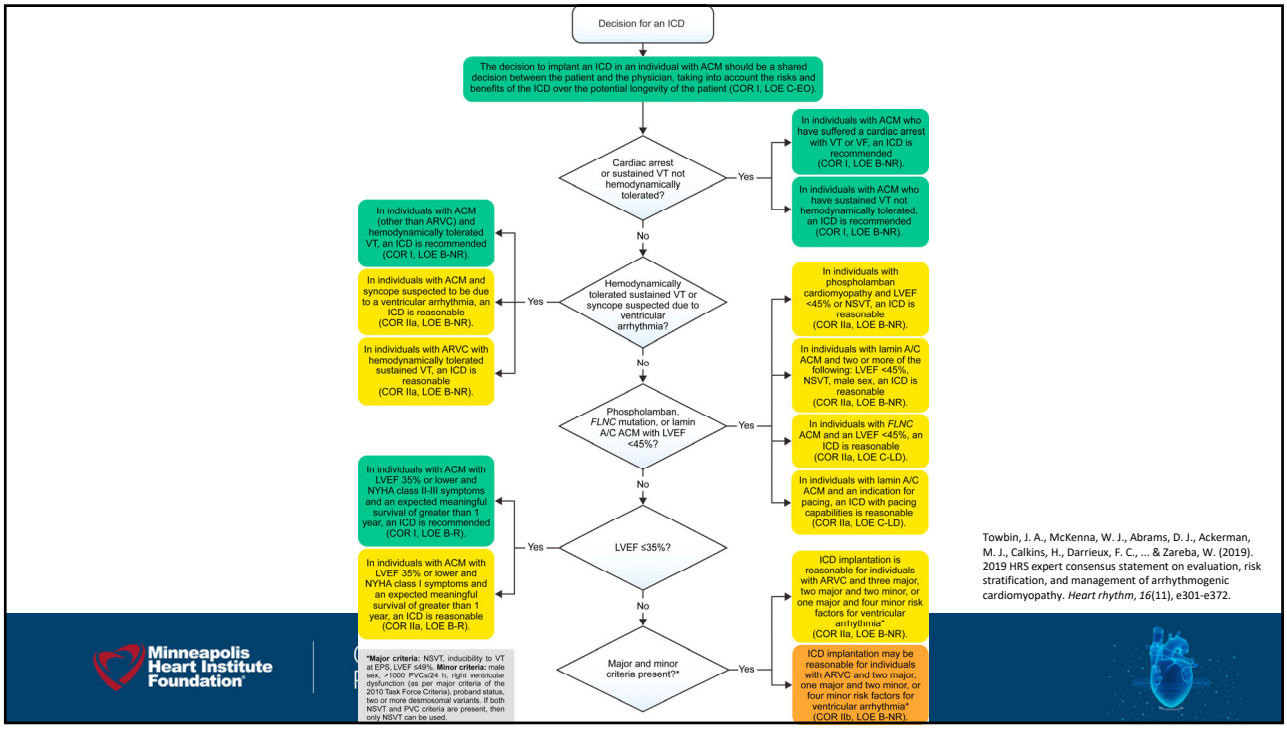

Towbin, J. A., McKenna, W. J., Abrams, D. J., Ackerman, M. J., Calkins, H., Darrieux, F. C., ... & Zareba, W. (2019). 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. *Heart rhythm*, *16*(11), e301-e372.

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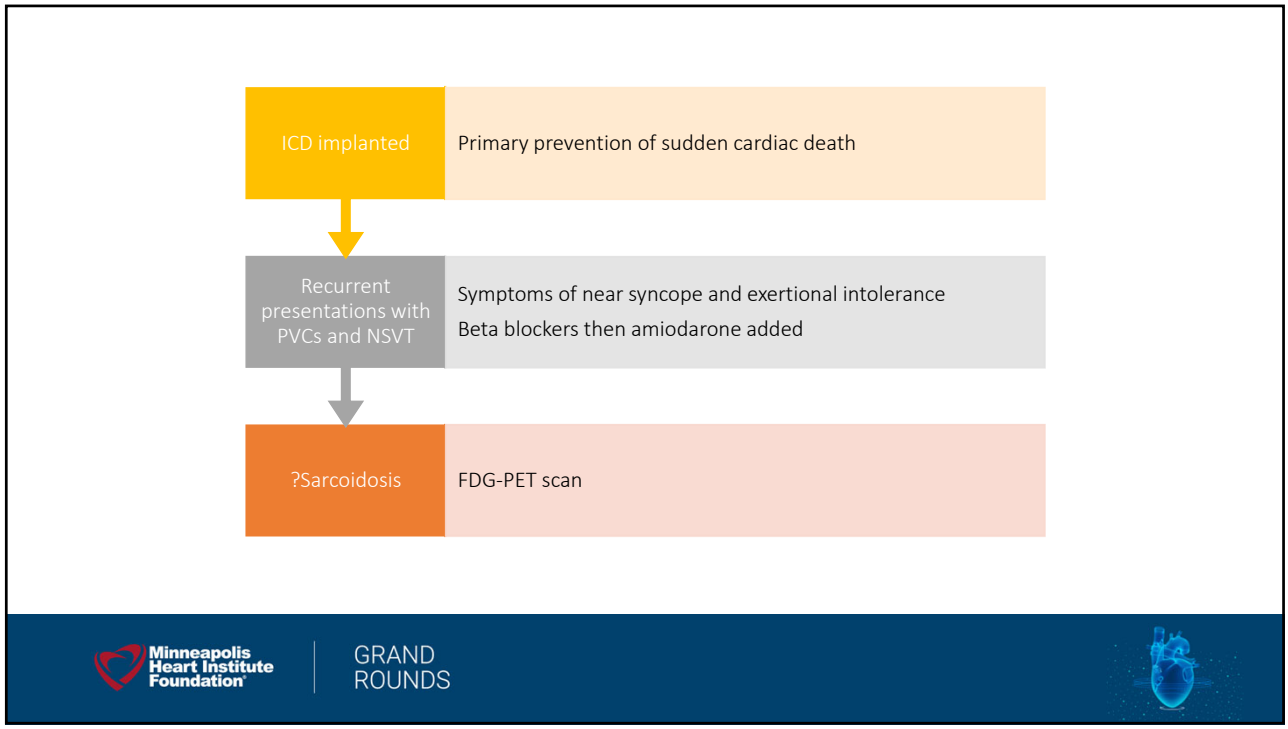
ICD implanted
Primary prevention of sudden cardiac death
Recurrent presentations with PVCs and NSVT
Symptoms of near syncope and exertional intolerance
Beta blockers then amiodarone added
?Sarcoidosis
FDG-PET scan
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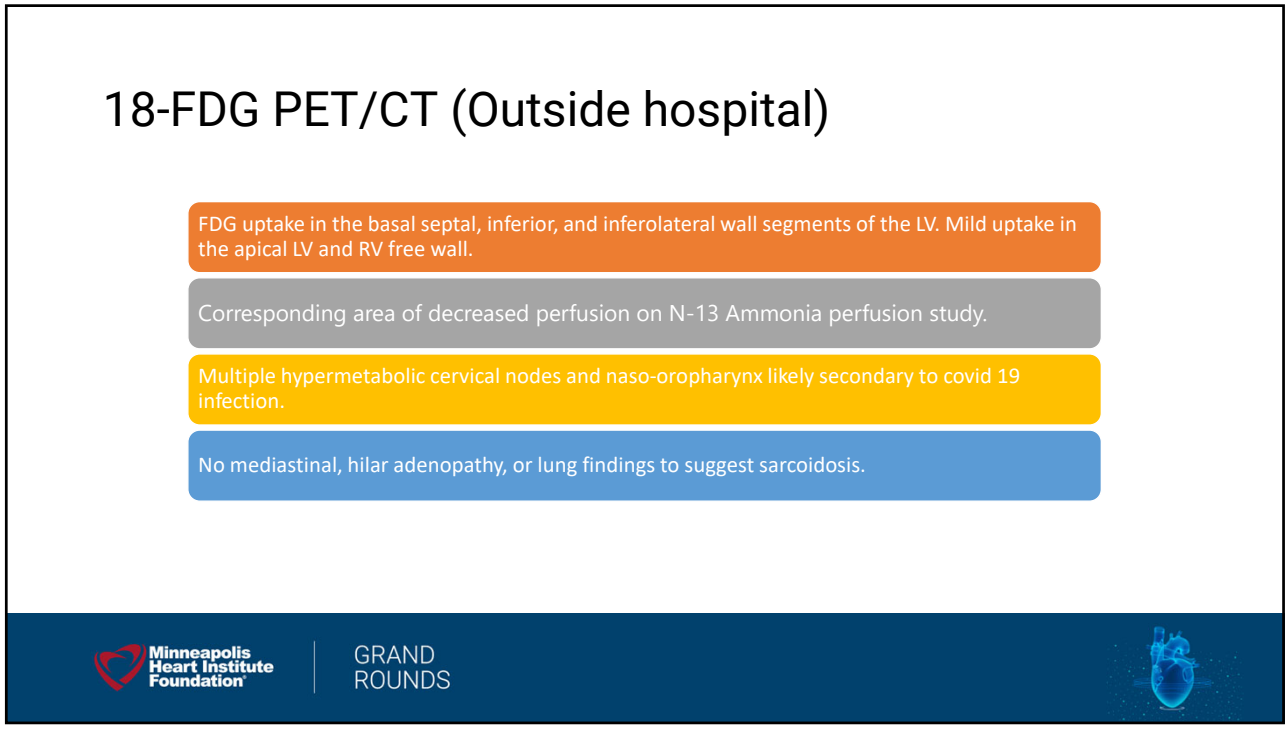
18-FDG PET/CT (Outside hospital)
FDG uptake in the basal septal, inferior, and inferolateral wall segments of the LV. Mild uptake in the apical LV and RV free wall.
Corresponding area of decreased perfusion on N-13 Ammonia perfusion study.
Multiple hypermetabolic cervical nodes and naso-oropharynx likely secondary to covid 19 infection.
No mediastinal, hilar adenopathy, or lung findings to suggest sarcoidosis.
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Biopsies

Cervical lymph node: Negative for metastatic tumor and granuloma.

Myocardium: patchy fibrosis. Negative for myocarditis, sarcoidosis, iron overload, amyloidosis, and ischemic changes.

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Type	ATP Seq	Shocks	Success	ID#	Date	Time hh:mm	Duration hh:mm:ss	Avg bpm A/V	Max bpm A/V
VT-Mon				7	12-Mar-2023	21:48	:03:38	89/164	107/171
VT-Mon				6	12-Mar-2023	21:42	:06:05	91/162	97/167
VT-Mon				5	12-Mar-2023	21:41	:36	98/156	---/167
VT-Mon				4	12-Mar-2023	21:37	:03:49	96/159	125/167
VT-Mon				3	11-Mar-2023	00:55	:12:30	110/173	130/182
VT-Mon				2	11-Mar-2023	00:40	:12:55	99/169	122/182
VT	5		Yes	1	10-Mar-2023	22:07	:39	125/194	---/200

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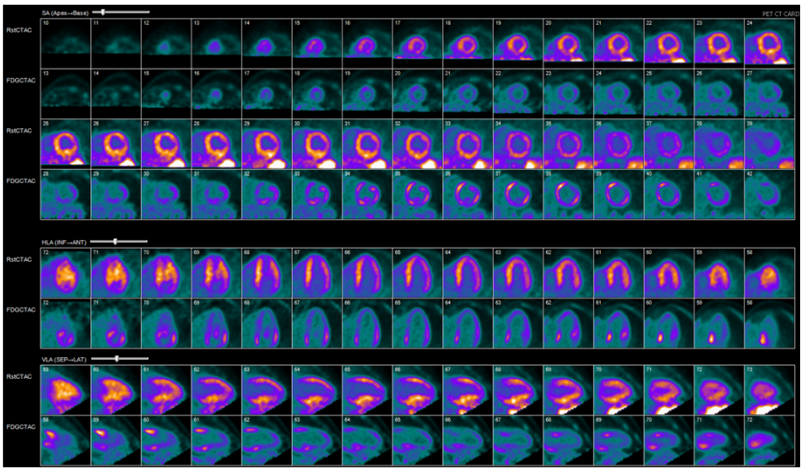
18-FDG-PET/CT

- Resting myocardial perfusion with rubidium was normal.
- With 18 FDG imaging there was uptake in the basal anteroseptal, anterolateral, inferior, inferoseptal, inferolateral, lateral and septal walls. SUV max: 5.4.
- There was no evidence of abnormal extracardiac activity.



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13

Expert Consensus Recommendations on ***Criteria for the Diagnosis of CS***

There are 2 pathways to a diagnosis of Cardiac Sarcoidosis:

1. Histological Diagnosis from Myocardial Tissue
   CS is diagnosed in the presence of non-caseating granuloma on histological examination of myocardial tissue with no alternative cause identified (including negative organismal stains if applicable).
2. Clinical Diagnosis from Invasive and Non-Invasive Studies:
   It is probable* that there is CS if:
   a) There is a histological diagnosis of extra-cardiac sarcoidosis
   and
   b) One or more of following is present
   - Steroid +/- immunosuppressant responsive cardiomyopathy or heart block
   - Unexplained reduced LVEF (<40%)
   - Unexplained sustained (spontaneous or induced) VT
   - Mobitz type II 2nd degree heart block or 3rd degree heart block
   - Patchy uptake on dedicated cardiac PET (in a pattern consistent with CS)
   - Late Gadolinium Enhancement on CMR (in a pattern consistent with CS)
   - Positive gallium uptake (in a pattern consistent with CS)

   and
   c) Other causes for the cardiac manifestation(s) have been reasonably excluded

*In general, 'probable involvement' is considered adequate to establish a clinical diagnosis of CS.[33]

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Birnie, D. H., Sauer, W. H., Bogun, F., Cooper, J. M., Culver, D. A., Duvernoy, C. S., ... & Soejima, K. (2014). HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. *Heart rhythm*, *11*(7), 1304-1323.

14

Treatment for cardiac sarcoidosis

Corticosteroids	First line agents High rates of recurrence in monotherapy
Immunomodulators	Upfront addition to conticosteroids in cases of rapidly progressive heart failure, life-threatening arrhythmias, and extensive inflammation on cardiac PET Methotrexate, azathioprine, mycophenolate mofetil, leflunomide, and cyclophosphamide
Biologic anti-tumor necrosis factor (TNF)	Third line agents Infliximab and adalimumab

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Lehtonen, J., Uusitalo, V., Pöyhönen, P., Mäyränpää, M. I., & Kupari, M. (2023). Cardiac sarcoidosis: phenotypes, diagnosis, treatment, and prognosis. European Heart Journal, 44(17), 1495-1510.

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Sarcoidosis treatment started

- Immunosuppression: Prednisone and Mycophenolate Mofetil
- Prophylactic TMP/SMX, nystatin, calcium + Vitamin D supplements.

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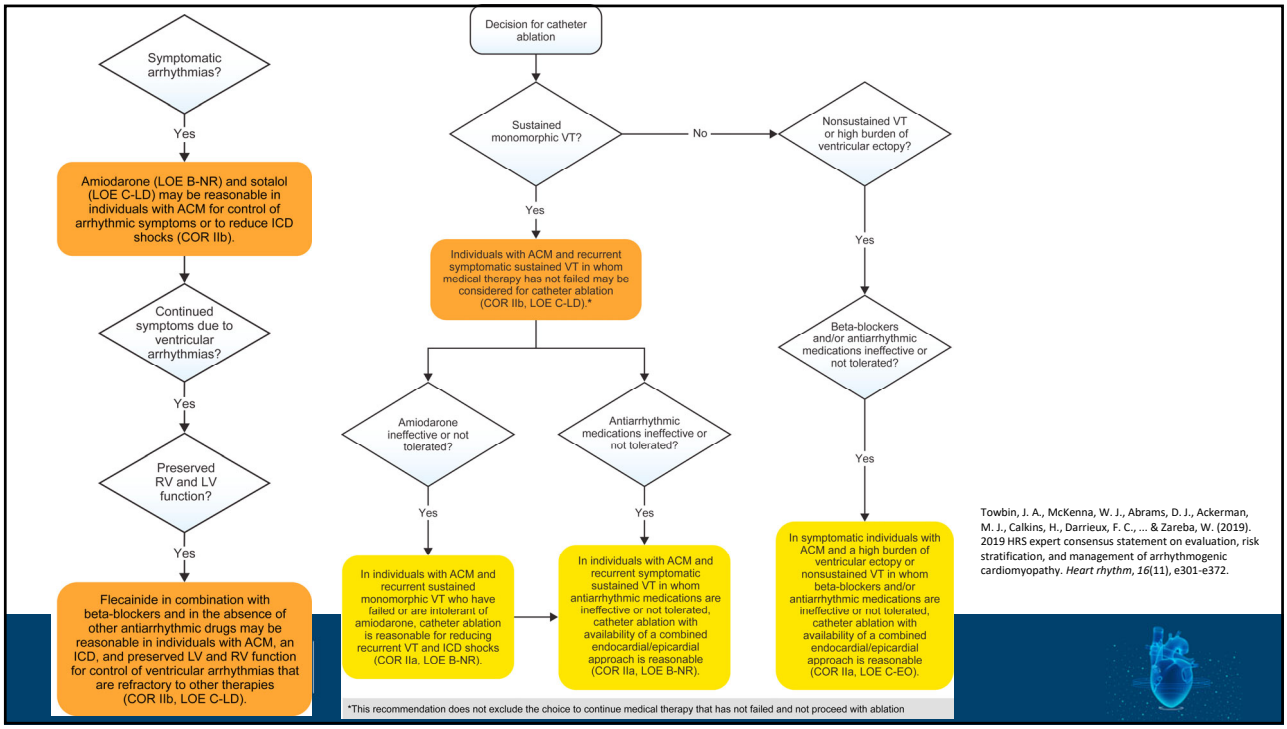

Towbin, J. A., McKenna, W. J., Abrams, D. J., Ackerman, M. J., Calkins, H., Darrieux, F. C., ... & Zareba, W. (2019). 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. *Heart rhythm*, *16*(11), e301-e372.

17

Expert Consensus Recommendations for the Management of Ventricular Arrhythmias

Class IIa

1. Assessment of myocardial inflammation with FDG-PET **can be useful** in CS patients with ventricular arrhythmias.
2. Immunosuppression **can be useful** in CS patients with frequent ventricular ectopy or nonsustained VT and evidence of myocardial inflammation.
3. Immunosuppression **can be useful** in CS patients with sustained ventricular arrhythmias and evidence of myocardial inflammation.
4. Antiarrhythmic medication therapy **can be useful** in patients with ventricular arrhythmias refractory to immunosuppressive therapy.
5. Catheter ablation **can be useful** in patients with CS and ventricular arrhythmias refractory to immunosuppressive *and* antiarrhythmic therapy.
6. Catheter ablation **can be useful** in patients with incessant ventricular arrhythmias.

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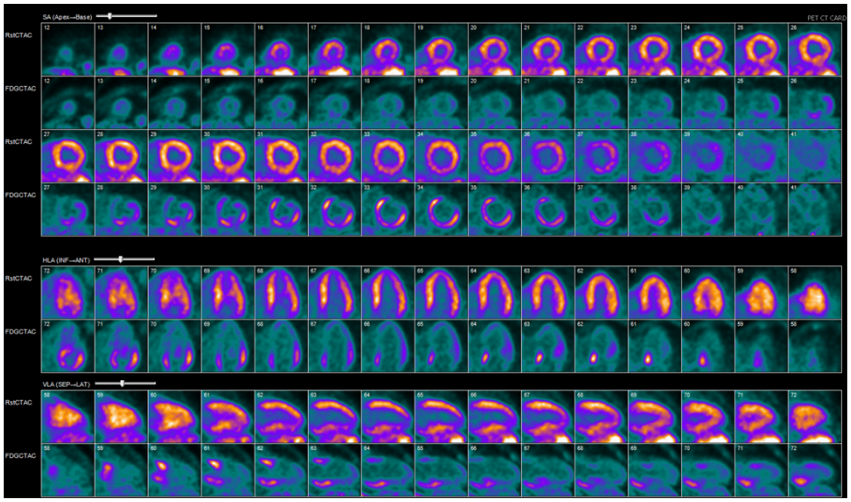
Birnie, D. H., Sauer, W. H., Bogun, F., Cooper, J. M., Culver, D. A., Duvernoy, C. S., ... & Soejima, K. (2014). HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. *Heart rhythm*, *11*(7), 1304-1323.

18

18-FDG-PET/CT - 5 months later

- Resting myocardial perfusion with rubidium was normal.
- With 18 FDG imaging there was uptake in the basal anteroseptal, inferior, inferoseptal, and inferolateral walls. SUV max: 4.7.
- There was no evidence of abnormal extracardiac activity.



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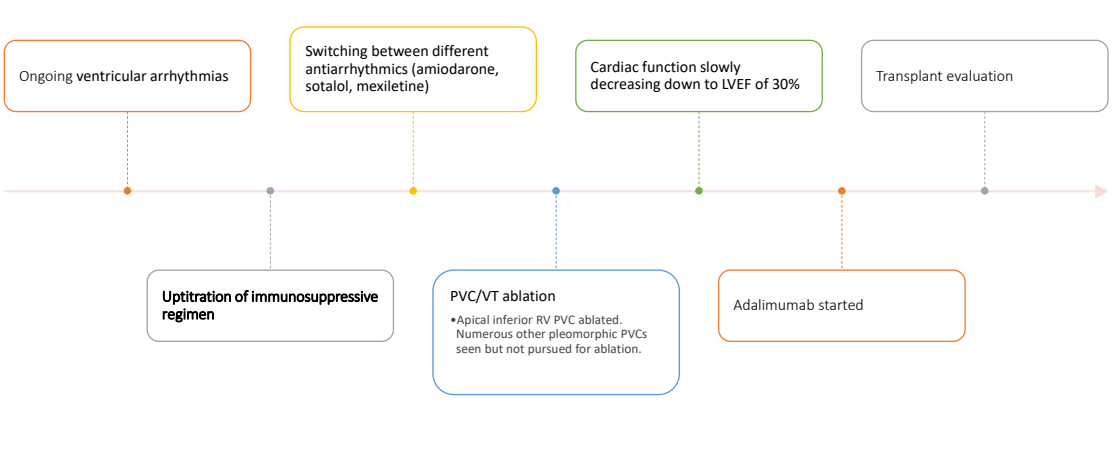

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HIV infection still considered relative contraindication to heart transplantation at most centers...

Perception of HIV-positive patients as high risk recipients to be avoided given scarce organ supply

Concern for immunosuppression-triggered progression of HIV to AIDS

Drug interactions which could worsen outcomes

- Impact of certain antiretrovirals on cytochrome P-450 metabolism

From kidney transplant data:

- HIV remained stable post-transplantation
- HIV positive recipients have higher rejection rates

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Doberne, J. W., Jawitz, O. K., Raman, V., Bryner, B. S., Schroder, J. N., & Milano, C. A. (2021). Heart transplantation survival outcomes of HIV positive and negative recipients. The Annals of thoracic surgery, 111(5), 1465-1471.

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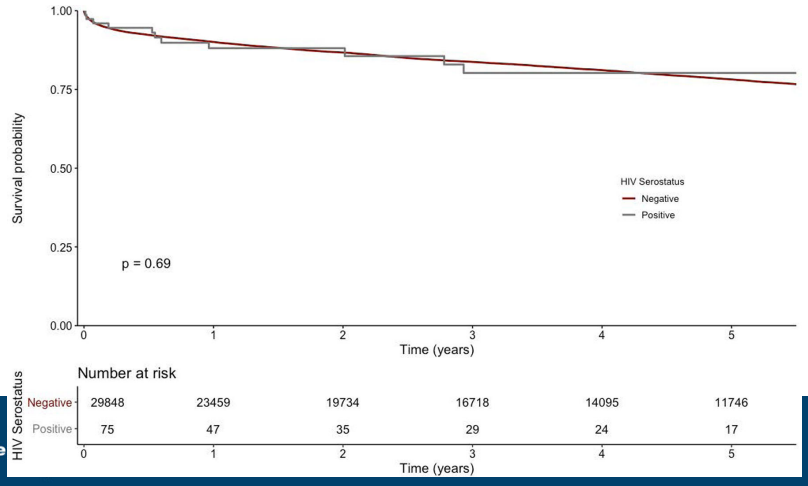
Heart Transplantation Survival Outcomes of HIV Positive and Negative Recipients

Check for updates

Julie W. Doberne, MD, PhD, Oliver K. Jawitz, MD, MHS, Vignesh Raman, MD, MHS, Benjamin S. Bryner, MD, MS, Jacob N. Schroder, MD, and Carmelo A. Milano, MD

Division of Cardiovascular and Thoracic Surgery, Department of Surgery, Duke University Medical Center, Durham, North Carolina



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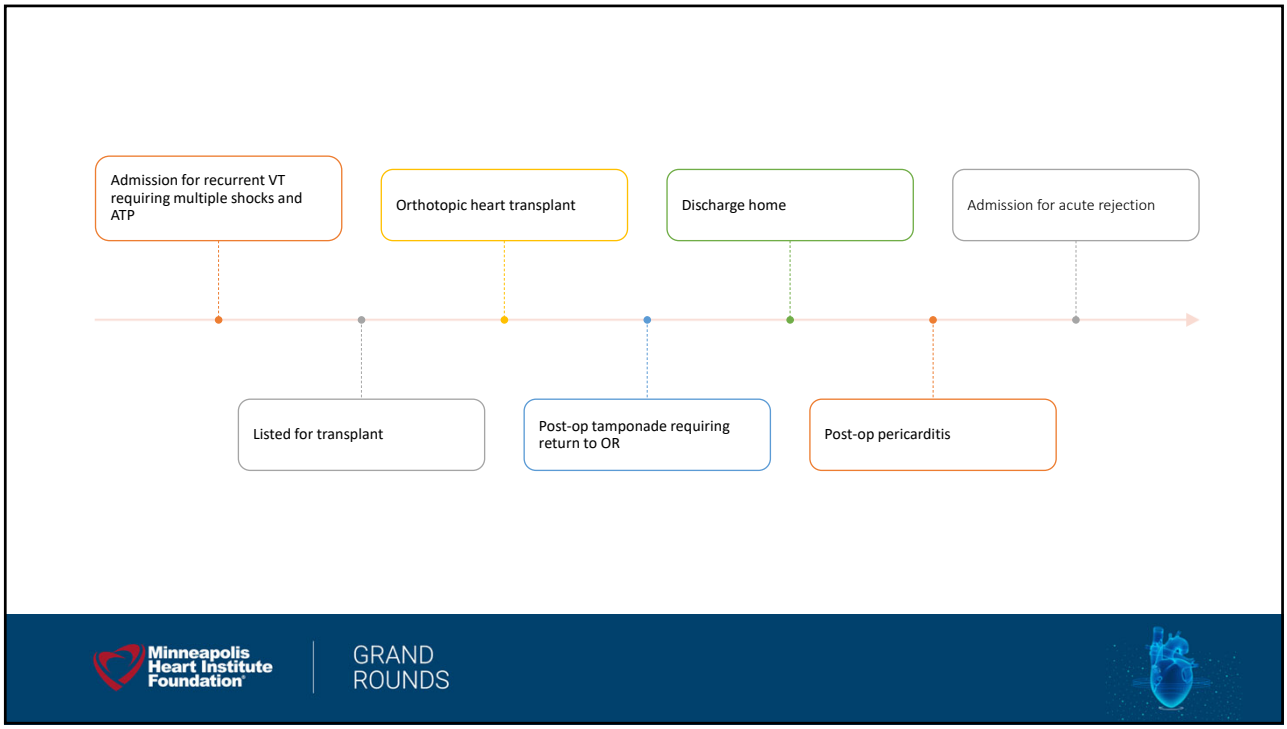
Admission for recurrent VT requiring multiple shocks and ATP
Orthotopic heart transplant
Discharge home
Admission for acute rejection
Listed for transplant
Post-op tamponade requiring return to OR
Post-op pericarditis
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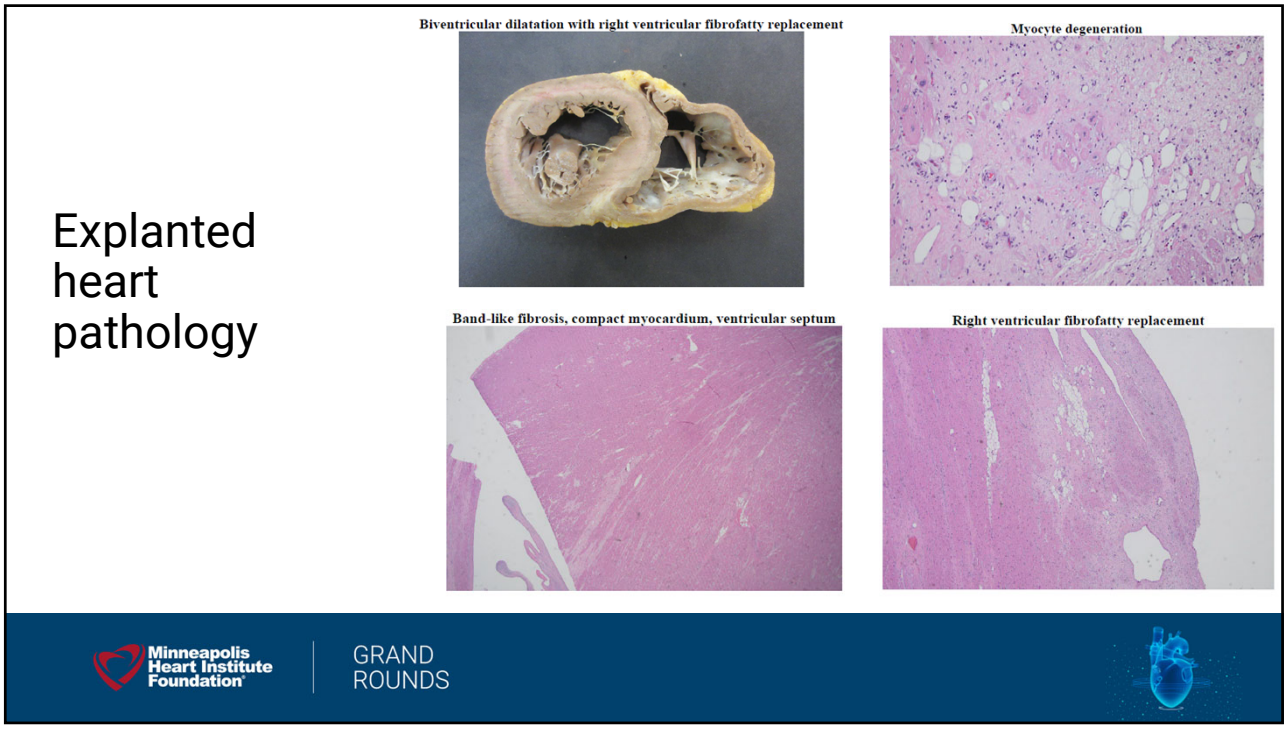
Explanted heart pathology
Biventricular dilatation with right ventricular fibrofatty replacement
Myocyte degeneration
Band-like fibrosis, compact myocardium, ventricular septum
Right ventricular fibrofatty replacement
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How sensitive and specific is 18-FDG PET for cardiac sarcoidosis?

Diagnostic performance of F-18 FDG PET for detection of cardiac sarcoidosis; A systematic review and meta-analysis

Seong-Jang Kim, MD, PhD,[a,b,c] Kyoungjune Pak, MD, PhD,[d] and Keunyoung Kim, MD[d]

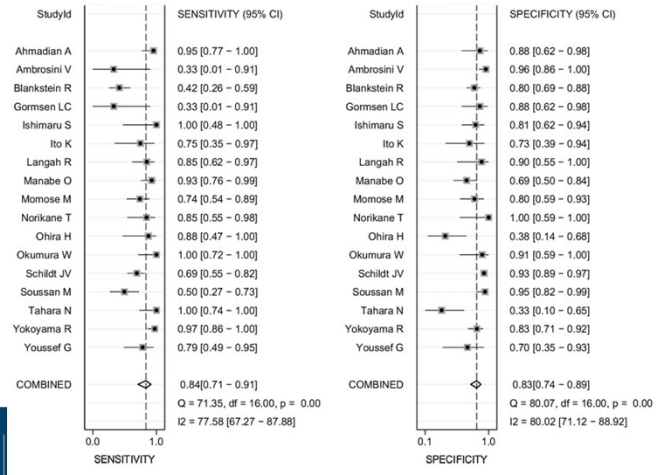

Figure 4. Forest plot of pooled sensitivity and specificity of F-18 FDG PET or PET/CT for the diagnosis of CS.

Kim, S. J., Pak, K., & Kim, K. (2020). Diagnostic performance of F-18 FDG PET for detection of cardiac sarcoidosis; A systematic review and meta-analysis. Journal of Nuclear Cardiology, 27, 2103-2115.

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How common is inflammation on 18-FDG PET in patients with ARVC?

Prevalence of ^{18}F–fluorodeoxyglucose positron emission tomography abnormalities in patients with arrhythmogenic right ventricular cardiomyopathy

Alexandros Protonotarios [1], Eleanor Wicks [2], Michael Ashworth [3], Edward Stephenson [4], Oliver Guttmann [5], Kostas Savvatis [4], Neha Sekhri [6], Saidi A Mohiddin [4], Petros Syrris [7], Leon Menezes [8], Perry Elliott [9]

Affiliations + expand

PMID: 30409737 DOI: 10.1016/j.ijcard.2018.10.083

Abstract

Background: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a heritable heart muscle disease that causes sudden cardiac death in the young. Inflammatory myocardial infiltrates have been described at autopsy and on biopsy, but there are few data on the presence of myocarditis in living patients with ARVC using non-invasive imaging techniques. FDG-PET is a validated technique for detecting myocardial inflammation in clinically suspected myocarditis. We aimed to determine the prevalence of myocardial inflammation in patients with ARVC using ^{18}F-fluorodeoxyglucose positron emission tomography (FDG-PET).

Methods and results: We performed a retrospective analysis of a single centre cohort of patients with ARVC referred for FDG-PET scans between 2012 and 2017 for investigation of symptoms or suspected device infection. Sixteen patients (12 male; age 42 ± 13 years) with a definite diagnosis of ARVC were identified. Seven had positive FDG-PET scans, two of whom had cardiac sarcoidosis on endomyocardial biopsy. Of the remaining five, two carried pathogenic desmoplakin mutations. FDG uptake was found in the left ventricular myocardium in all cases. One patient also had right ventricular uptake.

Conclusion: In this exploratory study, we show that some patients with ARVC have evidence for myocardial inflammation on FDG-PET, suggesting that myocarditis plays a role in disease pathogenesis.

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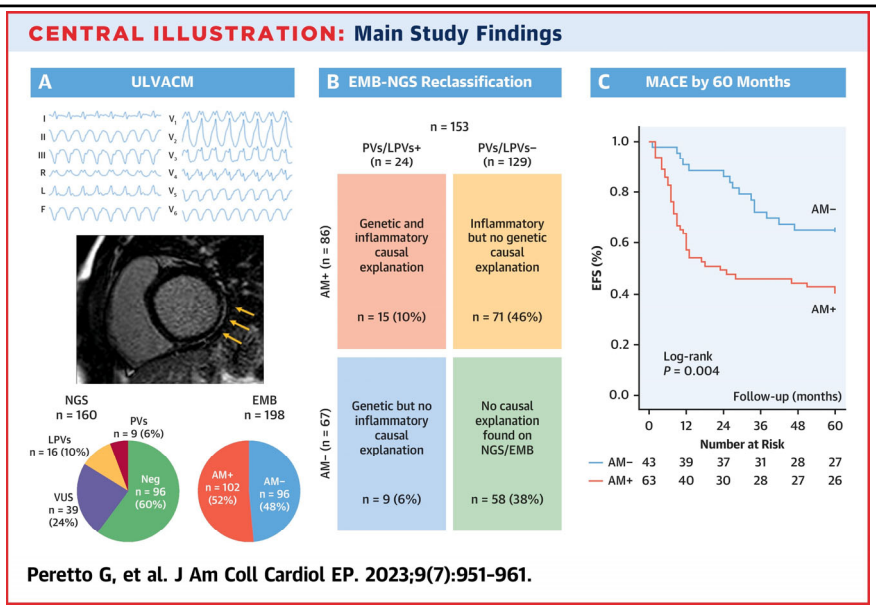
Protonotarios, A., Wicks, E., Ashworth, M., Stephenson, E., Guttmann, O., Savvatis, K., ... & Elliott, P. (2019). Prevalence of 18F-fluorodeoxyglucose positron emission tomography abnormalities in patients with arrhythmogenic right ventricular cardiomyopathy. *International journal of cardiology*, *284*, 99-104.

26

How common is inflammation in patients with ACM?

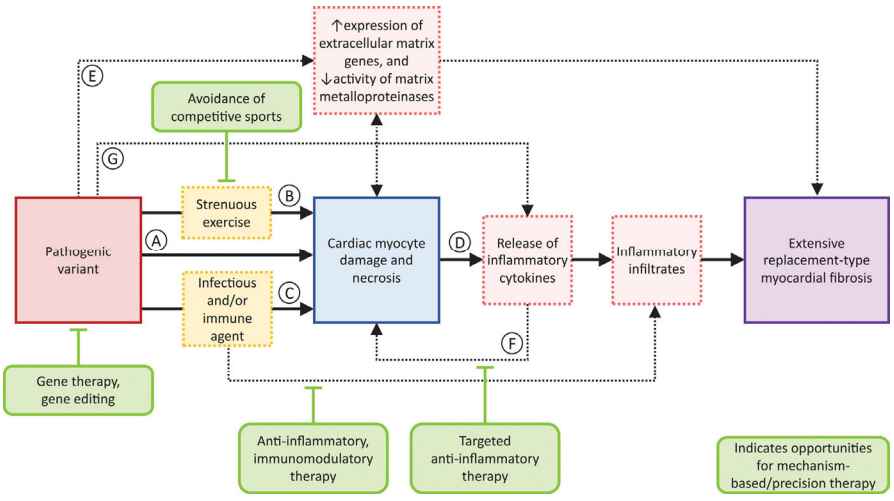


Peretto G, et al. J Am Coll Cardiol EP. 2023;9(7):951-961.

Peretto, G., Casella, M., Merlo, M., Benedetti, S., Rizzo, S., Cappelletto, C., ... & Cooper Jr, L. T. (2023). Inflammation on endomyocardial biopsy predicts risk of MACE in undefined left ventricular Arrhythmogenic cardiomyopathy. *Clinical Electrophysiology*, *9*(7_Part_1), 951-961.

27

Where does inflammation fall in the pathogenesis of ACM?



Asatryan, B., Asimaki, A., Landstrom, A. P., Khanji, M. Y., Odening, K. E., Cooper, L. T., ... & Chahal, C. A. A. (2021). Inflammation and immune response in arrhythmogenic cardiomyopathy: state-of-the-art review. *Circulation*, *144*(20), 1646-1655.

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- Thank you!

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Cardiovascular Disease in Kenya

Joseph D. Steffens, MD
Ron Johannsen, MD
12/18/2023

HennepinHealthcare

Allina Health
MINNEAPOLIS HEART INSTITUTE

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Disclosure

- **I have no financial disclosure or conflicts of interest with the presented material in this presentation.**
- **Any photos of patients have been used with permission and are for teaching purposes only**

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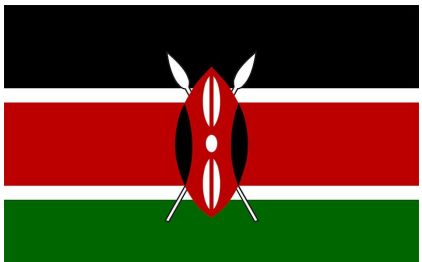
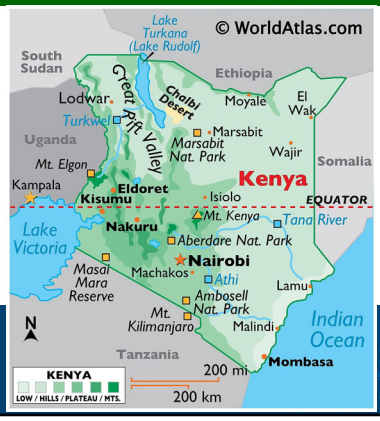
Kenya

- **Gained independence from Great Britain in 1963**
- **Population: 53.1 million people (2021)**
- **Official languages are Swahili and English**





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Epidemiology

- **Life expectancy 67.7 years**
- **Median age is 19.6 years**
- **Prevalence of rheumatic heart disease (RHD) is ~15% in East Africa (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10522432/)**
- **300,000-400,000 deaths per year worldwide (vast majority are women, ~85%, and common ages of death second and third decade of life and in pregnancy)**

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World Health Organization Data: Kenya

Top causes of death for females

Deaths per 100 000 population. Kenya, 2019

Neonatal conditions	42.5
Lower respiratory infections	39.3
HIV/AIDS	35.1
Stroke	29.8
Diarrhoeal diseases	25.9
Tuberculosis	24.6
Malaria	21.8
Maternal conditions	21.3
Cirrhosis of the liver	20.4
Ischaemic heart disease	20.4

Top causes of death for males

Deaths per 100 000 population. Kenya, 2019

Neonatal conditions	55.2
Tuberculosis	50.3
Lower respiratory infections	46.6
HIV/AIDS	44.4
Road injury	42.4
Diarrhoeal diseases	32.8
Cirrhosis of the liver	30.6
Stroke	30.6
Malaria	26.3
Ischaemic heart disease	25.2

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World Health Organization Data: United States

Top causes of death for females

Deaths per 100 000 population. United States of America, 2019

Ischaemic heart disease	135
Alzheimer disease and other dementias	116
Chronic obstructive pulmonary disease	62
Stroke	55
Trachea, bronchus, lung cancers	43
Breast cancer	29
Kidney diseases	25
Hypertensive heart disease	20
Lower respiratory infections	19
Colon and rectum cancers	18

Top causes of death for males

Deaths per 100 000 population. United States of America, 2019

Ischaemic heart disease	172
Alzheimer disease and other dementias	58
Chronic obstructive pulmonary disease	57
Trachea, bronchus, lung cancers	52
Stroke	41
Drug use disorders	29
Kidney diseases	27
Self-harm	25
Diabetes mellitus	22
Prostate cancer	22

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Eldoret and Kapsowar

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RHD Screening

2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease

Joselyn Rwebembera, James Marangou, Julius Chacha Mwita, Ana Olga Mocumbi, Cleonice Mota, Emmy Okello, Bruno Nascimento, Lene Thorup, Andrea Beaton, Joseph Kado, Alexander Kaethner, Raman Krishna Kumar, John Lawrenson, Eloi Marijon, Mariana Mirabel, Maria Carmo Pereira Nunes, Daniel Piñeiro, Fausto Pinto, Kate Ralston, Craig Sable, Amy Sanyahumbi, Anita Saxena, Karen Sliwa, Andrew Steer, Satupaitea Viali, Gavin Wheaton, Nigel Wilson, Liesl Zühlke & Bo Reményi — Show fewer authors

Criteria for pathological MR (requires all):
- Observed in two views
- Minimum MR jet length (1.5 cm for patients weighing <30 kg and 2.0 cm for patients weighing ≥30 kg) observed in one view
- Velocity >3.0 m/s[b]
- Pan-systolic jet[b]

Criteria for pathological AR (requires all):
- Observed in two views
- Velocity >3.0 m/s[b]
- Pan-diastolic jet[b]

RHD morphological criteria:
- MV anterior leaflet thickening and/or MV chordal thickening
- MV leaflet restriction and/or excessive anterior leaflet tip motion
- AV thickening, prolapse or restricted leaflet motion

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12 year old male presents with his father for RHD Screening

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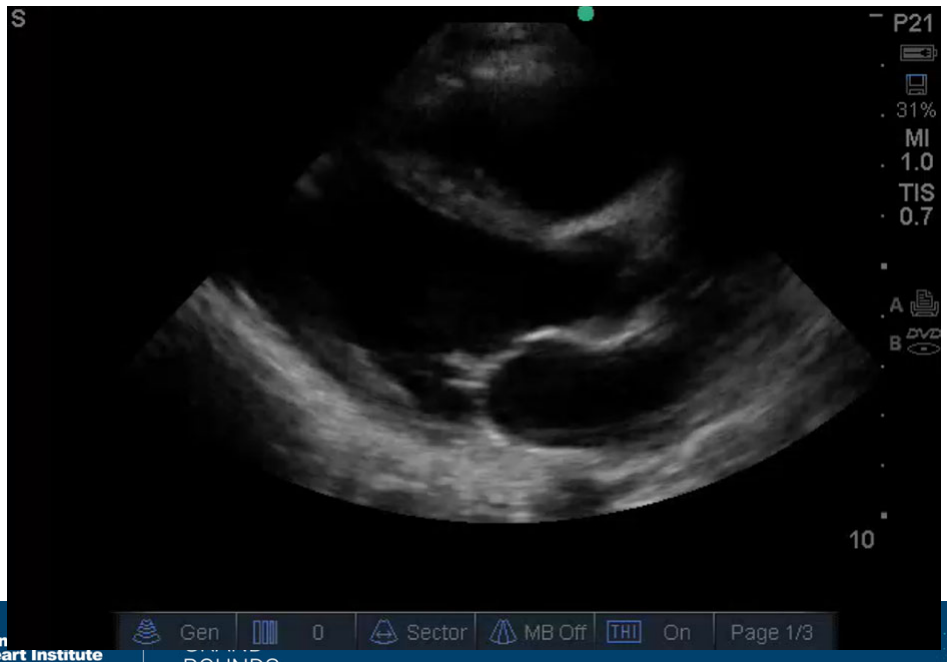

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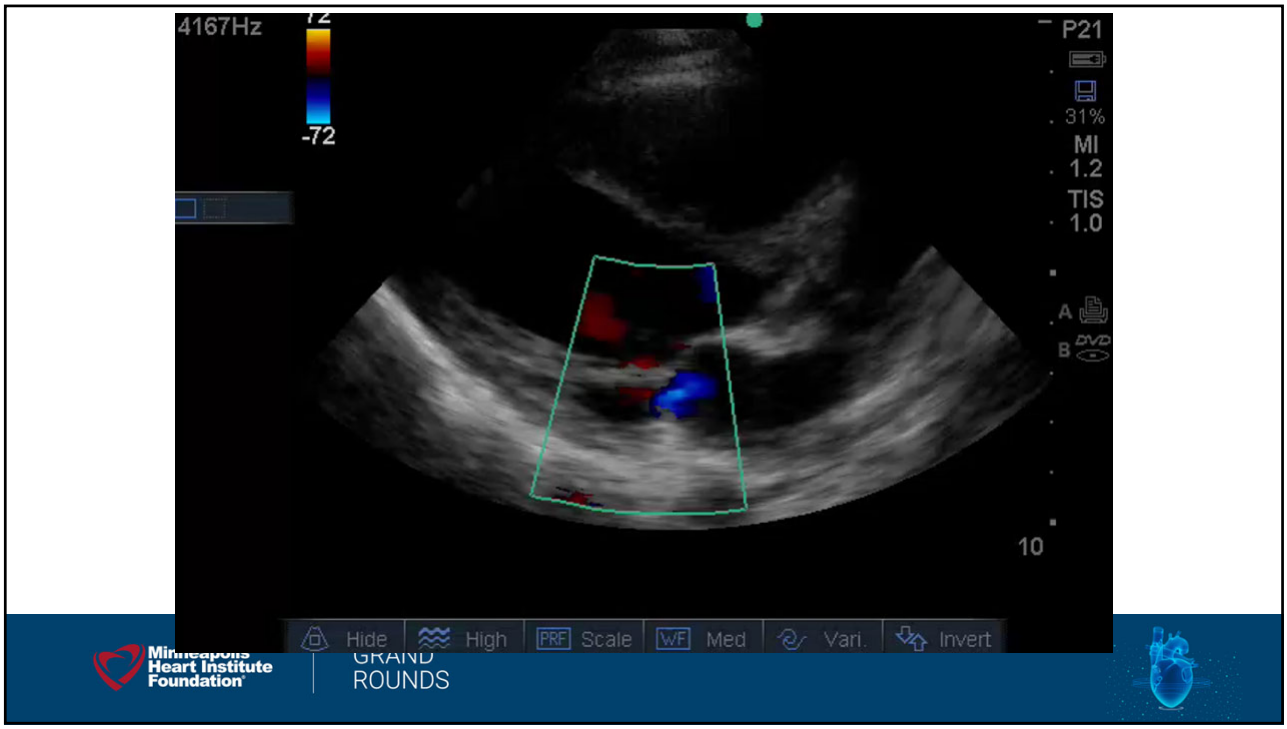
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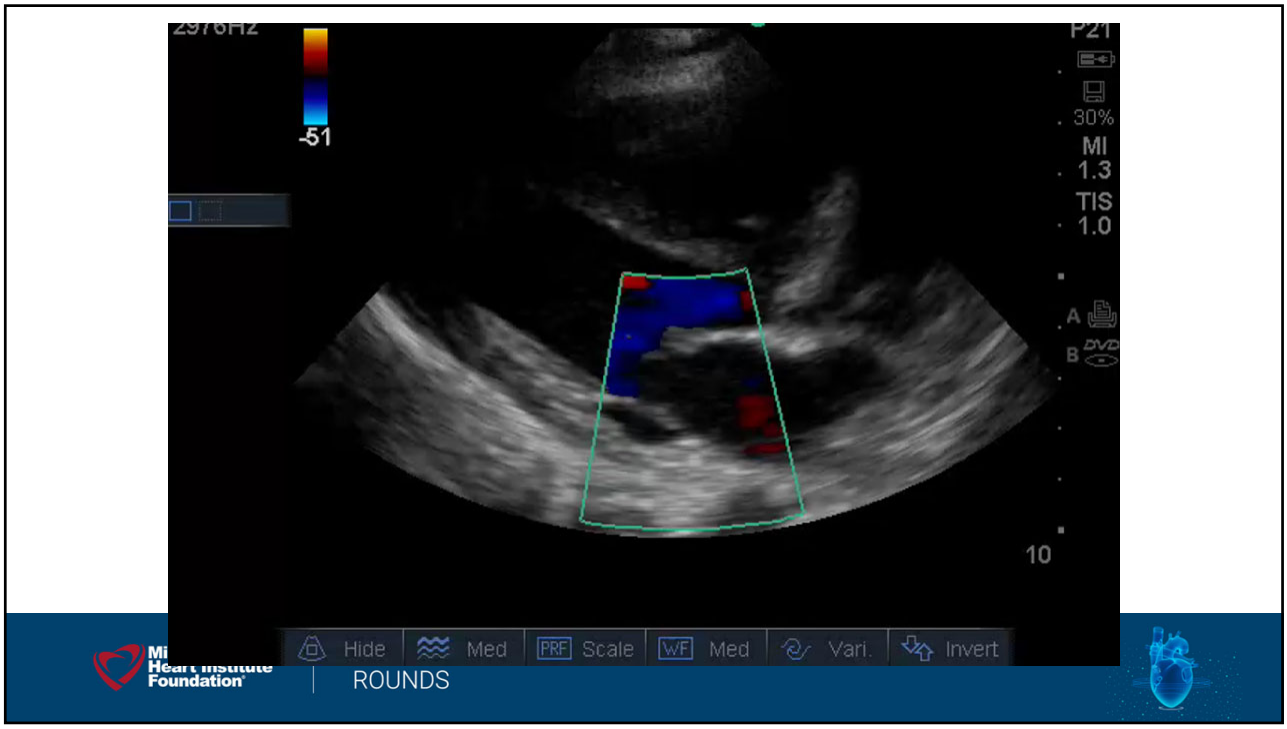
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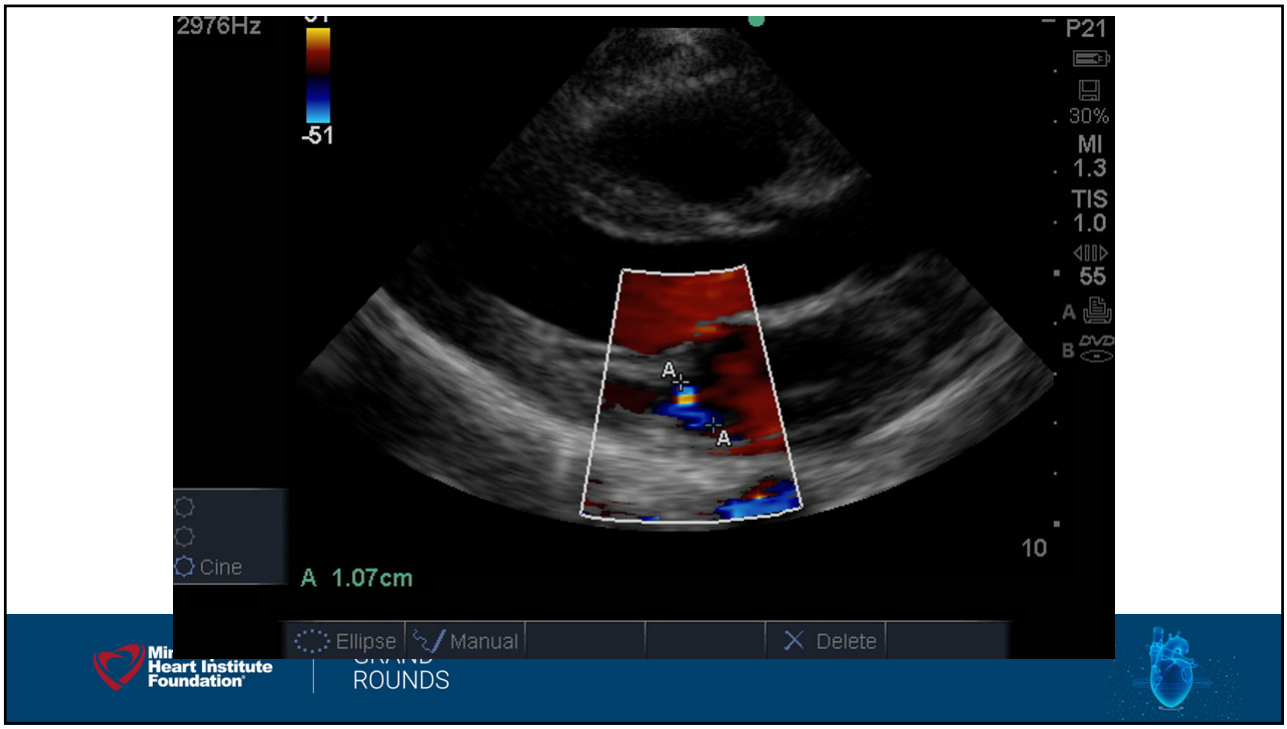

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Saving time saves lives! A time focused evaluation of a single-view echocardiographic screening protocol for subclinical rheumatic heart disease

Ronald A. Johannsen [a], Zachary P. Kaltenborn [b,*], Gautam R. Shroff [a,c]

[a] Division of Cardiology, Department of Internal Medicine, Hennepin County Medical Center, 716 S 7th St, Minneapolis, MN 55415, United States of America
[b] Division of General Internal Medicine and Hospital Pediatrics, Department of Internal Medicine and Pediatrics, University of Minnesota Medical School, Division Mailbox - MMC 741, 420 Delaware Street SE, Minneapolis, MN 55455, United States of America
[c] Department of Medicine, University of Minnesota Medical School, 420 Delaware Street SE, Minneapolis, MN 55455, United States of America

Background: Rheumatic heart disease affects 33 million people in low and middle income countries and is the leading cause of cardiovascular death among children and young adults. Evidence increasingly supports that simplified screening protocols can identify at risk children with good accuracy. One of the more proximal and pragmatic hurdles that has not been completely explored is the time required for executing the screening exam.
Methods: We conducted an observational study comparing three different echocardiographic strategies in four separate school-based screening programs in Kenya and Cameroon.
Results: In a sample of 911 children, we found that a single-view screening strategy can be obtained in an average time of 1.2 min/child, the two-view in an average of 2.1 min/child, and multi-view in an average of 5 min/child.
Conclusions: Our study demonstrates that there are significant differences in the time required to execute different screening protocols and is an essential consideration in the feasibility of large scale populations based rheumatic heart disease screening programs.

[15]. However, we used a modified version of the WHF criteria that did not require the application of continuous-wave Doppler, rather identifying visually pandiastolic AR or pathological pansystolic MR. [6,12] We additionally considered eccentric, visually identified pansystolic mitral regurgitation abnormal if the length was more than 1 cm. These

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RHD Secondary Prevention

Table 10.1. Recommended antibiotic regimens for secondary prophylaxis

ANTIBIOTIC	DOSE	ROUTE	FREQUENCY
First line			
Benzathine benzylpenicillin G (BPG)	1,200,000 units (≥20 kg) 600,000 units (<20 kg)†	Deep intramuscular injection	Every 28 days‡ Every 21 days for selected groups‡
Second line (if IM route is not possible or consistently declined)			
Phenoxymethylpenicillin (penicillin V)	250 mg	Oral	Twice a day
Following documented penicillin allergy			
Erythromycin	250 mg	Oral	Twice a day

† For children weighing less than 10 kg, a dose of 600,000 units is still generally recommended but seek paediatric advice & careful planning of the regimen of secondary prophylaxis.
‡ People on 28-day regimens can be recalled from 21 days to help ensure that injections are given by day 28.
§ BPG given every 21 days may be considered for a) patients who have breakthrough ARF despite complete adherence to a 28-day regimen, or b) are at high risk of adverse consequences if ARF occurs (have severe RHD or a history of heart valve surgery).

Mild RHD [c]	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF, or until age 21 years (whichever is longer) If NO documented history of ARF and aged <35 years: [d] Minimum of 5 years following diagnosis of RHD or until age 21 years (whichever is longer)
Moderate RHD [c, e]	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 35 years (whichever is longer) If no documented history of ARF and aged <35 years: [d] Minimum of 5 years following diagnosis of RHD or until age 35 years (whichever is longer)
Severe RHD [e, f]	If documented history of ARF: Minimum of 10 years after the most recent episode of ARF or until age 40 years (whichever is longer) If no documented history of ARF: [g] Minimum of 5 years following diagnosis of RHD or until age 40 years (whichever is longer)

ews/clinical-update-secondary-prevention-arf

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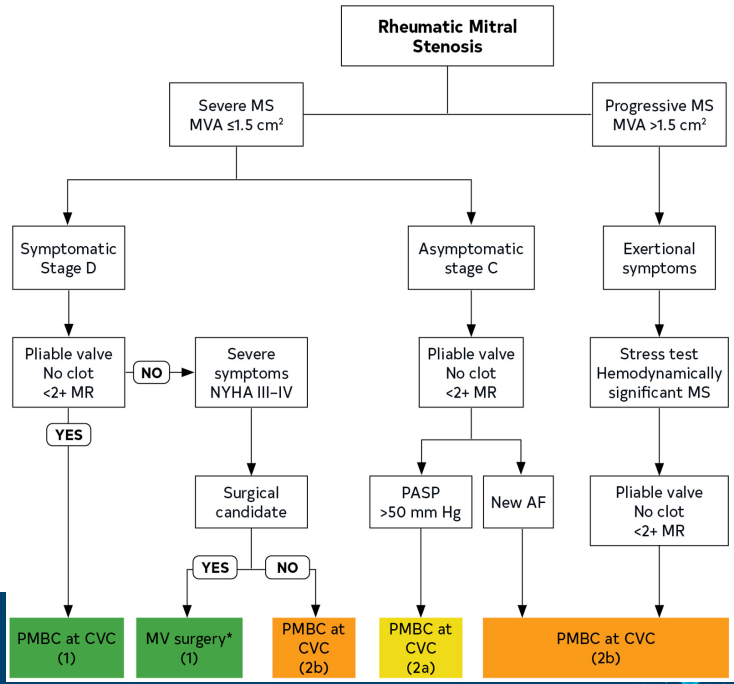
2020 ACC/AHA Guidelines



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Wilkins Score

Grade	Mobility	Thickening	Calcification	Subvalvular thickening
1	highly movile valve with only leaflet tips restricted	Leaflets near normal in thickness (4–5 mm)	A single area of increased echo brightness	Minimal thickening just below the mitral leaflets
2	Leaflet mid and base portions have normal mobility	Mid leaflets normal, considerable thickening of margins (5–8 mm)	Scattered areas of brightness confined to leaflet margins	Thickening of chordal structures extending to one third of the chordal length
3	Valve continues to move forward in diastole, mainly from the base	Thickening extending through the entire leaflet (5–8 mm)	Brightness extending into the mid portions of the leaflets	Thickening extended to distal third of the chords
4	No or minimal forward movement of the leaflets in diastole	Considerable thickening of all leaflet tissue (>8–10 mm)	Extensive brightness throughout much of the leaflet tissue	Extensive thickening and shortening of all chordal structures extending down to the papillary muscles

The total score is the sum of the four items and ranges between 4 and 16

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Case 1

- **12 year old male with history of rheumatic heart disease**
- **Worsening mitral stenosis with shortness of breath and weight loss**
- **Medically managed with metoprolol, furosemide, and spironolactone**

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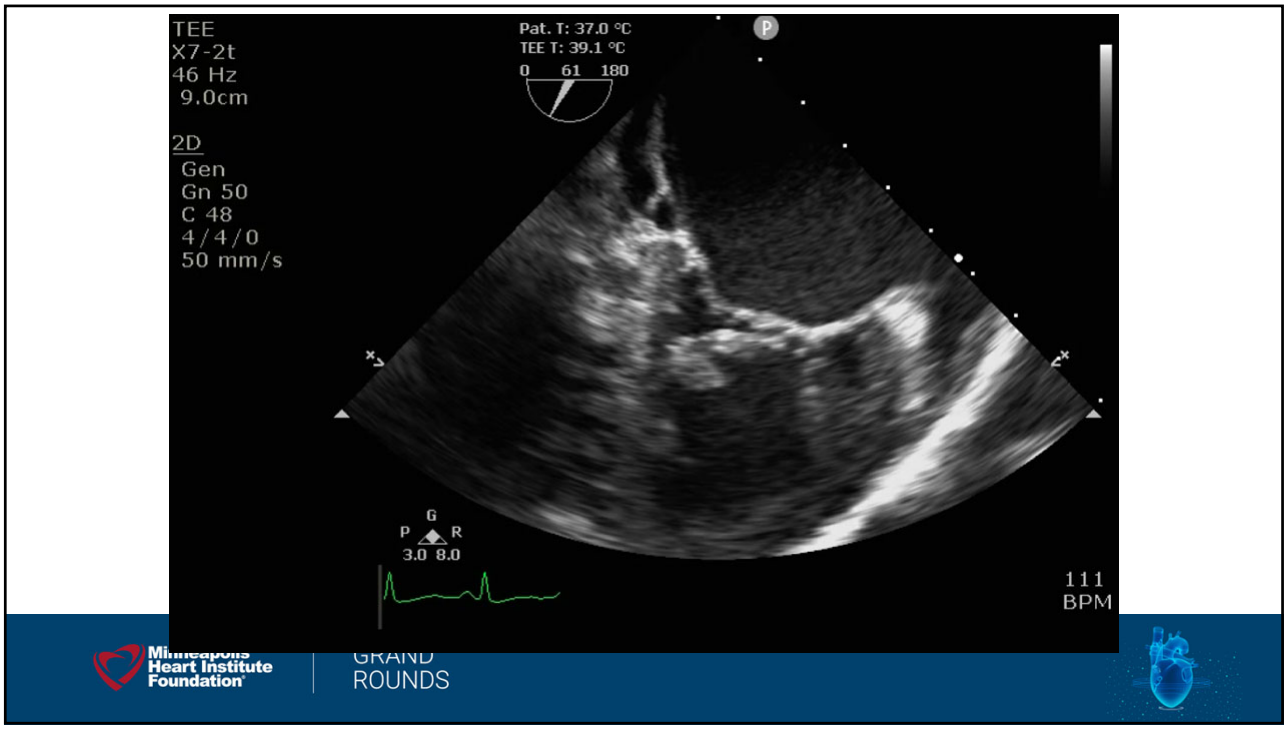
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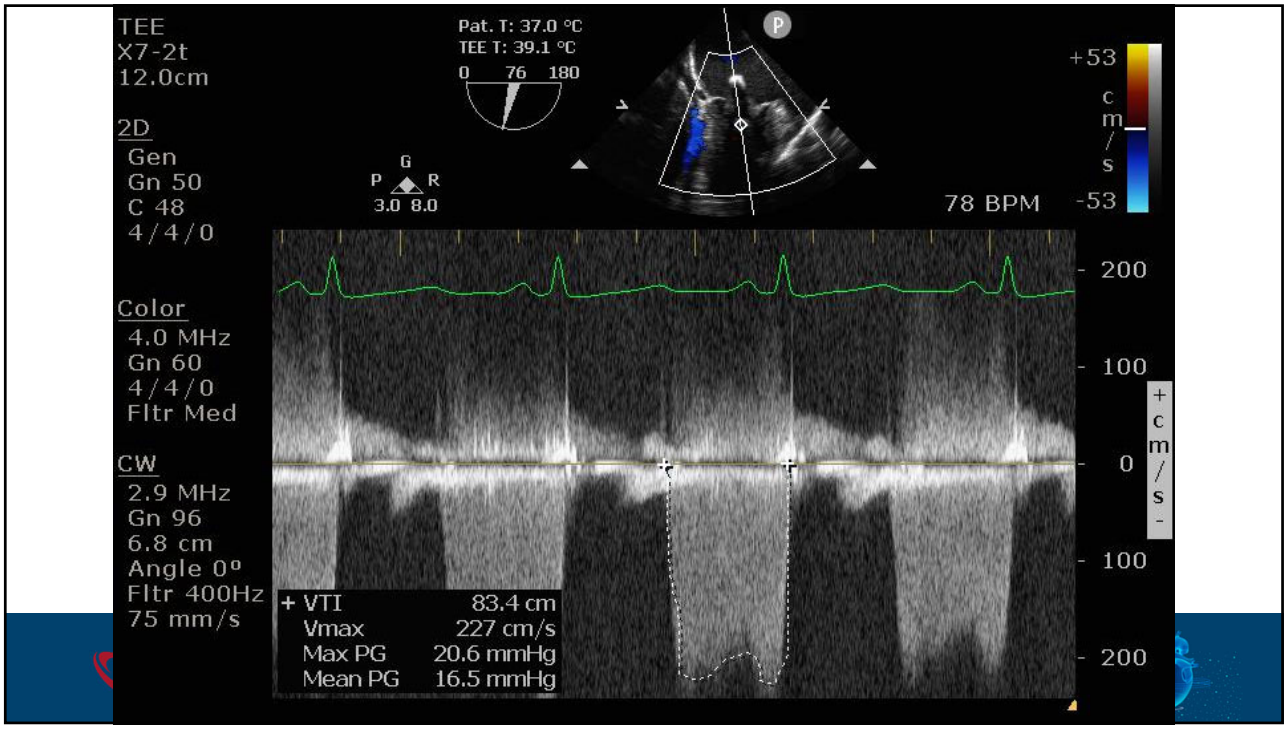
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CW
2.9 MHz
Gn 96
6.8 cm
Angle 0°
Fltr 400Hz
75 mm/s
Pat. T: 37.0 °C
TEE T: 39.1 °C
0 76 180
G
P R
3.0 8.0
78 BPM
+53
cm/s
-53
200
100
0
100
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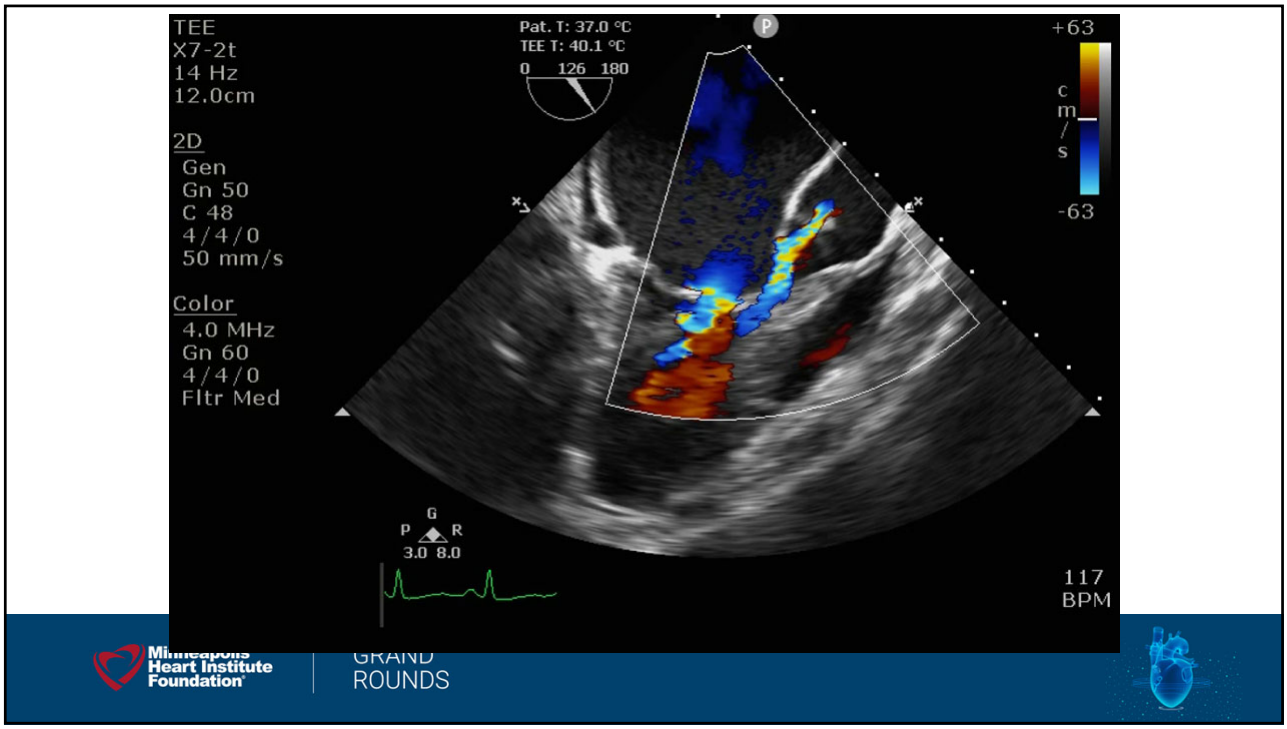
+ cm/s -
+ VTI 83.4 cm
Vmax 227 cm/s
Max PG 20.6 mmHg
Mean PG 16.5 mmHg

50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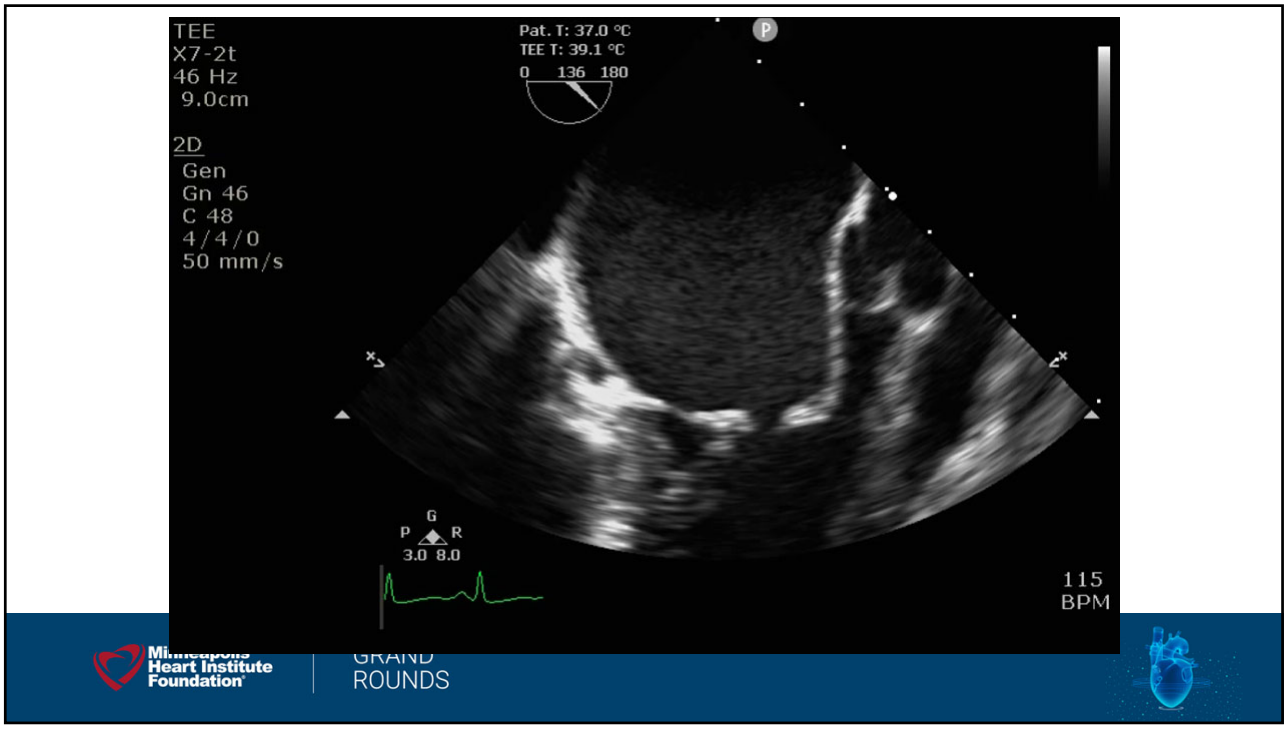
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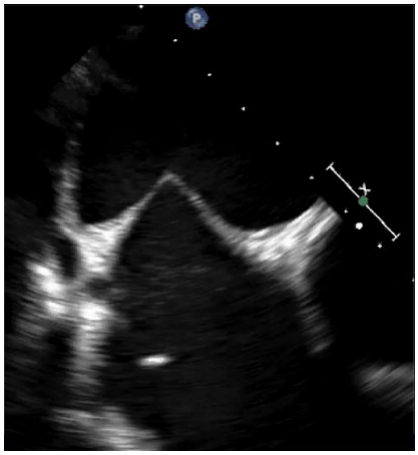
52

Transseptal Puncture



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Courtesy, Dr. Jason Rogers, UC Davis

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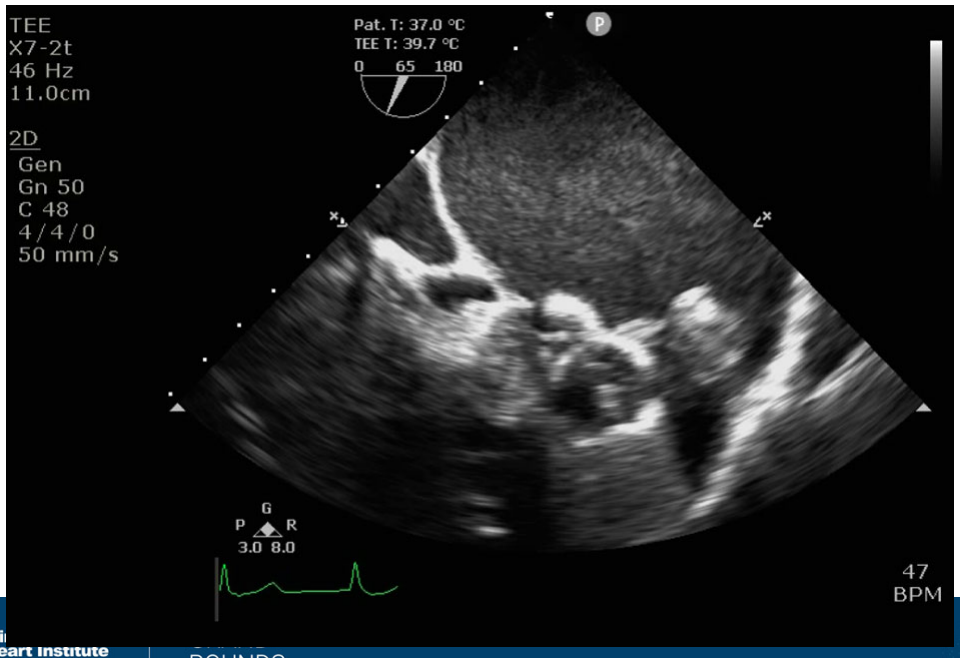

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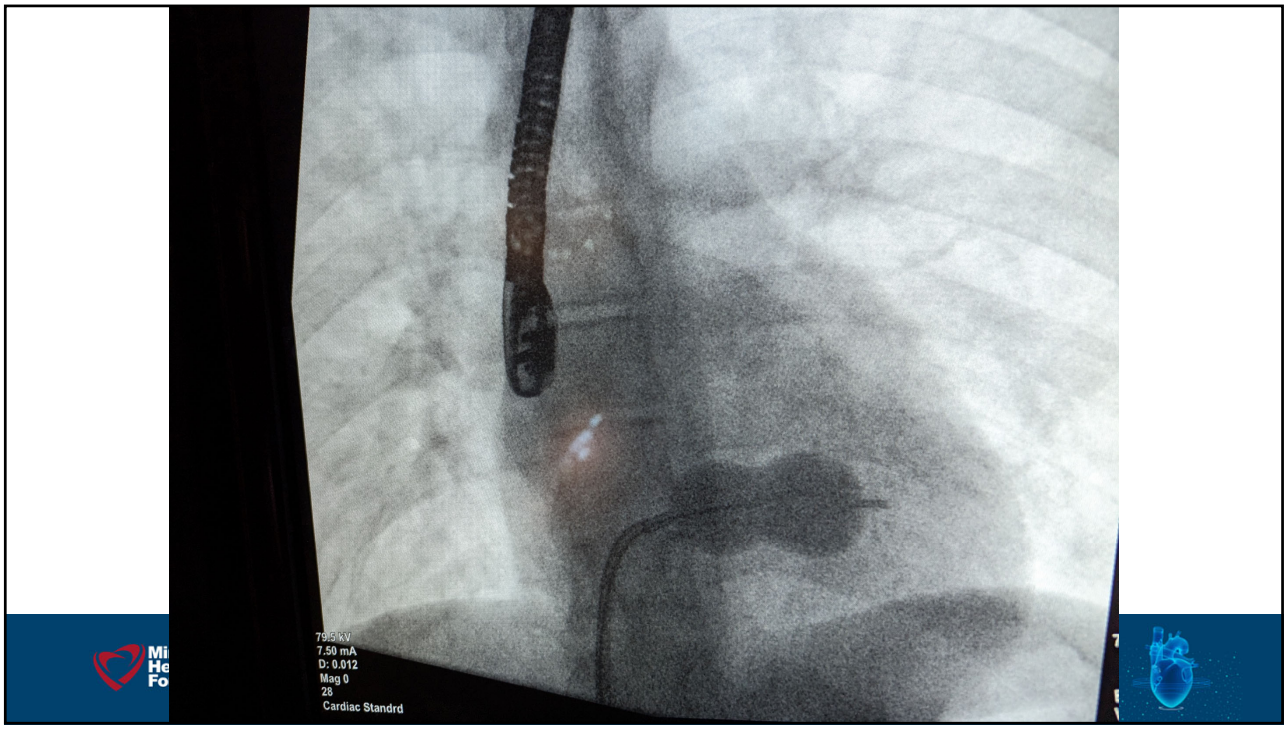

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Post mitral balloon valvuloplasty

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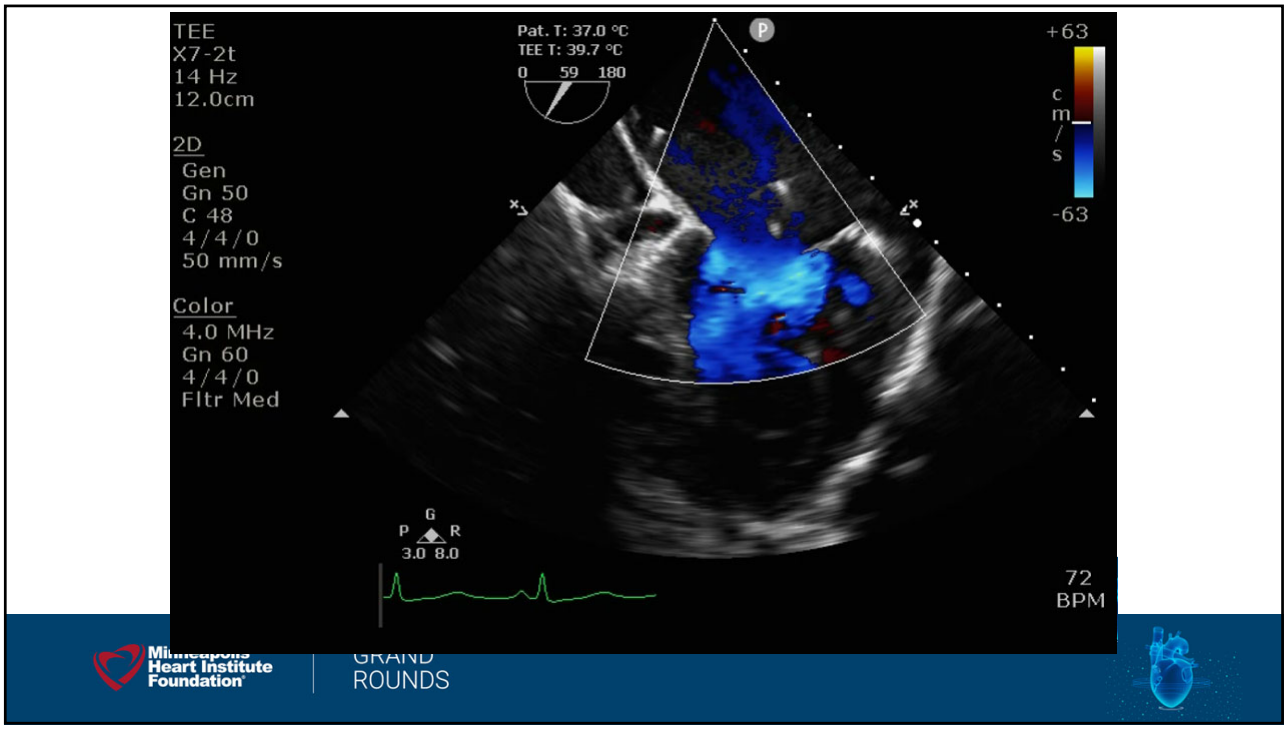
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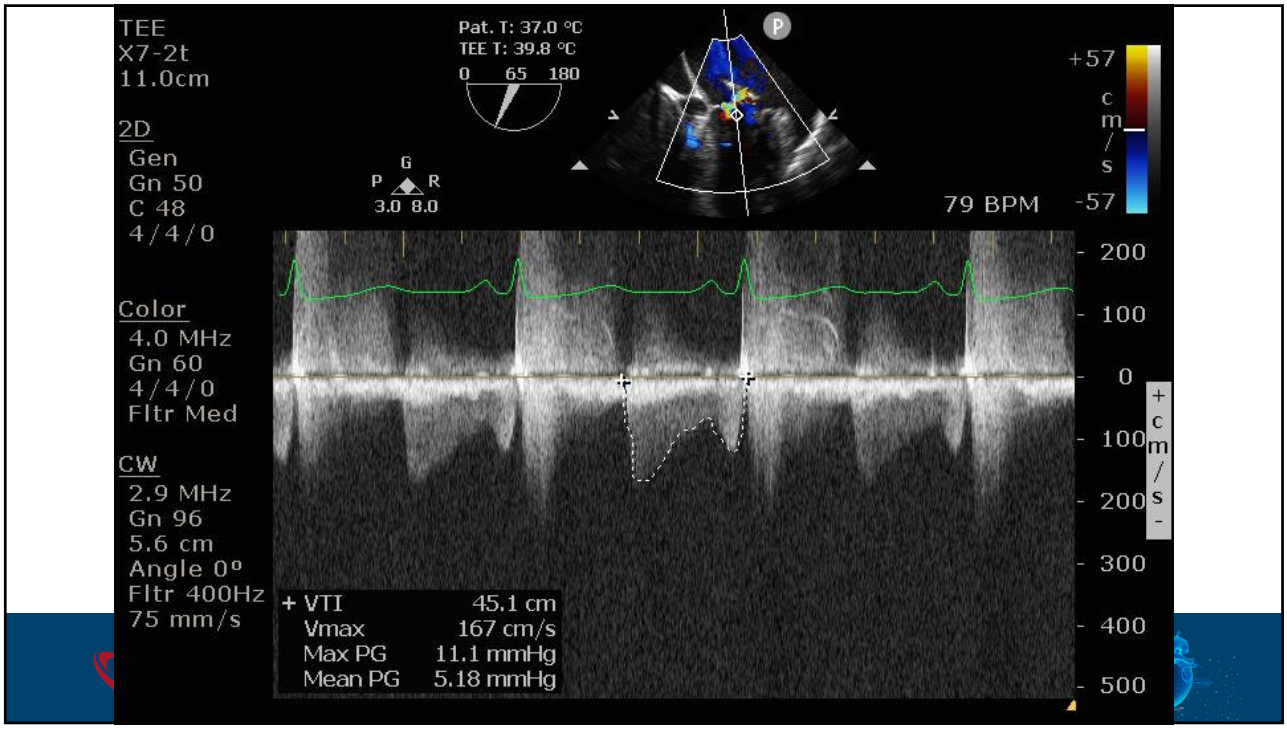
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100
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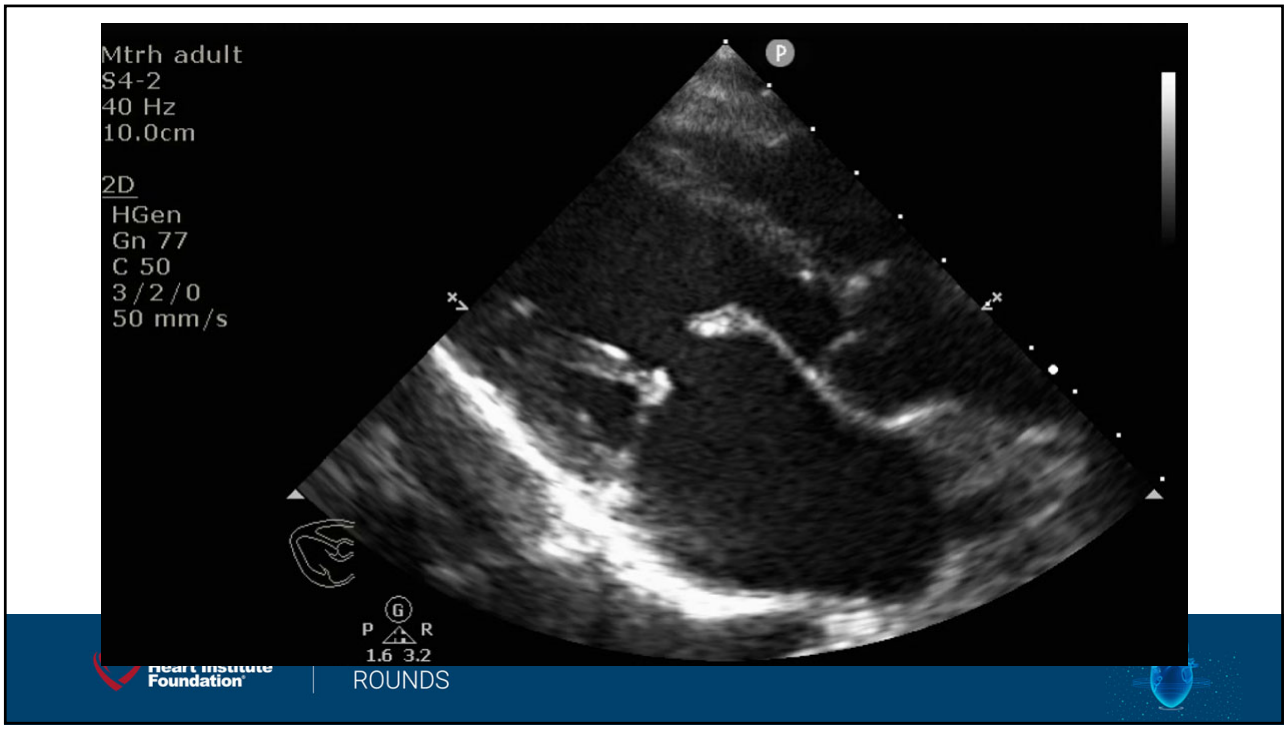
+ VTI 45.1 cm
Vmax 167 cm/s
Max PG 11.1 mmHg
Mean PG 5.18 mmHg

58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ROUNDS

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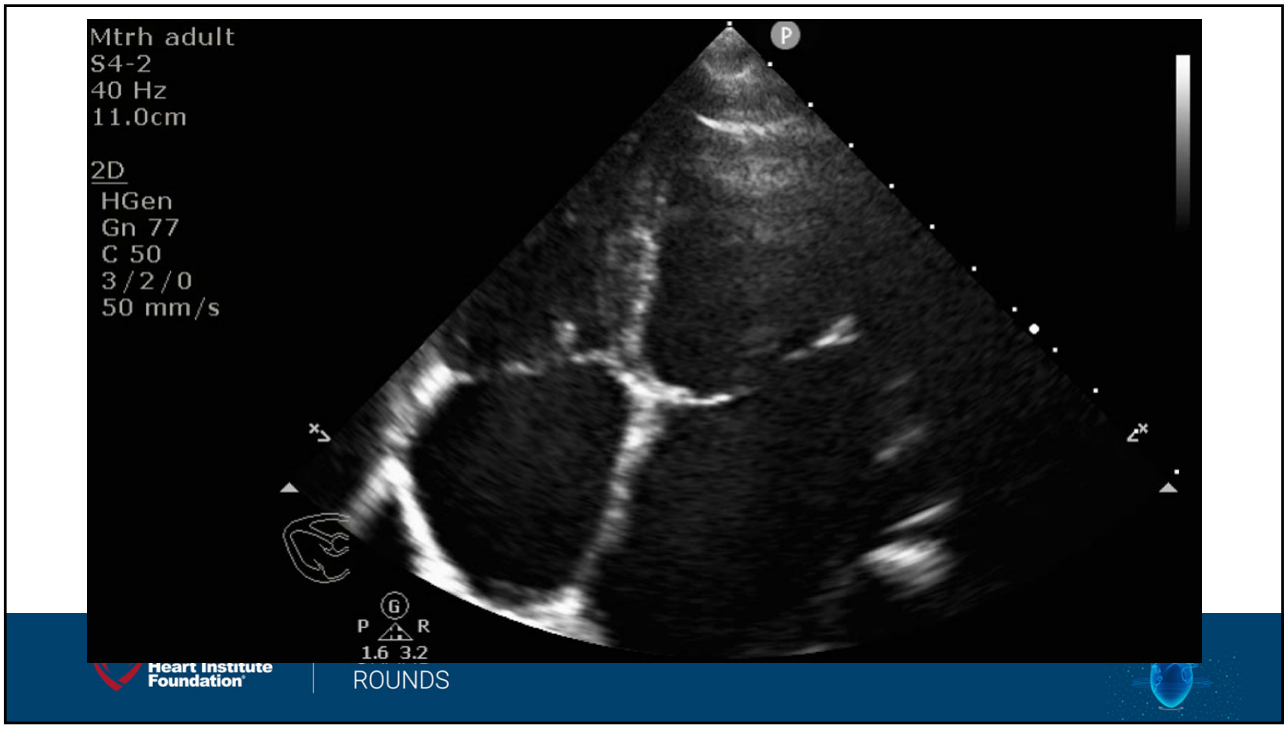
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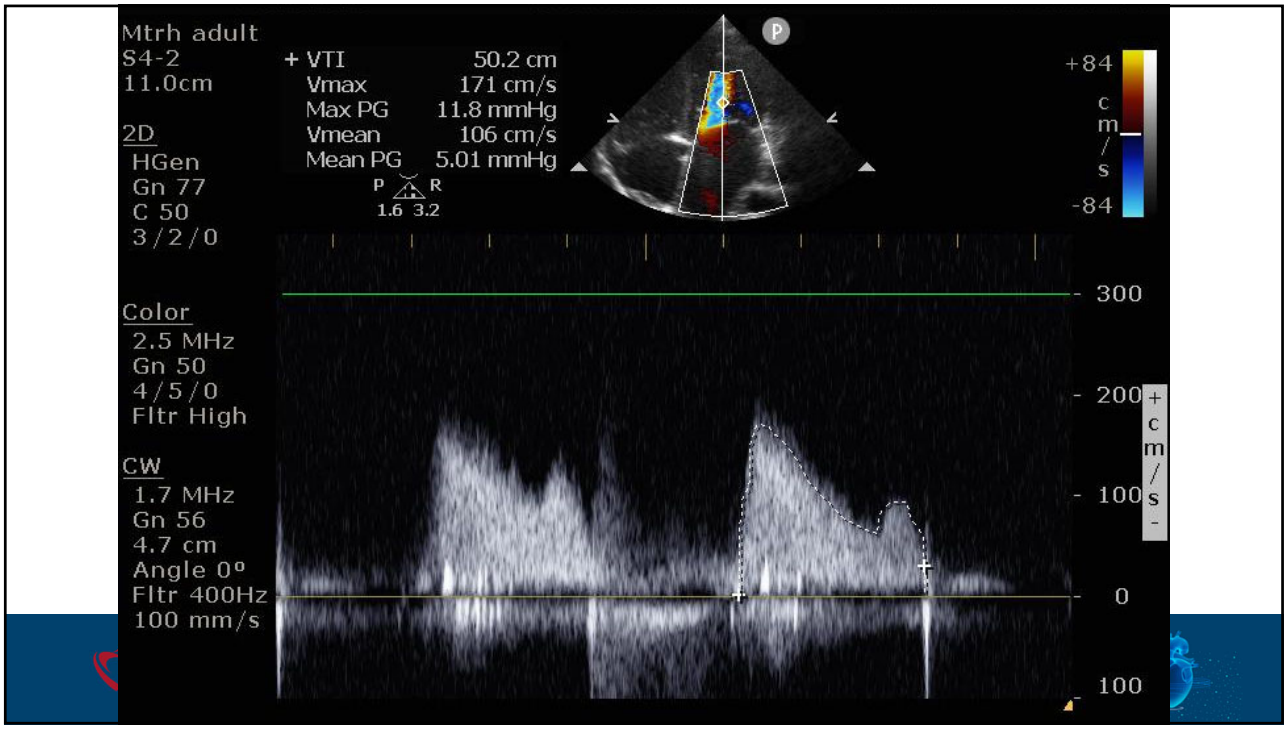

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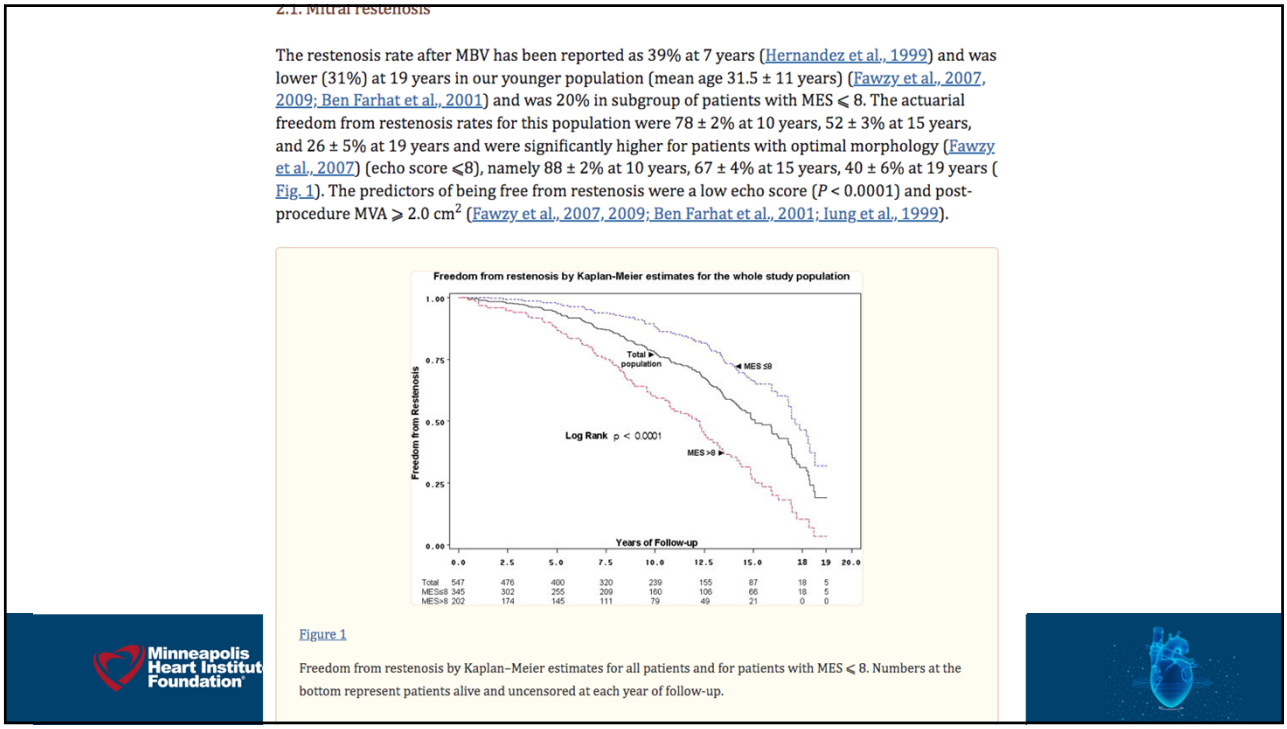

2.1. Mitral restenosis

The restenosis rate after MBV has been reported as 39% at 7 years (Hernandez et al., 1999) and was lower (31%) at 19 years in our younger population (mean age 31.5 ± 11 years) (Fawzy et al., 2007, 2009; Ben Farhat et al., 2001) and was 20% in subgroup of patients with MES ⩽ 8. The actuarial freedom from restenosis rates for this population were 78 ± 2% at 10 years, 52 ± 3% at 15 years, and 26 ± 5% at 19 years and were significantly higher for patients with optimal morphology (Fawzy et al., 2007) (echo score ⩽8), namely 88 ± 2% at 10 years, 67 ± 4% at 15 years, 40 ± 6% at 19 years (Fig. 1). The predictors of being free from restenosis were a low echo score ($P < 0.0001$) and post-procedure MVA ⩾ 2.0 cm^2 (Fawzy et al., 2007, 2009; Ben Farhat et al., 2001; Iung et al., 1999).

Figure 1

Freedom from restenosis by Kaplan–Meier estimates for all patients and for patients with MES ⩽ 8. Numbers at the bottom represent patients alive and uncensored at each year of follow-up.

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Case 2

- **25 year old woman who was in the hospital to schedule an appointment**
- **Has been feeling short of breath and unable to walk on any incline**
- **Her father recently passed away from RHD**

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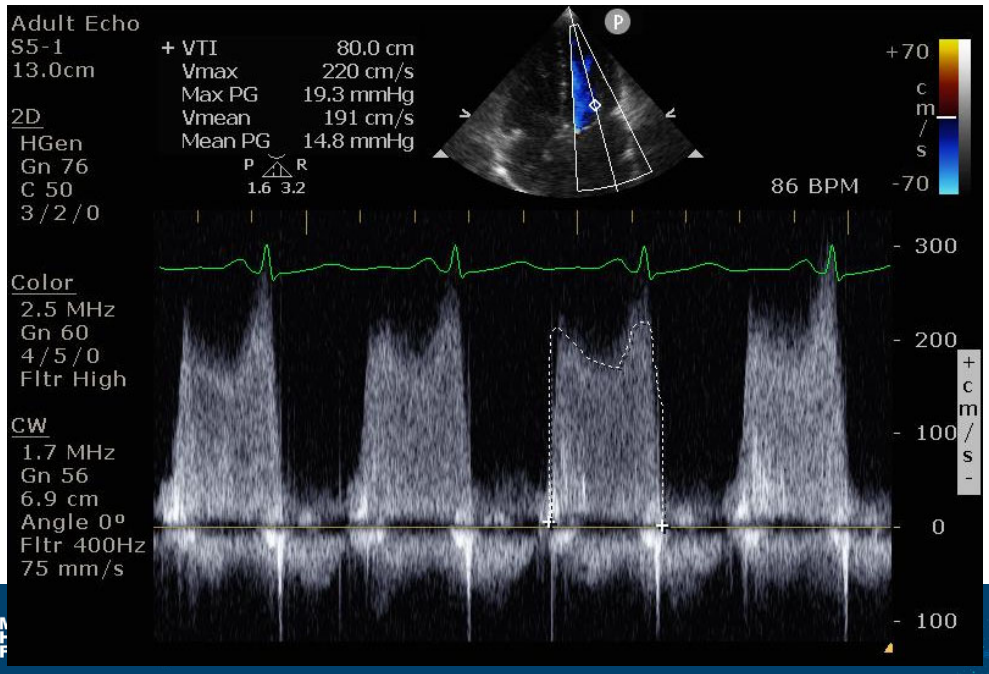

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Course

- **She was admitted overnight, diuresed, and beta blockade was initiated**
- **She underwent PBMV the following morning**
- **Mean gradient improved to 4 mmHg with no mitral regurgitation**

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Thank you!

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Thank you!

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