#ASEchoJC Twitter Chat
Tuesday, January 10, 2023 – 8 PM ET

- **Recommendations for the Use of Echocardiography in the Evaluation of Rheumatic Heart Disease: A Report from the American Society of Echocardiography** (JASE, January 2023)

Authors and JASE Editor-in-Chief

- Stephen H. Little, MD, FASE (@SLittleMD)
- Shantanu P. Sengupta, MD, DNB, FASE (@senguptasp)
- Patricia A. Pellikka, MD, FACC, FAHA, FASE (@pattypellikka)

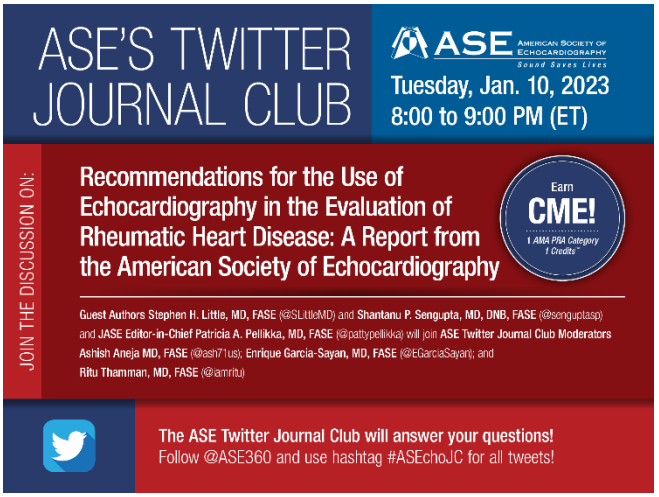
Moderators:

- Ashish Aneja MD, FACC, FASE, FSCMR (@ash71us)
- Enrique Garcia-Sayan, MD, FACC, FASE (@EGarciaSayan)
- Ritu Thamman, MD, FASE, FACC (@iamritu)

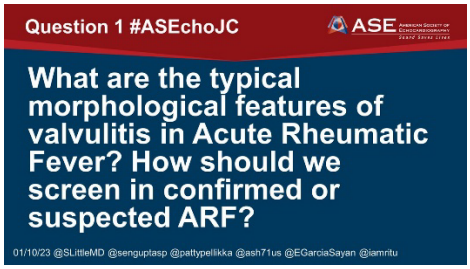
Introduction and Welcome: 📣Welcome to tonight's #ASEchoJC! 💥We'll discuss new recommendations for #EchoFirst in Rheumatic Heart Disease w/ authors @SLittleMD @senguptasp, @JournalASEcho EiC @pattypellikka & co-moderators @ash71us @iamritu

💥 10 ❓for discussion to follow

👉Use #ASEchoJC in all Tweets



Q1:



A1 Notable Responses:

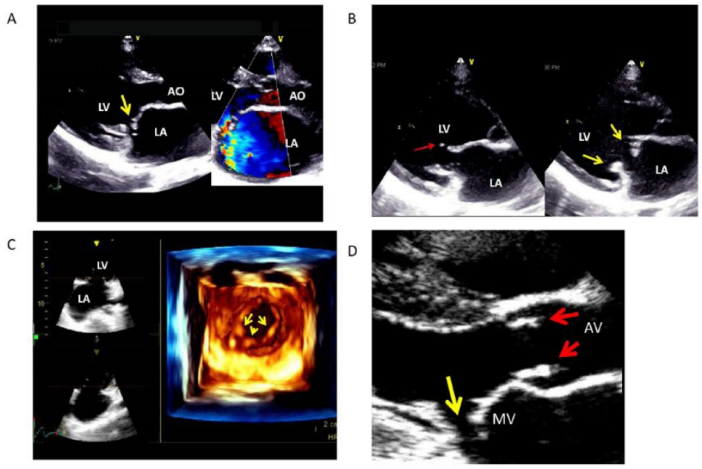
@EGarciaSayan: What are the typical morphological features of valvulitis in Acute Rheumatic Fever (ARF)? How should we screen in confirmed or suspected ARF?

@EGarciaSayan: 🔺Immune-mediated carditis in 50-70% cases of ARF

🔺#EchoFirst Dx valvulitis is major Jones criteria for carditis in ARF

🔺Look for mitral or aortic regurgitation

🔺Valve thickening, MV>AV, may be absent in early phase 👉MV thickness >3.5 mm in adults & 3 mm in children



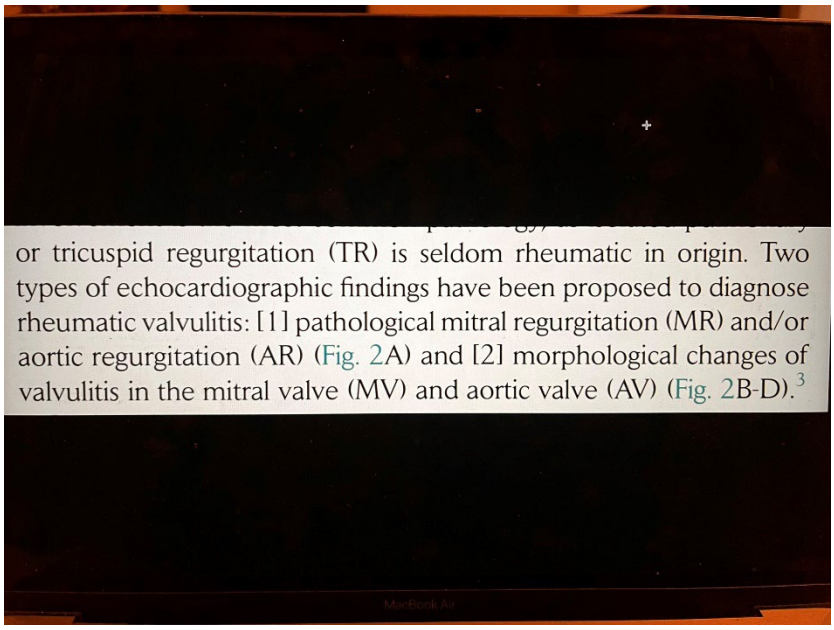
@SLittleMD: You guys are so fast with your detailed, and very informative tweets! I really need to up my game.

@iamritu: In ARF if pt has severe AR (without significant MR) with signs of inflammation like high ESR & prolonged PR(vascular AR) suspect Takayasu h/t @DrRajeshG1

@SLittleMD: Leaflet thickening in the right clinical context (after confirmed or suspected GAS infection - tonsilopharyngitis).

@EGarciaSayan: features of valvulitis in acute rheumatic fever. Clinical context + valve regurgitation or significant leaflet thickening indicating carditis

@senguptasp: Valvular thickening and restricted mobility seen in MV and AV are the main morphological features



@iamritu: immune-mediated carditis major criterion for ARF Dx

LV dysfunction 🚫common in ARF though carditis is pancarditis.If pt has HF usually from acute severe MR or AR(valvulitis) If pt w LV dysf &🚫 much MR or AR but fever/joint pain more likely🥚 myocarditis 📷 @thiagoecho

@EGarciaSayan: What are the typical morphological features of valvulitis in Acute Rheumatic Fever? How should we screen in confirmed or suspected ARF?

@SLittleMD: Wow. What a beautiful image!

@thiagoecho: Thank you !

@DrRajeshG1: Excellent teaching points.

Isolated pericarditis with fever and arthritis without valvular lesions = likely viral. Pericarditis in ARF occur in severe form of carditis.

@DocStrom: The MV leaflet tips thicken/fuse before the base so the result is a hockey stick like deformity of the valve.

@DocStrom: 2/ as the disease progresses, the whole leaflet can be thickened with commissural fusion and the opening snap can be lost.

@mariovar55: Dilated mitral.annulus

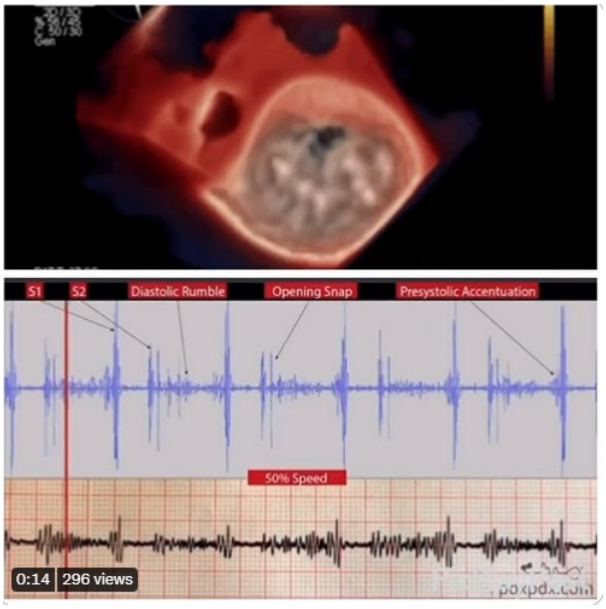
Many times absent mitral coaptation

Aortic reg

@iamritu: valvular thickening at free edge of leaflet with or without restricted leaflet mobility is most common morphology of Mitral RHD

Screen with 🩺 or pocus

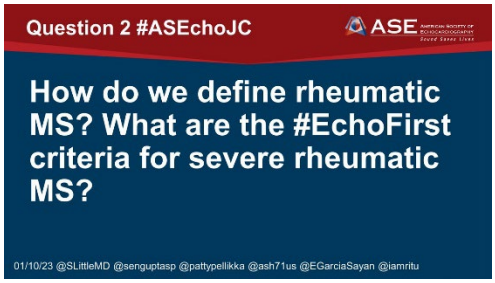
http://bit.ly/3W1cPde



@pattypellikka: More than trace mitral regurgitation in a child is a tip to pathology

@Pardhu6627: Beading of mitral valve is typical of rheumatic valvulitis

Question 2:



A2 Notable Responses:

@EGarciaSayan: 🔺RHD is most common global etiology of MS

🔺Defined as mean gradient >4 mmHg + typical morphological characteristics

🔺Thickening (distal>basal), commissural fusion, diastolic doming🖌️, chordal involvement

🔺Remember MVA<1.5 cm2 IS severe MS (not moderate) since 2014

@SLittleMD: The MVA cut off of < 1.5cm2 is the new one to remember for severe MS. Aligned with the ACC/AHA VHD guideline. Before was <1.0cm2 which some of us older folks still remember.

@SLittleMD: Always look for more than one parameter to confirm severe MS. Ideally will find MVA <1.5cm2; long PHT (>150ms); and mean diastolic gradient >10mmHg. But the diastolic parameters are flow dependent. The MVA is essentially fixed for moderate or severe rheumatic MS.

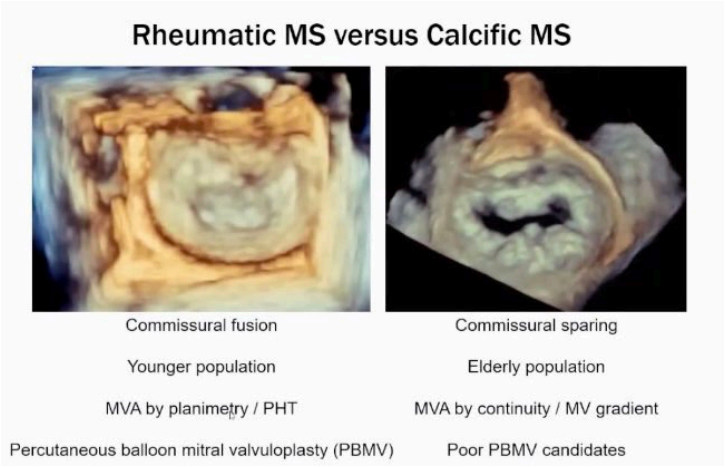
@EGarciaSayan: @ASE360 president @SLittleMD highlights the importance of multi-parametric approach for MS assessment. Careful on relying on one single parameter (especially mean gradient). Must know caveats of planimetry and PHT measurement.

@pattypellikka: See this letter for differentiation of rheumatic from calcific MS

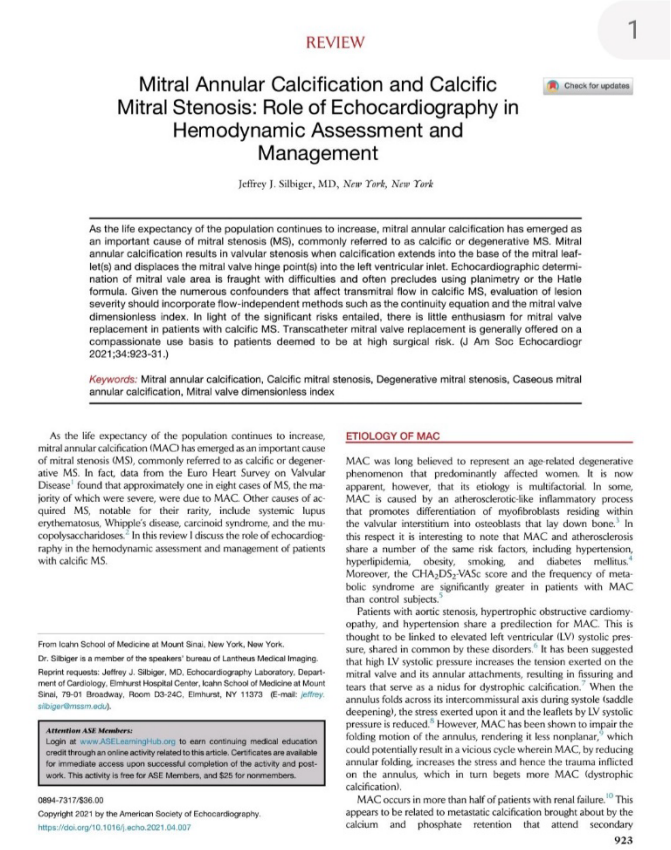
Insights from Simultaneous Echo and Cath Gradients in Rheumatic Compared to Calcific Mitral Stenosis - Journal of the American Society of Echocardiography (http://onlinejase.com)

@EGarciaSayan: differentiating calcific from rheumatic mitral stenosis

@iamritu: 🔑 differences b/w rheumatic & calcification mitral stenosis



@DavidWienerMD: Here's a review of calcific MS, whose assessment is different, from @JournalASEcho



REVIEW

1

Mitral Annular Calcification and Calcific Mitral Stenosis: Role of Echocardiography in Hemodynamic Assessment and Management

Jeffrey J. Silbiger, MD, *New York, New York*

As the life expectancy of the population continues to increase, mitral annular calcification has emerged as an important cause of mitral stenosis (MS), commonly referred to as calcific or degenerative MS. Mitral annular calcification results in valvular stenosis when calcification extends into the base of the mitral leaflet(s) and displaces the mitral valve hinge point(s) into the left ventricular inlet. Echocardiographic determination of mitral vale area is fraught with difficulties and often precludes using planimetry or the Hatle formula. Given the numerous confounders that affect transmitral flow in calcific MS, evaluation of lesion severity should incorporate flow-independent methods such as the continuity equation and the mitral valve dimensionless index. In light of the significant risks entailed, there is little enthusiasm for mitral valve replacement in patients with calcific MS. Transcatheter mitral valve replacement is generally offered on a compassionate use basis to patients deemed to be at high surgical risk. (J Am Soc Echocardiogr 2021;34:923-31.)

Keywords: Mitral annular calcification, Calcific mitral stenosis, Degenerative mitral stenosis, Caseous mitral annular calcification, Mitral valve dimensionless index

As the life expectancy of the population continues to increase, mitral annular calcification (MAC) has emerged as an important cause of mitral stenosis (MS), commonly referred to as calcific or degenerative MS. In fact, data from the Euro Heart Survey on Valvular Disease[1] found that approximately one in eight cases of MS, the majority of which were severe, were due to MAC. Other causes of acquired MS, notable for their rarity, include systemic lupus erythematosus, Whipple's disease, carcinoid syndrome, and the mucopolysaccharidoses.[2] In this review I discuss the role of echocardiography in the hemodynamic assessment and management of patients with calcific MS.

From Icahn School of Medicine at Mount Sinai, New York, New York.

Dr. Silbiger is a member of the speakers' bureau of Lantheus Medical Imaging.

Reprint requests: Jeffrey J. Silbiger, MD, Echocardiography Laboratory, Department of Cardiology, Elmhurst Hospital Center, Icahn School of Medicine at Mount Sinai, 79-01 Broadway, Room D3-24C, Elmhurst, NY 11373 (E-mail: jeffrey.silbiger@mssm.edu).

0894-7317/$36.00

https://doi.org/10.1016/j.echo.2021.04.007

ETIOLOGY OF MAC

MAC was long believed to represent an age-related degenerative phenomenon that predominantly affected women. It is now apparent, however, that its etiology is multifactorial. In some, MAC is caused by an atherosclerotic-like inflammatory process that promotes differentiation of myofibroblasts residing within the valvular interstitium into osteoblasts that lay down bone.[3] In this respect it is interesting to note that MAC and atherosclerosis share a number of the same risk factors, including hypertension, hyperlipidemia, obesity, smoking, and diabetes mellitus.[4] Moreover, the CHA_2DS_2-VASc score and the frequency of metabolic syndrome are significantly greater in patients with MAC than control subjects.[5]

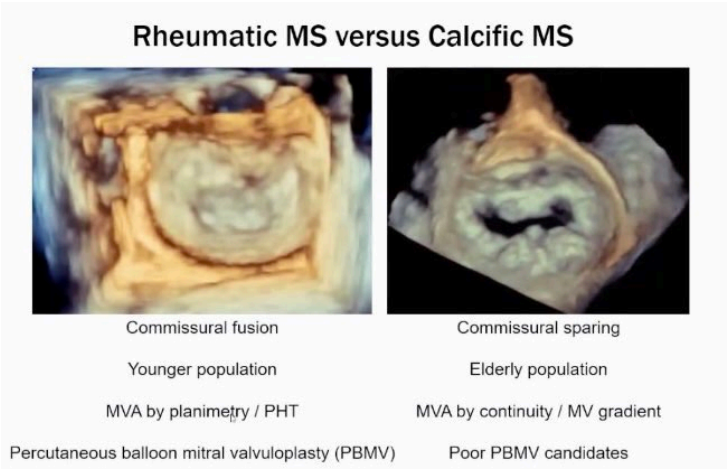
Patients with aortic stenosis, hypertrophic obstructive cardiomyopathy, and hypertension share a predilection for MAC. This is thought to be linked to elevated left ventricular (LV) systolic pressure, shared in common by these disorders.[6] It has been suggested that high LV systolic pressure increases the tension exerted on the mitral valve and its annular attachments, resulting in fissuring and tears that serve as a nidus for dystrophic calcification.[7] When the annulus folds across its intercommissural axis during systole (saddle deepening), the stress exerted upon it and the leaflets by LV systolic pressure is reduced.[8] However, MAC has been shown to impair the folding motion of the annulus, rendering it less nonplanar,[9] which could potentially result in a vicious cycle wherein MAC, by reducing annular folding, increases the stress and hence the trauma inflicted on the annulus, which in turn begets more MAC (dystrophic calcification).

MAC occurs in more than half of patients with renal failure.[10] This appears to be related to metastatic calcification brought about by the calcium and phosphate retention that attend secondary

@senguptasp: Commissural fusion , leaflet thickening , restricted leaflet motion causing hockey stick appearance , chordal thickening and shortening

@EGarciaSayan: morphological characteristics or rheumatic mitral stenosis

@iamritu: MV thickening & commissural fusion ➡progressive⬇ in mitral valve area characteristic of rheumatic MS 🆚 Calcific MS,stenosis rarely >mild unless annular calcification extensive & involves basal parts of leaflets. Rheumatic MS involves tips & margins of leaflet



@DavidWienerMD: Rheumatic MS is a funnel (restriction at leaflet tips)

Calcufic MS is a tunnel (restriction at annulus)

@senguptasp: Severe MS has valve area<1.5cm2, PHT > 150 marc, Mean gradient > 10 mmHg with PASP > 50

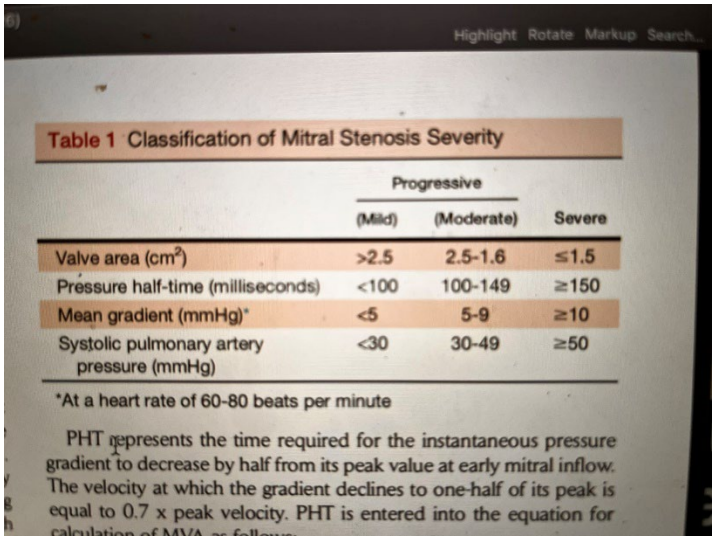


Table 1 Classification of Mitral Stenosis Severity

	Progressive		
	(Mild)	(Moderate)	Severe
Valve area (cm^2)	>2.5	2.5-1.6	≤1.5
Pressure half-time (milliseconds)	<100	100-149	≥150
Mean gradient (mmHg)*	<5	5-9	≥10
Systolic pulmonary artery pressure (mmHg)	<30	30-49	≥50

*At a heart rate of 60-80 beats per minute

PHT represents the time required for the instantaneous pressure gradient to decrease by half from its peak value at early mitral inflow. The velocity at which the gradient declines to one-half of its peak is equal to 0.7 x peak velocity. PHT is entered into the equation for

@pattypellikka: Watch out for high output states or concurrent mitral regurgitation which will cause a disproportionate increase in the transmitral flow velocity and pressure gradient

@pattypellikka: Transmitral gradients are flow &HR dependent. Use projected transmitral gradient for Doppler hemodynamics: A Novel Assessment Using Projected Transmitral Gradient Improves Diagnostic Yield of Doppler Hemodynamics in Rheumatic and Calcific Mitral Stenosis. | Semantic Scholar

@Drmonicaa_D: Ma'am .. with really high regards n respect .. a great tip.. wil be really kind enough if u could explain how to do projected transmitral gradient

@mariovar55: Opening area reduced

Comisural fusion

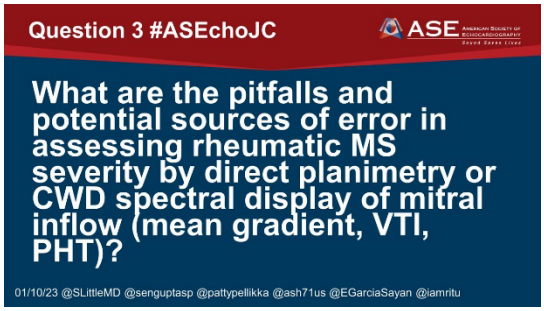
Thickened valves usually associated with shortened ,fusionadas and thickened chordae

Compromise simultaneously of other valves

@iamritu: MV most common valve involved in first & recurrent carditis.Nodularity (beading) along leaflet seen. Severe MR dominates almost 90% of times. if severe AR & only trivial MR less likely ARF

@AhmedDa77274329: MV area

Question 3:



A3 Notable Responses:

@pattypellikka: When measuring the mitral valve area by planimetry, the short axis imaging plane must be positioned at the tip of the valve leaflets to avoid over estimation. Examine the valve closely in SAX view, slowly tilting the transducer to the level of the valve tip.

@DavidWienerMD: Measuring at the tip of the funnel!

3D echo MPR helps measure it accurately

@DrCCaroli: Other interesting way: mitral separation index.

📏PLAX maximal diastolic distance between mitral valve tips (cm) x 1.6. This formula estimate mitral area and correlates with planimetry (SAX).

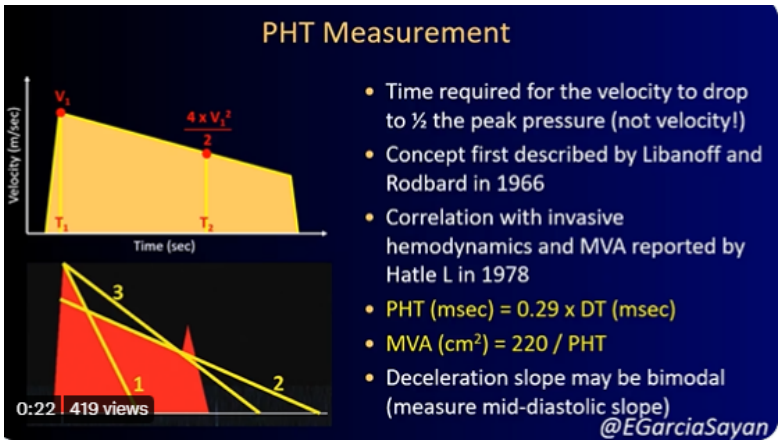
@EGarciaSayan: 🔺MG influenced by HR (ok only between 60-80 bpm), cardiac output, mixed valve disease

🔺Always report rhythm and heart rate

🔺PHT >150 ~ MVA <1.5 cm2 in RHD, assuming normal LV & LA compliance (unreliable in calcific MS)

🔺Trace mid-diastolic slope

🔺Avoid after PMBV

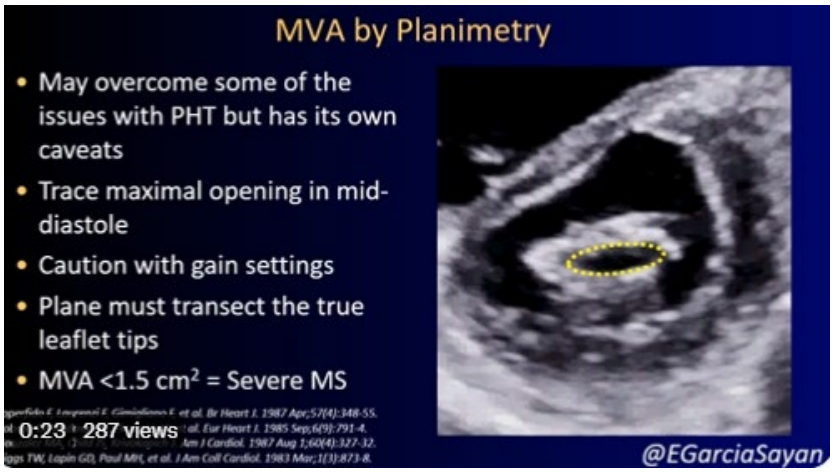


@EGarciaSayan: 🔺2D planimetry should transect leaflet tips perpendicular to LV axis

🔺Overestimation is common; look at the shape of LV in SAX (⭕vs🥚)

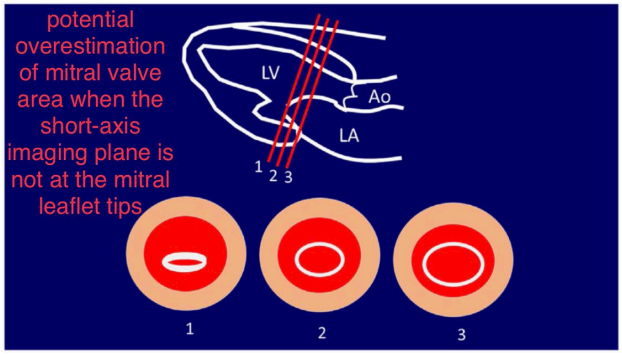
🔺Caution with gain settings

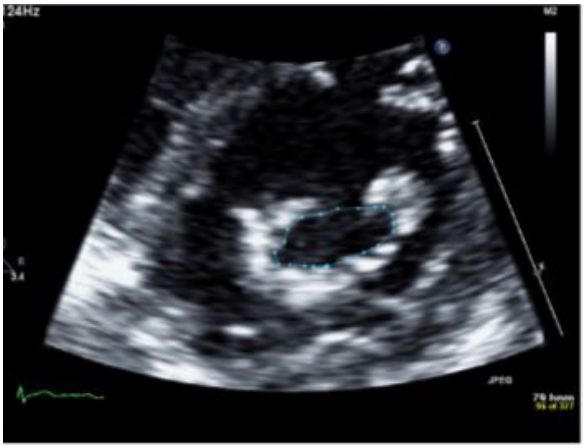
🔺Consider biplane imaging & 3D



@iamritu: When measuring MVA by planimetry, short-axis imaging plane should be positioned at tip of valve to avoid overestimation

New in these guidelines severe MS MVA ≤ 1.5 cm2 (old guidelines ≤ 1.0 cm2) Image subvalvular region in SAX & tilt transducer to level of valve tip





@SIwa23288585: I am sure 1gt tip of valve🤔

@EGarciaSayan: Pitfalls and potential sources of error in assessing rheumatic MS severity by direct planimetry. Severe MS is defined as MVA <1.5 cm2 (not 1 cm2!)

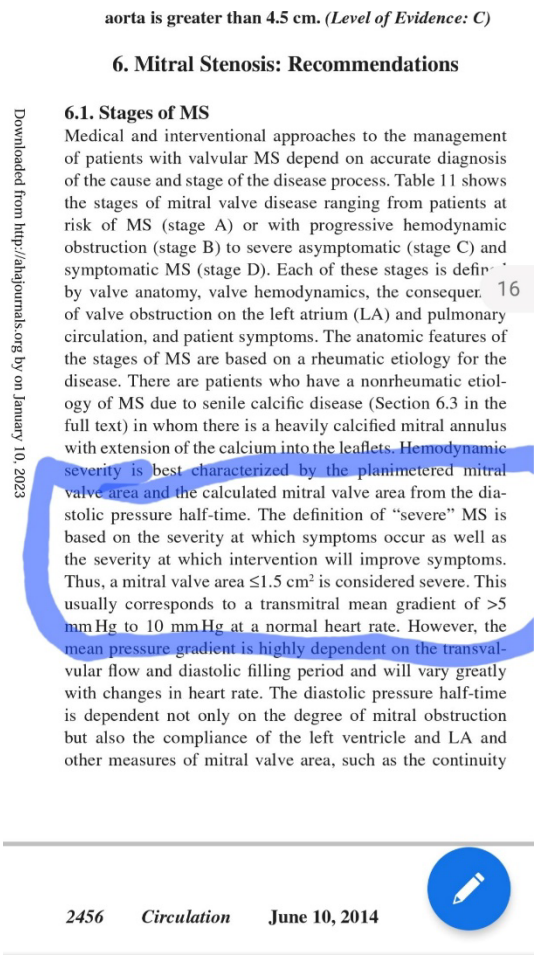
@rajdoc2005: We use <=1.5 cmsq for severe MS. I am curious WHY the severity of MS changed in the guidelines from 1 to 1.5? Thoughts?🤔🤔

@EGarciaSayan: Good question, I've wondered that too. My guess is that unlike in other valve diseases where the rule is to intervene if severe & symptomatic, those previously known as "moderate MS" (MVA 1-1.5) are often symptomatic and had an indication for intervention even in old guidelines.

@DrCCaroli: New MS guidelines are more close to the clinical reality of the patients👥. Symtomatic MS with an area between 1-1.5 cm2 are frecuent in practice🫁.

@rajdoc2005: Thanks @EGarciaSayan That's a reasonable explanation. Makes sense.

@DavidWienerMD: Reaching back to the 2014 ACC/AHA valve guideline



aorta is greater than 4.5 cm. *(Level of Evidence: C)*

6. Mitral Stenosis: Recommendations

6.1. Stages of MS

Medical and interventional approaches to the management of patients with valvular MS depend on accurate diagnosis of the cause and stage of the disease process. Table 11 shows the stages of mitral valve disease ranging from patients at risk of MS (stage A) or with progressive hemodynamic obstruction (stage B) to severe asymptomatic (stage C) and symptomatic MS (stage D). Each of these stages is defin[illegible] by valve anatomy, valve hemodynamics, the consequer[illegible] of valve obstruction on the left atrium (LA) and pulmonary circulation, and patient symptoms. The anatomic features of the stages of MS are based on a rheumatic etiology for the disease. There are patients who have a nonrheumatic etiology of MS due to senile calcific disease (Section 6.3 in the full text) in whom there is a heavily calcified mitral annulus with extension of the calcium into the leaflets. Hemodynamic severity is best characterized by the planimetered mitral valve area and the calculated mitral valve area from the diastolic pressure half-time. The definition of "severe" MS is based on the severity at which symptoms occur as well as the severity at which intervention will improve symptoms. Thus, a mitral valve area ≤1.5 cm^2 is considered severe. This usually corresponds to a transmitral mean gradient of >5 mm Hg to 10 mm Hg at a normal heart rate. However, the mean pressure gradient is highly dependent on the transvalvular flow and diastolic filling period and will vary greatly with changes in heart rate. The diastolic pressure half-time is dependent not only on the degree of mitral obstruction but also the compliance of the left ventricle and LA and other measures of mitral valve area, such as the continuity

16

@rajdoc2005: Great info! Consolidates what @EGarciaSayan was referring to earlier…Revised guidelines based on symptoms! Thanks all! 🙏🙏

@EGarciaSayan: Another question is how do we "grade" non-severe progressive mitral stenosis. The RHD guidelines we just discussed propose moderate 1.6-2.5 and mild >2.5 whereas ACC/AHA guidelines do not suggest grading further from severe. Thoughts?

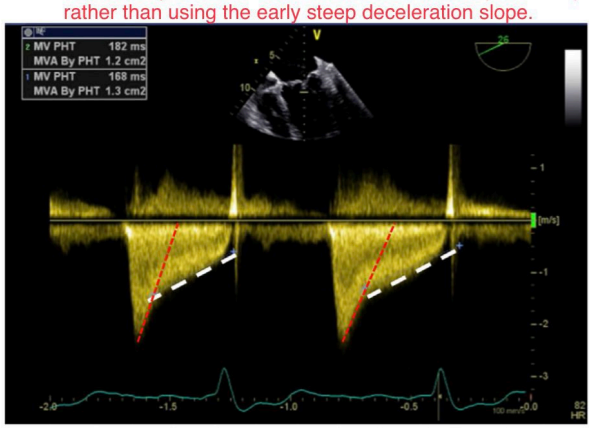
@abhinuv23: I think a very frequent conundrum is discrepancy between calculated or derived area and gradient.

@EGarciaSayan: Agree. Current guidelines definitely favor MVA over gradient in these settings, and Dx primarily based on MVA, due to the many factors that can influence MG.

@senguptasp: We need to make simpler for clinicians.. that's why made it simple

@iamritu: When the deceleration slope of the MV inflow is biphasic with a rapid decay (red line) followed by a more gradual decay, the deceleration slope should be traced in mid-diastole (white line) rather than using the early steep deceleration slope. http://bit.ly/3vWT4Je

When the deceleration slope of the MV inflow is biphasic with a rapid decay (red line) followed by a more gradual decay, the deceleration slope should be traced in mid-diastole (white line) rather than using the early steep deceleration slope.



Case #1

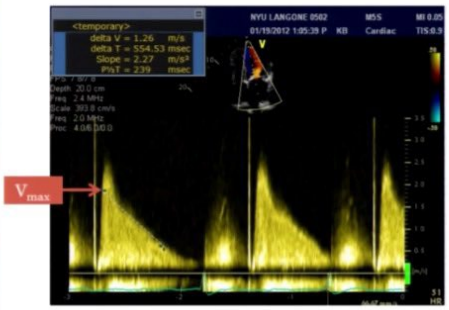
Answer:

Avoid the initial slope.

Use the mid-diastolic slope.

Extrapolate V_{max} based on mid-diastolic slope

This is the correct way to measure PHT when deceleration slope is bimodal.



PHT = 239 msec

MVA = 220 / 239
= **0.9 cm²**

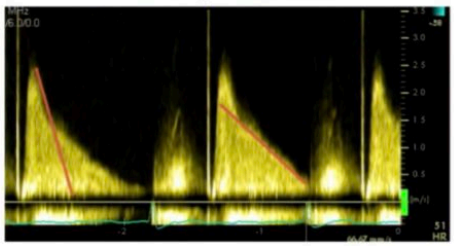
Case #1

Question:

My patient with mitral stenosis has a bimodal deceleration slope.

Can I still use the pressure half-time method to calculated the mitral valve area?

Which slope should I use?



PHT = 91 msec

MVA = 220 / 91 =
2.4 cm²

PHT = 239 msec

MVA = 220 / 239
= **0.9 cm²**

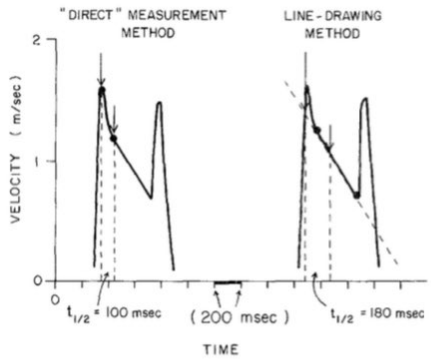


FIGURE 3. Direct measurement vs mid-diastolic line-drawing methods to determine Doppler pressure half-times ($T_{1/2}$). Direct method measures absolute time (ms) for the initial peak velocity to decrease to peak velocity/$\sqrt{2}$; in this example, $T_{1/2}$ (time between *dotted lines* from *arrow* at directly measured peak velocity and *arrow* at peak velocity/$\sqrt{2}$) is 100 ms. Line-drawing method uses the linear midportion of the velocity profile (*between the 2 dots*) whether or not it includes the peak velocity. The *first arrow* gives the extrapolated initial peak velocity and the *second arrow* the extrapolated peak velocity/$\sqrt{2}$, $T_{1/2}$ = 180 ms.

@rajdoc2005: This is such a crucial point to remember! Its a very common situation. Need to pay attention HOW and WHERE to measure the deceleration slope on the Doppler profile.

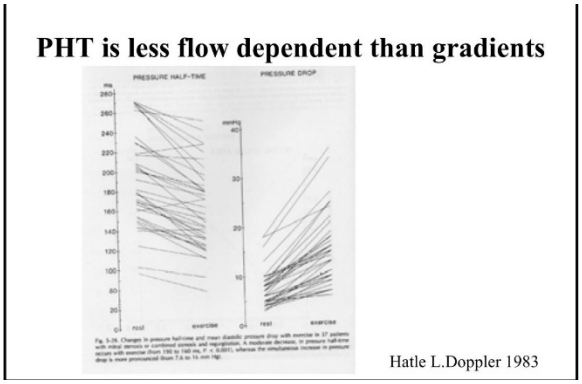
@senguptasp: CW Doppler for MS is flow and HR dependent , May change in high output states and ass. MR

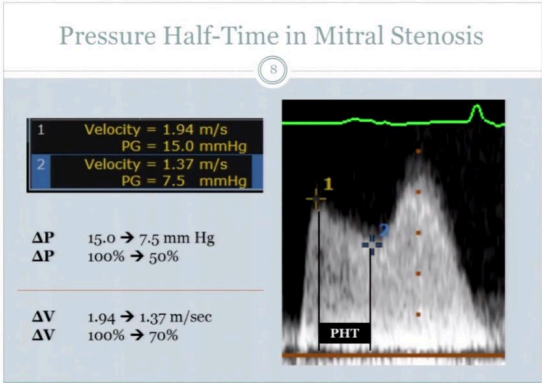
@EGarciaSayan: Pitfalls and potential sources or error in assessing MV severity based on mean transmitral gradient. MG influenced by HR (ok only between 60-80 bpm), cardiac output, mixed valve disease. Always report rhythm and heart rate

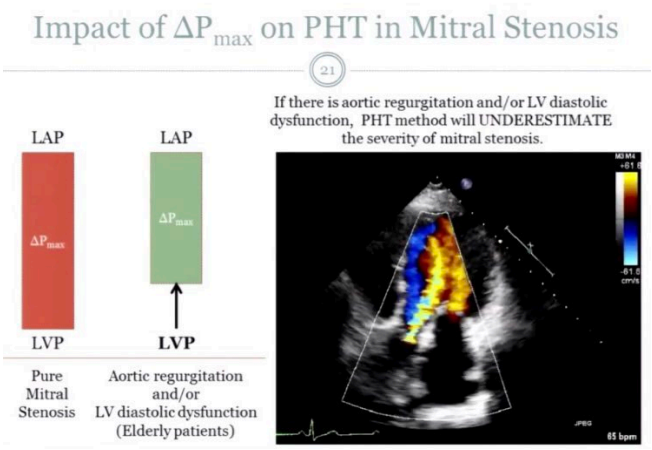
@iamritu: pHT:

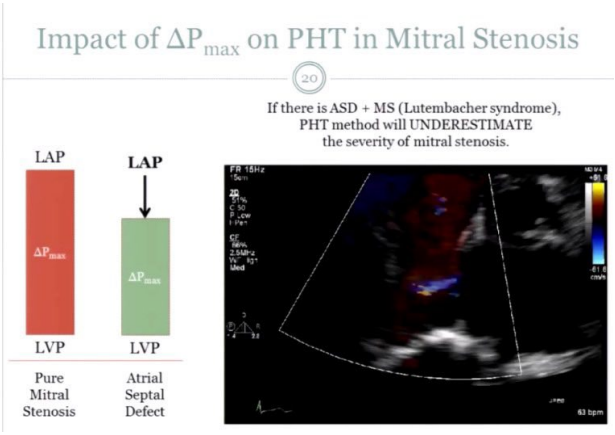
Less flow dependent than Doppler gradients

220/PHT formula not accurate for MVA when concomitant AR or ASD(changes ▲Pmax) or sudden shift in LA/LV compliance









@EGarciaSayan: Caveats of PHT measurement. Not accurate with conditions that impact LA or LV compliance or filling pressures. Do not use in calcific MS, immediately after PMBV, severe AR, etc.

@rajdoc2005: This is a classical board review question for fellows!

So fellows pay attention!!

@DavidWienerMD: MS+ASD = Lutembacher's syndrome. Great trivia question for #echofirst nerds, right @rajdoc2005

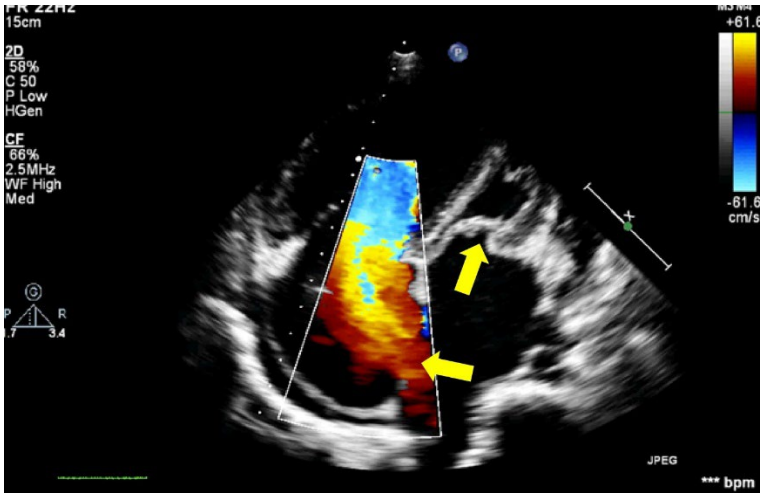
@SIwa23288585: Oh~ #CHD #ACHD

👏Lutembacher症候群

1916年にLutembacher がASDに先天性僧帽弁狭窄症 (MS) 合併症例報告しか し, MS原因 として は, 当初考 えられ ていた先天性病変よ りもリウマ チ性変化が多 く~

@DavidWienerMD: ありがとうございます

@EGarciaSayan: Lutembacher syndrome (congenital or acquired ASD + mitral stenosis). As a result PHT underestimated and MVA overestimated.



@SIwa23288585: good image, impressive👏👏👏

@rajdoc2005: Yup! You read my mind @DavidWienerMD And it is "RHEUMATIC" mitral stenosis in that syndrome!😁😁

@nilsonlopezmd: Several years later, this talk by @SLittleMD about mitral stenosis remains current and deals with several pitfalls in evaluating MS. Worth watching. Outstanding speaker!

https://www.youtube.com/watch?v=xbJB9fQdszo

@SLittleMD: Thank you for the shout out @nilsonlopezmd Great to know that the achieved talks can still be appreciated. 🙏

@DavidWienerMD: Ping @ASE360: our esteemed President

@senguptasp: Pitfalls of Planimetry by 2D - needs that we should cut at the MV tip which may be difficult at times .. 3D echo helps with multi planar imaging

@EGarciaSayan: Pitfalls and potential sources of error in assessing rheumatic MS severity by direct planimetry highlighted by @senguptasp. Image the tip of the leaflets, consider biplane imaging or 3D, avoid overestimation.

@SLittleMD: Important to consider that the anterior leaflet is longer than the posterior leaflet so diastolic flow is typically directed posteriorly into the LV. This means that the true-short axis of the MVA may not be parallel to the MV annular plane (or perpendicular to the LV long axis)

@SIwa23288585: Indeed👏👏👏

@iamritu: CWD mean gradient in mmHg defines severity

Mild <5

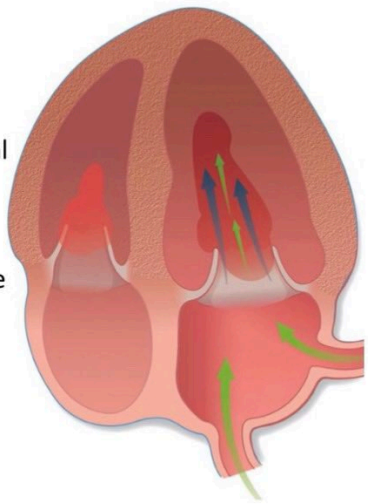
Moderate 6-10

Severe ≥10

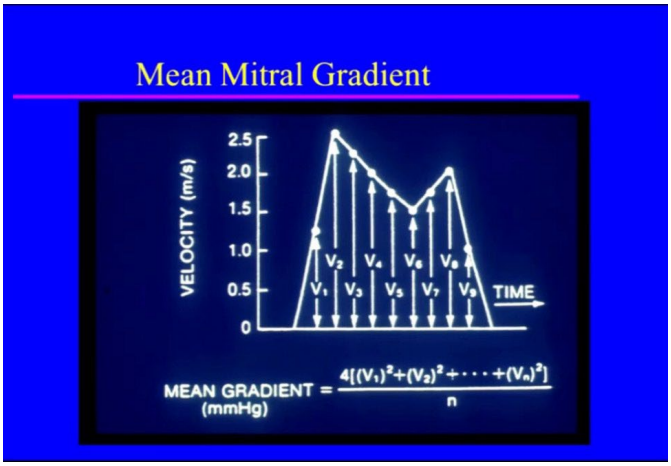
Measure CWD flow at mitral valve tips which reflects the maximum pressure difference between LA & LV

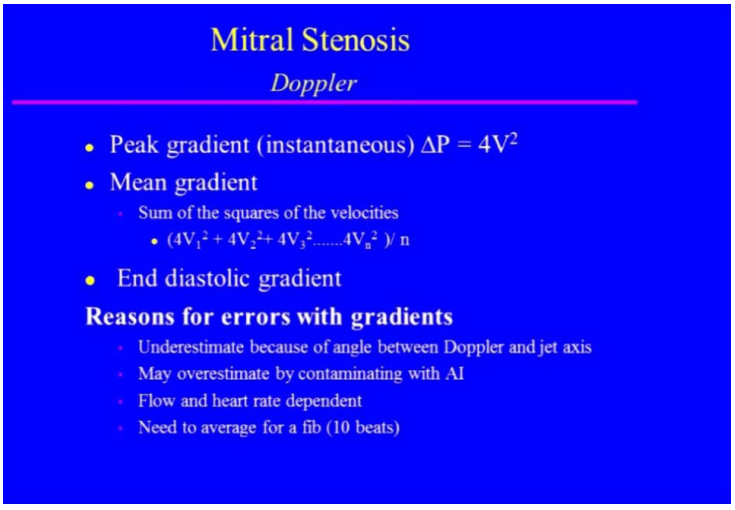
varies with heart rate,flow

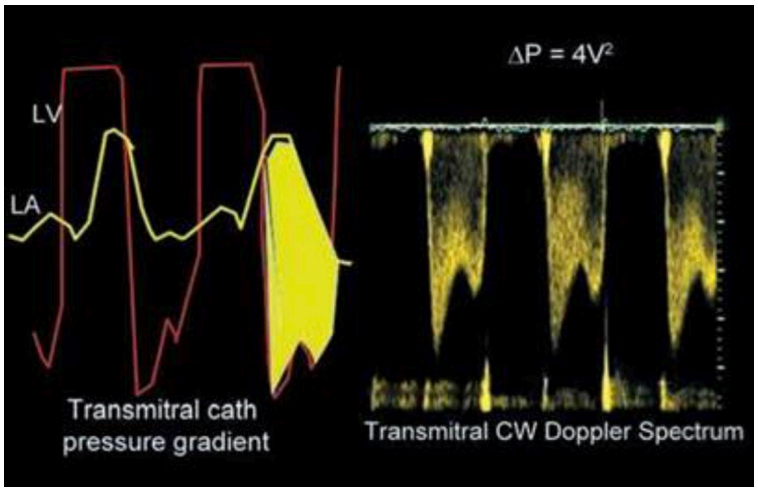
Doppler flow measured at the mitral valve tips reflect the maximal pressure differences between the left atrium and the left ventricle



Frank A. Flachskampf et al. JIMG 2015;8:1071-1093

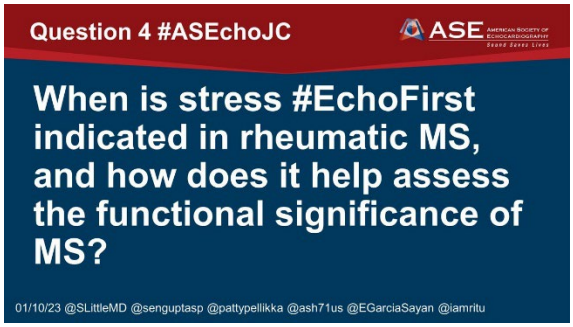






@RdickeyACS: CWD spectral display pitfalls: gradients are flow and heart rate dependent and should not be relied upon as the only parameters to assess MS severity. High output states or concurrent MR causes a disproportionate increase in the transmitral flow velocity and pressure gradient.

Question 4:

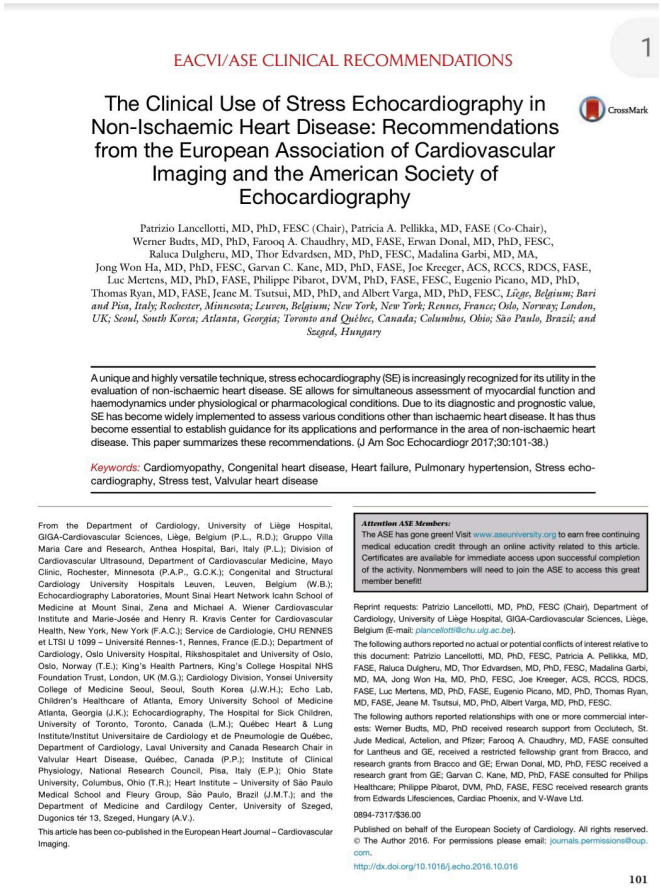


A4 Notable responses

@pattypellikka: Exercise echo is useful to assess the hemodynamic significance of mitral stenosis and its relationship to symptoms.

@EGarciaSayan: @pattypellikka highlights the use of stress #EchoFirst to evaluate mitral stenosis.👉Must read 2017 @ASE360 guideline for stress #EchoFirst in non-ischemic heart disease that Dr. Pellikka co-chaired: https://asecho.org/wp-content/uploads/2017/02/Stress-Echo-in-Non-Ischaemic-Guideline-2-2-17.pdf

@DavidWienerMD: And @ASE360 has a guideline for that! Right @pattypellikka?



EACVI/ASE CLINICAL RECOMMENDATIONS

1

The Clinical Use of Stress Echocardiography in Non-Ischaemic Heart Disease: Recommendations from the European Association of Cardiovascular Imaging and the American Society of Echocardiography

Patrizio Lancellotti, MD, PhD, FESC (Chair), Patricia A. Pellikka, MD, FASE (Co-Chair), Werner Budts, MD, PhD, Farooq A. Chaudhry, MD, FASE, Erwan Donal, MD, PhD, FESC, Raluca Dulgheru, MD, Thor Edvardsen, MD, PhD, FESC, Madalina Garbi, MD, MA, Jong Won Ha, MD, PhD, FESC, Garvan C. Kane, MD, PhD, FASE, Joe Kreeger, ACS, RCCS, RDCS, FASE, Luc Mertens, MD, PhD, FASE, Philippe Pibarot, DVM, PhD, FASE, FESC, Eugenio Picano, MD, PhD, Thomas Ryan, MD, FASE, Jeane M. Tsutsui, MD, PhD, and Albert Varga, MD, PhD, FESC, *Liège, Belgium; Bari and Pisa, Italy; Rochester, Minnesota; Leuven, Belgium; New York, New York; Rennes, France; Oslo, Norway; London, UK; Seoul, South Korea; Atlanta, Georgia; Toronto and Québec, Canada; Columbus, Ohio; São Paulo, Brazil; and Szeged, Hungary*

A unique and highly versatile technique, stress echocardiography (SE) is increasingly recognized for its utility in the evaluation of non-ischaemic heart disease. SE allows for simultaneous assessment of myocardial function and haemodynamics under physiological or pharmacological conditions. Due to its diagnostic and prognostic value, SE has become widely implemented to assess various conditions other than ischaemic heart disease. It has thus become essential to establish guidance for its applications and performance in the area of non-ischaemic heart disease. This paper summarizes these recommendations. (J Am Soc Echocardiogr 2017;30:101-38.)

Keywords: Cardiomyopathy, Congenital heart disease, Heart failure, Pulmonary hypertension, Stress echocardiography, Stress test, Valvular heart disease

From the Department of Cardiology, University of Liège Hospital, GIGA-Cardiovascular Sciences, Liège, Belgium (P.L., R.D.); Gruppo Villa Maria Care and Research, Anthea Hospital, Bari, Italy (P.L.); Division of Cardiovascular Ultrasound, Department of Cardiovascular Medicine, Mayo Clinic, Rochester, Minnesota (P.A.P., G.C.K.); Congenital and Structural Cardiology University Hospitals Leuven, Leuven, Belgium (W.B.); Echocardiography Laboratories, Mount Sinai Heart Network Icahn School of Medicine at Mount Sinai, Zena and Michael A. Wiener Cardiovascular Institute and Marie-Josée and Henry R. Kravis Center for Cardiovascular Health, New York, New York (F.A.C.); Service de Cardiologie, CHU RENNES et LTSI U 1099 – Université Rennes-1, Rennes, France (E.D.); Department of Cardiology, Oslo University Hospital, Rikshospitalet and University of Oslo, Oslo, Norway (T.E.); King's Health Partners, King's College Hospital NHS Foundation Trust, London, UK (M.G.); Cardiology Division, Yonsei University College of Medicine Seoul, Seoul, South Korea (J.W.H.); Echo Lab, Children's Healthcare of Atlanta, Emory University School of Medicine Atlanta, Georgia (J.K.); Echocardiography, The Hospital for Sick Children, University of Toronto, Toronto, Canada (L.M.); Québec Heart & Lung Institute/Institut Universitaire de Cardiology et de Pneumologie de Québec, Department of Cardiology, Laval University and Canada Research Chair in Valvular Heart Disease, Québec, Canada (P.P.); Institute of Clinical Physiology, National Research Council, Pisa, Italy (E.P.); Ohio State University, Columbus, Ohio (T.R.); Heart Institute – University of São Paulo Medical School and Fleury Group, São Paulo, Brazil (J.M.T.); and the Department of Medicine and Cardiology Center, University of Szeged, Dugonics tér 13, Szeged, Hungary (A.V.).

This article has been co-published in the European Heart Journal – Cardiovascular Imaging.

Reprint requests: Patrizio Lancellotti, MD, PhD, FESC (Chair), Department of Cardiology, University of Liège Hospital, GIGA-Cardiovascular Sciences, Liège, Belgium (E-mail: *plancellotti@chu.ulg.ac.be*).

The following authors reported no actual or potential conflicts of interest relative to this document: Patrizio Lancellotti, MD, PhD, FESC, Patricia A. Pellikka, MD, FASE, Raluca Dulgheru, MD, Thor Edvardsen, MD, PhD, FESC, Madalina Garbi, MD, MA, Jong Won Ha, MD, PhD, FESC, Joe Kreeger, ACS, RCCS, RDCS, FASE, Luc Mertens, MD, PhD, FASE, Eugenio Picano, MD, PhD, Thomas Ryan, MD, FASE, Jeane M. Tsutsui, MD, PhD, Albert Varga, MD, PhD, FESC.

The following authors reported relationships with one or more commercial interests: Werner Budts, MD, PhD received research support from Occlutech, St. Jude Medical, Actelion, and Pfizer; Farooq A. Chaudhry, MD, FASE consulted for Lantheus and GE, received a restricted fellowship grant from Bracco, and research grants from Bracco and GE; Erwan Donal, MD, PhD, FESC received a research grant from GE; Garvan C. Kane, MD, PhD, FASE consulted for Philips Healthcare; Philippe Pibarot, DVM, PhD, FASE, FESC received research grants from Edwards Lifesciences, Cardiac Phoenix, and V-Wave Ltd.

0894-7317/$36.00

http://dx.doi.org/10.1016/j.echo.2016.10.016

@EGarciaSayan: 🔺Stress #EchoFirst in non-severe MS + symptoms

🔺or asymptomatic MS before major surgery or pregnancy

🔺Exercise supine bicycle preferred, DSE can assess MG but not SPAP

🔺Severe MS: MG>15 mmHg or SPAP > 60 mmHg

▲2017

@ASE360 guidelines: https://asecho.org/wp-content/uploads/2017/02/Stress-Echo-in-Non-Ischaemic-Guideline-2-2-17.pdf

@Slwa23288585: #リウマチ性 #僧帽弁狭窄症

#stresseho #運動負荷心エコー

㊗ASEもオススメ👏👏👏

@SLittleMD: I'm starting to think that a Twitter JC is a stress test. My MV gradient is increasing!

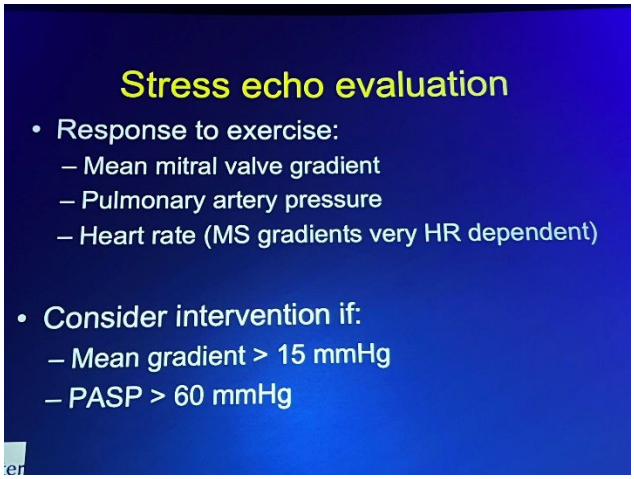
@senguptasp: Stress echo In MS when symptoms are inconsistent with echo data and for risk stratification . T mean gradient > 15 mm Hg or > 18 mmHg with Docutamine stress echo - Hemodynamic significant MS

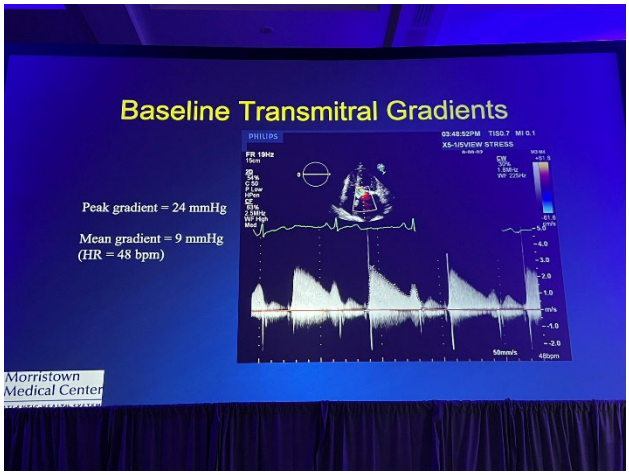
@EGarciaSayan: @senguptasp highlights indications for stress #EchoFirst in RHD. Non-severe MS + symptoms, or asymptomatic severe MS in certain situations. Look for MG>15 mmHg or SPAP > 60 mmHg with exercise.

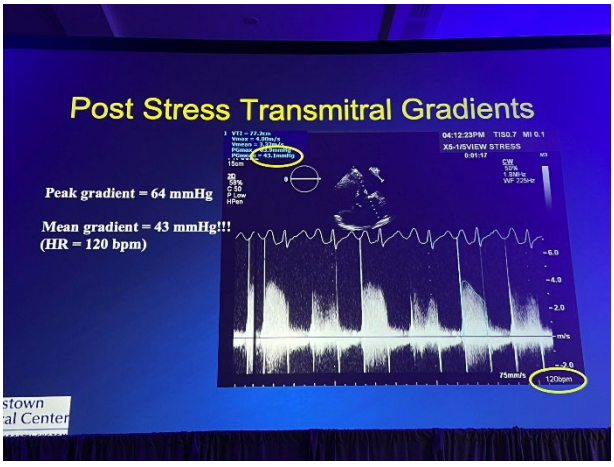
@pattypellikka: exercise echo helps when sx disproportionate to rest findings or if functional status is uncertain

@SLittleMD: DSE can be diagnostic with those criteria. But as in all conditions - exercise (bike, treadmill, handgrip) is preferred if the patient can perform.

@iamritu: if MVA between 1.0-1.5 & pt asymptomatic, can do stress #echofirst to assess hemodynamic significance & consider intervention if mean gradient increases with stress/exercise to >15 or PAP> 60 mmHg (note heart-rate)







Rheumatic MS is a slowly progressive disorder with an insidious natural history. The patient may be compensated and remain relatively asymptomatic at baseline despite the hemodynamic limitations posed by the stenosis. In some cases, this is due to patients limiting their activity. When imaging data and symptoms do not correlate with each other, stress echocardiography is useful to assess the functional significance of MS. Stress-induced increase in heart rate and cardiac output allows evaluation of the hemodynamic behavior of the valvular obstruction and its influence on the pulmonary circula-

@pattypellikka: The Clinical Use of Stress Echocardiography in Non-Ischaemic Heart Disease: Recommendations from the European Association of Cardiovascular Imaging and the American Society of Echocardiography - ScienceDirect

@doctormcintosh: Had a recent teenage complex #chd patient with an atrial switch and exertional symptoms where significant pulmonary venous baffle and lv inflow obstruction was uncovered with exercise stress echo and prompted baffle revision. Sx improved post op

@DavidWienerMD: Available in the newly reorganized Guidelines section of the @ASE360 website

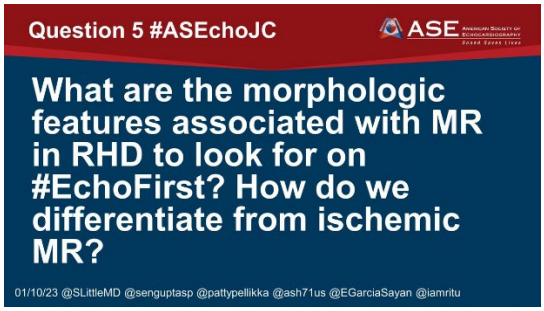
@EGarciaSayan: When is stress #EchoFirst indicated in rheumatic MS, and how does it help assess the functional significance of MS?

@SLittleMD: Exactly. An remember that the PAP at pk stress may be age dependent.

@EGarciaSayan: Another thing to consider is that when using DSE in patients that cannot exercise, can reliably look at mean gradient increase, but not PASP >60 mmHg (which should be looked at only with exercise)

@pattypellikka: agree. Also add color flow to make sure MR is not increasing--important when PMBV contemplated

Question 5:



A5 Notable responses

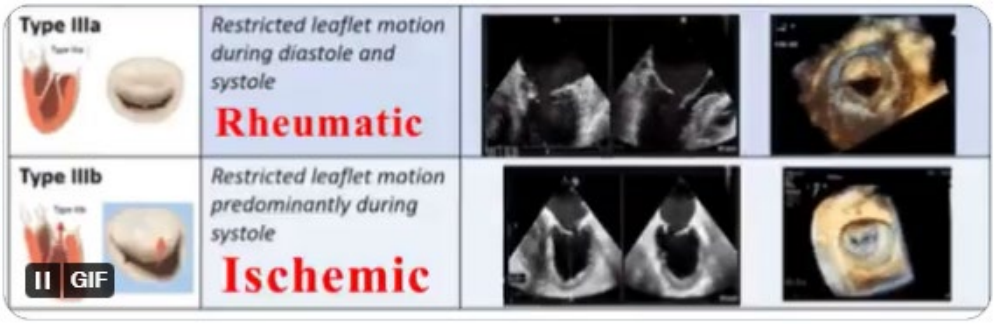
@pattypellikka: Hockey-stick deformity of mitral leaflets, with diastolic doming of the anterior leaflet and chordal thickening and calcification, well seen in the PLAX view, classic for rheumatic

@SLittleMD: Can be challenging to differentiate RHD MR from ischemic MR. Especially if posterior leaflet chords are shortened. Need to look for leaflet thickening with RHD (but which can also occur to a lesser degree with chronic secondary MR).

@iamritu: tough to tell apart Rheumatic VS ischemic MR sometimes. here's @fiore_corrado examining an explanted rheumatic MV with thickening & posterior leaflet chordal shortening /retraction

@EGarciaSayan: @SLittleMD highlights the challenges of differentiating RHD MR from ischemic MR, as these may coexist and overlap. Look for typical features of RHD, and the absence of RWMA.

@iamritu: Rheumatic Type 3A: excessive leaflet tip motion because of elongation or restriction & thickening of primary chords, displaced leaflet tips toward LA, leads to incomplete leaflet apposition MR VS Type 3B ischemic MR post leaflet tethering due to LV dysfunction w RWMA



@SIwa23288585: #リウマチ性 MR👉弁尖肥厚,過剰な動き,変位が原因

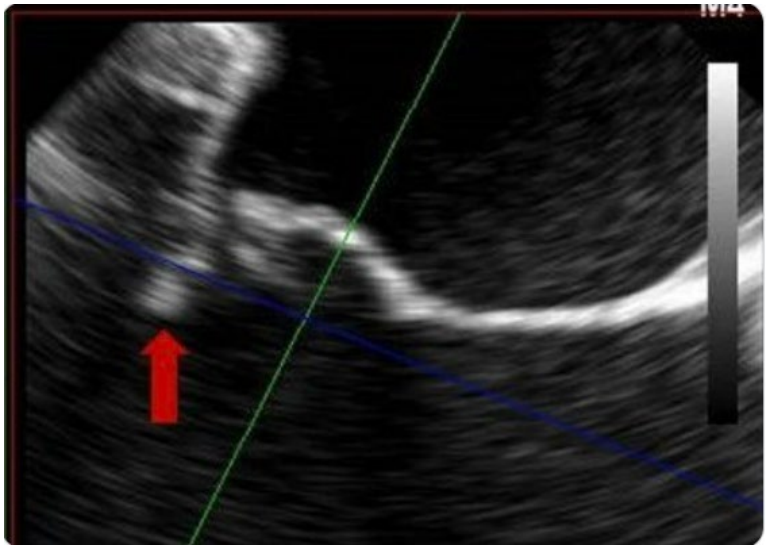
VS #虚血性 MR👉LV機能不全に伴う弁尖tethering

@SLittleMD: Really nice figures/video. Note how asymmetrically the posterior leaflet appears affected in the RHD case.

@rajdoc2005: Great point! Also look for other common valvular problems like Aortic Regurgitation which is more common in rheumatic MR. "You are the company you keep"😁

@SLittleMD: So true. The associated lesions can be very helpful to refine the etiology.

@iamritu: Can get pseudo prolapse with either ischemic or rheumatic MR but only ischemic will have LV dysfunction with RWMA that causes anterior leaflet pseudoprolapse

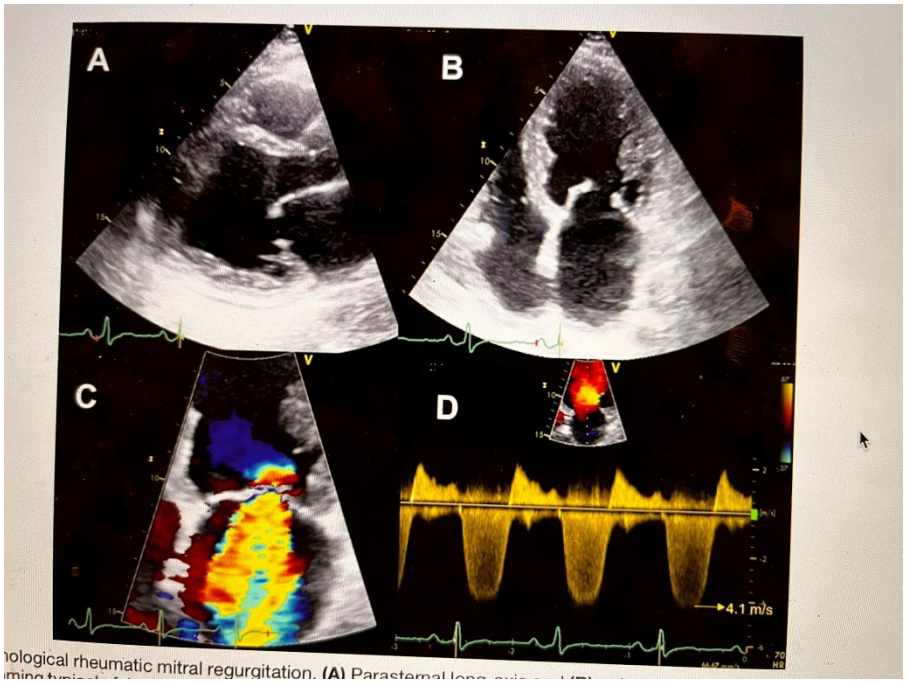


@SLittleMD: A great example of pseudo-prolapse (AKA end-to-side coaptation). If the basal posterior LV wall motion is normal (after a really good look), then RHD is more likely the cause of MR.

@senguptasp: RHD MR can be eccentric, central or wall jets

@ash71us: specifically MR from Acute Carditis? or MR from late manifestations of Carditis?

@senguptasp: Revised Jones criteria for pathological RHD MR - holodiastolic jet seen in at least 2 views and peak MR velocity > 3 m/ sec



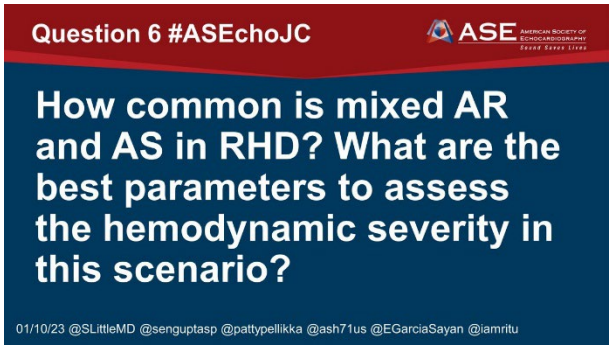
ological rheumatic mitral regurgitation. **(A)** Parasternal

@EGarciaSayan: Evaluation of pathological MR in RHD. Remember that valvular regurgitation is now part of the revised Jones criteria.

@ash71us: Jones criteria added #EchoFirst for the first time :)

@iamritu: MR is most frequent valvular abnormality in RHD thickened leaflets with doming in diastole. pathological mitral regurgitant jet in systole, w jet length ≥2 cm, CWD regurgitant flow w peak velocity ≥3 m/s

Question 6:



A6 Notable responses

@pattypellikka: Rheumatic AV disease -> commissural fusion, fibrotic thickening, retraction of the leaflet edges ->triangular or rounded orifice in systole. Mixed stenosis and regurgitation are common in rheumatic involvement of the aortic valve. Integrate multiple parameters to grade severity

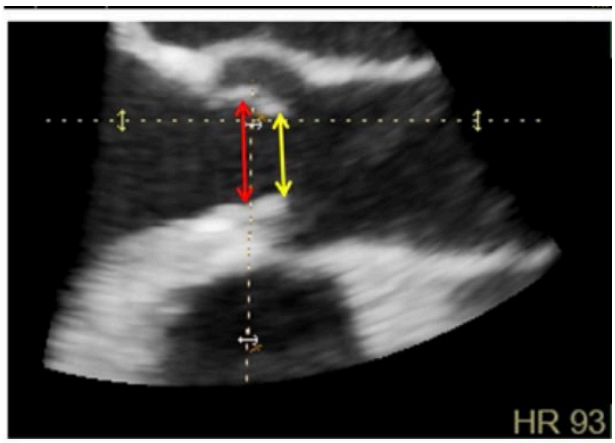
@senguptasp: RHD severe AS - transaortic flow V max > 4m/s, mean GR. > 40 mm Hg, AVA< 1cm2, indexed AVA <0.6 cm2/m2, LVOT/ AV velocity <0.25

@senguptasp: Aortic valve anatomy to be seen in zoomed view always - look for commisural fusion , thickening , calcification a cusp edges in PSAX

@senguptasp: RHD AV involved always occurs with RHD MV involvement. RHD AR is more common. Always look for Mixed AR and AS due of RHD

@EGarciaSayan: How common is mixed AR and AS in RHD? Regurgitation is more common (46.7%) than stenosis (14%) in rheumatic AV disease.

@iamritu: Rheumatic AS usually coexists w AR because of retraction of cusp edges Severe AR ↑ flow across LVOT & AV, thus peak velocity & gradients may be ↑ than expected for given valve area, use continuity equation rheumatic AV starts from edges rather than body/leaflet base



TEE view of AV marking the correct level of cut plane at the leaflet edges (yellow arrow) in contrast to a misplaced plane cut at the level of doming (red arrow) yielding a larger AVA.

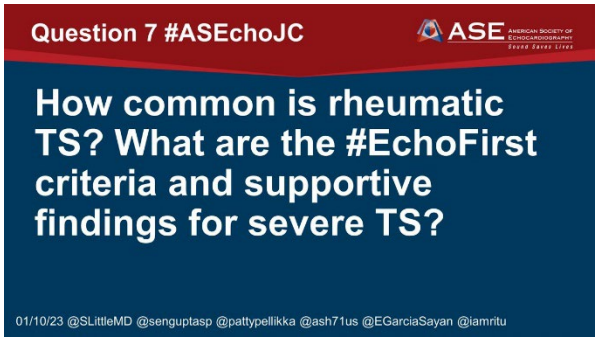
@EGarciaSayan: How common is mixed AR and AS in RHD? What are the best parameters to assess the hemodynamic severity in this scenario? @iamritu highlights caveats with using gradients in mixed valve disease

@SLittleMD: Another great image. RHD AV pathology also includes commissural fusion so in theory balloon-valvotomy should work for R AV stenosis - except AR so often coexists that BAV is not usually an option.

@SLittleMD: And if the MV is has normal structure and function, then the AV is not rheumatic.

@EGarciaSayan: Very important point. MV is almost always affected when RHD affects the aortic valve

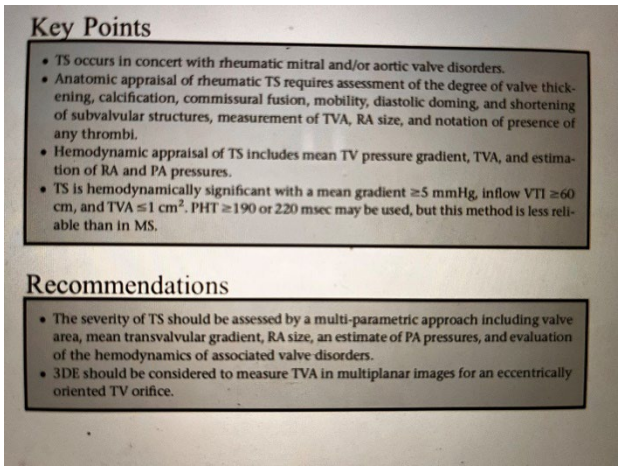
Question 7:



A7 Notable responses

@pattypellikka: Isolated rheumatic TS is uncommon; look for associated rheumatic mitral &/or AV involvement. Features of TV are similar to those of rheumatic MV but TV tends to be less calcified. Assess severity with multiparametric approach

@senguptasp: TV should always for looked in all RHD patients for TS . Here are the recommendations for TS

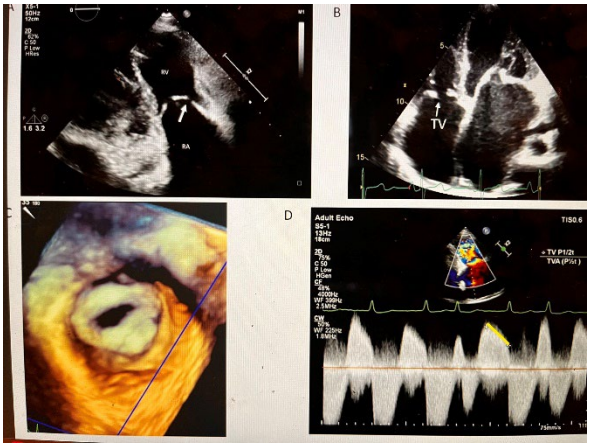


Key Points

- TS occurs in concert with rheumatic mitral and/or aortic valve disorders.
- Anatomic appraisal of rheumatic TS requires assessment of the degree of valve thickening, calcification, commissural fusion, mobility, diastolic doming, and shortening of subvalvular structures, measurement of TVA, RA size, and notation of presence of any thrombi.
- Hemodynamic appraisal of TS includes mean TV pressure gradient, TVA, and estimation of RA and PA pressures.
- TS is hemodynamically significant with a mean gradient ≥5 mmHg, inflow VTI ≥60 cm, and TVA ≤1 cm^2. PHT ≥190 or 220 msec may be used, but this method is less reliable than in MS.

Recommendations

- The severity of TS should be assessed by a multi-parametric approach including valve area, mean transvalvular gradient, RA size, an estimate of PA pressures, and evaluation of the hemodynamics of associated valve disorders.
- 3DE should be considered to measure TVA in multiplanar images for an eccentrically oriented TV orifice.



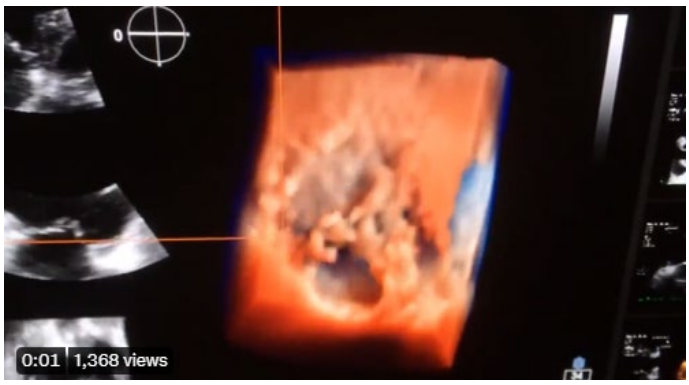
@iamritu: TS severe if mean gradient is ≥5 mmHg, & TVA ≤1 cm2.

TS is mild TVA >1.5 cm2, & moderate with a TVA 1-1.5 cm2

Isolated rheumatic TS is rare, usually with rheumatic mitral &/or aortic valve

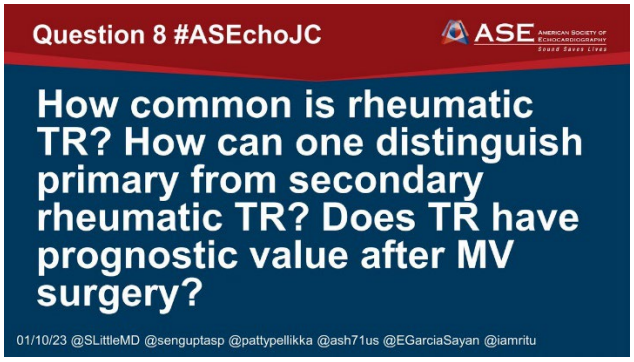
significant calcification of TV occurs less than MS

@iamritu: 🤩 3-D of rheumatic tricuspid valve by @omorenou



@ash71us: wish all tricuspid 3Ds looked like that!

Question 8:



A8 Notable responses

@pattypellikka: TR is estimated to have a prevalence of 70% in rheumatic heart disease and may be primary or secondary.

@SLittleMD: Begin with the question: primary or secondary TR? Look for TV thickening/diastolic doming. Recall that MV involvement "always" precedes other valve involvement so beware MV disorders with secondary TR from PH/RV dilation. And know that atrial TR can occur with chronic AF.

@senguptasp: Atrial TR is an important thing to be remembered

@pattypellikka: Tricuspid valve repair for moderate TR should be considered at the same time as mitral valve surgery. Moderate TR as a predictor of severe TR and severe TR, a predictor of heart failure

@Siwa23288585: #リウマチ性弁膜症 重症三尖弁閉鎖不全症も心不全予防の治療必要😳

@ash71us: Is the severity of TR mitigated by severe mitral valve disease?

@pattypellikka: more apt to be exacerbated

@iamritu: No & TR is a bad prognostic indicator post MVR & ongoing trial to look at prospectively treating TR at the time of MVR

@Mitrovalvology: regardless of annulal diameter ?

@jisaezdeibarra: Atrial fib., it's even a stronger predictor of severe TR later on and heart failure

@senguptasp: RHD TR is suspected when associated with diastolic doming or TV thickening

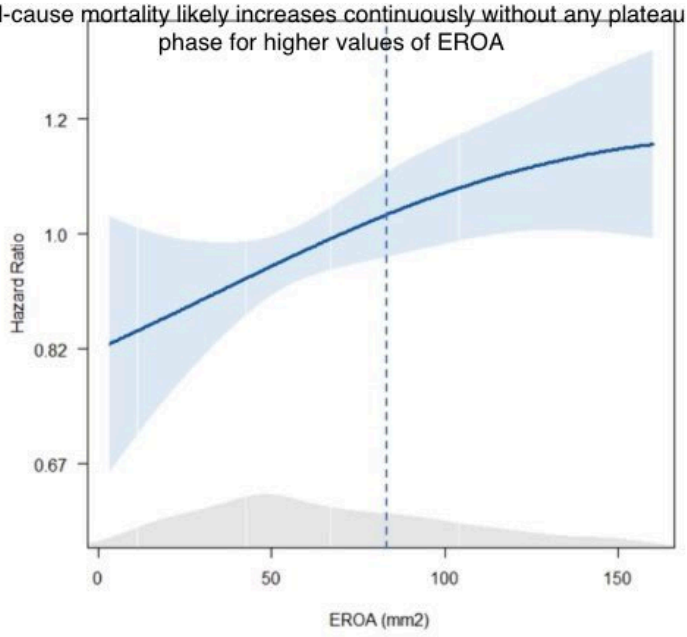
@senguptasp: Severe RHD TR - VC > 0.7 cm , VC > 0.4 cm2 via 3D Echo, PISA > 0.9 cm, EROA > 40 cm2, RG vol > 45 ml by PISA , reg Jet area > 10 cm2

@iamritu: TR 3rd most common valve lesion after MR & MS, almost always assoc w MS

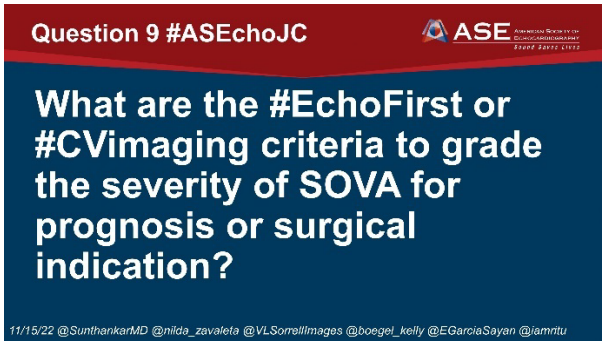
3DE allows for accurate measurements of tricuspid annulus & EROA of regurgitant jet

Continuum of risk with higher EROAs for TR no real plateau TR undertreated

All-cause mortality likely increases continuously without any plateau phase for higher values of EROA



Question 9:



A9 Notable responses

@pattypellikka: exercise echo can help to understand hemodynamic significance of mixed and multi-valve disease

@senguptasp: RHD AR and AS - AVA by continuity equation applicable

@pattypellikka: remember that volumetric quantification is inaccurate with multivalve disease.

@senguptasp: Don't use DSE to differentiate pseudo severe from True severe AS in presence of RHD severe MV disease

@EGarciaSayan: Very important point! LFLG AS can be due to severe mitral stenosis, and in general, we should avoid DSE in these settings.

@senguptasp: In combined AR and MS - proper align of Doppler signal and also timing of flow helps

@senguptasp: Combined MS and AS - best is measuring valve area by planimetry - 3D echo better than 2D echo

@swatigar: It seems the echo machine makers are least bothered about pediatric patients.

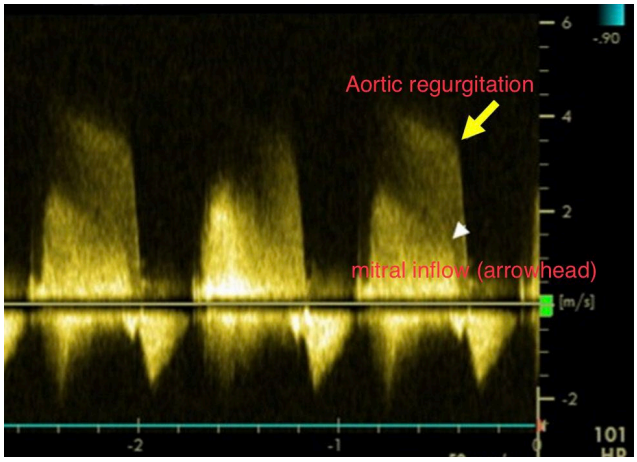
The 3d pediatric probe is not useable in most of our pts

- big footprint

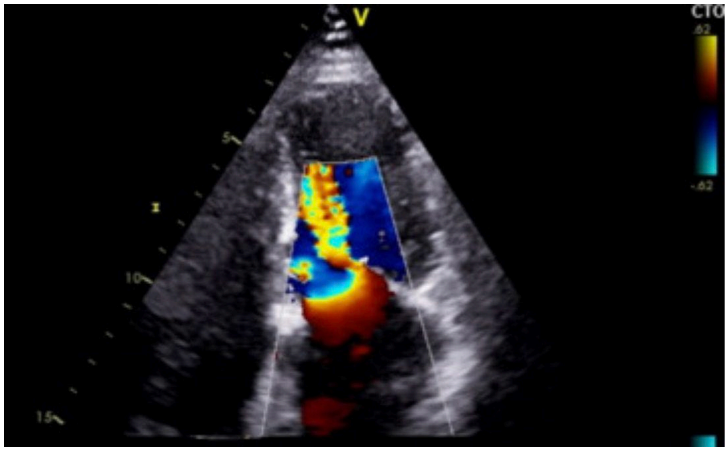
- terrible artefacts with resp and motion (uncontrollable in opd setting)

@iamritu: In mixed AR/MS

Timing of AR VS MS: AR jet starts earlier at aortic valve closure & lasts longer in diastole & has a higher velocity reflecting the higher Ao to LV pressures VS LV to LA pressures



@iamritu: aortic regurgitant jet can be mistaken for MS jet on color Doppler the funnel-shaped, stenotic MV orifice can be directed toward the interventricular septum, mimicking AR



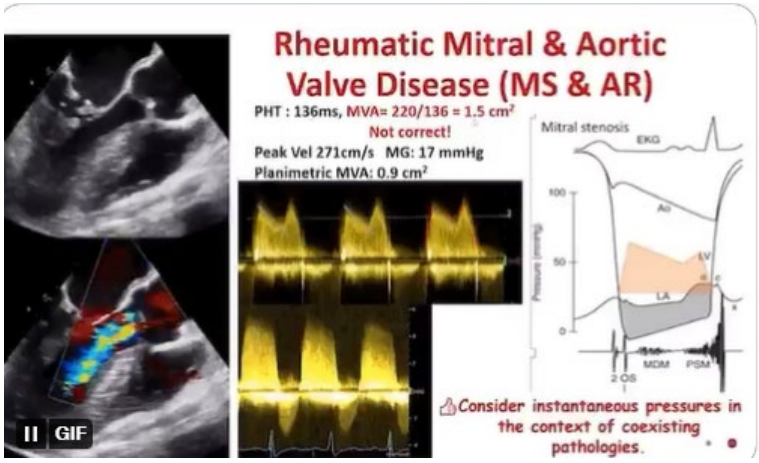
@rajdoc2005: Great point! This is not an uncommon occurrence. Need to pay attention while reading these echos.

Off axis views and panning through the LVOT can help - specifically looking for the exact origin of that diastolic flow into the LV!

@ash71us: Cool - not commonly seen in Adult labs in the US!

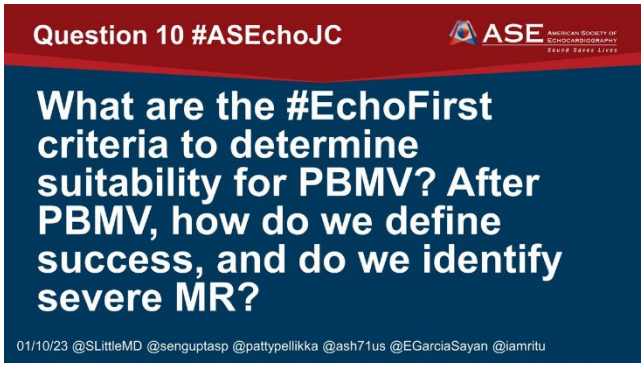
@iamritu: PHT will underestimate MVA in MS with severe aortic regurgitation because of rapid changes in compliance of LV & overestimation of MVA using PHT

preload is ↑ by AR,it's ↓ by MS opposing loading conditions may result in lower LV volumes c/w isolated AR



@leylaelifsade: Thank you for revisiting and twitting my presentation at the @escardio congress 2020 to learn more about traps in Doppler studies visit http://esc365.escardio.org/presentation/209993?

Question 10:



A10 Notable responses

@pattypellikka: •Degree of thickening & calcification of leaflets and commissures

•Extent, symmetry of commissure fusion

•Severity of subvalvular thickening, fusion

•Severity of MR

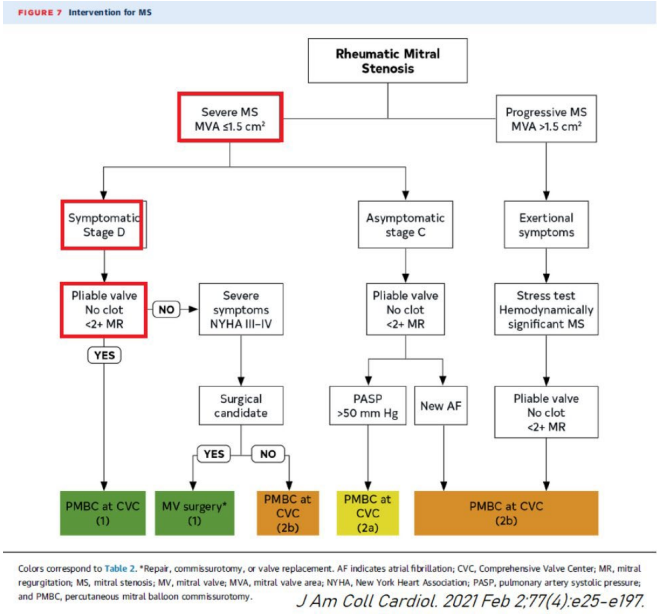
•LA or appendage clots

@EGarciaSayan: ⚠PBMV if symptomatic severe MS, pliable valve, <2+ MR & no LAA thrombus

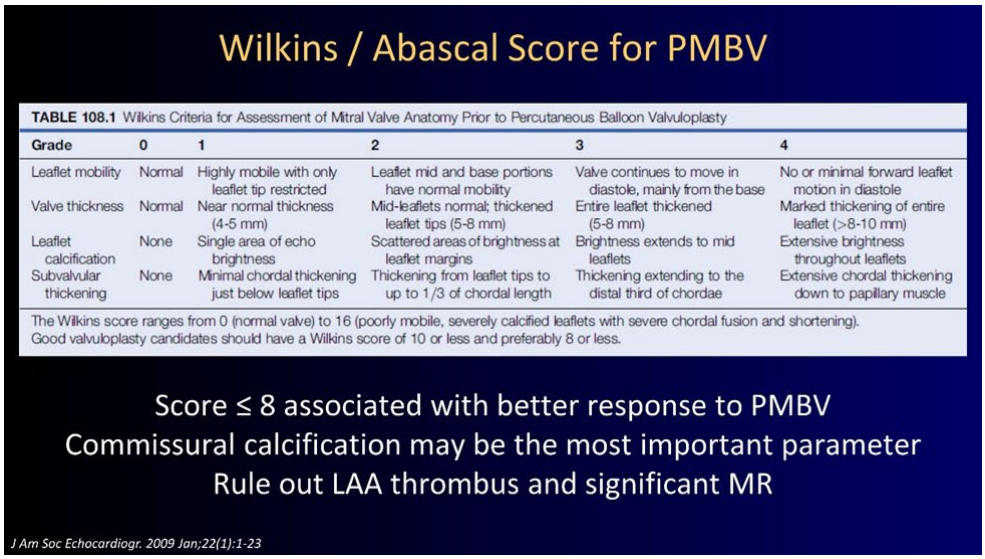
⚠Consider in asymptomatic severe MS & PASP>50 mmHg

⚠Wilkins score <=8

⚠Importance of careful #iEcho guidance, watch for severe acute MR



@EGarciaSayan: Wilkins score for assessing suitability for PBMV. Can consider >8 in very selected cases based on specific morphologic features.



@SLittleMD: thank you @EGarciaSayan @pattypellikka @iamritu @senguptasp and @JournalASEcho. What a fabulous forum for echo learning! I need to sign off now to let my mean MV gradient return to normal.

@EGarciaSayan: 😂Very fast-paced indeed! We should consider adding some guided meditation at the end of each #ASEchoJC

@ash71us: We are all a bit breathless 😊

@SLittleMD: I'll share the only mnemonic I know in Cardiology. Helps me remember the Wilkins score categories:

Can The Mitral Split?

C = Calcium

T = Thickening

M = Mobility

S = Sub-valvular apparatus

@EGarciaSayan: An excellent mnemonic to remember the Wilkins score shared by @SLittleMD. 0-4 points for each parameter, <=8 defines favorable morphology for PMBV

@iamritu: best mnemonic 😍for #Wilkins score

@DavidWienerMD: That's a great mnemonic. Thanks @SLittleMD

@EGarciaSayan: 👏This is fantastic, I've never heard this mnemonic before, but I will definitely share it with our fellows.

@TewanSuwanich: I remember it as Mi - Tral - S - Core 😂 Same meaning for each alphabet

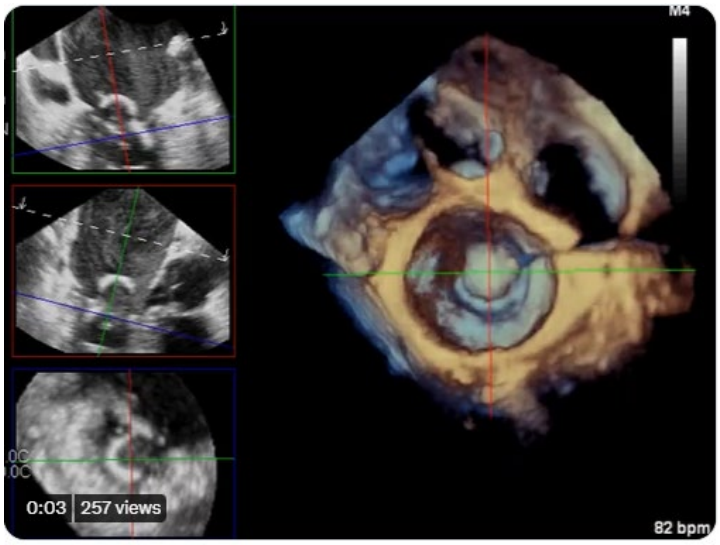
@EGarciaSayan: 🔺Interventional #EchoFirst (#iEcho) guidance for PBMV

🔺Reassess suitability, MR <2+, r/o LAAT

🔺Guide trans-septal puncture, balloon positioning & inflation (3D MPR can help)

🔺Assess gradient & MR after each inflation

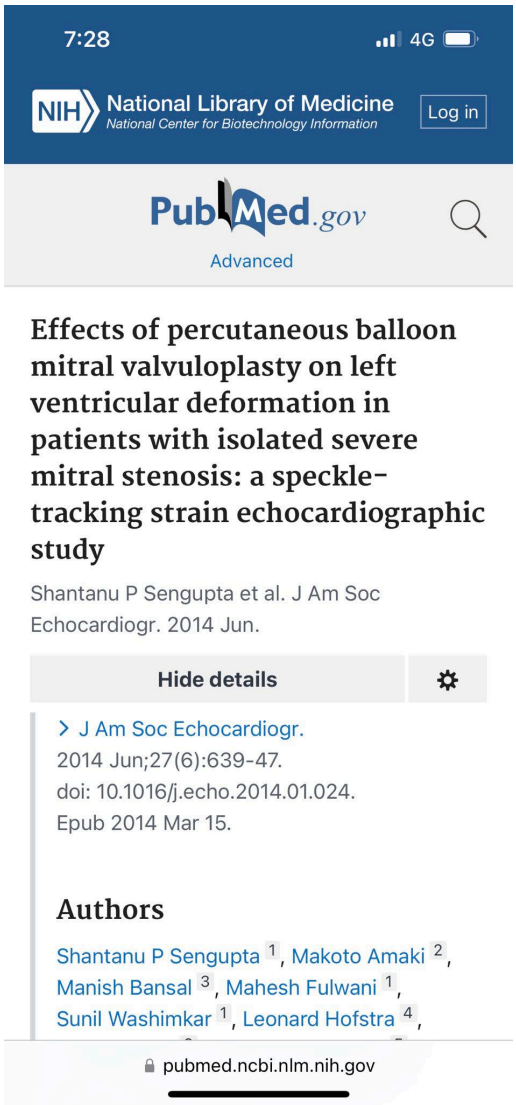
🔺Watch for acute severe MR, large residual ASD



@pattypellikka: After echo-guided PBMV, look for complications including tamponade, severe MR, extent of iatrogenic ASD

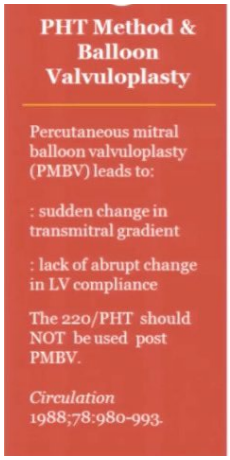
@EGarciaSayan: Importance of careful assessment for complications immediately following balloon inflation in PBMV. Assess gradient, MR, ASD, pericardium.

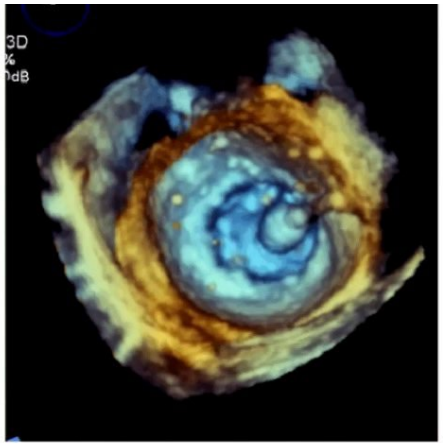
@senguptasp: Look for commisural fusion .. and be careful with presence of calcium . Irrespective of LV function try to go for PTMC if the MV are pliable as LV function improves after BMV

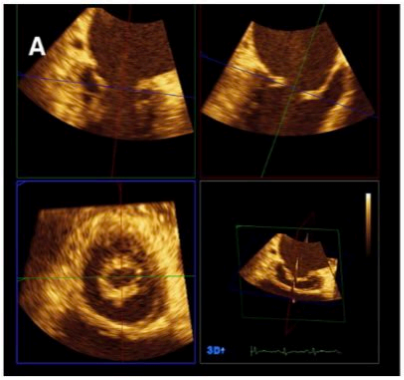


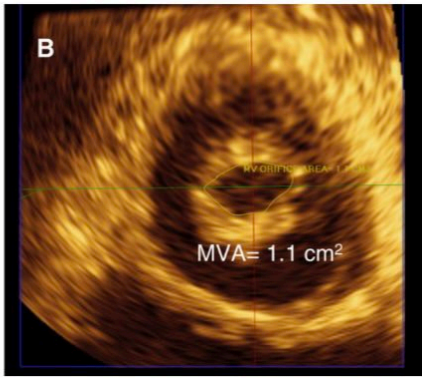
@iamritu: Successful PBMV is defined when a MVA ≥1.5 cm2 is achieved with no more than +2 MR

Post PMBV don't use PHT to calculate MVA, use planimetry from 3D









@EGarciaSayan: How do we define successful PMBV?

@iamritu: Several scoring systems, all almost equal in predicting outcomes

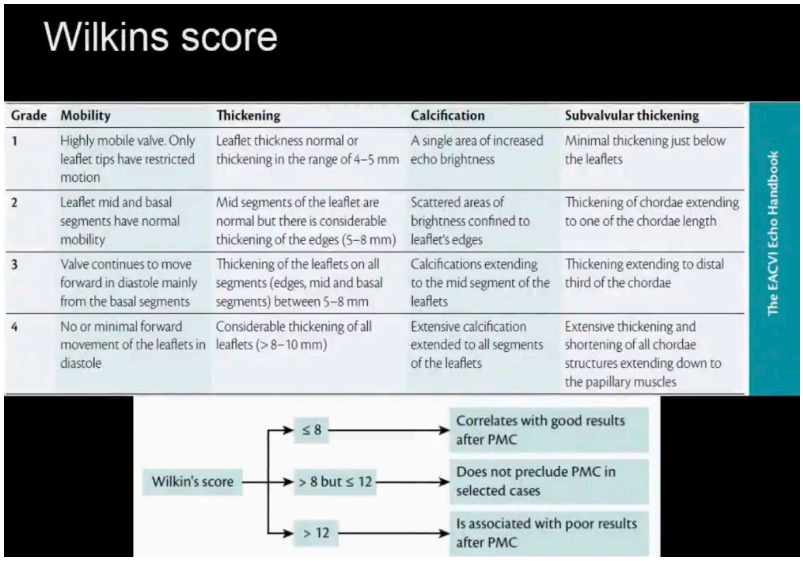
🔑 criteria:

degree of leaflet thickening, pliability/doming of leaflets (particularly anterior leaflet)

extent of calcification,

extent & symmetry of commissural fusion

severity of subvalvular disease



@EGarciaSayan: Wilkins score <=8 defines favorable anatomy for PMBV, but can consider intervention in those in 8-12 range in selected cases. Commissural calcification may be the most important parameter in these "borderline" cases.

@mariovar55: Wilkins score

Comisural calcification top on the list

Successful pbmv, opening 50% more the basal area, jo kore than 1+ MR

@EGarciaSayan: And that's all for tonight's #ASEchoJC!

🙏Thanks to authors @SLittleMD @senguptasp, @JournalASEcho EiC @pattypellikka & co-moderators @ash71us @iamritu

If you missed the live tweets, catch up on the discussion later by searching #ASEchoJC.

👉Claim CME: http://bit.ly/3VfuvBF

@EGarciaSayan: Another amazing #ASEchoJC! 🙏Thanks to authors @SLittleMD @senguptasp, @JournalASEcho EiC @pattypellikka & co-moderators @ash71us @iamritu. If you missed the live tweets, catch up on the discussion later by searching #ASEchoJC.

@pattypellikka: great session! Thank you, @EGarciaSayan @iamritu @senguptasp @SLittleMD @ash71us @JournalASEcho

@senguptasp: Fantastic session. Thank you.

@iamritu: Wow that was the fastest #ASEchoJC I was ever in! Awesome 😎 🤩 work by the guideline authors @SLittleMD @senguptasp tweeting from India! & our @JournalASEcho EIC @pattypellikka & #ASechoJC chair @EGarciaSayan & my co moderator @ash71us & friends @rajdoc2005 @DavidWienerMD & all

@rajdoc2005: Yet another great #ASEchoJC

@EGarciaSayan: Enjoyed participating and lots of fun tweeting - as always!!!😍😍

Thank you for joining @rajdoc2005! Your #ASEchoJC tweets are always excellent

@rajdoc2005: Thanks! Education is even more fun when you have your best friends around to share!🙏

Thanks again for another wonderful #ASEchoJC @ASE360 ❤️

@mswami001: Fantastic work! 👏🏾👏🏾👏🏾👏🏾

@RdickeyACS: Thank You @ASEchoJC!

@SIwa23288585: Thanks, Great time😍 fromJP