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Cardiomyopathies

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WHO definition of cardiomyopathy:

"Disease of the myocardium associated with cardiac dysfunction"

World Health Organization/International Society and Federation of Cardiology Task Force on the Definition and Classification of Cardiomyopathies

- ➤ Dilated Cardiomyopahty
- ➤ Hypertrophic Cardiomyopathy
- ➤ Restrictive Cardiomyopahty
- ➤ Arrhythmogenic Right Ventricular Cardiomyopathy
- >Unclassified Cardiomyopathies

Fibroelastosis
Noncompacted myocardium
Systolic dysfunction with minimal d

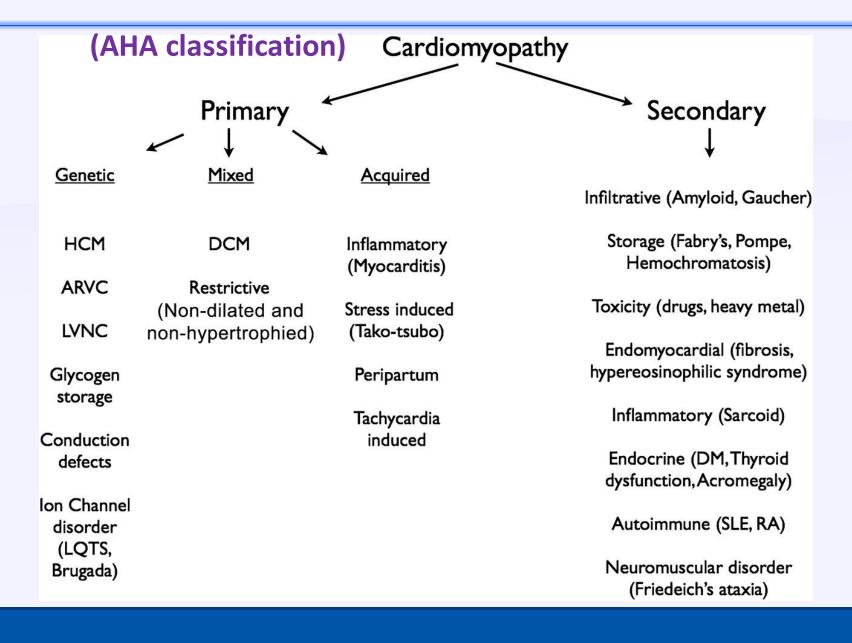
Systolic dysfunction with minimal dilatation Mitochondrial involvement

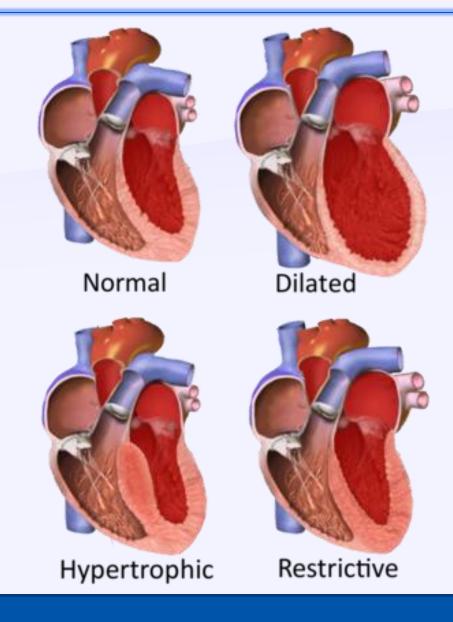
➤ Specific Cardiomyopahties

Ischemic cardiomyopathy
Valvular cardiomyopathy
Hypertensive cardiomyopathy
Inflammatory cardiomyopathy
Metabolic cardiomyopathy
General system disease
Muscular distrophies
Neuromuscular disorders
Sensitivity and toxic reactions
Peripartum cardiomyopathy

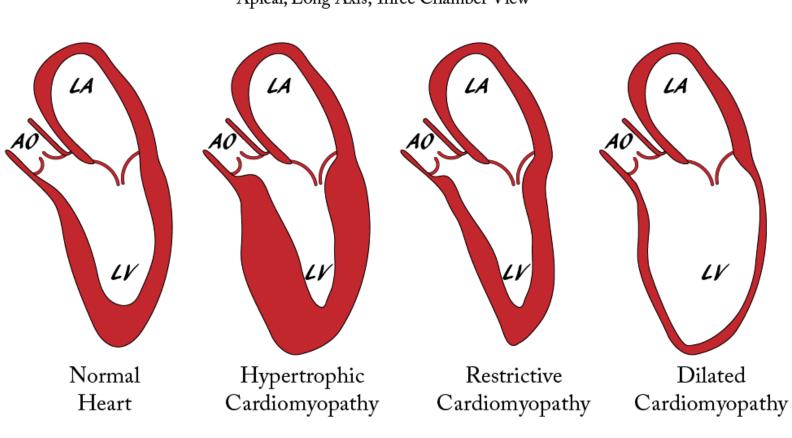
AHA Expert panel definition of cardiomyopathy:

- ➤ "A heterogeneous group of disease of the myocardium associated with mechanical and or / electrical dysfunction that usually (not invariably) exhibit inappropriate ventricular hypertrophy or dilatation and are due to variety of causes that frequently are genetic
- ➤ Cardiomyopathies are either confined to the heart or are part of generalized systemic disorders often leading to cardiovascular death or progressive heart failure- related disability".



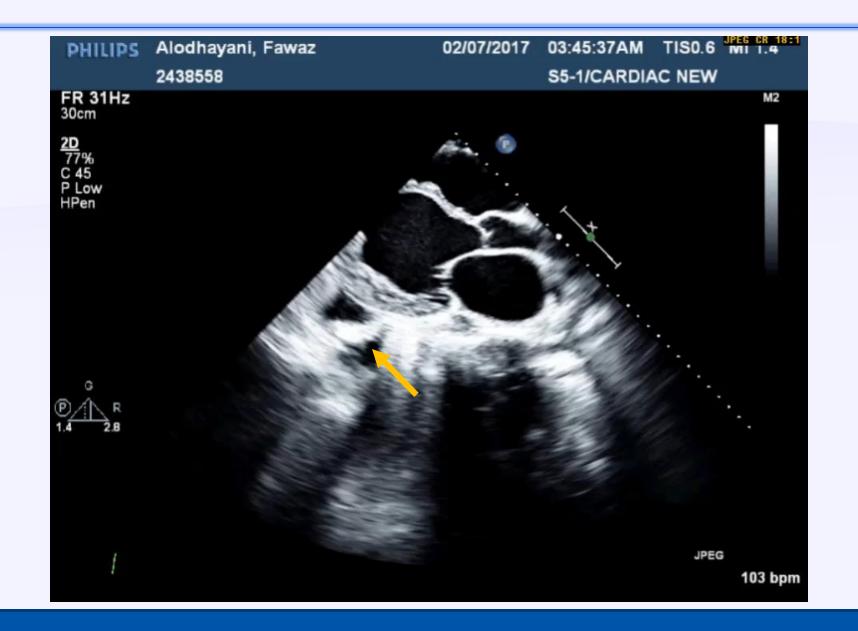


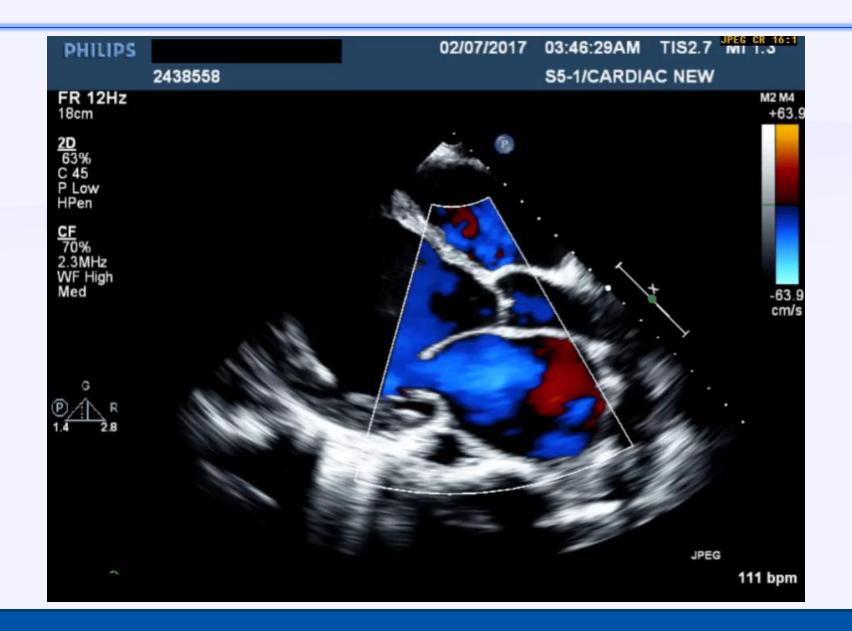
Cardiomyopathy Apical, Long Axis, Three Chamber View

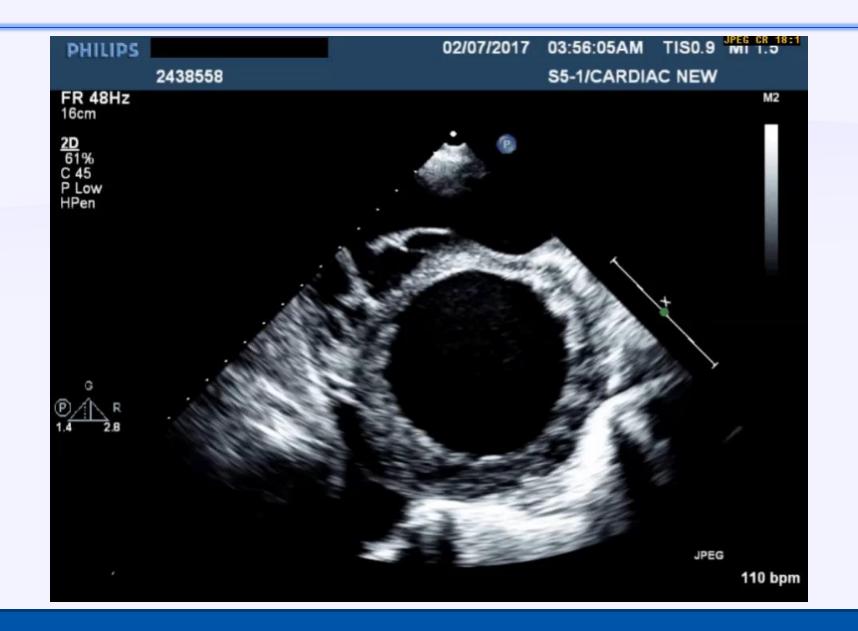


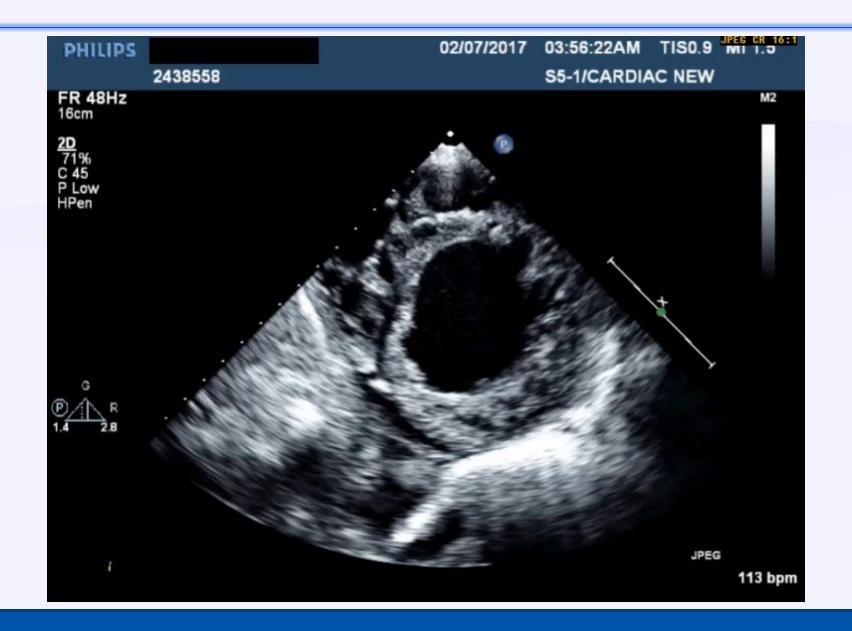
Case 1

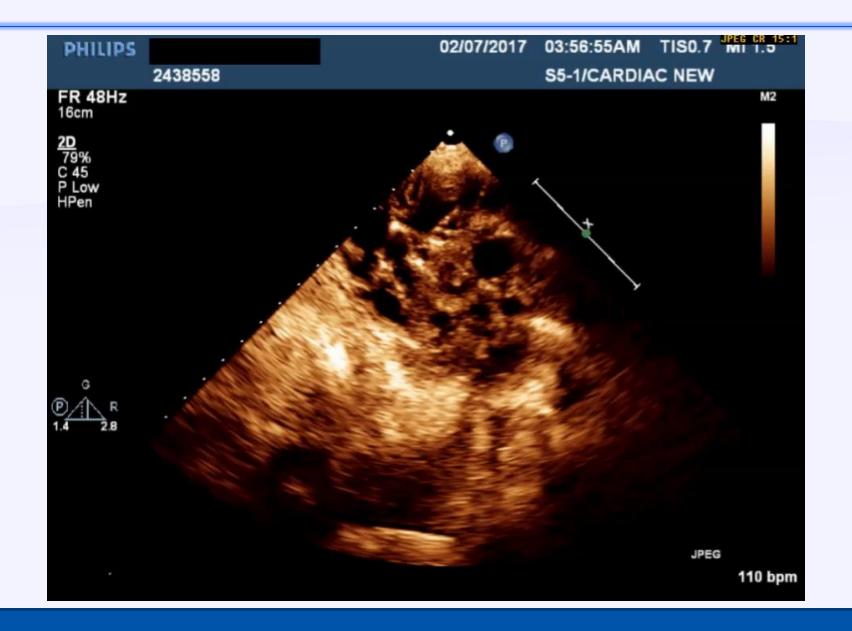
29-year-old man presented with SOB. Echocardiography was requested to rule out cardiac tamponade



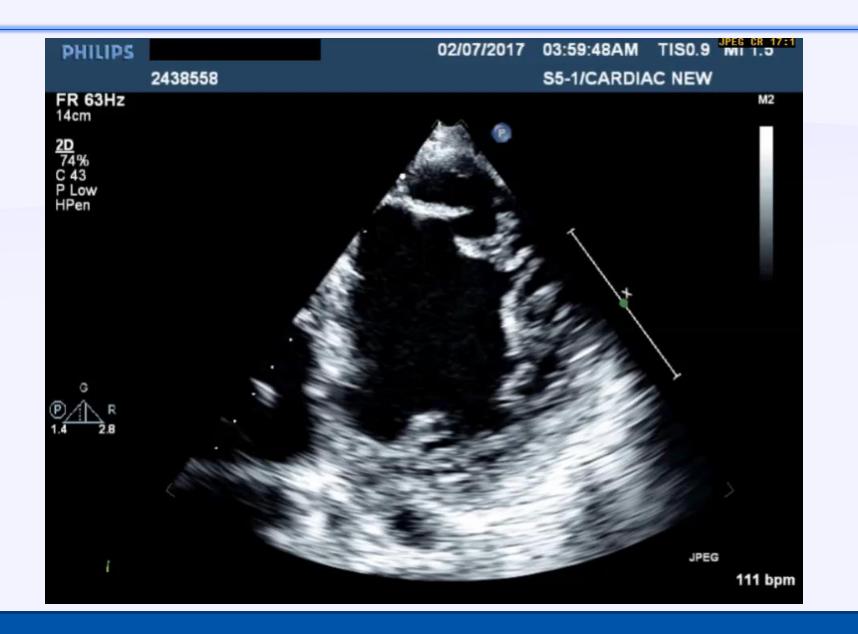


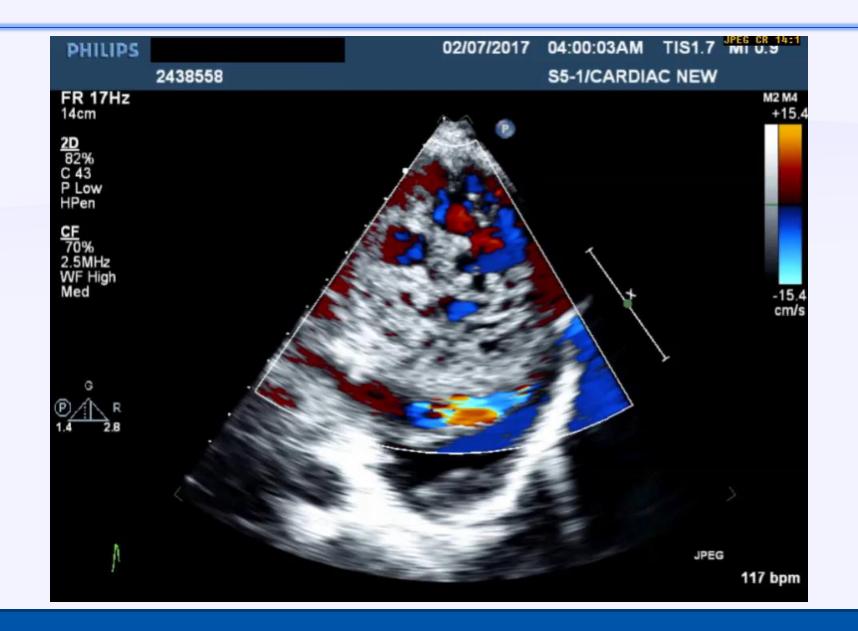




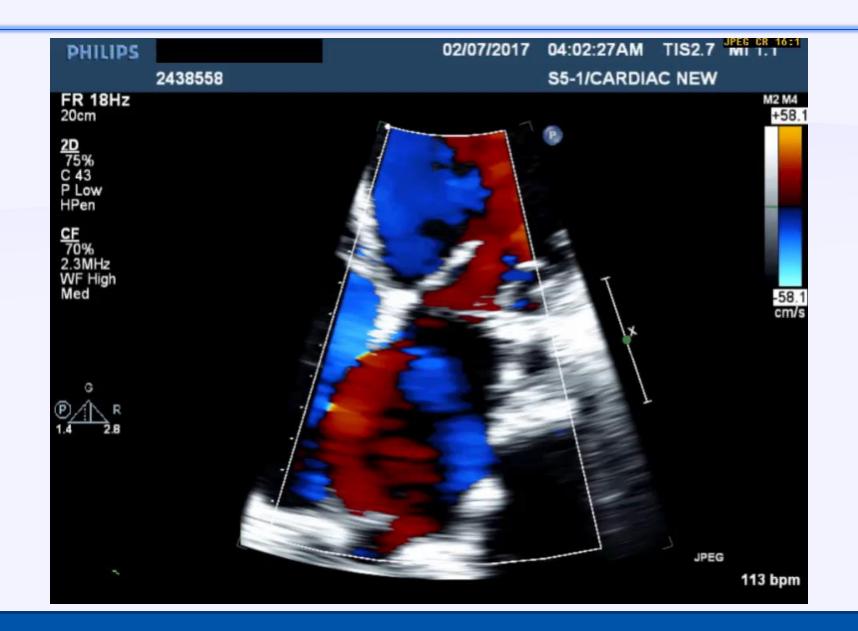


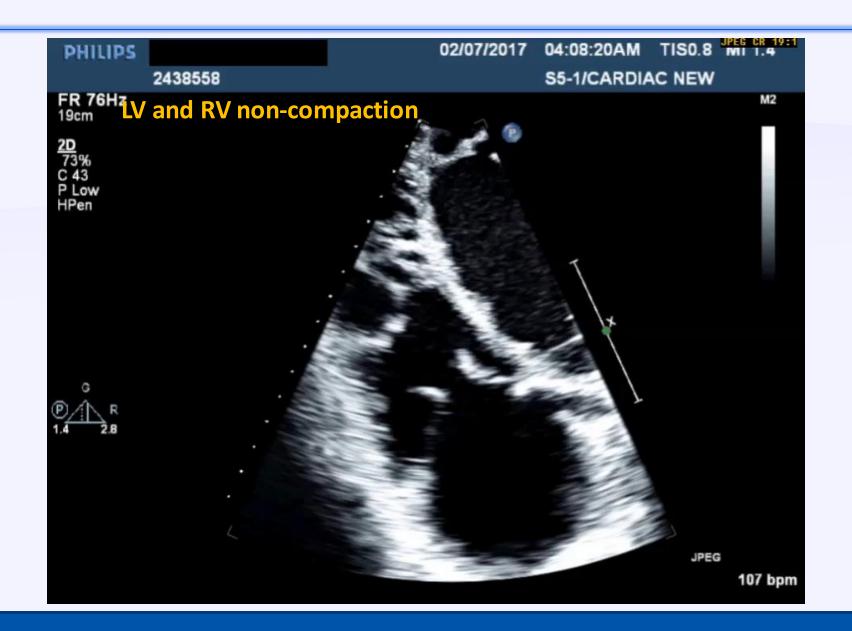


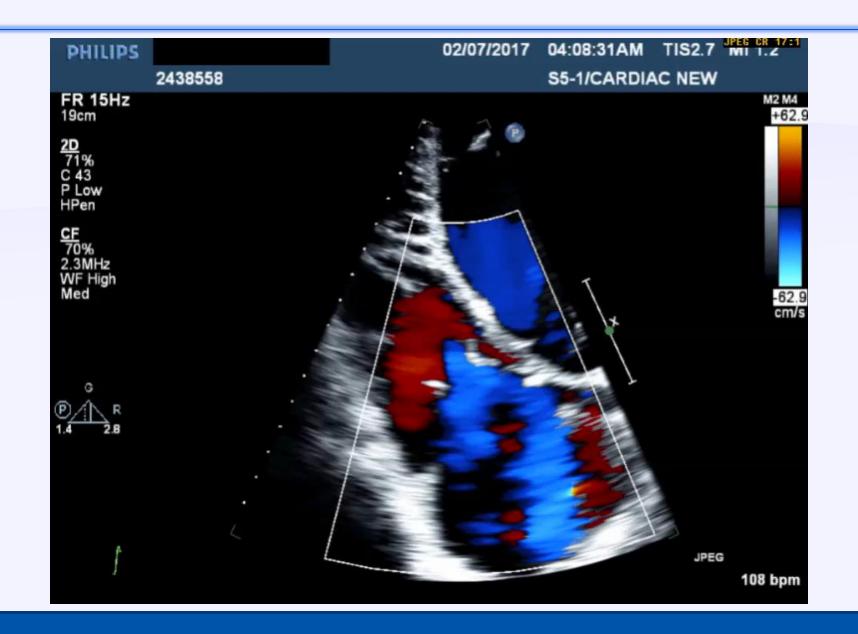


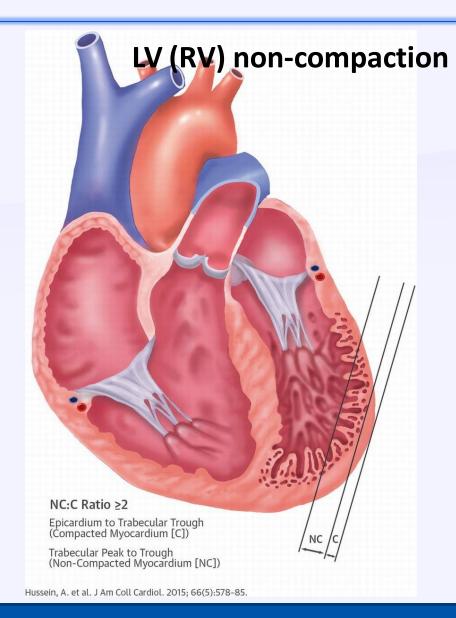










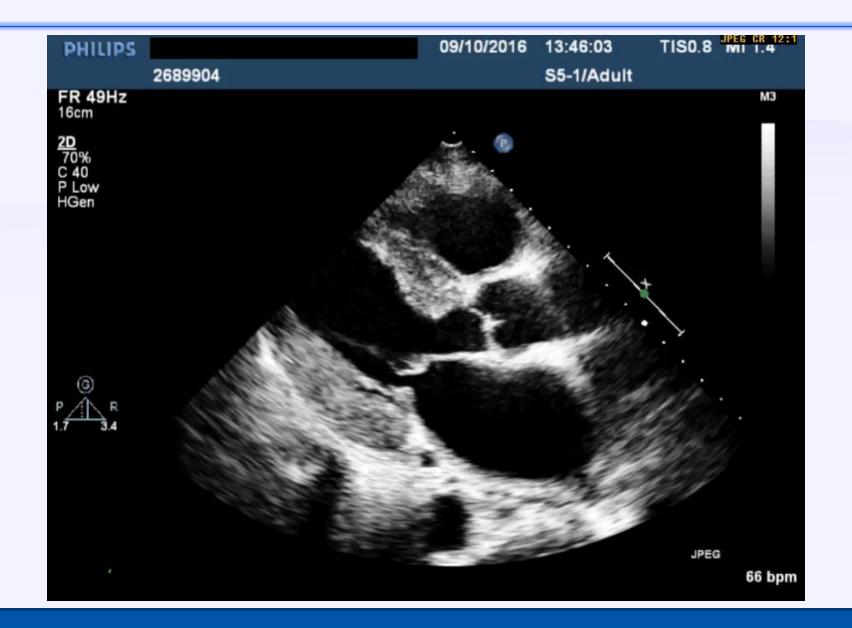


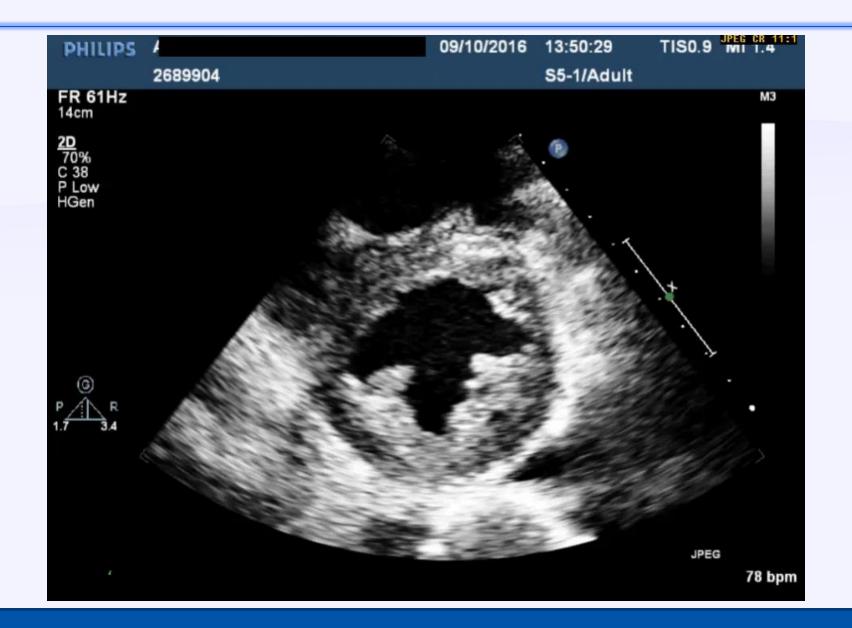


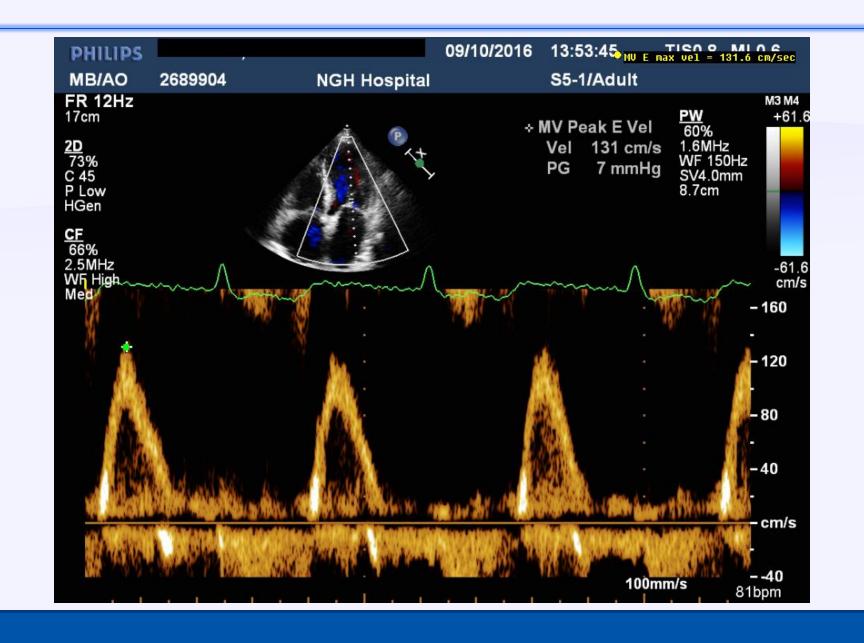


Case 2

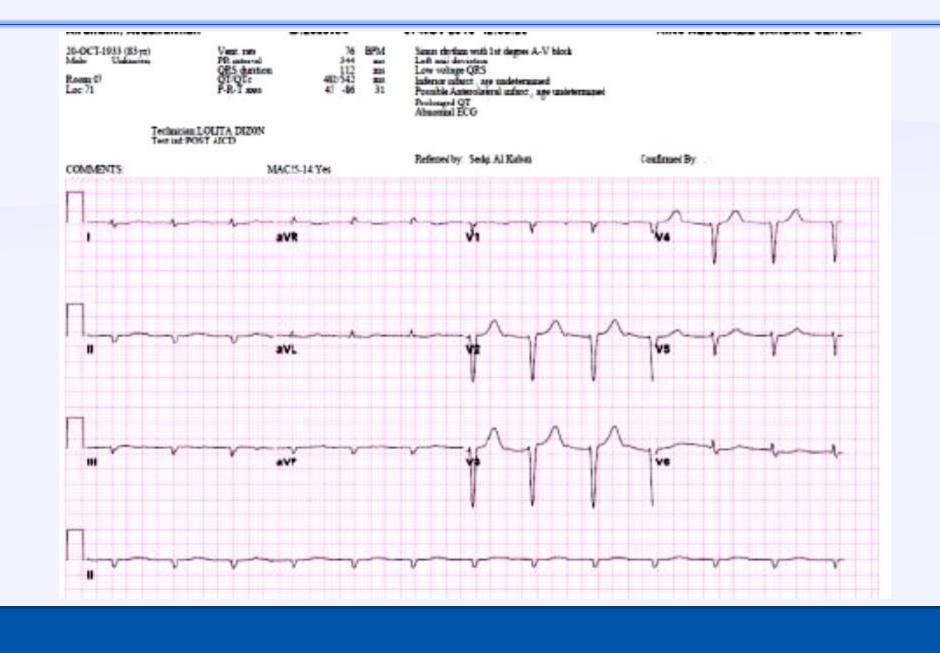
➤ 82-year-old man presented to our center due to severe SOB and atrial fibrillation. No history of hypertension

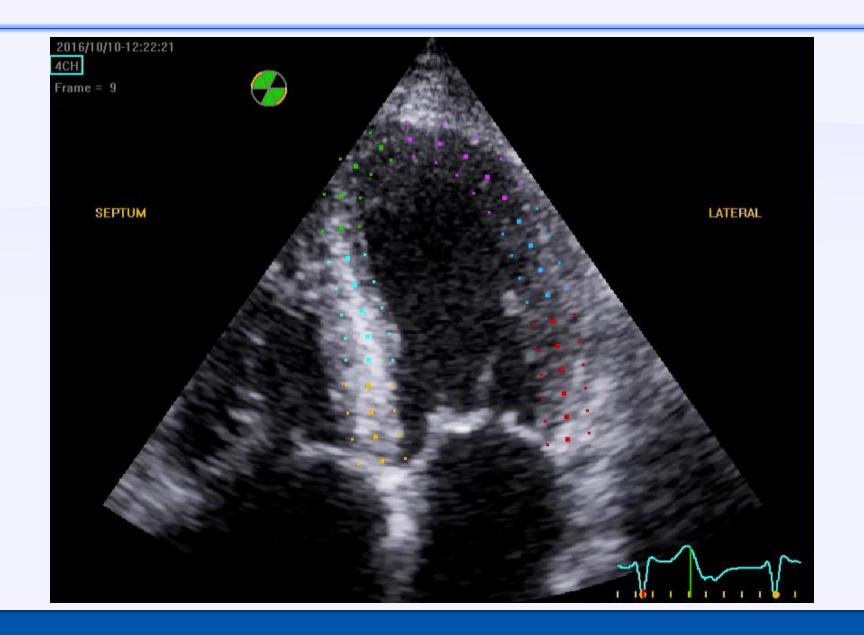


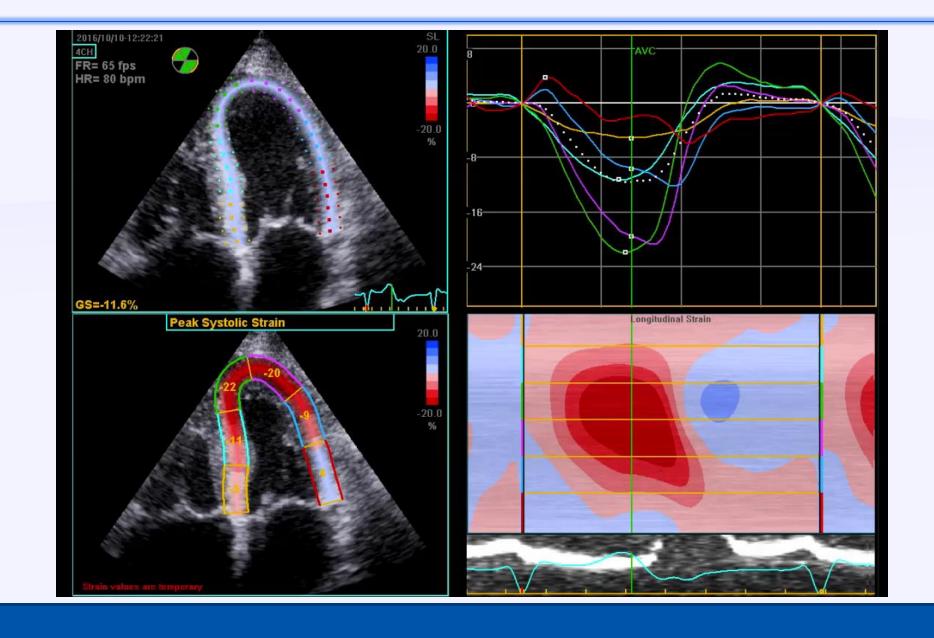


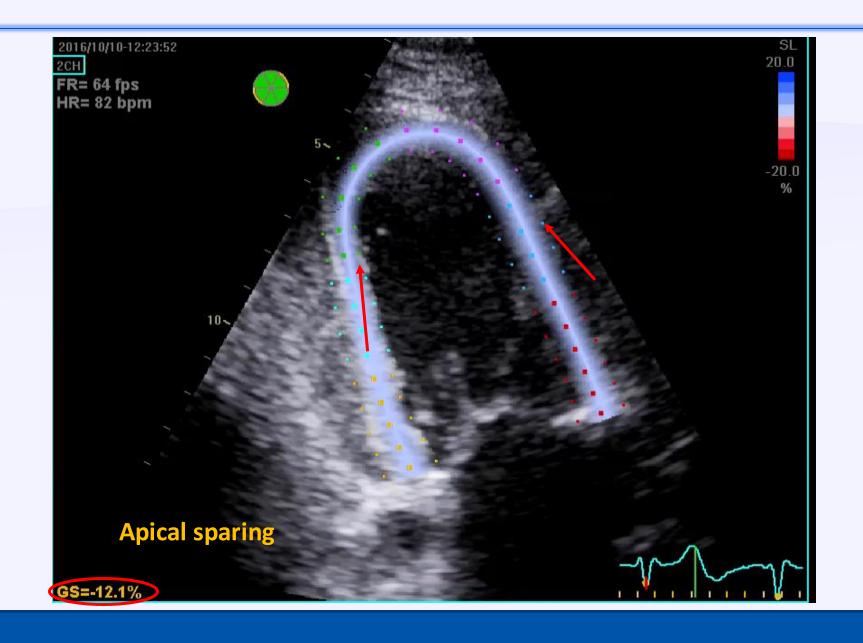


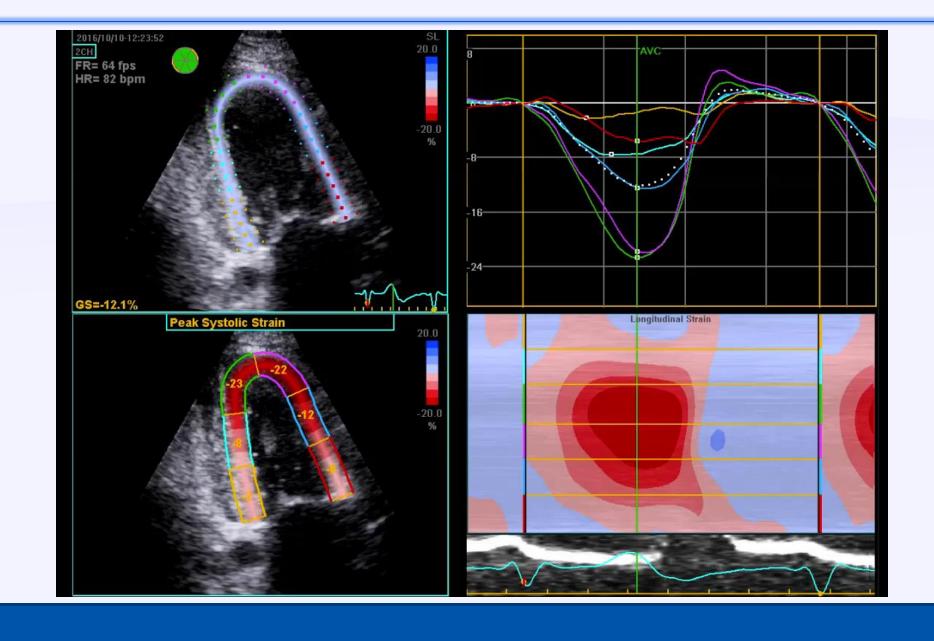


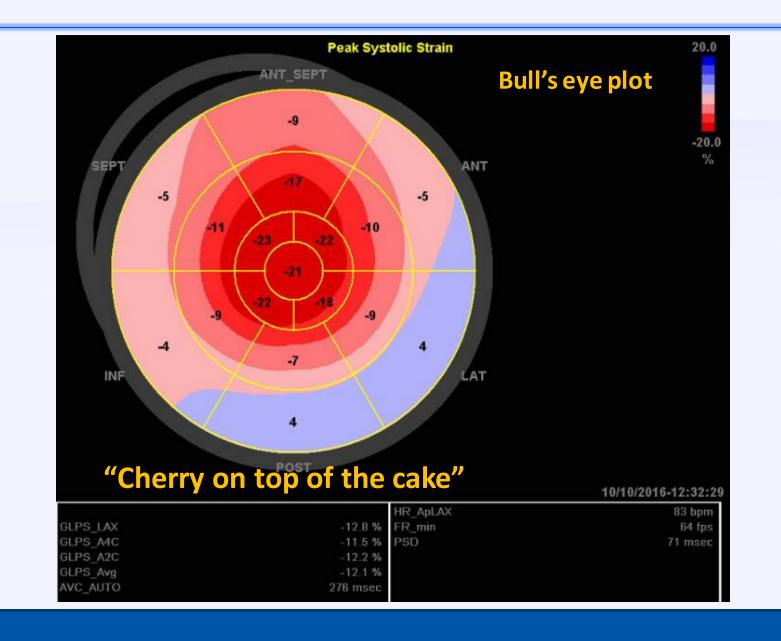


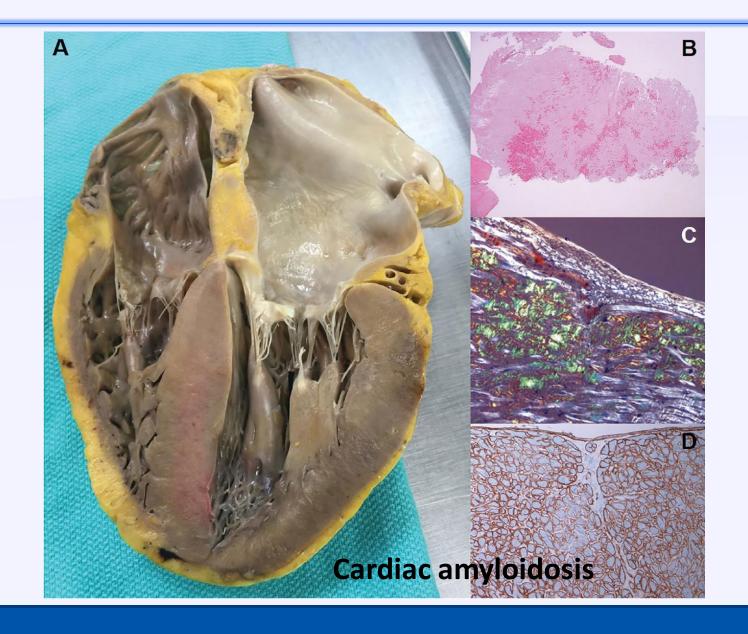




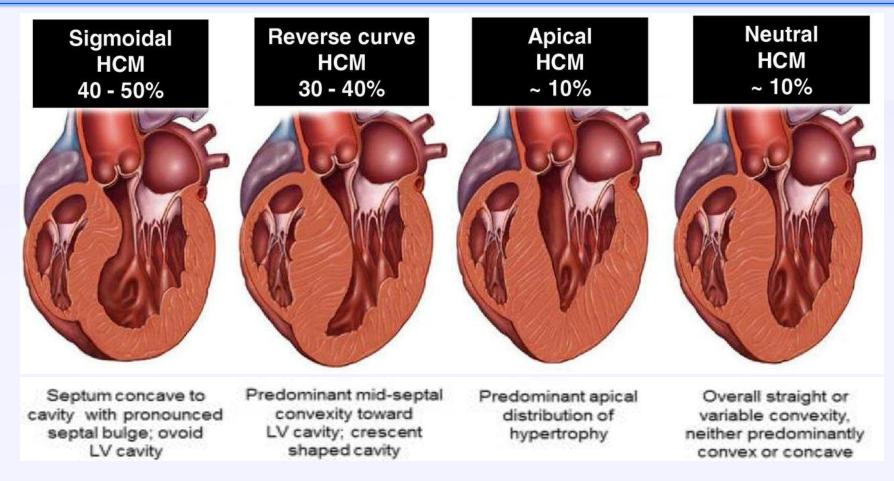








Hypertrophic cardiomyopathy (HCM)



HCM is a common genetic cardiovascular disease involving myocardium. HCM is affecting 1 in 500 general population which is equivalent to at least 600,000 (? 60,000 in Canada).

Isolated septal myectomy for hypertrophic obstructive cardiomyopathy: an update on the Toronto General Hospital experience

Rachel D. Vanderlaan¹, Anna Woo², Anthony Ralph-Edwards^{1,3}

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Background: Isolated septal myectomy is considered the gold standard for refractory left ventricular outflow tract (LVOT) obstruction at centers with dedicated hypertrophic obstructive cardiomyopathy (HOCM) surgeons. In this paper, we provide an update on the Toronto General Hospital (TGH) experience for isolated septal myectomy and comment on the safety and efficacy of myectomy in patients with thin basal septal thickness at our institution.

Methods: We retrospectively reviewed all patients undergoing surgical myectomy at our institution from January 2012 to August 2016. We analyzed patient characteristics, intraoperative variables, pre- and post-procedural echocardiographic parameters, and key outcomes including post-operative stroke, renal failure, iatrogenic ventricular septal defect (VSD), post-procedure insertion of permanent pacemaker, and mortality. Results: At our institution, 150 isolated septal myectomy surgeries were performed over the study period. Preoperative echocardiography demonstrated an average basal septal thickness of 2.10±0.43 cm with a resting gradient of 67±37 mmHg and a provoked gradient of 89±40 mmHg. Fifty percent of patients had significant systolic anterior motion (SAM) of their mitral valve and 53% had associated moderate to severe mitral regurgitation (MR). Of note, 24% of patients had a thin septum of ≤1.7 cm. Discharge echocardiograms demonstrated significant septal reduction to an average basal septal thickness of 1.04±0.26 (P<0.05), with negligible resting and provokable LVOT gradients. At the time of discharge, none of the patients had significant SAM and only 5.3% of patients had residual greater than mild MR. Patients undergoing isolated myectomy with a thin basal septum had similar outcomes to those with a >1.7 cm septal thickness. In our contemporary cohort, there were no iatrogenic VSDs, 5.3% of patients required a permanent pacemaker and there was one early death.

Table 1 Major ca	ardiac surgical	procedures for	or HOCM I	patients
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Cardiac surgical procedures	Cohort (n=291)
Isolated septal myectomy	150
Myectomy + CABG	65
Myectomy + mitral repair/replacement	17
Myectomy + other	59

HOCM, hypertrophic obstructive cardiomyopathy; CABG, coronary artery bypass grafting.

Table 2 Preoperative characteristics for isolated septal myectomy cohort

Patient preoperative variable	Value (cohort n=150)	
Median age (years)	52 [42–61]	
Sex (male)	93 (62%)	
Median age for females (years)	55 [40–63]	
Median age for males (years)	51 [44–58]	
BSA	2.04 (1.85–2.2)	
NYHA class (III-IV)	98 (65%)	
CCS class (III-IV)	87 (58%)	
Syncope	40 (27%)	
Basal septal thickness ≤1.7 cm	36 (24%)	
Concomitant atrial fibrillation procedure	9 (6%)	
DSA body surface area: NVHA New York Heart Association:		

BSA, body surface area; NYHA, New York Heart Association; CCS, Canadian Cardiovascular Society.

Table 3 Echocardiography parameters			
Parameter	Preoperative Echo	Discharge Echo	
Basal septal thickness (cm)	2.10±0.43	1.04±0.26*	
Resting LVOT gradient (mmHg)	67±38	11±7*	
Provoked LVOT gradient (mmHg)	89±40	13±9*	
Presence of significant SAM	75 (50%)	0 (0%)*	
Moderate or severe mitral regurgitation	79 (53%)	8 (5.3%)*	
Left atrial size (cm)	4.5±0.7	4.2±0.7	
RVSP (mmHg)	33±10	34±10	

^{*,} P<0.05. LVOT, left ventricular outflow tract; SAM, systolic anterior motion; RVSP, right ventricular systolic pressure; Echo, echocardiogram.

Conclusions

Our cohort demonstrates that isolated septal myectomy remains a safe and effective operation that can achieve excellent results, irrespective of basal septal thickness, when performed by experienced surgeons in a dedicated HCM center.



OPEN ACCESS

Review article

Alcohol septal ablation in hypertrophic cardiomyopathy

Juan José Santos Mateo¹, Juan R. Gimeno²*

ABSTRACT

Alcohol septal ablation (ASA) has become an alternative to surgical myectomy in obstructive hypertrophic cardiomyopathy since it was first introduced in 1994 by Sigwart. The procedure alleviates symptoms by producing a limited infarction of the upper interventricular septum, resulting in a decrease in left ventricular outflow tract (LVOT) gradient. The technique has been improved over time and the results are comparable with those of myectomy. Initial concerns about long-term outcomes have been largely resolved. In this review, we discuss indications, technical aspects, clinical results and patient selection to ASA.

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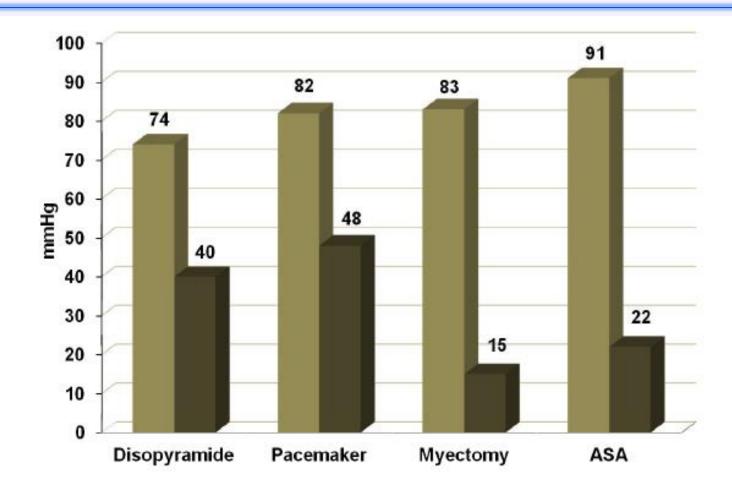


Figure 1. Reductions in the mean resting LVOT gradient in different treatments. Light grey bars represent pre-treatment gradients. Black bars represent post-treatment gradients.

Table 2 Considerations for selection of septal reduction therapies.

Septal myectomy	Septal ablation
Patient choice (immediate results)	Patient choice (less invasive and shorter recovery)
Concomitant cardiac disease	High surgical risk (comorbilities)
Longest follow-up data	Relatively shorter follow-up evidence
Expertise limited to few HCM centres	More reproductive results between centres
Massive hypertrophy	Mild-moderate hypertrophy (16-25 mm)
Mid-ventricular obstruction	
Younger patients	Elderly patients
	Cost of double risk of pacemaker and reinterventions.

Surgical myectomy, history





The Father of Septal Myectomy for Obstructive HCM, Who Also Had HCM

The Unbelievable Story

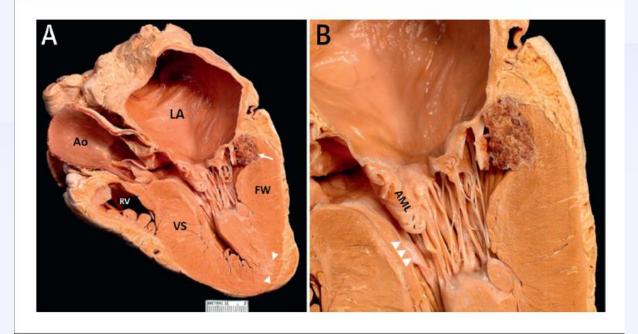
Barry J. Maron, MD,^a William C. Roberts, MD^b

JACC 2016



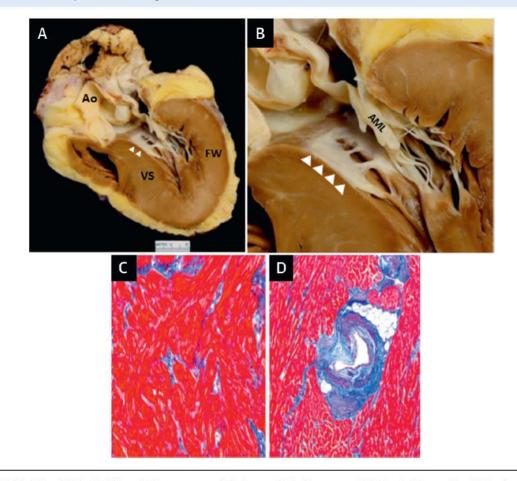
Dr. Andrew Glenn Morrow, chief of cardiac surgery at NIH. He was the first surgeon to perform the surgical myectomy in the early 1960s. He was diagnosed himself to have HOCM at the age of 40 by Dr Eugene Brawnwald. He refused to have surgery or even medical treatment and died suddenly at the age 60.

FIGURE 3 The Heart of Dr. Morrow



Weight, 645 g. (A) The ascending aorta (Ao) is free of atherosclerotic plaque, with a structurally normal aortic valve, nondilated left ventricular (LV) cavity, and greatly dilated left atrium (LA). Ventricular septal (VS) thickness is 20 mm, asymmetrically hypertrophied with respect to LV free wall (FW). Behind posterior mitral leaflet is a large calcific deposit (arrow), frequent in older patients. Several small scars are evident in the VS and at the apex (arrowheads). (B) In the subaortic region of the hypertrophied septum is a fibrous plaque (arrowheads) in apposition to the thickened anterior mitral leaflet (AML), evidence of prior outflow obstruction. RV = right ventricle.

FIGURE 4 The Explanted Heart of Daughter



Weight 510 g. (A) The V5 thickness is 23 mm, asymmetrically hypertrophied with respect to LV FW, and with a nondilated LV cavity. (B) Septal endocardial fibrous plaque (white arrowheads) adjacent to the thickened AML, and prior evidence of mitral valve systolic anterior motion and obstruction. (C) Typical disorganized myocyte arrangement in septum, and areas of interstitial fibrosis (blue); myocardial scarring was absent (×400). (D) Intramural coronary artery with disrupted media and intimal fibrous thickening (×100). Masson trichrome stain. Abbreviations as in Figure 3.

Art of Operative Techniques

Transaortic septal myectomy: techniques and pitfalls

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Hypertrophic cardiomyopathy (HCM) is the most common congenital cardiac disease, affecting up to 1 in 200 individuals. When it causes left ventricular outflow tract (LVOT) obstruction, treatment is indicated to reduce symptoms and the risk of sudden cardiac death. Pharmacologic therapy is the first line treatment, however if it fails, surgical myectomy or percutaneous ablation of the hypertrophic myocardium are the standard therapies to eliminate subaortic obstruction. Both surgical myectomy and percutaneous ablation have been demonstrated as safe and effective treatments; however, myectomy is the gold standard with a significantly lower complication rate and more complete and longstanding reduction of LVOT obstruction.

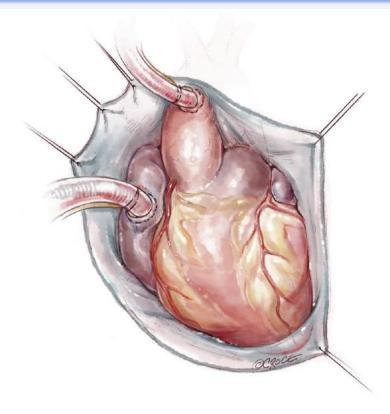


Figure 1 Multiple pericardial sutures are used to suspend the right pericardium to the adjacent superior sternal edge. Left pericardial suspension sutures are placed from the aortic annulus level to the pericardial reflection only, this allows the LV apex to drop into the left chest. This maneuver elevates the aorta to the sternal level and facilitates visualization during the procedure.

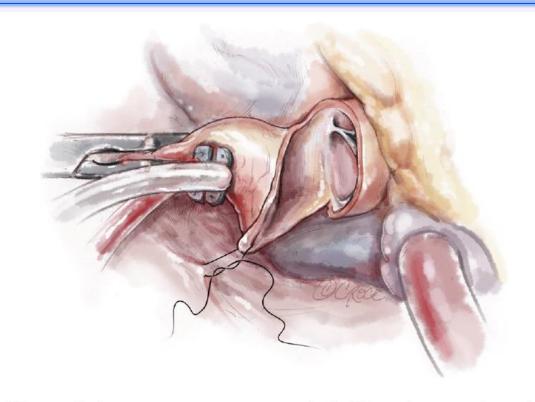


Figure 2 A transverse aortotomy is fashioned approximately 0.5 cm above the sinotubular junction. It is important not to cross the STJ with the incision as this prevents distortion of the aortic valve with closure of the aortotomy, which can result in aortic insufficiency. The distal aorta at the incision line is then tacked to the pericardial reflection at the level of the SVC for exposure.

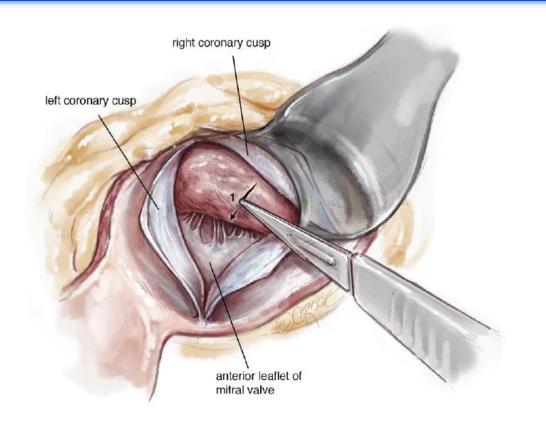


Figure 3 The first myectomy incision is made oriented towards the LV apex beginning 2mm below the right coronary leaflet hinge point and 2 mm towards the membranous septum. The depth of the incision is usually 1-1.5 cm and length 3.5-5 cm as guided by the preoperative echocardiogram.

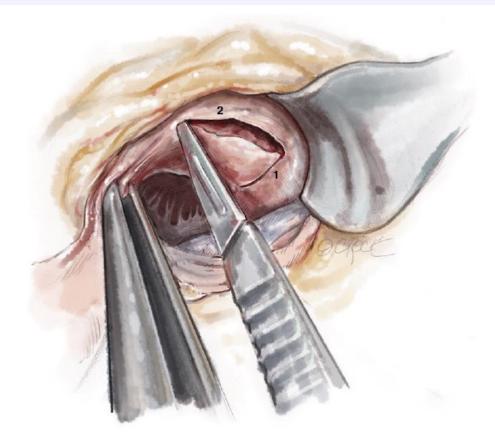


Figure 4 A series of 2.0 mm deep incisions running parallel to the LCC and RCC insertions are created to generate a myocardial flap corresponding to the desired resection thickness. Once the desired thickness has been generated, progress is made into the ventricle.

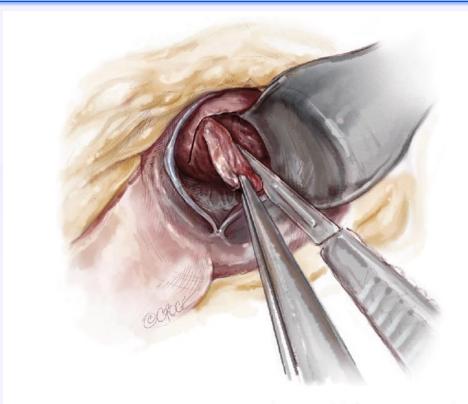


Figure 5 Using Argentine forceps, the 'LVOT flap' is grasped and distracted inferiorly. Now a second 2.0 mm deep incision is made at the superior border of incisions 2 and 3. In the region of the sub-commissural area the two original incisions are rounded with subsequent incisions. The process is repeated eventually forming a flap in the LVOT resection is terminated by placing downward traction on the specimen and incising across the base.

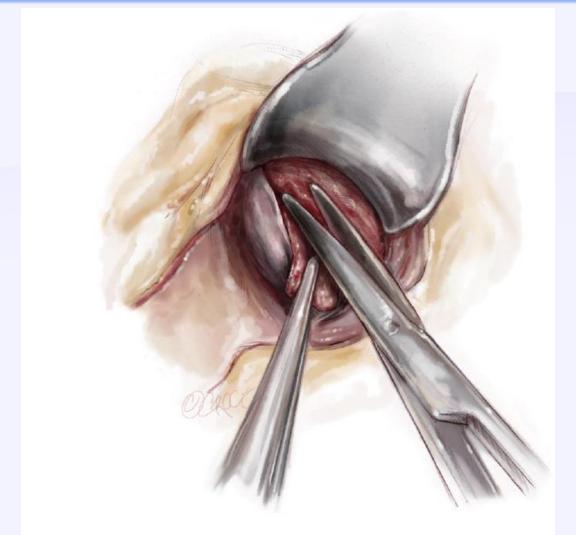


Figure 6 Final distal division made with scissors.

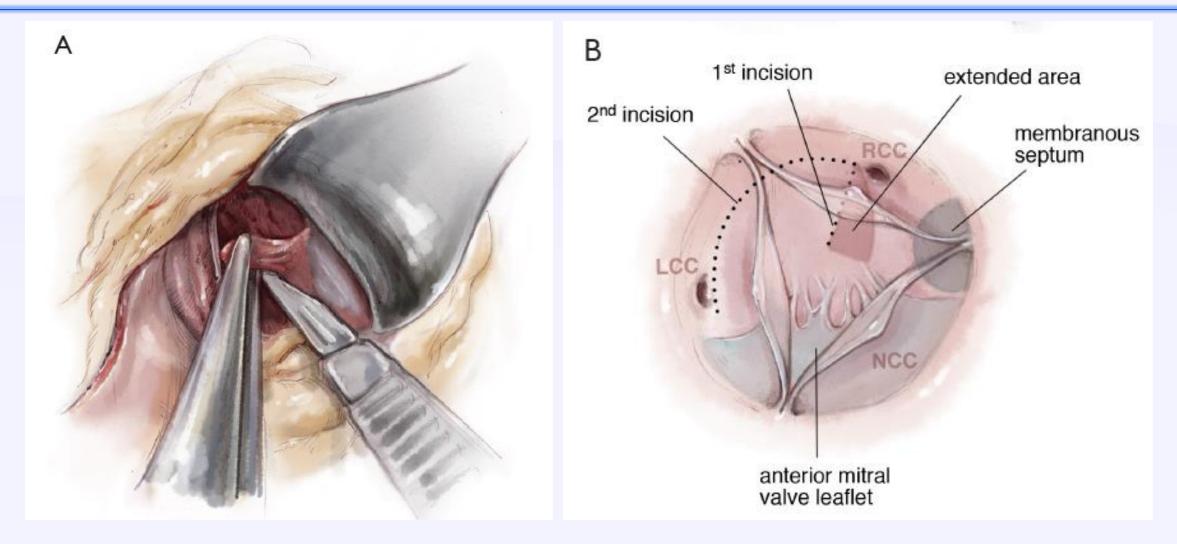


Figure 7 Extension of the resection under the membranous septum.



Figure 8 Completed resection. Note the papillary muscle heads are easily visible at the base of the resection.

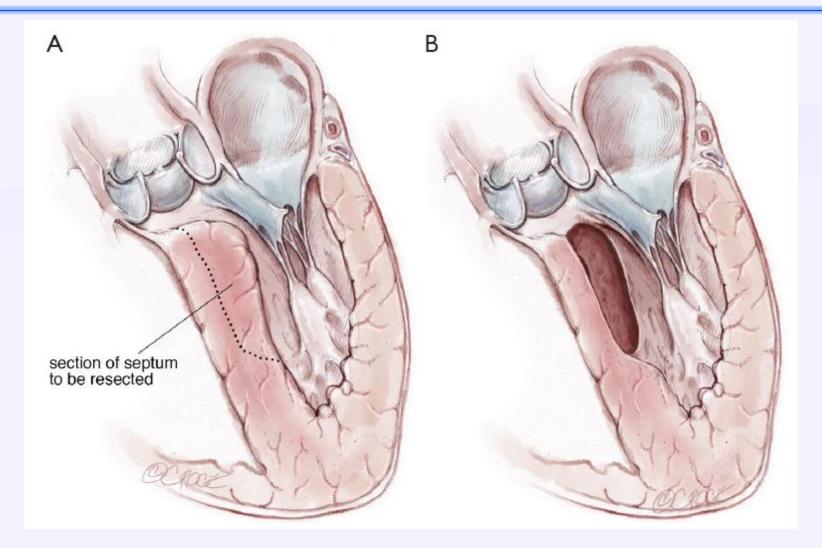


Figure 9 Illustration of septum resected.

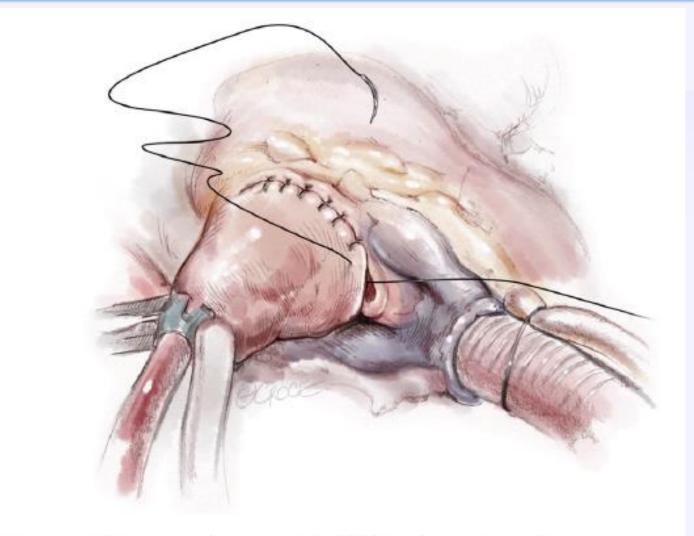


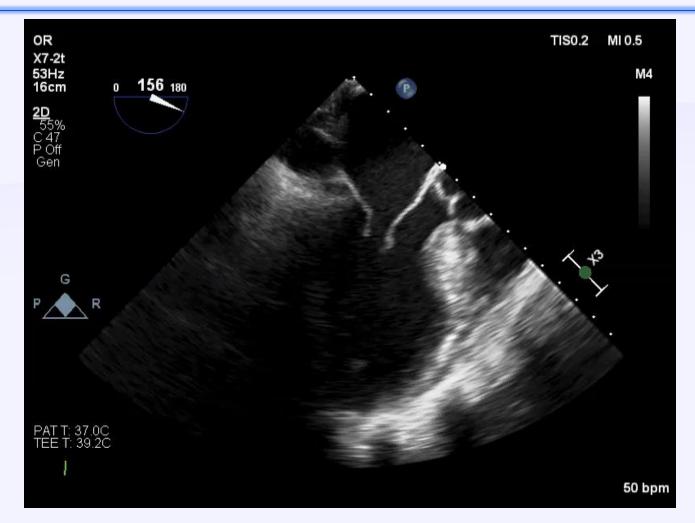
Figure 10 Aortic closure with 5-0 Prolene single layer suture.

Conclusions

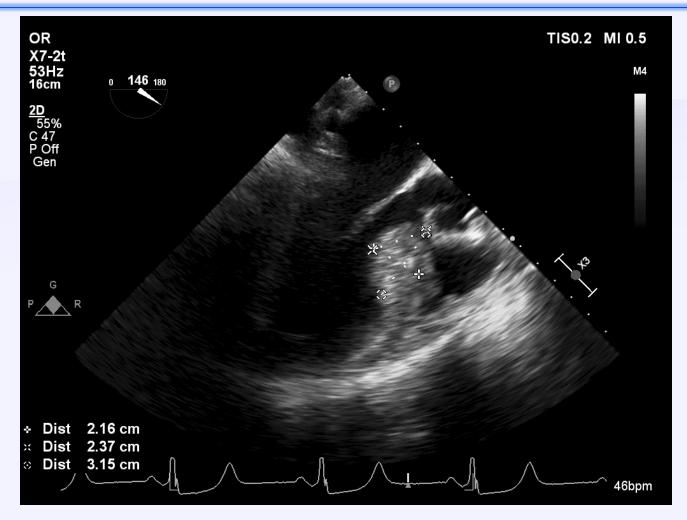
Myectomy is the gold standard for treatment of LVOT obstruction. Excellent results can be obtained at centres with dedicated HCM programs. Careful attention throughout the preoperative, intraoperative and postoperative settings can ensure low complication rates and favourable long-term results.

Case 3

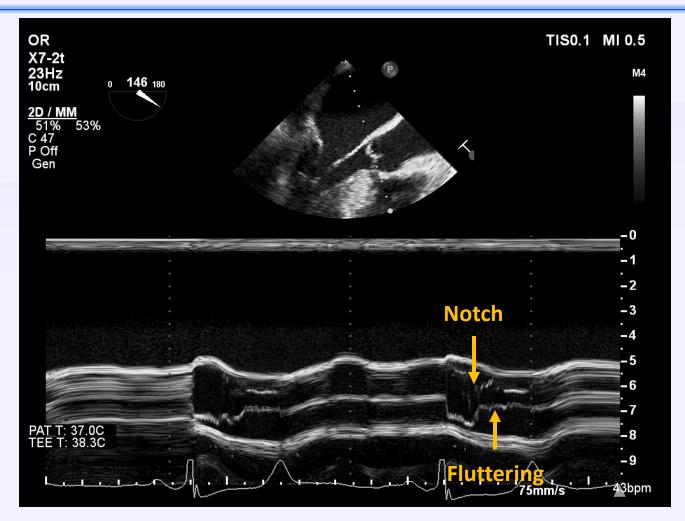
55-year-old woman with a known history of hypertrophic obstructive cardiomyopathy



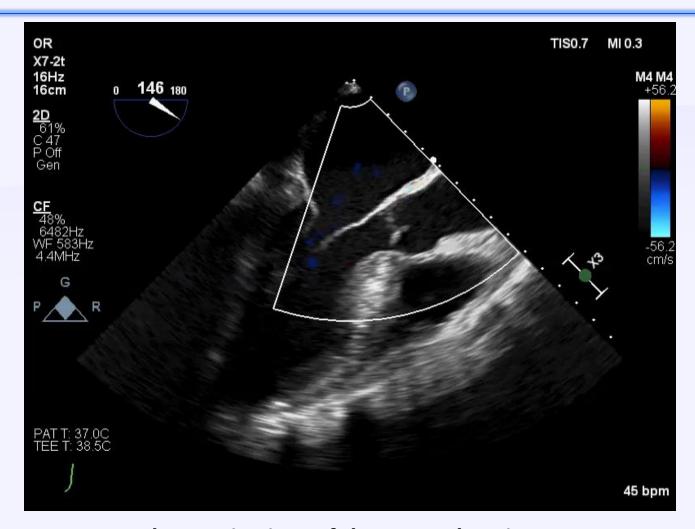
Pre op, long axis view of the LVOT showing severe thickening of the basal septum, severe SAM



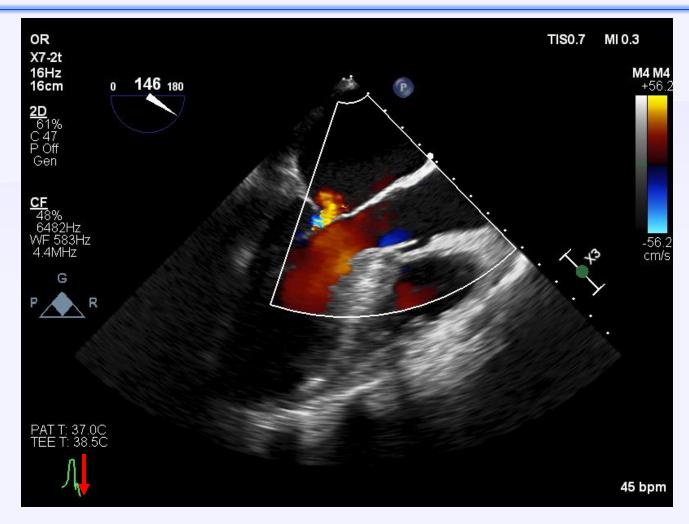
Pre op measurements: maximum septal thickness = 2.2 cm, distance of RCC to the SAM contact = 2.4 cm, and distance of RCC to the downward extension of thickening = 3.2 cm



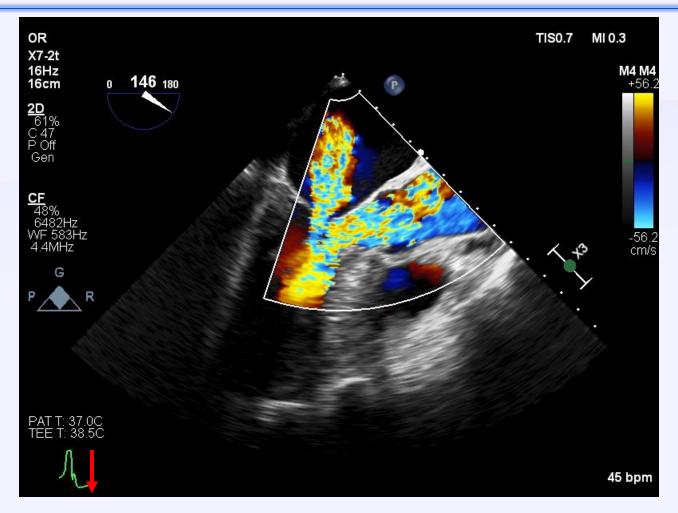
M-mode across the aortic valve showing classic sign of LVOT dynamic obstruction (mid systolic notch and aortic cusps fluttering)



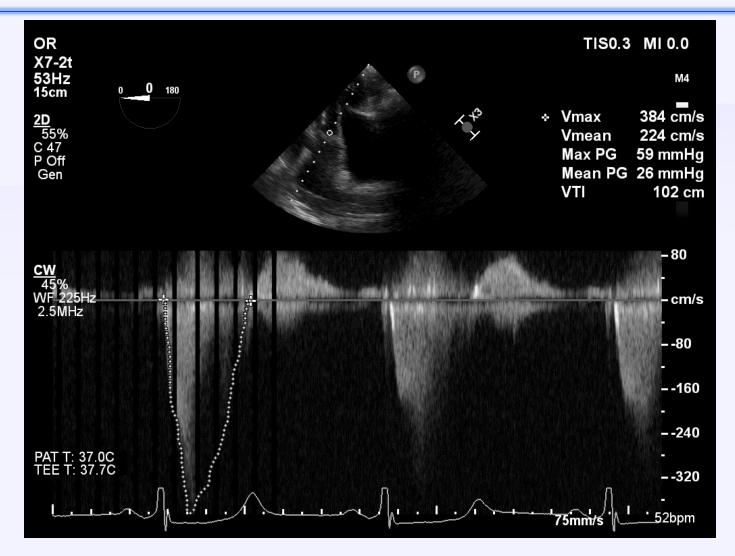
Pre op, long axis view of the LVOT showing severe posteriorly directed jet of MR following SAM



Trace early systolic MR which will remain after myectomy

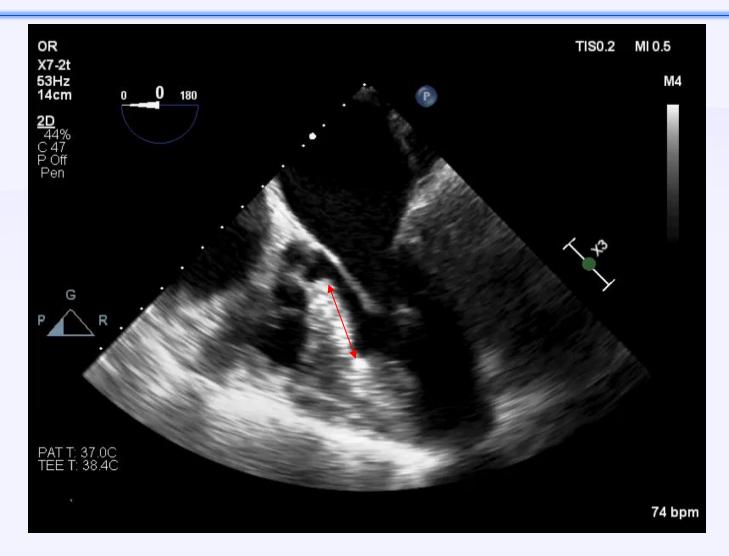


Late systolic MR following SAM which should disappear after a successful myectomy

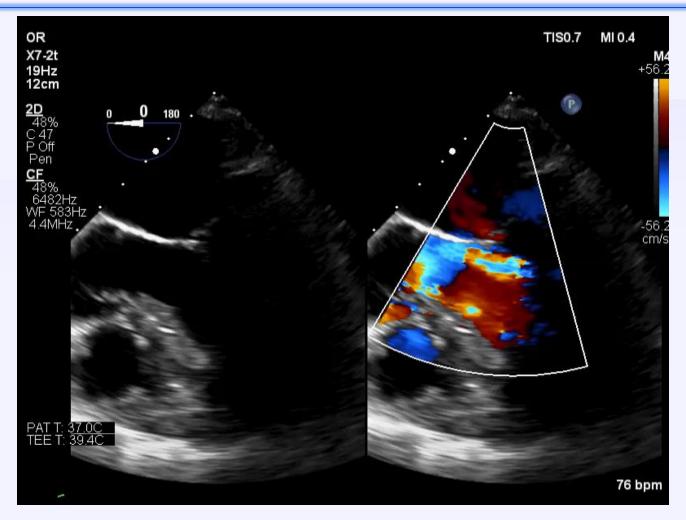


LVOT gradient, PIG = 59 mmHg (contaminated with MR)

Post op



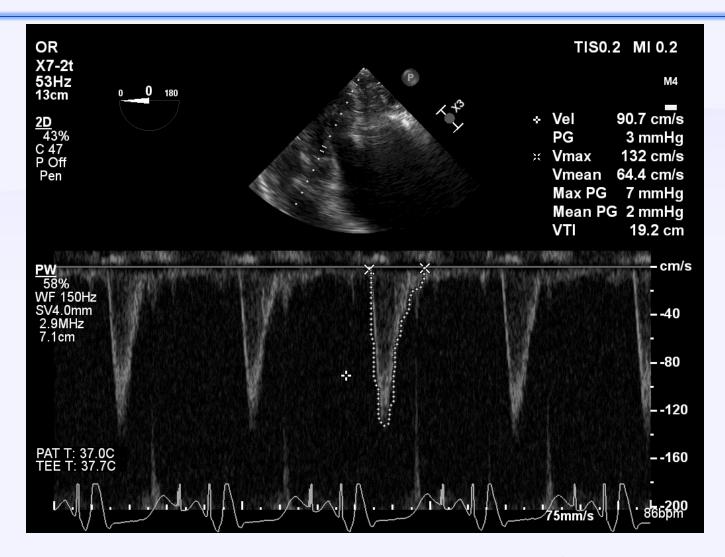
Extensive septal myectomy, very mild SAM



Post op, septal perforator at the site of myectomy, no LVOT systolic turbulence, no VSD, only trivial MR left



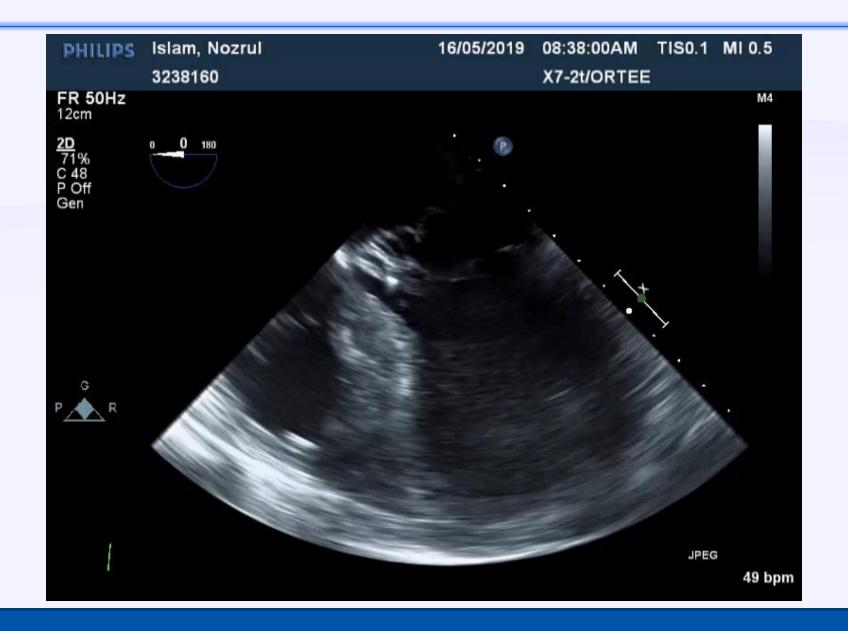
Post op long axis view, trivial early systolic MR

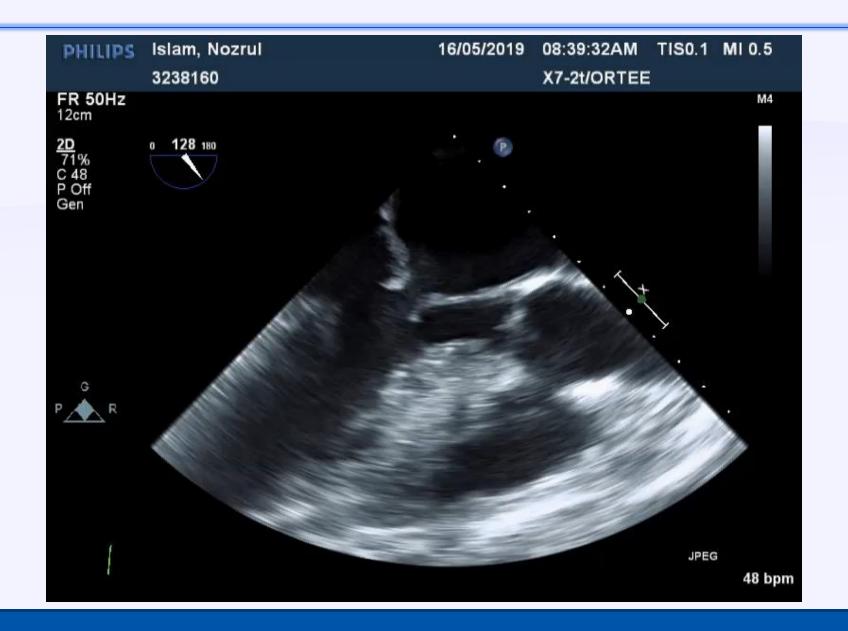


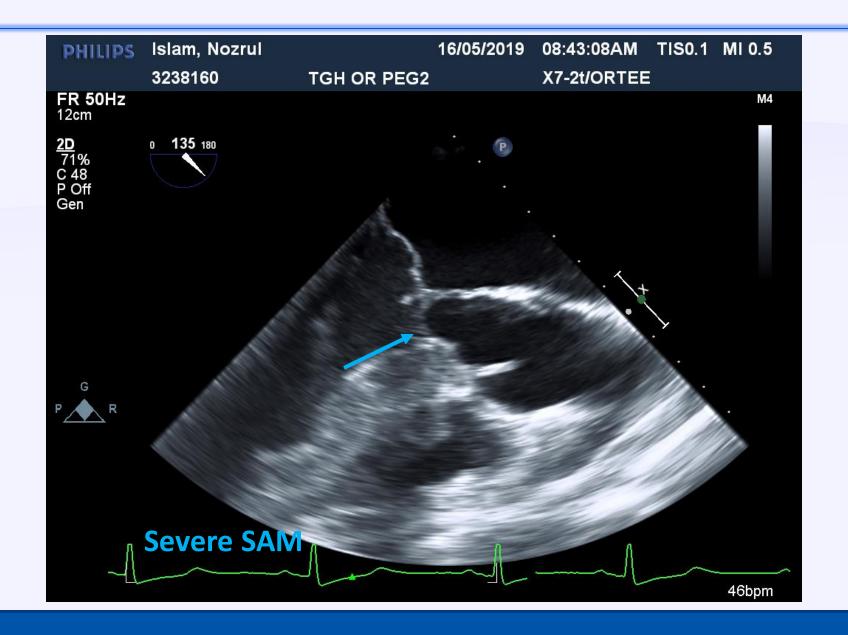
Post op LVOT gradient, PIG = 7, mean of 2 mmHg

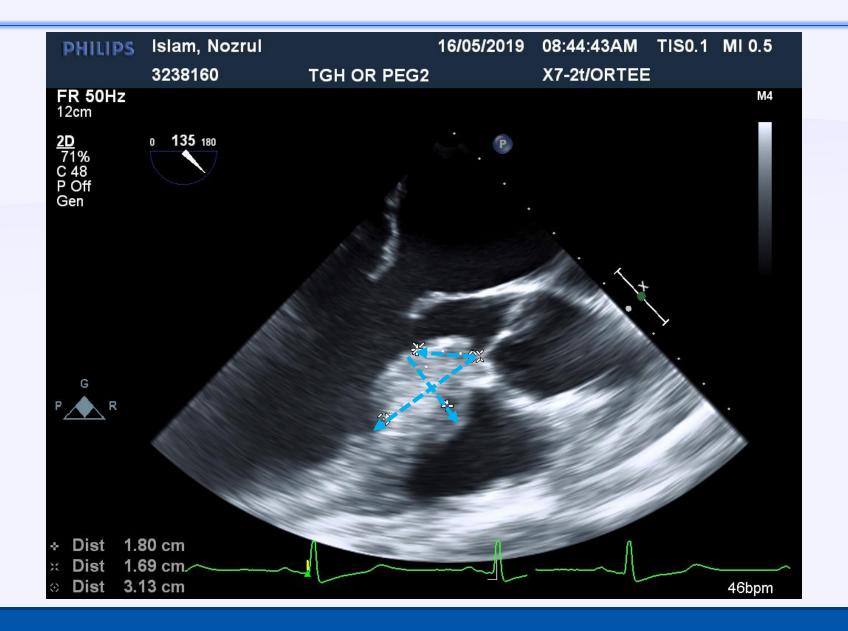
Case 4

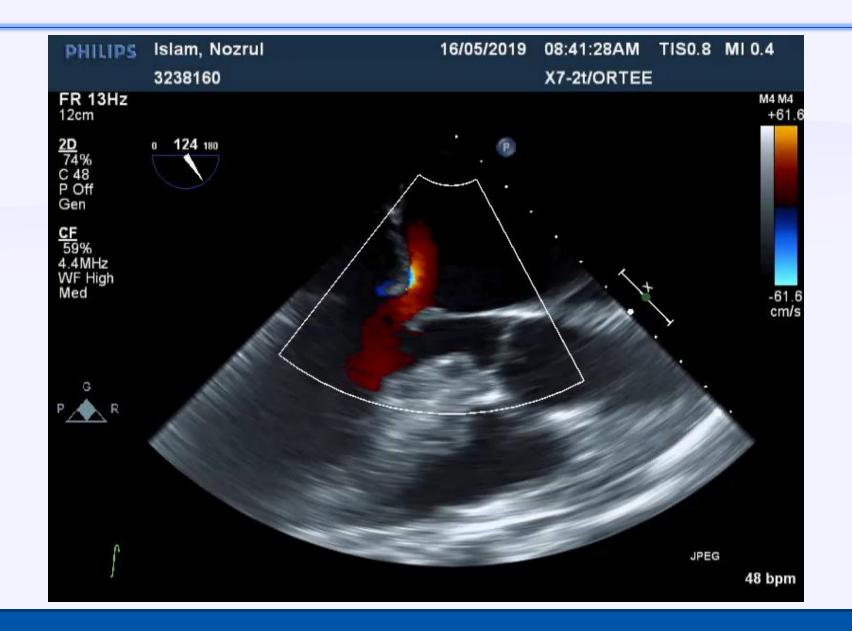
54-year-old man with a known history of hypertrophic obstructive cardiomyopathy

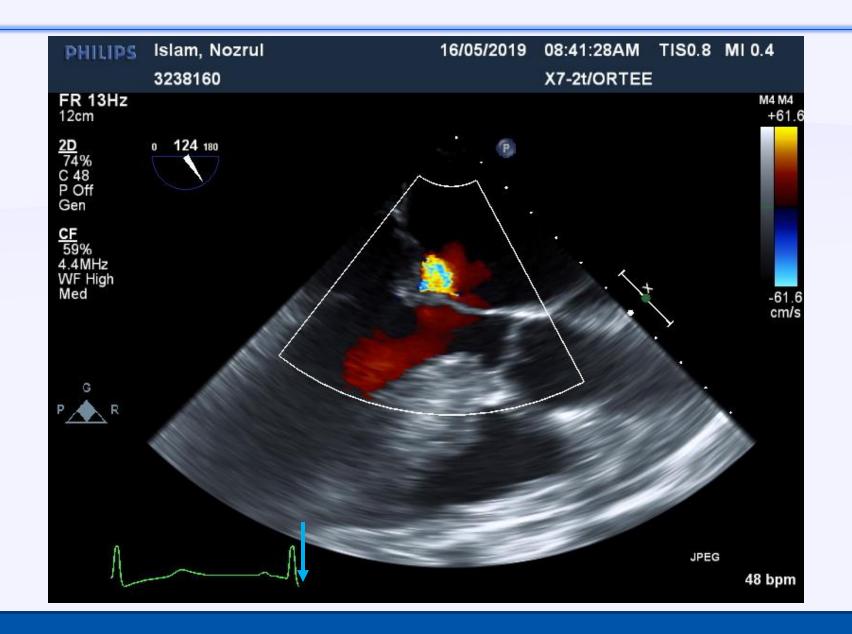


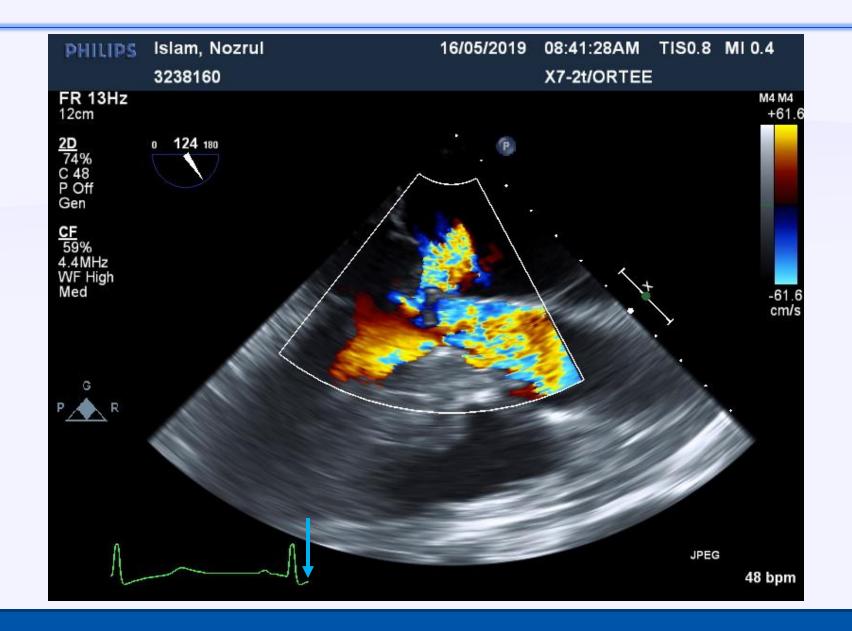


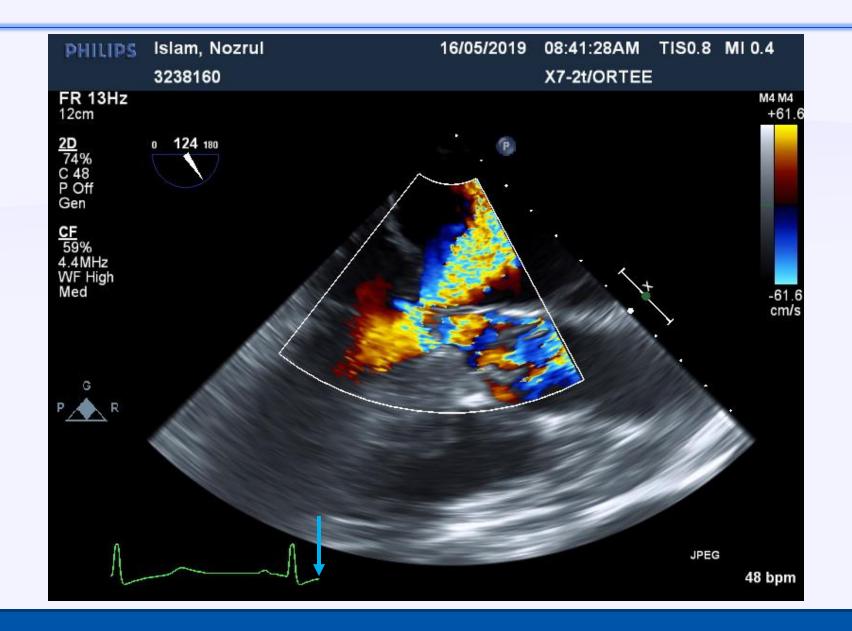


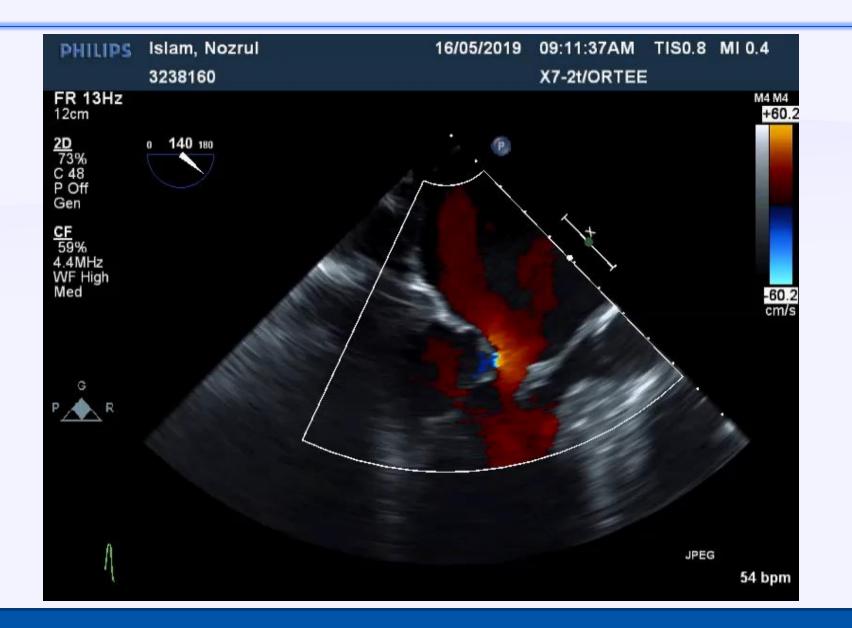


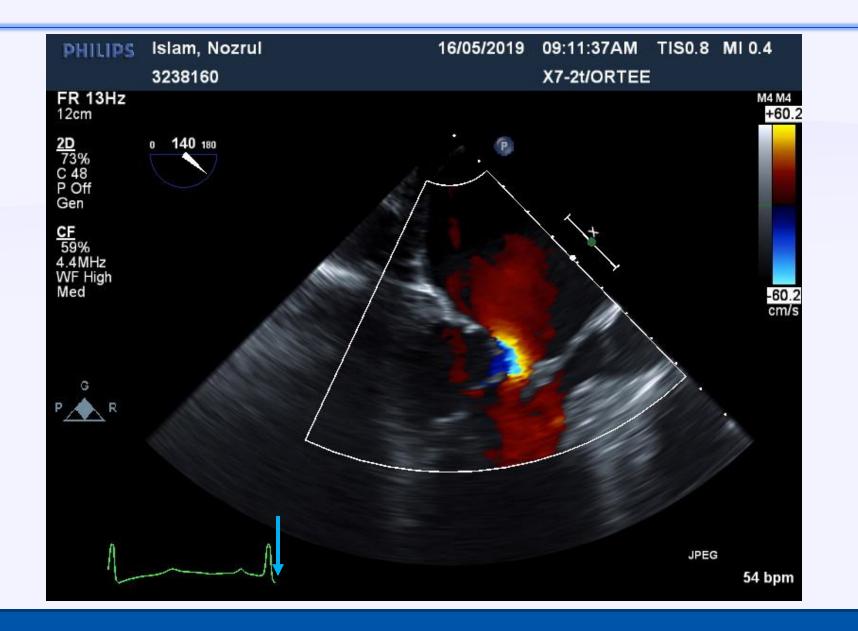


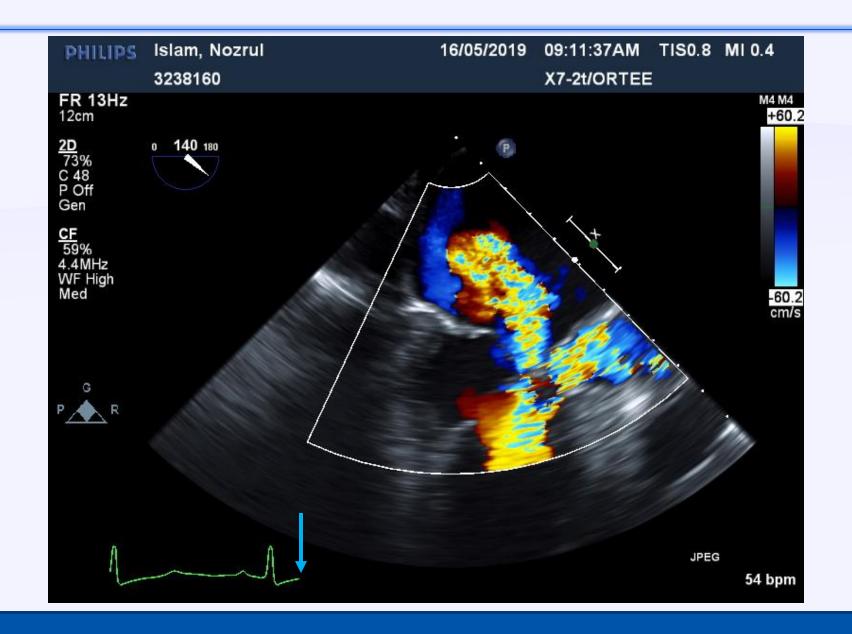


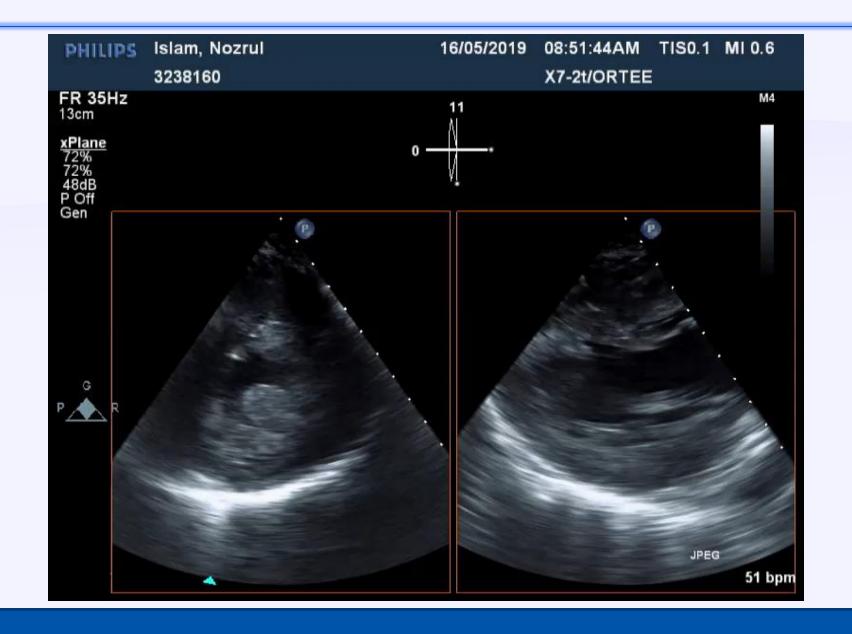


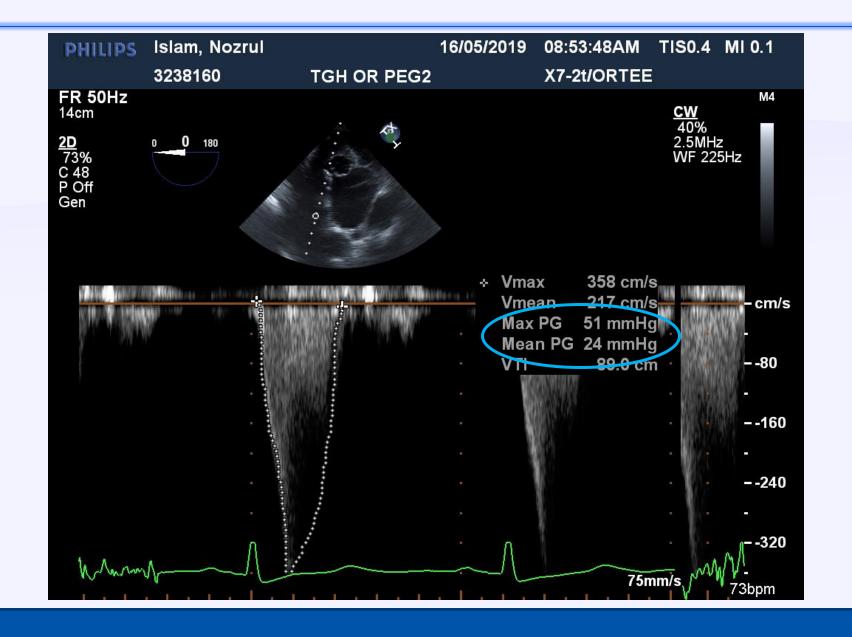




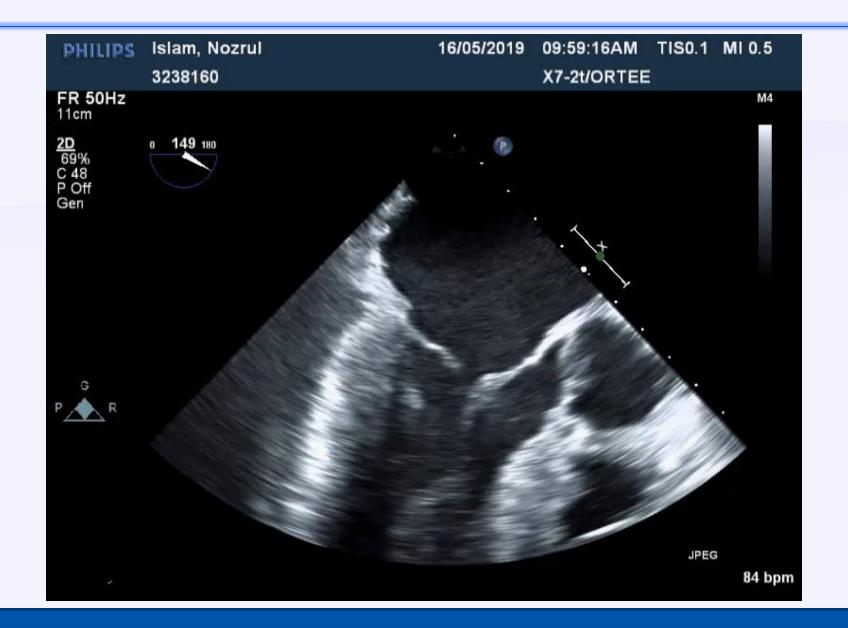


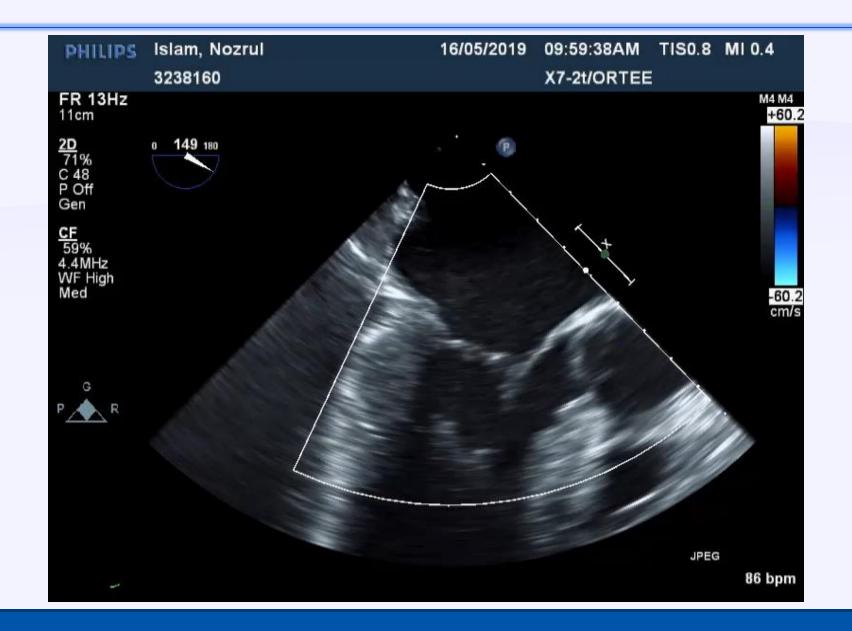


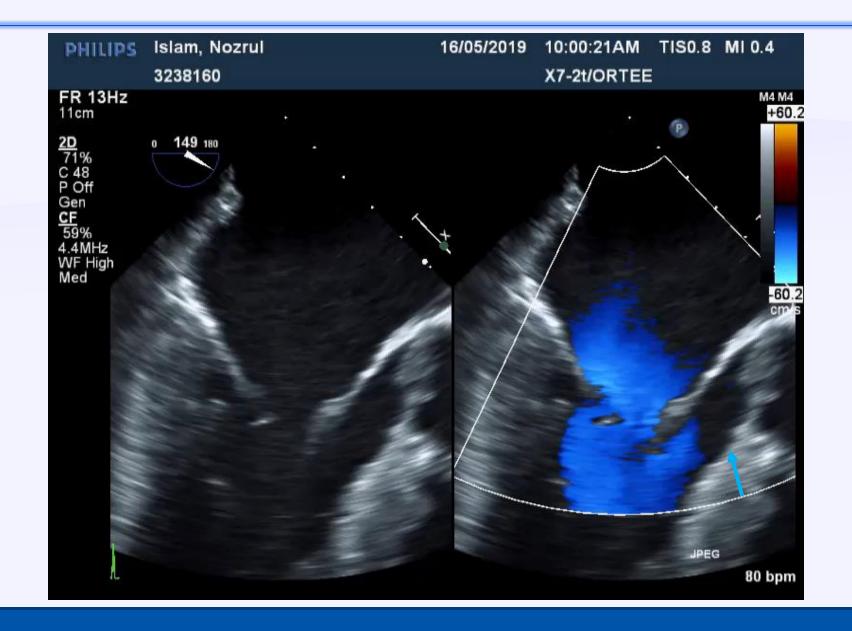


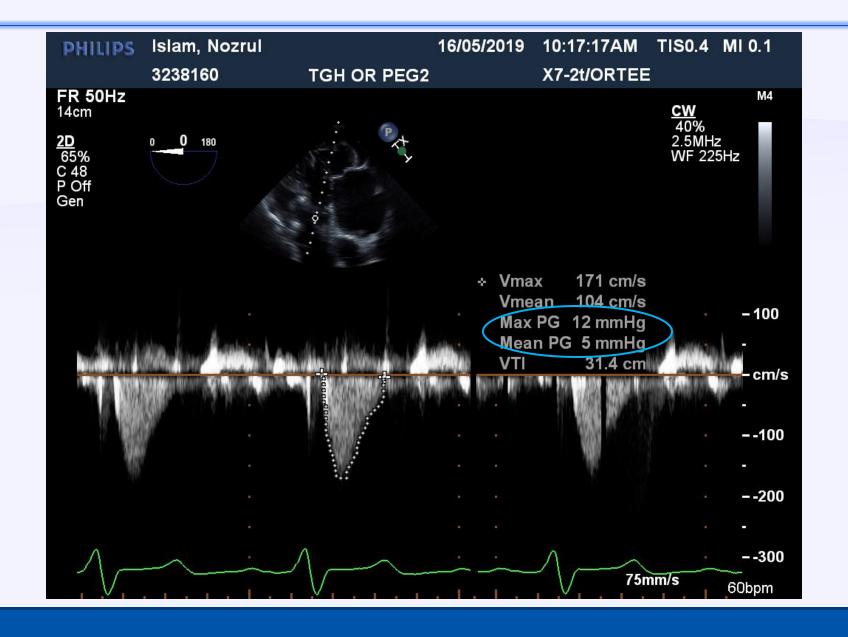


Post op



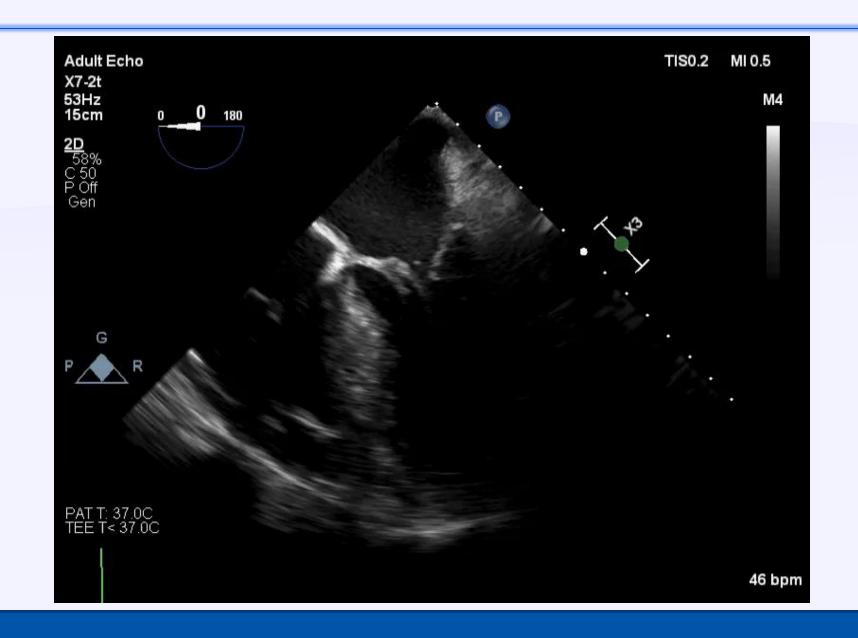


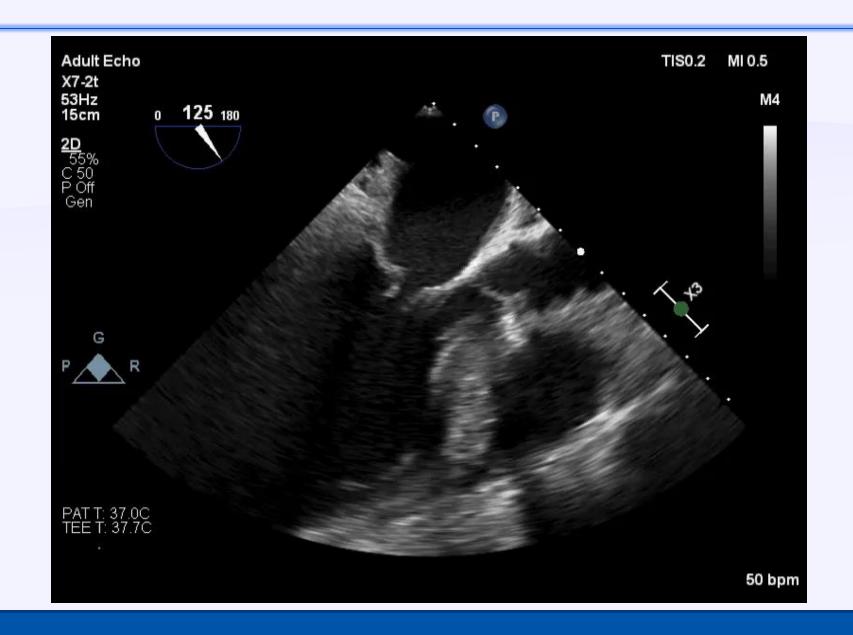


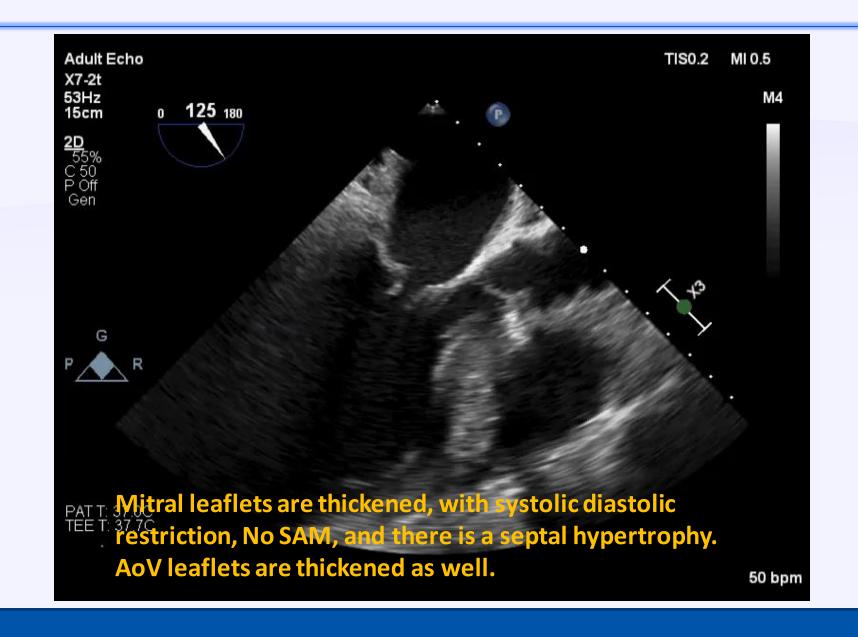


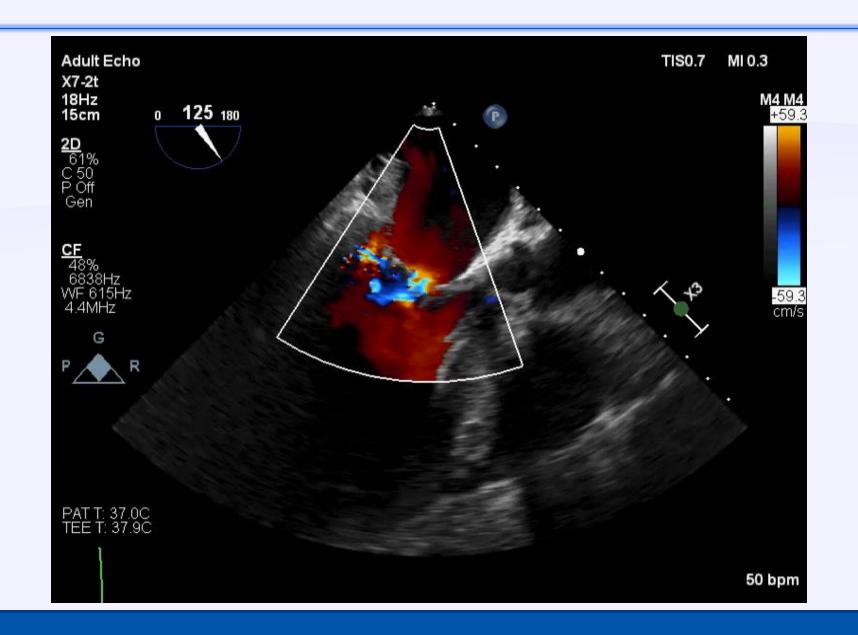
Case 5

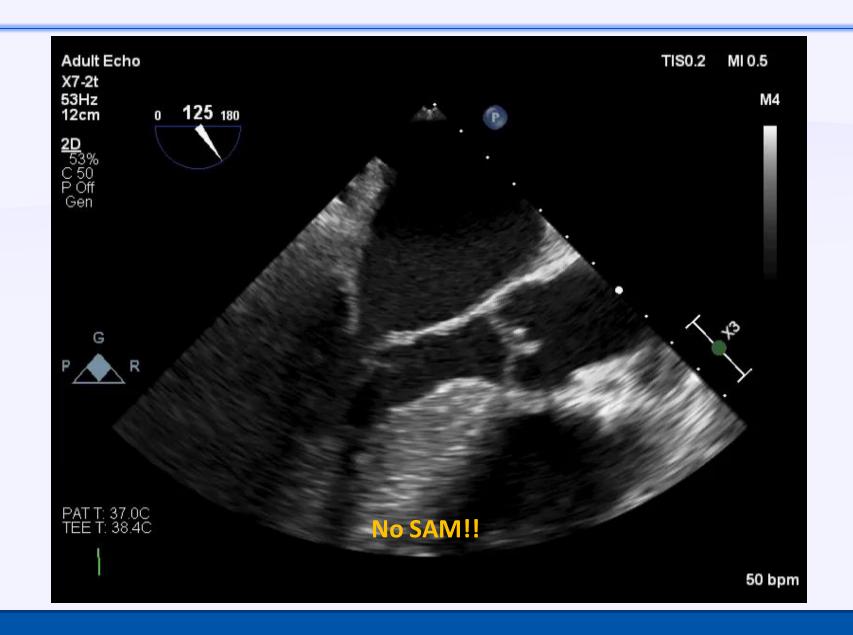
66-year-old woman was referred from BC. for myectomy

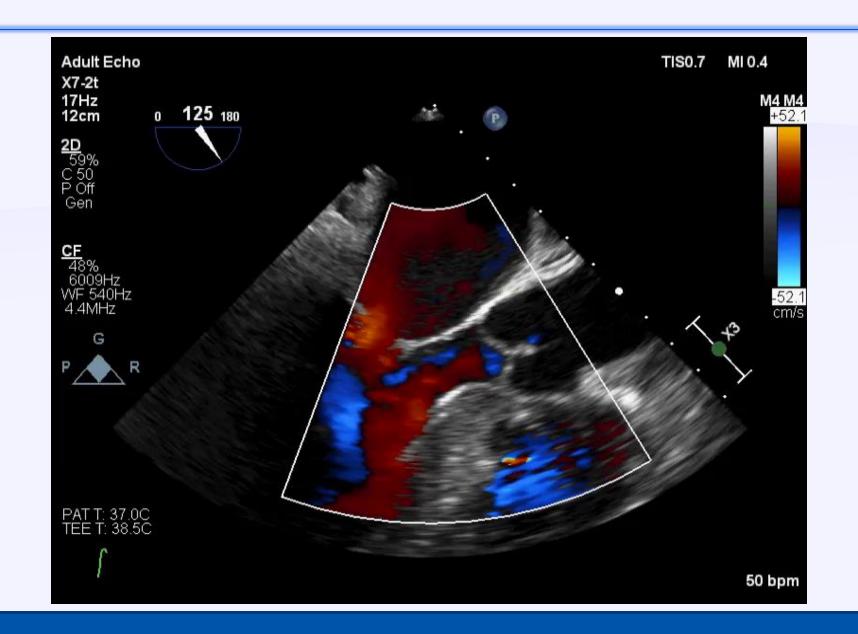


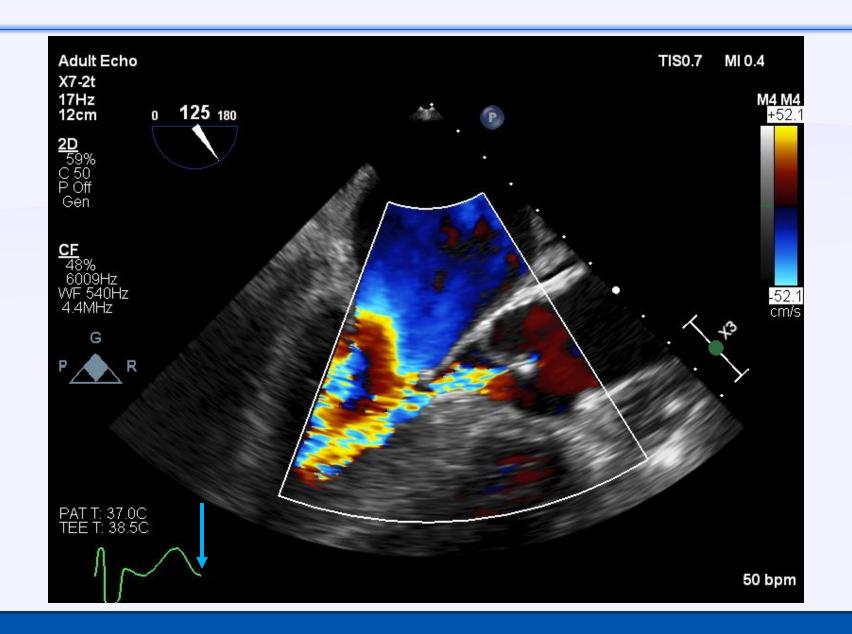


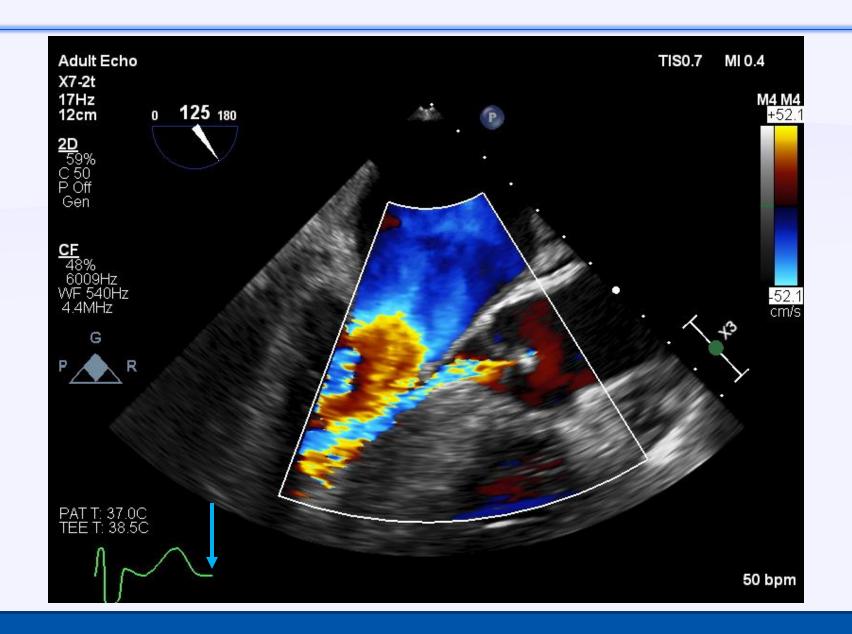


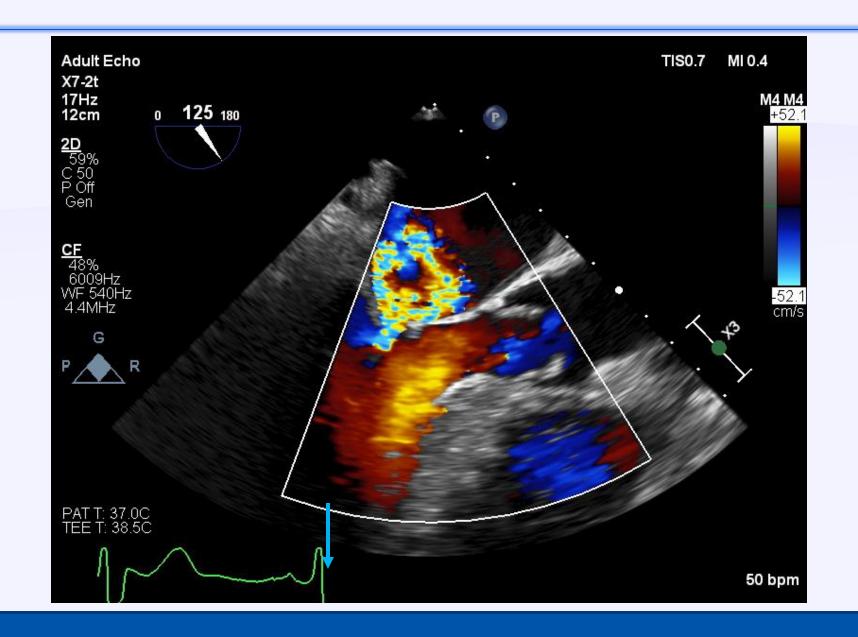


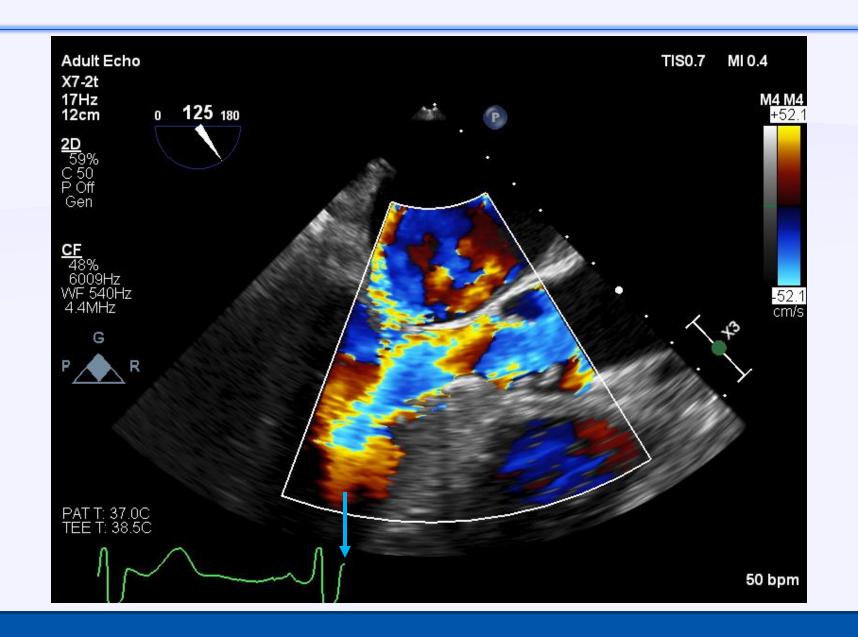


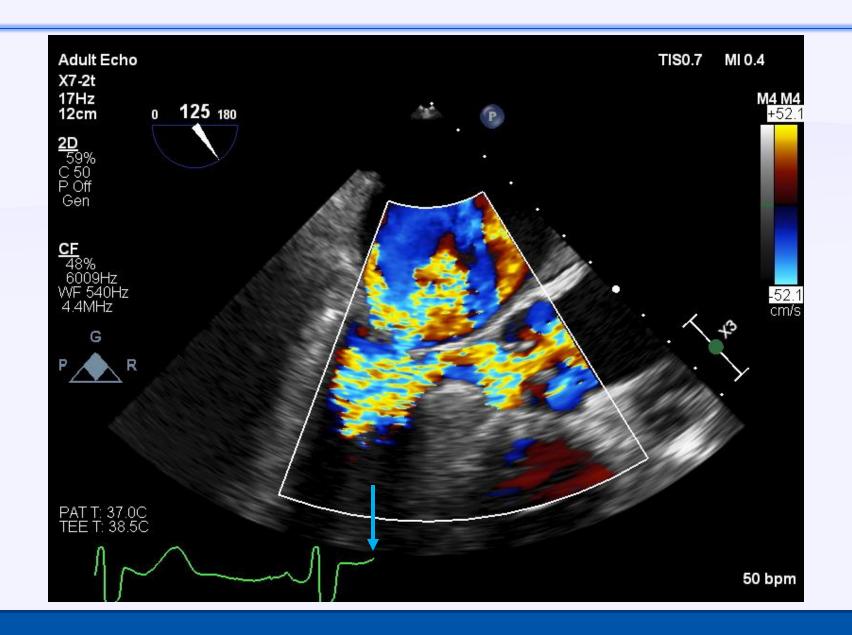


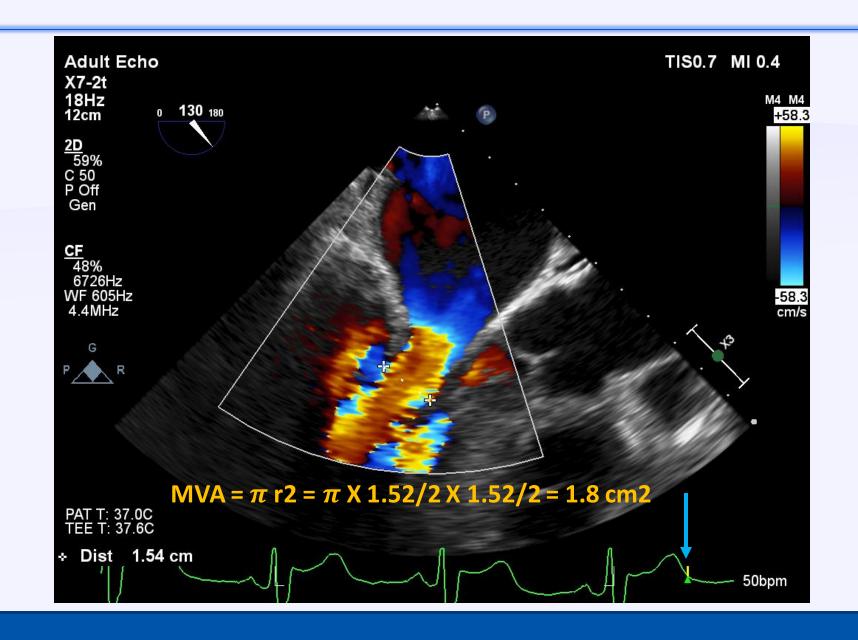


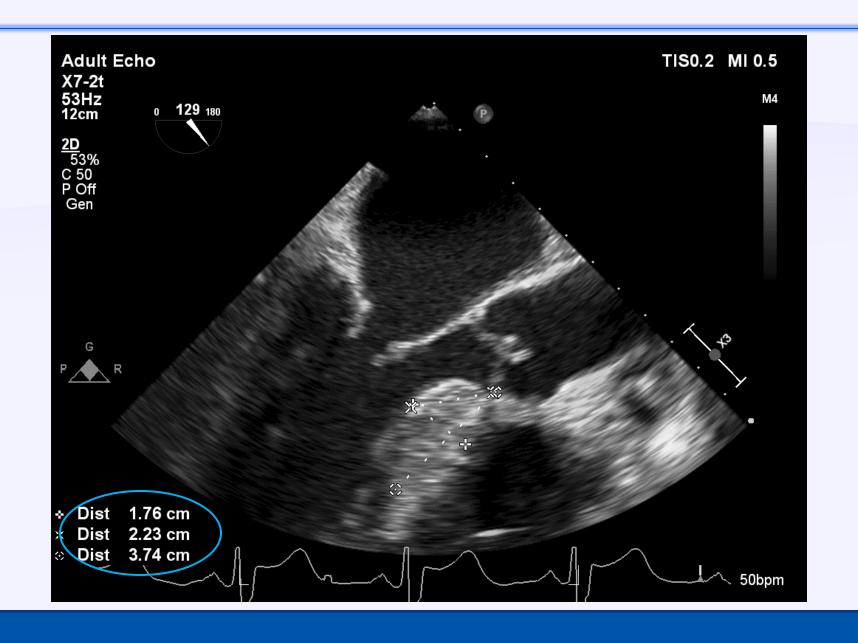


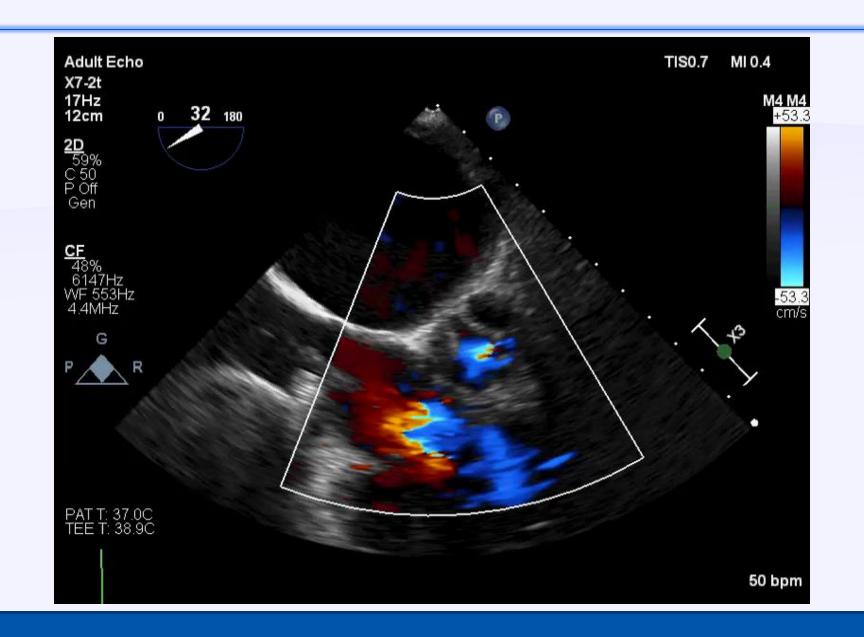


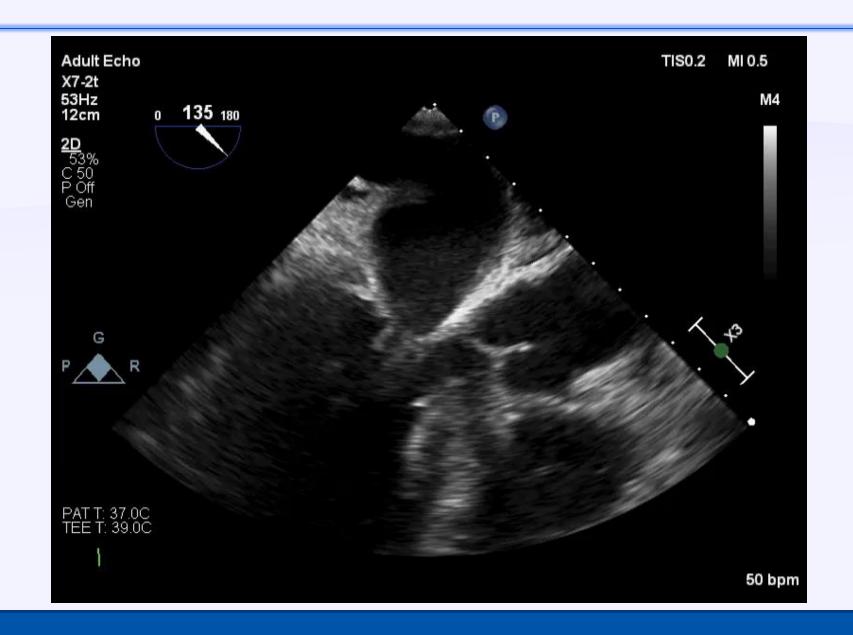


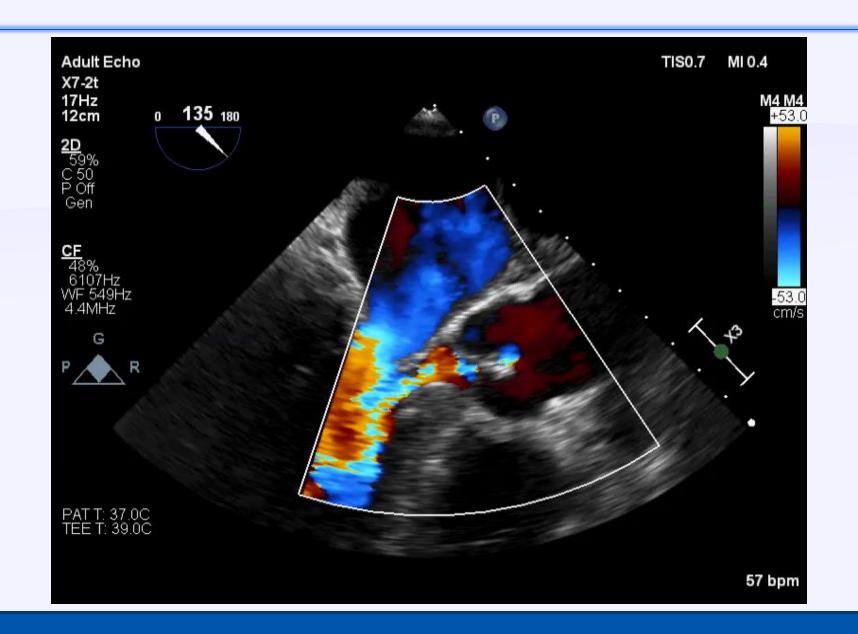


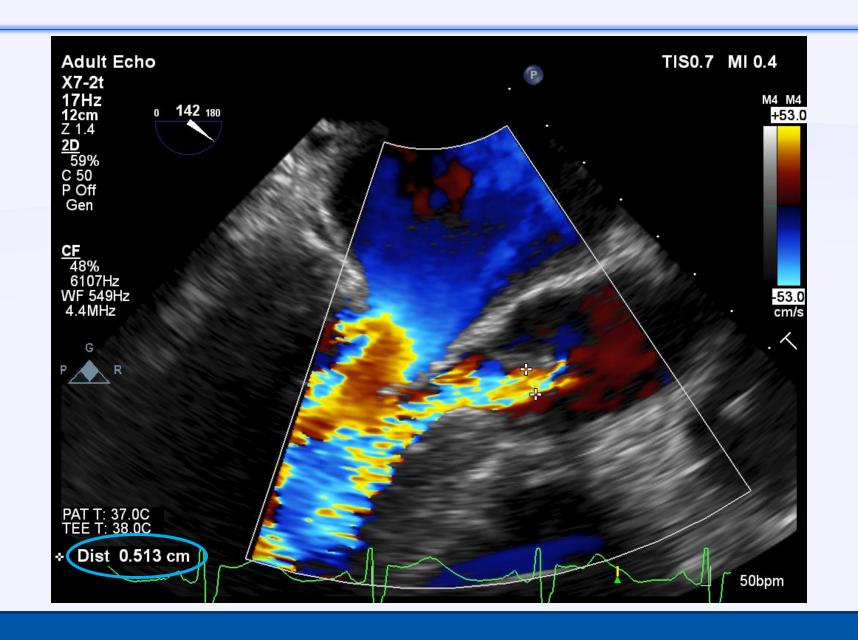


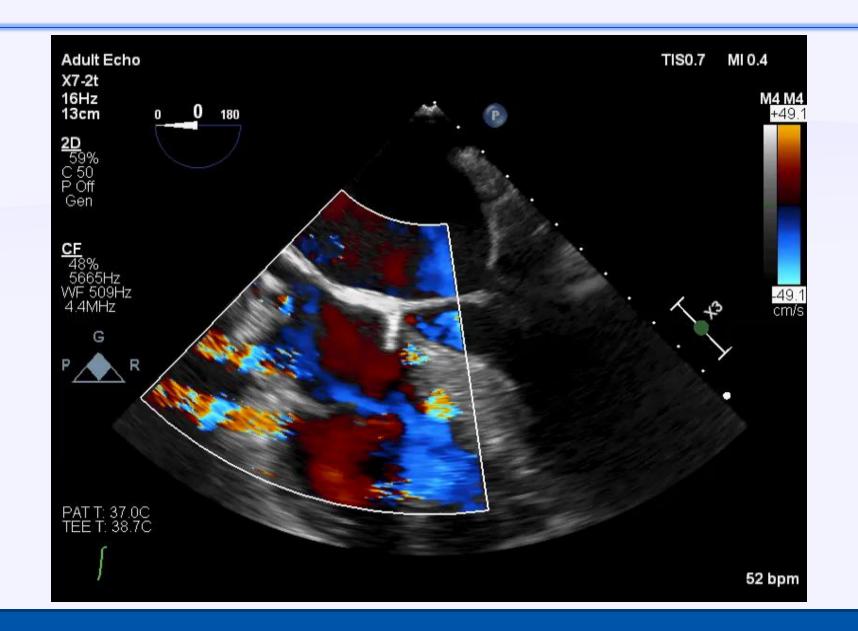


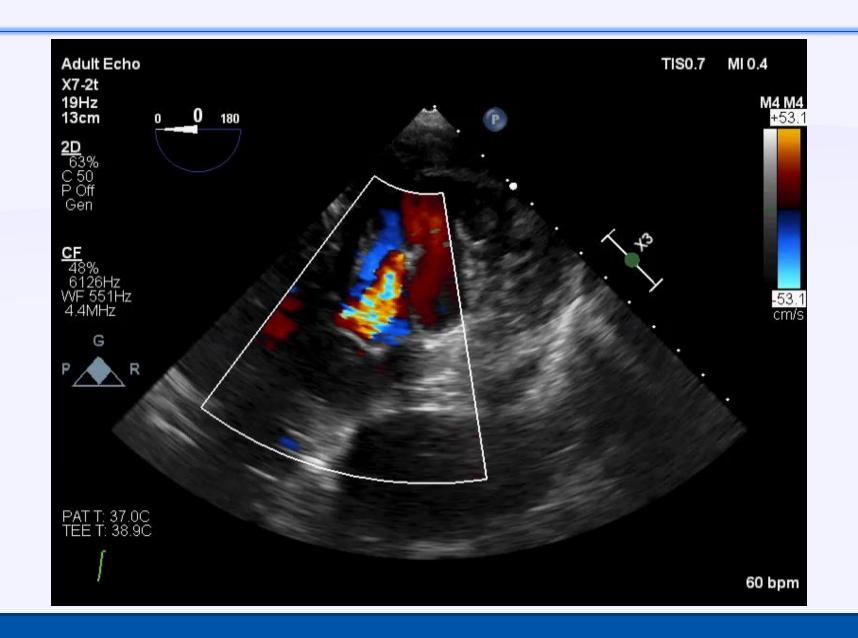


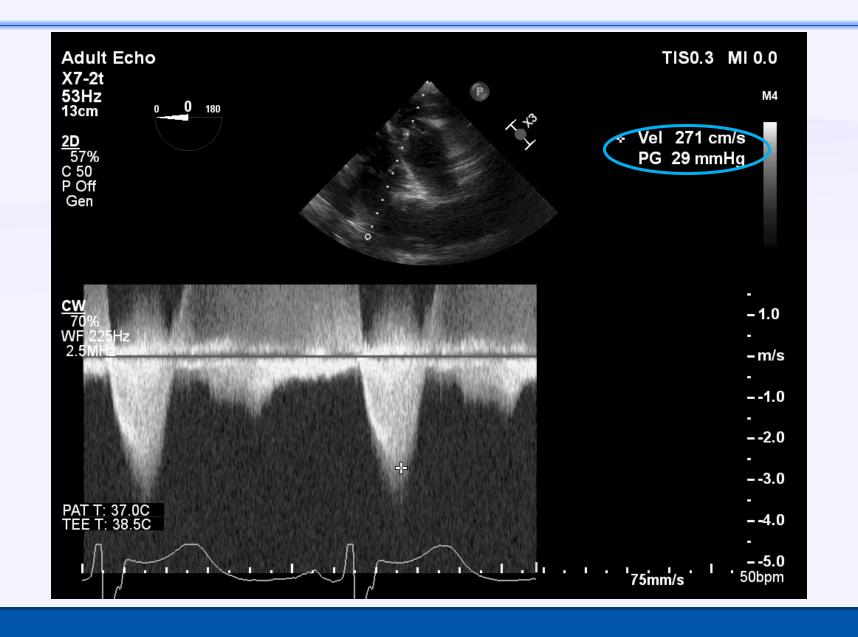




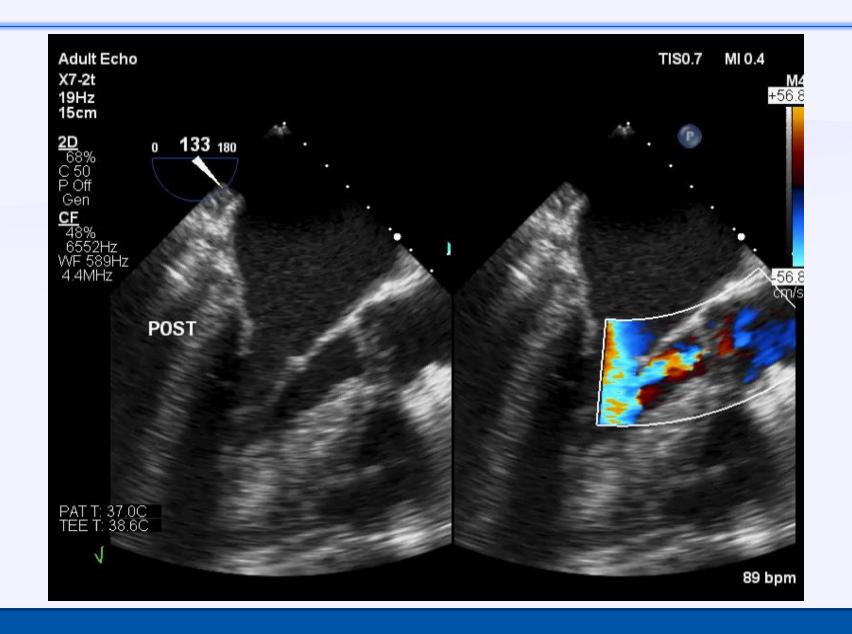


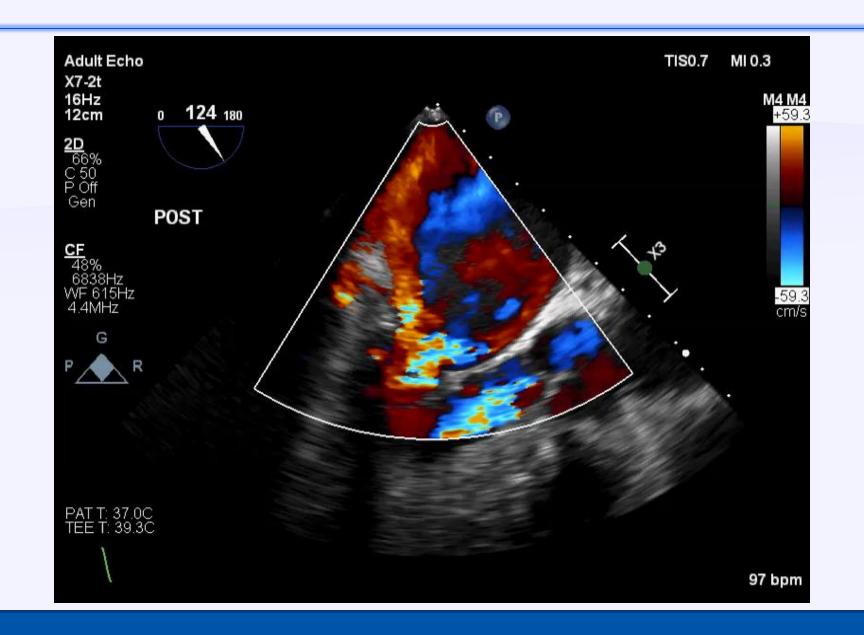


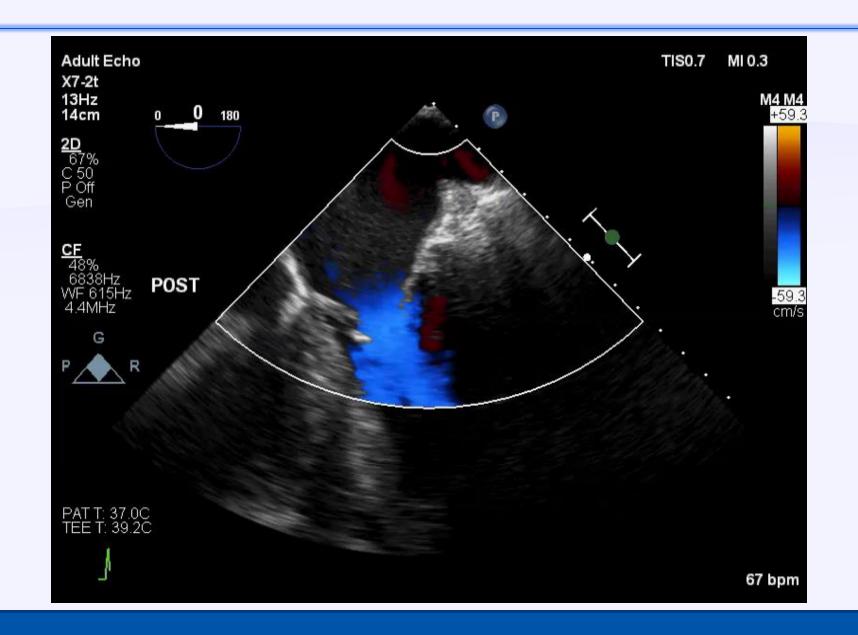


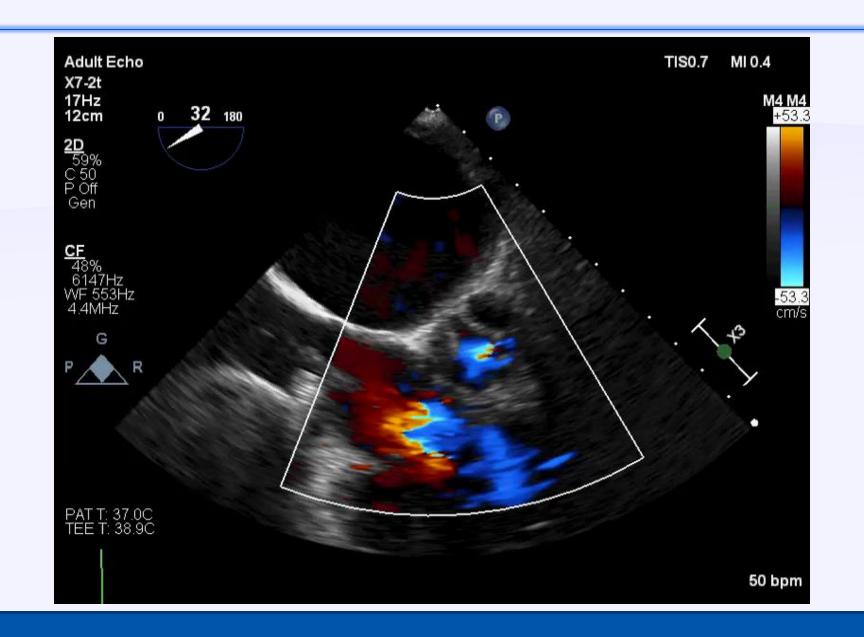


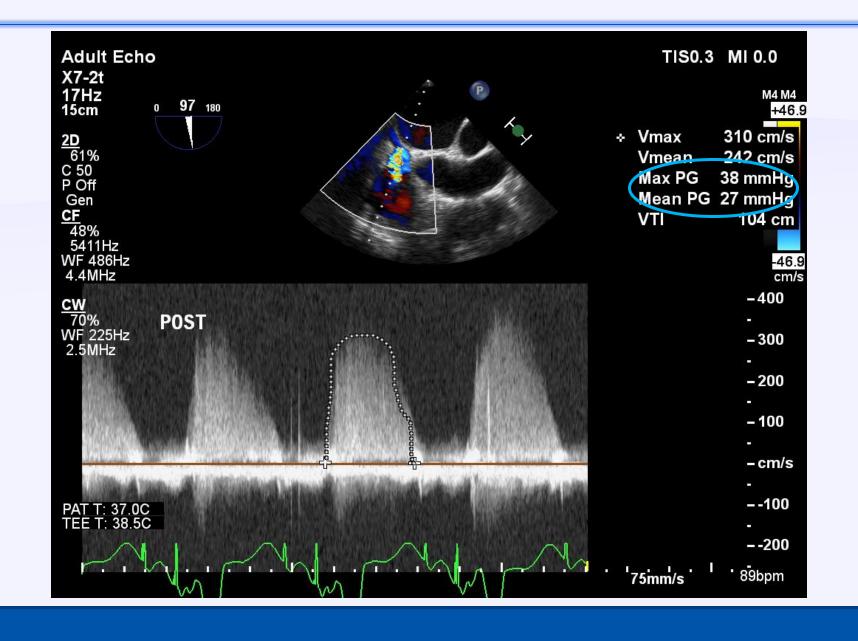
Post op

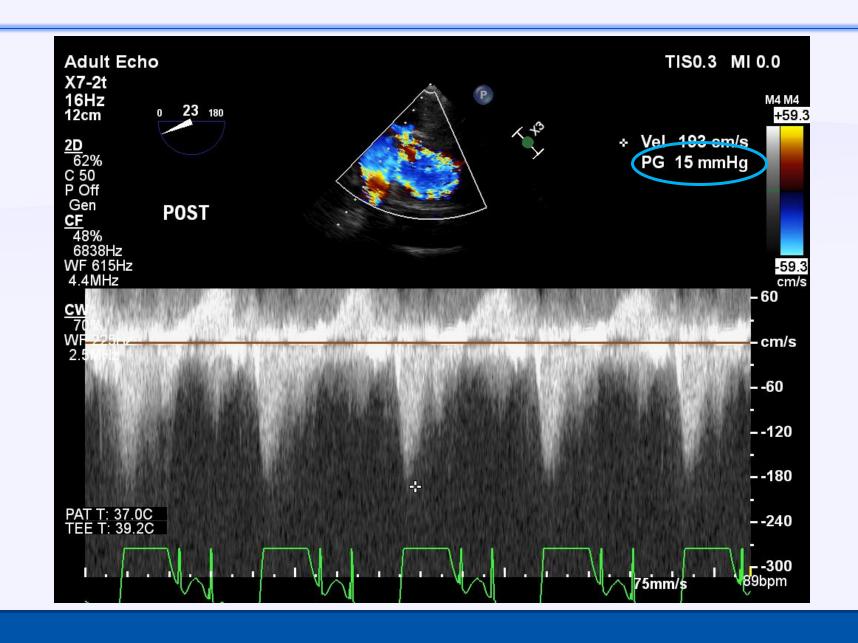












Patient had cardiac arrest in ICU the day after surgery and was resuscitated. She was discharged home one week later with moderate to severe MR.

Suggested reading materials

- 1. 2011 ACCF/AHA Guideline for the diagnosis and treatment of hypertrophic cardiomyopathy. JACC 2011.
- 2011 American Society of Echocardiography clinical recommendations for multimodality cardiovascular imaging in patients with hypertrophic cardiomyopathy. JASE 2011.
- 3. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy. European Heart Journal 2014.

Based on AHA classification which of the following cardiomyopathy is a primary cardiomyopathy?

- A. Stress induced cardiomyopathy (Takatsubo cardiomyopathy)
- B. Cardiac amyloidosis
- C. Duchenne cardiomyopathy
- D. Hemochromatosis

All of the followings are complications of surgical septal myectomy EXCEPT

- A. VSD
- B. Complete A-V block
- C. Pulmonary regurgitation
- D. Flail mitral valve

Which of the following statements about results of surgical myectomy IS CORRECT?

- A. Mortality of this surgery in an experienced center is about 2-3%
- B. In group of patients with thinner septum (septal thickness less than 1.7 cm), chance of creating VSD is much higher
- C. Chance of complete A-V block is less than alcohol septal ablation
- D. Chance of residual severe SAM is about 5%

Which of the following treatments in HCM is less effective in terms of reducing mean LVOT gradient?

- A. Surgical myectomy
- B. Pacemaker
- C. Alcohol septal ablation
- D. Disopyramide

All of the following statements about alcohol septal ablation (ASA) in HCM are correct EXCEPT

- A. It is less invasive compared to surgical myectomy
- B. Chance of complete A-V block is about 10%
- C. In elderly patients with comorbidities is a better choice than surgery
- D. Relief of the gradient at the LVOT is faster than surgical myectomy

Correct Answers

1- A

2- C

3- C

4- B

5- D

