What to Do for a 75 Year Old Man of Severe Ebstein Anomaly with Decreased RV Function, Atrial Fibrillation

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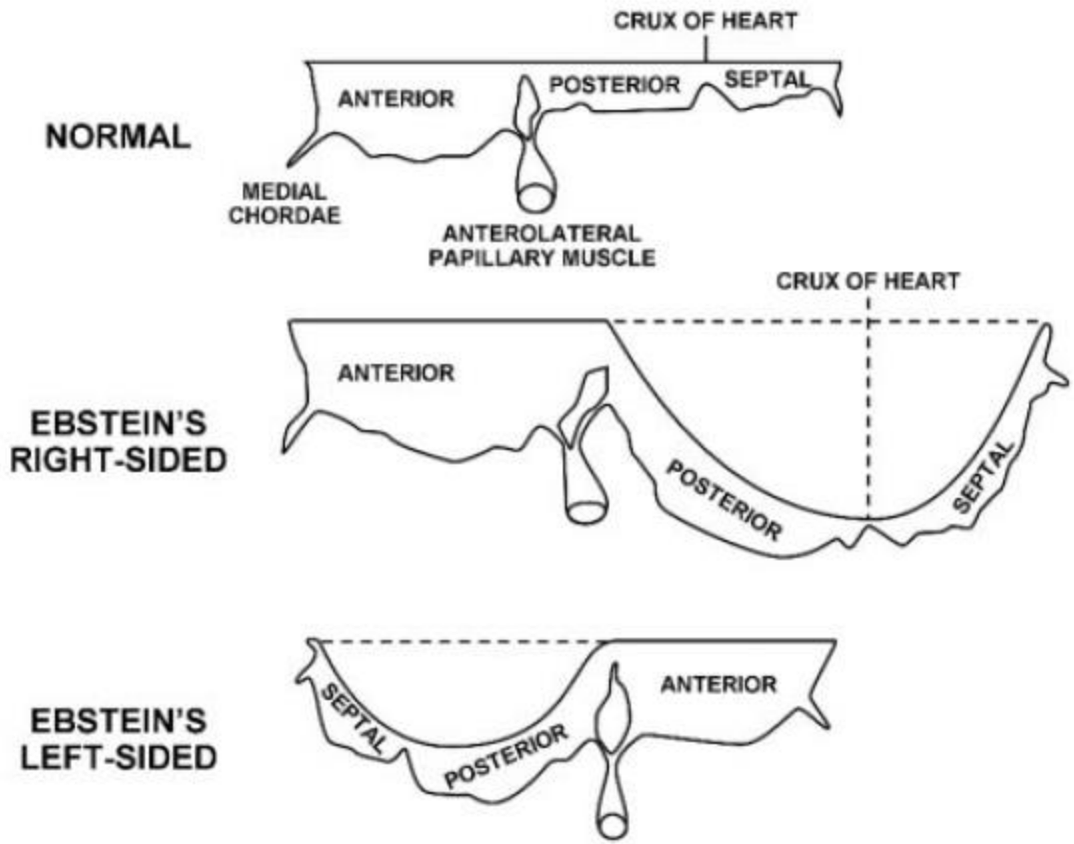
COI Disclosure

Name of First Author: ***Jong-Min Song***

The authors have no financial conflicts of interest to disclose concerning the presentation

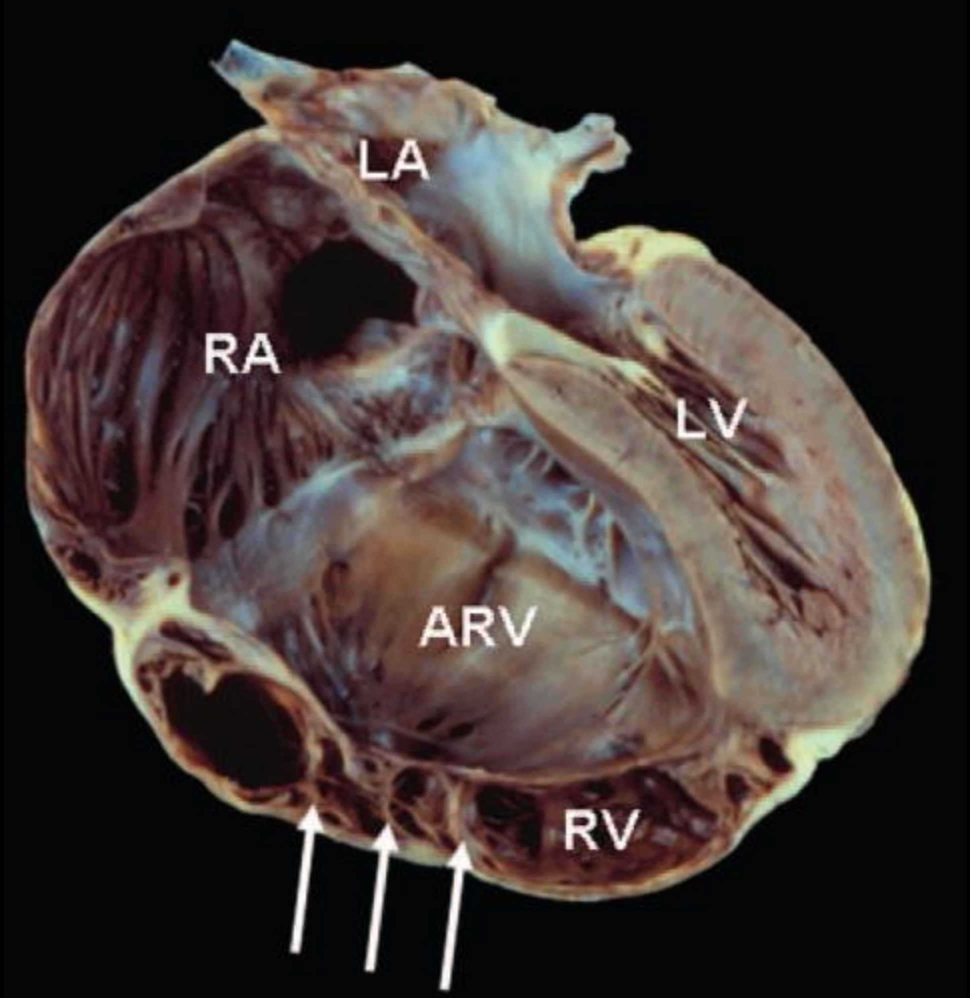
Ebstein Anomaly

Ebstein's Anomaly



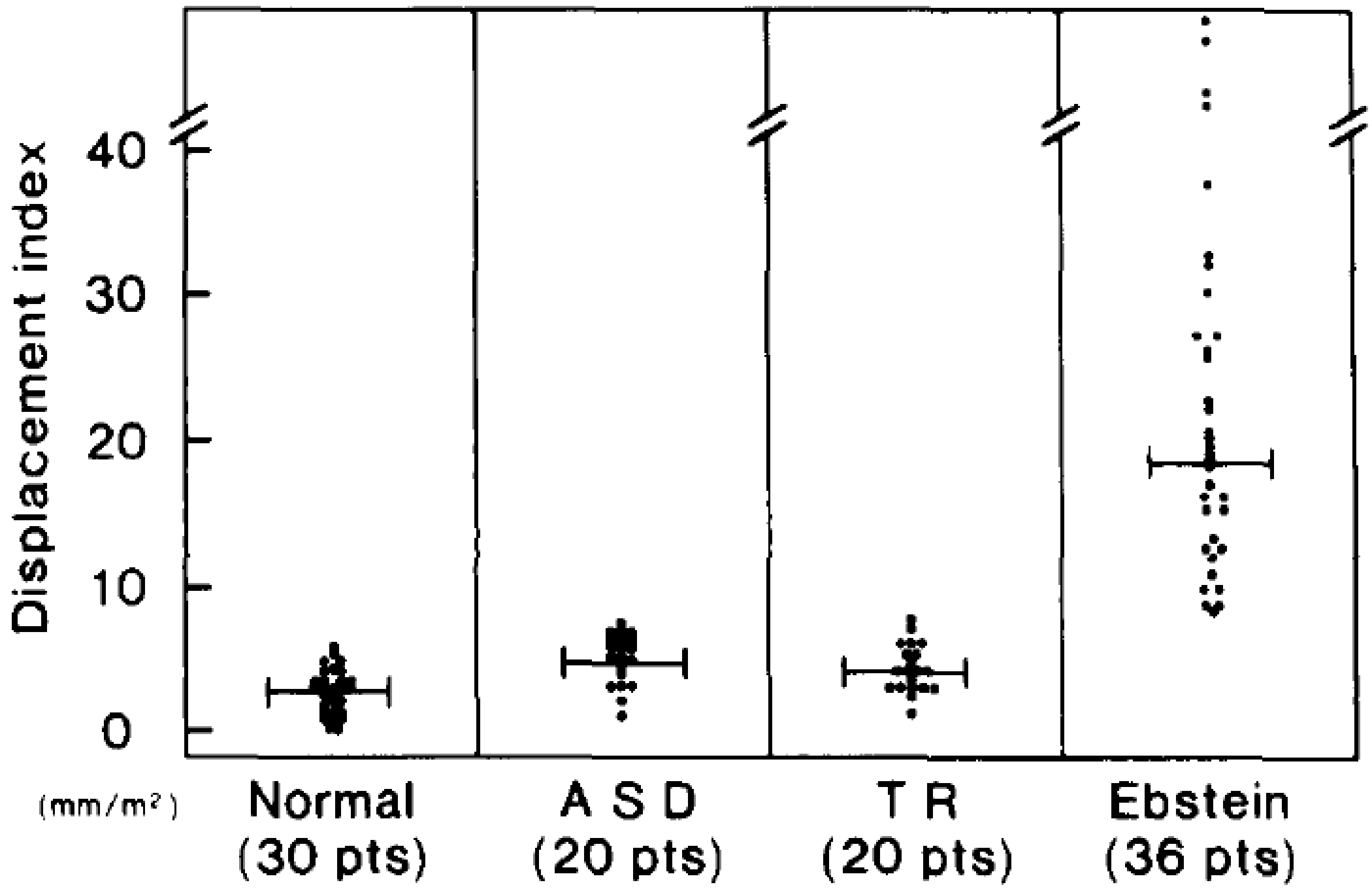
Attenhofer Jost CH, Circulation. 2007;115:277-285

Severe Ebstein's Anomaly



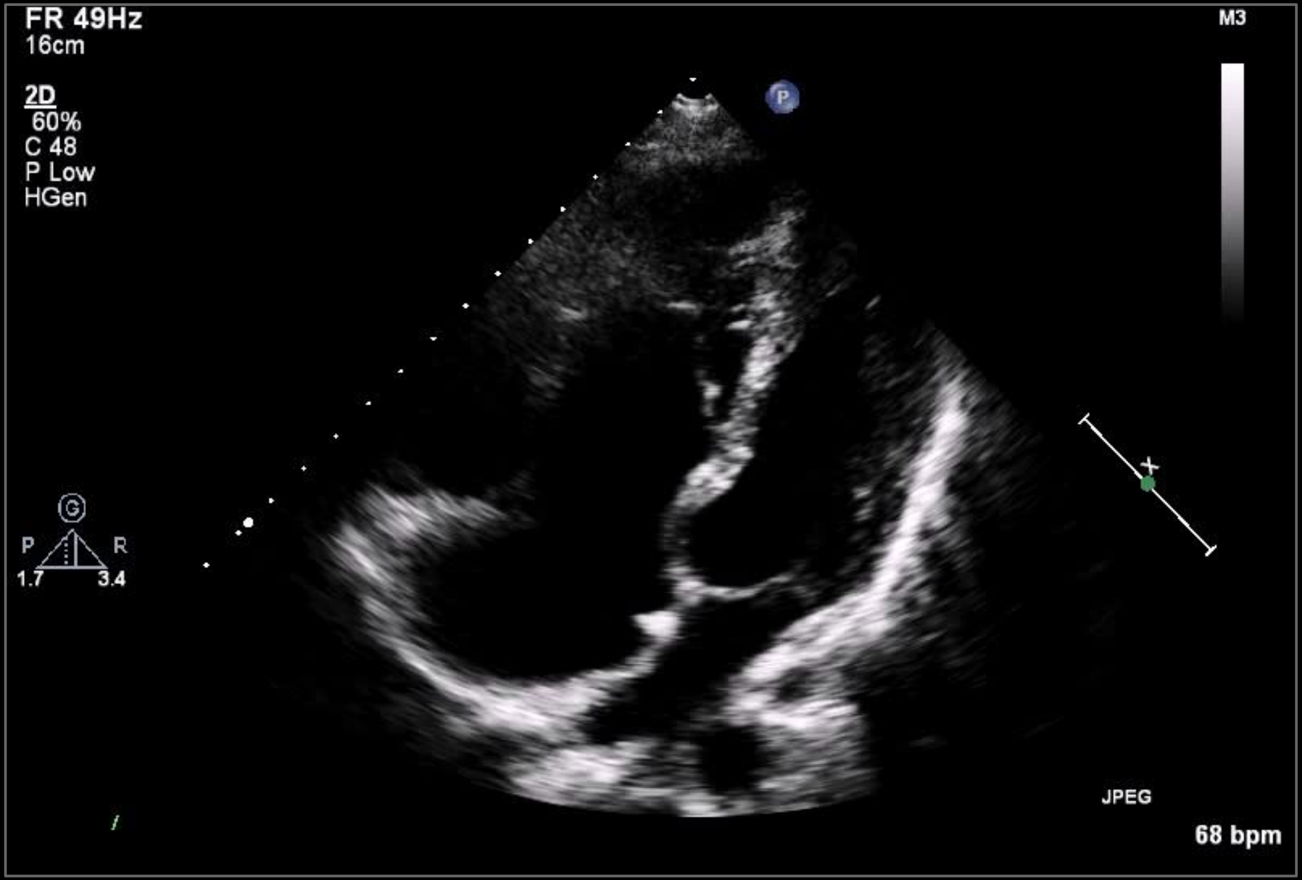
Attenhofer Jost CH, Circulation. 2007;115:277-285

Displacement of Septal Leaflet

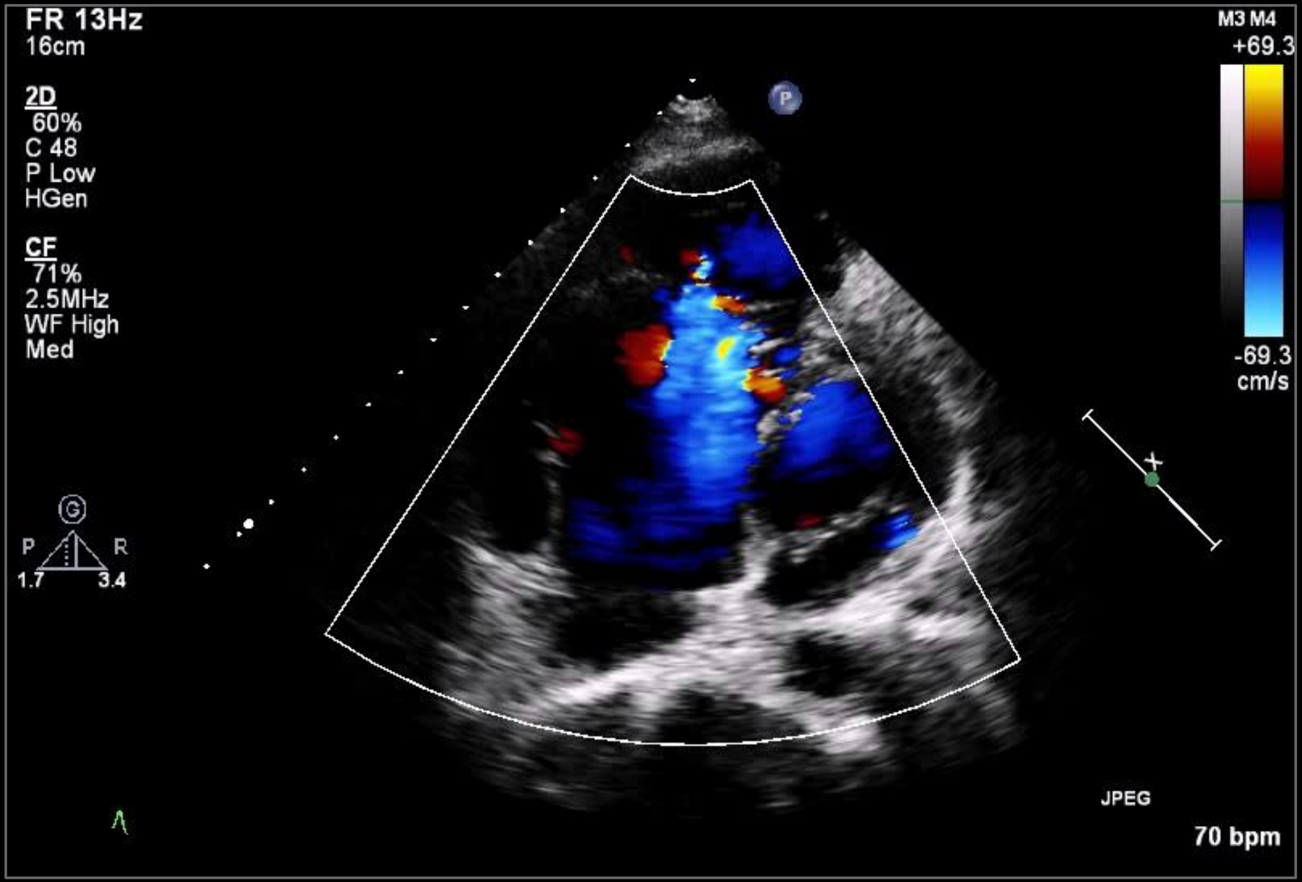


Shiina A, JACC 1984:356-70

Ebstein Anomaly



Ebstein Anomaly



Ebstein Anomaly

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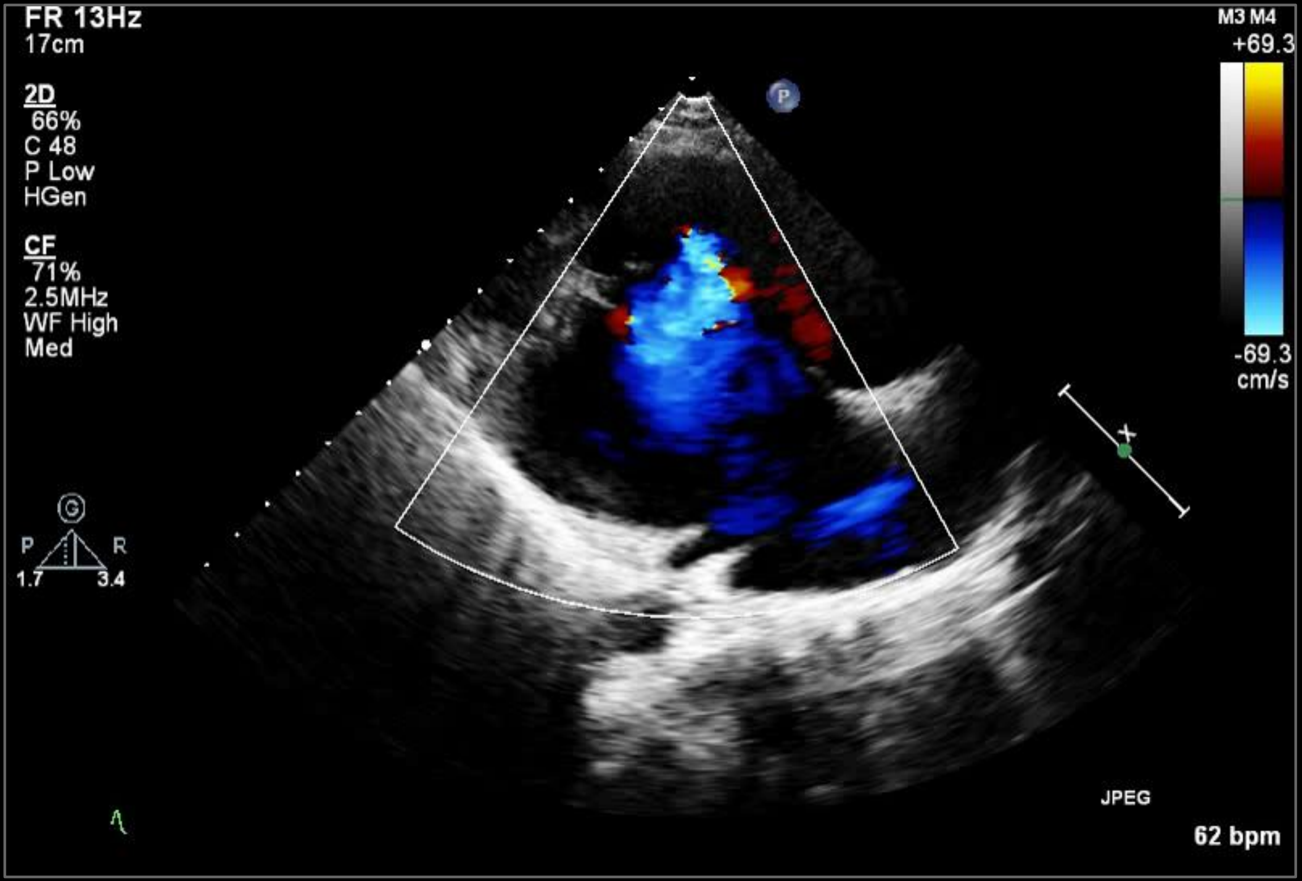

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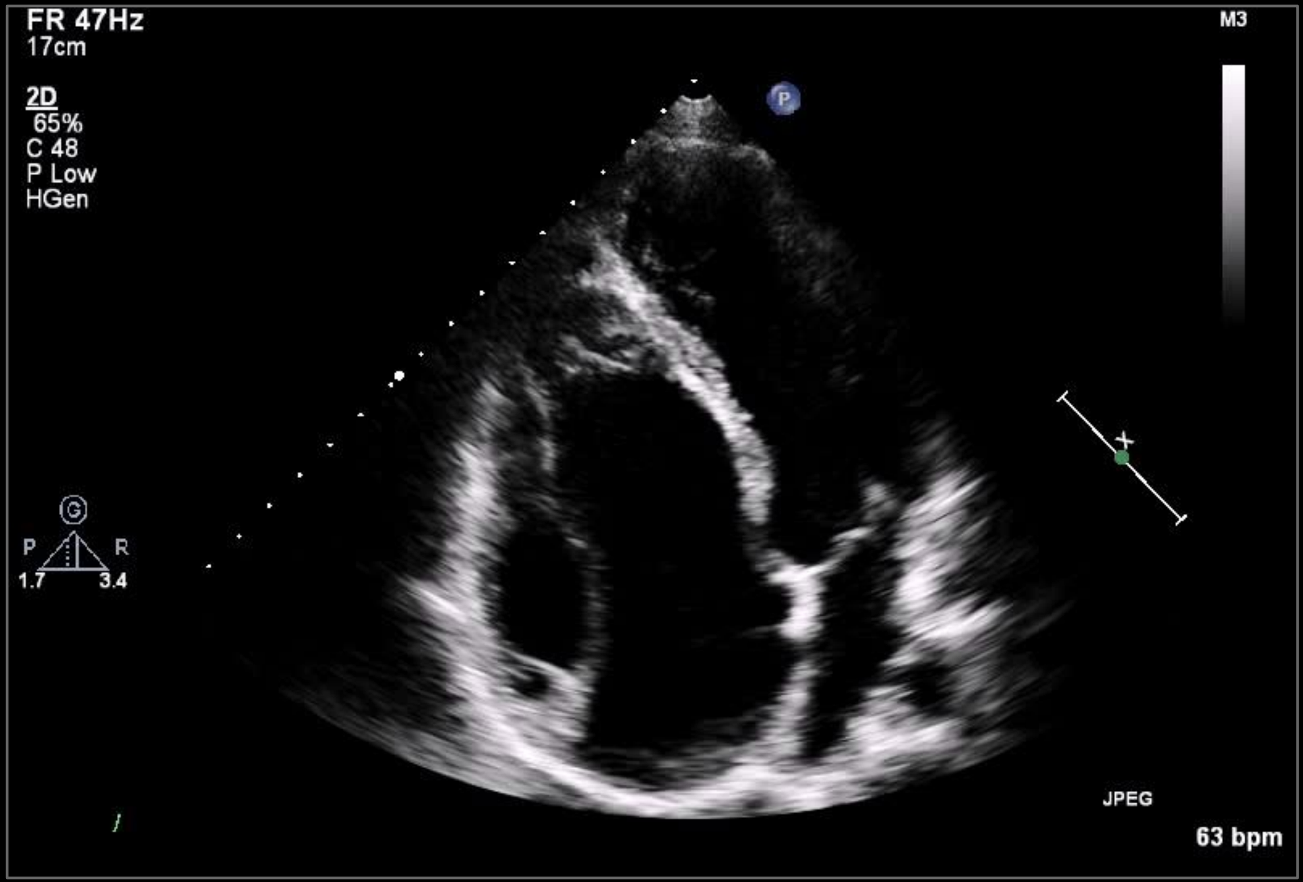

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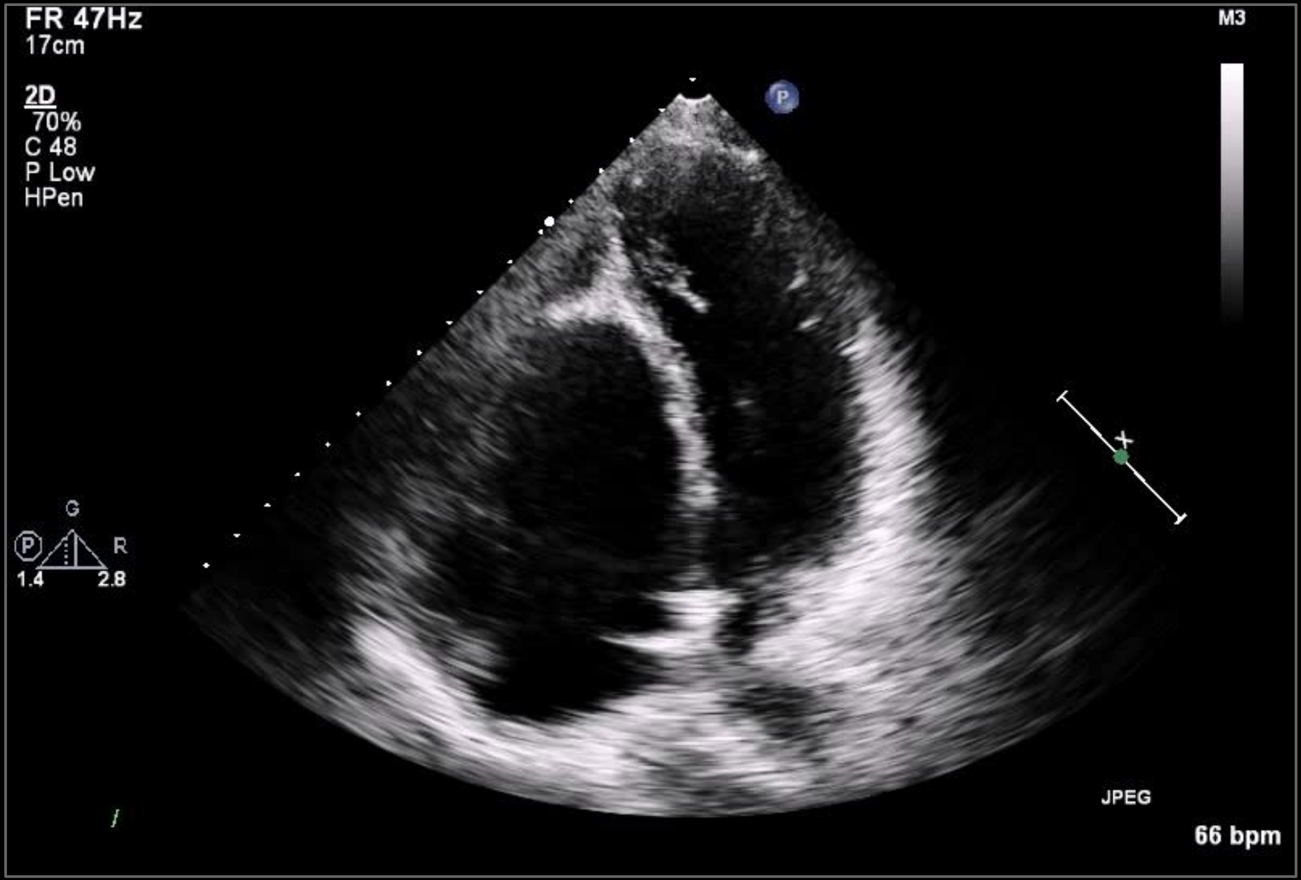

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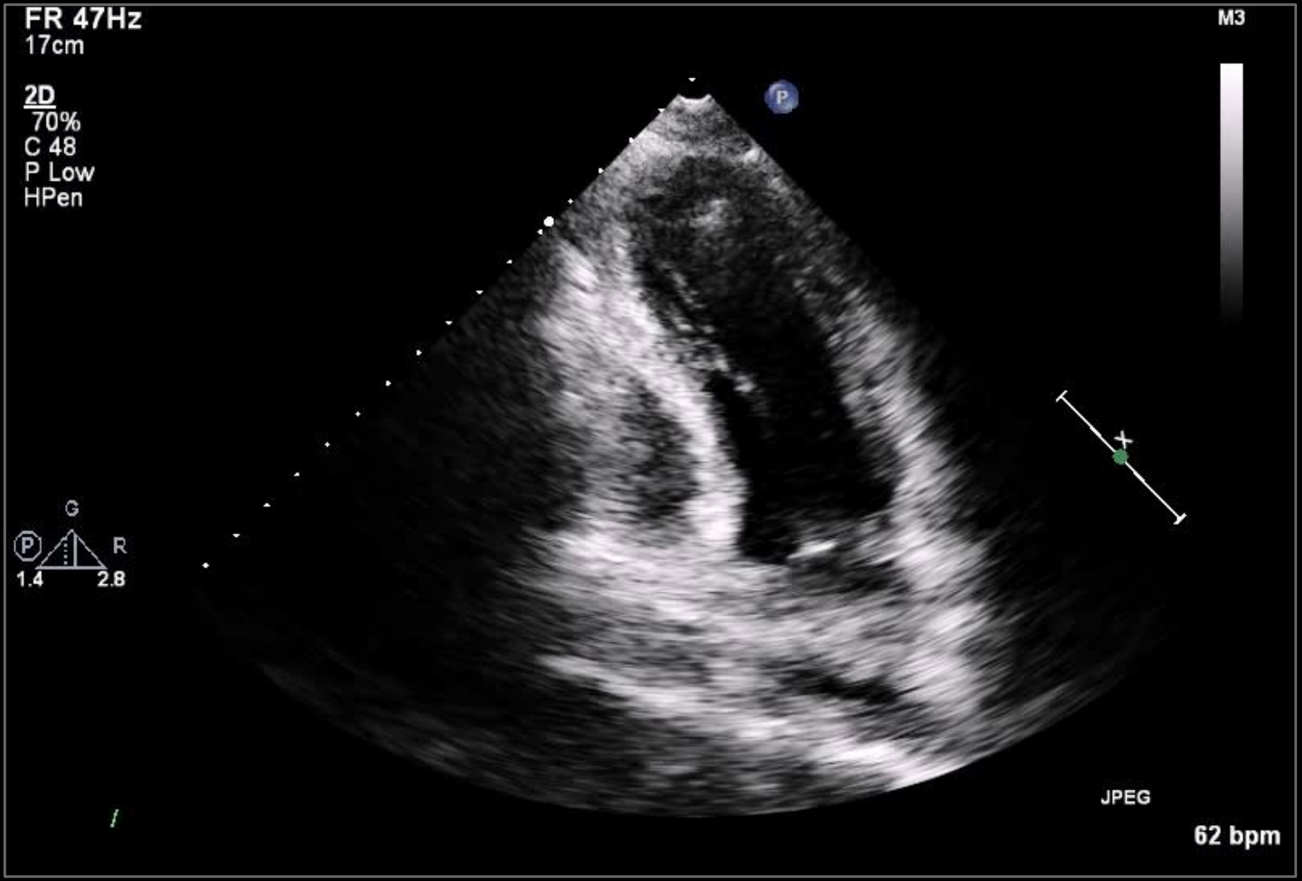


Anatomic Assessment

❍ Tricuspid valve leaflets

- Apical displacement of septal/posterior leaflet (> 8 mm/m^2 or 20 mm in adults)
- Tethering, elongation, absence of the leaflets
- Leaflet fenestrations (anterior leaflet)
- Accessory leaflet tissue (muscular shelf)
  - Between septal and anterior leaflets
- Annulus dilatation

Anatomic Assessment

Right ventricle

- Atrialized right ventricle
- Right ventricular dilatation
  - Between annulus and posterior leaflet
- Posterior aneurysm/RVOT aneurysm
  - 20%

Left ventricle

- Size and shape of the left ventricle
- Mitral valve prolapse
  - Elongated chordae

Functional Assessment

❍ Tricuspid valve

- Tricuspid regurgitation or stenosis
- Pressure gradient across the tricuspid valve
- Severity of tricuspid regurgitation
- Right ventricular inflow tract obstruction

Functional Assessment

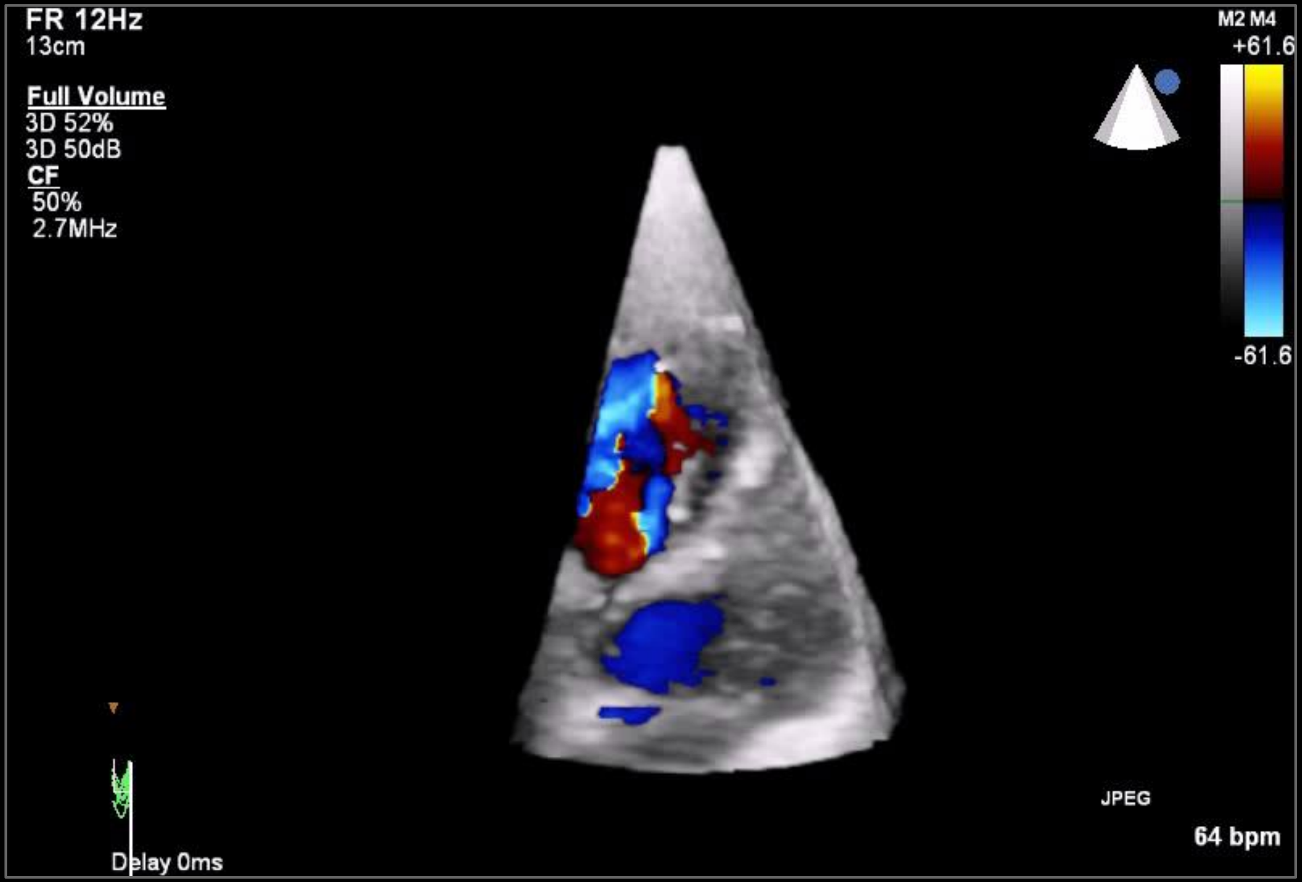
- **Right ventricle**
  - Progression of chamber enlargement
  - Right ventricular function

- **Left ventricle**
  - Left ventricular function
  - Mitral regurgitation (severity)

Ebstein Anomaly

Ebstein Anomaly

Ebstein Anomaly



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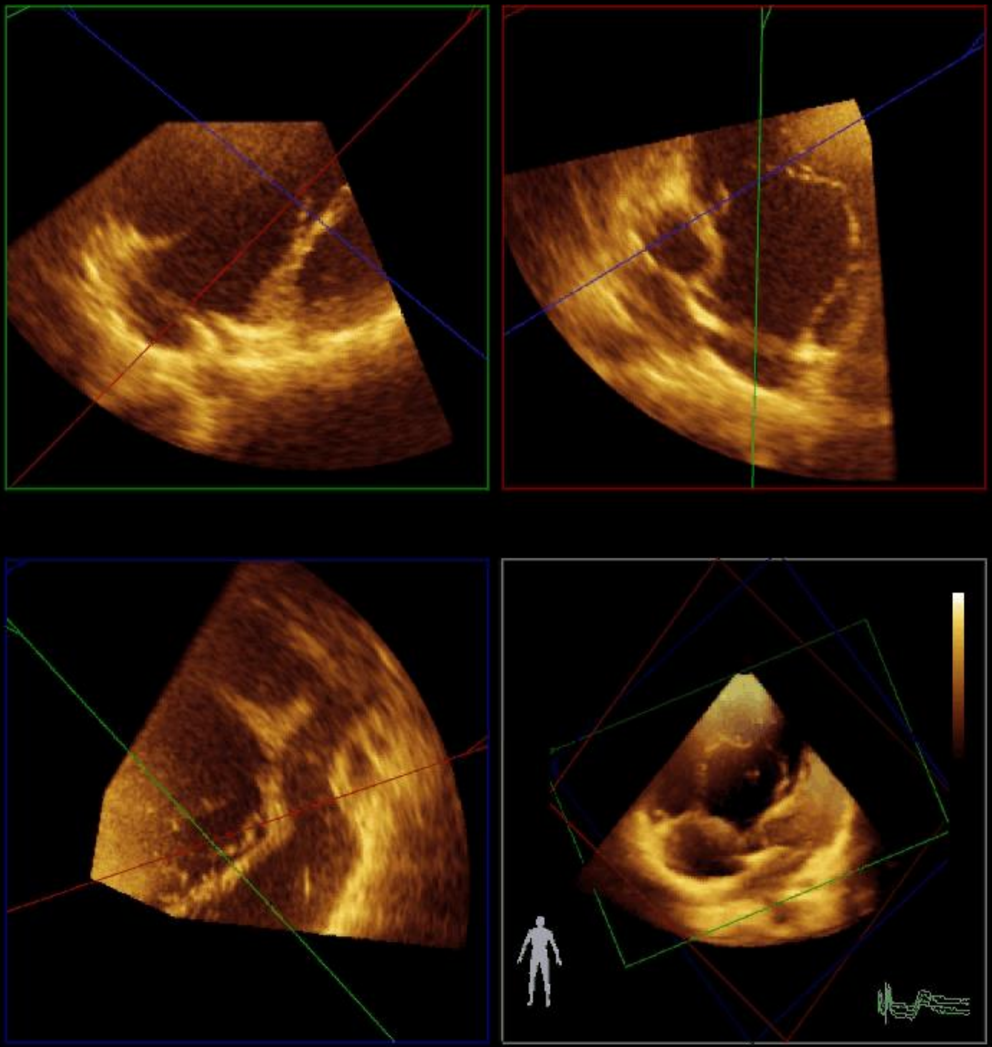


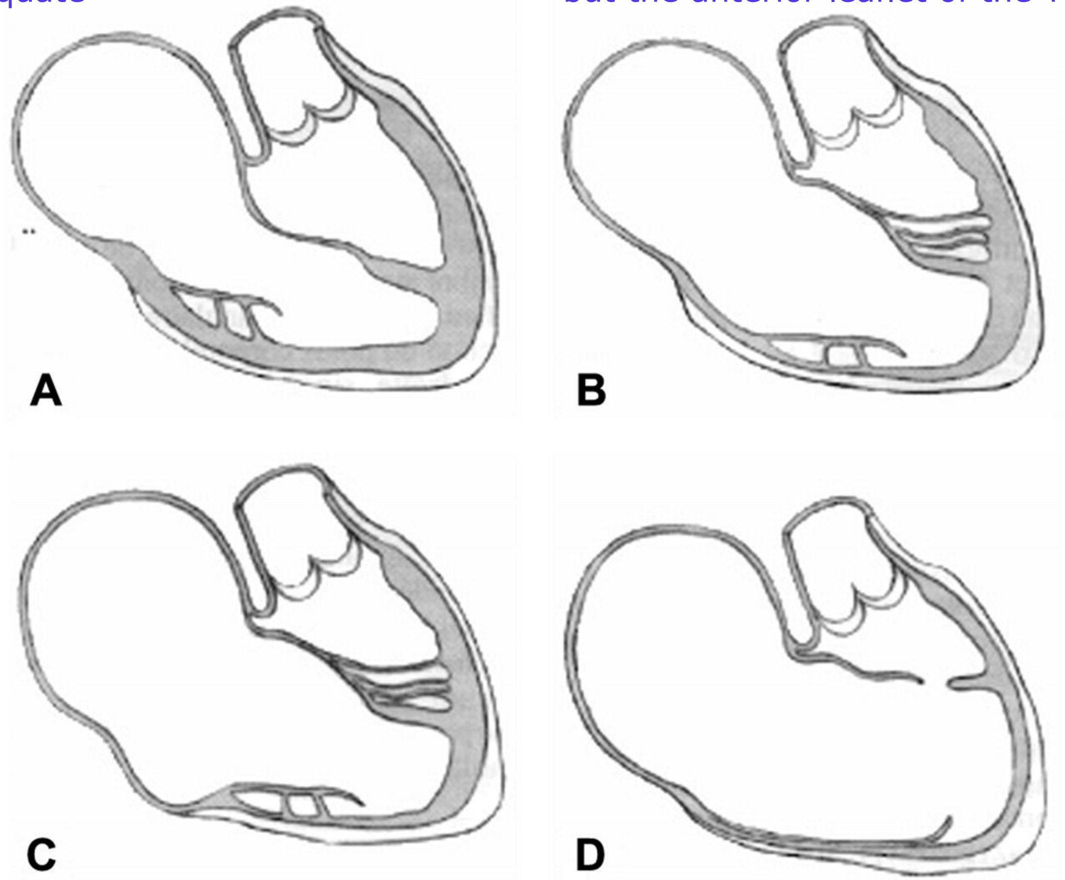
PHILIPS

Severe
Ebstein Anomaly

Carpentier Classification

The volume of the true RV is adequate

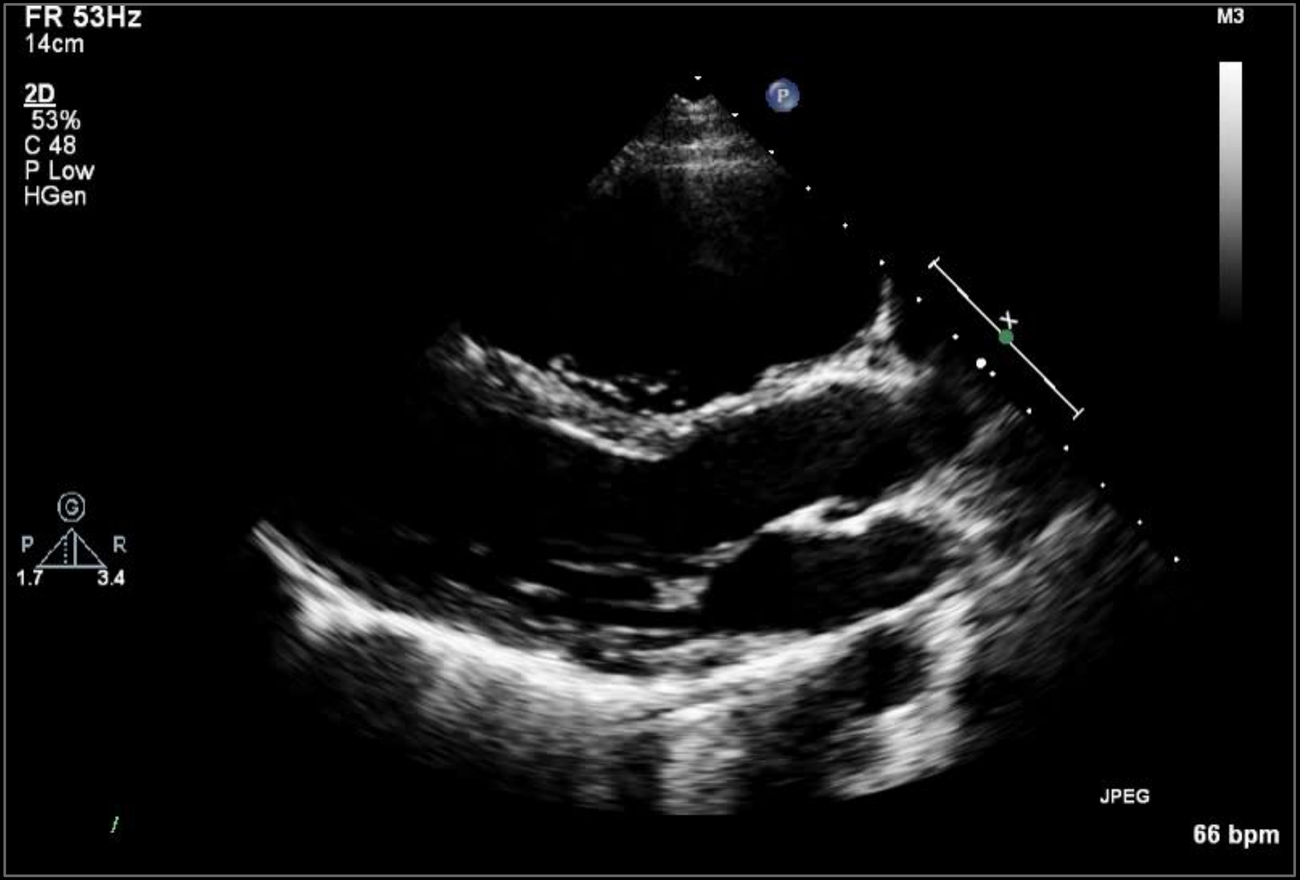
A large atrialized component of the RV exists, but the anterior leaflet of the TV moves freely



The anterior leaflet is severely restricted in its movement and may cause significant obstruction of the RVOT

Almost complete atrialization of the RV except for a small infundibular component

Ebstein Anomaly



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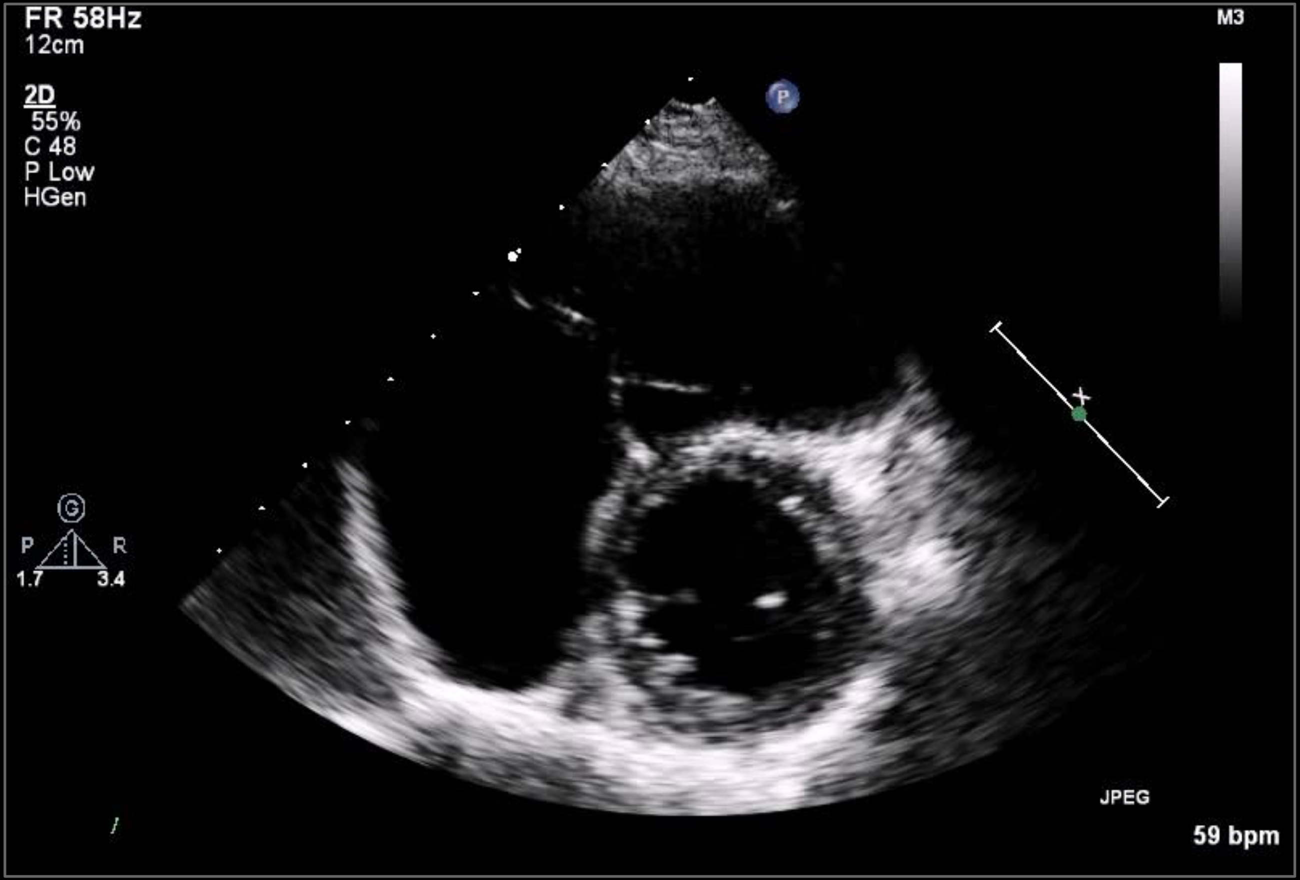
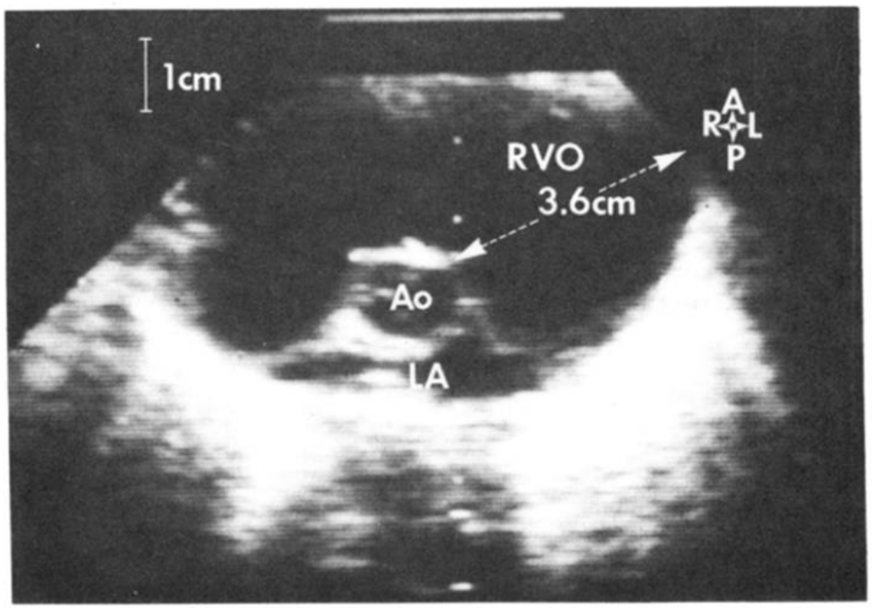
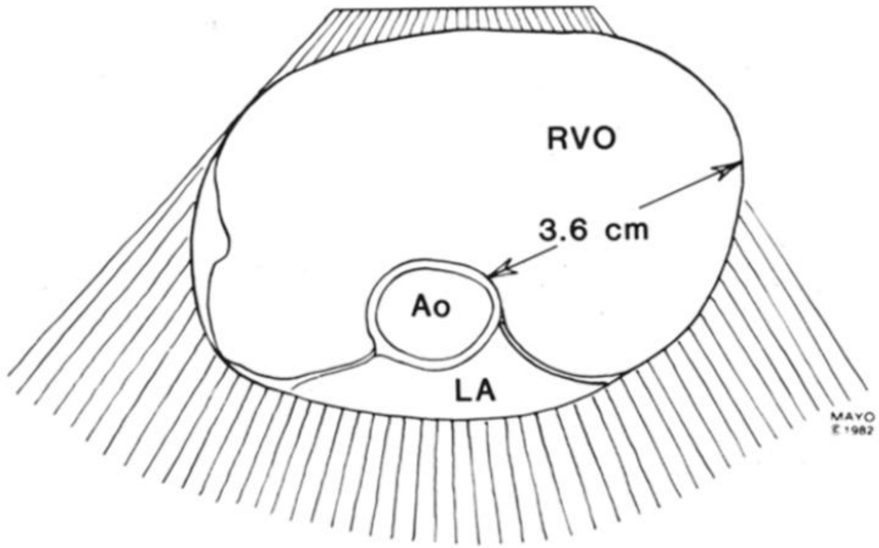


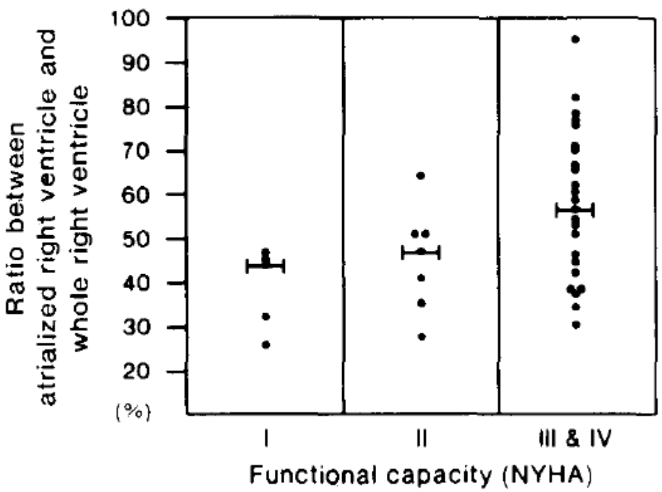
Aneurysmal Dilatation of RVOT



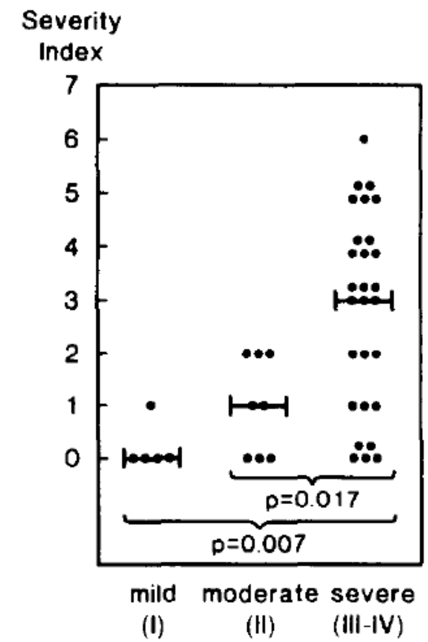


Shiina A, JACC 1984:356-70

Anatomy & Functional Disability



	2-D echo features	Index Number
Septal leaflet	absent	1
	displacement ≥ 25 mm/m^2	1
Anterior leaflet	displacement	1
	displacement of free edge	1
	restricted motion	1
	severe prolapse	1
Intracardiac cavities	aneurysmal RVOT	1
	aRV/RV ≥ 50%	1
	tricuspid anulus ≥ 45 mm/m^2	1
	right atrium ≥ 60 mm/m^2	1
	TOTAL	10

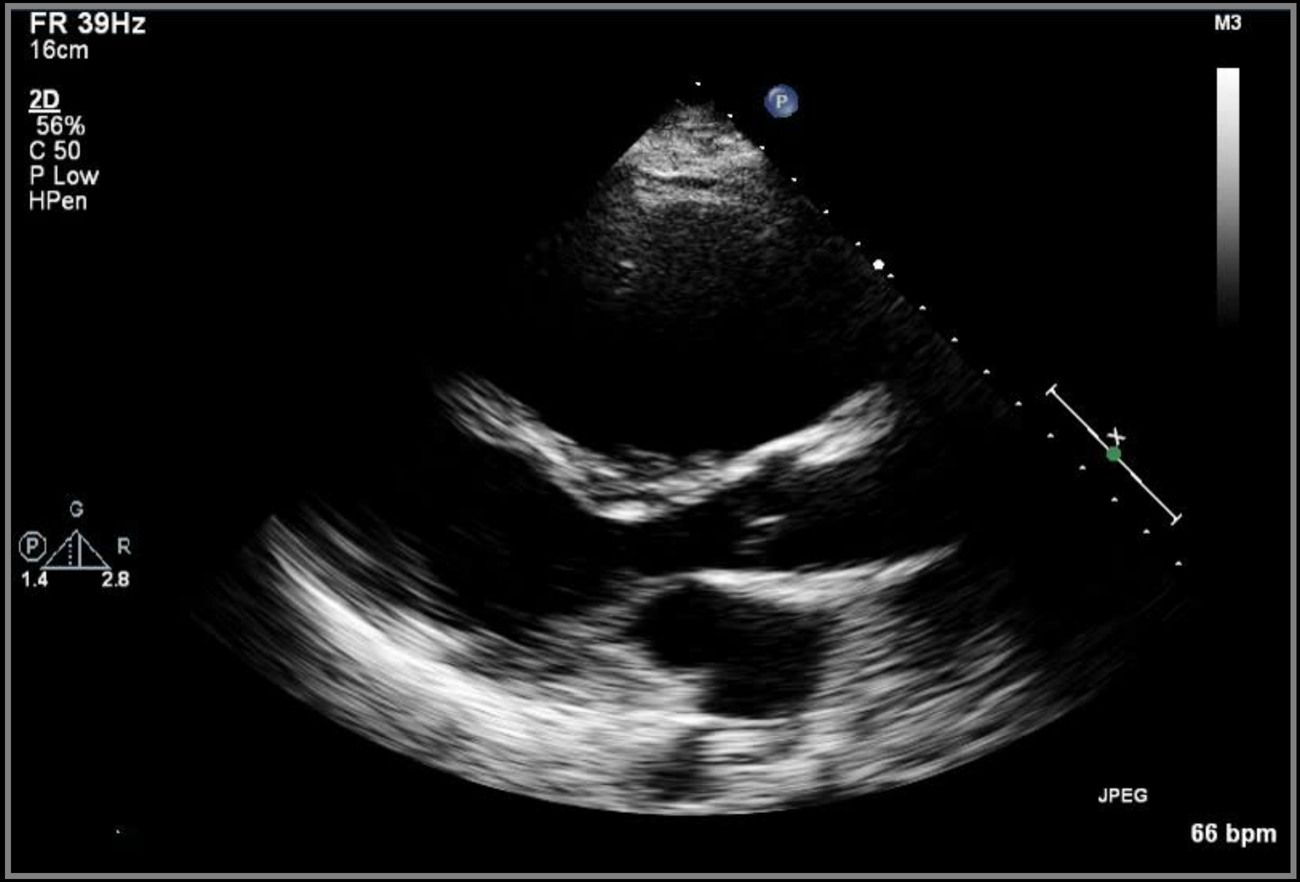


Shiina A, JACC 1984:356-70

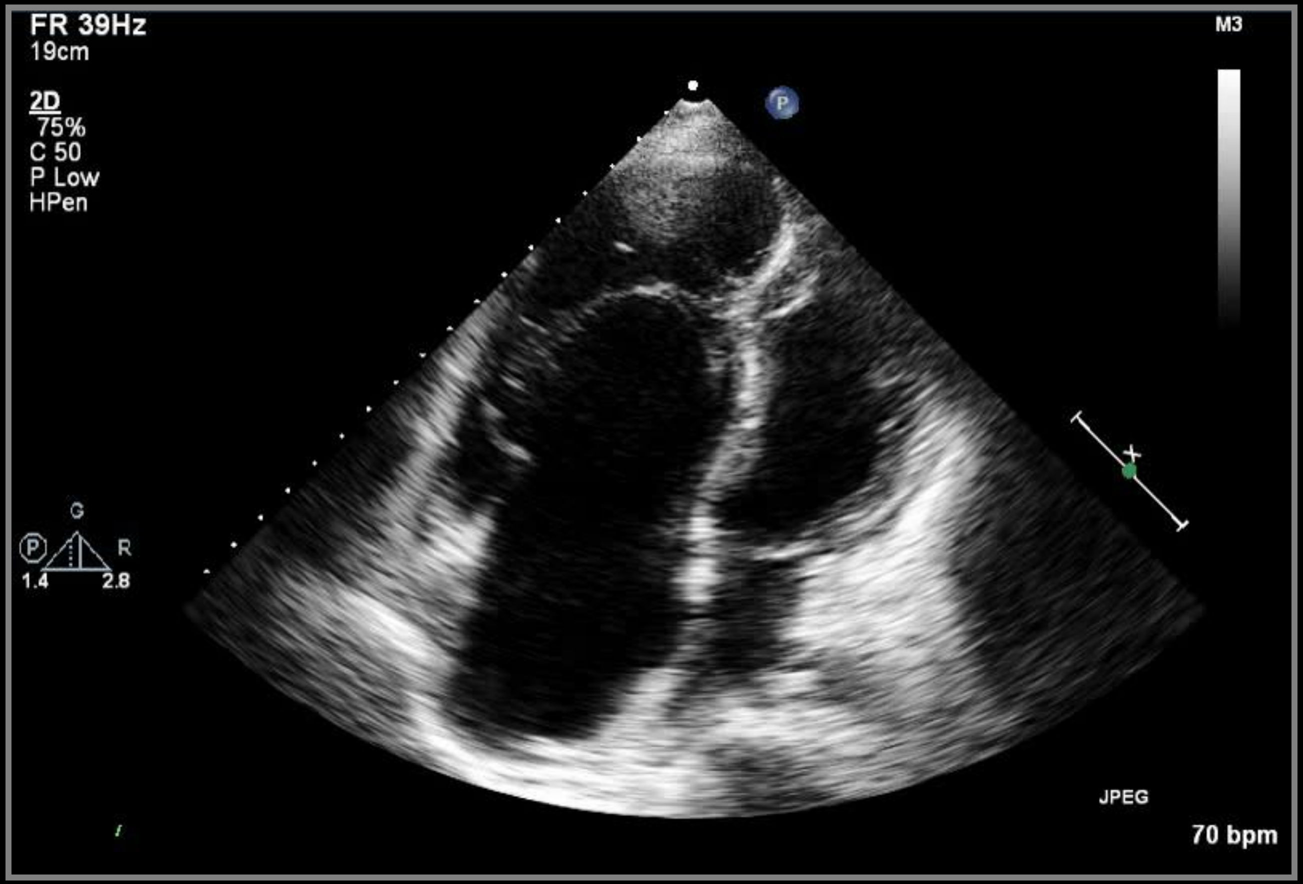
Surgery
vs.
Observation

50 YO / M

- Alleged CHD, DOE Fc I



50 YO / M



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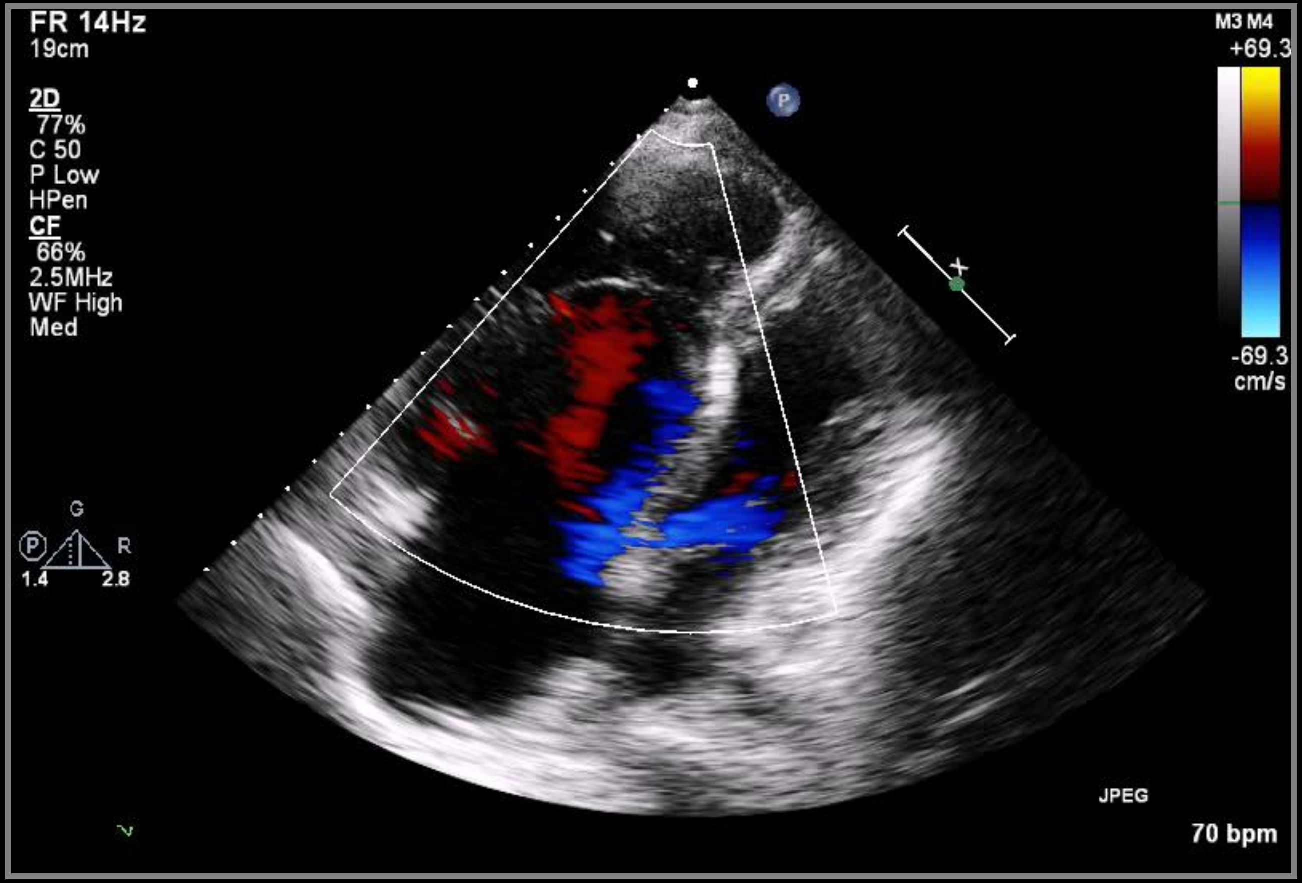
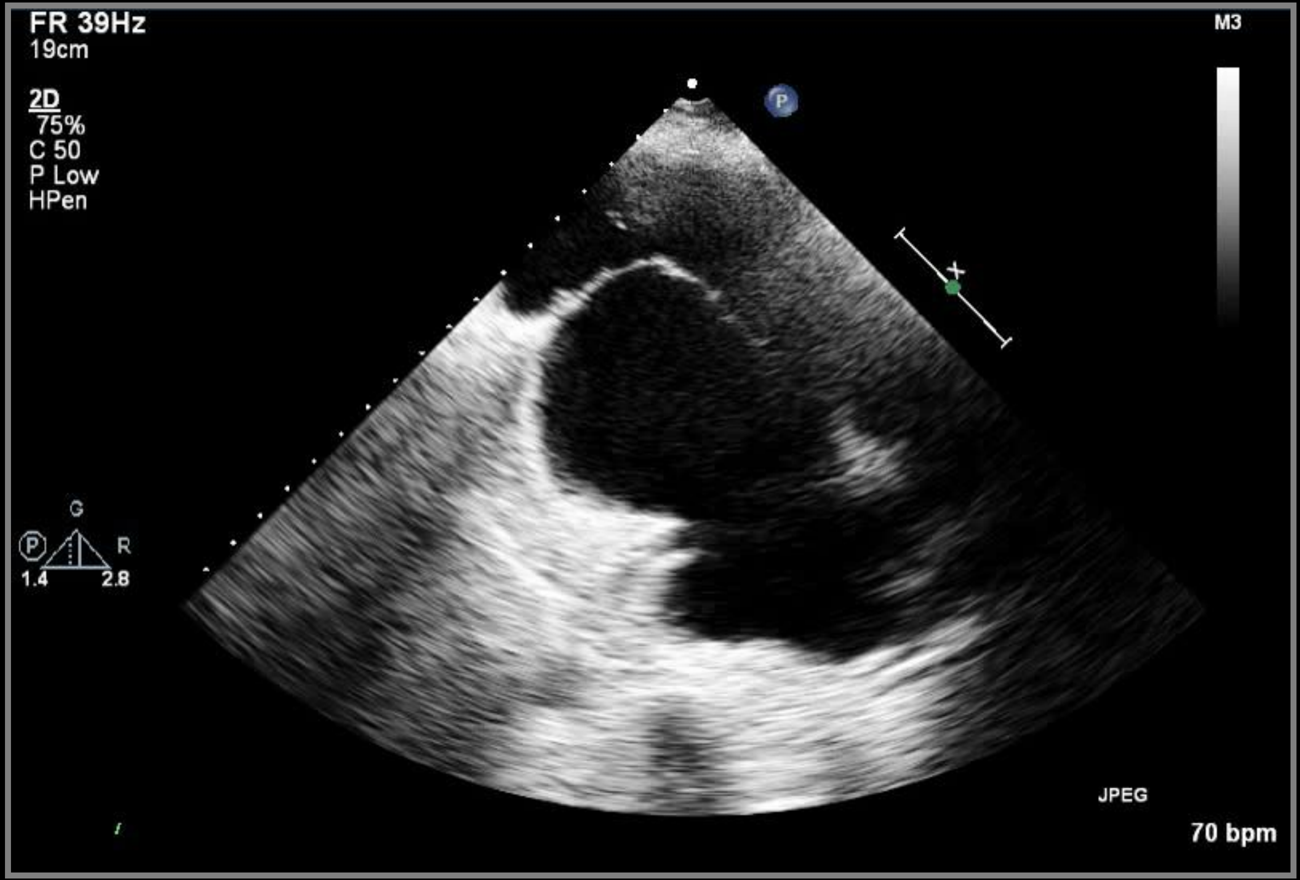
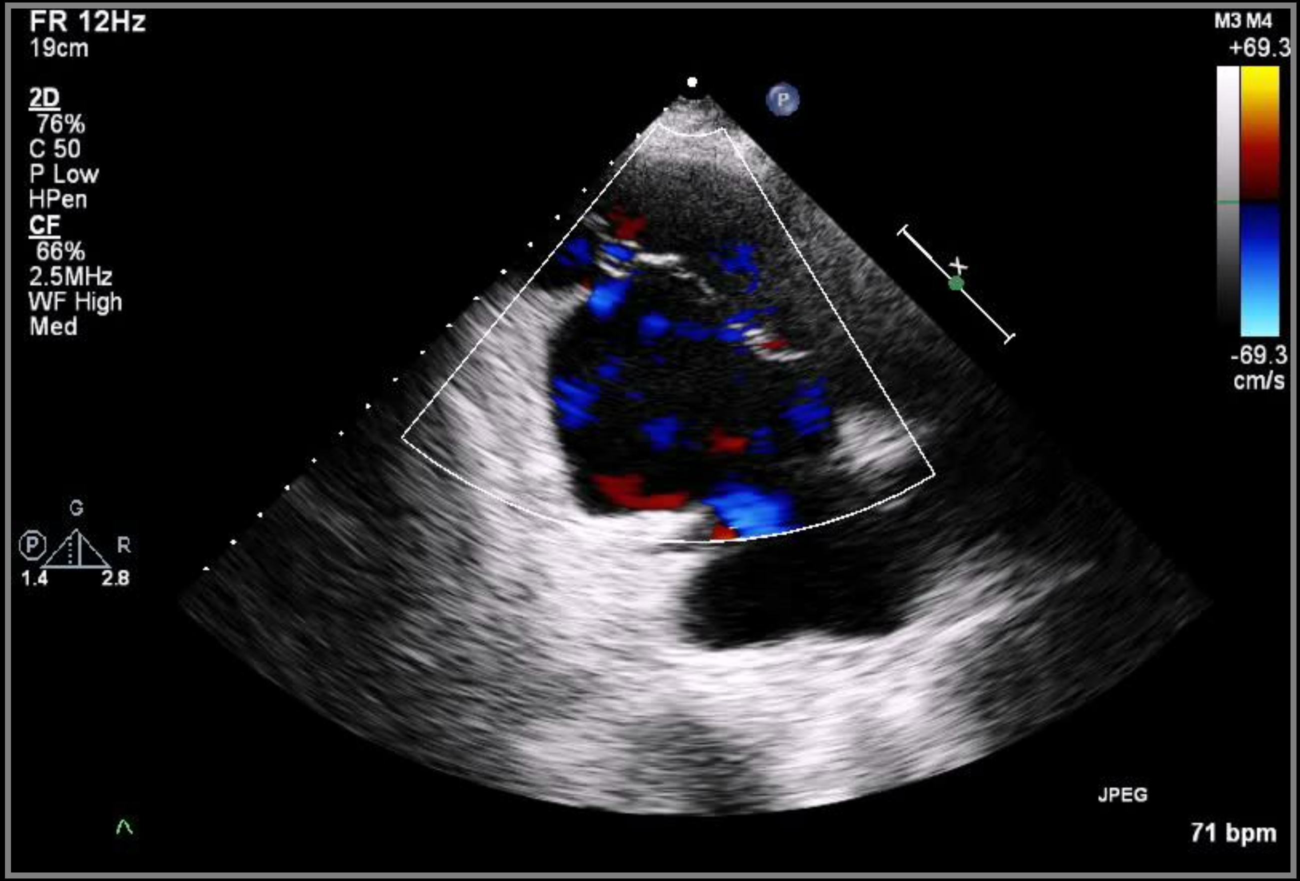


FR 14Hz
19cm
2D
77%
C 50
P Low
HPen
CF
66%
2.5MHz
WF High
Med
M3 M4
+69.3
-69.3
cm/s
G
R
1.4
2.8
JPEG
70 bpm

50 YO / M

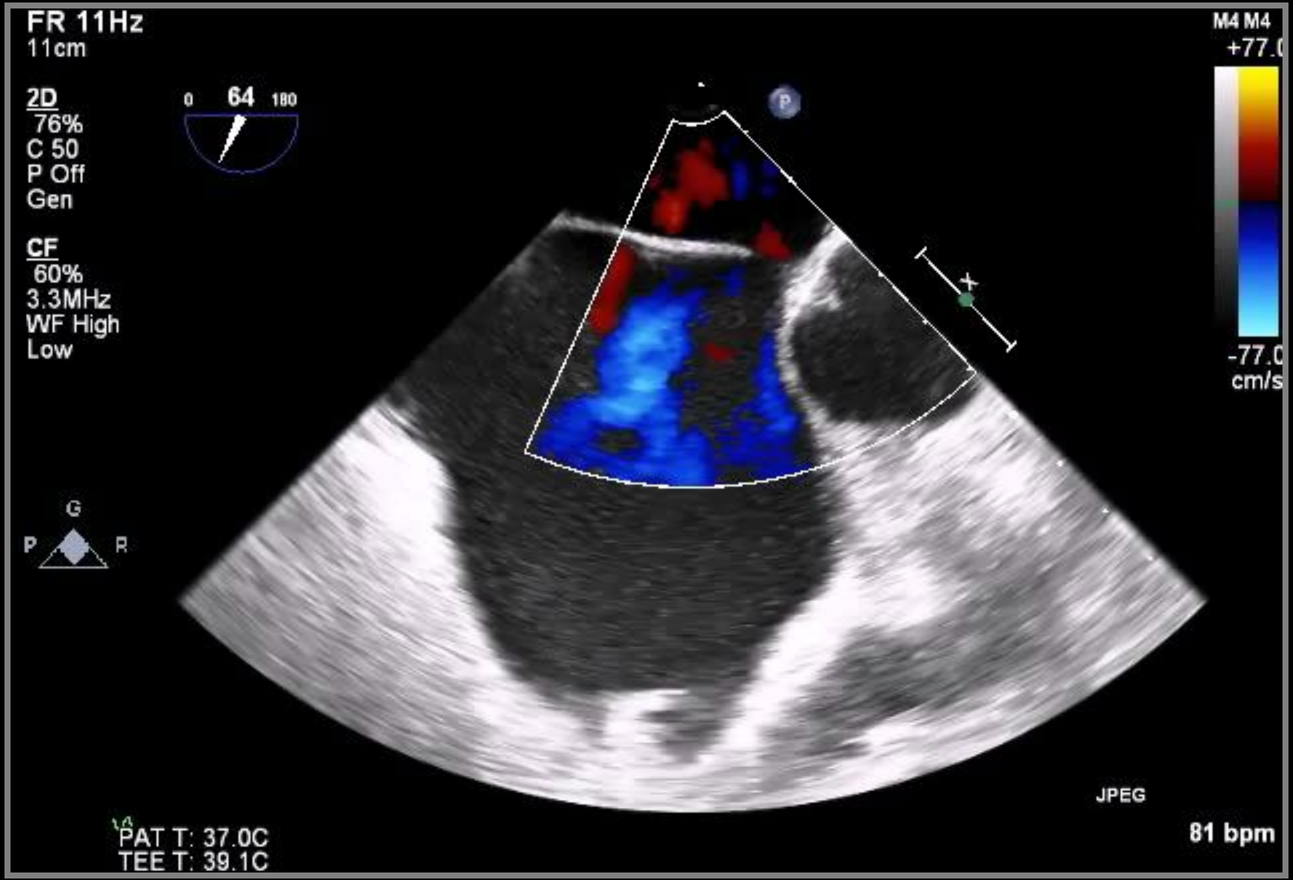


50 YO / M



FR 12Hz
19cm
2D
76%
C 50
P Low
HPen
CF
66%
2.5MHz
WF High
Med
M3 M4
+69.3
-69.3
cm/s
G
R
1.4
2.8
JPEG
71 bpm

50 YO / M


FR 11Hz
11cm
2D
76%
C 50
P Off
Gen
CF
60%
3.3MHz
WF High
Low
0 64 180
M4 M4
+77.0
-77.0
cm/s
G
P R
JPEG
81 bpm
PAT T: 37.0C
TEE T: 39.1C

50 YO / M

FR 35Hz
9.0cm
4:47:03
M4
2D
69%
C 50
P Off
Gen
0 64 180
G
P R
Dist 1.53 cm
76bpm

50 YO / M


FR 34Hz
6.0cm
Full Volume
3D 40%
3D 40dB
0
90
180
M4
JPEG
78 bpm
PAT T: 37.0C
TEE T: 38.1C

Anatomic Assessment

Associated anomalies

- Patent foramen ovale/atrial septal defect
- RV inflow and outflow tract obstruction
- VSD
- Accessory conduction pathway (WPW syndrome), increasing risk of atrial tachycardia
- PS
- TOF
- CoA
- Mitral valve abnormalities

Surgical Intervention

- **Surgeons with training and expertise in CHD should perform tricuspid valve repair or replacement with concomitant closure of an ASD, when present, for patients with Ebstein's anomaly with the following indications:**

  - **Symptoms** or deteriorating exercise capacity. (*Level of Evidence: B*)
  - **Cyanosis** (oxygen saturation less than 90%). (*Level of Evidence: B*)
  - **Paradoxical embolism**. (*Level of Evidence: B*)
  - Progressive **cardiomegaly** on chest x-ray. (*Level of Evidence: B*)
  - Progressive **RV** dilation or reduction of RV systolic function. (*Level of Evidence: B*)

Circulation. 2008;118:2395-2451

Surgical Intervention

Indications	Class[a]	Level[b]
Indications for surgery		
• Surgical repair should be performed in patients with more than moderate TR and symptoms (NYHA class >II or arrhythmias) or deteriorating exercise capacity measured by CPET	I	C
• If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair	I	C
• Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest X-ray	IIa	C

EHJ 2010;31:2915–2957

Surgery vs. Observation

Decreased RV function

Surgery vs. Observation

Decreased RV function

75 years old

Natural Course in Exercise Capacity

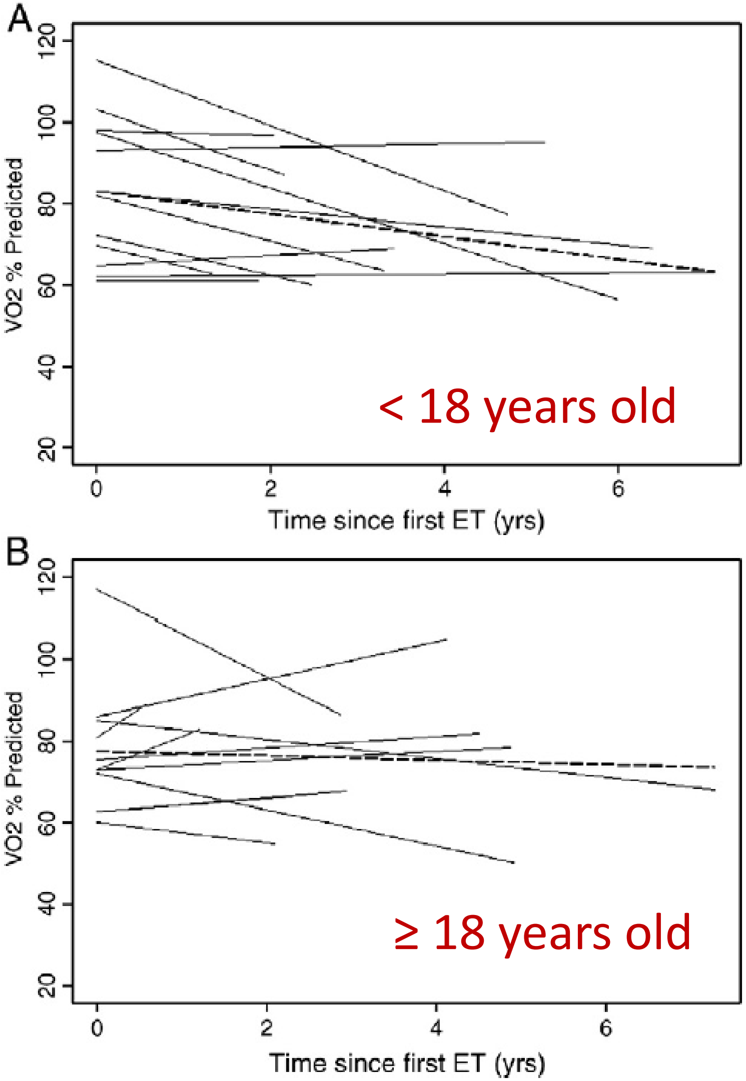


Table I. Demographics

Characteristic	
Male gender, n (%)	13 (57)
Age at first CPX (y), median (range)	17.9 (8.1-52.5)
Time from first to last CPX (y), median (range)	3.3 (0.6-7.3)
No. of CPX	
2	12 (52%)
3	6 (26%)
4	3 (13%)
5	2 (9%)
Mean ± SD	2.8 ± 1.0
Severe tricuspid regurgitation*	9 (39%)
Atrial level right to left shunting †	7 (30%)
ES grade[1,10]	
1	7 (32%)
2	10 (45%)
3	4 (18%)
4	1 (5%)

N = 23 patients; 64 exercise tests.
*Based on echocardiogram within 6 months of the first exercise test.
†Oxygen saturation at rest or with exercise ≤93%.

Am Heart J 2012;163:486-91

Natural Course in Exercise Capacity

Table V. Average rate of change per year for Vo_2% and O_2 pulse%

	Vo_2%: change/y			O_2 pulse%: change/y		
	Mean ± SD	**P***	**P†**	**Mean ± SD**	**P***	**P†**
Overall (n = 23)	−1.87 ± 8.04	.05		−2.49 ± 9.96	.03	
Age at initial CPX						
Age <18 y (n = 12)	−3.04 ± 6.78	.01	.15	−4.06 ± 9.33	.01	.12
Age ≥18 y (n = 11)	−0.43 ± 8.79	.77		−0.58 ± 9.88	.73	
Tricuspid regurgitation						
Mild or moderate (n = 14)	−1.59 ± 8.85	.18	.57	−1.92 ± 8.93	.11	.44
Severe (n = 9)	−2.67 ± 6.92	.11		−4.11 ± 11.56	.13	
ES grade						
1 (n = 7)	−1.91 ± 9.87	.36	.82	−2.26 ± 8.46	.22	.84
2 (n = 10)	−3.43 ± 7.94	.04		−3.8 ± 12.48	.12	
3 or 4 (n = 5)	0.02 ± 4.23	.99		−1.64 ± 6.53	.30	

Overall and by age, severity of tricuspid regurgitation, and ES grade. Other abbreviations as in Table III.
* *P* value from 1 sample *t* test comparing mean slope to 0.
† *P* value from 2 sample *t* test comparing mean slopes in the groups.

Am Heart J 2012;163:486-91

Functional Status after Surgery

21 patients with Ebstein anomaly (between 6 and 59 years of age)

TABLE 1. Changes in CPET: Comparing Baseline Testing with Follow-up After Surgical Intervention

		Baseline test	Follow-up	*P* values*
Sex	♂/♀	5/16	5/16	–
Age	Years	24.5 (13.7 – 43.6)	26.7 (14.8 – 44.9)	–
Tricuspid regurgitation	I/II/III/IV	0 / 2 / 16 / 3	9 / 10 / 2 / 0	< .001
RER	Peak	1.07 (1.03 – 1.13)	1.07 (1.03 – 1.13)	.501
SpO_2† (%)	Rest	98 (95 – 99)	99 (97 – 100)	**.004**
	Peak	92 (84 – 98)	97 (95 – 100)	**.010**
Peak Vo_2	mL/min/kg	21.0 (17.6 – 23.5)	20.7 (18.8 – 27.9)	**.009**
	% of predicted	68.4 (52.2 – 83.3)	77.3 (57.2 – 91.4)	**.009**
V_E/V_{CO_2}	Slope	32.5 (30.3 – 43.8)	29.3 (27.8 – 33.1)	**.001**

CPET, Cardiopulmonary exercise test; *RER*, respiratory exchange ratio. *2-sided Wilcoxon test. †SpO_2 could be assessed in only 20 patients.

J Thorac Cardiovasc Surg 2011;141:1192-5

Functional Status after Surgery

21 patients with Ebstein anomaly (between 6 and 59 years of age)

TABLE 2. Changes in CPET variables during follow-up by patients undergoing primary surgery and patients being reoperated

		Primary surgery (group 1) n = 14, Median (Q1-Q3)			Reoperation (group 2) n = 7, Median (Q1-Q3)		
		Baseline test	Follow-up	*P* values*	Baseline test	Follow-up	*P* values*
Sex	♂/♀	3/11			2/5		.557†
Age	Years	18.3 (10.8–46.3)			31.1 (21.1–41.3)		.224‡
TR	I/II/III/IV	0/2/11/4	5/8/1/0	**.002**	0/0/5/2	4/2/1/0	**.014**
RER	Peak	1.04 (1.01–1.09)	1.04 (1.01–1.12)	.248	1.12 (1.08–1.23)	1.10 (1.07–1.16)	.672
SpO_2 (%)§	Rest	95 (92–98)	99 (99–100)	**.003**	99 (98–100)	99 (98–100)	.279
	Peak	88 (78–94)	99 (97–100)	**.003**	99 (98–100)	99 (98–100)	.317
peak VO_2	mL/min/kg	21.0 (17.8–22.1)	20.3 (18.5–27.8)	.059	20.3 (16.4–24.1)	21.3 (18.5–29.5)	.091
	% predicted	68.8 (51.0–83.0)	77.8 (58.0–92.1)	.064	56.4 (53.1–86.7)	73.6 (55.5–92.1)	.063
V_E/VCO_2	Slope	32.3 (30.8–50.5)	29.4 (27.8–32.9)	**.005**	32.5 (30.0–36)	29.3 (26.5–34.5)	**.018**

CPET, Cardiopulmonary exercise test; *RER*, respiratory exchange ratio; *TR*, tricuspid regurgitation. *2-sided Wilcoxon test. †Chi-squared test comparing the sex distribution of the 2 subgroups. ‡2-sided Mann-Whitney-U test comparing the age of the 2 subgroups at baseline test. §SpO_2 saturation could be assessed in only 20 patients.

J Thorac Cardiovasc Surg 2011;141:1192-5

Functional Status after Surgery

Functional Status After Operation for Ebstein Anomaly

The Mayo Clinic Experience

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Rochester, Minnesota

Objectives The objective of this study was to review the long-term functional outcome of patients with Ebstein anomaly who had cardiac operation at our institution.

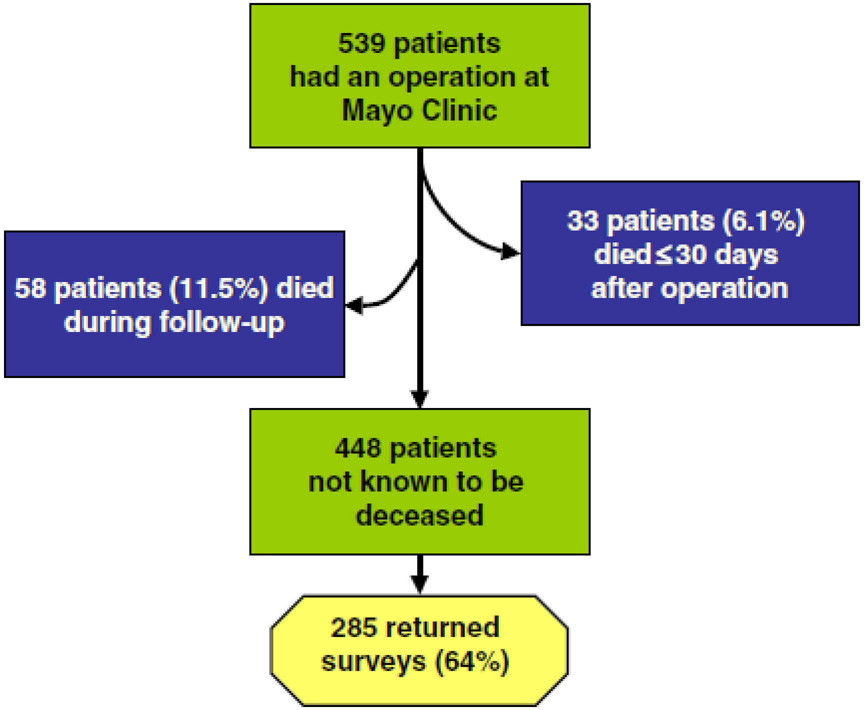
Background Ebstein anomaly is a spectrum of tricuspid valvular and right ventricular dysplasia. Many patients will require operation in an attempt to improve quality of life.

Methods From April 1, 1972, to January 1, 2006, 539 patients with Ebstein anomaly underwent 604 cardiac operations at the Mayo Clinic in Rochester, Minnesota. Patient records were reviewed, and all patients known to still be alive were mailed a medical questionnaire or contacted by telephone.

Results At the initial operation at our institution, the mean age of the patients was 24 years (range 8 days to 79 years) and 53% were female patients. Survival at 5, 10, 15, and 20 years was 94%, 90%, 86%, and 76%, respectively. Survival free of late reoperation was 86%, 74%, 62%, and 46% at 5, 10, 15, and 20 years, respectively. Surveys were returned by 285 of 448 (64%) patients known to be alive at the time of this study. Two hundred thirty-seven (83%) patients were in New York Heart Association functional class I or II, and 34% were taking no cardiac medication. One hundred three patients (36%) reported an incident of atrial fibrillation or flutter, 5 patients (2%) reported having had endocarditis, and 1 patient (<1%) reported having a stroke. There were 275 pregnancies among 82 women. The recurrence of congenital heart disease was reported in 9 of 232 (3.9%) liveborn children.

Conclusions Patients have good long-term survival and functional outcomes after undergoing surgery for Ebstein anomaly. Atrial arrhythmias are common both before and after surgery. Many patients have had one or more successful pregnancies with a low-recurrence risk of congenital heart disease. (J Am Coll Cardiol 2008;52:460–6)

Functional Status after Surgery



Health Status Questionnaires

NYHA functional class

- I (43%)
- II (40%)
- III (12%)
- IV (4%)

J Am Coll Cardiol 2008;52:460–6

Functional Status after Surgery

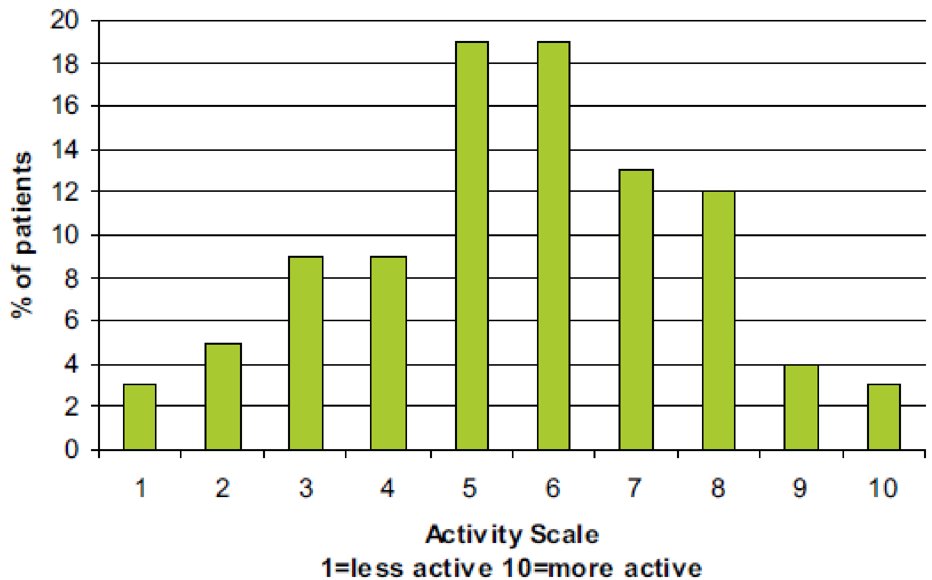


Figure 3 Self-Reported Activity Scale After Operation for Ebstein Anomaly

Shown are the percentage of survey responders who rated their exercise tolerance related to other people their own age on a scale of 1 to 10.

Table 3 Self-Reported Exercise Tolerance

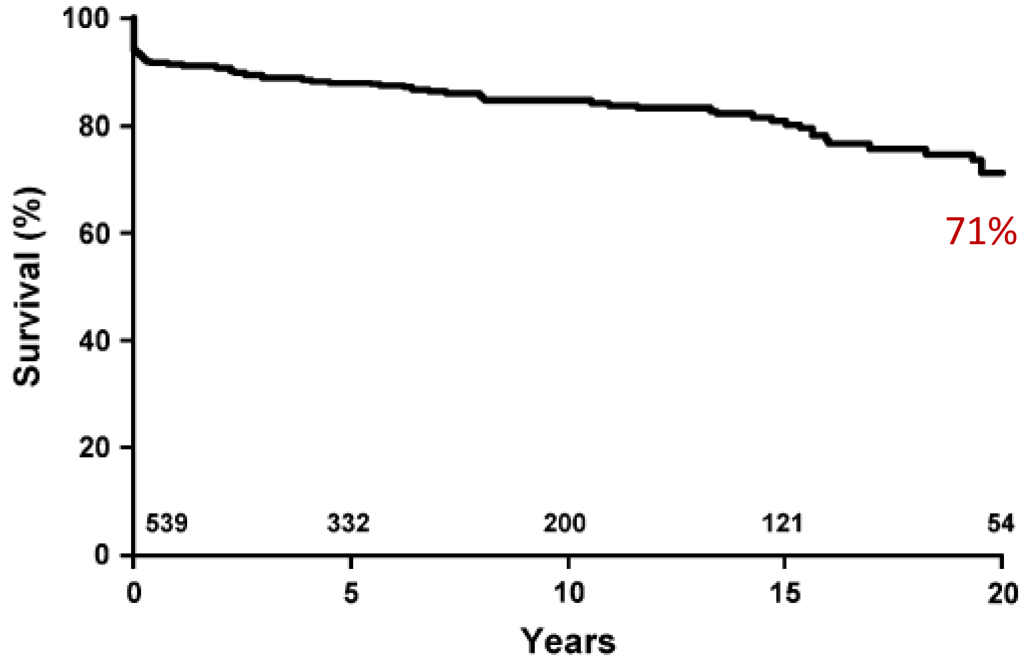
My Ability of Exercise Relative to Peers Is	n = 285	%
Much greater	7	2.5
Slightly greater	29	10.2
About the same	105	36.8
Slightly less	89	31.2
Much less	43	15.1
I am unable to exercise	5	1.8
Unknown	7	2.5

J Am Coll Cardiol 2008;52:460–6

Survival after Surgery

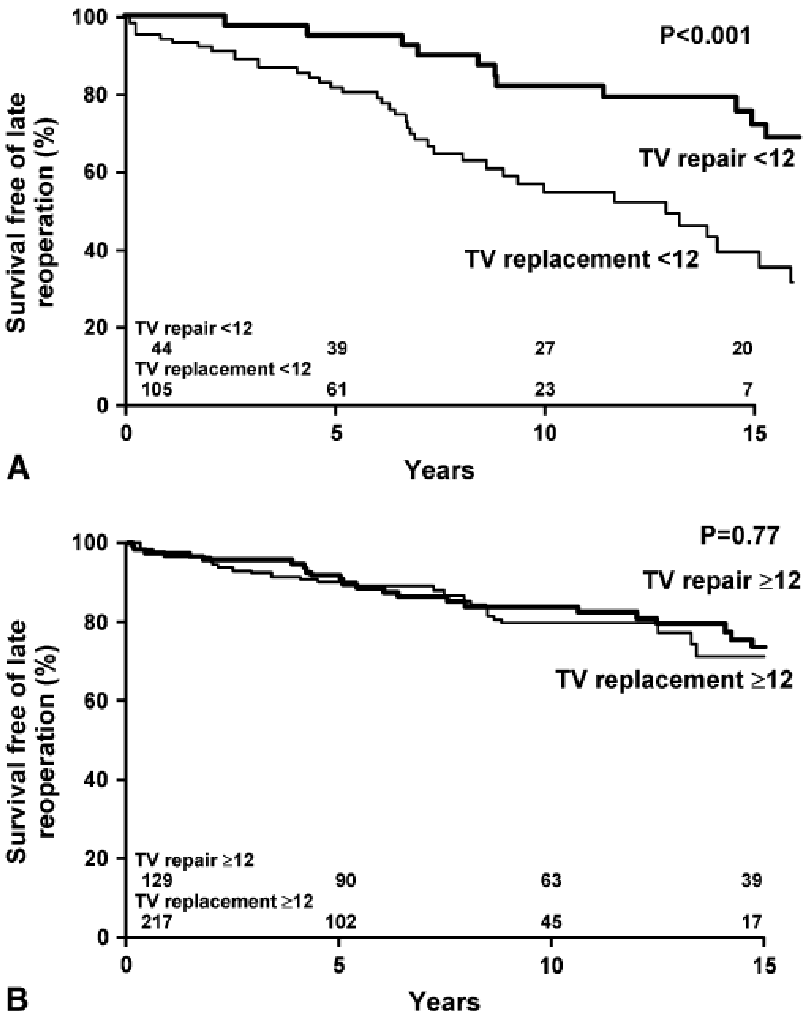
539 Patients with mean [range] age at surgery 24 years [8 days to 79 years]

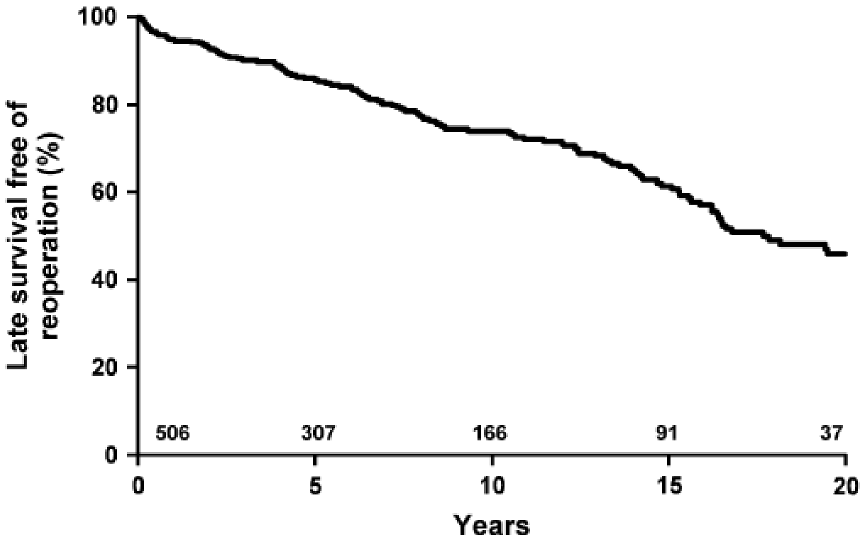
Early mortality: 4.8%



JTCVS 2008;135:1120-36

Reoperation after Surgery





JTCVS 2008;135:1120-36

50 Years of Age or Older

11 patients were > 60 years of age

Table 1 Clinical Characteristics

Patient characteristics (n = 81)	
Age at diagnosis, yrs	43 ± 19
Women	51 (63)
Pre-procedural characteristics (n = 89)	
Patient age at surgery, yrs	59 ± 8
New York Heart Association functional class	
I/II	13 (15)
III/IV	76 (85)
Dyspnea	74 (83)
Palpitations	69 (78)
Edema	30 (34)
Cyanosis	21 (24)
Stroke/TIA	21 (24)
Dizziness	16 (18)
Right-sided heart failure	13 (15)
Syncope	7 (8)
Clubbing	6 (7)

Values are mean ± SD or n (%).
TIA = transient ischemic attack.

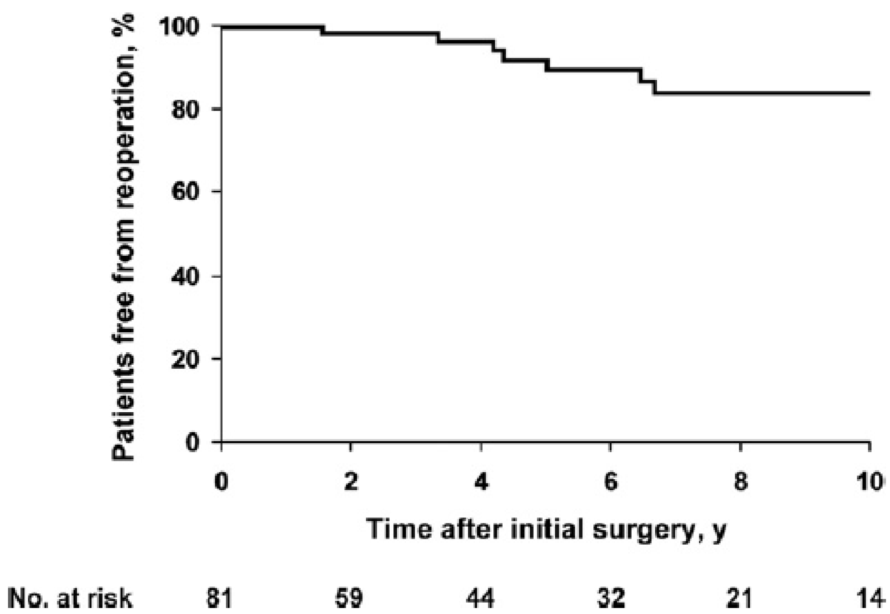
Table 2 Surgical Procedures Performed (N = 89)

Procedure Type	No. of Procedures (%)
Tricuspid valve operation	
Repair	22 (25)
Replacement	65 (73)
None	2 (2)
ASD/PFO closure	58 (65)
Right reduction atrioplasty	41 (46)
Anterior right pericardectomy	18 (20)
Plication of atrialized RV	12 (13)
Right-sided maze procedure	18 (20)
Ablation of accessory pathway	9 (10)
CABG	9 (10)
Mitral valve surgery	6 (7)
Repair	4 (5)
Replacement	2 (2)
Bidirectional Glenn operation	2 (2)
Aortic root surgery	2 (2)
Repair of PS	1 (1)
Permanent pacing	1 (1)

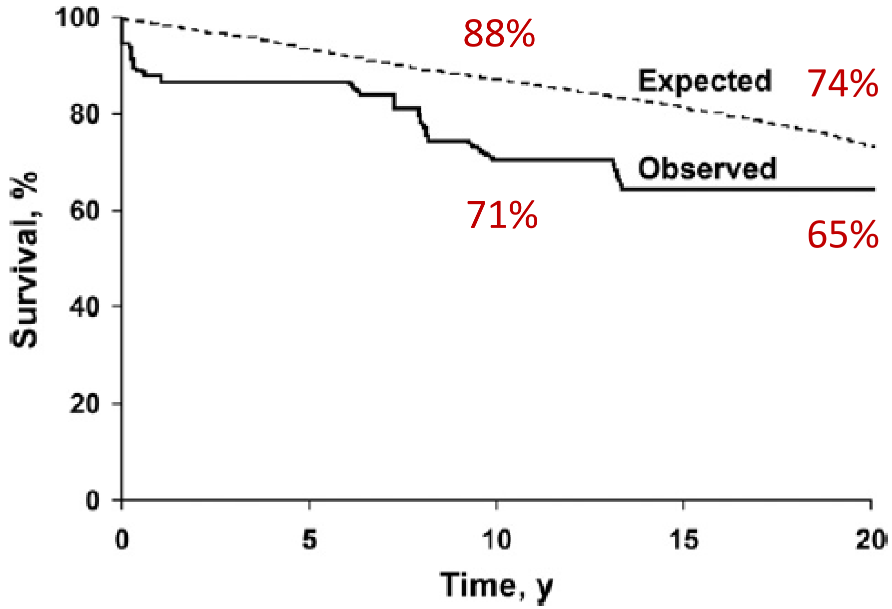
ASD = atrial septal defect; CABG = coronary artery bypass grafting; PFO = patent foramen ovale; PS = pulmonary stenosis; RV = right ventricle.

J Am Coll Cardiol 2012;59:2101–6

50 Years of Age or Older



Early mortality: 4%



For the 71 patients with clinical follow-up, improvement in functional class occurred in 63 patients **(89%)**. Postoperatively, only 8 patients remained in functional class III or IV (11%).

J Am Coll Cardiol 2012;59:2101–6

50 Years of Age or Older

Table 4 Univariate Predictors of Death During Follow-up

Parameter	HR (95% CI)	p Value
No post-operative improvement	10.0 (3.01–33.4)	<0.001
Pre-operative history of heart failure	4.42 (1.42–13.7)	0.01
Pre-operative LVEF <50%	3.59 (1.20–10.7)	0.02
Diabetes mellitus	6.76 (1.43–31.9)	0.02
Pulmonary hypertension	1.21 (0.36–4.08)	0.75
History of atrial fibrillation	0.95 (0.26–3.46)	0.94
Tricuspid valve replacement	1.63 (0.44–5.99)	0.46
Male	1.96 (0.65–5.89)	0.23
Age at surgery, per 10 yrs	2.53 (1.28–5.00)	0.007

CI = confidence interval; HR = hazard ratio; LVEF = left ventricular ejection fraction.

J Am Coll Cardiol 2012;59:2101–6

Surgery + Maze operation
Vs.
Catheter ablation

Catheter Intervention

- *Class I*
  - Adults with Ebstein's anomaly should have catheterization performed at centers with expertise in catheterization and management of such patients. (*Level of Evidence: C*)
- *Class IIa*
  - Catheter ablation can be beneficial for treatment of recurrent supraventricular tachycardia in *some patients* with Ebstein's anomaly. (*Level of Evidence: B*)

Circulation. 2008;118:2395-2451

Catheter Intervention

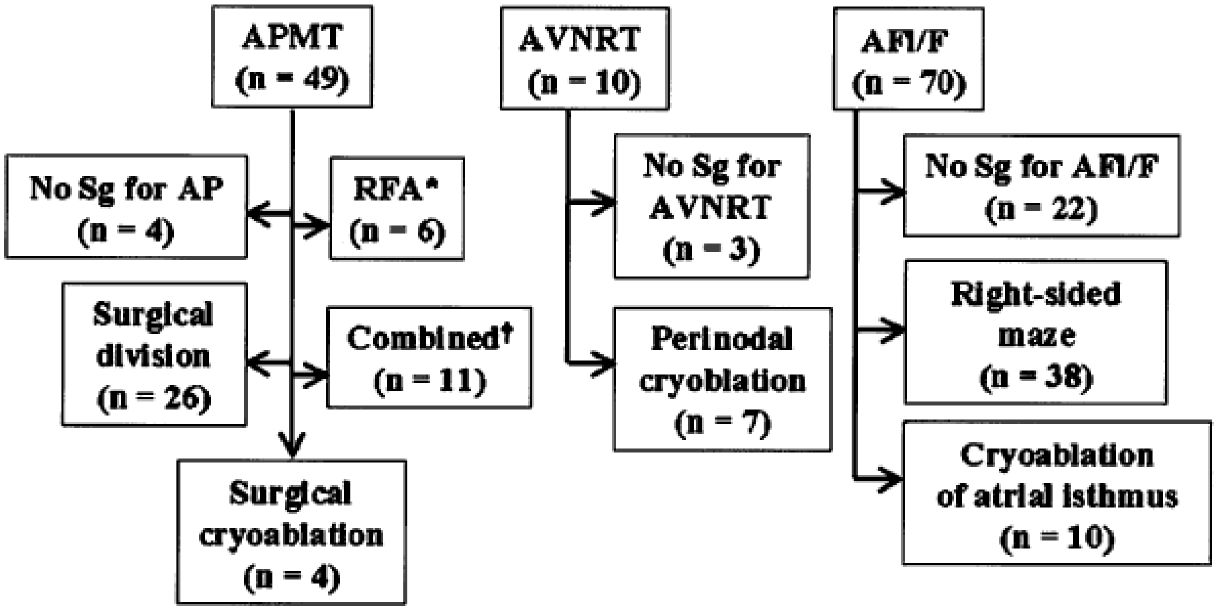
Indications for catheter intervention		
• Patients with relevant arrhythmias should undergo electrophysiologic testing, followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery	I	C
• In the case of documented systemic embolism probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered	IIa	C
• If cyanosis (oxygen saturation at rest <90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention (see text)	IIb	C

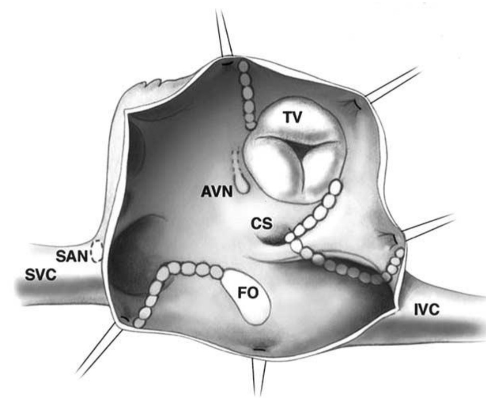
EHJ 2010;31:2915–2957

Accessory Pathway Mediated SVT

- Localization of accessory pathways is often challenging & > 50 % of patients have multiple accessory pathways
- The success rate for catheter ablation is lower in Ebstein anomaly (≤81 %) patients compared with patients with structurally normal hearts (≥95 %)
- Rarely, right coronary artery stenosis has been seen following catheter ablation

Ebstein Surgery & Maze





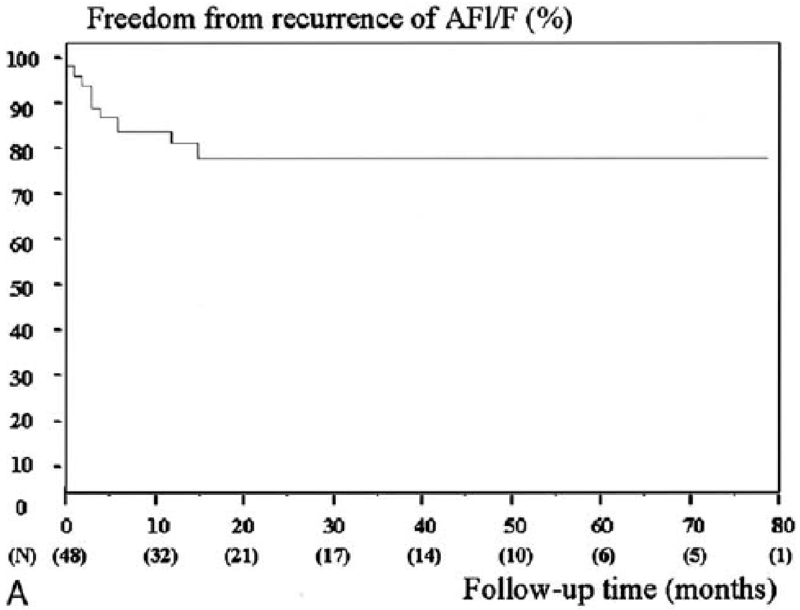
JTCVS 2004;128:826-33

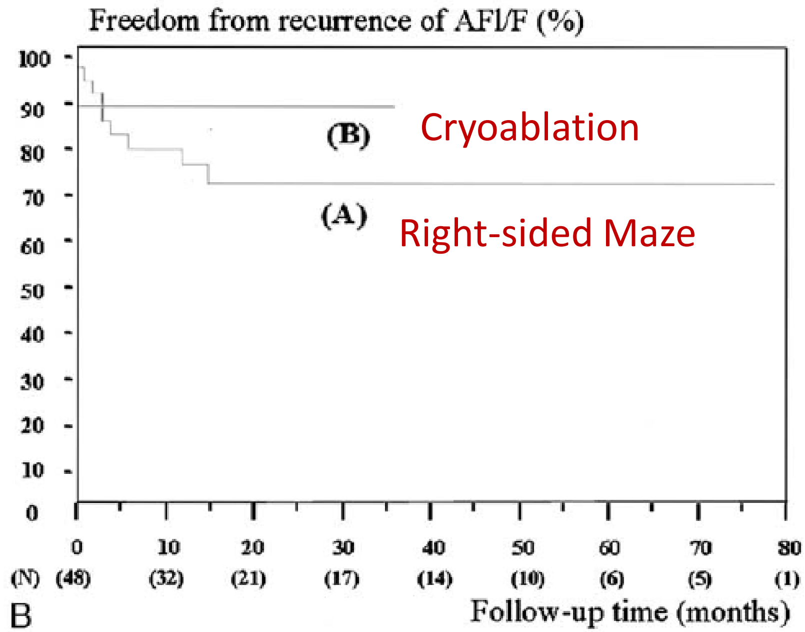
Circulation. 2015;131:1110-1118

Ebstein Surgery & Maze

Recurrence of atrial fibrillation

- 6/14 (43.0%) without Maze op
- 12/48 (25.6%) with Maze or cryoablation





JTCVS 2004;128:826-33

Circulation. 2015;131:1110-1118

Ebstein Surgery & Maze

86 patients who had corrective surgery and concomitant maze procedure

Table 5. Late Outcomes According to Type of Preoperative Arrhythmia

Outcome	Paroxysmal	Persistent	*p* Value
No AFl/F + no AAM			
RSM	88%	71%	0.17
BAM	77%	86%	0.08
	0.08[a]	0.053[a]	
Freedom from warfarin			
RSM	83%	86%	0.87
BAM	75%	71%	0.86
	0.47[a]	0.31[a]	

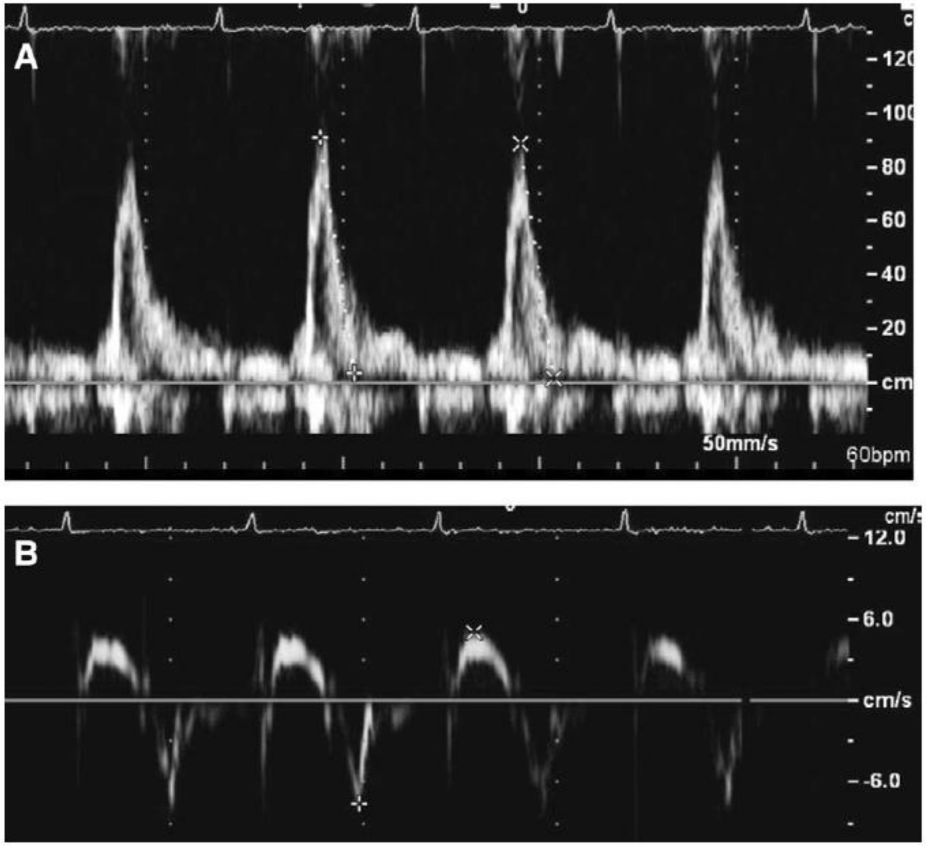
[a] Probability value for column comparison.

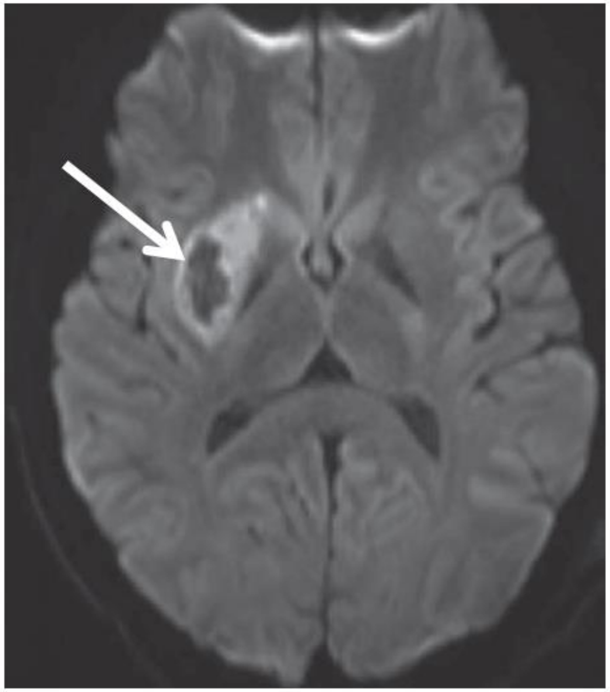
AAM = antiarrhythmic medications; AFl/F = atrial flutter or fibrillation; BAM = biatrial maze; RSM = right-sided maze.

Ann Thorac Surg 2015;99:1700–5

Ebstein Surgery & Maze

The absence of LAMF after the Cox maze III procedure is as high as 39% and has been demonstrated to persist up to 56 months.





Circulation. 2015;131:1110-1118

What to Do for a 75 Year Old Man of Severe Ebstein Anomaly with Decreased RV Function, Atrial Fibrillation

Surgery + Maze operation
Anticoagulation

Earlier surgery should have been considered.

Thank you for your attention.