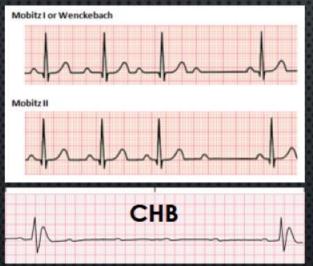
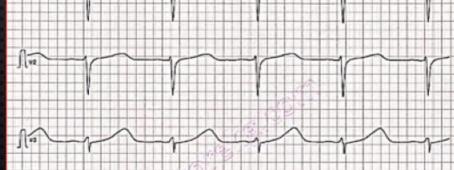
MANAGEMENT OF CARDIAC ARRHYTHMIAS AND CONDUCTION DISORDERS-2018









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56 y/o male with 2 vessel ischemic heart disease (cath 2 months before) presents from the ER with unexplained syncope. Patient is then transferred to CCU and myocardial infarction is ruled out.

Patient is then referred for electrophysiology testing and found to have **inducible sustained monomorphic VT** on electrophysiological study which of the following best describes his appropriate care.





BOR

QUESTION 1

- Patient needs further electrical monitoring with beta blocker treatment.
- 2. Patient should have ICD implanted ... a class 1 indication
- Patient instructed to have further follow up with primary physician with global risk reduction
- Patient requires echocardiogram to further assess his need for ICD implant
- Patient needs no further treatment except for his ischemic heart disease





6.1. Ischemic Heart Disease

ICD implant

6.1.1. Secondary Prevention of SCD in Patients With Ischemic Heart Disease

Recommendations for Secondary Prevention of SCD in Patients With Ischemic Heart Disease

References that support the recommendations are summarized in Online Data Supplement 17 and 18.

COR LOE Recommendations		Recommendations		
	B-R	1. In patients with ischemic heart disease, who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) (1-4) or stable VT (LOE: B-		
1	B-NR	NR) (5) not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.		
State Interr Va	alue ement: nediate alue E: B-R)	 A transvenous ICD provides intermediate value in the secondary prevention of SCD particularly when the patient's risk of death due to a VA is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status (6). 		
ı	B-NR	 In patients with ischemic heart disease and unexplained syncope who have inducible sustained monomorphic VT on electrophysiological study, an ICD is recommended if meaningful survival of greater than 1 year is expected (7). 		



Mr Gomez is asymptomatic with neuromuscular disorder and ask if he needs a ICD, your recommendation would be

- 1. He is not a candidate for ICD because he is asymptomatic
- Mr. Gomez needs heart catheterization to evaluate his CAD status first before considering ICD
- 3. Patients with neuromuscular disorder, primary and secondary prevention, ICDs are recommended
- 4. Patient needs EP study and depending on results may need ICD
- Patient would need to have documented complete heart block before further work up





6.8. Neuromuscular Disorders

ICD

Recommendations for Neuromuscular Disorders

References that support the recommendations are summarized in Online Data Supplement 38.

COR	LOE	Recommendations	
1	B-NR	 In patients with neuromuscular disorders, primary and secondary prevention ICDs are recommended for the same indications as for patients with NICM if meaningful survival of greater than 1 year is expected (1, 2). 	
lla	B-NR	 In patients with Emery-Dreifuss and limb-girdle type IB muscular dystrophies with progressive cardiac involvement, an ICD is reasonable if a meaningful survival of greater than 1 year is expected (3-8). 	
lla	B-NR	 In patients with muscular dystrophy, follow-up for development of cardiac involvement is reasonable, even if the patient is asymptomatic at presentation (9-12). 	
IIb	B-NR	 In patients with myotonic dystrophy type 1 with an indication for a permanent pacemaker, an ICD may be considered to minimize the risk of SCA from VT if meaningful survival of greater than 1 year is expected (9, 13, 14). 	

Table 9. Neuromuscular Disorders Associated With Heart Disease

Muscular Dystrophy	Inheritance	Gene/ Protein Affected	Primary Cardiac Pathology	Frequency of Cardiac Involvement	Causes of Death	Associated With Sudden Death?
Duchenne	X-linked recessive	Dystrophin	NICM	>90%	Respiratory, HF	Yes, uncertain etiology
Becker	X-linked recessive	Dystrophin	NICM	60%-75%	HF, respiratory	Yes, uncertain etiology
Limb-girdle type 1B	Autosomal dominant	Lamin A/C	Conduction system disease and NICM	>90%	Sudden, HF	Yes
Limb-girdle type 2C-2F	Autosomal recessive	Sarcoglycan	NICM	<25%	Respiratory, HF	Uncertain
Limb-girdle type 21	Autosomal recessive	Fukutin- related protein	NICM	20%-80%	Respiratory, HF	Uncertain
Myotonic type 1	Autosomal dominant	CTG repeat expansion	Conduction system disease and NICM	60%-80%	Respiratory, sudden, HF	30% of deaths, uncertain bradycardia versus tachycardia
Myotonic type 2	Autosomal dominant	CCTG repeat expansion	Conduction system disease	10%-25%	Normal causes	Reported
Emery-Dreifuss	X-linked and autosomal dominant or recessive	Emerin, Lamin A/C	Conduction system disease and NICM	>90%	Sudden, HF	Yes
Facioscapulohu meral	Autosomal dominant	D4Z4 repeat contaction	Possibly conduction disease	5%-15%	Normal causes, respiratory rarely	Not reported

HF indicates heart failure; and NICM, nonischemic cardiomyopathy. Adapted with permission from Groh, et al. (15).



Miss Johnson is a asymptomatic 34 y/o women who presents for breast biopsy and EKG finds long QT interval >470 ms which best describes best treatment option

- 1. Patient needs to be started on beta blocker and followed for symptoms
- 2. Patient needs no further treatment since asymptomatic
- 3. Patient is a candidate for biventricular pacemaker set at heart rate faster than her intrinsic rate
- 4. Patient is a candidate for ICD
- 5. Patient needs ETT first to make further decisions



6.9.1.1. Congenital Long QT Syndrome

			-
		Recommendations for Long QT Syndrome]
		at support the recommendations are summarized in Online Data Supplement 40.	1
COR	COR LOF Recommendations		
1	B-NR	 In patients with long QT syndrome with a resting QTc greater than 470 ms, a beta blocker is recommended (1-5). 	
į	B-NR	 In high-risk patients with symptomatic long QT syndrome in whom a beta blocker is ineffective or not tolerated, intensification of therapy with additional medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation, and/or an ICD is recommended (2, 6-12). 	1
ı	B-NR	 In patients with long QT syndrome and recurrent appropriate ICD shocks despite maximum tolerated doses of a beta blocker, intensification of medical therapy with additional medications (guided by consideration of according to the particular long QT syndrome type) or left cardiac sympathetic denervation, is recommended (6, 7, 10, 13-16). 	
(1)	B-NR	 In patients with clinically diagnosed long QT syndrome, genetic counseling and genetic testing are recommended (17-21). 	
lla	B-NR	 In patients with suspected long QT syndrome, ambulatory electrocardiographic monitoring, recording the ECG lying and immediately on standing, and/or exercise treadmill testing can be useful for establishing a diagnosis and monitoring the response to therapy (22-29). 	
lla	B-NR	 In asymptomatic patients with long QT syndrome and a resting QTc less than 470 ms, chronic therapy with a beta blocker is reasonable (3, 30, 31). 	
IIb	B-NR	7. In asymptomatic patients with long QT syndrome and a resting QTc greater than 500 ms while receiving a beta blocker, intensification of therapy with medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation or an ICD may be considered (2, 8, 11, 30).	
III: Harm	B-NR	 In patients with long QT syndrome, QT-prolonging medications are potentially harmful (5, 12, 32-34). 	



HIGH RISK FEATURES: CONGENITAL LONG QT SYNDROME

- LQTS include those with QTc >500 ms
- 2. Genotypes LQT2 and LQT3
- 3. Females with genotype LQT2
- 4. <40 years of age
- 5. Onset of symptoms at <10 years of age
- 6. Patients with recurrent syncope



Figure 9. Prevention of SCD in Patients With Long QT Syndrome QT prolonging drugs/ LQTS hypokalemia/ hypomagnesemia Resuscitated (Class III: Harm) cardiac arrest QTc <470 ms QTc ≥470 ms and/ or symptomatic ICD Beta blocker candidate* (Class I) Beta blocker Beta blocker (Class IIa) (Class I) ICD (Class I) Persistent symptoms Asymptomatic and Recurrent ICD and/or other high-risk QTc >500 ms shocks for VT features† Treatment intensification: Treatment intensification: Treatment intensification: additional medications, additional medications, additional medications, left cardiac sympathetic left cardiac sympathetic left cardiac sympathetic

denervation and/or an ICD

(Class I)

denervation

(Class I)



denervation and/or an ICD

(Class IIb)



Mr Phillips is having ETT and develops this rhythm



- 1. Patient has monomorphic VT and needs CCU admission
- 2. Patient needs ICD immediately
- Patient has exercise-induced polymorphic VT in catecholaminergic polymorphic ventricular tachycardia and needs beta blocker (class 1 indication)
- 4. Patient will not benefit from beta blocker and is contraindicated
- Patient needs iv amiodarone





6.9.1.2. Catecholaminergic Polymorphic Ventricular Tachycardia

Recommendations for Catecholaminergic Polymorphic Ventricular Tachycardia

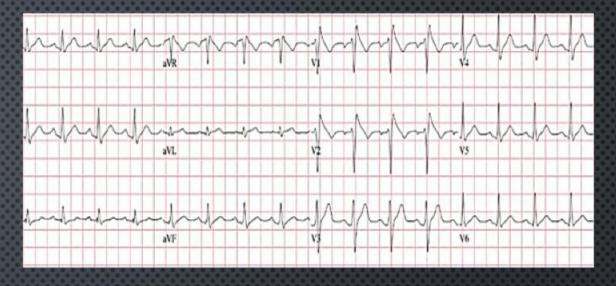
References that support the recommendations are summarized in Online Data Supplement 41.

COR	LOE	Recommendations
1	B-NR	 In patients with catecholaminergic polymorphic ventricular tachycardia, a beta blocker is recommended (1, 2).
1	B-NR	 In patients with catecholaminergic polymorphic ventricular tachycardia and recurrent sustained VT or syncope, while receiving adequate or maximally tolerated beta blocker, treatment intensification with either combination medication therapy (e.g., beta blocker, flecainide), left cardiac sympathetic denervation, and/or an ICD is recommended (2-6).
lla	B-NR	 In patients with catecholaminergic polymorphic ventricular tachycardia and with clinical VT or exertional syncope, genetic counseling and genetic testing are reasonable (7).





43 y/o male brought by ambulance to ER after apparent cardiac arrest, no chest pain, negative troponin and the following EKG, your diagnosis



- 1. Takotsubo heart disease
- 2. Brugada syndrome and has class 1 indication for ICD
- 3. Right ventricular infarction
- 4. Non STEMI
- 5. Left bundle branch block



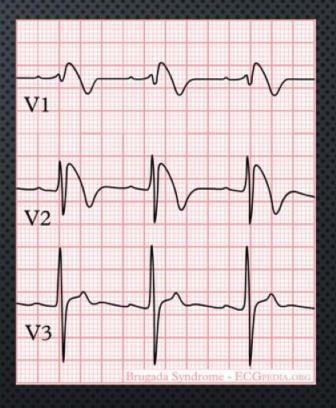
6.9.1.3. Brugada Syndrome

Recommendations for Brugada Syndrome

References that support the recommendations are summarized in Online Data Supplement 42 and Systematic Review Report.

- 1-	Systematic neview neport.					
	COR	LOE	Recommendations			
	1	B-NR	 In asymptomatic patients with only inducible type 1 Brugada electrocardiographic pattern, observation without therapy is recommended (1-5). 			
	1	B-NR	 In patients with Brugada syndrome with spontaneous type 1 Brugada electrocardiographic pattern and cardiac arrest, sustained VA or a recent history of syncope presumed due to VA, an ICD is recommended if a meaningful survival of greater than 1 year is expected (4, 6). 			
	1	B-NR	 In patients with Brugada syndrome experiencing recurrent ICD shocks for polymorphic VT, intensification of therapy with quinidine or catheter ablation is recommended (7-11). 			
	1	B-NR	 In patients with spontaneous type 1 Brugada electrocardiographic pattern and symptomatic VA who either are not candidates for or decline an ICD, quinidine or catheter ablation is recommended (7, 9-11). 			
	lla	B-NR	 In patients with suspected Brugada syndrome in the absence of a spontaneous type 1 Brugada electrocardiographic pattern, a pharmacological challenge using a sodium channel blocker can be useful for diagnosis (12-14). In patients with asymptomatic Brugada syndrome and a spontaneous type 1 Brugada electrocardiographic pattern, an electrophysiological study with programmed ventricular stimulation using single and double extrastimuli may be considered for further risk stratification (1, 6, 13, 15-17). 			
	IIb	B-NR ^{sr}				
	IIb	C-EO	7. In patients with suspected or established Brugada syndrome, genetic counseling and genetic testing may be useful to facilitate cascade screening of			

Brugada syndrome is definitively diagnosed when a type 1 ST-segment is observed in >1 right precordial lead (V1 to V3)





49 y/o male presents with <u>atrial fibrillation</u> with hypertrophic cardiomyopathy with low CHA2DS2-VASc score

- 1. Patient does not require any anticoagulation since score is low
- 2. Patient requires ticagrelor
- 3. Anticoagulation is indicated independent of the CHA2DS2-VASc score
- 4. Anticoagulation is only needed if the EF < 35%



	Score		Adjusted Stroke Rate (% per y)
CHADS ₂		CHADS ₂ *	
Congestive HF	1	0	1.9
Hypertension	1	1	2.8
Age ≥75 y	1	2	4.0
Diabetes mellitus	1	3	5.9
Stroke/TIA/TE	2	4	8.5
Maximum score	6	5	12.5
		6	18.2
CHA ₂ DS ₂ -VASc	(CHA ₂ DS ₂ -VASct	
Congestive HF	1	0	0
Hypertension	1	1	1.3
Age ≥75 y	2	2	2.2
Diabetes mellitus	1	3	3.2
Stroke/TIA/TE	2	4	4.0
Vascular disease (prior MI, PAD, or aortic plaque)	1	5	6.7
Age 65-74 y	1	6	9.8
Sex category (i.e., female sex)	1	7	9.6
Maximum score	9	8	6.7
		9	15.20

TABLE 12 Summary of Recommendations for Specific Patient Groups and AF			
Recommendations	COR	LOE	References
Hypertrophic cardiomyopathy			
Anticoagulation is indicated in HCM with AF independent of the CHA ₂ DS ₂ -VASc score	1	В	(169,170)
Antiarrhythmic drugs can be useful to prevent recurrent AF in HCM. Amiodarone or disopyramide combined with a beta blocker or nondihydropyridine calcium channel antagonist are reasonable	lla	С	N/A
AF catheter ablation can be beneficial for HCM to facilitate a rhythm-control strategy when antiarrhythmics fail or are not tolerated	lla	В	(171-174)
Sotalol, dofetilide, and dronedarone may be considered for a rhythm-control strategy in HCM	IIb	С	(12)



Acute coronary syndrome-Atrial fibrillation

AF complicating ACS		
Urgent cardioversion of new-onset AF in the setting of ACS is recommended for patients with hemodynamic compromise, ongoing ischemia, or inadequate rate control	1	С
IV beta blockers are recommended to slow RVR with ACS and no HF, hemodynamic instability, or bronchospasm	1	С
With ACS and AF with CHA₂DS₂-VASc score ≥2, anticoagulation with warfarin is recommended unless contraindicated	1	С



Atrial fibrillation: watch out for low EF (diltiazem-probably safest)

Pulmonary diseases		
A nondihydropyridine calcium channel antagonist is recommended to control ventricular rate with AF and COPD	I.	С
Cardioversion should be attempted for patients with pulmonary disease who become hemodynamically unstable with new-onset AF	1	С
Heart failure		
A beta blocker or nondihydropyridine calcium channel antagonist is recommended for persistent or permanent AF in patients with $HFpEF$	1	В
In the absence of preexcitation, an IV beta blocker (or a nondihydropyridine calcium channel antagonist with HFpEF) is recommended to slow ventricular response to AF in the acute setting, with caution in patients with overt congestion, hypotension, or HFrEF	1	В
In the absence of pre-excitation, IV digoxin or amiodarone is recommended to control heart rate acutely	1	В
Assess heart rate during exercise and adjust pharmacological treatment in symptomatic patients during activity	1	С
Digoxin is effective to control resting heart rate with HFrEF	1	С



Postoperative cardiac and thoracic surgery			
A beta blocker is recommended to treat postoperative AF unless contraindicated	1	Α	
A nondihydropyridine calcium channel blocker is recommended when a beta blocker is inadequate to achieve rate control with postoperative AF	1	В	

WPW and pre-excitation syndromes		
Cardioversion is recommended for patients with AF, WPW syndrome, and RVR who are hemodynamically compromised	1	
IV procainamide or ibutilide to restore sinus rhythm or slow ventricular rate is recommended for patients with pre-excited AF and RVR who are not hemodynamically compromised	1	
Catheter ablation of the accessory pathway is recommended in symptomatic patients with pre-excited AF, especially if the accessory pathway has a short refractory period	1	
IV amiodarone, adenosine, digoxin, or nondihydropyridine calcium channel antagonists in patients with WPW syndrome who have pre-excited AF is potentially harmful	III: Harm	



*Catheter ablation is only recommended as first-line therapy for patients with paroxysmal AF (Class IIa recommendation).

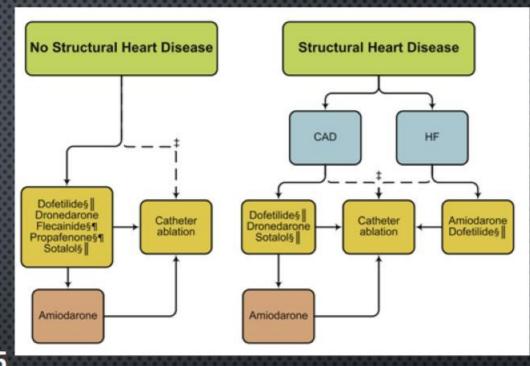
†Drugs are listed alphabetically

‡Depending on patient preference when performed in experienced centers.

§Not recommended with severe LVH (wall thickness >1.5 cm).

||Should be used with caution in patients at risk for torsades de pointes ventricular tachycardia.

¶Should be combined with AV nodal blocking agents.



Atrial fibrillation





- ICD therapy is recommended for primary prevention of SCD to reduce total mortality in selected patients with nonischemic DCM or ischemic heart disease at least 40 days post-MI with LVEF of 35% or less and NYHA Class II or III symptoms on chronic GDMT, who have reasonable expectation of meaningful survival for more than 1 year.
- ICD therapy is recommended for primary prevention of SCD to reduce total mortality in selected patients at least 40 days post-MI with LVEF of 30% or less, NYHA Class I symptoms while receiving GDMT, who have a reasonable expectation of meaningful survival for more than 1 year.

Recommendations

- Implantation of an ICD for primary prevention is not recommended within the first 3 months after initial diagnosis of NICM.
- If recovery of left ventricular function is unlikely, implantation of an ICD for primary prevention can be useful between 3 and 9 months after initial diagnosis of NICM.

TABLE 4. Characteristics of Patients in Whom CRT Is Strongly Supported by Randomized Trials

Sinus rhythm

LVEF ≤0.35

Ischemic or nonischemic cardiomyopathy

QRS complex duration ≥120 ms

NYHA functional class III or IV

Maximal pharmacological therapy for heart failure

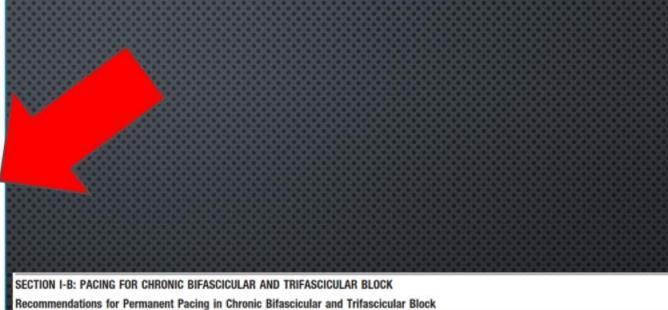
Kusumoto et al HRS/ACC/AHA Expert Consensus Statement on ICD Therapy 2013 Circulation. 2005;111:2146-2150



Indications for Permanent Pacing in Acquired Atrioventricular Block in Adults

Class I

- Third-degree AV block at any anatomic level associated with any one of the following conditions:
 - a. Bradycardia with symptoms presumed to be due to AV block. (Level of evidence: C)
 - Arrhythmias and other medical conditions that require drugs that result in symptomatic bradycardia. (Level of evidence: C)
 - c. Documented periods of asystole ≥3.0 seconds or any escape rate <40 beats per minute (bpm) in awake, symptom-free patients. (Level of evidence: B, C)
 - d. After catheter ablation of the AV junction. (Level of evidence: B, C) There are no trials to assess outcome without pacing, and pacing is virtually always planned in this situation unless the operative procedure is AV junction modification.
 - e. Postoperative AV block that is not expected to resolve. (Level of evidence: C)
 - f. Neuromuscular diseases with AV block such as myotonic muscular dystrophy, Kearns-Sayre syndrome, Erb's dystrophy (limb-girdle), and peroneal muscular atrophy. (Level of evidence: B)
- Second-degree AV block regardless of type or site of block, with associated symptomatic bradycardia. (Level of evidence: B)



Class I	Class I	Class I

 Intermittent third-degree AV block. (Level of Evidence: B) (27–33)

 Type II second-degree AV block. (Level of Evidence: B) (34–36)

3. Alternating bundle-branch block. (Level of Evidence: C) (37)

No change

No change

New Class I recommendation that adds alternating bundle branch block to the manifestations of fascicular block that indicate pacing therapy. This recommendation was not explicitly stated in the previous version.



SUMMARY HIGHLIGHTS

- ICD SEEMS TO BE USED EARLIER THAN PRIOR GUIDELINES.
- AFIB HR <110 (IF HEART NOT ENLARGING) OR 85 IF PATIENT TOLERATES DRUGS WELL
- WATCH FOR CHANNELPATHIES



Post test



QUESTION 1: PATIENT HAS CARDIAC ARREST.. NON ISCHEMIC HEART DISEASE NO MI

- Patient needs further electrical monitoring with beta blocker treatment.
- 2. Patient should have ICD implanted ... a class 1 indication
- Patient instructed to have further follow up with primary physician with global risk reduction
- Patient requires echocardiogram to further assess his need for ICD implant
- 5. Patient needs no further treatment except for his ischemic heart disease



6.2.1. Secondary Prevention of SCD in Patients With NICM

Recommendations for Secondary Prevention of SCD in Patients With NICM

References that support the recommendations are summarized in Online Data Supplement 25 and 26.

COR	LOE	Recommendations
ı	B-R	1. In patients with NICM who either survive SCA due to VT/VF or experience hemodynamically unstable VT (LOE: B-R) (1-4) or stable VT (LOE: B-NR) (5)
	B-NR	not due to reversible causes, an ICD is recommended if meaningful survival greater than 1 year is expected.
lla	B-NR	 In patients with NICM who experience syncope presumed to be due to VA and who do not meet indications for a primary prevention ICD, an ICD or an electrophysiological study for risk stratification for SCD can be beneficial if meaningful survival greater than 1 year is expected (6-11).
IIb	B-R	 In patients with NICM who survive a cardiac arrest, have sustained VT, or have symptomatic VA who are ineligible for an ICD (due to a limited life- expectancy and/or functional status or lack of access to an ICD), amiodarone may be considered for prevention of SCD (12, 13).



QUESTION 2

Mr Gomez is asymptomatic with neuromuscular disorder and ask if he needs a ICD, your recommendation would be

- 1. He is not a candidate for ICD because he is asymptomatic
- Mr. Gomez needs heart catheterization to evaluate his CAD status first before considering ICD
- 3. Patients with neuromuscular disorder, primary and secondary prevention, ICDs are recommended
- 4. Patient needs EP study and depending on results may need ICD
- Patient would need to have documented complete heart block before further work up



QUESTION 3

Miss Johnson is a asymptomatic 34 y/o women who presents for breast biopsy and EKG finds long QT interval >470 ms which best describes best treatment option

- 1. Patient needs to be started on beta blocker and followed for symptoms
- 2. Patient needs no further treatment since asymptomatic
- 3. Patient is a candidate for biventricular pacemaker set at heart rate faster than her intrinsic rate
- 4. Patient is a candidate for ICD
- 5. Patient needs ETT first to make further decisions



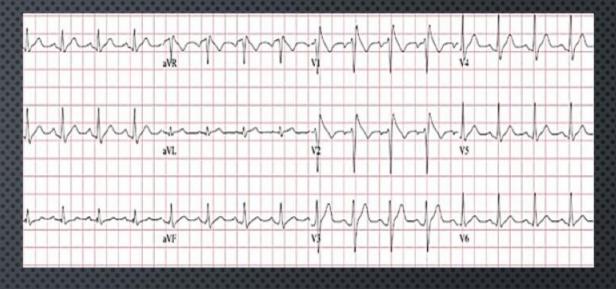
Mr Phillips is having ETT and develops this rhythm



- 1. Patient has monomorphic VT and needs CCU admission
- Patient needs ICD immediately
- Patient has exercise-induced polymorphic VT in catecholaminergic polymorphic ventricular tachycardia and needs beta blocker (class 1 indication)
- 4. Patient will not benefit from beta blocker and is contraindicated
- Patient needs iv amiodarone



43 y/o male brought by ambulance to ER after apparent cardiac arrest, no chest pain, negative troponin and the following EKG, your diagnosis



- 1. Takotsubo heart disease
- 2. Brugada syndrome and has class 1 indication for ICD
- 3. Right ventricular infarction
- 4. Non STEMI
- 5. Left bundle branch block



49 y/o male presents with <u>atrial fibrillation</u> with hypertrophic cardiomyopathy with low CHA2DS2-VASc score

- 1. Patient does not require any anticoagulation since score is low
- 2. Patient requires ticagrelor
- 3. Anticoagulation is indicated independent of the CHA2DS2-VASc score
- 4. Anticoagulation is only needed if the EF < 35%



	Score		Adjusted Stroke Rate (% per y)
CHADS ₂		CHADS₂*	
Congestive HF	1	0	1.9
Hypertension	1	1	2.8
Age ≥75 y	1	2	4.0
Diabetes mellitus	1	3	5.9
Stroke/TIA/TE	2	4	8.5
Maximum score	6	5	12.5
		6	18.2
CHA ₂ DS ₂ -VASc	(CHA ₂ DS ₂ -VASc	
Congestive HF	1	0	0
Hypertension	1	1	1.3
Age ≥75 y	2	2	2.2
Diabetes mellitus	1	3	3.2
Stroke/TIA/TE	2	4	4.0
Vascular disease (prior MI, PAD, or aortic plaque)	1	5	6.7
Age 65-74 y	1	6	9.8
Sex category (i.e., female sex)	1	7	9.6
Maximum score	9	8	6.7
		9	15.20

Thank you

