Sexual Activity in CHD & Channelopathies: The Pink Elephant No One Wants to Discuss!

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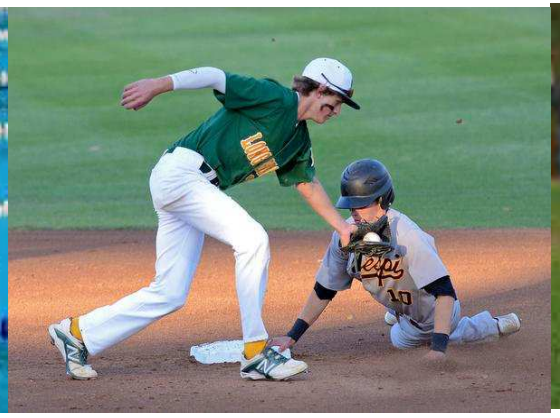
Cardiology 2018

16 year-old boy with HCM (septum 23 mm)
Can my son play the following sports?







17 year-old female with LQT type II (QTc 490 msec)
Can my daughter play the following sports?



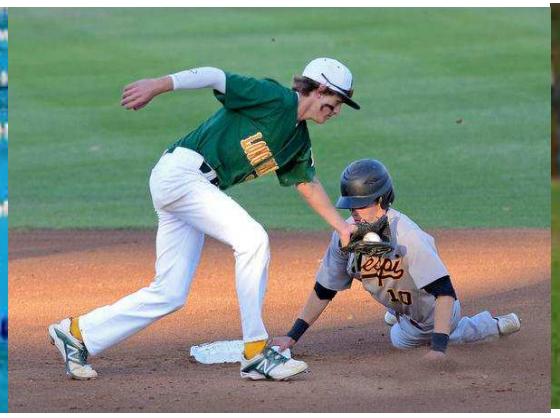





17 year-old male s/p Ross with mild AS/moderate AI
Can my son participate in the following sports?








yes
no
YES

What about Sex?

Shhh...

we are at a children's hospital

Case #1

- 21 year old-young lady with HCM
- Family History: Sudden cardiac death in her father at age 38
- At age 12 she underwent a dual chamber ICD for family history
- No shocks, battery reached end-of-life at age 19 year (**new ICD?)**
- Continued to be without symptoms
- Elected to replace the ICD generator
- 1 month prior to her wedding, practicing with her fiancée for their honeymoon, late dinner, glass of wine, at the end of sexual intercourse……


KABOOM

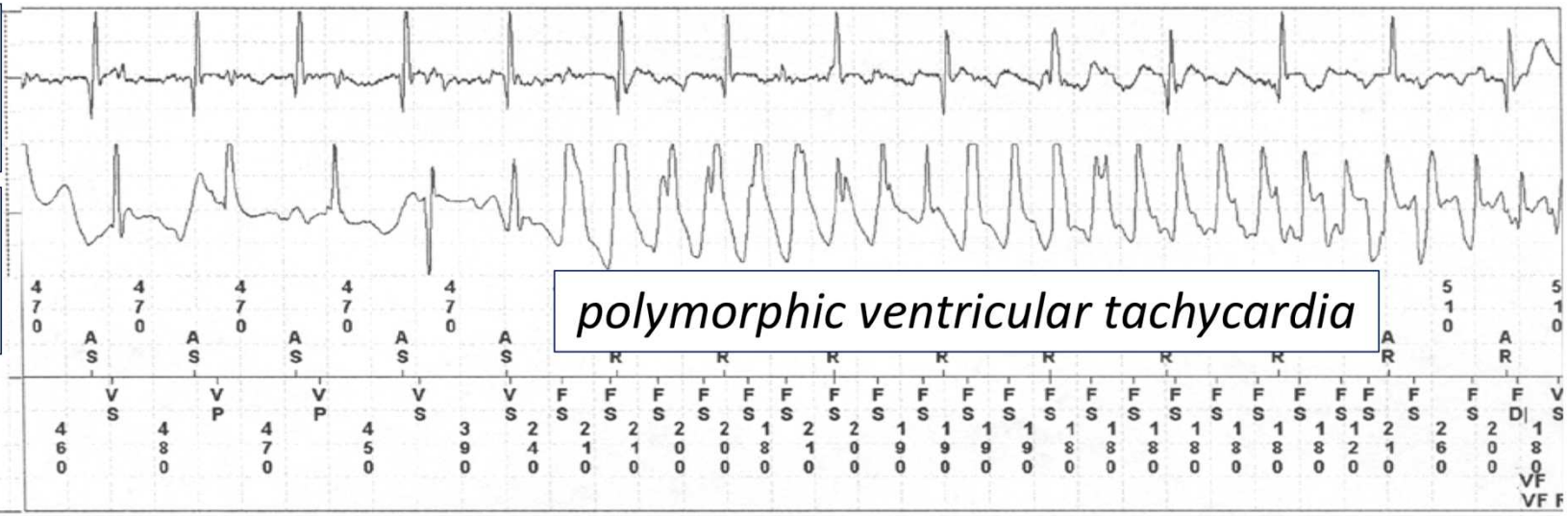
Case #1

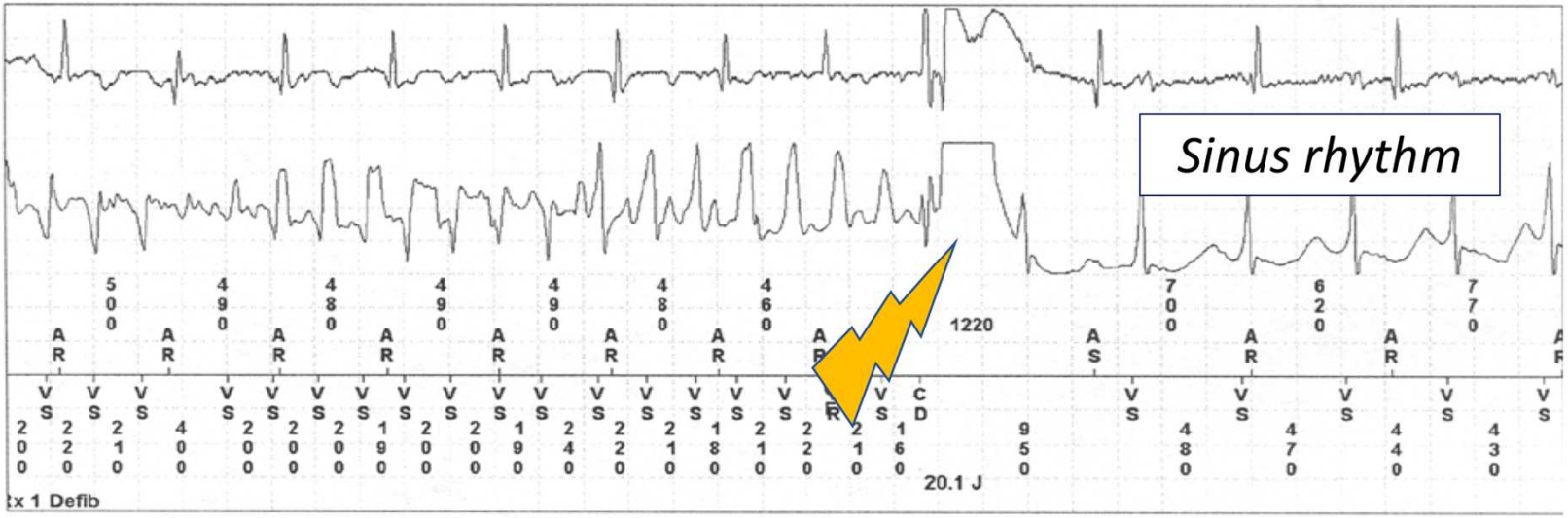
- She was pretty sure she “just got shocked”
- The fiancée thought he was “exceptional” and she should just accept that fact and that their honeymoon would be AWESOME!!!
- I received a phone call from my patient who lived in Tucson and I had them come up to the office.

Atrial Channel

Ventricular Channel

Marker Annotation





Case #1 (cont.,)

- The patient was fine, we talked about alcohol, dehydration, lack of sleep (she had been up the night before planning last minute wedding activities, seating arrangements etc)
- Increased verapamil SR from 180 mg to 240 mg PO QD
- 1 month after the wedding – received another phone call from my patient that her now husband was having performance anxiety *("and no Dr. Cohen it was not an AWESOME honeymoon between the sheets")* and was hoping I could recommend someone for therapy for him.
- 12 months after therapy, they appear to be doing well.

Case #2

- 14 year old boy found by grandmother sitting upright in a chair but blue and unconscious with pants down (*Playboy Magazine* next to the chair)
- **And no, I am sure he was not reading the articles!**
- Resuscitated by grandmother (a nurse)
- VF upon arrival by EMS (Epi, shock, and return of spontaneous rhythm)
- At an outside hospital an ECG revealed irregular pattern of bigeminy per report but no strips available, bradycardia also noted and one arrest might have been in the setting of bradycardia??
- Epi drip at outside hospital for a few hours and no mention of worsened arrhythmias

Courtesy, Susan Etheridge MD

Past Medical History:2 previous ? syncopal episodes

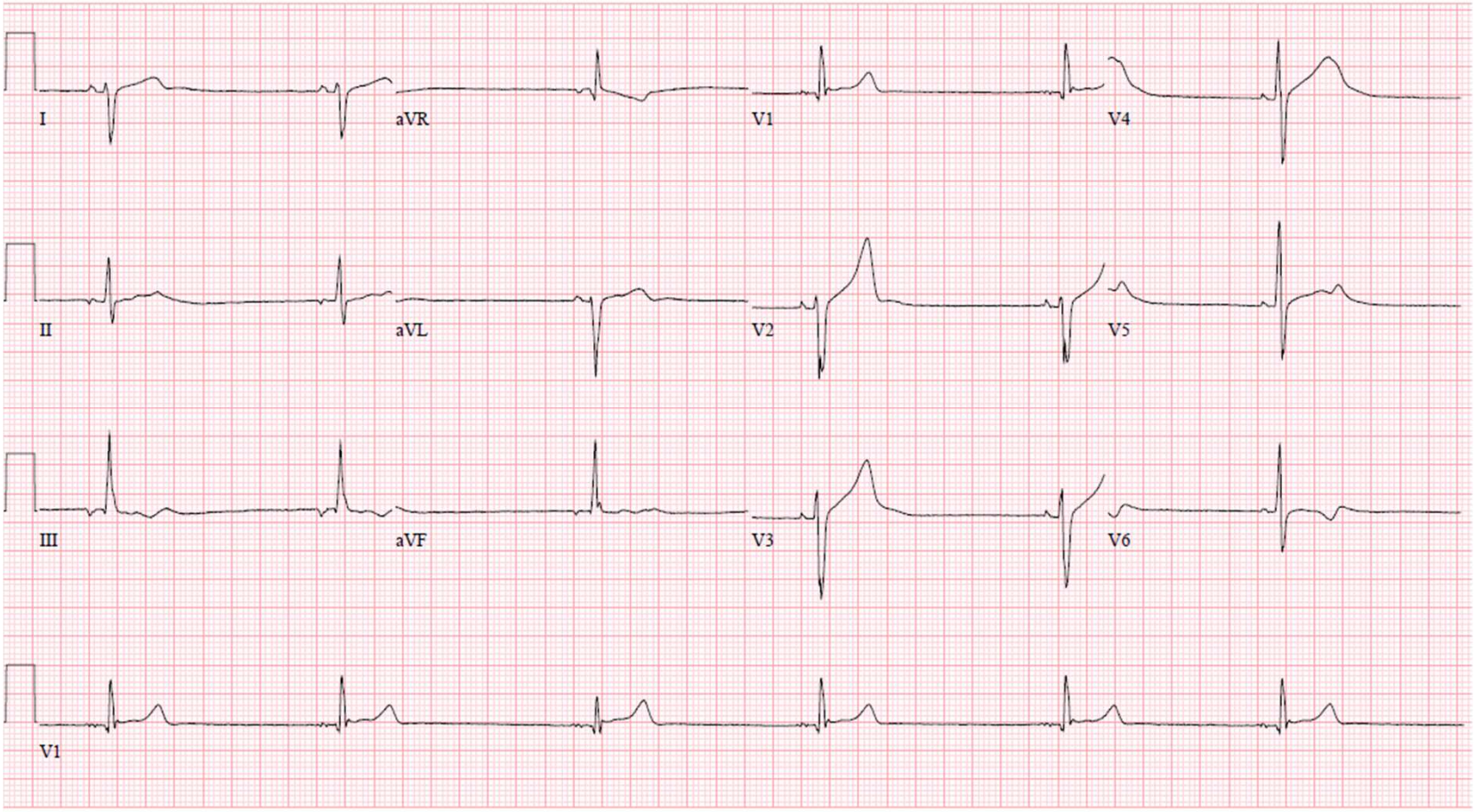
- **age 10, fainted while upset**
  - spontaneous return of consciousness
  - workup including a 24 hour Holter (-)

FH: 9-year-old second cousin suffered a near drowning episode and has an ICD.

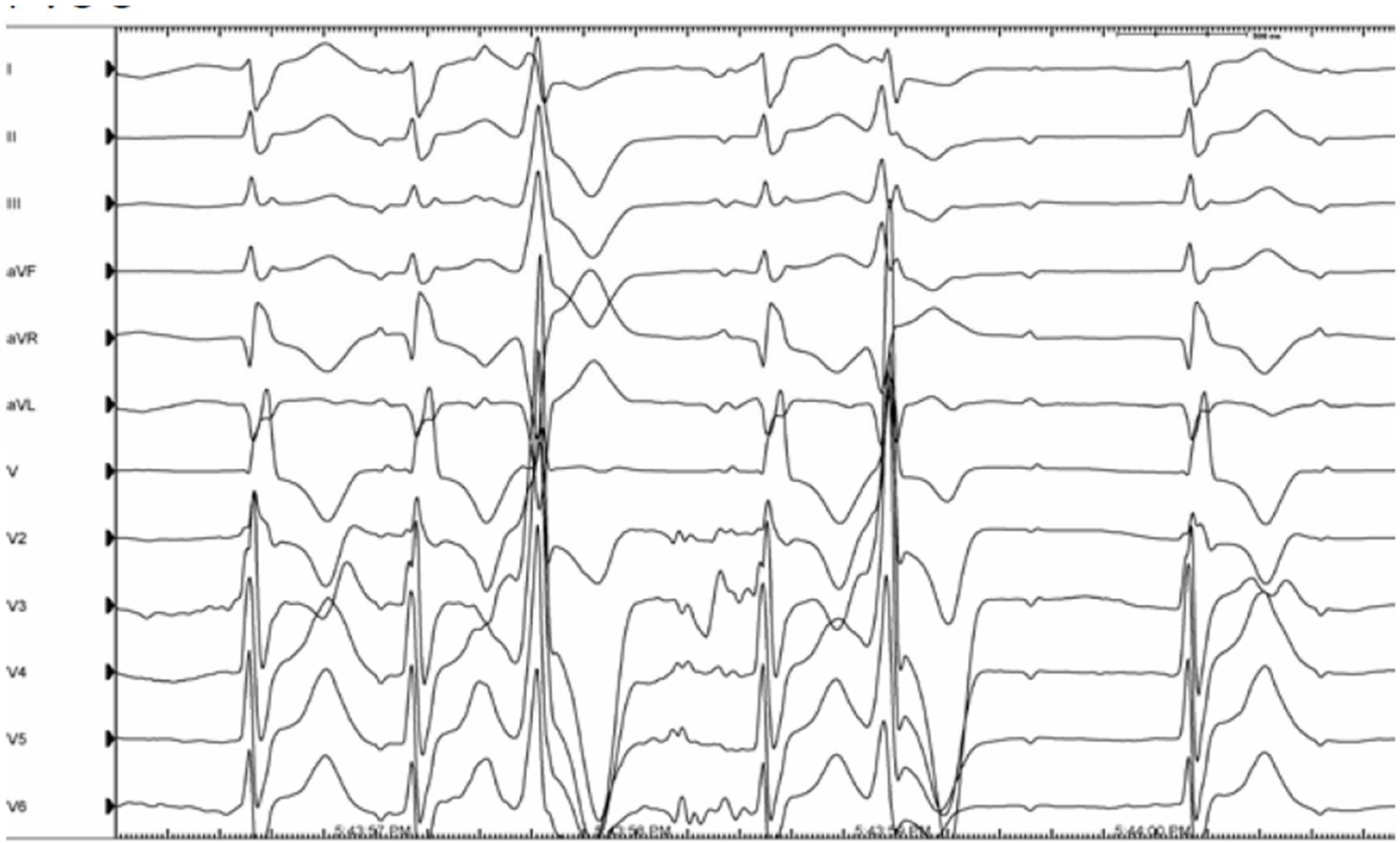
4 months prior to the VF arrest

- "all riled up" arguing with father and began breathing erratically
- unclear if complete LOC but shaking "as if he was having a seizure"
- After a few minutes regained full consciousness
- EMS was called but nothing done
- states he felt like things closed in and then he was watching the event from his father’s perspective

ECG on admission



Normal Echo, cath normal with normal coronary arteries



Uniform PVCs during epi challenge
Normal procainamide challenge

Normal MRI
Normal Biopsy

View Image -- Webpage Dialog

Notes View Image

Close

1 of 1

- Invitae Catecholaminergic Polymorphic Ventricular Tachycardia Panel

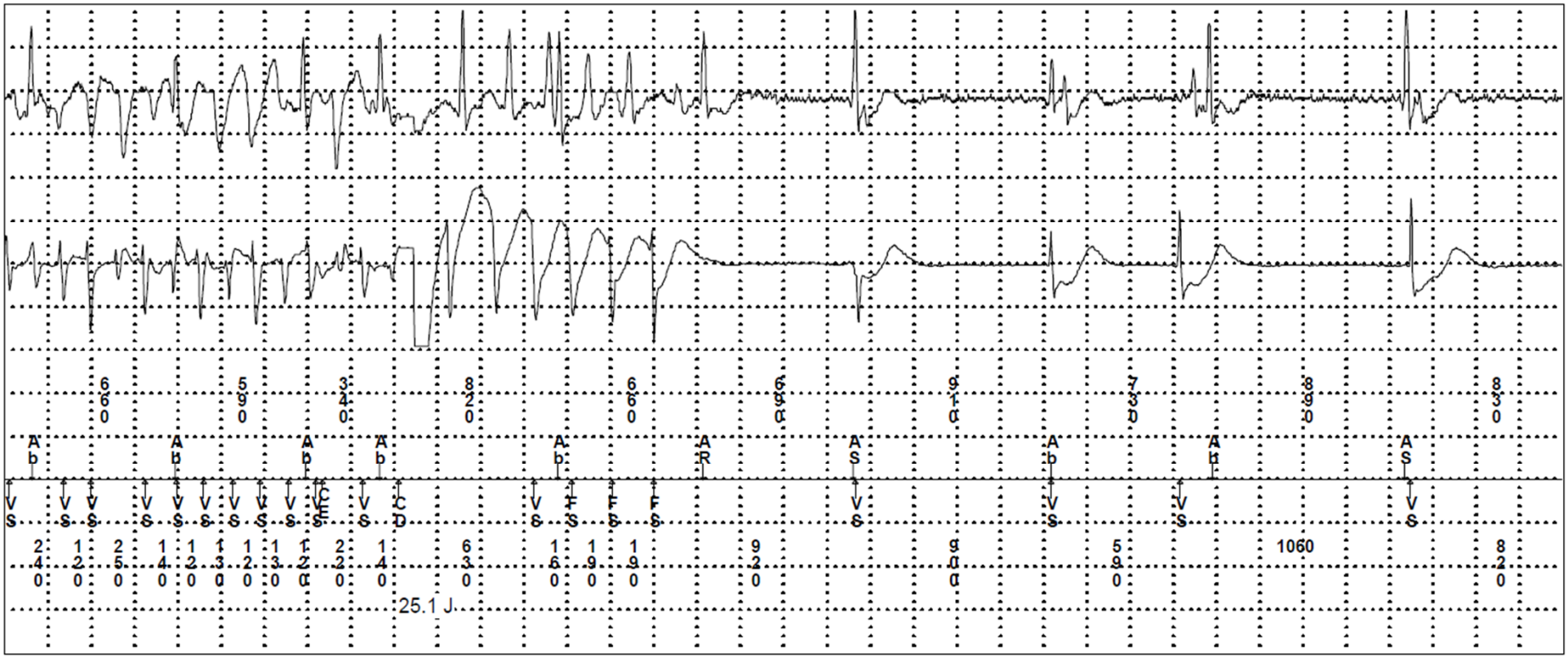
Summary

Variant of Uncertain Significance identified in RYR2. But probably CPVT

Clinical Summary

- A Variant of Uncertain Significance, c.11150A>G (p.Lys3717Arg), was identified in RYR2.
  - The RYR2 gene is associated with autosomal dominant catecholaminergic polymorphic ventricular tachycardia (CPVT) (MedGen UID: 351513), arrhythmogenic right ventricular cardiomyopathy (ARVC) (MedGen UID: 318748) and left ventricular noncompaction (LVNC) (PMID: 24394973).
  - The clinical significance of this variant is uncertain at this time. Until this uncertainty can be resolved, caution should be exercised before using this result to inform clinical management decisions.
  - This variant may qualify for complimentary family studies as part of our VUS Resolution Program. De novo variants are a known cause of RYR2-related conditions. Parental testing may clarify the inheritance of this variant. Please visit www.invitae.com for more information.
- These results should be interpreted within the context of additional laboratory results, family history, and clinical findings. Genetic counseling is recommended to discuss the implications of this result. For access to a network of genetic providers, please contact Invitae at clientservices@invitae.com, or visit www.nsgc.org or tagc.med.sc.edu/professional.organizations.asp.

Complete Results



Noncompliant on Meds Continued with VF Events

Case #3

- 14 year-old boy presents for 5th opinion after having repeated seizures with exercise
- Stress Test: PVCs and non-sustained runs of VT (3 beats)
- Genetic testing (research): VUS in RYR2
- Started beta-blockers
- Diagnosis: probable CPVT
- Restricted from exercise
- No symptoms
- Followed by a local cardiologist in his home town

Courtesy, Maully Shah MBBS

Case #3 (cont.,)

- Age 19, started becoming non-compliant with his BB
- Continued to be non-compliant with his medications
- Repeatedly missed follow-up appointments
- Age 20 fathered one child
- Despite missing appointments PMD refilled Atenolol
- One morning during sexual intercourse, arrested
- Girlfriend started CPR, EMS arrived, shocked a few times for VF, died.
- Note from local cardiologist told him to go "slow" during sex

Courtesy, Maully Shah MBBS

What Do We Know About Sudden Cardiac Death?

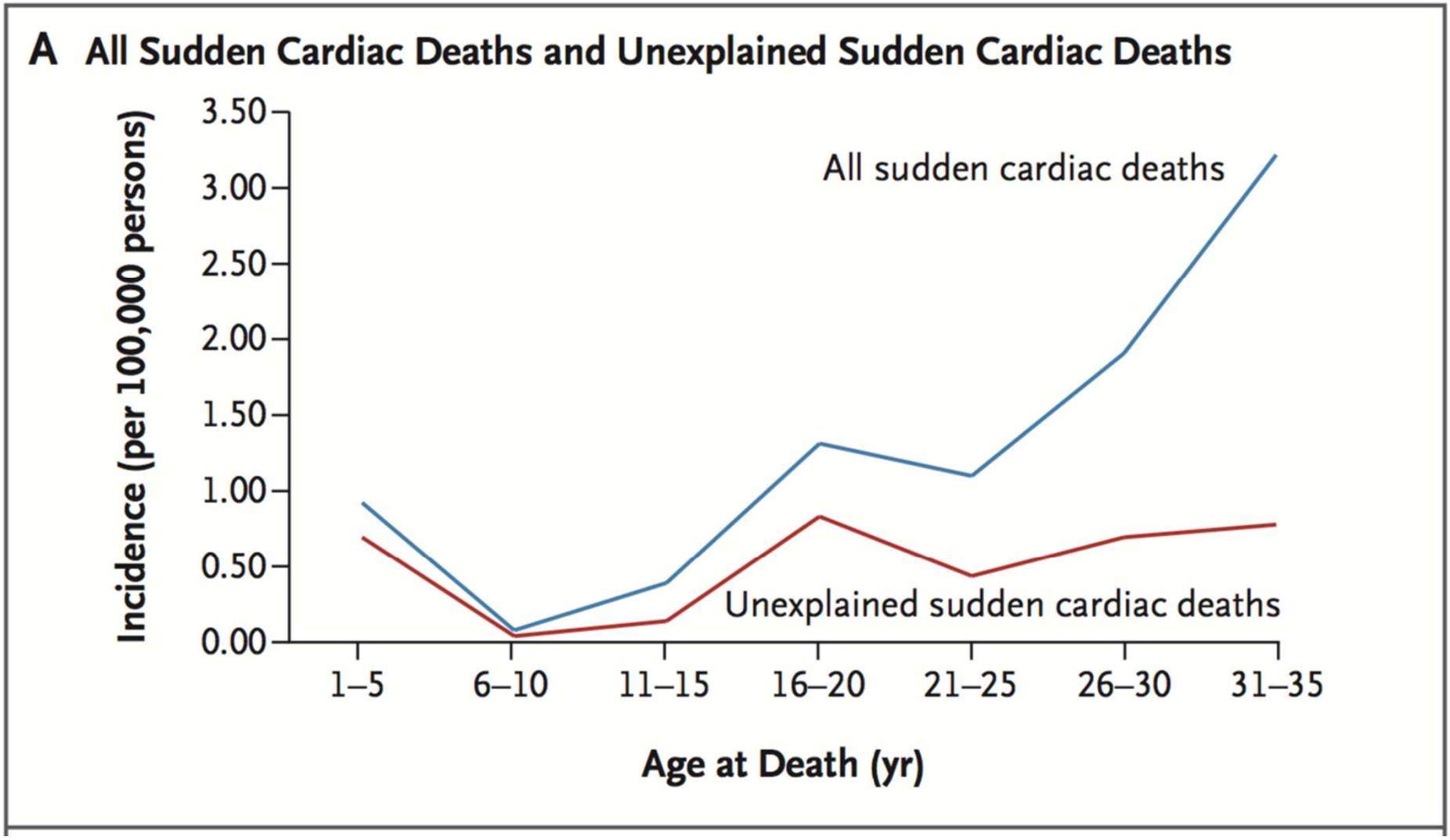
The NEW ENGLAND JOURNAL of MEDICINE

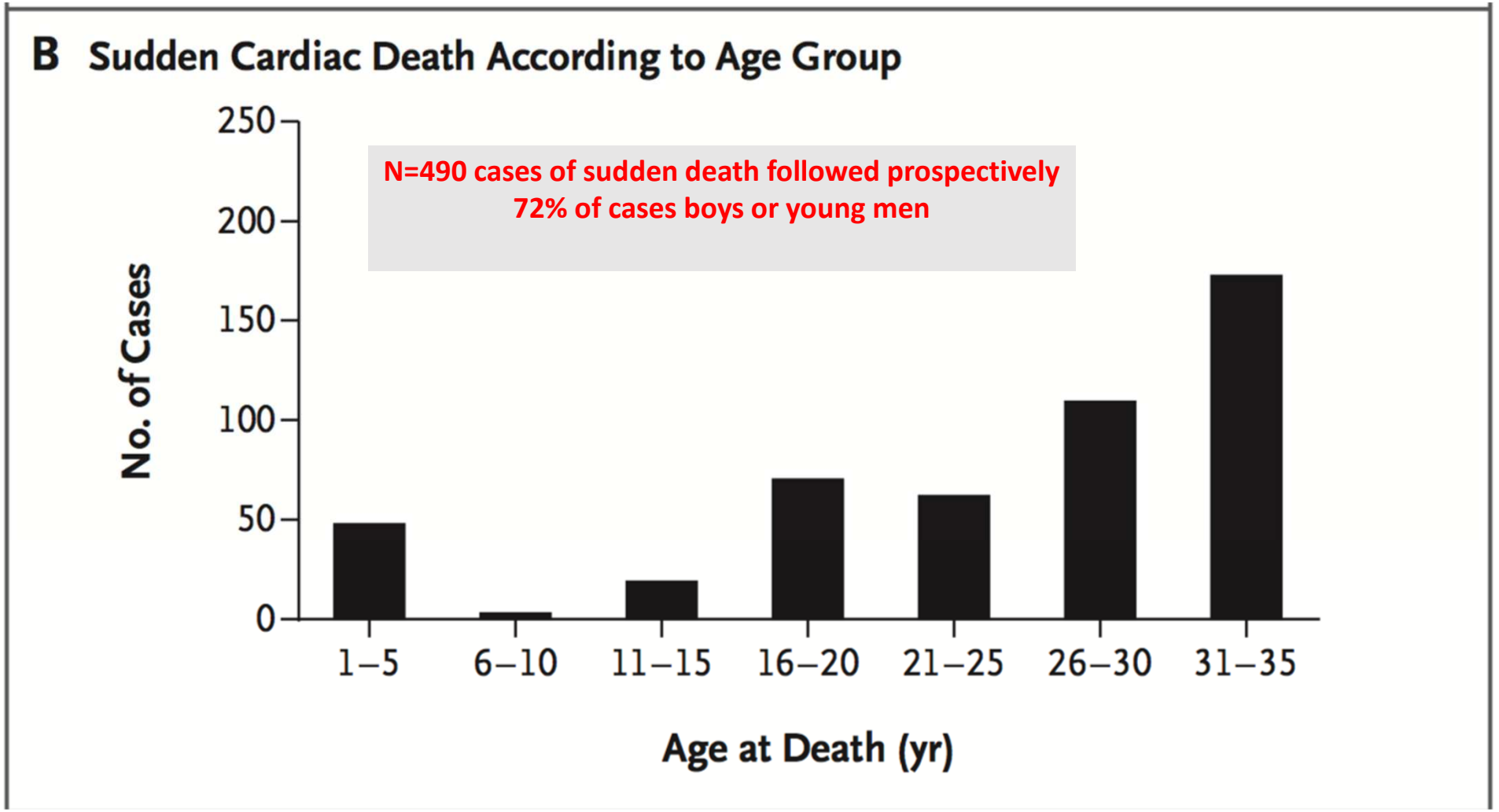
ORIGINAL ARTICLE

A Prospective Study of Sudden Cardiac Death among Children and Young Adults

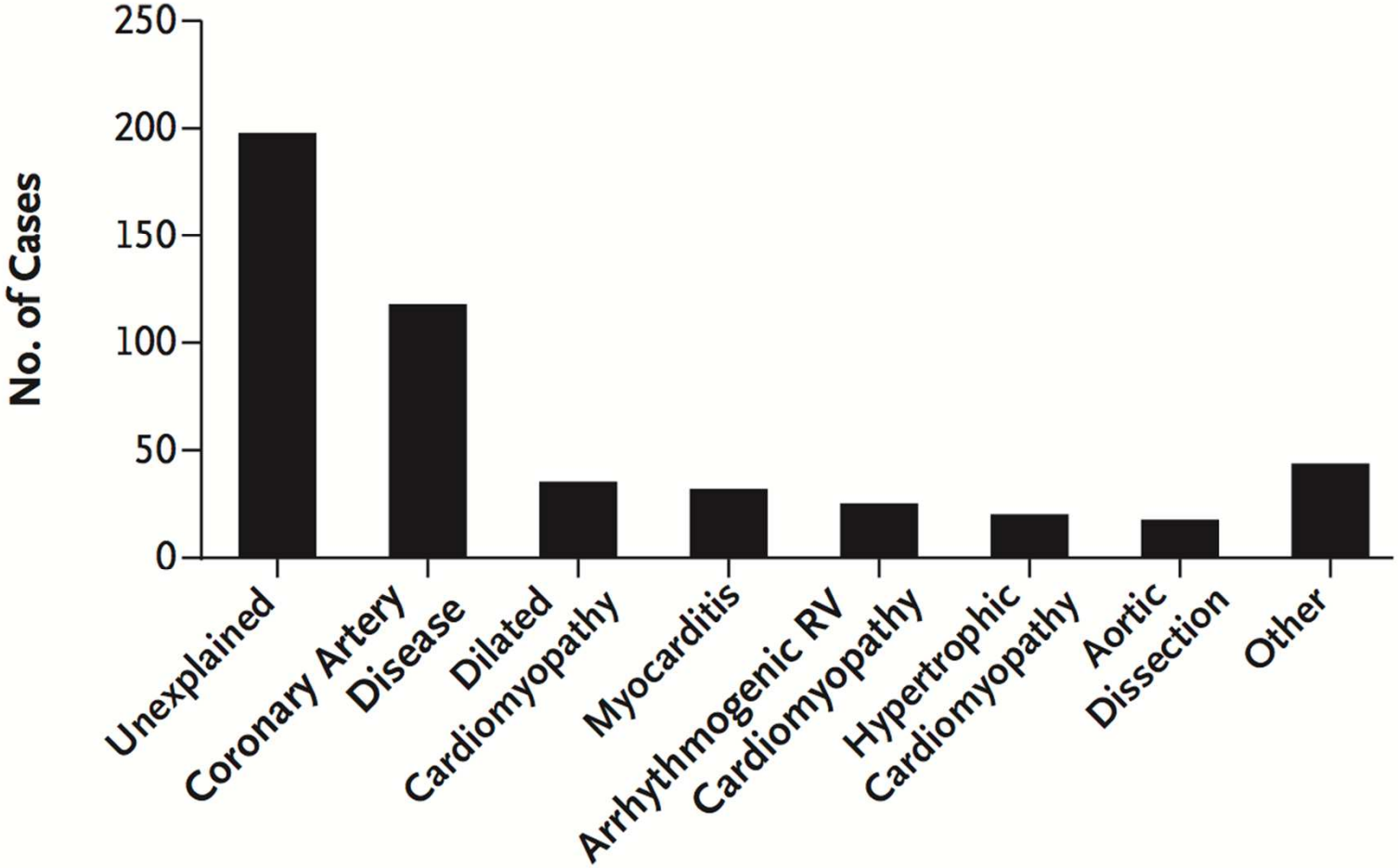
R.D. Bagnall, R.G. Weintraub, J. Ingles, J. Duflou, L. Yeates, L. Lam, A.M. Davis, T. Thompson, V. Connell, J. Wallace, C. Naylor, J. Crawford, D.R. Love, L. Hallam, J. White, C. Lawrence, M. Lynch, N. Morgan, P. James, D. du Sart, R. Puranik, N. Langlois, J. Vohra, I. Winship, J. Atherton, J. McGaughran, J.R. Skinner, and C. Semsarian

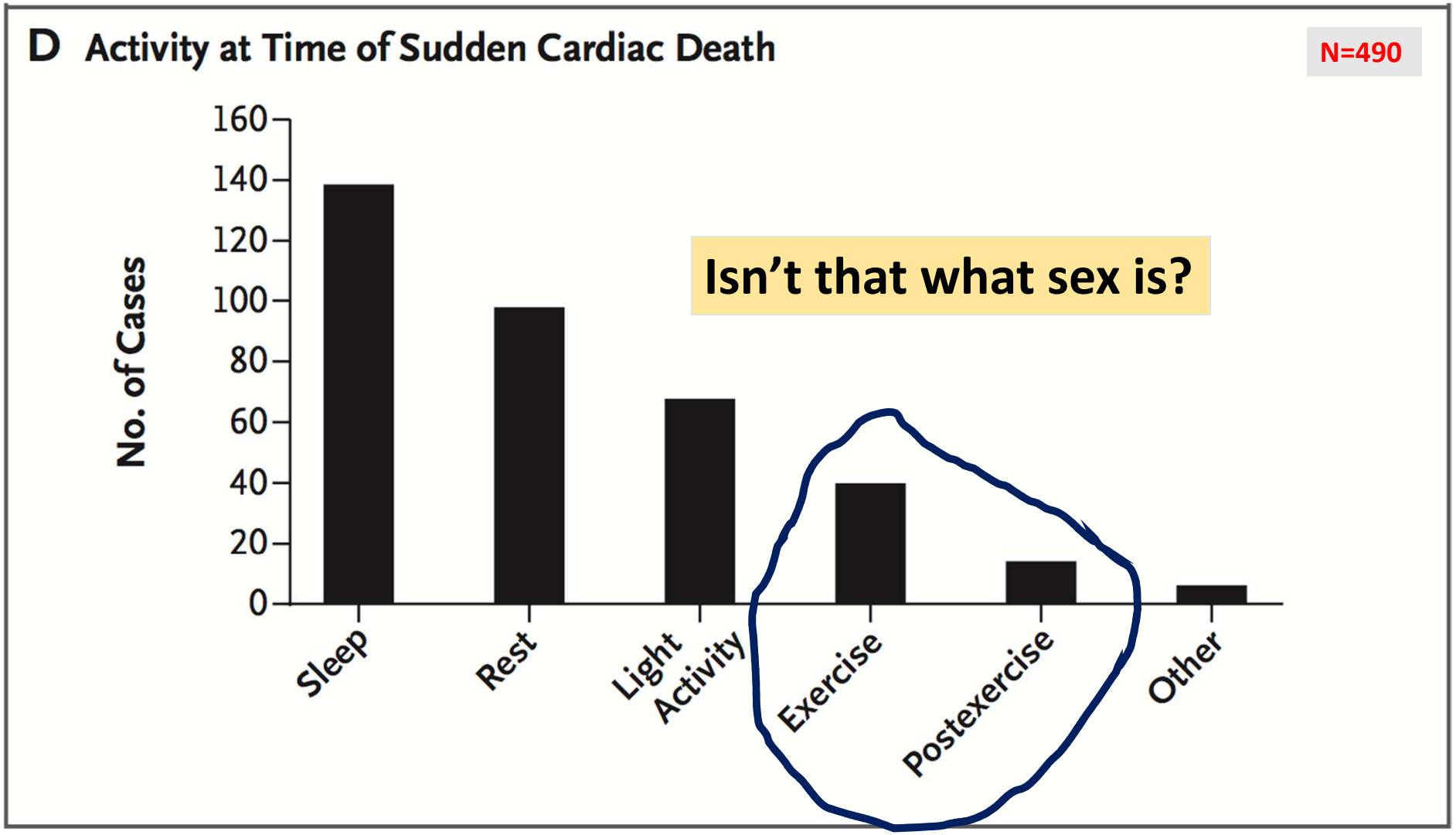
2016


A All Sudden Cardiac Deaths and Unexplained Sudden Cardiac Deaths
Incidence (per 100,000 persons)
3.50
3.00
2.50
2.00
1.50
1.00
0.50
0.00
All sudden cardiac deaths
Unexplained sudden cardiac deaths
1–5
6–10
11–15
16–20
21–25
26–30
31–35
Age at Death (yr)


B Sudden Cardiac Death According to Age Group
N=490 cases of sudden death followed prospectively
72% of cases boys or young men
No. of Cases
250
200
150
100
50
0
1–5
6–10
11–15
16–20
21–25
26–30
31–35
Age at Death (yr)

C Causes of Sudden Cardiac Death




D Activity at Time of Sudden Cardiac Death
N=490
No. of Cases
160
140
120
100
80
60
40
20
0
Sleep
Rest
Light Activity
Exercise
Postexercise
Other
Isn't that what sex is?

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S-e-x!!! #1

10-19-2010, 12:55 AM

Ok, I'm very conservative by nature, but not one who is afraid to talk about topics that can make people uncomfortable. I personally think that sex is a beautiful blessing.

I know this topic will get peoples attention... I also know it's kind of taboo. But what the heck, I like to have fun with the discussions and think this is a topic that should be covered. I ha

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I'm sure I will think of more questions/comments, but this should get us started. Let me know your thoughts/etc.

-Darren

- Is sex safe with HCM (no, I'm not talking about condoms)
- Why do we with HCM tend to sweat when we sleep and "ahem" when we are dancing in the sheets?
- If we are discouraged from doing activities that get our heart rate to change too rapidly, then how can we be safe?
- If we have sex and don't climax is that safer?
- If we are having sex regularly and are more relaxed, is that better?

Is that true for everyone?

AHA Scientific Statement

Sexual Activity and Cardiovascular Disease

A Scientific Statement From the American Heart Association

Endorsed by the American Urological Association, Society for Cardiovascular Angiography and Interventions, Society of Thoracic Surgeons, American Association of Cardiovascular and Pulmonary Rehabilitation, International Society of Sexual Medicine, American College of Cardiology Foundation, Heart Rhythm Society, and Heart Failure Society of America

Glenn N. Levine, MD, FAHA, Chair; Elaine E. Steinke, RN, PhD, FAHA; Faisal G. Bakaeen, MD; Biykem Bozkurt, MD, PhD, FAHA; Melvin D. Cheitlin, MD, FAHA; Jamie Beth Conti, MD; Elyse Foster, MD, FAHA; Tiny Jaarsma, RN, PhD, FAHA; Robert A. Kloner, MD, PhD; Richard A. Lange, MD, MBA, FAHA; Stacy Tessler Lindau, MD; Barry J. Maron, MD; Debra K. Moser, DNSc, RN, FAHA; E. Magnus Ohman, MD; Allen D. Seftel, MD; William J. Stewart, MD; on behalf of the American Heart Association Council on Clinical Cardiology, Council on Cardiovascular Nursing, Council on Cardiovascular Surgery and Anesthesia, and Council on Quality of Care and Outcomes Research

Acute Cardiovascular Effects of Sexual Activity

- During heterosexual vaginal intercourse, systolic and diastolic blood pressure increase mildly
- The greatest increases occur around the time of orgasm. Heart rate rarely exceeds 130 bpm (again this is an adult study) and systolic BP rarely exceeds 170 mmHg in normotensive individuals
- Men and women have similar responses
- Sexual activity in young healthy married men with their usual partner is comparable to moderate physical activity in the range of 3-5 METS (equivalent of easily climbing two flights of stairs).

Heart Failure Recommendations

- Sexual function correlates with symptomatic status (NYHA and 6 minute walk test) but not with an ejection fraction
- Most heart failure patients actually placed greater emphasis on the importance of quality of life, including sexual activity, than on improving survival

- **Sexual activity is reasonable for patients with compensated and/or mild (NYHA class I or II) heart failure *(Class IIa; Level of Evidence B)*.**
- **Sexual activity is not advised for patients with decompensated or advanced (NYHA class III or IV) heart failure until their condition is stabilized and optimally managed *(Class III; Level of Evidence C)*.**

Circulation 2012,125:1058-1072

Sexual Activity & ICD Recommendations

- In large autopsy series (again mostly adults), the incidence of sudden death during sexual intercourse in patients with ICDs was 0.6-1.7%
- In post-MI patients, the frequency of ventricular ectopy and ICD shocks was comparable during physical exertion, mental stress and sexual activity.

- **Sexual activity is reasonable for patients with an ICD implanted for primary prevention *(Class IIa; Level of Evidence C)*.**
- **Sexual activity is reasonable for patients with an ICD used for secondary prevention in whom moderate physical activity (>3–5 METS) does not precipitate ventricular tachycardia or fibrillation and who do not receive frequent multiple appropriate shocks.**

Circulation 2012,125:1058-1072
Can Med Assoc 1977,116:1250-1253
Clin Cardiol 2002;25:474-478

Sexual Activity & CHD Recommendations (pseudo-meta analysis)

- 9% ♀ with CHD reported symptoms during sexual activity, which included dyspnea, perceived arrhythmia, increased fatigue, or syncope
- Increases with complexity of CHD
- In a survey of ♂ with CHD, 9% reported subjective arrhythmias (**probably not the same 9%),** and 5% reported chest pain with sexual activity and it increased with worsening NYHA.
- **Be Careful** in patients with pulmonary HTN, cyanotic heart disease, LVTOTO, anomalous coronary artery between the aorta and pulmonary artery (R?L?)

Sexual activity is reasonable for most CHD patients who do not have decompensated or advanced heart failure, severe and/or significantly symptomatic valvular disease, or uncontrolled arrhythmias (*Class IIa*)

Circulation 2012,125:1058
Heart 2009,95:1179
Circulation 2009;119:1085

Sexual Activity & HCM Recommendations

- Most common cause arrhythmia related sudden cardiac death in the young
- Approximately 70% have LVOTO at rest or with physiological provocation
- Linkage between physical activity and sudden death events attributable to VT/VF raises concern that **vigorous sexual activity** may heighten the risk in patient with disease.
- ***??? What defines and who defines vigorous sexual activity????***

1. Sexual activity is reasonable for most patients with hypertrophic cardiomyopathy (HCM) *(Class IIa; Level of Evidence C)*.
2. Sexual activity should be deferred for patients with HCM who are severely symptomatic until their condition is stabilized *(Class III)*

Circulation 2012,125:1058-1072
Circulation 2006;114:2232-2282

Sudden Cardiac Arrest During Sex in Patients with Either Catecholaminergic Polymorphic Ventricular Tachycardia or Long-QT Syndrome: A Rare But Shocking Experience

ROBERT W. LOAR, M.D.,* J. MARTIJN BOS, M.D., PH.D.,† BRYAN C. CANNON, M.D.,* and MICHAEL J. ACKERMAN, M.D., PH.D.*,†,‡

From the *Division of Pediatric Cardiology, Department of Pediatric and Adolescent Medicine; †Department of Molecular Pharmacology & Experimental Therapeutics Windland Smith Rice Sudden Death Genomics Laboratory; and ‡Division of Cardiovascular Diseases, Department of Medicine, Mayo Clinic, Rochester, Minnesota, USA

Sudden Cardiac Arrest During Sex in Cardiac Channelopathies. ***Background:*** **Patients with catecholaminergic polymorphic ventricular tachycardia (CPVT) and long-QT syndrome (LQTS) are susceptible to cardiac events during sympathetic nervous system activation. Herein, we sought to determine the risk of cardiac events associated with sex in CPVT and LQTS patients.**

Methods and Results: **We reviewed the electronic medical record of patients seen in the Genetic Heart Rhythm Clinic. There were 445 patients ≥18 years diagnosed with LQTS (N = 402, age at diagnosis 30 ± 16 years) or CPVT (N = 43, age at diagnosis 25 ± 15 years). No sex-induced cardiac events occurred in the LQTS population, and 2 occurred in the CPVT population. Sex-induced events were more likely in CPVT (2/43, 4.7%) than LQTS (0/402, 0%, P = 0.008). One case involved a 22-year-old CPVT1 female with prior cardiac arrest, who experienced several appropriate implantable cardioverter defibrillator shocks during intercourse while taking β-blockers. The second case was a 52-year-old CPVT1 male with history of recurrent exercise-triggered syncope, who had syncope during sex in the setting of β-blocker noncompliance. Extrapolating from published estimates of intercourse frequency by age, the overall event rate was only 0.0004%, and 0.005% among the CPVT cohort.**

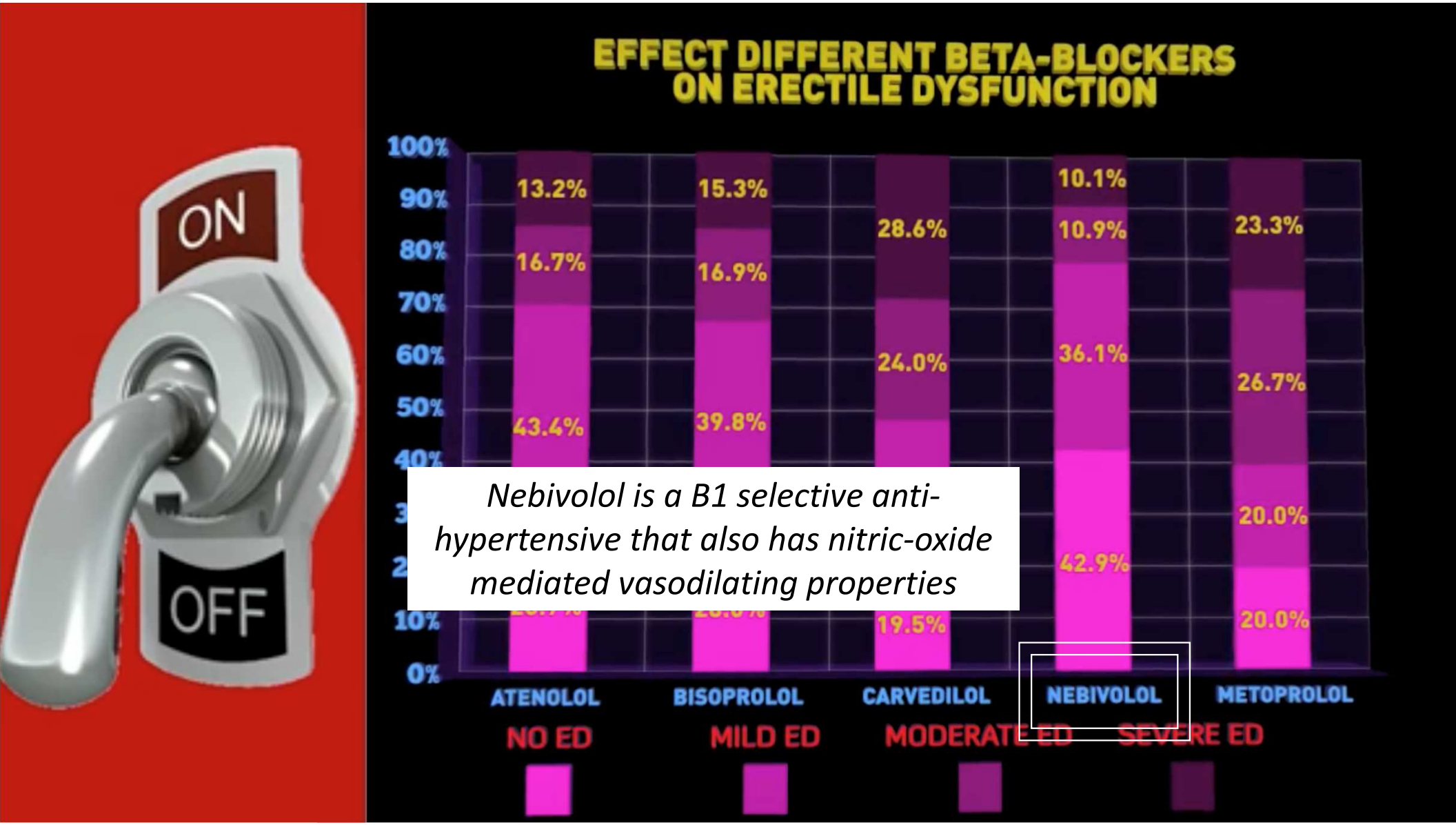
Conclusions: **Potentially life-threatening cardiac events during sex in patients with CPVT are rare and even rarer in LQTS. Overall, the cardiac event per intercourse rate is extremely low. Patients and their partners should be reassured that sex is a low-risk activity from a cardiac standpoint.** *(J Cardiovasc Electrophysiol, Vol. 26, pp. 300-304, March 2015)*

Sudden Cardiac Arrest During Sex in Patients with Either Catecholaminergic Polymorphic Ventricular Tachycardia or Long-QT Syndrome: A Rare But Shocking Experience

- Patients with LQT & CPVT are susceptible to cardiac events during sympathetic nervous system activation.
- Retrospective review of 445 patients ≥ 18 years with **LQTS** (N=402, age 30 ± 16 y) or **CPVT** (N=43, age 25 ± 15 years).
- Sex induced cardiac events (2/43 in CPVT 4.7%)
  - ICD storm, appropriate shocks during intercourse while taking beta-blockers
  - Recurrent exercise triggered syncope who had syncope during sex in the setting of beta-blocker non-compliance
- No sex induced cardiac events in the LQTS patients
- Overall cardiac event rate is low during sex in patients with CPVT and even rarer in LQT and patients should be reassured that sex is a low-risk activity??

True Statement: Patients with LQT, CPVT, and HOCM who are supposed to be on beta-blockers and stop taking them are at risk for life-threatening events.

Cardiovascular drugs that can improve symptoms and survival should not be withheld because of concerns about the potential impact on sexual function *(Class III: Harm; Level of Evidence C)*.


ON
OFF
EFFECT DIFFERENT BETA-BLOCKERS ON ERECTILE DYSFUNCTION
100%
90%
80%
70%
60%
50%
40%
10%
0%
13.2%
16.7%
43.4%
15.3%
16.9%
39.8%
28.6%
24.0%
19.5%
10.1%
10.9%
36.1%
42.9%
23.3%
26.7%
20.0%
20.0%
ATENOLOL
BISOPROLOL
CARVEDILOL
NEBIVOLOL
METOPROLOL
NO ED
MILD ED
MODERATE ED
SEVERE ED
Nebivolol is a B1 selective anti-hypertensive that also has nitric-oxide mediated vasodilating properties

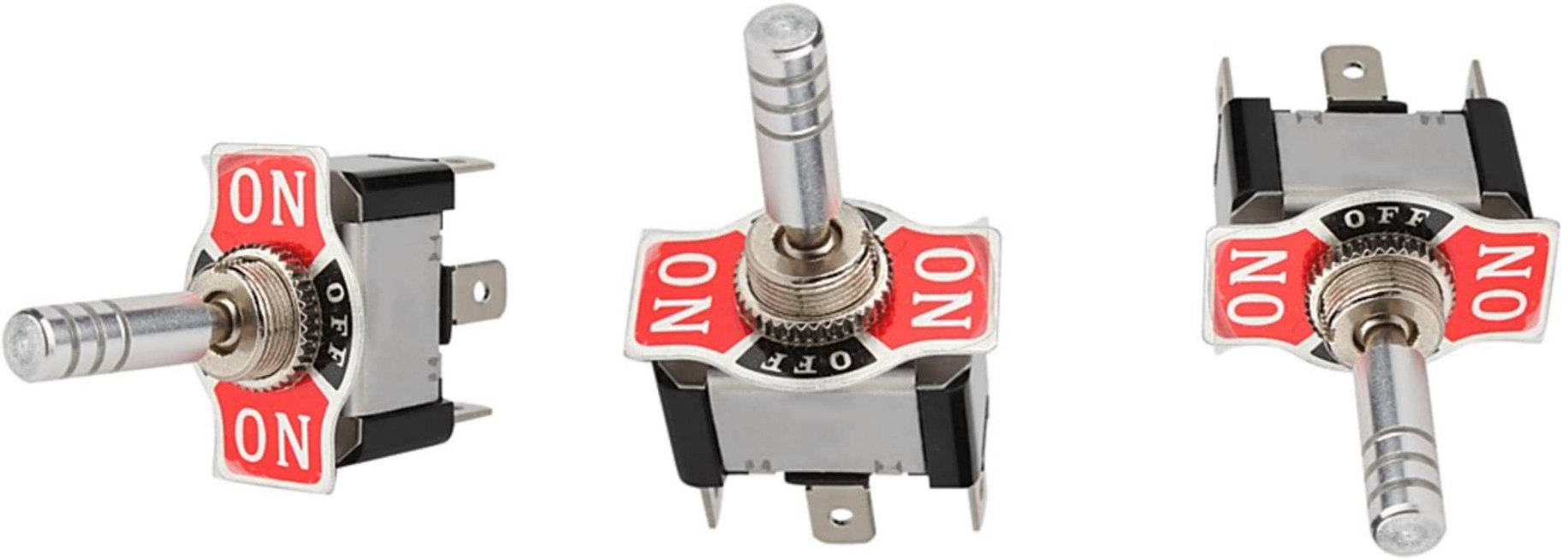
We give patients with heart disease beta-blockers which causes erectile dysfunction

So in order to have sexual intercourse and get to the point of having an orgasm one potentially has to work harder, last longer, and have more endurance?

So if you are non-compliant on your beta-blockers, you don’t have erectile dysfunction, you may perform better and not take as long?

Possible in the patient's eyes:

There is probably a healthy life switch balance




Low libido?

Pharmacotherapy for Sexual Dysfunction PDE5 Inhibitors

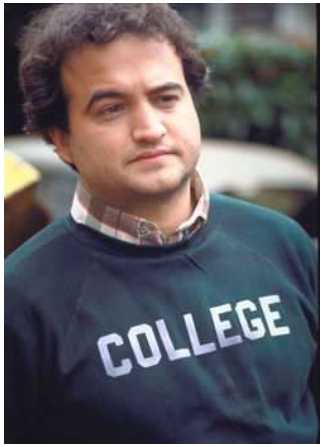
Recommendations

1. PDE5 inhibitors are useful for the treatment of ED in patients with stable CVD *(Class I; Level of Evidence A)*
2. The safety of PDE5 inhibitors is unknown in patients with severe aortic stenosis or HCM *(Class IIb; Level of Evidence C)*.

What Should We Do as Pediatric Cardiologists? (just my two cents)

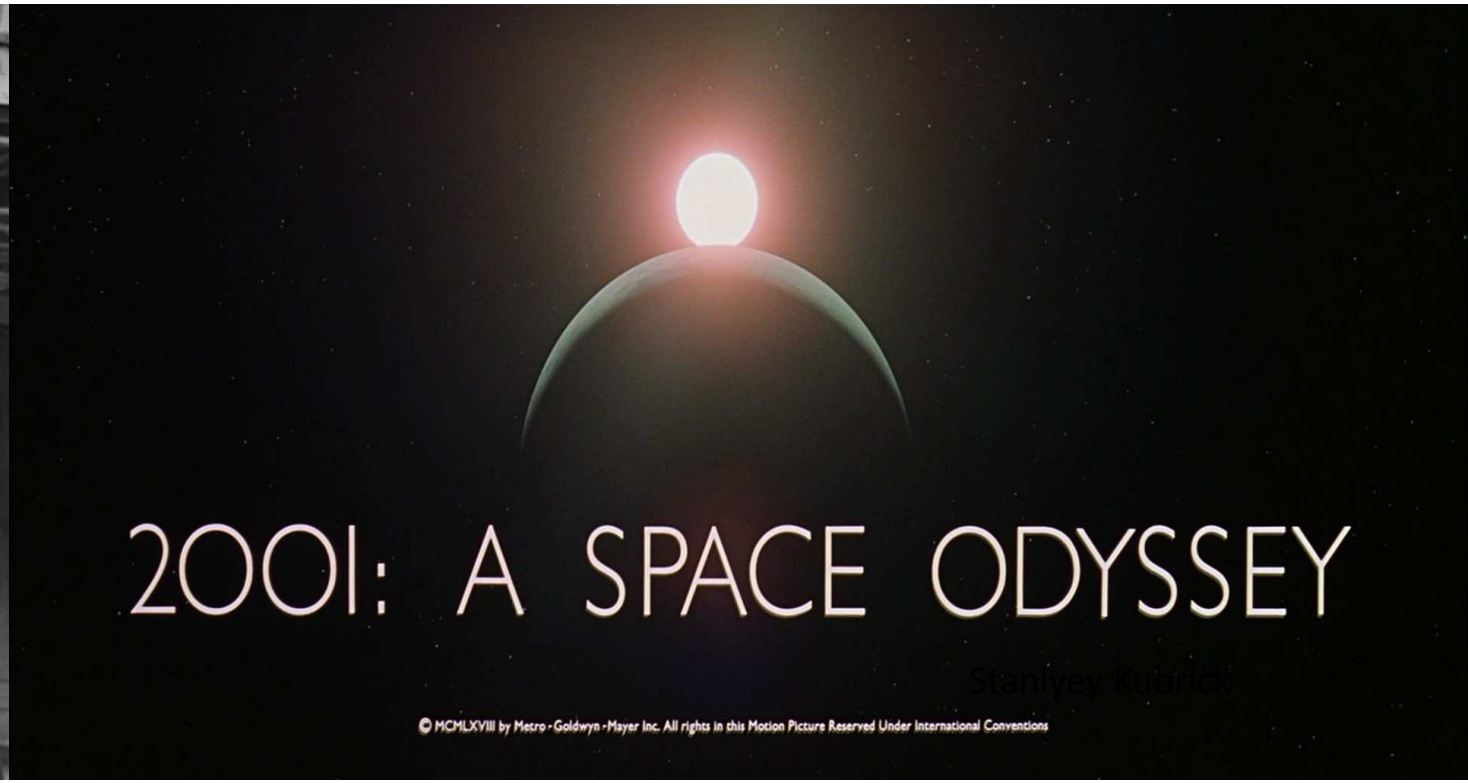
- We need to identify our high risk CHD, cardiomyopathy, LQT, CPVT adolescent and young adult patients and as we have discussions about sports restrictions we must also have discussions about sex.
- Restricting adolescents and young adults from masturbation and sex is probably unrealistic even if we could identify high-risk patients.
- So at a minimum we should educate them about the importance of staying hydrated, being compliant on beta-blockers, avoiding alcohol at the same time as sex, and educating their partners about their heart condition.
- If arrhythmias exist on a treadmill they likely will exist in bed

We must talk to our college bound CHD, channelopathy and cardiomyopathy patients about sex, drinking alcohol at the same time, stress of college, exams, sleep disruptions and not just condoms and STDs



What Should We Do as Pediatric Cardiologists? (just my two cents)

- If patients are at exceptionally high-risk (LQT with T wave alternans, QTc >550 msec, syncope on beta-blockers or CPVT with ongoing bidirectional ectopy on a treadmill) we should think about putting in an ICD.
- If patients are at moderate risk and there is any concern about noncompliance with beta-blockers, alternative strategies such as a left cardiac sympathetic denervation should be discussed.
- **If patients are at moderate risk and the patient and his/her partner would feel better having an AED in the bedroom we should help provide that level of comfort.**



Sir Arthur Clarke was a British science fiction writer, futurist, undersea explorer and inventor.

Wrote 2001: A Space Odyssey in 1968 and commented that "I don't have all the answers, but the questions are certainly worth thinking about"

***Screenplay for Space Odyssey 1968 (Stanley Kubrick Directed)**

But I do have one answer…… the wait is over


Mitchell Cohen, MD
Mitchell Cohen, MD
Thank You


The Philadelphia Inquirer
AT LAST!


E-A-G-L-E-S!
THE TEAM THAT FINALLY GAVE PHILLY ITS SUPER ENDING
The Philadelphia Inquirer
DAILY NEWS
COVER NOT FINAL