Recurrent Syncope of Unclear Etiology

Paul Stephens Jr.,MD

February 23, 2018

NO DISCLOSURESS

Definition of Syncope

- Sudden and transient LOC resulting from a reversible impairment of cognitive function, usually from cerebral hypoxemia
- Brief duration 3-15 seconds
- Spontaneous and complete recovery

Epidemiology

- Up to 50% of young adults can recall at least one past episode
- Females>males
- Peak age 10-30 years
- Recurrence rate 30-50% within five years
- Approximately 120 per 100,000 population seek medical attention

Table 1 Causes of pediatric syncope

Cardiovascular causes
- •Arrhythmias
  - Complete heart block, Sick sinus syndrome, Tachyarrhythmias, Long QT syndrome, Brugada syndrome
- •Structural heart disease
  - Valvular heart disease such as aortic stenosis, mitral valve prolapsed, coronary artery anomalies, hypertrophic obstructive cardiomyopathy, primary pulmonary hypertension, Eisenmenger syndrome
- •Cardiac tumors

Neurocardiogenic/ vasodepressor/ vasovagal/ neurally-mediated syncope

Orthostatic hypotension

Postural orthostatic tachycardia syndrome (POTS)

Convulsive syncope

Reflex syncope

Psychogenic syncope

Drug and toxin induced

Metabolic causes: Hypoglycemia, electrolyte imbalance, endocrine disorder

Special situations
- •Coughing, sneezing, micturition, defecation, deglutition
- •Hair grooming
- •Trumpet playing, suffocation, weight lifting, diving, stretching

Singhi,
Indian J Ped 2017

Diagnosis

- Far and Away, the most important aspect of the evaluation is a **DETAILED** personal history, even if it is painful for the examiner to listen to
- The answer is usually in the history
- Eyewitnesses or Earwitnesses can be of tremendous help
- Frequent episodes of recurrent syncope or an abnormal history = during or immediately after exertion, strong emotion, supine
- Family history may be helpful: Recurrent benign syncope, Arrhythmias, CM, Sudden Cardiac Death (Aborted)

Table 2 Essential elements during history-taking in a child with syncope

- Time of the day when event occurred
- What was the child doing at that time?
- Frequency and duration of the episode
- Premonitory symptoms
- Associated features during the event: light-headedness dizziness, palpitations, shortness of breath, chest pain, headache, pallor, nausea, sweating, visual problems, abnormal hearing
- Duration of episode (usually brief: few seconds to 1–2 min followed by rapid spontaneous recovery within an hour)
- Contributory environmental factors: upright posture, prolonged standing, change in posture, heat, fatigue, hunger, time of last meal, crowding, suffocation, dehydration, emotional surge, stress)
- Any concurrent illness or prior medical history
- Personal history inclusive of menstrual, exercise regimen, drug intake
- Family history of familial/congenital heart disease, metabolic disorder, deafness, psychiatric disorder/ sudden unexplained deaths in children or young adults, pacemaker implantation

Singhi, Indian J Ped 2017

Typical "Patho"physiology of Most Fainters

- Precipitating factors include diminished intravascular volume and gravity provoked decreased venous return →
- mechanoreceptors (C fibers) in atria and inferolateral wall of LV, pulmonary artery →
- Sympathetic stimulation with vigorous cardiac contraction of an underfilled ventricle→
- Sympathatic inhibition and parasympathetic stimulation →
- Paradoxical bradycardia and hypotension→
- Decreased medulla oblongata cerebral perfusion → prodrome and symptoms

Physiology of Most Syncopal Patients

- Typical ages : during and after growth spurt
- Typical times: 10 am-2 pm, Week-ends and holidays, Winter and Summer
- Typical people: Adolescents, females, middle and high schoolers, catholics, choir members,
- Typical sites, in and on way to kitchen, in and around bathroom, health class, church, planes
- Controversary over mechanism and sequence of events: cerebral hypoxia and subsequent lack of stimulation of alpha receptors, overstimulation of beta receptors, other circulating substances

2. CARDIOINHIBITORY VASOVAGAL SYNCOPE

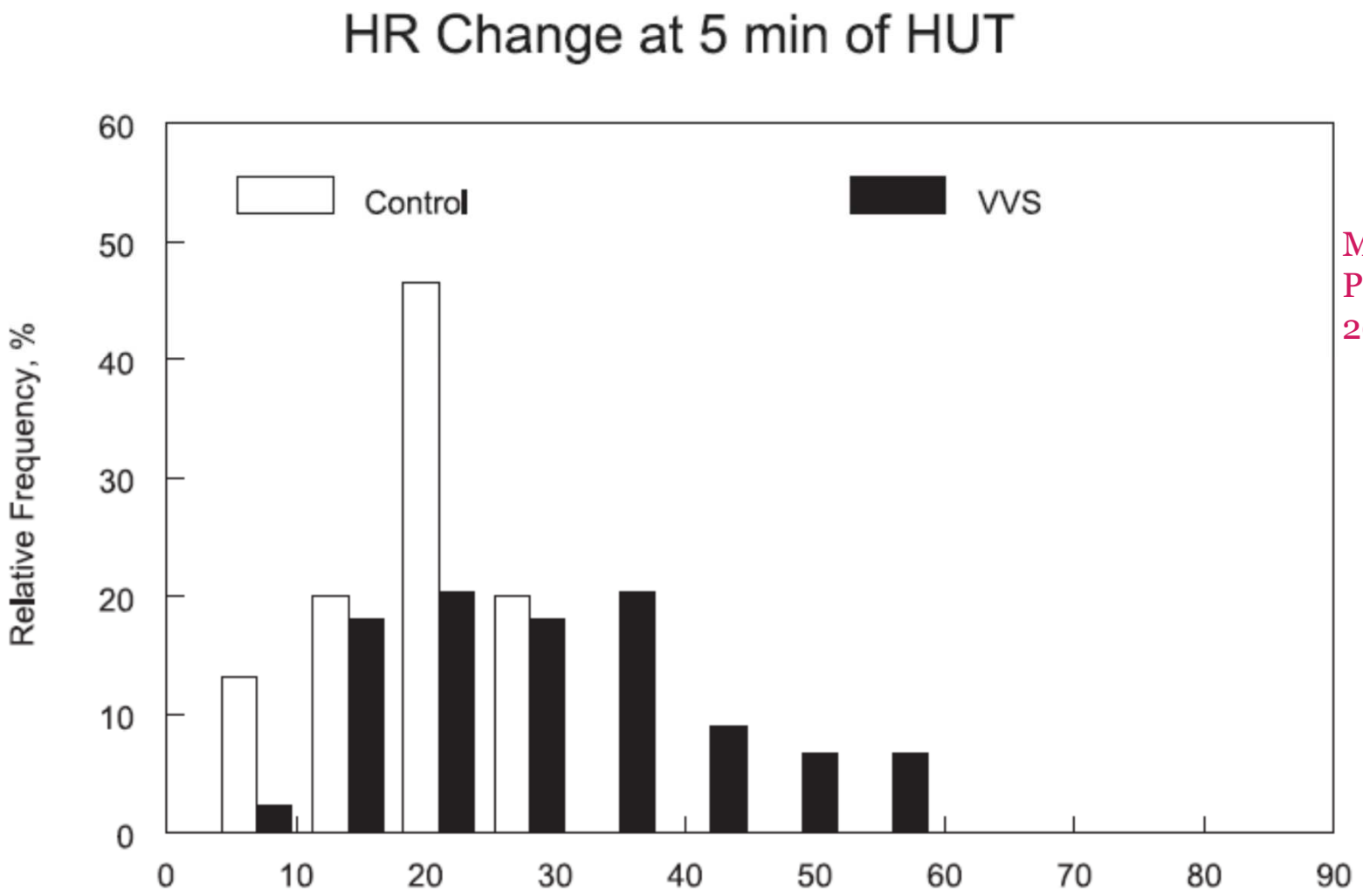
As modified VASIS classification [14], vasovagal syncope presents the following response patterns to tilt testing:

i. Type 1 mixed: there is drop in blood pressure (BP) and heart rate (HR). BP falls before HR falls. The ventricular rate does not fall to less than 40 beats per minute.
ii. Type 2A, cardioinhibition without asystole: there is a drop in HR, with ventricular rate less than 40 beats per minute for more than 10 seconds.
iii. Type 2B, cardioinhibition with asystole: there is asystole longer than 3 seconds duration. BP decrease occurs simultaneously or before HR fall.
iv. Type 3 vasodepressor: there is decrease in BP below 80 mmHg, but HR does not fall more than 10%, from its peak, at the time of syncope.

DaSilva, Pacemaker Indications in Syncope 2016

Postural Heart Rate Changes in Young Patients With Vasovagal Syncope

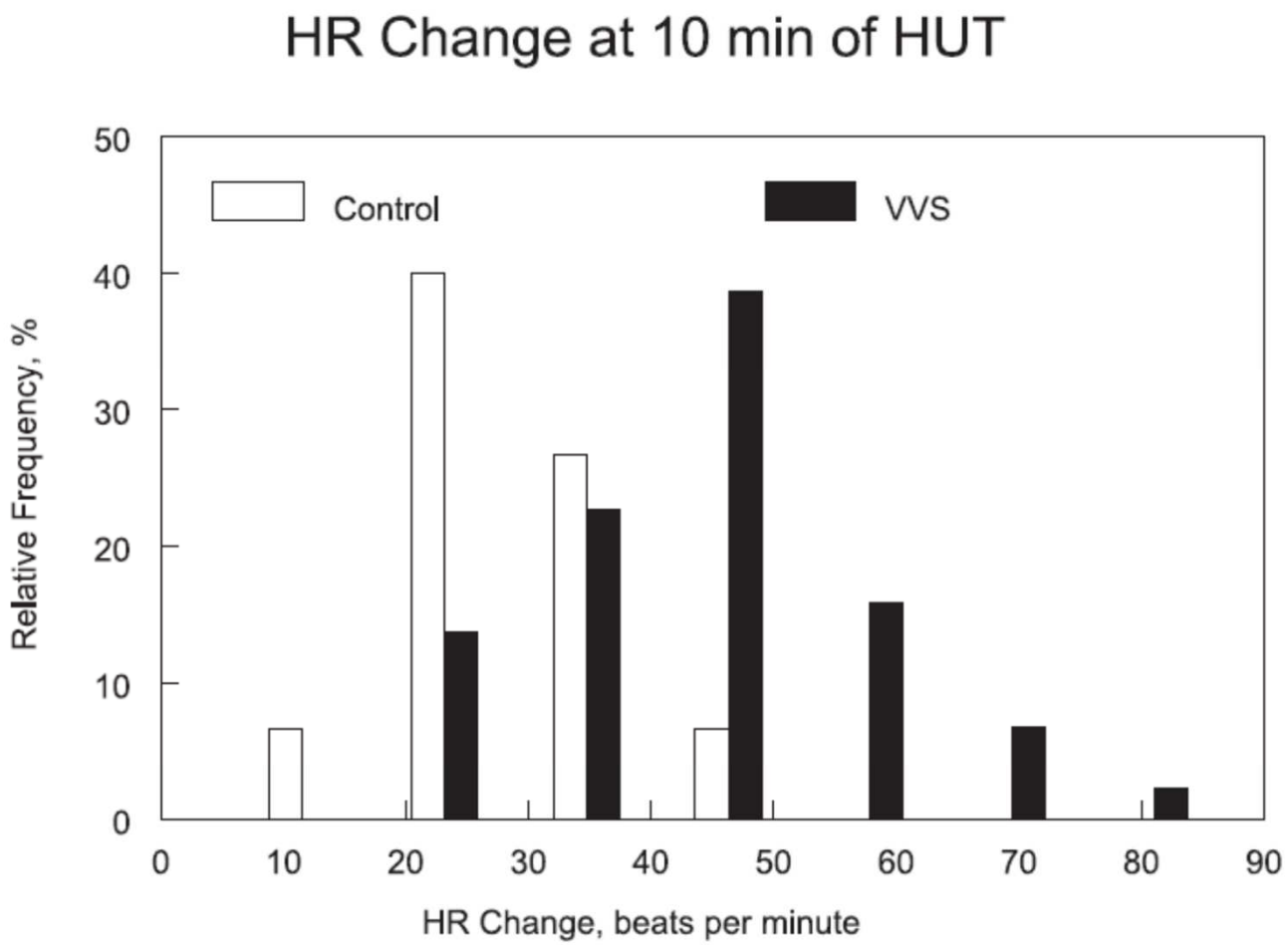
Marvin S. Medow, PhD,[a,b] Sana Merchant, MD,[a] Melissa Suggs, BS, MS,[a] Courtney Terilli, BSN,[a] Breige O'Donnell-Smith, MD,[a] Julian M. Stewart, MD, PhD[a,b]



Meadow Pediatrics 2017

FIGURE 3

Changes in HR (in beats per minute) binned, at 5 minutes (upper panel) and 10 minutes (lower panel) after the imposition of an HUT. HR changes are depicted as their relative frequency (%) of occurrence.



Meadow Pediatrics 2017

FIGURE 3
Changes in HR (in beats per minute) binned, at 5 minutes (upper panel) and 10 minutes (lower panel) after the imposition of an HUT. HR changes are depicted as their relative frequency (%) of occurrence.

Mechanisms of Vasovagal Syncope in the Young: Reduced Systemic Vascular Resistance Versus Reduced Cardiac Output

Julian M. Stewart, MD, PhD; Marvin S. Medow, PhD; Richard Sutton, MB, BS, DSc; Paul Visintainer, PhD; David L. Jardine, FRACP, MD; Wouter Wieling, MD, PhD

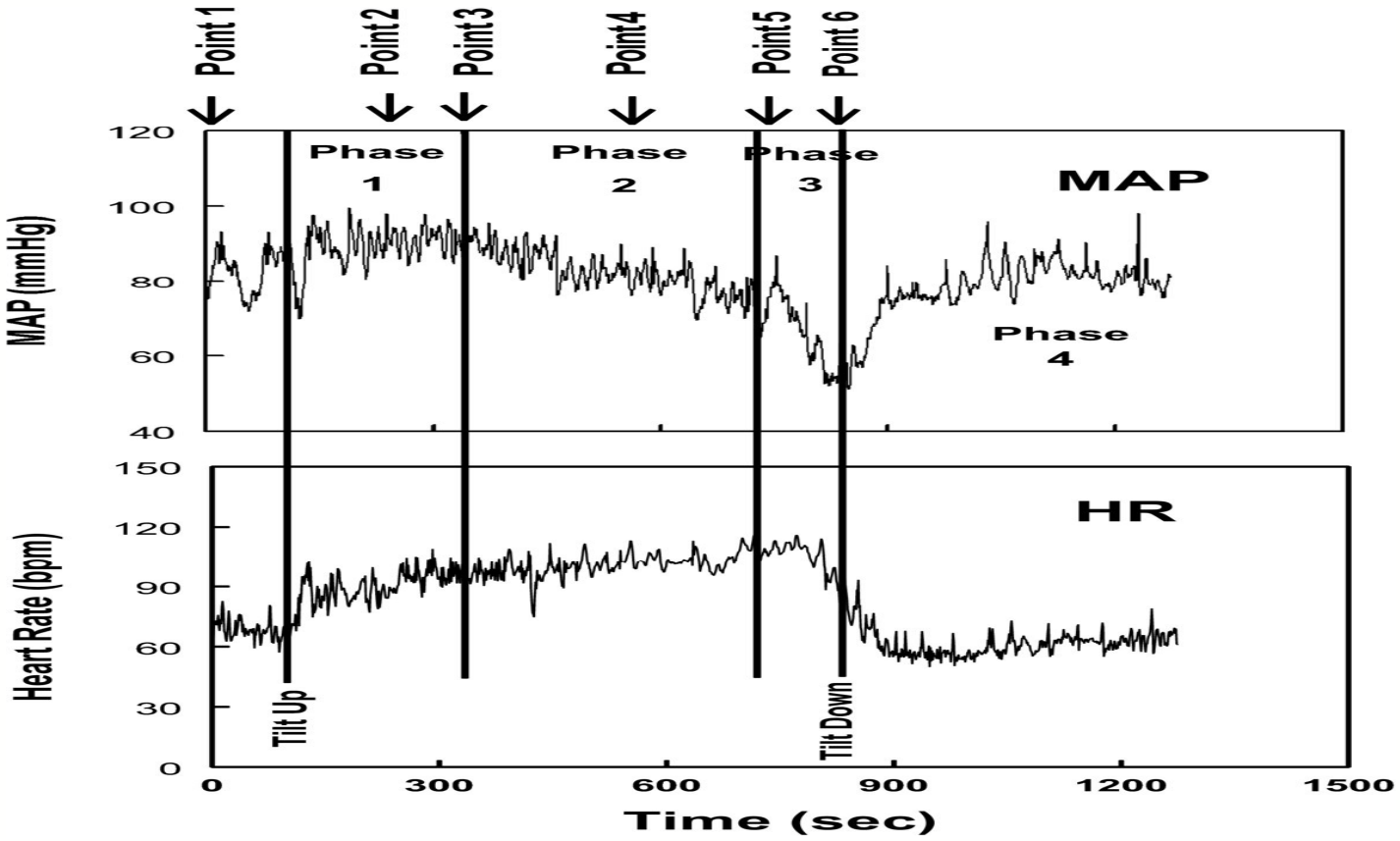
***Background*—**Syncope is a sudden transient loss of consciousness and postural tone caused by cerebral hypoperfusion. The most common form is vasovagal syncope (VVS). Presyncopal progressive early hypotension in older VVS patients is caused by reduced cardiac output (CO); younger patients have reduced systemic vascular resistance (SVR). Using a priori criteria for reduced CO (↓CO) and SVR (↓SVR), we studied 48 recurrent young fainters comparing subgroups of VVS with VVS-↓CO, VVS-↓SVR, and both VVS-↓CO&↓SVR.

***Methods and Results*—**Subjects were studied supine and during 70-degrere upright tilt with a Finometer to continuously measure blood pressure, CO, and SVR and impedance plethysmography to estimate thoracic, splanchnic, pelvic, and calf blood volumes, blood flows, and vascular resistances and electrocardiogram to measure heart rate and rhythm. Central blood volume was decreased in all VVS compared to control. VVS-↓CO was associated with decreased splanchnic blood flow and increased splanchnic blood pooling compared to control. Seventy-five percent of VVS patients had reduced SVR, including 23% who also had reduced CO. Many VVS-↓SVR increased CO during tilt, with no difference in splanchnic pooling, caused by significant increases in splanchnic blood flow and reduced splanchnic resistance. VVS-↓CO&↓SVR patients had splanchnic pooling comparable to VVS-↓CO patients, but SVR comparable to VVS-↓SVR. Splanchnic vasodilation was reduced, compared to VVS-↓SVR, and venomotor properties were similar to control. Combined splanchnic pooling and reduced SVR produced the earliest faints among the VVS groups.

Conclusions*—**Both ↓CO and ↓SVR occur in young VVS patients. ↓SVR is predominant in VVS and is caused by impaired splanchnic vasoconstriction. (J Am Heart Assoc.*** **2017;6:e004417. DOI: 10.1161/JAHA.116.004417.)**

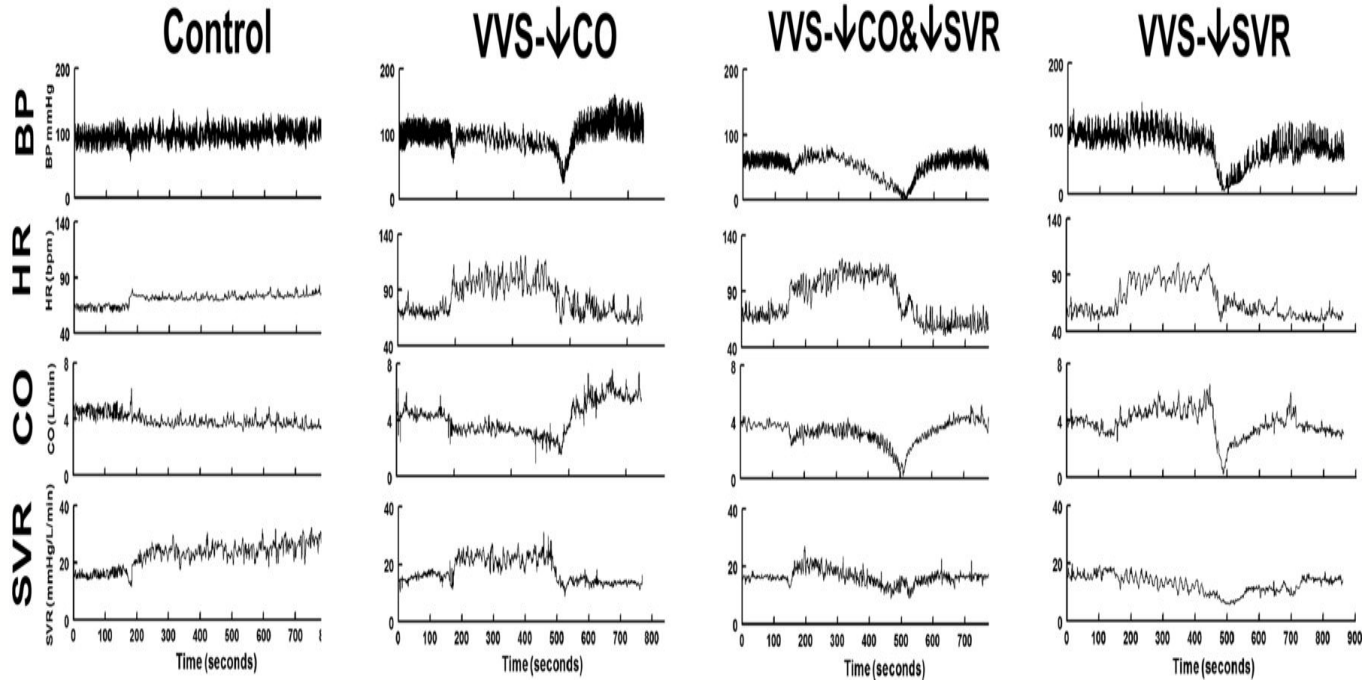
Key Words: cardiac output • syncope • tilt-table testing • vasoconstriction • vasovagal syncope • young

Head-up tilt table test for a vasovagal syncope patient.



Julian M. Stewart et al. J Am Heart Assoc 2017;6:e004417

Blood pressure (BP), heart rate (HR), cardiac output (CO), and systemic vascular resistance (SVR) for representative subjects during head-up tilt table testing.



Julian M. Stewart et al. J Am Heart Assoc 2017;6:e004417

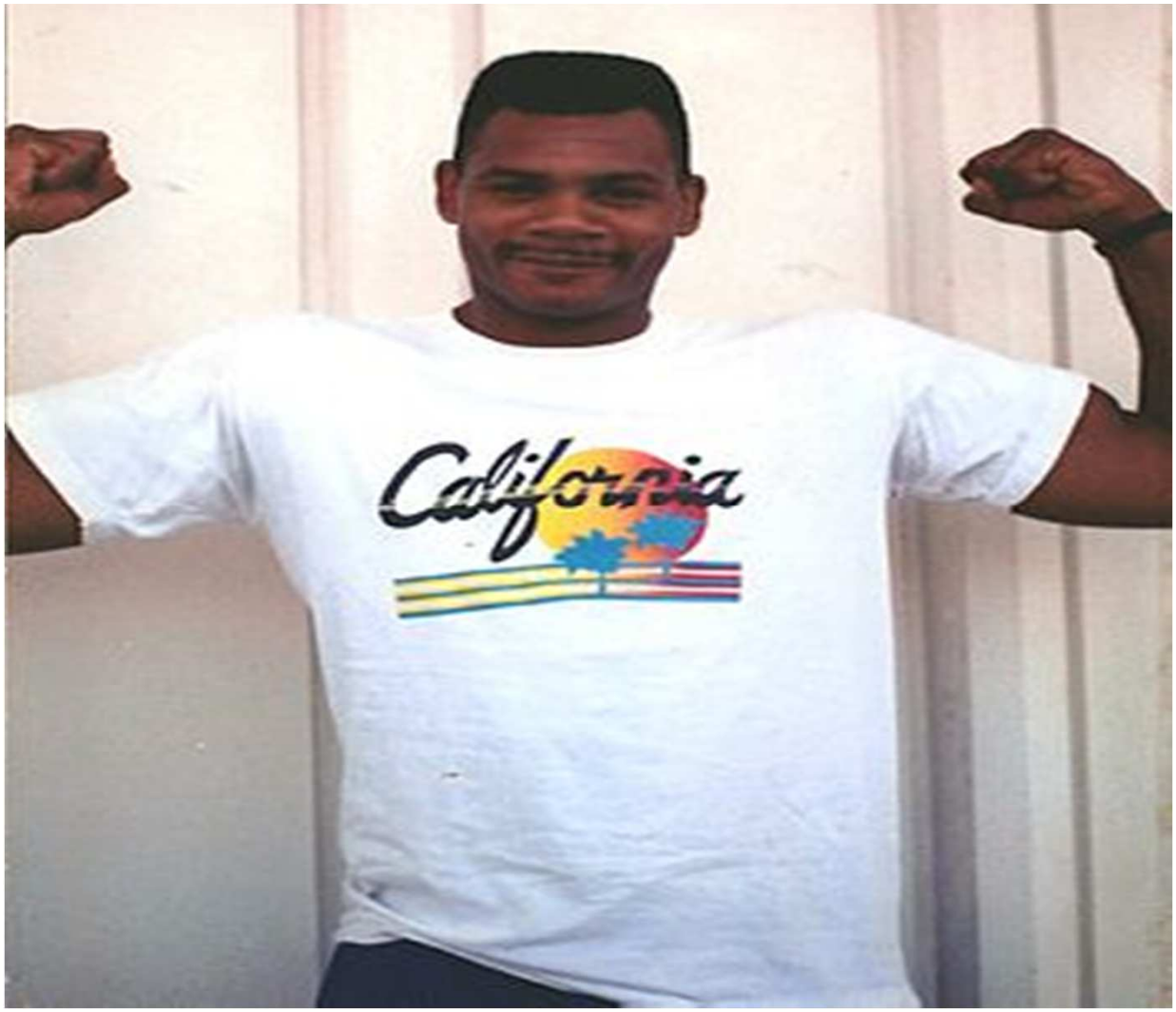
Pathologic Syncope - Etiologies

- Reduced cardiac output
  - Obstruction – LV output – Valvar, Sub-Valvar
  - Obstruction – RV output- Pulm HTN, Pulmonary Stenosis, TV disease
  - Arrhythmia – Long Qt, WPW, CPVT, ARVD, Heart block, Anomalous Coronaries

TABLE 2
Features suggesting a cardiac etiology

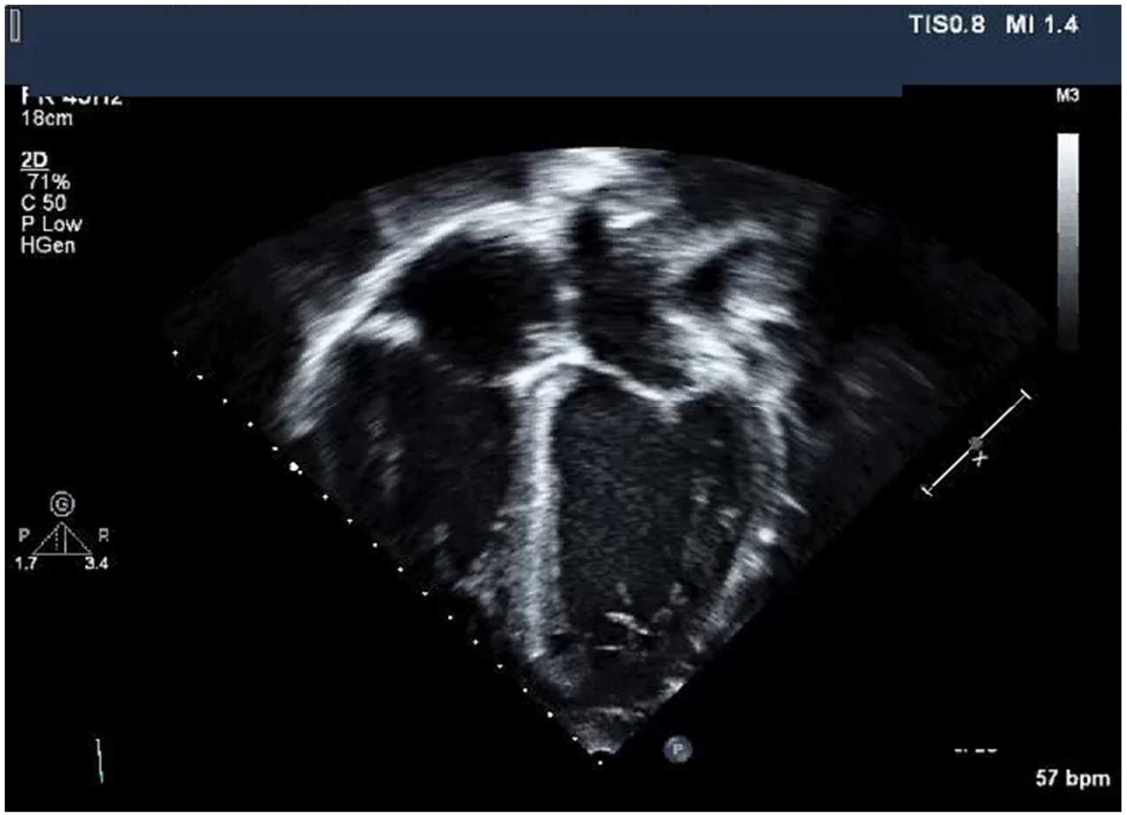
No warning prodromal symptoms
Event precipitated by exertion
Event triggered by emotion, fright or loud noise
Associated palpitations or chest pain
Known heart disease or arrhythmia
Family history: sudden death, cardiomyopathy, cardiac arrhythmia, long QT syndrome or unexplained accidents

Brna, Ped Child Health 2006

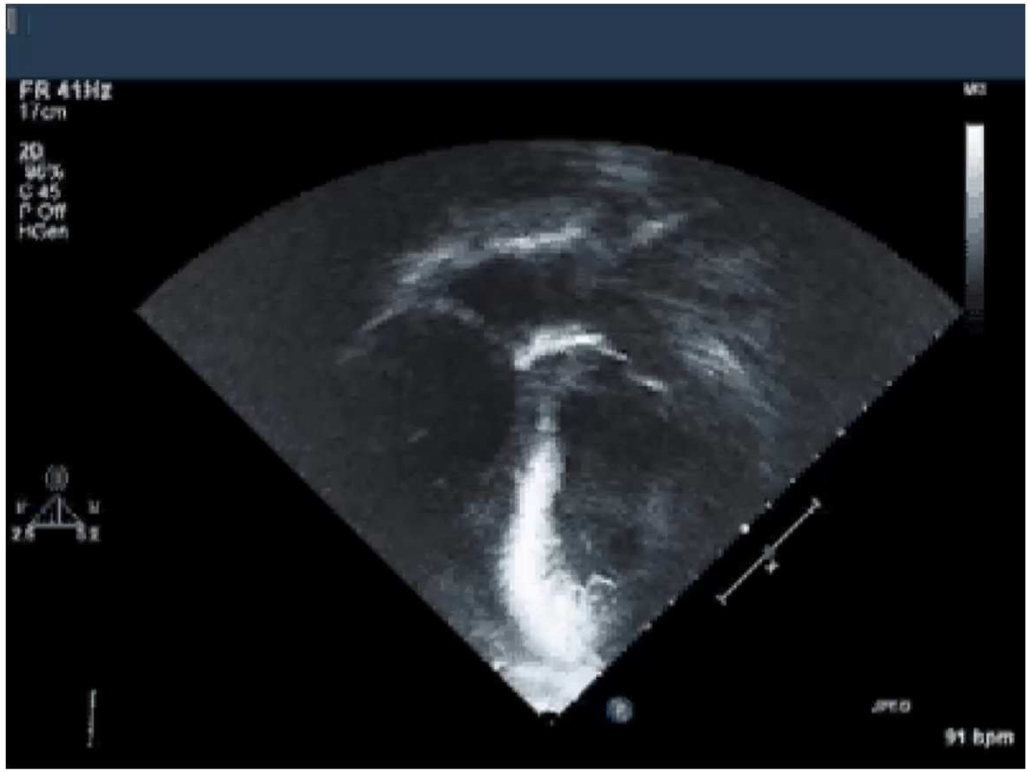

California

Hank Gathers (Feb 11, 1967 – March 4, 1990)

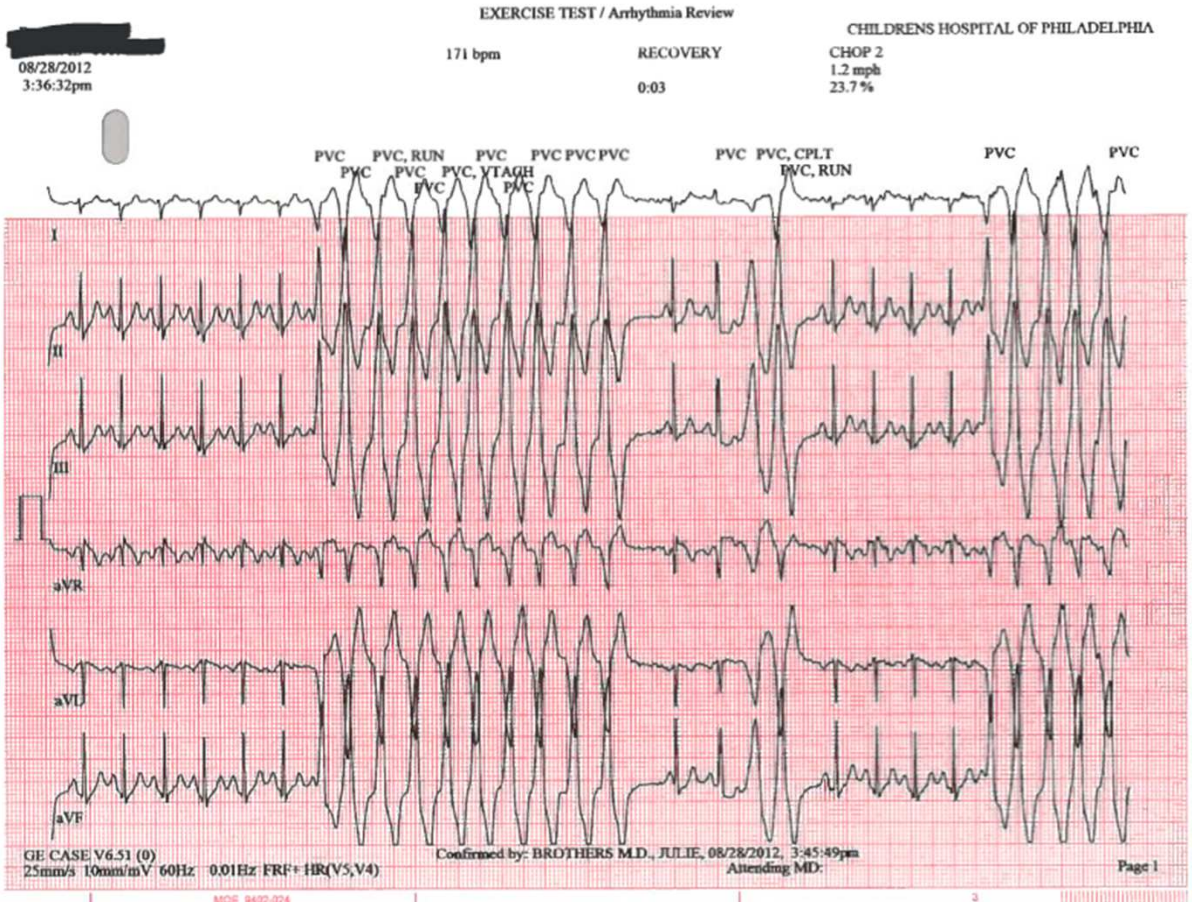
HCM or Athlete's Heart ?



EST : No arrhythmia
During exercise
No definitive
ischemic changes
VO2 >45 ml/kg/min


FR 41Hz
17cm
91 bpm

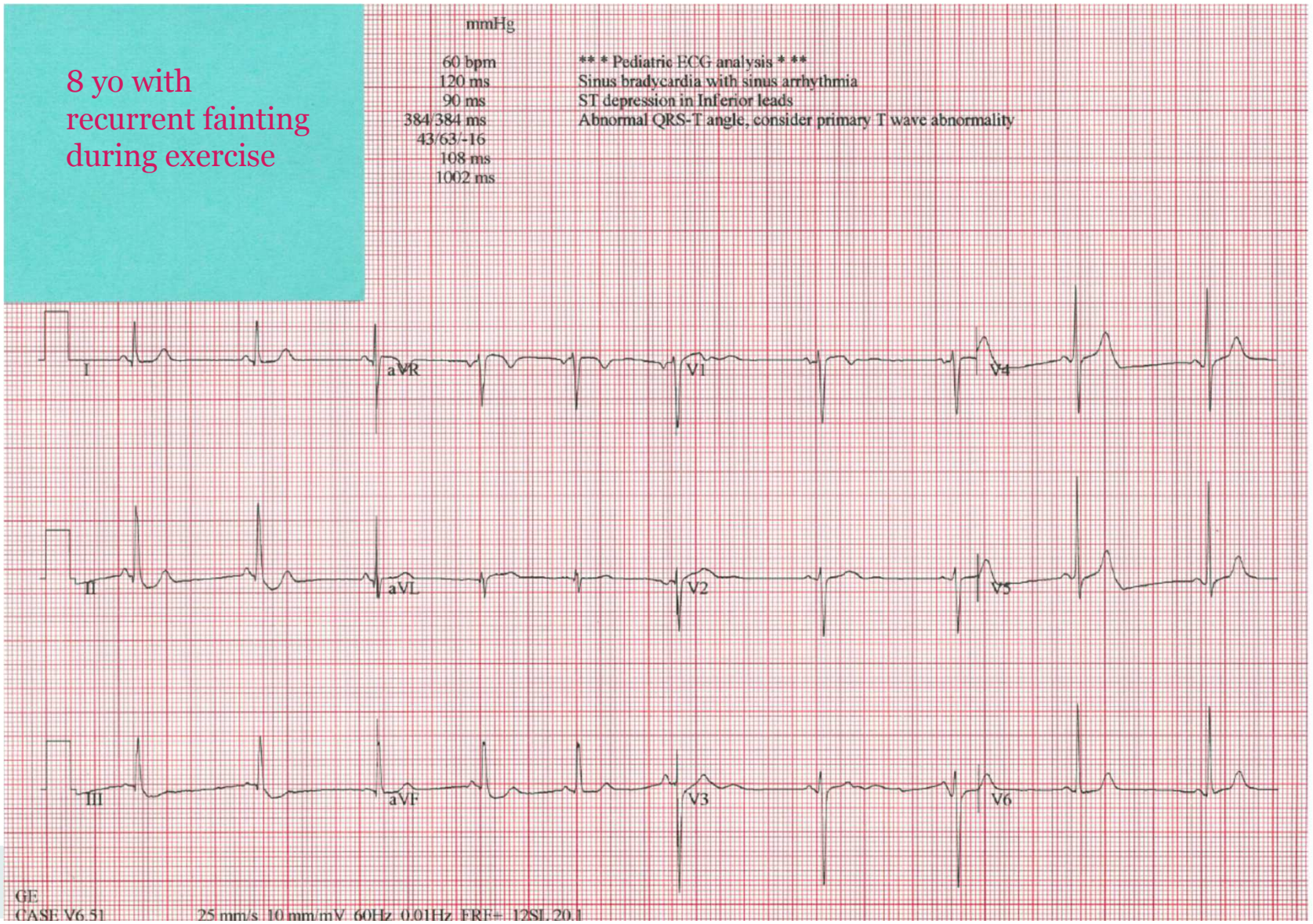
18 yo rower faints and falls out boat while rowing solo at practice. Normal ECG and ECHO



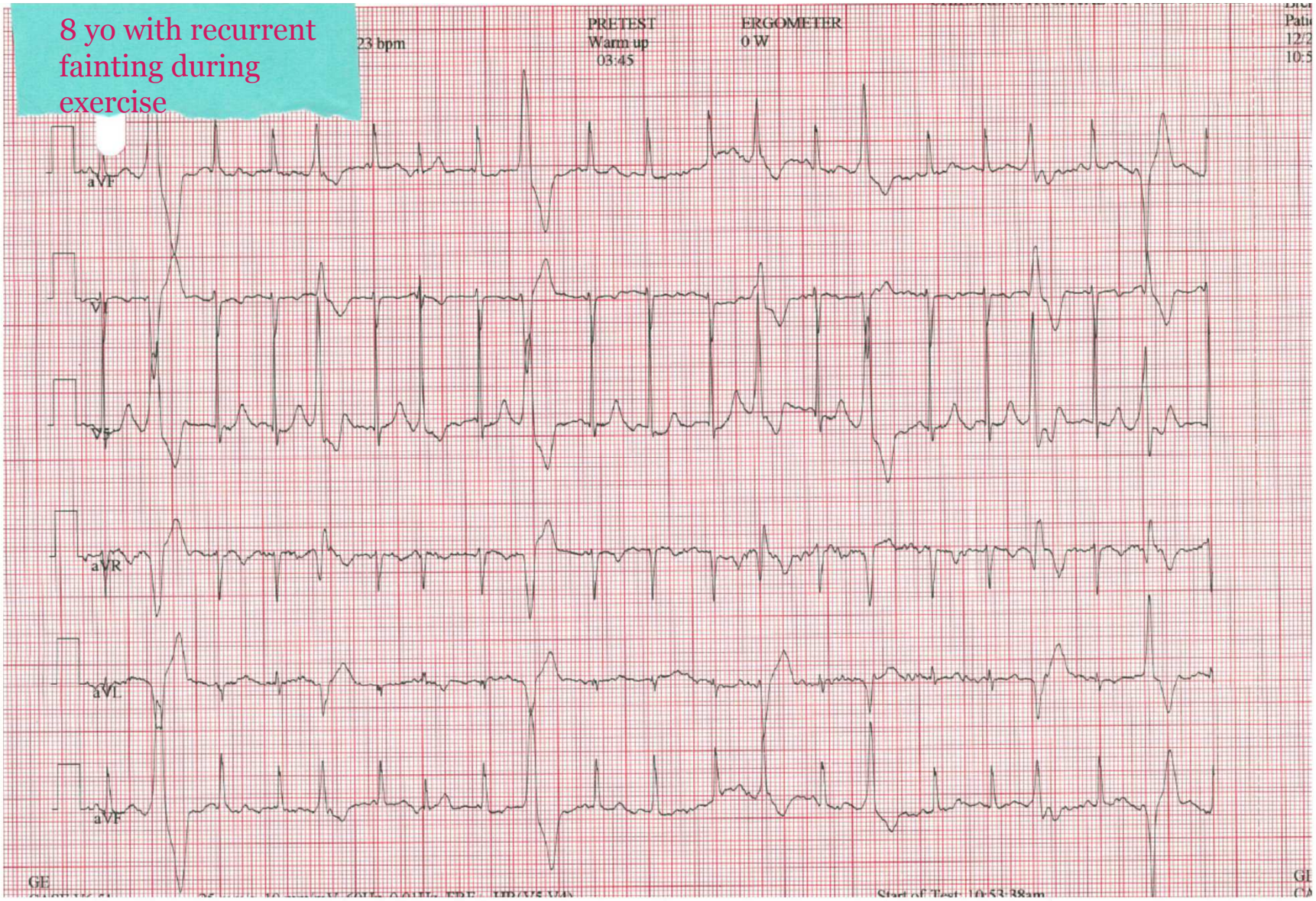
Cardiology 2018

Children's Hospital of Philadelphia Cardiac Center

8 yo with recurrent fainting during exercise



8 yo with recurrent fainting during exercise



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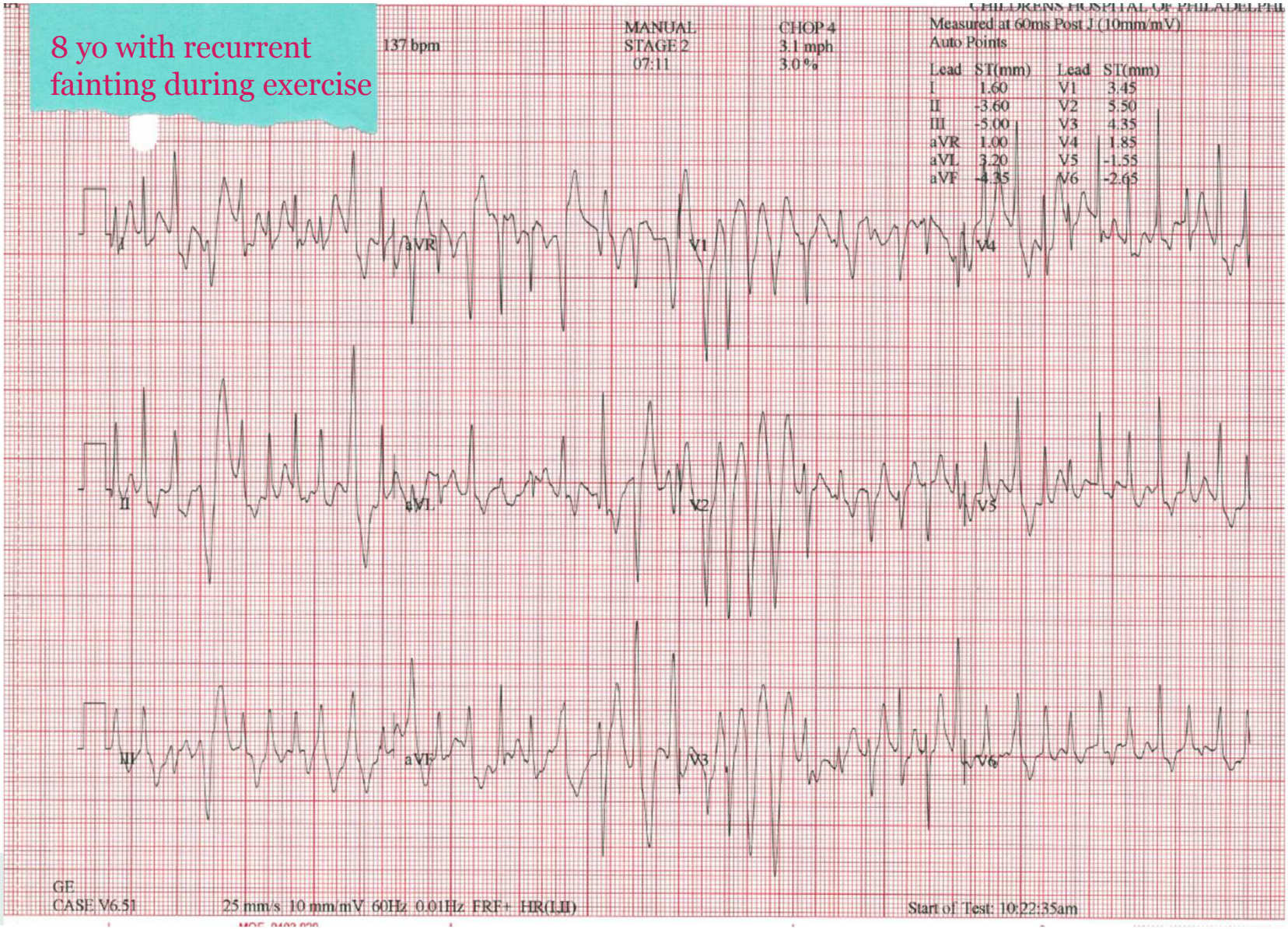
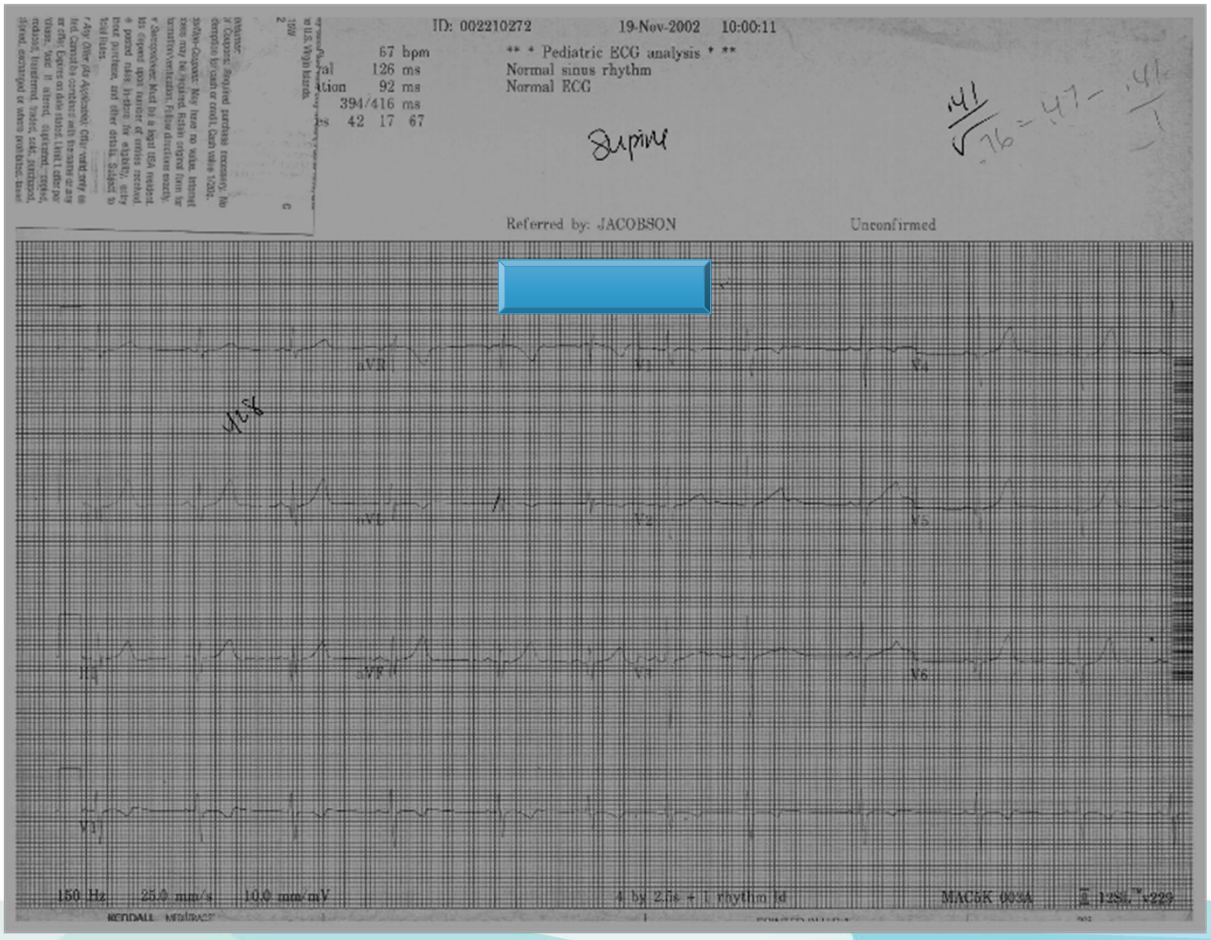


Cardiology 2018

Children's Hospital of Philadelphia
Cardiac Center

11 yo athlete evaluated for syncope at basketball game



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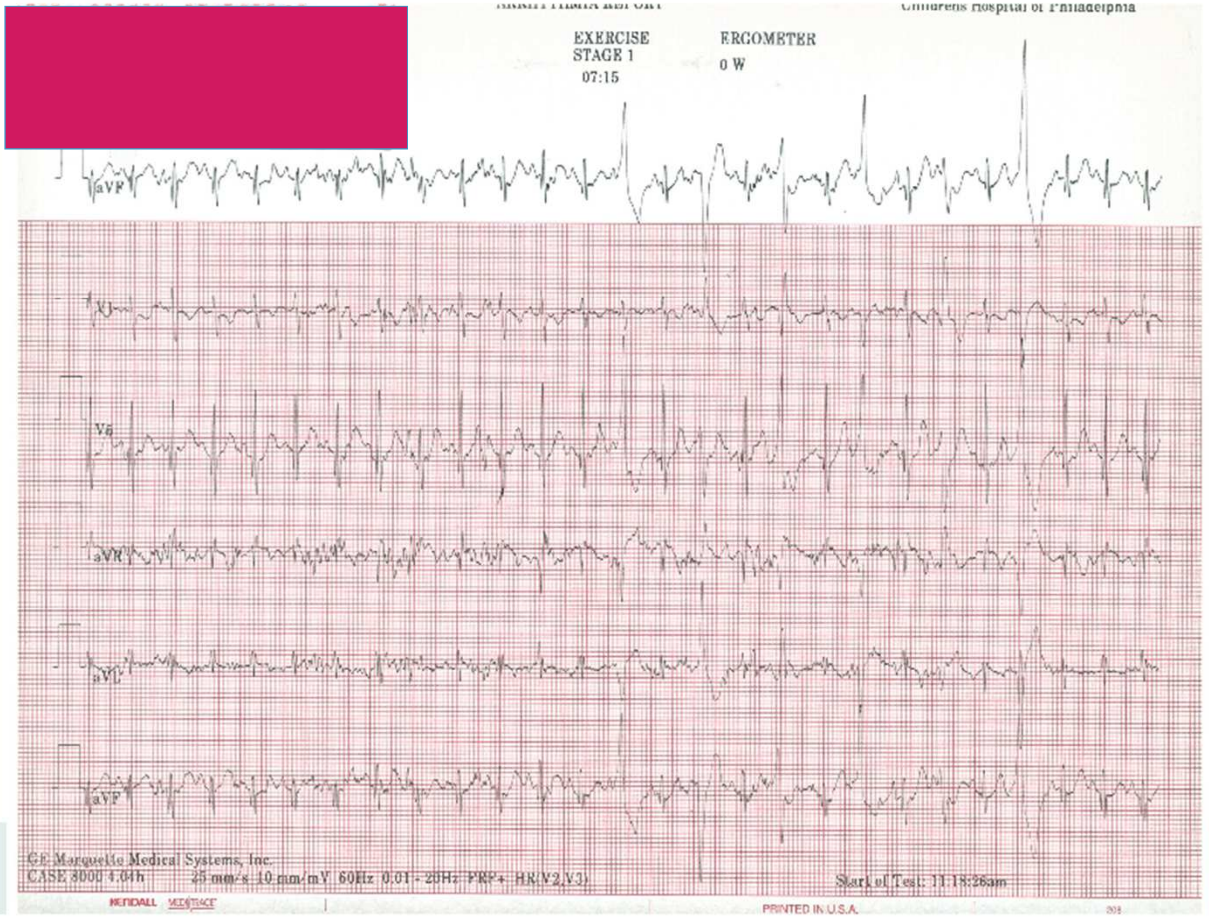

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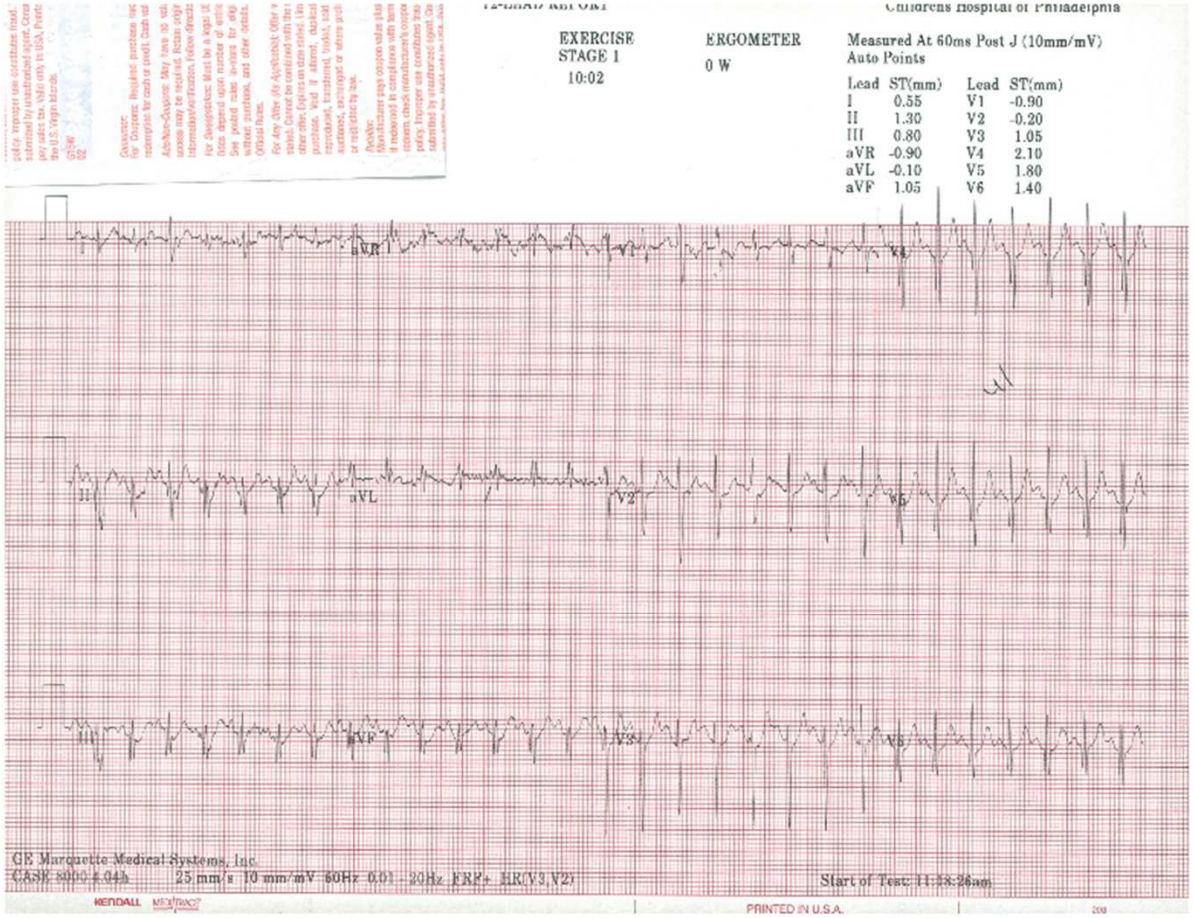

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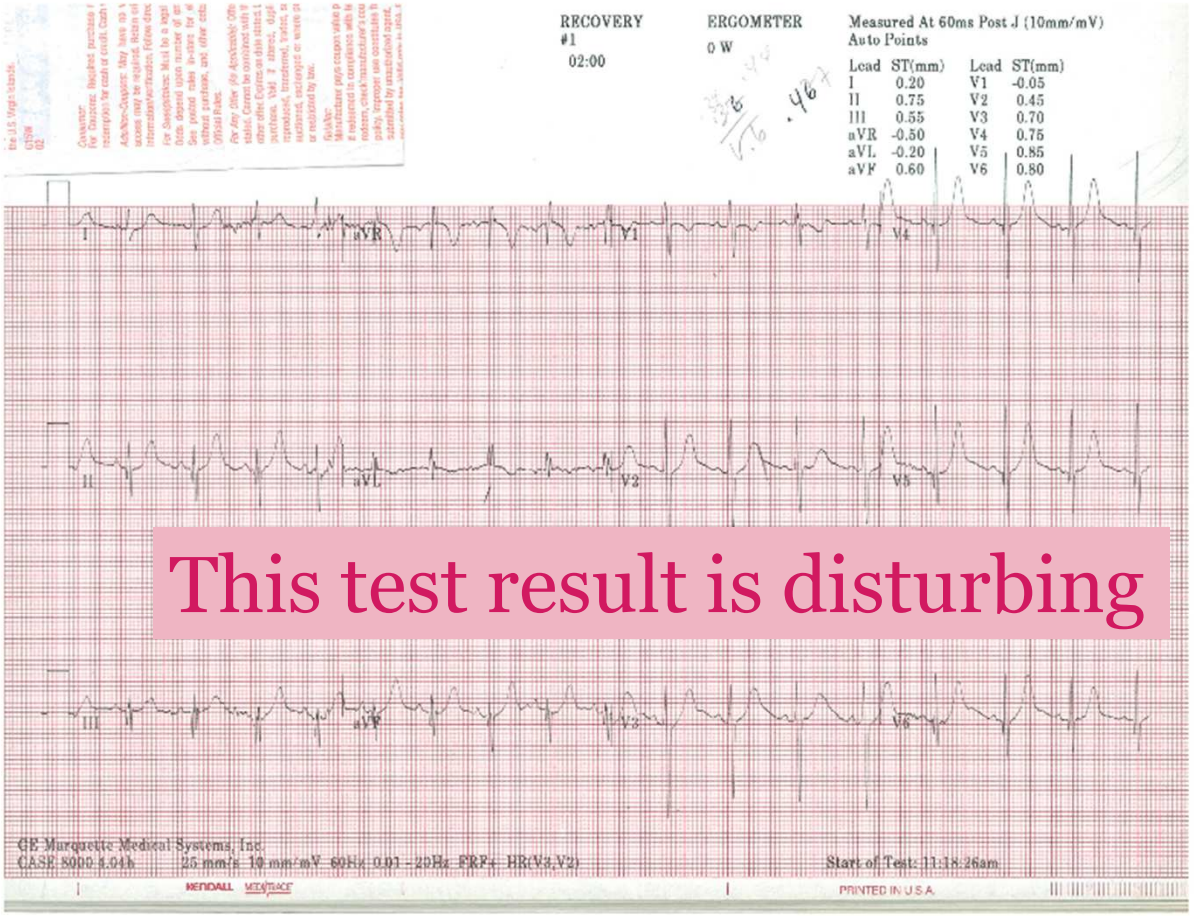


11 yo athlete evaluated for syncope at basketball game

- 11 yo collapsed 1 minute into basketball play-off game, generalized seizure. EMT responded, patient felt better, finished game and 2 additional games on same day. Not very hot August day, patient really "hyped" for game, and well-hydrated
- Referred to Cardiology, arrhythmia suspected.
- FH: 13 yo maternal uncle died in sleep, vomited, thought to have asphyxiated, three seizures prior to death
- Mother has seizure disorder brought on by anxiety/emotion. Dx'd with anxiety disorder, on Dilantin
- First cousin (maternal): seizure disorder brought on by anxiety, has "outgrown" the seizures

Clinical analysis of catecholamine sensitive polymorphic ventricular tachycardia in children

Ge Haiyan Li Xiaomei Rivers Zhang Yi Liuhai Ju Zheng Xiaochun Mei-Ting Li

Chinese Journal of Pediatrics, 2017,55 (12) : 926-931. DOI: 10.3760 / cma.j.issn.0578-1310.2017.12.012

Summary

purpose

To investigate the diagnosis and treatment of catecholamine-sensitive pleomorphic ventricular tachycardia (CPVT) and the causes of delayed diagnosis and misdiagnosis.

method

Retrospective analysis of the history, physical examination, body surface electrocardiogram, cardiogram, treadmill exercise test in 11 children (7 males) diagnosed with CPVT in the pediatric department of the First Affiliated Hospital of Tsinghua University from June 2014 to July 2017 were retrospectively analyzed. , Echocardiography and pathogenic gene information.

result

10 cases of children have repeated emotional or post-exercise syncope history, more than 1 cases of children showed agitation or after exercise palpitation, chest tightness. The first symptom mean age 8.4 (4.0 ~ 13.7) years old, delayed diagnosis time mean 2.4 (0.04 ~ 5.00) years. Two children had a family history of syncope or sudden death. Nine children were misdiagnosed, four were diagnosed with complex arrhythmias, three were diagnosed with syncope, two were diagnosed with epilepsy and were treated for several years with anti-epilepsy. All 11 patients had typical bipolar or pleomorphic ventricular tachycardia in their ambulatory or treadmill exercise tests. Nine cases of children found that CPVT-related pathogenicity or possible pathogenic mutations. Eleven patients were treated with beta-blocker, with an average follow-up of 0.97 years, 6 without clinical symptoms, 4 occasional syncope, and 1 sudden death.

-AVERAGE AT OF PRESENTATION 8 YEARS
-AVERAGE DELAY IN DX- 2 YEARS
9/10 CHILDREN MISDIANOSED
2 PATIENTS DX'D WITH EPILEPSY

Clinical and genetic profile of catecholaminergic polymorphic ventricular tachycardia in Hong Kong Chinese children

TC Yu *, Anthony PY Liu, KS Lun, Brian HY Chung, TC Yung

A B S T R A C T

Objective: To report our experience in the management of catecholaminergic polymorphic ventricular tachycardia in Hong Kong Chinese children.

Methods: This case series study was conducted in a tertiary paediatric cardiology centre in Hong Kong. All paediatric patients diagnosed at our centre with catecholaminergic polymorphic ventricular tachycardia from January 2008 to October 2014 were included.

Results: Ten patients (five females and five males) were identified. The mean age at presentation and at diagnosis were 11.0 (standard deviation, 2.9) years and 12.5 (2.8) years, respectively. The mean delay developed aborted or resuscitated cardiac arrest (n=2) and syncope (n=1). Left cardiac sympathetic denervation was performed in five patients and an implantable cardioverter defibrillator was implanted in another. There was no mortality during follow-up.

Conclusions: Catecholaminergic polymorphic ventricular tachycardia should be considered in children who present with recurrent syncope during exercise or emotional stress. Despite beta-blocker treatment, recurrent ventricular arrhythmias occur and may result in cardiac arrest.

Hong Kong Med J 2016;22:314–9
DOI: 10.12809/hkmj154653

FIG. Patient 1: (a) polymorphic ventricular ectopics, with bidirectional QRS complexes induced by exercise test; and (b) polymorphic ventricular tachycardia, with bidirectional QRS complexes recorded by Holter monitoring

CPVT Hong Kong experience 2016

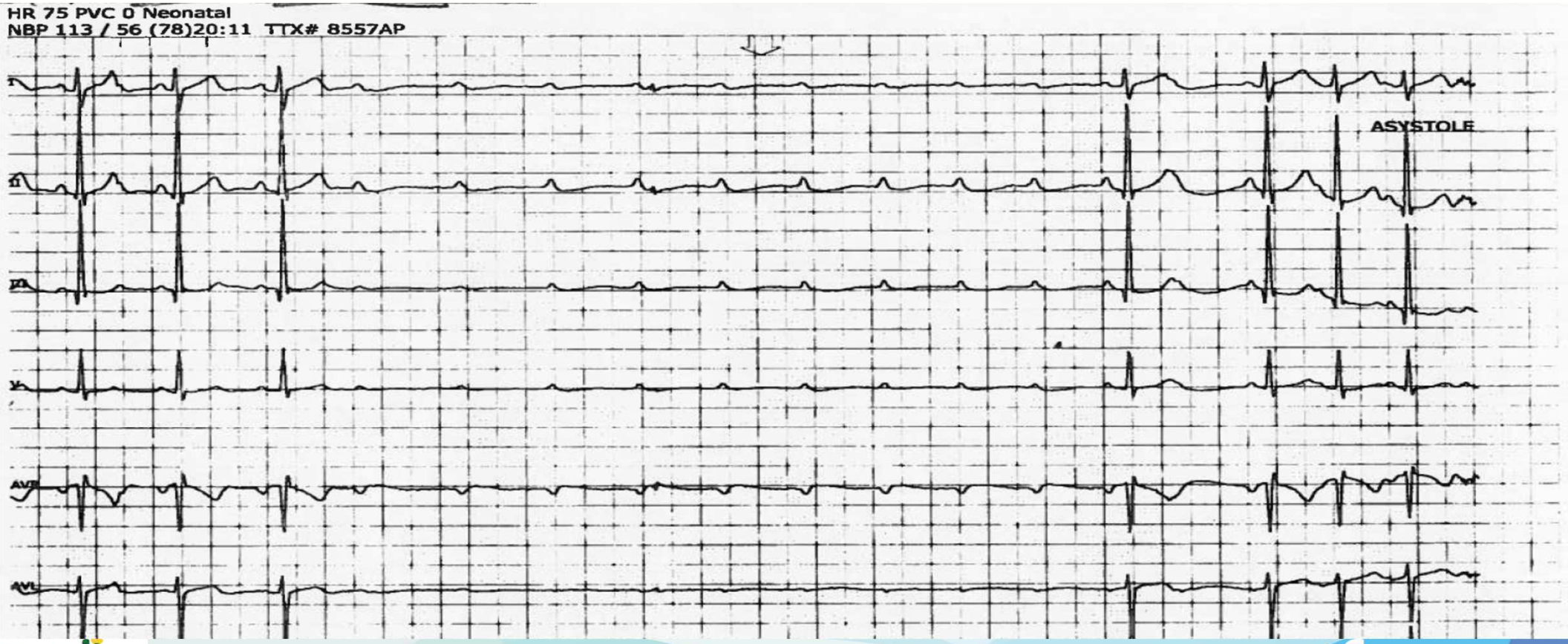
TABLE 1. Demographic data, clinical presentation, diagnostic meth d the threshold heart rate of polymorphic ventricular ectopic and ventricular tachycardia of the 10 patients

Patient No.	Sex	Age of onset (years)	Age at diagnosis (years)	Delay of diagnosis (years)	Follow-up duration (years)	P tation	Diagnostic method	QTc interval of resting ECG (ms)	VE threshold (heart rate, beats/min)	VT threshold (heart rate, beats/min)
1	Male	6.2	6.9	0.7	6.7	Syncope	Isoprenaline infusion	426	120	140
2	Female	11.0	14.1	3.1	5.5	Syncope	Adrenaline infusion	394	110	142
3	Female	9.7	13.0	3.3	6.4	Syncope	Treadmill	416	150	150
4	Female	14.2	14.3	0.1	4.7	Aborted cardiac arrest	Arrest ECG	422	90	126
5*	Male	11.6	14.9	3.4	2.9	Syncope	Treadmill	406	125	NA
6	Female	7.2	9.2	2.0	2.7	Aborted cardiac arrest	Adrenaline infusion	396	110	166
7	Female	13.7	15.1	1.4	2.7	Syncope	Adrenaline infusion	444	126	170
8	Male	13.2	13.2	0	2.4	Aborted cardiac arrest	Adrenaline infusion	380	117	138
9*	Male	14.0	14.4	0.3	2.0	Aborted cardiac arrest	Arrest ECG	423	NA	NA
10	Male	9.3	10.0	0.7	0.7	Syncope	Adrenaline infusion	404	120	130

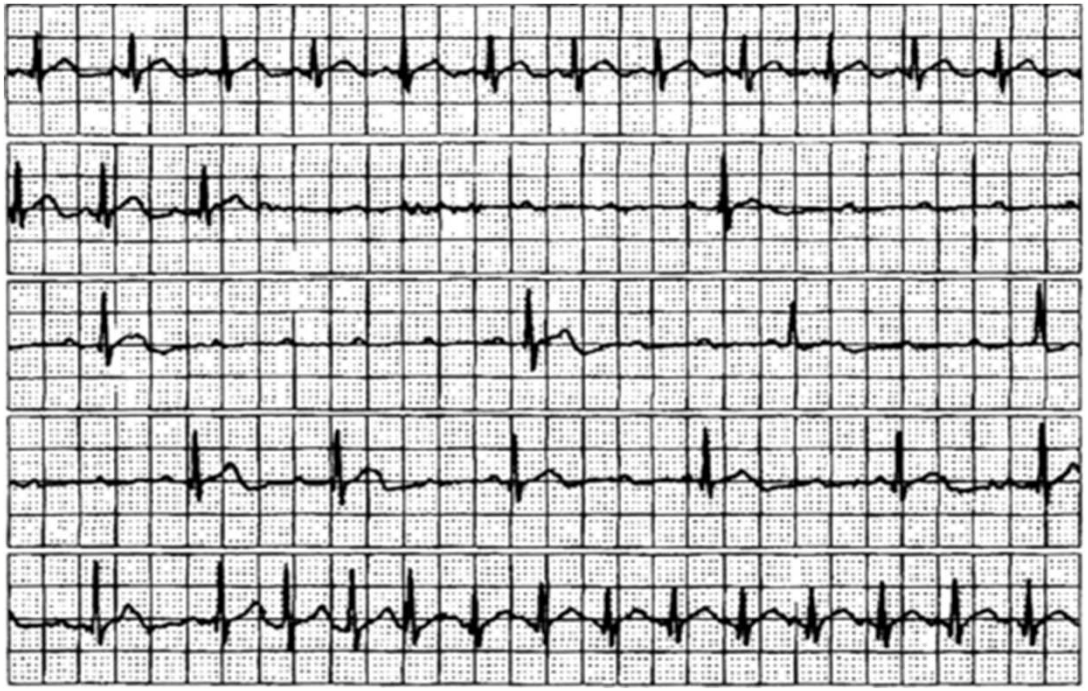
Abbreviations: ECG = electrocardiogram; NA t available; QTc = heart rate–corrected QT; VE = ventricular ectopic; VT = ventricular tachycardia

* Brothers of the same family

13 yo with clusters of atypical syncope



Paroxysmal AV Block in Children with Normal Cardiac Anatomy as a Cause of Syncope



Pass and Silver
Event Recording

Pacing and Clinical Electrophysiology
Volume 31, Issue 3, pages 322-326, 28 FEB 2008 DOI: 10.1111/j.1540-8159.2008.00992.x
http://onlinelibrary.wiley.com/doi/10.1111/j.1540-8159.2008.00992.x/full#f1

SP1

Paroxysmal AV Block in Children with Normal Cardiac Anatomy as a Cause of Syncope

Pass and Silver, Holter Tracing

Pacing and Clinical Electrophysiology
Volume 31, Issue 3, pages 322-326, 28 FEB 2008 DOI: 10.1111/j.1540-8159.2008.00992.x
http://onlinelibrary.wiley.com/doi/10.1111/j.1540-8159.2008.00992.x/full#f2

Slide 38

SP1 Stephens, Paul, 2/18/2018

Table

Characteristics of S Patients w h Paroxysmal A Block

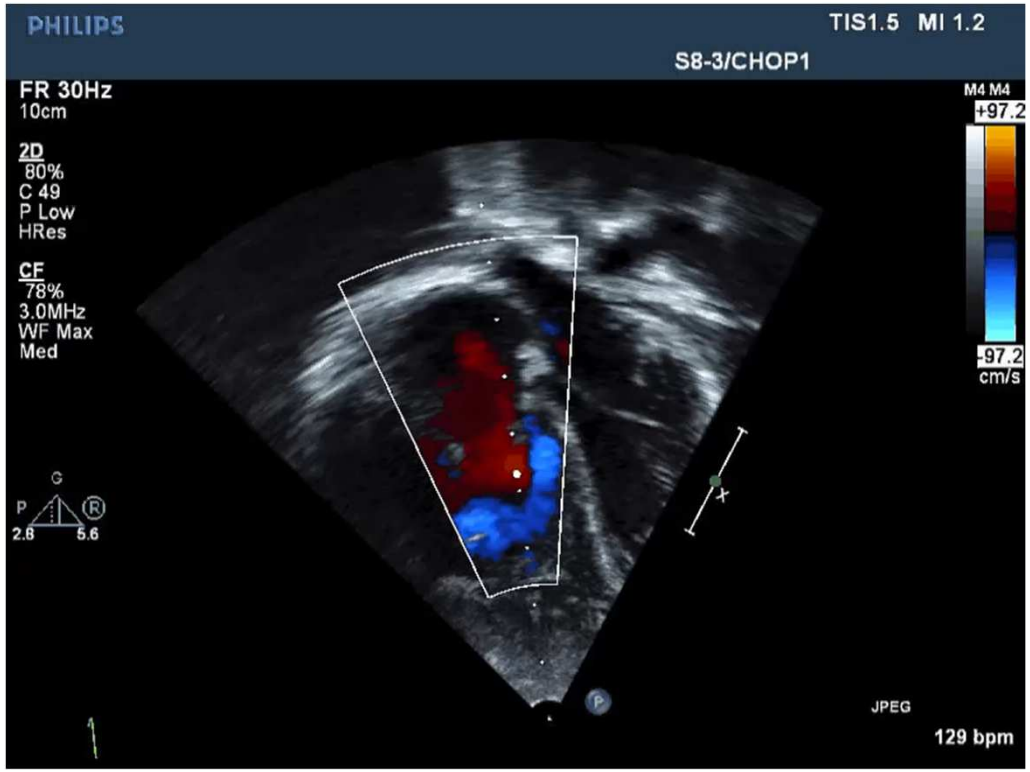
Patient	Age (years)	Sex	Medical History	Preceding Symptoms	Length of Symptoms (years)	Method of Diagnosis	Atrial Rate with AV Block (bpm)	Longest Pause (sec)	Pacing Mode	Follow-up (years)
1	2.5	F	None	Crying out, headache	5	Event recorder	150	17	VVI	0.5
2	8	F	ADHD, asthma	Nausea, syncope while lying flat	6	Cardiac monitor	115	22	VVI	1.75
3	8	F	None	Nausea, syncope while lying flat	4	Cardiac monitor	75	11	VVI	16
4	9	F	None	Crying out, abdominal pain	7	Cardiac monitor	150	14	DDD	10
5	13	M	ADHD	Blurry vision and room spinning while eating	1	Event recorder	75	7	DDD	1.6
6	15	F	None	Strong emotion, sight of blood	11	Holter monitor	75	16	DDD	1.5

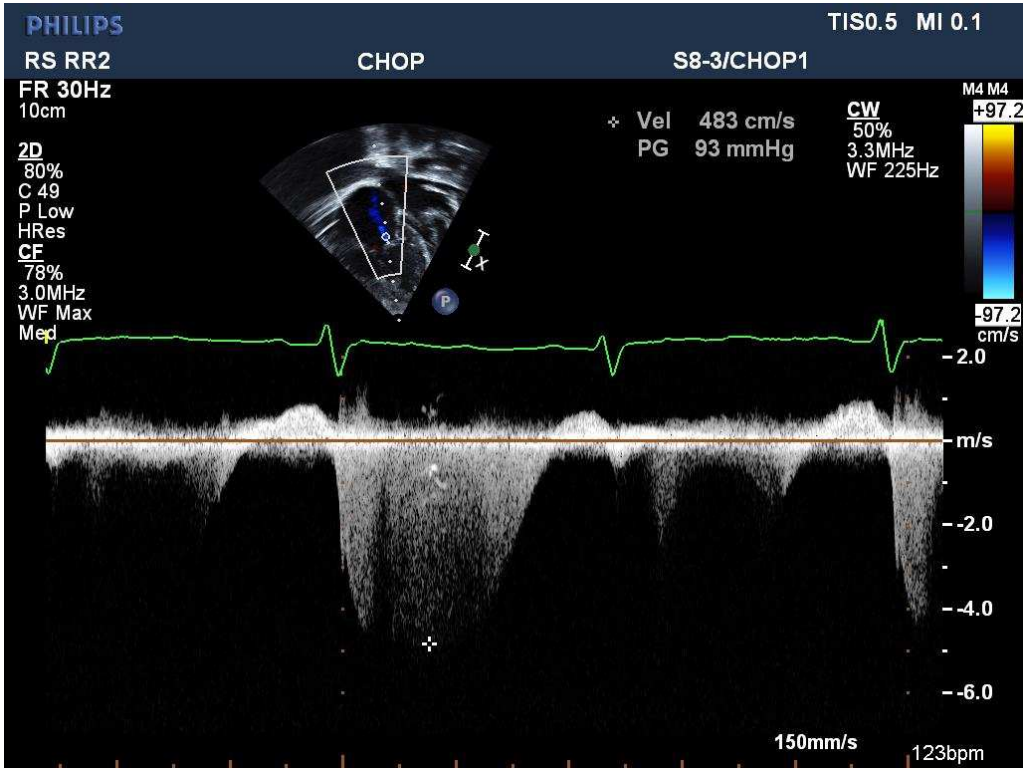
ADHD = Attention Deficit Hyperactivity Disorder.

Young females who have years of fainting, including fainting while lying flat may require an ILR.

Pass and Silver, AV block, 2008

14 mo admitted to Neurology for transient staring, loss of tone for 5-15 seconds




PHILIPS
TIS0.5 MI 0.1
RS RR2
CHOP
S8-3/CHOP1
FR 30Hz
10cm
2D
80%
C 49
P Low
HRes
CF
78%
3.0MHz
WF Max
Med
Vel 483 cm/s
PG 93 mmHg
CW
50%
3.3MHz
WF 225Hz
M4 M4
+97.2
-97.2
cm/s
2.0
m/s
-2.0
-4.0
-6.0
150mm/s
123bpm

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

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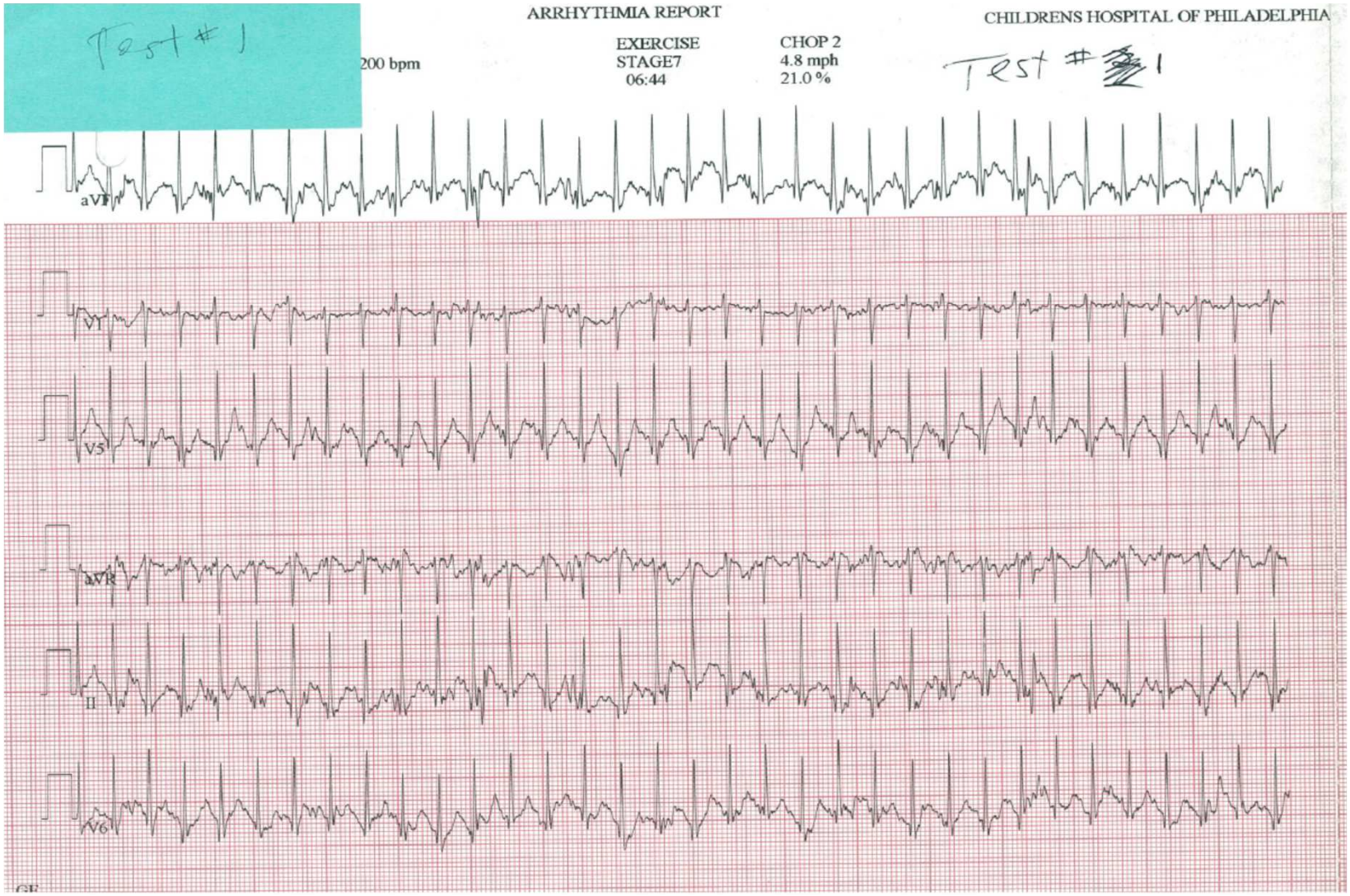
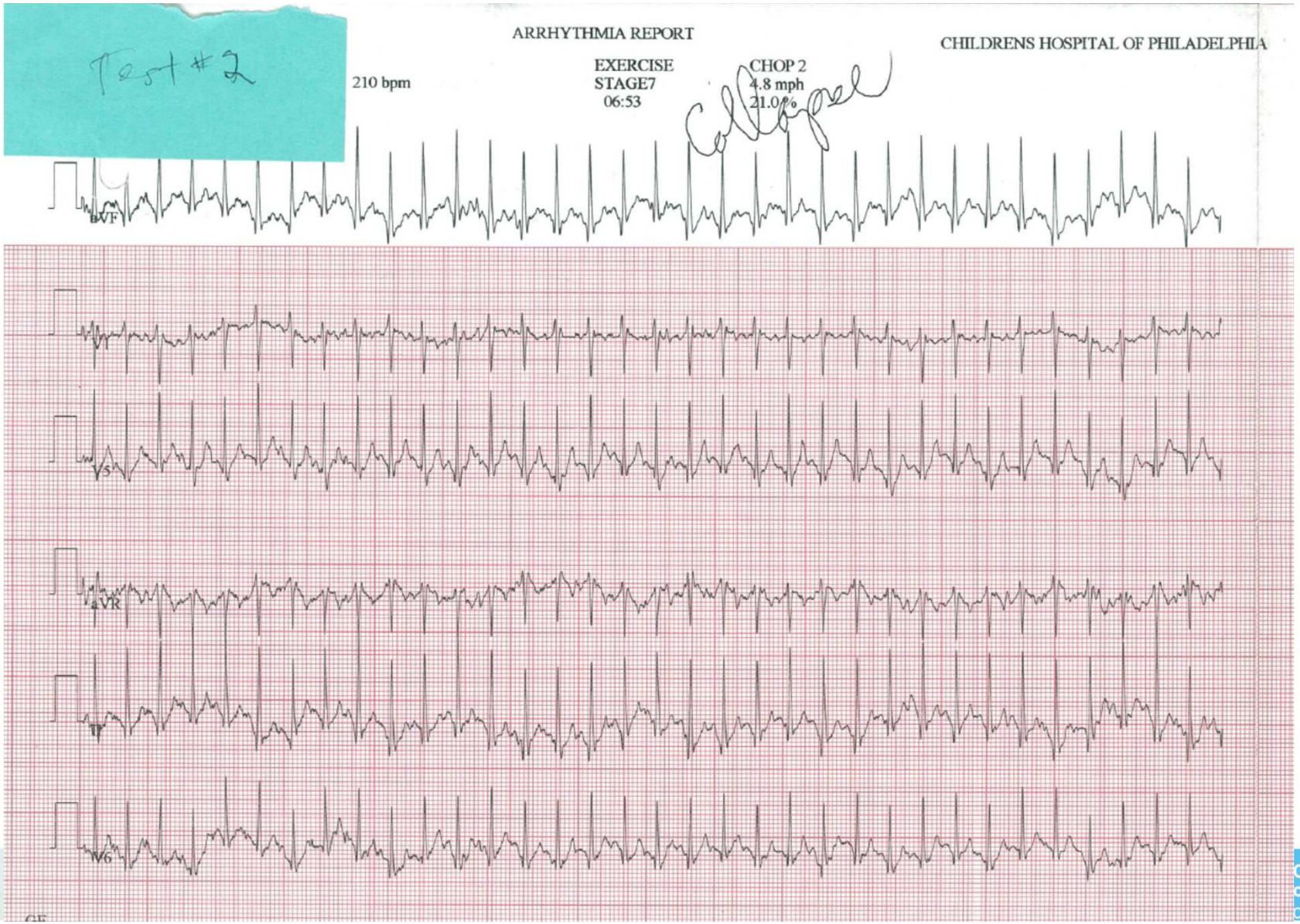
From the *Division of Pediatric Cardiology, Department of Pediatric and Adolescent Medicine; †Department of Molecular Pharmacology & Experimental Therapeutics Windland Smith Rice Sudden Death Genomics Laboratory; and ‡Division of Cardiovascular Diseases, Department of Medicine, Mayo Clinic, Rochester,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

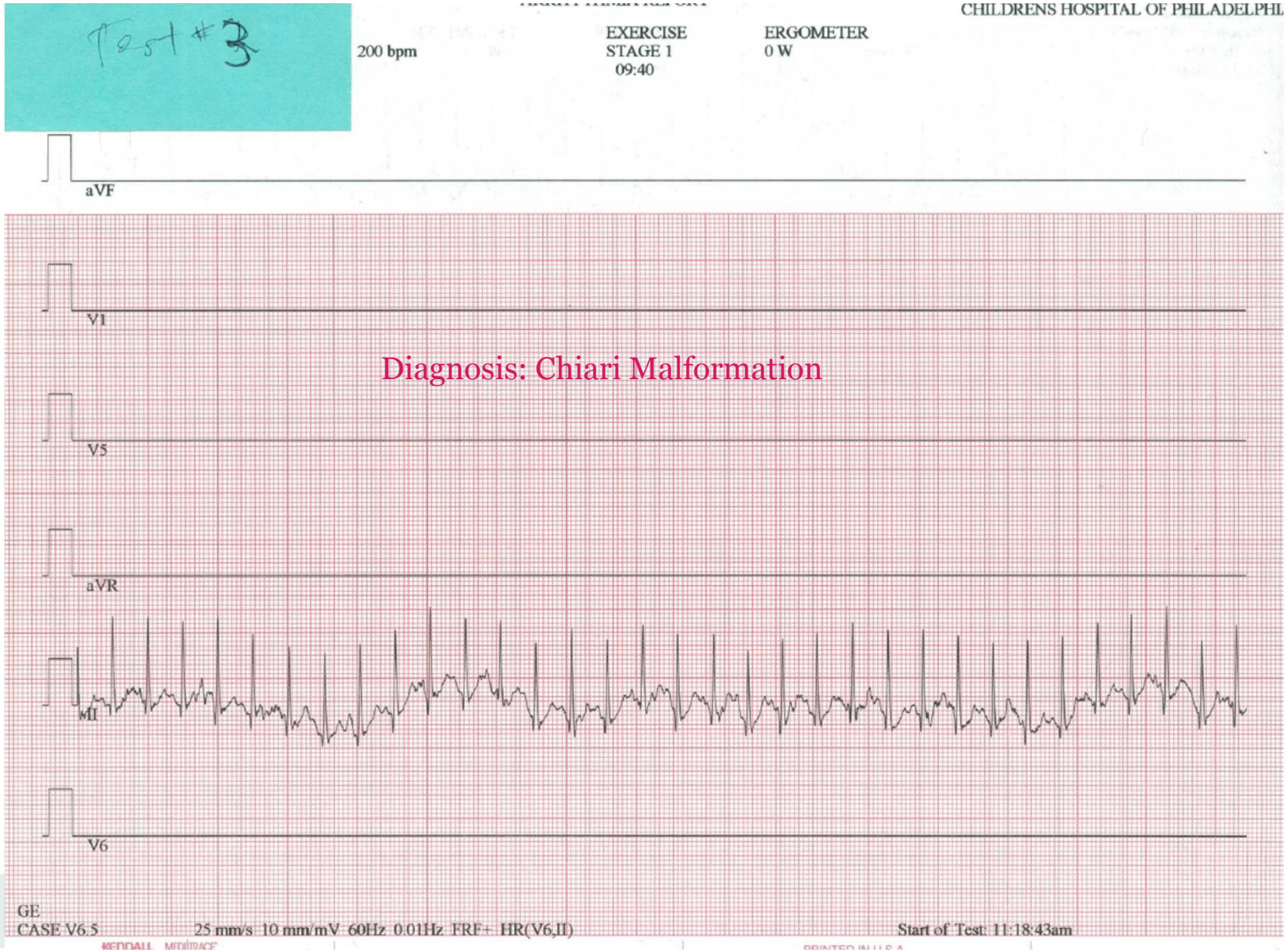
Sudden Cardiac Arrest During Sex in Cardiac Channelopathies. *Background:* **Patients with catecholaminergic polymorphic ventricular tachycardia (CPVT) and long-QT syndrome (LQTS) are susceptible to cardiac events during sympathetic nervous system activation. Herein, we sought to determine the risk of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


Test #1
ARRHYTHMIA REPORT
CHILDRENS HOSPITAL OF PHILADELPHIA
200 bpm
EXERCISE
STAGE7
06:44
CHOP 2
4.8 mph
21.0 %
Test #1
aVF
V1
V5
aVR
II
V6


Test #2
ARRHYTHMIA REPORT
CHILDRENS HOSPITAL OF PHILADELPHIA
210 bpm
EXERCISE
STAGE7
06:53
CHOP 2
4.8 mph
21.0 %
aVF
V5
aVR
II
V6



Evaluation and Tests

- PE is usually normal in Vasovagal syncope
- BP supine and standing
- Increase in heart rate with standing
- ECG
- ECHO, Holter
- TTM, Ziopatch or equivalent, MRI, EPS
- Tilt Table testing?
    - Class IIA indication in adults
- Implantable loop recorders
    - Class IIA in older patients

Treatment of Vasovagal Syncope

- Water
- Water
- Water
- Water
- Water
- Volume amount? Metric Clear Urine thrice daily

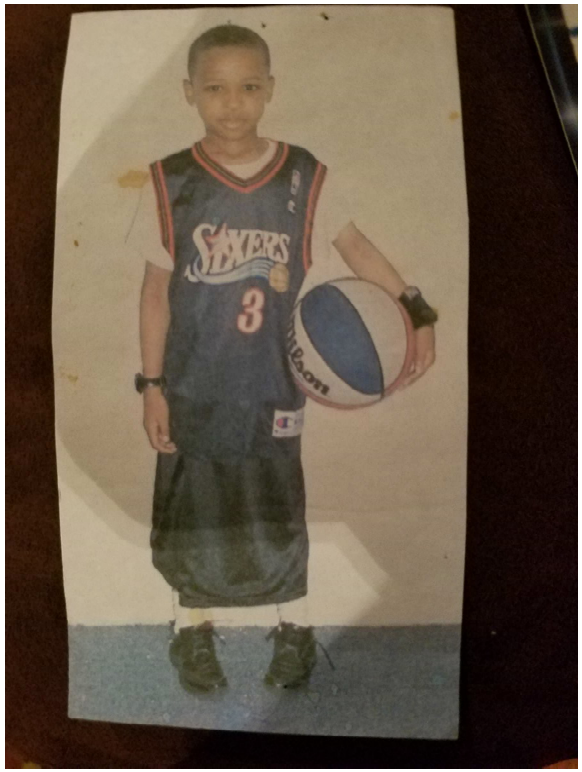
Treatment of Syncope

- Added Salt – Pretzels, healthy snacks, BACON
- Compressions socks, stockings
- Maneuvers prior to standing
- Venous Valve Training exercises
- Fludrocortisone, Mitodrine, Metoprolol
  - Class IIB
- Treatment of underlying Cardiac or Neurologic cause

Summary

- Recurrent syncope is common
- Patient history is essential for an accurate diagnosis
- Few diagnostic tests for “typical” syncope are required
- History that fails the smell test should be thoroughly investigated
- REMEMBER, You might not get it right the first time. Keep the doors open to your patient and primary care team!
- Appropriate therapy can be life-saving

Worrisome Syncope – Syncope and subsequent seizure playing basketball



8 yo with syncope

- Diagnosed with Anomalous left coronary artery from the opposite facing sinus
- Had surgical unroofing
- At age 11 years, ranked 36th in the USA in the 100 meters
- Attending college on full basketball scholarship
- Eternally grateful to PCP


AT&T LTE
1:29 PM
64%
Instagram
t.harp3
TROY 3 HARPER
35
STRAIGHT FT MADE
ALL-TIME DREXEL MBB RECORD

FLY EAGLES FLY

Many Thanks


The Children's Hospital of Philadelphia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