Autonomic investigation in children which test and when

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15% of children and adolescent experience at least 1 syncope

Reason for syncope in children

75% autonomic syncope

10% cardiac syncope

8-17% psychogenic or unexplained

Personal and family history

Physical examination

ECG

Exclude possible arrhythmic reason

Autonomic syncope

Types of autonomic mediated syncope

Breath-holding spells

Blood injury phobia

Inappropriate tachycardia

Postural tachycardia syndrome

Vaso-vagal syncope

Main autonomic reflex regulating circulatory

Type of reflex	Receptors	Localisation of receptors	Function	Effect
baroreceptors	mechanoreceptors	Aortic arch, pulmonary artery Coronary artery Zatoki szyjnej	↑ parasymaptetic ↓sympatetic	↓HR ↓BP
Bainbridge	mechanoreceptors	Vena cavie and pulmonary vein	↑sympatetic	Heart rate regulation during exercise
Bezold-Jarisch	Mechano and chemoreceptors	Left ventricle	↑parasympatetic ↓sympatetic	↓HR ↓BP
Arterial chemoreceptors	Chemoreceptors (O2, CO2, pH)	Aortic arch, right subclavian and carotid artery	↑sympatetic ↓parasympatetic	↑HR ↑BP Respiratory regulation
Central nuer chemoreceptors	Chemoreceptors C)2, pH	Medulla	↑↑↑ sympathetic ↓parasympatetic	↑HR ↑BP Respiratory regulation
Diving reflex	Thermoreceptors, chemoreceptors	Facial skin, rdzeń przedłużony	↑sympatetic ↓parasympatetic	↓HR ↑BP

Tests for autonomic investigation

1. Orthostatic (Schellong) and modify Schellong test
2. Head up Tilt test
3. Valsalva maneuver
4. Cold pressor test
5. Handgrip test
6. Thermoregulatory sweat test
7. Supine and upright epinefrine and norepinefirne level
8. Carotid massage
9. Diving reflex

2015 Heart Rhythm Society Expert Consensus Statement on the Diagnosis and Treatment of Postural Tachycardia Syndrome, Inappropriate Sinus Tachycardia, and Vasovagal Syncope

Heart Rhythm, Vol 12, No 6, June 2015

Recommendations—POTS and Vasovagal Syncope in the Young

	Class	Level
Pediatric patients presenting with suspected vasovagal syncope or POTS should undergo a detailed medical history review and physical examination and undergo a 12-lead ECG.	I	E
Pediatric patients with suspected POTS should undergo orthostatic testing.	I	E
Tilt-table testing is reasonable for highly selected pediatric patients with suspected vasovagal syncope.	IIa	C
It seems reasonable to treat selected pediatric patients with vasovagal syncope with midodrine.	IIb	B-R
It seems reasonable to treat pediatric patients with vasovagal syncope or POTS with interventions that are recommended for adults with these disorders.	IIb	E

Different mechanisms leading to OI

Mechanisms are not fully understand

Lack of systematic investigations

For majority of children with OI
Personal history is crucial for diagnosis
Non-pharmacological therapy causes symptoms disappear

1.
2. High sensitivity and specificity
3. High repeatability

Breath-holding spells

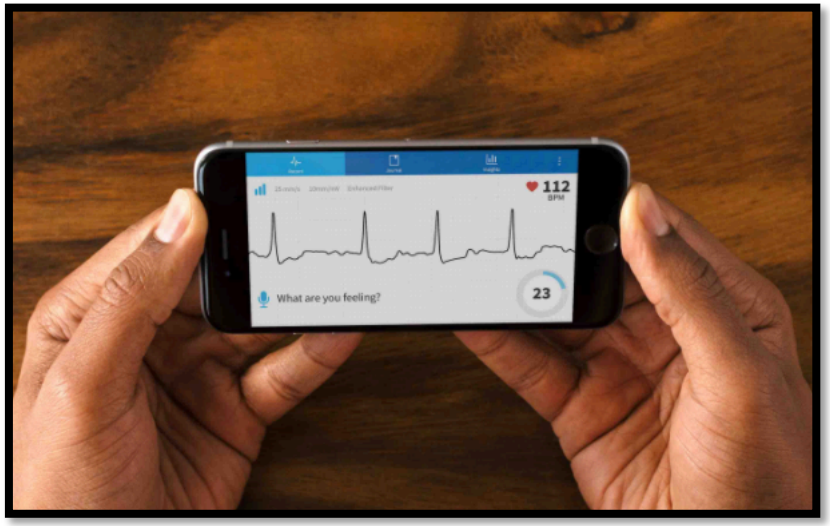
Usually occurs between 6 months of age and 5 years (peak 12-24 months)

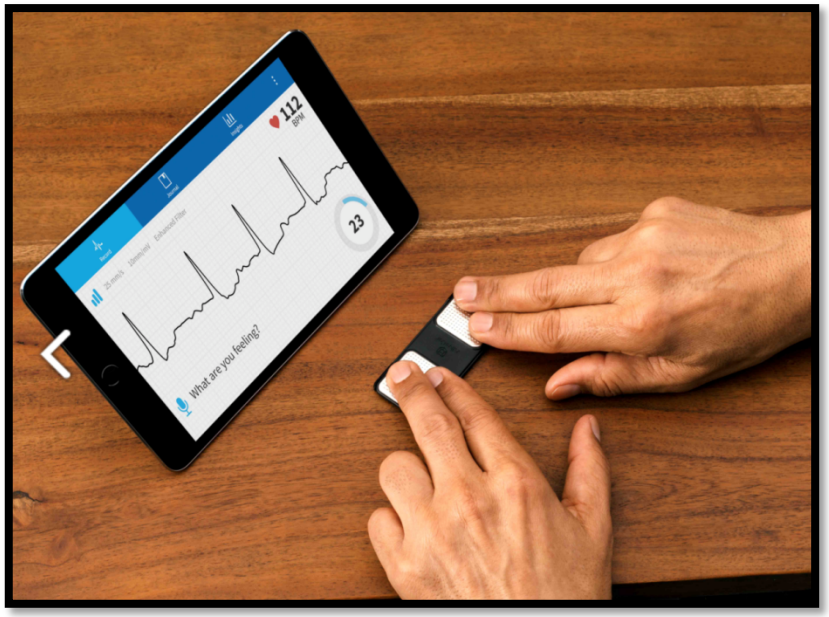
There is severe bradycardia and/or asystole

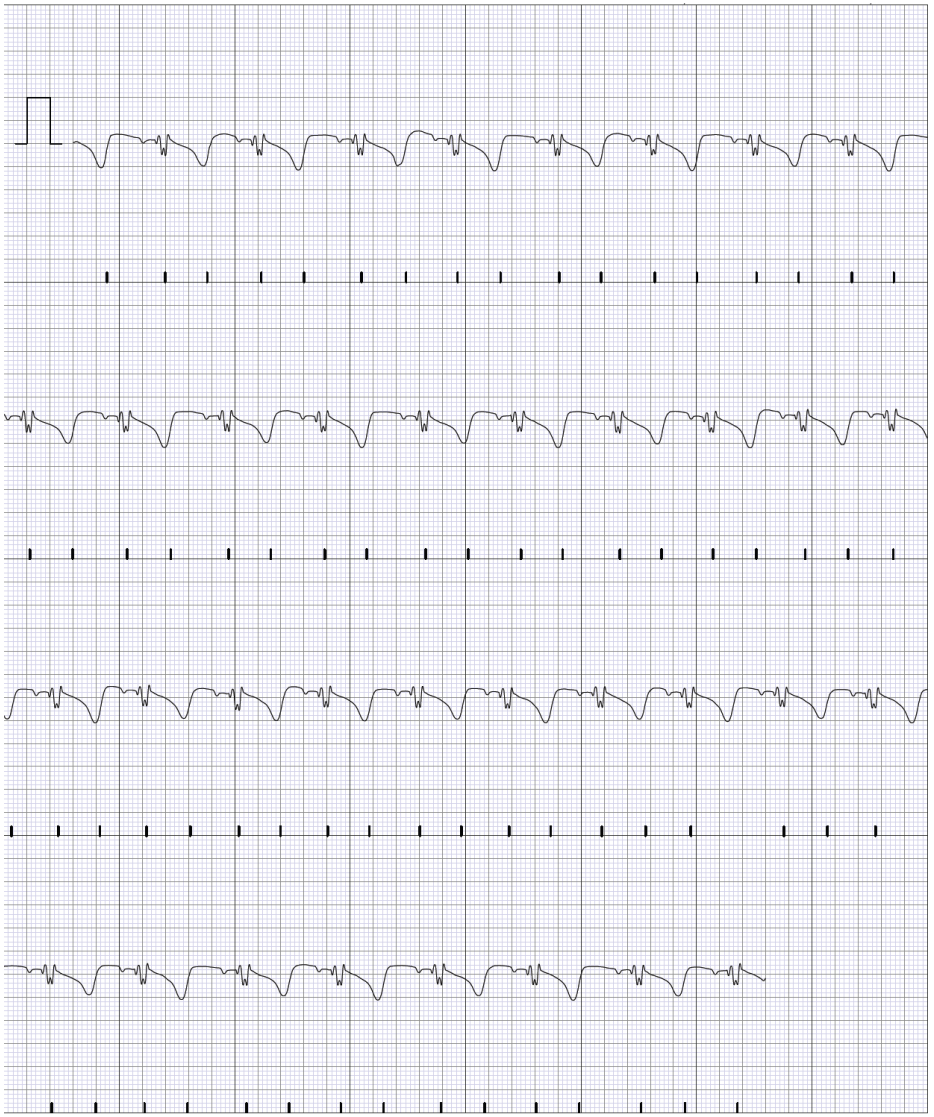
Infantile form of vaso-vagal syncope

Cyanotic-loud cry followed by apnea, may be associated with myoclonic movements

Pallid breath holding spells/reflex anoxic seizures-brought on by injury or pain. In contrast to cyanotic no crying



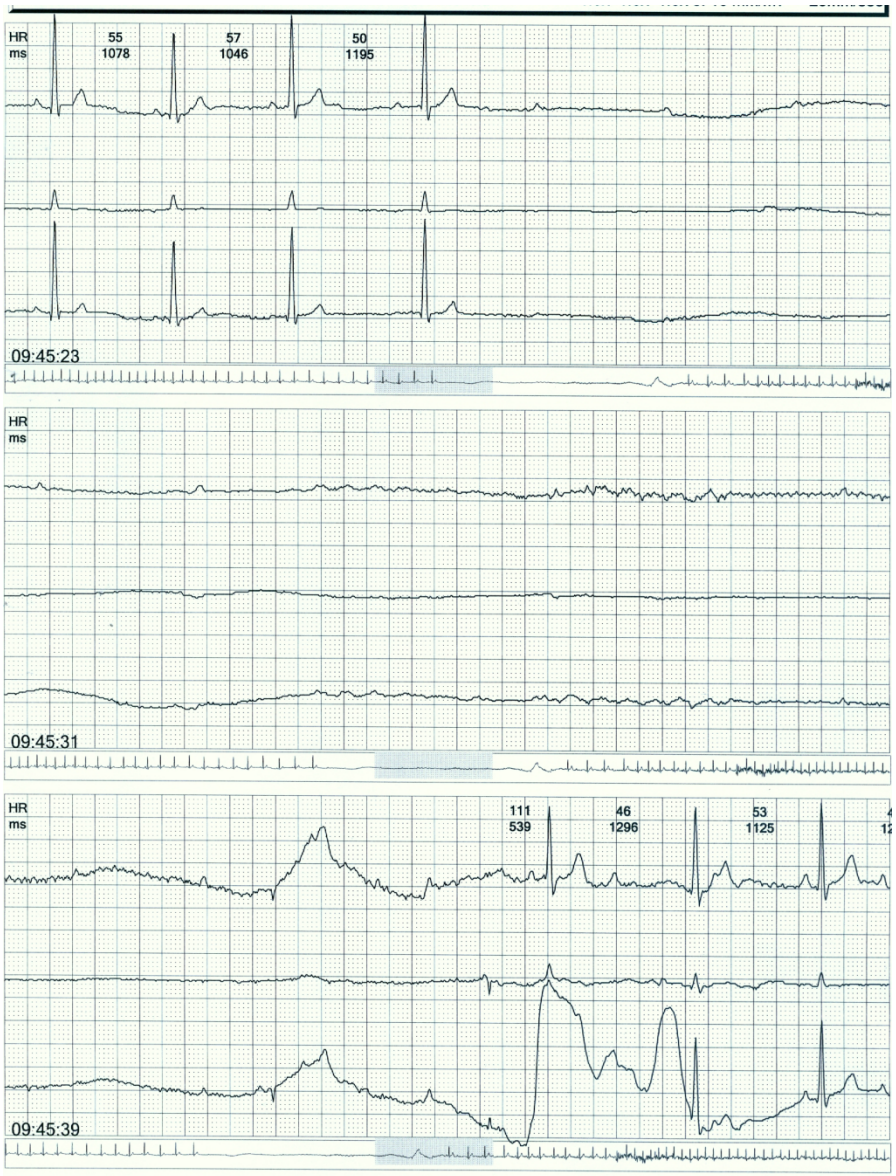




ECG from chest

10 month old boy with LQTS and Fallot

Blood injury phobia



13 years old girl

Inappropriate sinus tachycardia is define as sinus heart rate >100bpm (with 24-hrs monitoring >90 bpm-ADULTS!) and is associated with distressed symptoms

Inappropriate Sinus Tachycardia

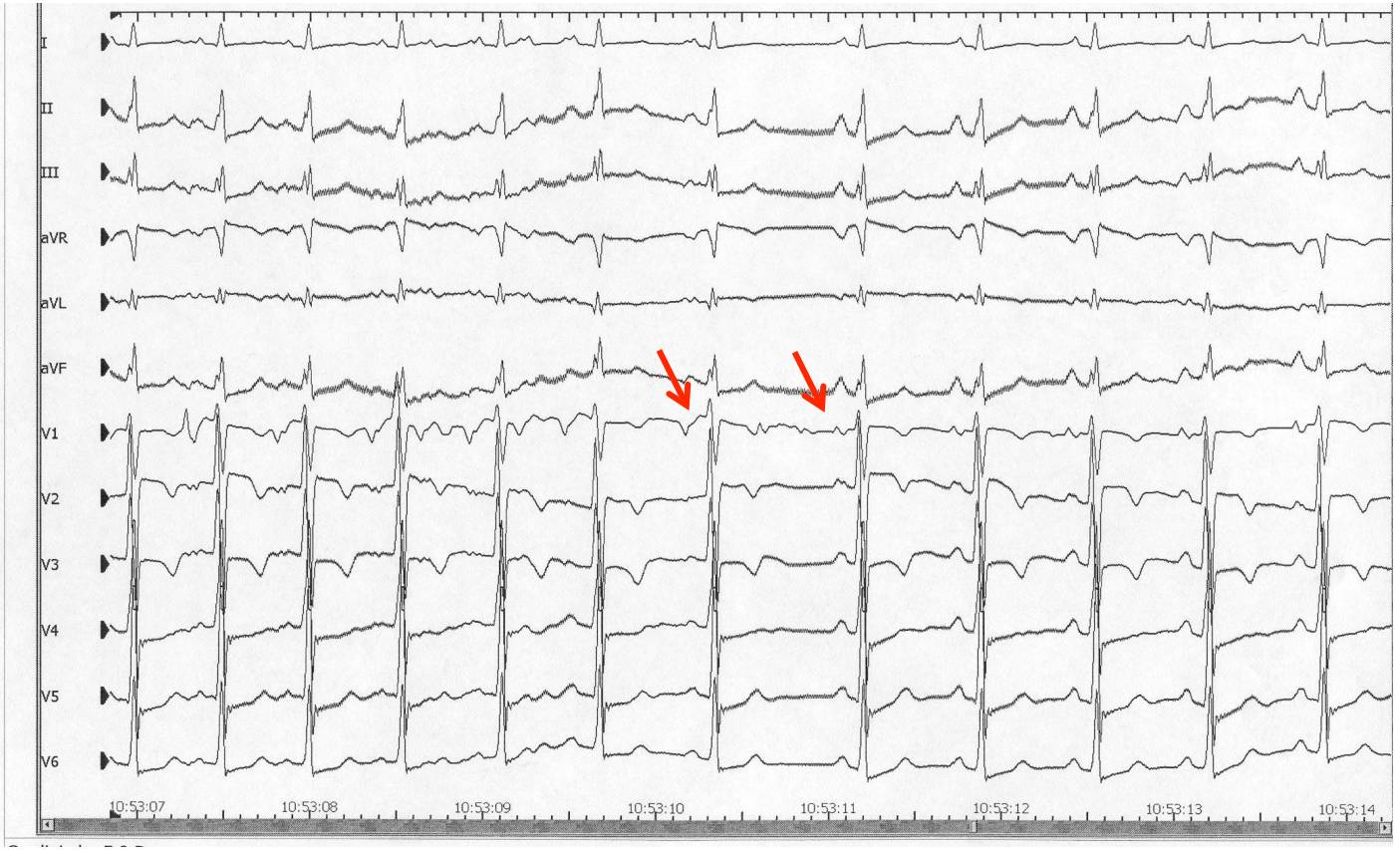
-increased sinus node automaticity

-increased beta-adrenergic activity

-decreased parasympatetic activity

Medical history and physical exam focus on
-possible cause of sinus tachycardia
-psychological triggers
-panic attacks

The standard 12 – leads ECG and 24-Holter monitoring
-proofed the diagnosis
-exclude FAT
Autonomic test: HR response to deep breathing, Valsava maneuvers, cold water test, HRV **have unproven usefulness**


I
II
III
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10:53:12
10:53:13
10:53:14

Postural tachycardia syndrome: frequent symptoms occur with standing: lightheadedness, palpitations, tremor, weakness, exercise intolerance and fatigue. Increased >40 beats /min when changing positions with absence of orthostatic hypotension

Postural Tachycardia Syndrome-physiology

-peripheral autonomic denervation

-hypovolemia

-hyperadrenergic POTS

-deconditioning

These mechanism often co-exist

POTS

Detailed history:

-symptoms chronicity

-possible causes

-potential triggers (diet and exercise)

-symptoms of autonomic neuropathy (feeling full, bloating, nausea, sweating)

-family history of sudden cardiac death <50 yr

-comorbidity: infection, diabetes, hypothyreosis, side effects from cancer treatment, autoimmun disease, Ehlers-Danlos

Orthostatic test

10-15 min rest in supine position

RR and HR measure 1,2, 3 and 10 min after upright position

Positive: HR increased >40bpm (POTS)

RR decreased within 3 min s≥20mmHg

d≥10mmHg (orthostatic hypotonia)

IF ORTHOSTATIC TEST IS NORMAL BUT CLINICAL SYMPTOMS MAKE POTS VERY POSSIBLE HEAD UP TILT TEST IS HELPFULL

VV syncope is a syncope syndrome that usually occurs with upright posture held for more than 30 sec or with exposure to emotional stress, pain, worm, nausea and pallor. It is associated with hypotension and relative bradycardia, and followed by fatigue

Vaso-vagal syncope- physiology

-periphelal venous pooling

-reflex from mechanoreceptors

Vaso-vagal syncope

Detailed history:

-predisposing situations (prolonged standing 2-3 min)

-prodromal symptoms (diaphoresis, warmth, flushing, nausea)

-physical signs

-recovery time (1-2 min, but patient could be tired for minutes to hours)

Head UpTilt Test

Heart Rhythm recomendation for evaluation of vv syncope

Recommendation	CoR	LoE
Tilt table testing can be useful for the investigation of patients with suspected vasovagal syncope who lack a confident diagnosis after initial assessment.	IIa	B-NR
Tilt table testing is reasonable for distinguishing convulsive syncope from epilepsy; to establish a diagnosis of pseudosyncope; and in patients with suspected vasovagal syncope but without clear diagnostic features.	IIa	B-NR
Tilt table testing is not recommended for predicting the response to specific medical treatments for vasovagal syncope.	III	B-R
Implantable loop recorders can be useful in the investigation of older patients with infrequently recurrent and troublesome syncope who lack a clear diagnosis and are at low risk of a fatal outcome.	IIa	B-R

Head Up Tilt Test

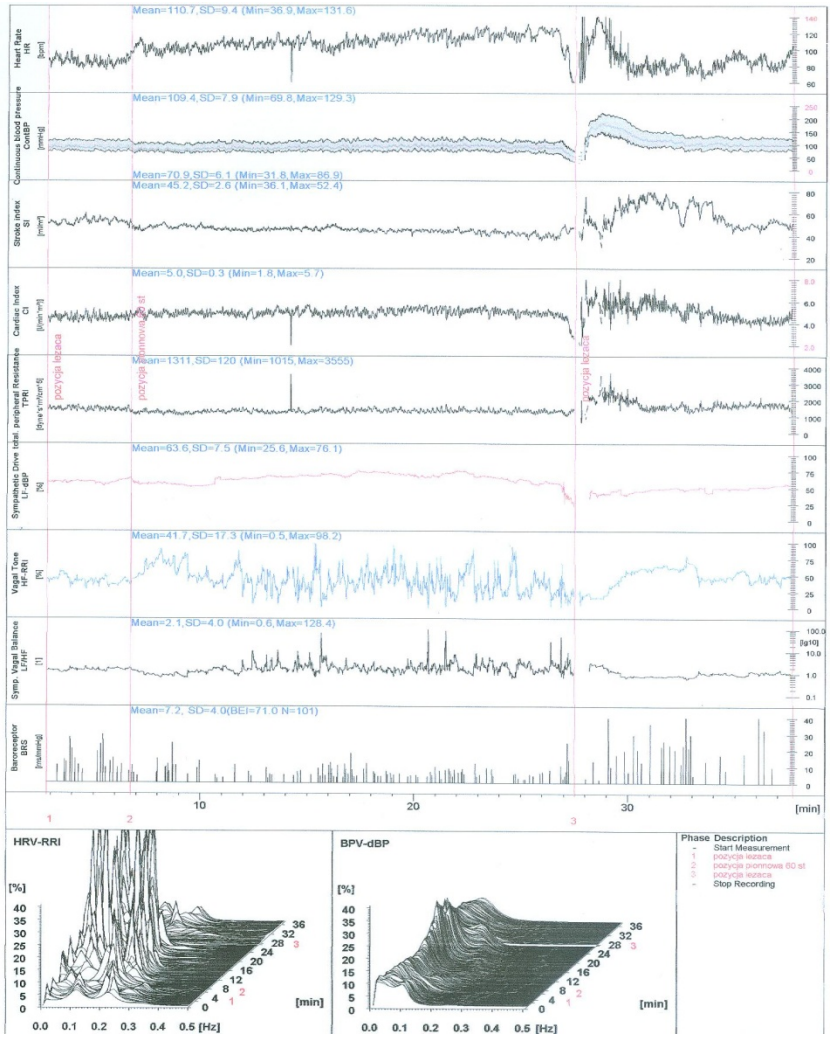
Used for children older than 6 yrs

HUTT has not been prospectivly validated

There is no ideal protocol

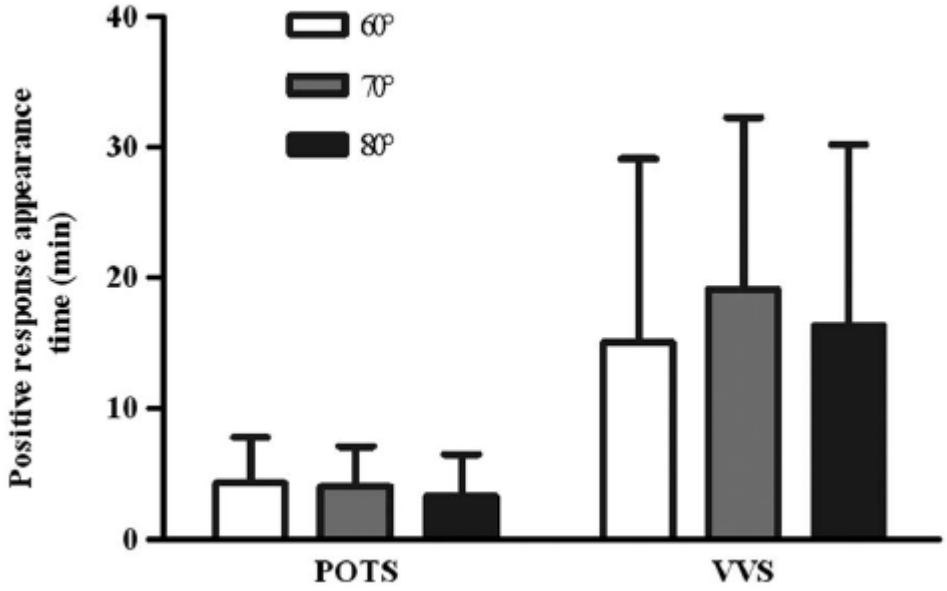
With increasing sensitivity there is decrease in specyfity

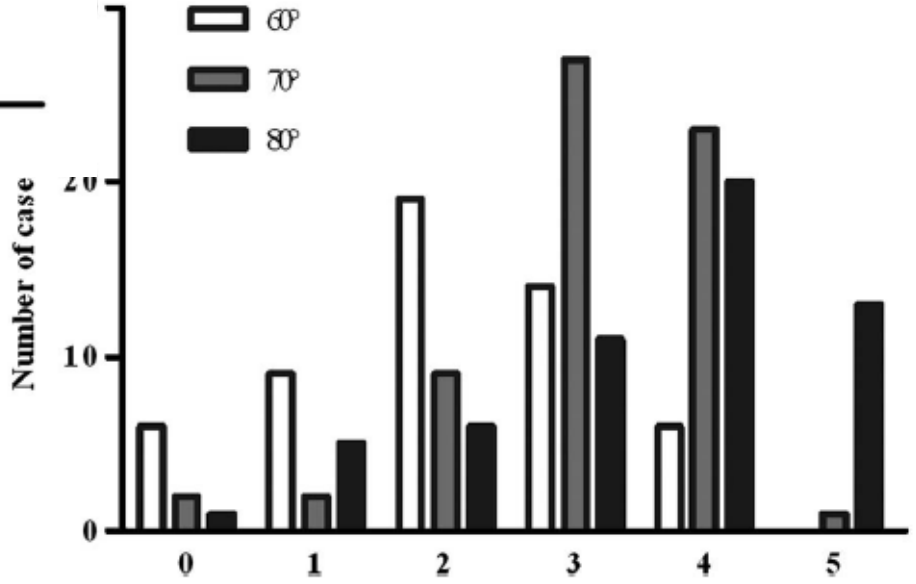
Most labs used 70 ° for 20 min



Tilt angles and positive response of head-up tilt test in children with orthostatic intolerance

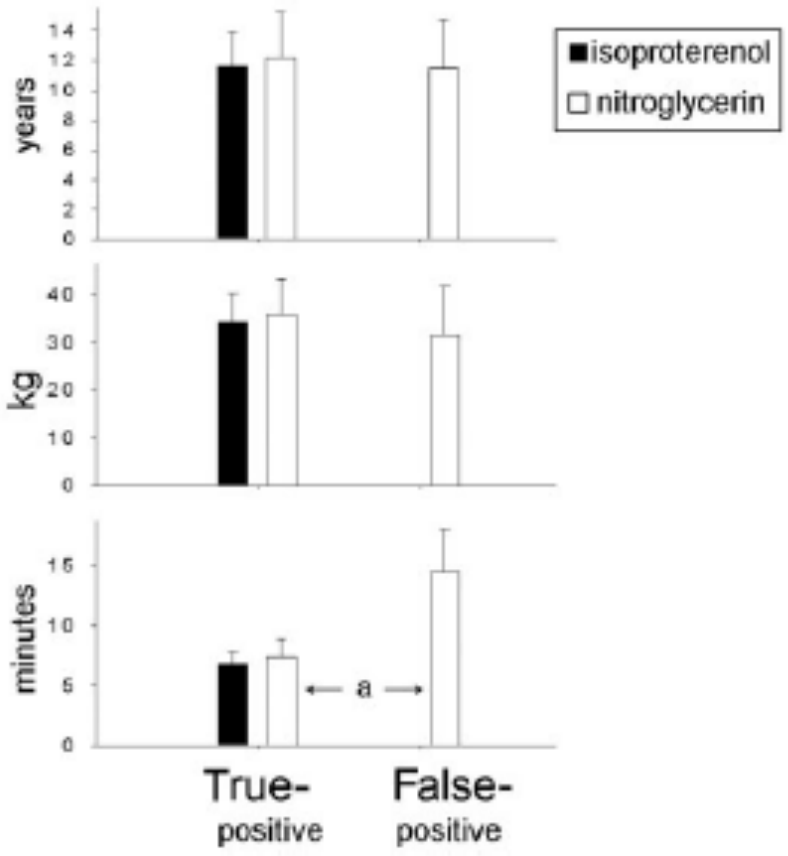
Jing Lin,[1] Yuli Wang,[1] Todd Ochs,[2] Chaoshu Tang,[3,4] Junbao Du,[1] Hongfang Jin,[1]





Provocation of Neurocardiogenic Syncope During Head-up Tilt Testing in Children: Comparison Between Isoproterenol and Nitroglycerin

Antonios P. Vlahos, MD[a], Meropi Tzoufi, MD[a], Christos S. Katsouras, MD[b], Theodora Barka, RN[b], Irene Slonti, MD[c], Lampros K. Michalis, MD[b], Antigoni Siamopoulou, MD[a], Theofilos M. Kolettis, MD[b]



Pediatrics 2007

POTS/VVS

When others autonomic tests should be done?

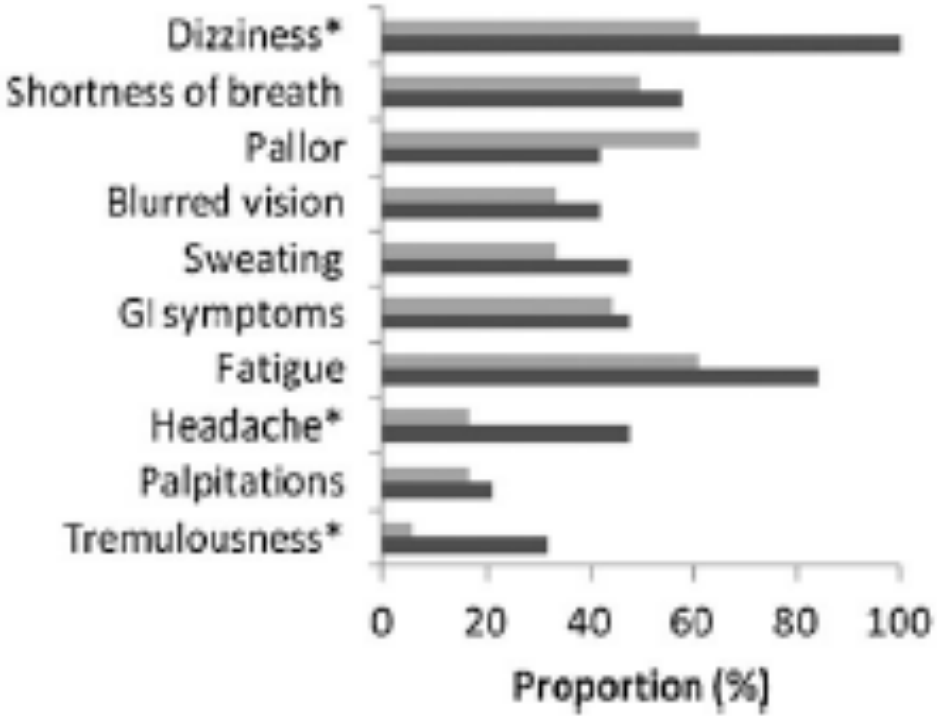
-diagnosis is uncertain

-symptoms persists

Clinical features of hyperadrenergic postural tachycardia syndrome in children

Qingyou Zhang,[1]* Xia Chen,[2]* Jiawei Li[1] and Junbao Du[1]

[1]*Department of Pediatrics, Peking University First Hospital, Beijing, and* [2]*Department of Pediatrics, Langfang Municipal People's Hospital, Langfang, Hebei, China*



Pediatrics International, 2014

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Items	Hyperadrenergic POTS ($n = 19$)	POTS alone ($n = 18$)	*P*
Age (years)	11.47 ± 2.06	10.61 ± 2.55	0.264
Sex ratio (M/F)	7/12	7/11	0.140
BMI, kg/m^2	19.39 ± 3.78	18.13 ± 3.72	0.316
Supine SBP (mmHg)	102.44 ± 9.05	97.89 ± 8.10	0.121
Supine DBP (mmHg)	57.28 ± 7.71	56.22 ± 8.29	0.695
Supine HR (b.p.m.)	73.78 ± 10.15	73.78 ± 10.55	1.000
Standing SBP (mmHg)	119.39 ± 8.66	105.50 ± 7.91	0.000
Standing DBP (mmHg)	66.67 ± 6.25	64.89 ± 5.45	0.370
Standing HR (b.p.m.)	127.47 ± 13.07	117.94 ± 13.19	0.034
ΔHR (b.p.m.)	54.05 ± 13.51	44.16 ± 12.54	0.027
Standing serum norepinephrine (pg/mL)	961.61 ± 343.07	420.81 ± 145.34	0.000

Pediatrics International, 2014

When we suspect hypovolemic…

Urinary sodium exertion

<124 mmol/24 hrs is suitable for salt supplement

B.G.

Syncope since 2 yr

Constipation

Defecation often with crying and syncope

Any manouvers in anal region – crying and syncope

Hypotherosis

B.G

Neurological Department

- ✓ **EEG syncope provocation by stimulation in anal region**
- ✓ **ECG channel- asystole 7 sek**
- ✓ **24 h Holter EKG – proof the asystole**

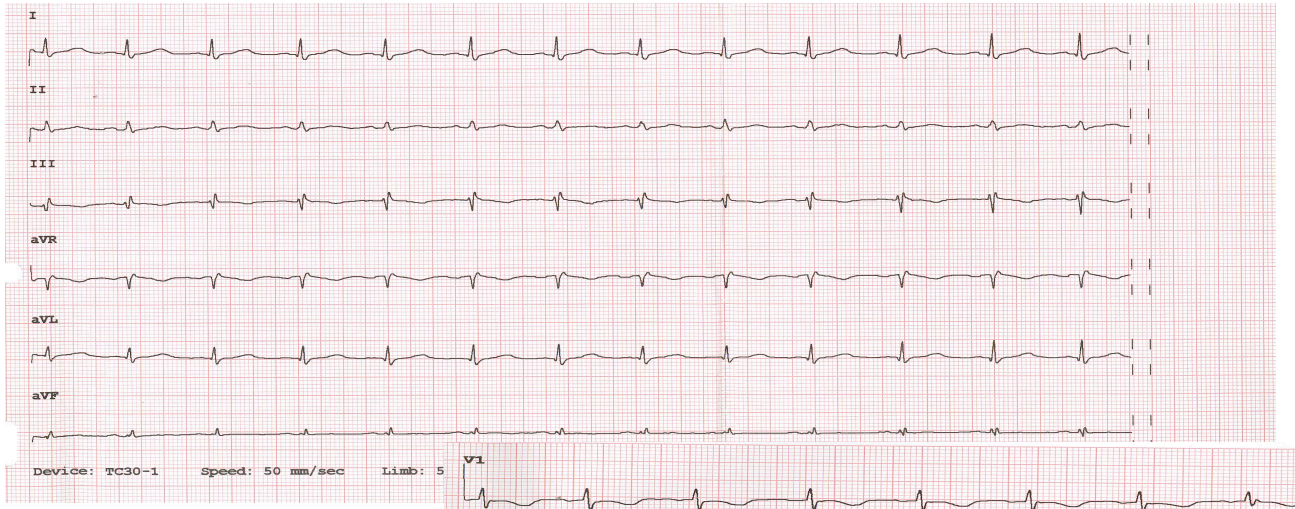
Cardiologic evaluation:

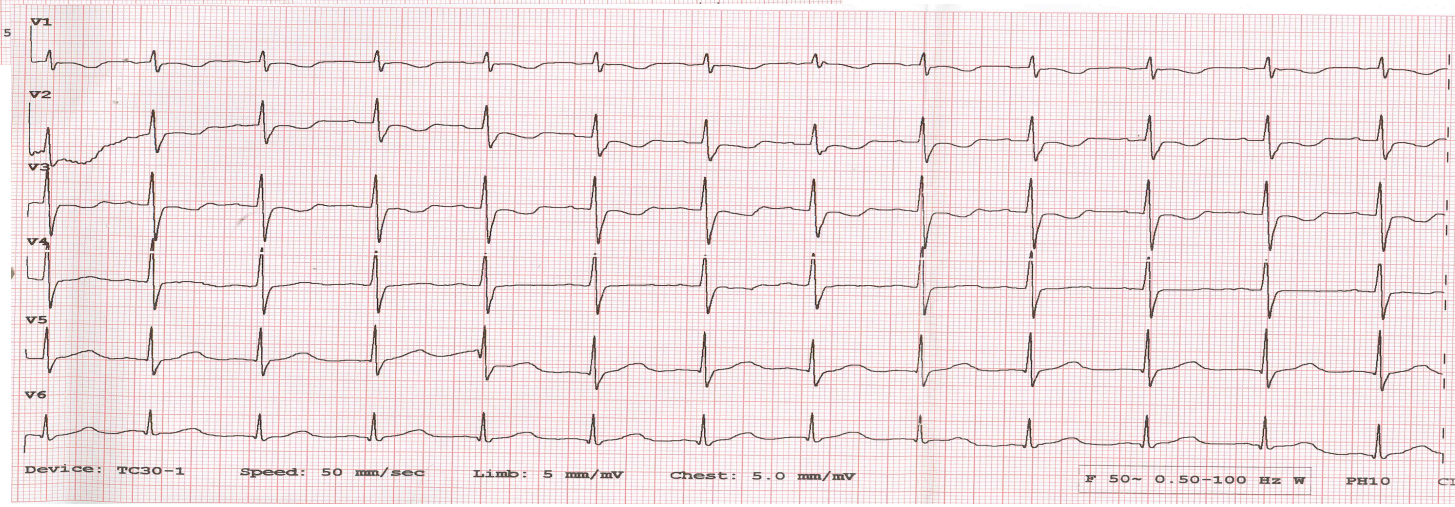
O/E normal heart sounds, no HF

B.G.

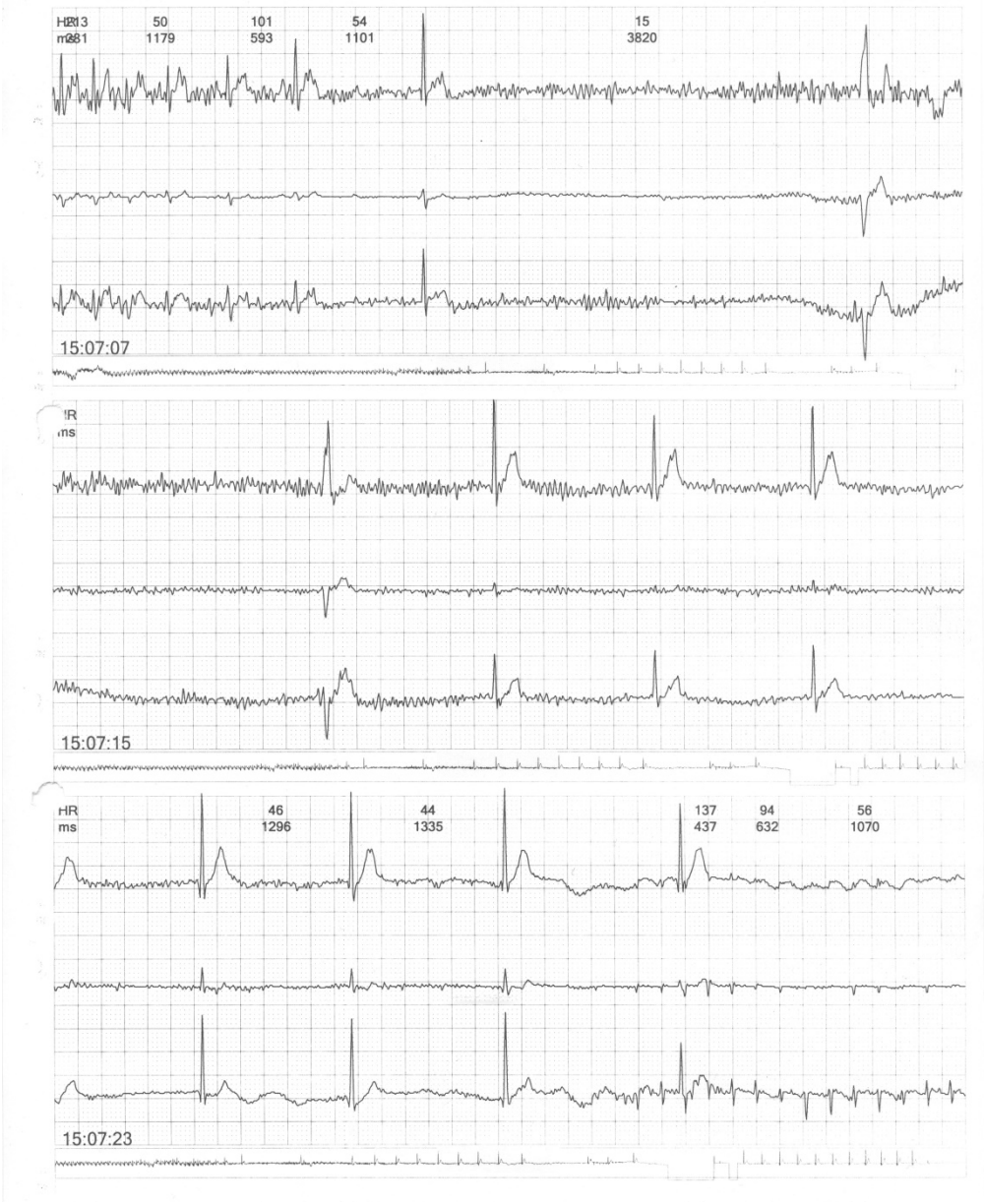
ECHO- normal

ECG





- B.G.
- Holter EKG



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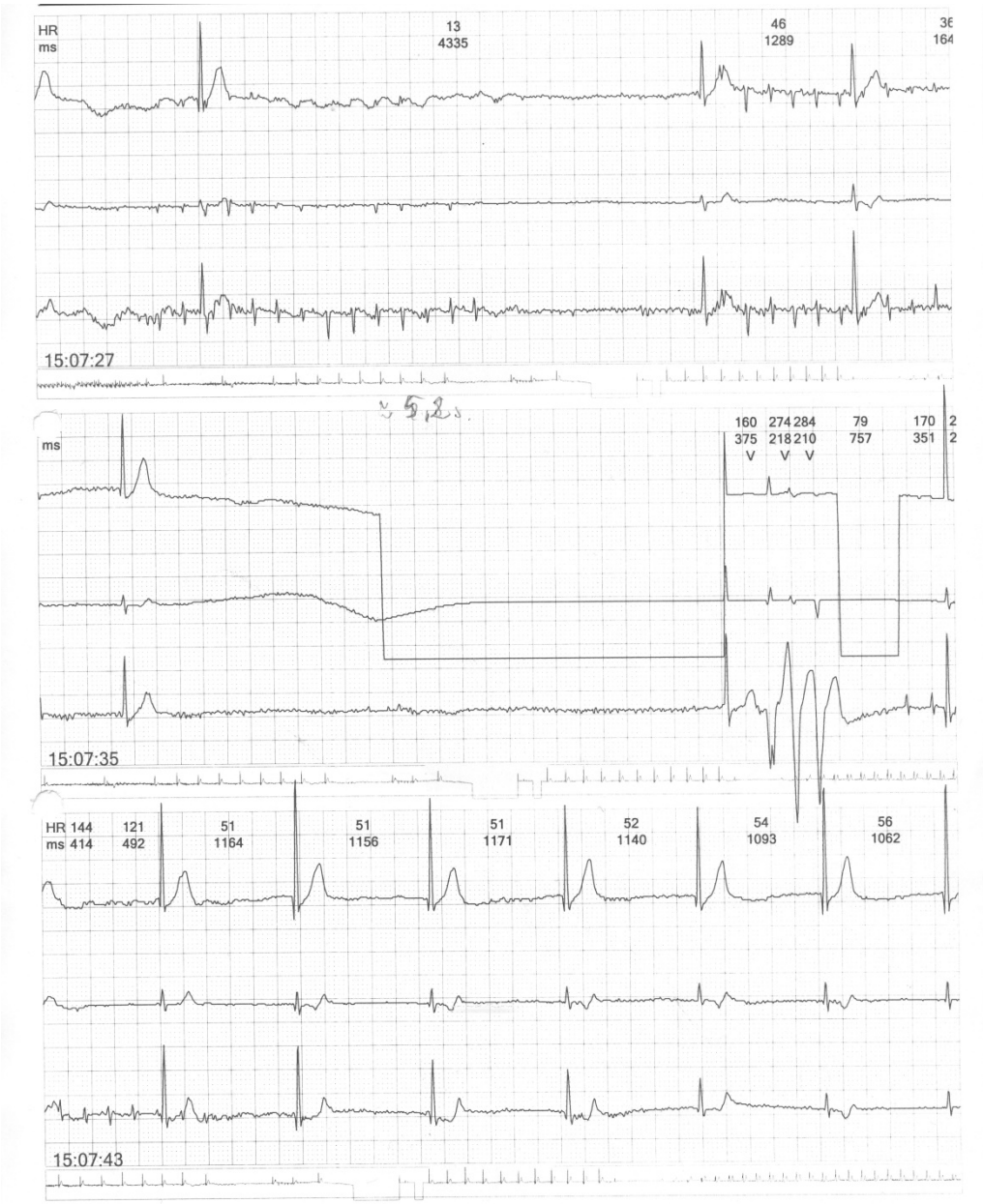

Crucial for diagnosis is detail personal history

There are only a few tests suitable for autonomic investigation

Regrading to specific symptoms other than orthostatic and HUTT test can be used