Arrhythmias in children

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Case Report

- Female, 17 years
- No previous illness
- Slight dizziness at rest
- Paroxysmal palpitations felt
Teenager with dizziness a. palpitations

Questions:
• Diagnosis?
• Responsible for symptoms?
• Treatment?
Overview of frequent and clinically relevant arrhythmias:

- Extrasystole
- Supraventricular tachycardia
- Ventricular tachycardia
Basics

Normal conduction

SA node
AV node
Bundle of His
Left bundle branch
Right bundle branch
Purkinje fibers

Normal ECG

Delay in A-V node
Depolarization of atria
Depolarization of ventricles
Repolarization of ventricles
Basics
12 standard leads

Einthoven
I,II,III

Goldberger
aVR, aVL, aVF

Wilson
V1-6
12-lead scalar electrocardiogram
Domaines of ECG
• Myocardial ischemia
• Arrhythmia
Rhythm and heart rate
• Rhythm regular or irregular?
• Heart rate for age: normal? too slow? too fast?

QRS duration and morphology
• QRS duration normal or prolonged for age?
• Morphology of QRS complex?

P wave morphology and relation to QRS complex
• P-waves visible?
• Normal morphology?
• How related to QRS complex?
Teenager with dizziness a. palpitations

- SR
- Monomorphic
- VES
- Triplets
- Fusion beat

**Diagnosis**
- VES
Extrasystole

- Supraventricular (SVES)
- Ventricular (VES)
Supraventricular Extrasystole
Supraventricular Extrasystole (SVES)

ECG – Criteria
- P-wave premature
- deformed
- short PQ-interval
- normal QRS complex
Origin of SVES

Sinus extrasystole

Atrial extrasystole

“upper” AV-Extrasystole

“middle” AV-Extrasystole

“lower” AV-Extrasystole
Extrasystole

- Supraventricular (SVES)
- Ventricular (VES)
Ventricular Extrasystole
ECG-Criteria of VES

- Premature prolonged QRS complex
- Mostly no P wave visible
- Origine from LV -> right bundle branch block
- Origin from RV -> left bundle branch block
VES - Definition

- Bigeminus
  - 1 N – 1 ES
- Trigeminus
  - 2 N – 1 ES
- Quadrigeminus
  - 3 N – 1 ES
- Couplet
- Triplet
- >3 = Ventricular Tachycardia (VT)!!
Exercise test in pat. with extrasystole

At rest
- HR 74/min

During exercise
- HR 144/ min
- No VES

-> Exercise test for risk stratification of ES
Conclusion
• Almost always benign
• Patients asymptomatic
• Disappear during exercise
• No treatment required

-> BUT
Attention if

• Polymorphic VES

• Increase of ES during exercise testing

-> Exclude underlying heart disease e.g. myocarditis (echocardiography)
Case Report

- Girl, 5 years
- gastrointestinal infection 4 weeks before
- present problems
  - dyspnoea
  - fatigue
  - irregular heart beats

**Diagnosis**
Dilative cardiomyopathy/myocarditis

**Echocardiography**
- Reduced LV function
- Dilated LV
- Mitral valve insufficiency
Case Report

• Infant, 6 weeks
• No previous illness
• Suddenly restless
• Poor feeding
• Swetting

→ Diagnosis: sepsis/ infection?

• Ad admission: heart rate 260/min
Tachycardia

- HR 260/min
- Small QRS complex
- No P–wave
Upper limit of normal resting heart rate, e.g.

- Infants 180 beats/min
- Adolescents 130 beats/min
Tachycardia

Definition
• >3 consecutive beats above upper normal range of age adjusted heart rate

Terms
• Paroxysmal: sudden begin and end of tachycardia
• Sustained: persistent tachycardia >30 sec
• Non-sustained: tachycardia <30 sec
• Incessant: tachycardia lasts for days, weeks
• Repetitive: change between SR a series of paroxysmal tachycardia
Tachycardia

- Supraventricular (frequent)
- Ventricular (rare)
Supraventricular tachycardia

Definition

• Origin above the bundle of His
• Normal duration of QRS-complex
  - duration in children <60 msec
  - duration in adults <80 msec
• 2 forms

Re-entry tachycardia
Via accessory pathway

Focal atrial tachycardia
## Relative Incidence – Age

<table>
<thead>
<tr>
<th>Author</th>
<th>Population</th>
<th>AV-Reentry</th>
<th>AVN-Reentry</th>
<th>Primary atrial</th>
</tr>
</thead>
<tbody>
<tr>
<td>Naheed et al.</td>
<td>Fetuses</td>
<td>73%</td>
<td>0%</td>
<td>27%</td>
</tr>
<tr>
<td>Ko et al.</td>
<td>Children</td>
<td>73%</td>
<td>13%</td>
<td>14%</td>
</tr>
<tr>
<td>Case et al</td>
<td>Children, 50% CHD</td>
<td>33%</td>
<td>24%</td>
<td>42%</td>
</tr>
<tr>
<td>Josephson, Wellens</td>
<td>Adults</td>
<td>34%</td>
<td>51%</td>
<td>15%</td>
</tr>
</tbody>
</table>
Atrioventricular Reentry Tachycardia

- Electrical impulse:
  AV node → ventricles → accessory pathway → atrium retrogradely excited

- Normal QRS complex

- Inverted P-waves following QRS complex
1. Narrow complex regular tachycardia at 180 beats/min (norm <130/min)
2. Retrograde P-wave following QRS complex
Preexcitation

- Antegrade accelerated AV-conduction through accessory pathway (Kent bundle) bypassing the AV-node (sometimes concealed conduction)

- **WPW syndrome**
  the classic form of preexcitation
Criteria of WPW-syndrome

- Short PR-interval due to an anomalous rapid AV-conduction pathway (adults <120 msec)
- Delta wave
- Wide QRS complex
Wolff-Parkinson-White Syndrome

- Presence of a short PR interval (<120 msec)
- A wide QRS complex >120 msec with a slurred onset of the QRS waveform producing a delta wave in the early part of QRS
Risk of WPW-syndrome

- Paroxysmal AV-reentry tachycardia
- Small QRS tachycardia = orthodromic conduction
Acute Therapy of SVT

Diagnosis
• 12 lead ECG
• 12 lead ECG recording during therapeutical intervention

Therapy, if patient haemodynamically stable
• Vagus manoeuvre, e.g. ice on face
• Rapid bolus of i.v. adenosine (0.1-0.3 mg/kg)
• i.v. propaphenone (1-2 mg/kg) slowly over 5-10 min

Guidelines of DGPK, 2011
Conversion of SVT with Adenosine

Adenosine
• Causes transient AV block
• Interrupts SVT, if AV node is part of the reentry circuit
Diagnosis of underlying tachycardia after bolus of Adenosine

- Transient AV block due to adenosine
- Unmasking atrial flutter
Acute Therapy of SVT

Equipment
• 12 lead ECG recording during therapeutical intervention
• Stable i.v. line
• ICU back-up

Therapy, if patient haemodynamically instable or unsuccessful conversion with drugs
• ECG triggered synchronised cardioversion
  0,5-1 J/kg
• If unsuccessful
  - increase to 2 J/kg
  - i.v. amiodarone 5mg/kg
    over 10-20 min

Guidelines of DGPK, 2011
Long term treatment for SVT

Repetitive paroxysmal SVT can result in heart failure due to tachycardia induced cardiomyopathy.

-> treatment is mandatory

RV
- dilated
- reduced function
- TV insufficiency
Long term treatment for SVT

Frequently used drugs
- Propaphenone (10mg/kg/d)
- Flecainide (3-7mg/kg/d)
- dl- Sotalol (2-6mg/kg/d)
- Propranolol (2mg/kg three times a day)
- Amiodarone (maintenance dosage 3-5mg/kg/d)

Ablation of accessory pathway
- Indicated for paroxysmal reentry SVT at age >5 years
Case Report

- Male adolescent, 14 years
- No previous illness
- Suddenly palpitations
- Presyncope
- Dyspnoea
- Dizziness
• Heart rate 142/min (norm <120/min)
• Broad QRS tachycardia
Tachycardia
  • Supraventricular
  • Ventricular
    (rare)
Supraventricular Tachycardia (SVT)
• Origin above His bundle
• Mostly:
  - Small QRS (<80 msec) complex tachycardia
  - Patient haemodynamically stable
  - No underlying heart disease
  - More frequent in children than in adults
  - Can be interrupted by bolus of i.v. adenosine

Ventricular Tachycardia (VT)
• Origin below His bundle
• Broad QRS (>80 msec) tachycardia
• Cannot be interrupted by bolus of i.v. adenosine
• Mostly:
  - Patient haemodynamically instable (emergency!)
  - Frequent underlying heart disease
Wide QRS complex tachycardia

**Diagnosis**
- 12 lead ECG recording
- Bolus of i.v. adenosine
  - if patient is stable
  - if VT, no response to adenosine
- Often underlying heart disease

**Acute therapy**
- Emergency, on ICU
- Synchronised cardioversion
  (0.5-1 J/kg, up 2 J/kg)
- Defibrillation if ventricular fibrillation
- If no success i.v. amiodarone
  (5mg/kg over 20 min)
Case report

- Male, 11 years
- Tetralogy of Fallot
- after surgical repair

- Acute problems:
  - palpitation
  - dizziness
  - breathlessness
Non sustained VT
Haemodynamic relevance
nonsustained ventricular tachycardia
Follow-up therapeutical options

• Drug treatment
  - beta blocker, e.g. metoprolol
  - amiodarone

• ICD implantation after cardiac resuscitation

• Ablation of focus in ventricle
Ventricular tachycardia

Special form: Genetic syndrome of arrhythmia = ion-channel disease

- Long QT syndrome
- Short QT syndrome
- Brugada syndrome
- Catecholamine sensitive polymorph VT (CPVT)

Typical signs
- ECG
  - polymorph VT (torsade de point), ventricular fibrillation
- Trigger
  - physical or emotional strain
- Symptoms
  - *syncope*, sudden death
• Male, 12 years
• **Syncope during exercise** at the age of 11y
• No previous illness, uneventful family history
• Present problem
  - **Syncope during exercise** causing fracture of the mandibular
• During anesthesia for surgery of mandibula fracture
  - ventricular tachycardia
  - ventricular fibrillation
Torsade de point VT
QTc: 585ms!
Long QT syndrome

Typical features

- Prolongation of QTc on ECG (best on Lead II and V5)
- Predisposition to life-threatening ventricular tachyarrhythmias
- Inherited disease, mainly autosomal dominant
- Prevalence 1:2,000-10,000 subjects
QTc

Represents the duration of activation and recovery of the ventricular myocardium

Bazett’s formula:

\[
QTc \text{ (sec)} = \frac{QT \text{ (sec)}}{\sqrt{R-R \text{ (sec)}}}
\]
<table>
<thead>
<tr>
<th>Rating</th>
<th>1-15y</th>
<th>Adult male</th>
<th>Adult female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&lt;440ms</td>
<td>&lt;430ms</td>
<td>&lt;450ms</td>
</tr>
<tr>
<td>Borderline</td>
<td>440-460ms</td>
<td>430-450ms</td>
<td>450-470ms</td>
</tr>
<tr>
<td>Prolonged</td>
<td>&gt;460ms</td>
<td>&gt;450ms</td>
<td>&gt;470ms</td>
</tr>
</tbody>
</table>

Goldenberg et al., J Cardiovasc Electrophysiol 2006
Genetically caused syndromes of arrhythmia

- Genetic testing recommended
- LQTS 1-3 account for 85% of all genetically detected LQTS

**TABLE I. Inherited Arrhythmic Disorders**

<table>
<thead>
<tr>
<th>Disease</th>
<th>Inheritance</th>
<th>Chromosome Locus</th>
<th>Gene</th>
<th>Ion Channel</th>
</tr>
</thead>
<tbody>
<tr>
<td>LQT3</td>
<td>AD</td>
<td>3p21–p23</td>
<td>SCN5A</td>
<td>Sodium</td>
</tr>
<tr>
<td>LQT4</td>
<td>AD</td>
<td>4q25–q27</td>
<td>ANKB</td>
<td>Ankyrin</td>
</tr>
<tr>
<td>LQT2</td>
<td>AD</td>
<td>7q35–q36</td>
<td>KCNE2</td>
<td>Potassium</td>
</tr>
<tr>
<td>LQT1</td>
<td>AD</td>
<td>11p15.5</td>
<td>KCNE1</td>
<td>Potassium</td>
</tr>
<tr>
<td>LQT5</td>
<td>AD</td>
<td>21q22.1–p22.2</td>
<td>KCNE1</td>
<td>Potassium</td>
</tr>
<tr>
<td>LQT6</td>
<td>AD</td>
<td>21q22.1–p22.2</td>
<td>KCNE2</td>
<td>Potassium</td>
</tr>
<tr>
<td>Brugada</td>
<td>AD</td>
<td>3p21–p23</td>
<td>SCN5A</td>
<td>Sodium</td>
</tr>
<tr>
<td>JLN1</td>
<td>AR</td>
<td>11p15.5</td>
<td>KCNE1</td>
<td>Potassium</td>
</tr>
<tr>
<td>JLN2</td>
<td>AR</td>
<td>21q22.1–p22.2</td>
<td>KCNE1</td>
<td>Potassium</td>
</tr>
</tbody>
</table>

AD = autosomal dominant; AR = autosomal recessive; JLN = Jervell and Lange-Nielsen syndrome


Nader et al, Tex Heart Inst J 2007
Risk stratification

*Figure 1.* Kaplan–Meier Estimates of Survival Free of Cardiac Events among the 580 Patients with the Long-QT Syndrome in the Risk-Stratification Analysis, According to the Genetic Locus of the Mutation. The difference among the groups was significant (P=0.007 by the log-rank test).

Therapy of long QT-syndrome

**Acute treatment**
- Cardioversion / defibrillation
- i.v Magnesium (25-50mg/kg over 30 min)

**Long term treatment**
- Oral Betablockers, e.g. Metoprolol
- Implantation of ICD, e.g. after first cardiac arrest
- Surgical left cervicothoracic sympathetic denervation
Case Report:
“To treat or not to treat….”
Case Report

- 9 year old female
- healthy
- asymptomatic
- routine ECG before adenotomy

ECG: Monomorphic VES, triplets
Holter:
- Heart rate: ~90/min
- Diagnosis?
Normal
- Impulse from sinus node faster than AV-node or ventricles

Accelerated ventricular rhythm
- Impulse from ventricle slightly faster than from sinus node

DD:
Ventricular tachycardia
# Clinical differentiation of AVR from VT in adults

<table>
<thead>
<tr>
<th>Feature</th>
<th>AVR</th>
<th>VT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discovery</td>
<td>Chance</td>
<td>Illness</td>
</tr>
<tr>
<td>Symptoms</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Hemodynamic effects</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Sinus isochronicity</td>
<td>&lt;10 (-20)%</td>
<td>&gt;10 (-20)%</td>
</tr>
<tr>
<td>Heart rate</td>
<td>&lt; 120bpm</td>
<td>&gt;120bpm</td>
</tr>
<tr>
<td>Exertion</td>
<td>Converts to sinus rate</td>
<td>Nonconversion</td>
</tr>
<tr>
<td>Length of run</td>
<td>Short bursts</td>
<td>Long runs</td>
</tr>
<tr>
<td>Drug treatment</td>
<td>Not effective</td>
<td>Effective</td>
</tr>
<tr>
<td>Bundle branch block</td>
<td>LBBB</td>
<td>LBBB or RBBB</td>
</tr>
</tbody>
</table>

AVR, accelerated ventricular rhythm; bpm, beats per minute; LBBB, left bundle branch block; RBBB, right bundle branch block; VT, ventricular tachycardia

Reynolds and Pickoff, Pediatr Cardiol 2001
1. 12 lead ECG recording for diagnosis of arrhythmia

2. Frequent arrhythmia
   • Extrasystole
     - SVES and VES mostly harmless
     - Disappear during exercise
     - Cave: ES more frequent during exercise and patient symptomatic
       -> consider underlying heart disease

3. Clinically relevant arrhythmia
   • Supraventricular tachycardia
     - Small QRS tachycardia
     - Frequently reentry tachycardia due to accessory pathway
     - Mostly patients haemodynamically stable
     - Mostly i.v. bolus of Adenosine can interrupt reentry tachycardia
     - Long-term treatment of paroxysmal SVT with antiarrhythmic drugs
       or ablation of accessory pathway
• **Ventricular tachycardia**
  - Broad QRS complex tachycardia
  - Patient haemodynamically instable
  - i.v. adenosine cannot interrupt tachycardia
  - Emergency treatment with cardioversion
  - Frequently underlying heart disease (e.g. after cardiac surgery)

• **Important form of ventricular tachycardia**
  - Genetically inherited ion-channel disease
  - Long QT syndrome with risk of polymorph VT/ VF
    syncope and sudden death

4. **Differential diagnosis of broad QRS “tachycardia”**
   • Accelerated ventricular rhythm
     - Harmless
     - No treatment
<table>
<thead>
<tr>
<th>ECG findings</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Electrocardiographic</strong></td>
<td></td>
</tr>
<tr>
<td>QTc (Bazett’s formula)</td>
<td></td>
</tr>
<tr>
<td>? 480ms</td>
<td>3</td>
</tr>
<tr>
<td>460-470ms</td>
<td>2</td>
</tr>
<tr>
<td>450ms (in males)</td>
<td>1</td>
</tr>
<tr>
<td>Torsades de pointes</td>
<td>2</td>
</tr>
<tr>
<td>T- wave alternans</td>
<td>1</td>
</tr>
<tr>
<td>Notched T- wave in 3 leads</td>
<td>1</td>
</tr>
<tr>
<td>Low heart rate for age</td>
<td>0,5</td>
</tr>
</tbody>
</table>

**Clinical history**

<table>
<thead>
<tr>
<th>Syncope</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>With stress</td>
<td>2</td>
</tr>
<tr>
<td>Without stress</td>
<td>1</td>
</tr>
<tr>
<td>Congenital deafness</td>
<td>0,5</td>
</tr>
</tbody>
</table>

**Family history**

<table>
<thead>
<tr>
<th>Family members with definite LQTS</th>
<th>1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unexplained SCD in immediate family members &lt; 30yrs old</td>
<td>0,5</td>
</tr>
</tbody>
</table>

1. Findings in the absence of medications or disorders known to affect these electrocardiographic findings.
2. Torsades de pointes and syncope are mutually exclusive.
3. Resting heart rate below the second percentile for age.
4. The same family member cannot be counted in both categories.

Schwartz et al., Circulation 1993
Scoring:

<1 point: low probability of LQTS
2 to 3 points: intermediate probability of LQTS
>4 points: high probability of LQTS
Stimuli for events

LQT1: exercise-related arrhythmic events (related to swimming may be specific) Moss et al, Am J Cardiol 1999; Batra et al, J Pediatr 2002

LQT2: events triggered by auditory stimuli, such as an alarm clock or telephone ringing. Moss et al, Am J Cardiol 1999

LQT1/ LQT2: acute arousal events (such as exercise, emotion or noise)

LQT3: highest risk of events when at rest or asleep Schwartz et al, Circulation 2001
Stimuli for events

Triggers for cardiac events according to 3 genotypes. Numbers in parentheses indicate number of triggers, not number of patients.

Schwartz et al; Circulation 2000
Risk stratification categories for LQTS patients based on published event rates; more specific risk subsets by age group are detailed in Table 3. Kaplan-Meier (KM) estimates are based on a series of 869 LQTS patients (52). CFR = cardiopulmonary resuscitation; TdP = torsades de pointes; other abbreviations as in Figure 2.
Beta-blockers

- first-line prophylactic therapy
- should be administered to all intermediate or high-risk affected individuals
- considered on an individual basis in low-risk patients (Goldenberg et al.; JACC 2008)
- associated with a significant reduction in the rate of cardiac events in LQT1 and LQT2
- no evident reduction in LQT3 mutations (Moss et al., Circulation 2000)

→ high rate of residual cardiac events under beta-blocker therapy (Priori et al., JAMA 2004)
Implantable cardioverter-defibrillator (ICD)

- indicated for secondary prevention in LQTS patients
- for primary prevention in high-risk patients who remain symptomatic despite beta-blocker therapy Goldenberg and Moss, JACC 2008
- highly effective in high-risk LQTS patients Zareba et al., J Cardiovasc Electrophysiol 2003
- should be considered in high-risk Jervell and Lange Nielsen patients (limited efficacy of beta-blocker therapy) Schwartz et al., Circulation 2006; Goldenberg et al., J Cardiovasc Electrophysiol 2006
- should be considered when there is a strong family history of SCD or when compliance or intolerance to drugs is a concern Epstein et al., JACC 2008
Surgical left cervicothoracic sympathetic denervation

- introduced for the treatment before beta-blockers became available Moss et al., N Engl J Med 1971
- considered in patients with recurrent syncope despite beta-blocker therapy and in patients who experience arrhythmia storms with an ICD Schwartz et al., Circulation 2004
- 46% ! remained asymptomatic Schwartz et al., Circulation 2004
Pacemaker
- Used in selected LQTS patients with sinusbradycardia
→ Long-term follow up studies indicate a high rate of SCD Dorostkar et al., Circulation 1999

Ablation Haissaguerre et al., Circulation 2003
→ Further experience is needed

Gene specific therapies
- LQTS 3: beta-blocker + mexitilene (sodium channel blocker)!
- potassium channel blockers, protein kinase inhibitor,…
→ experience limited
• Female, 8 years old
• Incidental finding pre-op: wide QRS complex rhythm
Diagnosis?
Accelerated ventricular rhythm

- relatively slow monomorphic, benign VT
- The rate tends to be just slightly above sinus rate; not exceeding 200 beats per minute
- generally diagnosed only in patients with a structurally and functionally normal heart
- It resolves spontaneously and should not be treated
How to diagnose Arrhythmia: Rhythm method

Regular rhythm
• Normal rate
  - regular sinus rhythm
• Rapide rate, e.g.
  - supraventricular tachycardia
  - ventricular tachycardia
• Slow rate, e.g.
  - complete AV block

Irregular rhythm
• Regular arrhythmia, e.g.
  - AV block (Wenkebach)
• Irregular arrhythmia, e.g.
  - atrial fibrillation
• Single or infrequent arrhythmia, e.g.
  - premature beats

Long pause
- e.g. Sinus arrest
How:
NODAL RAVT

ORTHODROMIC

Fast limb

Slow limb

ANTIDROMIC

Fast limb

Slow limb

RV

LV

R R R

P P P

R R R

P P P
Supraventricular Tachycardia: AV nodal reentrant tachycardia

"slow-fast" conduction

P-wave follows the QRS complex
ECG analysis: the RP interval

• Useful in distinguishing AV reentry from AV node reentry tachycardias

\[
\leq 70 \text{ msec} \quad \text{traditionally associated with AVNRT}
\]

• > 70 msec accessory pathway

• > PR interval PJRT or atypical AVNRT