Silent Cardiac Killers in the Young

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Sudden Cardiac Death

- **Adolescents**
  - Incidence: 10/100,000 per yr
  - Incidence ↑
  - Exercise/stress induced
  - Congenital/Familial

- **Adults**
  - Incidence: 1/1000 per yr
  - Incidence ↓
  - Often occurs at rest
  - Acquired/Familial
Electrocardiography in the Patient With Syncope

- Acute coronary syndromes
- Tachydysrhythmias (SVT, CPVT, PJRT)
- Bradydysrhythmias and AV blocks
- Abnormalities of the intervals
  - prolonged QT, short QT (!!!)
  - short PR, wide QRS ( = WPW)
- Hypertrophic cardiomyopathy
- Brugada syndrome
- Arrhythmogenic RV dysplasia
## Basics: Intervals

*Davignon 1980*

<table>
<thead>
<tr>
<th>Group</th>
<th>Age*</th>
<th>Male</th>
<th>Female</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0 to 24 hours</td>
<td>85</td>
<td>104</td>
<td>189</td>
</tr>
<tr>
<td>2</td>
<td>1 to 3 days</td>
<td>85</td>
<td>94</td>
<td>179</td>
</tr>
<tr>
<td>3</td>
<td>3 to 7 days</td>
<td>88</td>
<td>93</td>
<td>181</td>
</tr>
<tr>
<td>4</td>
<td>1 to 4 weeks</td>
<td>59</td>
<td>60</td>
<td>119</td>
</tr>
<tr>
<td>5</td>
<td>1 to 3 months</td>
<td>51</td>
<td>64</td>
<td>115</td>
</tr>
<tr>
<td>6</td>
<td>3 to 6 months</td>
<td>44</td>
<td>65</td>
<td>109</td>
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<tr>
<td>7</td>
<td>6 to 12 months</td>
<td>60</td>
<td>78</td>
<td>138</td>
</tr>
<tr>
<td>8</td>
<td>1 to 3 years</td>
<td>94</td>
<td>98</td>
<td>192</td>
</tr>
<tr>
<td>9</td>
<td>3 to 5 years</td>
<td>114</td>
<td>98</td>
<td>212</td>
</tr>
<tr>
<td>10</td>
<td>5 to 8 years</td>
<td>114</td>
<td>112</td>
<td>226</td>
</tr>
<tr>
<td>11</td>
<td>8 to 12 years</td>
<td>110</td>
<td>124</td>
<td>234</td>
</tr>
<tr>
<td>12</td>
<td>12 to 16 years</td>
<td>105</td>
<td>142</td>
<td>247</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td></td>
<td>1,009</td>
<td>1,132</td>
<td>2,141</td>
</tr>
</tbody>
</table>

* The term “to” specifies the upper limit of each age range in the sense of “less than” logic. For instance, age group 2 (1 to 3 days), includes children from 24 to less than 72 hours.
Basics: Intervals

Rijnbeek 2001

| Table 2: Lead-independent ECG measurements for boys (upper row) and girls (lower row): median (2nd percentile, 98th percentile) |
|---|---|---|---|---|---|---|---|---|---|---|---|
| Lead | 0-1 months | 1-3 months | 3-6 months | 6-12 months | 1-3 years | 3-5 years | 5-8 years | 8-12 years | 12-16 years |
| Heart rate (beats min⁻¹) | 160 (129,192) | 152 (126,187) | 134 (112,165) | 128 (106,194) | 119 (97,155) | 98 (73,123) | 88 (62,113) | 78 (55,101) | 71 (48,99) |
| P axis (°) | 56 (13,99) | 52 (10,73) | 49 (−5,70) | 49 (9,87) | 48 (−12,78) | 43 (−13,69) | 41 (−54,72) | 39 (−17,76) | 40 (−24,76) |
| P duration (ms) | 78 (64,85) | 79 (65,98) | 81 (64,103) | 80 (66,96) | 80 (63,113) | 87 (67,102) | 92 (73,108) | 98 (78,117) | 100 (82,118) |
| PR interval (ms) | 79 (69,106) | 78 (62,105) | 78 (63,106) | 80 (64,97) | 83 (62,104) | 84 (66,101) | 89 (71,107) | 94 (75,114) | 98 (78,122) |
| QRS axis (°) | 99 (77,120) | 98 (85,130) | 106 (87,134) | 114 (82,141) | 118 (86,151) | 121 (99,162) | 129 (99,160) | 134 (105,174) | 139 (107,178) |
| QRS duration (ms) | 97 (75,140) | 87 (57,138) | 66 (−6,107) | 68 (14,122) | 64 (−4,118) | 70 (7,112) | 70 (−10,112) | 70 (−21,114) | 65 (−9,112) |
| QTc interval (ms) | 101 (91,121) | 99 (78,133) | 106 (84,127) | 109 (88,133) | 113 (78,147) | 123 (99,153) | 124 (92,156) | 129 (103,163) | 135 (106,176) |
| *Corrected QT interval, according to Bazett’s formula: QTc = QT \* \sqrt{heart rate / 60} |

Bold values indicate that the 95% confidence intervals of the percentile estimates for boys and girls do not overlap.
Basics: T wave

- Limited value
- (+) DOL 5-7
- (-) until 8 yo/early adolescence
- (-) 10 yo-adult “juvenile T wave”
- (+) T wave V1 = RVH
Basics: LVH

- S V1 and R V6
  - S V1 > 98th for age*
  - R V6 > 98th for age*
  - Adult R wave progression in young kid*
  - (-) T inferolateral = “strain”*
  - Deep Q V5-6 (>5mm)
#1: 18 yo presents after a syncopal episode
Hypertrophic Cardiomyopathy

- Many other names
  - IHSS, ASH, HOCM

- Characteristic anatomic abnormalities
  - Hypertrophied, non-dilated LV (normal CXR)
  - Septal hypertrophy (asymmetric, concentric, apical)

- Familial incidence in 55% of cases

- Average age at diagnosis 30-40 yo.

- Mortality 1-3.5% per year
Clinical features

- Syncope, chest pain, palpitations, dyspnea, sudden death
- Often associated with exertion (not always!)
- Attributable to dysrhythmias or sudden reductions in cardiac output
- Systolic murmur at apex or LLSB
- Increases with valsalva, standing
- Decreases with trendelenburg, squatting
Hypertrophic Cardiomyopathy

- ECG abnormalities present in 85-93%
- Definitive diagnosis — doppler ECHO
  - Doppler helps assess severity of obstruction at rest and with provocative maneuvers
- Treatment
  - Beta blockers, calcium channel blockers
  - Amiodarone if ventricular dysrhythmias
Hypertrophic Cardiomyopathy

ECG abnormalities
- HLVV (“LVH”), LAE most common
- Tall R wave in V1 (mimics posterior MI)
- Deep narrow Q-waves in inferior, lateral leads
- Apical HCM: giant precordial T-wave inversion
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy
ALCAPA
ALCAPA
ALCAPA
Apical Hypertrophic Cardiomyopathy
Apical Hypertrophic Cardiomyopathy

“Giant” T-wave inversions
Take Home Point

High voltage plus narrow Q waves in inferior/ lateral leads

= "Abnormal"

Hypertrophic cardiomyopathy or ALCAPA
#2: 16 yo presents after a syncopal episode
#2: Brugada Syndrome
The Brugada Syndrome

- First described in 1992 by Pedro and Josep Brugada
- Associated with sudden cardiac death
- Typically healthy patients with structurally normal hearts
- Generally considered a hereditary disease
The Brugada Syndrome

- More common cause of sudden death than previously recognized
  - Responsible for up to 20% of sudden deaths in pts. without structural heart abnormalities
  - Responsible for 4-5% of all sudden deaths
    - Incidence varies in different populations
- Most common in young males
- Age at diagnosis: 2 days to 84 years
- First onset of symptoms (syncope, sudden death) ~ 40 yo.
The Brugada Syndrome

- Mortality ~ 10% per year if not treated with internal cardioverter-defibrillator (ICD)
  - Antidysrhythmics have no effect on prognosis
- Syndrome characterized by
  - ECG abnls. in leads V1-V3
  - Polymorphic or monomorphomorphic VT
  - Structurally normal heart
  - Familial occurrence in ~ half of patients
The Brugada Syndrome

ECG findings in V1-V3
- RBBB or IRBBB pattern
- ST-segment elevation — 2 types
  - “Coved-type” (most common)
  - “Saddle-type”
- Findings can vary and be transient
- Obtain “high precordial” leads
- Definitive diagnosis — EPS
Brugada and Brugada, *J Am Coll Cardiol*, 1992
Brugada Syndrome

Coved Type

Saddle Type
Brugada Syndrome
Brugada Syndrome
Brugada Syndrome
Brugada Syndrome
Brugada Syndrome
Uncomplicated RBBB
Brugada Syndrome
Brugada Syndrome
The Brugada Syndrome

NORMAL SINUS RHYTHM
INCOMPLETE RIGHT BUNDLE BRANCH BLOCK
ST ELEVATION CONSIDER ANTERIOR INJURY OR ACUTE INFARCT
ST ELEVATION CONSIDER ANTERIOR INJURY OR ACUTE INFARCT
PROLONGED QT INTERVAL OR T U FUSION, CONSIDER MYOCARDIAL DISEASE, ELECTROLYTE IMBALANCE, OR DRUG EFFECTS
* * * * * * * * * ACUTE MI * * * * * * *
ABNORMAL ECG
WHEN COMPARED WITH ECG OF 07-FEB-2007 16:21, (UNCONFIRMED)
FUSION COMPLEXES ARE NO LONGER PRESENT
ST LESS ELEVATED IN INFERIOR LEADS...

Referred by:  

Confirmed By:  

M.D.
Brugada Syndrome
40 yo man with syncope
Brugada Syndrome

Courtesy Chris Touzeau

<table>
<thead>
<tr>
<th>Name:</th>
<th>12-Lead 1</th>
<th>HR 86 bpm</th>
<th>*** ACUTE MI SUSPECTED ***</th>
<th>acute infarct</th>
</tr>
</thead>
<tbody>
<tr>
<td>ID:</td>
<td>110107000109</td>
<td>00:28:03</td>
<td>** Abnormal ECG <strong>Unconfirmed</strong></td>
<td></td>
</tr>
<tr>
<td>Patient ID:</td>
<td>01 Nov 07</td>
<td>PR 0.130s</td>
<td>Normal sinus rhythm</td>
<td></td>
</tr>
<tr>
<td>Incident:</td>
<td>QRS 0.102s</td>
<td>QT/QTc</td>
<td>Incomplete right bundle branch block</td>
<td></td>
</tr>
<tr>
<td>Age:</td>
<td>40</td>
<td>P-QRS-T Axes</td>
<td>ST elevation consider lateral injury or</td>
<td></td>
</tr>
<tr>
<td>Sex:</td>
<td>aVR</td>
<td>0.348s/0.416s</td>
<td></td>
<td></td>
</tr>
<tr>
<td>P-QRS-T Axes</td>
<td>47° 19° 26°</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

MONTGOMERY CO EMS 13811371-138 29G6MOM 1GN6G7J LP1233515687
P/N 805319
PRINTED
40 yo man with chest pain

Courtesy Chris Touzeau

<table>
<thead>
<tr>
<th>12-Lead ECG</th>
<th>HR 54 bpm</th>
<th>• Abnormal ECG<strong>Unconfirmed</strong></th>
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<tbody>
<tr>
<td>30 Sep 08</td>
<td>15:17:59</td>
<td>• Sinus bradycardia</td>
</tr>
<tr>
<td>PR 0.168s</td>
<td></td>
<td>• Right bundle branch block</td>
</tr>
<tr>
<td>QT/QTc</td>
<td></td>
<td>• Inferior infarct, age undetermined</td>
</tr>
<tr>
<td>Sex: 40</td>
<td></td>
<td></td>
</tr>
<tr>
<td>P-QRS-T Axes</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Note:** The ECG shows characteristics of sinus bradycardia, which may be an indication of underlying cardiac issues. Further diagnostic evaluation and consultation with a cardiologist are recommended.
### STEMI!

*Courtesy Chris Touzeau*

<table>
<thead>
<tr>
<th>Name:</th>
<th>12-Lead 3</th>
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<tbody>
<tr>
<td>Ent ID:</td>
<td>893008145883</td>
</tr>
<tr>
<td>Dent:</td>
<td>30 Sep 08</td>
</tr>
<tr>
<td>PR</td>
<td>0.168s</td>
</tr>
<tr>
<td>HR</td>
<td>54 bpm</td>
</tr>
<tr>
<td>QRS</td>
<td>0.124s</td>
</tr>
<tr>
<td>QT/QTc</td>
<td>0.446s/0.422s</td>
</tr>
<tr>
<td>Sex:</td>
<td>P-QRS-T Axes</td>
</tr>
<tr>
<td>aVR</td>
<td>55° -6° 75°</td>
</tr>
</tbody>
</table>

- Abnormal ECG **Unconfirmed**
- Sinus bradycardia
- Right bundle branch block
- Inferior infarct, age undetermined

---

0.05-40Hz 25mm/sec
Brugada (3 yo)
Brugada (10 yo)
(In)complete right bundle branch block with ST elevation = Brugada syndrome
#3: 16 yo presents after a syncopal episode
Arrhythmogenic Right Ventricular Dysplasia (ARVD)
Arrhythmogenic Right Ventricular Dysplasia (ARVD)

- 2nd most common cause SCD
- Up to 20% SCD in patients < 35 yo
- Characterized by (fibro)fatty infiltration and thinning of the RV
- Non-ischemic cardiomyopathy
- Higher incidence in Italian and Greek descent
ARVD

Clinical features

- Syncope, chest pain, palpitations, dyspnea, diaphoresis, sudden death
- Often associated with exertion (not always!)
ARVD

Diagnosis
- Study of choice: Cardiovascular MRI
  ECG/Signal-averaged ECG
  Echocardiogram, RV angiogram
  Genetic testing, biopsy, autopsy
- Treatment: implantable cardiac defibrillator
ARVD

90% ECG abnormalities
- Epsilon wave (most specific)
- T-wave inversions V1-V3
  - Juvenile TWI typically resolves in puberty
- Prolonged S-wave upstroke V1-V3
- Localized widening QRS V1-V3
- Paroxysmal VT w/LBBB morphology
Epsilon Wave

Distinct deflection clearly separated from previous QRS

“BLIP”
Prolonged S-wave
ARVD
Teenager ARVD/C
Presents with palpitations (AVRD VT)
ARVD

Nasir et al, *Circulation*, 2004
ARVD
ARVD

Jain et al, Circulation, 2009
ARVD

ARVD

Jain et al, Circulation, 2009
Take Home Point

Epsilon wave or Precordial TWI in post-pubertal individual = Arrhythmogenic right ventricular dysplasia
Take Home Points

ECG is the most important screening tool in any youth w/ syncope
ECG does not rule in nor out structural heart disease
When in doubt recommend refraining from exercise
Summary

HCM
- High voltage + narrow Q in lateral leads

Brugada
- (In)complete RBBB with ST elevation

ARVD
- Epsilon wave
- Post-pubertal with precordial TWI
Special Thanks

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- Dr. Peter Gaskin
- Dr. Jane Crosso
- Dr. Hugh Calkin
Questions?

For a PDF copy of all slides, go to:

lectures.umem.org/AAEM
[will be posted after AAEM for 1 month]

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