Neonatal and infantile Ventricular Arrhythmia

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Seoul National University Children’s Hospital

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Neonatal and infantile ventricular arrhythmia

- PVC
- Accelerated ventricular rhythm

- Ventricular tachyarrhythmia
  - Idiopathic VT
  - Long QT syndrome
  - Myocarditis / Cardiomyopathies
  - VT d/t cardiac tumor
Premature ventricular complexes

• Common finding in the neonatal period: 18% of neonates by 24 hr holter

• Clinical evaluation to exclude the presence of structural or functional heart disease and a careful measurement of the corrected QT interval.

• Isolated monomorphic PVCs in the absence of associated heart disease:
  ➔ favorable prognosis: Disappeared during follow-up in all neonates at a mean age 2.1 months. no treatment

• Immaturity of cardiac conduction tissue and the autonomic nerve system may be the main causes of frequent premature beats

Pediatr Cardiol 1987;8(2):103–8
Arch Dis Child Fetal Neonatal Ed 2006;91:F419–F422
Progress in Pediatric Cardiology 11 2000. 39]45
Korean J Pediatr 2017;60(11):344-352
Accelerated ventricular rhythm

• The rate is not more than 10-20 beats/min faster than the normal sinus rate

• Generally considered to be a benign arrhythmia without clinical Sx or hemodynamic consequences

• Frequently resolved without any treatment

Ventricular tachycardia

- Wide QRS tachycardia, >120 bpm and ≥ 3 consecutive PVCs
- Rare in newborn and infant.
- Does not have to be particularly ‘wide’ (mean 0.08 sec)
- VT needs to be differentiated from other SVT/AF with BBB and SVT with anterograde conduction across an accessory pathway.
- If the tachycardia QRS is wide in neonate/infant then the diagnosis of VT is more likely.

- Presentation: incidental finding ~ cardiogenic shock

Causes of VT in newborn and infant

- Idiopathic VT (m/c)
- Hereditary channelopathy, including LQTS
- Myocarditis, cardiomyopathy
- Cardiac tumor
- Structural heart disease
- Electrolyte imbalance
Idiopathic VT

• No etiology of the arrhythmia can be identified in normal structural heart

• In general, when VT is idiopathic and asymptomatic, a good long-term prognosis can be anticipated.

• Symptomatic infants: higher HR (226 vs 196 bpm)

• VT onset in newborn without heart disease: Disappeared at a mean age 1.7 -2 months without complications

Arch Dis Child Fetal Neonatal Ed 2006;91:F419–F422
European multicenter study, 98 children from 12 centers

- 27%: infancy onset VT (17 newborn)
- 22% presented with symptoms
- Better Prognosis when VT occurred during the 1st year of life (VT resolution in 89% of infancy onset VT vs. 56% of beyond)

- RBBB 4/27 (14%), LBBB 23/27 (85%) in infancy VT
- No mortality

Table 2. Comparison of Clinical Characteristics of VT Depending on the Age at Initial Manifestation

<table>
<thead>
<tr>
<th></th>
<th>Infants</th>
<th>Children</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>27</td>
<td>71</td>
<td></td>
</tr>
<tr>
<td>Symptoms initially</td>
<td>6 (22%)</td>
<td>30 (38%)</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Sustained VT</td>
<td>12 (44%)</td>
<td>24 (34%)</td>
<td>NS</td>
</tr>
<tr>
<td>RBBB</td>
<td>4 (14%)</td>
<td>23 (32%)</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>VT resolution</td>
<td>24 (89%)</td>
<td>40 (56%)</td>
<td>&lt; 0.01</td>
</tr>
<tr>
<td>Never treated</td>
<td>9 (33%)</td>
<td>16 (23%)</td>
<td>NS</td>
</tr>
<tr>
<td>Still treated</td>
<td>2 (7%)</td>
<td>21 (30%)</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Follow-up</td>
<td>48 months</td>
<td>46 months</td>
<td>NS</td>
</tr>
</tbody>
</table>

RBBB = right bundle branch block morphology.
Ventricular tachycardia in infants with structurally normal heart: a benign disorder

1Division of Cardiology, The Children’s Hospital of Philadelphia, Pennsylvania
2Division of Cardiology, Department of Pediatrics, St. Louis Children’s Hospital, Washington University School of Medicine, St. Louis, Missouri

- 31 infants
- median onset: 1 day, range 1-255 days)
- Median duration of treatment: 13 months (3-105 months)
- VT resolved spontaneously in all( med 59 days)
- 20 LBBB, 4 RBBB in 24 infants
- No infant had sx of cardiovascular compromise
- Good ventricular function

Cardiol Young. 2010 December ; 20(6): 641–647.
10 patients < 12 months among 81 pts

Idiopathic VT (RBBB): 5 patients

Idiopathic VT with RBBB had 2 peaks – infancy and adolescence

Onset at infancy and monomorphic type were related to the successful resolution of VT.
Treatment

• DC cardioversion for hemodynamically unstable patients
• IV lidocaine
• beta-blocker, flecainide, mexiletine
• Amiodarone, or sotalol in absence of LQTS
Cause of VT in newborn and infant

- Idiopathic VT
- Hereditary channelopathy, including LQTS
- Myocarditis, cardiomyopathy
- Cardiac tumor
- Structural heart disease
- Electrolyte imbalance
Long QT syndrome

- Inherited channelopathy, Leading cause of sudden cardiac death.

- Presentation during neonate and infancy: higher risk of development of life threatening arrhythmia, higher mortality than older age group

- Prolonged QTc had a high prevalence of sudden infant death syndrome
  - 9% of SIDS victims had genetic mutation in cardiac ion channel, esp. in sodium current related genes.

Circulation. 2007 Jan 23;115(3):361
Clinical Characteristics and Genetic Background of Congenital Long-QT Syndrome Diagnosed in Fetal, Neonatal, and Infantile Life
A Nationwide Questionnaire Survey in Japan

Hitoshi Horigome, MD; Masami Nagashima, MD; Naokata Sumitomo, MD; Masao Yoshinaga, MD;

*Circ Arrhythm Electrophysiol. 2010;3:10-17

- 58 cases were registered from 33 institutions.
- Diagnosis of LQTS: fetal life (n=18), the neonatal period (n=31) → 84%
- Clinical presentation: sinus brady (n=37), VT/TdP (n=27), AV block(n=23), family hx (n=21), SCD/cardiac arrest (14), convulsion (5), syncope (5)
- Genotype confirmed in 29/41(71%): LQT1 (11), LQT2 (11), LQT3(6), LQT 8 (1)
- Tx: BB, mexiletine, pacemaker/ICD
- Death 7

<table>
<thead>
<tr>
<th>Parameter</th>
<th>LQT1 (n=11)</th>
<th>LQT2 (n=11)</th>
<th>LQT3 (n=6)</th>
<th>Negative (n=12)</th>
<th>Global Test</th>
<th>Pairwise Comparison</th>
</tr>
</thead>
<tbody>
<tr>
<td>HR, bpm</td>
<td>109±12</td>
<td>95±34</td>
<td>100±31</td>
<td>104±32</td>
<td>NS</td>
<td></td>
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<tr>
<td>(n=10*)</td>
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<tr>
<td>QTc, ms</td>
<td>560±24</td>
<td>538±74</td>
<td>592±79</td>
<td>575±86</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>(n=10*)</td>
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Proportion with family history, % 82 27 50 17 P<0.05
Proportion with sinus bradycardia, % 73 82 83 75 NS
Proportion with VT/TdP, % 0 91 100 42 P<0.05
Proportion with AVB, % 9 55 83 25 P<0.05

LQT1–Negative, P<0.05
LQT1–LQT2, P<0.001
LQT1–LQT3, P<0.005
(LQT1–LQT3, P=0.068)
Cardiac Channelopathies Associated with Infantile Fatal Ventricular Arrhythmias: From the Cradle to the Bench

KOICHI KATO, M.D.,* TAKERU MAKIYAMA, M.D., Ph.D.,† JIE WU, Ph.D.,*,†

- 7 cases had survived torsades de pointes (TdP) or ventricular fibrillation at ages < 1 year. Genetic test and functional assay to clarify genotype-phenotype correlation.

 Fatal ventricular arrhythmias in the early period of life is associated with cardiac channelopathies.

Functional assays revealed both gain and loss of function in SCN5A mutations, and loss of function associated with the KCNH2 mutation.

• Fetal sono: r/o PVC, VT
• 41 weeks, NFSD with 3.3kg
• VT and TdP on 1st day of life
• O/S hospital: IV lidocaine and amiodarone, defibrillation

→ transfer to SNUCH
Case

Amidarone d/c, lidocaine and propranolol

KCNH2 Novel sequence variant: c.1862G>T, p.S621I, heterozygote

QTc 533ms

→ FU with Propranolol for 8 yrs
Cause of VT in newborn and infant

- Idiopathic VT
- Hereditary channelopathy, including LQTS
- Myocarditis, cardiomyopathy
- Cardiac tumor
- Structural heart disease
- Electrolyte imbalance
Myocarditis

• Affected infants are seriously ill, with poorly tolerated incessant arrhythmias, congestive heart failure, and even cardiac arrest.
• usually have polymorphic incessant VTs.
• Myocardial function generally is severely depressed.
• extremely poor prognosis.
4 Months/male, poor oral intake, URI Sx
HR 60bpm, hypotension
EKG: cAVB, cardiac enz. elevation
Temporary PM insertion
Amiodarone, lidocaine, esmolol
DC cardioversion #11
→ Intubation and full sedation
VT controlled
HD#14 IV drug d/c
HD#20 discharge with oral BB
Myocardial tumors

- Incessant VT (occurring >10% of the day)

**Figure 2.** Same case. Operative photograph of left ventricular myocardioma. The thumb is above and index finger below. The arrow marks the gray-white area of "tumor."

**Figure 3.** Same case. Photomicrograph of myocardioma: Normal myocardium is present on the top right of the photograph. The large, pale cells of the hamartoma are found at the left and bottom. Original magnification × 900, reduced by 50%.

- 21 infants with incessant VT
- Mean 10.5 months
- EPS localized VT to the LV in 17 and to the RV septum in 4
- Medical tx failed to eliminate VT in all
- All had surgery at an age of 3 to 30 months
- Myocardial harmatomas in 13, Rhabdo in 2, myocarditis in 1 and myocardial fibrosis in 4, normal in 1
- Three died
- Histiocytoid cardiomyopathy

*Incessant Ventricular Tachycardia in Infants: Myocardial Hamartomas and Surgical Cure*

ARTHUR GARSON, JR., MD, FACC,* RICHARD T. SMITH, JR., MD, FACC,* JEFFREY P. MOAK, MD, FACC,* DEBRA L. KEARNEY, MD,† EDITH P. HAWKINS, K L. TITUS, MD, PhD,† DENTON A. COOLEY, MD, FACC,‡ DAVID A. OTT, MD on, Texas

*Progress in Pediatric Cardiology 2000:11;39-45*
*J Am Colll Cardiol 1987;10:619-26*
Conclusion

• VT is not common in neonate and infant but may be associated with significant morbidity and mortality

• Treatment and prognosis depend on the VT mechanism and pattern, the hemodynamic impact, and associated conditions.

• Idiopathic VT, monomorphic, normal QTc, narrow QRS during SR → Good prognosis

• Fatal arrhythmia such as Polymorphic VT and VF in infancy is associated with cardiac channelopathies
  → careful follow-up and management for the high risk of SCD
Thank you for your attention!