Catherine,

The attached announcement for the upcoming EMS Advisory Board Executive Committee is

Children with Broken Hearts

💖 Neonates with CCHD
💖 Sudden Cardiac Death

William B. Moskowitz, MD, FAAP, FAAC, FSCAI, FAHA
Professor and Chair, Pediatric Cardiology
Vice Chair, Clinical Operations,
Department of Pediatrics

Notebook.
Thanks! Irene
More infants are being diagnosed with critical congenital heart defects that require stabilization and transportation to regional centers for care.

As older children and adolescents with and without CHD participate in competitive athletics at many venues, sudden unexpected cardiac death remains a tragic and sometimes unpreventable event.
Objectives

• Therefore:
  – immediate care
  – appropriate care
  – stabilization
  – rapid transportation

• are essential to optimize outcome.
United States
4,000,000
Births Per Year

40,000 Births
All Congenital
Heart Disease

10,000 Births
Severe Congenital
Heart Disease

The Incidence of Congenital Heart Disease
Julien I. E. Hoffman, MD, FACC,* Samuel Kaplan, MD, FACC†

JACC Vol. 39, No. 12, 2002
June 19, 2002:1890–900
CONGENITAL HEART DISEASE

DEFINITION

Congenital Heart Disease is an anatomic malformation of the heart and/or its vessels which occurs during intrauterine development.
CONGENITAL HEART DISEASE

PREVALENCE

- 1% of live births (8-12/1000) if Family Hx negative
- 40-50% of Trisomy 21; 1 in 20 if mother has CHD
- 1 in 39 if mother diabetic
- Increased risk if father has CHD
CONGENITAL HEART DISEASE

FETAL CIRCULATION
There are some aspects of the fetal circulation that makes it quite different from the neonatal or adult circulation.

- The placenta is the major route of gas exchange, excretion, and acquisition of essential fetal chemicals.
- The placenta provides a LOW resistance circuit.
Two important events take place immediately following birth.

* The infant takes his/her first breath of air.

* The placenta is separated from the infant.
The cardiovascular system of the infant changes in the following ways.

- Pulmonary vascular resistance decreases.
- Pulmonary blood flow increases.
- Systemic vascular resistance increases.
- Blood flow through the ductus becomes primarily left to right.
- The foramen ovale functionally closes.
CONGENITAL HEART DISEASE

Children with Broken Hearts
CONGENITAL HEART DISEASE

These programmed changes take place regardless of whether the heart is normal or anatomically abnormal.
Ductal Dependency

I. Pulmonary Blood Flow
II. Systemic Blood Flow
III. Adequate Mixing in TGA
Ductal Dependency

With severe obstruction to pulmonary blood flow, maintenance of a PDA is essential for adequate PBF.

- Severe tetralogy of Fallot
- Critical valvar PS
- Pulmonary atresia
- Tricuspid atresia
- Severe Ebsteins disease of TV
- Complex single ventricle with PS
Children with Broken Hearts

Ductal Dependency

PA- IVS
Children with Broken Hearts

Ductal Dependency

TETRALOGY OF FALLOT
**Heart Disease**

**Ductal Dependency**

**With severe obstruction to systemic blood flow, maintenance of a PDA is essential for adequate SBF or cardiac output.**

- Critical valvar AS
- Hypoplastic Left Heart Syndrome
- Critical Coarctation of the Aorta
- Interrupted Aortic Arch
- Complex Single Ventricle with subAS
COARCTATION OF THE AORTA
HYPOPLASTIC LEFT HEART
Ductal Dependency

With obstruction to free mixing across the atrial septum in TGA, maintenance of a PDA is essential to improve atrial mixing and arterial saturation.
Some children with critical congenital heart disease will have no symptoms and have an entirely normal physical examination at the time they are sent home from the hospital after birth.

These children may become critically ill or die in a few days as their PDA constricts and closes if their congenital heart disease is not recognized.
The Law

- Infants born in Virginia shall be screened for critical congenital heart defects in accordance with the provisions set forth in §32.1-65.1 and 67 of the Code of Virginia and as governed by 12VAC 5-71-200 et seq.

- The Virginia Newborn Screening System consists of three components: (i) Virginia Newborn Screening Services (ii) Virginia Early Hearing Detection and Intervention Program, and (iii) Virginia Critical Congenital Heart Defect Screening Program.

- Currently, most hospitals in Virginia are screening!
Pulse Oximetry Screening Is:

- An assessment of oxygen level to check for cyanosis in newborns before they leave the hospital.
- Low blood oxygen levels may indicate the presence of congenital heart defects or other serious health conditions.
Ductal Dependency

Prostaglandin E

I. “Cyanotic” CHD where PBF is dependent on PDA.

II. “Acyanotic” CHD where SBF is dependent on PDA.

III. Transposition of the great arteries to improve mixing and arterial saturation.
Children with Broken Hearts

PGE-1

- Recommended starting dose **0.05-0.1 mcg/kg/min**.
- The dose may be titrated to maintain an open ductus with use of clinical signs of adequate perfusion, in addition to arterial blood pH and PO2, blood pressure, pulse oximetry, urine output, and echocardiography.
- In most infants, the ductus will reopen within 30 minutes to 2 hours after starting PGE-1.
- Once the ductus is open, the dose can be reduced to **0.002-0.05 mcg/kg/min**.
Many of the adverse effects of PGE1 are dose-related. Apnea, flushing, fever, bradycardia, and/or hypotension may indicate the need for dose reduction.

- Apnea occurs in approximately 12% of neonates receiving PGE1; not typically observed with doses < 0.01 mcg/kg/min. **Apnea is most likely to occur early, often within the first hour, and in infants weighing less than 2 kg.**

- Hyperthermia occurs in 10-14% of patients treated with PGE1. Cutaneous vasodilation (resulting in flushing and edema) occurs in approximately 10% of infants, and hypotension in up to 4%.
Adverse Effects of PGE-1

• When possible, the PGE1 dose be reduced to the lowest effective rate prior to transport to avoid having to initiate artificial ventilation during transport.

• Even with the increased safety associated with lower doses, **all hospitals considering the use of PGE1 should be capable of providing adequate ventilatory support in the event that apnea does occur, including intubation.**

• In cases where a reduction in dose is not tolerated, elective intubation prior to transport should be considered.
**Background:** Setting goals for monitoring and initiating life-saving interventions such as PGE1 during transport stabilization are dependent on establishing an accurate clinical diagnosis. **Objective:** The aim was to determine the accuracy of clinical diagnosis of suspected congenital heart disease (CHD) and the decision to initiate PGE1 in neonates presenting with hypoxemia. **Methods:** A retrospective cohort study (2002-2004) on hypoxemic neonates who were transported to an outborn NICU.

- Provisional diagnosis established by the transport team was categorized as suspected CHD (group 1), suspected persistent pulmonary hypertension of the newborn (group 2), and suspected CHD and/or persistent pulmonary hypertension of the newborn (group 3) based on history, physical examination, laboratory test, chest radiograph, and initial response to treatment.
- A definitive diagnosis was established on arrival to NICU by echocardiography.
# Clinical guidelines used by the transport team to establish a diagnosis of suspected CHD or PPHN

<table>
<thead>
<tr>
<th>Finding</th>
<th>Suspected CHD</th>
<th>Suspected PPHN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Associated clinical risk factors</td>
<td>Dysmorphic features</td>
<td>Fetal distress, meconium stained liquor, low Apgar score, cord pH &lt; 7.1</td>
</tr>
<tr>
<td>Cardiovascular examination</td>
<td>Murmur common</td>
<td>Murmur occasionally</td>
</tr>
<tr>
<td></td>
<td>Upper and lower limb systolic blood pressure gradient of ≥ 10 mm Hg</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Weak pulses</td>
<td></td>
</tr>
<tr>
<td>Oxygen saturation</td>
<td>Persistent low Spo₂ in the absence of significant respiratory distress</td>
<td>Lability (desaturation of &gt;10% on handling)</td>
</tr>
<tr>
<td></td>
<td>Reversed preductal vs postductal Spo₂ gradient</td>
<td>Preductal vs postductal Spo₂ gradient &gt; 10%</td>
</tr>
<tr>
<td>Hypoxia test</td>
<td>PaO₂ &lt; 50 mm Hg</td>
<td>PaO₂ &lt; 100 mm Hg</td>
</tr>
<tr>
<td>Oxygen weaning</td>
<td>Tolerates weaning to room air</td>
<td>Intolerance to attempted weaning</td>
</tr>
<tr>
<td></td>
<td>in many cases</td>
<td></td>
</tr>
<tr>
<td>Lactic acidosis</td>
<td>Common</td>
<td>Common</td>
</tr>
<tr>
<td>Chest radiograph</td>
<td>Cardiomegaly (cardiothoracic ratio &gt; 60%) or abnormal shape of heart</td>
<td>Abnormal lung parenchyma (collapse, consolidation, nonhomogenous lung opacity, or pneumothorax)</td>
</tr>
<tr>
<td>Responsiveness to intravenous PGE₁ or iNO</td>
<td>Dramatic response to PGE₁</td>
<td>Dramatic response to iNO</td>
</tr>
<tr>
<td></td>
<td>Minimal response to iNO</td>
<td>Occasional response to PGE₁</td>
</tr>
</tbody>
</table>
**Hyperoxia test** — The hyperoxia test is useful in distinguishing cardiac from pulmonary causes of cyanosis.

- In CHD associated with obligate right-to-left shunting resulting in cyanosis, blood in the pulmonary veins is fully saturated with oxygen in ambient air. Administering higher concentrations of inspired oxygen increases the amount of dissolved oxygen but has minimal effect on oxygen tension levels because there is no effect on the deoxygenated blood that is shunted to the systemic circulation.

- In contrast, patients with pulmonary disease have pulmonary venous desaturation. Supplemental oxygen administration in pulmonary disease typically increases pulmonary venous oxygen levels and improves systemic oxygenation.
Results: 115 neonates were included.

- Mean gestational age at birth 38.2 + 2.4 weeks
- Median age at admission to NICU 1 (1-26) day

The interventions provided during transport stabilization included:

- mechanical ventilation (n = 86, 75%)
- PGE1 (n = 70, 61%)
- inotropes (n = 41, 36%)
- fluid bolus (n = 50, 43%).

The accuracy of a provisional diagnosis of CHD by transport team was 87.7% and the positive predictive value was 88.1%.

- 60 neonates (88%) received PGE1 appropriately. 8 neonates (12%) with duct-dependent CHD (n = 68) did not receive PGE1 and were considered as missed opportunities.

- Ventilated neonates in groups 1 and 3 were identified as the groups that can potentially benefit from more liberal use of PGE1.
Why do we care?

- Low PaO2 is closely related to white matter cerebral damage.

- Delayed initiation of intravenous PGE1 for neonates with hypoxemia secondary to a duct-dependent pulmonary blood flow lesion may worsen the cerebral insult.

- Patients with provisional diagnosis of CHD but not treated with PGE1 had the lowest rate of mechanical ventilation and remained clinically stable on arrival to the tertiary center.
Opportunities for enhanced transport stabilization

- The decision to not commence PGE1 in these patients may relate to concern of apnea and need for intubation en route. **The administration of a lower dose of PGE1 may be a more desirable strategy in managing relatively stable neonates with suspected CHD.**

- It is well recognized that lower doses of PGE1 are **equally** effective in maintaining the ductal patency, but at lower risk of inducing apnea.

- In clinical situations where an infusion of PGE1 is commenced, it is important that there are skilled transport personnel available to deal with any adverse effects of therapy, most notably the onset of apnea.
To Intubate or Not to Intubate? Transporting Infants on Prostaglandin E1

- Some physicians prophylactically intubate infants on PGE before transporting them to tertiary referral centers.
- This is the first study to compare the clinical transport complications of unintubated and prophylactically intubated infants receiving PGE for CHD.
- A retrospective chart review of 202 infants receiving PGE1 during transport from 2000-2005. PGE1 adverse effects were described as likely or possible and transport complications as major or minor (requiring no intervention).
- Identified risk factors for major transport complications, and subgroup analysis compared risks among unintubated and prophylactically intubated infants.
To Intubate or Not to Intubate? Transporting Infants on Prostaglandin E1

- **Sixty-four percent** of infants were intubated before transport:
  - 34% emergently before PGE1
  - 14% for PGE1-related adverse effects
  - 11% prophylactically
- Likely PGE1 adverse effects were noted in 38% of infants
  - 18% with apnea.
- **Major complications occurred during 42% of all the transports**
  - 7 (10%) of 73 unintubated infants
  - 14 (61%) of 23 prophylactically intubated infants.
- After controlling for multiple factors, **elective intubation was a significant predictor of major transport complications.**
# To Intubate or Not to Intubate? Transporting Infants on Prostaglandin E1

*Pediatrics* 2009;123:e5-e30

## Multivariate Analysis of Major Transport Complications

<table>
<thead>
<tr>
<th>Variable</th>
<th>OR</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medical comorbidity</td>
<td>2.22</td>
<td>1.02–4.08</td>
</tr>
<tr>
<td>$\text{PGE}_1$ dose</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;0.05 μg/kg per min</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>0.05 μg/kg per min</td>
<td>4.80</td>
<td>1.60–14.40</td>
</tr>
<tr>
<td>&gt;0.05 μg/kg per min</td>
<td>3.72</td>
<td>1.10–12.63</td>
</tr>
<tr>
<td>Intubation type</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unintubated</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Emergent</td>
<td>15.68</td>
<td>3.85–63.83</td>
</tr>
<tr>
<td>Elective</td>
<td>7.44</td>
<td>2.82–19.68</td>
</tr>
<tr>
<td>CHD physiology</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Single ventricle</td>
<td>1.42</td>
<td>0.66–3.07</td>
</tr>
<tr>
<td>Transport mode</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ground</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Helicopter</td>
<td>1.17</td>
<td>0.49–2.78</td>
</tr>
<tr>
<td>Fixed wing</td>
<td>0.20</td>
<td>0.02–2.59</td>
</tr>
<tr>
<td>Transport time</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;30 min</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>30–60 min</td>
<td>0.89</td>
<td>0.36–2.19</td>
</tr>
<tr>
<td>60–90 min</td>
<td>0.58</td>
<td>0.18–1.89</td>
</tr>
<tr>
<td>&gt;90 min</td>
<td>3.73</td>
<td>0.44–31.39</td>
</tr>
<tr>
<td>Gender, EGA</td>
<td>NS</td>
<td></td>
</tr>
</tbody>
</table>

EGA indicates estimated gestational age; NS, not significant.
CONCLUSIONS

• Despite high rates of prostaglandin E1 adverse effects, elective intubation of infants for transport significantly increased the odds of a major transport complication.

• The risks of prophylactic intubation before the transport of otherwise stable infants on prostaglandin E1 must be weighed carefully against possible benefits.
Pre-transport stabilization: Available models for pre-transport stabilization and care during transport are:

- **STABLE**: Sugar, Temperature, Artificial breathing, Blood pressure, Laboratory work, Emotional support.

- **SAFER**: Sugar, Arterial circulatory support, Family support, Environment, Respiratory support.

- **TOPS**: Temperature, Oxygenation (Airway & Breathing), Perfusion, Sugar
Summary

1. The neonate with critical CHD decompensates as a result of altered hemodynamics or oxygen delivery as its circulation transitions from that of a fetus to an adult.

2. Accurate diagnosis of defects and predominant physiology is routine.
Summary

3. Maintenance of **Ductal Patency** is usually attainable with **PGE**, providing stability for interventional catheterization or surgical procedures (and transport to tertiary center).
Questions?
Part 1
Sports Participation with Congenital Heart Defects

- Physical activity is important for patients with congenital heart disease.
- Regular exercise at recommended levels can be performed and should be encouraged in all patients with congenital heart disease.
- Physical performance and exercise tolerance is close to normal in patients with simple lesions with successful repair or no need for therapy.
- Most patients with complex lesions have some degree of residual disease, making them less suitable for participation in competitive sport.

“Exercise Prescription”
Sports Participation with Congenital Heart Defects

Classification of Sports

- **Type of exercise**
  - Dynamic
  - Static
- **Level of intensity**
  - Low
  - Medium
  - High

Danger of bodily collision

Increased risk if syncopal

36th Bethesda Conference Eligibility Recommendations for Competitive Athletes With Cardiovascular Abnormalities
JACC Vol. 45, No. 8, 2005
### Bethesda Conference

**Children with Broken Hearts**

<table>
<thead>
<tr>
<th>Increasing Static Component</th>
<th>A. Low (&lt;40% Max O₂)</th>
<th>B. Moderate (40-70% Max O₂)</th>
<th>C. High (&gt;70% Max O₂)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Low (&lt;20% MVC)</td>
<td>Billiards, Bowling, Cricket, Curling, Golf, Riflery</td>
<td>Baseball/Softball*, Fencing, Table tennis, Volleyball</td>
<td>Badminton, Cross-country skiing (classic technique), Field hockey*, Orienteering, Race walking, Racquetball/Squash, Running (long distance), Soccer*, Tennis</td>
</tr>
<tr>
<td>II. Moderate (20-50% MVC)</td>
<td>Archery, Auto racing†, Diving†, Equestrian†, Motorcycling†</td>
<td>American football*, Field events (jumping), Figure skating*, Rodeoing†, Rugby*, Running (sprint), Surfing†, Synchronized swimming†</td>
<td>Basketball*, Ice hockey*, Cross-country skiing (skating technique), Lacrosse*, Running (middle distance), Swimming, Team handball</td>
</tr>
<tr>
<td>III. High (&gt;50% MVC)</td>
<td>Bobsledding/Luge†, Field events (throwing), Gymnastics†, Martial arts*, Sailing, Sport climbing, Water skiing†, Weight lifting†, Windsurfing†</td>
<td>Body building†, Downhill skiing†, Skateboarding†, Snowboarding†, Wrestling*</td>
<td>Boxing*, Canoeing/Kayaking, Cycling†, Decathlon, Rowing, Speed-skating†, Triathlon†</td>
</tr>
</tbody>
</table>

* Increasing Dynamic Component
Does Sports Activity Enhance the Risk of Sudden Death in Adolescents and Young Adults?

Domenico Corrado, MD, PhD,* Cristina Basso, MD, PhD,† Giulio Rizzoli, MD,‡ Maurizio Schiavon, MD,§ Gaetano Thiene, MD†

Padua, Italy
Relative risk of SD
Young athletes (55) vs non-athletes (245)
(Veneto region of Italy; 1979-1999)

RR = 2.5
CI = 1.8-3.4
p < 0.001

<table>
<thead>
<tr>
<th><strong>Sudden Death in Young Competitive Athletes</strong></th>
</tr>
</thead>
</table>
| • Sport activity in adolescent and young adults is associated with an increase in the risk of sudden death *(relative risk=2.5)*  
• Given the substrate of underlying cardiovascular disease such as **congenital coronary anomaly, hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, channelopathies, and premature coronary atherosclerosis**, strenuous physical activity may trigger life-threatening ventricular arrhythmias  
• Therefore, every effort should be made to recognize the cardiac abnormalities implicated in sudden death during preparticipation screening examination |
Sudden Cardiovascular Death (SCD) During Sports Participation:

Goals

• Prevent the event
• Prevent death due to the event

Wes Leonard's Last Shot
Fennville High School basketball
Rate of sudden death during sports participation in the U.S. is not known

- No central registry for sudden death
- Unclear number of sports participants
  - >7 million high school athletes
  - >400,000 NCAA athletes
  - >8 million + recreational athletes (?)
Rate of sudden cardiac death during sports participation in the U.S. is not known

- Generally accepted U.S. estimate is 0.5-2.0/100,000 high school athletes.

- Despite the placement of automatic external defibrillators (AEDs) in public areas including schools and athletic venues, SCD continues to occur despite resuscitative efforts.
Causes of Sudden Cardiac Death in Young Competitive Athletes in the U.S.

**Most common:**
- Hypertrophic Cardiomyopathy
- Congenital coronary artery anomaly

**Less common:**
- Myocarditis
- Aortic rupture (Marfan syndrome)

**Uncommon:**
- Arrhythmogenic RV Cardiomyopathy
- Atherosclerotic coronary artery disease
- Channelopathies
- Aortic valve stenosis
Hypertrophic Cardiomyopathy

HCM is a heterogeneous disease genotypically, phenotypically, patho-physiologically, clinically and therapeutically.

A genetic defect is invariably present:

single-gene disorder with autosomal dominant pattern

> 13 genes
>
> 300 mutations identified

Familial transmission is frequent (50-60%)
Mutations Affecting Contractile and Z-disc Proteins


Children with Broken Hearts
Children with Broken Hearts

HCM

Myocardial Disarray
Hypertrophic Cardiomyopathy
Hypertrophic Cardiomyopathy

**Syncope**

- Atrial or ventricular tachyarrhythmias or bradyarrhythmias
- Heart Block
- Obstruction to LV outflow
- Diastolic dysfunction
- Myocardial ischemia
Hypertrophic Cardiomyopathy Screening Investigations - ECHO

- Most important test
- Location and extent of LVH
- Systolic and diastolic function
- Presence and severity of SAM
- Severity of obstruction (response to Rx)
- Degree of MR, anatomic abnormalities
Anomalous Coronary Artery

Children with Broken Hearts
Marfan Syndrome

This person with the Marfan syndrome is tall and thin and has an arm span that exceeds her height.
Children with Broken Hearts

Marfan Syndrome
Right Ventricular Dysplasia

ARVD is defined as “a cardiac disease mainly involving the right ventricle, characterized by variable replacement of myocardium with adipose tissue, and associated with ventricular arrhythmias and a risk of sudden cardiac death.”

- Familial pattern in ~50%; Autosomal Dominant
- Variable penetrance
- Multiple loci on chromosomes 14, 1, 10

- ARVD, a disease of myocardial stretch?
- Impairment of cell-cell adhesion (desmosome)
- Disruption of cardiomyocytes in response of mechanical stress or stretch

Histology: Fatty infiltration with remaining strands of myocardial fibers + thin fibrosis
Natural History: ARVD

- "Concealed" phase - subtle RV changes; +/- minor ventricular arrhythmias; rarely SD during competitive athletics
- "Overt electrical disorder" – overt RV structural/functional changes; symptomatic RV arrhythmias, syncope, SCD
- "Right heart failure" – global RV dysfunction with preserved LV function
- "Biventricular pump failure" – significant LV dysfunction
12-lead ECG: ARVD

- Localized (right-sided prolongation of QRS – 58%)
- Epsilon waves – 30% “intraventricular myocardial defect”
- TWI beyond V3 – 80%
- Prolonged S-wave upstroke > 55ms ~ 90%
MRI: ARVD

- The definitive test?
- Requires experienced cardiac imaging specialist
- Can distinguish fat from muscle
- Cine MRI helps qualitate free wall function
Estimated that **1 in 10,000 persons** are gene carriers. LQTS causes **3000 to 4000** sudden deaths in children and young adults each year in the US. Prolonged ventricular repolarization and a propensity for life-threatening ventricular tachyarrhythmias resulting in syncope and sudden death is characteristic.
### Table 2. Clinical Characteristics in Common Genotypes of Congenital Long QT Syndrome*

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Genotype</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>LQT1</td>
</tr>
<tr>
<td>Gene mutated</td>
<td>KCNQ1 (= KvLQT1)</td>
</tr>
<tr>
<td>Current affected</td>
<td>I_Ks</td>
</tr>
<tr>
<td>Estimated prevalence, %</td>
<td>42</td>
</tr>
<tr>
<td>Events occurring with exercise or emotional stress, %</td>
<td>97</td>
</tr>
<tr>
<td>Exercise-related trigger</td>
<td>+++</td>
</tr>
<tr>
<td>Other triggers</td>
<td>Diving</td>
</tr>
<tr>
<td>Events before age 10 y, %</td>
<td>40</td>
</tr>
<tr>
<td>Events before age 40 y, %</td>
<td>63</td>
</tr>
<tr>
<td>Median age at first event, y</td>
<td>9</td>
</tr>
<tr>
<td>Mean corrected QT interval, ms</td>
<td>490 ± 43</td>
</tr>
<tr>
<td>QT shortening with exercise</td>
<td>Below normal</td>
</tr>
<tr>
<td>Efficacy of β-blockade to prevent events</td>
<td>+++</td>
</tr>
<tr>
<td>Efficacy of mexilentine to shorten QT interval</td>
<td>—</td>
</tr>
</tbody>
</table>

* Adapted from Wilde AA, Roden DM. Circulation. 2000;102:2796-98 (58), with permission. Clinical data derived from reference 36. +++ = strong association; ++ = moderate association; + = weak association; — = no effect.
American Academy of Pediatrics
Section on Sports Medicine and Fitness

• SCREENING EXAMINATION
  • Before participating in any sports, young athletes should have a complete physical exam that includes a detailed personal and family history of any heart conditions.
  • Exam should be done by a health care provider with the training, medical skills, and background to recognize heart disease.
  • Preparticipation Cardiovascular Screening of Competitive Athletes AHA/ AAP Recommendations (14-point screening)
SUDDEN CARDIAC DEATH IN YOUNG ATHLETES
Can the Cardiac Pre-participation Examination Save Lives?

YES

But not every life at risk.
Children with Broken Hearts

Comprehensive care saves teenager following sudden cardiac arrest

On April 19, 2007, while walking across the cafeteria at Powhatan High School in Virginia, junior Brittnay Worsham dropped dead to the floor. A heart arrhythmia sent the healthy 14-year-old cheerleader unexpectedly into cardiac arrest. Two teachers, both volunteer firefighter-paramedics,一听 CPR, grabbed the automated external defibrillator off the wall, and administered shocks to restart her heart in a normal rhythm.

Worsham was rushed to a nearby hospital, which sent her to the VCU Children’s Medical Center for treatment, where, on the cardiac surgery unit, she received an extensive workup for the cause of the arrhythmia.

Pediatrics and internal medicine chair of the VCU Division of Pediatric Cardiology, Dr. R. Russell Hepplewhite, said Worsham might have been experiencing a form of short-coupled atrial arrhythmia, which can occur in the absence of any identifiable cause.

Once stabilized, doctors induced Worsham into a state of mild hypothermia, a lifesaving technique called “cooling.” This procedure brings the body temperature down to about 91 degrees Fahrenheit, not only giving the patient a better overall chance of surviving after a cardiac arrest but also increasing the chances of improved long-term function.

Worsham spent three weeks at the heart center. She underwent a cardiac catheterization and underwent an electrophysiology study during which an arrhythmia she was ablated with a radiofrequency catheter. She was then implanted with a defibrillator to protect her from future life-threatening arrhythmias.

Today, Worsham, now 17, is a picture of health. After making a full recovery, she’s back in school, cheering for the Powhatan Indians and graduating with her class this June.

“She is a remarkable survivor,” Moskowitz said. 

VCU becomes one of the first in the U.S. to offer a Doctor of Nurse Anesthesia Practice degree.
VCUHS has seen first hand the survival benefit of children that received immediate CPR and defibrillation in surrounding school systems.

A number of these older children have been transported to VCUHS and have received the benefit of therapeutic hypothermia through VCUHS’s ARCTIC Program.

Since its inception in February 2008, the Advanced Resuscitation, Cooling Therapeutics, and Intensive Care (ARCTIC) program has admitted > 700 patients suffering from cardiac arrest.
Children with Broken Hearts

Post-Cardiac Arrest Care

Circulation. 2013;127:244-250.
The ARCTIC program partners with other community hospitals to provide advanced post resuscitation care for their patients as well through the ARCTIC referral center.

Goes well beyond providing just hypothermia treatment.

Begins with the EMS responders, who have been trained to begin decreasing the body temperature of a cardiac arrest patient immediately during and following resuscitation.

Studies have shown the quicker the cooling process begins, the better the outcomes. **Therapeutic Hypothermia**

In the past, the temperature was lowered by wrapping the patient in cooling blankets - took four to six hours to decrease the body temperature sufficiently.

Now, paramedics begin cooling during resuscitation. They start running in two liters of ice-cold saline through an IV right away.

A very aggressive resuscitation program within Richmond’s EMS system.
Hypothermia Defined

- Mild hypothermia = 32-35°C
- Moderate = 28-32°C
- Severe = 20-28°C
2005 Recommendation
AHA & ILCOR

- Recommended inducing & maintaining for 12 to 24 hours, Therapeutic Hypothermia (33°C) after ROSC in patients experiencing OHCA who remain comatose hours after resuscitation & in whom the initial cardiac rhythm is VF.
Pathophysiology of Post-Cardiac Arrest

Brain ischemia during cardiac arrest

(Global vs Focal)

Inflammation and injury

Increased ICP

Poor neurological outcome
Physiologic Effects of Hypothermia

- Desired effects:
  - Decreases the cerebral metabolic rate (1°C = 5-8%)
  - Inhibits influx of Ca & glutamate accumulation
  - Suppresses ischemia-induced inflammatory cytokines
  - Reduces disruptions in BBB & vascular permeability
<table>
<thead>
<tr>
<th>Physiologic Effects of Hypothermia</th>
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<tr>
<td>• <strong>Decreased oxygenation</strong></td>
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<td>• Shifts oxyhemoglobin-dissociation curve to left</td>
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<td>• Vasoconstriction</td>
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<td>• VQ mismatch</td>
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<tr>
<td>• Increased blood viscosity (Hematocrit increases 2% per 1 degree C decline in temperature.)</td>
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<tr>
<td>• <strong>Metabolic acidosis</strong></td>
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<tr>
<td>• Lactate generation due to shivering and decreased tissue perfusion</td>
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<td>• Impaired hepatic metabolism and impaired acid excretion.</td>
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</table>
• ARCTIC and programs like it consider cooling to be only a single component of a comprehensive, care-bundled strategy paralleling traumatic brain injury (the neurocognitive defects are almost identical).
• Much of its benefit is likely to be in prevention of fever as well as minimizing reperfusion injury.
• Continuous EEGs (up to 30% of these pts have seizures - including status -- and they are not identified during mechanical ventilation if paralytics are used unless continuous EEGs are used).
Pediatric Cardiac Arrest

- About **16,000** children suffer from cardiac arrest each year in the United States.¹

- Survival rates in children are around **27%** after in-hospital cardiac arrest and **5% to 10%** after out-of-hospital cardiac arrest.¹

- Cardiac arrest in children outside of the hospital is often caused by events like near drowning or SCD. Such events typically lead to severe hypoxia and, if of sufficient duration, lead to cardiac arrest. Low cardiac output, pulmonary hypertension, hypoxia, and arrhythmias are some of the potential causes leading to cardiac arrest. **VF is uncommon to rare.**

- Successful resuscitation in children with full cardiac arrest without long term brain damage is **low**, especially in out-of-hospital arrests, primarily because of the length of time the child is hypoxic and subsequent tissue damage and metabolic acidosis.

The THAPCA study “Therapeutic Hypothermia after Pediatric Cardiac Arrest (THAPCA)” investigated whether body temperature control improves outcomes in children after cardiac arrest.

About 900 children were enrolled in this study at 41 clinical centers throughout the US and Canada. Enrollment in the THAPCA Trials began in September, 2009.
Results

In comatose children who survive of out-of-hospital cardiac arrest, therapeutic hypothermia (33 degrees for 120 hours), as compared with therapeutic normothermia (36 degrees, fever prevented), did not confer a significant benefit with respect to survival with good functional outcome at 1 year.

Survival at 12 months did not differ significantly between the treatment groups.
Pediatric Cardiac Arrest

The Chain of Survival

Early Access → Early CPR → Early Defibrillation → Early Advanced Care → Comprehensive Cardiac Care
AHA/ACC SCIENTIFIC STATEMENT

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Preamble, Principles, and General Considerations

A Scientific Statement From the American Heart Association and American College of Cardiology

Barry J. Maron, MD, FACC, Co-Chair*  Douglas P. Zipes, MD, FAHA, MACC, Co-Chair*  Richard J. Kovacs, MD, FAHA, FACC, Co-Chair*
Thank you for your attention.

QUESTIONS?