Sudden Unexplained Death In Childhood (SUDC)

Erin Bowen, MD
11/05/19
Connecticut American Academy of Pediatrics Webinar
"the sudden death of a child older than one year of age which remains unexplained after a thorough case investigation, including review of the clinical history and circumstances of death, and performance of a complete autopsy with appropriate ancillary testing."

Krous et al., Ped Dev Path 2005
Conor’s Story

- Born at 37 weeks
- Repeat C-section
- Normal development
- 4 ear infections in first year of life
- 1 episode of wheezing
Conor’s Story

- 17 months old
- Fever 102, Rhinorrhea
- Unresponsive after 1 hour nap
- CPR started at home
- Transported to ER
- Autopsy normal

Cause of death = Undetermined (R99)
Goals

• To review the epidemiology of SUDC
• To discuss the challenges
• To review the research
• To describe the role of the pediatrician after these deaths
• To discuss SUDC Foundation Resources
Epidemiology

- **Category** (NOT a diagnosis) of death in children ages 1 through 18
- Remains **unexplained** after a thorough investigation, including autopsy
- Most often during **sleep**
- Unpredictable and **unpreventable** at this time
Incidence

- Approximately 400 children annually in U.S.
- Exact incidence unknown
  - Varied investigation/certification practices
    - Sudden unexplained or SUDC=R96
    - Undetermined=R99
    - Mild Pathologic Findings= Explained Cause of Death (COD)
    - Possible or Probable Cause Listed= Coded as that specific COD
Causes of Death (Ages 1-4)

Leading causes of death ages 1-4: U.S. 2017

- Motor vehicle
- Drowning
- Other unintentional injury
- Unintentional Injury
- Congenital Anomalies
- Malignant Neoplasms
- Homicide
- Ill-defined and unknown causes

Source: CDC, National Vital Statistics System, 2017
### U.S. SUDC Death Rates (2017)

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Deaths</th>
<th>Crude Death Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-4</td>
<td>243</td>
<td>1.5/100,000</td>
</tr>
<tr>
<td>5-9</td>
<td>33</td>
<td>.2/100,000</td>
</tr>
<tr>
<td>10-14</td>
<td>39</td>
<td>.2/100,000</td>
</tr>
<tr>
<td>15-18</td>
<td>74</td>
<td>.4/100,000</td>
</tr>
</tbody>
</table>

**CDC Wonder Database: ICD codes R96-99**

**Similar crude death rates for northeast census region**
## Sudden Infant Death vs SUDC

<table>
<thead>
<tr>
<th></th>
<th>Sudden Infant Death</th>
<th>SUDC (Age 1-4)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Male Sex</strong></td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td><strong>Ethnicity</strong></td>
<td>African American+</td>
<td>African American+</td>
</tr>
<tr>
<td></td>
<td>American Indian/Alaska Native+</td>
<td>American Indian/Alaska Native+</td>
</tr>
<tr>
<td><strong>Sleep Related</strong></td>
<td>++</td>
<td>++</td>
</tr>
<tr>
<td><strong>Seizures</strong></td>
<td>- =</td>
<td>+ (Febrile)</td>
</tr>
<tr>
<td><strong>Prone Position</strong></td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td><strong>Other Associations</strong></td>
<td>Smoke exposure, Bedsharing, unsafe sleep environment</td>
<td>Illness/fever in 48 hours prior to death</td>
</tr>
</tbody>
</table>
Challenges

- Category of death; not a diagnosis
- Rare: Difficult to identify trends
- No known risk factors or prevention strategies
- Limited Awareness
  - Limited medical training around pediatric deaths
- Variable certification practices may lead to inaccurate statistics
  - Some unexplained certified as “explained” (e.g. mild URI)
  - “Ill-defined” causes not included in reports
- Autopsy limitations- cannot detect arrhythmia/seizure
- Research Funds Scarce
Addressing the Challenges:

Research, Consensus Guidelines & Family Support
Research
151 Case Referrals to the San Diego SUDC Research Project (2001-2011)

123 Case Referrals to the SUDC Foundation (2011-2014)

*Limited Retrospective Reviews, not population based

<table>
<thead>
<tr>
<th>SUDC Profile</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
</tr>
<tr>
<td>Age 1-3</td>
</tr>
<tr>
<td>Term, singletons</td>
</tr>
<tr>
<td>Prone, often face down</td>
</tr>
<tr>
<td>Most unwitnessed, during apparent sleep</td>
</tr>
<tr>
<td>30% with febrile seizures</td>
</tr>
</tbody>
</table>

Hesdorffer et al, *Epilepsia* 2015
Epidemiology Profile (Ireland, N=44)

Population based analysis, Ireland

Unexplained Deaths aged 1-4 from 1994-2008

SUDC Death Rate *increased* from 0.08 to 0.18 per 10,000 population aged 1-4

SIDS Death Rate decreased from 0.71 to 0.34 per 1000 live births

McGarvey, et al Arch Dis Child 2012
31.7% of SUDC children with Febrile Seizures

Hesdorffer et al, Epilepsia 2015
SUDC and Febrile Seizures (N=391)

622 Children aged 1 to 17 years decedents registered with the SUDC foundation

414 Children aged 1 to 17 years decedents whose families completed interviews

391 Children aged 1 to 6 years decedents whose families completed interviews

127 SEDC deaths

12 Accidental deaths

115 Natural deaths

73 Infectious deaths

42 Noninfectious deaths

264 SUDC deaths

181 SUDC without seizure history

76 SUDC with febrile seizure only

7 SUDC afebrile seizure history

28.8% General Population: 2-5%

Crandall LG, et al. JAMA 2019
### SUDC Associations

<table>
<thead>
<tr>
<th>Association</th>
<th>SUDC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male Sex</td>
<td>146/264 (55.3%)</td>
</tr>
<tr>
<td>Prone Position</td>
<td>173/211 (82%)</td>
</tr>
<tr>
<td>Face Down</td>
<td>122/192 (63.5%)</td>
</tr>
<tr>
<td>Apparent Sleep</td>
<td>253/264 (95.8%)</td>
</tr>
<tr>
<td>Terminal Fever</td>
<td>112/245 (45.7%)</td>
</tr>
</tbody>
</table>

**Adjusted p = .008**

Median age 20 months

No repeat SUDC death among 3144 sibling life years

Crandall LG, et al. JAMA 2019
Death Investigation Timeline

Length of Time to Receive Cause of Death

- < 4 weeks: 19
- 4-6 Weeks: 31
- 7-10 Weeks: 32
- 3-4 Months: 30
- 5-6 Months: 8
- > 7 Months: 13

Crandall, et al 2012
Research: Future Directions
“Finding out about the SUDCRRRC (SUDC Registry & Research Collaborative) allowed us to start picking up the pieces. Enrolling in the study made us feel like we were doing something. It gave us hope, and hope is a powerful thing when your healthy child goes to bed and doesn’t wake up. I cannot imagine receiving a death certificate with cause of death unknown and feeling like there was no one trying to find answers.”

-SUDC Parent
SUDC Registry and Research Collaborative

- Created in 2014 at NYU Langone Health
- To understand risk factors and causes and develop preventative strategies
- Multisite collaboration: NYU Langone Health, Columbia University, Mayo Clinic and Clinical and Forensic Collaborators
- **Inclusion Criteria**: Sudden unexpected death of child 11 months to 18 years where an investigation was performed and the cause of death was considered undetermined
- ~ 200 enrolled
- Neuroimaging, Neuropathology, Genetic Analyses and Case Reviews

**To Refer a Case:**
laura.crandall@nyulangone.org
Consensus Guidelines
SUDC is a Multidisciplinary Issue

- EMTs/Paramedics
- Law Enforcement
- Fire Department

- Physicians
- Social Workers
- Clergy

First Responders

Emergency Room

Pediatricians & Clinical Specialists

Death Investigation Agencies

- Primary Care
- Neurology
- Cardiology
- Genetics
- Psychiatry

- Medical Examiners
- Coroners
- Death Investigators
- Child Welfare
Grant funded by SUDC Foundation

Collaboration between National Association of Medical Examiners (NAME) and American Academy of Pediatrics (AAP)

Guidelines for death investigation, death certification, family needs and research needs for sudden death in pediatrics

In Press: sudpeds.com
Protocols and Education for Family Care

- Trauma informed protocols
- Training for first response teams
- Collaboration and communication between professionals and agencies
- Education for hospital teams about local medicolegal death investigation system

Unexplained Pediatric Deaths In Press
Immediate Family Care

- **Resuscitation**
  - Family presence
  - Social work and clergy
  - Designated point person

- **Communication with Families**
  - Private room
  - Use child’s name
  - Allow family to hold child (with supervision)

- **Communication with Professionals**
  - Notify primary care physician
  - Communicate with medical examiner

Memorial keepsakes (handprint, footprint, lock of hair)

*Pediatrics 2014*  
*Unexplained Pediatric Deaths In Press*
"We left the hospital with a sheet of paper. No one even told us about the medical examiner or Child Protective Services."

-SUDC Parent
Immediate Resources for Families

Bereavement Information
- Grief Literature
- Support Groups

Medical Examiner Information
- Role of medical examiner
- Contact information

Emergency Department
- Contact information
“After receiving great care for the past 30 hours, when it came time to leave the hospital shortly after he died, we felt we were left alone. We were in shock and confused and no one walked us out or said goodbye. We even got lost in the hospital trying to find our way out. Also, no one mentioned SUDC.”

SUDC Parent
Ongoing Family Care

- Ongoing Care
- Child Death Review Teams
- Home Visits for Families
- Ongoing communication throughout investigation

Unexplained Pediatric Deaths In Press
Role of the Pediatrician
Role of the Pediatrician: Immediate

Express condolences

Attend funeral

Face to face meeting

Refer to SUDC Foundation

Assess and re-assess need for support and referrals

AAP Policy: Supporting the Family after the Death of a Child, 2012
Unexplained Pediatric Deaths 2019
Role of the Pediatrician: Death Investigation and Beyond

- Communicate with medical examiner
- Provide updates throughout investigation process
- Offer to read autopsy with family
- Assess and re-assess need for support and referrals

Unexplained Pediatric Deaths, In Press
“To receive the autopsy results by mail with no warning was to be perfectly honest, like a kick to the gut when I was already down. Reading this extremely technical document filled with findings and determinations in complex medical language was also completely overwhelming.”

-SUDC Parent
Siblings: “The Forgotten Mourners”

- Varied responses by developmental age (anger, indifference, sadness)
- Guidance for parents on how to talk to children about death
- Age Appropriate Resources: books, grief support groups, mental health referrals
  - SUDC.org
  - Dougy Center
  - National Alliance for Grieving Children
- Reassess at annual visits and around anniversaries/birthdays
- Consider a flag in the chart
- Screen subsequent siblings as well

AAP Policy 2012
Unexplained Pediatric Deaths, In Press
Siblings: Medical Care

• Potential for more visits
  • Balance compassionate care without over-medicalizing
• Cardiac Referral for all first degree relatives
• Additional Referrals as indicated:
  • Neurology, Genetics
  • Research opportunities (SUDCRRC)
• Utilize resources from SUDC Foundation at sudc.org:
  • “Caring for the SUDC Family: Medical and Bereavement Information for the Clinician’s Consideration”

AAP Policy 2012
Unexplained Pediatric Deaths, In Press
Resources for SUDC
SUDC Foundation Resources: Advocacy, Information, Research & Support

- Resources for Professionals and Families
- Published Research
- Family Liaison for MDI Office
- Case Advocacy
- Virtual Support/Peer Support Program
- DNA Banking Assistance

Refer a Family: SUDC.org
Conclusions

- SUDC is not a diagnosis; it is a category of death
- Surveillance limited- Is it underestimated?
- Comprehensive death investigations are vital
- More research is needed to identify risk factors and prevention strategies
- Clear and compassionate communication is critical
- Pediatricians can act in a unique role as a liaison
- Refer all sudden pediatric deaths to SUDC Foundation early

Refer a Family: SUDC.org
“When a child dies in this way, we all want answers. But we don’t want speculations or guesses. We want true answers. Professionals need to remember that this child in front of them has a group of people that love him or her dearly and that the child isn’t the only patient. The family is too.”

-SUDC Parent
“You are a part of this family’s medical history forever. How do you want to be remembered?”

-Laura Crandall
SUDC Foundation Executive Director
Thank You

NYU Langone Team:
Orrin Devinsky, MD (PI)
Laura Gould Crandall, MA
Daniel Friedman, MD
Mikhail Kazachkov, MD
Heather Lau, MD
Jennifer Lighter, MD
Cynthia Loomis, MD, PhD
John Pappas, MD
Colin Phoon, MD
Timothy Shepherd, MD, PhD
Matija Snuderl, MD
Tom Wisniewski, MD
Christopher William, MD
Dominique Leitner, PhD
Joyce Lee
Jenny Ng

External Collaborators:
Columbia University Medical Center
David Goldstein, PhD
Dale Hesdorffer, PhD

Mayo Clinic, Rochester, MN:
Peter Lin, MD
Joseph Maleszewski, MD
Ross Reichard, MD
Michael Ackerman, MD, PhD
David Tester, PhD

University of North Carolina-Chapel Hill
Matthew Halvorsen, PHD

External Collaborators:
Declan McGuone, MBBCh
Tom Andrew, MD
Kelly Lear, MD
Kristen Landi, MD
Kristin Roman, MD
Keith Pinckard, MD, PhD
Alex Williamson, MD, PhD
Katherine Maloney, MD
Heather Jarrell, MD
Kathy Pinneri, MD
Inma Cobos, MD

Funding:

Special thanks to the SUDC families who have entrusted us with learning from their life’s greatest tragedy. And thank you to the clinicians and medical examiner partner offices for their case referrals.
Questions?