



THE ROLE OF EPILEPSY IN CHILD FATALITIES

Telling Stories to Save Lives



KEY FUNDING PARTNER

FEDERAL ACKNOWLEDGEMENT

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HOUSEKEEPING

Before we get started

- This webinar is being recorded and will be available on the National Center's webpage (URL: www.ncfrp.org).
- Participants are muted. Use the question-and-answer box to ask questions.
- Due to the large number of participants, the speakers may be unable to answer all questions. Unanswered questions will be answered and posted with the recording.
- Contact the National Center (email: info@ncfrp.org) for any tech problems.





EVALUATION

<https://www.surveymonkey.com/r/32BRMMX>

Vicky Whittemore, Ph.D.

Welcome and Introductions

Program Director

National Institute of Neurological Disorders and Stroke

National Institutes of Health





Describe epilepsy and Sudden Unexpected Death in Epilepsy, or SUDEP.



Discuss the relationship between epilepsy and fatalities in infants, children, and youth.



Illustrate lessons learned about epilepsy-related fatalities through the child death review process.



Highlight opportunities to participate in the Sudden Death in the Young (SDY) Case Registry.



PRESENTATION OBJECTIVES

Speakers



Daniel Friedman, M.D., M.Sc.
New York University
Grossman School of Medicine



Chase Foster, B.S.
Child and Infant Fatality Program Director
Tennessee Department of Health



Epilepsy & Premature Mortality

Daniel Friedman, MD, MSc

Professor of Neurology

Feb 14, 2023

What is Epilepsy?

- A spectrum disorder where the cardinal feature is a predisposition to **recurrent unprovoked seizures**
- **Spectrum disorder:** multiple etiologies, multiple coexisting conditions, multiple severities
 - *Well-controlled seizures in an otherwise healthy adult to child with severe intractable seizures, neurodevelopmental delay and autism due to a de novo mutation in the SCNA1 gene*

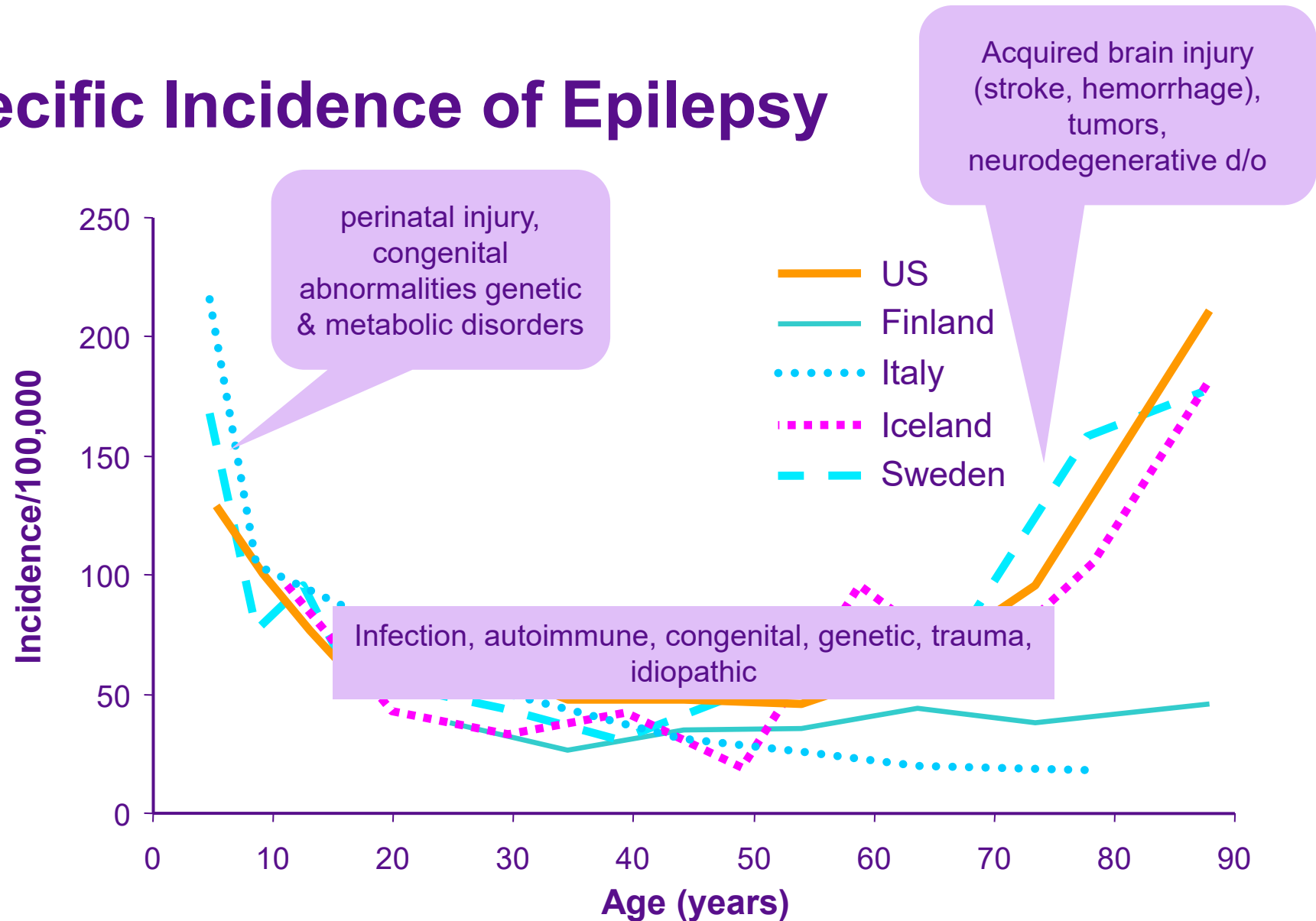
Prevalence of Epilepsy

- By a conservative estimate, 50 million people worldwide have epilepsy¹
- **One in 26 people** will have epilepsy at some point in their lives, and 5% of persons report a seizure at some time in their lives
- Epilepsy is more common than **autism spectrum disorder**, cerebral palsy, **multiple sclerosis** and **Parkinson's disease** combined.



1. Brodie MJ and Dichter MA. *N Engl J Med*. 1996;334(3):168-175.

Age-Specific Incidence of Epilepsy

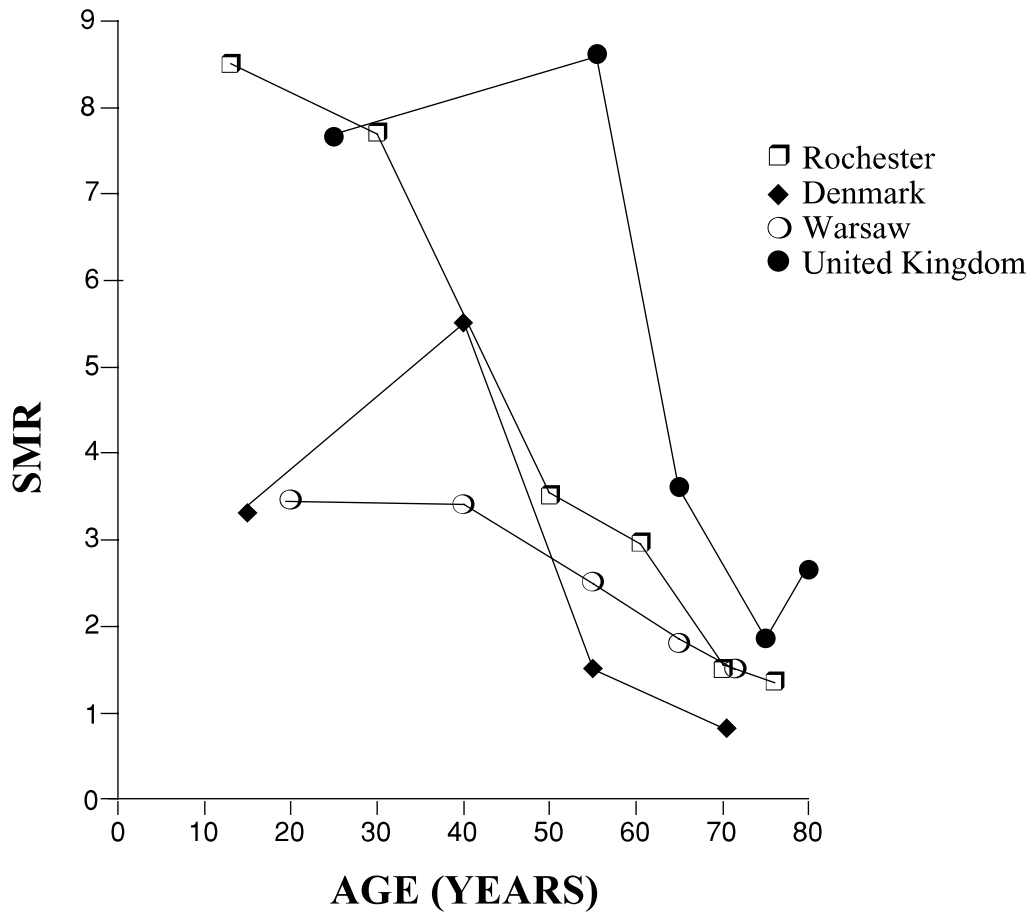


Adapted from Hauser WA. In: Engel J Jr, Pedley TA, eds. *Epilepsy: A Comprehensive Textbook*. Philadelphia, PA: Lippincott-Raven Publishers; 1997:47-57.

Epilepsy Mortality

- **1.6-3** fold higher mortality rates in people with epilepsy
- Many deaths related to underlying cause of epilepsy (metabolic d/o, tumor, etc)
 - Reduced life-expectancy of ~10 yrs
- People without a known cause of epilepsy have a 50% increase in mortality rate compared to the general population
 - Reduced life-expectancy of ~2 yrs

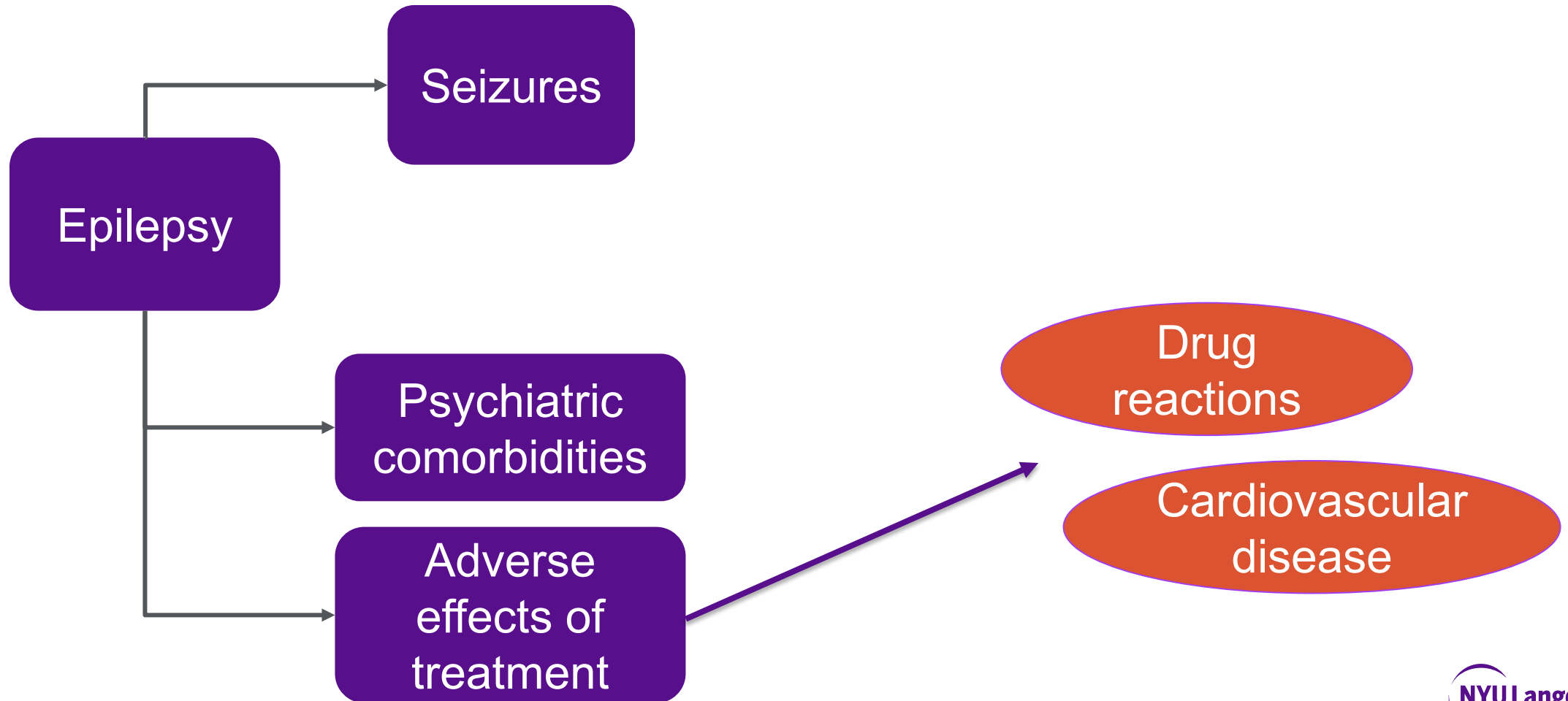
Epilepsy mortality by age



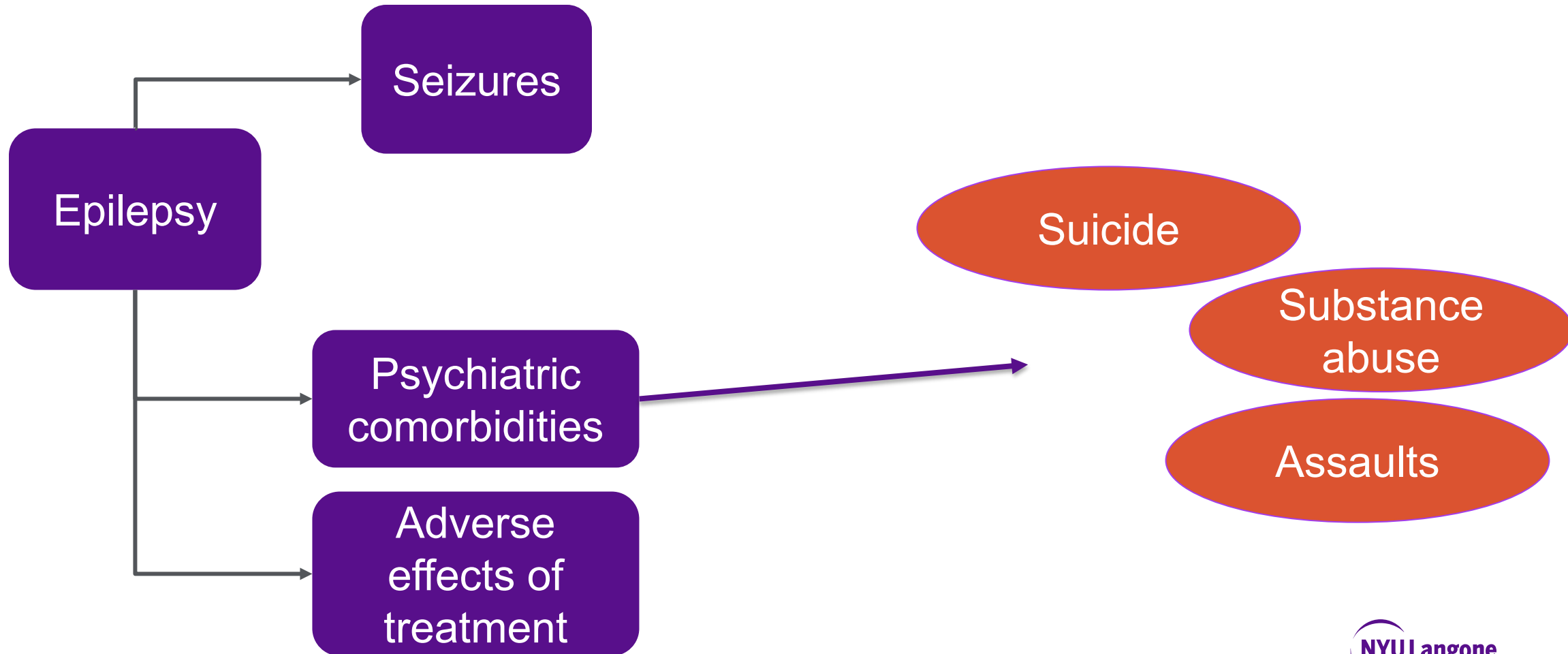
The increased mortality in epilepsy compared to the general population is proportionally greater in young people

FIG. 1. Standardized mortality ratios (SMR) of epilepsy by age in four studies: Warsaw, Poland (4); Rochester, Minn., U.S.A. (5); United Kingdom (6); and Denmark (17).

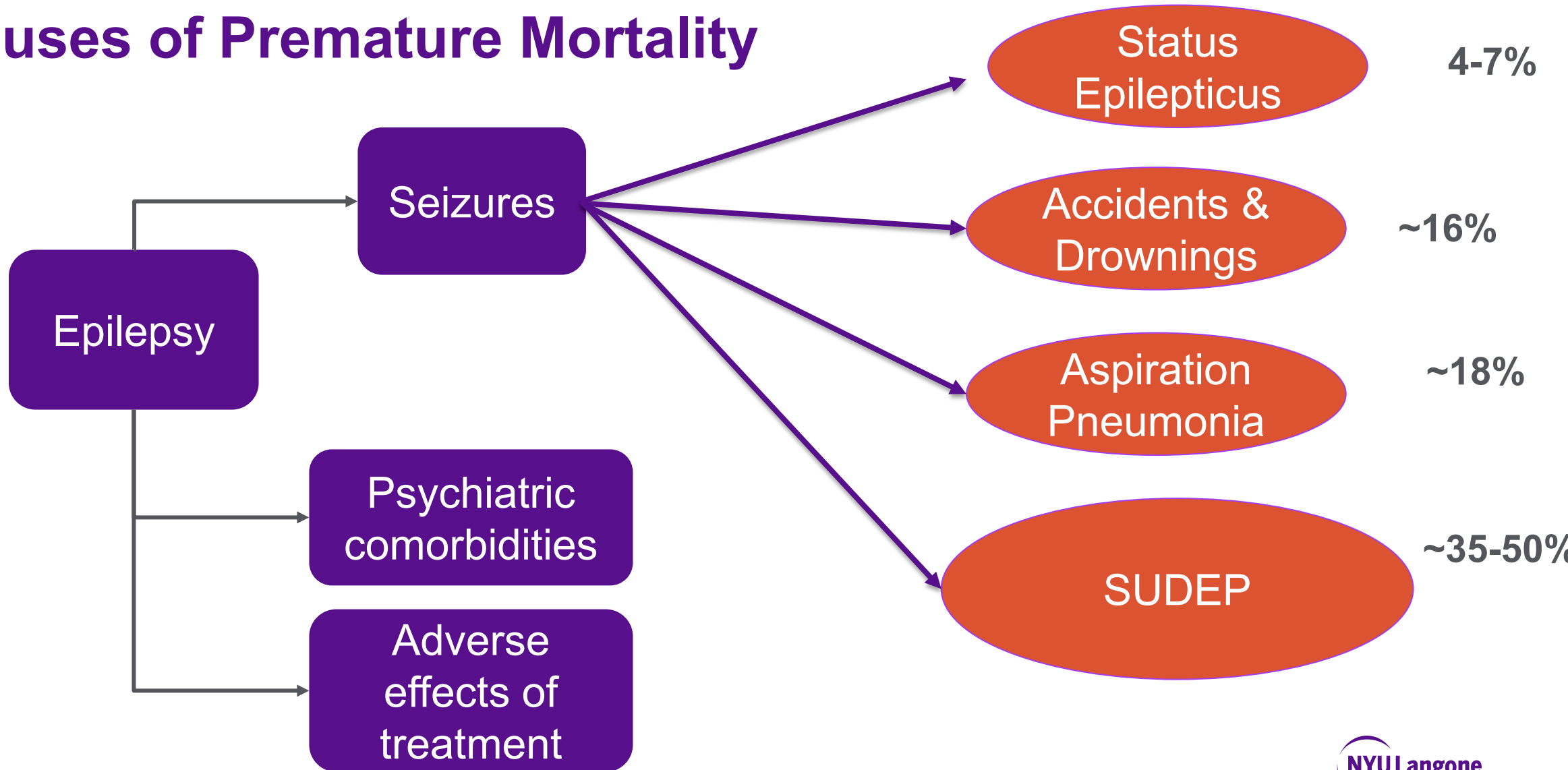
Causes of Premature Mortality



Causes of Premature Mortality



Causes of Premature Mortality



Sudden Unexpected Death in Epilepsy (SUDEP)

- Sudden and unexpected nontraumatic and non-drowning death in a patient with epilepsy (excluding status epilepticus)
 - *Definite* if: autopsy doesn't reveal structural or toxicological cause
 - *Probable* if: no autopsy performed but there is no other plausible cause for death
 - *Possible* if: limited information regarding death circumstances or there is a plausible competing explanation for death
- Designation of SUDEP Plus
 - Coexisting condition that alone would not lead to death but may be mediator
- Near-SUDEP
 - Successfully resuscitated SUDEP (survival > 1 hr)

Nashef Epilepsia 2012;53(2):227-33

Early Descriptions of SUDEP



Mount Vernon, June 20, 1773.

Dear Sir,

It is an easier matter to conceive, than to describe the distress of this Family; especially that of the unhappy Parent of our Dear Patsy Custis, when I inform you that yesterday removed the Sweet Innocent Girl Entered into a more happy and peaceful abode than any she has met with in the afflicted Path she hitherto has trod.

She rose from Dinner about four o'clock in better health and spirits than she appeared to have been in for some time; soon after which she was seized with one of her usual Fits, and expired in it, in less than two minutes without uttering a word, a groan, or scarce a sigh.

This sudden, and unexpected blow, I scarce need add has almost reduced my poor Wife to the lowest ebb of Misery; which is encreas'd by the absence of her son, (whom I have just fixed at the College in New York from whence I returned the 8th Inst) and want of the balmy consolation of her Relations...



SUDEP Incidence

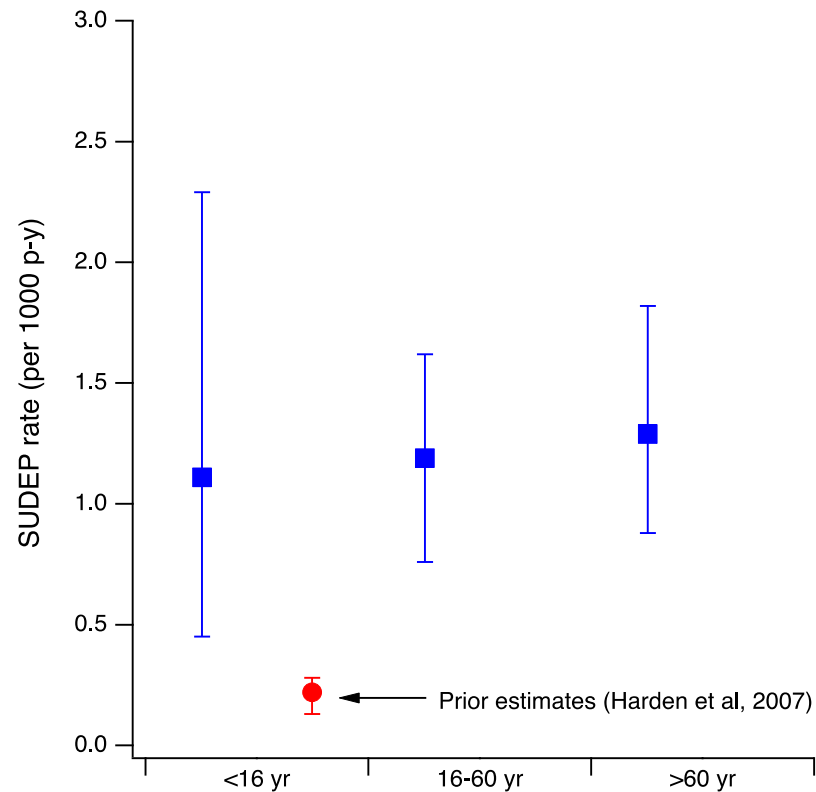


Article

The incidence of SUDEP

A nationwide population-based cohort study

Olafur Sveinsson, MD, Tomas Andersson, BSc, Sofia Carlsson, PhD and Torbjörn Tomson, MD



SUDEP Incidence



ARTICLE

Incidence of sudden unexpected death in epilepsy in children is similar to adults

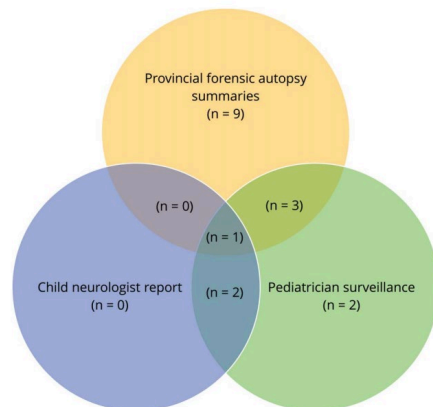
Anne E. Keller, MPH, Robyn Whitney, MD, Shelly-Anne Li, MSc, Michael S. Pollanen, MD, PhD, and Elizabeth J. Donner, MD, MSc

Neurology® 2018;0:1-5. doi:10.1212/WNL.0000000000005762

Correspondence

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Figure Identification of the 17 pediatric cases of sudden unexpected death in epilepsy by data source



- Multiple methods of case ascertainment in Ontario btwn **2014-2015**
- All methods missed some cases
- Used capture-recapture methods to account for incomplete ascertainment
- Estimated incidence:
1.45 (0.90–2.22) per 1,000 pt-years

Risk Factors

**Ongoing Tonic-Clonic
Seizures**

1–2 /yr = **OR 5**

≥3 yr = **OR 15**

Early onset

Long duration

Non-adherence

Remote
Symptomatic/Abnormal
Neurological status

Seizure freedom

Nocturnal
supervision/listening device
OR = 0.1 - 0.4

Practice guideline summary: Sudden unexpected death in epilepsy incidence rates and risk factors

Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society

Harden et al, Neurology 2017

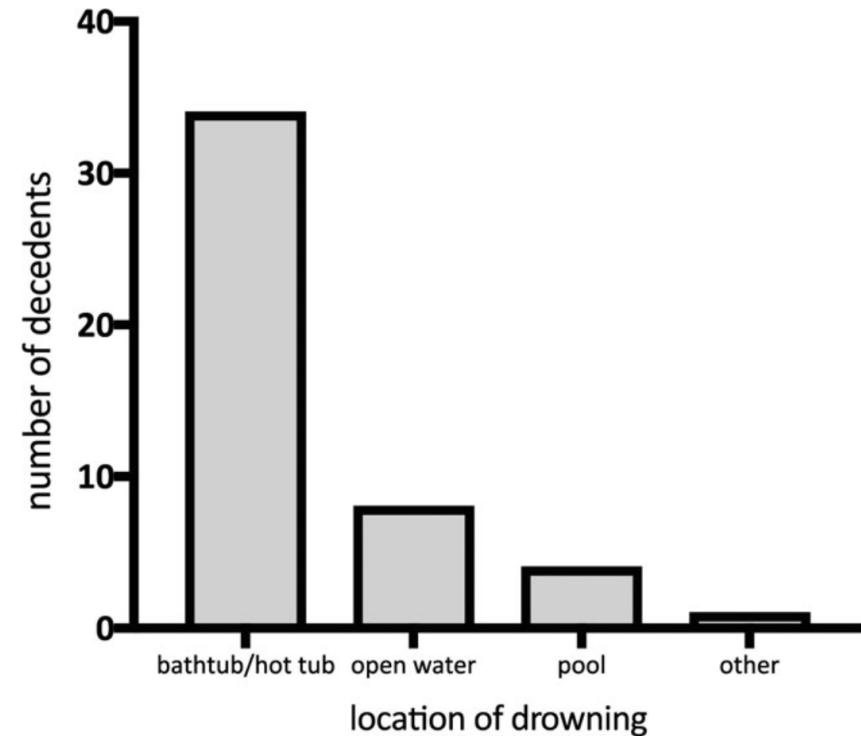
Beyond epilepsy?

SUDEP likely part of spectrum of seizure-related sudden death

- Sudden unexpected death in childhood (SUDC) – ages 1-18
 - 32% had history febrile seizure, mostly uncomplicated FS
- Acute symptomatic seizures - potential COD in 5/14 patients with alcohol-related seizures in one SCD series.

Drowning vs SUDEP?

- PWE – risk of drowning 15-19 x
- 3rd most common cause of bathtub drowning
- Post-mortem findings of drowning are not specific
- No difference in qualitative or quantitative autopsy findings between drowning and SUDEP



Cihan et al, *Epilepsia* 2018

Approach to Death Investigation

- Environment: location, position, witnessed seizure, circumstantial evidence of seizure
- Medical history: epilepsy history, seizure types (convulsive?), medications, adherence
- Toxicology: medication levels, drugs of abuse
- Gross: lung weights, airways, evidence of aspiration, evidence of recent seizure (tongue bite), alternate explanations for death
- Neuropathology: cause of epilepsy
- Micro: competing causes of death



Accepted: 24 January 2018

DOI: 10.1111/epi.14030

SPECIAL REPORT

Epilepsia®

National Association of Medical Examiners position paper: Recommendations for the investigation and certification of deaths in people with epilepsy

Owen Middleton¹ | Daniel Atherton² | Elizabeth Bundock³ | Elizabeth Donner⁴ |
Daniel Friedman⁵  | Dale Hesdorffer⁶  | Heather Jarrell⁷ | Aileen McCrillis⁸ |
Othon J. Mena⁹ | Mitchel Morey¹ | David Thurman¹⁰ | Niu Tian¹¹ |
Torbjörn Tomson^{12,13} | Zian Tseng¹⁴ | Steven White¹⁵ | Cyndi Wright¹⁶ | Orrin Devinsky⁵

Consider seizures and epilepsy when...

- Diagnostic delay in epilepsy
 - Median time of 12 mo to make diagnosis in teens/adults
 - If initial seizures are non-convulsive
 - Decedent may not carry epilepsy diagnosis at time of death
- Screen for history of seizures if:
 - Sudden unexpected death
 - Drowning in benign circumstances
 - Single vehicle accidents

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SDY & SUDEP in Tennessee

Chase Foster | SDY Coordinator

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Overview

- Child Fatality Review
- Sudden Death in the Young
- SUDEP in TN
- Safe Stars Initiative
- SUDEP Dashboard Vision

Child Fatality Review (CFR)

- To review child deaths in order to promote an understanding of the causes of childhood deaths as well as to create and carry out recommendations in order to prevent future childhood deaths.
- 34 Local Teams
 - Multidisciplinary teams meets quarterly and reviews all child deaths (around 900 deaths annually)
 - 17 years and under and 23 weeks or 500 grams
- 1 State Team
 - High level state officials, meet quarterly
 - Reviews aggregate data and develops prevention recommendations

Sudden Death in the Young Case Registry (SDY)

- Tennessee is 1 of 13 states to receive funding for this project
 - Subset of CFR
- Goals
 - Establish the incidence of sudden death in the young in the U.S. using a population-based approach through state public health offices
 - Investigate etiologies and risk factors for sudden death in the young, including Sudden Unexplained Infant Death, Sudden Cardiac Death, and Sudden Unexplained Death in Epilepsy

SDY Criteria

Nationally, all deaths in young people under age 20 may be considered for inclusion in the registry, except in cases where death was due to any of the following:

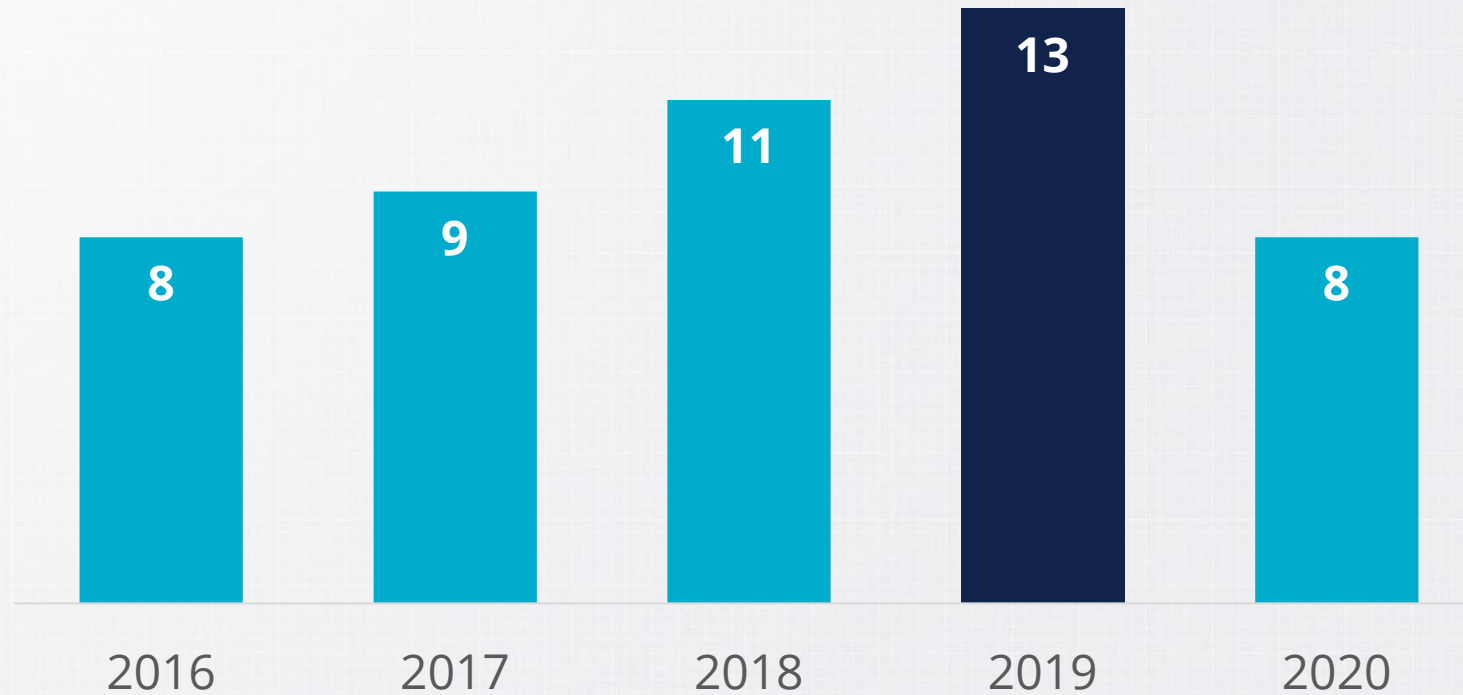
- Accident in which the external cause was the obvious and only reason for the death, **except infant suffocation**
- Homicide
- Suicide
- Accidental or intentional overdose of drugs even if this caused cardiac or respiratory arrest with no prior history of other possible chronic disease or autopsy findings suggestive of another cause
- Terminal illness in which the death was reasonably expected to occur within 6 months

SDY Categorization

- Unexplained, SUDEP
- Unexplained, possible cardiac & SUDEP
- Explained neurological
 - (non-epilepsy related)
- Explained other: status epilepticus
- Other categories
 - Excluded
 - Incomplete Case Information
 - Explained cardiac
 - Explained infant suffocation (under age 1)
 - Unexplained, possible cardiac
 - Unexplained Death

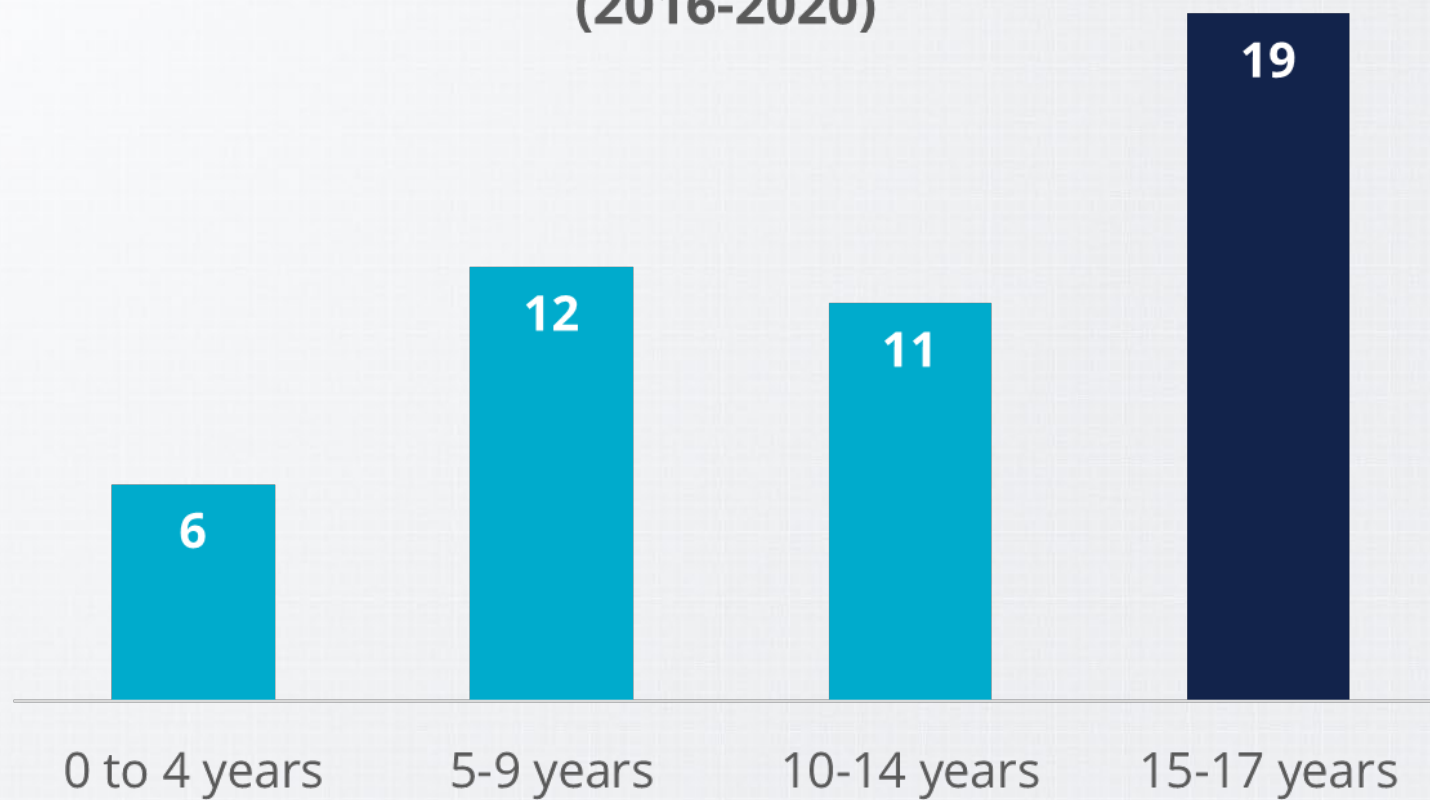
SUDEP in Tennessee

Unexplained, Sudden Unexpected Deaths in
Epilepsy (SUDEP) per Year



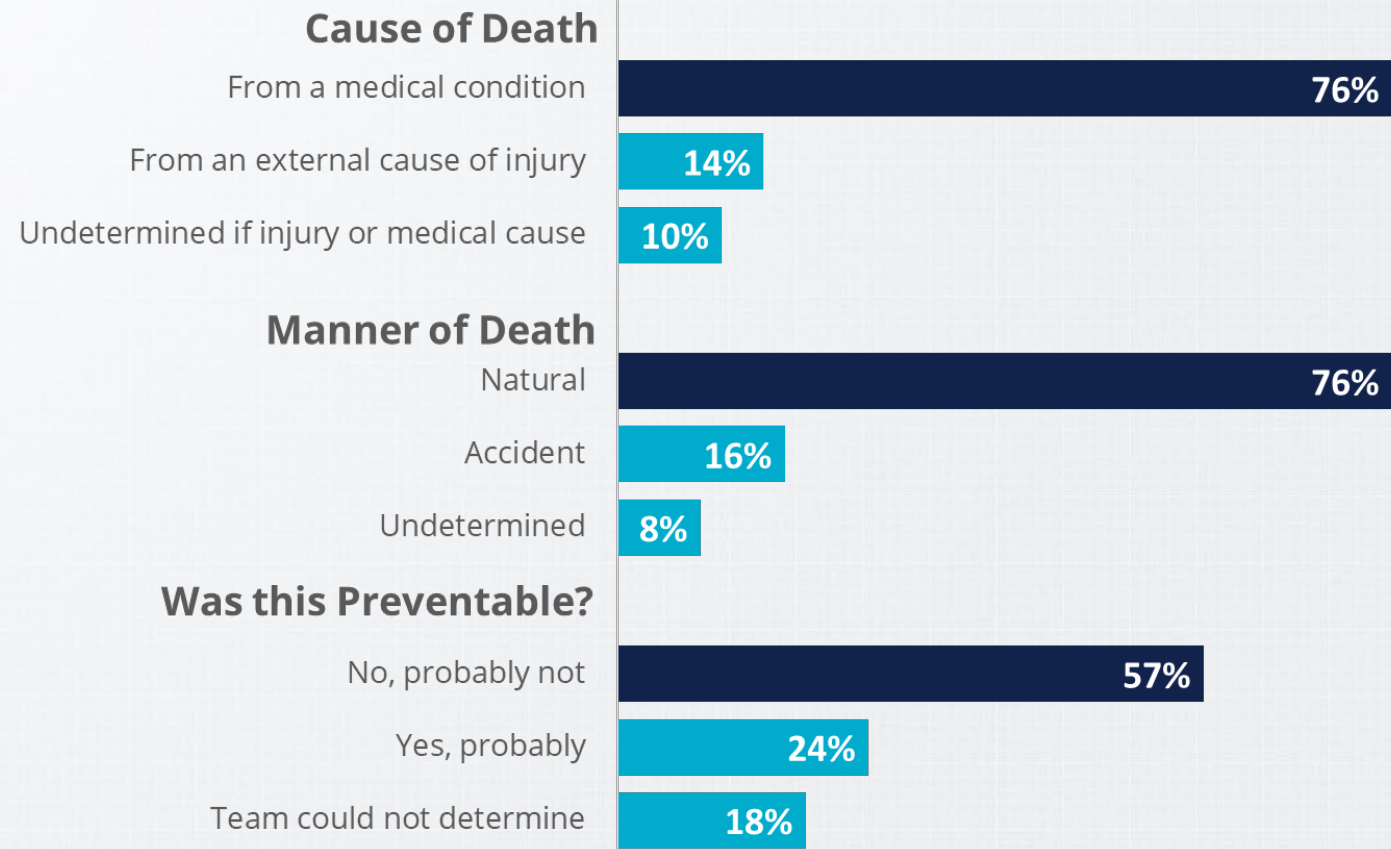
SUDEP in Tennessee

Age at Death for Unexplained, SUDEP Cases
(2016-2020)



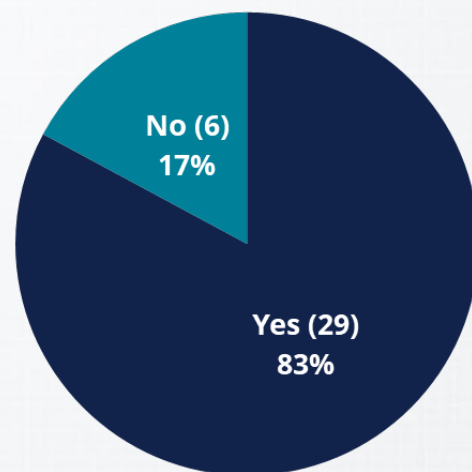
SUDEP in Tennessee

Fatality Review for Unexplained, SUDEP Cases (2016-2020)

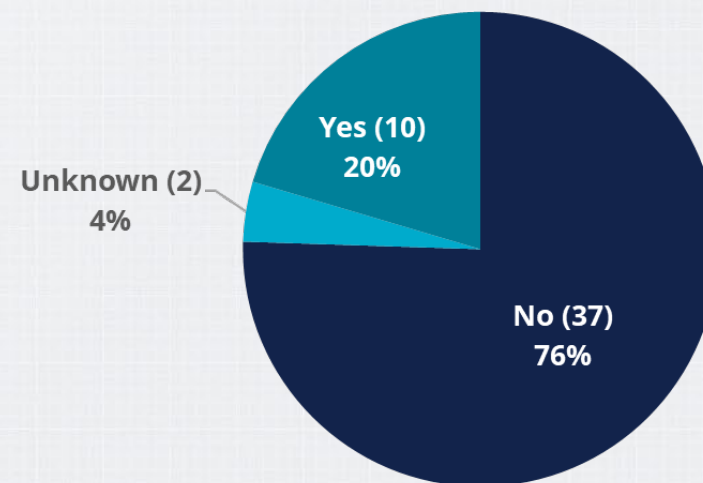


SUDEP in Tennessee

Use of Anti-epileptic Drugs in Unexplained,
SUDEP Cases (2016-2020)



History of Epilepsy or Convulsion/Seizure in Blood
Relatives of Unexplained, SUDEP Cases (2016-2020)



Safe Stars

- Free program that recognizes youth sports leagues throughout Tennessee for providing the highest level of safety for their young athletes.
- Safe Stars consists of 3 levels: gold, silver, and bronze, and involves the implementation of policies around topics such as concussion education, weather safety, and injury prevention.
- Safe Stars goal is to provide resources and opportunities for every youth sports league to enhance their safety standards.



Community Success Story



SUDEP Dashboard Vision

- Medication adherence in SUDEP
 - Obstacles
- Safety education dissemination & follow-through
- Conduct and analyze a national needs assessment for individuals diagnosed with epilepsy
- Conduct and analyze a national needs assessment for families that have experienced a loss from SUDEP

Questions?



Contact Information

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QUESTIONS

WHAT ADDITIONAL INFORMATION WOULD BE HELPFUL?



USE THE QUESTION-AND-ANSWER BOX

The box is located at the bottom of the screen



UNANSWERED QUESTIONS

All unanswered questions will be answered and posted on the National Center's website (URL: www.ncfrp.org).



EVALUATION

<https://www.surveymonkey.com/r/32BRMMX>

UPCOMING WEBINARS

Additional Opportunities



CELEBRATING SAFE SLEEP SUCCESS STORIES: Stories from Fatality Review Teams

- Wednesday, February 22, 2023, 12:00-1:00 PM ET



BUILDING RELATIONSHIPS BETWEEN FATALITY REVIEW TEAMS AND SCHOOLS

- Wednesday, March 15, 2023, 2:00-3:00 PM ET



CONTACT INFORMATION



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A dark silhouette of a person with long, flowing hair, positioned in the center of the frame. The person appears to be looking away from the camera, with their hair blowing in the wind. This silhouette serves as a background for the text and the button.

THANK YOU FOR YOUR TIME!

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