Increasing Awareness of Sudden Death in Pediatric Epilepsy Together

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The death of any child is tragic. When the death is sudden and unexpected, it can seem especially incomprehensible. Henry was 4 years old when he died only a few weeks after his epilepsy diagnosis; his parents were devastated and never knew that death could occur; no physician had discussed the possibility with them. Henry was an otherwise healthy child, had a history of febrile seizures, and died in his sleep before his epilepsy workup was complete and before his medication was likely therapeutic. Since Henry's death 8 years ago, together and independently, Henry's parents, pediatrician, and neurologist have sought understanding and opportunities to advance awareness and prevention around epilepsy-related mortality. These efforts have launched a multidisciplinary partnership between Children's National Health System and the University of Virginia to develop an educational research program to systematically raise provider and patient awareness of sudden unexpected (or unexplained) death in epilepsy persons (SUDEP) and other risks associated with epilepsy. This article gives voice to these different experiences and underscores the value of families and providers working together to improve care and prevent death.

Although epilepsy is a common childhood disorder, all of us were impacted by Henry, who died, suddenly and unexpectedly, as a result of his seizure disorder. Our partnership started with Henry's mother, Henry's pediatrician, and Henry's neurologist and now includes an educational research program that incorporates parent advocacy, nursing, neuropsychology, psychiatry, neurology, and basic science expertise.

HENRY'S MOTHER

As a bereaved mother, I was desperate to make sense out of our tragedy. I searched for understanding and connection to others who shared my experience and ultimately connected with other parents at Citizens United for Research in Epilepsy. I joined their efforts to fund epilepsy research, including SUDEP, and raise awareness about the devastating effects of epilepsy. Through this experience, I learned that rigorous scientific pursuit can affect change and that families can play an important role in the process. One effort that I am especially proud of is my work with other families and professionals to create Partners Against Mortality in Epilepsy, a biannual scientific meeting for families and professionals that seeks to advance SUDEP research and prevention strategies by bringing different stakeholders together to learn from one another. Through these efforts and others, I have met many bereaved family members and the most consistent thing I hear is that they wish they had known about SUDEP. I have more recently had the opportunity to engage with my son's care team, who also cares deeply about this fundamental gap in knowledge. Most families want to know all risks associated with epilepsy and providers face barriers (ie, lack of knowledge, lack of time) when meeting these needs. Together, we are working to educate

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Mrs Lapham and Drs Gaillard, Sexter, and Berl conceptualized the article, each wrote an initial draft of their perspective, and all contributed to the final version to reflect a collaborative voice.

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local providers and families about the risks associated with epilepsy and arm them with knowledge to mitigate harm and death.

HENRY'S PEDIATRICIAN

As Henry's pediatrician, it was not until a conversation with Henry's mother that I heard the term SUDEP for the first time. I had no experience with a child ever dying from epilepsy, nor was this something I ever discussed with parents. Since Henry's death, I am more cautious during the early stages of a diagnosis of epilepsy, when medication may not yet be at a therapeutic level. A specific change that I have instituted since learning about SUDEP is stressing, especially to parents of children with nocturnal and tonic clonic seizures, the importance of medication adherence, because seizure control is the best way to minimize SUDEP risk. I make sure that parents are working with their neurologist to achieve this goal. I also now talk with parents about nighttime supervision and monitoring options, which I previously had not discussed. There is 1 study that demonstrates the benefit of nighttime supervision,[1] and I discuss with families the range of possibilities, including using an infant monitor, having the child sleep in the same room, or having a nighttime sitter. The benefit of sleeping in the same bed rather than using an infant monitor is that the adult may be able to detect the child's seizure better if the seizure can't be seen or heard on an infant monitor. With all supervision methods, there is the opportunity to be able to intervene to stop the seizure and assure safety. The drawbacks of nighttime supervision in the form of cosleeping include potential disruption to family dynamics and developmental socialization and that it is considered one of the greatest risks in sleep-related infant deaths. Nighttime supervision is costly when hiring a nighttime sitter that is not the parent, and disrupts sleep schedules if it is the parent; however, closer monitoring may only be needed on a short-term basis until there is confidence regarding the level of control and frequency of seizures. In addition to supervision, there are ongoing studies regarding monitoring devices, which include monitoring heart rate and respiration. The hope is that an alarm could alert the parents, who could then go in the child's room and ensure that the child is in a safe position and administer a rescue medication as needed. The drawbacks are that these devices are still under investigation and current devices may give families a false sense of security or produce too many false alarms. Ultimately, the family will decide how to monitor seizures by considering the costs and benefits of available options. Regardless, I recommend that the family have rescue medications on hand in case of prolonged or clusters of seizures.

Discussing these options with patients and their families introduces them to the term SUDEP and underscores the importance of treatment adherence. During these conversations, I encourage parents to discuss mortality risk with their child's neurologist. Collaboration among pediatricians and pediatric neurologists is critical to the successful treatment and education of our patients with epilepsy and their families; however, we appreciate that in more rural environments, this may present a challenge due to distance. Yet another reason that the primary care physician needs to be well informed.

HENRY'S NEUROLOGIST

As Henry's neurologist, I was taught during my pediatric and neurology training that epilepsy is a benign disease and that when children die, the cause is primarily from the underlying disease (eg, brain tumor, degenerative, or metabolic disease). Henry proved, sadly, that epilepsy can be deadly and is not a benign disease. In fact, 24% of patients with childhood onset epilepsy will die within 40 years, 3 times the expected mortality rates.[2,3] Before Henry's death, I would wait for families to raise their fears, to the extent they occurred, by asking them nonspecifically if they had any questions. If the concern for mortality was not raised, I would often defer the conversation for another visit, if at all. After Henry's death, I still wait patiently for families to raise their own questions and fears, but if that question does not arise before the close of a clinic visit, I now directly ask what their greatest fear is, and it is invariably harm from seizures and death. When there is no response to my question, then I ask directly, "Do you fear your child will be harmed or die from their seizures?" Capable and skilled physicians do not often discuss mortality.[4,] Yet, families wish to discuss these matters at the first visit, not later, and expect their doctor to do so.[5,6] This is part of our duty and responsibility as physicians. For many children who have "uncomplicated" epilepsy, the data may be reassuring because most children's risk of death is similar to all children, although death in this low-risk population can occur.[3] SUDEP incidence is lowest in young children, but is higher in adolescence (cumulative risk, 1.6%), and peaks in younger adulthood (ages 31–40 years).[7] The overall risk is 33 per 100 000 patient years, but 9 per 100 000 patient years if uncomplicated (normal exam, normal MRI, no developmental disability).[3] For other children, if epilepsy begins in early childhood and never fully remits, the average cumulative risk of SUDEP is ~8% by age 70 years.[7] The large majority who die will do so from their underlying disease and from pulmonary failure. Of the 15% to 20% of epilepsy-related deaths, the large majority are SUDEP-related,

but suicide should not be forgotten.[3] Those at highest risk of death (97 per 100 000 patient years) have early epilepsy onset and frequent generalized tonic clinic seizures (the risk declines with <3 seizures per year), especially nocturnal seizures, and polytherapy (perhaps reflecting epilepsy severity).[8,9] It is thought that successful efforts to achieve optimal seizure control reduces mortality risks.

DEVELOPING OUR PARTNERSHIP

Henry's story and those of others who have died unexpectedly are a wake-up call that there is too much we do not know. It became clear to all of us that education is our most powerful tool and that we do not need to wait for all of the answers before we start preventing SUDEP. At the time of Henry's death, searches for information on the internet came up empty; since then, there are resources for providers and families with information about the facts, prevention strategies, printed brochures, and how to talk about SUDEP (www.epilepsy.com/learn/impact/mortality/sudep).[10] Despite this progress, work still remains to be done. Over several meetings, we developed our first project that brings together many areas of expertise and seeks to change primary and specialty care provider knowledge and practice. On a local level, we are implementing systems that will lead to improved quality of care and, ultimately, we aim to change outcomes. Our work has leveraged local foundation funding and we are connected with broader, federally funded efforts that delineate the risks of SUDEP, examine possible mechanisms of SUDEP (including cardiac failure, cardiac arrhythmia [eg, channelopathies], central apnea, autonomic instability, and pulmonary edema), and, most importantly, identify effective monitoring and devise interventions to decrease mortality. We support efforts to work with medical examiners to recognize epilepsy as a cause of death, promote registries that include DNA collection, and develop clinical and translational research to advance understanding.

Our partnership between families and providers has played a significant role in highlighting care and education areas that needed strengthening and, in the case of SUDEP, needed to be brought out of the shadows entirely. Moreover, our work together has shaped a program of research that includes a novel first finding that 85% of pediatricians do not know about SUDEP.[11] Through their energy and drive, Henry's family has worked closely with our institution and private foundations to promote important SUDEP initiatives. Their efforts, along with those of so many other families and providers, have accelerated the study of an important field (mortality in epilepsy) and, in doing so, have enabled the National Institutes of Health to fund a significant multicenter SUDEP research effort (http://csr.case.edu/index.php/Main_Page). Furthermore, these efforts have helped to create a scientific committee within the American Epilepsy Society dedicated to SUDEP, additionally solidifying the importance of this issue within our community. When families and providers unite, they can more efficiently impact multiple levels of research and care that advance understanding and action around the full scope of a disease.

ABBREVIATION

SUDEP: sudden unexpected (or unexplained) death in epilepsy persons

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