

CARING FOR THE SUDC SIBLING WITH FEBRILE SEIZURES

Facts on Febrile Seizures

- Febrile Seizures (FS) occur in 2-4% of children in the general population between the ages of 6 months and 5 years
- Simple FS: generalized seizures that last < 15 minutes and do not recur within 24 hours. They are generally accepted to be benign and not associated with increased mortality, hemiplegia, or intellectual disability. The risk of developing epilepsy in children with FS is only slightly higher than the general population. One of three children with simple FSs develop one or more recurrent FS(s). The older the child or higher the fever at the time of the first seizure correlates with a lower risk of recurrence.
- Complex FS: FS that are focal (for example: convulsing of only one side), prolonged (≥ 15 minutes), and/or recurrent within 24 hours. Among those with complex FS, 2-4% develop epilepsy.
- FS correlate with illness and usually with a rising fever, but the seizure may precede the fever.
- FS presentations are variable and may present as: tonic-clonic (stiffening followed by shaking), tonic (stiff), atonic (very limp), or staring during the seizure. Breathing may also be affected.

The Current Standard of Care for the treatment of FS generally includes:

- Finding the source of infection and providing appropriate clinical intervention for the infection
- Educate parents regarding recurrence
- Use of fever-lowering drugs
 - Note: Most doctors recommend fever-lowering drugs for children with prior FSs. These medications can reduce fever and discomfort, although their effectiveness in reducing seizures remains unproven.
- Rescue medications: Sometimes the use diazepam during periods of febrile illnesses for those with complex FS or to stop a seizure from being prolonged (See more below)

SUDC and FS

- SUDC is defined as a sudden death of a child older than 12 months that remains unexplained after a thorough investigation.
- Recent SUDC research and referrals to the SUDC program demonstrate a higher incidence of FSs in the child who died (24.4%) and/or in their family history (25%) than we would expect to see in the general population. Notably, both the cases in the SUDC research and those registered with the SUDC are not population-based data, so the incidence known to the program may or may not reflect and accurate incidence occurring in the general population.
- Published references on SUDC and FS are listed at the end of this document.

Considerations for SUDC Siblings with FS

Given these facts, we offer the below suggestions for you to discuss with your Pediatrician and/or Pediatric Neurologist who is treating a SUDC sibling who has experienced simple or complex FS or in family with a history of SUDC and FS.

1) Consider “Rescue Medications”: These are anti-seizure medications that are kept readily available in case of a FS that can stop the seizure. These medications should be used if the FS lasts more than 3-5 minutes or earlier if the child has a history of prolonged FSs. Since the duration of the FS, especially >10-15 minutes correlates with the risk of subsequent epilepsy (ie, seizures not related to fever), prolonged FSs should be stopped.

- Diastat is a brand of a rectal diazepam (aka ‘valium’) and is commercially available at pharmacies. Dosage: based on bodyweight.
- Midazolam can be given as a nasal spray or between the lip and gum (buccally). The nasal spray is prepared by a compounding pharmacy while buccal midazolam is given as a liquid with a syringe. Dosage based on body weight.

2) Should home monitors be used to detect seizures to allow for prompt care?

- There is currently no evidence monitors are needed or will change outcome
- While monitoring may detect convulsive seizures, they probably do not detect all seizures
- False alarms can occur and can be disruptive and anxiety-provoking for caregivers.
- Monitors can help reduce anxiety for some caregivers by providing some peace of mind but they can increase anxiety in others. As the grief of the loss of a child is unique to an individual-so is there coping of siblings with medical concerns. The support of mental health professionals can be helpful in this regard.
- The market on wireless pulse oximetry monitors with alarms, motion sense monitors, seizure monitors and related biometric monitors is exploding with growing technology.
- Consider the type of monitor that would help to safely detect a seizure for the sibling.
- Resource list: www.dannydid.org/sudep/devices-technology/
- Monitor types include, but are not limited to:
 - Wireless Pulse Oximetry monitors with alarms
 - Motion Sensor Seizure Monitors
 - Motion Sensor and other devices that recognize various physiological markers

3) Pediatric Neurologist Consultation

- Given the possible role of FS in some cases of SUDC based on personal and familial occurrence of FS in many cases, consultation with a pediatric neurologist may be considered for the SUDC sibling who had a FS.

- A Pediatric Neurologist consultation can provide a thorough evaluation of the child to confirm the diagnosis of a FS and differentiate the episode from other neurological disorders or a possible cardiac event (which would require a pediatric cardiologist referral-see #5). They can also provide specific clinical recommendations for their care.
- SUDC siblings who have not experienced a FS themselves: Since FS are almost always benign, most pediatric neurologists would not routinely recommend any testing or preventive strategy for a sibling who has never experienced a FS. However, parents may want to discuss a rescue medication if a prolonged FS were to occur.

4) Prepare an emergency plan

- Create a plan and place it in easy viewing (i.e. door of the refrigerator) and a copy in each caregiver's wallet, child's backpack etc.
- The list might include: the names and telephone numbers of who to call when a problem arises, emergency department of preference for EMS, what items to take with the child to the ER (health and insurance card, medical history file, parent's name and contact information, medications, and snack foods that the child prefers to assist during long ER visits.

5) Review the autopsy findings of the child who died and confirm cardiovascular screening of first-degree relatives

- Fever may provoke specific electrocardiographic (EKG) changes leading to ventricular arrhythmias (life-threatening heart rhythm disturbances like Long QT syndrome, Brugada Syndrome and CPVT), often misdiagnosed as seizures. The etiology is thought to relate to the effect of fever on ion channels in the cardiac cell membrane responsible for excitation and relaxation of the cell. Post mortem diagnosis requires a molecular autopsy which is routinely not performed. Therefore, in the absence of any neurological findings on autopsy or clinical review of the deceased sibling, detailed evaluation of first-degree family members (parents and siblings) in a dedicated family cardiovascular genetics clinic is warranted to exclude the possibility of these conditions in other family members.

Related References:

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- Contributing Authors: Orrin Devinsky MD, Laura Crandall, Annapurna Poduri MD, Carol Camfield MD, Gregory L. Holmes MD, Elaine Wirrell MD, and Dominic Abrams MD

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