

Compassionate Allowance Outreach Hearing on Brain Injuries

Social Security Administration

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Good afternoon. I would first like to thank you for the opportunity to speak to you today. My name is Rachel Berger. I am a pediatrician at Children's Hospital of Pittsburgh and an associate director of the Safar Center for Resuscitation Research at the University of Pittsburgh. I will be speaking to you today wearing two hats. First, I am here as a medical expert in pediatric brain injury, specifically inflicted traumatic brain injury, often referred to as shaken baby syndrome. Second, I am here as a physician who specializes in the care of children who have been abused and neglected. In that capacity, I believe it is also my duty to advocate for the medical needs of this vulnerable population.

My goals in the next 10 minutes are therefore

- (1) to use 'shaken baby syndrome' as a paradigm disease to discuss some of the important issues related to diagnosis, evaluation and assessment of disability after pediatric brain injury
- (2) to convince you that injury to a developing brain is very different than injury to a developed brain and that understanding outcome and disability after pediatric brain injury requires that one recognize and understand this
- (3) to discuss why I believe that the diagnosis of 'shaken baby syndrome' should be added to the SSI list of impairments

For the purposes of this testimony, I will define pediatric traumatic brain injury - TBI - as a brain injury which occurs outside of the neonatal period and is the result of trauma. I will therefore not include injuries such as neonatal hypoxia or near-drowning. I will use the term 'shaken baby syndrome' to refer to a specific type of pediatric TBI which occurs as a result of being violently shaken by a caretaker. There are other terms for shaken baby syndrome including abusive head trauma, non-accidental head trauma, and shaken impact syndrome, but I will use the term 'shaken baby syndrome' since it is the term recognized by the public and the term associated with the ICD-9 code for the condition. Shaken baby syndrome is the cause of up to 90% of severe TBI in children under one year of age and the majority of severe TBI in children less than two. It therefore contributes significantly to the morbidity and mortality of TBI in young children and to the number of children who receive SSI in this age-group. As I will discuss, virtually all children with shaken baby syndrome who survive have significant life-long impairment.

In order to understand issues related to diagnosis, evaluation and outcome assessment after pediatric TBI, it is critical to recognize that children are *not* little adults and that injury to a developing brain is very different than injury to a developed brain. There is a misconception among both lay persons and some medical professionals that children with brain injuries have a better recovery than adults. A child with *localized* brain injury such as a stroke is likely to have better recovery than an adult because the uninjured areas of the brain can 'take over' the function of the injured brain. In these cases, young age, even within the pediatric age range is an advantage. In contrast, a child who sustains a *diffuse* brain injury generally has a *worse* prognosis than an adult with a similar injury. When large areas of the brain are injured, there is not enough normal brain to compensate for the injured brain. But perhaps even more importantly, when the brain

is diffusely injured, it affects the ability of the seemingly normal areas of the brain to develop and mature correctly.

The least mature area of the brain in an infant or young child and the part which appears to be most vulnerable to early injury is the prefrontal cortex. This area of the brain is critically important for executive function which is responsible for planning, abstract thinking, rule acquisition, initiating appropriate actions and inhibiting inappropriate actions. *One of the most important predictors of the ability to function in society is the level of executive function.* Executive function emerges in early childhood and continues to mature through adolescence. Since normally developing young children do not have this function, it is not possible to assess it or the extent to which it is impaired as the result of a TBI until they are older. Understanding how brain development affects when injury-related deficits can be assessed is critical for experts who review disability claims for children with TBI. It is also an important consideration when reviewing disability claims for young adults who suffered a pediatric TBI and who initially appeared to have recovered, but who are now unable to function in the work place

I would now like to touch briefly on the issues of neuroimaging and assessment of injury severity. As in adults, head CT is the most common imaging technique in children with TBI. MRI is used as well, though not as frequently in young children because of the sedation risk. Functional MRI (fMRI) and PET scanning, which I know were discussed in this morning's session, are infrequently used after pediatric TBI and age-appropriate norms are just being established in children. These techniques are best for evaluating gross abnormalities and in many pediatric TBI cases, they tell us remarkably little about the true extent of brain injury. The injury sustained in shaken baby syndrome serves as an excellent example of the apparent disconnect between neuroimaging and injury

severity. These children have an injury which is usually a combination of shearing of the axons in the brain, tearing of the bridging veins and lack of oxygen to the brain. Yet, their head CTs are often only mildly abnormal. As I have mentioned, however, virtually all these children are significantly disabled.

The lack of a robust scale to assess injury severity is one of the most problematic issues in pediatric TBI. The Glasgow Coma Scale score, the gold standard severity assessment scale, was developed in adults. In that population and in teenagers it is well correlated with outcome. It is a notoriously poor assessment score, however, in infants and young children. The reason is that assigning a Glasgow Coma Scale score or GCS requires assessment of verbal, motor and eye function. This is simply not possible in children who when not injured, do not yet know how to speak and/or respond to commands. The lack of a strong correlation between the GCS score and outcome in young children is therefore not surprising. Unfortunately, there is no other standardized method of assessing injury severity in young children at this time.

Finally, I would like to touch upon the subject of outcome assessment after pediatric TBI. As you are aware, in order for a child to be labeled disabled, the injury needs to preclude them from performing age appropriate activities for at least 1 year. This definition fits best for children with severe injury for whom it is clear even before hospital discharge that they will have severe lifelong disability. Demonstrating disability is more problematic for children who appear to have recovered or to be only mildly impaired at the time of discharge, but who later begin to fail to develop appropriate milestones and ultimately cannot perform age-appropriate activities. Though disabilities can become apparent at any time after injury, it is more common when new types of demands are being placed on the brain. Typically, these times are when children start school, move from 3<sup>rd</sup> to 4<sup>th</sup>

grade, go through puberty or get their first job. For example, if child has a TBI at age two and injures the part of the brain which is needed to learn to read, then this disability will not be recognized until the child goes to kindergarten and reading becomes an age-appropriate expectation.

Two of the important issues in outcome assessment are when and how to assess outcome. In an ideal world, all children who suffer a brain injury would be assessed soon after injury and then closely tracked by the age-appropriate system (e.g. Alliance for Infants for children <3 yrs, the public school system for older children). A full assessment could then be performed as soon as this tracking suggests that the child is not developing appropriately relative to his or her own previous developmental trajectory. The earlier the recognition of the disability, the more likely it is that therapies can be initiated which may decrease the likelihood that the child will ultimately require SSI. Early documentation of a disability is also important because it makes it easier to link the TBI to the emerging disability which is imperative if the child should ultimately apply for SSI.

For some children, the extent of disability is such that it can be identified on a standard IQ test. In children in whom the disability is more subtle or involves primarily executive function, IQ may be normal. In those children, complete neuropsychological testing is important. Unfortunately this is expensive and rarely covered by medical insurance and the number of qualified pediatric neuropsychologists is very limited. When it is performed, however, it can be extremely helpful in establishing the extent and type of disability and the type of therapy which is likely to be most useful. There are times, however, when even neuropsychological testing does not demonstrate disabilities that are so obvious to the adults and even other children in the child's life. These cases often involve disabilities which are, at least initially, directly related to socialization, a

developmental skill, which can ultimately impact a person's ability to develop and sustain relationships, a critical skill needed to function in society.

Before I end, I would like to explain why I believe shaken baby syndrome should be on the list of SSI impairments. Unlike children with other diseases, children who have been victims of abuse often do not have parents advocating for them and it is incumbent upon others to ensure that society does what is best for these most vulnerable children. Close to 80% children with shaken baby syndrome meet disability criteria by the time of hospital discharge because of cortical blindness, seizure disorder or cerebral palsy. Virtually all children with shaken baby syndrome are ultimately approved for SSI - though this is often only after one or more appeals. In many cases, appeals are never filed after an initial denial. The reasons children with this syndrome have such poor outcome are likely related to two of the issues we have discussed previously: the type of injury, which in this case is diffuse, and the age of children when the injury occurs, which in this case is very young. The only other children with outcome which is comparably poor are very young children who sustain a non-inflicted diffuse brain injury which is complicated by severe brain swelling.

In would like to end by summarizing what I hope you take away from this testimony

- (1) that injuring a developing brain is very different than injuring a developed brain
- (2) that outcome after pediatric TBI is often worse than one might predict based on neuroimaging and the GCS. This is particularly true in children with diffuse injury and those who were very young at the time of injury
- (3) that deficits from pediatric TBI may not be apparent for months or years after injury

(4) that shaken baby syndrome is a unique disease in which the interaction between age and mechanism appears to be particularly toxic to the brain and results in almost universal disability

As a result of the all of the above and in order to best serve the needs of children with TBI, I recommend that serious consideration be given to having a pediatric brain injury expert review the SSI application for children who apply as the result of a TBI. I believe that many of the frustrations for families who go through this process as well as the large number of denials - at least initially - are in large part due to a system which is adult-oriented and which reviews pediatric TBI without the understanding of the issues we have discussed above.

Thank you for your time. I would be happy to answer any questions.