

COMMISSIONER ASTRUE: I hate to break up all the good conversations again, but we're trying to keep everything on time and try to get you all out of here at 4:30. We have another outstanding panel to kick off the afternoon. Our three panelists will be Dr. Rachel Berger, who is associate director of the Safar Center for resuscitation research at the University of Pittsburgh medical center, Greg Ayotte, who's director of consumer services at the brain injury association of America, and Kelly Lang, who is the parent of a child with a traumatic brain injury. And we'll start with Dr. Berger.

RACHEL BERGER, M.D.: Good afternoon. I first want to thank you for the opportunity to speak to you today. As Dr. Astrue said, my name is Rachel Berger. I'm a pediatrician at children's hospital in Pittsburgh and associate director of the Safar center for resuscitation research at the University of Pittsburgh. I'll be speaking to you today wearing two hats. First, I'm here as a medical expert in pediatric brain injuries, specifically inflicted traumatic brain injury, which is often referred to as shaken baby syndrome. And, second, I'm here as a physician who specializes in the care of children who have been abused or neglected. And, in that capacity, I think it's also my duty to advocate for this group of very vulnerable children. My goal in the next ten minutes are first to 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 injuries. Second, 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 you recognize and understand this. And, third, to discuss why I think that the diagnosis of shaken baby syndrome should be added to the SSI list of impairments. For the purposes of this testimony, I'm going to define pediatric traumatic brain injury, TBI, as a brain injury which occurs outside of the neonatal period and is due to trauma. So I'm not going to include injuries such as neonatal hypoxia or near drowning. And I'm going to use the term shaken baby syndrome to refer to a very specific type of pediatric brain injury, which occurs as result of being violently shaken by a caretaker. There are many other terms for shaken baby syndrome, such as abusive head trauma, nonaccidental head trauma, shaken impact syndrome but I chose to use the term shaken baby syndrome because I think that's what the lay public recognizes and it's a term that's associated with the ICD 9 code. Shaken baby syndrome causes up to 90% of the severe brain injury in children less than one and the majority of severe brain injury in children less than two. And it, therefore, contributes very significantly to morbidity and mortality of the traumatic brain injury in young children and to the number of children who receive SSI for brain injury in this age group. As I will discuss, virtually all children with shaken baby who survive have significant long-term impairment. In order to understand issues related to diagnosis, evaluation and outcome assessment after pediatric brain injury, it's critical to recognize that children are not little adults And that injury to a developing brain is very different than injuring a developed brain. There is a misconception among both lay people and some medical professionals that children with brain injury have better recovery than adults. And, indeed, a child with a 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 area. And in these cases, young age even within the pediatric age range is an advantage. In contrast, however, a child who sustains a diffuse brain injury generally has a worse prognosis than an adult with a similar injury.

Because when large areas of the brain are injured, there is not enough normal brain to take over or compensate for the injured brain. And perhaps even more importantly, when the brain is diffusely injured, it affects the ability of the seemingly normal part of the brain to develop and mature correctly. The least mature area of the brain in an infant or child and the part which appears to be most vulnerable to early injury is the pre-frontal cortex. This area of the brain, as you know from this morning, is critically important for executive function. And that's important for planning, abstract thinking, rule acquisition, initiating appropriate actions and probably most importantly, inhibiting inappropriate actions. One of the most important predictors of the ability to function in society is the level of executive function. An executive function emerges in early childhood and continues to mature through adolescence. Since normal developing children do not have executive function in the young ages, it is not possible to assess the executive function or the extent to which it is impaired as a result of brain injury until children 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 brain injury. It is also an important consideration reviewing a disability claim for a young adult who suffered a pediatric brain injury and who initially appears to have recovered but who then is unable to function in the workplace. I want to touch briefly on the issues of neuro imaging and assessment of injury severity. As an adult, head ct is the most common imaging technique in children with brain injury. MRI is used as well, although not as frequently in young children because of the issues of sedation risk. [inaudible] or pet scanning, which I know were discussed this morning, are very infrequently used in children and even the standards are not well established yet. CT and MRI are best for evaluating gross abnormalities. But in many pediatric cases, they tell us remarkably little about the true extent of injury. The injuries sustained in shaken baby syndrome serves as an excellent example of the apparent disconnect between neuro imaging and injury severity. These children have an injury which is usually a combination of shearing of the axons of the brain, tearing of the bridging vein and a lack of oxygen to the brain. Yet, their head CT's are often only mildly abnormal. And as I mentioned, virtually all of these children end up being significantly disabled. The lack of a robust scale to assess injury severity is one of the most problematic issues in pediatric brain injury. The GCS score, physical standard, as you discussed this morning, was developed in adults. And in that population, it's very well-correlated with outcome. But it is notoriously poor outcome assessment in infants and young children. The reason is that assigning a GCS requires verbal, motor and eye function. And it's certainly not possible in a child who does not yet know how to speak or cannot even respond to commands. The lack of a strong correlation between GCS and outcome in young children is, therefore, not surprising. But, unfortunately, there's no other standardized method of assessing injury severity in young children at this time. Finally, I want to touch on the issue of outcome assessment. As you are aware, for a child to be labeled disabled, the injury needs to preclude them from performing age appropriate activities for at least a year. This definition sits well for children with severe brain injuries for whom it's clear even before hospital discharge that they will have severe disability. Demonstrating disability is more problematic for children who appear to have recovered or to be only mildly impaired at discharge but then who later fail to develop milestones and who ultimately cannot perform their age appropriate activities. Though disabilities can occur at any time or become apparent any

time after injury, it's more common when new types of demands are placed on the brain. Typically, these are times such as when the child starts school. When they move from the 3rd to 4th grade, when they go through puberty or when they get their first job or need to drive. For example, if a child gets a brain injury at age two and injures the part of the brain which is required for reading, disability cannot be recognized until the child goes to kindergarten and reading becomes an age appropriate activity. Two of the important issues about outcome assessment are when and how to assess it. In an ideal world, all children who have a brain injury would be assessed soon after injury and "closely tracked" by the age appropriate system. A full assessment can then be performed as soon as the tracking suggests 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 therapy can be initiated, which may decrease the likelihood of requiring SSI in the future. Early documentation of any disability is also important because it makes it easier to link the brain injury to the emerging disability if at a later time the child does require SSI. For some children, the extent of disability can be identified on a standard IQ test. In children in whom it's more subtle or involves primarily executive function, IQ may be normal. In those children, complete neuropsychological testing is important. Unfortunately, this is expensive, rarely covered by medical insurance and the number of qualified pediatric neuropsychologists is limited. When it is performed, however, it can be extremely helpful in establishing the extent and the type of disability and the type of therapy which will be most helpful. There are times; however, that even neuropsychological testing does not demonstrate disabilities that are so obvious to the adults and even to the other children in the child's life. These cases also 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, which is critical to life. Before I end, I want to explain why I believe that 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. It's incumbent upon others to ensure that society does what is best for these most vulnerable children. Close to 80% of children with shaken baby meet disability criteria by the time of hospital discharge because of cortical blindness, seizure disorder or cerebral palsy. Virtually all children with shaken baby are ultimately approved for SSI, though this is only often after one or more appeals. And in many cases for various reasons, appeals are never filed for these children. The reasons the children with this syndrome have such poor outcomes are related to the main issues we have discussed: Type of injury, which in this case is diffuse, and the age of the children in which the injury occurs, which in this case is very young. The only other children with comparable outcomes are very young children with a nonafflicted diffuse brain injury, complicated by severe brain swelling. I'd like to summarize what I'd like you to take away from this testimony. First, injuring a developing brain is very different than injuring a developed brain. Two, that outcome after pediatric traumatic brain injury is often worse than you might expect based on neuro imaging and a Glasgow coma scale score, which is the way we can assess injury severity. And this is particularly true in children with diffuse injury and those who are very young at the time of the injury. Three, the deficits for pediatric brain injury may not be apparent for months or even years after the injury. And, finally, that shaken baby syndrome is a unique disease in which the interaction

between each mechanism appears to be particularly toxic to the brain and results in almost universal disability. As a result of all of the above, in order to best serve the needs of children with brain injury, I would recommend that serious consideration be given to having a pediatric brain injury expert review the SSI applications for children who apply as a result of 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 brain injury without the understanding of the issues we have just discussed. Thank you for your time.

COMMISSIONER ASTRUE: Thank you. Greg.

GREG AYOTTE: Thank you. Good afternoon. My name is Greg Ayotte and I'm the director of consumer services for the brain injury association of America. The brain injury association of America was founded in 1980 by individuals who wanted to improve the quality of life for individuals who had sustained brain injuries and their families. Today, the association encompasses a nationwide network of more than 40 state affiliates sharing in the mission of creating a better future through brain injury prevention, research, education and advocacy. On behalf of BIAA, its nationwide network of affiliates and the millions of Americans affected by brain injury, I want to express sincere appreciation for the opportunity to speak before you today. The brain injury association applauds the compassionate allowance initiative and the social security administration's desire to identify information to help better determine when a child or adult with a brain injury is most likely to meet the SSA disability standard. The brain injury association also applauds the openness of the administration in proactively seeking information about how its disability determination process can be improved for individuals with brain injury. According to the CDC, each year at least 1.5 million civilians sustain a TBI and TBI is the leading cause of death and disability in children and young adults. Males are twice as likely to sustain a brain injury as females. But people of every age, race, gender and socioeconomic status are at risk for brain injuries. Brain injuries are caused by falls, motor vehicle crashes, assaults, sports and recreation accidents and most recently blasts from IED's. The RAND corporation, center for military health policy, estimates that 320,000 members of the U.S. armed forces has sustained a brain injury while serving in Iraq and Afghanistan. I know that the earlier session this morning devoted special attention to this specific population of heroic men and women. All in all, with a new injury occurring every 23 seconds in the United States, TBI clearly ranks as a public health crisis. Brain injuries can lead to physical, cognitive and psychosocial behavioral impairments ranging from balance and coordination problems to loss of hearing, vision or speech. Fatigue, memory loss, concentration difficulty, anxiety, depression, impulsivity and impaired judgment are also common following a brain injury. The even so-called mild injuries can have devastating functional consequences that require intensive treatment and long-term care. BIAA state affiliates regularly work with individuals with brain injury who encounter obstacles in trying to access important public programs, including social security disability benefits. Obviously, it would be impossible for me to recount every type of challenge that these individuals have faced. For this reason and because these voices are so important to hear, the brain injury association has circulated news of today's hearing to all affiliates and is actively encouraging individuals

to submit their personal accounts into form of written testimony to the social security administration. I would, though, like to highlight a few themes which have emerged as the association heard from and worked with individuals with brain injuries.

COMMISSIONER ASTRUE: Greg, could I ask you to move just a little closer to the mic? I think I'm picking up a little sign in the back row they're having trouble picking up.

GREG AYOTTE: Sorry about that. Closer? I'm sorry. All right. I would like to highlight a few themes which have emerged as BIAA has heard from and worked with individuals with brain injuries who are struggling to gain access to social security disability benefits. As you know, social security determinations rely in large part on medical information. Perhaps the biggest challenge when it comes to medically addressing brain injury is the fact that the brain is infinitely more complex than any other organ or organ system in the human body. Add to that the fact that every brain and every brain injury is unique and it's easy to understand why it's often said that there is much more that is unknown about the brain and brain injury than is known. This unknown surely plays a complicating role in social security disability determination process in cases of brain injury. However, even if we await further advances in diagnostic and prognostic technology, a substantial amount of expertise in identifying and treating brain injury has developed within the past few decades. It is critically important that clinicians and other professionals involved in social security disability determination processes in cases of brain injury have specialized experience in brain injury. As one brain injury professional recently noted to the brain injury association, it has been my experience that without proper training as to how to read the pattern of results obtained through an assessment, patients are often undiagnosed or misdiagnosed as to the severity and nature of specific deficits. Similarly, it would be ideal if all social security examiners making disability determinations in brain injury cases had training resulting in strong working knowledge of the unique aspects of brain injury. Many individuals with substantial disability resulting from brain injury have reported that their application for disability benefits was rejected and that they were told they should be able to hold down some kind of job even after they detailed and documented extensive evidence of impairment. Many of the characteristics of brain injury impairments are not obvious or familiar to the general public, likely including social security examiners. Such characteristics include balance problems, car sickness, sensitivity to bright lights and visual stimuli, irritability, fatigue, sensory overload related to difficulty filtering out background noise, inability to initiate or to follow through on tasks and quite commonly a lack of self-awareness of one's own cognitive deficits. It is important that those making the determination truly understand the impact that these various impairments have on the lives of individuals with brain injury, as well as the implications such impairments can have for employability and the ability to participate in the workforce. Several brain injury support professionals and case managers communicated to the brain injury association that the individuals with brain injury that they were working with would not have been able to complete the paperwork necessary to apply for benefits without their assistance. This poses a difficult problem in terms of bureaucratic hurdles, but it is the reality for many individuals with brain injury. One individual with a brain injury noted that the lawyer

asked me to bring records of some events. He did not consider that record keeping as one of my most difficult tasks and I am unlikely to find most of the records he requested within the next two days. Another brain injury survivor likened asking a person with injury-related cognitive impairments to navigate the social security disability benefits process to asking a person with a badly broken leg to run to get help, except that the person with the damaged leg would know that they couldn't run, which is more than many people with brain injury may be able to do. One case manager stated the process of applying for disability benefits with the social security administration for patients with brain injury needs to be refined to consider the cognitive deficits more and to expedite the approval and appeals process. One theme above all is quite clear for both children and adults, obtaining social security benefits serves as a gateway facilitating access to additional much needed healthcare and support services. The brain injury association of America looks forward to continuing to work with the social security administration to making the social security disability system more accessible to individuals with brain injury who meet the eligibility criteria. Thanks.

COMMISSIONER ASTRUE: Thank you, Greg. Kelly.

KELLY LANG: Thank you. Good afternoon. I'm Kelly Lang, and I would like to thank you for allowing me to tell you about my family's introduction into the world of pediatric brain injury. Seven years ago this month I was driving with two of my daughters to my oldest daughter Hanna's nutcracker rehearsal. The last thing I remember is pulling out of my driveway. I can describe the accident only through witness testimony. We approached a traffic light at an intersection. As the light was turning yellow, I slowed to stop. At the same time, another car hit two others behind me. Then hit me and pushed us 60 feet through an intersection until we came to a guardrail. The first thing I remember is hearing my five-year-old Hanna yell, Mommy, Mommy, wake up. The next thing was an EMT asking me if there was anyone sitting in the third row of my minivan. Luckily, there was not. However, I instantly knew there was something wrong with my three-year-old Olivia. They asked me how old she was and I started screaming at them to do something to save her. She was sitting directly behind me in a five point harness car seat. The three of us were taken in separate ambulances to Loudoun hospital center and immediately after arriving the doctors decided they could not treat Olivia since the facility was not an equipped trauma center. She was immediately transferred to Fairfax hospital. Hanna was discharged with the advice to give her Tylenol if she had any back pain. She went home with a friend. Olivia was transported to Fairfax while I had a ct and x-rays. Upon arriving at Fairfax, Olivia was in the pediatric intensive care unit and was not responding to anything. My husband, Michael, had arrived immediately after I did. The neurosurgeon informed us that there was bleeding and swelling in Olivia's brain and they put in an intracranial pressure monitor into her skull. We were told that we should not touch her or try to over stimulate her since the pressure of her brain was being closely monitored and if the pressure escalated too high, she would have to have brain surgery. Luckily the pressure did decrease and the monitor as well as the c-spine collar was removed after the 8th day in the hospital. When my husband and I began looking into inpatient rehabilitation facilities for Olivia, she remained in the pediatric intensive care unit and had not opened her eyes until this point. She was then transferred to Kennedy Krieger

institute in Baltimore after two weeks of being at Fairfax. Once we arrived at Kennedy Krieger, Olivia really began to show some improvement. As soon as we arrived in her room and placed her on the bed, she rolled over for the very first time. Her rehabilitation team originally predicted that she would be in rehab for three to six months. She was discharged on December 29th, 2001, exactly two weeks after we arrived. During our stay, we worked with occupational therapists, physical therapists, speech pathologists and recreational therapists. She regained mobility and worked to regain some of her fine motor and speech abilities. Upon discharge, we received in-home physical therapy for one month. At the end of January, two months post-accident, Olivia asked when she could go back to preschool. Prior, she had attended a preschool two mornings a week for three hours a day. However, after the accident, she could only attend for two hours two days a week due to her fatigue. She was on a wait list for private speech and occupational therapy and at the same time we had an individualized educational plan eligibility hearing through the school program and learned that she was eligible for services. No big surprise. Her services with the public school system began in April of 2002 and simultaneously we started private speech therapy two times per week in addition to two mornings of private preschool, two mornings of special education preschool where she also received physical therapy, occupational therapy and speech services. Olivia has made great strides through her speech therapy. She continued with private speech for three years and still receives speech therapy at school. It took a year and a half until we were able to find a private occupational therapist, and she remained in therapy for three years. She no longer receives occupational therapy through the school system even though her neuropsychological reports have continued to recommend it. She recently began therapeutic writing this year after being on a wait list for over two years. Although Olivia appears to be a very well – appears to be a normal well-adjusted ten-year-old, she has significant cognitive difficulties. She has an individualized educational plan that outlines services the school system provides, such as one-on-one instruction, extra response time, small group testing. Olivia has also struggled with anxiety since her injury. She's afraid of the dark, balloons, loud noises and enclosed spaces, such as movie theaters. These anxieties have prevented her from participating in many activities during the past seven years. She also has some social anxieties and at times has trouble communicating with her peers, because she's not able to keep up with the conversation or the game that is currently being played. Olivia has made significant gains since the day of our accident seven years ago. In fact, her neurologist has commented that she is a miracle case. When he viewed her MRI, he commented that based on the injuries she sustained; she should not be walking or talking. We are hopeful that Olivia will continue to improve and be an inspiration for others. Thank you for allowing me this time to share her story.

COMMISSIONER ASTRUE: Thank you very much. I'd like to start with a couple questions on shaken baby syndrome. I'm not sure -- maybe David can tell me for sure. We're playing catch up on a lot of the updating of our regulations. We're trying now to make sure we're updated every five years. But this area of the regulation I think it's probably been a couple decades. I'm not even sure shaken baby syndrome was recognized the last time we did a systematic review in this area. So help us with some things that probably you consider very basic. What would you consider to be really the threshold

medical observations for a diagnosis of shaken baby syndrome? What's the minimum that you need to have, in your view, in order to have a diagnosis of shaken baby syndrome?

RACHEL BERGER, M.D.: Well, that's a hard question to answer. Insofar as -- we used to think, for example, that you needed to have sub Ural hemorrhages or you needed to have an abnormal ct. But now there are many, many cases, in fact we've had multiple in our own hospital, where the head ct's and even the brain MRI's are normal, but the children have retinal hemorrhages or fractures and someone admits to having shaken them. So, in those children, it appears that the threshold to have retinal hemorrhages, which is bleeding in the eye, for that child is lower than the threshold to have a brain injury. But I say that for someone to make the diagnosis of shaken baby syndrome, probably only occurs in half if not fewer of the few cases because the stigma attached to making the diagnosis and the medical-legal implications of the diagnosis are so significant.

COMMISSIONER ASTRUE: I'm going to get to that in a second. That's my other question.

RACHEL BERGER, M.D.: You wouldn't even make it unless you probably met a much higher threshold than you actually need. So I think there's no one -- you don't have to say you have to have brain injury, you have to have retinal hemorrhages. There's nothing you need to have except for probably an expert to say, this constellation of symptoms in light of the history that the person is providing us about how it happened are not consistent. And that's probably the best definition that the history does not match the physical exam findings.

COMMISSIONER ASTRUE: So let me ask you about some of the complications. I mean, generally, our self identification is a social insurance agency. So part of our culture is we tend to want to take in facts, analyze facts, and then send a check when appropriate. We're not a traditional social service agency, but the world is more complicated than that. So one of the difficult judgments we often have to make is when it's appropriate to point what we call a representative payee, a third person who controls the benefit for the individual. We often do that in cases where -- we do that for homeless people. We do that for people with addiction problems if they've qualified for some other reason, because we're not allowed to give benefits just on the basis of addiction. In these shaken baby cases, I imagine there's some cases where it's been a caretaker or someone other than the parents that are responsible for the abuse. But when it is one of the parents that is responsible for the abuse, should we go operate, for instance, on just a presumption that we should come in with a rep payee? If one of the parents is responsible for shaken baby syndrome, should we assume that they can't manage the child's benefit for the child's good?

RACHEL BERGER, M.D.: That's a good question. But almost all these cases where the parent is one of the perpetrators or a perpetrator by omission, meaning that it was perhaps a significant other and this person didn't protect them, usually the person applying for the social security is actually the foster care agency and not the parents. So, for these

children, either the medical rights have been terminated or the child is in foster care, in which case the person applying is actually a soc and as you know, there's lots of rules about what you do for a child in foster care who receives SSI. But to make a very long story short, it allows for the social service agency to place the child in a higher level of foster care home such that there are fewer children there, the person has more specialized medical care so that the child gets additional care through that system because of the other system. The parent almost never is the -- if they're the perpetrator or the perpetrator by omission, they are virtually never the person applying or collecting.

COMMISSIONER ASTRUE: Okay. That's helpful. Thank you.: Someone else?

DAVID RUST: I guess for Dr. Berger, I'd go back to that same question again. And that is, when we get a case like this, how do we know -- in other words, the concept of the compassion allowance is that it is with the correct medical evidence you can make a quick decision, that you can make a decision much more quickly than we normally would. I guess the question -- and it seemed to me your answer to the commissioner was that there is no single set of tests or there is no single way to quantify the degree of the damage to the brain that would allow us to make that decision?

RACHEL BERGER, M.D.: You're asking specifically in shaken baby syndrome or pediatric brain injury?

DAVID RUST: Can you pull the microphone closer?

RACHEL BERGER, M.D.: What did you say?

DAVID RUST: Can you pull the microphone a little closer?

RACHEL BERGER, M.D.: Specifically in shaken baby syndrome or pediatric brain injury in general are you asking?

DAVID RUST: I'd start with shaken baby.

RACHEL BERGER, M.D.: So in shaken baby syndrome, I would say that it's a medical diagnosis, it's a clinical diagnosis. So if children don't qualify -- say if your child was among the most severe cases, presumably you will be labeled with that condition. And so it will be clear that way. If you're not labeled with the condition, the way it is now is children will become eligible because they're cortically blind, they have cerebral palsy, or they have a severe seizure disorder, or they end up being mentally retarded, developmental delay, some other reason. So that's usually how they're qualifying in that way, as opposed to -- because of the actual term, shaken baby syndrome. In terms of other children where that's not the medical diagnosis, I think it becomes very difficult because of the difficulty in giving an assessment at injury. There are many children with a Glasgow coma scale of three who actually turn out to be quite relatively functional and don't end up getting SSI and, Likewise, quite a few children with a GCS score of 13 to 15 who end up being far more disabled than you would ever expect. So because we can't

really do a good job assessing by the Glasgow coma scale score, I think it becomes harder, on the social security end, to have a cut point because that assessment is not very sensitive or specific. And likewise, even with a ct scan, diffuse cerebral edema is probably the only sign I would say that at least is very well correlated with a poor outcome. But even in the same way, there's a fairly significant overlap between children who have diffuse brain injury but turn out to be fairly functional and children who don't have diffuse brain injury that turn out not to be functional. I think it's probably one of the areas where pediatrics becomes much more complicated than adults, because we can't assess well initially, we can't use neuroimaging. And so the outcome, as the neurosurgeon that I work with, Dr. Allison, always says, I hate saying this to parents but I've been doing this for 20 years and I'm really not much better than I was 20 years ago at telling parents how this child is going to be in five years. I just think it's a really difficult question in pediatrics.

DAVID RUST: You mentioned a couple of times that damage to a developing brain is more severe and more permanent than damage to a developed brain.

RACHEL BERGER, M.D.: It's just different because if you haven't developed – we always say, children going into a brain injury, they come in for example a six month old being able to roll or being able to babble and with no other function. And as you injure the rest of the -- diffusely injure the brain, there wasn't very much there before that the child can recover. And so that's part of the problem. Even things that looked normal, particularly the prefrontal cortex, it looks normal on neuroimaging. But then the child cannot inhibit any functions. And they get to school and none of the kids want to play with them because they still, you know, yell things like, you're stupid, to another child or they can't hold their place in line. All of a sudden, it becomes a huge social issue even though it seemingly, when they were injured, that part of the brain was normal. And so it's just very difficult because the brain itself has not developed, the wiring has not taken place. And somehow the brain just is not able to properly wire once you have that diffuse brain injury.

COMMISSIONER ASTRUE: I apologize for the other two witnesses for focusing on Dr. Berger, but I think we're trying to sort a couple of things out to try to figure out how to fix things. A couple of questions. Is there agreement in the medical literature about how many cases of shaken baby syndrome there are each year? And can you also talk to me, again, definitionally -- I'm trying to figure out how to give my -- I suspect that what we've done historically is not looked at it as a syndrome but looked symptomatically and tried to just evaluate by symptom. That's not uncommon in the agency. Is there an upper age on shaken baby syndrome? What's right above that? And what looks like -- I'm just trying to figure out some of the definitional issues and how we might handle some of these things.

RACHEL BERGER, M.D.: That's a really interesting question. The answer is, you can be a shaken baby at any age. And, in fact, there's some great published reports with the Israeli army that used to use it as a form of torture. You can have an adult who looks exactly like a shaken baby for all intents and purposes. If you shake an adult through a

form of torture, they will get retinal hemorrhages, they will get brain swelling, they will get subdurals, and they can die. That said, most of the cases are less than a year of age. We have certainly had cases up to five or six years of age. It's just that it becomes more difficult to physically shake a child who is heavier.

COMMISSIONER ASTRUE: And is there anything from -- you have a general sense of what we try to do and why. Is there any dramatic difference in these cases between age one and younger in the older children?

RACHEL BERGER, M.D.: That's a great question. In fact the answer is when we do see older kids with shaken baby; they do seem to recover better than the younger children. And that is pointing, I think, to the issue of age. If the children talked before and walked before, they do tend to do better, although they clearly are still significantly disabled. But they are much less likely to be vegetative. I think there are so few cases of the four, five, six year olds, relative to the infants, that I think it's hard to say that for sure. But clearly, anecdotally and in our experience, at least at UPMC, the older children with shaken baby do, I'd say, have less severe disability than the younger children.

COMMISSIONER ASTRUE: And just one more follow up. The incidence rate, do we have any sense of what we think it is?

RACHEL BERGER, M.D.: Actually, there's two really good population-based studies. In the United States, it's about 1 in 3300 per case of severe or fatal in children less than one. So if you assume there's four million births a year, then you would say there's probably about 1500 cases in kids less than one. But about a third up to half of those kids die. So from an SSI perspective --

COMMISSIONER ASTRUE: It's a relatively small percentage of cases. And there are going to be a certain percentage that aren't going to qualify for SSI for other reasons too.

RACHEL BERGER, M.D.: Right, exactly.

COMMISSIONER ASTRUE: So it's a relatively Small --.

RACHEL BERGER, M.D.: Right. And then between ages one and two, it drops off significantly. And then after age two, there really aren't any incidents figures because it's more we see the cases once or twice a year. They're really rare.

COMMISSIONER ASTRUE: Okay. That's helpful. I'm sorry for hogging the forum. I'll give my colleagues a chance.

WALTER KOROSHETZ, M.D.: This is for Greg. I was just going to ask -- you know, this morning we were talking about these different time periods and the -- I just wondered. The issues that you've brought up in terms of people negotiating the system and having trouble, do you think that that's paramount in people in the first year after injury? Or is that something you think is a lifelong problem.

GREG AYOTTE: That's a good question. I was actually talking with a person the other day from North Carolina who was four years post injury who was just applying for disability and a lot had to do with the fact that for the four years after she didn't have access to good care, didn't have access to good testing. She couldn't realize the changes that were going on, and she wasn't understanding the feedback she was getting. So I would say, even at four years, she was just starting to apply. And I know she was going to have issues cognitively with kind of figuring out getting medical records, getting assessments and following through. So I would say no, it's not necessarily just in that first year. It definitely could be longer post injury where those issues can present. I want to kind of add that point, that time comment too with the pediatric injury. We get a lot of phone calls from folks who, you know, the child is injured, they go to rehab, they go home, and it's not until 12, 14, sometimes two years post injury where you really start to see the impact of the injury, especially with the frontal lobe injury with a younger child where you may not see the kind of academic and kind of cognitive effect of that injury until they get to middle school when you really have to start using some abstraction. And all of a sudden, this kid who was kind of getting through school just plummets. So you have to kind of -- sometimes, I think, we hear that sometimes from parents when they decide to finally apply for SSI. They're getting denied in part because it's three years between the time of the injury and the time that you're applying. That's how pediatric injuries present.

COMMISSIONER ASTRUE: That's a valid point. That may be -- I'm not going to pretend that I can immediately recall our exact standards in this area, but it would be -- you can understand, looking from an examiner's point of view, that in most cases they're conditioned that if there's a big time gap between what appears to be the injury and what appears to be the symptom, there's a presumption against causation. And that's right probably for the vast majority of diseases and conditions. But here, I think that it does seem to be pretty common and I don't think there's very much doubt that, you know, you might not see the symptoms for a long period of time. And that might be something that, you know, would bear some clarification on our part for the people that are making the decisions in the field. But I'll have to go back and check exactly what the instruction says.

NANCY GRISWOLD: In older children, I know you said

COMMISSIONER ASTRUE: -- Can you get a little closer to the mic? I think they can't hear you in the back. NANCY GRISWOLD: I know you said a moment ago that mostly these seem to appear in children, I think you said, under one year. But in older children, is there any kind of constellation of symptoms or anything diagnostic that you look for that relates back or might relate back to a shaken baby incident that perhaps went undetected?

RACHEL BERGER, M.D.: That's a great question. And I have to say, as early as the early 1970s, Kathy, the person who first described shaken baby made the statement that I wonder if some of the mental retardation cases in older kids were actually missed cases

of shaken baby. And I have to say, as a pediatrician who used to run a foster care clinic, the rate of mental retardation in the siblings of abused children is remarkably high. And it's hard not to think, was that a missed case. If you go over time though, the findings on MRI are so nonspecific later on, if brain Atrophied, anything that it would be really hard to go back and make the diagnosis retrospectively. But presumably, the children would still qualify under other criteria such as mental retardation or seizure disorder or cerebral palsy. So I think it's very hard to go back. If the diagnosis isn't given at the time, I think it would be almost impossible at a later date to say, you know, three years ago this was a shaken baby.

NANCY GRISWOLD: So you're really looking at a contemporaneous kind of a diagnosis.

RACHEL BERGER, M.D.: Right. It's a clinical diagnosis, probably an underused diagnosis. But it really is -- at this point, we've learned more and more. There's no single finding that you can say this is a shaken baby. It's more that there's a constellation of findings. Probably, most importantly, that the history the parent is giving you, such as, nothing happened, and what you're seeing on ct just don't match. A parent who says the child was fine and started vomiting and has huge subdural and retinal hemorrhages, those two clearly don't match. If there was a car accident and then you see the same thing, that's not a shaken baby because now the two match. So it's really still a clinical diagnosis.

WALTER KOROSHETZ, M.D.: So I was going to ask, in terms of your daughter, what do you think are the things that you could put your finger on that are most beneficial in trying to get her recovery going?

KELLY LANG: I think the most beneficial thing was how much therapy we put her into immediately. Even though we had a two-year wait for occupational therapy that we pushed -- we got her into speech therapy immediately and kept that going. And even then, her neuropsych reports were coming back abysmal. It was very upsetting to see that, but I think that has really helped her. And the thing that is a little bit different with her is that a lot of children will -- their behavior will show that there's something wrong, and she's the opposite. There's no behavior issues. In fact, she'd rather, you know, fall back to the back of the room and disappear. She wouldn't let anyone know there's something wrong. So we have to keep on top of what is going on with her.

LYNDA DAVIS PhD.: Do you expect her condition to stabilize or is she getting better or worse?

KELLY LANG: That's hard to say, because Usually she'll take two steps forward in one area and then she'll fall back two steps in another area. It becomes cyclical. The doctors have always said we can't tell you what her prognosis is. It's just a wait and see. Now, she is in 4th grade now and I see things are starting to slide down a little bit academically and cognitively. I mean, she's still not reading well. That's been a very difficult thing for her. Executive functioning is very badly damaged. So it's just -- we just take one day or one year at a time.

COMMISSIONER ASTRUE: Greg, do you have any sense, for the whole universe of the pediatric cases, how many TBI's we'd be talking about generally? The incident rate, order of magnitude?

GREG AYOTTE: Can you say that again?

COMMISSIONER ASTRUE: How many pediatric brain injury cases do you think we probably see in the country each year? Do you have any sense generally? We got a rough number on the shaken baby syndrome, but you have accidents, car accidents, other forms of injury. Do you have any sense for how many cases we're talking about generally?

GREG AYOTTE: For pediatric injury, there's less data available. The CDC is probably the best kind of data source, and even their stuff doesn't really flesh out. There's another group, the traumatic brain injury model systems, which really kind of tries to get some idea of the scope of brain injury and outcome, but they just do 16 and up. So there's that big gap. I would say the best estimate is probably around a million a year is what I've read, and That's based on CDC numbers. That's kind of extrapolating from smaller groups so it's hard to gauge the accuracy of it.

COMMISSIONER ASTRUE: Okay.

NANCY GRISWOLD: I think that what I heard you saying was that there may be a later onset of some symptoms or recognition of some symptoms in these cases because the brain is developing and then when a skill is needed, perhaps that part of the brain has been damaged in some way and then the skill simply isn't there. So I'm curious to know, how far into the developmental stage do you expect to keep seeing symptoms of one sort or another appear?

RACHEL BERGER, M.D.: I'm sorry, I didn't hear the ending.

NANCY GRISWOLD: How far into development could you expect to see some sort of deficiencies or lack beginning to appear, some sort of symptoms?

RACHEL BERGER, M.D.: I think after you have an injury, pretty much all the way through -- it depends where you end. Pediatric kind of depends if you end at 15 or you end at 18. But really, the frontal lobes develop until age 21, so the Full brain has not developed until you're almost 21 years old. So I think any deficiencies that occur at the time of brain injury can almost be recognized at any point as new -- basically new different types of challenges the brain needs to perform. I think, as you stated also, reading is probably the best example because you can't assess reading in a two year old. You cannot assess it until the child basically falls behind. Until they fall behind. But at other points, the third to fourth grade turns out to be a really big problem because For the first time they can't copy an answer. They're actually starting to do some abstract thinking. And for some kids, that's where everything falls apart. You know, for other children, it's where the peer relationships change, when they're going into middle school,

around then. I mean, all children are awkward at that time. But for these children, they're not developing executive function anything like the peers and things just fall apart. So I think there's not a certain time. And probably the earlier the injury, you could say there's even many, many, many years. The other thing is how well you're assessing it along the way. I mean, you're the parent who is doing multiple neuropsychological testing. You have children who have no access to that testing. And it's not until literally they test with an IQ of 70 on an IQ test that anyone notices there's a problem. But there probably have been problems all along. It just doesn't get noticed until the IQ test six years after the brain injury. And I think that becomes very problematic for the system because causation becomes very, very difficult which is why I said I think, ideally -- there's tracking available. Lyons, for instance, tracks anyone up to age three in the United States. We don't use it. But if you track children along the way, it would be much clearer when something develops or emerges. And then, of course, you could also treat it. But I think that's one of the biggest issues is, you know, how long after and how do you link it back, how do you prove causation.

NANCY GRISWOLD: So problems with age-appropriate functioning might become more severe with age –

RACHEL BERGER, M.D.: Exactly.

NANCY GRISWOLD:-- related to this.

RACHEL BERGER, M.D.: And I think it is. The one year old doesn't need to do very much. And so if the one year old doesn't develop speech, for example, until 20 months, that's a little behind the curve but not terribly. But if your third grader still doesn't read, everything after that is dependent on everything before and things fall apart. So I think that usually it does get worse as time goes on, not better. The question is just when does someone recognize this child has a problem. And it can be remarkably late. I had a child in my office not that long ago in the foster care system who came in because he was having all these behavior problems. And I pulled his chart and they sent me volume 8 of 8. Why does he have eight volumes? The kid had been near drowning. He had been in our ICU for almost three months and nobody -- he was in foster care. Nobody caring for him realized that for three months he had been in the ICU with a brain injury. Clearly, these were related, but it had been years since the injury. So I think it seems obvious when you have parents that are on top of it, but I think it can go for a long period of time when the resources aren't there and the parents don't realize that this is the link.

NANCY GRISWOLD: Thank you.

DAVID RUST: If we were going to look at something from a policy point of view, shaken baby is a particular definition of a particular type of injury deliberately inflicted. But are there – I mean, is there more generic pediatric brain injury that would be very similar that we should look at more broadly or should we restrict it to this one area?

RACHEL BERGER, M.D.: That's a really hard question. I was thinking a lot about that. Is there any other thing that's equivalent to the shaken baby syndrome? I don't think there is. The only thing I could think of was young children, very young children with diffuse brain injuries which are complicated by severe cerebral edema. Generally, those children have a terrible outcome. And that's the only other group. That's not a diagnosis. That's a constellation of their increased intracranial pressure, and there are clinical problems with that. I can't think of any other single syndrome where you could say almost universally this child doesn't do well. I think it may be that we're not doing a good job at the beginning diagnosing or understanding how severe the actual injuries are. And if we had a better way of assessing them, other than the Glasgow coma scale score, it might become clear there is a way to predict. But I think at this point we don't have a way to do that.

WALTER KOROSHETZ, M.D.: Why would you separate out, you know, the car accident from the shaken baby in terms of SSA's business? Aren't they really variations on the same theme?

RACHEL BERGER, M.D.: The mechanism of a car accident is usually very different. The reason is that usually a car accident, first of all, is a one-time event. And many times shaken babies, at least half the time, have evidence of prior brain injury. And as everyone knows, the worst thing you can do to a brain is reinjure it. So one of the other reasons for bad outcome may be that they're injured before. And usually it's a single impact in a car accident. Even though it's an acceleration/deceleration, it's clearly a different injury. And its degree of hypoxia, for example, which is very bad for the developing brain, is much worse in the shaken babies. And there clearly is something different insofar as in motor vehicle crashes very few kids have retinal hemorrhages, whereas a significant proportion of the shaken babies do. So there is something very different about those two mechanisms of injury. And it's probably a combination of age, the actual mechanism, how many times it happened, how quickly they come to care. So it's probably a constellation, but it's hard to say motor vehicle accidents. Even in a severe one, some of the children do remarkably well. And likewise, some of them seem well and do very badly. So I think it would be hard to do that.

LYNDA DAVIS PhD.: This morning we discussed the relationship between PTSD and TBI. Is there a similar relationship among children between post-traumatic stress disorder and shaken baby syndrome?

RACHEL BERGER, M.D.: Interesting question. I just came actually from the international conference on shaken baby syndrome, and there were some issues raised about PTSD. I have to say the degree of impairment in most of these kids is so severe that PTSD almost isn't an issue because they're severely disabled and vegetative and so PTSD, it almost can't be defined because they're too severely injured. There are a few kids who 15, 20 years later, aside from cortical blindness, seizure disorder are cognitively fairly intact. They don't have PTSD. Now, you wonder if it's because they were four months old when it happened. But I think there are so few cases where the children are

cognitively intact enough to even talk about PTSD that it's a hard -- it's something hard to define in that group.

COMMISSIONER ASTRUE: We've got time for one more. I think we're set. Thank you all. I know this has been very helpful. A lot of things for us to think over, and we're very grateful to all of you for the time you spent to help us on this. Thank you very much.
[applause]