OFFICE OF SPECIAL MASTERS

90-1630 V

(Filed: August 21, 1997)

RITA LORD, parent and legal representative of CHARLES E. LORD II, a minor,	*	
Petitioner,	*	
vs.	*	PUBLISHED
SECRETARY OF THE DEPARTMENT OF HEALTH AND HUMAN SERVICES,	*	
Respondent.	*	

Anthony Bottar, Syracuse, New York, for petitioner.

Caroline Elmendorf, U.S. Department of Justice, Washington, D.C., for respondent.

DECISION ON ENTITLEMENT

On September 27, 1990, petitioner filed her petition seeking compensation on behalf of her son, Charles Lord, II, alleging that as a result of a DPT vaccination administered on May 2, 1975, her son sustained a vaccine related encephalopathy. Petitioner alleges further that as a result of his injury, Charles has sustained permanent neurological damage. Respondent defends, arguing that the infant's injury and subsequent neurological deficits are due to a factor unrelated, namely Reye's syndrome, or a Reye's-like condition triggered, more likely than not, by an underlying inborn error of metabolism.

A hearing was held on October 30, 1996 in Washington, D. C. Inasmuch as the parties did not disagree as to facts, the hearing was limited to expert testimony. Petitioner presented the testimony of Dr. Leon Charash, of Hicksville, New York, a board certified pediatrician and, since 1956, a specialist in pediatric neurology. Respondent presented the testimony of Dr. Mary Anne Guggenheim, of Helena, Montana, a board certified pediatric neurologist. The court concludes that petitioner is entitled to compensation.

FINDINGS OF FACT

The evidence supports the following findings of fact. Charles Lord was born on February 17, 1975 and was a normal healthy infant until a few hours after his first DPT shot administered on May 2, 1975. On May 6, 1975, the infant was taken to see Dr. Brown, the family doctor in Cleveland, New York, with symptoms of tachypnea (rapid labored breathing), cyanosis, respiratory depression, reduced responsiveness developing into a coma-like state, with a three-day history of extreme irritability, vomiting, hypotonicity, lethargy, and other symptoms. His doctor arranged for him to be driven immediately to Oneida City Hospital by ambulance, but because his condition worsened to include onset of tonic/clonic seizures, cerebral edema with no corneal response, decorticate rigidity, and decrease in muscle tone followed by deep coma, he was intubated and transferred to Crouse-Irving Memorial Hospital in Syracuse, New York. His condition was considered critical.

His doctors diagnosed Reye's syndrome. The diagnosis was based on the fact that the child had an acute onset of hepatic dysfunction and encephalopathy without any other identifiable cause. See Respondent's Exhibit H at 3. Charles remained hospitalized until May 19, 1975. Thereafter he began to improve slowly. According to his mother's statement, Charles seemed to recover from the acute event, but his development thereafter was slow.

When Charles was approximately one year of age, he was found to be deaf. He demonstrated evidence of clumsiness, lagging speech, behavioral problems, and hyperactivity. Following his initial hospitalization at the age of ten weeks, no further seizures were observed until the approximate age of eight and one-half years. At the time of this writing, he is 22 years of age, deaf, moderately mentally retarded, does not speak, read or write, and has poor eye/hand coordination. He has been seizure-free for a significant period of time but recently developed cardiomyopathy that was not present in early childhood years, although it was known that he had a heart murmur in infancy.

STATUTORY SCHEME

Petitioner enjoys a statutory presumption in her favor if she is able establish that Charles sustained an injury listed in the Vaccine Injury Table found at §14 of the Vaccine Act and that the first manifestation of onset of symptoms occurred within the three-day time frame specified in the statute for that injury. Respondent may rebut petitioner's claim of an on-table injury by demonstrating, by a preponderance of the evidence, that the injury, illness, or condition was caused by a factor unrelated to the administration of the vaccine. §13(a)(1)(B). In proving a factor unrelated, however, respondent is bound by the following sections of the Act:

(2) . . . [T]he term "factors unrelated to the administration of the vaccine" --

(A) does not include any idiopathic, unexplained, unknown, hypothetical, or undocumentable cause, factor, injury, illness, or condition, and

(B) may, as documented by the petitioner's evidence of other material in the record, include infection, toxins, trauma (including birth trauma and related anoxia), or metabolic disturbances.

§13(a)(2).

Section 14(b)(3)(B) contains a related provision:

(B) If in a proceeding on a petition it is shown by a preponderance of the evidence that an encephalopathy was caused by infection, toxins, trauma, or metabolic condition the encephalopathy shall not be considered to be a condition set forth in the table. If at the time a judgment is entered on a petition filed under section 300aa-11 of this title for a vaccine-related injury or death it is not possible to determine the cause . . . of an encephalopathy, the encephalopathy shall be considered to be a condition set forth in the table. . . .

§14(b)(3)(B).

Sections 13(a)(2) and 14(b)(3)(B) are both set forth here because they are relevant to the allegation that a metabolic condition is the more likely cause of Charles' condition.

ISSUES

The parties agree that on May 6, 1975, Charles was in an encephalopathic state. At hearing, respondent's expert, Dr. Guggenheim, acknowledged that the description of his clinical course over the immediate 72-hour period following vaccination makes it likely that the encephalopathy had its onset prior to May 6, 1975, placing the first manifestation of the onset of symptoms within the table time frame. Petitioner, therefore, is entitled to a statutory presumption of a vaccine-related cause of his injury. The burden of showing a factor unrelated thus shifts to the Government.

Respondent's case in rebuttal argues that Charles' condition is consistent with Reye's syndrome or, more accurately, a Reye's-like syndrome with a metabolic disorder suspected as its cause. Respondent takes the position that a diagnosis of Reye's syndrome is consistent with his clinical course, including symptoms emerging in mid-childhood and recent years, the onset of seizures eight years after the initial hospitalization in 1975 and, more recently, onset of cardiac problems. Respondent's expert is of the opinion also that a careful analysis of the medical records identifies what she believes to be a progressive dementia -- a neurological deterioration over the years. Both experts agree that if the facts support a finding of progressive dementia, petitioner's claim of a vaccine-related encephalopathy cannot be sustained. Petitioner challenges the allegation of progressive dementia, arguing that the facts simply do not support a progressive condition. Petitioner argues further that Reye's syndrome is not a disease; it is merely a classification given to a specific constellation of symptoms, and that no tests have ever indicated the existence of a metabolic condition.

To successfully rebut the presumption of a vaccine causation enjoyed by dint of respondent's concession that a table injury exists, respondent's statutory burden of proof is to demonstrate its claim by a preponderance of evidence and is subject to §§13(a)(2) and 14(b)(3)(B) relating to proof of factors unrelated. The central issue in this case is whether respondent's rebuttal evidence is sufficient to establish a factor unrelated. If the court finds that respondent has failed to rebut the presumption of a table injury, petitioner is required to establish that Charles' present disabilities are causally related to a vaccine injury.

REYE'S SYNDROME[(1)]

Charles' treating physicians concluded that his condition was consistent with Reye's syndrome. Reye's syndrome is an appellation given to a constellation of symptoms that culminates in a "hepatic encephalopathy:" Reye's syndrome is not "a unique specific disease, but simply reflects a recognizable pattern of acute liver and brain dysfunction." See e.g. Dr. Guggenheim's 1993 Report, Respondent's Exhibit H at 6. The syndrome was first described in 1963, is very serious, and frequently results in death. Prior to the early 1960s, it was called "acute toxic encephalopathy of unknown origin." The

clinical course and symptoms of a vaccine-encephalopathy and Reye's syndrome mirror each other, but Reye's syndrome is distinguishable by the presence of not only brain dysfunction, but also of liver disease. The existence of positive acute liver studies constituted a crucial laboratory finding in Charles' illness and led the treating physicians to diagnose his condition as Reye's syndrome.

The diagnosis of Reye's syndrome led the court to consider initially a speedy decision in respondent's favor. A more careful review of the evidence, however, reveals the complexity of this case based on evidence that Reye's, in fact, is not a cause, but the result of an earlier event. Medical science is not always able to identify the specific cause and a variety of events may "trigger" its onset.

Dr. Guggenheim testified that Reye's syndrome "or a Reye's-like syndrome" in this case was probably caused or "triggered" by a congenital metabolic disorder. Respondent submitted medical articles that support a relationship between Reye's syndrome and metabolic disorders. Dr. Guggenheim's comprehensive medical report, written November 22, 1993 (Respondent's Exhibit H), argues that although Reye's syndrome has never been thought to be triggered by a DPT immunization, "a variety of infections and even live virus vaccines have been suggested as a trigger." Id. at 5,6.

The medical literature supports a finding that multiple causes or triggers for Reye's syndrome have been implicated including the following: metabolic disorders; toxins found in a number of viruses and other infectious diseases;[(2)] and para-infectious diseases secondary to immunologically mediated illnesses. Static Encephalopathies of Infancy and Childhood, Miller and Ramer Eds., Raven Press, N.Y. (1992) at 307-308. Ingestion of aspirin and even immunizations have been implicated. According to Kenneth F. Swaiman in his textbook, Pediatric Neurology, 2d Ed. (1994):

At present it is believed that Reye syndrome generally results from a complex interaction between the toxicity of certain viruses and salicylates [aspirin]. . . . The mechanism of the encephalopathy in Reye syndrome remains controversial.

Id. at 1240-1241.

EXPERT OPINION

Petitioner argues, and respondent does not deny, that no identifiable metabolic disturbance has been identified in this case. Four metabolic tests were performed, and results were negative. Respondent requested additional testing to rule out other possible disorders, but admits that such disorders are legion, and to test for all possibilities is not feasible. No factual evidence exists, therefore, to establish the existence of an inborn error of metabolism as claimed. Nonetheless, Dr. Guggenheim's opinion is that a metabolic condition can be supported as more likely than not. Her statement was summarized as follows:

I find that the preponderance of evidence in this case strongly indicates that Mr. Lord has some kind of disorder, probably metabolic in nature, that resulted in the Reye-like event in infancy (unusual) [sic], the progressive dementia, the development of seizures in mid-childhood, and a cardiomyopathy that was not present in his early childhood years. It seems to me highly unlikely that this complex disorder could be explained by a one time reaction to a vaccine. Unfortunately, none of the laboratory tests done to date identify the specific nature of his disorder.

Respondent's Exhibit A, December 1, 1995 letter of Dr. Mary Anne Guggenheim to Dr. Ellison, Medical Analysis Branch, Division of Vaccine Injury Compensation Program.

In summary, respondent's evidence consists of two factors: First, respondent relies on the testimony of their expert, Dr. Guggenheim, that an inborn error of metabolism is capable of causing all Charles' symptoms during the acute event, his subsequent clinical course, and his present deficits; a vaccine encephalopathy would not account for the liver dysfunction during the acute event, in her opinion, and could not account for certain deficits subsequently acquired. Second, both respondent's expert and petitioner's expert agree that if the evidence supports the existence of a progressive dementia and downhill deterioration, a vaccine-related encephalopathy cannot be sustained, because a vaccine-related encephalopathy is an acute event rather than a progressive injury. The issue of a progressive condition will be addressed hereafter.

Petitioner's expert, Dr. Leon Charash, argues that no evidence of metabolic disease has been established. He is of the opinion that a vaccine-related encephalopathy is more likely for the following reasons: 1) The results of four laboratory tests for metabolic disease were negative -- none was able to identify any specific metabolic disorder; 2) Reye's syndrome is rarely, if ever, found in children under the age of two; in fact he has never seen it in his 40-odd years of practice nor heard of a case in a child of such tender years; (3) 3) the infant demonstrated no signs of neurologic abnormalities or symptoms prior to vaccination, and it is unlikely that a metabolic disorder could exist without earlier symptoms being observed; 4) the onset of symptoms occurred abruptly, within hours of the shot; 5) the liver dysfunction component of the injury, hallmark of a diagnosis of Reye's syndrome, is explained by the infant's ingestion of significant levels of aspirin and exposure to Benzene (Kerosene) fumes prior to the encephalopathy -- the presence of liver dysfunction, therefore, does not rule out a co-existing vaccine-related encephalopathy;(4) and 6) no inference of a progressive deterioration can be credibly sustained. Tr. at 40,41.

DISCUSSION

The hallmark of Reye's syndrome is liver damage, and it is this component, although not exclusively, that leads respondent to reject the incidence of a vaccine related encephalopathy. Dr. Charash, for petitioner, does not claim that the vaccine is responsible for the liver damage, as stated earlier, but believes that the relatively large doses of aspirin, a known trigger for the syndrome, with the possible compounding influence of the Benzene, was the factor that accounts for the "hepatic" component of the acute event: "The first sign of aspirin intoxication would be liver malfunction. . . . For a small baby, he was given a good deal of aspirin. He was given that aspirin because he was having reactions to his vaccination." Tr. at 19-21. Additionally, according to test results, the liver abnormalities were relatively mild, "only minimally elevated," (Tr. at 38)(5) certainly not significant enough to cause the level of brain pressure observed. Dr. Charash ascribes Charles' liver damage to the aspirin and the brain damage (encephalopathy), that is, cognitive deficits, motor deficits, hyperactivity and related difficulties, to the vaccine. (6)

Much of the evidence presented at hearing explored whether Charles suffers from a progressive condition that would argue against a vaccine-related etiology, or a static condition that would support a vaccine-encephalopathy. Dr. Guggenheim finds evidence of a decline in cognitive abilities based on two "Leighter" tests, one at age 6 and the second at age 14. The first evaluation, performed at the Rochester School for the Deaf at age six years and three months, documents an overall I.Q. of 89, which is low average but within normal range. The second evaluation was performed at the New York School for the Deaf eight years later. The later test ascribed to Charles, at age 14 years, an I.Q. of 57. Dr. Guggenheim thus identifies what she perceives to be a definite and visible lag in cognition which would be consistent with a progressive condition and an underlying metabolic disturbance.

Conversely, Dr. Charash, argues that one must suspect the success of an I.Q. evaluation of a deaf child,

particularly in a six-year old.[7]

It is his opinion that the effectiveness of the first exam should be discounted in its entirety thereby discounting any evidence of a decline in cognitive abilities. Dr. Charash cites the following, written by the evaluator of the first test, in support of his opinion:

[A]ll of the psychologists who have tested Charles report a developmental lag in visual motor skills and in motor coordination. With minor variations he has been found to have low average intelligence. All are in agreement that his deafness and his visual motor lag contribute to his experience of frustration and thus render it difficult to get an accurate measurement of his intelligence.

Psychological Evaluation: Age 6 years, 3 months. P.Ex.15 at 1.

(Emphasis supplied).

Dr. Guggenheim cites the following from the same test report in rebuttal:

He was relatively calm throughout the examination, and showed great desire to remove the materials and replace them in their boxes following completion of the test items. He was at almost all times in significant interaction with the examiner, with good eye contact and good response to the examiner's direction. He showed frustration and immature withdrawal (along with squirming and moving to another part of the room) when he experienced failure on an item. In spite of this, he was able to be brought back to the next item. . . . I felt that he devoted sufficient effort to the tasks presented to render the results valid.

Id. at 2. (Emphasis supplied.)

These two statements appear to be inconsistent although they were made by the same evaluator. The evaluator, Forrest C. Orr, Ph.D, a clinical psychologist, states finally that Charles "most probably" had the potential for "average or bright normal intelligence" as he grew older. Id. He felt, however, that it was too early to tell the extent to which the child had sustained brain damage from his acute illness (at ten weeks of age). He explained that learning disabilities would become more apparent later, particularly in the area of linguistic skills, and recommended that an evaluation of linguistic skills be postponed till later. His opinion is more consistent with that of Dr. Charash:

All [psychologists] are in agreement that his deafness and his visual-motor lag contribute to his experience of frustration and thus render it difficult to get an accurate measurement of his intelligence. . . . It is too early to tell the extent to which he has been damaged or the presence of learning disabilities, which may become more apparent later. I do not believe it is possible to assess the linguistic skills of a deaf child of this age with any degree of accuracy, and I therefore suggest postponing such an evaluation for another two years or so.

Id. at 1,3.

Based on the evidence, the court cannot conclude with any degree of confidence, that Charles' first test at age six was an accurate assessment of cognitive ability. The evidence, therefore, is equivocal and does not preponderate in support of cognitive deterioration. Petitioner's expert staunchly denies any allegation that Charles is getting progressively worse; his seizure disorder, for example, has been improving markedly. Tr. at 32.

The next issue to be addressed is the significance of the onset of seizures at age 8. Dr. Guggenheim believes the late onset of seizures would be more consistent with a metabolic disorder and would likely rule out any relationship between the seizures and a vaccine. Dr. Charash disagrees. He testified that one cannot rule out a causal relationship explaining that after an injury to the brain, scar tissue develops and the brain may not discharge electrically for several years. Tr. at 43. "[S]eizures may appear any time up to . . . puberty. They can start at nine or ten from an injury either at birth or before birth, because scar tissue doesn't always discharge electrically right away." Id. The following is from the transcript:

Q. Dr. Charash, it's your testimony that an insult at age two and a half months can lead to seizures developing perhaps seven years later?

A. Yes. Particularly if the child convulses overtly during the encephalopathy and has a chaotically abnormal EEG at that time [as in this case]. That certainly makes it highly likely that the subsequent appearance of seizures would be causally related to the encephalopathy, whatever its cause.

Tr. at 43-44.

As to this particular issue, again, the court cannot discern a preponderance for either party. I do not know whether ascribing the late onset of seizures to an encephalopathy at ten weeks of age is any less possible than ascribing such seizures to an inborn error in metabolism. The court has heard numerous experts testify that signs of prenatal or neonatal injuries, whatever their cause, may not show up for several years.

Finally, Dr. Guggenheim cites the following facts as additional support for her theory of a progressive metabolic condition. In 1988, at the age of 13 years, Charles developed significant breathing difficulties and chest pain and was found to have enlargement of the heart, which turned out to be due to recurrent pericardial and pleural effusions (escape of fluids into the those parts of the body). The usual causes of a progressive cardiac problem were ruled out and no cause has been identified. Neither party suggests that these breathing and cardiac problems could be vaccine related. Respondent, however, believes they could support a finding of an inborn error of metabolism. Dr. Guggenheim finds support in the April 7, 1989 letter written by Dr. William Hannan to one of Charles' treating physicians. Dr. Hannan reviewed the emergence of Charles' cardiac problems and raised the question of whether this child might have an underlying metabolic disease which, in his opinion would explain the whole clinical picture, the "presumed Reye's syndrome" that occurred at ten weeks of age, the cardiac condition, and other symptoms as well. Respondent's Exhibit H at 5. He raised specifically the possibility of "carnitine deficiency abnormality." He acknowledges that the type of cardiac problem that the patient had is not the typical type for carnitine deficiency, but it nevertheless seemed to him "an interesting possibility." Id. Dr. Hannan's opinion is speculative and hypothetical, raising only the possibility of a metabolic disorder. For that reason, it merits limited weight, an insufficient basis for finding that such disease or disturbance exists in fact.

Identification of a specific metabolic disorder by name is not required for respondent to prove a factor unrelated. Sections 13(a)(2) and 14(b)(3)(B) require no more than a showing that a metabolic disorder exists and that more likely than not it caused the encephalopathy. See e.g. Dieudonne v. Secretary of HHS, No. 90-1695V, slip op. at 17, 1996 WL 101594 (Fed. Cl., Spec. Mstr., Nov. 17, 1996); Lassiter v. Secretary of HHS, No. 90-1036V, slip op. at 6, 1996 WL 749708 (Fed. Cl., Spec. Mstr., Dec. 17, 1996). Respondent has not demonstrated that a metabolic disorder exists.[(8)] The evidence remains hypothetical and undocumented within the intent of §13(a)(2)(A) showing only that an inborn error of metabolism is capable of causing Charles' deficits and is suspected. The evidence does not meet the level of proof required for concluding that such a metabolic condition exists in fact and that, in fact, caused Charles'

current condition. Section 14(b)(3)(B) says "If at the time a judgment is entered . . . it is not possible to determine the cause, by a prepondrance of the evidence, of an encephalopathy, the encephalopathy shall be considered to be a condition set forth in the table."

CONCLUSIONS

Petitioner has satisfied all jurisdictional and substantive requirements for establishing a Table case and is entitled to compensation. The court concludes that the DPT vaccine administered on May 2, 1975 resulted in a table injury, namely an encephalopathy, which is the proximate cause of Charles' past and present deficits with two exceptions. No evidence exists to support a vaccine-related cause of his existing cardiomyopathy, and insufficient evidence exists to determine whether Charles' deafness is congenital or vaccine-related. The issue of the etiology of Charles' deafness is reserved for further evidence to be presented during the damages phase of these proceedings.

The parties are directed to discuss issues relating to damages. If the parties are unable to agree as to terms, the parties shall seek experts for the purpose of developing a life care plan or plans for future care and treatment of Charles' vaccine related deficits.

IT IS SO ORDERED.

E. LaVon French

Special Master

1. "Reye's" syndrome appears also in some texts as "Reye" syndrome.

2. "Reye syndrome is an acquired toxic encephalopathy . . . generally following a viral prodrome. . . . Numerous metabolic disorders and intoxications may produce a Reye-like syndrome." Swaiman, Pediatric Neurology, 2d Ed. (1994) at 1238,1239.

3. Dr. Guggenheim acknowledges that onset of a Reye's-like event in a child of this tender age would be unusual, although, she points out, it can happen. Respondent's Exhibit A., She cites in support Shuttenlocker, Peter, and Trauner, "Reye's Syndrome in Infancy," Nov. 1993 Pediatrics Vol. 62, 1978, at 84.

4. In the four-day interval between the date of vaccination and admission to the hospital, Charles received five or six doses of aspirin, and had been exposed to Benzene (Kerosene) household fumes. R.Ex.H at 2. Aspirin ingestion plays a major role in Reye's syndrome, and recent recognition of that fact has resulted in discontinuation of aspirin use for children with the result that Reye's syndrome is now rarely encountered. See e.g. Ronald E. Baird et al., "Reye's Syndrome: Lessons for Family Physicians," American Family Physician, Vol. 50, No. 7 at 1454. Respondent's Exhibit J, at 1.

5. Dr. Charash states: " The enzyme levels went up somewhat and then they came back down. That may be seen in Reye's syndrome, but it also can be seen after aspirin and after cleaning fluid or Benzene . . . ." Tr. at 20.

6. Swaiman's textbook proposes that a complex interaction between aspirin and other toxins may be implicated. See discussion on p. 5.

7. At the time of his second test, when he was 14 years old, Charles was taking medication. Dr. Charash seems to suggest that medication could have a negative impact on scoring. Tr. at 46.

8. See Strother v. Secretary of HHS., 18 Cl. Ct. 816 (1989) (for a discussion of respondent's burden in establishing a factor unrelated).