J Forensic Sci, September 2003, Vol. 48, No. 5
Paper ID JFS2003009_485
Available online at: www.astm.org

TECHNICAL NOTE

Rachel S. Rosen[1]*; Vernon Armbrustmacher,*[1] *M.D.; and Barbara A. Sampson,*[1] *M.D.*

Mortality in Cerebral Palsy (CP): The Importance of the Cause of CP on the Manner of Death

ABSTRACT: Cerebral palsy (CP) is a nonprogressive motor deficit present or sustained in the perinatal period. We reviewed the files of the Office of Chief Medical Examiner of the City of New York for the 1997–2001 interval seeking those with any mention of cerebral palsy. There were 26 such cases, including 18 natural deaths, three accidents, two homicides, two therapeutic complications, and one death classified as undetermined. Proper reporting and careful investigation of these deaths is required for accurate certification of cause and manner of death, as well as for adequate tracking of these deaths for public health purposes.

KEYWORDS: forensic science, cerebral palsy, mortality

Cerebral palsy is a clinical diagnosis defined as a nonprogressive motor deficit present or sustained in the perinatal period. The most common cause of the neurological deficit is anoxic-ischemic encephalopathy (up to 23%) (1), but other conditions such as malformations, infections, and birth injury can produce the same manifestations. The term is not always applied accurately because some patients with progressive conditions or disorders acquired later are sometimes misclassified as cerebral palsy (2–5).

The medical examiner regularly evaluates decedents with this diagnosis, because these patients often die under unexplained circumstances or in public institutions. If the diagnosis is not confirmed as a nonprogressive condition present at the perinatal period, the medical examiner must be alert to the possibility of an acquired condition that may not be natural.

The CP rate has held steady in developed countries over the past 30 years (6). Several studies of the life expectancy and causes of death of patients with cerebral palsy, using registries of those with disabilities and information on death certificates, have been published (7–10). Strauss and Shevelle found that a lack of mobility and an inability to lift the head placed an individual at a higher risk of mortality than those who were able to self-feed, roll, or sit (7). Respiratory infections and primarily pneumonia were a major cause of death in those with cerebral palsy (8,9). Individuals with cerebral palsy also were found to have a greater risk for cancers and diseases of the digestive system (9). However, it is difficult to determine the mortality rate of cerebral palsy because the clinical spectrum is nonspecific and wide. Williams and Alberman found the most common causes of death on death certificates to be respiratory infections, neurologic complications of cerebral palsy, other infections, and drowning (8).

[1] Office of the Chief Medical Examiner of the City of New York, 520 First Avenue, New York, NY.

Received 20 Jan. 2003; and in revised form 10 May 2003; accepted 10 May 2003; published 4 Aug. 2003.

We reviewed the files of the Office of Chief Medical Examiner of the City of New York for the 1997–2001 interval, seeking those with any mention of cerebral palsy. During those years approximately 25,000 medical examiner autopsies were performed, 26 of which were on persons with cerebral palsy. The results of the investigations and autopsies are described, illustrating the value and need for such investigation.

Homicides and Undetermined

Case 1

A 14-year-old boy was given the diagnosis of CP at the age of 3 months following a seizure. Since then he lived in multiple nursing care facilities in a severely debilitated state, being blind, nonverbal, nonambulatory, and with only primitive reflexes. His immediate cause of death was beta-hemolytic streptococcus septicemia and bronchopneumonia, a complication of his debilitated condition.

Examination of the 840-g brain and dura revealed an adherent, bilateral 2.0 to 3.0-mm-thick, yellow-brown subdural neomembrane over the cerebral convexities including the sides of the falx. Calcified plaques up to 2.0 mm thick and 25.0 mm in diameter were embedded in the neomembrane over the parietal convexities. The brain had cystic encephalomalacia in the right occipital/temporal and left occipital/parietal areas. The cerebral surface was soft due to underlying cavitation. The brain stem and cerebellum were not remarkable, and the arteries at the base of the brain followed a normal distribution and were patent.

Coronal sections of the cerebrum revealed bilateral, smooth-lined, porencephalic cavities replacing all but the subarcuate white matter. The cavities communicated with the ventricles and extended to the subarachnoid space, but there was no cortical scarring or ulegyria. Finger-like extensions of the cavities extended through the subcortical white matter as far posteriorly as the occipital lobe on the right, but most projections ended at the mid-cerebral level. The corpus callosum was thin.

Microscopic sections of the frontal and parietal lobes showed the porencephalic cavity lined by dense gliosis, extending from the ependymal surface of the lateral ventricle to the pia mater. There was also extensive periventricular gliosis, as well as a subdural fibrous membrane representing an organized subdural hemorrhage.

Because of the history of an apparently abrupt onset of his condition at 3 months of age and the presence of an organized subdural hemorrhage, a search of his early medical records was undertaken. It revealed a normal labor, delivery, and post-natal course. The mother had a history of drug abuse. At a routine well-baby clinic visit at 3 months of age, the baby was noted to be lethargic. Further evaluation revealed acute bilateral retinal hemorrhages, subarachnoid hemorrhage, and subdural hemorrhage. The diagnosis of inflicted head injury (Shaken Baby Syndrome) was made, and the child became a ward of the state. The manner of death was classified as homicide.

Case 2

A 5-year-old girl carried a diagnosis of CP since birth. Her delivery by cesarean section at 43 weeks gestation was complicated by hypotonia, apnea, and tonic-clonic seizures. Her birth weight was 6.8 lb and her length was 20 in. She had severe psychomotor and mental retardation, spastic quadriparesis, and was bedridden. At the time of her death, she was found unresponsive, face down on her bed. Autopsy disclosed severe malnutrition with body weight of 17 lb and a length of 31 in. Her skin was unevenly pigmented with lanugo hair on the back and extremities. There was marked atrophy of fat and skeletal muscle, as well as the solid organs. She also was found to be dehydrated with a vitreous urea nitrogen of 57 mg/dL and creatinine of 0.2 mg/dL. Multiple abrasions and ulcers were present on the head, torso, and extremities.

Neuropathological examination of the 340-g brain and dura disclosed a thin, bilateral, slightly golden brown subdural neomembrane, mainly on the right. There was loss of white and gray matter of the frontal, parietal, and occipital lobes, bilaterally. There were scattered islands of residual cortex in the frontal lobes and strips of relatively preserved cortex in each parietal lobe. The temporal lobes were relatively unremarkable. The brainstem and cerebellum were not remarkable, and the arteries at the base of the brain followed a normal distribution and were patent.

Coronal sections of the cerebrum revealed almost total cystic encephalomalacia of the white matter with multiple glial strands in the frontal, parietal, and occipital lobes, consistent with a remote ischemic event. Transverse sections of the spinal cord revealed degeneration of the lateral columns.

Microscopic examination of the brain and spinal cord showed multiple glial strands in the frontal lobe, spongy degeneration of the lateral columns in the spinal cord, and loss of cells within the dentate fascia of the hippocampus, consistent with an old ischemic event.

The neuropathologic abnormalities in this case are consistent with CP. The cause of death was determined to be complications of severe malnutrition including dehydration and bronchopneumonia due to cerebral palsy (hypoxic ischemic encephalopathy). Because of parental neglect, the manner of death was classified as homicide.

Case 3

An 8-month-old girl was placed in a nursing home where she was eventually diagnosed as having profound mental retardation, chronic organic brain syndrome, cerebral palsy, asthma, seizure disorder, left hemiparesis, and blindness. She remained in nursing homes until her death at age 19 years.

Neuropathologic examination of the 500-g brain and dura disclosed that the brain was soft and the cerebral hemispheres were flaccid due to ex-vacuo enlargement of the ventricles. Most of the cerebral mantle ranged from 2.0 to 5.0 mm thick. The cerebral gyri were markedly widened and flattened except over the dorsal-medial occipital lobes, where the surface was smooth and tan. The cerebellum was small, and the inferior cerebellar cistern was 3.0 by 4.0 by 2.5 cm. The brain stem was also small.

Coronal sections of the cerebrum showed marked enlargement of the lateral ventricles with almost complete loss of the cerebral white matter and thinned cortex. The diencephalon was relatively intact. The spinal cord dura revealed a narrowing to 4.0 to 5.0 mm in diameter of the distal 4 to 5 cm of thoracic cord in the thoracolumbar segment.

The final neuropathologic diagnoses were hydranencephaly, consistent with a remote traumatic and/or anoxic/ischemic encephalopathy and remote transverse thoracic myelopathy.

The decedent had been the product of a full-term pregnancy and was born by cesarean section. She was developing normally until she had two episodes of head trauma. At 3 months of age, while under the care of her father, the decedent fell off a tabletop and was hospitalized with seizures, bilateral retinal hemorrhages, and intracranial bleeding. A CT scan showed leucomalacia in the frontal and occipital lobes and hydrocephalus. The child was placed temporarily in foster care. Two months later, the decedent acquired a compression-type injury while supposedly being clutched under the father's arm in order to protect her from being struck by another male playing football. The decedent was hospitalized again with skull fractures, subarachnoid hemorrhage, and a seizure disorder. She again was placed in foster care. At 8 months of age, she was permanently placed in a nursing home. The underlying cause of death in this case is head trauma. The clinical diagnosis of CP was inappropriate. The manner of death was classified as undetermined because of reasonable doubt about whether the trauma was accidental or abusive.

Accidents

Of the 26 cases reviewed, three deaths were classified as accidents. The first was a 14-year-old boy with CP who fractured his femur after falling from his wheelchair. The next day while at home the decedent had difficulty breathing and became cyanotic. He was taken to the hospital where he was pronounced dead. At autopsy, the cause of death was determined to be pulmonary and systemic fat emboli due to fracture of the left femur. The brain revealed anoxic/ischemic encephalopathy with diffuse neuronal loss and degeneration of the corticospinal tracts.

The second instance was an 18-year-old male who, while home for the holidays from his long-term care facility, was found unresponsive in his bedroom after having a restless night. He was found, according to the family, prone on his bed with the right side of his face partially in a towel, obstructing his nose and mouth. Due to a severe vertebral deformity, the decedent could not turn his head from left to right. The cause of death was certified as positional asphyxia with upper airway obstruction complicating cerebral palsy with severe kyphoscoliosis. His brain showed slight sulcal widening and white matter atrophy/loss.

The final instance was an 11-year-old girl burned in her bathtub when her sister accidently turned on the hot water while temporarily unsupervised. The girl had second and third degree burns to 65% of her body. Investigation disclosed no evidence of child

abuse or neglect; therefore, the death was classified as an accident. The cause was scald burns of approximately 65% of the total body surface area with bronchopneumonia and sepsis.

Neuropathologic examination of the 890-g brain showed polygyria in each cerebral hemisphere, thickening of the leptomeninges around the brain stem and cerebellum, and a tenting deformity of the superior cerebellar vermis, suggesting an upward transtentorial herniation due to obstructive enlargement of the fourth ventricle. Coronal sections of the cerebrum showed marked, generalized diminution of the cerebral white matter, a thin corpus callosum, and a periventricular cavity surrounding the left frontal pole of the lateral ventricle communicating with the body of the lateral ventricle, forming an area of marked dilatation with severe loss of white matter in the temporal and occipital lobes. The cerebellum exhibited severe enlargement of the fourth ventricle with secondary compression of the cerebellum and brainstem.

Natural Deaths

The majority of deaths of those with cerebral palsy were natural (18 of 26). Their ages ranged from 20 months to 49 years. Only two individuals were institutionalized at the time of their death. The two major causes of death were infectious complications such as bronchopneumonia (five deaths), sepsis (two deaths), and pneumonitis (one death) and seizure disorder (seven deaths). In addition, there was one death due to myxomatous degeneration of the mitral valve and one death from pulmonary thromboembolism due to deep venous thrombosis of the lower extremity. This decedent had decreased mobility complicating a congenital lower extremity defect of unknown etiology. One death was due to acute subarachnoid hemorrhage of undetermined etiology. Importantly, trauma was ruled out. Comparison of the neuropathologic examinations in these cases showed the most common findings to be degeneration or diffuse neuronal loss, gliosis, and atrophy (in nine cases), hydrocephalus (five cases), organized subdural hemorrhages (four cases), degeneration of corticospinal tracts (three cases), cystic encephalomalacia (three cases), anoxic/ischemic encephalopathy (three cases), microcephaly (two cases), and cerebellar degenerative changes consistent with remote hypoxia (two cases).

Therapeutic Complications

In New York City, therapeutic complication as a manner of death is defined as a fatality that arises from predictable complications of diagnostic and therapeutic procedures. There were two such cases in our review. The first was a 44-year-old man with CP whose cause of death was complications (including peritonitis, acute pancreatitis, and aspiration pneumonia) following laparoscopic cholecystectomy for cholelithiasis. The contributing cause of death was cerebral palsy with seizures. In addition, a remote bilateral cerebral border zone infarct was found during the neuropathologic exam. The second case was a 29-year-old man who died of complications of aspiration with respiratory failure following extubation after femoral hernia repair. The contributing cause was cerebral palsy. Examination of the brain showed bilateral atrophy of the hippocampi. Both of these individuals lived at home prior to hospitalization.

Discussion

The Office of Chief Medical Examiner of the City of New York performs approximately 5000 autopsies per year. Between 1997 and 2001, 26 decedents who carried the diagnosis of cerebral palsy were autopsied. Of these, 14 were males and twelve were females with a mean age of 18 years (age range 1½ to 49 years old). Fifty-four percent were black, 23% were Hispanic, and 23% were Caucasian. The manner of death was broken down as follows: 18 natural (69%), three accidents (11%), two therapeutic complications (8%), two homicides (8%), and one undetermined (4%). Of the 18 natural deaths, over half were due to infectious complications (Fig. 1). Overall, this is consistent with previously published reports.

In a study by Strauss et al., the mortality of a California population with cerebral palsy was analyzed in an effort to identify diseases that may have their etiologies rooted in cerebral palsy. Individuals were evaluated annually using the Client Development Evaluation Report (CDER), which was used to identify the severity, type, and location of CP. Over a 9-year period, 4028 of the 45,292 people identified with CP died. Of these deaths, 40 were due to drowning, 17 due to pedestrian motor vehicle accidents, and 29 were homicides. Of the 29 homicides, four were due to child battering and other maltreatment and 14 were due to the "late effects of injury purposely inflicted by other persons" and may have been the cause of the cerebral palsy (9).

One problem encountered in tracking deaths due to cerebral palsy is that the diagnosis often is not mentioned on the death certificate. Evans and Alberman found that cerebral palsy is more likely to be mentioned on the death certificate if the child dies in the first few years of life. They found cerebral palsy mentioned in less than half the cases of children aged ten or more who died of complications of cerebral palsy (11). It is also vital that cerebral palsy be included on the death certificate so that state and national government statistics are accurate and programs and resources for the developmentally disabled are appropriately directed and funded. When cerebral palsy is listed as the cause of the death, the etiology of the cerebral palsy must be included on the death certificate to clarify the manner of death.

In the investigation of alleged cerebral palsy fatalities, the key question to ask is whether neurologic impairment was present since birth and was not progressive. If the answer is no to either question, further investigation must be done. The importance of this is illustrated clearly by the case of the 14-year-old boy described previously. If investigation into the cause of his cerebral palsy had not been done, his death would have been misclassified as natural

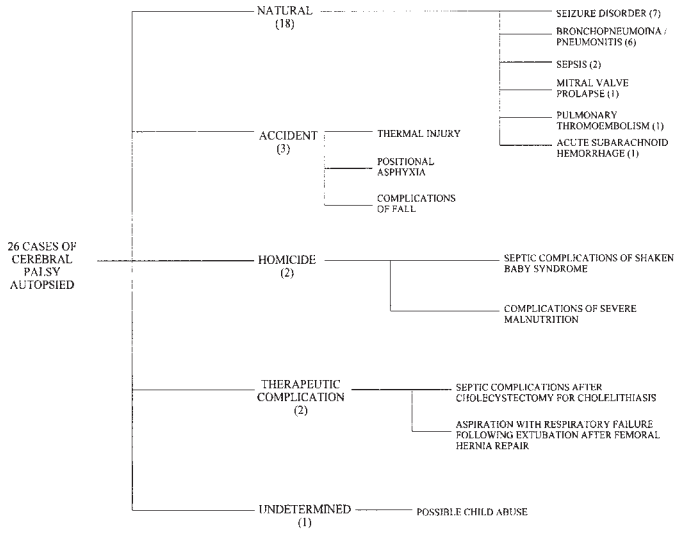

FIG. 1—*Causes of death.*

rather than homicide. Evidence of old injury, such as healing fractures or a subdural neomembrane, also should prompt further investigation. In addition, the challenge of caring for these children places them at risk for abuse and neglect. This is illustrated by the case of the 5-year-old girl with severe CP who was starved to death because of the difficulties involved in caring for her. Although the majority of true cerebral palsy deaths are natural, these children are also at risk for accidental deaths and therapeutic complications due to their debilitation.

Acknowledgments

We thank Dr. Charles Hirsch, Chief Medical Examiner of the City of New York, for helpful discussions and a critical review of the manuscript.

References

1. Nelson KB, Ellenberg JH. Antecedents of cerebral palsy: multivariate analysis of risk. N Engl J Med 1986;315:81–6.
2. Paneth MD. Etiologic factors in cerebral palsy. Ped Ann 1984:191–201.
3. Nelson, KB. What proportion of cerebral palsy is related to birth asphyxia? J Pediat 1988;112(4):572–4.
4. Nelson KB, Leviton A. How much of neonatal encephalopathy is due to birth asphyxia? AJDC 1991;145:1325–31.
5. Paneth MD. Birth and the origins of cerebral palsy. New Engl J Med;1986 July:124–6.
6. Clark, SL, Hankins, GD. Temporal and geographic trends in cerebral palsy—fact and fiction. Am J Obstet Gynecol 2003;188(3):628–33.
7. Strauss D, Shavelle R. Life expectancy of adults with cerebral palsy. Dev Med & Child Neur 1998;40:369–75.
8. Williams K, Alberman E. Survival in cerebral palsy: the role of severity and diagnostic labels. Dev Med and Child Neur 1998;40:376–9.
9. Strauss D, Cable W, Shavelle R. Causes of excess mortality in cerebral palsy. Dev Med and Child Neur 1999;41:580–5.
10. Strauss D, Shavelle R, Anderson TW, Baumeister A. External causes of death among persons with developmental disability: the effect of residential placement. Am J Epidemiol 1998;147:855–62.
11. Evans PM, Alberman E. Certified cause of death in children and young adults with cerebral palsy. Arch Dis Child 1991;66:325–9.

Additional information and reprint requests:
Barbara A. Sampson, M.D.
City Medical Examiner
Office of Chief Medical Examiner of the City of New York
520 First Ave.
New York, New York 10016