Original Paper

Pediatric Neurosurgery

Pediatr Neurosurg 1996;24:292–298

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Long-Term Outcome in Infants with the Shaking-Impact Syndrome

Key Words
Head injury
Infants
Shaking-impact syndrome
Child abuse
Subdural hematoma
Outcome

Abstract
Nonaccidental injury accounts for nearly one quarter of all hospital admissions for head injury in infancy, and is associated with significant morbidity and mortality. Long-term outcome in survivors, however, has been incompletely studied. In this series, 84 infants 2 years of age and younger with the shaking-impact syndrome consecutively admitted to a single hospital between 1978 and 1988 were identified. A questionnaire detailing current medical, developmental, and behavioral status was developed, and attempts were made to locate the 62 children surviving the acute injury. Family instability and strict confidentiality restrictions precluded locating the majority of children, but 14 children with demographic and injury characteristics similar to those of the overall group were contacted at an average of 9 years after injury. Seven children were severely disabled or vegetative, 2 were moderately disabled, and 5 had a good outcome. Of the latter group, 3 had repeated grades and/or required tutoring. Acute factors associated with poor outcome included unresponsiveness on admission, need for intubation, age less than 6 months, and bilateral or unilateral diffuse hypodensity on CT scan. All children with bilateral diffuse hypodensity and loss of gray-white differentiation on CT scan remained blind, retarded, nonverbal, and nonambulatory in spite of aggressive medical and surgical management. This study suggests that the majority of children surviving the shaking-impact syndrome suffer major permanent morbidity, and that acute factors predicting long-term outcome may help guide aggressiveness of care.

Introduction

The shaking-impact syndrome, also referred to as the shaken-baby syndrome, has become recognized as one of the most frequent causes of traumatic mortality and morbidity in infants [1, 2]. Patients with this condition typically present with neurologic findings ranging from irritability and lethargy to coma and death, and are found to have subdural and subarachnoid hemorrhage frequently accompanied by retinal hemorrhages and fractures of the

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1016–2291/96/0246–0292$10.00/0

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Received:
August 15, 1995
Accepted in revised form:
July 10, 1996

ribs and long-bone metaphyses. The mechanism is a nonaccidental inflicted injury probably involving rapid angular acceleration-deceleration forces of a major magnitude [3, 4]. While progress has been made in the acute recognition of the clinical and radiographic findings of the syndrome, less is known about the long-term consequences to survivors [5–8]. In particular, while it is not difficult to predict significant disability in those infants who demonstrate massive radiographically evident parenchymal loss, the long-term prognosis in infants with less acutely severe injuries remains largely unknown. In this study, follow-up of a large cohort of infants with strictly defined shaking-impact syndrome whose injuries occurred between 5 and 15 years previously was attempted in order to provide information regarding long-term outcome. Acute factors were analyzed to identify predictors of eventual outcome.

Materials and Methods

All aspects of the study protocol were approved by the Institutional Review Board. The study population consisted of 84 consecutively admitted children 2 years of age or younger with the diagnosis of shaking-impact syndrome during a 10-year period (July 1978 to June 1988). Patients admitted between 1978 and 1985 formed the basis of a previously reported study on the acute findings in this group; these patients had been identified through records of all inpatients evaluated for suspected child abuse or neglect [4]. Patients admitted during the subsequent 3 years were identified by the same mechanism. All children had clinical and/or radiographic confirmation of subdural and/or subarachnoid hemorrhage. Based on the best history given after multiple interviews with caretakers, the lack of other medical conditions (such as coagulopathy), and supportive findings such as soft-tissue injury, fractures, and retinal hemorrhages, all children were considered by the child abuse evaluation team to have findings consistent with inflicted injury. In equivocal cases an algorithm for the determination of inflicted injury was used to decide inclusion; children in whom insufficient evidence was present to confirm the suspicion of nonaccidental injury were excluded from the study (20 additional patients had injuries consistent with shaking-impact syndrome during this time period, but did not meet criteria for inclusion) [2].

From the total group of 84 patients who met criteria for inflicted injury and shaking-impact syndrome, 62 children surviving the acute injury were identified. Medical records from the acute hospitalization were reviewed and patients were classified by the following variables: age; gender; intracranial hemorrhage type (subarachnoid hemorrhage, acute or chronic subdural hemorrhage); presence of retinal hemorrhages; reported mechanism of injury/presenting complaints; presence of associated injuries including skull fractures, bruises, other fractures and soft-tissue injuries, and old fractures; level of consciousness (coma, lethargy or irritability, or alert and active), and presence of seizures. Children were considered comatose if they exhibited spontaneous posturing, showed either no response to noxious stimulation or a stereotyped response (such as decerebration), did not open their eyes or opened their eyes without visual responsiveness, and did not cry.

A telephone questionnaire was developed for the childrens' caretakers. Last known addresses and telephone numbers were gathered from inpatient and outpatient medical records, rehabilitation facilities, social-service and child protection agencies, local pediatricians, and public telephone directories for all surviving patients. Families were contacted by the investigators and asked if they would like to voluntarily participate in a follow-up study of children with head injuries by answering questions about the patient by telephone. The interview took approximately 30 min, and included information in four spheres. Medical information included questions regarding general health, current medications, seizures, surgery, and pediatric and specialty clinic follow-up. Functional information included questions regarding vision and hearing, ambulation and language function, ongoing physical, occupational, or other therapies, and activities of daily living such as feeding, dressing, and toileting. Educational information included school history, placement (regular classroom, special education, socially/emotionally disturbed, or combination), special services utilized, and presence of memory and attention difficulties. Social information included questions regarding behavior and social skills.

All available acute and follow-up medical records for participating patients were reviewed. In addition to the acute-injury variables described above, more detailed information regarding follow-up hospital course, radiographic findings, and subsequent medical follow-up was compiled for each patient with an interview completed. Factual information from the interview was validated whenever possible by comparing responses with information available from the medical record.

Outcome at the time of the telephone interview was classified using categories parallel to those in the Glasgow Outcome Score [9]. Since this score was designed for adults and is based largely on level of function compared to premorbid state, it is not directly applicable to patients injured as infants. For this reason outcome categories in this series were assigned according to comparison to age-appropriate activities in normal children, and included good outcome (independent in normal activities, mild impairments permitted), moderate disability (independent but with impairment, not at age level), severe disability (not independent in normal age-appropriate activities, cognitive and/or physical disability), and vegetative.

In order to test for differences between the total group of 62 survivors and the subset of children who could be located for long-term follow-up and for whom follow-up interviews were conducted, a χ^2 analysis was performed to look for differences between these two groups using all the demographic and acute-injury variables outlined above.

Results

Of the 62 surviving patients identified, whose injuries had occurred an average of 10 years previously, successful contact with current caregivers was made for only 14 patients (22.6%). Because the study group of children had suffered inflicted injuries, most were discharged to temporary foster care. Locating families was hampered by several factors, including lack of long-term follow-up at

Table 1. Patient data

Pt. No./ sex	Age at injury/ follow-up	History	Physical examination/ intubation	Radiology	Outcome
1/M	2 mo/15.5 yrs	seizures, apnea	opisthotonic, unresponsive; blood on LP; bilateral retinal detachment (treated acutely outside hospital; not intubated)	bilateral SDH	Severe: blind, retarded, ambulatory and verbal; seizure disorder
2/M	4 mo/13.1 yrs	lethargy, poor oral intake	irritable but arousable; bilateral retinal hemorrhages	bilateral occipital SAH/SDH; bilateral parietal fractures	good: regular school, A–B grades
3/F	11 mo/11 yrs	fell off sofa, down 3 steps in walker; seizures	sleepy but arousable, left retinal hemorrhage; occipital cephalo-hematoma	left acute SDH, decreased density left temporal lobe (resolved on later scans)	good: last seizure at age 2; regular school; tutored
4/M	2 mo/9.9 yrs	poor feeding, seizures; fall 2 weeks ago	left hemiparesis; bilateral retinal hemorrhages; scalp bruise; intubated	SDH/SAH; multiple focal areas of hypodensity/infarction and contusion; bilateral skull fractures; femur and rib fractures	severe: left hemiparesis; speech impediment; special education; behavioral problems
5/F	7 mo/10.1 yrs	disciplined for crying; irritable	irritable but responsive; transient apnea/bradycardia; bilateral retinal hemorrhages	small SDH/SAH; fractures ribs, clavicle, humerus, vertebra	good: repeated 1st grade; obese; eneuresis
6/F	4 mo/9.1 yrs	fell from swing 10 days ago; now lethargic	irritable; bilateral 6th n. palsies, bulging fontanelle; bilateral retinal hemorrhages	bilateral SDH	moderate: 'perceptually impaired' – motor, memory, attention; several years behind grade level
7/M	1 mo/8.8 yrs	irritable, poor feeding, 'jittery'	lethargic; swollen knee; bilateral retinal hemorrhages	small SDH, SAH; diffuse hypodensity; femur and rib fractures	severe: blind, nonverbal, wheelchair-bound, seizures
8/F	6 mo/7.8 yrs	found limp, poor breathing	posturing, unresponsive; bilateral retinal hemorrhages; intubated	diffuse SAH/SDH; right parietal contusion	severe: blind, nonverbal, wheelchair-bound, seizures
9/M	2 mo/7.5 yrs	irritable, vomiting, seizures	seizures, apnea; intubated	left SDH, SAH, left parietal contusion; multiple rib, clavicle, metaphyseal fractures	moderate: language delay, mild right hemiparesis; short attention span; tutored
10/M	3 mo/7.1 yrs	'funny leg position; not breathing well'	decerebrate; apnea; bilateral retinal hemorrhage; intubated	interhemispheric subdural; diffuse hypodensity	severe: blind, nonverbal, wheelchair-bound
11/F	2 yrs/8 yrs	assaulted by mother with psychiatric illness	unresponsive; seizures; bruises at iliac crests, frontal scalp; intubated	interhemispheric subdural; diffuse hypodensity	vegetative: died from respiratory compromise age 8
12/F	4 mo/5.6 yrs	irritable	irritable; thigh swelling; bilateral retinal hemorrhages	bilateral small occipital SDH; femur fracture	good: tutor for speech; 'behavior problems'
13/F	3 mo/5.5 yrs	lethargy, breathing problems	unresponsive; multiple bilateral retinal hemorrhages; seizures/apnea; intubated	right SDH, skull fracture, right frontal contusion; posterior interhemispheric blood; right hemispheric hypodensity; fractures ribs, humerus, tibia, skull	severe: hemiparesis; school for develop-mentally impaired; poor attention; aggressive behavior
14/F	9 mo/5.9 yrs	lethargy	lethargic but arousable; full fontanelle; increased tone in legs; bilateral retinal hemorrhages; multiple bruises	left SDH; interhemispheric blood; left temporal contusion; hypodensity left ACA distribution (left pericallosal infarct); skull fractures; metacarpal and acromial fracture	good: regular 1st grade

M = Male; SDH = subdural hemorrhage; LP = lumbar puncture; SAM = subarachnoid hemorrhage; F = female; ACA = anterior cerebral artery.

the parent institution, and by a high incidence of underlying family instability. The most significant obstacle to locating children was the existence of strict legal and confidentiality restrictions which did not allow the child-protective service agencies to disclose the patients' whereabouts, even to medical practitioners. All caregivers who were located participated fully in the study, and were uniformly willing to discuss the subjects without reservation.

Patient Characteristics

Of the 14 patients available for follow-up, there were 6 boys and 8 girls (table 1). Mean age at injury was 6.4 months. This group did not differ significantly from the overall population of 62 children with respect to any of the variables studied, including age, gender, history/presenting complaints, intracranial hemorrhage classification, level of consciousness, or presence of retinal hemorrhages, skull fractures, or associated extracranial injuries.

Presenting symptoms in the 14 patients included lethargy or irritability in 6 children, seizures without coma in 3, and coma with or without seizures in 5. Twelve patients had retinal hemorrhages. Acute CT scans showed subarachnoid and/or subdural blood in all children; 9 patients also parenchymal abnormalities including diffuse or focal hypodensity or contusions (table 1).

Seven children were intubated in the acute period for apnea, seizures, and/or unresponsiveness. Children were treated in the acute period with standard management for head injury, including intensive care management, intracranial pressure monitoring when indicated (6 patients), and tapping or drainage of subdural collections as needed.

Long-Term Follow-Up

One child who had remained vegetative, with decerebrate posturing and severe seizures, died 5 years after injury from respiratory compromise. The remaining children had a mean age of 9 years at the time of the follow-up survey (range 5.5–15.5 years). Of these, 6 were severely disabled, 2 were moderately disabled, and 5 had a good outcome. The surviving children with severe disability include 3 who are blind, nonverbal, and wheelchair-bound, 1 child with blindness, mental retardation and seizures, and 2 with hemiparesis, severe learning disabilities and behavioral problems. The 2 children with moderate disabilities have perceptual impairments and difficulties with memory and attention requiring special education; 1 also has a mild hemiparesis. The 5 children with good outcome are functioning in regular school, although 3 have repeated grades or require tutors, and 2 have behavioral problems (table 1).

Table 2. Acute factors and outcome

Acute factors	Outcome		
	good	moderate	severe/veg.
Unresponsive (n = 5)	0	0	5
Intubation (n = 6)	0	1	5
Seizures (n = 5)	1	1	3
Apnea (n = 6)	1	1	4
CT findings			
Diffuse hypodensity (n = 4)	0	0	4
Focal hypodensity or contusion (n = 5)	2	1	2
Age			
<6 months	2	2	6
>6 months	3	0	17

Relationship between Acute Factors and Outcome

The small number of patients available for long-term follow-up precludes detailed statistical analysis of correlations between acute factors and outcome; however, several trends are apparent and will be described (table 2).

Level of Consciousness. All 5 children who presented with unresponsiveness (patients 1, 8, 10, 11, and 13) remained vegetative or severely impaired at follow-up.

Intubation. All 6 children (patients 4, 8, 9, 10, 11, 13) who required intubation in the acute period, whether for apnea, seizures, or unresponsiveness, have severe (5 children) or moderate (1 child) disabilities.

Seizures. Of the 5 children with acute seizures (patients 1, 3, 5, 9, 13), 1 had a good outcome, 1 has moderate disabilities, and 3 are severely disabled.

Apnea. Of the 6 children presenting with apnea (patients 1, 5, 8, 9, 10, 13), 1 had a good outcome, 1 has moderate disabilities, and 4 are severely disabled.

Computerized-Tomography Findings. All children with bilateral diffuse hypodensity with loss of gray-white differentiation (patients 7, 10, and 11) or unilateral hemispheric hypodensity (patient 13), which was apparent on the initial study or within the first few days of injury, had severe disability at long-term follow-up (fig. 1, 2). All children with bilateral diffuse hypodensity became vegetative or blind, nonverbal, and wheelchair-bound. Of children with focal areas of hypodensity or contusion (patients 3, 4, 8, 9, 14), 2 had a good outcome, 1 has moderate disabilities, and 2 are severely disabled (fig. 3).

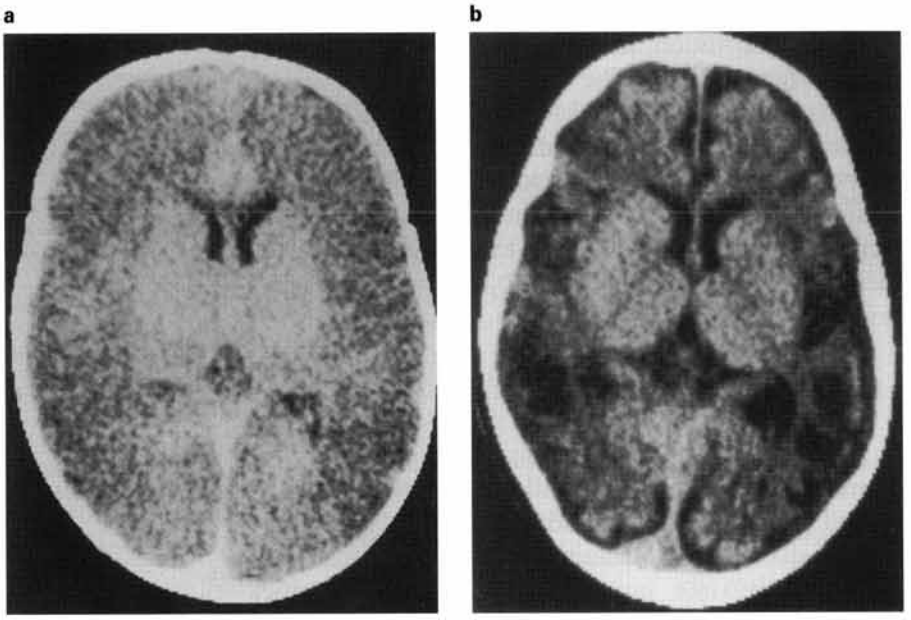

Fig. 1. a Acute CT scan, patient 7. Interhemispheric and tentorial subdural hemorrhages were identified, along with diffuse bilateral hypodensity and loss of gray-white differentiation. **b** Follow-up CT 1 year after injury showing severe bilateral cortical atrophy. The child remains severely impaired (blind, nonverbal, nonambulatory) 9 years later.

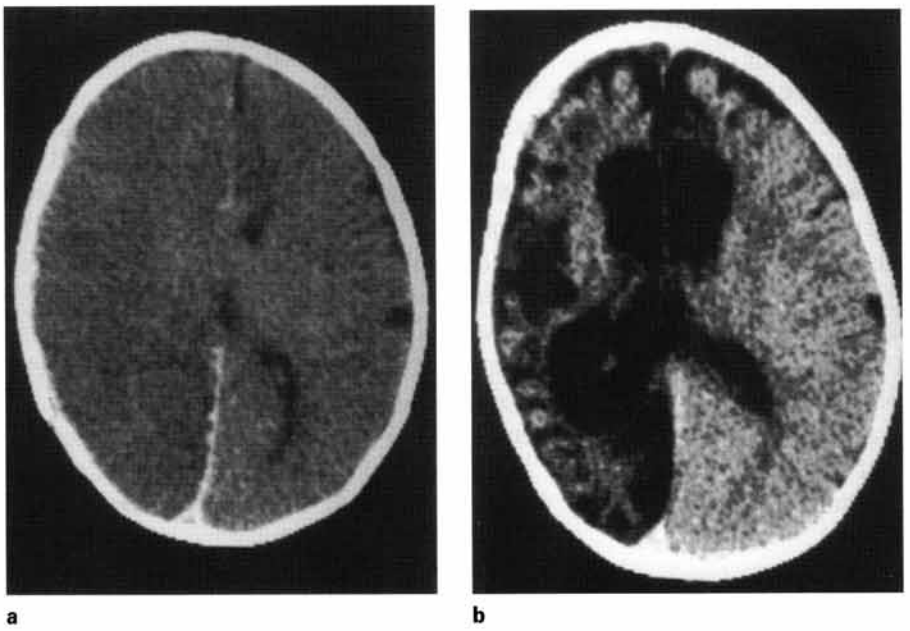

Fig. 2. a Acute CT scan, patient 13. Skull fracture, subdural blood and diffuse hypodensity of the right hemisphere is noted. **b** Follow-up study, 3 months after injury, showing severe atrophy of the right hemisphere and contralateral medial frontal lobe. The child has a hemiparesis, developmental delay and behavioral disorder 5 years later.

Age at Injury. Six of 7 children with severe disability were 6 months or younger when injured; 3 of 5 with good outcomes were older than 6 months at the time of injury (table 2).

Discussion

It has been recognized that outcome after severe head injury is better in children than in adults, with the notable exception of the very young [10, 11]. Inflicted injury is the most common cause of mortality in very young children in some series, and this entity is likely responsible for the higher mortality seen in infants in large series of pediatric head injury [2, 12]. Series of acutely injured children with the shaking-impact syndrome have provided mortality rates ranging from 15 to 27% [4, 13]. The acute mortality rate of 26% in the present series of 84 patients with strictly defined shaking-impact injuries during a 10-year period in a single institution is consistent with rates reported in previous series.

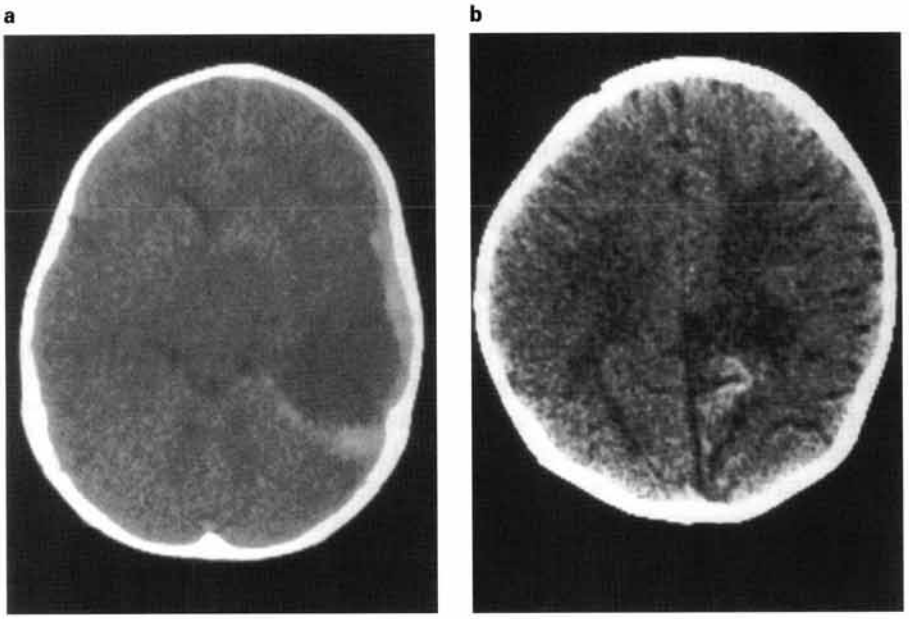

Fig. 3. a Acute CT scan, patient 14. Skull fractures, left subdural hematoma, left temporal contusion, and hypodensity in the distal left anterior cerebral artery distribution were noted. **b** Follow-up study, 2 weeks after injury, showing hyperdensity at the site of the focal distal anterior cerebral artery infarction. The child is performing at age level in regular public school.

Long-term outcome data in survivors of the shaking-impact syndrome, particularly in the less severely injured children, have been lacking. Caffey [3] postulated that many cases of mental retardation in children might be due to unrecognized abusive head injury sustained in infancy. Follow-up studies of children surviving brain injuries from a variety of mechanisms have suggested poorer outcome in those children who have suffered nonaccidental injury [14]. Some series reporting follow-up of abused children have disclosed incidences of mental retardation greater than 40%, but the injury characteristics or other factors contributing to this outcome are not well delineated [15, 16]. In fact, there exists no standardized, validated assessment tool to measure long-term outcome in children injured as infants; the current study required construction of such an instrument, and validation will require further prospective studies.

Children with the shaking-impact syndrome have many factors other than brain damage resulting from the traumatic event itself that can be expected to influence long-term outcome. These include premorbid factors, such as prematurity or other conditions predisposing to neurologic disability, and environmental factors such as family instability and socioeconomic conditions [8, 17, 18]. Such influences would seem to have the greatest effect in children with the less severe injuries, since in children with severe acute brain damage the injury itself would likely predominate as the factor most responsible for severe neurologic disability. Although 5 children in this series had a good outcome, 3 of these have repeated grades or require tutoring; the relative contributions of brain injury versus other factors affecting these children remains unknown.

Among types of pediatric head injury, the shaking-impact syndrome is homogeneous in that all patients have subarachnoid and/or subdural hemorrhage. However, parenchymal abnormalities, including diffuse or focal areas of hypodensity or contusion seen on CT scan, vary among patients and correlate with clinical injury severity. While the present series is limited by the small number of patients who could be located, these patients appear to represent a cross-section of the larger group with respect to acute clinical severity of injury and the typical spectrum of associated parenchymal findings. As might be expected, both acute clinical indices (such as level of consciousness and need for intubation) and radiographic findings (such as diffuse or multiple areas of hypodensity) appear to predict poor long-term outcome.

Among practicing neurosurgeons who manage infants with this disorder, the appropriate treatment is not always clear, for it is still unknown whether the damage is primarily traumatic or hypoxic/ischemic. Whether aggressive management for increased intracranial pressure and brain swelling influences outcome in this condition remains unknown. However, in this series the outcome of patients who were unresponsive on admission or whose CT scans showed diffuse unilateral or bilateral hypodensity and loss of gray-white differentiation remained poor, even with aggressive management, many years after the injury. Additional indicators of poor prognosis were age

less than 6 months and need for intubation. Of all factors analyzed, outcome was the most consistent and most unfavorable (vegetative or blind, nonverbal, and nonambulatory) in those children with CT findings of diffuse bilateral hypodensity. Though more recent series have shorter durations of follow-up, we have not seen any exceptions to the rule that children with the so-called 'big black brain' on CT scan after nonaccidental injury always die or remain severely impaired [19]. This condition is apparent at presentation or within the first 48–72 h after admission using standard unenhanced CT scan. This information should raise the question as to whether aggressive medical intervention is warranted when diffuse hypodensity and loss of gray-white differentiation is seen, as it appears to predict death or permanent severe disability in infants with the shaking-impact syndrome.

The results of this study suggest that shaking-impact injuries in infants result in major morbidity, with only approximately one third of patients realizing a good outcome. Prospective long-term follow-up in this mobile population will be needed if we are to define prognostic indicators further and evaluate the effectiveness of therapeutic intervention.

Acknowledgements

The authors would like to thank Gary Magram, MD, and Faith Waxman, MSW, for their help in locating patients included in this project, and Chris Boardman, Biostatistics and Epidemiology, Children's Hospital of Philadelphia, for help with statistical analysis.

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