OUTCOME AND PROGNOSIS OF WHIPLASH SHAKEN INFANT SYNDROME: LATE CONSEQUENCES AFTER A SYMPTOM-FREE INTERVAL

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Whiplash shaken infant syndrome (WSIS) derives its name from Crowe's mechanical concept of whiplash (or recoil) injury (Olsnes 1989). The two main causes are motor accidents and child abuse. Shaking infants by the shoulders, trunk or limbs may cause hyperextension and/or hyperflexion of the head, which moves like the lash of a whip (Kleinman *et al.* 1989, Kleinman 1990, Brown and Minns 1993). In many respects, however, the concept of WSIS (Guthkelch 1971; Caffey 1972, 1974) is still hypothetical. Child-abuse traumas are almost never adequately witnessed, and the role of impact and acceleration–deceleration is still somewhat controversial (Ommaya *et al.* 1968, Duhaime *et al.* 1987, Brenner and Fischer 1988, Hadley *et al.* 1989, Alexander *et al.* 1990*a*). So far the literature is limited to descriptions of the short-term follow-up of about 100 cases of WSIS and 20 to 50 per cent of the children are symptom-free a year after the incident (Ludwig and Warman 1984, Frank *et al.* 1985, Sinal and Ball 1987, Brenner *et al.* 1989, Hadley *et al.* 1989, Wilkinson *et al.* 1989, Zepp *et al.* 1992). To our knowledge, no long-term study of a significant series of WSIS has been published, although the need for such studies was emphasized two decades ago (Caffey 1974, Oliver 1975). Non-medical experts in child abuse and even physicians and judges often uncritically apply the term 'shaking', which is very convenient. Nevertheless, despite the hypothetical aspects of WSIS, its usefulness is established: the mechanism does exist, and its significant representation in cases of abuse is highly plausible. Brown and Minns (1993) have recently published an excellent review of the literature on the whiplash shaking injury.

The present article reports on the long-term follow-up of 13 cases of WSIS. The analysis of our material benefits from three clinical advances of recent decades, two of which occurred after the papers by Guthkelch (1971) and by Caffey (1972). First, a better understanding has been acquired of the stages of the postnatal neurodevelopment, including growth and differentiation of neural tissue and synaptic stabilization (Evrard *et al.* 1992); this paper analyses how the severe lesions found in WSIS interfere with these developmental targets. Second, it has often been suggested that whiplash injury in adults and adolescents, when severe enough, is a complex cervical–encephalic syndrome carrying true neuropsychological consequences (Olsnes 1989, Kischka *et al.* 1991, Ettlin *et al.* 1992, Radanov *et al.* 1993). Such a derangement could also occur in shaken infants, in addition to the contusional and haemorrhagic lesions. Awareness of the sign-free interval between lesions and clinical conse-

TABLE I
Main diagnostic features on admission

Diagnostic features	*Present series (N=13)*	*Estimated percentage from previous studies**
History		
First child of both parents	12†	62[3]
Admission upon request of parents	9	Not found
Recent minor accident reported by parents as explanation	5	25–100[1,3,4,5,10]
Proven prior abuse, neglect or shaking	8	39–71[8,12]
Clinical findings		
Weight <50th centile	13	33[13]
Weight <10th centile	9	Not found
Traumatic tegumental lesions§	7	20–50[2,4,6,7,13]
Vomiting, anorexia	7	25–100[3,4,5,7]
Anaemia (<9g/DL haemoglobin)	9	50–100[5,11]
Coma or stupor	12	55–100[4,7,9,11]
Seizures upon admission	9	20–100[2,3,4,5,7,9,11,13]
Increased intracranial pressure**	11	55–75[3,9]
Grossly haemorrhagic CSF (subdural and/or lumbar)	12	75–83[3,4]

*Estimates from literature derive from Harcourt and Hopkins 1971[1], Zimmerman *et al.* 1979[2], Frank *et al.* 1985[3], Ludwig and Warman 1984[4], Alexander *et al.* 1986[5], Sinal and Ball 1987[6], Giangiacomo *et al.* 1988[7], Benzel and Hadden 1989[8], Brenner *et al.* 1989[9], Bruce and Zimmerman 1989[10], Hadley *et al.* 1989[11], Alexander *et al.* 1990*a*[12], Jaspan *et al.* 1992[13].
†13th case was a second child of same couple. Their first child died at nine months of age from acute and severe head injury due to abuse.
§Multiple ecchymoses or scars, scalp haematomas.
**Bulging fontanelle, conspicuous enlargement of cranial sutures, macrocephaly above 2 SD, paralysis of upward vertical gaze.

quences, the third clinical tool for our analysis, has only slowly emerged in the literature since 1955 (Lyon 1961, Lyon and Evrard 1987), and its neurobiological bases are still incompletely understood. Nonetheless, the study of the sign-free interval in WSIS is crucial for prognosis, retrospective diagnosis and rehabilitation strategy.

Subjects and method

The 'SOS-Enfants' programme was launched in 1979 as a multidisciplinary child-abuse programme involving a team composed of a paediatric neurologist, child psychiatrists, psychologists, social workers and a lawyer, all associated with the Hôpital Saint-Luc, University of Louvain Medical School at Brussels, Belgium. This programme is supported by the Office de la Naissance et de l'Enfance, a government agency for mothers and children, which provides resources for diagnosis, guidance, treatment, follow-up and prevention. We estimate that our programme recruits a 10th of all recognized severe abuses with major neurological lesions occurring in Belgium. Among the 449 cases in our child-abuse programme between 1982 and 1989, 22 cases were candidates for a diagnosis of WSIS. In this group, 13 cases displayed intra-ocular haemorrhages and intracranial lesions that satisfied all the criteria for a diagnosis of WSIS, including the absence of any other traumatic or non-traumatic mechanism (Table I). The shaking was clearly due to abuse in these 13 cases. The violent shaking was explicitly admitted by the parents in two cases. In five cases, the parents described an unconvincing recent traumatic event as an explanation, and we interpreted their effort as an indirect and partial avowal.

TABLE II
Ocular, nervous and pericerebral lesions at acute phase

Lesions	*Present series (N=13)*	*Estimated percentage from previous studies**
Retinal and preretinal bleeding	10[†]	68–100[3,4,6,7,9 12,14,15]
Pericerebral collections	12	20–72[5,10]
Arachnoidal haemorrhage	4	10–77[4,8,10,12,14,18]
Subdural haematoma	3	20–100[1,2,4 6,8,10,12,14,17,18]
Interhemispheric collection	2	41–61[2,10]
Early hypodense collection	4	25–40[10,11]
Bleeding in pericerebral collections previously constituted	3	Not found
Hemispheric lesions	8	
Intraparenchymal bleeding	1	5–30[3,8,11,18]
Acute oedema		65[8]
hemispheric	1	Not specified
bilateral	3	16–100[6,8,11]
local	4	Not specified
delayed	3	Not specified
Early contusional oedema	2	5–69[4,12,14]
Contusional tears[§]	1	100[17]
Traumatic strokes	2	50[2]
sylvian	1	Not specified
other territories	1	Not specified
Lesions due to associated strangulation	1	Not found
Brainstem lesions	1	Not found
Skull fracture	4	0–67[1–3,14,16–18]

*Estimates from literature derive from Harcourt and Hopkins 1971[1], Zimmerman *et al.* 1978[2], 1979[3], Ludwig and Warman 1984[4], Merten *et al.* 1984[5], Frank *et al.* 1985[6], Alexander *et al.* 1986[7], Cohen *et al.* 1986[8], Duhaime *et al.* 1987[9], Sinal and Ball 1987[10], Giangiacomo *et al.* 1988[11], Brenner *et al.* 1989[15], Alexander *et al.* 1990*a*[16], Jaspan et al. 1992[17], Brown and Minns 1993[18].

[†]Full eye examination was not performed in the one patient who died soon after admission.

[§]Visualized with neuropathological study. At acute phase, high-resolution cranial sonography, which is capable of detecting tears (Jaspan *et al.* 1992), was not performed.

All the WSIS cases were admitted in our service a few hours or days after the shaking. 12 of the infants were aged between three weeks and six months (mean 5.5 months), while the 13th infant was 21 months old at the time of the attack. One infant died during the acute period after the shaking, and a postmortem examination was performed. The multidisciplinary follow-up of the 12 other cases covered a period of four to 14 years after the last shaking (with a mean follow-up of 7 years 2 months). At a mean frequency of once a year, it included general, neurological and psychological examinations and a social evaluation. The Gesell (Gesell 1940), WISC (de Ajuriaguerraand Marcelli 1989) and Terman (Cesselin 1968) were among the most frequently used developmental tests.

The control group consisted of 10 age-matched children, admitted to our service between 1982 and 1989 for abuse without suspicion of WSIS (fractures of the skull, rib and long bones, cutaneous haematomas and failures to thrive). These children were assessed by our multidisciplinary team and submitted to a long-term follow-up. The intention was not to provide a complete statistical comparison, but to look at the effects of socio-economic status.

Results

The main purpose of this paper is to

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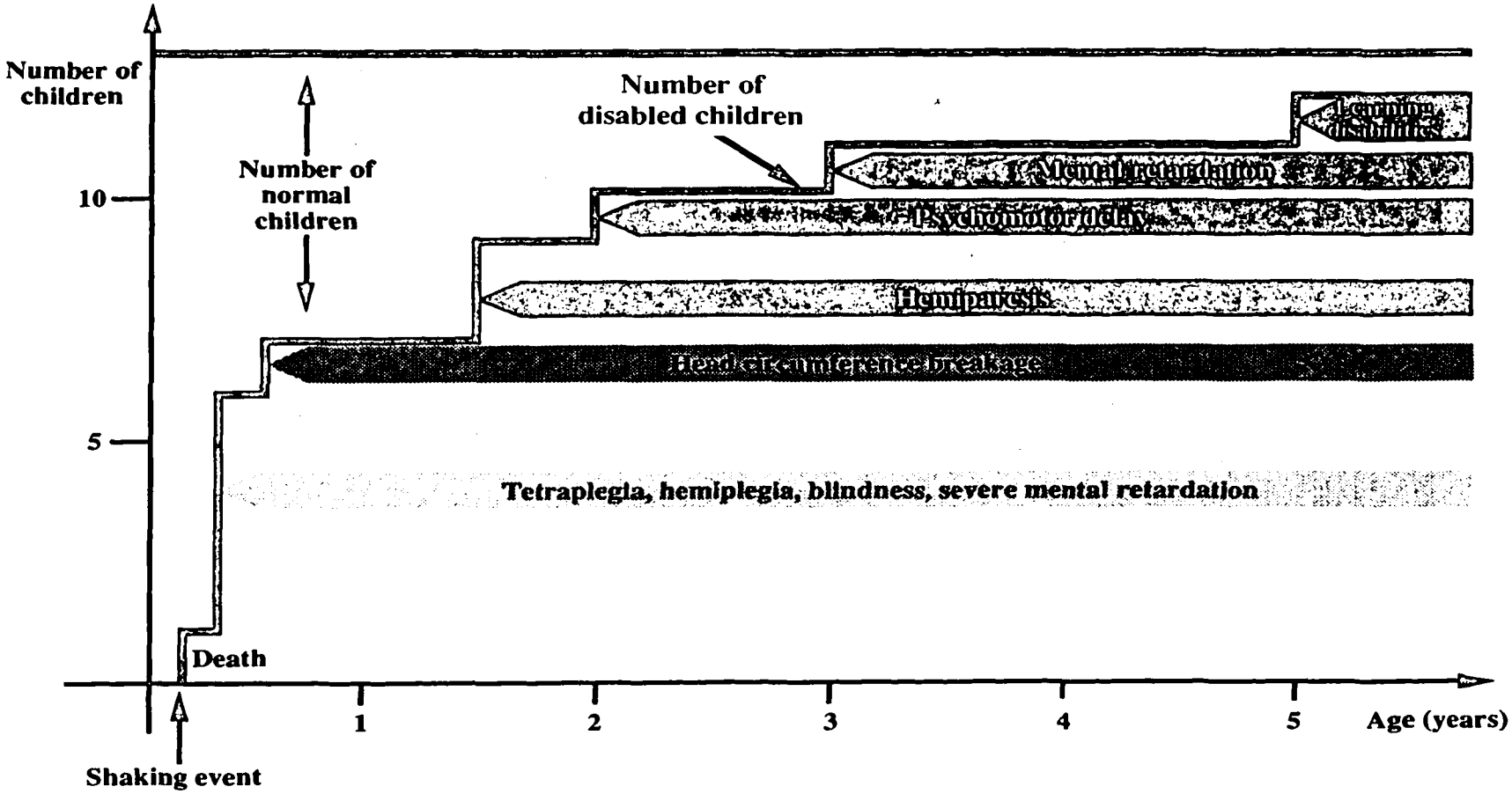

Fig. 1. *Chronology of appearance of developmental defects after WSIS.* Horizontal upper line *represents the total number of children.* Step curve *corresponds to number of disabled children; this number increases gradually with time. Difference between horizontal upper line and step curve corresponds to number of unaffected children remaining. Psychomotor delays and mental retardation are specifically indicated when they were first disability to appear; they were also conspicuous in cases when other neurological disabilities appeared earlier.*

TABLE III
Delayed lesions and permanent neurological and developmental signs with their interval of appearance

Clinical and radiological sequels	*Present series*	*Estimated percentage from previous studies**	*Interval of appearance in present series*
Mortality	1/13	12–62[4,6,12]	No interval
Blindness	2/12	12–20[4,7]	No interval
Visual impairment	2/12	6–40[4,7,8,12]	No interval
Neopallial atrophy	6/12	75–100[2,3,5,6,7,12]	Within 1mth
Isodense secondary pericerebral collection	1/12	Not found	Within 10 days
Tetraplegia, hemiplegia	3/12	5–40[4,7,8,12]	Within 1 mth
Transitory hypodense collection following arachnoidal disruption	1/12	25[7]	Within 1mth
Lesions at junction between grey and white matters (supposed to be tears)	3/5	100[12]	Within 3 wks
Impairment of head circumference growth	4/12	100[1]	Visible after 4 mths
Epilepsy	4/12	25–47[4,7,8]	Within 2 yrs
Hemiparetic status	2/12	12–15[7,8]	Within 1 yr
Psychomotor retardation	9/12	27–30[4,7]	Within 1 yr
Psychiatric problem	6/12	7[7]	Within 4 yrs
Mental retardation	11/12	93[7]	Within 5 yrs
Learning disabilities	11/12	Not known	Within 6 yrs

*Estimates from literature derive from Oliver 1975[1], Zimmerman *et al.* 1978[2], Zimmerman *et al.* 1979[3], Ludwig and Warman 1984[4], Frank *et al.* 1985[5], Cohen *et al.* 1986[6], Sinal and Ball 1987[7], Brenner *et al.* 1989[8], Hadley *et al.* 1989[9], Wilkinson *et al.* 1989[10], Alexander *et al.* 1990[11], Jaspan *et al.* 1992[12].

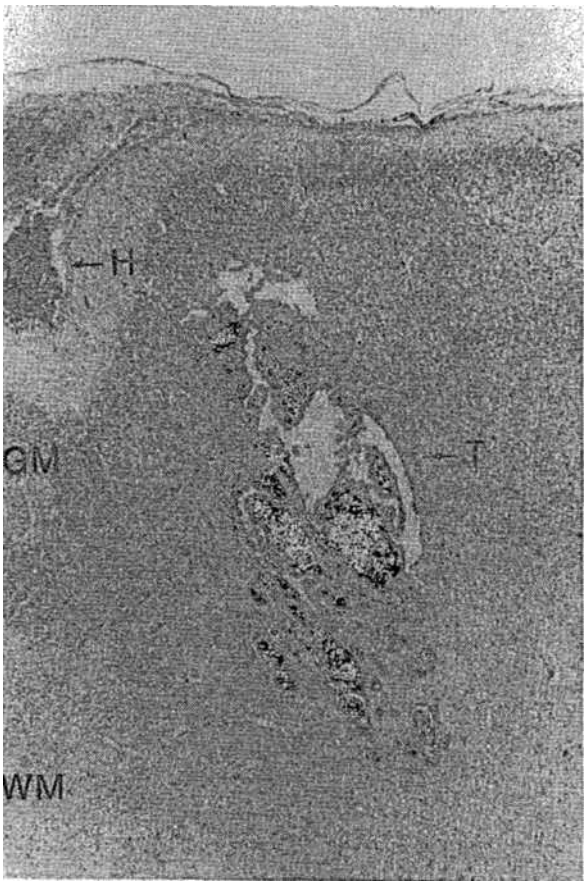

Fig. 2. *Cortical–subcortical tear and arachnoidal haemorrhage. Neuropathological study of infant who died in acute period after shaking event. Coronal section, parietal lobe. H=arachnoidal haemorrhage; T=cortical–subcortical tear; GM= neocortex. WM=white matter. 10 tears disseminated in brainstem and cerebral hemispheres were observed in examined blocks. When in cerebral hemispheres, most tears are typically cortical–subcortical, tangential or radial, as illustrated above. Mechanism of this type of tear is discussed in text. (Cresyl-violet staining of 20μm thick section., ×25.)*

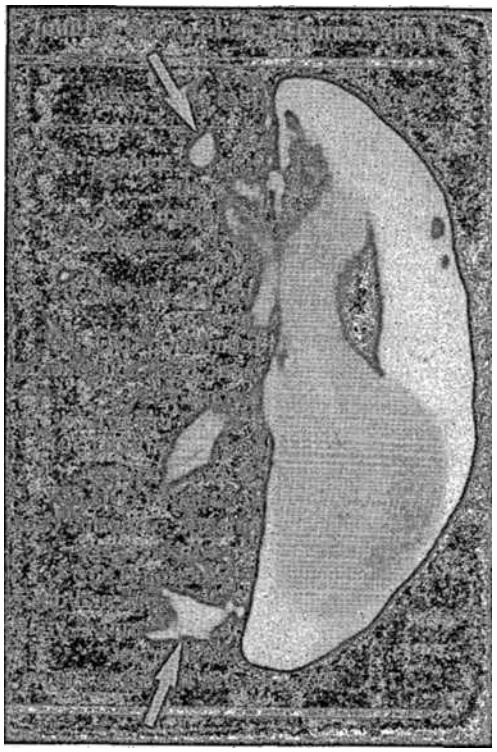

Fig. 3. *Magnetic resonance study (axial T_2-weighted-image) performed four years after shaking event. Whole left-hemispheric atrophy associated with sequels of tears (at junction between white and grey matter -arrows) is highly characteristic of strangulation during shaking assault.*

TABLE IV
Epilepsy in WSIS

	N
Convulsions at admissions	9/13
Long-term epilepsy	4/13
Early onset after initial convulsions	2
Late onset without initial convulsions	2*

*4 and 18 months delay

report the long-term follow-up of patients with WSIS. The clinical signs and laboratory examinations belonging to the early period of WSIS are reported in the previous section and in Tables I and II. They summarize the pathophysiological and lesional patterns in the acute period, which are crucial for the understanding of the long-term developmental outcome.

The chronological appearance of signs and lesions, during the follow-up and up to a maximum of 13 years, is shown in Figure 1 and Table III.

Within 24 hours of the shaking, the neuropathological data revealed a contusional tear in the brainstem (probably due to sectioning of the cerebral peduncles by the local dislocating forces at the axis of the lash of the whip) and contusional tears in the brain hemispheres (which suggests a 'creeping' of the immature cortex with axonal fractures over the underlying white matter). Such lesions are probably induced by out-of-phase anteroposterior displacements of the neopallial layers, which in the developing brain display differential properties of elasticity and reaction to acceleration between the cortex and the concentric white matter strata. The neuropathological observation confirmed other previously reported lesions and mechanisms: the shearing of blood vessels within and around the brain with massive subarachnoidal bleeding and acute and generalized brain oedema, sparing the brainstem (Lindenberg and Freytag 1969, Calder *et al.* 1984, Leestma 1988, Hadley *et al.* 1989, Brown and Minns 1993). The main clinical pattern in the early post-shaking period of the 12 cases without neuropathological control was due to various causes: the shearing of brain, pericerebral and ocular vessels; brain oedema; sectioning forces of parenchyma due to acceleration of adjacent and adherent neural compartments with differential elastic and mechanical properties; local contusions; and contusions by 'contrecoup'. Our report of an association between shaking and strangulation stresses the asymmetrical consequences of strangulation when due to unilateral carotid compression by one thumb with incomplete blood compensation through the polygon of Willis (Feldman and Simms 1980, Bird *et al.* 1987) (Fig. 2).

Skull fractures involving parietal or occipital bones were associated with other criteria of WSIS in four of 13 cases. One fracture was stellate; none was depressed or growing.

During the month after the traumatic shaking event, blindness, visual impairment, tetraplegia or early detectable hemiplegia, neopallial cortical-subcortical atrophy, secondary collections and

TABLE V
Long-term deficits in group without sign-free interval

Child	*Actual clinic status*
1	Died during the acute period after the shaking
2	Vegetative status Spastic tetraplegia Microcephaly (-4SD) Total blindness Severe epilepsy for four yrs after trauma
3	Total blindness Mental retardation (DQ 50) Hyperkinetic behaviour and depressive status
4	Spastic tetraparesis Bilateral visual impairment Mental retardation (DQ 50) Cranial growth breakage
5	Mental retardation (DQ 60) Major epileptic syndrome (Lennox-Gastaut) Cranial growth breakage
6	Mental retardation (DQ 74) Pervasive developmental disorder Severe epilepsy during 4 yrs after trauma Cranial growth arrest
7	Described in legend to Figure 2

DQ = developmental quotient (Gesell).

TABLE VI
Long-term deficits in group with sign-free interval

Child	*Neurological examination*	*Cognitive development*	*Behavioural problems*	*Educational level*	*Rehabilitation needs*
1	Normal	IQ 77		Failure with 3 yrs retardation	Speech theapy
2	Right hemiparesis	IQ 80 (Verbal 79, Performance 85)	Anxiety disorder	Failure with 2 yrs retardation	Speech, psychological and neurodevelopmental therapy
3	Normal	DQ normal		Adequate level	
4	Left hemiparesis	IQ 60 (Verbal 70, Performance 56)	Pervasive developmental disorder, hyperkinetism	Special educational school	Speech, psychological and neurodevelopmental and occupational therapy
5	Normal	IQ 80 (Verbal 69, Performance 95)	Pervasive developmental disorder	3 yrs retardation	Psychological therapy
6	Normal	IQ 53 (Verbal 67, Performance 47)		Special educational school	Speech, and neurodevelopmental therapy

IQ = Intelligence Quotient (WISC, Terman)

TABLE VII
Long-term outcome in 'control' group

Feature	*Cases with cranial fracture (N=14)*	*Cases without cranial fracture (N=6)*
Neurological examination	4 normal	6 normal
Cognitive development	4 normal	4 normal 2 with mild mental retardation
Behavioural problems	4 with none	3 with none 3 requiring psychological therapy
Educational level	4 adequate	4 adequate 2 at special educational school

lesions at the junction between the grey and white matter (assumed to be tears) appeared in half of the cases of this series.

During the next four months, cranial growth ceased in five cases (Fig. 3) and delayed epilepsy appeared in one case (Table IV).

Regarding the long-term follow-up, the 13 cases reported in this series can be divided into two groups according to outcome. The group without a sign-free interval (N=7) remained severely and permanently abnormal from the time of the shaking; one died. Their long-term deficits are listed in Table V.

The group with a sign-free interval (N=6) apparently recovered fully after the shaking, and their detailed neurological, psychomotor and general examinations were quite normal when performed two months after the shaking (Table VI). All but one of the children in this group left the normal cohort and became disabled after a delay ranging between six months and five years (see Fig. 1). This interval was free from any detectable abnormal clinical sign: all the children in this study were evaluated at three-month intervals by paediatric neurologists and psychologists aware of the initial diagnosis and alert to developmental abnormalities. Delayed hemiparesis was detected around 18 months of age, 12 months after the shaking, in two cases. Psychomotor retardation, especially in language, adaptability and social behaviour, became apparent around 24 months in one case. Mental retardation was diagnosed in five of the six cases within five years of the event. All five required special education. Severe behavioural disorders appeared in three of the six cases: severe anxiety disorder (N=1), pervasive developmental disorder (N=2), and severely hyperkinetic behaviour (N=1). Only one child, now five years old, seemed to be clinically normal and problem-free.

Discussion

The short-term follow-up of about 100 cases of WSIS has been previously published in the literature (Ludwig and Warman 1984, Frank *et al.* 1985, Sinal and Ball 1987, Brenner *et al.* 1989, Hadley *et al.* 1989, Wilkinson *et al.* 1989, Zepp *et al.* 1992). The number of our cases reported in this long-term follow-up study is similar to the average number described in the seven series mentioned above.

Impairment of conciousness, seizures, intracranial hypertension, and retinal and pericerebral bleeding were the initial or immediate signs of WSIS in this series and are also reported in the literature (Caffey 1972, 1974, Ludwig and Warman 1984, Sinal and Ball 1987, Brenner *et al.* 1989, Wilkinson *et al.* 1989, Alexander *et al.* 1990*a*, Brown and Minns 1993, Committee on Child Abuse and Neglect 1993). Additional data from our series, however, show that it is not always easy to make an early diagnosis. In most cases, children are admitted to the emergency room by request of the parents, who report a history of very benign traumatic events incompatible with the condition of the infant (Kravitz *et al.* 1969, Helfer *et al.* 1977, Billmire and Myers 1985, Joffe and Ludwig 1988, Rivara *et al.* 1988, Williams 1991, Duhaime *et al.* 1992). Two-thirds of the cases had a history of

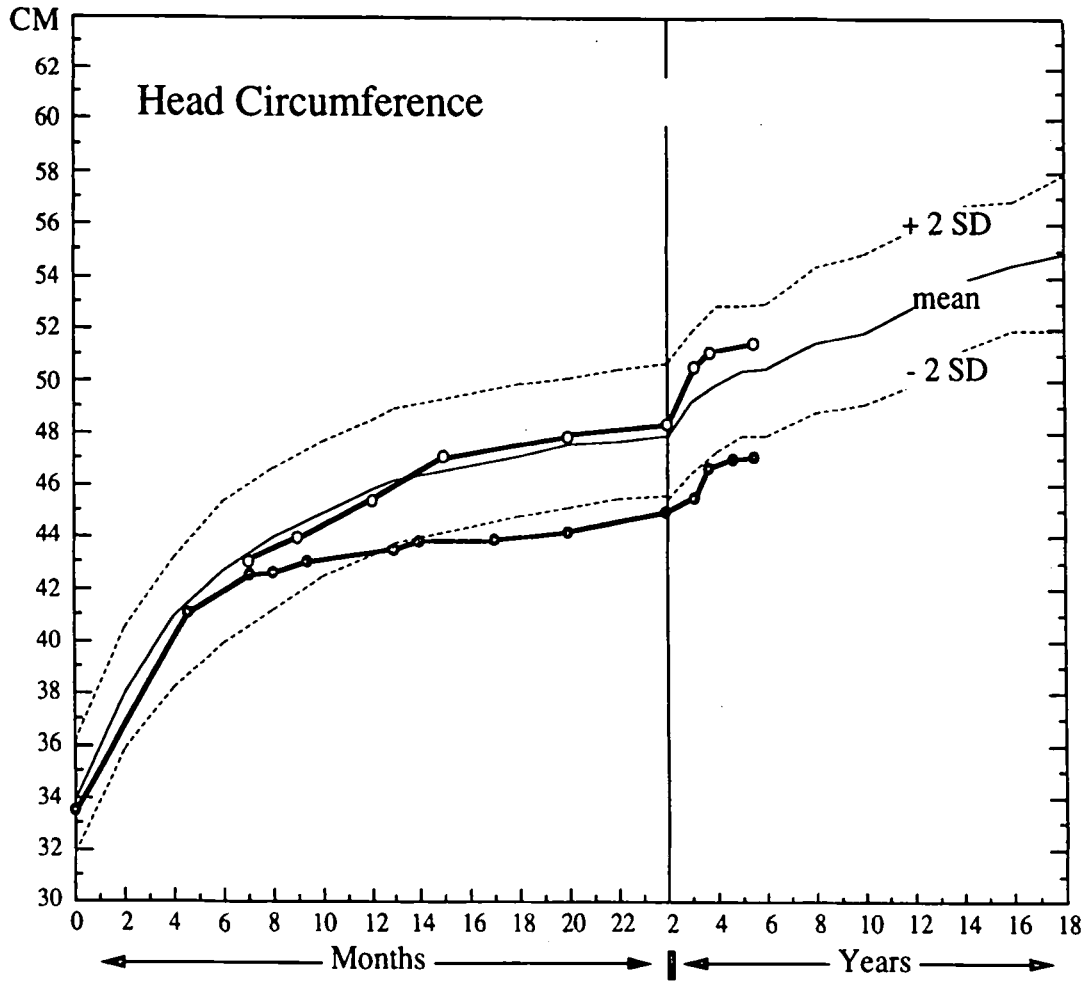

*Fig. 4. Cranial growth curves in pair of twins. One (*lower curve*) was severely shaken at four months of age. Hemiplegia was detected 10 days after event; mental retardation was detected at age of 22 months; IQ at five years was 80. History of her twin sister (*upper curve*) was uneventful, with perfectly normal examination and development and IQ of 110. Head circumference curves illustrate arrest of cranial growth occurring in WSIS: in five cases, it appeared within 4 months and brain growth did not restart for 12 months; it reached 4SD in one case, 2SD in three cases, and -1SD under initial centile of head circumference in one case (curve from Nelhaus 1968).*

abuse or neglect, and most of the other cases had other lesions related to abuse (tegumental lesions and bone fractures) (Alexander *et al.* 1990*b*). In all the cases in our series, shaking was clearly due to violent behaviour within an abusive context, even if the parents tried (consciously or not) to convince the physician and perhaps themselves otherwise by offering a mild and credible history. However, Guthkelch (1971) reported that "a good shaking", is felt, at least by British parents, to be socially more acceptable and physically less dangerous than a blow on the head or elsewhere.' The same clinical features may also be caused accidentally by dangerous games (like throwing a baby up in the air and trying to catch it) or inadequate folk practices (Guarnaschelli *et al.* 1972), rather than resulting from deliberate abuse. Early convulsions occur much more frequently than after any other traumatic conditions at this age and confirm the severity of the lesions (Annegers *et al.* 1980). In addition to the classic interhemispheric and parieto-occipital subdural haematomas often reported in WSIS (Zimmerman *et al.* 1978, Levin *et al.* 1989, Kleinman 1990), early CT-hypodense pericerebral collections with intracranial hypertension were found in seven of our cases (Giangia-

como *et al.* 1988). All these seven cases with hypodense collections benefited from a surgical drainage, which in all cases demonstrated the presence of blood.

The sequelae detectable during the first four months have been described in several papers (Oliver 1975, Ludwig and Warman 1984, Frank *et al.* 1985, Sinal and Ball 1987, Brenner *et al.* 1989, Hadley *et al.* 1989, Wilkinson *et al.* 1989), and were present in half of the cases of our series (Table III and Fig. 4). Only Oliver (1975) has reported previously an impairment of cranial growth following shaking. This is a conspicuous feature in one-third of our cases, with a significant and abnormal decrease in cranial growth—*i.e.* head circumference curve dropping to two to four standard deviations less than the initial value (Jaffe *et al.* 1992) occurring within four months of the shaking. Several mechanisms are probably involved in this microcephalic evolution. Areas of necrosis and more diffuse neuronal death including cytotoxic oedema can be provoked by contusion and/or by traumatic vascular accidents (see Fig. 2). Neuronal death can be secondary to an excitotoxic post-traumatic disorder (Lipton and Rosenberg 1994). Post-traumatic encephalopathy following the shaking can also severely interfere with the very active neurodevelopmental processes of dendrogenesis, axogenesis, synaptogenesis and synaptic stabilization, gliogenesis and myelination that occur during the first postnatal year. These ontogenic events are responsible for dramatic brain growth, with the brain's weight being increased by a factor of 1.5 between 5.5 and 12 months of age (Yakovlev and Lecours 1967, Huttenlocher 1979, Changeux 1983, Changeux and Dehaene 1989, Huttenlocher 1990, Evrard *et al.* 1992). Each case with head-circumference growth deceleration as defined above had very severe neurological sequelae and belonged to the first group (without a free interval). In our series, the most severe and constant predictive factor of long-term neurological sequelae is the deceleration of brain growth; retinal haemorrhage and intracranial lesions are two other reliable predictors of permanent neurological deficits (Sinal and Ball 1987, Wilkinson *et al.* 1989).

The comparative roles of dissecting lesions *vs* direct and 'contre-coup' contusional lesions in WSIS have been the focus of much debate in the literature (Ommaya *et al.* 1968, Duhaime *et al.* 1987, Brenner and Fischer 1988, Hadley *et al.* 1989, Alexander *et al.* 1990*a*). The first lesional type, due to acceleration/deceleration, is specific to WSIS; the second lesional type, due to head impact (sometimes revealed by a skull fracture) can be associated with WSIS but is not specific to it; in this respect, the term 'contusional tear' is confusing, because it belongs to the first lesional type. In this study, the group of patients with immediate evidence of neurological impairment (the group 'without a sign-free interval') includes three infants without direct nor 'contre-coup' contusion and without skull fractures, and three infants with fractures and contusions due to impact. Shaking only is sufficient to produce severe and permanent neurological and psychological sequelae as already described (Ludwig and Warman 1984, Frank *et al.* 1985, Sinal and Ball 1987, Brenner *et al.* 1989, Hadley *et al.* 1989, Wilkinson *et al.* 1989, Alexander *et al.* 1990*a*, Zepp *et al.* 1992). It does not exclude the common-sense hypothesis that the association of lesions of both types can worsen the outcome.

Our series reports long-term data demonstrating the existence of a sign-free interval for several neurological and mental manifestations. When appearing with a sign-free interval, hemiparesis is the first condition that can be detected clinically. The clinical onset of hemiparesis is similar to that of cerebral palsy of perinatal origin (Lyon 1961, Lyon and Evrard 1987). At 18 months of age, after a sign-free interval, psychomotor retardation may be revealed by deficits in the emergence of language, constructional ability and spatiotemporal exploration. In our study, major behavioural disorders were the next clinical sequelae to appear in the group with a sign-free interval: they become detectable three to six years after the shaking and were always associated with neurological signs in this series (see Table VI). It is difficult to say exactly what may have been due to the

shaking itself and what was caused by the general history of abuse. Goldson (1991) reviewed the different problems following 'child maltreatment' and found a higher frequency of anxiety and depression, low self-esteem and difficulty forming social relationships in the abused population.

In all but one case in the group with a sign-free interval, IQ dropped below 80 between the ages of three and six years old. This almost invariable intellectual and neuropsychological deficit once described previously (Oliver 1975) requires special education and rehabilitation. Multiple tiny but obvious residual lesions were found on magnetic resonance imaging, particularly at the border zone between neopallial grey and white matter, which suggests sequelae of tears at this level with disturbances of the associative pathways (Fig. 5). Disturbances of dendrogenesis, synaptogenesis and synaptic stabilization could also be hypothesized and checked by positron emission tomography (Wanet-Defalgue *et al.* 1988). For ethical reasons, we did not perform such studies. Remote neopallial consequences of brainstem lesions have been hypothesized to explain several deficits in whiplash injuries in adult humans and monkeys (Ommaya *et al.* 1968, Olsnes 1989). Such mechanisms have never been evoked in WSIS but they may be a possibility. Other aetiological factors may explain the high incidence of mental retardation in our WSIS series. Unfavourable prenatal genetic or epigenetic factors may exist in abuser populations. However, the thorough clinical study of all the children and the families of this series did not disclose any prenatal factors that could have provoked mental retardation in these cases. Unfavourable postnatal environmental factors must be taken into consideration, as suggested by the fact that the average birthweight of our subjects was at the 30th centile, while the average bodyweight at admission for shaking was below the 10th centile. Deficits in the quality of parenting, the emotional environment and the affective relationship constitute a crucial prognostic factor for cognitive development in child abuse, as revealed by our control cases of abuse without WSIS (Table VII), but these factors are very difficult to test statistically (Elmer and Gregg 1967, Gregg and Elmer 1969, Morse *et al.* 1970, Friedman and Morse 1974, Smith and Hanson 1974, Oates 1984, Goldson 1991).

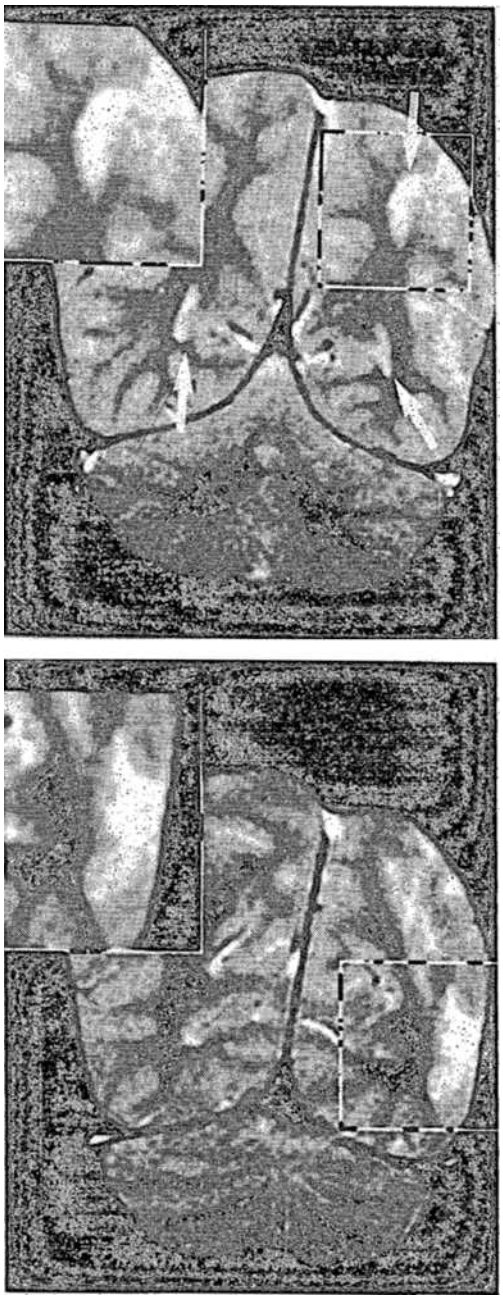

Fig. 5. *Magnetic resonance study (coronal T_2-weighted images) performed seven years after shaking event. Three hyperdense areas scattered in both hemispheres could signify reactional gliosis around tears* (upper). *More posterior view shows contusional cortical–subcortical sequel* (lower)

Conclusion

Several short-term follow-up series of WSIS have reported a prognosis of disabilities in half of the survivors. In this study, long-term follow-up enabled us to demonstrate that severe developmental consequences appear after a sign-free interval in the cases that had seemed to escape short-term sequelae. This makes the long-term prognosis of WSIS severe in almost all cases. It emphasizes the importance of educating parents, future parents and child-minders about the danger of shaking babies, both abusively and non-abusively. Several pathophysiological and therapeutic aspects have also emerged, including the importance of detecting isodense and hypodense pericerebral collections in the acute period, the delay of appearance of brain-growth deceleration after shaking, and the usefulness of late MRI for the detection of traumatic tears. Moreover, the notion of a sign-free interval carries medico-legal implications: no final favourable prognosis can be given before the age of six years old, or perhaps even later.

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SUMMARY

Long-term follow-up five to 13 (mean seven years) of 13 cases of whiplash-shaken-infant syndrome (WSIS) demonstrated long sign-free intervals. Full clinical appearance of neurological deficits takes four months for the interruption of brain growth, six to 12 months for lesions of the central nervous system long pathways, up to two years for epilepsy, and three to six years for behavioural and neurospychological signs. In our series, WSIS ocurred at a mean postnatal age of 5.5 months and caused intracranial, retinal and preretinal haemorrhages, intracranial haematomas, oedema, contusional tears, and developmental disturbances interfering with the growth and differentiation of neural tissue and with synaptic stabilisation. These mechanisms account for the long sign-free interval that makes its impossible to formulate a precise and final neurological prognosis before the age of school entrance. Only one of our patients seems to have remained normal even several years after the shaking.

RÉSUMÉ

Devenir et pronostic du syndrome des enfants secoués (WSIS): conséquences tardives après un intervalle libre

Le suivi à long terme (cinq à treize ans avec une moyenne de sept ans) de 13 enfants secou´s a montré de longs intervalles libres. L'expression complète des déficits neurologiques exigeaient quartre mois pour l'interruption de la croissance cérébrale, de six à 12 mois pour les lésions des voies longues du système nerveux central, jusqu'à deux ans pour l'épilepsie et de trois à six ans pour les signes comportementaux et neurospychologiques. Dans notre série le WSIS apparaissait à un âge postnatal moyen de 5,5 mois et provoquait des hémorraiges intracrâniennes, rétiniennes et prérétiniennes, des hématomes intracrâniens, de l'oedème, des cicatrices de contusion, et des perturbations de développement interférant avec la croissance et la différentiation du tissu neural et avec la stabilistation synaptique. Ces mécanismes rendent compte d'un long intervalle libre qui rend impossible un pronostic précis et final avant l'âge d'entrée à l'école. Un seul de nos patients semble être resté normal même plusieurs annéss après les mauvais traitements.

ZUSAMMENFASSUNG

Outcome und Prognose beim Whiplash-Shaken-Infant-Syndrom (WSIS): Späte Konsequenzen nach einem Symptom-freien Interval

Bei den Langzeituntersuchungen (5–13, im Mittel 7, Jahre) an 13 Kindern mit Whiplash-Shaken-Infant Syndrom (WSIS) zeigten sich lange Symptomfreie Intervalle. Das volle klinische Bild der neurologischen Ausfälle zeigt sich nach vier Monaten mit der Unterbrecheung des Hirnwachstums, in 6–12 Monaten treten Läsionen der langen Bahnen des Zentralnerven-systems, in bis zu zwei Jahren Epilepsie und in 3–6 Jahren Verhaltensund neuropsychologische Störungen auf. Bei unseren Patienten trat das WSIS im mittleren postnatalen Alter von 5.5 Monaten auf und es fanden sich intrakranielle, retinale und periretinale BLutungen, intracranielle Hämatome, Ödeme, Kontusionszeichen und Entwicklungsveräanderungen, wodurch die Differenzierung von

Nervengewebe und die Stabilisierung der Synapsen beeinträchtigt wurden. Durch diese Vorgänge sind die langen Symptom-freien Intervalle zu erklären und dadurch ist es unmöglich, eine genaue und endgültige Prognose vor dem Schulalter zu stellen. Nur einer unserer Patienten scheint sogar mehrere Jahre nach dem Schütteln normal gblieben zu sein.

RESUMEN

Curso y pronóstico de niños con síndrome de trallazo (ST): consecuencias tardías tras un intervalo mudo

Un seguimiento a largo término (5–13 años con un promedio de 7) de 13 casos con síndrome del trallazo (ST) demostró la existencia de largos intervalos silenciosos. El plazo de aparición de claros sígnos de déficits neurológicos es de cuatro meses para la interrupción del crecimiento cerebral, de 6 a 12 meses para las lesiones de vías largas del SNC, hasta dos años para la epilepsia y de 3 a 6 años para los signos neuropsicológicos y del comportamiento. En nuestras series el ST tuvo lugar a una edad promedio postnatal de 5.5 meses y causó hemorragias intracraneales, retinianas y preretinianas, hematomas intracraneales, edema, desgarros contusionales y alteraciones del desarrollo que interferian con el crecimiento y diferenciación del tejido neural y la estabilización sináptica. Estos mecanismos explican el largo intervalo sin signos clínicos que hace imposible hacer un pronóstico neurológico final antes de la edad escolar. Sólo uno de nuestros pacientes parece que se mantuvo normal incluso varios años después de la sacudida.

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