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Clinical case / Cas clinique

Long-term outcome of the shaken baby syndrome and medicolegal consequences: A case report

Conséquence à long terme du syndrome du bébé secoué et incidence médicolégale : à propos d'un cas

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Abstract

Introduction. – Studies of long-term outcome of the shaken baby syndrome (SBS) are scarce, but they usually indicate poor outcome.
Objectives. – To describe long-term outcome of a child having sustained a SBS, to ascertain possible delayed sequelae and to discuss medicolegal issues.
Methods. – We report a single case study of a child having sustained a SBS, illustrating the initial clinical features, the neurological, cognitive and behavioural outcomes as well as her social integration.
Results. – The child sustained diffuse brain injuries, responsible for spastic right hemiplegia leading to secondary orthopaedic consequences, as well as severe cognitive impairment, worsening over time: the developmental quotient measured at 15 months of age was 55 and worsened as age increased. At 6 years and 8 months, the child's IQ had fallen to 40. Behavioural disorders became apparent only after several months and precluded any social integration. The child eventually had to be placed in a specialised education centre at age 5.
Discussion and conclusion. – The SBS has a very poor outcome and major long-standing sequelae are frequent. Cognitive or behavioural sequelae can become apparent only after a long sign-free interval, due to increasing demands placed on the child during development. This case report confirms severity of early brain lesions and necessity for an extended follow-up by a multi-disciplinary team. From a medicolegal point of view, signaling the child to legal authorities allows protection of the child, but also conditions later compensation if sequelae compromise autonomy.

Keywords: Non accidental head injury; Shaken baby syndrome; Prognosis; Diagnosis; Child protection

Résumé

Introduction. – Les études rapportant un suivi à long terme après syndrome du bébé secoué (SBS) sont rares mais suggèrent toutes un pronostic sombre.
Objectif. – Décrire l'évolution clinique à long terme d'une enfant victime d'un SBS, afin d'objectiver la gravité et l'apparition progressive des séquelles et de discuter les implications médicolégales.
Méthode. – Nous rapportons un cas unique illustrant le tableau clinique initial d'une enfant victime d'un SBS, son évolution neurologique, cognitive, comportementale et son insertion sociale.
Résultats. – L'enfant présentait des lésions cérébrales diffuses, responsables d'une hémiplégie spastique compliquée de troubles neuro-orthopédiques secondaires et d'un retard sévère du développement psychomoteur, se majorant avec le temps : le quotient de développement évalué à 55 à l'âge de 15 mois s'aggravait avec l'âge. Enfin, à l'âge de six ans et huit mois, le quotient intellectuel avait chuté à 40. Des troubles du

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comportement d'apparition retardée et d'aggravation progressive ont entravé toute tentative d'insertion, l'enfant étant finalement intégrée dans un établissement médicoéducatif à l'âge de cinq ans.
Discussion et conclusion. – Le SBS peut laisser des séquelles majeures et définitives, d'installation parfois très différée, par altération des capacités d'apprentissage, devenant plus évidentes lorsque les exigences environnementales envers l'enfant augmentent. Cette observation confirme la gravité des conséquences de lésions cérébrales précoces et la nécessité d'un suivi très prolongé par une équipe multidisciplinaire. Au plan médicolégal, le signalement judiciaire initial permet, outre la protection de l'enfant, la reconnaissance de l'infraction pénale par une expertise judiciaire, conditionnant ainsi une indemnisation ultérieure en cas de séquelles.

Mots clés : Syndrome du bébé secoué ; Pronostic ; Diagnostic ; Signalement ; Réparation médicolégale

1. English version

1.1. Introduction

Non-accidental head injury is the most frequent cause of mortality and morbidity in neonates [1–3,15]. In its minimal form, shaken baby syndrome (SBS) consists of subdural haematoma:

- in the absence of any concurrent, accidental injury reported by the baby's parents or legal guardians;
- following a minor accident which is incompatible with the extent of the damage.

In 75 to 90% of cases, the subdural hematoma is associated with uni- or bilateral retinal haemorrhage [3,32,36]. SBS is the most severe example of non-accidental head injury trauma and was only described relatively recently: in 1860, Tardieu described a series of 32 cases of battered children, including 18 with pericerebral bleeding at autopsy [49]. In the United States in 1940, Caffey noticed the association between long bone fractures and subdural haematoma in the absence of external signs of maltreatment and (in 1972) coined the name ''whiplash shaken baby syndrome'', which unambiguously identified shaking as the causal mechanism. Caffey's definition of SBS encompassed a set of symptoms, including the presence of a subdural haematoma or sub-arachnoid haemorrhage and retinal haemorrhage which contrasts with the absence of external signs of head injury or the presence of only minor signs [18,19]. At the same time, Guthkelch reported observations of babies in England having suffered subdural haematoma through shaking [28]. This syndrome can occur in any sociocultural milieu and mainly affects children under the age of 12 months (and under 6 months in particular). Boys are consistently more involved than girls (with a typical gender ratio of 2:1), despite the lack of any real explanation for this phenomenon. Although SBS is not infrequent (with a reported incidence of 24 to 30 cases per 100,000 inhabitants per year in the United States and in Scotland [10,34]), it is not always diagnosed, and this is true even of the most severe forms [31]. In France, the incidence of SBS (180 to 200 diagnosed cases per year) is certainly underestimated [46]. SBS can lead to death or permanent handicap [30,46]. The prognosis for shaken babies is not well characterized and there are few studies on the outcomes (and especially the long-term outcomes) in these children. Moreover, the few published studies have used very different recruitment modes and methodologies. Nevertheless, all cite a poor vital, neurological and cognitive prognosis. Hence, the reported mortality varies between 15 [43] and 38% [2], with a median at 20 to 25% [3,12]. In terms of neurological and cognitive sequelae, one can observe severe psychomotor development delays, spastic quadriplegia, severe motor disorders, epilepsy, cortical blindness, microcephalus and severe cortical and subcortical atrophy [3,11–13,16,46,48]. A recent literature review on the prognosis for non-accidental head injury [12] covered 489 cases; it indicated a mortality rate of 21.6% and a morbidity rate in the survivors that varied from 59 to 100% (depending on the series' recruitment mode) and averaged 74%. This means that only a quarter of the children were free of sequelae. The prognosis for SBS is significantly correlated with the initial Glasgow score, the presence of major retinal haemorrhage, the presence of a cranial fracture and the extent of the parenchymatous lesions identified in the first 3 months [17].

Over the years, many SBS victims have been hospitalized in our Physical Medicine and Rehabilitation service, which specializes in acquired brain damage in the child. The huge majority of cases are referred to us by Necker Children's Hospital, which has been the Paris Île-de-France region's main hospital for referral of children with severe head injuries since 1994 [37].

We published an initial, retrospective study on 28 SBS victims subsequently hospitalized in our service between 1995 and 1999 [14]. All these children have since been followed prospectively. The extent of the neurological damage became apparent very early on, after a median follow-up period of 18 months (range: 6 to 43 months) in children with a median age of 2 (range: 1 to 4 years). Brain atrophy was evidenced by a change in the head circumference curve. A break-point in the curve was seen in all children, with an average loss of 2.2 standard deviations (S.D.) for the group as a whole. The average change was 0.5 S.D. for those who seemed otherwise unaffected and 4.4 S.D. for the most severe cases, with one extreme value of 8 S.D. – meaning that brain growth stopped completely in this child. We identified convulsive status as a major prognosis factor.

Here, we report in detail on the case of a female patient with 6 years of follow-up, in order to illustrate the long-term impact of SBS on a child's development and the resulting legal consequences. We hope that this clinical case history will