SHAKEN BABY SYNDROME: OCULAR AND ASSOCIATED SYSTEMIC FINDINGS, MEDICAL AND SURGICAL MANAGEMENT, AMBLYOPIA PREVENTION

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Traumatic sequelae of nonaccidental injuries occurring in infants and young children as a consequence of violent shaking are called shaken baby syndrome. Synonyms are whiplash shaken infant syndrome, battered child syndrome or child abuse syndrome. It commonly results in intracocular and intracranial hemorrhages. As the mortality rate is 15% it is important to recognize this form of child abuse.

The typical victim of shaken baby syndrome is a male infant younger than six months of age who is alone with the perpetrator at the time of injury. The injury is unrelated to race, gender, socioeconomic status, or education. An infant is more likely to suffer from intracranial and intracocular bleeding as a result of shaking, because the head is proportionately larger and heavier relative to the body than that of an older child or adult and is stabilized less well by neck muscles. The condition is still underdiagnosed to a significant extent: Less than 5% of abused children become known to the authorities. On the other hand, in central Europe, 3.5% of parents confessed to have used such violence on their children that it might have resulted in severe injury. 10% of children that were admitted to hospital because of an injury show evidence of physical violence. 2/3 of the abused children are babies.

The diagnosis of child abuse requires a high index of suspicion: child abuse is usually not reported on. Characteristically a history of shaking is lacking. In addition, the shaken infant may present with minimal external signs of trauma.

In child abuse, the eye is involved as presenting sign in 4-6% of cases. While after accidental head injuries nearly all babies (≤3 years of age) had normal funduscopic examinations, the majority of babies with nonaccidental head injuries were found to have varying degrees of retinal hemorrhages. Retinal hemorrhages occur in 11-23% of all physically abused children and in 50-80% of shaken babies. Indirect ophthalmoscopy shows intracocular hemorrhage in various locations - subretinal, intraretinal, preretinal (subhyaloid), and intravitreal, concentrated in the posterior pole region and usually bilateral. The amount of intracocular blood correlates with the degree of acute neurologic damage. Cotton-wool spots, white-centered hemorrhages, macular edema, papilledema, and retinoschisis are less common.

Computed tomography or magnetic resonance imaging detects subarachnoid or intracerebral blood, cerebral edema, and cerebral atrophy. Elevated intracranial pressure often is present. A variety of neurologic symptoms, ranging from irritability and lethargy to seizures, coma, and death, can occur. In infants retinal hemorrhages are mostly seen post partum or in shaken babies. 24 hours after a normal birth, 19 to 32% will show retinal hemorrhage in one or both eyes.

By 72 hours the rate drops to 11-13%. These hemorrhages usually completely resolve within 6 weeks without further treatment. However, if intravitreal of retinal hemorrhages are seen later on, one may not conclude, that this is solely related to shaken baby!

The suspect of a child abuse requires the initiation of a multidisciplinary approach and a careful documentation of all findings. The documentation should include photography and ultrasonography. When violence is suspected, a careful general examination has to be performed to evaluate other signs of trauma also, when other symptoms are lacking. The ophthalmologists will be routinely consulted also when external signs were found because of the pathognomonic appearance of the fundus. Because of its medicolegal consequences, other causes of retinal hemorrhages have to be carefully ruled out.

Treatment is most commonly supportive. After the abuse has stopped, hemorrhages and other acute changes resolve within several months. Late manifestations include perimacular retinal folds, chorioretinal atrophy or scarring, optic atrophy, and retinal detachment. A retinal detachment or a nonclearing vitreous hemorrhage may require surgical treatment. Enzyme assisted vitrectomy may facilitate to create the posterior vitreous detachment which otherwise is very difficult in these infants. The clinical course of shaken baby retinopathy ranges from complete clearing, to severe visual loss (secondary to optic atrophy), or macular scarring. Postmortem examination of the optic nerves in shaken babies often reveal perineural hemorrhage, which may contribute through a mechanism of nerve fibre compression to the development of optic atrophy in survivors.

A 50% incidence of gazing disorders is reported in shaken baby syndrome reflecting nervous system insults. Even without evidence for retinal damage visual disturbance may occur. In 1959, Hollenhurst reported visual loss in 35% of 23 children with subdural or subarachnoidal hemorrhage who showed no evidence of retinal damage.

As a proportion of these visual losses is related to secondary amblyopia, it is mandatory to carefully observe the victim regularly to prevent the development of an amblyopia.