

MEDICINE

IDENTIFYING MUNCHAUSEN SYNDROME BY PROXY

—by Alex V. Levin

Editor's Comments

—by Martin Finkel

Dr. Alex Levin has developed considerable expertise in a very difficult and usually bizarre form of maltreatment—Munchausen Syndrome by Proxy. He has done a superb job of characterizing this syndrome by providing the essential components of the syndrome and a profile of the perpetrators. This syndrome represents another form of maltreatment for which a multidisciplinary team is essential.

Munchausen Syndrome by Proxy (MSP) is a form of child abuse in which the perpetrator, who is almost always the

mother, causes her child to come under intense and prolonged medical scrutiny as a result of her falsification of history, covert alteration of laboratory specimens, or covert creation of unusual physical findings which create the appearance of illness in the child.

The most common manifestations of this syndrome include: 1) repeated covert suffocation which creates a clinical picture of recurrent cyanosis, apnea, or gastroesophageal reflux, 2) factitious bleeding from body orifices due either to direct covert injury or to the use of substances such as maternal menstrual or animal blood, which create the appearance of what is thought to be the patient's blood, or 3) laboratory manipulations such as the addition of sugar or salt to body fluid samples. Covert administration of drugs such as insulin, anticonvulsives, and even common household foods such as table salt or pepper have been well recognized as manifestations of MSP.

Although these bizarre clinical scenarios have been noted by many authors, a more common form of MSP may involve the behavior of "doctor shopping," wherein the child is taken from physician to physician with vague and nonspecific complaints, in particular symptoms related to allergic illness or fatigue. Well-meaning medical professionals become entangled in the scenario created by the perpetrator in an attempt to understand what seem to be symptoms of an elusive and rare medical disorder. Therefore, it is especially important for medical caretakers to be aware of this syndrome, as early recognition will prevent continued abuse of the child which can potentially lead to death in approximately 10% of cases.

The profile of an MSP perpetrator is very characteristic, and in and of itself should raise the level of suspicion regarding the possibility of this diagnosis. She most often

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SHAKEN BABY SYNDROME

—by Alex Levin

Editor's Comments

—by Martin Finkel

Parents and non-medical professionals may be unaware of the significant morbidity and mortality which can result from the shaking of a baby. This parental response to the unrelenting cries of a baby frequently presents without obvious injuries and may create a diagnostic dilemma for the clinician. This article by pediatric ophthalmologist, Alex Levin, MD, is an excellent introduction to the problem of Shaken Baby Syndrome.

The Shaken Baby Syndrome (SBS) is a form of child abuse almost exclusively affecting children under the age of three years, with the majority of victims under 18 months of age. The clinical triad of brain injury, skeletal injury, and retinal hemorrhages is the hallmark of SBS. However, only brain injury is required when making this diagnosis. There is currently much controversy regarding the role of blunt head trauma in the causation of the injuries seen in SBS (2,3). However, my own clinical experience, and that of many others, suggests that head trauma is not required to generate the types of brain injuries seen in SBS. These injuries can result from the violent shaking of an infant during which the head is caused to move in many directions in an uncontrolled fashion. Children of this age have relatively large heads, immature brains, and weak neck muscles, all of which contribute to the injuries

As the brain is caused to accelerate and decelerate within the skull, veins may be torn, resulting in the accumulation of blood within (parenchymal) and around (subdural and subarachnoid) the brain. Contusion and laceration of the brain may also occur. Although the brain injuries of SBS may be fatal, a "subclinical" form of shaking may occur in which the infant presents to a caretaker with more nonspecific complaints such as irritability or vomiting. These injuries are most often detectable by CAT scan. However, in some patients, these hemorrhages will only be apparent with MRI scans. MRI also allows for better dating of these injuries.

The skeletal manifestations of SBS include small chip fractures of the ends (epiphyses) of the bones of the arms and legs and/or multiple posterior rib fractures. These latter fractures are caused by compression of the chest between the perpetrator's hands. The extremities are affected when a child is grasped by the arm or leg while the shaking occurs. Despite these multiple fractures, most children who are victims of SBS present with no external evidence of trauma. It should be remembered that rib fractures are an extremely unusual injury in this age range and are rarely produced even with the most severe blunt trauma.

Retinal hemorrhages within the eye occur in over 80% of affected children. They may be few in number or widespread throughout the retina. Dome-shaped blood-filled cystic cavities within the layers of the retina (traumatic retinoschisis) may be diagnostic for SBS. Although the retinal hemorrhages usually resolve without visual sequelae, visual loss or blindness is not uncommon in this syndrome due to injuries to the optic nerve or brain. An ophthalmologist should be consulted to perform complete

retinal examination in all cases of suspected SBS. Postmortem removal of the eyes, and perhaps the entire orbital soft tissue contents, can be extremely important in recognizing that nonaccidental injury has occurred. In particular, our recent research suggests that hemorrhage behind the eyeball may be characteristic of SBS.

It is always important to rule out accidental trauma or medical illness when faced with findings compatible with SBS, in particular the brain and eye injuries. Coagulopathies, spontaneous intracranial hemorrhage (e.g., aneurysm), or meningitis might rarely mimic SBS without fractures. These can usually be ruled out by noting the patterns of physical, radiologic, and ophthalmologic findings. Of course, social history is of utmost importance when trying to rule out accidental injury. The astute clinician must recognize the possibility of abuse based on the total clinical scenario. Whenever retinal hemorrhages are seen, a CAT scan or MRI should be performed. If the CAT scan is normal, I suggest that an MRI be used. A complete skeletal survey, looking for occult fractures, is also indicated.

SBS is a devastating form of child abuse, responsible for the majority of infant homicides. Survivors may be left with severe neurological and visual handicaps. Detection and prompt intervention may be life-saving.

References

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