



XII CONGRESSO NAZIONALE SIMEUP

**DALLE EVIDENZE
ALLA PRATICA CLINICA
IN EMERGENZA-URGENZA
PEDIATRICA**

News dalla letteratura

Emanuela Piccotti

DEA Pediatrico - IRCCS G. Gaslini

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- Il lavoro propone una nuova denominazione di Munchausen syndrome by proxy (Roy Meadow 1977)
- Riporta manifestazioni neurologiche comuni in caso di child abuse suggerendo di tenere in conto questa causa nella diagnosi differenziale infatti studi dimostrano che le manifestazioni neurologiche sono presenti nel 40-50 % dei casi di MCA

Medical Child Abuse Beyond Munchausen

Syndrome by Proxy Thomas A. Roesler, MD and Carole Jenny, MD,

Condizione clinica che si realizza quando un bambino riceve cure mediche non necessarie e/ o dannose / potenzialmente dannose a seguito di istigazione da parte del caretaker .

Si tratta di forma diagnosticabile di child abuse che pone il bambino e i suoi fratelli in situazione di rischio significativo di danno , conseguenze a lungo termine fino alla morte

Common neurological manifestations

- Apparent life-threatening events – ALTE
- Seizures/spells
- Ataxia/gait abnormality
- Weakness/paralysis
- Chronic headaches
- Nystagmus/loss of vision

- **Inappropriate diagnosis of metabolic/mitochondrial disorder**
- **Induction of neurological symptoms by poisoning**

- Apparent life-threatening events

An “apparent life-threatening event” (ALTE) is a relatively common occurrence in infancy that alarms a witnessing caregiver and often leads to the infant presenting to a medical provider. ALTEs are estimated to occur in up to 9.4 per 1,000 live-born infants and **account for nearly 1% of all emergency department visits for infants**

- According to Kahn less than 3% of ALTEs appear to be related to child abuse, but Bonkowsky et al. more recently estimated this prevalence to be closer to 11% . It's important to keep in mind that a presentation of ALTE as a manifestation of MCA can fall into one of two categories : fictitious ALTE versus induced ALTE . There are numerous case reports of confirmed recurrent suffocation as the etiology for symptomatic presentation often initially labeled as ALTE

- Recurrent ALTEs, those that only occur in the presence of one caretaker after which the child is brought to care while still symptomatic, as well as those with oral or nasal bleeding are more likely **problematic**. Covert video surveillance (CVS) can be helpful in proving that reported symptoms are not really occurring and in catching actual acts of ALTE induction. Included in the ALTE differential diagnosis are real or falsified seizures.

Seizures/spells

Although ALTE presentation as apnea, cyanosis, choking, or gagging is a relatively common presentation of MCA, **one of the leading clinical presentations of MCA, at around 42% of cases, continues to be fictitious epilepsy.**

- Seizures can be falsely reported or induced by carotid sinus pressure, asphyxia or poisoning. Atypical seizure histories, ones that are resistant to all treatment, those for which all treatments are not tolerated, and seizures in the context of complex/multisystem diseases, are concerning for MCA.

Ataxia/gait abnormality/ headache

Ataxia or gait problems can be falsely reported or commonly induced by intoxications.

Chronic headache complaints are common MCA manifestations. These are resistant to usual therapies and the children are not observed to have clinical signs. **Nystagmus** is another intoxication-induced symptom.

- The vogue these days in MCA is mitochondrial disease since objective tests such as muscle biopsy and metabolic studies cannot rule it out. Also, its multisystem manifestations are ripe for symptom fabrication.

- Intentional poisoning should be part of the differential diagnosis for any patient with altered consciousness, and other obscure neurological symptoms such as muscle weakness, ataxia, nystagmus, apnea, or syncope. The majority of medications used in intentional poisoning have been prescriptive medications with the most common being anticonvulsants. In one study of 55 cases of this condition, 71% of poisoning agents were prescribed medications and of this group the most commonly used medications were anticonvulsants.

Sequelae

- The potential damage that a victim of MCA may suffer can range from mild to fatal.
- Damage to the CNS is also a risk considering the toxic effects of drugs and poisons administered to induce symptoms. In one case study, an 8-week-old infant with no significant past medical history presented with profuse diarrhea and excessive diuresis due to severe hypernatremia: after close observation, the mother admitted to providing exogenous salt to the infant.

- Long-term morbidity and mortality is harder to estimate. In one review of 451 patients with this condition, 6% of victims died and 7.3% were believed to have suffered long-term or permanent injury. Injury type was both physical and psychological. In another literature review, the mortality rate of this condition was believed to exceed 30%.⁵

Making the diagnosis

- Cases of MCA are so varied and bizarre that the diagnosis is often difficult to make. As with any condition, the first step in making the diagnosis of MCA is to consider the possibility of MCA in the context of a differential diagnosis

- The key to the diagnosis is an index of suspicion. Detailed records review may reveal inconsistencies and falsehoods. Often children who are victims of MCA have had many providers from multiple specialties and multiple health facilities. This makes records collection difficult and onerous.

- The authors recommend multidisciplinary team review and coordination as part of diagnosis and management. Covert video surveillance (CVS) may be appropriate in limited circumstances. Once consensus is reached about the diagnosis, a caretaker informing session, backed up by Child Protective Services (CPS) involvement, is recommended.