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Brief Communication

Nodding syndrome in northern Uganda: Overview and community perspectives

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ABSTRACT

The increasing prevalence of nodding syndrome in northern Uganda has generated a wide range of speculations with respect to etiology and natural history of and best possible medical treatment for this mysterious seizure disorder. Despite in-depth investigations by the United States Centers for Disease Control and Prevention and the Ministry of Health in Uganda, no clear causal factors have emerged. At the same time, northern Uganda communities are voicing concern for their lack of knowledge about nodding syndrome. The purpose of this commentary is to summarize northern Uganda community perceptions of this syndrome. These reflections demonstrate the need for larger investigations into the impact of nodding syndrome and other seizure disorders on local communities both in northern Uganda and throughout the world, in particular rural areas of resource poor countries.

1. Introduction and overview

Nodding syndrome is a new epilepsy disorder that mainly affects children and only has been reported and confirmed from three African regions at this point in time: northern Uganda, South Sudan, and southern Tanzania [1–4]. It is characterized by paroxysmal and repetitive head nodding, most likely resulting from loss of neck muscle tone [5]. Nodding syndrome initially was described in southern Tanzania in the early 1960s and later studied extensively in that region [5–9]. It is associated with clinically supportive signs of epilepsy and seizure activity on electroencephalography [1,5]. Winkler et al. established in Tanzania the definitions of "head nodding" and "head nodding plus" that categorize patients as having solely nodding features or nodding with other types of epileptic seizures; this terminology continues to be utilized among healthcare workers in the northern Uganda region. The most effective treatment to reduce the occurrence of nodding attacks and other associated seizures remains anticonvulsants such as sodium valproate, phenobarbital, and phenytoin [5,10].

Since 2009, nodding syndrome has become increasingly prevalent in South Sudan and northern Uganda, although single reports as well as unpublished data point to a much earlier occurrence in the mid 1990s [2,11,12] and has since generated significant scientific, political, and media attention. In 2012, the United States Centers for Disease Control and Prevention (CDC) announced its plans for a clinical trial on potential treatments [4]. As demonstrated in historical and current studies, nodding syndrome in northern Uganda continues to affect children, primarily in the age range of 5–15. The most recent statistics from the Ugandan Ministry of Health reported approximately 3000 affected children and 170 deaths [13].

In these children, nodding syndrome appears to represent a progressive encephalopathy leading to cognitive decline. Death may result from mainly secondary causes, such as injuries sustained during seizures or malnutrition; but thus far, mortality and its causes have not been reported from prospective epidemiological studies [4,12]. In affected children in Tanzania, mental retardation was present in approximately 40% of the children ranging from mild to severe [9]. Contrary to what has been observed in northern Uganda, in the majority of cases in Tanzania, the syndrome is not rapidly progressive and does not result in severe encephalopathy. This difference makes us wonder whether nodding syndrome in Uganda and Tanzania represents the same entity or whether the witnessed head nodding may be the result of different underlying pathological processes.

The CDC 2011 investigation into the South Sudan outbreak corroborates earlier theories that infestation with *Onchocerca volvulus* is higher in affected children than in unaffected children [1]. Nevertheless, the evidence still does not speak definitively in favor of this as an etiology, as no studies have yet determined whether exposure occurred before or after the onset of nodding syndrome in the affected individuals. In addition, the only large-scale investigation of cerebrospinal fluid in people with head nodding and other epilepsy syndromes in southern Tanzania, an area endemic for *O. volvulus*, did not show any evidence of present or past infection with the parasite, this study concluded that direct invasion of the central nervous system with *O. volvulus* was highly unlikely

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[14]. However, indirect mechanisms of a causative link cannot be fully excluded at this time.

Preliminary results of the CDC investigation in South Sudan show no association with patient demographics or exposure to munitions. The CDC currently is studying other potential etiologies related to food supply, vitamin deficiency, and heavy metals [1]. Local World Health Organization authorities currently are pointing to nutritional deficiencies [2]. Nevertheless, to date, no published studies have demonstrated a clear statistically significant relationship of nodding syndrome to any of these variables.

Finally, the geographical and familial clustering of nodding syndrome has led multiple different investigators to suggest a genetic etiology for the syndrome. In their 2008 Tanzania study, Winkler et al. discuss the possibility that nodding syndrome may represent a distinct genetic epilepsy syndrome. Head nodding may also be part of epilepsy and non-epilepsy disorders with unknown etiology, such as paroxysmal tonic upgaze with coexistent absence epilepsy [15] or spasmus nutans [16,17]. They also note the presence of gliotic changes and hippocampus sclerosis on cerebral MRI in several affected children with nodding syndrome. Though patient numbers were small and, therefore, results may not be representative, they are identified as a feature for future investigation [5].

Overall, while much-needed new attention on nodding syndrome has developed in recent years, the syndrome, nevertheless, remains puzzling for investigators. While no clear etiology for nodding syndrome has been identified, future studies may offer new insights. At the same time, the Lutheran World Federation (LWF) Uganda Country Program, a registered non-governmental organization serving northern Uganda for nearly a decade, has been addressing the psychosocial impact of nodding syndrome on these communities. Its observations offer important insight on community beliefs about disease etiology, transmission, and treatment. The purpose of this report is to summarize the northern Ugandan community's perceptions of this new syndrome, as may be applicable to other seizure disorders throughout the world.

2. Methods/results

The Lutheran World Federation works under the auspices of and in cooperation with the government of Uganda, and has well-established relationships with local leaders and community members. Additionally, as Humanitarian Accountability Partnership (HAP) certified, LWF operates in a way that ensures the accountability and quality of its work to the community members.

Before conducting group dialogs with adult caregivers of children with nodding syndrome at Atanga Health Centre III in Atanga subcounty, Pader District in the spring of 2012, LWF field workers explained in local language the purpose and content of the discussions. A written consent was read to the participants as a group, and each adult participant then provided individual approval and understanding with a fingerprint. In this way, each participant acknowledged his willingness to participate in the discussion and approval to publish the contents of the discussion. Participants understood that they could leave the group discussion at any point and were not required to answer the questions asked. Consent forms subsequently were filed in the LWF head office in Kampala, Uganda.

Approximately 10–12 adult caregivers were surveyed together in each focus group dialog; there were five total dialogs conducted. The groups were not divided according to age or gender, and more than half of the participants were female. Participants were allowed to express individual opinions to LWF fieldworkers after the conclusion of the group dialogs. Lutheran World Federation field workers transcribed these quotes and reviewed field notes from their community work over the past 3 years. This ensured that the opinions of participants in the group dialog represented those of the community at large. Given the descriptive and qualitative nature of this study, no statistical software was used for analysis. The Gulu University Institutional Review Committee approved the work.

2.1. Perceptions about etiology

Despite the medical theories regarding potential causal agents in the etiology of nodding syndrome, the communities of Kitgum, Lamwo, and Pader strongly associate the temporal relationship of the war with the onset of nodding syndrome in their children. The LWF field officer assigned to address the impact of nodding syndrome in northern Uganda explains that, “Conclusively, people say that this is a result of the war. Before the camps, there was no nodding syndrome. There were a few cases of epilepsy treated successfully with local herbs. The community believes that nodding syndrome resulted from a condition of the [internally displaced person] IDP camps.” He also adds that community members definitely state, “The black fly brings blindness, not nodding syndrome.”

Many of the community members believe that food provided to them in the camps triggered nodding syndrome. They endorse theories involving maize flour, cowpeas, porridge, and cooking oil. Most do not think that water caused nodding syndrome, affirming that the borehole supply was safe in the camps.

Despite being concerned about potential causal relationship between wartime food and nodding syndrome, the community members do not believe that the Lord's Resistance Army (LRA) itself poisoned any food supply. “The LRA just killed people,” one community member said. Nevertheless, they do believe that the weapons used by the LRA may have contributed to the development of nodding syndrome. “The force that the gun releases can cause an abortion, and bombs and explosions released chemicals,” one parent explained.

Aside from the food and potential chemical poisoning, the northern Uganda community demonstrates religious and spiritual beliefs about the etiology of nodding syndrome. One parent of an affected child explained, “It's a plan from God, because not all families have it. God is tired of people, and is punishing in many ways. This is for the many sins they have committed … we don't know what the sins are, but God does.” The spiritual beliefs are intertwined with the effect of war on their region. Many community members assert that fighting in the war led to the development of nodding syndrome. “Many people were killed,” one parent said. Another added, “If you have a quarrel at home, then you have problems … if you quarrel and then something bad happens, it is the result of the quarrel.”

2.2. Perceptions about transmission

Despite associating the appearance of nodding syndrome with the decade of war, most community members believe that the syndrome is transmissible from one child to another. Many parents attempt to isolate ill from healthy children. The village schools indicate similar measures to keep affected children physically separate from others. One parent expressed that adults are not susceptible to the transmission of the syndrome because “The blood of elders is stronger.” Another added, “Adults can resist because they are strong. Adults who are experiencing nodding syndrome may still have the spirits or toxins in them.”

2.3. Perceptions about treatment

Though community members believe that nodding syndrome started as a result of some condition in the IDP camps or war itself, they, nevertheless, followed the advice of the camps to obtain medical attention at local health centers. One parent explained, “Maybe the medical centers have the medicine for a cure. Nodding medicine is the only cure, the only option. There are no other options besides modern medicine.” Others believe that the only positive change in their children has resulted from antiepileptic drugs provided at medical centers. Finally, one parent affirmed, “the medicine doesn't chase the spirits away, but stops the fits.”

3. Conclusions

While the CDC and other medical personnel investigate the causes and the most effective treatment of nodding syndrome, the LWF experience suggests that addressing cultural conceptions about the disease and providing non-medical support to the community are equally important. These voices illuminate how a history of regional conflict affects family understanding of illness, highlight the relationship between disease and culture, and suggest how community health initiatives could be focused to best educate the community. Larger studies investigating the cultural understanding of nodding syndrome and other seizure disorders, their impact on the communities, and realistic interventions to mitigate their effects are warranted.

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