Investigation into the Nodding syndrome in Witto Payam, Western Equatoria State, 2010

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Introduction

'Nodding syndrome' is found in South Sudan mainly in Western Equatoria where it was first reported in the 1980s (1) and where WHO did a series of assessments in 2001, 2004 and 2006 (2). It has also been described in Western Uganda (3) and Tanzania (4).

Nodding syndrome is a progressive condition characterised by head nodding (hence the name), mental retardation and stunted growth (4) affecting mainly children and young adults.

Some reports suggest that the episodes of nodding occur when the child begins to eat food or feels cold (1, 4). These episodes are said to be brief and disappear when the child stops eating or feels warm again.

Little is known about the prognosis of the nodding syndromebut it is thought to be a very debilitating physically and mentally. Attacks can cause children to collapse and injure themselves or die, for example, by falling into a fire.

Materials and Methods

In September 2010 a team led by the Ministry of Health, Government of Southern Sudan (MoH-GoSS) visited Witto Payam in Western Equatoria State in order to investigate reports from UNOCHA and some INGOs of nodding syndrome in Witto Internal Displaced People (IDP) camp. The objective of the team (composed of the authors of this report) was to provide the MoH-GoSS with workable recommendations on how to tackle the syndrome. The people now living in the IDP camp had been displaced from Diko, Tore and some surrounding villages by the Lord's Resistance Army. Most of the IDPs are originally from Witto Payam who moved and settled in Diko during the war time displacements. The team met community leaders, administrators and families at Witto IDP camp, Lui town and Jambo town.

At Witto IDP camp the community leader explained the purpose of our visit (the first government-led delegation to investigate the mysterious nodding phenomenon) to a large crowd of affected children and their parents. We then had a one-hour brainstorming session with a focus group of 25 parents, elders of the community and church leaders. We used unstructured questions and answers to find out:

- when the community noticed the first case of nodding syndrome
- events that happened, including eating habits, during the civil war and
- what the community thinks causes the disease.

We asked individuals to give a history of the syndrome and to suggest possible causes and ways the condition may be transmitted. We also interviewed five parents with affected children and we tried to elicit the nodding symptom in four children by asking their parents to feed them local foods.

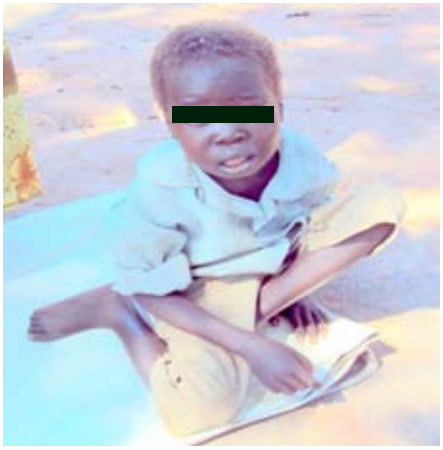

Figure 1. A 15-year old boy with stunting and mental retardation in Witto Payam (credit: Thomas Akim)

In Jambo town we repeated a similar brain storming session with a fifteen-member group and we interviewed

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the Clinical Officer at Jambo Health Centre.

At Lui Hospital we met the administrator and interviewed two doctors, the head nurse, the statistician and five cases at the outpatient department (OPD) and Medical Ward. Some of us briefly visited a nearby co-education primary school and collected a list of pupils reported to have the nodding syndrome.

Figures 1 and 2 show some affected children.

Figure 2. Mental retardation in the young adult in Witto Payam (credit: Thomas Akim)

Findings

We registered a total of 96 cases of nodding syndrome: 70 in Witto IDP camp and 26 at Jambo. Of the affected children:

- 46% were categorised as stunted according to our observation. However we do not know the prevalence of stunting in children without the syndrome.
- 52% of their parents said that "nodding is induced by the sight of food".

Figure 3 shows that:

- Most cases were aged between 5 and 20 years and therefore born between 1980 and 2005.
- The greatest number was in the 10 -15 year age group.

Figure 4 shows the increasing trend of nodding syndrome cases in Witto Payam and Jambo town over time.

Of the 96 documented cases:

- 96% had been treated at Lui Hospital by a Medical Officer or at Jambo PHCC by a Clinical Officer. The drugs mainly used were anticonvulsants: carbamazepine and phenobarbitone - that had been issued monthly and provided by MoH-GoSS.
- 74% were the first, second, third or fourth borne of the mother.
- In some families more than three children were reported to be affected.

Four of the affected children were given food under our observation but we did not see any 'nodding phenomenon'.

Information from the community

There appeared to be two types of presentations of the syndrome:

1. The community reported that, "the nodding symptom usually begins with the sight of food" and over time changes to generalised tonic–clonic convulsions (i.e. partial seizures with secondary generalization).

2. Some patients were said to initially present with generalised seizures characterised by sudden loss of consciousness, tonic-clonic convulsions, rolling back of eyes, salivation, loss of sphincter control, confusion and finally a deep sleep (generalized seizures). The parents reported that the condition is preceded by an aura that could suggest a focal origin of an epileptic discharge.

The community could not provide a credible theory for the emergence of the syndrome. Some elders speculated that it could have resulted from relief food that might have been contaminated, childhood vaccinations or large or small flies which had invaded the areas of wartime displacement. They had observed that people became blind when bitten by these flies.

Some community members could not suggest any plausible theory while some thought it could have supernatural causes. Others thought the disease was caused by the settling amongst, and subsequent intermarriage between, the local community and other war-displaced Southern Sudan communities. Notably, traditional healers who had been consulted by the community had not succeeded in treating the syndrome.

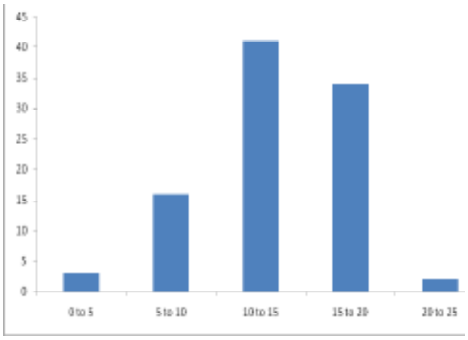

Figure 3. Distribution of numbers of affected persons in Witto Payam by age group (years)

Management

At present the syndrome is treated with common anti-convulsants although prognosis is said to be poor. According to health workers, children who start treatment early and on a regular basis have seizures less frequently. Therefore they have normal growth and no observable complications whereas their cohorts on irregular or no treatment have more complications.

In fact the drugs are in short supply. Some parents of the affected children bought medicines from a private pharmacy but said they were ineffective. This could have resulted from drug intolerance or under-dosing. Many health workers are not familiar with the syndrome and do not know how to treat and care for patients - indicating the need for more training.

As a remedy, the community in Witto Payam had tried to socially isolate their affected children (i.e. drinking, eating and sleeping separately) but said the syndrome continued to occur in healthy children. The school administration was in a dilemma as to whether or not they were required to have separate classes for affected children. If one pupil in the class showed symptoms, by the end of the year about seven other children were reported to have the same condition. This raises the question of whether this is a communicable disease.

Discussion

The team considered the following questions.

1. Is nodding syndrome a form of epilepsy?

Observations and reports of the nodding episodes both from this study and from others in Tanzania and Uganda lead us to speculate that the nodding syndrome may be a particular form of epilepsy found mainly, perhaps only, in this area of Africa. The study in Tanzania, which did MRI scans and EEG recordings (4), concluded that head nodding is "possibly a new epilepsy disorder in sub-Saharan Africa". A previous study in Lui indicated that EEG results were consistent with a specific encephalopathy, which progresses in well-defined stages, and nodding represents the onset of symptoms and the ictal events common to all stages of epilepsy (5). However until this condition can be further investigated by a clinical neurologist and by doing more EEGs it is difficult to come to definite conclusions.

Children with 'nodding' are said to have mental retardation, and nodding is often seen as a habit or 'comfort' activity in children with learning disability, autism, etc. The link between nodding and mental retardation needs to be clarified.

That the nodding syndrome may be a type of epilepsy is strengthened by the fact that the health workers reported that children who start treatment with anti-convulsants early and on regular basis have reduced frequency of the seizures compared to affected children on irregular or no treatment.

Some parents reported that nodding was triggered by the sight of food and this has been reported previously (1, 4). However it did not occur when we watched affected children being offered food. So this observation needs more investigation.

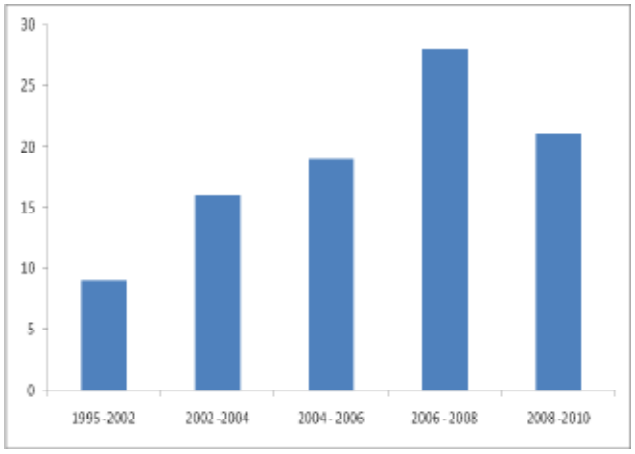

Figure 4. Trend in prevalence of nodding syndrome between 1995 - 2010

2. What are the causes of nodding syndrome?

The team considered whether the condition could be caused by:

1. Ingesting chemicals from biological and chemical weapons previously used in the area. Had these affected the parents so that the syndrome was genetically passed to their children? Although some families had more than one child with the syndrome this seems unlikely as the condition has been described in Tanzania and Uganda where there have been no wars.

2. Eating seeds covered in toxic chemicals that were provided by relief agencies and meant for planting – as suggested by the community. However toxicological investigations on relief foods, such as lentils and sorghum, gave insignificant results in a case control study done in Lui (5).

3. Infection by a parasitic worm, Onchocerca volvulus, which is carried by a black fly and which causes river blindness. Most children in South Sudan suffering from nodding syndrome live close to the Yei River and

93% of the victims carry the parasite (6). A link between river blindness and other cases of epilepsy (7) as well as retarded growth (8) had been proposed previously, although the evidence for this link is inconclusive (9). In the Tanzanian study MRI lesions were associated with positive skin polymerase chain reaction (PCR) for O. volvulus despite negative PCR of the cerebrospinal fluid (4).

According to the majority of the people we interviewed, the nodding syndrome was first noticed around 1997 in IDP camps near Tore, a village near Yei, and on the banks of Yei River. However, some community members claimed that, even before displacement took place in the bushes of Kediba County, cases were already present in the Jambo area (or in Witto Payam) around 1992.

The team considered that the involvement of a vector was not supported as the syndrome affected only children and young adults born around 1989 to 2005. Also it seems to occur in specific areas despite there being the same ecosystem in most of Western Equatoria State and part of Central Equatoria State (Yei County).

Before the role of Onchocerca volvulus can be ruled out further investigations are needed to find out whether children with the nodding syndrome have a greater prevalence of Onchocerca volvulus than non-affected children.

Conclusions

Based on the team's observations at Witto Payam and a review of the literature it is tentatively speculated that nodding syndrome may be a particular type of epilepsy, found mainly in certain areas of Eastern Africa, which begins with focal symptoms and later progresses to generalised convulsions. However its causes remain obscure.

The present study relied mainly on data from interviews with local people, and the team recommends that further studies be carried out to investigate this syndrome (including its causes, clinical presentation and prevalence) in more detail. These should include laboratory investigations, brain scans and EEGs, use objective research methods (e.g. case controls) and involve experienced clinicians.

Note: After returning from Witto Payam we found that more than 500 cases of nodding syndrome were being treated in Usratuna PHCC in Juba. The PHCC record review indicated that the cases were coming from the wider community of Southern Sudan.

Recommendations

The Ministry of Health should:

- provide adequate medicine for the symptomatic treatment of affected persons
- urgently conduct research into the possible causes of the nodding syndrome to allow better management of the patients with the illness and to help undertake preventive measures.

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