Original article

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The natural history of nodding syndrome

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Introduction

- Nodding syndrome is a poorly understood acquired neurological disorder affecting children in sub-Saharan Africa
- The aetiology and pathogenesis are unknown. Preliminary studies suggest the syndrome may be a neuro-inflammatory disorder with antibodies to *Onchocerca volvulus* cross reacting with neuron proteins.
- The distinctive feature is repeated clusters of head nodding (that occur at the sight of food, with cold weather or baths, or spontaneously). Patients progressively develop multiple other features (seizures, cognitive and behavioural problems, growth failure, and severe disability).

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Objective

- This study aimed to describe the early features and natural history of nodding syndrome, and to refine the proposed clinical stages.

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Methods

This study was part of a larger epidemiologic study of nodding syndrome in Uganda and was a cross-sectional survey of a retrospective cohort of patients living in Pader district in Northern Uganda.

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Results

- A total of 210 children were recruited.
- The mean age at the onset of head nodding was 7.5 (SD 3.0) years.
- Five overlapping clinical stages were recognised; Prodromal, head nodding, convulsive seizures, multiple impairments, and severe disability stages.
    - Clinical features before the onset of head nodding (prodromal features) included periods of staring blankly or being inattentive, complaints of dizziness, excessive sleepiness, lethargy, and general body weakness, all occurring 2 weeks -24 months before nodding developed.
    - After the onset of head nodding, patients progressively developed convulsive seizures, cognitive and psychiatric dysfunction and physical deformities, growth arrest, and eventually, in some patients, severe disability.

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Conclusion

- Nodding syndrome is probably a multisystem disorder in which symptoms develop over several overlapping and progressively severe stages, starting with a non-specific prodromal period of variable length.
- A high index of suspicion and prompt recognition of especially the early features may guide in the early identification of at-risk patients and promote the prompt initiation of interventions before extensive brain injury develops.
- The wide spectrum of symptoms and complications emphasises the need for multi-disciplinary investigation and care.

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