Official Journal of the European Paediatric Neurology Society

Original article

Paroxysmal tonic upgaze in normal children: A case series and a review of the literature

Cinzia Salmina [a], Ilaria Taddeo [a], Mattia Falesi [a], Peter Weber [b], Mario G. Bianchetti [a,*], Gian Paolo Ramelli [a]

[a] Department of Pediatrics, Mendrisio and Bellinzona Hospitals, and University of Bern, Switzerland
[b] Division of Neuropediatrics and Developmental Medicine, University Children's Hospital, Basel, Switzerland

ARTICLE INFO

Article history:
Received 16 October 2011
Received in revised form
2 April 2012
Accepted 21 April 2012

Keywords:
Abnormal eye movements
Epileptic seizures
Non-epileptic seizures
Paroxysmal tonic upgaze of childhood

ABSTRACT

The purpose of this review was to update the clinical characteristics of paroxysmal tonic upgaze in neurodevelopmentally normal children. We made the diagnosis (between 2008 and 2010) in 8 infants referred to us with suspected epilepsy. We found 38 further cases in the literature.

In the 46 children (29 boys and 17 girls) tonic upward ocular deviation was first noticed between the age of 2 weeks and 90 months (median: 9 months). This tendency persisted for between 1 and 48 months (median: 7 months). The duration of paroxysmal events was highly variable: brief events lasted between 3 s and 10 min in 50% of the cases, intermediate events between 5 s and 30 min and long events between 10 s and 2 h. The frequency ranged from one every 3 months to 10 per day. In 15 children the episodes of upward deviation of the eyes were associated with an impaired movement coordination. In 2 further children an impaired movement coordination was noted during febrile illnesses.

The results of this review in normal children characterize the entity as follows: onset under 2 years of age, a small predilection for the male gender, eventual improvement and recovery, and impaired movement coordination.

1. Introduction

First reported by Ouvrier and Billson in 1988, paroxysmal tonic upgaze of childhood is considered a distinctive non-epileptic disorder of abnormal ocular movements with onset during infancy that can occur in otherwise neurologically normal subjects.[1] The condition is characterized by episodes of conjugated upward eye deviation with neck flexion, down beating saccades in attempted downgaze, preserved horizontal eye movements, and normal consciousness, which are often associated with an impaired ability to coordinate movements.[1,2]

We recently made the diagnosis of paroxysmal tonic upgaze in 8 neurologically normal children. However, we were impressed by the paucity of information in available reviews that deal with paroxysmal nonepileptic events in childhood.[3] To integrate efficiently the existing information on benign paroxysmal tonic upgaze, we present our experience and the results of a systematic review of the corresponding original literature.

* *Corresponding author*. San Giovanni Hospital, 6500 Bellinzona, Switzerland.
E-mail address: mario.bianchetti@pediatrician.ch (M.G. Bianchetti).
1090-3798/$ – see front matter © 2012 European Paediatric Neurology Society. Published by Elsevier Ltd. All rights reserved.
doi:10.1016/j.ejpn.2012.04.004

2. Case series

Over a period of 3 years, between 2008 and 2010, 8 apparently healthy Swiss children (4 boys and 4 girls) with paroxysmal tonic upgaze were ascertained by two of us (GPR and PW). The children had been born at term ($N = 7$) or near term ($N = 1$) without perinatal or neonatal disease indicators after an uneventful pregnancy with a body weight appropriate for gestational age. They had been referred for diagnostic evaluation of suspected epileptic seizures.

The diagnosis of paroxysmal tonic upgaze of childhood was made when a child had recurrent episodes of sustained conjugate upward deviation of the eyes lasting 3–6 s with normal horizontal eye movements, which had been documented by video-recording, and absent epileptiform activity on electroencephalography. Since no child with a clinical picture of paroxysmal tonic upgaze had concurrent electroencephalographic abnormalities, none was excluded from the report on this basis.

In all cases, one of us (IT) recently (July of 2011) performed an interview with the parents of the children, whose age ranged between 20 and 36 months. In addition to age at onset, age at maximal expression and age at resolution, particular inquiry was made into possible onset trigger factors, frequency, exacerbating factors, diurnal fluctuation, and associated features. Furthermore, follow-up assessment involved inquiry into other paroxysmal neurological phenomena. Specific inquiry was also made into the family history of eye movement abnormalities and of paroxysmal neurological disorders. Finally, a standardized neurological examination and the Griffiths test were performed.

First-degree relatives of 2 children were affected with seizure disorders, while 1 child had a history of simple febrile convulsions. In the 8 children, episodes of conjugate upward deviation of the eyes, with neck flexion apparently counteracting for the abnormal eye position, were first noted between 2 and 18 months of age (Table 1). In all cases the episodes were relieved by sleep and only reappeared one to 3 h after awaking. In two children first episodes of tonic upgaze appeared within 2–3 days after an immunization respectively an acute upper respiratory tract illness. The frequency of observed episodes of upward deviation of the eyes ranged from 2 to 3/day to ≥10/day. Stereotypic movements of the hands were associated with paroxysmal tonic upgaze in 2 children. No brain imaging studies were performed in the eight children. Their parents were given reassurance regarding the functional and self-remitting nature of the condition and no drug treatment was prescribed.

In the 8 children, the tendency towards paroxysmal tonic upgaze disappeared 1–7 months after onset. At last follow up all children were normal.

3. Review of the literature

Two of us (CS and MF) recently performed a computer-based search of the terms “tonic upgaze” and “tonic upward gaze”. Only cases occurring in neurodevelopmentally normal children without seizure disorders that were published after the

Table 1 – Clinical features in 8 Swiss patients (4 girls and 4 boys) affected with paroxysmal tonic upgaze of childhood. In all cases the episodes were relieved by sleep.

Number	Gender	Age at Onset (Months)	Onset Trigger Factor	Age at Maximal Expression (Months)	Age at Resolution (Months)	Frequency	Exacerbating Factor and Diurnal Fluctuation	Associated Features	Age at last follow-up (months)	Neurodevelopmental Outcome[a]
1	Male	2	None	2–8	8	2–3/day	None	None	29	Normal
2	Male	5	None	6	8	4–6/day	Supine Position	None	22	Normal
3	Female	6	None	6	7	2–3/day	Attacks after crying	None	37	Normal
4	Male	7	6th month vaccination	9	14	≤10/day	While sitting on high chair	Uncoordinated movements of the hands	28	Normal
5	Female	7	None	9	11	4–6/day	Baby’s nutrition and changing diapers	None	36	Normal
6	Male	12	None	12–13	16	4–6/day	While playing	None	20	Normal
7	Female	15	None	15–19	19	2–3/day	Fatigue (especially evening hours)	Uncoordinated movements of the hands	22	Normal
8	Female	18	Upper respiratory illness	18–22	22	4–6/day	While playing	None	39	Normal

a Griffiths mental development test.