Movement Disorders
CLINICAL PRACTICE
CLINICAL VIGNETTE

Dystonia in Pediatric Huntington's Disease; Prominent and Possibly Painful

Marjolein M.A. Ketels, MD,[1,] Oliver William Quarrell, MD, FRCP,[2] and Mayke Oosterloo, MD, PHD[1]*

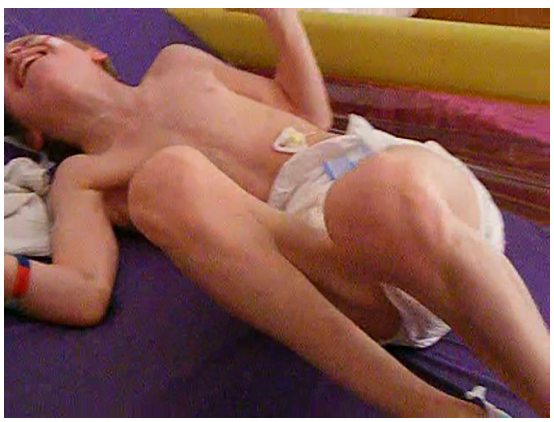
Pediatric Huntington's disease (PHD) is defined as affected with Huntington's disease under the age of 18.[1] Common clinical features at the presentation of PHD are cognitive impairment, behavioral changes, gait disorder, cerebellar signs and oral motor dysfunction.[1] Dystonia and pain have previously been reported as possibly common, but unrecognized features in PHD, which are not necessarily intertwined.[2] Our two videos suggest that painful generalized dystonia can be a prominent feature in PHD (Videos 1 and 2). Dystonia can be paroxysmal (Video 2) and non-paroxysmal (Video 1). When correctly recognized, symptomatic treatment can improve quality of life of these patients. Unfortunately, in case 1 (Video 1) pain medication including opiates were not sufficient and ultimately, pain led to palliative sedation. In case 2 (Video 2), opiates were prescribed to improve the pain, with limited effect (Videos 1,2).

Author Roles

(1) Research project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript: A. Writing of the first draft, B. Review and Critique.

M.K.: 3A.
O.Q.: 3B.
M.O.: 3B.

Video 1. 15-year old girl with PHD and prominent dystonia of both arms and legs. She is screaming and seems to be in pain. She verbally confirms that she is in pain, however this is not on tape.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/mdc3.13850



Video 2. 11-year old boy with PHD and dystonic posturing of the limbs. He is screaming and seems to be in pain.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/mdc3.13850

[1]*Department of Neurology, Maastricht University Medical Center, Maastricht, The Netherlands;* [2]*Department of Clinical Genetics, Sheffield Children's NHS Foundation Trust, Sheffield, UK*

***Correspondence to:** Marjolein M.A. Ketels, Department of Neurology, Maastricht University Medical Center, Maastricht, The Netherlands; E-mail: marjoleinketels@gmail.com
Keywords: pediatric Huntington's disease, dystonia, pain.

Received 24 February 2023; revised 22 June 2023; accepted 16 July 2023.
Published online 18 August 2023 in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/mdc3.13850

Disclosures

Ethical Compliance Statement: The authors confirm that the approval of an institutional review board was not required for this work. Declaration of patient consent: written consent was received from the next of kin of the presented patients. The parents were verbally informed about the manuscript and also had the opportunity to read it before submission. Their signed consent was saved as a pdf file. Journal's Ethical Publication Guidelines: We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Funding Sources and Conflicts of Interest: No specific funding was received for this work. The authors declare that there are no conflicts of interest relevant to this work.

Financial Disclosures for the Previous 12 Months: The authors declare that there are no additional disclosures to report. ■

References

1. Cronin T, Rosser A, Massey T. Clinical presentation and features of juvenile-onset Huntington's disease: a systematic review. *J Huntingtons Dis* 2019;8(2):171–179.
2. Moser AD, Epping E, Espe-Pfeifer P, et al. A survey-based study identifies common but unrecognized symptoms in a large series of juvenile Huntington's disease. *Neurodegener Dis Manag* 2017;7(5): 307–315.