Functional tics and echophenomena

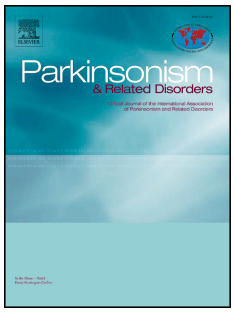
Ganos, Christos; Erro, Roberto; Cavanna, Andrea E.; Bhatia, Kailash P.

DOI:
10.1016/j.parkreldis.2014.10.001

Document Version
Peer reviewed version

Citation for published version (Harvard):
Ganos, C, Erro, R, Cavanna, AE & Bhatia, KP 2014, 'Functional tics and echophenomena', *Parkinsonism and Related Disorders*, vol. 20, no. 12, pp. 1440-1441. https://doi.org/10.1016/j.parkreldis.2014.10.001

Accepted Manuscript



Functional tics and echophenomena

Christos Ganos, Dr. Roberto Erro, Andrea E. Cavanna, Kailash P. Bhatia

PII: S1353-8020(14)00370-8

DOI: 10.1016/j.parkreldis.2014.10.001

Reference: PRD 2454

To appear in: *Parkinsonism and Related Disorders*

Received Date: 1 June 2014

Revised Date: 18 July 2014

Accepted Date: 2 October 2014

Please cite this article as: Ganos C, Erro R, Cavanna AE, Bhatia KP, Functional tics and echophenomena, *Parkinsonism and Related Disorders* (2014), doi: 10.1016/j.parkreldis.2014.10.001.

Functional tics and echophenomena

Christos Ganos[1,2], Roberto Erro[1], Andrea E. Cavanna [1,3,4], Kailash P. Bhatia[1]

[1] Sobell Department of Motor Neuroscience and Movement Disorders, UCL Institute of Neurology, University College London, London, United Kingdom
[2] Department of Neurology, University Medical Center Hamburg-Eppendorf (UKE), Hamburg, Germany
[3] Department of Neuropsychiatry, BSMHFT, and University of Birmingham, Birmingham, United Kingdom
[4] School of Life and Health Sciences, Aston University, Birmingham, United Kingdom

Correspondence to:
Dr. Christos Ganos
Clinical Movement Disorders Group
Sobell Department of Motor Neuroscience and Movement Disorders
UCL Institute of Neurology
Email: cganos@gmail.com

Keywords: Echophenomena; Functional (psychogenic) tic disorders; Gilles de la Tourette syndrome;

Title character count: 51

Manuscript word count: 794

No conflicts of interest

Main Text

Echophenomena denote the stimulus-triggered repetition of actions (echopraxia) or sounds (echolalia). Gilles de la Tourette syndrome is the most commonly known neuropsychiatric syndrome associated with the presence of echophenomena. However, several other conditions, including the bizarre and less well-explored culture-specific startle syndromes (i.e., Latah, Jumping Frenchmen and Miryachit) may also present echophenomena [1, 2]. To date, there has been no report of echophenomena in functional movement disorders. Here we describe the case of a young female patient with a functional tic disorder and abnormal stimulus-triggered behaviours with striking echophenomena.

This 18-year-old female developed jerky movements at the age of 16. The first episode was noted during a driving lesson, when she felt pain in her right arm, followed by uncontrollable jerky movements, which lasted for several minutes. A few weeks later, at nighttime, she again complained of pain in her right arm, and during that time the movements reappeared for several hours. Since then, she has been experiencing jerky movements in all parts of her body. She also developed an uncontrollable impulse to repeat other people's words or actions several times, for up to hours. This has been in fact her biggest problem as it has led to difficulties with her social life and academic performance. Further, she reported irregular episodes of right arm shaking during writing or carrying objects. There was no history of substance abuse and no history of attention-deficit hyperactivity or obsessive-compulsive disorder. The patient's younger sister was born with Down's syndrome. The rest of the family history with regard to neuropsychiatric conditions was unremarkable.

On clinical examination (segments shown in video), complex, brief and sudden jerky, but in their majority non-repetitive, tic-resembling movements were noted. Most of the movements interfered with the execution of voluntary actions leading to either exaggerated responses or action interruptions. Some, but not all movements were preceded by inner tension and

although voluntary control reduced their severity and frequency, they could not be inhibited completely. In contrast, cognitive or emotional engagement led to their brief cessation. Startle responses were inconsistently exaggerated. Words, such as “no” or “caffeine”, but also other words not embedded in a certain context triggered prolonged periods of echolalia. Similarly, sudden unforeseen actions of a third party would elicit echopraxia. Previous investigations for secondary causes of tic disorders (peripheral blood smear for acanthocytes; cranial MRI) were unrevealing.

Historically, echophenomena were emphasized by Georges Gilles de la Tourette, who considered them essential diagnostic features for his newly described disorder [3]. Subsequently, it became clear that echophenomena in early life constitute normal elements of healthy development, but their persistence or re-emergence at a later developmental stage may denote an underlying neuropsychiatric condition, including autism spectrum disorder, catatonia, speech disorders, as well as different types of dementia and (culture-bound) startle-syndromes [1, 2]. Interestingly, the latter group of syndromes, which is characterized by exaggerated spontaneous and startle-induced echophenomena, as well as irregular jerky movements resembling tics, shares some phenomenological features with the characteristics of the motor behaviour in our presented case here. However, the main complaint of our patient was not that of abnormal startle behaviour, as the movements she echoed were largely dependent on contextual embedment of stimuli and not their intensity. Also, other clinical features encountered in the culture-bound startle-syndromes, such as automatic obedience or further startle-induced abnormal motor behaviours (e.g. throwing objects) were not present here.

The manifestation of sudden, brief and uncontrollable jerky movements alongside the prominent presence of echophenomena, in the presence of normal early development and in the absence of cognitive decline or further neuropsychiatric features, in an adolescent, should

prompt the consideration of a primary tic disorder. However, several clinical characteristics were not in favour of this. First, the onset of the movement disorder here was in late adolescence and was sudden, rather than insidious and during childhood, which is typical for primary tic disorders. Also, the presented movements were in their vast majority not repetitive and complex and in contrast to primary tics did not show the characteristic rostro-caudal distribution gradient. In addition, although most tics usually lessen during action execution, here most movements significantly interfered with voluntary actions. Further, although the jerky movements could not be completely voluntary inhibited, they were clearly distractible.

Taken together, the aforementioned features of the jerky movements were atypical for a primary tic disorder and alongside the reported episodic right arm tremor prompted the consideration of a functional (psychogenic) movement disorder. In fact, similar clinical features have been highlighted in a case-series of nine patients diagnosed with functional movement disorders resembling tics corroborating given diagnosis[4]. Of note, functional tic disorders are rare [4]. However, this case demonstrates that echophenomena may also be encountered in this context and widens the differential diagnosis of clinical presentations involving echophenomena across the neuropsychiatric spectrum of movement disorders.

Author Contributions

A: Drafting/revising the manuscript for content, including medical writing for content. B: Acquisition of data. C: Study supervision or coordination.

CG; RE; AEC; KPB: ABC

Financial Disclosures:

Christos Ganos
Receives academic research support by the Deutsche Forschungsgemeinschaft (DFG; GA2031/1) and received commercial research support in form of travel grants by the Movement Disorders Society, Actelion, Ipsen, Pharm Allergan and Merz Pharmaceuticals.

Roberto Erro
Has been partly supported by COST Action BM1101 (reference: ECOST-STSM-BM1101-160913-035934

Andrea E. Cavanna
Nil

Kailash P. Bhatia
Funding for travel from GlaxoSmithKline, Orion Corporation, Ipsen, and Merz Pharmaceuticals, LLC; serves on the editorial boards of Movement Disorders and TherapeuticAdvances in Neurological Disorders; receives royalties from the publication of Oxford Specialist Handbook of Parkinson's Disease and Other Movement Disorders (Oxford University Press, 2008); received speaker honoraria from GlaxoSmithKline, Ipsen, Merz Pharmaceuticals, LLC, and Sun Pharmaceutical Industries Ltd.; personal compensation for scientific advisory board for GSK and Boehringer Ingelheim; received research support from Ipsen and from the Halley Stewart Trust through Dystonia Society UK , and the Wellcome Trust MRC strategic neurodegenerative disease initiative award (Ref. number WT089698), a grant from the Dystonia Coalition and a grant from Parkinson's UK (Ref. number G-1009).

Video 1

Neurological examination. A. Spontaneous jerky and on occasion patterned movements of the upper body interfering with normal action execution on finger tapping. Also, palilalia (repetition of number “ninety-nine”) during loud counting. B. Exaggerated stimulus-triggered repetition of hand clapping and foot tapping. C. Bouts of echolalia, self-triggered with the word “no”, and triggered by the voice of the patient’s mother for the words “Christmas” and “Icecream”. D. Stimulus triggered echopraxia with sudden arm elevation following the examiners movement; startle-akin response on hand clapping with immediate repetition; delayed and exaggerated imitation of examiners right arm movement during stance. E. Unusual jerky movements during gait.

References:

[1] Ganos C, Ogrzal T, Schnitzler A, Munchau A. The pathophysiology of echopraxia/echolalia: Relevance to Gilles De La Tourette syndrome. Mov Disord. 2012;27:1222-9.
[2] Bakker MJ, van Dijk JG, Pramono A, Sutarni S, Tijssen MA. Latah: an Indonesian startle syndrome. Mov Disord. 2013;28:370-9.
[3] Gilles de la Tourette G. Etude sur une affection nerveuse charactérisée par de l'incoordination motrice accompagnée d'écholalie et de coprolalie. Arch Neurol. 1885;9:19–42,158–200.
[4] Baizabal-Carvallo JF, Jankovic J. The clinical features of psychogenic movement disorders resembling tics. J Neurol Neurosurg Psychiatry. 2014;85:573-5.