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Gilles de la Tourette syndrome in a cohort of deaf people

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ABSTRACT

We present six patients with Gilles de la Tourette syndrome (TS) who are also deaf. TS has been observed previously, but rarely reported in deaf people, and to date, so called "unusual" phenomenology has been highlighted. TS occurs almost worldwide and in all cultures, and the clinical phenomenology is virtually identical. In our cohort of deaf patients (we suggest another culture) with TS, the phenomenology is the same as in hearing people, and as in all other cultures, with classic motor and vocal/phonic tics, as well as associated phenomena including echo-phenomena, pali-phenomena and rarer copro-phenomena. When "words" related to these phenomenon (e.g. echolalia, palilalia, coprolalia or mental coprolalia) are elicited in deaf people, they occur usually in British Sign Language (BSL): the more "basic" vocal/phonic tics such as throat clearing are the same phenomenologically as in hearing TS people. In our case series, there was a genetic predisposition to TS in all cases. We would argue that TS in deaf people is the same as TS in hearing people and in other cultures, highlighting the biological nature of the disorder.

1. Introduction

Gilles de la Tourette syndrome (TS) is a childhood onset neurodevelopmental disorder characterized by multiple motor and vocal (phonic) tics, lasting longer than a year and with a distinct spectrum of complex tic-related behaviours and associated co-morbid conditions and co-existent psychopathologies (Robertson, 2000, 2011, 2012; Singer, 2005).

TS is common with a prevalence of 1% in youngsters aged 5–18 years world-wide apart from Sub-Saharan Black Africa; TS is much more common in people with special educational needs and in those with autistic spectrum disorders (Eapen et al., 1997; Robertson, 2008a,b; Knight et al., 2012). Most individuals who are diagnosed with TS in both community and even clinical studies, are often mild as far as tics are concerned, but by far the majority nevertheless have co-morbid disorders, co-existent psychopathologies, behavioural and/or learning difficulties (Freeman et al., 2000; Khalifa and von Knorring, 2003, 2005).

We suggest that the co-morbid conditions which occur in approximately 90%, include attention deficit hyperactivity disorder (ADHD), obsessive–compulsive behaviours (OCB), disorder (OCD), and autistic spectrum disorder (ASD). We also suggest that the co-existent psychopathologies are also common and include depression, anxieties and phobias, Oppositional Defiant Disorder (ODD), Conduct Disorder (CD), Personality Disorder, aggressive behaviours, episodic rages and, less commonly, Bipolar Affective Disorder (Freeman et al., 2000; Khalifa and von Knorring, 2003, 2005; Robertson, 2000, 2006, 2012). However, these co-existent psychopathologies are thought to be not an integral part of the condition but rather the result of other genetic and environmental factors (Eapen and Robertson, 2008; Robertson et al., 2014). Self-injurious behaviours are integral to GTS, predominantly obsessional in nature, and differ from the self-mutilation and deliberate self-harm in other disorders (Robertson et al., 1989), but they have also been suggested to be associated with poor impulse control (Mathews et al., 2004) if severe. Both mild and moderate SIB are associated with increased tic severity (Mathews et al., 2004).

GTS, in the majority of cases, is genetically determined, although the genetic mechanisms are more complex than originally thought (Fernandez and State, 2013). Other suggested aetiological factors include pre- and peri-natal difficulties (Hoekstra, 2013) and neuro-immunological abnormalities (e.g. Mell et al., 2005; Murphy, 2013). In the United Kingdom (UK), 4% of

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school aged children are affected by conductive deafness and 0.3% school aged children have sensorineural deafness. In addition, 0.1% of children have permanent hearing impairment. In the UK an estimated 125,000 adults and 20,000 children use British Sign Language (BSL). There are several types of deafness including congenital and acquired, with a variety of causes and also with varying degrees of severity (see Table 1).

Few reports of an individual with both TS and deafness have been documented. (Cipolotti et al., 1999; Morris et al., 2000; Rickards, 2001; Dalsgaard et al., 2001; Sedel et al., 2006; Chovaz, 2013). In some of these there has been an implication that the TS is phenomenologically different to that encountered in TS in hearing people. Signing coprolalia (Morris et al., 2000; Rickards, 2001) and signing complex "vocal tics" (Dalsgaard et al., 2001) such as echolalia have been reported. The most recent documentation (Chovaz, 2013) highlighted how knowledge of both deaf people and TS is important for recognizing the condition and if not, TS diagnosis may be missed.

We report a series of a further 6 individuals with TS who are also deaf. The phenomenology of the tics, co-morbid disorders, the self-injurious behaviours (SIB) and the co-existent psychopathology, in our opinion, are the same as in hearing people and, in all cases, there was also a positive family history of TS, tics or OCB. We suggest that despite the previous reports of signing vocal tics and signing coprolalia, TS in deaf people is not different from either an aetiological nor phenomenological point of view to that encountered in hearing people. Deafness as a cultural issue is discussed with regards to TS, highlighting the fact that TS occurs in all most cultures (Robertson et al., 2009; Knight et al., 2012).

2. Methods and case reports

Each patient/subject underwent a detailed clinical interview by MMR using three published schedules for assessing individuals with TS including: The National Hospital Interview Schedule (NHIS) for TS (Robertson and Eapen, 1996), the Diagnostic Confidence Index (DCI: Robertson et al., 1999) and for the severity of tic symptoms, the Yale Global Tic Severity Scale (YGTSS) (Leckman et al., 1989) as well as one-self report scale, the MOVES scale (Gaffney et al., 1994). The NHIS was also used as a structured method for obtaining relevant family history, as well as the diagnosis of TS-associated conditions including obsessive–compulsive behaviours (OCB), disorder (OCD), attention-deficit hyperactivity disorder (ADHD), and the possible co-existent psychopathology, incorporating the relevant questions and items from the Diagnostic Interview Schedule where necessary.

All initial and subsequent interviews were conducted with the assistance of a BSL interpreter. The patients were all diagnosed with TS according to DSM-IV-TR (American Psychiatric Association, 2002) and ICD-10 criteria (World Health Organisation (WHO), 1992). All patients gave written consent for the anonymous publication of the paper. We have anonymised any identifying information.

2.1. Case 1 (AA, male)

Patient 1 is a 15 year old male, AA, was assessed at the request of the Child and Adolescent Mental Health Services (CAMHS), for confirmation of the diagnosis of TS. He had been referred to CAMHS 2 years before, with a long history of violence and aggressive behaviour. The CAMHS team diagnosed him as having TS and with psycho-education, support and behavioural management, his TS symptoms and behaviours improved markedly.

Histories were obtained from the patient (via a signing interpreter), the mother and father, as well as the deaf unit and CAMHS staff. Developmental history was that he was an unplanned pregnancy. He was born full term following normal delivery, and fed well. He never slept well and demanded much attention from his mother. He was a boisterous child, and his mother found the "terrible 2's" very difficult. At the age of 2 years he was hospitalized with meningitis, after which he became deaf and lost his balance: he also lost all his speech. He was eventually given a hearing assessment and as a result, hearing aids.

Between 2 and 4 years AA was described as being very "violent" and aggressive, being difficult at nursery and preschool as well. He was educated in a "total communication unit" (an educational environment which makes use of a number of modes of communication such as signed, oral, auditory, written and visual aids, depending on the particular needs and abilities of the child). Secondary school was very difficult for AA and after one month he began to be excluded regularly. He was bullied at school, despite his being placed and educated in the specialized unit. There is no other significant past history including streptococcal infections as a youngster.

AA was given a cochlear implant at the age of 4 years (a surgically implanted electronic device that provides sense of sound to a person who is profoundly deaf or severely hard of hearing).

Table 1
Current case series and reports from the literature on TS & deafness.

Current series case number & authors from the literature	Age	Age onset GTS	Sex	Type of deafness	Severity deafness	YGTSS score	Diagnostic Confidence Index (%)	Talking ability	Vocal tics	Signing tics	Rx deafness
1. AA	15	10	M	Acquired	Severe	58	94	After CI@5 years	Yes	No	
2. BB	13	11	M	Congenital	Profound	70	88	No	Yes	No	
3. CC	33	12	M	Cong (recessive)	Profound	65	84	No	Yes	Yes	
4. DD	31	12	F	Congenital	Profound	55	44	Not properly	Yes	No	
5. EE	18	12	F	Congenital	Profound	60	71	No	Yes	No	
6. FF	19	6	M	Congenital	Profound	60	70	No	Yes	No	
Cases from the literature											
Cipolotti et al.			M	Congenital							
Morris et al.			M	Pre-lingual					No	Coprolalia	
Rickards			M	Pre-lingual						Coprolalia	
Dalsgaard et al.	10		M						No	Vocal tics	
Sedel et al.	18	Childhood	M	Hypoacousia	Not stated				Yes	Not stated	
Chovaz*	n/s	Childhood		Not stated	Not stated						

CI—cochlear implant; ADHD—attention deficit and hyperactivity disorder; CD—conduct disorder; ASD—autistic spectrum disorder; SIB—self injurious behaviour; OCB—obsessive compulsive behaviour; OCD—obsessive compulsive disorder.

* Chovaz – last case report – E pub 23rd March 2013—so only abstract available.