Recognising Stereotypies and Tics in Co-Existing Autism Spectrum Disorder

Children and Young People with Autism Spectrum Disorder (ASD) commonly present with well recognised unusual body movements, which are often addressed as part of the consultation and diagnostic work-up. Movements such as rocking, spinning and toe-walking have long been thought to serve the function of regulating sensory arousal along with the movements subjectively liked by the child or young person. Concerned parents and care-givers are often reassured by discussions along these lines.

Occasionally, a child or young person may present either during or following the ASD diagnostic process with movements that appear to be more intrusive and/or complex than those commonly observed. As such, these movements may not fit into a simple or well recognised schema. This may lead to diagnostic dilemma and uncertainty regarding appropriate clinical management.

In this report, we will briefly highlight two recent cases to consider how some of the common movement disorders may be encountered, in children with ASD. The aim of this is provide information of relevance when considering the differential diagnosis and management of common, but often misdiagnosed, paediatric movement disorders.

**Case One**

‘A 3 year old boy is referred for assessment with concerns regarding his social communication.

He is reported to have a preference for playing by himself. He is observed to display facial grimacing and writhing hand gestures when playing on his favourite iPad game, when he has favourite toys or in the back of the car. These movements are reported to occur on a daily basis.

His parents have a video of the movements and they can be seen to be paroxysmal, rhythmical movements that occur in a fixed pattern and involve the same muscle groups each time – facial mouth stretching and leg ‘spasms’. He appears alert and happy during the movements and is easily distracted out of the movements, though appears somewhat annoyed at being interrupted. There is a paternal uncle with epilepsy, so his parents are requesting an EEG as they are concerned the movements may be seizures.’

**Clinical Considerations**

1. What differential diagnoses should be considered?
2. Would you request an EEG?
3. What advice will help parental understanding and management?

These movements are typical **Motor Stereotypies**, a general term that
refers to repetitive patterns of movements.

**Definition of Stereotypies**
Clinically, in the Tics and Neurodevelopmental Movements (TANDeM) service at Evelina Children’s Hospital, London we define motor stereotypies as:

- Rhythmical, repetitive and sometimes complex movements that (unlike tics) are relatively constant in pattern and predictable within individuals
- The child presents as engaged and comfortable with the movements and may report enjoyment during them
- No immediate purpose to the movements is evident, though they are generally observed at times of excitement or boredom
- Movements of this nature frequently elicit concern in the observer

We are interested in defining the clinical phenotype of stereotypies, with preliminary evidence that for some children the movements are associated with interesting cognitive processes.¹

**The Differential Diagnoses**
Restricted and repetitive patterns of behaviour contribute to the diagnostic criteria for ASD²; it is therefore essential to distinguish motor stereotypies from circumscribed interests, routines, preoccupations with certain objects, unusual sensory responses and obsessive compulsive behaviours³. In contrast to stereotypies, these behaviours are typically performed at times of anxiety or to reduce feelings of apprehension, whilst stereotypies are associated with excitement or boredom. Identifying the underlying thoughts (e.g. obsessions or worries) is essential to the differential diagnosis of stereotypies from other ritualistic and obsessive behaviours in children with ASD. Importantly, an EEG is not required if movements are consistent with a diagnosis of stereotypies, though it is important to consider other movement disorders (see Box 1).

**Box 1: Differential Diagnoses**
- Myoclonus
- Tics
- Focal seizures
- Restless leg syndrome
- Dystonia
- Chorea
- Drug-induced movements
- Hyperekplexia or startle
- Paroxysmal dyskinesia
- Cataplexy

**Pathophysiology and Prognosis**
Repetitive movements are reported in approximately 60% of typically developing children between 2 and 5 years of age, with a reduction in the frequency of observed movements with age.⁴ In contrast, for children with developmental difficulties (e.g. ASD, sensory impairments, learning disabilities or social isolation) stereotypies are more likely to persist into later childhood and adolescence.

The reduction in movements for typically developing children is thought to reflect a process of privatisation (i.e. an awareness of
It is not damaging to suppress stereotypies, and although initially the child or young person may find it slightly uncomfortable, this sensation will reduce over time in the majority of children. To help children suppress these movements difficulties it will be important to identify when/where the behaviours occur (e.g. during assembly, walking to school). Increasing cognitive load/stimulation at these times may be useful, as can providing the child with a private space where they can engage at an appropriate time (e.g. a quiet area away from main playground). Involving the child in these discussions will be important to increase motivation and engagement (see Box 2).

**Management Strategies**

The most important thing to inform parents is that "stereotypies do not harm the brain." The main area to address in clinical management is the potential impact on social interactions (e.g. peer perceptions) and functional development (e.g. if the child is missing/failing to attend to information in the classroom) due to engaging in stereotypies. Behavioural strategies may be useful to support the natural process of privatisation or to increase the child or young person’s ability to control stereotypies when social awareness is lacking, these include:

1. **Providing an Explanation**

   It may be useful for the child to have an automatic response that they can give if peers ask questions about the movements. For example, “they are movements that I do when I am excited, I find them very hard to control and can’t always stop myself from doing them”. Supporting children to practice their response and educating teaching staff about stereotypies may be required.

2. **Managing the Movements**

   It is important to remember that children find these movements enjoyable, so should not be prohibited from engaging in them completely. Management of parental anxiety may be required. There seems to be an interesting

---

**Box 2: Habit Reversal Tips for Managing Stereotypies**

1. Increase the child’s awareness of when they are engaging in stereotypies by calmly pointing them out. A ‘code word’ or ‘gesture’ can be used to discretely let the child know when they are performing the movements.

2. Provide an alternative response or behaviour that the child can engage with that occupies the hands (e.g. a small toy, blue-tac to fiddle with).

3. Encourage the use of the alternative response though positive praise and reinforcement (e.g. behavioural reward charts).
association with a family history of OCD (often involving counting or checking).

3. Pharmacological Management

Isolated stereotypies do not usually warrant pharmacological management. However, medication may be considered in those with self-injurious or particularly restrictive movements. Selective Serotonin Reuptake Inhibitors (SSRIs) have been previously trialled for use in this area, though the response to medication is inconsistent.

Referral to a Specialist Service

Referral to a specialist service is recommended when there is doubt regarding diagnostic classification or if medication is being considered.

Lesson from Case One

The most useful form of management is to reassure and ‘name’ the movements, whilst increasing parents or caregivers knowledge of the common life course of stereotypies and behavioural strategies.

Case Two

A 9 year old boy attends for review. He has severe ASD, learning disabilities and is in a specialist school. His parents inform you that his behaviour has deteriorated at home and school, despite specific ASD support being in place. He has recently begun spitting, swearing and repeatedly shouting out in an offensive nature. He shows no remorse when swearing, with his parents reporting embarrassment when this recently occurred at church. His teacher described him as lacking concentration and fidgeting in the classroom.

At the clinic, he is observed to have frequent eye blinking and throat clearing. When discussing the swearing he suddenly has a bout of loud swearing, which appears out of his control. Despite his mother’s disapproval he shows no embarrassment following the episode. When you ask about spitting, he spits, his mother apologises for her son being rude and ‘naughty’.

Clinical Considerations

1. Does the behaviour have its origins in a movement disorder?
2. Why does the child not appear distressed by the behaviour?
3. Does the severity of the symptoms reflect the diagnosis?

These movements are likely to be Tics, of both motor and phonic variety.

This case is interesting as the identification of tics is made complicated by the presence of ASD and lack of social appropriateness. Therefore these children and young people do not show the usual remorse or embarrassment seen in children with Tourette Spectrum or tic disorders. This is common in clinical practise.
Due to the highly suggestible nature of tics, the coprolalia (swearing tic) and spitting are exacerbated by discussions and punishments regarding these ‘bad behaviours’, as both actually ‘suggest’ them further and increase anxiety. Therefore they need to be managed in a different way.

It is important to note that coprolalia occurs in less than 20% patients with tics. Unfortunately, this tic is largely over represented in the Media when Tourette syndrome is mentioned, which contributes to misconceptions and parental anxiety.

**Definition of Tics**

Tics are defined as ‘sudden, rapid, non-rhythmic motor movements or vocalisations usually appearing in bouts and waxing and waning in frequency, intensity and location.’  

If the child presents with multiple motor tics and at least one phonic tic, with the tics occurring for a period of over one year then a diagnosis of **Tourette syndrome** can be made. If the tics are either motor or phonic then a diagnosis of **Chronic Motor or Phonic Tic Disorder** is appropriate.

**The Differential Diagnosis**

There are a number of key clinical features that enable the clinician to differentiate tics from other similar repetitive movements (see Box 3)

Diagnosis can be made on the basis of clinical features, however, more detailed investigations may be required if there is: late adolescent onset, the presence of an additional movement disorder, an uncharacteristic deterioration in daily motor and phonic tics who do not find them disabling or impairing, as well as children with a single eye blinking tic that they find extremely anxiety provoking and distressing.

Assessment of tic severity and impairment is best made using the clinician rated Yale Global Tic Severity Scale for Children (YGTSS-C). The MOVES, PUTS and GTS-QoL may also be useful to gain a better understanding of the child’s

---

**Box 3: Key Features of Tics:**

1. Tics are associated with a premonitory urge/feeling to perform the movement (e.g. itch, muscle tension)
2. The premonitory urge is relieved when the tic is performed
3. Tics are highly suggestible
4. Tics are suppressible (though some children find this easier than others)
5. Suppressing tics is not harmful but may be associated with a feeling of discomfort.
6. Onset is typically later than stereotypies in early childhood

**The Severity of Symptoms**

The severity of symptoms does not influence the diagnosis. However, identifying the impact of tics on the child’s functioning is of importance to inform clinical management. For example, in the TANDeM clinic we commonly see children with multiple

---

**Note:**

- "Due to the highly suggestible nature... increasing anxiety. Therefore they need to be managed in a different way.
- "It is important to note that coprolalia occurs in less than 20% patients with tics. Unfortunately, this tic is largely over represented in the Media when Tourette syndrome is mentioned, which contributes to misconceptions and parental anxiety."
awareness of their tics, associated premonitory urge and impact on daily functioning / quality of life.

Given the high co-morbidity of symptoms and difficulties, the term **Tourette spectrum** is useful in clinic, with severity determined by the degree of impairment or quality of life experienced by. There is a common misconception that ‘Tourettes’ is a term reserved for ‘severe cases’, which is not true. The notion should be avoided as it may prevent families from accessing useful resources or support.

**Pathophysiology and Prognosis**

Tourette disorder is organic and has a neurological basis. Terms such as habit, behaviour, functional and psychological should be avoided when explaining tics. Tourette’s has a prevalence rate of approximately 1%, with a male to female ratio of 3:1. Typical age of tic onset is between 4 and 6 years of age. Tics ‘wax and wane’ across development with increased tic severity typically reported between 10 and 12 years of age, followed by a reduction during adolescence and into adulthood. Co-morbidities are common and present in 90% of children with tics, with Attention Deficit Hyperactivity Disorder (ADHD), Obsessive Compulsive Disorder most commonly reported, though other mood and sleep disorders also frequently co-occur. Tics commonly increase in frequency and intensity around periods of excitement (e.g. Christmas) or stress (e.g. exams).

The pathophysiology of tics is linked to the fronto-striatal-thalamo-cortical circuits, so again increased tic control with age is thought to reflect the on-going maturation of frontal structures and associated neural networks. Close links between the limbic system and striatal networks are likely to account for the increase in tics with extremes of emotion. The role of the cerebellum and its connections are still to be determined but likely to be more important than we recognise at present.

**Management Strategies**

There is no ‘cure’ for tics, though there are number of ways to manage the movements and vocalisations. Psycho-education for the child, parents and school is an essential first step to ensure the tics are appropriately managed and the reasons behind the management strategies are understood by all.

The main treatment approaches adopted at the TANDeM clinic are discussed below and summarised in Box 4.

1. **Ignoring the Tics (but with good explanation as to why)**

Due the highly suggestible nature of tics, talking about tics will contribute to an increase in their frequency. So, one effective treatment approach is to ‘ignore’ the tics, though it is important that children and young people feel that they can talk to their parents about the tics, if needed. Ignoring tics can be difficult, especially when they occur at times of relaxation (e.g. whilst watching TV with the family). At these times, it is up to other people to remove themselves from the room, rather than the child with tics being excluded. It is also important that children with tics do not ‘play’ on the notion of not talking about tics, as
again this may increase tic frequency.

2. Behavioural Strategies for Tics

There are a number of behavioural techniques available for managing tics. The main treatment adopted within the TANDeM clinic is Habit Reversal Therapy. This treatment approach encourages children to inhibit the tic and replace it with a movement that is less noticeable or annoying to them and prevents them from performing the tic. For example, for a sudden arm jerking tic, an alternative response might be to tense the muscles and push the arm inwards. Another similar strategy is that of Exposure with Response Prevention (E/RP) where the child learns to tolerate the premonitory urge and resists performing the tic. Awareness of the premonitory urge is required for these techniques and holding the competing response until the urge has passed is essential.

Behavioural techniques of this nature require practice and patience, but over time appear to contribute to a reduction in tic severity and frequency for many children. There are a number of useful self-help resources available, though often specialist psychological support is required to effectively implement these strategies. These therapies should ideally be offered when requested by the child and the pros and cons of delivering intervention need to be considered. Additional ‘therapy homework’ may be a burden and add anxiety increasing tics and and is not always the appropriate approach.

3. School Liaison

School liaison is often useful to aid understanding and provide teachers with helpful strategies to manage tics. For example, it is important that tics are not mistaken for ‘naughty’ behaviours, as it is not uncommon for children to be punished for tics (e.g. echolalia, eye rolling), or excluded from activities due to ‘annoying’ others (e.g. assembly). This is not acceptable and may be discriminatory.

Some children may unconsciously suppress tics during the day at school, probably as a result of being engaged in other cognitive tasks. This is important to know as intuitively one assumes that this involves effort but some children ‘just don’t tic’ at these times and have no idea why. However, for other children the suppression of tics may be effortful and engagement in tic suppression may lessen attentional control and cognitive resources available to complete academic work. For these children, a ‘class pass’ where they have the opportunity to leave the class room to engage in their tics in private may be useful.

Children with tics often present with problems with attention, inhibition and specific learning difficulties. It is therefore important that level of work is appropriate and support is provided if needed. Effective environmental management for attention problems is often essential to help children with tics focus in the classroom (e.g. not sitting near a window, distracting wall displays/things hanging from the ceiling). Similarly, effective management of autism specific difficulties can help the movement disorder due to reducing levels of anxiety.
4. Pharmacological Management

For some children pharmacological management of tics may be appropriate. This is a particularly useful approach if there are co-morbidities that are impairing the child’s functioning at home and/or school (e.g. attention problems, anger outbursts/ rage attacks, sleep problems).

As only a limited number of rigorous studies have taken place most centres use clinical experience to guide their choices. The first line medication favoured in TANDeM is Clonidine; it is useful in coexisting behavioural disorders, sleep onset difficulties and in the presence of comorbid ADHD.

Risperidone and Aripiprazole are also helpful. Haloperidol and pimozide have both been examined in randomised double blind controlled trials and Haloperidol in particular was the favoured choice for many years but these have both in more recent years been over taken by Risperidone which has also passed rigorous trials and has an improved safety profile. We avoid haloperidol now due to the side effect profile.

Interested parties may wish to read further in the European clinical guidelines for Tourette syndrome and other tic disorders. Surgical treatment including deep brain stimulation options are also available in very severe cases, though given the waxing and waning nature of tics these treatments are not a main treatment approach.

In cases where co-morbidities such as attention problems, anger outbursts/ rage attacks, anxiety, learning difficulties are present it is often these that are most impairing. In these situations psychological (e.g. Cognitive Behavioural Therapy) and/or pharmacological management of these difficulties is essential and often has a positive impact on tics.

Information and resources about tics are available through Tourettes Action, a charitable organisation that supports young people, parents, teachers and other professionals www.tourettes-action.org.uk

Referral to Specialist Service

For children with pre-existing co-morbidities a referral to a paediatrician, neurologist and clinical geneticist may be useful. Rare genetic and epigenetic factors may account for these heterogenous disorders, with on-going research in this area. For example, in the above

**Box 4: Tips for Managing Tics**

1. Ignore tics to reduce suggestibility
2. Help children work out an ‘explanation’ to give peers about the tics
3. Encourage the use of suppression and habit reversal strategies (if appropriate)
4. Liaise with the school to ensure appropriate management of tics
5. Consider pharmacological management (if appropriate)
6. Ensure co-morbid symptoms are appropriately assessed and managed (e.g. learning, attention, anxiety, sleep)
Lesson from Case Two:
Tic disorders in people with ASD can be difficult to define as symptoms may be masked. It is key to identify them and differentiate them from stereotypies or behavioural difficulties as management differs.

Take Home Message
The purpose of this report was to highlight the importance of the accurate assessment and management of movement disorders in children with ASD. The two cases reported presented with stereotypies or tics, though it is important to remember that these movement disorders frequently co-occur. Given the contrasting management approaches (i.e. increasing awareness in the child with stereotypies vs. ignoring and not suggesting the tics) it is essential that time is spent carefully defining the movements so that parents and other professionals working with the child adopt appropriate management strategies.

Although beyond the scope of this brief report, children with ASD also often present with epilepsy, so this should always be considered as a potential differential diagnosis to ensure appropriate management of seizures and commonly co-occurring behavioural and psychiatric symptoms.

We hope that this report has highlighted the importance of viewing movement disorders during childhood as a "spectrum", with co-existing behavioural and psychiatric problems being the norm, rather than the exception. As such, management of these co-occurring symptoms often has the largest impact on emotional well-being and quality of life, which in turn has a positive impact on any movements.

References
5 Gogtay, N., Giedd, J. N., Lusk, L., Hayashi, K. M., Greenstein, D., Vaituzis,


Journal of psychosomatic research, 67(6), 497-501


Sarah Mills
Specialist Paediatric Registrar, Lewisham

Sally Robinson
Highly specialised Clinical Psychologist

Tammy Hedderly
Consultant Paediatric Neurologist

TANDeM Service, Evelina Children’s Hospital London

Tammy.Hedderly@gstt.nhs.uk