

13 Tic Disorders

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Introduction and History

Gilles de la Tourette (under the tutelage of Charcot at the Salpêtrière) described the syndrome of tics, echolalia and coprolalia in Paris in 1885, initially in 9 patients (not all of whom he had met). The same syndrome had in fact been previously described by Trousseau 20 years earlier. Interest in this syndrome in Paris was sparked by descriptions of “culture bound” startle syndromes (latah, myriachit and the “Jumping Frenchmen of Maine”). These syndromes were characterised by echolalia, coprolalia and automatic obedience in response to startling stimuli. Tourette noted in his patients that the behaviours occurred in some people in the absence of a clear stimulus. In most of the latter part of the 19th Century and the first 60 years of the 20th Century, psychoanalytical explanations for tics were in vogue. However, in 1961, the first successful treatment of tics with haloperidol was recorded by Seignot and an explosion in biological research into the causes of TS began. The familial/genetic aspects of TS were documented in the ensuing years although a single gene cause for TS looks unlikely. Epidemiological studies have now suggested that TS, as it is currently defined, is common in children (point prevalence around 0.5%), although often mildly expressed. Associated problems with attention, concentration, obsessionality and mood changes are also commonly described. Finally, research into autoimmune mechanisms, has renewed interest in the relationship between TS and Sydenham’s chorea (St. Vitus’ dance).

Clinical Features

Tics are stereotyped movements which may be jerky or dystonic. They are often experienced with premonitory sensation (an itchy, tense or tight feeling which is experienced around the area of the tic or as a mental phenomenon). They can be suppressed at the expense of internal tension. The movement itself often brings relief of the tension and suppression can lead to a “rebound” phenomenon. Sometimes tic-like movements can be entirely voluntary in order to reduce the premonitory sensations. Tics may be present in motor or phonic domains and can be simple (eye blinks) or complex (squatting, jumping or twirling). Diagnosis of tics in children is more difficult as premonitory sensations are harder to elicit (children find them more difficult to describe).

Differential Diagnosis

Current classification covers two areas; idiopathic tic disorders and neurological disorders in which tics present as a symptom.

Idiopathic tic disorders are divided into the following categories:

- Transient tic disorders (disorders lasting less than 1 year)
- Chronic multiple tics (motor or phonic but not both)
- Tourette syndrome (onset before 18 years, motor and phonic tics, no other neurological explanation)
- Tic disorder NOS (for example, a syndrome identical to TS but onset over 18 years)

These distinctions do not have a clear basis in epidemiology or pathology, and the categories are open to a number of criticisms. For instance, the category “transient tic disorder” can only be diagnosed when it has ceased, which diminishes its validity. One criticism of the TS category, levelled at other disorders which have been defined by their “idiopathicness”, is that when a cause is found for some people with the syndrome, they automatically cease to have it by definition.

More recent factor analytical studies have suggested reclassification along the following lines:

- TS simplex (just tics)
- TS complex (tics plus echo-, pali- or copro-phenomena)
- TS plus (TS plus co-morbid conditions such as ADHD or autistic spectrum disorders)

Further advances in brain research will inevitably lead to a reclassification of tic disorders along aetiopathological lines.

Other Conditions with Tics

There are many other disorders of childhood and adulthood which have tics as a symptom. These include neurodegenerative disorders such as Huntington's disease or Wilson's disease, metabolic disorders such as Lesch-Nyhan syndrome and homocysteinuria, neuroacanthocytosis and intoxication with drugs such as amphetamines and cocaine. Tics can appear as a tardive syndrome in those treated with neuroleptic medication and can occur as part of neurodegenerative processes in later life such as cerebrovascular disease. Motor disorder in myoclonus-dystonia related to the DYT11 gene can appear tic-like but is usually faster and not accompanied by premonitory sensations. People carrying this gene often have task-specific dystonias in addition (such as writer's cramp) and may have had periods of alcohol dependence.

In the majority of cases, the diagnosis of TS can be made without extensive investigation. Factors in the history or examination which would trigger further investigation would include:

- Intellectual impairment or developmental delay
- Late onset of symptoms
- Presence of myoclonus
- Progressive (rather than "waxing and waning") course
- No "sensory" symptoms
- Fixed dystonias or task-specific dystonias such as writer's cramp
- Other focal neurological signs
- Specific dysmorphic features (e.g. indicating Fragile X or Down syndrome)
- Short lived period of motor disorder with long symptom-free periods (could be drug intoxication)

Tourette Syndrome (TS)

Diagnostic Criteria

The DSM-IV criteria which currently define TS are:

- Multiple motor and one or more phonic tics
- Onset before 18 years of age
- Symptoms present for 1 year at least
- Not due to detectable neurological disorder or substance abuse

The definition includes non-mandatory characteristics:

- Tics occur many times a day in bouts
- Tics tend to "wax and wane", changing in frequency, severity, type and location over time.

There has also been argument about whether to include the idea of "significant impairment of function" into the diagnosis, as there are many people who fulfil the current criteria for diagnosis of TS but don't show any impairment in function. Many clinicians want to include impairment in the diagnostic criteria but researchers (especially geneticists) do not as it may cloud the search for the most appropriate phenotype.

Epidemiology

Recent community-based epidemiology studies indicate that the prevalence of Tourette syndrome in school age children is 0.5%. This is relatively stable in different countries including Scandinavia, South Korea, China and Mauritius.

Clinical Course

Typical age of onset of TS is between 6 and 8 years with tics starting in the head and neck. Tics tend to wax and wane with symptoms replacing each other over time. The tendency is also for tics to become more complex with time. The average age for the most severe tics is between 12 and 14 years with tics tending to improve in late adolescence or early adulthood. Recent follow-up studies comparing video taped interviews indicate that most people with TS tend to still have tics during adulthood even if they are not aware of having them. There have been few studies looking at the factors that determine whether there is remission or whether the TS symptoms persist into adulthood.

Co-morbidity

Studies of co-morbidity are once again hampered by the problem of ascertainment bias as most are conducted in clinic settings. Obsessive-compulsive behaviours and OCD are very common in TS and share many clinical characteristics with tics. OC behaviours may just be more complex, more goal directed tics. Self-injurious behaviours are seen in a third of clinic patients with TS and it has a compulsive (and sometimes symmetrical) nature. Studies from the veterinary world (especially of horses) show similar patterns of tic-like behaviours and self-injurious behaviours.

Attention deficit problems are very common in TS and may affect around 40% of children attending clinic. However, attentional problems in TS may be caused by tic suppression, obsessional rumination, by medications, depression or anxiety. The recent, community-based epidemiological studies have shown a true co-morbidity with ADHD (not due to ascertainment bias or “pseudo-ADHD”)

Both depression and anxiety are considerably higher in people with TS and these syndromes have a multi-factorial aetiology

Aetiology

Tics probably represent a final common pathway of a variety of developmental problems. The aetiology of TS is unknown. Family studies indicate that TS appears to be heritable alongside an early-onset form of obsessive-compulsive disorder but the commonest form of attention deficit hyperactivity disorder (ADHD) does not share genetic factors with TS. Large genome-wide association studies have confirmed multiple genes of small effect and moderate overlap with Obsessive-compulsive disorder. Genes involving dopamine, histamine and the SLITRK1 gene confer significant risk. Imaging studies have shown changes in basal ganglia volumes and symmetries but no clear, reliable abnormality has been reliably demonstrated. Perinatal events appear to confer extra risk but are neither necessary nor sufficient. Recent research using the Sydenham’s chorea model has shown encouraging results (people with TS having a high positive rate for anti-basal ganglia antibodies) and there is a mild epidemiological relationship between tic onset and streptococcal infection, although this is a common thing in childhood. So far, treatments based on this model have not demonstrated success. A new study, EMTICS, is designed to answer the questions around the auto-immunity hypothesis in TS. A high quality cohort study of children (the ALSPAC study) indicated that maternal alcohol and cannabis use and low maternal weight in pregnancy may also be risk factors.

Management

Management should be holistic and consider the quality of life of the patient and family at the centre.

Education of the patient and family about the illness is vital. This may include genetic counselling where appropriate. Close liaison with schools is important from the point of view of educating teachers about TS and monitoring progress. Statements of special educational needs may be necessary.

Treatment should be focussed on the symptoms that most impair quality of life, development or learning. These may not necessarily be tics. Psychological treatments include habit reversal. Habit reversal may be useful in people with a small number of disabling tics and a number of recent studies support its efficacy, although it requires effort and effect sizes are modest. Drug treatments for which there is reasonable evidence include clonidine (relatively low effect size but better tolerated), risperidone and aripiprazole. Other drugs which have been used include haloperidol (which is poorly tolerated on the whole) and sulpiride (not available in many countries). There has been controversy about whether stimulants used to treat ADHD (such as methylphenidate) have made tics worse. The best evidence, from the US TS Study group, is that stimulants do not make tics worse. This finding needs replication.

For most people, TS is a self-limiting illness and medication should be used sparingly. Side effects can often impair mood, concentration and learning.

Botulinum toxin has been used to good effect in specific tics. Use of Botulinum toxin appears to reduce premonitory sensations in addition to reducing tics. A number of cases have been reported of successful use of Botulinum toxin injections into the laryngeal muscles to minimise coprolalia.

Deep brain stimulation (DBS) has now been reported in around 150 cases of severe TS worldwide. The rationale for its use is sketchy and controlled trials are infrequent. Around nine different targets have been used without a clear rationale for any of them. Targets include the thalamus and both motor and limbic parts of the GPi. In general, results in a relatively intractable group have been good.

Further Reading

Tourette Syndrome (2006) *Advances in Neurology* Volume 99. Eds. Walkup, Mink, Hollenbeck.

Journal of Psychosomatic Research (2009) Volume 67. This is an invited Special edition dedicated to Tics and Tourette syndrome.

Paschou, P (2013) The genetic basis of Tourette syndrome. *Neuroscience and Biobehavioural Reviews*. 37(6):1026-1039.

Martino, D & Leckman J.F (2013) *Tourette syndrome*. Oxford University Press