The Otolaryngologic Manifestations of Tourette syndrome

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OTL & TS

Objectives
Review of Tics
Review of TS in general
Video clips
Review of TS & OTL literature
Case presentations
Resources
What is a Tic?

- Movement that is:
  - Involuntary (but often has a premonitory sensation)
  - Usually rapid
  - Repetitive
  - Nonrhythmic
- Is more easily recognized than precisely defined in words
What is Transient Tic Disorder of Childhood?

- Usually a single tic
- Occurs in the early school years
- Seen in up to 20% of children (M>>F)
- Do not persist for more than one year (by definition)
- Less likely than TS to be associated with other problems or disorders
- Vocalizations are rare
Examples of Transient Tics

- Eye-blinking or -winking
- Eye-rolling
- Shoulder-shrugging
- Head-shaking
- Facial grimacing
- Throat-clearing

Are more noticeable with fatigue or excitement
Single Chronic Tic Disorder

- Last many years
- Unchanging character
- Thought to be closely associated with TS
- Presentation may mask TS if all tics are not observed or part of history is forgotten
What is Tourette Syndrome

- Tourette Syndrome (TS) is a neurodevelopmental disorder characterized by multiple motor and at least one vocal tic.
- Onset is before age 18.
- Tics occur daily (not necessarily simultaneously).
- Wax and wane.
- Must be present for at least 1 year.
Gilles de la Tourette
How Common is TS?

- 0.5 – 1% prevalence
  
  \[ = \frac{1}{100-200} \]

- Majority of cases are mild

- Inheritance – possibly AD, bi-lineal, modifying genes, is genetically heterogeneous
Is there a Differential Diagnosis?

- Seizure disorder
- Autism-like stereotypical behavior
- Stereotypical movement in mental retardation or blindness
- Neural sarcoidosis
- Hemifacial spasm
- Stuttering (secondary manifestations)
- In older patients
  - Tardive dyskinesia
  - Sydenham chorea
  - Amphetamine abuse
  - Schizophrenia
Common Motor Tics

- Eye blinking
- Flicking hair out of eyes
- Mouth opening
- Arm extending
- Facial grimacing
- Bruxism
- Shrugging
- Licking lips
- Rolling eyes
- Squinting
- Hopping
- Skipping
<table>
<thead>
<tr>
<th>Vocal Tics</th>
<th>Inspiratory sounds</th>
</tr>
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<tbody>
<tr>
<td>Throat clearing</td>
<td>Inspiratory sounds</td>
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<tr>
<td>Throat squeaking</td>
<td>Humming</td>
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<tr>
<td>Coughing (patterned)</td>
<td>Expiratory laryngeal noises</td>
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<tr>
<td>Sighing</td>
<td>Shrieking, squealing</td>
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<tr>
<td>Wheezing</td>
<td>Grunting</td>
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<tr>
<td>Gulping</td>
<td>Nasal sounds</td>
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<td>Gagging</td>
<td>Growling</td>
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<tr>
<td>Blowing</td>
<td>Hooting, yelping, barking</td>
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<td>Gasping</td>
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<td>Snorting</td>
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</table>
Common Vocal Tics

- Throat clearing
- Screaming
- Barking
- Snorting
- Spitting
- Grunting
- Sniffing
- Squealing
- Coughing
- Humming
### Head and Neck Motor Tics that may present to the Otolaryngologist

<table>
<thead>
<tr>
<th>Head and Neck Motor Tics</th>
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<tbody>
<tr>
<td><strong>Mouth</strong> stretching/opening</td>
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<tr>
<td><strong>Nose</strong> twitching/wrinkling</td>
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<tr>
<td><strong>Grimacing</strong></td>
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<tr>
<td><strong>Bruxism</strong></td>
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<tr>
<td><strong>Tongue protrusion</strong></td>
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<tr>
<td><strong>Platysma stretching</strong></td>
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<tr>
<td><strong>Spitting</strong></td>
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<tr>
<td><strong>Teeth clicking</strong></td>
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<tr>
<td><strong>Cheek puffing</strong></td>
</tr>
<tr>
<td><strong>Sucking</strong></td>
</tr>
<tr>
<td><strong>Lip smacking, licking, curling, stretching</strong></td>
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<tr>
<td><strong>Jaw movements</strong></td>
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TS Symptom Classification

- Mild
- Moderate
- Severe

Based on frequency, complexity and degree to which they cause impairment or disruption of the patient’s ongoing activities of daily life
Complex Vocal Tics

- Echopraxia – what you’ve seen
- Echolalia – what you’ve heard
- Palilalia – what you’ve said
- Coprolalia – foul language
  - 5% in TS-Only
  - 15% in TS+
  - best known symptom via media
When are Symptoms the Worst and Will they Disappear?

- Worst - usually 2 years before puberty
- Symptoms diminish in adolescence
- Continue to decrease in adulthood
- Some patients do not have a change
Can the Suppression of Tics Occur?

Automatic suppression

Recent study – the doctor’s office is the least likely place for accurate tic observation

At end of visit or while leaving they may be more in evidence

Videotaping can help significantly when there are doubts
TS Prognosis (Tics)

- 1/3 complete remission through adolescence and early adulthood
- 1/3 significant improvement through early adulthood
- 10% unchanged through adulthood
Famous People with TS
Mozart
Samuel Johnson
Michael Wolf
Jim Eisenreich
Mahmoud Abdul-Rauf
Syndromes Associated with TS

TS+

- Obsessive Compulsive Disorder (OCD) – 30%
- Attention Deficit Hyperactivity Disorder (ADHD) – 65%
- Depression, Anxiety – 17%-25%
- Academic problems – 30%
- Rage attacks, behavior difficulties
What is the Etiology of TS?

The most convincing evidence for dopaminergic involvement has come from the dramatic response to neuroleptic treatment.
What is the Etiology of TS?

- Reduced inhibition in motor pathways at multiple levels of the cortical-striatal-thalamo-cortical circuit, particularly the basal ganglia.
TS and the Otolaryngologic Literature


1998 – Abwender et al Severe Stutterers have an increased risk of developing TS. Brain Lang. 1998 May;62(3):455-64


Tourette syndrome and Otolaryngology
(Kozak et al, 1989)

- 72 patients
- Retrospective study – questionnaire
- 82% response rate
- 70% of patients had a tic involving the nose, throat, head or neck
- 56% of patients had throat clearing at some time during course of their disorder
Of those with an “ENT” tic 42% had seen an Otolaryngologist in the past for various complaints (23/54)

15/23 saw an Otolaryngologist after the onset of TS but before the diagnosis was made

1/23 was dx by an Otolaryngologist

1/23 was seen after the dx of TS

6/23 were seen prior to the onset of TS symptoms
7/16 patients saw an Otolaryngologist with a specific complaint related to TS and the diagnosis was not made.

20% had TMJ complaints.

20% had a history of car sickness.
Examples of TS Patients’ Complaints to the Otolaryngologist in our Study

- Sniffing and throat clearing
- Back of throat and nose noises
- Snorting and difficulty breathing
- Facial expressions, tongue thrusting
- Noises like gulping sounds, coughing

Rickards H. Signing Coprolalia and Attempts to Disguise in a Man with Prelingual Deafness. Movement Disorders Vol 16, No. 4, 790-791.
Treatment of TS

- The vast majority of patients do not require pharmacologic treatment
- Education is the key
Treatment of TS

Decision to treat depends on the degree to which TS symptoms are interfering with the child’s normal development.

Emphasis must be on assisting with the normal developmental tasks such as feeling competent in school, developing friendships, etc.
Treatment and TS

- **Education and Reassurance**
- **Monitoring**
- **Family Counseling**
  - natural parental upset about the tics requires lengthy, calm discussion and education, particularly for fathers
- **Academic and Occupational Intervention**
  - tic tolerance can be improved through the development of appropriate understanding by the patient, family and school or work environment
Medical Treatment of TS

- The goal is to bring the tics into a tolerable range, not to eliminate them completely
- Treatment of co-morbid disorders
- Psychiatry and Neurology and Pediatric specialties usually treat medically
TS and Medication

- Clonidine (Catapres)
- Neuroleptics
  - Haloperidol (Haldol)
  - Pimozide (Orap)
  - Fluphenazine (Prolixin)
  - Others
- Atypical antipsychotics
  - Risperadone
  - Olanzapine
Treatment

- Botulinum Toxin
TS + OCD

- Clomipramine (Anafranil)
- Fluoxetine (Prozac)
- Sertraline (Zoloft)
TS + ADHD

- **Clonidine** (Catapres)
- **Stimulants**
  - Methylphenidate (Ritalin)
  - Pemoline (Cylert)
  - Dextroamphetamine (Dexedrine)
- **Tricyclic antidepressants**
Case Report

- 6 year old male referred by local pediatrician
- CC was chronic throat clearing unresponsive to medical treatment x 2 years and Allergy w/u was negative
- Seen by local Otolaryngologist for Adenoid problem which was ruled out
- Parents very upset (particularly the father)
Case Report con’t

- Problems at school and with friends because of the disruptive noise
- History of eye blinking, mild OC Behavior
- Exam was normal but was suppressing throat clearing, however not completely and also had mild shoulder tic
- “I don’t know why, but I just do it”
- Diagnosis: TS
You have made the diagnosis or you think you have made the diagnosis. What now?

How you present this to the family at this point in time is crucial

- Habit, child is anxious, nervous, doing it for attention or ask if there are problems at home

Refer to an expert in your community

- Psychiatry
- Neurology
Case Report 2

- 9 y.o. boy referred for eustachian tube dysfunction
- 3 years of near continuous autoinsufflation of ears and lower jaw movements
- No improvement with tympanostomy tubes elsewhere
- Throat clearing, sniffing and complex limb movements
- Ear exam and audiogram normal
- Refused referral
Consider TS of the child says:

- I have a tickle in my throat
- I have something stuck in my throat
Video
Video
Conclusions

- Patients with Tourette syndrome do present to Otolaryngology
  - Tics often involve the upper aerodigestive tract/head and neck region
- Heightened awareness may allow for earlier diagnosis and prevent unnecessary delays in treatment
Conclusions

Patients with TS can also have other otolaryngologic medical problems such as chronic adenoiditis, allergy, reflux, etc.
Resources

Canada: Tourette Syndrome Foundation of Canada
www.tourette.ca

B.C.: Dr. Roger Freeman, Department of Psychiatry, B.C. Children’s Hospital
www.tourette-confusion.blogspot.com
Gracias