Tics and Tourette Syndrome

Tics & Tourette

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I have nothing to disclose

Objectives
- Define Tics and Tourette Syndrome
- Differential Diagnosis of tics
- Treatment of Tics and Tourette Syndrome
  - medications
  - behavioral therapies
  - surgical options

Objectives
- Identify Common Tourette Co-morbidities
  - ADHD/ADD
  - Anxiety
  - Headache/Migraine
  - OCD/OC Tendencies
  - Stuttering/Language D/O
  - Anger Control/SIB
  - LD
  - LID
  - Sleep Disorders
  - Autism/ASD
- Overview of Co-morbidity Treatments
  - “2 birds with 1 stone”
  - Relative contraindication
- Mistaken associations/non-co-morbidities

What is a tic?
- Repetitive
- Stereotyped
- “Movement Fragment”
- Involuntary (with some volitional control – i.e. suppressability)
- Frequent premonitory urge
- Waxes and wanes
- Migratory
- Cranial to Caudal onset
Tics and Tourette Syndrome

ICD9: 307.21
Transient Tic Disorder of Childhood

- 1 in 10 school age children will have a tic at some point
- Usually self limited and often not recognized by family

ICD9: 307.21
Transient Tic Disorder of Childhood

- Frequently thought to be allergies
  - often seen by A+I
- Frequently thought to be poor vision or dry/irritated eyes
  - often seen by ophthalmology
- Frequently thought to be enlarged tonsils or nasal abnormality
  - often seen by ENT

Tic Differential Diagnosis

- Sterotypies
- Myoclonus
- Dystonia
- Self stimulation
- Complex Partial Seizures/
  Focal Seizures/ Drop Attacks
  Absence Seizures/ Myoclonic Seizures

- RLS
- Cramps
- Chorea
- Akathesia

Pediatrian Neurology and Developmental Neuroscience
4.1

Tics and Tourette Syndrome

ICD9: 307.2
Tics/Tic Disorder NEC

ICD9: 307.22
Chronic Tic Disorder (motor or phonic)

- Persist greater than a year
- May be subtle and only exacerbate when stressed or excited
- May migrate to various tics or same tic persist
- Often not recognized by the individual or his family as abnormal or have justified due to some long ago injury etc.
- Likely not biochemically different than Tourette Syndrome
- Only defined differently by DSM IV

ICD9: 307.23
Tourette Syndrome

- Both Motor and Phonic Tics
- No longer use term verbal tics because this implies made by the mouth and they are not necessarily (e.g. throat clearing, sniffing)
- Lasting > 1 year, most days
- May wax and wane during that year
- Beginning before the age of 18 years
- May have been subtle in childhood then completely resolve and re-occur as adults

Phonic Tics

Breathing or Diaphragmatic Tics

Coprolalia/Echolalia
**ICD9: 307.23**

**Tourette Syndrome: Time Course**

- **Onset**
  - avg 5-6 years
  - frequent by 3 years
  - I have personally seen at 13 months
- **Peak**
  - avg 10-12 years
  - no clear connection to age of puberty or onset months
- **Resolution**
  - majority: minimal to no tics after 18-24 years,
  - may reoccur in later life

**Genetics**

- Bi-Lineal Transmission
- Typically strong FHx of OCD, ADHD (often denied unless asked specifics)
- Often tics in family not recognized (power of observation in clinic)

**Recent Genetic Findings in Tourette Syndrome**

- L-Histidine Decarboxylase — *NEJM*, 5/2010
- Tourette disorder spectrum maps to 14q31.1 in Italian Kindred — *Neurogenetics*, 5/2010
- Tourette syndrome is associated with recurrent copy number variants — *Neurology*, 5/2010
  (same variant implicated previously in autism)
Tics and Tourette Syndrome

**Tic Treatments**
- Alpha2 CNS Agonists
- D2 receptor Antagonists (Traditional Neuroleptics, e.g. Fluphenazine)
- Dopamine Depletion (Tetrabenazine)
- Topiramate
- BNZ
- Habit Reversal Therapy
- DBS

**Tic Complications**
- Peripheral Nerve Compression – Ulnar or Radial
- Radicular Injury – Sciatic Notch Neuropathy Secondary to Hip-Thrusting
- Whiplash Tic Spinal Cord Injuries – Frequently C8-T1
- Repetitive Blunt Trauma
- Self-Mutilating Tics or Obsessions
- Social Stigma

**Tic Complication**

**Tic Exacerbating Meds**
- Stimulants (ADHD)
- Cold Medications / Decongestants
- TCA’s
- SSRI’s
- Levodopa
- AED’s: Carbamezapine, Lamotrigine, PB
- Bupropion
- Street Drugs: Cocaine, Amphetamines
Malignant Tourettes

DBS Pediatric Indications

- Numerous case reports in adults and children with marked tic and OCD improvement (GPI)
- 2009 HED approval for OCD in NA for > 18 y/o
  - No FDA / HED approval for Tourette's
  - European Tourette Guidelines Specify > 18 y/o
- TCH Multidisciplinary Clinic / Team
  (Neurosurgery, Neurology, PM+R, PT/OT, Psychiatry, Neuropsychologist, Child Life, Psychology)
- Baylor DBS Consensus Conference

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Tic Co-Morbidities

- ADHD/ADD > 60%
- Often huge discrepancy between IQ and school/job performance due to concentration
- Fine motor impairments often greater with higher degree of ADHD
- Chorea Minima correlates more to ADHD/ADD than the presence of tics

“Fidgety”
Tics and Tourette Syndrome

Chorea Minima

Anxiety/OCD/OC Tendencies

- 50% with OCD tendencies
- 34% with fully met OCD criteria
- 24% with ADHD and OCD
- Other forms of anxieties
  1) phobias
  2) preoccupations
  3) "my little worrier"
  4) compulsions
  5) separation anxiety
  6) Agoraphobia
  7) night time fears
  8) Trichotillomania

Tic Co-Morbidity Treatments

ADHD
- Alpha2 CNS agonists
- Norepinephrine Reuptake Inhibitor:
  Atomoxetine (Strattera)
- Stimulants
  Dextroamphetamine, methylphenidate
  Dexamethasone, Lisdexamphetamine

OCD

PANDAS/CANS

- Pediatric Autoimmune Neuropsychiatric Disorders Associated With Streptococcal Infections
  1) presence of OCD and/or a tic disorder
  2) prepubertal symptom onset
  3) episodic course of symptom severity
  4) association with GABHS infection
  5) assoc. w/ neurological abnormalities.
    (e.g. ADHD, chorea minima)
**Tics and Tourette Syndrome**

### PANDAS/CANS
- No case of PANDAS resulting in the valvular abnormalities that happens in up to 1/3 of cases of RF who develop Sydenham’s Chorea
- Streptococcal Infection and Exacerbations of Childhood Tics and OCD Symptoms: A Prospective Blinded Cohort Study - Only 5 of 64 exacerbations over 2 years of the 40 PANDAS cases compared to controls were temporally associated (within 4 weeks) with a group A β-hemolytic streptococcus infection. 75% of the clinical exacerbations in cases had no observable temporal relationship.

### PANDAS Consensus Statement
(2009, AHA endorsed by the AAP)
“The PANDAS hypothesis has stimulated considerable research, as well as considerable controversy… [and] should be considered only as a yet-unproven hypothesis… the committee does not recommend routine laboratory testing for GAS, long-term antistreptococcal prophylaxis to prevent, or immunoregulatory therapy (e.g. intravenous immunoglobulin, plasma exchange) to treat exacerbations of this disorder.”

### Headaches in Tourette Syndrome
- Referred neck pain from neck/shoulder tics - Whiplash tic
- Premonitory Urge
  - Patients sometime say the urge itself can be painful
- Migraines
  - 5x the rate of the general population
  - Topamax may be good treatment option

### Myoclonic (Whiplash) Tics

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  - Sleep Disorders

- Fine Motor issues
- OCD/OC Tendencies
- Stuttering/Language D/O
- Anger Control/SIB
- Cognition
- Autism/ASD?

- Mistaken associations/non-co-morbidities

### Tourette Syndrome Language Problems
- Echolalia/Echopraxia
  (very frustrating/embarrassing and they often say they can’t explain why they do this)
- Palilalia
- Coprolalia/Copropraxia
  (Media focuses on this and often parents biggest fear, yet < 10% and probably < 5% in kids)
- Blocking
  (frequently misdiagnosed as stuttering)
Tourette Syndrome

Language Problems

- TS and Stuttering – 7.7% (9% males)
  - Bimodal distribution of stuttering at age 2 to 3 years and ages 4 to 6 years. Many similarities between “non-developmental” stutters and tics
  - Both much more likely to have ADHD and another co-morbidity
  - Suggestion that both may be related to dopamine and may respond to dopamine antagonists
  - Similar CBT techniques effective for both
  - Very high rate of tics in severe stutters

Stuttering and Tics

Behavioral Disorders in Tourette Syndrome

- ODD or CD – 15%
  - males >> females
- Multiple Co-morbidities
  - On average, individuals with TS have just over two additional disorders (comorbidity score of 2.06),
  - Significantly higher in males (2.11) than in females (1.83)
  - Behavioral problems greater among patients with TS and one or more comorbid diagnosis
Learning Disability in Tourette Syndrome

- 27% with specific LD
  - Discrepancy between specific domain and other IQs
  - Math and reading most common
- Dyslexia
- IEP often necessary
  - OHI forms
  - Letters of Diagnosis

Cognition in Tourette Syndrome

- Overall lower IQ
  - Several articles have suggested overall lower IQ but may include ASD and other tic associated disorders
  - IQ often above average
  - Concentration difficulties not a reflection of intelligence
  - May have mismatch between overall IQ and math (LD)
- MRI changes
  - Cortex volumetric changes

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Sleep Disorder in Tourette Syndrome

- 27% w/ Tourette have a Sleep Disorder
  - Multiple Sleep articles quoting multiple sleep disorders
  - Insomnia the most problematic
  - Consider Clonidine qhs
  - Consider higher doses at night of sedating meds (e.g. neuroleptics)
  - Stimulants may contribute to insomnia
- PLM vs. nocturnal tics
  - Tics have been recorded in all stages of sleep

Autism and Tics

- Can you have Tourette and ASD?
  - Mutually exclusive or co-morbid?
- Stereotypies vs. Tics
  - Tics: cranial to caudal vs. often extremities or whole body
  - Age of onset: in stereotypies < 2 y/o vs. tics 4 to 6 y/o
  - Can have both
- OCD vs. ASD
  - Lining up toys
  - Anxious if not following routine

Autism/ASD
Tics and Tourette Syndrome

4.1

Tics and Stereotypies

Autism/ASD/OCD

Autism and Genetics

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Mistaken associations/non-co-morbidities

Mistaken for TD

Mistaken for ADR
**Mistaken for GI disturbance**

- **Todd's Syndrome Resources**
  - www.tsa-usa.org
  - I Have Tourette's but Tourette's Does Not Have Me
    - Parent/Teacher Tourette teaching resource guide
  - www.tourettetexas.org
    - Camp du Balloon Rouge
    - Regional meetings/activities
    - Local school advocacy

**Mistaken for Allergies**

- Frequently thought to be allergies
  - Often seen by A+I
- Frequently thought to be poor vision or dry/irritated eyes
  - Often seen by ophthalmology
- Frequently thought to be enlarged tonsils or nasal abnormality
  - Often seen by ENT

**References**

- 2009 Prevention of Rheumatic Fever and Diagnosis and Treatment of Acute Streptococcal Pharyngitis
- A Scientific Statement From the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young, the Interdisciplinary Council on Functional Genomics and Translational Biology, and the Interdisciplinary Council on Quality of Care and Outcomes Research. Endorsed by the American Academy of Pediatrics
- Continuum (Lifelong Learning in Neurology), “Movement Disorders”, Vol 13, Number 1, Feb. 2007. American Academy of Neurology, Lippincott Williams & Wilkins, Hagerstown, MD.