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
  - ODD/Conduct D/O
  - LD
  - Sleep Disorders
  - Fine Motor issues
  - OCD/OC Tendencies
  - Stuttering/Language D/O
  - Anger Control/SIB
  - Cognition
  - Autism/ASD

• Overview of Co-morbidity Treatments
  - “2 birds with 1 stone”
  - Relative contraindication

• Mistaken associations/non-co-morbidities
What is a tic?
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
Cranial to Caudal
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
ICD9: 307.21
Transient Tic Disorder of Childhood
Tic Differential Diagnosis

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

- RLS
- Cramps
- Chorea
- Akathesia
Tic Differential Diagnosis
Tic Differential Diagnosis
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 then 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)
Like Father Like Son
Like Father Like Son
Genetics

- 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)*
Genetics: High Sibling Rate
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 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
Malignant Tourettes
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  - ADHD/ADD
  - Anxiety
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  - LD
  - Sleep Disorders
  - Fine Motor issues
  - OCD/OC Tendencies
  - Stuttering/Language D/O
  - Anger Control/SIB
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• Mistaken associations/non-co-morbidities
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”
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
  Dexamethylphenidate, Lisdexamfetamine
# Alpha2 CNS Agonist Studies

<table>
<thead>
<tr>
<th>Author/year</th>
<th>N</th>
<th>Study sample</th>
<th>Design</th>
<th>ADHD outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scahill et al.(^{[18]}) 2001</td>
<td>34</td>
<td>ADHD + tic disorders</td>
<td>Guanfacine vs placebo (parallel groups)</td>
<td>Significantly active &gt; placebo on teacher-rated ADHD Rating Scale; not on Conners Parent Rating Scale</td>
</tr>
<tr>
<td>TS Study Group(^{[19]}) 2002</td>
<td>136</td>
<td>ADHD + tic disorders</td>
<td>Clonidine, MPH combination, placebo (parallel groups)</td>
<td>Monotherapies &gt; placebo on Conners Teacher and Parent Rating Scales; combination showed large effect on Conners Teacher and Parent Rating Scales</td>
</tr>
<tr>
<td>Cummings et al.(^{[20]}) 2002</td>
<td>24</td>
<td>TS</td>
<td>Guanfacine vs placebo (parallel groups)</td>
<td>Active no different from placebo but only a few subjects had ADHD</td>
</tr>
<tr>
<td>Palumbo et al.(^{[21]}) 2008</td>
<td>122</td>
<td>ADHD</td>
<td>Clonidine, MPH combination, placebo (parallel groups)</td>
<td>Monotherapies not superior to placebo on Conners Parent or Teacher Rating Scales; combination showed large effect on Conners Parent and Teacher Rating Scales</td>
</tr>
<tr>
<td>Handen et al.(^{[22]}) 2008</td>
<td>11</td>
<td>PDD + ADHD symptoms</td>
<td>Guanfacine vs placebo (crossover)</td>
<td>Guanfacine &gt; placebo on Conners Parent Rating Scale of hyperactivity</td>
</tr>
<tr>
<td>Biederman et al.(^{[23]}) 2008</td>
<td>345</td>
<td>ADHD</td>
<td>Guanfacine XR vs placebo (parallel groups)</td>
<td>All active doses &gt; placebo on clinician-rated ADHD Rating Scale</td>
</tr>
<tr>
<td>Sallee et al.(^{[24]}) 2009</td>
<td>324</td>
<td>ADHD</td>
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<td>All active doses &gt; placebo on clinician-rated ADHD Rating Scale</td>
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**MPH** = methylphenidate; **PDD** = pervasive developmental disorder; **TS** = Tourette’s syndrome; **XR** = extended-release formulation.
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)
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.
“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 a good treatment option
Myoclonic (Whiplash) Tics
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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 vs. Tics
Stuttering vs. 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
Objectives

• Identify Common Tourette Co-morbidities
  ✓ ADHD/ADD
  ✓ Anxiety
  ✓ Headache/Migraine
  ✓ ODD/Conduct D/O
  ✓ LD
  ◦ Sleep Disorders
  ✓ Fine Motor issues
  ✓ OCD/OC Tendencies
  ✓ Stuttering/Language D/O
  ✓ Anger Control/SIB
  ✓ Cognition
  ◦ Autism/ASD?

Mistaken associations/non-co-morbidities
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 Stereotypies
Autism/ASD/OCD
Autism and Genetics
Objectives

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✓ ADHD/ADD✓ Fine Motor issues
✓ Anxiety✓ OCD/OC Tendencies
✓ Headache/Migraine✓ Stuttering/Language D/O
✓ ODD/Conduct D/O✓ Anger Control/SIB
✓ LD✓ Cognition
✓ Sleep Disorders✓ Autism/ASD?

Mistaken associations/non-co-morbidities
Mistaken for TD
Mistaken for ADR
Mistaken for GI disturbance
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
Tourette 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
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*
• Breedveld GJ et al., Tourette disorder spectrum maps to chromosome 14q31.1 in Italaian Kindred, Neurogenetics. 2010 May 2. [Epub ahead of print]
References

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• Continuum (Lifelong Learning in Neurology), “Movement Disorders”, Vol 13, Number 1, Feb. 2007. American Academy of Neurology, Lippincott Williams & Wilkins, Hagerstown, MD.