Tourette Syndrome-a clinical perspective

Jessica R. Oesterheld, MD

Pre-Lecture Exam Question 1

1. **In order to qualify for a diagnosis of Tourette Syndrome, which of the following statements is true?**

A. Coprolalia must be present.

B. Vocal tics should be present for less than 1 year.

C. Only children with chromic motor and phonic tics and one year history of OCD qualify.

D. The tics must be present before the age of 7.

E. Multiple motor tics and one or more vocal tics must be present for more than 1 year.

Question 2

2. **If a boy is diagnosed with PANDAS, which of the following statements is true?**

A. Place him immediately on prophylactic penicillin.

B. His symptoms typically develop slowly over time.

C. He should be treated with plasmapheresis.

D. Monitor future symptoms and if a throat culture is positive, treat with antibiotics.

E. He will likely develop Sydenham's chorea.

Question 3

3. **A 7-year-old boy has mild vocal tics and mild motor tics. Which of the following statements is true?**

A. His symptoms will likely remain about the same.

B. His symptoms will worsen as he approaches his late teens.

C. The phonic tics will tend to lessen over time.

D. His tics will likely peak in the next 2-3 years.

E. His motor tics will worsen as he approaches his teen years.

Question 4

4. An 8-year-old girl meets criteria for mild TS and ADHD. She is failing in school because of the severity of her ADHD. Which of the following statements is true?

A. The presence of tics contraindicates the use of psychostimulants.

B. Clinician should treat her tics first before treating her ADHD symptoms.

C. Dextroamphetamine sulfate is the psychostimulant that should be avoided.

D. Bupropion would help ADHD symptoms and would improve the tics.

E. Treating her ADHD symptoms with psychostimulants or atomoxetine.

Question 5

5. Which of the following statements about the pharmacotherapy of TS is true?

A. Haloperidol is the drug of choice.

B. Drugs shown to be effective may reduce symptoms about 10-40%.

C. ECG's must be done before treatment with pimozide, clonidine, and risperidone.

D. Drugs that improve tics will make ADHD symptoms worse.

E. All tricyclic antidepressants are contraindicated.

Question 6

6. An 8-year-old girl has chronic throat clearing, nose twitching, eye widening, abdominal tensing. She must enter each doorway and walk backwards through it again before entering any room. She "pulls" at her shirt collars and her socks must be "evened up 4 times" each morning. She meets criteria for which of the following?

A. Tourette Syndrome

B. Tourette Syndrome and Obsessive Compulsive Disorder

C. Tourette Syndrome and Atypical Psychosis

D. Chronic phonic tics and Obsessive Compulsive Disorder

E. Chronic motor tics and Obsessive Compulsive Disorder

Question 7

7. **A 12-year-old boy has had improvement in his chronic motor and phonic tics, but he has recently started to worry about "catching the AIDS virus" and he has been washing his hands many times each day. Which of the following statement is true?**

- If his symptoms of OCD worsen, his tics will also worsen.
- If his symptoms of OCD improve, his tics will also improve.
- If this symptoms of OCD impair his functioning, he should be treated either with CBT or an SRI.
- If his symptoms of OCD worsen, medications that might be tried will worsen his tics.
- His symptoms of OCD are likely to be insignificant.

Question 8

8. Parents of a child with TS are asking about the use of a clonidine patch. Which of the following statements is true?

A. The Catapress TTS patch only comes in one strength.

B. The Catapress TTS patch lasts for about 10-14 days.

C. Before using the Catapress TTS patch, oral dosing of clonidine helps establish the needed dosage.

D. The Catapress TTS patch cannot be used while in the shower.

E. If the Catapress TTS patch falls off, the child will have serious hypertension.

History of TS

- 1885 Georges Gilles de la Tourette, student of Charcot
- from 1885-1965 only 50 cases described and treatment was entirely psychoanalytic
- in 1965, TS diagnosis “re-medicalized” with haloperidol: Shapiros and TSA

DSM-IV Criteria for TS

- Fluctuating course of multiple motor and one or more vocal tics
- Tics many times a day for more than 1 year
- Variable locations & frequency of tics over time
- Onset before 18 years (range 2- 15 years)
- Exclude substance abuse and CNS diseases like Huntington Chorea, postviral encephalitis
- Exclude common transient motor or phonic tics by duration and chronic motor **or** chronic phonic tics

Secondary Causes of Tic Disorders (MTTINN) "empty tin"

- Metabolic - Wilson Disease, hyperthyroidism
- Trauma - head trauma
- Toxic - amphetamines, cocaine and speed abuse, neuroleptic tardive Tourette, CBZ, L-DOPA (phonic), CO, manganese
- Infectious - viral encephalitis, Sydenham chorea
- Neoplasm - basal ganglia tumor
- Neurologic - Mitochondrial encephalomyopathy, Parkinsonism, Huntington Chorea, neurocanthocytosis, Meiges Syndrome

Characteris-tics

- clonic or dystonic or tonic
- brief intermittent movements or sounds
- lessen with sleep, (but present through all stages) and purposeful activity
- variable degree of suppression possible: “relatively irresistible” or “temporarily suppressible”
- “sensory tics”-anticipatory urges
- “disinhibition” or complex behaviors known to be dangerous
- “site sensitization”

Common Simple Tics

- Phonic or Vocal: throat clearing, sniffing, coughing, screeching, yelling out, hiccoughing

- Motor: eye blinking, shoulder jerking, head turning, eye widening, dystonic eye movement, abdominal tensing, tongue movements

Common Complex Tics

- Phonic or vocal: repeating words or phrases, echo self or others, changes of prosody or intonation: **echolalia, palilalia, coprolalia**
- Motor: facial grimacing, complex touching movements, kissing, jumping, **echopraxia, copropraxia, "blocking tics"**

Prevalence of Tics and TS

- Tic prevalence-- 1 in 5 children, more motor
- TS prevalence DSM-IV = 4-5/10,000
- Most authors = 1/1,000 for full blown TS in males and 1/10,000 in girls
- Milder variants occur in an unknown percentage of children: in one community study 1/95 boys and 1/759 girls (Burd et al 1986, Hornse et al 2001)

Natural History of TS

- Behavioral problems often develop first with ADHD
- First simple tics 7 years
- Complex tics emerge years later
- Waxing and waning course with volleys of tics replaced by new and different tics
- Sometimes possible to find triggers (anxiety) for exacerbations
- Generally, children with earliest onset are most severely involved, but sometimes, mild cases-> severe and severe virtually vanish
- Worst tics occur between 9-11 years (Leckman et al 2001)
- OC symptoms in late latency of early teens
- Tends to improve in late adolescents (Leckman et al 2001)

Clinical Course of Tourette's Syndrome

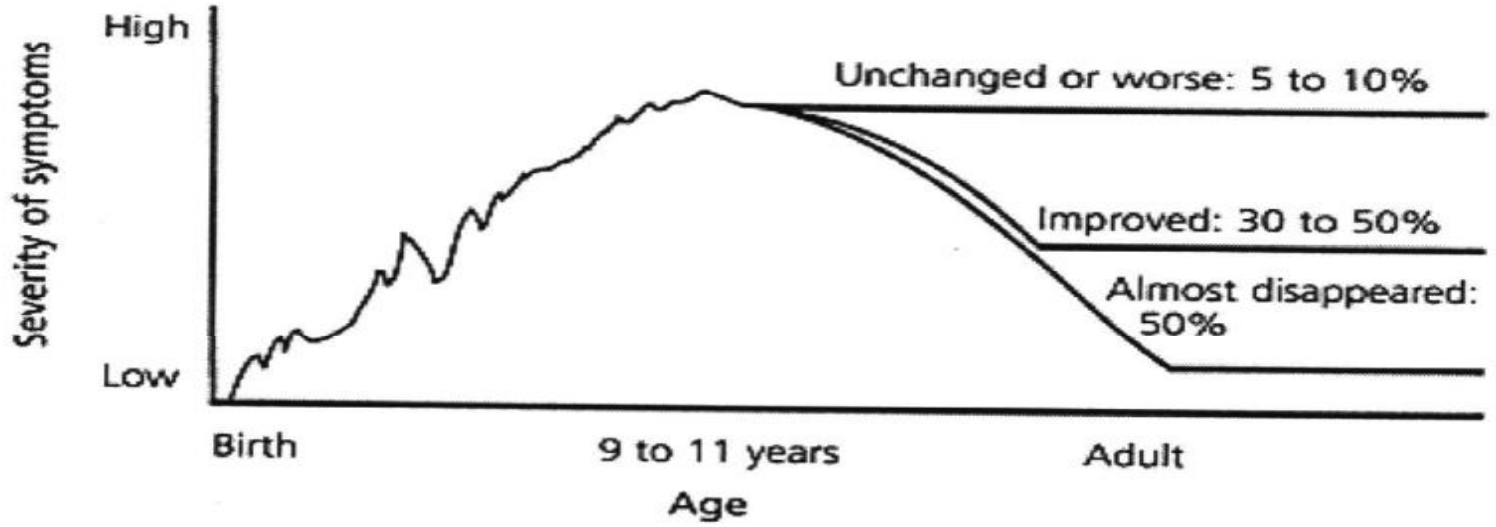

FIGURE 1. The clinical course of Tourette's syndrome. Onset typically occurs before seven years of age and the disorder is usually recognized two to three years after onset. In most children, the severity peaks at nine to 11 years of age. About 5 to 10 percent of patients have an intensifying course with little or no improvement. In about 85 percent of patients, symptoms diminish during and after adolescence.

TS Comorbidity-(King et al 2003)

- ADHD 60-70%
- OCD 40-50%
- Other anxiety disorders 20-30%
- Learning disorders 20- 30%
- Mood Disorders 20-30%
- Aggression or oppositional behavior, enuresis, sleep disturbances, mood lability, SIB, migraines

Obsessions and Compulsions (OC)

- obsessions are repeated mental violent images, or mental repetition of words or phrases, or repeated counting or preoccupation with numbers which despite efforts cannot be completely controlled
- compulsions are repeated actions which are experienced as necessary to do to ward off impending disaster, or, for young children, just done without a sense of why
- it can be difficult to distinguish tics from compulsive behaviors

PANDAS

- Some TS and OCD with acute onset may be caused by antibodies to Strep that coat the basal ganglia (model Sydenham's chorea)
- Lymphocyte BD 8/17 marker associated with rheumatic fever and PANDAS
- Check Strep serologic studies (anti-DNase B and ASO titers) and Strep cultures in kids who develop tics or OCD suddenly
- Consider antibiotic treatment if positive Strep cultures only (Swedo 1997, 1998)
- Research only: immunomodulary therapy (Perlmutter 1999) intravenous immunoglobulin (IVIG) and plasmapheresis

"We must see Tourette's as a window, not merely on the pathophysiology of the basal ganglia and diencephalon, but on what it means to be human and to live in the world."

— Oliver Sacks

General Principles of Evaluation and Treating TS

Treat the whole child and his family

- **assess history and current functioning and family style**
- **need to address prior misunderstanding of symptoms, including parental guilt**

Source of support: http://www.tsa-usa.org

General Principles of Treating TS

Importance of psychoeducational and psychotherapy to family and child

- **Family:** Don't forget what a challenge it can be to raise a TS kid
- **Child:** Who can be bewildered by tics or OCD symptoms; need coherent sense of self: ?control
- **Siblings:** May have feelings about "special treatment" of TS sib

General Principles of Treating TS

Counsel hope and forebearance

- **Most individuals will have a better adulthood than childhood**
- **80-90% will improve substantially**