Outgrowing the Child Neurologist: Transition in Tourette Syndrome

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Road Map

- Why is transition for Tourette syndrome so challenging?
- What are the important considerations in transition planning?
- How can we improve the situation?

First: a few facts about Tourette Syndrome

- Changes in DSM V Criteria vs DSM IV
  - Multiple motor and > 1 vocal tic at some time
  - Clusters of different tics, daily or intermittently for >1 year, with no tic-free period ≥ 3 months
  - Onset < age 18 years
  - Tics cause significant distress or impaired functioning (school, social or occupational)
  - Not caused by direct effect of substance abuse, stimulants or general medical condition
A few more facts about Tourette Syndrome

• Best estimate of Tourette prevalence somewhere between 0.1-1.0%
• Lower bound includes estimated 600,000 children with impairment
• Upper bound includes all cases of multiple tics
• Tics are more common in boys
• Isolated tics occur in 20-25% of all children

And even more facts about Tourette Syndrome

• Symptoms often begin with ADHD then motor tics followed by verbal tics and OCD
• Cephalo-caudal spread and simple to complex progression of tics
• Maturation of circuitry in basal ganglia may explain tendency for tics to diminish with puberty
• Even latest DSM V definition of Tourette syndrome is limited to tics and does not include co-morbid conditions

Natural History of Tourette Syndrome

<table>
<thead>
<tr>
<th>Age Range</th>
<th>Condition</th>
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<tbody>
<tr>
<td>4-6 years</td>
<td>ADHD symptoms</td>
</tr>
<tr>
<td>6-8 years</td>
<td>Simple motor tics</td>
</tr>
<tr>
<td>7-9 years</td>
<td>Obsessive-compulsive behaviors</td>
</tr>
<tr>
<td>8-10 years</td>
<td>Complex motor tics</td>
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<tr>
<td>10-11 years</td>
<td>Simple verbal tics</td>
</tr>
<tr>
<td>12-14 years</td>
<td>Complex verbal tics</td>
</tr>
<tr>
<td>Late Adolescence</td>
<td>Improvement/resolution</td>
</tr>
</tbody>
</table>
Common sequence of symptoms associated with Tourette Syndrome

Slide 7

Common Neuropsychiatric and Developmental Co-Morbidities with TS

- ADHD 60-75%
- Obsessive-compulsive Disorder 20-30%
  - 50-60% have OC symptoms
- Anxiety disorders 20-30%
  - Separation anxiety, panic attacks, generalized anxiety
- Intellectual disability and specific learning disabilities 20-25%
- Autism 5%
- Mood disorders, emotional lability, aggression, rage attacks – near 100% at some point

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Why emphasize developmental disabilities and co-morbidities in TS?

- Tics define the disorder, but co-morbidities often more disabling and longer lasting
  - Only 12% have isolated tics, according to survey of 3500 patients by the Tourette International Consortium
  - Tics are often outgrown while other symptoms often persist

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ADHD in Tourette Syndrome

Treatment of ADHD in Tourette Syndrome

- Behavioral/educational interventions
- α-adrenergic antagonists
  - Guanfacine, clonidine
- Atomoxetine
- Stimulants
  - Methylphenidate, dextroamphetamine
  - Amphetamines (Adderall, Vyvanse)
- Antidepressants
  - Imipramine, buproprion
- Neuroleptics + stimulant as last resort

Potential Impact of Untreated ADHD

<table>
<thead>
<tr>
<th>Low self-esteem</th>
<th>Academic limitations</th>
<th>Smoking and substance abuse</th>
<th>Legal problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>Childhood</td>
<td>Adolescence</td>
<td>Adulthood</td>
<td></td>
</tr>
<tr>
<td>Impaired family and peer relationships</td>
<td>Motor vehicle accidents</td>
<td>Occupational/vocational difficulties</td>
<td></td>
</tr>
</tbody>
</table>
OCD in Tourette Syndrome

Treating OCD

• Cognitive Behavior Therapy (CBT)
• SSRI
  – Fluoxetine (Prozac)
  – Sertraline (Zoloft)
• Combination of CBT and SSRI
• TCA
  – Clomipramine (Anafranil)
• Consider adjunctive atypical neuroleptics

Importance of Premonitory Urges
Strategies for Managing Tourette Syndrome

• Develop a team to address all of the child’s needs
  – Medical (primary care, specialist), psychological, educational, family, community
  – Insist upon open communication between all members of the team

• Involve the child in any treatment plan
  – He/she must understand, participate and “own” the plan, especially by adolescence

• Always maintain optimism
  – Remember the natural history of waxing and waning symptoms plus tendency to improvement over time

What do we mean by Transition?

• The process beginning in childhood to prepare children with chronic illness and their families for adult care

• Transition must be distinguished from transfer - the formal act of handing over care from pediatric to adult health system

As a neurologist, I must remember that for parents, caring for a child with Tourette syndrome can seem like traveling through a long tunnel….
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… but there is always daylight at the other side.

The child neurologist is as skeptical about transition as the patient and family

- We have often been working with the family for years, and we have bonded with the child and family

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- The culture of pediatric care is different from adult medicine – we see ourselves as comprehensive and collaborative vs narrow and focused
- Few adult practitioners seem interested in our complex and behaviorally challenging patients (especially if they are intellectually disabled with limited insurance)

Not all transition is bad

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Not all transition is bad

Transition is a necessary fact of life that should not be like falling off a cliff

Goals of Medical Transition

- To prepare young adults with ability to understand and take responsibility for management of his/her chronic disorder
- To achieve maximal independence in activities of daily living including educational, vocational and social relationships
- To transfer to adult medical providers and ancillary supportive services
Transition has become a part of pediatric care

Core Developmental Issues to Address in Adolescence

- Personal responsibility
- Autonomy (Independence)
- Personal identity
- Body Image
How does Tourette syndrome impact these issues?

- Personal responsibility
  - “Why do I have to take medication? nobody else does.”
- Autonomy
  - “I am not supposed to do most of the things that my friends do—drinking, drugs, sex.”
- Body Image
  - “The pills will make me fat.”
- Personal identity
  - “No one will go on a date with me.”

Transition takes planning

Transition: General Concepts

- Transition planning should start by age 12-16, and should include:
  - Preparation for higher education, vocational support, employment and maximal independence
  - Awareness of available resources and how to access them for on-going support
  - Recognition that adult services can be limited (especially for those with intellectual disability or behavioral disorders)
Transition in Tourette Syndrome

- Most teens outgrow tics, but persistent problems are common with complex Tourette syndrome (i.e. ADHD, OCD and anxiety disorders)
- Even if tics and behavior are under control, must anticipate risk of sub-threshold problems leading to academic challenges, difficulty maintaining job, substance abuse

Perceived Barriers to Successful Transition

- Patient and/or family unwilling to transition (72%)
- Adult providers lack experience (71%)
- Not enough adult providers (68%)
- Multiple Providers (Fragmented care) (62%)
- No time to discuss transition (58%)

Goldberg B, Unpublished 2008

Other barriers to successful transition

- Lack of knowledge of community resources
- Difference in pediatric vs adult culture
- Aging out from pediatrics, but patient unready to take adult responsibility
- Funding changes
- Transportation
- Lack of reimbursement for transition services
How can we overcome perceived barriers to successful transition?

- Develop explicit transition policy
  - goal as part of lifelong preparation for successful adult life (not “kicking to the curb”)
- Discuss differences between pediatric and adult model of care starting at early age
  - Shift from family to individual
  - Youth as decision-maker

My personal Transition advocacy agenda: What can be done to improve the process?

- Identify barriers to transition
- Establish a local transition program
  - Develop collaborations with adult neurology, primary care providers, disability services
- Create national transition resources under auspices of Child Neurology Foundation
  - Transition Consensus Panel

CHOP Neurology Transition Model

- Clinician referral
- Review of medical chart and preparation of detailed summary by transition specialist
  - Diagnosis, evaluation including past medications, most recent psychoeducational evaluation or school records, if available
- Assessment of readiness for independence
- Identification of appropriate adult resources within Penn system and in local community

CHOP Transition Program

REACH and REACH for College
(Rapport, Empowerment, Advocacy through Connections and Health)

- Self-care
- Self-advocacy
- Care coordination
- Wellness
- Relationships
- Eligibility for insurance and federal/state programs
- Navigating the medical system
- Advocating for support services

Opportunity for young people and their parents to gather information, network and socialize.

Behind the Scenes

- Transition Special Interest Group
  - Representatives throughout the hospital from every department
  - Partnership with the electronic medical record
- Chair’s Initiative on Transition
  - Development of hospital wide policies tailored to needs of individual programs
  - Utilize EMR to reduce time factor and facilitate compliance
  - Empower patients and families to pursue transition
Initial Collaborations with UPenn

- Epilepsy
- Neuromuscular program
- Movement disorders
- ADHD
- Autism
- Tourette syndrome
  - Adult neurologist at CHOP
  - Penn Adult Developmental Disabilities program

Transition is very gratifying when it works
Transition is very gratifying when it works.
...and even though it seems that transition can be an impossible feat...

...it is necessary, all patients deserve to graduate to adulthood...

...and we must remember that the goal is TRANSITION – NOT JUST TRANSFER
With appreciation to all of my wonderful, challenging patients ....
.... in the hope that they will all grow to their full potential