Outgrowing the Child Neurologist: Transition in Tourette Syndrome

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ROAD MAP

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

FIRST: WHAT WE KNOW ABOUT TOURETTE SYNDROME

A FEW FACTS

- Definition - 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

- Even latest DSM V definition of Tourette syndrome is limited to tics and does not include co-morbid conditions
- Best estimate of prevalence of chronic motor-verbal tics (i.e. Tourette syndrome) between 0.1-1.0%
- Lower bound includes estimated 600,000 children with impairment
- Upper bound includes all cases of multiple tics

AND EVEN MORE FACTS

- Occasional, isolated tics in 20- 25% of all children
- Tics frequently progress downward and from simple to complex
- Etiology of tics is incompletely understood – most affected have familial basis, but latest data suggest minor genetic changes in de novo cases
- Tics more frequent in males and OCD in females in affected families
- Maturation of basal ganglia circuitry may explain tendency for tics to diminish with puberty

LASTLY: IMPACT OF CO-MORBIDITIES ON TS

- Even latest DSM V definition of Tourette syndrome is limited to tics and does not include co-morbid conditions
- According to a large survey of individuals with TS, over 90% have co-existing neuropsychiatric disorder (ADHD, OCD, mood/anxiety disorder) as well as sleep disorder, migraine, etc.

ADHD: MOST COMMON CO-MORBIDITY

- ADHD often precedes tics by 2-3 years
- Typical age of onset of ADHD is 4-6
- Criteria for ADHD met in 60% of individuals with TS
- Stimulants may exacerbate and/or provoke tics in predisposed children (but do not cause them)
- Tics unlikely to worsen with stimulants, but waxing and waning course may coincide
- Co-morbid ADHD predicts academic problems, even after factoring out learning disabilities and tic severity
- In addition, untreated ADHD itself contributes to car accidents, substance abuse, vocational difficulties

OCD AND OBSESSIVE-COMPULSIVE BEHAVIOR

- Any OC mannerisms in 50-60% of individuals with TS; can be difficult to distinguish from repetitive tics
- 20-30% meet full DSM V diagnosis for OCD
- Typical age of onset 7-9 years
- Repetitive or ritualistic behaviors can be age appropriate; some children are shy, anxious, timid or resistant to change
- OCD may overlap with anxiety, depression, school problems, difficulty with routine tasks
- Diagnosis of OCD requires excessive time, distress and interference with daily activities

OTHER COMMON CO-MORBIDITIES: ANXIETY

- Some symptoms seen in 50% of individuals with chronic tic disorders
- Anxiety disorders take different form – separation, social, selective mutism, specific phobia, generalized, panic
- Youth with anxiety have more severe tics; youth with more severe tics have more anxiety
- Tics worsen with increased stress/anxiety
- Anxiety associated with lower quality of life

OTHER COMMON COMORDITIES: DEPRESSION

- Teens and adults with chronic tic disorders at increased risk
- Associated with increased tics and functional impairment
- Predictive of need for psychiatric hospitalization
- Associated with lower quality of life
- Persistence of tics into adulthood is strongest predictor of suicide

OTHER COMMON COMORDITIES: RAGE

- Definition: out of proportion to inciting event and atypical of baseline character
- Seen in 20-40% of youth with TS
  - Often associated with co-morbid OCD ± anxiety
- Most distressing and disabling symptom reported in adolescents with TS
  - Increases family dysfunction and social conflict
  - Leading cause of psychiatric hospitalization

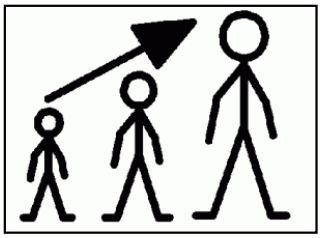
WHAT PARENTS SHOULD REMEMBER

- Tourette syndrome is disorder to be managed, not disease to be eradicated
- Tics common, typically "wax and wane" and rarely cause functional impairment or injury.
- Therefore, understanding always necessary but treatment not always indicated.
- All treatments symptomatic rather than curative
- When intervention indicated, CBT recommended as first line for mild-moderate tics, assuming child mature enough and willing, treatment accessible
- Safe and effective drugs available; recommended for moderate-severe tics, preferably in conjunction with CBT

STRATEGIES FOR PARENTS TO CONSIDER

- Develop a team to address **all** needs and insist upon open communication between all members
  - Medical (primary care as well as specialist)
  - Behavioral health
  - Educational
- Involve the child in any treatment plan
  - He/she should understand, participate and "own" the plan, especially by adolescence
- Optimism is always important
  - Natural history includes "waxing and waning" symptoms
  - Most individuals improve over time
  - Positive attitude has been shown to minimize impact

MOVING ON TO TRANSITION



WHAT DO WE MEAN BY (MEDICAL) TRANSITION?

- Transition is a process beginning in early adolescence to prepare children and their parents/caregivers with chronic medical conditions
- Distinguished from transfer - the formal act of handing over care from pediatric to adult health system
- Transition may look different for every patient, depending on medical complexity and whether a patient has intellectual or physical disabilities

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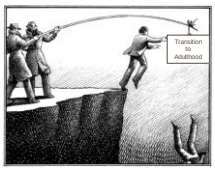
GOALS OF MEDICAL TRANSITION

- 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 avoid the transition cliff of missed opportunities and lost services



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WHY FOCUS ON TRANSITION?

- Adulthood is inevitable
- Youth need to establish independence to the best of their potential, often despite physical or intellectual disabilities
- Young adults with neurological (and neuropsychiatric) disorders do not preclude general medical issues that are best managed by adult physicians
- Delaying transition simply "kicks the can down the road"

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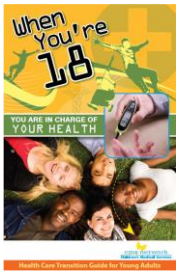
TRANSITION IS NOW AN ACCEPTED PHASE OF PEDIATRIC CARE



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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- Few adult practitioners seem interested in our complex and behaviorally challenging patients (especially if they have limited insurance or intellectual disability)

TRANSITION: GENERAL CONCEPTS

- Transition should start by age 12-14
- There should be annual reevaluation of goals and progress
- Age appropriate concerns will change during process
- Importance of legal considerations if youth unable to manage affairs due to intellectual or physical disability
- It takes time to identify adult provider
- Transfer is only the last step

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WHY IS TRANSITION SUCH A LONG PROCESS

- Adolescence is a challenging time, and any medical condition, especially one with neuropsychiatric dimensions, makes it more difficult
- It takes time to develop knowledge of disorder, self-management skills, limitations, consequences of poor choices
- Patients and families need to learn about available resources and how to access them for on-going support

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WHAT MAKES TS TRANSITION SO DIFFICULT

- Tics define the disorder but co-morbidities often more disabling and longer lasting
- Most common comorbidities are ADHD and OCD
- Other frequent issues include mood/anxiety disorders, sleep disorders, learning disabilities, ASD
- Tics and behavioral issues make challenges of adolescence more difficult

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WHAT MAKES TS TRANSITION SO DIFFICULT

- Most teens outgrow tics, but ADHD, OCD and anxiety disorders may persist
- During adolescence, mood disorders, emotional lability, aggression, rage approach 100% at some point
- Even if tics and behavior are outgrown or controlled, must consider sub-threshold issues leading to academic challenges, difficulty in maintaining relationships, substance abuse
- Recognition that few adult neurologists will manage TS , ADHD or behavior disorders; may need to switch to primary care or psychiatry

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PRACTICAL CONSIDERATIONS FOR THE PRACTITIONER

- Importance of clear office transition policy
- Need to encourage adolescent to accept responsibility for self-care and self-advocacy
- Transition is ongoing process with need for periodic reevaluation
- Consider guardianship/power of attorney in cases of IDD and physical disability
- Medical home is important for all, but critical for complex or challenging patients
- Transition model is equally valuable even if provider unchanged

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HOW PARENTS CAN SUPPORT TS TRANSITION

- They can encourage knowledge of medical condition
  - Identify all aspects of disorder, medications, emergency plan, when to seek medical attention
  - Importance of healthy habits and making good choices
- Parents can accept an evolving role
  - Shift from youth's "advocate" to "ally"
  - Need to remember that it's about the youth and not about them
  - Abandon idea that the youth will make same choices as they would
  - "Letting go" might look different in each case

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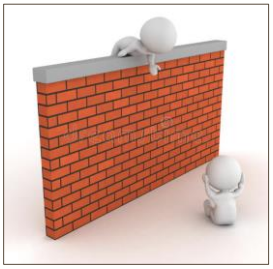
FROM BARRIERS TO SOLUTIONS



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FROM BARRIERS TO SOLUTIONS:
NOT YET READY FOR ADULT MODEL OF CARE

Barrier:

- Lack of understanding of difference between pediatric (family-centered) and adult (patient-centered) care
- Youth's ignorance/denial of consequences
- Overprotective parents

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Solution:

- Gradually introduce individual office time beginning in early teens
- Provide training to meet psychosocial needs
- Utilize local resources (e.g. NJCTS) to encourage independence and self-advocacy

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FROM BARRIERS TO SOLUTIONS:
TEEN ISSUES

Barrier:
- Lack of adherence to medical plan
- Risk-taking behaviors
- Impact of psychiatric co-morbidities (i.e. mood, anxiety)

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TEEN ISSUES

Barrier:
- Lack of adherence to medical plan
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Solution:
- Screen for psychosocial problems
- Visits can include opportunity to practice 3 sentence summary
- "Health Passport" on flash drive or cell phone app

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FROM PRINCIPLES TO PRACTICE: RESOURCES OF THE CHILD NEUROLOGY FOUNDATION

- Office transition policy
- Transition readiness checklist
- Self-care assessment
  - Separate forms for individuals with IDD
- Transfer packet
  - Transfer Letter Sample
  - Plan of Care
  - Medical Summary

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CNF MEDICAL SUMMARY

- Contents
  - Diagnosis/etiology
  - Current medications/laboratory results
  - Previous treatments and evaluations
  - Significant past procedures
  - Protocol for emergency care
- In addition
  - Assessment of youth's knowledge of medical condition, self-management skills and decision-making capacity
- Summary best completed at least one year prior to transfer

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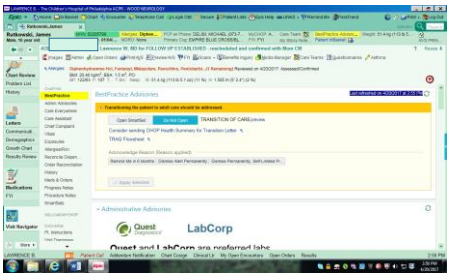
LOCAL SOLUTIONS:
UTILIZING THE EMR: BEST PRACTICE ADVISORY



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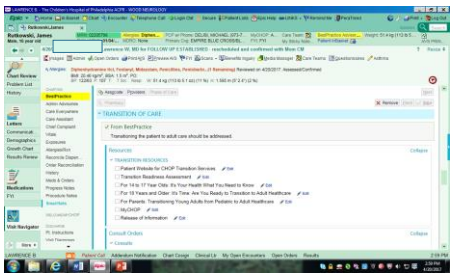
LOCAL SOLUTIONS:
UTILIZING THE EMR: TRANSITION RESOURCES



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NEUROLOGY MATTERS (AN ADVERTISEMENT)

- Seeking 15 volunteers for pilot project to help middle school age children (age 10-14) with early transition
- Free, 6-week on-line program designed to develop critical writing skills, provide opportunities to confront personal challenges and to share with similarly affected youth
- In addition to increasing disease self-knowledge and self-management skills, the sessions will encourage expression of feelings which target the social and emotional impact of living with TS
- Please contact NJCTS or me to sign up; feel free to ask any questions at brownla@.chop.edu

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WHERE TO GO FOR RELIABLE ON-LINE RESOURCES

www.childneurologyfoundation.org/transitions

www.gottransition.org

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SO, EVEN THOUGH IT SEEMS THAT TRANSITION IS AN IMPOSSIBLE TASK….

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…AND IT IS OFTEN AN EXPLOSIVE PROCESS…

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…. ALL PATIENTS DESERVE TO GRADUATE TO FULL ADULTHOOD

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