TOURETTE SYNDROME IN THE PEDIATRIC POPULATION

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Objectives

- Review current literature on TS with an emphasis on the diagnostic criteria and symptomology, as well as identify prevalence and etiology of the disorder.
- Explore and identify co morbidity conditions affecting children with TS.
- Identify current treatment practices for TS
- Identify implications for the future pediatric nurse or school nurse role and practice for children and families affected by TS.

Gilles de la Tourette Syndrome

- Georges Gilles de la Tourette
  - French physician, known today as a neurologist
  - 1885> described nine patients with motor & vocal tics
  - His major contribution was to clearly define this movement disorder

Most famous

- 86-year old woman
  - Marquis de Dampierre
- Exhibited chronic motor and vocalization tics throughout her lifetime
  - Starting at age 7 until her death

Definition of Tourette's

- DSM-IV-TR defines TS as:
  - Multiple motor tics and one or more vocal tics.
- American Psychiatric Association (2000) defines tics as:
  - "sudden, rapid, recurrent, nonrhythmic, stereotyped motor movements and vocalizations" (p.108).

Tics

- Need to rule out other conditions
  - Wilson's disease
  - Huntington's chorea
  - Lesch-Nyphan disease
  - Post-viral encephalitis
  - Substance abuse
  - Any other underlying medical conditions

2 Classification of tics

- 1. Simple tics
  - Related to chronic motor movements
    - Blinking of the eyes
    - Abdominal tensing
    - Nose wrinkling
    - Clearing of the throat
    - Shoulder shrugging
    - Facial movements
    - Coughing
    - Yelling
    - Hiccupping
    - Barking
    - Humming
    - Snorting
    - Sniffing

- 2. Complex tics
  - Prolonged movements or noises that utilize several muscle groups
    - Hitting
    - Jumping
    - Smelling an object
    - Deep knee bends
    - Squatting
    - Twirling
    - Repeating parts of a sentence
    - Echolalia
    - Coprolalia
    - Copropraxia

Tics

- Can experience a combination of different tics OR just one specific type of tic at a time
- Motor & vocal tics occur several times a day for several months
- Severity & frequency of tics is different for each child
  - Some tics barely noticeable
  - Some tics occur 30-100 repetitions in a minute

Tics continued

- Frequency of tics does not indicate severity of TS
  - This is because many of the tics are intermittent
    - They come and go over certain periods
  - TS is based on whether the tics are simple, complex, or a combination of the two
  - Onset of phonic tics occurs much later than motor tics

Characteristics of TS

- Begins in early childhood
  - Mean age is 6 to 7 years of age
  - Can begin as early as 2 years of age
  - The earlier a child is affected, the more severe the symptoms (Gilbert, 2006).
  - Symptoms may diminish as child ages

Etiology

- Cause is unknown
  - 60% of all cases may have a hereditary link
  - Autosomal dominant trait (Gelmann & Selekman, 2006)
    - Males are 4 times as likely to exhibit symptoms than females

Theories related to TS

- Decrease in dopamine in the brain
  - Dopamine
    - Neurotransmitter responsible for transmitting information from one nerve cell to another
    - Abnormal levels trigger the development of tics

Streptococcal Infections

- Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS)
  - Chronic group A beta-hemolytic streptococcal infections
    - Decreases brain activity in the basal ganglia, which then decreases the production & use of dopamine (Cavanna, Servo, Monaco, & Robertson, 2009).
    - Present with an abrupt onset of motor tics that occurs with strep infection
    - Continued periods of remission & exacerbation

Prevalence in children

- 2% of the pediatric population
  - 5-30 per 10,000 children affected
  - 10-20% of school age children have transient motor tics for about one year
  - Affects all ethnic groups
    - Shapiro, 2002

CoMorbidity

- 90% of children diagnosed with TS have one or more underlying psychiatric conditions
  - Attention-deficit hyperactivity disorder (ADHD)
  - Obsessive compulsive disorder (OCD)
  - Anxiety disorders
  - Behavioral/emotional problems
  - Mood disorders

TS and ADHD

- 2-12% of children diagnosed with ADHD have TS (Cavanna, et al, 2009)
  - These children often experience an increase in tics
  - Alternative expression of the TS gene, which links these two conditions together (Gaze et al, 2006)

Prognosis

- 50% of children who develop TS early in life will outgrow the tics by age 18 (Shapiro, 2002)
  - Related to brain maturation after pubescence
  - This increases dopamine production
- Presence of co-morbid conditions
  - ADHD & OCD will continue with tics
  - Based on severity of co-morbid conditions

Assessment

- Complete medical history
- Physical examination
  - Rule out other conditions
- Motor and/or vocal tics
  - Present for one year with no tic-free periods greater than 3 months
  - Tics may decrease during periods of sleep
  - Increased fidgeting
  - Difficulty concentrating

continued

- Tics may increase during periods of excitement, stress, or fatigue
- Some children will show an increase in tics due to no underlying cause
- Tics can wax and wane (Gilbert, 2006)

Assessment continued

- Neuroimaging studies
  - Computerized tomography (CT scans)
  - Electroencephalogram (EEG)
  - Magnetic resonance imaging (MRI)
- Historical observations
  - Parents
  - School officials
  - practitioners

Other symptoms

- Antisocial behavior
- Extreme changes in personality
- Inappropriate sexual activity
- Exhibitionism
- Sleep disturbances
- Experience negative responses by others
  - Bullying
  - Ridicule
  - Isolation

Treatment

- 4 main components of treatment for tics associated with TS (Gilbert, 2006)
  - 1. Obtaining a correct diagnosis
  - 2. Understanding of the disorder and concerns with impulsivity and obsessions
  - 3. Collaborating with family members
  - 4. Understanding treatment modalities

Main Goals of Treatment for TS

- Treatment depends on severity of tics & any underlying co-morbid conditions
  - Improve social functioning
  - Improve self-esteem
  - Increase child's quality of life

Pharmacological Treatments

- Neuroleptics
  - Block dopamine production, which controls tics
  - Many undesirable side effects, such as sedative effect, weight gain, impaired cognitive ability, extra pyramidal side effects
  - Examples:
    - Pimozide (Orap)
    - Fluphenazine (Prolixin)
    - Trifluoperazine (Stelazine)
    - Thioridazine (Mellaril)
    - Haloperidol (Haldol)

Pharmacological treatments

- Alpha-adrenergic agonists
  - Inhibit dopamine production, which suppresses tics
  - Clonidine (Catapres)
  - Guanfacine (Tenex)
- Non-dopamine receptor blocking
  - Atomexetine (Straterra)
  - Sertraline (Zoloft)
  - Fluoxetine (Prozac)

TS & ADHD

- Stimulant medications
  - Stabilizes dopamine production in brain
  - Methylphenidate

Behavioral Therapy

- Behavioral therapy
  - Tics are involuntary, but children can be taught techniques to suppress or decrease the negative effects of tics
  - Teaching child & family how to avoid triggers, then provide techniques to lessen the severity and occurrence of tics
  - Family support
  - School interventions

Habit Reversal Therapy

- Awareness training
- Self-monitoring of tics
- Relaxation techniques

Psychotherapy

- Psychotherapy
  - Psychological problems can occur due to TS
  - Due to rejection, isolation, bullying, etc.

Implications for School Nursing

- Open communication
- Safe environment
- Treatment plan
  - Indivdualized Education Plan (IEP)
- Educating students and school personnel on TS

Individualized Student Health Plan

- Provides clear instructions for school personnel
- Behavioral therapy
  - Relaxation techniques
  - Teaching initial triggers of tics
- Monitor height, weight, BMI
  - Start of medication regime
  - Monitor every month for 3 months
  - Monitor every 6 months while receiving treatment

Bullying

- TS children are more prone to experience violence and isolation from peers
  - Reinforcing socially acceptable behaviors in classroom
  - Conducive to positive learning environment
- Bullying prevention programs
  - Focus on empowerment & conflict resolution

Care for the Hospitalized Child with TS

- Open communication
- Family support
- Maintaining routines

Family Support

- Maintain open communication with families
- Provide support services

Community Resources

- Tourette Syndrome Association
  - http://www.tsa-usa.org
- National Institute of Neurological Disorders and Stroke
  - www.ninds.nih.gov
- National Tourette Syndrome Association, Inc.
  - http://tourette-syndrome.com
- Local community resources

Conclusion

- TS is a debilitating neurobehavioral disorder that begins in early childhood
- Diagnosis is difficult
- Treatment is essential to well-being of child and family

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