Tourette syndrome and chronic tic disorders

Psychoeducation for children, adults and families

Objectives

1. To review the clinical phenomenology and diagnostic criteria for tic disorders
2. To discuss the natural history of tic disorders
3. To describe the epidemiology of tic disorders and common comorbid disorders
4. To review recommendations on assessment and treatment of tic disorders

Clinical Phenomenology and Diagnostic Criteria

Tics: Definition

A sudden, rapid, recurrent, non-rhythmic motor movement or vocalization

Fragments of normal motor actions or vocalizations that are misplaced in context

Motor Tics

Simple Motor Tics

- Sudden, brief, meaningless movements
  - Eye blinking
  - Eye movements
  - Grimacing
  - Nose twitching
  - Mouth movements
  - Head jerks
  - Shoulder shrugs
  - Abdominal tensing

Complex Motor Tics

- Slower, longer, more “purposeful” movements
- Rarely seen in absence of simple motor tics
  - Touching objects or self
  - Gestures with hands
  - Dystonic postures
  - Obscene gestures (copropraxia)

Vocal Tics

Simple Vocal Tics

- Sudden, meaningless sounds or noises
  - Throat clearing
  - Coughing
  - Sniffing
  - Screeching
  - Barking
  - Grunting

Complex Vocal Tics

- Syllables, words, phrases or statements
- Odd patterns of speech
  - Sudden changes in rate, rhythm, volume
- Echolalia
- Obscene, inappropriate and aggressive words or statements (coprolalia)

History of Tics

- Suppressibility
- Distractibility
- Suggestibility
- Variability
  - Character of tics
  - Frequency of tics – waxing and waning
- Exacerbation with stress or excitement
- Premonitory Urge

Tourette's Disorder: DSM 5

A. Both multiple motor and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently.

B. The tics may wax and wane in frequency but have persisted for more than 1 year since first tic onset.

C. Onset is before age 18 years.

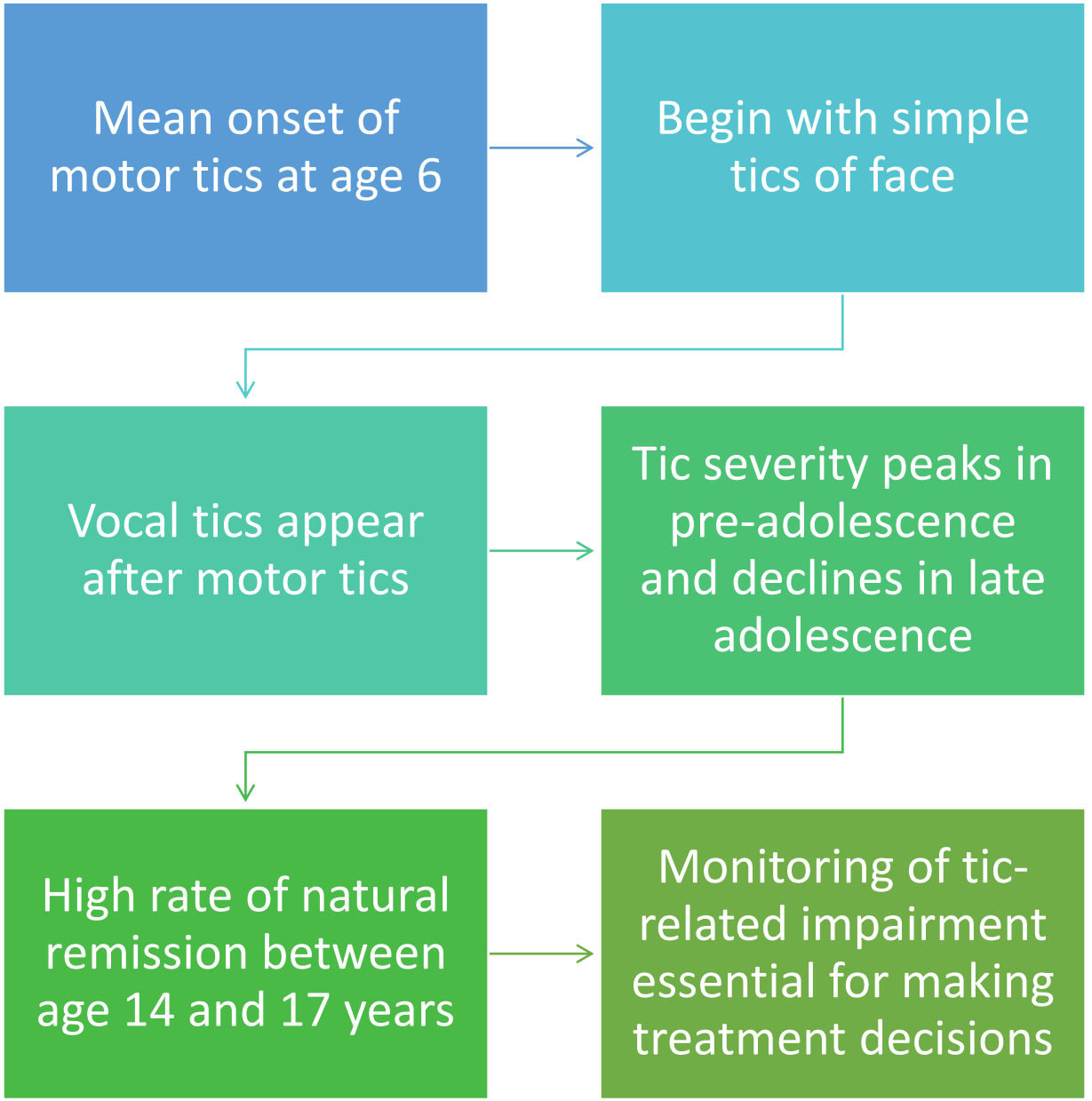
D. The disturbance is not attributable to the physiological effects of a substance or another medical condition

Neurobiology of tic disorders

- Neurochemical and neuroimaging studies suggest dysfunction of the dopaminergic pathways in the cortico-striato-cortico-frontal circuitry
- Other neurotransmitter systems implicated – glutamatergic, GABA-ergic, noradrenergic, histaminergic pathways
- Highly heritable but genetically heterogenous condition
- Environmental factors may play a contributory role
    - Autoimmune dysfunction
    - Pre/perinatal adversity

Natural history of Tourette syndrome

Natural History of Tourette Syndrome



Course of Tourette Syndrome and Comorbidities in a Large Prospective Clinical Study

Camilla Groth, MD, Nanette Mol Debes, MD, PhD, Charlotte Ulrikka Rask, MD, PhD, Theis Lange, PhD, MSc, Liselotte Skov, MSc

Objective: Tourette syndrome (TS) is a childhood-onset neurodevelopmental disorder characterized by tics and frequent comorbidities. Although tics often improve during adolescence, recent studies suggest that comorbid obsessive-compulsive disorder (OCD) and attention-deficit/hyperactivity disorder (ADHD) tend to persist. This large prospective follow-up study describes the clinical course of tics and comorbidities during adolescence and the prevalence of coexisting psychopathologies.

Method: The clinical cohort was recruited at the Danish National Tourette Clinic, and data were collected at baseline (n = 314, age range 5–19 years) and at follow-up 6 years later (n = 227) to establish the persistence and severity of tics and comorbidities. During follow-up, the Development and Well-Being Assessment (DAWBA) was used to diagnose coexisting psychopathologies. Repeated measures of severity scores were modeled using mixed effects models.

Results: Tic severity declined yearly (0.8 points, CI: 0.58–1.01, on the Yale Global Tic Severity Scale [YGTSS]) during adolescence; 17.7% of participants above age 16 years had no tics, whereas 59.5% had minimal or mild tics, and 22.8% had moderate or severe tics. Similarly, significant yearly declines in severity of both OCD (0.24, CI: 0.09–0.39, on the Yale–Brown Obsessive Compulsive Scale for Adults [Y-BOCS] and Yale–Brown Obsessive Compulsive Scale for Children [CY-BOCS]) and ADHD (0.42, CI: 0.32–0.52, *DSM-IV*) were recorded. At follow-up, 63.0% of participants had comorbidities or coexistent psychopathologies, whereas 37.0% had pure TS.

Conclusion: Severity of tics, OCD, and ADHD were significantly associated with age and declined during adolescence. However, considerable comorbidities and coexisting psychopathologies persist throughout adolescence and require monitoring by clinicians.

Key words: Tourette syndrome, prospective study, clinical course, OCD, ADHD

J Am Acad Child Adolesc Psychiatry 2017;56(4):304–312.

Course of Tourette Syndrome

Tic severity declined yearly by 0.8 points on the YGTSS (0.58-1.01) during adolescence

18% of participants above age 16 years had no tics, whereas 60% had minimal or mild tics (YGTSS < 19), and 23% had moderate or severe tics

Significant yearly declines in severity of both OCD (0.24, CI: 0.09–0.39, on the CY-BOCS) and ADHD (0.42, CI: 0.32–0.52, DSM-IV) were recorded

Predictors of the Clinical Course of TS

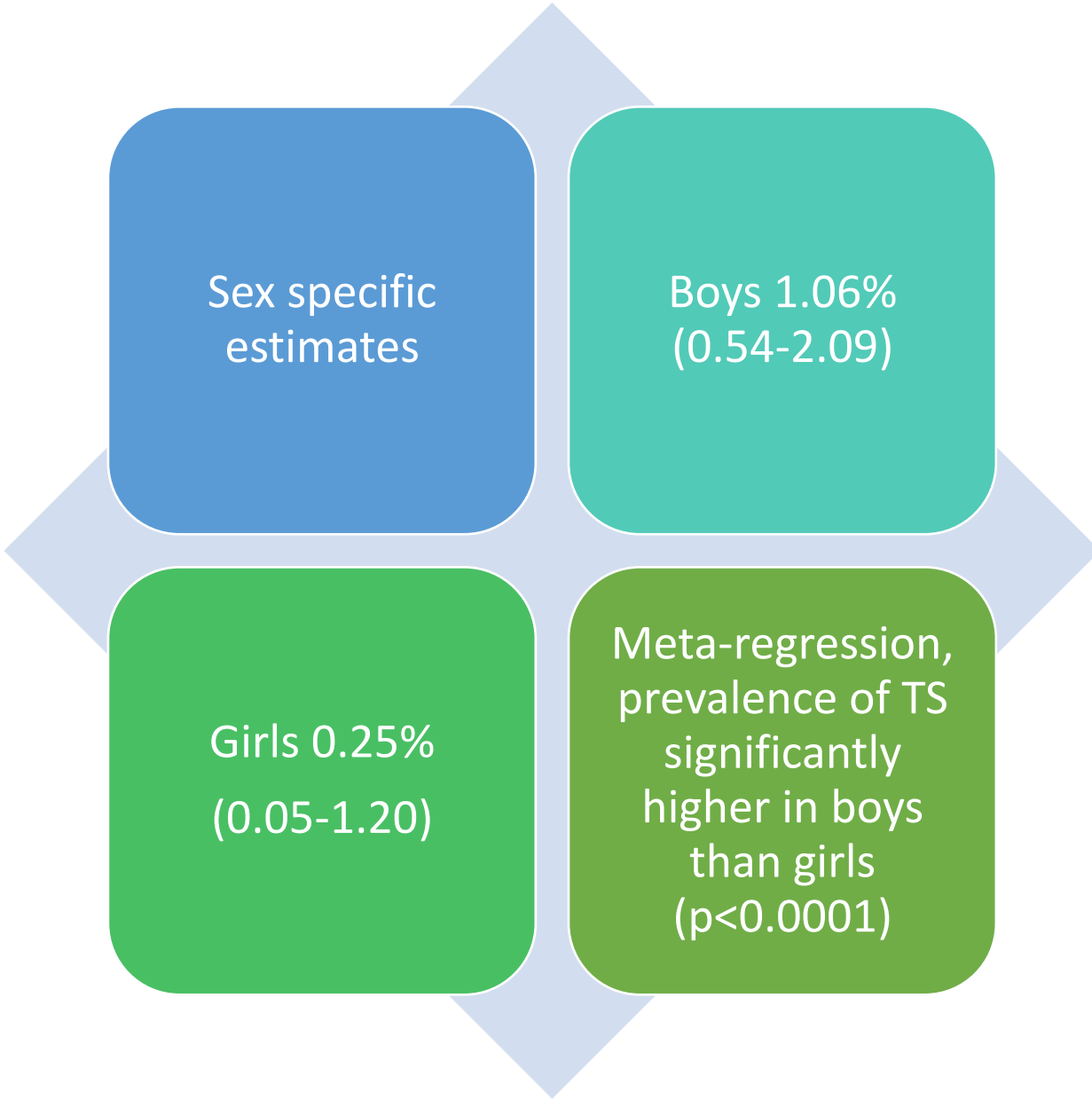
- The strongest predictors of high tic scores, and OCD or ADHD diagnoses in adulthood were the corresponding tic, OCD and ADHD severity scores in childhood
- Being female and childhood ADHD severity predicted future emotional disorders

Quality of Life

- The presence of comorbidity has a greater impact on quality of life than tic severity in children and adults with TS
- Psychosocial domains are more affected than physical domains
- Children
  - Presence and severity of ADHD has greatest impact
- Adults
  - Presence of anxiety and depression have greatest impact

Epidemiology of Tourette syndrome and comorbidities

Prevalence: Children, School Based Studies



Knight et al 2012 Pediatric Neurology

Prevalence of Diagnosed TS in Canada

- Data from Canadian Community Health Survey 2010/2011
- Population-based survey of Canadians age 12+
- Prevalence of diagnosed TS in adolescents was 3.33 per 1000
  - Male 6.03 per 1000
  - Female 0.48 per 1000
- Prevalence of diagnosed TS in adults was 0.66 per 1000
  - Male 0.89 per 1000
  - Female 0.44 per 1000

Sociodemographic characteristics of Canadians with TS

- Compared to the general population, Canadians with TS
  - Were less likely to attain post-secondary education
  - Had lower household income
  - Were less likely to be employed
  - If employed, were less likely to be employed full-time

Comorbidity

Original Investigation

Lifetime Prevalence, Age of Risk, and Genetic Relationships of Comorbid Psychiatric Disorders in Tourette Syndrome

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Objective: to characterise the lifetime prevalence, clinical associations, ages of highest risk of psychiatric comorbidity among individuals with TS

JAMA Psychiatry. 2015;72(4):325-333. doi:10.1001/jamapsychiatry.2014.2650
Published online February 11, 2015.

Comorbid Psychiatric Disorders in TS

- Phenotypic data collected for genetic studies from TS-affected individuals aged 6 years +
- Recruited from tic disorder specialty clinics in North America, Great Britain and the Netherlands
- 1374 participants with TS
- 85.7% met DSM IV-TR criteria for 1 or more comorbid disorder; 57.7% met criteria for 2 or more comorbid disorders

OCD and ADHD

OCD 50% (more common in females)

ADHD 54% (more common in males)

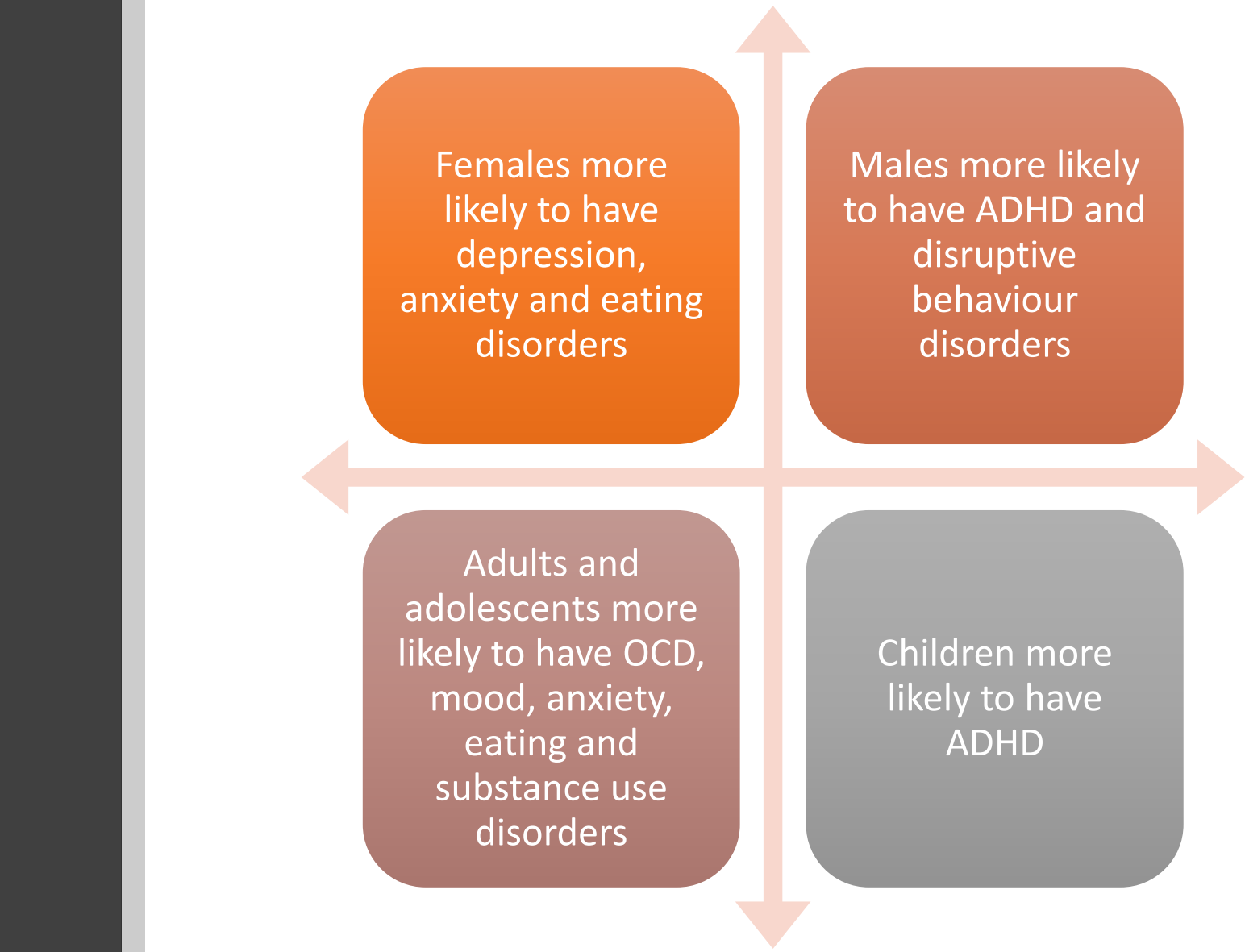
30% had TS+OCD+ADHD

28% had TS only

Other Psychiatric Comorbidities

- Mood Disorders (depression, dysthymia, bipolar I and II) 30%
- Anxiety Disorders (GAD, panic, agoraphobia, PTSD, SAD, social phobia, specific phobia) 36%
- Disruptive Behaviour Disorders (ODD, CD) 30%
- Eating Disorders (anorexia, bulimia) 2%
- Psychotic disorders 1%
- Substance use 6%

Comorbid Psychiatric Disorders in TS



Recommendations on the assessment and treatment of Tourette syndrome

American Academy of Neurology Clinical Practice Guidelines

Guidelines on the assessment and treatment of TS

Published in 2019

Evidence-based

Counseling recommendations: Natural History of TS

- Providing information to families about the natural history of a disorder can help inform treatment decisions
- Tics begin in childhood and demonstrate a waxing and waning course
- Peak tic severity usually occurs between the ages of 10 and 12 years, with many children experiencing an improvement in adolescence
- There is no evidence that treatment is more effective the earlier it is started.

Counseling recommendations: Natural history of TS

- 1a: Clinicians must inform patients and their caregivers about the natural history of tic disorders
- 1b: Clinicians must evaluate functional impairment related to tics from the perspective of the patient and, if applicable, the caregiver
- 1c: Clinicians should inform patients and their caregivers that watchful waiting is an acceptable treatment approach in individuals who do not experience functional impairment from their tics

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Recommendations: Natural history of TS

- 1d: Clinicians may prescribe the Comprehensive Behavioural Intervention for Tics (CBIT) as an initial treatment option relative to watchful waiting for people with tics who do not experience functional impairment, if they are motivated to attempt treatment
- 1e: Physicians prescribing medications for tics must periodically re-evaluate the need for ongoing medical treatment

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Psychoeducation: Teacher and Classroom

- Psychoeducation about TS with peers can result in more positive attitudes toward a person with TS
- Psychoeducation about TS with teachers can improve knowledge about the condition
- Improving peers' attitudes about and teachers' knowledge of TS may positively affect people with TS

Psychoeducation Recommendation

- 2. Clinicians should refer people with TS to resources for psychoeducation for teachers and peers, such as the Tourette Association of America
- In Alberta, you can refer them to us!
- www.touretteocdalbertanetwork.ca

Clinical Assessment of People with Tics

- Past medical history
- Medication use
- Developmental history
- Family history
- History of tics
- Neurological exam
- Assessment of tic severity

Secondary causes of tics

Neurodevelopmental disorders

Neurodegenerative disorders

- Huntington Disease
- NBIA
- Neuroacanthocytosis

Postinfectious and infectious

- Sydenham chorea
- PANS

Structural

- Vascular
- Post-traumatic

Toxic/Medication-induced

- Antipsychotics (tardive tics)
- Psychostimulants
- Antiepileptics – lamotrigine, phenytoin, carbamazepine

Clinical Assessment of People with Tics

- Screening for comorbidities essential
  - ADHD
  - OCD
  - Generalized Anxiety Disorder
  - Oppositional Defiant Disorder
  - Mood disorders

Assessment and treatment of ADHD in people with tics

3a: Clinicians should ensure an assessment for comorbid ADHD is performed in people with tics (B)

3b: Clinicians should evaluate the burden of ADHD symptoms in people with tics (B)

3c: In people with tics and functionally impairing ADHD, clinicians should ensure appropriate ADHD treatment is provided (B)

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Assessment and Treatment of OCD in people with tics

Cognitive behavioural therapy is considered first line treatment of OCD in individuals with tics disorders

4a: Clinicians should ensure an assessment for comorbid OCD is performed in people with tics (B)

4b: In people with tics and OCD, clinicians should ensure appropriate OCD treatment is provided (B)

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Other Psychiatric Comorbidities

There is an increased risk of dying by suicide and attempting suicide in people with TS which persists after controlling for psychiatric comorbidity

Persistence of tics beyond young adulthood, previous suicide attempts and comorbid personality disorders increase the risk of death by suicide

Fernandez de la Cruz et al 2016, Biological Psychiatry

Other Psychiatric Comorbidities

5a: Clinicians must ensure appropriate screening for anxiety, mood, and disruptive behavior disorders is performed in people with tics (A)

5b: Clinicians must inquire about suicidal thoughts and suicide attempts in people with TS and refer to appropriate resources if present (A)

Treatment expectations

- While medication and behavioural therapies can result in meaningful reduction in tics, these interventions rarely result in complete cessation of tics

When should we treat tics?

- No set symptom severity threshold to determine when we should treat
- An individual choice
- Recommend that the decision be based on disability
  - Physical
  - Social
  - Emotional
- The choice of which treatment to use should be a collaborative decision between patient and clinician

Behavioural Treatments

- CBIT is a manualized treatment program consisting of habit reversal training (HRT), relaxation training, and a functional intervention to address situations that sustain or worsen tics
- People with tics receiving CBIT are more likely than those receiving psychoeducation and supportive therapy to have reduced tic severity
- A randomized controlled trial comparing behavioural and pharmacological treatments in children with tics found that behavioural therapy is as effective as pharmacological therapy

Behavioural Treatments

- 7a For people with tics who have access to CBIT, clinicians should prescribe CBIT as an initial treatment option relative to other psychosocial/behavioral interventions
- 7b For people with tics who have access to CBIT, clinicians should offer CBIT as an initial treatment option relative to medication
- 7c Clinicians may prescribe CBIT delivered over teleconference or secure voice over Internet protocol delivery systems if face-to- face options are unavailable in a patient care center. If CBIT is unavailable, other behavioral interventions for tics may be acceptable, such as exposure and response prevention

Antipsychotic treatment for tics

Recommendations

Rationale

- Haloperidol, risperidone, aripiprazole, and tiapride are probably more likely than placebo to reduce tic severity
- Pimozide, ziprasidone, and metoclopramide are possibly more likely than placebo to reduce tic severity
- There is insufficient evidence to determine the relative efficacy of these drugs.
- Higher risk of drug induced movement disorders, weight gain, adverse metabolic side effects, prolactin increase and QT prolongation with both 1st and 2nd generation antipsychotics in both children and adults across psychiatric and neurological conditions

Recommendations

9a: Physicians may prescribe antipsychotic medications for the treatment of people with tics when the benefits of treatment outweigh the risk

9b: Physicians must counsel patients on the relative propensity of antipsychotic medications for extrapyramidal, hormonal and metabolic adverse effects to inform decision making on which antipsychotic should be prescribed

Recommendations

9c: Physicians prescribing antipsychotic medications for tics must prescribe the lowest effective dose of medication to decrease the risk of adverse effects

9d: Physicians prescribing antipsychotic medications for tics should monitor for drug-induced movement disorders and for metabolic and hormonal adverse effects, using evidence-based monitoring protocols

Recommendations

9e: Physicians prescribing antipsychotic medications for tics must perform ECGs and measure the QTc interval before and after starting pimozide or ziprasidone, or if antipsychotics are co-administered with other drugs that can prolong the QT interval

9f: When attempting to discontinue antipsychotic medications for tics, physicians should gradually taper medications over weeks to months to avoid withdrawal dyskinesias

Alpha agonists for the treatment of tics

Recommendations

Rationale

- In children with tics and ADHD, clonidine and guanfacine have demonstrated beneficial effects on both tics and ADHD symptoms
- Effect size for tics appears larger in children with comorbid tics and ADHD
- No evidence regarding relative efficacy of clonidine and guanfacine
- Systematic review on the use of alpha-2-adrenergics in children with ADHD demonstrated hypotension, bradycardia, and sedation with both agents, and QTc prolongation with guanfacine extended release
- Abupt withdrawal may cause rebound hypertension

Recommendations

8a: Physicians should counsel individuals with tics and comorbid ADHD that alpha-2-adrenergic agonists may provide therapeutic benefit for both conditions

8b: Physicians should prescribe alpha-2-adrenergic agonists for the treatment of people with tics when the benefits of treatment outweigh the risks

8c:Physicians must counsel patients regarding common side effects of alpha-2-adrenergic agonists, including sedation

Recommendations

- 8d: Physicians must monitor heart rate and blood pressure in all patients with tics treated with alpha-2-adrenergic agonists
- 8e: Physicians prescribing guanfacine extended release must monitor the QTc interval in patients with a history of cardiac conditions, patients taking other QTc-prolonging agents, or patients with a family history of long-QT syndrome
- 8f: Physicians discontinuing alpha-2-adrenergic agonists must gradually taper them to avoid rebound hypertension

Botulinum toxin injection for tics Recommendations

- Rationale
  - No evidence for toxins other than onabotulinum toxin A
  - Adverse effects include weakness and hypophonia when injected into laryngeal muscles
  - Effect lasts 12-16 weeks

Recommendations

10a: Physicians may prescribe botulinum toxin injections for the treatment of older adolescents and adults with localized and bothersome simple motor tics when the benefits of treatment outweigh the risks

10b: Physicians may prescribe botulinum toxin injections for the treatment of older adolescents and adults with severely disabling and aggressive vocal tics when the benefits of treatment outweigh the risks

Recommendations

- 10c: Physicians must counsel individuals with tics that botulinum toxin injections may cause weakness and hypophonia, and that all effects are temporary

Topiramate Recommendations

- Rationale
  - In patients who are not obtaining a satisfactory response or experience adverse effects from other medical treatments, topiramate may be a useful alternative
  - Adverse effects include cognitive and language problems, somnolence, weight loss, and it may increase the risk of renal stones

Recommendations

11a: Physicians should prescribe topiramate for the treatment of tics when the benefits of treatment outweigh the risks

11b: Physicians must counsel patients regarding common adverse effects of topiramate, including cognitive and language problems, weight loss, and an increased risk of renal stones

Cannabis-based medication for the treatment of tics

- Rationale
  - Many patients with TS use cannabis as a self-medication for tics
  - Current RCTs limited to THC
  - Cannabis is associated with an increased risk of short-term dizziness, dry mouth and fatigue
  - No evidence that controlled treatment will induce addiction

Cannabis-based medication for the treatment of tics

- Rationale
  - Should be avoided in children and adolescents due to the association between cannabis exposure and harmful cognitive and affective outcomes in adulthood
  - Not to be used in women who are pregnant or breastfeeding, or in patients with psychosis
  - Prescription and access varies by region; practitioners must abide by regional legislation

Cannabis Recommendations

- 12a: Due to the risk associated with cannabis use and widespread self-medication with cannabis for tics, physicians must offer to direct patients to appropriate medical supervision when cannabis is used as self-medication for tics
- 12b: Physicians may consider treatment with cannabis-based medication in otherwise treatment resistant adult patients with TS suffering from clinically relevant tics
- 12c: Physicians may consider treatment with cannabis-based medication in adult patients with TS who already use cannabis efficiently as a self-medication in order to better control and improve quality of treatment

Cannabis Recommendations

- 12d: Physicians prescribing cannabis-based medication must prescribe the lowest effective dose to decrease the risk of adverse effects
- 12e: Physicians prescribing cannabis-based medication must inform patients that medication may impair driving ability
- 12f: Physicians prescribing cannabis-based medication to patients with TS must periodically re-evaluate the need for on-going treatment

Tourette Syndrome Clinic at Alberta Children's Hospital

- The only dedicated clinic for children affected by TS in Alberta
- Adults with TS seen at Movement Disorders Clinic at Foothills Medical Centre
- 3 neurologists
  - Davide Martino, Justyna Sarna and myself
- 1 nurse
  - Tracy Hammer
- Limited psychology, physical therapy and occupational therapy support from ACH Neuroscience Clinics

Tourette OCD Alberta Network

- Funded by MNCY SCN from 2019 to 2022, then will transition to non-profit society
- Provide care navigation for people affected by TS and OCD province wide
- Provide education to mental health therapists across Alberta in behavioural therapies for TS and OCD
- Provide education, support and resources for people affected by TS and OCD
- Work with educators to facilitate school success
- touretteocdalbertanetwork.ca

Summary

Tourette syndrome is a common childhood onset neurodevelopmental disorder

Many children will have improvement in tic severity in late adolescence

All individuals with Tourette syndrome should be screened for comorbid psychiatric disorders

Behavioural therapies are recommended first-line for tics which cause functional impairment

There are several medications that can help decrease tic severity