Tourette Syndrome and Autism-what are the implications?

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KING'S HEALTH PARTNERS

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Tourette syndrome

- Multiple motor and phonic tics

onset < 18 years

frequency +++ per day > 1 year

no remission > 3 months

+/-Significant impairment

- M:F 3: 1
- Family history +ve

ESSTS- European Guidelines

- Assessment and investigation
- Pharmacology
- Psychology-Behavioural/psychosocial and Neurocognitive
- Deep Brain Stimulation
- Role of streptococcal disease and PANDAS

Eur Child Adolesc Psychiatry (2011) 20:155–171


Psycho-Social
Education
Psychology
Neuro-
Psychiatry
Movements
Development
Genetics

- Tics
- Myoclonus
- Tremor
- Dystonia
- Hyperekplexia
- Chorea
- Athetosis
- Stereotypy
- ?toe walking

- focal and segmental dystonias (blepharospasm, retro-, latero- en torticollis, Meige's syndrome and oculogyric crises).
- Myoclonic dystonias, e.g. DYT 11
- "Culture Bound" Startle disorders -Latah, Myriachit, Imu, Goosy, and Ragin Cajuns)
- Paroxysmal kinesiogenic choreoathetosis.
- Multifocal and focal myoclonias.
- Tardive oro-bucco-lingual dyskinesias
- Drug induced movement disorders
- Epilepsy-e.g.rolandic/occipital./MAS
- Tremors of thalamic glioma
- Post-encephalitic movement disorders
- Restless legs syndrome
- Hemifacial spasms / blepharospasm
- Choreas e.g. Huntington's disease, Chorea Acanthocytosis
- Sydenhams Chorea / PANDAS
- Tremor of Wilson's Disease
- Psychogenic tics.(rare)

Co-Morbidities

- ADHD 60%
- OCD/OCB 30%
- Anger control problems 28%
- Sleep disorder 18%
- Learning problems 22%
- Mood disorder 17%
- Anxiety disorder 25%
- Self injurious behavior 15%-60%
- Autism spectrum disorder 30%

ocb

- Checking
- Counting
- Washing
- Smelling
- Rearranging
- Touching
- Hoarding
- symmetry

Neurobiology

- *Conventional imaging* normal-?caudate size
- *Circuitry postulated* is fronto-cortico-striatal
- *Transmitters implicated*—dopamine excess release or hypersensitivity of postsynaptic d2 receptors
- Neuronal nicotinic acetylcholine receptors
- P.m. low serotonin in brainstem
- Cholinergic interneurones

Genetic factors

- Mz twin 87% conc DZ 20%
- Linkage to 4,5,8,11,17
- Bilineal transmission

- SLITRK1 Candidate gene on chromosome 13q31.1 identified due to proximity to inversion. (dentritic growth in cortex) (science 2005)

- L-Histidine Decarboxylase Complex

Anti neural antibodies and CNS autoimmunity

Antibodies against intracellular antigens

- Onconeural antibodies; Hu, Yo, Ri, CV2, amphipysin, Ma2.
- GAD

Antibodies against neuronal surface antigens

- VGKC, AMPA, GABA, NMDA, Glycine receptors

Graus at al., 2010 J Neurol. 257(4); 509-17.

VGKC antibodies in children

- 12 paediatric patients whose serum IgG immunoprecipitated VGKC complex.
- VGKC titres 50-1420
- Diverse clinical spectrum; 3 global developmental regression, 6 movement disorder, 4 dysarthria, 3 seizures, 1 neoplasia.
- Benefits from early immunotherapy, with tendency to relapse.
- Diagnosis delayed

Dhamija at al 2011. Pediatric Neurology 44; 275-281

Some Cases

Tic vs. Stereotypy

Tic	Stereotypy
Onset 5-7 years	Onset typically <3 years
Migration of tics over months or years	Typically a single stereotypy which remains relatively stable
Premonitory urge which is relieved by performing tic	Lack of premonitory urge, but associated with engrossment in a task
Common location – eyes, face, head, shoulders	Involve hand, arm or entire body frequently

- 11 YR OLD BOY
- SEVERAL YEARS OF TICS GOING UNDIAGNOSED.
- CONCERNS OVER SOCIALISING AND CONFIDENCE.

Trend Report

Patient: 0072-7527 (legocaptainrexfan)

Report Parameters

Parameter Name	Value
Scale Completions	Display all
Display Items	Difficulty with daily activities, Felt anxious, Felt needed more help, Lack of self-confidence, Tics caused pain or injuries

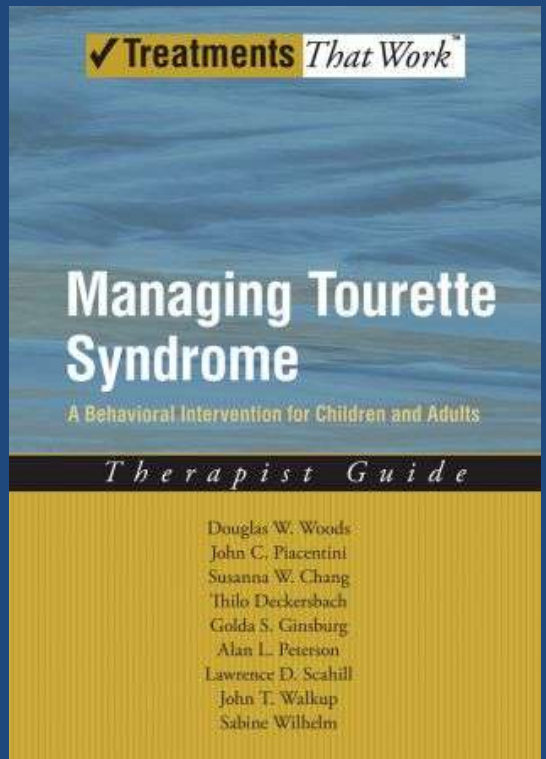
Psychological treatments

- Mass practice
- Habit reversal
- Self-monitoring
- Contingency management
- Relaxation training
- Exposure with response prevention (ER/P)
- neurofeedback

CBITS trial and manual

(Comprehensive Behavioural Intervention for Tics)

- Includes multiple strategies
- Subject of large RCT cf education only
- Outcome in acute phase ~ 50% reduction in impairment score
- ES=0.68 (cf meds max 1.0)
- Published treatment manual
  - Woods et al OUP



Other Treatments

- Clonidine
- risperidone
- aripiprazole
- Nicotine; mecamylamine
- Benzodiazepines, baclofen
- Tetrabenazine
- Topiramate,
- Cannabinoids
- Botulinum toxin
- pimozide
- Immunomodulation; penicillin prophylaxis
- Transcranial magnetic stimulation
- Neurosurgery; deep brain stimulation

Naloxone + other opioids

Olanzapine

Antiandrogen

SSRIs

Na Valproate levetiracetam

Nifedipine

Atomoxetine

Haloperidol

TOURETTE	ASD
Problems in SOCIABILITY –not always	Problems with sociability-core deficit
Language delay/disorder-rare	Core deficit
OCD/OCB	Rigidity, routines, restricted interest
Specific learning difficulty-often mild	Learning difficulties-uneven profiles
ADHD often	Some hyperfocus
Infrequent abnormal EEG	0.3 Abnormal EEG
Frequent Tics	Some Tics , more stereotypies

Stereotypies

Repetitive, seemingly driven, non-functional behaviours present for >4 weeks that interferes with normal activities or result in self injury

DSM–IV

- Not better accounted for by a compulsion or a tic.
- Not due to physiological effects of a substance or a general medical condition.

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