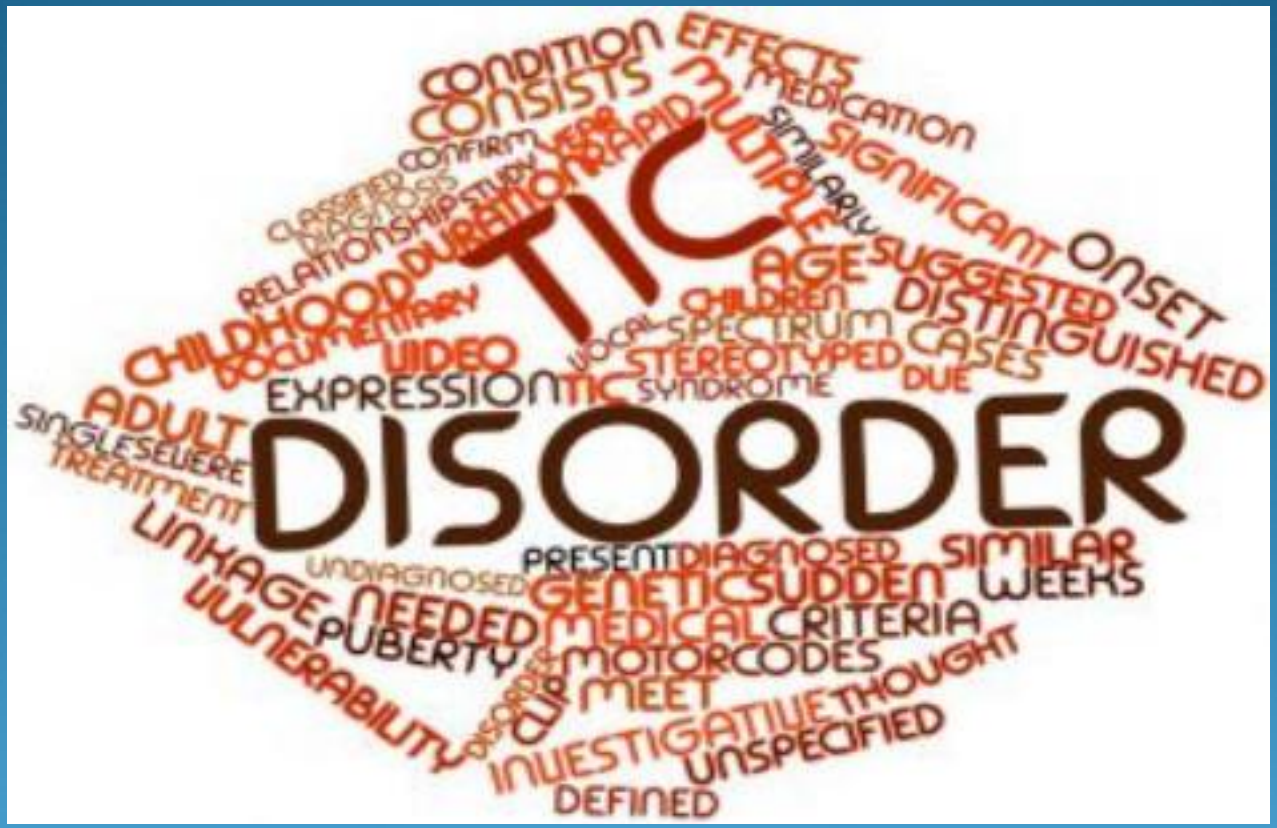
TIC DISORDERS



INTRODUCTION

- Perhaps the best known of the tic disorders is **Tourette's disorder** (TD).

- Tic disorders have emerged as complex, heterogeneous, neuropsychiatric movement disorders of **childhood onset** that are the manifestation of an intricate interplay of genes and environment.

- The understanding of the etiology, genetics, neuroanatomy, and epidemiology of tic disorders has been significantly enhanced.

HISTORY

In 1825, the French physician **Jean-Marc Gaspard Itard** provided the first clinical description of Tourette's disorder in a French noblewoman, the Marquise de Dampierre, who displayed body tics, barking sounds, and uncontrollable utterances of obscenities.

HISTORY

In 1885, **Georges Gilles de la Tourette**, a French neurologist, described eight patients, in addition to the patient originally reported by Itard, with childhood-onset tics and coexisting emotional symptoms and behavioral problems.

Gilles de la Tourette coined the term coprolalia for vocal tics that manifested as uncontrollable utterances of obscenities.

HISTORY

Psychoanalytic views of Tourette's disorder, introduced in the 1920s and 1930s, interpreted tics as expressions of repressed childhood sexual conflict.

In the 1940s, **Margaret Mahler** attributed tic symptoms to infantilization, parental overindulgence, and overprotection, as well as sexual conflict.

HISTORY

Serge Lebovici, the leading French child psychoanalyst during the 1950s and 1960s, asserted that neurotic mothers were the cause of tics in their children.

COMPARATIVE NOSOLOGY

- It depends on the presence and classification of their primary symptom—tics.

- The DSM-IV-TR and the ICD-10 criteria, are based primarily, on **the type of tics present** (motor, vocal, or both) and **the duration of symptoms** (greater or less than 1 year).

COMPARATIVE NOSOLOGY

- Over the years, individual criteria have changed slightly as advances in understanding of the prevalence and course of tic disorders has emerged.

- DSM-IV-TR, has eliminated the criterion that tics cause distress or impairment.

EPIDEMIOLOGY

- Transient tic disorder-up to 20 percent of school-age children
- Chronic tic disorders is between 2 and 4 percent.
- Prevalence estimates for Tourette's disorder also vary.

EPIDEMIOLOGY

- Boys are more commonly affected.
- The reported male-female ratio varies from 1:1 to 10:1 across epidemiological studies (Scahill *et al.* 2013)
- Tourette's disorder is manifest by 11 years of age, typically beginning between ages 3 and 8 years. The average age of onset is 7 years of age.

ETIOLOGY & PATHOPHYSIOLOGY

- The etiology and pathophysiology of Tourette's disorder and other tic disorders are unknown.

- Factors that are considered to be involved in it are-

A. Genetic factors
B. Immunological
C.Psychosocial stress
D. Neurochemical and neuroanatomical

ETIOLOGY & PATHOPHYSIOLOGY

A. GENETIC FACTORS-

- Twin studies- concordance rate for monozygotic twins are greater than dizygotic twins(Prince *et al.* 1985)

- Family studies- first degree relatives are at high risk(Pauls *et al.* 1991).

ETIOLOGY & PATHOPHYSIOLOGY

- Chromosome studies-Regions near the centromere of chromosome 2, as well as on 6p, 8q, 11q, 14q, 20q, 21q, and X, have been implicated.

- Cytogenetic studies- rare sequence variant in SLITRK1 on chromosome 13q31 (Abelson *et al.*2013).

- Androgen exposure is speculated to play a role in the development of tics, given the higher prevalence of Tourette's disorder in males.

ETIOLOGY & PATHOPHYSIOLOGY

B. IMMUNOLOGICAL-

- A post infectious mechanism for the development of Tourette's disorder and OCD has been postulated since the late 1800s.

- An autoimmune process that is secondary to the paediatric autoimmune neuropsychiatric disorders associated with streptococcal infection (PANDAS), is considered to be the potential mechanism for TS.

ETIOLOGY & PATHOPHYSIOLOGY

C.PSYCHOSOCIAL STRESS-

- Tic disorders have long been identified as “stress sensitive” conditions.

- Patients with TS experience more stress than healthy controls(Findley *et al*.2003) and that antecedent stress may play a role in subsequent tic exacerbation

  (Lin *et al*., 2007, 2010)

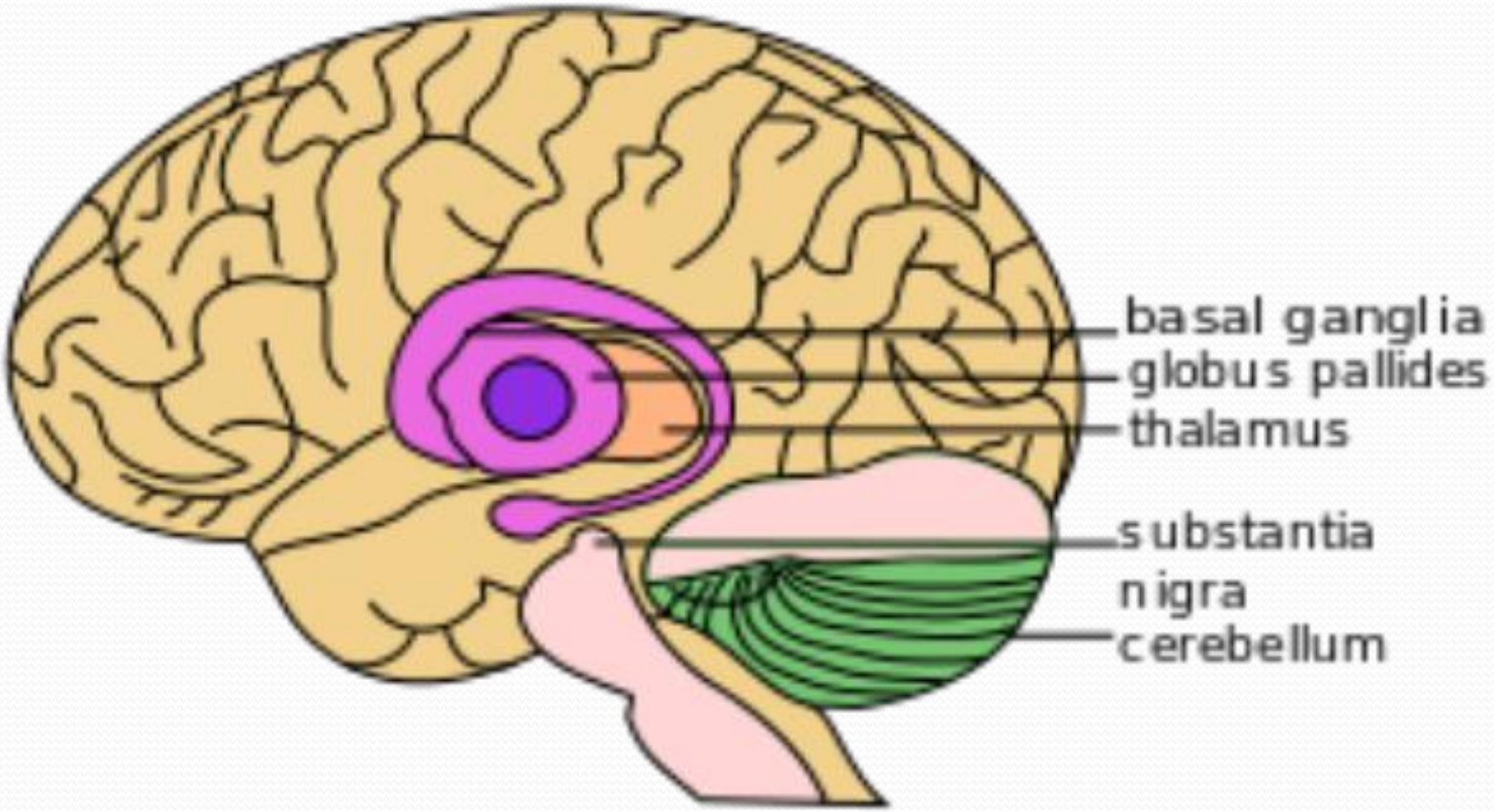
ETIOLOGY & PATHOPHYSIOLOGY

D. NEUROCHEMICAL AND NEUROANATOMICAL-

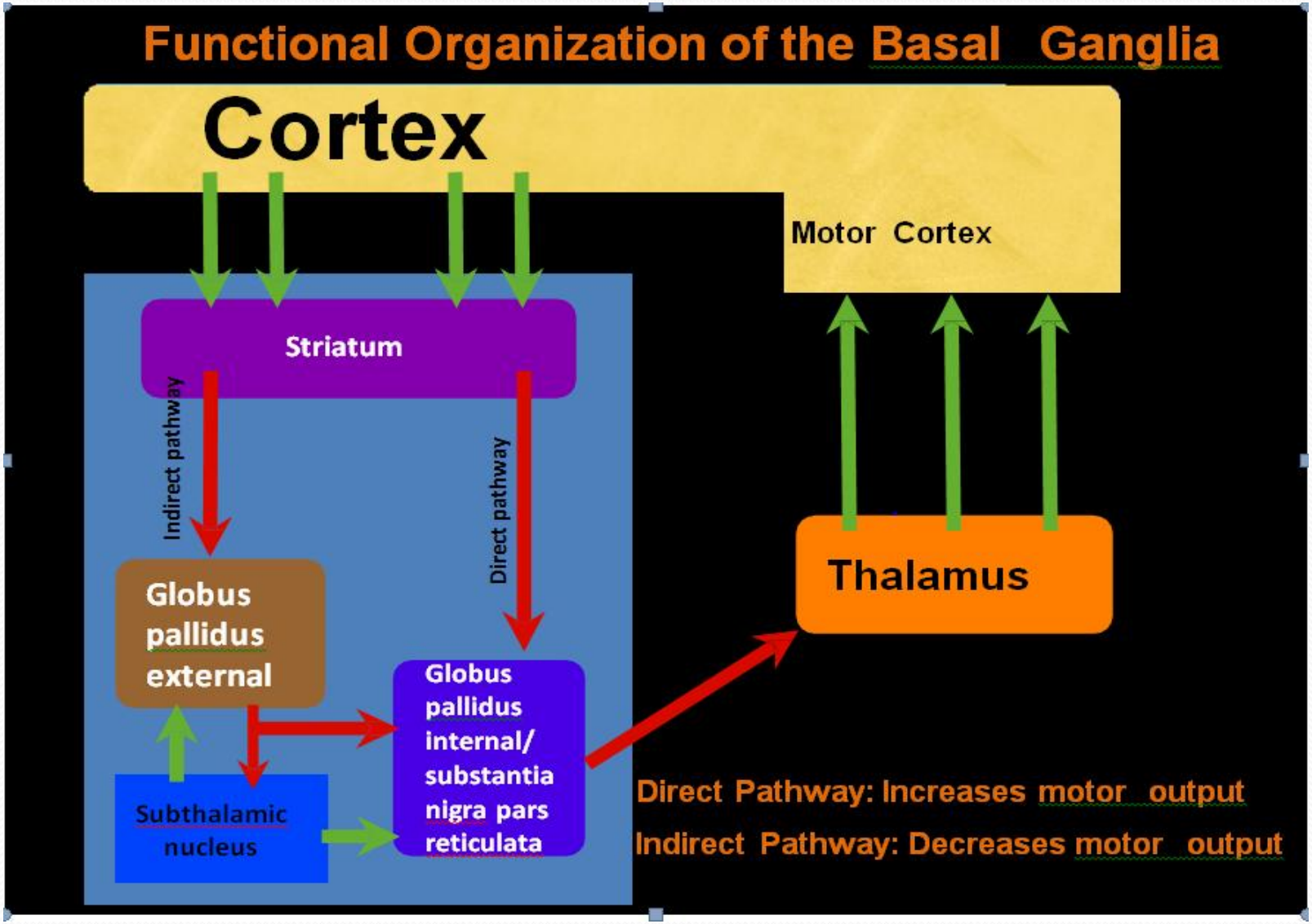
- Glutamatergic, serotonergic, cholinergic, noradrenergic, and opioid systems may also be implicated, given that they are present in CSTC circuits and interact with the dopaminergic system.

- Choline and n-acetylaspartate reduced in left putamen and frontal cortex.

Basal Ganglia and Related Structures of the Brain



CORTICO-STRIATO-THALAMO-CORTICAL-CIRCUIT



ETIOLOGY & PATHOPHYSIOLOGY

- MRI studies have indicated- volume or asymmetry abnormalities in caudate or lenticular nuclei (putamen and globus pallidus).

- Larger parieto-occipital region volumes and smaller inferior occipital volumes have also been observed.

ETIOLOGY & PATHOPHYSIOLOGY

- Increased gray-matter volumes bilaterally in the ventral putamen and decreases in gray matter in the left hippocampal gyrus have also been shown.

- Taken together, structural and functional neuroanatomical findings support the understanding that Tourette's disorder results from abnormal activity in the basal ganglia.

CLINICAL FEATURES

- Tics are stereotyped, rapid, recurring motor movements (motor tics) or vocalizations (phonic or vocal tics) that are nonrhythmic, involuntary or semivoluntary, and sudden in onset.

- Tics typically involve one muscle or a group of muscles

- May be characterized by their anatomical location, number, frequency, duration, and complexity

CLINICAL FEATURES

- They can be classified as-

-**Simple** (involving one muscle, muscle group, or sound)

- **Complex** (slower, more purposeful movements involving multiple muscle groups or multiple sounds).

CLINICAL FEATURES

- **Simple motor tics** cause-

-a brief, jerking movement (clonic tic);

-a briefly sustained abnormal posture (dystonic tic); or an isometric contraction (tonic tic).

CLINICAL FEATURES

- **Examples** of simple motor and vocal tics are-
  - -eye blinking
  - -shoulder shrugging
  - -sustained eye closure (blepharospasm)
  - -ocular deviations
  - -teeth grinding (bruxism)
  - -mouth opening
  - -throat clearing, coughing, and sniffing

CLINICAL FEATURES

- **Complex motor and vocal tics** may include-

  -touching objects

  -Jumping

  -Rotating

  -imitating others' gestures (echopraxia)

  -repeating syllables, words, or one's phrases (palilalia)

  -repeating others' words or phrases (echolalia).

CLINICAL FEATURES

- The most common initial presentation of a tic disorder- Simple motor tics
- The most common initial tic- eye blinking
- Tics are experienced as irresistible but can be suppressed for varying periods of time.

CLINICAL FEATURES

- Several tics separated by variable periods of no tic activity.
- Change in frequency, anatomical location, number, complexity, and severity over minutes, hours, and days, as well as over longer periods of time.
- May persist during sleep.

CLINICAL FEATURES

- Many individuals with Tourette's disorder characterize their tics as a voluntary response to an uncomfortable feeling that precedes them.

- Many individuals report a "**just right phenomenon**" that to relieve the uncomfortable urge they must repeat a particular movement until "it feels good" or "it feels just right."

CLINICAL FEATURES

-Both multiple motor and one or more vocal tics are there.

-Begins with a simple motor tic on the face (e.g., blinking) Eventually, vocalizations (e.g. throat clearing, barking etc) ensue and are typically "explosive."

CLINICAL FEATURES

-**Copopraxia** = a sudden, tic-like vulgar, sexual, or obscene gesture.

- **Coprolalia** = the sudden, inappropriate expression of a socially unacceptable word or phrase that may include obscenities

DIAGNOSIS

- It is diagnosed primarily on the basis of history and tics that may be observed during the examination, reliable sources of information are essential.

- Both DSM-5 and ICD-10 contains 3 major categories-
- 1.Tourette's Disorder
- 2.Persistent (Chronic) Motor or Vocal Tic Disorder
- 3.Provisional Tic Disorder

DIAGNOSIS

Tourette's Disorder 307.23 (F95.2)

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

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

DIAGNOSIS

- Onset is before age 18 years.

- The disturbance is not attributable to the physiological effects of a substance (e.g., cocaine) or another medical condition (e.g., Huntington's disease, postviral encephalitis).

DIAGNOSIS

Persistent (Chronic) Motor or Vocal Tic Disorder 307.22 (F95.1)

- Single or multiple motor or vocal tics have been present during the illness, but not both motor and vocal.
- The tics may wax and wane in frequency but have persisted for more than 1 year since first tic onset.
- Onset is before age 18 years.

DIAGNOSIS

- The disturbance is not attributable to the physiological effects of a substance (e.g., cocaine) or another medical condition (e.g., Huntington's disease, postviral encephalitis).

- Criteria have never been met for Tourette's disorder.

- *Specify* if:

  With motor tics only

  With vocal tics only

DIAGNOSIS

Provisional Tic Disorder 307.21 (F95.0)

- Single or multiple motor and/or vocal tics.
- The tics have been present for less than 1 year since first tic onset.
- Onset is before age 18 years.
- The disturbance is not attributable to the physiological effects of a substance (e.g., cocaine) or another medical condition (e.g., Huntington's disease, postviral encephalitis).
- Criteria have never been met for Tourette's disorder or persistent (chronic) motor or vocal tic disorder.

DIAGNOSIS

- Other Specified Tic Disorder **307.20 (F95.8)**
- Unspecified Tic Disorder **307.20 (F95.9)**

RATING SCALES

- Yale Global Tic Severity Scale (YGTSS)
- Abnormal Involuntary Movement Scale (AIMS)
- Tic Symptom Self-Report

DIFFERENTIAL DIAGNOSIS

1. CHOREA-

- Spontaneous movements, irregularly timed, non-repetitive, randomly distributed and abrupt in character.

- Motor impersistence (ie, they cannot maintain a sustained posture) & involves both proximal and distal muscles.

DIFFERENTIAL DIAGNOSIS

2.ATHETOSIS-

- Slow, irregular, writhing movements, usually in the hands and fingers but often involving the neck.

- It is a slow form of chorea.

DIFFERENTIAL DIAGNOSIS

3.BALLISMUS-

- Intermittent, coarse, large-amplitude, jerking, shaking, flinging movements.
- Typically unilateral

DIFFERENTIAL DIAGNOSIS

4.COMPULSION-

- Repetitive behavior undertaken to prevent or reduce anxiety or distress.
- It is important to determine whether the behavior is being pursued as a strategy to relieve anxiety or as a preventive measure rather than simply to satisfy an urge.

DIFFERENTIAL DIAGNOSIS

5.DYSKINESIA-

- Choreiform or dystonic movement that is involuntary, stereotyped, and not suppressible .

DIFFERENTIAL DIAGNOSIS

6.DYSTONIA-

- Sustained, tonic contraction that progresses to abnormal postures
- Repetitive twisting and sustained involuntary movements that may be slow or rapid.
- Movements are not preceded by any premonitory sensation and they are unable to be temporarily suppressed.

DIFFERENTIAL DIAGNOSIS

7.MYOCLONUS-

- Sudden, brief, clonic, shock-like jerks, usually involving the limbs.
- Myoclonus is not suppressible.
- The condition may be physiologic (such as sleep myoclonus) or pathologic (such as seen with myoclonic epilepsies).

DIFFERENTIAL DIAGNOSIS

- It is characterized by sudden unidirectional movements that are often non rhythmic .

- It can be differentiated from tics by its rapidity, lack of suppressibility and absence of premonitory urges.

DIFFERENTIAL DIAGNOSIS

8.RESTLESS LEG SYNDROME-

- The most prominent symptom of RLS is the overwhelming urge to move your legs, especially when patient sitting still or lying in bed.

- Feel unusual sensations like a tingling, crawling, or pulling sensation in the legs.

DIFFERENTIAL DIAGNOSIS

- Symptoms get worse at night and are mild or absent in the early part of the day.

- Symptoms ease up when the patient move.

DIFFERENTIAL DIAGNOSIS

9.SPASM-

- Stereotypic, prolonged movement involving the same group of muscles.

DIFFERENTIAL DIAGNOSIS

10.AKATHESIA-

- Motor restlessness (unpleasant need to move), usually in the lower extremities.
- Affects trunk and leg muscles

DIFFERENTIAL DIAGNOSIS

11.TREMOR-

- Unintentional, somewhat rhythmic, muscle movement involving oscillations of one or more parts of the body

DIFFERENTIAL DIAGNOSIS

12.STEREOTYPY-

- Repetitive, usually meaningless, gestures, habits, or automatisms.
- Movements are repetitive, stereotyped, and purposeless, elicited by excitement, anxiety, and occasionally boredom. The characteristics of the movements change little in quality over time.

DIFFERENTIAL DIAGNOSIS

- Stereotypies are common in children with autism or intellectual deficiency, although may be present in children with no other conditions.
- Typically do not start in the face or neck.
- Usually starts at a younger age, such as in the first 1 to 2 years of life.

COMORBIDITY

- Symptoms associated with ADHD & OCD have received the most attention.
- 30-50% children with TS diagnosed with comorbid ADHD(Khalifa & Von Knorring,2006)
- More than 40% individuals with TS experience recurrent OC symptoms (Leckman *et al.* 1994, 1997)

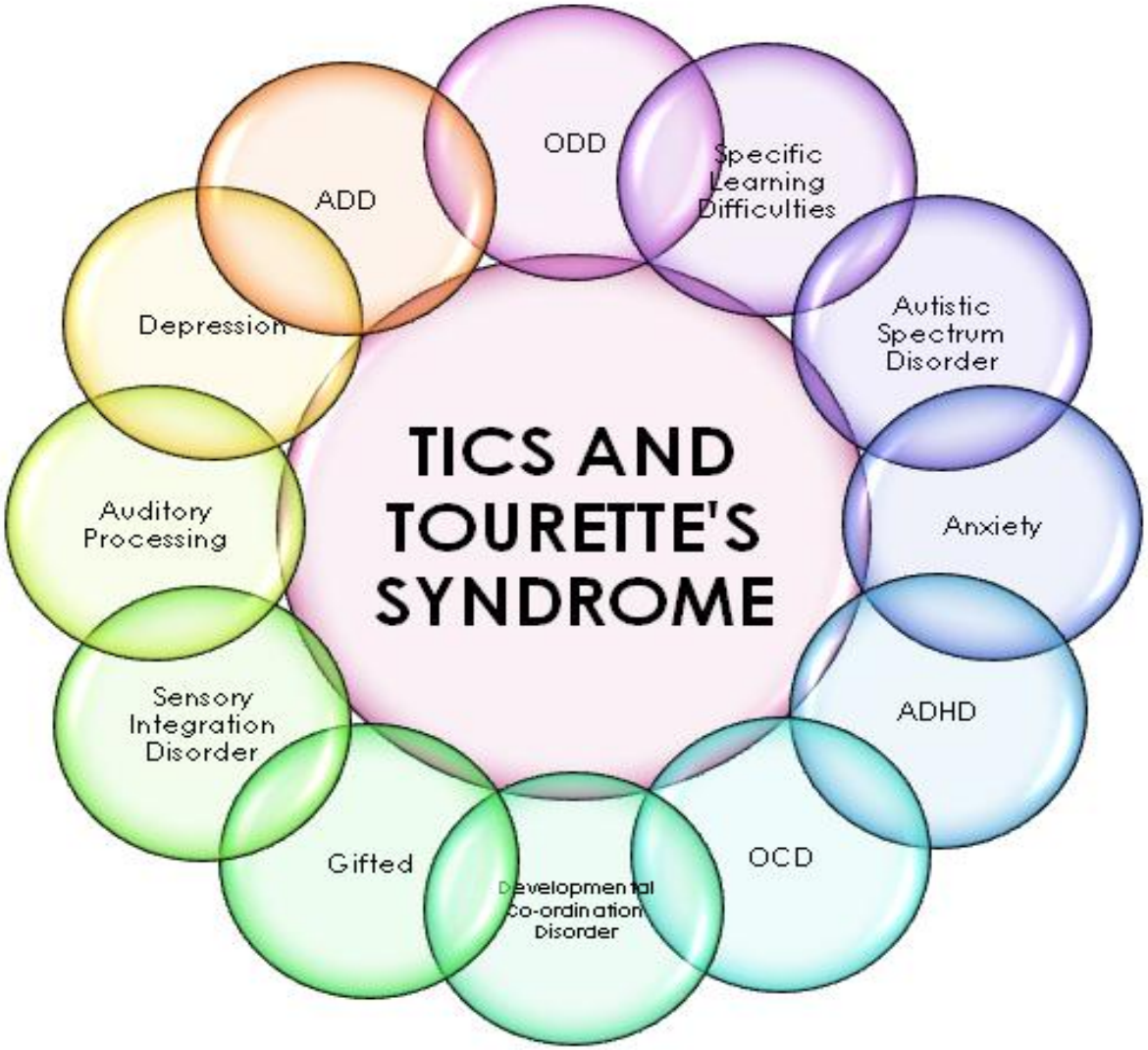
COMORBIDITY

- In the several cohorts study conducted-

-From 20 to 40 % of Tourette's disorder patients have been reported to meet full criteria for OCD

-Up to 90 % have been reported to have subthreshold symptoms such as repetitive counting, touching, or symmetry needs.

COMORBIDITY



COURSE AND PROGNOSIS

- The typical course of chronic motor tic disorder and Tourette's disorder is characterized by the onset of simple motor tics of the eyes, face, head, or neck at about age 6 or 7 years.

- Followed by a rostral-caudal progression of motor tics over several years to involve the shoulders, trunk, and extremities.

COURSE AND PROGNOSIS

- Vocal tics typically start at age 8 to 9 years; more-complex tics often begin later.
- Tics range from mild to severe and tend to stabilize over time.
- Frequency ranges from nonstop bursts(>100 tics per min) to rare events that occur only a few times a week(Peterson & Leckman,1998)

TREATMENT

- Individuals with mild and even some moderate tics may not need treatment at all, if the tics are not causing significant distress or impairment.

- Simple, active monitoring may be all that is necessary for some individuals with mild tics.

TREATMENT

- **Indications** for treatment of tics include-

  -interference with social interactions,

  -physical pain or discomfort, and

  -impairment in any aspect of educational or occupational functioning.

TREATMENT

- Medications are usually considered only when symptoms interfere with academic or job performance, peer relationships, social interactions, or activities of daily living.

- **The goal of treatment** is not to eliminate all tics, but to relieve tic-related discomfort or embarrassment and to achieve a degree of control of tics that allows the patient to function as normally as possible.

TREATMENT

GENERAL GUIDELINES-

- Pharmacotherapy is the cornerstone of effective treatment for tic disorders.
- Treatment goals should be to reduce symptoms or achieve symptom control, support adaptive functioning and strengths, and enhance developmental progress.
- Self-esteem at all times should be supported

TREATMENT

- Containment is another cornerstone of treatment.
- Use of a “tic room” or a “time out” area provides an opportunity to contain problematic tics or compulsions and to “de-stimulate.”
- Because emotional conflicts and stress frequently increase symptom intensity and frequency, time-limited withdrawal from stressful situations can be beneficial.

Medication	Typical Range (mg)	Starting Dose (mg)	Maximum Dose (mg)	Common Side Effects1.
1.NEUROLEPTICS-				
Haloperidol	.25–5.0	.25–.5	5.0	Sedation, weight gain, dysphoria, extrapyramidal effects, dry mouth, blurred vision, constipation, urinary retention
Pimozide	1.0–10	.5–1.0	15–20	Sedation, weight gain, dysphoria, extrapyramidal effects, dry mouth, blurred vision, constipation, urinary retention, ECG changes (QTc prolongation)
Risperidone	1.0–3.0	.25–.5	4–6	Weight gain, sedation, lipid and glucose metabolism abnormalities, extrapyramidal effects
Ziprasidone	20–80	5–10	120–160	Weight gain, sedation, lipid and glucose metabolism abnormalities, extrapyramidal effects, ECG changes (QTc prolongation)
Aripiprazole	2.5–15	1.25–2.5	20	Akathisia, sedation, agitation, weight gain, lipid and glucose metabolism abnormalities, extrapyramidal effects

Medication	Typical Range (mg)	Starting Dose (mg)	Maximum Dose[a] (MG)	Common Side Effects
2.Partial α2-adrenergic agonists				
Clonidine	.05–.45	.025–.05	.45	Sedation, headaches, insomnia, stomachaches, hypotension, dry mouth, midsleep awakening, irritability
Guanfacine	1.0–3.0	.25–.5	3–4	Sedation, headaches, hypotension, midsleep awakening, constipation
3.TCAs-Desipramine, Clomipramine				
4.SSRIs- Fluoxetine, Sertraline, Paroxetine, Fluvoxamine, Citalopram, Escitalopram				

PHARMACOTHERAPY

NEUROLEPTIC AGENTS-

- Since the late 1960s when haloperidol was first introduced as a treatment for Tourette's disorder patients.

- Haloperidol and pimozide have been the only FDA approved agents for the treatment of Tourette's disorder.

PHARMACOTHERAPY

- The newer, atypical neuroleptics, or second-generation antipsychotic agents, are recommended as **first-line treatment** when a neuroleptic is indicated for moderate to severe tics.

- These agents, which block both D_2 dopamine and 5-HT_2 serotonin receptors, have the potential advantage of fewer extrapyramidal side effects.

PHARMACOTHERAPY

α-Adrenergic Agonists-

- These are recommended as first-line treatment for most patients with mild to moderate Tourette's disorder.
- Clonidine activates presynaptic autoreceptors in the locus coeruleus to reduce norepinephrine release and turnover in the cerebral cortex.
- In addition, clonidine also reduces the disinhibition, impulsivity, inattention, and hyperactivity often present in young Tourette's disorder patients.

PHARMACOTHERAPY

- Guanfacine (Tenex), an alternative α2-adrenergic agonist, may also be efficacious for hyperactivity, impulsivity, and tics.

- Guanfacine activates postsynaptic prefrontal α-adrenergic cortical receptors

OTHER AGENTS

STIMULANTS-

- Stimulants are first-line agents for the treatment of ADHD.
- The current recommendation is to provide stimulant treatment for ADHD symptoms with comorbid tic disorder, monitoring for the exacerbation of tics.
- Side effects of stimulant use include insomnia, appetite suppression, and weight loss.

OTHER AGENTS

- Immune therapies such as IVIG, plasmapheresis
- Corticosteroids
- Opioid antagonists (e.g., naltrexone)
- Botulinum toxin

SURGICAL TREATMENT

- Stereotaxic neurosurgical techniques
- Bilateral capsulotomy
- Deep brain stimulation (DBS)
- Transcranial magnetic stimulation

BEHAVIOURAL THERAPY

- Relaxation techniques, such as deep breathing, guided imagery, and use of relaxation tapes.
- Habit-reversal training (HRT)
- Parent management training

BEHAVIOURAL THERAPY

- **SIMPLIFIED HABIT REVERSAL-**

  -Awareness Training

  -Competing Response Training

  -Relaxation Training (optional)

  -Social Support

PSYCHOTHERAPY

- Individual supportive therapy
- Family therapy
- Group therapy

EDUCATIONAL INTERVENTIONS

- Learning problems and classroom difficulties occur commonly in tic disorder patients.
- Specific developmental disorders and ADHD- or OCD-related symptoms may interfere with academic performance.
- Provide consultation, guidance, and education to teachers or employers of patients with tic disorders.

EDUCATIONAL INTERVENTIONS

- Creating moderate, task-oriented structure in the classroom, preferential seating
- One-to-one support, or individualized educational or work plans.
- Writing aides, such as silent typewriters and computers.

EDUCATIONAL INTERVENTIONS

- Specific workplace interventions include-

  -structured tasks

  -organization of tasks into smaller units

  -flexible time limits


sniff!
sniff!
Illustration by Park Gee-young
The Korea Herald

THANK YOU