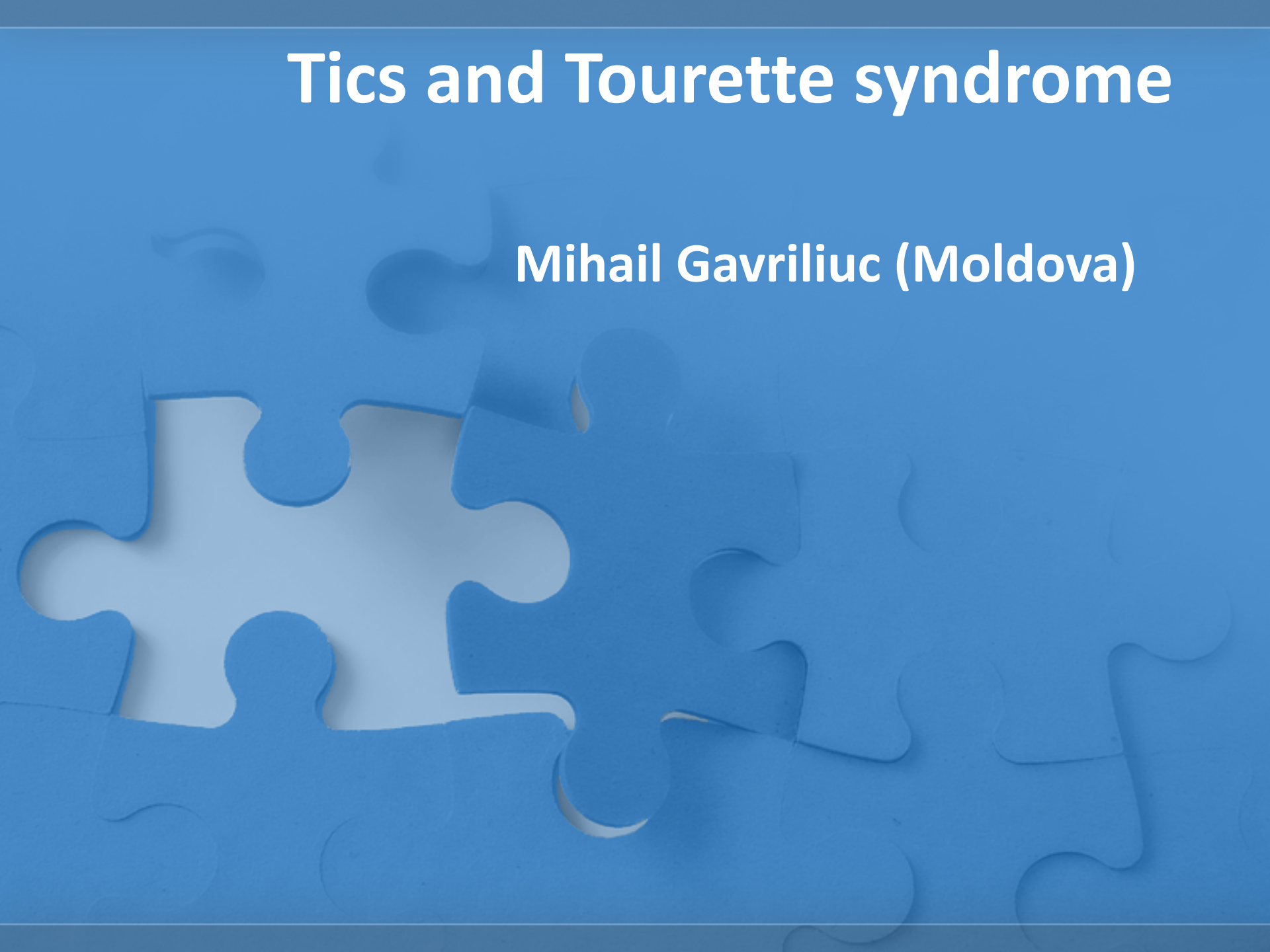


Tics and Tourette syndrome

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DECLARATIONS

Competing interests

- Wife - Chief of Neuro Lab
- Son – Neurologist
- Daughter-in-law - Neurologist
- Daughter – Neurologist
- Nephew - Neurologist
- Son-in-law – Neurosurgeon
- Granddaughter – the future Neuroscientist...

Funding

- No external funding was sought or granted for this work

TICS

Definition

Rapid brief repetitive purposeless semi- or involuntary movement, occurring at irregular intervals.



<https://en.wikipedia.org/wiki/Tic#>

TICS

Definition

Tic can be temporarily suppressed by an effort of will.

idiopathic (primary) tics

- Simple tic
- Multiple (complex) tics
- Multiple (complex) tics with vocalization
(Gilles de la Tourette's syndrome)

TICS

| | | Simple (examples) | Complex (examples) |
|--------|--------------------|----------------------|-----------------------|
| | | MOTOR | |
| | Head jerking | | Touching people |
| | Shoulder shrugging | | Touching objects |
| | Hand clapping | | Ecopraxia |
| | Neck stretching | | Copropaxia |
| | | | |
| PHONIC | Vocal | Throat clearing | |
| | | Sniffing | |
| | | Grunting | |
| | Verbal | | Echolalia |
| | | | Palilalia |
| | | | Lexilalia |
| | | | Coprolalia |

Symptomatic (secondary) tic syndromes

| Congenital | | Acquired | | |
|-------------------------------|---|--|--|---------------------------|
| Inherited | Huntington's disease | Vascular | Cerebral hemorrhage or infarction | |
| | Generalized idiopathic torsion dystonia | | Trauma | Birth trauma |
| | Choreoacanthocytosis | Head injury in childhood or adult life | | |
| Tuberous sclerosis | Peripheral injury | | | |
| Citrullinaemia | Infections | Encephalitis | | |
| Richner-Hanhart | | PANDAS | | |
| Genetic | Down's syndrome | Neoplastic | Brain tumour | |
| | Anomaly: XYY | | 'Degenerative' | Senile tic |
| | Anomaly: XXX/9p | Toxic | | Carbon monoxide poisoning |
| | Anomaly: Fragile-X | | | Mercury intoxication |
| | Anomaly: 9p monosomy | Metabolic | Hypoglycemia | |
| Anomaly: 18q22 translocations | Drugs | | a. L-dopa. b. Antipsychotics. c. Stimulants. d. Antiepileptics. e. Antidepressants. f. Alcohol withdrawal. g. Illicit drugs: amphetamines, cocaine, heroin, methylphenidate, pemoline. | |
| Developmental | | Autism | | |
| | Asperger's syndrome | | | |

simple tics

- estimates of children showing such tics have varied from 4 to 25%, but most are in the range of 10% or more
- appear from 2 years onwards and mean age of onset has been found to be about 7 years
- may show a familial tendency

simple tics

- most frequently involve the face, particularly around the eyes
- winking, unilateral blinking, raising the eyebrows, frowning, moving the scalp, flaring the nostrils, grimacing, and wriggling the ears
- sniffing, snorting, throat clearing, coughing, humming, and making clicking noises with the tongue



<https://en.wikipedia.org/wiki/Tic#>

simple tics

- intelligible speech is uncommon
- shoulder shrugging, head nodding, and head banging may occur
- tics involving the arms, hands, lower limbs, or trunk are less frequent but can result in touching, slapping, gesturing, stamping, kicking, and the like

simple tics

- involve a single movement of the face or head, appearing spontaneously and lasting weeks or months before clearing
- sometimes they may recur or be followed by a similar movement
- occasionally, different types of tic can be contemporaneous

simple tics

- the vast majority of these movements eventually disappear without trace
- infrequently simple tics persist into adult life
- may also remain and appear as a mannerism or part of the person's personality

simple tics

- treatment is usually not required for simple tic although botulinum toxin injections can be effective (Marras *et al.* 2001).
- explanation and reassurance should suffice
- advise parents to overlook the symptoms and avoid emphasizing them by attempts at correction

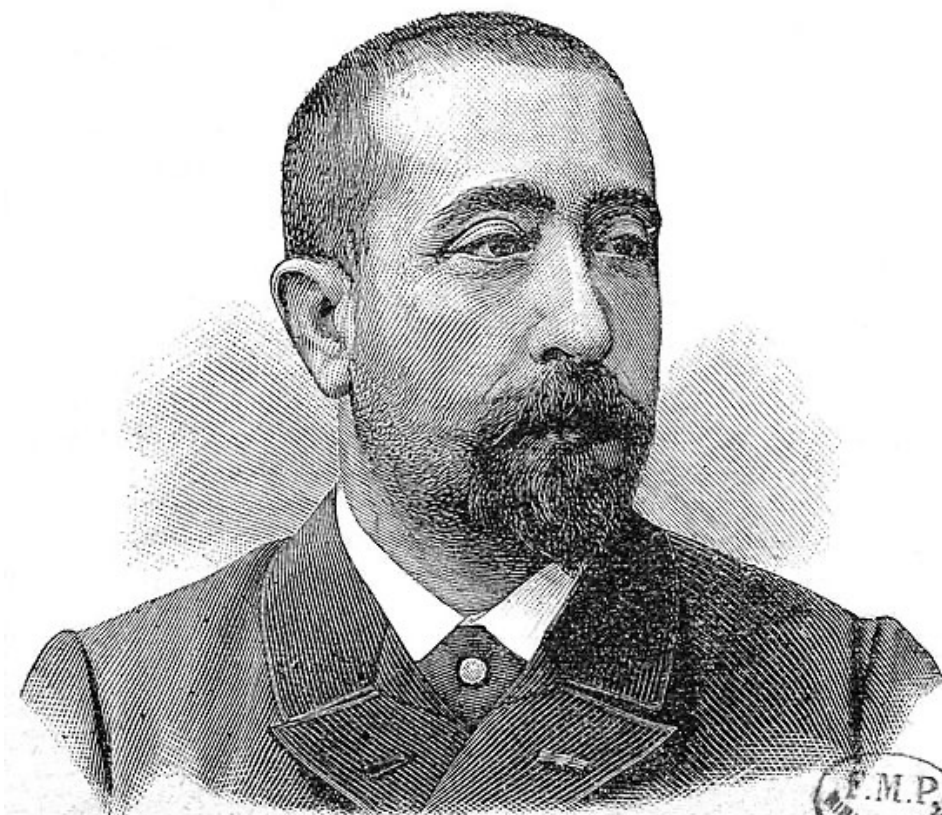
Gilles de la Tourette's syndrome

Definition

Chronic disorder characterized by multiple involuntary tics, including at least one vocal tic.

Georges Albert Édouard Brutus Gilles de la Tourette

(1857 – 1904)

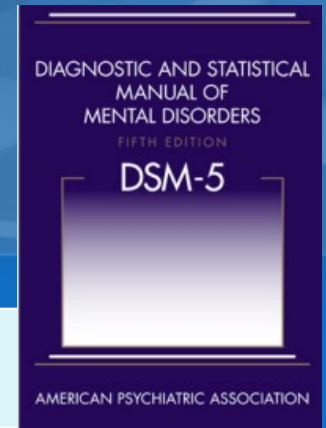


M. le D^r GILLES DE LA TOURETTE,
Médecin des Hôpitaux de Paris, directeur en chef
du service médical de l'Exposition de 1900.

Cliché E. Pirou.



Diagnostic criteria (DSM-V) for Gilles de la Tourette's syndrome



1. Age of onset during childhood or adolescence; before age 18.

2. Multiple involuntary motor tics, or involuntary sensory tics that may induce voluntary tonic or other movements.

3. One or more involuntary vocal tics. Motor and vocal tics have been present at some time during the illness, although not necessarily concurrently.

4. Tics irresistible but can be suppressed for seconds to hours.

5. Variations in the number, frequency, type, location, or severity of tics occur over time ('waxing or waning'). The tics occur many times a day (usually in bouts) nearly every day or intermittently throughout a period of more than 1 year, and during this period there was never a tic-free period of more than three consecutive months.

6. The disturbance is not due to the direct physiological effects of a substance (e.g. stimulants) or a general medical condition (e.g. Huntington's disease or post-viral encephalitis).

Gilles de la Tourette's syndrome

- complex motor tics consist of more coordinated and complicated movements, such as touching, smelling, and copropraxia (obscene gestures)
- motor tics usually first appear in the face, but multiple body regions can be involved
- over time, tics often recede from one body part and evolve elsewhere

Gilles de la Tourette's syndrome

- simple vocal tics include inarticulate noises and single sounds, such as throat clearing and grunting
- complex vocal tics have linguistic meaning and consist of full or partial words, such as echolalia and coprolalia (obscene words)
- coprolalia may be mild and transient, and it occurs in only a minority of cases

Gilles de la Tourette's syndrome

| Types of vocalizations | | | |
|------------------------|------------------------------|---------------|-------------------------|
| Grunt | <i>grohăit</i> | Whoop | <i>strigăt</i> |
| Throat clearing | <i>curățatul gâtului</i> | Hiss | <i>sâsăit</i> |
| Bark | <i>lătrat</i> | Growl | <i>mârăit</i> |
| Snort | <i>sforăit</i> | Wa Wa | <i>(va va)</i> |
| Ugh | <i>(expresie de dezgust)</i> | Sucking noise | <i>zgomot de sugere</i> |
| Ah | <i>(interjecție)</i> | Sh Sh Sh | <i>(ș ș ș)</i> |
| Gulp | <i>înfulecat, înghițit</i> | Pant | <i>gâfăit</i> |
| Hiccough | <i>sughiț</i> | Wail | <i>geamăt</i> |
| Click | <i>pocnet</i> | Eh | <i>Eh</i> |
| Hum | <i>zumzet</i> | Quack | <i>măcăit</i> |
| Squeak | <i>chițăit</i> | Gasp | <i>icnit</i> |
| Shriek | <i>urlet</i> | Cluck | <i>cotcodăcit</i> |
| Burp | <i>răgăit</i> | Yelp | <i>schelălăit</i> |
| Hoot | <i>chiuit</i> | Yahoo | <i>(neplăcut)</i> |
| Ooh | <i>(Uuh)</i> | T, T, T, T | <i>T, T, T, T</i> |
| Moan | <i>meaunat</i> | Clug | <i>Bulbuc</i> |

1 op 100 doet het

Vocal tics in motion – Tourette Awareness
<https://vimeo.com/97056475>

Gilles de la Tourette's syndrome

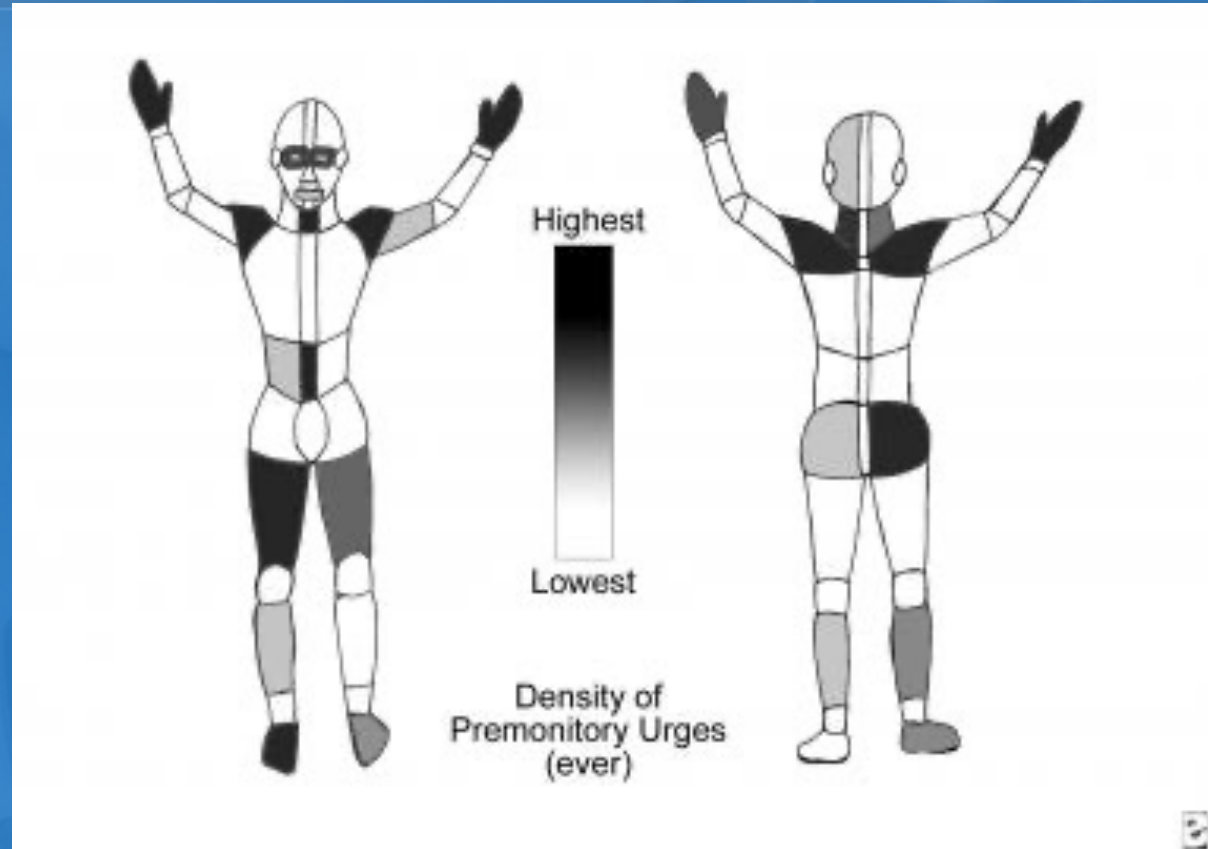
Copropoxia is the performance of obscene acts and gestures. Like coprolalia, copropoxia does not usually commence until adolescence. Such complex tics are common, occurring in at least 20-30% of cases (Nee *et al.* 1980, Lees *et al.* 1984). Actions include the pal-backed 'V' sign, exhibiting and erect forefinger with a clenched fist, pelvic thrusts, and hand movements simulating masturbation.

Gilles de la Tourette's syndrome

Coprophagia?

Gilles de la Tourette's syndrome

some patients describe focal uncomfortable sensations (sensory tics or premonitory urge) from which they attempt to obtain relief with movements often interpreted as voluntary.



The feeling of urge is similar to the need to yawn, sneeze, blink or scratch on it.

Gilles de la Tourette's syndrome

- ~1/2 of patients report symptoms of obsessive compulsive disorder, eg, checking, counting, perfectionism, and another 1/2 have evidence of attention deficit hyperactivity disorder, eg, inattention, impulsivity, distractibility, hyperactivity



Gilles de la Tourette's syndrome

Echo-phenomena consist of forced repetition of other's words and phrases (echolalia) or actions (echopraxia).

Echolalia is present in about 40% and echopraxia - in about 20% of patients.

Mean age of onset is approximately 13 years .

Patients may repeat single words, phrases, or sentences. Sometimes they imitate accent or speech peculiarities and famous people may be mimicked. Animal sounds can be a part of the repertoire.

Gilles de la Tourette's syndrome

Other neurological abnormalities

- Stuttering (over 30%)
- Associated dystonia
- Startle response with generalized body jerks and blinking after a loud unexpected noise
- 'Soft' neurological signs (altered tone, mild incoordination, dysdiadokochinesia, equivocal or extensor plantar response without weakness, intermittent postural abnormalities, and cramp on rapid movement. Migraine has been reported to be more common in children with Gilles de la Tourette's syndrome than in controls.
- Restless legs
- Compressive neuropathy and radiculopathy, cervical myelopathy.

Gilles de la Tourette's syndrome

- a variety of other behavioral disorders, eg, anxiety disorders, phobias, have been associated with Tourette's syndrome as well
- obsessive-compulsive disorder, and perhaps other behavioral disturbances as well, may represent alternative expressions of the Tourette's syndrome trait

Gilles de la Tourette's syndrome

Sleep disturbances, including enuresis, somnambulism, and night terrors, have been noted in between 30 and 50% of children with Gilles de la Tourette's syndrome (Corbett *et al.* 1969, Nee *et al.* 1980, Glaze *et al.* 1983, Barabas *et al.* 1984, Comings and Comings 1987).

Gilles de la Tourette's syndrome

Diagnostic Workup:

- simple observation
- neuroimaging or other diagnostic studies are generally not required
- psychiatric evaluations and testing with standardized neuropsychologic measures of attention and obsessive-compulsive behavior, may be helpful for diagnosing associated behavioral disturbances

Gilles de la Tourette's syndrome

Management:

- most patients can avoid the use of medications
- educating patients, family members, peers, and school personnel regarding the nature of Tourette's syndrome, restructuring the school environment, and providing supportive counseling are measures that may be sufficient

Gilles de la Tourette's syndrome

Medical therapy

- ***Dopamine receptor blocking drugs:***
 - HALOPERIDOL (0.25 to 15 mg/day)
 - PIMOZIDE (1 to 10 mg/day)
 - FLUPHENAZINE (2 to 15 mg/day)
 - RISPERIDONE (0.5 to 16 mg/day)
 - ARIPIPRAZOLE (5 to 30 mg/day)

Gilles de la Tourette's syndrome

Medical therapy

- ***Alpha agonists:***

- CLONIDINE (0.05 to 0.5 mg/day)
- GUANFACINE (0.5 to 10 mg/day)

- ***Other drugs:***

- CLONAZEPAM (0.5-10 mg/day)
- TETRABENAZINE (25-200 mg/day)

Gilles de la Tourette's syndrome

Medical therapy

- ***Antidepressants:***

- FLUOXETINE (20 to 80 mg/day)
- CLOMIPRAMINE (25 to 250 mg/day)
- SERTRALINE (50 to 200 mg/day)
- PAROXETINE (20 to 50 mg/day)

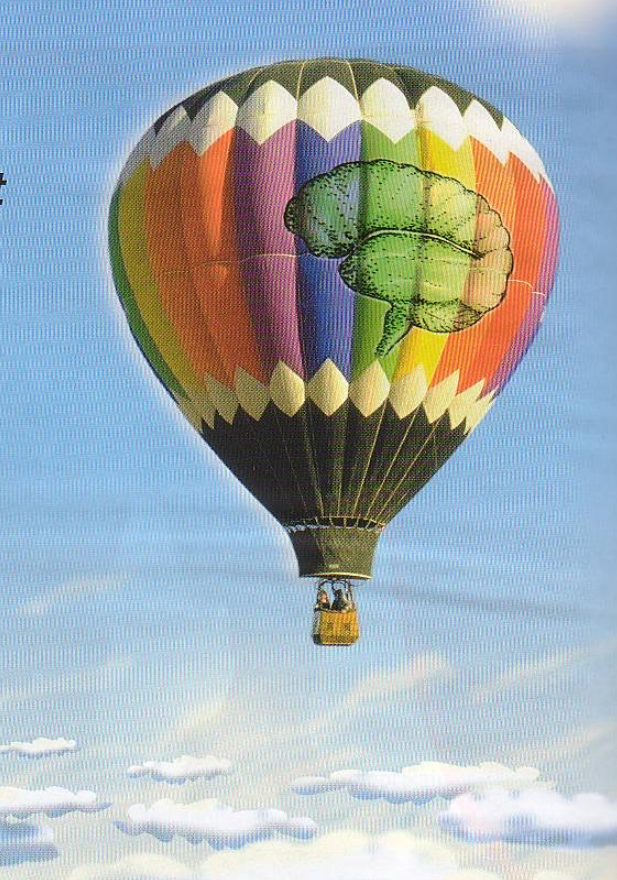
Gilles de la Tourette's syndrome

Surgical therapy

- Stereotactic thalamotomy (in the past)
- Deep brain stimulation
- Stereotactic surgery to the limbic system

Other therapy

- Botulinum toxin injections



"Knowledge is of two kinds. We know a subject ourselves, or we know where we can find information on it."

-- *Samuel Johnson*

thank you

merci

obrigado

grazie

gracias

kiitos

спасиби

danke

tack

köszönöm

хвала

teşekkür ederim

ačiū

hvala vam

tak

þakka þér

σας ευχαριστώ

ddiolch 'ch

Dank u

dëkuj

dziękuję

ありがとう

תודה

Ďakujem

hvala

Takk

благодаря

Tapadh leibh

falemmnderit

شكرا

trugārez

mulțumesc

tānan teid

дзякуй

спасибо

Ви благодарам

谢谢

धन्यवाद

Go raibh maith agaibh

Paldies