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REVIEW ARTICLE

Tourette Syndrome: a literature review

Síndrome de Tourette: una revisión de la literatura

Síndrome de Tourette: uma revisão bibliográfica

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Abstract

The aim was to review the historical, and clinical aspects, symptoms, diagnosis, and treatment of Tourette Syndrome (TS) in the literature. The study is a bibliographic review, written in an integrative and systematic manner. (TS) was described from the observation of people who had tics and obscene behavior, in which the sufferer was unable to control them. The "International Statistical Classification of Diseases and Related Health Problems" allocates TS to the group of disorders with emotional disturbances. TS is a chronic neuropsychiatric condition characterized by stereotyped actions, multiple motor tics, and/or vocal tics, which generally appear during early adolescence without specific etiology. The treatment of TS is carried out through a pharmacological and non-pharmacological combination and, in some cases where the treatment is ineffective, surgical treatment. The results demonstrated that pharmacological treatment is effective, but for more satisfactory results it is necessary to associate it with non-pharmacological treatment.

Descriptors: Tourette's Syndrome; Tics; Medication; Social Behavior; Quality of Life.

Resumén

El objetivo fue revisar los aspectos históricos, clínicos, síntomas, diagnóstico y tratamiento del Síndrome de Tourette (ST) en la literatura. El estudio es una revisión bibliográfica, escrita de manera integradora y sistemática. (TS) se describió a partir de la observación de personas que presentaban tics y conductas obscenas, en las que quien los padecía era incapaz de controlarlos. La "Clasificación Estadística Internacional de Enfermedades y Problemas de Salud Relacionados" clasifica el ST en el grupo de trastornos con alteraciones emocionales. El ST es una condición neuropsiquiátrica crónica caracterizada por acciones estereotipadas, múltiples tics motores y/o tics vocales, que generalmente aparecen durante la adolescencia temprana sin una etiología específica. El tratamiento del ST se realiza mediante una combinación farmacológica y no farmacológica y, en algunos casos en los que el tratamiento resulta ineficaz, mediante tratamiento quirúrgico. Los resultados demostraron que el tratamiento farmacológico es eficaz, pero para resultados más satisfactorios es necesario asociarlo a un tratamiento no farmacológico.

Descriptores: Sindrome de Tourette; Tics; Medicamento; Comportamiento Social; Calidad de Vida.

Resumo

Objetivou-se revisar na literatura os aspectos históricos, clínicos, sintomatologia, diagnóstico e tratamento para a Síndrome de Tourette (ST). O estudo trata-se de uma revisão bibliográfica, redigida de maneira integrativa e sistemática. A (ST) foi descrita a partir da observação de pessoas que apresentavam tiques e comportamentos obscenos, em que o portador é incapaz de controlá-los. A "Classificação estatística internacional de doenças e problemas relacionados à saúde" aloca a ST no grupo de transtornos com perturbações emocionais. A ST é uma condição neuropsiquiátrica crônica caracterizada por ações estereotipadas, tiques motores múltiplos e/ou tiques vocais, que geralmente aparecem durante o início da adolescência sem etiologia específica. O tratamento da ST é feito pela associação farmacológica e não farmacológica e em alguns casos em que o tratamento está sendo ineficaz, o tratamento cirúrgico. Os resultados demonstraram que o tratamento farmacológico é eficaz, porém para resultados mais satisfatórios é necessário a associação com o tratamento não farmacológico.

Descritores: Síndrome de Tourette; Tiques; Medicação; Comportamento Social; Qualidade de Vida.

Introduction

Tourette's Disorder, Tourette's Syndrome (TS) or Gilles de la Tourette's disease was best described in 1885, by George Gilles de la Tourette, a medical student of Jean-Martin Charcot, who observed cases of people who presented symptoms such as multiple tics, coprolalia, palilalia, echolalia and non-obscene socially inappropriate behaviors (NOSIBs). Currently, the "International Statistical Classification of Diseases and Related Health Problems" allocates TS to the group of disorders with emotional and behavioral disturbances that begin in childhood and adolescence[1,2].

This disorder is a chronic neuropsychiatric condition characterized by stereotyped actions: multiple motor tics and/or vocal tics. These tics are defined as sudden, rapid, recurrent, and non-rhythmic motor movements or vocalizations and patients report an inability to control them and feel a premonitory sensation whose relief occurs after the tic has occurred, which tends to reduce Quality of Life. (QoL) of these individuals. Studies indicate that TS has a prevalence of 1%, being more common in young males, with an approximate ratio of 3 to 4 affected men for every woman[1,3,4].

The objective of the present study was to review the literature on aspects of TS and the consequences on the quality of life of its sufferers, in addition to demonstrating the benefits of early diagnosis and treatment.

Methodology

The study is a bibliographical review and for its execution the terms were researched: "Tourette Syndrome", "Tics", "Medication", "Social Behavior" and "Quality of Life" on the platforms: Virtual Health Library and its databases, Online Electronic Scientific Library (SciELO) and Online Medical Literature Analysis and Retrieval System (MedLine).

The inclusion criteria were original articles, revisions, and experience reports in Portuguese, English, or Spanish, published between 2010-2022, available online, and relevant to the topic sought. Articles in languages other than Portuguese, English, or Spanish and articles that were published before 2010 were excluded. Furthermore, printed articles were not used.

Results and Discussion

General aspects and epidemiology

TS is a disorder in which there is a chronic neuropsychiatric condition characterized by stereotyped actions: multiple motor tics and/or vocal tics. These tics are defined as motor movements or vocalizations that are sudden, rapid, recurrent, and non-rhythmic, and patients report being unable to control them and feel a premonitory sensation, the relief of which only occurs after the tic has occurred, which tends to reduce the Quality of Life (QoL) of these individuals. Studies indicate that TS has a prevalence of 3 to 8 cases per 1000 children and 1% of the world population, being more common in young males, with an approximate ratio of 3 to 4 affected men for every 1 woman[1,3-6].

Etiology

The etiology of Gilles de la Tourette's disease remains nonspecific, initially, studies pointed to an autosomal dominant genetic disorder. Another factor analyzed regarding the cause of tics is streptococcal infections. According to this hypothesis, infections may lead to the formation of anti-neuronal antibodies. Although there is analysis of environmental factors, genetic factors are considered the primary contributors to the pathogenesis of this disease, however, genetic mutations or risk alleles have not yet been identified. While the first studies on the genetics of the syndrome focused on multigenic lineages and suggested Mendelian inheritance, subsequent segregation analyses point to a more complex inheritance pattern. After the identification of rare genetic mutations associated with the TS phenotype, a monogenic inheritance model was proposed. In models of monogenic disorders, there is variation in the dominant or both recessive alleles. In research into TS, although no specific mutation has been found, genetic findings may allow the identification of the affected pathways and may lead to the development of new treatment strategies[3,7].

Pathophysiology

There is evidence that TS is manifested by dysfunctions in the cortico-striatum-thalamo-cortical circuit and by hyperexcitability in cortical motor areas, which leads to the perception of symptoms on a broad spectrum. Furthermore, it has been demonstrated that the neurotransmitter gamma-aminobutyric acid (GABA) plays an important role in the manifestation and clinical control of TS. GABA concentrations in the supplementary motor area are paradoxically elevated in individuals with tics. Scientific evidence indicates that GABA concentrations in the supplementary motor area have a negative correlation with cortical excitability in the primary motor cortex. As a result, it is proposed that GABA contributes to the management of TS, due to tonic inhibition located in the supplementary motor area, which could lead to the suppression of tics[8].

Diagnostic method

The American Psychiatric Association, based on criteria from the Diagnostic Manual and Statute of Mental Disorder (DSM-5), defines that to diagnose Tourette's Disorder, patients must present motor and/or vocal tics. , concomitant or not; and the frequency of these stereotyped movements can increase or decrease for more than a year since the first tic, and the onset of manifestations of these symptoms must be before 18 years of age. In general, in the clinical profile of this syndrome, the initial manifestations generally occur between 11 and 12 years of age, and the disturbances (tics) must not be attributable to the physiological effects of any substance or to any other medical condition that could explain[3,4,6].

Clinical manifestations

The individual with TS can often go through embarrassing situations in the eyes of those around them and who do not know about what they are dealing with, as

the patient will present motor movements and decontextualized vocalizations, so it is necessary to pay attention to the tendency towards isolation, in cases where patients are ashamed of their symptoms and fear that they will be judged inappropriate. Therefore, it can be said that there are impacts on both the sociocultural and educational aspects of individuals living with this disorder, mainly because this syndrome is clinically related to other characteristic comorbidities, for example, attention deficit disorder, hyperactivity, obsessive-compulsive disorder (OCD), sleep disorders, anxiety, depression, aggressive behavior, oppositional disorder, defiant, emotional lability, and impulsivity. In this sense, early diagnosis is extremely important to ensure progressive improvement in the clinical condition of individuals, ensuring the best professional approach, avoiding the impacts of the syndrome on social life, individual development, and improvement in QoL[3,9].

Treatment

Due to all the symptoms caused by TS, such as social difficulties, anxiety, obsessive-compulsive disorder, and attention deficit hyperactivity disorder. Treatment for TS must correlate pharmacological and non-pharmacological treatment, with surgical treatment being in some cases non-resolutive. When approaching pharmacological treatment, some drug classes are proposed, such as dopamine receptor blockers, alpha-2-agonists, antiepileptic drugs, GABA agonists, and the injectable use of botulinum toxin. Despite the different classes, dopamine receptor blockers (BRD) are the only ones approved by the US Food and Drug Administration (FDA). The BRDs used are Haloperidol, Pimozide, and Aripiprazole, but they have some side effects that can lead to treatment abandonment, for example, drowsiness, increased QT interval, weight gain, and increased appetite, among others that are less prevalent[10].

Alpha-2-agonists are used as first-line for tics considered to be of moderate intensity, the main representatives of this class are Clonidine and Guanfacine. Despite being from the same drug class, each one has a different action, Clonidine acts on the locus coeruleus reducing the amount of noradrenaline, while Guanfacine stimulates the alpha-2-adrenergic center to inhibit the sympathetic impulse to blood vessels and the heart. In a previous clinical trial, the use of Clonidine was proposed as a means of improving tics in children with attention deficit hyperactivity disorder, however, the use of Clonidine in patients with TS is preferable, in patients who are also affected by HDA if there is this synergism, there is a significant improvement in tics, but it showed that the use of Clonidine in patients with TS alone, there was no significant improvement in tics[11].

The use of antiepileptics has shown a satisfactory response, especially with the use of Topiramate at a dosage of 118 mg per day versus placebo, there was an improvement of 53.6% in tics in the group using Topiramate. Therefore, the use of Topiramate has become a promising alternative for ST, even if isolated. There are reports on the use of benzodiazepines (BZ) as an adjuvant treatment for TS, related to anxiety, however, there is still no evidence for this, BZ can cause side effects such as drowsiness and slowed thinking, in addition to causing tolerance and dependence[12,13].

Another reported treatment for TS is the injectable use of botulinum toxin, which consists of applying botulinum toxin to the places where the tic occurs, seeking to inhibit their occurrence, but there is no improvement in the sensation of premonition. According to a study carried out with 18 subjects divided into intervention and control groups, botulinum toxins were applied to people with tics in the facial, neck and shoulder regions. The results showed an improvement of 39% in patients who received the toxin and a worsening of 5.8% in patients who did not receive the toxin[14].

In addition to pharmacological treatment, non-pharmacological treatment must be carried out to achieve more satisfactory results. The main non-pharmacological treatment is Habit Reversal Training (HRT). Cognitive behavioral intervention for tics (ICCT) and cannabis use. However, treatment with Cannabis is still questioned, as its side effects are largely greater than the effects related to tics. TRH is divided into three components, these are awareness training, relaxation training, and competitive response training; the first consists of identifying when the so-called tics will occur, and the competitive response is identifying acts that are inhibitory to the tics and practicing them when there is a premonition of the tics, and finally the relaxation exercise is exercises carried out to reduce the symptoms. of stress and anxiety. ICCT is the practice of HRT associated with psychoeducation and behavioral interventions. It is worth mentioning that ICCT replaces relaxation therapy with functional interventions, such as changing lifestyle habits to reduce symptoms of stress and anxiety[9,15].

The use of Cannabis is associated with a significant reduction in the number and intensity of tics in RCTs. However, the use of Cannabis as a non-pharmacological treatment for Tourette's Syndrome is still questioned, as its production is not standardized, in addition to presenting significant side effects such as drowsiness, increased appetite, dizziness, headache, ocular hyperemia, dry mouth, anxiety, decrease short-term memory and decreased concentration, psychosis and euphoria[15,16].

Surgical treatment for Tourette's Syndrome is still very limited and is not easily accessible, as it is necessary to meet detailed criteria and as it is a treatment that is in the study phases. A study was carried out reviewing the use of ablative surgery in ST between the years 1960 and 2000, finding that the use of this surgical technique did not cause great benefits, and its use was questioned, due to the exposure of patients to high-risk surgeries, being minimal benefit, showing little or no decrease in tics[10,17].

However, new surgical expectations have emerged with the study of Deep Brain Stimulation (DBS), which works to stimulate three main areas, which are the centromedian-parafascicular complex of the thalamus, the internal anteromedial globus pallidus and the posteromedial globus pallidus. internal ventral. These stimuli have been related to good outcomes, that is a considerable reduction in tics, being

an excellent therapeutic option in refractory cases, in open reports. However, when the research criteria are applied, in a randomized and blind manner, the results obtained are not robust and positive, so there are doubts regarding the use of the treatment and its cost-benefit[10,18].

However, the use of ECP should not be done indiscriminately, but rather after thorough evaluation, application of criteria, and use only in cases refractory to conventional treatments. The criteria are manual diagnoses and statistics of mental illnesses; age over 18, except in cases of greater severity, this criterion being not absolute and dependent on the ethics council of the health center used; tics graded as severe by the Yale Global Tic Severity Score, being greater than or equal to 35 out of 50 points; tics as the primary cause of dysfunction; refractoriness to conventional treatment; if other medical, psychiatric or neurological conditions have been stable for more than 3 months; patient acceptance for treatment; patient with a neuropsychological situation to support the surgical procedure, post-surgery and likely non-improvement of the condition, these are the eligibility criteria for surgical treatment in patients suffering from Tourette's Syndrome[18].

There is also discussion about the use of traditional Chinese medicine as an adjuvant treatment and this has been used to complement other treatments. Therefore, the use of Ningdong granules, whose mechanism of action is the elevation of homovanillic acid, which is related to the regulation of Dopamine, proved to be effective. Additionally, other practices I see being used involve the Qunfeng Zhidong recipe and the use of the active compound from Gastrodia elata to inhibit stereotypical behaviors and alleviate symptoms in TS[19,20].

Conclusion

This article sought to demonstrate what aspects of patients with Tourette Syndrome are, what criteria are used for diagnosis, and what treatments are used. Even in the face of a syndrome whose etiology is still unknown, some pharmacological and non-pharmacological measures have been demonstrated, which can help in the treatment of this syndrome, also highlighting the use of surgical treatment for cases refractory to pharmacological and non-pharmacological treatment, being, therefore, cited and weighed the benefits of surgical treatment.

The need to carry out follow-ups to help control tics and improve the quality of life of these patients was also highlighted. It is worth highlighting that to better elucidate the etiology of the syndrome and to achieve better outcomes, continued research in the area is necessary.

However, this bibliographical review has limitations, as more studies are needed with new diagnostic and therapeutic approaches so that people with TS can benefit and improve their QoL.

References

1. Greydanus DE, Tullio J. Tourette's disorder in children and adolescents. Translational Pediatrics. 2020;9(Suppl 1):S94-S103. doi: 10.21037/tp.2019.09.11
2. Organização Mundial da Saúde (OMS). CID-10 Classificação Estatística Internacional de Doenças e Problemas Relacionados à Saúde. 10 ed. São Paulo: Universidade de São Paulo; 1997.
3. Ferreira ACGR. Revisão da Literatura sobre a Síndrome de Tourette. Fed. Nac. das Apaes (Fenapaes) [Internet]. 2019 [acesso em 11 nov 2023];12(2):22-34. Disponível em: https://apaeciencia.org.br/index.php/revista/article/view/126
4. Robertson MM. Tourette syndrome, associated conditions and the complexities of treatment. BRAIN. 2000;123(3):425-462, 2000. DOI: 10.1093/brain/123.3.425
5. Mattos JP, Rosso ALZ. Tiques e síndrome de Gilles de la Tourette. Arq. Neuro-Psiquiatr. 1995;53(1):141-146. https://doi.org/10.1590/S0004-282X1995000100022
6. American Psychiatric Association. Manual diagnóstico e estatístico de transtornos mentais: DSM-5. 5. ed. Porto Alegre: Artmed; 2014.
7. Deng H, Gao K, Jankovic J. The genetics of Tourette syndrome. Nature Reviews Neurology. 2012;8(4):203-213. DOI: 10.1038/nrneurol.2012.26
8. Draper A, Stephenson MC, Jackson GM, Pépés S, Morgan PS, Morris PG, Jackson SR. Increased GABA contributes to enhanced control over motor excitability in tourette syndrome. Current Biology. 2014;24(19):2343–2347. DOI: 10.1016/j.cub.2014.08.038
9. Gonçalves DM, Silva NG, Estevam ID. Síndrome de Tourette e terapia cognitivo-comportamental: um estudo de caso. Rev. bras.ter. cogn. [online]. 2019;15(1):51-58. DOI: 10.5935/1808-5687.20190008
10. Jimenez-Shahed J. Medical and Surgical Treatments of Tourette Syndrome. Neurol Clin. 2020;38:240-366. DOI: 10.1016/j.ncl.2020.01.006
11. Weisman H, Qureshi IA, Leckman JF, Scahill L, Bloch MH. Systematic review: pharmacological treatment of tic disorders–efficacy of antipsychotic and alpha-2 adrenergic agonist agents. Neurosci Biobehav. 2013;37(6):162-1171. DOI: 10.1016/j.neubiorev.2012.09.008
12. Jankovic J. Treatment of tics associated with Tourette syndrome. Journal of Neural Transmission. 2000;127(5):843-850. DOI: 10.1007/s00702-019-02105-w
13. Cheng T, Wallace DM, Ponteri B, Tuli M. Valium without dependence? Individual GABAA receptor subtype contribution toward benzodiazepine addiction, tolerance, and therapeutic effects. Neuropsychiatric disease and treatment. 2018;14:1351-1361. DOI: 10.2147/NDT.S164307
14. Marras C, Andrews D, Sime E, Lang AE. Botulinum toxin for simple motor tics: a randomized, double-blind, controlled clinical trial. Neurology. 2001;56(5):605-610. DOI: 10.1212/wnl.56.5.605
15. Müller-Vahl KR. Treatment of Tourette syndrome with cannabinoids. Behavioural neurology. 2013;27(1):119-124, 2013. DOI: 10.3233/BEN-120276

16. Sokratous S, Kaikoush K, Mpouzika MD, Alexandrou G, Karanikola NM. Attitudes, beliefs and knowledge about medical cannabis among nurses and midwives in Cyprus: a cross-sectional descriptive correlational study. BMC Nurs. 2022;21(1):120. DOI: 10.1186/s12912-022-00887-1
17. Temel Y, Visser-Vandewalle V. Surgery in Tourette syndrome. Movement disorders: Official Journal of the Movement Disorder Society. 2004;19(1):3-14. DOI: 10.1002/mds.10649
18. Jimenez-Shahed J. Design challenges for stimulation trials of Tourette´s syndrome. The Lancet. Neurology. 2015;14(6):563-565. DOI: 10.1016/S1474-4422(15)00043-5
19. Wang N, Qin DD, Xie YH, Wu XC, Wang DY, Yang H, Li XX, Xiong L, Liang JH. Traditional chinese medicine strategy for patients with tourette syndrome based on clinical efficacy and safety: a meta-analysis of 47 randomized controlled trials. Biomed Res Int. 2021;6630598. DOI: 10.1155/2021/6630598
20. Wang Y, Zhao L, Li A. Gastrodin-A potential drug used for the treatment of Tourette Syndrome. Journal of Pharmacological Sciences. 2021;145(3):289-295. DOI: 10.1016/j.jphs.2021.01.005