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## CRANIOFACIAL DISORDERS IN THE COURSE OF TOURETTE'S SYNDROME - A REVIEW OF THE LITERATURE

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### ABSTRACT:

**INTRODUCTION:** Tourette's syndrome is a neurological disorder in which the patient presents first symptoms, such as motor and voice tics, in childhood. If the disease significantly impairs proper functioning in society, symptomatic treatment is applied. Antipsychotics, dopaminergic receptor blockers, VMAT2 inhibitors, and non-pharmacological treatment are used, leading to a reduction in the incidence of tics by up to 50%.

**AIMS:** The aim of the work is to present the symptoms of the disease, in particular concerning the craniofacial region and the possibility of implementing any treatment.

**MATERIALS AND METHODS:** All data comes from the analysis of the PubMed database. The paper presents cases in which the structures of the oral cavity, eyes and the brain have been damaged.

**DESCRIPTION OF THE STATE OF KNOWLEDGE:** Treatment with TS must take place on many levels and includes pharmacological and non-pharmacological treatment, therefore, patient care should be interdisciplinary.involving specialists such as psychologists, nutritionists, neurologists, anesthesiologists and dentists. The task of such a team is to diagnose the Tourette syndrome, improve patients quality of life, even by limiting the effects

of medications (weight gain) or specialist treatment of damage and discomfort caused by symptoms. Psychological therapy remains the primary one, which is why a qualified team of psychologists and psychotherapists is important. Dentists are an important part of the interdisciplinary team, due to the high frequency of tics covering the oral cavity and the associated consequences. Decisions on treatment should be established jointly, referring to a comprehensive look at individual needs and priorities in patient therapy.

**SUMMARY:** It was found that there is a variety of types and intensity of tics in patients with Tourette's syndrome. The clinical team supervising such a patient should be an interdisciplinary team including dentists, neurologists, psychologists or orthopedics to provide patients with comprehensive and multidirectional treatment.

**KEY WORDS:** Tourette syndrome treatment; tics; head injury; oral injury; eye injury

### **Introduction:**

Tourette's syndrome is a disorder from the borderline of neurology and psychiatry, manifested by the presence of motor and vocal tics that last at least a year. In the past, Tourette's syndrome was thought to be a mental, neurological and even infectious disorder. Structural deformation showing neurological symptoms was rarely considered. Nevertheless, the origin of the disease remains unknown. The first symptoms in the vast majority of cases can be observed in 5-8 years of age. These include muscle tics, ie repetitive, non-rhythmic and stereotypical involuntary movements (eg blinking) of individual muscle groups and phonetic tics, ie cough, palylalia, echolalia, koprolalia<sup>1,2,3,4,5</sup>. In most cases, tics persist until adulthood, but with age, a reduction in their severity is observed<sup>3</sup>. Tourette's syndrome is often accompanied by obsessive-compulsive disorder (OCD) and attention deficit hyperactivity disorder (ADHD)<sup>5</sup>. Additional clinical aspects are episodes of anger, anxiety disorders, mood disorders and learning disorders<sup>6</sup>. The patient and his environment complain about a reduced quality of life due to difficult interpersonal interactions, social exclusion despite the fact that patients do not show intellectual disability<sup>3,4</sup>. Adolescents and adults report the need to perform tics as the most onerous aspect of the disease<sup>4</sup>. Patients have a higher risk of fractures in the neck and limbs, which can be reduced by implementing antipsychotic or antidepressant treatment. The increased risk of fractures is associated with widespread self-injury of patients with Tourette's syndrome. Therefore, the side effects of antipsychotics, such as sedation and weight gain, are important, probably affecting the reduction of injuries<sup>7</sup>. According to the guidelines - behavioral therapy is the basic method of treatment for patients with Tourette syndrome<sup>4</sup>. Comprehensive Behavioral Intervention for Tics (CBIT) is a type of behavioral therapy in which patients undergo psychoeducation about tics, relaxation training, tics awareness training. An important part of the therapy is training, which helps to recognize and intervene before the tick is fully developed<sup>4</sup>. Pharmacological treatment focuses on alleviating symptoms by taking antipsychotic drugs from the group of neuroleptics and those acting on noradrenergic receptors. The above examples are considered the most effective options for pharmacological treatment and are often prescribed<sup>7</sup>. The application of a botulinum toxin (BTX) is also used, which inhibits neurotransmission resulting in muscle contraction<sup>3</sup>. If the mentioned attempts of symptomatic treatment are not effective, surgical treatment can be recommended - deep brain stimulation (DBS). As a result of using DBS nerve signals that cause abnormal movements in the form of ticks are blocked<sup>3</sup>.

**Aim of study:**

The aim of the study is to present, on the basis of a literature review, the symptoms that may occur in the craniofacial region in patients with Tourette syndrome with particular emphasis on the treatment and rehabilitation of this anatomical region in these patients.

**Material and methods:**

An analysis of the PubMed internet database literature in the last 17 years was performed by entering the keyword: Tourette - assuming of its presence in titles or abstracts of searched publications. 5137 works have been obtained. After adding the word oral or tongue or ear or eye, 90, 5, 10 and 97 entries were received respectively. After reading the literature seven publications which reported on craniofacial defects in Tourette's syndrome were qualified to paper. The rejected works did not refer directly to the craniofacial disorders or the description of abnormalities in this region were too superficial.

**Results:**

Reports in which the place of the disorder was the oral cavity

Shimoyama et al. <sup>8</sup> in 2003 described a 13-year-old boy whose main complaint was chronic upper lip inflammation. The clinical picture of the Tourette syndrome in the patient consisted of: licking and touching the mouth, biting the upper lip, applying a towel over the head, putting on handkerchiefs and hitting the head. During consultations at the maxillofacial surgery clinic, ulcerative stomatitis was diagnosed most intensively in the upper lip with pain around the gum-labial connection. The boy was referred to a psychiatric consultation, which suggested the presence of the Tourette syndrome in the patient. EEG and MRI performed after a pediatric consultation were within the normal range. The boy received antibiotics, haloperidol at a dose of 3 milligrams three times a day, bromazepam at a dose of 6 milligrams three times a day, and fulnitrazepam at a dose of 1 milligram at bedtime. For sedation 5 milligrams of diazepam was injected. An acrylic splint was applied to the teeth of the mandible to protect the upper lip against chewing. Pain relief was obtained and the further process of damaging the upper lip was stopped. The biting of the upper lip after applying the splint has stopped.

Hood et al. <sup>9</sup> presented in 2004 the case of a 16-year-old female patient who was diagnosed with Tourette syndrome at the age of 12. Manifestations of this team included: stabbing the eyes with a finger, scratching the forehead, touching the teeth and biting the tongue. The patient was previously treated with clonazepam 0.5 milligrams twice a day, clonidine 0.1 milligrams twice a day and olanzapine 15 milligrams a day. In the intraoral examination, swelling of the tongue and 3-4 cm lacerations of the dorsal and lateral region of the tongue were observed. The oral mucosa on the left side contained numerous ulcers, and the lower lip was violet-blue covered with scabs. The patient was secured with tracheal intubation through the nasal cavity. During her stay in the emergency department, she received a 2-milligram benzotripine dose of 50 milligrams in two doses intramuscularly divided, diphenhydramine 100 milligrams in two intravenous doses, lorazepam in a total dose of 7 milligrams, 1 milligram intravenously, morphine in a dose a total of 6 milligrams for 2 milligrams intravenously and risperidone in a dose of 4.5 milligrams. Antibiotics were also given intravenously. Bilateral botulinum toxin was applied to the masseter muscles and the mouthpiece was adjusted to prevent chewing. After 6 days of stay, lesions within the oral

cavity were significantly improved. The patient reported more control over her behavior and was transferred to a psychiatry department in a stable state.

Lee et al.<sup>10</sup> from 2016 describes the case of a 9-year-old boy with Tourette syndrome diagnosed at age 8. The boy presented the following symptoms: humming, persistent sniffing, excessive eyelid blinking, shaking of the limbs, head banging and biting the tongue. Previous treatment has included drugs such as benztropin, escitalopram and risperidone. In the examination of the oral cavity, ulcerations of the right side of the tongue, including the tongue back, were observed. The patient experienced severe pain and burning which contributed to the difficulty in chewing and swallowing food. As part of the treatment, a 3 millimeter soft polyvinyl appliance fitted to both the maxillary and mandibular arches was used. Topical ointment and gargle with chlorhexidine were recommended. After 3 weeks of using the appliance, the language changes have improved significantly. Due to the reduction of pain and burning sensation his nutritional status improved.

#### Reports in which the disorders included the eyeball and orbital area

Margo<sup>11</sup> described in 2002 two cases: a 21-year-old and a 24-year-old man with Tourette syndrome diagnosed at age 12 and 11 years respectively. In both patients, the damage to the eyeball was due to an episode of tics during the medical examination. In a younger patient the lesion occurred as a result of hitting the doctor's left finger with the left eye. This resulted in corneal abrasions and corneal haemorrhage. The patient used topical antibiotics. The eye was healed without further consequences. In the older patient, the damage resulted from hitting the right eyeball in the tonometer's footplate. Ophthalmologic consultation revealed corneal abrasion. Antibiotic ointment was prescribed.

In 2008, Kovacich<sup>12</sup> described a case of a 25-year-old man, whose main complaint was the loss of contact lenses associated with excessive blinking. The clinical picture of the Tourette syndrome consisted of: flicking of the wrists, making humming sounds and echolalia. Haloperidol was prescribed at a dose of 1 to 2 milligrams per day, and it was recommended to use finer (thinner) lenses with steeper base curve. In the follow-up examination, the patient reported that he had not blinked them out since the lenses were changed. Occasionally, when the lenses were dried and irritated the eyeball, he noticed increased blinking frequency.

The publication of Francisconi et al.<sup>13</sup> describes the case of a 14-year-old boy, in whom Tourette's syndrome was found 6 years earlier. The boy took on permanent haloperidol, clonazepam and clonidine. The damage to the left eye occurred as a result of repeated hitting with no fist. In the examination, with the help of ultrasonography, subluxation of the lens was found with the loss of ciliary processes and the presence of posterior hyaloid detachment. Retinal detachment was not found. A 23 gauge pars plana vitrectomy (PPV) was performed to remove the lens with attached ciliary processes. There were no retinal defects. Due to the patient's inclination to self-harm, the placement of a scleral fixated intraocular lens (IOL) implant has been postponed and contact lenses have been recommended for his aphakia. Issues as proper hygiene, suitable lenses and the importance of regular follow-up were discussed with a patient.

#### Reports in which disorders related to craniofacial injuries

Fassano and Galluccio<sup>14</sup> in 2017 described the case of a 17-year-old boy diagnosed with severe Tourette syndrome. The manifestations of the syndromes consisted of the following symptoms: flexion-extension of his head, sometimes connected with the extension of the

arms. He took low doses of pimozide and oxcarbazepine. After admission to the intensive care unit, his condition was rated on a Glasgow Coma Scale and the score was 5. Physical examination revealed bilateral mydriasis with a poor response to light. MRI showed acute subdural and subarachnoid haemorrhage. Due to massive edema decompressive craniectomy was performed. Six months after the event, MRI in the FLAIR sequence showed necrosis of white matter in the fronto-orbital cortex, diffuse hemispheric hyperintensities and signs of diffuse axonal injury (DAI). After a year, the patient presented symptoms of bradyphrenia, apathy and simple tics of the hand and face. Treatment with risperidone, valproic acid and oxcarbazepine was recommended.

### **Description of the state of knowledge:**

Tourette's syndrome (TS) is often associated with at least one additional, chronic health problem, such as asthma, visual disturbances, musculoskeletal disorders and traumatic brain injury <sup>21</sup>. An especially dangerous complication of TS is traumatic brain injury (intracranial injury) due to the presence of motor tics. For example, we distinguish severe subdural and subarachnoid haemorrhages due to head bumps and rapid neck tics resulting in dissection of the vertebral artery and bilateral cerebellar infarction. There are many causes of disability in children and young adults, but the main one is due to traumatic brain injury. Moreover it should be remembered that this morbidity impacts negatively on the lives of patients and their families, therefore it is important to implement measures that minimize the risk of traumatic brain injury. Intracranial injuries include fracture of the skull, shock and brain contusion, and intracerebral hemorrhage. An important observation is that patients with Tourette Syndrome taking regular antipsychotic drugs compared to non-complying with the recommendations patients are less likely to experience traumatic brain injury. This is due to the direct action of drugs, which suggests significant treatment efficacy.

The explanation of the relationship between Tourette's syndrome and the increased risk of intracranial injury is trivial, because it results from direct damage during movement tics. The intensity of motor tics is mostly low, however, a sudden increase in the frequency of occurrence, in this case uncontrolled movements of the head and neck causing intracranial injury, is possible. However, there is strong evidence from long-term observation that patients with Tourette Syndrome are at increased risk for the above-mentioned injury <sup>21</sup>.

Another aspect is the huge struggle for dentists with TS patients as an often problem during routine control or major treatments. Symptoms of the disease may increase due to stress and anxiety associated with the visit, which is why it is common to use sedation or rarer general anesthesia <sup>16</sup>. Patients with TS aim of dental appointments caries and less trivial injuries resulting from symptoms characteristic of the syndrome – self-mutilation of the tongue and upper lip or extraction of teeth. Various sedative medications are available, among which one should distinguish commonly used benzodiazepines (midazolam, diazepam). Morphine sulphate in combination with droperidol is much less frequently used. It has been observed that benzodiazepines and general anesthesia show high efficacy in suppressing the occurrence of tics, therefore it seems appropriate to suggest a reduction in patient stress and anxiety before and during activities performed at the visit <sup>16</sup>. In patients with TS, cases of tooth self-extraction can be observed. They were caused by strong grinding of teeth or biting (more typical for small children). In order to prevent further incidents, the use of an overlay should be suggested <sup>15</sup>.

Recent years have brought progress in the treatment of TS. The interest in antipsychotics, dopaminergic blockers, drugs not affecting adrenergic transmission and non-neuroleptics, eg

VMAT2 inhibitors (valbenazine) increased significantly <sup>17,18</sup>. In the treatment of TS, not only neuroleptics, both classical and atypical, such as haloperidol or aripiprazole are used, but also drugs from other groups with anxiolytic, sedative, anticonvulsant or sympatholytic activity, i.e. clonidine, topiramate, clonazepam and also botulinum toxin. Treatment with D2 receptor antagonists (haloperidol) carries a high risk of adverse effects including dyskinesias (parkinsonian syndrome), hyperprolactinemia and effects on other receptors, eg. histamine or serotonin, which may lead to weight gain, nausea or sedation. Treatment of tics is, however, only recommended in some situations, including when tics cause self-harm, cause psychological or physical discomfort and contribute to problems in dealing with other people or interfere with the daily functioning of the patient. Pharmacological treatment contributes on average to controlling 25-50% of symptoms <sup>18</sup>. Treatment with TS should not be limited to pharmacological agents, but should also include behavioral therapy. Broadly understood psychotherapy is the primary and obligatory therapeutic treatment <sup>17,19</sup>. It is highly effective and free of side effects, as opposed to pharmacotherapy. The limitation in the use of behavioral therapy results from several causes – the reluctance of patients to attend such meetings and the lack of qualified staff <sup>18</sup>. At the same time, comprehensive behavioral intervention (CBIT) is more effective than supportive psychotherapy <sup>20</sup>. This type of therapy helps patients identify pre-symptomatic factors (tics), which can help to reduce their symptoms and discomfort (habituation process) accompanying the patient when he or she does not perform the tick. At the same time, work in areas such as everyday life of the patient, i.e. contacts with people, discomfort caused by the occurrence of symptoms, low self-esteem or stigmatization (especially in the school-age group) contribute to the improvement of quality of life and tics <sup>19</sup>. People who have been diagnosed with tics can be treated with deep brain stimulation (DBS). In the case of people with TS, the implant is placed in the internal part of the pale knob and the posterior-abdominal, medial-orantero-medial part of the hill. This method has numerous contraindications including severe depression, significant damage to brain structures in the magnetic resonance image (MRI), underweight or tics that can damage the neurostimulator, i.e. compulsive touching the implant site and compressing this area <sup>17,19</sup>. It was shown that in people treated actively for DBS there was a significant decrease in the intensity of tics in comparison to people who were apparently subjected to stimulation <sup>20</sup>. It is a promising method, while its effectiveness is open (further clinical trials are needed), therefore it should be included in the treatment only in the case of ineffectiveness of all other available treatments <sup>19</sup>.

In the case of tics involving lip biting and tongue, a thermoformable overlay is helpful, the task of which is to limit further damage within these structures, because pain can have a self-tapping effect. Any damage involving the oral cavity should be consulted by a qualified dentist who is part of an interdisciplinary team. Dental procedures in persons with TS may contribute to the increase in the intensity of tics resulted from stress, therefore anesthesia consultation is recommended before the procedure. Sedation and general anesthesia significantly reduces the risk of complications associated with symptoms during dental procedures <sup>16</sup>.

### **Summary:**

The analysis of publications presenting case reports of patients with Tourette's syndrome leads to conclusion that patient care requires an interdisciplinary approach. Depending on the intensity and type of tics, neurological, dental, orthopedic, psychological, pediatric or anesthetic consultations are necessary. The main problem is the situation in which tics make it

difficult or impossible for the patient to live in society. The basic method of treatment is psychotherapy. In selected cases, positive results can be achieved by pharmacological treatment.

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