Congenital Trismus

Abstract
Temporomandibular disorders come in many different forms. Congenital trismus is one of these forms that is a newly diagnosed phenomenon in the medical world. This disorder can often be misdiagnosed and confusing to medical professionals. The etiology behind trismus is contributed to a wide range of factors. There are also several signs and symptoms that accompany this condition as well as many complications. Treatment options are still being discovered for this disorder. A case-report of a 6-year-old boy shows the importance of correctly diagnosing and treating trismus.

Introduction
There is a rare condition in which the mouth is unable to open fully. The exact cause behind this condition is unknown and varied symptoms make for a difficult diagnosis. “There are few studies concerning treatment and no standard of care exists.”¹ This condition is known as congenital trismus and it is defined as any restriction to mouth opening or the inability to fully open the mouth.¹⁻³ Trismus is a condition that is frequently overlooked by oncologists, surgeons, and clinicians. Patients with trismus often assume that a reduction in the mobility of their jaw is normal, or that the condition will resolve on its own.² Maximal mouth opening varies greatly from person to person. The opening is dependent on age and gender and can range from 40 to 60 mm, measured between the incisors of the lower and upper jaw.³⁻⁴

Anatomy of Mouth and Face
Congenital trismus affects the structures of mastication including the masseter and pterygoid muscles, the temporomandibular joint (TMJ), nerves that innervate the mouth, and other supportive tissues.¹⁻³ When the muscles and surrounding tissue are damaged, limited mouth opening will occur. This limited mouth opening will lead to muscles atrophying and joint degeneration.² The temporomandibular joint articulates between the condyle of the mandible and the squamous portion of the temporal bone. The condyle is elliptical in shape and the articular surface of the temporal bone is composed of a concave articular fossa and a convex articular
eminence. The meniscus is a fibrous, saddle shaped structure that separates the condyle and the temporal bone. The meniscus is separated into an anterior band and a posterior band. When the TMJ is functioning normally, the condyle and meniscus move together beneath the articular eminence. When the mouth opens in a normal fashion, the condyle may lie beneath the anterior band of the meniscus.

When the meniscus reduces, the patient will often feel a pop or click in the TMJ. The meniscus can remain anteriorly displaced at full mouth opening. This condition is termed anterior displacement without reduction. Patients with anterior displacement without reduction cannot fully open their mouth. Sometimes a tear or perforation of the meniscus is also present. Grinding noises in the TMJ are frequently associated with a TMJ abnormality as well.

Etiology

Trismus is a progressive contraction of the mastication muscles resulting in reduce mobility of the mouth. Trismus can result from various problems, including trauma, surgery, radiation treatment, tetanus, or from TMJ problems. Muscle damage, nerve damage, joint damage, scar tissue, or a combination of these factors can also result in trismus. There are three subcategories of factors that can be attributed to trismus: external factors, internal factors, and iatrogenic factors. External factors include:

- Neoplasms.
- Acute infection.
- Myositis.
- Systemic diseases (lupus, scleroderma, and others).
- Pseudoankylosis.
- Burn injuries.
- Trauma to the muscles or joint.

Internal factors include:

- Bony ankylosis (bony growth within the joint).
- Fibrous ankylosis.
- Arthritis.
- Infections.
- Brusixm (a form of internal trauma).

Iatrogenic factors include:

- Third molar extraction (muscles of mastication torn).
- Joint hyperextended.²

Trismus can also be caused by a tumor that invades the surrounding tissues and causes joint stiffness. In patients with cancer, the combination of radiotherapy and surgery increases the patient’s risk for developing trismus. “Radiotherapy causes fibrosis and contracture within the treatment area from damage to the blood supply; there is evidence that stiffness or fusion of the TMJ occurs.”⁶(p. 33) The degenerative effects in the TMJ could become permanent over time.⁶

**Signs and Symptoms**

Congenital trismus comes with varied signs and symptoms. These symptoms affect the quality of life that the patient is able to achieve. After trismus occurs, it is progressive and long-lasting.⁶ Oral hygiene becomes compromised, chewing and swallowing becomes more difficult therefore increasing the risk of aspiration. Other symptoms include:

- Reduced nutrition intake due to impaired mastication.
- Difficulty in speaking.
- Difficulty in brushing teeth.
- Abnormal facial appearance.²-⁴

In the case of radiation therapy, associated symptoms include headaches, jaw pain and stiffness, ear aches, deafness, and pain on moving the jaw.²-⁴ Patients with congenital trismus can also be very difficult to intubate if needed.⁴ These symptoms often present very difficult and challenging cases.

**Diagnosis and Complications**
Trismus develops very slowly over a long period of time. There is a simple test that helps in diagnosing this condition referred to as the ‘three finger test’. This test is comprised of having the patient insert three fingers into the mouth. If all three fingers fit between the upper and lower incisors, then the mouth opening is considered normal. However, if the fingers do not fit or less than three fingers fit, then restriction is likely. Accurate measurements are vital when diagnosing trismus. If a patient has a mouth opening of 35 mm or less, this is an indicator that the patient has trismus.

There are four major complications associated with limited mouth opening. The first of these complications is eating issues. Restriction in the mouth opening results in only small amounts of food being eaten. Due to the lack of nutrition, patients often experience significant weight loss and nutritional deficits. Along with eating issues comes compromised airway clearance. The tongue is not able to properly move food around the mouth and form a proper bolus; this in combination with the compromised mastication can lead to aspiration. Second, oral hygiene is severely diminished. Poor oral hygiene can result in infection which can lead to further complications that could be fatal. Osteoradionecrosis, a condition where the bone of the mandible dies from radiation or infection, can also result. Third, speech impairment and the inability to create normal sounds are also present. The fourth complication is the most apparent when assessing trismus, the inability to open the mouth due to joint immobilization. If left untreated, degenerative processes will become permanent, muscles will atrophy and the muscle fibers will also become shortened within days.

Imaging

Imaging congenital trismus is mostly done with panoramic x-ray, computed tomography (CT), and magnetic resonance imaging (MRI). The diagnostic procedures that are considered necessary in diagnosing TMJ dysfunctions include: diagnostic x-rays, tomograms, arthograms, CT or MRI scans, cephalograms (x-rays of the jaw and skull), and pantograms (x-rays of the maxilla and mandible). Diagnostic imaging is crucial in the treatment plan for those who have congenital trismus. The following case studies show how computed tomography was used to diagnose trismus with different etiologies.

In the first case study, a 3-year-old boy sustained a mandibular condyle fracture after falling while playing. The fracture was never treated and a year later the patient had increased
trismus along with difficulty eating. A CT scan confirmed the temporomandibular joint
dysfunction and the patient underwent a coronoidectomy and aggressive physiotherapy. In case
the second case, a 22-month-old infant presented with limited interincisal opening. Hospital
records showed that within the first month of life, the patient had sustained a parotid infection
that caused a septic arthritis resulting in craniomandibular dysplasia and micrognathia. A CT
scan revealed that the patient had ankylosis of the temporomandibular joint along with complete
fusion of the mandibular condyle to the cranial base. The patient was treated with ipsilateral
condylectomy, reverse-L mandibular osteotomy, transport distraction osteogenesis, and
coronoidectomy, followed by physiotherapy. The third case study was performed on a premature
newborn infant. The patient was not able to be intubated due to limited mandibular excursion.
The patient required an emergent tracheostomy. When the patient was 9-months-old she was re-
evaluated. Mandibular excursion was limited to an interincisal opening and a craniofacial
computed tomographic scan confirmed bilateral bony ankylosis. Surgical treatment is planned
when the patient turns 3. “Preoperative computed tomographic scanning is recommended to aid
in classification and selection of treatment modalities.”

Magnetic resonance imaging (MRI) is often used for patients who have undergone
radiation therapy to help in displaying abnormal findings in masticator structures. Studies are
being performed using MRI to try and predict the severity of trismus in patients who are
receiving radiation doses. In a study performed by Hsieh et all\(^9\), 22 patients were evaluated for
trismus after receiving radiation therapy to the head or neck. Seventeen patients (77.3\%) had
varying degrees of trismus during a 24-month period after receiving doses of radiation. Several
abnormalities of the masticator muscles were diagnosed with the use of MRI, suggesting that
trismus is a multifactorial disease. The study found that when using a scoring system based off
the MRI images, the severity of trismus could be predicted.\(^9\)

Case Report

There is a form of trismus that is a rare autosomal dominant inherited disorder known as
trismus-pseudocamptodactyly syndrome or Hecht-Beals’ syndrome. This syndrome presents
with “variable expressivity, presenting partial inability to open the mouth and limited extension
of the interphalangic joints while attempting dorsiflexion of the wrists.”\(^10\) The following
case study was performed on a 6-year-old boy from Guatemala who presented with an unknown
form of congenital trismus. The family history was not complete due to the mother being single and the loss of contact between her and the boy’s father, furthermore, the mother’s family had no similar conditions. The initial exam showed that the boy’s maximal mouth opening was 23 mm. The boy also had carious lesions due to his dental hygiene problems that could not be fixed at the time of the examination because of the limited mouth opening. The boy also had difficulty with eating. A panoramic radiograph showed that the cause of the trismus was severe bilateral enlargement of the mandibular coronoid processes (see Figure 1). A 3-D CT was ordered confirming the diagnosis (see Figure 2). The restricted mouth opening was secondary to the overgrowth of the coronoid processes which was interfering with the maxillae. A physical examination showed limited function in performing dorsal flexion of both hands. This restriction was due to shortened finger-flexor-tendons. The boy’s condition was finally diagnosed as trismus-pseudocamptodactyly syndrome (Hecht-Beals’ syndrome.)

In the case presented, the patient underwent intraoral bilateral coronoidectomy with general anesthesia. Post-surgery, a mouth-opening device was placed to prevent a new opening problem from fibrosis or scarring. The boy did obtain a normal mouth opening and no complications were noted 31 months after surgery. The boy also had dental restoration and is currently on orthopedic rehabilitation. As for the inability to dorsal flex the hands, it was determined that there was no interference with normal function and therefore no treatment was necessary. In this particular case, the autosomal dominant inherited trait was never confirmed due to the loss of contact with the boy’s father. The genes that cause this condition have not been identified and analysis of blood markers has failed in showing any linkage to any chromosomes.10

**Treatments**

Early treatment of trismus is vital to helping prevent further complications or minimize those complications that are already present. Passive motion, performed several times per day has been more effective than static stretching. A good stretching practice for patients with trismus is the ‘7-7-7’. Patients will open their mouth seven times with assistance. Maximal mouth opening that does not cause pain will be held for seven seconds. The exercises will be performed seven times per day. The devices used to treat trismus have ranged from cages over the head, heavy springs that fit between the teeth, screws placed between the central incisors,
tongue depressors that are stacked together and placed between the teeth, and hydraulic bulbs placed between the teeth.\textsuperscript{2} Patients may also be referred to a speech and language therapist or physiotherapist to help with treatment.\textsuperscript{3} The most commonly used treatment options for congenital trismus include physical therapy, exercises (see Figure \textit{3})\textsuperscript{6}, as well as devices to help post-surgery. Physical therapy is generally the mainstay treatment and can be used alone or in conjunction with other modalities. Forced mouth-opening during surgery can help improve trismus, but the effect is often short-lived and can lead to potential complications. There are a variety of jaw opening devices that can also be used to treat trismus. Devices that are currently used include stacked tongue depressors, corkscrew devices, TheraBite Jaw Motion Rehabilitation System, and the DTS (see Figure \textit{4}).\textsuperscript{1}

\textit{TheraBite Jaw Motion Rehabilitation System}

The TheraBite provides balanced, gentle stretching to the jaw muscles without using the patient’s own muscles. This is called ‘passive movement’ because the device is allowed to move the muscles which are restricted.\textsuperscript{3} The TheraBite is a plastic device placed in the mouth and then manual force is applied in a lever-aged fashion using plastic handles. This device operates on the principle of high-torque, short duration, and passive stretching. The force used to increase the mouth opening is proportional to how hard the device is squeezed. The TheraBite has demonstrated efficacy in studies done within 6 weeks of surgery for patients diagnosed with oropharyngeal carcinoma. TheraBite has also proven helpful when combined with unassisted exercise for patients who have undergone radiation therapy within 5 years.\textsuperscript{1} In one study performed during a ten week period, a group of patients that had radiation-induced trismus, were studied while using the TheraBite System. At the end of the ten weeks, the group had improved an average of more than 13 mm compared to a group of patients using the tongue depressors that had only improved less than 5 mm. One of the benefits of using the TheraBite is that it not only stretches the connective tissues, but also properly immobilizes the temporomandibular joint.\textsuperscript{2}

\textit{DTS}

The DTS is a commercially available jaw opening device that operates on the principle of low-torque, prolonged duration stretching. The DTS provides a very low level dynamic jaw opening and is used for up to 30 minutes 3 times a day. Once the maximal use schedule is
tolerated, patients may be instructed to adjust the tension to provide increased stretch to prevent rebound spasms of the muscles of mastication. Pilot studies are being performed to determine the efficacy of the DTS among patients with head and neck cancer.¹

**Other Exercises**

Other exercises can be used to help patients regain full jaw motion. These exercises include:

*Springy bites*

- Place the index finger between the top and bottom front teeth with the nail of the finger under the top front teeth.
- Gently bite down on your finger so that the top and bottom teeth are just barely squeezing the index finger.
- Perform this exercise for one minute frequently throughout the day.³

As the exercise becomes easier, the range of motion can be increased by turning the index finger onto the side or by using the flat of your thumb to increase the stretch. Two fingers can also be used between the upper and lower teeth.

*Wooden spatulas*

A speech and language therapist may also have the patient use a wooden spatula that is measured in one millimeter thickness. The therapist will give the patient a recommended number of spatulas to insert between the teeth. This will help to provide a stretch through passive jaw movements.³

**Conclusion**

Congenital trismus is a temporomandibular joint disorder that often presents with limited mouth opening. Congenital trismus is frequently accompanied with confounding symptoms and no working diagnosis that connects the disease with the symptoms that are observed. Due to these factors, it is difficult to formulate an appropriate treatment plan. “This difficulty is compounded when the rarity of the condition results in no guidance from the literature.”¹¹(p. 652)
The exact cause of congenital trismus is unknown. Early treatment of this condition is better for the patient and can greatly improve quality of life. The most important treatment options include daily jaw exercises as well as physiotherapy. Patients with trismus will often have to undergo several surgeries throughout life. This condition is manageable with the appropriate intervention and treatment plan. Knowing that the discovery of this condition is relatively recent, and therefore immature in its diagnosis as well as treatment, offers patients optimism that future innovations will lead to greater success in its treatment.
References


Figure 3. Jaw exercise sheet for patients receiving radiotherapy that were to be performed twice a day, every day until their follow-up appointments. Image courtesy of: Rose T, Leco P, Wilson J. Development of simple daily jaw exercises for patients receiving radical head and neck radiotherapy. Journal of Medical Imaging and Radiation Sciences. 2009;40(1):32-37.
Figure 4. Devices used to treat trismus include (A) stacked tongue depressors, (B) corkscrew devices, (C) the TheraBite Jaw Motion Rehabilitation System, and (D) the DTS. Image courtesy of: Stubblefield MD, Manfield L, Riedel ER. A preliminary report on the efficacy of a dynamic jaw opening device (dynasplint trismus system) as part of the multimodal treatment of trismus in patients with head and neck cancer. Archives of Physical Medicine and Rehabilitation. 2010;91(8):1278-1282.