



Review Article

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Neuromuscular disorders affecting the Oral and Maxillofacial region and their Dental management

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Abstract

A large number of Neuromuscular disorders have their manifestations in the oral and maxillofacial region. Oral physicians come across abnormalities in the dentition, soft tissues and occlusion caused by these disorders during the intra-oral and extra-oral examination. Also, patients with Neuromuscular disorders need to be treated with special precautions during dental treatment. Since a dental practitioner plays an important role in the early diagnosis and management of these conditions, this review aims to provide concise and relevant information about Neuromuscular disorders affecting the Oral and Maxillofacial region and their dental management.

Keywords: Neuromuscular disorders, Maxillofacial region, Dental management.

INTRODUCTION

Neuromuscular disorder literally means caused due to muscle impairment because of pathology in the associated nerves.

These neuromuscular disorders have a collective lifetime prevalence rate of 3% to 5% which makes their encounter common with the dental practitioner.

These disorders affect the dentition, soft tissues and the occlusion and hence we should be aware of their oral manifestations, dental management and treatment.

This chapter focuses on the most common neuromuscular disorder affecting the orofacial region and their management, pertaining to oral physicians.

- CVD
- Multiple sclerosis
- Parkinsonism
- Huntington's disease
- Seizure disorders
- Bell's palsy
- Myasthenia gravis

1. Cerebrovascular Disease or CVA or Stroke

Refers to disorders that cause damage to cerebral blood vessels due to impaired cerebral circulation.

➤ Aetiology-

A. It is caused due to sudden impairment in central leading to death or focal neurologic deficit.

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B. Strokes result from-

Hypertension

Trauma

Substance abuse

Aneurysmal rupture

C. 85% strokes result from ischemia due to atherosclerotic disease, thromboembolic event, occlusion of cerebral vessels.

➤ Clinical features-

It depends on the size and location of the affected brain region. Common signs and symptoms are-

-sensory and motor deficit

-paresis in eye movements

-visual defects

-sudden headache

-dizziness

-nausea

-seizures

-impaired memory

➤ Diagnosis

1. Stroke should be considered whenever a patient experiences the aforementioned clinical features.
2. Other diagnostic methods are MRI brain, non-contrast CT scan
3. Lab investigations include complete blood count, urinalysis, coagulation profile and when indicated blood culture, ECG and lumbar puncture.

➤ Treatment

1. Management of acute stroke includes medical therapy to reduce bleeding or thromboembolic occlusion.
2. Thrombolysis with intravenous tissue plasminogen activator (TPA) from 3-4.5 hours after stroke onset.

➤ Oral health considerations-

1. Following stroke patients may experience oral problems like masticatory and facial muscle paralysis, impaired taste sensation, diminished gag reflex and dysphagic which need to be managed according to the presenting complaint.
2. Maintenance of oral hygiene replacement of missing teeth should be done.
3. Blood pressure should be monitored to prevent stroke and history should be taken about previous episode of stroke.

4. Patients with a history of stroke are usually using aspirin and warfarin, hence use of NSAIDs may increase the risk of bleeding and their long term use may reduce the protective effect of aspirin.

5. Stress reduction during dental visits should be done for which pre operative inhalation N₂O-O₂ or oral anxiolytic can be used.

6. Local anaesthesia containing epinephrine can be used in stroke patients but should be used judiciously.¹

2. Multiple Sclerosis

It is a relapsing remitting autoimmune inflammatory demyelinating disease of the CNS.

➤ Aetiology-

1. It can be caused secondary to trauma.

2. Though the exact cause of M.S. is unknown, genetic susceptibility clearly exists.

3. It is mostly an autoimmune reaction in which major histocompatibility complex (MHC) on chromosome 6p21 has been identified.

4. Infections agents like Epstein Barr virus and human herpes virus 6 are also implicated in the pathogenesis of M.S.

➤ Clinical features-

1. Age of onset of MS is typically between 20-45 years.
2. It is more common among women than men (2:1 ratio).
3. Clinical features of MS depend upon the area of the CNS involved and frequently affected areas include optic chiasma, brainstem, cerebellum and spinal cord.
4. The sudden onset of optic neuritis without any other CNS signs or symptoms could be the first symptom of MS.
5. Diplopia, blurring, nystagmus is also commonly seen.
6. Limb weakness is characteristic of MS and can be manifest as loss of strength, fatigue or gait problems.
7. Ataxia may affect the head and neck of MS patients and may cause cerebellar dysarthria.
8. These patients often show sensory impairment including paraesthesia and hyperesthesia.
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➤ Diagnosis-

1. MS lesions are called 'plaques' which are characterised by perivenular cuffing with inflammatory mononuclear cells, which are seen in the white matter and periventricular area of the CNS.
2. These plaques are visible as hypertense and hypotense areas on T2 and T1 weighted images respectively, suggestive of chronic and active lesion which is diagnostic of MS.

3. Other advanced imaging techniques that are being evaluated for diagnosis of MS are diffusion tensor imaging, magnetization transfer imaging, proton magnetic resonance spectroscopy and functional MRI.

➤ Treatment

1. Glucocorticoids can be used to manage both initial attacks and acute exacerbation of MS.
2. Intravenous methylprednisolone is administered at a dose between 500-1000mg per day for 3-5 days
3. Disease modifying agents include injectable interferon IFN- beta/a, IFN-beta/b, and glatiramer acetate.
4. Mitoxantrone (Novantrone) is a chemotherapeutic agent administered intravenously that is effective in reducing, neurologic disability.
5. Some other common agents used for management of MS are anticonvulsants, benzodiazepines, tricyclic antidepressants, smooth muscle relaxant, anticholinergic agents and analgesics.

➤ Oral health considerations-

1. MS patients can present with trigeminal neuralgia (TN) with possible absence of trigger zones and continuous low intensity pain which should be managed similar to typical TN.
2. Patients may present with neuropathy of the maxillary(V2) and mandibular branch (V3) of the trigeminal nerve which may result in burning, tingling or reduced sensation.
3. Neuropathy of the mental nerve can cause numbness of the lower lip and chin.
4. Facial weakness and paralysis may be seen in MS patient.
5. Dysarthria may be seen as scanning speech in these patients.
6. Elective dental treatment should be avoided in MS patients during acute exacerbation.
7. These patients may require dental treatment in operating room under general anaesthesia.
8. Patients relatives or nurse should be appraised about the importance of daily home care of oral hygiene.²

3. Parkinsonism-

It is a neurodegenerative disorder characterized by rigidity, tremors, brady kinesis and impaired postural reflexed.

➤ Aetiology –

1. In idiopathic parkinsonism, there is dopamine depletion due to degeneration of the dopaminergic nigrostriatal system in the brainstem.
2. This leads to an imbalance of dopamine and acetylcholine which are neuro-transmitters that are normally present in the corpus striatum.

3. Symptoms similar to parkinsonism may also be induced by drugs that cause a reduction of dopamine in the brain, the most common drugs being phenothiazine derivatives.
4. Although, a definite aetiology has not been established, the most likely explanation is that the disease results from a combination of accelerated aging, genetic predisposition, exposure to toxins and abnormality in oxidative mechanisms.

➤ Clinical features-

1. It affects people older than 50 years of age.
2. The 4 cardinal motor signs of parkinsonism are resting tremors, rigidity or stiffness, bradykinesia and postural instability or impaired balance and coordination.
3. 50% of patients with this develop dementia, depression, anxiety, apathy and irritability.
4. Anatomic dysfunction can cause orthostatic hypotension, constipation, urinary frequency and urgency and abnormal sweating.

➤ Diagnosis of parkinsonism-

1. The diagnosis is based on the health history, neurologic examination and response to levodopa therapy.
2. Differentiating classic parkinsonism from a variety of parkinsonian syndromes characterized by motor decline and dementia can be challenging.
3. Anatomic and functional brain imaging, CSF evaluation can be useful to exclude other diagnosis.

➤ Treatment-

1. Dopamine replacement therapy using levodopa combined with carbidopa remains the initial gold standard.
2. Anticholinergics such as scopolamine may help control tremor and rigidity.
3. Dopamine agonists such as bromocriptine, per-golide, pramipexole alone or in combination with levodopa may control parkinsonism.
4. Rivastigmine, a cholinesterase inhibitor, is effective in treating parkinsonism dementia.
5. Clozapine is effective for treating parkinsonism psychosis.
6. Exercise has demonstrated significant benefit in physical conditioning, gait, balance, leg strength and walking speed.

➤ Oral health considerations-

1. Patients experience increased salivation and drooling, making maintenance of dry field difficult for some dental procedures.
2. Angular cheilitis is common in these patients.
3. Use of anticholinergic drugs cause xerostomia which leads to damage to teeth and PDL and also difficulty in

retaining dentures, mucosal ulcerations, denture sores, increased chance of bacterial and fungal infections.

4. Patient loses facial expressions and has slow speech that is soft and fading.
5. Tremor of the head, lips and tongue are common.
6. These patients require periodontal recall every 4-6 months owing to poor oral hygiene.
7. Enameloplasty or mouth guard is required to prevent injury to the tongue due to tardive dyskinesia.
8. Salivary substitutes and topical fluoride application are necessary in patients with xerostomia.
9. During dental procedures, chair should be positioned at 45 degree to limit muscle rigidity and breathing difficulties.

Appointments should be short and relaxing with nitrous oxide sedation which helps reduce stress and prevalence of tremors.³

4. Huntington's disease-

➤ Aetiology-

1. It is hereditary degenerative disease of the central nervous system characterized by chorea (involuntary movements) and dementia.

➤ Clinical features-

1. The earliest manifestation of this disease consists of depression and irritability coupled with slowing of cognition.
2. There are subtle changes in coordinated and minor choreiform movements appear.
3. There could be progressive nonsensing of choreic movements that are observed in the face, tongue and head.
4. In advanced cases, the hyperkinesia becomes aggravated and movement can become violent, with difficulty in speech and swallowing.

➤ Diagnosis-

1. The diagnosis is based on neurological, neuropsychological and psychiatric evaluation.
2. MRI or contrast enhanced CT scan of brain can be done to reveal structural changes at particular sites in the brain.
3. Genetic testing is a confirmatory test for this disease.

➤ Treatment-

1. There is no cure for Huntington's disease, progression cannot be halted and treatment is purely symptomatic.
2. Treatment is usually involving blocking of dopamine receptor such as haloperidol and phenothiazines, which temporarily reduce the hyperkinesia and the behavioural disturbances.

➤ Oral health considerations-

1. Dysphagia and choreic movements of the face and the tongue will make dental treatment quite challenging.
2. Sedation with diazepam can be considered.
3. Whenever possible, dentures for such patients should be avoided because of the danger of fracture or its accidental swallowing.⁴

5. Seizure disorders –

Seizures are paroxysmal disorders of cerebral function characterized by an attack involving changes in the state of consciousness, motor activity or sensory phenomenon.

➤ Aetiology-

1. Isolated, nonrecurrent, generalized seizures among adults are caused by metabolic disturbances, toxins, drug effects, hypotension, hypoglycaemia, uremia, encephalopathy drug overdoses and drug withdrawal.
2. Cerebrovascular disease may account for approximately 50% of new cases of epilepsy in patients older than 65 years.
3. Other aetiologies for epilepsy include degenerative CNS disease, developmental disabilities and familial / genetic factors.

➤ Clinical features-

There are seven types of seizures-

a. Grand mal –

1. It is most common type of seizure which can occur alone or with other types.
2. This seizure begins with aura in which patient experiences epigastric discomfort or hallucinations of hearing, vision or smell.
3. It is followed by the tonic phase in seconds by unconsciousness, cry and tonic muscle spasm.
4. The tonic phase is followed by the clonic phase composed of convulsive jerky movements, incontinence and tongue biting.
5. Jaw is clamped shut and there is foaming at the mouth.

b. Petit mal-

1. These seizures occur exclusively in children and frequently disappear during second decade of life.
2. The patient loses his consciousness, but will continue his normal activity immediately after the seizure.

c. Psychomotor-

1. During the seizure, the patient exhibits purposeless movements and bizarre behaviour.

2. Patient may wander about aimlessly or may exhibit violent behaviour during the seizure.

d. Jacksonian –

1. The seizure begins with clonic movements of a distal portion of extremities or the face.
2. The convulsive movements spread to the limbs, become generalized and causes loss of consciousness.

➤ **Diagnosis-**

1. History and physical examination are critical to diagnose a seizure.
2. Complete neurologic examination is essential which includes the testing of cranial nerve function, assessment of mental status and testing of motor function.
3. Blood studies like complete blood count, electrolytes, glucose, magnesium, calcium are done to identify any metabolic cause of seizure.
4. All the patients should undergo MRI and CT scan for detection of any pathology in the brain responsible for seizures.
5. An EEG (electroencephalogram) is an important tool for classifying seizure disorder.

➤ **Treatment –**

A. Immediate management during seizure-

1. The patient should be moved away from danger like fire, water or sharp objects.
2. After convulsion ceases, the patient should be positioned supine with legs slightly elevated.
3. Basic life support should be given if indicated.
4. If convulsion continues for more than 5 minutes urgent medical help should be summoned.
5. Intravenous anticonvulsant like diazepam should be given.

B. Pharmacologic management-

1. Lamotrigine, carbamazepine and phenytoin are indicated for the treatment for partial seizures.
2. Valproic acid is indicated for the treatment of generalized tonic-clonic seizures.

➤ **Oral health considerations-**

1. These patients have high rate of physical injuries, including dental and facial trauma.
2. Hence, precipitation of seizures during dental treatment should be avoided by reducing psychologic stress. Inhalation sedation with nitrous oxide (upto20%) and oxygen is highly recommended.
3. Phenytoin induced gingival hyperplasia is commonly seen in anterior labial surfaces of the maxillary and mandibular

gingiva. Maintenance of oral hygiene by chlorhexidine mouthwash can reduce the inflammation or surgical reduction can be done if recession.

4. Patients taking AED's(antiepileptic drugs) have marked bone marrow suppression which causes increased chances of infection and prolonged bleeding. Hence, a complete blood count should be done prior to any dental treatment.
5. Aspirin and other NSAIDS should be avoided in patients taking valproic acid and it can possibly cause increased bleeding.⁵

6. Bell's palsy-

It is also called facial paralysis or seventh nerve paralysis.

➤ **Aetiology-**

1. The exact aetiology is not known but viral infection like herpes simplex affecting the facial nerve causes inflammation and hence paralysis of the nerve.
2. Other causes like trauma during dental extraction, surgical procedures like parotidectomy, tumours of the cranial base, parapharyngeal space and infratemporal fossa can lead to seventh nerve palsy.

➤ **Clinical features-**

1. Bell's palsy begins with slight pain around one ear, followed by an abrupt paralysis of the muscles on that side of the face.
2. The eye on the affected side stays open, corner of the mouth drops, and there is drooling.
3. As a result of masseter weakens, food is retained in both the upper and lower buccal and labial fold.
4. The face becomes expressionless and the creases of the forehead are flattened.
5. Due to impaired blinking, corneal ulcerations from foreign bodies can occur.
6. Involvement of the chorda tympani nerve leads to loss of taste perception on the anterior two-thirds of the tongue and reduced salivary secretion.

➤ **Diagnosis-**

1. A thorough history taking and physical examination helps in diagnosing bell's palsy and the extent of the weakness of the facial muscles.
2. Blood tests to detect viral or bacterial infection can diagnose the aetiology for bell's palsy.
3. MRI or CT scan can be advised to rule out any pathology in the brain.

➤ **Treatment-**

1. Spontaneous improvement is generally seen within 6 months in most cases.

2. Combination of acyclovir 400mg 5 times daily with prednisolone (40-60mg daily) for a week is believed to be more effective than steroid alone.
3. Supportive measures like protecting the eye with eye patches, artificial tear substitutes should be given.
4. Injection of botulinum toxin has shown to be effective in treating this disease.
5. Physiotherapy can be advised for the muscles of face.
4. Patient's tongue may be supple and flaccid with bilateral grooves on the dorsal surface due to atrophy of tongue musculature.
5. Palatal muscles may be affected causing patient to have difficulty in elevating soft palate.

➤ Oral health considerations-

1. Patient has drooping corner of mouth due to which there is drooling of saliva. Hence dental treatment becomes difficult.
2. There may be for oral hygiene due to retention of food in the upper and lower buccal and labial folds due to weakness of buccinator.
3. Patient experiences difficulty in speech and mastication.⁶

7. Myasthenia Gravis –

➤ Aetiology-

1. It is a chronic neuromuscular disease caused by autoimmune destruction of the skeletal neuromuscular junction resulting in impaired neurotransmission and muscle weakness
2. In this condition, autoantibodies are produced to the acetylcholine receptors on the motor end plates of muscles, thus binding of acetylcholine is blocked muscle activation is inhibited.
3. The most common autoantibody is anti-acetylcholine receptor(AChR) and less commonly a muscle specific receptor tyrosine kinase(MUSK).

➤ Clinical features-

1. Patients with ocular symptoms like diplopia and /or ptosis.
2. Oropharyngeal, facial and masticatory muscle weakness is common and results in dysphagia, asymmetry and dysarthria.
3. In severe cases, respiratory difficulty arises.

➤ Diagnosis-

1. The clinical examination and history are highly suggestive of myasthenia gravis.
2. Diagnosis is confirmed by a variety of bedside, electrophysiological and immunological tests.
3. The most commonly used immunological test to establish a diagnosis of myasthenia gravis quantifies serum anti-AChR, with a reported sensitivity of 85%.

➤ Treatment-

1. Anticholinesterase drugs such as neostigmine and pyridostigmine bromide increase acetylcholine availability and receptor binding and provide symptomatic benefit.
2. Plasma exchange and high-dose intravenous immunoglobulin can rapidly and temporarily reduce circulating antibodies.
3. Autoantibody production can be reduced using corticosteroids and non-steroid immune suppressants.

➤ Oral health considerations-

1. Patient may have difficulty with prolonged mouth opening and swallowing.
2. Aspiration risks can be high and can be reduced by adequate suction, the use of rubber dam and avoiding bilateral mandibular anaesthetic block.
3. The patient may be at risk for respiratory crisis due to overmedication.
4. Drugs that may affect the neuromuscular junction such as narcotics, tranquilizers and barbiturates should be avoided.
5. Certain antibiotics like tetracycline, streptomycin, sulphonamides and clindamycin can affect neuromuscular activity and should be avoided.^{1,7}

Table 1: Guidelines for oral healthcare of patients with neuromuscular disease-

MEDICAL CONSULTATION	STRESS REDUCTION/ANXIOLYTIC GUIDELINES	CHAIR POSITION GUIDELINES	ANALGESIA GUIDELINES
It should be summoned when-patient's status, severity and level of control is uncertain.	A rapport should be established to reduce stress and anxiety.	Local anaesthesia can be safely used in patients with seizure disorder.	NSAIDs can be used safely in majority of patients with neurologic disease
Patient who has not seen a physician within last year	The type of steroid taken, dose and duration of treatment should be ascertained.	Intraoral anaesthetics should be administered slowly following aspiration	Major concern is bleeding tenderness which is most prevalent in patients with cerebrovascular disease who take anticoagulants.
Corticosteroids have been taken within last 12 months	Patient should obtain proper rest the night before treatment and should reduce work on the day of treatment.	In patients with impaired mental capacity, lip biting after LA administration is a common postoperative problem.	Respiratory depressant narcotics should be used with caution in patients with neurologic respiratory difficulties

Medications and dosage are not well known by the patient	Appointment should be in the morning and kept short		
Need for additional medication or change in medication during dental treatment	Benzodiazepines, which have minor CNS depressant activity, are the sedative/anxiolytic drugs of choice.		
There are plans for stressful or complex oral surgical procedures.			
Iatrogenic problem arises from dental treatment eg. Bell's palsy that persists more than 6 hours after dental injection.			
ANTIBIOTIC GUIDELINES	AIRWAY MAINTENANCE AND GASEOUS ADMINISTRATION	DRUG EFFECTS AND INTERACTIONS GUIDELINES	INFECTION CONTROL GUIDELINES
Culture and sensitivity testing should be done whenever oral infection is present.	Rubber dam is recommended for routine operative procedures in patients with neurologic disease especially in patients with drooling saliva.	Valproic acid inhibits platelet aggregation and can produce palatal petechiae and abnormal bleeding.	Antibiotic prophylaxis and oral antibiotic rinses should be used when the patient is severely immunocompromised.
Oral penicillin can be used safely as long as the patient is not hypertensive to it.	Nitrous oxide therapy is an excellent anxiolytic.	Carbamazepine depresses bone marrow leading to bleeding tenderness and increased incidence of oral infections.	Contact with blood, saliva and aerosol should be minimized.
Broad spectrum antibiotics should be prescribed with caution in epileptic women who use oral contraceptives.		Primidone and phenobarbital cause ataxia and drowsiness. Additional sedatives and respiratory depressants should be avoided.	Routine aseptic protocol should be followed.
In patients with myasthenia gravis, aminoglycosides who have a neuromuscular blocking effect, should be avoided.		Patient with history corticosteroid therapy may have adrenal suppression. Consultation with the physician is mandatory	

Conflict of Interest

The author reports no conflicts of interest.

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