



REVIEW ARTICLE

**NEUROMUSCULAR DISORDERS AFFECTING THE ORAL AND MAXILLOFACIAL REGION:
INSIGHTS FROM A NARRATIVE REVIEW**

Manda Jasmine¹, Badari Ramakrishna Botu², Rahul Marshal Vaddeswarapu³, Kosuri Venkata Lokesh⁴, Anupama Pottam⁵, Srinivasa Chakravarthi⁶

¹Department of Oral Medicine and Radiology Anil Neerukonda Institute of Dental Sciences Visakhapatnam, Andhra Pradesh, India, drjasminemanda@gmail.com

²Professor and Head of the department Department of Oral Medicine and Radiology Anil Neerukonda Institute of Dental Sciences Visakhapatnam, Andhra Pradesh, India, badari.botu@gmail.com

³Professor Department of Oral Medicine and Radiology Anil Neerukonda Institute of Dental Sciences Visakhapatnam, Andhra Pradesh, India vmars002@gmail.com

⁴Associate Professor Department of Oral Medicine and Radiology Anil Neerukonda Institute of Dental Sciences Visakhapatnam, Andhra Pradesh, India, lokesh.kosuri123@gmail.com

⁵Assistant Professor Department of Oral Medicine and Radiology Anil Neerukonda Institute of Dental Sciences Visakhapatnam, Andhra Pradesh, India dranupamabaratham@gmail.com

⁶Department of Oral Medicine and Radiology Anil Neerukonda Institute of Dental Sciences Visakhapatnam, Andhra Pradesh, India, dr.dentoplus@gmail.com

***Corresponding Author** Professor, Dr. Rahul Marshal Vaddeswarapu Department of Oral Medicine and Radiology Anil Neerukonda Institute of Dental Sciences Dr.NTR University of Health Sciences, Visakhapatnam-531162, Andhra Pradesh, India, +919848720046, vmars002@gmail.com

ABSTRACT

Background: Neuromuscular diseases are a group of disorders affecting the body's muscles and nerves, often leading to muscle weakness, which is the most common clinical feature. These conditions interfere with the body's adaptive systems, reducing overall functional capacity.

Objectives: This article aims to provide a brief review of some common neuromuscular disorders, highlighting their clinical manifestations and their implications for oral health, particularly within the oral and maxillofacial region.

Results: Neuromuscular disorders present with varying degrees of muscle weakness and dysfunction, which can significantly affect oral and maxillofacial structures. These may include difficulties in mastication, swallowing, speech, and maintaining oral hygiene, thereby increasing the risk of oral health complications.

Conclusion: Understanding the manifestations of neuromuscular diseases is essential for early recognition and comprehensive management in dental practice. Special attention is required to address oral health challenges in affected patients, ensuring improved quality of care.

Keywords: Motor neuron disease; Muscular diseases; Muscle weakness; Oral and Maxillofacial region; Muscle Atrophy

INTRODUCTION

The terms 'muscle disease', 'muscular dystrophy', 'neuromuscular conditions', and 'neuromuscular disorders' all describe a large group of conditions that affect either the muscles, such as those in the arms and legs or the heart and lungs, or the nerves which control the muscles. They can cause problems with:

- The nerves that control the muscles
- Muscles
- Communication between the nerves and muscles.¹

These disorders result in longstanding functional deficits that result in substantial utilization of healthcare resources in addition to the emotional, financial and social burden to those affected individuals and their families. NMDs of the maxillofacial region also affect the aesthetic facial appearance of the patients. Some forms of NMD are hereditary and manifest at birth or early childhood whereas the incidence of other forms increases with age due to spontaneous mutation in the gene or due to immune system disorders.¹

Neuromuscular disorders 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 dentition, soft tissues, and occlusion and hence we should be aware of their oral manifestations, dental management, and treatment. This focuses on some of the most common neuromuscular disorder affecting the orofacial region and its management, by oral physicians.²

- CVD (Cerebrovascular Disease)
- Multiple sclerosis
- Parkinsonism
- Huntington's disease
- Seizure disorders
- Bell's palsy
- Myasthenia Gravis

CEREBROVASCULAR DISEASE (CVD)

Cerebrovascular disease includes all disorders that cause damage to the blood vessels supplying the brain, producing neurologic damage.³ The most common form is ischemic heart disease, a fatal condition that can lead to acute myocardial infarction.²³

Etiology:

Approximately 80% of strokes are associated with the development of atherosclerosis leading to cerebral ischemia and infarction and the remaining 20% of cases are caused by cerebral hemorrhage.³

Clinical manifestations and treatment of stroke:

- Strokes due to ischemia may be classified clinically either as a stroke in evolution or as a completed stroke. "Stroke in evolution" is a descriptive term used to indicate condition in which symptoms associated with cerebral ischemia become progressively worse while the patient is under observation.³
- Symptoms of complete stroke caused by an embolus develop suddenly.³
- It is not preceded by transient ischemic attacks; rather, the stroke itself evolves rapidly because the clot originates elsewhere and suddenly blocks a cerebral vessel. Whatever the cause of the neurologic damage, the resultant infarct may enlarge for a period of 4 to 5 days because of cerebral edema.³
- Repair is dependent on good collateral circulation and is accomplished by the formation of fibroblastic scar tissue.³

After a completed stroke, treatment focuses on the prevention of further neurologic damage, through the reduction of underlying risk factors and by rehabilitation procedures, including speech and physical therapy.³

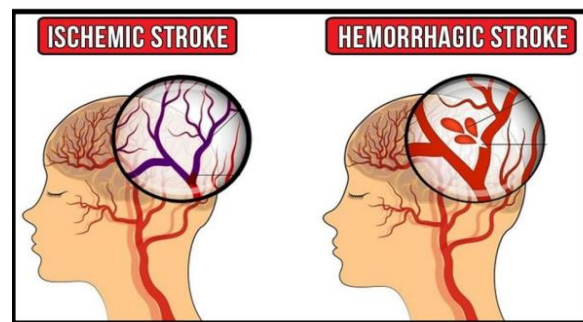


Figure 1. Types of cerebrovascular stroke²⁵

MULTIPLE SCLEROSIS

Multiple sclerosis (MS) is a chronic neurological disease characterized by the demyelination of axons within the central nervous system. The disease occurs more frequently among women.⁴

Etiology: An immunologic basis is strongly suggested by the presence of activated T lymphocytes and autoantibodies to glycoproteins detected in MS lesions.⁴

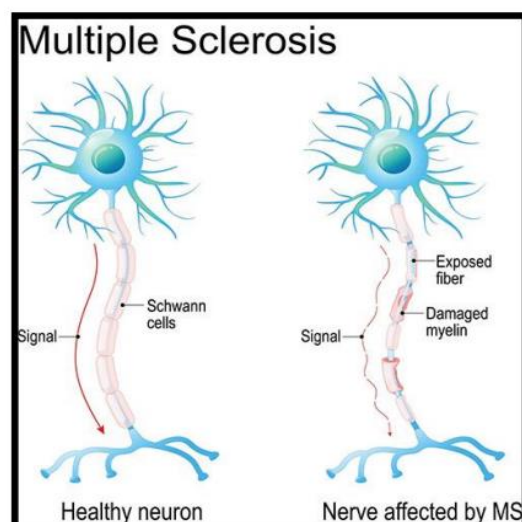


Figure 2. Healthy neuron and nerve affected by multiple sclerosis²⁶

Clinical Manifestations

- Uhthoff's sign is characterized by rapid vision loss following a body temperature increase that is associated with strenuous exercise.⁵
- Another important sign of ocular disturbance is Marcus Gunn's pupillary sign, which can be elicited in patients with unilateral optic neuritis in the following manner: a bright light is shone into each eye separately; when this light is moved from the normal to the affected eye, the pupil of the latter dilates rather than constricts.⁵
- Weakness or paresthesia of the extremities is noted.⁵
- An important feature of motor nerve function is the relative fluctuation of symptoms daily.⁵

Pharmacotherapy of multiple sclerosis:

Monoclonal antibodies:

Natalizumab is given as a 300 mg IV infusion over one hour, every four weeks, Rituximab, Ofatumumab is given by subcutaneous injection, starting with 20 mg administered at weeks zero, one, and two.⁶

Oral therapies:

Diroximel fumarate: The starting dose for oral dimethyl fumarate is 120 mg given twice daily. After seven days, the dose should be increased to 240 mg given twice daily. It is available in 120 and 240 mg preparations.⁶

Oral Health Considerations

- Pain is normally severe and lancinating, but trigger zones may be absent. In time, the pain often becomes less severe but more continuous.⁶
- Neuropathy to the mental nerve can cause numbness of the lower lip and chin. Facial paralysis appears later in the course of the disease.⁶
- It may be difficult to distinguish between the paralysis caused by MS and that due to Bell's palsy, but up to 24% of MS sufferers may experience facial paralysis.⁶

PARKINSONISM: PARKINSON'S DISEASE

Parkinsonism is a neurodegenerative disorder characterized by rigidity, tremors, bradykinesia, and impaired postural reflexes.⁷ Parkinson's disease is a progressive neurodegenerative condition linked with α -synuclein deposition.²⁴

Etiology and Pathogenesis:

In idiopathic parkinsonism, dopamine depletion due to degeneration of the dopaminergic nigrostriatal system in the brainstem leads to an imbalance of dopamine and acetylcholine, neurotransmitters that are normally present in the corpus striatum.⁷

Clinical Manifestations: Tremor, rigidity, bradykinesia, and postural instability are the cardinal features of parkinsonism and may be present in any combination.⁸

Treatment:

Pharmacotherapy of Parkinson's disease: Pramipexole – Pramipexole IR is usually started at 0.125 mg three times a day. The dose should be increased gradually by 0.125 mg per dose every five to seven days.⁸



Figure 3. Parkinsonism Symptoms

Ropinirole and Rotigotine are other drug of choice.

Oral Health Considerations

- Patients must often be treated in the upright position, making access to certain areas of the oral cavity difficult for the dentist.⁹
- In addition, anxiety in a Parkinson's disease patient can increase both the tremor and the degree of muscle rigidity.⁹
- Due to dysphagia and an altered gag reflex, special precautions must be taken to avoid the aspiration of water or materials used during dental procedures.⁹
- In patients who suffer from hypersialorrhea, maintaining a dry field in procedures that require such can be especially difficult.⁹
- Xerostomia is a common side effect of anti-parkinsonism medications; the consequent root caries and recurrent decay must be diligently treated.⁹

HUNTINGTON'S DISEASE (HUNTINGTON'S CHOREA):

Huntington's disease is a hereditary degenerative disease of the central nervous system, characterized by chorea (involuntary movements) and dementia.¹⁰

Clinical Manifestations

The clinical manifestations include declination of cognitive behaviour, mood swings, involuntary movements, speech difficulties.

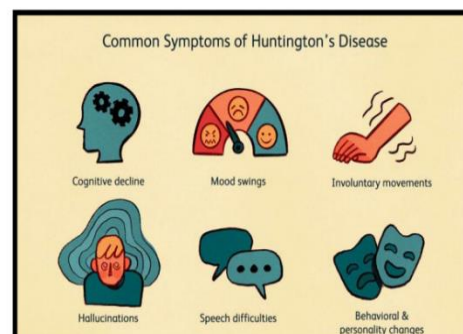


Figure 4. Common symptoms of Huntington's disease²

Treatment: Pharmacotherapy: Tetrabenazine, Deutetrabenazine, Antipsychotics, Benzodiazepines.¹¹

Oral Health Considerations:

- Dysphagia and choreic movement of the face and tongue will make dental treatment especially challenging.¹¹
- Sedation with diazepam may be considered.¹¹
- Whenever possible, dentures should be avoided because of the danger of fracture or the accidental swallowing of the dentures.¹¹

EPILEPSY:

Epilepsy is a condition characterized by abnormal, recurrent, and excessive neuronal

discharges precipitated by many different disturbances within the central nervous system.¹²

Etiology: Epilepsy in older adults may occur as a complication of any of the previously mentioned causes but is more often associated with cerebrovascular diseases such as stroke and tumor.¹²

Clinical Signs

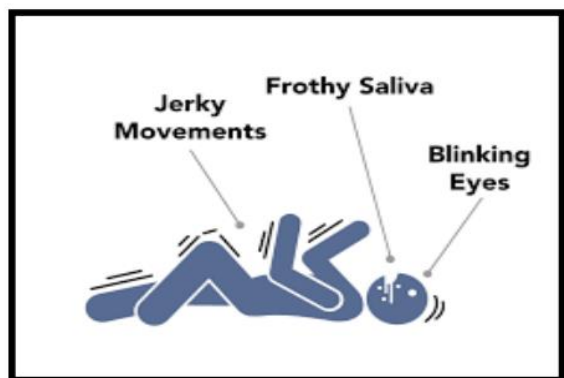


Figure 5. Signs of epilepsy²⁷

DISCUSSION

Treatment

A. Immediate management during a seizure:

- The patient should be moved away from dangers like fire, water, or sharp objects.¹³
- After convulsion seizes, the patient should be positioned supine with legs slightly elevated.¹³
- Basic life support should be given if indicated.¹³
- If convulsion continues for more than 5 minutes urgent medical help should be summoned.¹³
- Intravenous anticonvulsants like diazepam should be given.¹³

B. Pharmacologic management

1. Lamotrigine, carbamazepine, and phenytoin are indicated for the treatment of partial seizures.¹³

2. Valproic acid is indicated for the treatment of generalized tonic-clonic seizures.¹³

Oral health considerations:

1. These patients have a high rate of physical injuries, including dental and facial trauma.¹²
2. Hence, precipitation of seizures during dental treatment should be avoided by reducing psychological stress. Inhalation sedation with nitrous oxide (upto20%) and oxygen is highly recommended.¹²
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 occurs.¹²

BELL'S PALSY:

It is also called facial paralysis or seventh nerve paralysis.

Etiology: This is associated with edema of nerve and subsequent mechanical compression of the facial nerve, leading to paralysis or weakness in the distribution of the facial nerve.¹⁹ Other etiologic conditions include herpes simplex viral infection, parotidectomy, tumors of the cranial base, parapharyngeal space, and infratemporal fossa can lead to seventh nerve palsy.^{14,15,16}

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.¹⁷
2. The eye on the affected side stays open, the corner of the mouth drops, and there is drooling.¹⁷
3. The face becomes expressionless and the creases of the forehead are flattened.¹⁷

Oral health considerations

1. The patient has a dropping corner of the mouth due to which there is drooling of saliva. Hence dental treatment becomes difficult.¹⁸
2. There may be oral hygiene due to retention of food in the upper and lower buccal and labial folds due to weakness of the buccinator.¹⁸
3. Patient experiences difficulty in speech and mastication.¹⁸

MYASTHENIA GRAVIS: A chronic autoimmune disorder causing weakness of skeletal muscle.²⁰

Etiology: it involves loss of acetylcholine receptor (AChR) function on the muscle membrane.²⁰

Clinical features

1. Patients with ocular symptoms like diplopia and /or ptosis.
2. Oropharyngeal, facial, and masticatory muscle weakness is common, resulting in dysphagia, asymmetry, and dysarthria.
3. In severe cases, respiratory difficulty arises.²¹

Oral health considerations

1. The 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 a rubber dam, and avoiding bilateral mandibular anesthetic block.
3. The patient may be at risk for a respiratory crisis due to overmedication.
4. Drugs that may affect the neuromuscular junction such as narcotics, tranquilizers, and barbiturates should be avoided.²²

CONCLUSION

NMD cases are characterized by progressive muscular impairment leading to loss of mobility, being wheelchair-bound, swallowing difficulties, respiratory muscle weakness, death from respiratory failure, and other symptoms.

Neuromuscular diseases are classically divided by localization into disorders that involve cranial and spinal motor neurons, spinal nerve roots, nerve plexuses, peripheral nerves, neuromuscular junctions, and muscles. The evaluation of cerebrospinal fluid (CSF) composition is not routinely needed to help facilitate a diagnosis of a suspected neuromuscular disease. Rather, electromyography, nerve conduction studies, and nerve and muscle biopsies are often of greater diagnostic importance.

DECLARATIONS

Acknowledgments

We thank everyone who supported and contributed to this study.

Funding

This research did not receive any specific grant or financial support from funding agencies in the public, commercial, or not-for-profit sectors.

Competing Interests

The authors have no competing interests to declare.

Ethical Approval

The study was approved by the appropriate ethics committee and conducted according to relevant guidelines and regulations.

Informed Consent

Not applicable.

REFERENCES

1. Potikanond, S., et al. (2018). "Muscular Dystrophy Model." *Adv Exp Med Biol* 1076:147- 172.
2. Quinn N, Schrag A. Huntington's disease and other chores. *J Neurol* 1998;245(11):709-16.
3. Adams HP Jr, Brott TG, Furlan AJ, et al. Guidelines for thrombolytic therapy for acute stroke: a supplement to the guidelines for the management of patients with acute ischemic stroke. *Stroke* 1996; 27:1711–8.
4. 19. Sadovnick AD, Ebers GC. Genetics in multiple sclerosis. *Neurol Clin* 1995;13:99-118.
5. Mumford CJ, Wood NW, Kellar-Wood H, et al. The British Isles survey of multiple sclerosis in twins. *Neurology* 1994;44:11–5.
6. Rudick RA, Cohen JA, Weinstock-Gutman B, et al. Management of multiple sclerosis. *N Engl J Med* 1997 337:1604–11.
7. Lang AE, Lozano AM. Parkinson's disease. Part 1. *N Engl J Med* 1998;339:1044–53.
8. Lang AE, Lozano AM. Parkinson's disease Part 2. *N Engl J Med* 1998;339:1130–43.
9. Olanow CW, Koller WC. An algorithm for the management of Parkinson's disease: treatment guidelines. *Neurology* 1998;50: S1–57.
10. Quinn N, Schrag A. Huntington's disease and other chores. *J Neurol* 1998;245(11):709-16.
11. Kieser J, Jones G, Borlase G. Dental treatment of patients with neurodegenerative disease. *N Z Dent J* 1999;95(422):130-4.
12. Browne TR, Holmes GL. Epilepsy. *N Engl J Med*

2001; 344(15):1145–51

13. Bradie MJ, Dichter MA. Antiepileptic drugs. *N Engl J Med* 1996;334:168–75.

14. Murakami S, Mizobuchi M, Nakashiro Y, et al. Bell's palsy and herpes virus: identification of viral DNA in endoneurial fluid and muscle. *Ann Intern Med* 1996;124:27–30.

15. Sanchez Rodriguez A. Bell's palsy in association with herpes simplex virus infection. *Arch Intern Med* 1998;158(14):1577–8.