Neuromuscular Disorders

أ. د. تغريد فاضل

Definition

Are diseases that affect both nerve and muscle tissue .Neuromuscular disorders represent a spectrum of nerve-related diseases and conditions that affect the body's voluntary muscles. Causes weakening of muscles in the body because of interrupted communication between the nervous system and the muscles it controls. Typically, these diseases can be managed to improve quality and length of life, but are incurable.

Classification of neuromuscular disorders:

- CEREBROVASCULAR DISEASE
- MULTIPLE SCLEROSIS
- ALZHEIMER'S DISEASE
- SEIZURE DISORDERS
- PARKINSON DISEASE
- MYASTHENIA GRAVIS

Cerebrovascular disease:-

Cerebrovascular disease includes all disorders that cause damage to the blood vessels supplying the brain, leading to impaired cerebral circulation thereby producing neurologic damage.

"complete Stroke" and "cerebrovascular accident" (CVA) is a sudden impairment in cerebral circulation resulting in death or a focal neurologic deficit lasting more than 24 hours, are terms used to describe an acute neurologic injury resulting from a severe interruption in the flow of blood to the brain.

Complete cessation of the flow may render an irreversible cerebral infarct within a period of 3 or 4 minutes.

Neurologic events related to CVA include:

Transient ischemic attack (TIA):- defined as reversible, acute, short-duration, focal neurologic deficit ("mini stroke") resulting from transient (reversible within 24 hours) and localized cerebral ischemia

Reversible ischemic neurologic defect (RIND):- defined as reversible, acute, focal neurologic deficit due to transient and localized cerebral ischemia but resulting in neurologic deficits that last more than 24 hours

Symptoms of cerebrovascular disease

Clinical Manifestations

The clinical manifestations of stroke vary depending on the size and location of the affected brain region. The most common signs and symptoms include sensory and

motor deficits, changes (paresis) in extraocular muscles and eye movements, visual defects, sudden headache, altered mental status, dizziness, nausea, seizures, impaired speech or hearing, and neurocognitive deficits such as impaired memory, reasoning, and concentration .

General symptoms following stroke:-

- variable motor paralysis
- sensory loss
- visual difficulties
- speech impairment

Types of cerebrovascular diseases

Cerebrovascular Accident (CVA) or Stroke either due to:-

1- atherosclerosis(85%) leading to cerebral ischemia and infarction result from ischemia due to atherosclerotic disease, thromboembolic events, and occlusion of cerebral blood vessels, with neurologic deficits related to the loss of neural function in tissues distal to the event.

2- Cerebral hemorrhage (15%) result from hemorrhagic events leading to infarction, most often related to hypertension, trauma, substance abuse, or aneurysmal rupture.

Three major types of ischemic stroke syndromes have been described:

1- small vessel (lacunar),
2- large vessel (cerebral infarction)
3- Brainstem stroke

Lacunar strokes:- result from obstruction of the small (<5 mm diameter) penetrating arterioles.

Age and uncontrolled hypertension are the greatest predisposing factors. Symptoms usually include unilateral motor or sensory deficit without visual field changes or disturbances of consciousness or language. The prognosis for recovery from lacunar infarction is fair to good, with partial or complete resolution usually occurring over four to six weeks.

Cerebral infarction (large vessel):- is characterized by extensive downstream ischemia, usually due to a thromboembolic event along the distribution of the internal carotid artery and cerebral arteries. Emboli often originate from the heart after acute myocardial infarction or in hyperdynamic conditions such as chronic atrial fibrillation.

Hypertension is an important risk factor in the development of thrombosis, particularly at the carotid bifurcation, and treatment of severe hypertension is essential for the prevention of stroke. High level brain functions are affected, and the prognosis is poor.

Brainstem infarction :- results from occlusion of small or large vessels supplying the brainstem, resulting in variable deficits ranging from motor and sensory deficits to death when respiratory centers are affected.

Transient lschemic attack

A transient ischemic attack (TIA) is a sudden but reversible neurologic deficit that lasts from a few minutes to 24 hours. Approximately 30% of individuals with a history of TIA experience a completed stroke within a 5-year period. An important cause of transient cerebral ischemia is embolization a source is readily apparent in the heart or a major extracranial artery to the head.

Clinical manifestations

The symptoms of TIAs vary markedly among patients. Onset is abrupt and without warning, and recovery usually occurs rapidly, often within a few minutes.

During the attack, a wide variety of neurologic signs and symptoms can develop, depending on which site of the brain is affected by ischemia.

1- Repeated short periods of arm and hand weakness are associated with focal ischemia in the contralateral frontal lobe.

2- If the vertebrobasilar arterial system is involved, short episodes of dizziness, diplopia, dysarthria, facial paresthesia, and headache are common symptoms.

Treatment of TIA

Treatment of TIAs should be initiated as soon as the diagnosis is established and should be directed towards the:-

1- Correction of the immediate pathologic problem (e.g., embolism).

2- Measures to control the primary underlying problem (e.g., hypertension or coagulopathy).

3- Anticoagulant therapy with either heparin or coumadin is often used.

4- Treatment with aspirin, however, significantly reduces the frequency of TIAs and the incidence of stroke in high-risk patients.

D.D. of CVA

Seizures, hypoglycemia, intracranial tumors, trauma, infection, encephalitis, multiple sclerosis (MS), and prolonged migrainous Aura.

Diagnosis

In addition to a thorough neurologic and cardiovascular examination, anatomic and functional brain imaging is central to the diagnosis of stroke. Time is of the essence for instituting treatment to manage acute stroke. Intracranial hemorrhage must be quickly excluded before life-saving thrombolytic therapy can begin. Although brain magnetic resonance imaging (MRI) provides greater anatomic detail and sensitivity for detection of early infarction, non contrast computed tomography (CT) scan is the first line of imaging.

Laboratory evaluation of the stroke patient includes compete blood count, comprehensive metabolic panel, urinalysis, coagulation profile, and, when indicated, blood culture, echocardiography, and lumbar puncture.

Treatment in general:-

The outcome of stroke and related TIAs and RIND is significantly affected by the timeliness of treatment. Early intervention is critical to prevention, treatment, and recovery.

TIAs and RIND are treated by reduction in hypertension (lifestyle changes such as diet, exercise, smoking cessation, and stress reduction; medical therapy for hypertension; and anticoagulant or antiplatelet medications).

Management of acute stroke includes medical therapy to reduce bleeding or thromboembolic occlusion, medical therapy to reduce brain edema and neurotoxicity/nerve injury, and surgical interventions (revascularization, hemorrhage control).

Once intracranial hemorrhage has been excluded as the source of acute cerebral ischemia, thrombolysis with intravenous tissue plasminogen activator (t-PA) can improve reperfusion, minimize infarction, and reduce disability.

After a completed stroke, treatment focuses on:

1. the prevention of further neurological damage, through the reduction of underlying risk factors
2. Rehabilitation procedures, including speech and physical therapy.
3. An intracranial hemorrhage should also be treated as a medical emergency of airway maintenance and requires the transfer of the patient to an intensive care unit with close monitoring.
4. The surgical treatment of a hemorrhaging aneurysm consists of closing off the blood vessels that supply the area and removing the abnormality.

Oral Health Considerations

Following stroke, patients may experience several oral problems, including masticatory and facial muscle paralysis, impaired or lost touch and taste sensation, diminished protective gag reflex, and dysphagia. These problems can lead to impairment of food intake, poor nutrition, and weight loss due to diminished taste satisfaction, chewing capacity, and swallowing; choking; and gagging.

Diminished motor function of masticatory and facial muscles may also reduce food clearance from the mouth and teeth with the presence of diminished dexterity of the arms or hands may adversely affect oral hygiene and increase the risk for caries and periodontal disease.

Prior history of TIA or stroke increases the risk of a future or second stroke, with the highest risk during the first 90 Days. With optimal medical monitoring and poststroke

care patients can safely undergo invasive dental treatment, with appropriate consideration for stress reduction, medication interactions and adverse effects, neurologic deficit management, and control of underlying cardiovascular/ cerbrovascular risk factors.

Use of antiplatelet and anticoagulant medications is common in patients with a history of stroke, TIA, and RIND. This includes oral aspirin; oral antiplatelet drugs such as subcutaneous low-molecular- weight heparin, and, less commonly, warfarin. These medications taken in therapeutic dosages, and for warfarin with an international normalized ratio ≤ 3.5, rarely require dose modification before routine dental and minor oral surgical treatment.

Concomitant use of non-steroidal anti-inflammatory drugs (NSAIDs) may increase the risk for bleeding, and their long-term use may reduce the protective effect of aspirin.
Stress reduction and confidence building for the patient during dental visits are important behavioral goals to make the patient comfortable and minimize anxiety-related elevation in blood pressure.
Pre- or perioperative inhalation- N2O-O2 or oral anxiolytic medication can aid in reducing treatment-related stress and anxiety.
Use of epinephrine-containing local anesthetics is not contraindicated, but they should be used judiciously and follow guidelines recommended for patients with cardiovascular disease; epinephrine-containing impression cord should not be used. Blood pressure should be monitored at every visit

Important points of Oral Health Considerations:-

1- As the first line of medical management of stroke patients is often on anticoagulant therapy, the patient may have a predisposition to excessive bleeding. It may be necessary to confer with the patient's physician to obtain current coagulation values (PT, INR) so as to ensure that the patient is stable for more invasive dental treatment.
2-Xerostomia is a common side effect of the medications used in the management of cerebrovascular disease. Patients can then be susceptible to a higher caries rate.
3- Stroke patients have physical disabilities, which can affect the orofacial area.
4- Patients with weakness in the muscles of the orofacial area may have poor control of oral secretions, a reduced gag reflex, and changes in their ability to masticate, leading to poor nutrition.
5- Patients with apraxia affecting the orofacial region may have impaired voluntary movements, such as protruding the tongue, expectorating.
6- Careful history taking, checking of blood pressure prior to treatment, avoidance of lengthy appointments.

Cavernous sinus thrombosis

Cavernous sinus thrombosis, usually secondary to dental, nasal, or ocular infections, is a rare but severe complication because of its possible fatal outcome.
Infections of the maxillary dentition may spread to the cavernous sinus through openings in the cranial bones or through emissary veins connecting the extra- and intracranial systems.
Venous propagation begins with the facial vein and proceeds through the ophthalmic vein, which is an affluent of the cavernous sinus.

Signs and symptoms of cavernous sinus thrombosis

- Severe headache often accompanied by tearing
- Swelling, redness, or irritation around one or both eyes
- Drooping eyelids and inability to move the eye
- High fever
- Pain or numbness around the face or eyes
- Fatigue
- Vision loss or double vision
- Seizures
- Altered mental status that can range from confusion to coma

* seizures are rare.

In most cases, patients experience rapid swelling of the face and eyelids.
The classic neurologic signs of acute cavernous sinus thrombosis are:-
Exophthalmos, periorbital edema, retinal vein thrombosis, and involvement of the ophthalmic division of the trigeminal nerve , trochlear and abducent nerves, leading to ptosis, dilated pupils, and lack of corneal reflexes.

Treatment of cavernous sinus thrombosis:

Treatment consists of immediate antibiotic therapy and the removal of the source of infection whenever possible.
Staphylococcus aureus is the most common pathogen, identified in approximately 70% of cases and is the pathogen implicated in nearly all cases of facial infections and sphenoid sinusitis. Streptococci are cultured less commonly. Anaerobes are found occasionally, especially with sinus, dental, or tonsillar infections. Rarely, fungal infections from Aspergillus fumigatus or mucormycosis have been implicated.
Therefore, for most etiologies, empiric therapy should include:

- Vancomycin used until the actual culture results are available, plus
- A third-generation cephalosporin, such as ceftriaxone. In patients with documented true allergy to penicillin, a fluoroquinolone should be used instead.

- Intravenous metronidazole should be added if dental or sinus infection is suspected. Antifungal therapy has been advocated only in cases of biopsy-confirmed invasive fungal infection. However, in at-risk patients, antifungal treatment should be considered as fungi may cause devastating neurological complications beyond cerebral venous thrombosis.

High doses of intravenous antibiotics are required because thrombus may limit penetration of antibiotics. Bacteria, sequestered within the thrombus, may not be killed until the dural sinuses have started to recanalize.

Antibiotics also need to be administered over an extended period, for at least 2 weeks beyond the time of clinical resolution. This aims to insure complete sterilization and prevent relapses.

Concurrent supportive therapy is necessary alongside antibiotic treatment, and includes resuscitation, oxygen support, and local eye care.

Multiple sclerosis (MS)

Is a chronic neurologic disease characterized by multiple areas of central nervous system (CNS) white matter inflammation, demyelination, and gliosis (scarring). Myelin is critical for propagation of nerve impulses, and when it is destroyed in MS, slowing and/or complete block of impulse propagation is manifested by abnormal muscular and neurologic signs and symptoms, associated with the myelination of axons within the central nervous system.

The disease occurs more frequently among women. The average age of onset is during the fourth decade of life, but MS may occur at any age.

The disease presents in the form of recurrent attacks

Etiology

1- An immunologic (autoimmune disease) basis is strongly suggested by the presence of activated T lymphocytes and autoantibodies to glycoproteins detected in MS lesions.

2- Environmental exposure in MS , and two common infectious agents to be implicated in the pathogenesis of this disease are Epstein–Barr virus and human herpesvirus 6. Other viruses that have been implicated in the pathogenesis of MS include measles, mumps, rubella, parainfluenza, vaccin, and human T-lymphotropic virus

3- increased antibody titers against measles virus, rubella virus, mumps virus, Epstein-Barr virus, herpes simplex viruses 1 and 2, and human herpes virus 6 (HHV-6) have been found in the cerebrospinal fluid and serum.

4- Genetic influences also appear to play a significant role in the development of MS.

Clinical Manifestations

The most common symptoms following an acute exacerbation include impairment of vision, muscular incoordination, and bladder dysfunction.

1. The clinical signs and symptoms of MS depend on the site of the demyelinating lesion of the CNS involved, and frequently affected areas include the optic chiasm, brainstem, cerebellum, and spinal cord.
2. More than 60% of individuals with MS have visual disturbances caused by demyelinating lesions of the second cranial nerve. The loss of vision usually occurs over a period of several days, with partial recovery within 1 month.
3. Other ophthalmic symptoms include "color blindness" and diplopia caused by involvement of the third, fourth, and sixth cranial nerves.
4. **Uhthoff's sign**, found in MS, is characterized by rapid vision loss following a body temperature increase that is associated with strenuous exercise.
5. MS patients frequently complain of electric shock–like sensations that are evoked by neck flexion and radiate down the back and into the legs. This is referred to as **Lhermitte's symptom** and is generally self-limiting but may persist for Years
6. Weakness or paresthesia of the extremities, with an increase in the deep tendon reflexes, is another common early finding in cases of MS.
7. bladder dysfunction, euphoria, ataxia, vertigo, and generalized incoordination
8. The majorities of cases of MS are chronic and are characterized by exacerbations and remissions over a period of many years.
9. During acute episodes, severe neurologic involvement is evident. This slowly resolves, but some permanent neurologic involvement remains after each episode

Diagnosis

1. Clinical and is based on the age of the patient, the presence of neurologic signs that cannot be explained by a single lesion, the progressive nature of the disease, and a history of exacerbations and remissions.
2. There are no definitive laboratory tests for MS, but demyelinating changes can be seen on (MRI) in more than 90% of cases. MRI demonstrates characteristic abnormalities of MS in >95% of patients. MS plaques are visible as hyperintense
3. Evoked potentials measure CNS electrical potentials, and abnormalities are detected in up to 90% of patients with MS.
4. CSF is often analyzed in patients suspected of having MS, and positive findings include an increase in total protein and mononuclear white blood cells.

Treatment

1- High doses of **intravenous corticosteroids** may arrest the progress of MS; about 85% of patients with relapsing-remitting MS show objective signs of neurologic improvement during treatment with intravenous corticosteroids. Glucocorticoids are used to manage both initial attacks and acute exacerbations of MS. Intravenous methylprednisolone is typically administered at a dose between 500 and 1000 mg/d for three to five days to reduce the severity and length of an attacks

2- Long-term treatment with immunosuppressants may reduce the frequency of relapse in patients with MS. Azathioprine is probably the safest drug in this category and has reduced relapse to 70% of study patients in 3 years. Administration of methotrexate appears to be the best therapy for slowing deterioration in patients with chronic progressive MS.

3- The use of interferon-γ-1b and -1a has shown promise; both have been shown to reduce clinical attacks and lesions

Oral Health Considerations

Individuals may present with signs and symptoms of MS.

1- Trigeminal neuralgia (TGN), which is characterized by electric shock–like pain, may be an initial manifestation of MS in up to 3% of cases. MS-related TGN is similar to idiopathic TGN . Features of MS-related TGN include possible absence of trigger zones and continuous pain with lower intensity.
2- Medications often used to manage TGN are similar to those used for treatment of idiopathic TGN.
3- Patients with MS may also demonstrate neuropathy of the maxillary (V2) and mandibular branches (V3) of the trigeminal nerve, which may include burning, tingling, and/ or reduced sensation.
4- Neuropathy of the mental nerve can cause numbness of the lower lip and chin.
5- **Myokymia** may be seen in patients with MS and consists of rapid, flickering contractions of the facial musculature secondary to MS lesions affecting the facial nerve.
6- Facial weakness and paralysis may also be evident in MS patients.
7- Dysarthria that results in a scanning speech pattern is often seen in patients with MS.
8- Temporomandibular disorder and headache.

Evaluate cranial nerve function, if cranial nerve abnormalities are detected, the individual should be referred to a neurologist for further evaluation.

It is recommended to avoid elective dental treatment in MS patients during acute exacerbations of the disease due to limited mobility and possible airway compromise.

Patients with significant dysfunction may require dental treatment in an operating room under general anesthesia due to the inability to tolerate treatment in an outpatient setting.

In addition, electric toothbrushes and oral hygiene products with larger handles may be necessary for completing oral hygiene in patients with significant motor impairment. be aware of possible interactions of these medications with those commonly used and prescribed in dentistry, as well as oral and systemic side effects of these agents.

ALZHEIMER'S DISEASE (AD)

Dementia is defined as an acquired deterioration in cognitive abilities that impairs the successful performance of activities of daily living. Memory is the most common cognitive ability lost with dementia; other mental faculties affected include problem-solving skills, judgment, visuospatial ability, and language.

The genetic basis of AD has been studied extensively, and specific genetic mutations have been implicated in both the familial and sporadic forms of the disease. Familial AD is an autosomal dominant disorder with onset typically prior to age 65 year

Clinical Manifestations

AD is a slowly progressive disorder represented by a continuum recognizes three stages of AD: (1) preclinical AD, (2) mild cognitive impairment due to AD, and (3) dementia due to AD.

Preclinical AD occurs before changes in cognition, and everyday activities are observed and primarily used for research purposes.

Cognitive impairment (CI) due to AD is characterized by mild changes in memory and other cognitive abilities that are noticeable to patients and families but are not sufficient to interfere with day to-day activities.

Dementia due to AD is characterized by changes in two or more aspects of cognition and behavior that interfere with the ability to function in everyday life. The initial signs of AD involve retrograde amnesia from progressive declines in episodic memory. This may initially go unrecognized or be viewed; however, as the disease progresses, memory loss begins to affect performance of daily activities, including following instructions, driving, and normal decision making.

As AD progresses, the individual is often unable to work, gets confused and lost easily, and may require daily supervision. Also language impairment, loss of abstract reasoning and skills. Advanced AD is characterized by loss of cognitive abilities, agitation, delusions, and psychotic behavior.

Patients may develop muscle rigidity associated with gait disturbances and often wander aimlessly.

In end-stage AD, patients often become rigid, mute, incontinent, and bedridden. Help is needed for basic functions, such as eating and dressing, and patients may experience generalized seizure activity. Death often results from malnutrition, heart disease, pulmonary emboli, or secondary infections.

Diagnosis

Diagnosis of preclinical AD primarily utilizes biomarker assessment, including markers of Aβ protein deposition in the brain, and markers of downstream neurodegeneration (elevated CSF tau protein and brain atrophy on MRI.

Clinical diagnosis of AD is based on an individual's medical history together with the clinical and neurologic examination findings.

Criteria include a history \of progressive deterioration in cognitive ability in the absence of other known neurologic or medical problems.

Possible AD refers to those who meet the criteria for dementia but have another illness that may contribute to the neurologic status, such as:- hypothyroidism or cerebrovascular disease, vitamin deficiency, depression, delerium, side effects of drugs and toxicity and excessive use of alcohol .

Diagnostic analysis of CSF may show a slight increase in tau protein and a lower concentration of Aβ peptide compared with healthy individuals or those with other dementias.
Electroencephalographic (EEG) studies typically demonstrate generalized slowing without focal features. Neuroimaging is important in evaluating suspected AD to exclude alternative causes of dementia, such as cerebrovascular disease, subdural hematoma, or brain tumor.
MRI and CT typically reveal dilatation of the lateral ventricles and widening of the cortical sulci, particularly in the temporal regions.
Volumetric MRI uniformly demonstrates shrinkage in vulnerable brain regions(brain atrophy).

Treatment
There is no cure for AD, and therapy is aimed at slowing the progression of the disease.
Cholinesterase inhibitors are approved to treat mild to moderate cases of AD and are considered the standard of care.
Memantine, a noncompetitive *N*-methyl-d-aspartate receptor antagonist believed to protect neurons from glutamate-mediated excitotoxicity, is used for treatment of moderate to severe AD.

Studies have demonstrated greater cognitive and functional improvement when memantine is used in conjunction with cholinesterase inhibitors compared to monotherapy.
Antidepressants, such as selective serotonin reuptake inhibitors, are commonly used to treat depression, which is often seen in the mild to moderate stages of AD. Antipsychotic agents are used for those patients who display aggressive behavior and psychosis, especially in the later stages of the disease.
Other agents that have been reported to be of clinical value in the treatment of AD include antioxidants, such as α-tocopherol (vitamin E), cholesterol-lowering drugs, anti-inflammatories, and herbal

Oral Health Considerations
Oral and dental health is a major issue in patients with AD because significant deterioration in oral health status is commonly observed with advancing disease.
Patients with AD appear to be at higher risk for developing coronal and root caries, periodontal infections, temporomandibularn joint abnormalities, and orofacial pain compared to healthy subjects.

Patients with AD should be placed on an aggressive preventive dentistry program, including an oral examination, oral hygiene education, prosthesis adjustment, and a three-month recall.

It is recommended to complete restoration of oral health-care function in the earliest stages of AD because the patient's ability to cooperate diminishes as cognitive function declines. Time-consuming and complex dental treatment should be avoided in persons with severe AD.
Medications used to treat AD can cause a variety of orofacial reactions and potentially interact with drugs commonly used in dentistry. Cholinesterase inhibitors may cause sialorrhea, whereas antidepressants and antipsychotics are often associated with xerostomia. In addition, dysgeusia and stomatitis have been reported with use of antipsychotic agents. Antimicrobials, such as clarithromycin, erythromycin, and ketoconazole, may significantly impair the metabolism of galantamine, resulting in central or peripheral cholinergic effects.

Anticholinesterases may increase the possibility of gastrointestinal irritation and bleeding when used concomitantly with NSAIDs.
Local anesthetics with adrenergic vasoconstrictors should be used with caution in AD patients taking tricyclic antidepressants due to potential risk of cardiovascular effects, such as hypertensive events or dysrhythmias

Parkinsonism

Is a neurodegenerative disorder characterized by:
1- Rigidity 2- tremors, 3- bradykinesis, and 4-impaired postural reflexes (postural instability).
The most common form of parkinsonism is Parksinson's disease (paralysis agitans), but parkinsonism is seen in a variety of disorders such as postencephalitic parkinsonism, and post-traumatic parkinsonism following closed head injury.
Many of the signs of Parkinson's disease are found in the head and neck. The typical "masklike" facial appearance with infrequent blinking and lack of expression is caused by bradykinesis.
The muscle rigidity also causes difficulty in swallowing, resulting in saliva drooling.
Speech affected because of the lack of muscle control, and mandibular tremor results in masticatory difficulties, especially in those with removable dental appliances.
Abnormalities in oral behavior, such as purposeless chewing, grinding, and sucking movements, are also well recognized in patients with Parkinson's disease and make dental treatment especially difficult.

Treatment

Drug treatment is often not required early in the course of Parkinsonism.

1- Patients with mild symptoms but no disability may be helped by amantadine. This drug improves all of the clinical features of Parkinsonism.
2- Anticholinergics are more helpful in alleviating tremor and rigidity than in alleviating bradykinesia, but these drugs have many side effects.
3- Levodopa, a dopamine precursor that can cross the blood-brain barrier, improves all the major features of Parkinsonism.

Bell's palsy

Bell’s palsy is recognized as a unilateral paresis of the facial nerve. The dysfunction has been attributed to an inflammatory reaction involving the facial nerve.

A relationship has been demonstrated between Bell’s palsy and the isolation of herpes simplex virus 1 from nerve tissues.

Bell’s palsy begins with slight pain around one ear, followed by an abrupt paralysis of the muscles on that side of the face. The eye on the affected side stays open, the corner of the mouth drops, and there is drooling.

As a result of masseter weakness, food is retained in both the upper and lower buccal and labial folds. The facial expression changes remarkably, and the creases of the forehead are flattened.

Due to impaired blinking, corneal ulcerations from foreign bodies can occur.

Causes

Although the exact reason Bell's palsy occurs isn't clear, it's often linked to exposure to a viral infection. Viruses that have been linked to Bell's palsy include the virus that causes:

- Cold sores and genital herpes (herpes simplex)
- Chickenpox and shingles (herpes zoster)
- Mononucleosis (Epstein-Barr)
- Cytomegalovirus infections
- Respiratory illnesses (adenovirus)
- German measles (rubella)
- Mumps (mumps virus)
- Flu (influenza B)

Symptoms

Signs and symptoms of Bell's palsy come on suddenly and may include:

1. Rapid onset of mild weakness to total paralysis on one side of face — occurring within hours to days
2. Facial droop and difficulty making facial expressions, such as closing eye or smiling
3. Drooling
4. Pain around the jaw or in or behind ear on the affected side
5. Increased sensitivity to sound on the affected side
6. Headache
7. A decrease in ability to taste
8. Changes in the amount of tears and saliva
9. In rare cases, Bell's palsy can affect the nerves on both sides of face

Diagnosis

There's no specific test for Bell's palsy. Look at face and ask to move facial muscles by closing eyes, lifting brow, showing teeth and frowning, among other movements.

Other conditions — such as a stroke, infections, Lyme disease and tumors — can also cause facial muscle weakness, mimicking Bell's palsy, may recommend other tests, including:

Electromyography (EMG). This test can confirm the presence of nerve damage and determine its severity. An EMG measures the electrical activity of a muscle in response to stimulation and the nature and speed of the conduction of electrical impulses along a nerve.
Imaging scans. Magnetic resonance imaging (MRI) or computerized tomography (CT) may be needed on occasion to rule out other possible sources of pressure on the facial nerve, such as a tumor or skull fracture

Treatment

Commonly used medications to treat Bell's palsy include:

Corticosteroids, such as prednisone, are powerful anti-inflammatory agents. If they can reduce the swelling of the facial nerve, it will fit more comfortably within the bony corridor that surrounds it. Corticosteroids may work best if they're started within several days of when symptoms started.

Antiviral drugs. The role of antivirals remains unsettled. Antivirals alone have shown no benefit compared with placebo. Antivirals added to steroids .However, despite this, valacyclovir (Valtrex) is sometimes given in combination with prednisone in people with severe facial palsy.

Physical therapy

Paralyzed muscles can shrink and shorten, causing permanent contractures. A physical therapy by massage and exercise of facial muscles to help prevent this from occurring.

Surgery

In the past, decompression surgery was used to relieve the pressure on the facial nerve by opening the bony passage that the nerve passes through. Today, decompression surgery isn't recommended. Facial nerve injury and permanent hearing loss are possible risks associated with this surgery.

Myasthenia gravis

Is a disease characterized by progressive muscular weakness on exertion, secondary to a disorder at the neuromuscular junction.

It is autoimmune disease ,autoantibodies combine with and may destroy the acetylcholine receptor sites at the neuromuscular junction, preventing the transmission of nerve impulses to the muscle .The initial signs of this disease commonly occur in areas innervated by the cranial nerves (frequently, the eye muscles).
Patients present with

1. ptosis, diplopia
2. difficulty in chewing or swallowing
3. respiratory difficulties
4. limb weakness
5. or some combination of these problems.

Oral and facial signs

1. The facial muscles of expression are involved
2. Tongue edema making eating difficult for patients
3. difficulty in chewing; these patients will be unable to finish chewing a bolus of food because of the easy fatigability of the muscles

Treatment

1. Anticholinesterase drugs such as neostigmine and pyridostigmine bromide
2. thymectomy
3. Long-term cortico-steroids and immunosuppressive drugs are necessary.

Dental management

1-A respiratory crisis may develop from the disease itself or from over medication.
2- Dental treatment should be performed in a hospital where endotracheal intubation
3-The airway must be kept clear because aspiration may occur in patients whose swallowing muscles are involved.
4-Adequate suction and the use of a rubber dam are aids in these cases.
5-The dentist should avoid prescribing drugs that may affect the neuromuscular junction, such as: Narcotics, tranquilizers, and barbiturates.
Certain antibiotics, including tetracycline, streptomycin, sulfonamides, and clindamycin, may reduce neuromuscular activity and should be avoided.

SEIZURE DISORDERS & Epilepsy

A seizure is a paroxysmal event due to abnormal, excessive, hypersynchronous discharges from neuronal aggregates in the CNS. The term *epilepsy* describes a group of neurologic disorders characterized by recurrent seizure activity.
1- Focal, 2- generalized and 3- unknown seizures are currently the three major categories of seizure activity used in clinical practice.

1-The focal seizure category (Partial Seizures)
Includes partial seizures; this type of seizure activity originates within networks limited to one hemisphere and clinical manifestations of these seizures depend on the site of origin. Simple partial seizures reflect neuronal discharge from a discrete cortical locus, such as the motor cortex of the frontal lobe, or in subcortical structures, and generally not associated with impaired consciousness.
Simple partial seizures consist of clonic activity, which are rapid jerks that also can be accompanied by somato-sensory phenomena, visual changes/distortions, and auditory, olfactory, and gustatory
2- Generalized seizures arise from both cerebral hemispheres simultaneously and have distinctive clinical features that facilitate diagnosis. The underlying pathophysiology of generalized seizures is attributed to abnormal neuronal excitability.
a- Absence seizures (petit mal) are a type of generalized seizure that is characterized by sudden, brief lapses of consciousness without loss of body tone and may be attributed to abnormal oscillatory rhythms generated during sleep by circuits connecting the thalamus and cortex.
b- **Tonic-clonic (grand mal)** seizures are generalized seizures that present with dramatic clinical features, most notably, tonic contracture and uncoordinated clonic muscular movements.

Other types of generalized seizures include atypical absence, atonic, and myoclonic seizures. 3- Those seizures that cannot be classified as either focal or generalized are termed **unknown seizures**.

Etiology usually varies according to patient's age.

The most common seizures arising in late infancy and early childhood are febrile seizures without evidence of associated CNS infection; these usually occur between 3 months and 5 years of age and have a peak incidence between 18 and 24 months.

Isolated, non recurrent, generalized seizures among adults are caused by multiple etiologies, including metabolic disturbances, toxins, drug effects, hypotension, hypoglycemia, hyponatremia, uremia, hepatic encephalopathy, drug overdoses, and drug withdrawal.

Cerebrovascular disease may account for approximately 50% of new cases of epilepsy in patients older than 65 years. Other etiologies for epilepsy include degenerative CNS disease, developmental disabilities, and familial/genetic factors. Epilepsy occurs more frequently in individuals who have neurologic-based disabilities, such as cerebral palsy and autism.

Epilepsy

Epilepsy is a condition characterized by abnormal, recurrent, and excessive neuronal discharges precipitated by many different disturbances within the central nervous system.

These aberrant discharges may cause episodes of sensory and motor abnormalities as well as loss of consciousness.

Common causes of epilepsy:

1-Infants are much more likely to suffer from epilepsy after complications at birth, such as anoxia lack of oxygen traumatic brain injury during delivery, intracranial injury, metabolic disorders, abnormal brain development and congenital malformations.

2- Predominant causes in children and adolescents include head trauma and acute or febrile infections, fever, brain tumors, genetic disorders and brain scarring.

3- Young adults with alcohol or drug abuse commonly suffer from generalized seizures after periods of severe abuse.

4- Epilepsy in older adults occurs as a complication of any of the previously mentioned causes but is more often associated with cerebrovascular diseases such as stroke, brain scarring, abnormal brain development, head trauma and brain tumors.

International Classification of Epileptic Seizures

1- Partial (focal) seizures
   Simple partial seizures
   Complex partial seizures
   Partial seizures leading to secondarily generalized seizures
2- Generalized Seizures
   Absence seizures (petit mal)
   Typical

Atypical
Tonic-clonic seizures (grand mal)
Myoclonic seizures
Clonic seizures
Tonic seizures
Atonic seizures (astatic seizur

Generalized seizures

The majority of generalized seizures are called either:-

1 -tonic-clonic seizures (grand mal).(MOST COMMON TYPE 90% of epileptics experience it alone or in combination with another type of seizure)

2 -absence (petit mal) seizures

Tonic-clonic seizures (A grand mal seizure) :characteristically begins with an aura. The aura may be experienced as epigastric discomfort, as an emotion, or as a hallucination of hearing, vision, or smell.

The aura is followed seconds to minutes later by unconsciousness, or a cry, then tonic muscle spasms; this rigid phase lasts about 30 seconds. Because of the spasm of the respiratory muscles, the patient does not breathe and becomes cyanotic during this period.

The tonic phase is followed by a clonic phase composed of convulsive jerky movements, incontinence, and tongue biting.

Absence seizures (petit mal):

Is the second most common type of seizure and it occurs without an aura and with few or no clonic or tonic movements.

Absence seizures present almost exclusively in children and frequently disappear during the second decade of life.

Diagnosis:

1. History & physical examination are critical because the diagnosis may based on clinical findings.
2. A complete neurological examination (testing of cranial nerves)
3. Blood studies: complete blood count, mg, calcium , glucose to identify metabolic cause
4. Toxins screen: to identify seizure due to drugs, lumber puncture to exclude any infectious cause
5. Brain imaging: underlying CNS structural abnormalities or pathology

MRI and CT

6. EEG (to classify the seizure & to determine the type of anticonvulsant)

Treatment

Antiepileptic drugs (AEDs)

*phenytoine: long half life,dosed less frequently cause gingival over growth, hirsutism, coarsening of facial featuers

*carbamazepine: hepatotoxicity, leukopenia,aplastic anemia
*Lamotrigine: skin rash
*Valproic acid: treatment of G.tonic clonic,can cause bone marrow suppressison & hepatotoxicity
*Additional drugs as topiramate, gabapentin & oxcarbazepine
*surgical procedures: limited removal of hippocompus & amygdala, temporal lobectomy or hemispherectomy
*Vagus nerve stimulation: placement of an electrode on the left vagal nerve leading to wide spread activation of cortical & sub cortical pathways

Discontinuation of pharmacologic therapy is considered when seizure control has been achieved. The following patient characteristics yield the greatest chance of remaining seizure free after discontinuation of drug therapy:
(1) Complete medical control of seizures for one to five years;
(2) Single seizure type; (3) normal neurologic examination, including intelligence; and (4) a normal EEG.
Many patients are often withdrawn successfully from medication after an interval of two to four years without seizures who meet the above criteria and who clearly understand the risks and benefits.
Patients may use three or more drugs to successfully treat refractory epilepsy; however, up to 30% of patients are resistant to all medical therapies. Surgical procedures may be indicated for these patients.
Deep brain stimulation (DBS) and responsive neurostimulation systems are also currently used for treatment of refractory epilepsy.
Gene therapy is currently being investigated as an alternative treatment modality for epilepsy refractory to standard therapies.

Oral health consideration
* Uncontrolled Patients should be referred to a hospital
*Patient with implanted vagus nerve stimulator do not require antibiotic prophylaxis
*we must avoid any triggers of the patient seizures activity
*Placement of fixed prosthesis is recommended rather than removable prosthesis.
*Patient taking the medication mentioned requires laboratory evaluation prior to dental treatment
*Aspirin& NSAD should be avoided in patient taking valproic acid
*Gingival over growth intraoral lesion & lips enlargement
*Xerostomia:
Reduced salivary flow may result from the use of AEDs, may observe increased dental caries and oral candidiasis in patients using these agents.
Topical fluoride should be considered for patients with seizure disorders who are at increased risk of developing dental caries, and antifungal agents should be prescribed if oral candidiasis develops. Additional oral findings in patients taking AEDs may include stomatitis, glossitis, and ulcerations.

12 Cranial Nerve

Cranial Nerve	Assessment
I olfactory	Smell
II optic	Vision
III oculomotor	Eye movements, PERRLA, eyelids
IV trochlear	
V trigeminal	Facial sensations, corneal reflex
VI abducens	Assessed with III and VI
VII facial	Taste, smile, frown, close eyes tightly
VIII acoustic	hearing
IX glossopharnxgeal	Gag reflex, swallowing, taste;
X vagus	
XI spinal accessory	Shrug shoulders, turn head against resistance
XII hypoglossal	Stick out tongue, move tongue side to side