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Cranial Nerves

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Introduction:

Hi everyone! My name is Alekhya Nimmagadda and I am a medical student at the Apollo Institute of Medical Sciences and Research. I would like to thank Dr. Janette Mailo, a Pediatric Neurologist at the University of Alberta , for guiding me and helping me develop this podcast on cranial nerves**.** I would also like to thank Dr. Melanie Lewis and the Pedscases team for their support in creating this podcast.

Objectives:

By the end of this podcast, listeners should be able to

1) Define and Identify cranial neuropathies in children

2) Discuss the etiology and clinical manifestations of various cranial nerve disorders

3) Demonstrate how to examine each cranial nerve

4) Review the investigations and management of cranial nerve disorders

Clinical cases:

Let's look at some clinical cases affecting the cranial nerves.

Clinical case 1:

A 4 month-old previously healthy infant presents with a 3 day history of poor sucking, drooling, weak cry and constipation. Today she cannot hold her head up without support.

On the exam, the pertinent cranial nerve findings include restricted range of eye movements, non-reactive pupils, bilateral ptosis, absent gag reflex and a weak suck.

Brief past medical history reveals a healthy, infant born at 39 weeks of gestation by spontaneous vaginal delivery after an uncomplicated pregnancy to a 31 year old G3P2 mother. The infant met all her early developmental milestones as expected for age.

There is no pertinent family history. The family has recently moved to a new home in an area under development with lots of surrounding construction. The maternal grandmother recently watched the baby for a few hours and she mentioned she had dipped the soother in honey to calm the crying infant.

Clinical case 2:

A 7-year-old girl presents with a sudden onset of right unilateral facial paralysis. Prior to the onset, she felt intermittent ear pain, which has since resolved.

On examination there is facial asymmetry and loss of facial expressions on the right side, involving both the upper and lower face with flattening of the forehead and inability to raise the eyebrow on the affected side. Blinking is also absent on the right and she can't fully close her right eye. Nasolabial fold appears flattened and the right corner of the mouth is drooping. Otoscopic examination of the ear shows erythema and bulging of the tympanic membranes.

There is no other cranial nerve involvement. No facial numbness. Full extraocular eye movements. Normal external auditory canal and soft palate. No hyperacusis or hearing impairment. No cerebellar signs or any other neurological abnormalities are present.

Definition:

What is a cranial neuropathy?

Let's start with a definition

A cranial Neuropathy is a dysfunction or problem with a nerve. It can lead to pain, tingling, numbness or weakness of the muscle supplied by that nerve.

There are 12 pairs of cranial nerves in the human body. Cranial nerves 1 and 2 arise from the cerebral neurons and the remaining cranial nerves 3-12 arise from the neurons of the brainstem.

Cranial Nerves disorders are caused by damage to the cranial nerves. This damage may be congenital or acquired.

In this podcast we will be discussing various disorders of cranial nerves. We will discuss etiologies, approach to physical exam and management.

Etiology and Clinical Presentation of Cranial Nerve disorders:

Cranial Nerves 1-12

In general the etiology of any cranial nerve disorder can be broadly attributed to: infection, inflammation, degenerative disease, tumor or trauma.

Now, I will be discussing each individual cranial nerve.

Cranial Nerve 1: Olfactory Nerve

The cranial nerve 1 is called the olfactory nerve. This nerve is responsible for the sensation of smell. Damage to this nerve can lead to loss of smell, a rare condition called anosmia.

Congenital anosmia can occur due to a condition called Kallmann syndrome, a genetic syndrome also associated with hypothalamic hypogonadism**.** In this condition the GnRH neurons fail to migrate from the olfactory bulb to the hypothalamus**.** Affected individuals present with loss of smell as well as hypogonadism that delays or prevents puberty.

Anosmia can also occur due to traumatic damage to cranial nerve 1. This type of damage can often lead to permanent loss of smell.

Some other causes of dysfunction of cranial nerve 1 include acute or chronic rhinitis, smoking, cystic fibrosis, or secondary to certain medications.

Cranial Nerve 2: Optic Nerve

The optic nerve is responsible for vision. Dysfunction of the optic nerve can lead to vision loss.

There are several reasons for optic nerve dysfunction. Optic neuritis is an inflammation of the cranial nerve 2. Other causes include optic nerve atrophy and papilledema due to increased intracranial pressure. Papilledema means swelling of part of the optic nerve due to increased intracranial pressure.

Optic neuropathy can be seen in patients with multiple sclerosis which is an autoimmune demyelinating disorder of the CNS neurons.

Damage to the optic nerve can also occur due to trauma and lead to post traumatic blindness.

Cranial Nerves 3,4 and 6:

Oculomotor, Trochlear and Abducens nerves. We usually think of these three nerves together, because they are jointly responsible for coordinated movements of the eye.

The oculomotor nerve which is cranial nerve 3, supplies the superior rectus, medial rectus, inferior rectus and the inferior oblique muscles. It also supplies the levator palpebrae superioris. Injury to this nerve can then lead to the eye turning down and out. There may also be a ptosis which is drooping of the eyelid. The parasympathetic nerves lie in close proximity to cranial nerve 3, and therefore parasympathetic innervation can be disrupted when there is damage to cranial nerve 3. Loss of parasympathetic innervation would lead to miosis as well.

The trochlear nerve which is cranial nerve 4, is the smallest nerve only supplying the superior oblique. This nerve has an important function in downward gaze. Therefore, damage to cranial nerve 4 leads to impairment in everyday activities like reading and walking down the stairs.

The abducens nerve, the cranial nerve 6, supplies the lateral rectus, which moves the eye laterally, or out to the side.

Causes of dysfunction of cranial nerve 3, 4 and 6 include infantile botulism, congenital horner's syndrome, ophthalmoplegic migraine and myasthenia gravis.

Cranial Nerve 5- Trigeminal Nerve- This nerve is responsible for providing sensation to the face and motor innervation to the jaw.

It has three branches: Ophthalmic, maxillary and mandibular.

Damage to the trigeminal nerve can be caused by neoplasms like schwannomas and meningiomas, cavernous sinus thrombosis, a blood clot in the cavernous sinuses which can be life threatening, or lateral medullary syndrome, which is the result of the infarction of the lateral medulla.

Trigeminal Neuralgia is a disorder causing extreme pain in the face. This pain often causes the patient to wince, therefore another name for it is tic douloureaux.

Cranial Nerve 7: This is called the facial nerve.

Neoplasms, stroke and infection can all cause damage to cranial nerve 7.

Bell's Palsy is one of the most common causes of facial nerve palsy.

Bell's palsy can be idiopathic or it can be related to an infection with the Herpes Zoster virus. Lyme disease has also been found to be a cause of Bell's palsy.

As we demonstrated in our second clinical case, Bell's palsy usually begins in children as pain behind the ear. Paralysis of the face typically follows. Bell's Palsy is characterized by a weakness of both, the upper and the lower parts of the face. As a lower motor neuron lesion, Bell's palsy affects the entire side of the face including the forehead. Careful physical exam can easily distinguish a lower motor neuron lesion from an upper motor neuron lesion. An upper motor neuron lesion would cause the forehead to be spared, since upper motor neuron innervation to the forehead is coming from both sides of the cortex.

Besides weakness of the forehead, a child with Bell's palsy will have difficulties closing the eye on the affected side and they will have an asymmetrical smile. Incomplete eye closure can lead to eye dryness.

The facial nerve is also responsible for the sensation of taste from the anterior 2/3 of the tongue, and for the motor portion of the corneal reflex.

Cranial Nerve 8- This is the vestibulocochlear nerve.

Damage to this nerve can result in hearing loss if the cochlear portion of the nerve is injured.

Disruption of the vestibular portion can lead to vertigo, nausea, vomiting and nystagmus.

Common causes of cranial nerve 8 disorders include acoustic neuromas, labyrinthitis and vestibular neuronitis.

Cranial Nerves 9 and 10- Glossopharyngeal and Vagus nerves

The glossopharyngeal nerve plays an important role in swallowing as well as providing taste sensation to the posterior 1/3 of the tongue. Cranial nerve 9 also supplies the stylopharyngeus and it elevates the pharynx. Innervation of the parotid gland by glossopharyngeal nerve is responsible for salivation.

The vagus nerve plays an important role in swallowing, talking, coughing and elevating the palate. The vagus nerve also keeps the uvula in the midline position and therefore,

damage to this nerve can cause deviation of the uvula, ipsilaterally, meaning to the side of the lesion.

These nerves can have their normal function altered as a part of cerebral palsy and they can also be affected in pseudobulbar palsy.

Moreover, cranial nerve 9 can be affected by botulism as well.

Cranial Nerve 11- Spinal Accessory nerve

The spinal accessory nerve supplies the sternocleidomastoid and the trapezius muscles, which are involved in turning the head and shrugging the shoulders, respectively.

Dysfunction of spinal accessory nerve can lead to weakness and atrophy of these muscles, leading to inability to shrug the shoulders and shoulder pain. The etiologies include trauma and iatrogenic causes.

Our last cranial nerve is Cranial Nerve 12:

Hypoglossal Nerve:

This nerve supplies the tongue.

Damage to this nerve can be due to trauma or iatrogenic causes.

Loss of this nerve's function can lead to weakness, fasciculations and atrophy on one side of the tongue.

Examination of Cranial Nerves:

Cranial Nerve 1:

The olfactory nerve is usually not tested in infants due to their limited ability to communicate. In older children the olfactory nerve can be tested by asking the patient to sense a known type of smell. If there is cranial nerve 1 dysfunction, the patient will not be able to smell.

Cranial Nerve 2: The optic nerve can be assessed by testing of the patient's vision.

In infants, a common cranial nerve 2 test is to shine a bright light into the infant's eyes and observing if the infant blinks in response to the light.

In older children the optic nerve can be tested by the following:

Assessment of visual acuity. A Snellen's chart is typically used to test visual acuity.

Visual Field testing- This can be assessed by visual field confrontation. Each eye should be tested separately. The child is asked to focus on the examiner's nose and as the examiner moves their fingers within visual fields superiorly and inferiorly, the child with the intact visual fields should still be able to see the examiner's fingers out of the corner of their eyes.

When we are testing pupillary responses, we are looking for pupillary constriction in response to bright light shining into the eyes.

Fundoscopy will look for papilledema.

Cranial Nerves 3,4 and 6:

We have a few tests that can assess if these nerves are intact. First is the pupillary response test, as mentioned before, we are looking for pupillary constriction in response to bright light.

In a cooperative child, cranial nerves 3, 4 and 6 can be assessed by examining coordinated movements of the eyes. The examiner can ask the child to focus on an object, or the examiner's finger, as the examiner moves the object, or finger, horizontally and vertically. If these nerves are intact, the eyes should move smoothly and fully in all directions.

Doll's eye maneuver is typically used in comatose patients. The examiner turns the patient's head horizontally and vertically while watching for conjugate movement of the eyes in the opposite direction.

Cranial nerve 5:

In an infant, the trigeminal nerve can be easily tested by the sucking reflex. If a nipple or a pacifier is placed near the infants mouth, they will start to suck on it.

In an unresponsive patient, cranial nerve 5 can be assessed by testing the corneal reflex. The examiner will gently touch the tip of a cotton applicator to the edge of the cornea. Intact cranial nerve 5 (and 7) will result in a blink. The trigeminal nerve is responsible for the sensation and the facial nerve is responsible for the blink response of the eye.

In a cooperative older child, the trigeminal nerve can be tested by examining the sensation of the face. This can be done by touching various areas of the face and asking the patient if they can feel the sensations.

The Jaw jerk can also be assessed. The examiner places their finger on the child's jaw and taps it. The jaw will move slightly in the presence of an intact nerve.

Cranial Nerve 7:

In infants, the facial nerve can be easily tested by observing the movements and symmetry of the face while the infant cries or grimaces.

Cooperative children can be asked to blow up their cheeks, raise their forehead and smile.

Abnormality in the facial nerve can lead to an asymmetric smile, drooping of the face and difficulties closing the eye on the affected side. The child will not be able to puff their cheeks or raise their forehead.

In an unresponsive child, corneal reflex can be tested as we already explained.

Cranial Nerve 8:

In an infant vestibulocochlear nerve can be tested by audiometry.

In an older child the cranial nerve 8 can be tested by Weber or Rinne tests. In the Webber test, a vibrating tuning fork is placed on the patients forehead. The vibration should be equally distributed to both sides of the head and should be perceived equally in both ears. If the vibration is localised to one side, the patient might be suffering from hearing loss. In conductive hearing loss, the vibration is more prominent in the affected ear. In sensorineural hearing loss, the vibration is perceived stronger in the normal ear.

In the Rinne test the examiner places the vibrating tuning fork on the patient’s mastoid bone and waits until the sound from the vibrating tuning fork stops completely. Then the examiner positions the same vibrating tuning next to the patient’s ear on the same side, but without touching it. If the patient continues to hear the vibrations of the tuning fork,

then air conduction is better than bone conduction. This is seen with a normal cranial nerve 8 and in sensorineural hearing loss. If bone conduction is better than air conduction, then the patient has conductive hearing loss.

Cranial Nerves 9,10-

In infants, these cranial nerves can be tested by assessing sucking and swallowing, as well as by the quality of the infant's cry.

Gag reflex can also be tested in an unresponsive patient. Alternatively, the uvula can be touched by the cotton tip or a tongue depressor, and if the cranial nerve 9 is intact, the uvula will symmetrically elevate.

Cranial nerve 11: Spinal accessory nerve

In infants, this nerve can be examined by observing the movements of the neck, looking especially for symmetry and in young infants, head control.

In cooperative children cranial nerve 11 can be assessed by asking the patient to shrug their shoulders against resistance. Next, resistance is applied on the side of the head and the patient is asked to turn their head and push against the resistance.

Cranial Nerve 12:

This nerve can be tested in infants by observing the movement of the tongue during sucking.

Older children can be asked to stick out their tongue. Tongue deviation, if present, is towards the side of the lesion. The weakness of cranial nerve 12 can lead to atrophy, weakness and fasciculations

Supportive management and Treatment of Cranial Nerve Disorders:

There are 12 cranial nerves and many diseases that can impact the function of these important nerves. The etiologies of the disorders of cranial nerves can be broadly categorized as congenital or acquired. Acquired caused include infection, inflammation, trauma, neoplasms and neurodegenerative disease.

Management of the cranial nerve disorders depends on the cause. Therefore, the very first step is to diagnose the disorder and identify the underlying cause.

Infection can be a common cause of disorders of cranial nerves. If infection is suspected, serology can be helpful to identify an infectious agent. Once the infection is identified the appropriate treatment can be initiated.

In the case of a suspected infantile botulism, the confirmatory test is a stool culture. However, obtaining a stool sample for an analysis can be difficult in a constipated child suffering from botulism. Therefore if Clostridium botulinum infection is suspected in an infant, the botulinum antitoxin should be given immediately while the confirmatory test is pending.

Bell's palsy can be caused by Herpes zoster virus, Lyme disease or associated with Guillan Baree Syndrome. The appropriate treatment must be given without delay even before the results are available, using careful clinical assessment.

In Bell's palsy specifically, mainstay of treatment is Prednisone 1-2 mg/kg/day orally for 7-10 days. Pain management is important and NSAIDs are frequently used. Most children recover from Bell's palsy but in rare cases, residual impairment persists.

Inflammatory disorders including optic neuritis, labyrinthitis, or vestibular neuritis can affect several cranial nerves, and the treatment is corticosteroids to supress inflammation.

Neoplasms can also lead to cranial neuropathies due to their mass effect. CT and MRI can identify the mass. Surgical decompression or radiation may be indicated to remove or reduce the size of the mass and alleviate the symptoms.

For cranial neuropathies that are caused by trauma, conservative management including pain management is the first step. Sometimes surgical techniques can help in selective cases where conservative management fails and the symptoms are particularly bothersome.

Trigeminal neuralgia is an incompletely understood and rare disease in children. It causes severe neuropathic pain in the distribution of the trigeminal nerve. It is very difficult to treat but medications such as carbamazepine or gabapentin can alleviate the pain in some patients.

Kallmann syndrome can be diagnosed by brain MRI and treated by hormonal replacement.

Discussion of our Clinical Cases:

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Now that we have some insight into the function of cranial nerves and how to test them let's revisit our clinical cases.

Our first case was Infantile botulism. Infantile botulism is caused by a bacteria called Clostridium botulinum following ingestion of preformed spores of clostridium botulinum. The spores can be found in raw honey, but also in dirt and construction dust. The botulinum neurotoxin prevents acetylcholine release from the pre synaptic terminal of the motor neuron causing progressive muscle weakness. Infantile botulism presents with multiple cranial nerve palsies.

As described, patients often show symptoms such as ptosis of the eyelids and pupils unreactive to light, both characteristic of dysfunction of cranial nerve 3. In addition the baby is not sucking very well. This demonstrates that cranial nerve 5 is affected. There is an absent gag reflex indicating that cranial nerve 9 is affected as well.The diagnosis is confirmed by stool sample analysis, however, since this can be difficult to obtain in a constipated weak infant, the treatment should not be delayed and the infant should promptly receive immunoglobulin antitoxin.

The diagnosis in our second case was Bell's palsy, describing a previously well 7 year old girl who recently developed acute otitis media, followed by facial asymmetry and loss of facial expression involving cranial nerve 7. The physical exam confirms the involvement of both the upper and lower face indicating it is a lower motor neuron lesion. Since the facial nerve is the efferent nerve involved in the corneal reflex, facial nerve palsy can cause absence of blinking as seen in our case. Facial nerve palsies can also cause drooping of the mouth and inability to raise eyebrows as described. The treatment of facial nerve palsy is oral glucocorticoids such as prednisone.

Key points and Conclusion:

1.There are 12 pairs of cranial nerves.

2. Cranial nerves can have sensory function, motor function or both.

3. Cranial nerve disorders occur when there is dysfunction or damage to a cranial nerve.

4. The majority of cranial neuropathies are caused by infections, trauma, inflammation or a neoplastic process. This can lead to loss of cranial nerve function. There can be sensory loss or motor loss to the muscles the individual cranial nerves supply.

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5. In order to diagnose a cranial nerve dysfunction, a careful examination of each cranial nerve must be performed. Specific tests can be done for each cranial nerve to diagnose the cranial neuropathy.

6. The treatment of cranial neuropathies is specific to each individual cause. Treatment is directed toward the underlying etiology.

7. Broadly, infections can be treated with the appropriate antibiotics or antiviral medications. The main treatment for botulism is antitoxin and supportive care.

8. Inflammatory disorders are treated with corticosteroids to suppress inflammation.

9. For cranial neuropathies caused by neoplastic processes and trauma, supportive care, pain management and in selective cases surgical management may be considered.

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