Chapter 175 – Pediatric Neurologic Disorders

Episode Overview

- Causes of altered mental status in children are broad. Use a comprehensive approach! (e.g. DIMES)
    - Includes vascular events (e.g., stroke, arteriovenous malformation with bleed), infection (e.g., meningitis, sepsis, encephalitis), trauma, toxic ingestion, anatomic or structural abnormality (e.g., intracranial mass or tumor), metabolic derangements, intussusception, or seizures, which may be subclinical.
- Altered mental status in children has a varied spectrum of clinical presentations and may include any of the following: altered level of consciousness, excessive sleepiness, irritability, lethargy, and abnormal behavior.
- Signs and symptoms of bacterial meningitis vary by age and can include nonspecific manifestations in infants and young children, such as irritability and lethargy.
- Antibiotic therapy for the patient with suspected bacterial meningitis, should not be delayed by an LP. Initial antibiotic coverage should be broad and guided by the most likely pathogens in the patient's age group.
- When treating the patient with seizures:
    - A careful and detailed history is instrumental in determining whether an event was a seizure. If the event was a seizure, the history should delineate which type of seizure occurred (partial or generalized) and whether the clinical event fits into a known epilepsy syndrome.
    - Status epilepticus constitutes a neurologic emergency that carries high morbidity and mortality rates. Initial treatment is typically with IV lorazepam, followed by fosphenytoin.
- A simple febrile seizure is generalized, lasts less than 15 minutes, and occurs in a neurologically and developmentally normal child between 6 months and 6 years of age.
- Children with simple febrile seizures do not require blood and urine testing, other than as needed lab work for the evaluation of the fever source. In immunized infants 6 to 18 months of age with a first-time simple febrile seizure, a lumbar puncture (LP) can be avoided if the child is not-ill appearing, has returned to baseline, and there are no clinical signs of meningitis.
- The possibility of provoked seizures should be considered because many causes are treatable.
- Breath-holding spells occurs in children 6 months to 6 years of age and are triggered by pain or emotional upset. After a trigger, the child becomes pale or cyanotic and may lose consciousness, sometimes with a brief period of clonic movements or opisthotonos.
- When treating the patient with headaches, consider the following:
    - Most headaches in children have benign causes that usually can be diagnosed by a careful history and physical examination. Radiologic evaluation by CT, MRI, or both may be necessary to rule out secondary causes of headache such as intracranial hemorrhage, subarachnoid hemorrhage, brain tumor, or brain abscess.
    - Warning signs of secondary headaches include sudden onset, occurrence with straining or exertion, association with neurologic symptoms, change in headache pattern, nocturnal awakening, and bilateral occipital headaches.

- When treating the patient with ataxia:
    - In children, 40% of ataxia cases are caused by acute cerebellar ataxia.
    - A toxicology screen is the test with the highest diagnostic yield for acute-onset ataxia in children.
    - Approximately 45% to 60% of all childhood brain tumors arise in the brainstem or cerebellum and can manifest as slowly progressive ataxia.
- The cause of vertigo with hearing loss is significantly different from that of vertigo without hearing loss. This differentiation can be helpful in guiding the diagnostic evaluation.
- When assessing an infant or child with motor weakness, it is important to distinguish presentations consistent with upper motor neuron pathology from lower motor neuron processes.
- When treating the pediatric patient with either a suspected or confirmed stroke, consider the following:
    - Strokes are less common in children than in adults. They represent a neurologic emergency and may be hemorrhagic or ischemic in nature. Imaging with CT and/or MR can help confirm the diagnosis of stroke. An important component of stroke management involves the prevention of secondary brain injury.
- Spinal cord compression is a medical emergency regardless of cause and requires prompt diagnosis and treatment. It may arise from trauma, infection, inflammation, or malignancy.
- The diagnosis of Guillain-Barré syndrome (GBS) is largely clinical, although LP may be helpful in confirming the diagnosis. Patients with GBS are at risk for respiratory compromise and should be admitted to the hospital for observation and supportive care.
- The diagnosis of infant botulism is largely clinical. If there is high clinical suspicion, treatment should be initiated promptly, without awaiting laboratory confirmation. Given the risk of respiratory compromise, infants with botulism should be admitted to the hospital for observation and supportive care.
- Diagnosis of myasthenia gravis in the ED may be confirmed with the Tensilon test. (remember: injection of an acetylcholine esterase inhibitor to assess for increased strength on repetitive movements). The disorder can often be treated on an outpatient basis, but patients with truncal involvement and concern for respiratory compromise should be admitted to the hospital for observation and supportive care.

Core Questions

1. List ten causes of provoked (acutely symptomatic) seizures.
2. List four episodic disorders that may mimic seizures in neonates and four in non-neonates.
3. Give an approach to acute seizure control in a 9-month-old and in a 5-year-old.
    a. How does it differ if you are unable to obtain IV or IO access?
4. What is the definition of status epilepticus?
5. List 6 medical treatments for status epilepticus.
6. What is the definition of a simple febrile seizure?
7. Describe the management of febrile seizure.
8. List 5 reasons for CT Head after seizure and describe management after the 1st peds seizure.
    a. Which patients should have outpatient imaging and neurology follow-up?
    b. Which children with seizure should be admitted to hospital?
9. List 10 differential diagnoses for headache in peds.

10. List 8 indications for radiologic imaging in patients with headache.
11. With regards to presentation and management, how are migraines different in children?
12. Describe the criteria which define migraine headache (review).
13. List 10 causes of pediatric ataxia.
14. Describe an approach to the pediatric patient with ataxia.
15. List 5 central and 5 peripheral causes of vertigo. Which is the more common cause of vertigo in children?
16. List 8 risk factors for pediatric stroke.

Show Notes only:

- Spinal Cord Disorders
- GBS
- Myasthenia Gravis

Wisecracks

1. Describe each of the following:
    a. Infantile Spasms
    b. Absence Epilepsy
    c. Benign Rolandic Epilepsy of Childhood
    d. Lennox-Gastaut Syndrome
2. What is the most common cause of status epilepticus in children? In adults?
3. List five side effects of therapeutic dilantin use.
4. When is LP indicated in children with febrile seizures?
5. Give causes of acute, acute recurrent, chronic progressive and chronic non-progressive headaches.
6. Describe the presentation of infantile botulism.

Rosen's In Perspective:

This chapter includes the following pediatric neurologic disorders—altered mental status, seizures, headache, ataxia and derangements of balance and motor dysfunction. In this RIP, we want to quickly touch on "altered mental status in children". It's a topic covered in this chapter that we've discussed in the past.

Really, the key point is keep a BROAD DIFFERENTIAL! Use an approach like VITAMIN C&D, AEIOUTIPS, DIMES, or if it is the neonate: think through THE MISFITS:

- Vascular events (e.g., stroke, arteriovenous malformation with bleed)
- Infection (e.g., meningitis, sepsis, encephalitis)
- Trauma
- Toxic ingestion
- Anatomic or structural abnormality (e.g., intracranial mass or tumor)
- Metabolic derangements (e.g., DKA, hypoglycemia, urea cycle defect)
- Intussusception
- Seizures

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It is important to note that we will not be covering pediatric CNS infections in this podcast. However, we have included a couple of tables that are will act as a small refresher.

Check out Episode 109 on CNS infections for a deep discussion on pediatric meningitis/encephalitis.

Also, check out Tables 174.2 and 174.3 in Rosen's 9th Edition for a more comprehensive review of causative agents of pediatric bacterial meningitis and their associated antibiotic therapy.

Bacterial Meningitis: Common Causative Pathogens by Age	
Age Group	**Pathogens**
Newborns	GBS, E. Coli, Listeria
Infants and Children	Strep. pneumo, N. meningitidis, H. Flu B
Adolescents and Young Adults	Strep. pneumo, N. meningitidis

Bacterial Meningitis: Initial Empirical Antibiotic Coverage by Age	
Age Group	**Pathogens**
0-28 days	Ampicillin PLUS gentamycin OR cefotaxime
28 days – 3 months	Ampicillin OR vancomycin PLUS cefotaxime OR ceftriaxone
>/3 months	Cefotaxime OR ceftriaxone PLUS vancomycin

Core Questions

[1] List ten causes of provoked (acutely symptomatic) seizures.

To review, let's talk about the classifications for symptomatic seizures:

REMEMBER: A seizure is defined as a paroxysmal event characterized by temporary involuntary changes in the patient caused by abnormal and excessive activity of a group of cortical neurons.

Classifications of Seizures	
Type:	**Sub-Type:**
Generalized	Tonic-clonic Absence Myoclonic Clonic Tonic Atonic
Focal	Simple Partial (normal mental status) Complex Partial (altered mental status)
Unknown	Epileptic Spasms

For a more comprehensive classification of seizures, please refer to Table 174.4 in Rosen's 9th Edition.

Provoked seizures are caused by an identifiable trigger and stem from a broad array of disturbances, including fever, metabolic derangements, and trauma. Unprovoked seizures have no clear immediate precedent. Epilepsy is commonly defined as the occurrence of two or more unprovoked seizures.

DIMES for anything related to altered LOC:

- **Fever**
  - **Infection**
    - Don't forget about atypical febrile seizures (no fever)
    - Don't forget about gastrointestinal-related seizures
  - **Hemorrhage**
- **Toxin**
  - **Drug intoxication or withdrawal**
- **Metabolic**
  - **Electrolytes**
  - **Inborn-EOM**
  - **Liver/renal disease**
- **Neoplastic**
- **Vascular**
  - **AVM rupture**
- **Degenerative**
  - **Hypoxia**

For a more comprehensive list of causes for acute symptomatic seizures, please refer to Table 174.5 in Rosen's 9th Edition.

[2] List four episodic disorders that may mimic seizures in neonates and four in non-neonates.

Neonates:

- Jitteriness
- Benign neonatal sleep myoclonus
- Nonepileptic apnea
- Opisthotonos (hyperextension, back arching, spasticity - either physiologic or pathologic in cases of meningitis, tetanus)
- Normal movement

Non-neonates:

- Syncope
- BRUE
- Breath-holding spells
- Migraine with aura (vomiting, motor deficits, altered LOC)
- Sydenham's chorea
- Various sleep disorders - narcolepsy, cataplexy
- Tics

- Psychogenic non-epileptic seizures
- Panic attacks

For a more comprehensive list of mimics of epilepsy disorders, please refer to Table 174.6 in Rosen's 9th Edition.

[3] Give an approach to acute seizure control in a neonate and in a 5-year-old.

According to the Canadian Pediatric Society, the objectives for the acute management of CSE are as follows:

1. Maintenance of adequate airway, breathing and circulation (ABCs).
2. Termination of the seizure and prevention of recurrence.
3. Diagnosis and initial therapy of life-threatening causes of CSE (e.g., hypoglycemia, meningitis and cerebral space-occupying lesions).
4. Arrangement of appropriate referral for ongoing care or transport to a secondary or tertiary care centre.
5. Management of refractory status epilepticus (RSE).

Specific seizure control:

Let's talk about the drugs **- THIS ASSUMES YOU'VE EXCLUDED ANY CORRECTABLE METABOLIC CAUSES!!** (e.g. glucose < 2.6 mmol/L)

- **First line:**
  - **Lorazepam (IV/IO/Buccal/PR)**
    - **Dose**: 0.1 mg/kg, maximum one-time dose 4 mg
    - **Interval:** Every 5 minutes x 2
    - **Note:** STOP after two proper doses of benzo's (subsequent ones aren't effective and probably just lead to respiratory depression)
- **Second line:**
  - **Fosphenytoin/phenytoin**
    - **Dose:** 20 mg/kg
    - **Interval:** IV over 5-10 minutes (in NS or D5W)
  - **Phenobarbital**
    - **Dose:** 20 mg/kg
    - **Interval:** 1 mg/kg/min over 20 minutes
  - **Valproic acid**
    - Note: probably has a role, but not as fully researched in kids

How does it differ if you are unable to obtain IV or IO access?

Remember: Effectiveness (compared to IV) in descending order: IN >>> Buccal >> PR

- **IN route**
  - Midazolam
    - **Dose:** 0.2 mg/kg (max 5 mg/nostril)
- **IM route**

    - Midazolam
        - **Dose:** 0.2 mg/kg
    - Fosphenytoin
        - **Dose:** 20 mg/kg IM (max 1 g)
- **PR:**
    - Diazepam
        - **Dose:** 0.5 mg/kg
    - Lorazepam
        - **Dose:** 0.1 mg/kg
- **Buccal:**
    - Midazolam
        - **Dose:** 0.5 mg/kg
    - Lorazepam
        - **Dose:** 0.1 mg/kg

Pro tip: just memorize lorazepam 0.1 mg/kg for any route except IM or IN; in which case its 0.2 mg/kg for midazolam

Note: "During the administration of medications, pulse rate, respiratory rate, BP, cardiac monitoring and oxygen saturation via pulse oximeter should be followed on a regular basis. Anticonvulsant medications may cause loss of airway reflexes, respiratory depression, hypotension and cardiac arrhythmias**." - CPS Statement.**

[4] What is the definition of status epilepticus?

Remember: status epilepticus is typically defined as:

- 5 minutes or more of continuous seizure activity (clinical or electroencephalographic)
- Recurrent seizure activity without return to baseline between seizures.

NOTE: The number one cause of status epilepticus is a febrile illness**.**

NOTE: Never forget about non-convulsive status epilepticus. Non- convulsive status epilepticus is marked by an altered mental status. Patients may demonstrate confusion, unresponsiveness, abnormal motor movements, twitches, lip smacking, or automatisms. An electroencephalogram (EEG) can confirm the diagnosis and should be obtained if non-convulsive status is suspected.

[5] List 6 non-pharm. and six pharm. treatments for status epilepticus.

1. Ensure the patient stays safe (fall prevention; clear the area of hard objects; don't put anything in the mouth!)
2. Maintain ABC's
    a. Side lying if secretions or vomit; on the back if assisting with ventilation
    b. Suction supplies ready
    c. NPA
    d. Apply oxygen via non-rebreather or BVM if supporting ventilation
    e. End tidal C02 if available
3. Managing bradycardia, hypotension or delayed capillary refill with volume resuscitation and oxygenation/ventilation
4. Terminate the seizure

   a. In animal models, ischemic and excitotoxic neuronal cell loss starts to occur after 30 min of seizure activity. Seizures that last longer than 5 min to 10 min are at high risk of continuing for at least 30 min, so early treatment is associated with the best outcome. - CPS Statement
5. Search for and correct any reversible provoking triggers - hypoglycemia, hyponatremia, hypocalcemia

Order of preference:

1. 1st line: Benzos
2. 2nd line: Fosphenytoin (or phenobarbital if patient is already on phenytoin)
   a. Fosphenytoin can be given 3x faster than phenytoin and has fewer side effects; can be given IM
   b. Phenobarbital has greater risks for apnea, depressed consciousness, and hypotension,
3. If refractory to a first line agent and a second line agent = you have refractory status epilepticus*; consider these agents (in consultation with PICU):
   a. Phenobarbital
   b. Pentobarbital
   c. Midazolam infusion (post-intubation)
   d. Levetiracetam
   e. Propofol
   f. Topiramate
4. Full laboratory investigations for metabolic abnormalities
5. Treatment of hyperthermia

NOTE: CSE that is unresponsive to two different antiepileptic medications (e.g., a benzodiazepine and phenytoin) is considered to be refractory, although some authorities have added a duration criterion such as longer than 30 min or longer than 60 min.

[6] What is the definition of a simple febrile seizure?

Definition: generalized seizure occurring in the presence of a fever without CNS infection.

- Occurring in a developmentally and neurologically normal child
- Less than 15 minutes
- Between 6 months and 5 years
- Normal neurologic examination pre and post-seizure

What makes a simple febrile seizure complex:

- Multiple seizures occurring within 24 hours
- Seizure lasts longer than 15 minutes
- Seizure occurs outside of normal age range
- Focal component to seizure.

NOTE: Regardless of the seizure being febrile, we should always consider whether there is an acute provoked cause of the seizure. Overall, 33% of children who have a febrile seizure will have another one.

NOTE: Temperature does not need to be markedly elevated; almost 50% of children with a febrile seizure have a documented temperature below 39°C.

NOTE: Meningitis should be considered in any patient with seizures and fever, although a child whose mental status is normal before and after the seizure is very unlikely to have meningitis. We recommend evaluation and treatment for meningitis in infants younger than 3 months presenting with febrile seizures. They are not only at higher risk for serious bacterial infections, including meningitis, but their mental status is difficult to assess accurately.

[7] Describe the management of febrile seizure.

NOTE: The acute management is similar to most other seizures!

According to the CPS:

The objectives for the acute management of CSE are as follows:

1. Maintenance of adequate airway, breathing and circulation (ABCs).
2. Termination of the seizure [intervene if > 5 mins] and prevention of recurrence.
3. Diagnosis and initial therapy of life-threatening causes of CSE (e.g., hypoglycemia, meningitis and cerebral space-occupying lesions).

NOTE: Electroencephalography and neuroimaging generally are not required after a first simple febrile seizure.

[8] List 5 reasons for CT Head after seizure and describe management after the 1st peds seizure.

NOTE: According to CPS: “history of trauma, evidence of increased ICP, focal neurological signs, unexplained loss of consciousness or suspicion of cerebral herniation are some of the indications for a computed tomography (CT) scan of the head.”

You may be asking about which patients should have outpatient imaging and neurology follow-up:

ANSWER: Most kids who need imaging - need it in the ER (or admission); EEG’s are a less urgent investigation.

NOTE: According to UptoDate: “A CT scan is most often used for emergency department assessment. In the setting of a first seizure, a non-contrast CT scan reveals findings that change acute management in 3 to 9 percent of children and infants Among children with a first seizure that is apparently unprovoked, the risk of an urgent finding may be even lower: in a prospective multicenter study of 475 children (age 1 month to 18 years) with an unprovoked first seizure, CT or MRI identified an emergent/urgent finding in 0.8 percent of patients and non-urgent findings in an additional 10 percent.”

You may also be asking which seizures warrant EEG’s:

ANSWER: Complex febrile seizures, new unprovoked seizures, recurrent unprovoked seizures

Which children with seizure should be admitted to hospital?

ANSWER: New focal neurologic deficits, persistent altered mental status, recent trauma, persistent headache, or partial seizures, neonatal seizures, high parental anxiety.

NOTE: Infants and children who present in status epilepticus not related to fever should have neuroimaging.

[9] List 10 ddx for HA in peds.

By age 15, 75% of children will have experienced a headache; most of these headaches are benign primary headache syndromes.

REMEMBER: Most of the pediatric headaches are migraines.

Migraine headaches likely come about as a result of an interaction between genetic redisposition and environmental stressors. The mechanism of migraine headaches is thought to involve a primary dysfunction of the brain in which a wave of spreading cortical neuronal depression is accompanied by vascular changes. Derangement of the trigeminovascular reflex results in alterations of regional blood flow, and this neurovascular interaction is thought to contribute to neurogenic inflammation and the development of migraine headaches.

The primary goal of the ED evaluation is to differentiate life-threatening causes of headaches from primary headaches, such as migraine or tension headaches. A few different terms are used to characterize headaches: acute, acute recurrent, chronic non-progressive, chronic progressive, and mixed.

REMEMBER: The child's history is the most important component to an accurate diagnosis

Primary headache causes:

- Migraine headache
- Tension headache
- Cluster headache
- Chronic daily headache

Secondary headache causes:

- Trauma (e.g., intracranial bleed, concussion, skull fracture)
- Structural (e.g., neoplasm, AVM)
- Systemic (e.g., hypertension, metabolic)
- Infection (e.g., meningitis, abscess, etc…)
- Toxic (e.g., medication ingestion)

For a more comprehensive list of causes for secondary headaches in pediatric populations, please refer to Table 174.8 in Rosen's 9th Edition.

Remember: In the absence of other signs of CNS involvement (e.g., nuchal rigidity, alteration in level of consciousness, focal neurologic findings), headaches in febrile children usually do not constitute evidence of CNS infection. Non-specific viral illnesses represent most diagnoses in children presenting to the ED with an acute headache.

[10] List 8 indications for radiologic imaging in patients with HA.

For a more comprehensive list of indications for radiologic imaging of patients with headaches, please refer to Box 174.3 in Rosen's 9th Edition

Indications for Radiologic Imaging in Patients with Headache	
Strongly Indicated if:	• Abnormal neurologic examination • Signs and symptoms of elevated ICP • Meningeal signs PLUS focal neurological findings or altered mental status • Significant head trauma • Progressive neurological deficits • Significant head trauma • Severe nocturnal headache waking the patient from sleep or is present upon waking • Severe headaches • Chronic progressive worsening headache • Presence of VP shunt
Consider if:	• Headache or vomiting upon waking • Unvarying location of headache, especially if occipital • Persistent headache with no family Hx of migraines • Neurocutaneous syndrome • Age<3

While the obvious physical exam findings will prompt the average health care worker to get imaging; some of these things are easily missed if you aren't doing a FULL deliberate history!

REMEMBER: Red Flags

- History of head trauma
- Symptoms of increased ICP - Valsalva/cough/strain
- Awakening from sleep
- Chronic progressive h/a
- Headache on awakening
- Occipital h/a
- No family headache of migraines

[11] With regards to presentation and management, how are migraines different in children?

ANSWER:

- Pediatric migraine headaches are more commonly bilateral
- Atypical or migraine variants are more common in children:
    - Hemiplegic migraine
    - Ophthalmoplegic migraine
    - Basilar migraine
    - "Alice in wonderland syndrome" (acute confusional state)
    - Ocular migraine
    - Abdominal migraine

[12] Describe the criteria which define migraine headache.

Migraine without an aura:

Also known as common migraine, is the most frequent type of pediatric and adolescent migraine and includes the following criteria: more than five attacks that last 2 to 72 hours (untreated or unsuccessfully treated), accompanied by nausea, vomiting, photophobia or phonophobia—and including a minimum of two of the following criteria: unilateral or bilateral location, pulsing quality, moderate to severe intensity, and aggravated by routine physical activities.

Migraine with an aura:

Previously known as classic migraine, is diagnosed when at least two attacks fulfilling the diagnosis of migraine occur accompanied by a variety of sensory warning symptoms, such as flickering lights, loss of vision, and tingling or numbness. The aura typically develops over 5 or more minutes and completely resolve within 60 minutes.

[13] List 10 causes of pediatric ataxia.

Definition: abnormal modulation or organization of movement; can be congenital or acquired

Congenital ataxia is associated with CNS abnormalities. Acquired ataxia can be acute, episodic, or chronic. The chronic ataxias are usually caused by inherited metabolic or genetic disorders. Usually, ataxia is caused by cerebellar dysfunction, but lesions in the corticospinal tract or dorsal columns of the spinal cord may also be causative.

In children, 40% of ataxia cases are caused by acute cerebellar ataxia:

- Boys are predominantly afflicted at 2-4 years of life
- A history of recent illness with multiple causative agents is seen in 70% of patients, but Varicella virus is the most common, associated with up to 26% of cases. The disease is thought to be due to an autoimmune phenomenon leading to cerebellar demyelination.

Other causes of childhood ataxia include:

- Acute post-infectious demyelinating encephalomyelitis
- Brainstem encephalitis
- Drug ingestion
- Guillain-Barré Syndrome
- Metabolic disorders
- Aminoacidopathies
- Mitochondrial disorders
- Organic acidopathies
- Urea cycle disorders
- Migraine headaches
- MS
- Neoplasm
- Opsoclonus-myoclonus syndrome
- Recurrent and chronic genetic ataxias
- Seizure
- Stroke
- Vertebral artery dissection

[14] Describe an approach to the pediatric patient with ataxia.

Usually we'll see these kids in the first few days after onset, usually because of a refusal to walk, unsteadiness of arm movements, or sudden development of a wide-based so-called drunken gait.

REMEMBER: Diagnosis is in the HX and Physical!

Key questions to ask:

- Does this child or family have any congenital or family history of ataxia?
- Does this kid have normal mental status?
    - If yes:
        - Ask about any recent viral infections
    - If no:
        - Be worried!
        - Differential and workup needs to include:
            - Toxic ingestions / overdoses
                - Up to 32% of cases of acute childhood ataxia are due to drug toxicity, usually anticonvulsants, benzodiazepines, alcohol, or antihistamines or, less commonly, from exposure to organic chemicals or heavy metals.
            - Meningitis / encephalitis
            - Hydrocephalus
            - CNS lesions
                - Post trauma / neck dissection
                - Malignancy
                    - Approximately 45% to 60% of all childhood brain tumors arise in the brainstem or cerebellum and can be manifested with slowly progressive ataxia.

        - Acute decompensation can occur, with the development of hydrocephalus or hemorrhage into the lesion.
    - Strokes
    - Acute post-infectious disseminated demyelinating encephalomyelitis,
        - Alteration in consciousness and multifocal neurologic deficits, as well as by fever and frequent occurrence of seizures.

NOTE: Urine and serum toxicology studies are the highest yield laboratory studies. CT and MRI findings are usually normal in patients with post-infectious ataxia, but demyelination, tumor, hydrocephalus, or traumatic injuries may be identified. CSF analysis may show mild pleocytosis or lymphocytosis in acute post-infectious ataxia; findings are normal in most other cases.

[15] List 5 central and 5 peripheral causes of vertigo. Which is the more common cause of vertigo in children?

REMEMBER: Vertigo = illusion of movement**.** Disease processes that effect the balance of the vestibular, visual, and proprioceptive systems can cause vertigo by impairing the neural activity of the vestibular nucleus. Diseases of the ear, eighth cranial nerve, neck, brainstem, or eye can lead to vertiginous symptoms. Vertigo is characterized as central or peripheral, depending on whether the cause is in the CNS.

NOTE: Key things to ask about on history:

- Ear symptoms: such as otalgia, hearing loss, and tinnitus
- Other important historical features that should be determined include headache, loss of consciousness, head trauma or barotrauma, and family history of migraine or seizure disorders

Patients can be divided into those who have hearing loss and those who have normal hearing. In the group with hearing loss, further characterization of the loss as conductive or sensorineural (using the Weber and Rinne tests) can help localize the peripheral lesion from the middle ear, labyrinth, or eighth cranial nerve.

The differential diagnosis for vertigo is as follows:

- **Central** - think infectious, inflammatory, vascular, neoplastic
    - AVM's
    - Brain abscess
    - Chiari malformations
    - Encephalitis
    - Meningitis
    - Seizures
    - Trauma

- **Peripheral** - think outside in: wax, cholesteatoma, OM, labyrinthine concussion/infection, vestibular neuronitis, congenital, benign

    - BPPV
    - Cholesteatoma
    - DM
    - Otitis media
    - Vestibular neuronitis

For a more comprehensive list of differential diagnoses for central and peripheral vertigo, please refer to Box 174.5 in Rosen's 9th Edition

NOTE: A few pediatric-specific disease entities to know about:

- The most frequent cause of benign paroxysmal vertigo of childhood is a migraine headache, with vertigo occurring as the aura of an episode.
- Patients with basilar artery migraines also present with vertigo, hemiparesis, ataxia, palsies of the third, sixth, or seventh cranial nerve, drop attacks, and blindness in various combinations, followed by migraine headache.
- Benign paroxysmal positional vertigo is rare in children,but can occur spontaneously as well as after trauma. The earliest age at which it has been reported is 11 years. It is believed to be due to otoliths that have moved out of their normal positions in the utricle and is corrected by canalith repositioning maneuvers (e.g., Epley maneuver).
- Ménière's disease, a syndrome with a combination of vertigo, fluctuating hearing loss, and tinnitus, is responsible for 1.5% to 4% of cases of pediatric vertigo.
- Vestibular neuronitis, thought to be caused by viral infections, is manifested as vertigo without hearing loss. A preceding cold is found in 60% of patients. It is manifested with severe vertigo that resolves in a few days, they can get persistent vertigo with rapid head position changes lasting a few months until their brain adapts to it.
- Labyrinthitis is an inflammatory process involving the inner ear membranous labyrinth; it manifests with vertigo, hearing loss, and tinnitus. Cytomegalovirus, rubella virus, and rubeola viruses are common causative agents. Bacterial labyrinthitis usually occurs in association with meningitis and should be suspected in any ill child with vertigo and high fevers, especially in combination with a perforated tympanic membrane.
- Neurofibromatosis can be manifested with vertigo if it involves the superior vestibular nerve.
- Other genetic syndromes such as Alport syndrome are also associated with vertigo
- Ototoxic drugs are also associated with vertigo
- Cranial nerve deficits associated with vertigo may indicate a brainstem lesion or tumor. Vertigo is the presenting symptom in 5% to 12% of cases of multiple sclerosis.

Laboratory tests in the vertiginous patient should be dictated by the history and physical examination and may include glucose and electrolyte level assessment, thyroid function tests, and viral titers or serologic studies (e.g., for Lyme disease or syphilis). CT or MRI is indicated for patients for whom an underlying CNS abnormality (central vertigo) is suspected.

Management of the vertiginous patient depends on the underlying cause, which may not be evident in the ED. For acute symptomatic relief, vestibular suppressants such as meclizine and diazepam may be helpful.

NOTE: Of all these, which are the most common causes of vertigo in children?

1. Otitis media
2. Migraine Disorders

[16] List 8 risk factors for pediatric stroke.

REMEMBER: Strokes in children are rare; unlike adults the incidence of hemorrhagic and ischemic strokes is roughly equal - 50-50. (~1 in 100k). SAH is also much less common (0.4 in 100k) and is often aneurysmal.

One could think categorically about the common aetiologies and risk factors of ischemic stroke, as recommended by UpToDate:

- Cardiac abnormalities
- Vascular lesions
- Hematologic abnormalities
- Infection
- Head and neck trauma
- Genetic conditions.

Additionally, you could also break it down with Virchow's Triangle:

- Venous stasis
- Endothelial injury
- Hypercoagulable

Or, you could consider the risk factors for stroke in pediatric patients by thinking about common causes:

- Sickle cell disease (puts you at risk for BOTH hemorrhagic and ischemic strokes)
    - This is the most cause of strokes in children
- Structural heart disease (embolic vs. complication of anticoagulation)
- Moyamoya disease

For the hemorrhagic strokes, typically these are due to:

- Ruptured AVM's
- Hematologic abnormalities (including thrombocytopenia or platelet dysfunction, hemophilia and other congenital or acquired coagulopathies, and hemoglobinopathies)

For a more comprehensive list of the risk factors for pediatric stroke, please refer to Box 174.6 in Rosen's 9th Edition

Show Notes Only:

[1] Spinal Cord Disorders

REMEMBER: Think of intrinsic (e.g. viral-induced transverse myelitis) vs. extrinsic spinal cord disorders (e.g. epidural abscess)

Clinical findings:

- Paraplegia
- Hyporeflexia
- Sensory deficits
- Complete sensory loss or paresthesias below the level of the spinal cord lesion(s)
- Also, if the distal portion of the cord is affected, patients will have bowel and bladder incontinence.

Management: Based on the underlying process - surgery vs antibiotics vs steroids.

[2] Guillain- Barré Syndrome

Definition: An acute, demyelinating polyneuropathy that typically presents as transient, symmetric, ascending paralysis in the setting of a recent infection.

It is thought to be autoimmune-mediated and classically causes demyelination of motor and sensory nerves. Children of all ages may be affected; however, it is uncommon in young toddlers and infants. Often, there is a history of a preceding minor viral or gastrointestinal illness in the weeks prior to presentation.

NOTE: Campylobacter jejuni is the most common infectious agent associated with GBS.

Management: Watch for rapid respiratory deterioration; IVIG or plasma exchange.

[3] Myasthenia Gravis

Definition: Myasthenia gravis is an autoimmune disorder characterized by autoantibodies directed against the acetylcholine receptor of the neuromuscular junction. This action produces intermittent and fatigable weakness. Myasthenia gravis is usually seen in adults; however, there are three types that affect children—neonatal (transient), congenital, and juvenile.

REMEMBER: MG has a waxing and waning weakness, worsens as the day goes on and often presents with bilateral ptosis.

The Tensilon test may be used in the ED to confirm the diagnosis. Edrophonium (Tensilon), an acetylcholinesterase inhibitor, is administered IV. By blocking the action of acetylcholinesterase at the neuromuscular junction, the presence of acetylcholine is prolonged, and muscle weakness transiently improves. Atropine should be available during the administration of edrophonium to treat possible cholinergic reactions (e.g.,bradycardia).

NOTE: Watch for the myasthenic crisis precipitated by an acute stressor (may need intubation!) [see CRACKCast episode on neuromuscular disease].

Management: Oral cholinesterase inhibitor therapy is the maintenance therapy.

Wisecracks

[1] Describe each of the following:

- **Infantile Spasms**
    - UptoDate defines infantile spasms (IS) is an age-specific convulsive disorder of infancy and early childhood.
    - Most children with IS present between three and seven months of age; onset after 18 months is rare.
    - Spasms are symmetric contractions of flexor or extensor axial or limb muscles. They vary in pattern, intensity, duration and extent. Most spasms occur in clusters of two to more than 100 over one to several minutes.
    - EEG is required to make the diagnosis. - will show hypsarrhythmia (very disorganized EEG rhythm) Management is controversial and complex. Prognosis is poor.

- **Absence Epilepsy**
    - According to UptoDate, the hallmark seizure type is the typical absence seizure (TAS), characterized by profound impairment of consciousness, abrupt in onset and termination, lasting an average of 10 seconds.
    - TAS are frequently associated with early arrest in activity, staring, repetitive eyelid movements, and automatisms.
    - Usually occurs in children 4 - 10 yrs of age.
    - The overall prognosis of [childhood absence epilepsy] CAE is favorable. In most cases, seizures respond well to the first-line drug monotherapy and remit before puberty, without cognitive sequelae.

- **Benign Rolandic Epilepsy of Childhood**
    - Benign (childhood) epilepsy with centrotemporal spikes (BCECTS or BECTS, also called benign Rolandic epilepsy) is the most common, presenting at a mean age of eight years, with focal motor or secondarily generalized seizures that occur mostly at night.
    - These kids usually outgrow their seizures by adolescence.

- **Lennox-Gastaut Syndrome**
    - According to UptoDate:
    - Benign occipital epilepsy of childhood (Gastaut syndrome) presents at a mean age of nine years with seizures that have prominent visual symptoms including blindness and hallucinations.
    - Seizures usually occur during the daytime and are brief, but can be frequent, indicating antiseizure drug treatment in most individuals. EEG demonstrates

occipital spikes and waves that are activated by eye closure. Seizures often remit prior to adulthood

[2] What is the most common cause of status epilepticus in children? In adults?

Children:

- Febrile status epilepticus (~30%)

Adults:

- Acute symptomatic causes (> 50%)
    - Structural brain lesion (acute or longstanding)
    - Toxic cause
    - Metabolic cause
- Remote symptomatic causes / low antiepileptic drug levels

[3] List five side effects of therapeutic Dilantin use.

Long term:

- Gum hyperplasia
- Facial coarsening
- Neurologic changes: confusion, slurred speech, diplopia, ataxia
- Rash

Rare but serious s/e:

- Agranulocytosis
- SJS/TEN
- Aplastic anemia
- Hepatic failure
- Dermatitis/rash
- Serum sickness
- Adenopathy
- Pseudolymphoma
- Neuropathy
- Ataxia
- Lupus syndrome
- Hirsutism

[4] When is LP indicated in children with febrile seizures?

NOTE: According to newer AAP recommendations, LP is not necessary in children older than 12 to 18 months in whom clinical findings are not suggestive of meningitis.

In immunized infants 6 to 18 months of age with a first-time simple febrile seizure, LP can be avoided if the child is not ill-appearing, has returned to baseline, and there are no clinical signs of meningitis.

Additional consideration of the need for LP should be made for children:

- Between 6 and 12 months of age whose vaccination status for H. influenzae type b or Streptococcus pneumoniae is incomplete
- In children who have been pretreated with antibiotics.
- Patients presenting with unprovoked seizures who demonstrate persistent abnormal mental status, do not return to baseline, or show signs of meningitis.

[5] Give causes of acute, acute recurrent, chronic progressive and chronic non-progressive headaches.

Acute Headaches:

- AVM's
- Acute intracranial bleed
- Sinusitis
- Otitis media
- Traumatic head injury

Acute Recurrent Headaches:

- Cluster Headaches

Chronic Progressive Headaches:

- Intracranial neoplasms
- Pseudotumor cerebri
- Hydrocephalus
- Brain abscess

Chronic Non-progressive Headache:

- Muscle Contraction Headache
- Conversion Headache

[6] Describe the presentation of infantile botulism.

REMEMBER: Infant botulism typically affects infants younger than 6 to 8 months. It results from intestinal colonization with Clostridium botulinum. A neurotoxin produced by C. botulinum impairs acetylcholine release from the presynaptic membrane, thereby affecting skeletal muscle, smooth muscle, and autonomic function.

It usually has an insidious onset, with progressive symptoms including poor suck, lethargy, and hypotonia. Cranial nerve exam may also be abnormal. Infants develop constipation and poor feeding, with subsequent hypotonia and weakness, which may require respiratory support.

Sources:

- Unpasteurized honey (potential reservoir for C. botulinum)
- Most US cases of infant botulism are thought to arise from ingestion of environmental dust particles containing C. botulinum spores and may be associated with active construction areas in which there is disruption of the ground.
    - Infants have higher stomach pH and so these spores are not killed as they are in adults - which is why honey is contraindicated in infants < 1 yo

Diagnosis:

The diagnosis of botulism may be confirmed through isolation of botulinum toxin in the stool; EMG's can also confirm the dx.

Management:

- Supportive care
- Preparation in case of invasive ventilation requirements
- BABY BIG
    - Initial dosing for botulism immunoglobulin in infants less than 1 year of age is 50 mg/kg of body weight in a single IV infusion.