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**Foundation for the Education and Research in
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***Challenging Pediatric
Neurological Emergency Patients***

Generalized Weakness in a Ten Month-old Infant

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Case Presentation

After her parents had noted some progressive weakness over the preceding twenty-four hours, a ten-month-old female infant was brought in to her pediatrician for evaluation. Other symptoms included drooling, difficulty latching onto her mother's breast, a poor suckle, and "droopy" eyelids. Due to a history of a recent upper respiratory infection and a physical exam that was significant only for a serous otitis media, the pediatrician's working diagnosis was viral syndrome. She was discharged to home on amoxicillin, but presented that evening to the emergency department with her parents because she was no longer moving her right hand.

The infant had not been previously hospitalized but had required treatment for frequent ear infections, with her last course of amoxicillin having been completed one week prior to presentation. Her only other recent illness had been an episode of croup in the past two months. She was the product of a term pregnancy and born via spontaneous vaginal delivery, without complications. Other than the amoxicillin, she was on no other prescription or over the counter medications. She had not been given any herbal preparations. She lived with her parents and an older sibling, who had also recently recovered from a similar upper respiratory infection. There were no pets in the household and no history of recent travel or camping. Their house was undergoing a remodel and waterline

construction had been taking place outside their home for approximately three weeks. Immunizations were up to date, and a review of systems was negative except for some “loose” stools.

On physical exam, vital signs included the following: a rectal temperature of 36.6 degrees Celsius, pulse 147, respiratory rate 34, with an oxygen saturation of 99% on room air, and a blood pressure of 126/73. On her neurological examination, her face was symmetrical and gag reflex was intact. She would reach toward an object with her right arm but could not move any of the digits of her hand. She was only able to hold objects briefly in her left hand. In addition she had difficulty sitting and poor head control. Bicep and patellar tendon reflexes were 2+ and symmetrical. Other than bilateral middle ear effusions, there were no other significant findings, with no evidence of respiratory compromise, focal infection, or trauma.

Laboratory studies included a complete blood count, basic metabolic panel, a C-reactive protein, and spinal fluid analysis. Blood, urine, stool, and cerebral spinal fluid cultures were obtained. The results were notable only for a white blood cell count of 13,000 and a CO₂ of 17. Computed tomography of the head without contrast did not reveal any evidence of radiographic abnormalities.

The infant was admitted to the neurology inpatient service but continued to have deterioration in her clinical status over the next forty-eight hours. She became increasingly weak to the point that she lost her gag reflex and required feedings via a nasogastric tube. She developed increasing hypotonia, loss of facial expressions, and ptosis. Magnetic resonance imaging of her head was normal. After diagnostic studies were obtained, definitive treatment was initiated.

A Case of Whole Body Pain in a 12 Year-old Girl

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Case Presentation

M.E. is a 12 year old Caucasian female who presents to the emergency department accompanied by her parents with a chief complaint of pain over her entire body.

Patient notes that the pain in her body began one week ago, and was initially only in her toes, and fingers, and gradually progressed to now involve all four extremities, her trunk, but spares her head/neck/face. History is difficult to obtain, as the patient is extremely tearful and requesting pain medication throughout the history. Pain is described as intense, and has a burning quality. Her parents note that she has stayed home from school for the past 4 days due to the pain. She states she has trouble walking, but when asked regarding weakness, she denies it. No bowel or bladder changes.

PMH: No serious medical problems.
Her mother notes a recent gastrointestinal illness lasting 2-3 days 6 weeks ago.

Meds: none, NKDA

Social history: parents state that they recently moved, and she has been having some difficulty adjusting to her new school. They didn't bring her to a doctor earlier because they thought she just needed a "break" from school.

Family history: negative

ROS: Pt denies nausea/vomiting. When asked, she states that she sometimes "can't get her breath", especially when the pain is bad. No chest pain, no rashes, no fever/chills, no urinary symptoms.

Physical exam: VS: BP 98/60, HR 110, temp 98.9

1. In general, very tearful young female, some regressive behaviors noted (clinging to mom, refusing to cooperate)
2. Pulmonary: lungs clear to auscultation bilaterally
3. Cardiovascular: S1, S2, no rubs/murmurs/gallops
4. Abdomen: soft, non-distended, non-tender
5. Extremities: no clubbing, cyanosis, or edema

Edward P. Sloan, MD, MPH

6. Neuro: Cranial nerves: Extraocular movements intact, pupils equal round and reactive to light. Facial sensation and strength intact. Sensory exam notable for extreme pain to even cotton wisp sensation in lower extremities which the patient unable to tolerate, and the pain involved the trunk as well as extremities. Motor exam limited by patient effort, but appeared to have full strength in all extremities. Tone examination was normal or slightly decreased. Deep tendon reflexes could not be obtained. Patient refused to walk.

Laboratory examinations:

1. CBC, renal panel normal, mildly increased transaminases noted, with normal cholestatic parameters
2. Chest x-ray normal

The patient was sent home from the emergency department, with the diagnosis viral syndrome and with follow-up planned by her pediatrician within a week. However, 2 days later, the patient returned with complaints of profound weakness, and shortness of breath. Her repeat examination is as follows:

VS: BP 130/78 HR 125 temp 100.6

1. General exam unchanged from previous exam, except that the patient appears extremely short of breath.
2. Neuro exam: Cranial nerves: pupils equal, round and reactive, extraocular movements intact. Facial strength is decreased bilaterally, and patient has difficulty closing her eyes. Sensory exam: pt has less sensitivity to light touch than previous exam, but still has burning pain to light touch in all 4 extremities and her trunk. Motor exam: bilateral lower extremities were flaccid, with no movement noted. Upper extremities were extremely weak, and the patient was unable to raise arms off of the bed. Tone was markedly decreased in all 4 extremities, and deep tendon reflexes could not be obtained.

What is your differential diagnosis and how would you proceed?

Headache with Right Upper Extremity Weakness and Dysphasia in a 10 Year-old Boy

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Case Presentation

A 10-year-old boy was brought to the emergency department (ED) for evaluation of headache, garbled speech, and right upper extremity weakness. One half hour prior to arrival the family had been at a shopping mall when the boy had suddenly grabbed his head, cried out that he had a terrible headache, and sank to the floor without striking his head. There was a questionable momentary loss of consciousness, but no motor activity or urinary incontinence. Immediately afterwards the parents noted that the boy was not using his right arm. He had two episodes of forceful, projectile vomiting. Parents noted that they could not understand his speech. The parents immediately transported him to the hospital.

The patient had no past medical history except for a heart murmur echocardiogram-proven to be due to a bicuspid aortic valve. There was no history of trauma, headaches, or drug use. There was no significant family history.

On initial ED evaluation the patient appeared alert and was in moderate distress. He was holding his forehead in his left hand. Vital signs were blood pressure 116/76, pulse 120, respiratory rate 18, oral temperature 97.6° C, and his skin was cool and dry. On physical examination there was no sign of trauma, pupils were equal and reactive to light, fundi had sharp disk margins, and the neck was supple and non-tender. Cardiac examination revealed a I/VI systolic ejection murmur, and the lungs were clear. The abdomen was soft, and there were no lesions on the skin. On neurological examination, cranial nerves II -XII were intact. Motor examination showed 5/5 strength in all muscles on the left side and the right leg, but there was 0/5 strength in all the muscles groups in the right upper extremity. Deep tendon reflexes were +2 and symmetrical; toes were down going. Sensation was intact (localized and withdrew from pin prick) There were no meningeal signs.

The patient could understand and carry out three-step commands; however he had difficulty with naming and repetition.

The patient was placed in a semi-prone position with his head elevated at 45 degrees, and airway management equipment was readied at the bedside. He was placed on oxygen and an ECG monitor showed normal sinus rhythm. An 18-gauge intravenous line of 5% dextrose in 0.45% normal saline was placed in the left antecubital vein; when the intravenous line was started, blood was obtained for a bedside glucose determination, which registered 80 mg / dL. Blood was sent for a complete blood count, electrolytes, calcium, magnesium, BUN, creatinine, glucose, and PT, PTT. Urine was sent for a toxicology screen looking specifically for phencyclidine, cocaine, and amphetamines. Arrangements were made for a stat head computed topography (CT) scan.

The patient remained stable over the first hour in the emergency department except for one more episode of vomiting. His expressive dysphasia persisted. The arm weakness began to resolve and by the time he was sent for the CT scan his strength had improved to 3/5. All of the laboratory tests returned within normal limits except for the toxicology screen and sedimentation rate which were still pending.

A 16 Year Old Male That “Can’t See & Can’t Hear”

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Case Presentation

A 16 year old male presents to the pediatric emergency department 1 day PTA with the chief complaint “I can’t see.” Per the initial triage assessment, the patient had grossly normal vision. He was able to walk independently. He could get up on the scale and find his way back to the waiting room without assistance. When asked to get up on the scale, the patient responded “Why do I need to get up on the scale?” The patient left the waiting room prior to a physician evaluation.

The patient represented to the ER 24 hours later accompanied by his cousin. His chief complaint was “can’t see, can’t hear.” Again, the patient was able to walk to the triage area without assistance and he was able to follow verbal cues. His vision and hearing seemed grossly intact.

In the treatment area, the patient denied recent illnesses. He denied nausea, vomiting and diarrhea. He denied fevers, chills, or recent URI's. He was eating and drinking, although much less over the past 24 hours. He admitted to occasional ETOH and marijuana use, but denied other illicit drug use. The patient's only complaint was that he could not see and could not hear. He could not elaborate any further. However, the patient could turn his head when asked in a normal tone of voice. He could see, identify, and pick up a pen when asked.

On exam, the patient was bright and alert. He was afebrile with a HR=72 and a BP=130/70 with a RR=14. His EOM were intact and without nystagmus. His pupils were mid-sized and reactive. His sclerae were muddy. His TM's were clear bilaterally. His neck was supple. His heart was RRR and without murmurs, rubs, or gallops. His lungs were clear to auscultation. His abdomen was soft and non-tender. His skin was without purpura, petechiae, or rashes. He had several tattoos on his arms and back. His extremities were well perfused with good cap refill. The patient demonstrated a normal gait and had equal strength throughout. It was unclear whether the patient was unable or unwilling to cooperate with further neurological exam. Directions needed to be repeated during the physician-patient encounter. The patient answered 'huh' and 'don't know' to most questions. He seldom spoke in full sentences. The patient appeared annoyed and frustrated with further encounters and serial exams. He stated 'going home.'

After the physical exam, the patient jumped off the gurney and found the sign marked restroom without assistance. Afterward, he walked out of the restroom and into the hallway. When asked why he was standing in the hallway, he replied "huh". When asked again, he replied "huh" while placing his cupped hand around his ear. He was asked the same question a third time and stated "waiting for mom." (His mother was called at home and asked to come to the ER). He was redirected to his bed and the patient fell asleep.

The cousin offered further history. The patient was in his normal state of health until 2 days PTA. The patient was arrested by the police for possession/smoking of marijuana. He was released in the custody of his mother in the early morning hours. The patient was not acting right, but the family thought that his behavior was secondary to his jail experience. 1 d PTA, the patient awoke complaining of "can't see". Over the next 24 hours, the patient also complained of headache and he was bumping into things at home. He was squinting and was intermittently covering his one eye, then covering the other eye. At times, he did not make sense when he talked. He spent the majority of the day in bed and was not acting like his normal self. Per the cousin his PMH/PSH was unremarkable. He had no allergies and took no medications. His social history revealed that he lived at home with his mother. He had one child. He does not do well in school because of frequent delinquencies but is otherwise developmentally normal. He smokes marijuana and drinks hard liquor occasionally. He had a new tattoo placed less than one month ago.

In summary, we have a 16 yr old male who was in his usual state of health until 1 day prior to admission. He complained of a headache and decreased hearing and vision. The cousin reports that the patient is not acting like his normal self, he is sleeping a lot, and sometimes he does not make sense when he talks.

What is the differential diagnosis?

“I Think My 17 Month-old Baby Is Drunk!”

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Case Presentation

A 17-month-old female was brought by her parents with the chief complaint of inability to walk without staggering and falling to the right. She had been seen at a small, community ED earlier that same day and was referred to our center for further work-up and evaluation. The patient had been in good health until three days prior, when she developed vomiting and diarrhea that lasted about 24-36 hours. On the day of presentation, the patient was being looked after by her grandmother, who noted that she was having difficulty ambulating and called the parents. They, too, noted her unsteady gait and observed that she was falling to the right. There was no known ingestion and no history of trauma. She was eating well, and her mental status appeared normal to the parents. The patient did not appear to be in pain per the mother.

The patient had no significant past medical history besides the gastrointestinal symptoms reported above. A fever had not been noted during the last week, and no medications had been given. She had a normal birth history with normal developmental milestones achieved. The parents keep no medications in the house, although the grandmother has some antihypertensive and some “back pills” at home, which she does not believe were accessed by the patient. Review of systems was negative except as noted above, and the patient’s immunizations were up to date.

On presentation, her vitals were: temperature 37.7 rectally, heart rate 144 while crying, respiratory rate 25, and capillary refill <2 seconds. In general, she was crying but appeared awake and alert, in no apparent distress, sitting on her mother’s lap, appropriately anxious at being in the ED. She was easily consoled by the parents. Her HEENT exam revealed no external evidence of head

trauma, normal TM's bilateral, and no discharge with moist mucous membranes and a supple neck. Her cardiopulmonary exam was unremarkable. Her abdomen was soft and non-tender, and her extremities were without bruises or deformity, and she had full range of motion in all joints without discomfort. Her skin was without rash or pallor, and skin turgor was normal. On neurological exam, she appeared to be moving all extremities freely, with grossly normal strength and sensation. When asked to reach and handle a small object with either hand, she was symmetric with no ataxia noted. Her eyes tracked normally without nystagmus and her face was grossly symmetric. When placed on her feet, she did not appear hesitant or in any pain, but on attempting to ambulate, she consistently staggered and fell to the right side several times into her mother's arms. She had no apparent pain during these episodes.

Routine laboratory studies were obtained and were as follows: WBC count 17,400 with a left shift but no bands; Hgb/Hct 12.2/36.8; electrolytes within normal limits; urinalysis negative. A head CT scan was performed and was read as negative; this was done without contrast. A lumbar puncture was grossly bloody with final cell counts of 1400 WBC and 240,000 RBC and a negative Gram stain. A urine toxicology screen was negative for drugs of abuse, and a serum ethanol level was undetectable.

The patient remained stable while in the ED, with no change in her symptoms. Her mental status remained normal, and follow-up temperature recordings were afebrile. Neurology consultation was obtained and the patient was discharged home with no specific therapy to obtain an outpatient MRI scan and neurological clinic follow-up.

What was the final diagnosis in this patient?

A Nine Year-old Who Is Walking “Like He Is Drunk”

Edward P. Sloan, MD, MPH

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Case Presentation

A 9 year-old young man was brought to the Emergency Department because he was “walking like he was drunk”, with speech difficulties and illegible handwriting. The child had had these symptoms for over two days, and his pediatrician had obtained a plain brain CT, which was interpreted as negative. He denied any headache or visual changes.

The child had fallen while riding his bike 12 days earlier, landing on his face, without LOC, as witnessed by his siblings. His mother saw him 30 minutes later, and noted him to be “ a little dazed”, but otherwise OK. He had amnesia to the event, and complained of a diffuse headache for three days following the accident. His history was otherwise not contributory.

On physical exam, he had some resolving abrasions and ecchymosis of the right malar eminence. Although he comprehended speech adequately, his own speech was sparse. He had diffuse weakness (4/5) of all extremities, and slightly hyperreflexia on his R side, including a positive Babinski’s reflex on the R. He was noted to have cerebellar ataxia as noted on finger-to-nose and heel-to-shin movements of the R extremities. He had a moderately ataxic gait in the ED, especially with heel-to-toe walking.

What is your differential diagnosis and how would you proceed?