IV Immunoglobulin in the Treatment of Neuromuscular Disorders

Case Presentation: Initial Emergency Room Consultation

A 35-year-old male with no significant past medical history presents to the emergency room (ER) with rapidly progressive weakness and sensory changes.

The patient developed a respiratory infection 3 weeks prior, from which he made a full recovery. His primary care physician suspected a viral upper-respiratory tract infection, and the patient was not treated with antibiotics. About 4 days before presentation at the ER, the patient noticed tingling in his feet that started rapidly and soon progressed to involve his hands. Over the next couple days, he noticed his legs felt heavy, and he was tripping when he walked. Later, he had difficulty lifting his arms and noticed he was dropping things. The day before presenting to the hospital, the patient had difficulty catching his breath and therefore decided to seek medical attention. He denies any neck pain, diplopia, swallowing problems, or bladder/bowel incontinence. He denies any recent immunizations.

His past medical history is unremarkable. He takes no prescription or over-the-counter medications or supplements. He has no known drug allergies. He does not smoke, drink alcohol, or use illicit substances. He is an attorney. His sister has rheumatoid arthritis but there is no family history of peripheral neuropathy or other neurologic diseases.

In addition to what is noted above, a complete 14-topic review of systems is obtained and is unremarkable. On physical examination, he is a well-developed and well-nourished male in no acute distress. He is afebrile. His blood pressure is 110/70, pulse is 90, and respiratory rate is 16. His oxygen saturation on room air is 99%. There are no murmurs or abnormal heart sounds. His lungs are clear. There are no skin lesions.

He is alert and oriented to person, place, and date. Registration and 5-minute recall are normal. He follows commands and names and repeats without difficulty. His speech is fluent.

Cranial nerve testing reveals PERRLA; optic discs are sharp, visual fields are full to confrontation, and extraocular muscles are intact. Facial sensation is intact. Facial strength is mildly impaired, as evidenced by a horizontal smile and mild problems with eye closure. Hearing is intact bilaterally to finger rub. Palate, tongue, and uvula are midline. Shoulder shrug strength is normal.
Motor strength is MRC grade 3/5 in the legs. In the arms, he has MRC grade 4/5 strength throughout. There is no evidence of spasticity in the arms and legs. There is no significant muscle atrophy.

There is decreased pinprick and temperature perception distal to the knees and elbows bilaterally. The patient has absent vibratory perception in the toes. Proprioception is mildly reduced in the toes.

Reflexes are absent throughout. Plantar responses are flexor bilaterally. Coordination is normal on finger-nose-finger and heel-knee-shin testing bilaterally. His gait is very unsteady, and he has difficulty arising from the bed. Telemetry in the ER shows normal sinus rhythm.

You discuss with the patient that his presentation is suggestive of Guillain-Barre syndrome (GBS), which is a rapid-onset inflammatory peripheral neuropathy or peripheral nerve dysfunction. However, you want to run some tests, including labwork, lumbar puncture, and electrodiagnostic studies to confirm your suspicions. In the meantime, he will be admitted to the hospital for close monitoring in case of any further deterioration. You ask that the patient have a baseline and serial forced vital capacities (FVC) to assess his respiratory status. Rehabilitation services, including physical, occupational and speech therapy, are consulted.

**Evaluation and Management Coding**
If the patient is a neurologic consultation for someone admitted to a medicine service, the correct code would be 99255 for a level 5 initial inpatient consultation. If the patient is admitted to the service of the neurologist, the code would be 99233 for an initial hospital care billing. In both cases, the history and physical are comprehensive, and the medical decision making is high complexity.

**Hospital Follow-up Visit**
You evaluate the patient in follow-up the next day. Overnight, the patient was admitted to the Intensive Care Unit because his FVC was borderline low. Unfortunately, his weakness progressed. Furthermore, his FVC continued to decrease, and he developed worsening dyspnea, prompting the use of noninvasive positive pressure ventilation.

On examination, you find that his strength has significantly deteriorated. Motor strength is MRC grade 0–1/5 in the legs. In the arms, he has MRC grade 3/5 strength throughout. Otherwise, his cranial nerves examination, sensory examination, and reflexes are unchanged. He is nonambulatory.

His lab work is negative for HIV and Lyme disease. His lumbar puncture shows elevated protein level with normal white cell count. His nerve conduction studies and electromyography are consistent with an early generalized demyelinating polyneuropathy.
You discuss with the patient that his presentation, examination, and diagnostic studies are consistent with GBS. You would like to start him on treatment and understand that both IV immunoglobulin (IVIg) and plasma exchange are both efficacious. At this hospital, IVIg is more routinely performed and is therefore chosen. You counsel the patient that the infusion will be dosed at 0.4 mg/kg/day for 5 days. You advise him that potential side effects include hypercoagulability and renal failure. The patient asks why he cannot have both IVIg and plasma exchange as a measure of hastening his recovery. You counsel him that this combination is probably not better than either treatment alone. You further explain that while he is receiving the infusion, he will continue to have supportive care, including FVC monitoring and rehabilitation therapies.

**Evaluation and Management Coding**

One could choose one of two codes for this visit. Many would choose the subsequent hospital care code of 99233, as your history and physical are detailed, and the medical decision making is high complexity. If one spent 35 minutes or more with the patient and 50% or more of the time was expended counseling the patient and coordinating care, then one could use time as the determinant for coding 99233. The neurologist could also use code 99291 for critical care services as long as 30 minutes were expended in the visit.

**Outpatient Neurology Clinic Follow-up – 1 Month**

The patient returns to your outpatient clinic 1 month after your initial consultation (about 5 weeks after symptom onset). After he received IVIg, his neurologic course had stabilized. He was then transferred to an Acute Rehabilitation Center where he has worked daily with physical and occupational therapy. After therapy, he has improved and is now ambulating with a walker. He denies any worsening neurologic symptoms or dyspnea.

On examination, you find that his strength has significantly improved. His facial weakness has improved. Motor strength is MRC grade 4/5 in the proximal legs. In the distal legs, he has 3/5 ankle dorsiflexion strength. In the arms, he has MRC grade 4/5 strength throughout. Otherwise, his cranial nerves examination, sensory examination, and reflexes are unchanged. His gait is steady with a walker.

The patient is counseled that he has followed the expected course for GBS treatment. He asks if there is any benefit to repeating IVIg, and you advise him that this would likely not help and that you expect that he will continue to improve as time passes. He is encouraged to call if there is any deterioration, as deterioration might suggest a more chronic inflammatory neuropathy.

**Evaluation and Management Coding**

Most neurologists would bill an established outpatient code of 99213 (level 3 established patient). The patient is markedly improved, and medical decision making is straightforward. If one spent 25 minutes with the patient, and more than 50% was spent in counseling the patient about the use of IVIg, plasma exchange, other therapies, prognosis, and the differentiation from CIDP, then one could choose 99214 (level 4...
established patient) as long as the note concluded with a statement regarding counseling and coordination of care and the topics discussed.

**Outpatient Neurology Clinic Follow-Up – 6 Months**

The patient returns to your outpatient clinic 6 months after your initial consultation. He is dramatically improved and walks in without any gait assist devices. Since your last visit, he continued to improve at the Acute Rehabilitation Center and then was transitioned home with outpatient rehabilitation services. He denies any worsening of neurologic symptoms. He has no specific complaints.

On examination, you find that his strength has significantly improved. Motor strength is MRC grade 5/5 in the legs with the exception to toe dorsiflexion and plantar flexion which are 4/5. In the arms, he has MRC grade 5/5 strength throughout. His sensory examination shows improved but decreased vibratory perception in the toes. Distal pin prick and temperature perception are still reduced as before. His reflexes are still absent. His gait is steady.

The patient is quite happy with his overall improvement. He is encouraged to call if there is any deterioration, which might indicate a more chronic inflammatory neuropathy. Otherwise, he has elected to follow up as needed.

**Evaluation and Management Coding**

Most neurologists would bill for a level 3 outpatient established patient code of 99213. Medical decision in this case would be low complexity, driving the decision to use this code.

**Questions**

1. Which of the following is true for IVIg in the treatment of GBS in adults according to the recent guideline “Intravenous immunoglobulin in the treatment of neuromuscular disorders”\(^1\)?
   - A. IVIg is less effective than plasmapheresis
   - B. IVIg is ineffective for the treatment of GBS
   - C. IVIg should be followed by treatment with plasmapheresis
   - D. IVIg is equally efficacious as plasmapheresis
   - E. IVIg is optimally given at a total dose of 1gm/kg
   **The correct answer is D.**

2. What do the guidelines indicate for the role of steroids in the treatment of GBS?
   - A. Steroids have proven benefit as adjunctive therapy with IVIg
   - B. There is no proven benefit for the use of steroids with IVIg
   - C. Steroids are effective therapy when used alone
   - D. High-dose steroids are more effective than low-dose steroids
   - E. Steroids are more effective with plasmapheresis than with IVIg
   **The correct answer is B.**
3. If this patient were to develop a more chronic, progressive course of neuropathy consistent with a diagnosis of CIDP, what do the guidelines recommend about treatment with IVIg for this condition?
   A. IVIg is effective for the long-term treatment of CIDP
   B. Treatment of CIDP is not covered in the guideline
   C. There is inconclusive information on the treatment of CIDP with IVIg
   D. Plasmapheresis is more effective than IVIg
   E. IVIg is not effective for the treatment of CIDP

   The correct answer is A.

4. Which of the following conditions has a Level A recommendation for treatment with IVIg?
   A. Post-polio syndrome
   B. Inclusion body myositis
   C. IgM paraproteinemia
   D. GBS
   E. Diabetic neuropathy

   The correct answer is D.

Diagnosis Coding

This is a case presentation and not a patient record. The diagnosis code choice for each visit would depend on the diagnostic statement in the record.

In the initial emergency room consultation, the case description states the symptoms of "rapidly progressive weakness and sensory changes," and the patient is told the "presentation is suggestive of Guillain-Barré syndrome (GBS)." We do not actually see the official recorded diagnosis here. Though the diagnostic statement, "suggestive of Guillain-Barré syndrome" or "rule out Guillain-Barré syndrome" is commonly seen in records, ICD-9-CM Official Guidelines for Coding and Reporting specifically advises against using such uncertain terms as the diagnosis for billing. There is no means for capturing the concept of "rule out," "probable," "possible," or similar terms with ICD-9-CM. For physician claims (Part B Medicare or other third party), instead use the diagnosis to the highest level of specificity available. For some physicians, the data present on admission here would be enough to use the diagnosis of GBS for that day’s billing claim. For those not comfortable with using GBS, then use as the diagnosis the presenting symptoms.

If the initial diagnosis is uncertain, and symptoms are the highest level of specificity, code:

   728.87   Muscle weakness
   782.0    Disturbance of skin sensation

If the initial diagnosis is GBS, code:

   357.0   Acute infective polyneuritis
Guillain-Barre syndrome

For the follow-up hospital visit, use the GBS code as above.

For the two outpatient visits, the acute phase and treatment of GBS is complete, and the symptoms are sequelae of the acute illness. There are no “late effects” codes in ICD-9-CM for the sequelae of GBS. This appropriate ICD-9-CM code for these visits is once again 357.0.

Though the implementation date for ICD-10-CM is in reconsideration, this classification is still expected to replace ICD-9-CM at some point in the future. The rules for using these codes are very similar to those for ICD-9-CM. For the symptoms listed above, the ICD-10-CM codes are:

- **M62.81** Muscle weakness
- **R20.8** Other disturbance of skin sensation

And for GBS:

- **G61.0** Guillain-Barre syndrome

In ICD-10-CM there is a code for the late effects of GBS, and the code instructs to “code first” the sequelae. For the outpatient visits code:

- **M62.81** Muscle weakness
- **G64.0** Sequelae of Guillain Barre syndrome

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