

**Case Report**

# Case Series of Severe Abdominal Sepsis, AKI with Anuria and Fulminant Guillain-Barre Syndrome (GBS) Mimicking Brain Stem Death

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**Abstract**

The presence of coma, flaccid quadriplegia without sedation and absence of most of the brain stem reflexes (fixed dilated pupil, no response to light, no corneal reflex, absent cough and gag reflexes) suggests brain stem death. However, polyneuropathy in nerve conduction studies (NCS), a high protein concentration in the CSF, and EEG suggested that the patient actually had GBS and all three patients recovered from GBS after receiving therapeutic plasma exchange (TPE) with / without IV immunoglobulin.

Brain death determination includes exclusion of reversible cerebral injury, confounding conditions, and it is vital to perform electroencephalography (EEG) when uncertainty exists about the reliability of the clinical exam to enable appropriate early diagnosis and therapeutic management.

We report these cases as an important reminder to clinicians to consider immune mediated post infectious polyneuropathy like GBS before accepting a diagnosis of brain stem death, especially in patients with sepsis. Variant GBS can be associated with Bickerstaff brainstem encephalitis and this explains coma, loss of brain stem reflexes.

**Introduction**

GBS is one of the most common causes of acute, acquired weakness and is often provoked by a preceding infection. In some cases, it may be complicated by respiratory failure or autonomic dysfunction. The acute polyneuropathy of GBS is often triggered by an immune response to an antecedent infection or other event where cross-reacting epitopes on peripheral nerve are similar to

those of the pathogen (molecular mimicry), and all myelinated nerves (motor, sensory, cranial, sympathetic) can be affected.

Bickerstaff brainstem encephalitis (BBE) is syndrome characterized by encephalopathy with ophthalmoplegia and ataxia [1], and is associated with the presence of antibodies to particular gangliosides that are present in neuronal tissue with homologues in some pathogens (anti-GQ1b antibodies).

The exact cause of Guillain-Barre syndrome is unknown; however, two-thirds of patients report symptoms of an infection in the six weeks preceding diagnosis and such infections include COVID-19, respiratory or a gastrointestinal infection, and Zika virus.

There is no known cure for Guillain-Barre syndrome, but several treatments appear to improve symptoms and reduce the duration of the illness. Although most people recover completely occasionally severe cases can be fatal. Whilst recovery may take several years, most people are able to walk six months after symptoms. Some people may have lasting effects from it, such as weakness, numbness or fatigue. Variant GBS can be associated with Bickerstaff brainstem encephalitis which may mimic brain death [1].

Diagnosis of brain death is established by criteria first established at a conference of the Royal Colleges and Faculties of the United Kingdom in 1976, namely that all brain stem reflexes are absent (the pupils are fixed in diameter with no response to light, no corneal reflex, oculovestibular reflexes are absent, no response to painful stimulation within the cranial nerve distribution, cough and gag reflexes are absent, and there is apnea). Electroencephalography (EEG) and cerebral angiography were not initially included although they may assist in difficult cases [2,3].

Sepsis and septic shock are the causes of AKI and are known as Sepsis-Associated AKI (SA-AKI) and accounted for more than 50% of cases of AKI in the ICU, and have a poor prognosis [4].

In our rare case series three cases of abdominal sepsis with AKI and GBS mimicking brain stem death are presented.

All patients presented to Madinat Zayed Hospital (MZH), SEHA-Abu Dhabi-U.A.E., this is a secondary hospital with some tertiary subspecialties and 20 ICU beds between 2009 to 2023.

### **Case 1:**

A 38-year-old male patient from Bangladesh was admitted in 2009 with an acute abdomen. Surgical exploration revealed a sealed peptic ulcer. The patient developed severe AKI (creat 930  $\mu$ mol/l, urea 20 mmol/l bic 8 mmol/l) with anuria from admission and required renal replacement therapy in the form of intermittent hemodialysis. On the day three of admission, he developed unexplained respiratory failure and required intubation and mechanical ventilation. At day 7, he developed flaccid quadriplegia without sedation, and fixed dilated pupils, together with autonomic neuropathy in the form of low blood pressure and bradycardia. On day 15 he remained unwell with flaccid quadriplegia without sedation for 10 days, and no brainstem reflexes. An apnea test was performed and he was declared brain dead.

On day 33, he passed a large amount of urine and the dialysis was ceased. He started to move his lower jaw, and his pupils

began reacting to light. The diagnosis was revised and the possibility of GBS (unexplained respiratory failure, autonomic neuropathy, a high CSF protein of 4.4 G/l) was entertained and he received IV immunoglobulin. Nerve conduction studies (NCS) confirmed diffuse symmetrical mixed sensory and motor axonal polyneuropathy.

On day 48 he started to respond to verbal stimuli, moving head right and left, but still had flaccid quadriplegia. By day 56 he was fully awake and communicating, but still had a flaccid quadriplegia, and he required continued mechanical ventilation because weak respiratory muscle until day 70. On day 78 he was transferred from ICU to the rehabilitation center with good outcome but with some residual neurological disability and moderate weakness mainly lower limbs.

### **Case 2:**

A 28-year-old male African patient without any previous history of severe or chronic illness was admitted to the ICU in 2020 with an acute abdomen, ascites, high inflammatory markers (CRP 290 mg/l, WBC 34,000 cells/mm<sup>3</sup>), high liver enzymes (ALT 309 IU/l, AST 306 IU/l), severe metabolic acidosis (pH 7.2, bic 14 mmol/l) and anuria with serum creatinine 872  $\mu$ mol/l and urea 19 mmol/l. A CT abdomen with contrast revealed normal sized kidneys, but with irregular medullary enhancement and hypodense non-enhancing renal cortex (reverse rim sign) [5], Figure 1, consistent with renal cortical necrosis, requiring renal replacement therapy, initially with daily intermittent hemodialysis (IHD) from day 1 until day 5. However due to increasing hemodynamic instability he started continuous renal replacement therapy (CRRT) with regional citrate anticoagulation. On day 5 he required paracentesis of his tense ascites. On day 6 he developed unexplained type 2 respiratory failure, and was intubated and ventilated. On day 10 he was noted to have flaccid quadriplegia with no gag reflex, sluggish pupillary reactions, areflexia, ophthalmoplegia, and was not responding to painful stimuli and with absent doll's eye, corneal and gag reflexes. Nevertheless, he had a high CSF protein (of 1.7 g/l) and normal EEG. His NCS showed features consistent with acute progressive demyelinating polyneuropathy. Anti-GQ1b antibodies were requested but were unavailable in our pathology department as we suspected a GBS, Miller-Fisher, Bickerstaff brainstem encephalitis overlap syndrome and he started therapeutic plasma exchange (TPE). After five sessions of TPE followed by intravenous immunoglobulin (IVIG), he started to obey commands with improvement of the power in all limbs.

Ascitic fluid analysis revealed high white cell count with 97% lymphocytes and a high protein (of 22 g/l) and tuberculous peritonitis was suspected, and so he started antituberculosis treatment on the day 18. Given the cortical necrosis we suspected irreversible renal disease and he started intermittent hemodialysis

thrice weekly from day 37. After 64 days, he was transferred from the ICU to the rehabilitation center and required maintenance hemodialysis. On day 211, he had partial recovery of AKI with sufficient renal function to become dialysis independent, with serum creatinine about 140  $\mu\text{mol/l}$  without renal replacement therapy (RRT).

**Case 3:**

In 2023, a 44-year male patient from Bangladesh with history of hypertension, dyslipidemia was admitted with an acute abdomen, a serum creatinine of 444  $\mu\text{mol/l}$  anuria, high inflammatory markers (CRP 152 mg/l) and liver enzymes (AST 150 IU/l ALT 142 IU/l) with a known previous baseline normal serum creatinine (81  $\mu\text{mol/l}$ ). A diagnosis of abdominal sepsis with secondary AKI was made and he required admission to ICU and started daily intermittent HD then CRRT. On day 9 the patient had an unexplained cardiac arrest, likely due to respiratory failure. He was successfully resuscitated but developed flaccid quadriplegia without sedation,

with fixed dilated pupils, loss of deep reflexes, ophthalmoplegia, no response to light, no corneal reflex, absent cough and gag reflexes and brain death was considered. On day 18, the patient started to pass more urine suggesting some renal recovery. On day 22 NCS revealed severe axonal sensory and motor polyneuropathy affecting mainly lower limbs (Table 1) and he had a normal EEG. The CSF protein was high (1 g/l), again suggesting the diagnosis of GBS with overlap syndrome. He commenced TPE on day 24 and received 10 sessions with volume of 3 liters and replacement with albumin 5%, on alternate days, followed by IVIG 26 g/d for 5 days. We noted gradual improvement in his condition and he started to open his eyes, obey commands and power improved in all limbs (upper > lower). He was weaned from ventilator support on day 93 and transferred from ICU to a medical ward on day 96. He was discharged from the hospital after 171 days walking with assistance with mild weakness in the lower limbs and serum creatinine of 120  $\mu\text{mol/l}$  without renal replacement therapy.

**Motor Nerve Conduction Study:**

Site	Latency (ms)	Amplitude (mv)	Area (mv/ms)	Segment	Distance (mm)	NCS (m/s)
Median L						
Wrist	4.8	4.3	18	Wrist		29.7
Elbow	12.5	0.3	2	Wrist-elbow	230	(RR 49–68)
Median R						
Wrist	4.5	3.3	12.5	Wrist		44
Elbow	9.5	2.9	12.3	Wrist-elbow	220	(RR 49–68)
Ulnar L						
Wrist	3.6	4.0	14.5	Wrist		40
Elbow	9.3	3.6	14.5	Wrist-elbow	230	(RR 51–70)
Ulnar R						
Wrist	3.3	4.7	15.5	Wrist		45.3
Elbow	8.6	4.3	16.1	Wrist-elbow	240	(RR 51–70)
Peroneal L						
Ankle			No response			
Head of fibula			(RR 36–69)			

Peroneal R						
Ankle			No response			
Head of fibula			(RR 36–69)			
Tibial L						
Ankle			No response			
Popliteal			(RR 35–68)			
Tibial L						
Ankle			No response			
Popliteal			(RR 35–68)			

### Sensory Nerve Conduction Study

Site	Latency (ms)	Amplitude (uv)	Area (uv /ms)	Segment	Distance (mm)	NCS (m/s)
Median L Wrist	3.0	8.3	0.7	Wrist	120	39.5 (RR 50–74)
Median R Wrist	3.2	5.6	0.3	Wrist	130	40.1 (RR 50–74)
Ulnar L Wrist	2.7	2.3	0.2	Wrist	110	41 (RR 50–74)
Ulnar L Wrist	2.7	2.3	0.2	Wrist	110	41 (RR 50–74)
Ulnar R Wrist	2.3	5.3	0.4	Wrist	100	43.1 (RR 50–74)
Sural Left			No response (RR 40–70)			
Sural Right			No response (RR 40–70)			

**Table 1:** NCS for case three-revealed severe axonal sensory and motor polyneuropathy affecting mainly lower limbs (RR = Reference range).



**Figure 1:** CT abdomen with contrast hypodense non-enhancing renal cortex, characteristics for (reverse rim sign) consistent with renal cortical necrosis.

## Discussion

We present 3 patients with abdominal severe sepsis, AKI requiring renal replacement therapy who developed Guillain-Barre syndrome (GBS) mimicking brain stem death.

AKI complicating abdominal sepsis is not an uncommon finding for patients in the ICU, and sepsis is found in about 40% to 50% of patients with AKI in the ICU and amongst the critically ill, sepsis is the most common cause of AKI [4].

Cortical necrosis (CN) is a rare cause of acute kidney injury (AKI) in developed countries accounting for ~2% of all patients with AKI [6]. CN may commonly be caused by severe hemodynamic perturbation including obstetric causes like abruptio placentae, but also sepsis, burns, snake bites, pancreatitis, HUS, toxins and drugs [6]. AKI with acute anuria (urine output < 100 mL/24 h) is uncommon, and more commonly related to obstruction, shock or a catastrophic vascular event but in these cases the anuria was secondary to abdominal sepsis. Other studies have also reported that sepsis was the main cause of anuria with incidence of about 60% in hospital acquired AKI [7-10].

We report the three cases of fulminant Guillain–Barré syndrome (GBS), mimicking brain death, who were previously healthy, admitted to hospital ICU with abdominal sepsis. All patients quickly developed respiratory failure, requiring mechanical ventilation. When pharmacological sedation was withdrawn, patients were found to have a coma with absent brainstem reflexes, resembling a state of brain death.

A GBS was diagnosed, based on the NCS demonstrated motor and sensory responses unexcitable in upper and lower extremities. An EEG showed normal brain function, high CSF protein. Treatment with therapeutic plasma exchange followed by IV intravenous immunoglobulins appeared to be effective. While the flaccid quadriplegia often persisted for some time, ocular movements

appeared to come back initially, along with signs of recovering consciousness. Then, after some weeks, small amplitude and long duration motor response were detectable in the upper limbs. Despite the very long ICU stays and prolonged recovery the eventual outcomes appeared favourable, after rehabilitation. Severe GBS may thus mimic brain death and it is important to stress that other testing is important when considering the possibility of Bickerstaff brainstem encephalitis (BBE). BBE is characterized by progressive ophthalmoplegia, ataxia and disturbance of consciousness, has an overlap with Miller Fisher syndrome and GBS [1-3].

## Conclusion

Sepsis is the most common cause of hospital AKI rarely can associate with anuria or renal cortical necrosis (CN), with poor outcome but delayed recovery with late resumption of sufficient renal function to become dialysis independent still possible [6]. The exact cause of Guillain-Barre syndrome is unknown, but two-thirds of patients report symptoms of an infection in the six weeks preceding. GBS can overlap with anti-GQ1b antibody syndromes (Miller Fisher, Bickerstaff brainstem encephalitis) which may mimic brain stem death. We strongly recommend an EEG and NCS in such cases, along with assay of anti-GQ1b antibodies where available.

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**Patient consent:** The patients provided written informed consent to publish their case and any associated images.

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## Author statements

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**Data availability:** All data generated during cases are included in the article further inquiries to be address to the correspondence author.

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