

ICTAL ASYSTOLE IN A PATIENT WITH POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES) AND SEIZURES: A CASE REPORT

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Background

- Ictal asystole is rare, seen in less than 1% of patients with epilepsy, and typically seen in patients with temporal or frontal lobe seizures, rarely in occipital lobe seizures, and has never been described in a patient with PRES.
- PRES is a clinical-radiologic diagnosis in which there is a breakdown of the blood brain barrier caused by abrupt changes of blood pressure, or cytokines causing endothelial damage and this creates brain edema.
- The typical neurological symptoms of PRES include encephalopathy (50-80%), seizure (60-75%), headache (50%), and visual disturbance (35%).
- Brain imaging in PRES usually reveals vasogenic edema in the bilateral parietal-occipital regions, and may include the frontal, temporal, basal ganglia, brainstem or cerebellum.
- Most patients with PRES have a favorable outcome but mortality can be as high as 3-6%, persistent neurological sequela in 10-20% of patients, and recurrent PRES in 5-10%.

Case Report

History of presenting illness: A 75 year old right-handed woman initially presented after having a generalized tonic-clonic seizure at home. Her imaging showed bilateral parieto-occipital edema consistent with PRES. She returned to the hospital a week later with encephalopathy and her MRI brain showed a new infarct in the right occipital pole.

Past medical history: chronic kidney disease, diabetes mellitus, neuropathy, morbid obesity, hypertension, breast cancer and depression

Home medications: carvedilol, furosemide, gabapentin, glipizide, losartan, oxycodone, sertraline, simvastatin and sitagliptin.

Neurological Exam: She was awake and oriented to person and place. She had difficulty following commands and was inattentive. Her cranial nerve examination was normal. Her motor strength examination revealed normal strength in all extremities except for bilateral plantar weakness 4+ (chronic). Her reflexes were 2+ in the upper extremities and absent in the lower extremities. She had upgoing toes on the left and downgoing on the right. She had a length dependent neuropathy in the lower extremities.

Laboratory Values: white blood cell count 15.2, creatinine 1.5, glucose 198 and urine culture positive for E. coli.

Imaging:

MRI brain: Confluent signal abnormality in the cerebral white matter predominantly posteriorly, compatible with evolving PRES with superimposed new areas of acute infarction, largest in the right occipital pole.

Hospital Course: Eight days into her admission, she had two episodes of bradycardia, AV block, and asystole for up to 9 seconds within 5 hours of each other. Her heart rate was 30 bpm and her blood pressure was elevated to 208/95 with both episodes. Staff found her to be unresponsive and bleeding from the mouth. These episodes were not immediately recognized as seizures and were initially attributed to PRES and transient cerebral hypoperfusion related to bradycardia/asystole. However, due to the concern for seizures, she was later placed on continuous video-EEG monitoring. Four hours later, she had a secondarily generalized tonic-clonic seizure with AV block and sinus arrest resulting in 20 seconds of asystole, followed by severe bradycardia for 69 seconds. The staff initiated cardiopulmonary resuscitation five seconds after the asystole ended with return of sinus rhythm. Her video EEG revealed a seizure arising from the right parietal lobe with secondarily generalization. She had no further seizures after treatment. She had complications due to her acute respiratory failure and was placed on comfort care. She unfortunately died a week later.

Diagnostic Work-up

Imaging

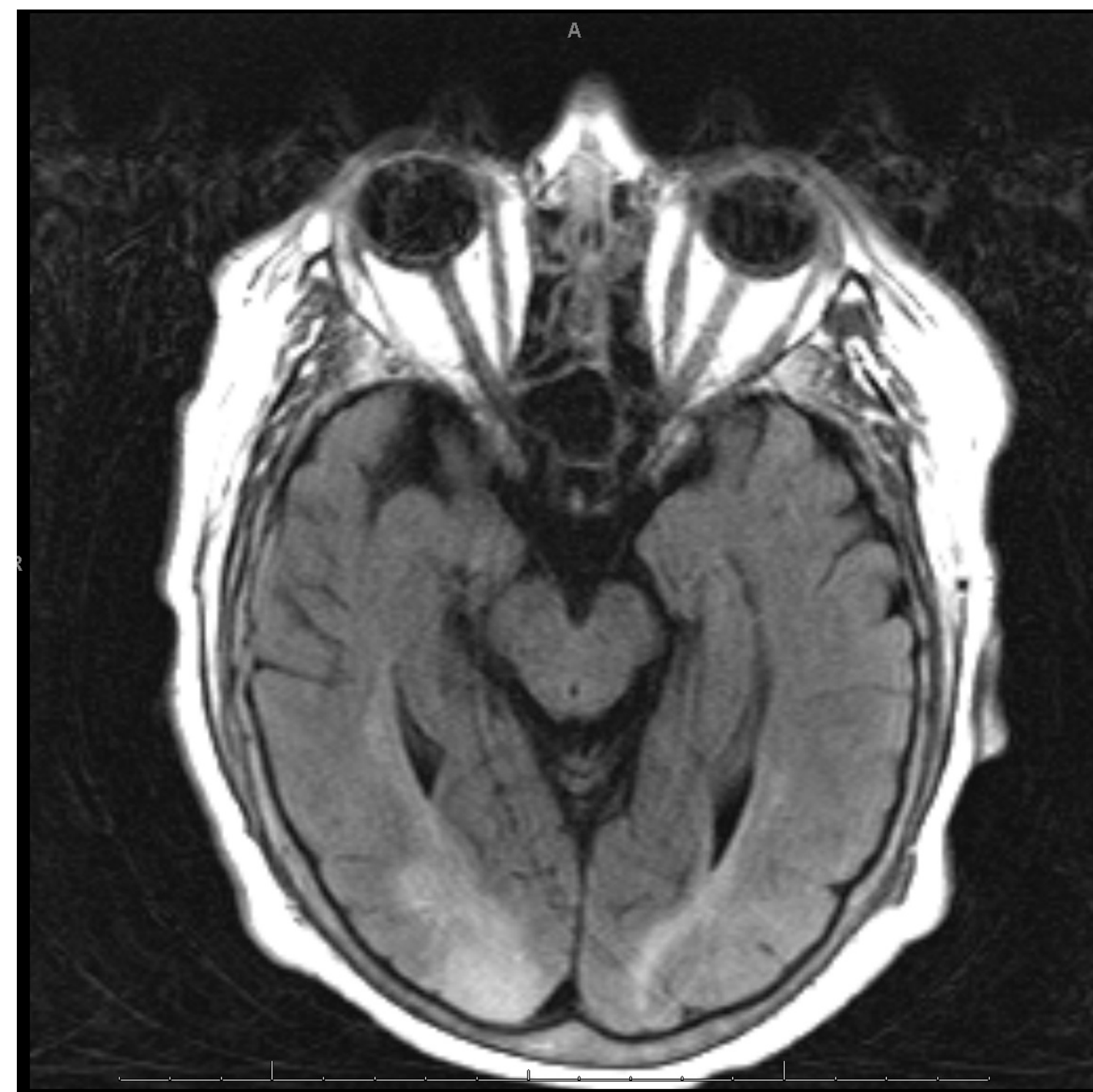


Figure 1: Brain MRI scan, axial T2 fluid-attenuated inversion recovery (FLAIR) sequences showing abnormal T2 signal bilaterally in the right greater than left occipital lobe, consistent with vasogenic edema.

Video Electroencephalogram (EEG)

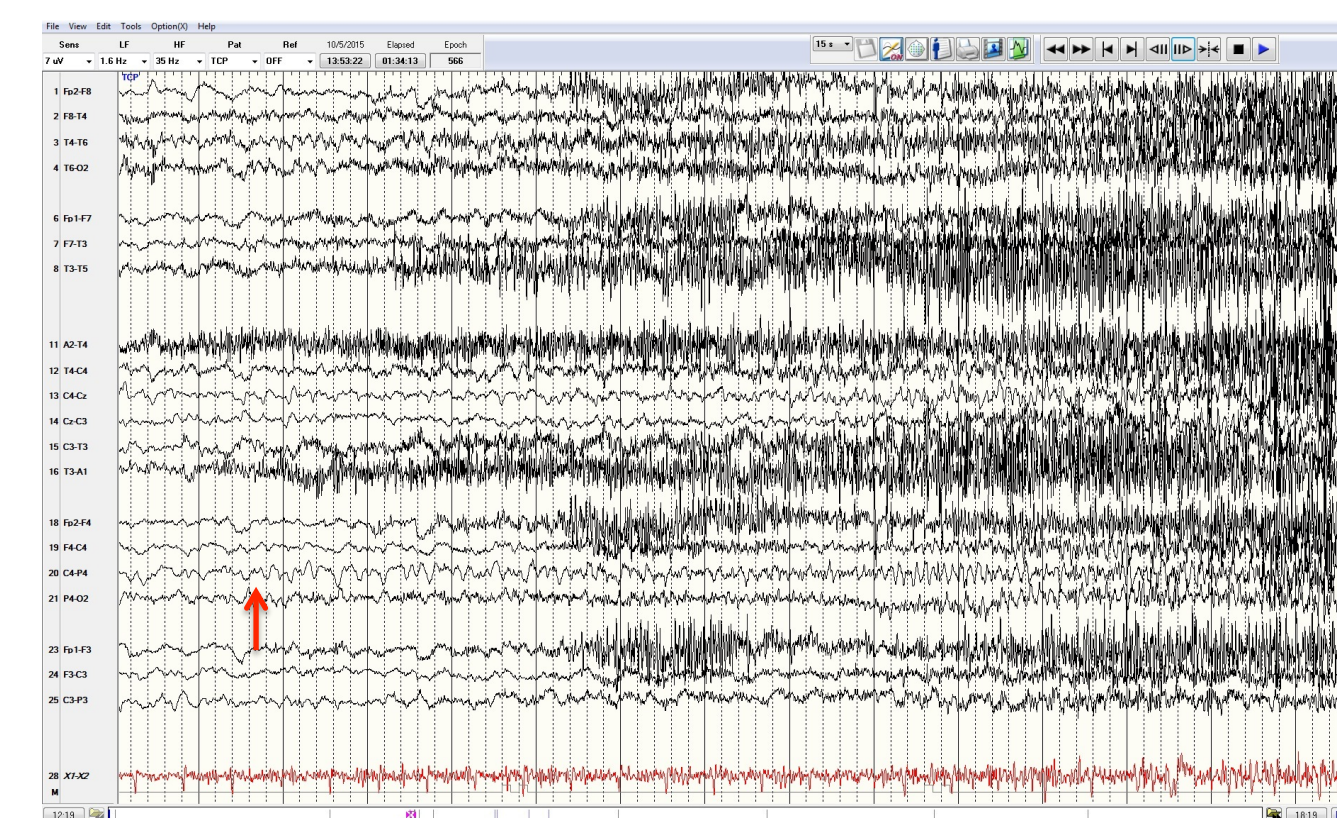


Figure 2: The seizure starts maximally in the right parietal region (P4) with rhythmic theta frequencies (red arrow) with some ictal spread to the right posterior temporal region (T6).

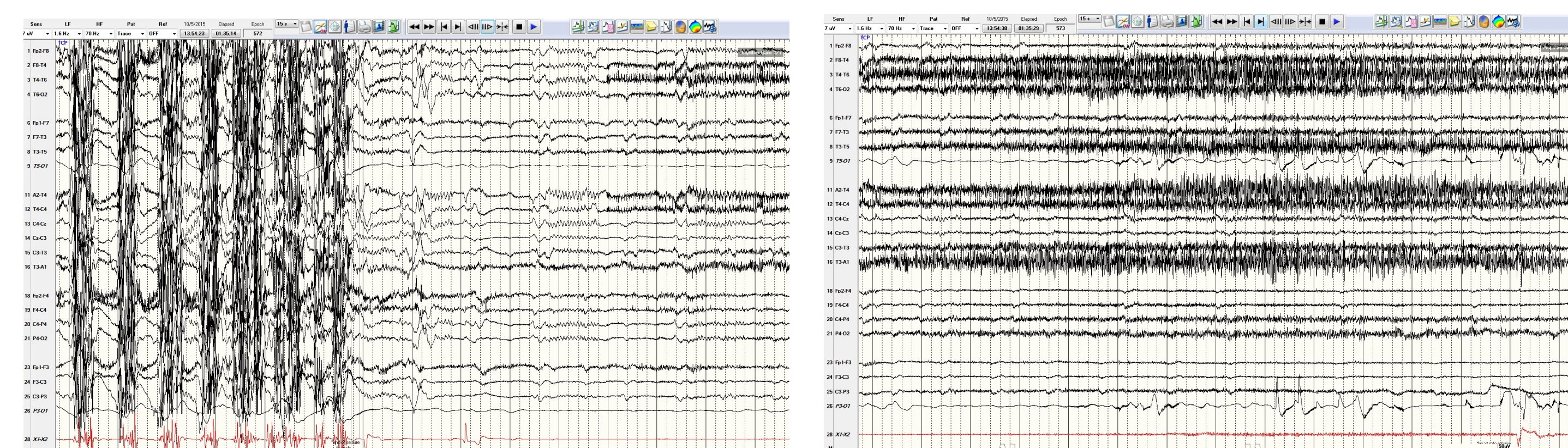


Figure 3: Asystole. The first page records the end of a secondarily generalized tonic clonic seizure. At the end of the seizure, the ECG shows asystole which continues on the next EEG page. The cardiac asystole continues for 20 seconds.

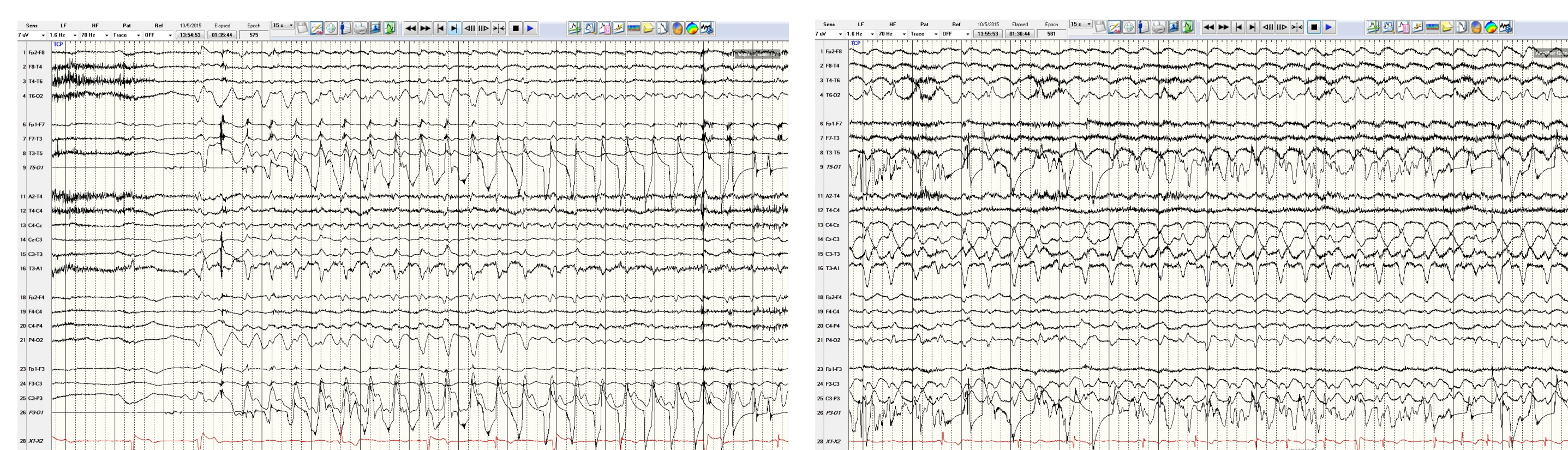


Figure 4: Bradycardia. Consecutive EEG tracings from above. The first page shows bradycardia in the ECG lead 20 seconds after the end of the seizure. This continued for 69 seconds until the heart rate returned to sinus rhythm as seen in the second page of EEG. The EEG shows chest compression artifact in both pages.

Conclusions

- Our patient had recurrent ictal asystole, likely due to PRES.
- Ictal asystole has possible serious medical consequences, including syncope, and may be a potential mechanism for SUDEP.
- Parasympathetic functions responsible for bradycardia/asystole are thought to be lateralized primarily to the left hemisphere. Contralateral seizure spread to the left insular cortex or temporal lobe may have caused the bradycardia and asystole in our patient.
- PRES is not frequently associated with epilepsy and the risk for epilepsy has been reported to be less than 2 percent.
- In contrast, seizures are very common in PRES. There are no data to guide how long therapy should be in patients with seizures and PRES but short term therapy during the acute stage of PRES is recommended.
- Prompt treatment of seizures and recognition that PRES can cause recurrent ictal asystole may help to prevent asystole-associated complications, possibly including death.

References

- Britton JW, Ghearing GR, Benarroch EE, Cascino GD. The ictal bradycardia syndrome: localization and lateralization. *Epilepsia* 2006;47:737-744.
- Datar S, Singh T, Rabinstein AA, Fugate JE, Hocker S. Long term risk of seizures and epilepsy in patients with posterior reversible encephalopathy syndrome. *Epilepsia* 2015; 56(4):564-568.
- Fugate JE, Rabinstein AA. Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions. *Lancet Neurol.* 2015;14(9):914-25.
- Moon SN, Jeon SJ, Choi SS, et al. Can clinical and MRI findings predict the prognosis of variant and classical type of posterior reversible encephalopathy syndrome? *Acta Radiol* 2013;54:1182-90.
- Nei M, Sperling MR, Mintzer S, Ho RT. Long-term cardiac rhythm and repolarization abnormalities in refractory focal and generalized epilepsy. *Epilepsia.* 2012;53(8):e137-40.
- Oppenheimer SM, Gelb A, Girvin JP, Hachinski VC. Cardiovascular effects of human insular cortex stimulation. *Neurology* 1992;42:1727-32.
- Schuele S, Bermeo AC, Alexopoulos AV, Locatelli ER, Burgess RC, Dinner DS, Foldvary-Schaefer N. Video-electrographic and clinical features in patients with ictal asystole. *Neurology.* 2007;69:434-441.
- Tinuper P, Bisulli F, Cerullo A, Carcangiu R, Marini C, Pierangeli G, Cortelli P. Ictal bradycardia in partial epileptic seizures: autonomic investigation in three cases and literature review. *Brain.* 2001;124:2361-2371.