Introduction: Congenital Long QT syndrome (LQTS) is a rare congenital disorder of ventricular repolarization characterized by QT interval prolongation. The prevalence in Asia ranges from 0.02% to 0.04%. LQTS predisposes patients to seizure like activity, syncope, life threatening arrhythmias and sudden cardiac death. Diagnosis is based on a good clinical history and electrocardiographic findings. The epinephrine stress test provides a safe and reliable way of unmasking Type 1 LQTS.

Methods: Objective: To present a case that will highlight the clinical utility of provocative testing with Epinephrine in a young female with history of recurrent seizure and syncope.

Result: Case Presentation: A 27-year old female who was having episodes of seizure and syncope for the past two months. Neurologic work-up and initial cardiac work up were all negative and was managed with seizure disorder. She came in with another episode of seizure followed by syncope sustaining a hematoma on her right frontal area. She was documented having supraventricular tachycardia on monitor which reverted to sinus rhythm after giving doses of Adenosine and Verapamil. Given the history of seizure and syncope with associated arrhythmia in a young patient, long QT syndrome was highly suspected. An epinephrine stress test was done to induce long QT and catecholaminergic polymorphic ventricular tachycardia. Initial electrocardiogram before the stress test showed sinus rhythm with normal QTc interval of 401 msec. Patient was started in graded Epinephrine infusion under continuous ECG monitoring and noted progressive prolongation of actual QT interval and QTc during stage 1 recovery. There was no note of arrhythmia. The QTc reverted to pretest level at the end of test. She was discharged on Beta-blocker therapy and advised for ICD implantation.

Conclusion: The diagnosis of seizure and syncope with documented associated arrhythmia is a challenge to every clinician, warrants a good clinical history and further investigation. Among these, congenital long QT syndrome as a cause in a young patient with recurrent seizure and syncope with negative initial diagnostic work up and normal QTc on initial electrocardiogram could pose a dilemma. Early diagnosis and treatment can prevent sudden cardiac death in patients with long QT syndrome. Provocative testing with catecholamine, such in this case, Epinephrine, can unmask concealed type 1 long QT syndrome with a high level of accuracy. It may help differentiate patients with suspected LQTS from normal and may distinguish one genetic defect from another.