## AUTOSOMAL DOMINANT PARTIAL EPILEPSY WITH AUDITORY FEATURES: DESCRIPTION OF A TUNISIAN JEWISH FAMILY NOT LINKED TO LGI1 GENE W. Simri

Israel

waleeds@netvision.net.il

BACKGROUND AND OBJECTIVES: Autosomal dominant partial epilepsy with auditory features (ADPEAF) is rare familial epilepsy with onset in adolescence or early adulthood. In some families mutations in Leucine-rich glioma inactivated (LGI1) gene have been identified. We describe a new Israeli Tunisian Jewish family with ADPEAF.

METHODS: Clinical characteristics and results of EEG, computed tomography (CT), and magnetic resonance imaging (MRI) were evaluated in four affected members of the family. Genetic counseling was performed and a four generation pedigree drawn. Sequencing of the LGI1 gene was performed on one affected individual.

RESULTS: Four affected members from two generations had rare and drug-responsive adult onset tonic-clonic seizures constantly preceded by an auditory aura. The most common auditory symptoms were simple; talking, buzzing, ringing and loss of hearing bilateral less with visual, olfactory or vertiginous sensory disturbance. Routine and sleep electroencephalograms were normal. MRIs of brain were normal. Sequencing of the LGI1 gene did not reveal the causative mutation in the affected individual tested.

CONCLUSIONS: ADPEAF is a distinct condition with homogeneous clinical features. Genetic findings are consistent with linkage to chromosome 10q24,transmission with high penetrance, benign course, and seizures well controlled with standard antiepileptic drugs.LGI1 is not homologous to any known ion channel; the mechanism by which it causes epilepsy is unknown.

LGI1 is not responsible for the disease in the family presented here. Further genetic investigations will follow to hopefully identify a possible novel causative gene and mutation responsible for ADPEAF in this family