Landau-Kleffner syndrome is a rare disorder that affects twice as many males as females. Affected siblings and discordant monozygotic twins have been reported rarely. In additional to language regression, the diagnosis requires the presence of severely epileptiform activity on EEG, particularly during non-REM sleep. Additional testing may include magnetoencephalography. Brain imaging with magnetic resonance imaging (MRI) is recommended to exclude structural lesions since several cases have resulted from brain tumors. Other testing including behavioral and/or brainstem evoked audiometry and standardized psychometric and speech/language testing are helpful to exclude hearing loss and provide the basis for therapies to aide in recovery.