

# Ohtahara syndrome

Early infantile epileptic encephalopathy syndrome (EIEE), also known as Ohtahara syndrome, is an age-dependent epileptic encephalopathy syndrome defined by clinical features and electroencephalographic findings. Epileptic disorders with refractory seizures beginning in the neonatal period and/or early infancy have a potential risk of premature mortality, including sudden death. We aimed to identify the causes of death in EIEE and conducted a literature survey of fatal outcomes.

We performed a literature search in MEDLINE, EMBASE, and Web of Science for data from inception until September 2017. The terms “death sudden,” “unexplained death,” “SUDEP,” “lethal,” and “fatal” and the medical subject heading terms “epileptic encephalopathy,” “mortality,” “death,” “sudden infant death syndrome,” and “human” were used in the search strategy. The EIEE case report studies reporting mortality were included.

The search yielded 1360 articles. After screening for titles and abstracts and removing duplicate entries, full texts of 15 articles were reviewed. After reading full texts, 11 articles met the inclusion criteria (9 articles in English and 2 in Japanese, dated from 1976 to 2015). The review comprised 38 unique cases of EIEE, 17 of which had death as an outcome. In all cases, the suppression-burst pattern on electroencephalographies (EEGs) was common. Most cases (55%) involved male infants. The mean (standard deviation [SD]) age at onset of seizure was  $19.6 \pm 33$  days. The mean (SD) age at death was  $12.9 \pm 14.1$  months. Most infants (58.8%) survived less than one year. The cause of death was described only in eight (47%) patients; the cause was pneumonia/respiratory illness or sudden unexpected death in epilepsy (SUDEP).

The results show EIEE as a severe disease associated with a premature mortality, evidenced by a very young age at death. Increasing interest in the detection of new molecular bases of EIEE is leading us to a better understanding of this severe disease, but well-reported data are lacking to clarify EIEE-related causes of death <sup>1)</sup>.

<sup>1)</sup>

Radaelli G, de Souza Santos F, Borelli WV, Pisani L, Nunes ML, Scorza FA, da Costa JC. Causes of mortality in early infantile epileptic encephalopathy: A systematic review. *Epilepsy Behav.* 2018 Jun 12;85:32-36. doi: 10.1016/j.yebeh.2018.05.015. [Epub ahead of print] Review. PubMed PMID: 29906699.

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