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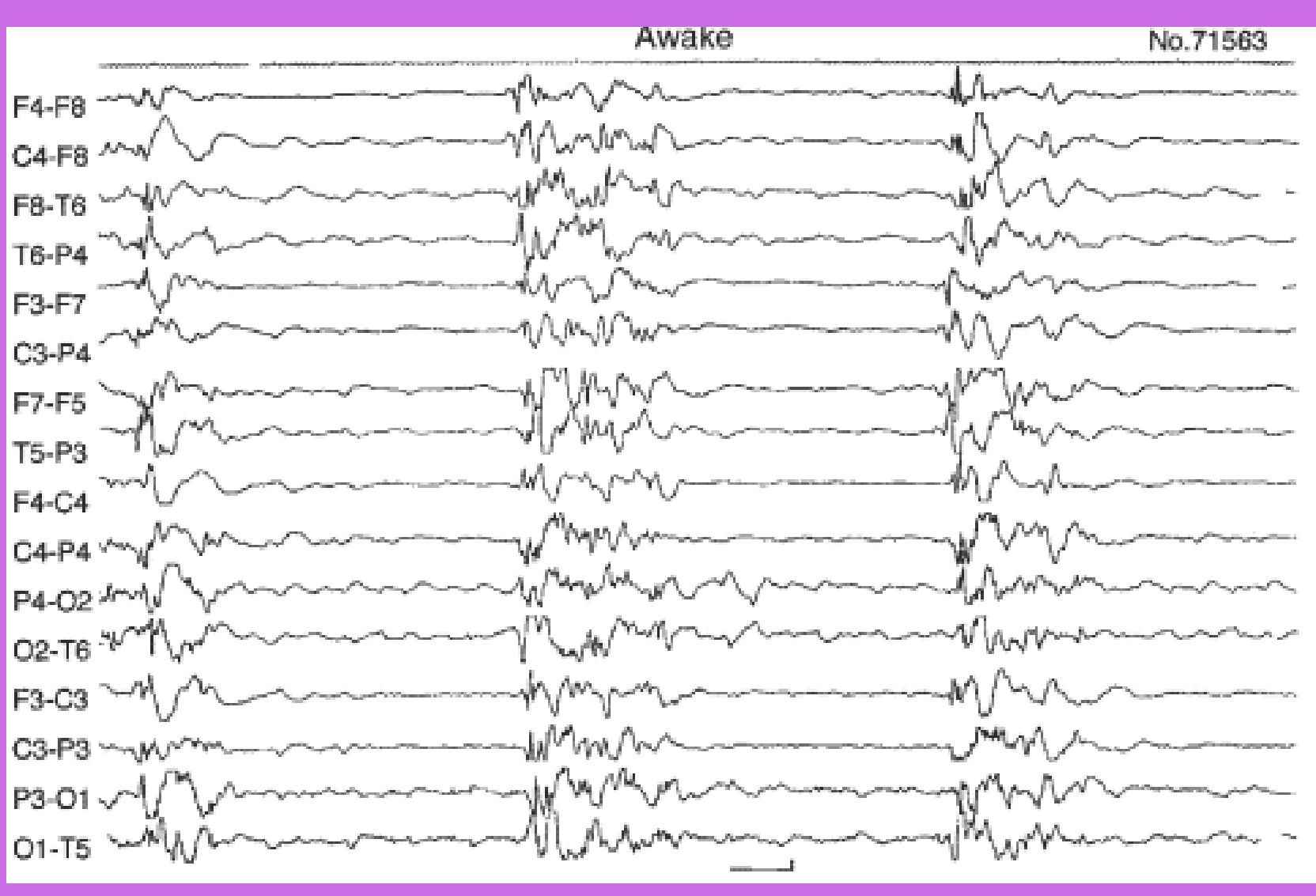
Ohtahara Syndrome

Ohtahara syndrome is also called early infantile epileptic encephalopathy (EIEE) a progressive epileptic encephalopathy. Painful spasms and partial seizures characterize Ohtahara syndrome. Symptoms present within the first few months of life. EIEE is a debilitating progressive neurological disorder involving cognitive disabilities and not easily managed seizures.

The majority of Ohtahara syndrome seizures are shock-like jerks of muscle, tonic, and myoclonic. These attacks are uncommon, setting this syndrome apart from early myoclonic encephalopathy.



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Symptoms

Children with Ohtahara syndrome seizures often exhibit an upward eye gaze, dilated pupils, stiffening of the muscles, and altered breathing, but may also experience focal seizures. Ohtahara syndrome has a characteristic pattern recognizable on an electroencephalogram (EEG). Brain malformation or genetic mutations often cause Ohtahara syndrome. Metabolic causes are less common.

Infants with Ohtahara syndrome can experience several categories of seizures. The most common seizure category in Ohtahara syndrome includes Focal seizures, Tonic-Clonic seizures, and Myoclonic seizures.

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Symptoms

Focal seizures, usually involve one part of the body or one side of the body. Repetitive jerking movements are common, and they last for a few seconds. Focal seizures are characterized by clonic episodes accompanied by impaired consciousness. It is often difficult to recognize whether an infant's consciousness is impaired.

Generalized tonic-clonic seizures involve movements of the legs and arms, usually involving both sides of the body. Babies experience head movements or a jerking eye during a generalized tonic-clonic seizure. The seizures also involve impaired consciousness, but it can be challenging to recognize that an infant has impaired consciousness.

Infantile spasms are quick, sudden movements that may last for only a second. Infantile spasms often involve the neck and both arms or legs.

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Symptoms

Myoclonic seizures involve repetitive jerking movements affecting one or both sides of the body. They may last for a few minutes and are generally followed by exhaustion and a period of sleeping.

Tonic seizures are the most common type of seizure in Ohtahara syndrome—characterized by a stiffening of the arms and legs, usually lasting a few seconds.

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Treatments

An early diagnosis and start of treatment can be crucial in achieving a better long-term prognosis. Consult with a palliative care team to ensure the caregivers and medical team work toward the same set of goals. Several medications help control and lessen the severity of Ohtahara Syndrome. Unfortunately, anti-seizure drugs are often not very effective in controlling this disorder.

Corticosteroids, ACTH, or prednisolone, are occasionally helpful. The ketogenic diet (high fat, low carbohydrate) is appropriate in a minority of cases. Epilepsy surgery may be beneficial for those with focal brain lesions (abnormal development of one area/side of the brain)
The most current research suggests Chloral hydrate is effective for kids with cryptogenic Ohtahara syndrome.

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Treatments

Seizures diminished or disappeared within 36 hours after chloral hydrate therapy and have not recurred. Electroencephalogram (EEG) results demonstrate marked improvement after treatment. Chloral hydrate may be helpful as a treatment for severe epileptic encephalopathies.

There is no cure for Ohtahara syndrome. Sadly many children afflicted with Ohtahara syndrome will not survive beyond early childhood. Survivors are often left with cognitive and severe physical disabilities.