Alternating Hemiplegia of Childhood

A rare neurological disorder marked by **recurrent episodes of paralysis** affecting each side of the body.

Etiology: majority caused by a heterozygous mutation in the ATP1A3 gene.
Rarely caused by mutations in ATP1A2.

PRESENTATION

Phase 1 (birth-1 year)	Phase 2 (1-5 years)	Phase 3 (>6 years)
▪ **Dystonia** ▪ Abnormal **ocular movements** ▪ Occasional **hemiplegic spells**	▪ **Hemiplegic spells**: unilateral or bilateral, lasting minutes to days ▪ **Loss of developmental milestones,** recovered over weeks or months as episode clusters subside ▪ **Focal seizures**	▪ Persistent **developmental delay** ▪ Fixed **neurological deficits** ▪ **Attacks** of dystonia, hemiplegia, and epileptic seizures

Migratory hemiplegic episodes (unilateral to opposite side or bilateral) are pathognomonic.
Episodes of hemiplegic attacks tend to **decrease later in life**

TRIGGERS

- Excitement
- Emotional stress
- Exposure to water
- Fatigue
- Trauma
- Hot weather
- Physical activity
- Cold weather
- Illness
- Loud noise
- Bright light
- Menstruation

DIAGNOSTIC CRITERIA

1) Onset of paroxysmal symptoms **<18 months**
2) Repeated **attacks of hemiplegia that** *alternate in laterality*
3) Episodes of **quadriparesis** or **plegia** as a separate attack or as a generalization of a hemiplegic event
4) Other **paroxysmal symptoms** either concurrent with or independent of hemiplegic attacks
5) Relief from symptoms upon **sleep**
6) Evidence of **developmental delay** or **neurological findings**

COMORBID CONDITIONS

- Epileptic seizures
- Developmental delay
- Intellectual disability
- Migraine
- Fine and gross motor delays
- Cardiac dysfunction
- Sleep disorders
- Movement disorders

INVESTIGATIONS

- Genetic testing confirms diagnosis
- EEG + MRI
- Others: sleep studies, ECG, echo, developmental, neuropsychological and psychiatric evaluations as indicated

MANAGEMENT

Acute Management	Preventative Therapy
▪ Trigger avoidance ▪ Sleep *(sedatives may be used)*	▪ Flunarizine *(reduction in frequency, severity, and duration of spells)* ▪ Antiseizure medications *(if seizures)*

Multidisciplinary approach

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