Infantile Spasms Care Unit: Pediatric Acute Care, Epilepsy Monitoring Unit, Emergency Department

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DISEASE OVERVIEW

Infantile Spasms, or epileptic spasms, is a rare (2-4 per 10,000 live births) epileptic encephalopathy that occurs before age 2 and presents with a constellation of the following characteristics:

- Brief myoclonic jerks, usually occurring in clusters. Characteristic spasms consist of bilateral arm extension and leg flexion when lying supine. When held in a sitting position, caregivers will often describe head drop and/or a "crunching" motion.
- Hypsarrhythmia, a chaotic high-amplitude background pattern on EEG.
- Developmental plateau or regression

Presentation with all three is often referred to as West Syndrome of Infantile Spasm Syndrome.

Infantile Spasms commonly presents before 12 months of age, with the mean onset of 4 months.

Etiology can be varied and includes structural brain malformations (congenital and acquired), genetic, and metabolic causes. For almost a third of cases, an underlying cause is not identified.

NEURO EXAM PEARLS

Detailed description and frequency of abnormal movements should be monitored for and documented.

Thorough cutaneous exam should be performed, including Wood's lamp, to rule out any possible neuro-cutaneous etiology.

Developmental milestones should be assessed.

Assessing signs or symptoms of concurrent infectious illness is also necessary and may impact treatment decisions.

MANAGEMENT STRATEGIES/ NURSING IMPLICATIONS

Timely, and effective, treatment of Infantile Spasms is correlated with better neurodevelopmental outcomes, thus Infantile Spasms is often treated with urgency.

Routine or continuous EEG will be performed to confirm diagnosis. Infantile Spasms is often initially misdiagnosed as GERD, Sandifer Syndrome, or hyperactive Moro reflex.

MRI may be performed to identify any underlying structural causes. Genetic and/or metabolic testing may be indicated based on history and physical.

Traditional seizure rescue medications are not indicated for infantile spasms.

Close monitoring for therapy side-effects and EEG follow-up are necessary.

MEDICATION /SPECIALIZED LABS

There are three commonly accepted treatments for Infantile Spasms:

- Adrenocorticotropin hormone (ACTH, Acthar Gel)
- High dose corticosteroids (prednisolone)
- Vigabatrin (Sabil, Vigadrone)

Other traditional anti-seizure medications are not indicated for Infantile Spasms.

Adrenocorticotropin hormone (ACTH, Acthar Gel)

- Traditional 4-week course (2-week high-dose with a 2-week wean)
- Intramuscular injections
- Substantial side-effect profile with close monitoring during therapy required
- Immunosuppression, rare cardiac hypertrophy, and possible wean-related adrenal insufficiency are the most concerning possible side effects
- Costly, only available through specialty pharmacies

High-dose oral corticosteroids (OCS)

- Prednisolone
- Traditional 4-week course (2-week high-dose, 2-week wean)
- Substantial side-effect profile (although less than ACTH) with close monitoring during therapy required
- Immunosuppression and possible wean-related adrenal insufficiency are the most concerning possible side effects

Vigabatrin (Sabil, Vigadrone)

- Titrated up for lowest effective dose, duration of treatment varies
- Powder formulation that requires mixing prior to oral administration
- FDA Risk Evaluation and Mitigation program due to risk of permanent peripheral vision loss
- Baseline and periodic vision testing is required
- Costly, only available through specialty pharmacies

Based on results of a recent study, some centers are now prescribing dual therapy, hormonal therapy (ACTH or OCS) and vigabatrin.

Ketogenic diet has also been used, however, not widely accepted as a first-line treatment.

TEAM QUESTIONS/COMMUNICATION

Close coordination of follow up monitoring and care is required.

If ACTH or OCS is chosen, close follow up during therapy must be coordinated through either home health, primary care provider, or the neurology clinic. If inpatient, the follow-up plan should be confirmed prior to discharge.

If ACTH or vigabatrin is chosen as treatment, insurance prior authorization is usually required and access to medication must be coordinated with a specialty pharmacy.

If ACTH or OCS is started, ensure uninterrupted therapy is available prior to discharge. ACTH or OCS should not be abruptly discontinued due to risk of adrenal insufficiency.

PATIENT/FAMILY/CAREGIVER TEACHING SUPPORT

Adrenocorticotropin hormone (ACTH, Acthar Gel)

- Instructed on how to give IM injections
- Follow-up and side effects
- Weaning schedule

High-dose oral corticosteroids (OCS)

- Follow-up and side effects
- Weaning schedule

Vigabatrin (Sabil, Vigadrone)

- Titration schedule and how to mix
- Follow-up and side effects
- Vision testing

Neurology and EEG follow-up plan, along with any further diagnostic plan should be coordinated and communicated to caregivers.

Infants diagnosed with Infantile Spasms qualify for Early Intervention and should be provided with appropriate referrals.