



Management of infantile spasms (IS) during the COVID19 pandemic

Timely diagnosis and management of new onset infantile spasms (IS) is among the highest clinical priorities for a child neurologist. However, the COVID-19 pandemic requires clinicians to minimize in-person healthcare visits. Below, please find guidance to assist with clinical decision making for IS during this public health crisis.

Diagnosis

- The initial clinical visit may be performed by telemedicine or video conference.
- Obtain home video that captures several consecutive events.
- Ask about light spots on the skin (i.e., as a possible indicator of tuberous sclerosis complex.¹)
- EEG confirmation is strongly encouraged.² If obtained, include at least one sleep cycle. Outpatient EEG is preferred over inpatient admission.
- After diagnosis, if the etiology is uncertain, brain MRI should be performed urgently (if available).

Key elements of history for the diagnosis of infantile spasms

Age of Onset 2 - 9 months, occasionally up to 2 years of age

Characteristics Often flexor movements (though can be extensor) of arms, legs, and/or neck lasting about 1 second; slower than myoclonus, but briefer than a tonic seizure

Clustering Infantile spasms occur at regular intervals (every 15 - 45 seconds) within clusters lasting several minutes

Timing Infantile spasms often occur after awakening

Treatment

- Select from among ACTH (adrenocorticotropic hormone), high dose prednisolone (6-8mg/kg/day), and vigabatrin, unless contraindications to all three.
- For etiologies other than tuberous sclerosis complex, prefer outpatient initiation of prednisolone.³
- For tuberous sclerosis complex, prefer vigabatrin if immediately available. If anticipating a treatment delay, consider initiating prednisolone until vigabatrin can be obtained.
- Avoid non-standard therapy as first treatment choice (i.e., avoid topiramate, ketogenic diet, etc.).
- For prednisolone or ACTH, consider GI prophylaxis with a proton pump inhibitor or H2 blocker.
- For prednisolone or ACTH, write one prescription with 2 weeks at a high dose *and* a 2-week taper.

Suggested protocol for prednisolone dosing:

Days	Dose – Prednisolone
1-14	8 mg/kg/day (÷ QID, max 60 mg/day)
15-17	6 mg/kg/day (÷ TID)
18-20	4 mg/kg/day (÷ BID)
21-23	2 mg/kg/day (QD)
24-26	1 mg/kg/day (QD)
27-29	0.5 mg/kg/day (QD)

Follow up

- Home measurement of blood pressure and/or laboratory testing can be deferred, unless there are clinical indications beyond routine monitoring. Abnormalities typically occur with clinical changes (i.e., lethargy or reduced feeding). Follow closely via telehealth, video conference, or phone calls.
- Assess treatment response via telehealth or video conference 10-14 days after treatment initiation.
 - If clinical spasms *continue*, add or modify treatment *without* confirmatory EEG.
 - If clinical spasms have *resolved* (or if the caregiver is uncertain), a repeat EEG, including at least one sleep cycle, is strongly encouraged. Outpatient EEG is preferred over inpatient admission.

Additional References

Quality of care for infantile spasms (AAN). <https://bit.ly/2X9ympq>

Neurophysiology studies during COVID19 (ACNS). <https://www.acns.org/practice/covid-19-resources>.

Epilepsy care during COVID19 (AES). <https://bit.ly/2UVLcoE>

¹ Tuberous sclerosis complex is a clinical diagnosis based on exam and imaging and does not require genetic confirmation.

² Hypsarrhythmia is not required for diagnosis of IS. Some epileptiform discharges are typically present. A normal EEG, including sleep, is reassuring against IS. Follow up frequently if clinical suspicion is high.

³ Prednisolone is inexpensive, readily available, and orally administered. It can be started outpatient. It does not require training of parents or caregivers, use of subspecialty pharmacies, or pre-approval by insurance. Note that ACTH and vigabatrin are still acceptable treatments, especially for admitted patients, so long as the duration of hospitalization can be minimized. This document should in no way be used to justify denial of insurance coverage for ACTH or vigabatrin.