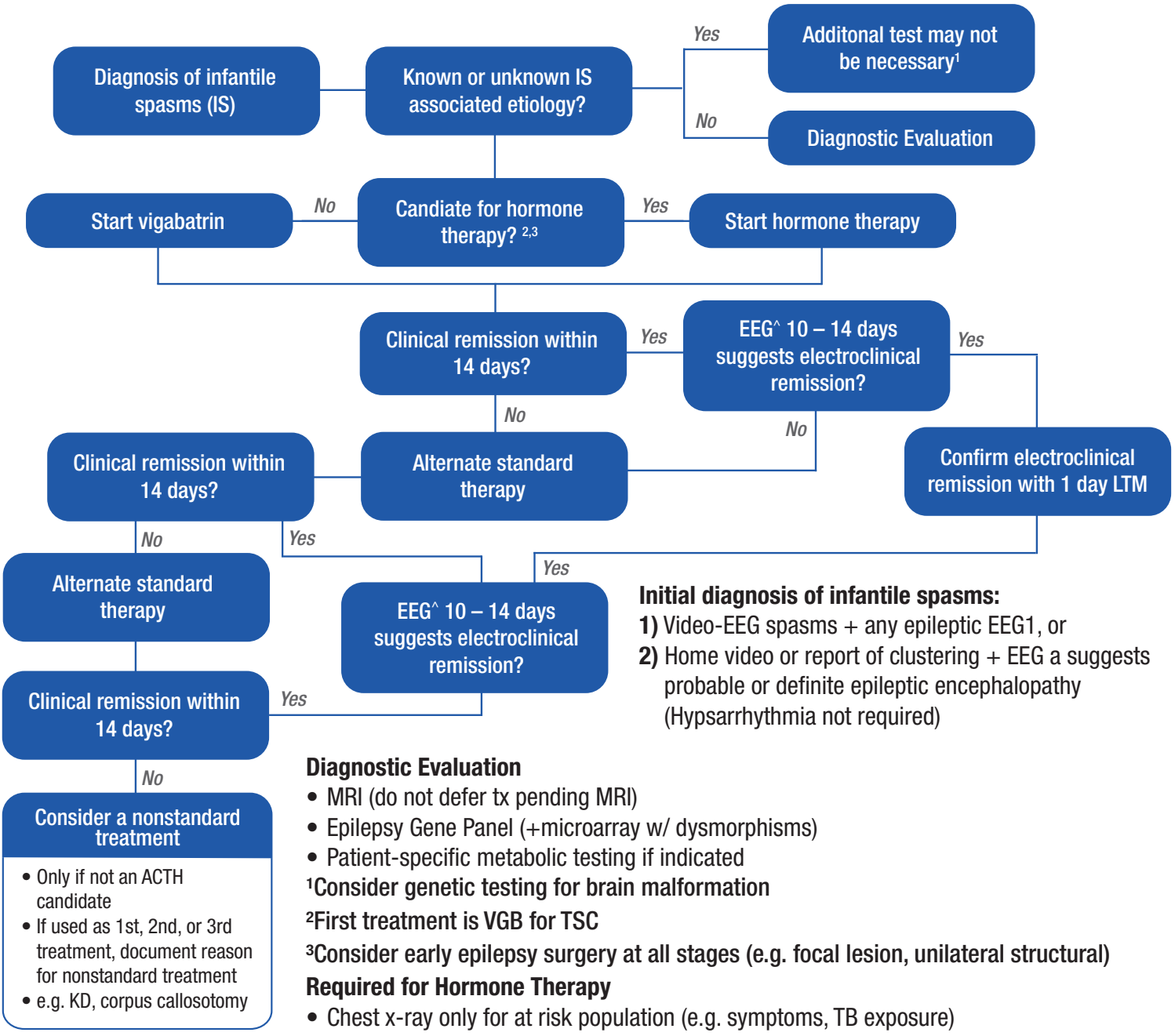
Infantile Spasms Algorithm 2022

- Hormone therapy as 1st treatment if eligible
- Standard therapy as 1st, 2nd & 3rd treatment if eligible (Standard therapy: prednisolone, ACTH, VGB, epilepsy surgery)
- Clinic and EEG evaluation within 10 – 14 days if eligible
- Start next standard treatment within 2 weeks if eligible



Initial diagnosis of infantile spasms:

1) Video-EEG spasms + any epileptic EEG1, or

2) Home video or report of clustering + EEG a suggests probable or definite epileptic encephalopathy (Hypsarrhythmia not required)

Diagnostic Evaluation

- MRI (do not defer tx pending MRI)
- Epilepsy Gene Panel (+microarray w/ dysmorphisms)
- Patient-specific metabolic testing if indicated

[1]Consider genetic testing for brain malformation

[2]First treatment is VGB for TSC

[3]Consider early epilepsy surgery at all stages (e.g. focal lesion, unilateral structural)

Required for Hormone Therapy

- Chest x-ray only for at risk population (e.g. symptoms, TB exposure)
- Add immunocompromised to problem list
- Famotidine 0.5 mg/kg/dose BID or similar

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Days	Dose – Prednisolone (3mg/ml)	Dose – Natural ACTH
1-14	8 mg/kg/day (÷ QID, max 60 mg/day)	150 IU/m^2/day (÷ BID)
15-17	Same individual ml dose TID	30 IU/m^2/day (QD AM)
18-20	Same individual ml dose BID	15 IU/m^2/day (QD AM)
21-23	Same individual ml dose QD	10 IU/m^2/day (QD AM)
24-26	½ individual ml dose QD	10 IU/m^2/day (QOD AM) 3 total doses
27-29	¼ individual ml dose QD	

Days	Vigabatrin*
1	About 50 mg/kg/dose once
2-4	About 100 mg/kg/dose (÷ BID)
>5	About 150 mg/kg/dose (÷ BID)

*Slow titration or reduce dose for side effects (e.g. sedation, airway)

Vigabatrin

- Script from VGB-approved doc, send to insurance-appropriate specialty pharmacy (consult NCH pharmacist)
- Ophthalmology eval within 4 weeks, f/u intervals per ophthalmology
- Avoid using w/ PB & benzodiazepines (e.g. clobazam): aspiration risk
- Without early clinical or EEG improvement, consider weaning off
- D/C after 6 months treatment if possible^

^Longer courses (e.g. 9 – 12 months) may be recommended

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Critical Documentation

- Day of spasm onset (if unknown, estimate or report unknown)
- Document risk of death with hormone therapy
- Add immunocompromised to problem list
- If hormone not used as initial treatment, document reason
- If nonstandard used as 1st, 2nd, or 3rd treatment, document reason
- If no follow-up in 10-14 days, document reason
- Neurology resident to create/confirm treatment calendar
- Inpatient team to ensure EEG and clinic f/u or document handoff
- Send standardized e-mail w/ hormone calendar to IS team infantilespasms@nationwidechildrens.org

Important Points

- Avoid reflex diagnostics, consider relevance to patient
- B6 100 mg infusion for select Pt (e.g. unknown etiology, early-onset)
- RN will call at 1 week for update
- Avoid benzodiazepines for clusters (generally ineffective)
- Post-treatment EEG may be deferred for ongoing spasms
- LTM to confirm remission (unless would not change management)
- Vaccines after hormone therapy: non-live may start after completion of hormone, live vaccines may start after one month of completion
- Consider alternate hormone with prior Pred or ACTH failure
- Referral for developmental evaluation at baseline and follow-up

Special Populations

- Patients with congenital CMV and HSV are eligible for hormone therapy, see ID treatment algorithm
- Patients with TSC are eligible for hormone therapy after failed VGB, monitor cardiac rhabdomyoma for enlargement

Monitoring for Hormone Side Effects may Include

- BP monitoring at 10-14d neurology f/u*
- Physician (PCP, ER) for fever >100.4 F (ER with lethargy)

*Low incidence of endorgan dysfunction, false positives common – often due to irritability/movement or improper technique

Blood Pressure Monitoring and Treatment

- Use right arm when child is sleeping or calm
- For BP >120 systolic, repeat in 15 minutes when calm
- If repeat >120, give isradipine 0.1 mg/kg PO, recheck BP 1 Hr later
- If still >120 with treatment, neurology to call nephrology
- If <120, repeat BP 6 Hr later, may repeat isradipine q6 Hr PRN
- If requiring repeated doses of isradipine, give amlodipine 0.1 mg/kg/day, may be increased to BID to target systolic 100 – 110

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Infantile Spasms Shared Decision-Making

- ACTH (Acthar Gel), prednisolone (Orapred), and vigabatrin (Sabril) are all standard treatments for infantile spasms.
- However, hormone therapy (prednisolone or ACTH) is more effective than vigabatrin and is the preferred initial therapy for eligible patients.
- Prednisolone is a liquid given by mouth for one month (available at any pharmacy).
- ACTH is a refrigerated liquid given by intramuscular injection for one month (specialty pharmacy only).
- Vigabatrin is a powder mixed with water, milk or juice given by mouth for approximately six months if effective (specialty pharmacy only).

Possible Side Effects of Prednisolone and ACTH	
Common	Irritability, poor sleep, increased appetite, weight gain, water retention with puffy appearance
Less Common	Elevated blood pressure but low risk for related complications
Rare	Bleeding ulcer, temporary diabetes, low potassium, adrenal insufficiency, weakened immune system, death due to infection

Possible Side Effects of Vigabatrin	
Common	sedation, reduced tone (more floppy or limp)
Rare	peripheral vision loss (no known risk with a trial of less than 2 months, up to 5% at six months)

- ACTH and vigabatrin are FDA-approved for infantile spasms. Prednisolone is not FDA-approved because it is an inexpensive medication and there is no incentive for a drug company to pursue FDA approval. The lack of FDA approval does not mean that prednisolone is less effective.
- Vigabatrin and prednisolone have a much shorter length of hospitalization because of challenges with insurance approval and specialty pharmacy delivery for ACTH. For many patients, prednisolone is an ideal choice because it is more effective than vigabatrin as a first treatment, is easier to give than ACTH, and has a shorter length of hospitalization compared to ACTH.
- Natural ACTH contains porcine (pig) products. This fact may be important to some families with religious restrictions.

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