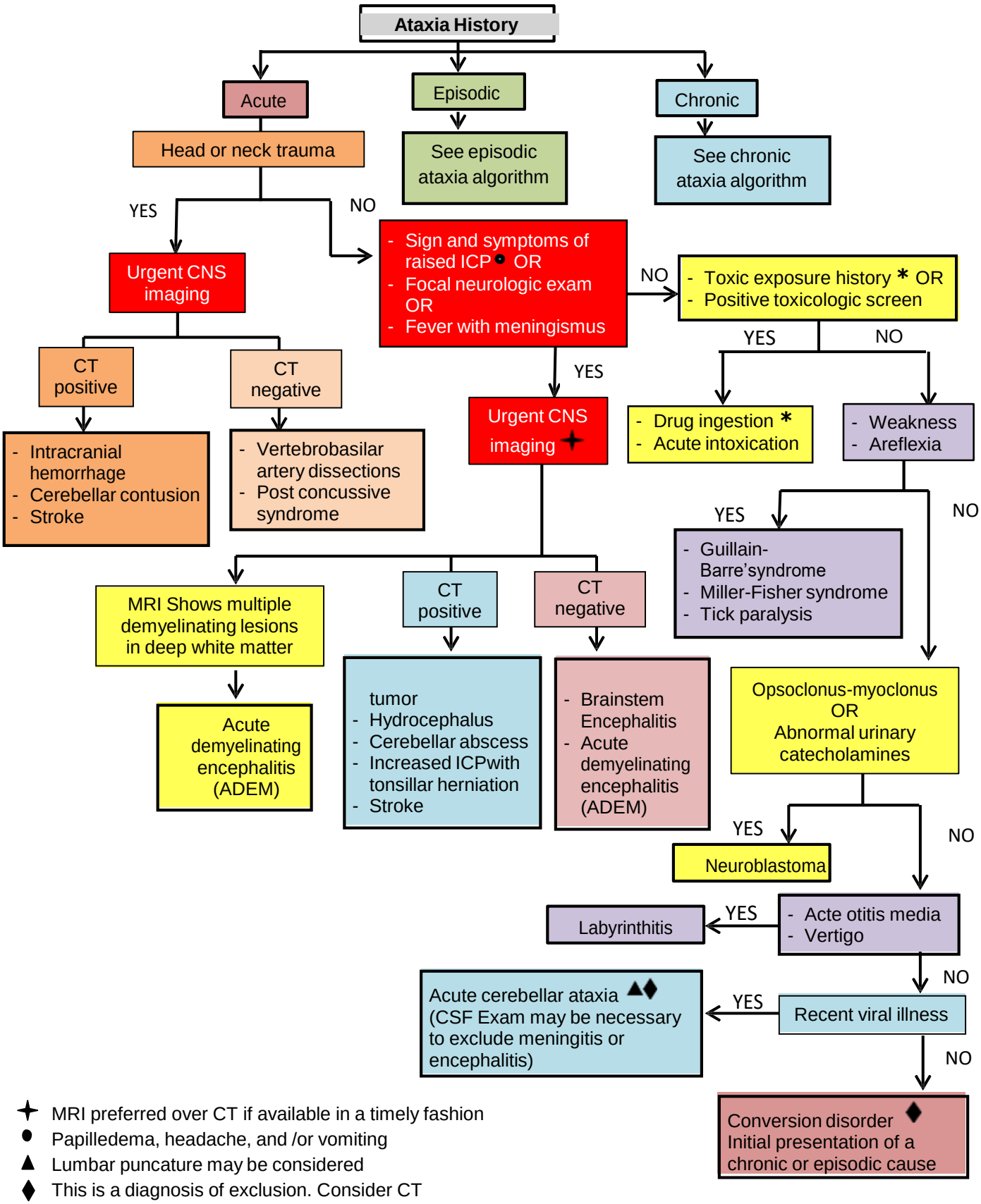
Approach to Acute Ataxia in Children



✦ MRI preferred over CT if available in a timely fashion

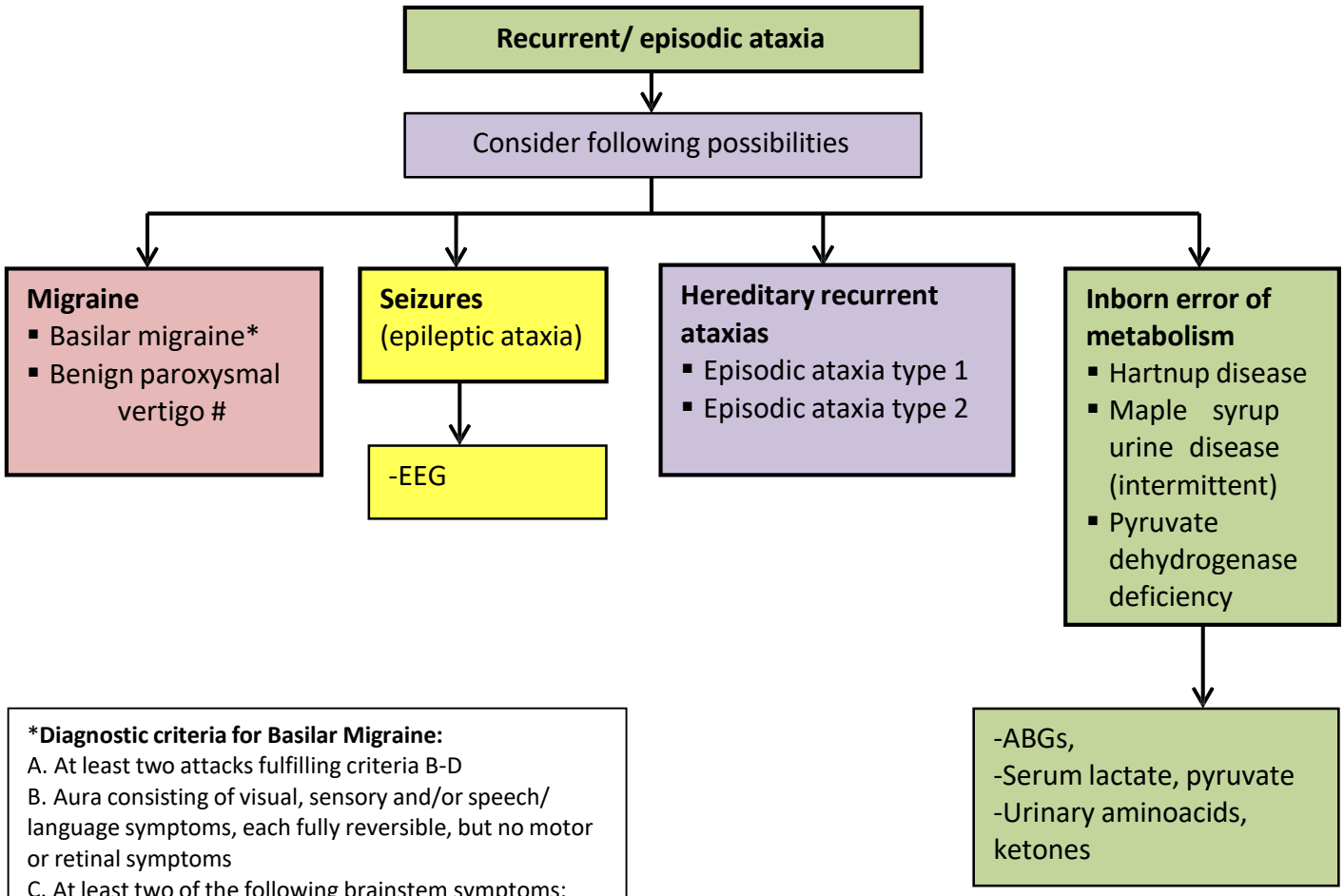
● Papilledema, headache, and /or vomiting

▲ Lumbar puncture may be considered

♦ This is a diagnosis of exclusion. Consider CT

*drugs (anticonvulsants), heavy metal poisoning

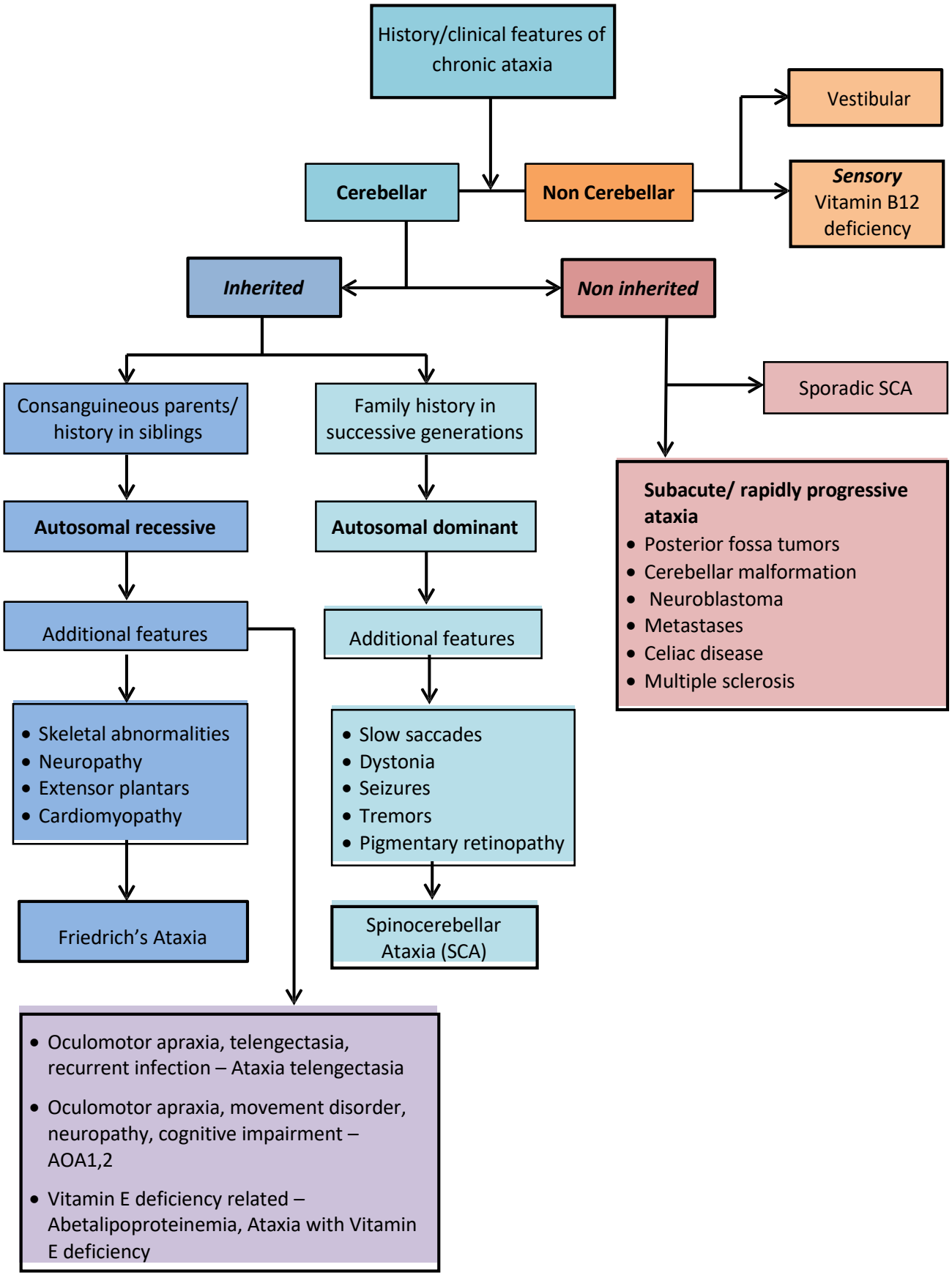
Approach to Recurrent/ episodic Ataxia in Children



***Diagnostic criteria for Basilar Migraine:**
A. At least two attacks fulfilling criteria B-D
B. Aura consisting of visual, sensory and/or speech/ language symptoms, each fully reversible, but no motor or retinal symptoms
C. At least two of the following brainstem symptoms:
1. Dysarthria
2. Vertigo
3. Tinnitus
4. Hypacusis
5. Diplopia
6. Ataxia
7. Decreased level of consciousness
D. At least two of the following four characteristics:
1. At least one aura symptom spreads gradually over _5 minutes, and/or two or more symptoms occur in succession
2. Each individual aura symptom lasts 5-60 Minutes
3. At least one aura symptom is unilateral3
4. The aura is accompanied, or followed within 60 minutes, by headache
E. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.

 # Diagnostic criteria for Benign Paroxysmal Vertigo:
A. At least five attacks fulfilling criteria B and C
B. Vertigo1 occurring without warning, maximal at onset and resolving spontaneously after minutes to hours without loss of consciousness
C. At least one of the following associated symptoms or signs:
1. Nystagmus
2. Ataxia
3. Vomiting
4. Pallor
5. Fearfulness
D. Normal neurological examination and audiometric and vestibular functions between attacks
E. Not attributed to another disorder.

Approach to Chronic Ataxia in Children



KEY NOTES:

Ataxia is incoordination or clumsiness of movement that is not the result of muscle weakness.

Cerebellar ataxia cause irregularities in the rate, rhythm, amplitude, and force of voluntary movements, especially at initiation and termination of motion resulting in

- Intention tremors
- Past pointing
- Inability to perform rapid alternating movements (dysdiadochokinesia).
- Dysarthric speech
- Nystagmus
- Ataxic gait

Sensory ataxia has no vertigo or dizziness, also spares speech, worsens when the eyes are closed (positive Romberg sign), and is accompanied by decreased vibration and joint position sense.

Vestibular ataxia has prominent vertigo (directional spinning sensations)and may cause past-pointing of limb movements, but spares speech.

Diagnostic Approach

When treating a child with ataxia, a flexible, stepwise approach is helpful.

- Clarify that ataxia is the movement disorder.
- Localize the lesion. Unilateral or predominantly midline cerebellar signs may indicate focal cerebellar pathology. Consider brain MRI. Possible causes of focal cerebellar disease include congenital malformation, neoplasm, demyelination, abscess, or vascular event. Treatment of focal neoplasms may be surgical and depends on the cause identified or suspected.
- Stratify into a time-course group—acute/subacute, static, chronic progressive, episodic.
- Look for associated features outside of the cerebellum that narrow the differential.
- For acute/subacute, likely acquired ataxias, assess for intoxications, signs of infection, or inflammation.
- For static ataxias, monitor clinically, consider brain MRI.
- For all other ataxias that could be inherited, take a careful family history and examine parents and siblings, if possible.
- In many cases where the problem appears to be static but is relatively mild, and no skills have been lost, a detailed diagnostic evaluation with laboratory testing and neuroimaging is not necessary. A follow-up examination in 6 to 12 months to ensure that there is no regression suffices.
- Neuroimaging should usually be obtained in

    1. In cases where other abnormalities are identified on general examination, suggesting the presence of a syndrome. In these cases, karyotyping or genomic hybridization studies should be considered as well.
    2. In cases with clearly asymmetric motor findings, microcephaly, or more severe motor problems. “Ataxic cerebral palsy” has lower prevalence than spastic forms. The American Academy of Neurology Practice Guideline for the evaluation of the child with cerebral palsy recommends that neuroimaging be obtained in children diagnosed with cerebral palsy.
    3. In cases where nystagmus, headaches in a young child, and acquired ocular malalignment are present.
    - The imaging modality of choice is magnetic resonance imaging (MRI), although ultrasound through the open fontanel in infants is sometimes a good choice.

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