■ Multimedia teaching material

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Epileptic Disorders

Early detection and evolution of hypsarrhythmia in a patient with subcortical band heterotopia

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- A 2-week-old full-term baby with XY genotype but female phenotype and bilateral ventriculomegaly on prenatal ultrasound presented with failure to thrive and episodes of right arm shaking
- Interictal EEG was initially normal (Figure 1A)
- Two months later, repeat EEG after a witnessed bilateral clonic seizure revealed independent bitemporal spikes (Figure 1B)
- Brain MRI showed band heterotopia along the lateral ventricles and mild ventriculomegaly (Figure 2)
- At 5 months, EEG obtained after breakthrough bilateral clonic seizures was significant for hypsarrhythmia (Figure 1C)
- After initiation of prednisolone, there was a notable improvement in hypsarrhythmia background (Figure 1D) after two weeks

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Figure 1. Evolution of hypsarrhythmia on serial EEG. A) Normal EEG at two weeks of age on neonatal montage. B) Abundant bitemporal spikes at two months of age on neonatal montage. C) Hypsarrhythmia at five months of age. D) Improvement in background, two weeks after initiation of prednisolone, although with persistence of interictal abnormalities. EEG settings: (A, C) sensitivity 10 uV/mm, LFF 1 Hz, HFF 70 Hz; (B, D) sensitivity 15 uV/mm, LFF 1 Hz, HFF 70 Hz.

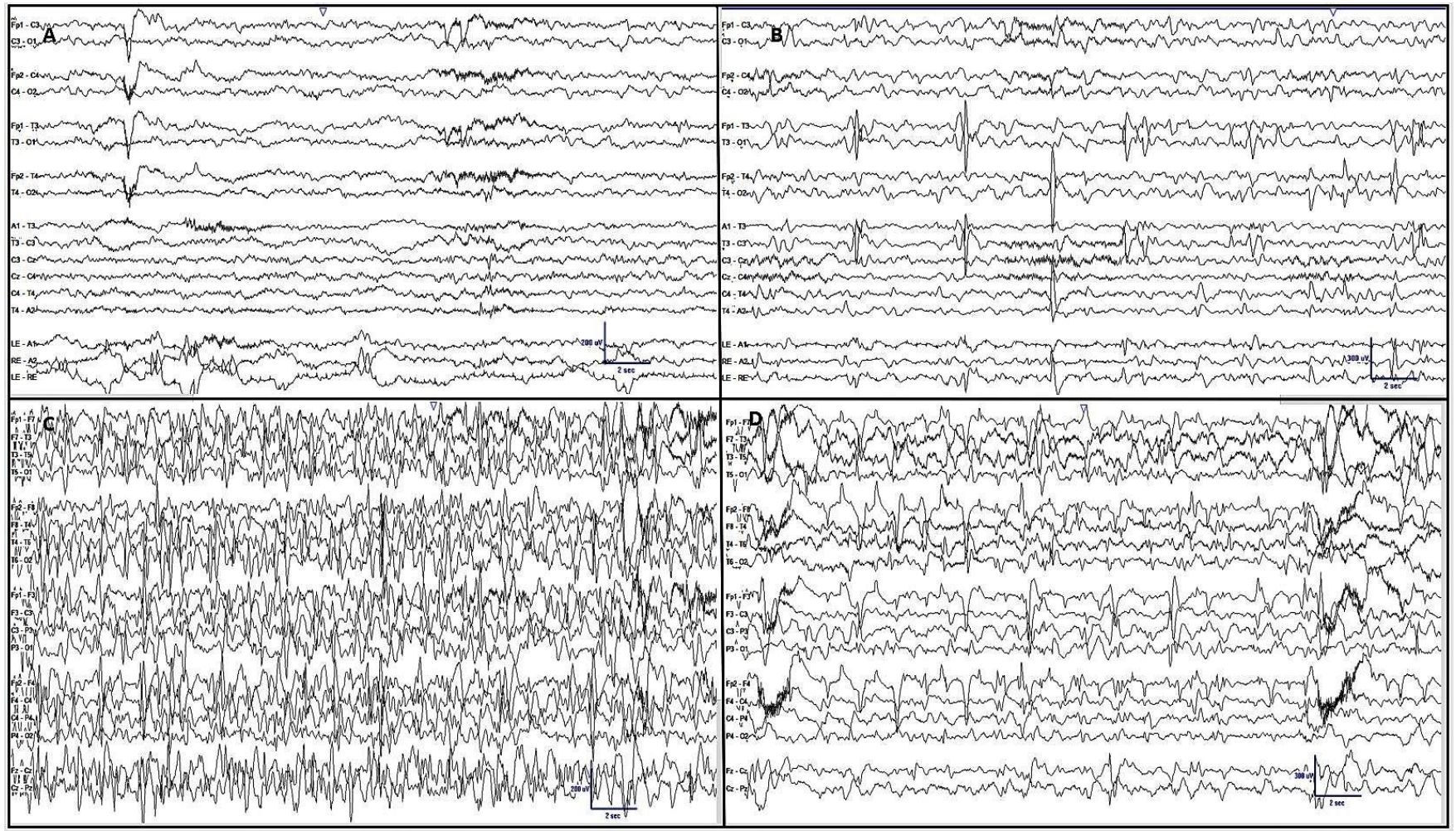

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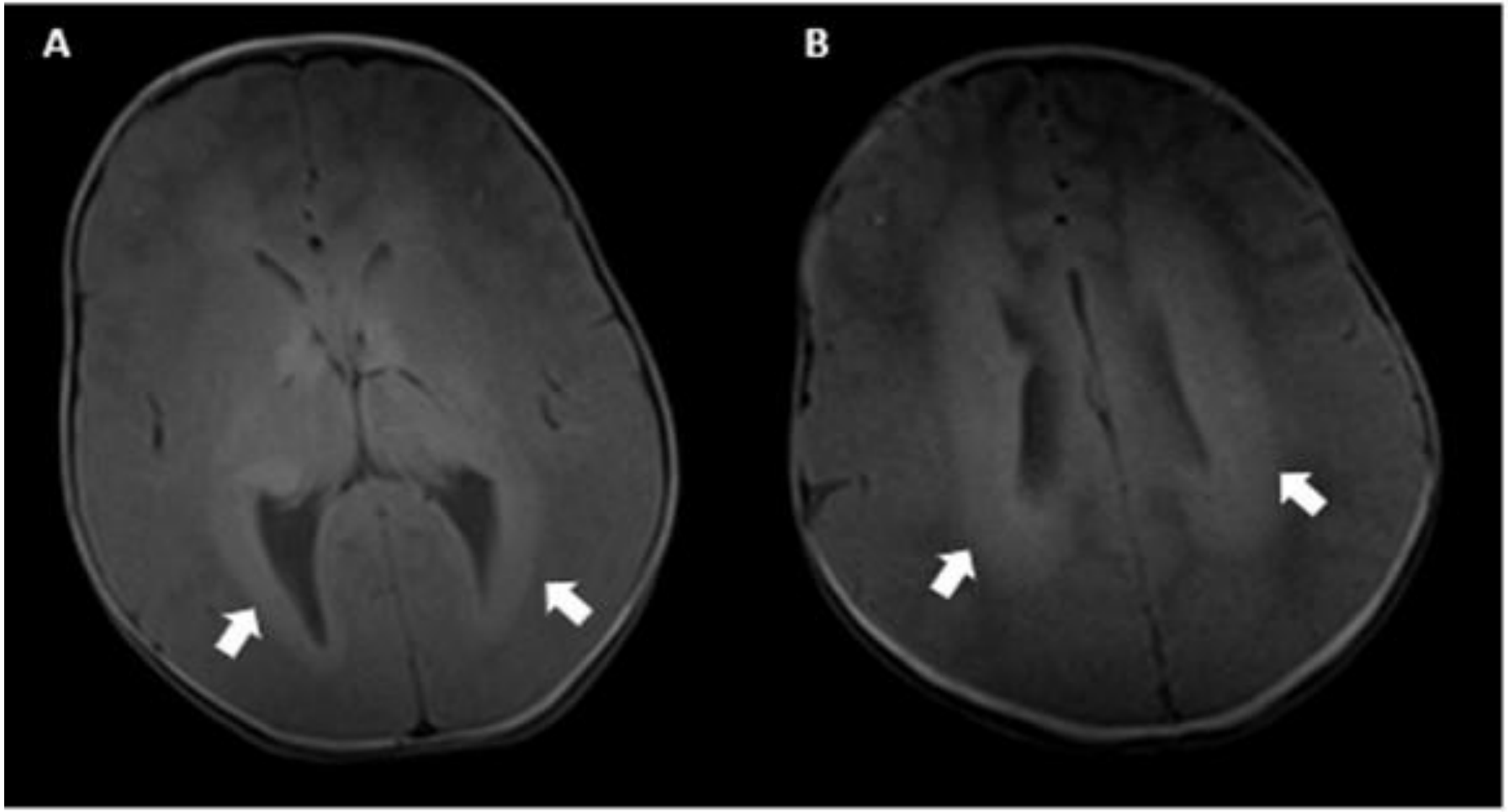
Figure 2. A, B) Brain MRI showing a band of abnormal signal surrounding the lateral ventricles symmetrically, which follows gray matter on T1 axial images, and bilateral ventriculomegaly.



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- No additional seizures were noted. The patient had global developmental delay since birth, however, no regression
- Genetic testing with epilepsy and brain malformation gene panels was unrevealing for an etiology of subcortical band heterotopia (SBH)
- SBH can present with infantile spasms with or without hypsarrhythmia and is often related to mutations in the *PAFAH1B1* or *XLIS* genes [1]
- Notably, our patient did not develop infantile spasms
- These findings support the importance of maintaining a low threshold for follow-up EEGs for hypsarrhythmia in patients with malformations of cortical development, such as band heterotopia despite unrevealing initial EEGs and an inconsistent semiology
- Early detection and treatment of hypsarrhythmia have been associated with an improved prognosis depending on the underlying etiology [2, 3]

References

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