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Hypermotor-tonic-spasms seizure sequence related to *CDKL5* deficiency disorder: a typical case

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Typical Hypermotor-Tonic-Spasms Sequence in CDKL5 Deficiency Disorder

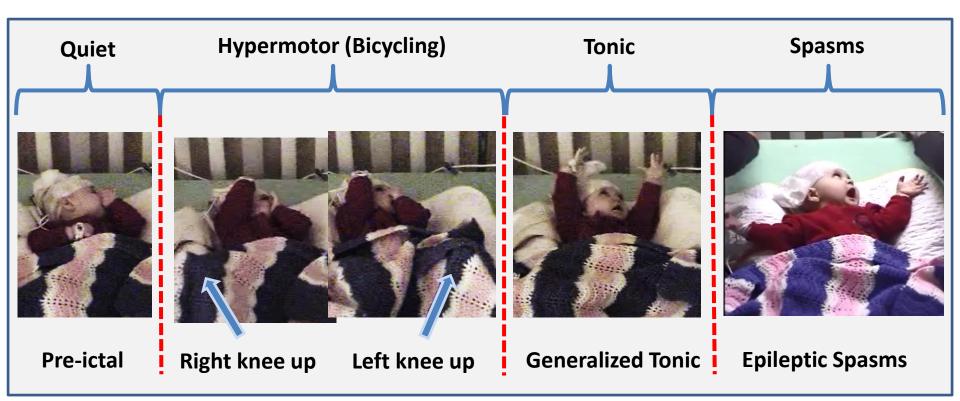


Figure 1: Patient with a typical hypermotor-tonic-spasms sequence associated with *CDKL5* deficiency disorder (CDD). Each phase was as follows: hypermotor for only 2 seconds with a pedaling movement of the legs; tonic for 20 seconds, and spasms for 4:35 minutes (shortened in the video).



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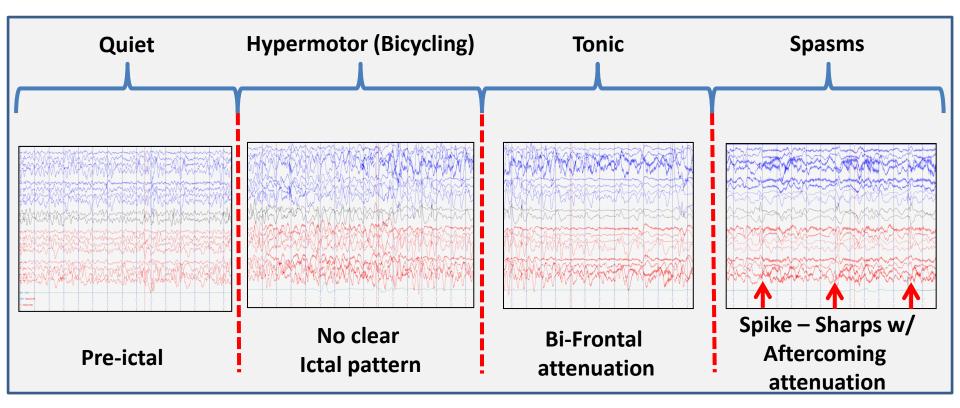
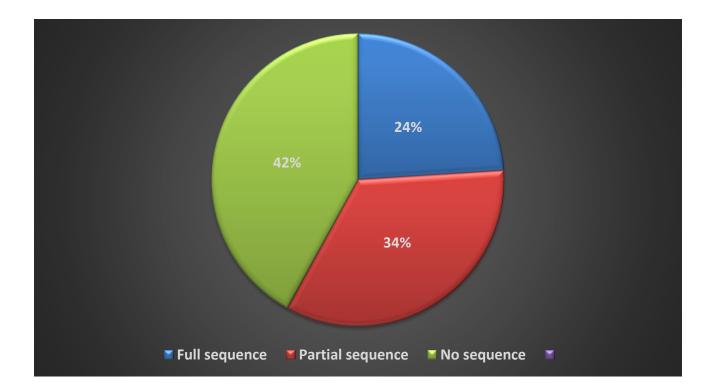


Figure 2: EEG settings: anterior-posterior bipolar montage, LFF: 1Hz, HHF: 70Hz, Notch: 60Hz, Sensitivity: 7uV/mm (except for spasms sampled at 15 uV/mm), paper speed: 30 mm/sec. Red arrows depict occurrence of spasms.



Percentage of patients with *CDKL5* deficiency disorder (CDD) showing complete, partial or no sequence



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Not all patients with CDD will have an ictal Hypermotor-Tonic-Spasms sequence; in fact, many will not. However, it can be fully seen in one of 4 patients with CDD; and partially present (mostly a combination of tonic – spasms) in 1 of 3 patients. Recognizing this pattern early can help in diagnosing and counselling families regarding CDD.