Background for NavNeuro Pediatrics Fact-Finding Case 2 - Jane Doe

Identifying Information:
Age: 6 years
Sex: Female
Handedness: Left
Education: Homeschooled in a homeschool co-op, family experienced with homeschooling
Race/Ethnicity: Asian/Chinese

Patient Overview:
Jane is a 6 year-old left-handed female with a history of Sturge-Weber syndrome with left-sided brain involvement associated with left hemispheric brain atrophy and focal epilepsy, described as likely consistent with electrical status epilepticus in sleep (ESES). Consistent with her history of Sturge-Weber syndrome, her medical history is consistent for leptomingeal angioma in the left hemisphere, right-sided hemiparesis and left-sided facial port-wine birthmark.

Reason for Referral
She was referred by Dr. Neurologist 1 of the Hospital Name for a baseline neuropsychological evaluation to evaluate her neurocognitive functioning in light of her medical history.

Family History/Early Developmental History
Jane was placed into an urban orphanage in China around 2 months of age and little information is available regarding her early history. She was adopted around 20 months of age. When adopted, was not speaking but displayed good problem-solving with how to open containers. Limited use of right hand and significant right-sided weakness. Walked at 26 months and started using 2 word phrases around age 3 years.

Medical History
Followed by 2 neurologists.
Brain MRI at 3 years notes left cerebral atrophy, left meningeal enhancement and a left sided venous angioma in the left frontal lobe. Findings reported left cerebral peduncle atrophy at the level of the midbrain and mild flattening of the left medullary pyramid. MRI report indicated normal appearance of Jane's right hemisphere. While at age 3 years Jane was not noted to have seizures, her history prior to adoption was largely unavailable.

Around age 4 ¼ years, Jane displayed episodes of new onset nighttime enuresis after previously sleeping through the night without bedwetting. Episodes of nighttime incontinence have continued to present. Neurology evaluation at age 5 years noted concern for complex partial seizures and an electroencephalogram (EEG) was ordered. Results of the EEG (age 5 years) indicated near continuous spike waves during sleep in the left hemisphere. Jane was started on Levetiracetam, and has since taken various anti-epileptic agents in an attempt to manage her seizures, though multiple EEG's have indicated persistent spike and wave in sleep with treatments attempted. Most recent EEG and note of evaluation findings from Dr. Neurologist 1 indicated likely electrical status epilepticus in sleep (ESES). Following consultation with Dr. Neurologist 2, recommendations for baseline neuropsychological assessment and 24-hour EEG monitoring was proposed to assist her treatment team with decision making and monitoring of Jane's neurocognitive status. Hearing and vision have been screened and are reportedly normal. She is followed by ophthalmology closely for possible glaucoma and visual changes associated with Sturge-Weber Syndrome.

Medications:
baby aspirin (once daily); Onfi (clobazem) tablet (10mg/day); levetiracetam (5mL twice daily), folic acid (400 mcg/day); and vitamin D (400 IU/day). She took her medications as prescribed on the day of the evaluation.

Educational History
Homeschool cooperative and mother teaching. Mother experienced in homeschooling. Notes strength with memorization.

Social-Emotional History
Has some friends, not a best friend. Since starting levetiracetam, parents described that Jane had increased difficulty with behavioral outbursts, though this has reportedly decreased since her dosage has been changed. Emotionally, Jane's mother reported no concerns with symptoms of depression or anxiety and indicated that Jane "loves herself" and has a generally sweet disposition. Jane has many important strengths and is described as having a good sense of humor and she enjoys music, dancing, helping her mother in the kitchen and playing outside.