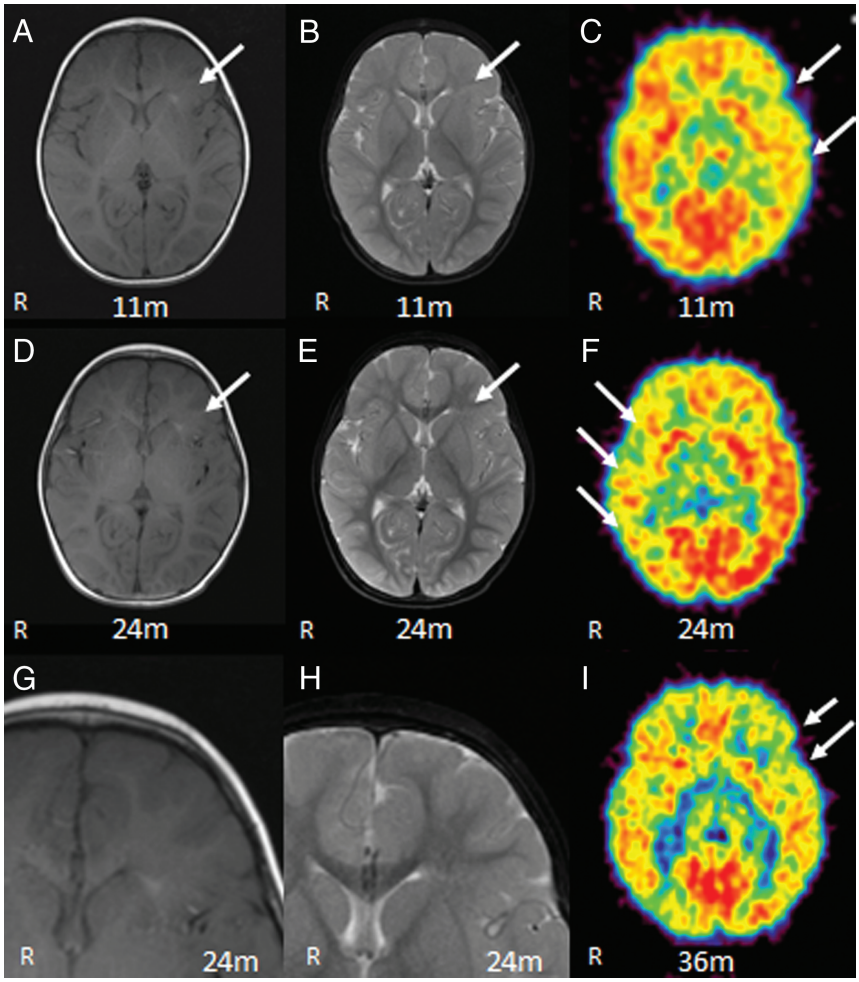
On-line Table 1: Patient characteristics

Patient No.	Sex	Age at Onset of Spasms (mo)	Age at Diagnosis of WS (mo)	Age at Starting ACTH Therapy (mo)	Treatments	Seizure Outcome after Initial Treatment	Seizure Outcome at Last Follow-Up	DQ or Intelligence Quotient at Last Follow-Up
1	Female	7	11	12	Clobazam, ACTH, valproic acid, zonisamide, topiramate, levetiracetam	Spasm recurred at 17 mo of age	Seizure-free with AEDs (topiramate, levetiracetam) (5 yr of age)	57 (KIDS, 5 yr of age)
2	Female	18	19	19	Valproic acid, ACTH	Seizure-free from 20 mo of age	Seizure-free without AEDs, except febrile seizures (5 yr of age)	95 (KIDS, 5 yr of age)
3	Male	2	4	4	Clonazepam, ACTH	Seizure-free from 5 mo of age	Seizure-free without AEDs (7 yr of age)	84 (Wechsler Intelligence Scale for Children, 4th ed, 7 yr of age)
4	Female	3	4	5	Clonazepam, ACTH	Seizure-free from 5 mo of age	Seizure-free without AEDs (5 yr of age)	91 (KIDS, 5 yr of age)

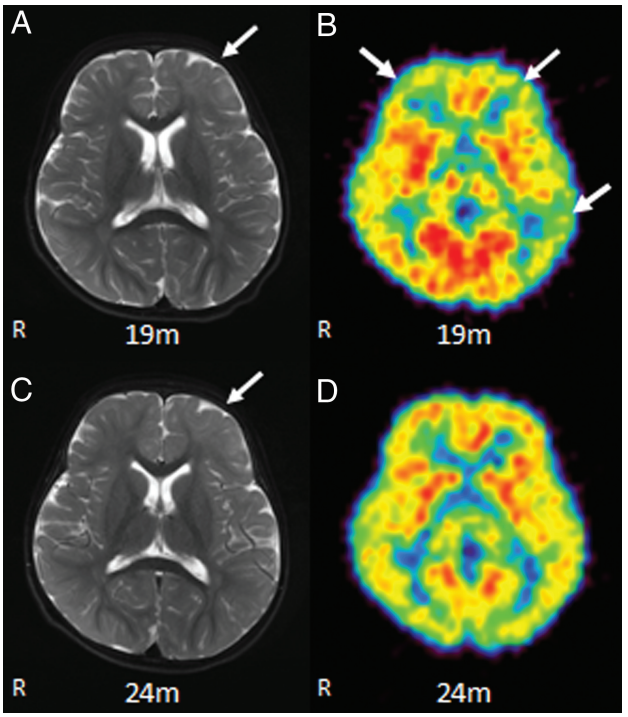
Note:—ACTH indicates adrenocorticotropic hormone; AEDs, antiepileptic drugs; KIDS, Kinder Infant Development Scale; DQ, developmental quotient.

On-line Table 2: Summary of MRI, PET, and EEG findings

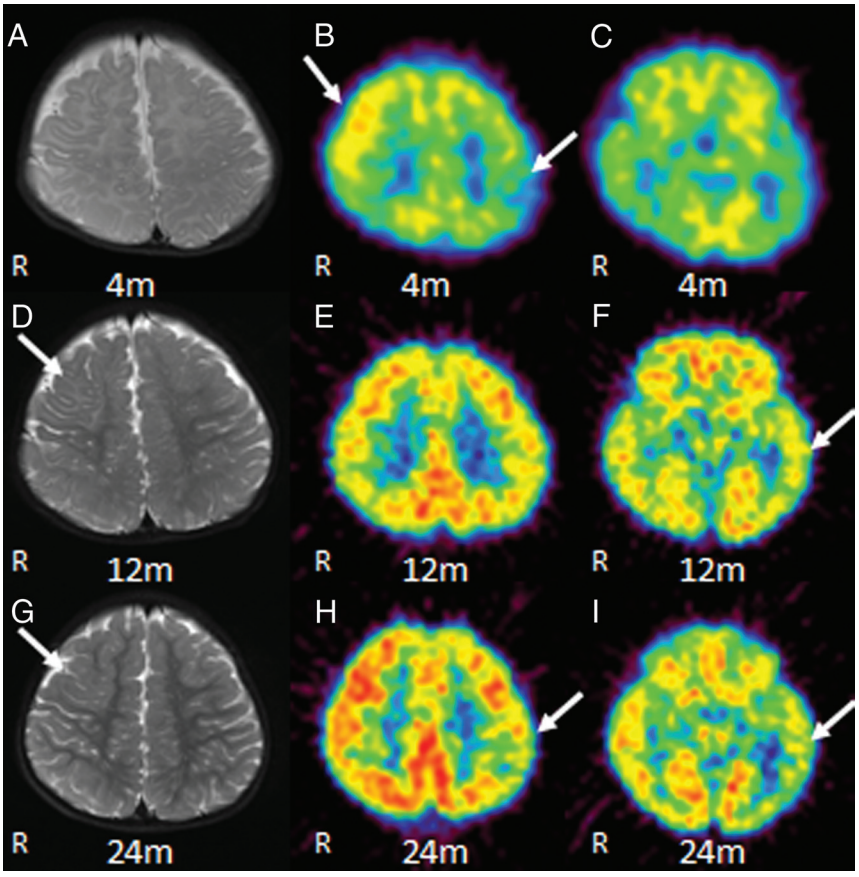
Patient No.	Age at Examination (mo)	MRI Lesion	PET		EEG	
			At Lesion	Extralesion	At Lesion	Extralesion
1	11	Left frontal	Hypometabolism	Hypometabolism left temporal	Hypsarrhythmia, spikes, spike and waves	Hypsarrhythmia
	24	Left frontal	Normal	Hypometabolism right frontotemporal, hypermetabolism left temporal	Spikes, polyspikes, slow waves	None
	36	Left frontal	Hypometabolism	Normal	Spikes, polyspikes, slow waves	None
2	19	Left frontal	Hypometabolism	Hypometabolism left temporal, right frontal	Hypsarrhythmia	Hypsarrhythmia, spikes in left temporal
	24	Left frontal	Normal	Normal	Spikes, polyspikes, sharp waves	None
3	4	Invisible	Hypermetabolism	Hypometabolism left parietotemporal	Hypsarrhythmia	Hypsarrhythmia
	12	Right frontal	Normal	Hypometabolism left temporal	Spikes, fast waves	Spikes, fast waves in left frontal
	24	Right frontal	Normal	Hypometabolism left parietotemporal	None	Polyspikes in left temporal
4	4	Invisible	Normal	Normal	Hypsarrhythmia	Hypsarrhythmia
	12	Invisible	Normal	Normal	Spikes, sharp waves	None
	24	Right frontal	Hypometabolism	Hypometabolism left temporal	Sharp waves	Sharp waves in left temporal



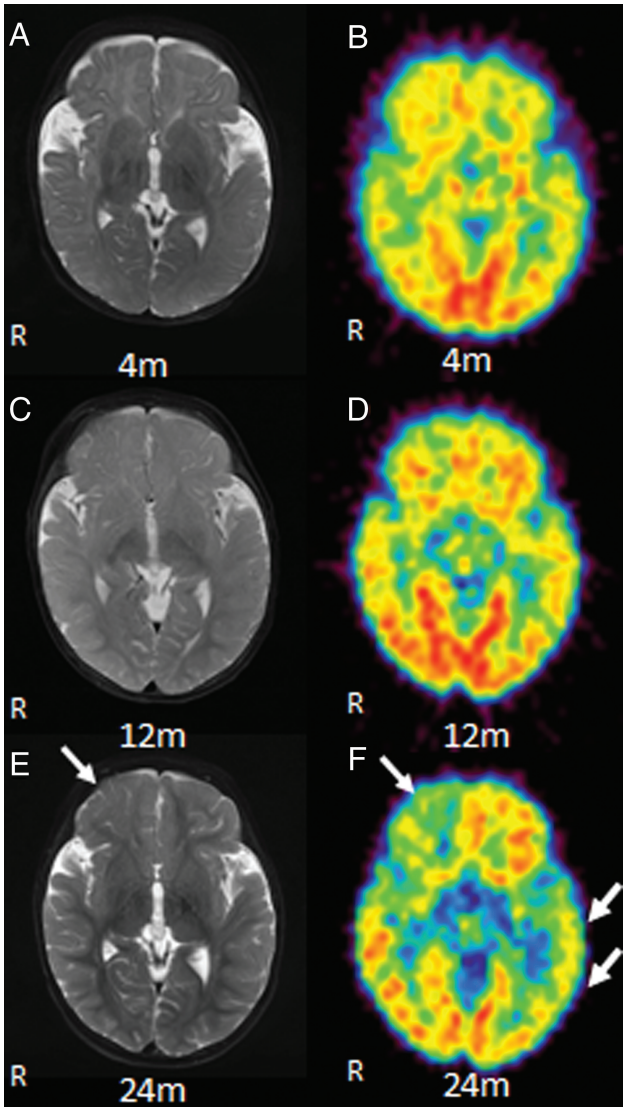
ON-LINE FIG 1. MR imaging and PET of patient 1. *A*, T1WI at 11 months of age shows a hyperintense area in the subcortical white matter of the left frontal lobe (arrow). *B*, T2WI shows a hyperintense area at the same site as the T1WI (arrow). *C*, PET at 11 months of age shows hypometabolism in the left frontotemporal lobes (arrows). *D–E* and *G–H*, MR imaging at 24 months of age still shows hyperintensity in the subcortical white matter of the left frontal lobe (arrow). *F*, PET at 24 months of age shows hypometabolism in the right frontotemporal lobe contralateral to the MR imaging lesion (arrows). *I*, PET at 36 months of age shows regional hypometabolism at the site of the MR imaging lesion in the left frontal lobe (arrows).



ON-LINE FIG 2. MR imaging and PET of patient 2. *A*, T2WI at 19 months of age shows a hyperintense area in the subcortical white matter of the left frontal lobe (arrow). *B*, PET at 19 months of age shows multiple regions of hypometabolism in the bilateral frontal and left temporal lobes (arrows). *C*, MR imaging at 24 months of age shows hyperintensity in the left frontal white matter that is the same as at 19 months of age (arrow). *D*, On PET at 24 months of age, hypometabolism is normalized.



ON-LINE FIG 3. MR imaging and PET of patient 3. *A*, MR imaging at 4 months of age does not show any abnormality. *B* and *C*, PET at 4 months of age shows regional right frontal hypermetabolism and left parietal hypometabolism (arrows). *D*, T2WI at 12 months of age shows a hyperintense area in the subcortical white matter of the right frontal lobe (arrow). *E* and *F*, PET at 12 months of age shows regional hypometabolism in the left temporal lobe (arrow). *G*, On T2WI at 24 months of age, a hyperintense area is more clearly seen than at 12 months of age (arrow). *H* and *I*, PET at 24 months of age shows left temporoparietal hypometabolism (arrow).



ON-LINE FIG 4. MR imaging and PET of patient 4. *A* and *B*, MR imaging and PET at 4 months of age do not show an abnormality. *C* and *D*, MR imaging and PET findings at 12 months of age are normal. *E*, T2WI at 24 months of age shows a hyperintense area in the white matter of the right orbitofrontal area (arrow). *F*, PET at 24 months of age shows regional hypometabolism in the right frontal and left temporal lobes (arrows).