Table 2. Epilepsy history of the 9 ARX patients with epilepsy.

Patient	Type of seizure	Age at onset	Frequency and circumstances of seizures	EEG	medications	Outcome of epilepsy	Severity of cognitive deficit
1 (Family I)	Infantile spasms	4 months	daily	hypsarythmia	GVG, HC Topiramate	At 20 months: free of seizure and meds	Moderate
2 (Family III)	Partial complex	4 years and 4 months	Total of 8 seizures (1 watching TV)	Occipitotemporal spikes & slow waves, generalized spike wave discharges	VPA	At 17 years and 9 months: free of seizures on VPA	Moderate
3 (Family V)	- Infantile spasms - Febrile status epilepticus - generalized tonic clonic seizure - partial seizure	- 5 months - 14 years and 9 months - 17 years and 3 months - 17 years and 7 months	- daily - Rare generalized tonic clonic seizures	hypsarythmia	ACTH, CZP VPA	At 18 years and 8 months: last seizure at 17 years 7 months old, on VPA CZP	profound
4 (Family VII)	Short febrile tonic clonic (once without fever)	3 years and 4 months	Total of 3 seizures within six months	normal	VPA	At 4 years and 6 months: free of seizures on VPA	Moderate
5 (Family X)	- Infantile spasms - partial complex seizures - noctural tonic- clonic - generalized tonic- clonic seizures - status epilepticus	- 7 months - 22 months - 3 years and 2 months - 5 years and 8 months - 6 years and 1 month	- Daily - Twice a week - Once a week - every two months	Na	- HC ACTH - CBZ - GVG - VPA - LTG LVT	At 11 years and 4 months: generalized tonic clonic seizure once a month, on VPA, LTG, LVT	profound
6 (Family XI)	Absences	4 years	Some watching TV	Na	VPA	At 11 years and 10 months: free of seizures and meds	Moderate
7 (Family XI)	- Febrile seizure - Absences	- 14 months - 7 years and 9 months	watching TV	Na	no	At 7 years and 11 months: free of seizures and meds	Moderate
8 (Family XII)	?	14 years			Ethosuccimide VPA, CLB	At 54 years: occasionally	severe

9	- Hypotonic	- 22 months	- once	Normal			severe
(Family XII)	seizures			at 4 years old			
	- Tonic seizures	- 5 years	- 2 to 3 times a		- VPA CBZ	At 18 years and 6	
	with eye rolling,		week		CLB	months: free of	
	pallor and cyanosis					seizures on VPA	
						CBZ	

CBZ: carbamazepin; CZP: clonazepam; CLB: clobazam; LTG: lamotrigin; VPA: valproate; GVG: vigabatrin; LVT: levetiracetam; HC: hydrocortisone.