

Table e-1: Clinical, paraclinical and immunological features of patients with VGKC-complex Abs and primary non-inflammatory diagnoses

Demo-graphics	Presentation	Investigations	Diagnosis	VGKC-complex-Ab (pM)	Immunotherapy	Outcome (Follow-up, months)
9 yr M Caucasian	Focal epilepsy with worsening seizures.	MRI: Left hippocampal sclerosis CSF not done	Focal epilepsy	105	Steroids	Good response to steroids (11)
3 yr M Caucasian	Transient Hemiplegia	MRI brain normal	Undiagnosed	111	Not treated	Full recovery (25)
14 yr M Asian	Motor tics	Not investigated	Tic disorder	117	Not treated	Ongoing symptoms (23)
8 yr M Caucasian	Long standing seizure disorder	MRI brain normal CSF not done	Primary generalized epilepsy	120	Not treated	On-going seizures (13)
16 yr F Caucasian	Bilateral lower limb pain. Long standing Hypothyroidism	MRI brain and spine normal. NCS and EMG normal	Undiagnosed	125	Not treated	Ongoing symptoms (12)
10 yr M Caucasian	Headaches, history of allergies	MRI brain normal	Headaches	128	Not treated	Full recovery (24)

4 yr F African Caribbean	New onset myoclonic seizures	MRI normal CSF no cells	Myoclonic astatic epilepsy	143	Not treated	On-going seizures. Developmental concerns (26)
9 yr M Asian	Encephalopathy with worsening seizures. Background learning difficulties	MRI: Bilateral grey matter heterotopia. CSF no cells	Pyridoxine-dependent epilepsy (compound heterozygote mutation in ALDH7A1)	158	Not treated	On-going seizures (26)
14 m M Caucasian	Encephalopathy with prolonged seizures	MRI: Cortical swelling	Mitochondrial disorder with POLG mutation	394	Not treated	Patient died

Table e-2. Clinical, paraclinical and immunological features of patients with VGKC-complex Abs levels under 400pM and primary inflammatory diagnoses

	Demo-graphics	Presentation	Investigations	Diagnosis	VGKC-complex-Ab (pM) and additional antibodies	Immunotherapy	Outcome (Follow-up, months)
1	9yr F Asian	Encephalopathy, seizures	MRI: Increase leptomeningeal enhancement CSF: WCC 15	Autoimmune encephalopathy	103	Not treated	Cognitive Behavior Seizures (48)
2	16yr F Caucasian	Encephalopathy, seizures, hallucinations	MRI: Signal change both hippocampi and amygdala, swelling of mesial temporal structures. CSF no cells	Limbic encephalitis	107 TPO Abs 464 IU/mL	Steroids, IVIG	Behavioral problems (51)
3	13yr F Caucasian	Muscle weakness and leg cramps Background of scleroderma	Not done	Myositis.	112, hippocampal neurons positive	Escalation of IT	Myositis resolved (42)
4	2yr F Caucasian	Encephalopathy, seizures and movement disorder	MRI: Cortical restricted diffusion. CSF no cells	Autoimmune encephalopathy	113 TPO Abs 247 IU/ml	Steroid, PLEX, IVIG	Cognitive, behavior (20)

5	11yr F African Caribbean	Subacute onset weakness of both lower limbs with frequent falls	MRI: multiple high signal lesions in the brain and spine CSF: no cells, intrathecal OCB	Clinically isolated syndrome	126	Steroids	Clinical relapse (relapsing remitting multiple sclerosis) (26)
6	6yr M Asian	Ascending paralysis ophthalmoplegia and ataxia	MRI brain: Normal MRI spine: Diffuse thickening and enhancement of ventral and dorsal roots (cauda equina)	GBS Miller Fisher variant	130	Steroids	Full recovery (14)
7	13yr M African Caribbean	Encephalopathy, aphasia, hallucination, parkinsonism	MRI brain: Normal. PET reduce uptake in the cortex CSF no cells	Autoimmune encephalopathy	136 ASOT 400 IU, TPO Abs 235 IU/ml	Steroids IVIG	Full recovery (15)
8	10yr F African Caribbean	Encephalopathy, seizures, movement disorder	MRI brain: Widespread cortical signal change CSF no cells	NMDAR-Ab encephalitis	138, hippocampal neurons positive NMDAR abs	IVIG	Quadraparesis Visual failure Seizures (60)
9	15yr F African Caribbean	Leg weakness and sensory loss	Multiple shot segment myelopathy	Transverse myelitis	139	Steroids	Prolonged recovery course (27)

10	13yr F Asian	Acute confusional state with strange behavior and hallucination and bradykinesia.	MRI brain: Normal CSF no cells	Autoimmune encephalopathy	140 ASOT 400 IU	Steroids	Relapse at 8months with good response to steroid (29)
11	3yr F Caucasian	Ataxia, urine incontinent, aphasia, seizure, hallucinations.	Reversible cerebellar signal change. CSF no cells, mixed OCB	Autoimmune encephalopathy	149	Steroids	Emotional liability, Behavioral problems (22)
12	2yr F Caucasian	Encephalopathy, confusion, seizures	MRI brain: Bilateral hippocampi and amygdala, swelling of mesial temporal structures. CSF no cells	Limbic encephalitis	156	Steroids	Full recovery (9)
13	4yr M Caucasian	Encephalopathy, seizures, hallucinations	MRI brain: Normal CSF no cells	Autoimmune encephalopathy	161	Steroids, IVIG, Azathioprine	On-going seizures. Cognitive (60)
14	9yr F Caucasian	Confusion, seizures, nystagmus, dysmetria and ataxia	MRI brain: Normal CSF no cells	Autoimmune encephalopathy	163	Not treated	Full recovery (23)
15	15yr F Caucasian	Relapsing chorea despite treatment with Pen V and valproate Background of SLE	MRI brain; Normal CSF no cells	Sydenham chorea.	175	MMF for SLE	On going (12)
16	16yr F	Weakness, unsteadiness and	EMG and nerve conduction study	Lambert–Eaton	175	Not treated	On going

	Caucasian	double vision	indicative of presynaptic neuromuscular junction defect	myasthenic syndrome			(63)
17	12yr F Caucasian	Encephalopathy, confusional state and seizures	MRI brain: Focal temporal cortical signal change CSF WCC 20	Limbic encephalitis	185	Not treated	Ongoing seizures and memory prob (60)
18	2yr M African Caribbean	Encephalopathy with disautonomia	MRI: bilateral signal change in the medulla and corticospinal tracts CSF no cells	Acute disseminated encephalomyelitis	265	Steroids	Full recovery (29)