28th Annual meeting of Epilepsy Society of Thailand

Autoimmune-associated epilepsy & Seizures secondary to autoimmune encephalitis

Two sides of the same coin

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Disclosures

- Not directly related to this talk

Overview

01 Key facts about autoimmune-associated seizures

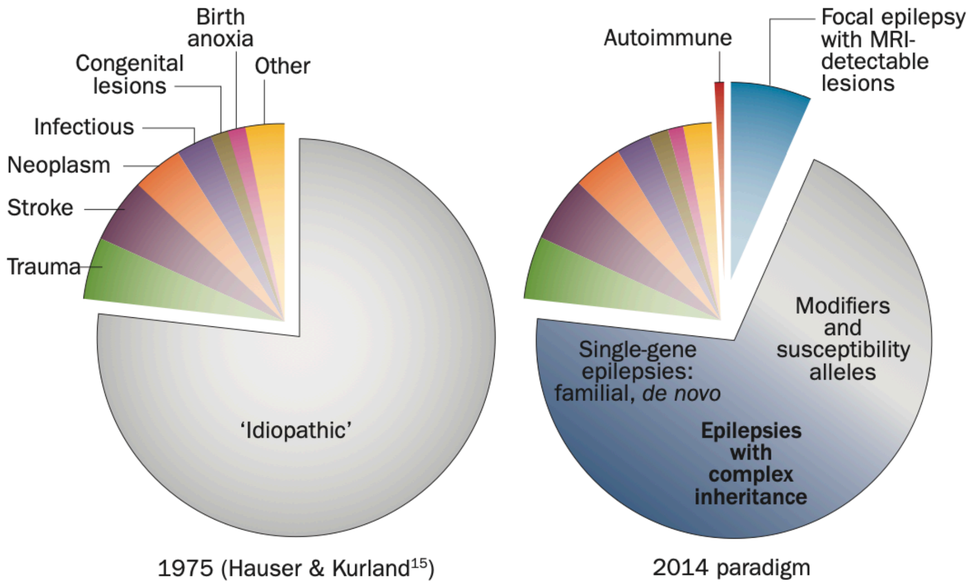
02 Acute symptomatic seizure & autoimmune-associated epilepsy

03 Patient selection for immune etiology evaluation

04 Treatment approach of autoimmune-associated seizures

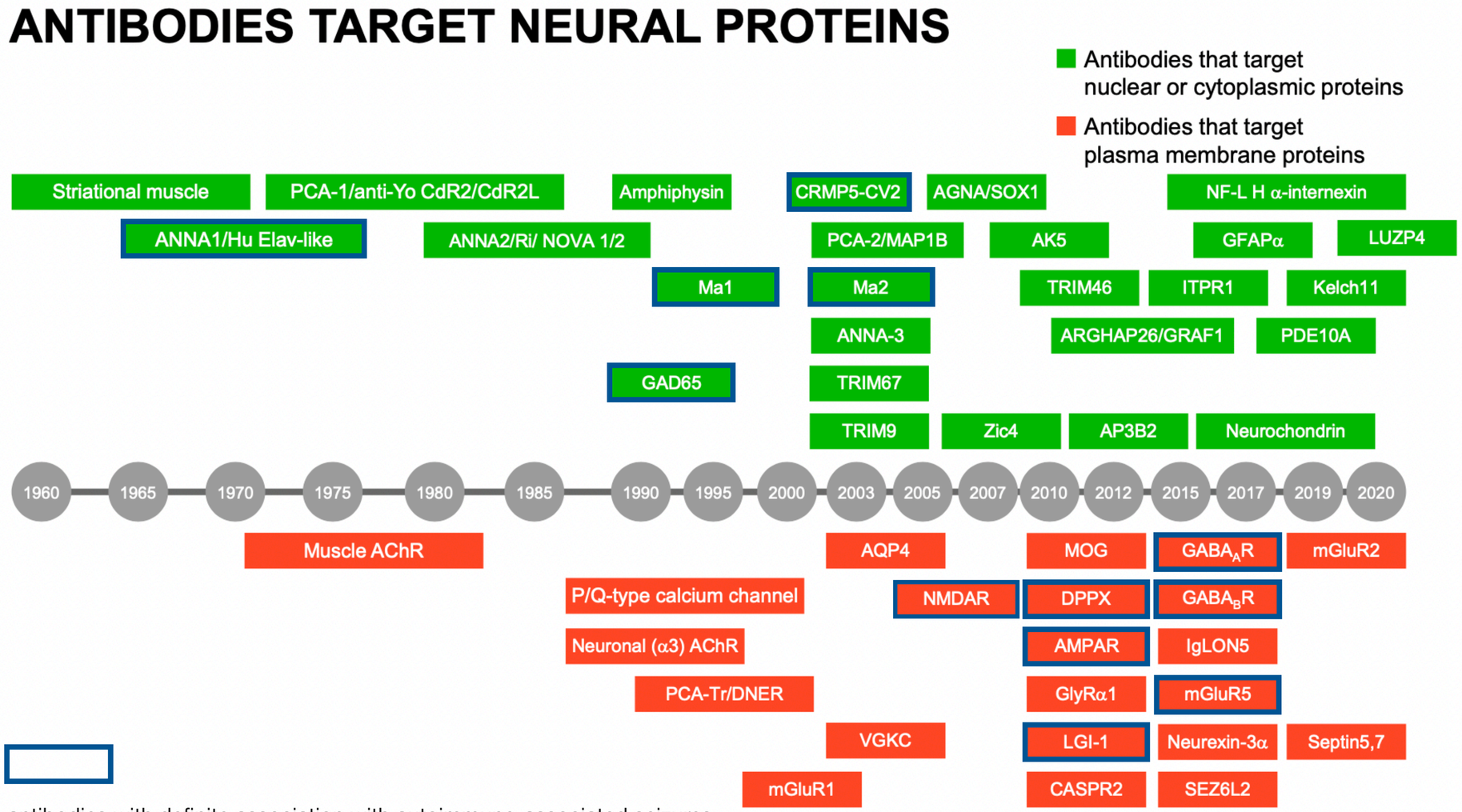
Key facts

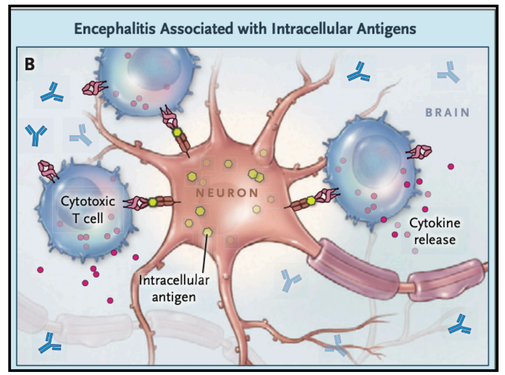
- A third of patients suffer ongoing seizures and mechanisms are incompletely understood.
- Neuroinflammation likely involved in many epilepsies, regardless of etiology.
- Seizures commonly occur in autoimmune encephalitis, initial presentation 20%.
- Focal epilepsy of unknown cause, 3-16%

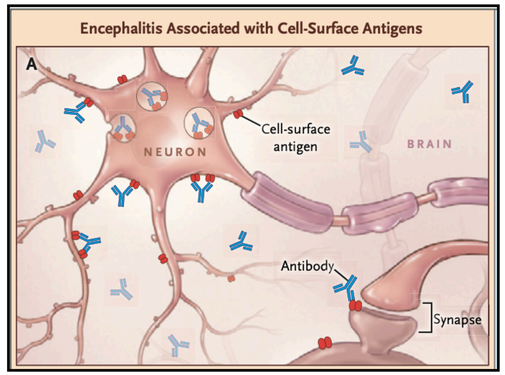


Thomas & Berkovic. *Nat Rev Neurol* 2014; Feyissa et al. *Neurol Neuroimmunol Neuroinflamm* 2017; Steriade et al. *JAMA Neurol* 2021

Increase awareness of autoimmune causes of seizures







courtesy Divyanshu Dubey. summer conference 2022

Acute symptomatic seizure & Epilepsy

"**Epileptic seizure**: occurrence of signs and/or symptoms due to *abnormal excessive or synchronous neuronal activity* in the brain."

"may be characterised by *sensory, motor or autonomic phenomena* **with** or **without** the loss of consciousness."

–ILAE 2005

"**Epilepsy**: a disorder of the brain characterised by an enduring predisposition to generate epileptic seizure, and by neurobiologic, cognitive, psychological, and social consequences."

–ILAE 2005

"**Acute symptomatic seizure**: seizure occurring at the time of systemic insult or in close temporal ass. with a documented brain insult."

–ILAE 2010

Seizure, from Latin sacire, "to take possession of"

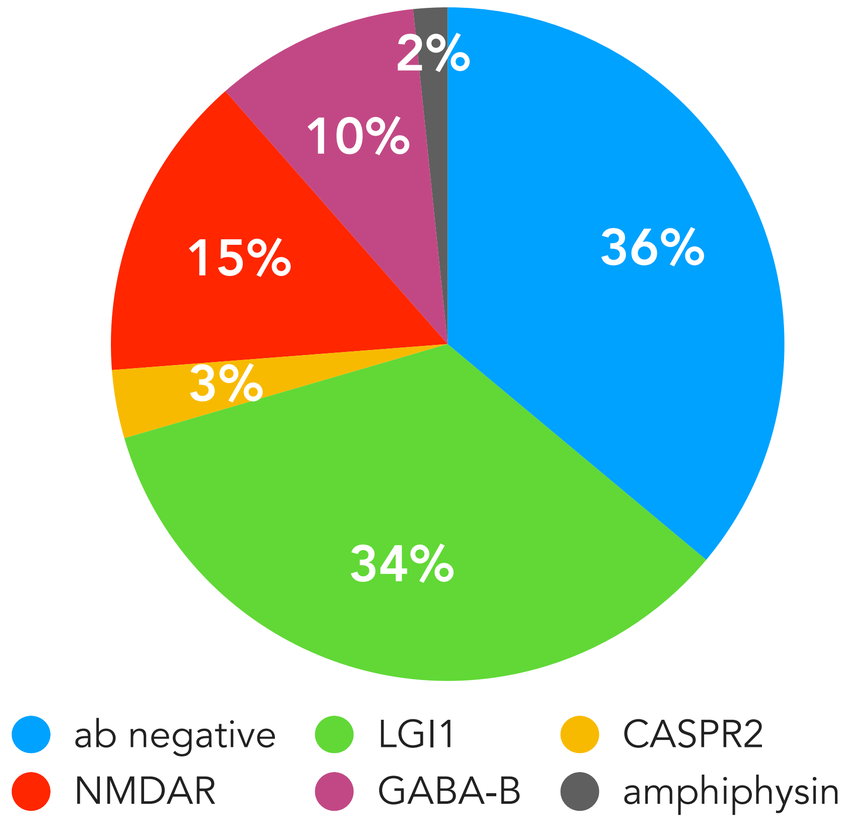
Acute symptomatic seizures secondary to autoimmune encephalitis and autoimmune-associated epilepsy: Conceptual definitions

	Acute symptomaic seizures secondary to autoimmune encephalitis	Autoimmune-associated epilepsy
Pathophysiology	seizures during active phase of encephalitis antibody-mediated ictogenesis	structural postencephalitic injury and/or ongoing T-cell-mediated inflammation
Underlying antibodies	Antibodies against surface antigens (LGI1, NMDAR, CASPR2, GABAR, GABABR, mGluR5, DPPX, AMPAR)	Antibodies against intracellular antigens (GAD-65, onconeural ab) Rasmussen encephalitis Persistent epilepsy after acute AE
Outcome	Seizures terminate with remission of encephalitis Potential for ASM discontinuation	Pharmacoresistant focal epilepsy common

modified from Steriade et al. *Epilepsia* 2020

Seizure semiology

Observational retrospective case series
A tertiary epilepsy center
Autoimmune-associated seizures [ab+ (n 39), ab- (n 22)] vs MTLE with HS (n 22)
Median age 50 years, 82% male



	Positive ab	MTLE-HS
Focal aware seizure	23.3%	4.5%
Focal unaware seizure	50%	100%
Postictal confusion	0	82%
Focal to bilateral tonic-clonic	77% (78% during sleep)	36% (13% during sleep)
Sz duration (sec)	11	95
Seizure frequency	93% daily no monthly	60% weekly 40% monthly

Regarding seizure semiology
: NO significant differences were noted between ab+ and ab-

Lv et al. *Ann Clin Transl Neurol* 2018

Seizure semiology

Observational retrospective case series

Mellen Center at the Cleveland Clinic

Autoimmune encephalitis with seizures

Median age 58 years (14-83), 63% female

Ab+ 74%

	Total cohort (n = 19)
Seizure semiology	
Subclinical (%)	11 (58%)
Generalized tonic-clonic (%)	11 (58%)
Focal unaware (%)	9 (47%)
Focal aware motor (other than faciobrachial dystonic) (%)	4 (21%)
Faciobrachial dystonic[a] (%)	4 (21%)
Focal aware nonmotor (%)	11 (58%)
Somatosensory (%)	5 (26%)
Visual (%)	3 (16%)
Gustatory (%)	2 (11%)
Olfactory (%)	1 (6%)
Pilomotor (%)	2 (11%)
Psychic (%)	8 (42%)
Abdominal (%)	3 (16%)
Autonomic (%)	5 (26%)
Multimodal auras[b] (%) (n = 11 – patients with auras)	9 (82%)
Median number of aura types (range, SD)	3 (1-4, 1.1)
Status epilepticus upon presentation (%)	10 (53%)
Refractory status epilepticus (NORSE syndrome) (%)	9 (47%)

Where is the symptomatogenic zone?

- Rapidly refractory of ASMs
- Median time to 2nd ASM of 9.5 days, median number of ASM of 3 (1-6)

Steriade et al. *Seizure* 2018

Perisylvian regions are susceptible to immune-mediated epileptogenesis

Refractory chronic epilepsy associated with neuronal auto-antibodies: could perisylvian semiology be a clue?

Lisa Gillinder [1,2], Linda Tjoa [1,2], Basil Mantzioris [1,2], Stefan Blum [1,2], Sasha Dionisio [1,2]

[1] Mater Advanced Epilepsy Unit, Mater Adults Hospital,
[2] Princess Alexandra Hospital, Department of Neurology, Brisbane, Australia

"Series of 10 patients with autoimmune-associated epilepsy all has perisylvian semiology"

Table 1. Summary of the semiological features of perisylvian seizures, which are largely classified into five groups.

SOMATOSENSORY*	VISCERO-SENSITIVE	AUDITORY	LANGUAGE	AUTONOMIC
Parasthesias	Pharyngeal discomfort	Auditory hallucinations	Dysarthria	Tachycardia
Pain	Abdominal discomfort**	Palinacousis	Dysphasia	Bradycardia
Warmth	Thoracic sensations	Tinnitus		Hyperventilation
	Taste	Vertigo		Hypoventilation
	Tenesmus			Piloerection
				Flushing
				Sweating
				Salivation

Retrospective review of case series

Outpatient department of a tertiary centre

Chronic epilepsy (>2 years) + antibody positive

N 10, mean age 37 y (22-43), 80% female

50% GAD65, 30% GLY-R, 10% NMDAR,

10% LGI1

Non-lesional epilepsy with perisylvian semiology

- high level of suspicion for autoimmune etiologies

Gillinder et al. *Epileptic Disord* 2017

New-onset refractory status epilepticus

Etiology, clinical features, and outcome

Retrospective review of RSE without etiology identified within 48 h
13 academic medical centers

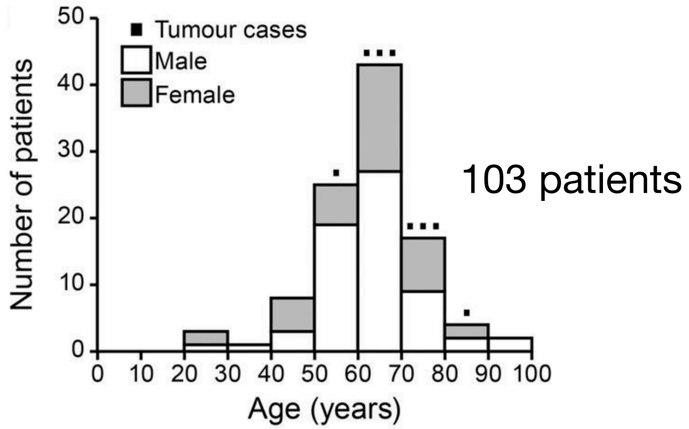
- 48% etiologies were identified
- 19% autoimmune encephalitis
- 18% paraneoplastic encephalitis (tumours should be removed)

Table 1 Eventual etiology of new-onset refractory status epilepticus after extensive evaluation

Etiology	No. (%)
Cryptogenic	67 (52)
Nonparaneoplastic autoimmune	25 (19)
Anti-NMDA receptor	7 (5)
Anti-VGKC complex	5 (4)
SREAT	5 (4)
Cerebral lupus	4 (3)
Anti-GAD65	3 (2)
Anti-striational	1 (1)
Paraneoplastic	23 (18)
Anti-NMDA receptor	9 (7)
Anti-VGKC complex	3 (2)
Anti-Hu	3 (2)
Anti-VGCC	2 (2)
Anti-CRMP5	1 (1)
Anti-Ro	1 (1)
Seronegative	4 (3)
Infection-related	10 (8)

Gaspard et al. *Neurology* 2015

LGI1-antibody limbic encephalitis



- age: 64 (31-84)
- ♂ : ♀ = 2 : 1
- Amnesia, disorientation, emotionality
- Hyponatremia, sleep disturbance,
- rare cancer association: thymoma, lung, adenoCA

Criteria for AE	
Subacute onset < 3 months	**40%**
Focal CNS finding	10%
Seizures	**99%**
CSF pleocytosis	20%
MRI suggestive of encephalitis	40-75%

Thompson et al. *Braine* 2018; Gadoth et al. *Ann Neurol* 2018; Rocamora et al. *Seizure* 2017

Seizures in LGI1-antibody limbic encephalitis

- Seizures are frequent: multiple per day
- Subclinical seizures, motor/sensory seizures “wave” sensation
- **Faciobrachial dystonic seizures (FBDS) are highly specific, present in 34-53%**
- Ipsilateral piloerection
- Bilateral TCS can occur, usually infrequent
- Semiology can change throughout course, multifocal localization described

Aurangzeb et al. *Seizure* 2017; Smith et al. *Neurology* 2021

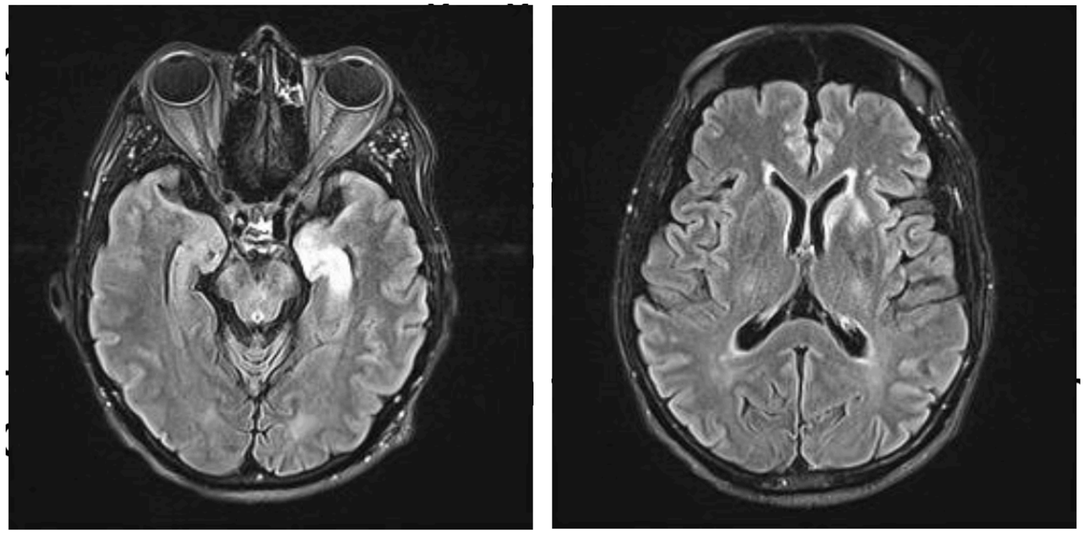
EEG in LGI1-antibody limbic encephalitis

- May be completely normal
- FBDS typically have no EEG correlate
- Subclinical seizures are frequently seen
- Interictal epileptiform discharges may be multifocal, frequently involve temporal regions
- *Seizures with hyperventilation* (all six patients were dx with autoimmune encephalitis)

Wennberg et al. *Clinical Neurophysiol* 2018; Wennberg RA. *Ann Clin Transl Neurol* 2022

Investigations & Outcomes

- MRI brain can be normal, signal abnormalities in medial temporal structures or basal ganglia



- CSF, can be normal (50%), antibody more sensitive in serum than CSF
- Most patients respond to immunotherapy, may have residual cognitive impairment
- Relapses are not uncommon (40%)(3-94 months)

Gadoth et al. *Ann Neurol* 2017; Smith et al. *Neurology* 2021

Seizure characteristics, treatment, and outcome in autoimmune synaptic encephalitis: A long-term study

Wuqiong Zhang, Xue Wang, Na Shao, Rui Ma, Hongmei Meng *

Department of Neurology, The First Hospital of Jilin University, Changchun, Jilin 130021, China

To report potential factors associated with persistent seizures

Patient with autoimmune synaptic encephalitis

First Hospital of Jilin Univerisity, 2015-2017

Seizure outcomes, median follow-up 30 mo (8-40)

63% exhibited seizure remission after immunoRx

Risk of developing epilepsy, **LGI1 & $GABA_B$ >>> NMDAR**

Factors:

- older age at onset
- status epilepticus
- high protein in CSF
- antibody type

ImmunoRx delay associated with development of epilepsy[2]

Statistical analysis of risk factors for patients who developed persistent seizures after initial immunotherapy.

	Short-term seizures (n = 27)	Persistent seizures (n = 16)	P-value
Age (years)	41 (18–73)	58 (38–68)	<0.001
Sex (female)	11 (40.7)	8 (50.0)	0.555
Interval before starting initial treatment (days)	31 (7–160)	48 (8–180)	0.308
Seizure as initial symptom	18 (66.7)	13 (81.3)	0.303
Absence of α-dominated rhythm	3 (11.1)	3 (18.8)	0.655
IEDs	12 (44.4)	10 (62.5)	0.252
SE	4 (14.8)	9 (56.3)	0.004
Abnormal MRI findings	14 (51.9)	10 (62.5)	0.497
CSF tests			
Hing protein level	7 (25.9)	9 (56.3)	0.047
Leukocytosis	17 (63.0)	9 (56.3)	0.663
Therapy selection			0.138
IVIG alone	2 (7.4)	4 (25.0)	
Corticosteroids alone	6 (22.2)	1 (6.3)	
Combination	19 (70.4)	11 (68.8)	
Neuronal antibody type			0.001
NMDAR	13 (48.1)	0	
LGI1	8 (29.6)	7 (43.8)	**LGI1 : 47%**
$GABA_BR$	6 (22.2)	9 (56.3)	**$GABA_BR$: 64%**

1. Zhang et al. *Epilepsy Behav* 2019; 2. Shen et al. *Ann Clin Transl Neurol* 2020

Autoimmune-associated epilepsy remain a core diagnostic challenge

- Can occur in several context:

  1. after resolution of active encephalitis: up to 29%[1] (wide variability)

  2. chronic unresolving encephalitis

  3. chronic epilepsy (often drug resistance) and not carry typical encephalitic features

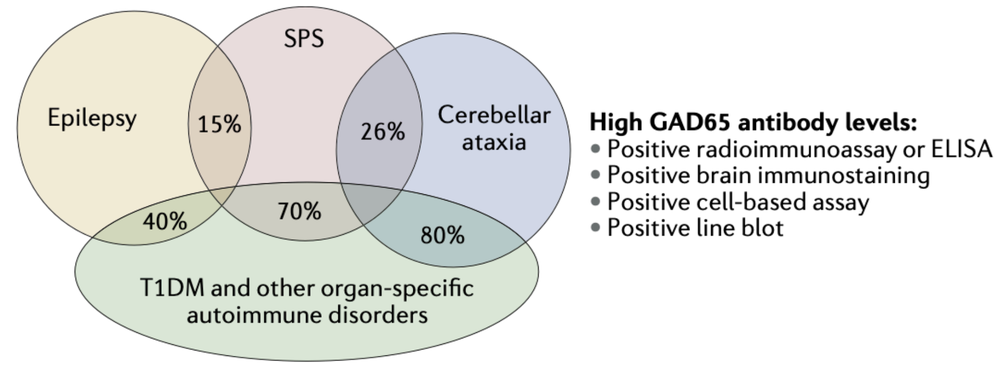
- Systemic review of studies reporting % neural autoantibody in people with epilepsy (PWE)

  - antibody frequencies varied significantly across studies

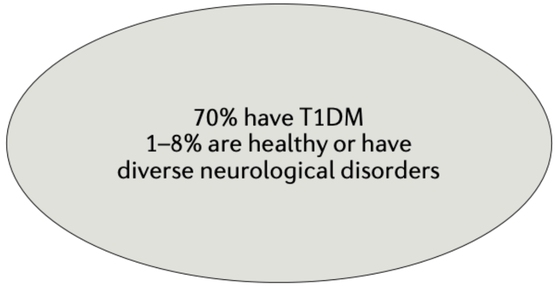
  - 0-24.1% antibody positivity; 0-12.5% GAD65 antibody positivity

1. Falip et al. *Seizure* 2020; 2. Steriade et al. *JAMA Neurol* 2021

Anti-GAD65 antibody-associated epilepsy



High GAD65 antibody levels:
- Positive radioimmunoassay or ELISA
- Positive brain immunostaining
- Positive cell-based assay
- Positive line blot



Low GAD65 antibody levels:
- Positive radioimmunoassay or ELISA
- Negative brain immunostaining
- Negative cell-based assay
- Positive or negative line blot

RIA titer > 20 nmol/L
ELISA > 1000 U/mL

- **Young adults (median 30yr), ♀ : ♂ = 4:1**
- **Autoimmune disorder** (57%): T1DM, thyroid disease
- Frequent seizure
- Focal aware seizure are common
- **TLE "plus"**
- Sometimes encephalitic onset
- MRI: vary over the course
- CSF: typically noninflammatory, OCB
- *<20% become seizure-free*

Graus et al. *Nat Rev Neurol* 2020

Epilepsia

BRIEF COMMUNICATION

Musicogenic epilepsy: Expanding the spectrum of glutamic acid decarboxylase 65 neurological autoimmunity

Kelsey M. Smith[1] | Nicholas L. Zalewski[2] | Adrian Budhram[1,3] | Jeffrey W. Britton[1] | Elson So[1] | Gregory D. Cascino[1] | Anthony L. Ritaccio[4] | Andrew McKeon[1,5] | Sean J. Pittock[1,5] | Divyanshu Dubey[1,5]

Retrospective chart review

9 musicogenic epilepsy patients

All had high-titer GAD65-IgG

(median 294 nmol/L)

#	handedness	Age at epilepsy onset, years	Cognitive concerns	Type of music	Highest seizure frequency	Main seizure type	Bilateral tonic–clonic ever	Seizures induced by music on EEG	Any seizures captured on EEG	Interictal discharges on EEG	Brain MRI
1	M/25/R	18	No	Pop	1/day	FIA	Yes	RT	RT	BT	Normal
2	F/36/L	36	No	Country	10–11/day	FA	No	None	LT	BT	Normal
3	F/31/R	17	Yes	Pop	1/week	FIA	Yes	None	RT	RT	Normal
4	F/28/L	23	No	No specific music	5–6/month	FA	Yes	None	RT	RT	Normal
5	F/61/R	46	No	Melancholic, church hymns	1/week	FIA	Yes	RT	RT	BT	Normal
6	F/26/R	23	No	Pop and soft rock	4–5/week	FIA	No	LT	LT	None	Normal
7	F/34/R	4	Yes	Unfamiliar hymns, classical	4/day	FIA	Yes	None	BT	BT	Normal
8	F/36/R	34	Yes	Pop and techno	Once daily	FIA	No	RT	RT	None	Cystic lesions RT
9	F/52/R	14	Yes	Organ music	1/2 weeks	FIA	Yes	None	BT	BT	R MTS

GAD65-IgG should be tested in patients with musicogenic epilepsy

Smith et al. *Epilepsia* 2021

Clinical characteristics that suggest underlying autoimmune origin

1. High seizure frequency (daily)
2. Short seizure duration (<30 sec)
3. Changing seizure type over time
4. Early intractibility to high number of ASM
5. Preceding febrile illness, history of systemic autoimmunity or malignancy
6. Perisylvian semiology, FBDS, NORSE +unidentified cause, musicogenic seizures

No absolute pathognomonic clinical feature of autoimmune-associated seizures

Lv et al. *Ann Clin Transl Neurol* 2018, Quek et al. 2012, Steriade et al. *Seizure* 2018, Aurangzeb et al. 2017, Irani et al. 2011, Thompson et al. 2018, Falip et al. 2018

Precautions about antibody test result

Antibodies with uncertain association with encephalitis

- Low-titer GAD65 (<20 nmol/L)
- Voltage-gated potassium channel (without reactivity to LGI1 or CASPR2)
- Low-titer CASPR2
- Ganglionic nicotinic acetylcholine receptor
- VGCC

Careful to always correlate clinical to antibody detected

***Panel 7*: Criteria for autoantibody-negative but probable autoimmune encephalitis**

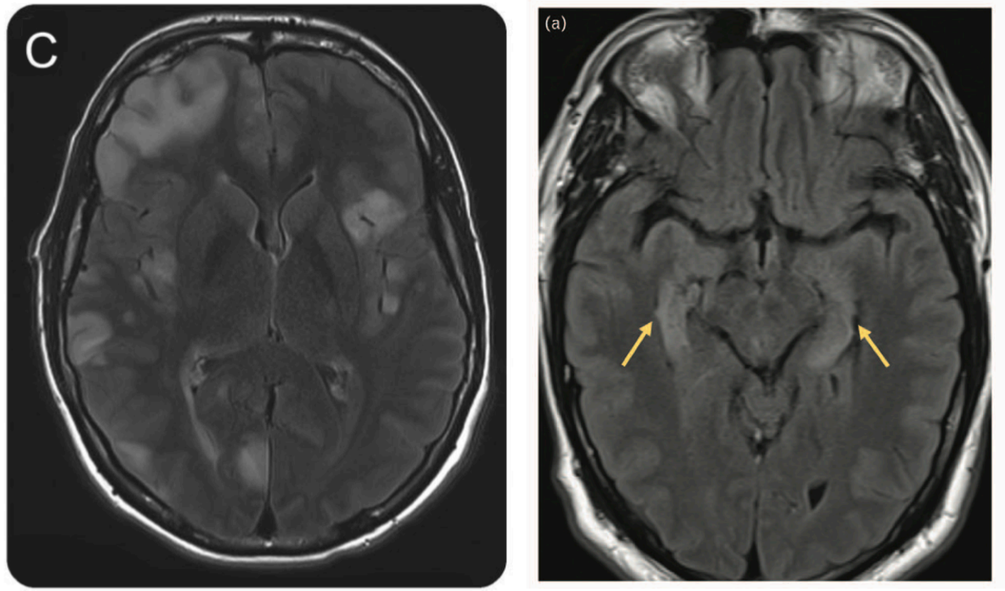
Diagnosis can be made when all four of the following criteria have been met:

1. Rapid progression (less than 3 months) of working memory deficits (short-term memory loss), altered mental status, or psychiatric symptoms
2. Exclusion of well defined syndromes of autoimmune encephalitis (eg, typical limbic encephalitis, Bickerstaff's brainstem encephalitis, acute disseminated encephalomyelitis)
3. Absence of well characterised autoantibodies in serum and CSF, and at least two of the following criteria:
   - MRI abnormalities suggestive of autoimmune encephalitis*
   - CSF pleocytosis, CSF-specific oligoclonal bands or elevated CSF IgG index, or both*
   - Brain biopsy showing inflammatory infiltrates and excluding other disorders (eg, tumour)
4. Reasonable exclusion of alternative causes

Graus et al. *Lancet Neurol* 2016

MRI brain

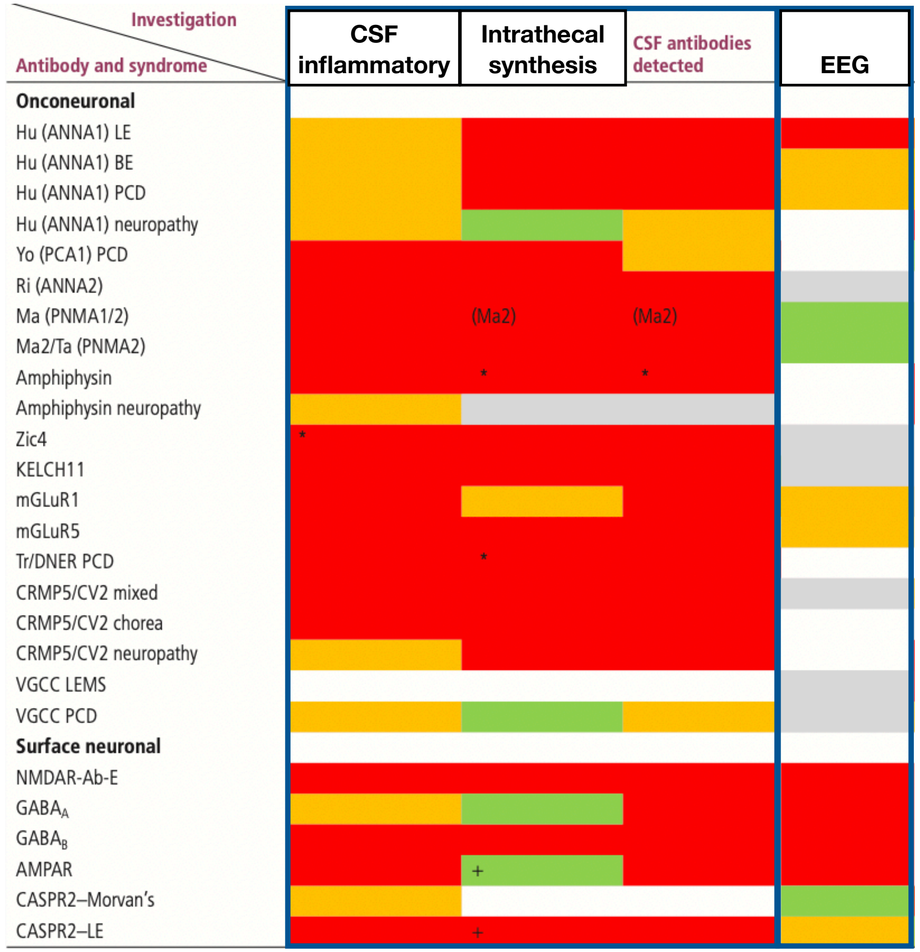
- Often abnormal in anti-$GABA_A$ receptor encephalitis: multifocal, affecting both gray and white matter (ddx of fulminant MS and ADEM)
- Abnormal in 60-70% of limbic encephalitis
- Normal in many cases of seronegative AE



Normal MRI does not exclude the diagnosis of autoimmune-associated seizure

Spatola et al. *Neurology* 2017; Venkatesan A. *Curr Opin Inf Dis* 2019

CSF & EEG evaluation



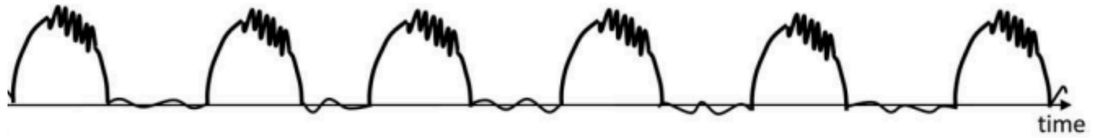
	RDA+F; or PDs+F if (and only if) the PDs are blunt delta waves	
	Continuous/ Abundant (≥50% of record/epoch)	**Frequent/ Occasional (≥1 to 49% of record/epoch)**
Fast activity WITH stereotyped relationship to delta wave	Definite EDB	Possible EDB
Fast activity WITHOUT stereotyped relationship to delta wave	Possible EDB	RDA+F or PDs+F, but NOT EDB

<u>30% of patients with anti-NMDAR encephalitis</u>[2]

more prolonged hospitalization and trend toward worse scores of mRS[2]

NOT unique to anti-NMDAR - ICU (HIE, brain tumor, stroke, metabolic)[3]

CSF: pleocytosis, OCBs, high protein but <u>normal glucose</u>



1. Binks et al. *Pract Neurol* 2022; 2. Schmitt et al. *Neurology* 2012; 3. Baykan et al. *Clin EEG Neurosci* 2018; 4. Hirsch et al. 2021

Scoring focus on features of encephalitis

- Antibody Prevalence in Epilepsy (APE), 2017
- Predicts neural antibody positivity in patient with seizures
- Later revised (APE^2) in population presenting with cognitive dysfunction
- **Score ≥4**, sens 98%, spec 85%

1A: Antibody prevalence in epilepsy and encephalopathy (APE^2 score)	Value
New onset, rapidly progressive mental status changes that developed over 1–6 weeks or new onset seizure activity (within one year of evaluation)	(+1)
Neuropsychiatric changes; agitation, aggressiveness, emotional lability	(+1)
Autonomic dysfunction [sustained atrial tachycardia or bradycardia, orthostatic hypotension (≥20 mmHg fall in systolic pressure or ≥ 10 mmHg fall in diastolic pressure within three minutes of quiet standing), hyperhidrosis, persistently labile blood pressure, ventricular tachycardia, cardiac asystole or gastrointestinal dysmotility][a]	(+1)
Viral prodrome (rhinorrhea, sore throat, low grade fever) to be scored in the absence of underlying systemic malignancy within 5 years of neurological symptom onset	(+2)
Faciobrachial dystonic seizures[e]	(+3)
Facial dyskinesias, to be scored in the absence of faciobrachial dystonic seizures	(+2)
Seizure refractory to at least to two anti-seizure medications	(+2)
CSF findings consistent with inflammation[b] (elevated CSF protein > 50 mg/dL and/or lymphocytic pleocytosis > 5 cells/mcL, if the total number of CSF RBC is < 1000 cells/mcL)	(+2)
Brain MRI suggesting encephalitis[b] (T2/FLAIR hyperintensity restricted to one or both medial temporal lobes, or multifocal in grey matter, white matter, or both compatible with demyelination or inflammation)[c]	(+2)
Systemic cancer diagnosed within 5 years of neurological symptom onset[c] (excluding cutaneous squamous cell carcinoma, basal cell carcinoma, brain tumor, cancer with brain metastasis)	(+2)
	Total (max: 18)

May miss patients with autoimmune-associated epilepsy!!

Dubey et al. *Epilepsia* 2019

More recent tools aim to be relevant to epilepsy presentations

To identify antibodies in pt with focal epilepsy of unknown etiology
Prospective multicenter cohort study
Adults with focal epilepsy of unknown etiology, without recognized AE
n= 582 pt, 20 (3.4%) had autoimmune etiology of seizures

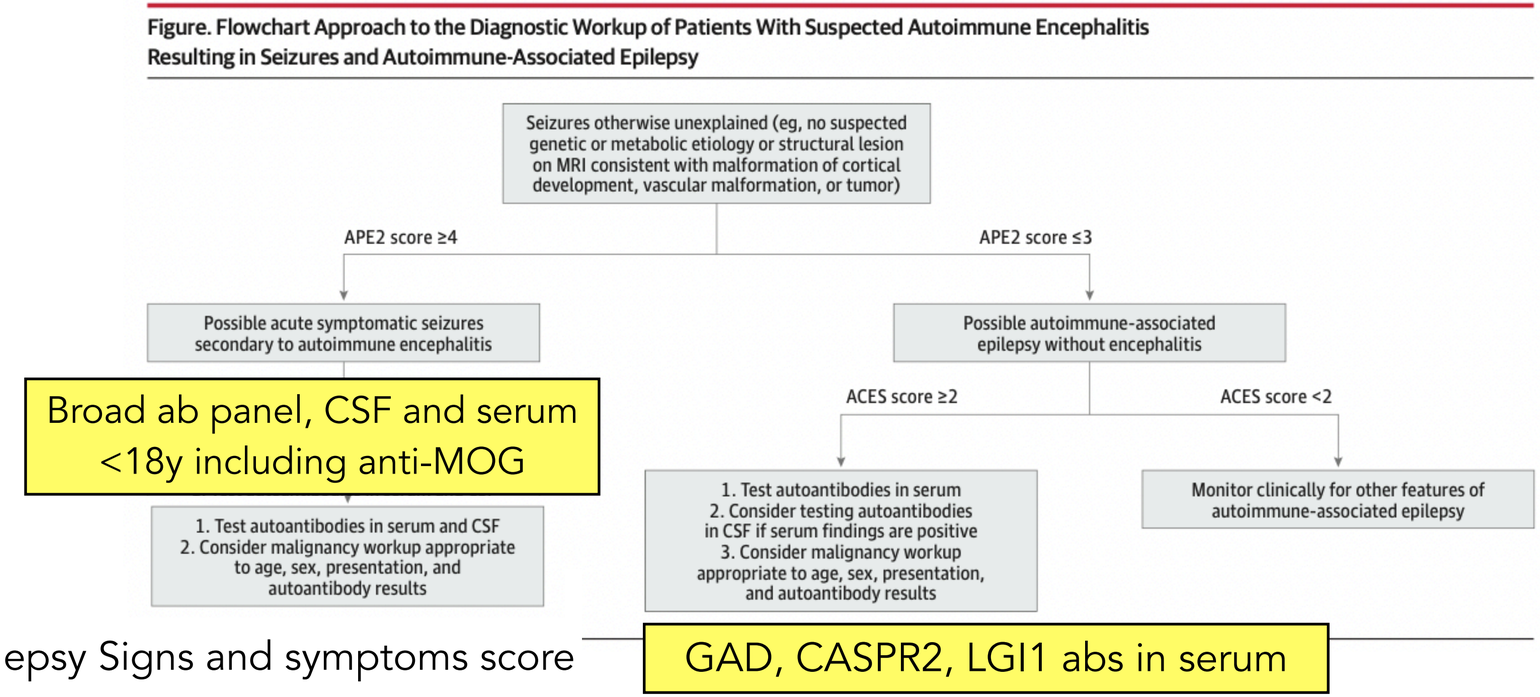
ACES SCORE

	Point
Cognitive symptoms	1
Behavioral changes	1
Autonomic symptoms	1
Speech problems	1
Autoimmune diseases	1
Temporal MRI hyperintensities	1

score max 6
score ≥ 2
sens 100%, spec 84.9%

ACES score,
Antibodies Contributing to focal Epilepsy Signs and symptoms score

Flowchart approach to diagnostic workup



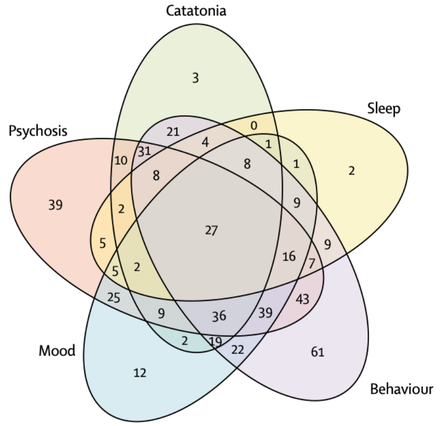
Broad ab panel, CSF and serum <18y including anti-MOG

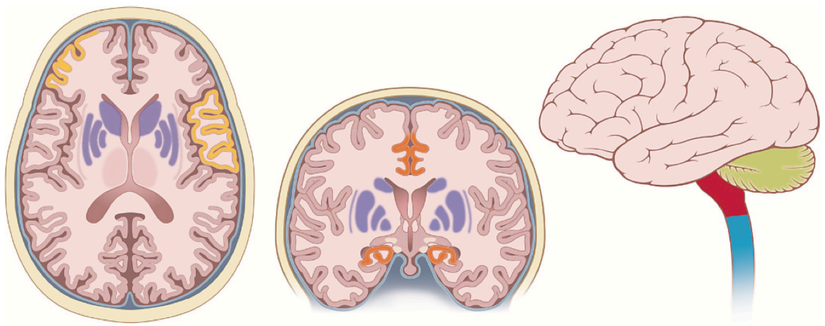
GAD, CASPR2, LGI1 abs in serum

De Bruijn et al. *Ann Neurol* 2021; Steriade et al. *JAMA Neurol* 2021

Treatment approach

Specific treatment	Symptomatic treatment
Addressing underlying etiology with immunotherapy	**Antiseizure medications (ASM)**: ASM alone, seizure control in 15% higher responder rates with SCB[1] (CBZ, LCS, PHT, OXC)
Likely paraneoplastic? prompt identification/Rx of tumor	movement disorders, gait instability psychosis/behaviour, mood, cognition, sleep, autonomic dysfunction





Limbic System	Extra-limbic Cortices	Basal Ganglia	Cerebellum	Brainstem	Spinal Cord
Anti-Hu **Anti-VGKC** **Anti-GAD65** **Anti-GABA B** **Anti-AMPAr** Anti-LGI1 Anti-VGCC	**Anti-NMDAr** **Anti-VGCC** **Anti-GABA-A** **Anti-GLuR3**	**Anti-CV2** **Anti-D2** Anti-NMDAr	**Anti-Yo** **Anti-GLuR1** Anti-Hu Anti-GAD65 Anti-VGCC	**Anti-Ma** **Anti-Ri** Anti-Hu Anti-Yo	**Anti-GLyR** Anti-Hu Anti-GAD65 Anti-CV2

1. Feyissa et al. *Neurology* 2017; 2. Al-Diwani et al. *Lancet Psychiatry* 2019; 3. Ball et al. *Clin Imaging* 2022

Immunotherapy

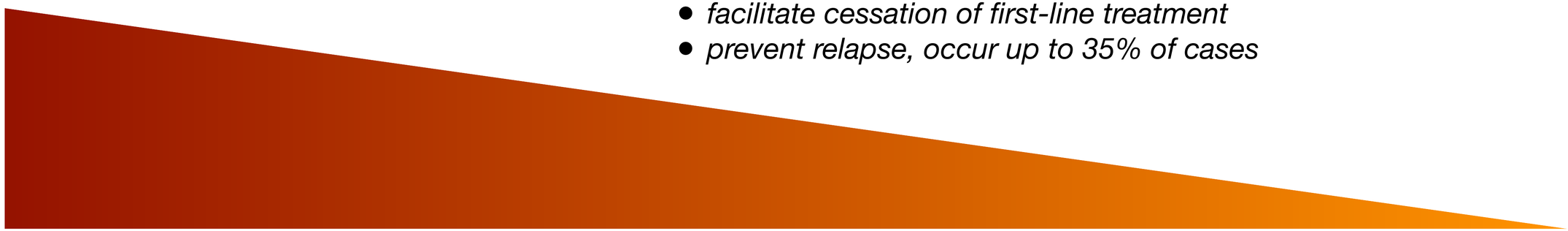
First-line
(isolation or combinations)
- IVMP, 3-5 d
- IVIG, 3-5 d
- Plasma exchange, 5-7 Rx

Second-line
- Rituximab
- Cyclophosphamide

Therapies used in induction with gradually increasing intervals over ensuing 6-12 mo
e.g.
- IVMP 1000 mg wkly, gradually increase intervals
- oral prednisolone1MKD 3 mo, gradually reduce
- IVIG 0.4 g/kg once wkly, gradually increase intervals

oral mycophenolate mofetil or oral azathioprine
- after establishment to response to induction therapy
- 3-5 years

Induction therapy	Maintenance therapy



- *facilitate cessation of first-line treatment*
- *prevent relapse, occur up to 35% of cases*

None of these treatment have been subject to placebo-controlled trials,
except single-center study comparing IVIG to placebo in anti-LGI1 ab encephalitis

Gillinder & Britton. *Continuum (Minneap Minn)* 2022

When the use of immunotherapy is uncertain?

RITE score

(Response to Immunotherapy in Epilepsy score)

- **Score ≥ 7**, sens 87.5%, spec 83.8%

	Score
New-onset, rapidly progressive mental status changes that developed over 1-6 weeks or new-onset seizure activity (within 1 year of evaluation)	1
Neuropsychiatric changes: agitation, aggressiveness, emotional lability	1
Autonomic dysfunction (sustained atrial tachycardia or bradycardia, orthostatic hypotension [≥20 mm Hg fall in systolic pressure or ≥10 mm Hg fall in diastolic pressure within 3 minutes of quiet standing], hyperhidrosis, persistently labile blood pressure, ventricular tachycardia, cardiac asystole, or gastrointestinal dysmotility)	1
Viral prodrome (rhinorrhea, sore throat, low-grade fever), only to be scored in the absence of underlying malignancy	2
Faciobrachial dystonic seizures	3
Facial dyskinesias, to be scored in the absence of faciobrachial dystonic seizures	2
Seizure refractory to at least to two antiseizure medications	2
CSF findings consistent with inflammation (elevated CSF protein >50 mg/dL and/or lymphocytic pleocytosis >5 cells/mm^3, if the total number of CSF red blood cells is <1000 cells/mm^3)	2
Brain MRI suggesting encephalitis (T2/FLAIR hyperintensity restricted to one or both mesial temporal lobes, or multifocal in gray matter, white matter, or both compatible with demyelination or inflammation)	2
Systemic cancer diagnosed within 5 years of neurologic symptom onset (excluding cutaneous squamous cell carcinoma, basal cell carcinoma, brain tumor, cancer with brain metastasis)	2
Initiation of immunotherapy within 6 months of symptom onset	2
Neural plasma membrane autoantibody detected (*N*-methyl-D-aspartate [NMDA] receptor antibody, γ-aminobutyric acid A [$GABA_A$] receptor antibody, γ-aminobutyric acid B [$GABA_B$] receptor antibody, α-amino-3-hydroxy-5-methylisoxazole-4-propionic acid [AMPA] receptor antibody, dipeptidyl-peptidase–like protein 6 [DPPX], metabotropic glutamate receptor 5 [mGlur1], mGluR2, mGluR5, leucine-rich glioma inactivated protein 1 [LGI1] antibody, IgLON5, contactin-associated proteinlike 2 [CASPR2] antibody or myelin oligodendrocyte glycoprotein [MOG])	2
Maximum score	**22**
Cutoff score predicting favorable seizure outcome	**≥7**[b]

Dubey et al. *Epilepsia* 2017

Prompt identification and treatment of tumor is paramount

Unusual presentation of more common disease/injury

CASE REPORT

Rapid recovery from catastrophic paraneoplastic anti-NMDAR encephalitis secondary to an ovarian teratoma following ovarian cystectomy

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SUMMARY

This report is aimed to describe a life-threateni
anti-*N*-methyl-D-aspartate receptor (NMDAR) e
secondary to ovarian teratoma with rapid recov
1 day after the removal of the tumour. A 23-ye
woman presented with sudden headache, pers

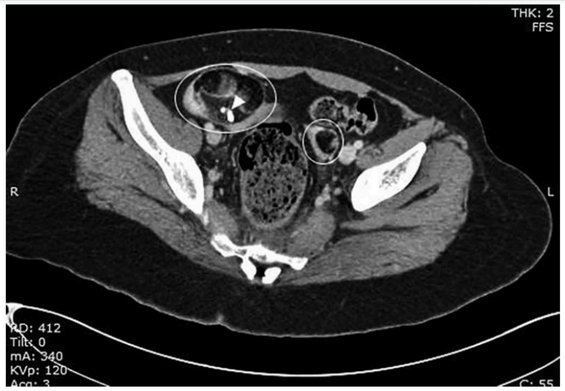


Figure 1 Abdomen CT scan shows bilateral ovarian cysts suggestive of teratoma, measuring 5.5×4.3 cm (right) and 2.6×2.2 cm (left), note the calcifications in the right cyst (arrow head).

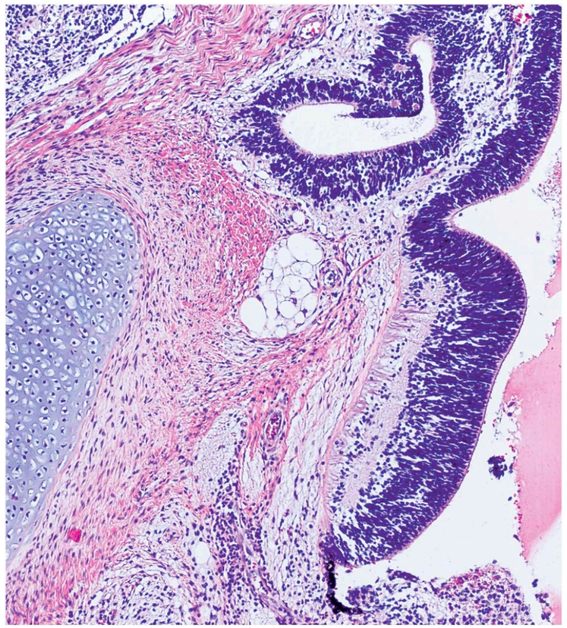
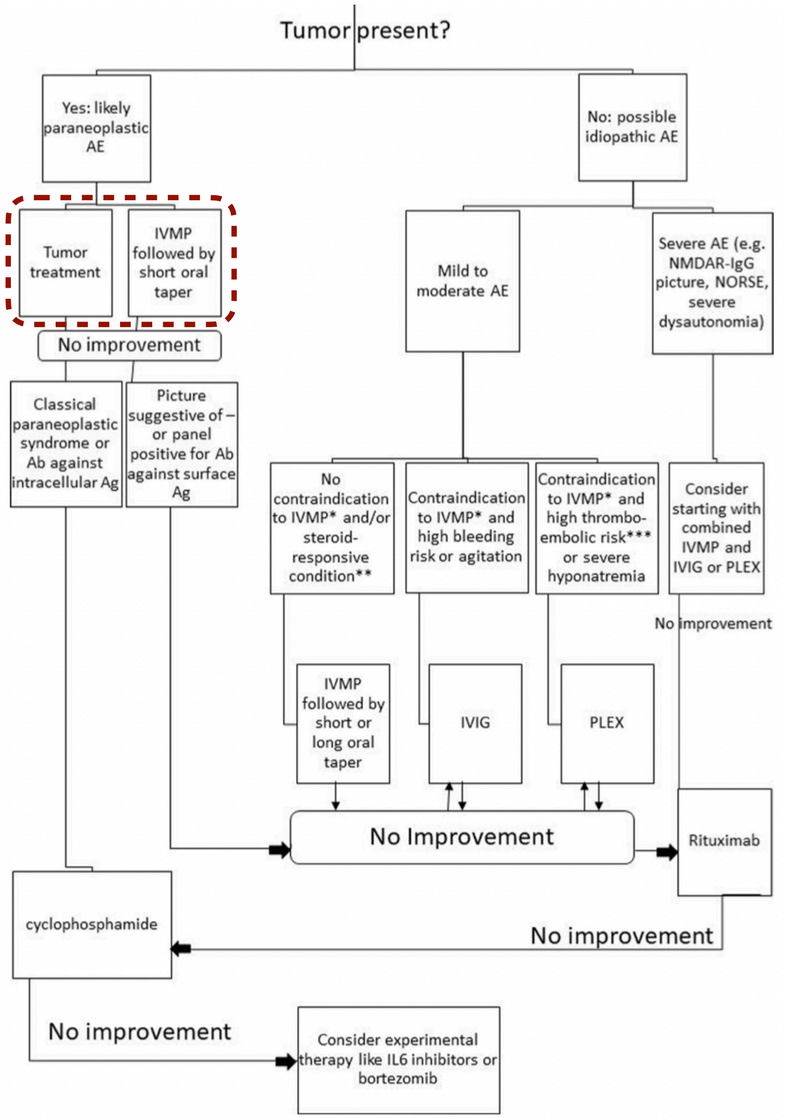


Figure 5 Pathological examination: the left ovarian tumour shows both immature neuroepithelial and mesenchymal components.



1. Tantipalakorn et al *BMJ Case Rep* 2016; 2. Abboud et al. *J Neurol Neurosurg Psychiatry* 2021

Conclusions

- Autoimmune-associated seizures are increasingly encountered in clinical practice
- Acute symptomatic seizure behave differently than autoimmune-associated epilepsy
- Critical recognizing early in their course because delays can result in poorer outcomes
- Scoring have been developed to improve certainty of dx and therapeutic decision making

ขอบคุณครับ

สมัครเป็นสมาชิกครอบครัว Neuro CMU

สแกน qr code

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