



Evidence Review:

Intravenous immunoglobulin for autoimmune encephalitis

NHS England

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1. Introduction

Autoimmune encephalitis (AIE) is used to describe a group of disorders characterised by symptoms of limbic and extra-limbic dysfunction occurring in association with antibodies against synaptic antigens and proteins localised on the neuronal cell surface. These autoimmune conditions include, but are not limited to, VGKC-complex antibody associated encephalitis, NMDA-receptor antibody associated encephalitis, GAD antibody associated encephalitis, MOG antibody disease and Hashimoto's encephalitis. AIE symptoms include amnesia, seizures, psychosis, abnormal movements, autonomic dysregulation, hemiplegia, visual loss and coma.

Human immunoglobulin is a sterile preparation of concentrated immunoglobulins recovered from pooled human plasma or serum obtained from outside the UK, tested and found non-reactive for hepatitis B surface antigen and for antibodies against hepatitis C virus and human immunodeficiency virus (types 1 and 2). Intravenous immunoglobulin (IVIg) is proposed as a treatment option for the above indications where the condition is unresponsive to first line treatments such as steroid therapy or plasma exchange (PLEX), or first line treatments are contra-indicated.

A global shortage of human immunoglobulin and the rapidly increasing range of clinical indications for treatment with immunoglobulins has resulted in the need for a Demand Management programme for IVIg in the UK. IVIg is commissioned by NHSE in line with Clinical Guidelines for Immunoglobulin Use (Department of Health, Second edition update, July 2011). ADEM and AIE are classified as grey indications, and IVIg is not currently routinely commissioned by NHS England for these conditions. Grey indications are those diseases where the evidence is weak, in many cases because the disease is rare, and treatment should be considered on a case by case basis.

2. Summary of results

What is the clinical effectiveness of IVIg for autoantibody-associated neurological encephalitis syndromes, when used:

- a) instead of PLEX in those who haven't responded to steroids alone,
- b) are critically ill and need to optimise treatment urgently (i.e. in combination with steroids), or c) in poor responders to steroids and PLEX?

Evidence for the effectiveness of IVIg in autoimmune encephalitis due to autoantibodies targeting neuronal cell surface proteins comes from one large systematic review by Nosadini et al 2015 which includes all articles published up to September 2015 and three other small sized studies not included in this review. There are no randomized controlled trials of treatment, and the majority of studies are small sized retrospective case series, except for the study by Titulaer et al which included 577 patients.

The review does not include subgroup analysis by treatment groups but provides information comparing patients receiving immunomodulatory therapy with those receiving no immunomodulatory therapy. It appears that IVIg when used in combination with other immunomodulatory treatments has better outcomes compared to patients with no immunotherapy. This appears true for encephalitis syndromes due to anti-NMDAR antibodies, anti-Capr2 antibodies, anti-GABAR antibodies, anti-GABAR antibodies, anti-GPPX antibodies, anti-GlyR antibodies. Anti-IgLON5 encephalitis appears to be different from the other autoimmune encephalitides, with poor response to immune therapy and a high mortality rate.

Nosedini et al 2015 also concluded that early commencement of immune therapy is more commonly associated with better outcomes, and that the use of second-line immune therapies is more commonly associated with better outcomes and a lower rate of relapse, but is influenced by severity bias, as sicker patients are more likely to receive second-line therapy.

Evidence for the use of IVIg in autoimmune encephalitis due to paraneoplastic syndrome (PND) is derived from one review article by Sadeghian et al 2010 and a number of small sized studies (Vodopevic et al 2015, Moon et al 2014, Omlez et al 2013). Based on this Clinical Evidence Review, there is some evidence that the use of IVIg might have a positive impact, but as IVIg was used with other immunomodulatory treatments it is not possible to assess the specific impact of IVIg on patient outcomes. Sadeghian et al 2010 recommend IVIg use in PND affecting peripheral nervous system. Evidence level 3-4.

The main limitations of the evidence were the limited number of patients studied and the retrospective and non-standardised nature of both data and outcome measures. Most studies did not include a precise definition of patient outcomes, though a few studies used the Modified Rankin Scale to measure this. It also appears that a standardised protocol for the use of IVIg was lacking, and the majority of the studies did not present detail on the sequence of use of drugs/therapies used. In a very small number of patient studies IVIg was used as first line treatment but in most of the studies, IVIg was used in combination with steroids and/or PLEX. Due to the lack of subgroup analysis, and lack of details on sequence of use of drugs used in the treatment, the available evidence does not make it possible to reach definitive conclusions about the clinical effectiveness of IVIg in AIE.

What is the cost effectiveness of IVIg for autoantibody-associated neurological encephalitis syndromes, when used a) instead of PLEX in those who haven't responded to steroids alone, b) are critically ill and need to optimise treatment urgently (i.e. in combination with steroids), or c) in poor responders to steroid and PLEX?

There are no published studies evaluating cost effectiveness of IVIg in autoantibody-associated neurological encephalitis syndromes.

3. Research questions

What is the clinical effectiveness of IVIg for autoantibody-associated neurological encephalitis syndromes, when used a) instead of PLEX in those who haven't responded to steroids alone, b) are critically ill and need to optimise treatment urgently (i.e. in combination with steroids), or c) in poor responders to steroid and PLEX?

What is the cost effectiveness of IVIg for autoantibody-associated neurological encephalitis syndromes, when used a) instead of PLEX in those who haven't responded to steroids alone, b) are critically ill and need to optimise treatment urgently (i.e. in combination with steroids), or c) in poor responders to steroid and PLEX?

4. Methodology

A review of published, peer reviewed literature has been undertaken based on the research questions set out in Section 3 and a search strategy agreed with the lead clinician and public health lead for this policy area. This has involved a PubMed search and search of the Cochrane database for systematic reviews, in addition to review of any existing NICE or SIGN guidance. The evidence review has been independently quality assured.

An audit trail has been maintained of papers excluded from the review on the basis of the inclusion and exclusion criteria agreed within the search strategy. The full list has been made available to the clinicians developing the policy where requested.

5. Results

A detailed breakdown of the evidence is included in the Appendix.

Appendix One

Grade	5	Study des	ign and interven	tion		Outcom	nes			Reference			Other
Grade of	Study design	Study	Intervention	Clinical	Category	Primary Outcome	Primary Result	Secondary	Secondary Result	Reference	Complicatio	Benefits	Comments
evidence	, ,	size		Population	0 ,	· ·		Outcome			ns noted	noted	
2-	Systematic	1167	IVIg, review	Auntibody	Clinical	Varied form study to study but mainly assessed in terms of Modified	IVIg was part of the treatment for all of	None	-	Nosadini,	no separate	as in	This is a well designed systematic review of studies
	-		includes an	related	effectiveness	Rankin Scale where 0 indicates no symptoms post treatment to 5	the autoantibody related encepahilaitis.			Margherita;	reporting for	primary	on encephalitis due to autoantibodies targeting
			analysis of other	encephaloomyel	of the	indicating sever disability and 6 indicating death.	The proportion of patients treated with			Mohammad,	IVIg	outcome	neuronal cell surface proteins. The review includes
			treaments	itis	intervention		IVIg varied from 27% to 54%. Generally			Shekeeb S.;			studies published upto Nov 2015 on nine different
			including			Anti NMDAR: 92.4% received immune therapy (766/829) and IVIg was	for most of the conditions patients			Ramanathan,			types of antibodies:
			steriods, PLEX,			used in 66% (502/761). In the large case series by Titulaer et al.,	treated with combination of immune			Sudarshini; Brilot,			There are no randomized controlled trials of
			and immune			steroids and IVIg were often given together (202/462, 44%). It is	therapy had better outcomes compared			Fabienne; Dale,			treatment, and the majority of studies are small
			therpaies			reported that people who didn't receive immune therapy had significantly	to patients who had no immune therapy.			Russell C			sized retrospective case series except study by
			including			poorer outcomes than those who received immune therapy. Also in the	Due to lack of subgroup analysis by IVIg			Immune therapy			Titulaer et al which include 577 patients.
			azthiprine,			largest case series consisting by Titulaer steroids and IVIg were often	it wsa not possible to asses the impact			in autoimmune			The review does not include subgroup analysis by
			methotrexate,			given together (202/462, 44%). It appears that IVIg when combined with	of IVIg on the outcomes.			encephalitis: a			treatment groups but provides information
			tacrolimus			other treatments had better outcome but is not possible to evaluate				systematic			comparing patients receiving immunomodulatory
						effectiveness of IVIg on its own due to lack of subgroup analysis.				review. Expert			therapy vs no-immunomodulatory therapy.
										Rev Neurother			However it appears IVIg when used in combination
						Anti LGI1: 97.2% (103/106) received immune therapy and IVIg was				2015;0(0):47119.			with other immunomodulatory treatments has better
						used in in 50% (53/106) There is no subgroup analysis by IVIg group.							outcomes when compared to patients with no
						Good outcome (full recovery or mRS 0) was 27.8% (20/72) in the							immunotherapy. These include encephalitis
						studies using neurological status as an outcome measure, 86.4%							syndromes due to anti-NMDAR antibodies, anti-
						(19/22) patients were seizure-free in the studies using seizure status as							Capr2 antibodies, anti-GABABR antibodies, anti-
						the main outcome measure and relapses occurred in 18% patients							GABAAR antibodies, anti-DPPX antibodies, anti-
						(16/89), and death in 2.5% (4/158) patients. As above It is not possible							GlyR antibodies.
						to evaluate effectiveness of IVIg on its own due to lack of subgroup							Anti-lgLON5 encephalitis appears to be different
						analysis.							from the other autoimmune encephalitides, with poor
						Anti-Caspr2:92.5% immune therapy (37/40, and IVIg was used in							response to immune therapy and high mortality rate.
						38.1% (8/21) There is no subgroup analysis by IVIg group. Outcome							Authors reommend that based on evidence early
						data was poorly reported and authors report relapse was uncommon,							commencement of immune therapy is more
						and full recovery occurred in about one-fourth of patients, whereas							commonly associated with a better outcome and the
						12.1% died (4/33 with adequate information). As above it is not							use of second-line immune therapies is more
						possible to evaluate effectiveness of IVIg on its own due to lack of							commonly associated with a better outcome and a
						subgroup analysis.							lower rate of relapses but is influenced by severity
													bias, as sicker patients are more likely to receive
						Anti-AMPAR: 95.2% (40/42), received immune therapy and IVIg was							second-line therapy.
						used in in 52.4% (22/42)There is no subgroup analysis by IVIg group							The main limitations are the limited number of
						.10.8% of patients had a full recovery (mRS 0) (4/37), whereas most							patients and the retrospective and non-standardized
						cases recovered partially (25/37, 67.6%). As above It is not possible to							nature of data and outcome measures. Majority of
						evaluate effectiveness of IVIg on its own due to lack of subgroup							the studies did not include definition of an outcome
						analysis.							except few studies which uses Modified Ranskin Scale to measure outcomes.
						Anti GABAAR: 54.5% (18/33) received first line immune therapy : and							Also it appears there is no one standard protocol
						IVIg was used in 27.3% (9/33) There is no subgroup analysis by IVIg							for use IVIg and majority of the studies did not detail
		l				group. Patients receiving immune therapy had better outcomes than							on the sequence of use of drugs/therapies used in
		l				those who did not receive immune therapy (mean Rankin Score 0: 2/18,							the treatment. In very small number of patients
		l				11.1% vs. 0/12, 0%), though there was a higher rate of relapse (3/18,							studies IVIg was used as first line treatment but in
		l				16.7% vs. 1/12, 8.3%). As above It is not possible to evaluate							most of the studies it IVIg were used in combination
		l				effectiveness of IVIg on its own due to lack of subgroup analysis.							with steroids +/- PLEX.
		l				and a subgroup analysis.							Due lack of subgroup analysis and lack of details
		l				Anti-GlyR antibodies: 77.3% (58/75) of patients with available data							on sequence of use drugs used in the treatment it is
		l				received immune therapy and IVIg in 42.6% (29/68). In the largest							not possible to answer the PICO question 1.
		l				published series, approaches to first-line immune therapy were variable							Study has been graded evidence level 2+ because
		ı	l	ı	ı	Passioned series, approaches to machine minute merapy were variable	I	ı		l	ı	I	Olday has been graded evidence level 2+ because

3	Other	not	IVIg along with	paraneoplastic		but typically started with intravenous methylprednisolone followed by high-dose prednisolone, and sometimes by PE, IVIg or both. There is no subgroup analysis by IVIg group but patients who received immune therapy had higher rate of good recovery than those who were not treated (mRS 0-1: 25/44, 56.8% vs. 1/10, 10%). As above due to lack of subgroup analysis and considering IVIg were used in combination with other drugs it not possible to draw conclusions on effectiveness of IVIg from these studies. Anti-DPPX antibodies: 64.3% (18/28) of patients received immune therapy during the first episode of disease and IVIg in 28.6% (8/28). There is no subgroup analysis by IVIg but authors report patients who did not receive immune therapy at the first episode had worse outcomes than patients who did receive immune therapy at the first episode had worse outcomes than patients who did receive immune therapy at the first episode had worse outcomes than patients who received second-line treatments at the first episode had better outcomes than patients who received second-line treatments at the first episode had better outcomes than patients who did not receive second line therapies (mRS 0-1: 4/14, 0.4% vs. 3/17, 1.7.6%) and lower rates of death (0/10, 0% vs. 3/17, 17.6%) despite similar rates of relapses (3/10, 30% vs. 5/17, 29.4%). Due to lack of subgroup analysis and considering IVIg were used in combination with other drugs it not possible to draw conclusions on effectiveness of IVIg from these studies. Anti-IgLON5 antibodies: The majority of patients received immune therapy (9/10, 90%), even though most presented late. First-line treatments were used in 90% (9/10) (steroids in 3/10, 30%; IVIg in 4/10, 40%) and second-line therapies in 70% (7/10) (rituximab in 3/10, 30%; cyclophosphamide in 4/10, 40%). Authors report a poor outcome despite use of intense immunotherapy. 70% (7/10) (rituximab in 3/10, 30%; cyclophosphamide in 4/10, 40%). Authors report a poor outcome despite use of intense immunotherapy. 70% (7/10) (ritux	Authors report that IVIg had been used	none	-	Sadeghian,	none		of the case series and case reports with small sample sizes included in the review. However the evidence on anti NMDAR antibodies which are based on large sample size are generalizable but due to lack of subgroup analysis for IVIg subgroup it is not possible to generalise results to this subgroup.
3	Other	not imentio ed		neurological	Clinical effectiveness of the intervention	not mentioned but reported as improvement in symptoms	Authors report that IVIg had been used in variety of PNDs but mechanism of action of IVIg in PND is uncertain. Thet recommend based on and based on experience with othermetrological disorders IVIg seems most appropriate for PND affecting the peripheral nervous syste.	none		Sadeghian, Hamid; Vernino, Steven. Progress in the management of paraneoplastic neurological disorders. Ther Adv Neurol Disord 2010;3(1):43-52.	none reported	primary outcome	This is an overview article with reference to 13 articles that have information relating to immunomodulatory therapy in paranroplastic syndrome. With regard to IVIg in Paraneoplastic neurological disorders (PND), Authors report that IVIg had been used in variety of PNDs but mechanism of action of IVIg in PND is uncertain. Thet recommend based on and based on experience with otherneurological disorders IVIg seems most appropriate for PND affecting the peripheral nervous syste

		16	I		In	T		1	 			I=
	Systematic		IVIg	none	Clinical refrictiveness of the intervention	Varied from study to study but main outcomes include Effect of IVIg: on transient seizure frequency improved neurologic condition	Granata et al 2003 N=11)-1/11(9%) transient \(\) seizure frequency >50%, improved neurologic condition, \(2/11(18%) transient \(\) seizure frequency \(\) up to 50% \(5/11 \) (46%) not assessable. \(6/11 \) Allid improvement in symptoms \(Frucht et al 2001 \) (n=1) -Mild improvement in symptoms \(Frucht et al 2001 \) (n=1) -Marked improvement in hyperkinette movements \(\) Villani et al 2001 \((n=1) \) Marked improvement (>75% \(\) seizure \) frequency, improved cognition \(\) Leach et al 1999 \((n=2) \) 2/2 \((100%) \) Marked improvement in seizure \(\) frequency, hemiparesis, cognition	none	Feasby, Tom; Banwell, Brenda; Benstead, Timothy; Bril, Vera; Brouwers, Melissa; Freedman, Mark; Hume, Heather; Freedman, John; Pi, David; Wadsworth, Louis. Guidelines on the use of intravenous immune globulin for neurologic conditions. Transfus Med Rev 2007;21(2 Suppl 1):S57- 107.	none reported	as in primary outcome	Rasmussen's encephalitis is one of the condition that is included in the list of condtions approved for use of IVIg. The approval was based on one case series and four case reports of IVIg use for Rasmussen's encephalitis. Overall, 31% (5/16) of patients showed marked improvement in symptoms following IVIg alone or in combination with additional therapies.
2-	Systematic + Meta Analysis	0	IVig	Fisher Syndrome and related disorders	Clinical effectiveness of the intervention	none reported			Overell, J. R.; Hsieh, S. T.; Odaka, M.; Yuki, N.; Willison, H. J. Treatment for Fisher syndrome, Bickerstaff's brainstem encephalitis and related disorders. Cochrane Database Syst Rev 2007;0(1):CD004 761.	-	-	This Cochrane reviewand authors conclude that there are no randomised controlled trials of immunomodulatory therapy in Fisher Syndrome or related disorders on which to base practice.
3	Systematic	38 patients	īVig	Cerebellar ataxia	Clinical effectiveness of the intervention	Not defined but outcomes reported as outcomes in clinical symptoms	1. Gluten ataxia,-Improved cerebella ataxia symptoms but not completely 2. Patients Resistance to gluten therpaytransient response to IVIg and relapse after disconituration of treatment. 3. Cerebellar ataxia with paraneoplastic cerebellar degeneration -no response to IVIg in combination with IV methylprednisolone and cyclophosphamide, 4. Cerebellar atxia with anti-CAD antibodies-high response to combination of immonotherpaies including IVIg but remision in longterm. Hashimotos encephalopathy- good response to steroids and no difference bewteen different types of immunotherpaies.	none	Mitoma, Hiroshi; Hadjivassiliou, Marios; Honnorat, Jérôme. Guidelines for treatment of immune- mediated cerebellar ataxias. Cerebellum Ataxias. 2015;2(0):14.	none reported	as in primary outcome	This is a systematic review of studies on cerebellar ataxia. The review identified 32 studies, majority of studies were case reports with 1 patients. There analysis showed IVIg and other immunotherapies had variable responses on various cerebellar ataxic conditions. 1.Gluten ataxia, Improved cerebellar ataxia symptoms but not completely 2. Patients Resistance to gluten therapy-transient response to IVIg and relapse after disconitunation of treatment. 3. Cerebellar ataxia with paraneoplastic cerebellar degeneration -no response to IVIg in combination with IV methylprednisolone and cyclophosphamide, 4. Cerebellar attaxia with anti-GAD antibodies-high response to combination of immonotherapies including IVIg but remisition in longterm. Hashimotos encephalopathy- good response to steroids and no difference bewteen different types of immunotherapies. Overall the level of evidence is 3 as the majority of studies included had just one patient and lack of comparator.

-	r	r		-			Tarana a managaran				1		I=
3	Case series	5	glucocrticods	Susac	clinical	clinical response	Patients treated with glucocrticoids and	None	-	Vodopivec,	none	as in	This is retrospective study of 5 patients with Susac
			and IVIg	syndrome	effectiveness		IVIg experienced clincial pregression and			Ivana; Venna,	reported	primary	syndrome. All received corticosteroids and IVIg.
							further required additional			Nagagopal;		outcome	Patients had no response to IVIg and corticosteroids
							immunospressive therpies			Rizzo, Joseph F.;			and were further treated with additional
										Prasad.			immunosupressants. Level pf evidence=3
										Sashank, Clinical			
										features.			
										diagnostic			
										findings, and			
										treatment of			
										Susac syndrome:			
										A case series, J.			
										Neurol. Sci.			
										2015;357(42036)			
										:50-57.			
										.50-57.			
2	Case series	34	IVIg was one of	autoimmune	Clinical	seizure frequency	Specific autoimmune antibodies were	none	none	Dubey,	none	as in	This is a retrospective case series of 34 patients with
3	Case series	34				Seizure frequency		none	none				
I	l	l	immunomodulat	epilepsy	effectiveness		detected in 76.5% (26) of patients; anti-			Divyanshu;	reported	primary	autoimmune epilepsy.Specific autoimmune
I	l	l	ory drug used		of the		VGKc in 23.5% (8); anti-			Samudra,	1	outcome	antibodies were detected in 76.5% (26) of patients;
	l	l	along with		intervention		NMDA-R in 20.6% (7); anti-thyroid/TPO			Niyatee; Gupta,	1	l	anti-VGKc in 23.5% (8); anti-NMDA-R in 20.6% (7);
			corticosteroids,				in 14.7% (5); anti-GAD in 11.8% (4); anti-			Puneet; Agostini,			anti-thyroid/TPO in 14.7% (5); anti-GAD in 11.8%
			plasmapheresis,				GABAB in 5.9% (2). Nine patients			Mark; Ding, Kan;			(4); anti-GABAB in 5.9% (2). Nine patients (26.5%)
			and				(26.5%) included in the study had an			Van Ness, Paul			included in the study had an underlying malignancy;
			chemotherapeut				underlying malignancy; 2 ovarian			C.: Vernino.			2 ovarian teratomas, 2 breast cancer, 1
			ic agents such				teratomas, 2 breast cancer, 1			Steven; Hays,			adenocarcinoma of the lung, 1 small cell lung
			as rituximab,							Ryan.			
							adenocarcinoma of the lung, 1 small cell			*			cancer, 1 testicular cancer, 1 papillary thyroid cancer
			cyclophosphami				lung cancer, 1 testicular cancer, 1			Retrospective			and 1 thymoma.
			de,				papillary thyroid cancer and 1 thymoma.			case series of the	:		94.1% (32) patients received immunomodulatory
			mycophenolate.				94.1% (32) patients received			clinical features,			therapies, including high dose corticosteroids
							immunomodulatory therapies, including			management and			(96.8%), plasmapheresis (62.5%), and IVIg (34.4%).
							high dose corticosteroids			outcomes of			9 (28.1%) patients received only high dose
							(96.8%), plasmapheresis (62.5%), and			patients with			corticosteroids as immunomodulatory therapy for
							IVIg (34.4%). 9 (28.1%) patients			autoimmune			acute management of recurrent seizures, whereas
							received only high dose corticosteroids			epilepsy. Seizure			the remaining patients received a combination of
							as immunomodulatory therapy for acute			2015;29(0):143-			corticosteroids with plasmapheresis and/or IVIg
										147.			[Corticosteroids with plasmapheresis and/or rvig
							management of recurrent seizures,			147.			
							whereas the remaining patients received						+ plasmapheresis: 12(35.3%), Corticosteroids + IVIg
							a combination of						+ plasmapheresis: 8 (25%), Corticosteroids + IVIg 3
							corticosteroids with plasmapheresis						(9.4%)]. 63.3% (19) of patients had 50% reduction in
							and/or IVIg [Corticosteroids						seizure frequency (RR) at the first clinic visit,
							+ plasmapheresis: 12(35.3%),						following inpatient management of acute episode. 6
	l	l	1		l		Corticosteroids + IVIg + plasmapheresis:				1	l	(17.6%) patients had complete resolution of seizures
1	l	l	1		l		8 (25%), Corticosteroids + IVIg 3				1	l	on initial clinic follow up.
	l	l	ĺ		1		(9.4%)].			l	1	l	Overall it appears that IVIg may have some influence
l	l	l	1		l		(9.4%)]. 63.3% (19) of patients had 50%				1	l	
l	l	l	1		l						1	l	on the prognosis. However the generalisability of the
	l	l	ĺ		1		reduction in seizure frequency			l	1	l	results of IVIg is very limited because IVIg was given
	l	l	ĺ		1		(RR) at the first clinic visit, following			l	1	l	in combination with other treatments and also due to
l	l	l	ĺ		1		inpatient management of			l	1	l	patient selection methods which lacks a control
l	l	l	1		l		acute episode. 6 (17.6%) patients had				1	l	group and it is difficult estimate the effectiveness of
l	l	l	ĺ		1		complete resolution of			l	1	l	IVIg.
	l	l	1		l		seizures on initial clinic follow up.				1	l	
1	l	l	1		l						1	l	
	l	l	1		l						1	l	
	l	l	1		l						1	l	
	l	l	1		l						1	l	
I	l	l	ĺ		1					l	1	l	
1	l	l	1							l	1	ı	

3	Case series	36	-	anti NMDAR	Clinical	improvement in neurological symptoms measured using mRs score	All patients received first-line treatment	none		keridou,	none	as in	This is a prospective case series of 36 children with
				encephalitis	effectiveness		and Median time to first line treatment			nastasia;	reported	primary care	anti NMDAR -ab. All patients received first-line
					of the		was 19days. The types of first-line			arantoni,			treatment and median time to first line treatment was
					intervention		treatment were including corticosteroids			genia;			19 days. The types of first-line treatment were
							in 86 %, Intravenous immunoglobulins in			accoz,			corticosteroids in 86 %, Intravenous
							89 %, and Plasma exchange in 39 %.			ırélien; Ducray,			immunoglobulins in 89 %, and Plasma exchange in
							81% received second-line treatment and			ançois; Gitiaux,			39 %. 81% received second-line treatment and
							median time from first-line to second-line			ril; Villega,			median time from first-line to second-line treatment)
							treatment) was 26 days. The types of			édéric; Deiva,			was 26 days. The types of second-line treatments
							second-line treatment included rituximab			ımaran;			included rituximab in 72 %, Cyclophosphamide in 14
							in 72 %, cyclophosphamide in 14 %, and			ogemond,			%, and 17% received long-term immunosuppression
							17% received long-term			eronique;			(does not mention with what drugs).
							immunosuppression (do not mention			athias, Elodie;			Outcome was measured using Modified Rankin
							with what drugs).		Pica	,			scale and were considered to have good outcome if
							Outcome was measured using Modified			éraldine;			mRs score was =2. At 12 months, 83 % had a</td
							Rankin scale and were considered to			ırdieu, Marc;			Good outcome and at 24 months same proportion
							have good outcome if mRs score was			ntoine, Jean-			were reported to have good outcome. 8% had
							=2. At 12 months 83 % had a Good</td <td></td> <td></td> <td>nristophe;</td> <td></td> <td></td> <td>relapses after diagnosis. The results of outcome are</td>			nristophe;			relapses after diagnosis. The results of outcome are
							outcome and 24 months same			elattre, Jean-			not available by IVIg or any other treatment
							proportion were reported to have Good		Yve	es; Honnorat,			subgroups. Evidence level 3 due to lack of
							outcome. 8% had relapses after		0010	rome.			comparator, case selection methods and small
			1]	Ì		diagnosis. The results are of outcome			eatment and			sample size.
			1]	Ì		are not available by IVIg or any other			tcome of]
			1]	Ì		treatment subgroups.			ildren and			This study was included in study Nosadini et al,
			1]	Ì					lolescents with			2015.
			1]	Ì					methyl-D-			
			1		1					partate			
					1					ceptor			
										cephalitis. J.			
									Neu	eurol.			
									201	15;262(8):185			
									9-18	1866.			
		31	F	NMDAR-Ab-	Clinical	F	All 1: 1 - 1 - 1 - 00 (740)		147 :	right, Sukhvir;			71
3	Case series	31	First line-			clinical improvement in symptoms based on modified Rankin scale	All patients received steroids; 22 (71%)	none			authors	as in primary	This is a retrospective case series of 31 children who had NMDAR-Ab-associated neurological disorders.
1			Steroids,IVIg,	associated	effectiveness	0=full recovery, 5=dead	received IVIg, 9 (29%)			acohen, Yael;	report no	F	
			PLEX, second	neurological	of the	U=rull recovery, 5=dead	received PLEX and 10 (32%) received		Jaco	cobson, Leslie;	significant	outcome	All patients received steroids; 22 (71%) received
			PLEX, second line -			U=full recovery, 5=dead	received PLEX and 10 (32%) received second-line immunotherapy.Four		Jaco Agra	cobson, Leslie; grawal, Shakti;	significant treatment	F	All patients received steroids; 22 (71%) received IVIg, 9 (29%) received PLEX and 10 (32%) received
			PLEX, second line - cyclophosphami	neurological	of the	U=full recovery, 5=dead	received PLEX and 10 (32%) received second-line immunotherapy.Four treatment groups were identified within		Jaco Agra Gup	cobson, Leslie; grawal, Shakti; upta, Rajat;	significant treatment complication	F	All patients received steroids; 22 (71%) received IVIg, 9 (29%) received PLEX and 10 (32%) received second-line immunotherapy.Four treatment groups
			PLEX, second line - cyclophosphami de, rituximab,	neurological	of the	U=Tuli recovery, 5=dead	received PLEX and 10 (32%) received second-line immunotherapy.Four treatment groups were identified within the whole		Jaco Agra Gup Phili	cobson, Leslie; grawal, Shakti; upta, Rajat; nilip, Sunny;	significant treatment complication were	F	All patients received steroids; 22 (71%) received IVIg, 9 (29%) received PLEX and 10 (32%) received second-line immunotherapy.Four treatment groups were identified within the whole cohort of 31 patients.
			PLEX, second line - cyclophosphami	neurological	of the	U=full recovery, 5=dead	received PLEX and 10 (32%) received second-line immunotherapy.Four treatment groups were identified within the whole cohort of 31 patients. The most		Jaco Agra Gup Phili Smi	cobson, Leslie; grawal, Shakti; upta, Rajat; nilip, Sunny; nith, Martin;	significant treatment complication	F	All patients received steroids; 22 (71%) received IVIg, 9 (29%) received PLEX and 10 (32%) received second-line immunotherapy. Four treatment groups were identified within the whole cohort of 31 patients. The most frequently used (61%) were IVIg and
			PLEX, second line - cyclophosphami de, rituximab,	neurological	of the	U=full recovery, 5=dead	received PLEX and 10 (32%) received second-line immunotherapy. Four treatment groups were identified within the whole cohort of 31 patients. The most frequently used		Jaco Agra Gup Phili Smi Lim,	cobson, Leslie; grawal, Shakti; upta, Rajat; nilip, Sunny; nith, Martin; m, Ming;	significant treatment complication were	F	All patients received steroids; 22 (71%) received IVIg, 9 (29%) received PLEX and 10 (32%) received second-line immunotherapy. Four treatment groups were identified within the whole cohort of 31 patients. The most frequently used (61%) were IVIg and steroids (Group A); PLEX was added to these in two
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3	Case series	20	Thirteen	patients with	Clinical	Clinical response as improvement in symptoms	Of the 20 patients, 10 patients were	none		Moon, Jangsup;	0	as in	This is retrospective case series of 20 patients with
			patients were	anti	effectiveness		diagnosed with limbic encephalitis, 9			Lee, Soon-Tae;		primary	anti-amphiphysin syndrome. Of the 20 patients, 10
			treated with	amphiphysin	of the		with dysautonomia, 6 with cerebellar			Shin, Jung-Won;		outcome	patients were diagnosed with limbic encephalitis, 9
			immunotherapy,	syndrome	intervention		dysfunction, and 4 with brainstem			Byun, Jung-lck;			with dysautonomia, 6 with cerebellar dysfunction,
			which included	-			encephalitis, 4 patients had peripheral			Lim, Jung-Ah;			and 4 with brainstem encephalitis, 4 patients had
			intravenous				neuropathy and 1 had myelitis. 11/13			Shin, Yong-Won;			peripheral neuropathy and 1 had myelitis.
			immunoglobulin				patients had favorable responses. One			Kim, Tae-Joon;			Thirteen patients were treated with immunotherapy,
			(IVIg) (n = 12),				patient			Lee, Keon-Joo;			which included intravenous immunoglobulin (IVIg) (n
			corticosteroids				did not improve after 10 months of			Park, Kyung-II;			= 12), corticosteroids (n = 9), tacrolimus (n = 4),
			(n = 9),				immunotherapy, and in one case			Jung, Keun-Hwa;			rituximab (n = 3), cyclophosphamide (n = 1),
			tacrolimus (n =				the effect of immunotherapy could not			Jung, Ki-Young;			tocilizumab (n = 1), and mycophenolate mofetil (n =
			4), rituximab (n				be assessed because the			Lee, Sang Kun;			WR to outcome 11/13 patients had favorable
			= 3),				patient was lost to follow-up.			Chu, Kon. Non-			responses. One patient did not improve after 10
			cyclophosphami							stiff anti-			months of immunotherapy, and in one case the
			de $(n = 1)$,							amphiphysin			effect of immunotherapy could not be assessed
			tocilizumab (n =							syndrome:			because the patient was lost to follow-up.
			1), and							clinical			Evidence level 3 and with limitation to generalisability
			mycophenolate							manifestations		1	of results due retrospective nature os study, lack of
1	l			l	1							l	
			mofetil (n = 1) (and outcome		1	comparator and small sample size. There is no
										after			subgroup analysis to assess effectiveness of IVIg.
										immunotherapy.			
										J. Neuroimmunol.			
										2014;274(42036)			
										:209-214.			
3	Case series	15	IVIg along with	autoimmune	Clinical	clinial improvement in symptoms and seizure free	NMDA receptor antibody positivity was	None	-	Pandit, Awadh	none	as in	This is a prospective case series of 15 patients with
3	Case series	15	IVIg along with steroids and	autoimmune encephalitis	Clinical effectiveness	clinial improvement in symptoms and seizure free	NMDA receptor antibody positivity was found in seven (50%) patients, VGKC	None		Pandit, Awadh Kishor; Ihtisham,	none	as in primary	This is a prospective case series of 15 patients with autoimmune encephalitis presenting with status
3	Case series	15				clinial improvement in symptoms and seizure free		None			none		autoimmune encephalitis presenting with status
3	Case series	15	steroids and	encephalitis	effectiveness	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC	None		Kishor; Ihtisham, Kavish; Garg,	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline
3	Case series	15	steroids and	encephalitis presenting with status	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati,	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE)	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found.	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma,	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%)
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha;	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done.	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari.	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant improvement and didnot have further	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune encephalitis: A	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in 9 patients, of whom 3/9 revolved IVIg followed by
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune	none	primary	autoimmune encephalitis presenting with status epilepticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in
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3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune encephalitis: A potentially reversible cause of status epilepticus,	none	primary	autoimmune encephalitis presenting with status epilepiticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in 9 patients, of whom 3/9 revoived IVIg followed by streoid and in rest IVG was given along with steroids and foolowed steroid tratement.
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3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one	None		Kishor; Ihtisham, Kavish; Carg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune encephalitis: A potentially reversible cause of status epilepticus, epilepsy, and cognitive decline.	none	primary	autoimmune encephalitis presenting with status epilepiticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in 9 patients, of whom 3/9 revoived IVIg followed by streoid and in rest IVG was given along with steroids and foolowed steroid tratement. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one death.
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one	None		Kishor; Ihtisham, Kavish; Garg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune encephalitis: A potentially reversible cause of status epilepticus, epilepsy, and cognitive decline. Ann Indian Acad	none	primary	autoimmune encephalitis presenting with status epilepiticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dSDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in 9 patients, of whom 39 revider IVIg followed by streoid and in rest IVG was given along with steroids and foolowed steroid tratement. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one death. Evidence leve 3 and limited general;isability due lack of comparator, small sample size and lack of
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one	None		Kishor; Ihtisham, Kavish; Carg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari, Autoimmune encephalitis: A potentially reversible cause of status epilepticus, epilepticus, epilepsy, and cognitive decline. Ann Indian Acad Neurol	none	primary	autoimmune encephalitis presenting with status epilepiticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in 9 patients, of whom 3/9 revoived IVIg followed by streoid and in rest IVG was given along with steroids and foolowed steroid tratement. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one death.
3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one	None		Kishor; Ihtisham, Kavish; Carg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune encephalitis: A potentially reversible cause of status epilepticus, epilepsy, and cognitive decline. Ann Indian Acad Neurol 2013;16(4):577-	none	primary	autoimmune encephalitis presenting with status epilepiticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dSDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in 9 patients, of whom 39 revider IVIg followed by streoid and in rest IVG was given along with steroids and foolowed steroid tratement. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one death. Evidence leve 3 and limited general;isability due lack of comparator, small sample size and lack of
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3	Case series	15	steroids and	encephalitis presenting with status epilepticus (SE) epilepsy, and cognitive	effectiveness of the	clinial improvement in symptoms and seizure free	found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dsDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one	None		Kishor; Ihtisham, Kavish; Carg, Ajay; Gulati, Sheffali; Padma, Madakasira Vasantha; Tripathi, Manjari. Autoimmune encephalitis: A potentially reversible cause of status epilepticus, epilepsy, and cognitive decline. Ann Indian Acad Neurol 2013;16(4):577-	none	primary	autoimmune encephalitis presenting with status epilepiticus (SE), epilepsy, and cognitive decline NMDA receptor antibody positivity was found in seven (50%) patients, VGKC antibody in five (36%) patients, and two patients had anti-GAD and in another anti-dSDNA antibodies were found. None of the patients showed any evidence of malignancy in the periodic tumor screening done IVIg was given in 9 patients, of whom 39 revider IVIg followed by streoid and in rest IVG was given along with steroids and foolowed steroid tratement. 67% (10/15) had significant improvement and didnot have further seizures or relapse and there was one death. Evidence leve 3 and limited general;isability due lack of comparator, small sample size and lack of

		1				I	I=						Te
٦	Case series	14	Steroids, IVIg,	anti LG1	Clinical	clinical improvement in symptoms based on modified Rankin scale	7 patients received corticosteroids only.	brain image	Patients without	Shin, Yong-Won;	none	as in	Retrospective case series of 12 paatients with anti
		patients	plasmapheresis,	encephalitis	effectiveness	0=full recovery, 5=dead	All patients responded positively with 2	findings and	medial temporal	Lee, Soon-Tae; Shin, Jung-Won;	reported	primary	LG1 encephalitis. 7 patients received corticosteroids
		with anti LG1	Rituximab, tacrolimus		of the intervention		showing partial response who improved with addition of IVIg. Five patients	treatment outcome	hypermetabolism on FDG- PET	Moon, Jangsup;		outcome	only. All patients responded positively with 2 showing partial response who improved with addition of IVIg.
		encepha	tacrominus		intervention		received combined therapy 4/5 has	outcome	scans were	Lim, Jung-Ah;			Five patients received combined therapy 4/5 has
		litis (12					complete recovery and no relapse. The		associated with	Byun, Jung-Ick;			complete recovery and no relapse. The difference
		included					difference between two groups was not			Kim, Tae-Joon;			between two groups was not statistically different
		in the					statistically different (p=0.271).		lower mRS	Lee, Keon-Joo;			(p=0.271). Treatment initiated within 1month of
		final					Treatment initiated		(TC=-0.480, p=0.02;				symptom onset seemed to be associated with
		analysis)					within 1month of symptom onset			Park, Kyung-II;			clinical outcome p=0.058; Fig. 2A). However,
		analysis)					seemed to be associated with clinical		with mRS=0 was	Jung, Keun-Hwa;			outcome measured by achievement of mRS=0
							outcome p=0.058; Fig. 2A). However,		also related to	Lee, Sang Kun;			failed to show statistical significance (p=0.221).
							outcome measured		medial temporal	Chu. Kon. VGKC			Level of evidence 3 due to lack of comparator, lack
							by achievement of mRS=0 failed to		hypermetabolism	complex/LGI1-			of sample size and retrospective nature of study
							show statistical significance (p=0.221)		$(\tau b = -0.535,$	antibody			causing bias.
							, ,		p=0.02; Fig. 2D).	encephalitis:			9
									Among patients	clinical			This study is also included in review by Nosadini et
									with medial	manifestations			al, 2015.
									temporal lesions, a	and response to			
									unilateral lesion was	immunotherapy.			
									associated with a	J. Neuroimmunol.			
									favorable outcome	2013;265(42036)			
									(tc=0.898, p b	:75-81.			
									0.001; and				
1		1		1	Ì			1	achievement of				
					1				mRS = 0 (tb =				
									0.750, p = 0.001. In				
					1				contrast, basal				
									ganglia				
									hypermetabolism				
									had no association				
									with mRS (TC =				
									-0.040, p = 0.907)				
									or achievement of				
									mRS = 0 (tb =				
									-0.089, p = 0.774),				
									and medial				
									temporal lesions on				
					l l				MRI also failed to				
1									show association				
									show association with mRS				
									show association with mRS (rc=0.111, p=0.726)				
									show association with mRS (τc=0.111, p=0.726) or recovery to mRS				
									show association with mRS (τc=0.111, p=0.726) or recovery to mRS = 0 (τb = -0.169, p				
									show association with mRS (τc=0.111, p=0.726) or recovery to mRS = 0 (τb = -0.169, p = 0.552). None of				
									show association with mRS (rc=0.111, p=0.726) or recovery to mRS = 0 (rb = -0.169, p = 0.552). None of these image				
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2	Capa sorina	11	eteroids and	NIMDA recente	Clinical	olinical response to treatment	based an abstract authors most 500°	2002	show association with mRS (rc=0.111, p=0.726) or recovery to mRS = 0 (rb = -0.169, p = 0.552). None of these image parameters were	Chakraharty	2002	acin	Outcopoditio caso parios of 41 patients wath NPMA
3	Case series	11	steroids and	NMDA recepto	Clinical	clinical response to treatment	based on abstract authors report 58%	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Chakrabarty,	none	as in	Retospective case series of 11 patients woth NDMA
3	Case series	11	steroids and intervention	NMDA recepto encephalitis	effectiveness	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop;	none reported	primary	repetor encephalitis. Authors report 58% responded
3	Case series	11			effectiveness of the	clinical response to treatment		none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari;			repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and
3	Case series	11			effectiveness	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali;		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan,		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha;		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha,		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D-		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D- aspartate		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D- aspartate (NMDA) receptor		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D- aspartate (NMDA) receptor encephalitis:		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari, Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D-aspartate (NMIDA) receptor encephalitis: experience of a		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari, Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh, Pediatric anti-N-methyl-D- aspartate (MMDA) receptor encephalitis: experience of a tertiary care		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop; Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh, Pediatric anti-N-methyl-D- sapartate (NMIDA) receptor encephalitis: experience of a tertiary care teaching center		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari, Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh, Pediatric anti-N-methyl-D- aspartate (MMDA) receptor encephalitis: experience of a tertiary care		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop,' Tripathi, Manjari; Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D-aspartate (NMDA) receptor encephalitis: experience of a tertiary care teaching center from north India.		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari, Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh, Pediatric anti-N-methyl-D- aspartate (NMDA) receptor encephalitis: experience of a tertiary care teaching center from north India. J. Child Neurol.		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari, Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D- aspartate (NMDA) receptor encephalitis: experience of a tertiary care teaching center from north India. J. Child Neurol. 2014;29(11):145		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari, Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D- aspartate (NMDA) receptor encephalitis: experience of a tertiary care teaching center from north India. J. Child Neurol. 2014;29(11):145		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup
3	Case series	11			effectiveness of the	clinical response to treatment	showed significant response to steroids	none	show association with mRS (τ c=0.111, p=0.726) or recovery to mRS = 0 (τ b = -0.169, p = 0.552). None of these image parameters were related to	Biswaroop, Tripathi, Manjari, Gulati, Sheffali; Yoganathan, Sangeetha; Pandit, Awadh Kishore; Sinha, Aditi; Rathi, Bhim Singh. Pediatric anti-N-methyl-D- aspartate (NMDA) receptor encephalitis: experience of a tertiary care teaching center from north India. J. Child Neurol. 2014;29(11):145		primary	repetor encephalitis. Authors report 58% responded to steroid and IVIg treatment. Evidence level 3 and with limitation to generalisability of results due retrospective nature os study, lack of comparator and small sample size. There is no subgroup

3	Case series	13	immunomodulat	Hashimoto	Clinical	improvement in neurological symptoms including stroke like symptoms,	Patient received steroids in the acute	none based on	- Olme	nez, Inan;	none based	as in	this is retrospective case over a 13 yrs period
			or treatments	encephalopathy	effectiveness	seizures, cognitive function	phase for 12 of 13 patients with rapid	abstract	Mose	ses, Harold;	on abstract	primary	consisting of 13 patients with Hashimoto
			including		of the		improvement in symptoms. Maintenance		Srira	ram,		outcome	encephalitis/ Patients received steriods and primary
			IVIg.Treatment		intervention		therapy was rituximab in 7 patients,		Subr	oramaniam;		measure	treatment with rapid in treatments. Patients also
			consisted of				intravenous immunoglobulin (IVIg) in 7,		Kirsh	shner,			received other immune treatments including IVIg in 7
			steroids in the				azathioprine in 4, mycophenolate mofetil		How	ward;			patients/. There is no subgroup analysis by IVIg
			acute phase				in 3, and methotrexate in 1 (some		Lagr	grange, Andre			group but authors report all 13 responded to
			Maintenance				patients received sequential therapy with		H.; F	Pawate,			treatment.
			therapy was				different agents). There was complete or			dharama.			Level of evidence is 3 and poor generalisability to
			rituximab in 7				near complete resolution of symptoms in			gnostic and			lack of comparator, patulent reslection methods and
			patients,				12 of the 13 patients			rapeutic			lack of subgroup analysis.
			intravenous							ects of			
			immunoglobulin							shimoto's			
			(IVIg) in 7,							ephalopathy.			
			azathioprine in							Neurol. Sci.			
			4,							13;331(42036)			
			mycophenolate						:67-7	-71.			
			mofetil in 3, and										
			methotrexate in										
			1 (some patients										
			received										
			sequential										
			therapy with										
1			different		1								
1			agents).		1								
1			agomo,		1								

3	Case series		IVIg (revie	Morvans	Clinical	Recovered- sponteneous or with treatment and death	15 patients (56%) had concurrent	none	-	Lee, Will; Day,	none	as inprimary	
		fromauth	includes an	Fibrillary \chorea			malignancy, 15 (79%) were positive for			Timothy J.;	reported	outcome	Movac fibrillary chorea. 9 (41%) patients had IVIg in
		ors	analysis of other		of the		VGKC-Ab. Associated autoimmunity			Williams, David			combination with other therapy. 3 patients had IVIg
		clinica	treaments		intervention		was reported in 11 patients (41%) with			R., Clinical,			for maintance. There is subgroup analysis to assess
		and 26	including				three having auto-antibodies only, while			laboratory and			the effectivenss of IVIg but the overall results
1 1		from	steriods, PLEX,	1	1		eight manifested clinical autoimmune	l l		electrophysiologi	1	1	showed 86% responded favorably with 3 patients
			and immune				diseases. Auto-antibodies to the			cal features of			dying.
		serach)	therpaies				acetylcholine receptor (n = 6), voltage-			Morvan's fibrillary			This is evedence level 3 study with limitaions to
			including				gated calcium channel (n = 2), glutamic			chorea. J Clin			generalisability of the study due lack of comaparator
			azthiprine,				acid decarboxylase (n = 1), phospholipid			Neurosci			patients selection and lack of subgroup analysis
			rituximab				(n = 1) and muscle specific kinase (n =			2013;20(9):1246-			
							1) were identified. The most common			1249.			
							associated autoimmune disorder was			12 10.			
							myasthenia gravis (n = 5) while others						
							included thyroid disease (n = 1),						
							psoriasis (n = 1), myositis (n = 1) and						
							anti-phospholipid syndrome (n=1). Four						
							patients (36%) had clustering of auto-						
							antibodies or autoimmune disorders that						
			l	l	1		are not classically related. Mortality			l	1		
			1	1	1		amongst all patients was 22% (6/27)	1		1	1	1	I
			l	l	1		and the prevalence of malignancy was			l	1		
			l	l	1		comparable between those who died			l	1		
			1	1	1		(50%) and those who recovered (57%).	1		1	1	1	I
			1	1	1		All patients with malignancies underwent	1		1	1	1	I
			l	l	1					l	1		
			1	1	1		surgery. Of those who recovered, two	l l		1	1	1	I
			1	1	1		(9%) showed spontaneous	1		1	1	1	I
							improvement. Immunotherapy was						
							instituted in 22 patients with 19 (86%)						
							responding favourably and three (14%)						
							dying. The most common first line						
							treatment (monotherapy or in						
							combination) was plasma exchange						
							(PEx) (55%) followed by intravenous						
							immunoglobulin (IVIg) (41%) and						
							corticosteroids (32%).IVIg. Four patients						
1							(18%) required second line treatment						
							after failing IVIg (n = 2), IVIg/						
							after failing IVIg (n = 2), IVIg/						
							after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and Ex/IVIg (n = 1). One patient was treated with rituximab						
							after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and Ex/IVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14						
							after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and EX/IVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance						
							after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and $ExIVIg$ (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in						
							after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and $ExIVIg$ (n = 1). One patient was treated with ritus/mab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids						
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							after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%);						
							after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%).						
3	Case series	10	steroids and	Non viral limbic	Clinical	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%).	none	-	Chou, IJun;	none	as in	This cross-sectional study describes the clinical
3	Case series	10	steroids and IVIg	encephalitis	effectiveness	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%).	none	-	Wang, Huei-	none reported in	as in primary	This cross-sectional study describes the clinical manifestation and the serological evidence of the
3	Case series	10		encephalitis	effectiveness	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%).	none	-				
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied.	none	-	Wang, Huei- Shyong; Lin,	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese
3	Case series	10		encephalitis	effectiveness	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%), azatibioprine (14%); PEx (7%) or cyclophosphamide (7%).	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo,	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azatibnoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin,	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExiVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou,	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), azatibnoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung,	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExiVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou,	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azatihoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng;	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExiVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent	none	·	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng-	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun-	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephaltis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExiVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%); IVIg (21%); azathioprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent	none	ī	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun- Tong; CHEESE	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun- Tong; CHEESE Study Group.	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephaltis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun- Tong; CHEESE Study Group. Limbic	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun- Tong; CHEESE Study Group.	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun- Tong; CHEESE Study Group. Limbic encephalitis in	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun- Tong; CHEESE Study Group. Limbic encephalitis in Taiwanese	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hais, Meng- Ying; Lin, Yun- Tong; CHEESE Study Group. Limbic encephalitis in Taiwanese children and	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng-Ying; Lin, Yun-Tong; CHEESE Study Group. Limbic encephalitis in Taiwanese children and adolescence: a	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuo, Chang-Fu; Lin, Khou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun-Tong; CHESS Study Group. Limbic encephalitis in Taiwanese children and adolescence: a single center	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Als
3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng-Ying; Lin, Yun-Tong; CHEESE Study Group. Limbic encephalitis in Taiwanese children and adolescence: a	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Alsr
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3	Case series	10		encephalitis	effectiveness of the	clincial improvement	after failing IVIg (n = 2), IVIg/ corticosteroid (n = 1) and ExIVIg (n = 1). One patient was treated with rituximab after failing cyclosporine. A further 14 patients required maintenance immunotherapy (monotherapy or in combination), using corticosteroids (57%): IVIg (21%), szatibinoprine (14%); PEx (7%) or cyclophosphamide (7%). 10 children with serological evidence of the presence of neuronal antibodies causing limbic encephalitis were studied. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90%	none	-	Wang, Huei- Shyong; Lin, Jainn-Jim; Kuo, Chang-Fu; Lin, Kuang-Lin; Chou, Min-Liang; Hung, Po-Cheng; Hsieh, Meng- Ying; Lin, Yun- Tong; CHEESE Study Group. Limbic encephalitis in Taiwanese children and adolescence: a sig	reported in	primary	manifestation and the serological evidence of the presence of neuronal antibodies in 10 Taiwanese children with limbic encephalitis. All of the 10 enrolled patients had acute onset of fever and rapid clinical deterioration. Six patients were treated with methylprednisolone pulse therapy or intravenous immunoglobulin (IVIg) at the acute stage. All had persistent neuropsychiatric symptoms and 90% developed refractory epilepsy. Evidence leve 3. due retrospective case selection, small sample size. Alsr

				clinical improvement measured modified Rankin scale	577 patients (1-85 years, median 21)	none separately		tulaer, Maarten	reported as		This is a large multinational, multicentre prospective
1	IVIg,	encephalitis	effectiveness		were studied, 212 were children (<18	reported			in primary	primary	study of 501 pateints with anti NMDAR antibody
	pasmapheresis)		of the		years). Treatment effects and outcome				outcome	outcome	encephalitis. 472 (94%) underwent first-line
	second line,		intervention		were assessable in 501 (median follow-		Ga	abilondo, lñigo;	(relapse)		immunotherapy or tumor removal, resulting in
	(rituximab,				up 24 months): 472 (94%) underwent		Arr	mangué,			improvement within four weeks in 251 (53%). Of 2
	cyclophosphami				first-line immunotherapy or tumor		Th	naís; Glaser,			patients who failed first-line therapy, 125 (57%)
	de) and tumour				removal, resulting in improvement within		Ca	arol; lizuka,			received second-line immunotherapy resulting in
	removal				four weeks in 251 (53%). Of 221		Tai	akahiro; Honig,			better outcome than those who did not (OR 2-69,
					patients who failed first-line therapy, 125		La	wrence S.;			1-24-5-80, p=0-012). During the first 24 months,
					(57%) received second-line		Be	enseler,			394/501 reached good outcome (mRS 0-2; media
					immunotherapy resulting in better		Su	usanne M.;			6 months), and 30 died. At 24 month follow-up
					outcome than those who did not (OR		Ka	awachi, Izumi;			204/252 (81%) had good outcome
					2-69, CI 1-24-5-80, p=0-012). During the		Ma	artinez-			Predictors of good outcome were early treatment
					first 24 months, 394/501 reached good		He	ernandez,			(OR 0.62, CI 0.50-0.76, p<0.0001) and lack of IC
					outcome (mRS 0-2; median 6 months),		Eu	ugenia; Aguilar,			admission (OR 0.12, CI 0.06-0.22,p<0.0001). 45
					and 30 died. At 24 month follow-up		Es	sther; Gresa-			patients had one or multiple relapses (representing
					204/252 (81%) had good outcome		Arr	ribas, Núria;			12% risk within 2 years); 46/69 (67%) relapses w
					Predictors of good outcome were early		Ry	/an-Florance,			milder than previous episodes (p<0.0001).
					treatment (OR 0-62, CI 0-50-0-76,			cole; Torrents,			Comparing children ,18 yrs with adults, the time
					p<0.0001) and lack of ICU admission		Ab	oiguei; Saiz,			between symptom onset and initiation of treatme
					(OR 0.12, CI 0.06-0.22,p<0.0001). 45		Alb	bert;			was shorter in children (21 versus 28 days, p=0-0
					patients had one or multiple relapses		Ro	osenfeld, Myrna			Overall the outcome was similar to that of adults
					(representing a 12% risk within 2 years);		R.;	; Balice-			(p=0.92,).
					46/69 (67%) relapses were milder than		Go	ordon, Rita;			Level of evidence=2 This is a well designed stu
					previous episodes (p<0.0001)		Gr	raus, Francesc;			with primary objective, patient selection methods
					Comparing children ,18 yrs with adults,		Da	almau, Josep.			and measurement of outcome using mRsclae. 1
					the time between symptom onset and		Tre	eatment and			study also considted of larger sample size and t
					initiation of treatment was shorter in		pro	ognostic			outcome reported for various subgroups includir
					children (21 versus 28 days, p=0.007,		fac	ctors for long-			type of treatment and age grooup. TThe only
					Overall the outcome was similar to that		ten	rm outcome in			limitation from a PICO perspective are that resu
					of adults (p=0.92,)		par	tients with anti-			are not valaibal; by IVIg treatment group so not
					4		NN	MDA receptor			possible to answer PICO question 1.
							en	cephalitis: an			, ,
							obs	servational			This is study was also included in the study of
							col	hort study.			Nosadini et al, 2015.
							Lai	ncet Neurol			·
							20	13;12(2):157-			
							16				
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3	Case series	20	streroids, IVIg,	anti NMDAR	Clinical	clinical improvement measured using Pediatric Cerebral Performance	During the first episode of encephalitis,	None reported		rmangue,	non	0	Thi is a retrospective caase series of 20 children with
			plasmapheresis	encephalitis	effectiveness	Category Scale" (PCPC, relapse	19 (95%) patients received first-line			haís; Titulaer,	eseparately		anti NDMAr encepahilitis. During the first episode of
					of the		immunotherapies (one patient was only		M	Maarten J.;	reported		encephalitis, 19 (95%) patients received first-line
					intervention		treated at third relapse). All patients		M	Málaga, Ignacio;			immunotherapies (one patient was only treated at
							received at least a short course of high-		В	Bataller, Luis;			third relapse). All patients received at least a short
							dose steroids (median 1, range 1-3		G	Sabilondo, Iñigo;			course of high-dose steroids (median 1, range 1-3
							courses), followed in 13 patients by oral			Graus, Francesc;			courses), followed in 13 patients by oral steroid
							steroid tapering for a median of 12		D	almau, Josep;			tapering for a median of 12 weeks (range 3-47). In
							weeks (range 3-47). In addition, 14		S	Spanish Anti-N-			addition, 14 patients received intravenous
							patients received intravenous		m	nethyl-D-			immunoglobulin (IVIg; median 2 cycles, range 1–12)
							immunoglobulin (IVIg; median 2 cycles,		A	spartate			and one patient had plasmapheresis. At last follow
							range 1-12) and one patient had			Receptor			up all patients had received immunotherapy: 20 had
							plasmapheresis. At last follow up all			NMDAR)			first-line therapies (steroids, IVIg and/or plasma
							patients had received immunotherapy:			ncephalitis			exchange), and 7 (35 %) second-line therapies
							20 had first-line therapies (steroids, IVIg			Vork Group.			(rituximab alone or combined with
							and/or plasma exchange), and 7 (35 %)			ediatric anti-N-			cyclophosphamide).
							second-line therapies (rituximab alone or	•	m	nethyl-D-			After a median follow up of 17.5 months (4-149), 17
							combined with cyclophosphamide).		a	spartate			(85%) patients had substantial improvement (PCPC
							After a median follow up of 17.5 months		re	eceptor			of 1 or 2: 60% complete recovery and 25% minimal
							(4-149), 17 (85%) patients had			ncephalitis-			residual deficits), 2 (10%) moderate or severe
							substantial improvement (PCPC of 1 or		cl	linical analysis			disability (PCPC of 3 or 4) and 1 died. The two
							2: 60% complete recovery and 25%		a	nd novel			patients with moderate or severe disabilities at follow-
							minimal residual deficits), 2 (10%)		fir	ndings in a			up of 4 and 9 months were showing improvement. 3
							moderate or severe disability (PCPC of		Se	eries of 20			patients developed relapse.
							3 or 4) and 1 died. The two patients with			atients. J.			Evidence level 3- due retspective case selection,
							moderate or severe disabilities at follow-			Pediatr.			lack of comparator and small sample size. The
							up of 4 and 9 months were showing			013;162(4):850-			results are also not available by IVIg so not possible
							improvement. 3 patients developed		8	56.e2.			to answer PICO question.
							relapse.						
													This study is study included in review by Nosadini et
													al 2015.
					l l								
2.	Casa sorios	02	IVIa	Children with	Clinical	mortality Heart Ejection fraction	Mortality was lower in the IV/Ig group In -	nono		hatt Girich	none based	ae in	Their is a well designed non randomicad prospective
2-	Case series	83	IVIg 400ma/ka/day	Children with	Clinical	mortality, Heart Ejection fraction	Mortality was lower in the IVIg group [n =	none		Shatt, Girish	none based	as in	Theis is a well designed non randomised prospective
2-	Case series	83	400mg/kg/day	acute	effectiveness	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the	none	С	handra; Sankar,	none based on abstract	primary	study with a comparator. A total of 83 consecutive
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group [n = 13 (22.8 %)]	none	C	handra; Sankar, huma;			study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were
2-	Case series	83	400mg/kg/day	acute encephalitis complicated by	effectiveness	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group [n = 13 (22.8 %)] with a relative risk of 0.17 (95 % CI =	none	C Ji K	Chandra; Sankar, huma; Kushwaha, K. P		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group [n = 13 (22.8 %)] with a relative risk of 0.17 (95 % CI = 0.02, 1.22). The difference in mortality	none	C Ji K U	Chandra; Sankar, huma; Kushwaha, K. P Use of		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis complicated by	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group [n = 13 (22.8 %)] with a relative risk of 0.17 (95 % CI = 0.02, 1.22). The difference in mortality reached borderline significance (p =	none	C Ji K U in	Chandra; Sankar, huma; Kushwaha, K. P Jse of htravenous		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram findings. Patients were allocated to the two groups
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis complicated by	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group [n = 13 (22.8 %)] with a relative risk of 0.17 (95 % Cl = 0.02, 1.22). The difference in mortality reached borderline significance (p = 0.05). At discharge, mean (SD) ejection	none	C Ji K U in in	Chandra; Sankar, huma; Kushwaha, K. P Jse of htravenous mmunoglobulin		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram findings. Patients were allocated to the two groups based on the days of the week: Those presenting on
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis complicated by	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group $[n=13\ (22.8\ \%)]$ with a relative risk of 0.17 $(95\ \%\ Cl=0.02,1.22)$. The difference in mortality reached borderline significance $(p=0.05)$. At discharge, mean (SD) ejection fraction improved from 32.8 % (6.31 %)	none	C JI K U in in	Chandra; Sankar, huma; Kushwaha, K. P Jse of htravenous mmunoglobulin ompared with		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram findings. Patients were allocated to the two groups based on the days of the week: Those presenting on Monday and Friday were allocated to IVIg treatment
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis complicated by	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group $[n=13 (22.8 \%)]$ with a relative risk of 0.17 (95 % Cl = 0.02, 1.22). The difference in mortality reached borderline significance (p = 0.05). At discharge, mean (SD) ejection fraction improved from 32.8 % (6.31 %) to 49.5 % (9.04 %) in group I patients,	none	C Ji K U in in c st	Chandra; Sankar, huma; Kushwaha, K. P Use of htravenous mmunoglobulin ompared with tandard therapy		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram findings. Patients were allocated to the two groups based on the days of the week: Those presenting on Monday and Friday were allocated to IVIg treatment (group I), and those presenting on the other days of
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis complicated by	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group [n = 13 (22.8 %)] with a relative risk of 0.17 (95 % CI = 0.02, 1.22). The difference in mortality reached borderline significance (p = 0.05). At discharge, mean (SD) ejection fraction improved from 32.8 % (6.31 %) to 49.5 % (9.04 %) in group I patients, which was significantly greater than that	none	C JI K U in in c s st	Chandra; Sankar, huma; Kushwaha, K. P Ise of htravenous mmunoglobulin ompared with tandard therapy is associated with		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram findings. Patients were allocated to the two groups based on the days of the week: Those presenting on Monday and Friday were allocated to IVIg treatment (group I), and those presenting on the other days of the week to standard care (group II). Group I (n =
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis complicated by	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group $[n=13 (22.8 \%)]$ with a relative risk of 0.17 (95 % Cl = 0.02, 1.22). The difference in mortality reached borderline significance (p = 0.05). At discharge, mean (SD) ejection fraction improved from 32.8 % (6.31 %) to 49.5 % (9.04 %) in group I patients,	none	C JI K U in or st is is	Chandra; Sankar, huma; Kushwaha, K. P Jse of htravenous mmunoglobulin ompared with tandard therapy is associated with improved clinical		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram findings. Patients were allocated to the two groups based on the days of the week: Those presenting on Monday and Friday were allocated to IVIg treatment (group I), and those presenting on the other days of the week to standard care (group II). Group I (n = 26) patients received IVIg at a dose of 400
2-	Case series	83	400mg/kg/day for 5 days plus	acute encephalitis complicated by	effectiveness of the	mortality, Heart Ejection fraction	1 (3.8 %)] patients compared with the standard care group [n = 13 (22.8 %)] with a relative risk of 0.17 (95 % CI = 0.02, 1.22). The difference in mortality reached borderline significance (p = 0.05). At discharge, mean (SD) ejection fraction improved from 32.8 % (6.31 %) to 49.5 % (9.04 %) in group I patients, which was significantly greater than that	none	C Ji K U in in c st is is in	Chandra; Sankar, huma; Kushwaha, K. P Jse of Intravenous inmunoglobulin ompared with tandard therapy is associated with improved clinical utcomes in		primary	study with a comparator. A total of 83 consecutive children with AES complicated by myocarditis were enrolled. Diagnosis of myocarditis was based on clinical, electrocardiogram, and echocardiogram findings. Patients were allocated to the two groups based on the days of the week: Those presenting on Monday and Friday were allocated to IVIg treatment (group I), and those presenting on the other days of the week to standard care (group II). Group I (n = 26) patients received IVIg at a dose of 400 mg/kg/day for 5 days in addition to standard care. A
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3	Case series	10	variuos	Children with	Clinical		Authors report only two patients showed	none	Haberlandt, E.;		This is retrospective case study of 10 children with
			immunomodulat		effectiveness		clinical improvement		Bast, T.; Ebner,		limbic encephalitis with with different type of
				encephalitis	of the				A.; Holthausen,		antibodies. All patients were treated with steroid and
			steroid and IVIg.		intervention				H.; Kluger, G.;		4 patients received IVIg. Only 2/10 patients showed
			Nine patients						Kravljanac, R.;		clinical improvement at the end of 24 months and
			received						Kröll-Seger, J.;		neither of them had received IVIg. This is a level 3
			corticosteroids						Kurlemann, G.;		evidence study with limited generalisability due to
			(seven						Makowski, C.;		small sample size, lack of comparartor and lack of
			intravenously,						Rostasy, K.;		subgroup analysis.
			two orally),						Tuschen-		
			intravenous						Hofstätter, E.;		
			immunoglobulin						Weber, G.;		
			s (IVIg) or						Vincent, A.; Bien,		
			combinations						C. G Limbic		
			of both Median	1					encephalitis in		
			time from						children and		
			disease						adolescents.		
			manifestation						Arch. Dis. Child.		
			until treatment						2011;96(2):186-		
			was 4 months						191.		
			(0-18 months)								
			for								1
			steroids and 18								
			months (0.5-33								
			months) for								
			IVIg. The								
			median								
			doses were 55								
			mg/kg								
			methylprednisol								1
			one-equivalent								
			(range								
			15-410 mg/kg)								
			and 2 g/kg IVIg								
			(range 2-2.8								
1			g/kg).								
											1
1											1
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Appendix Two

Literature search terms

Assumptions / limits applied	to search:
Original search terms:	n/a
Updated search terms - Population	Population part 1: (encephalitis OR encephalopathy) AND (antibody OR autoimmune OR autoantibody OR antibodies OR autoantibodies) OR population part 2: (NMDA OR NMDAR OR Caspr2 OR LGI1 OR VGKC OR hashimoto OR MOG) AND (encephalitis OR encephalopathy OR antibody OR autoimmune OR autoantibody OR autoantibodies)
Updated search terms - Intervention	intravenous normal human immunoglobulin OR IVIG OR intravenous immunoglobulin OR intravenous immune globulin OR immune globulin intravenous OR IGIV OR intravenous immunoglobulins OR intravenous normal human immunoglobulins OR intravenous immune globulins
Updated search terms - Comparator	n/a
Updated search terms - Outcome	n/a

	Conoral inclusion evitoria
Inclusion criteria	In order of decreasing priority, articles will be selected based on the following criteria. 1. All relevant systematic reviews and meta-analysis in the last 5 years and those in 5-10 years period which are still relevant (e.g. no further updated systematic review available) 2. All relevant RCTs and those in the 5-10 years period which are still relevant (e.g. not superseded by a next phase of the trial/ the RCT is one of the few or only high quality clinical trials available) >>>> If studies included reaches 30, inclusion stops here 3. All relevant case control and cohort studies, that qualify after exclusion criteria >>>> If studies included reaches 30, inclusion stops here 4. All relevant non analytical studies (case series/ reports etc.) that qualify after exclusion criteria >>>> If studies included reaches 30, inclusion stops here Specific inclusion criteria n/a
Exclusion criteria	General exclusion criteria Studies with the following characteristics will be excluded: 1. Does not answer a PICO research question 2. Comparator differs from the PICO 3. < 50 subjects (where studies with >50 subjects exist) 4. No relevant outcomes 5. Incorrect study type 6. Inclusion of outcomes for only one surgeon/doctor or only one clinical site (where studies with > one surgeon/doctor or one clinical site exist) 7. Narrative / non-systematic reviews (relevant referenced studies to be included) Specific exclusion criteria n/a