Management of Guillain-Barré Syndrome: A Systematic Review of Clinical Practice Guidelines

HSD4

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CONCLUSIONS

In the current review, IVIg and TPE emerged as the most effective and endorsed therapies for the management of GBS worldwide. Whilst IVIg and TPE were considered equally effective, there seemed to be a lack of globally recommended treatment alternatives for patients with poor outcomes to these therapies. Addressing these research gaps will aid in the development of informed decision-making and optimizing patient care strategies

INTRODUCTION

- Guillain-Barré syndrome (GBS) is a rare, potentially fatal, immune-mediated neuropathy, usually triggered by infections that results in autoimmune destruction of the peripheral nerves[1]
- With a global incidence of approximately 1-2 per 100,000 people per year, GBS can affect people of any age with incidence increasing linearly with age[2]
- The management of GBS is complicated due to the heterogeneity in clinical presentation, the absence of highly sensitive diagnostic tools, and the variability in the course of disease progression[1]
- The outbreak of infectious illnesses can lead to an increased incidence of GBS emphasising the importance of devising globally applicable clinical guidelines for managing GBS[1]

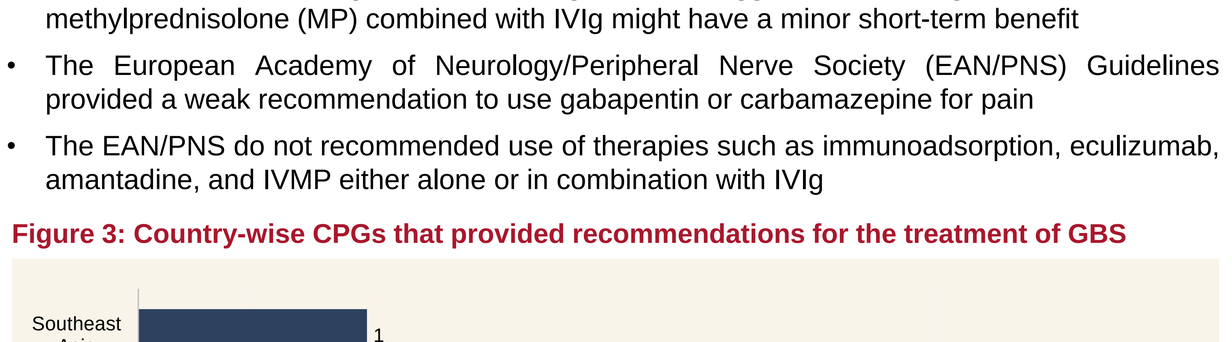
OBJECTIVE

- This systematic literature review (SLR) aimed to identify treatment recommendations based on clinical practice guidelines (CPGs) for GBS

METHODS

- This review followed the standard methodology for conducting an SLR as per the guidelines provided by the National Institute for Health and Care Excellence
- A systematic search was performed across key biomedical databases (EMBASE® and MEDLINE®) from database inception until June 2024 to identify relevant English language publications providing CPGs for GBS patients. The prespecified eligibility criteria are presented in **Figure 1**
- Each publication was reviewed by two independent reviewers, and disagreements were arbitrated by a third reviewer

Figure 1: Prespecified PICOS eligibility criteria for the selection of evidence



RESULTS

- Of the 458 publications retrieved, the SLR identified nine CPGs from 16 publications offering valuable information on the management of GBS (**Figure 2**)
- The majority of the studies (n=8) were published in peer-reviewed journals. Three CPGs each were identified from the USA and Europe followed by one each for Australia, Canada, and Southeast Asia (**Figure 3**). A list of the recommending bodies is provided in **Figure 4**
- Across all the CPGs, intravenous immunoglobulin (IVIg) at 0.4 g/kg/day for 5 days and therapeutic plasma exchange (TPE) emerged as effective first-line treatments, with IVIg often preferred due to its lower side effect profile
- Sequential therapy with TPE followed by IVIg was generally not recommended, whereas, for treating relapse, a second course of IVIg treatment was preferred
- Corticosteroids were largely discouraged in managing GBS; however, the European Federation of Neurological Societies guidance suggested that high-dose intravenous methylprednisolone (MP) combined with IVIg might have a minor short-term benefit
- The European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) Guidelines provided a weak recommendation to use gabapentin or carbamazepine for pain
- The EAN/PNS do not recommended use of therapies such as immunoadsorption, eculizumab, amantadine, and IVMP either alone or in combination with IVIg

Figure 3: Country-wise CPGs that provided recommendations for the treatment of GBS

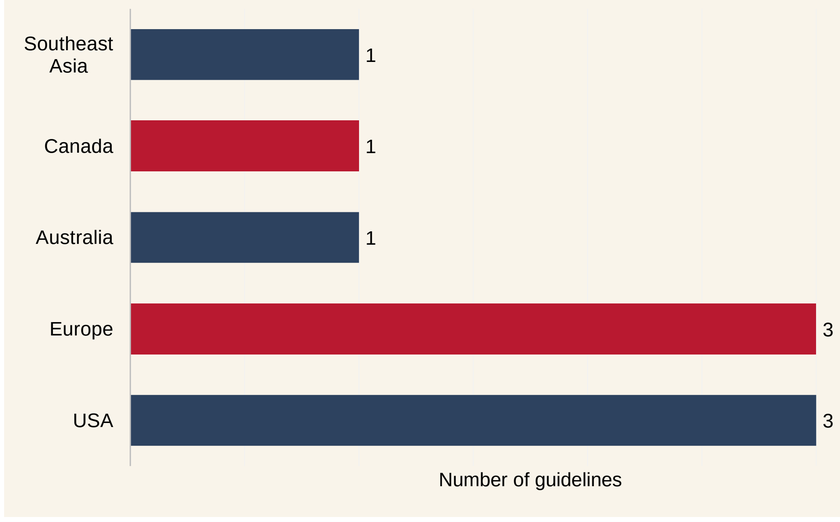
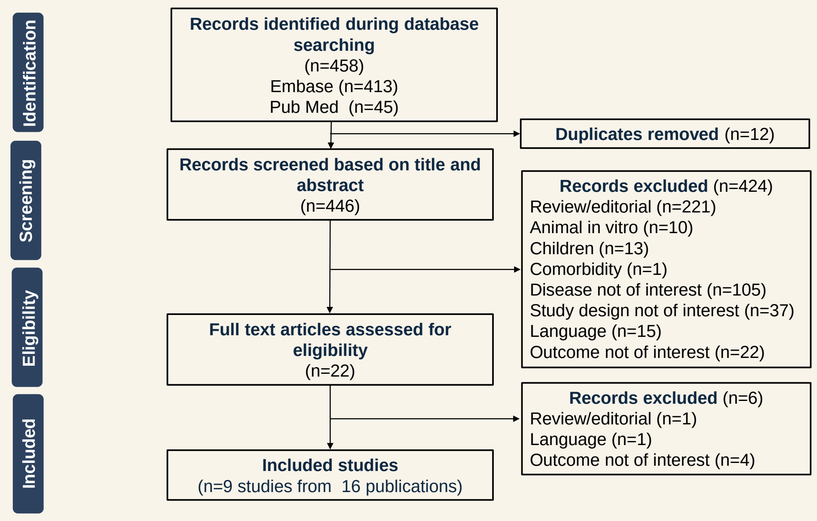
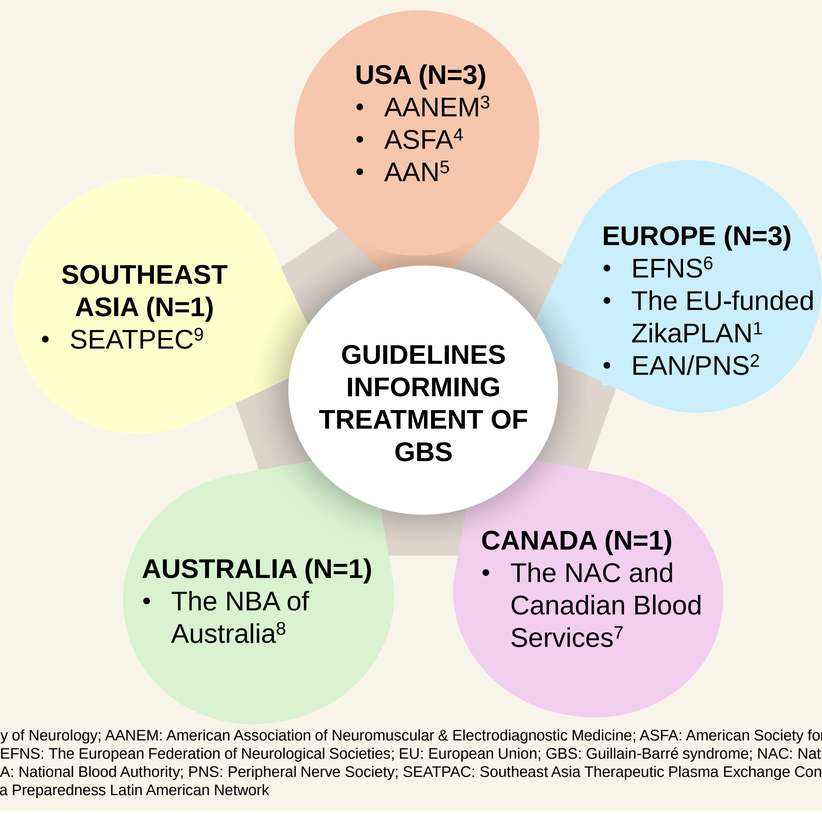


Figure 2: PRISMA diagram for the screening process



Embase: Excerpta Medica database; PRISMA: Preferred Reporting Items for Systematic Reviews

Figure 4: List of recommending bodies that provided treatment guidance for GBS



AAN: American Academy of Neurology; AANEM: American Association of Neuromuscular & Electrodiagnostic Medicine; ASFA: American Society for Apheresis; EAN: European Academy of Neurology; EFNS: The European Federation of Neurological Societies; EU: European Union; GBS: Guillain-Barré syndrome; NAC: National Advisory Committee on Blood and Blood Products; NBA: National Blood Authority; PNS: Peripheral Nerve Society; SEATPAC: Southeast Asia Therapeutic Plasma Exchange Consortium; USA: United States of America; ZikaPLAN: Zika Preparedness Latin American Network

Table 1: Country-wise treatment recommendations for treatment of GBS

Treatment								Recommending body
IVIg	TPE	CS	High dose IV MP with IVIg	Gabapentin or carbamazepine	IA	TPE followed by IVIg or IA followed by IVIg	Others^	
USA								
✓	✓	--	--	--	--	--	--	AANEM[3]
--	✓	--	--	--	--	--	--	ASFA[4]
✓	✓	✗	--	--	✗	✗	--	AAN[5]
Europe								
✓	✓	--	✓*	--	--	--	--	EFNS[6]
✓	✓	✗	✗	--	--	✗	--	The EU-finded ZikaPLAN[1]
✓	✓	✗	✗	✓*	✗	✗	✗	EAN/PNS[2]
Canada								
✓	--	--	--	--	--	--	--	The NAC and Canadian Blood Services[7]
Australia								
✓	--	--	--	--	--	--	--	The NBA of Australia[8]
Asia-Pacific								
✓	✓	✗	✗	-	--	----	--	SEATPEC[9]

*: Weak recommendation; ^Others: Alemtuzumab, eculizumab, brain derived neurotrophic factor, CSF filtration, cyclophosphamide, IFNβ-1a, muronomab-CD3, mycophenolate mofetil or tripterygium polyglycoside·AAN: American Academy of Neurology; AANEM: American Association of Neuromuscular & Electrodiagnostic Medicine; ASFA: American Society for Apheresis; CS: Corticosteroids; EAN: European Academy of Neurology; EFNS: The European Federation of Neurological Societies; EU: European Union; GBS: Guillain-Barré syndrome; IA: Immunoadsorption; IV: Intravenous; IVIg: IV Immunoglobulin; MP: Methylprednisolone; NAC: National Advisory Committee on Blood and Blood Products; NBA: National Blood Authority; PNS: Peripheral Nerve Society; SEATPAC: Southeast Asia Therapeutic Plasma Exchange Consortium; TPE: Therapeutic Plasma Exchange; USA: United States of America; ZikaPLAN: Zika Preparedness Latin American Network.

Table 2: Summary of treatment recommendations

Treatment Recommendation	Grade of recommendation
IVIg 0.4 g/kg/day for 5 days: first-line therapy, in patients with relapse[1,2,3]	Strong recommendation
TPE (200–250 ml/kg for 5 sessions)[2,4, 6,7,8]	Strong recommendation
High dose IV MP with IVIg[6]	Weak recommendation
Gabapentin or carbamazepine[2]	Weak recommendation
Corticosteroids[2,5,9]	Not recommended
Immunoadsorption[2,5]	Not recommended
Sequential treatment with TPE followed by IVIg or immunoadsorption followed by IVIg[5,6]	Not recommended
Alemtuzumab, eculizumab, brain derived neurotrophic factor, CSF filtration, cyclophosphamide, IFNβ-1a, muronomab-CD3, mycophenolate mofetil or tripterygium polyglycoside[2]	Not recommended

CD: Cluster of differentiation; CSF: Cerebrospinal Fluid; IFNβ: Interferon Beta; IVIg: Intravenous Immunoglobulin; MP: Methylprednisolone; TPE: Therapeutic Plasma Exchange

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Disclosures

Authors, A.S., G.K., and B.S. declare no conflict of interest

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