GBS | CIDP Foundation International

Speaker Series Summary Episode 6: Current Treatment Options for GBS, CIDP, MMN, and Variants

Overview

In this episode of our Speaker Series, we dive into the world of treatments for GBS, CIDP, and MMN as well as understanding them from Dr. Saperstein, a neurologist on our Global Medical Advisory Board.

Summary

GBS	**Two main treatments: Plasmapheresis or IVIg/Subcutaneous Infusion** • **Both are clinically proven to be equally effective with the same benefits** • **IVIg is more easily accessible in most hospitals whereas plasmapheresis requires more special equipment making it less practical for some patients** • **Most do well with each treatment, yet people can still be left with limitations years after their GBS episode** **Is there ever a circumstance where GBS is not treated with IVIg?** • **The recommended window of treatment is within four weeks of diagnosis, so consult your doctor on starting treatment if you are past the four week mark from the start of your symptoms** • **Be Aware: Some insurances will advocate that if a patient has some function they do not need any treatment. Understand that their perspective comes from a financial standpoint rather than a patient-support perspective. Therefore, continue to advocate for yourself and you are always welcome to ask for a doctor to doctor consult from the Foundation.** • **It is important to note that some countries do not have access to IVIg for a number of reasons, leaving patients without access to IVIg or Subcutaneous Infusion** ◦ **Small Volume Plasma Exchange is an option https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6104782/**

GBS	**What if a patient does not respond to the first round of IVIg?**
	• Most patients will not see results within a week, every patient is different so it is hard to predict the exact moment when patients will see results
	• If a first round of IVIg does not help a patient, most doctors will not approve a second round
	  ◦ The GBS treatment guidelines and clinical evidence do not show data favorable of a second infusion
	  ◦ They will often try to use steroids or see if a patient has a variant typically unresponsive to IVIg
	• Variants like Miller-Fisher, AMAN, and AMSAN are usually treated the same way as GBS with IVIg and do not have variant specific treatments
	
	Understanding GBS from CIDP
	• Only 3% of patients have relapsing GBS
	• If a patient's illness becomes chronic and progresses past 8 weeks, it is most likely CIDP
	
	Long Term Treatments
	• There aren't any great treatments for fatigue, yet most doctors recommend exercise and physical therapy.
	• Medications are prescribed for pain management, like Lyrica, modulating the immune system and treating nerve pain
	• If there as some residual effects present for more than a year, talk to your doctor because it may be another condition affecting your wellbeing
	• Please remember to stay in touch with specialists who originally helped you with your GBS diagnosis because if you develop long-term effects from GBS they can serve as a resource!

CIDP	**What is the difference between on label and off label and how does it affect CIDP?** • On label means that a medication is FDA approved where as off-label is not FDA approved • All immunoglobin comes from healthy donors, the only difference is a pharmaceutical's method of stabilizing the immunoglobin

CIDP	• All IG for GBS is off-label while some brands are on-label for CIDP, meaning they are FDA approved to treat the condition ◦ Even though IG is off-label for treating GBS, clinical evidence shows its effectiveness so that it is still currently the most effective treatment **Subcutaneous IG (SCIg)** • Self-injection of immunoglobin under the skin into fatty part of the body such as the thighs • A typical schedule with Subcutaneous IG involves a once-a-week infusion, and then the frequency can change as illness improves • Frequent infusions of IG at a lower dose can keep the body at a steady level without side effects like weakness, headaches, etc. that can occur with a higher dose • Gives patients more accessibility and convenience with self-injection • Most doctors see it as a second option to IVIg that patients can transition to **Concentration** • Each brand has different concentrations such as 5% and even more like 10% (10% is mostly for IVIg) • Some IVIG brans can be administered subcutaneously • Some brands use 20% for CIDP and can still be given subcutaneously, the determining factor is a patient's tolerance and responsiveness • Hydration is recommended to help with side effects from IG treatments • Concentration dosage is determined by a patient's weight in kilograms **Hyqvia (Brand of IVIG; administered Subcutaneously)** • Hyaluronidase is first injected to disrupt hyaluronic acid to make a pocket for fluid to go into • It is typically injected in the abdomen and is followed by the IVIg • It takes 2-3 hours which is shorter than IVIg and longer than SCIg, given every 3-4 weeks so there are less infusions • It is recommended for patients who get more side effects from typical IVIg • It can be self-administered or with a nurse's assistance **What are some alternatives to IG (Immunoglobin)?** • In some cases, the patient's body is not tolerating IVIg or SCIg rather than the treatment not working

CIDP	• Steroids can be an option and are less expensive, some patients take IVIg and Steroids at the same time • Plasmapheresis or plasma exchange: there are some risks like clogging veins and the equipment for this treatment is in limited settings • Immunosuppressant: results may vary and take months to occur, it is often used with other therapies • Rituxan or Rituximab (brand): target cells that will become plasma cells that will make antibodies so it prevents antibodies affecting myelin therefore preventing CIDP
MMN	• IVIG is the only proven treatment that works • Steroids either do not work or worsen the illness and condition • Immunosuppressants were used in the past • The focus of treatment is to tolerate IVIg or Subcutaneous IG
Anti-Mag	• Most patients do not respond to IVIg or steroids • Plasmapheresis does not work well • Immunosuppressants are tricky because they work depending on the patient • Rituxan is sometimes used to treat this condition with mixed results
Waiting for FDA Approval	**Other Potential Therapies** • Several potential therapies are developing for GBS, CIDP, and MMN • Some of these potential therapies target the complement cascade within the immune system • FCRN inhibitors are another potential therapy option being studied in clinical trials **Efgartigimod** • An FCRN receptor blocker pulls bad immunoglobin out of the blood stream • Once a week infusion for an hour or use hyaluronidase resulting in shorter infusion times • ⅔ of patient responded well but it is unclear if the long term effects lead to long-term remission, reverse a patient's progress, or is a superior treatment to IG therapy • Currently being reviewed by the FDA as of April 2024

Relevant Resources

Treatment guidelines - https://www.gbs-cidp.org/gbscidp-guidelines-for-treatment-and-diagnosis/

Treatments & Access page – https://www.gbs-cidp.org/treatments-access/

Centers of Excellence https://www.gbs-cidp.org/support/centers-of-excellence/

Doctor to Doctor consult - https://www.gbs-cidp.org/doctor-to-doctor/

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