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Guillain-Barré- Adding Insult to Injury

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Introduction

Guillain-Barré syndrome (GBS) is described as an acute peripheral neuropathy causing limb asthenia, or weakness, that progresses over a short period of time (Lehmann, Hughes, Kieseier, & Hartung, 2012). GBS is thought to be immune mediated, generally triggered by a prior viral or bacterial infection (Blum & McCombe, 2014). Rapid diagnosis and treatment of any neurologic condition is paramount to the recovery of the individual experiencing the assault. GBS was once thought of as a singular disease, but has since been distinguished as many subgroups and with varying diagnostic criteria. The most common subgroups are acute inflammatory demyelinating polyradiculoneuropathy (AIDP) and acute motor axonal neuropathy (AMAN), and Miller Fisher Syndrome (MFS) (Blum & McCombe, 2014).

It is strongly suggested that the risk of developing GBS is increased by a host of factors including familial genetics, particular pathogen exposure and certain polymorphisms (Rinaldi, 2013). Recent attention paid to GBS has been associated with the H1N1 vaccine. However, studies conducted between 2015 and 2016 suggest an increased incidence of disease associated with the Zika virus (Cao-Lormeau, et al., 2016). When the peripheral blood of patients with GBS have been examined, it reveals abnormalities in T cells, antibodies and gene expression (Blum & McCombe, 2014). Even with rapid treatment, approximately 20% of patients are left with severe disability (Kuwabara, 2004).

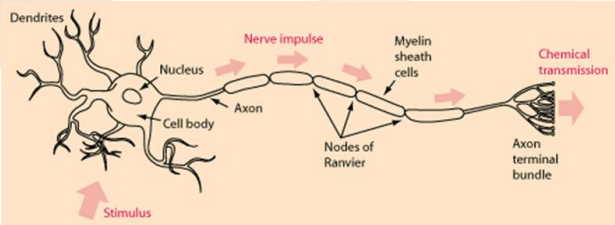

Image 1. Anatomy of a nerve cell. (Charand, 2000)

Signs & Symptoms

Symptoms tend to appear within the first 2 to 28 days, depending on the precipitating event (Kopytko & Kowalski, 2014). Classic diagnostic criteria and clinical presentation is as follows:

- Progressive motor weakness
  - More than one limb
  - Relative symmetry
  - Over a period of days, up to 4 weeks
- Areflexia
  - Universal (classic presentation)
  - Distal Areflexia with knee/bicep hyporeflexia (variable presentation)
- Autonomic dysfunction
  - Tachycardia
  - Postural hypotension
  - Hypertension
  - Vasomotor symptoms
- Afebrile

Diagnostic Testing & Criteria

In order to diagnose Guillain-Barré and the correct subtype, the following diagnostic testing must occur and results verified by the appropriate practitioners.

- Lumbar Puncture
- Cerebrospinal Fluid
  - Increased CSF protein
  - Fewer than 10 mononuclear leukocytes
- Electrodiagnostics
- Electromyography or Nerve Conduction Studies
  - Slowed or blocked nerve conduction

Underlying Pathophysiology

Guillain-Barre syndrome is described as "an acquired neuropathy characterized by inflammation of peripheral nerves," (Blum, Csurhes, Reddel, Spies, & McCombe, 2013, p. 92). Varying subtypes are mediated by different inflammatory responses. The pathophysiology of GBS ranges from antibody mediated disease to macrophage and T cell instigated destruction of peripheral nerve cells (Blum, Csurhes, Reddel, Spies, & McCombe, 2013).

T-cell receptors recognize antigens presented by HLA molecules and immune reactions are influenced by variations in T-cell genetics (Blum & McCombe, 2014). Depending on the subtype of GBS, peripheral nerve cells are presented to T-cells with the help of CD-1, CD-4 or CD-1 polymorphisms (Blum & McCombe, 2014).

Natural killer cells are lymphocytes that play an important role through chemokines and cytokines. Killer immunoglobulin-like receptors (KIR) are the largest group of natural killer (NK) receptors and exhibit roles in both inhibitory and activating processes (Blum, Csurhes, Reddel, Spies, & McCombe, 2013). KIR's are found on the surface of many NK cells but also on CD4+ and CD8+ T lymphocytes (Blum, Csurhes, Reddel, Spies, & McCombe, 2013). The importance of an alteration in the KIR/HLA combinations in GBS is the key to suggestions that innate immunity plays a significant role in the disease process.

The details of the pathophysiology vary by subtype. AIDP is the most common variant accounting for 85% of the cases and is caused by macrophage attraction to the myelin sheath (Panesar, 2014) . AMAN is most commonly seen following infection with campylobacter jejuni (figure 2) and is associated with macrophage invasion of the node of Ranvier (Panesar, 2014) . MSF cases present with positive anti-GQ1b antibodies 96% of the time but underlying pathophysiology is not completely understood (Panesar, 2014) .

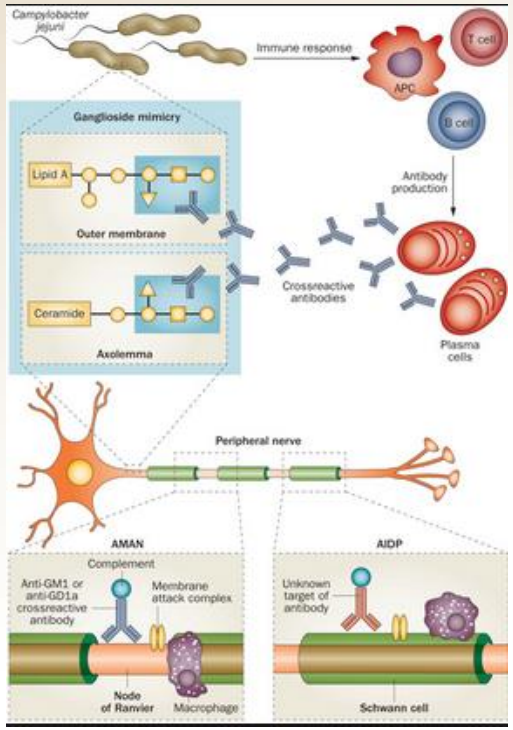

Image 2. Pathophysiology of C. Jejuni associated GBS (van den Berg, et al., 2014)

Significance of Pathophysiology

Overall, GBS is thought to be a disease due to mimicry between pathogens expressed on antigens and those on peripheral nerves. The body identifies "self" as foreign due to recent exposure to an antigen with similar properties and initiates the inflammatory response. With current research however, no specific HLA has been identified (Blum, Csurhes, Reddel, Spies, & McCombe, 2013). In demyelinating diseases, autoantibodies bind to myelin, triggering complement activation and mobilization of macrophages (Pasanen, 2015). In axonal disorders, the immune response occurs at the nodes of Ranvier, rather than the myelin sheath (Pasanen, 2015). Gangliosides appear to be the primary target in axonal disorders, and specific ganglioside antibodies have been identified in as many as 80% of patients (Pasanen, 2015). Specific targets for demyelinating syndromes have been more difficult to identify due to the nonspecific nature of macrophages.

Implications for Nursing

Nursing can make a difference in the recovery of their patients by understanding the clinical presentation, anticipating potential complications and attending to the specialized needs of the patient during the acute and recovery phase of illness. Symptoms can range in severity from mild weakness and paresthesia to total paralysis requiring mechanical ventilation. Complications can include compromised skin integrity due to limited mobility, respiratory infection due to prolonged intubation coupled with impair swallowing, and manifestations of depression due to emotional distress. Mortalities associated with GBS are generally secondary to respiratory processes.

While the clinical care a patient receives is important, emotional support and education regarding disease process are equally necessary. While motor function is significantly impacted by the pathophysiology of the disease process, cognitive function remains intact. Communication to the patient regarding what care is being provided, why and by whom is especially important to this patient population to support their emotional well-being.

The recovery phase of illness can last up to 18 months, with some patients having permanent disability.

- Appropriate Nursing Interventions
- Turning and repositioning at regular intervals
- Cough and deep breathing exercises
- Thorough skin care
- Oral and eye care
- Passive range of motion
- Provide emotional support as needed

Conclusion

Guillain-Barre syndrome diagnosis, treatment and management of relapse requires a multidisciplinary approach, supportive care and early mobility (Pasanen, 2015). Relapses are common and frequently follow vaccinations or illness; potentially occurring decades after the initial insult (Kopytko & Kowalski, 2014). Despite significant advances that have been made in treatment, GBS mortality remains steady at 4%-15% and thus, early diagnosis, appropriate treatment and prevention of complications are of the upmost importance.

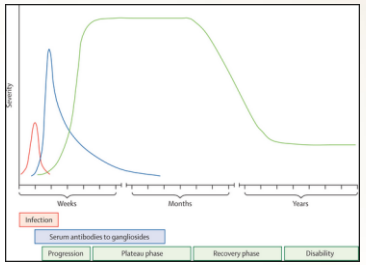

Figure 1. Course of Guillain-Barre Syndrome (Willison, Jacobs, & van Doorn, 2016)

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