

What is Guillain Barre Syndrome

- An acute immune-mediated polyneuropathy.
 - In most cases, GBS is preceded by an infection in the 3 weeks prior to onset of muscle weakness.
- Symptoms include numbness or tingling (paresthesia) in the toes and fingers, progressive weakness in the arms and legs over the next few days. Some patients experience paresthesia only in their toes and legs; others experience symptoms on one side of the body.
 - Symptoms may stay in this phase, causing only mild difficulty in walking. If the illness progresses, it can lead to complete paralysis of the arms and legs. About one quarter of the time, the paralysis continues up the chest and freezes the breathing muscles leaving the patient dependent on a ventilator. The swallowing muscles can also be affected.

What is CIDP

A chronic neuropathy resulting from damage to the myelin sheath in the Peripheral Nervous System. The peripheral nervous system is made up of sensory and motor neurons.

In CIDP, a relapse or worsening of disease is frequently preceded by an infection.

It is considered to be a related form of Guillain-Barre syndrome. It evolves much more slowly and usually is longer lasting. Some CIDP patients experience periods of worsening and improvement, and individual relapses can be confused with GBS.

Duration of progressive weakness is > 8 weeks.

Possible Causes of GBS/CIDP

- Thought to be caused by autoimmune processes
- Believed to be triggered by an event or insult such as an infection, vaccination, surgery or trauma.

IVIg and GBS/CIDP

Details to Follow

But First . . .

Immune System Players

- Lymph nodes
- Lymphatic vessels
- Thymus
- Liver
- Spleen
- Tonsils
- Blood
- Bone Marrow

Additional Immune System Components

Specialized cells include: Lymphocytes, Neutrophils, Macrophages

Specialized Proteins Include: Antibodies and Complement System

Immune System Function

- The goal
 - Recognize & destroy invading antigens
 - Differentiate between normal, healthy cells & foreign material, including cancerous cells

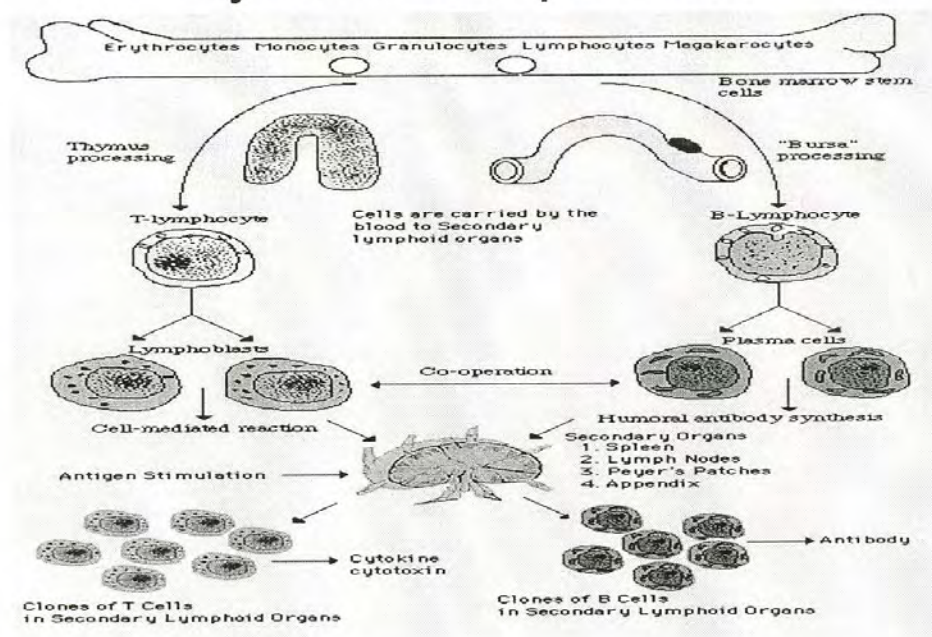
Immune System Malfunction

- The problem
 - Malfunction or absence of any of the immune system cells & proteins can lead to immune deficiencies or auto-immune diseases

How Does the Immune System Fit Into the Picture?

- If an infectious agent presents a protein, glycolipid, or other antigenic substance to the immune system that “looks” like a normal tissue component, the immune response to the infectious agent expands to include the normal tissue component.
- Whatever immune response would occur to the antigen also happens to the normal tissue.
- The result is damaged “self” tissue.

Humoral and Cellular Immune System Components



How Does IVIG Fit Into the Picture?

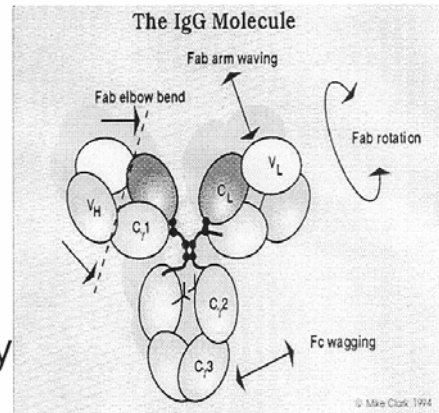
- IVIG Modulates immune system functions in auto-immune disorders:
 - By causing decreased production of antibodies including auto-antibodies produced by B lymphocytes
 - By having antibodies to defective immune proteins that cause inflammation and cell damage
 - By binding to certain damaging cells of the immune system to prevent their access to target tissues.

The Function of IVIG In Neuromuscular Disease is Complex

- It affects all the components in the immune regulatory network
- It can interfere with the expansion of certain cell lines or with the production of damaging immune globulins
- It can interfere with complement production to “turn down the volume” of the immune system
- It can interfere with membrane attack complex
- It can suppress other damaging components which act as helpers to the main players

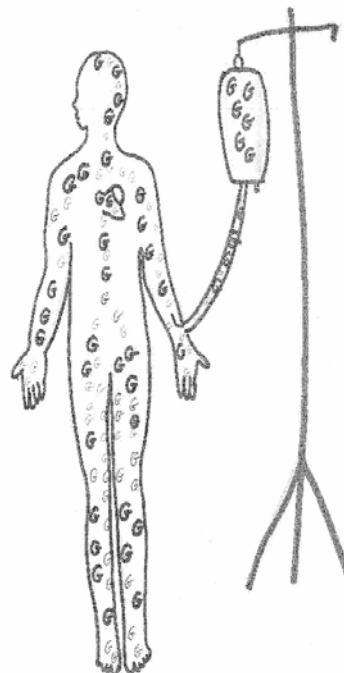
WHAT IS IVIG?

- Intravenous Immunoglobulin Preps Are “Antibody Concentrates” Purified From Large Pools of Human Plasma
- Immunoglobulins, Also known as antibodies, Are proteins that bind specifically to antigens and may help destroy them



What is IVIG Therapy?

- IVIG is administered from an IV bag or bottle directly into a vein
- Used for People Who Have Deficient or Dysfunctional Immune Systems or who Have an Autoimmune Disease



• IVIG is An Accepted Treatment

- CIDP – IVIG is first line therapy, especially in the early inflammatory phase
- Guillain Barre Syndrome –IVIG has become the treatment of choice as it is equally effective with plasma exchange and much more accessible

Considerations for IVIg Therapy

- How does effectiveness compare from one product to another?
- What do the products look like and how do they come?
- How do side effects compare from one product to another?
- Can side effects be prevented?
- Is there a potential for serious adverse effects?
- Do you have to go to the hospital for this therapy?

PRODUCT SELECTION

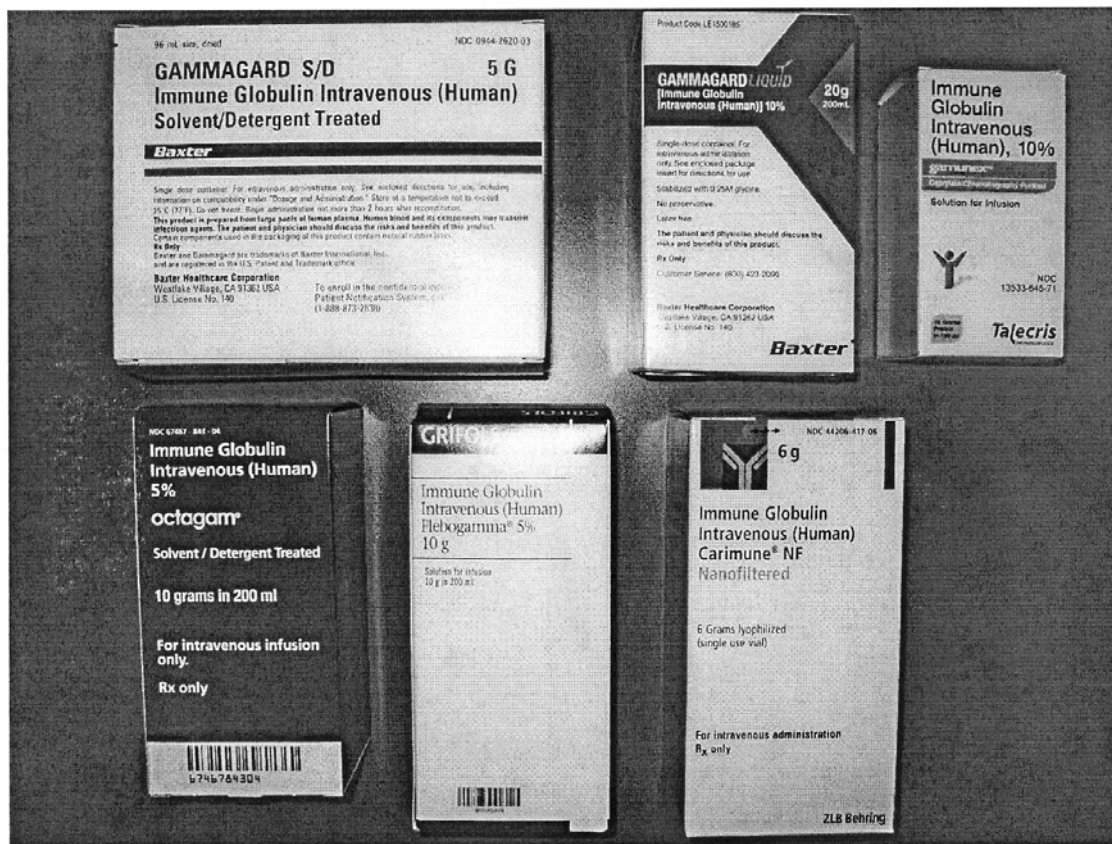
There are many brands of IVIG, all made by established pharmaceutical manufacturers.

There are no head to head studies between brands showing one to be more effective or better tolerated than another.

Products are dosed equivalently.

Each product is FDA approved for limited indications. No neuromuscular indication is FDA approved.

When choosing an IVIG Product, patient specific clinical criteria should be evaluated before making a brand and concentration selection.

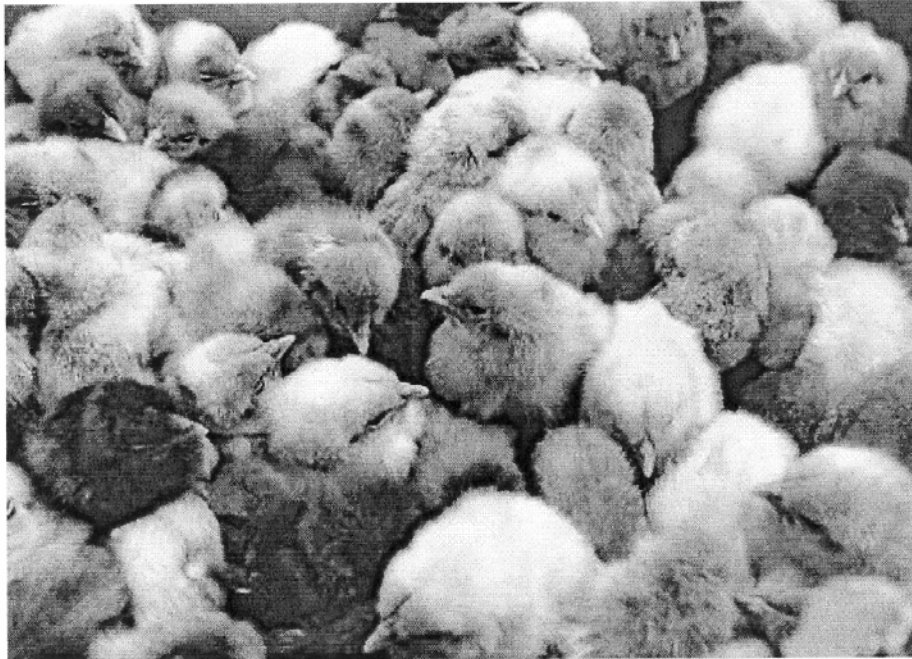


Examples of Patient Specific Considerations

- Potential for kidney problems
- Potential for blood clots
- Ability to handle extra fluid
- Age
- Need for hydration
- Potential IgA sensitivity
- Predisposition to headache
- Full medication profile, height, weight, nutritional status

DIFFERENCES AMONG AVAILABLE PRODUCTS

- LYOPHYLIZED (freeze dried) POWDERS
- LIQUID PRODUCTS
- PRODUCT STORAGE
- MIXING REQUIREMENTS
- CONCENTRATION DIFFERENCES
- STABILIZERS - SUCROSE
- COMPATIBILITY WITH FLUSH SOLUTIONS
- PURIFICATION STEPS



Common Side Effect of IVIG in Hens

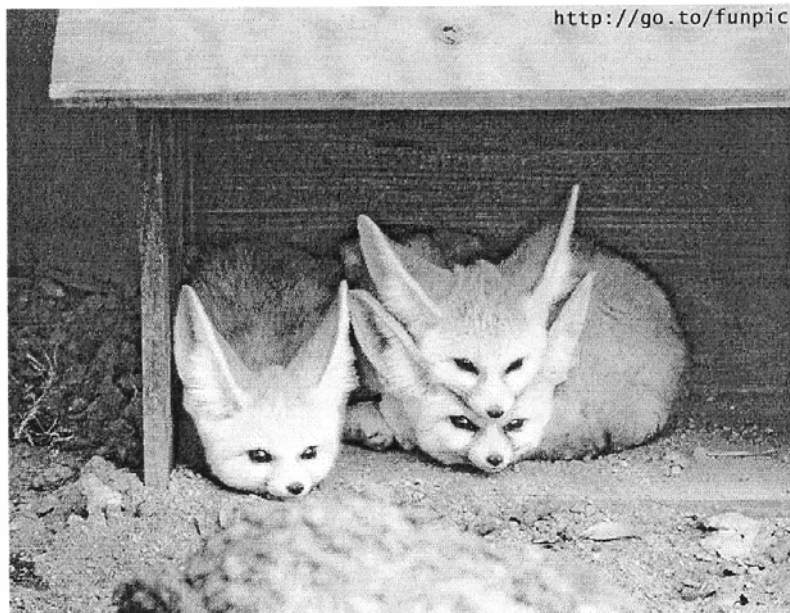
Common Side Effects of IVIG

- Headache
- Chills, flushing, fever
- Nausea
- Muscle aches or back pain
- Wheezing
- Chest tightness

Preventing Side Effects

- Patient's clinical information should be considered prior to therapy to determine potential for specific side effects to occur and allow for a plan to minimize or prevent those side effects.
- Premedication with antihistamines, corticosteroids, anti-inflammatories and anti-fever medications can reduce or prevent side effects.
- Manipulation of infusion rate by the RN can reduce severity of a side effect. Intervention may prevent a side from occurring during the infusion or one which is anticipated to occur after the infusion

Serious Adverse Effects of IVIG



Unusual and Potentially Serious Adverse Effects

- Anaphylaxis due to IgA sensitization
- Renal failure is an uncommon effect and is associated with administration of sucrose containing IVIg products.
- Rare incidences of thrombosis occur most often in patients with an underlying coagulation disorder
- Aseptic meningitis - A self limiting condition which manifests as fever, neck stiffness, headache, confusion, nausea and vomiting.
- Stroke

In What Settings Can IVIG Be Infused ?

- IVIg can be administered in the comfort and privacy of a patient's home.
- IVIg can be administered in the hospital.
- IVIg can be administered in an infusion center or in a cancer center where people can also get chemotherapy treatments.

ACCREDO

- Accredo is a home infusion company which specializes in administration of IVIG in the home.
- Accredo Pharmacists and Nurses are experts in administration of this therapy.
- Accredo carries all brands of IVIG products – as long as the product is available in the marketplace.
- Accredo pharmacists and nurses customize therapy to each patient's needs.



An IVIG Infusion from Accredo is Like a Day at the Beach

Accredo Model for Safety of Administration of IVIG in the Home

- IVIg Pharmacist and Nurse communicate with patient and physician to carefully evaluate clinical criteria to determine optimal IVIg dose, frequency and product selection.
- Accredo RNs are specially trained for IVIg therapy. They monitor blood pressure, infusion rate, potential for side effects, and manage all aspects of the infusion.
- Special medications are maintained in the home to stop a serious side effect before it becomes dangerous.

References

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