IVIg Therapy in Pemphigus

Donna A. Culton, MD, PhD
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What is IVIg?

- Intravenous Immunoglobulin
  - Intravenous (IV) – administered through an IV ( infusion)
  - Immunoglobulin (Ig) – fancy word for antibody

Viral particle Desmoglein
What happens in pemphigus...
How does IVIg work?

Bone Marrow: Pro-B, Pre-B

Immature → Mature

Germinal Center

Ag

Memory

Plasmablast

Bone Marrow: Plasma cell

Immunoglobulin production

X

Plaque formation

Teeth

Histological section
What is IVIg?

- Pooled antibodies collected from healthy volunteers
- Contains antibodies with specificities to common pathogens and vaccinations to which the donors have been exposed
How does IVIg work?

- Multiple proposed mechanisms
- Commonly thought to work by increasing catabolism (or the turnover) of antibodies through FcRn saturation
- Essentially, the body has mechanisms to control how many antibodies it has...if it gets too many it responds by getting rid of some
How does IVIg work?
How does IVIg work?

*How does FcRn fit in?
What are the benefits of IVIg?

- Works fairly quickly … typically within a week or two
- Rapid clearance of autoantibodies and clinical improvement
  - Within 1 week new lesions ceased to form
  - Within 2 weeks 80% of lesions had healed
- In addition to prednisone, IVIg is one of the ‘fast acting’ treatment options available

Amagai M et al. JAAD 2009;60:595-603.
What are the benefits of IVIg?

Day 0 – 1:640

2 weeks – 1:320

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What are the benefits of IVIg?

- Non-immunosuppressive … it contains healthy antibodies!
- Antibody levels to other common antigens remain stable
- Can be a good choice for patients with active infection

Amagai M et al. JAAD 2009;60:595-603.
What are the downsides of IVIg?

- Given over 3-5 days typically at an infusion center… so pretty time intensive
- Home infusion is an option, but has it’s own challenges
What are the downsides of IVIg?

- The positive effects only last about 1 month
How does IVIg work?

Bone Marrow

Pro-B  Pre-B  Immature  Mature  Germinal Center  Plasmablast

Spleen

Ag  Memory

Plasma cell

Bone Marrow

Pre-B  Immature  Mature  Germinal Center  Plasmablast

X

[Image of a person holding their mouth in discomfort]
What are the downsides of IVIg?

- The positive effects only last about 1 month
  - Use as a rescue
What are the downsides of IVIg?

- The positive effects only last about 1 month
- Use as a rescue
- Use as a bridge

![Graph showing the positive effects of IVIg lasting for about 1 month, with options for use as a rescue or bridge.](image_url)
What are the downsides of IVIg?

- The positive effects only last about 1 month
  - Use as a rescue
  - Use as a bridge
  - Use as maintenance therapy
What are the downsides of IVIg?

- Cost ....about $10,000 per cycle
- Can be less with home infusions
Things to consider beforehand…

- Large protein load – requires healthy kidneys
- Large volume load – requires a healthy heart
- Prothrombotic – risk of contributing to blood clot formation in patients that are prone to them
- IgA deficiency – your doctor will also check to make sure you are not IgA deficient (genetic condition that you could be unaware of) before giving IVIg
Common side effects

- Infusion reactions – blood pressure, heart rate, hives
  - Most common on first infusion
  - Usually not a deal breaker...can pre-medicate with tylenol and benadryl, slow the infusion rate
- Non-infusion related side effects – headache, fever, fatigue, chills, flushing, dizziness, hives, nausea/vomiting, cramping, changes in blood pressure
Summary of IVIg

- Works pretty fast
- Doesn’t last very long
- Can be used with other treatments
- IV infusion
- Costly
- Some possible side effects to be aware of