IG Therapy

How to Individualize Your Treatment plan to have best outcomes with least side effects, and

How to ensure your insurance plan will cover IVIG/SCIG

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Patient Advocacy & Provider Relations
CSI Pharmacy is a nationwide specialty pharmacy dedicated to servicing patients with chronic and rare illnesses requiring complex care. We are invested in improving the health—and the lives—of the people we serve by offering expert clinicians, passionate advocates, committed healthcare partners and strategic reimbursement experts. Our goal is to optimize patient outcomes, while treating patients as if they were members of our family.
CSI Pharmacy Specializes in Myositis

We treat...

- Dermatomyositis
- Polymyositis
- Necrotizing Myopathy
- Inclusion Body Myositis
- Juvenile Myositis

20% of our patients have a form of Myositis
Objectives / Goals

• What is Immune Globulin Therapy? IVIG versus SCIG

• How can IG Therapy be individualized?
  - Brand Selection
  - Hydration & Infusion Rate
  - Adverse Reactions (Prevention versus Treatment).

• How to choose the right insurance plan and make sure IVIG/SCIG is approved?
What is IVIG

- Immune Globulins = Antibodies (IgA, IgD, IgE, IgG, & IgM)

- Intravenous Immune Globulins (IVIG)

  - 10 brands of IVIG, 3 brands of SCIG in the U.S.
  - manufactured from plasma pools (1,000 to 60,000 donors)

  - primary component is IgG
  - brands generally considered equally effective, however have different tolerability profiles.
Plasma Products

Plasma (55% of Blood Volume)

Proteins (7%)

Water 91.5%

Factors, et.al. (1%)

Misc (7%)

Albumin (54%)

Ig (38%)

FII
FV
FVII
FVIII
FIX
FX
FXI
FXII
FXIII
VWF
ATIII
Prot C
Prot S
Plasmin
Plasmin-inhibitor
Antibody Function

Fight off harmful substances in the body.

Recognize antigens on the surface of pathogens and toxins.

Facilitate the neutralization, destruction, and elimination of pathogens and toxins.
Autoimmune Disorders

- Autoantibodies incorrectly label a healthy normal part of the body as harmful.

- IG Therapy (IVIG or SCIG) decreases the production of these autoantibodies.
# IVIG and SCIG Preparations Approved For Use In The United States

<table>
<thead>
<tr>
<th>Product</th>
<th>Route</th>
<th>Manufacturer</th>
<th>Indications</th>
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| Carimune NF 3%, 6%, 9%, 12% (once reconstituted) | IV    | CSL Behring                    | Primary humoral immunodeficiency  
|                        |       |                               | Immune thrombocytopenic purpura                                             |
| Cuvitru 20%            | SC    | Shire                         | Primary humoral immunodeficiency                                            |
| Flebogamma DIF 5%, 10% | IV    | Instituto Grifols, SA         | Primary humoral immunodeficiency                                            |
| Gammagard Liquid 10%   | IV/SC | Shire                         | Primary humoral immunodeficiency (IV/SC)  
|                        |       |                               | Multifocal motor neuropathy (IV)                                           |
| Gammagard S/D 5%, 10%  | IV    | Shire                         | Primary humoral immunodeficiency (IV/SC)  
| (once reconstituted)   |       |                               |  
|                        |       |                               | B-cell chronic lymphocytic leukemia                                         |
|                        |       |                               | Immune thrombocytopenic purpura                                             |
|                        |       |                               | Kawasaki syndrome                                                           |
| Gammaked 10%           | IV/SC | Kedron Biopharma              | Primary humoral immunodeficiency (IV/SC)  
|                        |       |                               | Immune thrombocytopenic purpura (IV)                                       |
|                        |       |                               | Chronic inflammatory demyelinating polynuepathy (IV)                        |
| Gammalux 5%, 10%       | IV    | Bio Products Laboratory       | Primary humoral immunodeficiency                                            |
|                        |       |                               | Immune thrombocytopenic purpura                                             |
| Gamunex-C 10%          | IV/SC | Instituto Grifols, SA         | Primary humoral immunodeficiency (IV/SC)  
|                        |       |                               | Immune thrombocytopenic purpura (IV)                                       |
|                        |       |                               | Chronic inflammatory demyelinating polynuepathy (IV)                        |
| Hizentra 20%           | SC    | CSL Behring                    | Primary humoral immunodeficiency                                            |
| HyQvia 10%             | SC    | Shire                         | Primary humoral immunodeficiency                                            |
| Octagam 5%, 10%        | IV    | Octapharma Pharmazeutika      | Primary humoral immunodeficiency (5%)                                       |
|                        |       | Octapharma USA                | Chronic immune thrombocytopenic purpura (10%)                              |
| Privigen 10%           | IV    | CSL Behring                    | Primary humoral immunodeficiency                                            |
|                        |       |                               | Immune thrombocytopenic purpura                                             |
|                        |       |                               | Chronic immune thrombocytopenic purpura                                    |

**July 2017** - Octapharma granted orphan drug status for the use of Octagam 10% in dermatomyositis

**Aug 2018** – FDA approves Octapharma’s new IVIG product ... Panzyga 10%

**PHASE III Trial** – proDERM Study in DM by Octapharma.

2gm/kg IVIG Q 4 weeks in refractory DM.
Individualized Therapy

• Prevention of adverse effects
  - Product Selection.
  - Hydration & Pre-medications
  - Taper up rate slowly.

• Treatment of adverse effects

• Customize future Infusions
General Statements on Product Differences

• IG Manufacturers have tweaked their formulations over the past decades in an effort to improve their product’s safety.

• The primary component of Ig products is immunoglobulin G (IgG). Brands of Ig can differ in IgG monomer, dimer, and aggregate concentrations, IgA and IgM content, stabilizers, additives, and levels of impurities.

• These differences result in different side-effect profiles.

• **By appropriate product-selection & utilization, the rate of adverse drug reactions and adverse events can be reduced!**
## Risk Factors Affecting Tolerability

<table>
<thead>
<tr>
<th>Risk Factors</th>
<th>IVIG Factors</th>
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<tbody>
<tr>
<td></td>
<td>Volume Load</td>
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<tr>
<td>Cardiac Impairment</td>
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<td>Renal Dysfunction</td>
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<td>Anti-IgA Antibodies</td>
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<td>Thromboembolic Risk</td>
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<td>Diabetes</td>
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<td>Geriatrics</td>
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<td>Neonates/Pediatrics</td>
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# IVIG & SCIG Dosing in DM & PM

| IVIG dose in dermatomyositis & polymyositis | 2gm/Kg in divided doses over 2 to 5 days.  
Typically dosed once month and then according to patient response* |
|---------------------------------------------|---------------------------------------------------------------------|
| SCIG dose in Dermatomyositis & Polymyositis | Varies greatly, and not well defined in the literature. In our experience;  
2gm/Kg per month divided into once weekly or twice weekly doses. |

- IgNS Immunoglobulin Therapy Standards of Practice  
  AANEM Consensus, IVIG 2009
IVIG Infusion Reactions

1) Aseptic Meningitis (Severe Post Infusion Headache)
2) Headaches during infusion or Hypertension/Hypotension
3) Flu-Like Symptoms
4) Dermatological
5) Anaphylactic reactions
6) Rigors
7) Severe Back and/or Leg pain
8) Thromboembolic events
9) Hemolytic Anemia
Prevention of Adverse Reactions

• Product Selection based on risk factors
• Pre-medicate with Tylenol & Benadryl
• Adequate Hydration
• Slow Infusion rate
Treatment of Infusion reactions

• Slow Infusion Rate
• Administer IV Fluids
• Solu-Medrol 1mg/Kg (max 125mg) IV push over at least 5 minutes. May add as a premedication for future infusions.
• Consider a brand change
• Inability to tolerate IVIG, consider switching to SCIG
Subcutaneous Immune Globulins (SCIG)

- Hizentra 20%
- HyQvia 10%
- Cuvitru 20%

- Gammagard Liquid, Gamunex-C & Gammaked may be administered IV or SC
SCIG Possible Advantages

• Subcutaneous weekly infusions keep IgG levels at a steady state – consistent
• No pre-meds required
• Side effects are mainly – localized*
• Autonomy and independence
• NO VENOUS ACCESS
Types of Health Insurance Plans

- Medicare
  - Traditional
  - Supplemental Plan
  - Part D Plan
  - Advantage Plans
- Medicaid
  - Expansion States
- Veterans Affairs
- Tricare
- Federal Employees Health Benefits Plan
- Employer-Sponsored Plans
  - Choice of plans
- Healthcare Market Place Plans (Affordable Care Act)
- Healthcare Sharing Plans (Faith Based Plans)
- Other Individual Plans
How Does Health Insurance Make You Feel?
What’s in Store for Health Care?
Complex problem consisting of: Destabilizing of the ACA; Driving up Costs of Health Insurance Plans in the Marketplace; Introduction of Cheap Health plans (Trump Plans) with no protections for the most vulnerable populations needing the most effective & expensive therapies = THE PERFECT STORM
What are the Current Threats for Patients with Myositis?

- Changes in the Affordable Care Act (ACA)
- Trump plans
- Healthcare Sharing Plans
- Understanding Medicare
- Knowing the Prior-authorization process
- Patient Choice
- Current Shortage of IVIG/SCIG
Affordable Care Act – How does it Help Rare Disease patients?

• **Elimination of Pre-existing Conditions:** Insurance providers are prohibited from denying coverage to children based on pre-existing conditions. Protections were expanded to all health insurance policies.

• **Out-of-pocket Cost & Deductibles:** Limits placed on the amount insurance companies can require policyholders to pay in out-of-pocket expenses such as co-payments and deductibles. *Changed.*

• **Individual Mandate:** Individuals will be required to have health insurance, with some exceptions, or will be subject to an annual financial penalty. Maximum fine of $2,085 per family or 2.5% of household income. *Eliminated*
  - *Causes the cost of all of the*
Affordable Care Act (cont.)

No Lifetime & Annual Caps: Plans are prohibited from using lifetime limits in issued or renewed policies. Annual Caps were phased out.

End Of Rescissions: Insurance providers can no longer drop an individual's coverage when he or she gets sick.

Donut Hole Rebate: Provides a 50% discount on all brand-name drugs in the donut hole and begins phasing in additional drugs to close the hole by 2020.
Affordable Care Act (cont.)

ALL IN JEOPARDY!!!

It is up to the courts...

Make sure you stay insured or Pre-existing Conditions will come into play Again!
**Trump Plans**

Short-term plans can exclude coverage for pre-existing conditions and can omit some benefits deemed essential in the Affordable Care Act. Short-term insurance policies were originally intended for people who were between jobs or needed temporary coverage for other reasons. There is nothing that would prevent companies from underwriting and issuing new policies to individuals at the end of the one-year coverage term.

UnitedHealth Group, does not cover the following benefits:

- Prescription drug coverage
- Expenses related to a normal pregnancy
- Treatment of mental disorders

National General, does not cover the following benefits:

- Outpatient prescription drugs
- Normal pregnancy or childbirth
- Routine well-baby care
- Costs resulting from a pre-existing condition
Healthcare sharing plans

• Faith-based programs which facilitate voluntary sharing among members for eligible medical expenses
  • Less expensive than health insurance ($300 to $500 per month/family)

• However...
  • Biggest problem with all healthcare sharing programs is their prescription drug policies.
    • People who require ongoing expensive prescriptions are only covered for a short duration. Maintenance prescriptions are not eligible for sharing at all. Members are encouraged to participate in prescription discount programs such as NeedyMeds, GoodRX, OneRX and LowestMed.
    • Exceptions, cancer and transplant recipients may be covered for only 6 months. Type 1 Diabetes would cover...
Medicare

Traditional Medicare

Part A – Hospital
Part B – Physician, Outpatient
Part D – Prescription Drug Plan
  • Home Infusion Therapies covered under Part D like IVIG. Donut Hole coverage plans are offered. If you are low income, assistance plans are offered.

Supplemental Plans

Medigap plans
  • If you are disabled and don’t choose a Medigap plan when offered, may not be eligible until 65 years old.
  • Pre-existing conditions may apply.

Medicare Advantage Plans

Plan C – Medicare HMO
  • Limits network of providers
  • Limits specialty drugs
  • Step Therapy
  • Need permission to go out-of-state
“Did you get prior authorization before you became ill?”
Prior-authorization

• A prior authorization is an extra step that most insurance companies require before they decide if they are going to pay for expensive specialists, procedures, and specialty therapies (medications).

• During this process, the insurer may request and review medical records, test results, and other information so they can make an informed decision.

• It will be determined if the service will be deemed medically necessary and identified as a covered service under your insurance policy based on what information was submitted.

• Usually, your doctor or pharmacist is notified in writing or by phone and you will receive a letter.
  • If you are denied you can appeal.
  • **YOU MUST APPEAL!**
  • If you are told you do not need a prior-authorization, make sure or your will be responsible for the bill if you get denied!
Patient Choice

Have you been forced to change providers, pharmacies, labs?

Have you been told you can no longer receive the brand of therapy you were getting?

Have you been told you can no longer receive the care you have been receiving from the specialist you like or the home care provider you have been with?

The Elimination of Patient Choice
IVIG/SCIG SHORTAGE

• BRANDS ON THE FDA SHORTAGE LIST
  • BIO PRODUCTS LABORATORY GAMMAPLEX
  • SHIRE/TAKEDA CUVITRU
  • SHIRE/TAKEDA GAMMAGARD LIQUID

• MOST DISTRIBUTORS HAVE PUT ALL BRANDS OF IVIG/SCIG ON ALLOCATION TO ENSURE THAT PATIENTS GET THE PRODUCT THEY NEED AND NO PRODUCT SITS ON THE SHELF OF ANY DISTRIBUTOR OR PROVIDER WITHOUT BEING USED WHILE WE ARE IN SHORT SUPPLY.

• SUMMER MONTHS USUALLY GET A LITTLE TIGHTER IN SUPPLY BECAUSE MANY OF THE MANUFACTURING PLANTS CLOSE TO BE CLEANED. SO WE WILL CONTINUE TO SEE TIGHT SUPPLY.

• IF YOU ARE BEEING TOLD THAT YOU CANNOT GET YOUR INFUSION BECAUSE OF A SHORTAGE OR THAT YOU ARE NOT HIGH ENOUGH PRIORTITY, THIS IS NOT ACCEPTABLE, PLEASE CONTACT ME AND I WILL HELP.
• Patients Need to be their own Advocates!
• Patients Need to be knowledgeable about what’s in their health insurance plan and how to choose the right plan for their needs.
  • Don’t assume your plan is the same as last year. READ YOUR PLAN!
• Employer-based plans are carving out prescription drug benefit plans.
• More High-deductible plans are being introduced.
• State plans have less protections.
• Medicare Advantage = less choice
Q & A

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