POSTER ABSTRACTS

IgNS 2018 Poster Session and Reception

Friday, October 19 • 5:00 - 7:00 • Landmark Circle

#1 Allergic Adverse Drug Reactions in IgA-Deficient Primary Immune Deficiency Disease Patients Treated with Subcutaneous Immunoglobulin and Facilitated Subcutaneous Immunoglobulin in the Home

Authors: Elissa Ritt, DHSc; Joseph DiStefano, RPh; Leslie Vaughan, RPh; Michelle Greer, RN

Introduction: Though intravenous immunoglobulin (IVIg) is considered standard of care in the treatment of patients with primary immune deficiency disease (PIDD), it is contraindicated in patients with low IgA levels and titers of anti-IgA antibodies due to the risk of anaphylactic and anaphylactoid reactions. Subcutaneous immunoglobulin (SCIg) is considered a safe, efficacious alternative to IVIg in the treatment of PIDD. A newer treatment option, facilitated SCIg (fSCIg), which is SCIg co-administered with recombinant human hyaluronidase to increase immunoglobulin absorption, has also been FDA-approved to treat PIDD. Previous literature has noted the safe treatment of PIDD patients who have anti-IgA antibodies with SCIg formulations when treatment with IVIg may not be possible. The purpose of this study is to assess the rate of allergic-type adverse drug reactions (ADRs) in PIDD patients with low IgA levels who have been treated with SCIg or fSCIg in the home. Methods: The medical records of all PIDD patients with low IgA levels (Results: Of the 15 medical records included, 13 patients were treated with SCIg, and two patients were treated with fSCIg. Of the SCIg-treated patients, 10 were treated with a 20% concentration SCIg product, and three were treated with a 10% concentration SCIg product. No allergic-type adverse events were noted in this population. Three patients experienced four other ADRs including local site reactions, headache, and elevated blood pressure. Discussion: In the 15 medical records examined, all patients were treated safely and without allergic- type ADRs. Anti-IgA antibody titers were not assessed, but anti-IgA antibodies can occur in up to 40% of PIDD patients with deficient IgA levels. Though no allergic-type ADRs were seen in this population, this study is limited by its small sample size. Conclusions: In the population studied, SCIg and fSCIg appear to be safe treatment options for PIDD patients with low IgA levels.

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#3 Design and Rationale of a Retrospective Analysis of the Impact of Rapid Rate Intravenous Immunoglobulin Infusion on Patient Safety, Tolerability, and Cost Reduction

Authors: Van Doan, Pharm.D. Candidate (1), Karen Kaczmarek, RN, BPS, RPT, CHC (2), Kelly Webb, RN, PHN, IgCN® (2), Dorit Freund (2), Jhonny Castrillon (2), Michael Rigas, Pharm.D. (2)

Introduction: A national patient-centered home infusion company has traditionally administered intravenous immunoglobulin (IVIG) at conservative maximum rates to minimize adverse events. However, the total time and cost required to infuse IVIG therapy at lower rates may be burdensome for patients and healthcare providers. Therefore, if infusion rates can be safely increased, this could represent both a time- and cost-saving measure that would benefit patients and providers. The objective of this study is to identify the impact of increased on-label infusion rates of a specific high-purity IVIG product on patient safety, tolerability, and potential cost reductions as a result of decreased needs for nursing care. Methods: This study was designed as a retrospective data collection and analysis. One specific high-purity IVIG product [Octagam] was administered at higher rates to a subset of patients, keeping within the manufacturer's recommendations for maximum infusion rates of 350ml/hr and 504ml/hr (in a 70kg patient) for 5% and 10% concentrations, respectively [1, 2]. The inclusion criteria included all patients who received the IVIG product from March 2008 to May 2018 with comprehensive data regarding the rate at which the product was administered as well as subsequent clinical outcomes. Patients will be divided into two groups that provide an even distribution for analysis: those who received ≤ 110 mL/hr IVIG infusion (Group 1, n=227) and those who received > 110 mL/hr (Group 2, n=210). Data will be accessed from the electronic medical records (CPR+) of the national home infusion company. Pharmacy and nursing progress notes will be evaluated for details regarding severity of adverse events and tolerability of the prescribed infusion regimen. Results: The primary endpoint of the study is to compare overall safety outcomes between Group 1 and Group 2, both by number of infusions (N=9,876) and by patient (N=437). The secondary endpoints include comparisons of safety outcomes by dose, infusion rate, number of infusions, product concentration, diagnosis, age, and comorbidities. The exploratory endpoint includes models of potential cost savings due to reductions in nursing care hours as a result of infusion rates > 110 mL/hr. Discussion: Pending the data analysis regarding the safety and tolerability of IVIG infusion at rates > 110 mL/hr, a determination can be made as to the initiation of revised infusion administration protocols that could potentially benefit patients and healthcare providers. In addition, the design of future prospective studies may be explored to measure the extent of overall cost savings as well as quality of life benefits to patients. Conclusions: Data analysis for this study is in progress. Preliminary results indicate that rapid infusion of the high purity IVIG product is safe and well-tolerated. Final results are expected in Q3, 2018 and will be available for review.

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#4 Clinical and Quality of Life Outcomes Following Infection Rate-Based Dosage Management in a Primary Immune Deficiency (PID) Population Subset: Data from the Immunoglobulin Diagnosis, Evaluation, and key Learnings (IDEaL) Patient Registry

Authors: Loretta Kristofek RN, BSN(1), Allyson Checkley PhD(1), Samantha Kile MS(1), William Bolgar PharmD(1), and Luqman Seidu MD(2)

Introduction: The IDEaL Patient Registry collects longitudinal information on patients receiving immunoglobulin (Ig) replacement therapy in an alternate care setting. This large real-world population allows for analysis of the clinical management of PID patients

and their health outcomes while receiving Ig treatment. PID patients treated with Ig replacement therapy often experience breakthrough infections. Management of dosage in relation to infection rate may improve the clinical and quality of life outcomes in this population. Our objective is to evaluate health outcomes associated with increasing Ig dosage from a focus group of PID patients experiencing above average infection rates. Methods: Patients were consented using an IRB-approved consent. Infection rates and dosages were analyzed for all study patients. Patients who were currently receiving Ig therapy and had greater than 3 infections and a) less than the minimum package insert dose of 100 mg/kg/wk, b) less than the IDEaL patient average dose of 129 mg/kg/wk, or c) less than the IDEaL patient dosage at which all infection rates fell below 3 infections per year (200 mg/kg/wk) were chosen for further analysis. The patients' physicians were presented with infection and dosage information and were offered the opportunity to increase the patient's dosage. Of the 10 eligible patients, 5 had their dosages increased. Patient data was obtained from pharmacist- and RN-completed Clinical Progress Reports, and the patient-completed SF-36v2 quality of life (QOL) survey and Life Quality Index Questionnaires (LQIQ) administered by mail every 6 months. Outcomes were tracked for 1 year post dosage change. Results: Three of the 5 patients had a decrease in their average annual infection rate of 50% or more (4.2, 5.6, and 4.7 infections/yr to 2.4, 0, and 0 respectively), and 2 had an increased infection rate (8.6, 6.7 to 15 and 12 respectively). The LQIQ response to whether treatment has improved their health remained at the highest rating for 1 patient, increased for 2 patients, and decreased for 1 patient. The SF-36v2 physical component score improved in 2 patients and decreased in 1, while the mental component score improved in 1 patient and decreased in 2. Discussion: In this pilot project, most patients experienced a decrease in infection rate as well as an improved perception of health and a higher physical component QOL score. The lack of outcomes improvement in 2 patients was potentially influenced by unhealthy body mass index related dosing issues and/or medications to treat autoimmune complications which may have further suppressed their immune system. More research is needed to better understand the influence of these factors on Ig dosing strategies. Overall, patients felt that their physical health had improved, though their mental health status slightly declined. For one patient, decreases in both scores may be attributed to placement in a rehabilitation facility due to congestive heart failure exacerbation and renal insufficiency. Conclusions: Clinical outcome should play a role in dosing strategies, and an individualized dosing approach to optimize treatment effects in this patient population can be achieved as a coordinated effort between pharmacists and physicians.

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#5 AN ANAYLSIS OF IG ADMINISTRATION PRACTICES AMONG IGNS MEMBERS

Authors: Susan A. Bauer, BSN, RN, CRNI®, VABC American Outcomes Management, L.P. (AOM)

Introduction: Intravenous Immunoglobulin (IG) is a therapy used for a multitude of disease processes including infectious, immune deficiencies, autoimmune diseases and inflammatory processes (Dourmishev, LA et al. 2016). IG has the potential for serious and life threatening consequences when administered incorrectly including acute renal failure, aseptic meningitis and thrombotic events including stroke and myocardial infarction (Scheinfeld, NS 2017). Each manufacturer has recommended infusion parameters varying in ramping frequency and rates based on patient weight. AOM contracts with agencies throughout the country to administer IVIG to patients in the home environment. This presents challenges in identifying nursing organizations skilled and knowledgeable in this specific aspect of care. During the 2017 IGNS Conference our goal was to get clarity and a consensus of how intravenous immune globulin therapy should be ramped during initial and subsequent infusions. We wanted to identify the practices that our colleagues were using for this very important aspect of care. What we anecdotally found was a lack of uniformity in our industry. Methods: We developed a survey to send to all IGNS members to identify specific infusion parameters used by each member's organization to identify current practices. This survey was made possible with the assistance of IGNS and BPL. The survey consists of 14 questionnaires, including such parameters as:

- Use of Premedications and Hydration
- IG administration weight vs volume based
- Ramping protocols
- Side effect limitations
- Client perceptions to IG infusion limitations

We will present the results of this survey, in detail, at this year's IGNS Conference. Results: Results to this survey have just begun coming in. Preliminary results suggest that 65.5% of the respondents administer IG based on the patient's weight. Moreover, 42.9% of the respondents use the same ramping protocol for all 10% products available although manufacturer recommendations vary. While 32.9% of the respondents follow a 15-minute ramping protocol, 47.1% follow a 30-minute ramping protocol. Almost half of respondents (49.1%) indicated that ramping parameters were provided to the nurse on either the prescription label or an alternate document. Other respondents may not have received the information or it was left to the nurse to calculate infusion rates or other communication methods used. While 62.2% used Ibuprofen as a premedication (based on use >50% of therapy administration), 55.5% used Acetaminophen and only 23.5% routinely used hydration with IG. Discussion: Administration practices vary by institution, clinician and prescribing practices as evidenced by the results of this survey. Standards for IG administration practices are necessary in order to achieve better patient outcomes. Conclusions: This survey identified that there are many variations used when administering IG therapy. IG can be safely administered to patients in all clinical settings and in the home. Clarity needs to be provided to the clinician to enable them to safely administer the prescribed therapy and minimize the potential for adverse reactions.

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Scheinfeld, N.S., (2017). Intravenous immunoglobulin. Medscape. June 21, 2017.

https://emedicine.medscape.com/article/210367

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#6 Onboarding Experience of Pediatric Patients With Primary Immunodeficiency Diseases With Subcutaneous Human Immune Globulin 20% (Ig20Gly)

Authors: Kenneth Paris (1), Iftikhar Hussain (2), Sudhir Gupta (3), Ping Wang (4), Barbara McCoy (4), Christopher J. Rabbat (5), Leman Yel (6)

Introduction: The safety and efficacy of subcutaneous immune globulin 20%, Ig20Gly, was demonstrated in a phase 2/3 North American study (NCT01218438) in patients with primary immunodeficiency diseases (PIDD). This post hoc analysis assessed the onboarding experience with Ig20Gly in pediatric patients. Methods: Patients aged ≥2 years received weekly Ig20Gly (Cuvitru) infusions at volumes of ≤60 mL/site and rates of ≤60 mL/h/site for ~1.3 years. To evaluate the Ig20Gly onboarding experience, adverse events (AEs), tolerability, and infusion parameters were assessed in patients aged 2 to Results: Most infusions (97.3%; 1134/1166) were not associated with a causally related local AE; 66.7% (14/21) of patients did not experience a causally related local AE. Five patients (23.8%) reached the maximum infusion rate of 60 mL/h/site for ≥2 infusions. A total of 54.3% and 95.2% of infusions were completed in Discussion: In patients aged Conclusions: Pediatric patients with PIDD demonstrated a positive onboarding experience with Ig20Gly supporting the immunoglobulin replacement treatment with Ig20Gly in children.

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#7 Onboarding Experience of Patients With Primary Immunodeficiency Diseases Who Switched to Subcutaneous Human Immune Globulin 20% From Intravenous or Subcutaneous Immune Globulin 10%

Authors: Sudhir Gupta (1), Iftikhar Hussain (2), Kenneth Paris (3), Mark Stein (4), Barbara McCoy (5), Ping Wang (6), Christopher J. Rabbat (7), Leman Yel (6)

Introduction: Subcutaneous immune globulin (SCIG) 20%, Ig20Gly, was safe and efficacious in a phase 2/3 North American study (NCT01218438) in patients with primary immunodeficiency diseases (PIDD). This post hoc analysis assessed the onboarding experience with Ig20Gly by examining infusion parameters based on prestudy treatment (intravenous IG [IVIG] or SCIG). Methods: Patients aged ≥2 years who were treated with IVIG (IV-switchers) or SCIG (SC-switchers) immediately before study entry received IVIG 10% (Gammagard Liquid) at the monthly dose equivalent to their recent prestudy treatment for 3 months. All patients were then switched to once-weekly Ig20Gly (Cuvitru) for ~1 year. Results: Infusion rates of ≥60 mL/h/site for more than one infusion were reached by 58.8% (30/51) of IV-switchers and 65.2% (15/23) of SC-switchers; the median infusion number when patients first reached 60 mL/h/site was 3 for both groups. Infusions were completed in Discussion: For both groups (IV-switchers and SC-switchers), the maximum infusion rate of 60 mL/h/site was reached early in the Ig20Gly treatment period (median infusion number, 3). IV-switchers reported slightly longer infusion durations and used fewer infusion sites by volume compared with SC-switchers. Conclusions: Patients with PIDD who switched to Ig20Gly from IVIG or SCIG therapy demonstrated generally comparable infusion parameters.

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#8 Few Adverse Drug Reactions When Switching Primary Immunodeficiency Patients to the New Intravenous Immunoglobulin – Post-Hoc Analysis Data from the GAM-01/GAM-03 Studies

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Introduction: Intravenous immunoglobulin (IVIG) and subcutaneous immunoglobulin (SCIG) represent the standard therapy for many types of primary immune deficiencies (PID), secondary immune deficiencies (SID) and some autoimmune diseases. Although IVIG solutions are in general well tolerated, several studies show that differences in IgG products lead to differences in tolerability and that switching IgG preparations triggers an increase in adverse drug reactions. Methods: A post-hoc analysis of the data obtained during the study and subsequent extension study on the efficacy, pharmacokinetics and safety of a new intravenous immunoglobulin 10% (panzyga®) in PID patients was performed. 51 patients were switched from their previous IVIG solution to treatment with the new intravenous immunoglobulin 10%. Initial infusion rate was 0.01 mL/kg/min (60 mg/kg/h). Infusion rate could be increased to max 0.08 mL/kg/min (480 mg/kg/h) in a predefined pattern. Results: Only 2 out of 51 (4%) patients experienced a treatment related adverse event during the first infusion after product switch from their previous IVIG to the new IVIG 10%. After 3 infusions of the new IVIG 10%, a total of 5 patients (10%) had experienced mild ADRs. 4 of these 5 patients experienced further ADRs during the study suggesting that they might be generally prone to reactions. Discussion: Several authors reported an increased ADR rate in their patients after being switched from one mostly well tolerated product to a different one. Ameratunga observed ADRs in 14% of PID patients after switch, Dashti-Khavidaki noted ADRs

related to 34 IVIG infusions after switch. Conclusions: In the present study a switch from any IVIG solution to the new IVIG 10% triggered a low number of treatment related AEs

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#9 Biochemical characterization, pathogen safety and stability of a new 16.5% subcutaneous immune globulin product

Authors: Nicola Gelbmann, Senior scientist; Alfred Zoechling, Senior scientist; Christa Mersich, Senior scientist; Torben Schmidt, Head of Virus and Prion validation; Eva Turpel-Kantor, Deputy Medical Directory Immunology; Thomas Ernegger, Head of CQC stability; Katharina Pock, Senior Director R&D Plasma; Juergen-Roland Roemisch, Senior Vice President R&D Plasma

Introduction: Immunoglobulin concentrates have been successfully used for decades to treat patients with primary or secondary immunodeficiency disorders. This treatment has substantially decreased the frequency of life-threatening infections in these patients. Biochemical and physico-chemical properties of a newly developed maltose-formulated subcutaneous immune globulin (human) 16.5% liquid for the treatment of patients with primary immune deficiency (PID) and secondary immune deficiency (SID) were investigated. Methods: Molecular size distribution of monomers, dimers, polymers and fragments were determined (according to European Pharmacopeia (EP) monograph 8. 0) by size exclusion chromatography (SEC). IgG and IgG subclass concentrations were quantified by respective nephelometric methods. Functionality of the IgG was demonstrated by measurement of Fc function, opsonophagocytosis and Fc gamma receptor binding assays. Dynamic light scattering measurement and size exclusion chromatography were used to characterize the integrity of the IgG molecule. Measurement of potential procoagulant activity was done by NATEM and TGA (FXIa-like activity). The capacity of the subcutaneous immune globulin (human) 16.5% liquid, Cutaquig (Octanorm), manufacturing process to robustly inactivate/remove pathogens was investigated in spiking experiments with prions and viruses. Results: The SCIG 16.5% contains 96% of human IgG and is characterized by an especially low content of polymers and aggregates, low viscosity, low isoagglutinin titres, low IgA and IgM contents with a broad spectrum of antibodies against infectious agents. It has a distribution of immunoglobulin G subclasses closely proportional to that in native human plasma. In the final product, potential procoagulant activity is not detectable. Discussion: Virus safety of the SCIG 16.5% is obtained via a combination of three validated orthogonal methods as part of the manufacturing process: cold-ethanol fractionation, Solvent/Detergent (S/D) and pH 4 treatment. A substantial depletion of prions during the manufacturing process was demonstrated. The intended shelf-life is 24 months stored at +2°C to +8°C protected from light. Within its total shelf-life the product can be stored at room temperature up to +25°C for up to six months. Efficacy and tolerability of this new subcutaneous normal immune globulin 16.5% were shown in a clinical phase III study performed in 18 centers in North America and Europe. Conclusions: This study demonstrated the functionality and physico-chemical properties of the IgG molecules, the pathogen safety and stability of the new subcutaneous human normal immunoglobulin 16.5% indicated for the treatment of patients with PID and SID.

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#10 Pharmacokinetics, efficacy, tolerability and safety of a new subcutaneous human immunoglobulin 16.5% in primary immune deficiencies

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Introduction: Patients with primary immune deficiencies (PID) require life-long replacement therapy with immunoglobulins (Ig) to prevent severe infections and irreversible complications. In addition to safety and efficacy, tolerability and convenience of administration of Ig products are essential factors in patient acceptance. A new 16.5% Ig preparation was developed for subcutaneous administration (SCIG). Methods: Primary outcome was to assess efficacy of a new 16.5% subcutaneous human immunoglobulin preparation (Cutaquig) in preventing serious bacterial infections. Secondary endpoints included evaluating tolerability and safety, determining the PK profile, the number and rate of other infections and changes in quality of life measurements. A prospective, open-label, single-arm phase 3 study involving 61 patients was conducted at 18 centers in North America and Europe. PID patients who were stable on IVIG treatment for at least 6 months and with IgG trough levels ≥5.0 g/L underwent a 12-week wash-in/wash-out period consisting of weekly SCIG doses 1.5 times the previous IVIG dose (based on published conversion rates for marketed SCIG products), followed by a 52-week efficacy period (64 SCIG infusions in total). 22 of 61 patients enrolled had complete pharmacokinetic assessments at different time points: before the switch from IVIG to SCIG (PKIV), after the wash-in/wash-out phase (PKSC1) and at week 16 of the efficacy period (PKSC2).

Results: 61 patients (age: 2-73 years; mean age 32.2 years; 54.1% female) receiving a total of 3,497 SCIG infusions (0.135 g/kg/week in young children (≥2 years and Discussion: Treatment with the new 16.5% SCIG product was well tolerated and had no serious bacterial infections during the study period. Conclusions: This study demonstrated that the new subcutaneous human normal immunoglobulin 16.5% is well tolerated, safe and effective in patients with PID.

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#11 Efficacy, safety and tolerability of a new intravenous immunoglobulin 10% in primary chronic immune thrombocytopenia.

Authors: Olga Arbach1; Astrid Birgit Taumberger2; Stefan Wietek2; Nicole Farina3; Abdulgabar Salama4*.*

Introduction: This pivotal trial assessed the efficacy of a new human normal IVIG 10% in correcting the platelet count in patients with primary chronic ITP. Methods: In this prospective, open-label, multicenter, phase III study, patients received a daily dose (1 g/kg) of the new IVIG 10% (panzyga®) for 2 consecutive days. Twenty centers in Bulgaria, Czech Republic, Germany, India, Poland, Romania, Russia and Ukraine enrolled patients. Primary endpoint was the clinical response rate defined as increase in platelets to ≥50x109/L within 7 days after first infusion; secondary endpoints included alternate response rate definitions, time to response, response duration, platelet counts, regression of bleeding, and safety. Analyses were performed on the full analysis set (FAS) consisting of all enrolled patients with at least one post-baseline platelet concentration measurement. Analyses were conducted using statistical software SAS (version 9.1 or higher). Results: Forty patients were enrolled (57.5% male, mean age 36.7 years, range 18–72); the FAS comprised 36 patients. Clinical response was seen for 29 of 36 patients (80.6%; mean maximum platelet count of 237x109/L); in 18 of 23 patients (78.3%) with bleedings at baseline, haemorrhages completely resolved by day 8, confirming the efficacy of the IVIG 10%. Median time to response and response duration was 2 days and 14 days, respectively. The new IVIG 10% was well tolerated at a maximum infusion rate of 8 mg/kg/minute in all but one patient; adverse events were mainly mild to moderate in severity. The most frequent was headache (32.5% of patients) followed by pyrexia (22.5% of patients). The death of two patients was assessed as unrelated to study drug. Discussion: Treatment with the new IVIG 10% was well tolerated with adverse events of mainly mild and moderate severity. Treatment also showed a clinical efficacy in 80.6% of patients, which led to increased platelet count and decreased frequency of haemorrhages Conclusions: Overall, this new IVIG 10% is well tolerated even at high infusion speed and induces a rapid platelet count increase, thus decreasing the bleeding rate and severity of bleeding events.

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#12 The Feedback Loop: SCIg Administration Improvement & Troubleshooting Techniques

Authors: Tyler Kilcoin, Anna Majapuro-Hirvonen

Introduction: Literature discussing local site reactions from subcutaneous immunoglobulin treatment (SGIg), states that swelling, redness, and drug leakage are typical and tend to disappear over time. Literature review also presents multiple techniques to mitigate SCIg infusion site reactions: adjusting the needle length to reach the subcutaneous tissue, using a tricuspid needle cut in place of a lancet needle for improved comfort and less scarring, inserting needles dry, and using an adhesive dressing to secure the needles in place. The key to improving patient experience and expectation is to establish a new standard for what is considered “normal” or “expected.” Methods: Patient studies were undertaken with groups of 57, 30, and 14 patients using a Constant Pressure System (CPS). The CPS is a type of infusion device technology that maintains constant pressure throughout the duration of the infusion. The CPS’ sensitivity to infusion-site saturation and its ability to provide accurate feedback, helps to reduce the flow of medication into the patient, and facilitates several cycles of patient improvement with each infusion, with consideration to the ancillary supplies (all patients used HIgH-Flo subcutaneous safety needle sets), the flow rate, and infusion-site perfusion. Three different questionnaires were used to gather patient data from the infusions. 57 participants were asked to report any leakage of drug during and/or directly after infusion when removing the needles from the infusion site. 30 patients who participated in the High-Flo needle validation study were asked to report on placement process, needle sharpness, and changes in infusions with the new needles, and costs. 14 patients were previously using the Freedom60 syringe driver and other manufacturers’ needles, all of them reported undesirable site reactions. In the study, patients reported on outcomes using the HIgH-Flo in a questionnaire. Results: In the group of 57 patients, 95.2% reported being satisfied with the infusions even though 73.9% experienced at least one adverse site reaction. In the 30 patients study, all the patients reported improvement after switching needles. The 14 patients study reported diminished or disappearance of site reactions. Discussion: The local site reactions are manageable and can be overcome. The performance of the CPS is known and accepted (FDA). Based on the device’s technology, the flow rate will decrease if the pressure at the patient’s tissue increases (site saturation). By measuring the first half of the infusion and comparing it to the second, it is possible to get feedback of the level of site saturation. Other factors such as the use of proper ancillary supplies, a flow rate calculator, and the guidance from an experienced clinician, all comprise the infusion assessment. These factors and feedback help to evaluate the infusion success, and modify the parameters to enhance the patient’s experience. Conclusions: During SCIg administration, a CPS responds to the site-saturation levels, effectively regulating the amount of drug received in proportion to the tissues’ ability to absorb it. Along with this built-in feedback technology, caregivers and patients can tailor the infusions using the appropriate ancillary supplies and the tools suggested in the literature for accurate and repeatable infusions.

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#13 The Influence of BMI on Efficacy of Immunoglobulin Replacement Therapy in Primary Immunodeficiency Patients Treated in the Home

Authors: Allyson Checkley PhD (1), Loretta Kristofek RN BSN (1), Samantha Kile MS (1), William Bolgar PharmD (1), and Luqman Seidu MD (2)

Introduction: Guidelines recommend dosing Ig replacement therapy according to ideal body weight, but it remains debated on whether this is the optimal dosing approach for patients with extreme body mass index (BMI) (lean/obese). Evidence suggests that the physiological characteristics of lean and obese patients have the potential to influence immunoglobulin (Ig) pharmacokinetics, but it is unclear whether weight-based dose changes are clinically relevant. The objective of this study was to evaluate whether BMI affects the rate of infection in primary immunodeficiency (PID) patients and if it should be a major factor in dosing considerations. Methods: Ig dosage and route of administration, annual infection rate, and BMIs of adult (> 18 years of age) PID patients were analyzed from the IDEaL patient registry database. This is a prospective, observational registry study of patients receiving Ig replacement therapy in the home with one national home infusion company. All data were collected following patient consent from nursing and pharmacist completed clinical progress reports from July 2010-March 2018. All data are presented as the mean ± standard deviation, and significance was set at $P \leq 0.05$. Results: Patients receiving subcutaneous (SC) Ig therapy (n=202) had an average BMI of 29.6±7.5, and 41.1% of these patients were obese (BMI 30-39.9) or morbidly obese (BMI ≥ 40). Only 1.5% of these patients were classified as underweight (BMI < 18.5). Average annual infection rates for overweight, obese, and morbidly obese SCIg patients were 2.3±1.7, 3.0±2.3 and 4.2±2.4, respectively compared to 2.1±1.8 for normal weight patients (BMI 18.5-24.9). Differences were statistically significant between morbidly obese and normal weight patients (p Discussion: Obese patients represent a growing proportion of the US population. The complexity of excess body weight on Ig pharmacokinetics suggests that efficacy may be influenced by a patient's BMI. We found obese patients had higher infection rates than normal weight patients, and this was more pronounced in patients receiving SCIg. Further, these patients received, on average, lower doses of Ig which is reflective of dosing according to ideal rather than actual body weight. Nonetheless, not all obese patients required higher doses to achieve good outcomes, suggesting that patient factors other than BMI are important. Conclusions: Significantly higher infection rates in obese patients suggest that dosing adaptations with respect to BMI should be considered to ensure Ig efficacy is maximized and the risk of adverse events is reduced.

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#14 Qualitative and Financial Impacts of a Fifteen Minute Titration Protocol for IVIG Infusions

Authors: Betsy Rothley, RN, MSN, FNP

Introduction: BPL's FDA approved, fifteen minute titration protocol for Gammaplex 10%, has significantly impacted the home infusion market, both in cost of nursing services at home and in ambulatory infusion suites. This time modification has also impacted patient reported outcomes in symptom control and time commitment for therapy. Methods: Comprehensive education will be provided to both staff RN's, as well as nursing agency partners to train on a 15 minute titration protocol. 2 cohorts will be identified, with same primary diagnosis and indication for Immunoglobulin infused therapy. One cohort will receive the therapy with the 15 minute titration protocol. The other cohort will not. At the end of the infusion, each patient within the cohorts will receive the same dosing questionnaire. Questions will be on a 0-10 rating scale, allowing for a patient's objective report. Results: Questionnaires will be reviewed and results recorded by identified cohort. Nursing time per infusion will also be recorded. Discussion: The goal of the review will be to determine if the outcomes of the qualitative questionnaire, as well as the nursing time are significant enough to encourage one therapy infusion protocol over another. The review will also determine if the outcomes by themselves would drive utilization towards one product, as well as enhance nursing economics. Conclusions: All data to be collected and collated by September 30, 2018.

Author Affiliations: BioScrip Infusion Services

#15 Observational study to determine the tolerability of IVIG 5% for the treatment of patients diagnosed with Primary Immunodeficiency Disorders

Authors: Isaac Melamed, MD, Melinda Heffron, Ruth Dana

Introduction: Intravenous immunoglobulin (IVIG) is relatively safe, with headaches and fatigue being common side effects. Recently, a subset of patients with common variable immunodeficiency (CVID) and low C1 esterase inhibitor (C1-INH) and/or low C1-INH function (C1-INHF) has been identified. Preliminary data indicates that low C1-INH levels may play a role in adverse drug reactions (ADRs) noted in a subset of CVID patients receiving IVIG. We designed this observational study to determine if 5% IVIG may be an alternative for patients who experience ADRs on 10% IVIG. Methods: Patients who had previously received 10% IVIG completed 6 infusion visits using 5% IVIG. At each visit, ADRs and data from patient diaries were recorded. Immune-biomarkers, including C1-INH/C1-INHF levels, were also evaluated. Results: Fifteen subjects completed the study; 12 with CVID and 3 with hypogammaglobulinemia. Switching to 5% IVIG reduced the number of ADRs by 40%. There were also reductions in mean fatigue index, headache score, and neuropathic pain score from IVIG 10% to 5%. Patients experienced an increase in physical function, greater energy, less fatigue, higher emotional rating and an increase in social function. The mean C1-INH|C1-INHF on 10% decreased from 31 to 13 mg/dL (normal 21-29mg/dL)|91% to 59% (normal >67%) while on 5 % the mean C1-INH|C1-INHF decreased from 27 to 21mg/dL|89% -76%. Discussion: This study demonstrated that C1-INH/C1-INHF level changes play a role in the incidence of ADRs for IVIG therapy; and a subset of patients may be more susceptible to C1-INH/C1-INHF downregulation by IVIG 10%. Conclusions: Our findings indicate that a 5% IVIG

preparation may be an alternative to SCIG patients who develop ADRs on 10% IVIG [various preparations]. In our study there was a lower incidence of ADRs with no changes in premedication or infusion rates. In addition, our findings also corroborate our earlier study indicating that C1-INH downregulation plays a role in the level of IVIG ADRs in PID, including incidence and severity of headaches and fatigue. Furthermore, there may be a subset of patients more susceptible to C1-INH downregulation by 10% IVIG who may benefit from switching to a 5% IVIG. We propose further studies.

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#16 Is NIR Vein-viewing Technology Beneficial in Home Infusion Therapy?

Authors: Brianna Regan, BSN, RN, IgCN®, Amanda Walker, CPXP, BSN, RN

Introduction: Studies show safety of home infusion therapy compares favorably to hospital infusion treatments with respect to adverse events. Added advantages include avoidance of hospital acquired conditions – especially for immunocompromised patients - and patient preference for care at home. One universal infusion therapy challenge is difficult venous access (DVA), typically the result of long-term infusion therapy. Vein-viewing technology can aid in identification of more venous options for venous access or IV catheter placement. This study evaluates the use of near-infrared (NIR) vein-viewing devices for infusion therapy in the home care setting. Methods: Over a six (6) month period, a home infusion company utilized NIR vein-viewing devices (VeinViewer®) to assess each patient's veins to determine the appropriate location for venipuncture. Catheter placement and therapy followed the standard of care. Care delivery parameters - including number of stick attempts, venipuncture location, device usage, time to catheter placement – and patient satisfaction were measured. Nurses collected data at care delivery via computer-based system (Envoy) for patient metrics. Results: Patients on long-term infusion therapy frequently tell nurses where to access their veins. This 'self-direction' by patients impacted the resulting data. In 75 patient encounters, 70 patients received IgG therapy and 5 received other therapy. Peripheral intravenous catheter (PIVC) was placed under the NIR light for 20 encounters, the vein was identified with NIR light and light removed for PIVC placement in 30 encounters and a 'known vein' was used in 25 encounters. Additional data was collected to evaluate whether an unknown vein was accessed after the 10th subject. Overall, a 'known vein' was accessed in 34 encounters and an unknown vein was accessed in 31 encounters. When asked if the NIR light device positively impacted the patient visit, 24 clinicians indicated Yes, 28 were neutral and 23 indicated no. Discussion: The majority of this organization's participating patients were established prior to initiation of the study. Clinicians noted throughout the study that DVA patients welcome vein-viewing technology more than patients with easy to access veins. In the case where the patient has easily accessible veins, the clinician and patient generally reported that technology was either "not necessary" or "added a variable that made access more challenging". For new home care patients and patients with dark skin, NIR light device was helpful. Patients also liked seeing their veins to understand other vein access options. Conclusions: Use of near-infrared vein-viewing technology in home infusion therapy provided both patient and clinician benefits, especially for difficult venous access (DVA) patients.

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#17 Interim Analysis of the Global Post Authorization Safety Study of Hyaluronidase-Facilitated Subcutaneous Immunoglobulin 10% Treatment in Patients With Primary Immunodeficiency Diseases

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Introduction: Recombinant human hyaluronidase (rHuPH20)-facilitated subcutaneous immunoglobulin (SCIG) 10% is an approved IG replacement therapy (IGHy) for patients with primary immunodeficiency diseases (PIDD). To acquire long-term safety data on IGHy, and assess prescribed treatment regimens and administration in routine clinical practice, a global postauthorization safety study (PASS) is being conducted. Methods: This is an ongoing prospective, non-interventional, open-label, uncontrolled, multicenter study initiated in the United States in November 2015 to assess local and systemic effects of IGHy (HyQvia) within a routine clinical setting. Patients aged ≥16 years with PIDD who have been prescribed and/or have started IGHy are eligible for enrollment. Patients are followed according to standard clinical practice and their treatment regimen is at the discretion of the treating physician. The presence of anti-rHuPH20 antibody titers is evaluated on a voluntary basis. Results: As of August 2017, 175 patients had been enrolled at 26 US study sites. There were no serious adverse events (AE) which were deemed treatment related. Sixteen patients experienced a causally related non-serious local AE (9.1%; 0.43 events/patient-year, 0.07 events per infusion) and 25 patients experienced a causally related non-serious systemic AE (14.3%, 0.88 events/patient year, 0.14 events per infusion). Of the 113 patients with immunogenicity data, 7 had ≥1 positive binding antibody test to rHuPH20 (titers ≥1:160); no neutralizing rHuPH20 antibodies were detected. Discussion: These prospectively-collected data indicate that IGHy is well tolerated in routine clinical practice in patients with PIDD. Conclusions: IGHy is well tolerated with no treatment-related serious AEs and has not been associated with neutralizing anti-rHuPH20 antibodies in patients with PIDD.

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#18 Overall and Pediatric Patient Preferences for Recombinant Human Hyaluronidase (rHuPH20)-Facilitated Subcutaneous Infusion of Immunoglobulin G in Patients With Primary Immunodeficiency Diseases

Authors: Diane Ito, PhD (1), Lisa M. Meckley, PhD (2) , Todd Berner, MD (3), Leman Yel, MD (2)

Introduction: Among the available routes of immunoglobulin (IG) administration for patients with primary immunodeficiency diseases (PIDD), recombinant human hyaluronidase (rHuPH20)-facilitated subcutaneous (SC) infusion of immunoglobulin G (IGHy) offers the option to self-administer at home on a 3-4 week basis. This analysis assessed treatment preferences of patients who switched to IGHy after previously receiving intravenous IG (IVIG) or conventional SCIG during the phase 3 study. Methods: In this prospective, non-controlled study, patients with PIDD were treated with IVIG for 3 months, followed by IGHy (HyQvia) at 3–4 week intervals for approximately 12 months. At the end of study, patients completed a preference questionnaire that assessed preference to continue IGHy and a range of treatment attributes, such as convenience, infusion time and frequency of administration. Treatment attributes were measured using a 5-point Likert scale (from "dislike very much" to "like very much"). Patients aged ≥14 years completed the questionnaire themselves and pediatric patients aged 2–13 years had a caregiver/parent complete the questionnaire. Results: A total of 87 patients with PIDD aged ≥2 years were enrolled in the study; 69 completed the questionnaire, of which 13 were aged Discussion: Real-world patient experience with IGHy may elucidate how these patient preferences might impact treatment adherence. Conclusions: Most patients, including children, preferred IGHy over IV or conventional SCIG administration.

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#19 Case Study: Evaluating the use of subcutaneous immune globulin therapy for the management of seizures in a pediatric patient with Lennox-Gastaut syndrome

Authors: Christine Miller, PharmD; Barbara Prosser, RPh; Monique Nelson, RN; Drew Doyle, RPh.

Introduction: BB is a 10 year-old male who is diagnosed with Lennox-Gastaut syndrome (LGS). His seizures began at age 5, and the types of seizures he experiences include grand mal, atonic, absence, and gelastic seizures. The seizures have been refractory to multiple oral anti-epileptic medications (AEDs). Vagus nerve stimulation (VNS) and maximum doses of lacosamide have helped to reduce the number of seizures the patient experiences, but, as of October 2017, BB was still experiencing at least one grand mal seizure and 4 to 12 gelastic seizures each day. BB also suffered from behavioral issues. His physician described his behavior as impulsive, intrusive, and defiant. At school, BB would need continuous one-on-one teacher interaction in order to perform his work, but he was eventually withdrawn from the classroom due to the amount of seizure activity he was experiencing. The discovery of low IG levels in this patient led to the attempt of using immune globulin (IG) therapy to manage his seizure activity. The purpose of this case study is to describe the use of IG therapy for the management of seizures in a pediatric patient diagnosed with Lennox-Gastaut syndrome. Methods: A multi-disciplinary team conducted a retrospective review of the patient's medical records, including the prescriber's office visit notes and assessments performed by clinicians within this home infusion organization during each contact with the patient's caregivers. Results: In October 2017, BB was initially prescribed immune globulin 20% liquid 10 grams subcutaneously every other week. In the week following the commencement of IG therapy, BB experienced 9 grand mal seizures. The frequency of the IG therapy was subsequently increased to 10 grams once weekly. By February 2018 the number of grand mal seizures the patient experienced had decreased to one per week. An improvement in BB's behavior and physical development were also observed, and he is able to participate in a classroom environment again. The dose of IG was increased to 15gm subcutaneously once weekly in February 2018. As of June 2018, BB continues to experience an average of one grand mal seizure per week, though he rarely experiences any gelastic seizures. The patient has expressed feeling "relaxed" since the frequency of his seizures has been reduced. Discussion: Treatment of Lennox-Gastaut syndrome with IG therapy is considered to be off-label use. Though BB's seizure activity has been significantly reduced, it has not been completely eliminated. He still experiences seizures with certain triggers, such as lack of sleep, emotional excitement, and change in weather. Adjustments to BB's oral AEDs and VNS are also being made. The dose or frequency of IG therapy may need to be adjusted in the future as the patient continues to grow and develop. Conclusions: Since the initiation of IG therapy for Lennox-Gastaut syndrome, BB has experienced a substantial reduction in seizure activity as well as an improvement in his quality of life. The number of grand mal seizures BB experiences has been decreased from at least one per day to one per week, and the occurrence of gelastic seizures has nearly been eliminated.

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#20 What is the "New Norm?" A prospective study plan for clinically managing teaching capabilities/utilization of nursing resources for new, "on-label" use of SCIG for the treatment of CIDP.

Authors: Cathy J. Taggart, RN, CRNI®, IgCN®, MBA

Introduction: With the recent roll out of on-label use of SCIG administration for those diagnosed with CIDP, we are experiencing an entirely new chapter in the care of this population. i. This new challenge offers a unique opportunity to provide care in a cost effective, productive manner. ii. The industry standard currently espouses teaching the home SCIG patient for self-care. We have yet to embrace what is the "new norm: what does taking care of this population, already challenged by escalating nursing costs, not always reimbursable, for a very needed therapy and population, look like? iii. Determining what this landscape will look like, providing this care, safely, and cost efficiently, will be best determined by utilizing

our education, screening, monitoring, and support the care of this population in a cost effective manner. iv. This organization is excited to work with industry providers, to research this need, report back to this industry, in a timely fashion, what this landscape will look like!

Methods: Our organization has developed an algorithm to represent this prospective study and what it will look like. ii. Population studied will focus strictly on those patients' serviced by our organization, and its contracted partners, specifically with the on-label designation for use of SCIG in CIDP patients. We feel this will give us a significant sample size. iii. There will be several arms within the study: a. Questionnaire upon referral b. Internal education and training, to screen further, i.e. insurance agencies, third party administrators, CMS. c. Outcomes data from recent roll out of new clinical documentation program. iv. Patients failing to tolerate SCIG, will be removed from study, but side effect profile will remain. v. Intent is to determine new industry standard for teaching methods, outside the box education ideas, and overall understand the costs this type of program may incur for the industry, and to be able to plan, and work toward optimal nurse productivity. vi. We intend to Includes statistics where appropriate such as teaching visits needed, patient tolerance, safety of self-infusion, etc. vii. Will monitor this for one year, with the intent to report out preliminary results at IGNS 2019. Results: i. Determine, by a variety of methods, what the "new norm" for nursing educational needs will be for this population and likely similar diagnoses in the future. ii. Will present on specific algorithm within the poster iii. Presentation in narrative fashion, saving all visual elements for the poster iv. Interested in feedback from clinicians within our space. Discussion: Our organization is excited to share lesson(s) learned from project/study at IGNS 2019 ii. Goal is to determine safest, most cost effective, best use of nursing commodities to adapt best practices for nursing within this population. iii. We feel this could help the industry as a whole to better define what the reality of servicing this population will cost, in manpower, efforts and commitment. Conclusion: Our organization proposes to study this patient population and their education needs to provide safe and reasonable administration of SCIG.

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#21 Port-A-Cath Usage in a Private Allergy and Immunology Practice

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Introduction: Since 1982, Port-A-Caths have been implanted beneath the skin, into the veins, for the injection of intravenous medications and for laboratory studies. Port-A-Caths were initially used for chemotherapy administration for the treatment of cancer, however the use of ports has widely increased to treat a variation of diseases. Patients that require intravenous Immunoglobulin G (IVIG) need treatment every 3-4 weeks. Hereditary Angioedema patients receive C-1 esterase Inhibitor and/or fresh frozen plasma (FFP) as needed for HAE prophylaxis or exacerbations. Due to the unpredictability of HAE exacerbations, some HAE patients require port access daily and some require extended periods of port access. The purpose of this abstract is to assess the safety, effectiveness, and patient outcomes of Port-A-Cath utilization in patients with Common Variable Immunodeficiency (CVID) and Hereditary Angioedema (HAE) in our practice. Methods: We compiled data using a population group of 20 established patients within this practice that require a port for the administration of intravenous medications. Results: Out of these 20 patients, 13 patients receive IVIG. All 13 IVIG patients have their ports accessed by our professional nursing staff within the practice. There are 7 patients that receive treatment for HAE. All 7 of these patients self-access and administer their medications at home after undergoing training by professional nursing staff. The complications that were seen with frequent port access included lack of blood return in 45% of patients which was resolved with Cathflo Activase, migration of port tail in 5.0% of patients which resolved with re-adjusting the port tail, leaking of medication in 10% of patients which resolved with increasing the huber needle size, and infection at the port site in 10% of patients causing sepsis which resolved with IV antibiotics. In this cohort, 35% of patients required port replacement, 25% of those patients being HAE and 10% IVIG. There were 2 HAE patients that had more than one replacement and 1 IVIG patient who needed a replacement. Some of the factors that may contribute to complications include an environment that is not sustainable for effective port care, inadequate port care including maintaining sterility, and improper placement. Discussion: There are many factors that may contribute to port complications and it is important to determine if the benefits outweigh the risks with each individual patient. Overall, we recommend that health care staff who are inquiring about long term port placement in patients consider the frequency of port access, the environment that the port is accessed in, the receptiveness of patient education and training, and the overall risk to the patient before determining if a port would be beneficial in their care and treatment. Conclusions: We conclude that an increased rate of infections and port replacements may be a result of more frequent port accessing. Our data shows that long-term Port-A-Cath placements, in general, are beneficial to our patients care and treatment. HAE patients specifically have verbalized that a port has increased their quality of life and has made the ability to independently manage their disease feasible.

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#22 A PROSPECTIVE, OPEN-LABEL, MULTICENTER STUDY OF THE EFFICACY, SAFETY, TOLERABILITY, AND PHARMACOKINETICS OF PLASMACAP IG (IVIG) IN ADULTS AND CHILDREN WITH PRIMARY IMMUNODEFICIENCY DISEASES (PIDD)

Authors: Jacinthe Guindon, MSM, Mark Krause, BSc, MBA

Introduction: We are presenting preliminary unconfirmed safety data from the adult portion of the Phase III multi-center study of PlasmaCap IG conducted in adults and children with primary immunodeficiency diseases (PIDD). Clinical Trial Identifier #NCT03238079. Methods: Patients enrolled at the time of the review included 48 adults aged

17 to 70 and 1 child aged 2 to 500 mg/dL at screening. All patients enrolled received the same dose they were receiving at the time of screening. An additional 23 children are expected to be enrolled on study. Results: At the time of the last safety review (June 2018), 48 adult subjects and 1 pediatric subject were enrolled. The adults were treated with PlasmaCap™ IG for a median of 162 (101-228) days; no subjects have completed the study but 5 withdrew between visits 3 and 6, due to personal reasons (n=3), generalized pruritus (n=1), and colon cancer diagnosis (n=1). There were no study drug-related serious adverse events. The adverse reaction rate was 0.129 per infusion; almost all were mild (0.09 per infusion; n=28), and the remainder were moderate (0.035 per infusion; n=11) and severe (0.003; n=1). 98.5% of the 309 infusions were completed without any administration changes, such as slowing, interrupting, or stopping the infusion. Discussion: Conclusions: In our ongoing clinical study, these preliminary data appear to indicate PlasmaCap IG is safe and well tolerated in the treatment of patients with PIDD.

Author Affiliations: Evolve Biologics

#23 Optimizing Patient Comfort during Delivery of Subcutaneous IgG (SCIg)

Authors: N. de Beer, J. Barbrie, C. Gutierrez, P. Lambert

Introduction: Studies have shown that home-based subcutaneous immunoglobulin replacement therapy achieves acceptable IgG trough levels and better health related quality of life and treatment satisfaction. While patients and caregivers are expressing a growing preference for home-based treatment, there remains a need to improve patient experience and comfort during delivery of SCIg to further enhance treatment satisfaction and maintain compliance. This study discusses the insertion and extraction of needle sets during self-administration of SCIg and highlights how treatment satisfaction can be improved with specific attention to needle penetration force, use of an insertion device and improved extraction techniques. Methods: Needle penetration force through various membranes is well accepted as a means to establish needle sharpness with a direct correlation to perceived pain. Tests were conducted to compare the dynamic penetration force of EMED's Soft-Glide needle sets (27ga and 24ga) which has a proprietary coating, with a selection of other market needles according to protocols based on ASTM F3014 test standard. Other features that contribute to needle sharpness and perceived pain were evaluated such as bevel design, needle coating and insertion or extraction techniques. In this regard, needle set features of EMED's OPT-Flow sets and inserter, as well as Soft-Site dressings were also evaluated. Results: Test results showed that the author's coated needle sets require the lowest amount of penetration force, both during initial puncture and the rest of the needle insertion. One comparison revealed that a different manufacturer's 26ga needle set required as much as 115% higher penetration force than the coated 27ga needle sets. On average, competitor's needles required between 12%-55% higher penetration forces for 27ga and 26ga respectively. The author's coated 24ga sets also performed better than other 24ga sets, and even other 26ga sets, requiring 40% and 30% less penetration force respectively. Discussion: Test results indicate that although the needle gauge influences penetration force, additional design features such as the quality and length of the bevel cut, and needle coating can reduce the penetration force and related pain. Furthermore, features on the needle set wings and hub that facilitates 90° insertion and extraction techniques reduces additional trauma to the injection site tissue. Some patients may benefit from using a needle inserter to assist in this process. Needle set removal can also be tedious and painful when dressings stick to the needle set wings, causing additional aggravation to site tissue. The dressing evaluated in this study demonstrated improved features allowing less adhesion and interference to the needle set during its removal and extraction. Conclusions: The main goal with any IgG replacement therapy is to achieve normalized serum IgG levels and, thereby, a reduced frequency and severity of infections. For home-based therapy, this requires patient compliance and treatment satisfaction. This study has shown that through improved needle design, penetration forces and expected pain during insertion is reduced. In addition, features on the needle set wings and hub are essential to facilitate a perpendicular insertion and extraction from the skin to reduce pain and improve patient comfort.

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#24 Improvement in myasthenia gravis symptoms with the use of subcutaneous immune globulin

Authors: Christine Miller, PharmD; Barbara Prosser, RPh; Monique Nelson, RN; Drew Doyle, RPh.

Introduction: JS is a 59 year-old female who was diagnosed with myasthenia gravis (MG) in 2005. Her major symptoms include fatigue, dyspnea on mild exertion, neck weakness and spasms, diplopia, and left ptosis. Past therapies used for JS's MG symptoms include rituximab, with which she developed serum sickness, and multiple intravenous immune globulin (IVIG) formulations given at a dose of 2 grams/kilogram once every 4 to 6 weeks. The IVIG infusions would help to improve her MG symptoms; however, the patient experienced serious adverse effects and would often require hospitalization due to aseptic meningitis and intractable vomiting. From 2015 through 2017, the patient experienced 15 hospitalizations. Within that same time period, JS also received 10 trigger point injections for neck spasms. In July 2017, due to the need for routine hospital admissions for the severity of adverse effects, the patient expressed her wishes to stop all treatments for MG. Because subcutaneous immune globulin (SCIG) is associated with less systemic adverse effects than IVIG, the patient was willing to attempt SCIG therapy. The purpose of this case study is to evaluate the effectiveness of subcutaneous immune globulin therapy in managing this patient's MG symptoms. Methods: A retrospective review of the patient's medical documentation was conducted by a multi-disciplinary team. The medical documentation comprised of office visit notes from the patient's neurologist, endocrinologist, and optometrist, as well as assessments conducted by a clinician within this

home infusion organization from July 2017 through July 2018. Results: In July 2017, the patient began receiving immune globulin (IG) 20% liquid 11 grams subcutaneously twice weekly. Since the initiation of subcutaneous IG therapy, the patient has experienced one hospitalization, which was a scheduled admission for a surgical procedure unrelated to myasthenia gravis. Additionally, the patient has required zero trigger point injections for neck spasms. JS did experience one MG exacerbation in November 2017. Documentation from the patient's optometrist describes a dramatic improvement in JS's vision along with the appearance of her eyelid and ocular structures. Her fatigue and dyspnea have also drastically improved, and she is able to take nightly walks for exercise. The patient describes this therapy as life-changing. Discussion: Because IG 20% does not carry an approved indication for MG, there were difficulties in obtaining insurance authorization for its use in this patient's case. Multiple subcutaneous infusions per week are required for this patient due to the high dose and high volume of IG necessary for the treatment of MG. The patient did experience local infusion site reactions initially until the infusion rate and number of infusion sites were tailored to this patient's tolerance. Conclusions: Since beginning SCIG infusions, JS has experienced an improvement in each of her major MG symptoms. The use of SCIG has helped to reduce the number of hospitalizations this patient experiences and has eliminated the need for trigger point injections. Despite the requirement to infuse SCIG more frequently than the IVIG infusions this patient had received in the past, SCIG has made a positive impact on this patient's quality of life.

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#25 PlasmaCap EBA™, an innovative method of isolating plasma proteins from human plasma

Authors: David Miller, BASc; Gillian Vanderlee, BEng, MASc; Maggie (Chunmei) Qin, M.S; Olivier Vaute, Ph.D, MBA; Mark Krause, BSc, MBA

Introduction: EBA (Expanded Bed Adsorption) chromatography technology has the potential to be the first major advancement in plasma protein extraction in over 75 years. It allows for isolation and purification of Immunoglobulin G (IgG) and other plasma-derived proteins at improved levels of quality, consistency, and purity compared to conventional methods, such as cold ethanol fractionation. It also provides a considerable cost- and time-saving manufacturing process. This technology offers greater efficiency in isolating plasma proteins and will allow us the potential to deliver improved therapies to patients with rare diseases. Methods: PlasmaCap EBA chromatography is the technology used by our Company to extract and purify IgG, as well as other valuable therapeutic proteins, from donor plasma, improving the yields of many commercially relevant proteins from each liter, as compared to the legacy technology in the plasma products industry. Results: EBA chromatography has been used in various commercial applications in the food and biologics industry at scales significantly larger than contemplated for our planned commercial facility. This technology has also been used for protein purification as an intermediate step in the development of numerous other clinical products. EBA chromatography is performed under mild conditions, without the use of ethanol. This reduces the potential for denaturation of proteins and allows for higher yields by removing inefficient processing steps. To our knowledge, no other commercial fractionators use a process similar to EBA. Discussion: In contrast to the legacy plasma manufacturing process involving cold ethanol fractionation, chromatography separates plasma-based proteins directly by specifically targeting the unique characteristics of each protein of interest (molecular size, charge or known interactions with specific molecules) through the use of a selective chemical matrix. By utilizing a more precise and accurate targeting mechanism for specific proteins, chromatography's gentler chemistry can result in superior product yields compared with cold ethanol fractionation, while allowing the remaining material to pass through unchanged. The remaining material can then be passed through subsequent columns to capture additional proteins. This is in contrast to traditional cold ethanol fractionation, where labile proteins can be denatured and scattered across other fractions beyond the one targeted. Our technology is a further improvement on traditional packed bed chromatography by utilizing EBA chromatography. EBA affords additional benefits over traditional chromatography by operating at ambient pressure levels rather than high pressure, handling high viscosity starting materials such as plasma and requiring fewer steps and less handling between manufacturing runs. As such, EBA chromatography is more amenable to large scale production than traditional chromatography, which has been shown in the production of other biological and food products. Conclusions: Plasma-derived products manufactured using this new technology will have competitive product profiles compared to existing products in the market. We also believe that in the future, our EBA technology will bring additional innovative plasma-derived products to the patients who need them.

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#26 Practical Application Of Subcutaneous Immunoglobulin For Maintenance Treatment In CIDP: The PATH Study

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Introduction: Patients with chronic inflammatory demyelinating polyneuropathy (CIDP) often require long-term intravenous immunoglobulin (IVIG) maintenance therapy. IVIG is associated with systemic adverse events (AEs) such as headaches, and can often require premedication. Subcutaneous immunoglobulin (SCIG) offers an alternative administration option with anticipated improvements in patient convenience and safety. The PATH study evaluated a 20% SCIG solution as a maintenance treatment and determined common SCIG infusion parameters for use in clinical practice. Methods: In a randomized, double-blind study, patients (n=172) received 0.2 or 0.4 g/kg a 20% SCIG solution (IgPro20) weekly, or placebo. The primary outcome was percentage of patients with CIDP relapse (determined by adjusted Inflammatory

Neuropathy Cause and Treatment (INCAT) score) or withdrawal during 24 weeks of treatment. Common infusion parameters for clinical practice were recorded, as were AEs per infusion. Treatment satisfaction was assessed via the Treatment Satisfaction Questionnaire for Medication (TSQM). Results: Both IgPro20 doses significantly reduced the percentage of CIDP relapse/withdrawal versus placebo, with most subjects preferring SCIG over their previous IVIG. Infusion of IgPro20 or placebo was performed over two sessions a week each averaging 1 hour and 20 minutes in the abdomen, thighs, and/or hip. Patients infused at a median of 4 (range: 1–8) sites in parallel; the number of sites was dependent on total volume administered and subject preference. Subjects infused a median of 4 g/20 mL per site (max. 10 g/50 mL), with a median infusion rate of 20 mL/hr/site (max. 50 mL/hr/site). The rates of AE were low (0.06/infusion), the most frequently occurring AEs were local site reactions (94.5% mild, 5.5% moderate), such as itching or swelling. The total rate of systemic AEs was 0.04 per infusion. Neither infusion rate nor volume had an effect on systemic or local reactions. Responses from the TSQM showed that the majority of subjects (88%) found SC administration of IgPro20 or placebo somewhat easy, easy, very easy or extremely easy. Discussion: SCIG administration was associated with a lower incidence of CIDP relapse and withdrawal for other reasons. The majority of subjects preferred SCIG over their previous IVIG therapy. SCIG can be performed over multiple sites, depending on subject tolerability. These results provide a basis for future SCIG infusion applications. Conclusions: In summary, IgPro20 is a flexible and efficacious maintenance therapy for CIDP, well tolerated at a range of infusion volumes and rates. Subcutaneous may be a preferred route of IgG administration for many patients.

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#27 Pharmacist completed infliximab risk assessment improves safety in the selection of home as site of care.

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Introduction: The development and implementation of a therapy specific risk assessments and infusion rate protocols can improve patient safety. Determination of premedication need based on patient history of tolerance and comorbid conditions which have potential to impact tolerance and adverse event risks are evaluated during the determination of home as an appropriate site of care. Additionally, infusion tolerance and safety may be further enhanced through utilization of titration guidelines based on the tolerance of the patient to therapy. Methods: This organization developed a risk assessment for patients receiving tumor necrosis factor-α blocker therapy (TNF-α blocker) which would further provide clinical guidelines for home as the appropriate site of care. Our process included the evaluation of our current immunoglobulin risk evaluation and modeling a unique risk assessment around those variable specific to infliximab and golimumab. Prior to the implementation of this risk assessment, 2 years of this organization's retrospective data around infusion reactions was studied to determine. A total of 540 infusions were reviewed and the number of mild, moderate and severe reactions recorded for maintenance doses of TNF blocker therapies. Results: The development of the risk assessment and additional infusion numbers allowed for the development of an infusion rate protocol which is based on a titration schedule determined by patient tolerance. With a infusion count of 520, no severe reactions were reported to have occurred, while a total of 1 moderate and 20 mild reactions were reported – a severe AE rate of 0%, a moderate AE rate of 0.2%, and a mild AE rate of 3.8%. This is consistent with or below adverse event rates reported on infliximab patients in multiple studies, across various sites of care.2-7 Two patients, over the course of their treatment, elected to return to an infusion suite to receive their infusions. One patient did not want medication delivered to the home or an alternate address, while the other did not want people in the home. Discussion: The use of a therapy specific risk assessment for initial determination of patient appropriateness for therapy and the site of care provides level sets both prescriber and patient expectations. It also allows for a clinical threshold to be better understood for moving a patient into home for treatment based on previous history and associated risk factors. Conclusions: Biologic therapy can be safely administered in the home when a systematic approach is utilized. The use of a therapy specific risk assessment for initial determination of patient appropriateness for therapy and the site of care provides a level set for both prescriber and patient expectations. It also allows for a clinical threshold to be better understood for moving a patient into home for treatment based on previous history and associated risk factors.

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#28 Creation of a Comprehensive SCIG Resource Tool for Clinicians

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Introduction: The subcutaneous (SC) route of immune globulin (IG) administration has been associated with a number of benefits for the patient, including a steadier serum Ig level; self-administration at the patient's convenience; and fewer systemic side effects compared to intravenous (IV) IG (Kobrynski, 2012). The number and variety of SCIG products available on the market today provide many options for treatment, necessitating a

thorough understanding by home infusion clinicians of how each drug may address a patient's specific needs. While SCIG administration guidelines are available from SCIG manufacturers specific to their individual products, a comprehensive resource that would facilitate comparison of all approved SCIG products and their considerations for use, has not been published. Methods: The goal of this project was to create a comprehensive, evidence-based SCIG resource tool to facilitate clinician review of available SCIG products, their administration considerations, as well as troubleshooting of side effects and prevention strategies. Methods included: (1) Research evidence-based best practice for management and prevention of SCIG-related side effects, and current product-specific instructions for use. (2) Survey clinicians active in the care of SCIG patients, both nurses and pharmacists, for their perspectives regarding current SCIG resource tools, the most frequently encountered SCIG complications and strategies to resolve them. (3) Identify trends in clinical practices for SCIG administration and troubleshooting from survey responses. (4) Develop clinical resource tool to address SCIG products, administration, and troubleshooting best practices. Results: Survey tool was developed and sent to 157 nurses with a 74% response rate. SCIG infusion experience of respondents ranged from 1 to 15 years, with an average of 5 years. Respondents identified a lack of concise and/or comprehensive information related to SCIG administration as a current challenge when providing SCIG therapy (n=22). The top four categories of unmet needs/learning opportunities identified by survey respondents included: drug/dose calculations; SC site selection; side effect troubleshooting/management; and training aids. Discussion: A concise evidence-based resource tool was developed to reflect manufacturer's instructions for use, published research findings, and clinician survey responses. The tool addresses a range of clinical considerations, including: administration frequency; IV to SC conversion rates; equipment and supplies for administration; SC site selection and preparation; therapy discontinuation; and troubleshooting of commonly reported side effects and administration-related issues. Keeping the tool updated as new drugs, supply or equipment options enter the market, and with newly published research findings, will be essential for the tool to remain relevant as a clinician resource. Conclusions: A comprehensive, evidence-based SCIG resource tool was developed and will be available for general access through IGNS.

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