The ORBCoN Report
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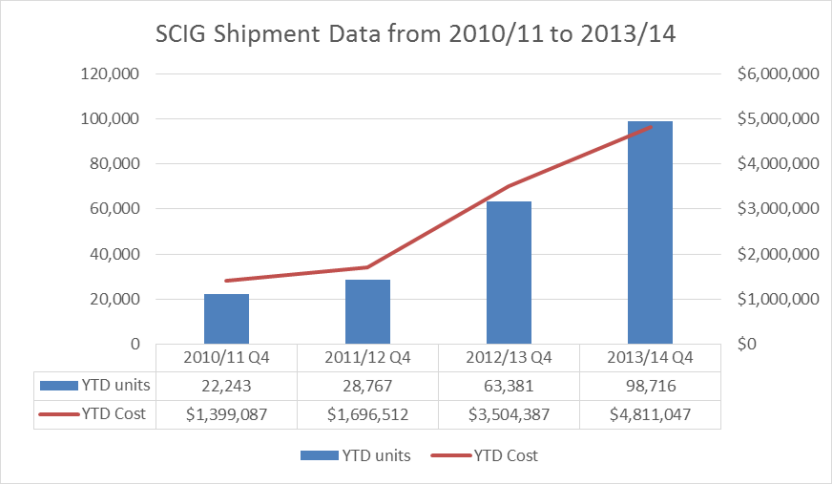
in this Issue

What's New at ORBCoN?

ORBCoN is pleased to provide this winter edition of The ORBCoN Report. The theme of this edition of the newsletter is 'Home Infusion Products'. Beginning in about 2009 certain blood products began to be made available to Ontario patients for self-administration. Both subcutaneous immune globulin (SCIG) and C1 Esterase Inhibitor are available for patients to self-administer at home. Some patients prefer this option that provides more freedom to receive their treatment on their own schedule and in their own home or while traveling. Fewer restrictions often results in improved quality of life for these patients as well as improved safety.

In this edition of the ORBCoN Report, we provide articles on the products, how they are ordered and issued and from a patient's perspective, the benefits of using them. To provide support for hospital transfusion medicine laboratories to order, store, issue and track these products, ORBCoN has developed a Home Infusion Toolkit. This resource is now available and can be found on the transfusionontario.org website. We hope that you will find this report both helpful and informative.



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Home Infusion Therapy Products Available in Canada

Adriana Martin, Infusion Specialist – CSL Behring

Subcutaneous immunoglobulin (SCIG) therapy was first initiated in the late 1970's by Berger and colleagues to permit administration of large volumes of immunoglobulin (Ig) versus painful intramuscular injections. In the 1980's IVIG preparations given once monthly were introduced and replaced the more frequent SCIG route. Serious systemic reactions with IVIG resulted in some investigators reconsidering SCIG treatment and returning to using IVIG products. New, more concentrated, SCIG products have been shown to decrease systemic reactions and offer patients increased quality of life[1].

The most frequently encountered patients receiving SCIG are those with Primary Immunodeficiency (PID). These patients require life-long Ig replacement therapy to prevent severe and reoccurring infection. Ig is traditionally administered intravenously (IVIG) on an outpatient basis, although in some Scandinavian countries subcutaneous administration of Ig as home self-infusion has become the predominant mode of delivery. SCIG therapy consists of smaller doses at more frequent intervals than IVIG resulting in smoother, more consistent serum Ig concentrations with greater stability in levels between infusions compared with IVIG.

A number of studies have reported that SCIG was comparable or superior to IVIG in maintaining sufficiently high trough levels of IgG[2,3,4]. SCIG therapy has been shown to be as effective as IVIG in preventing infections and has a better safety profile, with fewer systemic side effects. In patients with difficult vascular access and intolerable side-effects with IVIG therapy, SCIG therapy may be the only treatment option. While local tissue reactions are common with SCIG, they are usually mild, tend to improve over time and typically do not interfere with therapy.
Switching to SCIG therapy from IVIG can lead to significant improvements in health-related quality of life, appears to be more convenient for the patient, and can make it easier for the patient to travel. Selected patients can be expected to benefit greatly from SCIG therapy.

Hereditary Angioedema (HAE) is a very rare and potentially life-threatening genetic condition that occurs in about 1 in 10,000 to 1 in 50,000 people[5,6]. HAE symptoms include episodes of edema (swelling) in various body parts including the hands, feet, face and airway. In addition, patients often have bouts of excruciating abdominal pain, nausea and vomiting that is caused by swelling in the intestinal wall. Airway swelling is particularly dangerous and can lead to death by asphyxiation[7].

C1-Inhibitor (C1-INH) is a glycoprotein that regulates mediators of inflammation and vascular permeability. Angioedema (swelling) occurs because of local increases in capillary permeability causing extravasation (movement) of fluid into the deeper cutaneous or submucosal layers of tissue[8,9].

"Caring for patients with HAE is challenging due to the complexity of this disease. The care of patients with HAE in Canada is neither optimal nor uniform across the country. It was identified that early treatment likely leads to more rapid symptom resolution. Observational studies have suggested that early treatment can be efficacious in reducing the duration of an attack in some patients. Therefore, despite the absence of a high level of evidence, expert opinion was strong endorsing early treatment in an attempt to reduce morbidity and likely mortality. Because of the potential barriers in accessing therapy in a timely manner, patients should be trained on how to self-administer therapies appropriate for the treatment of acute attacks of HAE."[10]

In October 2014, The Canadian Hereditary angioedema guideline was published. The guidelines provide graded recommendations for the management of patients in Canada with HAE.[10] Recommendation #20 states "All patients should be trained on self-administration of HAE-specific therapies if they are suitable candidates. If patients cannot self-administer therapy, provisions should be made to ensure timely access to all appropriate therapies". As C1-INH

is dispensed through the blood banks in Canada, it is very important that there is access to C1-INH by all suitable candidates regardless of patient location.[10]

In 2010 Berinert® (C1-INH) became available through Canadian Blood Services. There has been an increased demand on Immunology clinics, Hemophilia clinics and the Berinert Assistance Program (Innomar) to train patients on self-administration of the life-saving therapy.

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Home Infusion Program; A Patient Perspective

Laura Paquet, SCIG Home and Infusion Patient

When Lynda Théoret, nurse coordinator of the SCIG program at The Ottawa Hospital, inserted a huge needle into a syringe, my eyes must have widened like saucers. She quickly reassured me that the needle went into the bottle of Hizentra—not into me. That was a relief.

I was a bit queasy about the idea of giving myself thrice-weekly SCIG infusions to manage my multifocal motor neuropathy. Even when the nurses at The Ottawa Hospital gave me IVIG, I had to look away when they inserted the needle into my arm. However, I was strongly motivated. I travel a lot for work, so not being tied to an IVIG schedule was a huge advantage.

After Lynda's cheery and thorough training, I was soon on my way home with a starter kit of supplies. Initially, I made just about every mistake in the book, from forgetting to attach the regulator to neglecting to prime the tubing. But no harm was done, and I soon got the hang of it.

I'm still shy about going out in public with my pump and tubes attached, but out of necessity I've gotten over that a bit. I've been to the store and driven our daughter to school while "wired up," as we say in our house. A long sweater or coat comes in very handy for hiding the tubing.

About a month after I started doing SCIG, I headed to Europe for a two-week work trip. I was quite nervous about going through airport security with all my needles and bottles, even though Lynda had given me an official hospital letter explaining my situation. I needn't have worried. I went through half a dozen airports and only one security agent asked me even a cursory question about my odd carry-on contents.

My biggest issue on the road was disposing of the used needles and syringes, as public sharps containers are non-existent in Belgium and Spain. In the end, I relied on hotel receptionists, who seemed a bit baffled by my request but were kind enough to take my equipment (solidly sealed inside freezer-strength Ziploc bags) for disposal.

Would I recommend the SCIG program to others? Absolutely, although it's not without its hitches. Occasionally, I hit a blood vessel, so I bleed or bruise. Sometimes, I get busy and forget to do a treatment, and end up infusing at midnight when I'd really rather be asleep. But overall, it works well at controlling my cramps and tremors, and the convenience is well worth it.

CASE STUDY: SCIG home infusion program at The Ottawa Hospital
Lynda Théoret, Nurse Coordinator - Subcutaneous Immune Globulin Program
Setting:
63 year old female outpatient
Background information:
Medical diagnosis is hypogammaglobulinemia, chronic obstructive pulmonary disease, and diabetes. Patient normally takes 15g or75 mL per week, of subcutaneous immunoglobulin (SCIG) injections three days per week.
Description of event:
Prior to receiving immunoglobulin treatment, patient had multiple chest infections requiring antibiotic treatment. She also suffered multiple hospitalizations for acute exacerbations of COPD requiring oxygen therapy, steroid treatments and antibiotics, at a rate of three or more times per year.
Program:
Candidates for this program include patients who are previously receiving intravenous immunoglobulin (IVIG), or new referrals. They are assessed by the program Nurse Coordinator, if the patient or significant other is a suitable candidate for the program. They are taught in 2 to 3 training sessions, and once the patient is proficient with the infusion technique and able to infuse safely in the home the nurse then orders 1 month of Ig product to reduce wasting in the event that patient doesn't want to stay on the Program. Orders are sent to Transfusion Medicine where the patient can pick up the product and necessary supplies to infuse by showing their hospital card or other I.D. The patient is informed by the nurse the importance of safe handling and of infusion logs which are reviewed with the patient after 1 month then after 3 months on product renewal. They are also taught the importance of reporting to the nurse any blood product adverse events. Patients will also get technique review periodically to make sure the patient continues to infuse safely. The patient is made aware to call a Nurse Coordinator if any questions or concerns arise regarding infusions.
Conclusion:
The patient has been on the program since 2012, and has done extremely well on subcutaneous immunoglobulin home therapy. She is now having less than one acute exacerbation per year, and describes them as mild and brief. She has had no hospitalizations since. The home infusion program has given her more freedom, and a better quality of life. She now has the ability to winter in Florida in good health, which is a very great improvement from her past problems of recurrent chest infections.

Upcoming Educational Events

EVENT	DATE	LOCATION
Vein to Vein 2015	March 20, 2015	NAIT Shaw Theater, Edmonton AB
NE CBS/ORBCoN Videoconference Symposium	April 15, 2015	Ontario Telemedicine Network/ The Ottawa Hospital
CBS/ORBCoN Spring Symposium	April 24, 2015	Li Ka Shing Knowledge Institute, St. Michael's Hospital, Toronto
Canadian Society for Transfusion Medicine (CSTM)	May 20 - 24, 2015	Fort Gary Hotel, Winnipeg, MB

"If you get health, then you have opportunity for literacy. Health first, then literacy. Once you have literacy, then you have a chance to bring in the new tools of communication. Let people reach out and have access to the latest advances."
- Bill Gates – 2010