Home Treatment for Children with PID

Christine McCusker, MD
Montreal Children’s Hospital
McGill University Health Center
We were asked to consult on a 3 year old girl with agammaglobulinemia. She had been diagnosed 6 months prior to the consultation and had started in IVIg replacement therapy at her treating hospital.

Her history prior to diagnosis including recurrent infections and hospitalization for intravenous antibiotic treatment.
IVIg

- Since starting IVIg this child complained of headaches beginning 24-48 hours after the infusions. They would last 1-2 days and were extremely debilitating.
- On at least one occasion she had a seizure during these headaches.
- We were asked to explore treatment options as pretreatment with antihistamines, steroids and NSAIDs were ineffective.
The Option of Sclg

- Newly available product (under Health Canada Special Access program) provided one option.
- Studies suggested that Sclg replacement may have less systemic side-effects including aseptic meningitis/headaches
- We decided to offer this treatment to the family
How to Set Up a Sclg Program?
The Quebec Experience

- Patient Selection
- Equipment
- Blood bank logistics
- Training
- Monitoring and record keeping
- Periodic reassessments
- Program Evaluation
Required Resources

- Nursing staff to train patients and monitor progress
- Process for tracking infusions
- Pumps, syringes, swabs, incidentals
- Sclg
SC infusions - required materials
Required Training-SCIG

- Storage and monitoring of SCIG at home
- Drug preparation and priming of infusion set
- Needle insertion using sterile technique
- Setting rate
- Removing cannulae and disposal of equipment
- Recognition and management of adverse events
- Record keeping
Practical aspects of SC infusions
Monitoring and Record Keeping

- Patients must keep a detailed log of infusions.
- Lot number, location of infusion and reactions must be recorded.
- Log must be signed by patient or partner.
- Logs are transmitted to the IACS nurse for review regularly at the 3 month visit.
Challenges

- Our learning curve to date has shown us
  1. Patient selection is essential (not all patients are appropriate for home therapy)
  2. One issue to be resolved is “who pays for pumps and equipment”. Overall funding for the program provided via the hospital IACS.
  3. Our goal is to provide options for our patients. While the home option is attractive, some patients are less happy with the increased frequency of SCIG use while others are pleased with the reduction in side-effects.
Patient Responses

➢ Our 3 year old began treatments on a weekly basis. Headaches and other side-effects completely resolved.

➢ But this is not our only success story…

➢ The following is a brief excerpt of an interview with one of our patients, age 9, 6 months after starting ScIg.
Interviewer: Is there something different about your belly when you, after receiving your treatment?
**Patient**: I get energy. I get lots of energy.

Interviewer: You get a lot of energy after?
**Patient**: Yeah

Interviewer: A, a lot of energy after receiving your treatment?
**Patient**: Yeah

Interviewer: Does, does the treatment at home?
**Patient**: Yeah

Interviewer: Ok. Right before having your treatment, how do you feel then?
**Patient**: I feel ok.
Interviewer: You feel ok. Then you get your treatment and after your treatment you feel?

Patient: Proud.

Interviewer: You feel proud.

Patient: And um...

Interviewer: You feel proud because?

Patient: I finished.
Interviewer: Ok. But now that you’ve experienced it, would you say anything more to the mommies and daddies (who may start the treatment) about what it’s going to be like at home?
**Patient:** It’s going to be better.

Interviewer: It’s going to be better?  
**Patient:** It’s more relaxing.

Interviewer: It’s *more* relaxing. Would you say it’s a good idea to have this treatment at home? 
**Patient:** Well, yeah.
Interviewer: And I can see a difference in you. You seem, ah, happier.

Patient: Yeah, I am.