

The Edmonton Rare Blood Disorders Programme

14 November, 2009

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University Hospital Stollery Children's





Dr. John Akabutu Centre for Bleeding Disorders

- Founded in 1978
- Patient group lobbied for provincial funding
- Home infusion program for hemophiliacs
- Annual assessment clinics



Dr. John Akabutu Centre for Bleeding Disorders

- Annual assessment clinics: hematologist, nurse, dentist, orthopod, physiotherapist
- One full-time nurse for kids and adults
- ½ time data clerk
- Full-time secretary

Dr. John Akabutu Centre for Bleeding Disorders

- 80 patients in 1978
- Expanded to cover all inherited bleeding disorders
- Now over 800 patients registered
- Home infusion of clotting factors
- On-demand
- Prophylaxis
- Keeps patients out of the ER
- Keeps joints healthy

The Edmonton Rare Blood Disorders Programme

- No formal funding for the programme
- Goals of providing comprehensive care, education, prevention of complications, health maintenance, and transition from peds to adults
- Home therapy for some

Edmonton RBD Programme

	Adults	Children
Hypogammaglobulinemias	92	1
Home therapy	73	1
HAE	53	5
Home therapy	23	1
Porphyria	13	
Sickle Cell Anemia	18	35
on RBC Exchange	7	1
Thalassemia	9	5
Diamond-Blackfan Anemia		6
Others on RBC exchange	2	
TOTALS	184	52

Edmonton RBD Programme: Geography



Hypogammaglobulinemia

Home Infusion

- First patient in August, 2006
- Now 73/92 patients on home-infusion of SCIG
- Seen in clinic by Drs. Ritchie and Pabst
- Taught in medical outpatient area individually
- Patient responsibilities: sign a contract, infuse 5-7 days a week, keep a log

Goals of SCIG Programme

- Prevent all severe infections
- Reduce mild infections to 1 or 2 per year
- Achieve an IgG level over 8





Edmonton and area

Healthier People in Healthier Communities

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Vivaglobin Home Infusion

I, _____ consent to participate in the Vivaglobin home infusion therapy program and further consent to care and treatment as deemed necessary within the program. I understand the risks and consequences of the treatment as well as the risks and consequences of not receiving treatment. I understand that Vivaglobin is a blood product, and the potential risks of blood borne infectious diseases have been explained to me.

1. I have received instruction in the following areas and have had the opportunity to ask questions and these have been answered to my satisfaction:
 1. Safe and appropriate initiation of subcutaneous injection, of mixing and self administration of subcutaneous Vivaglobin via syringe manual push.
 2. Action to take in the event of possible complications.
 3. Appropriate handling, use, and maintenance of equipment and supplies.
2. I have read the information package about subcutaneous administration of Vivaglobin, and I agree to carry out the infusions as I have been taught.

I agree to record the necessary infusion details and share the information regularly with my care team. All aspects of adverse or unanticipated events and how to contact medical assistance have been discussed with me.

I understand that in conjunction with this program, I must visit the PID clinic and physician who specializes in this disorder for regular follow-up appointments as required.

I consent to and confirm that the nature and anticipated effects of Vivaglobin home infusion therapy, including the significant risks and alternatives available, have been explained to me by Dr. _____, and I am satisfied with and understand the information provided to me.

Patient Printed Name: _____ Patient signature: _____ Date: _____

Witness Printed Name: _____ Witness Signature: _____ Date: _____

Physician Certification Statement

I hereby certify that I have explained the above procedure(s) to the above named patient/legal representative who, in my opinion, understands the nature, risks, and consequences.

Signature of Physician: _____ Printed Name: _____ Date: _____

Hereditary Angioneurotic Edema

- First patient taught self-infusion in Nov 2000
- 23/53 patients are on home infusion with C1esterase inhibitor
- Some on-demand, some prophylactically
- Easy compared to an ER trip, so more events are being treated
- Potentially life-saving

Sickle Cell Anemia

- 35 pediatric patients
- Comprehensive clinic with hematologist, nurse practitioner, pulmonologist, neurologist, social worker, Pain Service NP, and neuropsych
- Major challenges: new immigrants, some refugees, language and cultural barriers
- Backbone of the programme is EDUCATION

Sickle Cell Anemia

- Initial assessment
- Parents return for teaching, one-on-one
- Educational resources provided
- Transition to adult clinics:
 - Teens are taught about SCD, introduced to the nurse for the adult programme
 - Located in same hospital

RBC exchange in SCD

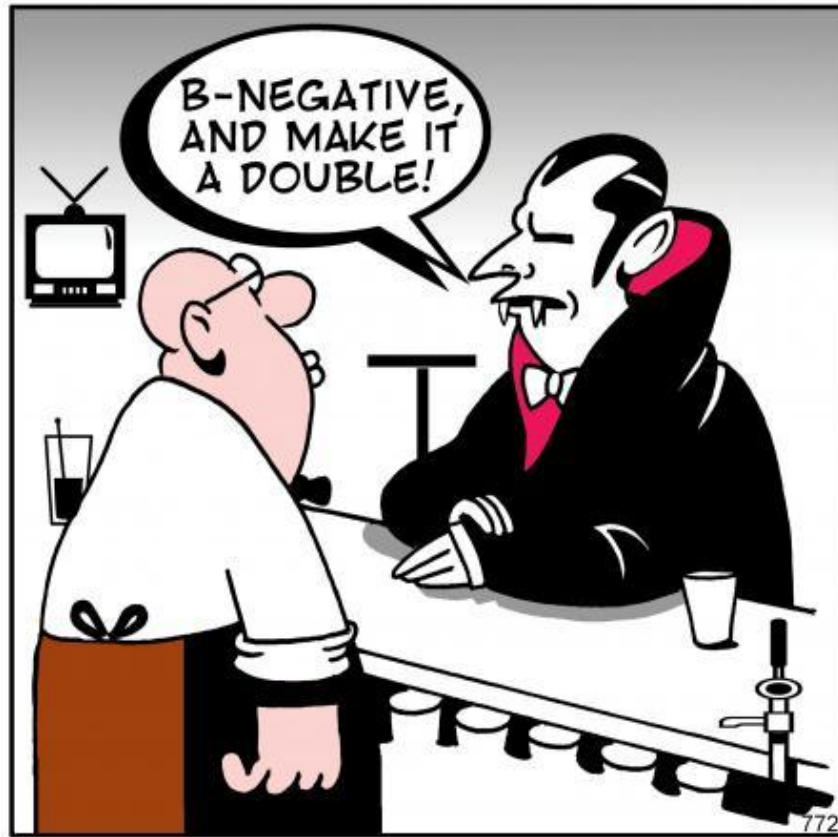


Red Blood Cell Exchange Programme

- 18 adult SCD patients: 7 are on exchange programme
- 1 thalassemia patient, 1 Hb Alberta patient

Red Blood Cell Exchange

- Challenges:
 - Huge amounts of blood
 - Blood groups (Duffy, Kell) not readily available in Edmonton
 - Campaign to encourage donation from other racial groups
 - Big venous catheters needed for exchange



Red Blood Cell Exchange

- Benefits:
 - Improved quality of life
 - Prevention of crises: painful, acute chest, stroke, priapism
 - Decrease in chronic pain, narcotic usage
 - Possible increase in life expectancy in patients with pulmonary hypertension

The Future

- Stable funding
- Evaluation of outcomes
- Registry/data management
- New facilities-The Edmonton Clinic





SHAKESPEARE