“Sickle Cell: What Every Nurse Should Know”

Presenters: Kate Uchendu (NP-Adult) & Melina Cheong (NP-Pediatrics)
Facilitator: Jacquie Dover, RN, BScN, MHSc, CCHN(C)
March 9, 2016
Agenda

• What is Sickle Cell Disease
• Epidemiology
• Newborn Screening
• Clinical Manifestations
• Pain in Sickle Cell Disease
• Psychosocial Impact of Sickle Cell Disease
• What can nurses do to help?
Sickle Cell Disease

- Group of inherited red blood cell disorders caused by abnormal hemoglobin.
- Sickled hemoglobin differs from normal hemoglobin.
- Substitution of one amino acid (valine in place of glutamic acid).
Sickle Cell Trait vs. Disease
Epidemiology

Births with a pathological Hb disorder per 1,000 live births
Ontario Newborn Screening

• 26 diseases screened.
• Sickle Cell added to newborn screening in Ontario since November 2006.
• First step in the prevention of morbidity and mortality due to severe bacterial infections.

(Gatson, M.H. et. Al, 1986)

http://www.newbornscreening.on.ca/bins/content_page.asp?cid=7-21-350&lang=1
Ontario Newborn Screening

• Currently all newborns in Ontario are screened for the following target hemoglobin diseases on their newborn screening test:
  – Sickle Cell Anemia (Hb SS)
  – SC disease (Hb SC)
  – Sickle-Beta-thalassemia (Hb S/B-thal)

• In Toronto, we see approximately 60 cases per year.
  – Started on penicillin prophylaxis.
  – Parental Education.
  – Offered Genetic counseling.
Screen Positive by Disease Category

- Hb: 93
- CF: 470
- Endo: 678
- Met: 440
- SCID: 29

Years:
- 2010
- 2011
- 2012
- 2013
- 2014

Registered Nurses' Association of Ontario
L'Association des infirmières et infirmiers autorisés de l'Ontario

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Hemoglobin

- Hemoglobin is the molecule that gives red blood cells its red color.
- Each red blood cell has several hemoglobin molecules.
- Oxygen is transported via hemoglobin molecules.
Sickle vs Normal Red Blood Cells

Normal red blood cells
- Disc-shaped.
- Looks like doughnuts with no hole.
- Flows easily through blood vessels.
- Lives for about 120 days.

Sickled red blood cells
- Sickle shaped.
- Hard.
- Gets stuck in small blood vessels causing pain and tissue damage.
- Lives for about 20 days.
Effects of Sickling

• Pain (Most common presenting complaint).
• Organ damage.
• Anemia.
Clinical Manifestations of SCD

- Retinopathy
- Asthma
- Acute Chest Syndrome
- Hepatopathy
- Cholelithiasis
- Pregnancy
- Complications
- Avascular Necrosis
- Aplastic crisis
- Stroke
- Pulmonary Hypertension
- Infection
- Asplenism
- Splenic sequestration crisis
- Renal Complications
- Priapism
- Leg ulcers
- Painful Vaso-occlusive Crises

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Pain in Sickle Cell Disease

- Most frequent presentation.
- Usually acute.
- Can have a chronic component.
- Important to differentiate between the two.
Acute Pain in SCD

- Vaso-Occlusive Episode.
- Cholecystitis.
- Splenic Sequestration.
- Priapism.
- Osteomyelitis.
Chronic Pain

• Leg Ulcers.
• Chronic Osteomyelitis.
• Avascular Necrosis of hips/shoulders.
• Depression/Anxiety.
Management of Acute Sickle Cell Pain - Recommendations

- Rapid Assessment.
- Focused history & physical exam.
- Awareness of other complications of sickle cell.
- Assess for causes of pain unrelated to sickle cell.
- Acknowledge pain, implement supportive measures.
Recommendations (cont’d)

- Use Institution Specific protocols.
- Use patient specific orders.
- Start patient on hydration.
- Rapidly initiate pain management with opioids for severe pain within 30 minutes.
- Reassess for complications, seek support from Hematology.
- Maintain best practices on in-patient unit if admitted.
Pain prevention & Intervention

The ‘3 Ps’ Approach

- **Pharmacological**
  - Medication

- **Physical**
  - Heat
  - Relaxation
  - Massage
  - Physiotherapy

- **Psychological**
  - Education
  - Distraction

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Analgesia Ladder

**Mild pain = Scale 0-3**
acetaminophen (e.g. Tylenol® or Tempra) OR ibuprofen (e.g. Advil® or Motrin®).

**Moderate pain = Scale 4-6**
acetaminophen AND ibuprofen AND If needed morphine OR hydromorphone.

**Severe pain = Scale 7-10**
acetaminophen AND ibuprofen AND morphine OR hydromorphone.
Seek medical advice if no improvement with pain medications.

**PAIN INCREASING**
Give medicines ‘around-the-clock’ every 4-6 hours in the following order.

**PAIN DECREASING**
Stop taking one medication at a time, starting with morphine or hydromorphone.

World Health Organization (WHO) Pain Relief Ladder

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WHO Recommendations on Pain Relief

• Balanced Analgesia
  – More than one class of analgesic.
  – Better pain relief with fewer side effects.

• Medications should be taken
  – Scheduled.
  – By the mouth.
  – By the ladder.
# Treatment Options for Sickle Cell Disease

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<th>Potentially Curative</th>
<th>Management of Complications</th>
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<td>Hematopoietic Stem Cell Transplantation.</td>
<td>Hydroxyurea (Hemoglobin F modulating agent).</td>
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<td>Regular Blood/Exchange Transfusions.</td>
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Blood Transfusions

- RBC Phenotype
- Absence of sickle hemoglobin.
- Used to remove sickle cells and replace with normal cells without increasing blood viscosity.
  - Autologous blood transfusion should be avoided.
- Alloimmunization (immune response to foreign antigens)
  - Increases with each transfusion.
  - Phenotypic incompatibility between donor and recipient.
  - 20%-25% incidence, greater than general public.
Most Importantly……

- Hematology should be notified before any blood products are given to a sickle cell patient.
Issues commonly encountered by Patients & Families

• Stigma.
• Poverty.
• Chronic pain and perception of addiction.
• Racism.
• Frequent visits.
• Transitions in Care.
• Patient & Provider knowledge deficit.
Psychosocial Impact

- Relationships.
- Workplace Issues & Finances.
- School.
- Anxiety about health.
- Guilt about passing on disorder.
What Nurses Can Do

• Educate yourself about Sickle Cell Disease.
• Timely assessment of patients.
• Advocate for rapid and effective treatment of pain.
• Develop trusting relationship with patient and family.
• Educate patients to use a pro-active approach in pain management.
• Educate patients and families on transition.
• Starting transition program from paediatrics to adult care.
Best Practice

• Utilization of the following:
  – Clinical protocols.
  – Medical directives.
  – Preprinted order sets.
  – Patient specific orders
Advocacy Groups

Associations
- Camp Jumoke
- Sickle Cell Disease Association of Canada
- Sickle Cell Awareness Group of Ontario
- The Sickle Cell Association of Ontario
- IASCNAPA

Community Health Centres
- Taibu Community Health Centre
- Black Creek Community Health Centre
- Unison Health & Community Services
CanHeam

http://canhaem.org/
Resources

– www.aboutkidshealth.ca
– www.scinfo.org
– http://www.who.int/en/
– http://www.newbornscreening.on.ca/bins/index.asp
– www.nhlbi.nih.gov
– http://www.uhn.ca/PrincessMargaret/PatientsFamilies/Clinics_Tests/Red_Blood_Cell_Disorders
– https://www2.rcn.org.uk/__data/assets/pdf_file/0004/372991/003874.pdf
Questions
References