Red Cell Exchange and Iron Overload Reduction – It comes out in the wash!

Sickle cell anemia case report

Kathy Grouchy RN
BC Children’s Hospital
Introduction

- Sickle cell anemia patient with severe iron overload secondary to red cell transfusions.
- Switching to automated red cell exchange (RBCX) procedures effectively lowered the iron overload.
Transfusion therapy indications for sickle cell anemia

- Ischemic Strokes
- Stroke prevention
- Elevated transcranial doppler velocity – frank cerebral vasculopathy
Types of transfusions

- Simple red cell transfusion
- Manual phlebotomy/exchange transfusion
- Automated red cell exchange (RBCX)
- RBCX most efficient to reduce HGB S%
- Usually long term therapy
Case report
18 yr old sickle cell anemia patient

- History of left sided ischemic stroke when 7 yrs old
- Initially poor peripheral venous access for automated red cell exchange (RBCX)
- Implanted IVAD not an option (high risk procedure)
- Simple red cell transfusion x 7 years (age 7-14) by peripheral vein
Case Report cont’d

- Non adherence to iron chelation treatment
- Severe iron overload from transfusions
- Switched to manual phlebotomy/exchange transfusion for 18 months (prior to RBCX)
- Peripheral antecubital veins adequate when 14 yrs old!
- Switched to RBCX on OPTIA every 4 weeks at age 15 by peripheral access 18g and 22g return
- 45 RBCX procedures
Procedure data entry

- Hct pre range 24 to 26
- End Hct range 25 to 26
- Hgb S% pre 44%
- Hgb S% post < 15%
- FB 100%
- Blood removed 2660ml
- Replaced 2370ml
- Average fluid replacement Hct 58
What’s in the bag?
Results

- RBCX procedures consistently lowered serum ferritin levels and Ferriscan results.
- Pre RBCX liver iron concentration (LIC) 17.0 to 17.8 mg/g dry tissue weight.
- LIC after RBCX – 1 yr = 9.6 mg/g, 2 years = 5.0 and 3 yrs = 2.4
- Ferritin levels pre 2800ug/L and currently 161ug/L.
Serum Ferritin Levels
Ferriscan results (MRI of Liver)

FerriScan Liver Iron Concentration Report

Report No: [Blank]
Patient ID: [Blank]
Name: [Blank]
Birth Date: [Blank]
Scan Date: 26 Feb 2011
Analysis Date: 01 Mar 2011
MRI Centre: UBC & BC Children’s Hospital

Average Liver Iron Concentration
17.8 µg/dry tissue (NR: 0.17-1.8)
319 mmol/kg dry tissue (NR: 3.3-33)

Transverse Relaxation Rate (2R) Images

Transverse Relaxation Rate (2R) Distribution

Note: The area of the liver image used for the FerriScan analysis excludes large vascular structures and other image artifacts. Approved by Service Centre Manager.

Rosonance Health Analysis Services Pty Ltd
www.resonancehealth.com
Ferriscan results

Liver Iron Concentration (LIC) mg/g dry weight over time.

- Increase from March 2010 to February 2011
- Decrease from February 2011 to February 2015
- Phlebotomy and RBCX events shown on graph.

Ferriscan

Liver Iron Concentration (LIC) mg/g dry weight

month/year

Phlebotomy
RBCX

LIC mg/g dry weight

Mar-10 Feb-11 Jan-12 Feb-13 Feb-14 Feb-15
Monthly red cell transfusions can result in severe iron overload

Switching to Automated RBCX procedures can not only lower Hgb S%, it can also effectively reduce iron overload secondary to red cell transfusions

Switching to RBCX can result in a positive outcome and improved quality of life for a sickle cell disease patient with iron overload
RED CELL EXCHANGE AND IRON OVERLOAD REDUCTION – IT COMES OUT IN THE WASH!

K Grouchy RN, G Chan RN, K Douglas RN, C Strahlendorf MD Hematology Program, British Columbia Children’s Hospital, Vancouver, BC Canada

**Case Report**

**Background**
- Sickle cell anemia (SCD) patients with stroke complications
- Red cell transfusions began at age 7 yrs
- Non-adherence to iron chelation treatment resulted in severe iron overload
- Poor peripheral VA for RBCX procedures (implanted access port not an option)
- Manual phlebotomy (10mls to 20mls/litre) exchange transfusion for 18 months prior to RBCX
- Adequate peripheral VA for RBCX when 14 yrs old
- RBCX on the Spectra Optia every 4 weeks – post Hbs< 15%
- Prescribed iron chelation stopped after RBCX started
- Annual ferriscan and cardiac MRI started 2 years prior to RBCX

**Problems**
- Red cell transfusions and non-adherence to iron chelation results in severe iron overload (IO)
- IO leads to organ damage, failure and death
- Inadequate vascular access (VA), for RBCX
- Implanted venous access ports: invasive high risk procedure - may not be an option for some patients

**Iron Levels Pre**
- Liver Iron Concentration: 17.6 mg/g = HIGH

**Iron Levels after Phlebotomy Treatment**
- Liver Iron Concentration: 17.0 mg/g - HIGH

**Iron levels after 1yr of RBCX**
- Liver Iron Concentration: 9.7 mg/g = HIGH

**Iron levels after 2yrs of RBCX**
- Liver Iron Concentration: 5.0 mg/g = HIGH

**Iron levels after 3yrs of RBCX**
- Liver Iron Concentration: 2.4 mg/g = NORMAL

**Results**
- RBCX procedures consistently lowered ferritin levels and Ferriscan results.
  - Pre RBCX Liver iron concentration (LIC) 17 to 17.8 mg/g dry tissue weight.
  - LIC after RBCX: 1 yr = 9.6 mg/g dry tissue weight, 2 years = 5.0 and 3 yrs = 2.4 mg/g dry tissue weight.
  - Ferritin levels pre 2800ug/L and currently 161ug/L.

**Conclusion**
- Monthly red cell transfusions can result in severe iron overload
- Automated RBCX procedures can significantly reduce iron overload secondary to red cell transfusions
- Switching to RBCX can result in a positive outcome, improved quality of life, and improved compliance for a sickle cell disease patient with iron overload.
Thank You

Questions ?