Experience of transfusional processings by Erythrapheresis in Adults Sickle Cell Center of Martinique

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Sickle Cell Disease

• Advances in care
• Improvement of quality of life and aging population
• Systemic disease
  o Acute and chronic complications: VOC, Infections, ACS, Stroke, Priapism, Leg ulcers, Retinopathy, Joints complications, Organ failures: renal, cardiac, liver ...
Therapeutics

- Early diagnosis since 1984
- Early prevention
- Therapeutic disease education
- Hydroxyurea
- Erythropoietin
- Bone marrow transplant
Therapeutics

- Early diagnosis since 1984
- Early prevention
- Therapeutic disease education
- Hydroxyurea
- Erythropoietin

.....Bone marrow transplant

And Transfusion
Transfusion treatment

Always case by case approach
Transfusion file

- Systematically at the beginning
  - Group ABO
  - Phenotype Rhesus, Kell
  - Systematic extended phenotype

- RBC bag = leukodepleted, Rhesus Kell phenotyped and compatibilised

- +++ , If alloimmunisation
  More similitude in Martinique between patients and donors)
Simple transfusion

• Indication = Worsening of a poorly tolerated anemia
  o Clinical
  o Reticulocytes rate

• Goal = Hb + Oxygen delivery
Transfusional Exchange

• *Goal* = replace the sickle erythrocytes by red blood cells containing Hb *A*
  - Decrease of %HbS
  - Avoid hyperviscosity
  - Delayed iron overload

• ⚠️ Never Hb should exceed 10 to 11 G/dL
Red Blood cell exchange: 2 techniques

- Manual Exchange: combination of phlebotomy and transfusion
- Automated Exchange with an apheresis device
# Red Blood cell exchange

<table>
<thead>
<tr>
<th><strong>Manual Exchange</strong></th>
<th><strong>Erythrapheresis</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Whole blood phlebotomy + transfusion</td>
<td>Elective subtracting of RBC Replacement by RBC Plasma restitution</td>
</tr>
<tr>
<td>1 or 2 venous access</td>
<td>2 good venous access</td>
</tr>
<tr>
<td>Limited volume</td>
<td>Ability to treat large blood volume</td>
</tr>
<tr>
<td>Hypovolemia risk</td>
<td>Constant hematocrit and volemia</td>
</tr>
<tr>
<td>Hyperviscosity risk</td>
<td>Viscosity controlled (Ht)</td>
</tr>
<tr>
<td>Ineluctable iron overload</td>
<td>No iron overload</td>
</tr>
<tr>
<td>No sophisticated equipment</td>
<td>But sophisticated material</td>
</tr>
<tr>
<td>Very time consuming</td>
<td>More comfortable for patients and nurses</td>
</tr>
</tbody>
</table>
Exchange indications
Punctual and Curative

- Stroke
- Acute Chest Syndrome
- Hepatic sequestration
- VOC hyperalgic not yielding to major analgesics > 5 to 8 days
- Acute Priapism: Etilefrine Failure and/or > 3 h
- Multi organ failure
Blood Exchange transfusion indications
Punctual and Preventive

- Preparation for surgery
- Cerebral Arteriography
- Pregnancy with severe sickle cell and/or obstetric history
Blood Exchange Transfusion indications

Long term

- Stroke: Primary prevention (cerebral vasculopathy)
- Stroke: secondary prevention
- Recurrent VOC and ACS if Hydroxyurea not tolerated
- Chronic organs failure: PAH, Kidneys, Heart, Liver
- Recurrent Priapism treatment-resistant
- Recurrent and rebel leg ulcers: less efficacy, but less painful
- Prevent secondary hemochromatosis (erythropheresis)
Manual exchange
Phlebotomy + Transfusion

- 3 phases
  - Phlebotomy 10ml/kg (5ml/kg in case of stroke)
  - Phlebotomy continued and start of the transfusion
  - Transfusion alone

<table>
<thead>
<tr>
<th>Initial Hematocrit</th>
<th>First Bleeding volume</th>
<th>Seconde Bleeding volume</th>
<th>Transfusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 7 g/dl</td>
<td>0</td>
<td>0</td>
<td>2 à 3 CG</td>
</tr>
<tr>
<td>7.5</td>
<td>0</td>
<td>0 à 150 ml</td>
<td>2 à 3 CG (900 ml)</td>
</tr>
<tr>
<td>8</td>
<td>0</td>
<td>200 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>8.5</td>
<td>0</td>
<td>250 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>9</td>
<td>200 ml</td>
<td>200 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>9.5</td>
<td>200 ml</td>
<td>250 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>10</td>
<td>250 ml</td>
<td>300 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>10.5</td>
<td>300 ml</td>
<td>300 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>11</td>
<td>300 ml</td>
<td>350 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>11.5</td>
<td>350 ml</td>
<td>350 ml</td>
<td>2 CG</td>
</tr>
<tr>
<td>12</td>
<td>350 ml</td>
<td>400 ml +/- 1 saignée le lendemain</td>
<td>2 CG</td>
</tr>
</tbody>
</table>

Recommandations 2015
Experience from ASCDC - Martinique
At March 30, 2016

772 patients

- 348 Hb SS: 45.08%
- 341 Hb SC: 44.17%
- 62 Hb Sβ+ Thal: 8%
- 5 Sβ0 Thal: 0.6%
- Hb ASAntilles, SDKorlebu, SOArab, SDPunjab ...
CRD-Adultes Martinique (2)

According to age

<table>
<thead>
<tr>
<th>Titre de l'axe</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>18-24</td>
<td>41</td>
</tr>
<tr>
<td>25-39</td>
<td>136</td>
</tr>
<tr>
<td>40-49</td>
<td>117</td>
</tr>
<tr>
<td>50-59</td>
<td>127</td>
</tr>
<tr>
<td>&gt; 60</td>
<td>79</td>
</tr>
</tbody>
</table>

- **Women**
- **Men**
Spectra Optia Apheresis System

- **GUI** - simple, high resolution touch screen, multiple languages
- **Cassette tray-load**
- **View port** - observe interface
- **Wheels/pedal** - optimized mobility
- **Dual IV pole** - accommodates multiple bags
- **Sealer** - convenient tube sealing device
- **Centrifuge door/chamber/filler** - access to centrifuge
- **Data port** - for printing and exporting data

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Automated Red Blood Cell Exchange

- Apheresis device: Spectra Optia / Terumo BCT

- Specific software
  - Patient parameters
    - Weight
    - Height
    - Ht
    - HbS (%)
  - Blood bag hematocrit is needed

- FCR: Residual level of HbS (%)
Pre transfusion Assessment

• 48 to 72 hours before
• Complete Blood Count+ Retic., HbS count, Calcemia, Ferritin
• Available RBC volume

• Order of 4 to 5 RBC bags, 7 to 8 in case of emergency
• Mean of **20 to 30 ml/kg** for iterative exchanges
• **50 ml/kg** for top up exchange
Procedures

- Duration of the procedure depends on the quality of venous access
  - In our ward: Peripheral venous access +++, 3 fistula, 2 fistula failure

- Depletion + Exchange: if Ht > 26-27% + Hb S (%) «favorable»
Post transfusionnel Assessment

At least 1 hour after the end of the erythropheresis

CBC, HbS
Since October 2012
More than 8 procedures/month

353 P./51 patients

- 2012: 15 Procedures
- 2013: 87 P.
- 2014: 106 P.
- 2015: 108 P.
- At 25 April 2016: 37 P.
Indications (1)

Prior Manual Exchange
12 patients/15

• 4 patients with fistula
• (+ 3 with difficult venous access)
Indications (3)
Punctual procedures

• **Pre Valvuloplasty**: 2 Procedures / 2 patients

• **In ICU**
  - Stroke: 4 Procedures / 2 patients
  - **ACS**: 2 Procedures / 2 patients (1 Hb SC)
  - **VOC**: 2 Procedures / 2 patients (1 Hb Sβ₀ Thal)
  - Priapism = 1 Procedure / 1 patient
Indications (4)
Iterative procedures: 37 p.

- VOC and/or failure or “waiting” Hydroxyurea: 7 patients
- Pregnancy: 7 (3 Hb SC)
- Cerebral Vasculopathy/Stroke: 6
- PAH: 6 (1 Hb SC)
- Multi organ failure: 6
- Renal failure: 4 (1 Hb SC)
- Liver failure: 2 (Hb Sβ+thal)
- Leg ulcers: 2 (1 HbSC)
- Desire for children: 2 (Stop Hydroxyurea)
- Post liver transplant: 1
- Priapism: 1 patient
Availability of the apheresis device and...

• Patients
  o Good venous access
  o Incentive, Information regarding fistula

• Nurses

• Physicians

• Device Failure fear
Benefits

• Stroke: No recurrence
• Known positive effects of automated RBC exchange
• Hemochromatosis
  • 4 patients with iron overload treated with chelation therapy (DFO) because of liver and renal failure: Ferritin between 2788 and 3400 mg/ml ➔ 452 and 633 within 21 to 30 months ➔ Chelation therapy can be stopped
• Targeted HbS reached
• PAH: 8 months
Initial progressive HTAP confirmed on Central line
Exchange started in February 2014
HbS = 35%, 07/10/2014

↓ Tricuspid leak 387 to 282 cm/s
↓ PAPs measured 70 to 42 mm Hg
Questions which remain

• Lower Risk of alloimmunisation? Less noisy?

• Alloimmunisation article
  – Michot, F. Driss et al, Transfusion, 2014
Patients feeling impact
Nurses feeling impact
Conclusion 1

- **TRANSFUSION** = unquestionable major therapeutic in some situations

- Effective Technique
  - decreased %HbS
  - long-term treatments avoided for iron overload
Conclusion 2

- Stop chelation?
- RBC bags use (punctual, long term)
- Targeted Ht: device/laboratory
- HbS low rate achieved

- Venous access: Fistula, …
Thanks

- R. IFRIM, MD
- S. ALEXIS-FARDINI, MD
- L. HAUSTANT-ANDRY, MD

Special thanks

- The entire center team
- Patients, their families et helpers

Thanks

- Françoise DRISS
- M. B.
- Y.A., J.I.
- P.B.

Thanks for your attention