

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



Gylna LOKO  
Centre de référence de la Drépanocytose  
[gylna.loko@gmail.com](mailto:gylna.loko@gmail.com)

# Sickle Cell Disease

- Advances in care
- Improvement of quality of life and aging population
- Systemic disease
  - 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
  - Clinical
  - 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

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

# Exchange indications

## Punctual and Curative

- Stroke
- Acute Chest Syndrome
- Hepatic sequestration
- VOC hyperalgetic 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 (erythrapheresis)

# Manual exchange Phlebotomy + Transfusion

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

Initial Hematocrit	First Bleeding volume	Seconde Bleeding volume	Transfusion
< 7 g/dl	0	0	2 à 3 CG
7.5	0	0 à 150 ml	2 à 3 CG( 900 ml )
8	0	200 ml	2 CG ( 600ml )
8.5	0	250 ml	2 CG
9	200 ml	200 ml	2 CG
9.5	200 ml	250 ml	2 CG
10	250 ml	300 ml	2 CG
10.5	300 ml	300 ml	2 CG
11	300 ml	350 ml	2 CG
11.5	350 ml	350 ml	2 CG
12	350 ml	400 ml+/- 1 saignée le lendemain	2 CG

# Experience from ASCDC - Martinique

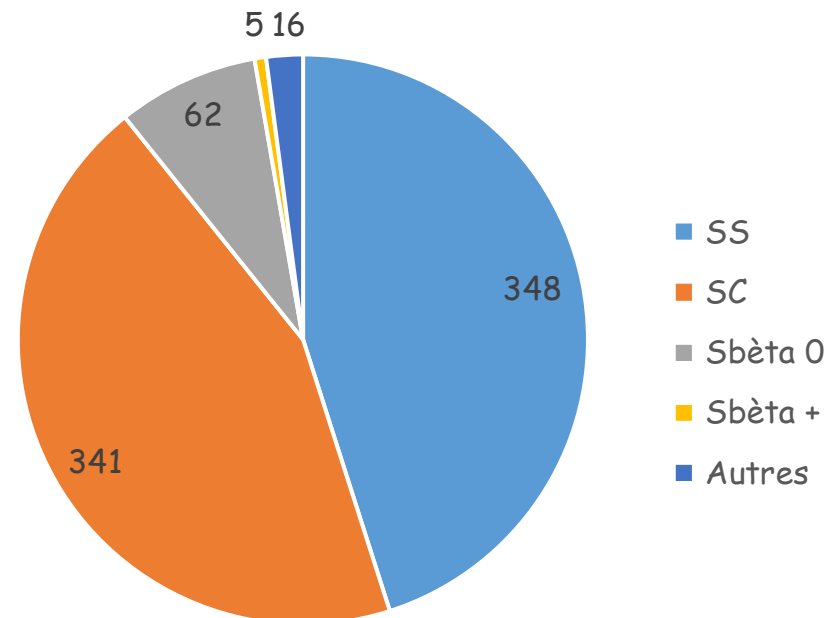
# CRD-Adultes Martinique (1)

At March 30, 2016

772 patients

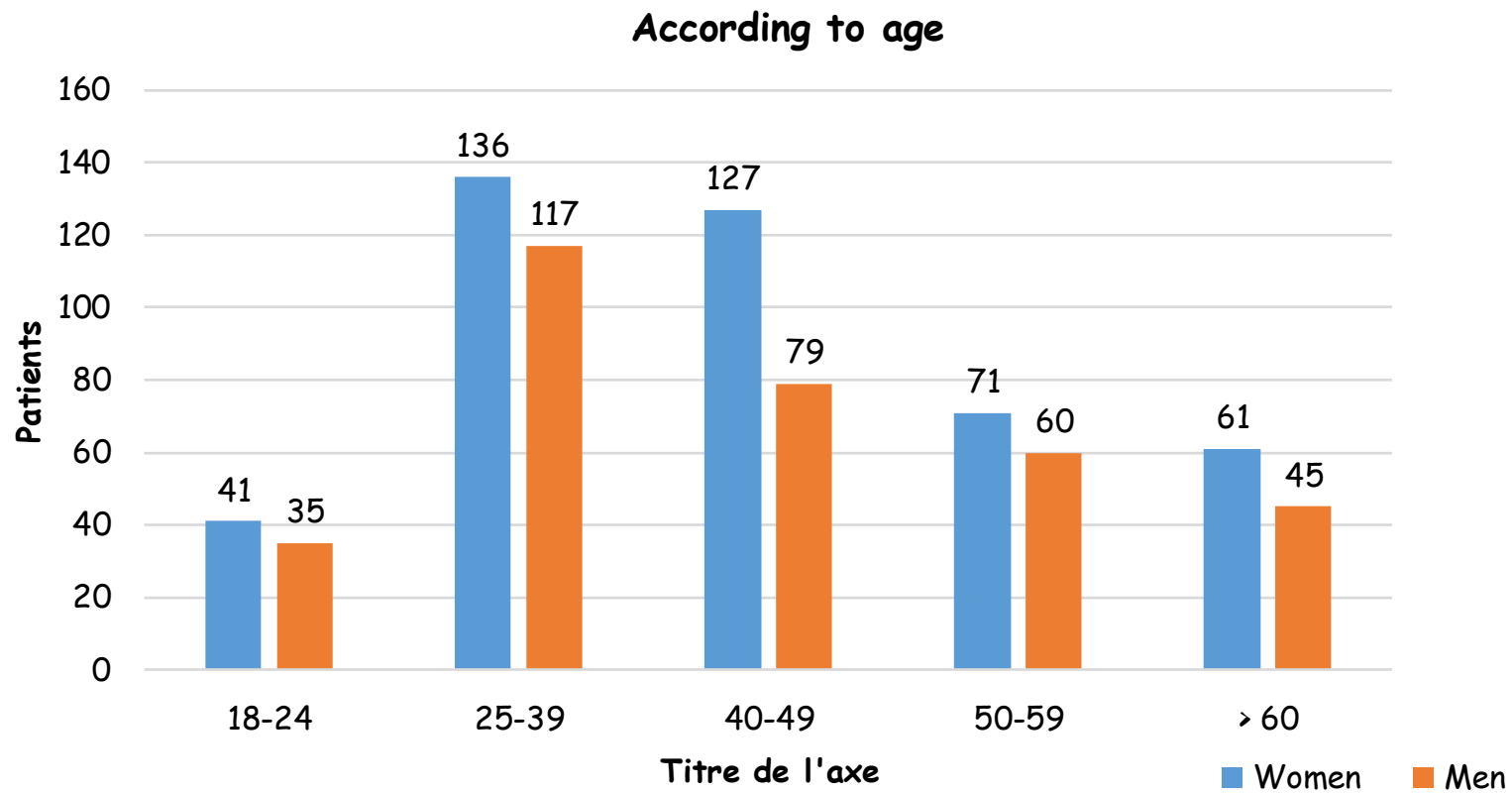
- 348 Hb SS: 45,08%
- 341 Hb SC : 44,17%
- 62 Hb S $\beta$ <sub>+</sub> Thal: 8%
- 5 S $\beta$ <sub>0</sub>Thal: 0,6%
- Hb AS Antilles, SD Korlebu, SO Arab, SD Punjab ...

According to genotype





# CRD-Adultes Martinique (2)



# Spectra Optia Apheresis System

GUI - simple, high resolution touch screen, multiple languages

Dual IV pole - accommodates multiple bags

Cassette tray-load cassette

Sealer - convenient tube sealing device

View port-observe interface

Centrifuge door/chamber/filler - access to centrifuge

Wheels/pedal - optimized mobility

Data port - for printing and exporting data



# 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



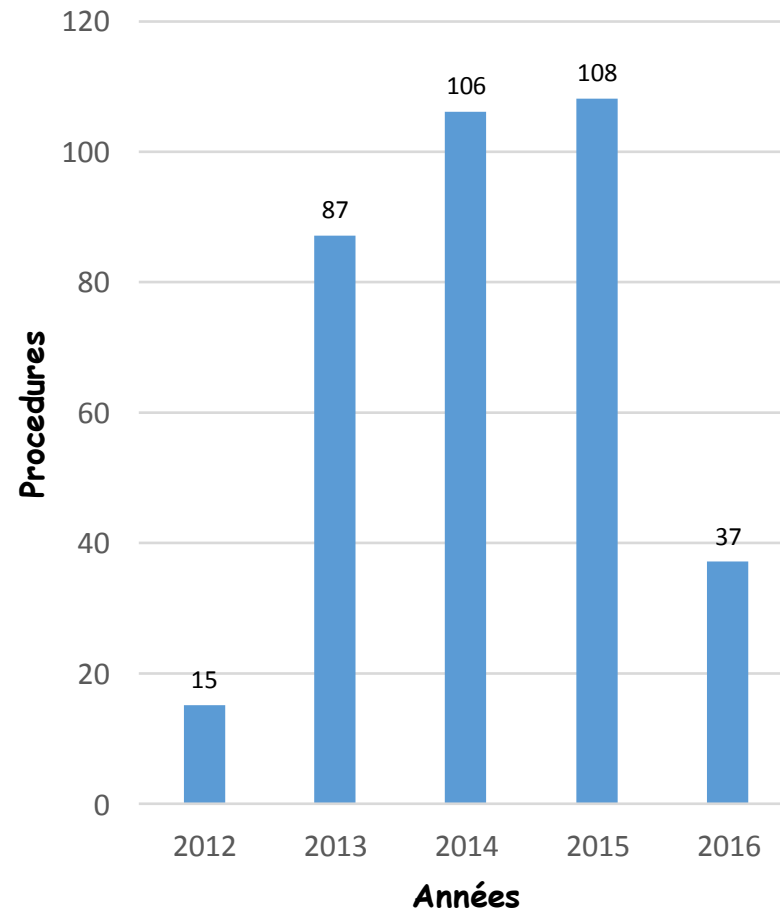
At least 1 hour after the end of the erythrocytapheresis  
CBC, HbS

# 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

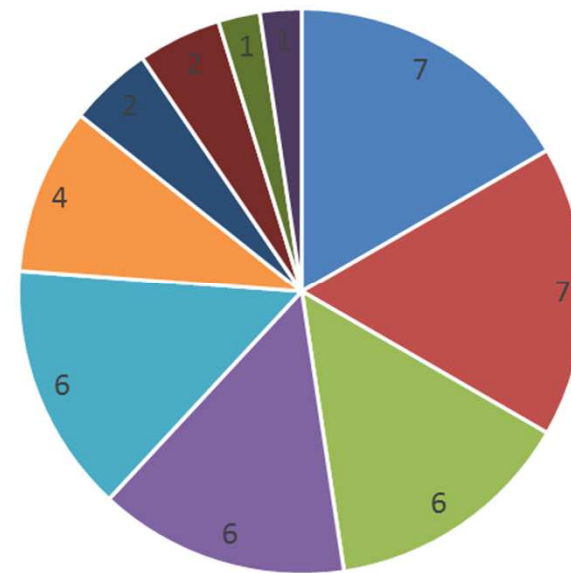
- Pre Valvuloplasty : 2 Procedures / 2 patients
- In ICU
  - Stroke : 4 Procedures / 2 patients
  - ACS : 2 Procedures / 2 patients ( 1 Hb SC)
  - VOC : 2 Procedures / 2 patients (1 Hb S $\beta_0$ Thal)
  - Priapism = 1 Procedure/ 1 patient

# 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 S $\beta$ +thal)
- Leg ulcers: **2** (1 HbSC)
- Desire for children: **2** (Stop Hydroxyurea)
- Post liver tranplant: **1**
- Priapism: **1** patient

Nombre de Patients



■ VOC ■ Pregnancy ■ Stroke ■ PAH ■ MOF ■ KF ■ LF ■ U ■ DC ■ Pp

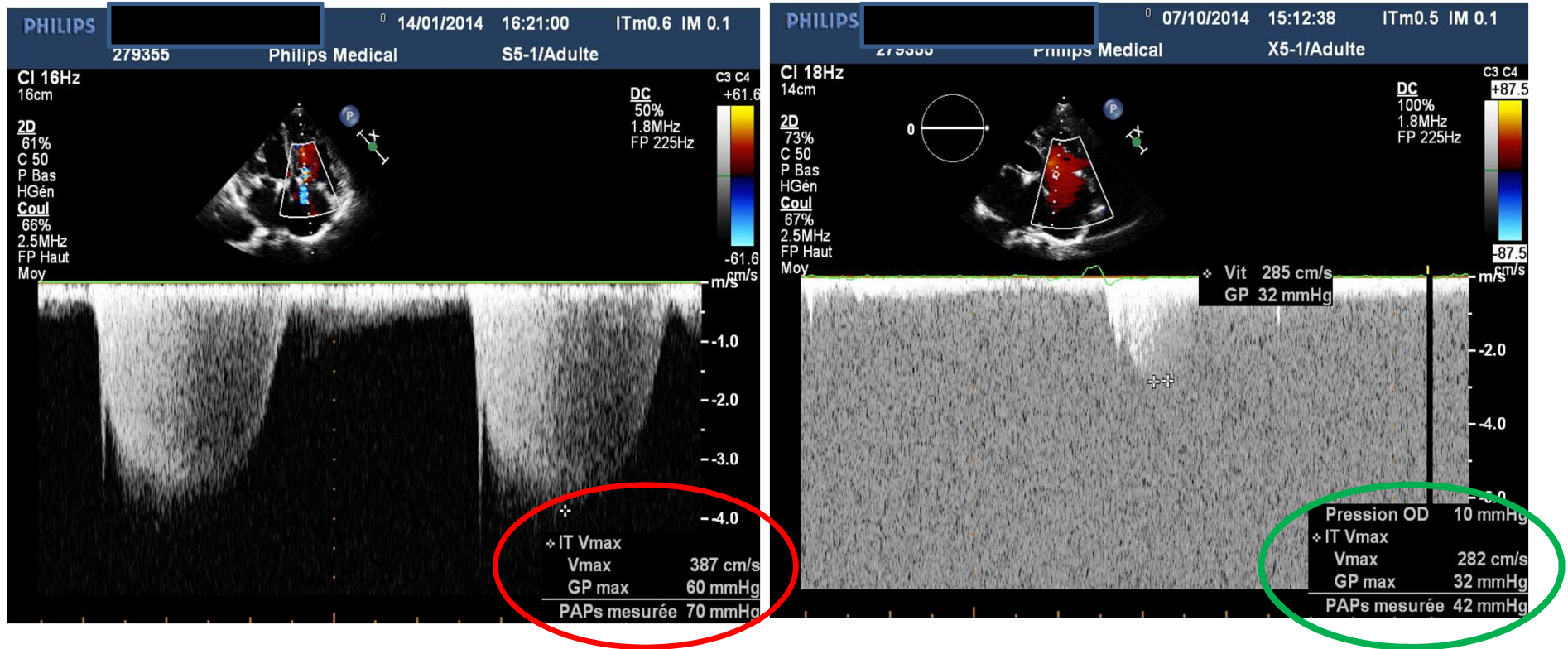
# Availability of the apheresis device and...

- **Patients**
  - Good venous access
  - 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

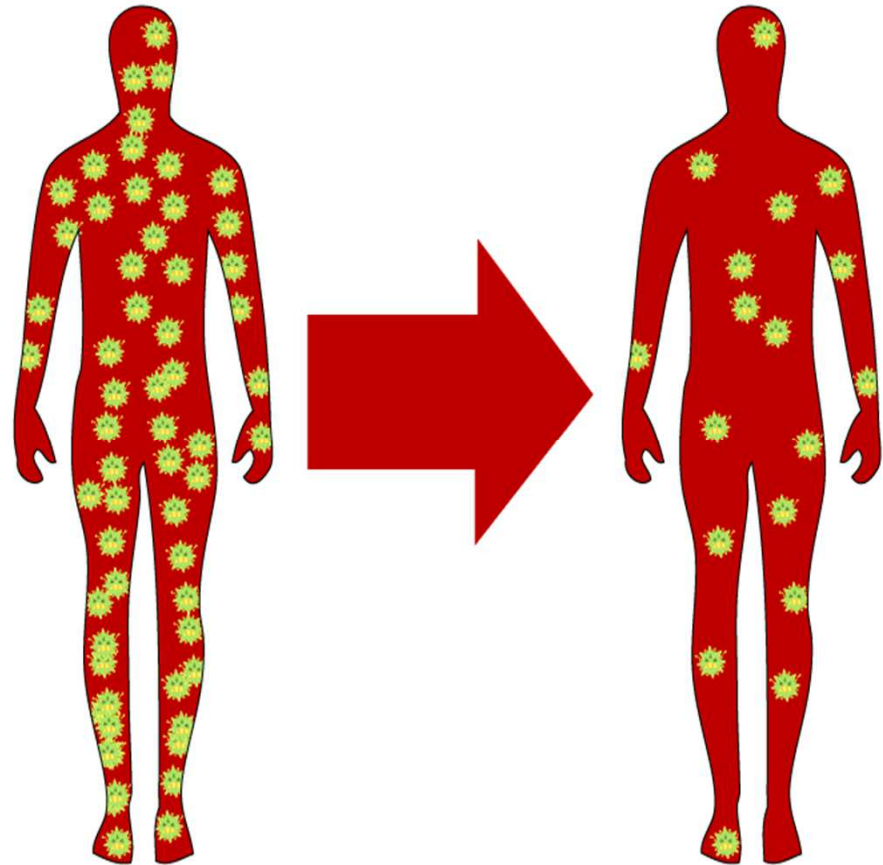
- Lower Risk of alloimmunisation ? Less noisy?
- Alloimmunisation article
  - Michot, F. Driss et al,  
Transfusion, 2014



**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

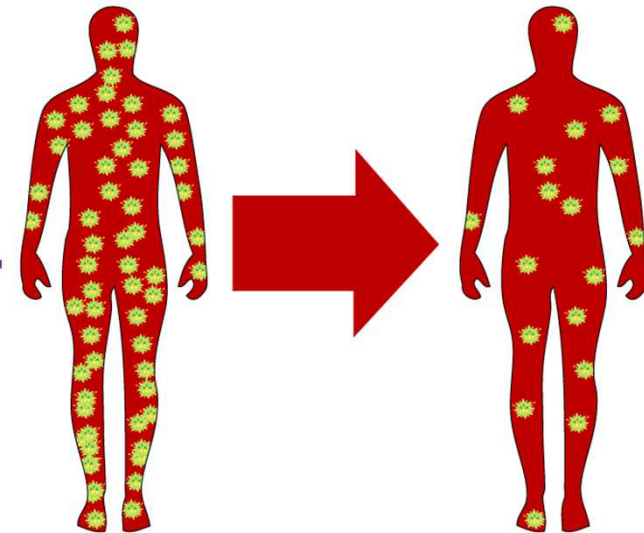




## Conclusion 2

- 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



## Thanks

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



## Special thanks

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

**Thanks for your attention**