GROUPE HOSPITALIER
HENRI MONDOR
ALBERT CHENEVIER - GEORGES CLEMENCEAU
JOFFRE-DUPUYTREN - EMILE ROUX

DHTR CASE REPORTS

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CENTRE DE RÉFÉRENCE MALADIES RARES
« SYNDROME DREPANOCYTAIRES MAJEURS »
HÔPITAL HENRI MONDOR – ALBERT CHENEVIER , AP-HP

CASE REPORT N 1

26 years old woman with Homozygot Sickle Cell Disease and a history of :

- Hb at steady state at 8 g / dl
- 4 Acute Chest Syndromes with intensive care admission and thrombosis
- Bilateral hip osteonecrosis
- No hydroxurea treatment

TRANSFUSION HISTORY

This patient has done several episodes of DHTR since 2008

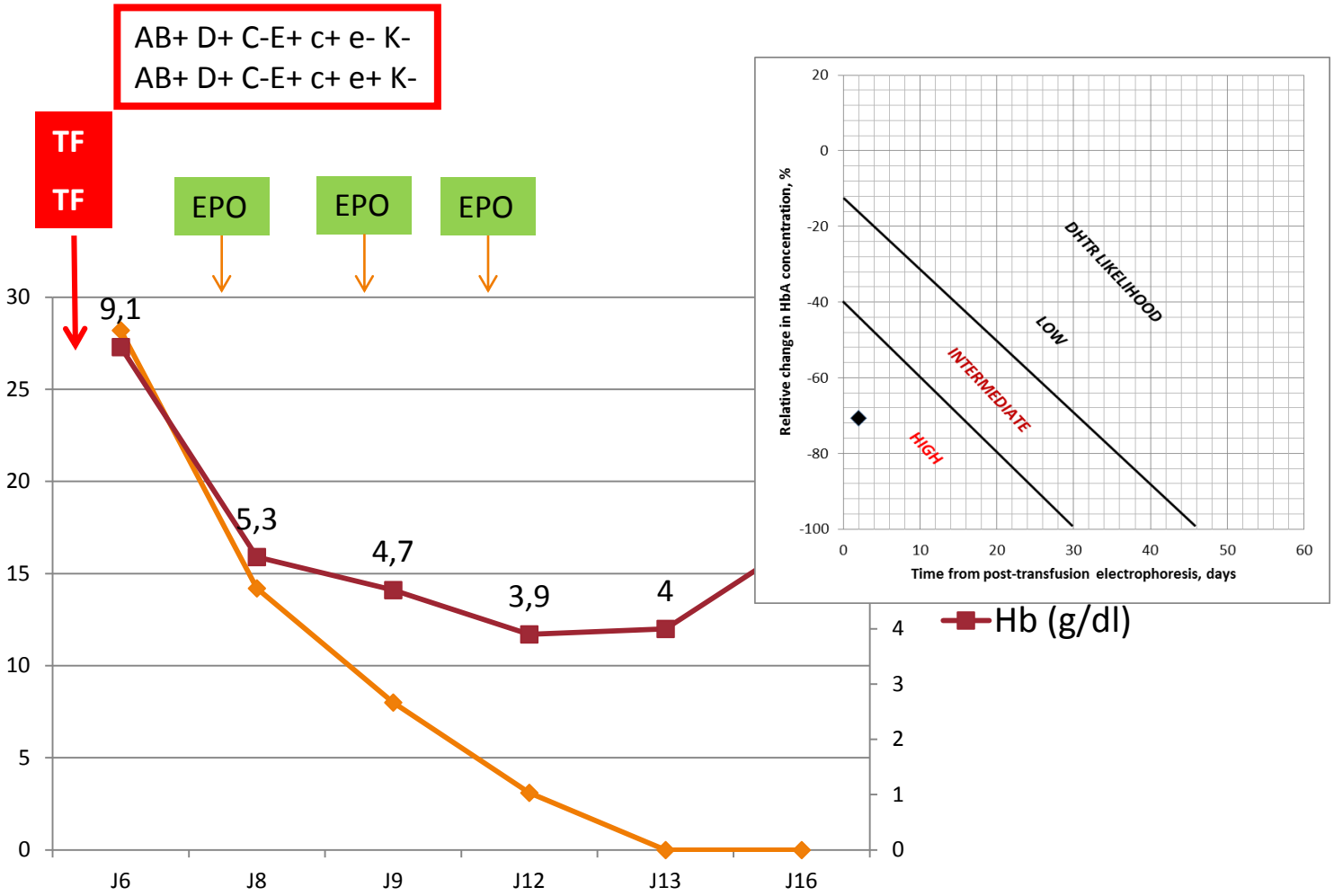
Since the first diagnosis of DHTR in 2008, she received a total of 19 RCB Units,

Probably other DHTRs have occurred previously and undiagnosed

Allo antibodys known : anti LeA, anti Hi, anti Cw

AB+ C-E+c+e+K-**Fya-Fyb-**Jka+Jkb+**M-** N+S+s+Lea-Leb-P1

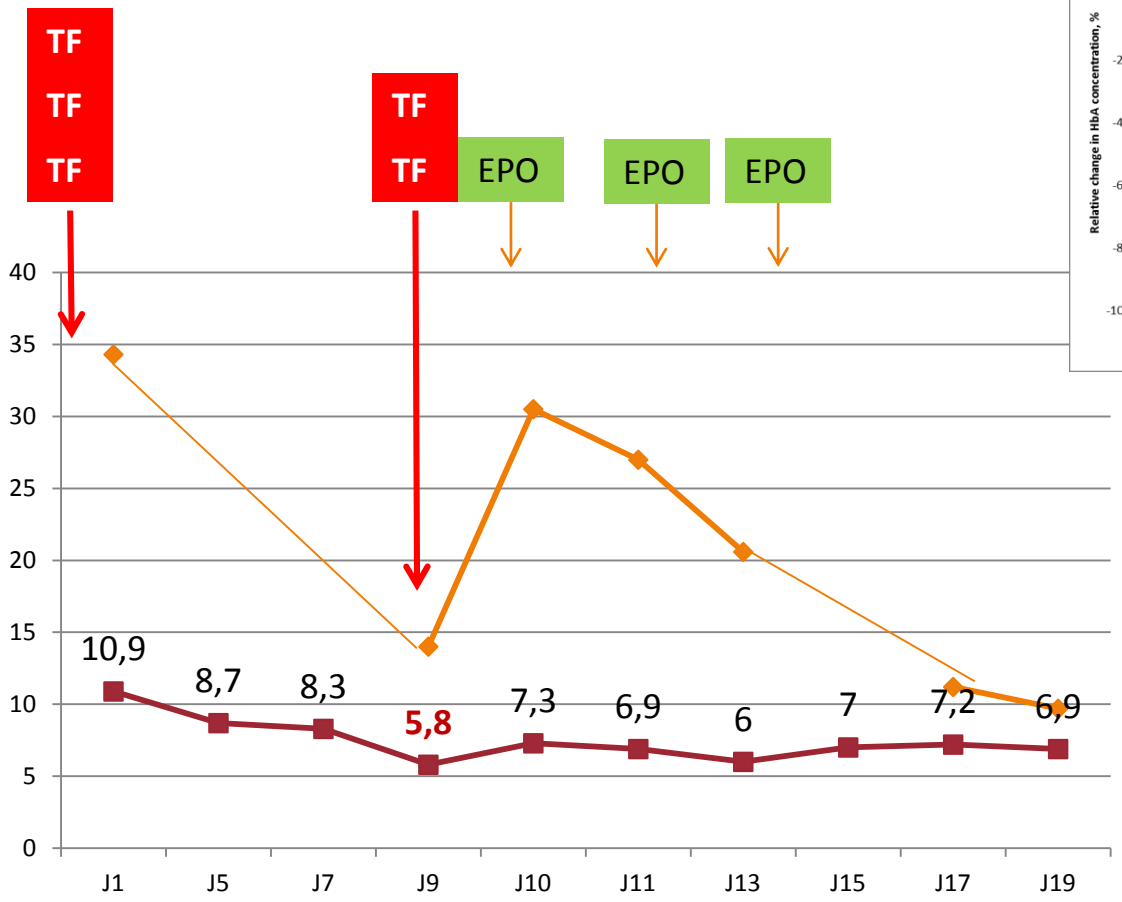
2010

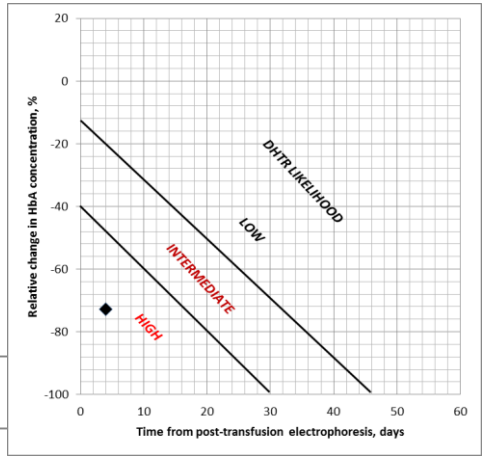


The patient was transfused during the surgical procedure
At Day 6 post surgery she has a ACS, dark urine and anemia.
Transfered in ICU , she received only EPO and symptomatic treatment

2011

M , Lea and Leb was not matched





Surgery of Hip Prothesis.

It required the transfusion of 3 RBC units during the procedure.

Occurrence of vaso-occlusive crisis with dark urine but the surgeon transfused the patient before her transfer.

2013

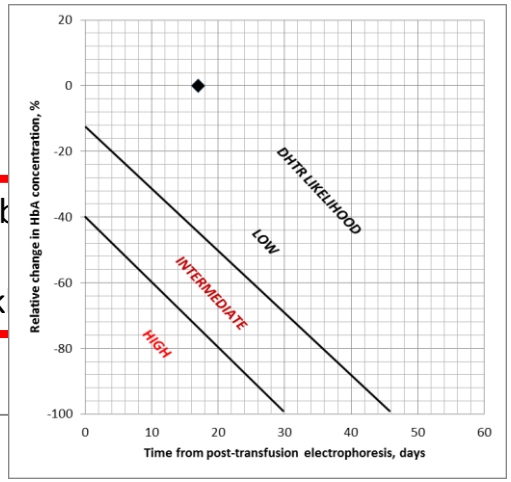
Rituximab

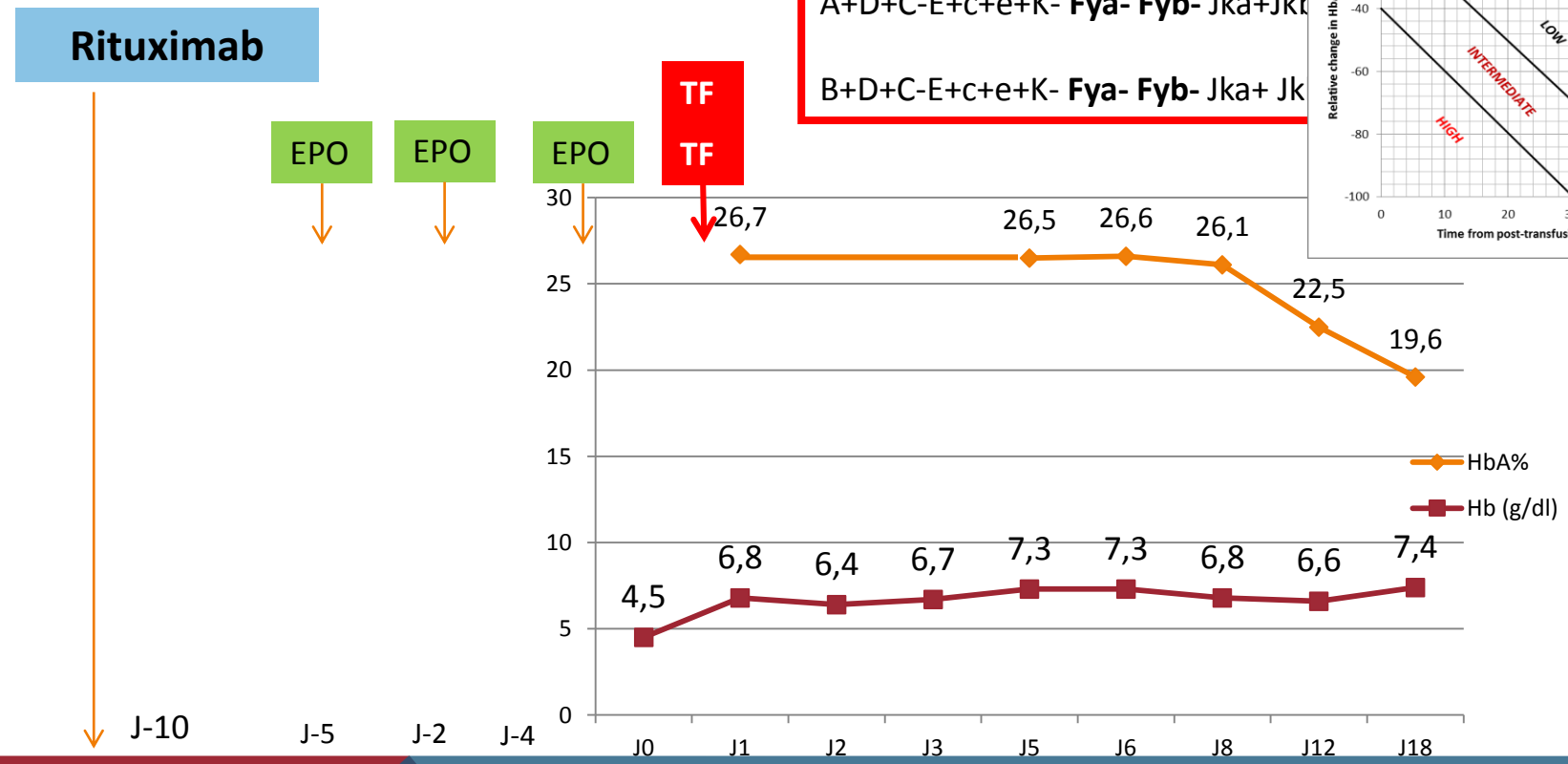
EPO EPO EPO

TF TF

A+D+C-E+c+e+K- **Fya- Fyb-** Jka+Jkb

B+D+C-E+c+e+K- **Fya- Fyb-** Jka+ Jk





Patient admitted for ACS with severity criteria and HTP in ICU

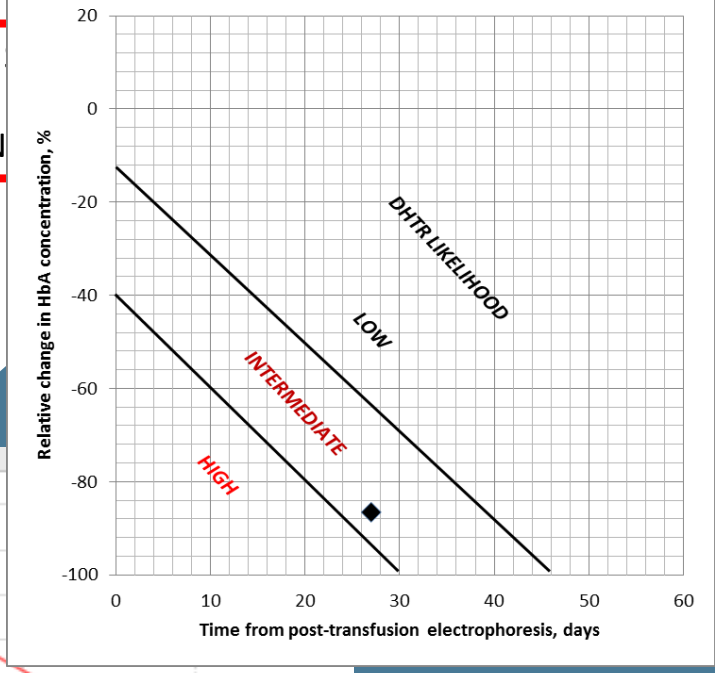
2018

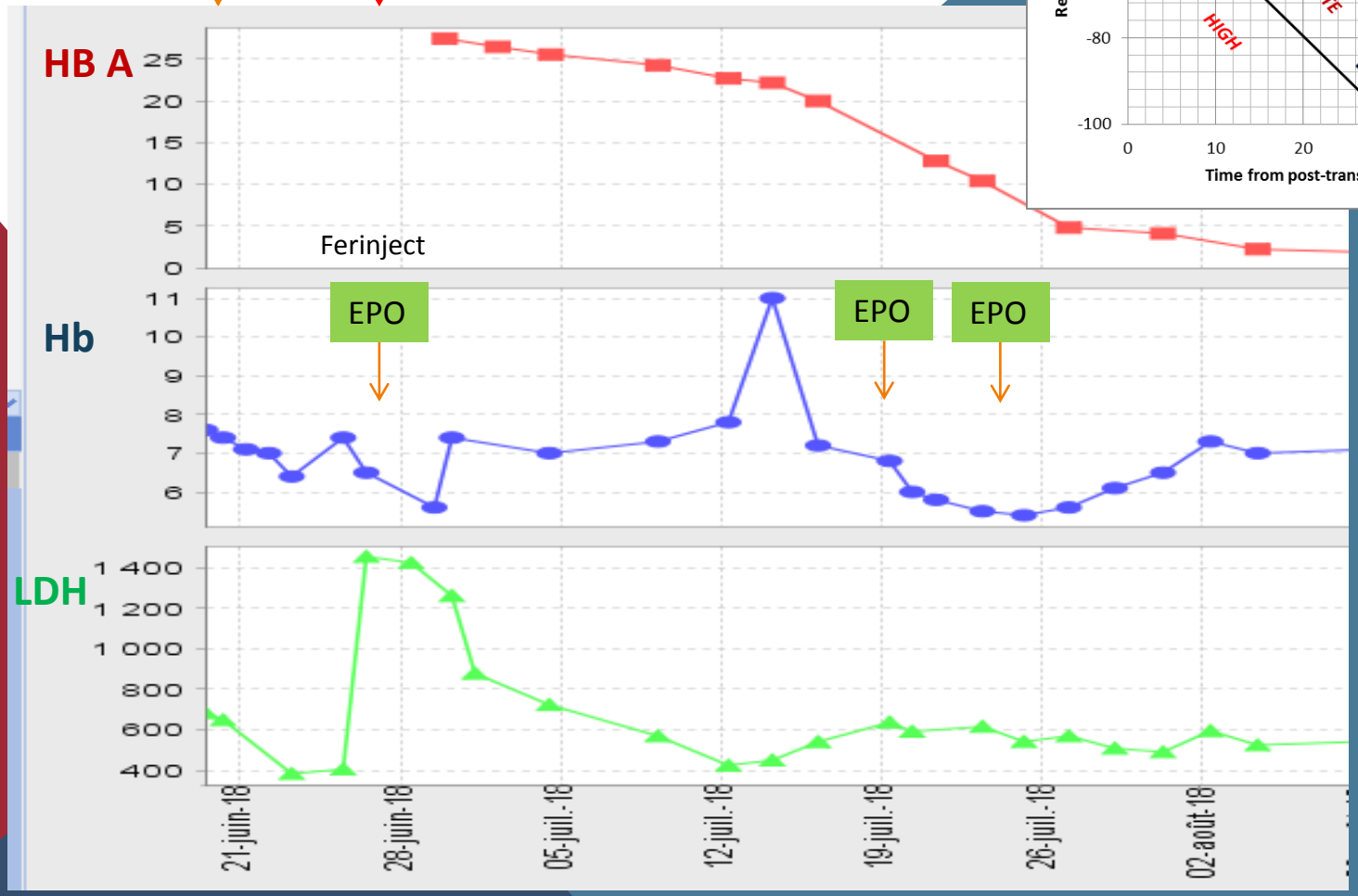
TF A+D+C-E- c+e+K- **Fya- Fyb-** Jka+Jkb-**M+** N-

TF B+D+C-E- c+e+K- **Fya- Fyb-** Jka+ Jkb+ **M+** N

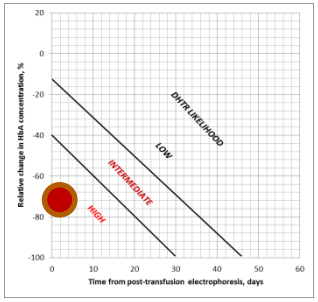
Rituximab

Steroids



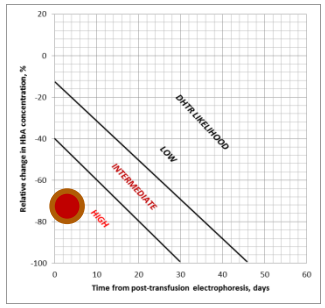


2010



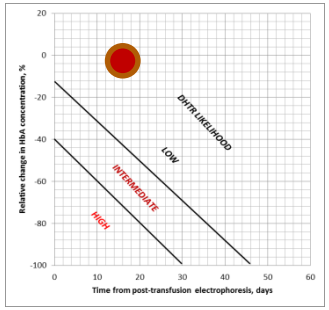
AB+ D+ C-E+ c+ e- K-
AB+ D+ C-E+ c+ e+ K-

2011



AB+ D+ C-E+ c+ e- K-
AB+ D+ C-E+ c+ e+ K-

2013

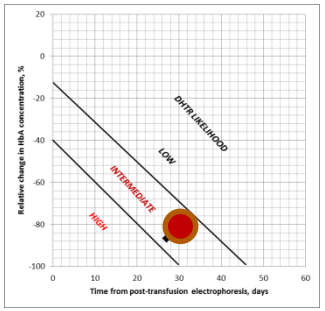


A+D+C-E+c+e+K- **Fya- Fyb-** Jka+Jkb-**M+** N+ S+ s+ Lea-**Leb+**

B+D+C-E+c+e+K- **Fya- Fyb-** Jka+ Jkb+ **M+** N+ S- s+ Lea-**Leb-**

Rituximab

2018



A+D+C-E- c+e+K- **Fya- Fyb-** Jka+Jkb-**M+** N- S- s+ Lea-**Leb+**

B+D+C-E- c+e+K- **Fya- Fyb-** Jka+ Jkb+ **M+** N+ S- s+ Lea-**Leb-**

Rituximab

AB+ C-E+c+e+K-**Fya-Fyb-**Jka+Jkb+**M-** N+S+s+**Lea-**Leb-P1

DAT: anti LeA, anti Hi, anti Cw

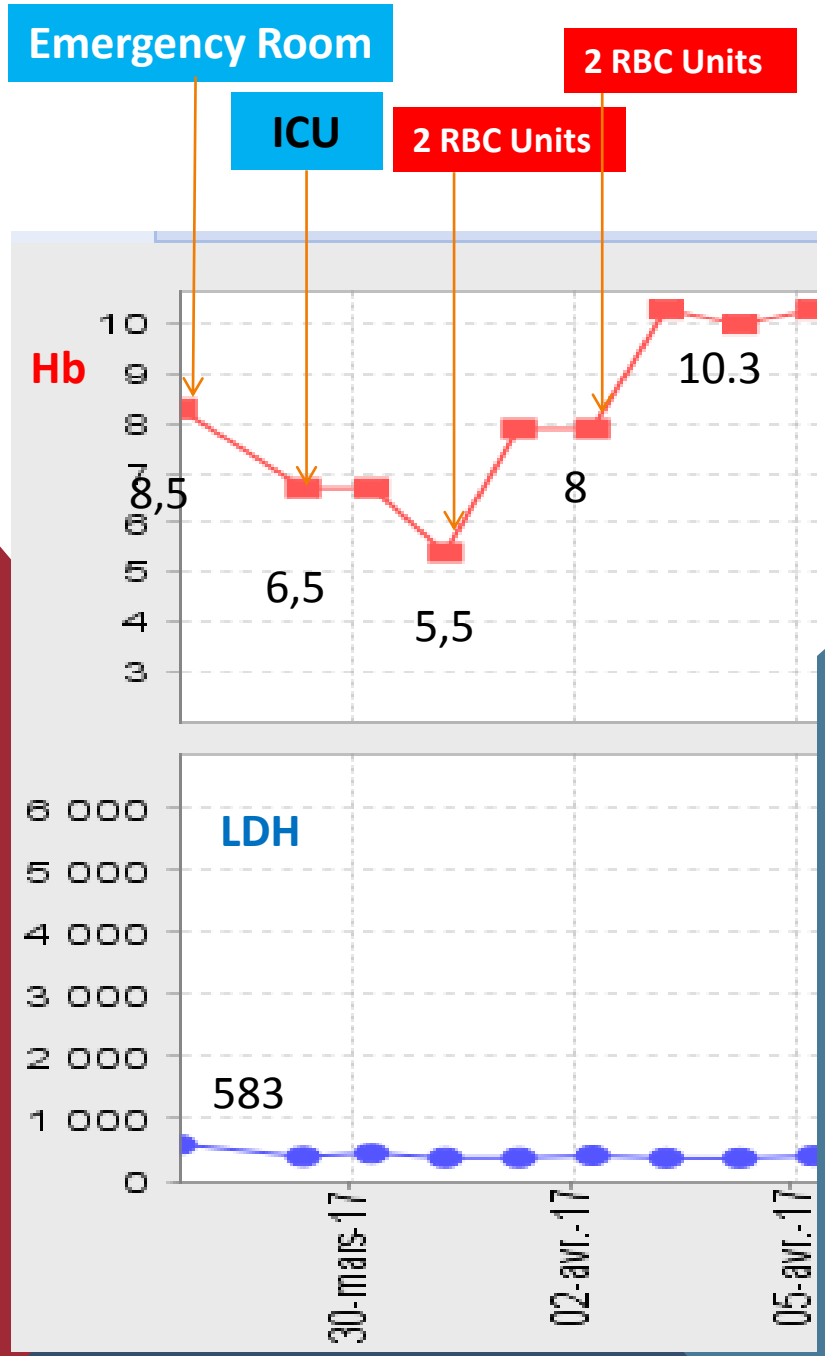
Since 2011 DAT are negative

CASE REPORT N°2

25-year-old woman, with Homozygous Sickle Cell Disease, is admitted in emergency unit for a vaso-occlusive crisis and chest pain

In her history :

- Hb at steady state : 9,5 g /d
- 5 ACS and multiple Crisis
- Splenectomy
- Number of transfusions: 40 Unites since 2000
- Prior screening test was positive in 2000 and since then negative
- Non History of transfusion problem
- Treatment by Hydroxycarbamid and folic acide



RBC Units : O-D-C-E-c+e+K- extended phenotype-matched crossmatch-compatible

post TF Hb : 10.3

post TF Hb A : 42.3%

post TF LDH :300

The clinical course was favorable after 3 days of treatment and two TF exchanges and the patient was transferred to internal medicine unit.

At Day 7 after the transfusion ,

reappearance of pain localized at both knees, without any other clinical sign or modification of the clinical examination.

Blood sampling on Day 9:

- anemia at 6.6 g / dL
- increase of LDH to 2800 UI/l

Suspicion of DHTR

Transfer to intensive care for monitoring and treatment

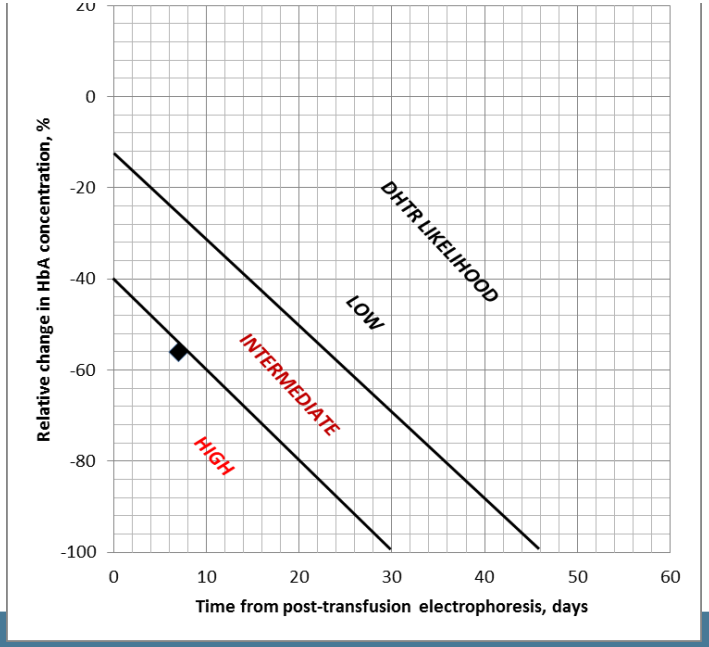
RESEARCH ARTICLE

AJH

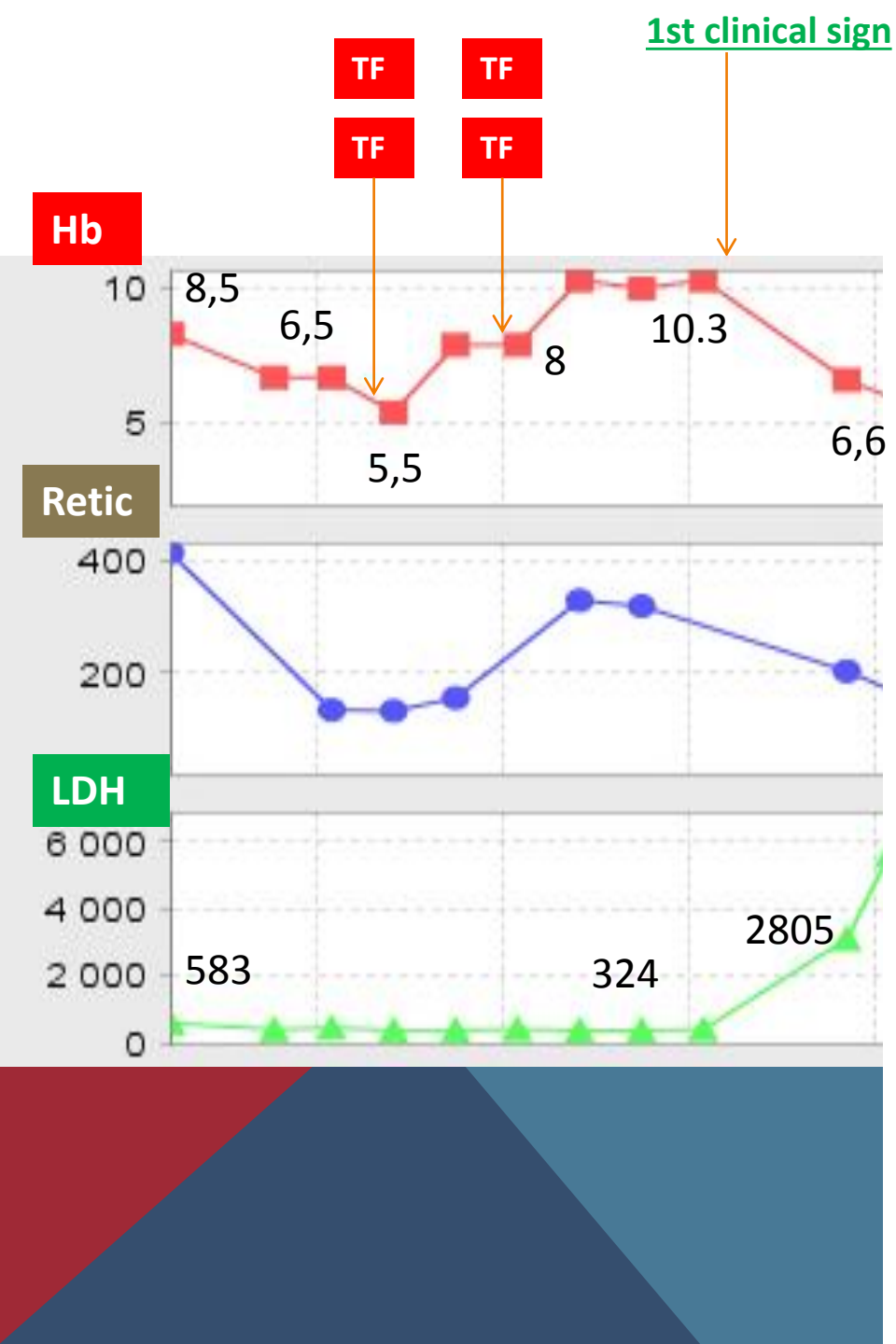
A diagnostic nomogram for delayed hemolytic transfusion reaction in sickle cell disease

Armand Mekontso Dessap,[1,2] France Pirenne,[3,4] Keyvan Razazi,[1,2] Stéphane Moutereau,[5] Shariq Abid,[1] Christian Brun-Buisson,[1,2] Bernard Maitre,[1,6] Marc Michel,[7] Frederic Galacteros,[4,8] Pablo Bartolucci,[4,8] and Anoosha Habibi[4,8]*

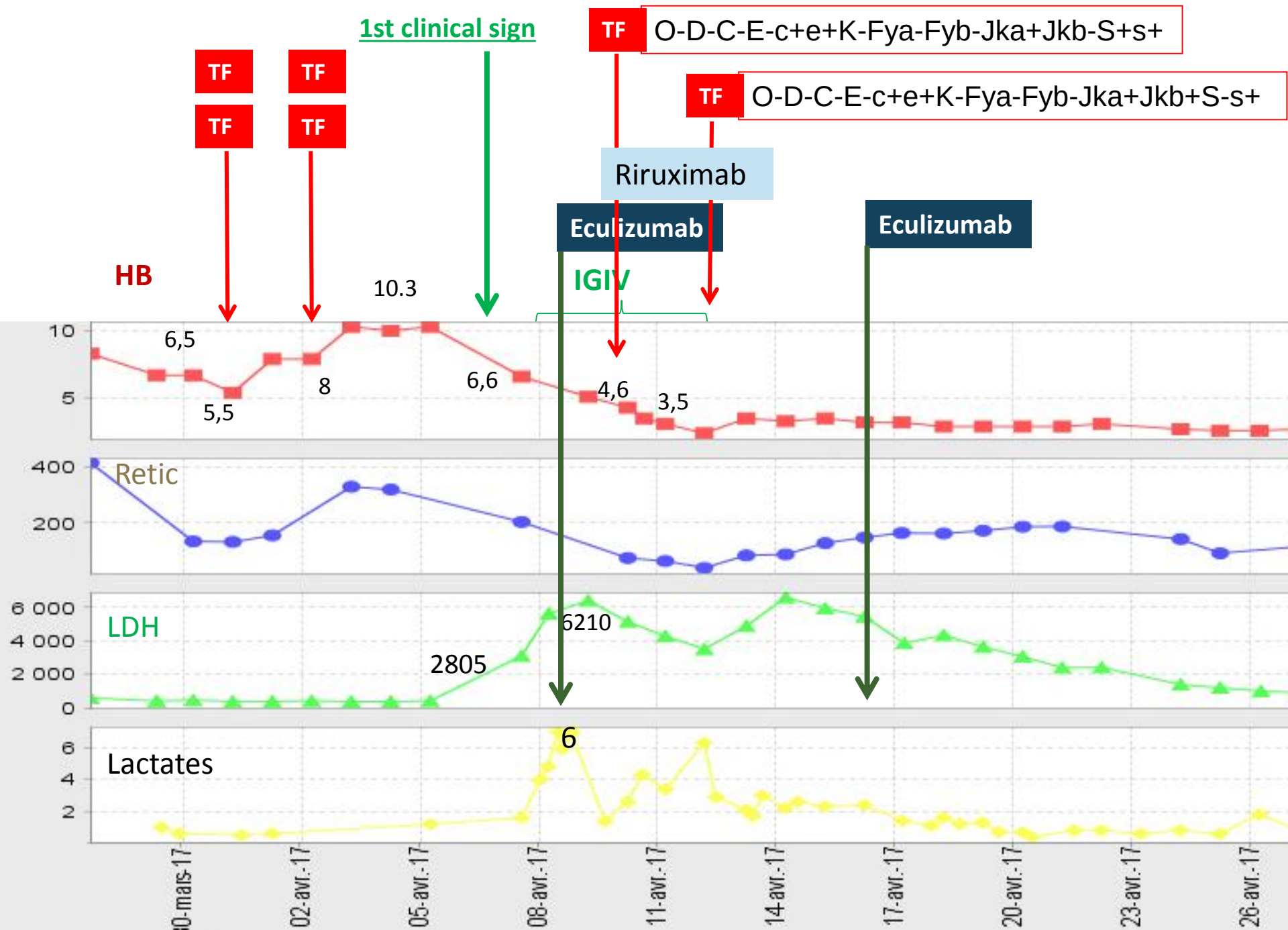
	1st assessment (AFTER the index transfusion)	2nd assessment (at DHTR suspicion)
Date	30/03/2017	07/04/2017
Total Hb, g/dL	10,3	6,6
HbA percentage, %	42,3	29,0



High risk of DHTR



Reapparence of pain in knee
High risk in Nomogram


1st clinical sign
TF
TF
TF
TF
TF
O-D-C-E-c+e+K-Fya-Fyb-Jka+Jkb-S+s+
TF
O-D-C-E-c+e+K-Fya-Fyb-Jka+Jkb+S-s+
Riruximab
Eculizumab
Eculizumab
IGIV
HB
10.3
6,5
5,5
8
6,6
4,6
3,5
10
5
Retic
400
200
LDH
6210
2805
6 000
4 000
2 000
0
Lactates
6
4
2
10-mars-17
02-avr.-17
05-avr.-17
08-avr.-17
11-avr.-17
14-avr.-17
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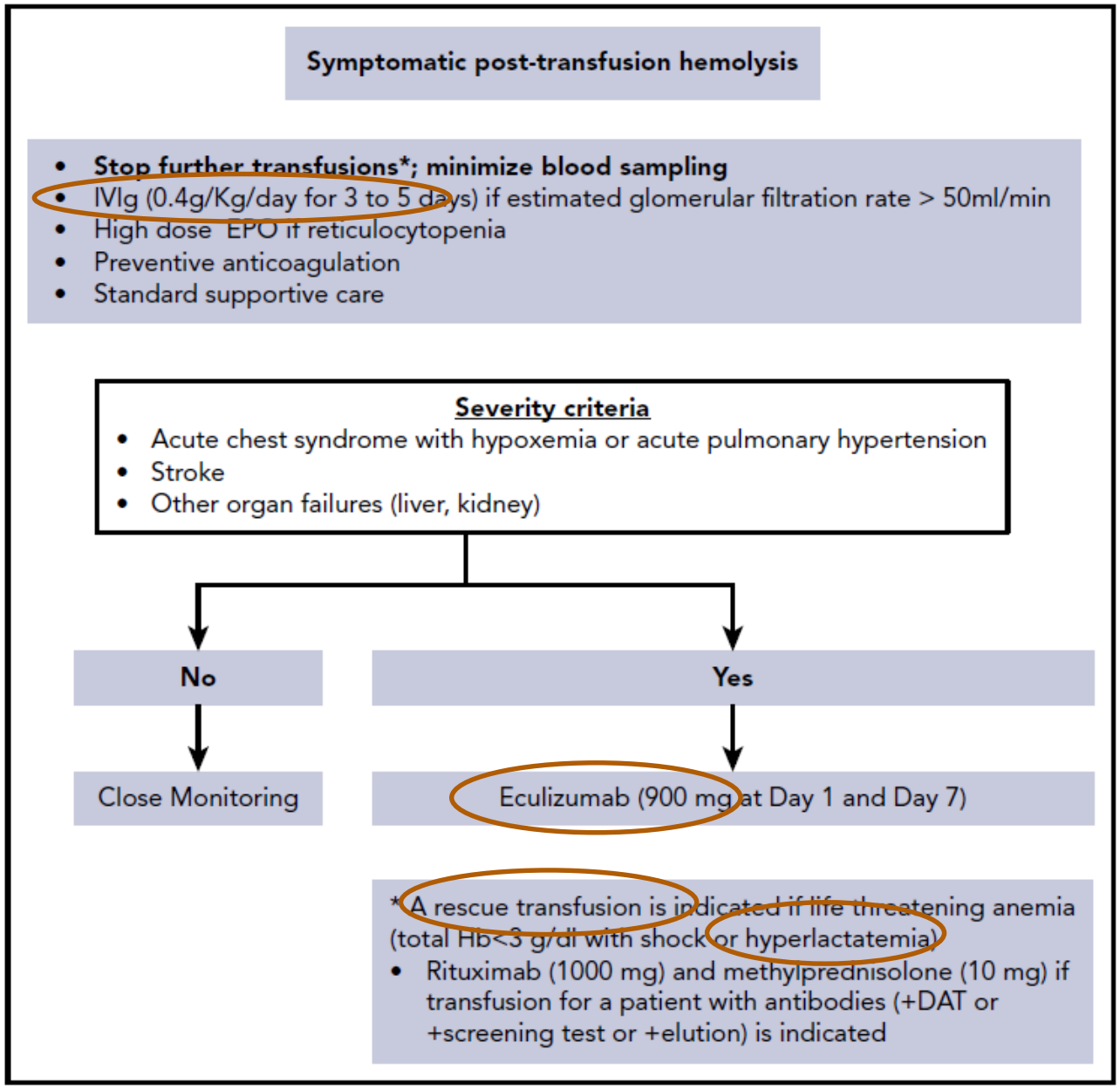
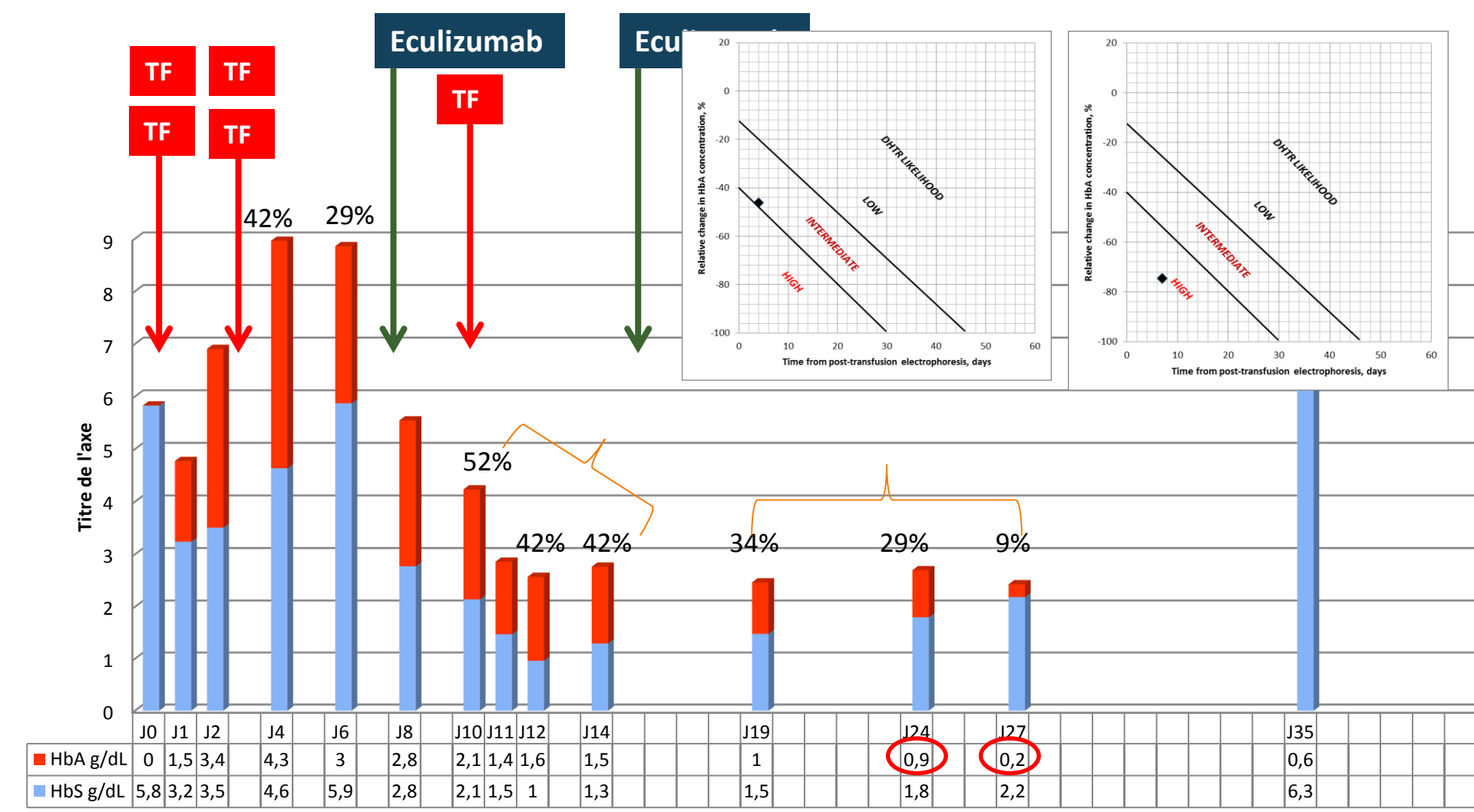
20-avr.-17
23-avr.-17
26-avr.-17



Pirenne Blood 2018



	J0	J1	J2	J4	J6	J8	J10	J11	J12	J14	J19	J24	J27	J35
HbA g/dL	0	1,5	3,4	4,3	3	2,8	2,1	1,4	1,6	1,5	1	0,9	0,2	0,6
HbS g/dL	5,8	3,2	3,5	4,6	5,9	2,8	2,1	1,5	1	1,3	1,5	1,8	2,2	6,3

TAKE AWAY

- Rituximab and Eculizumab improves randomly the effectiveness of the TF,
- however we have the impression that it mitigates the severity of reactions
- post TF HbA is required for early diagnosis
- Research on DHTR mechanisms is essential in order to offer other therapeutics

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