

Emoglobinuria Parossistica Notturna

Focus Clinico e Update Diagnostico

Caso Clinico - Ospedale di Legnano

- Uomo di 85 anni
- Diagnosi di MDS Trilineare nel 2005, responsivo a rH-EPO 40.000 U/settimana
- Emolisi Coombs-negativa subclinica, non indagata ma trattata con cicli di steroidi
- In TAO dal 2012 per episodi di FA
- Insufficienza renale nefrovascolare di grado moderato, ipotiroidismo.
- Diverticolosi con episodi di sanguinamento e infezione (Trasfusioni in fase acuta)
- Seguito presso altro centro, si riferisce a H Legnano nel 7/2018 per vicinanza.

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Sistema Sanitario

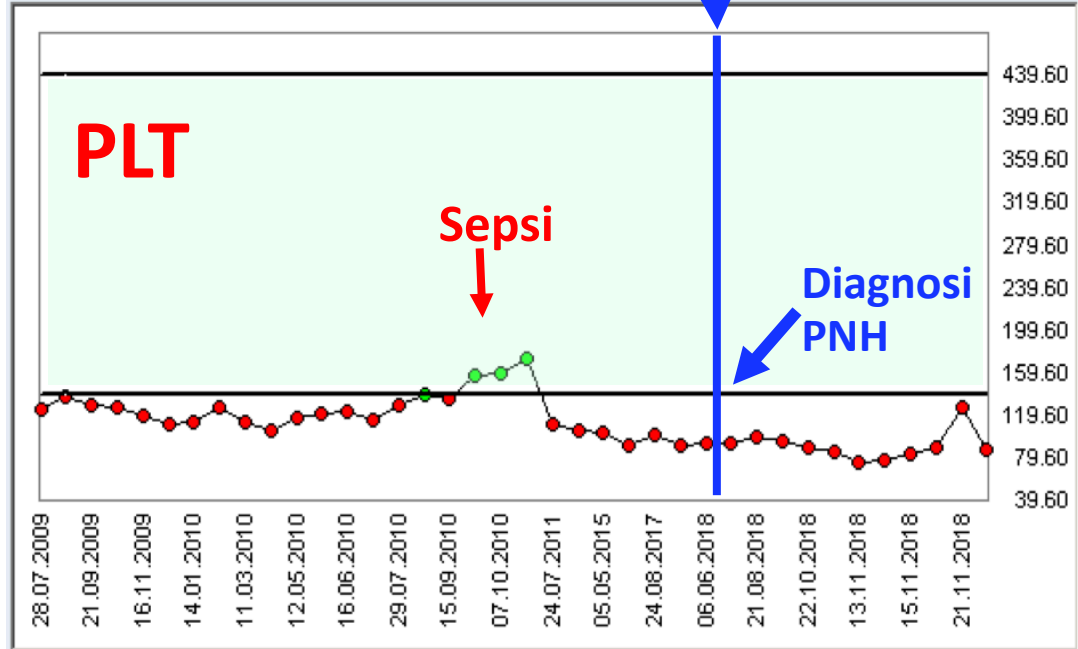
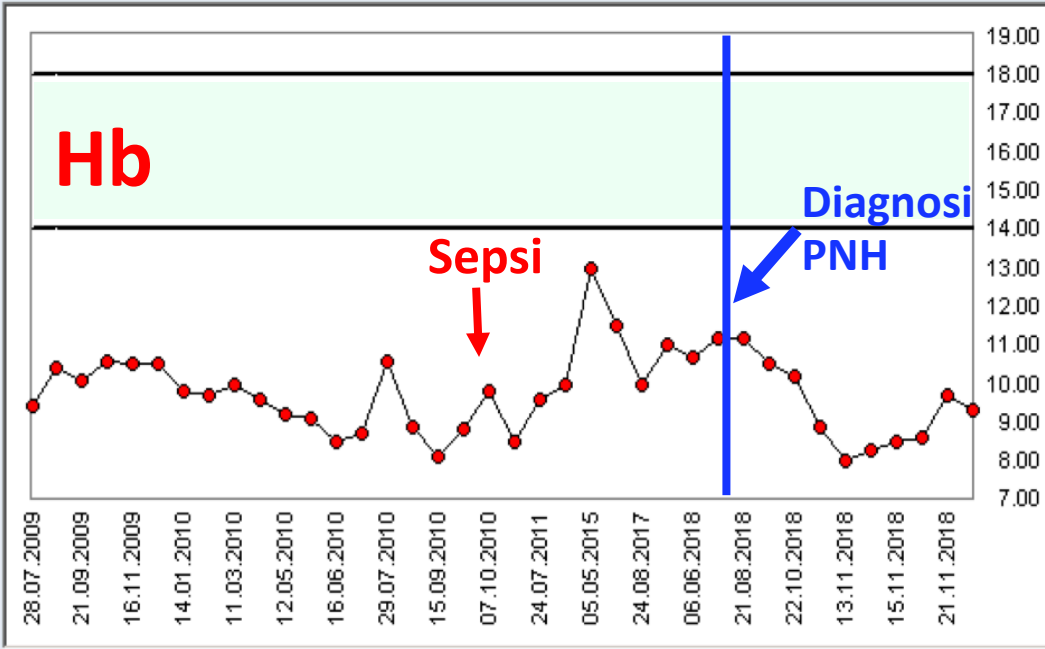


Regione
Lombardia

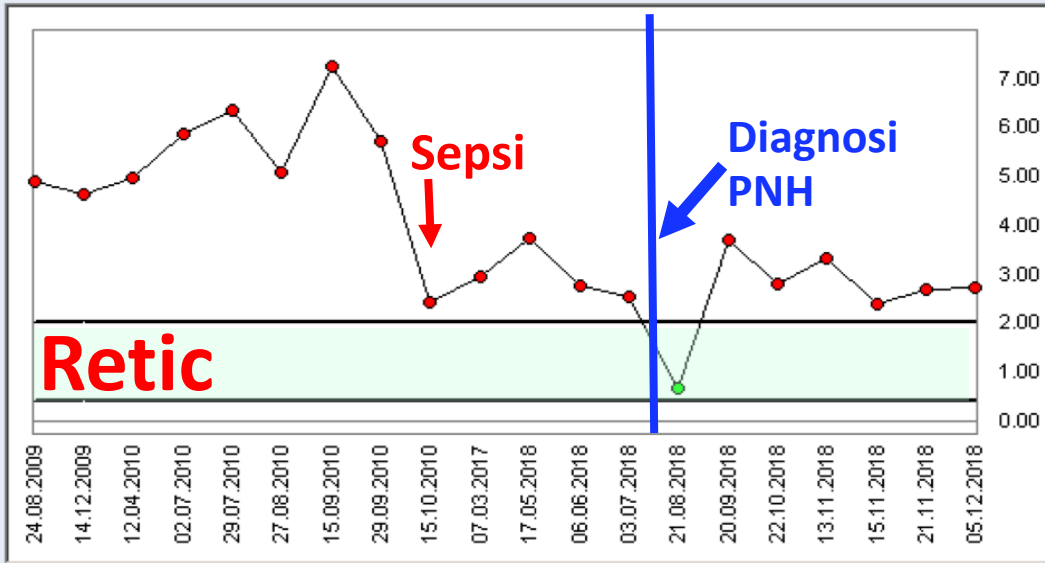


Caso Legnano - Emometria

Presca in Carico

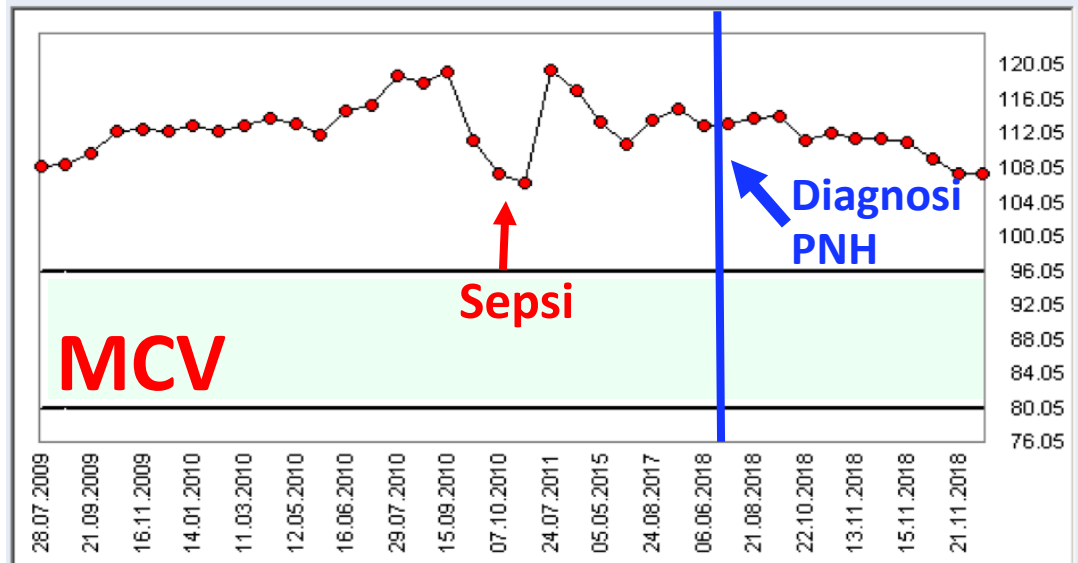


Analisi: 7001C - Emoglobina



Analisi: 7004 - Sg--CONTA RETICOLOCITI

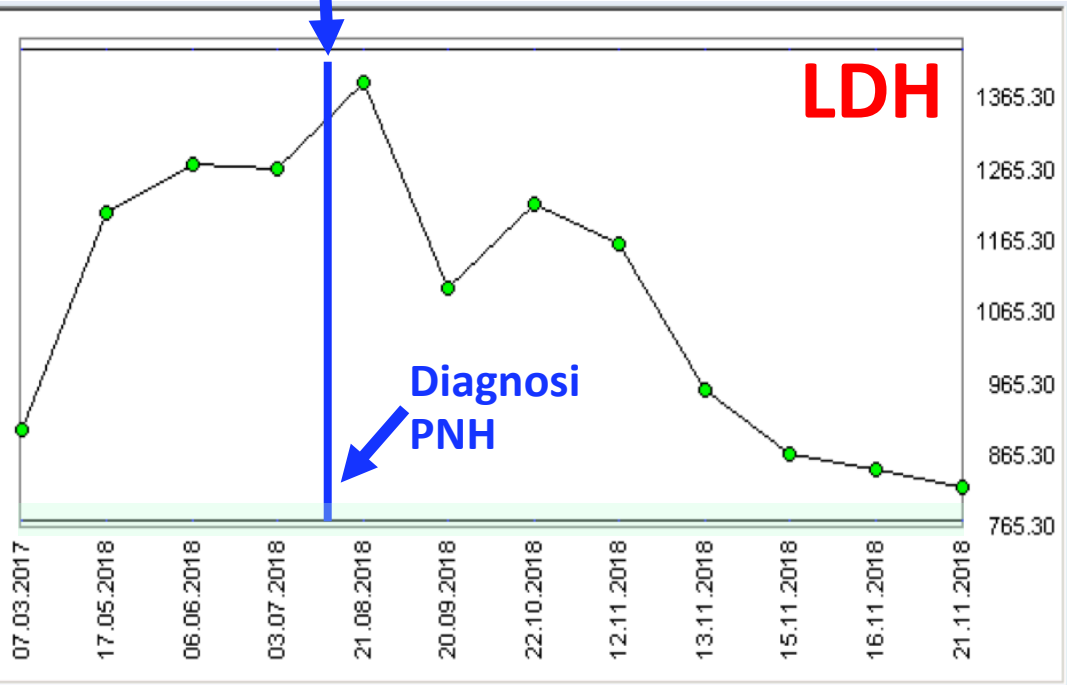
Analisi: 7001I - Piastrine



Analisi: 7001E - Volume Corpuscolare Medio

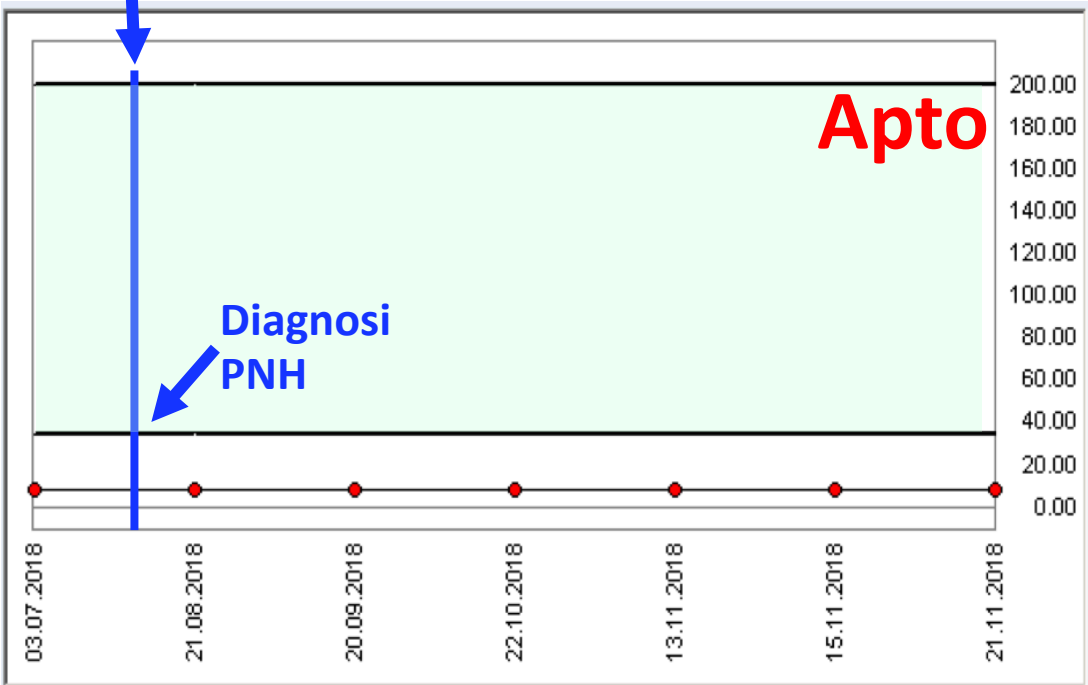
Caso Legnano - LDH e APTOGLOBINA

Presca in Carico



Analisi: 1170 - S--LATTATO DEIDROGENASI

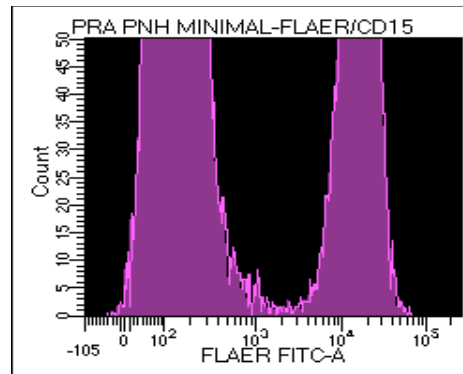
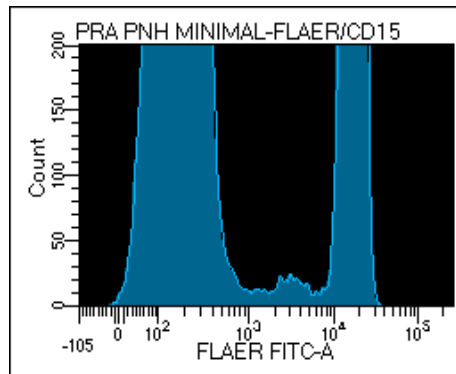
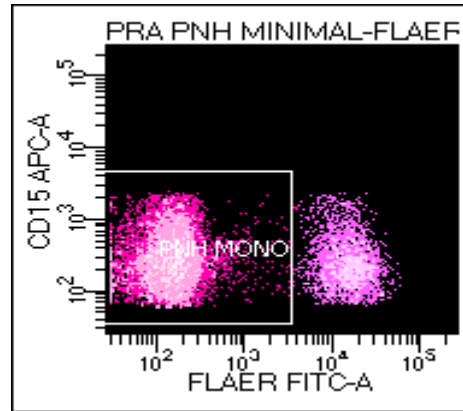
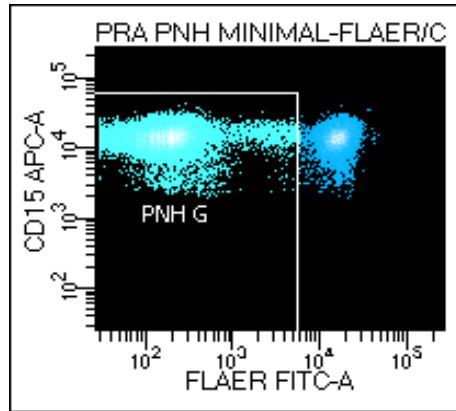
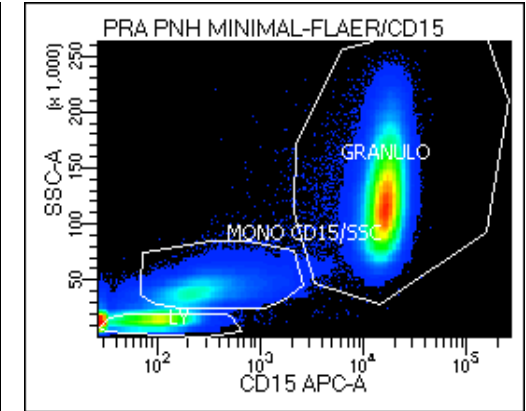
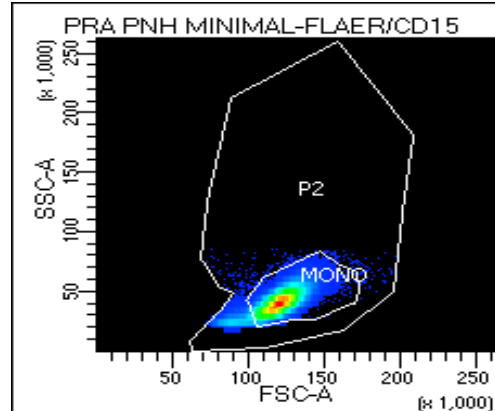
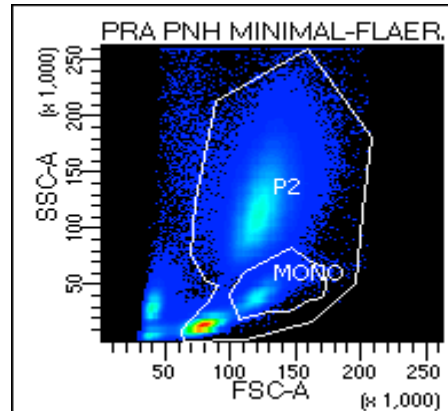
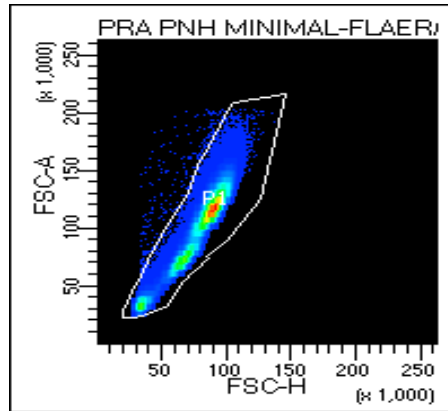
Presca in Carico



Analisi: 1417 - S--APTOGLOBINA

Caso Legnano - ANALISI CITOMETRICA

2-Color (FLAER/ CD15) Minimal Screening Analysis

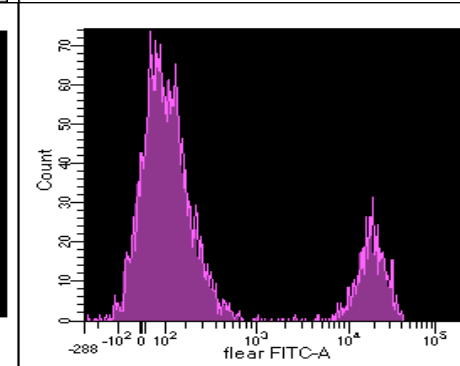
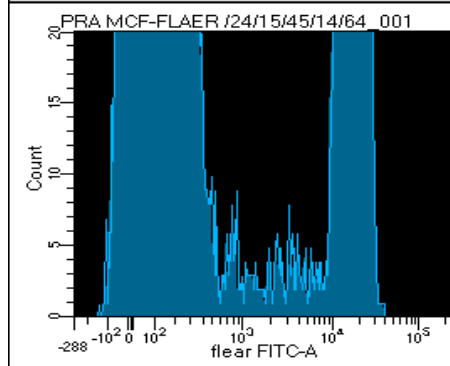
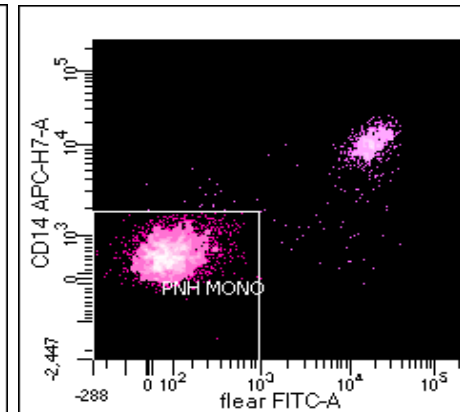
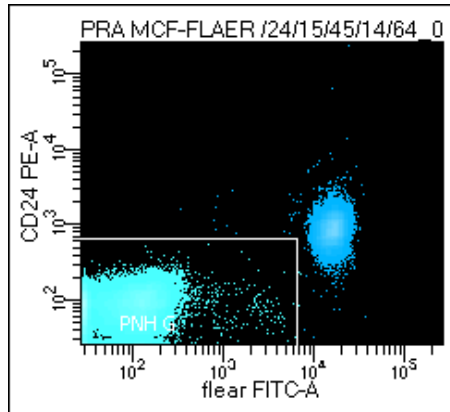
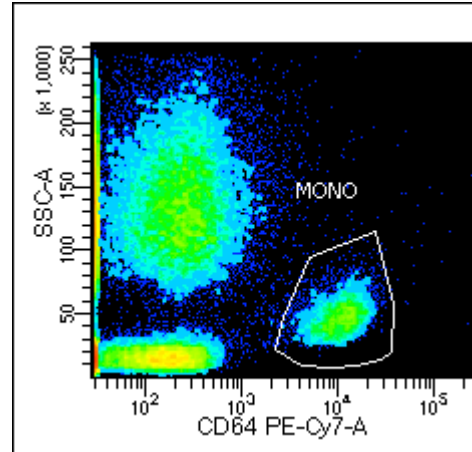
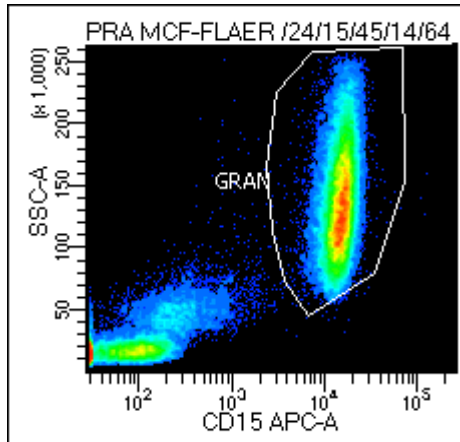
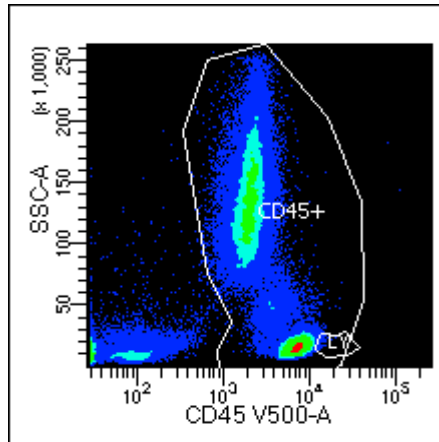


Tube: FLAER/CD15

Population	#Events	%Parent	%Total
All Events	219,190	####	100.0
P3	208,000	94.9	94.9
P1	207,536	99.8	94.7
P2	178,083	85.8	81.2
GRANULO	116,048	65.2	52.9
PNH G	83,346	71.8	38.0
LY	17,842	10.0	8.1
MONO CD15/SSC	20,772	11.7	9.5
MONO	18,377	88.5	8.4
PNH MONO	15,058	81.9	6.9

Caso Legnano - ANALISI CITOMETRICA

6-Color MCF Analysis

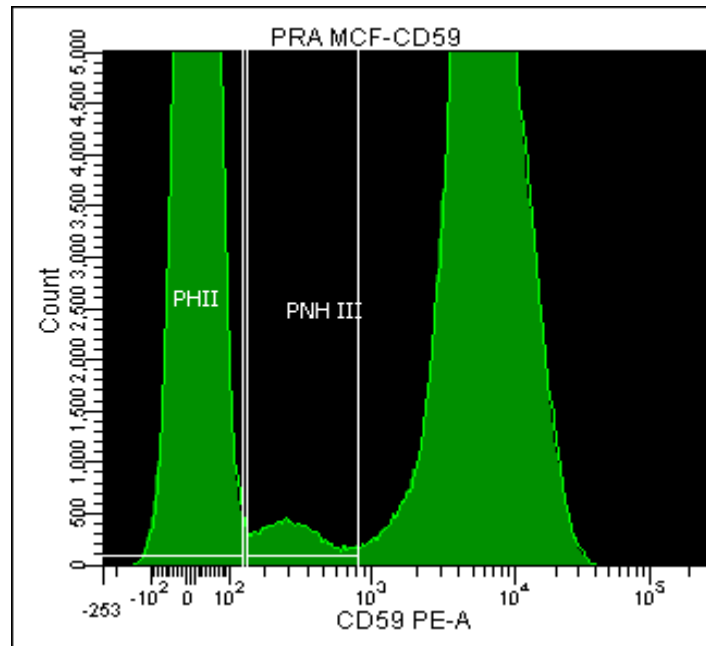
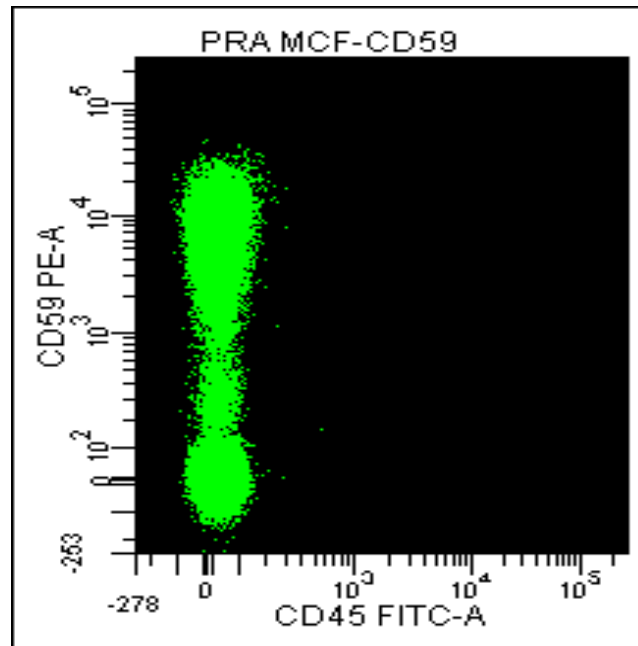


Tube: FLAER /24/15/45/14/64_001

Population	#Events	%Parent
All Events	42,412	###
P2 Singlets	42,067	99.2
P3 FSC/SSC	38,325	91.1
CD45+	32,834	85.7
GRAN	22,095	67.3
PNH G	15,869	71.8
MONO	2,891	8.8
PNH MONO	2,344	81.1
LY	71	0.2

Caso Legnano - ANALISI CITOMETRICA

RBC Analysis



Tube: CD59

Population	#Events	%Parent	%Total
<input type="checkbox"/> All Events	918,643	####	100.0
<input checked="" type="checkbox"/> P5	855,990	93.2	93.2
<input checked="" type="checkbox"/> P1	665,331	77.7	72.4
<input checked="" type="checkbox"/> P2	664,091	99.8	72.3
<input checked="" type="checkbox"/> P3	577,246	86.9	62.8
<input checked="" type="checkbox"/> PNH III	249,842	43.3	27.2
<input checked="" type="checkbox"/> PNH II	15,500	2.7	1.7

Caso Legnano - **CONCLUSIONI**

- Paziente anziano, con franca MDS chiaramente diagnosticata, passa **13 ANNI** con emolisi Coombs-negativa di basso grado, responsiva a rH-EPO e ben compensata, senza che nessuno se ne accorga, malgrado **'Reasons for Testing'**. *'Salvato' da TAO?*
- Solo occasionale fabbisogno trasfusionale, in occasione di episodi di sepsi o sanguinamento da diverticolite.
- L'analisi citometrica (Agosto 2018) mostra cloni PNH di grandi dimensioni nei tre compartimenti cellulari (Neutrofili= **72%**; Monociti= **81%**; Eritrociti= **43%** tipo III e **3%** tipo II).
- Caso sottoposto alla Commissione REL (Ottobre 2018) per segnali di progressiva resistenza a rH-EPO, giudicato **NON ELEGGIBILE** al trattamento con **Eculizumab** per l'età avanzata e la pluri-patologia.
- Dicembre 2018: Prosegue il follow-up con il solo supporto trasfusionale.