



**BOLOGNA**

**17 FEBBRAIO 2023**

**NH De La Gare**

# **POLICITEMIA VERA NEL 2023:**

**qualcosa è cambiato**

## **Paziente con aftosi del cavo orale**

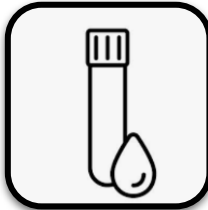
Camilla Mazzoni Istituto di Ematologia «L. e A. Seràgnoli», Università degli Studi di Bologna, IRCCS S. Orsola-Malpighi

**No Disclosures**

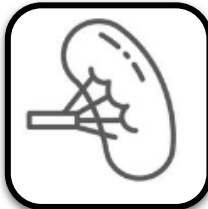


# CLINICAL CASE – Jul 2015

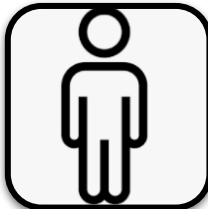
MAN 58 YRS OLD– Former smoker, previous eradicated IFN+ribavirin therapy for HCV infection, depression



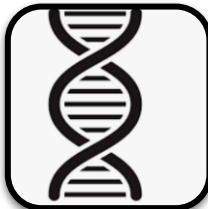
WBCs  $7.3 \times 10^9/L$  (normal differential count), Hb 16 g/dl, Hct 51.8%, PLT  $540 \times 10^9/L$ , LDH within normal limit, EPO 2.1 mIU/mL, normal iron deposits



Mild splenomegaly showed by abdomen scan (12,5 cm)  
moderate hepatosteatosi



Moderate itching after water contact, weight loss



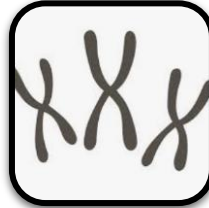
JAK2 V617F (VAF 21%)

# CLINICAL CASE – Sep 2015

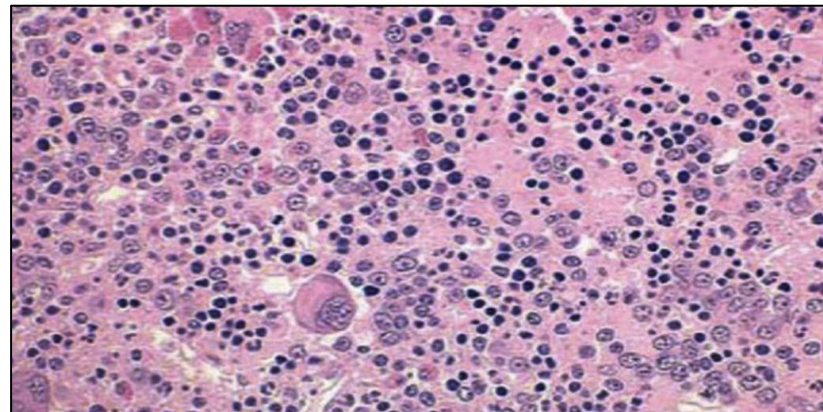
## PV low risk



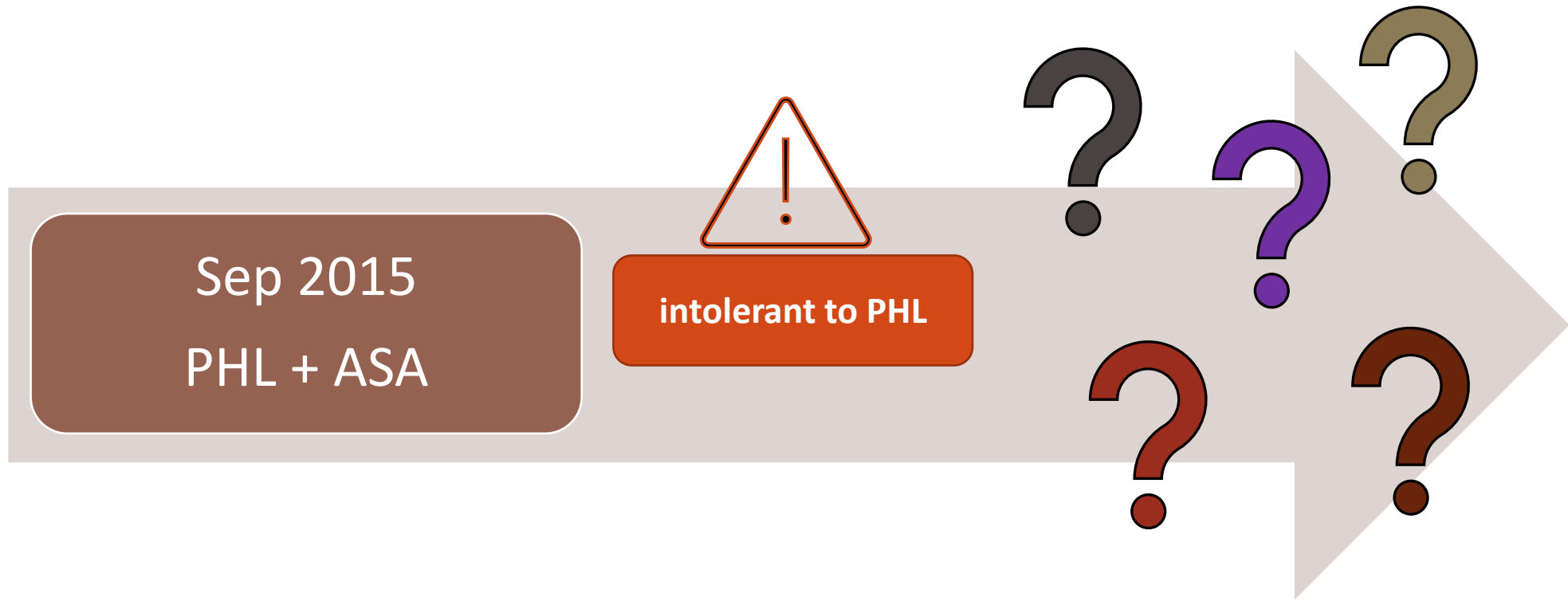
Age-matched hypercellular bone marrow with trilinear expansion, CD34+ precursors of 2-3%, fibrosis G0



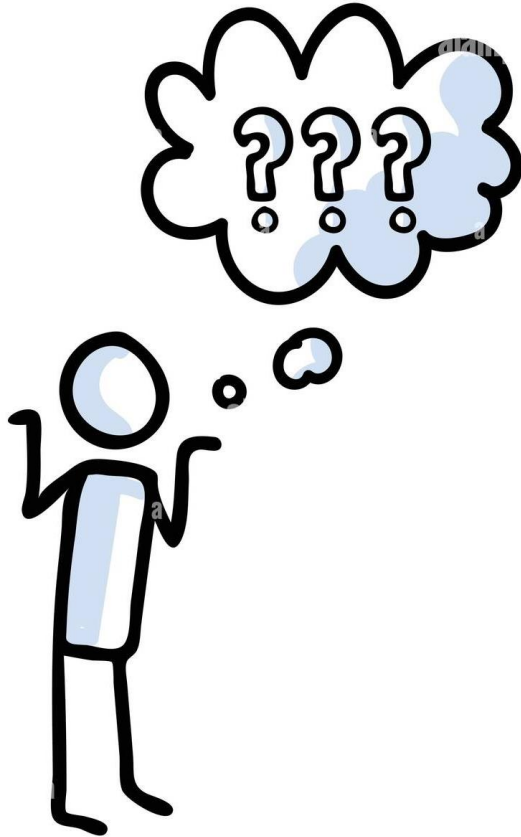
46, XY[22]/47, XY, +8[4]



# CLINICAL CASE



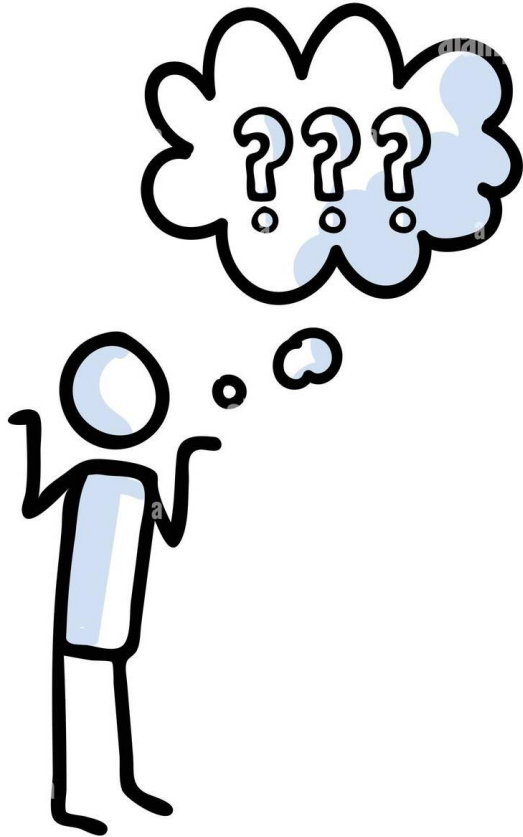
# CLINICAL CASE



## How to go on?

1. Just keep it up with PHLs, target hct 48-50%
2. Start HU
3. Start Peg-rIFN-a
4. Start RUX

# CLINICAL CASE



## How to go on?

1. Just keep it up with PHLs, target hct 48-50%
2. **Start HU**
3. Start Peg-rIFN-a
4. Start RUX



# CLINICAL CASE

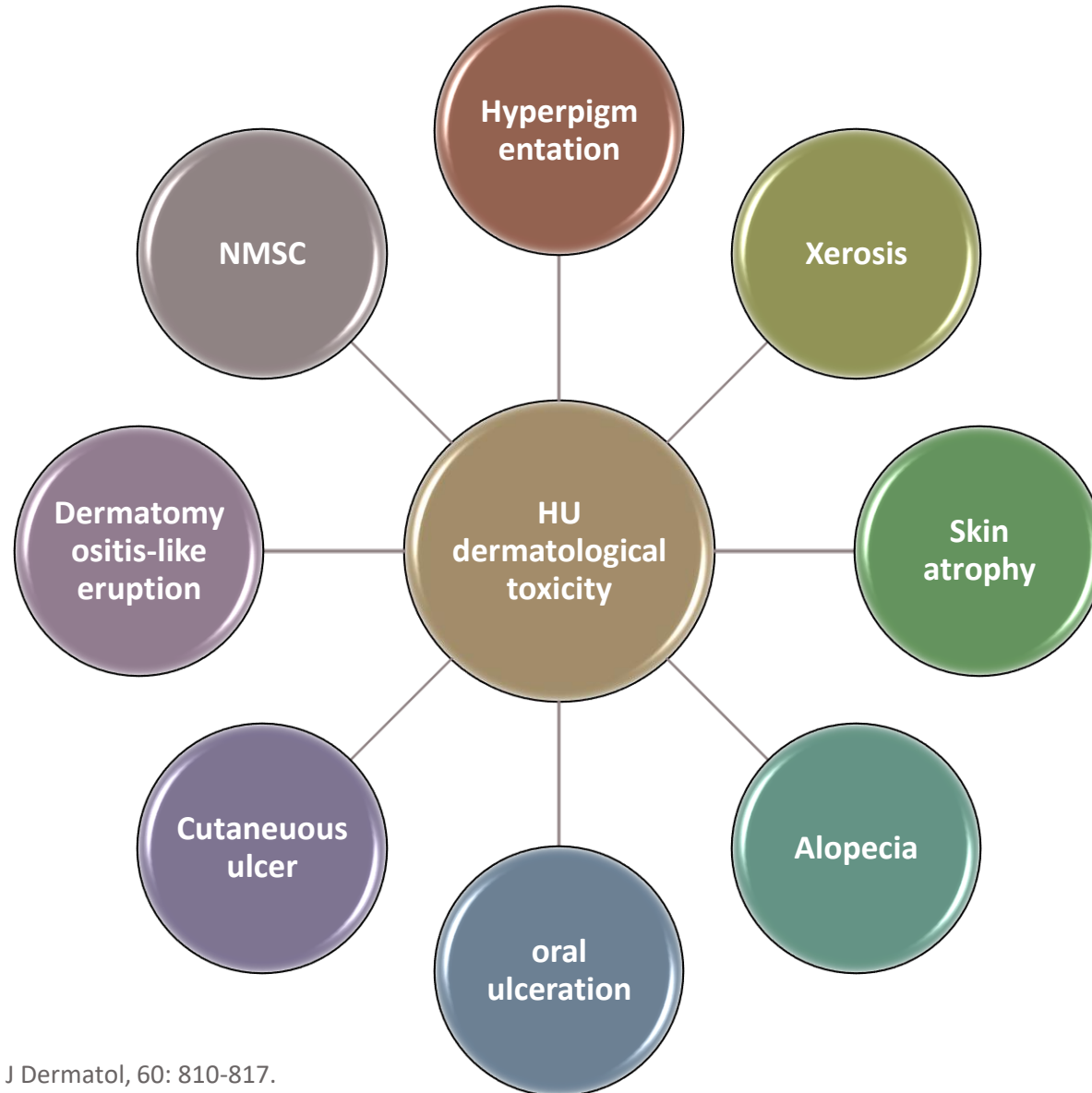


	Dec 2015 HU start	Jul 2017
<b>WBC</b>	6.8 k/ $\mu$ L	6.5k/ $\mu$ L
<b>Hgb</b>	17 g/dL	14.7 g/dL
<b>PLTs</b>	307 k/ $\mu$ L	334 k/ $\mu$ L
<b>Hct</b>	54.7 %	49.5%
<b>No of PHL/yr</b>	2	0
<b>MPN-related symptoms</b>	Itching, fatigue	Fatigue ++



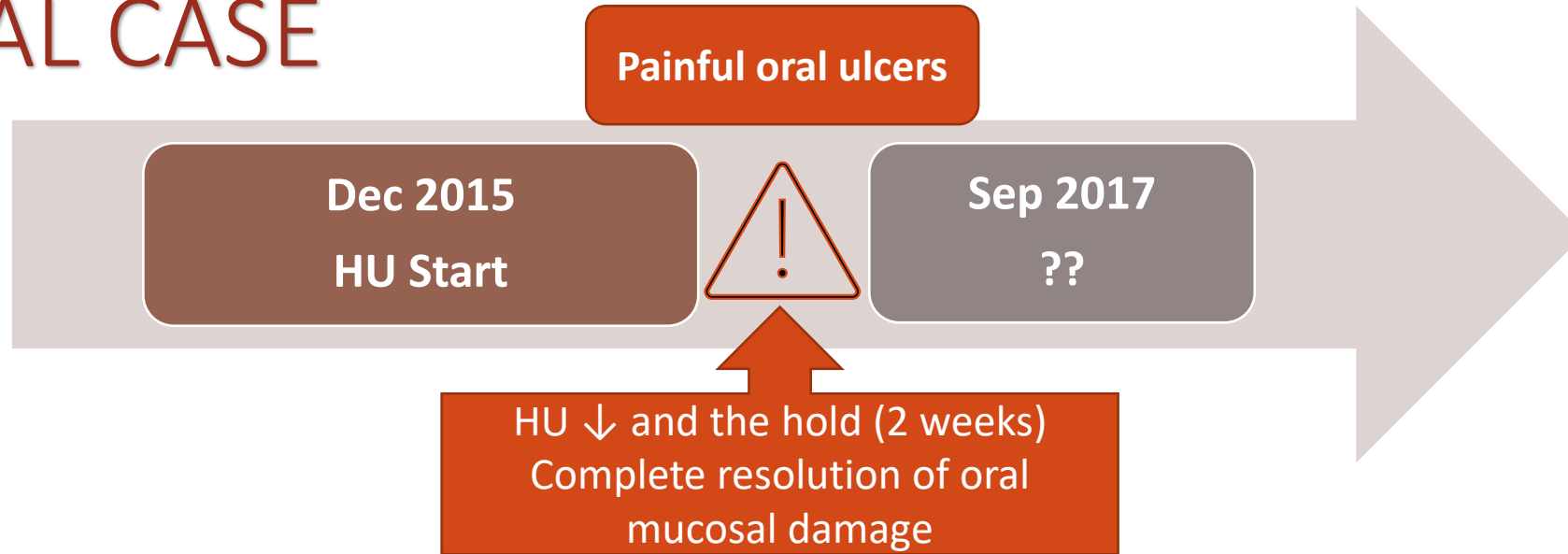


# Hydroxyurea: cutaneous side effects



Bulte, C.A. et al. Int J Dermatol, 60: 810-817.

# CLINICAL CASE



	Dec 2015 HU start	Jul 2017	Sep 2017 After HU hold
<b>WBC</b>	6.8 k/ $\mu$ L	6.5k/ $\mu$ L	6.5k/ $\mu$ L
<b>Hgb</b>	17 g/dL	14.7 g/dL	15,6 g/dL
<b>PLTs</b>	307 k/ $\mu$ L	334 k/ $\mu$ L	339 k/ $\mu$ L
<b>Hct</b>	54.7 %	49.5%	50 %
<b>No of PHL/yr</b>	2	0	0
<b>MPN-related symptoms</b>	Itching, fatigue	Fatigue ++	Itching, Fatigue ++

# CLINICAL CASE

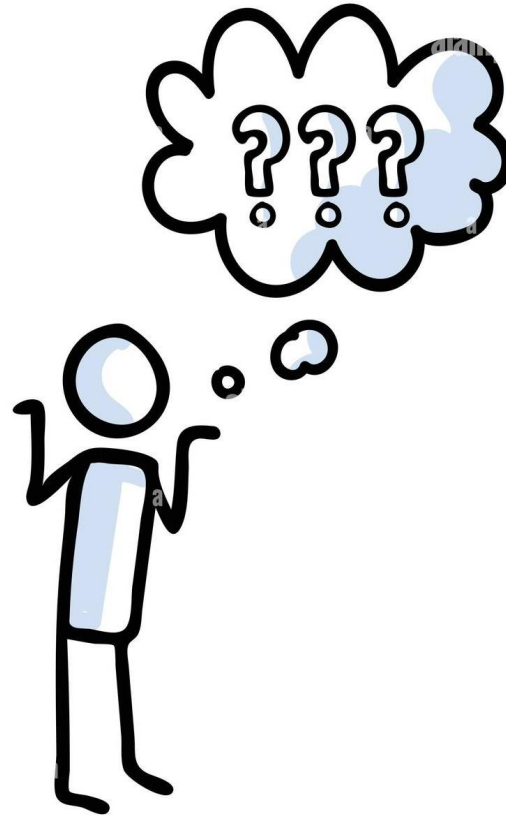
High risk PV patient (60 years-old)

No PHL possibility

Symptomatic for itching

Depression in active treatment

HU moderate cutaneous AE

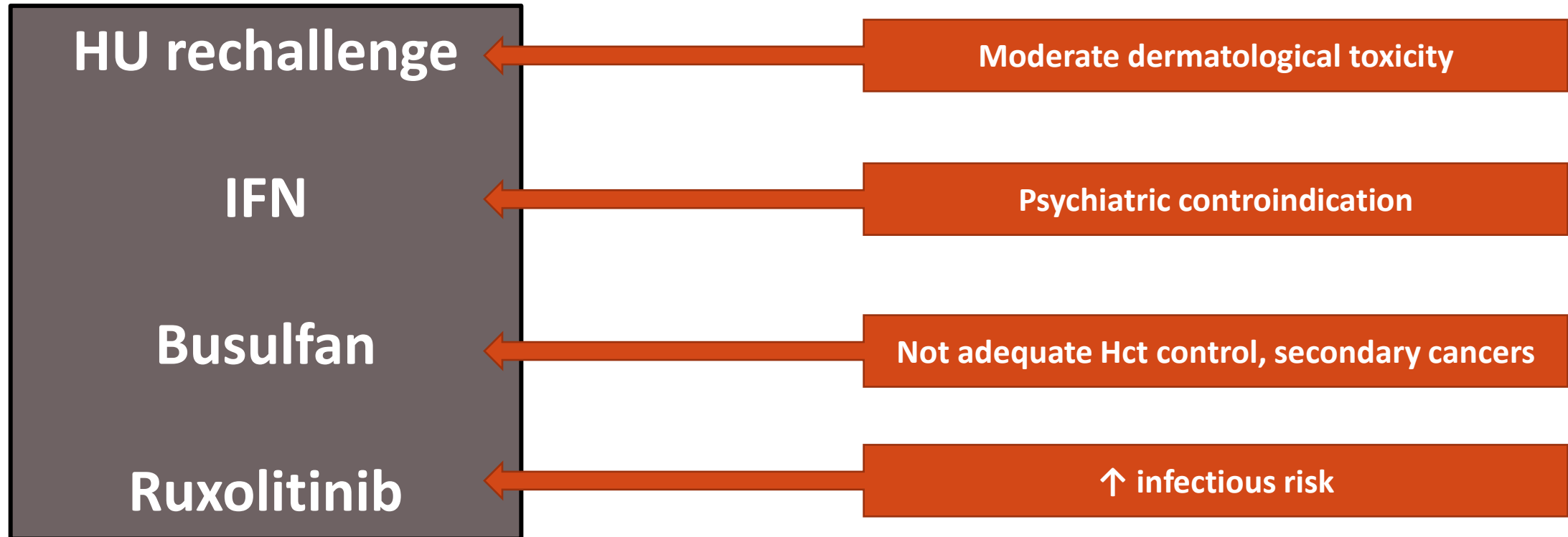


## What to do next?

1. HU rechallenge
2. IFN
3. Busulfan
4. Ruxolitinib



# CLINICAL CASE



# CLINICAL CASE

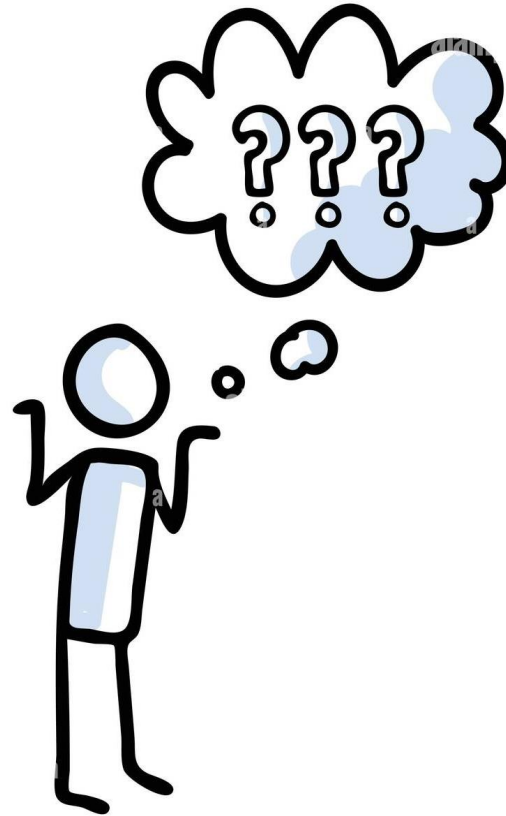
High risk PV patient (60 years-old)

No PHL possibility

Symptomatic for itching

Depression in active treatment

HU moderate cutaneous AE



## What to do next?

1. HU rechallenge

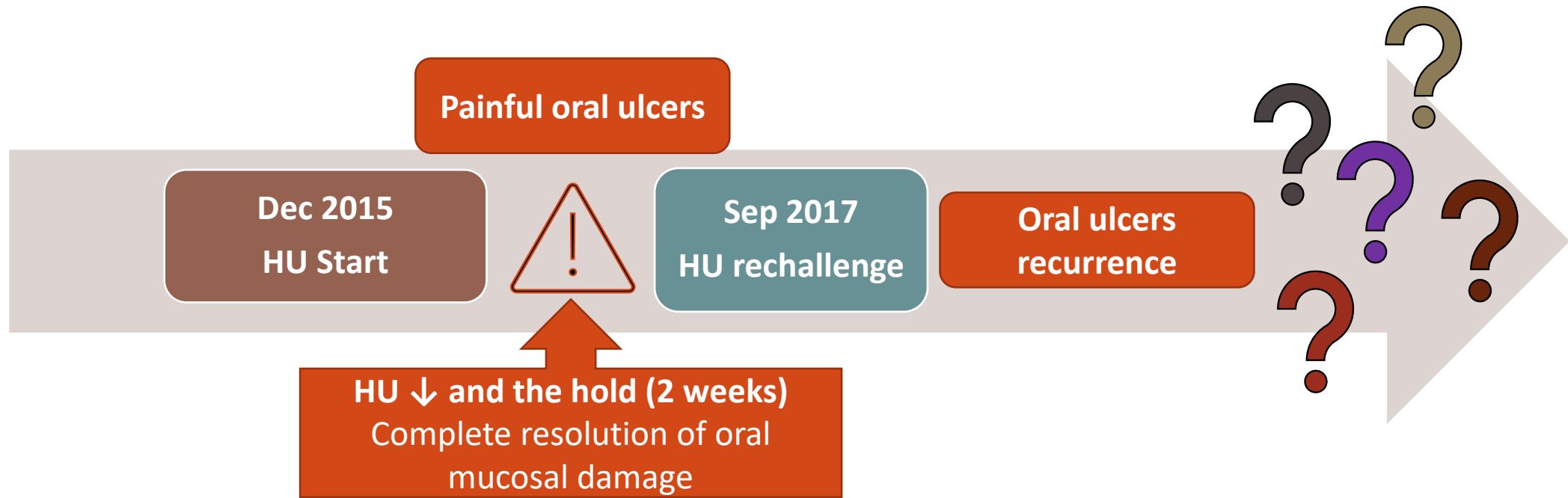
2. IFN

3. Busulfan

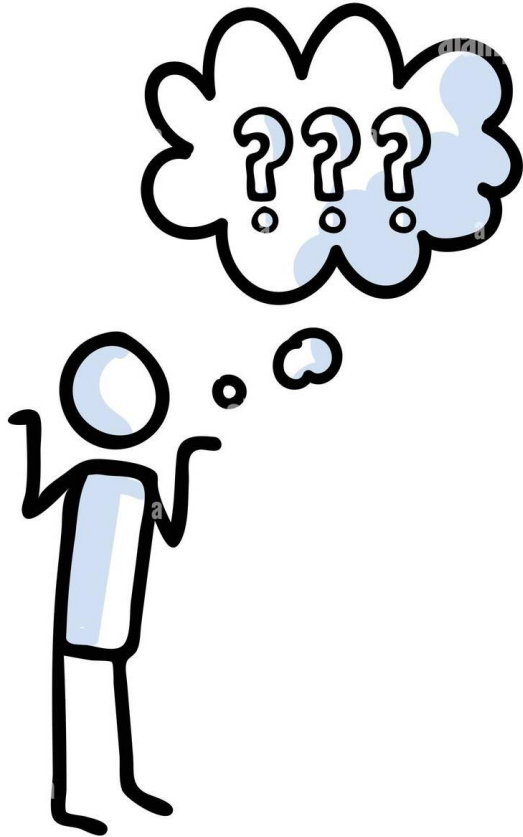
4. Ruxolitinib (pt refusal)



# CLINICAL CASE



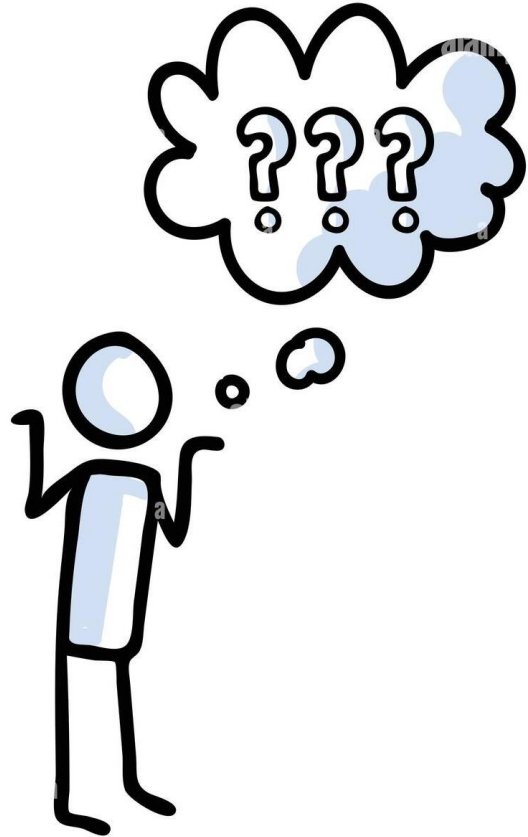
# CLINICAL CASE



## What about now?

1. Just keep it up HU, ulcers will resolve eventually
2. Start Peg-rIFN-a
3. Start RUX
4. Start BUS

# CLINICAL CASE



## What about now?

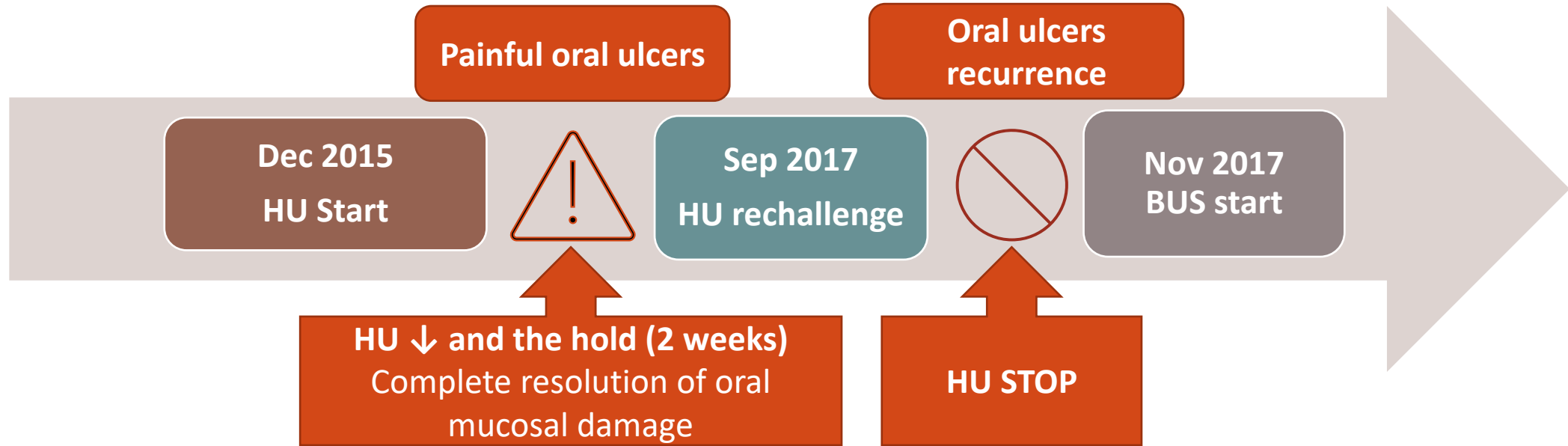
1. Just keep it up HU, ulcers will resolve eventually
2. Start Peg-rIFN-a
3. Start RUX
4. **Start BUS**

Not yet available!!

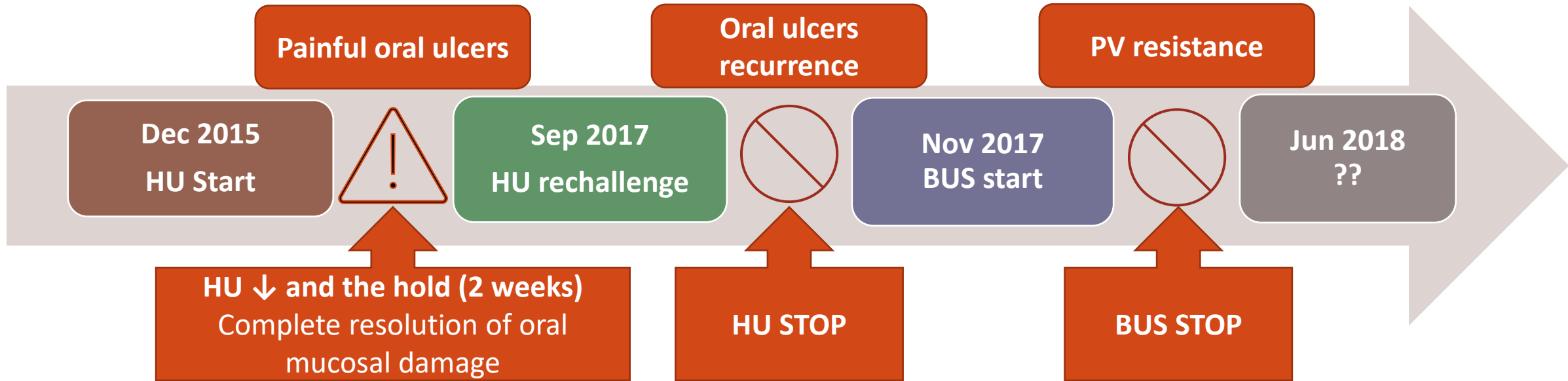




# CLINICAL CASE



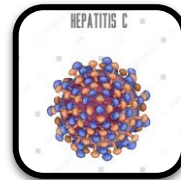
# CLINICAL CASE



	Dec 2015 HU start	Jul 2017	Sep 2017 After HU hold	Nov 2017 BUS star	Jun 2018
<b>WBC</b>	6.8 k/ $\mu$ L	6.5k/ $\mu$ L	6.8k/ $\mu$ L	7.3k/ $\mu$ L	7.3k/ $\mu$ L
<b>Hgb</b>	17 g/dL	14.7 g/dL	15,6 g/dL	14.7 g/dL	14.0 g/dL
<b>PLTs</b>	307 k/ $\mu$ L	334 k/ $\mu$ L	339 k/ $\mu$ L	275 k/ $\mu$ L	205 k/ $\mu$ L
<b>Hct</b>	54.7 %	49.5%	50 %	49.5%	48%
<b>MPN-related symptoms</b>	Itching, fatigue	Fatigue ++	Itching, Fatigue ++	Fatigue++	Itching++, Fatigue ++

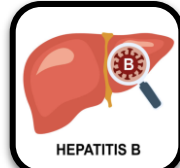
# CLINICAL CASE

## Screening for RUX start



HEPATITIS C

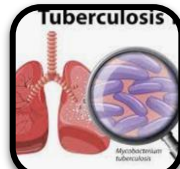
HCV RNA neg



HEPATITIS B

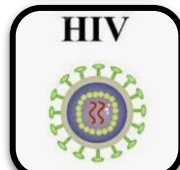
Anti Hbc pos  
HBV DNA neg

Lamivudine  
prophylaxis



Tuberculosis

Quantiferon neg



HIV

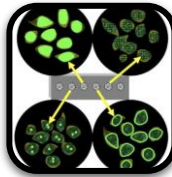
HIV neg



PV, fibrosis G1

# CLINICAL CASE

## Screening for IFN- $\alpha$ start



ANA reflex 1:160 (speckeled)



Anti thyreo-globulin and anti thyreo-peroxidase antibodies  
neg



Rheumatoid factor and anti-phospholipid antibodies within  
normal limits



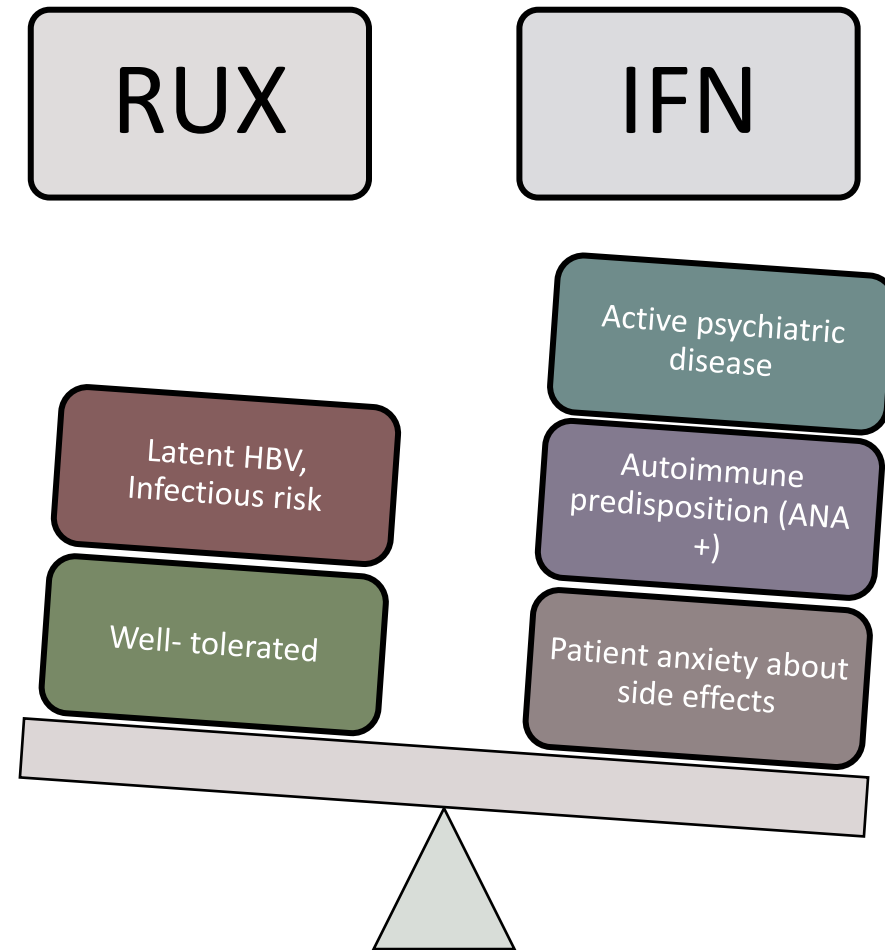
No hystory of diabetes or other autoimmune disease



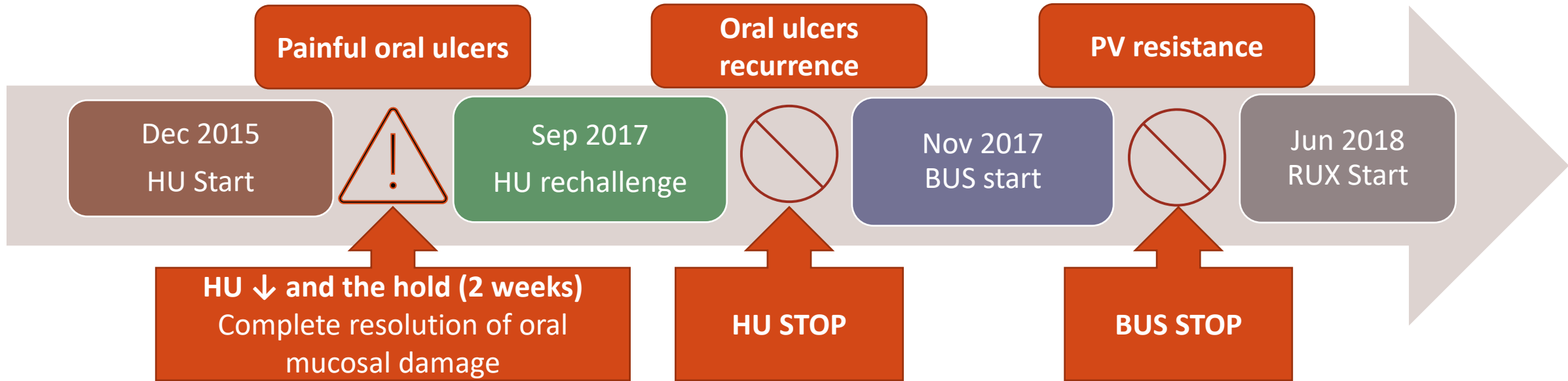
Depression still in active treatment



# CLINICAL CASE: the balance between RUX and IFN must be personalized



# CLINICAL CASE



	Dec 2015 HU start	Jul 2017	Sep 2017 After HU hold	Nov 2017 BUS start	Jun 2018 Rux Start	Dec 2018 + 6 months RUX
<b>WBC</b>	6.8 k/ $\mu$ L	6.5k/ $\mu$ L	6.8k/ $\mu$ L	7.3k/ $\mu$ L	7.3k/ $\mu$ L	4.5k/ $\mu$ L
<b>Hgb</b>	17 g/dL	14.7 g/dL	15,6 g/dL	14.7 g/dL	14.0 g/dL	12.2 g/dL
<b>PLTs</b>	307 k/ $\mu$ L	334 k/ $\mu$ L	339 k/ $\mu$ L	275 k/ $\mu$ L	205 k/ $\mu$ L	180 k/ $\mu$ L
<b>Hct</b>	54.7 %	49.5%	50 %	49.5%	48%	38.3%
<b>MPN-related symptoms</b>	Itching, fatigue	Fatigue ++	Itching, Fatigue ++	Fatigue++	Itching++, Fatigue ++	-



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# **GRAZIE!**