## **ELECTRONIC SUPPLEMENTARY MATERIAL**

## Annals of Hematology

Tracing the decision-making process for myelofibrosis: Diagnosis, stratification and management of ruxolitinib therapy in real-word practice

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## Supplementary Table 1. Questionnaire and related answers

	Questions	Possible answers	Percentages
1.	Where do you practice your clinical	a. University	43.75%
	activity?	b. Public hospital	56.25%
		c. Private practice	0%
2.	How many years of experience do you	a. <5	12.5%
	have in the treatment of chronic	b. 5-10	31.25%
	myeloproliferative syndromes?	c. >10	56.25%
3.	How many newly diagnosed myelofibrosis	a. <5	0%
	(MF) patients do you see per year?	b. 5-10	43.75%
		c. >10	56.25%
4.	You have experience in treating patients	a. Only MF	0%
	with:	b. MF and PV	0%
		c. MF and ET	0%
		d. MF, PV, ET	100%
5.	Do you use the IPSS score at baseline?	a. Yes	100%
		b. No	0%
6.	Do you use the MIPSS or MIPSS-70 score	a. Yes	25%
	at baseline?	b. No	75%
7.	Do you use the DIPSS score during	a. Yes	93.75%
	treatment?	b. No	6.25%
8.	Do you use the MYSEC-PM score for	a. Yes	81.25%
	secondary MF?	b. No	18.75%
9.	When do you decide to re-evaluate with	a. In the case of development/progression of	25%
	bone marrow biopsy a patient with PV in	splenomegaly	
	case of MF suspected evolution?	b. In the case of systemic symptoms not present before	ore 0%
		c. If phlebotomies are no longer needed	6.25%
		d. When at least two of the previous criteria are met	
10.	When do you decide to re-evaluate with	a. In the case of development of splenomegaly	18.75%
	bone marrow biopsy a patient with ET in	b. In the case of systemic symptoms not present before	ore 0%
	case of MF suspected evolution?	c. If cytoreduction is no longer needed	12.5%
		d. When at least two of the previous criteria are met	68.75%
11.	How often do you regularly visit a patient	a. Once a month	0%
	with IPSS low / intermediate-1 risk?	b. Once every 3 months	62.5%
		c. Once every 6 months	37.5%

		d.	Once a year	0%
12.	How often do you regularly visit a patient	a.	Once a month	50%
	with IPSS intermediate-2/high risk?	b.	Once every 3 months	43.75%
	(excluding transfusion-only access)	c.	Once every 6 months	6.25%
		d.	Once a year	0%
13.	When do you treat a patient with low IPSS	a.	Only in case of IPSS int-2 progression	6.25%
	risk?	b.	If splenomegaly is > 5 cm	31.25%
		c.	If symptoms are present	37.5%
		d.	I do not treat patients with IPPS low risk but I only	25%
			observe them	
14.	When do you treat a patient with	a.	If splenomegaly is > 5 cm	6.25%
	intermediate-1 IPSS risk?	b.	If symptomatic (TSS > 40 or very high single item	0%
			score)	
		c.	a+b	93.75%
		d.	I do not treat patients with IPSS int-1 risk	0%
15.	Do you have the opportunity to test for	a.	Yes	37.5%
	non-driver mutation in your center?	b.	No	62.5%
16.	Do you test non-driver mutation (i.e.	a.	No	25%
	ASXL1, etc) in your patients?	b.	Yes, always also for patients positive for driver	12.5%
	, , <u>, , , , , , , , , , , , , , , , , </u>		mutations	
		c.	Yes, only for young patients (< 50 years)	25%
		d.	Yes, but only for intermediate-1 risk patients to	37.5%
			decide the best therapeutic transplant strategy	
17.	Do you perform cytogenetic analysis in	a.	Yes, routinely for all	81.25%
- / •	MF patients?	b.	Never	0%
	m patients.	c.	Only for patients <50 years	18.75%
18	How do you routinely evaluate	a.	Manually	56.25%
10.	splenomegaly?	а. b.	With ultrasound	43.75%
	spicifolinegary:	c.	With abdominal CT/NMR	0%
19	For an int-2/high risk patient, do you use	a.	No, only after hydroxyurea	12.5%
1).	first-line ruxolitinib immediately?	а. b.	Yes, immediately	87,5%
20	Before starting ruxolitinib, in view of the	a.	Yes, but only for HBsAg	12.5%
20.	possible reactivation of hepatotropic	а. b.	Yes, I check for full panel and also HBV-DNA	87.5%
	viruses, do you check for hepatitis	c.	I do not perform for all patients but only in patients	0%
	serology?	C.	with previous positivity	070
21	In case of seropositive patient for hepatitis	a.	Yes	87.5%
۷1.	B, do you perform a prophylaxis?	а. b.	No	12.5%
22	Do you screen for previous TBC infection		Yes, always	93.75%
22.	before to start with ruxolitinib?	a. b.	No, never	0%
	before to start with ruxontimo?			6.25%
22	For TDC took what do you mostly you?	c.	Yes, only in case of a past medical history	
23.	For TBC test, what do you mostly use?	a.	QuantiFERON-TB Gold	81.25%
		b.	Mantoux intradermal test	12.5%
2.4	D. C	c.	CT scan	6.25%
24.	Before starting treatment with ruxolitinib,	a.	Never	50%
	do you perform viral serology for herpetic	b.	Yes, always	37.5%
2.5	viruses (CMV, HSV1-2, EBV, VZV)?	c.	Only for some frequent viruses	12.5%
25.	Do you perform antiviral prophylaxis	a.	No, never	6,25%
	during ruxolitinib treatment?	b.	Yes, always	0%
		c.	Yes, but only in patients with a history of previous	18.75%
		_	infections	
		d.	Yes, but only in secondary prophylaxis after one or	75%
			more episodes of herpes zoster reactivation during	
			treatment	
26.	How do you behave in case of infection	a.	Reduce temporarily the dose	62.5%
	during ruxolitinib?	b.	$\mathcal{E}$	37.5%
			dose after the resolution of the event	
		c.	Definitely discontinue the drug	0%
		d.	Discontinue the drug and the resume	0%
27.	How do you schedule the monitoring visits	a.	Once a week for the first 2-3 months	12.5%
	starting with ruxolitinib?	b.	Every 15 days	18.75%
	-	c.	Once a month	25%
		d.	Initially weekly and then depends on hematologic	43.75%
			toxicity	
				•
28.	In case of hematologic toxicity (grade 3	a.	Discontinue the treatment	0%
28.	In case of hematologic toxicity (grade 3 anemia):	a. b.	Discontinue the treatment Reduce the dosage	0% 18.75%

	<ul> <li>Transfuse the patient (even naïve) and continue with the same dosage</li> </ul>	81.25%
29. Do you use erythropoietin during	a. Never	12.5%
treatment with ruxolitinib in case of	b. Yes, always	25%
anemia?	c. Yes, but only in patients with serum EPO dosage	62.5%
	lower than then normal	
30. In case of hematological toxicity (grad-	a. Discontinue the treatment	18.75%
thrombocytopenia):	b. Reduce the dosage	81.25%
	c. Transfuse the patient (even naïve) and continue with the same dosage	0%
31. In case of toxicity and need for	a. Gradually reduce and then discontinue	87.5%
interruption, how do you carry out the suspension?	b. Discontinue the drug abruptly	12.5%
32. After how long on average patients rep	ort a. 1-2 weeks from the start of treatment	62.5%
a resolution / improvement in their symptoms?	b. 3-4 weeks from the start of treatment	37.5%
33. Do you use the MPN10 instrument to	a. Yes	75%
quantify symptoms?	b. No	25%
34. Based on your experience, how many	a. 30-40%	25%
patients with low / intermediate-1 risk	are b. Less than 30%	62.5%
symptomatic?	c. More than 50%	12.5%
35. How do you rate a splenic response to ruxolitinib?	<ul> <li>a. Consider the COMFORT studies criteria (&gt; 35% reduction in basal volume)</li> </ul>	25%
	b. Consider a reduction > 50% of the spleen length	31.25%
	c. According to 2013 IWG criteria	43.75%
36. Do you consider a dose reduction in the		25%
long-term responder?	b. Yes, always	31.25%
	c. Yes, but only if the patient has concomitant toxicity	43.75%
37. When do you consider a patient in therapeutic failure after treatment with	a. In case of absolute lack of splenic response and symptoms	18.75%
ruxolitinib?	b. Worsening of general conditions (increase in symptoms and splenomegaly) during treatment	43.75%
	c. There are no failure criteria and therefore I continue the treatment even if the patient has responded only from a symptomatic point of view	37.5%
38. Do you consider re-evaluating bone bio		50%
during treatment?	b. No	18.75%
	c. Yes, after at least one year of treatment	31.25%
39. Do you consider possible pharmacolog		81.25%
associations in patients with non-optimeresponse?		18.75%
40. What is the drug you most frequently	a. Erythropoietin	25%
associate with ruxolitinib?	b. Hydroxyurea	43.75%
	c. Anabolic	6.25%
	d. Vitamins	25%
41. Do you perform a transplant assessmer	t in a. At baseline, if IPSS Intermediate-2 /High	93.75%
patients under the age of 70 (more than	b. At progression, if DIPSS Intermediate-2 / High	56.25%
one answer is possible)?	c. If IPSS / DIPSS Intermediate-1 in the presence of	81.25%
	factors that negatively impact the prognosis  d. Only if the patient does not reach or lose a response to ongoing treatments and in the absence of	25%
	therapeutic alternatives	