

New Insights in Autoimmune Liver Disease

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Agenda

- Autoimmune hepatitis
 - Novelties in diagnosis and treatment
 - Treatment in patients with decompensated cirrhosis
 - Immunosuppression withdrawal
- Primary biliary cholangitis
 - Evaluation of treatment response
 - New treatment options

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Current Diagnosis of AIH

Simplified Criteria			
	0	1	2
ANA, SMA	Negative	1:40 ; 1:80	>1:80
IgG	Normal	> Normal	>1.5 ULN
Histology	-	Compatible	Typical
Viral hepatitis	Yes		No

▪ 6 points: Probable
▪ >7 points: Definite

None of these are pathognomonic of AIH
Requires the exclusion of other causes of liver disease

Floreani et al. Aliment Pharmacol Ther 2006,
Vergani et al. J Hepatol 2004, Hennes et al. Hepatology 2008

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Liver Biopsy Interpretation

✓ Typical: interface hepatitis, lymphoplasmacytic infiltrate, emperipoleisis, rosette.
✓ Compatible: chronic hepatitis without the typical features.

Concomitant liver diseases?
Acute presentation?

Chronic Hepatitis				
Portal inflammation	X	X	X	
Interface hepatitis	X	X	X	
Lobular inflammation		X	X	
Plasma cell groups	X	X	X	
+ signs of other disease	POSSIBLE	PROBABLE	PROBABLE	POSSIBLE
	POSSIBLE	POSSIBLE	POSSIBLE	UNLIKELY

Hennes et al. Hepatology 2008, Lohse et al. Liver Int 2022

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Liver Biopsy Interpretation

✓ Typical: interface hepatitis, lymphoplasmacytic infiltrate, emperipoleisis, rosette.
✓ Compatible: chronic hepatitis without the typical features.

Concomitant liver diseases?
Acute presentation?

Acute Hepatitis

Lobular hepatitis
Central perivenulitis
Plasma cell group
± portal inflammation

Hennes et al. Hepatology 2008, Lohse et al. Liver Int 2022

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Definition of Treatment Response

ALT and IgG normalization:
Histological Δ AI < 4

Complete biochemical response
Remission
Insufficient response
Non-response

Lack of ALT and IgG normalization after 6 months of starting IS.

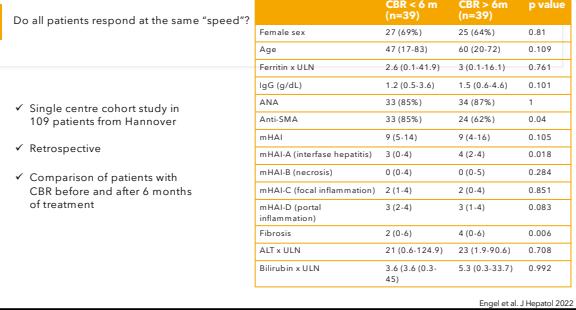
Less than 50% reduction in ALT after 4 weeks of treatment.

Normalization at 6 months

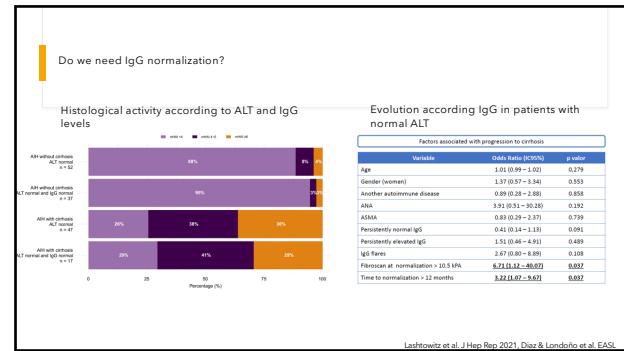
Pape et al. J Hepatol 2022, Slooter et al. Hepatology 2024

Survival (%) over time showing complete biochemical response (CBR) and no CRR.

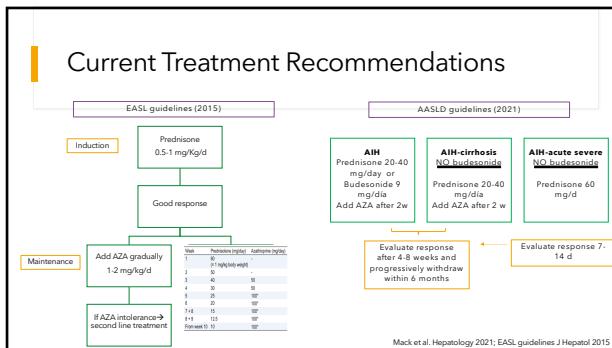
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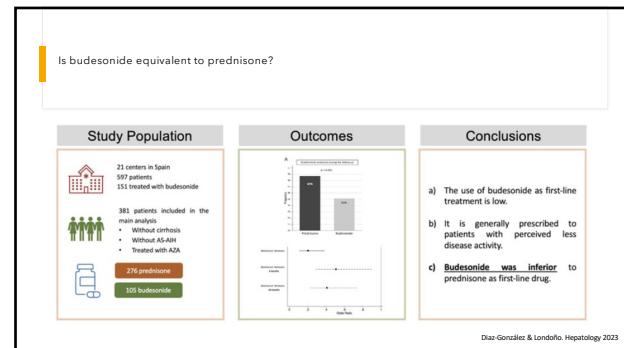
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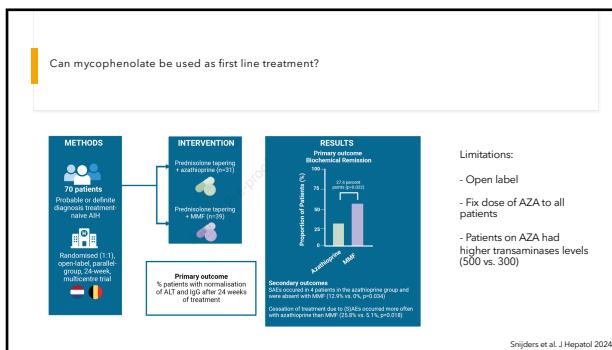
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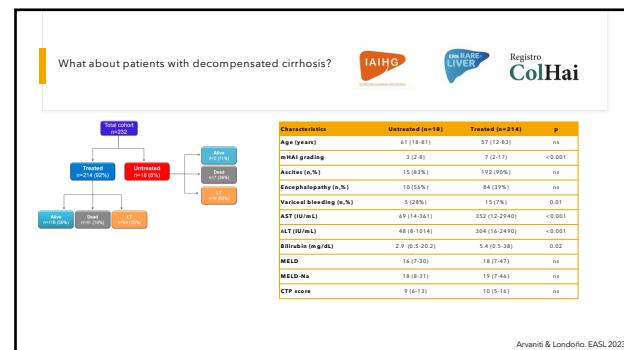
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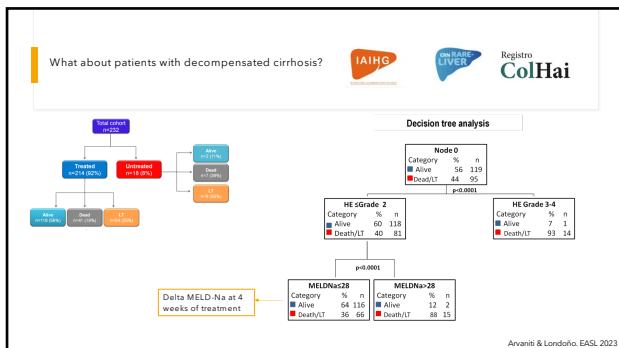
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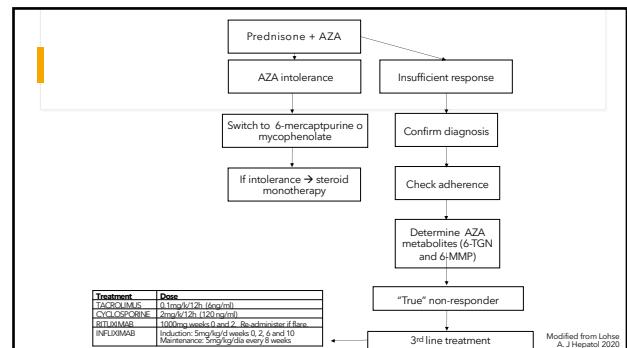
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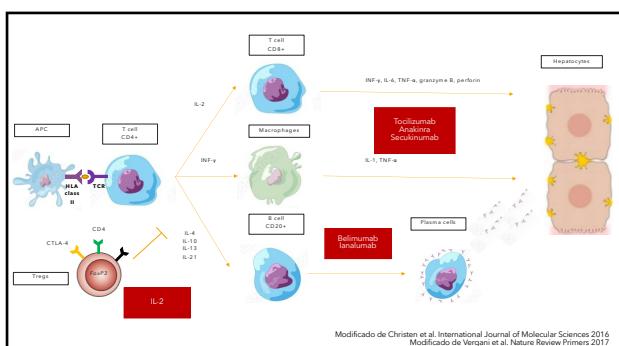
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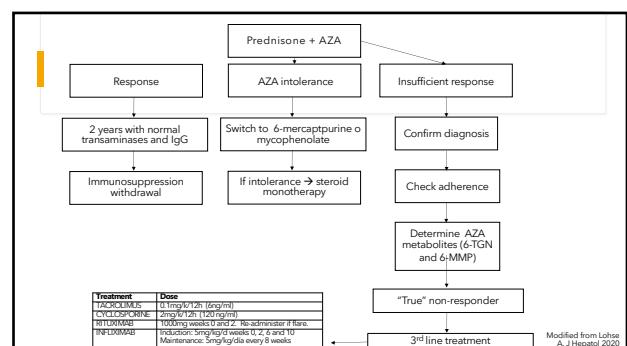
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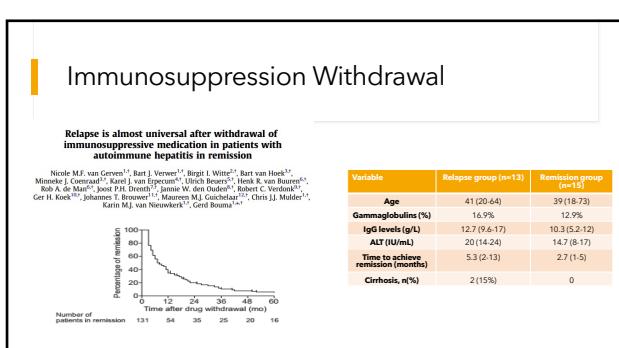
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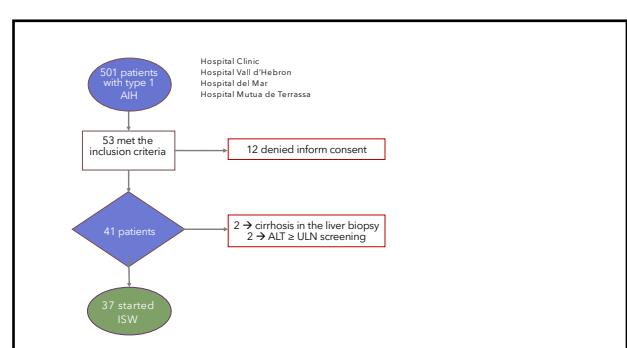
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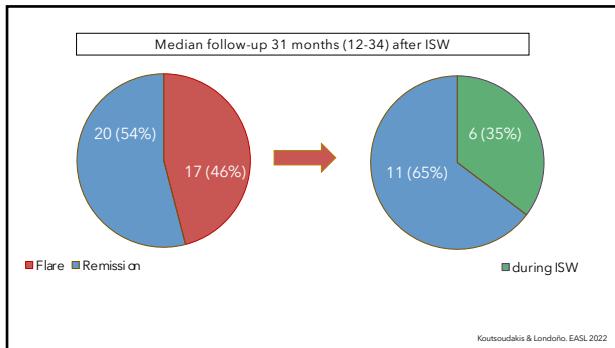
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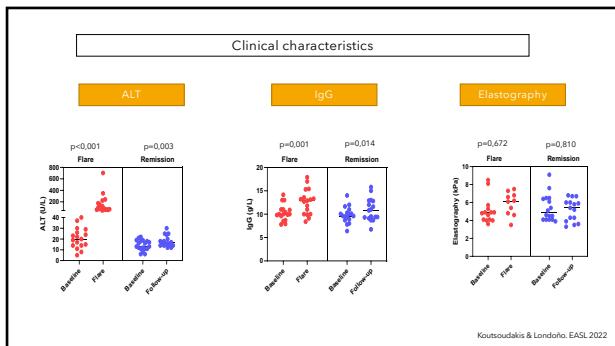
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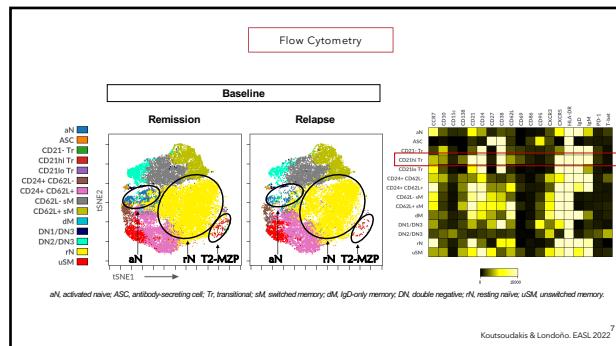
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Clinical characteristics			
	Flare (n=17)	Remission (n=21)	P
Age (years)	53 (42-75)	65 (22-80)	0.165
Female sex (n, %)	8 (50%)	11 (52%)	0.884
ALT (U/L)	20 (5-34)	14 (6-23)	0.021
IgG (g/L)	10 (8-14)	10 (6-14)	0.403
Time on remission (years)	4 (3-10)	3.5 (3-11)	0.728
nHAI	1 (0-3)	1 (0-2)	0.423
Transient elastography (kPa)	4.9 (3.6-8.5)	4.6 (3.9-9.1)	0.773
Immunosuppression(n, %)			
AZA	12 (75%)	15 (72%)	
Corticoids	1 (6%)	4 (19%)	
AzA + Corticoids	3 (19%)	2 (9%)	
Presentation (n, %)			
Chronic	10 (59%)	13 (65%)	
Acute	5 (29%)	7 (35%)	
Acute-severe	2 (12%)	0	
Median (range)			

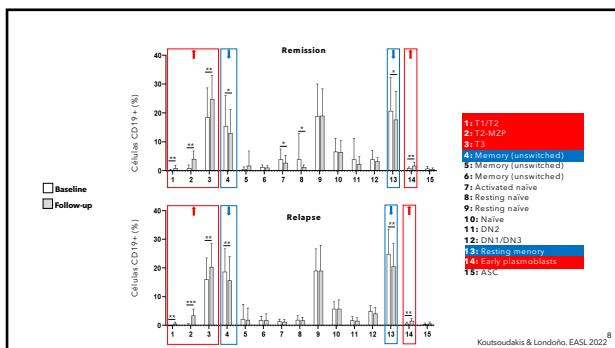
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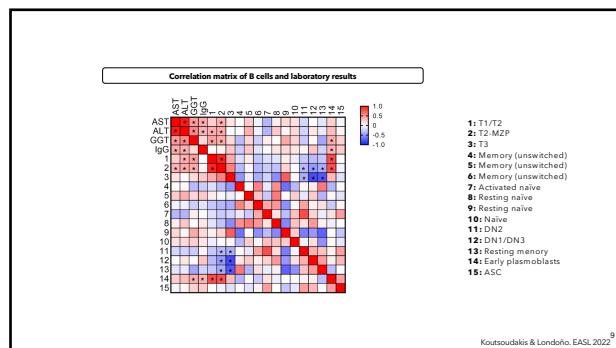
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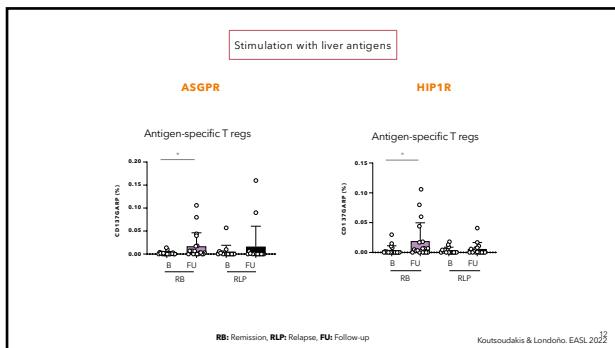
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Take home messages (1)

- Liver biopsy is essential for the diagnosis of AIH but should be interpreted by expert pathologist.
- IgG normalization might not be necessary for defining biochemical response.
- Mycophenolate could be a good alternative to azathioprine in selected patients.
- The presence of hepatic encephalopathy and MELD-Na score at AIH diagnosis serve to guide treatment decisions in patients with decompensated cirrhosis.
- There is a need for better second or third line treatment options in AIH.

van Gerven et al. J Hepatol 2013; Hartl et al. J Hepa 2015

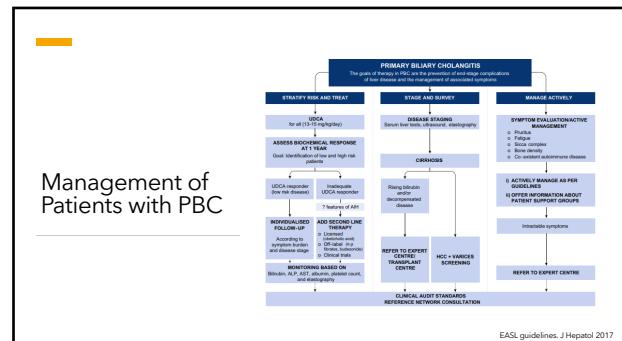
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Management of Patients with PBC



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Evaluation of Treatment Response

Criteria	Time	ALP	AST	Bilirubin	Albumin
Rochester-I	6m	>2 x ULN			
Rochester-II	12m	>2 x ULN		>1mg/dL	
Barcelona	12m	>ULN ↓ < 40%			
Paris-I	12m	>3 x ULN	>2 x ULN	>1 mg/dL	
Paris-II	12m	>1.5 x ULN	>1.5xULN	>1mg/dL	
Rotterdam	12m			>ULN	<LLN
Toronto	24m	>1.67 x ULN			

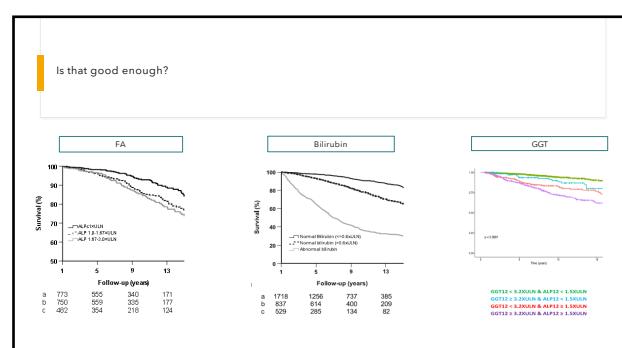
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UK-PBC
ALP, bilirubin, BL albumin, BL platelet count, BL age

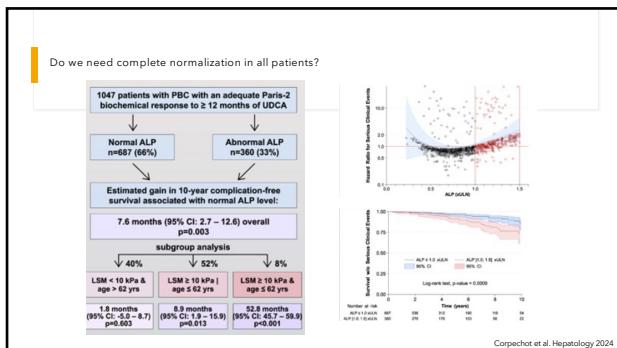
ALP, bilirubin, BL albumin, BL platelet count, AST (or ALT)

Parés A, et al. Gastroenterology. 2006; Corpechot C, et al. Hepatology. 2008; Kuijper EMM, et al. Gastroenterology. 2009; Kurogi T, et al. Am J Gastroenterol. 2010; Angulo P, et al. Liver. 1999; Monish N, et al. Liver Int. 2012; Corpechot C, et al. J Hepatol. 2011; Zhang LN, et al. Hepatology. 2013; EASL Clinical Practice Guidelines. J Hepatol. 2017

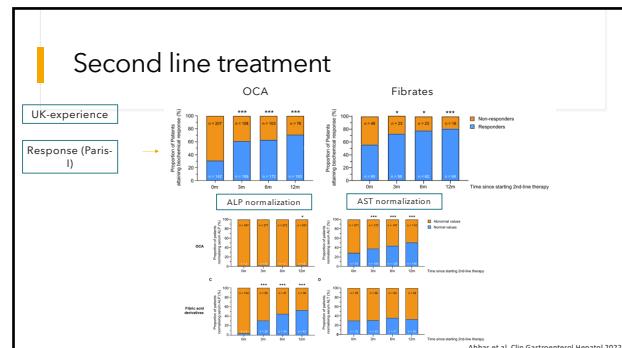
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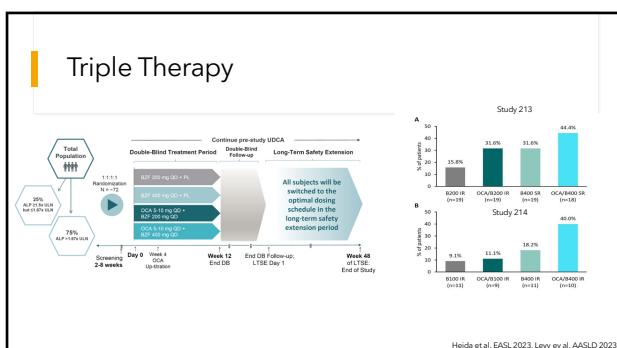
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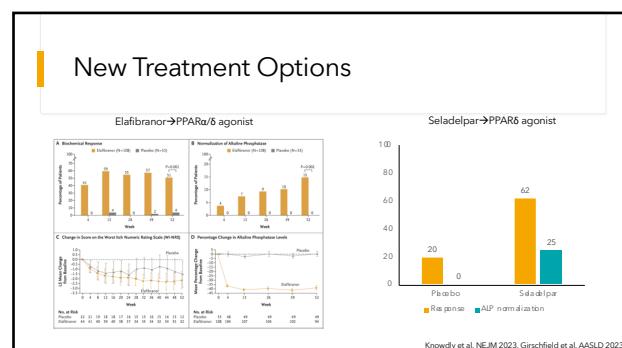
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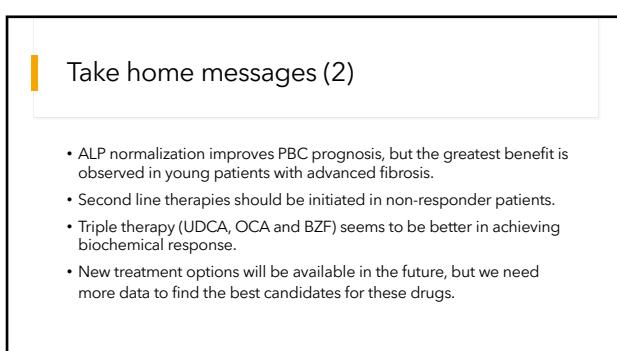
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