



BRIGHAM AND
WOMEN'S HOSPITAL

| The Lung Center |

LAM in TSC Surveillance, Treatment and Research

Souheil El-Chemaly, MD, MPH

*Associate Professor of Medicine
Harvard Medical School*



HARVARD MEDICAL SCHOOL
TEACHING HOSPITAL

2021 E-Webinar Series

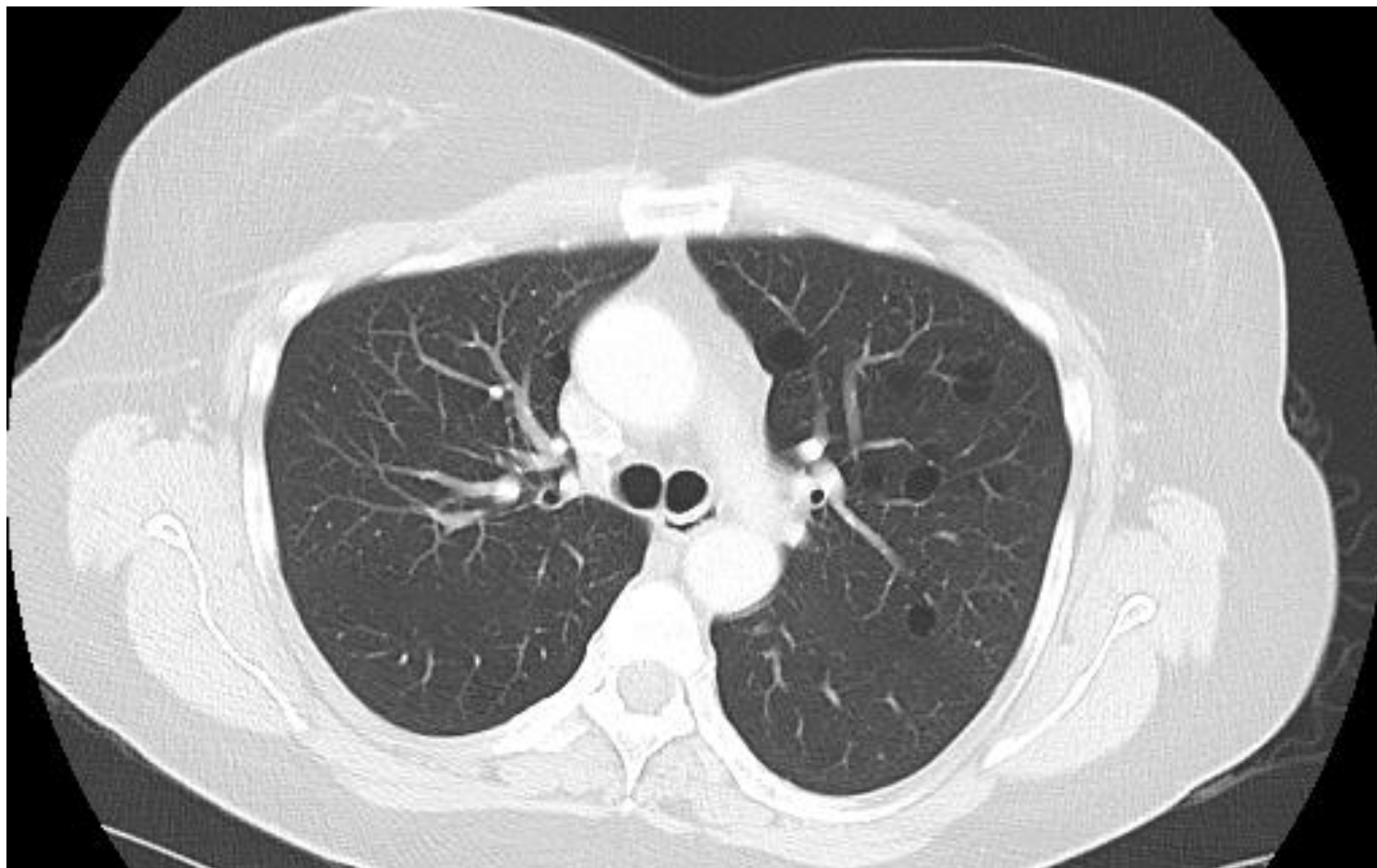
6/24/2021

No Conflict of Interest

What is LAM?

- LAM is a cystic lung disease

Cysts/ CT scan



Diagnosis of LAM

- Cystic lung disease
AND any of the following
 - TSC
 - AML (angiomyolipoma)
 - Milky effusions
 - VEGF-D > 800pg/ml

What are PFT?

ATS compliant tests are indicated by a ✓ : FVC ✓ FRC DLCO ✓ VTG ✓								
Spirometry (BTPS)		Predicted Range		Pre Bronchodilator		Post Bronchodilator		Percent Change
		Mean	95%	Actual	% Pred	Actual	% Pred	
FVC Effort Time		----	----	15:13	----	15:38	----	--
FEV ₁	L	2.81	2.20	2.80	100	2.86	102	2
FVC	L	3.50	2.75	3.64	104	3.66	105	1
FEV ₁ / FVC	%	81	70	77	95	78	96	1
FEV ₆	L	3.51	2.83	3.54	101	3.60	103	2
FEV ₁ / FEV ₆	%	83	74	79	95	79	95	0
FEF ₂₅₋₇₅	L/s	2.81	1.61	1.90	68	2.58	92	36
PEFR	L/s	6.83	5.10	7.16	105	7.87	115	10
FET	sec	----	----	8.03	----	7.29	----	-9
MVV	L/m	98.7	47.7	----	----	----	----	--
Lung Volumes (Box)		Predicted Range		Pre Bronchodilator				
		Mean	95%	Actual	% Pred			
VTG Effort Time		----	----	15:18	----			
TLC	L	5.10	4.02	7.33	144			
FRC	L	2.82	1.76	4.84	172			
IC	L	2.28	----	2.49	109			
ERV	L	1.07	----	1.17	109			
RV	L	1.75	0.97	3.67	210			
RV/TLC	%	34	26	50	147			
VC	L	3.36	2.68	3.66	109			
Diffusion		Predicted Range		Pre Bronchodilator				
Hb	Drawn	Mean	95%	Actual	% Pred			
DLCO Effort Time		----	----	15:23	----			
DLCO	mL/min/mmHg	22.95	16.45	23.01	100			
DLCO [Hb]	mL/min/mmHg	22.95	16.45	23.01	100			
Hb	g/dl	13.4	12.0	----	----			
VA [BTPS]	L	5.06	4.98	5.21	103			
DLCO/VA	mL/min/mmHg/L	5.10	3.78	4.41	86			

2 distinct scenarios

- TSC Diagnosed First- *In search of LAM*
- LAM Diagnosed First- *In search of TSC*

In Search of LAM

- Typical presentation no pulmonary symptoms

When should you get a chest CT scan?

Best way to follow? How often?

Is there a need for repeat CT scan?

What if cannot do PFT?

In Search of TSC

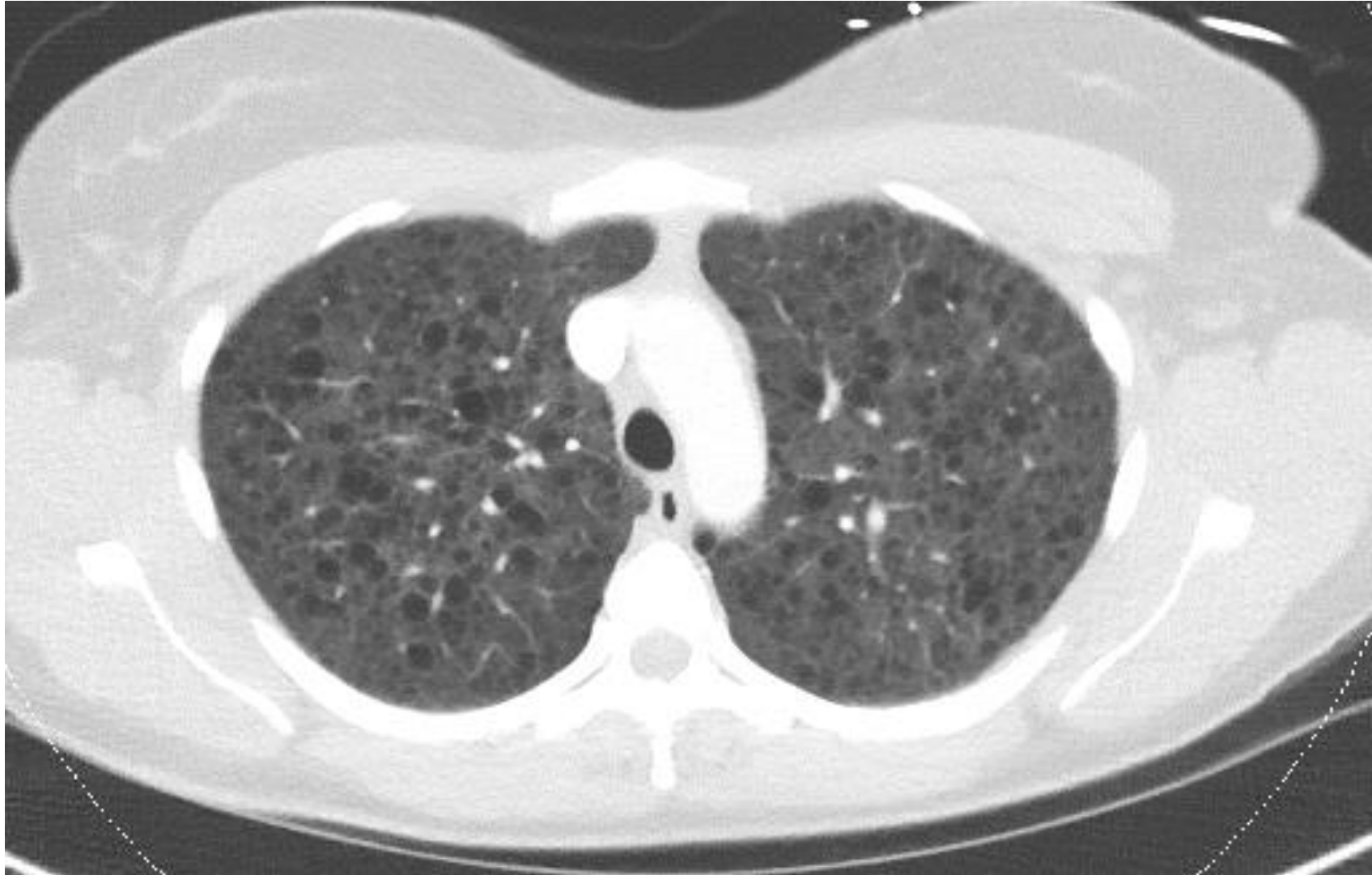
- Abdominal imaging (CT scan or MRI).

Bilateral AML high index of suspicion

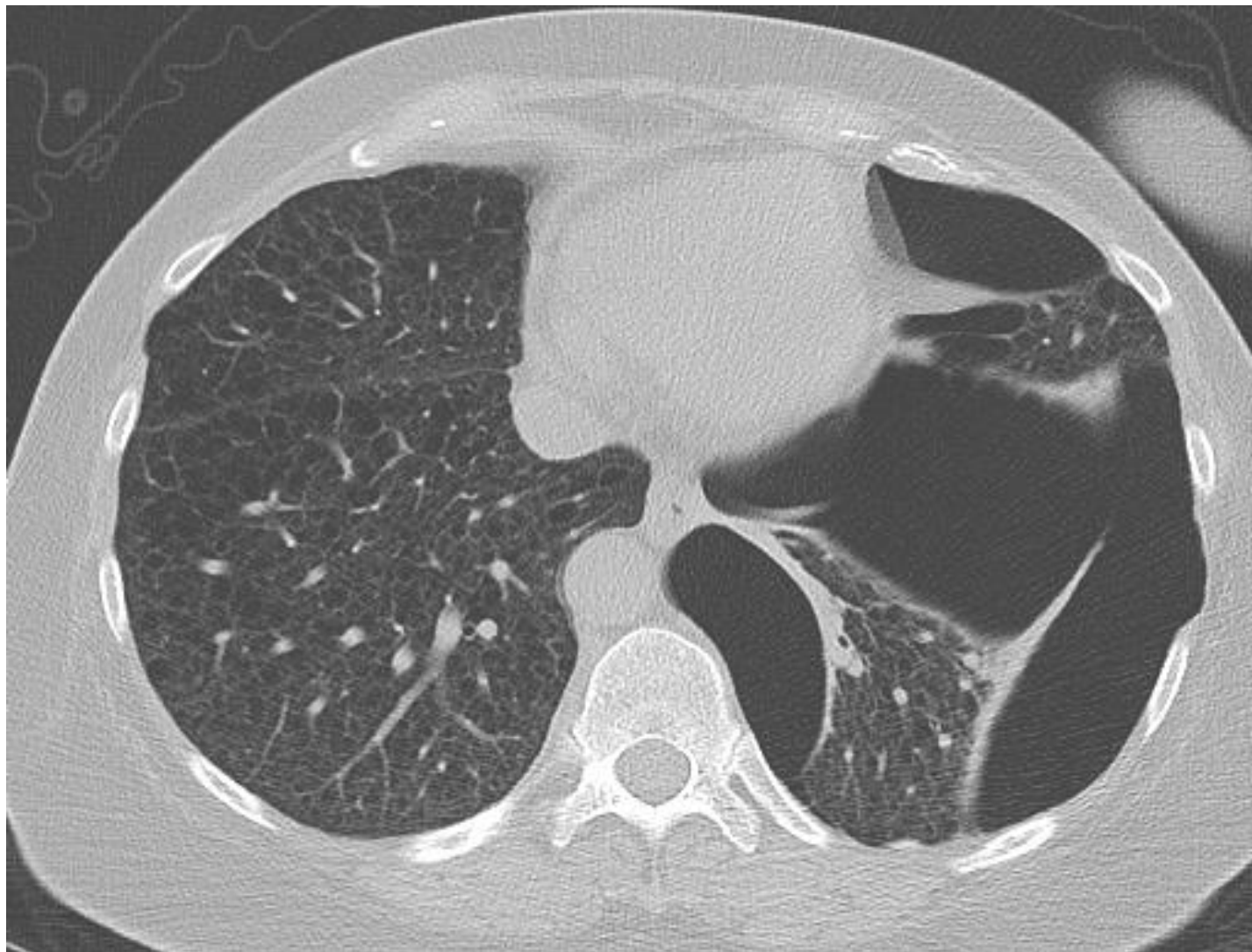
- Skin lesions
- Brain imaging
- VEGF-D levels?

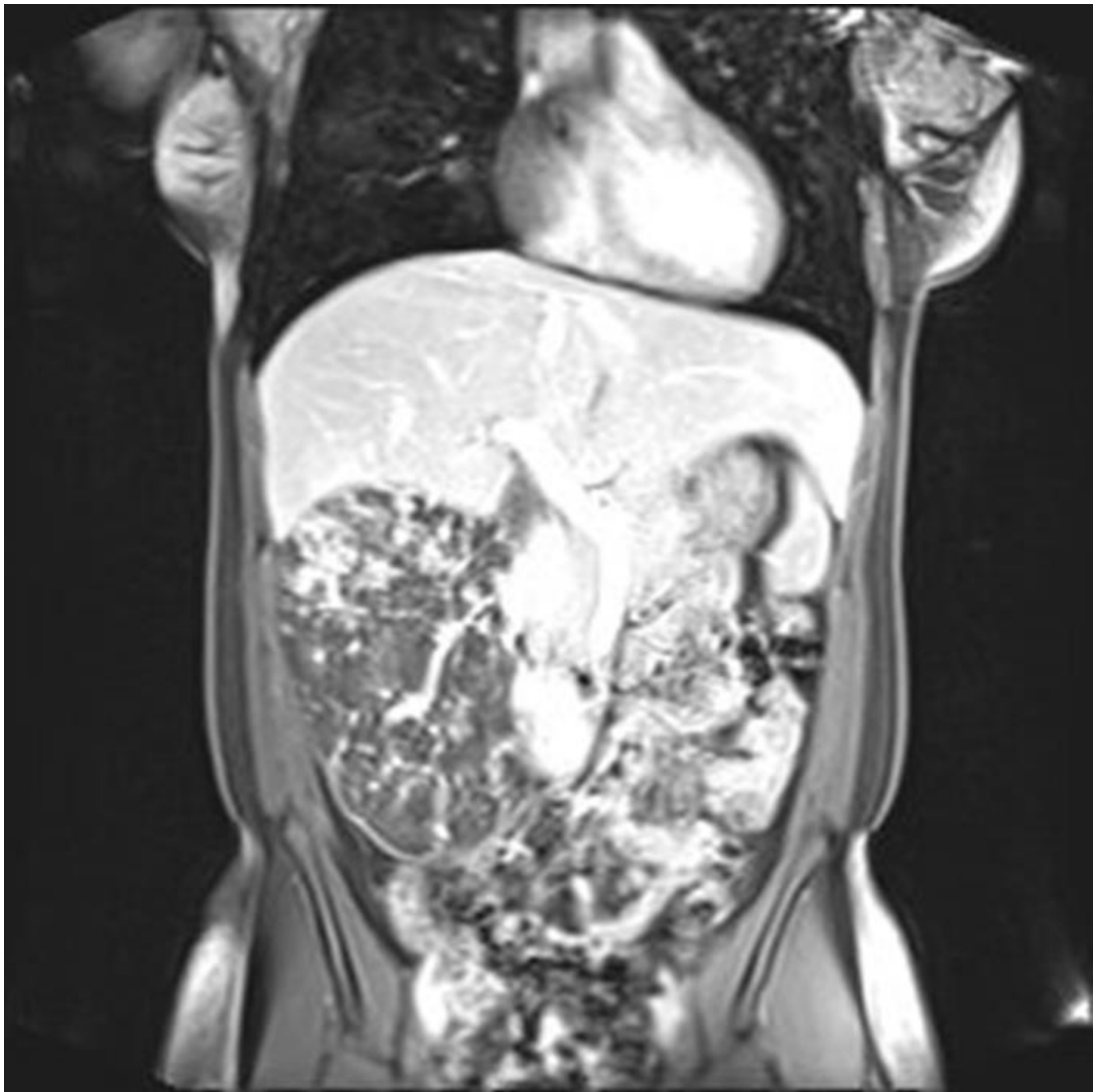
In Search of TSC

Typical presentation: Shortness of breath or chest pain



Pneumothorax



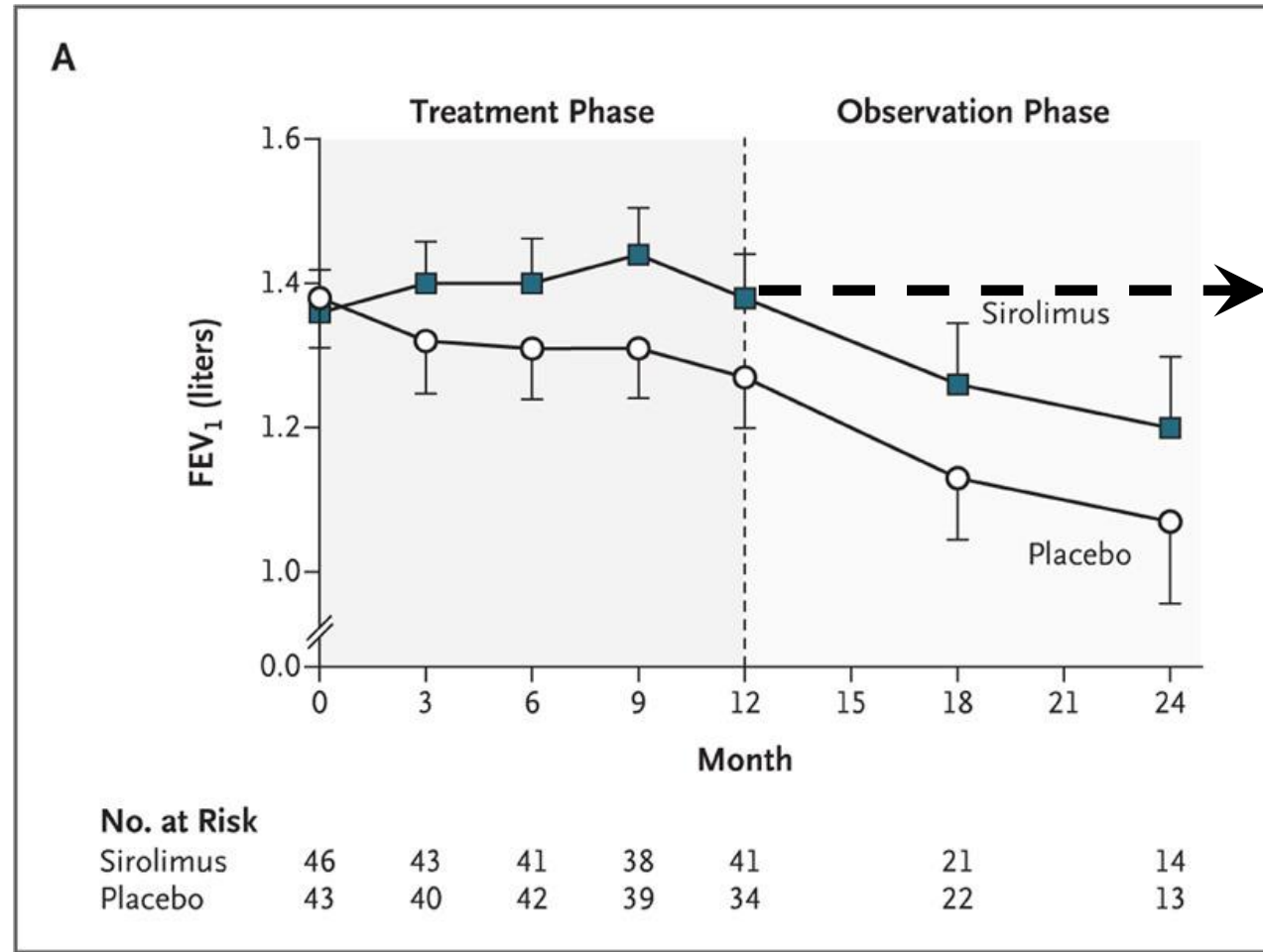


Careful skin examination



Brain imaging and careful history taking

Rapamycin works



Clinical trials in LAM

- MILED Multicenter Interventional LAM Early Disease Trial
- A Study of Nintedanib for LAM
- RESULT Resveratrol and Sirolimus in LAM Trial

Questions?