CAIRNS HOSPITAL

ALYSSA TIEPPO &
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Hamman Rich Syndrome

Case Study

58 yo male from Daintree

Previously well, presented with progressive SOB over 1 week

Associated with fever, chills, malaise, lethargy

Not on regular medication; smoked tobacco and marijuana

Worked as a firefighter previously

o/e: SpO2 90% RA, RR 26, BP 140/85, temp 36.4

Chest: fine crepitation over right lower zone

Laboratory

FBC: Hb 115, neutrophil 14.9, Plt 254

Na 126 (normal 135-145mmol/L, K 3.5 (normal: 3.5-5mmol/L), Cr 66 (normal: 65.4-119.3um/L)

LFT & coag profile within normal limits

VBG: pH 7.43, pCO2 28, pO2 54, HCO3 18

Sputum culture NAD

Impression

- Diffused alveolitis with broad differentials
- Treated with IV ceftriaxone, azithromycin
- Supplemental O2
- Microbiological screening
- Autoimmune screening
- Bronchoscopy & washing

Clinically improving

FEELING MUCH IMPROVED

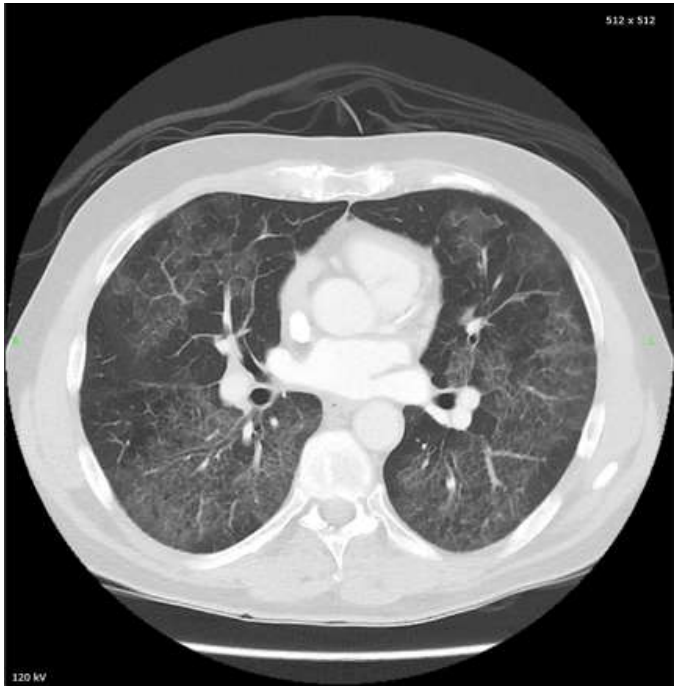
“BEST I’VE FELT IN YEARS”

HAPPY TO TRIAL OFF O2

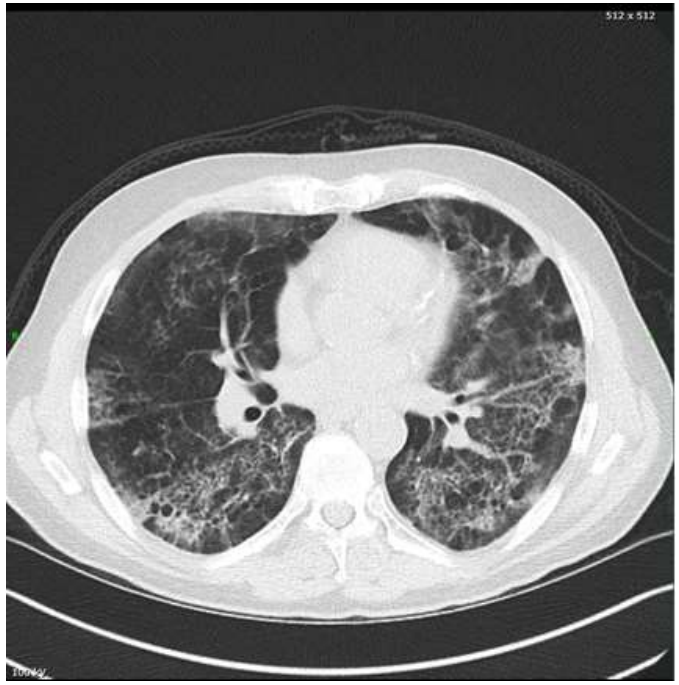
CLINICALLY IMPROVING ILD

ABS CEASED

Rapidly Progressive Fibrosis



Initial CT 07/02/2017



14/02/17

Spirometry

		Ref	Pre Meas	Pre % Ref
FEV1	Liters	3.44	3.36	98
FVC	Liters	4.34	4.39	101
FEV1/FVC	%	77	76	
FEF25-75%	L/sec	3.65	2.76	76
FEF50%	L/sec	4.57	3.66	80
PEF	L/sec	8.54	9.78	115

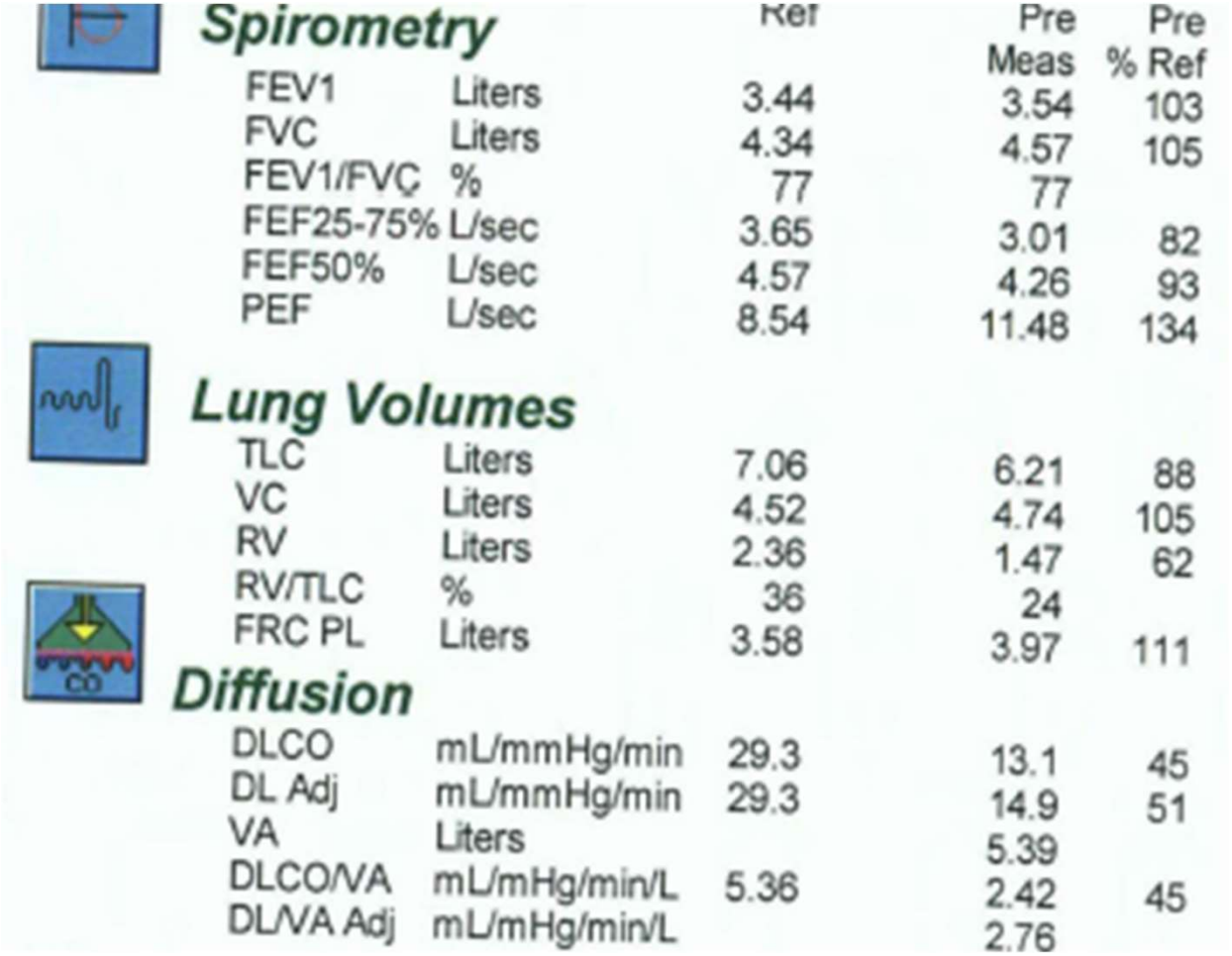
Lung Volumes

TLC	Liters	7.06	5.72	81
VC	Liters	4.52	4.39	97
RV	Liters	2.36	1.32	56
RV/TLC	%	36	23	
FRC PL	Liters	3.58	3.39	94

Diffusion

DLCO	mL/mmHg/min	29.3	10.2	35
DL Adj	mL/mmHg/min	29.3	11.9	40
VA	Liters		5.19	
DLCO/VA	mL/mHg/min/L	5.36	1.97	37
DL/VA Adj	mL/mHg/min/L		2.28	

Initial DLFTs 15/02/2017



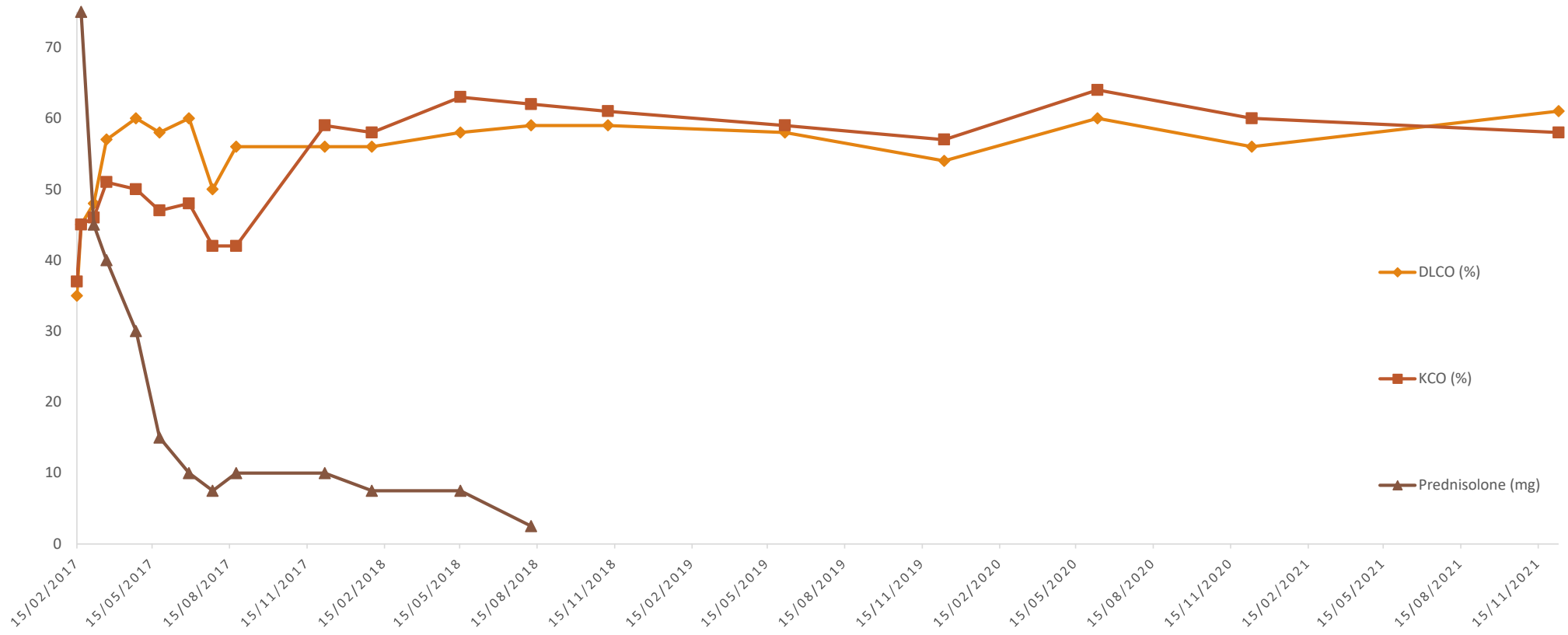
		Ref	Pre Meas	Pre % Ref
Spirometry				
FEV1	Liters	3.44	3.54	103
FVC	Liters	4.34	4.57	105
FEV1/FVC	%	77	77	
FEF25-75%	L/sec	3.65	3.01	82
FEF50%	L/sec	4.57	4.26	93
PEF	L/sec	8.54	11.48	134
Lung Volumes				
TLC	Liters	7.06	6.21	88
VC	Liters	4.52	4.74	105
RV	Liters	2.36	1.47	62
RV/TLC	%	36	24	
FRC PL	Liters	3.58	3.97	111
Diffusion				
DLCO	mL/mmHg/min	29.3	13.1	45
DL Adj	mL/mmHg/min	29.3	14.9	51
VA	Liters		5.39	
DLCO/VA	mL/mHg/min/L	5.36	2.42	45
DL/VA Adj	mL/mHg/min/L		2.76	

Sequential DLFTs 20/02/2017

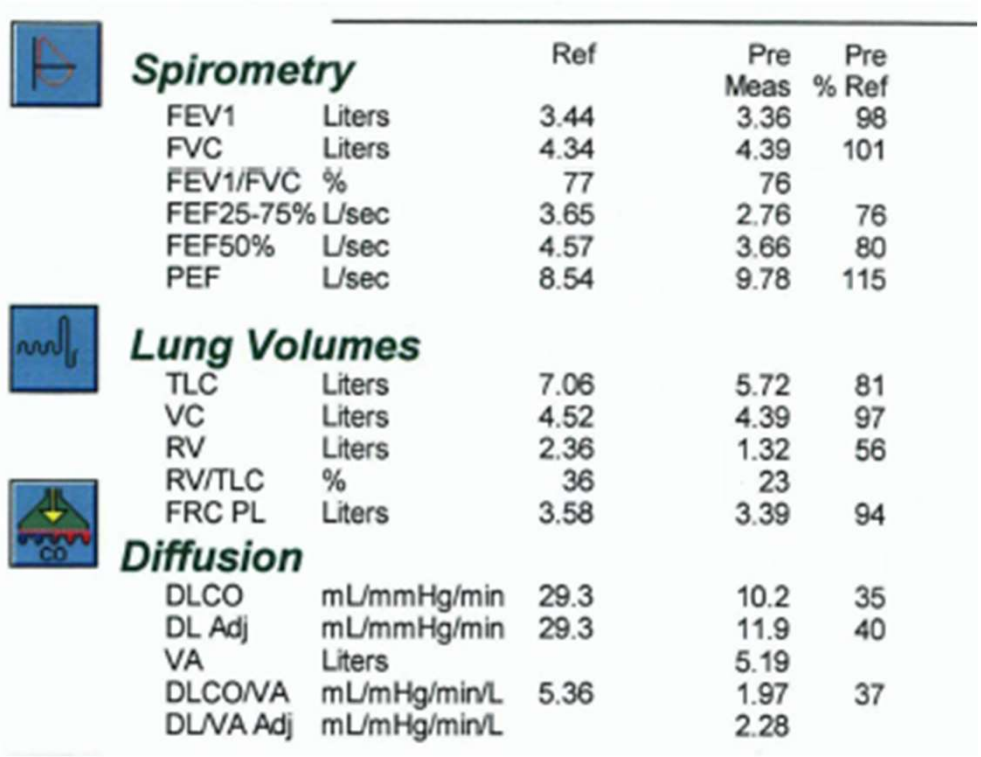
Discharged 24/02/17

- Clinically improving despite hypermania secondary to steroids
- Drop to 50mg prednisolone
- Plan for DLFTs every 2 weeks after discharge
- Repeat CXR before discharge and every 2 weeks after

DLCO & KCO Over Time



Final DLFTs – 09/12/2021



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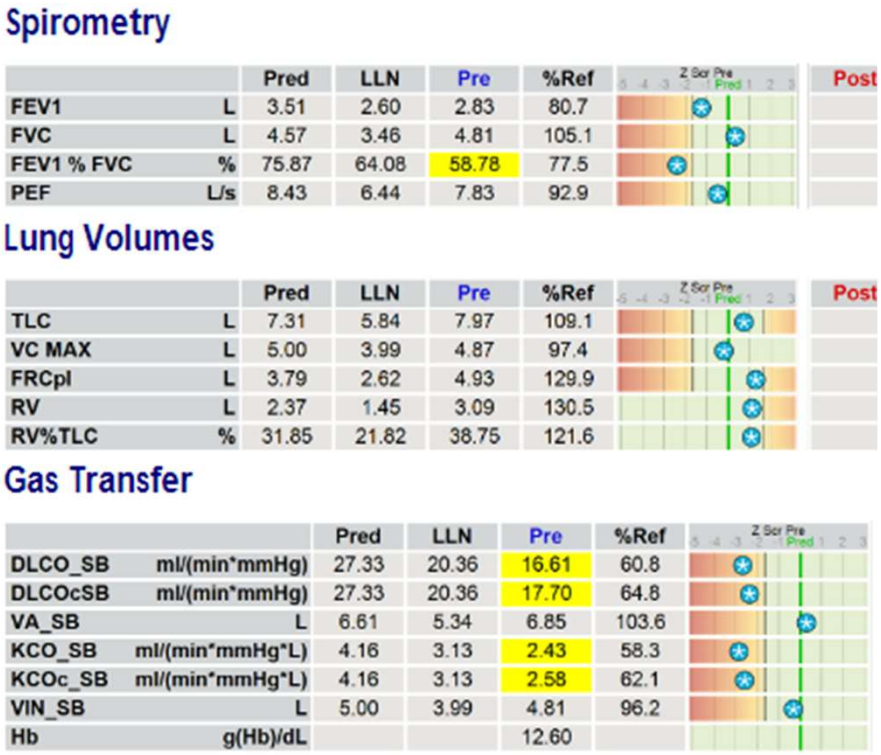
VA	Liters		5.19	
DLCO/VA	mL/mHg/min/L	5.36	1.97	37
DL/VA Adj	mL/mHg/min/L		2.28	

15/02/2017



Spirometry

		Pred	LLN	Pre	%Ref	Post
FEV1	L	3.51	2.60	2.83	80.7	
FVC	L	4.57	3.46	4.81	105.1	
FEV1 % FVC	%	75.87	64.08	58.78	77.5	
PEF	L/s	8.43	6.44	7.83	92.9	

Lung Volumes

		Pred	LLN	Pre	%Ref	Post
TLC	L	7.31	5.84	7.97	109.1	
VC MAX	L	5.00	3.99	4.87	97.4	
FRCpl	L	3.79	2.62	4.93	129.9	
RV	L	2.37	1.45	3.09	130.5	
RV%TLC	%	31.85	21.82	38.75	121.6	

Gas Transfer

		Pred	LLN	Pre	%Ref
DLCO_SB	ml/(min*mmHg)	27.33	20.36	16.61	60.8
DLCOcSB	ml/(min*mmHg)	27.33	20.36	17.70	64.8
VA_SB	L	6.61	5.34	6.85	103.6
KCO_SB	ml/(min*mmHg*L)	4.16	3.13	2.43	58.3
KCOc_SB	ml/(min*mmHg*L)	4.16	3.13	2.58	62.1
VIN_SB	L	5.00	3.99	4.81	96.2
Hb	g(Hb)/dL			12.60	

09/12/2021

Summary

Clinically and radiologically improved during admission

Discharged after 18 days admission on oral prednisone

Discharged from OPD December 2021 after remaining stable

Hamman Rich history

- Described by Louis Hamman and Arnold Rich in 1935
- Known as acute interstitial pneumonitis
- Acute onset and rapid progressive course

Epidemiology

Prevalence
1-9/100 000

Affects previously healthy individuals

Mean age 50-55 years

No gender predominance

Not associated with cigarette smoking

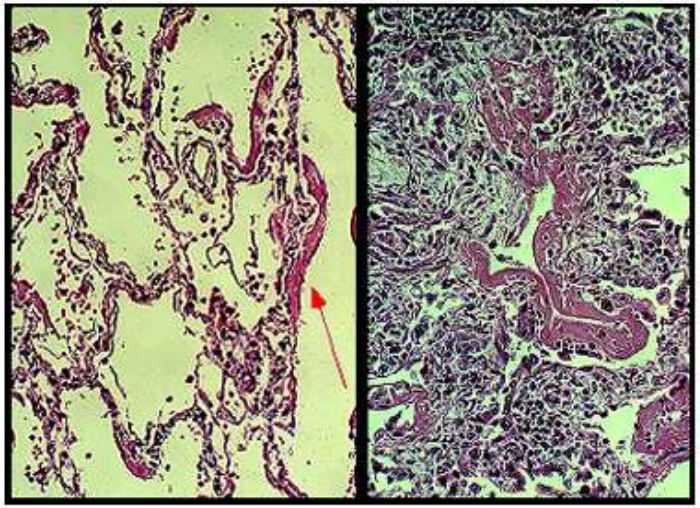
Clinical Presentation

- Rapid onset with myalgia, arthralgia, chills, malaise
- Fever, cough, progressive breathlessness
- Tachypnoea and diffused crackles on chest exam
- Hypoxemic respiratory failure requiring ventilatory support within a few days

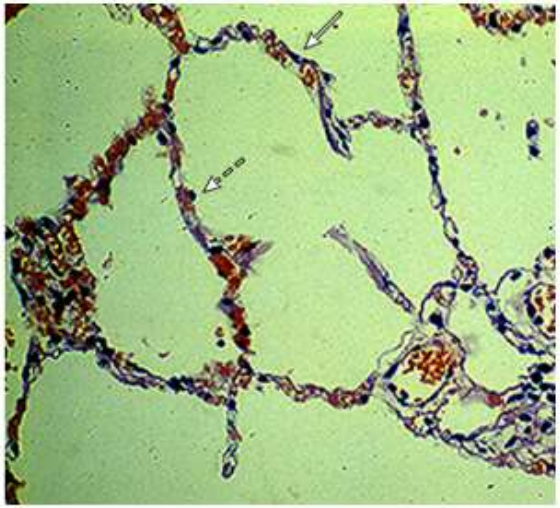
Broad Differentials for Diffuse Alveolitis

Ethchlorvynol
Aspirin
Radiation therapy
Oxygen toxicity
Heroin
Cocaine
Toxic inhalants
Chlorine gas
Nitrogen dioxide
Phosgene
Smoke
Ingestants
Paraquat
Kerosene
Rapeseed oil-toxic oil syndrome
Connective tissue disease

Lung Biopsy – Histology



AIP



Normal lung

Prognosis

- In-hospital mortality > 50%
- Most initial survivors die within 6 months of presentation
- Recurrence has been reported

Management

- Consider Hamman Rich early and treat aggressively
- Respiratory support
- Glucocorticosteroids
- Consider empirical antibiotics
- Consider Lung transplantation

Questions?