RESPIRATORY CASES

Airways and ILD

Donny Wong Respiratory Physician

AIRWAYS 1, 33 YO M

1 year of nocturnal dry cough with ?wheeze now nightly symptoms

Hx of Hay fever, multiple allergies & childhood Asthma

Never smoker and no Fhx of atopy

Clinic exam and spirometry normal, sniffing in clinic

What to do next?

AIRWAYS 1, INITIAL TESTS

Steroclear trial

Ix,

- CXR normal and CT sinus normal
- Formal lung functions

Baseline Spirometric Data

		Base	LLN	Pred	%Pred	Z Scr -5 -4 -3 -2 -1 Pred 1 2 3
FVC	L	4.80	3.34	4.19	114.3	
FEV 1	L	3.53	2.82	3.58	98.8	
FEV 1 % FVC	%	74	76	85	86.4	
MFEF 75/25	L/s	2.74	2.58	4.00	68.5	
PEF	L/s	8.98	7.11	9.10	98.6	

- Negative hypertonic saline challenge tests, while FeNO 124 (>50 elevated)

AIRWAYS 1, MORE TESTING

Steroclear some benefit

So for induced sputum analysis

		Pre	LLN	Pred	% Pred	Z-Score -5 -4 -3 -2 -1 Pred 1 2 3	Post	% Change
Spirometry								
FVC	**L**	4.76	3.34	4.19	114		4.95	4
FEV1	**L**	3.70	2.82	3.57	104		3.92	6
FEV1/FVC	**%**	78	76	85	91		79	2
MMEF	**L/s**	3.13	2.58	3.99	78		3.40	8
PEF	**L/s**	8.83	7.11	9.10	97		8.80	-0

- Post BD normal, and sputum eosinophils 13% (>3%)

Trial of Qvar 50 microgram 2 puffs bd

AIRWAYS 1, LATEST FU

Spirometry normal, FeNO 26

Bloods, Eosinophils 0.6, IgE 770

SPT +ve, HDM, cat, dog, Alternaria, Aspergillus, Perennial Rye,

Continued Qvar dose

EOSINOPHILIC BRONCHITIS

Possibly common

Diagnosis can be pragmatically treated with ICS

Otherwise through FeNO, Induced sputum analysis or airway biopsy

Spectrum of airways allergic disease

Long term outcome unclear.

AIRWAYS 2, 61 YO M

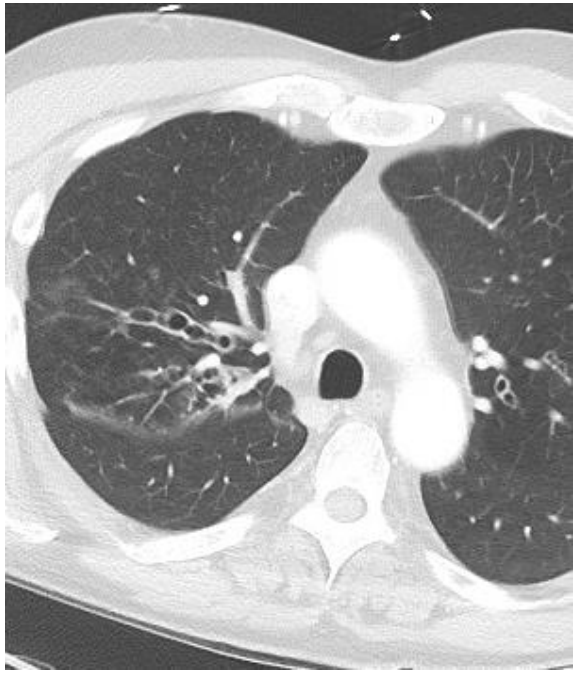
Long history of asthma, with MVA incidental CT picked up RUL varicose bronchiectasis 1/12 earlier with RML collapse

Hx of recurrent infections yearly, though not required antibiotics and possible sinus infections

Spirometry FEV1/FVC 3.14/4.32 (87/92%), ratio 73%,

On Symbicort 200/6 2 puff a day

What to do next?



AIRWAYS 2, TESTS

CT sinus excluded sinusitis

Labwork, IgE >2500, eosinophils 1.4, aspergillus RAST 4+, aspergillus precipitins ++++, normal immunoglobulins, anti-CCP and RF negative.

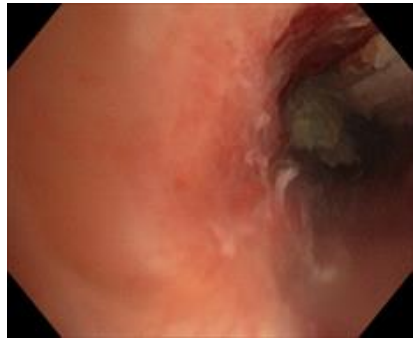
CXR PA and lateral still RML atelectasis

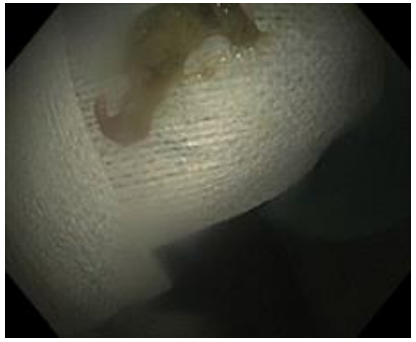
formal lung functions: FEV1/FVC of 3.07/4.61 (87/101%), ratio 67%, post BD non-significant, FeNO 27/28

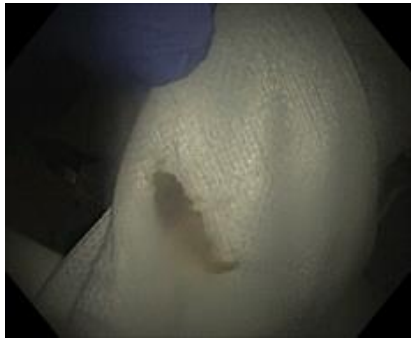
Went ahead for bronchoscopy

- Grown aspergillus

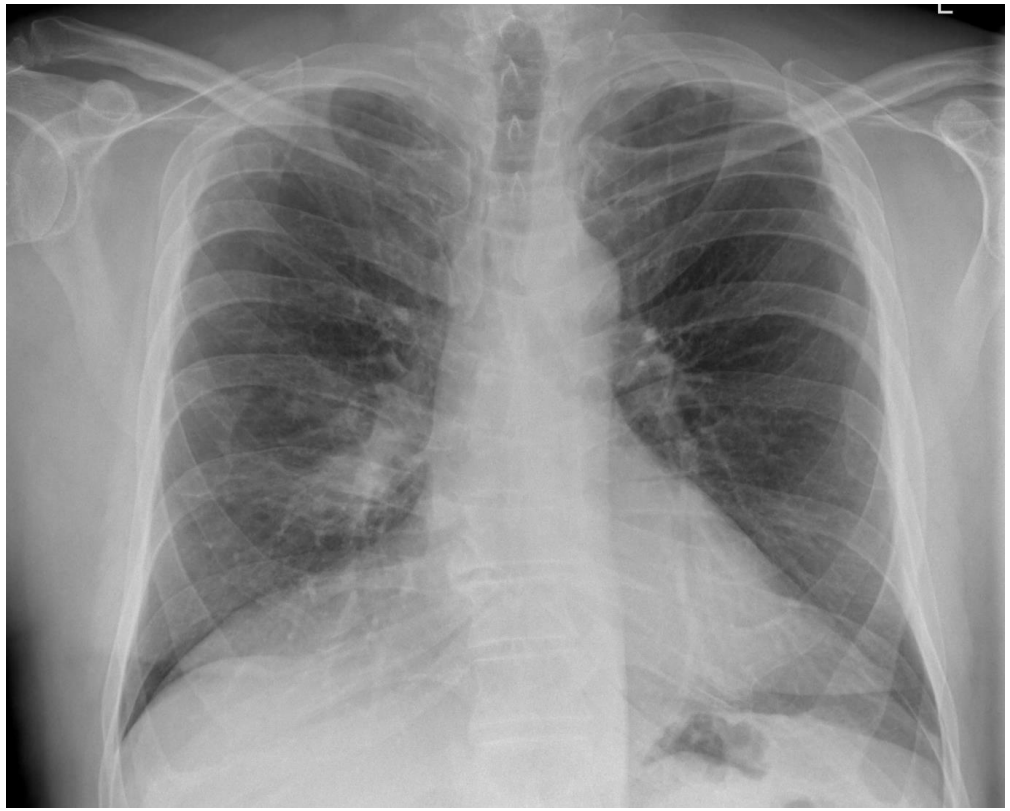
Treat with prolonged steroids

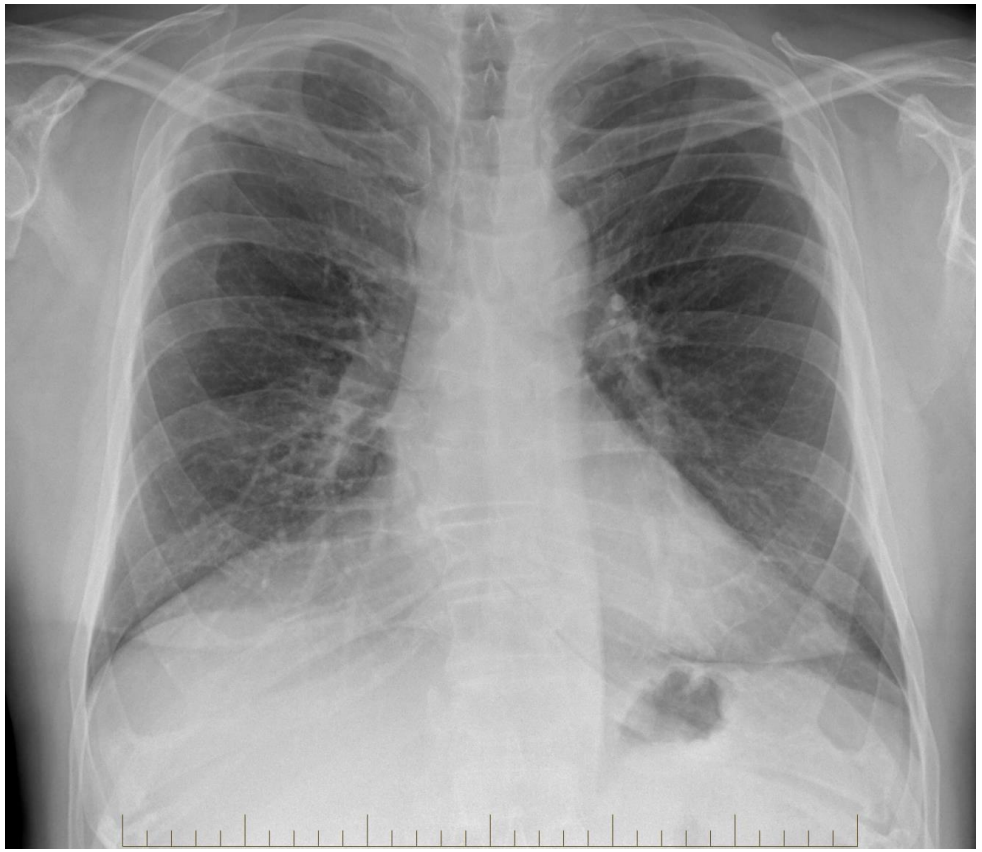






PRE AND POST BRONCHOSCOPY





ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS (ABPA)

Extreme form of asthma specific to aspergillus, can be to any fungi trigger in theory

Hx of recurrent difficult asthma, mucus plugs brief relief with steroids

Require asthma diagnosis, IgE >1000, high blood eosinophils, Aspergillus RAST/SPT and precipitins positive and ideally growth of aspergillus

Long course of steroids for flares and monitor with CBC and IgE.

AIRWAYS 3, 39 YO F

Poor control of asthma recently post viral infection, summer well

Hay fever, eczema and sinus issues in past, atopy in family

On high dose ICS/LABA and multiple steroids thus increased further ICS

Other history, depression and OSA (obesity BMI 50).

Lung functions, FeNO normal, IgE 94, no eosinophilia

		Pre	LLN	Pred	% Pred	Z-Score -5 -4 -3 -2 -1 Pred 1 2 3	Post	% Change
Spirometry								
FVC	**L**	1.66	2.24	2.78	60		1.80	9
FEV1	**L**	1.21	1.86	2.34	52		1.40	15
FEV1/FVC	**%**	73	74	84	87		78	6
MMEF	**L/s**	0.88	1.64	2.69	33		1.18	33
PEF	**L/s**	4.07	4.41	5.89	69		4.75	17

What now?

AIRWAYS 3

Severe asthma with obesity with repeated courses of steroids

Options oral steroids, ?montelukast, ?other triggers

Considered for biologics in NZ criteria as per pharmac

- Omalizumab- Anti-IgE
- Mepolizumab, Benralizumab – Anti IL-5

ILD 1, 70 YO M

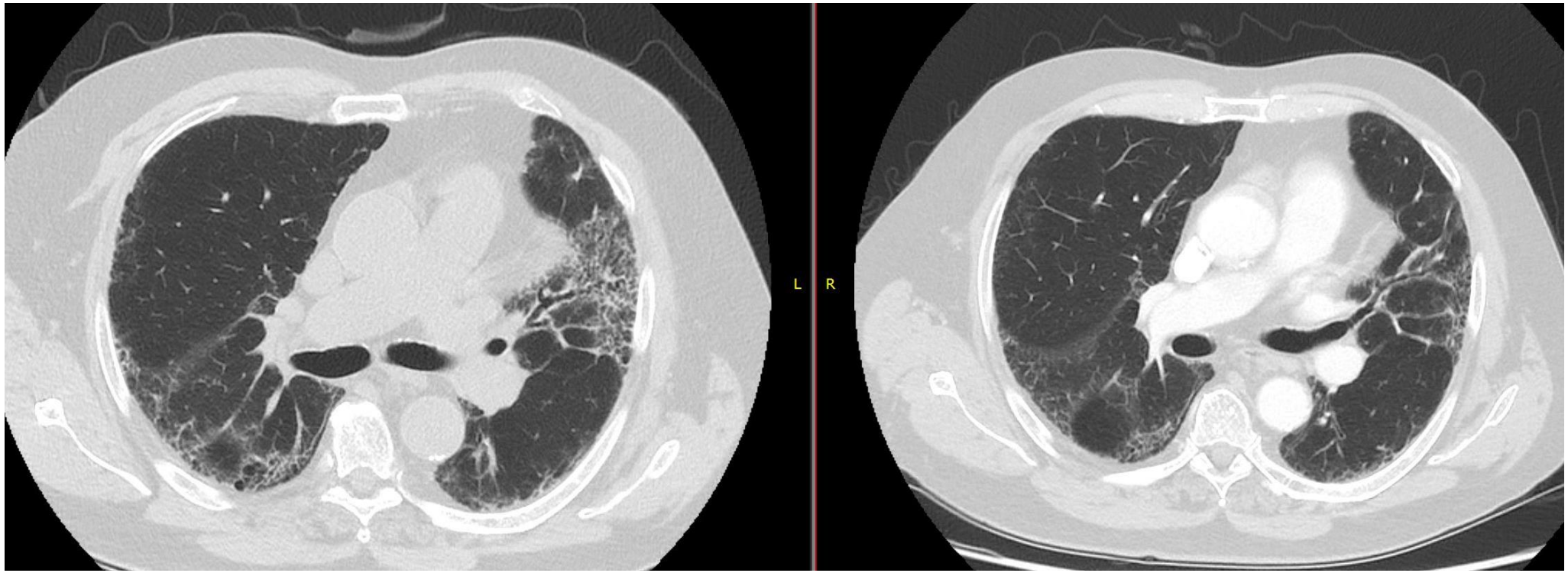
2 years earlier CT for weight loss with nodule completed follow up

Somewhat significant asbestos exposure with previous 30 pack year smoking

LFTs over 2 years

- FEV1/FVC of 2.38/2.99 (85/83% predicted), TLC of 4.15 (68% predicted) and DLCO of 4.18 (56% predicted)
- FEV1/FVC of 2.06/2.60 (76/73% predicted), TLC of 4.02 (66% predicted), DLCO of 3.85 (52% predicted)

ILD 1, CT OVER 2 YEARS



What's the diagnosis here?

ILD 1, IPF MANAGEMENT

Discussed at ILD meeting for consensus

Diagnosed as idiopathic pulmonary fibrosis (IPF) as UIP pattern on CT ie, traction bronchiectasis, honeycombing and lung volume loss

In NZ management only two antifibrotics agents require ILD consensus and FVC criteria

- Nintedanib and Pirfenidone, regular liver functions for both, diarrhoea and nausea/sunsentivity respectively,

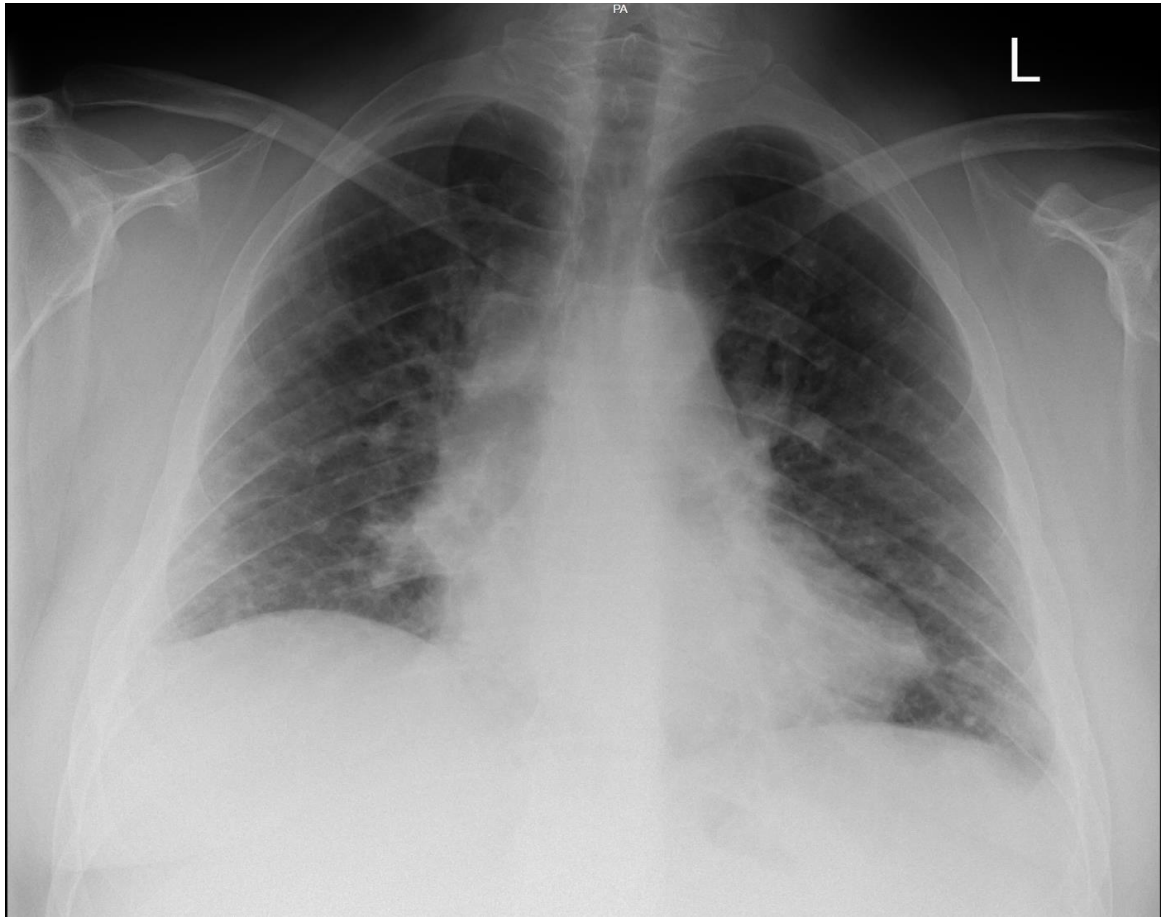
*Likely at some point for all progressive fibrosis ILD role for antifibrotic

ILD 2, 37 YO M

Presents with immigration medical after 3 x AFB sputa and repeat CXR already done

Fit and well asymptomatic, BMI 39, minimal ex smoker and no pulmonary insults and no FHx of autoimmune disease

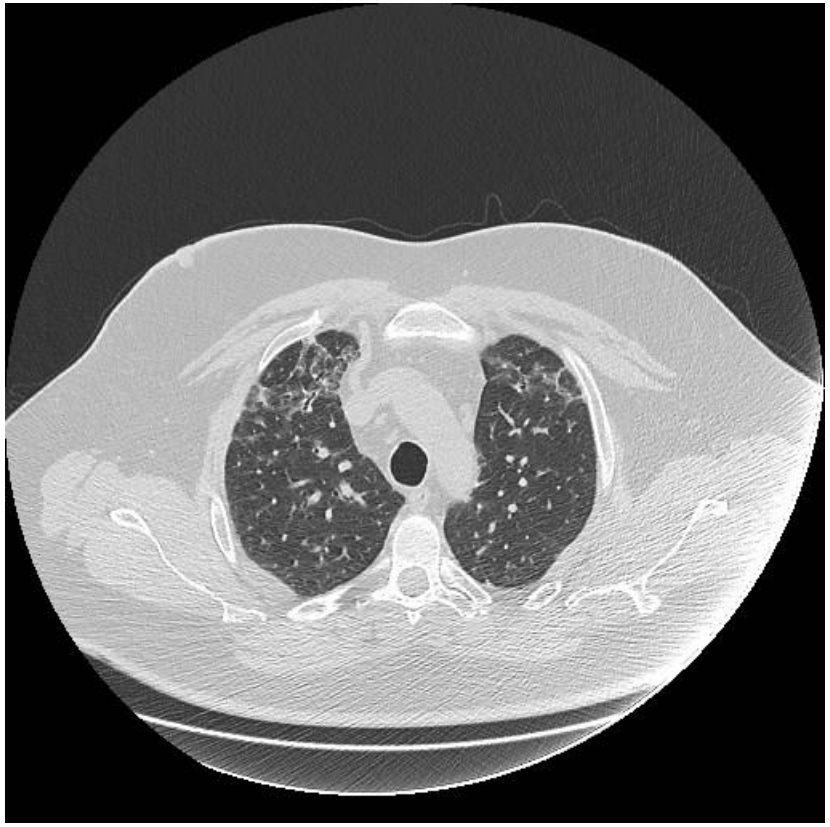
What is the CXR concerning for?



ILD 2, TESTS

Bloods, ANA 1280, Ku antibody Positive

		Pre	LLN	Pred	% Pred	Z-Score -5 -4 -3 -2 -1 Pred 1 2 3
Spirometry						
FVC	L	2.55	4.23	5.23	49	
FEV1	L	2.34	3.37	4.26	55	
FEV1/FVC	%	92	72	82	112	
MMEF	L/s	3.93	2.49	4.21	93	
PEF	L/s	9.90	7.80	9.80	101	
Lung Volumes						
TLC	L	3.25	6.39	7.54	43	
IC	L	1.73	3.93	3.93	44	
FRCpl	L	1.51	2.54	3.53	43	
ERV	L	1.09	1.54	1.54	71	
RV	L	0.42	1.31	1.98	21	
RV%TLC	%	13.04	19.41	28.39	46	
VC	L	2.82	4.56	5.48	52	
sR tot	kPa*s	1.00	1.18	1.18	85	
Gas Transfers						
DLCO_SB	ml/(min*mmHg)	16.45	26.20	33.51	49	
DLCOcSB	ml/(min*mmHg)	16.45	26.20	33.51	49	
VA_SB	L	3.23	5.86	7.16	45	
VIN_SB	L	2.60	4.56	5.48	48	
KCO_SB	ml/(min*mmHg*L)	5.09	3.73	4.71	108	
KCOc_SB	ml/(min*mmHg*L)	5.09	3.73	4.71	108	



GENERAL PRINCIPLES ILD

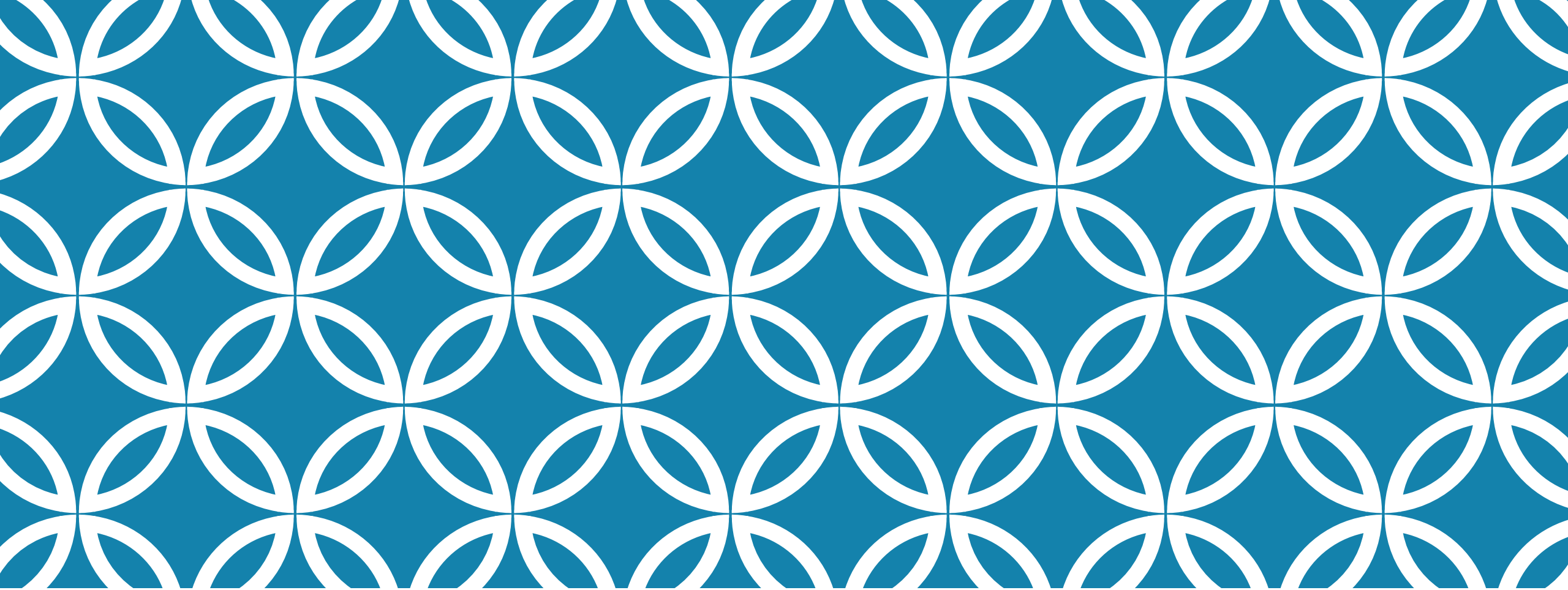
Rare disease in some respect.

Fine crackles ➔ CXR

Think of connective tissue disease and occupation or exposure history

Bloods can consider work up, ANA, ENA, ANCA, RF, Anti-CCP, myositis antibodies and CK.

Depends on driver treatment antifibrotic agents and/or immunosuppression (which can be quite aggressive)



QUESTIONS?

Thanks for your time