

Overview of DILD Cases (n=187)

CATEGORY

Diffuse Developmental Disorders

Growth Abnormalities

Pulmonary Interstitial Glycogenolysis

NEHI

Surfactant Dysfunction

Disorders of the Normal Host

Related to Systemic Disease Processes

Disorders of the Immunocompromised Host

Disorders Masquerading as ILD

Unclassified

23

12%

5

3%

28

15%

9

5%

20

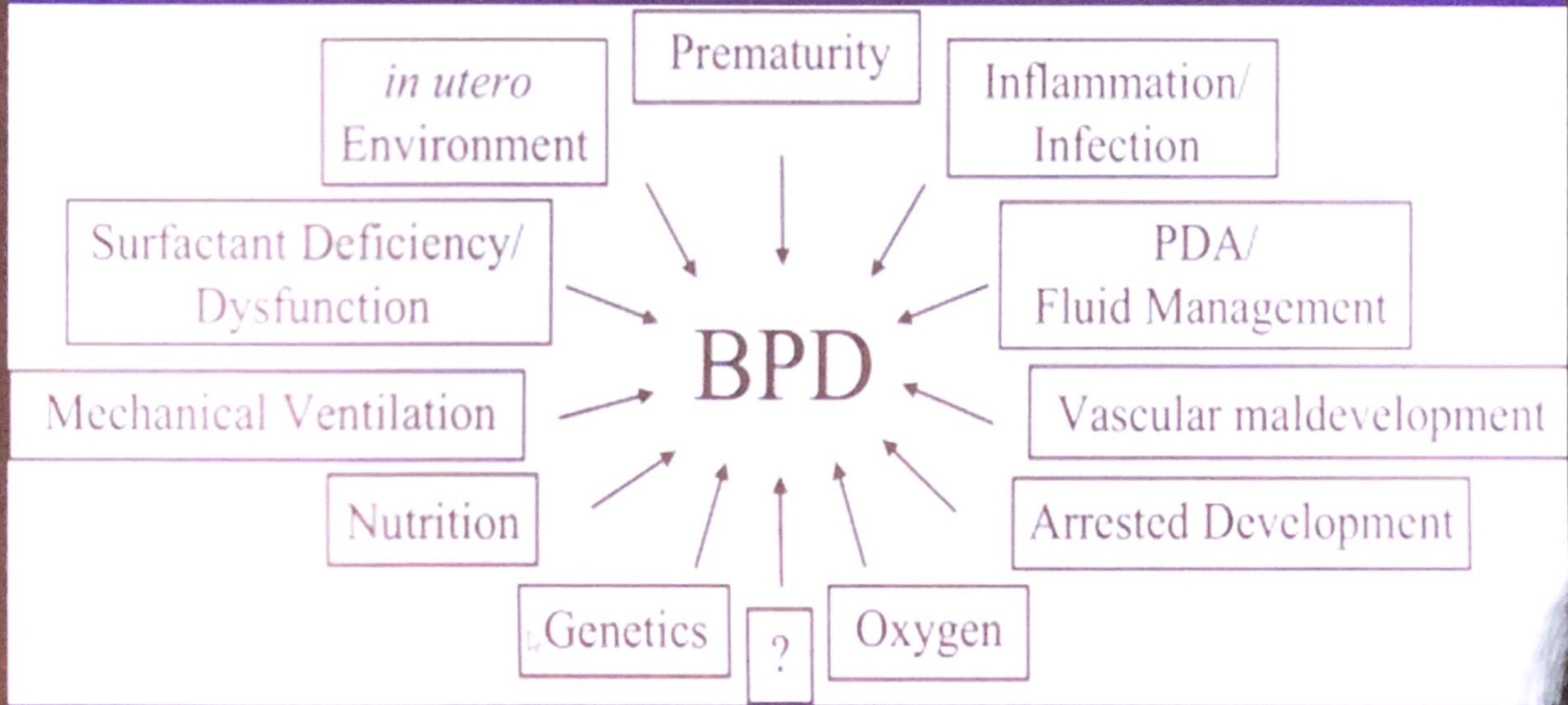
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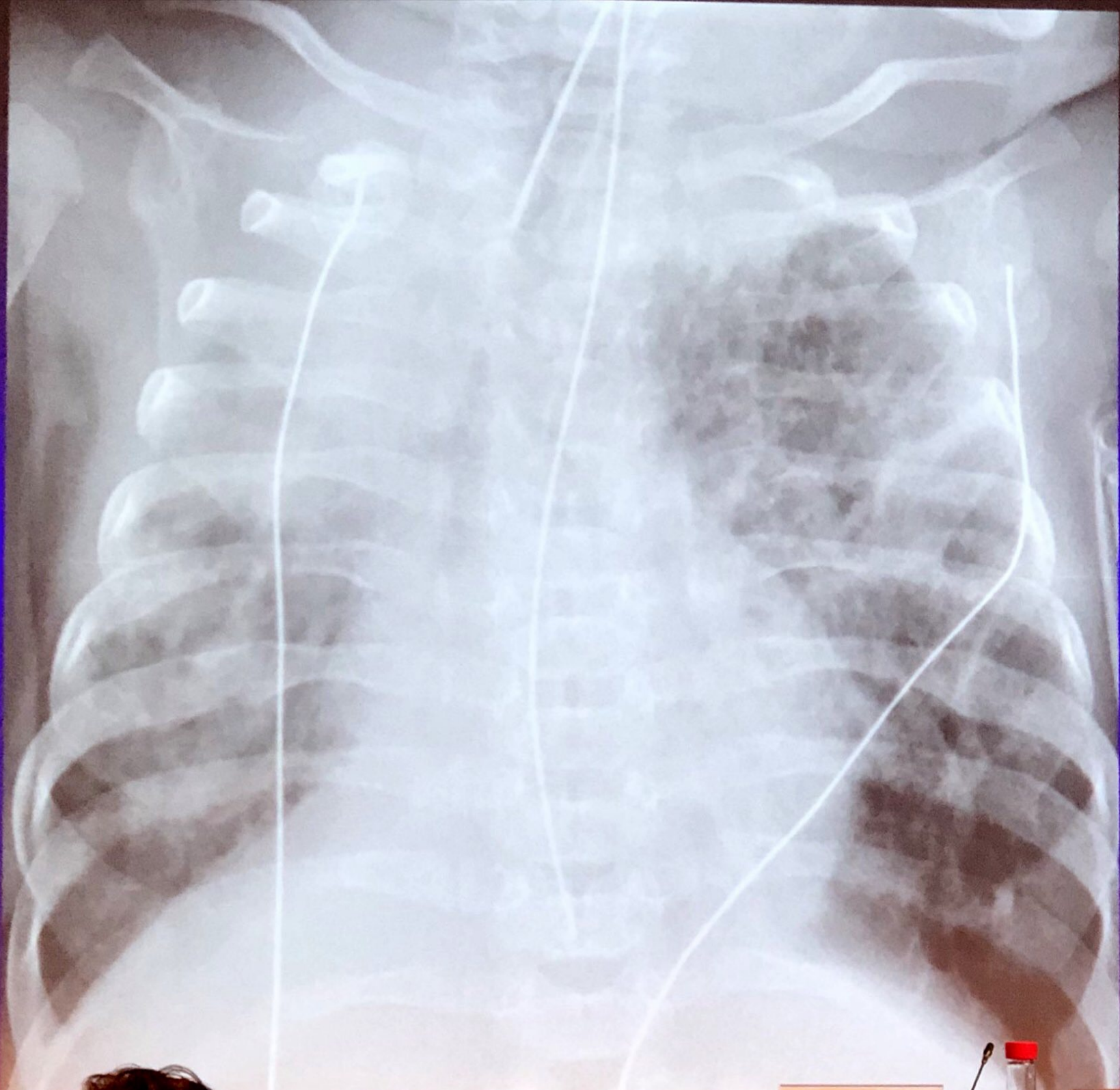
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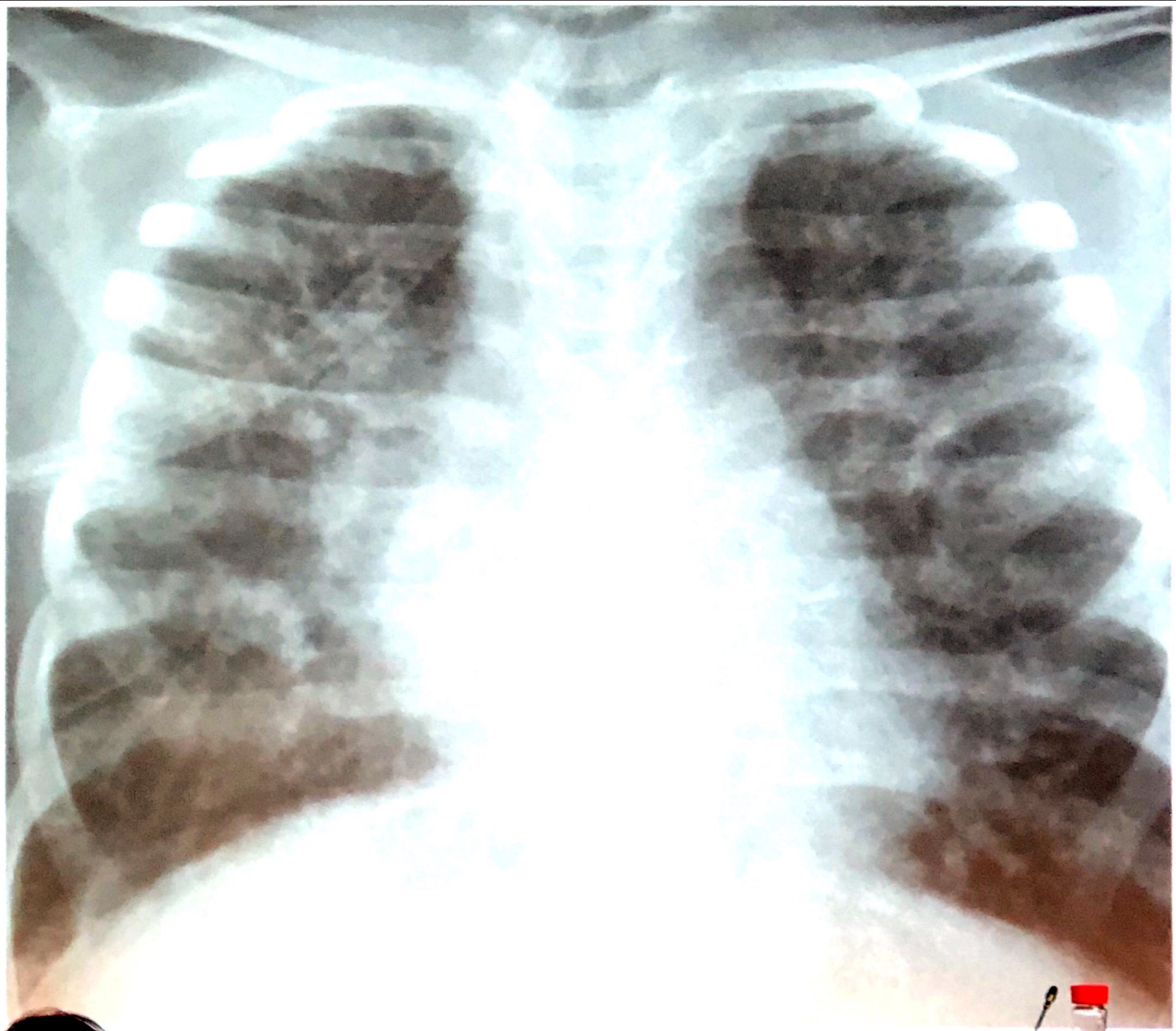
Overview of DILD Cases (n=187)

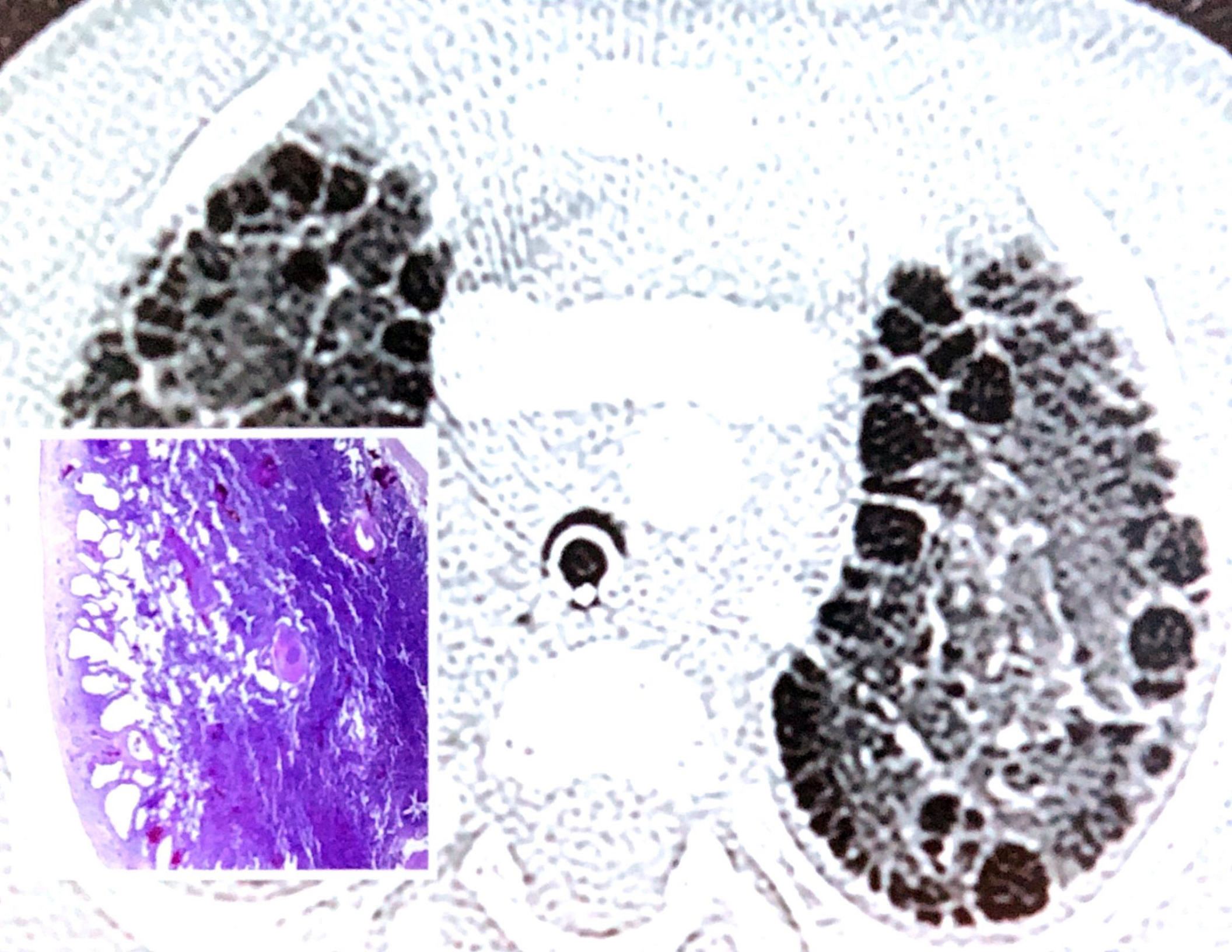
CATEGORY	# cases	%
Diffuse Developmental Disorders	11	6%
Growth Abnormalities	50	27%
PIG	6	3%
NEHI	17	9%
Surfactant Dysfunction	18	10%
Disorders of the Normal Host		16%
Related to Systemic Disease Processes		30%
Disorders of the Immunocompromised Host		11%
Disorders Masquerading as ILD	9	5%
Unclassified	20	11%

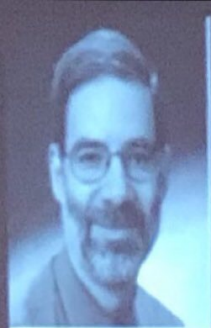
Pathogenesis of “new” BPD







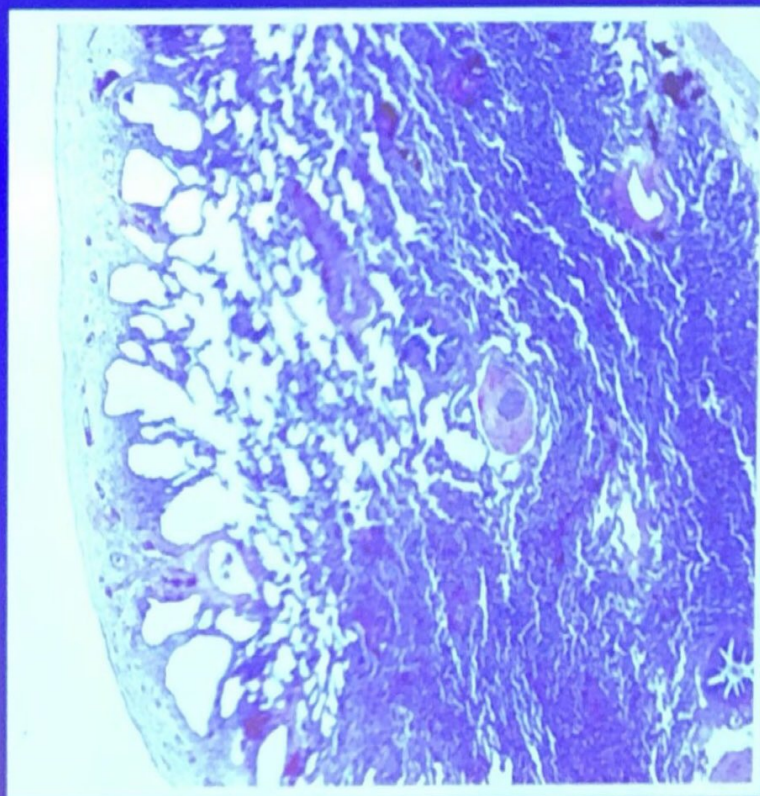




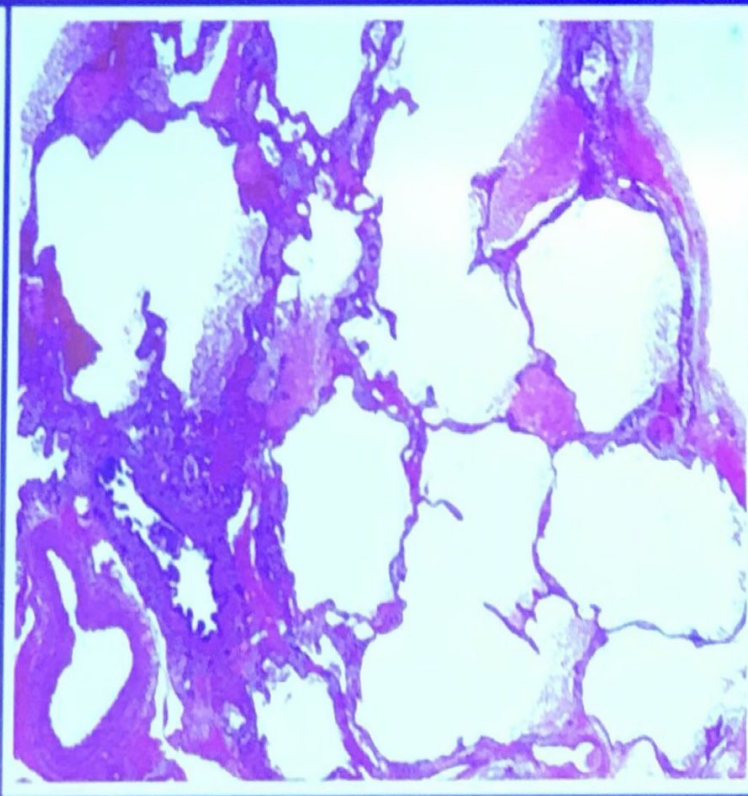
Growth abnormalities reflecting deficient alveolarization

46 total-24.6%

- Related to chromosomal disorders-eg trisomy 21
- Associated with congenital heart disease- (chromosomally normal)

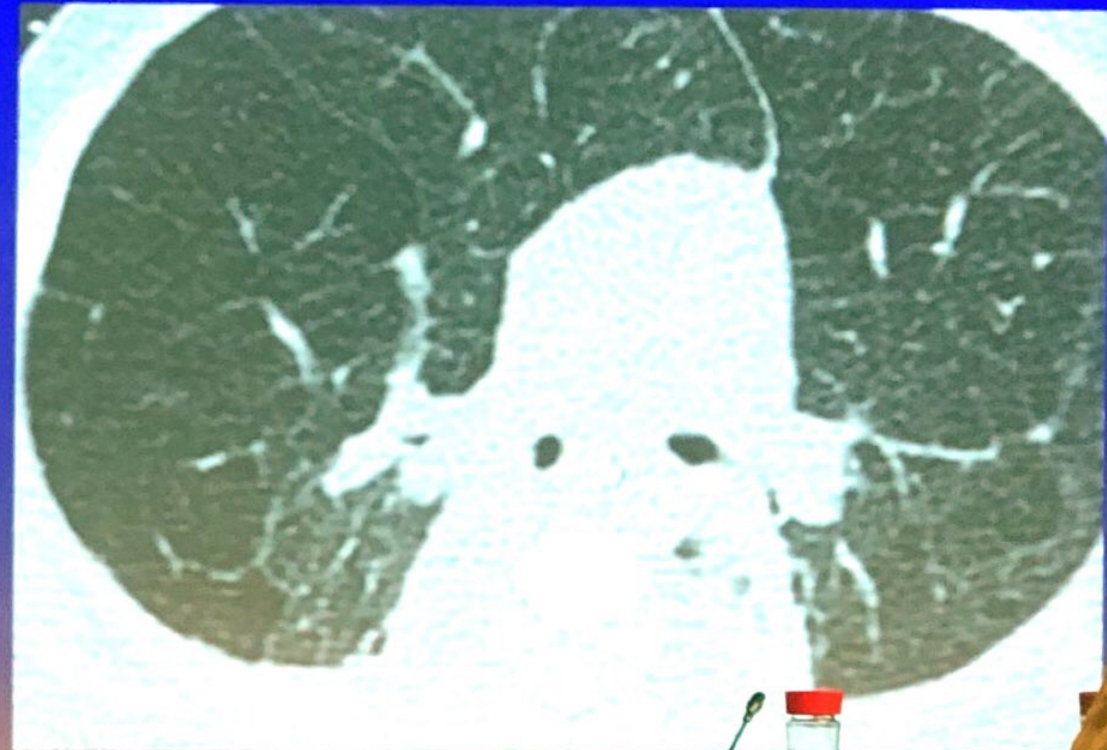
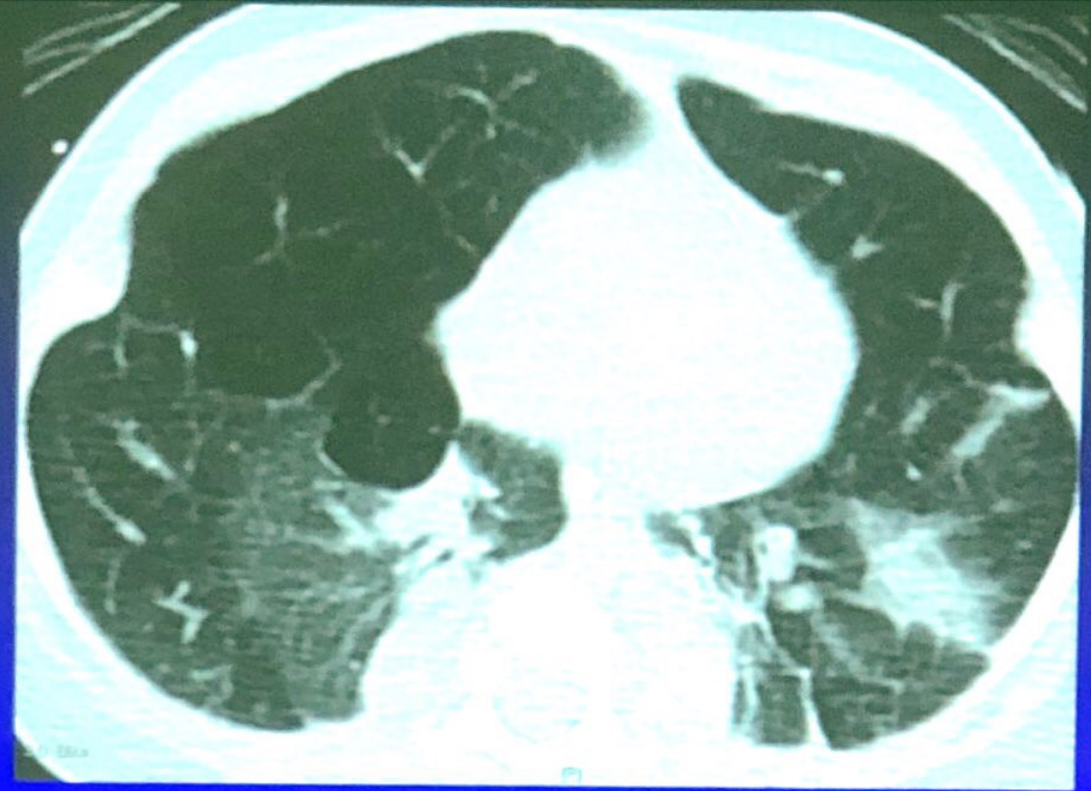


Down Syndrome

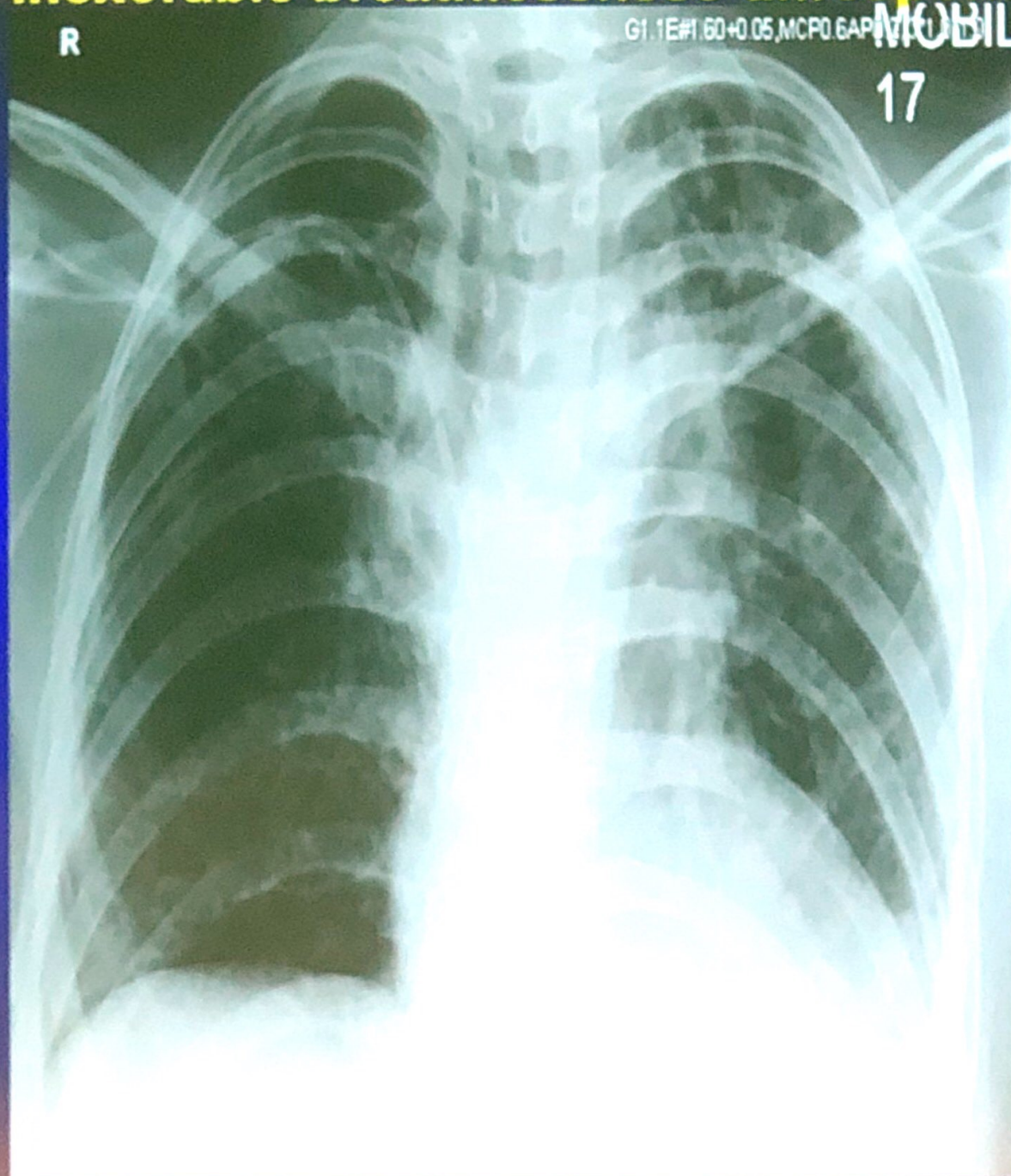


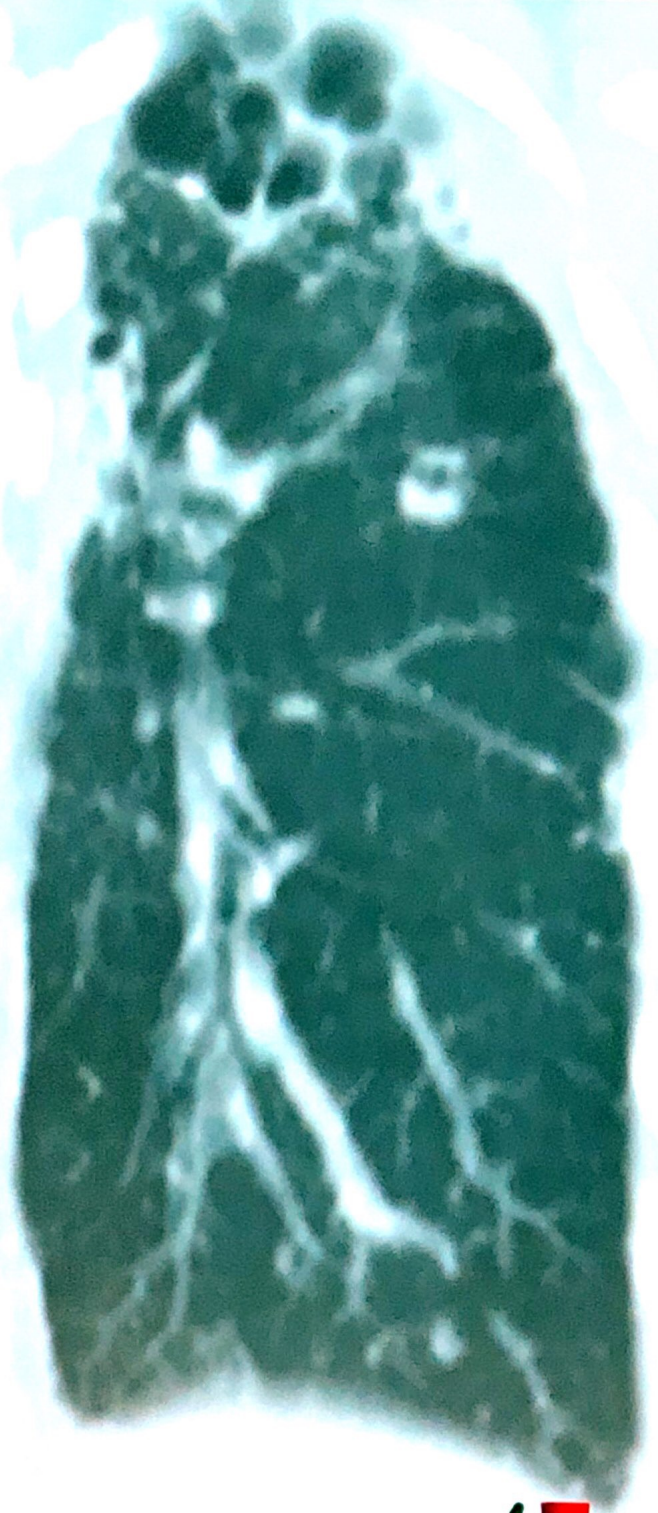
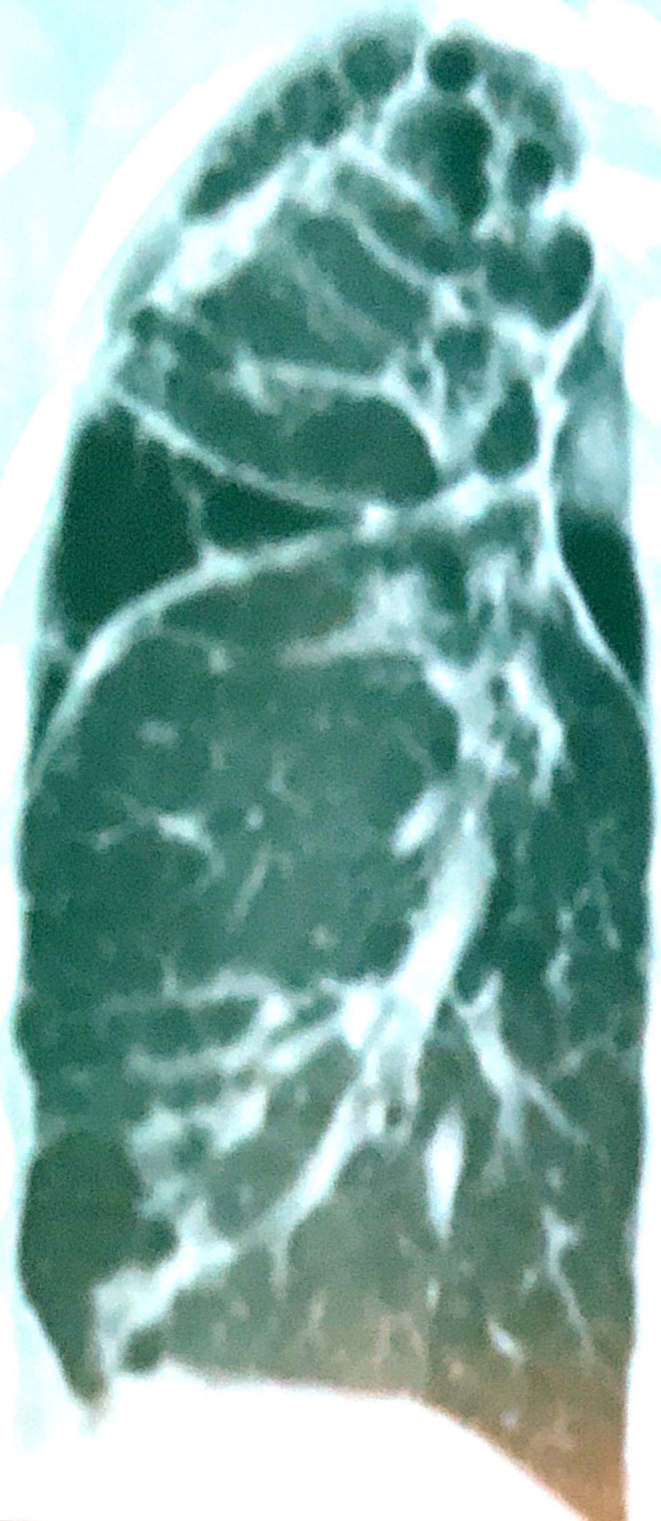
Term infant pulmonary stenosis
transplanted

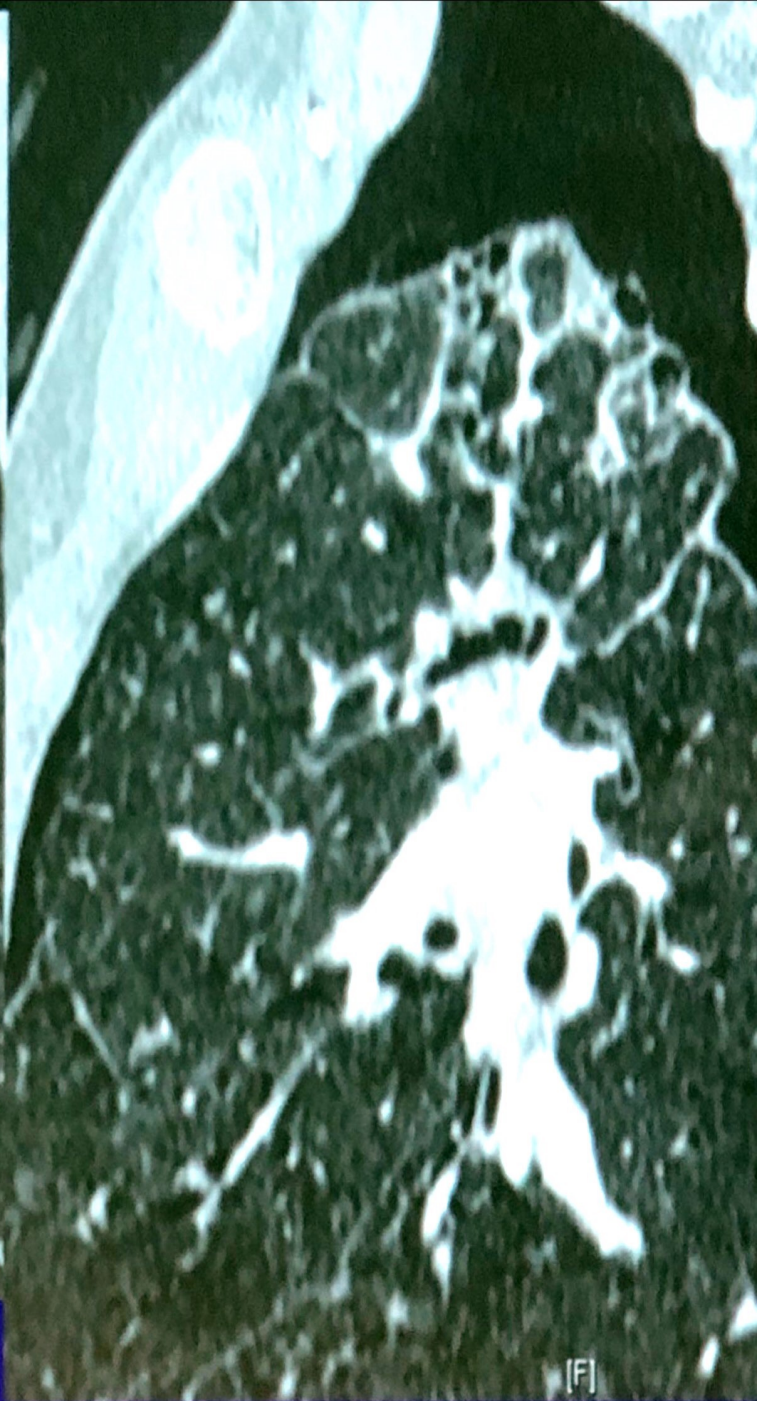
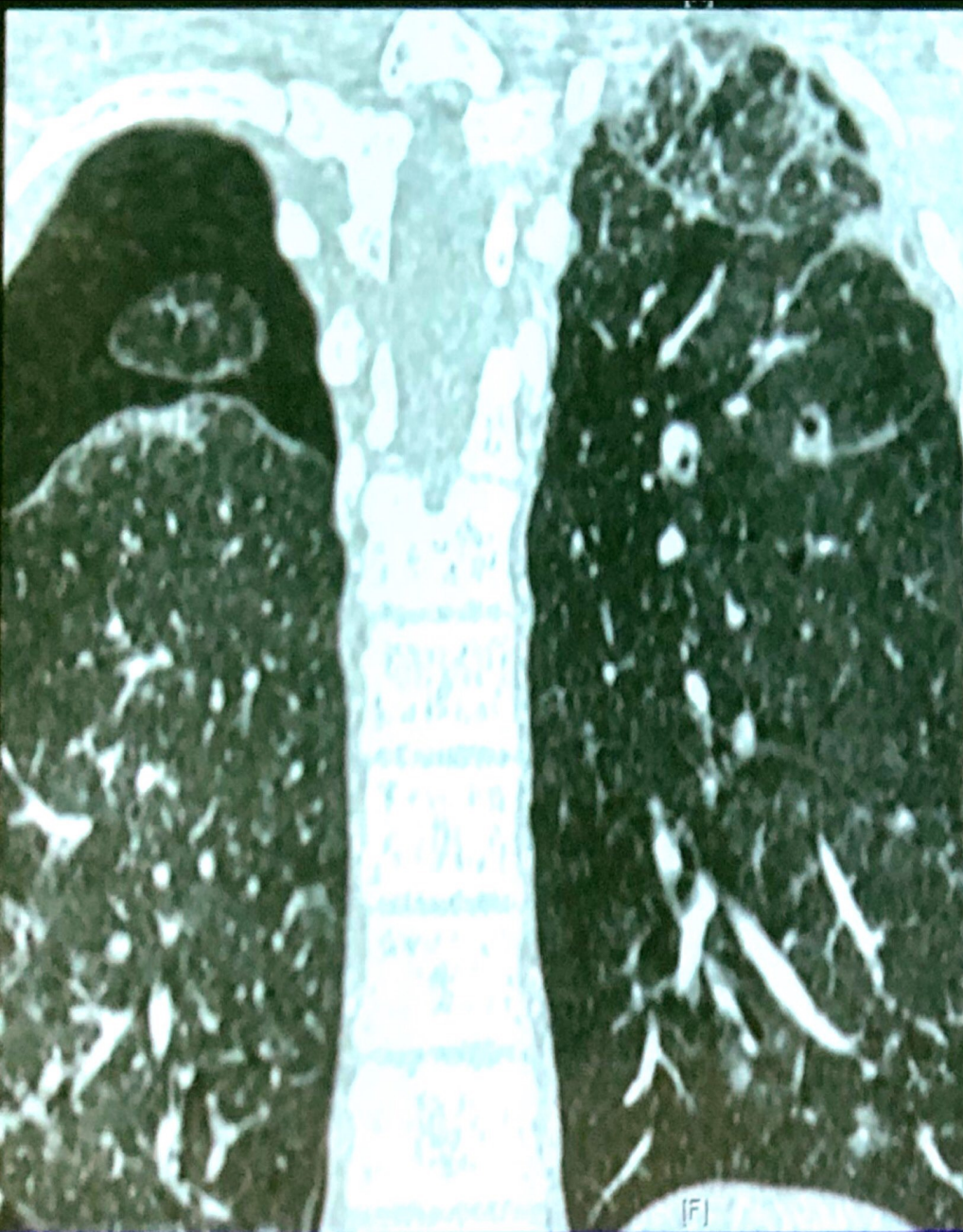
full term neonate with
nystagmus and deteriorating
respiratory function.



12-year-old girl post bone marrow transplant for immunodeficiency presents with inextorable breathlessness unresponsive to therapy.

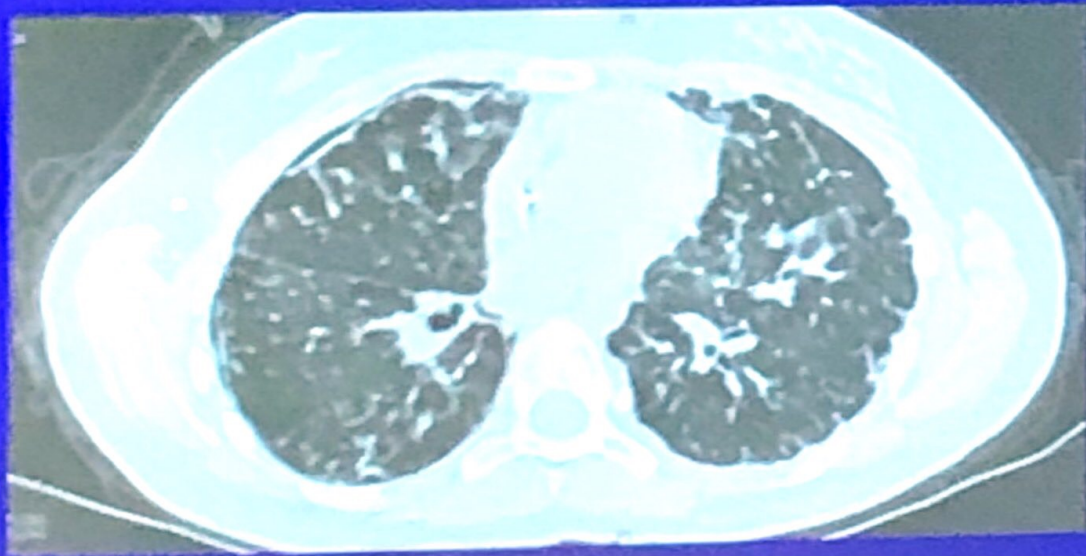




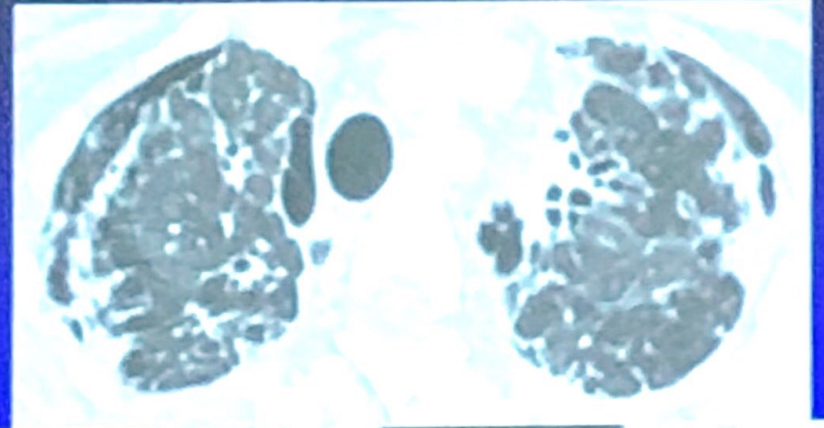


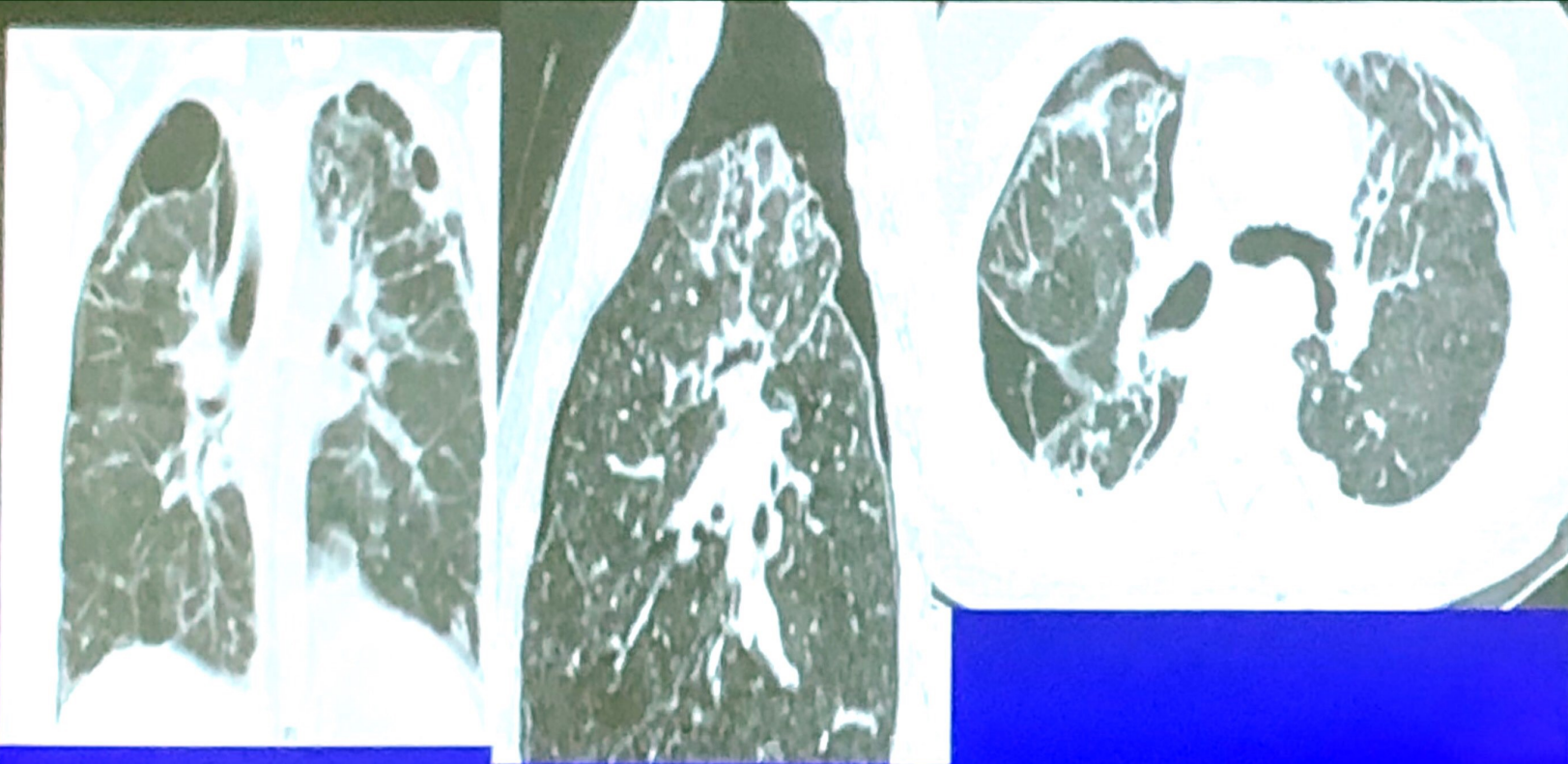
Idiopathic Pleuroparenchymal fibro elastosis iPPFE

- Fibrosis of the peripheral lung parenchyma with pleural thickening



- Predominantly affects the apices





Pleuroparenchymal fibroelastosis (PPFE)

PPFE (pleuroparenchymal fibroelastosis) post allogeneic bone marrow transplantation (Dyskeratosis congenita)

Previously reported as idiopathic

Presented with pneumothorax and sub pleural fibrosis, ABMT related constrictive bronchiolitis, histopathology showed coexistent sub pleural changes

Approach to Paediatric Lung Diseases: Hurdles



Parenchymal Patterns (I)

Normal lung



Mild LAA



Moderate LAA



Severe LAA



Ground glass



Reticulation



Honeycombing



New patterns.....

