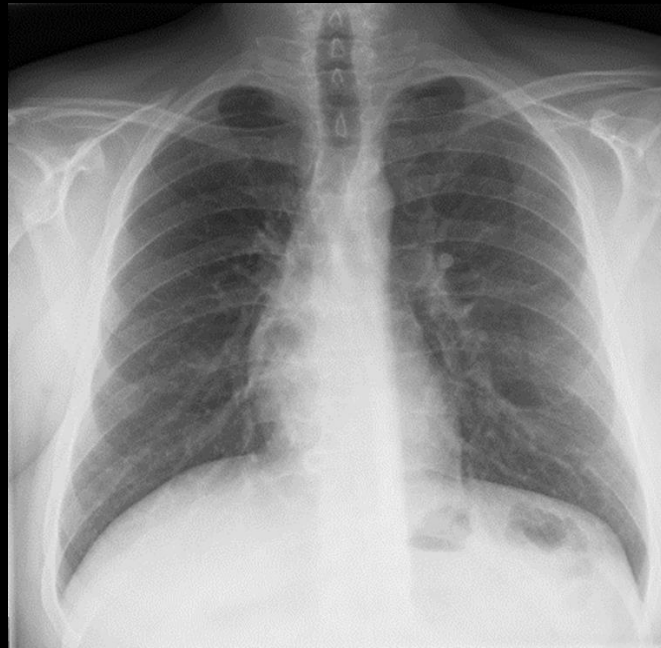


STAFF 5/3/2021

Damienne Vande Berg
Secteur: thorax
Superviseur: B. Ghaye

F, 60 ans, dyspnée
et amaigrissement

- Aplatissement AP thorax
- Élargissement AP thorax
- Attraction proximale des hiles
- Attraction distale des hiles
- Perte de volume des lobes supérieurs



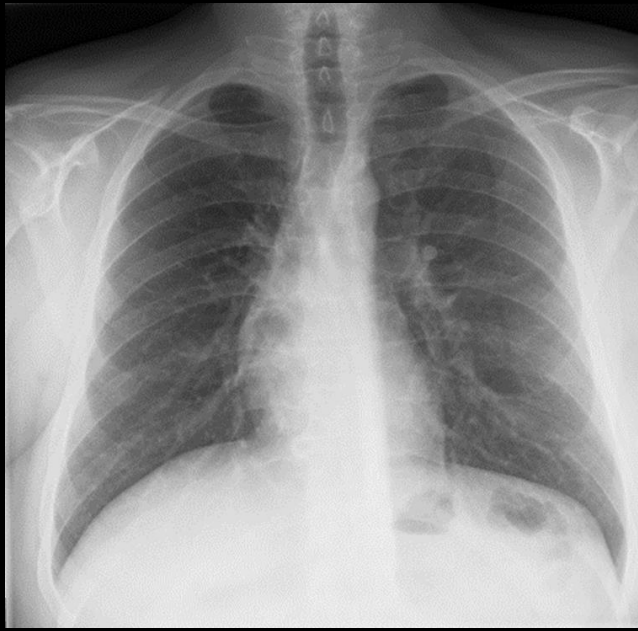
2005



2017



- Aplatissement AP thorax
- Élargissement AP thorax
- Attraction proximale des hiles
- Attraction distale des hiles
- Perte de volume Des lobes supérieurs



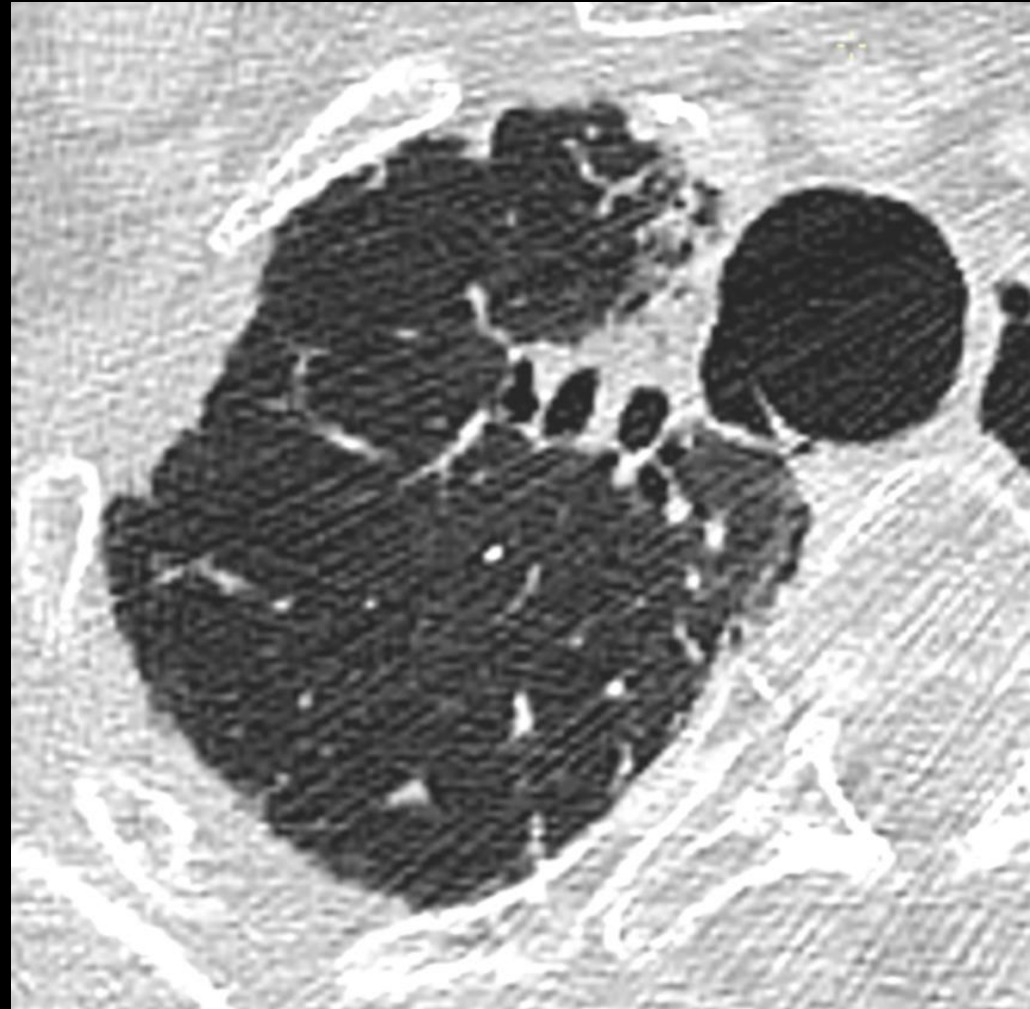
- Coiffe pleurale

- Fibroelastose

Pleuroparenchymateuse

- Wegener

- Sarkoïdose



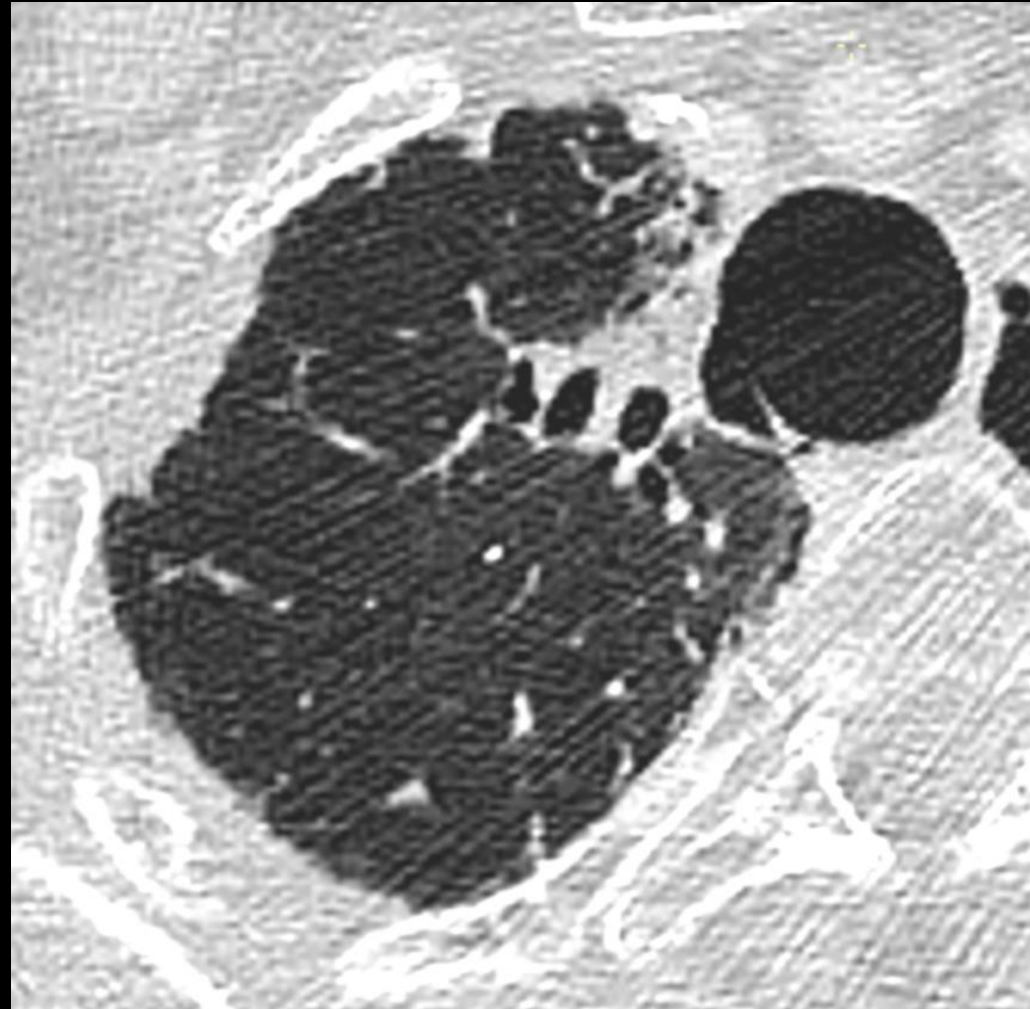
- Coiffe pleurale

- Fibroelastose

Pleuroparenchymateuse

- Wegener

- Sarkoïdose



Fibroelastose pleuroparenchymateuse

= Epaissement pleural + fibrose souspleurale +
perte de volume

Lobes supérieurs

→ Evolution PROGRESSIVE +/- rapide

→ insuffisance respiratoire restrictive

Causes

Type of PPFE	References
Idiopathic PPFE	
Nonidiopathic PPFE	
As a form of restrictive allograft syndrome complicating lung, bone marrow, and hematopoietic stem cell transplant (also known as “restrictive chronic allograft dysfunction”)	12, 13, 17, 20
Fibrotic interstitial lung disease (e.g., usual interstitial pneumonia, hypersensitivity pneumonitis)	23, 27, 31, 32
Chronic or recurrent bronchopulmonary infection (e.g., <i>Aspergillus</i> , nontuberculous mycobacteria)	1, 26, 27, 29, 30
Autoimmune or connective tissue disease (e.g., scleroderma, rheumatoid arthritis, inflammatory bowel disease)	22, 27
Familial history of pulmonary fibrosis	1, 2, 34, 35, 36
Short telomere lengths resulting from mutations of genes encoding the telomerase complex	35, 36
Anticancer/cytotoxic chemotherapy (e.g., cyclophosphamide and carmustine) and radiation therapy	21, 45
Occupational dust inhalation (e.g., asbestos and aluminum)	24, 25

Fibroelastose pleuroparenchymateuse

- H=F
- Tout âge
- Dyspnée, toux, douleur thoracique, amaigrissement
- Platythorax, encoche suprasternale



Diagnostic

- Clinique
- Histologique
- Radiologique
- Marqueurs biologiques ?

Critères diagnostics

Table 2. Proposed diagnostic criteria for pleuroparenchymal fibroelastosis

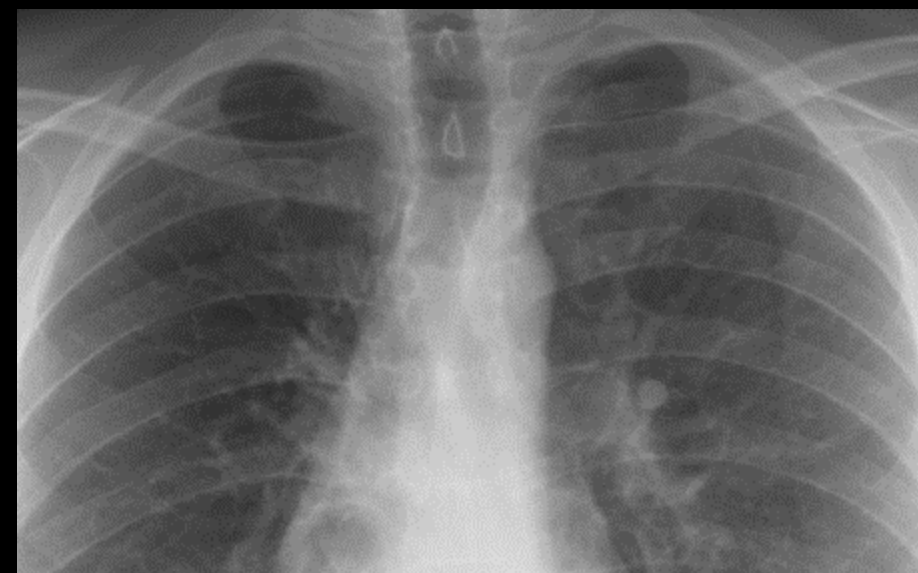
Category	Histopathology	High-Resolution Computed Tomography
Definite PPFE	Upper zone pleural fibrosis with subjacent intraalveolar fibrosis accompanied by alveolar septal elastosis	Pleural thickening with associated subpleural fibrosis concentrated in the upper lobes with less marked or no lower lobe involvement
Consistent with PPFE	Intraalveolar fibrosis present but 1) not accompanied by significant pleural fibrosis, 2) not predominantly subpleural, or 3) not present in an upper lobe biopsy	Upper lobe pleural thickening with associated subpleural fibrosis but 1) distribution not concentrated in the upper lobes or 2) with features of coexistent disease elsewhere
Inconsistent with PPFE	Absence of features in "definite PPFE" and "consistent with PPFE" categories	Absence of features in "definite PPFE" and "consistent with PPFE" categories

Definition of abbreviation: PPFE = pleuroparenchymal fibroelastosis.

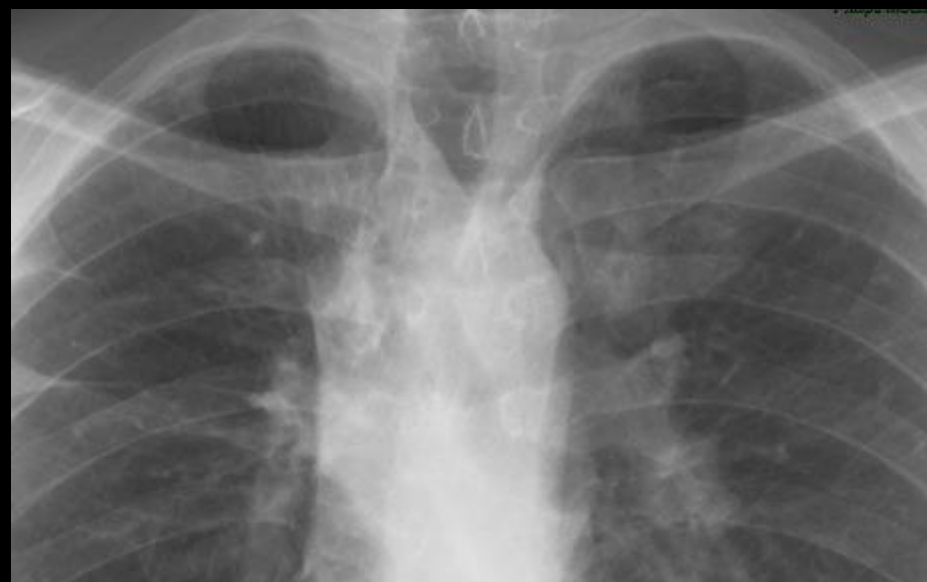
Reprinted by permission from Reference 27.

Rx

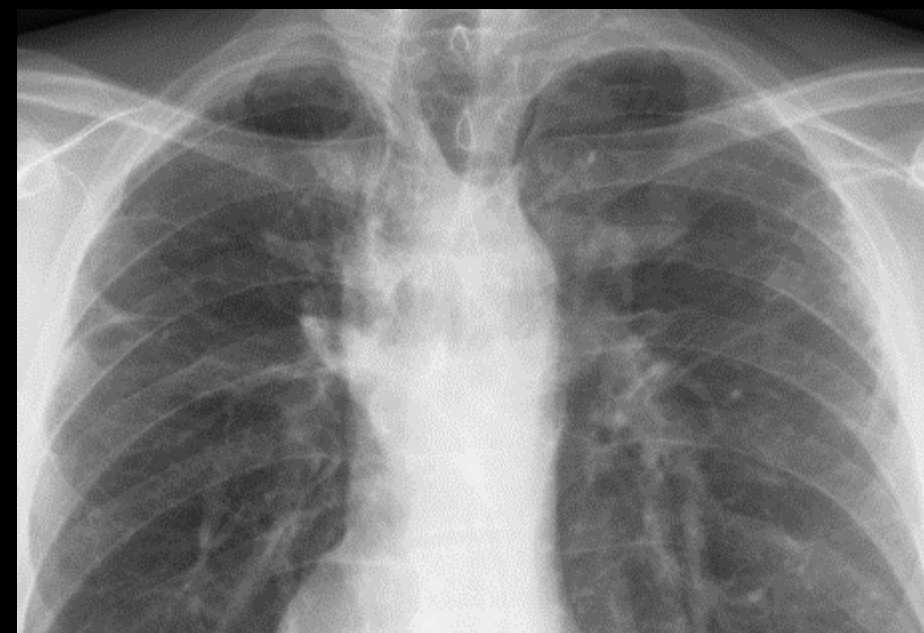
- Epaissement pleural lobes supérieurs
- Perte de volume LS avec ascension des hiles
- platythorax



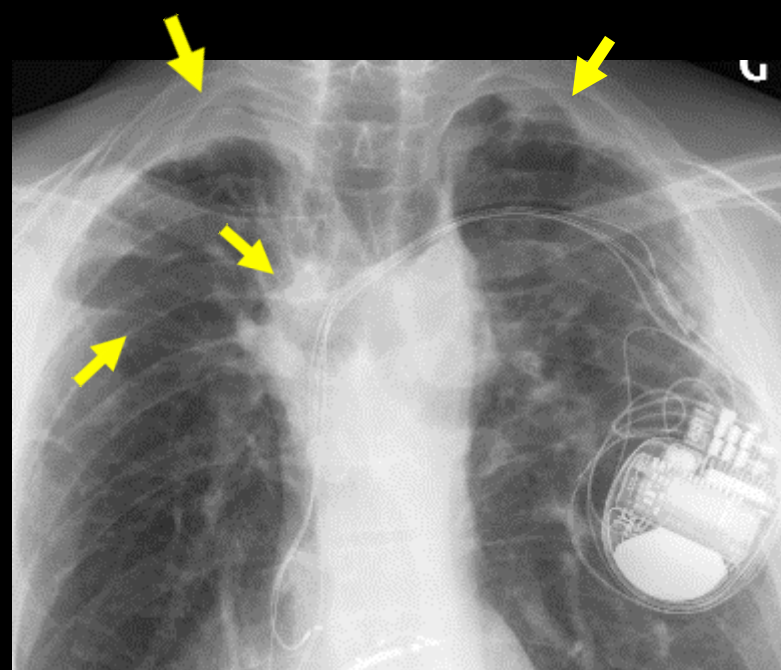
2005



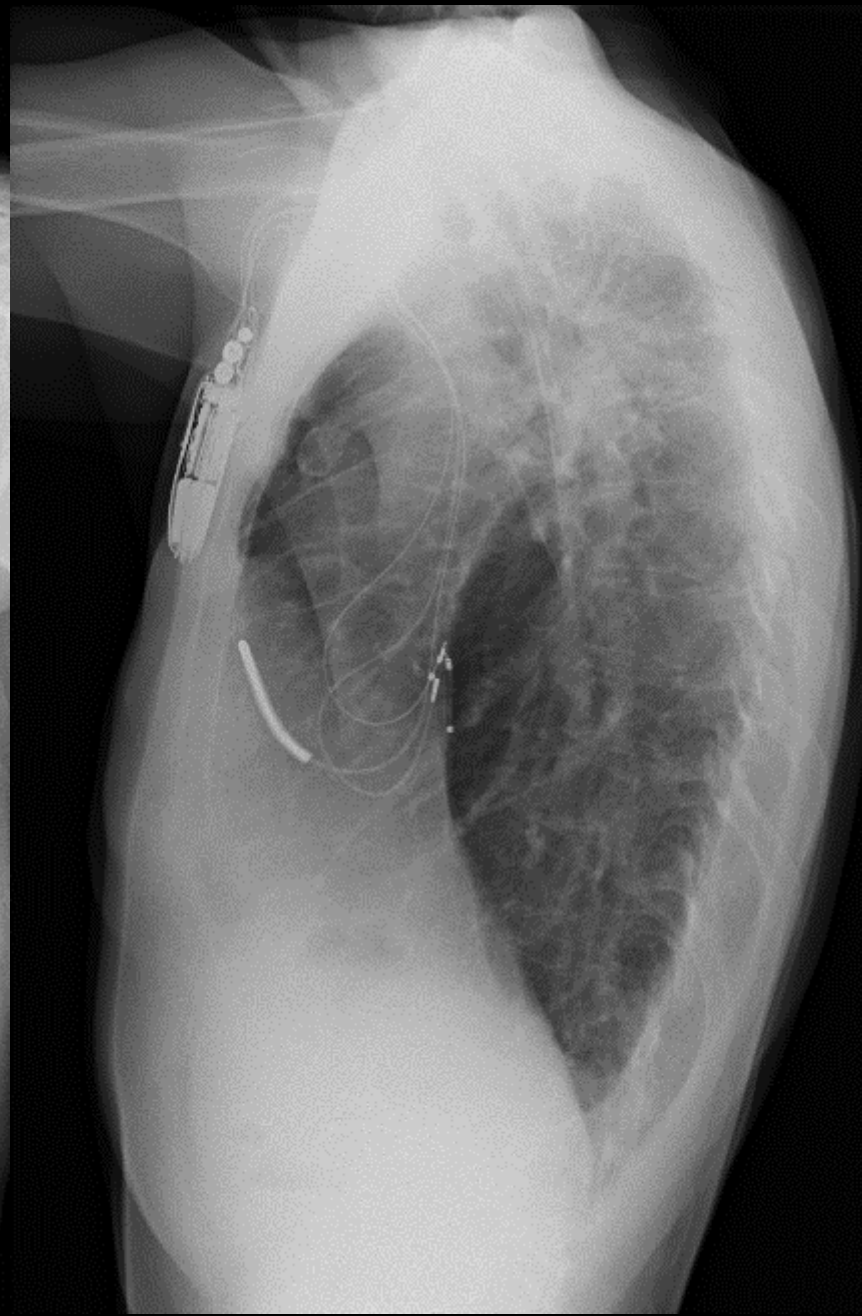
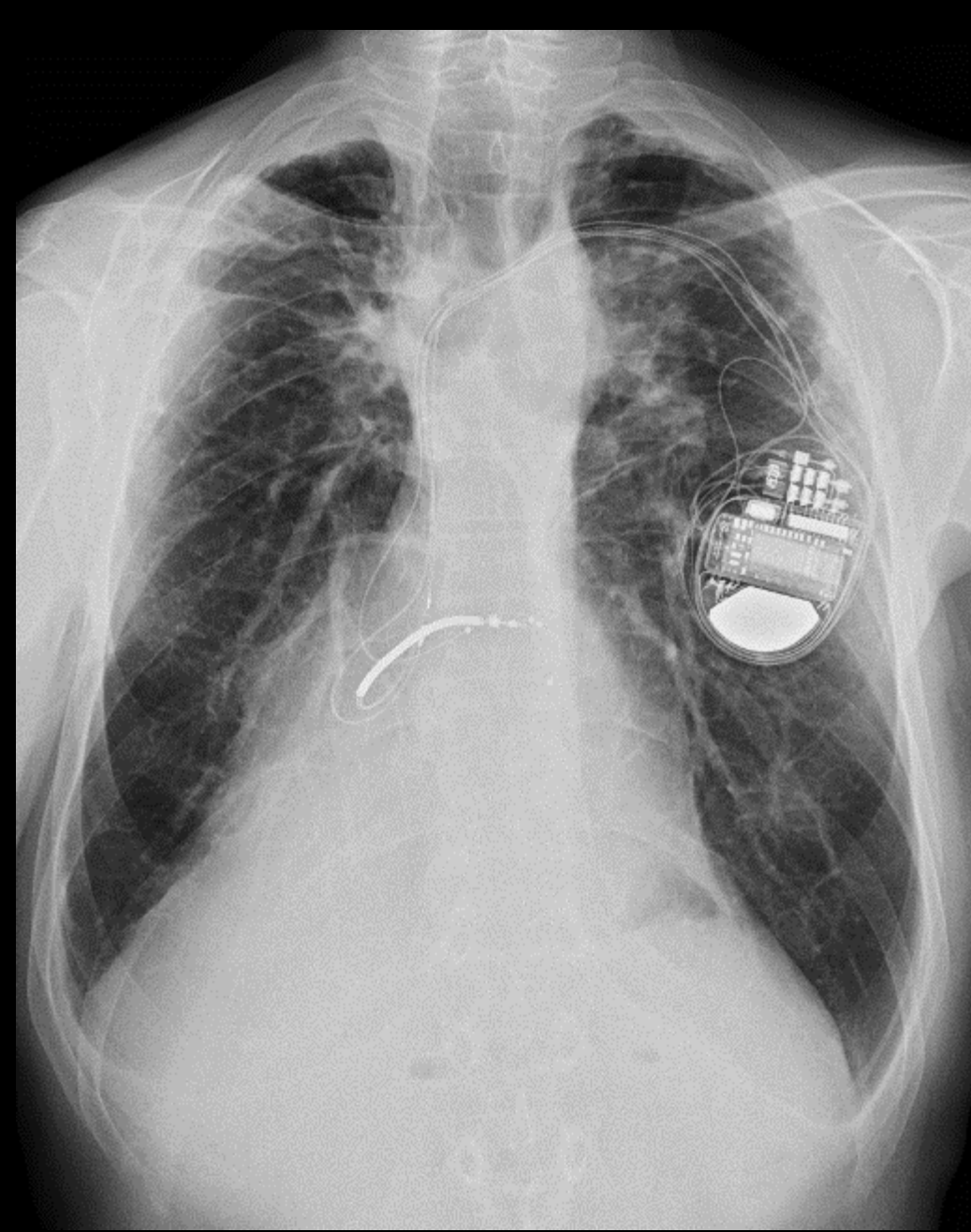
2008



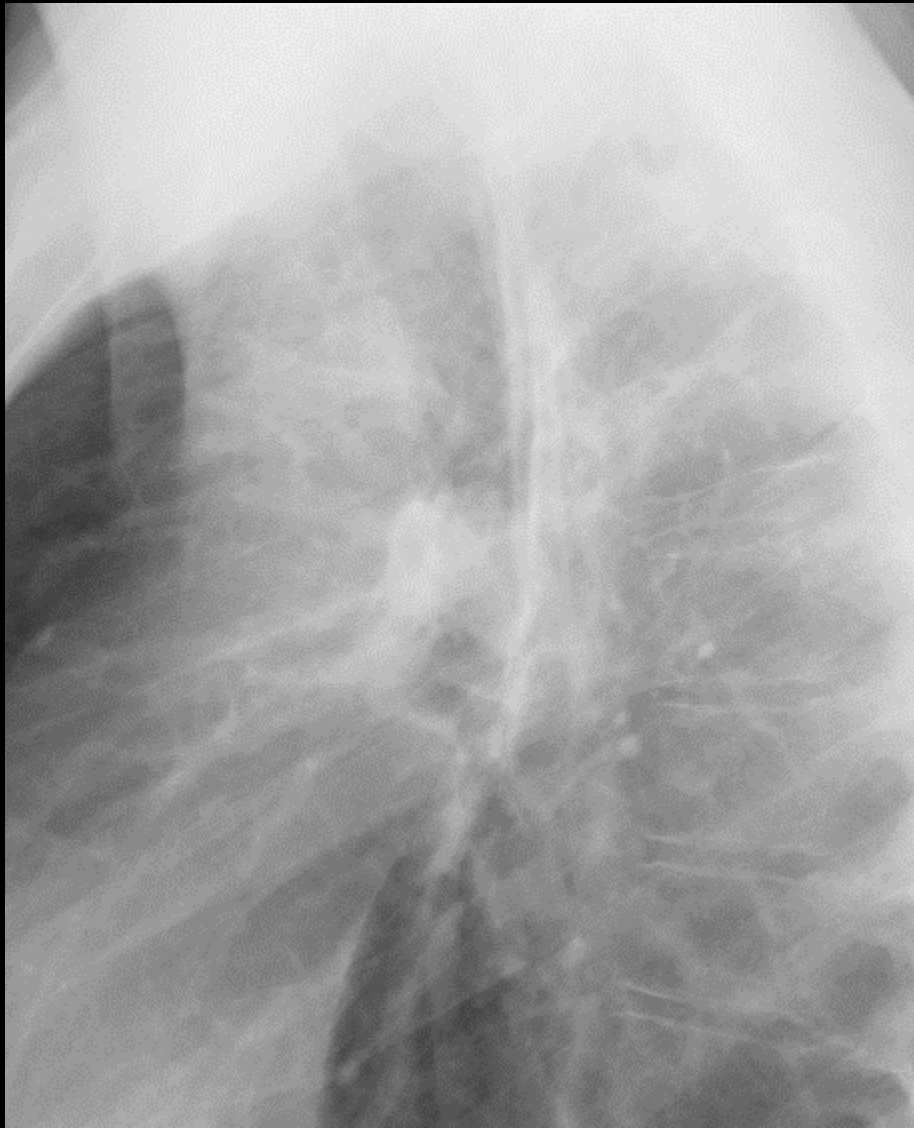
2012



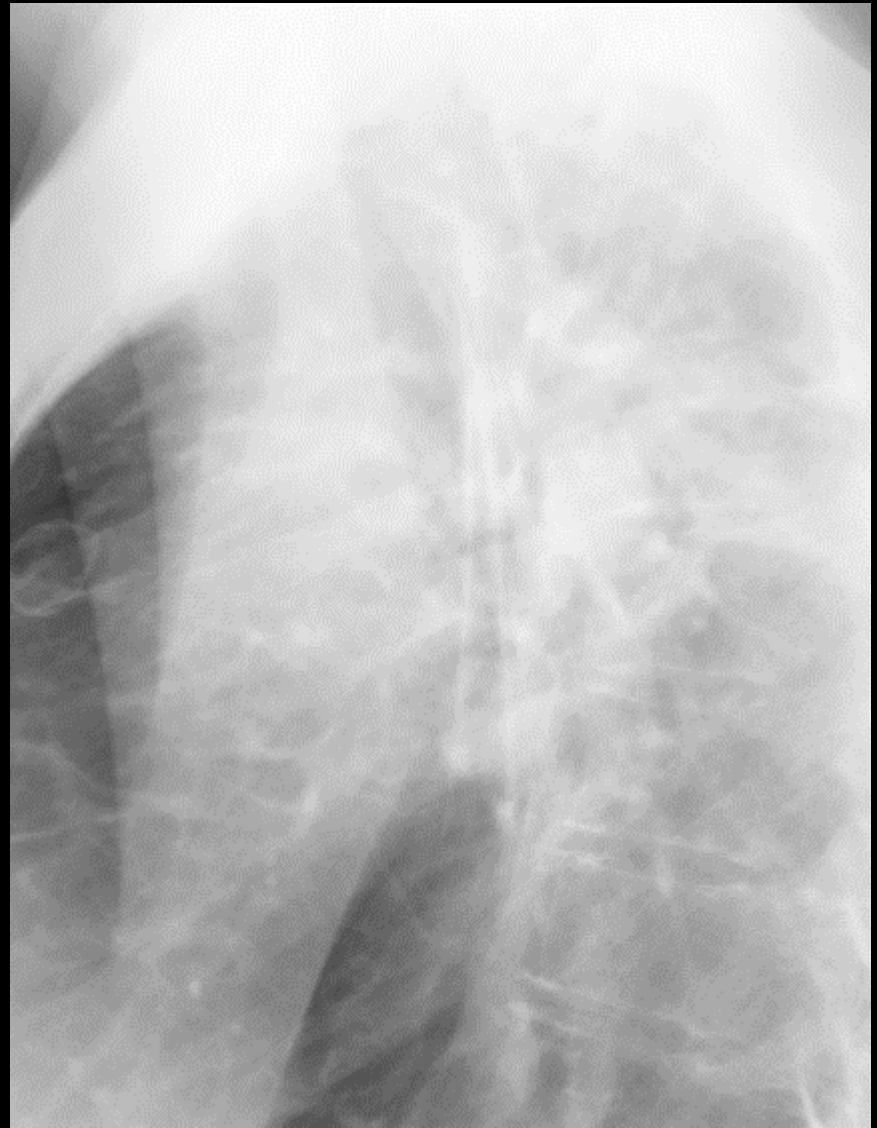
2019



2020



2005

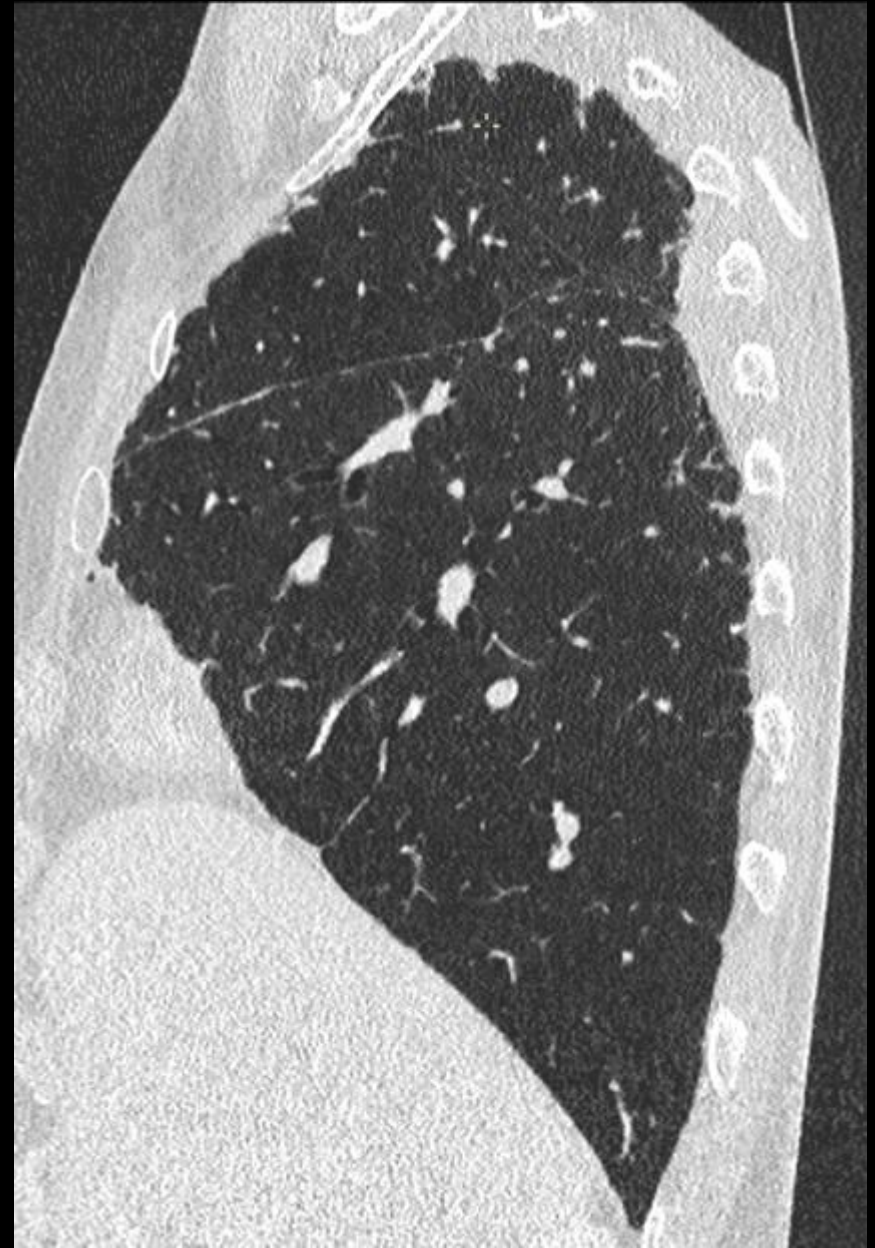
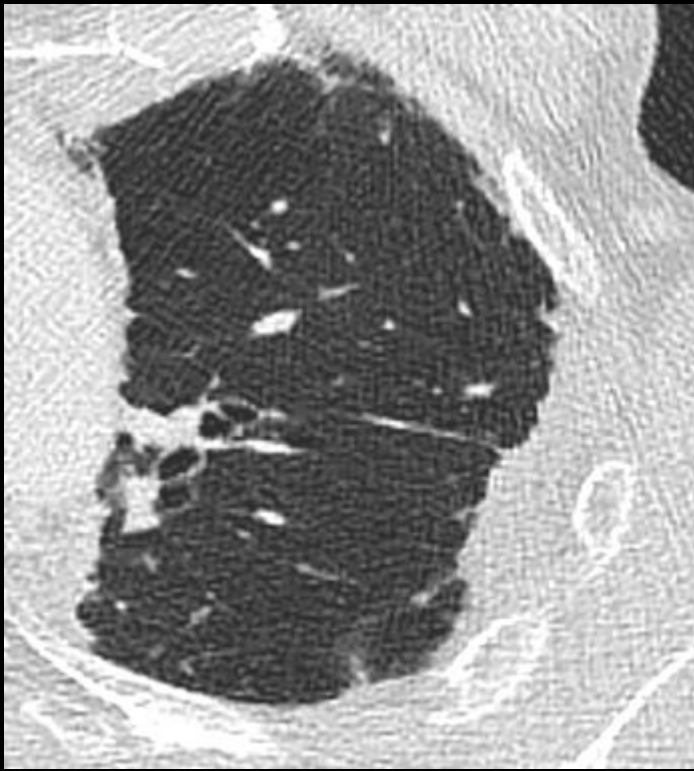


2019

TDM

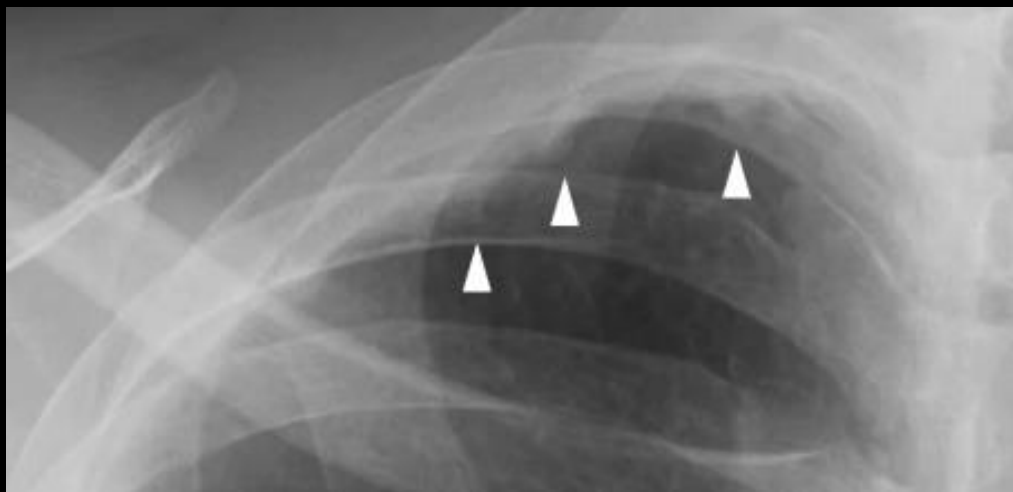
- Epaissement pleural LS
- Fibrose souspleurale
- Encoche suprasternale profonde
- Bord postérieur trachée dépasse bord antérieur vertèbre





Diagnostic différentiel

- Coiffe apicale
- Sarcoïdose
- PHS
- Mycobactéries
- Séquelles post-radiques
- Asbestose
- Connectivite
-



Ref. 3

Take home message

- FEPP= Epaississement pleural + fibrose souspleurale principalement lobes supérieurs

maladie EVOLUTIVE

-> Comparer aux examens antérieurs !

références

1. Chua, F, Desai, SR, Nicholson, AG, et al. Pleuroparenchymal fibroelastosis: a review of clinical, radiological and pathological characteristics. *Ann Am Thorac Soc* 2019; 16: 1351–1359
2. Philippe Camus, Jan von der Thüsen, David M. Hansell, Thomas V. Colby. *European Respiratory Journal* 2014 44: 289-296; DOI: 10.1183/09031936.00088414
3. Saito, A., Hakamata, Y., Yamada, Y. *et al.* Pleural thickening on screening chest X-rays: a single institutional study. *Respir Res* **20**, 138 (2019). <https://doi.org/10.1186/s12931-019-1116-9>