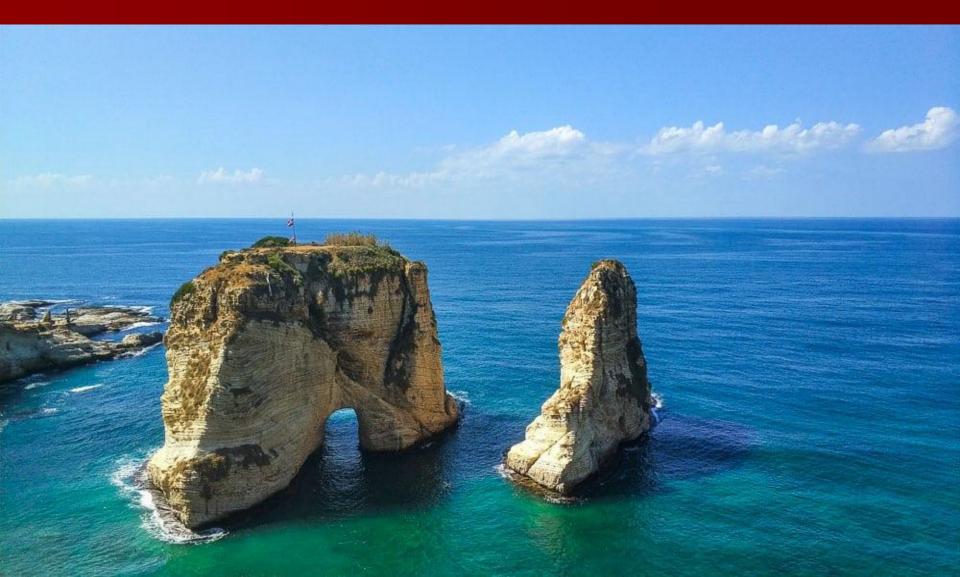
#### **Rare Encounters**

Dr A R Anani, FRCP Consultant Pulmonologist

#### Welcome All



#### Happy and Honoured



#### **Clinical First Rule**

Common cases are commonThey make our bread and butter

#### **Atypical Presentation**

- Some common diseases may have unusual features.
- Others may suggest a rare disease or syndrome.
- Some are more challenging, even Google can not help: Extra effort is needed.
- These are the spices of our work.

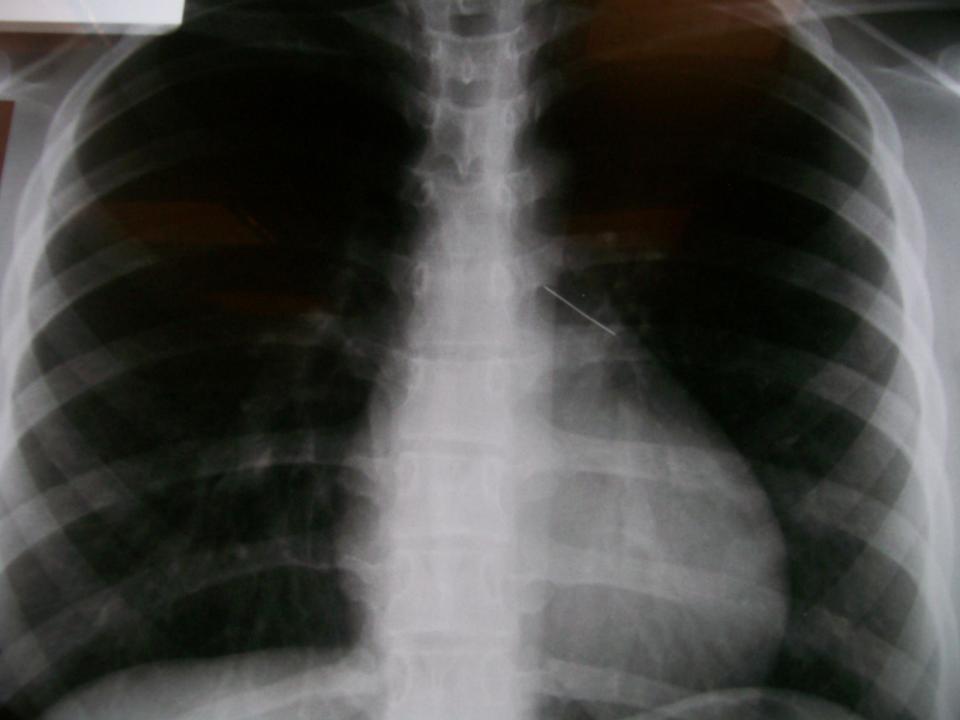
#### **This Presentation**

Purely clinical looking at some rare published encounters that needed extra effort

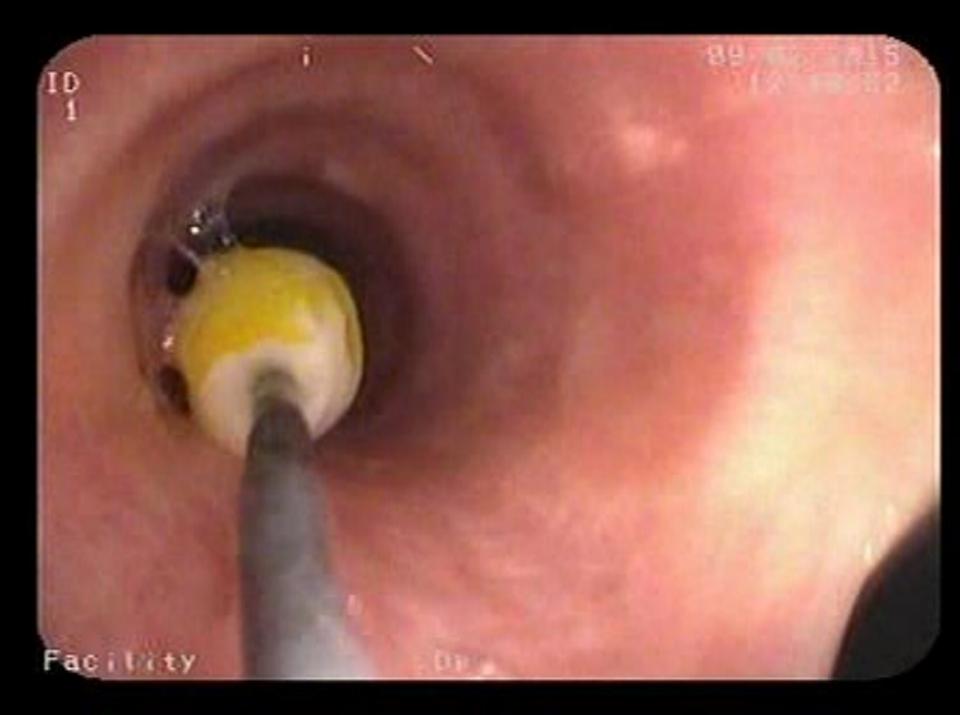
#### Simple Straightforward Problems

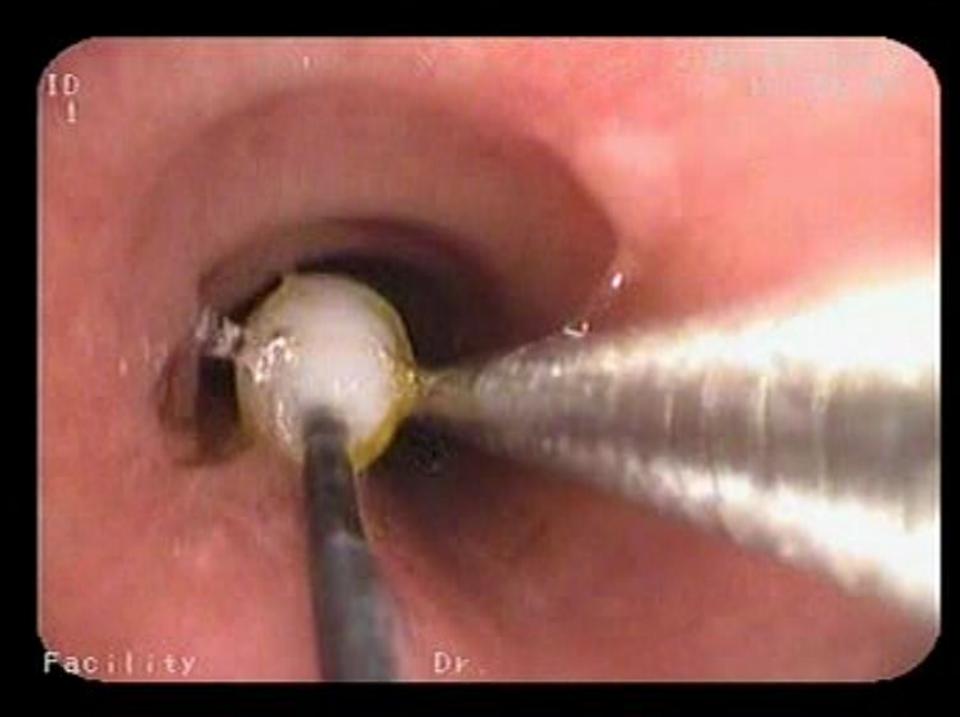
Or Are They?

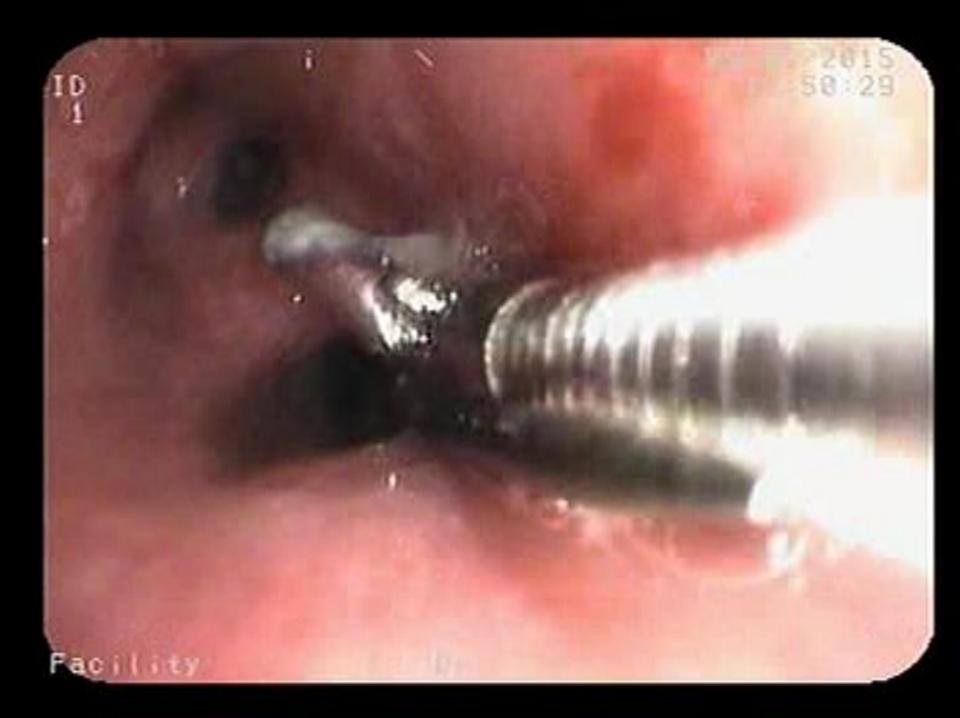


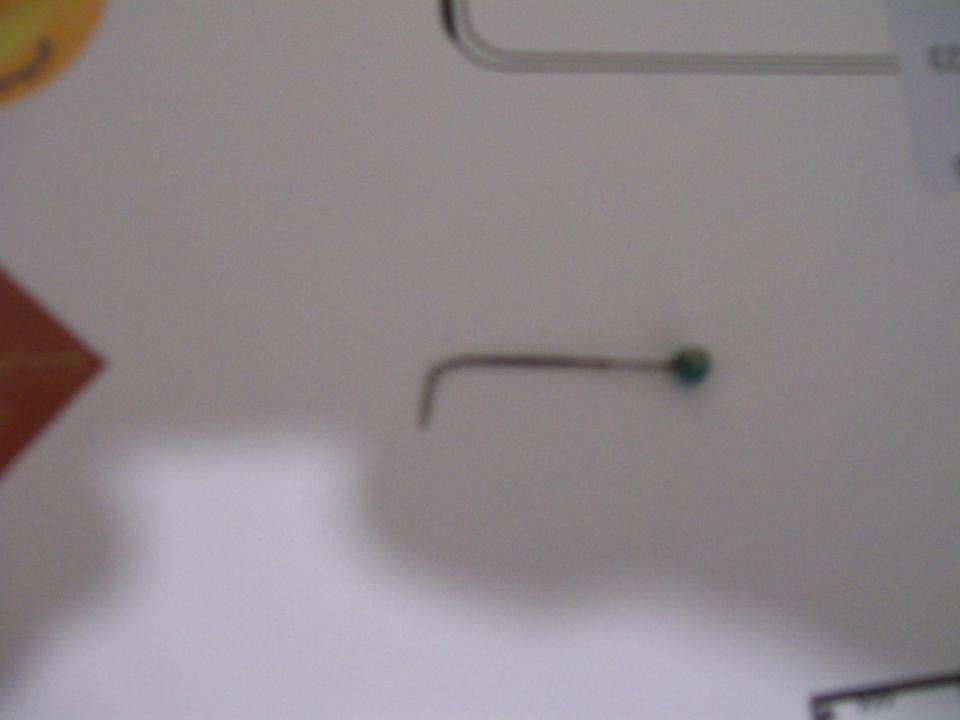














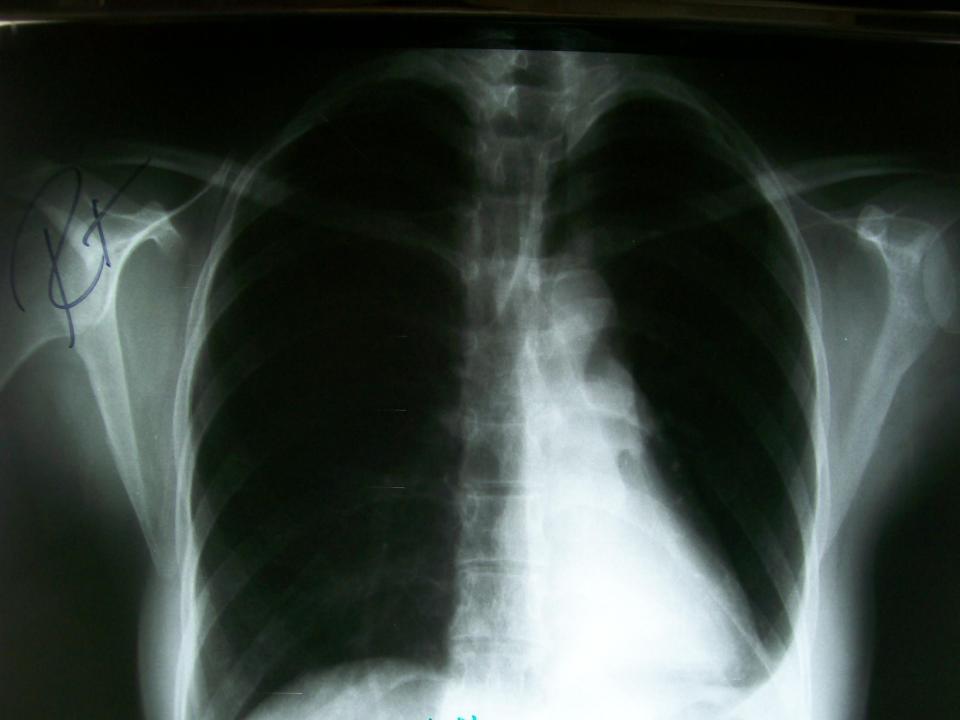




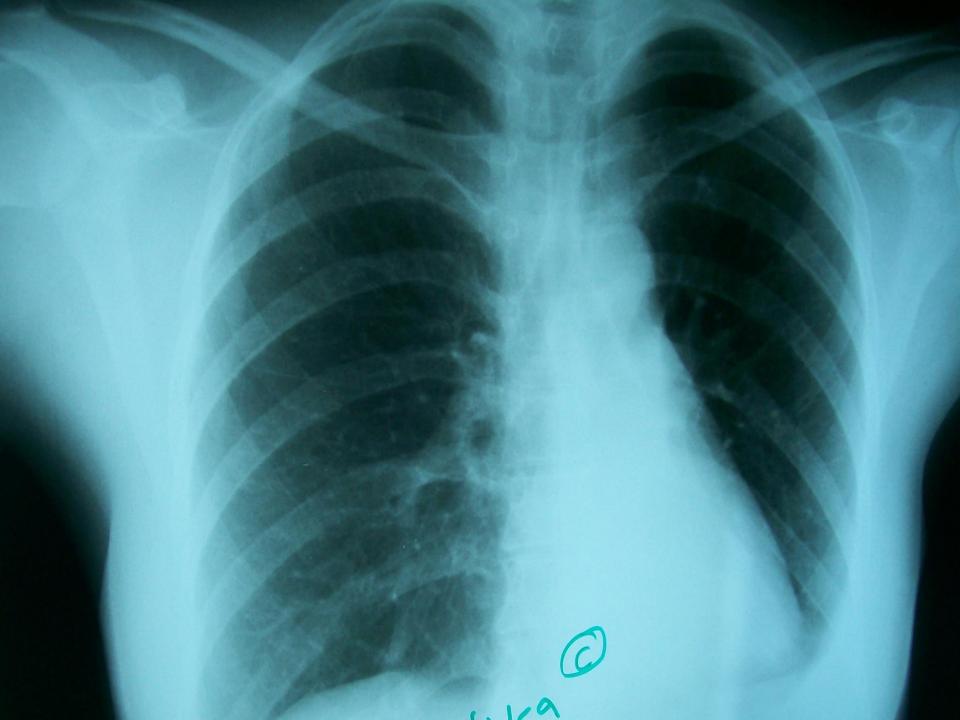


30 year old professional non smoker
Recently gave birth to a baby boy
She always enjoyed good health although pregnancy was troublesome

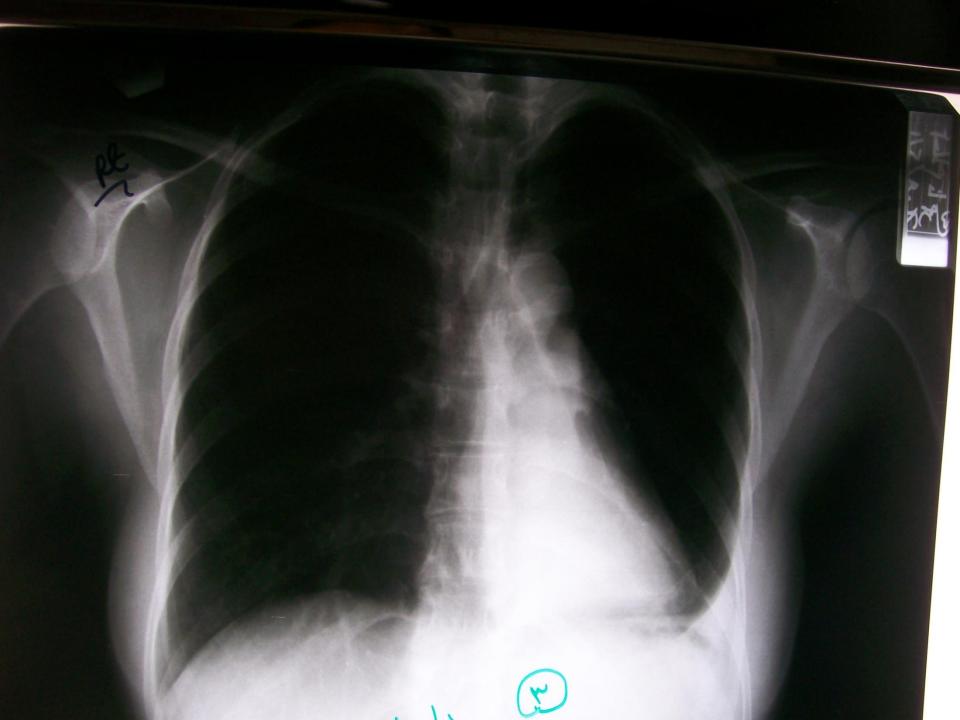
4 weeks ago she developed dry cough, no fever or other symptoms
GP prescribed antibiotic
Not better
He referred her for chest x ray



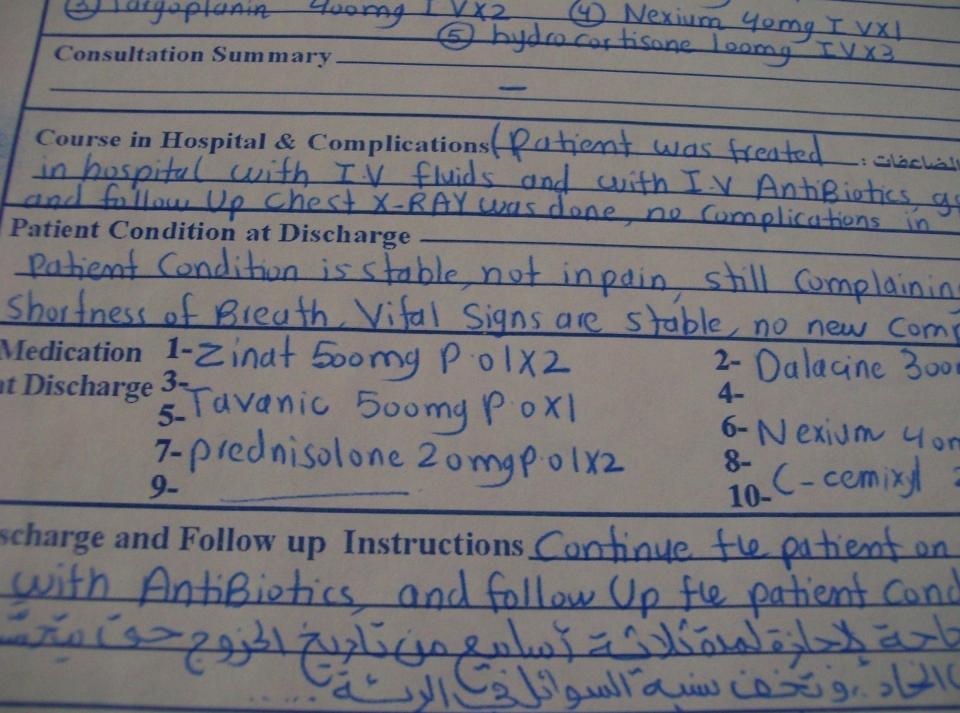
He gave a course of I V antibiotic
Chest x ray after one week: No change
Cough became troublesome



GP referred her for admission
She stayed in hospital for 3 weeks
She was tried on 5 different I v antibiotics with no improvement clinically or radiologically



 Her doctor, feeling stuck, decided to get rid of the problem by sending her home refusing to seek second opinion
 He sent her home on 2 other antibiotics!!!!

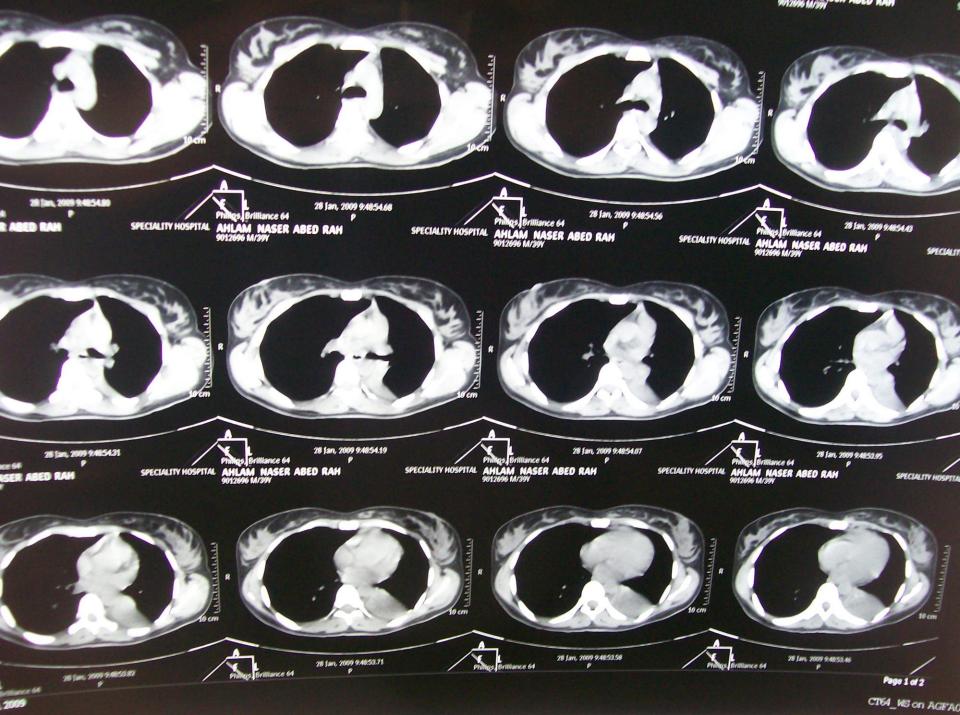


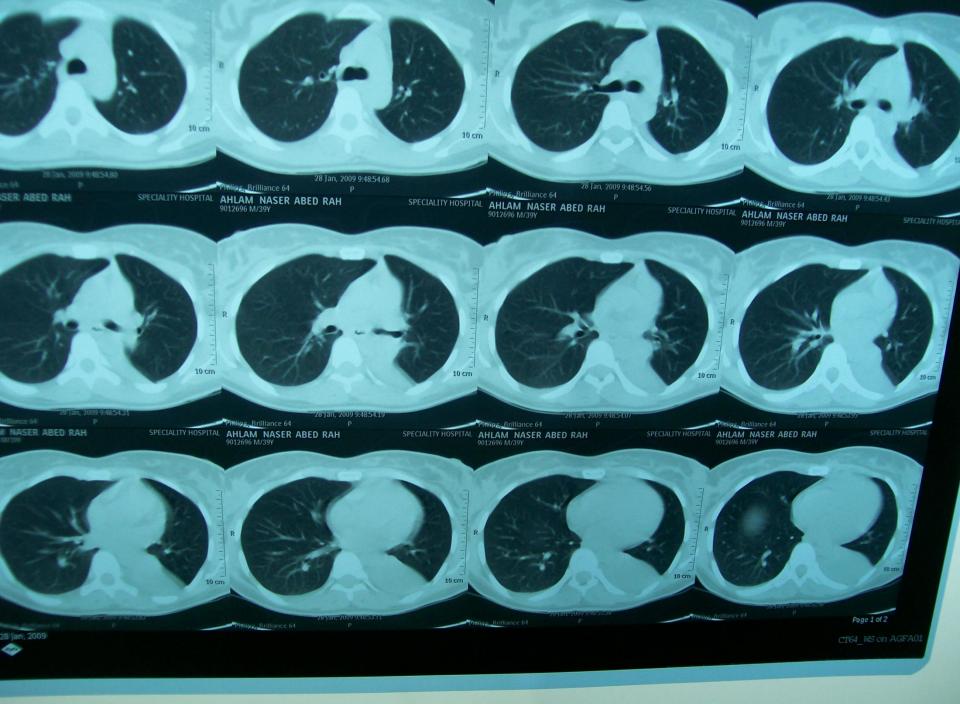
#### Lesson

# Insanity: doing the same thing over and over again and expecting different results.

- Albert Einstein

As she was really tired, her husband brought her to me
After reviewing her case, I requested CT scan





| The second second | <br>20 |     |    |
|-------------------|--------|-----|----|
|                   |        | 12: | 11 |

#### Report :

Chest CT

Multiple contiguous axial slices without contrast enhancement, coronal and sagittal reconstruction were obtained.

Evidence of consolidation with loss of volume is seen involving most of the of the LLL, in keeping with pneumonia in view of clinical history. Minimal shifting of the mediastinum to the Lt side is noted. Mild Lt pleural effusion is seen. The rest of the lungs are clear. No definite hilar or mediastinal adenopathy is visible.

No gross masses. However, this is a limited non-enhanced study

Conclusion:

========

-Consolidation in the the LLL with loss of volume and minimal Lt pleural effusion. - No gross adenopathy or masses.

Dr.Mustafa Al-Sabbagh

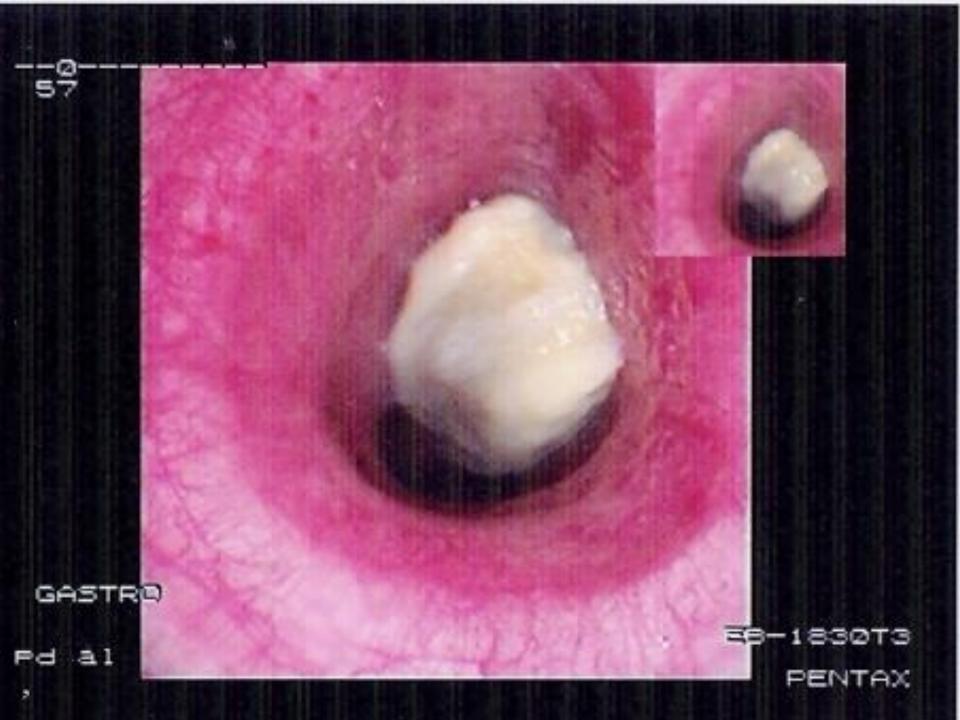


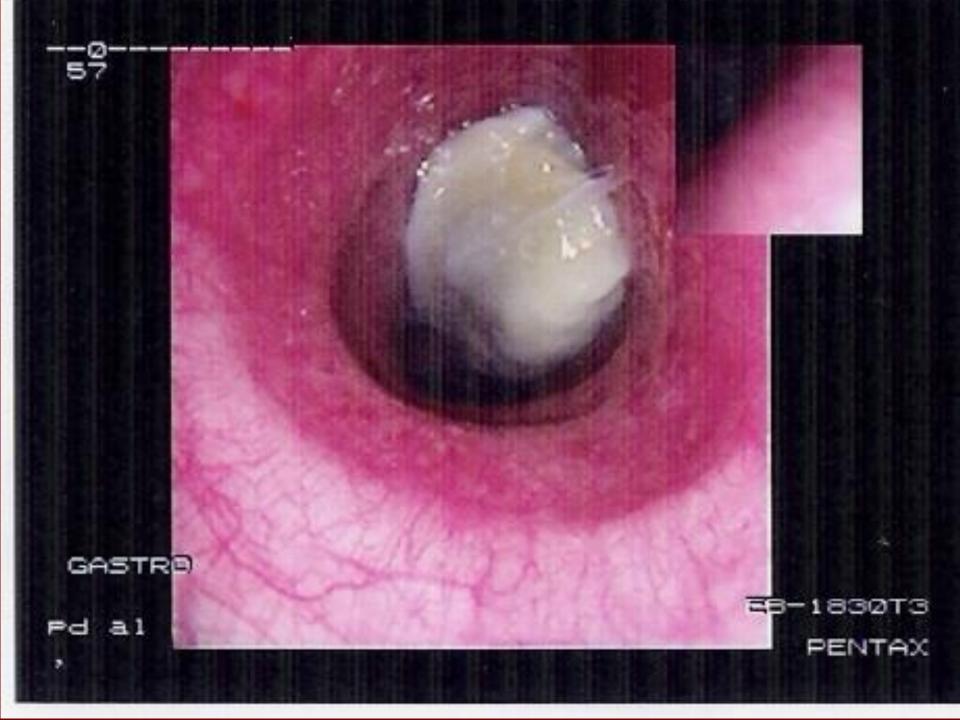
#### So I took her for bronchoscopy



# Bronchoscopy was amazingLet's see







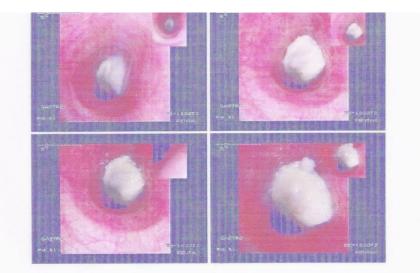


## Longing

The biopsy proved really not easy
The tips of biopsy forceps were "glued"
I managed to take few small pieces

#### **Bronchoscopy**

Dr AR Anani Consultant Pulmonary



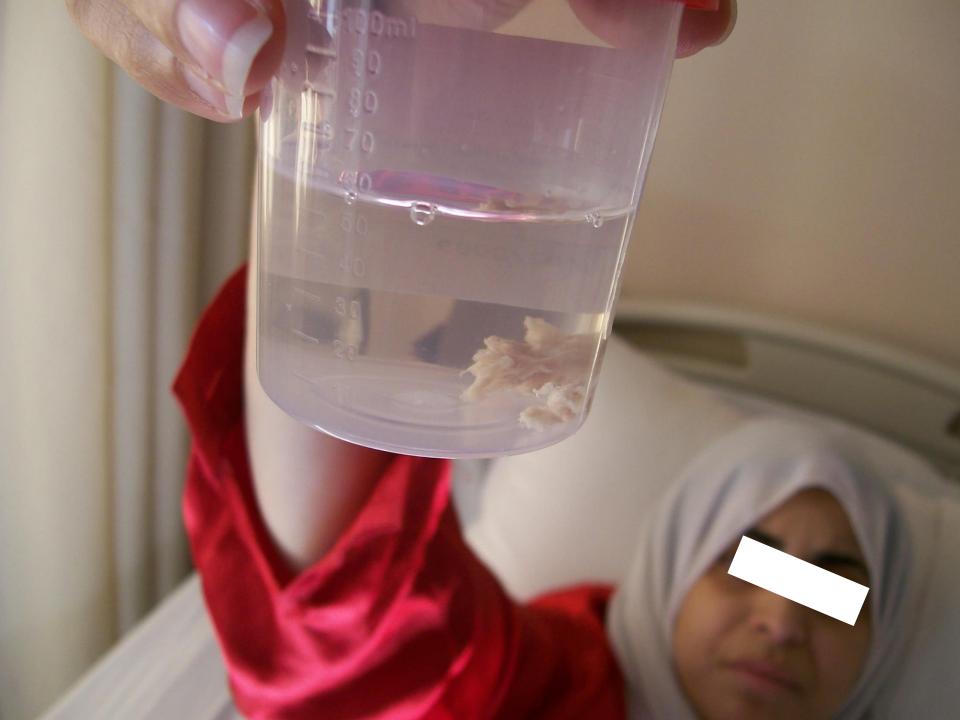
Vocal Cords, trachea, main carina, R lung: all normal.

The left lower lobe was almost totally blocked by very thick doughy material few cm in depth.

This is most likely aspirated food like rice but a thick necrotic tissue overlying a tumor cannot be ruled out.

Referred to thoracic surgeon.

Dr AR Anani Consultant Pulmonary





#### Biopsy report

Path. No.: 09-2-87

Origin of tissue: <u>Clinical history</u>: <u>Clinical diagnosis</u>: <u>Gross description</u>: Specimen received fixed & labeled with patient's name consist of Two linv soft tissue fragments. <u>Mic oscopic description</u>:

Sections show metaplastic stratified squamous epithelium with adjacent fibria, mixed inflammation mainly acute inflammatory cells with necrosis & keratin flakes. No malignancy.

Diagnosis: - Suggestive of reaction to foreign body.

## Longing

I called thoracic surgeon for help
We carried out rigid bronchoscopy
We managed to remove all the material from LLL

No other pathology like tumour

Pre operative diagnosis : CZ 205 e sea Post operative diagnosis : \_ Procedure perform Finding during surgery : \_ igid & love love by suction reps



# No one had a clue what that material wasI collected the material and sent it to





#### They called me and informed me



## Longing

I went back to patient, who was now symptom free, and asked her about choking with rice

## Longing

# I had longing for raw riceI choked with it frequently



#### Happy ending: she was back to work and happy



- A 48 year old lady, non-smoker, and had trouble with seasonal allergies from time to time.
- Twice had chest x rays and were normal
  Otherwise, she always enjoyed good health.

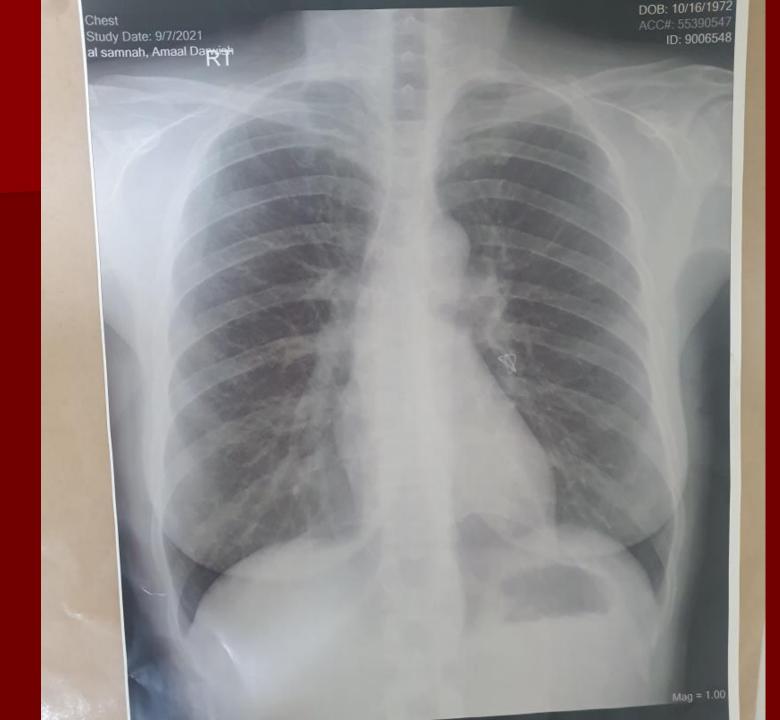
One week ago, she developed severe cough and purulent sputum.

- She tested herself for COVID-19, which was negative.
- She treated herself with antibiotics and usual medications.

As she had gotten worse, she visited A&E.
She had a chest x-ray, which revealed a total surprise.

- A foreign body was suspected.
- She had repeat x-rays to confirm the findings with clothes off. All radiologists at the Specialty Hospital confirmed that she had a metallic foreign body.
- She was referred to me.

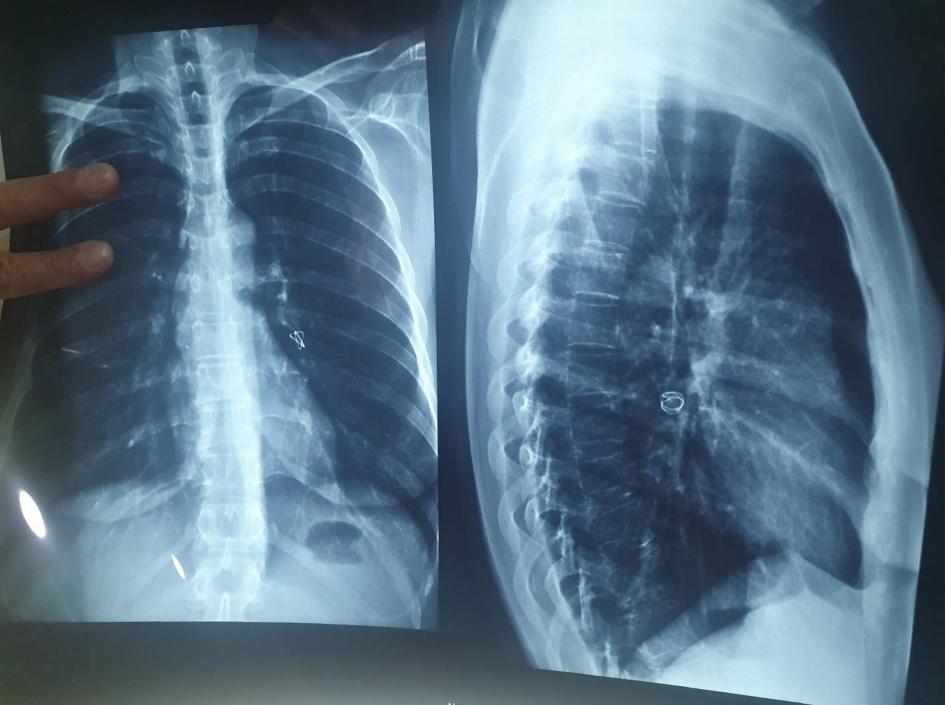
Let us look at the x-rays.











The patient denies any history of foreign body inhalation.
She cried " I did Not choke"

Definitely, she had a foreign body.
I took her for bronchoscopy, which was entirely normal.

- The patient is not happy and she is very anxious.
- I referred her to thoracic surgeon

 A very skilled thoracic surgeon, was consulted
 He did not trust my normal bronchoscopy

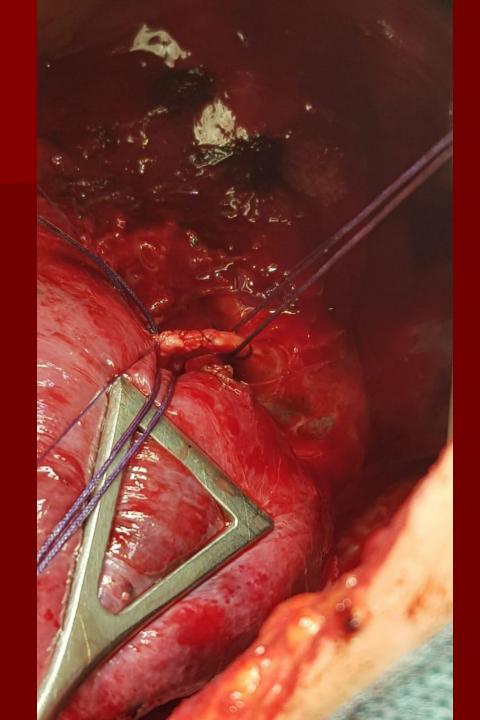
"I think this time you missed it"

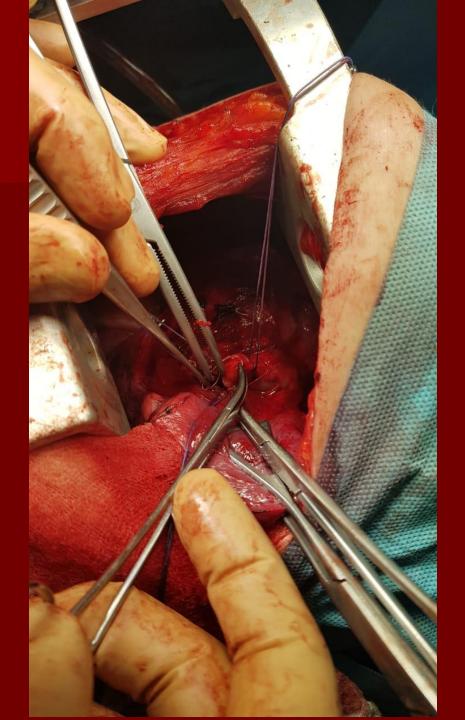
He repeated it
To his embarrassment he called me
"I searched all corners and found nothing"

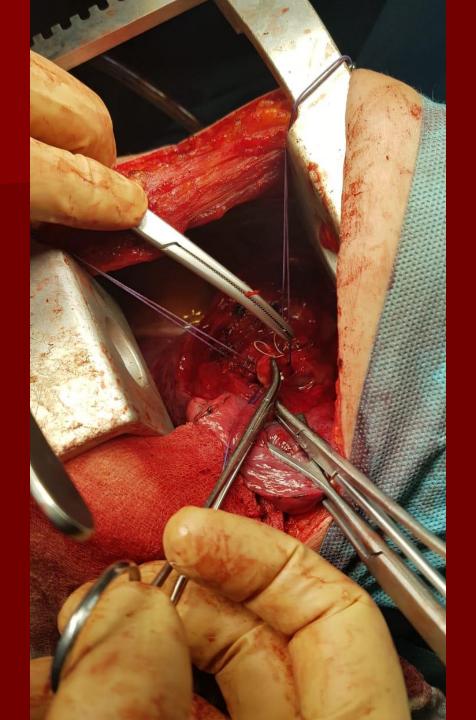
- As she continued to have chest pain and plenty of sputum, she demanded a definite solution.
- We agreed with surgeon to have thoracotomy.

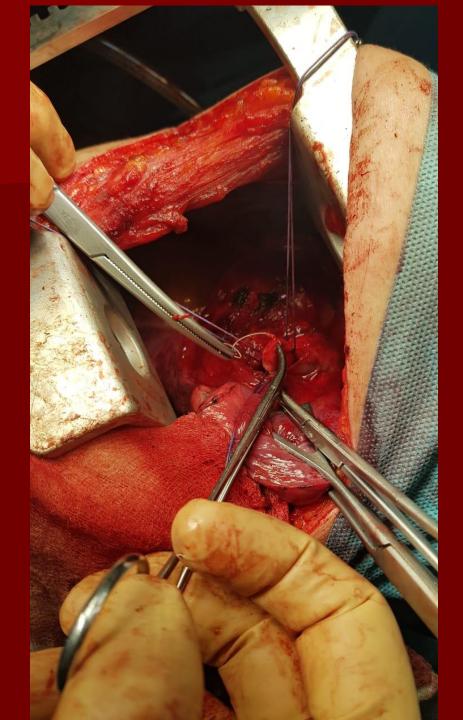
Careful inspection at thoracotomy revealed a metallic object in a pulmonary artery. With proper maneuvering, the metallic object was located and removed.
 It had already started to penetrate the adjacent left lower lobe bronchus.

#### Let's see some operative photos











Post operative course was excellent





Let us look at this metallic object:







Now, you see this objectRemember, did you choke with it?



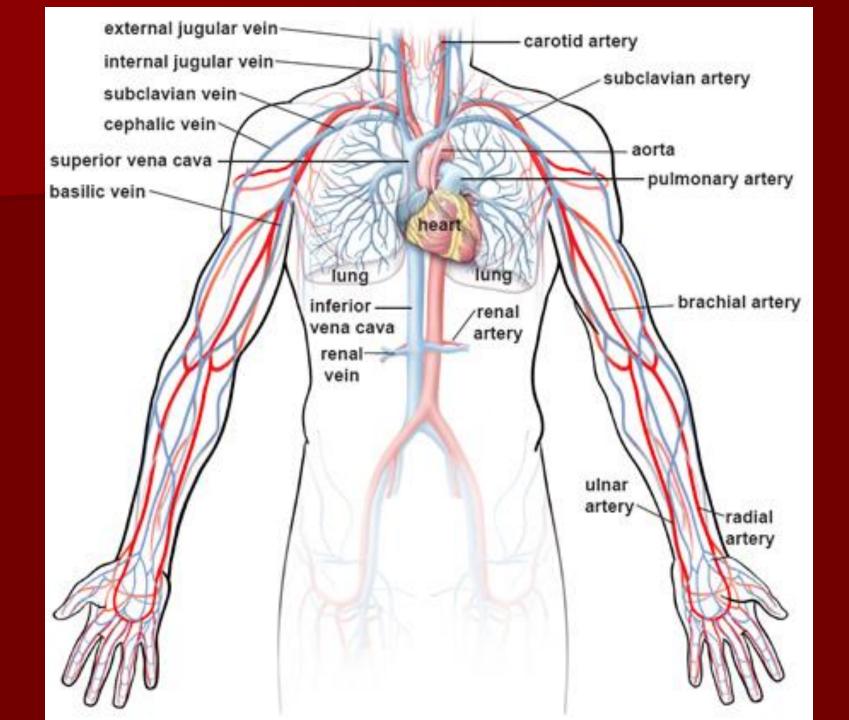


- The metallic object was identified by theatre nurse as a surgical wire used by vascular surgeons.
- Investigation with the lady about past medical history proved negative.

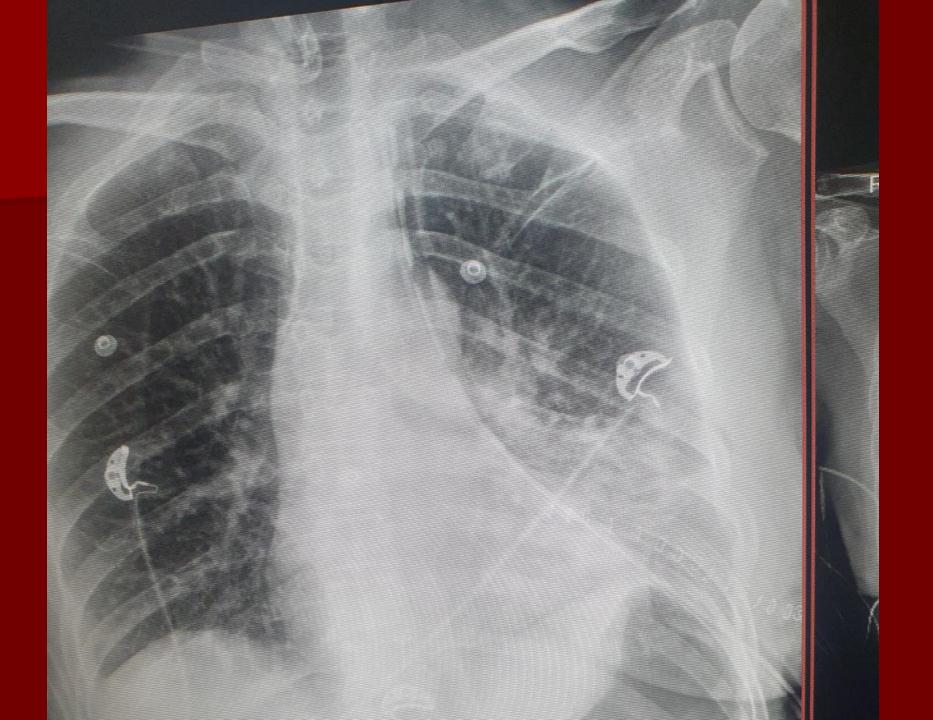
- Interrogating the husband, he remembered that eight years ago, she had pelvic surgery.
- She had pain in her legs, attributed to varicose veins in the uterus.
- She underwent surgery whereby those varicose veins were clamped by wires.

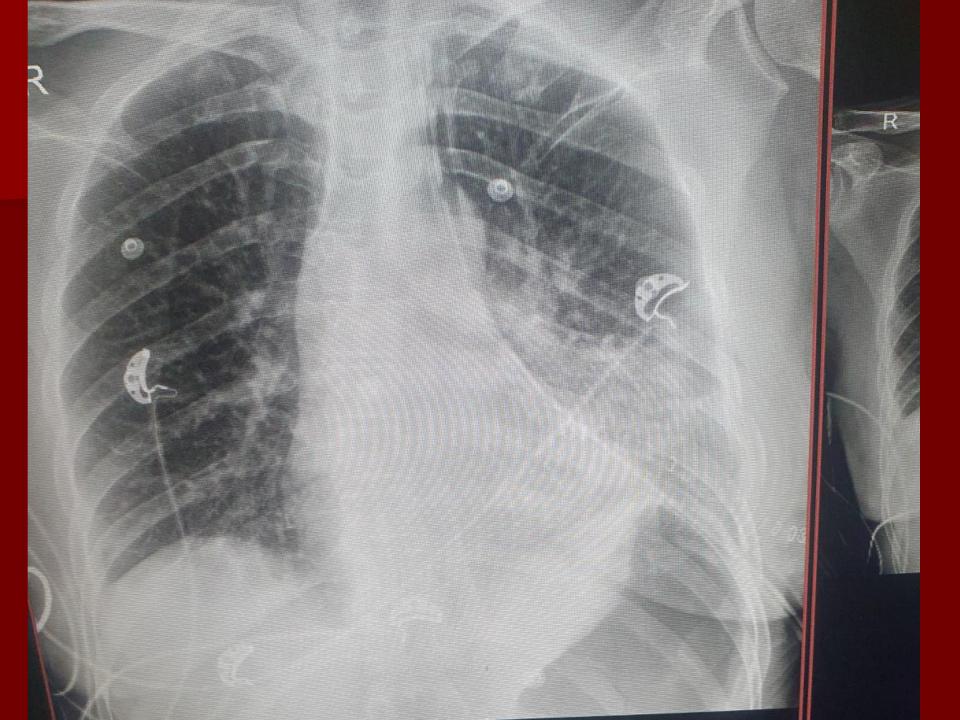
The vascular surgeon confirmed he used similar wires

We concluded that the wire penetrated through and went with circulation to the heart, then to the lungs, where it started to cause trouble.



These are her x-rays post-operation, which are wireless, with evidence of lung contusion on the left side.





Now she is totally symptom free except for pain in the wound









Troubled with eczema as a baby
 Early school years: eczema resolved but trouble with rhinitis started
 Aged 12: 2 nasal polyp operations



- At age 14 he started to have recurrent skin infections like boils and cellulitis
- In addition he had difficult asthma symptoms
- His nose became big with widening of the base and bridge
- He had also pneumonia requiring hospital admission

Hct. RBC WBC MCV MCH MCHC Platelets Neutrophils Lymphocytes Monocytes

Eosinophils

Erythrocyte Sedimentation Rate (ESR)

42 % 5.6 x10^6/µL 4.2 x103/µL 74 fL 25 pg 34 g/dL 176 x10<sup>3</sup>/µL 44 % 35 % 6 % 15 % 5 mm/1 hr







#### **ARS : University Years**

Continued to have rhinitis and asthma

Frequently had skin and chest infections requiring prolonged courses of antibiotics

# ARS: Working Man

# He came to see me for better asthma control Examination was remarkable





















## **ARS : Sudden Deterioration**

Admitted to hospital with severe cough and shortness of breath
No fever
Signs of consolidation R base and wheezy

#### Investigations

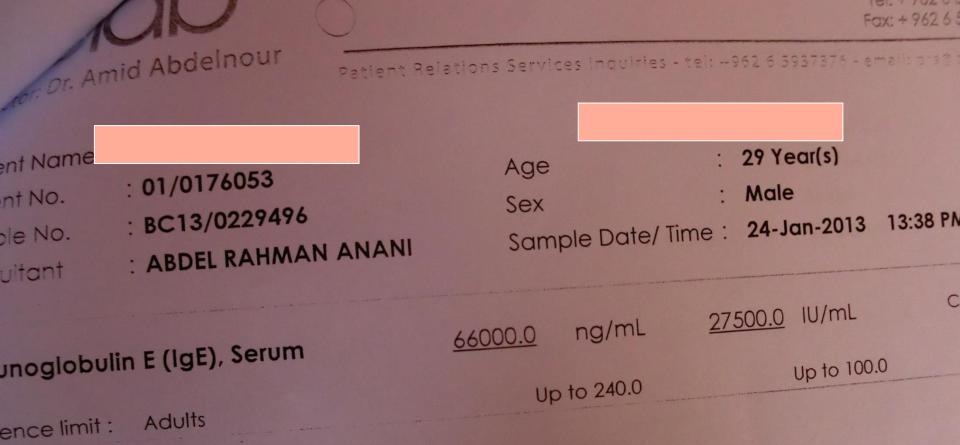


| Haemoglobin  | . 1  |       |        |         |                        | <u>Reference limit</u> |
|--------------|------|-------|--------|---------|------------------------|------------------------|
|              | : 14 |       | g/L    |         |                        | 138 - 170              |
| Haematocrit  | : 4  | 1.9   | %      |         |                        | 42.0 - 49.0            |
| RBC          | : 5. | .61   | x10^12 | 2/L     |                        | 4.7 - 6.1              |
| MCV          | : 74 | 4.7   | fL     |         |                        | 80.0 - 99.0            |
| МСН          | : 2  | 5.7   | pg     |         |                        | 27.0 - 32.0            |
| МСНС         | : 34 | 4.4   | g/dL   |         |                        | 32.0 - 36.0            |
| RDW          | : 16 | 6.3   | %      |         |                        | 11.6 - 15.0            |
| Platelets    | : 16 | 54    | x10^9/ | Ľ       |                        | 150 - 450              |
| MPV          | : 10 | ).6   | fL     |         |                        | 7.2 - 11.7             |
| WBC          | : 18 | 3.370 | x10^9/ | Ľ       |                        | 4.0 - 11.0             |
| Differential |      |       |        |         | <u>Reference limit</u> |                        |
| Neutrophils  | : 19 | %     | 3.490  | x10^9/L | 1.800 - 7.500          |                        |
| Lymphocytes  | : 9  | %     | 1.653  | x10^9/L | 1.200 - 4.000          |                        |
| Monocytes    | : 4  | %     | 0.735  | x10^9/L | 0.200 - 1.000          |                        |
| Eosinophils  | : 63 | 3 %   | 11.573 | x10^9/L | 0.040 - 0.500          |                        |
| Basophils    | :    | %     |        | x10^9/L | 0.015 - 0.100          |                        |
| Other        | : 5  | %     |        | x10^9/L | 0.000 - 0.000          |                        |

|                  |          | 1 1             | 6.3     | 8%     |                     |       |  |  |
|------------------|----------|-----------------|---------|--------|---------------------|-------|--|--|
| Platelets<br>MPV |          | 1 164<br>1 10,6 |         |        | 78<br>×10/9/L<br>fL |       |  |  |
| WBC              | : 18.370 |                 | x10^9/L |        |                     |       |  |  |
| Differential     |          |                 |         |        | /-                  |       |  |  |
| Neutrophils      | :        | 19              | %       | 3.490  | x10^9/L             | Ref   |  |  |
| Lymphocytes      | :        | 9               | %       | 1.653  |                     | 1.80  |  |  |
| Monocytes        | :        | 4               | %       | 0.735  | x10/9/L             | 0.200 |  |  |
| Eosinophils      | :        | 63              | %       | 11.573 | x10/9/L             | 0.040 |  |  |
| Basophils        | :        |                 | %       |        | x10^9/L             | 0.015 |  |  |
| Other            | :        | 5               | %       | 0.919  | x10^9/L             | 0.000 |  |  |



### The conclusion: Considering all the facts



| OF UI. AINO    | Abdemour                   | Patient Relatio |             |            |              |            | Fax: + 962 6 592 67 |
|----------------|----------------------------|-----------------|-------------|------------|--------------|------------|---------------------|
| ple No. :      | 01/0176053<br>BC13/0229880 | Patient Relatio |             |            |              |            |                     |
| sultant :      |                            |                 | Sar         | nple Date/ | Time : 26-   | Jan-2013   | 14:45 PM            |
| unoglobulin    | E (IgE), Serum             | <u>674</u>      | <u>64.0</u> | ng/mL      | <u>28110</u> | IU/mL      | Confirme            |
| ious Result    | 24-Jan-2013                | 27500.0 IU/mL   |             |            |              |            |                     |
| erence limit : | Adults                     |                 | U           | o to 240.0 |              | Up to 100. | .0                  |
|                |                            |                 |             |            |              |            |                     |



# Hyper IgE Syndrome (HIES) : JOB (Job ,II,7)

### So went Satan forth and smote Job with sore boils from the sole of his foot unto his crown

# **HIES : Facial Features**

- Patients resemble each other more than they resemble their family members
- Broad nasal base and bridge
- Frontal bossing
- Thickening of soft tissues of face, ear and nose "coarse faces"
- Two rows of teeth due to retaining of primary teeth

## **HIES : Manifestations**

Severe Allergic Manifestations Eczema , Rhinitis , Asthma

Recurrent Skin , Sinus and Pulmonary infections

High IgE levels 100 folds of upper normal limit



## 5 Year Messi

- Healthy boy who spends all time playing football
- Recently parents noticed excessive coughing during playing
- No fever , No wheeze , No night awakening
- Extremely well otherwise

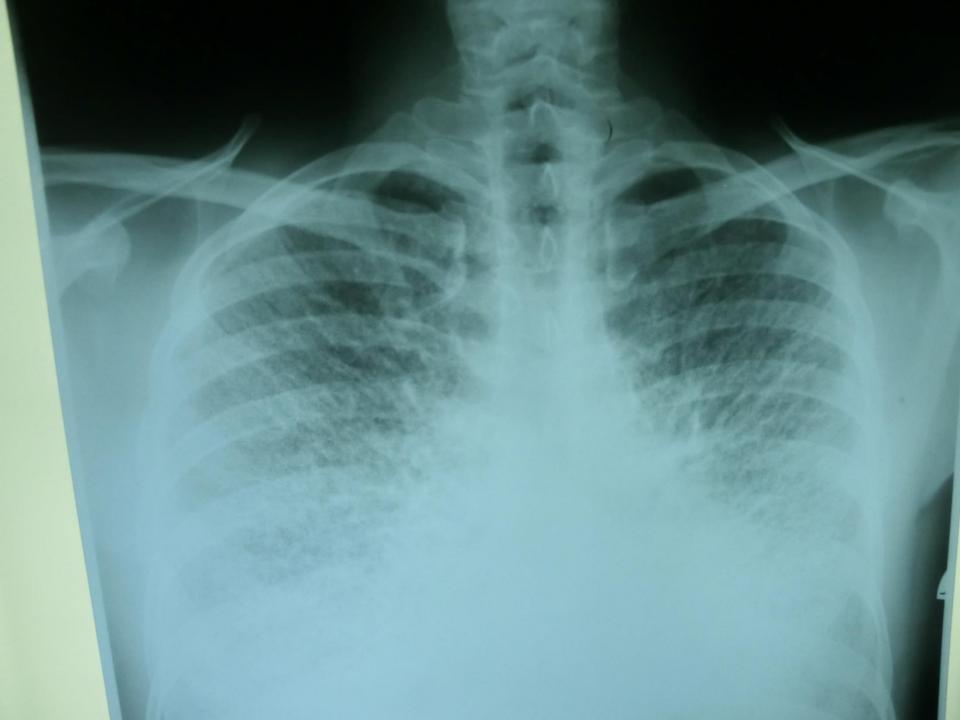
# 5 Year Messi : GP consultation

- Exercise induced asthma diagnosis was made
- Treatment with inhaled steroid and montelukast was of no benefit
- Parents noticed that he frequently rested during games
- Otherwise , he was very well

5 Year Messi : Pediatric Pulmonologist

Messi was banned from exercise and was miserable

- GP referred him to Chest X Ray
- Pediatrician admitted him to hospital



# 5 Year Messi : Inpatient

- A diagnosis of massive pneumonia was made !!!!!
- Cocktail of i.v antibiotics for a week and continued on oral for another week
- A chest x ray thereafter showed no change
- The pediatrician suggested treatment for TB and family not convinced



# 5 Year Messi In my Clinic

Clinical / Radiological mismatch
 Q) What diagnosis ?

#### Asthma NO

### Infection NO

#### T B NO



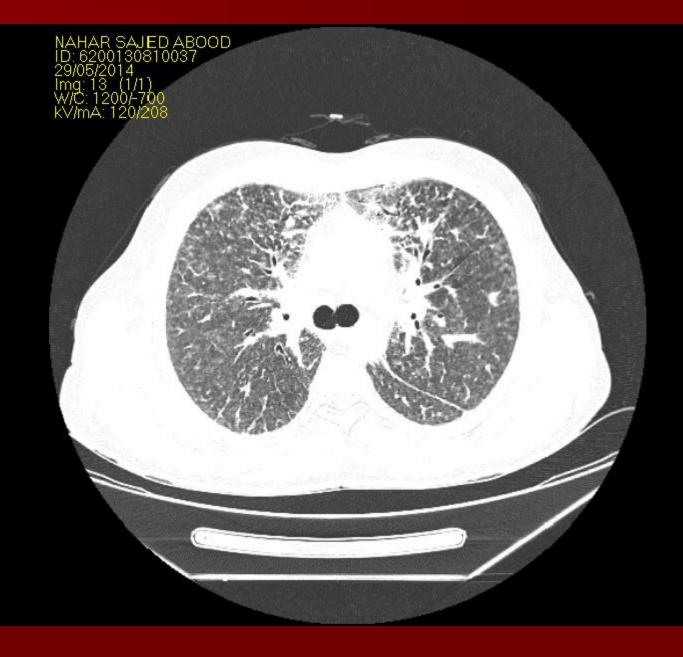
## 5 Year Messi : Caring Father

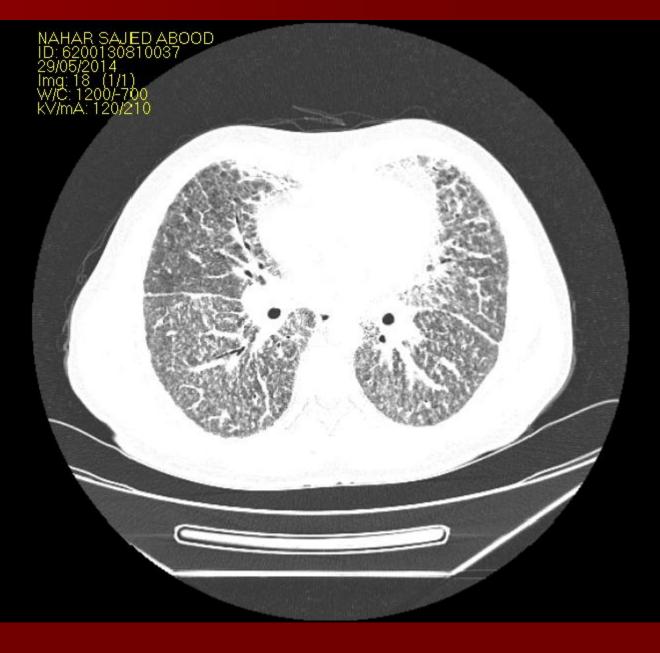
 I explained to father that Messi has special problem and needed investigations starting with chest C T

As father was also coughing I asked him to do chest x ray " just in case"



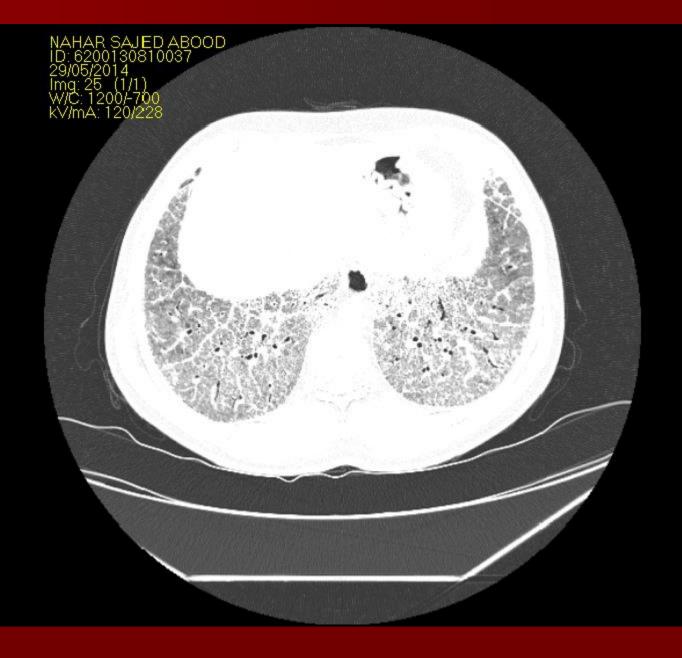












# Q) Diagnosis?



# 5 Year Messi : Father X Ray



# 5 Year Messi : Next Plan

## Lung Biopsy obtained from father

| Cytology no: S14/ 9301            | القسم: |
|-----------------------------------|--------|
| SDecime \$14/ 9301                |        |
| Specimen: Rt lung bx              |        |
| Clinical information:             |        |
| 38 y/o male case of lung fibrosis |        |
| and the case of lung fibrosis     |        |

### Microscopic Description:

The sections taken from the lung tissue reveal diffusely dilated alveolar s contain calcified laminated structures A mild interstitial lymphoid cell infiltrate and mild focal interstitial f with no evidence of active formation of fibroblastic plugs

#### Final Diagnosis:

RT lung, wedge biopsy:

- Pulmonary alveolar microlithiasis 🦸
- No evidence of granulomas or malignancy

cytotechnologist signiture: specialist signiture:

result date

Clinical information: 38 y/o male case of lung fibrosis

## Microscopic Description:

The sections taken from the lung tissue reveal diffusely d contain calcified laminated structures

A mild interstitial lymphoid cell infiltrate and mild foca with no evidence of active formation of fibroblastic plug

#### Final Diagnosis:

RT lung, wedge biopsy:
 - Pulmonary alveolar microlithiasis /
 - No evidence of granulomas or malignancy

spec sign

result date

cytotechnologist signiture:

## **Pulmonary alveolar microlithiasis**

## Clinical presentation

Often discovered incidentally on a chest radiograph. The radiographic features are out of proportion to clinical symptoms, It is the only disease that causes extensive alveolar shadow with minimal symptoms



# 5 Year Messi : The Plan

## Lung Transplant

What is unusual ?

# Difficult Cases may be

### "Difficult roads often leads to beautiful destinations. "

