THE SIX MILLION DOLLAR MAN

Alfred Chan

HKSCCM grand-round 16th Jan 2017

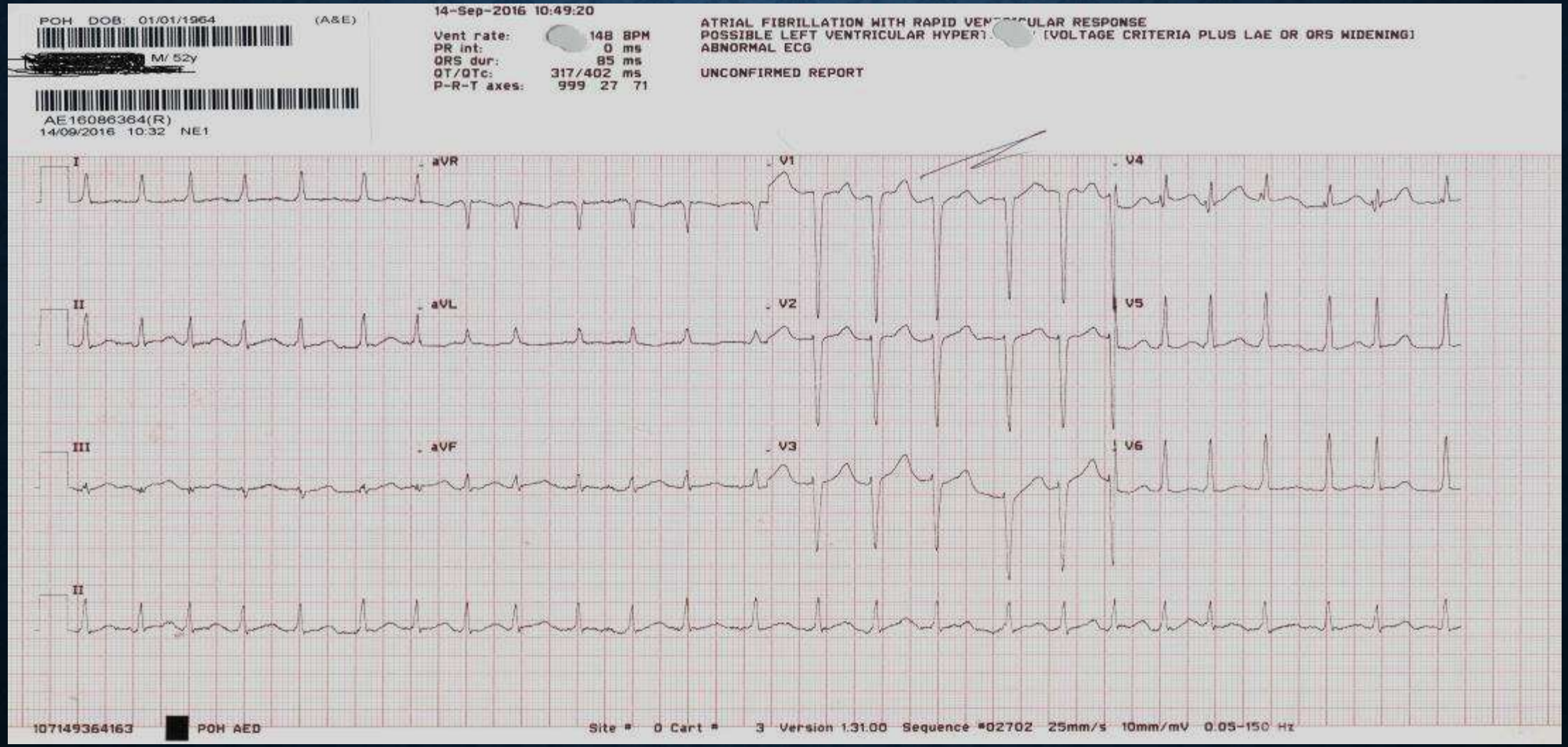
52-YEAR-OLD MAN, PAKISTAN

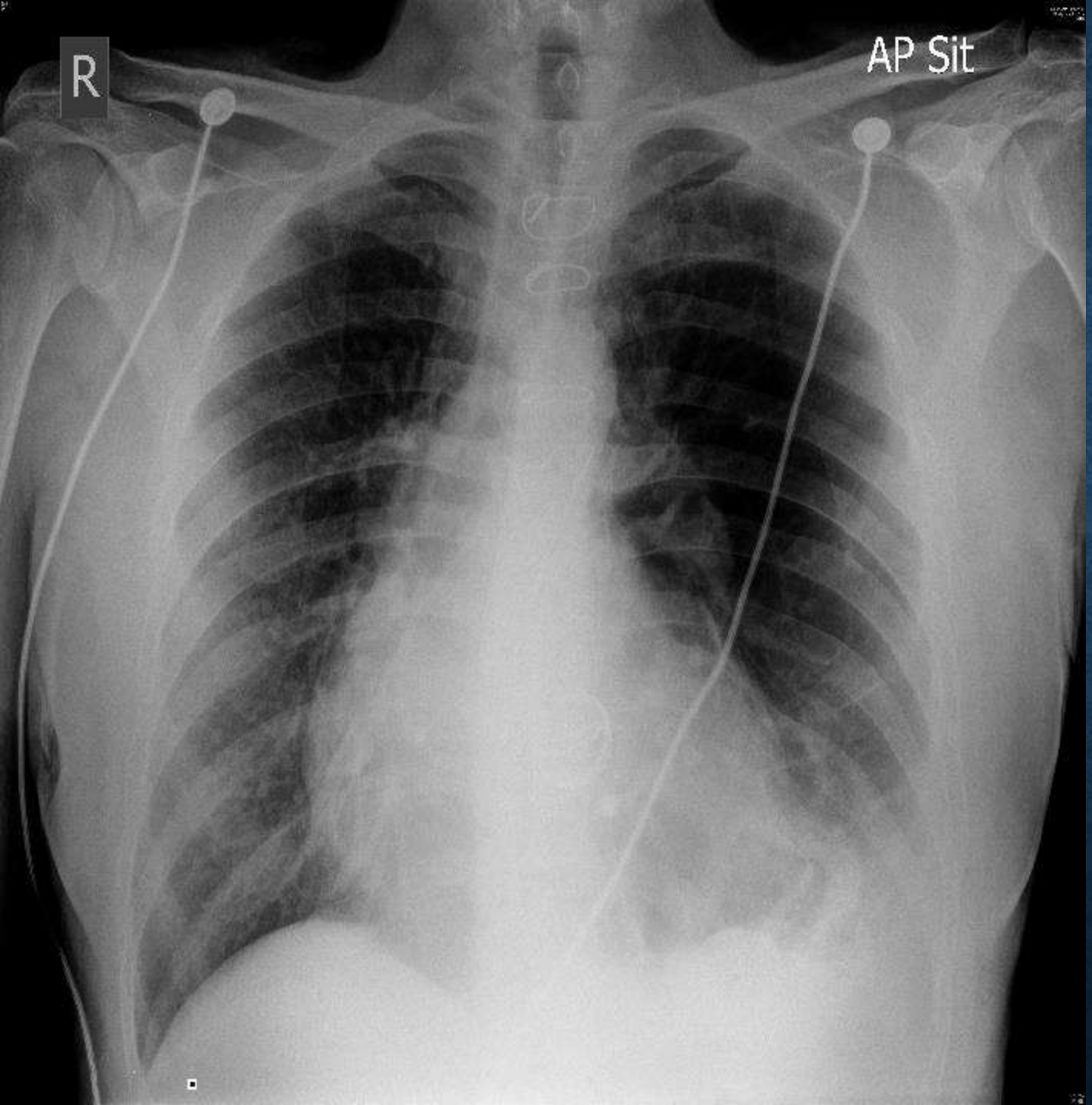
- Non-smoker; non-drinker
- Business man; resident of Pakistan
- Travel to HK for purchase of goods to be sale in own country
- Past medical history
  - COPD on bronchodilator
  - Known MS with PTMC May 2015 and then MVR Nov 2015
  - Echo June 2016: normal prosthetic valve function, LVEF 45%
  - On Warfarin/ Amiodarone/ Aspirin/ Lasoride (Lasix + Amiloride)

ATTENDED AED ON 14TH SEPT

- C/O: cough for 3 days then severe SOB today + chest pain
- Deny URI symptoms
- Vital signs at triage:
  - GCS 15/15 orientated
  - Pulse 155 irregular; BP 128/67
  - SaO2 at 78% on room air; Resp rate 18/ min
  - Appeared cyanotic. No Club/ LN. Short systolic murmur at apex.
  - SaO2 at 95% with NRM in "R" room

ECG


POH DOB: 01/01/1964 (A&E)
[redacted] M/ 52y
AE16086364(R)
14/09/2016 10:32 NE1
14-Sep-2016 10:49:20
Vent rate: [redacted] 148 BPM
PR int: 0 ms
QRS dur: 85 ms
QT/QTc: 317/402 ms
P-R-T axes: 999 27 71
ATRIAL FIBRILLATION WITH RAPID VENTRICULAR RESPONSE
POSSIBLE LEFT VENTRICULAR HYPER[redacted] [VOLTAGE CRITERIA PLUS LAE OR QRS WIDENING]
ABNORMAL ECG
UNCONFIRMED REPORT
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II
107149364163
POH AED
Site # 0 Cart # 3 Version 1.31.00 Sequence #02702 25mm/s 10mm/mV 0.05-150 Hz



CXR at attendance

LLZ Haziness

? Pleural effusion

WORKING DX: FAST AF CAUSING CHF

- Amiodarone 150mg in 100ml D5 over ½ hour IV
- vBG: pH 7.38; pCO2 5.73; pO2 2.67; BE -0.4

- CCU assessment:
  - ECG fast AF no definite ischemic change
  - V-scan → LVEF 40%; IVC collapsible on inspiration; MV no leaking
  - Impression → pneumonia with chronic impaired LV function
  - Advise to intubate patient then consult ICU

QUESTION 1

Would you intubate the patient?

POST-INTUBATION AT AED

- Connect to ***Oxylog 3000***, TV 400ml; PEEP 4; RR 12; O2 at 100%
- Peak pressure 24; SaO2 static at 85%
- ABG or vBG: pH 7.18; pCO2 8.2; pO2 6.6; BE -6

- ICU assessment:
  - Lung pathology cannot explain hypoxemia
  - Acute chest pain + SOB with desaturation → ? Pulmonary embolism
  - Request CT pulmonary angiogram but declined by radiologist

QUESTION 2

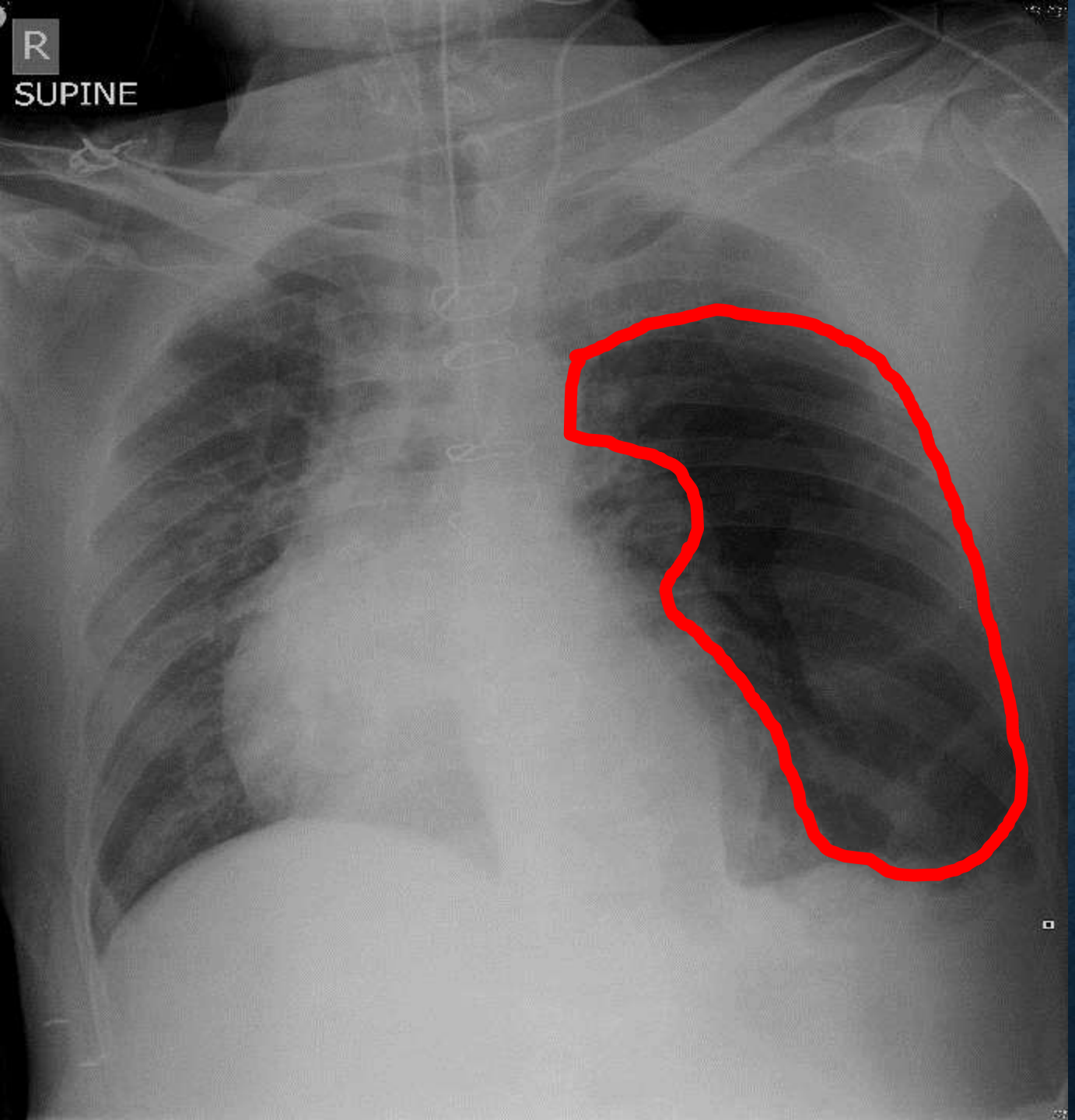
Would you admit this patient to ICU?

With what working diagnosis?

MANAGEMENT AT ICU

- Working diagnosis: CAP

- Air-born precaution
- NPA and ET aspirate for a panel of PCR study towards common respiratory tract infection pathogens
- Empirical IV Rocephin/ Doxycycline/ Tamiflu
- Deep sedation with Midazolam/ Morphine infusion
- Ventilator setting as protective lung strategy for ARDS
- CXR after intubation: no pneumothorax/ significant mass effect



CXR After intubation

Anything new ?

PROGRESS AFTER ICU ADMISSION (1)

- Investigation
  - Co-Hb 1.5%; Met-Hb <1%
  - Troponin 19.1 then 32 (ref < 34) at 6th hour
  - INR 1.7
  - Hb 18.2; Hct 0.629; MCV 74.5; WCC 9.4
  - Na 131; K 4.1; Urea 4.4; Creat 67; LFT normal
  - NPA flu A & B rapid antigen negative
  - Sputum AFB smear negative; Urine Legionella Antigen Negative
  - ABG (PEEP 8 FiO2 1.0) → pH 7.19; pCO2 8.43; pO2 7.18; BE -6; HCO3 23.5

PROGRESS AFTER ICU ADMISSION (2)

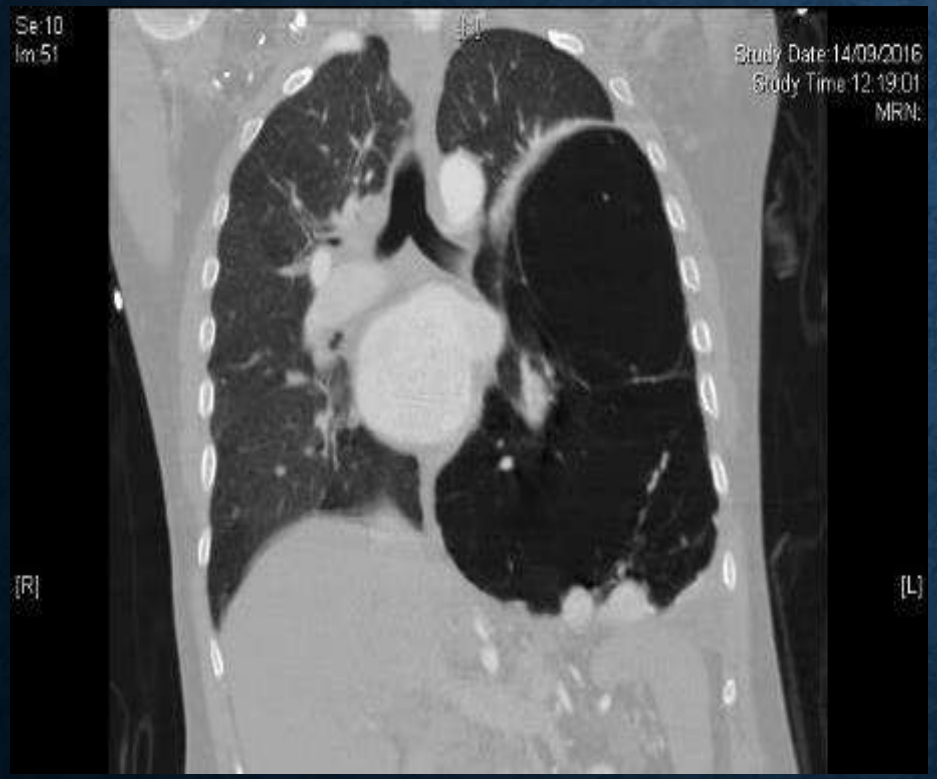
- Ventilator parameters
  - SIMV (PC) + PS mode
  - PEEP 8; PC 14 PS 14
  - RR 14; Insp time 1.7 second; FiO2 at 100%
- Physiology variables
  - Tidal volume 480ml; Minute Ventilation 6.8L
  - Peak airway pressure 22; Plateau pressure 18
  - Auto-PEEP 0

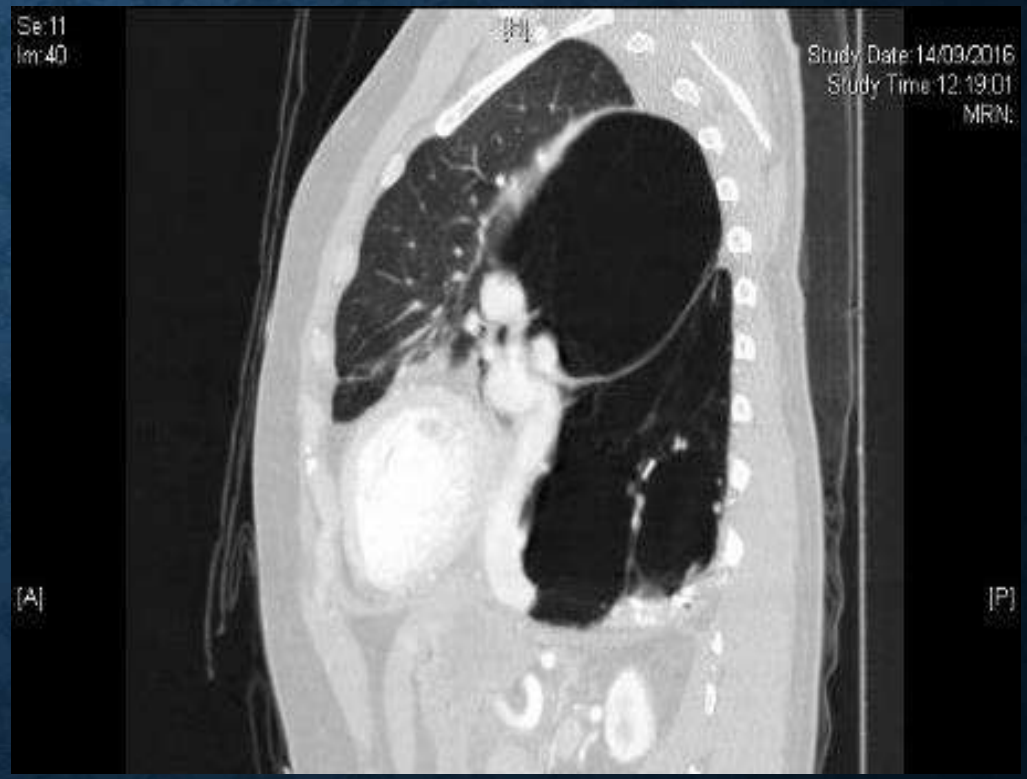
INTERVAL CHANGE IN ICU

- Further worsening of desaturation after ICU admission
- Developing hypotension/ obliguria requiring vasopressor
- Persistent AF 150-160/ min relying on amiodarone infusion
- Poor urine despite bolus Lasix 40 + 40mg within 2 hours

- Intervention:
  - Adjustment of ventilator setting with PEEP increment
  - CT Aortogram for ?dissection: done but image/ report pending

QUESTION 3: IS THE PATHOLOGY THERE?





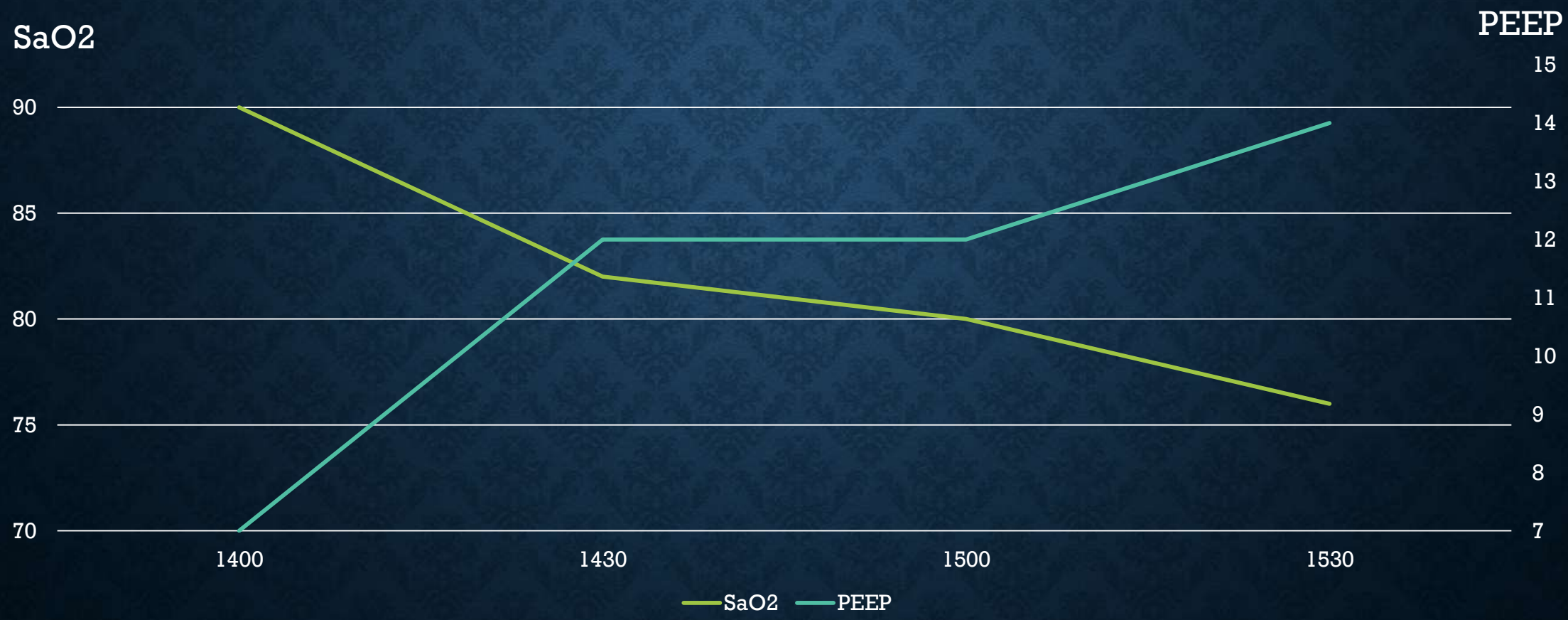
Loculated pneumothorax vs. bullae?

CLASSICAL CONTRAINDICATIONS TO PEEP

- [redacted]
- [redacted]
- [redacted]
- [redacted] of increased afterload

- Tension Pneumothorax - it will get worse

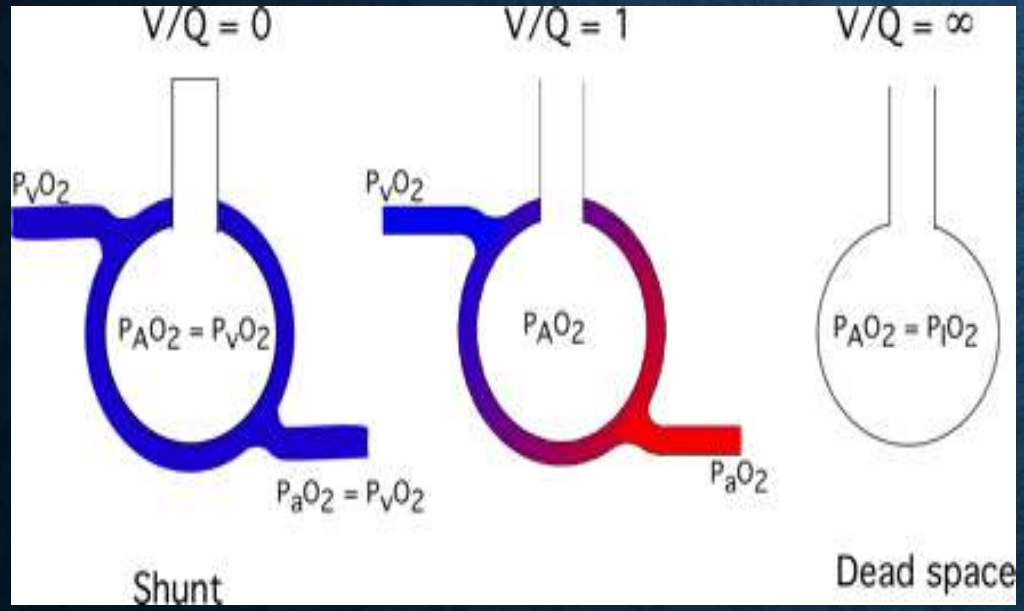
WHILE THINKING, NON-STOP WORSENING

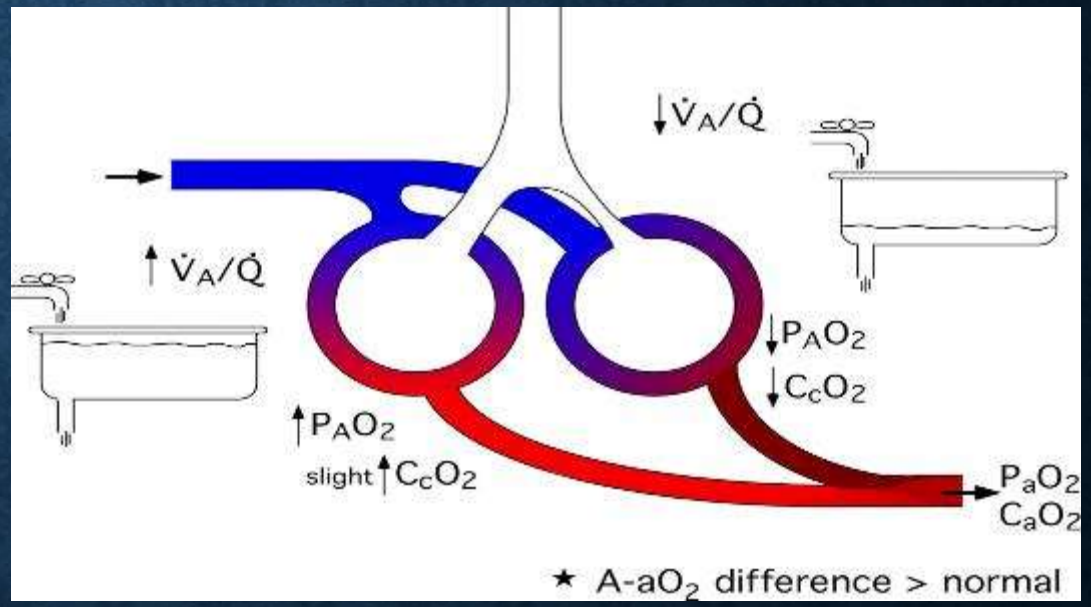


QUESTION 4

Why deteriorated with higher PEEP?

PHYSIOLOGY DISTURBANCE IN ARDS

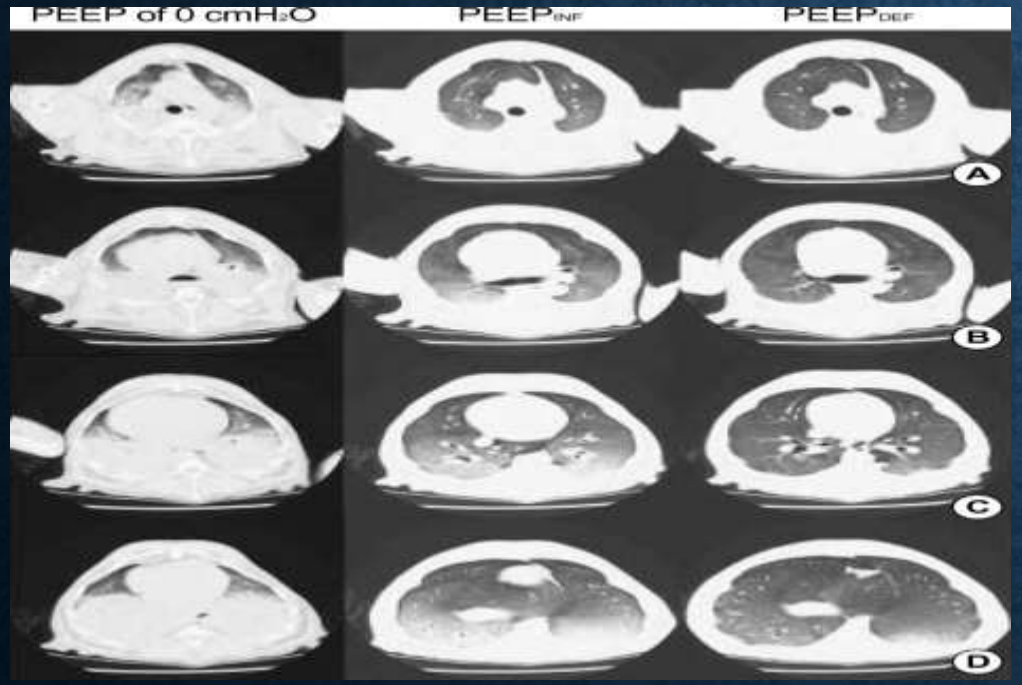




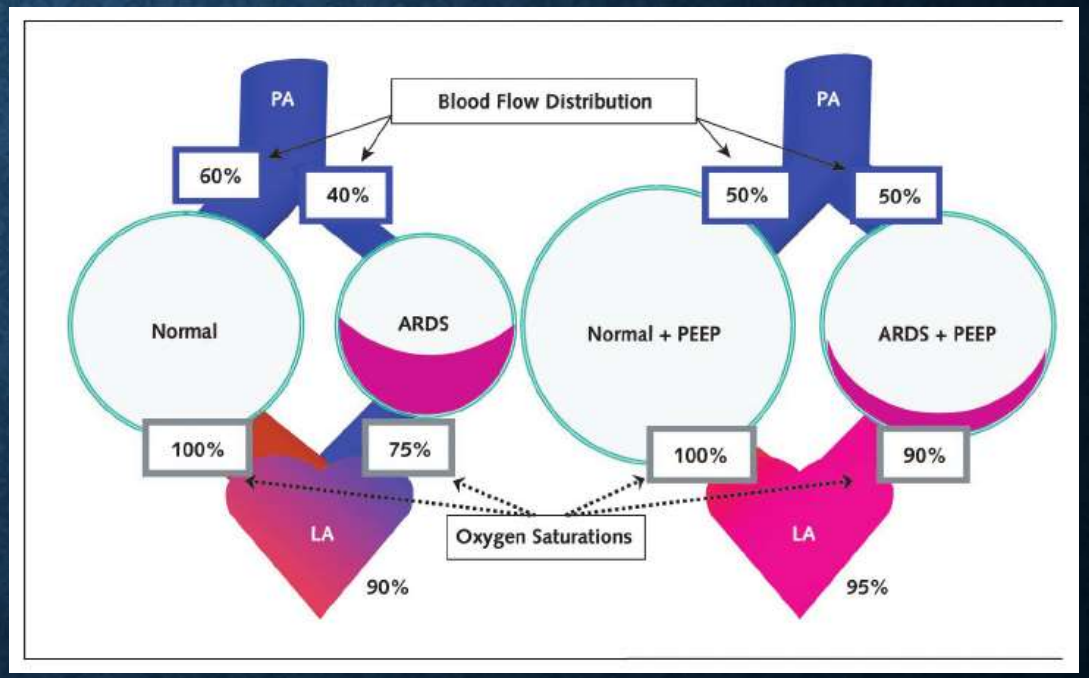
- Increased shunt and physiological dead space

- Mixing of hypoxemic blood with oxygenated blood

HOW PEEP MAY HELP

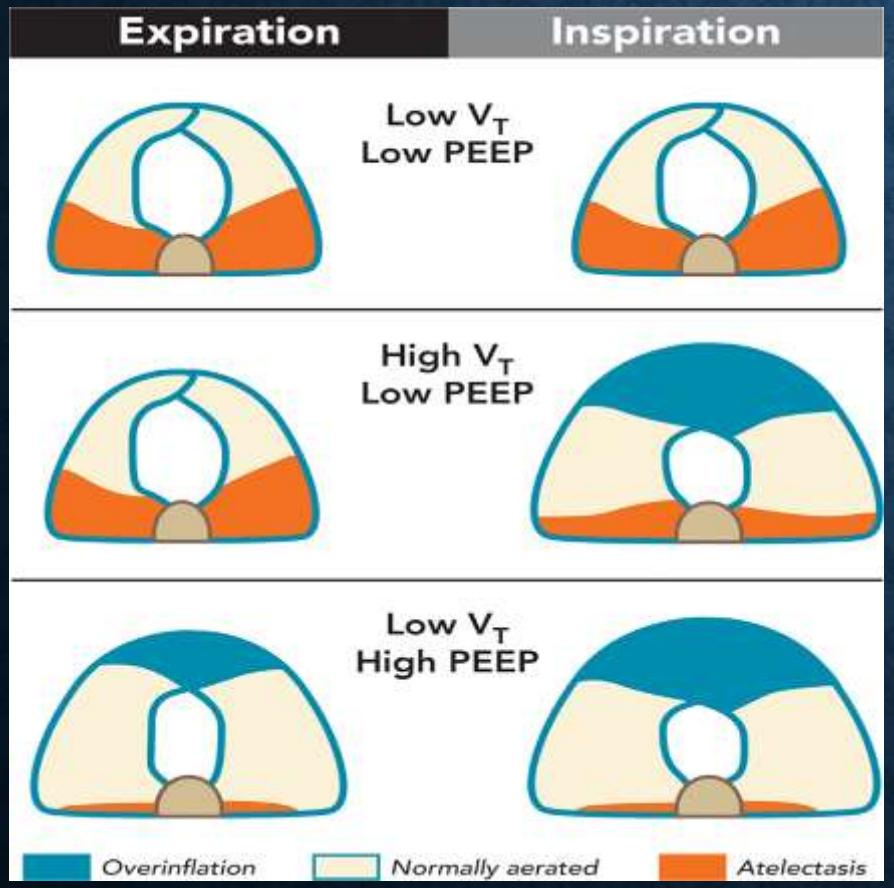


- Recruitment of atelectatic alveolar units for gaseous exchange



- Ventilation-perfusion matching improved via vasodilation at less hypoxic units

COMBINING HIGH PEEP AND LOW TV

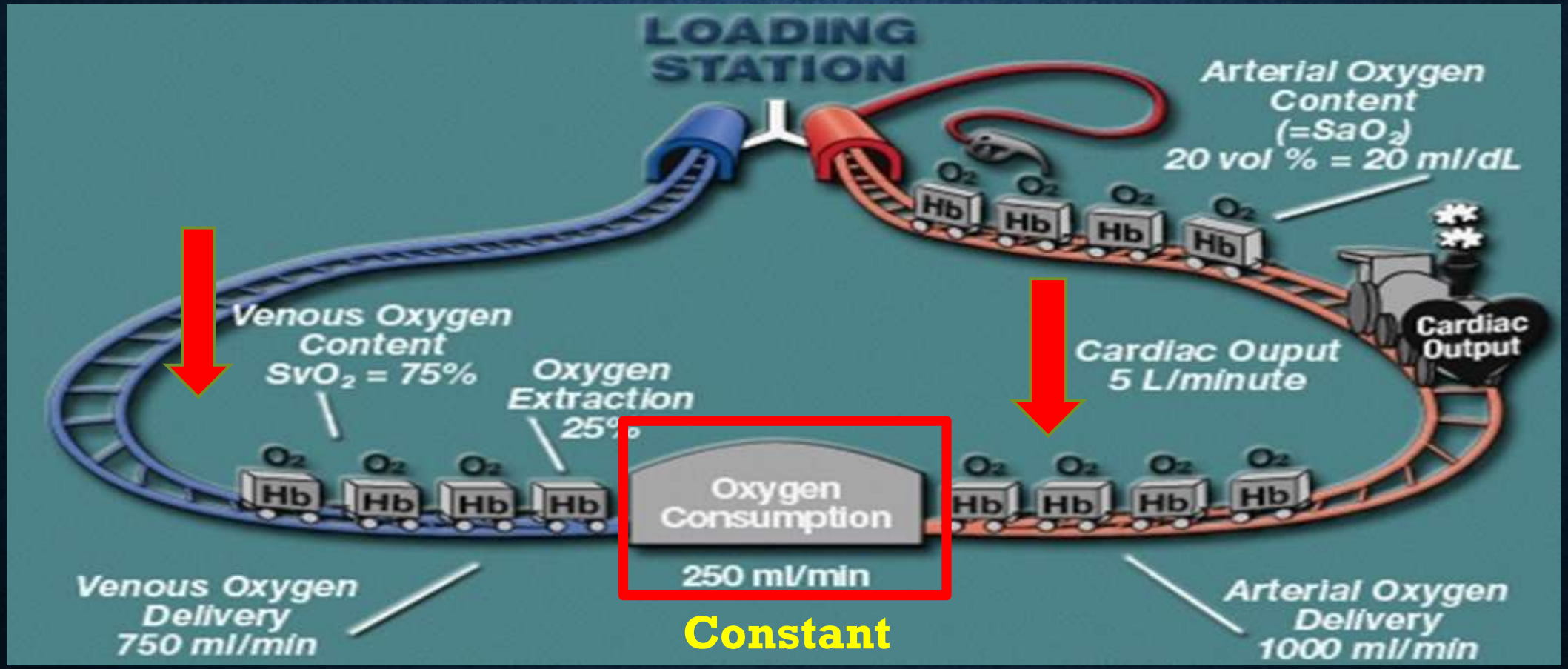


- PEEP can minimize atelectasis throughout the breathing cycle
- Low tidal volume causes less over-inflation of alveoli and hence less barotrauma, from which less cytokines may be produced

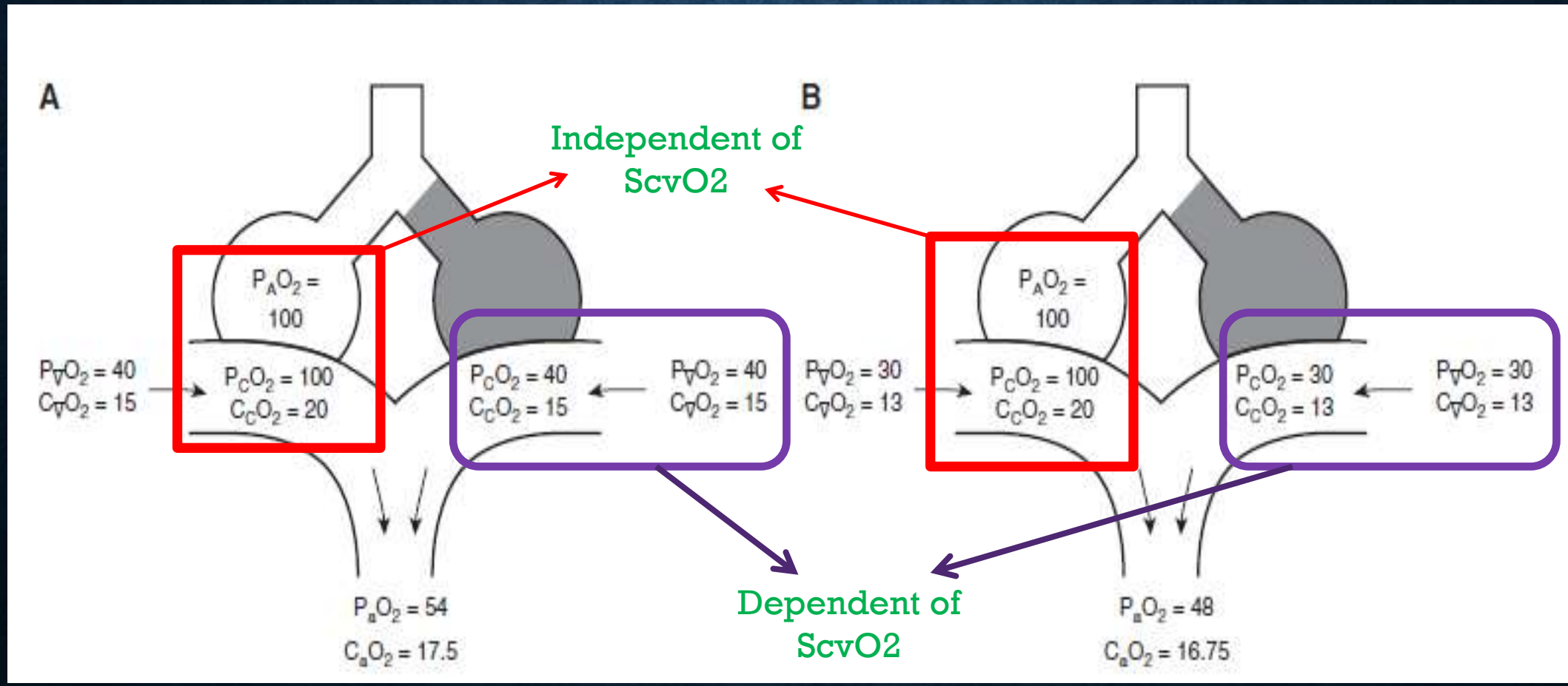
PEEP MAY WORSEN SAO2 IF ……

- **Occasion of barotrauma**
- **Tissue hypoxia due to low LV cardiac output**
  - PEEP-> ↑intrathoracic pressure ->↑ RV preload + RV afterload -> ↓CO -> O2 delivery -> ↑O2 extraction ->↓ mixed venous oxygenation
- **Patient with pre-existing R. heart failure / cor pulmonale**
  - Worsening of RV and RV dilatation -> ↑compression on LV (ventricular interdependence)
- **Patient with intracardiac shunt** e.g. ASD, patent foramen ovale
  - ↑Increase in R. heart pressure ->↑ R to L. shunt
- **Patient with focal lung pathology** e.g. AV malformation, telengiectasia, pneumonia
  - Increase PEEP -> overdistension of normal lung unit -> ↑regional pulmonary vascular resistance -> ↑ diversion of blood to shunt area -> worsening of V/Q and↑ shunt

LOW CO → MORE TISSUE O2 EXTRACTION

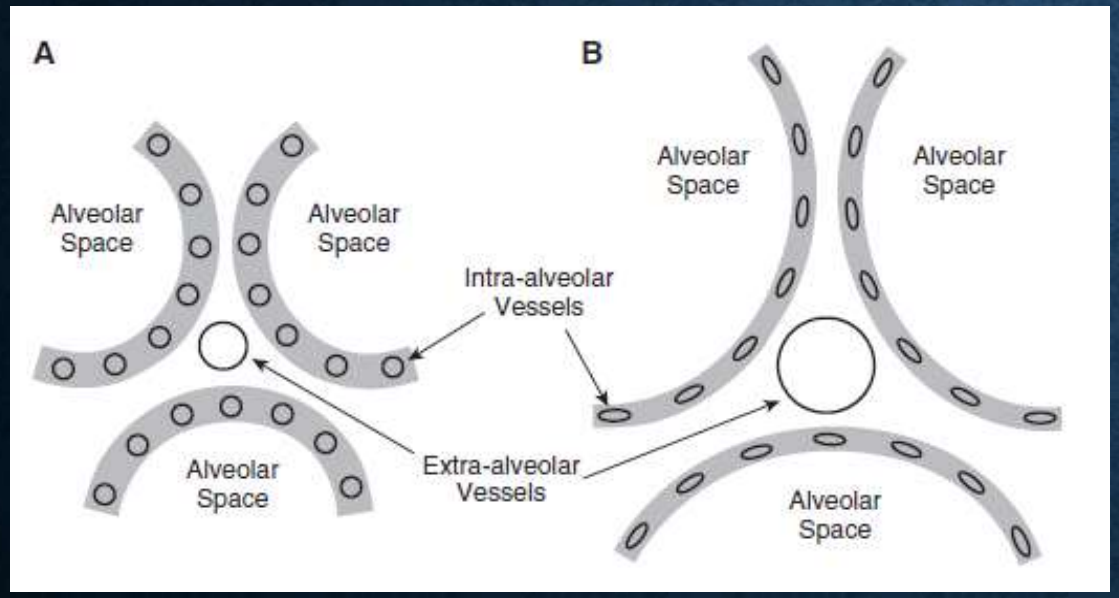


MIXING OF VENOUS BLOOD OF LOW ScvO2



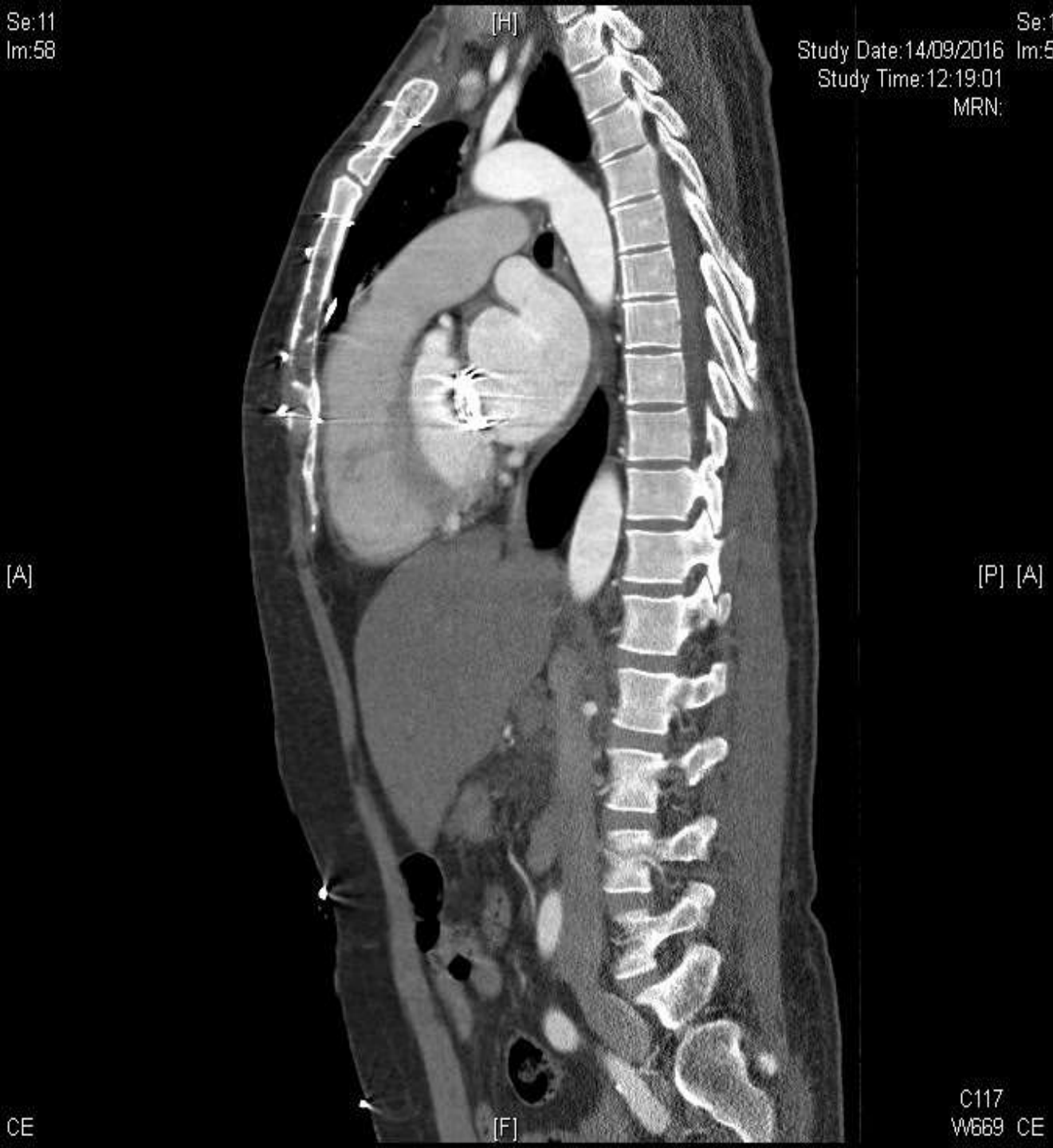
PEEP WORSEN HETEROGENEOUS LUNG

Collapse **Inflation**



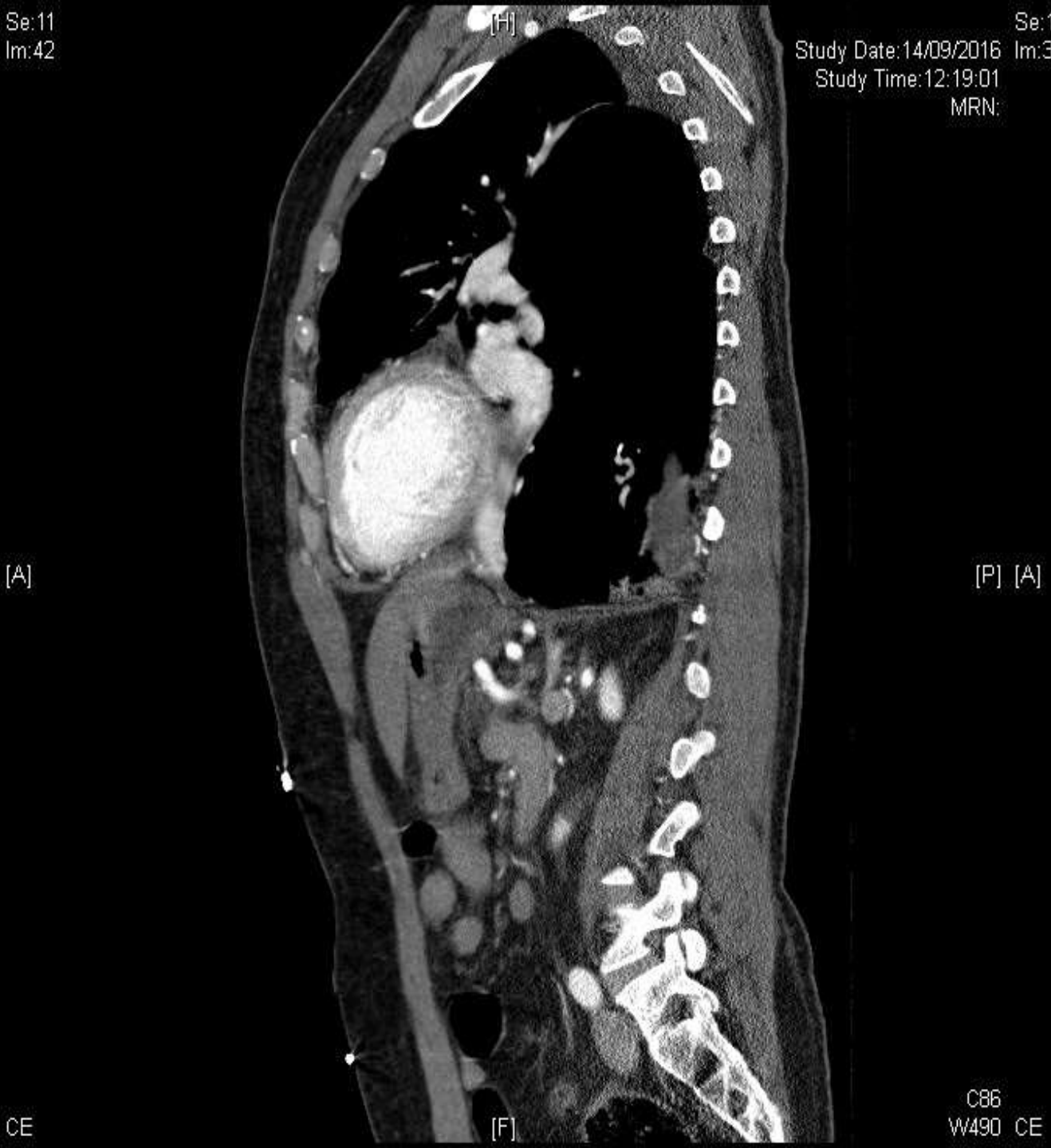
Pulmonary vasculature of alveoli

- Pulmonary vascular resistance (PVR) of a collapsed segment will decrease if PEEP recruit the segment and the extra-alveolar capillaries being opened
- PVR of a normal segment will not improve as it is optimally inflated, or may even raise if the intra-alveolar vessels under tension by hyper-inflation.
- Differential change in PVR increase shunting of blood to more diseased unit


Se:11
Im:58
[H]
[A]
[F]
CE

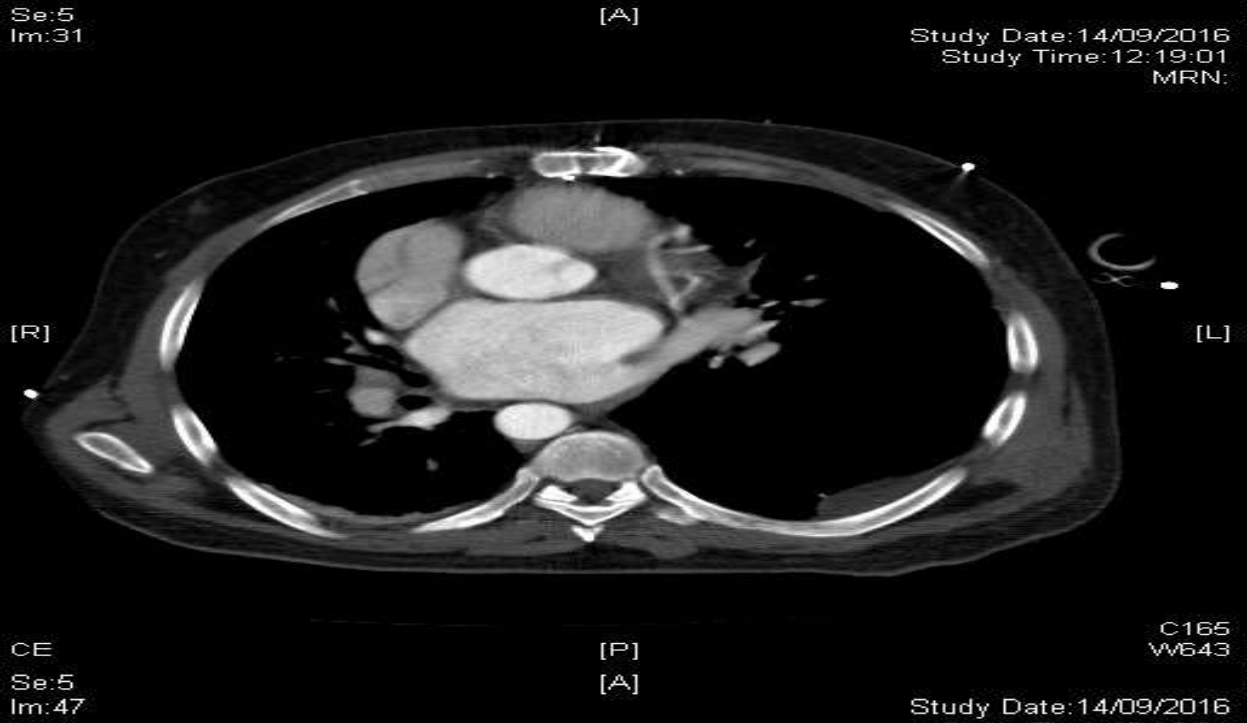
Se:11
Im:54
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[P]
[A]
C117
W669
CE

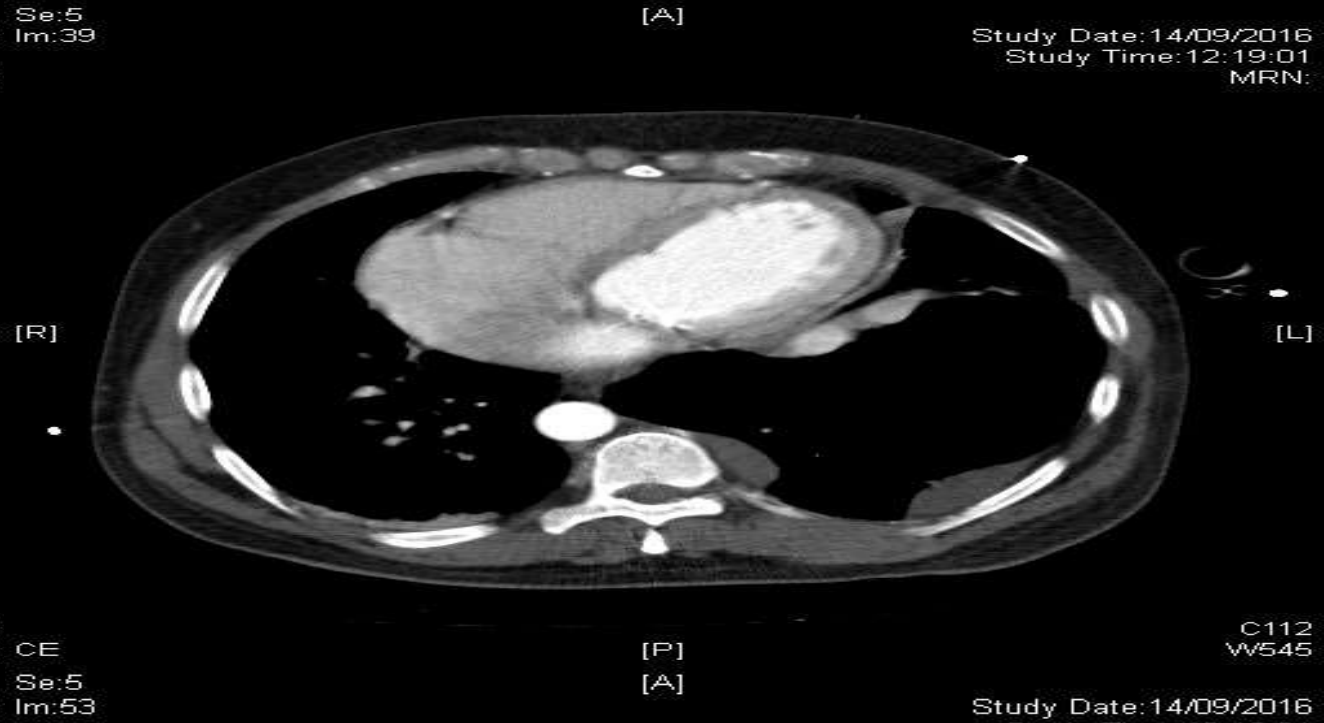
[H]
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[P]
[F]
C117
W669

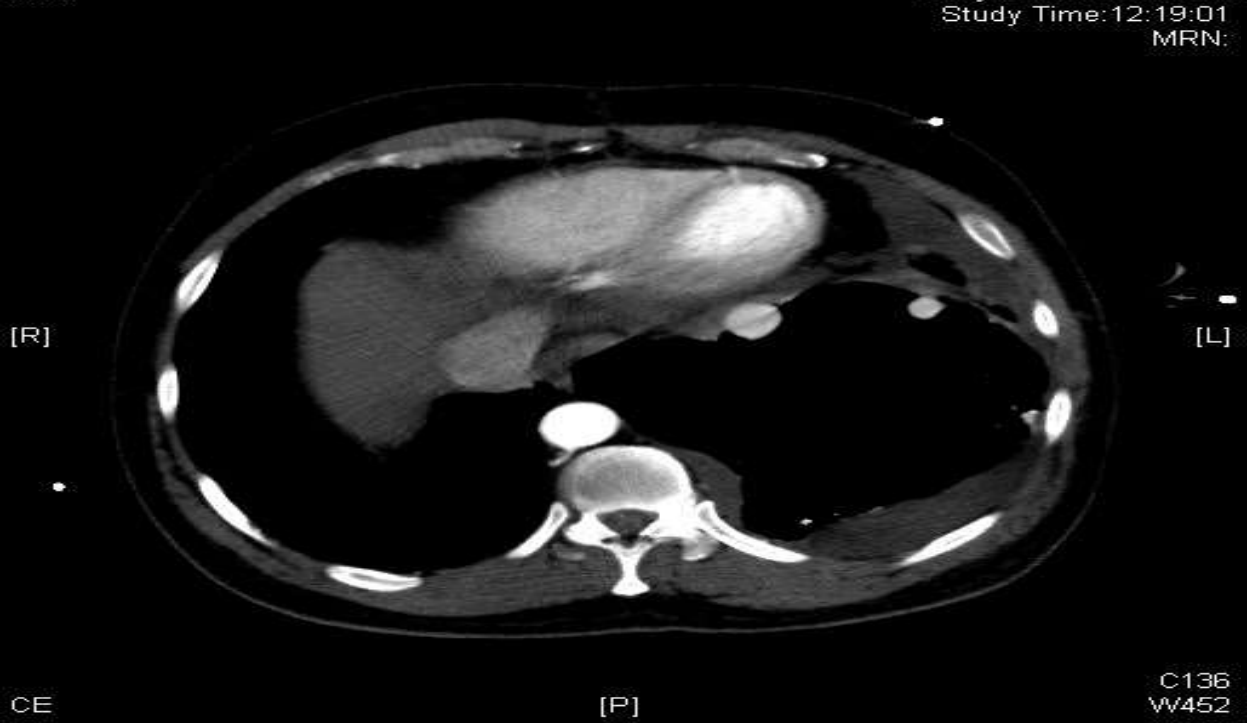

Se:11
Im:42
[H]
[A]
[F]
CE

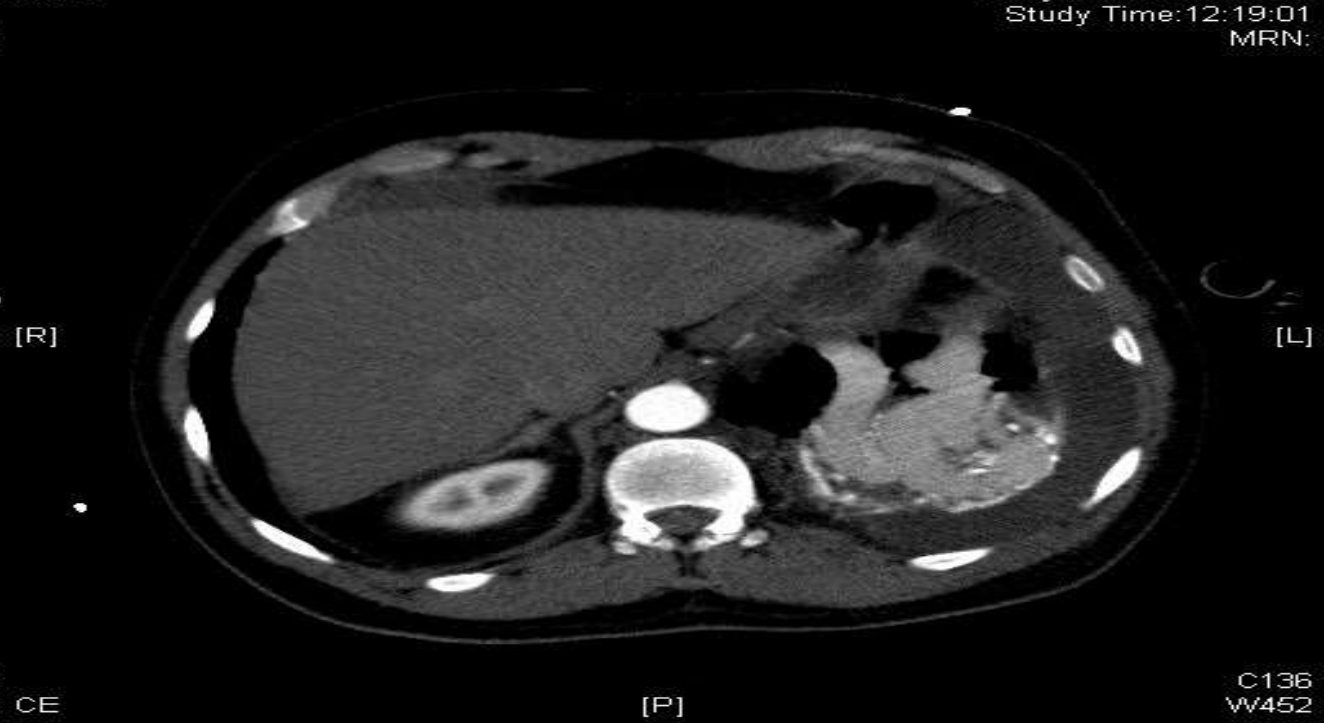
Se:11
Im:37
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[P]
[A]
C86
W490
CE

[H]
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[P]
[F]
C86
W490


Se:5
Im:31
[A]
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[R]
[L]
CE
[P]
C165
W643


Se:5
Im:39
[A]
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[R]
[L]
CE
[P]
C112
W545


Se:5
Im:47
[A]
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[R]
[L]
CE
[P]
C136
W452


Se:5
Im:53
[A]
Study Date:14/09/2016
Study Time:12:19:01
MRN:
[R]
[L]
CE
[P]
C136
W452

CT THORAX + AORTOGRAM

- No acute mural haematoma and no evidence of aortic dissection
- Cardiomegaly with dilated left atrium
- A large bulla and severe emphysematous change at whole Left LL
- A 7.7cm x 9.0cm x 3.1cm mass-like lesion consisting of a tangle of serpiginous prominent vasculature lying at the left lung base. It is connected to two prominent vasculature running anteriorly towards the hilum, joining left inferior pulmonary vein and left inferior pulmonary artery.
- Conclusion: Left lung huge bulla + pulmonary AVM

QUESTION 5

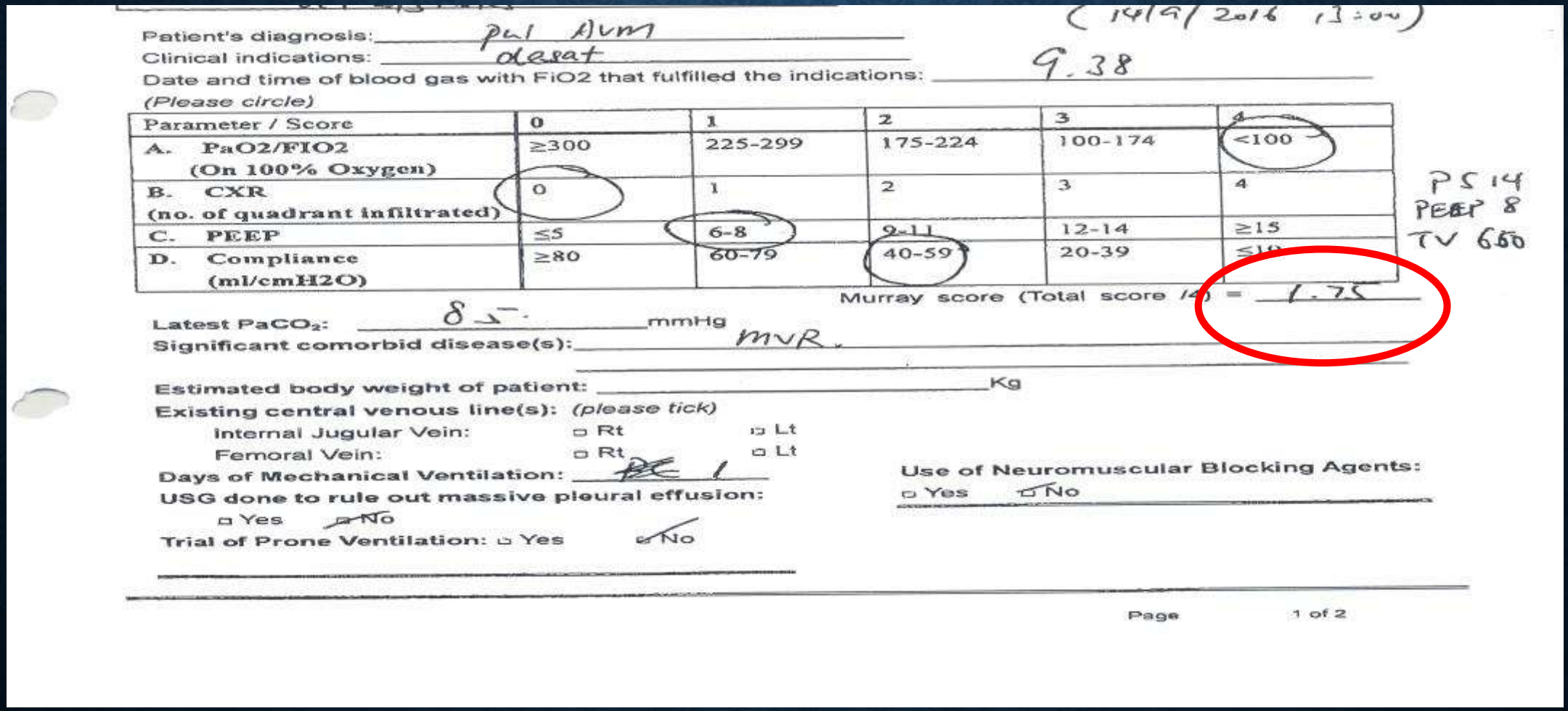
What can we do to reverse hypoxemia ?

What can we offer to treat the cause?

SEEK DIFFERENT SPECIALTIES' OPINION

- Cardiology review:
  - Echo with agitated saline: +ve after 3 cardiac cycles
  - Can give diuretics to promote urine output
- Cardiothoracic surgery for operative treatment of pul AVM
  - Bullae may have ruptured, but operative risk too high
  - Pul AVM for non-operative treatment
- Intervention radiologist
  - Embolization not feasible at POH; need transfer to TMH

CONSULTED ECMO CENTER……



Patient's diagnosis: Pul AVM (14/9/2016 13:04)
Clinical indications: desat
Date and time of blood gas with FiO2 that fulfilled the indications: 9.38
(Please circle)

Parameter / Score	0	1	2	3	4
A. PaO2/FIO2 (On 100% Oxygen)	≥300	225-299	175-224	100-174	<100
B. CXR (no. of quadrant infiltrated)	0	1	2	3	4
C. PEEP	≤5	6-8	9-11	12-14	≥15
D. Compliance (ml/cmH2O)	≥80	60-79	40-59	20-39	≤19

PS 14
PEEP 8
TV 650

Murray score (Total score /4) = 1.75

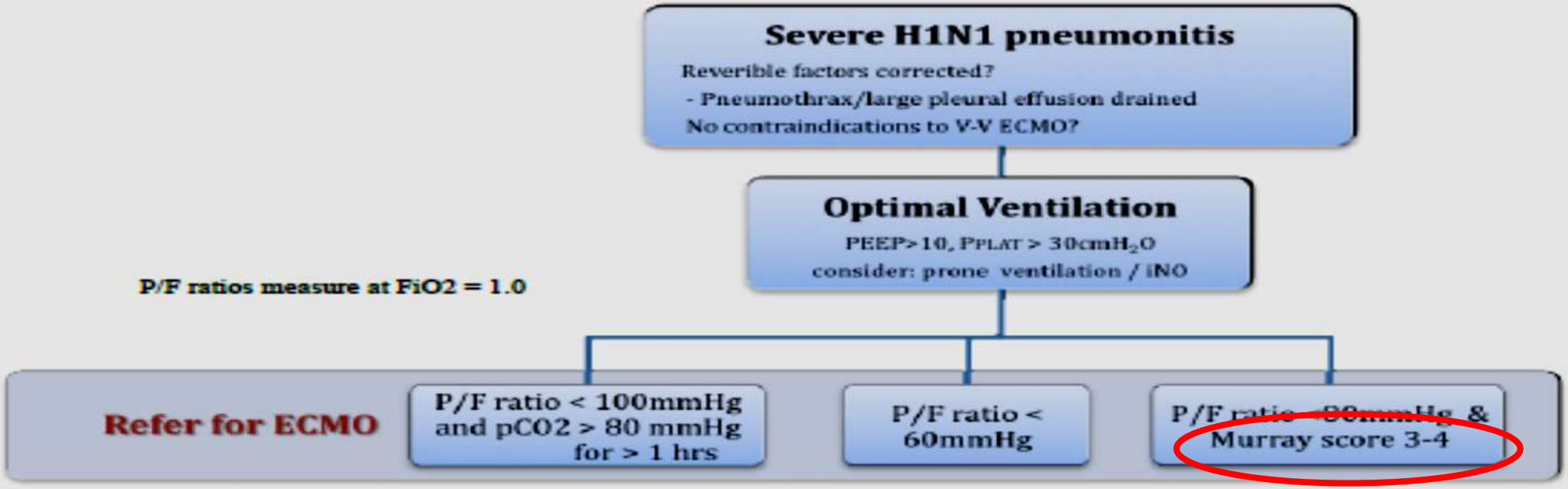
Latest $PaCO_2$: 8 ↓ mmHg
Significant comorbid disease(s): MVR.

Estimated body weight of patient: _____ Kg
Existing central venous line(s): (please tick)
Internal Jugular Vein: ☐ Rt ☐ Lt
Femoral Vein: ☐ Rt ☐ Lt
Days of Mechanical Ventilation: 1
USG done to rule out massive pleural effusion:
☐ Yes ☑ No
Trial of Prone Ventilation: ☐ Yes ☑ No

Use of Neuromuscular Blocking Agents:
☐ Yes ☑ No

Page 1 of 2

Indications[1, 2]



OOPS, NOT THIS TIME

ECMO CENTER REPLIED

- Not take-over for ECMO because
  1. Not fulfill Murray score being 3 or above
  2. No input from cardiothoracic surgery
  3. No detail of previous medical history
  4. Presence of large bullae/ AVM

CONSENSUS

"Continue expert care at POH ICU"

"Treat reversible cause i.e. infection"

from cardiologist/ cardiothoracic surgeon/ ECMO centers

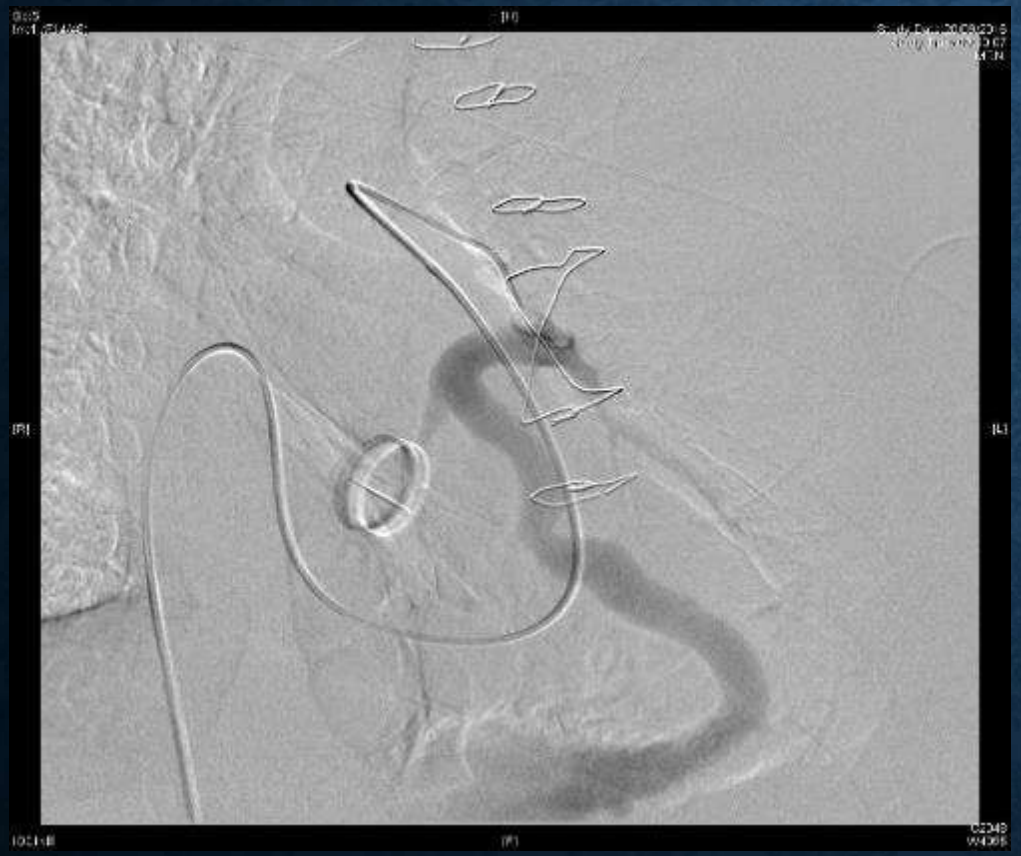
5PM: ICU STRATEGY AFTER "CONSENSUS"

- Acute: no evidence of pneumonia or CHF on CT thorax
- Chronic: lung function showed FEV1 is 1.79; FVC is 2.35 moderate airflow obstruction but to the degree of causing refractory hypoxia
- Cardiac: No frank RV failure or cor pulmonale; mechanical valve is not in malfunction
- Lung: bullae size static in serial CXR, unlikely contribute hypoxia significantly
- Conclusion: acute on chronic hypoxia which worsens after mechanical ventilation, likely related to positive pressure aggravating shunting
- Plan: to de-sedate and gradually withdraw ventilatory assist

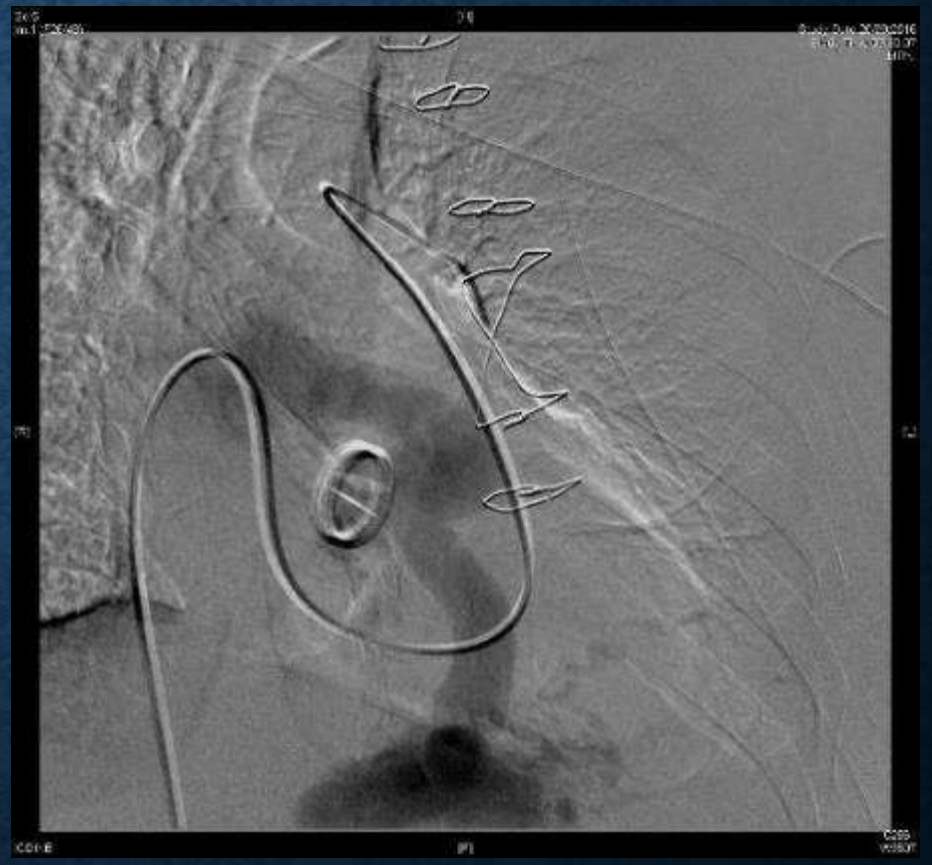
FURTHER ICU PROGRESS

- PEEP stepwise decrement form 14 to 4
- Switch of SIMV (PC) + PS mode to PS mode
- Stop further diuretics and give PRN bolus IVF
- At 6am 15th Sept
  - SaO2 was 90% with 0.5 FiO2 and PEEP 4
  - Wean off vasopressor
  - ABG showed pH 7.33; pCO2 6.19; pO2 8.38; BE -2.3; SaO2 91%
- He was extubated at 11am, SaO2 at 94-95% with 50% mask

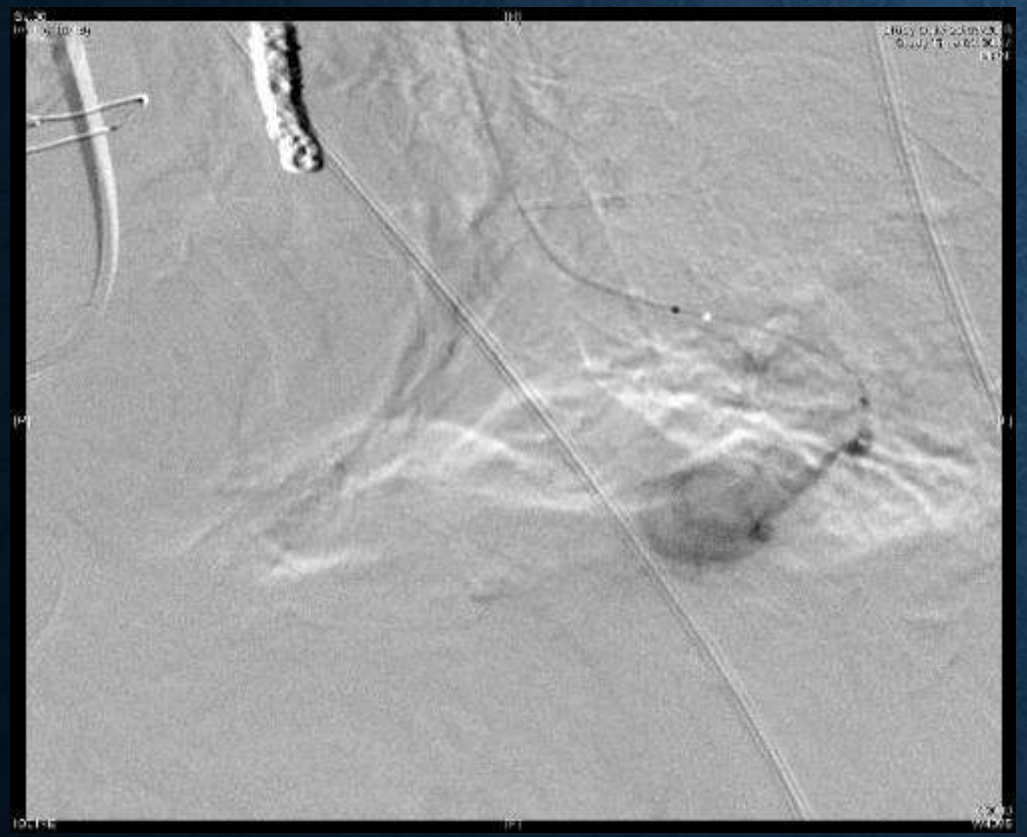
TO TMH FOR PULMONARY ANGIOGRAM



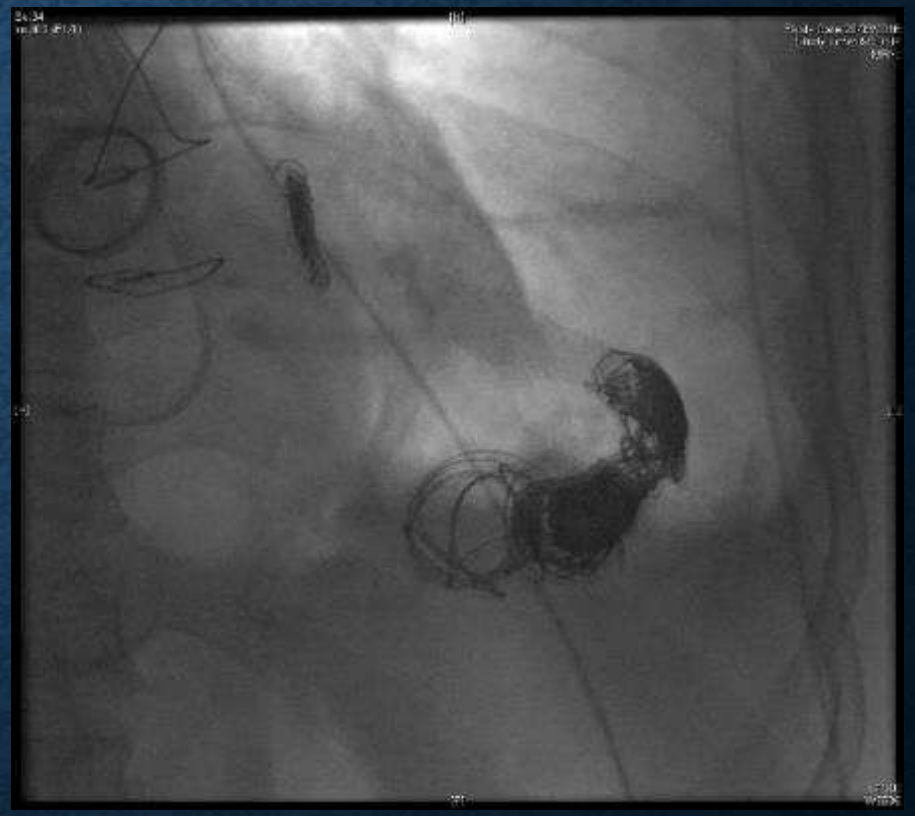
Left pulmonary artery



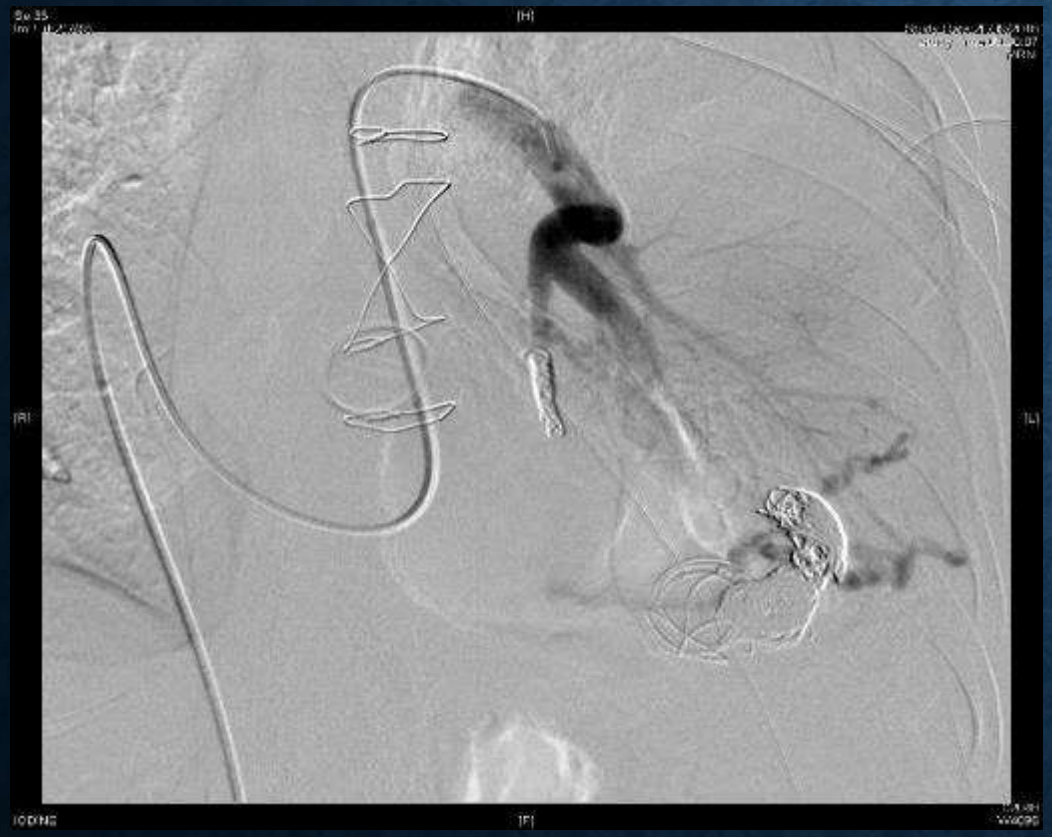
Big AVM draining to LA

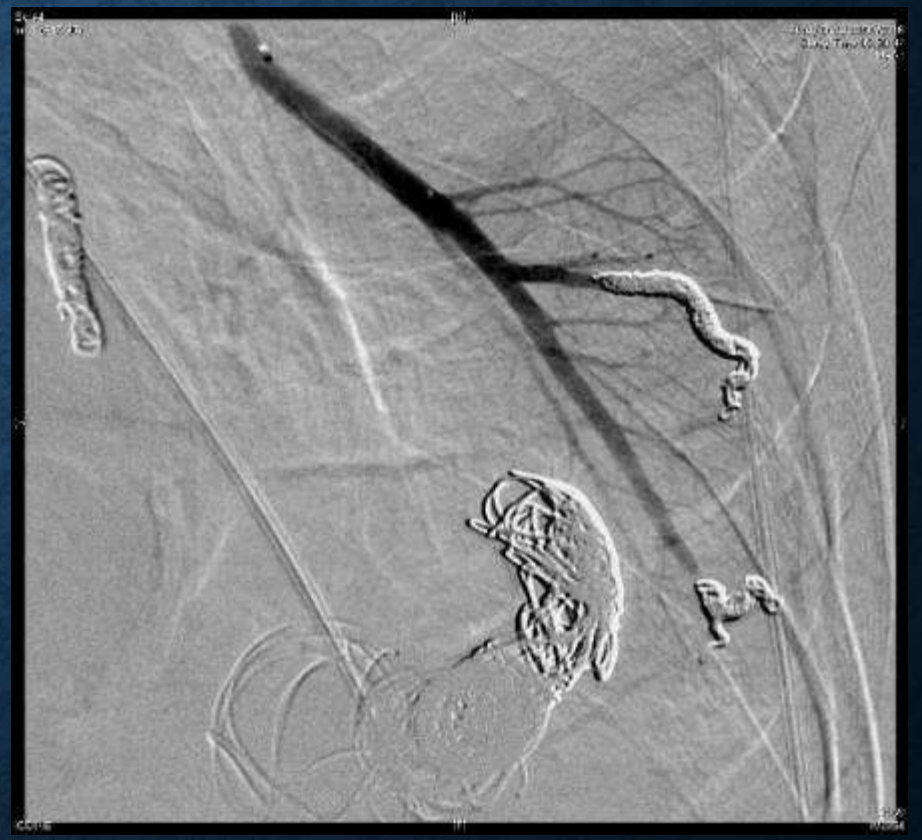


Emboli the feeding arm LPA



Occluding aneurysmal sacs





Still one feeding artery and was being emboli

ANGIOGRAM AND EMBOLIZATION

- Mean pulmonary arterial pressure before intervention is 34mmHg
- PAVM at base of left lung. It consists of hypertrophic segmental pulmonary artery, aneurysmal sacs and dilated draining vein
- Ruby coils 24mmx57cm, 24mmx57cm, 20mmx60cm, 20mmx60cm, 20mmx60cm, 16mmx60cm, 18mmx57cm, 14mmx60cm
- POD packing coils - J soft 60cm (x 2)
- Satisfactory occlusion of the PAVM was achieved. The mean pulmonary arterial pressure after intervention in 44mmHg. SaO2 after coil embolization is 98% on RA.

FROM ICU TO PAKISTAN

- Sputum PCR +ve for H3 Flu A and 5-day Tamiflu was completed
- Sputum/ blood and urine all negative growth

- ECMO center: PRN re-consult if necessary
- Cardiology: take over to POH Cardiac for rate control and re-warfarinization
- CTS: offer VATS bullectomy. Patient declined because of finance problem
- Patient was discharged to airport on 28th Sept

PULMONARY AVM (PAVM)

- Characterized by direct communication of pulmonary artery and pulmonary vein, without intervening capillary
- Incidence of PAVM is about 1:50,000 cases *Chin Med J (Engl) 2010;123:23-8.*
- Mayo clinic: 38 cases over 8.5 year (annual incidence 4.5 cases) *Mayo Clin. Proc.* 58:176–181
- Less than 400 cases were described in the literature. May be missed in autopsy.
- Women : men incidence = 2:1, but male predominant in newborns
- 10% presented in infant/ child, followed by a gradual increase in 5^{th}/ 6^{th} decades
- 70% of the cases are associated with Hereditary Hemorrhagic Telangiectasia (HHT)

DISTRIBUTION OF PAVM

- 75% unilateral; 80% sub-pleural; 50-70% at lower lobes

- Simple (80%) : between one pulmonary artery and one pulmonary vein
- Complex (20%) : two or more different segmental arteries supplying the aneurismal sac and one or two draining veins.

- Small (less than 5 cm) vs. Big (more than 5 cm) and may occupy whole hemithorax
- Primary (congenital) vs. Secondary (acquired)

PRESENTATION OF PAVM

Symptoms	Percentage
Symptomatic	91
Epistaxis	79
Dyspnea	82
Hemoptysis	11
Telangiectasis	69
Bruit	87
Clubbing	78
Cyanosis	79

Am. J. Surg. 89:1054–1080

SECONDARY (ACQUIRED) AVM

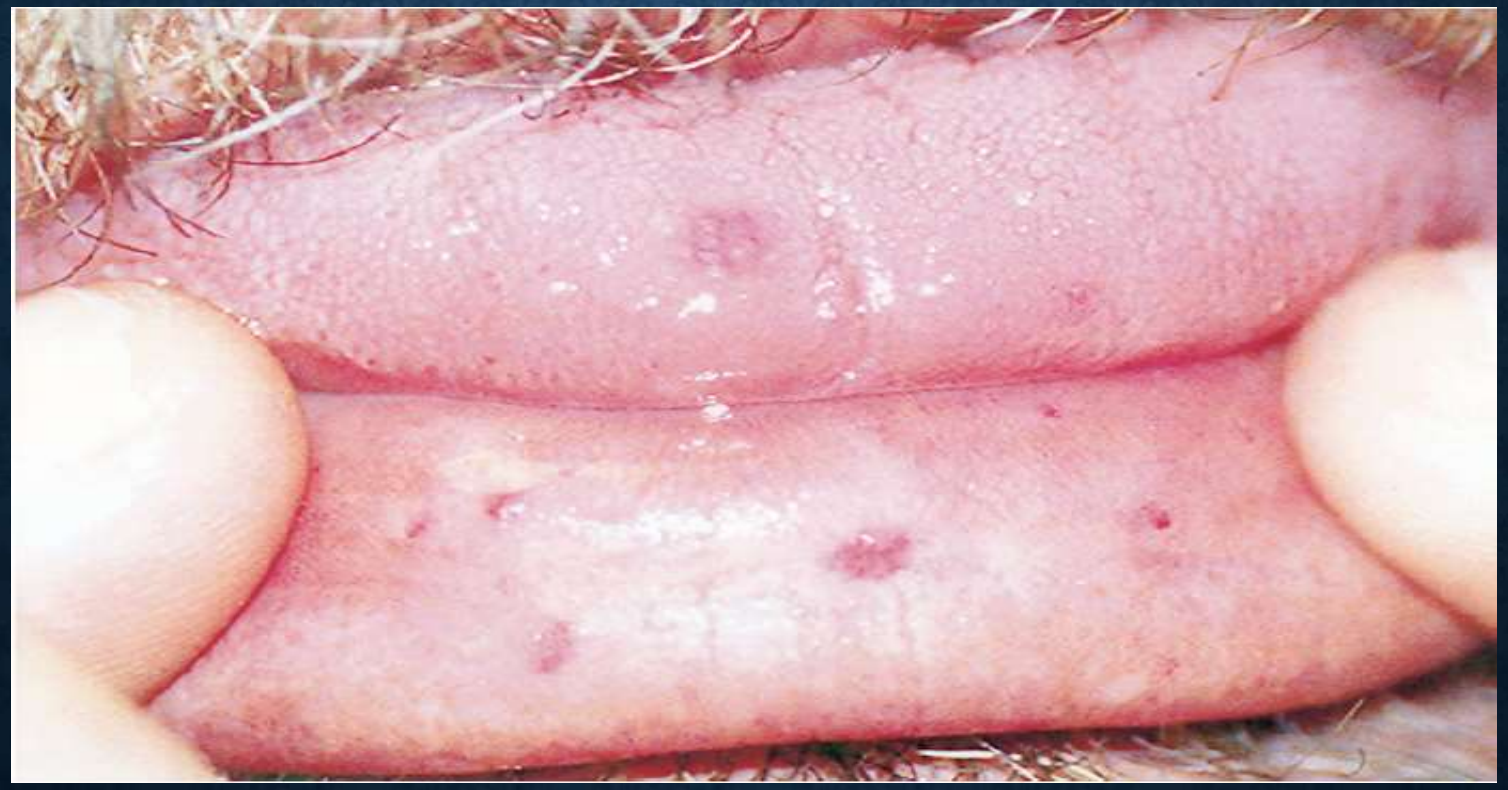
- Caused by presence pf hyperplastic changes in bronchial arteries or abnormal communication between pulmonary artery and vein

1. Post-traumatic
2. Chronic infections: TB/ Schistosomiasis/ Actinomycosis
3. Infiltrative: carcinomatosis (e.g. thyroid)
4. Cirrhosis: hepatopulmonary syndrome
5. Mitral stenosis
6. Iatrogenic: post-repair of congenital heart disease/ other open heart procedure

COMPLICATION OF AVM → NEED TX

- CNS: Seizure; Migraine headache; TIA/ CVA; Brain abscess
- Hypoxemia/orthodeoxia
- Resp: Hemothorax; Life-threatening hemoptysis; Pulmonary hypertension
- CVS: Congestive heart failure
- Polycythemia
- Anemia
- Infectious endocarditis/ cardiac emboli

Hereditary Hemorrhagic Telangiectasia (HHT)



Rendu-Osler-Weber syndrome

Hereditary Hemorrhagic Telangiectasia (HHT)

- Population incidence between 1/39,216 and 1/2,351
- Inherited as autosomal dominant pattern
- AVM in the skin, mucous membranes, and visceral organs
- 15 to 35% of patients with HHT have PAVM
- Asymptomatic until adult, usually 100% penetrance at 40-year-old
- Presented as cutaneous telangiectases or epistaxis.
- Of 80 patients with visceral involvement, only 9% had signs *Am. J. Med. Genet.* 32: 291–297

DIAGNOSTIC CRITERIA OF HHT

- Includes at least 3 of the following:

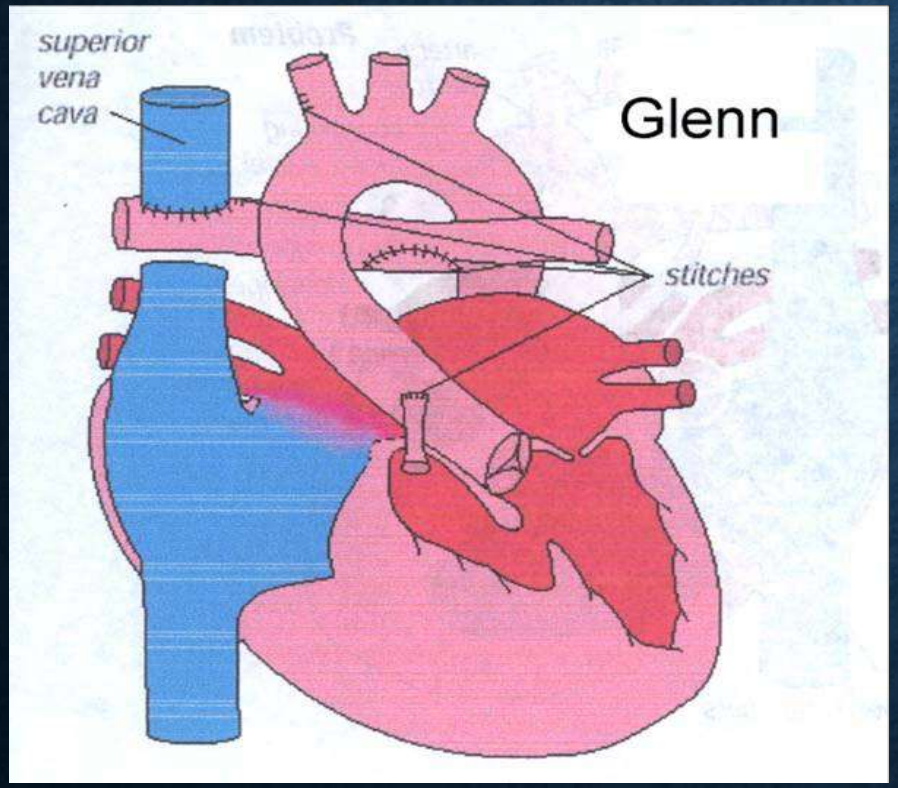
1. Recurrent and spontaneous epistaxis
2. Multiple mucocutaneous telangiectases
3. Visceral lesions (eg, GI arteriovenous malformations, pulmonary arteriovenous malformations)
4. First-degree relative with HHT by these criteria

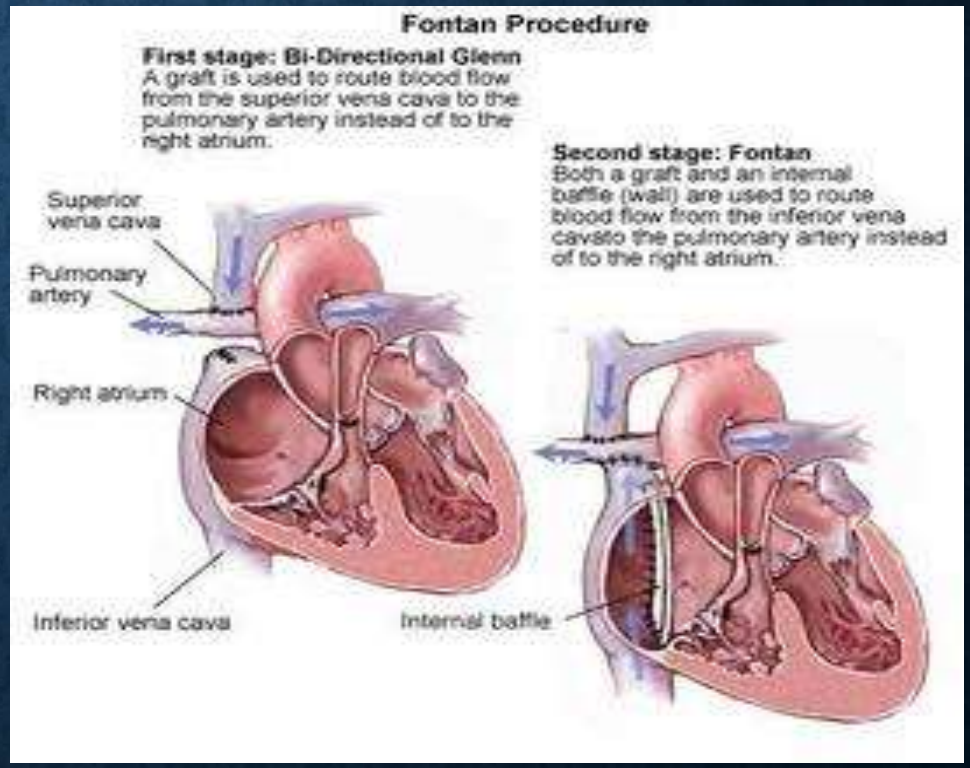
POST-OP FOR CYANOTIC HEART DISEASE

- Pulmonary AVM may develop after Glenn or modified Fontan procedures
- Glenn anastomosis (ie, superior vena cava [SVC] to right pulmonary artery [RPA]): PAVM occur in as many as 25% of cases
- Fontan operation (ie, SVC to right atrium and proximal RPA; hepatic veins to left pulmonary artery): designed as a surgical repair for congenital tricuspid atresia

- Need embolotherapy
- ECMO as salvage therapy to await embolization

JUST A REFRESH





86 YEAR-OLD WOMAN WITH SOB FOR 1/52

- Diastolic murmur at LLSB. Bilateral basal fine creps+
- Transthoracic echocardiogram (TTE) found severe mitral stenosis (valve area 0.88)
- Treat as acute CHF with diuretics
- Deteriorated with SaO2 only 88% with 10L O2; PaO2 56mm Hg

- CT showed Right infra-hilar large (4 cm) PAVM . A contrast echocardiogram (TTE with bubble study) confirmed presence of intrapulmonary shunt

Physicians Practice 2011

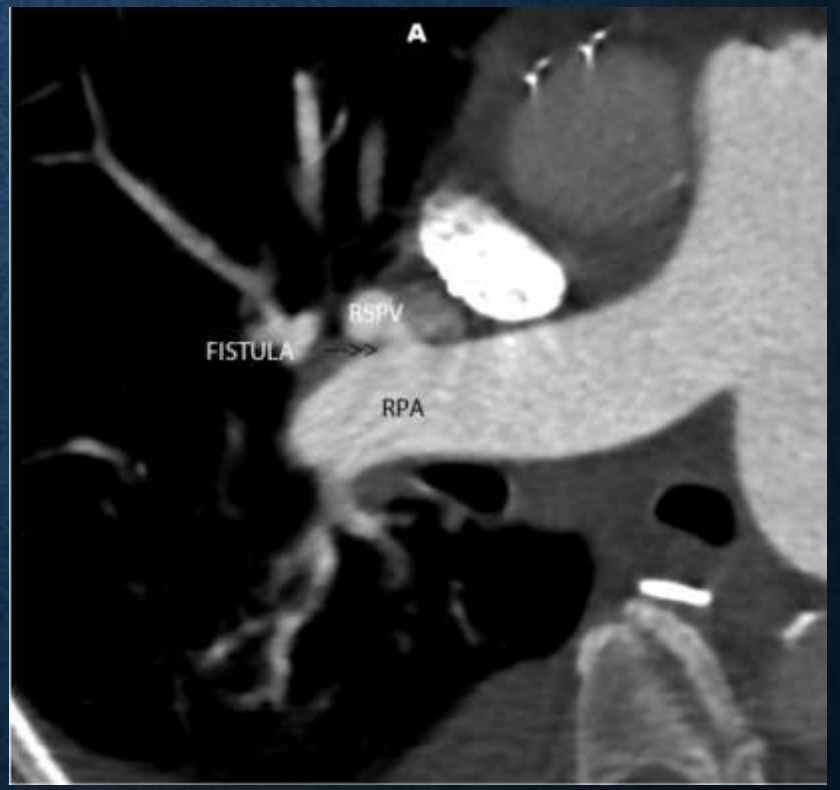
26 YR-OLD PAKISTAN BOY ACUTE SOB 2/7

- Oxygen saturation of 85% on room air
- No clubbing/ telangiectasis or murmur
- Polycythaemia with haemoglobin of 19.2 g/dL and haematocrit of 55%.
- Echocardiography was normal
- PO2=64 mm Hg on 0.28% FiO2 with an A–a gradient of 108
- CT revealed multiple arteriovenous malformationns (AVMs) in bilateral LL

BMJ Case Rep 2014

76 YEAR-OLD WOMAN WITH MVR AND CABG

- 2-week SOB 10 years after procedures
- SaO2 was 80% on 100% NRM
- Cyanosis. No clubbing
- Echo normal study with +ve bubble test after 3rd cardiac cycle
- Rt heart cath: NO intracardiac shunt
- CTA showed 1.1 cm right pulmonary artery to superior pulmonary vein fistula



BMJ Case Rep 2014.

84-YEAR-OLD FEMALE WITH HHT & PAVMS

- Admit ICU for severe SOB
- ABG on NRM 100% O2 revealed a pH of 7.39, a pCO2 of 33, and a PaO2 of 63
- Contrast echocardiography confirmed right-to-left intrapulmonary shunting
- [illegible] for anticipated need of transport to [illegible]
- Paralyzed and sedated: PCPS mode with peak inspiratory pressure at 20: PEEP at 5 cm H20; and FIO2 at 100%, and her SpO2 dropped to 60%
- Off relaxant, put on PS mode with ZEEP, SpO2 > 80%. Successful embolization

Chest. 2009;136(4_MeetingAbstracts)

TREATMENT OF PAVM -- EMBOLIZATION

- Embolotherapy appears to be the treatment of choice
- Coil(s) placed distal to any branch of feeding vessel
- Amplatzer duct occluder (ADO) for feeding vessels larger than 7-10 mm
- Early complications rupture of blood vessels, arrhythmias, and vascular occlusion.
- Pleuritic chest pain is the most common symptom is observed in 12%
- Average success rate of 98.7%
- Symptomatic recanalization was observed with 0.5% of procedures
- Rare to develop new pulmonary HT or worsened existing pulmonary HT

LONG TERM OUTCOME: EMBOLOTHERAPY

- 150 patients of technically successful PAVM embolization were reviewed
- *Total 415 AVMs were treated in 205 sessions*
- Involution in 97% of embolized lesions
- 97 previously small pulmonary arteriovenous malformations had enlarged to a significant size in 28 patients (18%)
- Complications included respiratory symptoms (n = 13), cerebral ischemia (n = 4), brain abscess (n = 5), hemoptysis (n = 3), and seizure (n = 1).

J Vasc Interv Radiol. 2006 Jan. 17(1):35-44

TREATMENT OF PAVM -- SURGERY

- Dropped out of favor since late ’70
- Any sort of resection: segmentectomy/ lobectomy/ pneumonectomy
- Recurrence or enlargement of the pulmonary AVM in as many as 12%
- Perioperative mortality has varied from 0-9.1%
- Indication: untreatable allergy to contrast

HOW MUCH DID “WE” PAY

Pakistan Rupee

Six Million

HK Dollars

400,000


THE
SIX MILLION
MAN
RUPEE

TAKE-HOME MESSAGE

- CXR imaging needs to be interpreted
- Be familiar with the causes of worsening with PEEP
- When things go wrong, always take time to consider making a turn
- PAVM, although not common, can be a diagnostic challenge, especially when it presents as life-threatening complication e.g. hypoxemia/ hemoptysis/ emboli
- ECMO may be an interim measure to await more definite therapy
- Surgical treatment of complex PAVM may be one day needed