

CASE SHARING

Dr Calvin Hoo

Trainee in Haematology and Oncology

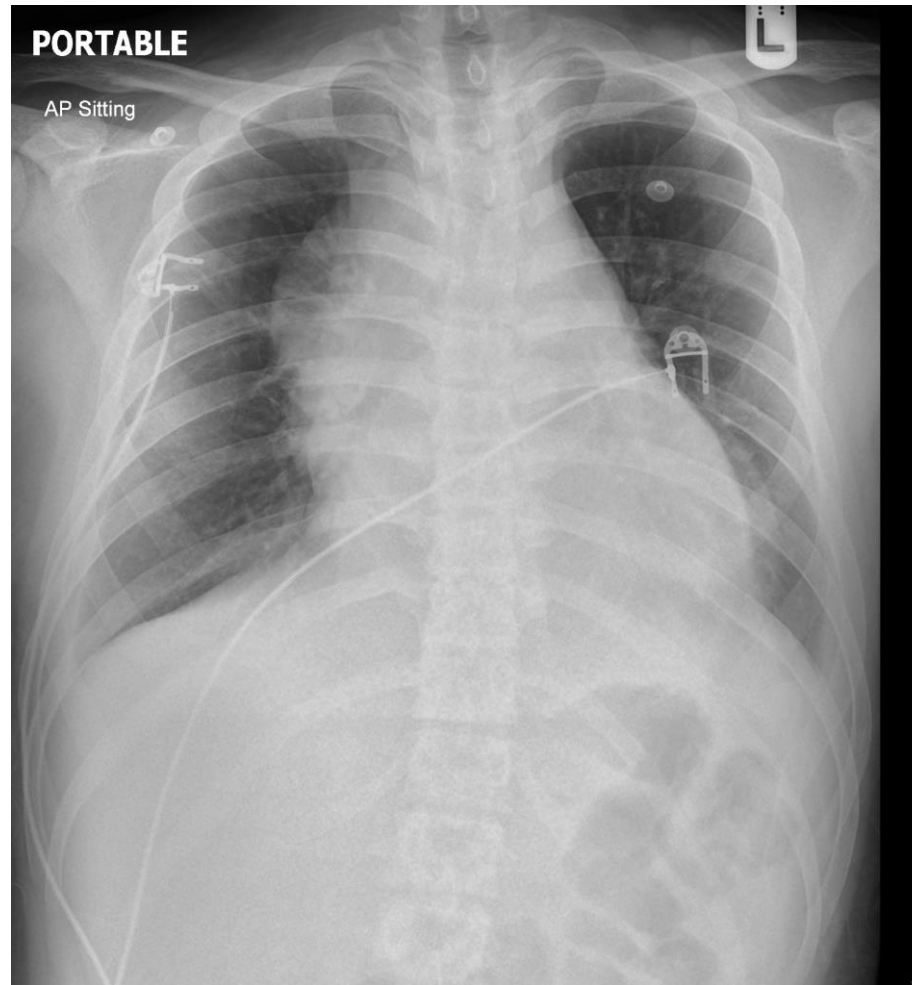
Hong Kong Children's Hospital



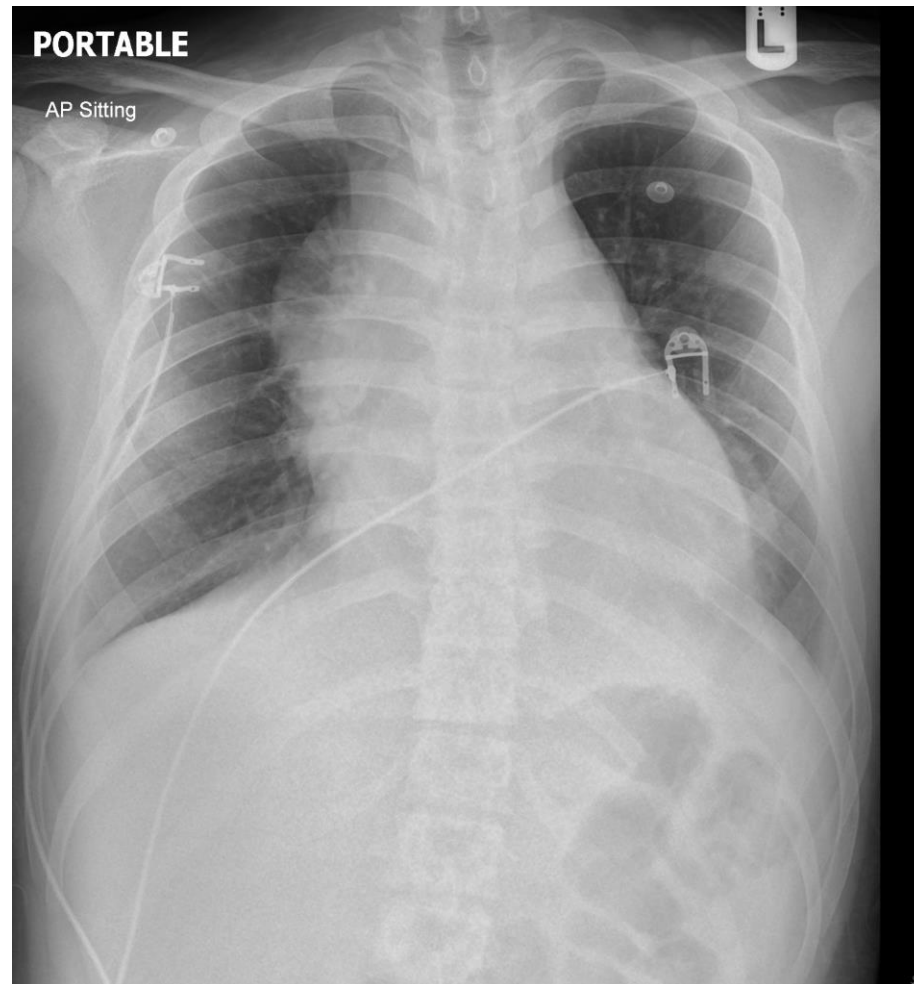
香港兒童醫院

Hong Kong Children's Hospital

Consultation...



1. What are the abnormalities?
2. How to confirm your diagnosis?
3. What are your worries?
4. What to do?



Case history

Diagnosis:

- Stage III mediastinal T-cell Lymphoblastic Lymphoma

Basic demographics:

- 16y/M
- Good past health
- No family history of oncological or haematological diseases
- Onset of symptoms for 2 weeks
 - Progressive cough, SOB, low grade fever
- No B symptoms
 - No significant weight loss
 - No high fever
 - No drenching night sweating

Physical exam

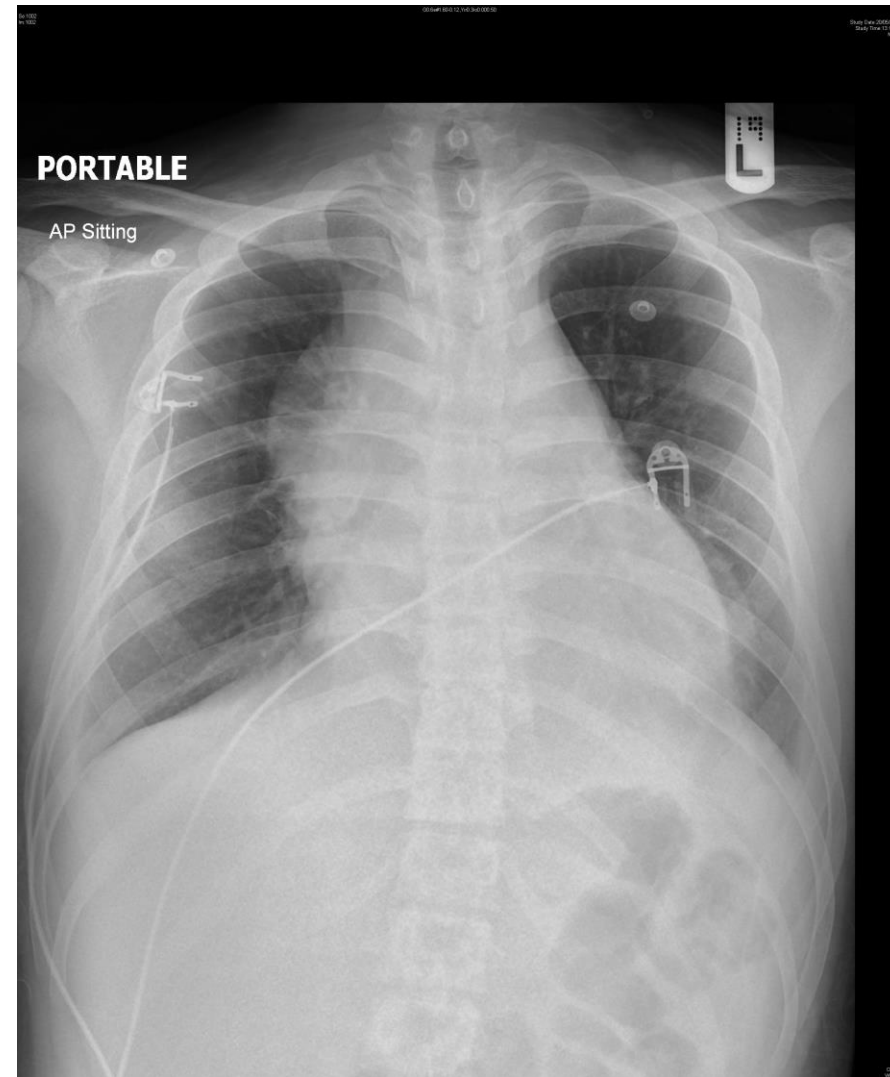
- Clinically well, not septic-looking
- CVS:
 - Tachycardia 130/min, BP normal
 - Signs of **SVCO**
 - **Distended neck vein**
 - No facial plethora, upper limb edema, Pemberton's sign
 - Signs of **cardiac tamponade**
 - **Cardiac apex deviated laterally**
 - **Muffled heart sound**
- Resp:
 - Tachypnea 24/min
 - On 2L O₂/min via NC
 - Trachea central, not deviated
 - No percussion dullness
 - AE satisfactory and symmetrical
- Abd: soft, non-tender, no hepatosplenomegaly
- Testes normal
- No enlarged cervical/ axillary/ groin LNs

Investigations

- Blood tests at presentation:
 - Hb 11.4, WBC 13.4, Plt 521
 - Smear: no blast
 - RFT, electrolyte normal
 - **LDH 517 (H), urate 0.47(H)**

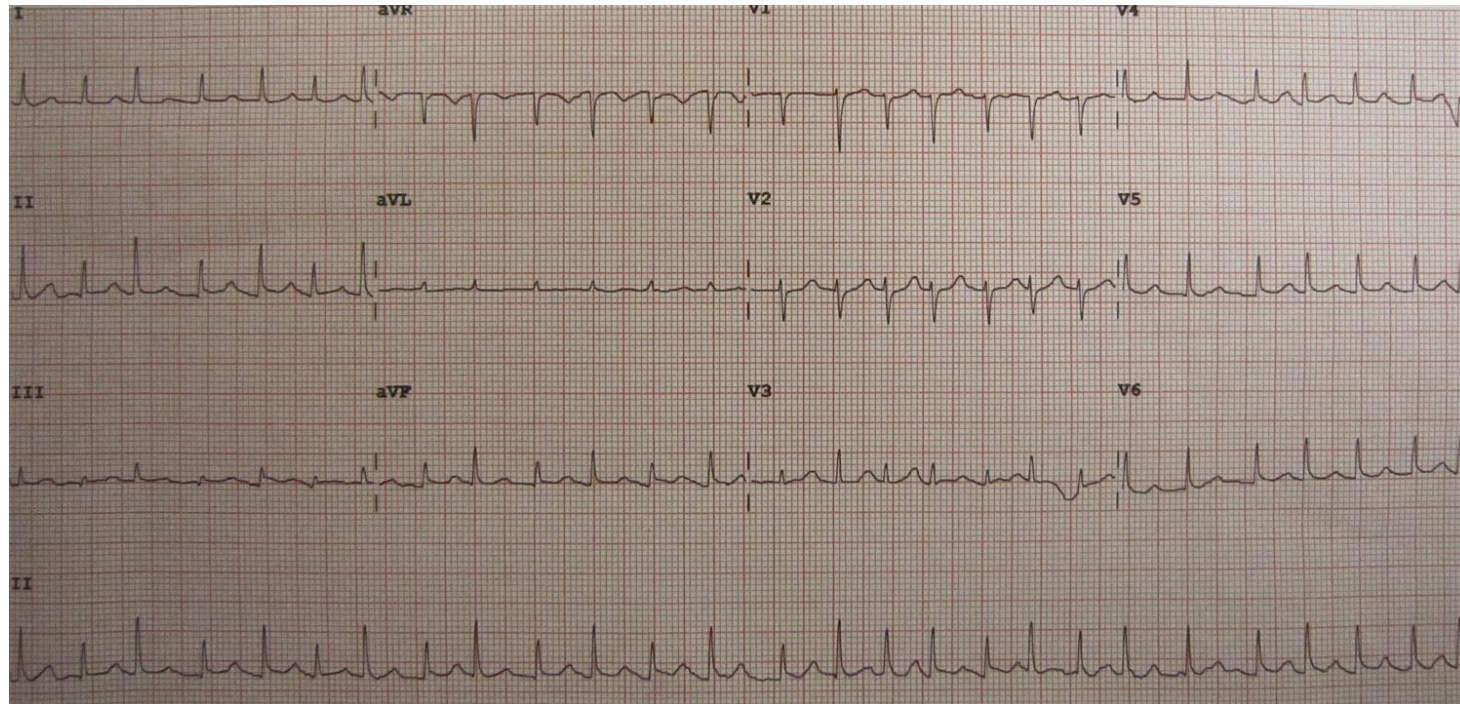
Investigations

- CXR:
 - Cardiomegaly
 - Mediastinal mass
 - Blunted left costophrenic angle



Investigations

- ECG:
 - Sinus tachycardia
 - **Low QRS voltage with electrical alternans -> cardiac tamponade!**
 - No ischemic changes



Progress

- Cardiac echo done by adult ICU:
 - massive pericardial effusion with features of **tamponade**
- USG-guided pericardiocentesis by adult cardiologist
 - 500ml blood stained fluid tapped out
 - Connect to pericardial drain with output ~450ml on first day
- Pericardial fluid: haematolymphoid malignancy of T cell lineage

Progress

- Transferred to HKCH PICU for further management on D3
- Echo done on admission:
 - minimal pericardial effusion
 - significant bilateral pleural effusion
 - **normal** cardiac structure and systolic function
- Urgent CT thorax:
 - 12cm mass at anterior and middle mediastinum
 - Tumor encases and compresses
 - **SVC** (lumen 0.7cm) and bilateral brachiocephalic veins
 - **Bilateral atria**
 - **Pulmonary trunk** and proximal part of bilateral pulmonary arteries
 - **Carina** and **bilateral main bronchi**

Progress

- Bilateral chest drain insertion
- Pleural fluid: T-lymphoblastic lymphoma
- BM aspirate + trephine: no invasion of lymphoma cells
- Diagnostic LP: no blasts

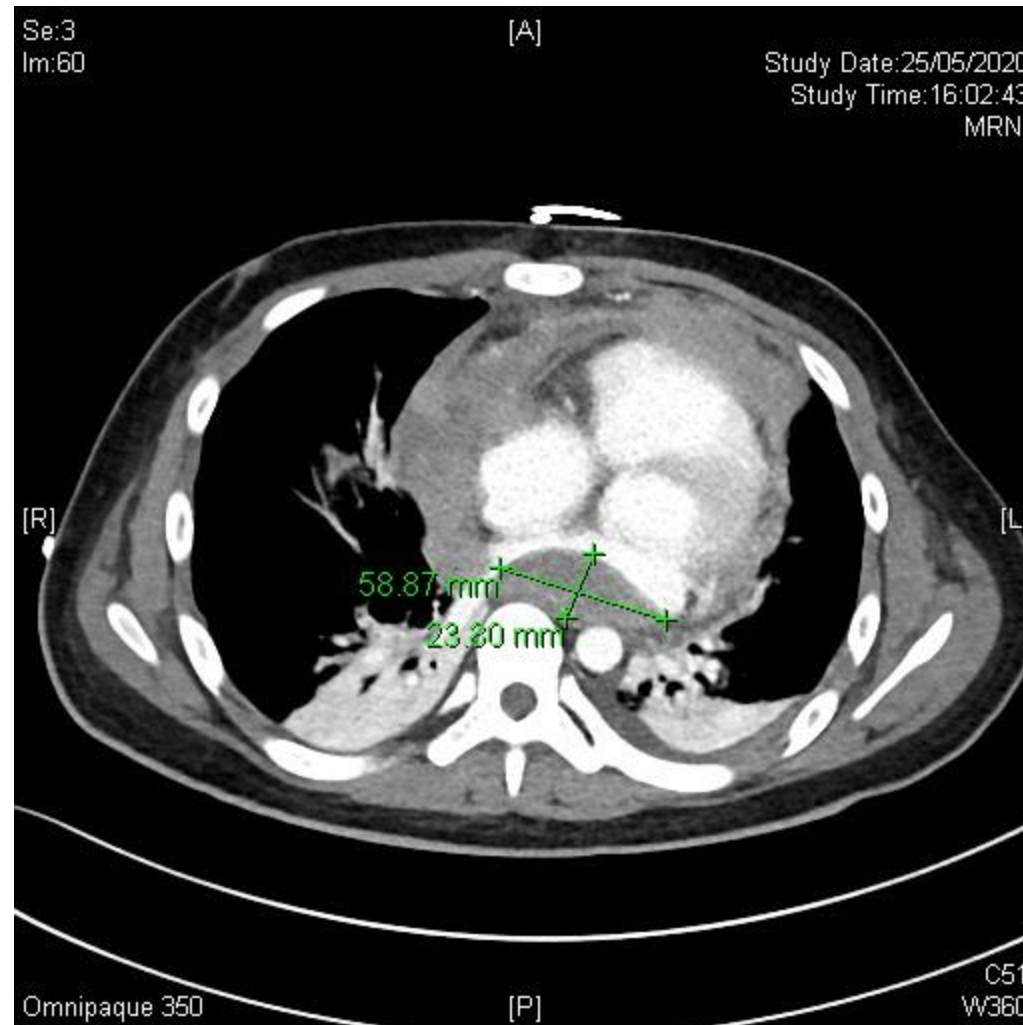
- Continue **hyperhydration**
- **Rasburicase** total given for 5 days, then to **allopurinol**
- Treated according to HK LBL 2008 protocol
- Started cytoreductive prephase **Prednisolone**
- No clinical or biochemical features of **tumor lysis syndrome**
- Also cover with broad-spectrum antibiotics

Progress:

- Progress CT thorax 3 days after
 - Increase compression to **SVC**, brachiocephalic veins and **left atrium**,
 - New development of **thrombus** extending from left brachiocephalic vein to left subclavian vein

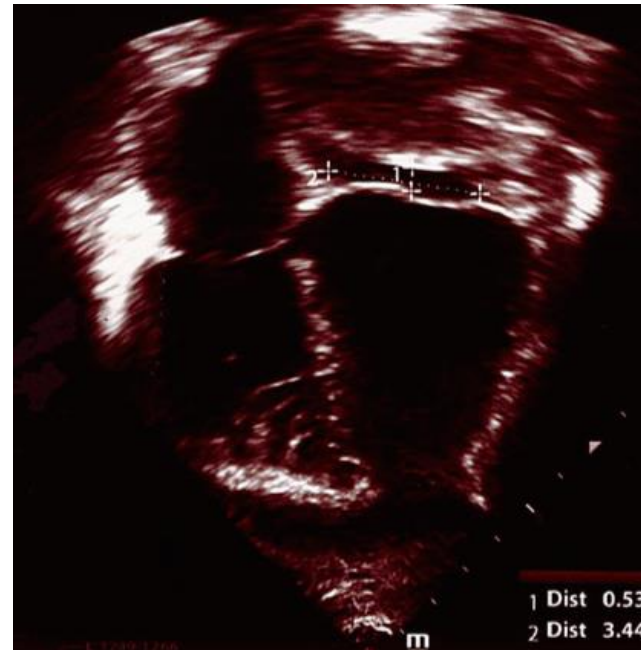
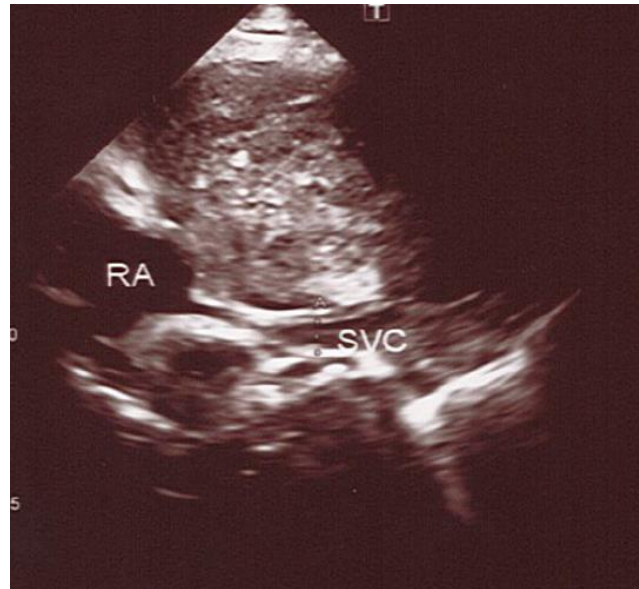


Progress:



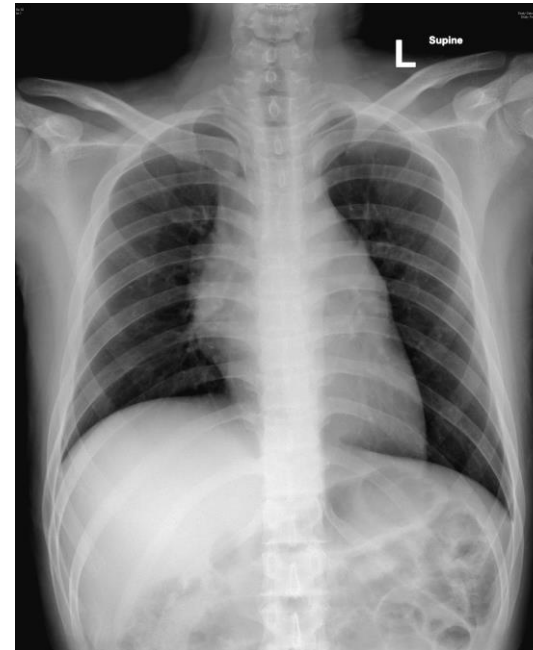
Progress

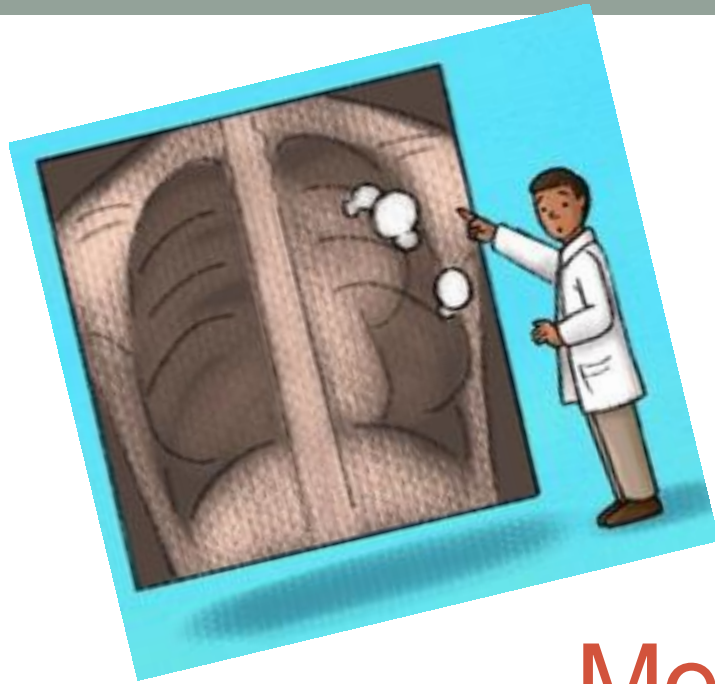
- Progress cardiac echo on the same day
 - Mass compressing **LA** 4 X 1.8cm
 - No evidence of pulmonary hypertension
 - No tamponade effect on RV/LV
 - **SVC** stenosis 3.5mm



Progress

- Take over to QMH PICU for observation for any further mass progression causing haemodynamic/ respiratory collapse
- Transfer back to HKCH in view of no further progression of tumor
- Continue chemotherapy
- Progress CXR 15days after chemo
 - Interval decrease in size of tumor





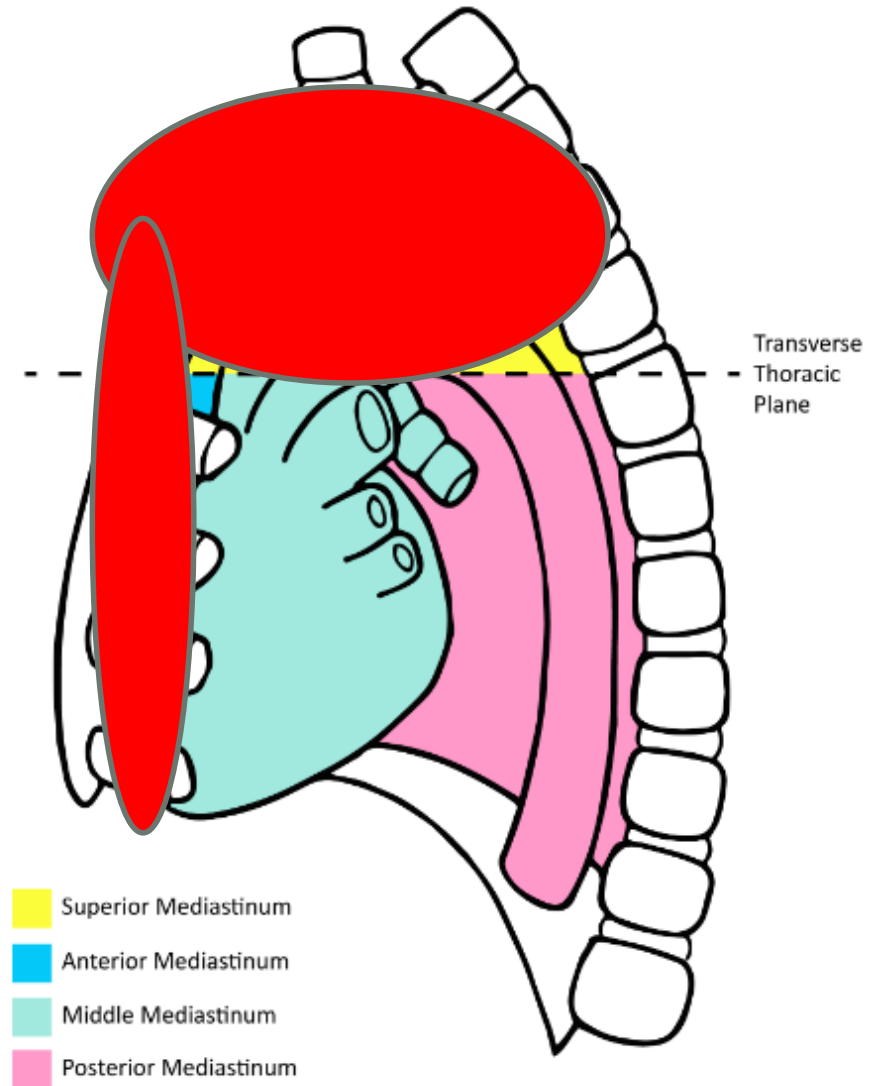
Mediastinal mass



Mediastinal mass - overview

- Children more prone to development of complications
- **Anesthetic deaths** mainly reported in children
 - Smaller caliber of blood vessels and airway
 - More compressible cartilaginous structure of airway
 - Small intrathoracic volume
- **Severity**
 - Pace of tumor growth
 - Size of tumor
 - Location of obstruction
- Not uncommon – **6 cases** since oncology service commenced in HKCH in Apr 2019

Anatomy



Etiologies

Benign

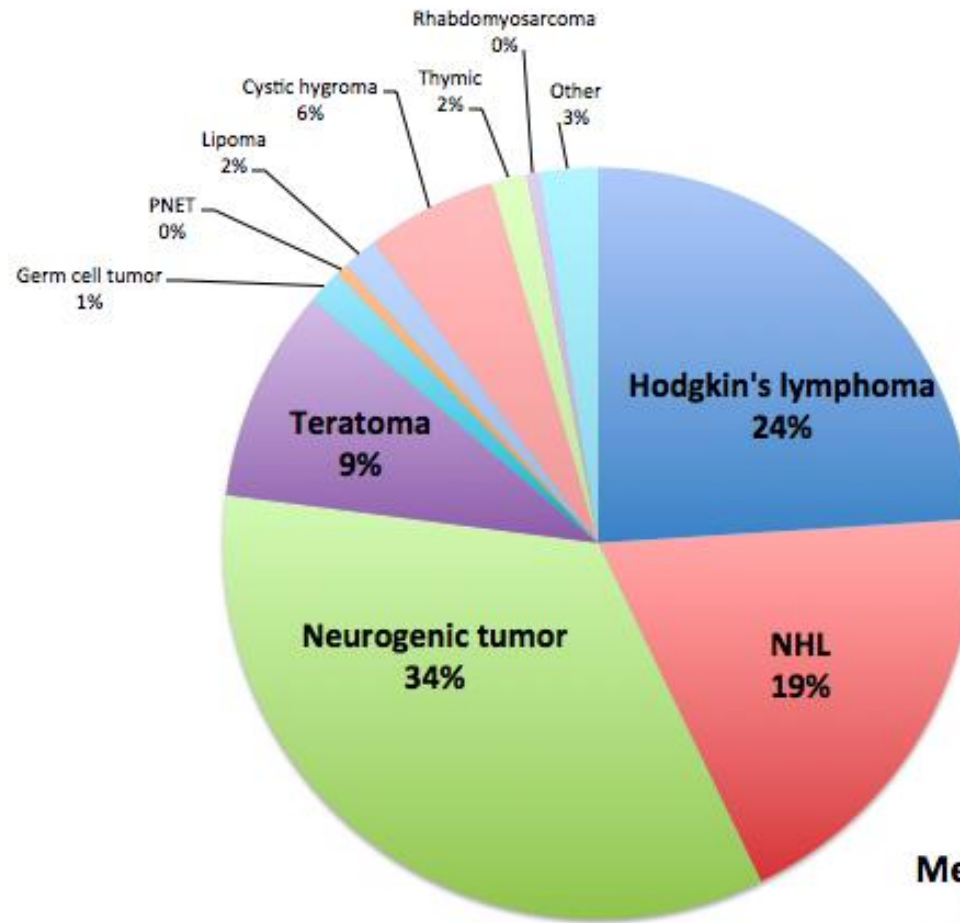
- Teratoma
- Lipoma
- Cystic hygroma

Malignant

- Lymphoma
 - Hodgkin lymphoma
 - NHL – T-LBL
- T-acute lymphoblastic leukaemia (T-ALL)
- Neurogenic tumor
- Malignant germ cell tumor

Adult: 4”T” (Thymoma, Thyroid cancer, Teratoma, Terrible lymphoma)

Etiologies



**Mediastinal mass in Children -
Experience in 196 cases**

Grosfeld et al. Annals of Surgical Oncology - 1994

Clinical emergency

- Life-threatening!
 - Potential **airway obstruction**
 - Potential **haemodynamic collapse**



Superior Vena
Cava Obstruction
(SVCO)

Mediastinal Mass
Syndrome (MMS)

Cardiac tamponade

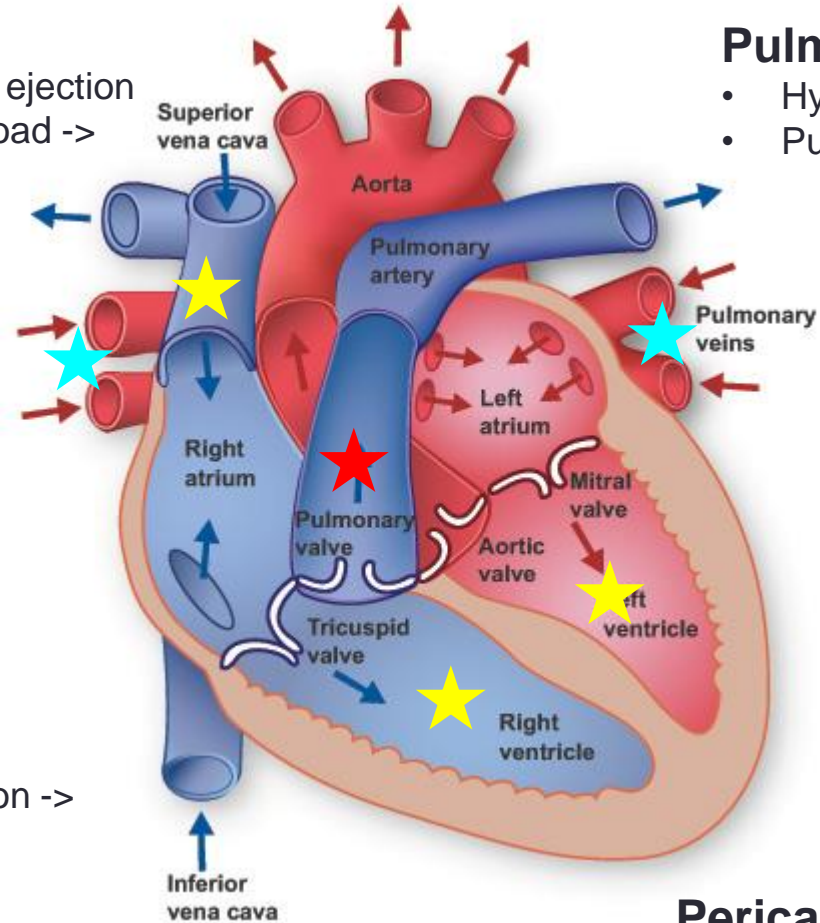
MMS effects on respiratory system

1. Obstructive physiology
2. Restrictive physiology
 - Reduced total lung capacity and functional residual capacity by mass compression, atelectasis, pleural effusion
3. Ventilation-perfusion mismatch

MMS effects on haemodynamic functions

Cardiac chambers, SVC

- Decrease ventricular filling or ejection
- Decrease left ventricular preload -> decrease cardiac output



Pulmonary veins

- Hypoxaemia
- Pulmonary edema

Pulmonary arteries

- Decrease pulmonary perfusion -> V/Q mismatch
- Right ventricular failure

Pericardial effusion -> cardiac tamponade

Who are the at risk patients?

- Orthopnea

- King DR, Patrick LE, Ginn-Pease ME, et al : Pulmonary function is compromised in children with mediastinal lymphoma. J Pediatr Surg. 1997;32:294-9

- Stridor

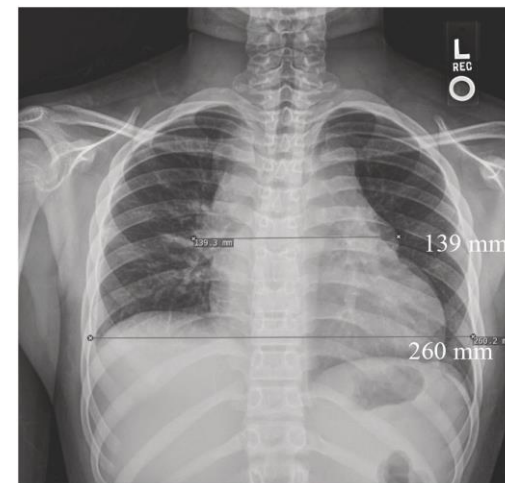
- Hack HA, Wright NB, Wynn RF. The anaesthetic management of children with anterior mediastinal masses. Anaesthesia. 2008; 63:837-46.

- Upper body oedema

Signs or symptoms correlate poorly with degree of airway or vessels compression

Who are the at risk patients?

- Major vessel compression, mainstem bronchus compression
 - Anghelescu DL, Burgoyne LL, Liu T, et al : Clinical and diagnostic imaging findings predict anesthetic complications in children presenting with malignant mediastinal masses. Paediatr Anaesth. 2007 ;17:1090-8.
- Mediastinal mass ratios (MMR)
 - Maximum width of the mediastinal mass divided by the maximum intrathoracic width (usually at the level of the diaphragm)
 - Cut-off value **>0.45**

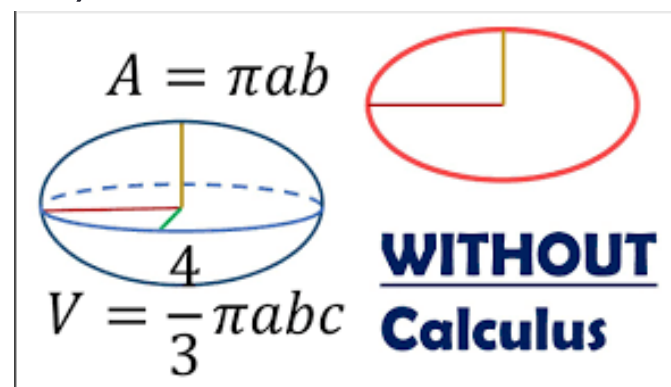


Who are the at risk patients?

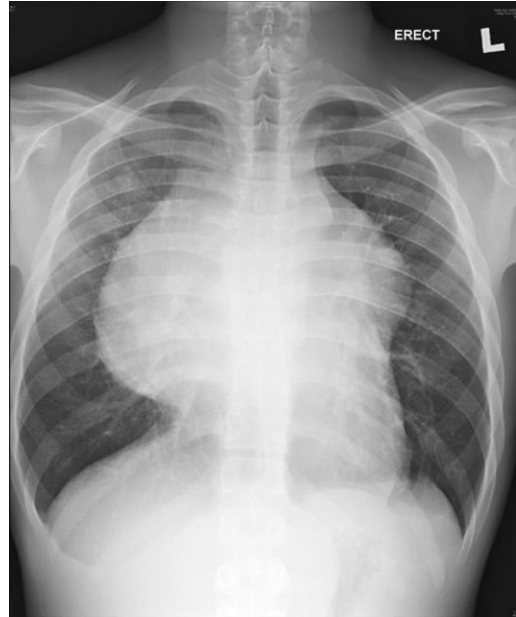
- Standardized Tumor Volume (STV)
 - Measurements by CT
 - Equation of the volume of an ellipsoid

$$\text{STV} = \frac{4}{3}\pi \times \left(\frac{1}{2} \text{ tumor height} \times \frac{1}{2} \text{ tumor width} \times \frac{1}{2} \text{ tumor depth}\right) / \text{body height (in cm)}$$

- Cut-off value = **2.5cm³**



Exacerbation of MMS



M15yr cough for 6 months and dyspnoea for 2 months

Facial swelling after waken up from sleep

SaO₂ 93% in air

Deteriorated on sedation after line access and BM; further deteriorated after intubation.

Emergency operation: mature teratoma

1. Sedation or anaesthesia

- Acute compression of airway or blood vessels
- Supine position
 - Increase intrathoracic pressure + decrease functional residual volume
- Muscle relaxant
 - loss of muscle tone with collapse of airway structures
- Loss of spontaneous ventilation

2. Positive pressure ventilation

- Complete obstruction of major airways or vessels
- Acute right ventricular failure
- Reduce ventricular preload

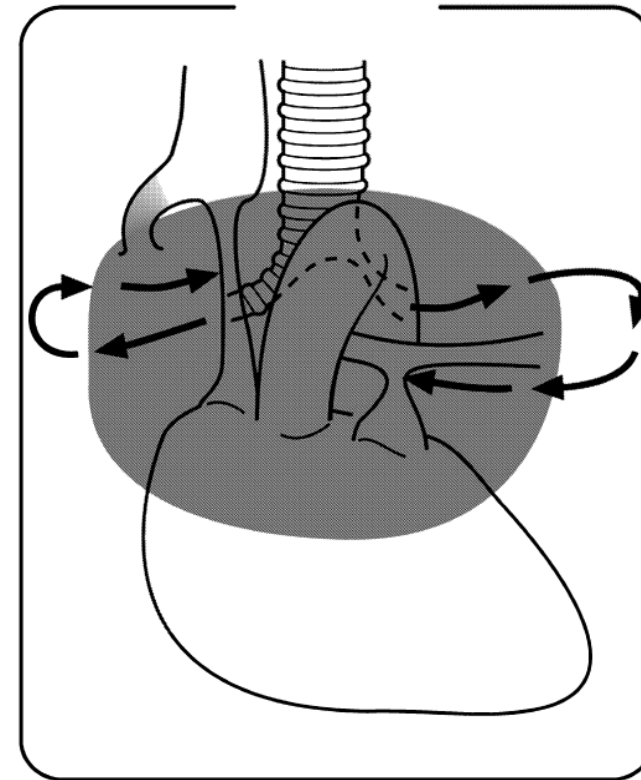
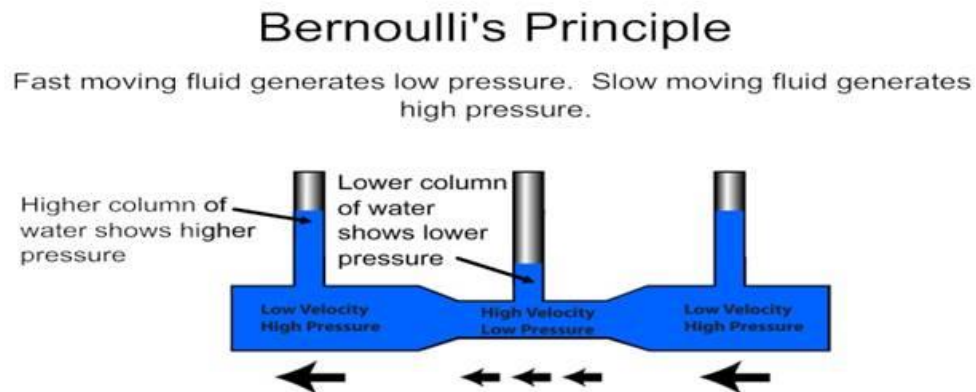


Figure 2
IPPV. Inspiration.

How to avoid?

Do not lie the patient flat!

- Allow child to remain supported on his/her comfortable position
 - a parent's lap, preferably in an upright or semi-upright position,
 - on bed at 45deg



Safe tissue diagnosis

- **Avoid** anesthesia or sedation, wherever possible
- Use of **alternative tissues**
 - Peripheral blood - ALL, germ cell tumor
 - Lymph node biopsy
 - Pleural/ pericardial fluid
- If anaesthesia is necessary
 - maintain **spontaneous breathing**
 - **local anaesthesia**
 - **Sedation technique** over GA technique e.g. ketamine
 - Minimize need of general anesthesia, muscle paralysis, controlled ventilation

“standby” cardiopulmonary bypass

- Previous paper suggest “standby” ECMO during induction of anesthesia
 - RISKY!!!
 - Need at least 5-15mins to cannulate and establish adequate circulation and oxygenation
- If unsafe for induction of anesthesia
 - Better to establish cardiopulmonary bypass **prophylactically**
 - Or cannulate and connect to machine **before induction**

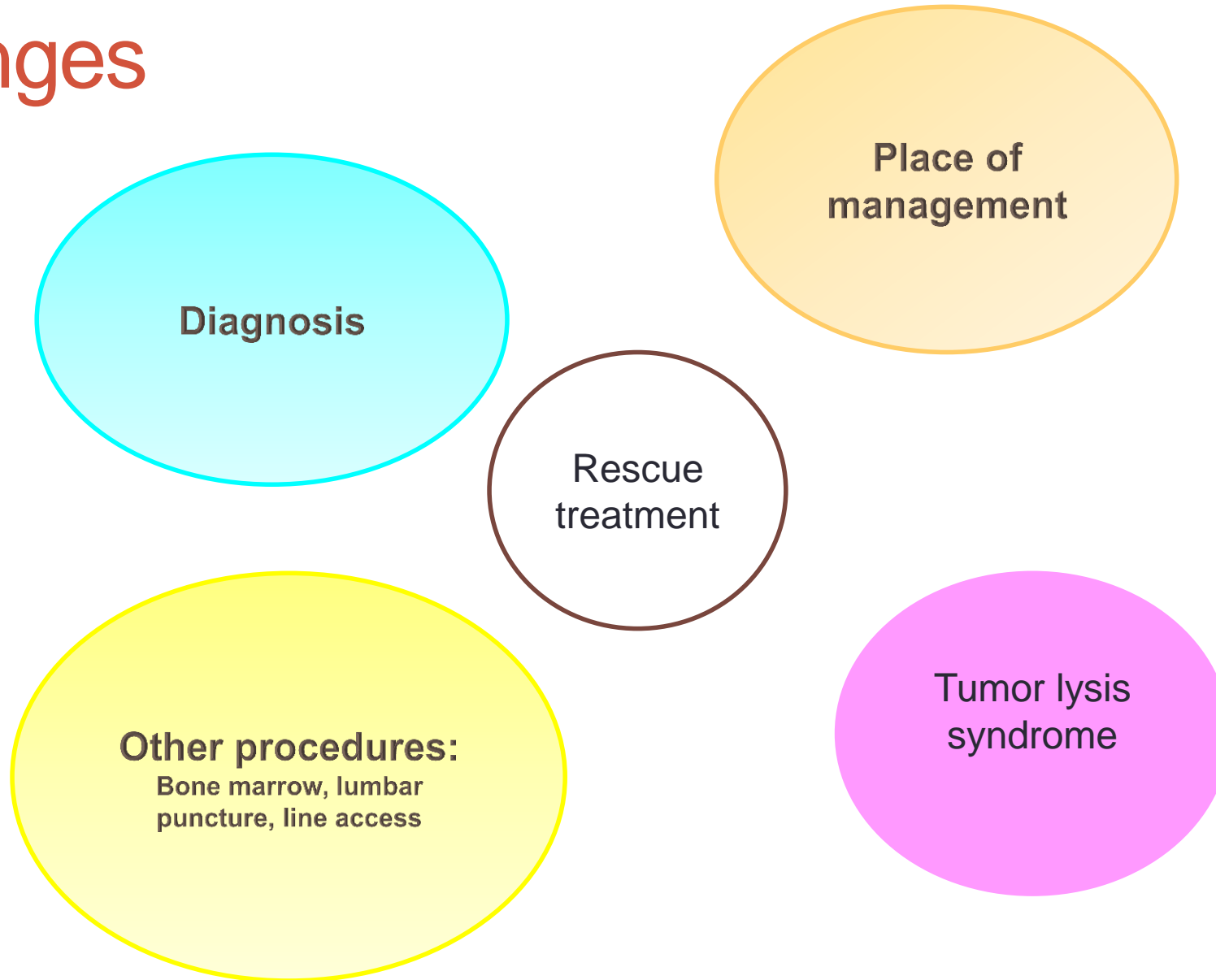
Steroid use

- Pre-diagnostic period - **controversial**
- Most studies recommend against -> interfere with the histological diagnosis
- Steroid has **no impact** on the diagnosis of benign tumor or less steroid sensitive tumor

For suspected lymphoma/leukaemia case, prophylaxis for **Tumor Lysis Syndrome** is important before steroid use

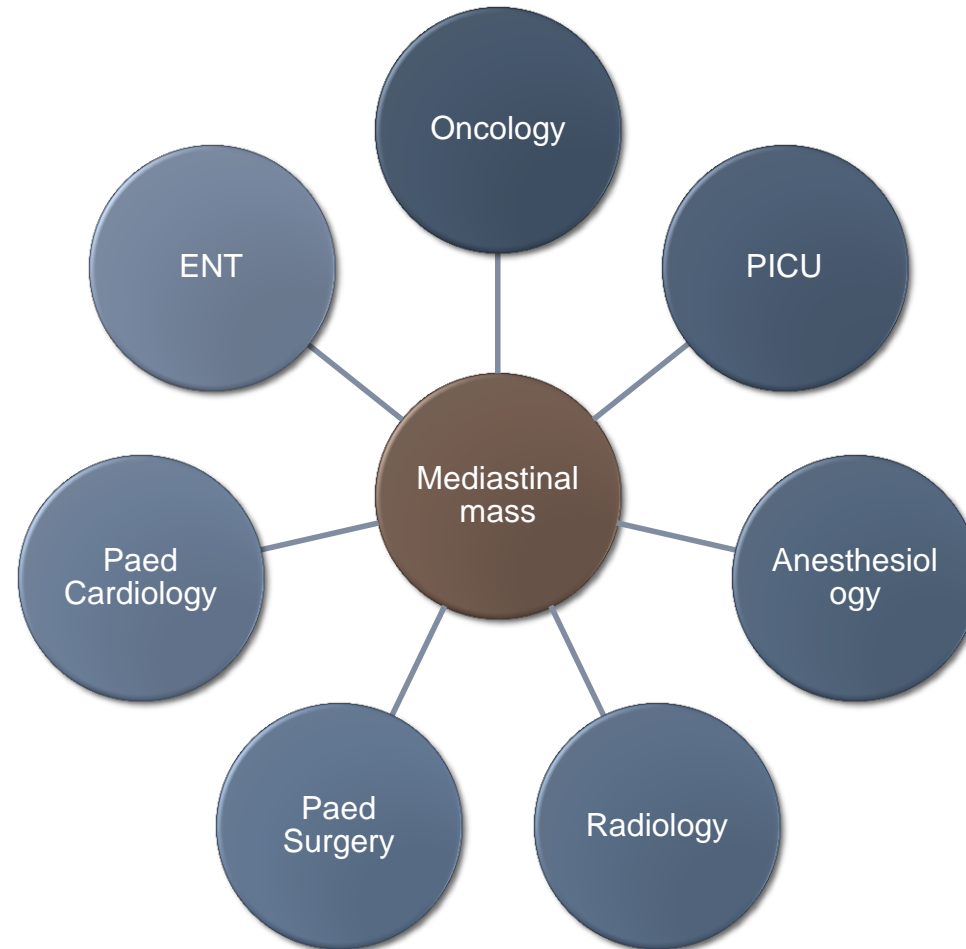
- Close monitoring of electrolytes, urate level, renal function
- Hyperhydration
- Pharmacological agents for hyperuricemia
 - E.g. allopurinol, rasburicase

Challenges

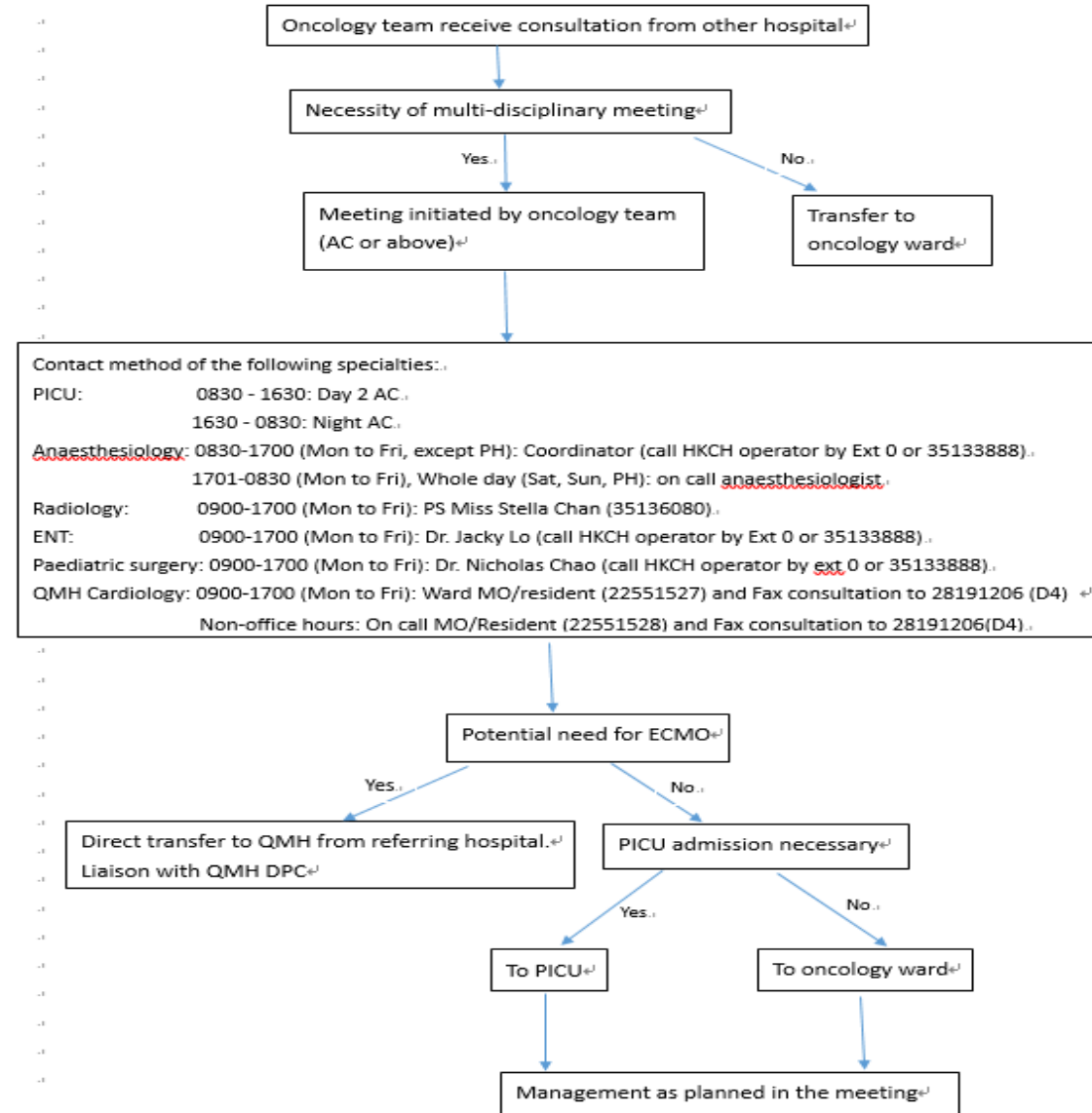


Multi-disciplinary care

- **Case by case** discussion



Flow chart for multidisciplinary meeting for mediastinal mass and upper airway mass



Take home message

- Mediastinal mass is **not uncommon** in our locality
- Mediastinal mass is **clinical emergency** and can cause morbidity or mortality
- Try to **avoid sedation/anesthesia** or **positive pressure ventilation**
- Try to obtain a **safe** tissue biopsy for diagnosis
- Steroid is **not** the key to every mediastinal mass case
- **Tumor lysis syndrome prophylaxis** is important
- Do consult us for any suspected or confirmed case



Thank you



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