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Fifteen Minute Consultation: Obvious and Not-So-Obvious Mediastinal Masses

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15 Minute Consultation: Obvious and Not-So-Obvious Mediastinal Masses

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"Fifteen Minute Consultation: Obvious and Not-So-Obvious Mediastinal Masses"

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Objectives:

To present a structured approach to the management of a child with a mediastinal mass presenting to the emergency department

To raise awareness of presenting features of less-obvious mediastinal masses and to encourage consideration of mediastinal masses in differential diagnoses

Methods:

Review of the relevant literature and review of London Paediatric Cancer Network supportive guidelines and subsequent description of the approach to a child presenting with features suggestive of a mediastinal mass.

Conclusions:

A systematic approach to history taking, clinical examination, and investigation of a child presenting with a mediastinal mass will assist in the safe and timely management of children presenting when they are critically unwell. Anticipation of potential management complications and early transfer for ongoing management will improve patient outcomes and minimise morbidity.

15 Minute Consultation: Obvious and Not-So-Obvious Mediastinal Masses

Introduction

Mediastinal masses in children are a rare but life-threatening presentation. They are the most common chest mass in children, with the anterior mediastinum being the most common site¹, and lymphomas being the most common cause¹⁻³. Whilst some mediastinal masses may present with obvious symptoms and clinical signs and appear obvious on chest x-ray (CXR), such as in figure 1, many do not, as in figure 2 below. Children with anterior mediastinal masses are at significant risk of serious and potentially fatal complications at presentation, most commonly due to extrinsic compression of the airway, and obstruction to the venous return or obstruction of cardiac output. Not infrequently clinical symptoms may not correlate with radiological findings¹⁻⁷. Airway obstruction has been reported in approximately 60% of patients presenting acutely with mediastinal masses¹. Furthermore, one third of asymptomatic patients have a significant reduction in tracheal dimensions seen upon CT scanning⁴, with total consensus that sedation and general anaesthesia should be avoided and managed by experienced paediatric anaesthetists if considered essential¹⁻⁹.

Anterior mediastinal masses are a paediatric medical emergency and should be considered a haematological/oncological emergency until proven otherwise, especially given that lymphomas (the most common cause of a mediastinal mass) can rapidly grow and progress over hours to days. Children should be managed by experienced paediatricians with a low threshold for escalating to paediatric intensive care.

Obvious and Not-So-Obvious Mediastinal Masses

Case 1: An 'Obvious' Mediastinal Mass

Figure 1- A clinical case of 'obvious' mediastinal mass seen on CXR presenting to the emergency department.

Case 2: A 'Not-So-Obvious' Mediastinal Mass

Figure 2- A clinical case of a 'not-so-obvious' mediastinal mass seen on CXR presenting to the emergency department.

THINK! Could you have mistaken Figure 2 for a pleural effusion?

Mediastinal masses often present atypically with a homogenous 'white-out' on CXR, and can easily be mistaken for more common diagnoses such as pleural effusions or a large opacification ie. pneumonia. Although more uncommon, always consider a mediastinal mass in an unwell patient with a 'white-out' on CXR, especially where there is a lack of classical symptoms of infection. Remember, a widened mediastinum on CXR cannot be attributed to normal thymus in children over 5 years.

History

All medical consultations should commence with a detailed paediatric history. This may immediately identify symptoms of a mediastinal mass, airway compromise or life-threatening neurological or cardiovascular compromise. More specific symptoms that may suggest a mediastinal mass are listed below in figure 3, but it is not uncommon for a mediastinal mass to be a surprise finding on CXR as they are often not as obvious at presentation as may be expected.

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7 **Figure 3- Key presenting symptoms of a child with mediastinal mass**
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20 **Examination**
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23 Respiratory signs are the most common presentation of a mediastinal mass, with up to 60% paediatric patients
24 presenting with airway compromise.
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28 In a life-threatening situation, children should be managed using an airway, breathing, circulation,
29 disability/neurology approach as per APLS.
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33 Mediastinal masses may present obviously or inconspicuously depending on the size and origin of the mass. A
34 thorough full-systems paediatric examination should be performed, with specific attention given to the respiratory,
35 cardiovascular, ear nose and throat (ENT) and neurological systems; carefully assessing for evidence of airway
36 compromise, superior vena cava obstruction, cardiac or neurological compromise seen in less-obvious mediastinal
37 masses.
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45 The key obvious and less-obvious presenting signs of mediastinal masses are described in figure 4 below.
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48 It is not unusual for mediastinal masses (typically T-cell lymphoma or leukaemia) to be associated with pleural
49 and/or pericardial effusions, thus making the underlying mediastinal mass harder to identify. Such pleural effusions
50 may be bilateral and may further compromise the child's respiratory status, and should increase the clinicians
51 suspicions of what may lie underneath.
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17 **Figure 4-** *Obvious and less obvious signs on clinical examination suggesting a mediastinal mass*
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22 **Differentials of Paediatric Mediastinal Mass**

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26 There are a wide range of differential diagnoses for a child with a mediastinal mass, as summarised in figure 5.
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29 **Figure 5-** *Differential diagnoses of a mediastinal mass in a child*
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41 **Causes of a homogenous 'White Out' or homogenous opacity on CXR**

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- Chest wall mass, e.g. Ewing's sarcoma
 - Consolidation
 - Diaphragmatic hernia
 - Diaphragmatic rupture
 - Mediastinal mass
 - Pleural effusion

- Pneumonectomy
- Pulmonary agenesis or hypoplasia
- Total lung collapse, eg. post endobronchial intubation

Investigations

Investigations in a child with mediastinal mass will be directed primarily by how unwell the child is and by the continuous assessment of the child's airway status and any sign of airway, respiratory or neurological compromise. It is essential not to compromise the child's safety to pursue clinical investigations.

The delivery of sedation or anaesthesia for a child with an anterior mediastinal mass can be fatal. This is due to the loss of muscle tone and the subsequent loss of the compensatory physical effort of maintaining the airway which occurs when sedated or unconscious. This process can result in an acute loss of airway patency which can be fatal even in the hands of an experienced anaesthetist. For this reason children with a suspected mediastinal mass should undergo sedation or anaesthesia only under the care of an experienced paediatric anaesthetist fully aware of the child's situation, and if no other option is available.

As demonstrated in figure 6, there are surprisingly few essential investigations required to permit commencing emergency treatment for a mediastinal mass where haematological malignancy is suspected.

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11 **Figure 6- Recommended investigations and guidance on where and when to perform them in a child with suspected**
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13 *mediastinal mass*

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19 Essential Diagnostic Investigations for Mediastinal Masses

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- 22 • A **baseline full blood count** should be performed and may indicate an underlying diagnosis of acute
23 lymphoblastic leukaemia.
 - 24 • A **blood film** should be performed to investigate for the presence of blasts.
 - 25 • **Baseline bloods**- urea and electrolytes, urate, phosphate, liver function tests, and a coagulation screen should
26 be performed prior to commencing treatment.
 - 27 • **CXR** - erect anteroposterior (AP) and lateral CXRs should be performed to establish the size and location of the
28 mediastinal mass and are adequate imaging prior to commencing emergency treatment in a child too unstable
29 to have a CT chest at presentation. Figure 7 illustrates the anatomical division of the mediastinum on CXR.
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58 **Figure 7 - Anatomical division of anterior, superior, middle and posterior mediastinum on lateral CXR**
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- **CT chest**, in patients without concerns for airway compromise and who are able to lie flat safely, is a helpful diagnostic investigation. However, where there are concerns for potential acute airway compromise and in patients who are unable to lie flat safely, cross-sectional imaging is not required as an essential investigation and should not be performed. In general, imaging should be performed without sedation or general anaesthesia to prevent potentially life-threatening airway compromise, and can be combined with other diagnostic procedures such as surgical biopsy or central line insertion at a later stage if unsafe to perform at presentation.
- **Ultrasound chest** is considered an essential investigation when a CT chest is not possible or where there is suspicion of a large pleural effusion, to further assess the mediastinum for potential tumour and the volume of a pleural effusion which may require emergency tapping prior to transfer or further treatment. If pleural fluid is tapped, it is advisable to collect samples for histology and haematology cytospin. However, the drainage of large volumes of pleural fluid is not advisable due to the risk of expansion oedema and subsequent respiratory collapse.

Management:

In a life-threatening situation, children should be managed using an airway, breathing, circulation, disability/neurology approach as per APLS. All cases, especially those with life-threatening mediastinal masses, must be discussed urgently with the attending paediatric consultant.

Children with an anterior mediastinal mass are at risk of sudden and acute respiratory collapse and can deteriorate very rapidly; even if looking clinically well at presentation. The management of these patients is often very difficult and it is therefore vital to have a very low threshold to seek senior support during the initial management period.

Children should be transferred urgently to the tertiary haematology/oncology centre if a mediastinal mass is seen on imaging and/or there is evidence of respiratory distress. Intensive care retrieval to PICU should be considered early.

The specific management of acute lymphoma and leukaemia (the most common causes of a mediastinal mass) is beyond the scope of this article. However, steroids are the emergency treatment of choice for a malignant mediastinal mass, but must not be commenced without careful preparation and discussion with the tertiary centre.

1 Whilst steroids may be commenced prior to a definitive histological tissue diagnosis in children at significant risk of
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3 airway compromise or superior vena cava obstruction, it must be considered that steroids can impair the quality of
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5 diagnostic material sampled upon biopsy, with important implications for longer-term treatment, hence a need for
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7 discussion with the tertiary centre prior to their prescription. In addition, there is a significant risk of rapid onset
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9 tumour lysis following the initiation of steroid treatment, where the rapid destruction of the bulky mass (particularly
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11 if T cell malignancy is suspected) can be fatal and requires aggressive prevention and management with intravenous
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13 fluid hyper-hydration and concomitant allopurinol or rasburicase (where available). The management of an anterior
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15 mediastinal mass where a haematological underlying cause is suspected is summarised in figures 8 and 9 below.
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27 **Figure 8 - Management of an anterior mediastinal mass in a child in the paediatric emergency department**
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6 Key Management Do's and Don't's:
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14 **Figure 9-** *Key management 'dos' and 'don'ts' for a child with suspected or proven anterior mediastinal mass*
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20 Take Home Conclusions
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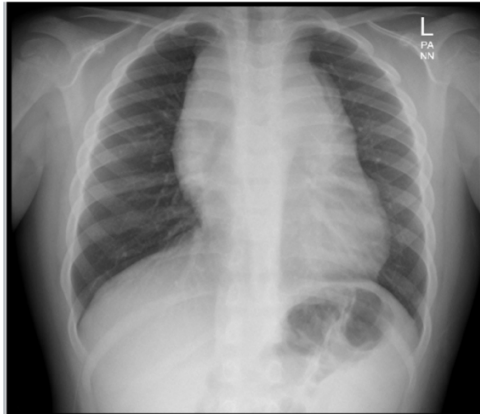
- 23 • Mediastinal masses are an emergency presentation and can cause life-threatening airway compression.
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- 26 • Simple investigations with CXR and blood tests may be sufficient to make a preliminary diagnosis at the
27 presenting hospital.
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- 31 • Staging and further diagnostic work-up, including cross sectional imaging, biopsy of the mass and bone
32 marrow sampling should usually be undertaken in the tertiary centre.
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- 37 • Steroids are the emergency treatment of choice for children with suspected lymphoma or leukaemia.
38 However, these should only be started after discussion with the tertiary centre and with careful preparation
39 given the high risk of tumour lysis syndrome.
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- 45 • Some mediastinal masses are truly asymptomatic.
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15 [use-before-this-date.pdf](http://www.londoncancer.org/media/81610/Supportive-Care-Protocol-4th-Edition-v1.0-Active-from-1st-July-2014-NOT-for-clinical-use-before-this-date.pdf)
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Case 1.
9 year old boy presented to A&E with a 6 week history of progressive difficulty breathing, orthopnoea, and neck swelling. No past medical history. The child was born uneventfully at term.

On examination the child had obvious stridor from airway compromise but without overt tracheal deviation. He had severe respiratory distress with tachypnoea, marked subcostal and intercostal recessions and tracheal tug with diffuse wheeze auscultated throughout the chest. There was neck swelling but no facial swelling, and no signs of neurological or cardiovascular compromise.

The child had an urgent CXR which showed a clearly enlarged mediastinum. He deteriorated very rapidly and was transferred urgently to paediatric intensive care for ventilatory support and commencing steroid treatment. It was not safe to perform a CT scan at the initial referral point- CXR and Ultrasound were imaging modalities used to make the initial diagnosis as both could safely be performed without lying the patient flat and without sedation.

A diagnosis of T-cell Acute lymphoblastic leukaemia was subsequently made. The child developed tumour lysis syndrome very shortly after commencing steroid treatment, requiring hyperhydration and Rasburicase therapy with forced diuresis in PICU.

Key Learning Points:

- Children with bulky mediastinal masses can deteriorate very rapidly
- Steroid therapy should not be started outside of the tertiary treatment centres due to the risk of rapid onset of tumour lysis syndrome which can be fatal and often requires intensive care
- A CT scan is not an essential investigation required at the point of assessment and stabilisation and should not be performed if the child is unwell or is at risk of sudden respiratory or cardiovascular collapse. A CXR +/- Ultrasound are sufficient and safer investigations until the child is stable enough to lie flat.

Figure 1- A clinical case of 'obvious' mediastinal mass seen on chest X-ray presenting to the emergency department.

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	<p>Case 2.</p> <p>An 8 year old boy presented to paediatric A&E with a 2 week history of wheeze with worsening dyspnoea over the preceding 72 hours. The child reported a 6 week history of chronic, non-productive cough; worse when lying flat. The child had been seen twice previously by his GP and prescribed salbutamol at the first visit and then oral prednisolone at the second, due to no benefit from bronchodilators alone. The child had no past medical history, was born at term by emergency caesarean section and was fully vaccinated, with no significant family history.</p> <p>On examination the child had tachypnoea with saturations of 90% in room air. Significant respiratory distress was seen with nasal flaring, subcostal and intercostal recession, tracheal tug, left-sided reduced air entry and a pleural friction rub. No crackles or wheeze auscultated. Abdomen was soft and non-tender with no masses or organomegaly. No lymphadenopathy was palpated and examination was otherwise unremarkable.</p> <p>CXR revealed a white-out of the left hemithorax with an associated meniscus, indicating a large effusion, and with mediastinal shift towards the right hand-side. By ultrasound (indicated by what was initially suspected to be an isolated pleural effusion) the anterior mediastinal mass was subsequently identified. A CT chest was not performed at presentation due to difficulty lying flat with a risk of airway compromise. Haemoglobin 116, white cells 14.7, platelets 367, urine and liver function tests unremarkable.</p> <p>The child was electively intubated due to risk of airway compromise, an emergency chest drain inserted and 1.6L pleural fluid drained before being transferred to the nearest tertiary oncology centre. The child was suspected to have lymphoma and was commenced on emergency treatment for this including intravenous antibiotics, dexamethasone and emergency chemotherapy. Risk of tumour lysis syndrome was managed with intravenous hyper-hydration fluids and Rasburicase. Biopsy of the mediastinal mass revealed a T-Cell Non-Hodgkin lymphoma.</p>
<p>Key Learning Points:</p> <ul style="list-style-type: none"> • It would be unusual for a school-aged child with no personal history of atopy to present with an exacerbation of asthma for the first time with no response to standard asthma treatment. • Children with bulky mediastinal masses are at significant risk of tumour lysis syndrome which can be precipitated by steroids. • This child had a very large mediastinal mass which was obscured by the associated large pleural effusion. In this case, it was the history which was crucial in suggesting an alternate diagnosis, and the ultrasound which allowed safe further radiological investigation of the underlying pleural effusion. A CT chest was not an essential investigation for the initial diagnosis and management of this child, and to the contrary would have compromised the patient's safety if performed at presentation. 	

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Figure 2- A clinical case of a 'not-so-obvious' mediastinal mass seen on chest X-ray presenting to the emergency department. Please note that this is not the initial presenting CXR but that taken on arrival at the tertiary oncology centre in PICU.

Key Symptoms of a Mediastinal Mass

- Cough (exacerbated lying flat)
- Chest pain
- Progressive Dyspnoea (especially without history of asthma/inter-current illness)
- Orthopnoea or sleeping with more pillows
- Difficulty Swallowing or choking when eating
- Reduced exercise tolerance (acute exertional breathlessness)
- Symptoms indicating Superior Vena Cava Obstruction- Swelling of the face/ head/neck
- Neurological symptoms indicating neurological compromise- headache, dizziness or confusion
- Red flag symptoms of haematological malignancy indicating the cause of mediastinal mass- Bone pain, easy bruising, weight loss

Figure 3- Key presenting symptoms of a child with mediastinal mass

More Commonly Associated findings with Mediastinal Mass	Less Commonly Associated Findings with Mediastinal Mass
Stridor or difficulty self-maintaining airway	Signs of mediastinal shift: Tracheal deviation, heart sounds heard loudest over mediastinum or right side of chest
Progressive Respiratory Distress: <i>Tachypnoea, nasal flaring, recessions, unilateral reduced or absent air entry, difficulty speaking in full sentences, hypoxia, cyanosis</i>	Signs of pleural effusion: <i>Reduced air entry, inability to lie flat, dry cough, pleurisy</i>
Unilateral Wheeze	Signs of Superior Vena Cava Obstruction: <i>Swelling of the face/head/neck or more prominent or engorged superficial head and neck veins</i>
Diffuse or Significant Lymphadenopathy	Signs of neurological compromise: <i>Confusion, syncope, reduction in Glasgow Coma Scale (GCS) Score</i>
Exertional Breathlessness with movement <i>(be aware of the patient being more 'still' than is appropriate for age)</i>	Signs of cardiac compromise (from obstructed venous return): <i>facial oedema or plethora, pulsus-paradoxus, tachycardia, engorged and distended head and neck veins.</i>
Inability to lie flat (Orthopnoea)	Signs of acute haematological malignancy: <i>Bone pain affecting gait or weight bearing, cachectic appearance, organomegaly, bruising, petechiae.</i>

Figure 4- More and less commonly associated findings with a mediastinal mass

Anterior Mediastinal Mass	Middle Mediastinal Mass	Posterior Mediastinal Mass
Lymphoma (<i>T-cell lymphoma, Hodgkin's lymphoma, Anaplastic large cell lymphoma, B cell lymphoma</i>)	Foregut duplication cysts, eg. <i>Bronchogenic cyst</i>	Thoracic Neuroblastoma
Leukaemia (<i>T-cell leukaemia</i>)	Tuberculosis	Neurofibroma
Solid Tumour (<i>Germ Cell Tumour, Teratoma</i>)	Fungal Chest Infection	Extra-medullary haematopoiesis
Thymic Cyst	Vascular Malformation	Vascular Malformation
Enlarged Thymus	Lymphadenopathy	
Tuberculosis		

Figure 5- Differential Diagnoses of a Mediastinal Mass in a child

Investigation	Essential Prior to Commencing Emergency Treatment	Essential Diagnostic, Additional Diagnostic, or Staging	Rationale for completing	Location to perform
Full Blood Count (FBC) & Blood Film	Yes	Essential Diagnostic	May indicate underlying diagnosis of acute lymphoblastic leukaemia or lymphoma	Presenting Hospital
Baseline Blood Tests (Urea & Electrolytes, Urate, Phosphate, Liver Function tests, Coagulation Screen, Group and Save).	Yes	Essential Diagnostic	Baseline bloods and assessment of renal and hepatic function required prior to commencing treatment.	Presenting Hospital
Erect Chest X-Rays (AP and Lateral Views)	Yes	Essential Diagnostic	Identification of a mediastinal mass (Size, Location). Adequate imaging prior to commencing treatment in an unstable child.	Presenting Hospital
Additional Blood Tests (Lactate Dehydrogenase (LDH), Alpha Feto Protein (AFP), and serum BHCg)	No	Additional Diagnostic	Exclusion of non-haematological diagnoses	Presenting or Tertiary Hospital
Ultrasound Chest	No	Additional Diagnostic <i>Essential Diagnostic if CT Chest unsafe to perform</i>	Evaluation of underlying pleural effusion. Further assessment of mediastinal mass when CT Chest unsafe to perform	Presenting or Tertiary Hospital
Unsedated CT Chest	No	Essential Diagnostic providing patient can lie flat safely. CT Chest NOT to be performed under sedation or general anaesthetic	Investigation of a mediastinal mass / white-out on CXR. <i>Although CT Chest is considered an essential diagnostic investigation, it is not required to commence emergency treatment and must be performed awake if prior to starting treatment due to risk of airway and respiratory compromise with sedation/anaesthesia.</i>	Presenting or Tertiary Hospital
Pleural Tap	No	Additional Diagnostic	Emergency drainage of life-threatening pleural effusion. <i>If chest drain insertion required, collect pleural fluid for histology and haematology cytospin. Ideally discuss with haematologist pre-emptively.</i>	Presenting or Tertiary Hospital
Echocardiogram	No	Essential Diagnostic	Investigation of potential Superior Vena Cava Obstruction and Pericardial Effusion	Tertiary Hospital
Lumbar Puncture	No	Staging	Evaluation of Central Nervous System (CNS) Involvement if underlying haematological malignancy	Tertiary Hospital
Bone Marrow Aspirate	No	Staging	Staging investigation for haematological cause of mediastinal mass	Tertiary Hospital
Urine Catecholamines (Posterior mediastinal mass only)	No	Additional Diagnostic	Excludes thoracic neuroblastoma as a cause of posterior mediastinal mass	Presenting or Tertiary Hospital

Figure 6- Recommended investigations and guidance on where and when to perform them in a child with suspected mediastinal mass

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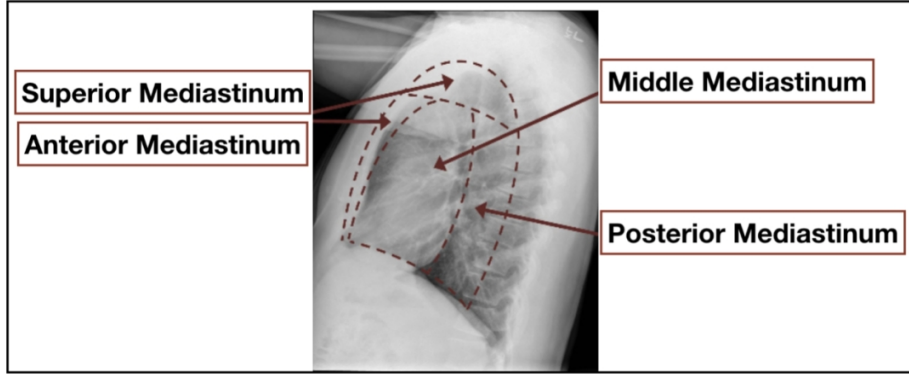


Figure 7- Anatomical division of anterior, middle and posterior mediastinum on lateral chest X-ray

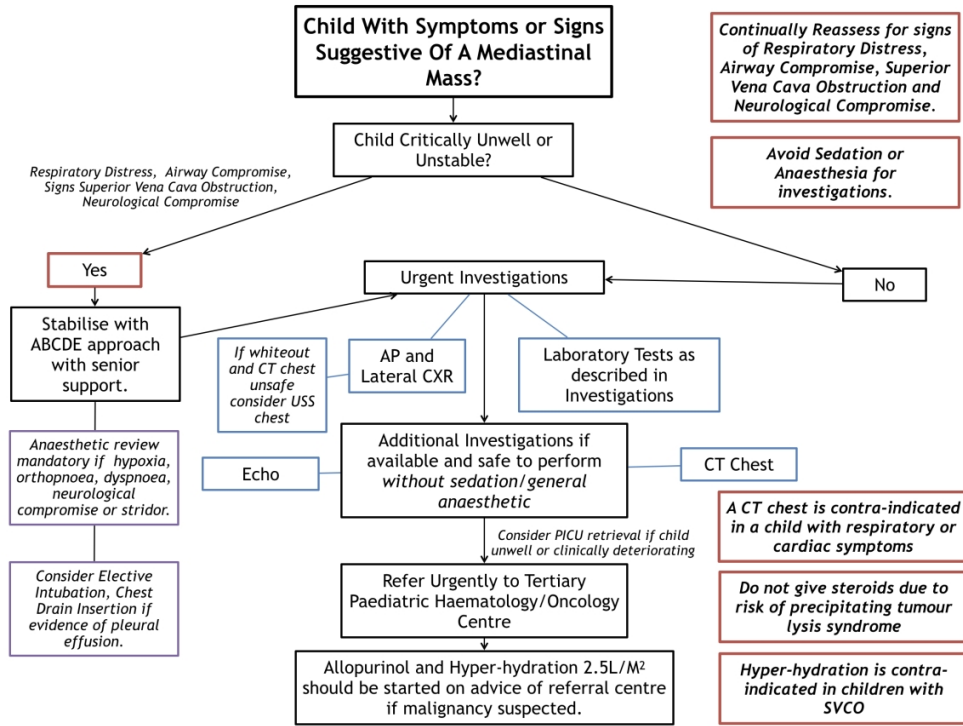


Figure 8- Management schema for a child with Anterior Mediastinal Mass in the paediatric emergency department

DO	DON'T
Manage and stabilise the child using an ABCDE approach as a priority over diagnostic investigations	Do Not give hyper-hydration to a child with signs of Superior Vena Cava Obstruction as this can lead to cerebral oedema and death.
Stabilise and transfer early to the tertiary treatment centre with a low threshold for early PICU retrieval.	Do Not commence steroid treatment without discussion with a tertiary paediatric haematologist, as steroids may precipitate life-threatening tumour lysis syndrome in bulky mediastinal masses.
Avoid Sedation / Anaesthesia for imaging and diagnostic procedures due to the high risk of airway compromise and respiratory arrest.	Perform a CT Chest in a child with respiratory or cardiac compromise
Continually Reassess the child for signs of airway compromise, respiratory distress, superior vena cava obstruction, and neurological compromise.	Struggle to manage a child with a mediastinal mass alone. This is a haemato-oncological emergency until proven otherwise and requires experienced senior supervision due to the risk of rapid deterioration and cardio-respiratory arrest.

Figure 9- Key Management 'Dos' and 'Don'ts' for a child with suspected or proven anterior mediastinal mass