

Guideline for management of patients with suspected anterior mediastinal mass

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INTRODUCTION

Mediastinal masses in children range from benign to malignant, with varying presentations and outcomes. Anterior mediastinal masses have potential for severe and life-threatening cardio-pulmonary compromise because of its impact on surrounding mediastinal structures such as the airway, heart, and great vessels.

While there is no single individual symptom or test that will predict risk of life-threatening complications, certain symptoms and signs may predict the increase in risk of undergoing general anaesthesia. Mediastinal masses present mainly with respiratory symptoms and may mimic common conditions such as asthma or croup. They may also present with SVC syndrome and superior mediastinal syndrome. The mass can also exert different effects depending on the patient's position- eg. orthopnoea can be associated with narrowing of trachea caused by combined effects of gravity in supine position and increasing intrapleural pressure at the end of expiration. Compression of great veins can also reduce cardiac preload which can result in a significant drop in cardiac output. Malignant mediastinal masses can also be associated with pleural or pericardial effusions, which can further exacerbate cardio-respiratory function.

Diagnostic work up and obtaining histological diagnosis can be challenging in this group of patients owing to the major risk of cardiopulmonary arrest following general anaesthesia.

A personalized multi-disciplinary approach with close collaboration between paediatric anaesthetists, intensivists, oncologists, and surgeons is essential to improve outcomes and avoid preventable complications.

Superior mediastinal syndrome	<p>Obstruction of airways (trachea, main bronchi) and/or cardiac or great thoracic vessel compression, usually from mediastinal mass.</p> <p>Manifests as: stridor, dyspnoea/tachypnoea, orthopnoea, wheeze, cyanosis, hypoxia, cardiac compromise/arrest</p> <p>This is a medical emergency</p>
SVC syndrome	<p>Onset can be insidious but can lead to sudden loss of cardiac output Can lead to rapid increases in ICP, with resulting cerebral edema</p> <p>Manifests as: Head and neck swelling and plethora, distended neck veins, Pemberton sign, syncope</p>

Investigations

BEWARE: inappropriately requested investigations may result in worsening respiratory compromise, rapid clinical deterioration, and cardiorespiratory arrest.

CXR	Include AP and lateral to establish size and whether mass is anterior or posterior
CT	<ul style="list-style-type: none"> Only perform if safe for patient Avoid sedation/GA if significant mass on CXR and/or significant respiratory symptoms-> this may precipitate respiratory failure leading to death If orthopnoea present- chest CT in supine position should be avoided. Consider alternative positions for scanning if patient can tolerate such as prone/lateral. To discuss with radiologist consultant. Determine degree of airway and/or great vessel compromise/compression
2DE	To discuss with paediatric cardiologist To consider if: <ul style="list-style-type: none"> Postural respiratory symptoms present Cardiomegaly (to exclude pericardial effusion) To exclude direct tumor invasion of pericardium or external atrial compression
Chest USS	If unable to perform chest CT, to discuss role of chest/mediastinal USS
Lab investigations	Initial investigations include: <ul style="list-style-type: none"> FBC, PBF, UECr, uric acid, LDH, AFP, HCG PT/PTT, GXM Urine VMA/Creatinine and HVA/creatinine (for posterior mediastinal mass) Consider monitoring for tumor lysis syndrome (discuss with oncologist)

Risk stratification for patients with anterior mediastinal mass

Risk category	Criteria	Recommendation guidelines
Low	Clinical: Asymptomatic Radiological: No evidence of significant compression of structures	Recommendations <ul style="list-style-type: none"> Close monitoring of clinical status including RR and saturations and evolving clinical symptoms. Administration IPPV likely to be safe, but still at risk of severe complications-> to discuss use of sedation/anaesthesia with anaesthetists
Intermediate	Clinical: Mild to moderate postural symptoms Radiological: Mild tracheal compression ($\leq 50\%$ cross-sectional area), small pleural effusion, small pericardial effusion	Mandatory Close monitoring of clinical status including RR and saturations Recommendations <ul style="list-style-type: none"> Consider HD/ ICU admission Consider large bore IV access in lower limb Maintenance of spontaneous

		ventilation desirable
High	<p>Clinical: Acute airway compromise: severe postural symptoms (orthopnoea), dyspnea, marked tachypnea, stridor, cough, wheezing, cyanosis, chest pain</p> <p>SVC syndrome (plethora, orbital suffusion, upper body oedema and/or prominent veins, neurological symptoms (dizziness, lethargy, stupor, impaired consciousness, seizures)</p> <p>Radiological: Airway - moderate to severe tracheal compression (>50% cross-sectional area) - tracheal deviation - main bronchial compression</p> <p>Vascular - compression of great vessels (esp. SVC, PA) - moderate to large pericardial effusion - Impaired ventricular function EF < 35% - mediastinal mass ratio >45%</p>	<p>Mandatory</p> <ul style="list-style-type: none"> • Maintain patient's position of comfort • Transfer to ICU for close monitoring • Secure large bore intravenous access in lower limb • Spontaneous ventilation should be maintained, avoid sedation • Standby rigid bronchoscope <p>Recommendations</p> <ul style="list-style-type: none"> • Consider pre-operative steroids or urgent chemotherapy/radiotherapy • Consider standby ECMO <p>If impending respiratory collapse:</p> <ul style="list-style-type: none"> • consider early referral / airway code for elective intubation • consider intubate past obstruction without paralysis • consider stent airway with rigid bronchoscope • activate CICU consultant for KIV ECMO <p>Resuscitation plan If cardiorespiratory decompensation:</p> <ul style="list-style-type: none"> - consider lateral position CPR - activate CICU for KIV ECMO

Management

- Do not force the patient to lie flat. In a situation of airway compromise, airway patency is best obtained in a semi-elevated, forward sitting, prone or lateral position, depending on the exact location of the mass in relation to the great vessels and airway.
- Avoid IV access in upper limb if evidence of SVC obstruction in view of risk of exacerbation of facial swelling and cerebral edema.
- **Consult oncologist, paediatric surgeons, anaesthetist, and intensivists immediately to discuss management plans for patient. A physician conference or group huddle is recommended.** Note that these conversations may be staged as more information becomes available (see "pre-operative huddle")
- Sedation is not recommended even if child is agitated.
- If pleural fluid drainage is needed, we recommend no more than 10mL/kg body weight (max

absolute of no more than 1L) to be drained at one setting.

- **Low threshold for CICU admission for close monitoring.**
- Systemic steroids can rapidly reduce size of a mass but significantly alters tissue quality, which can delay accurate diagnosis on biopsy and staging. Decision for systemic steroids should involve discussion with paediatric oncologist with multi-disciplinary inputs.

Pre-operative huddle:

- **Include: oncologist, paediatric surgeon, anaesthetist, interventional radiology, intensivist, OT nurses, ENT, CTS +/- cardio**
- Determine if procedure is high urgency
- Discuss location of biopsy site and established order of biopsy preference: eg. Bone marrow aspirate > pleural effusion > palpable lymph node > mediastinal mass biopsy
- If pericardial tamponade present, to discuss options- CTS for pericardial window vs cardio for pericardiocentesis
- Discuss venous access strategies: PICC versus central line
- Discuss need for standby blood products
- Ensure special equipment and expertise available:
 - Rigid bronchoscopy
 - Need for ECMO (especially if LVEF decreased) : CTS surgeon, intensivist, ECMO specialists, perfusionists, ECMO circuit on standby
- Oncology recommendations re: steroids, chemotherapy, radiotherapy

Anterior mediastinal mass suspected

CXR

Assess for high-risk factors:

Symptoms and signs

Airway compromise:

- Stridor
- Dyspnoea
- Orthopnoea
- Cyanosis/ hypoxia
- wheezing
- chest pain

SVCO:

- Facial plethora
- Orbital/upper body edema
- Distended neck veins
- Neurological s/s of cerebral edema

Others:

- Pleural effusion
- Pericardial effusion

YES

- Recommend HD/CICU admission
- To admit to CICU if:
 - Severe postural respiratory symptoms or evidence of significant airway compromise
 - Severe SVCO s/s
 - Moderate to severe tracheal compression (>50% cross sectional area)
 - Main bronchial compression
 - Compression of great vessels
 - Mod to large pericardial effusion
 - Mediastinal mass ratio > 45%

NO

- Consider HD admission
- Close monitoring of clinical status including respiratory rate and saturations and evolving clinical symptoms.

Activate urgent multi-disciplinary discussion with paediatric surgeons, anaesthetists, oncologists, intensivists, ENT, CTS and OT staff for planning of investigations and procedures (eg. CT thorax, 2DE, blood tests, pleural/pericardial drainage as indicated).

Evidence of severe airway compromise or compression of great vessels

YES

- Nurse in position of comfort
- Avoid sedation
- Transfer to CICU if not already done
- Secure large bore IV access in lower limb
- Consider need for intubation, with ENT and anaesthesia input
- Consider pre-operative steroids or urgent chemo/radiotherapy
- **Consider standby ECMO (as part of procedural planning and in event of acute cardio-respiratory compromise) -> involve CTS surgeon and ECMO team**

NO

- Assess fitness for GA
- Consider GW monitoring

In the event of respiratory or cardiorespiratory collapse
Resuscitation plan

Activate airway code

Consider lateral position for CPR

Activate CICU consultant for KIV ECMO within 5 min of cardiac arrest

Annex: Summary of published evidence supporting recognized risk factors for adverse clinical outcomes in acute presentation of mediastinal masses in children

Risk factor	Published supporting evidence
Tracheal cross-sectional area compression >50%	<ul style="list-style-type: none"> Shamberger RC, Holzman RS, Griscom NT, Tarbell NJ, Weinstein HJ. CT quantitation of tracheal cross-sectional area as a guide to the surgical and anesthetic management of children with anterior mediastinal masses. <i>J Pediatr Surg</i>. 1991 Feb;26(2):138-42. doi: 10.1016/0022-3468(91)90894-y. PMID: 2023069. King DR, Patrick LE, Ginn-Pease ME, McCoy KS, Klopfenstein K. Pulmonary function is compromised in children with mediastinal lymphoma. <i>J Pediatr Surg</i>. 1997 Feb;32(2):294-9; discussion 299-300. doi: 10.1016/s0022-3468(97)90197-4. PMID: 9044140. Azizkhan RG, Dudgeon DL, Buck JR, Colombani PM, Yaster M, Nichols D, Civin C, Kramer SS, Haller JA Jr. Life-threatening airway obstruction as a complication to the management of mediastinal masses in children. <i>J Pediatr Surg</i>. 1985 Dec;20(6):816-22. doi: 10.1016/s0022-3468(85)80049-x. PMID: 4087108. Reddy CSK, Phang DLK, Ng ASB, Tan AM. A simplified approach for anaesthetic management of diagnostic procedures in children with anterior mediastinal mass. <i>Singapore Med J</i>. 2020 Jun;61(6):308-311. doi: 10.11622/smedj.2019139. Epub 2019 Nov 4. PMID: 31680177; PMCID: PMC7905131. Halepota HF, Tan JSK, Reddy SK, Tang PH, Ong LY, Lee YT, Chan MY, Soh SY, Chang KTE, Ng ASB, Loh AHP. Association of anesthetic and surgical risk factors with outcomes of initial diagnostic biopsies in a current cohort of children with anterior mediastinal masses. <i>World Jnl Ped Surgery</i> 2021;4:e000303.doi:10.1136/wjps-2021-000303 Angheliescu DL, Burgoyne LL, Liu T, Li CS, Pui CH, Hudson MM, Furman WL, Sandlund JT. Clinical and diagnostic imaging findings predict anesthetic complications in children presenting with malignant mediastinal masses. <i>Paediatr Anaesth</i>. 2007 Nov;17(11):1090-8. doi: 10.1111/j.1460-9592.2007.02279.x. PMID: 17897276; PMCID: PMC4400730.
SVCO (orthopnea, upper body edema)	<ul style="list-style-type: none"> Lam JC, Chui CH, Jacobsen AS, Tan AM, Joseph VT. When is a mediastinal mass critical in a child? An analysis of 29 patients. <i>Pediatr Surg Int</i>. 2004 Mar;20(3):180-4. doi: 10.1007/s00383-004-1142-6. Epub 2004 Apr 3. PMID: 15064964. Reddy CSK, Phang DLK, Ng ASB, Tan AM. A simplified approach for anaesthetic management of diagnostic procedures in children with anterior mediastinal mass. <i>Singapore Med J</i>. 2020 Jun;61(6):308-311. doi: 10.11622/smedj.2019139. Epub 2019 Nov 4. PMID: 31680177; PMCID: PMC7905131. Angheliescu DL, Burgoyne LL, Liu T, Li CS, Pui CH, Hudson MM, Furman WL, Sandlund JT. Clinical and diagnostic imaging findings predict anesthetic complications in children presenting with malignant mediastinal masses. <i>Paediatr Anaesth</i>. 2007 Nov;17(11):1090-8. doi: 10.1111/j.1460-9592.2007.02279.x. PMID: 17897276; PMCID: PMC4400730. Ruellyn Cockcroft. National Child Cancer Network Child Cancer Clinical Guideline: Superior Mediastinal Compression (SMC) Syndrome. Ed.: Scott Macfarlane. 8 Jun 2016. Accessed 8 May 2022 (https://starship.org.nz/guidelines/superior-mediastinal-compression-smc-syndrome/) Oncological Emergencies. In: <i>Paediatric Haematology & Oncology Supportive Care Protocols</i>, 4th Edition version 2.0, 19 Oct 2018. Great Ormond Street Hospital for Children NHS Foundation Trust, The Royal Marsden NHS Foundation Trust, University College London Hospitals NHS Foundation Trust.
Great vessel compression (esp PA)	<ul style="list-style-type: none"> Lam JC, Chui CH, Jacobsen AS, Tan AM, Joseph VT. When is a mediastinal mass critical in a child? An analysis of 29 patients. <i>Pediatr Surg Int</i>. 2004 Mar;20(3):180-4. doi: 10.1007/s00383-004-1142-6. Epub 2004 Apr 3. PMID: 15064964.

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Large pericardial effusion	<ul style="list-style-type: none"> Lam JC, Chui CH, Jacobsen AS, Tan AM, Joseph VT. When is a mediastinal mass critical in a child? An analysis of 29 patients. Pediatr Surg Int. 2004 Mar;20(3):180-4. doi: 10.1007/s00383-004-1142-6. Epub 2004 Apr 3. PMID: 15064964. Reddy CSK, Phang DLK, Ng ASB, Tan AM. A simplified approach for anaesthetic management of diagnostic procedures in children with anterior mediastinal mass. Singapore Med J. 2020 Jun;61(6):308-311. doi: 10.11622/smedj.2019139. Epub 2019 Nov 4. PMID: 31680177; PMCID: PMC7905131. Lee SH, Oh BL, Kimpo M, Quah TC. Epidemiology of childhood malignant mediastinal masses and clinical factors associated with intensive care unit admission: A Singapore experience. J Paediatr Child Health. 2020 Jul;56(7):1039-1045. doi: 10.1111/jpc.14808. Epub 2020 Mar 12. PMID: 32162751.
Pleural effusion	<ul style="list-style-type: none"> Lam JC, Chui CH, Jacobsen AS, Tan AM, Joseph VT. When is a mediastinal mass critical in a child? An analysis of 29 patients. Pediatr Surg Int. 2004 Mar;20(3):180-4. doi: 10.1007/s00383-004-1142-6. Epub 2004 Apr 3. PMID: 15064964. Lee SH, Oh BL, Kimpo M, Quah TC. Epidemiology of childhood malignant mediastinal masses and clinical factors associated with intensive care unit admission: A Singapore experience. J Paediatr Child Health. 2020 Jul;56(7):1039-1045. doi: 10.1111/jpc.14808. Epub 2020 Mar 12. PMID: 32162751.

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