Provincial Clinical Knowledge Topic

Anterior Mediastinal Mass Management, Pediatric – Inpatient
## Document History

<table>
<thead>
<tr>
<th>Version</th>
<th>Date</th>
<th>Description of Revision</th>
<th>Revised By</th>
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<tr>
<td>1.0</td>
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<td>Sarah McKillop</td>
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Important Information Before you Begin

The recommendations contained in this knowledge topic have been provincially adjudicated and are based on best practice and available evidence. Clinicians applying these recommendations should, in consultation with the patient, use independent medical judgment in the context of individual clinical circumstances to direct care. This knowledge topic will be reviewed periodically and updated as best practice evidence and practice change.

The information in this topic strives to adhere to Institute for Safe Medication Practices (ISMP) safety standards and align with Quality and Safety initiatives and accreditation requirements such as the Required Organizational Practices. Some examples of these initiatives or groups are: Health Quality Council Alberta (HQCA), Choosing Wisely campaign, Safer Healthcare Now campaign etc.
Rationale

The presence of an anterior mediastinal mass (AMM) in a pediatric patient is considered an oncologic emergency. In children, the most common tumor type associated with an AMM is lymphoma. T cell leukemia, germ cell tumors and benign lesions also present with a mediastinal mass.

Ultimately, a timely diagnostic biopsy to establish a histologic diagnosis is essential to guide the appropriate therapy of a child with a mediastinal mass. Due to the relative rarity of a child presenting with a mediastinal mass, the decision making regarding the management of a child with a mediastinal mass from presentation until the procurement of a diagnostic sample should be done in conjunction with pediatric subspecialists including pediatric oncology. The procurement of a biopsy should be done only in a tertiary care pediatric hospital such as the Stollery Children’s Hospital or the Alberta Children’s Hospital.

The role of the primary care or emergency physician when a child presents with an AMM on chest x-ray is 1) to complete an immediate assessment of the hemodynamic and respiratory status of the child, 2) to provide emergent care to stabilize the child as needed, 3) seek urgent consultation from Pediatric Oncologist on call at the tertiary care pediatric hospital for discussion of decision making regarding the management of the child.

The role of the tertiary care pediatric hospital subspecialists (including but not limited to: pediatric oncology, pediatric intensive care, pediatric surgery, pediatric anaesthesia, pediatric radiology) is to coordinate multidisciplinary care of the child with AMM with goal of completing diagnostic investigations including biopsy in a safe and controlled manner and the timely initiation of appropriate disease directed treatment. Determining the most appropriate and effective location, approach and timing to obtain a diagnostic specimen for tissue diagnosis is important considering the need for anesthesia for most biopsy procurement options for children. The risk of life-threatening or fatal airway obstruction or cardiovascular collapse either acutely or during anesthesia in patients with AMM has been recognized for decades. The incidence of adverse events associated with anesthesia for pediatric patients with an AMM has decreased over the last two decades. This is likely due to an increased awareness of the nature and involvement of AMM by pediatricians, oncologists, intensivists, surgeons and anesthesiologists and the careful balancing of the safest and most effective approaches to obtaining a tissue biopsy.

Up to half of children will be asymptomatic at presentation with the AMM. This in itself does not preclude the child from a risk of airway obstruction or hemodynamic collapse with the administration of general anesthesia. Careful assessment of known predictive factors for anesthetic complications, and involving appropriate consultation services to assist in strategic planning for tissue biopsy is essential for the safe and timely management of a pediatric patient with an AMM.

This document is intended to summarize the initial management and workup of pediatric patients with suspected or confirmed AMM. A review of the literature, provides primarily level 4 and level 5 evidence (case series, case reports and expert opinion). Given the rarity of the problem, the diversity of presentations of pediatric AMMs, the variety of diagnostic modalities,
and the current infrequency of complications, higher level evidence to guide the initial management is unlikely. The differential diagnosis of an AMM is varied including malignant tumors, benign tumors and congenital anomalies. For further evidence in the management of these differential diagnoses, we refer the reader to topic specific protocols or literature.
Goals of Management

General Goals of Management of Pediatric AMM:

1. **ABC**: Protect airway and support oxygenation as appropriate. Establish “position of comfort” for individual patient to serve as “position of airway rescue” for symptom improvement. Maintain spontaneous ventilation. Consider establishing intravenous access and obtaining blood work if appropriate (See order set). **Do not use sedation without consultation with Anesthesia.**

2. **Ensure regular reassessment and adequate monitoring of respiratory and hemodynamic status of patient.** Acute changes in clinical status may occur with change in position, change in level of consciousness or progression of the mass.

3. **Chest X-ray** (Frontal and lateral) early during initial assessment if not done already to determine presence of AMM. Prompt interpretation of chest x-ray by radiology and review by the ordering clinician is required within a maximum of 24 hours if patient is clinically stable. Imaging should be forwarded to the receiving centre. Please note: cross section imaging will generally be completed at the pediatric tertiary care centre once patient is transferred/seen there.

4. **Perform patient directed history and physical examination** identifying symptoms and signs predictive of either impending or perioperative airway or respiratory compromise and/or hemodynamic instability due to mass effect on the airway, lungs, major cardiovascular structures and/or circulation. (Table 1)

5. **Urgent consultation with Pediatric Oncologist** at the tertiary care pediatric hospital to assist with immediate patient management and plan for transfer of care to the tertiary centre. Depending on the severity of the child’s symptoms and clinical status, contact through Rapid North or South with the pediatric oncologist on call may be essential.

Additional Goals for Pediatric Subspecialists:

1. **Timely multidisciplinary consultation to facilitate ongoing care and planning of diagnostic procedures** – including but not limited to Pediatric Oncology, Pediatric Anesthesia, Pediatric Intensivist, Pediatric Surgery, Pediatric Radiology, Interventional Radiology, Pathology.

2. **Safely obtain diagnostic sample diagnosis through least invasive approach as soon as is feasible.** (Initiation of empiric steroid therapy at the discretion of the pediatric oncologist or intensivist may be necessary to ameliorate symptoms of respiratory or cardiovascular distress and to facilitate diagnostic sample procurement. Once steroids are started the specimens for diagnosis must be collected within a maximum of 48 hours).

3. **When required, anesthesia should be undertaken with caution and with a reversible strategy that preserves spontaneous ventilation.**

4. **Timely initiation of disease directed therapy.**
### Table 1.0 Positive Predictive Factors for Anesthetic Risk with AMM

<table>
<thead>
<tr>
<th>Positive Predictive Symptoms/signs</th>
<th>Positive Predictive Echo Finding</th>
<th>Positive Predictive CT findings</th>
<th>Positive Predictive Supine Peak End Flow Rate (PEFR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Orthopnea</td>
<td>Pericardial Effusion</td>
<td>Tracheal cross sectional area &lt;50% predicted for age</td>
<td>&lt;50% predicted</td>
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<tr>
<td>Upper body edema</td>
<td>PA outflow obstruction</td>
<td>Severe carinal or bronchial compression</td>
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<tr>
<td>Syncope</td>
<td></td>
<td>SVC or great vessel obstruction</td>
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<td>Stridor</td>
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<tr>
<td>Pleural/pericardial effusion</td>
<td></td>
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<tr>
<td>Significant dyspnea</td>
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Algorithm 1.0: Initial Presentation and Management of Pediatric Patient with Suspected Anterior Mediastinal Mass (AMM)

Patient presents with signs or symptoms suggestive of AMM and is **clinically stable**

Patient presents with incidental finding of AMM on chest imaging and is **clinically stable**

Patient presents with signs of impending respiratory or cardiovascular collapse and signs or symptoms suggestive of AMM

1. Complete history and physical exam with focus on identifying positive predictive respiratory and/or cardiovascular symptoms (see table 1)
2. Obtain Chest X-Ray (Frontal and Lateral) if not already done. Follow-up within 24 hours or less if report not immediately available.
3. Obtain laboratory investigation (see order set)
4. Consult pediatric oncology at tertiary center as soon as AMM confirmed to determine management plan. Ensure all investigations are forwarded to the tertiary center with the patient.

1. **Urgently contact Pediatric Oncologist On Call at the Tertiary Care Pediatric Hospital through Rapid North or South for emergent consult** (depending on severity of symptoms/signs, the Rapid North or South call may also include the PICU) and **arrangement for transport to the tertiary centre**.
2. Support spontaneous ventilation (as possible) to assist in maintaining low intrathoracic pressure and minimizing the risk of major airway or vascular collapse - consider use of Heliox, CPAP, LMA with spontaneous ventilation if needed to temporize patient.
3. Establish position of comfort for patient
4. Avoid the use of neuromuscular blockade
5. If intubation is required anesthesia should coordinate this as bronchoscopy or awake intubation may be necessary. For intubation, a cuffed ETT tube should be placed distal to tracheal obstruction.*
7. Obtain laboratory investigation (see order set)
8. Empiric treatment with corticosteroids +/- chemotherapy should be considered only in conjunction with Pediatric Oncology when clinically indicated for respiratory or hemodynamic compromise.
9. Ensure all investigations are forwarded to the tertiary center with the patient.

*please note the level of obstruction may occur at the level of the carina inhibiting the ability to intubate distal to the obstruction

Ensure regular reassessment and adequate monitoring of respiratory and hemodynamic status of patient. Acute changes in clinical status may occur with change in position, change in level of consciousness or progression of the lesion.
Algorithm 2.0 Patient known to have Anterior Mediastinal Mass (AMM) and is in Tertiary Care Pediatric Hospital

Obtain necessary investigations and consultations to appropriately plan for diagnostic biopsy.
- Ensure regular reassessment and adequate monitoring of respiratory and hemodynamic status of patient. Acute changes in clinical status may occur with change in position, change in level of consciousness or progression of the lesion.
- Review Chest X-Ray and obtain cross sectional imaging of the chest in consultation with Pediatric Radiology. Avoid sedation for procedures. Ensure communication of the patient’s position of comfort to diagnostic imaging staff. Physician supervision for imaging should be considered.
- Obtain echocardiogram. Avoid sedation and ensure communication of the patient’s position of comfort* to the cardiology team.
- Obtain Supine Peak Expiratory Flow rate.
- Consult Anesthesia, Pediatric Surgery, Pediatric Oncology, Pediatric Radiology, Interventional Radiology and Pediatric Intensive Care to assist in planning for ongoing care and consideration of planning diagnostic biopsy procedure. Consider consultation with Pediatric Pulmonology and Pediatric Cardiology depending on clinical and imaging presentation.
- Prior to any planned diagnostic samples collection, pre-procedure communication with the pediatric pathologist is essential.
- The Clinician performing the biopsy procedure is responsible for booking and coordinating the procedures with the Operating Room/other teams (as needed).
- If concerned about impending airway or hemodynamic collapse, consider consultation and transfer to a center with ECMO/bypass capability.

Consider options to obtain diagnostic tissue sample by the least invasive option that will be result in diagnosis.
- Is it possible to consider diagnostic procedures using local anesthetic +/- anesthesia guided sedation in the operating room based on the patients age and developmental status?
- Is there an absence of positive predictive factors for anesthesia risk (see table 1)?

Proceed with least invasive diagnostic procedure to obtain biopsy using standard Anesthesia precautions for patients with known AMM.

Consider least invasive option that will provide diagnostic sample (consider available options)

Are there blasts on the CBC and differential (CB CD)?
Send peripheral smear and peripheral blood flow cytometry* to rule out leukemia.

Is the CBCD abnormal and suggestive of bone marrow infiltrative disease?
Consider bone marrow aspiration and biopsy With flow cytometry* using local anesthesia +/- anesthesia guided sedation in the OR. Consider if one sided bone marrow is adequate.

Is there a pleural effusion?
Consider thoracentesis under local anesthesia +/- anesthesia guided sedation in an OR and send samples for cytology, and flow cytometry*.

Are there palpable suspicious lymph nodes?
Consider open surgical biopsy under local anesthesia +/- anesthesia guided sedation in the OR.

Can an image guided biopsy of the mediastinal mass be completed under local anesthesia +/- anesthesia guided sedation in the OR by the radiologist?

Based on patient presentation or status no least invasive option for procuring a diagnostic sample is possible:
- In consultation with pediatric oncology consider steroid pretreatment for 24-48 hours maximum prior to procuring diagnostic sample
- Continue to reassess options to obtain diagnostic sample (must be obtained within 48 of starting steroids to optimize ability to obtain diagnosis)

*Flow cytometry should be requested from one diagnostic specimen unless agreed by pathologist
Name of Order Set: Anterior Mediastinal Mass Management Pediatrics Orders

Order Set Components

Order Set Restrictions: Pediatric patients
Order Set Keywords: Chest mass, Mediastinal

***Non-defaulted tests/consultations should be completed only once patient is received at the tertiary care pediatric hospital unless directed by the Pediatric Oncologist at tertiary center.

Laboratory Investigations

Hematology
✓ Complete Blood Count
☐ PT INR
☐ Fibrinogen
☐ Peripheral Blood Smear

Chemistry
✓ Electrolytes (Na, K, Cl, CO2)
✓ Urate
✓ Creatinine LEVEL
✓ LD (defer till tertiary care if difficulty with specimen collection)
✓ AFP (defer till tertiary care if difficulty with specimen collection)
✓ HCG Beta (Serum) (defer till tertiary care if difficulty with specimen collection)
✓ Phosphorous
✓ Calcium LEVEL
☐ Urea

Transfusion Medicine
☐ Type and screen

Diagnostic Investigations
✓ Chest X-Ray, 2 Projections (PA and Lateral)
☐ CT Chest
☐ Echocardiogram

Other Investigations
☐ Electrocardiogram- 12 lead
☐ Peak Expiratory Flow Rate

Consultations
✓ Pediatric Oncology
☐ Pediatric Anesthesia
☐ Pediatric General surgery
☐ Pediatric Radiology
☐ Interventional Radiology
Activity
- Keep head of bed greater or equal to 30 degrees. (Patient’s position of comfort, to serve as a rescue position for symptom improvement, may vary).

Respiratory Care
- Oxygen to keep oxygen saturations greater than or equal to 92%.
- Notify physician/nurse practitioner if O2 requirement is increased to maintain the same level of oxygenation, if there is a progressive increase in the work of breathing, or the patient is becoming more drowsy or lethargic.

Monitoring
Vitals signs: These orders need to be re-evaluated when the patient stabilizes or by two hours, whichever occurs first. Vital signs to include: respiratory rate (RR), pulse rate (P), blood pressure (BP), temperature (T), and oxygen saturation (O2 sat):
- routine
- every_____hourly
- every_____minutes
- continuous oxygen saturation monitoring

Diet
- NPO (due to respiratory distress)
- Clinical communication:
  - Patient can eat as usual until 8 hours before anesthesia
  - May have infant formula/light meal 6 hours before anesthesia
  - Stop solids at 6 hours before anesthesia
  - Breastmilk until 4 hours before anesthesia
  - Clear fluids until 2 hours prior to anesthesia
  - NPO 2 hours prior to anesthesia
- Other Diet:____________

Intravenous Orders (consider based on clinical status)
- Intravenous Cannula – Initiate IV (If evidence of superior vena cava syndrome carefully consider location of IV access to ensure unobstructed flow)

IV Maintenance (avoid potassium containing fluids):
- dextrose 5% in water - 0.9% sodium chloride IV at _____mL/hour
- 0.9% sodium chloride IV at _____mL/hour

IV bolus or rapid infusion including the following:
- 0.9% sodium chloride IV infusion _____ mL (10 to 20 mL/Kg) bolus
## Analytics

### Baseline Analytic - Outcome Measure

<table>
<thead>
<tr>
<th>Name of Measure</th>
<th>Order Set Usage for topic: Anterior Mediastinal Mass, Pediatric- Inpatient</th>
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<tbody>
<tr>
<td>Definition</td>
<td>For all patients admitted with suspected Anterior Mediastinal Mass, the number of times order set &quot;Anterior Mediastinal Mass Management Pediatrics Orders&quot; is being used. Overall, by region, by sites, and by units</td>
</tr>
<tr>
<td>Rationale</td>
<td>Intended to measure if the order set cited in the knowledge topic is being used. May indicate areas with adoption issues or gaps in topic</td>
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<tr>
<td>Notes for Interpretation</td>
<td>Site capacity, rural considerations, roll out of provincial CIS</td>
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### Clinical Analytics – Outcome Measure

<table>
<thead>
<tr>
<th>Name of Measure</th>
<th>Time to Pediatric Oncology Consult for Pediatric Patients with Anterior Mediastinal Mass on Imaging</th>
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<tbody>
<tr>
<td>Definition</td>
<td>In patients who have initial imaging confirming the presence of Anterior Mediastinal Mass (AMM), how long after imaging is pediatric oncology consulted?</td>
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<tr>
<td>Rationale</td>
<td>Timely referral (within 24 hours) enables the pediatric oncologist to coordinate multidisciplinary care of the child with AMM with goal of completing diagnostic investigations including biopsy in a safe and controlled manner and the initiation of appropriate disease directed treatment.</td>
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<tr>
<td>Notes for Interpretation</td>
<td>Variations in complexity of patients, rural considerations</td>
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Key Words

- Anterior mediastinal mass
- Chest mass
- AMM
- Anesthesia
- Lymphoma
References

Additional Reading and General References
Acknowledgements

We would like to acknowledge the contributions of the clinicians who participated in the development of this topic. Your expertise and time spent are appreciated.

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