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EDUCATIONAL REVIEW

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Anesthesia for children with anterior mediastinal masses

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Abstract

Children with an anterior mediastinal mass may have cardiopulmonary compromise that can be exacerbated under general anesthesia. Signs and symptoms such as cough, shortness of breath, stridor, orthopnea, accessory muscle use, a history of respiratory arrest, and the presence of a pleural effusion and upper body edema are predictive of perioperative complications. A larger mediastinal mass on imaging is predictive of perioperative complications. Risk stratification of patients, together with an individualized plan, will best guide operative management for patients with an anterior mediastinal mass. General anesthesia (GA) should be avoided if possible, but a spontaneous breathing technique is recommended if GA is required.

KEYWORDS
Anterior mediastinal mass, pediatrics

1 | INTRODUCTION

There is a lack of evidence in the literature to provide guidance on risk stratification and operative planning for the safe conduct of anesthesia and surgery in children with an anterior mediastinal mass. This focused review looks at the existing evidence and provides an evidence-based approach to their management.

2 | ANATOMY

The mediastinum can be divided into the anterior, middle, and posterior mediastinal compartments based on arbitrary landmarks on the lateral chest X-ray (CXR).[1] However, no anatomical or fascial planes exist to separate these compartments.[1] The heart and great vessels, tracheobronchial tree, esophagus, nerves, thymus, and lymphatics are all located in this area.[2]

A newer classification system based on anatomical boundaries defined by computerized tomography (CT) has been developed by the International Thymic Malignancy Interest Group, in conjunction with the Japanese Association.[2] This system emphasizes the anatomic interactions of mediastinal tumors with the surrounding mediastinal structures.[3]

3 | PRESENTATION

The most common cause of anterior mediastinal masses reported in children is lymphomas.[3] Other malignant causes include acute lymphoblastic leukemia and germ cell tumors.[4] Neuroblastomas and neurogenic tumors originate from the posterior mediastinal space and are therefore not generally associated with features of anterior mediastinal compression, unless they grow to such a size that they occupy part of the anterior mediastinal compartment, which can happen very occasionally. Non-malignant causes include bronchial cysts, teratomas, and vascular malformations.[4] The actual number and distribution of tissue diagnoses in children with anterior mediastinal masses are likely to be far more than what has been reported, as most of the published literature is retrospective case reports or limited case series.[5–10]

The incidence of perioperative complications ranges from 9% to 20% based on single-institution case series.[5–7,11] The reported complications range from transient and mild cardiorespiratory compromise to severe cardiopulmonary collapse. The authors' definitions of what constitutes a complication vary, with some having more stringent criteria than others, thus making comparisons between published studies difficult.[5–8,10–13] (Table 1).

A retrospective review over an 8 year period of children presenting with anterior mediastinal masses reported that 76% (34/45)

of children were symptomatic on presentation.[7] Signs and symptoms of airway compression include cough, chest fullness, dyspnea, hoarseness, stridor, and orthopnea. Cardiac compression can result in syncope, tachycardia, jugular venous distension, superior vena cava syndrome, and cyanosis. Patients can also present with constitutional symptoms such as fever, night sweats, and weight loss.[1]

4 | PHYSIOLOGY

Children with anterior mediastinal masses are at an increased risk of cardiopulmonary complications under general anesthesia (GA) in a supine position due to the physiological changes associated with it.

Respiratory changes under general anesthesia include ventilation/perfusion mismatch, a decreased functional residual capacity, decreased lung compliance, and the loss of bronchial muscle tone.[1,10] The supine position, subsequent increasing depth of anesthesia, and onset of IPPV in these children all contribute to the detrimental changes caused. In the supine position, the gravitational effects on the chest wall and anterior mediastinal mass reduce lung compliance. During spontaneous ventilation, the negative intrathoracic pressure during inspiration reduces the compressive effect of the anterior mediastinal mass.[1] Institution of positive pressure ventilation increases intrathoracic pressure and compresses the tracheobronchial tree during inspiration.[1] Neuromuscular blockade causes smooth muscle relaxation, a loss of airway muscle tone, and further worsens airway compression.[1]

Cardiovascular effects such as obstruction of the vena cava and venous return to the heart and direct cardiac compression may also play a role.[1,10] The dynamic effects on the heart and aorta are significantly less due to the higher operating pressures and muscular nature of these structures.[3] The pulmonary artery tends to be protected due to the anatomic location between the aorta and tracheobronchial tree[3]; nevertheless, there can be significant compression of the right side and pulmonary outflow tract with resulting decreased pulmonary perfusion, hypoxemia, and right ventricular failure.[1] These right-sided changes can result in a decrease in left-ventricular preload and subsequent reduction in cardiac output.[1]

Key points

- Children with an anterior mediastinal mass may have cardiopulmonary compromise that can be exacerbated under general anesthesia.
- Signs and symptoms such as cough, shortness of breath, stridor, orthopnea, accessory muscle use, a history of respiratory arrest, and the presence of a pleural effusion and upper body edema are predictive of perioperative complications.
- A larger mediastinal mass on imaging is predictive of perioperative complications.
- Risk stratification of patients, together with an individualized plan, will best guide operative management for patients with an anterior mediastinal mass.
- General anesthesia (GA) should be avoided if possible, but a spontaneous breathing technique is recommended if GA is required.

5 | SIGNS AND SYMPTOMS

Case reviews have tried to determine signs and symptoms which can predict perioperative complications (Table 2). Some of these include cough, shortness of breath, orthopnea, pleural effusion, accessory

TABLE 1 Complications

Study	Study size	Year	Complications
Ng et al. (2007)[11]	63	1964– 2002	Difficulty with ventilation, difficulty with intubation, hypotension, cardiac arrest, respiratory arrest, death
Anghelescu et al. (2007)[5]	118	1985– 2000	Transient respiratory problems[a], unplanned intubation, opioid overdose
Hack et al. (2008)[6]	56	1999–2006	Transient airway obstruction, lung collapse, stridor post-extubation, hypotension
Stricker et al. (2010)[7]	46	1998–2006	Airway obstruction, pneumothorax
Acker et al. (2014)[8]	69	2002–2012	Lung collapse, fulminant SVC syndrome
Garey et al. (2011)[10]	26	1994–2009	Transient desaturation[b], prolonged ventilation post-operatively
Reddy et al. (2000)[12]	25	2001–2013	Laryngospasm, difficulty with ventilation, hypotension
Azizkhan et al. (1985)[13]	50	1978–1984	Difficulty with ventilation

Abbreviation: SVC, superior vena cava.

[a]Transient respiratory problems that responded to airway suctioning, manual ventilation via mask, change in position to sitting or lateral decubitus, or change in ventilatory strategy.

[b]Transient desaturation likely secondary to airway obstruction.

TABLE 2 Signs and symptoms associated with risk factors for complications

Study	Study size	Year	Complication rate, number (%)	Risk factors for complications
Ng et al. (2007)[11]	63	1964–2002	7 (15%)	Evidence of tracheal or vascular compression, infection, and ≥3 respiratory S/S: cough, shortness of breath, orthopnea, pleural effusion, accessory muscle use, stridor, history of respiratory arrest
Anghelescu et al. (2007)[5]	118	1985–2000	11 (9.4%)	Orthopnea, upper body edema, great vessel compression, and main-stem bronchus compression
Hack et al. (2008)[6]	56	1999–2006	11 (19.6%)	Stridor, tracheal cross-sectional area ≤30% or ≤70% with bronchial compression on CT scan
Garey et al. (2011)[10]	26	1994–2009	3 (11.5%)	Correlation with symptoms was poor

Abbreviations: CT, computerized tomography; S/S, signs and symptoms.

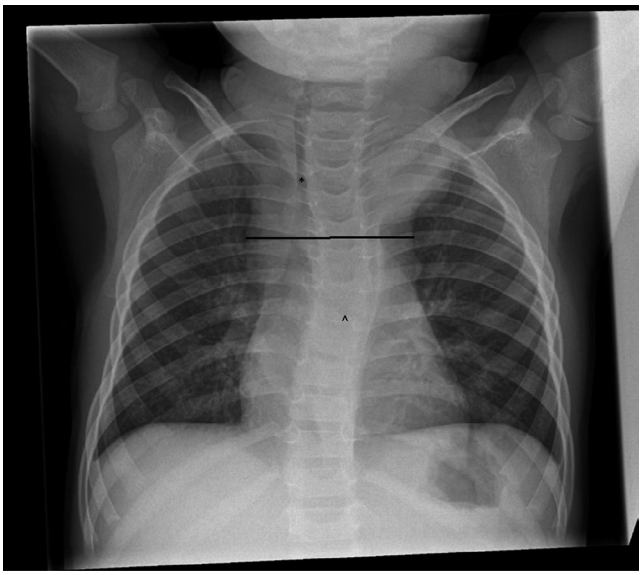

FIGURE 1 Chest X-ray of a child with an anterior mediastinal mass showing tracheal compression and deviation to the right- Anterior mediastinal mass. *Tracheal compression and deviation to the right. ^Left bronchus compression

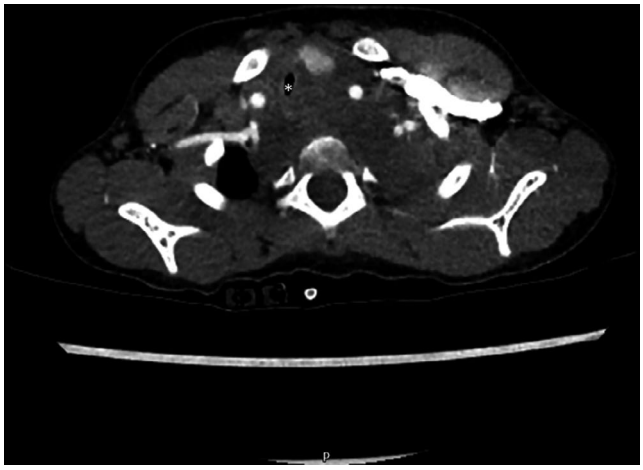

FIGURE 2 Computerized tomography scan of the same child with an anterior mediastinal mass showing tracheal compression and deviation to the right. *Tracheal compression and deviation to the right

muscle use, stridor, a history of respiratory arrest, and upper body edema.[5,6,11] Having ≥3 signs and symptoms also increases the risk of perioperative complications.[11]

It is also important to determine the "rescue" position which alleviates symptoms and in which the patient feels the most comfortable.[14] This is the position that relieves the mass effect on critical mediastinal structures.[15,16]

6 | IMAGING MODALITIES

The diagnosis of a mediastinal mass is usually made on a CXR (Figure 1). The mediastinal mass ratio (MMR), the ratio of the mediastinal mass width divided by the thoracic width, has been used to risk stratify patients. The average MMR of patients with perioperative complications was 0.56 in a pediatric population, suggesting that patients with a large mediastinal mass may be at a higher risk of perioperative complications.[17]

A CT scan makes it possible to measure the cross-sectional area (CSA) of the trachea at its narrowest (Figure 2). Patients with a smaller CSA are at a higher risk of perioperative complications (Table 3). However, this does not appear to be consistently associated with any signs or symptoms. Furthermore, Stricker et al.[7] found that although most patients with radiological evidence of cardiorespiratory compromise had signs and symptoms suggestive of mass effect, a small proportion of patients were asymptomatic (13.3%, 4/30). This is likely to be because the presence and severity of any sign or symptom are also dependent on other factors such as the rate of tumor growth, proximity to important structures in the mediastinum, and coexistence of other complications such as pericardial and pleural effusions.[6] Blank et al.[18] advocated the use of a CT scan to define the position and size of the anterior mediastinal mass once it is identified on a CXR.

An echocardiogram is generally indicated if the CT scan shows a pericardial effusion, or compression of the heart or a major vascular structure.[18] However, the results of an echocardiogram can be difficult to interpret. Many echocardiograms are performed on awake children, so the effects of anesthetic agents on the cardiovascular

TABLE 3 Imaging features associated with risk factors for complications

Study	Study size	Year	Complication rate, number (%)	Risk factors
Azizkhan et al. (1985)[13]	50	1978–1984	5 (11%)[a]	CSA <50%: significant respiratory symptoms CSA <65%: increased risk of perioperative complications
Shamberger et al. (1991)[26]	42	1980–1988	No anesthetic complications[b]	CSA <40%: orthopnea
Hack et al. (2008)[6]	56	1999–2006	11 (19.6%)	CSA <40%, or CSA <70% with bronchial compression: orthopnea, stridor, wheeze and superior vena cava obstruction CSA ≤30% or ≤70% with bronchial compression: increased risk of perioperative complications
Garey et al. (2011)[10]	26	1994–2009	3 (11.5%)[c]	Correlation with symptoms was poor

Abbreviation: CSA, cross-sectional area.

[a]General anesthesia with endotracheal intubation was administered in 45 patients.

[b]Local anesthesia was used primarily in patients with significant tracheal narrowing.

[c]General anesthesia was administered in 13 patients.

system may not be appreciated. The lack of "real-time" echo information during induction of anesthesia makes it difficult to determine whether an intraoperative complication was caused by reduced pulmonary blood flow (due to inadequate cardiac filling and/or right ventricular outflow) or airway compression causing inadequate ventilation.[1]

7 | RISK STRATIFICATION

Based on a risk stratification system derived for adults[18] and existing data on pediatric patients,[6] Pearson et al.[1] came up with a risk stratification system for children with an anterior mediastinal mass:

Low risk: Asymptomatic, no radiographic airway compression, and no cardiac or vascular compression;

Intermediate risk: Mild-to-moderate symptoms (dyspnea and orthopnea), mild tracheal compression <70%[a], and no bronchial compression;

High risk: Orthopnea, stridor or cyanosis, tracheal compression >70%[b] or tracheal CSA ≤70% with bronchial compression, great vessel compression, tamponade physiology on echocardiogram.

[a]Tracheal CSA ≥30% of normal.

[b]Tracheal CSA ≤30% of normal.

The authors concluded that patients in the low-risk group may undergo GA with positive pressure ventilation if necessary. Patients in the intermediate or high-risk groups are best managed with an individualized plan by a multidisciplinary team.[1] However, it is important to remember that a patient with mild or no symptoms could suffer cardiovascular and respiratory compromise under anesthesia, and the presence of a pericardial effusion or pulmonary outflow tract compression on imaging may pose more risk than expected. It is also important to discuss the management of complications, should any arise intraoperatively.[4]

8 | ANESTHETIC CONSIDERATIONS

It is often thought that obtaining an exact diagnosis is essential before initiating the appropriate treatment.[12,19] However, it may be prudent to first administer treatment if the patient is deemed high risk on presentation.[20] Hack et al. found that a clear diagnosis was found in 95% (17/18) of patients who required steroid pretreatment, and 100% (18/18) if the treatment course was limited to ≤5 days.[6] Discussion with the oncology team is extremely important with regard to administration of pre-diagnosis steroids (usually dexamethasone) as it is imperative that appropriate treatment to prevent tumor lysis syndrome is commenced and monitored beforehand. Case reports of tumor lysis syndrome have been associated with a dose of steroid[21,22] and anesthesia[23–25] in children.

Patients with an anterior mediastinal mass should have their procedures done in the main operating room, where support staff and rescue equipment are readily available.[1]

9 | ANESTHETIC MANAGEMENT

The degree of sedation or anesthesia required is dependent on the surgical procedure and the individual child. General anesthesia should be avoided if possible, and a superficial tissue sample can occasionally be obtained in cooperative children under sedation.[5,6,8,10,11,13,17,26] Agents such as dexmedetomidine and ketamine are less likely to cause respiratory depression[18] and have been used with success in the past.[27]

In some instances, a GA cannot be avoided. This includes procedures such as mediastinoscopy, video-assisted thoracoscopy, thoracotomy, sternotomy, or biopsy of deep tissue sites remote to the mediastinal mass.[14] All children must be thoroughly assessed clinically and the decision to proceed to general anesthesia should be made by a multidisciplinary team including senior anesthetists, surgeons, radiologists, and oncologists.

A spontaneous breathing technique is most commonly advocated in order to limit potential compression of important mediastinal structures.[1,3–5,12,14,18] The negative intrathoracic pressure during inspiration may potentially reduce the compressive effect of the anterior mediastinal mass.[1,12] If muscle paralysis is required, deepening the anesthetic with short-acting drugs and verifying that the patient can tolerate positive pressure ventilation before using a short-acting muscle relaxant is recommended.[1]

10 | Initial management in cardiorespiratory collapse

A plan should be devised and discussed with the team before commencing sedation or anesthesia. This will have taken into consideration the preoperative clinical assessment and radiological findings.

In the event of airway compression, application of continuous positive airway pressure and/or an attempt to ventilate with positive pressure ventilation should be considered.[4,14] A tracheal tube should ideally be advanced beyond the point of obstruction.[14] A rigid bronchoscope may be required if this fails,[1,4] which enables navigation over the narrowed region, hence splinting the airway open, and also allowing simultaneous ventilation through a side port. The rigid scope is also crucial, independent of the location of the stenosis, if other techniques are not successful. Repositioning the patient in order to alleviate the compressive effect of the mass may also be helpful.[4,14]

In the event of cardiovascular collapse, strategies include a rapid fluid bolus, reducing the depth of anesthesia and repositioning.[4,14] Despite the lack of good evidence in such cases, the early use of vasopressors to augment filling pressures would also be a sensible approach. As a last resort, sternotomy to lift the mass and relieve compression of the great vessels may be required.[1,4,14]

It is important to bear in mind that both respiratory and cardiovascular complications can occur simultaneously.[14] Although cardiovascular complications are less common, it is unknown to what degree a compromised cardiac output contributes to hypoxia[6] and vice versa. Management of one system may improve the other.[14]

Extracorporeal membrane oxygenation (ECMO), both as a preemptive and rescue technique, sounds theoretically appealing in high-risk patients.[28] However, both techniques come with their technical challenges: placing ECMO cannulae in children in a supine position without sedation, and the risk of prolonged hypoxia occurring while the ECMO circuit is being established in an emergency setting.[1,6,18]

11 | CONCLUSION

Pediatric patients presenting with an anterior mediastinal mass require a thorough history and physical examination. The presence and severity of cardiopulmonary compression and any associated signs and symptoms must be elucidated. Further imaging modalities such as a CT scan and echocardiogram are useful in determining the extent of the disease burden. A multidisciplinary team approach is paramount to understanding the patient-specific risks of general anesthesia and in determining the safest management approach. Even in patients with a low estimated risk prediction, severe complications can occur. Although multiple anesthetic techniques and drugs have been used successfully in the past, most emphasize the importance of maintaining spontaneous ventilation throughout the procedure. Adequate preoperative planning should include management of intraoperative complications. Backup plans should include rigid bronchoscopy, emergent sternotomy, and cardiopulmonary bypass.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable—no new data generated.

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