Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality of lung development with an incidence of 1 to 4 per 100,000 births.\(^1\) Other cystic lung abnormalities that manifest in children and constitute the differential diagnosis of CCAM include bronchogenic cyst, pulmonary sequestration, congenital lobar emphysema, pleuropulmonary blastoma, and cystic bronchiectasis. Chin and Tang reportedly first described CCAM in 1949.\(^2\) It is associated with cystic areas and adenomatous overgrowth of the terminal bronchioles. The lesion is purported to result from an embryologic injury to the developing lung bud during early pregnancy that affects development of terminal bronchiolar structures. It is classified into 5 subtypes, with type 1 being the most common.\(^1,2\) Type 0 CCAM (worst prognosis, least common [\(<3\%\)]) is known as acinar dysplasia consisting of microcystic disease throughout the lungs. Type 1 (most common [60\%], best prognosis) is a macrocystic type consisting of a localized lesion (single or several large cysts) that partially affects a single lobe of the lung. Type 2 (20\% of cases, poor prognosis) consists of a microcystic multicystic lesion with histologic features similar to the type 1 lesion. This variety is associated with other major congenital anomalies involving the renal or cardiac system and diaphragmatic hernia. Type 3 (\(<10\%\) of cases, poor prognosis) consists of microcystic and solid adenomatous tissue resembling a bronchiolar histologic structure (cuboidal ciliated and cuboidal nonciliated epithelium). The last variety of CCAM, type 4 (10\% of cases), is a macrocystic lesion lacking mucoid cells, presenting either in childhood or later in asymptomatic adults. Pleuropulmonary blastoma has been associated with this lesion.

Fetal cystic airway malformations including CCAM can be diagnosed with prenatal ultrasonography when the lesion is classified as a macrocystic (cyst >5 mm) or microcystic. A chest radiograph and computed tomographic (CT) scan of the chest constitute important diagnostic modalities, but a definitive diagnosis can be established only by histopathological examination of the excised tissue.\(^1,2\)

Clinically, CCAM presents in neonates or young infants with respiratory insufficiency or recurrent or persistent respiratory tract infection, and it may present as pneumothorax. The lesion can enlarge rapidly by ball-valve air entrapment by cysts, leading to a mediastinal shift and compression of the heart. Hence, surgical correction is necessitated in most patients.

Surgical management of CCAM can include endoscopic or open surgical repair, which would frequently require single-lung ventilation (SLV) for optimal surgical conditions. Options for lung isolation in infants include the use of a single-lumen endotracheal tube (SLT) inserted into the desired bronchus or balloon-tipped bronchial blockers, such as a Fogarty embolectomy catheter, which have their own limitations.\(^3\) Use of a microcuffed tube for this purpose has not been documented clinically, to the authors’ knowledge. We describe a novel simple technique of lung isolation in an infant with CCAM who was scheduled for bilobectomy.

**Case Summary**

An 11-month-old, 8-kg male infant was the product
of a normal vaginal delivery at term with a normal birth history. He was asymptomatic until 3 months of age, when he started having episodes of difficulty in breathing, persistent cough, irritability, poor physical activity, and intermittent fever. There was no history of nasal stuffiness, wheezing, orthopnea, or snoring. He had received antibiotics, cough syrup, and nebulization with salbutamol/albuterol (Asthalin) and budesonide (Budecort) on several instances in the past based on clinical suspicion of recurrent lower respiratory tract infections. A chest radiograph and CT scan of the thorax were ordered when symptoms were persistent and showed cystic lesions involving the upper and middle lobes on the right side along with a mediastinal shift toward the opposite side (Figure 1).

The patient received a diagnosis of CCAM of the right upper and middle lobe and was scheduled for bilobectomy. Preoperatively, the infant had tachycardia (160/min) and tachypnea (50/min) with normal capillary filling time. Results of arterial blood gas analyses and other blood investigations were unremarkable. Arterial oxygen saturation on oxygen administered by nasal cannula was 94% to 95%. On physical examination, mild chest retraction was present, and air entry was diminished on the right side along with bilateral rhonchi and harsh breath sounds.

Intravenously, dexamethasone, 1.5 mg, and acetaminophen (paracetamol), 120 mg (15 mg/kg), were administered. General anesthesia was induced with fentanyl, 15 μg, and propofol, 20 mg. Rocuronium, 10 mg, was administered intravenously, and after gentle positive pressure ventilation (PPV), the trachea was intubated with a 3.0-mm inner-diameter microcuffed tube (Microcuff, Kimberly-Clark). Following tracheal intubation, the head was turned to the right side, the bevel of the endotracheal (ET) tube was rotated 180°, and the ET tube was advanced into the left bronchus until the breath sound of the right lung diminished. The neonatal fiberoptic bronchoscope (FOB) was not working properly, so it could not be used. The ET tube cuff was inflated by slowly using the cuff pressure manometer until there was no audible air leak to the opposite lung. At this point, the cuff pressure was approximately 15 mm Hg. Effective lung isolation was also confirmed by the presence of a collapsed lung on the right side on thoracotomy. Cuff pressures were measured after lateral positioning and intraoperatively were maintained between 15 and 20 mm Hg. An ultrasound-guided caudal epidural catheter was threaded to the thoracic level to a premeasured distance corresponding to the T6 vertebrae using a 19-gauge Tuohy needle.

Intraoperative analgesia was maintained by an infusion of 0.25% bupivacaine (1.5 mL/h) following a negative test dose of 2% lignocaine with adrenaline 1:200,000 (0.8 mL). Invasive blood pressure monitoring was initiated following radial artery cannulation. Intraoperative arterial blood gases were maintained in the reference range. The operation lasted 4.5 hours. Anesthesia was maintained with sevoflurane in an air-oxygen mixture and fentanyl. The intraoperative anesthetic course was uneventful. The total blood loss was 85 mL, which was adequately replaced. After lobectomy, a right-sided intercostal drain was placed with an underwater seal. The cuff of the ET tube was deflated, and it was withdrawn to the trachea to check for any leaks. The collapsed lung tissue was reexpanded (see Figure 1).

The infant was electively ventilated and was extubated the next day in a stable condition. Intravenous acetaminophen, 15 mg/kg, was continued every 8 hours.
in the postoperative period. An epidural infusion of 0.125% bupivacaine (1.5 mL/h) was continued for 48 hours. Following extubation, the patient remained pain free with stable vital signs and required no additional analgesia. He was discharged home after 3 weeks. Histopathologic findings confirmed the lesion to be CCAM. Written informed consent was obtained from the patient’s parent for publication for academic purposes.

**Discussion**

Lesions in CCAM present complex anesthetic challenges because of the disease itself and the frequent need for SLV in these young infants. Use of SLV in a patient with CCAM prevents the contamination of normal lung from blood, debris, and infected material and makes the surgical approach more convenient.3-5 The various techniques for pediatric SLV include a double-lumen tube (sizes 26 and 28 for patients aged 8-12 years), a single-lumen (Univent) tube (ages 6-8 years), bronchial blockers (ages 0-18 years), and endobronchial intubation (age up to 18 years).3 The advantages and disadvantages of various techniques are described in the Table. Of these options, only endobronchial intubation and bronchial blockers are feasible in infants.

In infants, the simplest and quickest means of providing SLV is to intentionally intubate the mainstem bronchus of the nonoperated lung using an SLT.3,5 A neonatal FOB is greatly helpful in the guidance and confirmation of accurate ET tube placement. However, neonatal FOB is not easily available everywhere. Our FOB was not in working condition and, hence, we had to resort to a blind technique. In a study by Narayanasamy et al, most (80%) of CCAM cases were managed using endobronchial intubation. Endobronchial intubation has certain limitations for right-sided mainstem intubation, as the right upper lobe bronchus originates near the carina and can potentially be occluded by the endobronchial ET tube, leading to poor oxygenation and ventilation.3,7 However, in the study by Narayanasamy et al,9 of 10 patients had right-sided endobronchial intubations without any major difficulties in oxygenation or ventilation.

Other options to achieve blind left-sided bronchial intubation include using a stylet to curve the distal end of the tracheal tube to the left and using a distally curved rubber bougie that is directed blindly to the left bronchus, followed by railroading the tube over the bougie.3,8 However, these techniques can be potentially traumatic to the tracheobronchial tree.

Bronchial blockers are a useful technique for lung isolation in young infants, but their proper placement requires use of an FOB and if dislodged, they can block ventilation to both lungs.3,7,8 Guruswamy et al10 used a 3 Fr Fogarty catheter (guidewire in place) modified by bending the tip by 10°, for lung isolation in a neonate with CCAM using FOB guidance.

One of the considerations in using an SLT is the inadequate bronchial seal, especially if a smaller, uncuffed tube is used. As a result, this technique may not be able to provide a collapsed lung for the operative site or protect the normal lungs from contamination.3,7,8 Congenital cystic adenomatoid malformation may contain fluid varying from clear to purulent in nature. Nishimoto et al9 described the anesthetic management of 4 children with CCAM. In a 1-year-old boy with a cyst filled with purulent material, the ET tube became almost occluded with copious purulent material during the operative period; therefore, in another CCAM lesion suspected to be filled with purulent material they used bronchial blockers to prevent contamination of another lung. A cuffed ET tube inserted in the main bronchus of the nonoperative lung can be of advantage in such situations by providing an adequate seal.

A microcuffed ET tube has advantages over a conven-

<table>
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<tr>
<th>Technique</th>
<th>Advantages</th>
<th>Disadvantages</th>
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<tr>
<td>Endobronchial intubation using SLT</td>
<td>Simplest way of achieving SLV, inexpensive, can be used as lifesaving measure in emergency such as unilateral lung hemorrhage or pneumothorax</td>
<td>Incomplete contralateral lung collapse or protection from contamination, inability to suction the operated lung, and obstruction of right upper lobe bronchi</td>
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<td>Bronchial blockers (BB)</td>
<td>Easy insertion, best device for patients with a DA, no need to exchange a tube if mechanical ventilation is contemplated</td>
<td>Difficult suctioning and incomplete lung collapse due to narrow channel; shifting back and forth from SLV to 2-lung ventilation is difficult; dislodgment or loss of seal during procedure is possible</td>
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<td>Single-lumen tube (eg, Univent, Teleflex, LMA)</td>
<td>Easy to insert, as shaped like conventional SLT, fiberoptic intubation can be done in patients with a DA, can be left in place for postoperative ventilation</td>
<td>Besides disadvantages of BB, bronchial cuff pressures are higher, and tubes are more expensive than double-lumen tubes</td>
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<tr>
<td>Double-lumen tube</td>
<td>Large lumina facilitate suctioning and CPAP application, easy conversion from single- to double-lung ventilation, finest device for reliable lung separation</td>
<td>Difficulty in size selection, technically difficult to insert, tracheobronchial and tracheal cuff damage during insertion</td>
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Table. Techniques for Lung Isolation in Children

Abbreviations: CPAP: continuous positive airway pressure; DA: difficult airway; SLT, single-lung tube; SLV, single-lung ventilation.
tional cuffed ET tube in this regard. The cuff is short, cylindrical, and distally placed, as the Murphy eye has been eliminated. In a size 3 Microcuff ET tube, the tube tip is just 0.5 cm from the distal end of the cuff (Figure 2), thereby providing a greater margin of safety compared with a conventional cuffed ET tube. Thus, there is a lesser chance of the bronchial cuff obstructing the right upper lobe bronchus that may occur easily if an uncuffed SLT is used for this purpose. Also, a microcuffed ET tube with its ultrathin polyurethane cuff is known to achieve an adequate seal with low cuff pressures. This is very important during intubation of the bronchus, where because of a narrower lumen excessive cuff pressures can be generated with use of cuffed tubes and can lead to mucosal ischemia. Use of cuff pressure monitoring is important whenever such tubes are used in children.

Another important concern is a possibility of expansion of the CCAM with PPV, which places patients at risk of pneumothorax, airway collapse, and cardiovascular collapse. Hence, avoidance of a muscle relaxant for intubation until surgical incision has been suggested in the literature along with use of SLV. In a retrospective study by Narayanasamy et al., gentle PPV following administration of a muscle relaxant was used in 95% of the cases to secure the airway and was found to be safe. In our patient also, muscle relaxant was used for intubation after gentle PPV.

Good perioperative analgesia is of utmost importance in management of these cases. Narayanasamy and colleagues found that use of epidural analgesia facilitated extubation in the operating room in 94% of cases. In our case also, multimodal analgesia using acetaminophen, dexamethasone, and thoracic epidural anesthesia reduced perioperative opioid requirements to a minimum and improved breathing.

Certain limitations of using a microcuffed ET tube for SLV do exist, as with any other endobronchial technique. Limitations include the potential for inadequate collapse of the operative lung, inability to apply continuous positive airway pressure to the opposite lung to counter intraoperative hypoxemia, inability to suction the operative lung, and potential for limitation of ventilation to the right upper lobe bronchus during right-sided endobronchial intubation. However, as mentioned, the problems are clinically not significant and are lesser with a microcuffed ET tube technique than a traditional cuffed ET tube.

**Conclusion**

A microcuffed tube with its distally placed and short cuff has distinct advantages over conventional ET tubes for achieving SLV in infants where the choices are limited. Multimodal analgesia inclusive of continuous epidural analgesia should be used whenever feasible, especially for open techniques to improve the recovery of the child.

The following are learning points:

- Surgical management of CCAM lesions present complex anesthetic challenges and often needs SLV.
- The commonest, simplest, and quickest means of providing SLV is to intentionally intubate the mainstem bronchus of the nonoperative lung using an SLT.
- An uncuffed ET tube for SLV in children may not provide a proper seal and adequate ventilation.
- The microcuffed ET tube (Microcuff) with its shorter distal cuff than a conventional cuffed ET tube provides adequate seal at low pressures and should be preferred over a conventional cuffed ET tube for SLV in children.
- Regional anesthesia should be employed whenever feasible for these cases, to improve recovery.

**REFERENCES**


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