Airway Management in Patients with Tracheal Cartilaginous Sleeve

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Abstract

Introduction: Tracheal cartilaginous sleeve (TCS) is a rare congenital malformation with less than 40 cases described in the literature. It is seen almost exclusively in patients with craniosynostosis syndromes (CS), such as Pfeiffer, Crouzon, or Apert syndromes. It is characterized by fusion of a variable number of tracheal rings and lacks normal distensibility which can lead to difficulty in clearing secretions. The tracheal mucosa seems to be particularly sensitive in these patients and has a propensity to form granulation tissue following even minor trauma such as with suctioning.

Methods and Materials

Four patients in our tertiary academic institution were identified by bronchoscopy and started on a regimen of topical ciprofloxacin/dexamethasone through the tracheostomy. Five drops were applied twice per day for one week. The regimen was repeated every other week. Caregivers were educated about the disease process and were instructed to monitor for signs of airway obstruction and to avoid any trauma with suctioning.

The patients were monitored with frequent follow up in ENT clinic. These patients were also followed with periodic bronchoscopy performed in the operating room with photo documentation used to assess disease progress before and during treatment. Assessments were made as to the number of hospitalizations pre- and post-treatment as well as the number of infections treated at home. Families and caregivers were also questioned about their comfort level in caring for these patients as well as their quality of life.

Discussion

Making the diagnosis of TCS requires a strong clinical suspicion and visualization through direct laryngoscopy and bronchoscopy (DLB). The condition is seen almost exclusively in patients with CS. Typical presenting symptoms include biphasic stridor, increased mucous production, cough, repeated respiratory infections, cyanotic episodes, and failure to thrive (2). At our institution protocols have been put in place to ensure that this diagnosis is not missed. For instance, patients with CS undergo rigid bronchoscopy with ENT as part of their workup. Many previously reported cases were discovered on autopsy, and it has become clear that an effort should be made to avoid this, especially since about 58% of deaths in these patients are directly related to respiratory compromise (2). To improve its dismal mortality rate, discovery of the condition is paramount (1).

The next decision is whether to perform a tracheostomy. Just under 50% of cases described in the literature received a tracheostomy. Advances include bypassing upper airway obstruction and allowing for improved pulmonary toilet, while a disadvantage is the development of stoma granulation which is common and can cause obstruction (2). Lertsurbura et al. in their meta-analysis found that patients who undergo tracheostomy have a statistically significant survival advantage. All of our patients underwent tracheostomy.

Continued management with frequent follow up and bronchoscopy is recommended. Only two other cases in the literature described treatment with topical steroids. Improvement in granulation tissue formation was noted in both cases which is consistent with our findings. Given the severity of this condition and the potential for decreased hospitalizations, infections, and quality of life with the application of topical ciprodex via tracheostomy, we believe the addition of this medical regimen will benefit patients with TCS.

Conclusions

• TCS is a life-threatening condition with a reported 90% mortality rate by age two.
• Early recognition is important, and a DLB is necessary to make the diagnosis.
• Tracheostomy is recommended at least in more severe cases.
• Patient family/caregiver education is of utmost importance.
• It is important to limit trauma and aggressive suctioning to limit mucosal irritation and development of potentially obstructing granulation tissue.
• Application of topical ciprodex drops via the tracheostomy twice daily every other week is recommended to improve the number of hospitalizations, infections, and quality of life.

References


Table 1. Patient characteristics and outcomes.

<table>
<thead>
<tr>
<th>No.</th>
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<th>Age at tracheostomy</th>
<th>Pre-treatment admissions</th>
<th>Post-treatment admissions</th>
<th>Quality of life comments</th>
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</table>
| 1   | 2y M   | Crouzon, OSA, TCS (grade III) | 12m | 2 | 2 | "Post-travel in pain, no oxygen needed."
| 2   | 2y F   | Pfeiffer, OSA, Crouzon, TCS (grade III) | 22m | 2 | 2 | "Decreased secretions."
| 3   | 4y F   | Pfeiffer, OSA, Crouzon, TCS (grade III) | 28m | 6 | 1 | "Happy with [regimen]! Not as many trips to the ER."
| 4   | 4y F   | Pfeiffer, TCS (grade II) | 6m | 1 | 0 | "Not going to the ER as much."

Table 2. Patient characteristics and outcomes.

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