Congenital Isolated Agenesis of Epiglottis Presenting with Acute Life-threatening Event Successfully Managed by Temporary Supraglottic Closure and Tracheostomy

Ragahvendra G Prasad

ABSTRACT
Isolated agenesis of epiglottis is extremely rare. The exact incidence is unknown. Anecdotal presentations associated with Pierre Robin syndrome maxillary hypoplasia have been reported. Agenesis of epiglottis has also been published in an adult. Isolated agenesis of epiglottis with acute life-threatening event (ALTE) has not been specifically dealt with in the literature. Hence, this is the report of six contiguous cohort of isolated agenesis of epiglottis with ALTE.

Materials and methods: After the first case of isolated agenesis of epiglottis was diagnosed 2 years ago, five more such cases were treated by the group. These formed the index cases of the cohort. Decision for surgery was based on an arbitrary 3 ALTEs in 24 hours. All the patients were initially intubated. Supraglottic space was closed using interrupted unabsorbable sutures. Tracheostomy was added. At 3 to 6 months of follow-up, glottis was restored by Nd-YAG laser knife. Five out of six children are alive and thriving well. One child died before he could be actively managed.

Conclusion: Isolated agenesis of epiglottis with ALTE is extremely rare. Supraglottic closure saves lives and gives enough time for pharyngoesophageal coordination to develop.

Keywords: Agenesis of epiglottis, ALTE, Temporary supraglottic closure.

INTRODUCTION
Exact incidence of agenesis of epiglottis is not known. John Jacob Ballenger and James B snow Jr described five documented cases of anomalies of epiglottis. Ted L Tewjik, Vazken M Den Kaloustian also described development of epiglottis. Agenesis of epiglottis may either go unnoticed or present with respiratory distress and choking in children or even older people. Yang Jee Kim et al described a case of congenital aplasia of epiglottis in an adult.

Acute life-threatening event (ALTE) has rarely been reported with isolated absent epiglottis although absent epiglottis with Pierre Robin complex has been reported. Hoong reported epoglottoc anomales associated with maxillary hypoplasia and Pierre Robin sequence. Koempel J et al reported rudimentary epiglottis associated with Pierre Robin sequence. Habib et al isolated hypoplastic epiglottis with serious consequences. The exact management of absent epiglottis is, therefore, not standardized even in textbooks. After detecting the first case of agenesis of epiglottis 2 years ago, five other newborns with respiratory distress due to isolated agenesis of epiglottis were treated successfully with temporary supraglottic closure and tracheostomy. Hence, this attempts to highlight ALTE as a presenting feature of congenital agenesis of epiglottis and to suggest a method to save lives in such cases. Glottis can be restored later, once pharyngoesophageal coordination develops.

MATERIALS AND METHODS
This is an observational contiguous cohort of isolated agenesis of epiglottis treated by the group. Six cases were diagnosed and managed. These form the contents of this report. Table 1 shows the day of presentation and the number of ALTEs before referring to the surgeon. All the children were term new-born babies and presented with acute severe respiratory distress, cyanosis and bradycardia. There were no associated anomalies. Primary intubation was difficult, the help of an experienced pediatrician was needed. Indirect laryngoscopy finding of agenesis of epiglottis were confirmed by bronchoscopy. The inclusion for surgery was based on more than three ALTEs in 24 hours. All the babies were intubated and ventilated in a supine neck-extended position. A transverse suprahyoid incision was made. Agenesis of epiglottis conformed with clear picture of vocal cords on one side and nasogastric tube in pharynx on another (Figs 1A to E). Para-arytenoid and
Table 1: Age, sex, ALTE at time of presentation, confirmation and outcome

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>No. of ALTE</th>
<th>Difficult intubation</th>
<th>Bronchoscopy</th>
<th>Supraglottic closure</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Term baby (3 days)</td>
<td>Male</td>
<td>3 episodes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Undone and alive at 6 months</td>
</tr>
<tr>
<td>Term baby (1 day)</td>
<td>Male</td>
<td>3 episodes</td>
<td>Yes</td>
<td>Repeated</td>
<td>Yes</td>
<td>Undone at 8 months and doing well</td>
</tr>
<tr>
<td>Term baby (2 days)</td>
<td>Male</td>
<td>4 episodes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Undone at 4 months at doing well</td>
</tr>
<tr>
<td>Term baby (2 days)</td>
<td>Female</td>
<td>3 episodes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Awaiting 6 months, thriving well</td>
</tr>
<tr>
<td>Term baby (2 days)</td>
<td>Female</td>
<td>4 episodes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Undone at 5 months, doing well</td>
</tr>
<tr>
<td>Term baby (3 days)</td>
<td>Male</td>
<td>5 episodes</td>
<td>Attempted</td>
<td>Not done</td>
<td></td>
<td>Died before supraglottic closure</td>
</tr>
</tbody>
</table>

supra-arytenoid interrupted non-absorbable sutures were taken for temporary supraglottic closure. A temporary tracheostomy was added. Postoperative nasogastric tube feeding was continued and later shifted to exclusive breastfeeding. After 3 to 6 months, once clinically satisfied with pharyngoesophageal coordination, glottis was restored using Nd-YAG laser. Tracheostomy closed spontaneously. Remaining two patients are waiting for undergoing supraglottic closure.

RESULTS

All children who had 3 or more ALTEs in 24 hours were rescued and saved by this temporary glottic closure and tracheostomy. At 6 months follow-up, five out of six children were thriving well and pharyngoesophageal coordination re-established. One child succumbed before he could be operated.

DISCUSSION

Incidence of isolated agenesis of epiglottis is not clear\(^3\) associated with Pierre Robbin syndrome (absence epiglottis) hemigenesis of epiglottis, bifid epiglottis\(^1,3\) have been reported. Agenesis of epiglottis may either get unnoticed or may present with respiratory distress and choking and may even present in older people.\(^3\) Yang Jee Kim et al described a case of congenital aplasia of epiglottis
in an adult. Acute life-threatening event. ALTEs may be caused by systemic, CNS, TEF and other such conditions including massive oral tumors. Patients in this series were term babies and had respiratory distress and cyanosis at the time of presentation. Initial attempts at revival and intubation detected the absence of epiglottis which was later confirmed by bronchoscopy. Exact management of agenesis of epiglottis is unclear. The authors tried a temporary supraglottic closure by para- and supra-arytenoid spaces with temporary supraglottic sutures, protecting airway from digestive tract. A tracheostomy was added. This type of supraglottic closure saves lives and gives adequate time for newborn to develop pharyngo-oesophageal coordination.

Supraglottic closure is an effective way of preventing aspiration and temporary tracheostomy is very effective for airway management. Supraglottic closure facilitates easy feeding and helps baby to grow till pharyngo-oesophageal coordination develops. At 3 to 6 months of age, pharyngo-oesophageal coordination is checked and adequate supraglottic closure is undone using Nd-YAG laser. It is an extremely simple procedure. Five out of six children were revived with this technique and are thriving well. One child succumbed before he could be operated.

CONCLUSION
Isolated agenesis of epiglottis is extremely rare and when associated with ALTE, can be fatal. A temporary supraglottic closure with tracheostomy saves lives. Restoration of glottis is easy using Nd-YAG laser.

REFERENCES