Original article:

Study of association of congenital anomalies in patients with oesophageal atresia and distal tracheo-oesophageal fistula.

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Abstract:

Introduction: Oesophageal atresia is one of the commonest neonatal emergencies faced by paediatric surgeons. The incidence of oesophageal atresia varies from 1 in 2500 to 1 in 4500 live births with an average incidence of 2.4 per 10000 births. The most common type of oesophageal atresia is Vogt Type 3, in which there is a blind upper pouch with a fistula to the trachea or bronchus from the lower oesophagus. It occurs in about 85%--87% of patients.

Methodology: An Ongoing study is being carried out in a paediatric surgery unit in a teaching hospital in Mumbai. On an average, 20 newborns with oesophageal atresia are referred to the hospital per year. More than 80% are oesophageal atresia with distal tracheoesophageal fistula (Vogt’s type 3b; Gross type C).

Results: The present study consisted of twenty-five cases of oesophageal atresia with distal tracheoesophageal fistula. All 25 children have successfully undergone primary oesophageal anastomosis. All the cases are treated in a pediatric surgical unit of a tertiary teaching hospital. One patient of the present study had right sided Morgagni type of congenital diaphragmatic hernia. The association of these two conditions viz OA and congenital diaphragmatic hernia is rare.

Conclusion: Commonest congenital anomaly associated with oesophageal atresia with tracheoesophageal fistula was congenital heart disease.

Introduction:

Oesophageal atresia is one of the commonest neonatal emergencies faced by paediatric surgeons. The incidence of oesophageal atresia varies from 1 in 2500 to 1 in 4500 live births with an average incidence of 2.4 per 10000 births. The most common type of oesophageal atresia is Vogt Type 3, in which there is a blind upper pouch with a fistula to the trachea or bronchus from the lower oesophagus. It occurs in about 85%--87% of patients.

To anastomose the ends of an infant’s oesophagus, the surgeon must be as delicate and precise as a skilled watchmaker. No other operation offers a greater opportunity for pure technical artistry. Vast advances in the management, both surgical and non-surgical, have greatly improved the survival of babies born with oesophageal atresia.

Methodology:

An Ongoing study is being carried out in a paediatric surgery unit in a teaching hospital in Mumbai. On an average, 20 newborns with oesophageal atresia are referred to the hospital per year. More than 80% are oesophageal atresia with distal tracheoesophageal fistula (Vogt’s type 3b; Gross type C).
Babies with this type of oesophageal atresia who survived primary end-to-end oesophageal anastomosis are included in the study. All the survivors were contacted by mailing letters asking parents to follow up. Of the 82 parents contacted 25 responded and are included in the study. During follow-up, a detailed history, clinical examination and imaging studies are carried out. The study is done under following heads:

1. **Basic data:**
Data pertaining to patient viz. name, date of birth, sex, address, registration, date and day of life at primary oesophageal anastomosis are recorded.

2. **Waterstone’s group and associated anomalies:**
Waterstone’s group and associated anomalies such as cardiac, (detected by echocardiography), anorectal and its type, renal, (sonography and when required MCU) vertebral and rib anomalies, radial and rib anomalies and any other anomalies are noted.

**Results:**

**Waterstone’s Group**

Of the 25 cases majority i.e. 10 belonged to Waterstone’s group Bl.

Table No. 1

<table>
<thead>
<tr>
<th>Waterstone’s Group</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group A</td>
<td>9</td>
</tr>
<tr>
<td>Group B1</td>
<td>10</td>
</tr>
<tr>
<td>Group B2</td>
<td>5</td>
</tr>
<tr>
<td>Group C1</td>
<td>1</td>
</tr>
<tr>
<td>Group C2</td>
<td>-</td>
</tr>
</tbody>
</table>

None of the cases who followed up belonged to C2 category.

**Associated Congenital Anomalies :**

Clinical examination, radiographs, ultrasonography and echocardiography were used to detect following associated anomalies in the study group.
Table No. 2  Congenital anomalies associated with OA with distal TOF

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No. of cases</th>
<th>Percentage %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vertebral</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Rib</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Anorectal</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>10</td>
<td>40</td>
</tr>
<tr>
<td>Radial</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Renal &amp; urinary</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Congenital Diaphragmatic hernia</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Coloboma</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

Commonest congenital anomaly associated with oesophageal atresia with tracheoesophageal fistula was congenital heart disease.

Discussion:
One patient has thoracic hemivertebra with bird rib on the corresponding side. One patient with 13 ribs has undergone primary oesophageal anastomosis. Of the two Cases with associated anorectal malformation, one had anocutaneous fistula and underwent cut back anoplasty on the same day as TOF repair. The other case had high anorectal malformation with anorectal agenesis with rectoprostatic fistula which necessitated high sigmoid colostomy and subsequent Stephen’s sacroperineal pull through and colostomy closure. One of the patients had malrotated thumb and is awaiting tendon - transfer as advised by plastic surgeons. One other patient has dysplastic kidney primary grade I vesicoureteric reflux which on follow up has subsided. One case had right Sided Morgagni’s diaphragmatic hernia with a sac containing liver. It was Corrected at the same time as primary oesophageal anastomosis but by transpleural approach. None of the cases had trisomy. The present study consisted of twenty-five cases of oesophageal atresia with distal tracheoesophageal fistula. All 25 children have successfully undergone primary oesophageal anastomosis. All the cases are treated in a pediatric surgical unit of a tertiary teaching hospital. One patient of the present study had right sided Morgagni type of congenital diaphragmatic hernia. The association of these two conditions viz OA and congenital diaphragmatic hernia is rare. Anastomotic leak following primary oesophageal anastomosis in typeIII/b OA. Of the 25 patients of type III/b OA with distal tracheoesophageal fistula who survived following primary oesophageal anastomosis, one patient had developed anastomotic leak.
This case had severe anastomotic tension. Holder and Ashcroft noted that anastomotic tension is one of the important etiological factors in development of anastomotic leak.

**Conclusion:**

Commonest congenital anomaly associated with oesophageal atresia with tracheoesophageal fistula was congenital heart disease.

**References:**


