Esophageal atresia and Tracheoesophageal Fistula
A Study of 100 Cases
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Abstract
BACKGROUND: Esophageal Atresia presents a challenging congenital abnormality which is incompatible with life and was referred to as "hopeless from the beginning and not of much practical importance to the surgeon" [1] is now successfully repaired all over the world.
AIM OF THE STUDY: To study the clinical presentation, diagnosis, types, management and complications of esophageal atresia in children admitted to welfare Teaching Hospital.
PATIENTS AND METHODS: A retrospective analysis of data of 100 neonates with esophageal atresia with or without tracheoesophageal fistula treated and followed up in children welfare Teaching Hospital, between January 2003 to January 2005.
RESULTS: Prenatal history of Polyhydramnios was evident in 77(77%) cases.
(65%) were males and (35%) were females with male to female ratio was 1.8:1
The presenting features were frothy secretions from the mouth and failure to pass nasogastric tube in 98(98%) cases , chocking after feeding in 87(87%)cases, dyspnea in 12 (12%)cases, cyanosis in 50(50%)cases, chest infection in 40(40%)cases , most of the patients 92(92%) cases were of the common type (proximal atresia with distal fistula), while 7 (7%) cases were of pure atresia type(without fistula), 1(1%) case was of H-type fistula.
Associated anomalies were present in 23(23%) cases.
Post operative complications were anastomotic leak seen in 24.6%, stricture seen in 18.5%, recurrent tracheoesophageal fistula in 2.4% and mortality rate was high constituted 65%.
CONCLUSION: Esophageal atresia with distal fistula was the commonest type in this study. Frothy secretion from the mouth, failure to pass nasogastric tube, chocking after feeding and Radiological study (Plain X-ray) were the main diagnostic features. Delay in diagnosis and associated anomalies were responsible factors for increasing incidence of mortality and morbidity.

Introduction
Esophageal atresia is relatively common congenital malformation occurring in about 1 in 2500 to 3000 live births. The etiology of the malformation is likely to be multifactorial and remains unknown. With slight male predominance [1].
The 1st successful primary anastomosis for esophageal atresia was performed in 1941, which was followed by a steady improvement in survival for babies with this condition [1,2]. The progression in the treatment was enabled by improved neonatal support due to advances in neonatology, neonatal anesthesia, nutritional support & antimicrobial therapy [3].

Patients and Methods
This is a retrospective study of 100
patients with esophageal atresia, with or without tracheoesophageal fistula who were admitted to Children Welfare Teaching Hospital in Baghdad from January 2003 to January 2005. These cases were studied as regard to their age, gestational age, sex, weight, family history, prenatal history, age of the mother, clinical presentation and associated anomalies. Diagnosis was based on history taken from the mother, clinical examination, supported by radiological examination via plain x-ray after passing a nasogastric tube for common type and isolated type of atresia. Operative findings, post operative complications, secondary operation and follow up were studied. Patients with esophageal atresia and distal tracheoesophageal fistula were managed by right thoracotomy through the fourth intercostal space using an extrapleural approach, dividing the fistula and end to end esophagoesophageal anastomosis by single layer using silk sutures, a drain tube left beside the anastomosis. For isolated esophageal atresia gastrostomy and cervical esophagostomy was done. In H-type fistula ligation of the fistula done from neck.

Results
Sixty five patients were males (65%) and thirty five patients were females (35%), a male to female ratio 1.8:1. Ninety cases (90%) were full term babies. Seventy nine patients presented in the first three days of life, while eight patients presented between fourth to seventh days of life. This mean that the presenting age in the first week (87%), table-1.

Table 1 Age of patients at presentation

<table>
<thead>
<tr>
<th>Age of patients</th>
<th>No. (%) of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>(1-7) days</td>
<td>87 (87%)</td>
</tr>
<tr>
<td>(8-14) days</td>
<td>10(10%)</td>
</tr>
<tr>
<td>(15-25) days</td>
<td>3 (3%)</td>
</tr>
</tbody>
</table>

Out of 100 cases 7 patients were cases of pure atresia while 92 cases were common type and one case was H-type.

Most of the patients presented with frothy secretion from the mouth and failure to pass NGT. These and other presentations are shown in table2.

Table 2 Clinical features

<table>
<thead>
<tr>
<th>Signs &amp;symptoms</th>
<th>No. of Patients</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frothy Secretions from the Mouth &amp; Failure to Pass NGT</td>
<td>98</td>
<td>98</td>
</tr>
<tr>
<td>Chocking after feeding</td>
<td>87</td>
<td>87</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>12</td>
<td>12</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>50</td>
<td>50</td>
</tr>
</tbody>
</table>
Polyhydramnios was positive in mothers of 77 patients (77%). Maternal age ranged from 21-30 years in 78 cases (78%).

According to Gross classification of tracheo-esophageal anomalies we classified our patients into three types: Esophageal atresia with distal tracheoesophageal fistula (common type) constituted 92 cases (92%). Isolated esophageal atresia without fistula constituted 7 cases (7%). Isolated tracheoesophageal fistula 1 (1%) case.

Associated Anomalies:
Twenty three patients have associated anomalies (23%). Cardio-vascular anomalies were detected in 12 (12%) cases. Imperforate anus was detected in 8 (8%) cases. Musculoskeletal anomalies detected in 3(3%) case. Genitourinary anomalies found in 1 (1%) case had hypospadias with high type imperforate anus.

Chromosomal anomalies : one 1(1%) case of trisomy 21, have also anorectal and cardiac anomalies.

Out of 100 cases, 9 cases were of common type and not operated on, two patients were discharged on responsibilities of their families, 7 cases were critically ill and died before operation. Four patients presented with severe respiratory distress, all of them were premature and died within few hours after admission.

Operation:
For the common type of esophageal atresia and distal tracheoesophageal fistula (83 patients) for two patients gastrostomy and esophagostomy was done, while 81 patients underwent primary repair. For isolated esophageal atresia type (7 patients) all underwent gastrostomy & esophagostomy as a first step for further management. For H-type fistula ligation of fistula done through incision in the neck ,table (3).

\[\text{Table 3 Type of operation}\]

<table>
<thead>
<tr>
<th>Type of atresia</th>
<th>Gastrostomy &amp; esophagostomy</th>
<th>Primary repair</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Common type</td>
<td>2</td>
<td>81</td>
<td>83</td>
</tr>
<tr>
<td>Pure atresia</td>
<td>7</td>
<td>-</td>
<td>7</td>
</tr>
<tr>
<td>Total</td>
<td>9</td>
<td>81</td>
<td>90</td>
</tr>
</tbody>
</table>

\[\text{Table 4 Complications of surgery}\]

<table>
<thead>
<tr>
<th>Complication seen</th>
<th>No. of patients</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leak</td>
<td>20</td>
<td>24.6%</td>
</tr>
<tr>
<td>Stricture</td>
<td>15</td>
<td>18.5%</td>
</tr>
<tr>
<td>Recurrent TEF</td>
<td>2</td>
<td>2.4%</td>
</tr>
</tbody>
</table>

Follow Up:
Total number who survived after these different procedures were 32 (35%) cases. Esophageal dilatation
done for those cases with stricture once or more according to the severity of stricture using balloon catheter, help to resolve the problem in most cases, 2 cases need gasterostomy with dilatation using string method, 1 case developed recurrent TEF.

1 case developed recurrent chest infection proved to be recurrent TEF, reoperation done with no post operative complication.

Mortality Rate:
The mortality rate was (65%) / The most common cause of death was respiratory infection, sepsis and associated congenital anomalies.

Discussion
Tracheoesophageal fistula is one of the anomalies that are associated with high morbidity and mortality [4].

From this study certain observation had been made:
Frothy secretion from the mouth and failure to pass nasogastric tube were the main diagnostic criteria of esophageal atresia. Eighty seven (87%) out of the 100 patients had choking after feeding which is equal to other studies [5].

Cyanosis was present in 50(50%) cases and pneumonia in 40(40%) cases.

The age of presentation in this study was (1-3 days) in 72(72%) cases, while in the study done by spraye and et al. The age of presentation was in the 1st post natal day in 106 out of 119 live born infants [6], this is due to poor neonatal assessment either by mid wife after labor or by the attending medical staff.

The common type of esophageal atresia and distal tracheoesophageal fistula in our study was 92% which coincides with other studies [6].

Isolated esophageal atresia accounted for 7% in our study which is comparable to the study done by Josephine and et al. [7].

Male to female ratio was 1.8:1 in our study which almost similar to others [7]. Polyhydramnios was positive in 77 (77%) of our patient; this is similar to what was reported by James O'Neill [4].

As in Bianca study [8], maternal age has not got any statistical significance and no age group was immune from having such anomaly in their babies.

Associated anomalies were present in 23%, which is lower than 50% reported by Ashcraft & James O'Neill [1,4]. These cases were of common type which differ from other reports in which associated anomalies where most common with pure atresia [1,7].

Cardiovascular anomalies, constituted 12% which is lower than other studies [7, 9, 10, 11], this lower incidence may be due to lack of proper diagnostic procedure which can't be diagnosed on clinical basis.

It had been mentioned that right sided aortic arch is present in 4 (4%), which is approximated to our study (3%).

Anorectal malformation in our study constituted 8%, this is lower than 18% in European studies [10], and musculoskeletal anomalies were present in 3%of our patients. This is lower than others [7, 10].

Our study shows that cardiovascular anomalies were the most frequent associated congenital anomalies; imperforate anus was the second commonest anomalies which is similar to other studies [1, 7, 10].

Eighty three (83%) cases of the common type esophageal atresia underwent primary repair. In 2 patients there was a long gap between the two ends of esophagus, so that primary anastomosis was impossible. In all ligation of tracheoesophageal fistula done and gastrostomy was added as a staged procedure. This is in comparison to other studies [7,12, 13,14] in which similar approaches
were used for dealing with long gaps or high upper pouch. Anastomotic leak as an early postoperative complication was identified clinically by presence of saliva in chest tube without routine contrast study [15]. Anastomotic leak in our study occurred in 24.6% which is comparable to 36% in other studies [7].

Esophageal stricture as a late postoperative complication occurred in 18.5% patients which is lower than 25% in M. Said and et al [16], since esophageal dilatation is recommended for those cases developed leak[15,16,17,18] , we considered every patient with esophageal stricture need esophageal dilatation twice or more.

In our study Recurrent tracheoesophageal fistula found in 2/81(2.4%) this lower than 10% in other studies [7]. Our mortality rate was 65% which are high in comparision to other studies [7,11,12]and this is due to lack of intensive neonatal care unit which is important preoperatively [12,20] while assessing and investigating the patients and post operatively especially in premature patients those with RDS or associated congenital heart disease, and due to late referral cases.

Conclusions and Recommendations
This study shows that the tracheoesophageal atresia remain a major problem in pediatric surgery because of delay in diagnosis, referral and associated anomalies that are responsible factors for increasing incidence of mortality and morbidity. We recommend that every mother with poly hydramnios should be screened for the presence of esophageal atresia in their newborns. All of them should be screened for associated anomalies.

The presence of intensive care unit is mandatory in the management of these cases.

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