

# Management of Patients with Esophageal Atresia with or without Tracheoesophageal Fistula, A 7 Years Experience

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**Objective:** To see the short term outcome in patients treated for esophageal atresia with or without tracheo-esophageal fistula in our setup. **Design:** Descriptive and retrospective. **Place and Duration:** Department of Paediatric Surgery & Department of Cardiothoracic Surgery, Postgraduate Medical Institute, Lady Reading Hospital Peshawar from January 1998 to December 2004 with duration of 7 years. **Materials and Methods:** After diagnosis, patients were put on broad-spectrum intravenous antibiotics, intravenous fluids, vitamin K injection and throat suction in a normothermic environment. Diagnosis was established mainly on clinical grounds and supported by passing a big size nasogastric suction tube (size 10 Fr) through the mouth into the esophagus and taking a plain X-ray chest of the patient. After adequate preparation, through a right thoracotomy in 5<sup>th</sup> intercostals space, fistula repair and esophageal anastomosis was performed extrapleurally in patients with tracheoesophageal fistula while gastrostomy and cervical esophagostomy was performed in patients with pure esophageal atresia. Transanastomotic tube was passed as a nasogastric tube size 10 to act as a stent and later on used for tube feeding. Blood transfusion was the routine during surgery and chest tube inserted to drain the retro plural space for 3-5 days. Broad spectrum intravenous antibiotics were continued post-operatively. All the patients were treated in the general ward without neonatal intensive care facilities which is lacking in our hospital. After complete recovery, patients were allowed tube feeding 24 - 48hours after operation. Patients were discharged to home 5-7 days after surgery and tube feeding was continued for a maximum of two weeks. Oral feeds were allowed after removal of the nasogastric tube and confirming the patency with gastrograffin contrast esophagogram. They were asked to come for follow up to Out Patients Department (OPD) regularly. **Results:** A total of 60 patients with esophageal atresia with or without tracheoesophageal fistula were admitted. There were 40 males and 20 females. Age ranged from 1-7 days. Weight of the newborn patients was in the range of 2 - 3Kg. Four patients had cyanotic congenital heart disease, two were with imperforate anus and two with spina bifida. All patients had some form of bronchopneumonia due to aspiration of upper pouch contents out of whom 30 patients had severe pneumonia. Six patients left the hospital against medical advice and 6 patients died before operation. Forty-eight patients were operated. Esophagostomy and gastrostomy was performed for pure esophageal atresia (10 Patients), while in 38 patients, a right thoracotomy in the 5<sup>th</sup> inter costal space with fistula ligation & esophageal anastomosis was performed. Three out of ten patients with pure esophageal atresia died, while eighteen patients with tracheoesophageal fistula died after surgery. Twenty-seven out of total 48 patients survived and were discharged to home after an average hospital stay of 7 days after surgery. Eleven patients were seen in follow up and treated accordingly. **Conclusion:** This study shows that majority of these patients presented late because of improper referral system. Delay in diagnosis and management led to various complications such as aspiration pneumonia, dehydration and septicemia with great mortality which is further increased due to nonavailability of the neonatal intensive care facility in our setup. Survival of these patients can be improved by early and proper referral system, specialized medical and surgical team, specialized anesthesia with personnel trained in neonatal anesthesia.

**Key words:** Esophageal atresia, tracheoesophageal fistula, Thoracotomy, Esophagostomy, Gastrostomy,

Maternal Polyhydramnios is common in atresia of esophagus with or without tracheoesophageal fistula<sup>1,2</sup>. Postnatally the baby is usually premature with excessive salivation and frothing in the mouth. Attempts of feeding causes choking or cyanosis<sup>1,2</sup>. When a 10 Fr feeding tube is passed through the nose or mouth into the esophagus, it is arrested in the upper pouch at about 10cm distance from the mouth or nose. A plain radiograph of the chest and abdomen with the tube in place usually confirms the diagnosis. Gas in the bowel confirms the presence of tracheoesophageal fistula while a gasless abdomen indicates pure esophageal atresia<sup>1,2,3,4</sup>. Other congenital anomalies are also diagnosed which may be associated with it. The aim of treatment is to ligate the fistula and restore the esophageal continuity by end-to-end anastomosis. When this is not possible, the management is

staged, either to do delayed end-to-end esophageal anastomosis or esophagostomy & gastrostomy and later on esophageal replacement or stomach pull up procedures are performed<sup>1,2,5</sup>. Stricture esophagus, gastro esophageal reflux disease (GERD) and recurrent respiratory tract infections are long term complication associated with this condition<sup>2,5,6,7</sup>.

## Material and methods:

A total of 60 newborn patients were treated who were admitted to paediatric surgery ward through emergency and also shifted from other wards of this hospital and from other hospitals of the province. The diagnosis was mainly established on clinical grounds with high index of suspicion of this condition, which was confirmed by passing a 10 FR nasogastric tube through the nose or

mouth into esophagus and taking a plain X-ray chest and abdomen. Patients were put on broad spectrum intravenous antibiotics (3<sup>rd</sup> generation cephalosporin), intravenous fluids (175<sup>th</sup> 10% Dextrose/Saline) and vitamin K injection. Patients were kept nil per orally with regular throat suction with mucus extractor in a normothermic environment. After adequate preparation, patients were operated on elective list. Under general anesthesia with endotracheal tube, a right posterolateral thoracotomy in the 5<sup>th</sup> intercostals space was performed and esophagus approached extrapleurally. Tracheoesophageal fistula was ligated with 2/0 silk suture while end-to-end tension free anastomosis was performed with 4/0 silk suture in interrupted fashion. A transanastomotic tube was passed into the stomach to act as a stent and later on used for tube feeding. In patients with wide gap pure esophageal atresia, cervical esophagostomy and gastrostomy was performed and later on esophageal replacement after the age of 12 months.

Blood transfusion was routinely given to patients during surgery and chest tube inserted to drain the retroplural space for 3-5 days. Broad spectrum intravenous antibiotics and intravenous fluids were continued post-operatively. All patients were treated in the general ward without neonatal intensive care facilities which is lacking in our hospital. After complete recovery, patients were allowed tube feeding 24-48 hours after operation. Patients were discharged to home 5-7 days after surgery and tube feeding was continued for a maximum of two weeks in case of esophageal anastomosis while gastrostomy tube feeding was continued till esophageal anastomosis or replacement was performed.

After removal of the N/G tube a contrast esophagogram (Gastrograffin) was performed to see the patency and check any anastomotic leak. When esophageal integrity was confirmed, oral feeds were allowed. Parents were advised to bring their babies to outpatient department regularly. Any complaints or problems encountered in the follow up visit were recorded and treated accordingly.

**Results:**

Sixty newborn patients with esophageal atresia with or without tracheoesophageal fistula were admitted to paediatric surgery ward over a period of seven years. Ten patients had pure esophageal atresia while 50 had lower pouch tracheoesophageal fistula with atresia. with 40 male and 20 female patients (table-1). Age ranged from 1-7 days. Weight of these patients was in the range of 2-3 kg.

Four patients had cyanotic congenital heart disease, 2 were with imperforate anus, 2 with spina bifida and one with bilateral club feed (table-2). All patients had some form of bronchopneumonia. Thirty patients had severe bronchopneumonia and were declared unfit for anesthesia and surgery. Six patients died before operation and 6 patients left the hospital against medical advice. Forty

eight patients were operated. Cervical esophagostomy and gastrostomy was performed in 10 patients who were having pure esophageal atresia. Right thoracotomy in the 5<sup>th</sup> intercostals space with fistula ligation and esophageal anastomosis was performed in 38 patients who had esophageal atresia with lower pouch tracheoesophageal fistula.

Three patients out of ten pure esophageal atresia died after surgery while 18 out of 38 patients with tracheoesophageal fistula died post-operatively. Twenty-seven patients survived out of total 48 patients after surgery and were discharged to home after an average hospital stay of 7 days after surgery (table-3). Eleven patients with tracheoesophageal fistula repair were seen in follow up. Six patients had recurrent respiratory tract infection needing intravenous antibiotic therapy and repeated admissions. Two patients developed stricture esophagus who were treated by repeated esophageal dilatation. Patients with pure esophageal atresia did not come for definitive treatment and were lost to follow up.

Table 1: Type of esophageal atresia and sex distribution

Type of esophageal atresia	=n	Male	Female
Pure esophageal atresia	10(16.6%)	07	03
:Atresia with tracheoesophageal fistula	50(83.3%)	33	17
Total	60	40(66.6%)	20(33.3%)

Table 2: Anomalies and conditions associated with esophageal atresia.

Conditions	=n
Mild to moderate bronchopneumonia.	60(100%)
Severe bronchopneumonia	30 (50%)
Cyanotic congenital heart disease	04 (6.66%)
Anorectal malformation	02(3.33%)
Spina bifida	02 (3.33%)
Club feet	01(1.66%)

Table 3: Type of esophageal atresia and survival after surgery

Type of esophageal atresia	=n	Survival	expired
pure esophageal atresia	10	03(30%)	07(70%)
B:Esophageal atresia with tracheoesophageal fistula	38	18(47.3%)	20(52.6%)
Total	48	21(43.7%)	27(56.2%)
Pure esophageal atresia	10	03(30%)	07(70%)

**Discussion:**

Esophageal atresia is a congenital disorder that traditionally requires surgery in the first few days of life. With advances in the neonatal care there is tremendous

improvement in the survival of these patients needing thoracotomy and repair of the fistula and esophagus. However recently further advances in the new technology of thoracoscopic procedures have been introduced, which can decrease the technical difficulties of esophageal reconstruction<sup>6,7,8</sup>.

Repair of wide gap esophageal atresia remained a problem for a long time. Esophagostomy and gastrostomy and later on esophageal replacement with colon, jejunum, gastric tube and stomach has been performed by various authors and being practiced all over the world<sup>1,2,9,10</sup>. The problem of conduit redundancy, gastroesophageal reflux, stricture and epithelial metaplasia remains unavoidable<sup>2,11,12</sup>. There is no substitute for esophagus, which is better than esophagus and all efforts are made to reconstruct and preserve the native esophagus. Different methods have been adopted to restore esophageal continuity by doing delayed primary repair, intrathoracic or extrathoracic esophageal lengthening procedures such as longitudinal upper pouch flaps, circular myotomies or pulling on the upper pouch to increase the length of esophagus and aim a tension free esophageal anastomosis<sup>2,4,13,14,15</sup>.

The results of our study are different from the literature. Preoperative mortality and refusal by parents to get treatment and leaving the hospital against medical advice were strange and important factors to adversely affect the management of these patients. Majority of the patients were having respiratory tract infection leading to septicemia and death of these patients. Delay in the diagnosis, lack of awareness of the condition and late referral were common factors, which led to significant unfavorable consequences before and after surgery. Lack of neonatal intensive care facilities was another important factor, which can be the cause of great mortality and morbidity in these patients. Twenty-one out of 48 patients died postoperatively showing 43.75% mortality. Though the mortality rate is high in this study as compared to other studies but it is acceptable in our set up where the overall neonatal care facilities are deficient on one hand and delayed presentation, lack of awareness and poor referral system on the other hand. Low birth weight, aspiration pneumonia, hypothermia dehydration, metabolic disturbances and other congenital anomalies were the major contributing factors, which led to significant peri-operative mortality.

Survival rates have continued to improve recently with an expected rate exceeding 90% as reported by various studies<sup>1,2,16,17,18,19</sup>. Early thoracotomy and attempted primary repair of esophagus is advocated provided the patients condition permits<sup>2,20,21,22</sup>. Unavoidable conditions such as low birth weight and associated major congenital anomalies are well known factors that adversely affect the short term outcome of treatment in these patients<sup>23,24,25</sup>. However preventable conditions such as aspiration pneumonia, hypothermia

metabolic disturbances & fluid & electrolyte imbalance are considered to be important factors, which are of primary concern in our setup. These physiological derangements seen in our patients may not be a serious problems in other centers in this modern world but still of great concern which may partly explain the high perioperative mortality in these patients<sup>1,2,26,27</sup>.

Another strange observation was that there were only two types of esophageal anomalies found in these patients. Esophageal atresia was seen in 10 (16.6%) patients and atresia with distal fistula was seen in 50 (83.3%) patients. Other types such as H-type fistula, upper pouch fistula or both ends fistula were not encountered in this study. Distal tracheoesophageal fistula is the most common (85.8%), the next most common is pure esophageal atresia (7.8%) followed by the other types, which are collectively found in 6.4% of patients<sup>2,28</sup>. The reason of not finding patients with these rare types of esophageal malformations is not known. It is presumed that probably they were missed in patients who died before surgery or left the hospital without medical advice. If we restrict our study to only operated patients, then the findings are different, that is 20.83% patients with pure esophageal atresia and 79.16% with tracheoesophageal atresia, which are not in comparison with the literature<sup>2,4,8,29,30</sup>.

It was also observed in this study that majority of the patients had aspiration pneumonia and other major congenital anomalies. All the patients fell into Waterston group B and C. There was not a single patient who could be put in Waterston group A, which carries good prognosis. The survival of patients in-group B is 85% and C is 65% respectively.

The survival in these groups is low due to associated serious conditions and other major congenital anomalies<sup>2,4,31,32</sup>. This was the reason that our observation was not in comparison with the literature. In our study it was also seen that severe bronchopneumonia and congenital heart disease was the major associated problems leading to poor prognosis groups. Aspiration pneumonia was found in all patients with delayed presentation whose parents continued to feed their babies till they were admitted to the hospital.

Follow up of these patients showed that none of the patients with pure esophageal atresia came for further treatment after cervical esophagostomy and gastrostomy. They were counted in the early survivors in this study but might have died at home or might have gone to some other centers for further treatment. Eleven patients out of 20 survivors were irregularly seen in the follow up with various complications such as dysphagia, vomiting, recurrent chest infections, failure to thrive and malnourishment. Only two patients were regular in follow up and were found to have stricture esophagus and were treated with esophageal dilation.

It is clear from our study that the short term outcome, although not meeting the international standards but still

encouraging in our setup where there is lack of back up support which is needed for the management of a critical newborn patient. It is therefore concluded that a lot much can be done to improve the early results of treatment in these patients. Survival of these patients can be improved by the provision of early diagnostic, measures, proper referral system, specialized medical and surgical team, specialized anesthesia with personnel trained in neonatal anesthesia and with the advent of neonatal intensive care facilities. Prevention of aspiration pneumonia, control over infection, correction of dehydration and adequate treatment of septicemia in a critical newborn patient are the measures which shall be taken in the management of these patients apart from surgical skills and technicalities which are modified from time to time to improve the outcome of treatment in esophageal atresia.

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