# Assessment Of The Outcomes Of Neonates With The Diagnosis Of Esophageal Atresia And/Or Tracheoesophageal Fistula Admitted To Tikur Anbessa Specialized Hospital-Neonatal Intensive Care Unit

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## Abstract

***Background:***

*Esophageal atresia and/or tracheoesophageal fistula are a fairly common congenital anomaly which is associated with serious morbidity and high rate of mortality in newborns.*

***Objective:***

*To assess the outcomes of neonates with the diagnosis of esophageal atresia and/or tracheoesophageal fistula (EA and/or TEF) admitted to Tikur Anbessa Specialized Hospital, Neonatal Intensive Care Unit (TASH-NICU) from Sep., 2008 to Aug., 2013.*

***Methodology**:*

*This is a retrospective study in which clinical records of all neonates admitted to TASH-NICU with the diagnosis of EA and/or TEF from Sep., 2008 to Aug., 2013 were retrieved and those newborns whose charts had been filled with necessary information were selected as study subjects. A structured questionnaire was used to collect necessary data about each case. Outcome, possible determinant factors of outcome, associated congenital anomalies, treatment provided, and complications were analyzed.*

***Results**:*

*About 41 cases of EA and/or TEF were admitted to TASH-NICU during the study period. There were 34 eligible cases. Some of the cases whose charts were lost or incompletely filled were excluded from the study. Among the eligible ones, 19 (**55.9%)** were females and 15 (**44.1%)** were males. There were only 5(14.7%) newborns that were discharged alive. Most died during the neonatal age (85.3%). Surgical intervention was performed for only half of the cases. Aspiration pneumonia was the most common reason for delayed surgical intervention. It was also the leading cause of respiratory failure and death of the newborns. There was a strong association between mortality and birth weight (P-value < 0.05).*

***Conclusion***

*This study showed that most cases of EA and/or TEF admitted to TASH-NICU died during neonatal age. Low birth weight has been strongly associated with higher risk of mortality in neonates with the diagnosis of EA and/or TEF.*

*Supportive care provided to newborns with EA and/ or TEF was not sufficient as it didn't include life-saving treatments like mechanical ventilation and total parenteral nutrition.*

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***Recommendation***

*The department of pediatrics and child health should develop a management guideline which incorporates appropriate and sufficient supportive care in addition to early surgical intervention to improve the outcome of newborns admitted to TASH-NICU with the diagnosis of EA and/ or TEF. Early recognition and surgical management & intensive care are important in salvaging these newborns. Health care providers particularly those who are involved in newborns care immediately after delivery should always be aware of the fact that neonates with respiratory distress and frothy salivation could have EA and/ or TEF and should be assessed before they are fed. By doing so, aspiration can be prevented and outcome improved. Those who can provide obstetric care to pregnant mothers should also be cognizant of the fact that polyhydramnios with absent fluid-filled fetal stomach on ultrasonography could be associated with presence of EA and/ or TEF.*

## INTRODUCTION

Globally, about 4 million newborns die every year during the neonatal period. Almost all neonatal deaths occur in low and middle income countries. The highest numbers of neonatal deaths are in south-central Asian countries and in Sub-Saharan Africa (1).

Ethiopia is one of the countries with high neonatal and infant mortality rates. According to EDHS 2011, the neonatal mortality rate is **37/1000LBs** & IMR is **59/1000LBs**.

EA and/or TEF are relatively common malformations occurring in approximately 1 in 3500 births. In around half of the cases (syndromic EA), there are associated anomalies, with cardiac malformations being the most common. In the remainder (non-syndromic cases), EA/TEF occur in isolation (2).

This study has revealed that the outcome of management of EA and/or TEF in our hospital is very poor**.** Most cases died during neonatal period. This high mortality rate of neonates with esophageal atresia and/or tracheoesophageal fistula contributes to the neonatal & infant mortality rate of the country.

Early diagnosis, improved surgical technique, neonatal anesthesia, sophisticated ventilatory support, advanced intensive care management, early treatment of associated anomalies, responsiveness of anastomotic strictures to dilatation, and aggressive treatment of gastroesophageal reflux have influenced survival positively. With few exceptions, most infants with EA and/ or TEF should survive in the current era (3).

The management of newborns with EA and/or TEF has evolved considerably over the years. Currently, an overall survival of 85% to 90% has been reported from developed countries. In developing countries, several factors contribute to higher mortality rates. Factors contributing to mortality included **prematurity**, **delay in diagnosis with an increased incidence of aspiration pneumonia** and a **shortage of qualified nurses (4)**.

### Importance of the study

The results of the study will hopefully create awareness among health care professionals about the outcomes of cases of EA and/or TEF. This will remind health workers to improve their management and to prevent deaths in neonates admitted to TASH-NICU with the diagnosis of EA and/or TEF which in turn will partly help to reduce neonatal mortality rates.

## OBJECTIVES OF THE STUDY

### General objective

- To assess the outcomes of neonates admitted to TASH-NICU from Sep., /2008 to Aug.,/ 2013 with the diagnosis of EA and/or TEF.

### Specific objective

- To determine the prevalence of EA and/ or TEF in neonates admitted to TASH-NICU from Sept., 2008 to Aug./ 2013.
- To identify factors associated with poor outcome.
- To assess the pre- & postoperative care provided.
- To describe the type & prevalence of complications.
- To identify common possible causes of death.

## Materials and Methods

**Study Area**- The study was conducted at Addis Ababa University, College of Health Sciences, Department of Pediatrics and Child Health, TASH- NICU. This is the largest NICUs in the country.

TASH-NICU has six rooms and 40 beds: one room for preterms, two rooms for terms & post-terms (one room for critical neonates & one room for relatively stable ones), and 3 rooms for neonates under kangaroo mother care & others.

Different activities are conducted in the NICU:- undergraduate, postgraduate & fellowship programs; research; inpatient health service provision to admitted neonates & outpatient follow-up of high risk infants. There are two neonatologists and nineteen nurses trained in neonatal care. There are three pediatric surgeons & two pediatric surgery fellows in TASH.

**Study population**- All newborns admitted to TASH-NICU during the specified study period.

**Study subjects**: Newborns with the diagnosis of EA and/or TEF admitted to TASH-NICU during the study period.

**Study Design**- A retrospective study was conducted on the outcomes of cases of EA and/ or TEF admitted to TASH-NICU from Sep., 2008 to Aug., 2013. Data have been collected from the charts of study subjects using a questionnaire. Prevalence of EA and/ or TEF, factors associated with poor outcomes, types of therapy, congenital anomalies, complications, and causes of death were assessed.

**Study Period**: Sep., /2008 to Aug. /2013.

**Inclusion & Exclusion Criteria**

**Inclusion Criteria:** Neonates diagnosed to have EA and/ or TEF based on clinical & radiographic evidences and whose charts were available & filled with necessary information were included in the study.

**Exclusion criteria**: Neonates whose charts are incompletely filled & neonates whose charts are lost were excluded.

**Variables of the Study**

**Dependent variables:** postoperative complications, neonatal deaths and neonatal survival.

**Independent variables:** Gestational age, birth weight, sex, age at admission, time of surgery after admission, history of being fed.

**OPERATIONAL DEFINITIONS**

**Neonate**: A newborn from birth to the age of 28 days of life.

**Gestational age**- The length of gestation determined from last menstrual period (LMP) or

Ballard score or ultrasound determination if LMP is not reliable or unknown.

**EA**- is defined as a complete interruption in the continuity of the esophageal lumen.

**TEF**- may be defined as a congenital, fistulous connection between the proximal and/or distal esophagus, and the airway.

While EA and TEF may exist as separate congenital anomalies, great majority of patients with these congenital malformations have both EA and TEF. As EA, TEF, and EA and TEF generally have similar associations with other anomalies and complications, they will be considered together.

**Poor outcome**: postoperative complications or neonatal death.

**Ethical Considerations**

Approval was obtained from the Department of Pediatrics and Child Health research committee and IRB of College of Health Sciences, AAU.

**DATA ANALYSIS**

**Data quality management**: A structured questionnaire was prepared and the needed data collected from records of neonates who were diagnosed to have EA and/or TEF. Charts of each selected study cases were revised thoroughly in order to get the required data.

**Data entry and processing**: After data were coded and entered, SPSS version 20 was used to get important parameters, significance of testing, and data output was put in tables and graphs.

**Statistical methods**: Risk factors determination for association of predictors and dependent variables has been compared using chi-square test. Relative risk & odds ratio were also used to compare statistically significant associations (p-value <0.05).

**RESULTS**

There were about **41** newborns admitted to TASH-NICU during the specified study period diagnosed to have EA and/ or TEF but only 34 had charts with complete information. So they were selected to be the study subjects. Among the study subjects, 15(44.1%) were males and 19 (55.9%) were females.

Table 1. Patient Demographics and Parameters

| | Maternal Age in Years | Gravidity | Parity | GA in weeks | Neonatal Age in Hours | Birth weight in grams | Hospital Stay in Days |
|---|---|---|---|---|---|---|---|
| N | 34 | 34 | 34 | 34 | 34 | 34 | 34 |
| Mean | 26.35 | | | 38.1 | 60.62 | 2478.68 | 15.85 |
| Median | 26 | 2 | 1 | 39 | 48 | 2600 | 10.50 |
| Mode | 28 | 1 | 1 | 39 | 120 | 2800 | 4 |
| SD | 4.77 | | | | 57.57 | 625.01 | |
| Range | 20 | | | | 263 | 2400 | |
| Minimum | 18 | 1 | 1 | 28.4 | 1 | 1300 | 3 |
| Maximum | 38 | 7 | 4 | 43.6 | 264 | 3700 | 90 |

There were 8/34 (23.5%) preterm newborns with GA <37weeks; term newborns were 24/34(70.6%) with GA of 37-42 weeks; and post-terms were 2/34(5.9%) with GA>42 weeks. Only 11/34 (32.4%) of the newborns presented within the 1st 24hrs after birth and 24/34 (70.6%) within 72hrs; about 29.4% (10/34) presented after 72hrs of age.

Birth weight ranged from 1300 to 3700grams. 14/34 (41.2%) were LBW/VLBW & 20/34 (58.8%) had normal birth weight. About 66.7% (18 neonates) lost weight ranging from 0.4% to 22.5% of admission weight during their hospital stay. According to the birth weight in relation to the gestational age, there were 10 (37%) small for gestational age (SGA) and 17 (63%) were appropriate for gestational age (AGA).

Maternal age ranged from 18 to 38 years. Mean maternal age is 26 years. About 82.4% of the mothers were less than or equal to 30 years. Gravidity ranged from 1 to 7. Primigravidae constituted 47.1%. Parity ranged from 1 to 4; more than half were primiparae.

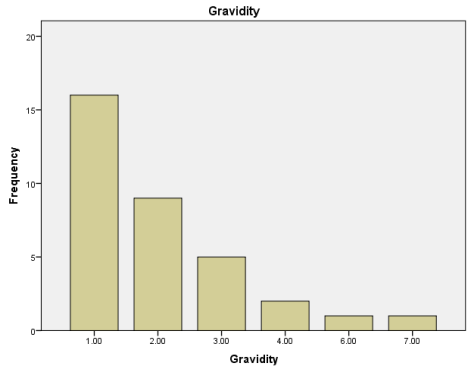


**Figure 1**

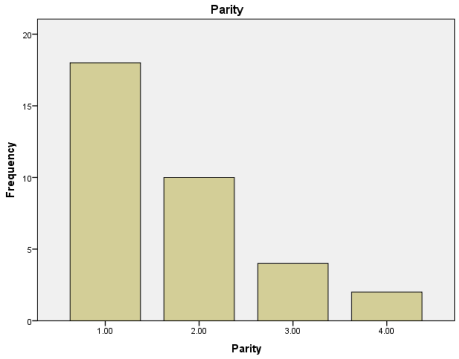


**Figure 2**

Most of the mothers had regular ANC follow-up (97.1%). Prenatal ultrasonography was done for 13 (38.2%) mothers of whom 10 (**76.9%**) were found to have polyhydramnios.

Mothers who delivered in a hospital were 27 (76.9%); in a health center 4(11.8%). Most 32 (94.1%) neonates were referred from a hospital. Nineteen (55.9%) neonates had been fed immediately after birth.

Respiratory distress and frothy salivation were reasons for referral in 23 (67.6%) newborns. Even though 9 (37.5%) neonates were either born in or taken to a health institution, diagnosis was not considered by the health professional. Distance from TASH was a reason for delayed presentation in 13 (54.2%).

Based on type of lesion, 24 (82.8%) cases were having EA with distal TEF (type A); 5 (17.2%) were cases of isolated EA (type B). In 5 cases, the type of lesion was not clearly described. Associated congenital anomalies were found in 11 (32.4%) neonates.

Most frequent admission diagnosis other than EA and/or TEF was aspiration pneumonia (81.3%).

The management provided includes oxygen, intravenous fluid, antibiotic, positioning, and suctioning in 58.8% of cases. Gastrotomy tube was placed for only 4 cases. No mechanical ventilatory support or total parenteral nutrition was given.

Definitive surgery was done for only 17 (50%), and 58.8% cases were operated after 3$^{rd}$ day of admission. Most common reason for delayed surgical intervention was the presence of aspiration pneumonia in 26 (81.3%) of the cases.

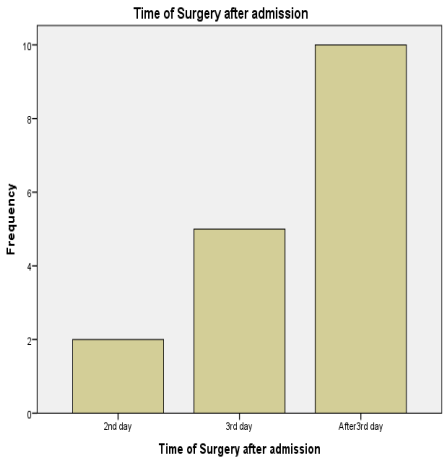


Figure 3

There were 6 cases (35.3%) with postoperative complications. Complications include sepsis in two cases, Anastomotic leak in 4 cases, and stenosis in two cases. Three of the cases with postoperative complication weredied.

Only 5 (14.7%) neonates were discharged alive and 29 (85.3%) died. Most (58.6%) of the cases died before surgical intervention and 41.4% died after definitive surgery was done. No neonate died during the time of surgery.

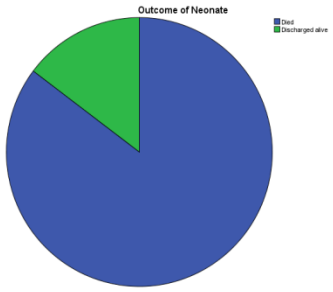


Figure 3

Respiratory failure secondary to aspiration pneumonia was mentioned as a possible cause of death in 21 (72.4%) of the cases and sepsis in 8 (27.6%) of cases. Early neonatal death occurred in 11 (37.9%) of the cases and late neonatal death in 17 (58.6%) of the cases.

Statistically significant association was found between low birth weight and poor outcome using Pearson Chi square (p-value<0.05). Prematurity is associated with a higher risk of mortality (RR=1.238, 95% CI 1.026-1.494). Low birth weight is also associated with higher risk of mortality (RR=1.333, 95% CI 1.035-1.717).

## DISCUSSION

Among the newborns in this study, associated congenital anomalies were diagnosed in 32.4%. The lower percentage of associated congenital anomalies in this study may be due to the fact that most neonates were not investigated exhaustively using imaging modalities in addition to the clinical assessment.

Respiratory distress, frothy salivation and difficulty of feeding were the main complaints that led to referral of most of the cases. There were more females than males unlike the male predominance in other studies elsewhere. Young maternal age is associated with having a baby with EA and/or TEF as is seen in this study in which most mothers (82.4%) were less than or equal to 30 years of age.

In one survey, the total prevalence rate of TEF and EA in 15 European countries registries covering 1,546,889 births during 1980-8 was 2.86 per 10,000. Sixty two per cent of cases were males. There was a significantly increased risk for mothers of less than 20 years of age (5).

The VACTERL complex refers to anomalies of the bony spinal column, atresias in the gastrointestinal tract, congenital heart lesions, tracheoesophageal defects, renal and distal urinary tract anomalies and limb lesions. In our setup, during the study period, associated congenital anomalies were found in 32.4% of cases with congenital heart disease being the most common of the anomalies (54.5%) which was also true in other studies. Anorectal (45.5%), renal anomalies (27.8%), vertebral (18.2%), and limb (18.2%) follow in that order. In a retrospective review conducted by Keckler SJ, et al, on all patients diagnosed with esophageal atresia between 1985 and 2005, a male predominance was seen with 62 males and 50 females. The categorical breakdown of anomalies were vertebral (24.1%), atresia (14.3%), cardiac (32.1%), TEF (95.5%), urinary (17.0%), skeletal (16.1%) and other (10.8%).The overall survival was 92.9% (6).

Congenital tracheoesophageal fistula (TEF) without esophageal atresia, commonly referred to as H-type fistula, comprises only 4.2% of all TEF's (7).

However, H-type TEF was not found in this study probably due to the fact that most of these cases present beyond neonatal age. In addition, there is difficulty in diagnosing them.

In this study, among the mothers who had an obstetric ultrasonographic examination, most of them were found to have polyhydramnios. However, the possibility of EA and/or TEF was not considered in any of these mothers. Most cases of esophageal atresia are not suspected

prenatally. Among fetuses with ultrasonographic features suggestive of esophageal atresia, 50% have the disorder confirmed postnatally. TEF or EA should be considered when there is polyhydramnios and an absent fluid-filled stomach.

Sonography will detect approximately one third of fetuses with TEF; an improved outcome is expected in these fetuses (8,9). In this study, 35.3% neonates had postoperative complications which include sepsis, anastomotic leak, and stenosis.

Early and late complications after repair of EA can be identified and potentially anticipated based on clinical findings at the time of repair and during the postoperative period (10).

Respiratory and gastrointestinal complications occur frequently, and may persist lifelong. Late complications of EA and/or TEF include tracheomalacia, a recurrence of the TEF, esophageal stricture, and gastroesophageal reflux (11).

The most common cause of death in our setup was respiratory failure secondary to aspiration pneumonia followed by sepsis. A retrospective review of newborns admitted to Hospital Sultanah Bahiyah, in Malaysia, from 1st January 2000 to 31st December 2009 was done. The mortality rate was 23% and the causes of death were pneumonia (36%), renal failure (18%), cardiac malformation (18%) and multiple congenital malformations (28%) (12).

In this study, the mortality was 85.3%, respiratory failure secondary to aspiration pneumonia being the most frequently documented possible cause of death with only 14.7% survivors who were discharged alive. This is exactly opposite to what is achieved in the developed world.

The improvement of survival observed over the previous two decades in the developed countries is multifactorial and largely attributable to advances in neonatal intensive care, neonatal anesthesia, **ventilatory** and **nutritional support**, antibiotics, **early surgical intervention**, surgical materials and techniques (13).

In this study, surgery was done only for half of the cases and most neonates were operated after the third day of admission. The supportive care that was provided includes supplementary oxygen, IV fluid, antibiotics, positioning, and suctioning. No mechanical ventilator or total parenteral nutrition was utilized. The combined effect of several factors has resulted in higher mortality of cases of EA and/or TEF in our setup.

## LIMITATIONS

- It was not possible to retrieve all the charts because the registration numbers of some of the cases were not documented in the logbook.
- This is a retrospective study and it is not possible to control the quality of data. Therefore, the quality largely depends on the already existing secondary data.

## CONCLUSION

Even though there were 41 cases of EA and/or TEF during the study period, only 34 of these cases were eligible and selected as study subjects. However, from the analysis of the eligible cases, it is found that respiratory failure secondary to aspiration pneumonia was the leading cause of death in neonates admitted to TASH-NICU with the diagnosis of EA and/or TEF during the specified study period.

## RECOMMENDATIONS

Prenatal obstetric ultrasonography is recommended especially in those pregnant mothers who are clinically suspected to have polyhydramnios.

Delivery room staff in health institutions should be oriented about the early signs of EA and/or TEF and they should be vigilant to detect cases before neonates are fed so that aspiration can be prevented.

Timely surgical intervention is mandatory as it is obvious that almost all cases die if surgery was deferred.

Postoperative intensive neonatal care needs to be improved so that neonates should not die after surgery. Mechanical ventilator, total parenteral nutrition, and well-trained and committed nursing staff should be available.

Ministry of Health of Ethiopia should create awareness of these cases among health care workers involved in maternal and newborn care at each level of the health care system.

It will be wise to conduct prospective study on this issue to have a better understanding of the extent of the problem and assess the current health service being given.

Recording and documentation in the neonatal intensive care unit should be improved.

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