# *Prospective study of outcome of Esophageal Atresia with or without fistula at Cairo University Children's Hospital*

THESIS

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# ABSTRACT

Esophageal atresia encompasses a group of congenital anomalies comprising an interruption of the continuity of the esophagus combined with or without a persistent communication with the trachea. Esophageal atresia is a relatively common congenital malformation occurring in one in 2500–3000 live births. During the last 40 years there has been a steady improvement in the overall survival rate due to early diagnosis and prompt referral, improvements in preoperative care and diagnosis and treatment of associated anomalies, advances in anesthetic techniques and sophisticated neonatal intensive care.

Results have improved with time, as the survival rates have improved, also the number of patients that had successful primary esophageal anastomosis have increased with a decrease in the rates of leakage. However, the mortality rate is still high in comparison to published results. Also, the survival rates in comparison to Waterston and Spitz classification are still low, indicating that our results are still unsatisfactory. Outcome was greatly affected by several preoperative factors such as delay in referral of patients affecting the age on admission and operation, chest condition preoperatively, cardiac anomalies, birth weight and other associated anomalies. The surgical technique also affected outcome as it affected post operative complications.

**KEY WORDS:** Esophageal Atresia. Primary esophageal anastomosis. Cardiac anomalies.

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# INDEX

# LIST OF ABBREVIATIONS

| | |
|---|---|
| EA | Esophageal Atresia. |
| TEF | Tracheoesophageal fistula. |
| VSD | Ventricular Septal Defect. |
| ASD | Atrial Septal Defect. |
| PDA | Patent Ductus Arteriosus. |
| IUGR | Intrauterine Growth Retardation. |
| GER | Gastroesophageal Reflux. |
| LES | Lower Esophageal Sphincter. |
| TAT | Transanastomotic Tube. |

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# INTRODUCTION

Esophageal atresia encompasses a group of congenital anomalies comprising an interruption of the continuity of the esophagus combined with or without a persistent communication with the trachea. Esophageal atresia is a relatively common congenital malformation occurring in one in 2500–3000 live births.[1]

During the last 40 years there has been a steady improvement in the overall survival rate due to early diagnosis and prompt referral, improvements in preoperative care and diagnosis and treatment of associated anomalies, advances in anesthetic techniques and sophisticated neonatal intensive care.

In light of the worldwide improvement in survival rates, and due to the large numbers of EA treated at Cairo University Specialized Pediatric Hospital, it is now mandatory to take a closer look at the management protocols and outcomes.

# REFERENCES

[1]. ***Spitz L:*** Oesophageal atresia. Orphanet Journal of Rare Diseases. 2007; 2: 24.

# AIM OF WORK

Proper assessment of preoperative and surgical approach of patients with Esophageal Atresia with or without fistula and their effect on early outcome, admitted in Neonatal Surgical Unit, Cairo University Hospital.

# I. HISTORICAL PERSPECTIVE

William Durston is believed to have provided the first description of esophageal atresia (EA) in the year 1670 when he published “a narrative of a monstrous birth in Plymouth...”in which he described a blind-ending upper esophageal pouch in a right infant of a set of thoracopagus-conjoined twins. The first description of EA with the typical form of tracheoesophageal fistula (EA-TEF) appeared in 1697 in the fifth edition of Thomas Gibson’s *The Anatomy of Humane Bodies Epitomized.* After that there were several anecdotal reports until the association of esophageal atresia with other structural abnormalities was recognized by Thomas Hill in 1840. Published in the *Boston Medical and Surgical Journal,* this report described an infant with EA-TEF and associated rectal agenesis with a rectourinary fistula.

In 1861 Harald Hirschsprung of Copenhagen brought together a series of 14 cases of EA-TEF, 4 cases of his own and 10 collected from the literature. A more complete account of the embryology, pathology and clinical diagnosis, including associated anomalies such as spina bifida, horseshoe kidney and imperforate anus was provided by Morrell McKenzie in 1880. He reported 57 cases of congenital esophageal malformation with 37 examples of tracheal or bronchial esophageal fistula. E. D. Plass, an instructor in obstetrics at the Johns Hopkins University, surveyed the literature in 1919 and reported 136 verifiable cases of EA, including 92 with an associated TEF. By 1931, Rosenthal had collected data on 255 patients and indicated, as Ladd emphasized in 1944,[2] “that atresia of the esophagus is a much more frequent anomaly than it has usually been considered to be”.

The history of the surgical treatment of EA-TEF is remarkable and covers 270 years linking the first description and first survivor. In 1869, Timothy Holmes of London, the author of *Surgical Management of Children's Diseases*, was the first to suggest the possibility of a surgical esophageal anastomosis in infants who had EA without TEF; however he added that "the attempt ought not, I think, be made". The first attempt to repair esophageal atresia without TEF was by Charles Steel in 1888 when he pushed a metal probe introduced through a gastrotomy up into the lower esophageal segment while another bougie was pushed downward from above. He assumed that the esophagus was blocked by a membrane but his procedure failed and autopsy revealed that the esophagus was found "to terminate above and below in a blind rounded ends an inch and a half apart and there was no cord or connection between the parts".

In 1899, Hoffman performed the first permanent gastrostomy in an infant with EA. In 1913, H. M. Richter of Chicago described two infants with EA-TEF, on whom he operated without success. Despite his results, Richter was optimistic about the eventual success of primary esophageal repair and stated, "I do not wish to dismiss the idea of immediate union of the two ends of the esophagus." Despite Richter's optimism, primary esophageal anastomosis and a myriad of other operations continued to fail; mortality rate for EA remained 100%.

The first published report of a case of EA-TEF treated by fistula ligation and primary esophageal anastomosis was presented by Robert Shaw of Dallas, Texas. Shaw performed the operation on September 25, 1938, and reported it in December 1939. In his report, Shaw referred to a personal communication from Paul C. Samson, who had also performed "an operation very similar" to the one Shaw described. Samson's patient died 12 hours after the operation. Shaw's patient died

on the 12th day after the operation, apparently as a result of transfusion reaction. Shaw performed the operation without knowing that Thomas Lanman in Boston has performed four primary esophageal repairs for EA-TEF between January 2, 1936, and July 27, 1937, with the use of extrapleural approach. In November 1940, Lanman reported these 4 cases, 1 other case of primary esophageal repair, and 27 other cases of EA seen in Boston Children's Hospital between September 1929 and February 1940. All 32 patients in the series died. However, the Lanman's fourth patient treated by fistula ligation and primary esophageal repair lived 9 days and at autopsy was found to have died of overhydration, not of mediastinitis or pneumonia. Remarkably Lanman summarized: "in spite of the fatal outcome in all the 30 operative cases, it is felt that considerable progress along rational lines is being made. The successful operative treatment of a patient with this anomaly is only a question of time."

According to Humphreys and Ferrer,[3] the first survivor of congenital atresia of the esophagus without TEF was a boy born in New York on February 16, 1935. The patient was initially treated by gastrostomy alone, with his "first thoracic operation performed in 1946." The first survivors of EA-TEF were a boy born in Minnesota on November 26, 1939, and a girl born in Massachusetts on the next day. After many failures in caring for infants with EA, N. Logan Leven of the University of Minnesota performed a gastrostomy on a 2500-g male infant with EA-TEF on November 29, 1939. After a failed attempt to close the TEF bronchoscopically with a coagulation electrode, extrapleural ligation of the fistula, was accomplished on January 5, 1940. When the infant's weight increased to 4630 g, a cervical esophagostomy was performed on March 27, 1940. The child thrived, and Leven proposed that a subsequent antethoracic esophagoplasty be performed to re-establish continuity of the gastrointestinal tract. In Boston, William Ladd