CONSIDERATIONS UPON A 29 CASES WITH ESOPHAGEAL ATRESIA

ES Boia[1], A Mittal[1]
[1]University of Medicine and Pharmacy "Victor Babes" Timisoara

Abstract

Esophageal atresia with or without tracheoesophageal fistula is a surgical emergency, presenting during first moments of extrauterine life. It is frequent associated with other congenital anomalies and it is usually complicated due to aspiration of gastric contents leading to pneumonia and respiratory distress. Ligation of fistula and reconstructive anastomosis of the ends of esophagus is mainstay of treatment.

Anesthesia and skill of anesthesiologist during pre, intra and post operative periods play an important role in successful treatment and survival of neonate.

Key words: esophageal atresia, tracheoesophageal fistula, reconstructive anastomosis

Introduction

Esophageal atresia (EA) is a congenital disorder when proximal and distal portion of esophagus do not communicate. The proximal blind pouch has thick musculature and bigger diameter, while the distal portion has thin musculature.

Tracheoesophageal fistula (TEF) is a congenital disorder, with abnormal connection between anterior esophagus and posterior membranous trachea.

These two congenital malformations can occur as separate entities but commonly they occur together and the five different combinations are:

1. EA with distal TEF
2. EA with proximal TEF
3. EA with proximal and distal TEF
4. Isolated EA (without TEF)
5. Isolated TEF (without EA)

Aims of study:

To evaluate:

- the incidence of EA/TEF, in a specific period, in the Department of Pediatrics Surgery and Orthopedics, Timisoara, Romania
- the distribution of EA/TEF in Western Romania
- the frequency of different types of EA/TEF
- the common used investigation for diagnosis of EA/TEF in our hospital
- the common used treatment for EA/TEF in our hospital
- the frequency of associated malformations and complications

Material and method

The study being done on 29 cases of esophageal atresia and tracheoesophgeal fistula during 1995- 2004, surgically resolved at Clinical Emergency Hospital for Children "Louis Turcanu" Timisoara, Department of Pediatrics Surgery and Orthopedics.

This study is an example of retrospective study, conducted with the help of:

- observation files
- operation protocols

The following parameters for every esophageal atresia - tracheoesophgeal fistula case where studied:

1. Clinical findings for each case of EA-TEF
2. Time distribution of EA-TEF - during years 1995-2004
3. Place distribution of EA-TEF - according to localities – either rural or urban
4. Person distribution of EA-TEF- according to sex
5. Age at presentation of EA-TEF
6. Frequency of EA-TEF - according to type of EA-TEF
7. Associated anomalies
8. Diagnosis
9. Management
10. Prognosis and complications

Results

1. Clinical findings.

a. Pregnancy history

Course

Most of pregnancies followed normal course and very few presented with complications. E.g. toxemia

Maternal age

In our study maternal age is <30 yrs in 70% of cases, while text says that frequency of EA-TEF increases with increase in maternal age (>30 yrs).

Polihydroamnios

Our study shows only 7% of pregnancies had having history of polihydroamnios. These were from cities, where follow up facilities are generally good. We know from experience that TEF presence decrease the amount of amniotic fluid.

Mode of Delivery

Most pregnancies had spontaneous or natural mode of delivery, very few needed cesarean section.

b. Prematurity

As we know EA/TEF is commonly associated with premature birth because of polihydroamnios.

Grade

Every case has demonstrated some degree of prematurity, according to APGAR SCORE, most common being prematurity grade 2.

Birth Weight

About 25% of cases weighed less than 2500 gm at birth.

c. General physical examination

Cutaneous Tissue

Most cases presented with generalized cyanosis, especially when crying and with attempted feeding, also physiological icterus was common, depending upon stage of prematurity.

Sub-cutaneous Tissue

Almost every case had some deficiency in subcutaneous adipose tissue.

Muscular Tissue

Every case presented with hypotonia, but about 4% cases presented opisthotonia because of associated nervous tissue disorder.

Lymphatic Tissue

No lymphadenopathy detected.

d. Respiratory system

Inspection

Most of cases presented with use of accessory muscles of respiration, cyanosis, and intercostals retractions with inspiration and choking especially with attempted feeding.

Palpation

There were no palpable deformities except those with vertebral column defect. E.g. scoliosis.

Percussion

Most of cases had dullness on percussion because of high association of aspiration bronchitis and pneumonia.

Auscultation

Most of cases presented with abnormal respiratory sounds bilaterally, rales or crepitations depending upon the underlying pathology.

e. Cardiovascular system

Inspection

Generally normal except when there was associated congenital cardiac malformation, leads to cyanosis.

Palpation

Apex beat normally palpable and peripheral pulses was normal.

Percussion

Almost every case had normal cardiac limits.

Auscultation

Most cases presented with normal cardiac sounds except in congenital cardiac malformation, there was holosystolic murmur.

f. Digestive system

Inspection

- Every case had been presented with excessive salivation and drooling.
- With attempted feeding there were episodes of gagging, choking and cyanosis.
- Most of cases had distended abdomen.
- 14% cases had anorectal atresia with no visible anal aperture.
- Most of cases had intestinal transit with history of passed meconium.
- 4% cases presented with anal ectopia

Palpation

Most of cases had soft abdomen on palpation with no hepatosplenomegaly.

Percussion

Most of cases had increased tympani on percussion because of distension, with normal hepatic limits.

Auscultation

Most of cases had normal borborygmy because history of present intestinal transit.

Those with absent intestinal transit had no audible borborygmy sounds.

g. Genito-urinary system

Most cases presented with normal micturation and normal aspect of urine, except in 4% cases with congenital renal malformations and also with normal genital system on inspection and palpation except 4% cases with bilateral ectopic testes.

h. Nervous system

Almost every case had normal nervous system examination with 4% cases of abnormal examination. E.g. absent spinal reflexes.

2. Time distribution of EA-TEF - during years 1995-2004 (fig. 1) was: 3 cases in 1995, 4 cases in 1996 and same number in 1997, 2 cases in 1998 and also 2 in 1999, 3 cases in 2000 and 3 in 2001, 5 cases in 2002, only 1 case in 2003 and 2 cases in 2004.

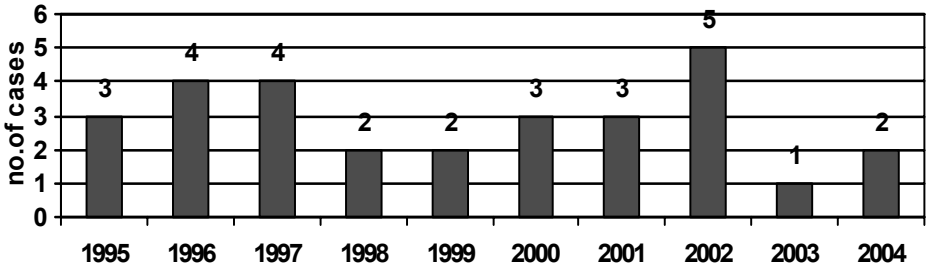

Fig. 1. - Time distribution of cases between 1995 – 2004.

3. Place distribution of EA-TEF cases according to localities indicated both the rural and urban areas had almost equal number of cases (fig. 2) , although urban areas had slightly more cases:

- rural area - 48% of cases
- urban area - 52% of cases

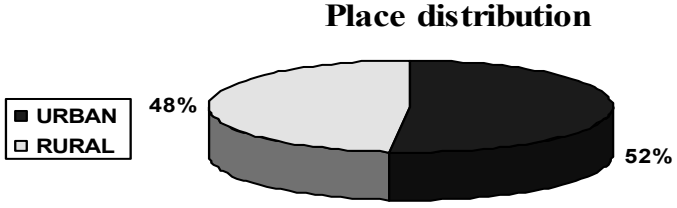

Fig. 2. – Place distribution of EA-TEF cases.

4. Distribution of EA-TEF cases according to sex in our study shows male (18) to female (11) ratio of 1.63.

5. Age at presentation of EA-TEF: according to time between delivery and presentation in our department, most of the cases (21) was hospitalized in the first 36 hours of life (fig. 3).

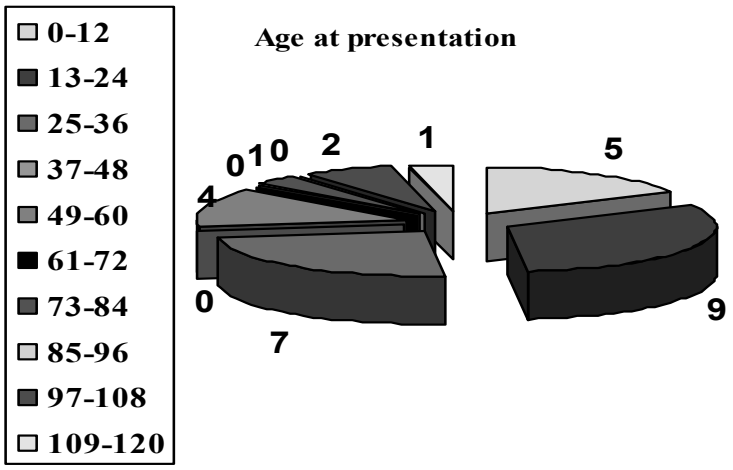

Fig. 3. – Distribution of cases according to age of presentation.

6. Frequency of type of EA-TEF in our study shows type 3 being most common:

- Type 3 (86%) 3A (20%) 3B (80%)
- Type 4 (7%)
- Type 1 (3%)
- Type 2 (3%)
- Type 5 (1%)

7. Associated anomalies – the presence of associated anomalies shows:

- 28% of cases had VACTERL association (fig. 4), most common being anal and cardiac. In contrast, studies conducted outside shows 50% VACTERL association.
- increased risk of VACTERL association with birth weight less than 2500 gms, which is consistent with outside studies.

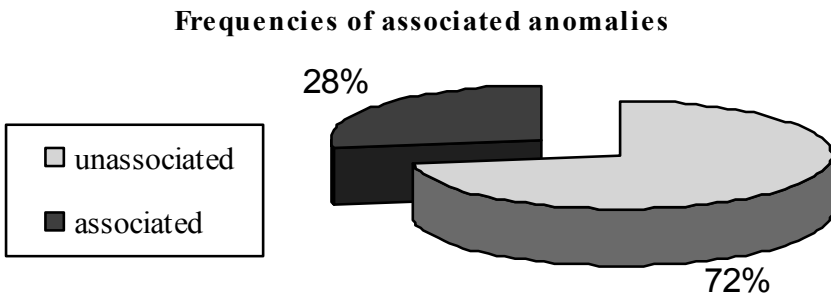

Fig. 4. – Frequencies of associated anomalies.

8. Diagnosis

As we know in new born prematurity, polihydroamnios and any demonstrable element of VACTERL, warrants a more thorough search for EA-TEF. Also, prenatal diagnosis of EA-TEF by maternal sonography is possible but requires highly experienced hand.

Postnatal nasogastric intubations followed by simple whole abdomen and chest radiography and if needed contrast studies are mainstay of diagnostic investigation for EA-TEF. Contrast studies helps to locate exactly the position of TEF, type of EA-TEF and to measure the distance between two ends of esophagus.

In our hospital, mainstay of postnatal diagnosis of EA-TEF is radiogarphy with nasogastric tube (NGT), with 41% of cases being diagnosed with this technique (table 1). This is consistent with diagnostic approach outside Romania.

Radiography with contrast medium being done on 38% of cases is not a preferred approach because it increases the chances of chemical tracheobronchitis and pneumonia, adding to further considerable morbidity.

Table 1. Radiological diagnosis in EA-TEF

Method of radiography	No. of cases
Radiography with NGT	12
Radiography with Contrast Medium	11
Radiography with NGT and Contrast Medium	06
TOTAL	29

9. Management

Immediate surgery is the only treatment of EA-TEF.

Approach adopted in our hospital to treat EA-TEF, is consistent with the approach adopted outside Romania and depends on the length of gap between proximal and distal ends of esophagus. Our study shows long gap situation 28% and small gap situation 72%, small gap situations being treated with single surgery approach and long gap situation being treated with stepped surgical approach.

a) *Small Gap Situation:*

- single surgery is required.
- 4th intercostals right space thoracotomy and extrapleural approach.
- azygos vein is divided.
- ligation of TEF.
- esophagus is mobilized and primary anastomosis is usually achieved.

b) *Long Gap Situation*

- stepped surgery is required.
- 1st ligation of TEF, cervical esophagostomy and gastrostomy.
- 2nd surgery is performed 2-3 weeks onwards includes esophagoplasty either with stomach or colon.

10. Prognosis and complications

Our study shows high Case Fatality Rate and low Survival Rate of 69% and 31% respectively.

A) *Case Fatality Rate* (CFR) = Total number of deaths due to particular disease / Total number of cases due to same disease x 100 = 69%.

B) *Survival Rate* = Total number of patients alive after five years / Total number of patients diagnosed or treated x 100 = 31%.

This high CFR is due to immediate postoperative life threatening complications. Most common being:

- cardiopulmonary failure
- acute septic shock- enterobacter species
- acute bronchopneumonia- staphylococcus aureus
- out of 31% who survived, very few have presented with postoperative stricture at site of primary repair of esophagus and recurrence of TEF.

Conclusions

After the study, we have reached some conclusions, which are as follow:

- there is approximate equal, rural and urban distribution of this condition.
- male to female ratio being 1.63, with male sex at increased risk of having this condition.
- 73% cases presented to us within first 36 hours of life.
- type 3 of EA is the most common form of the condition with 86% of total cases, rest is as follows type 4(7%), type 2(3%), type 1(3%), and type 5(1%).
- 28% of cases had been associated with VACTERL anomalies, out of which most common were anal and cardiac anomalies.
- 59% of cases were diagnosed with the help of contrast radiography, while rest 41% with simple whole thoracoabdominal radiography with NGT.
- choice of therapeutic approach depends upon length between two ends of oesophagus:
  - small gap situation- single surgery
  - long gap situation- stepped surgery
- now, in days of highly sophisticated aseptic surgical techniques, we still have severe complications with fatal evolution in approximative 70% of cases in our hospital, most common postoperative complications are:
  - cardiopulmonary failure
  - acute septic shock- due to enterobacter species
  - acute bronchopneumonia- due to staphylococcus aureus
  - hepatorenal failure
  - disseminated intravascular coagulation
- postoperative complications are main cause of mortality rather than EA-TEF itself

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Correspondence to:

Eugen Sorin Boia,
Gospodarilor Street, No. 42,
Timisoara 300778,
Romania
E-mail: eemboia@rdslink.ro