Pediatr Surg Int (2007) 23:767–771
DOI 10.1007/s00383-007-1964-0

ORIGINAL ARTICLE

# Prognosis of congenital tracheoesophageal fistula with esophageal atresia on the basis of gap length

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Accepted: 3 May 2007 / Published online: 20 June 2007


**Abstract** Congenital tracheoesophageal fistula (TEF) with esophageal atresia (EA) is not an uncommon disease of newborns. Several classifications have been advocated for predicting the outcomes of these patients but all are physiological and concentrated on associated medical condition that influences survival. We emphasize a new classification on the basis of gap between two esophageal pouches to define the magnitude of surgical problems in the primary repair and correlate them with the outcomes in terms of anastomotic leak, esophageal stricture and mortality, keeping other prognostic factors constant. A total of 50 cases of congenital TEF with EA were included and all underwent primary esophageal anastomosis after the ligation of TEF. The gap between the two pouches was meticulously measured intraoperatively using a vernier caliper before the ligation of TEF, and patients were divided into four groups on the basis of gap length. Group A, where gap length was >3.5 cm (ultralong), group B where gap length was 2.1–3.5 cm (long gap), group C where gap length was >1 cm but not more than 2 cm (intermediate group) and group D, where the gap between two esophageal pouches was 1 cm or less (short gap). The incidence of anastomotic leak was 80, 50, 28, 10.5% and the incidence of esophageal stricture was 100, 75, 22.5, 19% after successful primary repair, respectively, in groups A, B, C and D. The mortality was highest in group A (80%) followed by group B (50%) and 22% in group C and least 15.6% in group D. The incidences of esophageal leak and mortality were found to be statistically significant. This classification which is based on easily measurable criteria provides a useful method to predict morbidity, long-term outcome and mortality of EA with TEF surgery.

**Keywords** Esophageal atresia · Gap length · Prognostic criteria

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## Introduction

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is one of the challenging problems in pediatric surgery. Waterston's risk classification [1], traditionally used to identify neonates at risk for poor outcome, has become obsolete in the developed world [2–4]. In developing countries such as India, most of the babies of TEF with EA present late usually have pneumonitis because poor referrals and are of low birth weight. Because of these factors, although various new prognostic classifications are proposed, none is properly applicable for predicting the outcome. We present another classification where the gap between the two esophageal pouches is considered to be an important independent risk factor in the cases of TEF with EA. Measuring the gap length is more practical, and gives the proper prediction of short-term as well as long-term outcomes. In this study, the results of the 50 consecutive cases of TEF with EA, which were classified on the basis of gap between two esophageal pouches in four groups, keeping all other risk factors in consideration, were analysed in terms of esophageal leak, esophageal strictures and mortality. Other factors such as birth weight, age at presentation, pneumonitis and associated anomalies affecting morbidity and mortality in cases of TEF with EA; were comparable in all groups.

## Materials and methods

A total of 50 cases were studied during the period of 2 years from January 1999 to December 2001. We have included the patients of congenital TEF with EA only. The cases of pure EA were excluded from our study. Parameters significantly affecting the outcome were identified. These included gestational age, birth weight, age at presentation, and severity of pneumonitis along with associated anomalies. All these cases were subjected to single-stage, single-layer primary esophageal anastomosis using Vicryl 5.0 suture after ligation of fistula. The gap between the two esophageal pouches was meticulously measured with the help of a vernier caliper intraoperatively before ligating the fistula region. The gap length was measured in centimeter, and no attempts were made to measure the length preoperatively. The cases were divided into four groups according to gap. Group A (Ultralong gap) [5], i.e. the gap between two esophageal pouches was more than 3.5 cm, group B (long gap, i.e. gap was >2 to <3.5 cm), group C (intermediate, i.e. gap was >1 to <2 cm) and group D (short gap, i.e. <1 cm). All four groups were comparable in other parameters, such as the age of presentation, birth weight, pneumonitis and other associated anomalies. An outcome of these cases was analysed in terms of anastomotic leak, esophageal stricture and mortality.

Statistical analysis was done by using Fisher's exact test, Chi-square test and Chi-square tend.

## Results

There were 28 males and 22 females (M:F = 1.3:1), and their distribution was similar in different groups (Table 1). The average birth weight was 2,150 g (range 1,100–3,100). Th mean age of presentation was 58 h (range 10 h–9 days). Severe pneumonitis was present in 42% of the cases, 2, 4, 6 and 8 in ultralong, long, intermediate and short gap groups, respectively (Table 1); the difference in the incidence of pneumonitis in different groups was statistically insignificant. The associated malformation was 34% in present series, and was comparable in different groups (Table 2), but the incidence of associated anomalies in different groups was statistically insignificant. The number of the patients in groups A and B was less because we have selected the 50 consecutive cases of EA with TEF of which only five had ultralong gap and eight had long gap rather than selecting an equal number of patients in each group.

**Table 1** Distribution of patients on the basis of sex and incidence of pneumonitis in different groups

| | Ultralong group A | Long gap group B | Intermediate gap group C | Short gap group D |
|---|---|---|---|---|
| Male | 3 | 5 | 11 | 9 |
| Female | 2 | 3 | 7 | 10 |
| Pneumonitis present | 2/5 | 4/8 | 6/18 | 8/19 |

On statistical analysis the difference was not found to be statistically significant

In current series, overall anastomotic leak was observed in 30% (15/50) patients. Anastomotic leak was highest (80%) in group A and least (10.5%) in group D (Table 3). Esophageal stricture was observed in one case of group A and three cases each in groups B, C and D (Table 4). Mortality in our series was 30%, and was highest in group A (80%) followed by group B and was least (15.6%) in group D (Table 4). Of the 15 mortalities, 4 cases died of cardiac anomaly and 3 cases because of severe pneumonitis, rest due to septicemia following anastomotic leak. All the mortalities in group A were due to septicemia following anastomotic leak, whereas none of the patients in group D died of septicemia. The average hospital stay was 28 days (range 20–38 days), 20 days (range 16–27 days), 13 days (range 9–17 days) and 10 days (7–16 days), respectively, for groups A, B, C and D.

## Discussion

Waterston's risk classification [1] is important predictor of outcome in EA with or without TEF used three factors such as birth weight, severity of pneumonia and severity of associated anomalies. Early diagnosis, improved surgical technique, neonatal anesthesia, sophisticated ventilator support, advanced intensive care management and early treatment of associated congenital anomalies have nullified the effect of Waterston's risk factors to a great extent. Improved survival rates were noted irrespective of Waterston's categories [2]. In a historical review, Deurloo et al. [3] concluded that although the patients with EA treated nowadays are born earlier weigh less and have more associated anomalies than those treated 50 years ago; still the mortality rate is much lower due to early diagnosis, better supportive care and improved surgical techniques. In fact, Waterston's classifications have lost their prognostic usefulness and have become outdated in the western world [2–4]. In developing countries such as India, most of the babies are of low birth weight according to WHO standard, and most of the deliveries are conducted by untrained personnel, leading to a delay in the diagnosis of such cases. Most of the babies of TEF with EA presented at our centre were of low birth weight, had pneumonitis and with features of sepsis; this is the reason why we have chosen gap length as a new method for predicting prognosis of such patients.

**Table 2** Distribution of other associated anomalies in different groups

| | Ultralong | Long gap | Intermediate gap | Short gap | |
|---|---|---|---|---|---|
| Cardiovascular anomaly | 1 | 1 | 3 | 2 | 7 |
| Gastrointestinal anomaly | None | 1 | 2 | 1 | 4 |
| Skeletal anomaly | 1 | 1 | 1 | 3 | 6 |
| Total | 2/5 (40%) | 3/8 (37.5%) | 6/18 (33%) | 6/19 (31%) | 17/50 (34%) |

The distribution of other associated anomalies was statistically significant in different groups

**Table 3** Distribution of the cases according to gap length and incidence of anastomotic leak in different groups

| Gap length | No. of cases | Anastomotic leak |
|---|---|---|
| Ultralong (>3.5 cm) | 5 | 4 (80%) |
| Long gap (>2 to <3.5 cm) | 8 | 4 (50%) |
| Intermediate gap (>1 to <2 cm) | 18 | 5 (28%) |
| Short gap (<1 cm) | 19 | 2 (10.5%) |
| Total ($n$ = 50) | 50 | 15 (30%) |

On statistical analysis using chi-square tend for leak

| Groups | Odd ratio |
|---|---|
| D | 1 |
| C | 3.27 |
| B | 8.5 |
| A | 34 |

Value of $P$ = 0.002

**Table 4** Incidence of esophageal stricture and mortality in different groups

| Gap length | Mortality | Stricture |
|---|---|---|
| Ultralong (>3.5 cm) | 4 (80%) | 1 (100%) |
| Long gap (>2 to <3.5 cm) | 4 (50%) | 3 (75%) |
| Intermediate gap (>1 to <2 cm) | 4 (22%) | 3 (22.5%) |
| Short gap (<1 cm) | 3 (15.6%) | 3 (19%) |
| Total ($n$ = 50) | 15 (30%) | 10 (29%) |

On statistical analysis using Chi square tend for mortality

| Groups | Odd ratio |
|---|---|
| D | 1 |
| C | 1.5 |
| B | 5.33 |
| A | 21.33 |

Value of $P$ = 0.0025

The statistical result for esophageal stricture was not found to be statistically significant

In recent years, new prognostic classifications have been proposed. Montreal prognostic classification proposed by Poenaru et al. [4] suggested that only severe pulmonary dysfunction with preoperative ventilator dependence and severe associated anomalies have prognostic influence. This classification is also not suitable for developing countries because most of the patients presenting at tertiary centre had severe pneumonitis because of delay in diagnosis, poor transport and inadequate care.

Spitz et al. [6] observed that the birth weight of less than 1,500 g and the presence of major cardiac disease were the important predictor of outcome. Gidaro et al. [7] concluded that survival is related more to the presence of severe multiple associated malformations rather than broncho-pneumonic complications or birth weight. Vagyu et al. [8] suggested that respiratory distress syndrome and pneumonia are still essential preoperative risk factors for EA. Recognizable chromosomal abnormalities such as Down's syndrome and others have high mortality despite the proper treatment [9]. The presence of right-sided aortic arch adversely affects outcome by posing technical operative difficulties [10]. All these classifications stated above are also not suitable for the developing countries because most of the patients are of low birth weight, have pneumonitis, present late at tertiary centre and have higher incidence of other associated congenital anomalies. In this study, all the factors, which are considered as poor prognostic factors for EF with EA, are almost comparable in different groups.

Although the significant improvement in management and outcome of EA either with TEF or without TEF has been made in India during the past two decades, the survival rate of around 50% is still unsatisfactory. Two important reasons for unsatisfactory results are (i) delay in diagnosis leading to aspiration pneumonitis and septic complications and (ii) unsupervised transportation from long distances leading to hypothermia [11–14]. Most of the patients at our centre were of low birth weight, had pneumonitis, features of sepsis, had previous attempt of feeding and higher incidence of associated anomalies. These patients would be placed in a high-risk group according to any of the classifications stated above so we have to predict outcomes among these patients for which we used the gap length as a predictor for outcome in these cases. This classification is based on easily measurable criteria, provides a useful method to predict morbidity, long-term outcome. The gap between the two esophageal

pouches is an important predictor of survival [15] in the cases of TEF with EA; we also observed the same.

Age at presentation, not an important factor in the western world, had inverse relationship with outcome in many Indian series [11–14]. Increased age at presentation mostly due to delay in diagnosis and sometimes due to delay in transportation to a pediatric surgery centre is a common occurrence in Indian subcontinent. Similarly, prematurity which does not always lead to low birth weight does not find a place in prognostic classifications in common use, although it is a major risk factor in most of the reported Indian series.

Esophageal anastomosis leak is one of the common and most dangerous complications of surgery in TEF with EA. Anastomotic leakage into the mediastinum occurs in 14–21% of the children who have undergone a surgical EA repair. Leaks result from the small, friable lower segment, ischemia of the esophageal ends, excess anastomotic tension [16, 17], sepsis, poor suturing techniques, type of suture [18, 19], excessive mobilization of distal pouch [20] and increased gap length [16, 17, 19]. These studies show that gap length can predict the chances of anastomotic leak because in cases of long gap anastomotic site is under tension, and excessive mobilization of both pouches is required for proper anastomosis.

Anastomotic stricture is the most common complication following the surgical repair of EA and observed in 30–40% after successful repair [21]. A number of predisposing factors have been implicated in the pathogenesis of esophageal anastomotic stricture after successful primary repair of EA such as a two-layer anastomosis [22], increased gap length [16, 17], anastomotic tension [23], type of suture [24, 25], anastomotic leak [16, 17, 24] and GER [21, 24]. Anastomotic strictures after esophageal leaks vary from 70 to 100% in most of the reported series [24, 26, 27]. This suggests that gap length can also predict the chances of esophageal stricture after successful primary repair of esophagus because anastomotic site is under tension and the chances of anastomotic leak are more in the cases with long gap. Both these factors are associated with the increase incidence of esophageal stricture.

On the basis of these findings, we have developed a new classification for predicting the prognosis of TEF with EA based on the gap between the two esophageal pouches which is based on easily measurable criteria, provides a useful method to predict morbidity and long-term outcome. The gap length of 1 cm or less is considered as a short gap [11]. A gap length of more than 2 cm is considered arbitrary as a long gap [16]. A few studies consider a gap length of 3 cm or more as a long gap [28]. In another study [11], a gap length of more than 1 cm and less than 3 cm was considered as an intermediate group. When the gap between two pouches was more than 3.5 cm, it is called as ultralong gap [7, 8]. Taking all these studies in consideration we divided our patients into four groups as group A, ultralong gap (gap length > 3.5 cm), group B, long gap (gap length > 2 cm to <3.5 cm), group C, intermediate group (gap length > 1 to ≤2 cm), and group D as a short gap (gap length < 1 cm). Gap length is an important factor in determining the outcome of patients in the cases of TEF with EA, is demonstrated by a few other studies. Hagberg et al. in 1986 [10] reported a group of EA with a 3 cm gap and a complication rate of 100%.

In this study, anastomotic leak was observed in 30% (15/50) of the cases. The incidence of anastomotic leak was highest (80%) in group A, followed by group B (50%) (4/8) and was only 28% (5/18) and 10.5% (2/19) in groups C and D, respectively. On statistical analysis, the odd ratio of groups was 1, 3.27, 8.5, and 34 for groups D, C, B and A, respectively, and $P = 0.002$, which shows if anastomotic leak is 1 in group D, the chances will be 3.27, 8.5 and 34 times in groups C, B and A, respectively. These findings indicate that the chances of anastomotic leak are more in patients with long gap, suggesting that gap length can predict anastomotic leak.

An incidence of esophageal stricture was 35% (12/35) in the present series. The incidence was 100% in group A and 75% in group B where as it was only 22.5 and 19% in groups C and D, respectively. It is well known that increased anastomotic tension and esophageal leak increases the chances of esophageal stricture and so the measurement of the gap can predict the chances of esophageal strictures after successful primary esophageal repair and hence the long-term outcome. But in the present study, these values were not found to be statistically significant.

In the present series, we are not able to monitor gastroesophageal reflux because of lack of adequate measures, although the reported incidence of GER is 40–70% [23, 29]. The main reasons for GER are the shortage of intra-abdominal esophagus, stretch at anastomosis site, disturbance of esophageal motor function, manipulation during surgery, and esophageal motility problems [29]. In primary esophageal anastomosis of long gap EA, there is a considerable tension on anastomotic site as well as there is an extensive mobilization of distal esophagus, which can lead to the increase incidence of GER in a long term. Hence, gap length can also predict the chances of GER to some extent.

The mortality in the present study was 30% (15/50). Mortality was highest in group A (80%) followed by group B (50%), and was 22 and 15.6% in groups C and D, respectively. Of the 15 mortalities, 4 died because of associated congenital heart anomalies, 3 because of severe pneumonitis and 8 due to septicemia following esophageal leak, which accounted for almost 53% of the total mortalities. On statistical analysis, odd ratio for groups D, B, C

and A was 1, 1.5, 5.33 and 21.33, respectively, and $P = 0.0025$. The mortality was more in patients having long gap length; hence, the mortality can also be predicted by simply measuring the gap length. In the above discussion, it was observed that the gap between the two esophageal pouches can predict the chances of esophageal leak, the development of esophageal strictures, the chances of GER and mortality as well.

Anastomotic leak was maximum in group A (80%) and minimum in group D (10.5%), esophageal stricture was maximum in group A (100%) and minimum in group D (19%), and mortality was highest in group A (80%), most of them due to septicemia following anastomotic leak whereas mortality was minimum in group D (15%), none of them due to anastomotic complication. In this study, it was observed that although risk factors (i.e. low birth weight, prematurity, age at presentation, severe pneumonitis and other associated congenital abnormalities) used in the previous classifications predict the outcome of such patients, they are important factors for western countries but not for the developing countries such as India. Most of the patients in the present series were of low birth weight, had pneumonitis, sepsis, previous attempt of feeding, present late in hospital and a high incidence of associated anomalies. We think that gap between the two esophageal pouches can predict the short-term as well as long-term outcomes.

## Conclusion

This classification which is based on easily measurable criteria provides a useful method to predict morbidity, long-term outcome and mortality of EA with TEF surgery.

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