

## Clinical Outcomes of Esophageal Atresia: Comparison Between the Waterston and the Spitz Classifications

Rangsan Niramis,<sup>1,2,MD</sup>, Pitiporn Tangkhabuanbut,<sup>1,2,MD</sup>, Maitree Anuntkosol,<sup>1,2,MD</sup>, Veera Buranakitjaroen,<sup>1,2,MD</sup>, Achariya Tongsin,<sup>1,2,MD</sup>, Varaporn Mahatharadol,<sup>1,2,MD</sup>

### Abstract

**Introduction:** Preoperative prognostic predictors are important for surgeons and parents to estimate the survival of patients with esophageal atresia (EA). The aim of this study was to update the clinical outcomes of EA treatment by comparing between the Waterston and the Spitz classification. **Materials and Methods:** Medical records of the patients with EA treated at Queen Sirikit National Institute of Child Health from 2003 to 2010 were reviewed. All of the patients were categorised into 3 groups of the Waterston and 3 groups of the Spitz risk factor criteria for comparing of the differences in each group and each classification. **Results:** One hundred and thirty-two patients (81 males and 61 females) were treated for EA during the study period. Applying the Waterston classification, survival rate was 100% in group A, 91.5% in group B and 48.8% in group C. There was no statistical difference between the survival rate in group A and group B ( $P = 0.119$ ) but significant difference between group B and group C ( $P = 0.000$ ). Using the Spitz classification, survival rate was 97.4% in group I, 64.4% in group II and 27.3% in group III. There was obviously statistical difference of the survival rate between each group (group I vs group II,  $P = 0.000$ ; group II vs group III,  $P = 0.041$ ). **Conclusion:** Comparing with the prognostic predictors, the Spitz classification was more valid than the Waterston criteria. The Spitz classification is suitable to use for preoperative predictor to parental counselling and comparing of treatment outcomes of EA among paediatric tertiary care centres.

Ann Acad Med Singapore 2013;42:297-300

**Key words:** Tracheoesophageal fistula, Risk factor, Prognostic predictor, Classification

### Introduction

Esophageal atresia (EA) is the most common and serious anomaly of the esophagus. The abnormality was uniformly fatal throughout the world prior to 1939. The first survivors of esophageal repair were reported by Leven<sup>1</sup> and Ladd.<sup>2</sup> Later on, the survival rate of the disease continuously improved to more than 90% in this era.<sup>3-5</sup> The good results of esophageal treatment reflect the advances in neonatal intensive care, nutritional support, improvement of anesthetic and surgical techniques. Along with these advances, concepts of the prognosis and therapeutic strategy for EA management have been changing. Some investigators have questioned in the predictive validity of the Waterston classification<sup>6</sup> which is based on risk factors including low birth weight, pneumonia and associated congenital anomaly. A new risk classification was proposed by Spitz in 1990.<sup>2</sup> Risk factors in Spitz's study include only low birth weight and

cardiovascular anomalies. Many reports have found the Spitz classification useful for planning of the treatment, parental counselling and comparing outcomes in each centre.<sup>5,7-9</sup>

We have studied the results of EA management by using the Waterston risk factor criteria since 2000 and the outcomes were not satisfactory.<sup>10</sup> We herein evaluated this entity again. The aim of this study was to review the clinical outcomes of neonates born with EA and compare between the Waterston and the Spitz classification over the last 8 years from a single tertiary centre for paediatrics in Thailand.

### Materials and Methods

Medical records of all the neonates with EA treated at Queen Sirikit National Institute of Child Health between January 2003 and December 2010 were reviewed. Data were

<sup>1</sup>Department of Surgery, Queen Sirikit National Institute of Child Health, Thailand

<sup>2</sup>College of Medicine, Rangsit University, Thailand

Address for Correspondence: Dr Rangsan Niramis, Department of Surgery, Queen Sirikit National Institute of Child Health, Bangkok, Thailand 10400.

Email: nriramis@hotmail.com

obtained on birth weight, associated anomalies, operative procedures, complications and outcomes. Pneumonia and atelectasis were counted on the basis of radiological reports. The patients were categorised into the 3 Waterston<sup>6</sup> and the 3 Spitz risk groups.<sup>11</sup> The survival rates in each group were compared by Pearson's chi-square test. *P* values lower than 0.05 were considered statistically significant. The study protocol was approved by the Institutional Reviewer Board (Document No.53-060.1).

## Results

During the study period, 132 neonates with EA were admitted to our institute. There were 81 males and 61 females, hence the male to female ratio was 1.6 : 1. Ten patients were born at Rajavithi Hospital (previously known as 'Women's Hospital'), during which time there were 57,346 live births.<sup>12</sup> Therefore, the incidence of EA was 1.7 : 10,000 live births at Rajavithi Hospital. The mean gestational age was 36.6 ± 8.7 weeks (range, 27 to 41 weeks), whereas the mean birth weight was 2395.5 ± 1208.4 g (range, 1100 to 3510 g). The different anatomical variations of EA are shown in Table 1.

Table 1. Anatomical Variations of Esophageal Atresia in the 132 Patients

Type of esophageal atresia	No. of cases	Percentage (%)
Esophageal atresia with distal tracheoesophageal fistula	117	88.6
Esophageal atresia without tracheoesophageal fistula	13	9.8
Esophageal atresia with proximal and distal tracheoesophageal fistula	1	0.8
Isolated tracheoesophageal fistula	1	0.8

Table 2. Associated Anomalies of Esophageal Atresia (in 86 out of 132 Patients)

Congenital anomaly	No. of cases	Percentage (%)
Cardiovascular	63	47.7
Anorectal	23	17.4
Vertebral	14	10.6
Genitourinary	13	9.8
Limb	13	9.8
Duodenal atresia	8	6.1
Trisomy syndrome	7	5.3
Others	43	32.6

Associated anomalies were identified in 86 patients (65.2%) with a total of 182 various malformations. Sixty-three cases (47.7%) had cardiac anomalies with 49 major types, either cyanotic or non-cyanotic congenital heart diseases that require palliative or corrective surgery, including patent ductus arteriosus with surgical treatment. Seven patients had a trisomy syndrome, 4 with trisomy-21 and 3 with trisomy-18. Six patients presented with the VACTERL's association. Anorectal, vertebral and genitourinary malformations were noted in 23 (17.4%), 14 (10.6%) and 13 (9.8%), respectively (Table 2).

Among the total 118 cases of EA with tracheoesophageal (TE) fistula, the ratio of primary to staged esophageal anastomosis was 23 (19.5%) : 95 (80.5%) or 1 : 4. Seven patients died before definitive esophageal anastomosis. Thoracotomy with extrapleural approach was more popular than transpleural technique (65% : 35%). Of the 13 cases with isolated EA, 11 cases were definitely treated by gastric transposition, whereas 2 cases routinely underwent transthoracic esophageal anastomosis.

The patients were customarily grouped according to the Waterston classification.<sup>6</sup> Forty-two patients were allocated in group A, 47 in group B and 43 in group C. The survival rate in each group is shown in Table 3. Survival rates of the patients in group A and group B were not statistically different (100% vs 91.5%, *P* = 0.119), but there was a significant difference between group B and group C (91.5% vs 48.8%, *P* = 0.000).

Table 3. Survival Rate of the 132 Patients in Relation to Risk Classification

Classification	Definition	Total	Survival rate
Waterston			
Group A	Birth weight over 2500g and well	42	42/42 (100%)
Group B	Birth weight 1800 g to 2500 g and well or over 2500 g with moderate pneumonia and congenital anomaly	47	43/47 (91.5%)
Group C	Birth weight under 1800 g and well or 1800 g to 2500 g with severe pneumonia and severe congenital anomaly	43	21/43 (48.8%)
Spitz			
Group I	Birth weight over 1500 g with no major cardiac anomaly	76	74/76 (97.4%)
Group II	Birth weight less than 1500 g or major cardiac anomaly	45	29/45 (64.4%)
Group III	Birth weight less than 1500 g and major cardiac anomaly	11	3/11 (27.3%)

Regarding the Spitz classification,<sup>11</sup> 76, 44 and 11 patients were categorised in group I, group II and group III, respectively. There were significant differences in the survival rates between group I and group II (97.4% vs 64.4%,  $P = 0.000$ ) and group II and group III (64.4% vs 27.3%,  $P = 0.041$ ).

Complications of the patients are shown in Table 4. Causes of death in the 26 patients include major cardiac anomalies with congestive heart failure in 12 cases (46.1%), septicaemia in 10 cases (38.5%) and severe pneumonia in 4 cases (15.4%). Of the total 132 patients, 63 cases with cardiac anomalies had the survival rate of only 66.7%, compared with the 92.8% survival rate of the 69 cases without cardiac defects ( $P = 0.001$ ).

Table 4. Complications and Major Causes of Death

Complications	No. of cases	Percentage (%)
Severe pneumonia with atelectasis	25	18.9
Sepsis	24	18.2
Anastomotic leak	24	18.2
Congestive heart failure	20	15.2
Gastroesophageal reflux	13	9.8
Anastomotic stricture	11	8.3
Wound infection	7	3.8
Recurrent tracheoesophageal fistula	3	2.8
Patient death*	26	19.7

\*Major causes of death: Congestive heart failure = 12; sepsis = 10; and severe pneumonia = 4

## Discussion

The incidence of EA at Rajavithi Hospital, Bangkok, Thailand was slightly increased from 1.2 : 10,000 live births in the previous study (2000)<sup>10</sup> to 1.7 : 10,000 live births in the present study. However, the total survival rate had dramatically improved from 40.9% to 80.3%. The good results of EA management reflect the improvement of our teams in neonatal intensive care as a result of harnessing the use of modernised paediatric techniques. This study has updated the survival rate of EA treated at our institute by comparing between the Waterston and the Spitz classifications.

Preoperative risk factor classification is important in giving surgeons and parents a realistic prognosis for children. Waterston<sup>6</sup> proposed his classification for EA in 1962 which included low birth weight, pneumonia and associated congenital anomalies as the risk factors. The survival rates from Waterston's study were 95%, 68% and 6% in group A, group B and group C, respectively. He suggested that delayed definite repair should be performed for patients in

group B and group C. The Waterston classification seemed to be valid in the past decades. Other authors, who had used this classification, obtained similar results as Waterston's report.<sup>13-17</sup> In our previous study of 105 cases with EA in 2000,<sup>10</sup> patients in group A, group B and group C with the Waterston classification had statistically significant difference (88.2% : 50% : 10%;  $P < 0.05$ ).

In the present study of 132 cases with EA, the survival rate of the patients in group A and group B, categorised by Waterston, is not statistically different, but there was significant difference between group B and group C. In contrast, there were obvious statistical differences between group I, group II and group III in the Spitz classification (Table 3). Our results of the EA treatment indicate that the Waterston classification had the prognostic validity in the past with under improvement of neonatal intensive care including pneumonia and various associated malformations. With advances in low birth weight neonatal care management leading to improve outcomes of EA treatment, validity of the Waterston classification was changed. The survival rates of group A and group B were not different and increased to 100% in both groups from many reports.<sup>8,18,19</sup> This result shows that low birth weight of less than 1800 g to 2500g, pneumonia and congenital anomalies are not strong predictors of survival in the developing period.

Spitz<sup>11</sup> proposed a new classification based on low birth weight of 1500 g and only major congenital heart defects. The survival rates from Spitz's report were 97% in group I, 59% in group II and 22% in group III. The Spitz classification more accurately reflected the prognosis of our patients in each group: 97.4%, 64.4% and 27.3%, respectively. This was borne out by the apparent statistically significant difference among the Spitz groups, and is similar to the other reports.<sup>4,5,7,20</sup>

## Conclusion

The comparative study of both the Waterston and the Spitz risk factors in our patient groups demonstrated clear differences between the 2 categories. The Waterston classification has questioned the predictive validity in the developing period. The Spitz classification is more acceptable for preoperative prognostic predictor in this era. It may be used for parental counselling and comparing outcomes among paediatric tertiary care centres.

## Acknowledgements

The authors would like to thank Dr Siraporn Sawasdivorn, Director of Queen Sirikit National Institute of Child Health, for permission to publish this paper and Miss Sasichol Kamproh for assistance in statistical analysis.

## REFERENCES

1. Leven NL. Congenital atresia of the esophagus with tracheoesophageal fistula. Report of successful extrapleural ligation of fistulous communication and cervical esophagostomy. *J Thorac Cardiovasc Surg* 1941;10:648-57.
2. Ladd WE. The surgical treatment of esophageal atresia and tracheoesophageal fistulas. *N Engl J Med* 1944;230:625-37.
3. Spitz L. Esophageal atresia: past, present, and future. *J Pediatr Surg* 1996;31:19-25.
4. Lopez PJ, Keys C, Pierro A, Drake DP, Kiely EM, Curry JJ, et al. Esophageal atresia: improved outcome in high-risk groups? *J Pediatr Surg* 2006;41:331-4.
5. Driver CP, Shankar KR, Jones MO, Lamont GA, Turnock RR, Lloyd DA, et al. Phenotypic presentation and outcome of esophageal atresia in the era of Spitz classification. *J Pediatr Surg* 2001;36:1419-21.
6. Waterston DJ, Carter RE, Aberdeen E. Esophageal atresia : tracheo-oesophageal fistula. A study of survival in 218 infants. *Lancet* 1962;1:819-22.
7. Yagyu M, Gitter H, Richter B, Booss D. Esophageal atresia in Bremen, Germany-evaluation of preoperative risk classification in esophageal atresia. *J Pediatr Surg* 2000;35:584-7.
8. Konkin DE, O' hali WA, Webber EM, Blair GK. Outcomes in esophageal atresia and tracheoesophageal fistula. *J Pediatr Surg* 2003;38:1726-9.
9. Orford J, Cass DT, Glasson MJ. Advances in the treatment of oesophageal atresia over three decades: the 1970s and the 1990s. *Pediatr Surg Int* 2004;20:402-7.
10. Anuntkosol M, Watanatittan S, Niramis R, Rattanasuwan T, Buranakitjaroen V. Esophageal atresia: six-year experience with 105 cases. *Thai J Surg* 2000;125-8.
11. Spitz L, Kiely EM, Morecroft JA, Drake DP. Esophageal atresia: at-risk groups for the 1990s. *J Pediatr Surg* 1994;29:723-5.
12. Rajavithi Hospital, Department of Medical Service, Ministry of Public Health, Bangkok, Thailand. Statistical report, 2003-2010.
13. Holder TM. Esophageal atresia and tracheoesophageal fistula. In: Ashcraft KW, Holder TM, eds. *Pediatric Esophageal Surgery*. Orlando: Grune & Stratton; 1986.
14. Myers NA, Aberdeen E. Congenital esophageal atresia and tracheoesophageal fistula. In: Ravitch MM, Welch KJ, Benson CD, Aberdeen E, Randolph JG, eds. *Pediatric Surgery*. 3rd ed. Chicago: Year Book Medical Publishers, 1973.
15. Drainer IK, Dow GR. Esophageal atresia in the Western Region of Scotland. *J R Coll Edinb* 1974;19:276-81.
16. Grosfeld JL, Ballantine TV. Esophageal atresia and tracheo-esophageal fistula : Effect of delayed thoracotomy on survival. *Surgery* 1978;84:394-402.
17. Strodel WE, Coran AG, Kish MM, Weintraub WH, Wesley JR, Sloan H. Esophageal atresia: A 41-year experience. *Arch Surg* 1979;114:523-7.
18. Okada A, Usui N, Inoue M, Kawahara H, Kubota A, Imura K, et al. Esophageal atresia in Osaka: A review of 39 years' experience. *J Pediatr Surg* 1997;32:1570-4.
19. Louhimo I, Lindahl H. Esophageal atresia : primary results of 500 consecutive treated patients. *J Pediatr Surg* 1983;18:217-29.
20. Choudhury SR, Ashcraft KW, Sharp RJ, Murphy JP, Snyder CL, Sigalet DL. Survival of patients with esophageal atresia : influence of birth weight, cardiac anomaly, and late respiratory complications. *J Pediatr Surg* 1999; 34:70-3. discussion 73-4.