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Prognostic classification for esophageal atresia at King Chulalongkorn Memorial Hospital

Monthon Mekanantawat*
Somboon Roekwibunsri* Bidhya Chandrakamol*
Dusit Viravidhaya* Soottiporn Chittmittrapap*

Mekanantawat M, Roekwibunsri S, Chandrakamol B, Viravidhaya D, Chittmittrapap S. Prognostic classification for esophageal atresia at King Chulalongkorn Memorial Hospital. Chula Med J 2000 May; 44(5): 339 - 48

Objective : *Since 1962, the Waterston classification has been used to stratify neonates with esophageal atresia and/or tracheoesophageal fistula into three groups with different chance of survival. However, the results in the treatment of esophageal atresia, have steadily improved during the past 30 years. In effort to choose the appropriate prognostic classification for our institute, we analyzed the survival of a series of cases treated in the last 10 years.*

Setting : *Pediatric Surgical Unit, Department of Surgery, Faculty of Medicine, Chulalongkorn University.*

Design : *Retrospective descriptive study*

Materials and Methods : *All infants with esophageal atresia and/or tracheoesophageal fistula were treated by the pediatric surgical unit at King Chulalongkorn Memorial Hospital during a period of 10 years (January 1989 through December 1998).*

Result : *Survival according to Waterston risk cateories was 100 % for group A, 77.7 % for group B, and 30 % for group C. Survival*

*Department of Surgery, Faculty of Medicine, Chulalongkorn University

rate in Spitz risk categories was 74.2 %, 50 %, and 0 % for group I,II, and III respectively. And rearranged Spitz criteria (at birth weight 1,800 grams) in group I, II and III were 76.6 %, 50 % and 0 %;

Conclusion : *The confidence level of prognostic data by the modified Spitz classification subjected by the author was acceptable and may be another appropriate criteria for regional use in the meantime.*

Key words : *Esophageal atresia, Prognosis.*

Reprint request : Mekanantawat M, Department of Surgery, Faculty of Medicine, Chulalongkorn University, Bangkok 10330,Thailand.

Received for publication. November 15,1999.

มณฑล เมฆอนันต์ธวัช, สมบูรณ์ ฤกษ์วิบูลย์ศรี, พิทยา จันทรกมล, ดุสิต วีระไวทยะ, สุทธิพร จิตต์มิตรภาพ. การแบ่งกลุ่มตามลักษณะการพยากรณ์โรคของผู้ป่วยหลอดอาหารตีบตันในโรงพยาบาลจุฬาลงกรณ์. จุฬาลงกรณ์เวชสาร 2543 พ.ค; 44(5): 339 - 48

วัตถุประสงค์ : *เพื่อศึกษาและปรับปรุงวิธีการจำแนกกลุ่มที่เหมาะสมในการแบ่งกลุ่มตามลักษณะการพยากรณ์โรคของผู้ป่วยหลอดอาหารตีบตันในโรงพยาบาลจุฬาลงกรณ์*

รูปแบบงานวิจัย : *การศึกษาเชิงวิเคราะห์ย้อนหลัง*

สถานที่ทำการศึกษา : *หน่วยกุมารศัลยศาสตร์ ภาควิชาศัลยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย*

ตัวอย่างและวิธีวิจัย : *ผู้ทำการศึกษาได้ทำการวิเคราะห์ข้อมูลทางคลินิกและผลการรักษาผู้ป่วยหลอดอาหารตีบตันร่วมกับการมีท่อต่อระหว่างหลอดลมกับหลอดอาหารหรือผู้ป่วยหลอดอาหารตีบตันโดยลำพัง จำนวน 37 ราย ระหว่างปี พ.ศ. 2532 – พ.ศ. 2541 ผู้ป่วยได้ถูกทำการแบ่งเป็น 3 กลุ่มความเสี่ยงตามการจำแนกกลุ่มของ Waterston classification และ Spitz classification และการแบ่งกลุ่มโดย Modified spitz classification*

ผลการศึกษา : *ผลการศึกษาพบว่าอัตรารอดของผู้ป่วยตามการแบ่งกลุ่มของ Waterston เป็น 100 % ในกลุ่ม A, 77.7 % ในกลุ่ม B และ 30 % ในกลุ่ม C ในขณะที่อัตรารอดของผู้ป่วยตามการแบ่งกลุ่มของ Spitz เป็น 74.2 % ในกลุ่ม 1,50 % ในกลุ่ม 2 และ 0 % ในกลุ่ม 3 ตามลำดับ ผู้ทำการศึกษาได้ทำการแบ่งกลุ่มผู้ป่วยใหม่ออกเป็น 3 กลุ่ม โดยดัดแปลงจาก Spitz criteria (ใช้น้ำหนักแรกเกิด 1,800 กรัม) ผลปรากฏว่าอัตรารอดของผู้ป่วยเป็น 76.6 % ในกลุ่ม 1,50 % ในกลุ่ม 2 และ 0 % ในกลุ่ม 3 ซึ่งมีผลให้อัตรารอดระหว่างกลุ่มมีความแตกต่างอย่างมีนัยสำคัญทางสถิติ*

สรุปผล : *ผลการศึกษานี้อาจนำไปสู่การกำหนดการแบ่งกลุ่มผู้ป่วยตามลักษณะการพยากรณ์โรคใหม่ที่เหมาะสมขึ้นแทนการใช้ Waterston classification เดิม เนื่องจากผลของการพัฒนาการด้านการดูแลรักษาผู้ป่วยที่ดีขึ้น ทำให้การแบ่งกลุ่มแบบเก่าไม่สอดคล้องกับความเป็นจริงในปัจจุบัน*

Since 1962 [1] the Waterston classification has been used to stratify neonates with esophageal atresia and/or tracheoesophageal fistula into three groups "with different chances of survival "based on birth weight, the presence of pneumonia, and associated congenital anomalies. This classification continues to be widely used to compare results between different centers, but many investigators have questioned its validity. This is because results in the treatment of esophageal atresia have steadily improved during the past 30 years and early diagnosis and effective suctioning of the upper pouch have virtually eliminated aspiration pneumonia as a risk factor.[2,3] With modern neonatal intensive care, very low birth weight infants are expected to survive.[4,5] A number of technical advances have also contributed to reduce the operative morbidity and mortality, such as anesthetic advancement, choice of suture material,[6] use of elective mechanical ventilation post-operatively, and early recognition and treatment of anastomotic complications.[7] Many centers have reported successful primary surgical corrections at birth.[8,9] Alternative prognostic criteria from several centers have been reported and applied to modern pediatric surgical practice in stead of the Waterston classification.[10,11] One outstanding criteria among these studies has been reported by Spitz, and is based on birth weight and major congenital heart disease.[10] We analyzed the survival of a series of esophageal atresia and/or tracheoesophageal fistula patients treated in the last 10 years in an effort to choose the most appropriate prognostic classification for our institute.

Materials and Methods

During a period of 10 years (January 1989 through December 1998) 37 infants with esophageal atresia and/or tracheoesophageal fistula were treated by the pediatric surgical unit at King Chulalongkorn Memorial Hospital; all were managed by surgical correction. These cases were reviewed according to the statistical data: sex, gestation age, birth weight, type of esophageal atresia, associated congenital anomalies, type of surgical procedure and post - operative complications. Particular attention was directed at their birth weight, the presence of pneumonia during admission and associated congenital anomalies. One of 37 infants was excluded due to incomplete data.

A major cardiac anomaly was defined as a congenital heart disease that required palliative corrective surgery later, and excluded patent ductus arteriosus due to not requiring surgical ligation. The patients were categorized into three risk groups by both Waterston classification and Spitz classification. Survival rates in each group were determined. The outcome for each group in both categories were compared with Fisher's Exact test using a 95 % confidence level for statistical significance.

Results

Characteristics of the 37 patients

Twenty five of the 37 infants were male (67.5 %) and 12 were female (32.5 %). Twenty six cases (70.27 %) were the term – gestation age, 10 cases (27 %) were preterm infants and 1 case (2.7 %) was a post-term infant. Eleven cases (29.7 %) were born in the King Chulalongkorn Memorial Hospital and the other 26 (70.3 %) were referred from primary or secondary hospitals.

The maximum birth weight of the 37 infants was 3,500 gm.,and the minimum was 1,300 gm. (mean weight = 2,426.2 gm.).

Congenital anomalies recorded in 36 infants.

- esophageal atresia with distal trachoesophageal fistula was the most common anatomic variant, occurring in 30 cases (83.3 %) as shown in Table 1.

- associated congenital anomalies were detected in 18 patients (50 %) (Table 2.) Major cardiac anomalies were found in 4 infants. These were dextrocardia with ventricular septal defect (1 of 18), and ventricular septal defect alone (3 of 18). None had initial palliative or corrective cardiac surgery. Seven of the 18 cases had 2 associated congenital anomalies, 3 had 3 or more associated congenital anomalies, and 3 were VATER association (esophageal atresia + hemivertebrae + anorectal malformation in all 3 cases).

Table 1. Type of esophageal anomaly (1989 – 1998).

Type of esophageal anomalies	Number of patients
esophageal atersia with distal tracheoesophageal fistula	30
esophageal atersia alone	5
esophageal atersia with proximal tracheoesophageal fistula	1
Total	36

Table 2. Congenital anomalies in 18 infants.

Type of associated congenital anomalies	Number of anomalies
dextrocardia + VSD	1
VSD alone	3
PDA	4
hemivertebrae	3
ano – rectal malformation	10
polydactyly + syndactyly	1
radial dysphasia	1
duodenal atresia	1
hypospadias	2
craniosynostosis	1
cleft palate	1
Down's syndrome	1
pectus excavatum	1
single umbilical artery	1

Surgical procedure in 36 patients

Gastrostomy with or with out cervical esophagostomy was performed in all cases. Five infants (13.8 %) died before esophageal correction. Primary repair and delayed esophageal correction (post – gastrostomy) with in 10 days) were performed in 28 cases (77.7 %). These were 25 cases (69.4 %) for esophago – esophagostomy, 2 cases (5.5 %) for colonic interposition and 1 case (2.7 %) for ileocolic interposition. Staged esophageal correction was done in 3 cases (8.3 %). These were case for esophago – esophagostomy, 1 cases for gastric transposition, and 1 case for colonic interposition. Post operative complications occurred in 18 of these 36 cases (50 %), as shown in Table 3.

The overall survival rate in the 36 cases was 69.4 %. Survival statistics according to the Waterston classification were as follows: in group A, 8 of 8 patients survived (100 %); in group B, 14 of 18 patients survived (77.7 %); and in group C, 3 of 10 patients survived (30 %) (Table 4). This represented a significant difference between the 3 groups (Fisher's Exact test at p value < = 0.003).

The Spitz classification is based on birth weight and the presence of major congenital heart disease. The Spitz survival rates are shown in Table 5. The survival rates were 74.2 % for group 1, 50 % for group II, and 0 % for group III, but the confidence level was not statistically significant (Fisher's Exact test at p value < = 0.228).

However, we modified the birth weight in the Spitz criteria from 1,500 gm to 1,800 gm as shown in Table 6 and the corresponding survival rates for groups I,II and III were 76.6 %, 50 % and 0 %, respectively. This modified Spitz classification made a significant difference between these groups (Fisher's Exact test at p value < = 0.03).

The overall mortality rate was 30.5 %. The causes of death were septicemia, heart failure, upper gastrointestinal bleeding and pulmonary complications such as pneumonia, pneumothorax and persistent pulmonary hypertension. Three of the 4 cases of major cardiac anomalies (VSD) died from pneumonia (1 case), sepsis (1 case) and heart failure (1 case). And 2 of the 4 cases of PDA died from sepsis.

Table 3. Post operative complications in 18 patients.

Type of complications	Number of complications
pneumonia	7
anastomotic stricture	7
pneumothorax	1
persistent pulmonary hypertension	1
lung atelectasis	1
recurrent TEF	1
anastomotic leakage	6
upper GI bleeding	1

Table 4. Survival rates by Waterston classification.

Group classification	No. of survivors	No. of deaths	Total	Survival rate
Group A: birth weight > 2500 gm., no pneumonia, no anomalies	8	0	8	100 %
Group B: birth weight = 1800 - 2500 gm., no pneumonia, no anomalies ; or birth weight > 2500 gm, moderate pneumonia with anomalies	14	4	18	77.7 %
Group C: birth weight < 1800 gm., or birth weight > 1800 gm, severe pneumonia and anomalies	3	7	10	30 %
Total	25	11	36	69.4 %

Table 5. Survival rate in Spitz classification.

Group classification	No. of survivors	No. of deaths	Total	Survival rate
Group I: birth weight >=1500 gm., no major cardiac anomalies	23	8	31	74.2 %
Group II: birth weight < 1500 gm., or major cardiac anomalies	2	2	4	50 %
Group III: birth weight < 1500 gm. with major cardiac anomalies	0	1	1	0 %
Total	25	11	36	69.4 %

Table 6. Survival rates under the modified Spitz classification.

Group classification	No. of survivors	No. of deaths	Total	Survival rate
Group I: birth weight > =1800 gm., no major cardiac anomalies	23	7	30	76.6 %
Group II: birth weight < 1800 gm., or major cardiac anomalies	2	2	4	50 %
Group III: birth weight < 1800 gm. with major cardiac anomalies	0	2	2	0 %
Total	25	11	36	69.4 %

Discussion

King Chulalongkorn Memorial Hospital is a tertiary care center and about 70 % of the esophageal atresia patients are referred cases. Half of the infants with esophageal atresia had other congenital malformations. The management of infants born with multiple anomalies, such as VATER or VACTERL associations, should be conducted in a planned, logical manner provided that all abnormalities are manageable. Congenital cardiac anomalies are major health factors. All cases should be examined by echocardiography, but not necessarily before esophageal correction. The non - controllable severe cardiac conditions may be improved by initial palliative or corrective cardiac surgery. In casesof gastro - intestinal obstruction, such as imperforate anus, colostomy may be performed in the same operation. Gastrostomy was performed routinely for gastric decompression and early post - operative enteric feeding. But cervical esophagostomy was only done in selected cases. The optimal time for definitive esophageal correction depended on life – threatening conditions bound in some cases. The main procedure for esophageal correction was esophago – esophagostomy anastomosis (72 %). In the last few years, primary esophago – esophagostomy was a procedure was a procedure of choice, evern for a long gap esophageal atresia.

Post – operative aspirated pneumonia was found in 20 % of cases, and may be caused by swallowing difficulty, esophageal dysmotility,[12,13] gastro – esophageal reflux [13,14] and anastomotic stricture, especially in a long gap atresia.[8] All cases of esophageal anastomotic stricture were treated with cycle of dilatation. The most severe early post operative complication seemed to be anastomotic leakage that caused infective mediastinitis and was found in the presence of long gap atresia. In

these cases, we found that cervical esophagostomy diversion was unnecessary and only conservative treatment may be the optimum choice. Adequate suction drainage via extra – pleural tube drain and frequent oropharyngeal suction, including bread – spectrum parenteral antibodics, were enough. The results of esophagography after the complete healing phase of these cases were satisactory.

Waterston, et al [1] proposed a classification in 1962 to categorize infants with esophageal atresia and this showed survival rates of 95 %in group A, 68 % in group B and 6 % in group C. Even though the survival statistics according to the Waterston classification were significant in our series, it may not be appropriate. This is because the survival rate in our group A was 100 %. This likely results from greatly improved neonatal intensive care improvement and because the birth weight cut-off point at 2,500 gm was too high and/or because associated congenital anomalies were not significant risk factor. For these reasons, the newer Spitz criteria used lower birth weights and ignored the minor congenital anomalies.

Though group survival rates under the Spitz classification seem different among each group they were not statisfically distinguishable, thus we increased the birth weight in the Spitz criteria from 1.500 gm to 1,800 gm for another prognostic criteria, and the confidence level of prognostic data was acceptable. The reasons may be problems in low birth weight neonatal care, and pneumonia or non - cardiac anomalies that were still influential factors. And we had a small samples size that may have effected the statistical significance in this series. Nevertheless, the modified Spitz classification may be another appropriate criteria for regional use. In the future, if the treatments steadily improve especially in low – birth - weight neonatal care and respiratory preventive care, the original Spitz classification could be used to stratify accurately and could be compared with other centers.

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