The THAI Journal of SURGERY 2000; 21:125-128.
Official Publication of the Royal College of Surgeons of Thailand

Esophageal Atresia : Six-year Experience with 105 Cases

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Abstract

Background: **Care of infants with esophageal atresia has generally been improved. Data from previous review of 87 cases treated over a 10-year period (1983-1992) formed the basis for this follow-on study.**

Materials and Methods: **A retrospective study of 105 infants with esophageal atresia admitted to the Children's Hospital, Bangkok, during a 6-year period (1993-1998) was made.**

Results: **The incidence was 1.2 : 10,000 live births. The most common type, esophageal atresia with distal tracheoesophageal fistula, was noted in 90 percent of cases. Risk factors influenced against survival included birth weight below 2,000 grams and cardiac anomalies. Associated anomalies were detected in 65 percent of cases. Primary repair of the esophagus was done in 21 patients, while delayed repair was done in 57 infants. Esophageal replacement was necessary in 3 cases. The most frequent postoperative complication was atelectasis. The overall mortality rate was 59 percent.**

Conclusion: **Results from this current review were comparable with those of previous study. No significant improvement was discerned since the majority of patients were in poor-risk group. More effort has been made in detecting associated anomalies.**

Esophageal atresia (EA) is the most common congenital anomaly of the esophagus.[1] It was considered a uniformly fatal condition until 1939. Since then, many major centers have documented a decline in the overall mortality of EA with each passing decade.[2-4] In 1996, one of the authors published a review of 87 cases of EA treated at the Children's Hospital, Bangkok, between 1983 and 1992.[5] This current report documents the incidence, risk factors, and the overall management in a consecutive series of patients treated ever since.

Materials and Methods

The medical records of all infants with EA admitted to the Neonatal Surgical Unit at the Children's Hospital, Bangkok, during a 6-year period (January 1993 to December 1998) were reviewed. The patients were categorized into the three Waterston risk groups,[6] and survival rates were determined. Particular attention was directed at the influences on survival of birth weight and of associated anomalies, especially cardiac malformations. Operative procedures, complications,

and outcomes were delineated. Postoperative atelectasis was counted on the basis of radiological reports. The survival in each group was compared by chi-square test.

Results

Incidence

From 1993 through 1998, 107 neonates with EA were treated. Medical records were available for study in 105 cases. There were 60 males and 45 females, hence the male to female ratio was 1.33:1.

Eleven patients were born at Rajavithi Hospital, during which time there were 90,578 live-births,[7] therefore the incidence of EA was 1.2:10,000 live-births.

Type of anomaly

Only two types of EA were present. The most common one, EA with distal tracheoesophageal fistula (TEF), occurred in 95 cases (90.5%), and the other type, EA without TEF, occurred in 10 cases (9.5%).

Risk groups

The patients were customarily grouped according to Waterston's Classification.[6] Sixteen percent of infants were allocated to group A, 46 percent to group B, and 38 percent to group C. The survival rates according to the risk groups are shown in Table 1. The overall survival rate was 40.9 percent.

Table 1 Grouping of patients according to Waterston's Classification

Group	No. of Patients (%)	No. of Survivors	% of Survivors
A	17 (16.2)	15	88.2
B	48 (45.7)	24	50.0
C	40 (38.1)	4	10.0
Total	**105 (100)**	**43**	**40.9**

Table 2 Influence of birth weight on survival

Weight (gm)	No. of Patients (%)	No. of Survivors	% of Survivors
≥ 2,000	76	40	52.6
< 2,000	29	3	10.3

P < 0.005

Birth weight: Only 10 percent of infants weighing less than 2,000 grams survived, compared with 52 percent of those weighing more ($p < 0.005$) (Table 2).

Cardiac anomalies: Fifty infants had associated cardiac anomalies documented by electrocardiography, echocardiography, or color flow doppler. When these were considered together with birth weight, they further reduced survival rates desperately (Table 3).

Associated anomalies

Sixty-nine patients (65.7%) had a total of 123 associated anomalies. A classification of anomalies according to the various systems affected and survival in each group are shown in Table 4.

Operative procedures

Gastrostomy was performed in 84 infants, 57 of whom subsequently underwent esophageal anastomosis. Ten infants had long gap EA precluding

Table 3 Survival rate according to birth weight and congenital heart diseases (CHD)

Birth Weight (gm)	CHD	Total	Survivors	Survival Rate (%)	
≥ 2,000	No	44	32	72.7	$p < 0.005$
≥ 2,000	Yes	32	8	25.0	
< 2,000	No	11	2	18.1	$p > 0.05$
< 2,000	Yes	18	1	5.5	

Table 4 Associated anomalies versus survival

System Affected	No. of Patients (%)	Survivors (%)
No anomaly	36 (34.2)	24 (66.6)
Cardiac	50 (47.6)	9 (18.0)
Urinary tract	12 (11.4)	4 (33.3)
Gastrointestinal		
- anorectal	16 (15.2)	5 (31.2)
- other	3 (2.8)	1 (33.3)
Orthopedic		
- vertebral	17 (16.1)	9 (52.9)
- other	12 (11.4)	2 (16.6)
CNS	2 (1.8)	1 (50.0)
Chromosomal	1 (0.9)	- (-)
Miscellaneous		
- cleft lip / palate	4 (3.8)	- (-)
- other	6 (5.7)	2 (33.3)

Table 5 Operative procedures performed

Gastrostomy	84	
Ligation of TEF	10	
Anastomosis	78	
with preliminary gastrostomy		57 (73.1 %)
without preliminary gastrostomy		21 (26.9 %)
extrapleural approach		70 (89.8 %)
transpleural approach		8 (10.2 %)
Esophageal replacement	3	
gastric transposition		2
colon interposition		1

Table 6 Complications

	No. of Patients	%
Atelectasis	78	74.2 (N = 105)
Leak	30	38.4 (N = 78)
Stricture	8	10.2 (N =78)
Recurrent TEF	2	2.5 (N =78)

N = Total number of patients operated upon

anastomosis, so ligation of TEF was performed. Only two patients in this group survived esophageal bougienage and had successful anastomosis. Primary repair of the esophagus without preliminary gastrostomy was done in 21 infants. Esophageal anastomosis was attempted in 78 infants, 70 via the extrapleural approach and 8 transpleurally (Table 5).

Three patients needed esophageal replacement; two with isolated EA had gastric transposition, the other one with EA and TEF but long gap had colon interposition. All these three survived.

Complications

Atelectasis occurred in nearly 75 percent of the patients. Regarding anastomotic complications, leaks were identified in 30 patients (38.4%). The majority of leaks were minor ones which sealed spontaneously on conservative treatment. Strictures developed in 8 patients (10.2%), three of whom were so severe that they required resection of the strictures. Recurrent TEF was noted in 2 infants (Table 6).

Outcomes

Sixty-two infants died. Nearly half of these succumbed to sepsis. Others died of various causes as shown in Table 7.

Table 7 Causes of death in 62 patients

Cause	No. of patients
Sepsis	30
Respiratory failure	14
Heart failure	7
Pneumonia	5
Others	6

Discussion

From the previous review of EA treated at this hospital in the 10-year period,[5] the incidence of EA was around 1:10,000 live-births, risk factors were birth weight below 2,000 grams and associated cardiac anomalies.

This current study of 6-year period had shown rather the same incidence which was much lower than those of other series.[4,8,9]

Regarding the two risk factors of birth weight below 2,000 grams and cardiac anomalies, they still had considerable influences against survival. The doubly increased incidence of cardiac anomalies, with respect to that of the previous study,[5] indicated more effort to detect them by echocardiography and color flow doppler. However, echocardiography and color flow doppler were not routinely done in all cases, but selectively in ones with clinical pictures of congenital heart diseases. Therefore the actual incidence of cardiac anomalies might be higher.

Associated anomalies were classified according to the systems affected as advocated by Myers and coworkers.[10] Overall incidence of associated anomalies remained comparatively high in regard to other series.[4,10-15]

Primary repair of the esophagus without preliminary gastrostomy was done in more than 25 percent of patients receiving anastomosis, as opposed to none in the previous report.[5] Gastrostomy was considered mandatory as a vent in infants who had distal TEF and pneumonia precluding immediate primary anastomosis, and as a feeding route in ones without TEF. Thoracotomy by extrapleural approach was preferable to transpleural one, and was done in nearly 90 percent of infants, since it was considered to be safer in case of anastomotic leaks. For long gap EA requiring esophageal replacement in which anastomosis was not

possible, our experience was limited due to the small number of patients (only three) but thus far the outcome had been encouraging.

Concerning complications, atelectasis posed as a major threat postoperatively in three quaters of patients indicating nursing care inadequacy. Anastomotic complications, either leaks or strictures, were usually mild and required no subsequent surgical intervention.

The overall mortality rate was discouragingly high. Low birth weights and cardiac anomalies contributed to such outcome.

In conclusion, the lessons learned from this review are : (1) the incidence of EA was 1.2 : 10,000 live-births, (2) risk factors included birth weight below 2,000 grams and cardiac anomalies, (3) detection of associated anomalies, especially cardiac ones, were attempted more frequently than in the previous review, (4) primary repair of the esophagus was chosen when the patient's condition permitted, and extrapleural approach thoracotomy was preferable.

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