TWENTY FIVE YEARS OF INTERACTION WITH ESOPHAGEAL ATRESIA AND TRACHEO-ESOPHAGEAL FISTULA

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ABSTRACT

Clinical presentation and management modalities for 585 patients of esophageal atresia and/or tracheo-esophageal fistula reporting to Department of Paediatric Surgery, SMS Medical College, Jaipur over twenty five years (1972-1996) were analyzed in five phases of five years each retrospectively. Over the period of observation, the incidence of new cases as well the number of associated anomalies have shown a steady increase due to availability of modern diagnostic facilities. The operative mortality has shown a progressive decline from 95% early in the series to 40% in last few years. Post-operative anastomotic leaks occurred in 21% of cases, strictures in 8% and recurrent fistula in 1% of cases. The presence of severe associated anomalies, prematurity, pulmonary complications and sepsis still remain the major killers in our setup. Early detection and referral has lead to more infants reaching the specialized centres with less pulmonary complications. In the last decade, the survival rate for infants in Waterston's risk group A has improved to 86% but there is still a substantial difference in the survival rate reported from developed countries. The various factors resulting in poorer results in our set up have been highlighted.

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KEY WORDS: Esophageal atresia; Tracheo-esophageal fistula.

Introduction

Esophageal atresia has always presented a challenge to the treating paediatric surgeons. It is the epitome of modern surgery [1]. Embryologically, esophageal atresia is due to failed canalization of the solid rod of cells forming the esophagus throughout its length while the tracheo-esophageal fistula is result of failure of complete midline fusion of the tracheo-esophageal septum. William Durston in 1670 [2] is credited with first description of congenital atresia of esophagus. The usual form of the anomaly as associated with tracheo-esophageal fistula was first recorded by Thomas Gibson in 1697 [3]. The first successful primary repair was performed by Cameron Haight in 1941 [4]. The department of paediatric surgery was established in 1971 at SMS Medical College, Jaipur and first survival with this anomaly from our department was in 1972 [5]. Since then we have come a long way and present here a multiphase analysis of 585 patients with emphasis on the problems that tend to vitiate outcome in this anomaly.

Material and Methods

From 1972 to 1996, a total of 585 infants with atresia and/or tracheo-esophageal fistula were admitted to the Department of Paediatric Surgery, SMS Medical College & Hospital, Jaipur, India. The medical records of all patients with the diagnosis of esophageal atresia and tracheo-esophageal fistula admitted from

1972 to 1996 were analyzed with reference to the age, sex, birth weight, types, symptoms, complications, associated anomalies, methods of treatment and mortality. Each infant was also classified into risk group A, B & C according to Waterston's criteria. Patients were divided into 5 phases of 5 years duration each and analysis was made as regards to any change in the patient profile and the results over the time. Standard extrapleural single layer end to end anastomosis was used till 1990. But we have modified this technique by oblique oriented anastomosis in recent years. In this technique, a flap of upper pouch is constructed and after spatulation of the lower end, an oblique circumferential anastomosis is carried out (Fig 1). 5-0 silk was used initially for esophageal anastomosis, now we have changed to 5-0 vicryl/dexon. After completion of the anastomosis, if there was undue tension, circular myotomy was done with fine pointed scissors until the mucosa was exposed. We never used transanastomatic tube. The study with gastrograffin was performed on eighth postoperative day and if there was no leakage, oral feeding was started next day-initially with clear fluids and then gradually changing to milk. Extrapleural drain was removed on 10th post-operative day. Esophagus was routinely calibrated with a FG-10 red rubber catheter after 3 weeks without anaesthesia. This calibration was also done on routine follow up at monthly interval.

Results

The distribution of the 585 patients in this series was as follows: Phase I-65 patients, Phase II-60 patients, Phase III-84 patients, Phase IV-166 patients and Phase V-210 patients. The incidence was 12.5 cases per 1000 surgical admissions of the Institute. There was a male preponderance (4:1) in our series : 469 boys and 116 girls. The pattern of the anatomical types of the anomaly followed the usual proportional incidence: esophageal atresia with distal fistula in 90%, esophageal atresia without fistula in 7%, esophageal atresia with double fistula in 0.5% and tracheo esopha-

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Fig. 1 :

geal fistula without atresia (H type) in 2%. Two patients (0.34%) were diagnosed as having esophageal stenosis/ web. No patient with an isolated proximal fistula was seen in this series (Table 1).

Most of the patients in this series had low birth weight: 27.7% were over 2500 gms, 43.6% between 1800 and 2500 gms & 28.7% cases were below 1800 gms. Maximum number of patients were admitted within 24 hours of birth (46%). Out of the rest, 20%

reported between 1 to 2 days, 23% between 2 to 5 days and 11% after 5 days of birth. But, the oldest patient in our series presented at the age of 12 days. Thirty nine percent of patients had one or more associated anomalies (Table 2). The most frequently encountered anomaly was ano-rectal malformation seen in as many as 15% of cases. With improved diagnostic techniques like echocardiography, the incidence of associated congenital heart disease which was 3% in first phase has increased to 9% in the fifth phase.

Primary repair of the Esophagus was performed in 55% of patients in the entire series (Table 3). There was a significant number of patients (29%) with severe associated anomalies, prematurity and sepsis, which were incompatible with survival in these only gastrostomy was done and all patients died.

Two hundred cases were evaluated in relation to the gap between the two esophageal segments with special reference to complications and survival. Out of 200 cases, 154 had a gap of < 2 cm and 46 had a gap of > 2 cm. In the group with < 2 cm gap, fistula ligation and primary anastomosis was carried out in 112 cases with survival rate of 82% and gastrostomy and primary anastomosis was done in 42 cases with survival rate of 45%. In the group of > 2 cm gap, primary anastomosis was done in 30 cases with 60% survival and no case survived after fistula ligation and gastrostomy in 16 cases. Two cases of long gap isolated esophageal atresia survived after staged management. Complications in relation to long gap were much more than short gap. The incidence of the anastomotic leak was 16% in patients with < 2 cms gap and 33% in those with > 2 cms gap. Similarly incidence of post-operative stricture was 3% in patients with < 2 cms gap and 9% in those with > 2 cms gap. Recurrent fistula was in 0.65% in patients with < 2 cms and 3% in those with > 2 cms gap.

In overall complications, the incidence of anastomotic leakage has fallen from 37% in the first phase to 17% in last phase. Pulmonary complications have remained steady over the period: 64% in

TABLE 1
Percentage of anatomical types of esophageal anomalies

Type of atresia	Phase I (n=65)	Phase II (n=60)	Phase III (n=84)	Phase IV (n=166)	Phase V (n=210)	Overall (n=585)
Atresia alone	6	10	10	6	7	7
Upper fistula	—	—	—	—	—	—
Lower fistula	94	87	89	90	90	90
H type fistula	—	3	1	2	2	2
Double fistula	—	—	—	1	1	0.5
Stenosis / web	—	—	—	1	—	0.5

TABLE 2
Percentage of associated anomalies at presentation

Anomaly	Phase I (n=65)	Phase II (n=60)	Phase III (n=84)	Phase IV (n=166)	Phase V (n=210)	Overall (n=585)
Ano-rectal malformation	22	18	19	13	12	15
Congenital heart disease	3	2	8	9	8	7
Gastrointestinal	6	8	5	5	5	5
Skeletal	6	8	8	6	4	6
Genitourinary	—	2	2	4	4	3
Tracheomalacia	—	—	1	1	2	1
Right aortic arch	2	—	—	2	3	2

n = number of cases

TABLE 3
Percentage of patients who underwent various operative procedures

Operative Procedure	Phase I (n=65)	Phase II (n=60)	Phase III (n=84)	Phase IV (n=166)	Phase V (n=210)	Overall (n=585)
Primary anastomosis with or without gastrostomy	22	35	54	68	62	55
Cervical esophagostomy and gastrostomy	12	10	4	2	2	4
Gastrostomy alone	32	30	28	27	29	29
Gastrostomy and fistula ligation	12	7	5	1	4	5
Fistula ligation alone (H type fistula)	—	3	1	1	3	2
Gastrostomy and excision of esophageal web	—	—	—	1	—	—
No operative procedures	22	15	8	—	—	5

Note : All figures have been rounded off; n = number of cases

TABLE 4
Complication rate (%)

Complications	Phase I (n=65)	Phase II (n=60)	Phase III (n=84)	Phase IV (n=166)	Phase V (n=210)	Overall (n=585)
Anastomotic leak	37	30	21	16	17	21
Stricture	12	10	12	9	8	8
Recurrent fistula	2	—	—	0.5	1	1
Pulmonary	64	60	45	55	60	57
Wound infection	12	16	13	10	3	11

n = number of cases

TABLE 5
Distribution of cases according to the Waterston's criteria¹⁵ and their survival

Group	Phase I		Phase I		Phase III		Phase IV		Phase V		Overall	
	No of cases	Surviv (%)	No of cases	Surviv (%)	No of cases	Surviv (%)	No of cases	Surviv (%)	No of cases	Surviv (%)	No of cases	Surviv (%)
A	4	25%	5	40%	10	60%	31	87%	72	86%	122	80%
B	13	15%	12	25%	21	33%	45	58%	98	52%	189	47%
C	48	0%	43	2%	53	8%	90	27%	40	28%	274	15%
Overall survival	65	5%	60	10%	84	20%	166	46%	210	59%	585	39%

first phase and 57% in the last phase (Table 4).

The overall survival in different phases with regards to Waterston's risk group A, B and C was 80%, 47% and 15% respectively. The pattern of survival has shown a favourable trend in last decade with a primary survival rate of 86% for Waterston's group A, 52% for group B and 28% for group C in the period between 1992-1996. The mortality rate has shown progressive decline from 95% at the beginning to 40% in the last phase (Table 5).

Discussion

Esophageal atresia with or without a fistulous communication with trachea is a common pediatric surgical emergency. The first survival with this anomaly from our department was in 1972 [5] and on completion of 25 years 585 patients have been treated. In

latter decades, enormous progress has been made in the management of esophageal atresia and tracheo-esophageal fistula to the point of achieving survival in the vast majority of patients, including most premature infants. There is still a significant difference in the results of this entity in developing and developed countries [6].

Our policy for the treatment of esophageal atresia was that if the general condition is good and there were no major associated anomalies, we performed primary anastomosis without gastrostomy. In patients with severe pulmonary complications, gastrostomy under local anesthesia was done which was followed

by delayed primary anastomosis usually after 3-7 days when the chest condition improves.

The incidence of post-operative stricture formation was low in our series (8%) in comparison to series reported by Spitz et al [7,8] in 1987 (18%) and 1996 (10%). The contributory factor for the low incidence of stricture formation may be due to routine post-operative calibration and modified oblique anastomosis used by us in last phase [9].

In the developed western world birth weight and pneumonia are no longer considered as prognostic determinants [10] but in our set up low birth weight, chest infection and septicemia remain major causes of death apart from severe associated anomalies.

A long gap esophageal atresia (2cms) was managed by mobilization and upper pouch circular myotomy in 18 cases and esophageal flap esophagoplasty in 5 cases as described by Gough [11]. None of the patients survived where fistula ligation and gastrostomy was done for delayed primary anastomosis.

Our 30 patients with isolated esophageal atresia underwent gastrostomy alone or gastrostomy and cervical esophagostomy as the primary procedure. The majority of these succumbed to sepsis over the waiting period. Two such patients with gastrostomy and cervical esophagostomy underwent delayed definitive procedure and survived. As regards tracheo-esophageal fistula without esophageal atresia (H type), this is an entity that is frequently diagnosed but seldom confirmed. We have confirmed 10 such cases endoscopically (tracheobronchoscopy). These patients had fistula ligation using the right cervical approach.

The distal esophageal web was diagnosed in two cases by air contrast distal esophagogram performed through gastrostomy [12]. After confirmation of diagnosis, the perforation of membrane was done using bugbee electrode through the esophagoscope.

Overall survival which was a dismal 5% in 1972-1976, had increased to 57% in 1992-1996. Although we had only 28% survival in Waterston's category C during last phase, the survival in Waterston's risk group A was 86%; not much behind the developed countries (7,13,14,15). The transition through these two and half decades has seen a distinct change in referral pattern. Sixty six percent of the babies are now been referred to us within 48 hours of life (Fig 2). Over the years, better understanding of the physiology and availability of newer antibiotics, has resulted in better peri-operative care and improved survival.

Manning in 1987 [16] have remarked that a plateau has probably been reached as regards the outcome in

esophageal atresia. This may be true with regard to the developed countries, but many a hurdle will have to be surmounted before we achieve this in our set up. As the majority of the Indian people are poor, illiterate and live in distant remote areas where no health care has yet reached [17]. Most of the deliveries in rural areas are conducted by unskilled midwives who are not even aware of this anomaly. This results in continuance of oral feeding of these babies leading to aspiration into lungs thus further deteriorating their pulmonary status. Even after the diagnosis, transport of the baby to proper surgical centre takes long time due to inadequate facilities. Only a small fraction (less than 10%) of these babies reach major referral centres [18]. There are also no facilities to suck out the upper pouch of the baby during the long hours of transportation. All these factors result in the majority of patients coming late and having severe pneumonia at the time of admission. Most of these babies are also underweight. In our series 72% of patients were below 2.5Kg weight at the time of admission.

In spite of all these restraints, safer anaesthesia, improved post-operative care and more efficient management of complications and newer antibiotics have resulted in steady decline in mortality rate.

To sum up, the practice of medicine in developing countries differs substantially from that in developed countries, owing to socio-economic factors and geographical influence. The improvement in results of esophageal atresia over these last two and half decades is quite encouraging. We recommend following guidelines for further improvement in survival of these patients: (i) High degree of clinical suspicion is required in babies regurgitating all feeds or having continuous dribbling of saliva or in whom red rubber catheter could not be passed beyond 10 cms. (ii) Provide immediate supervised transport to the nearest tertiary care centre. (iii) Transport the babies in lateral or prone position with repeated suction by oral mucus aspirator or by syringe. (iv) Keep the patient fasting (v) Keep affected side of chest uppermost for dependent drainage and (vi) Keep the baby warm.

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