

Factors Influencing Survival in Esophageal Atresia

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ABSTRACT

There is little published literature from the third World countries that described the factors influencing survival of babies with esophageal atresia. We analysed 25 consecutive neonates treated for esophageal atresia. The overall survival rate was 36%. All 4 babies in Waterslon Group A, 37.5% in Group B, and 15.4% in Group C • survived. All 9 preterm babies died. Only 2 of the 16 babies who had pre-operative chest infection survived. The mean delay in diagnosis was 54 h in outborn babies and 20 h in hospital-born babies. We believe that a survival rate of 40% is easily achieved with minimum infras-truclural inputs. Simple methods and practices that would vastly improve operative results have been suggested.

Key words: Mortality, Esophageal atresia.

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The outcome of babies treated for esophageal atresia with or without tracheo-esophageal fistula (TEF) reflects the standard of neonatal surgical intensive care available at a particular institution. In the last four decades, results in the West have shown a remarkable improvement. Presently, a baby born with TEF should have a nearly 100% chance of survival if there are no associated major congenital anomalies(1). In India, however, most major institutions have overall survival rates of between 40-55% (2,4). Surprisingly, there is a lack of published Indian data that addresses the problems faced in the management of these children. We present our experience in the 25 cases that were treated at our new hospital that has only basic facilities for the care of surgical neonates.

Material and Methods

Twenty five consecutive cases of EA with or without TEF that were treated at our hospital between January 1988 and May 1992 were analysed. All clinical data was carefully recorded. All babies underwent surgical correction as soon as possible after diagnosis was established and the general condition of the baby allowed. Primary esophageal anastomosis was attempted whenever possible, except in the moribund patient.

Results

There were 12 male and 13 female babies. The mean birth weight was 1.79 kg (1.25-3.5 kg). Nine babies were preterm. Sixteen babies were born in hospital, of which 5 were born in another hospital and were referred for surgery. Esophageal atresia without TEF was seen in 2 patients, a gastrostomy and cervical esophagostomy was done and both are alive. Twenty two had EA with distal TEF. One patient had double fistula, from the proximal as well as the

distal esophagus. Associated anomalies were seen in 9 patients (36%) which included anorectal malformations in 6 (5 supralevator and 1 infralevator); Material talipes (1 case); absent azygos vein (1 case) and polydactyly (1 case). In 2 patients, the diagnosis was suspected on antenatal ultrasound; both underwent successful surgical correction immediately after birth.

In the 9 cases that were born at home, the mean delay in presentation to hospital was 33 h (range 3-96 h). There was a further delay in establishing the correct diagnosis in hospital (mean 21 h; range 4-48 h). The commonest symptom recorded was "failure to feed" which was interpreted and treated as neonatal septicemia till later, when the correct diagnosis was made. In one case, "frothing at the mouth" was misinterpreted as neonatal convulsions.

In the 16 hospital-born babies also there was a delay in diagnosis (mean 20 h; range 0-48 h).

Sixteen cases had pre-operative chest infection characterized by presence of crepitations, radiological findings or operative detection of atelectasis in the right lung. Six had mild chest infection while in 10 the infection was bilateral and extensive. Further, 3 cases had pre-operative apneic spells and/or cardiac arrests.

Nine babies survived (overall survival rate 36%). The break-up of the outcome of cases based on various criteria is given in *Table I*. All the 4 cases in Waterston Group A survived. The survival rate dropped significantly in Groups B and C cases. All 9 preterm babies died. Of the 16 cases with established chest infection pre-operatively, only 2 survived. The various factors contributing to mortality are listed in *Table II*. In only 1 case, in the initial part of our

TABLE I—Outcome of Operated Cases

	n	Alive (%)	Dead
Waterston group			
A	4	4 (100.0)	0
B	8	3 (37.5)	5
C	13	2 (15.4)	11
Associated anomalies	9	3 (33.3)	6
Preterm	9	0	9
Pre-operative chest infection			
Mild	6	1	5
Severe	10	1	9

experience, hypothermia contributed to the mortality. Lack of availability of ventilatory support precluded the salvage of some babies. Three of the 6 babies with associated anorectal malformations survived.

TABLE II—Factors Directly Contributing to Mortality

Factor	No.
Prematurity	9
Septicemia, chest infection	8
Anastomotic leak	1
Persistent tachycardia, cyanosis (? CHD)	1
Hypothermia	1

The operative procedures performed and their outcome is given in *Table II*. Primary esophageal anastomosis was attempted whenever possible, except in the moribund patient when a gastrostomy and cervical esophagostomy was done. One baby was referred to us with a gastrostomy. A right transverse colostomy was added in the 5 babies with supralevator anorectal malformations.

TABLE III—Outcome of Various Surgical Procedures

Operative procedure	n	Alive	Dead
Primary repair	15	5	10
Primary repair + colostomy	4	2	2
Gastrostomy + delayed primary repair	1	0	1
Esophagostomy + gastrostomy	4	1	3
Esophagostomy + gastrostomy + colostomy	1	1	0

Discussion

Results of treatment of TEF from the major centres in India are similar with the overall survival rates varying from 40-55%(2,4). These results are in sharp contrast to those from the West where the overall survival rates are over 85%(1). At most hospitals in our country the survivals are only anecdotal. Our survival figures compare favorably with the major Indian centres. One is tempted to conclude that a figure of 40% can be readily achieved even with a minimum input in terms of manpower and sophisticated gadgetry. How then can we improve upon these results? A careful scrutiny of the data provides the reasons for our poor results.

Prematurity: All 9 babies in this study that were preterm died. Both babies who had an antenatal diagnosis and were repaired immediately after birth survived. This underscores the role of antenatal suspicion and careful obstetric monitoring in the last trimester to carry the fetus to term as far as possible.

Delay in diagnosis: The most distressing observation in this study was the delay in presentation to the hospital compounded by a further delay in diagnosis in hospital. This was on an average 54 h in outborn babies and 20 h in hospital born babies. This delay

contributed substantially to the soiling of lungs and development of chest infection. Only 2 babies with pre-operative chest infection survived, in order to improve results, this is the factor that needs to be controlled. The standard nursery practice of passing a red rubber catheter into the esophagus of all hospital born babies will help. Similarly, resident pediatricians must think of this surgical cause when they are evaluating a neonate who presents with "failure to feed". In all such cases, and those with respiratory distress, no harm will be done if the patency of the esophagus is confirmed early.

Hypothermia: We were largely able to overcome the problem of hypothermia by simple measures in the absence of operating room heating. During the operation the baby was placed on a metal tray covered with a layer of cotton wool. Under the tray a rubber bag with hot water was placed which effectively maintained the baby temperature. The other technique was to incorporate a heat-over type of humidifier in the anesthesia circuit that delivered preheated, humidified anesthetic gases to the airway and also helped keep the core temperature up.

We chose to perform a primary esophageal repair in all except the moribund patients for three reasons: (1) we are unable to provide the intensive nursing care for prolonged periods in order to do a delayed esophageal

repair; (ii) it is our impression that of the babies in whom gastrostomy and esophagostomy has been done, few come back later for esophageal reconstruction, the remaining probably die in the waiting period due to various causes; (iii) esophageal substitution is a poor alternative and all attempts must be made to preserve the esophagus.

It was observed that the attitudes of residents and nursing staff contributed to the success in some of our cases. Since a majority of the staff had not seen a baby survive during their training period earlier, they all had a fatalistic approach towards these babies. However, when they did see the one that survived, their attitude changed and they looked after these babies with more enthusiasm and aggression.

Total Thoracic Stomach

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Large hiatus hernias are quite uncommon in children. "Total thoracic stomach", an extreme form in which the entire stomach herniates into the thorax is a very rare entity and few cases have previously been reported(1,2). Paradoxically, most of these patients present with minor symptoms at-

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tributable to gastro-esophageal reflux or recurrent chest infections(1). The diagnosis in such cases can frequently be made on chest X-ray although barium studies are confirmatory. The outcome of surgery is usually good, unless there is associated stricture of the esophagus and/or short esophagus(1,2).

We report two cases of "total thoracic stomach" which were associated with non-

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