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Esophageal Atresia with or without TEF-State of Art*

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Abstract

Esophageal Atresia (EA) encompasses a group of congenital anomalies comprising of an interruption of the continuity of the esophagus with or without a persistent communication with the trachea or bronchus.

In 90% of cases there is a distal tracheoesophageal fistula, in 7% there is no fistulous connection, while in 2% there is a tracheoesophageal fistula without atresia. EA occurs in 1 in 3500 admissions in the department of pediatric surgery. Infants with EA are unable to swallow saliva and are noted to have excessive salivation requiring repeated suctioning. Associated anomalies occur in 50% of cases, the majority involving one or more of the VACTERL association (vertebral, anorectal, cardiac, tracheoesophageal, renal and limb defects). A nasogastric tube should be passed at birth in all infants born to a mother with polyhydramnios as well as to infants who are excessively mucusy soon after delivery to establish or refute the diagnosis. Definitive management comprises disconnection of the tracheoesophageal fistula, closure of the tracheal defect and primary anastomosis of the esophagus. Where there is a "long gap" between the ends of the oesophagus, delayed primary repair should be attempted. Only very rarely will an oesophageal replacement be required. Survival is directly related to birth weight and to the presence of a major cardiac defect.

Keywords: Esophageal Atresia, Tracheoesophageal fistula

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Introduction

Although there has been a steady improvement in the management of patients with Esophageal Atresia (EA), a technical challenge still exists to improve the prospects of survival of these patients, in developing countries (1, 2). This report updated our experience with cases of EA with or without tracheoesophageal fistula (TEF) in the present scenario and highlights the problems faced in their management.

Material & Methods

During twenty-five years (1972 to 1996), a total number of 585 neonates with esophageal atresia, with or without tracheo-esophageal fistula were admitted in the Deptt. of Paediatric Surgery of SMS Medical College, Jaipur (India).

For the purpose of our present study, the patients have been grouped in five phases of five years each (1972 – 1976, 1977 – 1981, 1982 – 1986, 1987 – 1991 and 1992 – 1996). This division is arbitrary but roughly corresponds to the introduction of improved diagnostic tools like ultra-sonography, surgical methods of primary anastomosis even in cases with long gap, better suture material, newer anaesthetic techniques/ agents and antibiotics. The operative method used was standard extrapleural single layered end-to-end anastomosis. In 150 cases, the technique of oblique oriented anastomosis (3) was adopted, using 5.0 vicryl/dexon. The oblique anastomosis has a significant advantage of higher internal esophageal diameter, lower rate of break down and stricture formation.

Clinical description and diagnosis

The diagnosis of esophageal atresia may be suspected prenatally by the finding of a small or absent fetal stomach bubble on ultrasound scan performed after the 18th week of gestation. Overall, the sensitivity of ultrasonography is 42% but in combination with polyhydraminos the positive predictive value is 56% (4). Polyhydraminos alone is a poor indicator of oesophageal atresia (1% incidence). Available methods of improving the prenatal diagnostic rate include ultrasound examination of the fetal neck to view the blind-ending upper pouch (5) and to observe fetal swallowing and magnetic resonance imaging (6). The newborn infant of a mother with polyhydramnios should always have nasogastric tube passed soon after delivery to exclude oesophageal atresia. Infants with oesophageal atresia are unable to swallow saliva and are noted

to have excessive salivation requiring repeated suctioning. A plain X-ray of the chest and abdomen will show the tip of the catheter arrested in the superior mediastinum (T2-4) while gas in the stomach and intestine signifies the presence of a distal tracheoesophageal fistula. The absence of gastrointestinal gas is indicative of an isolated atresia.

Incidence of anatomical types of EA in our series is similar to the reported data (7, 8, 9, 10).

Over 50% of infants with esophageal atresia have one or more additional anomalies (11). The systems affected are as – cardiovascular 29%, anorectal anomalies 14%, genitourinary 14%, gastrointestinal 13%, vertebral/ skeletal 10%, respiratory 6%, genetic 4%, others 11%. The incidence of cardiac anomalies was less in our series but it went up with the increasing diagnostic facilities (10).

There is an increased incidence of associated anomalies in pure atresia (65%) and a lower incidence in H-type fistula (10%).

The VACTERL association consists of a combination of anomalies including vertebral, anorectal, cardiac, tracheo esophageal, renal abnormalities and limb defects (11). Other associations which may include esophageal atresia are the CHARGE association (coloboma, heart defects, atresia choanal, retarded growth and development, genital hypoplasia and ear deformities).

Pathophysiology of esophagus & trachea

The motility of the oesophagus is always affected in oesophageal atresia. The disordered peristalsis more commonly involves the distal oesophageal segment. The motility disorder is primarily due to abnormal innervation as evidenced by an abnormality in neuropeptide distribution (12, 13) though the Gangalion cells were normal (14). The resting pressure in the whole esophagus is significantly higher than in normal patients and the closing pressure of the lower esophageal sphincter is reduced.

The trachea is also abnormal in esophageal atresia. The abnormality consists of an absolute deficiency of tracheal cartilage and an increase in the length of the transverse muscle in the posterior tracheal wall (15). When severe, these abnormalities result in tracheomalacia with collapse of the trachea over a 1-2 cm segment in the vicinity of the fistula.

Risk categorization and prognosis (outcome):

In 1962, Waterston et al. (16) proposed a classification of infants born with esophageal atresia into three groups "with different chances of survival". The classification based on birth weight, associated anomalies and pneumonia:

Group A >2500 g birth weight and well

Group B
1. Birth weight (1800-2500 g) and well
2. Higher birth weight, moderate pneumonia and congenital anomaly.

Group C
1. Birth weight under (1800 g).
2. Higher birth weight and severe pneumonia and severe congenital anomaly.

Applying the Waterston classification to a series of 357 infants with esophageal atresia treated at Great Ormond Street from 1980 to 1992 (17), the results were : in Group A, 153 of 154 survived (99%), 72 of 76 in Group B survived (95%) and 101 of 142 in Group C survived (71%). In the last phase of our study, the survival rate improved to 78% Group A, 47% in Group B and Group C had only 27% survival (10). The new risk classification concerned birth weight and associated cardiac malformations which were previously identified as being responsible for most of the mortality.

The Spitz classification (17) for survival in esophageal atresia is:

Group I Birth weight over 1500 g with no major cardiac anomaly.

Group II Birth weight less than 1500 g or major cardiac anomaly

Group III Birth weight less than 1500 g PLUS major cardiac anomaly.

Major cardiac anomaly was defined as either cyanotic congenital heart disease that required palliative or corrective surgery or non-cyanotic heart anomaly that required medical or surgical treatment for cardiac failure.

Using the new risk classification scheme, survival was 97% Group I, 59% for Group II and 22% for Group III in the 1980's but has improved to 98%, 82% and 50% respectively (18). A study from Montreal identified only

preoperative ventilator dependence and severe associated anomalies as having prognostic significance (19).

Brown & Tam (20) suggested the measurement of gap length in esophageal atresia & provides a method of classification to predict morbidity & long-term out come.

We evaluated 200 cases with reference to the gap between two esophageal segments. Where the gap was >2 cm, the mortality and complication rate was significantly high (21).

Methods to overcome a wide gap

If there is a wide gap, the distal esophagus can be mobilized safely well down towards the diaphragm (22). Various maneuvers have been proposed to overcome a wide gap but in a very tense anastomosis can be achieved in most cases and if the infant is subsequently electively paralyzed and mechanically ventilated for approximately 5 days postoperatively, the anastomosis will heal without leakage (23, 24). Others have proposed tubularisation of the upper pouch after creating a flap (25), circular myotomy of the upper pouch (26), creation of posterior flap followed by end-to-end oblique anstomosis with a spatulated distal esophagus (3) or abandoning any attempt at initial primary anastomosis awaiting delayed primary anastomosis 6-12 weeks later (27).

Management of isolated esophageal atresia

The diagnosis of isolated esophageal atresia without a fistula should be suspected when on the initial radiograph there is no gas in the abdomen ("gasless abdomen"). We encountered two cases with esophageal web in our series which were diagnosed & successfully managed (28).

Once the diagnosis of isolated atresia is made, the next step is to perform a feeding gastrostomy and to estimate the extent of the gap between the proximal and distal esophagus.The gap between the esophageal ends is measured by injecting sufficient radio-opaque contrast into the stomach to allow it to enter the distal pouch or by passing a bougie (Hegar dilator or urethral sound) through the gastrostomy site into the distal esophagus. With a radio-opaque catheter in the proximal esophagus it is now possible to measure, in terms of vertebral body heights, the gap between the two ends. Where the gap is of three to six vertebra, delayed primary repair should be planned over a period of up to 12 weeks.

When the gap between the two ends is greater than six vertebrae, the options are to proceed as above and attempt delayed primary repair, to apply graduated tension on the esophageal ends over a period of 6-10 days and then perform a primary anastomosis (29), or to abandon any attempt to retain the esophagus and to perform a cervical esophagostomy and replace the esophagus at a later date.

Management of the H-type fistula

An H-type tracheoesophageal fistula is suspected when the infant experiences coughing with feeds or suffers from recurrent respiratory infections. The diagnosis is established on contrast esophagogram (ideally a tube esophagogram) and confirmed at bronchoscopy/esophagoscopy. It is extremely valuable to have a ureteric catheter passed across the fistula at preliminary bronchoscopy immediately prior to surgery. The operative division of the H fistula is most frequently performed via a low cervical approach.

Postperative complications

In our series sepsis and pulmonary complications still plays a major problem, the other complications are anastomotic leaks which occur in 15-20% (30). The major leaks occur in the early postoperative period (<48 hours) and present with life-threatening tension pneumothorax. Minor leaks may be detected on the "routine" contrast study usually performed on the 5th-7th day post-operatively. These will all seal spontaneously but there is an increased incidence of later stricture formation.

Anastomotic strictures develop in 30-40% (31) of cases, most of which will respond to one or two dilations. Risk factors which have been implicated in stricture formation include anastomotic tension, anastomotic leakage and gastroesophageal reflux. Oblique end-to-end anastomosis lowers the incidence of stricture formation. It was only 1.4% in our series. Endoscopic dilatation of the stricture can be carried out either at rigid esophagoscopy using semi-rigid bougies or by balloon dilatation introduced either at fluoroscopy or during flexible endoscopy.

Recurrent tracheoesophageal fistula:

The incidence of recurrent tracheoesophageal fistula is between 5-14% (32). A recurrent fistula should be suspected if the infant manifests respiratory symptoms (coughing during feeds, apnoeic or cyanotic episodes) or has recurrent respiratory infections after

"successful" repair of the esophageal atresia. Urgent investigations must be undertaken to positively diagnose or exclude the possibility of a recurrent fistula.

At surgery it is useful to define the esophagus above and below the site of the fistula and to insert stay sutures at both the esophageal and tracheal ends of the fistula before it is divided. The resulting defects in the trachea and esophagus are closed with non-absorbable interrupted full-thickness sutures.

Esophageal Replacement:

The need to replace the esophagus in esophageal atresia is extremely rare and should only be considered in very long-gap situations or where repeated attempts at retaining the host esophagus have failed and the infant's survival is at risk. There are basically three methods of esophageal replacement currently being practiced in children – gastric transposition (33), colonic interposition (34) and jejunal interposition (35). Each have merits and carry complications. Gastric transposition has more recently become a well established & successful substitute in infant with esophageal atresia.

All the problems of EA management in developing countries can be summarized under two broad categories – lack of awareness and lack of facilities. While lack of awareness leads mainly to delayed presentation and consequent complications, the inadequacy of facilities makes our setup vitally different from that of our counterparts in developed countries. For example – associated cardiac anomaly is regarded an important factor for poor prognosis in our setup. Limited facilities for carrying out pediatric cardiac surgery precludes the scope of improvement of survival rate in such cases. Poverty, illiteracy and ignorance lead to lack of awareness and delay in seeking medical advice. Nearly 95% of the deliveries in the rural and around 50% in urban areas are being conducted at home, mostly by untrained traditional birth attendants (TBAs) who are not even aware of this anomaly (EA). Concerted efforts through government and NGOs to train these TBAs are improving the scenario. The integrated child development services and better diagnostic, transport facilities and awareness with more specialized centers will ultimately improve the overall scenario.

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