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EARLY AND MIDTERM FOLLOW UP AFTER EARLY TOTAL SURGICAL REPAIR OF ESOPHAGEAL ATRESIA WITH TRACHEO- ESOPHAGEAL FISTULA

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ABSTRACT

Background / Study Objective: to asses and present our short and intermediate outcome i early repair of Esophageal Atresia with Tracheo-Esophageal Fistula

Patients: 24 patients diagnosed as EA with TEF treated at our hospital (Zagazig University Hospital, collected over a period of 4 years from Jan 2012 tell October 2016

Objectives: is to evaluate the short and long outcome of surgical repair of Tracheoesophageal fistula

Methods: From Jan 2012 tell October 2016, All cases diagnosed as EA with TEF treated at our hospital where included in this study. The follow up period was 2 year in average, age, sex, weight at birth, associated anomalies, surgical approach, postoperative complications and outcome.

Results: 24 newborns (16 males and 8 females) with EA/TEF were treated at our hospital with an average weight 2.1 kg (700 g to 3200 g). Aged from birth to 3 days. At the time of admission 1 (4.16 %) had aspiration pneumonia. Early esophago-esophageal anastomosis was decided for all cases

Conclusion: Factors contributing to mortality included prematurity, low birth weight, presence of associated serious anomalies, and delay in diagnosis or aspiration pneumonia.

INTRODUCTION

Esophageal atresia (E A) is a congenital abnormality in which the midportion of the esophagus is absent. Its live birth incidence was estimated to be between 1 in 3,570 and 1 in 4,500. Distal Tracheoesophageal fistula was found in most patients [(1, 2).]

EA appears to result from aberrations of differential growth rate, cellular differentiation, and apoptosis. The rate and timing of cell proliferation and differentiation can be altered by one or more factors. The apoptosis in the region of the esophagus as well as the developing lung bud can also affected before 34 days' gestation [(1).]

Esophageal atresia & Tracheo-esophageal fistula EA &TOF are correctable surgically, with generally good results. Cameron Haight in 1941 was the first to gain successful primary repair of an esophageal atresia with increased mortality in the following decades [(2).] The final outcomes were influenced by birth weight, additional malformations, and aspiration pneumonia caused by delayed diagnosis. The basic surgical management for most common type of EA with a lower fistula is uniform while babies with long-gap EA surgical strategy are controversial [(3).]

Maternal polyhydramnios was presented in about 35% of patients with a distal fistula, but also found in 95% of patients without distal fistula. Prematurity is also common. More than one-half of the patients with EA have other major congenital anomalies, of which congenital heart disease, urinary tract anomalies, and gastrointestinal tract abnormalities are the most common [(1,4).]

The aim of this prospective study is to evaluate the short and long outcome of surgical repair of Tracheoesophageal fistula from Jan 2012 tell October 2016 in our center with focus on complications following repair and to assess our surgical outcome for the last 4 years to in comparison with the international results.

PATIENTS AND METHODS

Our study included 24 patients diagnosed as EA with TEF treated at our hospital (Zagazig University Hospital, collected over a period of 4 years from Jan 2012 tell October 2016. followed for 2 years in average , age at diagnosis, sex, birth weight, associated anomalies, surgical approach, postoperative complications and outcome.

Diagnosis was suspected in some cases by prenatal ultrasound, and confirmed postnatally by clinical examination and (Chest x-ray, abdominal U/S, Echocardiography "ECHO ", lab study), (EA was suspected on antenatal ultrasonography when maternal polyhydramnios, a small stomach, a distended upper esophageal pouch, or abnormal swallowing is observed).

Any excessively drooling infant was assumed to have EA until proven otherwise. The diagnosis is made when a stiff 10-gauge French catheter introduced through the mouth, becomes arrested at about 10 cm from the gums. Failure to pass the catheter into the stomach confirms the diagnosis of EA; all cases were diagnosed within the first 36 hours postnatally (All patients proved to have distal trachea-esophageal fistulae with proximal EA “Gross classification C, Vogt classification 3B"). After proper pre-operative preparation and stabilization, repair was done for all cases in early infancy (within the first 72 hours) if applicable (pt sometimes came late referred from other hospital) with division of the trachea-esophageal fistulae and primary esophageal anastomosis using single layer of interrupted 5/0 &6/0 vikryl suture via standard postero-lateral thoracotomy through the 4th intercostal space intra-pleural approach. All patients were managed post-operatively in the neonatal intensive care unit. Intravenous fluids were administered together with trophic nasogastric tube feeding and prophylactic broad spectrum antibiotic were continued until Para-anastomotic intercostal tube drain was removed. Regular gentle naso-esophageal suction was done as required. Esophageal anastomosis was performed under tension in two patients.

Postoperative upper GI contrast study after esophageal repair is preferable to asses any narrowing. Esophagogram can be used to demonstrate postoperative complications such as leak, obstructive strictures, fistulous recurrence and gastro-esophageal reflux Barium makes a good visualization, but extra-luminal barium fibrous mediastinitis. Aqueous low-osmolality agents are preferred. They have no harmful effects on gastrointestinal tract, and are not absorbed. However, aqueous contrast materials have a decreased coating ability so they are diluted quickly and less sensitive; therefore up to 25% of thoracic perforations and 50% of cervical perforations may not be detected. Hyperosmolar agents are contraindicated because their aspiration causes pulmonary edema. Hypovolemia may result also as a result of intravascular fluids displacement into the gastrointestinal tract, severe dehydration, and marked tracheal and bronchial irritation.

The trans-anastomotic tube was removed after confirmation of anastomosis integrity with contrast study on the 7th post-operative day to rule out anastomotic leak before starting free oral feeding .after discharge patients was regularly assessed in the outpatient clinic

Statistical data analysis was performed with Student's t-test or Chi2 test, where appropriate. Statistical significance was determined at $P < 0.05$

RESULTS

24 newborns (16 males and 8 females) with EA/TEF were treated at our hospital. Their mean weight at birth was 2.1 kg (700 g to 3200 g). Age at diagnosis ranged from birth to 3 days. At the time of admission 1 (4.16 %) had aspiration pneumonia. Early esophago-esophageal anastomosis was decided for all cases, as there was no need for staged procedure for cases with long gap EA ,as we have only two cases with a gap more than 3 cm length and esophageal anastomosis was performed under tension for them . Associated anomalies were seen in 4 (16.66 %) patients ***(Table I).***

Post-operative complications and mortality:

Postoperative complications were comparable to other reports and overall operative mortality was (8.33 %).

Anastomotic strictures occurred in two of 24 patients (8.33 %), (stricture was defined as an anastomotic narrowing on an upper gastrointestinal film that required two dilations.

Anastomotic leaks (which were minor) were suspected if oral content was found in the chest drain , and confirmed by upper gastrointestinal series, the leakage rate was (4.16%), and this leak occurred for the same patient with stricture discovered later on during post-operative follow up and was managed successfully by dilation.

Gastro esophageal reflux was diagnosed by upper gastrointestinal endoscopy or pH probe in 3 patients, and was diagnosed between one month and two years post-operatively, all patients were treated medically.

The in hospital mortality was 2 of 24 patients (8.33 %) who underwent definitive surgical repair, one died immediately after surgery due to severe sepsis and cardio-pulmonary failure, and the other one died one year after surgery due to associated cardiac anomaly (ASD) ***(Table II).***

Survival rate in our study was compared with the published international survival rate using (Waterston prognostic classification), revealing non significant statistical difference except for group C in which the difference was significant (P =0.004), and this is shown in ***Table (III).*** Also our survival rate compared with the international published survival rate using (Spitz prognostic classification), revealing non significant statistical difference in all groups as shown in ***Table (IV).***

Four patients died before any surgical intervention and were not included in our study, those patients known to have severe associated anomalies.

Table I Preoperative data and operative approaches.

Total No. of patient	24 (100%)
Type of anomaly (EA &TOF	24 (100%)
Gender	16 boys (66.66%) and 8 girls (33.33%)
Associated anomalies	2 cardiac (8.3%) and 2 GIT (8.3%)
Gestational age	36.8 weeks (Range 34 to 38)
Birth weight	2.1 kg (Range 0.6 kg to 3.5 kg)
Esophageal gap	< 3cm (22 cases 91.66%) 3 to 5 cm (2 cases 8.33%)
Esophago-esophageal anastomoses	24 (100%)

Table II Postoperative complications and mortality.

	Number	%
Gastroesophageal reflux	3	12.5
Leakage	1	4.16%
Pneumonia	2	8.33
Stricture	2	8.33
Fistula recurrence	None	Zero
Redo	None	Zero
Mortality	2	8.33

Table III Comparison of our Survival rate with Waterston Prognostic Classification.

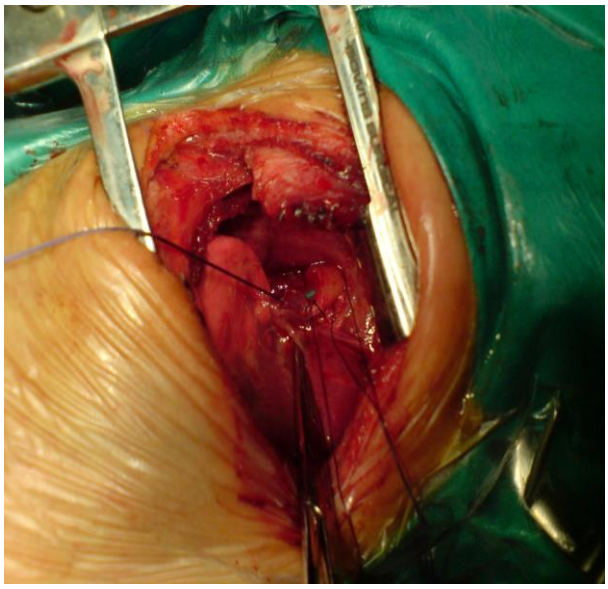
Waterston Prognostic Classification	Published SR	Our study SR
Birth weight > 2.500 kg and well	95 %	100 %
Birth wt. 1.800 to 2.500 kg and well or birth wt. > 2.500 kg but moderate pneumonia and other congenital anomalies	68 %	50 %
Birth wt. < 1.800 kg with severe pneumonia and severe congenital anomaly	6 %	33.33 %

1: P = 0.89 NS, ***2: P*** = 0.97 NS, ***3: P*** = 0.004 Significant

Table IV Comparison of our Survival rate with Spitz Prognostic Classification

Spitz Prognostic Classification	Published SR	Our study SR
I-Birth Weight > 1.500 kg without major cardiac disease	97 %	100 %
II-Birth wt. < 1.500 kg ,or major cardiac disease	59 %	50 %
III-Birth wt. < 1.500 kg and major cardiac disease	22 %	50 %

1: P = 0.81 (NS), ***2: P*** = 0.76 (NS), ***3: P*** = 0.49 (NS)



Surgical view

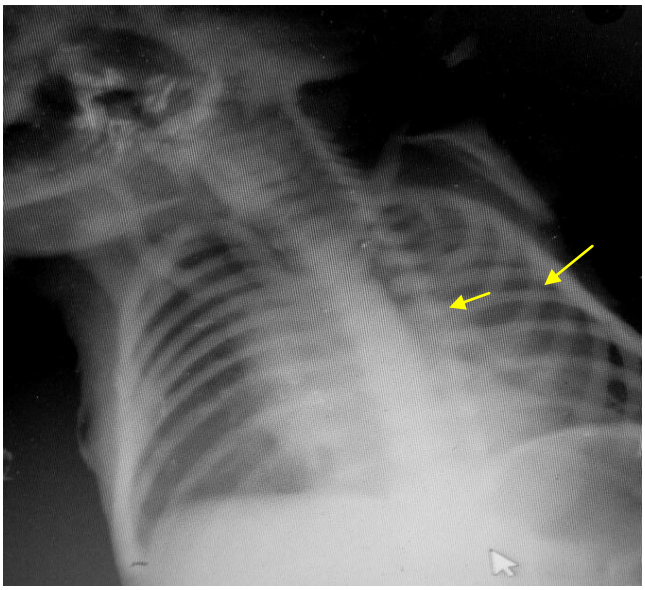
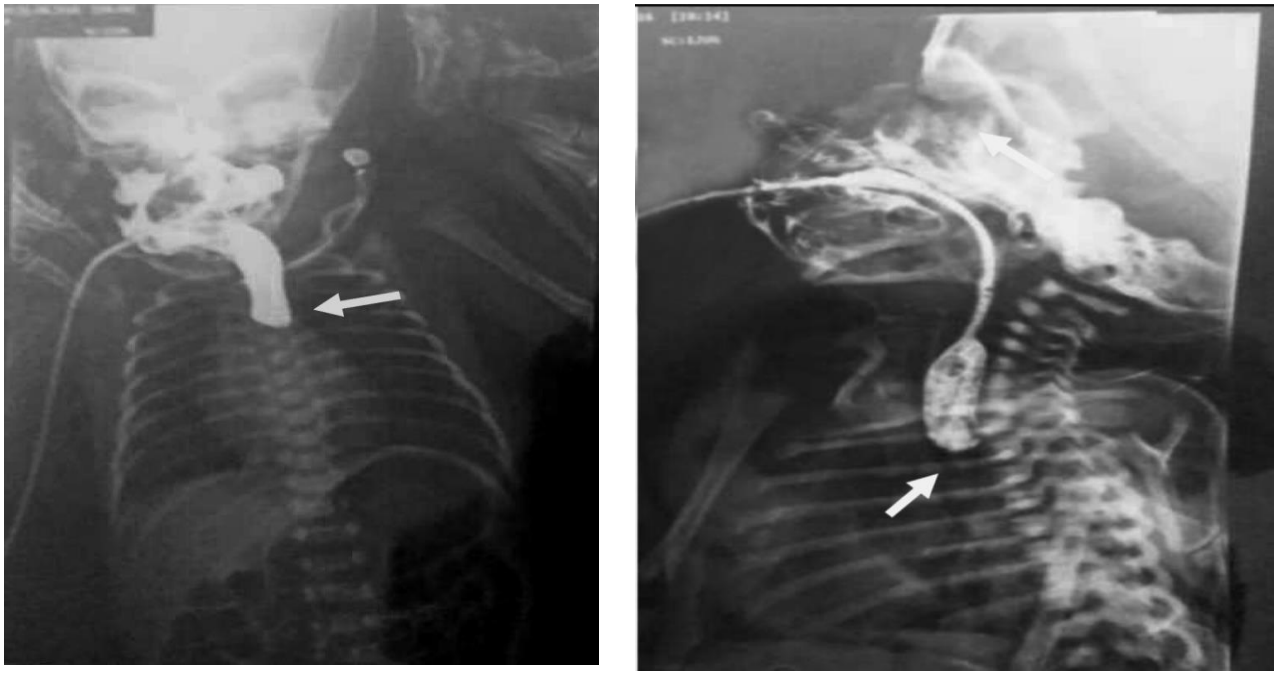
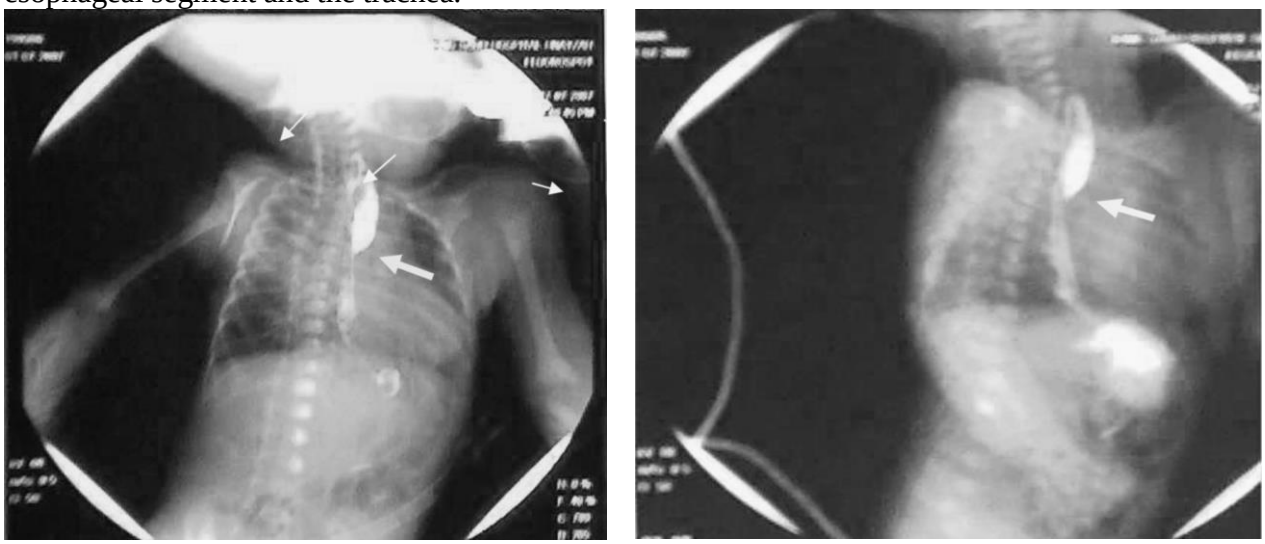


Fig. 1 Preoperative frontal radiograph of a case of OA with distal fistula shows a radioopaque tube (yellow arrows) coiled into the upper esophageal pouch. Air is present into the stomach indicating a distal fistula between the lower esophageal segment and the trachea.



A B

Fig.2 (A) Frontal & (B) Lateral preoperative views of upper GI contrast study of a case of OA with distal fistula shows arrest of contrast in the esophagus at the junction of its proximal and middle thirds(white arrows). Gas within the abdomen implies a fistulous communication between the lower esophageal segment and the trachea.



A B

Fig 3 Postoperative upper GI contrast study two months after operation show a short narrow segment of anastomotic stricture at the site of prior surgical anastomosis (white arrows) with proximal pouch dilatation of the esophagus.

DISCUSSION

In cases of suspected neonatal EA, frontal and lateral plain x-ray of the chest is indicated. Radiographic proof of a nasogastric tube failure to pass to the stomach is diagnostic of EA [24]

Brown et al stated that the incidence of first degree relatives having one or more components of the VACTERL association was 5.5 % in the EA [4].

Even though many system of casement was introduced [**The Waterston prognostic classification for survival of neonates**] with EA was proposed in 1962 and still widely used in many pediatric surgery centers [5, 6, 7].

The original survival rate (SR) in his 3 categories were 95 %, 68 %, and 6 %, whereas

the survival rate in this study in same 3 categories were 100 % , 50 % , and 33.33 %, respectively [8]. Our survival rate is better in class C as P = 0.004 which is significant, Table (III). We can attribute this difference which is significant to early diagnosis as well as, to the improvement in surgical techniques. The proper anesthetic management, perioperative management of those very low birth weight newborn and the proper management of pneumonia in neonatal intensive care unit (NICU) are also important factors.

The previously used Waterston classification has little relevance today in predicting outcome of neonates with EA as mentioned by other authors [9].

In 1994 Spitz [6], reported about that the prognostic classification and the original survival rate in his case categories were 97 %, 59 %, and 22 %, whereas in our work, the survival rate in comparison to the same categories of Spitz classification were 100 %, 50 %, and 50 %, respectively, Table (IV).

Our survival rate where comparable to published survival rate in all classes of Spitz series

The diagnosis nowadays can be established immediately after birth in most cases, and continuous suction of the upper pouch can prevent pneumonia. Survival of premature infants has significantly improved with progress in neonatal intensive care [3].

In 2006 Spitz [5] reported 10 % overall incidence of esophageal leak in early anastomosis which were mostly minor and sealed spontaneously. The strictures at the anastomotic site were recognized in 37% of cases, most also were minor and responded well to one or two dilations [10][11]. In our study, 1 (4.16%) case developed minimal leakage postoperatively which healed spontaneously by conservative treatment, and 2 (8.33%), cases developed stricture and was defined in our study as an anastomotic narrowing seen on upper gastrointestinal series , this case was treated by two settings of balloon dilations , and this was less than reported in other series (10). We suppose that the difference between our results and the previously mentioned studies may be attributed to the difference in (number of cases included in each study, gap length, suture material used and associated anomalies)

In the report of Spitz published in 2006, he mentioned that reflux occurs in 40 % of infants after repair of EA , half of them required anti-reflux surgery after failure of intensive anti-reflux therapy [3][12]. Another large series for Spitz in 1993, for 303 patients, he noted a 52 % rate of reflux in 87 patients with anastomotic stricture (14.5 % reflux rate from the total number of patients) [13][14]. In our study 3 (12.5 %) cases developed gastro-esophageal reflux, and all were treated conservatively. The true incidence of reflux in this study may be higher. This disparity can be awed to the difficulty in proper documentation and confirmation of the presence of GER postoperatively during follow up visits.

Although early repair is the favored method of treatment for most infant, a staged approach may be warranted hemodynamic instability, marked prematurity , and long gap EA [15][16]. The staged approach was consisted of feeding gastrostomy added to cervical esophagostomy in the neonatal period. However, the terminal esophagostomy leads to the need for an esophageal replacement procedure using the colon or the stomach in most cases. To preserve the native esophagus the alternative methods as lengthening procedures [14][17], or the lateral esophagostomy in the proximal pouch have been described [18].

Many surgeons prefer the delayed repair procedure after several months. The definitive surgery is delayed until the gap narrows enough to facilitate esophageo-esophageal anastomosis [19]. Prematurity or other complicating factors were the causes for delayed repair in another study [2]. Many authors favor primary esophago-esophageal anastomoses as compared with esophageal replacement, as they mentioned better results with primary esophago-esophageal anastomoses.

In our study, early esophago-esophageal anastomosis was performed for all cases, even for the two cases with long gap. One of the two patients with long gap died immediately post-operatively due to associated anomalies. The other patient with long gap and early esophago-esophageal anastomoses developed

post-operative leaks, and was successfully managed conservatively [19].

There is a group of infants with EA with lethal congenital anomalies for whom surgery would be of no benefit. Those include major cerebral anomalies , grade IV intraventricular hemorrhage , pulmonary atresia , trisomy 18 , and complex CHARGE (colorectal, heart anomaly , retardation , genital anomaly , and ear anomaly) syndrome [6][18]. These infants should not be included in survival statistics.

In our study, we excluded extremely low birth weight babies passed away within 6 hours after birth due to severe respiratory distress and the associated lethal cardiac anomalies, in spite of neonatal intensive care management.

The improved survival was the result of the advances in diagnosis of congenital anomalies early, and also the advances in the management of the high risk neonates in NICU, including the proper management of respiratory problems, improved neonatal transport systems and nutritional support [20]. The care in pediatric anesthesia has also was improved in the last period with accurate management of low birth weight patients, better monitoring capacity, and improved equipment to prevent hypothermia [20][21]. Accumulated surgical experience, the improvement in suture materials (we currently favor the use of prolene sutures), use of the finest instruments and the use of magnification tools have contributed to the improved outcomes of EA with trachea-esophageal fistula repair [22]. The thoracoscopic approach for EA repair might bring further improvement in the outcomes for these patients [23].

CONCLUSION

Prematurity, low birth weight, presence of associated serious anomalies are factors which can contribute to mortality. Also, delayed diagnosis, increased incidence of aspiration pneumonia, and a shortage of qualified nurses were considered.

The advances in neonatal intensive care with special attention to good ventilation and nutritional support can facilitate proper pre-operative preparation and treatment of associated anomalies as well as the advances in pediatric anesthesia, and refinements in surgical techniques have contributed to improved outcomes of EA.

Evaluation and overcoming the factors that lead to sepsis helps to improve the overall outcome,

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