Research Article

Determining the factors that predict the feasibility of one-stage primary esophageal repair versus staged repair in neonates with type C esophageal atresia: Developing some new criteria

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Abstract

Background: Several factors can predict the feasibility of primary repair or a staged repair should be done from the start in patients with esophageal atresia with distal tracheoesophageal fistula (Type C TEF) to avoid unnecessary lengthy operations which may result in high morbidity and mortality in such condition.

The aim of our work: The aim of our work was the assessment of the factors which can predict the feasibility of primary repair in case of type C TEF.

Material & methods: Our study is a case series study that included all patients who were admitted to our institute having type C TEF from June 2016 till June 2017. Patients with bad general or respiratory conditions were excluded from the start of our study. The age at operation, sex, birth weight, gestational age, associated congenital anomalies, and the findings in chest X-ray (CXR), as well as the intra-operative gap length, were recorded. The relation of the final surgical outcome with the previously recorded parameters was statistically tested.

Results: The study included 128 patients; from whom 84 patients had a primary esophageal repair and 44 patients had a staged repair in the form of esophagostomy and gastrostomy. The factors which predicted significantly the feasibility of primary repair were the gestational age (^{FE}p=0.02), the birth weight (p<0.001), the relation of the upper esophageal pouch to T3 (3rd thoracic vertebra) (^{FE}p<0.001*), and the measured intra-operative gap length (^{FE}p<0.001).

Conclusions: Patients having type C TEF who are full-term, having an average birth weight, having an upper esophageal pouch at or below the level of T3, and patients with gap length ≤3 cm as measured intra-operative have higher feasibility of primary esophageal repair.

Background

Patients who are presented with esophageal atresia with distal tracheoesophageal fistula (Type C TEF) may have a short or a long gap between the two esophageal pouches. Those patients with short gaps are usually repaired by primary anastomosis which is considered the optimum surgical option. On the other hand, patients having long gaps are considered challenging as they are usually treated in a staged procedure starting by ligation of the fistula in association with an initial feeding gastrostomy with or without cervical esophagostomy [1]. Cervical esophagostomy is done by some surgeons to allow sham feeding, to provide an exit for saliva, to allow the elongation of the proximal esophageal pouch, and to avoid the development of feeding difficulties if oral feeding was not attempted early in those patients. This is followed by esophageal replacement [1].

The case in which the surgeon decided not to do cervical esophagostomy is managed by the closure of the fistula with the construction of a feeding gastrostomy leaving the upper esophageal pouch intact with continuous suction by a Replogle tube allowing its spontaneous growth aiming at performing delayed primary repair later on [2].

Preoperative bronchoscopy has been described for the 1st time in the assessment of neonates with esophageal atresia in 1981 [3]. This procedure could help in determining the length of the gap between the two esophageal pouches by measuring the distance between the entrance of the fistula into the carina –the distal pouch- and the proximal pouch which can be suspected by the observation of the external compression of the pars membranacea. Also, a water-soluble inflated Fogarty catheter could be inserted into the fistula allowing for better assessment of the gap length as well as the intraoperative detection of the fistula [3]. Unfortunately, the unavailability of preoperative bronchoscopy in many centers especially in the developing countries makes its usage un-practical. Also, the benefit of this procedure is still debated [3].

Consequently; the gap length is usually measured intraoperative after the division of the fistula and then the decision to do either primary esophageal anastomosis or staged repair is made. Wrong decisions with failed primary anastomosis would result in a secondary long gap which results in increased morbidity and mortality rates due to repeated surgeries in such fragile neonates [4].

Aim of the work

Our study aimed to assess the factors that can predict the feasibility of primary repair in the case of Type C TEF.

Material & methods

Our study is a case series study that was carried out during one year period from June 2016 to June 2017 and included all neonates having type C TEF. The pre-operative data including the age at operation, the sex, the gestational age, associated anomalies, and the birth weight were recorded from the hospital records. The data integrity was maintained through these records. Any patient with pre-operative respiratory distress as well as having bad general condition was excluded from our study to assess only the anatomical and technical factors that might affect the achievement of a primary esophageal repair excluding any confusing factor of respiratory distress which usually needs staged procedure from the start. The presence of any associated congenital anomaly was also recorded. The diagnosis of the anomaly was confirmed by doing CXR while inserting an eight French nasogastric tube (NGT) in the upper esophageal pouch and the relation of the tip of this tube to the 3rd thoracic vertebra T3 was recorded.

All of the studied patients were explored by right posterolateral thoracotomy at the level of the right 4th intercostal space using an extra-pleural approach (after exclusion of right-sided aortic arch by pre-operative echo as a routine investigation). The fistula was then carefully dissected, circumferentially mobilized, and transfixed-ligated using Vicryl 4/0. The fistula was then transected and the proximal pouch was mobilized caudally to the distal pouch. The distance between the un-stretched fully mobilized proximal pouch and the distal pouch was measured using Vernier Caliper [5]. Then opposing traction sutures were applied to achieve an approximation of the two pouches.

At this point; the decision was taken: if primary anastomosis was feasible then an esophago-esophageal full-thickness end-to-end anastomosis was done using interrupted 5/0 Vicryl sutures over a trans-anastomotic eight French NGT reaching the stomach. This feasibility was defined as a possible anastomosis even under some tension. However, in cases in which the primary repair was not feasible; the distal pouch was ligated together with an open gastrostomy which was done through a supra-umbilical incision using a ten French, self-retaining tube being inserted into the anterior gastric wall near the lesser curvature. Also, cervical esophagostomy was done through a left transverse supraclavicular incision and secured to the skin. Once the procedure was completed, a draining chest tube was placed and secured to the chest wall. The chest wall was closed in layers with the appropriate suture materials. The surgical outcome whether primary or staged repair (esophagostomy and gastrostomy) was recorded and according to this outcome; the patients were divided into two groups; group A in whom a primary repair was feasible and group B in whom a staged repair was done. The two groups were compared according to the personal data, preoperative clinical and radiological findings, and the distance between the two esophageal pouches as measured intra-operative.

Statistical analysis

Data were fed to the computer and analyzed using IBM SPSS software package version 20.0. (Armonk, NY: IBM Corp) Qualitative data were described using the number and percent [6]. The Kolmogorov-Smirnov test was used to verify the normality of distribution Quantitative data were described using range (minimum and maximum), mean, standard deviation, and median. The significance of the obtained results was judged at the 5% level ($p \leq 0.05$).

The used tests were the Chi-square test (For categorical variables, to compare between different groups), the Fisher's Exact or Monte Carlo correction (Correction for chi-square when more than 20% of the cells have expected to count less than 5), the Student t-test (For normally distributed quantitative variables, to compare between two studied groups) and the Mann

Whitney test (For abnormally distributed quantitative variables, to compare between two studied groups).

Results

Our study included 128 patients diagnosed as having type C TEF; 84 patients (65.5%) had a primary repair and enrolled in group A and 44 patients (34.4%) had a staged repair and were included in group B. All of them had an average general and respiratory conditions on room air. There were 76 males (59.4%) and 52 females (40.6%) with a male: female ratio of 1.4:1. The difference in sex distribution between the two studied groups did not show statistical significance as shown in (Table 1). (Fisher exact, FEp=0.72).

Table 1: The difference in sex distribution between the two studied groups.

Gender	Total (No. = 128, 100%)		Group (A) (No. = 84, 65.5%)		Group (B) (No. = 44, 34.4%)		FEp
	No.	%	No.	%	No.	%	
Male	76	59.4	52	61.9	24	54.5	0.721
Female	52	40.6	32	38.1	20	45.5	

FEp: p-value of Fisher exact test

The gestational age varied between the two studied groups significantly as there were 84 full-term neonates (65.6%) and 44 preterm neonates (34.4%) with a higher percentage of full-term neonates in group A (68 patients; 81%) than in group B (16 patients; 36.4%) as shown in table 2. (Fisher exact, FEp=0.02*).

Table 2: Comparison of the two studied groups according to the gestational age (weeks).

Gestational age	Total (No. = 128, 100%)		Group (A) (No. = 84, 65.5%)		Group (B) (No. = 44, 34.4%)		FEp
	No.	%	No.	%	No.	%	
Full-term (> 37 week)	44	34.4	16	19.0	27	63.6	0.020*
Female	84	65.6	68	81.0	16	36.4	

FEp: p-value of Fisher exact test

The age at operation ranged between three and 33 days old with a mean of 14.91 ± 13.91 days. The age at operation was slightly higher in patients of group B than in patients of group A without showing any statistical significance as shown in table 3. (Mann Whitney, Z=98.0, p=0.506)

Table 3: The difference between the two studied groups according to the age at operation (days).

Age at operation (days)	Total (No. = 128, 100%)	Group (A) (No. = 84, 65.5%)	Group (B) (No. = 44, 34.4%)	Z	P
Min. – Max.	3.0 – 33.0	3.0 – 17.0	3.0 – 33.0	98.0	0.506
Mean ± SD.	14.91 ± 13.91	8.29 ± 5.17	14.91 ± 13.91		
Median	4.0	6.0	4.0		

P: p-value for comparing the two studied groups

The birth weight of the studied patients ranged between 1100 and 3600 gm with a mean of 2612.19 ± 630.4 gm. Patients of group B had a lower birth weight than patients of group A; this difference was statistically significant as shown in table 4. (Student t-test, T=4.472, p<0.001)

Table 4: The difference in birth weight between the two studied groups.

Birth weight (gm)	Total (No. = 128, 100%)	Group (A) (No. = 84, 65.5%)	Group (B) (No. = 44, 34.4%)	Z	P
Min. – Max.	1100 – 3600	2300 – 3600	1100 – 3000	98.0	0.506
Mean ± SD.	2612.19 ± 630.4	2896.19 ± 406.76	2070 ± 638.98		
Median	2700	2770	2100		

P: p-value for comparing the two studied groups

Associated congenital anomalies were present in about 28.1% of the studied patients (36 patients) with a higher percentage of patients having associated congenital anomalies managed by staged procedure (Group B). The difference in the incidence of such anomalies between the two studied groups did not show statistical significance as shown in table 5. (Fisher Exact, FEp =0.68)

Table 5: The difference in the incidence of associated congenital anomalies between the two studied groups.

Associated congenital anomalies	Total (No. = 128, 100%)		Group (A) (No. = 84, 65.5%)		Group (B) (No. = 44, 34.4%)		FEp
	No.	%	No.	%	No.	%	
Absent	92	71.9	68	80.95	24	54.56	0.681
Present	36	28.1	16	19.05	20	45.44	

FEp: p-value of Fisher exact test

The relationship of the upper esophageal pouch to the 3rd thoracic vertebra (T3) was assessed in the pre-operative CXR. This revealed that the upper pouch was below the level of T3 in about 80 patients (60%). A statistically significant higher incidence of patients with an upper esophageal pouch lower than the 3rd thoracic vertebra was noticed in group A (72 patients, 85.7%) than in group B (8 patients, 18.2%) as shown in table 6. (Fisher Exact test, FEp<0.001*)

Table 6: The relation of the upper esophageal pouch to the 3rd thoracic vertebra (T3) in CXR as compared between the two studied groups.

CXR	Total (No. = 128, 100%)		Group (A) (No. = 84, 65.5%)		Group (B) (No. = 44, 34.4%)		FEp
	No.	%	No.	%	No.	%	
Relation of the upper pouch to T3							
Above T3	48	37.5	12	14.3	36	81.8	<0.001*
Below T3	80	62.5	72	85.7	8	18.2	

FEp: p-value of Fisher exact test

The length of the gap between the two esophageal pouches as measured intra-operatively by the Vernier caliber [5] was less than 3 cm in 84 patients (62.5 %); from whom 72 patients were in group A (85.7%) and 12 patients were in group B (14.3%). This difference was statistically significant as shown in table 7. (Fisher Exact, FEp<0.001)

Table 7: The difference in the intra-operative gap length between the two studied groups.

	Total (No.=128, 100%)		Group (A) (No.=84, 65.5%)		Group (B) (No.=44, 34.4%)		FEp
	No.	%	No.	%	No.	%	
Intra-operative gap length (cm)							
>3 cm	44	34.4	0	0.0	44	100.0	<0.001*
<3 cm	84	65.6	84	100.0	0	0.0	

FEp: p-value of Fisher exact test

Discussion

The management of type C TEF can impose a very challenging situation, especially in cases with a long gap with higher morbidity and mortality rates than in the case of a short gap. This makes the importance of the predictor factors which can guide a proper decision on whether to do primary or staged repair to avoid a wrong decision with a resulting better outcome. Retaining the native esophagus by primary repair is almost always the target of any pediatric surgeon in this anomaly. Staged repair may be needed in patients with long gap atresia starting by ligation of the fistula in association with the construction of a feeding gastrostomy while preserving the proximal esophagus for delayed primary repair. This is usually done in case of patient instability either respiratory or generally. However, a cervical esophagostomy may be added to gastrostomy in case of anatomical and technical difficulties like long gap atresia planning for future esophageal replacement [7]. We resorted to doing esophagostomy and gastrostomy in our study in those patients in whom the primary repair was not feasible as we planned for a further esophageal replacement to reduce the hospital stay.

Many factors can affect the decision of either doing primary or staged repair in the case of type C TEF. These parameters would help the surgeon to make the decision, but they are not absolute rules as the final decision can be done only either by pre-operative bronchoscopy-which may not be available in many centers- or during intra-operative exploration. The gestational age is one of these factors that showed a statistically significant difference among our studied patients; this may be attributed to either the incompleteness of respiratory development which imposes a more rapid staged procedure rather than a lengthy primary repair or due to the long gap length in preterm patients. The study of [2] showed the same findings and concluded that premature neonates with this anomaly are better managed with staged repair with a resulting improvement in morbidity and mortality.

Another factor that may be taken into consideration while taking this decision is the age at the operation which although it was lower in patients with staged repair in our study, did not show statistical significance. This could be explained by the fact that patients with this anomaly who present lately usually have aspiration pneumonia with a bad general condition; both of these factors impose a major anesthetic challenge and enforce the surgeon to do a staged procedure to shorten the operative time. This explanation was also concluded by [8] in their study. Patients with low birth weight have a statistically higher incidence rate of staged procedures in our study. This could be explained by the fact that low birth weight patients who presented with type C TEF usually have a long gap due to the immaturity of the two pouches as also concluded by [9] in their study in 2016. [10] proposed that staged repair of this anomaly in both very low birth weight and premature neonates resulted in a significantly lower rate of anastomotic complications with a resultant lower morbidity rate. Also, they concluded that this should be considered the preferred surgical approach in this age group of patients. Their surgical point of view was that the extensive dissection of the esophageal pouches to achieve primary repair in this group of patients may result in significantly higher anastomotic complications due to premature friable tissues with borderline vascularity and possible risk of ischemia compared to full-term neonates. With increasing birth weight and age, the gap length decreases, and esophageal tissue is better developed thus making primary anastomosis easier to be performed and also to be tension free. Similarly, [11] in 2020 demonstrated also that patients with low birth weight have a higher rate of complications in case of the primary repair and recommended staged repair in such cases.

On the other hand, other surgeons recommend primary repair with satisfactory results in low and very low birth weight neonates as concluded by [12] in 2006 in their study. Also, [13] in 2017 concluded in their study that primary repair could be conducted in most low birth weight patients and saved the staged repair to unstable patients only. So average birth weight should only raise the feasibility of primary repair in type C TEF and can't be considered as a solid rule for proceeding to staged repair. The difference in the incidence of the associated congenital anomalies among our studied patients did not show statistical significance although a higher percentage of those who had associated congenital anomalies were treated by the staged procedure. The decision of doing a staged procedure in such patients could be attributed to the general as well as the respiratory instability and not due to the anatomical or the technical problem. [12] advised in their study that patients with associated congenital anomalies especially cardiac and pulmonary anomalies were better managed with the staged procedure as in this condition the patient can tolerate only the short procedure of fistula ligation but cannot tolerate a lengthy primary esophageal anastomosis with the lungs being retracted. Other surgeons proposed that the presence of associated congenital anomalies in patients with type C TEF does not affect the decision whether to do one or stage repair and that primary repair should be attempted whenever possible. They also concluded that the presence of associated anomalies affects only the prognosis but not the surgical procedure [14].

Patients with pre-operative respiratory distress or any respiratory support were excluded from our study to exclude any physiological confusing factors as we assessed the anatomical and technical factors that might affect the feasibility of a primary esophageal repair in such cases. [15] Considered that severe pulmonary dysfunction with preoperative ventilator dependence had a prognostic influence only and had no effect on the type of surgery. They explained the fact that the ventilator-dependent patients were at risk of deterioration under the long duration of anesthesia so it was better to ligate the fistula only and to postpone the definitive repair to reduce morbidity and mortality.

Preoperative assessment of the gap length is very important and helps the surgeon to be well prepared for the operation. The assessment can give an almost accurate prediction if the primary repair would be feasible or not and this would help in reducing the operative risk, the postoperative morbidity, hospital stay, and mortality, especially in developing countries with limited neonatal care facilities [16].

A significant difference was found in our study between the relation of the upper esophageal pouch to the 3rd thoracic vertebra regarding whether to do a primary anastomosis or staged repair. [9] mentioned similar results in their study as when the NGT in the upper esophageal pouch was arrested at the level of T1 (the level of the clavicle) or T2; the gap between both esophageal pouches was long, and primary esophageal repair was not feasible. On the other hand, when the NGT was arrested at or below the level of T3 the gap was short or even no gap when the NGT was arrested at the level of T4 so the primary repair was feasible. The gap length between the two pouches in the case of type C TEF was defined by several authors. [9] classified those patients in their study according to the gap length into a long gap length of > 2.1 cm, an intermediate gap of 1-2 cm, and a short gap of < 1 cm. Also, the gap between the two pouches in such cases was measured intra-operative by [17] by vernier caliper and classified these patients into four groups, the 1st group has an ultralong gap >3.5 cm, the 2nd group has a long gap of 2.1–3.5 cm, the 3rd group has an intermediate gap of 1-2 cm and the last group with a short gap < 1 cm.

Our study revealed a significant relationship between the distance between the two pouches and the feasibility of primary repair. [18] concluded in their study that patients with type C TEF who had a distance of less than 3.5 cm between the two pouches had a better prognosis than those with a longer distance.

Conclusion

The feasibility of primary repair of type C TEF could be predicted preoperatively depending on the presence of certain factors like full-term patients, average birth weight, and, the presence of upper esophageal pouch at the level of T3 in CXR as well as a gap length less than 3 cm as measured intraoperative. However, all of these parameters are rough parameters and not absolute rules as a further study with a larger number should be taken to reach an absolute figure. Also, we recommend another study in which we plan delayed primary anastomosis after gaining some length.

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