Surgical Treatment Of Esophageal Atresia: Our Experience After 24 Years

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Abstract

Introduction: The aim of this study was to critically analyse and evaluate the clinico-pathological epidemiological profile and the treatment of results for different types of esophageal atresia treated in our institution.

Material and methods: Data were collected retrospectively for 76 newborn children diagnosed with esophageal atresia in our institution between 1st June 1990 and 30th May 2014.

Results: The average birth weight was 2.4 kg. The mean gestational age was 36.1 ± 3.2 weeks. Types A, B, C and E of the EA (Gross Classification) were respectively represented 7 (9.2%) 1 (1.3%), 62 (81.6%) and 6 (7.9%). Malformations were associated in 53.9% of cases (n=41). The mean time between birth and the surgical cure was 61 days (0-3650 d). In types A, B and C, the mean time between the birth and surgical cure was 2 days (0-8 d). A tracheobronchoscopy was performed on 42 patients. It allowed us to confirm the diagnosis of oesophageal atresia (41 cases) and to find other bronchial tracheal abnormalities (5 cases). Seventy-four newborns underwent surgical correction of the EA. The mean time of hospitalisation was 48.9 days. Complications were oesophageal stenosis (35.5%), gastro-esophageal reflux (67.1%) with 9.2% who underwent a fundoplication. The mortality rate was 11.8% (n=9), mainly due to associated malformations (heart diseases). The long-term follow up (≥5 years) was complete for 24 patients and the mean follow up was 4.9 years. Forty-seven (68%) patients were alive at a year of decline.

Conclusion: Our results confirm the findings in the literature. The evaluation of the airways preoperatively proved an essential diagnostic mean in our series.

INTRODUCTION

Esophageal atresia (EA), resulting from a defect of embryonic septum between the trachea and oesophagus, is the most common oesophageal congenital malformation, estimated at 2.44 per 10 000 live births [1].

The surgical management of EA is complex and in case of long gap esophageal atresia, the effectiveness of different surgical techniques and strategies remain inconclusive and questionable. The aim of this study was to make a critical analysis to assess clinico-pathological epidemiological profile and the treatment of results for different types of esophageal atresia treated in our institution.

PATIENTS AND METHODS

This is a retrospective study conducted in the Cliniques Universitaires Saint Luc. All children born between 1st June 1990 and 30th May 2014 presenting an esophageal atresia and managed at the paediatric surgery unit were identified. Data collection was made from reports of hospitalisations and accounts of various consultations services and the operating records of the children concerned. We have analysed data from 76 patients. The parameters studied were the type EA, the birth term, birth weight, any associated malformations, the period of surgery, the preoperative tracheoscopy, the type of surgery and postoperative complications. Respiratory complications: tracheomalacia, recanalization of the tracheoesophageal fistula (TEF), pneumonia, mediastinitis, digestive (oesophageal stricture, clinic and/or documented gastro esophageal reflux, dysphagia), have been noted and their treatment were sought. Statistically, the data analysis was performed using SPSS ® software. The significance used for the tests was 0.05.

RESULTS

Population characteristics

Of the 76 children, 41 (53.9%) were born with an associated congenital malformation. The VACTERL association with 3 or more defects were identified in 21 patients (27.6%). Among patients with an associated VACTERL syndrome, 4 have shown an intestinal atresia (3 duodenal and 1 colic). Five patients presented a CHARGE syndrome. Among the associated abnormalities, the most frequent were cardiac abnormalities in 19 cases (25%) of which 6 received a surgery. The most common type of atresia according to the Gross classification was type C (62 patients, 81.6%), followed by type A (8 patients, 10.5%) and type E (6 patients, 7.9%). (Table 1)

Diagnostic

All patients with type A and type C were diagnosed by the identification of the abutment or of the winding of the probe suction to the chest radiography. A tracheobronchoscopy was performed on 42 patients (55.2%). It allowed us to confirm the diagnosis on 34 patients with EA of Type C and 4 patients EA of Type A. On one patient, the tracheoscopy did not objectify the TEF (type A), TEF found intra-operatively and so ranking this atresia in Type B. This also allowed finding other abnormalities of the tracheobronchial tree: 2 cases of tracheomalacia, 2 cases with 3 main bronchi and one case associating a diverticulum of the trachea. The diagnostic of the tracheoesophageal fistula H was revealed on 2 patients by opacification of the esophagus and 4 with the tracheoscopy alone.

Surgical management

The response time surgery was 61 days (0- 3560 d) for all EA. For the EA of type A, B and C, the response time was 2 days with extremes of 0 and 8 days. The side of the thoracotomy was almost always straight with extrapleural sides except in 2 cases: 1 case of thoracoscopy and one case in which the aortic arch was on the right side. A measure of the gap (distance between distal oesophagus and proximal) was performed on 6 of 8 patients with EA of type A. (Figure 1)

Figure 1

Algorithm for the surgical management based on the type of OA

Most patients with EA of type C underwent a primary surgical repair (59 of 62 patients).

On one patient with an EA of type A, an extension device (anterior flap) was necessary to realise the termino terminal oesophageal anastomosis. The recharge through the transanastomotic probe was performed within the first 3 days post-operative in 73.2% of cases and orally in the first 7 days in 62.8%. An esophagus opacification – stomach – duodenum – preceded the oral supply in 83.3%. The average hospitalization time was 48.9 days (0-208 days).

Postoperative complications

The complications observed were an anastomotic stenosis (35.5%), a recurrent esotracheal fistula (2.6%), anastomotic leakage (2.6%) and gastro-esophageal reflux (67.1%) associated with an esophagitis (10.5%). Of the 27 patients whose eso-duodenal transit has diagnosed an anastomotic stenosis, 25 were primarily treated by esophageal dilatation by balloon and/or bougie. However, for a specific patient, a segmental resection of the esophageal stenosis with termino terminal anastomosis was observed.

Also, an esophageal perforation during a dilatation session by Savary bougie has been surgically treated by transversal suture. The characteristics of patients with or without an esophageal stenosis are represented in Table 2. The TEF recurrences were treated by surgical revision. The medical treatment of the gastroesophageal reflux disease (GERD) consisted of the administration of inhibitors through the proton pump, prokinetic, H2 antagonist receivers and antacid. A surgical treatment by Nissen fundoplication or Toupet was necessary in 9.2%. One patient had Barrett's oesophagus and 6 patients with scoliosis.

Mortality

The number of deaths in this study was 9 patients, which translates into a mortality rate of 11.8%, mainly caused by associated malformations. Two patients died before the surgical treatment of their oesophageal atresia. The characteristics of the deceased patients were summarized in Table 3.

Follow up

The long-term follow-up (≥ 5 years) was complete for 24 patients, and the main follow-up was 4.9 years. Forty-seven patients (68%) were alive at one year of follow up.

Table 1

Clinical characteristics of patients

Characteristics	Value
Case	76
Sex (M/F)	37/39
Gestational Age (wk)	36.1 ± 3.2
≥37	46 (60.5)
<37	30 (39.5)
Birth weight (g)	2.4 ± 0.7
≥ 2500	40 (52.6)
1500 – 2500	29 (38.2)
<1500	7 (9.2)
Prenatal diagnosis	9 (11.8)
Associated anomalies	41 (53.9)
Cardiac	19 (25)
Gastro intestinal	4 (5.3)
Genito urinary	10 (13.1)
Vertebral and member	18 (23.7)
Chromosomal	2 (2.6)
Other	8 (10.5)
Type OA pre-operative	
A	8 (10.5)
C	62 (81.6)
E	6 (7.9)

Table 2

Comparison of children with and without esophageal stenosis.

Characteristics		Stenosis	No stenosis	Odds ratio (IC 95%)	P
Sex	F	12	25	0.768 (0.29 – 1.97)	0.583
	M	15	24		
Prematurity < 37 wk	Yes	8	22	0.516 (0.19 – 1.40)	0.192
	No	19	27		
Birth weight	< 2500g	11	25	0.66 (0.25 – 1.70)	0.3903
	≥ 2500g	16	24		
Prenatal diagnosis	Yes	4	5	1.530 (0.37 - 6.25)	0.5515
	No	23	44		
Associated malformation	Yes	13	28	0.696 (0.27 – 1.78)	0.4214
	No	14	21		
Type EA	C	21	41	0.682 (0.20 – 2.22)	0.5257
	Others	6	8		
Delay of surgery	≤ 2 days	23	40	1.006 (0.26 – 3.80)	0.3129
	> 2 days	4	7		
Type of 1st intervention	TT Anastomosis	22	38	1.042 (0.30 – 3.50)	0.809
	Others	5	9		
Anastomotic fistula / recurrent fistula	Yes	3	1	5.75 (0.56 – 58.29)	0.0999
	No	24	46		
Gastroesophageal reflux	Yes	18	32	0.937 (0.34 – 2.56)	0.9
	No	9	15		

TT anastomosis: termino terminal esophago esophageal anastomosis

Table 3

Characteristics of deceased patients

	Term (Wk)	BW (g)	Type EA	Age Death (d)	Combined anomalies	Etiology
1	37	2900	C	225	No	Sepsis
2	32	1750	C	17	Yes	Congestive heart failure, HTAP
3	37	2400	A	328	Yes	Sepsis
4	34	1700	C	1	Yes	Severe skin aplasia
5	36	2880	C	3	No	Respiratory failure
6	36	2310	C	45	Yes	Congestive heart failure
7	30	1140	C	53	No	Regurgitation / severe discomfort
8	37	3000	C	59	Yes	Cardiac, leukaemia
9	38	1650	C	0	Yes	Polymalformative syndrome

BW: Birth weight

DISCUSSION

The survival rate of EA in developed countries has reached a plateau in the 1980's and now appears to be stable with a rate of 95% [2,3].

The great frequency of abnormalities associated with EA (50%) can affect the treatment and the future of these children. The most common of related malformations occurs at the cardiovascular level, followed by musculoskeletal malformations, anorectal and intestinal deformities, genitourinary malformations, abnormalities of the head and neck, mediastinal abnormalities and chromosomal abnormalities [4]. In our study, more than half of the cases (53.9%) had an associated malformation, rate comparable to the centre of reference for congenital and malformative disorders of the esophagus [5] with 25% of heart defects. Thus, preoperative echocardiography was performed on our patients to seek for cardiac malformation and identify the position of the aortic arch. However, the echocardiography and chest X-ray are considered unreliable in identifying a right aortic arch in the EA [6,7]. Thus, the echocardiography appears to be more required for answering questions of preoperative anaesthesia than at the conduct of the surgical management [8].

In preoperative assessment, a tracheobronchoscopy was performed on more than half of our patients in order to observe one or many TEF, examine the tracheobronchial tree and diagnose a tracheomalacia or respiratory abnormalities. Although supported by several authors [9,10], the use of the endoscopy in perinatal remains controversial. The evaluation of airways preoperatively proved to be an essential diagnostic mean in our series. The primary surgical repair of EA with TEF is the best option of treatment in the absence of severe malformation. [4] Despite a favourable anatomy (short space between oesophageal cul-de-sac) and a meticulous surgical technique with excellent post-operative management, some potentially early or late post-operative complications can occur. Early complications include: anastomotic leaks (minor or major leaks), anastomotic stenosis, recurrent TOF and esophageal motility disorders with an associated risk of aspiration [11]. Anastomotic leaks can be identified during routine check of the opacification of the esophagus - stomach - duodenum or when viewing inside the chest drain saliva according to the extent of fistula. A chest tube was consistently implemented in our study meanwhile the routine opacification was performed at the discretion of the surgeon (83.3%).

Each surgeon does not systematically establish a chest tube and realise the opacification before the power start. The survey of several paediatric surgical centres revealed that 69% were implementing a thoracic drain and 72% performed a routine opacification. [8]

The incidence of anastomotic stenosis widely varies and is the most common cause of surgery among children with EA and TEF. The development of an anastomotic stenosis after an OA surgery continues to be a frustrating complication. A minimal tension of the termino-terminal anastomosis of the esophagus appears as a protective factor. The GERD treatment is essential to minimize the formation of

stenosis. We found in our series 35.5% of stenosis. Most studies report a stenosis rate of between 37% and 52% [14,15]. However, a study quotes a high rate of 69% [16]. The necessity for a dilatation for an anastomotic stenosis was 80% of patients in a large series [15,17]. In our series, it was 92.5%. The balloon dilatation under endoscopic control was the most common technique.

GERD is extremely common in children with a history of EA and can affect between 40% and 65% of patients [18,19]. The presence of a significant GERD is generally considered to be due in part to an intrinsic impaired motor of the esophagus [20]. However, it is likely that GERD is exacerbated by surgical repair and gastrostomy, causing an alteration of the anatomy of the gastro-esophageal junction and the His angle [11]. Although the majority of these patients are medically treated by H2 receptor antagonists or inhibitors of the proton pump [21,22], about 28% of them will benefit from surgical correction of their reflux [23]. The prolonged GERD of these children may cause an esophagitis, which can have consequences in the short and long-term [24]. In addition to the esophagitis, GERD may cause the appearance of a Barrett's metaplasia, which is a risk factor for oesophageal adenocarcinoma [24]. However, only 6 cases of esophagus cancer after oesophageal atresia were reported in the literature. We found in our series, a GERD in 67.1%, associated with an esophagitis in 10.5%. A surgical treatment by fundoplication was necessary in 9.2%.

The TOF recurrence rate among patients was consistent with other reports [12,14], two children (2.6%) developed a major anastomotic leakage and benefited surgical treatment. However, other authors have reported rates between 6-17% [12, 25].

Many surgical strategies have been reported for child management with EA of type A [26,27]. For EA without TEF (EA of type A), most respondents would prefer to do a delayed anastomosis with the establishment of a gastrostomy without esophagostomy [8]. This approach is consistent with a recent meta-analysis that found good results in the long-term [28]. In our series, we try to conserve the native oesophagus. We believe in a cautious approach, with 3 months waiting period to allow the oesophageal growth in relation to the thoracic spine chest, to facilitate an anastomosis between both ends [29]. Others have suggested that this growth is secondary to the swallowing reflex in the upper cul-de-sac and of the GERD in the lower cul-de-sac [26].

There is still no consensus on the optimal surgical approach for the esophagus replacement. The conservation of the native esophagus in atresia malformations with a very large esophageal gap is usually not possible, and a "conduit" is necessary to restore the continuity of digestive paths. The optimum behaviour for the replacement of the esophagus is a much-debated topic in the paediatric surgery spheres [30-31]. The most used conduits are the stomach, the colon and the jejunum. According to some authors, the stomach is considered to be the best conduit because of its resistance to acidity, its ability to maintain tubular shape without expansion, and its ability to bridge the long gap due to a good vascularization [32]. We have done on our patients an ileocolic esophagoplasty or esocoloplasty. Both ileocolic plasties performed were retrosternal with good results at 10 years. The need to consider surgical options available to determine the optimal surgical approach within this complex group of patients (EA without TEF) suggests that this type of surgery should be performed in centres with a considerable experience in surgery of the esophagus. In each case, there should be an examination of the relative benefits and risks associated with the preservation of the intrinsically abnormal native esophagus [34].

CONCLUSION

In recent years, advances in surgery and intensive care have greatly improved the prognosis of this malformation. The results of our series confirm the results found in the literature. Today, most children born with an esophagus atresia enjoy a normal life, although any remaining complications can sometimes have an impact on their quality of life. In the last few decades, efforts are being focussed on preventing long-term complications, including orality disorders.

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