Original Article

Oesophageal Atresia without Major Cardiovascular Anomalies: Is Management Justified at a District Paediatric Surgical Institution?

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Abstract

Background: Oesophageal atresia lacks sufficiently documented treatment approach, as guidelines are based rather on the opinion of experts than on systematic data. We aimed to answer the question if treatment of patients without major cardiovascular anomalies could be justified at a peripheral paediatric surgical institution, by evaluating the outcome of surgical correction. **Methods:** Thirty-three neonates underwent surgery for correction of oesophageal atresia during a period of 20 years. They were categorised into two time-period groups, to follow-up the evolution of surgical intervention and complications through time. Evaluation of post-operative outcome and morbidity was performed. The results were related to those of our recent cross-sectional study on families having experienced oesophageal atresia performed years after repair, regarding the long-term quality of life. **Results:** A shift from staged to primary repair occurred throughout time in the patients with a marginal long gap between proximal and distal oesophagus ($P = 0.008$). Anastomotic stenosis was the major short-term complication encountered, treated with post-operative dilation sessions. Dysphagia and reflux were the most common long-term complications. **Conclusions:** Oesophageal atresia without severe cardiovascular abnormalities could be treated at a peripheral paediatric surgical department with satisfactory outcomes. However, qualified paediatric surgeons, anaesthesiologists and neonatologists and the availability of neonatal intensive care unit should be definitively required.

Keywords: Management, morbidity, oesophageal atresia, outcome, stenosis

Introduction

Oesophageal atresia is a congenital anomaly with a frequency range from 1:2400 to 1:4500 births.[1,2] The year 2018 marked the 130th anniversary of the first-known surgical attempt to repair oesophageal atresia, performed by Charles Steele.[3] Cameron Haight, a thoracic surgeon trained in adult procedures, performed in 1941 the first successful primary repair of a patient with a distal fistula.[3,4] The founders of contemporary paediatric surgery set the classification and treatment principles of an anomaly that until today remains a challenge in many aspects.[2-4] It is said that oesophageal atresia is a malformation that defied primary surgical repair in the 20th century.[4]

Low birth weight, prematurity and associated anomalies compromise prognosis and survival rate. However, with neonatal intensive care management and evolution in surgery and anaesthesia, survival today exceeds 90%.[2,5] However, post-operative morbidity still remains an issue.[2] Recent attempts to establish guidelines for the treatment of post-operative complications, revealed the low level of evidence and the lack of systematic approach on treatment, which is often based on the opinion of experts.[2,6]

The aim of this study is to investigate the evolution of management throughout time, during a period of 20 years at a district paediatric surgical department that serves a population of 300,000, where congenital anomalies are not encountered as frequently as at major centres. The question posed was if all patients with oesophageal atresia should be transferred to major centres for surgical correction, or if treatment of patients without major cardiovascular anomalies or other severe malformations is acceptable at such facilities, under certain

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prerequisites. Post-operative morbidity was the primary factor under consideration.

Methods

The study included a cohort of 33 patients with oesophageal atresia without major cardiovascular anomalies (14 male and 19 female neonates) who were operated from 1 January 1994, to 31 December 2013. Patients with known major cardiovascular anomalies according to Spitz's definition of the term,[7] either with a antenatal diagnosis that leaded their mothers to give birth at a major institution, or if diagnosed postnatally, were transferred to a major centre with a paediatric cardiovascular surgical department. Information was acquired from the hospital archives, follow-up of patients and personal contact with patients and their parents.

Approval for the study was obtained from the bioethics committee of the institution where the treatment was performed. Informed consent was acquired from the parents of the patients and the patients themselves for those who are adults today.

Demographics, conception, gestation, clinical presentation, operative technique and complications were studied. To evaluate the evolution of outcome and post-operative complications throughout time, the cohort was divided into two subgroups, one including patients operated from January 1994 to December 2003 (Group A), and a second from January 2004 to December 2013 (Group B).

The results were related to those of our cross-sectional study[8] on a cohort of families ($n = 16$) having experienced oesophageal atresia, with participants both patients in person ($n = 8$) and their proxies ($n = 25$), which was performed years after repair and regarded their long-term quality of life.

Descriptive statistical analysis was applied. Chi-squared and Fisher's exact tests were performed for categorical data. The threshold for statistical significance was defined as $P < 0.05$. Statistical analysis was made using IBM SPSS version 25 software (IBM Corp, Armonk, NY, USA).

Results

Conception and pregnancy

Thirty patients conceived normally: two by *in vitro* fertilisation and one by assisted fertilisation. Two neonates came from twin pregnancies. The gestational period ranged from 30 to 40 weeks (mean: 36.13, standard deviation: 2.68 weeks). There was prematurity in 21 neonates (63.6%). Antenatal ultrasound evaluation was performed in 21 (63.6%); 11 of these patients (52.3%) presented with polyhydramnios. The birth weight ranged from 1179 to 3555 g (mean: 2436, standard deviation: 572.61 g). Two patients weighed < 1500 g at birth.

Clinical presentation

Saliva drooling ($n = 17$, 51.5%), choking ($n = 13$, 39, 4%) and vomiting at the first feeding attempt ($n = 4$, 12.1%) were the main clinical symptoms. Aspiration incidents were encountered in 12 neonates. The patients with aspiration ($n = 12$, ten of Group A and 2 of Group B) at birth, presented with pneumonia and respiratory distress. All patients were supported at the neonatal intensive care unit and were evaluated with cardiovascular and renal ultrasound. The Vertebral, Anal atresia, Cardiac, Tracheo-oesophageal fistula, Renal, Limb association criteria were screened in five patients. None of the patients presented with severe cardiovascular abnormality. Seven patients with congenital cardiopathies such as patent ductus arteriosus ($n = 3$), atrial septal defect ($n = 2$), ventricular septal defect ($n = 1$) and left pulmonary valve stenosis ($n = 1$), were non-haemodynamically significant and evaluated as non-compromising the prognosis and the post-operative course, and hence proceeded to oesophageal atresia correction at our institution. The associated extracardiac congenital anomalies in the study group are presented in Table 1.

Surgical repair

Surgical approach was performed with right posterolateral thoracotomy and extra-pleural exposure of the posterior mediastinum. In 24 patients, surgery was performed during the first 24 h, in four between 24 and 48 h and in five between 48 and 36 h. The anatomical types encountered were atresia with peripheral tracheo-oesophageal fistula in 30 patients (90.9%), isolated atresia in two (6.06%) and double proximal and peripheral fistula in one patient (3.03%). After closure of the tracheo-oesophageal fistula, primary anastomosis of the oesophageal segments was performed in 23 patients (69.6%) [Table 2].

The remaining ten patients had a long intersegmental gap, compromising the achievement of a tension-free primary anastomosis. The procedures performed in these patients included (i) oesophagostomy of the proximal segment combined with gastrostomy ($n = 4$), all performed before 2000; (ii) anastomosis combined with temporary gastrostomy ($n = 3$, circular myotomy performed in one patient);[9,10] and (iii) gastrostomy without primary anastomosis ($n = 3$) [Table 2].

Post-operative morbidity

The post-operative complications are shown in Table 3. There was no perioperative or post-operative mortality. The

Table 1: Extracardiac congenital anomalies presented in the study group

Type of anomaly	*n*
Cranial malformation	1
Lung hypoplasia	1
Tracheomalacia	6
Duodenal atresia	2
Annular pancreas	1
Jejunal atresia	1
Malrotation	1
Anorectal anomalies	3
Common urogenital tract	1
Inguinal hernia	3
Undescended testes	2
Disorder of sex development	1
Skeletal disorder	3

patients who presented with anastomotic leakage (n = 2), chylothorax (n = 1) and gastrointestinal reflux (n = 4) were treated conservatively. The post-operative short-term severe respiratory complication was pneumonia (n = 6 patients). All cases were treated effectively. During the long-term follow-up, four patients presented with pneumonia.

Anastomotic stenosis was the most common short-term complication. Five patients with anastomotic stenosis (n = 5, one of Group A and four of Group B) were successfully treated with dilations [Table 3]. In three patients with stenosis, one of Group A and two of Group B, additional temporary gastrostomy was required to obtain adequate nutrition.

Cervical oesophagostomy, an option in patients of Group A with a long gap between the oesophageal segments, was abandoned in the patients of Group B. There was a shift throughout time from staged to primary repair (P = 0.008), for the cases with a moderately long gap. This occasionally resulted in anastomoses that required post-operative dilations (n = 5). The dilation protocol consisted of one session under endoscopic inspection using oesophageal bougies, initiating after the 3rd post-operative month and repeated once in a month for 2 more months. Radiologic evaluation was performed prior to each dilation session.

Discussion

In the latest ESPGHAN-NASGHAN guidelines on oesophageal atresia evaluation, repair and complications, there were certain statements of interest: (i) the anomaly has shifted from an issue of mortality to an issue of morbidity, (ii) it is a lifetime and not only neonatal problem and (iii) there is lack of a systematic approach for long-term care.[2,6] These statements were an incentive to review our own experience on surgical repair and complications throughout time.

Table 2: Types of operations performed in the two groups of patients

	Group A	Group B	Total
Primary procedure			
Anastomosis (P=0.008)	7	16	23
Oesophagostomy and gastrostomy	4	0	4
Gastrostomy	1	2	3
Anastomosis and temporary gastrostomy	3	0	3
Secondary procedure			
Temporary gastrostomy	2	1	3
Anastomosis	4	1	5

Table 3: Post-operative complications following anastomosis of the oesophageal segments

	Group A (n=15)	Group B (n=18)	Total (n=33)
Short-term complications			
Stenosis	5	5	10
Leakage	2	0	2
Chylothorax	1	0	1
Long-term complications			
Dysphagia	1	3	4
Reflux	1	3	4

Major cardiac anomalies, severe prematurity and pre-operative estimation of long distance with bronchoesophagoscopy have been proposed as criteria to proceed to staged correction to avoid stenosis.[11-13] There is no precise definition of long-gap oesophageal atresia. Researchers proposed to consider as such, any atresia that cannot be repaired with a single operation.[14] Others suggested repair in two stages when the defect is longer than the length of three vertebrae. A spontaneous growth of the two oesophageal segments is anticipated in 6-8 weeks, followed by a secondary correction, either with anastomosis, or with a gastric or intestinal transplant.[13] There was a trend to attempt primary anastomosis in patients with a marginal gap throughout time at our institution.

Gastro-oesophageal reflux, peptic esophagitis, gastric metaplasia, Barrett oesophagus, anastomotic strictures, feeding disorders, dysphagia and oesophageal dysmotility are complications reported.[2] Adenocarcinoma and epidermoid carcinoma are life-threatening complications reported in later life.[15] The most common post-operative complication is anastomotic stenosis, with an impressive frequency variability between 10% and 80%.[2,5,6,16] A minor narrowing of the diameter of the anastomotic ring is often anticipated because of fibroblastic activity resulting in wound contraction.[17] Again, based on the opinion of experts, guidelines suggest that radiologic or endoscopic findings combined with the symptoms of clinical function impairment should be included to define anastomotic stenosis.[2] Anastomotic stenosis rate is considered proportionate to the distance between the oesophageal segments.[18]

There is no consensus on the proper measurement of the diameter of a post-operative stricture. Mathematical indexes have been proposed to achieve this task.[19,20] Radiologic evaluation followed by oesophagoscopy was performed in all the patients of our study to set the diagnosis of post-operative stenosis. Our results complied with a study reporting no association of type of surgery between primary and staged repair with post-operative morbidity.[21]

Management of post-operative stenosis is also an issue under debate. Questions have been posed on the efficiency of pharmaceutical anti-acid treatment and post-operative dilations.[2] The usefulness of proton pump inhibitors is still under debate, with controversial evidence on the protective effect on the oesophageal mucosa.[2,12,16,22,23] In a recent study, post-operative dilations were needed in 46% of corrected cases, with 51% of dilations performed in the 1st year of life, 16% during the 2nd and 33% from the 2nd to the 14th year.[12,16] Tambucci *et al*. provided an interesting and simplified algorithm for the management of anastomotic strictures following oesophageal atresia repair.[5]

We performed recently a cross-sectional study in 16 families of children who underwent oesophageal atresia treatment at our department during the last 25 years, and interviewed

both patients and their proxies on their quality of life in the physical, social and emotional domains.[8] These results reflected the patients included in the present study. In feeding issues, patients reported chocking while eating (37.9%), need of bigger fluid intake during lunch or dinner to surpass feeding difficulties (46.2%), a sense of chest burning (22.2%) and vomiting experiences (31.8%). Feeding issues paused to be an issue after the age of 4 years and no further dilation has been needed. Patients older than this age described their eating routine as satisfactory, except one who had feeding difficulties until the age of 6 years. However, patients were stressed (50%) when they had to eat at school or away from home.

Sleep issues (34.5%) and difficulties to find clothes of appropriate size for their age (18.5%) were also problems of the physical domain. In the social domain, they described themselves open (66.7%), confident (33.3%) and cautious (16.7%). All patients felt uncomfortable with their appearance because of their operative scars, though not disappointed. In the emotional domain, anxiety (100%) and disappointment (50%) were induced when dilations or medications were needed. They did not believe they would have any difficulties in finding friends, but 20% felt unsupported by their families. Finally, 33.3% of the patients felt that they would have difficulties in finding a partner.[8]

Limitations

The small number of patients and the retrospective character of the study are considerable limitations. This was anticipated, as the study comprised data from a district institution, with less congenital anomalies encountered compared to major institutions in big cities. Furthermore, two factors, that is, low birth rate and antenatal ultrasound which highlight the problem in foetal life often in favour of the decision of termination of pregnancy, have reduced frequency of congenital anomalies during the last decades.

Conclusions

Moderately, long-gap atresia treated with a single operation, results in anastomosis with a relative tension treated satisfactorily with dilations. Regarding the presentation of complications, it could be presumed that oesophageal atresia not associated with severe cardiovascular anomalies, may be treated successfully in any paediatric surgical department, with the prerequisite that there is availability of neonatal intensive care support, and treatment is performed by qualified paediatric surgeons, anaesthesiologists and neonatologists. This might affect decisions in paediatric surgical institutions which serve smaller population. Availability of certified healthcare workers and neonatal intensive care units may guarantee acceptable management.

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Conflicts of interest

There are no conflicts of interest.

References

1. Patria MF, Ghislanzoni S, Macchini F, Lelii M, Mori A, Leva E, *et al*. Respiratory morbidity in children with repaired congenital esophageal atresia with or without tracheoesophageal fistula. Int J Environ Res Public Health 2017;14:1136.
2. Krishnan U, Mousa H, Dall'Oglio L, Homaira N, Rosen R, Faure C, *et al*. ESPGHAN-NASPGHAN guidelines for the evaluation and treatment of gastrointestinal and nutritional complications in children with esophageal atresia-tracheoesophageal fistula. J Pediatr Gastroenterol Nutr 2016;63:550-70.
3. Mazurak M, Patkowski D. The visionaries, the virtuosos, and the history of congenital esophageal atresia surgery. Ann Thorac Surg 2018;106:305-8.
4. Muensterer OJ, Berdon WE. From Vogt to Haight and Holt to now: The history of esophageal atresia over the last century. Pediatr Radiol 2015;45:1230-5.
5. Tambucci R, Angelino G, De Angelis P, Torroni F, Caldaro T, Balassone V, *et al*. Anastomotic strictures after esophageal atresia repair: Incidence, investigations, and management, including treatment of refractory and recurrent strictures. Front Pediatr 2017;5:120.
6. Baird R, Lal DR, Ricca RL, Diefenbach KA, Downard CD, Shelton J, *et al*. Management of long gap esophageal atresia: A systematic review and evidence-based guidelines from the APSA outcomes and evidence based practice committee. J Pediatr Surg 2019;54:675-87.
7. Spitz L. Oesophageal atresia. Orphanet J Rare Dis 2007;2:24.
8. Athanasopoulou M, Sinopidis X, Koletsis E, Dougenis D, Karatza A, Dimitriou G, *et al*. Evaluation of long-term quality of life in children and young adults after neonatal surgery for esophageal atresia. Gazzetta Med Italiana Arch per le Sci Med 2018;177:253-9.
9. Livaditis A, Rådberg L, Odensjö G. Esophageal end-to-end anastomosis. Reduction of anastomotic tension by circular myotomy. Scand J Thorac Cardiovasc Surg 1972;6:206-14.
10. Uygun I, Zeytun H, Otcu S. Immediate primary anastomosis for isolated oesophageal atresia: A single-centre experience. Afr J Paediatr Surg 2015;12:273-9.
11. Valfrè L, Conforti A, Morini F, Bagolan P. Letter to the Editor in re: Foker process for the correction of long gap esophageal atresia: Primary treatment versus secondary treatment after prior esophageal surgery. J Pediatr Surg 2015;50:1807.
12. Bagolan P, Valfrè L, Morini F, Conforti A. Long-gap esophageal atresia: Traction-growth and anastomosis-Before and beyond. Dis Esophagus 2013;26:372-9.
13. Fall M, Mbaye PA, Horace HJ, Wellé IB, Lo FB, Traore MM, *et al*. Oesophageal atresia: Diagnosis and prognosis in Dakar, Senegal. Afr J Paediatr Surg 2015;12:187-90.
14. Bairdain S, Hamilton TE, Smithers CJ, Manfredi M, Ngo P, Gallagher D, *et al*. Foker process for the correction of long gap esophageal atresia: Primary treatment versus secondary treatment after prior esophageal surgery. J Pediatr Surg 2015;50:933-7.
15. Rintala RJ, Pakarinen MP. Long-term outcome of esophageal anastomosis. Eur J Pediatr Surg 2013;23:219-25.
16. Stenström P, Anderberg M, Börjesson A, Arnbjörnsson E. Dilations of anastomotic strictures over time after repair of esophageal atresia. Pediatr Surg Int 2017;33:191-5.
17. Manfredi MA. Endoscopic management of anastomotic esophageal strictures secondary to esophageal atresia. Gastrointest Endosc Clin N Am 2016;26:201-19.
18. Mousa H, Krishnan U, Hassan M, Dall'Oglio L, Rosen R, Gottrand F, *et al*. How to care for patients with EA-TEF: The known and the unknown. Curr Gastroenterol Rep 2017;19:65.
19. Said M, Mekki M, Golli M, Memmi F, Hafsa C, Braham R, *et al*. Balloon dilatation of anastomotic strictures secondary to surgical repair of oesophageal atresia. Br J Radiol 2003;76:26-31.
20. Sun LY, Laberge JM, Yousef Y, Baird R. The esophageal anastomotic stricture index (EASI) for the management of esophageal atresia. J Pediatr Surg 2015;50:107-10.
21. Osei-Nketiah S, Hesse AA, Appeadu-Mensah W, Glover-Addy H, Etwire VK, Sarpong P. Management of oesophageal atresia in a developing country: Is primary repair forbidden? Afr J Paediatr Surg 2016;13:114-9.
22. Kovesi T, Rubin S. Long-term complications of congenital esophageal atresia and/or tracheoesophageal fistula. Chest 2004;126:915-25.
23. Shawyer AC, D'Souza J, Pemberton J, Flageole H. The management of postoperative reflux in congenital esophageal atresia-tracheoesophageal fistula: A systematic review. Pediatr Surg Int 2014;30:987-96.