Journal of Pediatric Surgery 51 (2016) 1421–1425

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Outcome and management in infants with esophageal atresia – A single centre observational study

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ARTICLE INFO

Article history:
Received 28 December 2015
Received in revised form 14 February 2016
Accepted 20 March 2016

Key words:
Esophageal atresia
Tracheoesophageal fistula
Outcome
Morbidity

ABSTRACT

Background/Purpose: A successful outcome in the repair of esophageal atresia (EA) is associated with a high quality pediatric surgical centre, however there are several controversies regarding the optimal management. The aim of this study was to investigate the outcome and management EA in a single pediatric surgical centre.
Methods: Medical records of infants with repaired EA from 1994 to 2013 were reviewed.
Results: 129 infants were included. Median follow-up was 5.3 (range 0.1–21) years. Overall survival was 94.6%, incidences of anastomotic leakage 7.0%, recurrent fistula 4.6% and anastomotic stricture 53.5% (36.2% within first year). In long gap EA (n = 13), delayed primary anastomosis was performed in 9 (69.2%), gastric tube in 3 (23.1%) and gastric transposition in one (7.7%) infants. The incidences of anastomotic leakage and stricture in long gap EA were, 23.1% and 69.2%, respectively. Peroperative tracheobronchoscopy and postoperative esophagography were implemented as a routine during the study-period, but chest drains were routinely abandoned.
Conclusion: The outcome in this study is fully comparable with recent international reports showing a low mortality but a significant morbidity, especially considering anastomotic strictures and LGEA. Multicenter EA registry with long-term follow up may help to establish best management of EA.

Esophageal atresia (EA) is a rare congenital malformation affecting approximately 1 in every 3500 live births [1–5]. Advances in anesthesia, neonatal intensive-, surgical- and cardiac care have improved the overall survival rate in infants with EA since Haight and Towsley reported the first successful primary repair in 1943 [6]. Nevertheless, a significant morbidity has remained [7–12]. A successful outcome in the repair of EA has been associated with a high quality pediatric surgical centre, however there are several controversies regarding the best practice of EA [13].

The aim of this study was to investigate the outcome and management in infants with EA in a single pediatric surgical centre over the last two decades. Moreover, we aimed to compare the results from this study with recent reports from international pediatric surgical centres with a high caseload.

1. Patients and methods

This is a retrospective observational study of all the infants with repaired EA between 1994 and 2013 at the University Children's Hospital, Uppsala, Sweden. The use of patient data in this study was approved by the Regional Committee on Medical Research Ethics (Dnr 2014/119/1). Data was obtained from the medical records. EA was classified according to the Gross classification [14] and risk groups were defined according to Spitz classification [15]. Major cardiac anomaly was defined according to Spitz [15] (either cyanotic congenital heart disease that required palliative or corrective surgery or noncyanotic congenital heart disease that required medical or surgical treatment for heart failure). A diagnosis of VACTERL (Vertebra, Anorectal, Cardiac, Tracheo-Esophageal, Renal, Limb) association was made if three or more components of the association were present. An anastomotic stricture was defined as a symptomatic narrowing of the anastomosis that needed dilatation. We defined long gap EA (LGEA) as a gasless abdomen (Gross type A or B) with an initial distance of three vertebral bodies or more. We divided the study population into two subgroups; non-LGEA (Gross type C, D, E) and LGEA. The median follow-up time was 5.3 years (range 0.1–21).

1.1. Preoperative management

All infants had a chest and abdominal X-ray before surgery. Upper esophageal pouch decompression was performed by a nasogastric tube and preoperative antibiotics were given.

Echocardiography was done preoperatively. Peroperative tracheobronchoscopy was routinely performed since 2012.

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http://dx.doi.org/10.1016/j.jpedsurg.2016.03.010
0022-3468/

Table 1
Type of esophageal atresia (EA).

Gross type of EA	All infants		Infants with LGEA	
	(n = 129)	(%)	(n = 13)	(%)
A	10	7.75	10	76.92
B	3	2.33	3	23.08
C	107	82.95		
D	2	1.55		
E	7	5.42		
LGEA	13			

LGEA indicates long gap esophageal atresia.

1.2. Surgery of Gross type C, D and E (non-LGEA)

An upper tracheoesophageal fistula (TEF) was first located by tracheobronchoscopy and then cannulated before division of the fistula through a right cervical incision. The repair of the upper and lower esophageal segments in Gross type C was performed through an open thoracotomy (extrapleural approach). All the infants had a 5 or 6 Fr transanastomotic tube.

Since 2008 a chest drain was not routinely inserted. It was only inserted in infants with a tense anastomosis. From 2008 we have routinely applied a continuous 72-hour local anesthetic infiltration with levobupivacaine (Chirocaine 0.625 or 1.25 mg/ml; AbbVie AB, Boxs 1523, 171 29 Solna, Sweden) in the wound.

The repairs of EA were mainly performed by three senior surgeons.

1.3. Surgery of LGEA (Gross A and B)

All these patients had a feeding gastrostomy with an open technique without an esophagostomy with exception of one patient. The esophageal gap was assessed peroperatively at the same time as the gastrostomy was performed and then repeated prior to the esophageal reconstruction. Hegar dilators were used to identify the lower esophageal segment and at the same time an endoscope was placed in the upper esophageal segment, the segments were maximally pushed against each other and the gap was visualized with X-ray. After postoperative recovery the infants were transferred to their local hospital or home if the parents could have assistance with suction in the upper esophageal pouch. The surgical options for esophageal reconstruction were delayed primary anastomosis (DPA), gastric transposition or reversed gastric tube. The repairs of LGEA were mainly performed by three senior surgeons.

1.4. Postoperative management

Elective paralysis was only adopted in infants with severe tension in the anastomosis. Since 2011 oral feeding started and antibiotics were seponated after a routine contrast esophagography 7 days after the surgery. The nasogastric tube was removed when the infant could eat enough to gain weight. Prior to discharge from hospital the infant had a spinal X-ray and a renal ultrasonography.

Anastomotic leakage was treated with broad-spectrum antibiotics, chest drainage and total parenteral nutrition. Anastomotic stricture was treated with balloon dilatations. Recurrent TEF was treated endoscopically with glue or surgery.

1.5. Statistical analysis

To compare patients with LGEA to the non-LGEA study group, Fisher's exact test was performed on qualitative variables and Wilcoxon rank sum test with continuity correction on quantitative parameters. *P*-values less than 0.05 were considered statistically significant.

Statistical analyses were performed using R version 3.2.2, Copyright (C) 2015 The R Foundation for Statistical Computing, with Rcmdr Version 2.2-0.

2. Results

2.1. Patient characteristics

Totally there were 135 patients with EA during the study period at our centre, 6 patients were excluded from the study; 1 disapproved participation, 1 was operated at another centre, 2 had trisomy 18 and 2 had other severe multiple malformations and were not operated.

Totally 129 patients were included, 13 had LGEA and Gross type C was the predominant anatomical variation (Table 1). EA had been suspected on the basis of prenatal ultrasonography in 17 (13.2%) infants, because of absent stomach (Table 2). EA had been suspected on prenatal ultrasonography in 9 out of totally13 infants (69.2%) with LGEA.

Gestational age was lower in LGEA. (Table 2). Gender distribution, median maternal age, incidences of major cardiac anomalies, chromosomal abnormalities, CHARGE- and VACTERL association and very low birth weight were not significantly different between the non-LGEA and LGEA study groups (Table 2).

Median age at surgery was significantly higher in LGEA (3 days compared with 6.7 months). Median days on ventilator after surgery were 2 days in both groups. The median time to full enteral nutrition after repair of EA was 11 days in the non-LGEA and 17 days in LGEA group.

Table 2
Patient characteristics.

Patient characteristics	All infants (n = 129)	Non-LGEA (n = 116)	LGEA (n = 13)	p-value*
Boys	74 (57.4%)	65 (56.0%)	9 (69.2%)	0.55
Girls	55 (42.6%)	51 (44.0%)	4 (30.8%)	
Median age of the mother (years) (min; Q1; Q3; max)	29 (16, 26, 32, 41)	29 (16, 26, 32, 41)	29 (23, 27, 33, 40)	0.31
Prenatal diagnosis	17 (13.2%)	8 (6.9%)	9 (69.2%)	6.6e-7
Median gestation weeks (min; Q1; Q3; max)	38 (25, 35, 39, 42)	38 (25, 35, 39, 42)	36 (30, 34, 38, 39)	0.041
Median birth weight gram (min; Q1; Q3; max)	2685 (525, 2146, 3250, 4210)	2703 (525, 2264, 3299, 4210)	2184 (1272, 1950, 2844, 3350)	0.075
Major cardiac defects	13 (10.1%)	12 (10.3%)	1 (7.7%)	1
Chromosomal abnormalities	4 (3.1%)	4 (3.4%)	0 (0%)	1
CHARGE association	2 (1.6%)	2 (1.7%)	0 (0%)	1
VACTERL association	24 (18.6%)	22 (19.0%)	2 (15.4%)	1
Very low birth weight (<1500 g)	8 (6.2%)	7 (6.0%)	1 (7.7%)	0.58
Median age at surgery(days) (min; Q1; Q3; max)	3 (1, 2, 4, 578)	3 (1, 2, 3, 255)	202 (77, 137, 294, 578)	1.33e-9
Median days on ventilator (min; Q1; Q3; max)	2 (1, 1, 3, 21) (NA = 5)	2 (1, 1, 3, 21) (NA = 5)	2 (1, 1, 6, 17)	0.99
Median time to full enteral nutrition (days) (min; Q1; Q3; max)	11 (3, 9, 15, 451) (NA = 14)	10.5 (3, 9, 14, 451)	17 (10, 13, 28, 74)	5,82e-4
Overall survival rate	122 (94.6%)	109 (94.0%)	13 (100%)	1

* Reported p-value is for the comparison between non-LGEA and LGEA.