Primary repair in esophageal atresia

The results of long term follow-up

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Aim. **The aim of this study was to assess the impact of postoperative morbidity during a long-term follow-up (6-12 years) in children with esophageal atresia treated at birth by primary anastomosis.**

Methods. **Fifteen children with esophageal atresia and tracheoesophageal fistula were surgically treated at birth and their follow-up was extended to at least 6 up to 12 years. Data included clinical examination, evaluation of nutritional habit, continuous video recording of barium esophagogram, esophageal manometry, 24-h esophageal pH-monitoring and esophageal endoscopy.**

Results. **All the 15 patients completed the clinical evaluation and the set of tests. In the first 6 years, mild dysphagia and gastroesophageal reflux (GER) was observed in 3 cases whereas GER without dysphagia in 4 cases. These 7 patients were informed about simple nutritional behaviours to minimize symptoms and treated with H2-blockers. At long-term twelve-year analysis, all patients were between 50° and 75° percentile of expected growth. It was not referred peculiar food restrictions. Five patients showed mild dysphagia with solid foods; early satiety, epigastric burning and regurgitation were less frequent. Furthermore they showed multiple nonperistaltic body contractions at esophagogram and moderate impairment of esophageal motility at esophageal manometry. The 24-h esophageal pH-monitoring showed normal patterns in all patients. No major lesions of esophageal mucosa were detected at esophagoscopy.**

Conclusion. **Although GER and esophageal dismotility are reported as frequent findings in patients who underwent primary repair for esophageal atresia, these disorders don't cause any relevant impairment to the quality of their nutritional habit.**

Key words: **Gastroesophageal reflux - Esophageal atresia - Esophageal motility disorders.**

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Esophageal atresia (EA) is a fairly common congenital disorder whose incidence ranges between 1/4 000 and 1/5 000 births.[1] Nowadays, primary end-to-end anastomosis is carried out in the most of cases allowing a normal lifespan. Alternative surgical procedures are performed in a selected series of patients (long gap EA) or after failure of primary repair. Although the advances in neonatal intensive care and in surgical management have drastically reduced the mortality rate (8%) in infants weighing more than 2 500

g,[2] EA continues to be an important cause of morbidity. Gastroesophageal reflux (GER) is the main cause of postoperative morbidity occurring up to 35% to 58% of children with EA. Other complications related to EA (dysphagia, esophagitis, esophageal stenosis, respiratory distress, asthma, Barrett's esophagus) may impair daily nutritional habits influencing the quality of life.[3]

Aim of this study has been to assess the impact of postoperative morbidity in children with EA treated at birth by primary esophageal repair during a long-term (6 to 12 years) follow-up.

Materials and methods

We included in the study the patients who underwent primary repair for EA in our Department in the last 12 years. Long gap EA and/or delayed anastomosis were considered as exclusion criteria. The newborns who belonged to the most frequent group of EA (Vogt type III with the upper esophagus ending in a blind pouch and the lower esophagus joining the respiratory tract near the tracheal bifurcation with a tracheoesophageal fistula) were recruited for a long-term follow-up (6-12 years). The work-up has been tailored to physical examination and nutritional habits. Continuous video recording of barium esophagogram, esophageal manometry, 24-h esophageal pH-monitoring and esophageal endoscopy were performed to identify reflux recurrence and the cause of symptoms. We focused on the symptoms of GER and patients' eating habits.

Barium esophagogram was ensured to monitor the swallowing step and to assess the esophageal body motility and the sphincter function.

Esophageal manometry was carried out after an overnight fast. A water perfusion system with a 8 Fr manometric catheter was connected to a multichannel polygraph recorder. The proximal and distal esophageal pressures were measured as mmHg. All patients were studied for 30 min and at least 10 wet swallows were recorded. Esophageal motility was considered abnormal if middle esophagus peak contraction pressures were <30 mmHg and/or abnormal coordination between waves at esophageal body during swallowing occurred.

Twenty-four-hour esophageal pH-monitoring was carried out using a 2.1-mm diameter glass probe calibrated in standard buffer solution at pH 7 and 1, advanced 5 cm above the manometrically determined lower esophageal sphincter. pH reference electrode was connected to a data logger and kept in place for 24 h; this probe continuously recorded pH values at 6-s intervals. The patients were properly instructed to keep a written record of symptoms, meals and positioning and they were asked to avoid frozen or very hot foods. H2 blockers were suspended 2 days before the study. The computer software determined the pH-metric variables: acid reflux index (percentage of total time pH<4), number of reflux episodes, duration of longest episode pH<4 in minutes, number of reflux episode lasting more than 5 min.

TABLE I.—*Clinical evaluation and therapy in 15 patients.*

Patients	Clinical evaluation			
	Dysphagia		Other symptoms	
	6 years	12 years	6 years	12 years
1				Early satiety
2		Mild		Epigastric burning
3	Mild	Mild	Nausea	Nausea
4				
5				
6				
7	Mild	Mild	Regurgitation	Early satiety
8				
9				Epigastric burning
10	Mild	Mild	Regurgitation	
11		Mild		Regurgitation
12			Nausea	Early satiety
13			Epigastric burning	
14				
15				

Results

All patients (9 males, 6 females) completed the clinical evaluation (Table I) and the set of tests (Table II).

TABLE II.—*Results of esophagogram, manometry and pH-monitoring in 15 patients.*

	Esophagogram	Manometry	pH monitoring			
	Presence of GER 6 y /12 y	P/D * (mmHg) 12 years	Total time pH <4 (%) 6 y /12 y	No. of episodes 6 y /12 y	Longest episode (min) 6 y /12 y	No. of episodes >5 min 6 y /12 y
1	+/-	17/20	0/0	0/0	0/0	0/0
2	+/-	15/25	2.1/1.3	2/1	6/2	8/4
3	+/-	14/15	0.8/0.7	1/3	4/6	2/1
4	-/-	14/35	0/0	0/0	0/0	0/0
5	-/-	18/36	0.5/0.2	0/0	2/0	3/0
6	+/-	11/16	0.8/0.3	1/0	5/2	4/1
7	+/-	14/18	1.7/0.4	3/0	7/1	1/1
8	-/-	17/31	0.2/0	0/0	0/0	0/0
9	-/-	19/37	0/0	0/0	0/0	0/0
10	+/-	17/17	2/1	2/2	3/2	2/2
11	+/-	19/22	0.1/0	0/0	0/0	0/0
12	-/-	15/15	1.5/1.3	3/4	5/6	2/3
13	-/-	14/16	0.3/0.1	0/0	0/0	0/0
14	-/-	15/32	0.2/0.1	0/0	0/0	0/0
15	-/-	15/26	0/0	0/0	0/0	0/0

* P/D: Proximal and distal esophageal pressures (mmHg); mean and standard deviation (SD) are 15.6±2.19 and 24.06±8.22 for proximal and distal pressures respectively.

Three patients showed severe dysphagia, nausea and regurgitation requiring Nissen antireflux fundoplicatio at 10 months, 4 and 5 years after surgery respectively. None of these patients reported symptoms after fundoplicatio.

In the first 6 years dysphagia due to abnormal motility pattern was observed in 3 cases (20%). Regurgitation (13.3%), nausea (13.3%), epigastric burning (6.6%) were less frequent. None of the patients reported aspiration pneumonia, chronic cough or asthma. Barium esophagogram showed GER and an apparent weakness in progression activity of esophageal body with a distal dilatation in 7 patients. Seven patients with dysphagia or GER were medically treated whit H2-blockers.

At long-term twelve-year evaluation all patients were between 50° and 75° percentile of expected growth. It was not referred peculiar food restrictions. In 5 patients mild dysphagia occurred, in particular with solid foods requiring to drink a lot of water to solve swallowing impairment. In 7 patients regurgitation, early satiety, epigastric burning and nausea were sporadically observed, but they didn't cause any severe disturbance. At barium esophagogram, these 5 disphagic patients showed multiple nonperistaltic body contractions with a light slowdown of barium progression near the anastomosis that didn't show stenosis. In 10 patients barium boles passed through without any impairment.

Esophageal manometry confirmed the observations seen at video recording esophagogram. Although the duration of contractions was normal, a significant failure of the peristaltic coordination was found in all children. Peak amplitudes changed in relation to esophageal levels. In our series middle esophagus had peak contraction pressures averaged 15.6±2.19 SD (mmHg), whereas mean contraction pressures for normal peristaltic ones being near 30 mmHg.[4] In the distal esophagus contraction amplitude averaged 24.06±8.22 SD (mmHg) being about half of value in normal esophagus. Low distal esophageal pressure was found in 10 patients.

The 24-h esophageal pH-monitoring showed normal patterns in all patients at 6 and twelve-year follow-up (Table II). Rare acid-GER not correlated with meal were observed but none of these cases was linked to symptoms.

Esophagoscopy was performed in 5 dysphagic patients: no major lesions of

esophageal mucosa or esophageal strictures in body or cardias were detected.

Discussion

Many children treated for EA may show anastomosis leakage, esophageal strictures, respiratory distress, sudden infant death, GER, dysphagia, esophagitis, poor growth and esophageal cancer risk.[5-7] GER is so common after surgical treatment for EA that is considered as part of the disease. GER is responsible of severe nutritional and respiratory complications which are the causes of frequent hospitalizations. Most of symptoms caused by GER are present in almost 50% of the patients who underwent anastomosis within 5 years from surgical repair. The peak of incidence of swallow impairment occurs around 2 years of age and goes on up to 5 years. The dysphagia for solid foods caused by GER is referred in 16% of patients older than 15 years of age, having similar incidence to adults.

GER appears to be due to the motor dysfunction of the esophagus and to the incompetence of the lower esophageal sphincter.[8] Reduced pressures of the low tract and nonpropulsive low amplitude contractions are usual findings in patients with EA suggesting that dysmotility related to structural neuromuscular disorganization is a primary event. In these patients the propulsion of bolus and the clearance of refluxed fluid could be accomplished mainly by gravity. At esophageal manometry we confirmed the failure of the peristaltic coordination and low peak contraction pressures in all patients.

At six-year follow-up we observed 3 patients with mild dysphagia and GER at esophagogram and 4 patients with GER only. These 7 patients were treated with H2-blockers, usually adequate to achieve acid suppression with symptomatic relief, and informed about simple nutritional behaviours to minimize symptoms (avoid of few solid foods, no feeds 2 h before bedtime, eliminate caffeine, small and frequent feeds, intakes of a lot of water to "push down" foods). Furthermore, we think that the abnormal structure of the esophagus makes these patients poor candidates for successful prokinetic treatment.

At twelve-year follow-up weight and height percentiles of 15 patients were not reduced compared to sibiling pairs. Five patients showed mild dysphagia and minor symptoms that were considered as unremarkable problems. It happened because most of them had reached an acceptable way of eating, the esophageal propulsive impairment progressively decreases and they have got simple nutritional behaviours. None of 15 patients had GER after the twelve-year evaluation. Last observation should be considered surprising in view of the fact that uncoordinated esophageal peristalsis, responsible of poor esophageal acid clearance,[8] still remains. The absence of GER after a twelve-year follow-up could be due to the anatomical stability of the high pressure zone, to the improvement of gastric emptying and to the possible efficacy of drugs.

These 15 patients had dismotility at manometry but normal esophageal pH and few symptoms of GER. The shortened intra-abdominal segment due to anastomosis plays a role in reducing pressures into the esophagus and contributes to the pathogenesis of GER. The partial vagal denervation and the traction on cardia due to esophageal mobilization for anastomosis also account for the failure of the antireflux barrier.[9] A careful surgical approach may reduce intrathoracic traction and explain why dismotility is not always associated to reflux.

Recently it has been reported [10] a wider use of antireflux fundoplicatio in children operated for EA. In our opinion fundoplicatio should be offered if medical treatment fails, children fail to thrive, aspiration secondary to reflux happens or esophageal stricture develops. Moreover, a surgical fundoplicatio, even if well performed, could lastly be too tight for an essentially hypoperistaltic esophageal body with an impairment of peristaltic activity.

Conclusions

Clinical evaluation, radiographic techniques, esophageal manometry and endoscopy are

useful for long term follow-up in children with EA treated at birth by primary anastomosis. These patients frequently show the failure of esophageal peristaltic coordination nevertheless it doesn't cause any relevant impairment to the quality of the nutritional habits. Moreover, although the GER is reported as the main functional complication in children treated at birth for EA, in our long-term follow-up GER was not observed. In our opinion, surgical correction of GER in patients with EA treated at birth by primary anastomosis is mandatory if medical management fails, a stricture develops, the patient has a poor growth or shows respiratory disorders.

Riassunto

Anastomosi primaria nell'atresia esofagea: risultati del follow-up

Obiettivo. Lo scopo di questo studio è stato valutare, per un periodo di 12 anni, le possibili complicanze nei pazienti pediatrici affetti da atresia esofagea corretta alla nascita con anastomosi termino-terminale.

Metodi. Quindici bambini operati per atresia esofagea e fistola tracheo-esofagea sono stati seguiti per un periodo di 6-12 anni. Sono stati eseguiti: valutazione clinica, studio delle abitudini alimentari, esofagogramma con mezzo di contrasto, manometria esofagea, pH-metria ed endoscopia esofagea.

Risultati. I 15 pazienti hanno completato la valutazione clinica e la serie di test previsti durante il follow-up nei 12 anni. Nei primi 6 anni dopo l'intervento sono stati osservati in 3 casi lieve disfagia e reflusso gastroesofageo, in 4 solo reflusso. A questi 7 pazienti sintomatici è stato consigliato di modificare alcuni semplici abitudini nutrizionali associando la somministrazione di antagonisti dei recettori H2. A 12 anni dall'intervento tutti i pazienti hanno mostrato un trend di crescita compreso tra il 50° e il 75° percentile. I pazienti non hanno riferito alcuna restrizione dietetica. Cinque pazienti hanno presentato lieve disfagia per i cibi solidi e, più raramente, bruciore epigastrico, rigurgiti e senso di sazietà. Sono state osservate, inoltre, numerose contrazioni non peristaltiche all'esofagogramma e una moderata alterazione della motilità esofagea allo studio manometrico. La pH-metria e l'esofagoscopia non hanno mostrato alterazioni o lesioni significative in nessuno dei pazienti esaminati.

Conclusioni. Anche se il reflusso gastroesofageo e l'alterata motilità esofagea sono di frequente riscontro nei pazienti affetti da atresia esofagea operati con anastomosi primaria, non sembrano, comunque, influenzare il loro comportamento nutrizionale.

Parole chiave: Reflusso gastroesofageo - Atresia esofagea - Disordini della motilità gastrica.

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