# Dysphagia among adult patients who underwent surgery for esophageal atresia at birth

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**BACKGROUND:** Clinical experiences of adults who underwent surgery for esophageal atresia at birth is limited. There is some evidence that suggests considerable long-term morbidity, partly because of dysphagia, which has been reported in up to 85% of adult patients who undergo surgery for esophageal atresia. The authors hypothesized that dysphagia in this population is caused by dysmotility and/or anatomical anomalies.
**OBJECTIVE:** To determine the motor and anatomical causes of dysphagia.
**METHODS:** A total of 41 adults, followed at the Esophageal Atresia Clinic at *Hôpital Saint-Luc* (Montreal, Quebec), were approached to particpate in the present prospective study. Evaluation was completed using upper endoscopy, manometry and barium swallow for the participants who consented. The medical charts of respondents were systematically reviewed from the neonatal period to 18 years of age to assess medical and surgical history.
**RESULTS:** All 41 patients followed at the clinic consented and were included in the study. Dysphagia was present in 73% of patients. Esophagogastroduodenoscopy was performed in 32 patients: hiatal hernia was present in 62% (n=20); esophageal diverticulum in 13% (n=4); macroscopic Barrett esophagus in 31% (n=10); and esophagitis in 19% (n=6). Histological esophagitis was present in 20% and intestinal metaplasia in 10%. There were no cases of dysplagia or adenocarcinoma. Esophageal manometry was performed on 56% of the patients (n=23). Manometry revealed hypomotility in 100% of patients and included an insufficient number of peristaltic waves in 96%, non-propagating peristalsis in 78% and low-wave amplitude in 95%. Complete aperistalsis was present in 78%. The lower esophageal sphincter was abnormal in 12 (52%) patients, with incomplete relaxation the most common anomaly. Of the 41 patients, 29 (71%) consented to a barium swallow, which was abnormal in 13 (45%). The anomalies found were short esophageal dilation in 28%, delay in esophageal emptying in 14%, diverticula in 14% and stenosis in 7% of patients. There was more than one anomaly in 14% of patients.
**CONCLUSION:** Dysphagia was a highly prevalent symptom in adults who underwent surgery for esophageal atresia. The present study is the first to demonstrate that motor and anatomical abnormalities may be implicated in causes of dysphagia in this population. Furthermore, these anomalies may be demonstrated with simple investigations such as endoscopy, manometry and barium swallow.

**Key Words:** *Barium swallow; Barrett esophagus; Dysphagia; Endoscopy; Esophageal atresia*

## La dysphagie chez les patients adultes ayant été opérés à la naissance en raison d'une atrésie œsophagienne

**HISTORIQUE :** On a peu d'expérience clinique auprès des adultes qui ont été opérés en raison d'une atrésie œsophagienne à la naissance. Selon certaines données, la morbidité à long terme est considérable, en partie à cause de la dysphagie qui s'observe chez jusqu'à 85 % des patients adultes opérés en raison d'une atrésie œsophagienne. Les auteurs postulent qu'au sein de cette population, la dysphagie est causée par la dysmotilité ou les anomalies anatomiques.
**OBJECTIF :** Déterminer les causes motrices et anatomiques de la dysphagie.
**MÉTHODOLOGIE :** Au total, 41 adultes, suivis à la clinique d'atrésie œsophagienne de l'Hôpital Saint-Luc de Montréal, au Québec, ont été invités à participer à la présente étude prospective. Les chercheurs ont effectué l'évaluation par endoscopie œsogastroduodénale, manométrie et déglutition barytée chez les participants qui y ont consenti. Ils ont examiné systématiquement le dossier médical des répondants entre la période néonatale et l'âge de 18 ans afin d'évaluer leurs antécédents médicaux et chirurgicaux.
**RÉSULTATS :** Les 41 patients suivis à la clinique ont consenti et participé à l'étude. De ce nombre, 73 % avaient une dysphagie et 32 patients ont subi une œsophagogastroduodénoscopie : hernie hiatale chez 62 % (n=20), diverticule œsophagienne chez 13 % (n=4), œsophage de Barrett macroscopique chez 31 % (n=10) et œsophagite chez 19 % (n=6). De plus, 20 % avaient une œsophagite histologique et 10 % une métaplasie intestinale, mais on n'observait aucun cas de dysplagie ou d'adénocarcinome. Au total, 56 % des patients (n=23) ont subi une manométrie œsophagienne, qui a révélé une hypomotilité chez 100 % d'entre eux. Cette manométrie indiquait un nombre insuffisant d'ondes péristaltiques chez 96 % d'entre eux, une absence de propagation du péristaltisme chez 78 % d'entre eux et une faible amplitude des ondes chez 95 % d'entre eux. On constatait un apéristaltisme complet chez 78 % des patients. Le sphincter œsophagien inférieur était anormal chez 12 patients (52 %), la relaxation incomplète du sphincter étant l'anomalie la plus courante. Sur les 41 patients, 29 (71 %) ont consenti à une déglutition barytée, qui était anormale chez 13 patients (45 %). Les anomalies observées étaient une faible dilatation œsophagienne chez 28 %, un retard de la vidange œsophagienne chez 14 %, un diverticule chez 14 % et une sténose chez 7 % des patients. Enfin, 14 % des patients présentaient plus d'une anomalie.
**CONCLUSION :** La dysphagie était un symptôme hautement prévalent chez les adultes qui avaient été opérés en raison d'une atrésie œsophagienne. La présente étude est la première à démontrer que les anomalies motrices et anatomiques peuvent contribuer à la dysphagie au sein de cette population. De plus, ces anomalies peuvent être démontrées par de simples examens comme l'endoscopie, la manométrie et la déglutition barytée.

Esophageal atresia is a congenital anomaly defined by an interruption of the continuity of the esophagus with or without a communication with the trachea. This condition occurs in one in 2500 to 3000 live births (1) and is the most common congenital anomaly of the esophagus. There are five types of esophageal atresia; the most common is type C, with distal tracheoesophageal fistula occurring in 85% of cases. The majority of children with atresia undergo surgery at birth. Even if the continuity of the esophagus is anatomically replaced, the children are prone to several gastrointestinal problems including dysphagia and gastroesophageal reflux (GER) (2). Dysphagia is observed

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in more than one-half of children who undero surgery for esophageal atresia at birth, and may cause malnutrition and failure to thrive (3-5).

Clinical experiences of adults who undergo surgery for esophageal atresia at birth are limited. There is some evidence that suggests considerable long-term morbidity, partly because of dysphagia, which has been reported in up to 85% of adult patients who undergo surgery for esophageal atresia (6,7). Studies have shown that different anomalies may be implicated including dysmotility, stenosis, esophagitis and cancer (5). In fact, esophageal atresia is accompanied by intrinsic and extrinsic anomalies of neurological maturation, possibly leading to dysmotility. In addition, surgical repair may lead to stenosis and associated diverticula, which may contribute to dysphagia (5). However, there are no studies to guide the necessary investigations.

*Hôpital Saint-Luc* is a tertiary centre located in Montreal, Quebec, with a specialized clinic for adults who undergo surgery for esophageal atresia. We have previously demonstrated that dysphagia is a common symptom in this population (8,9); however, the etiology remains unclear. Our hypothesis was that dysphagia in this population is caused by dysmotility and/or anatomical anomalies. The objective of the current study was to determine the motor and anatomical causes of dysphagia.

## METHODS

Forty-one adult patients followed at the Esophageal Atresia Clinic at the *Hôpital Saint-Luc* were approached to participate in the present prospective study. After providing signed consent, all participants underwent an interview and clinical assessment. Evaluation was completed using upper endoscopy, manometry and barium swallow for individuals who consented. Medical charts of respondents were systematically reviewed from the neonatal period to 18 years of age to assess medical and surgical history.

### Interview and questionnaires

All participants were interviewed by an investigator and answered a standard questionnaire regarding gastrointestinal symptoms. Information regarding swallowing difficulties with different food textures, food impaction, dietary limitations, adaptive dietary habits and GER symptoms was requested. Dysphagia was defined as swallowing difficulties with one or multiple food textures and/or food impaction ≥1 per month. Criteria for GER were the presence of pyrosis and/or regurgitation ≥1 per month. Adaptive dietary habits included drinking water while eating, avoiding certain foods and usually eating slower than other people.

### Upper endoscopy and histology

After an overnight fast, esophagogastroduodenoscopy was performed under conscious sedation using an endoscope (EG2990i 2.8, Pentax Canada). Particular attention was devoted to the distal esophagus to locate the esophagogastric junction and the presence of mucosal injury. All abnormal endoscopic findings were reported. A fibrotic rim was defined as a narrowing of the esophagus, while a stricture was defined the inability to pass the endoscope through the narrowing (5). Esophagitis was graded according to the Los Angeles Classification (10). Barrett esophagus (11) was suspected when the squamocolumnar junction was ≥5 mm proximal to the esophagogastric junction. Biopsies were obtained for histological examination when an endoscopic anomaly was noted.

All biopsies were fixed overnight in Tissuefix (Qiagen, Canada) and subsequently embedded in paraffin. Six slices (3 µm each) were obtained for each biopsy and stained with hematoxylin phloxine saffron. The slides were reviewed by two pathologists blinded to the patients' clinical data. All cases were evaluated for the presence of esophagitis, metaplasia and dysplasia.

### Esophageal manometry

Stationary low-compliance perfusion manometry was performed using a round, four-lumen catheter. Lower esophageal sphincter (LES) pressure measurements were performed using the four distal openings of the catheter at a recording speed of 2.5 mm/s. The tip of the catheter was positioned in the stomach and then slowly withdrawn in 1.0 cm increments. LES pressure was recorded at midexpiration and end-expiration. Values were calculated as the mean of the three pressure channel readings. Contractions of the esophagus were recorded with the four pressure channels positioned 5 cm, 10 cm, 15 cm and 20 cm above the LES; 10 swallows of 5 mL each were then given at 30 s intervals. The measurement of each peristaltic parameter represented the mean of the 10 sequential swallows, and both amplitude and duration were individually determined for the different recording site above the LES. The data were transferred to a specifically designed software program (Gastrosoft, Synectics Medical Inc, USA) for analysis.

### Barium swallow

Barium swallow was performed according to a routine protocol. Participants stood for the examination. The fluoroscope (Philips, USA) was positioned and moved to capture the different phases of deglutition. Bolus transit patterns were assessed and anomalies were recorded in terms of intraesophageal stasis, dilation, diverticulum, stenosis and mass. All boluses were confirmed to have been cleared from the esophagus before the next bolus was administered.

### Statistical analysis

Data were collected in a database and analyzed using SPSS (IBM Corporation, USA). Differences between groups were analyzed using the $\chi^2$ test; P=0.05 was considered to be statistically significant.

### Ethics

The Ethics Committee of *Hôpital Saint-Luc* approved the present study and written informed consent was obtained from each patient. The study met the requirements of the Declaration of Helsinki.

## RESULTS

### Patients

All 41 patients followed at the clinic consented to participate and were included in the study; patient characteristics are summarized in Table 1. Esophageal atresia with distal tracheoesophageal fistula (type C, gross classification) was the most prevalent (85%) anomaly. At least one associated malformation was present in 26 (63%) patients. Eighty percent had a primary anastomosis and the others had either a gastric pull-up or a colonic interposition. The most frequent complications during childhood were failure to thrive (49%) and recurrent anastomotic stricture (29%).

All patients completed the questionnaire on gastrointestinal symptoms. Dysphagia was present in 30 (73%) patients (Table 1). Among these patients, dysphagia with solid food was present in 41%, with dry food in 32%, soft food in 10%, thick liquids in 10% and liquids in 10%. Blockages were present in six (15%) patients, but the episodes were self-resolved. Adaptive dietary habits were reported by 78% of the patients, the most common (61%) of which was to drink plenty between bites. Eating slower than other people was reported by 49% and avoidance of certain types of food by 15%. Twenty-two percent did not need to take precautions when eating. GER symptoms were recorded in 12 (29%) patients, including 17% under medical treatment and 12% without treatment. Pyrosis was present in 20% of patients and regurgitation in 24%.

### Endoscopic and histological findings (Table 2)

Esophagogastroduodenoscopy was performed in 32 (78%) patients. An esophageal anastomotic scar was recognizable in most (n=26 [81%]) cases of primary anastomosis; no strictures were found. Hiatal hernia was present in 20 (62%) patients, esophageal diverticulum in four (13%), macroscopic Barrett esophagus in 10 (31%) and esophagitis in six (19%), all of whom had grade A esophagitis. One-half of the patients with esophagitis and 80% of patients with macroscopic Barrett esophagus had no GER symptoms.

Biopsies were obtained in presence of macroscopic Barrett esophagus (n=10) and/or esophagitis (n=6). A total of 12 (38%) patients had

**TABLE 1**
**Characteristics of the study participants with surgically repaired esophageal atresia and the prevalence of selected gastrointestinal symptoms (n=41)**

| | |
|---|---|
| Age, years, mean (range) | 25 (18–44) |
| Male sex | 23 (56) |
| Body mass index, kg/m², mean (range) | 22 (15–38) |
| Dysphagia | 30 (73) |
| Swallowing difficulties* | 25 (61) |
| Blockage* | 6 (15) |
| Adaptive dietary habits | 32 (78) |
| Gastroesophageal reflux symptoms | 12 (29) |
| With medical treatment | 7 (17) |
| Without medical treatment | 5 (12) |

*Data presented as n (%) unless otherwise indicated. *One patient had both swallowing difficulties and a blockage*

**TABLE 2**
**Endoscopic findings in adult patients with surgically repaired esophageal atresia (n=32)**

| Endoscopic finding | |
|---|---|
| Esophagus | |
| Esophagitis | 6 (19) |
| Stricture | 0 (0) |
| Esophageal diverticulum | 4 (13) |
| Endoscopically suspected Barrett esophagus | 10 (31) |
| Stomach | |
| Hiatal hernia | 20 (62) |
| Aberrent pancreas | 8 (25) |

*Data presented as n (%)*

biopsies taken. No biopsies were taken in patients with a normal-appearing esophagus. Biopsies of the distal esophagus were examined to assess the presence of esophagitis, metaplasia, dysplasia or adenocarcinoma. Histological evidence of esophagitis was present in two patients who underwent biopsies without endoscopic esophagitis. Intestinal metaplasia was found in four of the 10 patients with endoscopically suspected Barrett esophagus. There was no dysplasia or adenocarcinoma in any biopsy specimen (Table 3).

**Esophageal manometry (Table 4)**

Esophageal manometry was performed in 23 (56%) patients and, of these, dysphagia was present in 13 (57%). Manometry revealed hypomotility in 100% of the patients and included an insufficient number of peristaltic waves in 96%, nonpropagating peristalsis in 78% and low wave amplitude in 95%. Complete aperistalsis was present in 78%. The LES was abnormal in 12 (52%) patients; the most common anomaly was incomplete relaxation and the upper esophageal sphincter was always normal.

**Barium swallow (Table 5)**

Of the 41 patients, 29 (71%) consented to a barium swallow, which was abnormal in 13 (45%). The anomalies found were: short esophageal dilation in eight (28% [four upstream and three downstream of the anastomosis]); delay in esophageal emptying in four (14% [two associated with aperistalsis in the inferior esophagus, one with an ileocolic transposition and one with a dilation]); diverticula in four (14% [one at the anastomosis level, three in the inferior esophagus]); and stenosis in two (7% [one in the proximal esophagus associated with a dilatation and one at the esophagocolic junction of a ileocolic transposition]). There was more than one anomaly in four (14%) patients. Dysphagia occurred in 48% of the 29 patients. Anatomical anomalies according to barium swallow were more prevalent in patients with dysphagia than in patients without dysphagia; however, this difference was not statistically significant (64% versus 27%; P=0.07).

**TABLE 3**
**Histological findings in esophageal biopsies from adult patients with surgically repaired esophageal atresia (n=12)**

| Histological finding | |
|---|---|
| Esophagitis | 8 (67) |
| Gastric metaplasia | 6 (50) |
| Intestinal metaplasia | 4 (33) |
| Dysplasia | 0 (0) |
| Adenocarcinoma | 0 (0) |

*Data presented as n (%)*

**TABLE 4**
**Manometric findings in adult patients with surgically repaired esophageal atresia (n=23)**

| Manometric finding | |
|---|---|
| Superior esophageal sphincter | |
| Normal | 23 (100) |
| Abnormal coordination and relaxation | 0 (0) |
| Esophageal body | |
| Normal | 0 (0) |
| Hypomotility | 23 (100) |
| Aperistaltism | 18 (78) |
| High amplitude waves | 3 (14) |
| Abnormal duration | 0 (0) |
| Lower esophageal sphincter | |
| Normal | 11 (48) |
| Hypotonia* | 4 (17) |
| Hypertonia | 1 (4) |
| Abnormal relaxation* | 8 (40) |

*Data presented as n (%). *One patient had hypertonia and abnormal relaxation*

**TABLE 5**
**Barium swallow findings in adult patients with surgically repaired esophageal atresia (n=29)**

| Barium swallow finding | |
|---|---|
| Normal | 16 (55) |
| Esophageal dilation | 8 (28) |
| Delay in esophageal emptying | 4 (14) |
| Esophageal diverticulum | 4 (14) |
| Esophageal stenosis | 2 (7) |

*Data presented as n (%)*

## DISCUSSION

In many studies, dysphagia is a common symptom in adults who undergo surgery for esophageal atresia (5,7). In our study, dysphagia was present in 73% of the cohort. To our knowledge, the present study was the first to investigate the causes of dysphagia in this population. We conducted a prospective study among adults followed in the esophageal atresia clinic using a standard questionnaire, gastroesophageal duodenoscopy, manometry and barium swallow.

The participation rate for endoscopy was 78%, which is high considering that some patients had already undergone an endoscopy during their pediatric follow-up and that this test was not prompted by a new symptom. In our study, the prevalence of histological esophagitis was 20%, which is slightly below the reported prevalence in adults who have undergone surgery for esophageal atresia (between 25% and 90%) (5). This could be explained by the fact that most of our population had undergone surgical antireflux therapy (37%) or were on antireflux medication (34%) compared with other studies (10% in the study by Sistonen et al [5]). There were no cases of cancer in our study, probably because of the young mean age of our population.

Manometry was performed in 56% of patients. All of the manometry findings in our study were abnormal. Hypomotility was demonstrated in 100% of patients and 78% had aperistalsis. The function of

the LES was abnormal in 52% and primarily related to incomplete relaxation (40%). The high prevalence of hypomotility caused by abnormalities intrinsic to the disease and surgical dissection during repair may, in part, explain dysphagia in adults who undergo surgery for esophageal atresia. Patients without dysphagia, despite dysmotility, are probably asymptomatic through adaptation (8).

Barium swallow was performed in 71% of participants and abnormalities were demonstrated in 45%. Abnormalities included dilations (28%), delays in esophageal emptying (14%), diverticula (14%) and stenoses (7%). Anatomical abnormalities were more common in patients with dysphagia than in those without dysphagia, although not a statistically significant difference (64% versus 27%; P=0.07). These anatomical abnormalities may play a role in the occurrence of dysphagia in this population (12) and the absence of a statistically significant difference was probably due to a lack of power.

## CONCLUSION

Dysphagia is a highly prevalent symptom in adults who undergo surgery for esophageal atresia. The present study was the first to demonstrate that motor and anatomical abnormalities could be implicated as causes of dysphagia in this population. Furthermore, these anomalies may be demonstrated with simple investigations such as endoscopy, manometry and barium swallow.

**DISCLOSURES:** The authors have no financial disclosures or conflicts of interest to declare.

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