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Long-term follow-up after esophageal replacement in children: 45-Year single-center experience

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ABSTRACT

Purpose: We evaluated the results of esophagocoloplasty (ECP) in children by performing a 45-year retrospective cohort study in a single center.
Materials and Methods: We collected all of the medical charts of patients who underwent ECP at our hospital from January 1969 to January 2014. We reviewed the medical history for each patient and then contacted the patients by phone to obtain responses to a questionnaire. The questionnaire evaluated the following four areas: overall general state, gastrointestinal function, dependence on the Health Sanitary System, and their aesthetic satisfaction.
Results: We contacted 96 patients, and 72 completed our questionnaire. There were 45 males and 27 female respondents, with a mean age of 34.5 years. The mean Karnofsky performance status index was 96.4%. There were excellent scores obtained from 55 patients and fairly good scores from 16 patients. There was 1 patient with a low score, and there were no scores reported below 70%. Most patients (58/72) reported regular bowel habits, and no patients required drugs or other measures to facilitate defecation. Gastroesophageal reflux was present in 51 patients, and 62.8% have bothersome symptoms. Twenty-five patients used pharmacological therapies. Forty-eight patients (66.6%) were satisfied with the aesthetic result of surgery (mean score in a scale from 0 to 5, is 3).
Conclusions: ECP can be used in children who require esophageal substitution. The resulting long-term QoL is acceptable. However, the aesthetic outcomes remain a problem.

Surgical reconstruction of the alimentary tract is used to restore the anatomy of children who have long gap esophageal atresia, severe caustic or peptic esophageal strictures, and various other esophageal disorders. The colon has been considered to be the best esophageal substitute in children for many years because it guarantees an adequate caliber and length. Unfortunately, there are only a limited number of publications investigating the long-term follow-up of colonic interposition [1]. We performed a retrospective cohort study to evaluate the results of ECP in patients treated in the last 45 years at a single center institution (Pediatric Surgery Department of Bologna).

1. Materials and methods

We reviewed all of the medical charts of patients who underwent ECP at the Department of Pediatric Surgery of the S. Orsola—Malpighi Hospital of Bologna from January 1969 to January 2014 and then assembled the patient medical history.

We then devised a questionnaire based on four areas of interest to estimate the effect of surgery on the quality of life (QoL):

1. The overall general state and functional outcome was assessed with the Karnofsky performance status (KPS) index as described by Burgos and colleagues [2] and Fernández-Sola et al. [4]. This index allows patients to be classified by their functional impairment. It is used to assess the prognosis and side effects of treatment in oncological patients and can be used to quantify patients' general well-being and everyday life activities. The Karnofsky score ranges from 100 to 0 and is described by intervals of 10 (see Table 1). A lower score indicates a worse survival rate. The KPS is both reliable and valid [3]. However, it is a unidimensional measure for the physical functional status and does not investigate all quality of life domains. Therefore, we decided to integrate the KPS index with other questionnaires (see the following areas) to obtain the maximum amount of information.
2. The gastrointestinal function assessment was based on the use of Gordon's functional Health Patterns (a guide based on the use of 11 categories that provide a systematic and standardized approach to data collection and a subsequent determination of aspects of health and human function), revised Rome III diagnostic criteria (a system developed to classify the functional gastrointestinal disorders) and gastroesophageal reflux disease Health Related Quality of Life Questionnaire (GERD-HRQL, a quantitative method of measuring symptom severity in gastroesophageal reflux disease). All of these assessments are valid, reliable and practical instruments [4–6]. We used elements from all of these questionnaires to create a valid and complete investigation tool.
3. The third area evaluates the dependence on the health system. We asked our patients about the number and type of medications that they use and their frequency of accessing health care facilities.

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Table 1
KPS content.

KPS content	
100%	Normal, no complaints, no signs of disease
90%	Capable of normal activity, few symptoms or signs of disease
80%	Normal activity with some difficulty, some symptoms or signs
70%	Caring for self, not capable of normal activity or work
60%	Requiring some help, can take care of most personal requirements
50%	Often requires help, requires frequent medical care
40%	Disabled, requires special care and help
30%	Severely disabled, hospital admission indicated but no risk of death
20%	Very ill, urgently requiring admission, requires supportive measures or treatment
10%	Moribund, rapidly progressive fatal disease processes
0%	Death

4. The last part of our questionnaire assesses aesthetic satisfaction through a numeric 0–5 scale evaluation (where 0 is a terrible result and 5 is an excellent result) and yes/no questions ("Are you satisfied with your appearance?" "Do you think that the opinion of others is important?" and "Have you ever felt discriminated because of your appearance?").

We constructed both a printed and online version (on GoogleDocs®) of the questionnaire. The patients were included after telephone contact and were invited for a clinical evaluation. Informed consent was obtained for all patients participating in the study.

The utility of Quality of Life scales, gastrointestinal function or self-image questionnaires is very limited at young ages. Thus, we decided to limit the results only to patients older than 16 years of age.

2. Results

We treated 133 patients and performed 141 esophageal replacements. All of the surgical procedures were performed by two experienced surgeons. The mean patient age at surgery was 54 months (range 12–208 months). The surgical indications are listed in Table 2. In cases of peptic and burn stenosis, an esophageal replacement was performed only after an unsuccessful cycle of dilatation. In three patients with portal hypertension, a total esophageal substitution was performed after failure of the portosystemic shunts. Table 3 describes the technical aspects of the surgeries performed. The most widely used procedure was retrosternal esophagocoloplasty. In the case of EA, we did not remove the distal esophageal stump. In other cases (38), esophagectomy was performed in 20 cases during the coloplasty, and in 5 cases, esophagectomy was performed after 2 months. There were 2 cases in which esophagectomy was performed before esophagocoloplasty (esophageal rupture due to chemotherapy and esophageal varicose vein in portal hypertension). Among the other 11 cases, 5 died from comorbidities and 6 have been entrusted to their centers of provenance with the recommendation to perform a deferred esophagectomy.

We did not use the left transverse colon in 19 patients (nine repeats and one unreliable left colic vascularization).

We believe that it is mandatory to place the intestinal bypass in the isoperistaltic direction. However, we used the antiperistaltic direction in one patient after multiple failures of previous esophageal replacements. In this case, the available tract was an ileocolic segment with a short vascular pedicle that made the isoperistaltic placement of the bypass impossible. The patients were discharged 15 days after the surgical procedure. Five patients died in the perioperative period between 1970 and 1981.

Table 2
Indications for surgery.

Indication	Number
Long gap EA with or without fistula	95
Peptic stenosis in GER	18
Burn stenosis	13
Portal hypertension	3
Congenital brachioesophagus	2
Diffuse esophageal leiomyomatosis	1
Postchemotherapy esophageal rupture	1

EA: esophageal atresia; GER: gastroesophageal reflux.

Table 3
Surgical details.

Surgical details	Number
Complete substitution	126
- Retrosternal	122
- Intrathoracic	4
- Redos	10
Segmental substitution	15 (all intrathoracic)
Left transverse colon (left colic vessels)	122
Right transverse colon (medial colic vessels)	8
Ileocecum (ileocolic vessels)	11

The complications are listed in Table 4 and are divided into 2 groups based on the necessity of surgery. Peptic complications occurred in the first few years of our experience before the introduction of an antireflux anastomosis.

We contacted 96 patients and found the following results: 9 died due to comorbidities, 8 did not want to participate, and 7 were excluded because the patient was less than 16 years of age. Thus, 72 patients completed our questionnaire. The mean follow-up was 345 months (range 180–552 months). The demographic data of the 72 patients are the following: the female/male ratio is 27/45, the mean age is 34.5 years (range 16–49) and 11 patients are married.

We report our results grouped in the four examined areas.

First area Overall general state and functional outcome—"Karnofsky performance status"

As far as working activity is concerned, 64 patients (88.9%) did not feel that surgery influenced their daily work activity, and 4 patients (5.6%) were able to perform normal work activities with minor difficulties. There were 3 patients (4.1%) that complained it took some effort to work, and 1 patient (1.4%) was unable to work. The last two categories ($n = 4$) included the following patients: one syndromic patient with 22q11 microdeletion, one patient affected by recurrent pulmonary infections, one patient retreated with surgery after an anti-peristaltic ECP performed at another hospital, and one patient disabled for reasons not related to ECP.

Regarding daily life activity, 60 patients (83.3%) had no limitations and 7 patients (9.7%) had minor difficulties. There were 4 patients (5.6%) who conducted normal activities with effort, and 1 patient (1.4%) was unable to perform normal activity.

Concerning personal care, 71 patients (98.6%) are autonomous and 1 patient (1.4%) has difficulties caring for himself. Thirty-six patients (50%) complained of clinical symptoms.

Table 4
Surgical complications.

Minor complications: number
- Cervical anastomosis leak: 9
- Wound infections: 2
- Segmental stenosis: 1
Major complications: number; surgical treatment
- Bypass necrosis: 8; redos
- Intestinal obstruction: 7; adhesiolysis
- Peptic diseases: 3; removal of the ulcerated colon and jejunal interposition
- Cervical anastomosis stenosis: 3; resection and reanastomosis
- Bypass stenosis: 1; redo