Long-Term Follow-Up and Quality of Life After Gastric Transposition

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Introduction

While we certainly subscribe to the principle that "the child's own oesophagus is best" and that the oesophagus can be preserved in a majority of cases of oesophageal atresia [1–3], we remain concerned that in some cases repeated attempts to preserve the oesophagus may be to the detriment of the child and that their own oesophagus may be a liability. In many of these children, their entire infancy and early childhood have been dominated by endless attempts to preserve the native oesophagus at all costs. Replacement of the oesophagus represents an irreversible decision to abandon further attempts at salvage of the oesophagus.

The ideal oesophageal substitute should function as closely as possible to the original structure. The patient should be able to swallow normally, consume normal amounts, and should not experience any reflux symptoms. An additional requirement in children is that the substitute should continue functioning for many years without deterioration.

Satisfactory results have been reported for all forms of oesophageal replacement [4], although the numbers reported are mostly small and long-term data are scanty.

At Great Ormond Street Hospital for children, in London in the past 25 years, we have used gastric transposition almost exclusively for oesophageal substitution. One-hundred and ninety-two infants and children underwent gastric transposition for oesophageal substitution [5]. There were 116 male and 76 female patients undergoing the procedure at a median age of 2 years (range 7 days to 17 years).

The indications for oesophageal replacement are shown in Table 53.1. Ninety-four patients were referred from centres abroad (49 %), and 62 from centres within the United Kingdom (32 %), while the remaining 36 (19 %) received all their treatment at Great Ormond Street Hospital. In total, 156 (81 %) of our patients were referred for their replacement from other centres.

A prior colonic interposition had been unsuccessful in 17 patients, six had a partial gastric transposition, three each had had a Scharli-type procedure [6] or a reversed gastric tube oesophagoplasty, and one child had a failed jejunal interposition. Previous extensive surgical attempts to retain the original oesophagus had been carried out in a total of 69 (36 %) patients.

The method of replacement [7, 8] was via the posterior mediastinum using blunt dissection in 98 patients, while 90 patients required an additional lateral thoracotomy due to extensive mediastinal fibrosis secondary to the original injury (caustic, perforation) or to previous

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Table 53.1 Indication for oesophageal replacement

Esophageal atresia	138
With distal tracheo-oesophageal fistula	76
Isolated atresia	48
With proximal fistula	12
H-fistula	2
Caustic stricture	29
Peptic stricture	9
Other	16
Achalasia	2
Laryngeal cleft	2
Congenital amotile oesophagus	2
Congenital stenosis	3
Congenital short oesophagus	1
Prolonged foreign body impaction	2
Diffuse leiomyoma	2
Inflammatory pseudo-tumour	1
Teratoma	1

attempts at oesophageal reconstruction. The stomach was placed in the retrosternal position in four patients, who previously had a failed colonic interposition placed in that site. A jejunal feeding tube was routinely inserted in patients who had not previously fed orally. A transanastomotic nasogastric tube was left in the intrathoracic stomach to provide postoperative gastric decompression. All patients with the exception of the first nine in the series were electively paralysed and mechanically ventilated for varying periods postoperatively.

Mortality: There were nine deaths in the series, a mortality rate of 4.6%. One child died intraoperatively from uncontrollable haemorrhage, five died in the early postoperative period from either respiratory (4) or cardiac (1) failure and three died over a year postoperatively, all of respiratory causes. Eight of these children had had complex courses prior to the transposition.

We believe that mortality can be reduced by submitting patients to oesophageal substitution earlier and refraining from endless attempts at oesophageal salvage. It is easy to become unduly focussed on saving the oesophagus at all costs, but repeated attempts at oesophageal salvage will substantially increase the operative difficulty encountered at the time of substitution procedure.

Anastomotic leakage at the oesophagogastric anastomosis in the neck occurred in 23 patients (12%), all except one of which closed spontaneously. The one child with a major disruption had a cervical oesophagostomy re-established. Secondary anastomosis was carried out uneventfully 6 months later. Four of these patients had undergone previous unsuccessful oesophageal replacement procedures (two colonic and two partial gastric transpositions), and nine had had multiple procedures carried out previously in an attempt to preserve their original oesophagus.

Anastomotic strictures developed in 40 patients (20%) all but three responding to endoscopic dilatations. In the three requiring stricture resection, the procedure was successfully completed via a cervical approach. In 17 cases, the original pathology was caustic oesophageal injury. Five children had previously undergone a colonic interposition.

Significant swallowing problems were encountered postoperatively in 55 patients (29%) half of whom had prolonged difficulties. Eighteen of these children had had major swallowing problems prior to the gastric transposition. The importance of sham feeding in maintaining a normal swallowing mechanism in infants having a cervical oesophagostomy for isolated oesophageal atresia where primary, or delayed, anastomosis is impossible cannot be overemphasised. The feeding difficulties can persist for many months during which enteral nutrition is provided by jejunal feeds, but improvement gradually occurs. It is important to persist with attempts at oral feeding and to try different consistencies of food. In the long term, the great majority of patients can eat and swallow normally. Although many prefer small frequent meals, those who have undergone oesophageal replacement in later childhood report a normal feeling of satiety after eating [2].

Respiratory problems: Reflux into the cervical oesophagus is common in the early months after gastric transposition. This can lead to regurgitation and aspiration especially in the recumbent position resulting in coughing episodes. It is recommended that the infant be propped up in bed and the older child sleeps on two or three cushions. The reflux may cause

mild oesophagitis for which an antacid should be prescribed.

Severe delay in gastric emptying occurred as a late complication in 16 (8.3 %) patients. Included among this group were three infants in whom an original pyloromyotomy was converted to a pyloroplasty and two who required a Roux-en-Y gastrojejunostomy. The delay in gastric emptying may be responsible for halitosis experienced by a few patients.

Dumping symptoms is frequently experienced in the early weeks postoperatively and usually responds well to simple measures such as small frequent meals, avoidance of sugar in the diet, the addition of starch as the main source of carbohydrate and separating the solid and liquid components of a meal. Dumping as a long-term problem only occurred in 2 % of cases but all eventually resolved.

Seven patients experienced problems with the jejunal feeding tube comprising leakage into the peritoneal cavity following traumatic re-intubation, volvulus, intussusception, internal fistula and adhesion obstruction.

Other complications included three infants with severe tracheomalacia, two of whom required aortopexy, two vocal cord paresis requiring temporary tracheostomy, two chylous effusions, two transient Horner's syndrome and one postoperative haemorrhage requiring re-thoracotomy.

The long-term outcome was considered excellent if the child had normal eating habits with the absence of symptoms. The result was considered good if the child had occasional dysphagia or had an altered eating habit such as a preference for a small, frequent meal. In 90 % of our patients, the long-term outcome was considered good to excellent in terms of the absence of swallowing difficulties or other gastrointestinal symptoms such as dumping or diarrhoea. Many patients prefer to eat small frequent meals. Unsatisfactory long-term outcome was present in eight patients (4.6 %), three of whom had chronic respiratory problems (CHARGE syndrome, laryngeal cleft, recurrent pneumonia). A poorer outcome was particularly associated with multiple previous attempts at oesophageal salvage. There was no evidence of deterioration in the function of the gastric transposition in 72 patients followed up for longer than 10 years.

Long-term nutritional and respiratory function [9]: Although the few children tested have a measurable respiratory compromise, they are generally asymptomatic.

The mean total lung capacity in one of our studies was around 68 % and forced vital capacity 64 % of expected. The ratio of forced expiratory volume in one second to the forced vital capacity was 87 %. This was not a longitudinal study, and therefore, it was not possible to determine whether the intrathoracic stomach was detrimental to lung growth. Of interest was that patients undergoing an uncomplicated primary gastric transposition had greater lung volumes than those subjects having a more complicated course involving multiple thoracotomies. We are aware that a bulky stomach in the chest of a child with borderline lung function may be a problem, and under these circumstances, gastric transposition may not be the optimal oesophageal substitute.

While most of our patients were in the lower centiles for weight, their heights remain within the normal range; we were unclear if this was related to their underlying problem or to the operation. Children who had caustic injury followed their previous percentiles.

Gastric emptying studies have shown that more than 50 % of both solid and liquid components of a test meal had left the stomach by the time of completion of the meal indicating that the transposed stomach act as a conduit rather than a reservoir. There is no correlation between diarrhoea or dumping symptoms and gastric empting.

It remains to be determined whether Barrett's metaplasia in the proximal oesophagus will be a longer-term problem. We have not encountered this so far but are aware of the problem when gastric tubes are used [10]. As the stomach has been vagotomised, the amount of acid produced may be insufficient to induce metaplasia.

We remain unsure of the best approach for those with CHARGE association, complete laryngeal clefts and caustic injuries to the upper

oesophagus and pharynx, as bolus gastrostomy feeds may be necessary in the long term in these children. Colonic interposition may be a better option under these circumstances.

We are encouraged that at least after the first two decades, there is no symptomatic deterioration in function of the transposed stomach.

Gastric transposition has replaced colonic interposition as the oesophageal replacement procedure of choice in many centres [5, 11, 12]. The excellent blood supply of the stomach, the fact that only one anastomosis is required and the relative technical ease of the procedure are clear advantages. In addition, the long-term follow-up of our patients has shown good growth and development and that the function of the replacement continues to be satisfactory in the immediate future.

Quality of Life Following Gastric Transposition

Previous research had shown that a significant proportion of children who underwent major neonatal surgery for life-threatening congenital abnormalities experienced psychosocial problems during childhood and early adolescence [13–16]. Since the outcome for children undergoing oesophageal replacement with gastric transposition for failed repair of oesophageal atresia has not previously been reported, we carried out a descriptive study in 2002 to assess in-depth functional outcome, psychosocial adjustment and health-related quality of life of patients following gastric transposition (GT) [17].

The rarity of the problem and the fact that many patients originated from a wide geographical area meant that our sample size was small. All the patients were resident in the United Kingdom and had undergone gastric transposition at Great Ormond Street Hospital for Children in London. Based on their operative history before gastric transposition, the 28 patients were divided into two groups. Group 1 consisted of 13 patients for whom gastric transposition was the primary reconstructive surgical procedure. The 15 patients in group 2 had undergone attempts at oesophageal repair or replacement, which had failed. Over two thirds of the patients in group 1, and more than half of those in group 2, had associated anomalies.

The total number of operative procedures for these 28 patients, including those performed in other institutions and those relating to procedures for associated anomalies, ranged from 2 to 91 (mean, 24 ± 22). The mean number of operative procedures in group 1 was 14 (SD, 15), and 32 (SD, 25) in group 2. The difference between the groups was significant ($p < .05$). In group 2, the mean number of operative procedures after gastric transposition was 19 (SD, 21) compared with a mean of 2 (SD, 2) in group 1 (Table 53.2).

Four patients (two in each group) were below school age, and eight patients (three in group 1,

Table 53.2 Patients' characteristics

	Group 1 $n = 13$	Group 2 $n = 15$
	Mean (SD)	Mean (SD)
AGE (years)	13 (5)	13 (6)
GA (weeks)	36 (3.4)	35 (3)
BW (kg)	2.14 (.54)	2.39 (.89)
Number of patients (%) with associated anomalies	9 (69)	8 (53)
None	4 (31 %)	7 (47 %)
Significant[a]	7 (54 %)	6 (40 %)
Cardiac	2	3
VATER	4	2
Sensory deficit	2 (15 %)[b]	2 (13 %)[c]
Age at GT (decimal yrs)[d]	0.95 (0.6)	4.56 (5)
Time since GT (decimal years)	12.26 (5.34)	8 (7)
Total no. of all operative procedures[b]	14 (15)	32 (25)
Related to oesophagus before transposition	1	19 (21)
Related to oesophagus after transposition	2 (2)	5 (7)
Body mass index: weight (kg)/length (m)^2 (z scores adjusted for age and sex)	−1.67 (0.98)	−1.70 (1.10)

[a]Includes conditions such as Fanconi anaemia (group 1), trisomy 21 (group 2)
[b]1 with bilateral anophthalmos and cerebral palsy; 1 congenitally blind
[c]2 with profound deafness
[d]$p < 0.05$

five in group 2) had left school. One was at university, three are at college, three were employed, and one had recently given birth to a healthy boy. Of those at school, five patients were in a special unit or in special schools, and four required special needs within normal schools (9/17, 53 %); in addition, two of the older patients had been in a special unit, and two had moderate learning difficulties (13/28, 46 %). With one exception, all the older patients, including the young mother, were still living with their parent(s).

The design of the study included a clinical review, in-depth interviews with patients and their parents and the use of self-report standardised questionnaires. For psychosocial outcomes, we used the Child Behaviour Checklists (Achenbach [18]): a questionnaire completed by parents (CBCL for patients aged 2–18 years), the parallel Teacher Report Form (TRF for patients aged 4–18 years) and the Youth Self-Report Form (YSR for patients aged 11–18 years). These Achenbach questionnaires [18–21] are designed to measure competencies and behavioural/emotional problems as seen by parents, teachers and youth, respectively, and make it possible to compare data from different respondents on a common set of problem items and scales. They have been used extensively in child/adolescent mental health research. Reliability and validity are well established. Health-related quality of life (QOL) was measured using a modified version of the Eypasch Gastrointestinal Index (GIQLI) [19, 22].

Behavioural and Emotional Outcome

The overall mean scores based on the parents' and teachers' report were similar to the norms. However, the distribution of the individual scores indicated a significant proportion of the patients in group 2 with scores in the clinical range. For example, based on the parents' report, only one (1/10, 10 %) patient in group 1 had scores in the clinical range on the total problem score, compared with 3/12 (25 %) patients in group 2 on both the total problem and internalising scales; externalising disorder scores were low in both groups. The teachers' views of their pupils differed from that of the parents. Although overall mean scores were similar to the normative data, the distribution of the individual scores indicated a significant proportion of the patients in both groups with scores in the clinical range. Depressive symptoms were foremost among patients in group 1 (3/7 43 %) and externalising disorders among patients in group 2 (3/7 43 %) [20]. The parents' and teachers' findings did not appear to be related to the presence of associated anomalies or the length of time since GT.

Twelve patients completed the Youth Self-Report Form—six in each group—and, with only one exception, they all rated themselves as functioning well within the normal range.

To sum up, based on these findings, a significant proportion of patients who underwent GT, after other procedures had failed, were judged by the adults who knew them well to have psychosocial adjustment problems. Disturbed behavioural adjustment, principally internalising disorders, was noted by teachers for a proportion of the patients in group 1.

Health-Related Quality of Life Outcomes

Nineteen patients aged between 10 and 22 years, ten in group 1 and nine in group 2, completed the questionnaire. The only difference between the groups was on disease-specific symptoms. For example, fewer patients in group 1 experienced dysphagia (30 % vs. 67 %) or pain after eating (20 % vs. 33 %), compared with patients in group 2. Similarly, a smaller proportion of patients in group 1 had gastro-oesophageal reflux symptoms such as heartburn or regurgitation during the day or at night compared with those in group 2 (40 % vs. 67 %). Some breathlessness was experienced during the day by over half the patients in each group, but breathlessness at night was more frequent in group 2. Differences between the groups were not related to the length of time since gastric transposition. These data were supported by the parents' perception of the QOL of their children. Based on parental responses, patients in group 1

experienced fewer disease-specific symptoms such as dysphagia, dumping symptoms and pain after eating. In addition, with the exception of psychological and physical/social symptoms, parents in group 1 perceived the health-related QOL of their children to be significantly better than did parents of patients in group 2.

QOL of the Young Children

The parents' responses to the questionnaire showed that they perceived the overall QOL of the five young patients, aged 2–4 years, especially for those in group 2, to be adversely affected by difficulties relating to all aspects of eating—their enjoyment of food, restrictions in types of food they could eat and the amount they were eating. However, these problems are often reported about healthy children of this age. One patient in each group (50 % vs. 33 %) experienced dysphagia, and one child (50 %) in group 1, compared with two in group 2 (67 %), experienced pain after eating. Both patients in group 1 and one patient in group 2 (33 %) were reported to have gastro-oesophageal reflux symptoms. Similar proportions experienced some breathlessness during the day. Almost half of this small group of young children (44 %) had associated anomalies, and this was an important factor affecting their lives.

Physical Characteristics

The physical growth characteristics of the 28 patients showed that, with the exception of one patient in each group, all the patients were below the 50th centile for weight, but five patients in group 1 (41 %) and two (12 %) in group 2 were above the 50th centile for height. When the standardised body mass index (z scores), adjusted for age and gender, was calculated, all the patients had a BMI below zero, ranging from –0.10 to –3.91.

Conclusion

In this study, we examined the psychological adjustment and the QOL of a small group of patients following GT for repair of long-gap oesophageal atresia. Based on the responses of the adults who knew them well, we found that psychological maladjustment was more prevalent among the patients who had unsuccessful reconstructive surgery prior to GT. Additionally, the parents of these patients viewed their overall QOL as less satisfactory compared with those for whom GT was the primary reconstructive procedure. They were said to cope less well when eating and experienced a greater number of symptoms such as dysphagia, regurgitation and dumping symptoms. The patients themselves reported greater difficulty swallowing. However, in contrast to their parents' perceptions, they viewed their overall quality of life and psychological adjustment as normal.

With one exception, all the patients and the families in this study reported that they were extremely satisfied with the outcome following gastric transposition. Based on the interviews with the patients and their parents, the patients without debilitating conditions led relatively normal lives, and many enjoyed sporting activities. However, they tended to be less socially and emotionally independent than their peers.

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