Cricopharyngeal myotomy in motor neurone disease

L A LOIZOU, M SMALL, AND G A DALTON

From the Departments of Neurology, and Ear, Nose and Throat Surgery, Queen Elizabeth Hospital, Birmingham

SUMMARY Twenty-five patients with dysphagia caused by neurological disorders, mainly motor neurone disease, underwent cricopharyngeal myotomy. Nineteen patients showed slight to dramatic improvement of swallowing for variable periods of time. There were five postoperative deaths. The results indicate that this simple procedure is of benefit to a substantial proportion of patients with neurological causes of dysphagia.

The development of dysphagia is one of the most distressing aspects of motor neurone disease as it leads to malnutrition in an alert patient and predisposes to attacks of choking and aspiration of food, with fatal consequences. Management is problematical, not least because of the ethical considerations posed by a fatal disease. Nutrition can be maintained in a number of ways.^{1–3} Liquid food can be given via a nasogastric tube, but this is often found to be uncomfortable and does not overcome the problem of pooling of saliva in the mouth and hypopharynx. Cervical oesophagostomy can be performed.^{3,4} This results in a cutaneo-oesophageal fistula, and pureed food is introduced into its origin by a syringe. Gastrostomy is seldom advised. Cricopharyngeal myotomy was first used by Kaplan in 1951⁵ in a patient with bulbar poliomyelitis and has been advocated by Mills^{6,7} in the United Kingdom, and Lebo *et al*⁸ in the USA.

The rationale for this procedure is based on the observation that the cricopharyngeus muscle behaves as a true sphincter and exhibits tonic contraction in the resting state.^{9,10} When a bolus of food is advanced towards the sphincter, the cricopharyngeus first relaxes to allow the bolus to enter the oesophagus. It then contracts to propel the bolus, and finally returns to its resting state of tonic contraction. In patients with bulbar palsy the sphincter fails to relax adequately. Section of the cricopharyngeus overcomes this mechanical obstruction, thus allowing food to enter the oesophagus under the influence of

gravity and minimising the danger of aspiration. The procedure has been applied successfully to patients with a variety of conditions leading to bulbar palsy.^{7–12}

Since 1973 the Department of Ear, Nose and Throat Surgery at the Queen Elizabeth Hospital, Birmingham has been providing the facility for cricopharyngeal myotomy in patients with neurological causes of dysphagia. The results of this procedure are presented in this report.

Patients and Methods

Between June 1973 and December 1978, 28 patients underwent cricopharyngeal myotomy. Three patients were excluded from this study because of inadequate documentation of pre-operative diagnosis and follow-up. The hospital records of the remaining 25 patients were reviewed. Where possible patients were seen by the authors. Alternatively additional information was obtained from the general practitioner. All but one had been investigated at a neurological unit in Birmingham before referral for surgery. The preoperative diagnosis was motor neurone disease in 20 patients, pseudobulbar palsy (stroke) in two patients, dystrophia myotonica in two patients, and multiple sclerosis in one patient.

The operative procedure was standardised and carried out under general anaesthesia.⁷ A nasogastric tube was inserted interoperatively, and maintained for two to four days postoperatively, while the patient was being changed to a near normal diet. Patients were discharged from hospital where possible on the seventh or eighth postoperative day. The table gives details of the patients and their outcome. All but two patients

Address for reprint requests: Dr L A Loizou, The Midland Centre for Neurosurgery and Neurology, Holly Lane, Smethwick, Warley, West Midlands B67 7JX.

Accepted 8 June 1979

Table Clinical details and outcome of 25 patients who underwent cricopharyngeal myotomy

Patient	Sex	Age (yr)	Length of history to operation (MO)			Respiration and coughing at operation	Diagnosis	Outcome
			Dysphagia	Dysarthria	Limb weakness			
MJ	F	57	6	6	12	Unimpaired	MND	Slight improvement. Died 14 mo after onset of MND, 5 mo after operation
GA	F	64	5	7	15	Unimpaired	MND	Improved. Died 2 yr after operation, 3 yr 3 mo from onset of MND
TP	M	63	26	26	12	Unimpaired	MND	Improved. Died 4 mo after operation, 2½ yr from onset of MND
RG	M	63	12	12	Nil	Impaired	MND	Improved. Died approx 1 yr after operation, 2 yr from onset of MND
HB	M	68	19	13	12	Unimpaired	MND	Improved for at least 2 mo. Further follow-up not available
HB	M	76	4	4	21	Impaired	MND	Improved. Died 1 mo after operation (pulmonary oedema) approx 2 yr from onset of MND
LJ	F	61	6	Nil	45	Unimpaired	MND	Improved. Died 8 mo after operation 4 yr 7 mo from onset of MND
IW	F	63	28	28	3	Unimpaired	MND	Improved. Died 1 yr 10 mo after operation, 4 yr 6 mo from onset of MND
NR	F	68	2	28	28	Unimpaired	MND	Improved. Died 1 yr 7 mo after operation, 4 yr from onset of MND
ME	F	63	10	10	6	Unimpaired	MND	Improved temporarily. NG tube required 7 mo postoperatively. Requiring continuous care
GH	F	24	18	18	36	Unimpaired	MND	Improved. Still alive 1 yr after operation
ChM	M	57	5	5	Nil	Unimpaired	MND	Slight improvement only. Still alive 8 mo after operation
GT	F	76	3	8	Nil	Unimpaired	MND	Alive to date. Improved for 4 mo but then gradual deterioration to swallowing fluids only 8 mo after operation
EEF	F	59	7	7	Nil	Unimpaired	MND	Improved at 2 mo but deteriorated 6 mo postoperatively. "Just managing" to date
SB	M	70	7	7	Nil	Unimpaired	MND	Improved temporarily. Died 3 mo after operation, 10 mo from onset of MND.
EJ	M	69	7	9	—	Impaired	MND	Died 36 hr after operation from respiratory failure, 9 mo from onset of MND
LP	M	62	2	2	14	Impaired	MND	Died 7th postoperative day from respiratory failure, 14 mo from onset of MND
JA	M	64	4	12	16	Impaired	MND	Died 7th postoperative day from respiratory failure, collapsed lung, 16 mo from onset of MND
WR	M	60	12	12	36	Impaired	MND	Died 14th postoperative day, 3 yr from onset of MND
BB	F	64	30	30	3	Impaired	MND	Died 10th postoperative day from respiratory failure, 2 yr 6 mo from onset of MND
ES	F	70	24	24	36	Recurrent chest infections	Pseudobulbar palsy. Right hemiparesis and left hemiplegia	No change. Still alive 3 mo after operation
RT	M	66	1½	1½	Mild right hemiparesis	Chronic bronchitis	Brainstem stroke	Improved from total aphagia to swallowing solids and liquids within days. Died 15 mo after operation
DM	F	43	12	12	27	Impaired	Dystrophia myotonica	Improved. Died 2 yr 3 mo after operation
JP	F	25	24	Nil	48	Impaired	Dystrophia myotonica	Improved. Still alive 6 yr after operation
CP	F	21	10	30	30	Recurrent chest infections	Multiple Sclerosis	Improved. Died 4 mo postoperatively, 2 yr 10 mo from onset of Multiple Sclerosis

MND = motor neurone disease; NG tube = nasogastric tube

had dysarthria associated with their dysphagia, which was manifested in prolonged eating time, inability to clear secretions from the mouth, and choking attacks both from accumulation of saliva and attempts to swallow food. All the patients with motor neurone disease had weakness and fibrillation of the tongue. Significant weight loss had occurred in 15 of the patients at the time of operation. Systemic disease coexisted with the primary pathology in some patients. Three had ischaemic heart disease, two patients had left ventricular failure, two had dementia, and one each had diabetes mellitus, myxoedema, and hypertension.

Results

There were 11 male patients with a mean age of 63.4 years (range 57–76 yr), and 14 female patients with a mean age of 54 years (range 21–76 yr). The two patients with dystrophia myotonica showed considerable improvement in their swallowing. One is alive to date, the other survived for 27 months after operation and died of bronchopneumonia at the age of 45 years. The patient with severe brainstem demyelination improved after operation and was able to swallow food and dispose of the nasogastric tube. She died four months after operation from recurrent chest infections. The patient with pseudobulbar palsy had suffered a right-sided hemiparesis and a left-sided hemiplegia three and two years respectively before operation. She showed no improvement in her swallowing, but was still alive three months postoperatively. The patient with brainstem stroke, on the other hand, benefited from the operation.

Five patients with motor neurone disease died in hospital, from 36 hours to 14 days after operation. This gives an overall mortality figure of 20%. All these patients had moderately severe impairment of respiration, and all had impairment of coughing and expectoration in addition to severe dysphagia and dysarthria. The remaining 15 patients with motor neurone disease showed some degree of sustained improvement in their swallowing, lasting from one month to two years, with a mean of nine months. One of these patients had section of the chorda tympani nerve to control sialorrhoea three months after myotomy, and had to revert to a nasogastric tube four months later. Only two of these 15 patients had respiratory insufficiency at the time of operation, and survived one month and one year postoperatively. The average duration of motor neurone disease from first symptoms to

Cricopharyngeal myotomy in motor neurone disease

death in patients of this series was two years and seven months, with a range of seven months to four years and seven months.

Discussion

The results of this study show that patients with motor neurone disease, and other neurological conditions causing dysphagia can be helped by the simple procedure of cricopharyngeal myotomy. This is more apparent if the results are viewed in the light of the natural history of the disease causing dysphagia, and the quality of life of the patient at the time of operation. In 19 of the 25 patients there was a successful outcome, with slight to substantial improvement in swallowing. Improvement was assessed on subjective grounds, as well as on observation of the patients during the postoperative period. Routine postoperative barium swallow studies were not carried out.

It is of interest that sustained improvement was produced in the two patients with dystrophia myotonica, an observation also made by Johnson and Kuwabara¹³ in patients with oculopharyngeal muscular dystrophy. There was also dramatic improvement in one patient with brainstem stroke and in the patient with brainstem demyelination.

The operative mortality in this series was 20%. Mills⁷ reported one death in 23 patients, and Lebo *et al*⁸ reported three deaths in 38 patients. The most important factor contributing to the postoperative deaths in this series was felt to have been the patients' severe limitation of coughing and ventilatory capacity. In future this may be avoided by advising myotomy at an earlier stage in the disease when respiratory involvement is limited. Even in these circumstances, however, the liability of the patient with bulbar palsy to inhale small amounts of secretions, and to develop laryngeal spasm, will result in deaths especially during sleep.

With a progressive disease such as motor neurone disease it is inevitable that eventually the muscles of the tongue and hypopharynx will become so weak that, even with a cricopharyngeal myotomy, swallowing will be very difficult. Until this happens, however, the patient with a myotomy can have a long period of fairly easy swallowing.

We would like to thank Mrs E Simpson and Miss M Fitton for preparation of the manuscript.

References

- 1 Editorial. Management of motor neurone disease. *Br Med J* 1976; 1:1422–3.

- 2 Editorial. Symptomatic care in motor neurone disease. *Br Med J* 1976; **2**:605-6.
- 3 Smith RA, Norris FH. Symptomatic care of patients with amyotrophic lateral sclerosis. *JAMA* 1975; **234**:715-7.
- 4 English GM, Mason Morfit H, Ratzer ER. Cervical oesophagostomy in head and neck cancer. *Arch Otolaryngol* 1970; **92**:335-9.
- 5 Kaplan S. Paralysis of deglutition, a post-polio-myelitis complication treated by section of the cricopharyngeus muscle. *Ann Surg* 1951; **133**: 572-3.
- 6 Mills CP. Dysphagia in progressive bulbar palsy relieved by division of the cricopharyngeus. *J Laryngol Otol* 1964; **78**:963-4.
- 7 Mills CP. Dysphagia in pharyngeal paralysis treated by cricopharyngeal sphincterotomy. *Lancet* 1973; **1**:455-7.
- 8 Lebo CP, Ü SK, Norris FH. Cricopharyngeal myotomy in amyotrophic lateral sclerosis. *Laryngoscope* 1976; **86**:862-8.
- 9 Lund WS. The cricopharyngeal sphincter: its relationship to the relief of pharyngeal paralysis and the surgical treatment of the early pharyngeal pouch. *J Laryngol Otol* 1968; **82**:353-67.
- 10 Blakeley WR, Garety EJ, Smith DE. Section of the cricopharyngeus muscle for dysphagia. *Arch Surg* 1968; **96**:745-60.
- 11 Mills CP. Cricopharyngeal sphincterotomy and bilateral division of the chorda tympani in bulbar palsy. *Proc R Soc Med* 1975; **68**:644-6.
- 12 Ü KS, Norris FH, Denys EH, Lebo CP. Surgery in patients with amyotrophic lateral sclerosis. Experience with cricopharyngeal myotomy in 100 cases. (abstract). *Excerpta Medica International Congress Series* 1977; **427**:45.
- 13 Johnson CC, Kuwabara T. Oculopharyngeal muscular dystrophy. *Am J Ophthalmol* 1974; **77**:872-9.