STAGED SURGERY IN OESOPHAGEAL ATRESIA

B. GAMPEL, F.R.C.S. (ENG.), *Department of Paediatric Surgery, Baragwanath Hospital and the University of the Witwatersrand*

The first successful repair of an oesophageal atresia was achieved by Haight in 1941, who divided the fistula and performed a primary oesophageal anastomosis.[1] This direct approach has become the standard form of treatment for the condition and its use has led to survival rates of over 90% in full-term, otherwise healthy, infants.[2]

However, only a minority of patients with oesophageal atresia are in this fortunate category. Most cases are complicated by pulmonary infection, prematurity or an additional congenital abnormality. These factors are so important to the prognosis that over-all survival in large series is seldom better than 65%[3] and may be as low as 6% in cases where the complications are of a severe degree.[2]

There is an increasing awareness in paediatric surgical centres that these complications are not entirely 'uncontrollable' and that if treatment is directed towards correcting them before undertaking major surgery, survival may be enhanced. This may require delaying of oesophageal repair in some cases or a return to the multiple-stage methods of Leven[4] and Ladd[5] in others. The problems concerned are complex and only constant reappraisal of methods and results will provide a basis for individualized management.

Sixty patients with oesophageal atresia, admitted over a 10-year-period, comprise the present series. Certain aspects of the mortality in the series will be discussed and an attempt will be made to define the indications and technique of surgical staging in the management of oesophageal atresia.

Transvaal Memorial Hospital Series

Between the years 1955 and 1964, 60 patients with oesophageal atresia were admitted to the Transvaal Memorial Hospital for Children (TMH), Johannesburg. The survival rate of 37% (22 patients) reflects the difficulties that have been encountered. That these difficulties are not being overcome by modern progress in neonatal care is shown by the fact that the survival rate dropped from 48% in the first 4 years of the series (14 out of 29 cases) to 26% in the latter 6 years of the series (8 out of 31 cases).

Types of Defect Encountered (Fig. 1)

Of the cases encountered, 54 of the 60 were of the 'usual' type—with atresia of the oesophagus commencing at D2 - D4 and a fistulous connection between the lower oesophageal pouch and the trachea (or right main bronchus).

Three patients had atresia without fistula.

Two patients had atresia with double fistula.

There was one case of 'H' fistula with oesophageal stenosis.

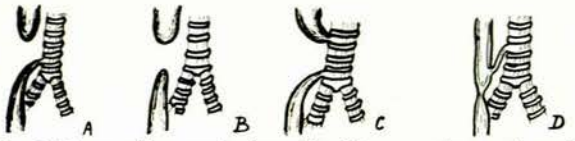

Fig. 1. Types of anomaly found in 60 cases of oesophageal atresia.
A. 'Usual' type —atresia with lower-pouch fistula—54 cases (90%).
B. Atresia with no fistula—3 cases (5%).
C. Atresia with double fistula—2 cases (3%).
D. 'H' fistula with lower-pouch stenosis—1 case (2%).

MORTALITY

The mortality of the TMH series will be discussed under the following headings:

1. Delay before admission.
2. Delay between admission and primary repair.
3. Birthweight.
4. Additional anomalies.
5. Staged procedures.

1. *Delay Before Admission*

The striking difference in survival between those cases where the diagnosis was made within the first 3 days of life (44% survival) and those in whom the diagnosis was delayed for more than 3 days (8% survival) is shown in Table I.

TABLE I. RELATIONSHIP OF SURVIVAL TO AGE AT DIAGNOSIS, TO PREVALENCE AND SEVERITY OF PULMONARY INFECTION AND TO PREMATURITY

Age at diagnosis	*Number*		*Under 5 lb.*	*Chest Signs* 'Clear'	'Moderate'	'Severe'
1st day	21 (11)	} (44%)*	2	7	12	3
2nd day	15 (6)		4 (1)	4	8	2
3rd day	12 (4)		6	2	6	4
4th day	7 (1)	} (8%)*	2	1	3	3
5th day	5 (0)		0	0	3	2
Total	60	(22–37%)*	14	14 (10–70%)*	32 (12–38%)*	14 (0)*

* Survivals.

Infection. The deciding factor in the mortality appeared to be the presence of pulmonary infection—more prevalent and severe the longer the delay in diagnosis. The over-all incidence of pulmonary infection was high (over 70%) and 'lipiodol pneumonitis' was noted in many cases.

That pulmonary infection is the principal cause of death in oesophageal atresia is unquestioned. De Borer and Potts found it to be the cause of nearly 80% of all deaths.[6] In the present series, of 14 patients where the lungs were judged to be 'clear' on admission, 10 survived (70%), where lung signs were 'moderate' 12 survived (40%), and of 14 who had severe pulmonary involvement, none survived.

Inhalation of saliva, feeds or lipiodol may all lead to pneumonitis, consolidation or atelectasis, but it was Waterston *et al.* who showed that the presence of a *patent tracheo-oesophageal fistula from the lower oesophageal pouch is the important lethal factor.*[7] In their series of 153 infants who had such a fistula, 78 (42%) developed pneumonia, whereas of 16 infants who had atresia without fistula, only 2 (13%) had pneumonia. The fistula has 2 effects. Firstly, it allows the reflux of gastric contents upwards into the bronchial tree, and, secondly, air passes down the fistula causing gastric distension and diaphragmatic elevation.

2. *Delay Between Admission and Primary Repair*

In 25 cases, primary repair was performed within 24 hours of admission. By the simple expedient of delaying surgery 24 - 72 hours beyond the time of admission, the state of the lungs was so improved in a comparable group of 26 patients as to result in doubling of their survival rate (Table II).

TABLE II. IMPROVEMENT IN SURVIVAL WHERE PRIMARY OESOPHAGEAL REPAIR IS DELAYED 24–72 HOURS (51 CASES)

	Total	*Survivors*	*Under 5 lb.*	*Severe chest infection*
1st day of admission	25	6 (24%)	5	6
2nd and 3rd days of admission	26	14 (52%)	5	6

Frequent suction on the upper pouch, maintenance of an upright position, antibiotics and bronchoscopy were all employed in the interval period. In spite of these measures, *deterioration* in the lung state was noted in the occasional case. Furthermore, that *these efforts were insufficient to obtain complete clearing and re-expansion of the lungs in all cases* is shown by the still modest survival rate of 52% in those delayed beyond 24 hours, despite the fact that only 5 premature infants were included in the group.

3. *Birthweight*

There were 14 infants weighing under 5 lb. at birth, with one operative survivor (this infant died at 6 weeks of gastroenteritis). Five of the 11 were under 4 lb. in weight. One premature baby also had a duodenal atresia. Only 3 infants in this group had clear lungs on admission.

25 - 40% of all cases with oesophageal atresia weigh under 5 lb. at birth, and are associated with a mortality of at least 60% in most series.[2,6,8,9] It is, however, incorrect to assume that the higher mortality is due to the prematurity *per se*. Pulmonary infection and additional anomalies are more common and more lethal in these smaller babies and it has been shown that if these complications are excluded from the reckoning, only those infants below 4 lb. in weight have a worse prognosis.[2]

4. *Additional Anomalies*

Autopsies were infrequently performed in this series and only 5 patients were recorded as having serious additional congenital abnormalities. These included 1 with duodenal atresia, 2 with imperforate anus (one of these had hydrocephalus), 1 Fallot's tetralogy and 1 partial truncus communis.

Major additional anomalies may occur in up to 33⅓% of patients with oesophageal atresia, but in less than 10% of all cases do they directly lead to death.[10] Systemic decompensation may only be expected in the neonatal period with such untreatable conditions as renal agenesis, ureteric atresia, pulmonary atresia, truncus communis and major cerebral defect. The higher mortality in these infants with additional anomalies is largely due, therefore, to difficulties in management and/or pulmonary infection (Table III).

TABLE III. DETAILS OF STAGED TREATMENT IN 9 PATIENTS

Case	*Birthweight*	*Chest signs*	*Reason for staging*	*Method of Staging*	*Comment*	*Result*
1	3½ lb.	'Clear'	Wide atretic gap	Fistula division, Gastrostomy, C. Oesophagostomy } 1st day	Pneumothorax and consolidation following failure to drain chest	Died
2	3½ lb.	'Severe'	Chest signs and prematurity	Gastrostomy—1st day	Gastrostomy catheter caused pyloric obstruction	Died
3	4½ lb.	'Moderate'	Atresia with no fistula;	Thoracotomy, Gastrostomy, C. Oesophagostomy } 2nd day	? Gastrojejunostomy the appropriate procedure	Died
			duodenal atresia	Gastrojejunostomy—4th day		
4	4¾ lb.	'Moderate'	Wide atretic gap	Fistula division, Gastrostomy, C. Oesophagostomy	All 3 procedures performed on day of admission	Died
5	5 lb.	'Severe'	Chest signs and small baby	Gastrostomy—1st day; Fistula division, C. Oesophagostomy } 3rd day	Gastrostomy wound disruption on 12th day	Died
6	5⅛ lb.	'Moderate'	Atresia with no fistula	Thoracotomy, Gastrostomy } 2nd day	Cause of death unknown	Died
7	5½ lb.	'Moderate'	Hydrocephalus; recto-vaginal fistula	Gastrostomy—3rd day		Died
8	5½ lb.	'Clear'	Imperforate anus; wide atretic gap	Gastrostomy—1st day; Delayed repair—4th day	Correctly staged	Lived
9	7 lb.	'Moderate'	Wide atretic gap	Fistula division, C. Oesophagostomy } 1st day; Gastrostomy—6th day	Correctly staged	Lived

5. *Staged Procedures*

Included here are 9 patients in whom some operative procedure was performed in order to postpone oesophageal repair. Two of these survived.

It appears that mortality in the remaining 7 was largely related to early thoracotomy or to technical difficulties—disruption of a gastrostomy wound; pneumothorax and consolidation following failure to drain the chest after fistula division; deliberate passage of a gastrostomy tube through the pylorus for feeding purposes (resulting in pyloric obstruction); difficulty in management of a duodenal atresia. In one case gastrostomy (as the initial procedure) was performed on the third day of admission.

CONCLUSIONS

It is clear that individualized management should concentrate on the prevention and treatment of pulmonary infection and the avoidance of oesophageal anastomosis in the poor-risk patient. In determining the basis for such management, the following conclusions are drawn from the series:

1. *Diagnosis is too often unduly delayed.* In confirming the diagnosis, 'lipiodol pneumonitis' should be avoided.

2. *Most of the infants admitted will have lung signs and will not be fit for early primary repair* (early oesophageal anastomosis).

3. *Where repair is delayed, immediate control of the effects of the fistula is mandatory.* Non-operative measures are not always adequate in preventing gastric reflux and in achieving maximum lung expansion. Simple suction gastrostomy is recommended as fulfilling these requirements.

4. *Delayed oesophageal anastomosis should await complete clearing of the lung fields.*

5. Every institution dealing with this problem should decide, on the basis of its experience and facilities, *if there is a minimum weight below which early primary anastomosis should never be performed.* Results here suggest the imposition of a weight limit between 4½ and 5 lb.

6. *Staged surgery is not simplified management.* Temporizing procedures require the closest attention to detail and enhanced vigilance on the part of both medical and nursing staff, if improved survival is to be obtained.

RECOMMENDED OUTLINE OF MANAGEMENT

1. *Diagnosis*

If oesophageal atresia is suspected because of increased salivation, cyanotic attacks and regurgitation while feeding, the defect is easily demonstrated by passing a fairly stiff, size 10 (French) rubber catheter down the pharynx—it will be held up about 10 cm. from the lips. There is no differential diagnosis. A straight X-ray examination of chest and abdomen should then be taken with the catheter in place. It will show whether the catheter is coiled or not, exactly how far down the tip extends, the state of the lungs, the presence of air in the stomach (indicating whether there is a patent fistula or not) and may demonstrate the presence of an intestinal obstruction. Air may be injected

down the catheter and serves as a good contrast medium: gastrografin (1 - 2 ml. only) is indicated where doubt persists, or to exclude the presence of an upper-pouch fistula. If used, the dye should be aspirated immediately after taking the X-ray photo.

2. *Immediate Control of the Effects of the Fistula*

(a) *If the patient weighs over 5 lb. and is fit* (clear lungs and no critical additional anomalies)—then immediate division of the fistula and oesophageal repair remains the treatment of choice.

(b) *If the patient weighs under 5 lb. or is otherwise unfit*—(pulmonary infection and/or critical additional anomaly)—then immediate *gastrostomy under local anaesthesia is performed*. This should be a simple 'inkwell' gastrostomy and should be a quick and safe procedure. The 2 important postoperative complications are spillage into the peritoneal cavity and obstruction of the pylorus by the gastrostomy tube. Attention to detail is therefore important—the opening in the stomach should be made as far away from the pylorus as possible; the catheter (Malecot type) should be pulled out as far as possible and tied to a skin suture; the stomach wall should be sutured to the anterior parietal peritoneum.

3. *Further Management of the Staged Patient*

Following gastrostomy, the infant is returned to a warm, moist atmosphere (preferably in an incubator if under 5 lb. in weight) and placed in a head-up position.

The gastrostomy is a deflation and suction gastrostomy. The catheter is left open and aspirated hourly.

The upper oesophageal pouch and pharynx are kept clear of saliva by intermittent (at least every 10 minutes) or continuous suction using a sump suction catheter.

Tracheal toilet should be started in theatre at the time of the gastrostomy and is repeated as often as necessary, usually once or twice daily. The preferred method is one that involves minimal trauma to the epiglottis and larynx—a small curved laryngoscope is used to pull the tongue forward and a fine suction catheter passed down into the bronchi. Bronchoscopy may be necessary in some cases.

Fluid requirements are maintained by scalp-vein infusion. Loss of saliva and gastric juice amounts to 200 - 300 ml. daily and this amount should be added to the normal daily requirements for the infant's weight and age.

Antibiotics are administered intramuscularly. We start with penicillin and streptomycin, then change to the antibiotic indicated by culture and sensitivity of the tracheal secretions.

This regime cannot continue beyond 48 - 72 hours since the infant must be fed. A decision must now be taken between (*a*) delayed primary repair and (*b*) division of the fistula.

Delayed primary repair. Most infants requiring an initial gastrostomy will be over 5 lb. in weight, and if their lungs have cleared within 48 - 72 hours, they will be suitable for delayed repair, which will proceed along the same lines as immediate division of a fistula and oesophageal anastomosis. A serious associated anomaly can usually also be controlled within 48 hours (e.g. colostomy for imperforate anus) and these patients are thus rendered fit for delayed primary repair.

Division of the fistula. This will be necessary if —

(*a*) The birthweight is below 5 lb.,

(*b*) evidence of pulmonary infection persists, or if

(*c*) the infant remains enfeebled by an additional major anomaly, or

(*d*) thoracotomy reveals an atretic gap too great for direct anastomosis.

The extra-pleural approach to fistula division should be cultivated since this may be performed under local anaesthesia, with minimal disturbance to the poor-risk patient and minimal compression of the right lung.[11]

Secondary oesophageal anastomosis can now be postponed indefinitely until the patient is fit and weighs at least 6 - 7 lb.

Cervical oesophagostomy is never an emergency operation and may be postponed weeks or even months. Suction on the upper pouch should be effective and needs to be less frequent as the baby's cough reflex develops and he learns not to inhale saliva.

Cervical oesophagostomy is performed either because the atretic gap is too great for direct anastomosis or because an anastomosis has broken down and cannot be repaired.

Reconstruction with colonic interposition will then be necessary and is usually postponed until the child reaches 1 year of age.

Atresia without fistula. This is suspected when the initial X-ray shows no air in the stomach. If the infant is fit, there is a good case for early thoracotomy and exploration, as an oesophageal anastomosis will be possible in some cases. In most, however, the atretic gap will be too great and gastrostomy (a difficult procedure because of the small stomach) with cervical oesophagostomy will have to be performed. If the patient is not fit for thoracotomy, gastrostomy is performed and at the same time Haight's method of measuring the lower oesophageal segment is used.[12] A radio-opaque catheter is passed up the lower oesophagus via the cardiac orifice. Postoperative radiographs will demonstrate how closely the tip of the catheter approximates the lower end of the upper pouch, and whether oesophageal anastomosis will be feasible.

DISCUSSION

This series contains a high incidence of cases complicated by pulmonary infection and illustrates the failure of early primary repair in the patient with severe complications. This supports Waterston's observation pertaining to this group that: 'It is clear that the policy of primary anastomosis has been tried and has failed.'[2] Most centres have found that results are better if primary repair is delayed 2 or 3 days after admission and have adopted the view that operation in oesophageal atresia is not an emergency. My impression, based on results in the present series, is that this view bears some risk and that control of the effects of the fistula is a matter of urgency.

The case for staged surgery in the management of poor-risk patients with oesophageal atresia has not yet been proved, since truly comparative series are not available. However, some remarkable successes with staging have been reported.

In 1958, noting the high mortality among premature babies with oesophageal atresia, Gross adopted a policy

of initial gastrostomy and fistula division, followed by oesophageal anastomosis when a weight of 6 - 7 lb. was reached.[13] He thereby improved his survival rate in this group from 28% to 62%.

Holder *et al.* treated 11 babies weighing less than 5 lb. along similar lines and saved eight.[11] Seven of the 11 weighed under 4 lb., only one of whom was lost. The smallest survivor weighed 2 lb. 13 oz. at 2 days. In 2 infants weighing less than 4 lb. diagnosis had been delayed more than a week. The delay before definitive oesophageal anastomosis varied from 2 weeks to 6 months.

Rehbein had 24 cases of atresia of the oesophagus with associated occlusions of the duodenum or anus and saved six.[14] He concluded that: 'Single-staged management of the oesophageal lesion and of the obstruction of the duodenum or anus should only—but should—be performed in the mature child with early diagnosis of the anomaly and in the absence of further complications.'

It is evident that in centres where experience can be accumulated, the highly specialized problem of complicated oesophageal atresia is being handled with increasing success. No longer need it be said: (Clatworthy, 1955) '. . . if the infant is in such poor condition and so seriously ill with complications that he cannot be prepared as a reasonable surgical risk within . . . (12 hours) . . . there is little hope of salvage'.[15]

SUMMARY

1. The mortality in a series of 60 patients with oesophageal atresia has been correlated with the presence of complicating factors and with the management of the patients in hospital.

2. It is suggested that oesophageal atresia with tracheo-oesophageal fistula constitutes a surgical emergency in that control of the effects of the fistula is urgent.

3. The rationale, scope and technique of surgical staging in the condition is discussed.

4. It is hoped that by outlining a plan for individualized management of oesophageal atresia we will stimulate enthusiasm and interest in the care of these patients and a determination to improve on hitherto modest survivals.

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