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Tracheo-oesophageal fistula and the respiratory distress syndrome

S. J. K. Holmes, E. M. Kiely, and L. Spitz

Hospital for Sick Children, Great Ormond Street, London, UK

Abstract. Four pre-term neonates with tracheo-oesophageal fistula required positive pressure ventilation because of the respiratory distress syndrome. Gastric rupture occurred in two patients, one of whom died. Ineffective ventilation was critically impaired by formation of a gastrostomy in a third patient, who also died. Direct ligation of the fistula in the fourth critically ill patient led to a dramatic improvement in gas exchange. When the resistance of the airways exceeds that of the fistula, gas escapes through the latter into the gastro-intestinal tract. Formation of a gastrostomy lowers intragastric pressure and thus the resistance to gas escape via the fistula; respiratory support is thus rendered ineffective. Effective ventilation is only possible after occlusion of the fistula, which we believe is best achieved by direct ligation.

Key words: Oesophageal atresia – Respiratory distress syndrome – Tracheo-oesophageal fistula

Introduction

Primary surgical correction of oesophageal atresia and tracheo-oesophageal fistula early in the neonatal period has been the goal since the success of Haight in 1941. Survival rates approaching 100% are now achieved [6, 9, 10] with patients in Waterston's groups A and B [14].

One-third of these babies are born prematurely and a significant number develop the respiratory distress syndrome (RDS). The high mortality rates associated with primary correction in these patients led to the development of staged correction following early gastrostomy [4, 8].

We report our experience with four such patients (Table 1) in whom tracheo-oesophageal fistula was associated with RDS in order to emphasise the dangerous interaction between the two conditions and the need for urgent occlusion of the fistula.

Offprint requests to: E. M. Kiely

Patients

Case 1. A female infant born at 29 weeks' gestation and weighing 835 g was born by emergency caesarean section. Respiratory distress was treated by continuous positive airway pressure to the naso-pharynx. A gastrostomy and jejunal feeding tube were inserted on the 2nd day of life. On the 5th day the child became acutely unwell with a pneumoperitoneum and was transferred to our institution, where urgent operation was undertaken to ligate the tracheo-oesophageal fistula and repair the ruptured stomach. After an initial improvement, the pneumonia present on transfer was complicated by heart failure and she died in spite of support at the age of 25 days.

Case 2. A female infant born at 32 weeks' gestation weighing 1400 g required respiratory support by intermittent positive pressure ventilation (IPPV) shortly after birth. Gas exchange remained inadequate in spite of increasing levels of support. The abdomen became markedly distended and a gastrostomy was performed at the age of 8 h. Thereafter ventilation became increasingly ineffective in association with a large gaseous leak through the gastrostomy. The child's condition deteriorated progressively and she died at the age of 20 h.

Case 3. A female infant born at 30 weeks' gestation weighing 1100 g required respiratory support by IPPV shortly after birth. Adequate gas exchange was achieved and her stable condition apparently favoured delayed primary repair. At the age of 36 h her condition deteriorated dramatically in association with massive abdominal distension. Urgent operation was undertaken to ligate the tracheo-oesophageal fistula and repair the gastric perforation. The patent ductus arteriosus (PDA) was ligated at the age of 7 days. Delayed primary repair proved impossible because of a wide gap and the child was discharged with a cervical oesophagostomy and gastrostomy for future gastric transposition.

Table 1. Patients with tracheo-oesophageal fistula and respiratory distress syndrome

Patient	Birth weight (g)	Gestational age (weeks)	Associated abnormalities	Primary procedure	Complications	Subsequent procedures	Outcome
1	835	29	PDA; PFO; VSD	Gastrostomy day 2	Gastric perforation day 5	Ligation TOF repair stomach day 5	Death age 25 days
2	1 400	32	PDA; A–V canal defect	Gastrostomy day 1	Progressive respiratory failure		Death age 20 h
3	1 100	30	PDA	Nil	Gastric perforation 36 h	Ligation TOF repair stomach 36 h	Alive (Gastric interposition 9 months)
4	1 600	33	PDA	Ligation TOF 16 h		Delayed primary correction 7 days ligation PDA 21 days	Alive and well (Fundoplication 6 months)

PDA = Patent ductus arteriosus
PFO = Patent foramen ovale
VSD = Ventricular septal defect
TOF = Tracheo-oesophageal fistula

Case 4. A female infant born at 33 weeks' gestation weighing 1600 g developed respiratory failure requiring support with IPPV at the age of 6 h. Gas exchange remained inadequate in spite of maximum conventional respiratory support. At the age of 16 h, with the child in extremis, emergency ligation of the fistula via a right thoracotomy was performed. An immediate improvement in ventilation occurred and her general condition became progressively better. Delayed primary correction of the oesophageal atresia was undertaken at the age of 7 days. Ligation of the PDA, which was well tolerated, was the only incident in an otherwise uneventful recovery. Failure to thrive at the age of 6 months was attributed to gastro-oesophageal reflux and corrected by fundoplication. The child now enjoys good health.

Discussion

The application of positive pressure to the airway in a patient with a tracheo-oesophageal fistula leads inevitably to escape of gas from the trachea to the oesophagus. The low pulmonary compliance associated with RDS necessitates increasing airway pressures to achieve effective ventilation. As this pressure increases, so does the amount of gas escaping. These events lead to loss of airway pressure, ineffective ventilation, and gastric distension. The potentially lethal consequences are exemplified by the cases described above.

Concern about the hazards of a tracheo-oesophageal fistula in staged management has largely been limited to the reflux of gastric contents into the airway. The desire to avoid allegedly dangerous thoracotomy and direct ligation of the fistula in the group C patient has led to the development of a number of alternative procedures. One of the earliest was gastric transection [3, 11], followed by lesser procedures attempting to achieve the same end by ligation of the gastro-oesophageal junction or occlusion of the distal oesophagus by a balloon catheter introduced via a gastrostomy. We have no experience of these techniques, but do not consider gastric transection a less traumatic procedure than direct ligation of the fistula and are concerned about damage to the distal oesophagus by attempts to occlude it.

Anaesthetists have emphasised the importance of positioning the endotracheal tube such that the tip lies distal to the fistula [1, 12]. The proximity of the fistula to the carina renders this ideal, with which we agree, difficult to achieve in practice.

Until recently, gastrointestinal and thus abdominal distension was thought to be the main cause of ineffective ventilation. The almost universally fatal outcome from rupture of the distended stomach [7, 12] gave credence to previous reports recommending early gastrostomy and staged management of these critically ill babies [4, 8].

Effective ventilation of the lungs is only possible if the airway resistance is lower than that of the tracheo-oesophageal fistula. Filston and his colleagues [2] were the first to emphasise that the tracheo-oesophageal fistula acts as a "low-resistance vent". This allows leakage of gas from the airway and is the main barrier to effective parenchymal ventilation in non-compliant lungs. The addition of a gastrostomy to this already inefficient system lowers the resistance of the vent so that ventilating gases pass preferentially into the atmosphere, by-passing the pulmonary parenchyma. Effective ventilation then becomes impossible

as in our case 1 and those reported by Filston [2] and Templeton [13].

Occlusion of the fistula provides the only solution. Filston achieved this without thoracotomy by ingenious use of a Fogarty balloon catheter. In this way, effective ventilation was possible until the pulmonary pathology resolved and delayed primary repair was possible. While impressed with these results, we are concerned about the difficulties of bronchoscopically introducing the catheter in the presence of respiratory failure and about securing the balloon in position in the fistula.

In case 4 above, direct ligation of the fistula was performed as an emergency with an immediate improvement in the efficiency of ventilation and gas exchange. Primary anastomosis was delayed as the child had been in a critical condition prior to ligation of the fistula. The dramatic improvement which follows ligation of the fistula may even allow immediate primary anastomosis, as evidenced by the excellent results of Templeton et al., with whose report [13] we wholeheartedly agree.

In conclusion, early ligation of the tracheo-oesophageal fistula is a prerequisite to the effective management of a neonate in respiratory failure. The greater the difficulty in achieving adequate gas exchange, the more urgent is the need to occlude the fistula.

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