

ATRESIA OF THE OESOPHAGUS WITH AN ABNORMAL TRACHEAL FOLD

BY

R. H. FRANKLIN AND A. J. P. GRAHAM

From the Department of Surgery, Postgraduate Medical School of London

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Refinements in diagnosis and increasing vigilance amongst obstetricians and paediatricians are leading to a more frequent and earlier diagnosis of congenital atresia of the oesophagus. Though the condition is comparatively uncommon its existence must be borne in mind in all cases of neonatal feeding difficulties, atelectasis, and cyanosis. The commonest cause of the last two conditions is the existence of a fistula between the trachea and the lower oesophageal segment, resulting in regurgitation of gastric juice into the bronchial tree. But it is becoming apparent that associated abnormalities of the trachea may lead to pulmonary complications, cyanosis, and congenital stridor. The following case is an example.

CASE REPORT

J. L., 6 lb. 13 oz., was born at 5 p.m. on August 18, with a normal delivery. Shortly afterwards it was noted that she had stridor and at intervals appeared cyanotic. Pharyngeal suction did not result in any improvement, and laryngoscopy showed no abnormality to account for the stridor or cyanosis. Sterile water by mouth was regurgitated. A soft rubber catheter was

arrested 10 cm. from the alveolar margin, and this finding led to the tentative diagnosis of congenital atresia of the oesophagus. The infant was examined under the x-ray screen, and air was seen in the stomach and intestines. The introduction of 1 ml. of iodized oil into the catheter demonstrated the typical blind upper oesophageal pouch. It was thus apparent that the infant had the common form of congenital oesophageal atresia in which there is a blind upper pouch ending about the level of the vena azygos arch, and a lower oesophageal segment communicating with the trachea by means of a fistula at its upper end.

The cause of the stridor and cyanosis was not obvious. Cardiac sounds and radiological appearance were normal.

The child was referred to Hammersmith Hospital by Dr. M. Baber for surgical treatment 24 hours after birth. Her general condition was good apart from the stridor and the cyanosis, which was relieved but not removed in an oxygen tent. Even in this, periodic cyanotic attacks and respiratory difficulty recurred. At one time shortly before operation tracheotomy was contemplated.

Operation (R. H. F.) was begun 28 hours after birth under endotracheal anaesthesia. The chest was opened through the right third intercostal space. It was then noted that with each inspiration the mediastinum was drawn sharply across to the opposite costal wall as though the air entry were obstructed and the operation could not proceed until a relaxant was given and controlled respiration secured. Thereafter the oesophagus was displayed and mobilized sufficiently to permit anastomosis of the upper and lower segments after section and closure of the tracheal fistula. At the close of the operation the infant was placed in a portable oxygen tent and returned to the ward. For a short period respirations appeared quietly normal, but after 45 minutes cyanosis and stridor suddenly recurred in the oxygen



FIG. 1

tent, cyanosis deepened, respiration ceased, and, despite resuscitative measures, the infant died.

NECROPSY FINDINGS.—The anastomosis was intact and appeared to be satisfactory. Both lungs were collapsed. The larynx and pharynx were normal. On opening the trachea a valve-like fold of mucous membrane was found overlying the tracheal opening of the fistula. The trachea and oesophagus were removed and examined more closely. The specimen is illustrated (Fig. 1), and consists of the larynx, trachea, both main bronchi, and their divisions anteriorly. Behind, the anastomosed oesophagus has been split longitudinally down the posterior wall and shows the dilated, thick-walled upper pouch and the narrow, thin-walled lower segment. The tracheal bifurcation is 4 cm. below the cords. The site of the fistula is in the mid-line posteriorly 0.5 cm. above the carina. Overlying this opening and extending 0.5 cm. above it is a semi-lunar, valve-like fold of tracheal mucous membrane, arising from the posterior third of the trachea at this point. The free edge is about 0.6 cm. long and freely mobile. From the lateral attachments of the cusp slightly raised ridges run upwards for 1 cm. in line with the posterior tips of the tracheal cartilages. In the mid-line posteriorly a raphe-like ridge extends from the posterior commissure of the cords to the site of the fistula. Along this line on the postero-external surface the trachea and upper oesophageal pouch are attached by a thin cicatricial band. Below the fistula the oesophagus and trachea were unattached and no median raphe can be seen. There were no abnormalities elsewhere.

DISCUSSION

The type of oesophageal atresia and fistula was that which accounts for about 80% of these cases. The interest in this case lies in the abnormal valve-like fold in the trachea, which has not hitherto been described in the literature. The fold was similar in shape to a thickened aortic valve cusp, and, since it was quite mobile, was presumably capable of

movement with the passage of air on respiration. On inspiration the cusp would be drawn out into the lumen of the trachea, obstructing air-entry and causing stridor and cyanosis. One may presume that with quiet respiration occlusion was only partial, but with stronger inspiratory efforts more complete obstruction would occur. By drawing out the centre of the free edge of fold it was possible to occlude approximately two-thirds of the tracheal lumen. Thus the cause of the stridor, the sudden attacks of cyanosis, and the violent inspiratory movements of the mediastinum at operation can be explained by obstruction of the tracheal lumen on inspiration.

Congenital stridor is an exceedingly rare condition, though according to Wilson (1952) it is possible that congenital laryngeal stridor may account for some deaths reported as "suffocation" or "over laying." Other congenital causes are webs or stenosis of the larynx or pressure on the trachea by mediastinal tumours or an enlarged thymus, anomalies of the great vessels or heart (White Franklin, 1952). Mercer (1945) has recorded acute stridor following successful surgical treatment for oesophageal atresia. One of us (R.H.F.) has encountered a congenital narrowing of the trachea at the site of an oesophageal fistula.

It is the tragedy of this case that an otherwise excellent prognosis was ruined by a congenital deformity which might have been corrected. Investigation of the stridor by pre-operative bronchoscopy might have revealed the additional abnormality.

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