Tracheal compression as a cause of respiratory symptoms after repair of oesophageal atresia

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SUMMARY A case of tracheal compression in an infant after repair of a tracheo-oesophageal fistula and oesophageal atresia is reported. Tracheopexy completely relieved the symptoms of apnoeic attacks, cyanosis, and convulsions. We suggest that tracheal compression in infants and children with repaired oesophageal atresia can not only cause life-threatening attacks but also be responsible for recurrent chest infections. All infants and children with signs and symptoms of tracheal compression as shown by the presence of a barking type of cough, recurrent chest infections, or persistent mild respiratory symptoms should be referred for thorough investigation of the tracheobronchial tree.

Respiratory problems after successful repair of oesophageal atresia may arise in the immediate postoperative period or present as recurrent respiratory infections in infancy and childhood. These later repeated episodes are common and may occasionally be so severe as to be life threatening. The treatment offered is often inappropriate since the precise causes of these recurrent infections have not been elucidated although tracheal abnormalities in children with oesophageal atresia have been described (Holinger and Johnston, 1963; Holinger et al., 1965; Benjamin et al., 1976) and their significance in terms of respiratory symptoms has been pointed out (Benjamin et al., 1976; Filler et al., 1976). Metaplasia of the tracheal mucosa and replacement of the ciliated squamous epithelium may also be of importance (Emery and Haddadin, 1971; Mithal and Emery, 1976).

The purpose of this report is to draw further attention to the occurrence of tracheal compression with consequent attacks of expiratory wheeze, cyanosis, apnoic spells, and recurrent respiratory infections, and to make a plea for a full investigation of the tracheobronchial tree (including bronchoscopy, bronchography, and if necessary angiography) in all children presenting with these symptoms of tracheal compression.

Case report

This first infant of healthy parents was born after a normal pregnancy and delivery, weight 3.147 kg. No resuscitation was needed at birth. Excessive oral mucus and the failure of passage of a tube into the stomach led to a diagnosis of oesophageal atresia and after transfer to this hospital she was operated on at the age of 8½ hours.

Bronchoscopy after the induction of anaesthesia showed a normal larynx without subglottic stenosis and a normal upper trachea. There was a marked flattening of the anterior and left lateral part of the trachea occluding over half of the lumen. The fistula to the oesophagus opened just below this bulge, 8 mm above the carina. The tracheo-oesophageal fistula was ligated through a right thoracotomy by an extrapleural approach and an oesophageal anastomosis made. Postoperative recovery was uneventful although she was noticed to have a harsh brassy cough or 'bark' when she cried. She was discharged home aged 16 days and made normal progress despite the barking cough.

When aged 4 months, she began to have 'colds' and to be snuffy, and a month later while being fed she had a bout of coughing, stopped breathing, and went blue. Recovery was rapid but three further episodes occurred in the ensuing weeks with cyanosis, apnoea, and generalised convulsions. Her paediatrician referred her back for investigation. Endoscopy showed a satisfactorily wide oesophageal anastomosis while in the trachea the findings were similar to those at birth except that the left lateral and anterior flattening was more marked and was seen to be markedly pulsatile (Fig. 1). Xerograms confirmed this flattening of the trachea at and just above the aortic arch. Angiograms and a ciné tracheogram showed that the aortic arch and great...
vessels were of normal configuration but that the tracheal lumen was virtually occluded with each pulse wave.

At age 7 months, via a left anterolateral thoracotomy the adventitia of the aortic and great vessels was sutured to the manubrium sterni in order to pull forward the anterior tracheal wall (Gross and Neuhausser, 1948; Mustard et al., 1969). Endoscopy immediately afterwards showed an almost normal tracheal shape (Fig. 2). Postoperative recovery was excellent and her mother volunteered the opinion that 'she hadn’t breathed so easily and freely in all her life'. The 'bark', although still present, was much less raucous and much less frequent. Tolerance of the operation and the rapid relief of symptoms and signs were gratifying and more than justified the decision to perform a tracheoectomy.

Discussion

Tracheoscopy led to a rapid and precise diagnosis and effective treatment in our patient with tracheal compression. Failure to perform endoscopy frequently means that the condition is misdiagnosed as tracheomalacia, recurrent respiratory infection, asthma, or oesophageal malfunction. 2 patients with similar life-threatening episodes of cyanosis and apnoea have had tracheostomies performed. This procedure prevented these episodes, but in the light of our recent experience the correct operative procedure was tracheoectomy. It is possible that the 'cot-death' of one of our cases after successful repair of oesophageal atresia was caused by this condition.

The incidence of tracheal compression in children who have had surgery for oesophageal atresia has not been accurately determined though a prospective study is now being undertaken. Filler et al (1976) found that symptomatic compression of the trachea occurred in 3 children and anatomic compression in 13 out of a total of 40 consecutive cases of oesophageal atresia. Touloukian et al. (1974) found that 2 out of 18 cases of oesophageal atresia had repeated episodes of acute respiratory distress due to tracheomalacia for periods up to one year of age.

Our initial analysis of infants and children with repaired oesophageal atresia suggests that the abnormality of the trachea is closely correlated with the clinical state of the patient and its recognition is essential to effective management. Thus, a persistent 'bark' is only found in those children with tracheal narrowing and in these patients recurrent pulmonary infections are common. It is our clinical impression that these respiratory symptoms improve after the age of 4 or 5 years but this can only be verified by regular follow-up of the cases in our study.

The management of these children must depend on their clinical condition. The majority of children with mild tracheal compression have minimal signs and suffer no consequences. However, any child with symptoms or signs of moderate or marked tracheal compression should be investigated. A history of recurrent pulmonary infections demands that bronchoscopy and bronchogram be performed to ascertain the current state of the diseased respiratory tract. Early tracheoectomy in patients with moderate tracheal narrowing may well prevent
recurrent chest infection in these children and avoid permanent tracheobronchial damage. Certainly, early operation is indicated in all patients whose lives are endangered by severe tracheal compression associated with apnoeic spells, cyanosis, or convulsions.

References


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