A CASE OF ATRESIA OF THE LARYNX

BY

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Congenital abnormalities of the larynx and trachea rarely cause death in the newborn. Milder anomalies are not uncommon and are compatible with survival. Holinger, Johnson and Schiller (1954) reported a series of 379 cases, of which 305 were congenital laryngeal stridor. Among 19 cases of congenital web of the larynx three required tracheotomy and none died. This condition may persist without symptoms into adult life. One case of membranous atresia of the glottis survived after immediate tracheotomy. Congenital subglottic stenosis was found in 34 cases, in two of which it was due to deformity of the cricoid cartilage. Fourteen of these required tracheotomy. One case of cartilaginous stenosis died at the age of 4 weeks in spite of tracheotomy. There were five cases of congenital cyst and 15 of laryngocoele. Congenital papillomata and tumours such as haemangioma and lymphangiomata have been reported. Kessel and Smith (1953) described a case of congenital absence of the trachea and reviewed the literature on tracheal anomalies.

Subglottic cartilaginous atresia or stenosis is the least common anomaly. O'Kane (1936) described a case of stenosis which survived after tracheotomy and Baker (1954) reviewed the same case at the age of 18 years. Gunn (1926) referred to one case of partial and one of complete stenosis. Potter and Bohlender (1941) reported a case of atresia which died 20 minutes after birth and Potter (1952) referred to two other cases. Bizza (1941) described complete atresia in a stillborn infant with 'hyperplasia' of the lungs and referred to eight other cases in the German literature. Sayre and Hall (1954) reported one case of stenosis which survived for two days and one of atresia. They referred to 11 cases in the German literature. Sandison's (1955) case was associated with partial absence of the trachea and tracheo-oesophageal fistula.

Of the above cases complete cartilaginous atresia was present in six and membranous atresia in one. Bizza (1941) and Sayre and Hall (1954) mention a further 11 cases in the German literature. The purpose of this paper is to describe a case of cartilaginous subglottic atresia and to discuss the embryology, diagnosis and possible treatment.

Case Report

The infant was the first of girl twins delivered of a healthy mother two weeks before the expected date following an uneventful pregnancy. The previous obstetrical history consisted of two full-term normal babies and one miscarriage at three and a half months. The mother is Rh positive and has a negative Wassermann reaction.

At birth the child was very asphyxiated, cyanosed and did not cry. Oxygen was administered by face mask and an occasional gasp was observed, in spite of which the condition rapidly became that of asphyxia pallida. The apex beat was firm and regular. Lobeline, 1 ml., was given without effect. Intragastric oxygen was then administered, causing a definite, though temporary, improvement. This was continued for one hour during which the apex beat became gradually weaker, and at the end of this period the child died.

The second twin was delivered 15 minutes after the first, cried well, and did not require any resuscitation. It was subsequently discovered to have a cardiac systolic murmur, although progress to date has been normal.

Necropsy. The body was that of a premature female infant weighing 4 lb. 6 oz. (2.05 kg.). The skin was pink. The only abnormalities were in the respiratory system. The mouth, pharynx and oesophagus were normal. The supraglottic part of the larynx was normal. Immediately below the vocal cords the larynx was completely occluded. This was found to be due to a mass of cartilage 1 cm. thick posteriorly and 0·3 cm. thick anteriorly. The upper surface was convex and the lower concave, forming a dome-shaped covering over the upper end of the trachea (Fig. 1). No communication could be found between the larynx and trachea. The trachea and bronchi contained a large amount of mucus; the lungs were of normal size for a newborn infant. The left lung consisted of one lobe, the right lung of two lobes. The consistency was solid and the cut surface was slimy due to the large amount of mucus in the air passages.

Histological examination of the two lungs showed normal development (Fig. 2): most of the alveolar ducts and alveoli were expanded but there were small areas...
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Fig. 1.—The larynx opened from behind showing the band of cartilage obstructing the lumen.

Discussion

Embryology. The development of the larynx is described in detail by Frazer (1910) and more briefly in the Manual of Embryology (Baxter, 1953). Only a short summary is needed here. The larynx develops in two parts, the infraglottic as a median sagittal cleft in the floor of the pharynx at the upper end of the pulmonary outgrowth, the supraglottic (transverse part) as a modification of the floor of the pharynx above the sagittal cleft. The vocal cords lie approximately at the boundary between the two parts.

It is the infraglottic part which concerns us now. During the third or fourth week the sagittal cleft is almost completely closed by overgrowth of the lateral masses (fifth branchial arches) at the sides. The epithelium fuses, leaving a small channel at the back (ductus pharyngo-trachealis) and sometimes a similar channel anteriorly. With the development of the cricoid cartilages in the lateral masses, the cleft reopens at the eighth-ninth week to form the infraglottic part of the larynx.

Fig. 2.—Section of lung showing normal development of the alveoli. Haematoxylin and eosin × 225.

Fig. 3.—Section of lung showing mucus in the air passages and alveoli. P.A.S. × 125.
ARCHIVES OF DISEASE IN CHILDHOOD

It is generally agreed that subglottic atresia is due to failure of the sagittal cleft to reopen, usually combined with an overgrowth of cartilage in the lateral masses. The small posterior fistula (ductus pharyngo-trachealis) has been found in many of the reported cases. It was not found in the present case, possibly because the larynx was opened from behind. The development of the lungs was normal in this case and in the others when the lungs are mentioned.

In Bizza's (1941) case the lungs were enlarged and he quotes other cases where this was found. He considered this to be due to retention of mucus. Although the lungs were of normal size in the present case there was considerable retention of mucus (Fig. 3). Snyder and Rosenfeld (1937) produced evidence that a tidal flow of amniotic fluid into the foetal lungs was necessary for their normal development. Potter and Bohlender (1941) denied this, giving a case of laryngeal atresia as an example. They considered that the fluid filling the alveoli and air passages could be transudate. They did not mention the presence of mucus, but noticed 'precipitated protein' in the alveoli histologically. Although it can play no part in the normal development of the lungs, it may be that retained secretions lead to dilatation of the alveoli and so assist the development of the lungs in cases where there is no outlet for the secretion.

Diagnosis. In the absence of any complications at birth which might lead to shock or narcosis, the outstanding symptoms are failure to breathe and cry. The infant is active and makes respiratory movements. Sternal retraction and indrawing of the intercostal spaces, supraclavicular fossae and epigastrium may be seen. The colour is usually normal at birth, but cyanosis soon appears after the cord is tied.

The presence of active movements and attempts at respiration distinguish atresia of the larynx from apnoea due to shock or narcosis, the normal colour at birth from asphyxia and cardiac cyanosis, and the absence of stridor from other laryngeal and tracheal anomalies.

The manoeuvre of clearing the respiratory passages will not help in diagnosis as the catheter seldom enters the larynx. The only certain method is laryngoscopy, which is not easy in a newborn infant. Also facilities for laryngoscopy are not always available at short notice.

Treatment. Without treatment the infant will not survive for many minutes. Potter and Bohlender's (1941) case survived for 20 minutes. The cases of Kessel and Smith (1953) and Sandison (1955) with complete and partial absence of the trachea survived for 66 minutes, and 4 hours 42 minutes. Both had a communication between the oesophagus and air passages and some aeration of the lung was found at necropsy.

Intragastric oxygen can maintain quite adequate oxygenation of the blood in the newborn. Sandison's case was given this treatment for 30 minutes and it produced an improvement in colour. A similar effect was obtained in the present case and survival was prolonged to 60 minutes.

Tracheotomy should be performed as soon as possible after the condition has been diagnosed. The case of membranous atresia reported by Holinger et al. (1954) was treated in this way and was alive and well at 10 months. Several other cases in their series were treated by tracheotomy soon after birth with good results. There still remains the danger of respiratory obstruction by the excess of retained mucus in the air passages. There might also be an increased susceptibility to pneumonia.

It has recently been suggested that hyaline membrane represents a highly concentrated secretion from the epithelial cells of the terminal bronchioles and alveolar ducts (Lynch and Mellor, 1955) or endogenous mucus (Wagner, 1954). If these conclusions are correct, an infant with laryngeal atresia and retention of these secretions should be very liable to develop hyaline membrane.

If the infant should survive, it is doubtful whether a satisfactory reconstruction of the larynx would be possible. Even if an airway could be established, permanent aphony would remain.

Summary

A case of cartilaginous subglottic atresia of the larynx in a twin newborn infant is described. The infant was given intragastric oxygen and survived for 60 minutes. The other twin had no respiratory difficulty but has a cardiac murmur. The embryology, diagnosis and treatment of laryngeal atresia are discussed.

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Addendum

Reconstruction has recently been successfully performed on a child of 2½ years with laryngeal stenosis (Sharp, 1956).
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