LARYNGEAL ATRESIA

BY

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(RECEIVED FOR PUBLICATION FEBRUARY 24, 1964)

Laryngeal atresia is amongst the rarest of congenital deformities. Thus, in the years 1948-1963 with approximately 50,000 births at St. Mary's Hospital, Manchester, there was only one with this deformity. While there is an abundance of reports of such conditions as laryngeal web, subglottic stenosis, and congenital stridor, only 4 case reports have appeared in the English literature of laryngeal atresia occurring in the absence of other congenital abnormalities. The infrequency of the condition is reflected by Holinger, Johnson, and Schiller (1954) in a review of a series of 379 cases of congenital laryngeal anomalies seen over a period of 10 years in Chicago. This series included 305 cases of congenital laryngeal stridor, 19 cases of congenital web, and 34 cases of congenital subglottic stenosis, but only 1 case of laryngeal atresia.

A review of the world literature shows that only 16 cases of laryngeal atresia have been recorded, and of these only 10 were infants without associated complex anomalies (Table). These reported cases probably represent a fairly high percentage of cases actually seen, but certainly not all, for MacGregor (1960) notes having seen several additional cases at necropsy and Potter (1961) described a further two cases seen in her practice. Nevertheless, the rarity of the anomaly is such that cases often fail to be recognized on the extremely infrequent occasions on which they occur. Most of the reported cases have been recognized only at necropsy and this is doubly unfortunate, for not only is the clinical picture resulting from the deformity highly characteristic, but if correctly diagnosed, prompt action may avert the otherwise inevitably fatal outcome.

It seems worth while, therefore, to report two further cases of laryngeal atresia; to review the previously reported cases; to discuss the embryology; and to describe the clinical picture and possible treatment.

Case Reports

Case 1. This male child was born to a healthy 24-year-old mother whose previous obstetric history consisted of one normal pregnancy that had resulted in a healthy living child. The mother's Wassermann reaction was

<table>
<thead>
<tr>
<th>Author</th>
<th>Live Born</th>
<th>Level of Atresia</th>
<th>Nature</th>
<th>Associated Abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rossi (1826)</td>
<td>+</td>
<td>Subglottic</td>
<td>Membranous</td>
<td>Atresia of posterior nares; tracheo-oesophageal fistula; scoliosis of trunk</td>
</tr>
<tr>
<td>Rose (1866)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of foot and hand; hypoplasia of right arm</td>
</tr>
<tr>
<td>Chiarri (1883)</td>
<td></td>
<td>Subglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of ear and nose</td>
</tr>
<tr>
<td>Gigli (1901)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of ear and nose</td>
</tr>
<tr>
<td>Frankenberger (1905)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of ear and nose</td>
</tr>
<tr>
<td>Kros (1915)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of ear and nose</td>
</tr>
<tr>
<td>Sieg (1922)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of ear and nose</td>
</tr>
<tr>
<td>Kovacs (1933)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of ear and nose</td>
</tr>
<tr>
<td>Fritz (1933)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Abnormality of ear and nose</td>
</tr>
<tr>
<td>Potter and Bohlender (1941)</td>
<td>+</td>
<td>Subglottic</td>
<td>Cartilaginous</td>
<td>Partial atresia of trachea; tracheo-oesophageal fistula</td>
</tr>
<tr>
<td>Bizz (1941)</td>
<td></td>
<td>Sub- and supraglottic</td>
<td>Cartilaginous</td>
<td>Partial atresia of trachea; tracheo-oesophageal fistula</td>
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<tr>
<td>Heineken (1952)</td>
<td></td>
<td>Subglottic</td>
<td>Cartilaginous</td>
<td>Partial atresia of trachea; tracheo-oesophageal fistula</td>
</tr>
<tr>
<td>Holinger et al. (1954)</td>
<td></td>
<td>Subglottic</td>
<td>Cartilaginous</td>
<td>Partial atresia of trachea; tracheo-oesophageal fistula</td>
</tr>
<tr>
<td>Sayre and Hall (1954)</td>
<td></td>
<td>Subglottic</td>
<td>Cartilaginous</td>
<td>Partial atresia of trachea; tracheo-oesophageal fistula</td>
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<tr>
<td>Sandison (1955)</td>
<td></td>
<td>Subglottic</td>
<td>Cartilaginous</td>
<td>Partial atresia of trachea; tracheo-oesophageal fistula</td>
</tr>
<tr>
<td>Rankin and Mendelson (1956)</td>
<td>+</td>
<td>Subglottic</td>
<td>Cartilaginous</td>
<td>Partial atresia of trachea; tracheo-oesophageal fistula</td>
</tr>
</tbody>
</table>
negative and she was Rhesus positive. There was no history of drug-taking during the pregnancy, which ran an uneventful course throughout. The child was delivered at 40 weeks, following a normal labour during which there was no evidence of foetal distress. At birth he appeared healthy, and his colour was normal. The original good colour was rapidly lost, however, and a gradually deepening cyanosis supervened. He was active and was making strong inspiratory efforts without any evidence of air entering into the lungs. The heart was beating regularly and strongly. Despite suction and oxygen his condition rapidly deteriorated with deepening cyanosis and weakening of the heart beat. An attempt at intubation was unsuccessful and death occurred 20 minutes after birth.

Necropsy. The child was of mature appearance and showed no external abnormality. The body weighed 2·6 kg. and the crown-heel length was 43 cm., the crown-rump length 34 cm. The stump of the umbilical cord contained two umbilical arteries. The skull and cranial contents were normal. The oral cavity, pharynx, thyroid, and oesophagus were normal. The larynx was occluded in the subglottic region and a probe could not be passed into the trachea. The trachea itself was, however, normal and patent throughout but contained a considerable quantity of whitish mucus. The right lung weighed 36 g. and left lung 26 g. Both lungs were voluminous and on sectioning appeared reddish brown in colour. There were no pleural petechial haemorrhages. The thymus was normal and there were no subcapsular haemorrhages. The heart was of normal size and shape, but there was an atrial septal defect due to failure of development of the septum secundum. The abdominal viscera and urogenital tract were normal. A centre of ossification was present in the os calcis but not in the lower end of the femur.

Larynx. Viewed from above the laryngeal lumen was occluded by firm whitish grey tissue at a point just below the vocal cord. The epiglottis and supraglottic regions were normal. There was a small dimple at the dorsal end of the medial line of the upper surface of the occluding mass. The larynx was opened posteriorly and the occluding mass cut sagittally (Fig. 1) when it could be seen that the occlusion was a solid plug of tissue extending for 1·25 cm. from just below the vocal cord to the beginning of the trachea. The central part of this plug consisted of cartilage and above and below this was a mixture of fibrous and muscular tissue.

Histology. The lungs were unaerated. In the peripheral portions of the lung the alveoli were collapsed and empty but in the more central areas the alveoli were widely distended and filled with mucus (Fig. 2). The bronchioles and small bronchi also contained considerable quantities of mucus. There was no evidence of infection and the histology of the remaining organs was normal.

Case 2. The mother of this male child was 22, and had had two previous normal pregnancies each of which had resulted in a normal child. The present pregnancy was
uneventful throughout and there had been no evidence of hydramnios. The mother’s Wassermann was negative and she was Rhesus positive. Labour was induced after 42 weeks' gestation and was normal throughout with no signs of foetal distress. The child appeared well at birth with a strong heart beat and a good colour, but became rapidly cyanosed after clamping of the cord. Although he was active and making respiratory efforts there was no air entry and spontaneous attempts at respiration soon ceased. Intubation was attempted but failed and cardiac massage proved of no avail. Following deepening cyanosis and weakening of the heart beat the child died 30 minutes after birth.

Histology. The lungs were normally developed and there was a moderate degree of aeration. The remaining organs showed no histological abnormality.

Discussion

The first report of a case of laryngeal atresia was by Rossi (1826). Since that date 15 cases have been reported. These cases are detailed in the Table, and it can be seen that they fall broadly into three groups, A, B, and C.

Group A consisted of two cases in which laryngeal atresia was only one of a constellation of unrelated congenital abnormalities (Rose, 1866; Chiari, 1883). Both these were stillborn and both had a cartilaginous atresia of the larynx; in one case subglottic and in the other both sub- and supraglottic regions of the larynx were occluded. In addition to laryngeal atresia, there was in Rose’s case a tracheo-oesophageal fistula, anophthalmia, an absent right ureter, and a complex malformation of the root of the tongue. The case reported by Chiari also showed, in addition to laryngeal atresia, atresia of the posterior nares, a hare-lip and cleft palate, an absent right kidney, an absent vagina, atresia of the cervix uteri, and a complete fusion of the eyelids. Cases of laryngeal atresia occurring against this background of multiple congenital abnormalities are likely to be stillborn, but the possibility of congenital abnormality of the respiratory tract would be readily considered if such an infant were born alive and showed respiratory difficulty. It is interesting to note the curious association in both cases with ocular and genito-urinary abnormalities, and tempting, therefore, to consider the possibility of this being a linked defect due to a chromosomal aberration.

Group B consisted of four cases in which laryngeal atresia was associated with deformities of the oesophagus or trachea, such as partial atresia of the trachea and tracheo-oesophageal fistula (Fritz, 1933; Sandison, 1955), oesophageal atresia and tracheo-oesophageal fistula (Sayre and Hall, 1954) or tracheo-oesophageal fistula (Heineken, 1952), but not with congenital abnormalities elsewhere in the body. Here the laryngeal atresia is only part of a more complex deformity. All the infants in this

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Fig. 3.—Case 2. The larynx has been opened from the dorsal aspect and the occluding plate of cartilage split sagittally.

Necropsy. The child was of mature appearance and showed no external abnormality. The body weighed 3.6 kg. The cranial contents were normal as was the cardiovascular system. It was impossible to pass a probe through the larynx into the trachea. The trachea itself was, however, patent and normal throughout and contained abundant white mucus. The lungs were heavy and voluminous (left 51 g., right 59 g.) but otherwise anatomically normal and showed no abnormality on section. The gastro-intestinal and genito-urinary systems were normal throughout. Centres of ossification were present in the lower end of the femur and in the os calcis.

**Larynx** (Fig. 3). The larynx was occluded by a thin plate of cartilage at a point 5 mm. below the vocal cords. The cartilaginous plate was 2 mm. thick. A passage, less than 1 mm. in diameter, was present at the posterior end of the medial line of the obstruction. This extended through from the larynx into the trachea and was thought to represent the ductus pharyngeo-trachealis. The duct was closed by a plug of thick white mucoid material in the upper part of the trachea.
group were born alive and in all cases the atresia was cartilaginous in type and in the subglottic area. In this group there is usually a communication between the oesophagus and trachea and hence there exists an opportunity for limited air entry into the lungs. Such infants do not, therefore, necessarily show the complete respiratory obstruction characteristic of an isolated laryngeal atresia. The mothers of infants falling into this group usually have had hydramnios during pregnancy, as distinct from mothers of cases of isolated laryngeal atresia in which hydramnios is most unusual.

Group C consisted of 10 cases in which laryngeal atresia was either the only congenital abnormality present (Gigli, 1902; Frankenberger, 1905; Krosz, 1915; Kovacs, 1933; Potter and Bohlender, 1941; Bizza, 1941; Holinger et al., 1954; Rankin and Mendelson, 1956) or was accompanied only by a simple cardiac or intestinal anomaly (Rossi, 1826; Sieg, 1922). In 7 of these the obstruction was subglottic and in 3 both supra- and subglottic areas of the larynx were atretic. In 7 the obstruction was cartilaginous and in only 2 was it membranous. In one (Fritz, 1933) the obstruction was partially cartilaginous and partially membranous. Only 3 of these infants were stillborn, the remaining 7 having been born alive and surviving for 10-60 minutes after birth. In this group the trachea and oesophagus were normal and in no case did the lungs show any structural abnormality. Bizza did suggest that in his case the lungs were hypertrophic, and though it is true that the lungs have been heavy, both in the previously reported cases and in our two cases, there seems little doubt that this excessive weight can be attributed to the retained mucus.

An understanding of the nature of this lesion requires some knowledge of the embryology of the larynx, a subject described in considerable detail by Frazer (1910). The respiratory system develops from a ventral sagittal cleft in the floor of the foregut. This cleft is eventually demarcated by a central mass at its cranial end and by fused lateral masses. Growth of these masses results in a Y-shaped entrance to the laryngeal anlage and with further growth this becomes T-shaped. The lumen of the original cleft becomes obliterated by overgrowth and fusion of the epithelial surfaces of the lateral masses. In many embryos the fusion is not complete and a small channel remains patent dorsally (the duc tus pharyngeal-trachealis). Less commonly, a similar channel remains open ventrally. The usual site of fusion of the epithelial surfaces of the lateral masses is just below the vocal cords, and this area of epithelial fusion eventually breaks down to form the laryngeal cavity, while the cricoid and arytenoid cartilages develop from paired centres in the lateral masses. There seems little doubt that subglottic cartilaginous atresia is due to failure of recanalization of the fused lateral masses, with resulting development of the cricoid cartilage as a solid mass rather than as a peripheral ring. The arytenoid cartilages may also present as a fused central mass. Failure of recanalization may be associated, however, with relatively normal development of the cartilage in the lateral masses, and this can result in membranous laryngeal atresia.

The clinical picture in infants born alive with isolated laryngeal atresia is highly characteristic. The course of pregnancy is usually normal and, in particular, hydramnios does not occur—this being in sharp contrast to the frequent presence of hydramnios during pregnancies resulting in babies with laryngeal atresia in association with tracheo-oesophageal abnormalities. There is, in the absence of any associated abnormality of labour, no foetal distress, and the infant appears normal at the time of birth. The colour is normal but the child rapidly becomes cyanosed after the cord is clamped. Despite this rapidly developing cyanosis the child does not appear shocked or asphyxiated, is active, and makes strong respiratory movements. There may be in-drawing of the intercostal spaces and suprasternal notch, but despite these respiratory efforts there is no air entry into the lungs and the infant does not cry. Without treatment the respiratory efforts gradually become weaker and the infant will usually die within 20 minutes, although it is rather remarkable that completely untreated cases may live for over half an hour. This unique clinical picture is easily distinguishable from that produced by shock, intracranial birth injury, asphyxia or narcosis and should serve as an urgent indication for laryngoscopy. If laryngoscopy proves difficult, and if laryngeal atresia is diagnosed clinically, a tracheostomy should be performed as soon as possible, even without visualizing the larynx, for such a failure should not be allowed to avert the decision. It is recognized that the nature of the obstruction is such that in most cases of laryngeal atresia perfect reconstruction of the larynx and adequate speech are unlikely to be obtained, and that one is going to achieve the saving of life but not the saving of speech. It is always possible, however, that the occasional case of simple membranous atresia may be susceptible to plastic treatment and produce eventually a reasonable degree of speech. During the neonatal period, it is usually impossible to say if the obstruction is cartilaginous or membranous, so intervention to save life is both justified and indicated.
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We should like to thank Dr. F. A. Langley for his advice and criticism; Professor W. I. C. Morris in whose department this work was done; the Department of Medical Illustration, Manchester Royal Infirmary for Fig. 3 and Mr. B. W. Figg for Figs. 1 and 2.

REFERENCES


INTERNATIONAL CONGRESS OF PAEDIATRICS

The 11th International Congress of Paediatrics will be held in Tokyo, Japan, from November 7 to 13, 1965, under the sponsorship of the International Pediatric Association, the Japan Pediatric Society, the Japanese Society of Child Health, and the Japanese Government.

The scientific programme will consist of plenary sessions, group sessions, group panel discussions, films, and scientific exhibits.

Registration for the membership-enrollment should be made by July 31, 1965. Registration fees are $50 for a regular membership ($60 after March 31, 1965) $30 for an associate (family) membership ($40 after March 31, 1965).

Those who wish to present papers at the Group Sessions are kindly requested to submit abstracts of 250 words or less before January 31, 1965, to the Congress Office, in triplicate.

The preliminary programme with application forms is obtained at any member Society of the International Paediatric Association in this country or at the Congress Office in Tokyo.

For further information, please contact: The 11th International Congress of Paediatrics, c/o Department of Paediatrics, University of Tokyo, 1 Motofujicho, Bunkyo-ku, Tokyo, Japan.