VASCULAR RING: A CAUSE OF CONGENITAL STRIDOR

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Stridor beginning at, or soon after, birth is not an uncommon symptom. Such stridor is seldom severe, has a natural tendency to become progressively less marked, and in the majority of infants disappears entirely at the end of the first, or early in the second, year of life. Occasionally, some cause of laryngeal or tracheal obstruction is found and can, where possible and when necessary, be eliminated. Finally, a number of babies with congenital stridor succumb to asphyxia or recurrent pulmonary infections, usually before the age of 1 year.

It must be admitted that in the majority of cases where recovery is spontaneous, and of those who die, no satisfactory cause for the stridor is ever found. The possible causes fall into two main groups according to whether the larynx or the trachea is the site of obstruction.

Laryngeal stridor in the newborn may be due to an exaggeration of the infantile shape of the upper aperture of the larynx. The aryepiglottic folds are almost in contact, producing a purring, inspiratory stridor. There is no dyspnoea, no distress and no dysphagia, and the condition is recognized on laryngoscopy. Webs, cysts and tumours of the larynx are all excessively uncommon, and can be seen on laryngoscopic examination. Neuromuscular incoordination of the larynx has been suggested as a possible cause of congenital stridor: the condition, if it exists, should be recognizable on laryngoscopy.

Tracheal stridor in the newborn may be due to tracheal stenosis, but this condition is extremely rare, and the majority of cases are not sufficiently severe to cause symptoms. Bronchoscopy and contrast radiography will demonstrate the condition. The trachea may be pressed upon by an enlarged thymus, a congenital goitre, or anomalous great vessels in the superior mediastinum. These three conditions will be considered in more detail.

Thymic stridor is described by Cockayne (1947) as

'a respiratory stridor which may be present at birth or may develop during the early weeks of life. The stridor is more pronounced during inspiration, but both inspiration and expiration are stridulous. It

may be continuous or paroxysmal. During a paroxysm of dyspnoea, the obstruction to respiration may be so great that cyanosis supervenes with lividity of the lips and tongue and retraction of the ribs. . . . In other cases there are syncopal attacks, in which the child becomes limp and cold and the respiration becomes almost imperceptible. The child usually recovers after a few minutes, but may remain unconscious for two or three hours. Some cases of hyperplasia (of the thymus) have been associated with a persistent hoarse cough, noisy nasal breathing, paroxysms of rapid respiration, or attacks of choking with cyanosis during feeds.'

Although occasional cases of thymic compression of the trachea are met, most authorities would agree with Nelson (1945) when he states:

'There may well be some doubt that the thymus causes compression of the trachea in any significant number of instances. There is no doubt that the thymus may cause widening of the upper mediastinal shadow, which may be eliminated by shrinkage of the thymic tissue by roentgen-ray therapy. It is of considerable moment, however, that the size of this shadow bears no relationship to the occurrence of symptoms of respiratory obstruction.'

Stone (1945), citing Hill (1933), states that

'congenital goitre is associated with inspiratory stridor which disappears in sleep but is aggravated by crying. Upon effort there is engorgement of the cervical veins and cyanosis. The cry is likely to be hoarse, and difficulty in swallowing is a common accompaniment. The infant tends to throw the head backward to facilitate respiration.'

Diagnosis is straightforward unless the thyroid gland lies in the superior mediastinum.

Compression of the trachea and oesophagus by a vascular ring causes stridor from birth, which is chiefly inspiratory and less marked during sleep. There is often a cough and particular susceptibility to pulmonary infections. These infants feed slowly, because of the respiratory difficulty, and tend to lie with the head retracted. Dysphagia usually becomes evident when semi-solid or solid foods are introduced into the diet.

The similarity between the clinical pictures of tracheal compression from these three causes is

obvious, and it is the purpose of this paper to suggest that a vascular ring may be responsible for congenital stridor in a greater proportion of cases than is generally recognized. Its recognition is important because affected infants with severe symptoms dating from birth almost invariably die if untreated, as shown in the review of Griswold and Young (1949), whereas diagnosis of the condition is straightforward, and the principles of treatment are clear.

Comprehensive classifications of the types of vascular ring have been worked out by Neuhauser (1946) and by Edwards (1948), but the multitudinous varieties fall into three main categories. Besides these, symptoms of oesophageal and tracheal compression may be caused by a single anomalous vessel, as in Bayford's (1789) original case of dysphagia lusoria.

In the first group there is a single aortic arch passing behind the oesophagus to descend on the opposite side (usually a right arch with a left descending aorta), the ring being completed by a ligamentum arteriosum or patent ductus arteriosus on the side of the descending aorta (Figs. 1*a* and 1*b*).

In the second group there is one complete aortic arch and one aortic rudiment, joining to form a descending aorta on either side (usually the left). The ring is completed by a ligamentum arteriosum or patent ductus arteriosus on the side of the aortic rudiment (Figs. 2*a* and 2*b*).

In the third group there is a complete double aortic arch encircling the trachea and oesophagus. The aorta may descend on either side (usually the left), and a ligamentum arteriosum or patent ductus arteriosus may or may not enter into the composition of the ring (Figs. 3*a* and 3*b*).

The embryology, symptomatology and diagnosis of vascular rings are discussed by Dolton and Jones (1952), who describe a child with double aortic arch and patent ductus arteriosus who underwent successful operation. The case presented here was distinguished by the severity of the symptoms which left little doubt that the child would not have survived without surgical intervention.

Case Report

Terence B., a boy born on April 17, 1952, was admitted to the Bradford Children's Hospital under Dr. R. L. Langley on May 12, aged 4 weeks. He was a second child, delivered normally at full term weighing 7½ lb. There was no history of maternal illness during pregnancy. He had been fully breast fed for three weeks, then his feeds were supplemented with a cow's milk-and-water mixture on which he was gaining weight satisfactorily.

The history was of difficulty in breathing since birth. For three days before admission there had also been

cough, fever and attacks of cyanosis and dyspnoea, for which systemic penicillin had been given.

He was a well nourished boy weighing 8½ lb., with a capillary naevus nearly 25 mm. in diameter on the forehead. The rectal temperature was 98·4° F., the pulse rate 138 per minute and respirations 36 per minute. He was breathing with difficulty; inspiration was harsh and wheezy, and there was a sharp, dry cough. There was cyanosis of the fingers and ears, but no clubbing of the fingers. The percussion note of the chest was unimpaired: the breath sounds were harsh in all areas. The heart sounds were pure, and no abnormality was detected in any other system. Stridor was present at all times until operation and was always less marked during sleep. The child showed no tendency to hold the head retracted.

The pre-operative course was marked by alarming attacks, invariably following feeds, which he took rather slowly. At times these attacks occurred two or three times a day for several days in succession, but on one occasion he went for four weeks without an attack. The attacks, which lasted between five and 20 minutes, started with choking and congestion of the face. The initial cyanosis then gave way to pallor, the child became limp, and respirations ceased. The heart action became rapid and feeble, and during one attack the heart sounds became inaudible. Recovery began with infrequent, gasping respirations, followed by a gradual return of normal colour, normal respiratory rhythm, and consciousness.

The mechanism of these attacks appears to be as follows. The regular occurrence after feeds suggests that the precipitating factor was a distended oesophagus which compressed the trachea. Choking caused the oesophageal contents to spill over into the trachea, thus completely obstructing respiration, and causing unconsciousness. At this stage the baby was to all appearances dead, except that the heart continued to beat feebly. With approaching death the blood pressure would fall, thereby relieving the pressure of the constricting aortic ring, and allowing air to pass through the trachea once more. When this mechanism was appreciated, the baby was held upright for 10 minutes after the completion of each feed. On this regime he had only one attack in four weeks, and this was cut short by inverting the child and thereby draining the oesophagus and trachea.

These attacks were frequently followed by pulmonary infection, and the child had eight attacks of bronchopneumonia in five months. These responded to penicillin at first, and to aureomycin later, but there seems little doubt that the responsible organisms would have become resistant to all available antibiotics if the condition had been allowed to persist.

Laryngoscopy by Mr. J. S. Davidson in June, 1952, showed no abnormality of the larynx or vocal cords. In August an attempt was made to introduce mixed feeding, but it was evident that the child had great difficulty in swallowing thickened feeds and the trial had to be abandoned.

X-ray photographs with barium in the oesophagus (Figs. 4, 5 and 6) were taken by Dr. P. P. Franklin and showed clear evidence of compression by a vascular

1a

FIGS. 1a and 1b.—Plasticine model of the vascular ring formed by the right aortic arch, right retro-oesophageal aorta and ligamentum arteriosum. Fig. 1a—Left anterior oblique view. Fig. 1b—Right posterior oblique view.

1b

2a

FIGS. 2a and 2b.—Plasticine model of the vascular ring formed by the right aortic arch, left aortic rudiment and patent ductus arteriosus. Fig. 2a—Left anterior oblique view. Fig. 2b—Right posterior oblique view.

2b

3a

FIGS. 3a and 3b.—Plasticine model of the vascular ring formed by the double aortic arch. Fig. 3a—Left anterior oblique view. Fig. 3b—Right posterior oblique view (pulmonary artery omitted for clarity).

3b

4a

FIG. 4a.—Barium swallow: anterior view. Note bi-lateral constriction by the double aortic arch, and displacement of the oesophagus to the left below the constriction.

FIG. 4b.—Plasticine model: anterior view.

4b

5a

FIG. 6a.—Barium swallow: left lateral view. Note the posterior indentation caused by the left retro-oesophageal aortic arch.

FIG. 6b. — Plasticine model: left lateral view.

6b

5b

FIG. 5a.—Barium swallow: left oblique view. Note long, shallow, posterior indentation caused by the left retro-oesophageal aortic arch, and smaller anterior indentation.

FIG. 5b.—Plasticine model: left oblique view.

6a

ring, with displacement of the lower oesophagus by a right-sided descending aorta.

Operation was undertaken by Mr. J. S. Davidson at the Bradford Royal Infirmary on October 8, 1952. The great vessels were approached from the left side and a left aortic arch identified. There was no evidence of a patent ductus arteriosus. The left arch was divided behind the origin of the left subclavian artery, and the cut ends sprang apart as if under considerable tension. Stridor and dyspnoea, which had hitherto made the anaesthetist's task extremely hazardous, were dramatically relieved, and the lung was seen to inflate and deflate much more readily.

The post-operative course was prolonged. Mixed feeding was introduced before the patient's discharge on the fourteenth post-operative day, and he swallowed semi-solids without difficulty. He was also able to take his feeds without any delay. He still had a slight cough on discharge, but it was an occasional, effective cough in sharp contrast to the ineffective cough before operation.

He was readmitted a few days later with mild bronchitis, but before he had fully recovered from this he developed very severe pneumonia, to which he almost succumbed. Convalescence from this illness was protracted, although he ate well, cut several teeth and gained weight satisfactorily.

When finally discharged three months after the operation, he was completely free from clinical and radiological signs of pulmonary disease.

Discussion

Although the great majority of vascular rings cause compression of both trachea and oesophagus, symptoms of oesophageal obstruction are usually inconspicuous while the infant is taking an entirely fluid diet. Such cases therefore present primarily with symptoms of tracheal compression, namely, stridor from birth, cough and an increased susceptibility to pulmonary infections. When these symptoms are encountered, the possibility of a vascular ring being responsible should be kept in mind, particularly if no abnormality is detected on laryngoscopy. Postero-anterior and lateral radiographs of the chest will usually show narrowing or

displacement of the trachea at the level of the aortic arch, and contrast films with barium in the oesophagus will demonstrate clearly any constriction and give valuable information regarding the anatomical composition of any vascular ring. Angiocardiography may be necessary, but the introduction of iodized oil into the trachea should be avoided if possible. The recognition of this anomaly will not only allow of operative intervention, where this is indicated by the severity of the symptoms, but will also ensure that the infant is not subjected to the hazard of tracheotomy, which would increase the liability to pulmonary infection without in any way relieving the obstruction.

Summary

The causes of congenital stridor are considered, and the suggestion is made that tracheal compression by vascular rings arising from congenital anomalies of the great vessels may not be as uncommon as hitherto supposed.

A case is described in which severe symptoms of tracheal and oesophageal compression due to a bilateral aortic arch were dramatically relieved by operation.

This case is published by permission of Dr. R. L. Langley. To him, to Dr. E. Rosenblum and Mr. J. S. Davidson, I wish to express my gratitude for helpful advice and criticism. The photographs are by Mr. P. Harrison, M.S.R., of the Photographic Department, Bradford Royal Infirmary.

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