Congenital Atresia of the Larynx in Association with Prune Belly Syndrome

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SUMMARY: A case of congenital atresia of the larynx in association with the Prune Belly Syndrome is reported. Some aspects of the aetiology and management of congenital atresia of the larynx are reviewed.

Introduction

I wish to report a case of congenital atresia of the larynx in association with absence of abdominal muscles. This is the first case reported of this anomaly in a child with "Prune Belly" Syndrome.

Case Report

A 1300 g, male infant was born by emergency Caesarian Section to a 22 year old Primigravid woman. The labour was spontaneous, with breech presentation, at 31 weeks gestation. Ritodrine infusion had failed to stop the labour and foetal heart monitoring for several hours before operation was normal.

At delivery, the baby made gasping movements but the lungs did not expand. Intubation was impossible because of a solid bar of tissue across the larynx. An airway was not established and the baby died after 10 minutes.

Several abnormal features were noted as shown in Figure 1. These included low set dysplastic ears and a soft, distended abdomen. Post mortem examination confirmed laryngeal atresia. The fusion appeared to be below the true vocal cords. There was a small pinhole opening through the posterior part of the atretic area. The lungs were hypoplastic but the base of the right lung was partly expanded.

The brain was smooth with no gyral pattern.

No muscle was found in the abdominal wall. There was only one umbilical artery but no obvious structural abnormality of the renal tract.

Chromosome analysis of blood taken just before death revealed a normal male karyotype. There was no detectable specific IgM for rubella, toxoplasma, cytomegalovirus or herpes virus.

Discussion

Congenital atresia of the larynx is generally regarded as being a rare anomaly and in a substantial proportion of cases it is associated with other potentially fatal abnormalities.

Smith and Baine¹ report nine cases of atresia of

the larynx. They suggest that the anomaly comprises a spectrum of malformation which include the congenital webs between the vocal cords.

Three types of atresia are distinguished: Type one in which the supraglottic and infraglottic parts of the larynx are atretic; Type two in which the atresia, as in this case, is infraglottic; and

Fig. 1 Showing abnormal features in the male infant.

three in which it is glottic. In all cases there is a persistence of a posterior pinhole opening in the atretic area.

Walender^{2, 3} describes the formation of the larynx from a solid epithelial outgrowth from the pharynx. His hypothesis is partly speculative and based on the embryology of the rat larynx, but, unlike the more generally favoured view of the development of the larynx, can explain the different types of congenital atresia. In particular he describes the formation of a posterior opening, the pharyngo-tracheal duct which normally merges with the cavity of the definitive vestibule. No aetiological factors have been reported in association with laryngeal atresia or "Prune Belly."

The larynx first appears in the fetus at 25-28 days gestation and its cavity is closed until about the third month. The mesodermal segmentation ultimately responsible for the formation of the abdominal musculature starts at approximately 23 days. A defect occurring between 20-30 days could be responsible for both abnormalities. No possible cause was found in this case.

Hereditary influences play a part in congenital laryngeal atresia and several members of the same family have been described with varying degrees of atresia⁴.

If an airway can be established at birth, the prognosis for the child often depends on the

severity of any associated congenital abnormalities. Tracheostomy may be needed in the initial management, but, in the long term, correction should not be undertaken too early as this may cause serious disturbances in the growth of the larynx and affect final voice production. The larynx continues to develop after birth and early reconstructive surgery may affect the growth centres of the cartilaginous structures. The best results in laryngeal atresia appear to be in cases where definitive surgery can be delayed as long as possible⁴.

Congenital webs are usually isolated anomalies and present less of a management problem. There is a case for treating them early, as persistence of the web can cause a disturbance in the development of the larynx⁴, probably as a result of the increased effort needed for voice production.

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