The Journal of Laryngology and Otology
November 1999, Vol. **113,** pp. 1026–1028

Severe congenital laryngeal hypoplasia in a 45-year-old man

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Abstract
Congenital laryngeal anomalies are rare. They usually present in infancy and early childhood. We present a case of severe congenital hypoplasia, with multiple anomalies of the laryngeal cartilages in a 45-year-old man.

Key words: Abnormalities; Larynx

Case report

A 45-year-old male, who smoked 20 cigarettes a day, presented with hoarseness of voice. He said his voice had always been 'gruff' causing him social embarrassment. He became worried about throat cancer when a friend of his recently died of cancer of the larynx. He gave no history of stridor, dysphagia, aspiration or respiratory infections.

On examination his voice was hoarse and reduced in volume but intelligible. Head and neck examination revealed hypoplastic maxillae, and bilateral lop ear deformity. The laryngeal prominence and tracheal rings were not palpable. The rest of his physical examination was normal (Figure 1).

Flexible laryngoscopy showed no epiglottis, absent aryepiglottic folds and rudimentary vestibular folds. The arytenoids were huge, adducting and abducting in a repetitive manner during phonation and swallowing; his vocal folds were shortened and there was some salivary pooling (Figure 2).

Direct laryngoscopy confirmed the above findings but no other abnormality. He had a smooth recovery from the general anaesthetic.

A normal full blood count, ESR and a negative autoimmune antibody screen excluded the diagnosis of polychondritis. His genetic screen was negative. Chest X-ray was normal. His hearing was normal.

A lateral neck X-ray confirmed absent epiglottis and thyroid laminae (Figure 3).

A computed tomography (CT) scan confirmed that the thyroid laminae were absent and the cricoid cartilage was grossly thickened and elliptical in shape.

Dr Peter Phelps' opinion was sought and he confirmed the above findings; he also commented that, 'this could either be a congenital anomaly or possible relapsing polychondritis.'

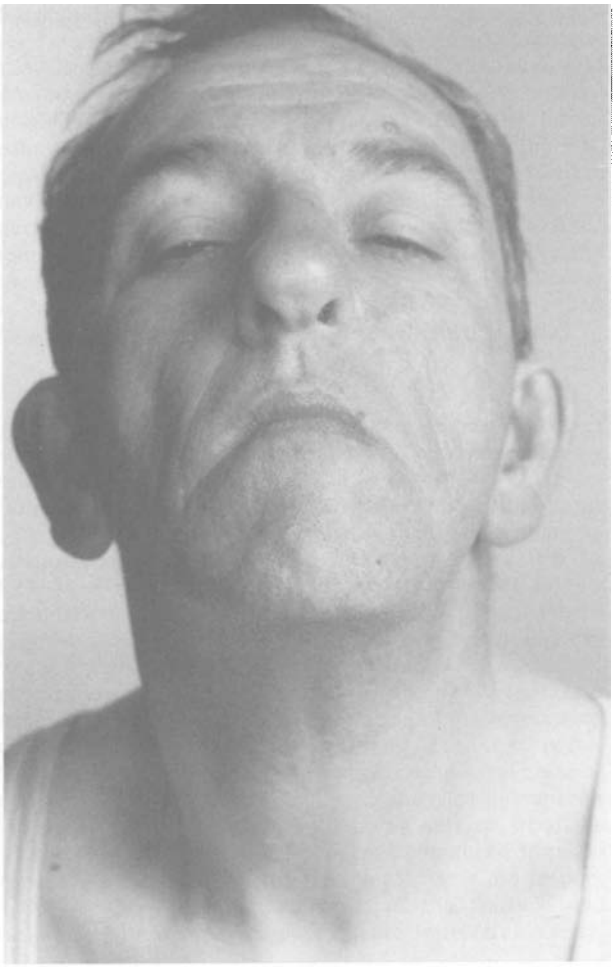

FIG. 1
Hypoplastic maxillae and lop ear deformity and absent laryngeal prominence.

Discussion

Congenital laryngeal anomalies are rare; usually presenting in infancy and early childhood (Goldenburge *et al.*, 1996; Tang *et al.*, 1996), with respiratory and feeding problems (Evans *et al.*, 1995).

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Accepted for publication: 1 July 1999.

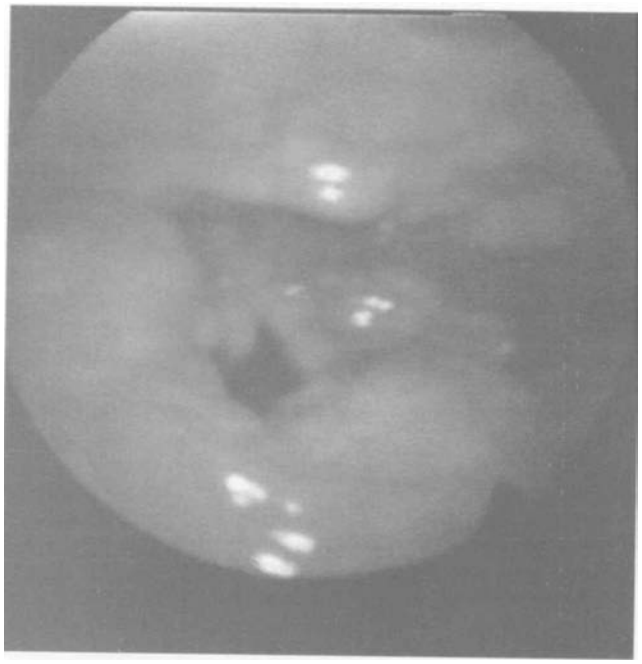

FIG. 2(a)

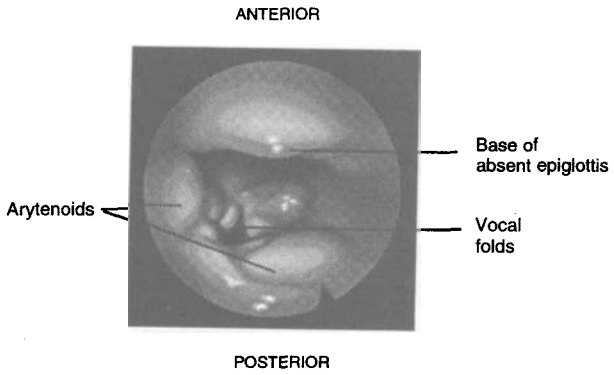

FIG. 2(b)

FIG. 2(a) and (b)
Endoscopic view of the larynx. Vocal cords during quiet breathing.

Our patient presented with severe multiple laryngeal anomalies at middle age. Despite the severity of his laryngeal hypoplasia, he had only a few symptoms, e.g. hoarseness of voice and frequent clearing of his throat due to some salivary pooling.

Embryologically the larynx develops in the floor of the cranial part of the foregut. An inverted U-shaped elevation, called the furcula (of His) develops behind the elevation of the tuberculum impar. The upper closed end forms the epiglottis, the open edge elongates into the aryepiglottic folds. More caudally a tube is formed by the fusion of the edges of the laryngotracheal groove, the extreme upper end of which remains patent as the laryngeal introitus. The adjacent caudal part forms the trachea, the lower end of which buds into stem bronchi of the future right and left lung buds, (Williams, 1989). Arrest or failure of growth during embryonic development would result in various laryngotracheal anomalies.

Laryngeal anomalies vary from laryngeal atresia which is incompatible with life (Gatti *et al.*, 1987; Tang *et al.*, 1996), to laryngomalacia which is the commonest cause of congenital stridor (Zeitouni and Manoukian, 1993). Varying stridor and feeding problems usually resolve by 18 months of age. However, progressive feeding problems with failure to thrive, stridor with cyanosis and apnoea require surgical intervention in the form of tracheostomy, hyomandibulotomy or epiglottoplasty (Zalzal *et al*, 1987).

Congenital absence of the epiglottis and bifid epiglottis usually present with infantile stridor (Reyes *et al.*, 1994; Goldenburge *et al.*, 1996), and are frequently associated with midline defects such as microcephaly, hypospadias, imperforate anus, laryngeal clefts and neuroendocrine disorders (Prescot, 1994). Morgan *et al.*(1993) studied 50 cases with Charge syndrome. They found a 38 per cent incidence of laryngeal abnormalities of whom 14 per cent required tracheostomy. They stressed the importance of early assessment of the upper airway to avoid the potentially lethal complications of aspiration.

Posterior laryngeal and laryngotracheal clefts are rare, they represent incomplete or failure of separation of the cranial part of the foregut from the larynx and trachea (Kaufman and Kohler, 1995). Congenital stridor and aspiration are the usual presentation (Evans *et al.*, 1995).

In our patient the larynx remained competent despite absence of supportive cartilaginous laminae. This emphasizes the importance of fibroelastic tissues as major factors in maintaining laryngeal patency.

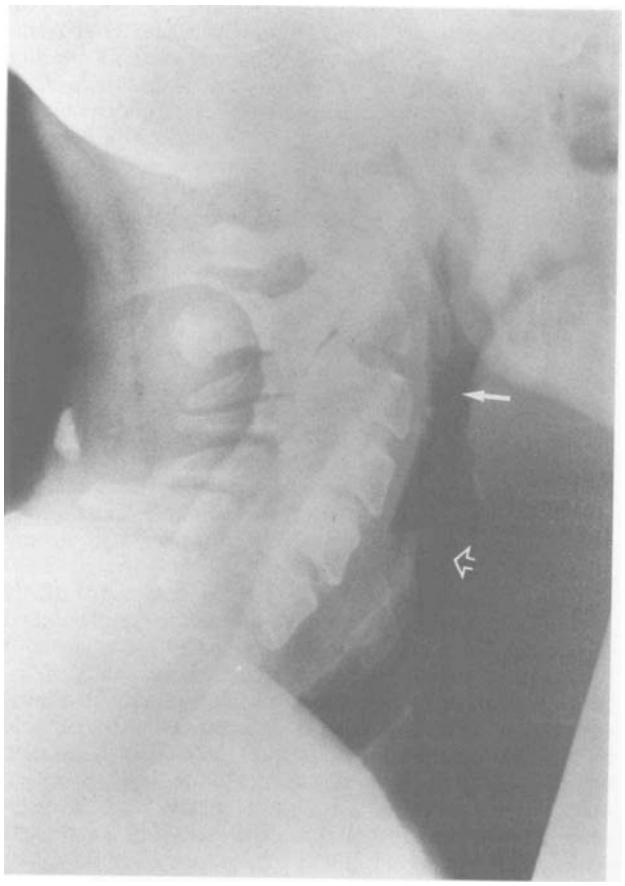

FIG. 3
Lateral soft tissue neck showing absent epiglottis (white arrow) and absent thyroid laminae (open arrow).

https://doi.org/10.1017/S002221510014589X Published online by Cambridge University Press

Conclusion

Severe congenital laryngeal hypoplasia is compatible with normal basic functions of the upper aerodigestive tract such as airway protection and swallowing although highly specialized processes, such as quality of voice may be affected.

Acknowledgements

The authors are grateful to Mr R. D. Leeming, F.R.C.S., Consultant ENT Surgeon for providing material for publication, to Dr D. Parker, Consultant Radiologist, Wrexham Maelor Hospital and Dr Peter Phelps, Consultant Radiologist, The Royal National Throat, Nose and Ear Institute, London for reviewing the scans. We also thank Miss Jocelyn Ashley for printing the text.

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