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# Lymphangioma of the tongue. A review of pathogenesis, treatment and the use of surface laser photocoagulation

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**Abstract**

Lymphangiomas of the tongue are rare tumours. Several reports of this interesting condition have appeared in the literature with varying modalities of treatment being employed to control tongue size. We present here our experience with seven children who have lymphangiomas of the head and neck with tongue involvement seen over the past eight years. If the tongue is large with protrusion outside the lip margins, we advocate early tongue reduction so as to promote proper speech and deglutition, reduce orthodontic problems and achieve good cosmesis. However, the natural history of these tumours is one of recurrent tongue enlargement secondary to infection and trauma, irrespective of surgical reduction. We describe our technique of surface $CO_2$ laser photocoagulation which has been successfully employed in controlling tongue size and removing superficial lymphangioma in all our patients. We propose therefore that this should be the mainstay of follow-up therapy in lingual lymphangiomas.

## Introduction

Lymphangiomas are uncommon developmental anomalies and involvement of the tongue is rare. Fewer than 50 cases have been reported in the English literature and a variety of treatment modalities have been advocated. We present here our experience with seven children over the last eight years who had lymphangioma of the head and neck with tongue involvement. The pathogenesis of these interesting tumours is reviewed and a treatment plan suggested. The carbon dioxide ($CO_2$) laser has been successfully employed in controlling tongue size both by wedge resection and by removing superficial lymphangiomas by surface laser photocoagulation.

## Pathogenesis

Although cystic hygroma was first described by Redenbacker in 1823 and Virchow gave the first accurate description of a lymphangioma of the tongue in 1854, it was not until 1901 when Sabin (1901; 1909) postulated the embryogenesis of the lymphatic system that the lymphatic origin of these unique tumours was recognized.

There are two main theories on the origin of the human lymphatic system. Sabin postulated that all lymphatic channels develop from venous endothelium. Peripheral lymphatic vessels then arise in a centrifugal fashion from endothelial sprouts, elongations and branchings (Batsakis and Rice, 1981). Huntington and McClure (1910) and later Kampmeier (1931) favoured the concept of centripetal development, wherein lymphatics originate from a confluence of perivenous mesenchymal spaces that secondarily open into the venous system. Whatever the correct theory, the aetiology of cystic hygromas also remains controversial (Ravitch and Rush, 1986). Their infiltrative nature seems to suggest a neoplastic process, although their lack of unremitting growth is strongly against it. Certainly the precursor appears to be a failure of the lymphatic spaces to join a central collecting system (lymphatic or venous) that results in sequestration from primary lymphatics. They are thus best regarded as hamartomas arising in embryonic sequestra (Batsakis and Rice, 1981).

Confusion also abounds regarding the classification of lymphangiomas. Landing and Farber (1956) recognized three groups:

1. The localized 'lymphangioma simplex' consisting of thin walled capillary-like channels.
2. 'Cavernous lymphangioma' which has large dilated lymphatic channels.
3. 'Cystic lymphangioma' or hygroma, which comprises endothelial-lined cysts ranging in diameter from a few millimetres to several centimetres.

Clinically, classifying cystic hygroma as a separate group may be justifiable. Pathologically, however, the finding of mixed patterns and the progression of one form to another strengthens the belief that these lesions are basically similar. Lymphangiomatous forms are found in tissues of a restrictive nature like skeletal muscle and fibrous connective tissue, whereas cystic lesions develop in areas where loose areolar tissue allows expansion (Bill and Sumner, 1965), for example in the neck.

The intimacy of the venous and capillary systems account in some instances for the active participation of the former in certain lesions, giving rise to terms like lymphohaemangioma. Generally, however, lymphangiomas are recognized by thin-walled endothelial-lined spaces containing lymph and a few or no erythrocytes.

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The neck appears to be the favoured site for lymphangioma. Tongue involvement is extremely rare and less than 50 cases have been reported in the literature. A review of 40 cases over a 30-year period by Kennedy (1989) yielded only one case located within the tongue. Lymphangioma, however, is the commonest cause of true macroglossia (Rice and Carson, 1985). Fifty to sixty per cent of all lymphangiomas present at birth and approximately 80 per cent will be detected before the age of two years (Bailey, 1990). Lymphangioma of the tongue may be localized or diffuse and this latter type constitutes the majority. Macroscopically the tongue takes a granular appearance being studded with multiple transparent lymph-filled cysts. Occasionally there is bleeding into these from trauma and they appear as interspersed haemorrhagic blebs. The enlarged tongue usually protrudes through the lip margins and becomes cracked and dry. Microscopically, there are abundant cystic endothelial-lined spaces containing blood or lymph found in the epithelial, subepithelial and muscular tissue. Due to dilatation of the enlarged lymphatics, atrophy of the striated tongue musculature occurs. The tongue enlarges with each episode of upper respiratory tract infection (URTI) and when protruding through the lip margins is also exposed to frequent trauma. These repeated episodes of infection and trauma lead to fibrous tissue being laid down. This thus leads to further dilatation of lymphatic channels and permanent enlargement of the tongue.

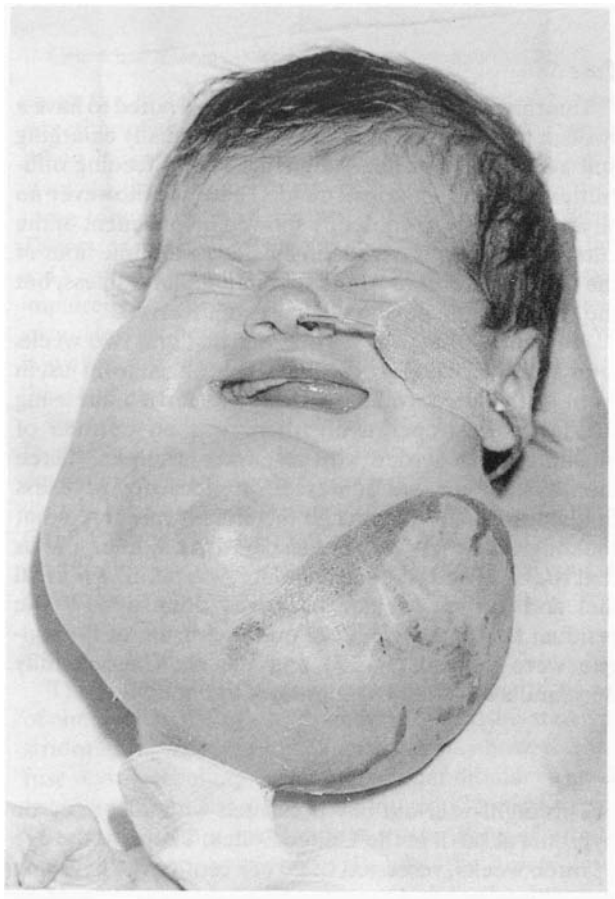

FIG. 1
Case 1 as she appeared at birth.

## Case reports

*Case 1*

This girl who is now eight years of age presented to us at birth with a huge cystic swelling under the jaw and extending into the floor of the mouth (Fig. 1). The tongue was not noted to be involved at this stage. A CT scan showed a huge multicystic mass crossing the midline and extending into the base of tongue. Six weeks later, a surgical debulking of the cystic hygroma was carried out using a high cervical incision. The mass was removed as completely as possible, leaving decapped cyst walls around the right carotid sheath, facial and hypoglossal nerves. Three days post-operatively she developed inspiratory stridor and had to be intubated urgently. Endoscopy under general anaesthesia showed lymphangiomatous involvement of the vallecula and supraglottis, and a tracheostomy was performed to secure the airway. A year later she contracted rubella with sudden enlargement of the residual cystic hygroma. At this time tongue involvement was seen. An excision of the lymphangioma from the right submandibular triangle and floor of mouth was attempted, but was made difficult by obscured landmarks due to the previous surgery and diffuse tissue infiltration by the hygroma.

In the next six years, there were 15 admissions for repeated episodes of tongue enlargement, usually following an URTI. The size of the tongue was however kept in check by surface laser photocoagulation (Fig. 2). A bronchoscopy done a year ago showed some lymphangioma involving the subglottic area with narrowing and she was therefore deemed unfit for decannulation. Her general condition appeared good, she was doing well in school and had no problems with speech. She was still having regular orthodontic follow-up (Fig. 3).

*Case 2*

This eight-year-old boy had noisy breathing noted at one week of life but this was attributed to laryngomalacia. The Health Visitor noted a mass in the neck at two weeks of age and the child was seen at the local hospital. A diagnosis of cystic hygroma was made and the child referred to us for further management two months later. On clinical examination, the child had a large mass in both submandibular triangles displacing the tongue. Endoscopy of the laryngopharynx showed a large lym-

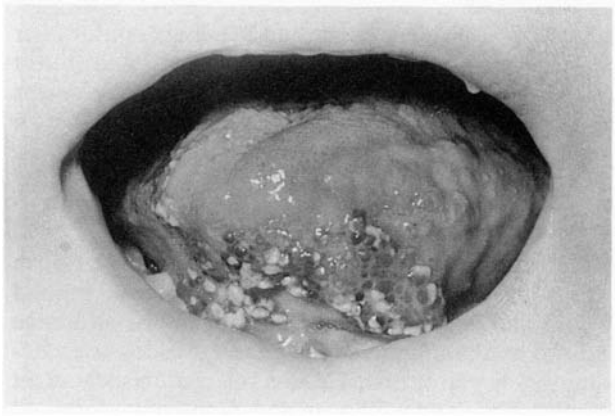

FIG. 2
Case 1. Her tongue size was controlled with repeated laser photocoagulation. Note residual lymphangioma on ventral surface.

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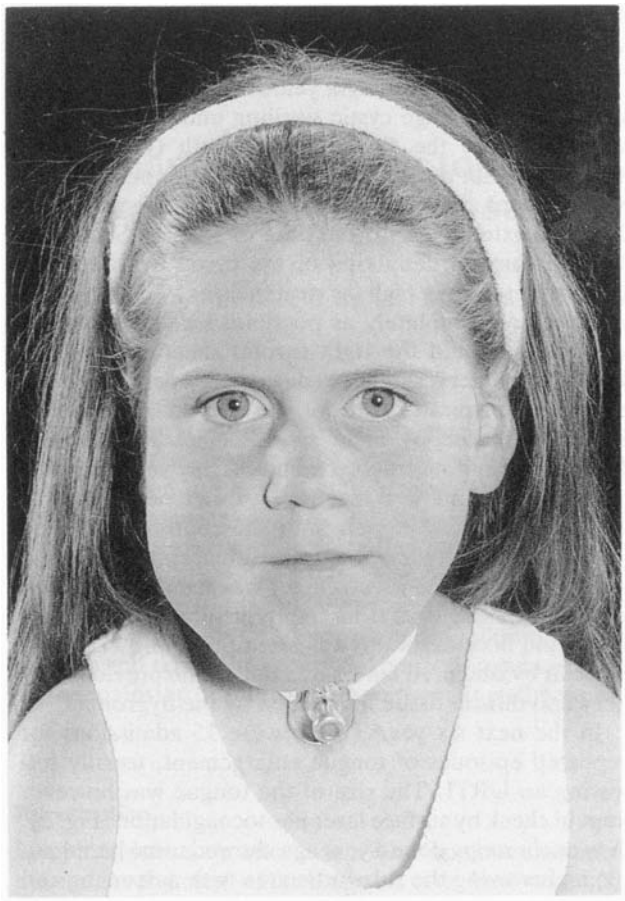

FIG. 3
Case 1 as she is now with a good general appearance and tongue wholly within lip margins.

phangioma involving the floor of mouth, base of tongue and vallecula. Post-operatively the child had increasing respiratory difficulties and required tracheostomy done three days later. After three months, a large cystic hygroma involving the left submandibular and parotid regions was seen with a small swelling on the right. Endoscopy showed lymphangiomatous involvement of the soft palate, both tonsils, tongue base, epiglottis and the supraglottis. Four months later, excision of almost the whole mass from the neck was achieved. Cysts around the carotid vessels, however, were marsupialized.

In the next seven years, he had 25 admissions for laser photocoagulation of his tongue. He was decannulated at the age of five. His speech is good, his dental problems are regularly being attended to and his tongue, although granular in parts, is of near normal size.

*Case 3*

This five-and-a-half-year-old boy presented at birth with a massive hygroma of the neck, both parotid regions and floor of mouth. The tongue was also huge and two weeks later, due to increasing stridor, a tracheostomy was performed. He was referred to us at eight months of age and a CT scan and endoscopy confirmed the clinical findings but also showed lymphangioma involving the pharynx with forward displacement of the tongue. The right side of the neck and cheek was more prominent than the left. Three months later he had the first of 11 laser photocoagulations stretching over four years. Just recently he underwent wedge resection of his tongue and it is now wholly within the lip margins with good functional and cosmetic results. He is continuing treatment for his other problems.

*Case 4*

This girl, now five years of age was noted to have a large tongue and left sided neck swelling at birth but was asymptomatic. Endoscopy two weeks later showed a large cervical cystic hygroma involving the floor of mouth, whole of the tongue and epiglottis. In the next six months she had two laser treatments to the tongue, but as there were signs of respiratory obstruction and no decrease in tongue size, a wedge resection of the tongue was undertaken a month later. She was well after this but had three further admissions for sudden neck enlargement and on the last occasion require a tracheostomy. Debulking of the left neck mass was carried out three months later. A year later, a tonsillectomy was carried out as she was having recurrent attacks of tonsillitis associated with tongue enlargements (Fig. 4). A tongue tip reduction was carried out and the child has been trouble-free for the past year with a tongue wholly within the mouth. Even the right neck swelling has partially subsided (Fig. 5). She is meanwhile on orthodontic and maxillofacial follow-up. If she continues to remain well and the hygroma remains stable in size, a decannulation will be attempted in the near future.

*Case 5*

This three-and-a-half-year-old girl was noted to have a swollen tongue at six months of age, gradually enlarging and associated with bleeding (Fig. 6) and feeding difficulties with speech impairment. There was however no airway difficulty. Endoscopy showed involvement of the anterior two-thirds of the tongue and part of the floor of the mouth. There was some submandibular fullness, but the rest of the head and neck was uninvolved.

A tracheostomy was carried out first and two weeks later a vertical wedge resection of the tongue from just in front of the circumvallate papilla was carried out using the laser. Post-operatively there was an episode of bleeding which settled with rest and antibiotics. Three months later she was admitted for endoscopy to assess fitness for decannulation. At this time it was noted that the tongue size was well reduced and the mother told us that the child had stopped drooling, was taking a normal diet and her speech had improved dramatically. The residual haemorrhagic blebs on the dorsum of the tongue were lasered (Fig. 7) and the child successfully decannulated. She now continues life normally.

*Case 6*

This eight-year-old boy presented with a large cystic hygroma at birth in the United States. There, at the age of three weeks, resection of 90 per cent of the hygroma was undertaken and a tracheostomy done. At 14 months of age, exploration of the left side of the face was undertaken, but abandoned due to encroachment of the mass

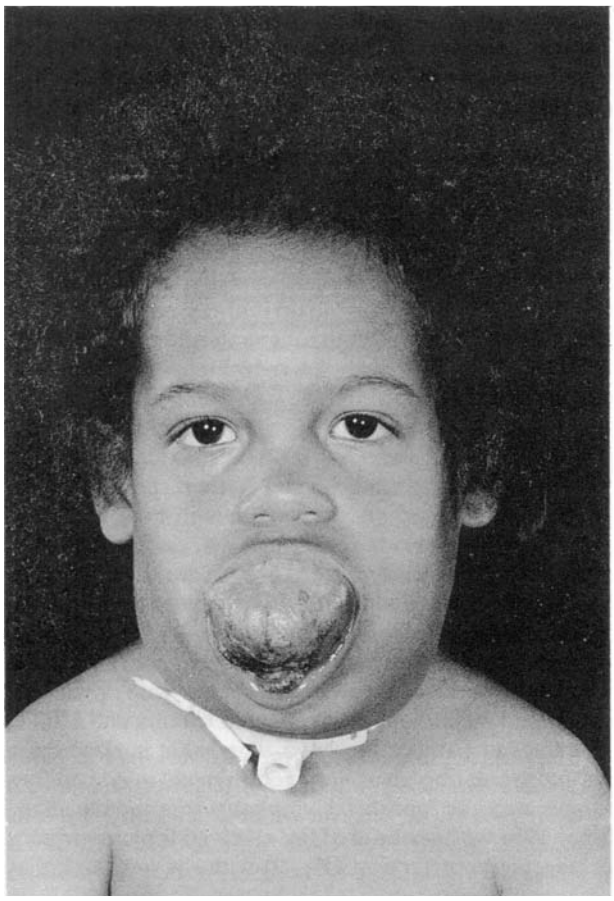

FIG. 4
Case 4 had a large protruding tongue after repeated URTI.

on to the facial nerve. At 22 and 24 months, further resection of the neck on the left side was undertaken. When he came to see us, the main mass of hygroma involved the left side of the face and upper neck with some tongue, floor of mouth and palate involvement. There was minimal involvement of the larynx but he had a IX and X cranial nerve palsy with consequently impaired swallowing, probably due to the previous surgery. He was being fed by a feeding gastrostomy. We, however, felt that no further surgery was warranted at this stage.

He was decannulated at the age of five and is now feeding orally. He had two admissions for laser photocoagulation to the tongue and left buccal mucosa and also required surgical closure of the tracheocutaneous fistula. When reviewed six months ago, the lymphangioma appears stable and he receives occasional courses of antibiotics for recurrent URTI.

*Case 7*

This four-year-old boy was transferred to us at the age of one week with facial and neck swelling associated with stridor. Examination under anaesthesia showed a diffuse cystic swelling of both submandibular regions extending on to the cheeks, right parotid and both lips. There was further involvement of the floor of mouth, base of tongue, epiglottis, vallecula, the aryepiglottic folds and ventricles. On observation in the Intensive Care Unit, his stridor improved and he was discharged home without a tracheostomy.

Two years from his first visit, he presented with some stridor and the face and neck hygroma appeared to be somewhat larger. The hygroma was found to be involving only the centre on the dorsum of the tongue (Fig. 8). The left external auditory meatus was also involved giving a cobblestone appearance. The tongue was lasered in the usual fashion. Six months later there was a further episode of enlargement. The large central tongue hygroma was excised using the laser and the rest lasered in the usual way. When seen six months ago, the ear involvement has disappeared and the tongue remains small.

## Discussion

The management of a child with lingual lymphangioma involves the various clinical features so well represented in our seven young patients. Respiratory obstruction was seen in five of them and necessitated a tracheostomy. A similar number had tongue protrusion which led to the secondary problems of drooling, difficulty in chewing and swallowing, poor speech and orthodontic abnormalities. The combined effects of these can sometimes make the child a social outcast (Dinerman and Myers, 1976) requiring psychological support. Management is therefore complex and requires a team approach involving different specialities.

Several methods of treatment have been advocated for the lesion itself. Radiation therapy, electrocoagulation, cryosurgery, steroids and injection of sclerosants

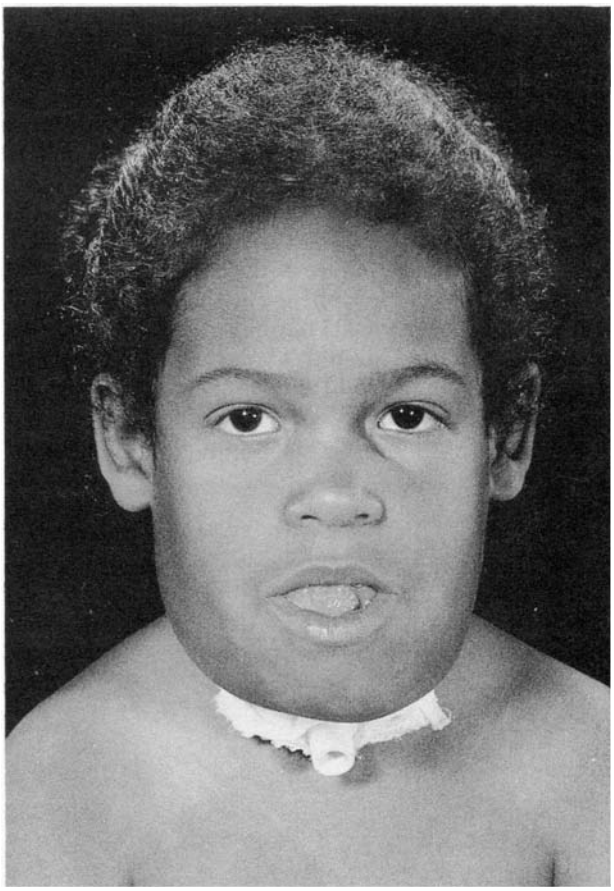

FIG. 5
Case 4 after a second tongue reduction.

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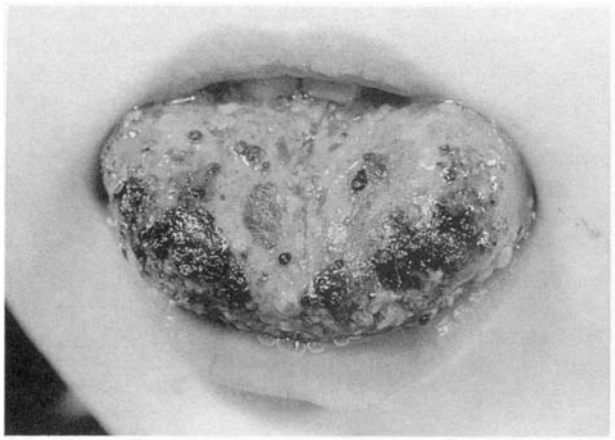

FIG. 6

Case 5 had a swollen tongue from six months of age. Note the haemorrhagic blebs on the lateral aspect.

are some of those which have been used. Some of these have fallen into disrepute because of poor results and unacceptable complications. The most effective treatment is surgery (Emery *et al.*, 1984; Cohen and Thompson, 1986; Ravitch and Rush, 1986; Kennedy, 1989) and has the following aims: (1) to restore adequate breathing and swallowing; (2) to leave a tongue capable of normal speech, taste, sensation and orofacial development (Velcek *et al.*, 1979) and (3) last but not least, to achieve a good cosmetic result.

Although sporadic cases of spontaneous regression have been reportd (Broomhead, 1964; Ninh and Ninh, 1974; Emery *et al.*, 1984), this is not an outcome to be anticipated. We recommend early surgery for the neck as it is technically easier before the tumour has grown and further involved normal tissue and before repeated infection has caused increased fibrosis. Complete excision is usually not possible, but as much as possible of the tumour must be removed while preserving vital structures. There is no justification for sacrificing these in view of the benign nature of the condition. Cysts left behind should be widely opened and as much of the lining as possible removed. Four of our patients had surgery to the neck and in a fifth who needed it, the parents declined, leaving the child with a significant neck mass.

When the tongue is grossly enlarged, the best approach is a vertical wedge resection, the amount to be removed being tailored to the needs of the individual. The initial incision is made in front of the circumvallate papillae using the laser as a scalpel (Sharplan 1025) at a distance of about 0.5 cm, using a spot size of 0.1–0.3 mm, a power of 20–25 W in continuous mode to give a power density between 50,000–75,000 W/cm$^2$. This power is ideal for both cutting and haemostasis (Carruth, 1985). The incision cuts through tongue tissue and a wedge is removed in the shape of a boat's keel, ensuring protection of the lingual and hypoglossal nerves which are deep. The tongue tip is always included in this technique and only enough tongue should be removed to allow it to fit back into the mouth so that occlusion, speech and swallowing are possible. The remnant tongue emargins are sutured in the midline. Except for one child (Case 7) who had a single episode of bleeding two days post-operatively which settled spontaneously, we have had no other complications in the immediate postoperative period. On follow-up, remnant tongue function is remarkably good. Three of our seven patients had the tongue size reduced with excellent results (Figs. 5 & 7).

In most instances, as was our own experience, the tongue can suddenly grow rapidly in size with or without associated bleeding due to repeated trauma and URTI. In all seven patients we have found that surface laser photocoagulation gives very good results in controlling tongue size, the superficial lymphangioma and bleeding. Since 1984 we have lasered the involved tongue surfaces in a serpiginous fashion (Fig. 9) with the laser at a dis-

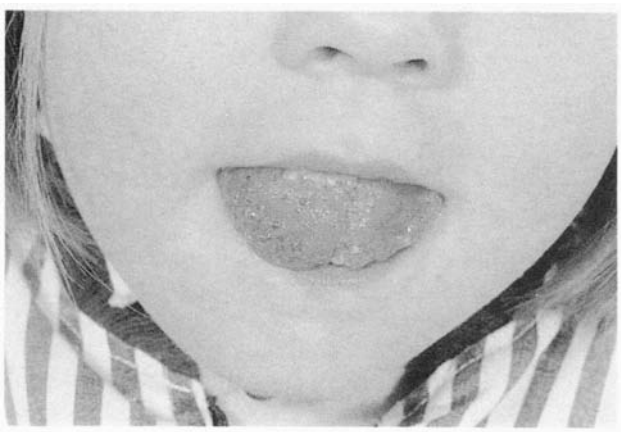

FIG. 7

Case 5 after laser photocoagulation.

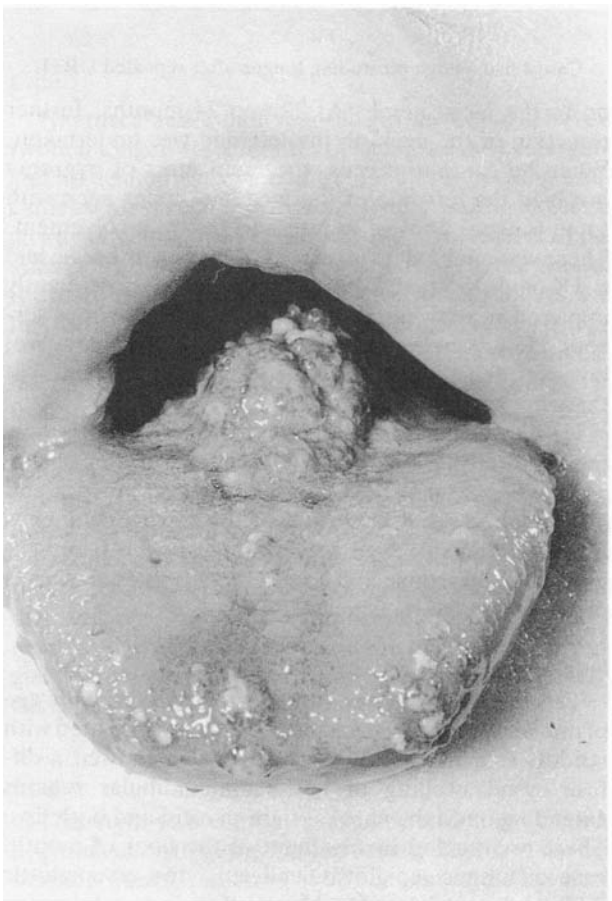

FIG. 8

Case 7 showing 'nodular' lymphangioma.

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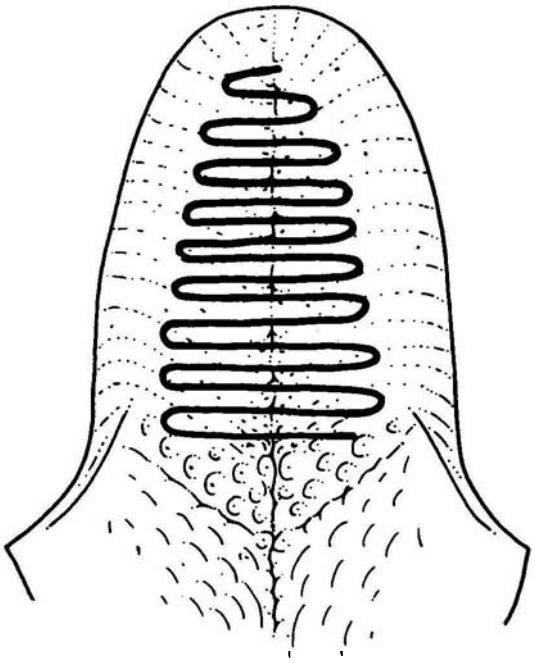

FIG. 9
Technique of laser photocoagulation.

tance of 1 cm from the target area, using a spot size of 0.5 mm and power of 5 W in continuous mode, leaving a gap of 1–2 mm between the margins to prevent areas of tissue necrosis resulting from overlapping deep penetration and scatter (Dixon, 1986). The power density is about 700 W/cm$^2$ and there is minimal post-operative oedema and pain and the patient is usually sent home the next day.

We have not seen bleeding as a complication after surface lasering as the laser seals all blood vessels up to 1 mm in diameter, and the lasered area heals beautifully. Repeat lasering is done at intervals varying between one to six months, depending on tongue size and the degree of haemorrhagic bubbling on its surface. Dixon *et al.* (1986) who had nine patients with various vascular malformations of the tongue and on whom they used the Argon and Nd:YAG laser in an almost identical fashion, reported similar results. In four of our children who did not have gross tongue involvement, this method of lasering has been the mainstay of management for periodic tongue enlargement over periods ranging up to six years.

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