

# CE Course: Abbey's Story: Cystic Hygroma

By April Turner, RDH, MSDH

## Learning Objectives:

- Define lymphangiomas and their cause.
- Discuss the oral/head and neck symptoms of cystic hygroma.
- Discuss risk management strategies for the cystic hygroma patient for dental/dental hygiene treatment and the potential complications.

Abbey had a sensitive tooth and was pretty sure it was a cavity and this caused anxiety because the last visit to the dentist was 15 years ago. Abbey has cystic hygroma lymphangioma and dental treatment is very difficult. Additionally, Abbey struggled finding a dentist who would even agree to treat the case. Abbey's condition is rare and most dentists were not willing to take the risk since those with cystic hygroma lymphangioma are prone to infection and swelling of the tongue.

Lymphangiomas are malformations of the lymphatic system and are believed to result from a blockage of lymph channels during fetal development.<sup>1</sup> Lymphangiomas are classified into three types based on the size of the lymphatic cavity: microcystic (capillary), macrocystic (cavernous), and cystic hygroma.<sup>1</sup> In the oral cavity, microcystic lymphangiomas present as clusters of vesicles on the buccal mucosa and tongue while macrocystic lymphangiomas and cystic hygromas appear below the mylohyoid muscles as swelling in the neck.<sup>2</sup> Macrocystic lymphangiomas are differentiated from cystic hygromas based on size and location; cystic hygromas are located in loose neck tissue and have multiple fluid-filled cysts.<sup>1</sup> Most lymphangiomas occur in the neck and face and are diagnosed at birth with the remaining diagnosed by age 2.<sup>3</sup> Most children born with cystic hygromas have a normal genetic makeup but

some studies have found increased incidence of Turner Syndrome, Down syndrome, or Noonan syndrome in these children.<sup>3</sup> Lymphangiomas are benign lesions and the treatment of choice is surgical removal particularly when lesions affect the respiratory abilities of the child. The lesions do not usually recur unless the surgeon is unable to remove all of the lesion.

When Abbey was born, there was a red lesion which the pediatrician thought was a strawberry hemangioma; strawberry hemangiomas usually resolve by age 3. Abbey's mother was advised to monitor it. When Abbey was 3, things changed. "I was 3<sup>1/2</sup> years old and my tongue grew, which happened pretty rapidly. Within about 1-2 months it was outside my mouth, about the size of a donut. We just covered it with gauze so it wouldn't dry out." During this time, a tracheotomy and feeding tube were placed since the tongue was affecting the ability to breathe and eat. Abbey had a series of laser and probe frequency treatments on the tongue to help shrink it. These treatments helped, but not enough for the tongue to go back into Abbey's mouth. "When I was 5, I had my tongue surgically cut and reshaped to fit in my mouth, [which] took about 3-4 months of healing. Afterwards injections in the two types of cysts (micro cystic and liquid filled ones) in my neck/under jaw area [were done] to help shrink them. In my teen years I had several radio frequency treatments on my tongue that helped maintain and shrink further but, since the main surgery when I was 5, [my



*Figure 1: Protrusion of the tongue similar to Abbey's case from Eivazi & Werner.<sup>7</sup>*

tongue] has stayed inside my mouth.” Prior to the tongue surgery, Abbey stated the majority of the primary teeth were extracted because they were causing the tongue to bleed. See Figure 1 for a photo similar to Abbey’s tongue condition.

Dr. Eugenius Redenbacher, a German physician, was the first to describe lymphangioma lesions in an article published in 1828. Then, in 1843, German surgeon, pathologist, and university lecturer Dr. Adolph Wernher first coined the term “lymphangioma,” described cystic hygromas, and, in 1877, proposed the first classification system for lymphangiomas.<sup>4</sup> Dr. Werner concentrated on pathology and lecturing since an eye infection restricted his ability to operate. He published multiple textbooks and articles on conditions of the head and neck as well as the benefits of vaccination.<sup>4</sup> Dr. Wernher’s classification system was used by most physicians until 1982 when Mulliken and Glowacki presented a cell-based system which classified lesions as hemangiomas and vascular malformations including lymphangiomas.<sup>2</sup>

The prevalence of cystic hygroma is 1 in 800 pregnancies and 1 in 8,000 live births; many pregnancies with lymphangiomas are lost before reaching full term.<sup>5</sup> Long term prognosis is good and depends on complete surgical removal and whether the lymphangioma is near critical structures such as vessels, nerves, and the airway.<sup>2</sup> Cystic hygromas and other lymphangiomas are usually removed surgically and do not recur. These patients, when seen in the dental office, may have residual scarring from surgery as a child, but no lasting effects. However, lymphangiomas can recur if not removed completely because the tumor was too close to vital structures. Lymphangiomas can have spontaneous regression, progress slowly, or develop hemorrhage in the cyst. If surgeons are unable to remove a cystic hygroma completely, they may choose to aspirate the cyst or use sclerotherapy which requires injection of medication into the lesion which causes it to shrink (See Figure 2). In some cases, the patient experiences macroglossia and has to have a glossectomy or has parotid gland involvement which needs excision.<sup>3</sup> Although most lesions present in the neck, cystic hygromas may also develop in the parotid gland and are the second most common congenital mass of the parotid gland.<sup>1</sup> Dental professionals need to be aware of past tongue surgeries or partial parotid gland removal in order to tailor dental treatment. In rare cases, patients who have had surgical excision at or near



*Figure 2: Female patient with lymphatic malformation treated over time with surgery, laser therapy, and sclerotherapy from Eivazi & Werner.<sup>7</sup>*

the parotid gland have experienced Frey Syndrome.<sup>6</sup>

Abbey, now 25, still has some residual effects from the lymphangioma surgery and lymphatic system involvement (See Figure 3). The dentist who agreed to treat Abbey was a pediatric dentist specializing in special needs cases. Abbey had multiple consults with the dentist and Abbey’s physician sent documentation outlining the details of Abbey’s condition. “Basically [my physician] said that as long as they’re able to avoid touching my tongue when possible everything would be fine.” Although Abbey is happy to have found a dentist willing to manage their care, dental appointments can be challenging. When asked about the last dental visit and what dental professionals



*Figure 3: Photo of Abbey today (used with permission)*

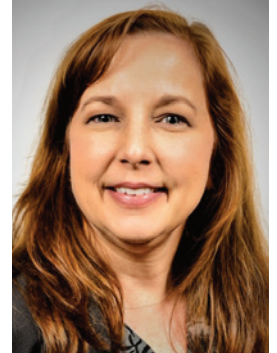
should know, Abbey said “It’s very difficult with dental work because [any] slight trauma toward my tongue can cause swelling, bleeding, or an infection. The main concern is [that] everything is sterile and having as little contact with my tongue as possible. During my last trip the vacuum tube they put in your mouth caught onto the side of my tongue for maybe half a second and that’s what triggered a two-week infection that caused me to miss work and go on steroids.” Abbey’s physician did not require antibiotic premedication for routine dental care such as hygiene services, however, Abbey has to have third molars removed. For this procedure, Abbey will need to take prophylactic medications and a two-week medical leave from work. Although cystic hygroma lymphangiomas are rare and most patients do not experience residual effects, dental hygienists need to take a careful medical history to determine how best to treat these patients. Further, it is valuable to seek medical clearance in order to minimize trauma and infection in patients with cystic hygroma lymphangioma.

## References

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## About the Author

April Turner, RDH, MSDH is the National Board Success Manager at West Coast University in Anaheim, California and currently teaches Clinical Seminar and Applied Research Lab. She has been a dental hygiene faculty member at WCU since 2011 and was in clinical practice from 1991 to 2019. April received her Bachelor of Science degree in Dental Hygiene from Loma Linda University and her Master of Science degree in Dental Hygiene from the University of Texas Health Science Center San Antonio. She is in the process of completing a Doctor of Education degree through Grand Canyon University. April is a member of the California Dental Hygienists’ Association, Orange County Dental Hygienists’ Society, American Dental Hygienist’s Association, and the American Dental Education Association.



Home Study Correspondence Course

# “Abbey’s Story: Cystic Hygroma”

Circle the correct answer for questions 1-10

- Lymphangiomas are malformations of the lymphatic system.
  - True
  - False
- Lymphangiomas are believed to result from a blockage of the lymph channels during fetal development.
  - True
  - False
- The prime risk for dental treatment for cases with cystic hygroma lymphangioma is:
  - Elevated blood pressure
  - Infection and swelling of the tongue
  - Gastric distress
  - Pigmentation of the tongue
- Lymphangiomas are classified into three types: macrocytic, microcytic and cystic hygroma.
  - True
  - False
- Most lymphangiomas occur in the face and neck and are diagnosed:
  - During childhood or by age twelve
  - During adolescence or by age eighteen
  - At birth or by age two
- Lymphangiomas are benign lesions with the treatment of choice being:
  - Radiation
  - Chemotherapeutics
  - Surgical removal
- When the treatment of choice is not successful, an alternative treatment for cystic hygroma is aspiration of the cyst or (sclerotherapy) injection of medications to cause shrinkage.
  - True
  - False
- Although most lesions present in the neck, cystic hygromas may also develop in the thyroid gland.
  - True
  - False
- Abbey’s case study and subsequent dental treatment resulted in:
  - no complications
  - Infection and steroid treatment
  - Loss of work for two weeks
  - Both b and c
- Considerations for dental and dental hygiene treatment for patients with cystic hygromas include:
  - Detailed medical and dental history
  - Medical consultation for potential premedication
  - Strategies for avoiding contact with the tongue
  - All of the above

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