

Oral Health Issues Facing Individuals with CdLS

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A daunting task facing parents of a child with multiple physical and developmental challenges is assembling a strong healthcare support team that can guide them through the maze of therapies, treatments and unknown difficulties.

Very important members of that team are the dental specialists who can address the oral health concerns affecting children and adults with Cornelia de Lange Syndrome (CdLS). Common oral and dental abnormalities can have a profound effect upon the overall health of patients. Feeding struggles (associated with a high prevalence of oral clefting), delayed eruption of the teeth, and proper swallowing, demand an early establishment of a “dental home.” An early consultation with a pediatric dental specialist is important. Pediatric dentists are trained in all aspects of assessment and treatment associated with children who possess special health care needs.

Physical Issues

CdLS presents an array of different abnormalities affecting the mouth and craniofacial structures. The jaws, teeth and oral soft tissues can be altered due to poor growth and development.

Children with CdLS are at a higher risk for cavities, orthodontic problems and self-inflicted oral injuries.

It is common to have clefting of the palate, which can affect infant feeding, speech development and proper jaw growth. Ankyglossia, or tongue-tie, is found in many children and can greatly affect speech development and proper eating. This condition can also be a factor in excessive drooling, making it difficult to swallow effectively. Periodontal, or gum, disease can be detected in a great number of individuals with CdLS. This becomes a serious threat to good oral health well into adulthood and can cause the early loss of the permanent teeth.

The lower jaw can be short. The upper jaw can be very narrow and positioned forward on the skull, causing misalignment and crooked teeth.

The upper and lower jaws are also usually quite narrow with a very high arch to the palate. This kind of arch configuration makes for very crowded teeth. Some people feel that this contributes to tongue thrusting due to such a small space for tongue positioning and resting.



There are many other factors involved with tongue thrusting, such as a constricted airway, large tonsils or neurological abnormalities leading to a poorly developed swallow mechanism. This can also lead to excessive drooling. Tongue thrusting and drooling are common in children with CdLS. A consultation with a speech pathologist, myofunctional therapist or physical therapist could be helpful.

Erosion

Enamel erosion can be seen in individuals who experience gastroesophageal reflux disease (GERD). The upward flow of stomach acids constantly bathe the teeth and cause the protective layer of enamel to dissolve away. This allows cavities to develop. When pitting of the enamel is seen on the backside of the front teeth, due to the acids, the pediatric dentist will use various medications, like fluoride, to control or even reverse the acid damage.

Tooth grinding, known as bruxism, compounds the erosion problem, causing further loss of tooth structure. The pediatric dentist can make a retainer-like device that can be worn to prevent the bad effects of tooth grinding.

Teeth Growth

Children with CdLS do have a great number of orthodontic concerns. The growth and development issues associated with the syndrome not only affect the height and weight of the children, but the growth and maturity of the mouth and dentition.

The primary, or baby, teeth have a tendency to come into the mouth later than we would typically expect. These teeth often do not fall out when they should. This causes the permanent teeth to erupt in an abnormal position and can block the permanent teeth all together.

Sometimes it is necessary to remove the baby teeth when they stay in too long. The dentist will determine if and when the over retained baby teeth, or even some of the permanent teeth, must be removed for a better bite.

Hygiene & Behavior

Home care and dietary concerns must be addressed early on. Appropriate oral hygiene practices can be taught to parents and other caretakers. Special toothbrushes, toothpastes and devices to help in the brushing process are available. Prevention of early childhood cavities, usually due to nighttime infant feeding practices, must be addressed as soon as possible. Monitoring for habitual retention of food in the cheeks, a common practice of children with CdLS, must be met with oral cleansing after each feeding. This can prevent the molars from decaying rapidly.



Self-inflicted oral and facial trauma may be noticed and addressed with various dental appliances and referrals to behavioral therapists. Routine six-month check-ups can assist in monitoring changes, detecting pathology and reinforcing proper home care.

Visiting the Dentist

An orthodontic consultation around the age of six years old is recommended. It can then be determined if the child is a good candidate for orthodontic care and when it should be started.

Compromises are often the answer. Obtaining a moderate improvement may be all that's needed or accomplished. When working with the pediatric dentist or orthodontist, we always need to be practical. We want to help improve the health of the child but, as always, we must weigh the risks and the benefits and come to a wise decision as to how to safely proceed.

The medical and developmental complications associated with CdLS make the delivery of dental therapy a challenge. When dental treatment, such as fillings, extractions and even some preventive procedures like dental sealants, are required, the best approach to accomplishing these tasks must be determined. Some children with CdLS receive their care in the dental office alongside their typical siblings with ease. For many others, this is not possible.

Conscious sedation, deep sedation and even general anesthesia are adjuncts to care that are often necessary in order to obtain cooperation and can take place in the dental office, surgery center or hospital environments. Airway abnormalities and the general health of the child must be taken into consideration to determine the safest approach to care.

The pediatric dentist can determine the best avenue to pursue in order to deliver the highest quality of care in a safe and effective manner. It's helpful to alert all of the child's healthcare providers any time the child will be placed under general anesthesia. Other physicians and dentists might desire to take advantage of the sedation and join together to perform multiple treatments, thus minimizing anesthetic experiences.

In Summary

The oral and dental concerns associated with CdLS necessitate early evaluation and intervention. A "dental home" must be established as early as possible so the child can consistently receive oral health care and prevention that is culturally sensitive, comprehensive and easily accessible.

