

Kabuki Syndrome and Cleft Palate

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What is a “Cleft Palate”?

The term “cleft” refers to a condition where the two sides of structure did not fuse or join together, and the word “palate” means the roof of the mouth. Thus “cleft palate” means a condition where there is an opening in the roof of the mouth (Fig.1). Cleft palate is a congenital defect, or birth defect, and it is often associated with cleft lip, which means a splitting in the lip.

Kabuki syndrome and cleft palate

In general, cleft lips or palates are reported to occur in about 500-700 births worldwide. It is reported that children with Kabuki syndrome have cleft lip/palate at a higher incidence (33-50%).

The chief symptoms of cleft palate are as follows:

Feeding problem: Babies with cleft palate are not able to suck and swallow normally because the opening in the roof of the mouth directly connects the mouth to the nasal cavity, resulting in milk and air escaping from the nose.

Speech and language problems: Children with cleft palates may develop their speech later and have difficulty in pronouncing several kinds of sounds such as “p,” “t,” and “k” because they cannot raise air pressure in the mouth due to the air leakage through the nose.

Dental problems: Teeth may not erupt normally; some teeth might be absent, malformed, or malpositioned.

Ear infections and hearing difficulties: The function of the auditory tube that connects the middle ear and the throat is often impaired and therefore ear infections can occur easier.

Cleft palate is a treatable condition by multidisciplinary approach. The “Cleft team” will take care of your kids and can help improve not only the function but also the appearance of the child.

Submucous Cleft Plate

Cleft palate is usually diagnosed shortly after birth because it is easy to find the cleft if you look into the baby's mouth. However, there is a special type of cleft palate called submucous cleft palate (SMCP). The term “submucous” means that the cleft is covered by the thin layer of mucosa at the center of the roof of the mouth, although the underlying muscles do not join together. Since



Figure 1: Cleft Palate

there is no apparent opening in the roof of the mouth, SMCP is sometimes difficult to find in infancy (Fig. 2) and might remain undiagnosed until they become older. One of our findings is that SMCP is observed at a much higher rate than has previously been reported. We treated six patients with cleft palate associated with Kabuki Syndrome at Shizuoka Children's Hospital. Three of them had an overt cleft palate and the other three had a submucous cleft palate.

The most important presenting symptom indicating that a child is suspected of having SMCP is abnormal and nasal speech. Another symptom of SMCP is a

uvula bifida, which means a splitting “uvula,” a small, soft piece of flesh that hangs down at the back of your mouth. If your child has these symptoms, we recommend you consult with a cleft palate specialist.

Treatment

Many medical professionals in different fields are involved in the treatment for your children because the skills of many different areas are necessary to solve the problems caused by cleft palate. A Cleft team, which usually includes a plastic surgeon, a dental surgeon, an ear-nose-throat (ENT) surgeon, a pediatrician, a speech-language pathologist, and a nurse, will take care of your child. Treatments include mainly surgery, speech therapy, and dental therapy.

Surgery

Surgery for cleft palate repair is usually performed between 10 and 18 months after birth. The surgery, which is called “palatoplasty,” consists of reconstruction of the splitting palate, including not only the mucosa but also the underlying muscle, which is most important for the speech and swallowing. There are several methods of palatoplasty. One of the most common procedures, “push-back” palatoplasty, is shown in Figure 3. In this

procedure, incisions are made on both sides of the palate. Then the palatal tissues, including mucosa and muscle, are moved from each side to the center back, and then sutured. With this procedure, the separated muscles are joined together and the palate can be reconstructed and elongated.

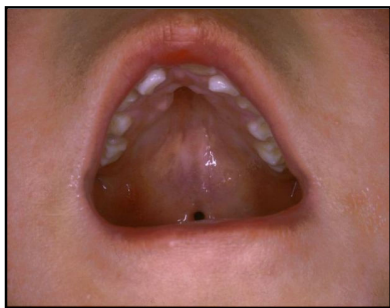


Figure 2: Submucous Cleft Palate

Speech therapy

After palatoplasty, children with cleft palate usually have speech therapy to learn how to use the reconstructed palate properly and acquire the correct pronunciation of sounds and words. The speech-language pathologist will evaluate your child's speech production and language development. The goal of speech therapy is to help them acquire correct sound and good speech habits.

Dental Care and orthodontic treatment

Children with a cleft palate often need dental and orthodontic treatment. Since the growth of the upper jaw is slower and less than the lower jaw, a child's upper teeth may not fit together properly with the lower teeth. In such cases, the orthodontist will help correct the alignment of the teeth and the relationship of the upper jaw to the lower jaw. If the tooth

alignments cannot be made normal by orthodontics alone, they may need orthognathic surgery, which is called an osteotomy, to reposition the upper jaw both forward and down.

Ear treatment

Children with a cleft palate are susceptible to ear infections, so it is important to have a regular examination by an ENT doctor for your child's ears. Since Children with severe ear infections are not able to hear language normally due to fluid collection in the middle ear, there is a risk for language delays and speech problems. To obtain proper drainage of the fluid in the middle ear, a small plastic tube is often inserted into the eardrum by an ENT surgeon.

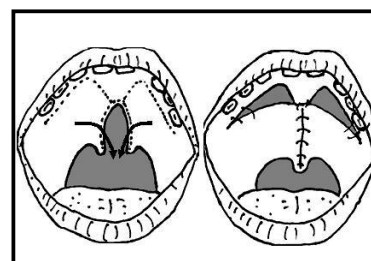


Figure 3: "Push-back" Palatoplasty

Figures:

1 Appearance of cleft palate

2 Appearance of a submucous cleft palate (cited from *The Cleft Palate-Craniofacial Journal*, Allen Press Publishing Services. 2006, Iida T et. al. Cleft palate in Kabuki syndrome: a report of six cases)

3 Schematic illustration of "push-back" palatoplasty

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