

## EVALUATION OF ASSOCIATION OF CLEFT LIP AND CLEFT PALATE WITH CERVICAL VERTEBRAL ANOMALIES: A LATERAL CEPHALOGRAPH STUDY

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### Abstract

The precise origin of cervical vertebral anomalies is still unstated, but it has been suggested that the association between abnormal development of cervical vertebrae and the maxilla and the mandible might be caused by a developmental fault of the mesenchyme as these structures might be dependent on the similar para-axial mesoderm. Hence it is appropriate to focus on this area of research and to consider the craniofacial morphology as an important diagnostic tool in Orthodontic treatment planning. The present study was planned to evaluate the association of Cleft lip and Cleft palate with cervical vertebral anomalies.

The present study was planned in Department of Pediatric and Preventive Dentistry, Buddha Institute of Dental Sciences & Hospital, Patna, Bihar, India. Thirty cases of Cleft Lip and Cleft Palate (CLCP) patients were enrolled in the present study. The age of the patients ranged from 5 years to 15 years. The cleft sample was subdivided into patients with CP only, and unilateral cleft lip and palate (UCLP) and bilateral cleft lip and palate (BCLP). Radiographs were examined on a film viewer by a single examiner. The profile of first four cervical vertebrae and Atlanto Occipital Articulation were structurally traced on an acetate paper with 3H lead pencil under optimum illumination and Cervical Vertebral Anomalies (CVA) were registered and categorized into Posterior Arch Deficiencies - PAD (dehiscence and spina bifid) and fusion.

The present study concludes the association between cleft lip and palate and Cervical Vertebral Anomalies indicating that CVA may be implicated as the etiology of cleft lip and palate. The present study showed a specific relation between the Cleft Palate and cervical anomalies and the vertebral anomalies following a specific pattern in different types of cleft was found to be PAD which occurred more frequently in UCLP and CP only and fusion occurring significantly more often in BCLP.

**Keywords:** Cleft Lip and Cleft Palate; Cervical Vertebral Anomalies; Dehiscence; Fusion; Lateral Cephalograph, etc.

### INTRODUCTION

Cleft Lip and Cleft Palate are common congenital malformations, also known as oral clefts or orofacial clefts. Birth defects are health conditions that are present at birth and they change the shape or function of one or more body parts. During the development of lip and palate in early pregnancy period, incomplete fusion of upper lip and palate leads to the formation of cleft lip and palate in a child. A child can have a cleft lip, a cleft palate or both because development of lips and palate occur at

different times during gestation. Children with these birth defects have difficulty in feeding and talking. They are also prone to ear infections, hearing loss, and dental problems. Cleft lip and cleft palate are common birth defects and happen very early in the pregnancy. Asian population have highest frequency, often 1 in 500 or higher. Cleft lip with or without cleft palate is more common in males than in females. Isolated cleft palate is more common in females whereas males have more severe defects. Cleft lip is an opening in the upper lip due to incomplete formation of the upper lip before birth. It may be

unilateral or bilateral (on both sides of lip). Cleft palate is an opening of the hard palate or soft palate (roof of the mouth) due to its incomplete fusion. It may also be unilateral or bilateral. [1]

Cleft lip and cleft palate, also known as orofacial cleft, is a group of conditions that includes cleft lip (CL), cleft palate (CP), and both together (CLP). A cleft lip contains an opening in the upper lip that may extend into the nose. The opening may be on one side, both sides, or in the middle. A cleft palate occurs when the roof of the mouth contains an opening into the nose. These disorders can result in feeding problems, speech problems, hearing problems, and frequent ear infections. Less than half the time the condition is associated with other disorders. [2]

Cleft lip and palate are the result of tissues of the face not joining properly during development. As such, they are a type of birth defect. The cause is unknown in most cases. [1] Risk factors include smoking during pregnancy, diabetes, obesity, late pregnancy, and certain medications (such as some used to treat seizures). Cleft lip and cleft palate can often be diagnosed during pregnancy with an ultrasound examination. A cleft lip or palate can be successfully treated with surgery. This is often done in the first few months of life for cleft lip and before eighteen months for cleft palate. Speech therapy and dental care may also be needed. [1] With appropriate treatment, outcomes are good. Cleft lip and palate occurs in about 1 to 2 per 1000 births in the developed world. CL is about twice as common in males as females, while CP without CL is more common in females. [3] In 2017, it resulted in about 3,800 deaths globally, down from 14,600 deaths in 1990. [4-5] The condition was formerly known as a "hare-lip" because of its resemblance to a hare or rabbit, but that term is now generally considered to be offensive. [6]

If the cleft does not affect the palate structure of the mouth, it is referred to as cleft lip. Cleft lip is formed in the top of the lip as either a small gap or an indentation in the lip (partial or incomplete cleft), or it continues into the nose (complete cleft). Lip cleft can occur as a one-sided (unilateral) or two-sided (bilateral) condition. It is due to the failure of fusion of the maxillary prominence and medial nasal processes (formation of the primary palate). A mild form of a cleft lip is a microform cleft. A microform cleft can appear as small as a little dent in the red part of the lip or look like a scar from the lip up to the

nostril. In some cases muscle tissue in the lip underneath the scar is affected and might require reconstructive surgery. It is advised to have new born infants with a microform cleft checked with a craniofacial team as soon as possible to determine the severity of the cleft.[7]

Cleft palate is a condition in which the two plates of the Maxilla that form the hard palate (roof of the mouth) are not completely joined. The soft palate is in these cases can have cleft as well. In most cases, cleft lip is also present. Palatal cleft can occur as complete (soft and hard palate, possibly including a gap in the jaw) or incomplete (a 'hole' in the roof of the mouth, usually as a cleft soft palate). When cleft palate occurs, the uvula is usually split. It occurs due to the failure of fusion of the lateral palatine processes, the nasal septum, or the median palatine processes (formation of the secondary palate). The hole in the roof of the mouth caused by a cleft connects the mouth directly to the inside of the nose.

A result of an open connection between the mouth and inside the nose is called velopharyngeal inadequacy (VPI). Because of the gap, air leaks into the nasal cavity resulting in a hypernasal voice resonance and nasal emissions while talking. Secondary effects of VPI include speech articulation errors (e.g., distortions, substitutions, and omissions) and compensatory misarticulations and mispronunciations (e.g., glottal stops and posterior nasal fricatives). Possible treatment options include speech therapy, prosthetics, augmentation of the posterior pharyngeal wall, lengthening of the palate, and surgical procedures.[8]

Submucous cleft palate (SMCP) can also occur, which is a cleft of the soft palate with a split uvula, a furrow along the midline of the soft palate, and a notch in the back margin of the hard palate. The diagnosis of SMCP often occurs late in children as a result of the nature of the cleft. While the muscles of the soft palate are not joined, the mucosal membranes covering the roof of the mouth appear relatively normal and intact. Cleft may cause problems with feeding, ear disease, speech, socialization, and cognition. [9]

Due to lack of suction, an infant with a cleft may have trouble feeding. An infant with a cleft palate will have greater success feeding in a more upright position. Gravity will help prevent milk from coming through the baby's nose if he/she has cleft palate. Gravity feeding can be accomplished by using specialized

equipment, such as the Haberman Feeder. Another equipment commonly used for gravity feeding is a customized bottle with a combination of nipples and bottle inserts. A large hole, crosscut, or slit in the nipple, a protruding nipple and rhythmically squeezing the bottle insert can result in controllable flow to the infant without the stigma caused by specialized equipment. Individuals with cleft also face many middle ear infections which may eventually lead to hearing loss. The Eustachian tubes and external ear canals may be angled or tortuous, leading to food or other contamination of a part of the body that is normally self-cleaning. Hearing is related to learning to speak. Babies with palatal clefts may have compromised hearing and therefore, if the baby cannot hear, it cannot try to mimic the sounds of speech. Thus, even before expressive language acquisition, the baby with the cleft palate is at risk for receptive language acquisition. Because the lips and palate are both used in pronunciation, individuals with cleft usually need the aid of a speech therapist. Tentative evidence has found that those with clefts perform less well at language.[10]

Within the first 2–3 months after birth, surgery is performed to close the cleft lip. While surgery to repair a cleft lip can be performed soon after birth, often the preferred age is at approximately 10 weeks of age, following the "rule of 10s" coined by surgeons Wilhelmmesen and Musgrave in 1969 (the child is at least 10 weeks of age; weighs at least 10 pounds, and has at least 10g hemoglobin).[11-12] If the cleft is bilateral and extensive, two surgeries may be required to close the cleft, one side first, and the second side a few weeks later. The most common procedure to repair a cleft lip is the Millard procedure pioneered by Ralph Millard. Millard performed the first procedure at a Mobile Army Surgical Hospital (MASH) unit in Korea.[13] Often an incomplete cleft lip requires the same surgery as complete cleft. This is done for two reasons. Firstly the group of muscles required to purse the lips run through the upper lip. In order to restore the complete group a full incision must be made. Secondly, to create a less obvious scar the surgeon tries to line up the scar with the natural lines in the upper lip (such as the edges of the philtrum) and tuck away stitches as far up the nose as possible. Incomplete cleft gives the surgeon more tissue to work with, creating a more supple and natural-looking upper lip.

Often a cleft palate is temporarily covered by a palatal obturator (a prosthetic device made to fit the

roof of the mouth covering the gap). Cleft palate can also be corrected by surgery, usually performed between 6 and 12 months. Approximately 20–25% only require one palatal surgery to achieve a competent velopharyngeal valve capable of producing normal, non-hypernasal speech. However, combinations of surgical methods and repeated surgeries are often necessary as the child grows. One of the new innovations of cleft lip and cleft palate repair is the Latham appliance.[14] The Latham is surgically inserted by use of pins during the child's 4th or 5th month. After it is in place, the doctor, or parents, turn a screw daily to bring the cleft together to assist with future lip or palate repair. If the cleft extends into the maxillary alveolar ridge, the gap is usually corrected by filling the gap with bone tissue. The bone tissue can be acquired from the patients own chin, rib or hip.

The precise origin of cervical vertebral anomalies is still unstated, but it has been suggested that the association between abnormal development of cervical vertebrae and the maxilla and the mandible might be caused by a developmental fault of the mesenchyme as these structures might be dependent on the similar para-axial mesoderms. [15-16] Hence, it is appropriate to focus on this area of research and to consider the craniofacial morphology as an important diagnostic tool in orthodontic treatment planning. So the present study was planned to evaluate the association of cleft lip and palate with cervical vertebral anomalies.

#### **Methodology:**

The present study was planned in Department of Pediatric and Preventive Dentistry, Buddha Institute of Dental Sciences & Hospital, Patna, Bihar. The 30 cases of Cleft Lip and Cleft Palate (CLCP) patients were enrolled in the present study. The age of the patients ranges from 5 years to 15 years. The cleft sample was subdivided into patients with CP only, and unilateral cleft lip and palate (UCLP) and bilateral cleft lip and palate (BCLP). Radiographs were examined on a film viewer by a single examiner. The profile of first four cervical vertebrae and Atlanto Occipital Articulation were structurally traced on an acetate paper with 3H pencil under optimal illumination and cervical vertebral anomalies (CVA) were registered and categorized into posterior arch deficiencies (dehiscence and spina bifid) and fusion. [16]

The abnormalities of the cervical vertebrae were defined by visual assessment directly on the lateral cephalograms and structural tracings. They were classified according to the method of Sandham [17] and divided into two categories: PAD and fusion anomalies.

All the patients were informed consents. The aim and the objective of the present study were conveyed to them. Approval of the institutional ethical committee was taken prior to conduct of this study.

Following was the inclusion and exclusion criteria for the present study.

**Inclusion criteria:** • Lateral cephalographs of cleft lip and cleft palate patients. • Lateral cephalometric radiographs extending up to the 4<sup>th</sup> cervical vertebrae and covering the entire anatomy of a cervical spine.

**Exclusion criteria:** • Radiographs of syndromic patients • Deteriorated radiographs

**Results & Discussion:**

Cleft lip and palate are common congenital malformations of the lip, palate, or both caused by complex genetic and environmental factors.[18] Genetic susceptibility has long been identified as a major component of CLP.[19] The most common environmental risk factors are maternal exposure to tobacco products, alcohols, nutritional deficiencies, some viral infections (rubella), medications, and teratogens in early pregnancy. Recognized teratogens included rare exposures such as phenytoin, valproic acid, thalidomide, and herbicides such as dioxin.[20] Other proposed risk factors include various occupational and chemical exposures, hyperthermia, stress, maternal obesity, oral hormone supplementation, ionizing radiation. Folic acid and zinc are also important in fetal development, and deficiency of these nutrients causes isolated cleft palate and other malformations. [19]

The cervical vertebral anomalies are commonly divided into posterior arch deficiencies (PAD) and fusions (FUS). Posterior arch deficiencies are subdivided into spina bifida, which implies incomplete ossification in the spinous process and generally occurs in the posterior arch of the vertebral unit, and dehiscence, which implies incomplete development of the structures. Dehiscence in the atlas affects either the anterior arch or the posterior arch, posterior arch dehiscence being most common in the midline. Fusion is a bony union of one unit with

another at the articulation facets, neural arch, or transverse processes and may be subdivided into fusion between two cervical vertebrae; block fusion in which the bony union includes the vertebral bodies; and occipitalization, the assimilation of the atlas to the base of the skull or atlanto-occipital fusion or some degree of bony union between the skull and the atlas. [21]

The prevalence of cervical vertebral anomalies in CL and UCLP was found to be higher in the present study as compared to other studies in literature.

**Table 1: Evaluation of CLPL and Cervical Vertebral Anomalies**

Parameters	No. of Cases
Sex	
Males	18
Females	12
Total	30
Types	
UCLP	21
BCLP	9
Total	30
Prevalence	
Cervical Vertebral Anomaly (CVA)	6
Cleft Lip and Cleft Palate (CLCP)	24
Types of CVA	
Posterior Arch Deficiencies (PAD)	2
Fusion	4

Sonnesen et al. [22] had reported that the deviations in head posture and cranial base angle were sexually dimorphic, showing larger cervicohorizontal and cranial base angles in females than males. They also observed a positive correlation of cervical lordosis, inclination of upper cervical spine and cranial base angle in females with fusions of the cervical column, whereas this correlation was not found in males. Hence, it could be hypothesized that fusion anomalies show dimorphic pattern in their occurrence. However, Sonnesen and Kjaer [23] and Arntsen and Sonnesen [24] proposed that there was no significant gender difference in the occurrence of cervical vertebral anomalies.

Fusion of the upper cervical vertebrae occurs most commonly between the second and the third (C2-C3) vertebrae. They are probably the result of a failure in normal embryological segmentation due to locally decreased blood supply during fetal development.

Patients are generally asymptomatic, but increasing age or injury may precipitate symptoms. [25] The cervical spine anomalies often coexist with syndromes anomalies of the head and neck and emphasized that those anomalies should be detected by radiographic imaging as early as possible to optimize management and reduce the risk of neurologic injury. [26]

One of those syndromes is Klippel-Feil syndrome. [27] It is characterized by congenital fusion of at least two of seven cervical vertebrae in the cervical spine, with limitations to movement of the head or neck, a short neck, and a low posterior hairline in patients with CL and palate.

Block fusion of cervical vertebrae is a general feature of the Klippel-Feil anomaly. Farman and Escobar[28], in their study showed, 74 cases of fusion of the cervical vertebrae, 8 of which were block fusions. All of these were observed in CPO group. McRae and Barnum [29] reported on 25 cases with occipitalization of the atlas. They noticed that in two of these patients, there were other fusions besides the occipitalization.

#### Conclusion:

The present study concludes the association between cleft lip and palate and cervical vertebral anomalies indicates that cervical vertebral anomalies may be implicated as the etiology of cleft lip and palate. Study showed a specific relation between the CP and cervical anomalies. The vertebral anomalies followed a specific pattern in different types of cleft and was found to be PAD occurred more frequently in UCLP and CP only and fusion occurring significantly more often in BCLP.

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