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# CLEFT PALATE LATERAL SYNECHIAE SYNDROME - A RARE CASE REPORT.

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**ABSTRACT** Oral synechiae is a rare craniofacial anomaly . This case report with findings of 15 days old Preterm male baby with birth weight of 1.6 kg , was admitted in our institution with the findings Of difficulty in opening mouth and feeding difficulty with bulging anterior fontanellae. Baby was maintaining saturation in room air. Careful examination of oral cavity reveals that baby is having Micrognathia , multiple bands connecting between alveolus and hard palate on both sides with incomplete cleft palate . Baby was evaluated thoroughly with routine blood investigation with MRI brain imaging. Diagnosed as a case of Cleft palate with lateral synechiae with tetraventicular hydrocephalus and synechiae was released.

**KEYWORDS :** synechiae , hydrocephlalus , cleft palate , Neonate

**Case History :**

15 Days old preterm , male baby , delivered via naturalis with birth weight of 1.6 kg.

Immediately after delivery child had mild respiratory distress . Baby was admitted in NICU for prenatal care and started of Nasogastric tube feeding . During that time baby Oral cavity examination shows multiple bands and incomplete mouth opening.

**Figure 1. Micrognathia**

**Figure 2. Lateral synechiae**

Baby was transferred to our institution. History wise Antenatal Ultrasound findings were normal. On examination child general condition , cry and activity was good. Anterior Fontanellae bulging with oral cavity findings of , Micrognathia ,multiple bands connecting alveolus and Hard palate on bothsides with incomplete cleft palate ( Fig 1 & 2).

Other systemic examination normal. Investigated with routine blood and MRI Brain imaging . MRI Shows tetra ventricular Hydrocephalus , cerebral atrophy & Dural venous thrombus (Fig.3). Head circumference monitoring Values are static . For hydrocephalus neurosurgeon opinion obtained and managed conservatively .

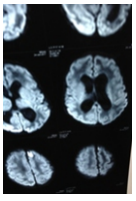

Figure 3.Hydrocephalus

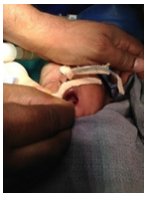

Figure 4. Synechiae release

Figure 5. Post synechiae release

Coagulation profile shows mild prolongation , which was corrected preoperatively. Cardiac Screening ECHO shows normal study. With this above finding baby was diagnosed as a case of Oral synechiae type I variety with hydrocephalus and planned for surgery .

Under general anaesthesia with Endotracheal tube - oral synechiae was released .

Post –op period was uneventful ( Fig 4 & 5).

**Discussion :**

Oral synechiae have been described in the literature as early as 1887 by Illera. This congenital Anomaly is very rarely encountered condition. The exact etiology behind oral synechiae is remain unclear , several theories have been proposed. Persistent of buccopharyngeal membrane is one of theory proposed by Longacre. Mathis theory says that due to secondary effect of epithelial rudiments that become adhered during the fusion of palatal shelf. Formation of subglasso palatal membrane that forms before the fusion process suggested by Gartlan.

The most common clinical presentation are neonatal respiratory distress and feeding difficulty. These are the major causes of morbitity in these patients and usually manifest in early period of life.

**Oral synechiae classified into five different types :**

1) Synechiae by an adhesion of the alveolar mucosa on one (or) both sides of the Upper and lower jaw.
2) Synechiae by an adhesion on the hard palate and floor of the mouth excluding the rear of tongue.
3 ) Synechiae in which hard palate and tongue are partially involved.
4) Synechiae in which soft palate and tongue are widely involved and interrupting Continuity between oral cavity and oropharynx.
5) Synechiae by adhesion between hard palate and lower lip.

Medial oral synechiae is the rarest variety.

Oral synechiae infrequently occur as an isolated finding . Usually they are associated With other congenital anomalies / syndrome . Cleft palate lateral synechiae syndrome

(CPLS) comprises of cleft palate , micrognathia and lateral intra oral synechiae.

Other syndromes associated with cleft palate and oral synechiae include congenital alveolar Synechiae syndrome , popliteal pterygium syndrome and Fryn's syndrome. In addition to that other congenital defects associated with oral synechiae are cardiac anomalies ,Hydrocephalus , Hypospadias and Bifid scrotum.

Our patient exhibited a case of oral synechiae type 1 variety with hydrocephalus and Feeding difficulties . No Respiratory distress child was started of oral feeding ( paaladai ) On 2nd post operative day . Baby was tolerating feeds and mouth opening was adequate .

needs regular follow up for hydrocephalus and cleft palate management.

**Conclusion :**

Oral synechiae is a rare craniofacial anomaly that usually occurs with other congenital Conditions. Early diagnosis and treatment is crucial for this condition . Feeding difficulty and respiratory distress are the major cause of morbidity. Management of airway is most important . Direct laryngoscope with oral intubation was successful in our case . Even after synechiae release ,these babies require regular follow- up for management of other associated anomalies.

**References :**

1) M.A. Mascarella, j.schwartz,j .Manoukian , Congenital intra- oral adhesions and surgical approach to cleft palate lateral synechiae syndrome . Int .j .paediatric otorhinolaryngol. 79(2015) 769 – 772.
2) A.R.Buche , V.R.Borade , N.Suman . Congenital oral synechiae : a case report and review. Int . j. Recent trends sci. Technol 10 ( 1) (2014) 184 – 186.
3 ) F. Shahin , B . Gholamreza . Cleft palate lateral synechiae syndrome - inheridtary or not ? Egypt . j. Ear Nose Throat Allied sci 15 (2) ( 2014) 173 – 175 .
4) S. K. Donepudi . R.M. Stocks , E.K. Pivnick Cleft palate lateral synechiae syndrome : an Opportunity for unique surgical closure. Int. j. Paediatr. Otorhinolaryngol. 73(6) (2009) 861-866.