Pediatric Oncall Journal (April-June 2019)
Volume 16, Issue 2: 57
DOI: https://doi.org/10.7199/ped.oncall.2019.28

PEDIATRIC ONCALL
CHILD HEALTH CARE

LETTER TO EDITOR (VIEWERS CHOICE)

CLEFT PALATE WITH LATERAL SYNECHIAE SYNDROME

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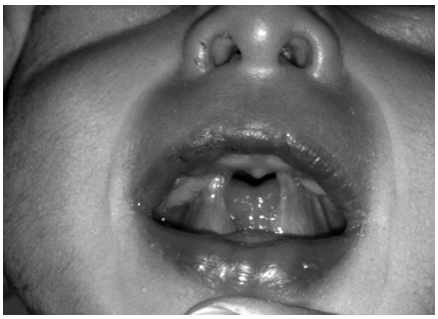
KEYWORDS
CPLS, Fryns Syndrome

ARTICLE HISTORY
Received 17 December 2018
Accepted 21 December 2018

A full term male neonate was born by normal vaginal delivery to parents of third degree consanguineous marriage. He had difficulty in opening of mouth right from birth. Examination of oral cavity showed a membranous band extending from floor of mouth to roof of mouth on both sides with a very minimal central aperture which made it difficult for the visualisation of internal structure of oral cavity (Figure 1). No other external congenital anomalies were noted. Systemic examination was normal. Paediatric surgical opinion was obtained and was taken for band excision. During surgery, after excision of the band, a central cleft palate and hypoplasia of tongue were noted. Child was started initially with nasogastric feeds followed by breastfeeds and discharged in a stable condition. Cleft palate surgery was planned at a later date.

Figure 1. Membranous band extending from floor of mouth to roof of mouth on both sides with a very minimal central aperture



Cleft palate with lateral synechiae syndrome (CPLS) is a rare autosomal dominant syndrome characterised by cleft palate associated with cord or membrane like adhesion between free borders of palate to floor of the mouth. In 1972, Fuhrmann et al described this rare syndrome.[1] Since then very few cases have been reported.[2,3] It is considered to be milder end of phenotypic expression of Fryns Syndrome spectrum.[2] The occurrence is influenced by environmental factors like temperature, viral infection and stochastic events. Genetic studies have failed to reveal any significant chromosomal defects. The etiology of synechiae has been debated with many theories. But none of the theories were conclusive. CPLS has been associated with popliteal pterygium syndrome, Van der Woude Syndrome (lower lip pits, cleft lip with or without cleft palate, hypodontia, congenital heart disease, syndactyly, ankyloglossia and cerebral anomalies) and Fryns syndrome (alveolar synechiae cleft palate and distal digital hypoplasia).[2,3] Cleft palate with oral synechiae are the rarest anomalies and usually the synechiae are lateral. But there have also been reports of cleft palate associated with a single midline synechiae.[4] CPLS can also be associated with hypoplasia of mandible, rarely with hydrocephalus and redundant lower lip tissue.[5] The symptoms are cleft palate, mouth adhesion, inability to fully open mouth and upper airway obstruction. In the year 2009, Donepudi et al suggested a novel approach in treating these patients.[6] They described a surgical closure of cleft palate with the synechiae tissue in a single stage procedure. They described this as a rare opportunity for unique surgical correction of these defects.

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Compliance with Ethical Standards

Funding: None

Conflict of Interest: None

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