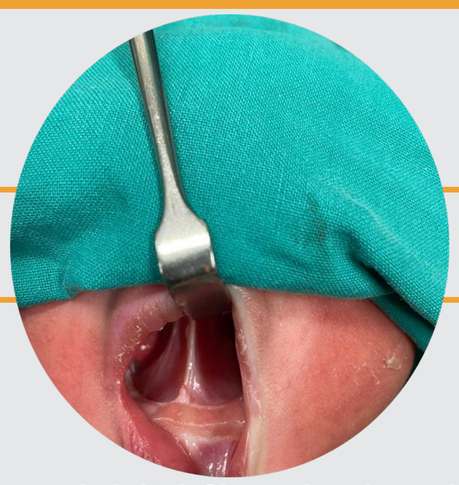
CONGENITAL SYNECHIAE OF THE ORAL CAVITY ASSOCIATED WITH CLEFT PALATE – CASE REPORT

Dr Lorraine Masotja
Limpopo (Private practice)

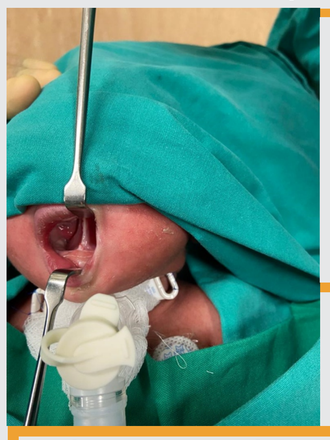
- *A case report of a new-born baby delivered by Caesarean- Section*
- *He was a second born and the first child did not have any abnormalities*
- *Pregnancy was uneventful*
- *Post delivery, he was in respiratory distress and was taken to Neonatal ICU*
- *Ventilation was immediately assisted by CPAP.*
  The baby was saturating well and stable in Neonatal ICU (NNICU)
- *On the first day of life, nurses and the Paediatrician were unable to feed him since they could not pass the bottle into the oral cavity*
- *They noticed "some tissue" obstructing the oral cavity*
- *ENT consultation was requested*

ENT CONSULTATION

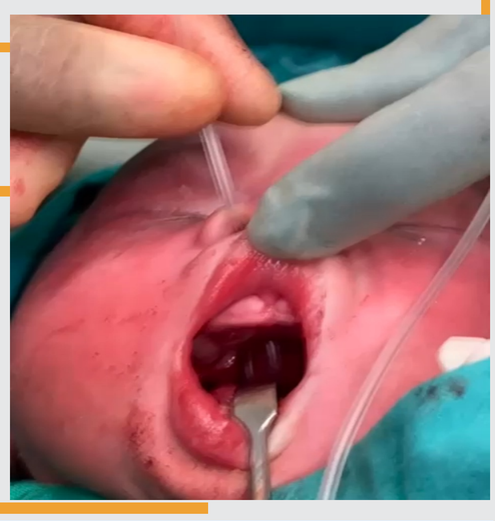


- *ENT Examination revealed thick fibrous band attached to upper alveolar ridge superiorly and to floor of the mouth inferiorly.*
- *The lateral edges were free on both sides*
- *The tongue was partially visible lateral and posterior to the thick tissue only on the right side*
- *The patient was desaturating significantly every time CPAP mask was removed*
- *Diagnosis of Oral Synechiae was made*

After discussion with the parents, and obtaining consent, the child was taken to theatre for proper examination and excision of the fibrous band. Difficult intubation was anticipated and consent for tracheostomy was obtained



- *Cardiology consultation did not reveal any cardiac abnormalities*
- *In theatre, Intubation failed and the patient was desaturating without supplementary oxygen*
- *Tracheostomy was done to secure the airway*
- *Synechiae excised*



- *After excision of the thick Synechiae, a complete Cleft Palate was found (CT Scans were not done preoperatively)*
- *Direct Laryngoscopy and Bronchoscopy were done. Not other abnormalities of the airway was found*
- *A Nasogastric feeding tube was inserted and patient was taken back to ICU in a stable condition*

CHALLENGES IN NNICU

- *Tracheostomy care – Neonatal tracheostomy tubes are small in calibre and may block easily . This may be potentially life threatening*
- *Cleft Palate and Feeding – In our institution, Cleft palates are only repaired after the age of nine months. The speech therapist was consulted to assist with feeding but this did not yield any positive results regardless of different feeding methods used*
- *Risk of Aspiration – The child was unable to swallow feeds and would frequently show signs of aspiration regardless of which feeding method was tried*
- *Parents – The patients condition affected the parents psychologically. Services of a Psychologist were used and this assisted them a lot*

DISCUSSION

- *Incidence of congenital birth defects in neonates is about 2.5%.*
- *Congenital fusion defects of the maxilla and mandible with other anatomical oral and orofacial abnormalities are usually rare.*
- *Though congenital fibrous bands commonly involve attachment between alveolar ridges of the maxilla and mandible or between the tongue and palate,*
  It can involve any part of the oral cavity.
- *Congenital fusion bands between maxilla and mandible and epithelium, connective tissue, muscle and bone all may be involved in the process.*
- *It is known as synostosis when it comprises of bones and soft tissues or synechiae when there is only involvement of soft tissues.*
- *Bony maxilla–mandibular fusion is an extremely rare anomaly.*
- *Congenital adhesion of the maxilla and mandible by fibrous bands is called Syngnathia*

CLASSIFICATION

Dawson divided the cases of syngnathia into:

- *Type I: Simple—with no other birth defects in head and neck*
- *Type II: Complex—with two subtypes*
  - *Type IIa: Syngnathia with aglossia*
  - *Type IIb: Syngnathia with agenesis or hypoplasia of the proximal mandible.*

Laster modified Dawson's classification system as:

- *Type Ia: Simple anterior syngnathia characterized by bony fusion of the alveolar ridges and without other congenital deformities in head and neck*
- *Type Ib: Complex anterior syngnathia characterized by bony fusion of the alveolar ridges only and associated with other congenital deformities in head and neck*
- *Type IIa: Simple zygomaticomandibular syngnathia characterized by bony fusion of the mandible to the zygomatic complex, causing only mandibular micrognathia*
- *Type IIb: Complex zygomaticomandibular syngnathia characterized by bony fusion of the mandible to the zygomatic complex and associated with clefts or temporomandibular joint ankyloses.*

- *The association of congenital oral synechia and cleft palate is an extremely rare syndrome*
- *Both classification systems do not classify simple anterior incomplete synechiae with cleft palate into any type.*
- *A new system of classification which takes into consideration the fusion of soft tissues and bones separately along with the presence of other congenital orofacial anomalies should be devised.*
- *A proper radiological investigation including high resolution CT scan is essential in complicated cases*

CAUSES

- *Various causes have been considered as a reason for the syndrome; however, none has been proven.*
- *Some described the cause to include persistence of the buccopharyngeal membrane.*
- *amniotic constriction bands in the region of the developing branchial arches*
- *environmental insults*
- *drugs such as large doses of vitamin A.*

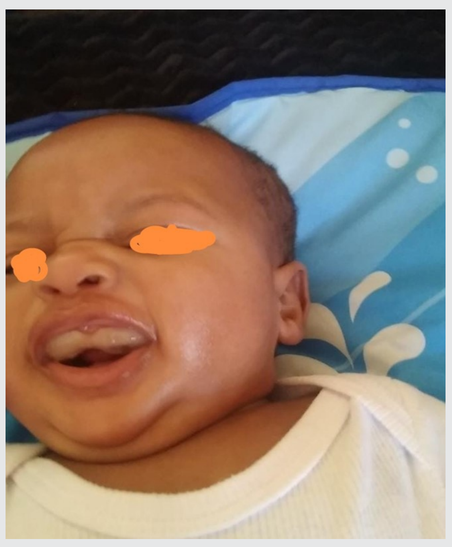
- *Congenital oral bands interfere with feeding, breathing, general health of the patient, growth and development,*
- *Poor growth of the facial skeleton*
- *Poorly aligned dental eruption*
- *Intubation for anaesthesia is challenging*
- *Aspiration pneumonitis*
- *Speech problems*
- *Early intervention is needed to secure the airway followed by management of feeding problems as delayed intervention in the case of maxillo-mandibular fusion can predispose the neonate to asphyxia, aspiration pneumonitis, malnutrition and growth retardation.*

Treatment varies as per the extent and nature of anomalies
Surgery is the mainstay of treatment

FURTHER MANAGEMENT OF THE PATIENT

- *Poor Feeding and Malnutrition - A maxillofacial surgeon assisted with an obturator and the child is bottle fed. At the time of discharge from the hospital, he was able to feed well with the obturator in situ. Speech therapist and dietician are continuing to follow up on feeding and nutritional status of the patient*
- *Possibility of OME –Cleft palate contributes to risk of Otitis media with effusion this will be followed up closely and managed accordingly as the cleft palate is repaired*
- *Cleft palate repair – The plan by the resident Plastic surgeon is to repair the cleft at the age of nine months*
- *Parent Counselling regarding future pregnancies – This was done by paediatrician and Psychologist*

The patient at four months with obturator in situ



*Consent to show images obtained from the parents

References

- *Synechiae of the Oral Cavity with Cleft Palate: A Rare Congenital Anomaly Hitendra P Singh1 , Sunil Kumar2 , Neha Rautela3 , Meraj Ahmad*
  International Journal of Head and Neck Surgery (2018): 10.5005/jp-journals-10001-13593
  Department of ENT and Head-Neck Surgery, King George's Medical University, Lucknow, Uttar Pradesh, India
- *Cleft palate with lateral oral synechiae presenting in late childhood: An extreme rarity*
  Department of Plastic Surgery, The Institute of Post-Graduate Medical Education and Research, Kolkata, West Bengal, Indi