CRANIOFACIAL DYSOSTOSIS (CROUZON SYNDROME)

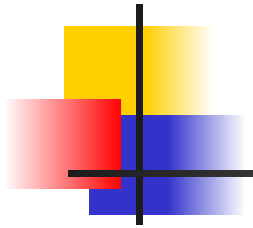
Dept.of Oral Pathology
&Microbiology

Learning Objectives

At the end of the lecture student should be able to describe

- Clinical features, oral manifestations, radiographic features,& surgical management of Marfan's Syndrome, Pierre Robin Syndrome,

MARFAN'S SYNDROME (MARFAN-ACHARD SYNDROME)



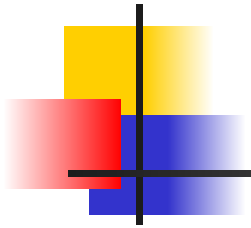
- Is a heritable disorder of connective tissue, characterized by abnormalities of the skeletal, cardiovascular, & ocular systems

Etiology:

- MS is transmitted by an autosomal dominant mode of inheritance
- Incidence : between 0.5 – 1 case in 10,000 births
- The Marfan gene is believed to produce a change in one of the proteins that provides strenght to a component of C.T. (probably collagen)

Clinical features:

- Tall, slender stature with relatively long legs & arms, large hands with long fingers, & loose joints
- The arms, legs & digits are disproportionately long compared with patients trunk
- Chest deformities include a protrusion or indentation of breast bone.

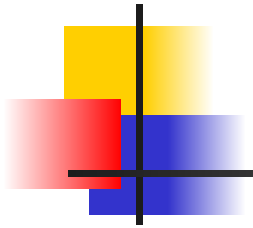


- Various degree of scoliosis is present
- Face appears long & narrow
- Oral finding: narrow, high-arched palate & dental crowding
- Cardiac : Mitral valve disease,
- Ocular : dislocation of lens (ectopia lentis); myopia

Treatment:

- Annual medical examination & treatment of cardiac & ocular defect.

PIERRE ROBIN SYNDROME (Robin anomalad)



- 3 essential components:
- Micrognathia or retrognathia
- Cleft palate
- Glossoptosis (often accompanied by airway obstruction)

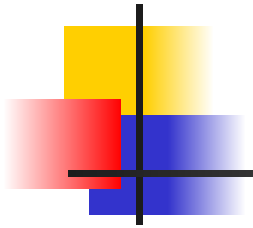
Etiology:

- Mechanical theory: most accepted
- Initially, mn hypoplasia occurs between 7^{th} & 11^{th} week of gestation. This keeps the tongue high in the oral cavity, causing cleft palate by preventing the closure of palatal shelves.

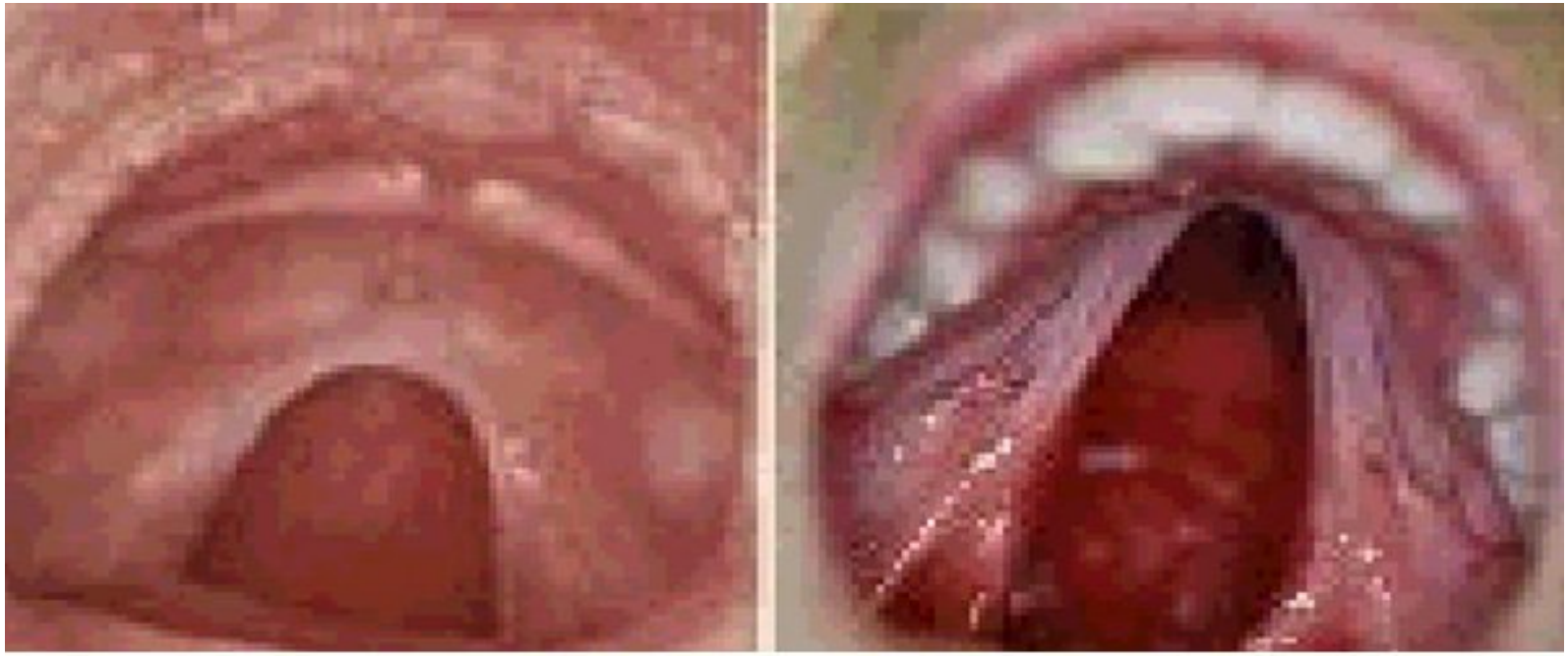
- It explains the classic inverted U- shaped cleft & the absence of an asso cleft lip.
- Neurological maturation theory
- Rhombencephalic dysneurulation theory

Clinical features:

- Prevalence: 1 per 8,500 live births.
- Equal sex predilection
- Micrognathia reported in majority cases (91.7%)
- Mn: small body, obtuse gonial angle, post located condyle. But mn hypoplasia resolves by age of 5-6 yrs

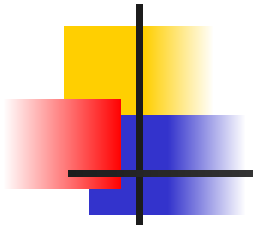


- Macroglossia & ankyloglossia are rare (10-15%)
- Resp & feeding difficulty in new born, sleep apnoea may be present
- Cleft palate (14-91%): can affect both hard & soft palate, usually (80%) U- shaped

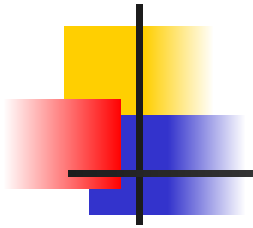


Pierre Robin malformation

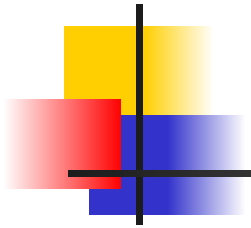
Fig 17-17. Pierre Robin malformation.
U- and V-shaped cleft palates. p. 989



- Occasionally, bifid or double uvula present
- Other asso anomalies: otitis media, hearing loss, nasal deformities, dental malformations



- Anomalies involving musculoskeletal system (70-80% cases) are most common systemic anomalies.: syndactyly, dysplastic phalanges, polydactyly, clinodactyly, hyperextensible joints, oligodactyly in upper limbs



- CNS defects: language delay, epilepsy, neurodevelopmental delay, hypotonia, hydrocephalus

Treatment & Prognosis:

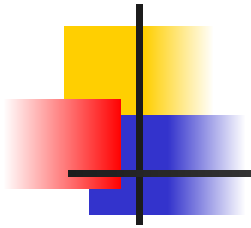
- Multidisciplinary approach

Summary

- Clinical features, oral manifestations, radiographic features,& surgical management of Marfan's Syndrome, Pierre Robin Syndrome,

BIBLIOGRAPHY

- Text book of oral pathology Shafer's, 5 & 6th edition
- Oral & Maxillofacial Pathology A Rationale for Diagnosis & Treatment. R E Marx 1st edition
- Color Atlas of Oral Diseases Cawson, R. 2nd & 5th edition
- Oral and Maxillofacial Pathology Neville, Brad W. 2nd
- Lucas's Pathology Of Tumor's of the Oral Tissues



Thank You