DEVELOPMENTAL DISTURBANCES

Dr Manasi Kale

ETIOLOGY

- Combination of genes:

A child may receive a particular combination of gene(s) from one or both parents, or there may be a change in the genes at the time of conception, which results in a craniofacial anomaly

- Environmental:

There is no data that shows a direct correlation between any specific drug or chemical exposure causing a craniofacial anomaly. However, any prenatal exposure should be evaluated.

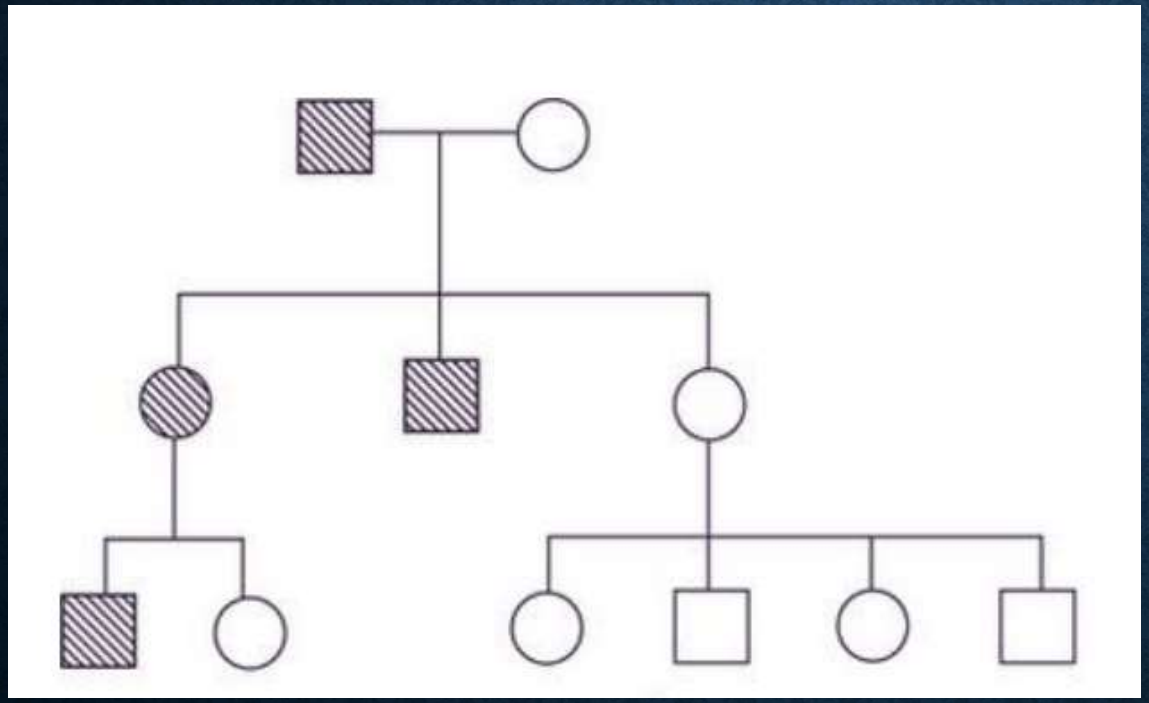
- Folic acid deficiency:

Studies have shown that women who do not take sufficient folic acid during pregnancy, or have a diet lacking in folic acid, may have a higher risk of having a baby with certain congenital anomalies, including cleft lip and/or cleft palate.

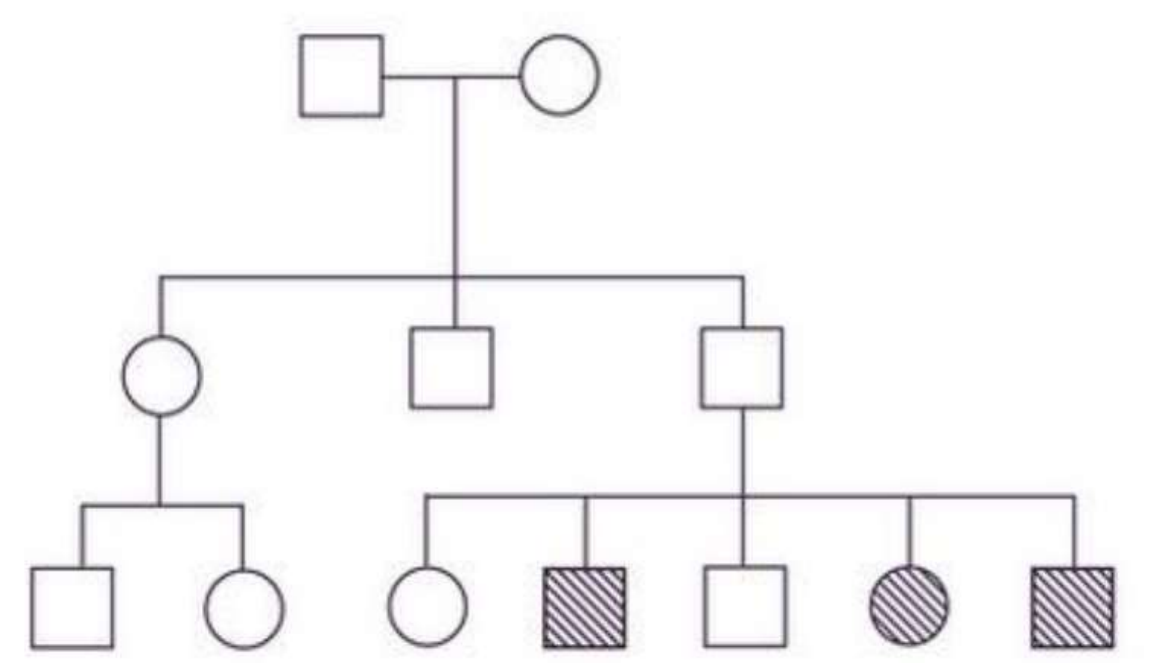
Genetics:

- Genotype is defined as the genetic constitution of an individual, and may refer to specified gene loci or to all loci in general.
- An individual's phenotype is the final product of a combination of genetic and environmental influences. Phenotype may refer to a specified character or to all the observable characteristics of the individual.

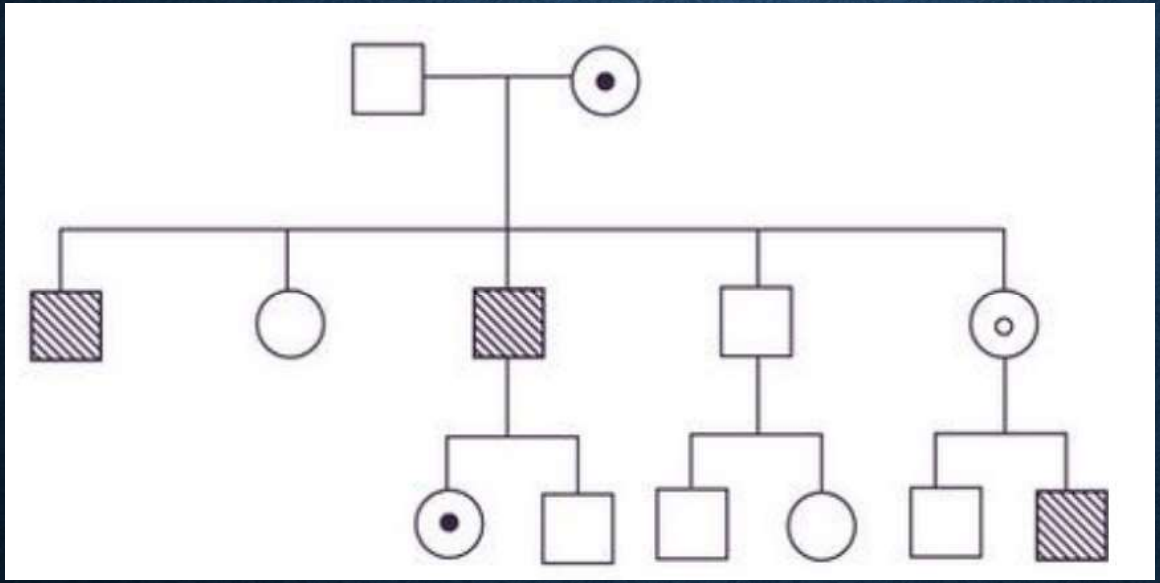
MODES OF INHERITANCE



Autosomal dominant pedigree



Autosomal recessive pedigree



X-linked recessive pedigree.

- Developmental Disturbances of Jaws .
- Abnormalities of Dental Arch Relations .
- Developmental Disturbances of Lips and Palate .
- Hereditary Intestinal Polyposis Syndrome .
- Developmental Disturbances of Oral Mucosa .
- Developmental Disturbances of Gingiva .
- Developmental Disturbances of Tongue .
- Developmental Disturbances of Oral Lymphoid Tissue .
- Developmental Disturbances of Salivary Glands .
- Fissural (Inclusion, Developmental) Cysts of Oral Region

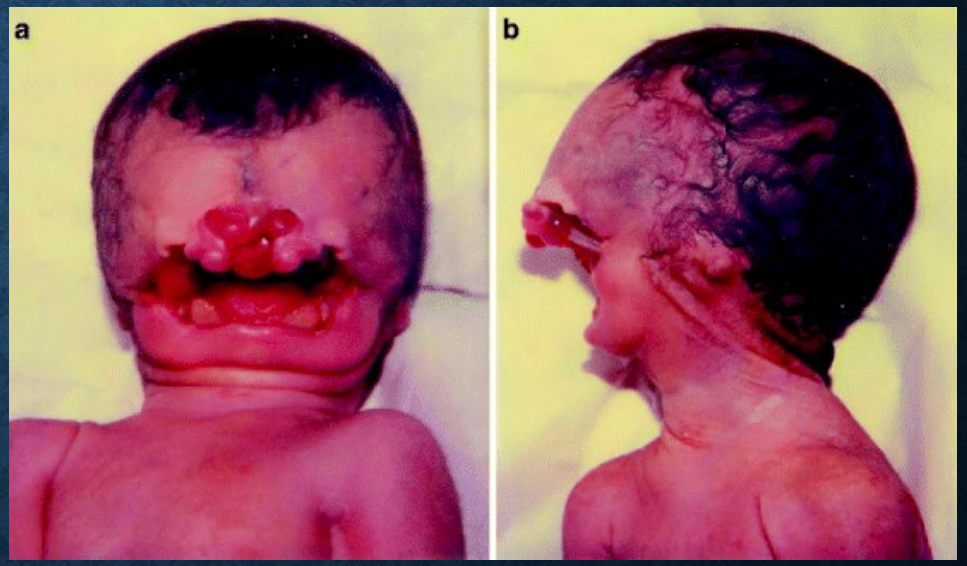
DEVELOPMENTAL DISTURBANCES OF TEETH

- Developmental Disturbances in Size of Teeth
- Developmental Disturbances in Shape of Teeth .
- Developmental Disturbances in Number of Teeth .
- Developmental Disturbances in Structure of Teeth .
- Disturbances of Growth (Eruption) of Teeth .

DEVELOPMENTAL DISTURBANCES OF JAWS

AGNATHIA (OTOCEPHALY, HOLOPROSENCEPHALY AGNATHIA)

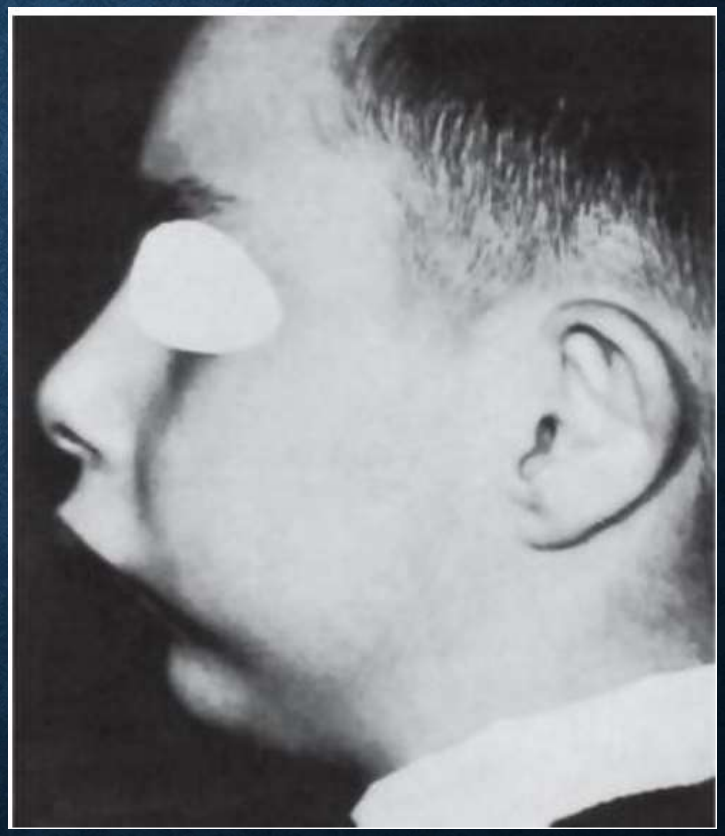
- Agnathia is a lethal anomaly characterized by hypoplasia or absence of the mandible with abnormally positioned ears.
- Autosomal recessive mode of inheritance.
- More commonly, only a portion of one jaw is missing. In the case of the maxilla, this may be one maxillary process or even the premaxilla.



- Partial absence of the mandible is even more common. The entire mandible on one side may be missing, or more frequently, only the condyle or the entire ramus, although bilateral agenesis of the condyles and of the rami also has been reported.
- It is probably due to failure of migration of neural crest mesenchyme into the maxillary prominence at the fourth to fifth week of gestation (postconception).
- The prognosis of this condition is very poor and it is considered to be lethal.

MICROGNATHIA

- Micrognathia literally means a small jaw, and either the maxilla or the mandible may be affected.
- Many cases of apparent micrognathia are not due to an abnormally small jaw in terms of absolute size, but rather to an abnormal positioning or an abnormal relation of one jaw to the other or to the skull, which produces the illusion of micrognathia.
- Bird face deformity



- True micrognathia may be classified as either congenital, or acquired.
- The congenital type is associated with other congenital abnormalities, including congenital heart disease and the Pierre Robin syndrome
- Micrognathia of the maxilla frequently occurs due to a deficiency in the premaxillary area, and patients with this deformity appear to have the middle third of the face retracted.
- Although it has been suggested that mouth-breathing is a cause of maxillary micrognathia, it is more likely that the micrognathia may be one of the predisposing factors in mouth-breathing.

- Agenesis of the condyles results in a true mandibular micrognathia.
- The acquired type of micrognathia is of postnatal origin and usually results from a disturbance in the area of the temporomandibular joint.
- Ankylosis of the joint, for example, may be caused by trauma or by infection of the mastoid, of the middle ear, or of the joint itself.
- The clinical appearance of mandibular micrognathia is characterized by severe retrusion of the chin, a steep mandibular angle, and a deficient chin button

- Causes of micrognathia

Congenital conditions for eg.

- Cerebrocostomandibular syndrome
- Pierre Robin syndrome

Intrauterine acquired conditions

- Syphilis, congenital Chromosomal abnormalities
- Trisomy 18
- Turner's syndrome

Autoimmune conditions

- Juvenile chronic arthritis

Mendelian inherited conditions

- CODAS (cerebral, ocular, dental, auricular, skeletal)
- Noonan's syndrome
- DiGeorge's syndrome

MACROGNATHIA

- Macrognathia refers to the condition of abnormally large jaws.
- An increase in size of both jaws is frequently proportional to a generalized increase in size of the entire skeleton, e.g. in pituitary gigantism.

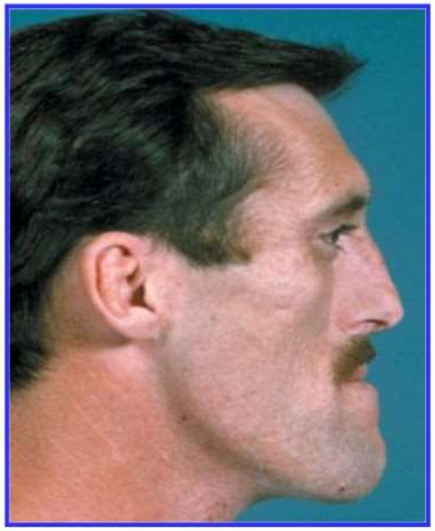
Macrognathia may be associated with certain other conditions, such as:

• Paget's disease of bone, in which overgrowth of the cranium and maxilla or occasionally the mandible occurs

• Acromegaly, in which there is progressive enlargement of the mandible owing to hyperpituitarism in the adult

• Leontiasis ossea, a form of fibrous dysplasia in which there is enlargement of the maxilla.

Clinical Appearance of Macrognathia



Macrognathia associated with Paget's disease of bone.

- In many instances the prognathism is due to a disparity in the size of the maxilla in relation to the mandible.
- In other cases the mandible is measurably larger than normal.
- The angle between the ramus and the body also appears to influence the relation of the mandible to the maxilla, as does the actual height of the ramus.
- Thus prognathic patients tend to have long rami which form a less steep angle with the body of the mandible.
- The length of the ramus, in turn, may be associated with the growth of the condyle. It may be reasoned, therefore, that excessive condylar growth predisposes to mandibular prognathism

General factors which favor mandibular prognathism are as follows:

- Increased height of the ramus
- Increased mandibular body length
- Increased gonial angle
- Anterior positioning of the glenoid fossa
- Decreased maxillary length
- Posterior positioning of the maxilla in relation to the cranium
- Prominent chin button
- Varying soft-tissue contours.

Treatment:

Surgical correction, Ostectomy.