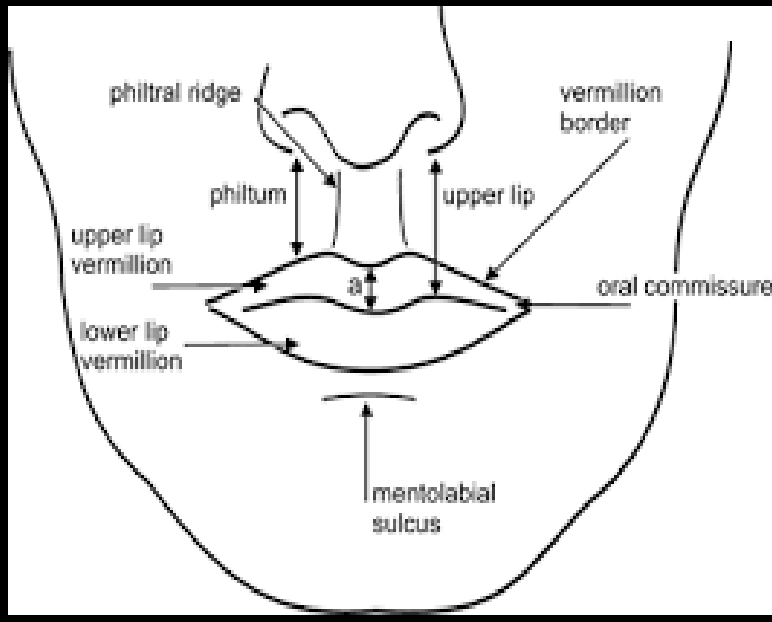
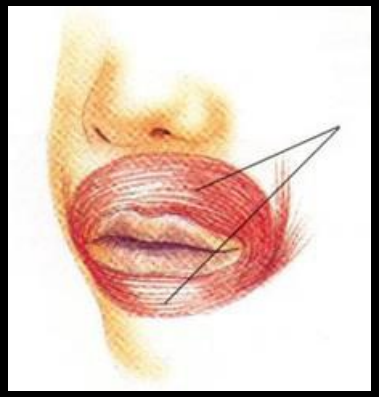
CONGENITAL ANOMALIES

CLEFT LIP
&
CLEFT
PALATE

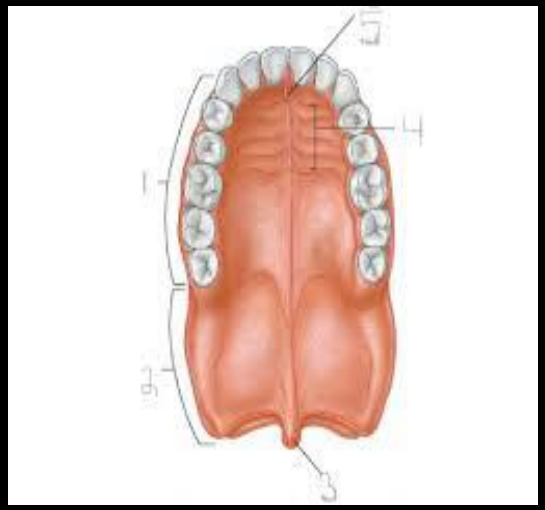
Anatomy

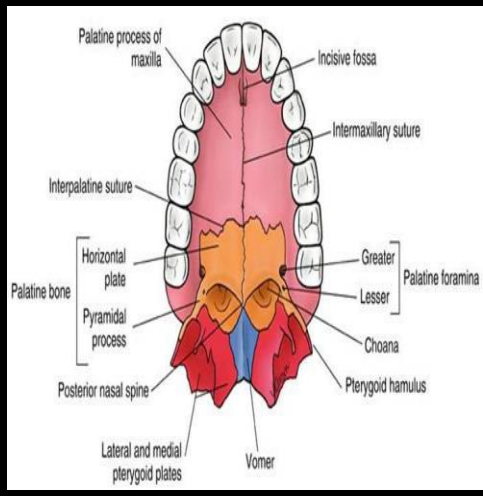
Boundaries of upper lip are nose, Nasolabial folds &Vermillion borders.
Vermillion is the red color dry mucosa.
White role is the line between skin & Vermillion.
Cupid's bow is part of vermilion.
Columella is the bridge between nasal tip & the upper lip.
Alae is the fold of skin(part of nose) joining the nasal tip & upper lip.
Nasal sill(nasal still, Simonard's band) is joining the alar base &columellar base.
Philitral column is the line joining the alar base & peak of cupid's bow.
Philitrum is the part of the upper lip between the Philitral colums.

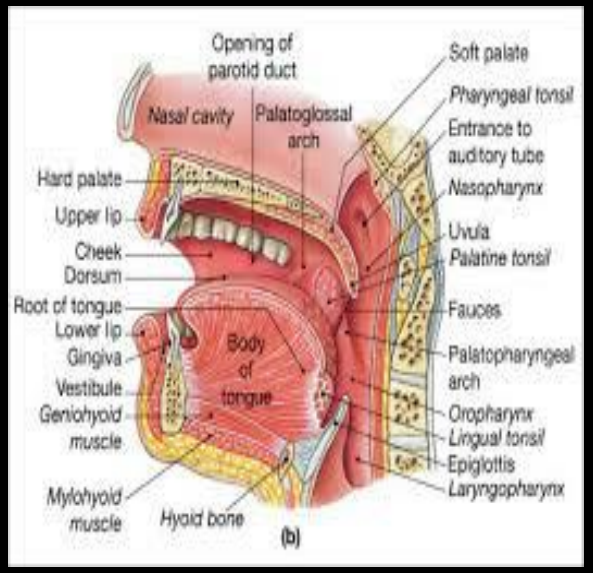


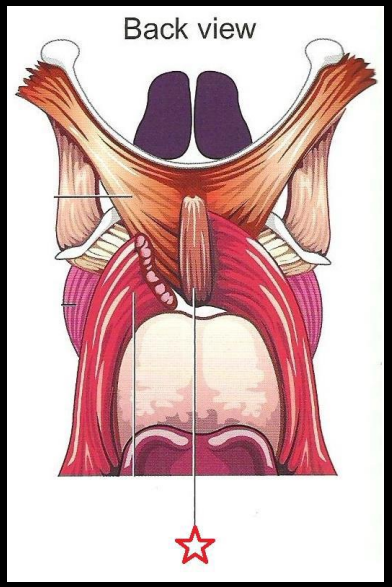


Palate is the roof the mouth& flour of the nose, composed of hard palate, bony part (anterior 2/3) & soft palate, muscular part (posterior 1/3). The function of the palate is by the soft palate upward & backward toward the pharyngeal walls result in complete separation of the nasal cavity & nasopharynx from the oral cavity & oropharynx. This function is important for sucking of milk which is essential for feeding in infants &during speech as it aids in building the necessary air pressure for letters formation.







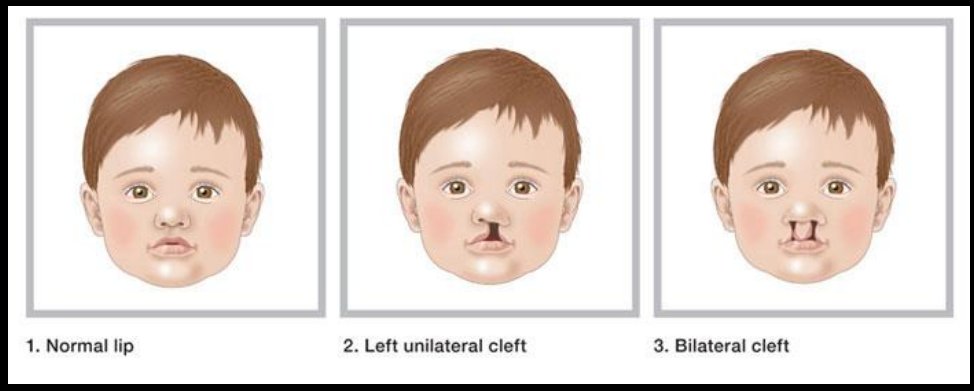


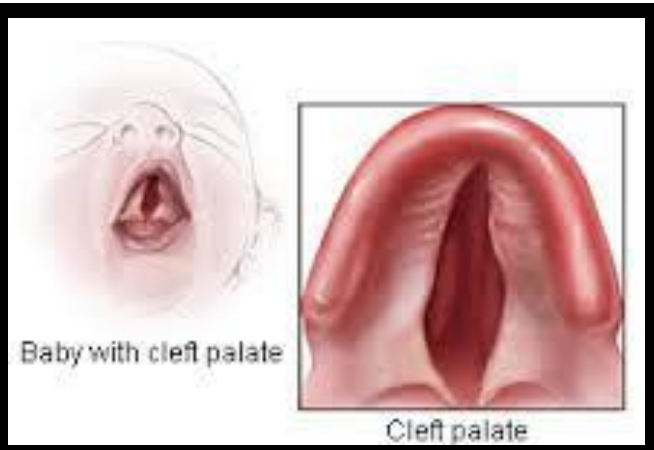
Cleft is a term used to describe failure of fusion or formation of parts during embryonic development e.g. cleft face, cleft lip, cleft palate, cleft hand, cleft foot.

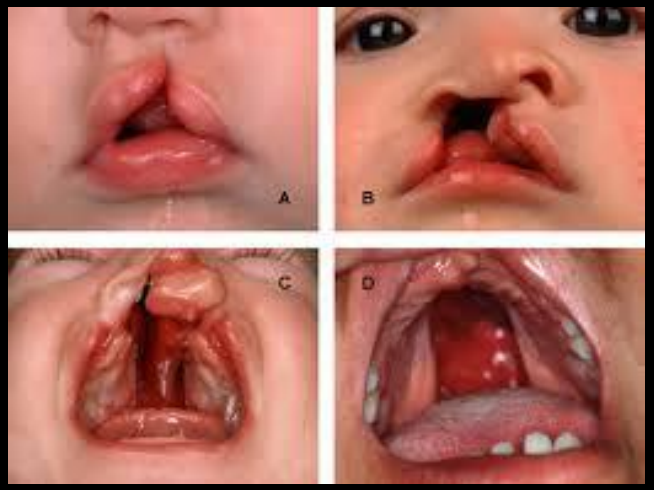
Cleft lip is usually occur at the line of Philitral column. It may be central, unilateral(left or right sided) or bilateral. Complete cleft lip is the cleft that extend beyond the nasal sill involving the nasal flour. Incomplete cleft is the cleft that not extend beyond the nasal sill.

Classification:

- **Cleft lip.**
- **Cleft palate.**
- **Cleft &cleft palate.**







Incidence:

-The second common congenital anomaly after club foot.

-If one of the parents is affected the possibility is 2%.

-One parent & one sibling the possibility is 14%.

-Normal parents &one affected sibling the possibility is 4.5%.

-Cleft sided cleft lip > right sided cleft lip.

- Cleft palate is more common in female while cleft lip & palate is more common in male.

Etiology:

1.Hereditary factors.

2.Environmental factors.

Drugs like steroids, anticonvulsants, alcohol.

Vitamins deficiencies or excess like vitamin A & riboflavin.

Radiation.

Problems:

1.Cosmetic----appearance.

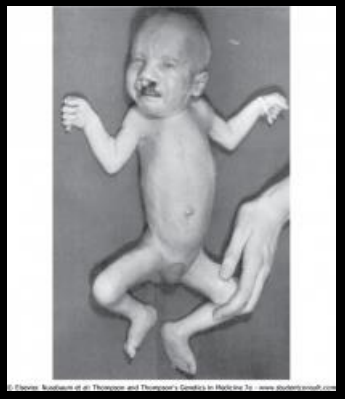
2.Feeding----defect in sucking mechanism.

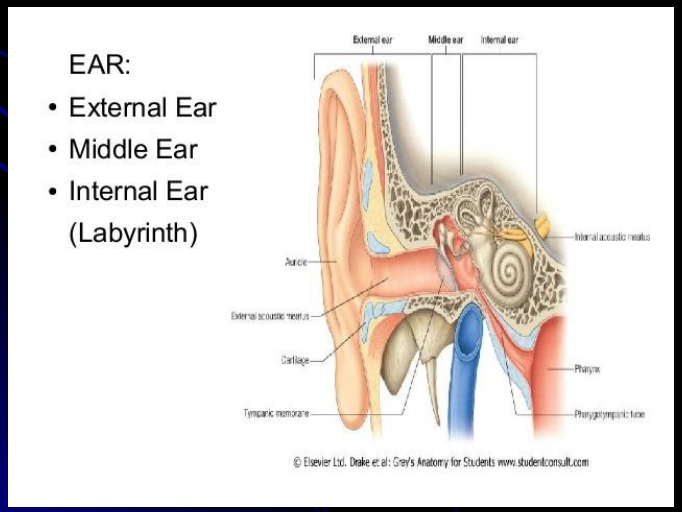
3.Failure to thrive--- needs follow up of growth as periodic checking of weight.

4.Speech problem-----nasal speech.

5.Otitis media(secretary type).

6.Respiratory problem.





Treatment:

-Management of problems.

-Surgery as repair of cleft.

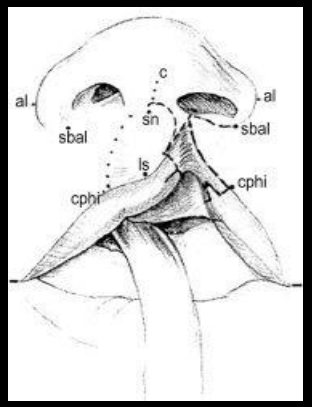
-cleft lip -------at age of 2.5- months.

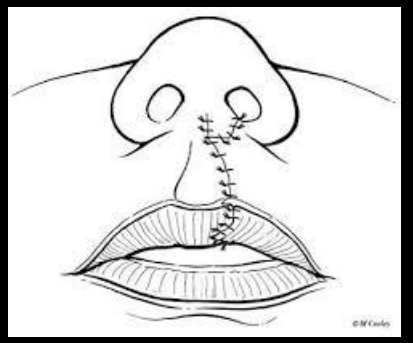
-cleft palate------at age of 1year.

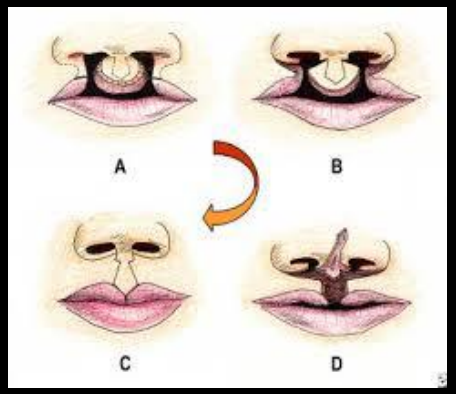
-nose ----------at age of 5year.

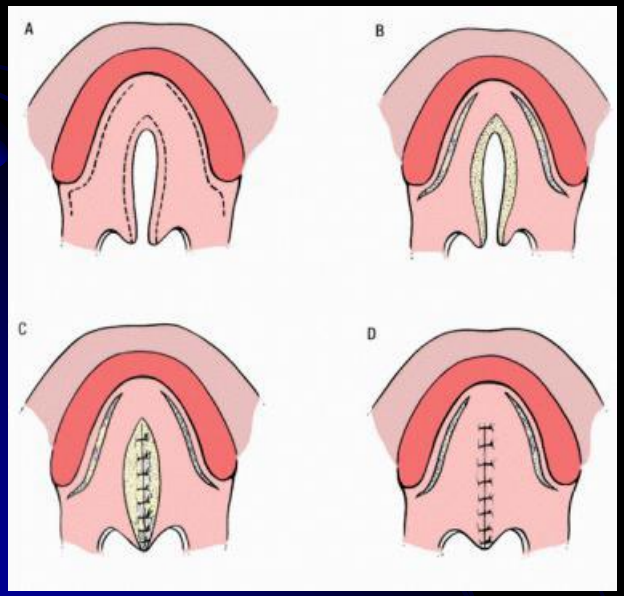
-alveolar cleft---at age of 7-8years.

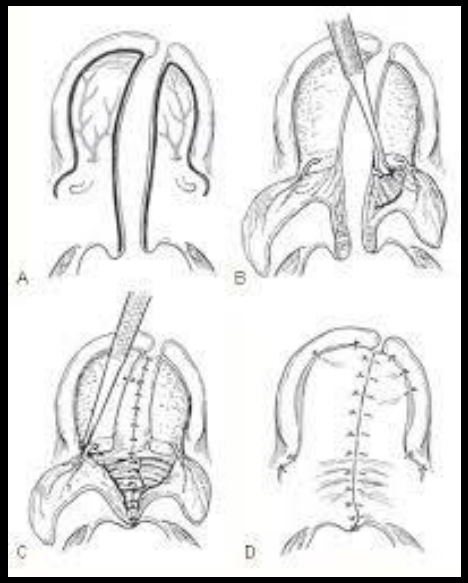
-Rhinoplasty----at age of 18years

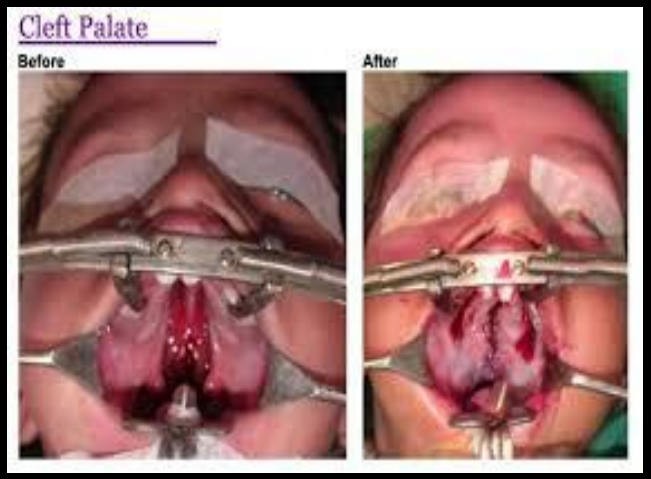












Congenital anomalies of the hand

1.Sydactyly:is fusion of two fingers or more.

-complete syndactyly:when fusion extend beyond the distal interphalangeal joint.

-incomplete syndactyly:when not reach the distal interphalangeal joint.

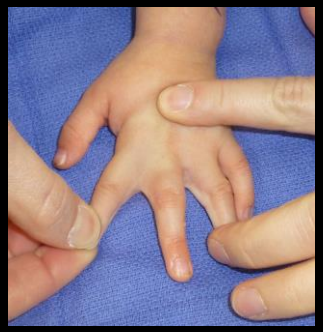
-simple syndactyly:is fusion of soft tissue only.

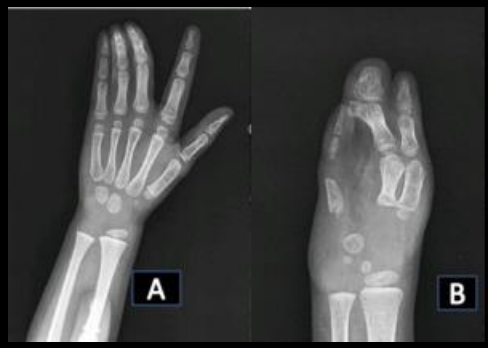
-complex syndactyly:is fusion of bone &soft tissue.

Treatment: is by surgery as separation and skin grafting at age of 3-4years.

2.Polydactyly: as presence of excess finger either on ulnar or radial side of the hand.

3.Macrodactyly:is big finger either primary involving all tissues or secondary involving single type of tissue as Neurofibroma or hemangioma.





Congenital anomalies of male external genitalia

Hypospadias

---------- 1.ventrally &proximally located meatus.

2.presence of chordee(fibrous cord) which the rudimentary part of corpus spongiosum.

3.meatal stenosis.

Types: according to the site meatus as proximal or distal OR ganular, coronal, penile, penoscrotal, scrotal perineal.

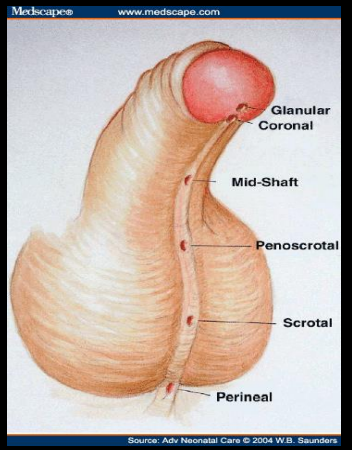
-Treatment is surgical repair in two stages:

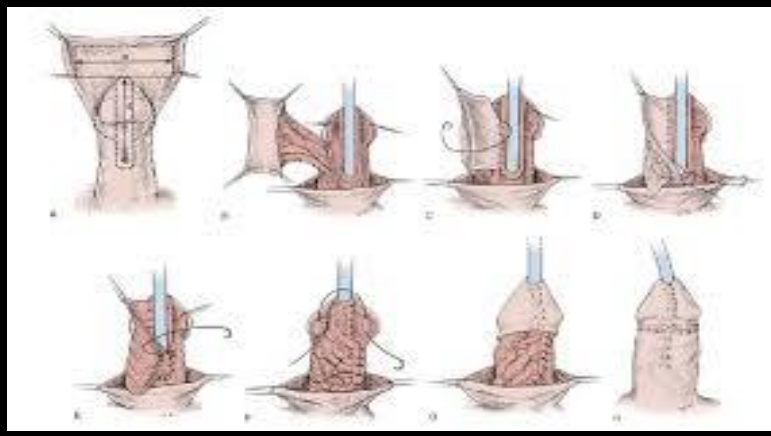
1.release of chordee.

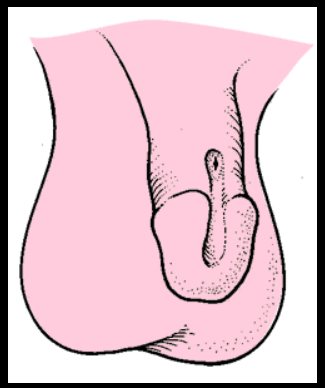
2.reconstruction of urethra.

The preferable age is the preschool age & the period between the 2 stages is at least 6 months.

Epispadias: is presence of meatus on the dorsal surface of the penis, it may be associated with ectopic vesicae.

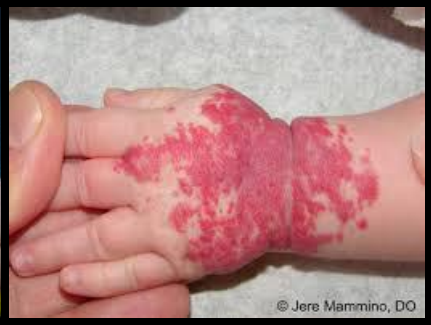


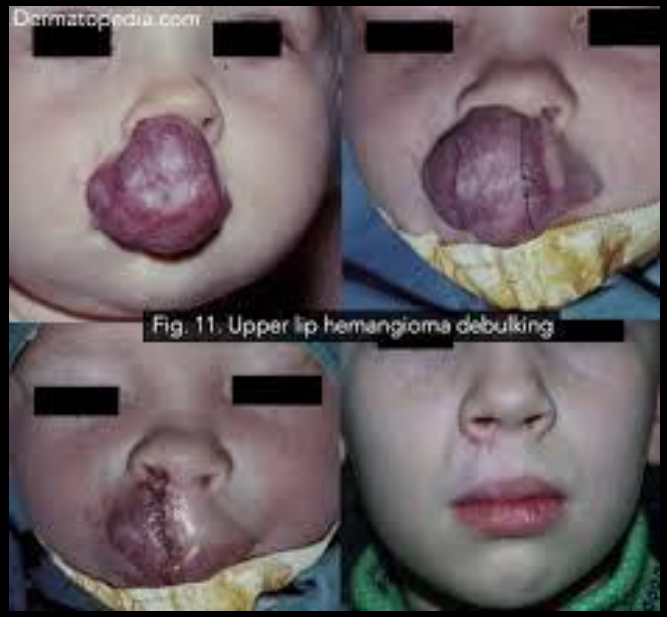




Cutaneous vascular malformation

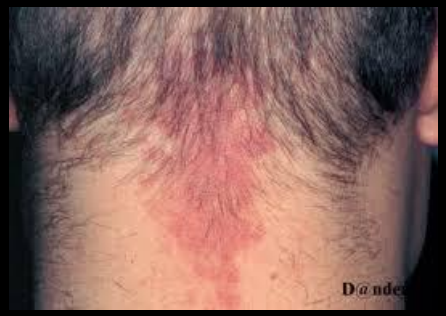
Straw-berry Hemangioma: is capillary hemangioma, composed of immature endothelium. It has 3 phases of growth; Growth phase as it appears in the few days of life as white or red color patch then changes to more red & purple color &elevates &increases in size gradually. It reaches its maximum size at age of one year. Platue phase as no change in color or size. Regression phase as it it decreases in size & change to white or skin color. This takes longer time up to age of 5-7 years more.





Port-Wine stain: is capillary hemangioma, composed of mature endothelium. It is not liable for regression. It appears as red-purple color patch, flat surface which may get granular appearance during puberty. The common site is the face on the distribution of the trigeminal (CN5) nerve, on part or whole of nerve distribution.

Salmon patch: is similar to port-wine stain but it regresses completely and the common site is the back of neck.

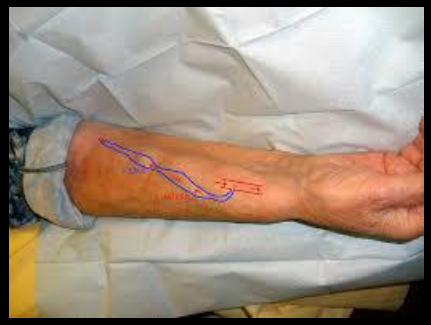


Pyogenic granuloma: is capillary hemangioma, may appears with port-wine stain or formed accidentally in incompletely healed wounds.

Cavernous hemangioma: is a venous type, less or not liable for regression. On examination it is compressible, deeply located, purple color masses.

A-V fistula: is an arterial type, associated with swelling or enlargement of the affected part & there are palpable thrill and brie.

Lymphatic malformation: like **Cystic hygroma** which is soft, transilluminable, poly cystic masses appear in the neck & face(or in the groin) after birth &increase in size gradually with age. It also increases in size when there is upper respiratory tract infections.



Complications of vascular malformations:

1.bleeding.

2.ulceration.

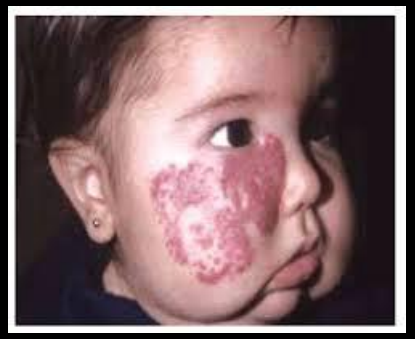
3.infections.

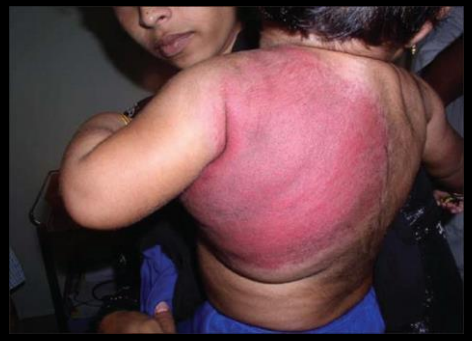
4.obstruction e.g. of airways, intestine, eyes.

5.disfiguring as enlargement (macrodactyly) or gigantism.

6.bleeding tendency as in casabach-merit syndrome in which there consumption coagulopathy & decrease in platelet count.

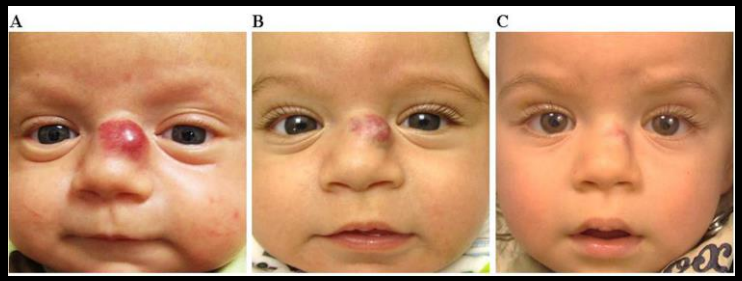
7. heart failure.

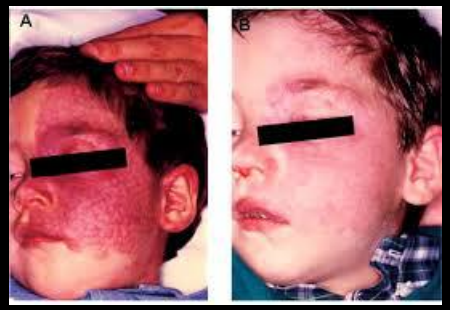




Treatment:

1. conservative as reassurance & wait for regression.
2. surgery as excision & reconstruction.
3. steroids.
4. Laser.
5. radiotherapy.
6. cryosurgery.
7. sclerosing agents.
8. embolization.





CAVERNOUS-HEMANGIOMA