

CLEFT LIP AND PALATE

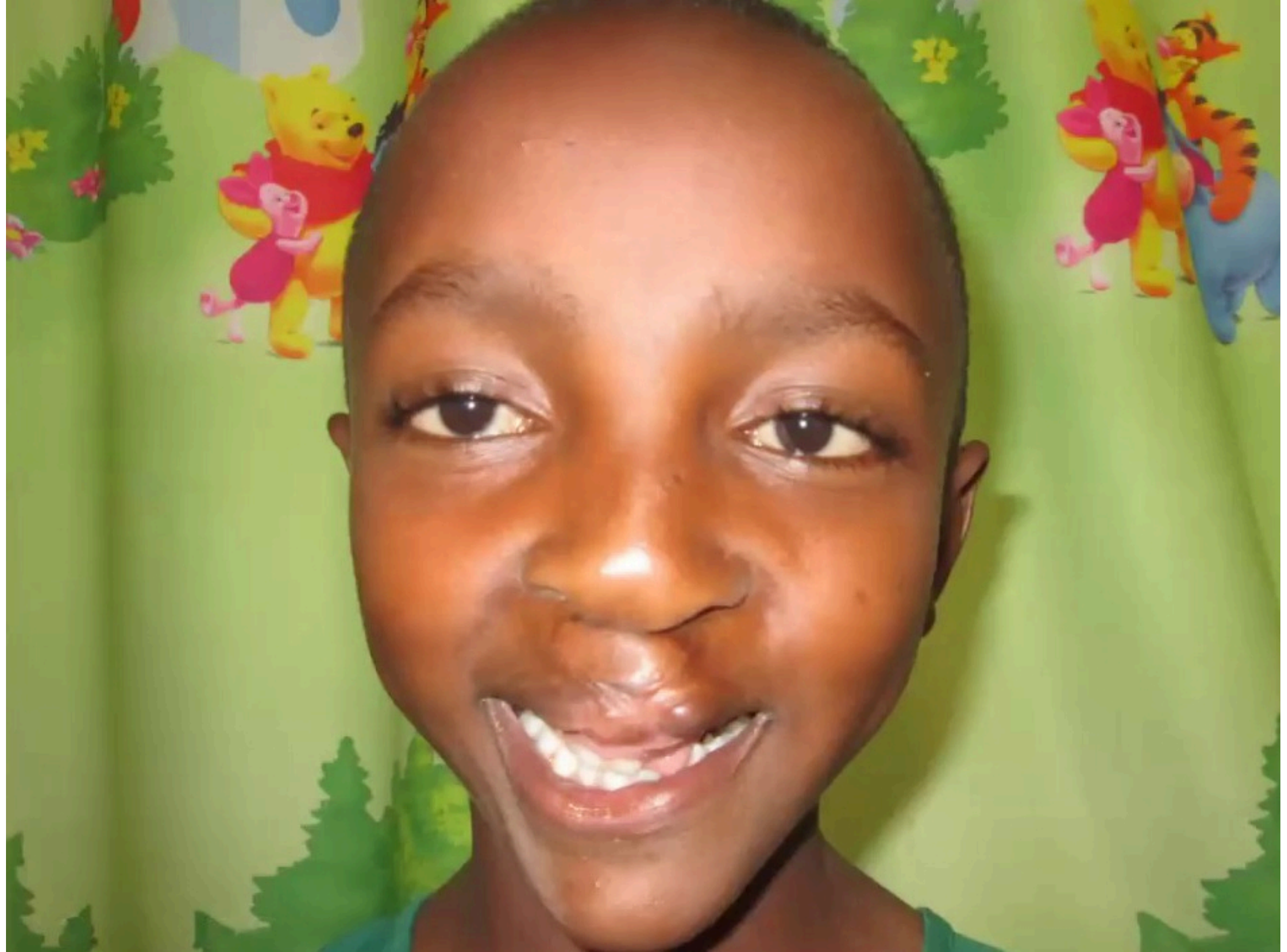
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BEFORE & AFTER PHOTOS.



4 MONTHS









CL & CP

- **One of the most common birth defects**
- **Certainly the most visible**
- **Worldwide overall incidence is estimated to be 1/700 with wide variability among races**
 - Low incidences seen among blacks
 - Highest incidence among American Indians, Japanese & Chinese

LECTURE OUTLINE

- **Defination,**
- **Cultural beliefs.**
- **Morphological/Anatomic
al Landmarks**
 - Face in general
 - Lip
 - palate
- **Embryology/Morphogen
esis**
- **Syndromes**
- **Diagnosis &
Management**



UNILATERAL COMPLETE CLEFT LIP & PALATE



24.09.2011 12:31

DEFINATION (HARE-LIP/MDOMO SUNGURA/MDOMO ULIYO NA MPASUKO/KAAKAA ILIYO NA MPASUKO)

Cleft lip and cleft palate – Clefing congenital deformities of the face. Caused by abnormal development during gestation.

A cleft is a sub-division in the body's natural structure, regularly formed before birth. This type of deformity is sometimes referred to as a *cleft*. The term hare lip is sometimes used colloquially to describe the condition because of the resemblance of a hare's lip.

Oxford dict def: a crack or split occuring naturally, eg in the ground or in the rock.

PATHOGENESIS

- Cleft lip occurs when there is failure of fusion of the frontonasal process and the maxillary process
- CP occurs due to the failure of fusion of the lateral palatine processes and the nasal septum (i.e failure of formation of the secondary palate)
 - When Cleft Palate (CP) occurs, the uvula is usually split.

BELIEFS ON CAUSE OF CLEFTS

Africa – Pregnant woman frightened after seeing/meeting a snake (Kikuyu) or an individual with protruding teeth (Borana/TZ)

Lun Heng (Chapter 6), Wang Chong says, "If a pregnant woman eats rabbit, the baby will have a cleft lip."

The Chinese word for cleft lip is *tuchun*, literally "harelip"

What does your community believe is the cause of clefts?

ANATOMY: NORMAL LIP ANATOMY

- **Muscles:**

Orbicularis oris, Levator labii superioris & Nasalis/depressor septi nasi

- **Vertical lip length:**

Newborns-10mm, age 3/12-13mm & adult-17mm

- **Distance btwn the peaks of cupid's bow is:**

3mm at 3/12.

- **Arterial bld supply:**

Labial art bilaterally

- **Sensory innervation:**

CN V (Trigeminal nerve) – Max Division (V2)

- **Motor innervation:**

CN V11 (Facial nerve) – Zygomatic 5 buccal branches

BASIC ANATOMY- LIP

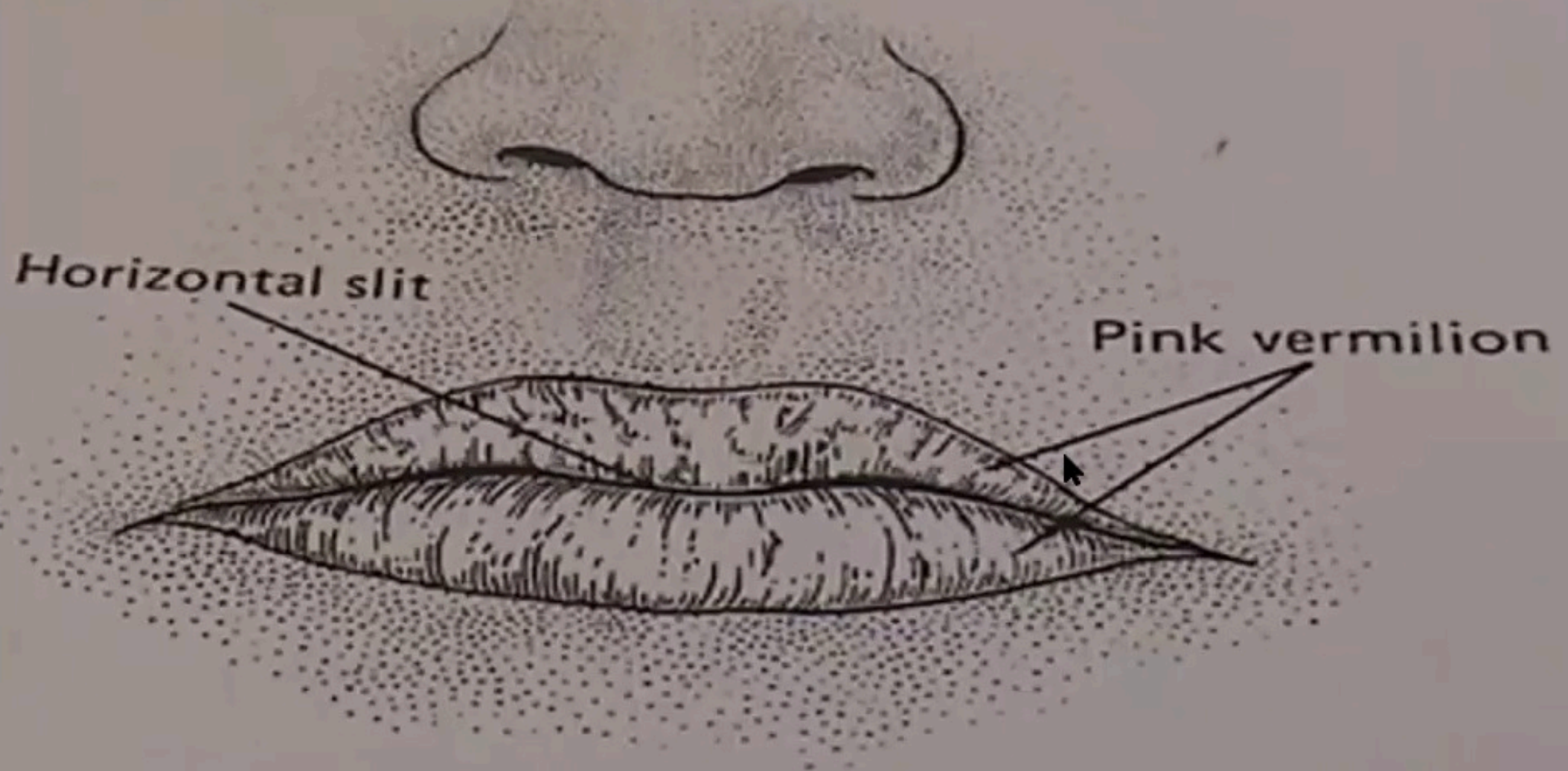
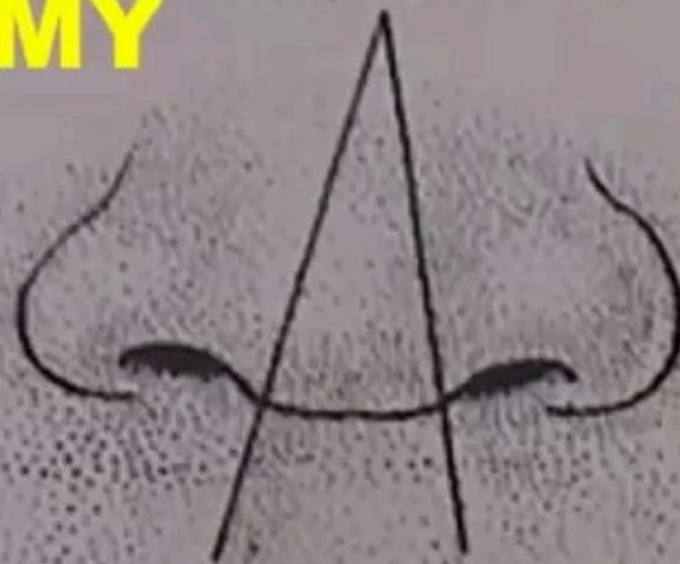


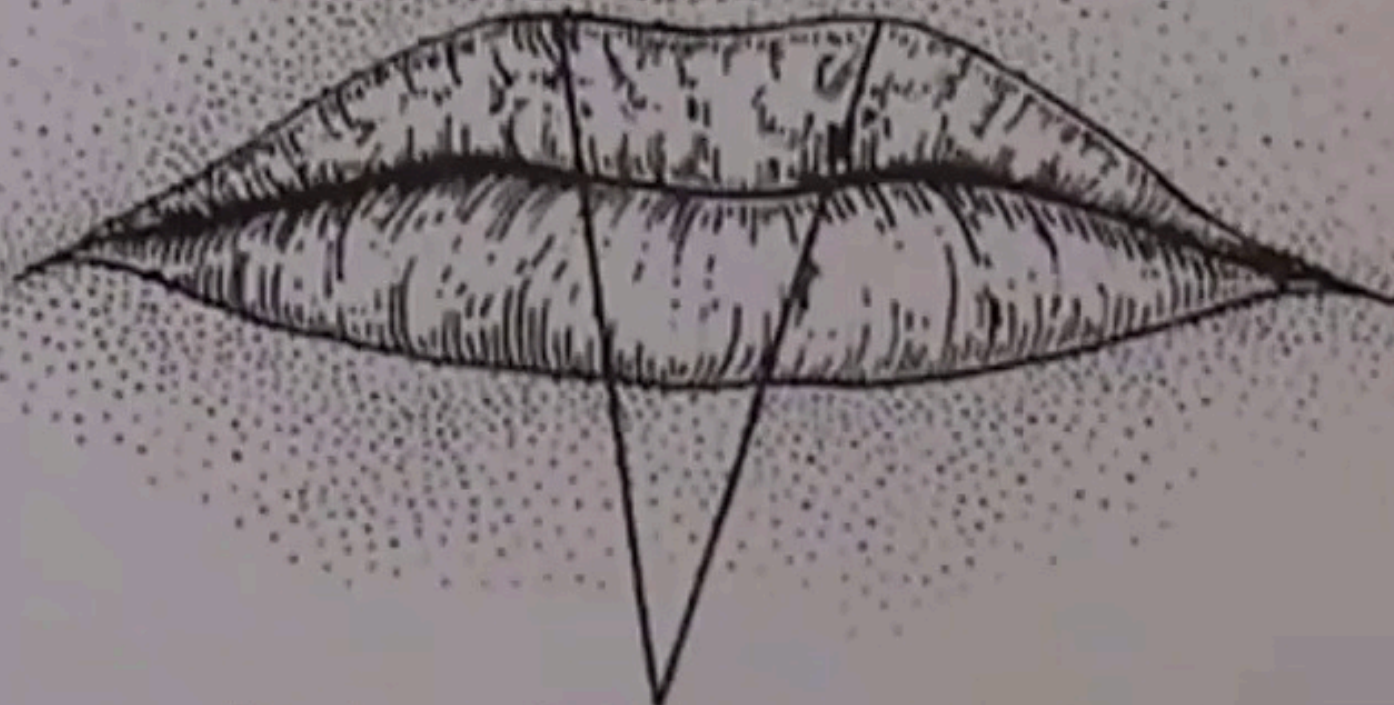
Figure 5-

LIP ANATOMY

Philtral lines

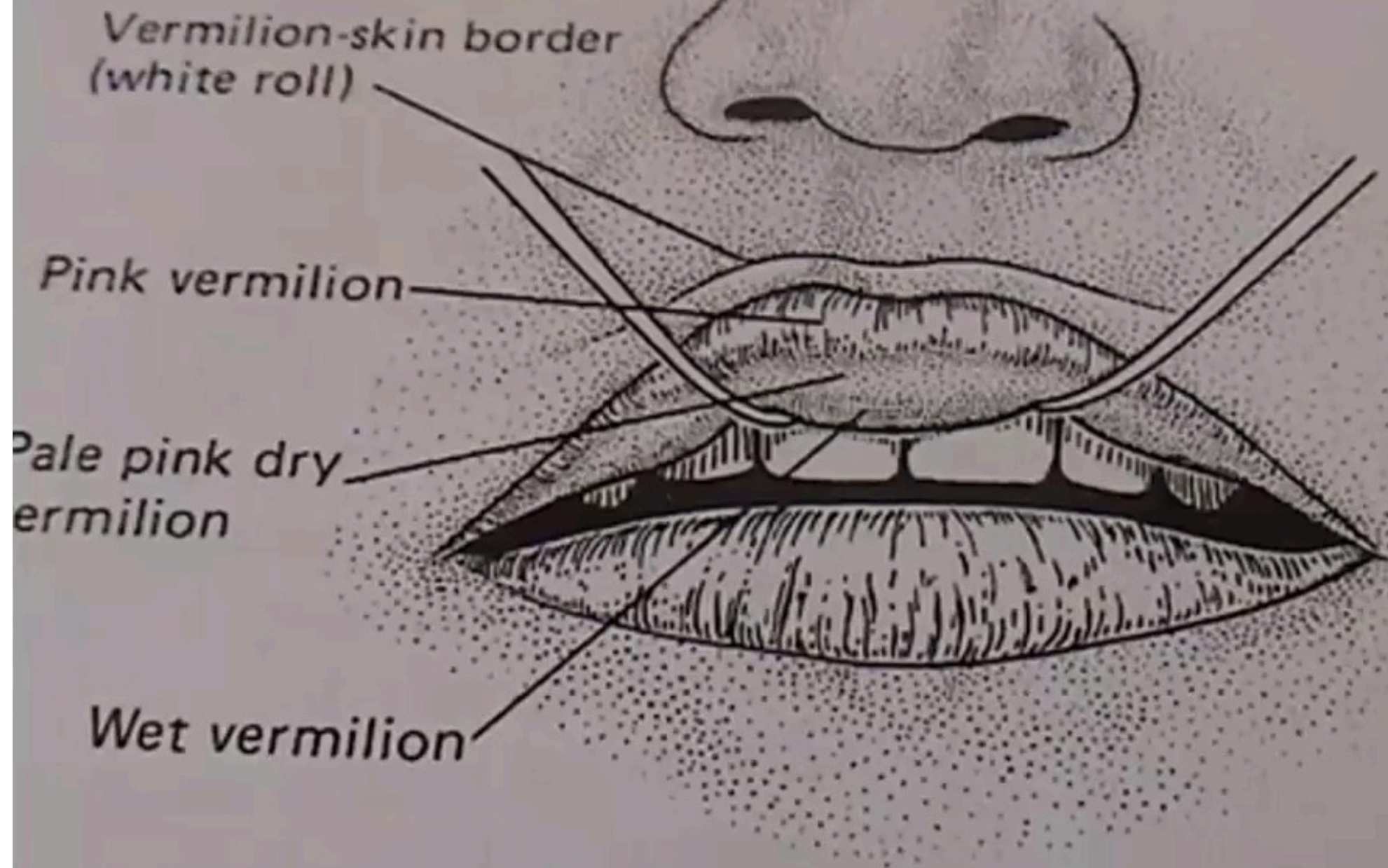


Philtral dimple

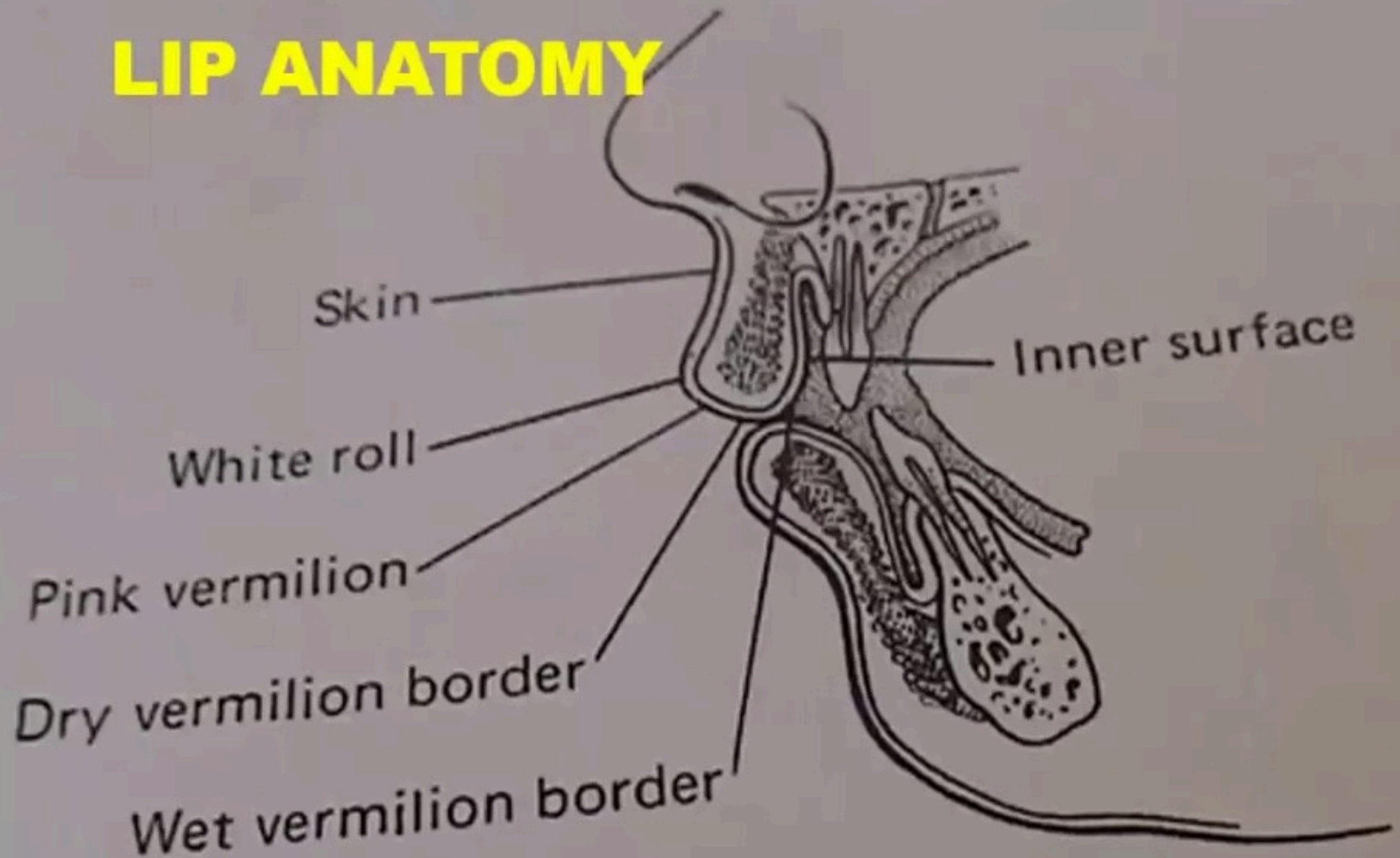


Peaks of Cupid's bow

LIP ANATOMY



LIP ANATOMY



LIP ANATOMY

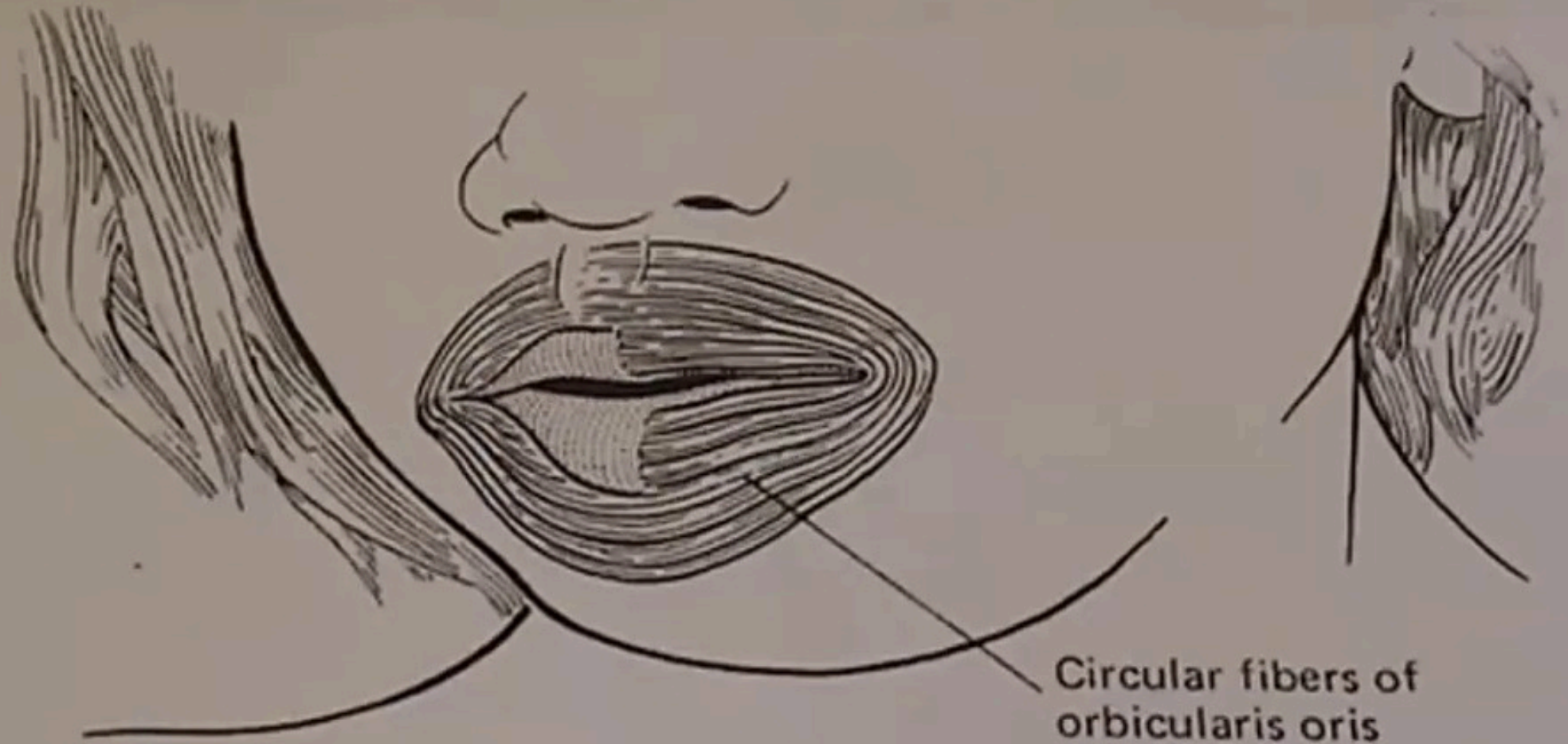


Figure 5-5

PALATE ANATOMY

The primary palate includes:

- the lip,
- alveolus &
- hard palate ant to the incisive foramen

Secondary palate includes:

- hard palate posterior to the incisive foramen &
- the soft palate

PALATE ANATOMY

ANATOMY OF THE VELOPHARYNX

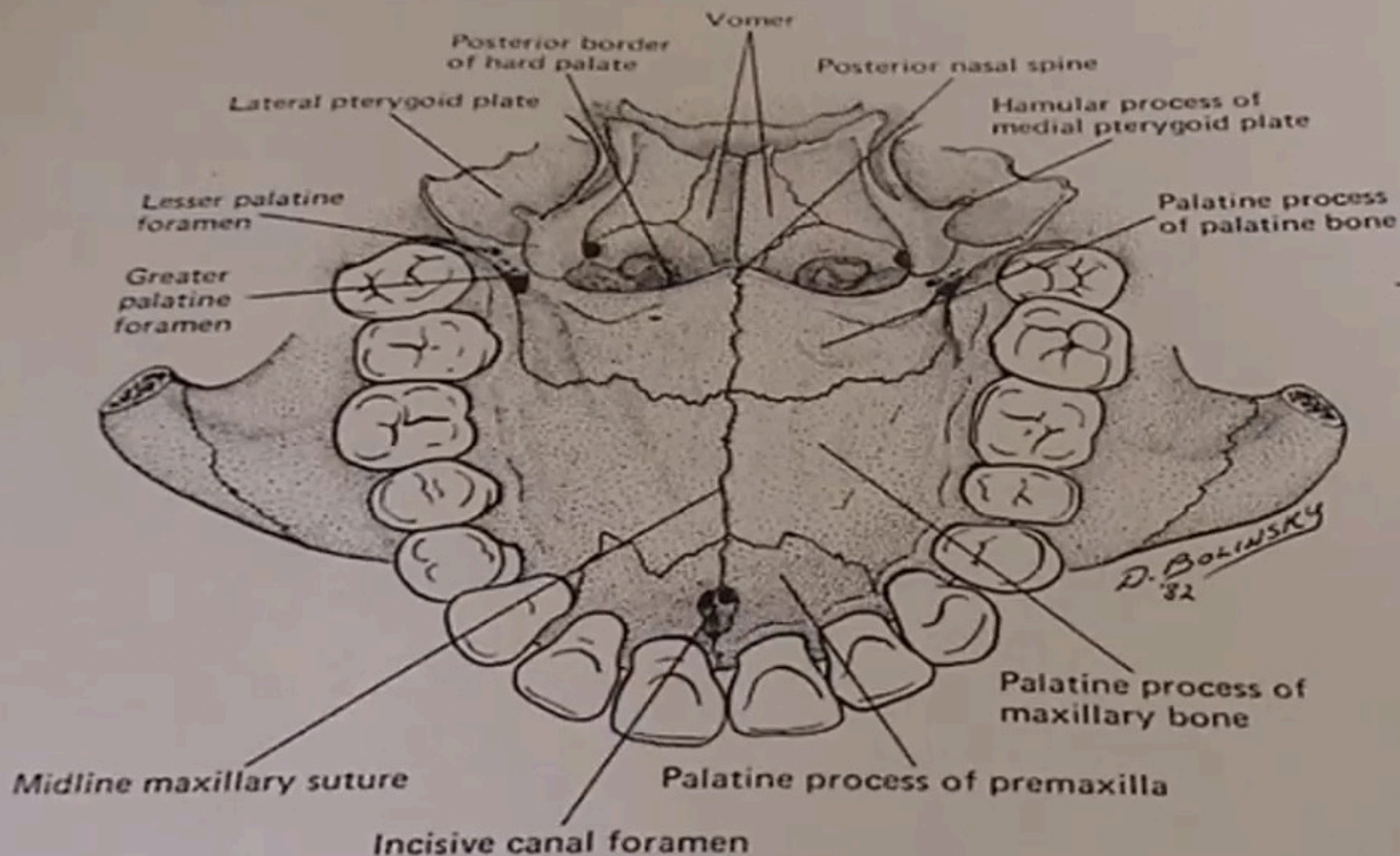
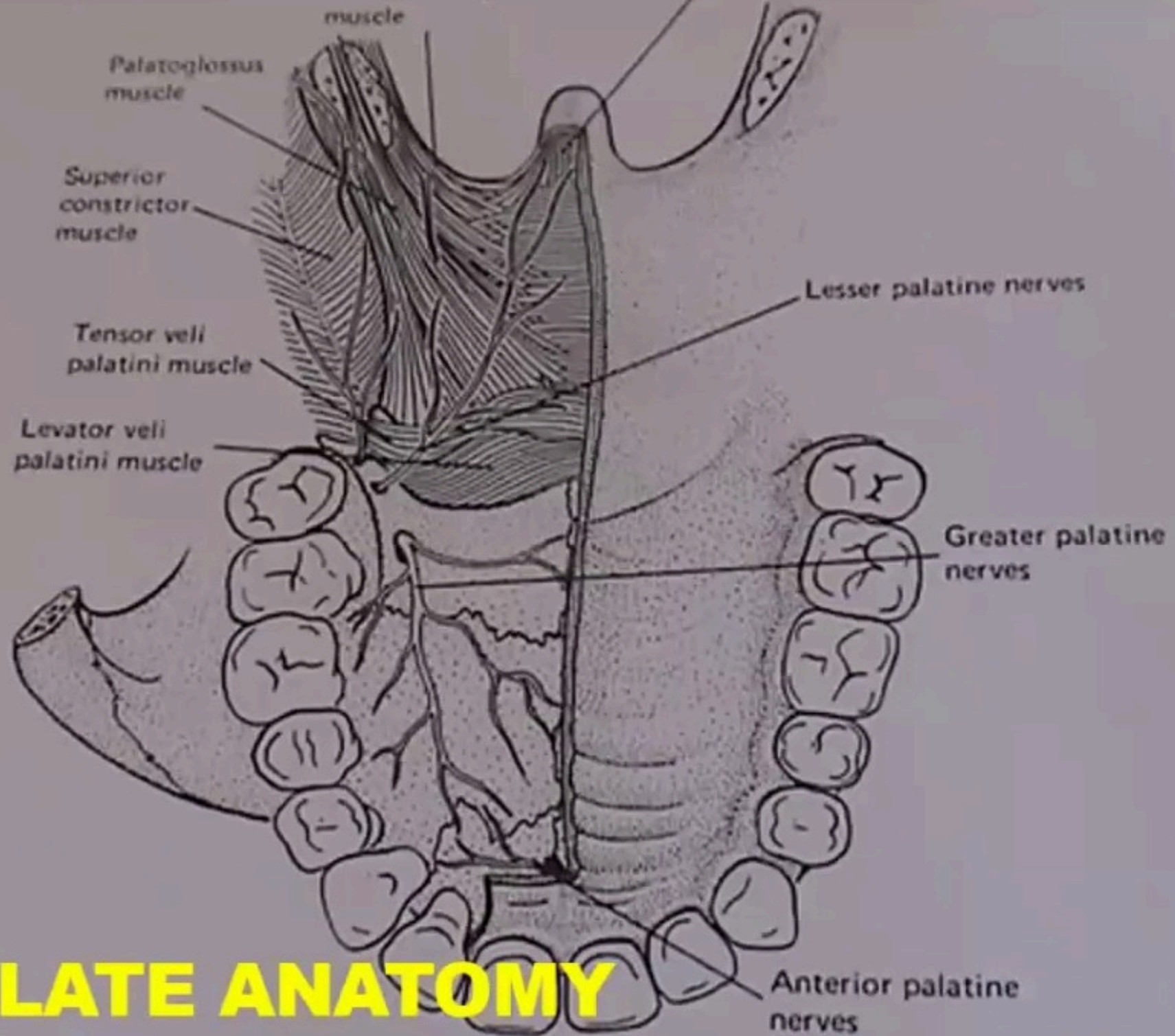


Figure 1-2

BONY ANATOMY



PALATE ANATOMY

EMBRYOLOGY OF CL&P

- **Face dvps from neurocrest cells**
- **Frontonasal process forms the nose & prolabium**
- **Primary palate starts to dvp on day 35 with formation of paired medial & lateral nasal & maxillary processes**
- **Fusion of these processes completes the dvpment of primary palate**

Cont.

- **After the development of primary palate, palatal shelves form as medial outgrowths from maxillary processes**
- **Initially they grow vertically but later elevate horizontally to fuse with each other**

Cont

- **Elevation due to hydration of hyaluronic acid.**
 - Interference with this step associated with CP
- **The left shelve elevates last & may be why left sided CP is commoner**
- **Failure of fusion, hypoplasia, breakdown or mechanical obstruction to fusion leads to CL**

ETIOPATHOGENESIS STILL NOT WELL UNDERSTOOD

- The etiopathogenesis of clefts has been widely studied but is still poorly understood
- It has been estimated that about half of the cases are nonsyndromic

MOLECULAR MECHANISMS OF CL&P

There is interplay bwn matrix metalloproteinase & tissue inhibitors metalloproteinase in palatogenesis.

Studies suggest that MMP are involved in palatal growth & in particular MMP-3 is involved in fusion process of secondary palate.

Loss of MMP may also be related to abn in the expression of regulatory proteins e.g. TGF-B & EGF.

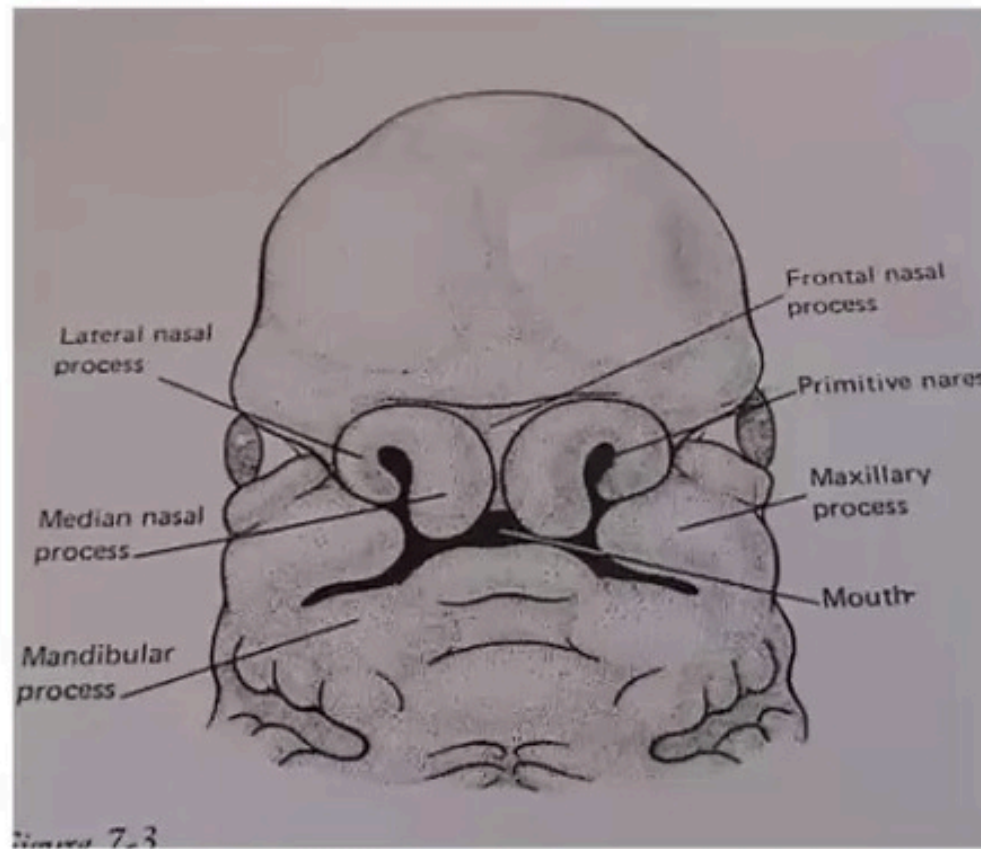
THEORIES ON EMBRYOGENESIS

Classic theory - CL&P resulted from failure of nasomedial & nasolateral processes of embryo to fuse

Mesodermal penetration theory - Notes that the embryo is an epithelial bilayer in the region of the face until mesoderm migration b2in the bilayer forming the facial processes

Failure of this results in clefting

EMBRYOLOGY



CLASSIFICATION (ACCORDING TO KERNAHAN & STARKE)

1. Clefts of primary palate

Unilateral – complete

- incomplete

A microform cleft is a very minor cleft where no surgery is required to correct it. A microform cleft can appear as small as a little dent in the red part of the lip or look like a scar.

Bilateral - complete

- incomplete

Median - complete (absent pre-maxilla)

- incomplete

CLASSIFICATION (CONT.)

2. Clefts of secondary palate

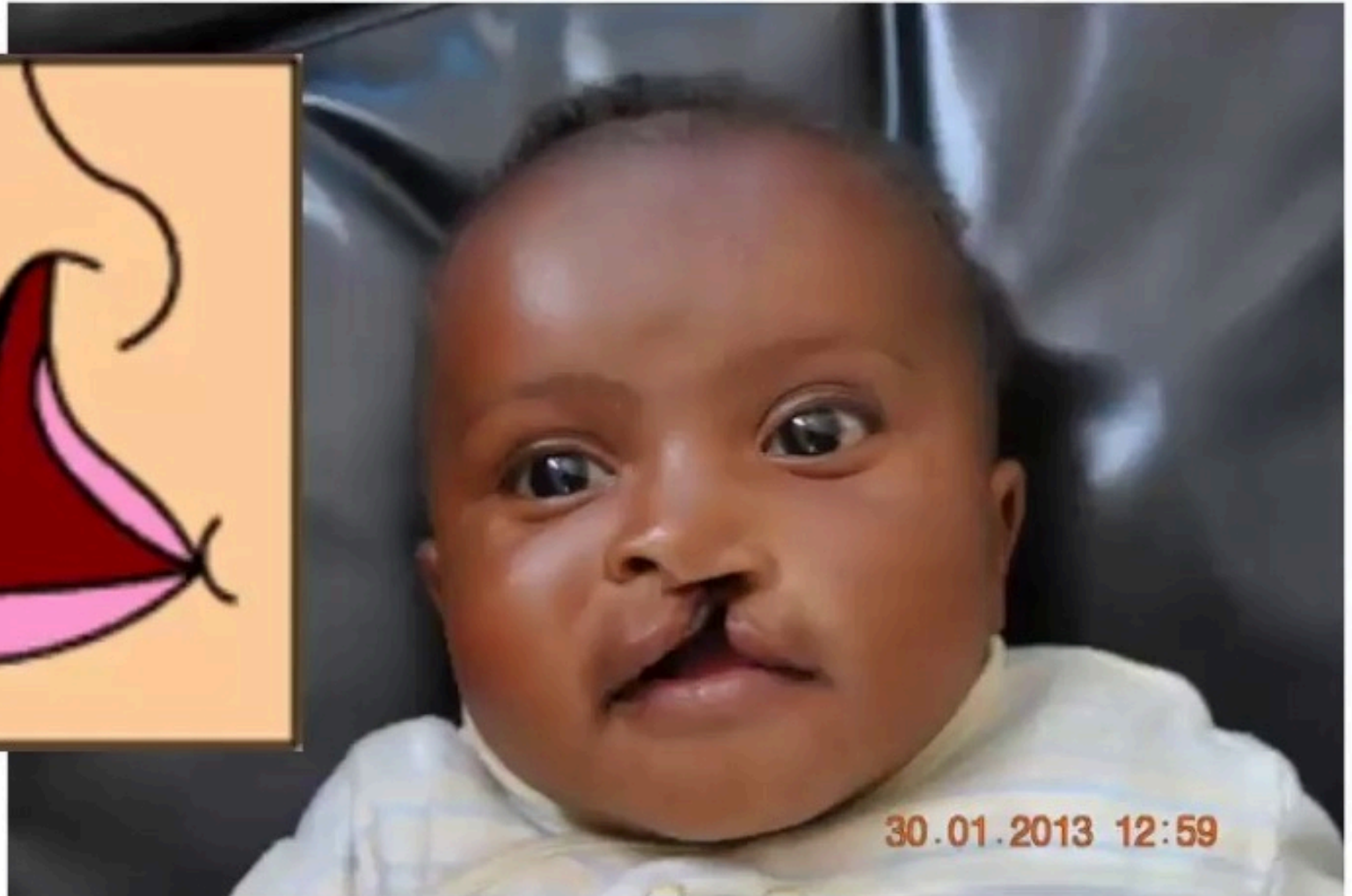
- a) complete**
- b) incomplete**
- c) submucous**

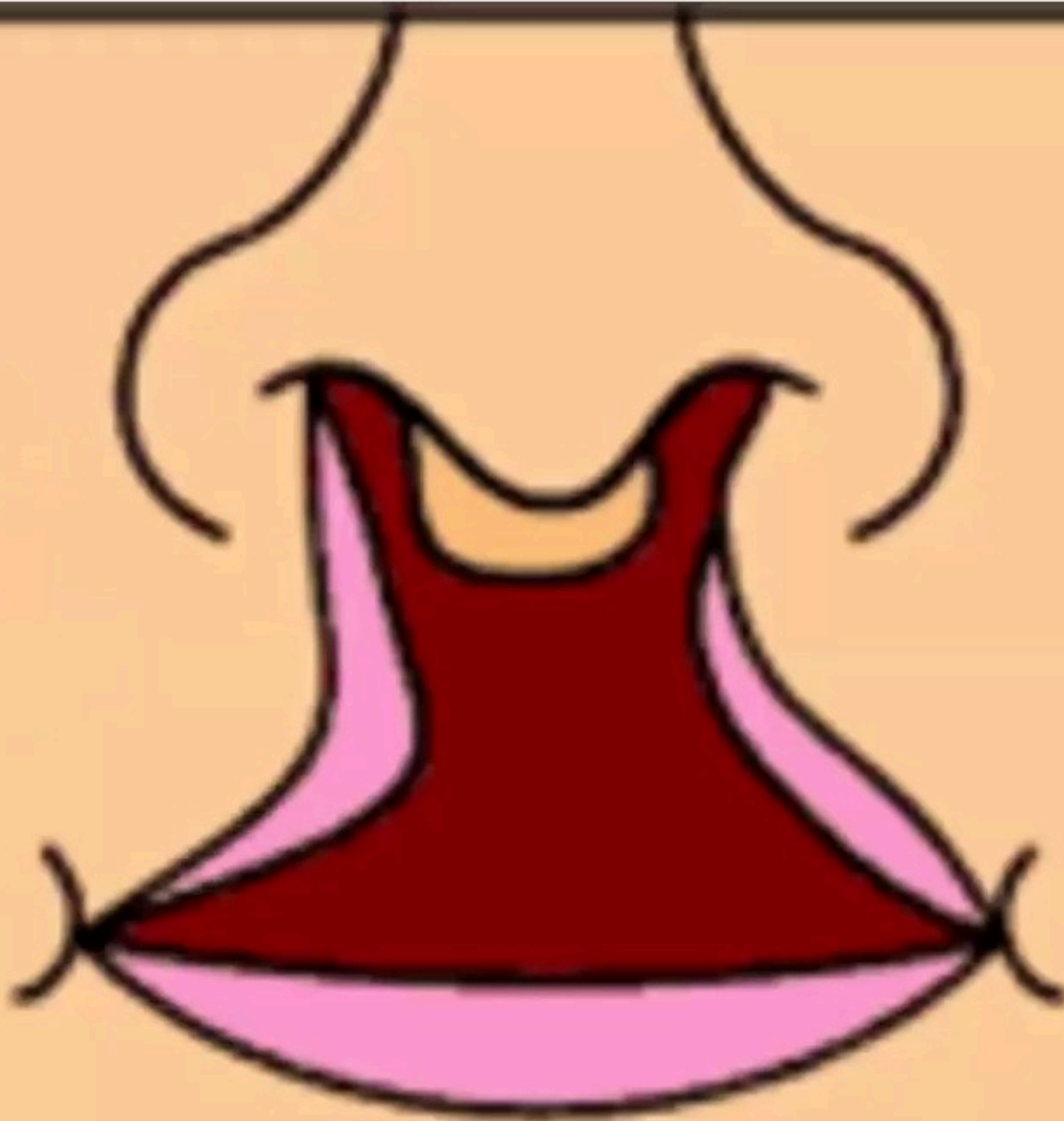
CLASSIFICATION (CONT.)

3. Clefts of both primary palate can be;

- a) unilateral**
- b) bilateral**
- c) median**

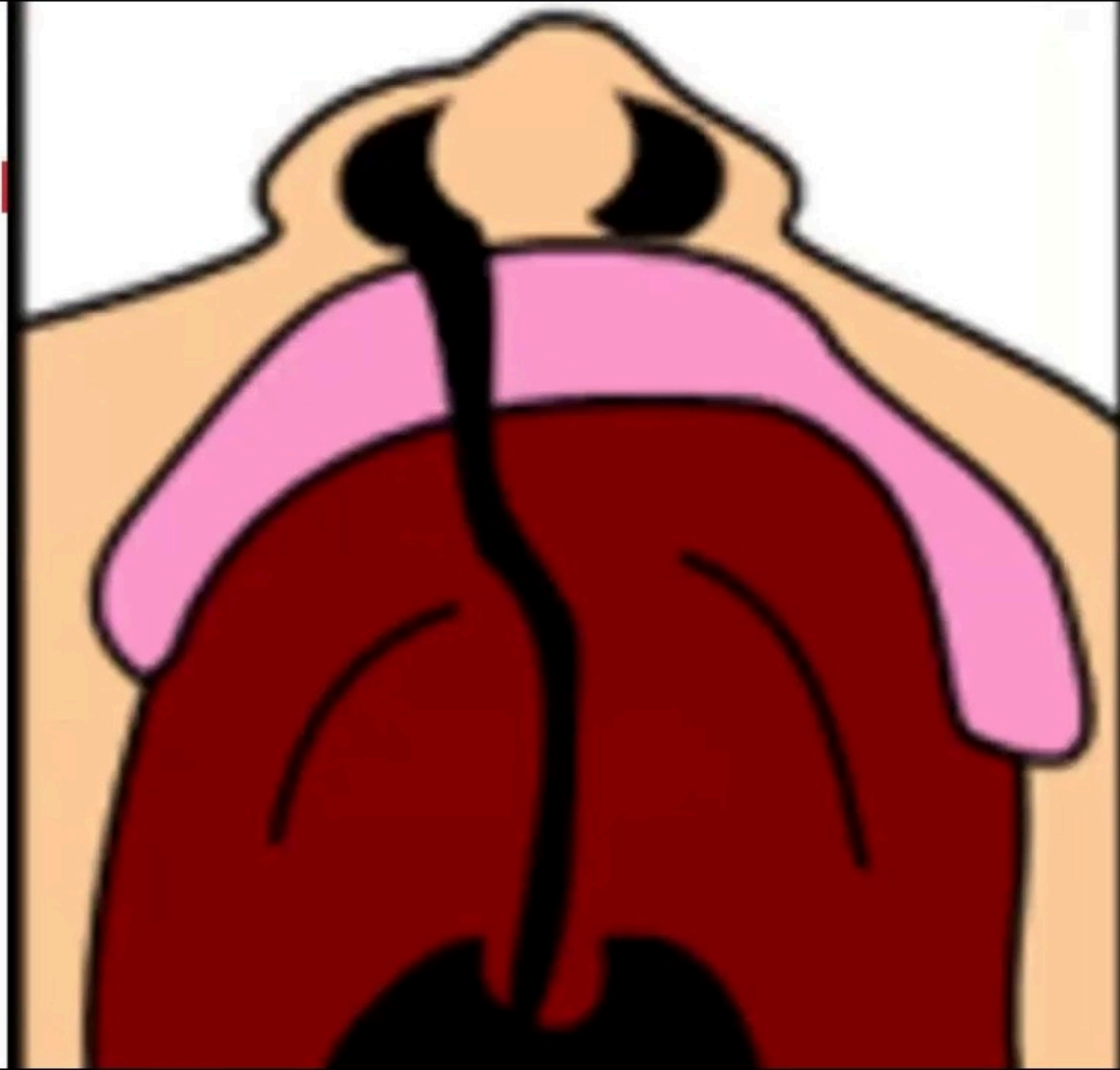
UCL – LT COMP



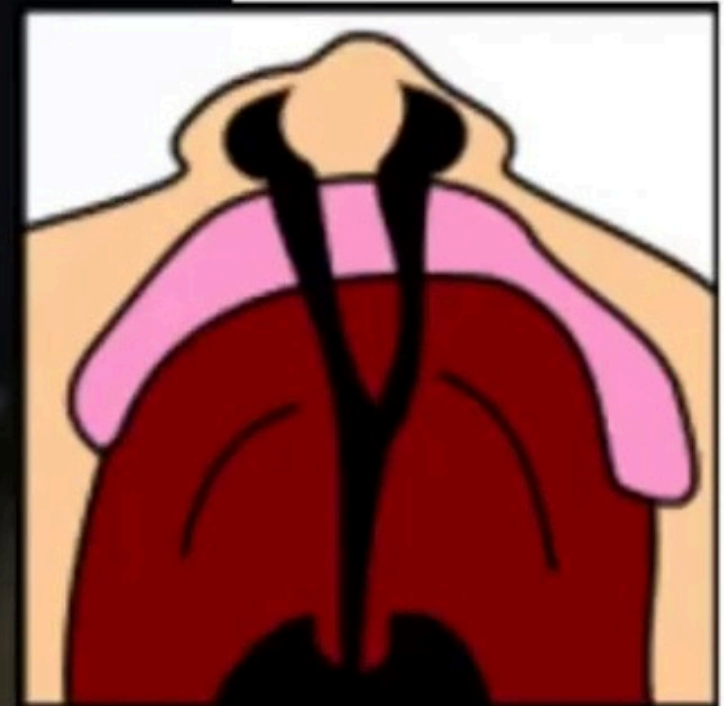


BCL - COMP

U

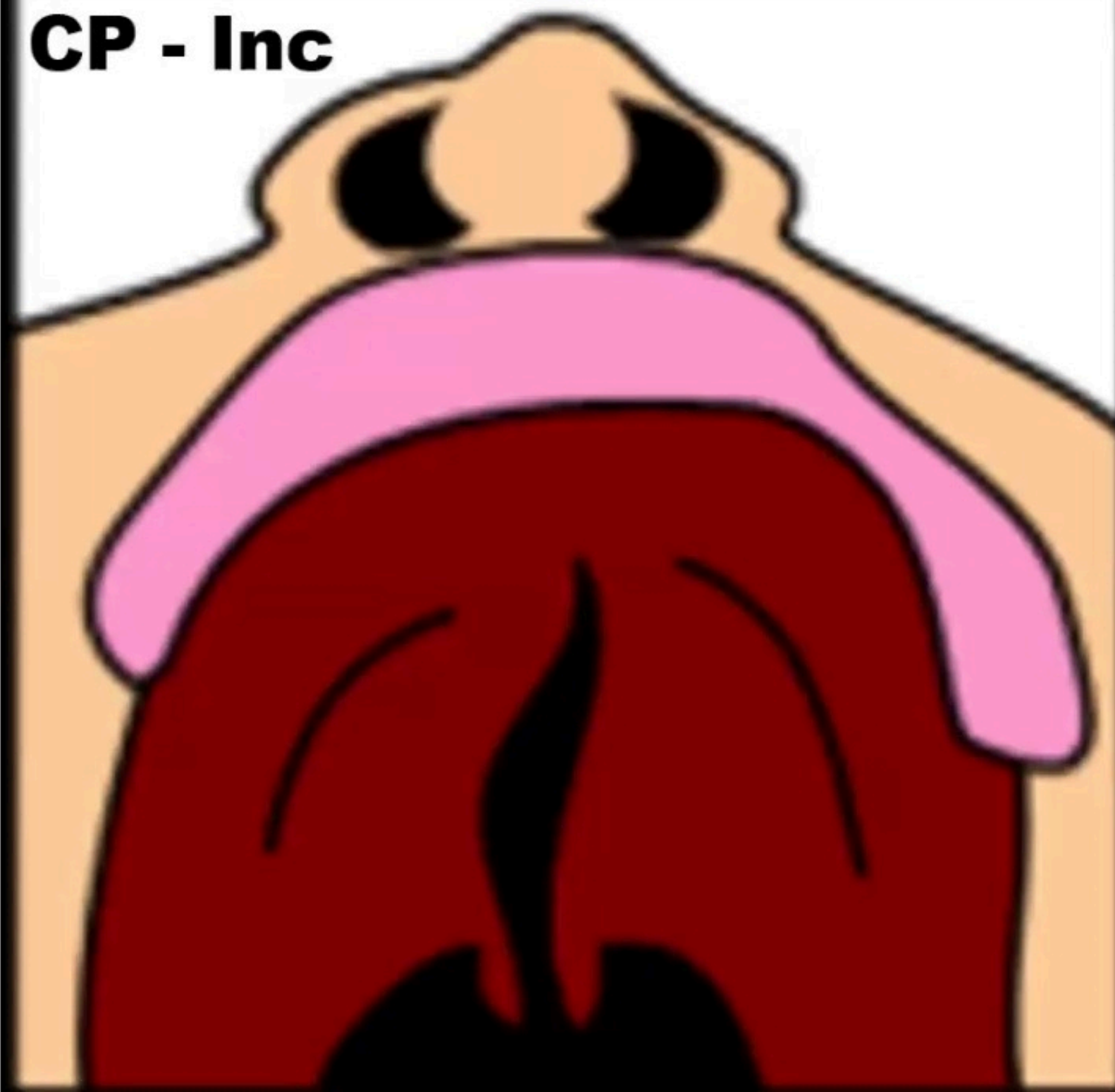


BIL CP - COMP





CP - Inc



UPPER LIP OF ANIMALS.



INCIDENCE: CLEFT LIP

- Incidence varies across the races
- Overall incidence of CL & CL&P is 1:1000
- CL
 - 1/500 live births in Asians,
 - 1/750 for whites &
 - 1/2000 for blacks
- No local statistics (?1:1000)
- M:F = 2:1
- CLP:CL = 2:1

DEMOGRAPHICS OF CL

M:F = 2:1

CLP:CL = 2:1

3% are syndromic

Risk factors;

1. Medications/Drugs – Phenytoin, methylprednisolone (Solu-Medrol), steroids, phenobarbital, diazepam & isotretinoin. Folate & Vit B6 defn.
2. Smoking & Alcohol.
3. Parental age esp father's age or both father & mother over 30 yrs old.
4. Family hx (Genetics) – parents have one child with CLP:4%, one parent has CLP:2-4%, parents with 2 children with CLP:9% & one child & one parent with CLP:14-17%.

SYNDROMES ASSOCIATED WITH CLEFT LIP & PALATE

- **Van Der Woud Syndrome**
- **Wardenburgs Syndrome**
- **Down Syndrome**
- **Trisomy 13**
- **Stickler Syndrome**
- **Pierre Robin sequence**
- **Velocardiofacial syndrome**
- **Median cleft lip**

DIAGNOSIS

In utero – Transvaginal US (*11 wks gestation*).

- Abdominal US (*16 weeks gestation*).

3-D US best. Sonographer dependent.

- MRI (*CP*).

Peri/Postnatal – Parents, Midwife, Doctor.

PRENATAL DIAGNOSIS



ASSOCIATED PROBLEMS

- **Speech**
- **Feeding**
- **Hearing problems due to middle ear infxn**
- **Cosmesis**
- **Dental irregularities**
- **Impaired mid-face growth**
- **Potential psychological problems**

CARE OF THE NEWBORN WITH C.P.

Feeding the infant with CP:

- No ability to create negative pressure

- Nipple pushed through cleft in hard Palate

- No coordination breathing and sucking,
»»lengthen the feeding

- Milk escape through the nose

FTT

TREATMENT

Surgery – Cheiloplasty, Palatoplasty or simply Cleft lip/palate repair.

Multi-disciplinary (Team concept) involving a Surgeon (Plastic & Maxillofacial), Speech/language pathologist, Paediatrician, Orthodontist, General Dentist, ENT/Otorhinolaryngologist, Audiologists, Geneticists, Social workers, Paediatric Nurse practitioners and Psychologists. All comprise members of the Cleft palate team. (First 3 is minimum requirement for Cleft team according to Amer Cleft Palate-Maxillofacial Ass. ACPA)

Neonate –

Feeding by cup & spoon, Feeding bottle. An infant with a cleft palate will have greater success feeding in a more upright position.

Gravity will help prevent milk from coming through the baby's nose if he/she has cleft palate

REPAIR OF CL

- **Before definitive lip repair is done pt may undergo a course of presurgical oral orthopedic**
- **Lip repair is carried out when the child's gc & wt permit safe GA induction**
- **Rule of 10 applies**
- **Repair may be done earlier in healthy babies**

TREATMENT PROTOCOL

Age 3 months – CL repair

Age 6 months – pre-surgical orthodontics

Age 9 months – speech therapy begins

Age 9 – 12 months – CP repair

1-7 yr – orthodontic treatment

7-8 yr Alveolar bone graft

REPAIR OF CL CONT

- **Effective repair must realign the vermillion & cupid's bow of the lip**
- **Reconstruct the upper lip & philtrum, re-approximate the orbicularis oris within the repair**
- **Millard's repair (rotational advancement) is the most popular for unilateral CL**
- **Bilateral CL more challenging**
- **Revision may be needed at 5yr**

GOALS OF REPAIR OF CP

(SOFT PALATE)

- **Improvement of speech**
- **Lengthening, muscles alignment and movement**
- **Improving Eustachian tube function**

REPAIR OF CP

- **Timing of repair controversial**
- **Usually repaired b/n 6-18 months**
- **Too early, you interfere with facial growth**
- **Too late, you interfere with speech development**
- **Von Langenbeck palatoplasty oldest method and still used**
- **Veau Wardill Kilner palatoplasty. W-shaped MP flaps advanced in V-Y fashion**

CONT

Furlow's double opposing Z-plasty technique

Complications like palatal fistula may result

Are treated by raising adjacent flaps of mucoperiosteum

If very large gingival or tongue tissues may be required

DETERMINANTS OF TREATMENT OUTCOME

- **Quality of the initial nasolabial and palatal repairs is the critical determinant of outcome, in terms of:**
 - appearance
 - Speech
 - facial growth
- **Centre based cleft care is best if available**

SURGICAL COMPLICATIONS PALATOPLASTY

Early

Bleeding

Airway obstruction/compromise

- * Pts should be monitored closely
post-operatively**

Late

Fistula

Reduced movement of the soft palate

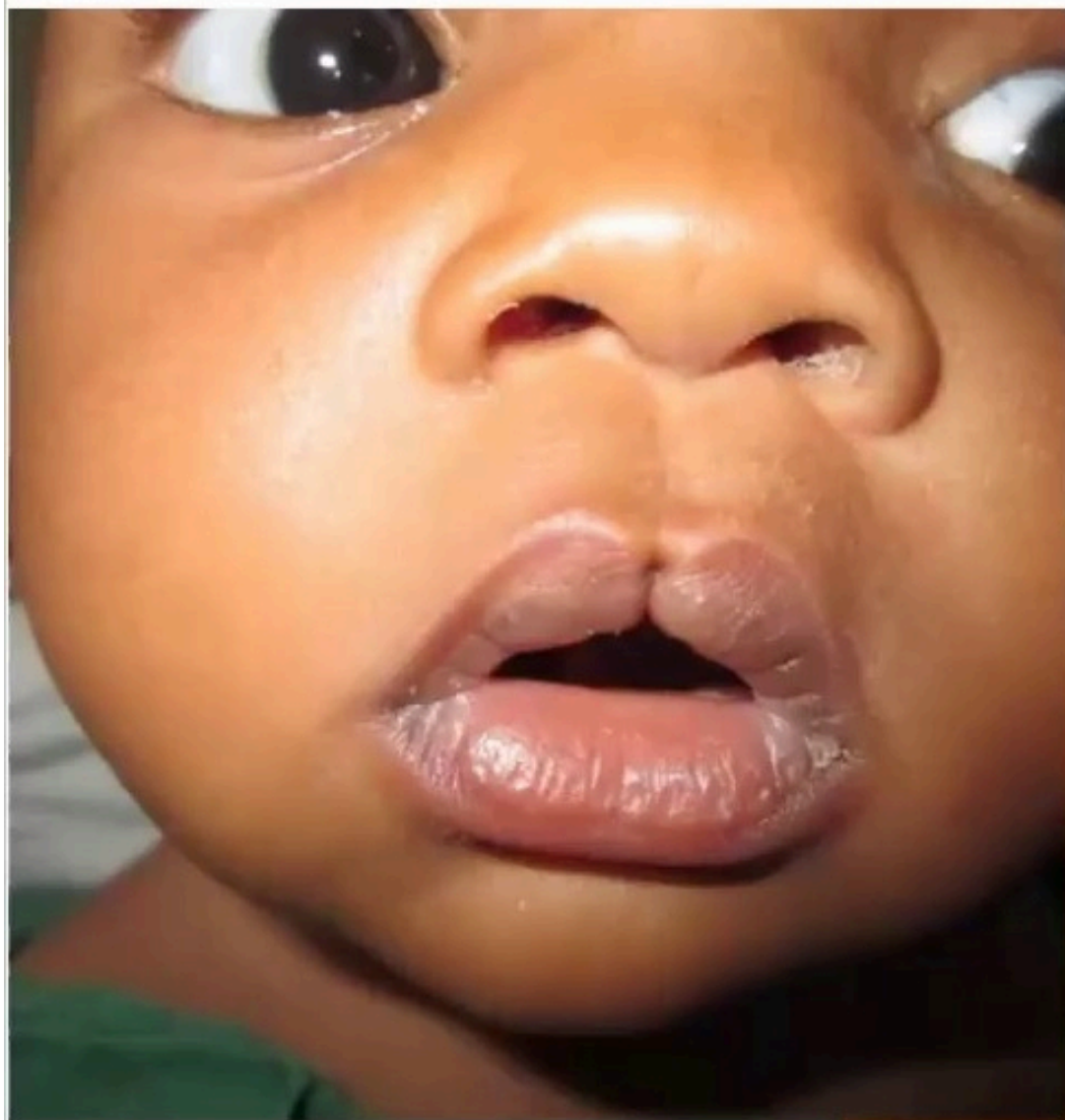
Persistent VPI

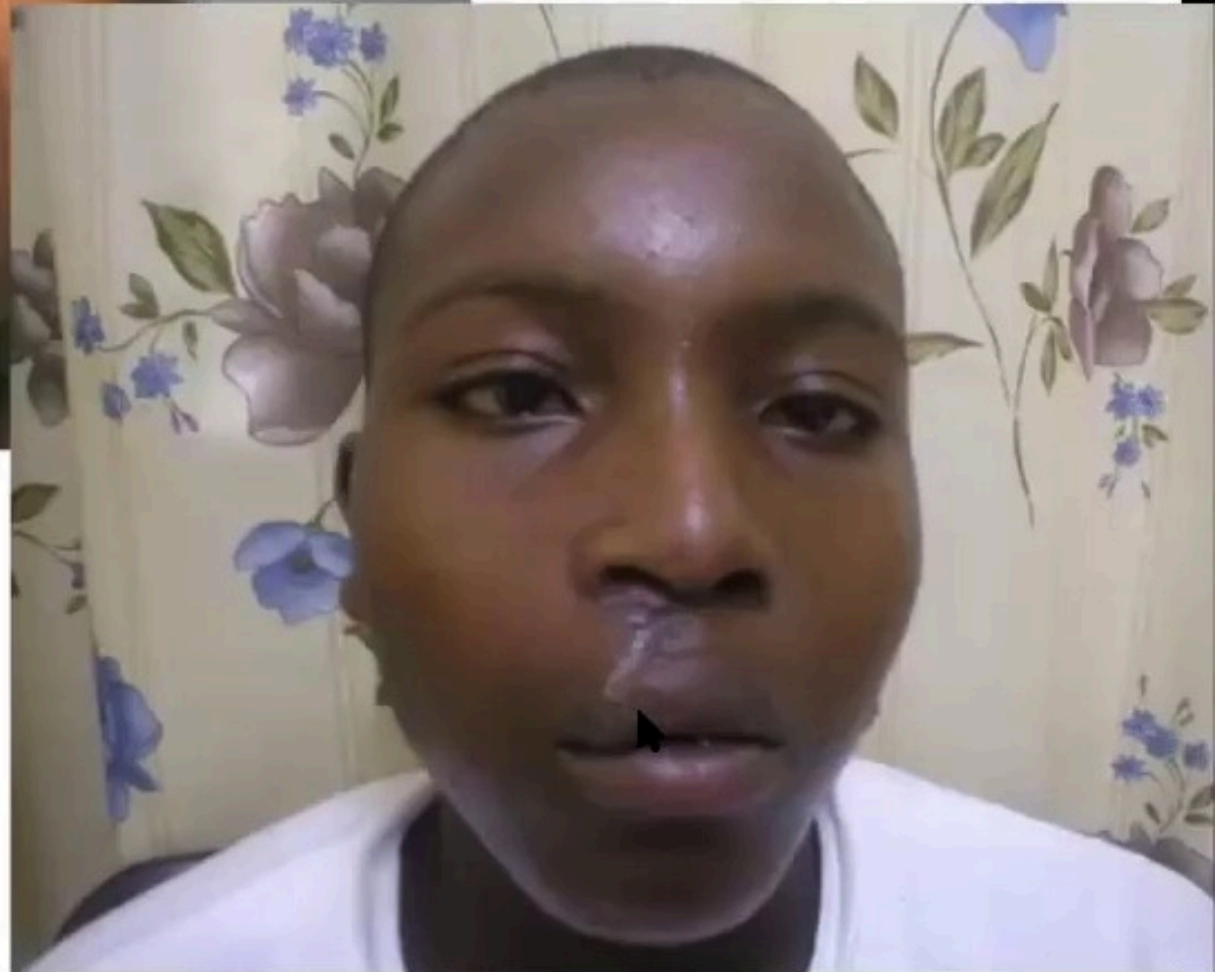
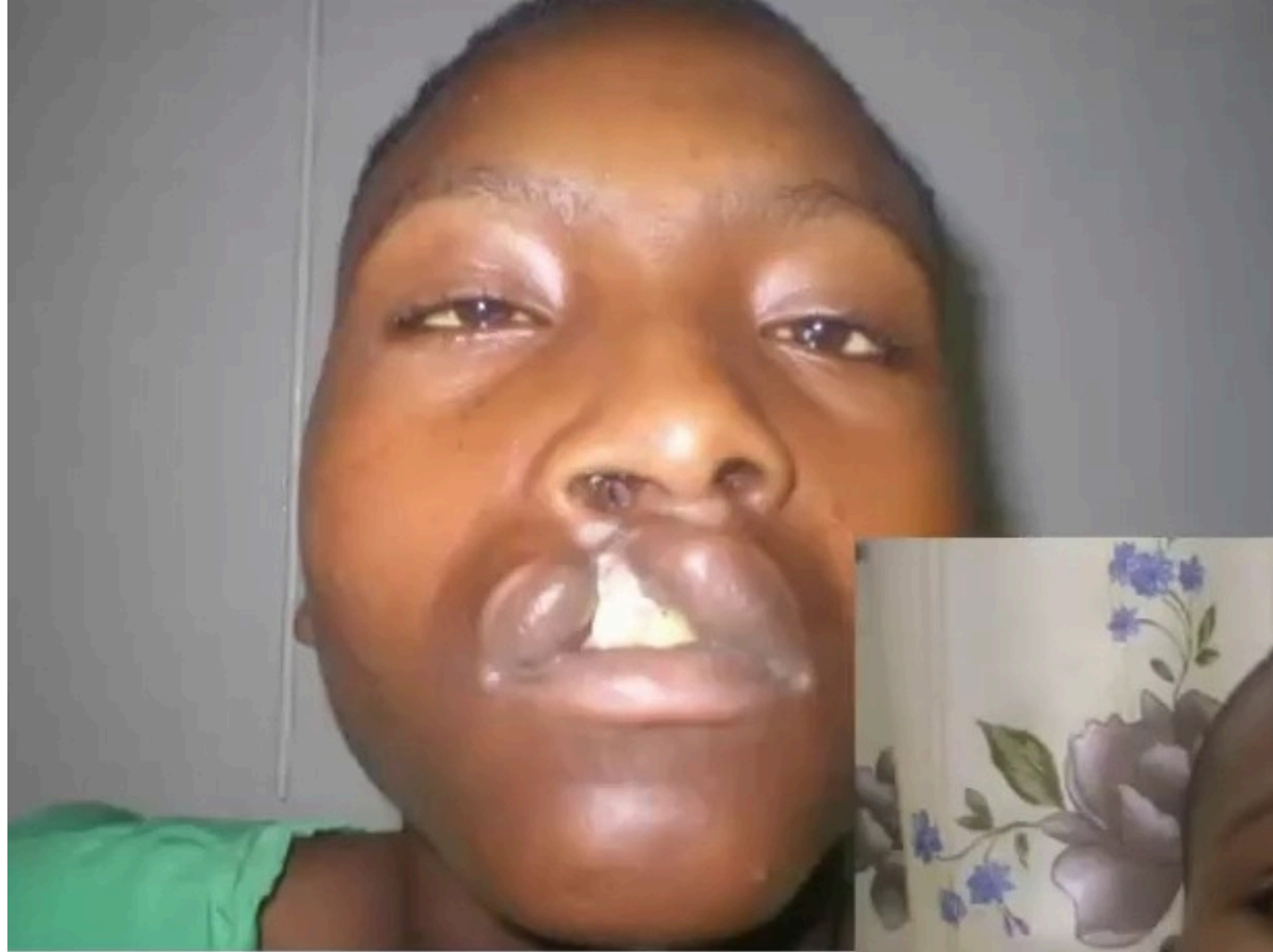
**Under developed facial skeleton/Midfacial growth
problems**

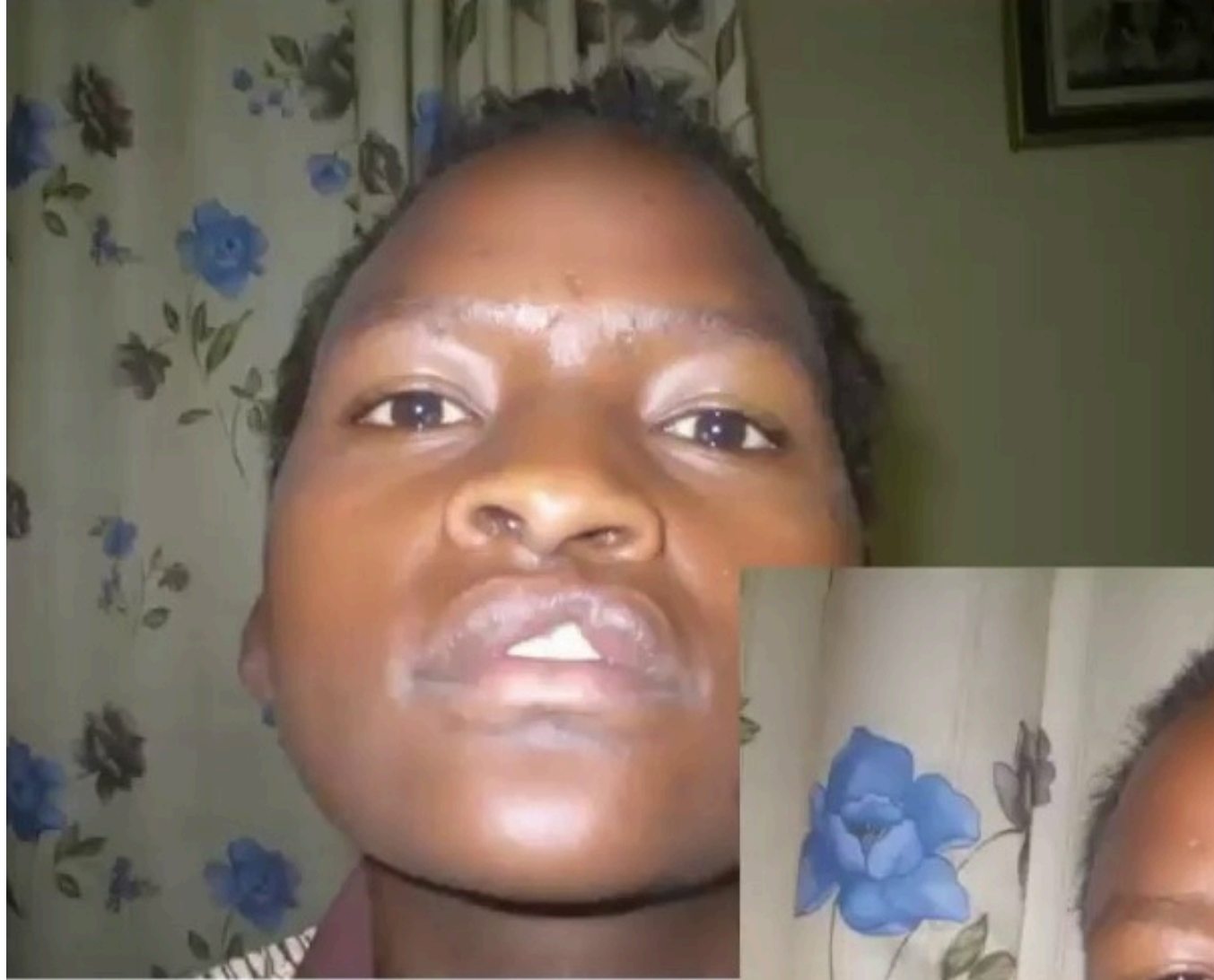
FUTURE

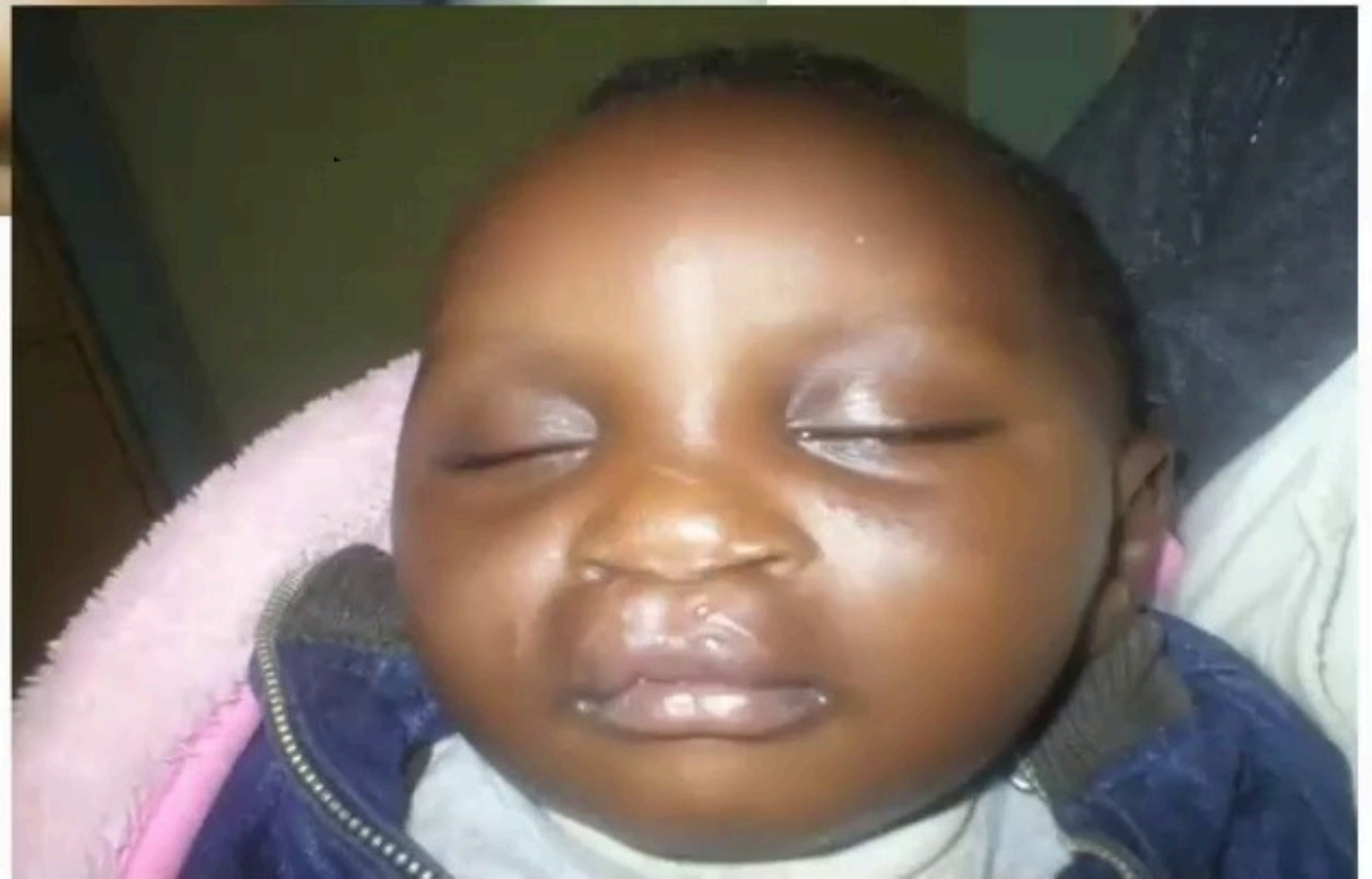
- 1. Vitamin administration –
*periconception***
- 2. High level folic acid during early
pregnancy (3 -5mg/day)**
- 3. Centre Based Care**
- 4. Registry of Congenital
malformations including clefts**

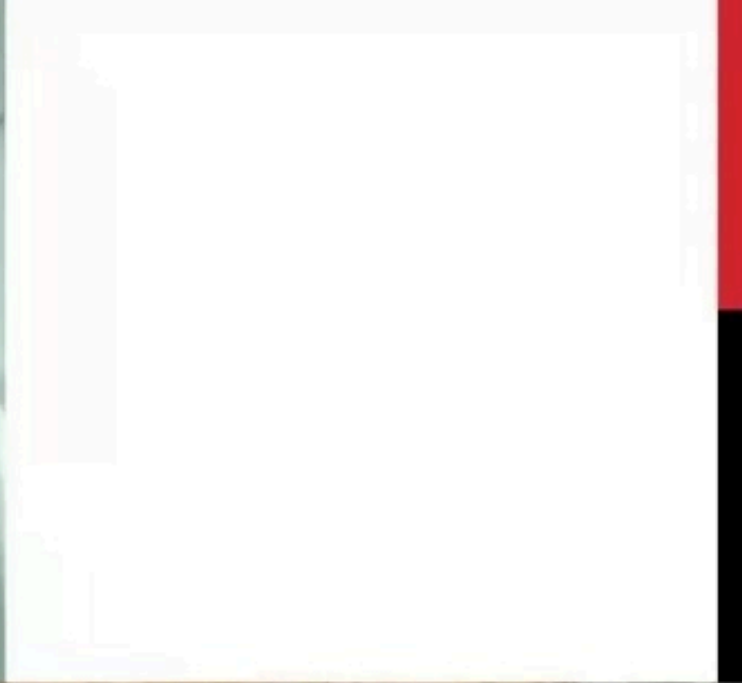












FAMOUS PEOPLE BORN WITH A CLEFT – *HISTORICAL.*

- Tutankhamen Egyptian Pharaoh who may have had a slightly cleft palate according to diagnostic imaging.
- Doc Holliday Dentist, gambler and gunfighter of the American Old West frontier.
- Tad Lincoln Fourth and youngest son of President Abraham Lincoln.
- Thomas Malthus 18th and 19th Century English demographer and political economist.

FAMOUS PEOPLE BORN WITH A CLEFT – *MODERN.*

Carmit Bachar American dancer and singer,

Eric Edgar Cooke Australian criminal, the last person to be hanged in Western Australia,

Jürgen Habermas German philosopher and sociologist,

Wendy Harmer Australian comedian,

Richard Hawley English guitarist, singer, songwriter and producer,

Michael Helm Canadian novelist,

Jesse Jackson American politician, professional civil rights activist and Baptist minister,

Stacy Keach American actor and narrator,

Tim Lott English novelist,

Rita MacNeil Canadian country and folk singer,

Cheech Marin American actor and comedian

Jesse Merz American actor and director

Ljubo Milicevic Australian football (soccer) player,

Nikki Payne Canadian comedian and actress,

Geoff Plant Canadian lawyer and politician, Attorney-General of British Columbia,

Lee Raymond American business executive - former CEO and Chairman of ExxonMobil,

Louis Wain English artist, best known for his drawings featuring anthropomorphised large-eyed cats,

CLEFT CHARITIES

(Top 3 in kenya)

1.



TheSmileTrain

The World's
Leading Cleft
Charity.

2. **Operation Smile**

3. **Cleft Kinder Hilfe**

THE END - THANK YOU

