Cleft Lip and Palate

Robert J. Tibesar MD
Children's Minnesota
ENT/Facial Plastic Surgery
September 27, 2019

Children's MINNESOTA

Disclosures: *I have no conflicts of interest or off-label usage to disclose.*

Children's MINNESOTA

Children's Minnesota Disclaimers and Confidentiality Protections

- Children's makes no representations or warranties about the accuracy, reliability, or completeness of the content. Content is provided "as is" and is for informational use only. It is not a substitute for professional medical advice, diagnosis, or treatment.
- You hold all responsibility for your use or nonuse of the content. Children's shall not be liable for claims, losses, or damages arising from or related to any use or misuse of the content.
- Please note that any peer review information and discussions are privileged and confidential under Minnesota's peer review statute (Minn. Stat. § 145.61 et. seq.) and therefore may not be disclosed unless appropriately authorized.
- Content that is subject to copyright or trademark law requires Children's permission for use.
- Content that includes patient protected health information requires compliance with all applicable state and federal laws protecting patient privacy and security including the Minnesota Health Records Act and the Health Insurance Portability and Accountability Act and its implementing regulations as amended from time to time.
- Please ask if you have any questions about these disclaimers and/or confidentiality protections.

Children's MINNESOTA

Cleft Lip and Palate (CL/P)

- The most common congenital anomaly of the face
- Affects approx. 1 in 700 live births

Children's MINNESOTA

Objectives

- Summarize important background information about CL/P
- Demonstrate surgical repair of CL/P
- Identify CL/P in prenatal U/S images
- Discuss the importance of prenatal counseling for CL/P

Children's MINNESOTA

CL/P Epidemiology: Incidence

Racial differences:

- Native Americans: 3.6/1000 live births
- Asians: 2.1/1000 live births
- Caucasians: 1/1000 live births
- Less in Africans & African Americans: 0.3/1000

Children's MINNESOTA

CL/P Epidemiology: Incidence

- CDC U.S. Annual Estimates: 2,650 babies born with cleft palate and 4,440 babies born with cleft lip +/- cleft palate.
- MN Dept of Health Annual Estimates:
  - 21 babies with cleft lip only
  - 42 babies with cleft lip and palate
  - 39 babies with cleft palate only
  - *=102 babies with some form of CL/P*

Epidemiology: Patterns

- Clefting: 1/3 is palate alone, 2/3 is lip with or without palate
- Bilateral Lip: 85% will have cleft palate
- Unilateral Lip: 70% have cleft palate
- Left side predominance
- Male predominance (1.5 – 2.0)

CL/P Genetics/Etiology

- Not well understood
- Multifactorial (several minor genes + environmental insults are additive)
  - Any disruption of programmed cellular proliferation, migration, and fusion will cause clefting.

Epidemiology: Risk Factors

- Fetal alcohol exposure, maternal drug use, maternal smoking
- maternal illness, infection
- Maternal medications: anticonvulsants, steroids, isotretinoin
- Maternal diabetes
- *Folate reduces risk*

Recurrence Risks

- Recurrence risks: Do not follow typical Mendelian inheritance patterns as seen with single gene conditions.
- 60%-70% of cases are sporadic, no family history whatsoever

importance of prenatal counseling

CL/P Recurrence Risks

-- FAMILIAL RISKS FOR NONSYNDROMIC CLEFT LIP AND PALATE

Family History	Cleft Lip ± Cleft Palate	Cleft Palate
No family history of cleft lip or **cleft palate**	0.1%	0.04%
Unaffected patients with		
One previously affected child	4%	2%
Two previously affected children	9%	1%
One affected parent	4%	6%
One affected parent and one previously affected child	17%	15%
± = with or without		

Associated Syndromes

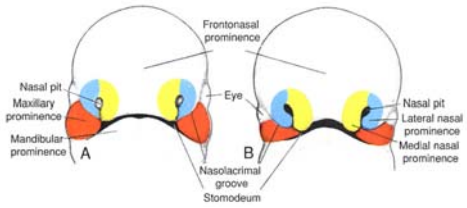
- About 15%-30% of all clefts are associated with a syndrome.
- More than 400 syndromes involve clefting.
  - Trisomy 13, 18
  - Van der Woude's syndrome
  - VCFS (22q microdeletion)
  - Pierre Robin Sequence

Isolated Cleft Palate

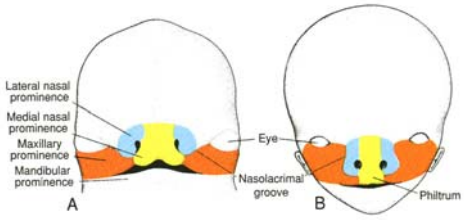
- Distinct entity, separate condition
- Incidence 0.5/1000, no racial differences
- Female predominance (60-80%)
- Higher association with congenital syndromes

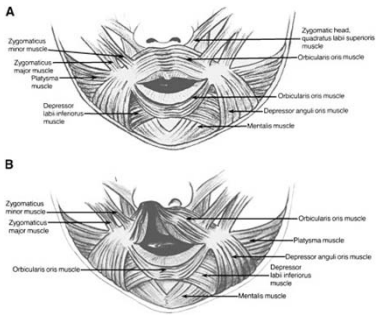
"U" shaped cleft palate "V" shaped cleft palate

Cleft Lip Embryology: 5-7 weeks



Cleft Lip Embryology: 5-7 weeks

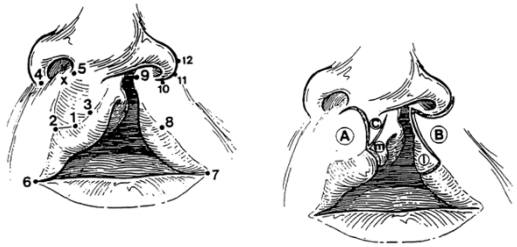




Cleft Lip Repair: Timing

- Old Rule of 10's
  - Hgb 10, 10 lbs, 10 weeks old
- Generally 8-12 weeks old
  - Anesthetic risks
  - Other congenital anomalies
  - Demonstrate feeding and growth
  - Parental bonding

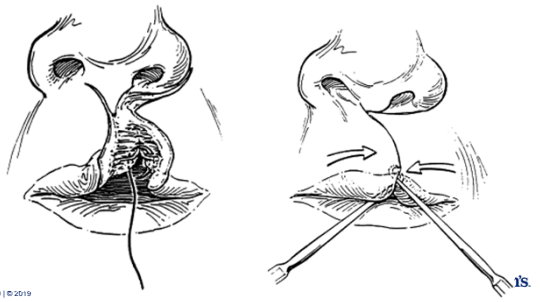
Unilateral Cleft Lip Repair: Rotation Advancement



19 | © 2019

Children's MINNESOTA

Cleft Lip Repair: Rotation Advancement



20 | © 2019

Unilateral incomplete cleft lip:

21 | © 2019

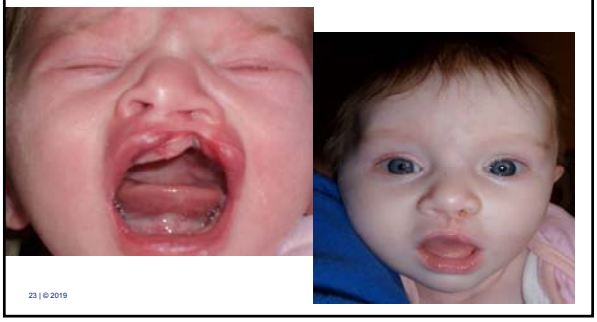
Children's MINNESOTA

Unilateral incomplete cleft lip

22 | © 2019

Children's MINNESOTA

Unilateral incomplete cleft lip



23 | © 2019

Unilateral incomplete cleft lip

24 | © 2019

Children's MINNESOTA

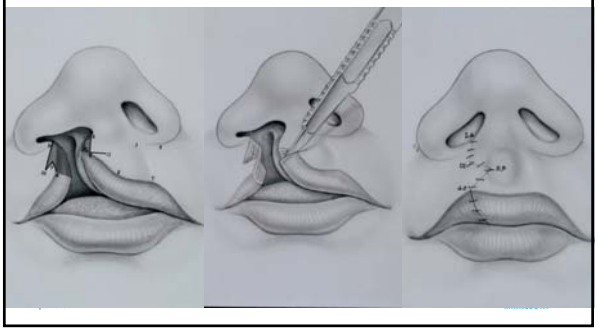
Unilateral complete cleft lip

Children's MINNESOTA

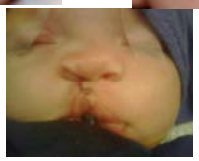
Unilateral complete cleft lip

Children's MINNESOTA

Unilateral Cleft Lip Repair: Triangle Flap



Unilateral CL: Triangle Flap

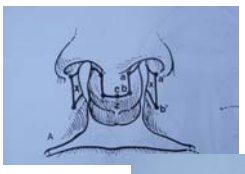


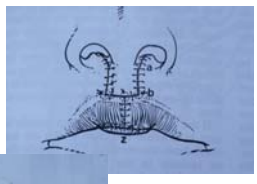
Children's MINNESOTA

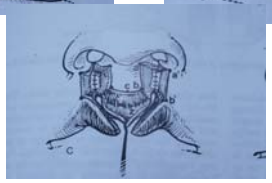
Bilateral cleft lip repair

hildren's MINNESOTA

Bilateral Cleft Lip

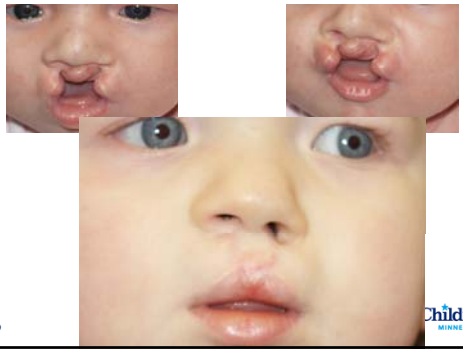






Children's MINNESOTA

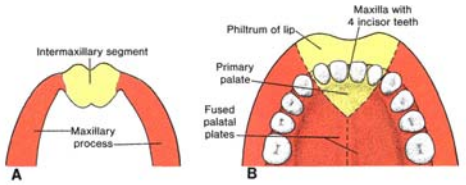
Bilateral Cleft Lip: before and after



Bilateral Cleft Lip: before and after

Cleft Palate Embryology

- Incisive Foramen is key to terminology.
- Primary Palate: Anterior to incisive foramen
  - Consists of lip and premaxilla.

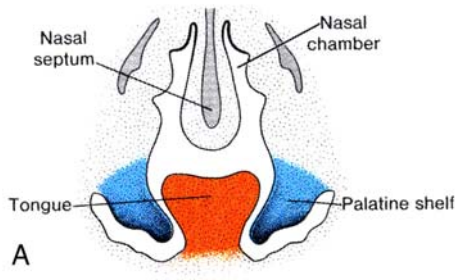


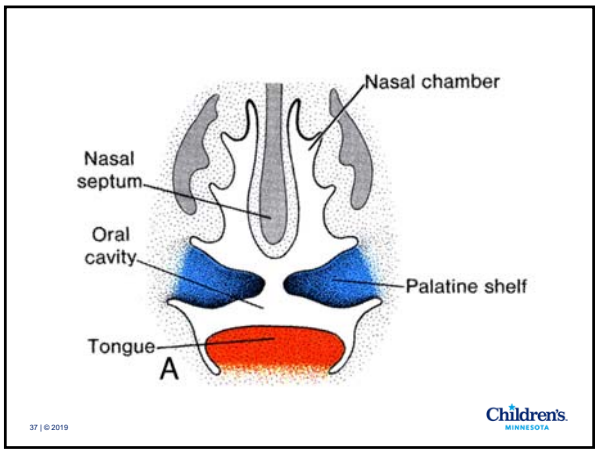
Cleft Palate Embryology

- **Palatogenesis:** 5-12 weeks gestation
  - Primary palate: lip and anterior palate (5-7 weeks)
  - Secondary palate: posterior palate (7-12 weeks)

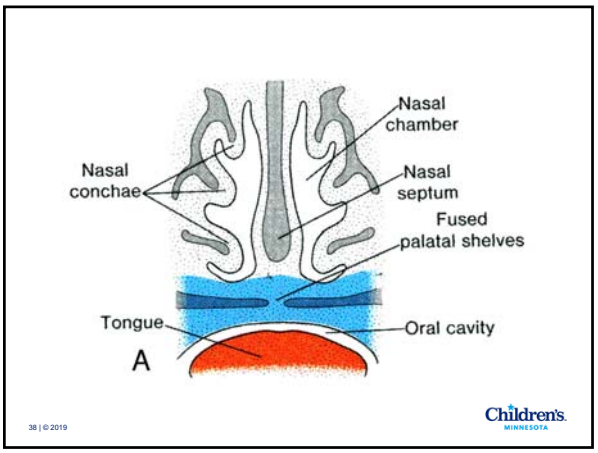
Cleft Palate Embryology

- Secondary Palate: Posterior to incisive foramen to uvula.
- Fusion of bilateral palatal shelves at 7-12 weeks gestation.
- Fuses from anterior to posterior.



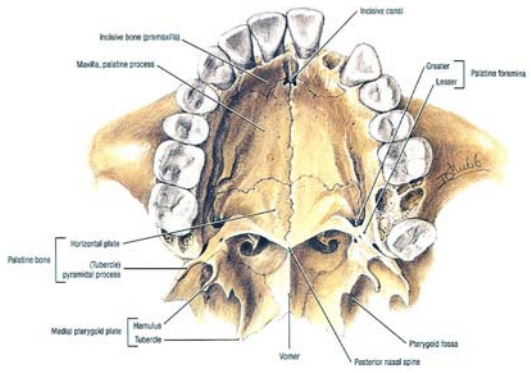


37 | © 2019 Children's MINNESOTA

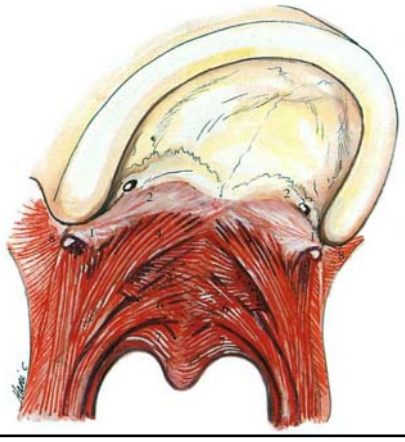


38 | © 2019 Children's MINNESOTA

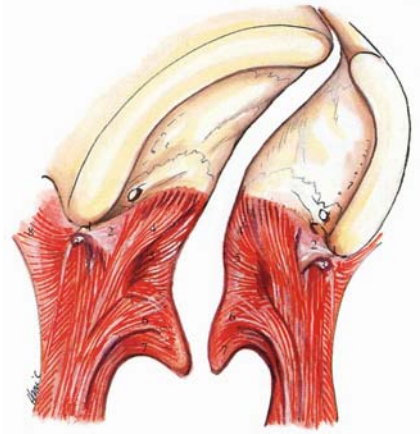
Normal Anatomy



3



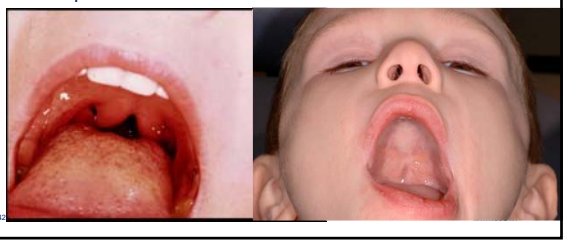
40 | © 2019 ildren's INNESOTA



41 | © 2019 ildren's INNESOTA

Submucous Cleft

- Diastasis of soft palate musculature
  - Bifid uvula, Zona pellucida, Notching of posterior hard palate



Problems Encountered in Cleft Palate

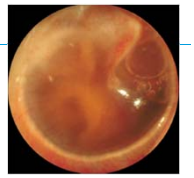
- Feeding difficulties
- Airway compromise if micrognathia & glossoptosis
- Hearing: Eustachian tube dysfunction
- Dental problems
- Speech: 45%-60% will have speech difficulty, velopharyngeal insufficiency

Feeding: infant with Cleft Palate

- Insufficient negative pressure
- Unsuccessful at breast feeding
- Pumped breast milk with special bottle & nipple is best
- Weekly weight checks, detailed feeding log

Eustachian Tube Dysfunction

- Tensor veli palatine muscle dysfunction
- Middle ear fluid, Need PE tubes

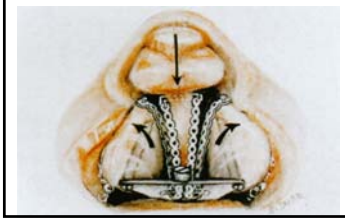


Timing of Cleft Palate Repair

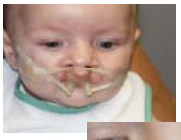
- Timing is controversial
- Early= better speech results and improved eustachian tube function
- Late= better midfacial growth
- Generally, 9-13 months old is optimal

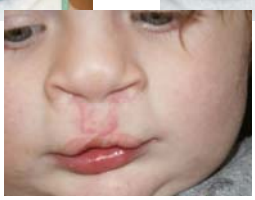
Adjuncts to Surgery

- Pre-surgical orthodontics
  - Re-approximate bony defects
  - Decrease the size of the cleft



Bilateral- Nasal Alveolar Molding





50 | © 2019

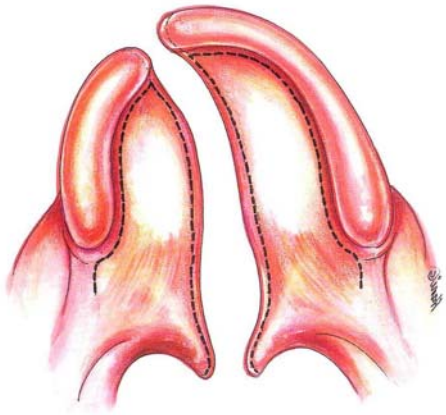
Children's MINNESOTA

Goals of Cleft Palate Surgery

- Reconstitute normal anatomy, separate nose from mouth
- Lengthen soft palate, improve speech
- Overcome eustachian tube dysfunction
- Improve dental occlusion
- Preserve midfacial growth

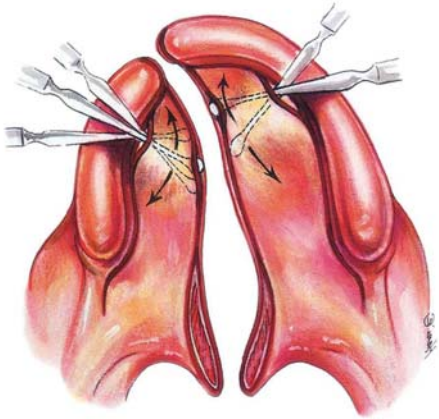
51 | © 2019

Children's MINNESOTA



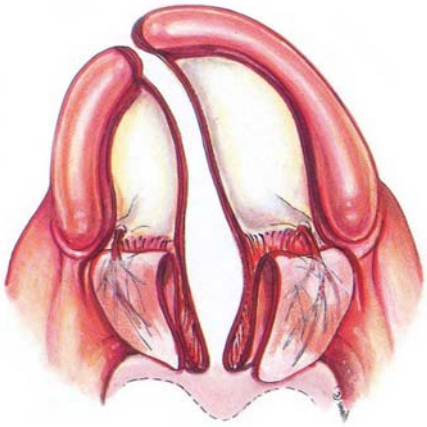
52 | © 201

en's



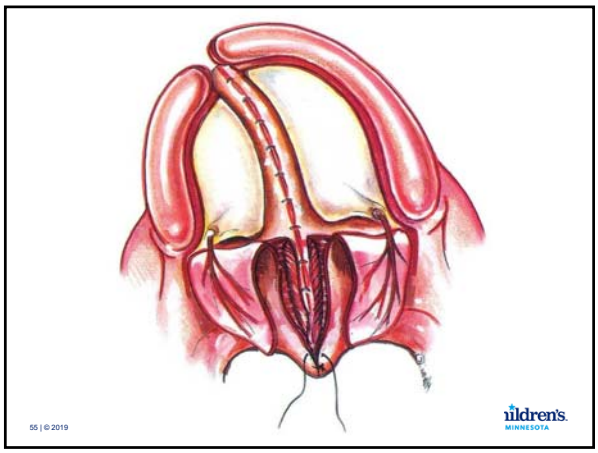
53 | © 201

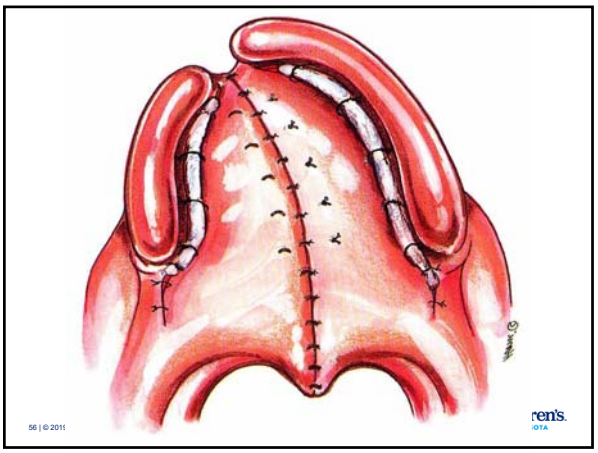
en's



54 | © 2019

lren's





Repaired Cleft Palate

Prenatal Cleft Diagnosis

- First reports of prenatal diagnosis of cleft lip via U/S in 1980's.
- Currently, routine U/S surveillance of the pregnancy includes one mid pregnancy anatomic examination at about 20 to 22 weeks.
  - –possible to detect cleft lip and palate.

Prenatal Cleft Diagnosis

- The ultrasound detection rate for CL/P varies between 17% and 70%
- Ultrasound detection rate increases with the severity of the cleft deformity.
- The ultrasound detection rate has increased with time.
  - Better equipment and technology
  - More experienced sonographers

Prenatal Cleft Diagnosis

- Goals of U/S screening for Cleft lip and palate:
  - detect the defect
  - precisely characterize its extent.
  - determine it's association with other structural anomalies.

Prenatal Cleft Diagnosis

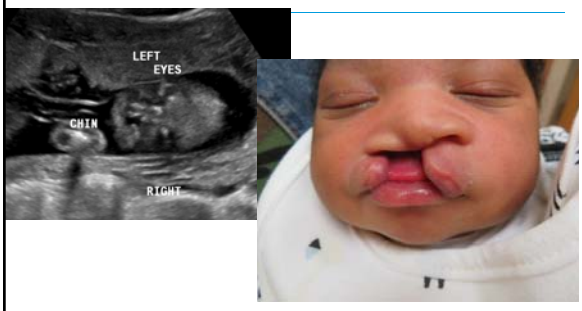
- Detecting cleft palate alone on U/S is difficult
  - MRI to further delineate the cleft defect.
- Clinical utility?
  - Currently there is no need for precise delineation of the cleft.

Prenatal Diagnosis: Fetal Surgery?

Not an option for cleft lip and palate as the risk for the fetus and mother is felt to be too great

Prenatal Diagnosis of Cleft Lip/Palate: Case example: R complete CL/P

Right complete cleft lip and palate



Right complete cleft lip and palate

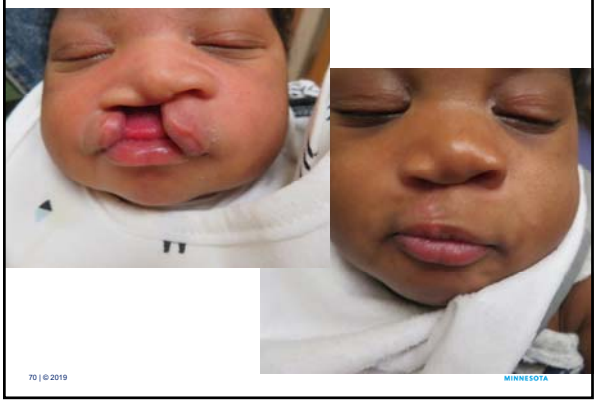
67 | © 2019

Children's MINNESOTA

68 | © 2019

69 | © 2019

Children's MINNESOTA



70 | © 2019

MINNESOTA

Prenatal Diagnosis of Cleft Lip/Palate: Case example: Left incomplete cleft lip and palate

71 | © 2019

Children's MINNESOTA

Prenatal Diagnosis of Cleft Lip/Palate: Case example: Left complete CL/P

72 | © 2019

Children's MINNESOTA

Prenatal Diagnosis of Cleft Lip/Palate: Case example: Left complete CL/P

73 | © 2019

Left cleft lip and palate

75 | © 2019

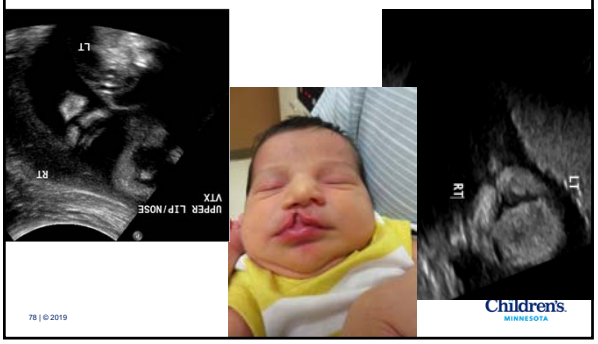
Right incomplete cleft lip

76 | © 2019

Right incomplete cleft lip

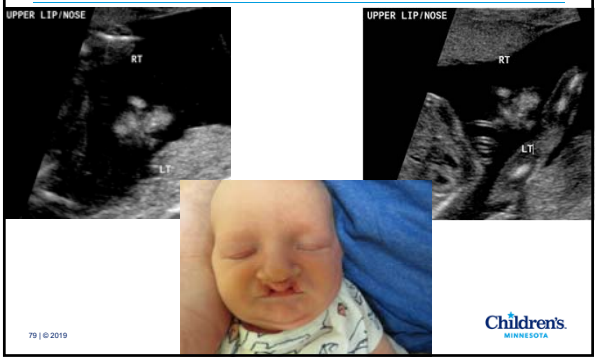
77 | © 2019

Right incomplete cleft lip



78 | © 2019

Bilateral cleft lip



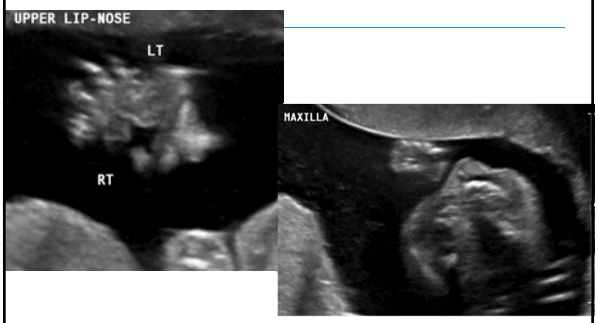
79 | © 2019

Children's MINNESOTA

80 | © 2019

Children's MINNESOTA

Right complete Cleft lip and palate



81 | © 2019

Children's MINNESOTA

82 | © 2019

Children's MINNESOTA

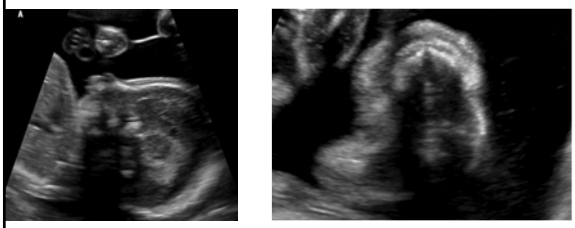
Right complete Cleft lip and palate

83 | © 2019

Children's MINNESOTA

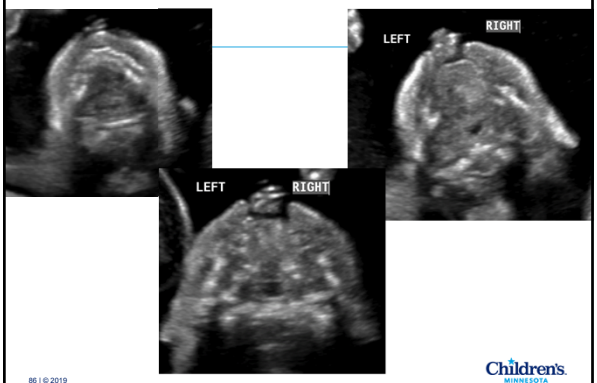
84 | © 2019

Bilateral cleft lip and alveolus likely palate



Children's MINNESOTA

Bilateral cleft lip and alveolus likely palate



Children's MINNESOTA

Prenatal Diagnosis

- Potential Negative: increased parental anxiety
- Potential Benefits
  - Allows further diagnostic evaluation
  - Prepare for the birth and feeding a newborn with cleft palate.
  - Prenatal counseling and teaching parents about cleft lip and palate.

Children's MINNESOTA

Prenatal Counseling

- Remember, the majority of cleft babies are born to families who do not have a history of cleft.
- Lack of knowledge about CL/P in the general public
- ***There is a need for prenatal counseling***

Children's MINNESOTA

Prenatal Counseling

Parental emotions: anxiety, distress, disappointment, fear, anger, confusion, feelings of helplessness

Parents are concerned about the future of their child.

Children's MINNESOTA

Prenatal Counseling

- Most moms would have a sense of guilt and want to blame themselves for their child's deformity.
- Many parents want to know what caused the cleft and why this happened.

Children's MINNESOTA

Prenatal Counseling

- Studies show that most families wish they did receive a prenatal diagnosis.
  - birth experience would be less traumatic
  - time to learn & psychologically prepare
- Vast majority of families say they never contemplated termination of the pregnancy based on the prenatal diagnosis of CL/P

What are the goals of the prenatal consultation?

Ease the parental anxiety and emotional distress

Offer reassurance that CL/P is very treatable.

Present a positive attitude to restore excitement and joy to the pregnancy.

Prenatal Counseling

- Meet with families in timely fashion
- Introduce them to team members
- Spend 60 minutes discussing CL/P

Team Members

- Audiologist
- Otolaryngologist/Cleft Surgeon
- Pediatrician/Team Coordinator
- Geneticist
- Oral Surgeon
- Orthodontist
- Prosthodontist
- Pediatric Dentist
- Psychologist
- Social Worker
- Speech Therapist

Team care

Prenatal Counseling

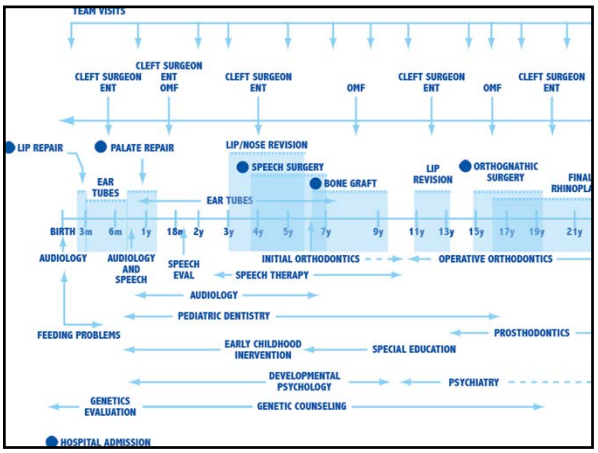
- Review everything from labor and delivery up through college years
- Where to delivery the baby?
- How to feed the baby?
- Hearing, Speech, Language
- Future surgeries

Prenatal Counseling

- Cleft lip and palate surgeries
  - Cleft lip repair around three months of age.
  - Cleft palate repair around one year of age.

97 | © 2019

Children's MINNESOTA



Goal for the child: to be a normal person who happens to have a cleft lip/palate

99 | © 2019

Children's MINNESOTA

Thank you for your time and attention

100 | © 2019

Children's MINNESOTA

101 | © 2019

Children's MINNESOTA