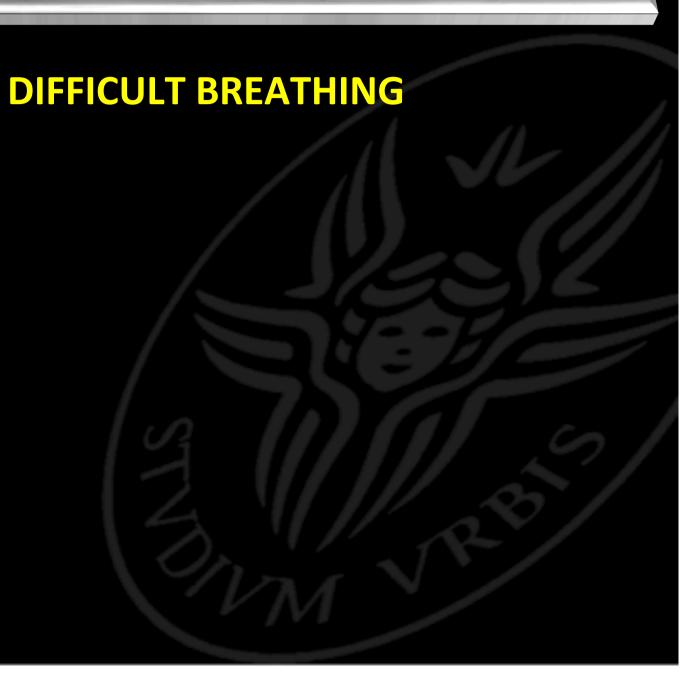
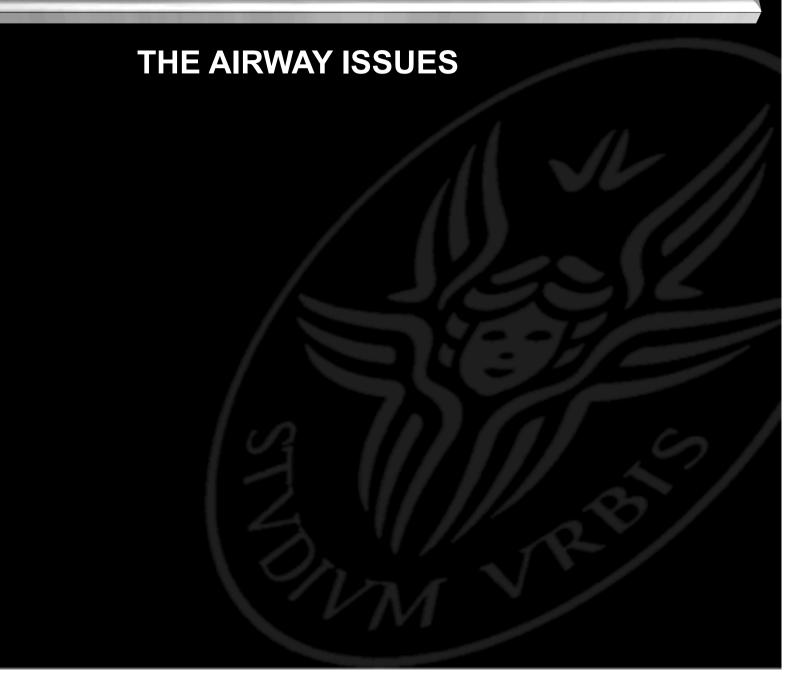
# Pierre Robin Sequence: treatment protocol at "Sapienza" University of Roma

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- Mandibular hypoplasia
- Glossoptosis
- ± Palate cleft





#### **PATHOGENIC MECHANISM**

**Abnormal position of the tongue** 

Mandibular hypoplasia



Glossoptosis

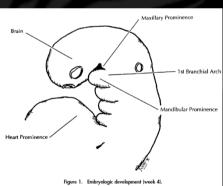
**Cleft palate** 











# TREATMENT GOALS

Life support:

- 1) Breathing
- 2) Feeding

Cleft palate closure

Maxillo-mandibular proper growth

#### **TREATMENT**

# Respiratory problems:

International literature provides many treatment options

- 1) Prone position
- 2) O<sub>2</sub> terapy
- 3) CPAP
- 4) Intubation
- 5) Tongue lip adhesion
- 6) Mandible distraction
- 7) Tracheotomy

## **TREATMENT**

# Feeding problems:

International literature provides different treatment options

- 1) Palatal plate
- 2) Gavage Nasogastric tube
- 3) Percutaneous endoscopic gastrostomy

## **TREATMENT**

At the present time there are no recognised international guidelines for the management of PRS

# **TREATMENT**

The priority in severe PRS is to resolve respiratory and feeding problems, ensuring normal growth

Eleven patients with severe Pierre Robin sequence.

All patients had mandibular hypoplasia, glossoptosis and palate cleft.

All patients had severe respiratory distress after delivery.

Our treatment protocol in severe Pierre Robin Sequence differs from others.



#### **OUR PIVOTS**

1) MANDIBULAR HYPOPLASIA: DISTRACTION OSTEOGENESIS

2) CLEFT PALATE:
ORTHOPAEDIC PLATE

3) CLOSURE OF CLEFT PALATE
EARLY CLOSURE

## **MANDIBULAR DISTRACTION**

#### International literature provides different treatment options

- McCarthy JG, Schreiber J, Karp N, et al. Lengthening the human mandible by gradual distraction. Plast Reconstr Surg 1992;89:1Y8
- Cohen S, Simms C, Burstein F. Mandibular distraction osteogenesis in the treatment of upper airway obstruction in children with craniofacial deformities. Plast Reconstr Surg 1998;101:312Y318
- A.D. Denny, R. Talisman, P.R. Hanson, R.F. Recinos, Mandibular distraction osteogenesis in very young patients to correct airway obstruction, Plast. Reconstr. Surg. 108 (2) (2001) 302—311.
- A. Denny, B. Kalantarian, Mandibular distraction in neonates: a strategy to avoid tracheostomy, Plast. Reconstr. Surg. 109 (3) (2002) 896—904.
- F.O. Monasterio, M. Drucker, F. Molina, A. Ysunza, Distraction osteogenesis in Pierre Robin Sequence and related respiratory problems in children, J. Craniofac. Surg. 13 (1) (2002) 79—83.
- F.O. Monasterio, F. Molina, F. Berlanga, et al., Swallowing disorders in Pierre Robin sequence: its correction by distraction, J. Craniofac. Surg. 15 (6) (2004) 934—941.
- R.B. Schaefer, J.A. Stadler, A.K. Gosain, To distract or not to distract: an algorithm for airway management in isolated Pierre Robin sequence, Plast. Reconstr. Surg. 113 (4) (2004) 1113—1125.
- F.D. Burstein, J.K. Williams, Mandibular distraction osteogenesis in Pierre Robin sequence: application of a new internal single-stage resorbable device, Plast. Reconstr. Surg. 115 (1) (2005) 61—67.
- Dauria D, Marsh JL. Mandibular distraction osteogenesis for Pierre Robin sequence: what percentage of neonates need it? J Craniofac Surg 2008;19:1237Y1243

# **MANDIBULAR DISTRACTION**

External mandibular distractor

Intraoral approach

Complete osteotomies with piezosurgery

Intraoperative activation (2mm)

Active distraction 3 days after surgery (1,5-2mm/day)

Callus consolidation period 4 weeks

Distractors removed without anesthesia

## **RESULTS**

- 11 patients with severe PRS underwent mandibular distraction
- Age at surgery varied from 12 to 100 days (mean 28 days)
- Duration of distraction ranged from 8 to 14 days (mean 10 days)
- Amount of distraction varied from 12mm to 22mm (mean 14mm)

# **MANDIBULAR DISTRACTION GOALS**

Advances the base of the tongue

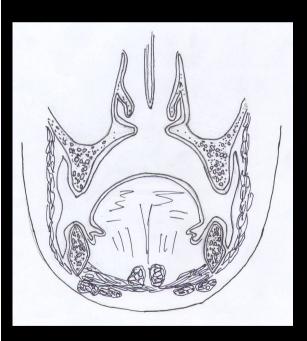
Enlarges the hypopharyngeal space

Eliminate airway obstruction

Avoid tracheotomy

# **ORTHOPAEDIC PLATE**

# Our palatal plate is not an obturator!

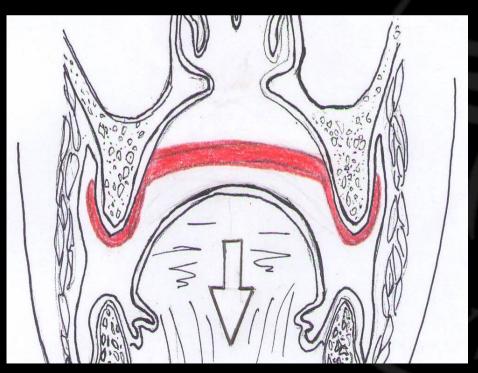


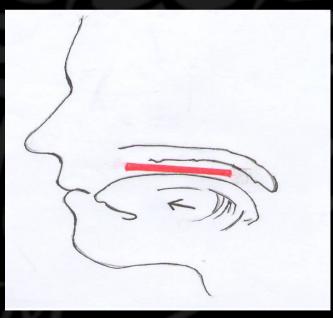




# **ORTHOPAEDIC PLATE**

It is an orthopedic device which is used to flatten the palatine vault to reposition the tongue in order to stimulate mandibular protrusion.





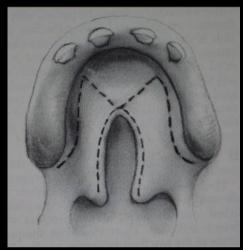
## **ORTHOPAEDIC PLATE**

- 1. Filling of the cleft
- 2. Redistribution of space in oral cavity with lowering of the palatine vault
- 3. Re-education of tongue posture
- 4. Improved breathing
- 5. Improved nutrition
- 6. Guide for the growth of the palatal processes
- 7. Pre-surgical reduction of the cleft

Leonardo ME, Mantuano MT, Silvestri A. Nuovo inquadramento patogenetico disfunzionale e ruolo della terapia funzionale nella sequela di Pierre Robin.

Ortognatodonzia Italiana 2000; 9-3: 339-344.

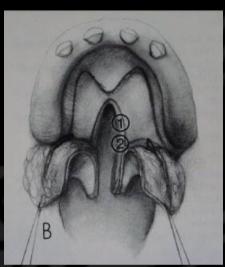
# **EARLY CLEFT PALATE CLOSURE**

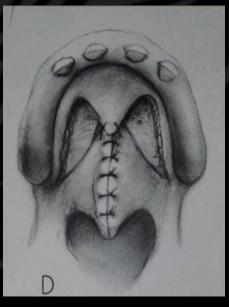


**PUSHBACK TECHNIQUE** 



This procedures flattens the palatal vault that is a desirable goal for these patients





## **PRENATAL DIAGNOSIS**

Prenatal diagnosis is possible from 20<sup>Th</sup> weeks of gestation by ultrasonography or RM.

These is important to give guidance to parents about possible respiratory distress after delivery.



Moshe Bronshtein, Shraga Blazer et all. Ultrasonographic diagnosis of glossoptosis in fetuses with Pierre Robin sequence in early and mid pregnancy American Journal of Obstetrics and Gynecology (2005) 193, 1561–4

