



# A CASE OF DIFFICULT AIRWAY IN A PREMATURE BABY

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## Case Details

- ❖ Name- Baby A, Sex- Male
- ❖ Gestational Age At Birth: 33+5 Weeks, Birth Weight: 1.845kg
- ❖ Admitted in NICU at birth with chief complaints of
  - ✓ Prematurity and low birth weight
  - ✓ Respiratory Distress
  - ✓ Facial anomaly

## Antenatal history

- ❖ 37 years old primigravida, Natural conception,
- ❖ Booked and Immunized
- ❖ HIV, HBsAg, HCV, VDRL, TORCH- negative
- ❖ Serum Screening for Fetal Aneuploidy- low risk
- ❖ TIFFA- No abnormality reported
- ❖ No h/o oligohydromnios
- ❖ No history of cleft palate or any syndrome in the family,
- ❖ Came with preterm labor, ANS partial covered

## Delivery room details

- Born by LSCS ( indication- non progress of labor),
- Baby cried immediately after birth, No resuscitation needed
- Apgar Score at 1 min - 8, 5 min - 9



- ▶ Facial features
  - ▶ Micro-Retrognathia
  - ▶ Cleft palate
  - ▶ Glossoptosis
- Prone nursing and NPA
- ▶ Shifted to NICU

## Examination

- Heart rate- 142/ min, All Peripheral Pulses Palpable
- Respiratory rate- 70/min, intercostal retraction present
- Spo2 – 76-80% in room air in supine, 80-85% in prone position. No Significant Pre-Post Ductal SpO<sub>2</sub> Difference
- NIBP- 70/40(53) mmHg RUL, No Significant Difference in Four Limb BP
- Temperature- 36.9degree Cel,

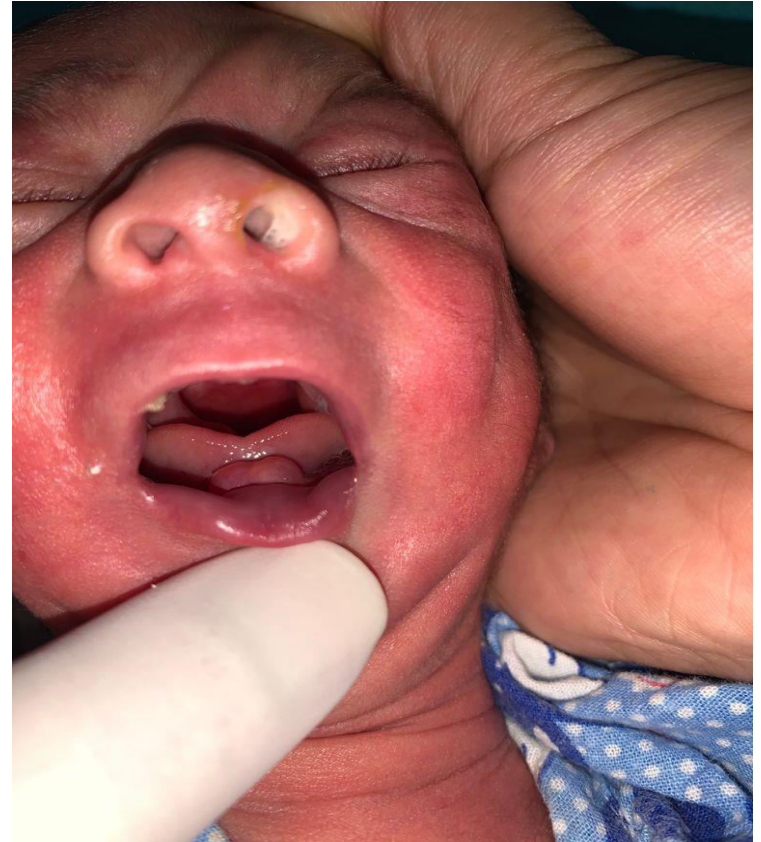
## Anthropometry

(As per fenton's 2013 chart)

- Birth weight -1845 gms  
( 10-50th percentile)
- Length-44cm ( 10-50<sup>th</sup> percentile)
- HC-31.5cm ( 50 -90<sup>th</sup> percentile)

## Head to toe examination

- ❖ Head normal in shape and size
- ❖ Facial anomaly present-as described
- ❖ Chest wall normal
- ❖ Limbs normal
- ❖ Spine and genitalia normal
- ❖ Anal opening patent and normally placed



## Systemic examination

### Respiratory system

- Respiratory distress present, B/L air entry equal and clear
- Silvermann Score-5/10

### Central Nervous System Examination

- Baby was active, alert
- Cry, Tone and Activity -- Normal
- Neonatal reflexes- Normal

### Per Abdomen Examination

- Soft, No Organomegaly

### Cardiovascular System

- S1,S2 normal
- No murmurs

## CASE PROGRESION IN NICU- Day 1

- ❑ Baby had respiratory distress: Desaturations upto 76% in room air, Retractions++
- ❑ Not relieved by prone nursing and temporary nasopharyngeal airway
- ❑ Put on CPAP via mask interface (Fio2-21%, PEEP-6) along with Lateral nursing
- ❑ Baby was on tube feeds (EBM)
- ❑ Baby tolerated feeds well, passed urine and stool adequately.

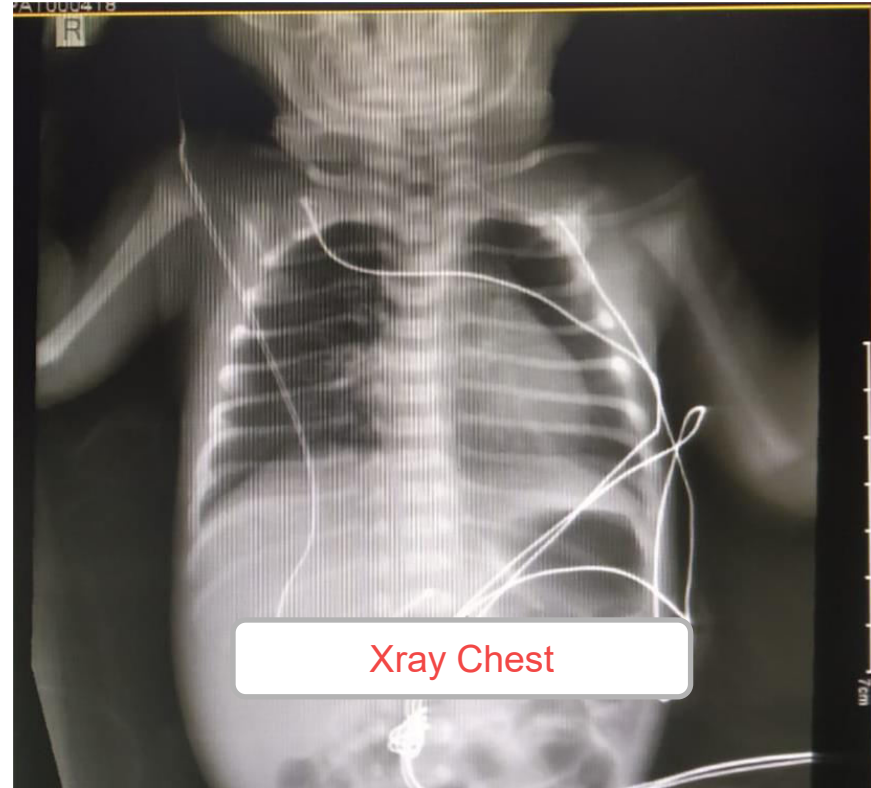


## Differential Diagnosis of Respiratory Distress

- ▶ Prematurity/ RDS
- ▶ Upper airway obstruction
- ▶ Congenital pneumonia
- ▶ Structural lung malformation

## Investigation

- ❑ CBC- Hb -20.7g/dl , TLC- 21,500/ mm<sup>3</sup> PCV-61%, Platelets-2,52,000/ mm<sup>3</sup>
- ❑ CRP- < 3mg/L
- ❑ USG Cranium and KUB- WNL
- ❑ ECHO- No structural cardiac lesion, mild PAH
- ❑ Arterial Blood gas- pH-7.39, pCO<sub>2</sub>-43.6mmHg, pO<sub>2</sub>-40mmHg, Lact-0.6 mmol/L, HCO<sub>3</sub> -22.9mmol/l



## CASE PROGRESION IN NICU-

### Day 2

- Baby on CPAP (FiO<sub>2</sub>-21%, PEEP-5) maintaining saturation on this settings.
- On tube feeds which tolerated
- Antireflux drugs started

### Day 3

- Baby continued on CPAP with PEEP-5, 21%Fio<sub>2</sub>, maintain saturation
- Tried to Wean off but distress increased, so again put on CPAP
- Repeat Xray was normal

## CASE PROGRESION IN NICU-

### Day 4

- ▶ Baby was completely comfortable on the minimal settings, but unable to wean off from CPAP
- ▶ No distress on proning
- ▶ Cause??
- ▶ Upper airway obstruction >> Pulmonary cause
  
- ▶ Decision for upper airway management

### Challenges

- Option for Upper airway management
  - Prone nursing
  - Nasopharyngeal airway
  - Glossoplexy
  - Tracheostomy

## CASE PROGRESSION IN NICU- Day 5

- ▶ Nasopharyngeal airway from modified ET tube placed

**CPAP weaned off Post NPT placement**  
**Could be nursed in supine position**  
**ABG- No hypercarbia**

## Case Summary

- ▶ Preterm, appropriate for gestational age baby boy, presenting at birth with syndromic appearance and respiratory distress

Preterm /AGA/Pierre Robin Sequence/RDS/Upper airway obstruction

## Discharge Planning

- ▶ Discharged on NPT on day 10
  - ▶ OG Tube Feeding
  - ▶ Home nurse staff
  - ▶ Antireflux medications
- ▶ Planned for corrective surgery
- ▶ Distraction osteogenesis of mandible considered
- ▶ Advised for airway evaluation on follow up



## Current Status

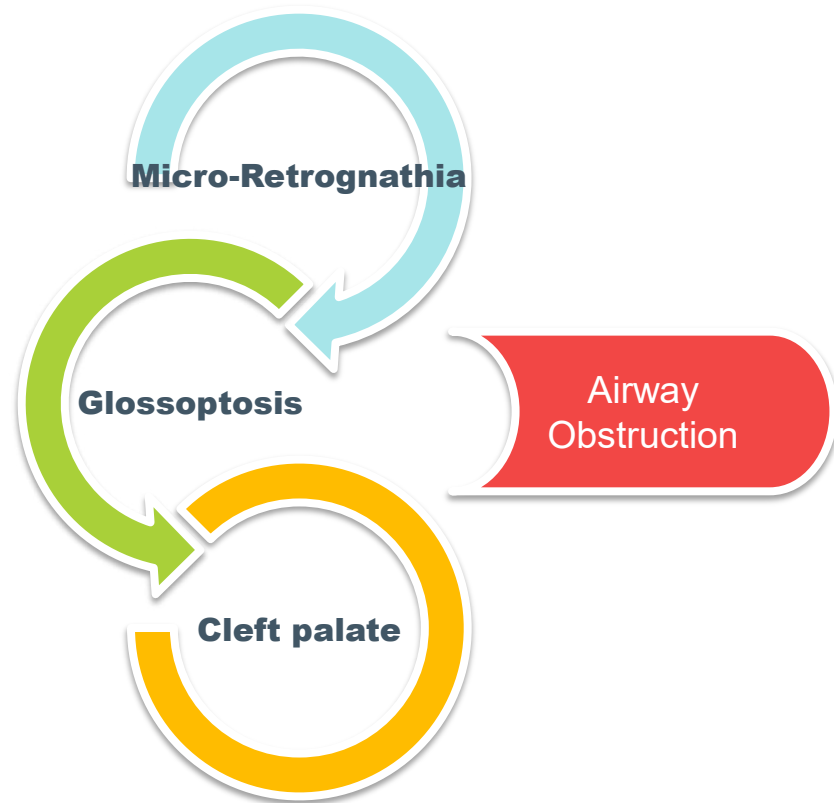
- ▶ Age- 4 months
- ▶ NPT discontinued at 3 month
- ▶ Growing adequately
- ▶ Feeding by OG tube
- ▶ No apparent life threatening event

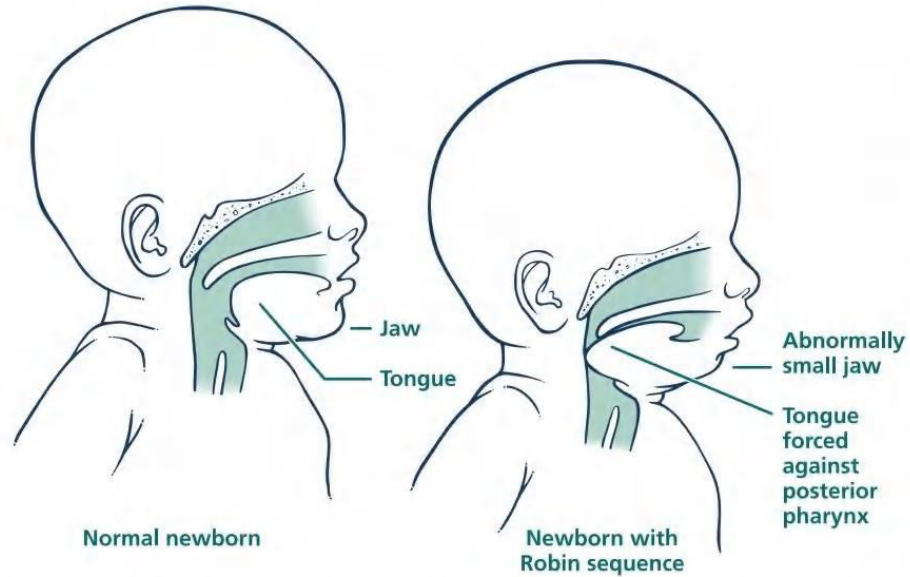
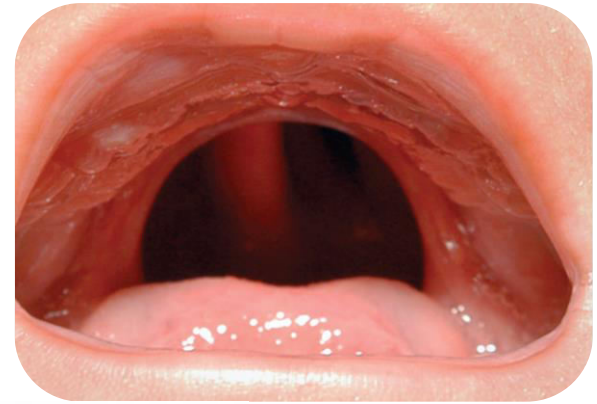


# Review of Literature

## Pierre Robin Sequence

- ▶ Described by Pierre Robbins
- ▶ As Sequence of events
- ▶ an occurrence around 1:8500 to 14,000 at birth
- ▶ Isolated RS or associated with other anomalies





## Etiopathogenesis

- ▶ Multiple theories proposed

Theory	Mechanism	Catch up growth
Neuromuscular abnormalities theory	A neuromuscular insufficiency inhibiting intra-uterine mandibular motion	Unlikely
Deformational theory	Mandible is intrinsically normal, but deformed by external factors. <b>ISOLATED RS</b>	Possible
Malformational theory	Primary failure of mandibular growth caused by a genetic defect.- SOX9 gene mutation <b>Syndromic RS</b>	Unlikely

## Clinical Associations

Type of RS	Description	Features
Isolated RS		
Syndromic RS	Stickler Syndrome	Cataract, Retinal detachment; Hearing loss Midfacial underdevelopment
	Treacher Collins Syndrome	Malar hypoplasia, Micrognathia, External ear abnormalities
	Marshall Syndrome	Midfacial hypoplasia, cleft palate, high myopia and cataracts, short stature
	Velocardiofacial syndrome	Developmental delay, cardiac anomalies, palatal anomalies, and immune deficiency
Associated Anomalies	Hypertelorism, Microtia, long and/or broad philtrum, frontal bossing, psychomotor retardation, ear malformations and hand malformations.	

## Clinical Associations Evidence

- ▶ M Holder-Espinasse et al: Out of 117 cases, isolated (48%), syndromic (35%), and with associated anomalies (17%)

Holder-Espinasse M et al J Pediatr. 2001  
Oct;139(4):588-90

- ▶ Izumi K, Konczal LL et al: Out of 125 cases, isolated (42%), syndromic (58%), Stickler syndrome 22%, Marshall Syndrome 3%

Izumi K, Konczal LL et al J Pediatr. 2012 Apr;160(4):645-650.e2.

## Clinical Presentation

### General

- Microretrognathia, Cleft palate
- SGA

### Resp

- Upper Respiratory tract obstruction
- Obstructive sleep apnea

### GIT

- Gastro esophageal reflux, Aspiration
- Feeding Difficulty, Failure to thrive

### Misc

- Anomalies of the musculoskeletal system, Recurrent otitis media, Natal teeth
- Pulmonary Stenosis, PPHN

## Diagnostic Evaluation

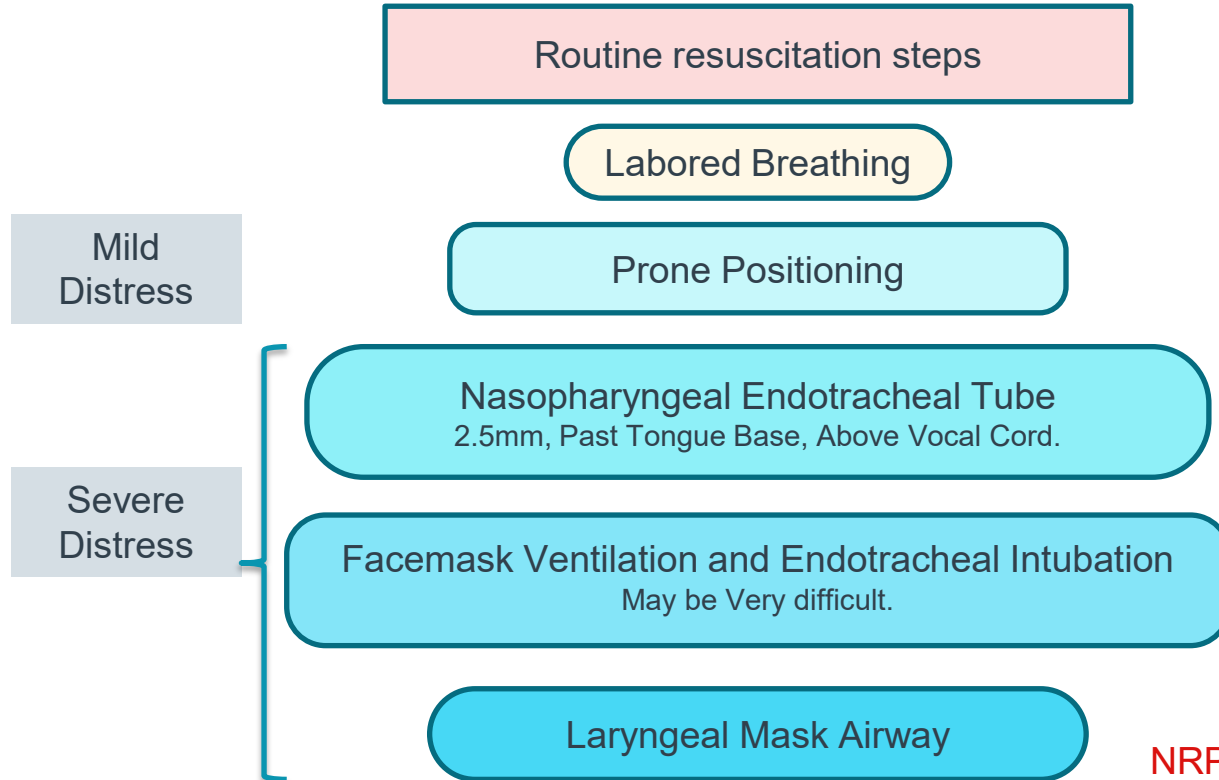
- ▶ Physical examination
- ▶ 2D and 3D photograph assessment
- ▶ Xray Chest and Infantogram
- ▶ Echocardiography, USG cranium, USG KUB
- ▶ ABG/Serum Bicarbonates
- ▶ Polysomnography, and Oximetry
- ▶ Airway evaluation-
  - ▶ Direct laryngoscopy
  - ▶ Bronchoscopy
- ▶ Genetic Testing



## Management

- ▶ Principle of Management :
  - ▶ Acute stabilization- Airway and Feeding
  - ▶ Definitive surgery: MDO, Cleft palate Repair
  - ▶ Management of associated factors: GE Reflux

# Delivery Room Management



# Airway Management in Neonatal Period

## Non Surgical Methods

### PRONE POSITION

- Success rate: 49-77%
- Mild, intermittent airway obstruction



### NASOPHARYNGEAL AIRWAY

- Success rate: 26-100%
- Single level of obstruction at tongue base



### ENDOTRACHEAL INTUBATION

- Success rate: 43%
- Temporary
- Technical difficulty



## Surgical Methods

### TONGUE LIP ADHESION

- Success rate: 33-100%
- Feeding issues
- Speech defects on long term



### MANDIBULAR DISTRACTION OSTEOGENESIS

- Success rate: 88-100%
- Injury to Bone/Permanent teeth, Nerve Damage



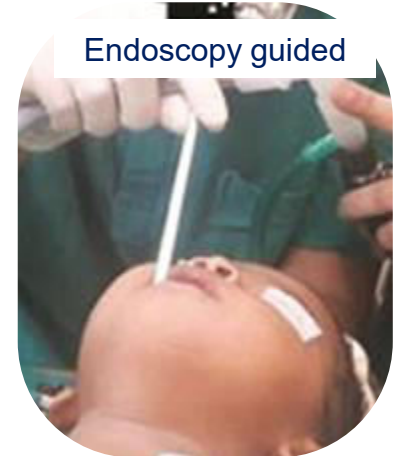
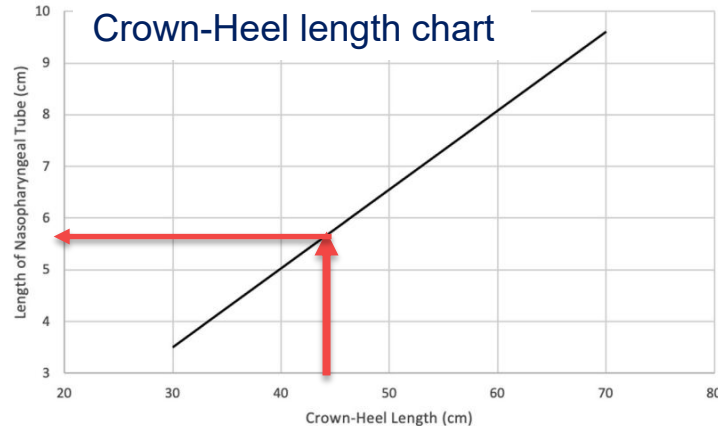
### TRACHEOTOMY

- Success rate: 100%
- For Multilevel airway obstruction
- Complications and Follow up care



# Nasopharyngeal Airway

- ▶ Indication:
  - ▶ Moderate/severe obstruction who failed conservative approaches
  - ▶ As a temporary solution while awaiting mandibular growth to avoid surgery
- ▶ **Size:** Widest that the nostril will accommodate without inducing tissue damage.
- ▶ **Length:**
  - ☒ length that ends just proximal to or above the epiglottis
  - ☒ but extends beyond the tongue base



## Preparation of Nasopharyngeal Airway

- Materials: 3 mm ET tube, Tegaderm, Dynaplast
- Choosing length: From Crown Heel Length chart + 4 cm
- Preparing wings
- Fixing of wings

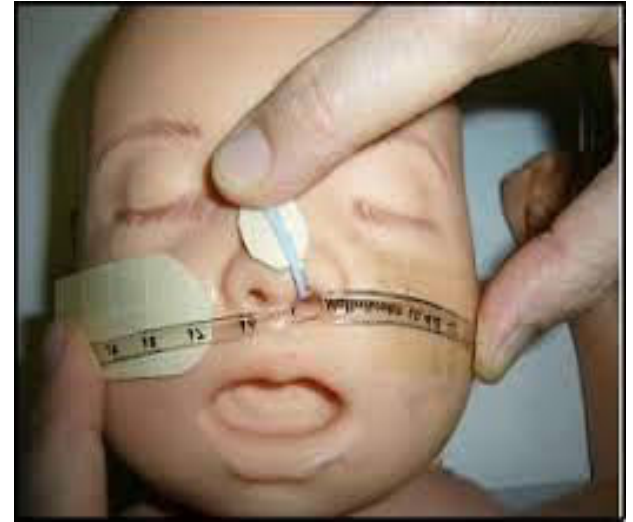
## Nasopharyngeal Airway Usage in Practice



ET tube Modification  
and tube holder



ET tube Modification  
without tube holder



ET tube with three wing split

## Monitoring and Follow up on NPA

### Care at Home

- ▶ Monitor respiratory rate/effort
- ▶ Humidification
- ▶ Regular suction
- ▶ See for skin erosion

### Follow up and weaning

- ▶ Multidisciplinary Team
- ▶ Pediatrician, Plastic Surgeon,  
Physiotherapist
- ▶ Reassess for weaning under  
admission with or without  
sleep studies

## The successful use of the nasopharyngeal airway in Pierre Robin sequence: an 11-year experience

Francois Abel,<sup>1</sup> Yogesh Bajaj,<sup>2</sup> Michelle Wyatt,<sup>2</sup> Colin Wallis<sup>1</sup>

- ▶ Abel et al, Great Ormond Street Hospital, UK, 2000-2010
- ▶ 104 PRS patients; Airway symptoms were managed
  - ☒ Conservatively in 27 patients (25.9%),
  - ☒ Use of NPA in 63 (60.6%)
  - ☒ Tracheostomy in 14 (13.4%)
- ▶ Average duration of NPA use was 8 months (3 weeks to 27 months)
- ▶ NPA improved Polysomnography results in all babies where used.



## Non invasive Ventilation in RS

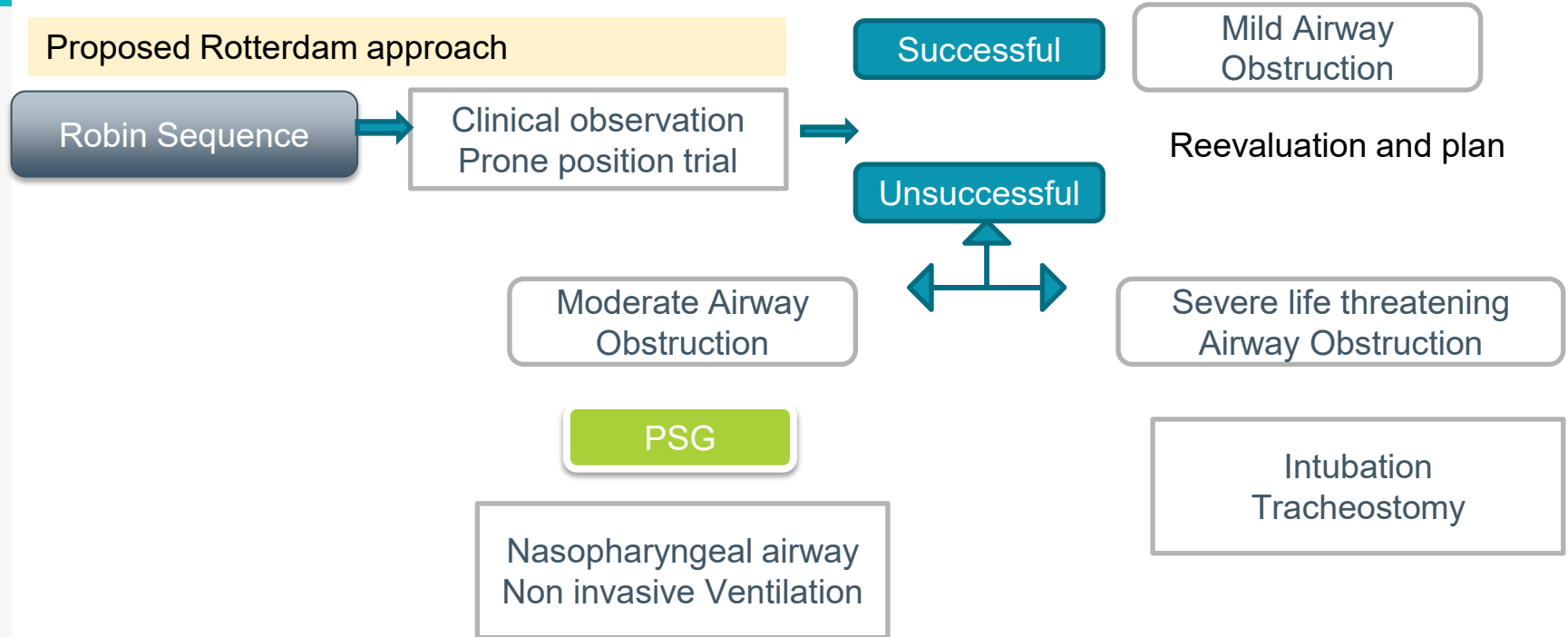
- ▶ Questionable in Presence of cleft palate
- ▶ Indication: RDS, Obstructive sleep apnea/Upper Airway Obstruction
- ▶ Alessandro Amaddeo et al.
  - ▶ Use of CPAP as first line management in a national PRS reference centre
  - ▶ Total 37 patient reviewed, Successful management with CPAP in 9 neonates

Alessandro Amaddeo et al, *European Respiratory Journal* 2015 46: PA1567

- ▶ Leboulanger et al,
  - ▶ Total 83 patient reviewed.
  - ▶ Noninvasive respiratory support- 7 babies. median age of use- 2 month
    - ☒ Decreased respiratory effort,
    - ☒ Improved gas exchange
    - ☒ Avoided Tracheotomy

Leboulanger et al *Pediatrics* 2010 Nov;126(5):e1056-63

# Summary of Respiratory Management- Proposed Rotterdam approach



## Management of associated factors- multidisciplinary team Approach

- ▶ **Neonate intensive care specialist**- clinical assessment, non-surgical treatment, GERD, feeding and growth
- ▶ **Plastic surgeon**- surgical management
- ▶ **Oral and maxillofacial surgeon** - surgical management
- ▶ **Otolaryngologist** - assessment of UAO by endoscopy, tonsillectomy
- ▶ **Geneticist** - genetic testing for syndromes
- ▶ **Speech and language therapist** - speech development, swallowing, feeding difficulties
- ▶ **Nutritionist** - feeding difficulties, growth

## Take Home Message

- ▶ Airway obstruction can be managed successfully by Noninvasive method
- ▶ Don't hesitate in exploring utility of NIV in Pierre robin sequence
- ▶ Nasopharyngeal Airway can be useful in avoiding surgical intervention

**Thank You**