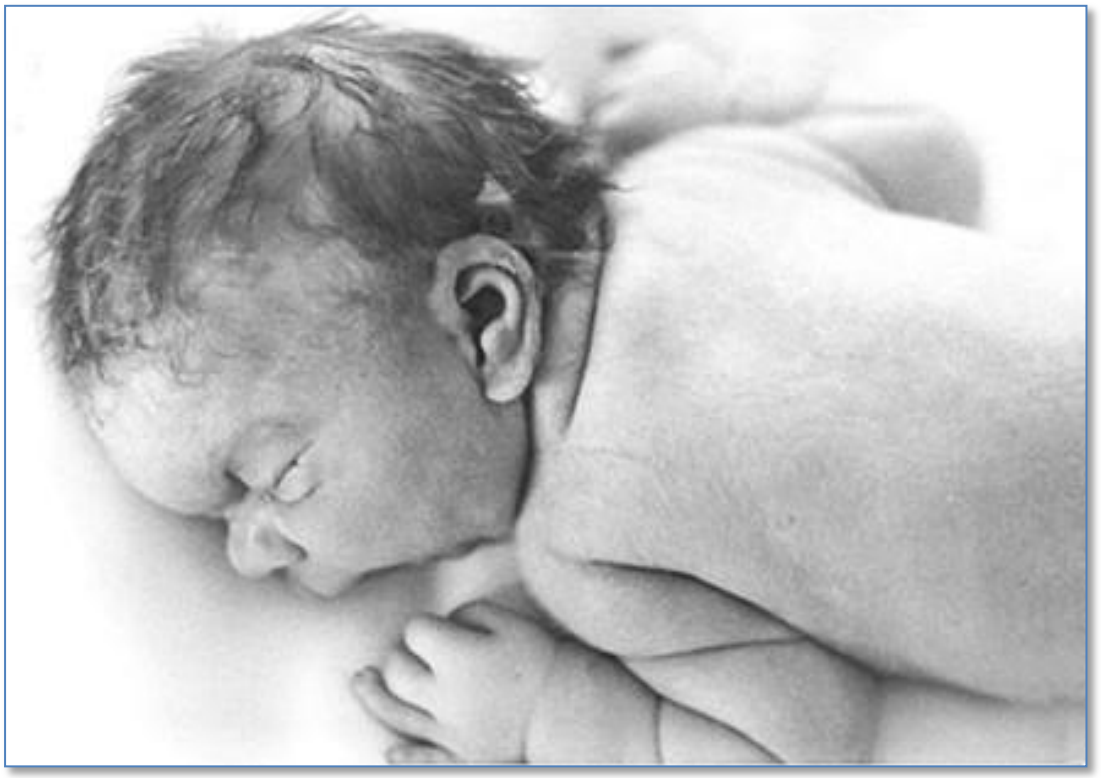
Clinical instruction report for newborns with

Pierre Robin Sequence



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2015

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Ganespalteafdelingen

For application at pediatric and obstetric wards in Denmark.

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This clinical instruction report has been approved by Consultant Plastic Surgeon Mikael Andersen, department S 3082, The Danish National Hospital, Copenhagen.

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Local Health Visitor specialized in cleft lip and palate in Denmark

Appendix 2 s. 33, LIST OF ADRESSES.

The clinical instruction exist on following homepages :

www.ikh.rm.dk/ganespalte /

www.lgcenter.dk

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Introduction

Background

Each year, 5-10 children are born with Pierre Robin Sequence in Denmark, which constitutes 1:14.000 per live-born child, equally distributed between boys and girls. This is demonstrated in a study by Printzlau and Andersen which features 50 children born between 1990 and 1999 (Andersen and Printzlau 2004).

These children are born with respiratory and nutritional deficiencies, which can be cured by means of intensive observation and respite care during the first months of life. In order to avoid permanent ill-effects, a highly competent pediatric staff and thorough parental training are imperative.

Due to the small number of cases of Pierre Robin Sequence, little evidence-based information on nursing and treatment of this patient group exists. However, some experience-based information has been provided by the few physicians, nurses, and health visitors responsible for this patient group in both Denmark and Europe. The clinical instruction provided in this report derives from evidence-based knowledge combined with 'best practice' obtained via the aforementioned experts. These experts will be mentioned by name in the text henceforth, and a list of their names as well as their professional relations to children with Pierre Robin Sekvens is provided at the end of the report. Pierre Robin Sequence will be abbreviated PRS henceforth.

This report centers on children with isolated PRS. Infants diagnosed with PRS may suffer from other congenital disorders which will not be addressed in this report. 62 per cent of the 50 children examined by Printzlau and Andersen have isolated PRS while 38 per cent suffer from other congenital disorders in addition to retro/micrognathia and cleft palate (Printzlau and Andersen 2004).

As described by Prow, in 80 per cent of cases the cause of PRS is to be found in various subjacent genetic circumstances (Prow 1999). Reduced intra-uterine growth of the mandible results in cleft palate and glossoptosis. The diagnosis PRS is made immediately post-birth, and the respite care process is initiated by a competent staff of which the majority are nurses. Since the respite care process also includes general infant care it is important that the parents take part from the outset and

receive thorough training in caring of the infant. They must be able to perform relevant nursing activities prior to discharge.

Diagnosis

PRS was first described by French physician Pierre Robin in 1934. According to his description the following conditions must occur in order for the diagnosis PRS to be made:

- **MICROGNATHIA**
  Underdeveloped mandible. Printzlau and Andersen (2004) apply the term **retro/micrognathia**.
- **U-SHAPED PALATE CLEFT**
  Printzlau and Andersen (2004) describe that merely two thirds of the 50 children included in their study had u-shaped palate clefts.
- **GLOSSOPTOSIS**
  Tongue retraction.
- **OBSTRUCTION OF THE PHARYNX**
  Caused by tongue obstruction in the palate cleft.

Breugem (2010) and Shprintzen (1992) describe how other similar diagnoses have emerged, including Pierre Robin Anomaly and Pierre Robin Sequence, but suggest that all physicians apply the original term in order to further research in and treatment of this group of children. Printzlau and Andersen (2004) ratify the required conditions for the original diagnosis with infants. Marcellus (2001) states that the majority of references describe PRS as a sequence, although many prefer to use the term 'syndrome'. PRS can be genetically determined or stem from intrauterine malformations.

Prow (1999) asserts that PRS is a sequence as it is caused by various genetic conditions. The underdeveloped jaw grows rapidly during the first 5 months, and the symptoms glossoptosis and obstruction of the pharynx will thus subside during this period and relieve the infant of some of its distress. A cleft palate can vary in size, from submucous cleft palate (cleft covered by skin) and cleft uvula, to a cleft in the soft and hard palates.

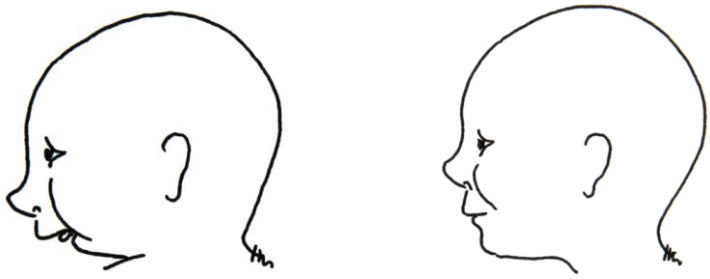
Symptoms

MICROGNATHIA can be detected in a prenatal ultrasonography, but this has not been described. Marcellus (2010) note that this may be due to former complexities involved in attaining an image in profile. However, as ultrasound instruments constantly improve, it is likely that micrognathia would be detectable on a scan today.

If the diagnosis micrognathia is made prenatally, the birth should be performed in a hospital with a pediatric department. The condition micrognathia involves an under-developed jaw as well as **weak jaw-joints**.

During the first 6 months the growth rate of the jaw is substantially higher than average. By the age of 6 years the jaw will have grown to normal size.

The weak jaw-joints stabilize gradually concurrently with the growth of the jaw. In frequent cases the respiratory and nutritional difficulties subside after approximately 4 months, improving the child's overall well-being and reducing impairments. It is imperative that the child makes it through the first months without injuries, as the various impairments 'heal' themselves.



Drawing based on photograph of new-born child with severe PRS – and the same child at the age of six

The **PALATE CLEFT** is most commonly referred to as being U-shaped, but according to Printzlau and Andersen (2004), one third of palate clefts have a narrower V-shape. A palate cleft can include most of the hard and soft palate and thus seem non-existing, or it can be a very small cleft in the soft palate with an indentation into the hard palate. The very large clefts and the very small clefts are at biggest risk of being overlooked (examination for palate cleft has been discussed in collaboration with Trisha Bannister, consultant nurse at Regional Cleft Lip and Palate Network, Northern England, Isle of Man, and Northern Wales, 2010) .

The palate cleft is operated on at the Danish National Hospital's Department of Plastic Surgery S 3082 during the child's second year of life in accordance with an individual estimate made amongst surgeon and anesthesiologist.

GLOSSOPTOSIS – the tongue of a child with PRS is placed in the middle of the oral cavity and has a broad tip. A newborn child with PRS is unable to point the tongue and push it forward into the gum. The infant makes stertorous sounds when in the supine position, and the respiratory difficulties can cause cyanosis. It is often difficult to establish contact with an infant with PRS when it is in the supine

position, as all energy and effort are exerted on breathing. When in the lateral position the tongue function improves but is still unable to reach gum or lips.

OBSTRUCTION OF THE PHARYNX – can be observed immediately post-birth but may aggravate during the first 2-3 weeks when the infant may have been discharged. Wilson (2000) describes the first case of an infant with PRS experiencing airway obstruction during its seventh week of life. Obstruction is caused by a negative inspiratory pressure retracting the tongue even further into the palate cleft.

Other congenital disorders in connection with PRS. Most common: Stickler Syndrome.

As previously mentioned, several infants with PRS suffer from other congenital disorders, at least one third according to the literature upon which this report is based. These disorders vary, from 2 intergrown toes to severe trisomy 18. The most common syndrome in connection with PRS is the hereditary Stickler Syndrome.

STICKLER SYNDROME:

Like all syndromes, Stickler Syndrome varies in severity. Infants with PRS are always referred to an eye specialist when approximately 2 months of age and when the worst respiratory problems have subsided. The eye specialist makes the diagnosis Stickler Syndrome.

The infant should thus be examined for the following symptoms:

- Retinal detachment and myopia (short-sightedness), seen in approximately 70-75 per cent of cases
- Hearing impairment
- Arthropathy and reduced longitudinal growth, early degenerative joint disease
- Palate cleft and micrognathia
- Heart defect (mitral valve prolapse)

In the study conducted by Printzlau and Andersen (2004) 6 out of 50 children had Stickler Syndrome. Marcellus (2001) confirms that Stickler Syndrome is the most common related syndrome.

Children born with 'isolated' cleft palate

Approximately 50 per cent of infants with cleft palate have slightly under-developed jaws and jaw joints. When in the supine position, stertorous sounds can be caused by the tongue's retraction into the pharynx, and the infant is thus more comfortable sleeping in the lateral position. In frequent cases this lasts for merely 1 to 2 weeks. This notion does not agree with the recommendation made by the National Health Service of Denmark that infants sleep in the supine position. Guidelines written by Bannister, Hudson, and Williams (2009) recommend the lateral position – the infant's individual circumstances and needs should determine the position. Heavy stertorous sounds are a sign of respiratory difficulties which will cause the infant to tire easily and become unable to concentrate. The staff at the postnatal ward must look carefully for signs of apnoea and if in doubt, the child should be transferred to the pediatric department.

Even with retracted tongues, infants with isolated cleft palate can be changed and nursed in the supine position. They should drink formula from a soft plastic bottle with a teat that fits the size of the mouth and a hole accommodated to the infant's capacities. The bottle is pressed concurrently with the infant's drinking. It is important that the infant does not tire before being full. Infants with cleft lip and palate receive breast milk like other infants in Denmark, and their prosperity rates in month five correspond to the average rate for Danish infants (Smedegaard, Marxen, and Moes, 2008).

Children with isolated cleft palate can be admitted into the postnatal ward along with their mothers and are examined for other disorders by a pediatrician prior to discharge.

A health visitor specialized in cleft lip and palate must always be contacted when a child is born with facial deformities. Her job is to assist the pediatrician in making the diagnosis, provide advice as regards observation and treatment, and train the parents, the physician, and the nurses in observing the infant's individual needs. She is to supervise the parents and nurses in handling the infant with as much care as possible while maintaining a natural environment and satisfying the infant's most basic needs.

The cleft lip and palate health visitor is part of the Joint Danish Cleft lip and Palate Team (App. 2).

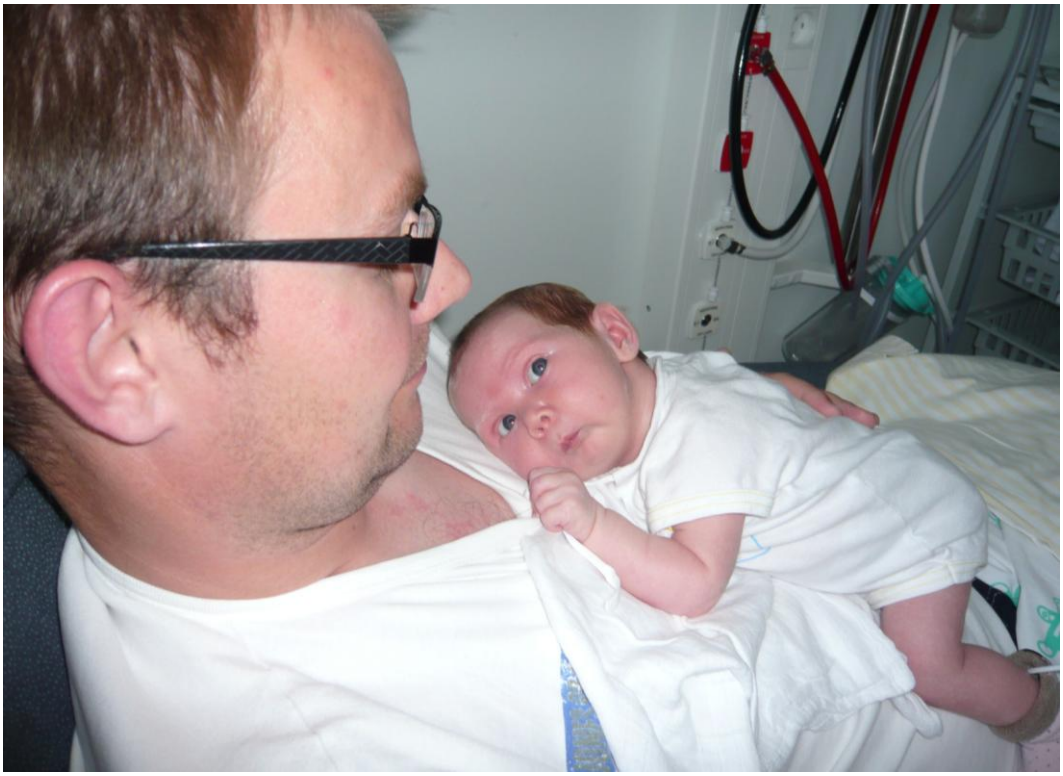
When a child with PRS is born – plan of immediate postnatal action

ALSO FOUND IN APPENDIX 2 – 'ACTIONCARD'

- Midwife calls pediatrician and neonatal nurse to the delivery room.
- Parents and infant are kept as close together as possible in order for the 'bonding' process to commence. The parents must learn to interpret and act upon the infant's signals whilst accepting the disorder.
- The infant must be kept in the lateral position and the stomach position. The infant may lie on the mother's breast and look up but may also suckle on the breast. While the infant is enable to breast-feed, the suckling stimulates bonding and milk production.
- Continuous SAO2 monitor on. Standard value: >95 per cent.
- Count of respiratory rate. Standard value: <58/minute.
- Examination of the infant should be made in the lateral or the stomach position. Use a light and a spatula to examine the oral cavity.
- The infant is placed in an incubator for the first couple of days and is observed for signs of cyanosis, indentations, and an overall pattern of movement. Does the infant prefer to bend backwards? The infant is observed for other congenital disorders or dysfunctions. The infant may of course lie in the stomach position on the parents' chests during observations.
- Random PCO2 measurement Standard value: 3,6-5,3
  Transcutaneous PCO2 is approximately 0,6 kpa above the arterial level at a calibrate temperature of 43 degrees Celsius. Transcutaneous PCO2 should be controlled at regular intervals (gas values).
- Haematological test, acid-base.
- With capillary gas values both pH and Pco2 are reliable and agree with artery blood when there is decent peripheral perfusion. (Pedersen 2006) and pediatrician Rasa Cipline.
- The infant is tube-fed during the first days of life in order to provide relief. The daily quantity should be less than normal 'early feeding', unless indicated by other factors.
- The mother is instructed in breast pumping, and the infant is slowly introduced to the feeding bottle with assistance from the specialist health visitor. It is important to make sure that the infant is able to suckle and swallow – this reflex is not well-functioning.
- Like with a premature infant, start with a moistened cotton applicator or a finger in the infant's mouth and milk in a syringe. The infant will expend a great deal of energy on breathing.
- In case the infant obstructs in the stomach position, a Nasal Airway is inserted from the nostril to just above the epiglottis with a nozzle enabling ventilation. If CPAP is deemed necessary,

finish with a Benveniste's Valve. In the case of a more long-term Nasal Airway treatment, the tube is fixed in a simpler and safer manner, refer to 'Nasal Airway' further on in the report.

- When a cleared respiratory tract is ensured, the infant should be transferred to the neonatal unit. A Nasal Airway must be inserted during transportation of the infant, and the infant must be in the stomach position in order to ensure a cleared air passages.
- The specialist health visitor is contacted (App. 2). Physician or midwife sends a report to the Cleft Lip and Palate Centre in either Aarhus or Copenhagen.



Problem areas 1 – 3

1. **MOTHER-CHILD RELATION/PARENT-CHILD RELATION**
2. **RESPITATION**
3. **NUTRITION**

1. The mother-child relation/the parent-child relation

The child is to live off the love of its parents.

During the first 2 months of life an infant's behavior is needs-based (Brodens 1991).

The mother of an infant with PRS has a natural concern for her child's well-being and is occupied with ensuring that it is breathing and growing.

The infant needs assistance in regulating sleep routine and concentrating; in this, eye- and body contact are vital means.

The child is born with a relational competence i.e. sensitivity towards faces, touch, and the voices of its parents. The interplay between mother and child opens the way for the bonding process; it enables the infant to integrate with its surroundings and develop into a social being.

Stern Daniel (2008) states that the basic elements of the early bonding process, i.e. security, affection, daily routine regulation, close bodily contact, and basic instruction in human relations should take place in the preverbal stage.

An infant with PRS and its parents face severe challenges, as the infant must be washed and changed in the lateral or the stomach position. Moreover, the infant will tire easily as it exerts all concentration on breathing. The infant has difficulties suckling and is unable to breast-feed. Parents will likely experience a fear of losing their new-born child in addition to feeling overwhelmed with the amount of time that must be put into nursing an infant with PRS.

This interplay between father and child helps the child integrate into its surroundings. Eye- and body contact are vital means.

All infants with PRS are discharged and brought to their home before 4 months of age when the impairments and difficulties should be abating. The majority of infants with PRS are discharged during the first to second month of life unless there are other congenital disorders. There is a general tendency for parents of a PRS patient to want to take over the full responsibility for their child's well-being; they are, however, often surprised at how demanding it is to nurse the child at home. Thus, early skin-to-skin contact is equally imperative as training in observation, treatment, and general nursing of the child.

Stubenitsky (2009) points out that parent/child bonding takes place during the first 5 years. It is important that the hospital staff provides sufficient supervision and support with the aim to enable the parents to nurture and treat the infant on their own and ultimately reduce hospitalizations.

1. a. Support of parental competencies.

- If the neonatal unit has a family room, this should be applied.
- Along with the specialist health visitor, the neonatal physicians and nurses, who are responsible for parental supervision prior to discharge, constitute the parents' group of permanent contact persons. If possible, a nurse and the local health visitor ought to visit the family at home while the child is hospitalized.
- The parents are referred to the medical social worker concerning such issues as loss of earnings, house-hold assistance, and remuneration of additional expenditures.
- Offer of psychological counseling. In many cases the parents do not experience a need for counseling before the child's symptoms have subsided. There are psychologists attached to the municipality, the hospital, and both Cleft Lip and Palate Centers.
- Thorough training in observation and nursing of the child. Refer to training form, App. 3.

1. b. The following health educational method can be applied for parental training

PRACTICAL TRAINING FORM IS OFFERED IN APPENDIX 3.

According to Antonovsky the human sense of coherence and meaning stems from various learning processes which the individual passes through during a lifetime. That is, the experience of predictability forms the basis of a sense of understanding, while the experience of an appropriate work load forms the basis of manageability, and participation and involvement provide basis for a sense of meaningfulness.

LEARNING PROCESSES	SENTIMENT RELATED TO COHERENCE AND MEANING
• Predictability	- understanding
• Work load	- manageability
• Participation	- meaningfulness

(Jensen and Johnsen 2000). The passage 'sundhedspædagogik og følelsen af sammenhænge'.

2. Respiration

Obstruction of the pharynx caused by retro/micrognathia, i.e. a retracted tongue and a cleft palate, causes a negative pressure in the pharynx during respiration. Obstruction of the upper airways can cause hypoxia and related disorders, CO2 accumulation in the blood, pulmonary vascular stasis, and cor pulmonale (Marcellus 2001).

Course of action in the case of tongue obstruction

The content of this paragraph is experience-based.

- The infant experiences a choking-sensation which is unpleasant for the infant as well as the parents.
- I have experienced a breath of wind from an ambulance door being shut causing the tongue to loosen.
- Hold the infant steadily on your left arm and near your stomach, place your right index finger below the root of the tongue, and pull. The tongue is hard like a cramping muscle and the SAO2 level is very low which causes cyanosis. According to Rolf Holm Knudsen, anesthesiologist at the Danish National Hospital, – and my own experiences – the finger is more lenient than a tongue depressor. Make sure to keep a tongue depressor near the infant, but attempt with the finger first. The Nasal Airway model 1. is simple and efficient if it is possible to insert it into the pharynx as a first aid.
- 'I had to ask a mother with very long nails to cut the nail of her index finger in case of obstruction'.
- Call in a physician (anesthesiologist or pediatrician). A Nasal Airway should be inserted.

The tongue can obstruct and create a blockage if the infant does not have a Nasal Airway or a palate plate during the first 4-6 weeks. In the most severe cases a tongue fixation is performed.

In Printzlau and Andersen's (2004) study of 50 infants with retro/micrognathia, 16 per cent of the cases were mild, 38 per cent were moderate, while 46 per cent were severe. Moreover it has been asserted that there is a certain correlation between the growth of the mandible and respiratory difficulties.

Cole, Lynch, and Slator (2008) agree with the above-mentioned assertion and provide a categorization scheme of PRS according to symptoms, treatment, and severity. The difference in mandible size is illustrated in 3 photographs.

For an examination carried out at Birmingham Children's Hospital, children with poor oxygen saturation were observed over 24-36 hours, and the saturation level proved to be below 90 during 5 per cent of the time (Andersen 2007). PCO2 and acid/base are evaluated on the basis of standard values. Pediatrician Rasa Cipline asserts that a child's saturation level should generally be > 92. Anderson (2007) states that continuous SAO2 monitoring is necessary during the first 4 months of life. Due to the risk of hypoxia-related disorders, an apnoea alarm is not sufficient – Pediatrician Rasa Cipline agrees with this notion.

Rasa Cipline asserts that control of respiratory rate, a random PCO2 measurement, and an acid/base review ought to be made even if the saturation level is > 90 per cent. These are initiated at the time of hospitalization and concluded at discretion.

Standard values for infants

- Respiratory rate < 58 times/minute
- PCO2 3,6-5,3 kpa
- Acid/base 7,37-7,45
- SAO2 > 95 per cent

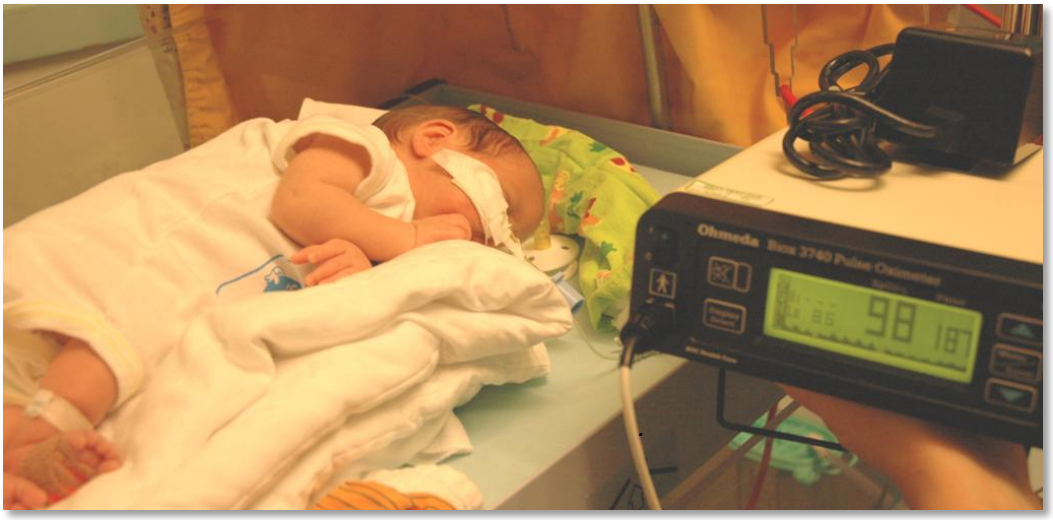
2. a. Step 1 in the respite care process: Placing the infant on its stomach or in the lateral position

The face must point downwards in order for the tongue to push forward. The infant's breathing should be as soundless as possible. In the supine position the infant will make stertorous sounds, tire, become restless and possibly cyanotic.

A comforter will help push the tongue forward into the mouth.

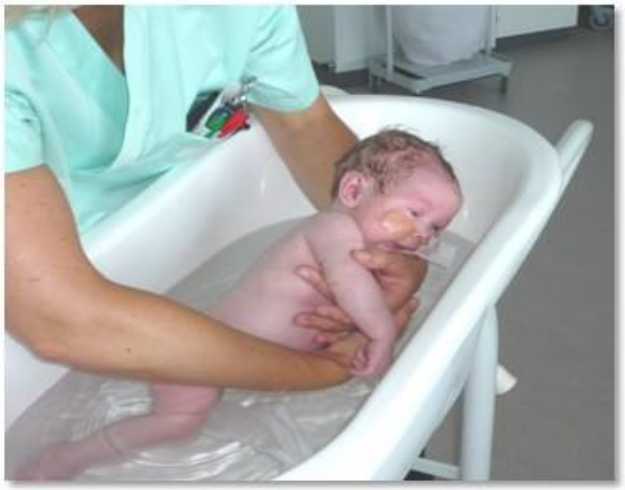
When lying on the stomach, the infant's head should be turned to the side but may shift from left to right, as with premature infants lying on a bulge to right above the shoulders.

Alternatively, a rolled diaper can be placed under forehead and shoulders and the face turned straight down towards the mattress. A cylindrical cushion under the back can prevent the infant from rolling over. At Skejby Hospital they apply a triangular cushion which can be purchased at 'Baby Paradise', Solsortevej 17, 9600 Aars, Tel. 51913958. English hospitals apply a model with a small cylindrical cushion on the front and a larger one on the back, but other alternatives may be useful for back support as well.



Supine position

When nursing, changing, washing, feeding, and communicating with the infant, the infant must lie on its stomach or in the lateral position with continuous saturation monitoring.



Bathing in the lateral position.

Andersen (2007) states that a Nasal Airway should be inserted if the saturation level tends to fall to approximately 80 per cent when the infant is lying on its stomach or in the lateral position (Pediatrician Rasa Cipline agrees). Control of PCO2 and acid/base status.

2. b. Step 2 in the respite care process: Fixed Nasal Airway

If in the stomach/lateral position the saturation level continuous to fall as described above, a Nasal Airway should be inserted:

- A Nasal Airway **ensures** cleared airways and prevents pharynx obstruction.
- A Nasal Airway can prevent the need of surgical procedure i.e. tongue fixation.
- A Nasal Airway can help enable the infant to suckle and swallow and improve its wellbeing.
- A Nasal Airway enables the parents to treat their child on equal basis with other infants, which makes the caring process more natural and furthers parent-child bonding.
- A competent staff can teach the parents to perform a Nasal Airway treatment. The practical training form offered in App. 3 is intended for parental training but can also be applied by the nursing staff.

In 2010 I visited a Danish mother, Tanja, who performed the Nasal Airway treatment at home after discharge from Hvidovre Hospital. She received guidance and supervision during house calls by a

neonatal nurse, a specialist health visitor, and the local nurse (Tanja Breitenkamp Kreuger 2007). She had no former experience but was pleased with the outcome. Tanja has created a personal website on which she shares the experience of looking after her son Marcus.[1]

Andersen (2007) describes 12 mothers and fathers who were trained on the basis of a training form at Birmingham Children's Hospital, refer to App. 3. As a final step in the process the parents were to evaluate their (in)abilities to treat and tend their child's respiratory and nutritional problems at home whilst maintaining their daily routines. Prior to discharge the staff was to make sure that the parents had received thorough training and supervision and were thus capable of tending the child at home. As a result, the average length of stay was reduced from 54 to 19, 5 days. The weight gain increased daily until it reached the average of other infants.

WHAT IS A NASAL AIRWAY?

Portex Tracheal Tube

- A tube size of 3.0 is suitable for a mature child. Select a tube size that fits the child's nostrils.
- For a mature child, the length should be approximately 7, 5-8 centimeters (length from the nose tip to tragus).
- Rolf Holm Knudsen, anesthesiologist at the Danish National Hospital department S 3082, cuts a 0,5 centimeter oval hole placed approximately 0,5 centimeters above the tip of the tube, a **Murphy Eye**, as it has been termed. The Portex 'Blue Line' tube has a Murphy Eye, and some smaller tubes may also have one as well – I personally have only seen a 3, 5 tube with a Murphy Eye.

  In his article, Andersen (2007) describes how he uses a perforator to cut the hole – I have not heard of such a procedure elsewhere. Pediatrician Rasa Cipline (2011) recommends the Murphy Eye.
- The tip of the tube must hang freely close to epiglottis. With advantage, a stent in the form of a small rubber tube lubricated with a local anesthetic can be applied for the passage.
- A stethoscope can be applied for air change monitoring – a tube that is too long will cause vomiting and choking sensations.
- In order to make an appropriate depth estimate, note the distance between the tip of the tube and the nostril on the observation form.
- During the first week of life (or longer if necessary) the tube is connected to a Portex nozzle enabling ventilation. This, however, is inept and unreliable. If the infant needs CPAP, a

[1] www.123hjemmeside.dk/Breitenkampkreuger.

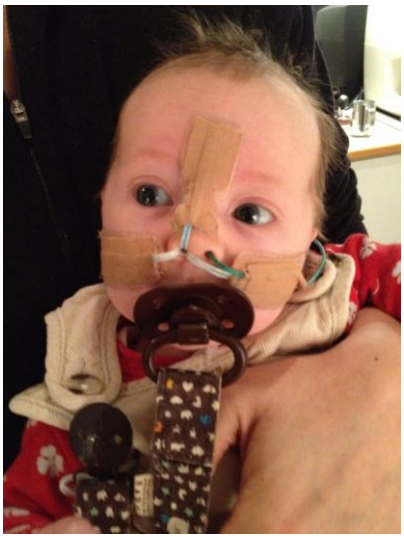
Benveniste's Valve should be applied as a nozzle. Note that this does not provide secure fixation either.

After this a more secure and reliable form of fixation is applied, refer to the images on the following page. The models were composed in collaboration with Pediatrician Rasa Cipline. According to our past experiences, scissors are the most suitable tool for cutting a Murphy Eye.

Inspired by specialist team of South Thames Cleft Service/Guy's and St. Thomas' Hospital, the book 'Guidance. Care for babies with Pierre Robin Sequence', and the personal experience of Emma Southby, specialist nurse in the team, 2010.

Images of Nasal Airway plus models on the following page.

Start model. Danish girl with start-nozzle on Nasal Airway.

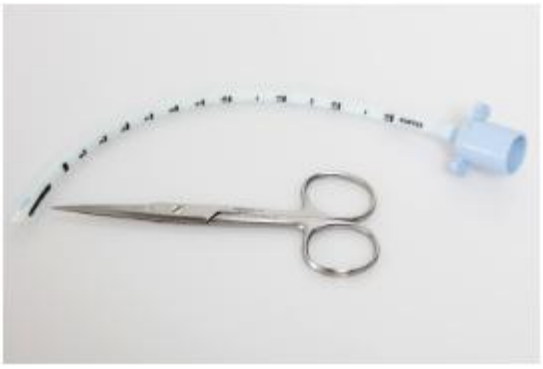


Model 1. – used by danish parents at home.

Model 3. English girl with a secured fixation – the tube is fixed to the grip

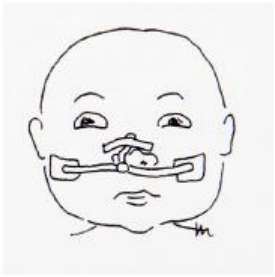
Nasal Airway

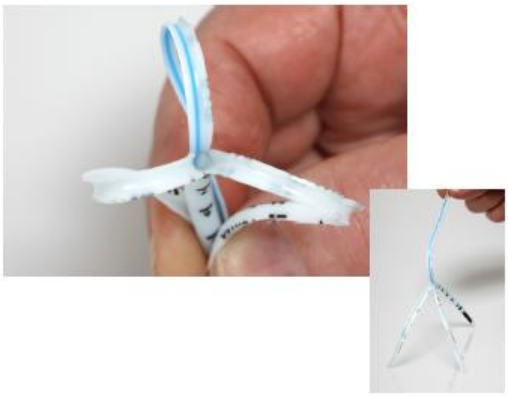
Start model



+ Murthy`s eye

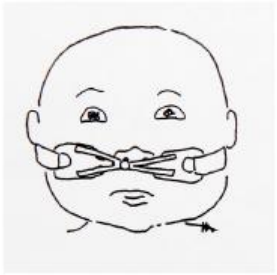
Model 1





1. 3 strips are cut out from the nostril. The first strip is bended upwards towards the eye and fixed on the bridge of the nose (on the same side as the tube), the other 2 are fixed on the cheeks with duoderm and plaster.

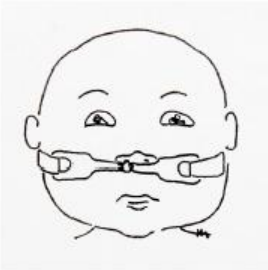
Model 2



2.a. The tube can also be cut 2,5 – 3 centimeters from the nostril – this piece is cut into 4 long strips which are fixed to the tube grip with leukostrip (4 strips in order to maintain a round tube hole). Pull the tube and fix it with duoderm plaster to the cheeks or on a hat.

2.b. The 4 strips can be longer and fixed without a grip, 2 strips on each cheek with a space of approximately 1 centimeter in order to maintain a round hole (model Aalborg, Jette Moes 2010)

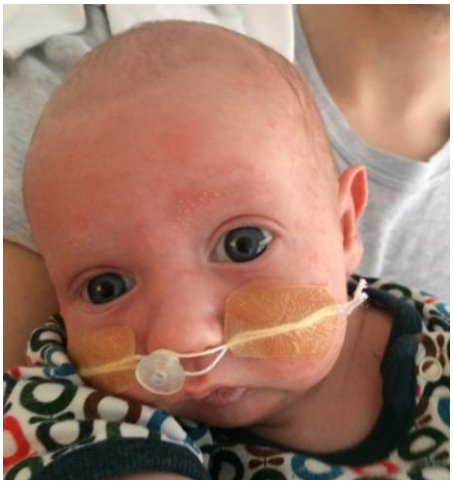
Model 3



3. A tube holder that is one number smaller than the tube is cut and sutured to the tube near the nostril (use a curved needle and a needle holder). The holder with flat cotton strips is fixed to the cheek with duoderm and a plaster or on a hat/ CPAP-hat, in order to protect the infant's skin

(Southby and Bannister 2).

- After discontinuation it may be necessary to re-apply the Nasal Airway when the tube withdrawal process initiates, as the infant needs strength and energy to be able to consume the daily portion via a feeding bottle and thus avoid sleep apnoea.



This Nasal Airway is easy and safe to use by hospital staff and parents. It has been used in Denmark since 2012.
Teleflex Medical , Ireland Ref. 125410 / Denmark MEDIQ tlf. 0045 36379100
Size 16 I.D. 3,5 mm O.D.5.3 mm normal newborn size. Smaller one Size 14 and size 12

NASAL AIRWAY NURSING

- The tube is changed once per week as a minimum, or as needed.
- Keep the tube clean, for instance by suctioning after every meal or as needed. Every 6 hours may be sufficient. Follow a regime in order to avoid obstruction.
- It may be necessary to detach and sanitize the tube, during ward rounds e.g.

The formation of mucus will decrease after a couple of days.

- Rinse with a maximum of 0,5 milliliters isotonic NACL prior to suction. Use for instance suction 6, depending on the tube size, and enter it into the tube (stop approximately 0,5 centimeters above the end of the tube). Move the suction through the tube with 'rolling' motions.
- After every meal, rinse nostrils and add a protective lotion. In the case of skin irritation, move the tube to the opposite nostril.
- The training form for parents offered in App. 3 may be useful.

Inspired by specialist team of South Thames Cleft Service/Guy's and St. Thomas' Hospital, the book 'Guidance. Care for babies with Pierre Robin Sequence', and the personal experience of Emma Southby, specialist nurse in the team, 2010

In case sufficient respiration is not attained after 2-3 weeks of Nasal Airway use, tongue fixation surgery can be performed in Denmark (Mikael Andersen, consultant plastic surgeon, department S 3082, the Danish National Hospital).

Marcellus (2001) relates about positive effects of Nasal Airway for children with PRS. He notes that if a Nasal Airway is not sufficient, a tongue fixation can be performed. Other pharynx anomalies may require other surgical procedures.

In several countries, including Switzerland and Sweden, gum-jaw plates are applied in order to prevent obstruction.

The Danish National Hospital department S 3082 perform tongue fixation surgery in severe cases of PRS.

2.c. Step 3 in the respite care process: Transfer to the Danish National Hospital in preparation for tongue fixation

According to Printzlau and Andersen's results (2004), one third of children with PRS have cleared airways when lying down; one third need Nasal Airways, and one third need tongue fixation.

Since this Clinical Instruction had been used I Denmark 2011 -2014, tongue fixation has not been done. (Consultant Mikael Andersen, Rigshospitalet)

If sufficient respiration is not attained when placing the infant on the stomach or in the lateral position, or by insertion of a Nasal Airway – or if long-term use of Nasal Airway prevents discharge – tongue fixation is the proper solution.

The infant must be at least 3-4 weeks old and weigh more than 3 kg in order for tongue fixation to be made. In the case of premature infants, tongue fixation must be made after the date of confinement.

Mikael Andersen, consultant plastic surgeon at the Danish National Hospital, recommends 2-3 weeks' use of Nasal Airway at the initial department prior to transfer.

The initial department's first call is made to the Department of Plastic Surgery S 3082. App. 1:

- Regarding tongue fixation: Consultant Surgeon Mikael Andersen,
- Regarding Nasal Airway: Consultant Anesthesiologists Rolf Holm Knudsen or Kirsten Eriksen.

Provided the above-mentioned surgeons and physicians find tongue fixation appropriate for the infant, the steps should be as follows:

As specified by Consultant Plastic Surgeon Mikael Andersen, department S 3083, the Danish National Hospital (2011):

The initial department makes contact with the Department of Plastic Surgery S 3082 at the Danish National Hospital and asks for Consultant Plastic Surgeon Mikael Andersen with regard to transfer.

Anesthesiologist and consultant plastic surgeon set a date for transfer in collaboration with the neonatal unit based on capacity in the operating room. The initial department is informed when a date has been set.

The infant carries a Nasal Airway and a saturation monitor during the transfer to the neonatal unit at the Danish National Hospital.

- Day 1. The infant is attended by consultant plastic surgeon and consultant anesthesiologist of department S 3082 at the neonatal unit.
- Day 2. Tongue fixation is performed. The lower side of the tongue is attached to the lower lip, pulling the tongue forward. The sutures are bound in a temporary 'binding plate' that is placed outside of the skin.
- Day 3. Postoperative observations are made at the neonatal unit.
- Day 4. The infant is transferred to plastic surgical department S 3082 (in which all surgeries on Danish children born with cleft lip and palate are performed).
- Day 5. The infant is still fed via a tube but is introduced to a feeding bottle.
- Day 8. The 'binding plate' is removed after the surgery. The tongue has now been pushed forward into the mouth, and the infant is able to move the tip of the tongue freely.

- 14 days after hospitalization the infant will likely be ready for discharge and is either moved to the initial pediatrics department or brought home along with a tube and a feeding bottle.

The tongue fixation is loosened at the age of 7-8 months. The appropriate time for loosening of tongue fixation is determined among surgeon, parents, and specialist health visitor at the Palate Council's cross-functional consultation. The Palate Council assembles at the Cleft Lip and Palate Centers in Aarhus and Copenhagen.

After fixation of the tongue the infant is able to eat from a spoon like other children and drink from a cup when the time is right. Infants with PRS are rarely accustomed to the feeding bottle. Tongue fixation does not remedy problems with vomiting which some children with PRS experience. The cause hereof has not been described.

After tongue fixation the infant feels energized and is better able to communicate. Breathing becomes effortless, and the parents are relieved of their fears and concerns as regards the risk of obstruction. The child can now lie on its back, which enables better communication, and the parents' contact with and nurture of the child is much less complicated.

As an immediate secondary effect of tongue fixation the lower gum flattens which causes the lower teeth to become horizontal. Both will correct themselves when the fixation is loosened.

According to the above-mentioned physicians of department S, a tracheostomy, which is rather difficult to close, should not be necessary.

A mother describes how her child had a tracheostomy prior to a tongue fixation, as the diagnosis PRS was not made immediately post-birth (Perlman 1992).

Palate cleft surgery is performed when the child is 1,5 – 2 years of age.

3. Nutrition

Infants with PRS experience nutritional difficulties due to reduced suckling- and swallowing abilities as well as respiratory distress.

Children with isolated cleft palate swallow approximately 5 times. In comparison, the average child merely swallows once (X-ray photography from a study conducted in Dresden and presented at an International Craniofacial Congress in Gothenburg 2001). Infants with PRS experience respiratory difficulties in addition to swallowing difficulties, which cause them to expend much energy on breathing and thus tire easily.

3.a. Growth curve

The weight curve of an infant with PRS can be expected to be below the Danish average until the age of 4 months. The length curve is normal, and the infant will thus appear long and thin.
Stubenitsky (2010) states that the weight increase of infants with Nasal Airways is normal during the first month of life, resulting in prolonged hospitalization.

Prodoehl (1995) describes how a tube in the pharynx can open up for air permeability. The size of the tube is not mentioned.

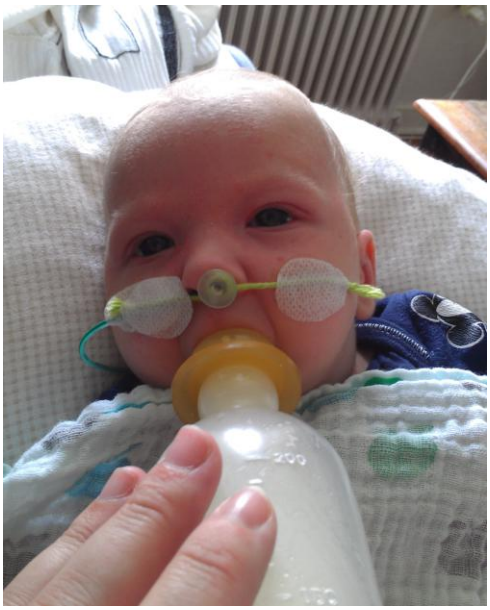
Unless other diagnoses have been made, the PRS patient is considered a healthy child with a lip and palate deformity. The deformities described above cause respiratory difficulties, **suckling- and swallowing difficulties, and weariness, which ultimately leads to nutritional problems, reduced weight increase, and potentially sitophobia.**

3.b. Conducting meals

- It is imperative that the eating environment is as 'normal' as possible i.e. that the child is comfortable and fed by either its mother or its father.
- The mother is supported and supervised while breast pumping. A breast pump can be borrowed free of charge (more on this later on in the report).
- The infant is fed solely via a tube during the first 24 hours of life, though not necessarily as large an amount as in 'early feeding'.
- The health visitor specialized in cleft lip and palate makes a first attempt to feed the infant while the parents and the nursing staff are instructed.
- A syringe containing milk is placed in the infant's mouth while the infant is suckling on either its mother's or its father's finger, and the milk is sprayed into the mouth in drops next to the finger. Alternatively a cotton applicator dipped in milk can be applied for starters.
- When the infant is ready an attempt is made to feed with a soft teat on a soft plastic bottle – a disposable bottle is preferable. At first, do not press the bottle. Observe the infant and the saturation monitor. Not until the infant is able to suckle and swallow safely may the bottle be pressed carefully. According to Shaw (1999), a soft bottle encourages the suckle/swallow reflex better than a hard bottle.
- The mother/father sits in a comfortable position in an armchair with a footstool, and the infant is laid on a duvet or a pillow in the lateral position (on the left side if the bottle is held in the right hand) with an elevated head. The mother/father must be able to see the infant's face as

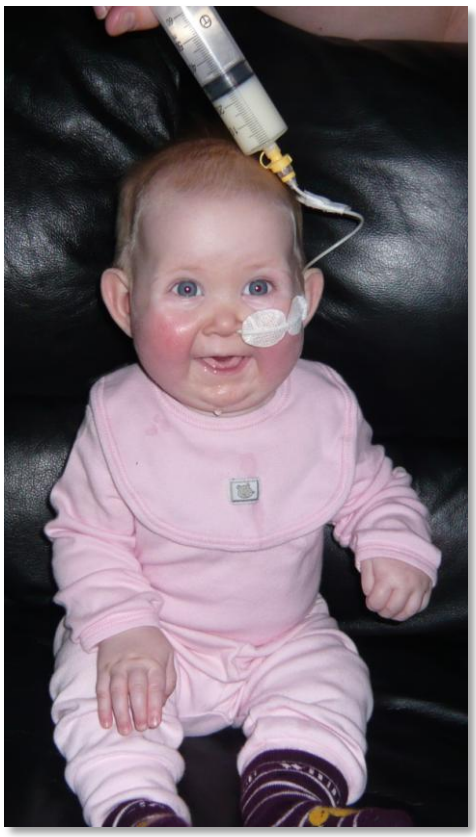
- well as the SAO2 monitor. As with breast feeding, joint collaboration is paramount. If the mother/father is unable to relax, so is the infant, which makes coordination of the suckling/swallowing reflex all the more difficult.
- The mother/father grabs the jaw joint from the back of the head with the middle finger and thumb and presses upwards and forward. Supporting the jaw will stabilize it and enable the infant to suckle properly on the teat, push it upwards into the palate, and move the mandible slightly. The infant might not be able to suckle and swallow the first couple of times, but practice makes perfect.
- When the infant is able to suckle and swallow safely and properly, the bottle may be pressed mildly while the infant is suckling. Due to the palate cleft the infant is unable to create a partial vacuum in its mouth. If SAO2 stays below 90, the infant is likely tired and the feeding should be discontinued.
- The infant should be full before tiring. However, the duration of the meal should be 30 to 45 minutes; if the infant is not properly fed after 45 minutes, apply a tube for supplementation.
- An amount of 8 meals per 24 hours is necessary during the first months of life, as the infant easily feels nauseous and fatigued.

Without Nasal Airway: Support of the mandibular angle during a meal. Eye contact and deep concentration.



3.c. Prolonged tube feeding, referral to dietician, and potential subsequent meal training

- If at discharge the infant is unable to consume the appropriate daily amount of milk based upon the instructions provided above, the parents can be trained in tube feeding – the training form in App. 3 can be applied. The tube is replaced once per week (or immediately if unstable). A permanent tube is changed every two weeks. A feeding pump should only be applied for larger amounts of milk or for a prolonged feeding process.
- Tube replacement takes place at the Department of Pediatrics.
- There is a risk that the infant may vomit and be unable to consume its daily amount. Prolonged tube feeding may cause vomiting as well. Fortification may become a necessity. Alternatively the infant can be fed with a highly nutritious dairy product as per agreement with a dietician in order for smaller amounts to be sufficient.
- **Prolonged tube feeding can cause the infant to develop sitophobia. In such a case, meal training will be provided at the Department of Pediatrics when the infant is ready.**



Danish girl of 7 months hospitalized for meal training after prolonged tube feeding.

The solution to nutritional problems as outlined above corresponds to the recommendations provided by Watson (2001) as well as Bannister and Southby's 'Guidance. Care for babies with Pierre Robin Sequence' (2009).

For a study on infant nutrition from 2005, 62 children – of which 12 had PRS – were observed (Reid 2005). During the first 2 weeks, 10 infants suffered from severe nutritional difficulties, while 2 infants experienced moderate nutritional difficulties. The 10 infants still had severe nutritional difficulties after 3 months; after 14 months, the number had reduced to 5, 2 of which were tube fed.

While it is difficult to nourish infants with PRS, it is my personal belief that an early effort by specialists will result in a reduction

of nutritional distress at the age of 14 months. This claim is supported by Bart (2009) who conducted a study on 30 infants with PRS in collaboration with a specialist team in Toronto, Canada. The weight increase amongst the 30 infants was lower than 700 g during the first 4 weeks, and the infants were continuously fed via stomach tubes, depending on the level of respiratory distress. Bart (2009) characterizes treatment of these infants as being highly challenging due to potential prolongation of hospitalization e.g.

3.d. Breast milk after all – borrowing a breast pump and purchasing a bottle and teat.

Recyclable soft plastic bottles for infants with cleft lip and palate can be purchased at Lyngby Svaneapotek (Lyngby Pharmacy), Tel. 45870096. Regular bottle teats are applied. The size and shape of the teat must fit the infant's mouth, and the size of the hole should be adapted according to the infant's suckling- and swallowing abilities. By purchasing both on their own, the parents become active participants in the treatment process while learning to observe the infant. Testing various teat sizes might be preferable. The Nuk teat is recommendable; first size 1, then size 2. If necessary, additional holes can be made. New teat models continue to emerge, e.g. the new Pigeon teat, which has a vent hole and is softer on one side. A selection of potential bottles and teats can be seen in 'Klinisk Vejledning for Børn med Læbe-ganespalte' ('Clinical Supervision for Children with Cleft Lip and Palate').

If the mother wishes to use a breast pump, a requisition form can be gathered from the cleft lip and palate health visitor. The Cleft Lip and Palate Centre in Aarhus will pay for the breast pump, which will be at the parents' disposal for as long as necessary.

A vast percentage of infants with PRS receive breast milk and for as long an average period as other Danish infants (Smedegaard 2008).

Gradual discharge and potential follow-up.

- Once the parents have attained the required competences and are able to nurse the infant at home, the infant is discharged by its primary physician and nurse. Please refer to App. 3.
- The clinical nurse specialist and the local health visitor are notified of the discharge, and it is recommendable that they participate in an evaluation meeting prior to discharge. The house

visits paid by an attending health visitor may take place more frequently than usually, as she is to check up on the infant's weight and the family's overall well-being. The specialist health visitor will maintain close contact with the family via house visits, e-mail, and telephone. Additionally she will constitute the interface between the family's health visitor and physician, the hospital, the local case worker as well as other social and medical bodies when necessary.

- Like all hospitalized infants the patient is examined and the hospitalization process is evaluated.
- The father should have at least 2 weeks off after discharge – the mother is on maternity leave. Household help and home nursing may be necessary.
- The hospitalization is now 'open', and the parents can contact the pediatric Department.
- Should the pediatric department have a 'take home nurse' like at Hvidovre Hospital e.g., it is recommendable that she pays the family 1 or 2 visits upon discharge as a connecting link between the primary and the secondary sector.
- The infant should be discharged with a saturation monitor, and it is imperative that the parents are able to use it. They must, however, also be able to observe the infant's breathing by watching its chest. During the first 4-6 months the saturation monitor is particularly vital during eating and sleeping.
- SAO2 monitors can be borrowed from the Central Aid Office of the North Denmark Region.
- **If the infant has a stomach tube** the parents must be cognizant of tube feeding. The pediatric department should offer to insert the tube.
- Until an agreement has been made between the hospital case worker and the municipality that the parents can either collect tubes from the local pharmacy or have them delivered from the hospital depot, they may collect the necessary remedies at the pediatric department.
- **If the infant has a Nasal Airway** the parents must be able to insert it, fix it, and rinse it via suctioning, ensuring its functionality.
- Home visits by the primary nurse or a health visitor within the first week upon discharge ought to be considered.
- New tubes can also be ordered fro the pharmacy and paid via the municipality, as can stomach tubes and other single-use materials.
- A movable suction device will be ordered, and the parents must be able to use it. In rare cases oxygenation can become necessary – the remedies must be ordered prior to discharge. The parents will receive instruction in application of the oxygen cylinder when it is delivered to the family's home.

- A referral to the Hearing Center is made, primarily in order to schedule the standard infant screening of which a positive result may be delayed due the palate cleft which causes the middle ear to suppurate. Moreover, in the case of Stickler Syndrome the infant may have a hearing impairment.
- A referral to the pediatric eye specialist is made. If the infant has Stickler Syndrome, this diagnosis will be made by the eye specialist.
- A referral to the Pediatric Outpatient Clinic is made by the primary physician. At least 3 examinations will be made – at the ages of 6 months, 2 years, and 5 years respectively, or as needed. The examinations will be scheduled in collaboration with the Department of Plastic Surgery S 3085 and the attending physician. The primary foci will be respiration, other potential congenital disorders, and the infant's overall well-being.
- The results of the FISH-test and the array CGH test are sent to the Department of Clinical Genetics if the infant suffers from other congenital disorders than cleft palate and micrognathia. The degree of severity of other disorders should determine when the tests are carried out, since all other infants go through a similar standard procedure. Furthermore, the bonding process is plenty problematic as it is.
- Once again the parents are offered psychological counseling, either at the hospital of via the specialist health visitor of the Cleft Lip and Palate Center. This offer will remain valid after the acute phase.
- A referral to a dietician should be made if the infant is unable to consume the proper daily amount of milk. This often becomes the case some time upon discharge.
- The local case worker is contacted, e.g. by the hospital case worker, and notified that an infant with special needs is discharged.
- Commentary for the attending physician.
- Nursing commentary for the attending nurse.

Discussion

I have been working as a health visitor specialized in cleft lip and palate in the North Denmark Region for 23 years and been employed part-time as a nurse at the neonatal department at Aalborg Hospital for 15 years. I have followed 10 children born with PRS, from birth till the age of 3; each time I have felt that I – and the rest of the staff – lacked the guidelines necessary for handling this group of patients, which practically 'heal themselves' but do, nevertheless, require help in order to avoid permanent ill-effects.

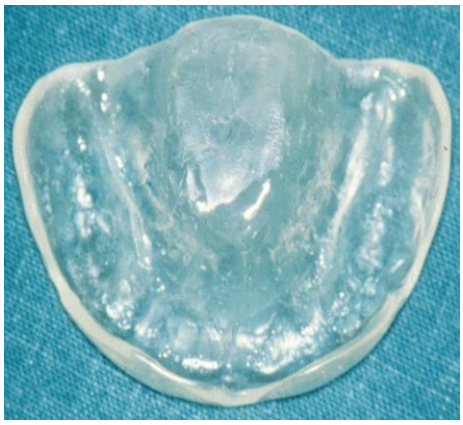
Additionally, being part of a small team of specialists working specifically with PRS patients, I have followed the treatment development and participated in international conferences.

THE TREATMENT OF CHILDREN WITH PRS VARIES GREATLY WORLDWIDE.

In other parts of the world they perform relief surgery on infants with PRS – these surgeries may damage the tissue structure. Large international craniofacial congresses often offer presentations regarding treatment of PRS; oral surgeons, from the United States e.g., display images of infants having had a mandibular distraction with subsequent jaw extension treatments or tracheotomy.

Consultant Plastic Surgeon Mikael Andersen is of the conviction that tracheotomy is unnecessary for infants with PRS but that a tongue fixation, on the other hand, is recommendable, as it can provide great relief for infants with severe PRS without permanent secondary effects.

In England the treatment of PRS is similar to that of Denmark and does not involve relief surgery. In severe cases a tongue fixation, which was formerly described based upon scientific articles and the experiences of Trisha Bannister, is performed.



PALATE/GUM PLATE

In Sweden and Switzerland a so-called 'palate/gum plate' is applied for all infants with cleft palate. In the video 'Mauro – yes, he can!' by Claus Herzog, MD. and Nurse Christi Herzog-Ilser, Switzerland, (2010) it is indicated that in addition to a palate plate, a Nasal Airway might be necessary for infants with PRS, but that tongue fixations or other forms of relief surgery are not performed on infants with severe PRS in Sweden or Switzerland. My interviews with cleft lip and palate specialists Karolin Ghazarian, orthodontist at Uppsala University Hospital (2010) and Karin Stabel Svensson, specialist nurse at Karolinska Hospital, Stockholm (2010) verified this indication.

NB. If palate/gum plates become sought after in Denmark, these can be produced by the dental surgeons at the Cleft Lip and Palate Centers. At the aforementioned Swiss clinic, the plates are made of a material which extends concurrently with the infant's growth; thus, merely one plate is needed per patient whereas in Sweden, several palate plates are produced for one infant. The gum/palate plate is already inserted during the infant's first week of life. Since it is problematic to transport an infant with PRS on a busy highway, much consideration should be put into selecting the location for the master impression to be made.

Stillizig (1998), orthodontist at Heidelberg Hospital in Germany, has manufactured a gum/palate plate with a small posterior curve for infants with extremely small mandibles.

Bannister, Hudson, and Williams (2009) point out that a gum/palate plate does not reduce the partial vacuum in the mouth. This is to say, the infant's suckling abilities do not improve. The major advantage of a gum/palate plate is thus its preventing the tongue from getting stuck inside the palate cleft.

Prenatal diagnostics

In Printzlau's and Andersen's (2004) study, 17 of the 50 infants examined suffered from various obstetrics-related prenatal problems. The average birth weight was 3100 g, which is below the Danish average for infants. 30 per cent of the infants were either 'small for date', premature, or both, and 10 per cent were premature, that is born before week 37. Ultrasonography devices constantly improve; in the future, then, obstetricians may become able to detect the reduced mandibular growth, make the diagnosis PRS prenatally and prepare the staff that are to deliver the infant.

Anesthesia

Rolf Holm Knudsen, anesthesiologist at the Department of Plastic Surgery, the Danish National Hospital, asserts that some infants with PRS have a narrow pharynx, which does not constitute a

nuisance for the infant but may cause problems for the anesthesiologist who is unaware that the infant is diagnosed with PRS. It has been suggested that infants with PRS should wear signs indicating their diagnoses. But in such as case, for how long should they wear the sign?

With this report, placing focus on the small group of children with PRS, I hope to raise awareness and appreciation that the lenient ways in which we treat these patients in Denmark should remain. It might be possible to make the treatment even less burdening for the child and less complicated for both the parents and the pediatric staff.

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Svensson, Katrin Stabel, specialsygeplejerske, Karolinska Sygehus, Stokholm, Sverige(Ganeplader)

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Layout

Audio-visual department, Aalborg Hospital – Aarhus University Hospital. Images of Nasal Airway, 2011.

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Pierre Robin Sequence, Pierre Robin Syndrome, cleft palate, micrognathia, glossoptosis, upper airway obstruction, Nasal Airway, respiratory distress, feedingtube, Stickler Syndrome, bonding, barn, health promotion, disease prevention.

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English Translation

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5-year-old with a normal jaw and chin. She is clever and ready for school. This is the Danish girl from the baby photos in this clinical instruction.

Appendix 1: Action card

PLAN OF IMMEDIATE POSTNATA ACTION WHEN A CHILD WITH PIERRE ROBIN SYNDROME IS BORN.

- Midwife calls pediatrician and neonatal nurse to the delivery room.
- Parents and infant are kept as close together as possible in order for them to experience 'bonding'. The parents must learn to interpret and act upon the infant's signals whilst accepting the disorder.
- The infant must be kept in the lateral position and the stomach position. The infant may lie on the mother's breast and look up but may also suckle on the breast. The infant will not be able to breast-feed but the suckling stimulates attachment and milk production.
- Continuous SAO2 monitor on. Standard value: >95 per cent.
- Count respiratory rate. Standard value: <58/minute.
- Examination of the infant should be made in the lateral or the stomach position. Use a light and a spatula to examine the oral cavity.
- The infant is placed in an incubator for the first couple of days and is observed for signs of cyanosis, indentations, and an overall pattern of movement. Does the infant prefer to bend backwards? The infant is observed for other congenital disorders or dysfunctions. The infant may of course lie in the stomach position on the parents' chests during observations.
- Random PCO2 measurement Standard value: 3,6-5,3
  Transcutaneous PCO2 is approximately 0,6 kpa above the arterial level at a calibrate temperature of 43 degrees Celsius. Transcutaneous PCO2 should be controlled at regular intervals (gas values).
- Haematological test, acid-base.
- With capillary gas values both pH and Pco2 are reliable and agree with artery blood when there is decent peripheral perfusion. (Pedersen 2006) and pediatrician Rasa Cipline.
- The infant is tube-fed during the first days of life in order to provide relief. The daily quantity should be less than normal 'early feeding', unless indicated by other factors.
- The mother is instructed in breast pumping, and the infant is slowly introduced to the feeding bottle with assistance from the cleft lip and palate health visitor. It is important to make sure that the infant is able to suckle and swallow – this reflex is not well-functioning.
- Like with a premature infant, start with a moistened cotton applicator or a finger in the infant's mouth and milk in a syringe. The child will expend much energy on breathing.

- In case the infant obstructs in the stomach position a nasal airway is inserted from the nostril to just above the epiglottis with a nozzle enabling ventilation. If CPAP is deemed necessary, finish with a Benveniste's valve. In the case of a more long-term nasal airway treatment, the tube is fixed in a simpler and safer manner, refer to 'Nasal Airway' further on in the report.
- When a cleared respiratory tract is ensured, the infant should be transferred to the neonatal department. A nasal airway must be inserted during transportation of the infant, and the infant must be in the stomach position which will ensure a cleared respiratory tract.
- Contact is made to the specialist health visitor (App. 2). Physician or midwife sends a report to the department for palate clefts in either Aarhus or Copenhagen.

Appendix 2: List of addresses for the Danish Cleft Lip and Palate Team

Cleft Lip and Palate Centre of West Denmark. Institute for Communication and Disabilities (Institut for Kommunikation og Handicap). Aarhus	**Peter Sabroesgade 4**	**8000 Aarhus C** Ikh.ganespalte@ps.rm.dk www.regionmidt.dk www.ikh.dk /ganespalte ?? www.sku.rm.dk /områder og afdelinger/ ganespalte	**Tel. +45 78412345**
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Aarhus Municipal Hospital **Department of Orthognathics**	Nørrebrogade 44	8000 Aarhus C	
Cleft Lip and Palate Centre of East Denmark Centre of Head and Orthopedics, The Danish National Hospital	Rygårds Allé 45	2900 Hellerup LGH@hav.1.regionh.dk www.lgcenter.dk ? ? www.rh.dk /klinikker/ born og unge –læbe-ganecenter	**Tel. +45 45114475**
The Danish National Hospital, **Department of Pediatrics, Centre of Head and Orthopedics 3082/**83	Blegdamsvej 9	2100 Copenhagen Ø **www.rh.dk /borneafdeling** DEPARTMENTS/Centre of Head and Orthopedics/Department of Pediatrics	**Tel. +45 35453083**
The Danish National Cleft Lip and Palate Association	www.llg.dk		

Øvrige specialsundhedsplejersker ansat i Aarhus:

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Vibeke Thinggaard	tlf. 5121 4541	vibeke.thingfaard@rm.dk	Region Syd (Fyn)
Susanne Langberg	tlf. 3074 2343	susanne.nielsen@rm.dk	Region Syd

Specialsygeplejersker ansat i Hellerup:

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Karen Marie Laadal	tlf. 4072 0936	karin.marie.laadal@rh.region.dk	Region hovedstaden/sjæll.

PALATE COUNCIL

- **Plastic surgeon, specialized in cleft lip and palate**
- **Orthodontist, specialized in cleft lip and palate**
- **Ear, nose and throat doctor, specialized in cleft lip and palate**
- **Health visitor, specialized in cleft lip and palate**
- **Speech therapist, specialized in cleft lip and palate**

Appendix 3: Training form for parents

Patient label

Parental training form – for appliance at discharge of newborns with Pierre Robin Syndrome Skills required prior to discharge with:	Shown	Conducted	Approved
Bottle/Tube feeding			
Demonstrates knowledge of the anatomy of the child's upper air passages.			
Is able to account for the principles determining the selection of bottle and teat plus the reason for potential tube feeding.			
Is able to place the child in an appropriate position, observe suckling abilities and level of strength and energy during meals, and apply an SAO2 monitor. Is able to determine when supplemental tube feeding is necessary.			
Is familiar with tube feeding remedies and how to obtain these after discharge.			
Is able to insert a tube into the correct location, fixate the tube, and feed the child through the tube.			
Nasal Airway			
Understands how/why to apply a Nasal Airway, based on knowledge of the anatomy of the child's upper air passages.			
Demonstrates understanding of why and when a Nasal Airway is necessary for this particular child. Is able to observe the child's breathing and apply an SAO2 monitor.			
Is familiar with the necessary remedies and how to obtain them after discharge.			
Is able to lay the child in an appropriate position and rinse the tube before removal.			

Is able to measure the right length of a new tube. Is able to insert the tube correctly and steadily while causing as little discomfort for the child as possible. Is able to ensure that the tube is in its correct location and fixate it simply and securely. Is able to prepare and conduct safe and gentle suction. Knows how to react in case the tube is obstructed by mucus.			
Knows when a specialist health visitor, the attending physician, or the pediatric department ought to be consulted.			
Is aware of the necessary course of acute action in case of tongue obstruction in the pharynx or severe respiratory distress.			

I/we have the following remedies at home and know how to use them:				
SAO2 monitor	Suction device	Feeding pump	Oxygen cylinder	Stethoscope

I am/we are conversant with the above-outlined procedure and prepared to nurse my/our child at home.

Date_____Signature_____

Filed in the child's journal along with a copy for the parents.