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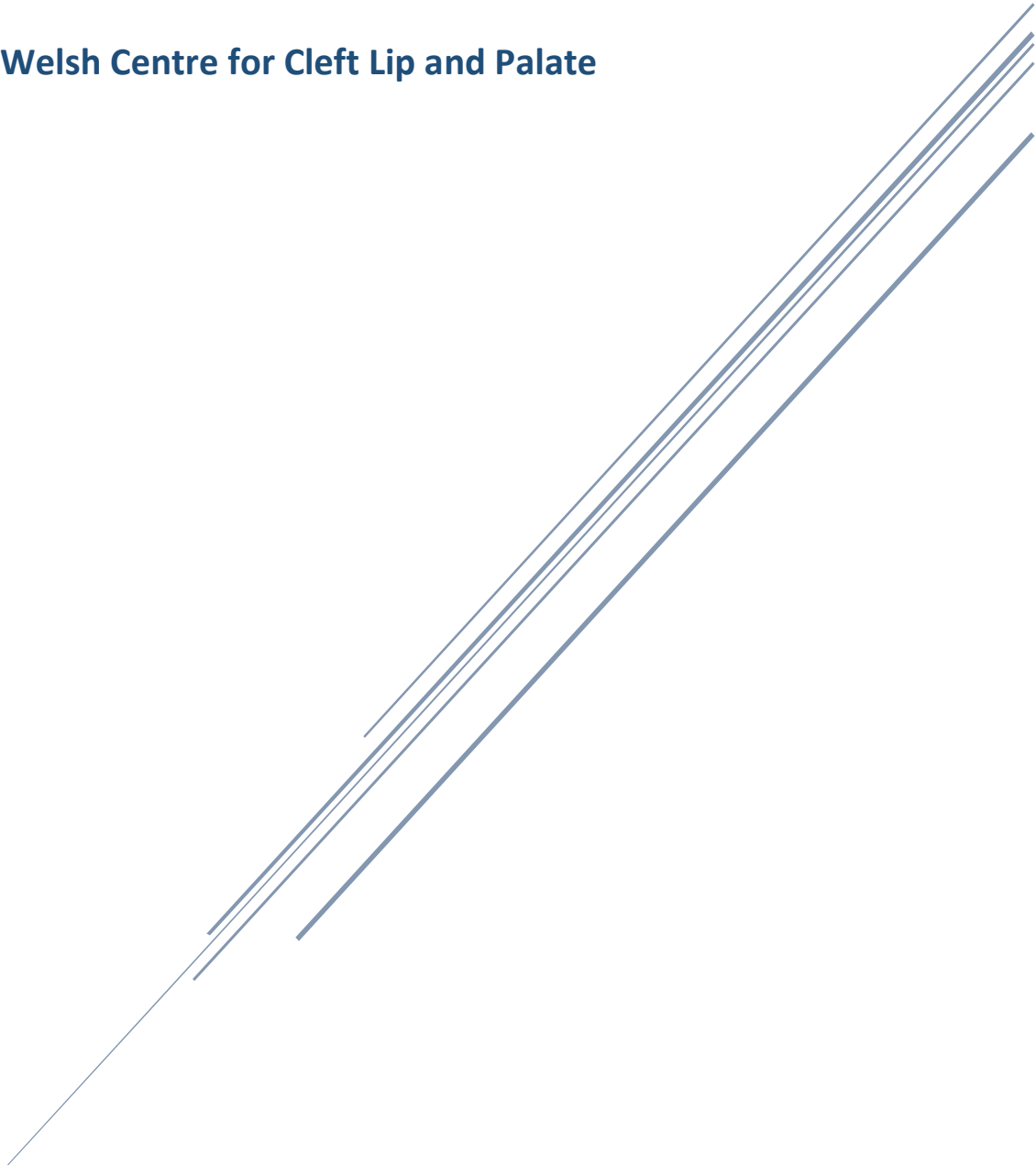
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The Welsh Centre for  
**Cleft Lip & Palate**

# South Wales Management Pathway for a Baby with Cleft Palate and Pierre Robin Sequence (PRS)

The Welsh Centre for Cleft Lip and Palate





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## Background

The Welsh Centre for Cleft Lip & Palate are based at Morriston Hospital, Swansea. We provide a service to those born with a cleft lip and/or palate who live in South, West and parts of Mid Wales.

We manage babies who are born with a Cleft Palate and Pierre Robin Sequence (PRS) in partnership with the Paediatric Respiratory Service who are based at the Childrens Hospital for Wales (CHfW), Cardiff.

Pierre Robin Sequence is the combination of micrognathia, glossoptosis and cleft palate (although cleft palate may be absent in some cases). These infants will have difficulty breathing and may have many obstructed breaths even if oxygen saturations are well maintained in air. They can be difficult to assess. They will have difficulty coordinating feeding and breathing and problems are likely to be exacerbated at times of viral infection. These infants will need to be referred to the paediatric respiratory service at The Children's Hospital for Wales for further management of breathing and feeding. A nasopharyngeal airway (NPA) is usually required. The jaw grows well over the first year in most children with PRS, and breathing and feeding problems usually improve over about 9 months. Children with other dysmorphic features may have a specific syndrome (e.g. Stickler syndrome, Goldenhar syndrome, Treacher Collins syndrome) and may need more complex management.

Cleft repair is usually between 9-12 months of age once the NPA has been successfully removed.

## Assessment and Referral

### Referral

Refer to cleft team: 01792 703810 [service will make contact within 24 hours]  
Consider transfer baby to neonatal unit  
NG feeds and Nil by mouth  
Oximetry and monitoring



Assessment by Cleft and Neonatal teams



Features consistent with Pierre Robin Sequence or other micrognathia syndromes



Discuss with Paediatric Respiratory Service @ The Children's Hospital for Wales

Respiratory distress	Some respiratory concerns	No respiratory concerns	
Oximetry and monitoring Insert nasopharyngeal airway	NG feeds Nil by mouth Oximetry and monitoring Optimise position (side)	Overnight oximetry study Discuss with Paediatric Respiratory service @ Children's Hospital for Wales	
NG feeds Nil by mouth	Referral and transfer to Paediatric Respiratory Service @ Children's Hospital for Wales for 10 day admission	Abnormal oximetry study	Normal oximetry study (consider repeating Day 5)
Referral and transfer to Paediatric Respiratory Service @ Children's Hospital for Wales for 10 day admission		NG feeds Nil by mouth Oximetry and monitoring Optimise position (side)	Feeding assessment by Cleft Nurse Training and provision of feeding equipment
		Referral and transfer to Paediatric Respiratory Service @ Children's Hospital for Wales for 10 day admission	Equipment Provision and training [Home Oximetry, BLS, car seat challenge]
			Audiology assessment Ophthalmology assessment Echocardiography Check antenatal ultrasound for renal abnormalities
			Discuss with Paediatric Respiratory Service @ Children's Hospital for Wales prior to discharge
			Sleep study @ Children's Hospital for Wales within 2 weeks

Link: [Children's Hospital for Wales guidance on NP insertion](#)



## Pre-cleft repair. Respiratory service management and timeline.

### Assessment at birth

Sleep polygraphy without NPA for assessment of obstruction  
Insertion of Vygon Nasopharyngeal airway [stick to a single nostril]  
Sleep study with NPA in-situ for assessment of NPA positioning

Nil by mouth and NG feeds  
Equipment Provision and training [NPA management, Home Oximetry, suction, BLS, car seat challenge]  
Feeding assessment by SLT. Training and provision of feeding equipment [liaison with local services]  
Audiology assessment, Ophthalmology assessment, Echocardiography, Renal assessment  
Airway management information leaflet for parents  
Respiratory / Cleft MDT

### Assessment at 3 months

Reassess size of NPA  
Sleep polygraphy with newly sized NPA in-situ  
Nil by mouth and NG feeds  
Feeding assessment by SLT. Potential to start remove then replace NPA for feeds during the day

### Assessment at 6 months

Reassess size of NPA  
Sleep polygraphy overnight with NPA in-situ, then extend into daytime with NPA removed  
If polygraphy acceptable through daytime naps, remove NPA during the day and reinsert each night  
Feeding assessment by SLT. Increase oral feeding as possible

### Assessment at 9 months

Sleep polygraphy overnight with NPA removed  
If polygraphy acceptable, remove NPA both day and night  
Parents keep NPA kit in case of problems with intercurrent infection  
Parents continue nighttime oximetry monitoring until after cleft palate repair

### Assessment at 14 months [post cleft repair]

Sleep oximetry post cleft palate repair  
If oximetry successful, remove home oximetry monitoring  
Discharge from paediatric respiratory and sleep service



## Post-operative care

The Cleft Specialist Nurse will contact the Respiratory Clinical Nurse Specialist to update about the outcome of surgery and the ongoing plan of care.

- If an NPA is needed post operatively, the duration required will be decided by the anaesthetist and cleft surgeon as this can change on an individual basis.
- On the day following palatoplasty, the surgeon and the anaesthetist together will review the baby, and if safe to do so they will remove the NPA for a trial period. Close observation and monitoring will be required during this time
- If observations are satisfactory for the following 24 hour period, then the patient can be discharged home without the NPA.
- If the observations are not satisfactory, then the Cleft Surgeon will re-pass the NPA. The baby will be prepared for discharge home with the NPA in-situ.
- The cleft surgeon will liaise with the paediatric respiratory consultants in the Childrens Hospital for Wales (CHfW) to update them on the patient's post-operative status and ensure that the discharge plan is agreed.
- The cleft nurse will ensure that the Parents have the necessary equipment to care for their baby safely at home. Items will include suction machine and catheters, pulse oximetry monitor and probes, appropriate sized NPA's and tapes.
- The cleft nurse will contact local hospitals to organise open access and liaise with the paediatric respiratory nurse specialist.
- The respiratory nurses will arrange ongoing support and a post-operative sleep study.



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## Parent information leaflet: Pierre Robin Sequence

### **Pierre Robin Sequence**

Your baby has been born with a small jaw and a cleft palate. This can make it difficult for your baby to breathe because their airway can become obstructed. This is called Pierre Robin Sequence.

The Welsh Centre for Cleft Lip and Palate, Morriston Hospital, and the paediatric respiratory team at The Children's Hospital for Wales will look after your child and make sure their breathing is safe as they grow. Most children grow out of their breathing problems as the jaw grows and can have their cleft palate repaired at the end of their first year.

### **The first year of life**

In the meantime, your baby will need some help with their breathing, and this is achieved using a nasopharyngeal airway (NPA). The NPA is a small tube that is passed down one nostril to reach behind the tongue so that the airway stays open and can't be obstructed as the tongue falls back. It's not safe for your child to feed while they have an NPA in position, so they will need to be fed with a nasogastric (NG) tube, a thin tube inserted through their nose into their stomach. If you were planning to breast feed your baby, the team will support you to express your breast milk to give to your baby via the NG tube. Your baby may feed by mouth again as soon as it is safe for them to do so, and this will be monitored with regular assessments by the speech and language therapists in the respiratory team.

### **Transfer to the Children's Hospital for Wales in Cardiff**

When you go to the Children's Hospital for Wales, your child will be assessed with breathing tests and overnight sleep studies, and will have the NPA positioned in the nostril. It takes a bit of time to get the NPA in the correct position, but when everything is set, you will be trained in how to manage your child with an NPA, how to change the NPA, how to use suction, how to use the monitoring equipment, and how to use the NG tube for feeding. There will be lots of time to ask questions and become confident with what is needed. Your first visit to the Children's Hospital will be for about 10 days and usually we can discharge you directly home from there.

### **Once you're at home**

When at home you will have the direct number to the paediatric respiratory team in Cardiff who will be able to help you with problems that arise. You will come to visit us for further assessments at 3 months, 6 months and 9 months of age. Hopefully by 9 months your baby won't need the NPA anymore and they can then have their cleft palate repaired.