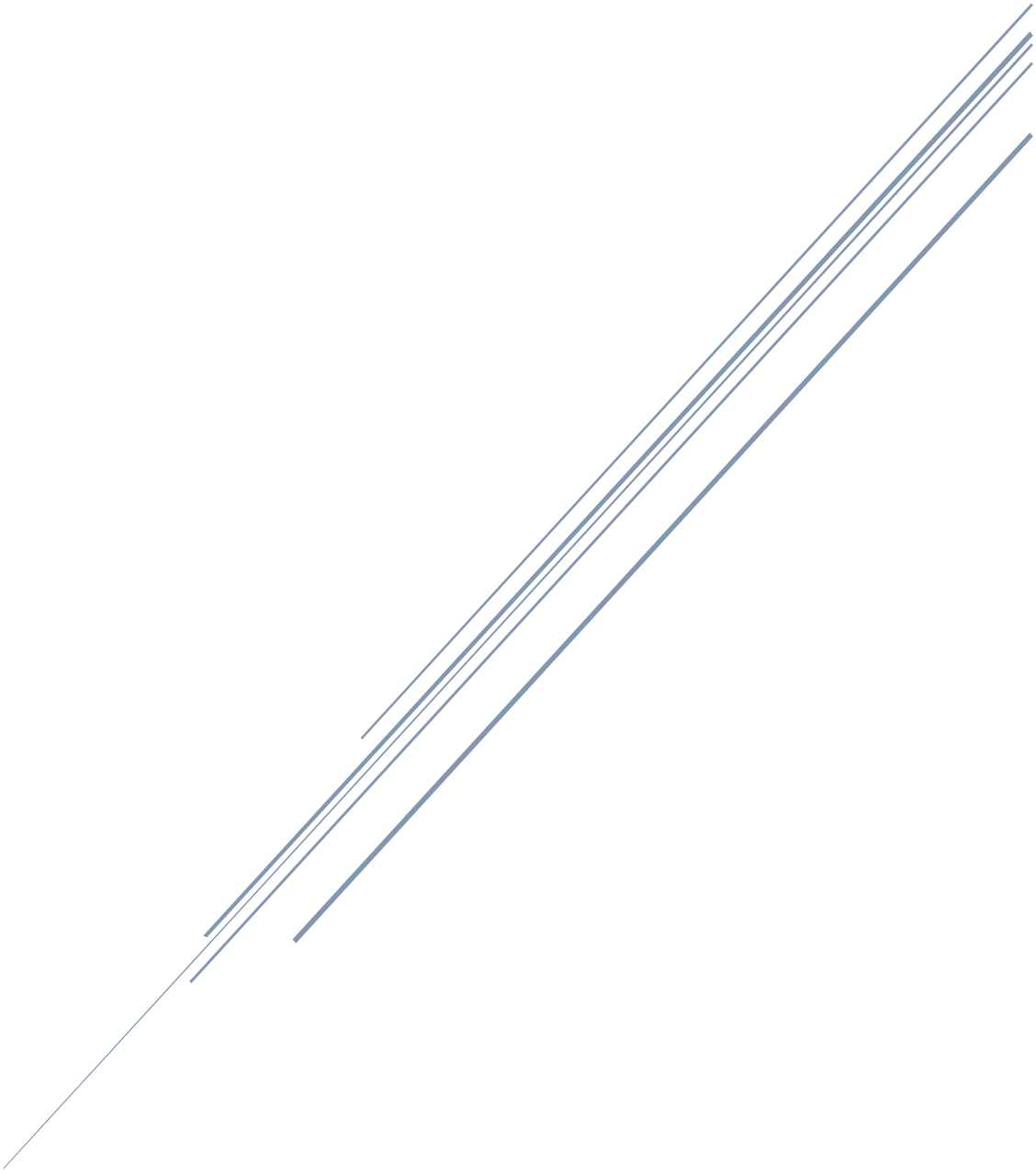
All Wales Paediatric Respiratory Network

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 care for 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 Children's Hospital for Wales (CHfW), Cardiff.

Pierre Robin Sequence (PRS) is the combination of ***micrognathia*** (a small lower jaw), ***glossoptosis*** (placement of the tongue towards the back of the mouth) and ***upper airway obstruction*** (difficulty with breathing). PRS is often associated with a ***cleft palate***, which is a condition present at birth where the tissues in the roof of the mouth have not fused together . These infants will have difficulty breathing and may have many obstructed breaths, even if oxygen saturations are well maintained in air. They may 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 assessment and further management of breathing and feeding. A nasopharyngeal airway (NPA) may be required. The lower jaw grows over the first year in most children with PRS, and breathing and feeding problems usually improve over about 9 months. Children with other congenital anomalies may have a specific syndrome (e.g. Stickler syndrome, Goldenhar syndrome, Treacher Collins syndrome) and may need more complex management.

Cleft palate reconstruction is usually performed between 8-12 months of age, once the NPA has been successfully removed and ideally with the NG tube removed and oral feeds established.

Assessment and Referral

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 nasopharyngeal airway if significant obstruction [stick to a single nostril]
Sleep study with NPA in-situ for assessment of NPA positioning

Patients with NPA stay nil by mouth and receive NG feeds and dummy dips
Patients not requiring NPA may or may not require NG tube feeding

For all patients
Equipment provision and training [Home Oximetry, suction, BLS, car seat challenge (and NPA training if NPA in situ)]
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

Patients without NPA receive all follow-up feeding advice from cleft team

Assessment at 3 months

Patients with NPA
Reassess size of NPA
Sleep polygraphy with newly sized NPA in-situ
Nil by mouth and NG feeds
Feeding assessment by SLT in all patients
Potential to commence oral feeds with NPA removed for short periods in the day around feeding

Patients without NPA attend for sleep study only. Feeding progression directed by cleft team

Assessment at 6 months

Patients with NPA
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 whole day and reinsert each night
Feeding assessment by SLT
Accelerate oral feeding as tolerated. Ongoing community SLT and dietetics with aim to remove NG by 9 months

Patients without NPA proceed to surgery if 6 month sleep study is adequate. Equipment returned. No respiratory follow-up

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
Feeding assessment by SLT
Parents continue nightime oximetry monitoring until after cleft palate repair

Assessment at 12 months [post cleft repair]

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

Parent information leaflet: Pierre Robin Sequence

Pierre Robin Sequence

Your baby has been born with a cleft palate and a small jaw which means his/her tongue may fall backwards towards the throat, especially when lying on their back. This can make it difficult for your baby to breathe. 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 baby and make sure their breathing stays safe as they grow. Most children grow out of their breathing problems as the jaw grows and can have their cleft palate repaired towards the end of their first year.

The first year of life

In the meantime, your baby may need some help with their breathing, and this can be 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 because there is a risk of choking, 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 if this is needed. 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 baby 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 reconstructed.

Taking care of yourself and getting support

The diagnosis of a baby with a Cleft Palate with PRS can be a difficult and overwhelming experience for parents. Being a parent of a baby is tiring in itself, and babies with PRS require even more attention. This can be a time of anxiety and stress for families. Taking time to look after yourself is important, but can often feel like it is not a priority. Try to engage your wider support network. Sometimes it helps to connect with others going through a similar experience as this can provide valuable emotional support and practical advice. CLAPA is one charity that can help you find a peer supporter (https://www.clapa.com/parent-support).