Management of infants born with a cleft lip and palate. Part 1

Cleft lip and/or palate is a relatively common facial anomaly occurring in babies, which varies in frequency according to racial or ethnic group. A holistic approach to care for the family and child involves many professionals at different times throughout the childhood years. This is the first of two articles which will look at nationally agreed standards of care for the immediate postnatal period, some of the problems which may occur in early infancy for both babies born with a cleft involving the lip and those born with an isolated cleft of the palate, and the specialist support services available to local healthcare professionals and parents.

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Key points

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1. National standards have been agreed by the Nurse Special Interest Group, Cleft Lip & Palate to provide an informed response for families following the birth of a baby with an oral cleft.
2. Cleft lip is often diagnosed during pregnancy and is immediately identified at birth, but cleft palate is more difficult to diagnose and may be missed at the initial birth examination.
3. Cleft lip or palate can be associated with other anomalies, which may affect the management of the baby.
4. Parents are profoundly affected by the attitudes of staff caring for their baby and require a lot of sensitive support in the immediate postpartum period.

Approximately 1000 babies are born in the UK each year affected by a cleft of the lip and/or palate, an incidence of about 1:700 live births[1]. In 1998, The Clinical Standards Advisory Group (CSAG) recommended the reorganisation of UK cleft care into nine Regional Specialist Managed Networks with established links to maternity units and obstetric departments. This has now been achieved and, for the main, specialist teams are in post and recommended links have been made.

The successful care of any child within the first postnatal year has an important long term impact on both the physical and emotional health of that child. Children born with an oral cleft require the skills of their parents and those of the local and specialised health teams. Early care is facilitated by early diagnosis and the prompt referral to a regional cleft unit.

National standards of care and ongoing support in the postnatal period

A set of national standards has been agreed by the Nurse Special Interest Group, Cleft Lip & Palate (SIGCLP) in order to provide a responsive, informed service to families

- Standard 1: All babies born with a cleft lip and/or palate are to be diagnosed at birth
- Standard 2: All babies are to be referred by a relevant professional to the cleft team within 24 hours of diagnosis, including antenatal diagnosis
- Standard 3: A Clinical Nurse Specialist (CNS) should visit within 24 hours of receiving the referral, or where there has been an antenatal diagnosis, make contact within 24 hours
- Standard 4: All babies should have a nationally agreed feeding assessment prior to the introduction of assisted feeding.
- Standard 5: At the first visit a feeding plan which supports the mother's preference for feeding, should be devised and documented.
- Standard 6: All mothers who choose to breast feed should be offered an electric pump for as long as they require, at no cost to themselves
- Standard 7: All parents are offered :
  – Counselling and support
  – Verbal and written information
  – Contact with a family of a child with a similar diagnosis
  – Written information about the parents' support group (CLAPA)
- Standard 8: All babies affected by cleft palate should be referred to a local audiology service
- Standard 9: All babies should be visited at home by the CNS within one week of discharge

TABLE 1 National standards for care of a baby with an oral cleft.

immediately following the birth of a baby with an oral cleft (**TABLE 1**).

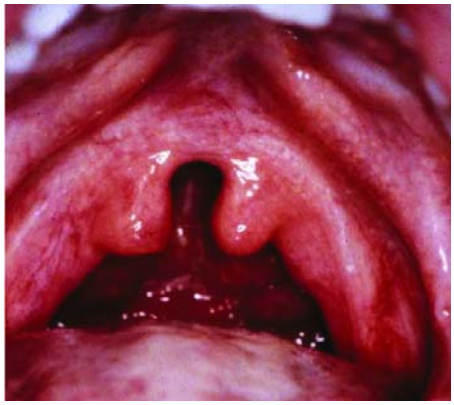
Care pathways have been developed by the regional cleft teams to ensure that the child and their family are introduced to the relevant specialist at appropriate times of assessment and treatment. A multidisciplinary team should consist of clinical nurse specialists, counsellors, speech and language therapists, surgeons, audiologists, psychologists, coordinators and dental therapists, dentists and orthodontists[2]. Primary and secondary surgery is undertaken at agreed ages, in an established paediatric setting, by a surgeon who receives between 30-40 new referrals per year. Links with paediatricians, geneticists and local acute and primary healthcare professionals are important. Parents and children are central to this pathway and are encouraged to be involved in decisions regarding the available options for treatment. A close liaison with the Cleft Lip & Palate Association (CLAPA) provides parent-to-parent support and a national voice when strategic decisions are being made about the provision of care.

Diagnosis and classification of cleft type

Clefts involving the lip are increasingly being diagnosed in pregnancy, at the 20 week anomaly ultrasound examination. Not all scanning departments routinely scan the face and clefts involving the palate remain very difficult to identify by ultrasound particularly in the absence of a cleft lip.

Facial clefts vary considerably in their presentation but can be simplified into those involving the lip, with or without the palate, and those involving the palate only. A cleft involving the lip can present as unilateral or bilateral and complete or incomplete. The nose is often distorted due to the abnormal insertion of the obicularis oris muscle into the alar margin. The most minor form is known as a forme fruste. This may present as a noticeable line from the base of the nose and involve a distortion of the lip vermillion. A cleft lip may occur in isolation or involve the primary and/or secondary palate[3]. This type of cleft is usually immediately identified at birth because of its visual impact.

In contrast isolated clefts of the palate can be easily missed at the initial birth examination. In a recent national audit



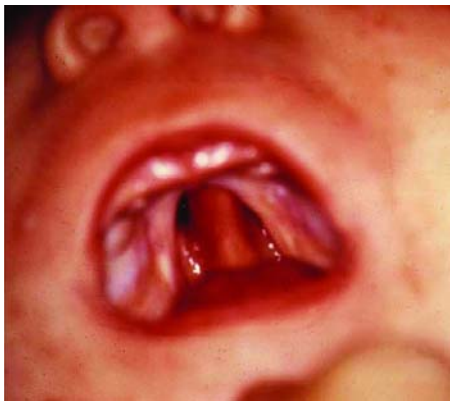


FIGURE 1 Examples of isolated cleft palate: Bifid uvula (left) and cleft palate involving the bony hard palate.

undertaken by the SIGCLP nurses, involving 963 babies born in the UK during a one year period, 23% of isolated clefts of the palate were missed at this examination[4]. This result highlights the inadequacy of the current digital examination, using a finger sweep of the palate, and identifies the importance of visualisation of the whole palate from the alveolar ridge back to the uvula with a torch and spatula.

Isolated clefts of the palate vary in size and shape. They may involve the posterior muscular soft palate and uvula only or include the bony hard palate (**FIGURE 1**). A submucous cleft of the palate occurs when there is incomplete muscle union across the velum of the posterior palate, but an intact oral mucosal surface. A baby with this type of cleft often presents as having feeding difficulties, failure to gain weight and nasal regurgitation of milk and, in the older child, speech problems. On examination a bifid uvula with a central translucent area through the posterior palate and a palpable notch at the union of the hard and soft palate can be good diagnostic markers. However a proportion of the population presenting with a bifid uvula, which is asymptomatic, will not require any further intervention.

Influence of genetic factors

At the present time clefts involving the lip or isolated to the palate only, can be divided into syndromic or nonsyndromic categories. A nonsyndromic cleft is diagnosed when affected individuals have no other associated anomalies, no family history, no environmental or teratogenic exposures known to predispose to clefting and normal cognitive and physical development. Infants with an oral cleft may present with other anomalies which are not associated with the cleft, but the differential diagnosis should involve a clinical geneticist for a full clinical assessment of the baby and their family history.

To date there are about 400 syndromes associated with clefts, some of which are very rare and others which run in generations of families and can be very subtle in their presentation. Where there is a cleft involving the palate only, the frequency of additional anomalies is much higher than for clefts involving the lip with or without a cleft palate. The literature doesn't always make this clear so an overall quoted figure of 60% additional anomaly rate, quoted by Shprintzen[5], can be misleading and lead to unnecessary investigations. Syndromes such as Sticklers and Velo Cardio Facial syndrome (22q11 deletion) are generally associated with isolated cleft palate and may have relatively mild additional manifestations, which can be easily overlooked and may only become apparent with increasing age.

Where there are no other markers further investigations may be unnecessary. However if other markers are present routine chromosomes may be helpful and where there is an isolated cleft of the palate with another abnormality a FISH test for 22q11 deletion is recommended. It is advisable that all infants affected by clefts have a cardiac assessment by a senior paediatrician and if a murmur is identified a cardiac echo should be arranged. Infants who present with an isolated cleft of the palate and have a family history of congenital non progressive myopia, detached retina and early onset of arthritis, require an ophthalmic assessment within the first six postnatal months to investigate possible Sticklers syndrome. Specialist genetic advice can offer targeted

chromosomal analysis and appropriate invasive testing.

A common disorder is Pierre Robin sequence, defined as an isolated cleft palate in association with micrognathia and glossoptosis. Upper airway obstruction occurs in varying degrees and may be assessed on a continuum of severity. Airway management becomes the priority for management and an approach to management will be considered in the second of these articles.

Unexpected neonatal behaviour, neurological deviations identified on feeding assessment and abnormalities identified on thorough paediatric examination are all indicators of possible problems. The presence of a syndrome can have a profound effect on a baby's growth potential and may influence early nutrition and ability to oral feed in infancy and must be considered as significant in the early management of these babies. Not all neonatal problems which occur concurrently with a cleft are of genetic origin. Additional factors which involve early management may be related to obstetric influences and difficulties, immaturity and parental support, adjustment and the development of compensatory parenting.

Early clinical considerations

- Identification of cleft at birth
- Paediatric examination by senior paediatrician
- Referral to regional cleft team at time of diagnosis
- Upper airway management if appropriate
- A lateral position for sleeping where palate involved
- Early assessment of oral feeding skills
- Appropriate investigations if other markers
- Consider involvement of clinical geneticist
- Support to parents

Immediate support

The attitudes of miwives, neonatal nurses and other staff profoundly affect parents' emotions and behaviour in the immediate postpartum and postnatal period. Parents are particularly sensitive to the responses of others and watch and evaluate the behaviour of those around them at this time[6]. Although parents may be in shock, they are often immensely protective of their newborn infant and any adverse remark, whether made directly or implied about the baby, might result in parents rejecting any further communication with that person. Where there is an unexpected anomaly the normal baby is often lost in the pursuit for other problems and it is not uncommon for parents to express an observation that professionals talk only of possible problems not of the normal aspects of their baby.

When a baby is born with a cleft, parents may experience feelings and reactions they did not anticipate and possibly have never experienced before. Such emotions are unlikely to have previously been built into their own construct system [7]. As parents gather resources these intense feelings dissipate[8] and they begin to reorganise and adjust to their parental role, which if successful results in acceptance. The time taken to reorganise their construct system varies for both parents and in this early period often occurs in an atmosphere of uncertainty.

An antenatal diagnosis will have allowed parents and their families time to adjust during the antenatal confinement, but the birth may initiate feelings of guilt and sadness about negative thoughts that they may have had before the birth. Whether the diagnosis is before or after birth, parents will often require the professional to "hold"[9] in a trusting relationship, whilst the process of reconstruction occurs. It is important that parents are given accurate information about their child's cleft and at a pace which is directed by the parents, in a language they can understand. In many regions parent-trained volunteers are available to offer parent-to-parent support and they can be identified either through the cleft team or CLAPA.

Early support and management

- Normal acknowledgement of their baby and successful birth
- Simple explanation of the problem and the normal aspects of their baby
- Avoid inaccurate information, but reassure parents the condition can be corrected
- Avoid panic reactions which can increase parental anxiety
- Assist skin to skin contact and normal exploration of the baby
- Encourage involvement of father – if not immediately available, offer to inform him
- Explore possible difficulty for father in informing relatives
- Establish mother's choice of feeding method and assist feeding if appropriate
- Acknowledge emotional response of both parents and encourage feelings to be expressed
- Avoid explanations which may be insensitive to parental feelings at the time
- Avoid stereotypical responses
- Be aware of your own feelings towards facial disfigurement and seek supervision if required
- Recognise initial dependency and only withdraw as parents regain control
- Avoid separation of mother and baby
- Discuss immediate plan of care with early referral to cleft team using agreed protocol
- Telephone cleft team for early advice and referral

Following discharge home a clinical nurse specialist from the regional team will continue to support, counsel and manage the ongoing feeding through the developmental stages of the first eighteen months. For all new parents the postnatal period from birth to two months is one of adaptation[10] and successful adjustment has been achieved when a mother expresses pleasure in her infant and in her own mothering activities.

Parents of babies with clefts need time and counselling to come to terms with unexpected feelings and help them reconstruct their approach to parenthood. They may move between successful adaptation and mal-adaptation throughout the childhood years if periods of treatment do not have the expected outcomes. The aim of this support is to help parents through the adjustment process, with frequent assessment and teaching to empower families to regain control of their baby's management. Following discharge home arrangements will be made for the infant and parents to meet the specialist multidisciplinary team within six weeks. A care plan will be discussed with parents/carers and times for surgery discussed. Timing may be affected by the type and size of the cleft and the presence of other anomalies which may take priority or affect care. Nurse led pre and postoperative assessment ensures that infants present for surgery having had the appropriate clinical screening, in a healthy condition with an adequate body weight. Infants are usually visited at home within a week following surgery to

assess recovery, the wound, re-establish normal feeding patterns and review the inpatient experience.

Conclusion

Although oral clefts vary considerably in their presentation, the early management of these babies can be successfully separated into two categories. The second of these two articles will consider some of the problems which can occur in infancy for both babies born with a cleft involving the lip and those with an isolated cleft palate.

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