Early care services for babies born with cleft lip and/or palate

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Introduction

In Europe around one in 700 babies is born with cleft lip and/or palate, the most common congenital anomaly of the head and neck region. The incidence is approximately 1,6 per 1,000 live births, but there is some variance across Europe [1]. Estimates indicate there are over 900,000 individuals (babies, children and adults) with clefts in Europe [2] - a significant figure, especially when one considers that not only the patients but also their families are affected in terms of psychosocial adjustment and having to endure the burden of a long treatment pathway.

In round figures the incidence by type of cleft may be summarised as follows [3]:

Table 1 — Incidence of Type of Cleft [1]

Type of Cleft	Percent of Total
Cleft palate only	50%
Cleft lip (± alveolus) only	20%
Cleft lip and palate	20%
Bilateral cleft lip and palate	10%

In some cases the cleft may be associated with other problems which need specialist management and these need to be identified early [4]. Accurate diagnosis (antenatal or post natal), the provision of appropriate information and support for the family, and the establishment of a structured care pathway, especially in the early months, will ensure that these infants thrive and develop like all other children. Access to good treatment varies widely throughout Europe, meaning that many children born with clefts are never given the opportunity to realise their full potential. The concept of a comprehensive specialist-team approach to care is not universal. Furthermore babies with clefts are still institutionalised in some countries in Europe [5].

The aim of this report is to provide an informative document which can be used by those countries where national protocols need to be established.

1 Scope

This Technical Report specifies recommendations for the care of babies born with cleft lip and/or cleft palate at time of diagnosis (ante- and/or postnatal) and the year following birth or diagnosis (whichever is later), including referral processes, establishment of feeding, parental support and care pathways.

Recommendations on all aspects of surgery, including timing and the use of pre surgical orthopaedics is excluded.

[1] For further information on different types of cleft see Annex A.

2 Terms and definitions

For the purpose of this document, the following terms and definitions apply.

2.1
assisted feeding
use of a soft, squeezable, bottle and/or adjusted teat and/or sipper spout to allow delivery of milk to the infant who is unable to generate suction to extract fluid independently

Note 1 to entry: It enables the infant to feed, effectively and safely, the required volume within an acceptable time frame.

Note 2 to entry: For further information on types of bottles and teats used for assisted feeding of babies born with clefts see Annex C.

2.2
cleft centre
hospital with a designated cleft team and paediatric facilities

2.3
cleft surgeon
surgeon trained in cleft surgery with a major commitment to cleft care and who practices cleft surgery on a regular and frequent basis

2.4
cleft team
multidisciplinary team which comprises the following members with proven competence in their field of expertise, paediatric experience and a major commitment to cleft care: A care coordinator/manager of the service; a surgeon trained in primary cleft surgery; a surgeon specialising in secondary cleft surgery such as bone grafting and orthognathic surgery; an orthodontist; a speech and language therapist; a nurse specialising in cleft care; a psychologist with recognised clinical training; an audiologist; an ENT surgeon; a geneticist; a restorative dentist; a paediatric dentist; a dental technician

Note 1 to entry: Whilst not all specialities will be required for every patient, access to all these practitioners is available when needed.

Note 2 to entry: If patients receive some aspects of care nearer home (e.g orthodontics, speech and language therapy) they receive care by trained specialists working in collaboration with the cleft team.

2.5
Eurocleft
Eurocleft Project 1996 – 2000 funded by the European Commission having the aim to improve management and understanding of cleft lip and palate and create a network of European researchers and clinicians to facilitate information exchange

2.6
cleft support organization
non-medical group with paid staff and/or volunteers offering advice and support to families affected by cleft

2.7
nurse specialising in cleft care
specialist responsible for planning and coordination of early cleft care, including feeding assessment, development of a feeding plan, providing support to the family and liaising with other health care professionals

Note 1 to entry: In the absence of a nurse, these services are provided by another trained professional with similar skills.

Note 2 to entry: An illustrative role of a nurse specialising in cleft care is provided in Annex D.

2.8
palatal plate
presurgical orthopaedics
intraoral appliance which can be used in the treatment of infants with cleft palate

Note 1 to entry: It can help with feeding and can be used to influence the maxillary growth and therefore to create better conditions for surgery.

Note 2 to entry: In newborns with Pierre-Robin-Sequence a plate with velar extension can be helpful to treat upper airway obstruction and avoid tracheostomy.

2.9
Pierre Robin Sequence
sequence of symptoms present in a newborn; micrognathia (small lower jaw), cleft palate, and glossoptosis (posteriarly placed tongue) that combine to cause airway obstruction

Note to entry: The back of the tongue falls back , particularly when the baby is supine, and occludes the airway.

3 Diagnosis and referrals

3.1 Antenatal diagnosis

3.1.1 Recommendations on making the diagnosis

The fetal face can be studied with ultrasound very early in gestation. Examination of the fetal face should be a component of guidelines for second trimester ultrasound examination [6].

3.1.2 Limitation of ultrasound

Although the correct ultrasound examination technique should enable clefting of the upper lip to be identified, it is difficult to detect an isolated cleft palate antenatally [7]. What is often described as a 'cleft palate' on ultrasound may be just a cleft alveolus. Training of the professional undertaking the ultrasound diagnosis should include information on basic embryology of clefting and how treatment of clefts is managed.

3.1.3 Suspected cleft lip and/or palate

If a cleft has been suspected, there should be a referral to a specialist in ultrasound diagnostics to confirm the diagnosis. As a cleft lip and/or palate may be associated with other anomalies, early assessment and diagnosis is necessary.

Clear information about cleft lip and palate should be given to families if a cleft is suspected. Genetic counselling for patients and families should be available on request. The main topic for discussion is the association with other anomalies and the recurrence risk for following pregnancy.

If the cleft is part of a complex syndrome, specific information about it and about the fetus viability should be given to parents during the meeting.

In the absence of other anomalies, a fetus with a cleft does not require a change in standard obstetric care.

3.1.4 Recommendations on referral to the cleft team

The following recommendations should be followed:

— a referral from the antenatal diagnostics unit to a multidisciplinary cleft team and to a cleft support organisation (where it exists) should be made as early as possible after diagnosis. The referring unit should ensure it maintains up to date contact information on the local or regional cleft team(s);

— parents should be contacted by a member of the cleft team as soon as possible after receiving the referral; and

— parents should be offered a face to face meeting with a member of the cleft team within one month of the confirmed diagnosis.

The aim of the referral to the cleft team is to provide parents with support and counselling, giving clear information about treatment and successful management of cleft lip and palate. Families don't generally expect the diagnosis or know anything about cleft lip and palate and they have a lot of questions and concerns about bringing up a child with a cleft. Some families may be thinking about terminating the pregnancy.

Psychosocial support for parents who are expecting a child with a cleft should be available. Parents may need help in normalising the pregnancy and understanding that the cleft does not define the baby. The following topics may need addressing:

— grief adjustment process;

— possible feeling of guilt;

— impact on the family;

— how to handle reactions from others; and

— whether or not it will be possible to breast feed.

Parents should be given contact details for the cleft team and be informed that the team is able to provide additional information at any stage during the pregnancy. The information needs of families after a diagnosis of a cleft are provided in chapter 11.

3.2 Postnatal diagnosis

3.2.1 Recommendations on making the diagnosis

Diagnosis of a cleft palate is often missed. Proper assessment of the palate is of paramount importance:

— a palpation of the palate is not sufficient on its own. Routine assessment of all newborn babies should include a visual inspection of the palate with a torch and spatula to depress the tongue;

— every newborn baby with a cleft should be seen by an experienced paediatrician as soon as possible.

3.2.2 Recommendations on referrals

Each maternity unit should designate a staff member to liaise with the cleft team to ensure that all staff are familiar with the contact arrangements and maintain an up to date knowledge and understanding of current practice in providing care for children with a cleft lip/palate.

Neonatal intensive care should be used only where specific needs justify it.

The maternity unit should provide basic information on cleft care such as general explanation of the condition, the reasons for treatment, treatment options, location of the cleft team, and details of support groups. This may be supplemented by leaflets, booklets or other kinds of information – websites, posters, and phone numbers.

Involvement of the cleft team around the birth will be necessary whether or not co-morbidities are present, such as Pierre Robin Sequence (see annex B).

All babies should be referred by relevant professionals to the cleft team as soon as possible after diagnosis. A cleft team member (a nurse specialising in cleft care where they exist) should visit at the earliest opportunity after receiving the referral. The team member will visit the mother at the maternity unit to observe the child during a feed, offer advice and supply any feeding equipment considered necessary.

4 Immediate Postnatal Care

4.1 Clinical assessment

It is important that a baby with a cleft lip/palate has a full clinical assessment by a neonatologist or paediatrician to rule out any possible comorbidities or associated syndromes. As a guide, the likelihood of co-morbidities according to cleft type are shown in Table 2:

Table 2 —– Co-morbidities associated with cleft type [8]

Cleft type [1]	***Association with co-morbidities (%)***
cleft palate only	*42.3* [2]
cleft lip and palate	*23.5*
cleft lip only	*7.6*
[1] Cleft types are described in Annex A. [2] Most commonly these are airway obstructions (Pierre Robin Sequence) or cardiac abnormalities	

If co-morbidities are present there needs to be a consideration as to whether the baby should be referred to neonatal intensive care. A referral to a genetics team at this stage may also be considered. The appropriateness of early feeding should be assessed and further investigations should be undertaken if required.

All children with cleft palate only should be assessed for airway obstruction and possible Pierre Robin Sequence prior to commencing oral feeding (see 4.1.1)

In the presence of a congenital heart anomaly consider 22q11 deletion syndrome, the second commonest syndrome after Pierre Robin Sequence associated with cleft palate only.

If no co-morbidities are found, the child can be treated as a well baby.

4.1.1 Pierre Robin Sequence – managing airway obstruction

The airway obstruction in these babies can be managed in a variety of ways including positioning (e.g nursing the baby in a side lying position), use of intra oral plates and insertion of a nasopharyngeal airway – see Annex B). Tracheostomy and jaw distraction in the absence of additional anomalies should be avoided. Pierre Robin Sequence is associated with Sticklers Syndrome so it is recommended that these babies have a genetic and ophthalmological assessment to exclude this.

4.2 Early parental involvement

The post natal period provides a window for parent/child bonding. This important aspect of early care should not be overlooked in the prioritising of clinical care and support for a child born with a cleft. The following recommendations should be respected:

— the child should be kept with the mother on the postnatal ward unless there are co-morbidities necessitating a neonatal intensive care environment;

— both parents should be involved in the care of the child and they should be kept informed of their child's diagnosis and treatment; and

— parents may need additional support to adjust to their child's diagnosis and help with the bonding process.

5 Feeding

5.1 Knowledge and skills of person providing feeding advice

Benchmarks for training should include the following items:

— paediatric skills;

— counselling skills; and

— knowledge of normal and abnormal feeding mechanisms.

5.2 Clinical assessment

Feeding should only be commenced after a full clinical assessment has been performed (see 4.1)

Premature babies may not manage oral or assisted feeding and may require nasogastric feeding initially. Oral feeding from the breast or a normal bottle can be commenced if there are no co-morbidities.

A feeding assessment should be undertaken by a health professional with special knowledge of feeding mechanisms and skills within cleft before commencing assisted feeding. Children without co-morbidities should be able to safely feed orally.

A detailed checklist for a clinical assessment of feeding is provided in Annex E.

5.3 Feeding plan

Prior to discharge of the mother and baby from hospital, the priority is to enable effective feeding of the child. A nasogastric tube should be used only in cases where comorbidities or specific needs exist. In some countries a palatal plate may be used to facilitate the feeding process.

The following recommendations should be followed when establishing a feeding plan:

— the cleft team should be involved in all decisions about feeding;

— a feeding plan should be developed preferably supporting the mother's preference for feeding;

— mothers should be encouraged to breast feed where possible;

— if assisted feeding is necessary, parents should be encouraged to use expressed breast milk;

— skin to skin contact to promote lactation and breastfeeding/breast milk feeding should be encouraged;

— both parents should be involved in all aspects of feeding and should be the principle feeders;

— appropriate tools (bottles/teats) and techniques to assist feeding should be available and parents instructed how to use them (see annex C);

— equipment to support the giving of expressed breast milk should be available where possible; and

— a cleft team member (a nurse specialising in cleft care where they exist) should ensure the baby receives necessary follow up and feeding reviews after discharge from hospital.

Additional support should be offered to parents with the introduction of solid foods at 4 – 6months.

5.4 Growth Measures

It is important that growth is closely observed and that the baby thrives and is well enough for first surgery. Length, head circumference and weight should all be monitored.

When monitoring the weight of the baby, the following information should be recorded:

— birth weight (noting any prematurity), and

— initial weight loss (loss of more than 10% of birth weight is abnormal).

Weight should be recorded at the following intervals:

— weekly for first month of life;

— two weekly for second month; and

— monthly until 6 months.

Some children may require increased calories and/or the temporary use of a nasogastric tube to maintain a healthy weight gain (particularly those with Pierre Robin Sequence and other co-morbidities).

6 Monitoring the baby and preparation for surgery

6.1 Monitoring

Ongoing monitoring of the child's care needs is imperative and should include input from the cleft team (cleft nurse if available). The following provides a checklist for assessment and action:

— continued paediatric surveillance for any co-morbidities such as cardiac anomalies, airway issues and associated syndromes;

— referrals should be made to other health care professionals as necessary i.e. Cardiologist, E.N.T, Geneticist etc.;

— a hearing test should take place as soon as possible after birth and again preoperatively. Repeat testing should take place at 18 months old, 3 years, 5 years and 10 years and as clinically indicated

— continuous assessment of dental and facial development should be provided by an orthodontist familiar with maxilliary and mandibular growth patterns. Dental health should also be monitored.

6.2 Preparation for Surgery

The following arrangements should be in place to ensure the child and family are prepared for surgery:

— pre surgery orthopaedics may be considered and this should be explained to the parents;

— all parents should have the opportunity of visiting the ward at the surgical centre prior to any surgery being undertaken and be offered counselling and support if needed;

— parents should be offered a consultation with the cleft surgeon who will perform the primary surgery, and all patients should be seen in outpatients at least once prior to any surgery being undertaken;

— an audiological assessment should be undertaken to determine whether ear tubes (grommets) should be inserted during surgery;

— an assessment should be made by the cleft team of the child's safety for surgery;

— the surgery in children should be done under general anaesthetic with a well trained paediatric anaesthesiologist;

— there should be additional clinical assessments prior to surgery if any other anomalies are present e.g. sleep observation (airway issues) and cardiac ECHO review.

— Informed parental consent should be sought prior to surgery.

7 Recommendations for involving and supporting parents

7.1 Involving parents

Shared decision-making about treatment has the potential to improve health outcomes, improve patient satisfaction and to save costs. However, there are barriers to involving parents: appointments may take longer, parents may find it hard to challenge decisions made by health professionals, health professionals may find it hard to have their decisions questioned.

Therefore, patient involvement should be built into the care process:

— parents should be encouraged by health professionals to take part in decisions about treatment and enough time should be allowed for this at medical appointments;

— parents should be helped to weigh up the pros and cons of different treatments;

— health professionals should give parents the opportunity to feedback on the care they have received and the outcomes of treatment; and

— parents should have access to timely information to help them make decisions about treatment. The quality of the information is important and it should be accurate, unbiased and evidence based (See 11.1).

7.2 Parent to parent support

Parents can find it helpful to receive support from other parents who have been in the same situation. This can include both practical and emotional support but should not include medical advice which should only be given by health professionals. This support is valuable from the time of diagnosis where parents share photos of their babies before and after surgery and through the whole treatment pathway.

The development of a parent-focused cleft support organisation should be encouraged. This organisation should be able to:

— provide non-medical advice and support to parents;

— create opportunities for parents to contact other parents either face to face, online or by telephone;

— bridge the gap between the parent and the healthcare provider; and

— act as an independent monitor of cleft care services.

The cleft team should provide information about the cleft support organisation to all parents at the time of first diagnosis and at other key stages of treatment.

Local, national and international patient support organisations can be helpful in supporting families through the long treatment process.

8 The longer term care pathway

8.1 Scope of a care pathway

Whilst the focus of this report is on the early care of babies born with clefts, it is important to understand that cleft care will last from birth to maturity, and in some cases well into adulthood. There are many aspects of care and treatment required. The overall objective of a cleft service is to:

- co-ordinate the total care of the patient from infancy through to adulthood;
- ensure that the cleft lip and /or palate is well repaired and the child‘s growth, development and function maximised;
- put in place arrangements for ongoing care and treatment of adult patients where clinically indicated;
- ensure that care is organised and provided in such a way that it:
  - respects the needs and wishes of the child, parent or guardian;
  - recognises the nature of the original birth anomaly;
  - optimises the psychological well-being of the patient and family;
  - facilitates speech and language progress;
  - ensures optimal hearing;
  - optimises facial appearance; and
  - enables appropriate and dental growth.

8.2 Long term care pathway

Care pathways will differ across different units. Table 3 contains a general guide illustrating the key interventions normally provided from birth to maturity, at specific age brackets, across different disciplines.

Table 3 — Care Pathway

Antenatal	Birth to 1,5 years	1,5 to 5 years	6 to 10 years	11 to 20 years	21 years +
Ultrasound diagnosis confirmed. Referral to cleft specialist team. Antenatal counselling and support. Provision of written information. Information on feeding options and presurgical infant orthopaedics (if used) Information about patient groups.	Diagnosis confirmed at birth. Assessment of presence of co-morbidities. Referral to cleft team. Visit by specialist advisor/nurse specialising in cleft care. Feeding assessment. Feeding plan. Meeting offered with parent/patient group. Registration on anomaly database. Post natal support and counselling. Provision of written information. Speech and language therapy assessment and advice. Pre-surgery visits. Pre-surgery infant orthopaedics (if used) Supported by nurse specialising in cleft care. Surgery according to local timing protocols. Routine dental care. Early orthodontic assessment. Audiological assessment. Psychological assessment and intervention where necessary.	ENT and audiology assessment and treatment. Speech and language therapy assessment and advice at 2-3 years, therapy as required. Psychological assessment and intervention where necessary. Paediatric dental care. Audit at 5 years. Possible revision surgery and velopharyngeal surgery. Routine dental care. Orthodontic assessment	ENT and audiology assessment/treatment. Speech and language therapy assessment and therapy as required. Paediatric dental care. Review of overall management. Audit data set at 10 years. Orthodontic care and treatment. Secondary surgery – alveolar bone graft. Possible revision surgery and velopharyngeal surgery. Routine dental care. Psychological assessment and intervention where necessary.	Orthodontic care and treatment. Preparation for possible orthognathic surgery. Psychological assessment and intervention where necessary. Paediatric and restorative dental care. Audit data set at 15 years and 20 years. Possible revision surgery and velopharyngeal surgery. Routine dental care. Genetic counselling.	Possible orthognathic surgery. Psychological assessment and intervention where necessary. Routine dental care.

9 Recommendations for the cleft unit team members and facility requirements

9.1 Team members

The composition of the cleft team is according to the definition of the term in 2.4.

9.2 Education and training

A main cleft centre should provide specialist staff training for all staff on the cleft team. Lead clinicians should be able to demonstrate a commitment to continuing professional education and training for all staff involved in the network of provision of cleft care. Where surgical and orthodontic trainees participate in care this should be as part of a defined training programme and in high volume centres.

9.3 Team management and responsibilities

9.3.1 Communication and coordination

The principal role of the multidisciplinary team is to provide case management to assure quality and continuity of patient care and follow-up. Each patient seen by the team requires comprehensive, multidisciplinary individualised treatment planning to achieve best outcomes with efficient use of parent/caregiver and patient time and resources. Teams should have a mechanism for achieving consensus with involved professionals and parents on treatment plans. Patient records, including images and x-rays, should be transferable in case of any family geographical movements.

9.3.2 Referral and communication with other professionals

The team should have a process for referring patients to local care providers when necessary and appropriate. The team should implement a process for information exchange with primary care professionals, outside agencies, and other professionals involved with the welfare of the patient. The team should also ensure referring bodies (such as diagnostic and obstetric units) are kept up to date with team contact details. Cleft teams should make a special effort to maintain contact with the paediatrician/family doctor of each child to support patient follow-up and coordinate healthcare needs, for as long as the patient needs it.

9.3.3 Research

Cleft teams should be encouraged to carry out research into treatment and prevention of cleft lip and palate and related syndromes in order that the management and treatment is evidence based.

NOTE 1: For further reference see ACPA guidelines in bibliography

10 Recommendations for organization of the cleft service, including clinical governance and audit

10.1 Service requirement

The complexities of cleft care management and treatment, from post natal care to adulthood, requires that the service should have:

- access to High Dependency Units (HDU)/Paediatric Intensive Care Units;
- access to paediatric surgical area including recovery area;
- dental clinics which facilitate orthodontics and paediatric dental services;
- access to ENT/Audiological services;
- computer support which facilitates web based data entry for submission to cleft registers/databases; and
- appropriate facilities for inpatients and multidisciplinary clinics (number and type of rooms, play areas, waiting areas) and facilities should at least meet paediatric standards.

10.2 Facilities for parents and children

10.2.1 Out-patient medical and dental care

Ideally, children receiving out-patient hospital care or dental care should be seen in a separate children's clinic area which is suitably equipped. If this is not possible, and children have to be seen during an adult clinic session, their appointments should be "clustered" preferably at the beginning of the session. A discrete area in the adult department should be designated for the exclusive use of children. Toys and diversionary activities should be available. The clinic staff should include a cleft specialist nurse or a nurse experienced and skilled in the management of children.

10.2.2 In-patient care

Resident paediatric staff, experienced in the management of fluid balance, post-operative pain control, and other postoperative problems should be available at all times.

The care of children who, in very exceptional circumstances, are not accommodated in a comprehensively staffed children's unit, or who are dependent on nursing care, should be provided by nurses who have up-to-date training in paediatric resuscitation.

The surgery in children should be done under general anaesthetic by a well-trained paediatric anaesthesiologist. The anaesthesiologist should have adequate opportunity to assess the child before surgery and there should be appropriate facilities and equipment available for post-operative monitoring. [9].

10.3 Clinical governance

The cleft team should act according to a strict protocol agreed by the team. The protocol describes the stages of treatment and also the assessment/evaluation of treatment outcomes and record taking.

10.4 High quality standards

The number of patients referred to the team should be sufficient to sustain the experience and specialist skills of all team members. For all members of the cleft team cleft care should be a major part of their clinical work. The Eurocleft guidelines [10] recommend no fewer than 30 new patients annually – this is based on the minimum number of patients required to undertake effective audit and comparison of outcomes. This has been endorsed by the World Health Organisation.

10.5 Audit, outcome measures and comparative studies

10.5.1 Monitoring short-term and long-term treatment outcomes

The team should document its treatment outcomes, including base-line performance and changes over time. Teams should conduct periodic retrospective or prospective studies to evaluate treatment outcomes and patient and family satisfaction. Evaluation of outcomes should also include patient satisfaction and family/care giver experiences using validated questionnaires.

Robust systems for clinical audit, agreed by the lead clinician(s), should enable collaboration at regional, national; and international levels.

The evaluation should be scheduled at regular intervals, the frequency and specific content of each of those evaluations being determined by the condition and needs of the individual patient. Monitoring treatment progress allows the updating of treatment plans.

The audit should be done in the team and also between teams. A multi-centred approach to comparing outcomes provides sufficient numbers of cleft sub types to produce meaningful results which can lead to recommendations for future good practice.

The results of evaluations should be publicly available in an accessible format.

National and international professional associations are also essential for facilitating communication between centres and disciplines, and also for disseminating research findings.

The Eurocleft recommendations on timing and scope of audit are displayed in Annex F.

10.5.2 National data sets

10.5.2.1 National Registers and Databases

The establishment of national registers or databases should be a priority. These are used to record cleft births and provide demographic data as well as detailing treatment protocols and outcomes. National registers or databases can be useful at governmental and non-governmental level for ongoing statistical analysis (incidence versus resources available) and enable comparisons of best practices across Europe.

10.5.3 Future developments

10.5.3.1 International collaboration

The monitoring and evaluation of outcomes in cleft care should be optimised through a system of national registers listing consecutive patients over their treatment lifespan and detailing outcomes. This would permit collaborative research at an international level to identify the best treatment protocols which should then be adopted by nationally designated centres of excellence in cleft care. To reduce the burden of care on patients, routine speech therapy and orthodontic treatment could continue to be provided near the patient's home but they are to receive care by trained specialists working in collaboration with the cleft team at the designated centre.

10.5.3.2 Costing cleft care

To ensure sufficient resources are available for basic cleft care there should be attempts to quantify the direct and indirect cost of cleft treatment at a national and international level.

11 Information and education needs

11.1 Families/Care givers

The information needs of the family will vary according to age of the infant/child and stage of the treatment pathway

11.1.1 At time of diagnosis (pre and post natal)

Cleft teams should provide information which includes:

— description of types of cleft and brief aetiology;

— before and after surgery photos;

— feeding methods;

— treatment pathways and what to expect at each stage of treatment;

— the long term impact of having a cleft;

— implications for planning future children; and

— introduction of the multidisciplinary team.

It is recommended that verbal information is backed up with written material either in leaflet form or with details of downloadable information from the internet. An approved list of websites should be provided. Full details of local, regional and national cleft support organisations should also be routinely provided, along with details on how to access contact information for national and international cleft teams.

11.1.2 Preparation for surgery

Advice and guidance shall be given to parents in preparation for any surgery. Information given should include:

— what the child will look like immediately following surgery;

— duration of operation;

— how pain will be managed;

— when they can visit the child in the recovery room;

— how long the child will remain in hospital;

— expected recovery trajectory post surgery; and

— care required following discharge after surgery.

11.1.3 Longer term care pathway

Teams should assist parents/caregivers in making informed decisions on the child's behalf and preparing the child and themselves for all recommended procedures. Parents/caregivers should be given information about recommended treatment plans and any alternatives, benefits, and risk factors. The mechanism for this communication can be either verbal or in writing and should follow after each team evaluation.

11.2 Health professionals

Midwives, nurses, paediatricians, general practitioners, dentists may all experience patients who have a cleft. Recommendations should be made to medical training schools to include an element on diagnosing and treatment of clefts. Front line health professionals should have:

— a basic understanding of cleft lip and palate;

— knowledge of how to “break the news” and explain the condition to parents;

— knowledge of care pathways so they can refer to cleft specialists at the appropriate time; and

— instructions on how and where to refer to specialist cleft services and support groups.

11.3 Social services and education providers

Bodies and institutions routinely involved with early child support and education should understand:

— the physical and psychological impact of having a cleft;

— how to direct families to appropriate support and treatment;

— that a child with a cleft will thrive like any other child given the right medical care and early assistance; and

— how the treatment pathway affects access to education.

11.4 Governments, healthcare service providers and the general public

Societal attitudes to clefts in some countries may need to be challenged. Government-led public awareness campaigns may be needed to remove the taboo and stigma of having a cleft. A better understanding of cleft lip and palate amongst the general public will ensure prospective parents are better able to cope with a diagnosis of cleft for their baby. In addition, health care service providers need to understand the complex treatment pathway and long term implications of having a cleft when designing and funding services.

12 Information production

Access to good quality information improves parents' ability to care for their child and take an active role in their treatment, empowering them to ask the right questions.

All scripted information should be clear, accurate, impartial, evidence-based and up-to-date.

Information should be presented in a range of formats and in appropriate languages, e.g. video, written material, downloadable documents, presentations, training courses, or websites.

Information should be free for parents and families and should be appropriate to their individual needs and treatment stage.

Information producers should consult with their target audience on topics, content and accessibility.

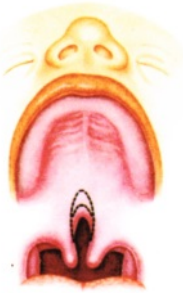
All information should include the patient experience through case studies and training should include presentations from parents and patients.

Information about specialist cleft services should be actively promoted to ultrasound specialists and maternity units.

Annex A
(informative)

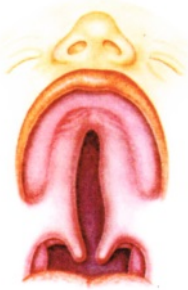
Types of cleft lip and/or palate[2]

Incomplete cleft lip



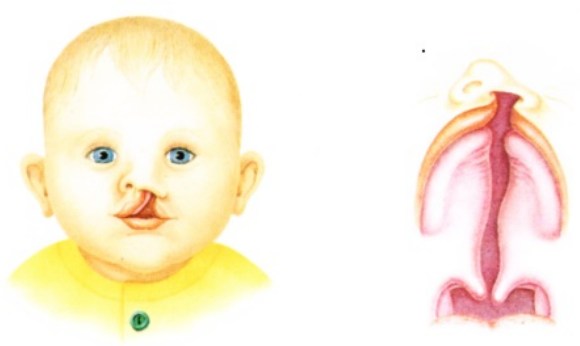
Cleft of the soft palate

Complete cleft lip
(alveolus (gum) not involved)

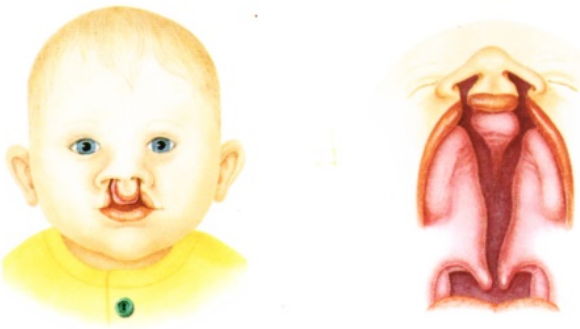


Cleft of the soft & hard palate

[2] Illustrations from Cleft Lip and Palate Association, UK



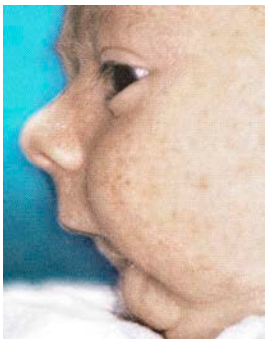
Unilateral cleft lip and palate
(alveolus involved)



Bilateral cleft lip and palate
(alveolus involved)

Annex B
(informative)

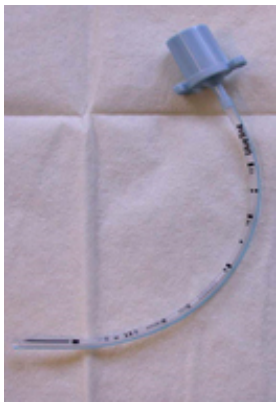
Pierre Robin Sequence and treatment options (clause 4.1.1)



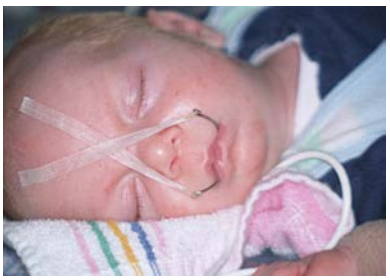
Pierre Robin Sequence occurs when a baby is born with a small lower jaw (micrognathia), a cleft palate and a tongue positioned toward the back. The tongue can fall into the throat causing total or partial airway obstruction, frequently heard as a ‘snorting’ or ‘grunting’ sound.

Suggested treatment options

1) An effective way to make breathing easier is to use a Nasal Pharyngeal Airway (NPA). This is a shortened breathing tube made from an adapted endotracheal airway (see illustration) which passes over the tongue and sits just above the epiglottis (wind pipe).



2) Another option recommended in some cleft centres is the use of plate with a velar extension (e.g Tübingen Plate) which is used to prevent the tongue slipping back and blocking the airway [11]



Annex C
(informative)

Bottles and teats used for assisted feeding of babies with cleft

C.1 Example of a squeezable bottle

Soft plastic enables gentle squeezing of the bottle to assist with feeding.

C.2 Example of squeezable teat

Sometimes it is the teat rather than the bottle which is squeezed.

C.3 Example of a sipper spout

A spout can be used on a bottle instead of a teat.

Annex D
(informative)

Description of the role of a nurse specialising in cleft care

A nurse specialising in cleft care is a core member of the multidisciplinary cleft team. The nurse will demonstrate a sound understanding of the issues related to the identification, assessment, treatment and management of cleft lip and palate patients, including those with associated anomalies, and play a key role in the management of the families with cleft babies throughout the treatment period. Specifically, the role of a nurse specialising on cleft care may include the following aspects:

— to be responsible for the ante-natal referrals made to the cleft service, reporting diagnostic findings and providing information and counselling regarding treatment, birth planning and feeding preparation;

— to undertake counselling around complex and sensitive issues e.g. the diagnosis of the cleft and other complex conditions;

— to assist in making a diagnosis and clinical assessment of newborn babies with clefts. They may present a variety of feeding, and respiratory difficulties;

— to undertake early clinical assessment and feeding assessment, including developing a feeding plan and providing necessary equipment (specialist bottles, breast pumps);

— to monitor the baby's feeding, growth and development over the first few months;

— to provide counselling, education and information for the parents and family;

— to undertake peri-operative assessments, offering support, information and advice to families pre and post surgery;

— to plan the longer term care and management of the needs of babies with clefts and additional complex conditions, in partnership with local paediatric services;

— to act as a liaison between hospital and home, providing information and advice to families, health professionals and extended carers, e. g. community nurses, paediatricians, patient support groups, obstetricians, ultrasonographers, general practitioners, health visitors midwives, ward nurses; and

— to actively support the transitional care of young people into adulthood through an agreed care pathway.

Annex E
(informative)

Sample form used for feeding assessment of babies with cleft

FEEDING ASSESSMENT OF BABIES WITH CLEFT

KEY: One red, seek guidance from core nurse.
Three or more orange, seek guidance

NAME .....

GENDER ☐ male ☐ female

DATE OF ASSESSMENT ..... **ASSESSING NURSE** .....

BIRTH HISTORY .....

TYPE OF CLEFT ..... **GESTATION** .....

MORBID CONDITIONS ☐ **yes** ☐ no

.....

OBSERVATION OF NEONATE

DYSMORPHIC FEATURES ☐ yes ☐ no

MUSCLE TONE ☐ normal ☐ **floppy**

MICROGNATHIA ☐ yes ☐ no

TONGUE POSITION ☐ anterior ☐ mid oral ☐ **posterior** ☐ **protruded**

AIRWAY MAINTAINED IN SUPINE ☐ yes ☐ **no**

TRACHEAL TUG **Supine** ☐ yes ☐ no **Prone** ☐ yes ☐ no

STERNAL RECESSION **Supine** ☐ yes ☐ no **Prone** ☐ yes ☐ no

SIGNS OF CYANOSIS ☐ yes ☐ no

COMMENTS .....

.....

NON NUTRITIVE ASSESSMENT

ORAL REFLEXES: Rooting ☐ yes ☐ no **Gag** ☒ yes ☐ no **Sucking** ☐ yes ☐ no

TONGUE MOVEMENT ☒ cupped ☐ humped ☐ protrudes ☐ flaccid

STRENGTH OF SUCK ☒ strong ☐ moderate ☐ weak

NO. OF SUCKING BURSTS ……

NO. OF SUCKS PER BURST ……

INFANTS RESPONSE ☐ relaxed ☐ tense ☐ avoidance ☐ gagging

☐ arm flailing ☐ sweaty ☐ cyanotic

NUTRITIVE ASSESSMENT

SUCKING PATTERN OVER 1 MINUTE

No of bursts .....

No of sucks per burst .....

Swallow triggered ☐ yes ☐ no

Sustained sucking ☐ yes ☐ no

Regular rhythm ☐ yes ☐ no

Suck/swallow/breathe co-ordination ☐ good ☐ poor

INFANTS RESPONSE ☐ relaxed ☐ tense ☐ avoidance ☐ gagging

☐ arm flailing ☐ sweaty ☐ cyanotic

KEY: One red, seek guidance from core nurse.
Three or more orange, seek guidance

SAFE TO PROCEED TO ORAL FEEDING ☐ YES ☐ NO

TYPE OF FEEDING :

- **BREASTFEEDING**
  - State of the breast ……
  - Milk flow ……
  - Position during the breastfeeding ……
  - Equipment to support breast / breast milk feeding e.g nipple shield, breast pump ……

- **BOTTLE FEEDING**
  - Rigid bottle
    - Type .....
    - Type of the nipple .....
    - Size of the nipple .....
    - Position of the holes .....
  - Soft bottle
    - Type .....
    - Type of the nipple .....
    - Size of the nipple .....
    - Position of the holes .....
    - Assistance required
      – no of squeezes per burst ……
      -- length of squeeze ……

- **COMBINATION NG/ORAL FEEDING**
  - Number of feeds through NG tube .....
  - Quantity .....
  - Number of oral feeds .....
  - Quantity .....
  - Duration of feed .....
  - Timing of feeding .....

- **NG TUBE FEEDING**
  - Number of feeds .....
  - Quantity .....
  - Timing .....

OTHER RECOMMENDATION .....

.....

.....

.....

THE SAFEST MODEL OF FEEDING CHOSEN AT THE MOMENT

.....

DATE OF NEXT FEEDING ASSESSMENT .....

Annex F
(informative)

Recommendations on treatment records

The following are consensus recommendations from the EUROCLEFT project [10] and are included as a general guide to record taking. It is acknowledged that in some countries records are routinely collected at first clinic visit, prior to first surgery.

F.1 Minimum treatment records

The team should document the procedures, its treatment outcomes and also changes over time. The timing and nature of the records depend on the clinical protocols followed by individual teams. Comprehensive records on each patient must include histories, diagnoses, treatment plans and reports of evaluation, all in a database. Supporting documentation may include photographs, radiographs, dental models and audio taped speech records. Cleft patients should be followed up routinely until 18-20 years of age. In order to compare results and monitor effectiveness of treatment it is important to have an agreed data set of what to record and at which intervals.

All patients' records must include the following elements:

— speech records;

— study models;

— relevant radiographs;

— clinical photographs;

— videotaped investigation of velopharyngeal function;

— audiometry;

— psychosocial assessment[3]; and

— oral health.

[3] The timings of psychological assessments are missing fron theEurocleft tables

F.2 Timing of minimum records

Table F.1 — Complete Cleft Lip, Alveolus and Palate (UCLAP & BCLAP)

Timing	Models	Lateral skull radiograph	Photographs	Speech	Audiometry/tympanometry
Primary surgery	√		√		
3 years				√*	√*
5/6 years	√		√	√	√
10 years	√	√	√	√	√
18+ years	√	√	√	√	

* If hard palate is closed.

Table F.2 — Complete Palate Only (CPO)

Timing	Models	Lateral skull radiograph	Photographs	Speech	Audiometry/tympanometry
Primary surgery	√		√		
3 years				√	√
5/6 years	√			√	√
15/16 years	√	√	√	√	√

Table F.3 — Cleft Lip Only (CLO)

Timing	Models	Photographs
Primary surgery	√*	√
3 years		
5/6 years	√*	√
10 years		
18+ years		√

* Only in patients with cleft of the alveolus as well as cleft lip.

F.4 — Alveolar Bone Grafting (ABG)

Timing	Intraoral radiograph	Photographs
Just before bone graft	√*	√
6 months after graft	√	
After canine fully erupted	√	√

F.5 — Pharyngoplasty

Timing	Speech
Just before operation	√
One year after operation	√

F.6 — Orthognathic Surgery

Timing	Models	Lateral cephalogram
Just before operation	√	√
One year after operation	√	√

F.6 — Psychological assessments[4]

Timing	Psychology
5 years	√
10 years	√
15 years	√
18+ years	√
Parent	√

F.7 — Psychological assessments: Orthognathic Surgery[5]

Timing	Psychology
Pre-operative	√
1 year post surgery	√

[4] This table has been added to the Eurocleft recommendations

[5] This table has been added to the Eurocleft recommendations

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