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

Fürsorgedienstleistungen für Babies mit Lippen-, Kiefer- und Gaumenspalten

Services de prise en charge précoce des bébés nés avec une fente labiale et/ou palatine

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EUROPEAN COMMITTEE FOR STANDARDIZATION
COMITÉ EUROPÉEN DE NORMALISATION
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Foreword

This document (CEN/TR 16824:2015) has been prepared by Technical Committee CEN/TC 424 “Project Committee - Care services for cleft lip and/or palate”, the secretariat of which is held by ASI.

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Introduction

In Europe around 1 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 summarized 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 realize their full potential. The concept of a comprehensive specialist-team approach to care is not universal. Furthermore babies with clefts are still institutionalized 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) For further information on different types of cleft see Annex A.

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.

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 specializing in secondary cleft surgery such as bone grafting and orthognathic surgery; an orthodontist; a speech and language therapist; a nurse specializing in cleft care; a psychologist with recognized clinical training; an audiologist; an ENT surgeon; a geneticist; a restorative dentist; a paediatric dentist; a dental technician

Note 1 to entry: While 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 specializing 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 specializing in cleft care is provided in Annex C.

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 improve physiological tongue position and to influence maxillary growth in preparation 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 invasive surgical procedures, e.g. tracheostomy.

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

Note 1 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 foetal face can be studied with ultrasound very early in gestation. Examination of the foetal 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 foetus viability should be given to parents during the meeting.

In the absence of other anomalies, a foetus 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 organization (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 do not 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 normalizing 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 Clause 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 specializing 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

4.1.1 General

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 [a]	***Association with co-morbidities (%)***
cleft palate only	*42,3* [b]
cleft lip and palate	*23,5*
cleft lip only	*7,6*
[a] Cleft types are described in Annex A.	
[b] 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 4.1.2).

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.

4.1.2 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 or other invasive surgical procedures in the absence of additional anomalies should be avoided. Pierre Robin Sequence may be associated with Sticklers Syndrome so it is recommended that these babies have a genetic, ENT 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 prioritizing 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

These 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 care 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.