



National Guideline for Cystic Fibrosis Nurses for the First Contact Telephone Call

Changing practice to support service delivery

Guideline for CF Nurses for the First Contact Telephone Call

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This document must be read and used in conjunction with:

An Bord Altranais (2000) Scope of Nursing and Midwifery Practice Framework. An Bord Altranais, Dublin.

An Bord Altranais (2000) The Code of Professional Conduct for Each Nurse and Midwife. An Bord Altranais. Dublin.

An Bord Altranais (2002) Recording Clinical Practice Guidance to Nurses and Midwives. An Bord Altranais. Dublin.

Health Service Executive (2007) Code of Practice for Health Care Records Management. Health Service Executive: Dublin.

Health Service Executive (2009) Services for People with Cystic Fibrosis in Ireland. Conclusions of a Working Group established by the HSE. HSE. Dublin.

An Bord Altranais (2010) Practice Standards for Midwives. An Bord Altranais. Dublin

Cystic Fibrosis Association of Ireland (2005). Towards a Better Service; the Treatment of Cystic Fibrosis in Ireland; Problems and Solutions. (Pollock Report). Cystic Fibrosis Association of Ireland. Dublin.

This is a controlled document and is subject to change before the review date should the need arise.

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Abbreviations

ANP	Advanced Nurse Practitioner
CF	Cystic Fibrosis
CNM 1	Clinical Nurse Manager 1
CNM2	Clinical Nurse Manager 2
CNS	Clinical Nurse Specialist
SECM	Self Employed Community Midwife
HSE	Health Service Executive
IRT	Immunoreactive Trypsinogen
NBS	Newborn Bloodspot Screening
NSCF	Newborn Screening for Cystic Fibrosis
NNBSL	National Newborn Bloodspot Screening Laboratory
NNBSP	National Newborn Bloodspot Screening Programme
NSC	Newborn Screening Card
PHN	Public Health Nurse

1. Background

Newborn Screening for Cystic Fibrosis (NSCF), when associated with early treatment, improves nutrition in childhood, reduces the burden of care for families and may improve survival (Castellani *et al*, 2009). An early diagnosis of Cystic Fibrosis (CF) has a beneficial effect on nutritional status, with improved growth, height and weight, and may prevent deficiency of fat-soluble vitamins and protein malnutrition (Castellani *et al*, 2009). Equally an early diagnosis of CF results in less parental stress compared to a delayed diagnosis (Castellani *et al*, 2009).

In June 2009, a National Steering Group was established to oversee the development and implementation of newborn screening for cystic fibrosis in Ireland. The HSE National Service Plan 2011 outlined that newborn screening for CF would be introduced as an integral component of the existing National Newborn Bloodspot Screening Programme (NNBSP).

There are six designated specialist CF centres offering care to which babies will be referred following a positive Newborn Bloodspot Screening (NBS) result. These six centres are located at Our Lady's Children's Hospital, Crumlin; Adelaide and Meath Hospitals incorporating the National Children's Hospital, Tallaght; the Children's University Hospital, Temple Street, Dublin; Cork University Hospital, Cork; University College Hospital, Galway; and the Midwestern Regional Hospital, Limerick. The Clinical Liaison Officer in the National Newborn Bloodspot Screening Laboratory will contact the CF Nurse in the designated CF centre with the positive result relating to CF. Following consultation with the CF Consultant, the CF Nurse will then make a telephone call to inform the parent(s) that the result of the heel prick test requires them to bring their baby into the designated centre the following day.

Between 72 and 120 hours after birth the heel prick test for newborn screening is carried out by the Midwife/Nurse in the Maternity Hospital/Unit, the Public Health Nurse (PHN) or the Self Employed Community Midwife (SECM). The blood sample collected from

the heel prick test is collected onto the Newborn Screening Card (NSC) as a blood spot. The NSC is completed by the Midwife/Nurse in the Maternity Hospital/Unit, PHN or SECM in advance of taking the sample. The details are verified by the parent(s) and consent is given. The sample is forwarded to the National Newborn Bloodspot Screening Laboratory (NNBSL) for testing.

A positive CF NBS result is not a diagnosis and should always be confirmed by a diagnostic test for CF (Comeau *et al*, 2007). A confirmatory sweat test is required for the diagnosis of CF and should be conducted on all babies with a positive CF NBS result, including those with two mutations detected (Comeau *et al*, 2007). A positive CF NBS result means the infant is at increased risk of having CF. The specific risk depends on the category of the positive result as indicated below. The result will be reported as:

“CF suspected” - when IRT > 99th centile and two mutations identified. The infant most likely has CF and requires diagnostic testing (i.e. a sweat test) to confirm the diagnosis of CF, and a clinical assessment by a CF consultant.

or

“Single CF associated mutation identified” - when IRT > 99th centile and one mutation identified. The infant has about a 1/16 chance of having CF and requires further testing (i.e. a sweat test) to either confirm, or rule out, the diagnosis of CF, and a consultation with a CF consultant.

2. Policy Statement

It is the policy of the Health Service Executive (HSE) that CF Nurses in the six designated specialist centres, employed by the HSE or other facilities providing care on behalf of the HSE, utilise this document as guidance to their practice. This national guideline refers specifically to the First Contact Telephone Call by a CF Nurse to a parent(s) following a positive result for their baby relating to CF. The guideline has been collaboratively developed with input from the six designated specialist centres and key stakeholders. The aim of the guideline is to provide standardised, evidence based guidance to support and guide the CF Nurse in making the First Contact Telephone Call

with the ultimate aim of providing clarity and supportive care to parent(s) at this time. It is the responsibility of each CF Nurse in the six designated specialist centres to read and comply with this guideline.

3. Rationale

Clear explanation to families of the process involved in screening and the implications of normal and abnormal results is central to the success of the NSCF programme. Effective communication by the CF team is especially important when parents are told that their baby is affected or is a carrier. When establishing a NSCF programme, attention should be given to ensuring timely and appropriate processing of results, to minimise potential stress for families (Castellani *et al*, 2009).

4. Purpose

This guideline aims to provide clear, evidence and consensus based guidance to nurses and midwives on the provision of information to service users when making the first contact telephone call following a positive result for a baby relating to CF.

5. Scope

The scope of this guideline applies to:

- 5.1** Registered general and children's nurses in the CF department/unit of the six designated specialist CF HSE centres
- 5.2** The implication of this guideline refers to newborn babies, parents, employees of the HSE or other facilities providing CF services on behalf of the HSE

6. Definitions

- 6.1 CF Nurse** – for this document the term CF Nurse refers to all nurses working in any one of the 6 designated specialist CF centres.
- 6.2 Consultant** – for this document the word Consultant refers to the Paediatric Consultant with a special interest in CF

- 6.3 The heel prick test** – this is a test that will be carried out between 72 and 120 hours following birth. A small sample of blood is obtained from the heel of the newborn baby. This blood is placed on a NSC and sent immediately by registered post or by courier to the NNBSL in the Children’s University Hospital, Temple Street, Dublin.
- 6.4 IRT** refers to immunoreactive trypsinogen. It is an enzyme produced in the pancreas and is raised in the blood of newborn babies with CF. IRT levels are measured in the bloodspot taken as part of the routine NBS programme. It forms the initial test in all CF NBS programmes. A raised IRT level is not diagnostic for CF; further testing is required such as genotyping and sweat testing in conjunction with a clinical assessment, to make a definitive diagnosis of CF.
- 6.5 National Newborn Bloodspot Screening Programme (NNBSP)** is a national public health screening programme that aims to identify certain serious medical conditions shortly after a baby is born. Most of these conditions have no immediate visible effect on the baby, but without treatment serious consequences can ensue. However, early effective treatment is available. The sample to date has traditionally been analysed for five conditions:
- Phenyketonuria
 - Congenital hypothyroidism
 - Classical Galactosaemia
 - Homocystinuria
 - Maple Syrup Urine Disease
- CF will be included in the NNBSP from 1st July 2011.
- 6.6 Cystic Fibrosis** - CF is Ireland's most common life-threatening inherited disease. Approximately 1 in 19 people are carriers of the CF gene and where two carriers parent a baby together, there is a 1 in 4 chance of the baby being born with CF (CFAI Annual Report 2009/10). CF affects many organs including the lungs, the

pancreas, the digestive tract and the reproductive system. It causes thick sticky mucus to be produced, blocking the bronchial tubes and preventing the body's natural enzymes from digesting food. The result is that people with CF are prone to chest infections and malnutrition. However as therapeutic options have expanded, significant advances have been achieved in both life expectancy and quality of life.

- 6.7 Mutation** – A mutation is a permanent alteration to a gene in a person's genetic code. The mutation damages the gene, and so the protein the gene codes for, is either absent or not functioning.
- 6.8 Sweat Test** - The sweat test measures the amount of chloride in sweat. There are no needles involved in this test. A sweat test is required for the diagnosis of CF. As part of the NNBSF the sweat test will be performed at a designated specialist centre. The sweat test can be carried out from three weeks of age onwards. However, some babies may not produce enough sweat to measure chloride levels. If a baby does not produce enough sweat the first time, the test should be repeated.
- 6.9 Clinical Assessment** - The clinical assessment of a baby with possible CF involves the Consultant Paediatrician taking a detailed history and performing a thorough physical examination.
- 6.10 Diagnosis** – the diagnosis of CF is made by a Consultant Paediatrician specialising in CF following a clinical assessment of the baby, interpretation of the results from the NSCF and the sweat test.

7. Roles and Responsibilities

- 7.1 Clinical Liaison Officer (NNBSL)** – is responsible for confirming the demographic data on the NBS form with the appropriate maternity service to

determine the relevant CF centre to which the baby should be referred. S/he identifies the parent's preferred first language and contacts the CF Nurse with the result by telephone and by faxing a hard copy of the data to the centre with the baby's contact details and the NBS result, including the identified CF mutations.

- 7.2 CF Nurse** –The CF Nurse is responsible for discussing a positive CF NBS result with the consultant and for informing the parent(s) of the need to bring the baby for assessment. S/he is responsible for arranging the sweat test and liaising with the consultant. The CF Nurse is also responsible for informing the Clinical Liaison Officer (NNBSL) of the result of the sweat test and confirming that the baby has been seen and assessed by the consultant who confirmed the diagnosis. S/he will fax a report to the Clinical Liaison Officer (NNBSL) as per local policy. Registered nurses / midwives must at all times practice within his/her scope of practice (An Bord Altranais, 2000).
- 7.3 Director of Nursing/Midwifery** is responsible for the provision of nursing staff in the CF department / unit, supporting the relevant nursing personnel to attend the Continuing Education Programme regarding the introduction of NSCF and the specific issues relating to the Guideline for the First Contact Telephone Call by CF Nurses. S/he is also responsible for the implementation of the Guideline for the First Contact Telephone Call by CF Nurses in the designated centre including support of staff to attend any required continuing education.
- 7.4 Consultant** – will liaise with the CF Nurse with regard to a positive NSCF result and agree a time for meeting the parents and undertaking a clinical assessment of the baby. S/he is responsible for the diagnosis of CF and for the overall care of the baby. It is important that the Consultant communicates with the parent(s) to ensure s/he has a clear understanding of the diagnosis The Consultant is responsible for referring the parent(s) for genetic counselling, where a baby is identified as being a carrier of a CF mutation.

8. Guidance

8.1 *Pre Call*

- 8.1.1** The aim of the First Contact Telephone Call is to contact the parent(s) of a baby who has been identified through the CF NBS process as having either one, or two, CF causing mutations, and to organise an appointment the following day for a sweat test and an assessment by the Consultant.
- 8.1.2** Prior to making the call the CF Nurse will speak with the Consultant to ensure the consultant is available the following day.
- 8.1.3** Prior to making the call the CF Nurse will organise the sweat test
- 8.1.4** Prior to making the call the CF Nurse will ascertain from the information supplied by the NNBSL Liaison Officer whether or not English is the preferred first language of the parent(s). In the event that English is not the preferred first language the CF nurse will contact an official interpreter for the First Contact Telephone Call and for the subsequent visit. The CF Nurse will brief the interpreter about the situation before the telephone call.

8.2 *The Call*

- 8.2.1** The CF nurse who makes the call will have participated in the Continuing Education Programme regarding the introduction of NBS for CF and the specific issues relating to the Guideline for the First Contact Telephone Call by CF Nurses.
- 8.2.2** The CF Nurse will speak clearly and identify herself as a nurse from the named hospital.
- 8.2.3** The CF Nurse will ascertain she is speaking to the parent(s) of the correct baby by checking the name of the baby, the date of birth and location of birth of the baby and the address with the parent(s).
- 8.2.4** The CF Nurse will refer back to the NBS (heel prick test) and documentation given to the parent(s) at antenatal and postnatal visits and ask the parent(s) how much they remember from what they were told. In supporting the parent(s) s/he

will actively listen, acknowledge concerns, provide meaningful and accurate answers and allow silences where appropriate.

- 8.2.5** The purpose of the call is to request that the parents attend an appointment for a sweat test and a clinical assessment. It is not to give a diagnosis. The CF Nurse will deliver the message that the heel prick result indicates that the baby has an increased risk of having CF and now requires a sweat test and assessment the following day so that a diagnosis for CF can either be confirmed or ruled out.
- 8.2.6** The desired outcome of the call is that the parent(s) understands the purpose of the call and the importance of attending for the visit the next day. This is potentially the beginning of a long and trusting relationship between the parent(s), the baby and the CF Nurse. S/he will endeavour to answer any questions in an open and honest manner.
- 8.2.7** Details of when and where the appointment will take place will be given.
- 8.2.8** The CF Nurse must emphasise clearly the importance of the sweat test / assessment and ensure that the parent(s) is aware of their responsibility to attend with their baby for the test and clinical assessment the following day.
- 8.2.9** The CF nurse will suggest that the parent comes to the appointment with their partner or a support person and comes prepared to stay at the hospital for a number of hours. This allows time for the completion of the sweat test, the results to come back and their appointment with the consultant and the CF nurse.
- 8.2.10** The CF Nurse may suggest to the parent(s) that they write down any questions or concerns that they have and bring them to the hospital the following day.
- 8.2.11** The CF nurse may direct the parent(s), if asked, to a dedicated website for newborn screening/sweat test (CFAI or HSE website) and explain that a lot of information on the internet is not accurate and there will be an opportunity at the visit for a full discussion.
- 8.2.12** In the event that the parent is distressed and unable to communicate, the CF Nurse may ask for the name and number of a support person and will request permission from the parent to contact that person.

8.2.13 In exceptional circumstances, as agreed with the consultant, alternative arrangements may be made for an assessment within a locally agreed timeframe and this will be clearly documented.

8.2.14 The CF Nurse will endeavour to reassure the parent(s) and speak in a calm and clear manner.

8.2.15 The CF Nurse will ask the parent(s) have they written down the instructions for attending the appointment and will ask the parent(s) to repeat back what they have written down.

8.3 *Post Call*

8.3.1 In the event of a parent(s) calling the CF Nurse back following the First Contact Telephone Call, the CF Nurse will approach the follow-up call in a similar manner to the initial First Contact Telephone Call.

8.3.2 In the unlikely event of a parent(s) refusing to attend for the appointment or to have any further interactions with the CF Nurse or the hospital, the CF Nurse will inform the consultant who will then pursue the matter according to local policy.

8.3.3 When the First Contact Telephone Call is complete, the CF Nurse will complete the documentation record (section 9) and file in the appropriate healthcare record.

9. Documentation

Documentation will comply with An Bord Altranais (2002) Recording Clinical Practice Guidelines for Nurses and Midwives, the Health Service Executive Code of Practice for Healthcare Records Management (2007) and other local policies / guidelines pertaining to documentation. It is proposed to use the following template to document the First Contact Telephone Call:

First Contact Telephone Call Record

Date: _____

Time call began: _____

Patient Details

Name:

DOB:

Address:

NNBSL Reference Number:

Details of Call from NNBSL

Name of Clinical Liaison Officer (NNBSL)	
Date and time of call	
Date and time of fax results received	

Interpretative Services

Preferred first language of the parent(s)?		
Is an interpreter required?	Yes	No
If yes, please give details		

Arrangements made prior to the call

Name of Consultant	Date & Time of visit
With whom was the appointment made for the sweat test?	Date & Time of Sweat test

Patient name and NNBSL reference number:

Please record the following from the telephone conversation

PLEASE CONFIRM	
1. Name of Mother:	
2. Baby's date of birth:	
3. Baby's place of birth:	
4. Address:	
5. Have you given your name and designated specialist centre?	
6. Informed the parent(s) the result of the heel prick test indicates further testing is required	
7. Explained the baby needs to come to hospital for a sweat test and an assessment tomorrow	
8. Asked the parent(s) to write down all the details of the visit	
9. Asked the parent(s) to read back details of the visit	
10. Was the name and number (and permission to contact same) of a support person obtained from the parent? If yes, please give details	
11. Are there any exceptional circumstances that would delay the visit?	
12. If so, were these discussed this with the consultant?	
13. Were alternative arrangements made? Please detail	

14. Did the parent(s) refuse to attend?	
15. If yes, was this documented and discussed with the consultant?	
16. Has an action been proposed? Please detail	

Time call ended: _____

Signed: _____

Please print name -

Comments:

For the purpose of audit and ongoing evaluation, please record after the First Contact Telephone Call, any questions the parent(s) asked during the call.

10. Implementation

10.1 Dissemination - The guideline will be disseminated by the Office of the Nursing and Midwifery Services Director to the Directors of Nursing and Midwifery in the six designated specialist centres. It is available to download from:

<http://www.hse.ie/eng/about/Who/ONMSD/Intro.html>

10.2 Education - All nurses working in the six designated specialist CF centres will be required to participate in a Continuing Education Programme regarding the introduction of NBS for CF and specifically issues relating to the Guideline for the First Contact Telephone Call by CF Nurses. Education will be provided as an initial immersion programme by the Centre for Children's Nurse Education, and thereafter will be available on an annual basis or when required.

11. Review and Audit

The guideline will be reviewed in June 2012. This is a controlled document and is subject to change before the review date should the need arise.

12. References

An Bord Altranais (2000) Scope of Nursing and Midwifery Practice Framework. An Bord Altranais. Dublin

An Bord Altranais (2002) Recording Clinical Practice Guidelines for Nurses and Midwives. An Bord Altranais: Dublin.

Castellani C., Southern K.W., Brownlee K., Roelse J.D., Duff A., Farrell M., Mehta A., Munck A., Pollitt R., Sermet-Gaudelus I., Wilcken B., Ballamann M., Corbetta C., de Monestrol I., Farrell P., Feilcke M., Ferec C., Gartner S., Gaskin K., Hammermann J., Kashirskaya N., Loeber G., Macek M., Mehta G., Reiman A., Rizzotti P., Sammon A., Sands D., Smyth A., Sommerbug O., Torresani T., Travet g., Vernooij A. and Elborn S. (2009) European best practice guidelines for cystic fibrosis neonatal screening. Journal of Cystic Fibrosis doi10.1016/j.jcf.2009.01.004.

Comeau A.M., Accurso F.J., White T.B., Preston W.C. III, Hoffman G, Parad R.B., Wilfond B.S., Rosenfeld M., Sontag M. K., Massie J., Farrell P.M. and O'Sullivan B.P. (2007) Guidelines for implementation of cystic fibrosis newborn screening programs: cystic fibrosis foundation workshop report. Pediatrics **119**(2), 495-518.

Cystic Fibrosis Association of Ireland (2010) Annual Report 2009/2010, Progress and Plans. CFAI, Dublin.

Health Service Executive (2007) Code of Practice for Healthcare Records Management. HSE. Dublin.

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