Great Ormond Street Hospital for Children
NHS Foundation Trust

Great Ormond Street
London WC1N 3JH
Tel: 020 7405 9200

Great Ormond Street Hospital Cystic Fibrosis Service

Quality Improvement Information Letter

We felt that it was time that we once again updated everyone on our quality improvement programme and also on some of our other research projects. Ongoing quality improvement is an integral part of our service – we are continually looking for ways in which we can improve the care we provide and ultimately improve outcome. Involving families who use the service is vital if we are to improve the care we provide and some of you will already have been or are involved in the projects below. We would like to take this opportunity to thank you all for your continued help and support.

QUALITY IMPROVEMENT (QI)

Improving network care

Many of you do not receive all of your cystic fibrosis (CF) care at Great Ormond Street Hospital (GOSH) but also attend your local hospital. We have been working to try and improve communication between the teams at local networks and ourselves. The work on our new clinical database has incurred some delay but is now in the final stages of development. We very much hope that by spring we will be recording all appointments in the same way and using the same system (whether at one of our networks or at GOSH) and information from these will be accessible between sites.

You may also be aware that the way in which CF care is funded has changed following a large piece of work by the CF Trust and Department of Health, which we were also involved in. We continue to work closely with the Specialist Commissioners and network hospitals to plan the future and put into place changes that have resulted from this work.

Contacting Us

We know it is very important that we are available to the families for advice and help. A very large number of telephone calls come through the unit every day and in order to deal with these efficiently and effectively it is important that you know the correct means of contacting us. We have introduced a new departmental e-mail which can be used for non-urgent queries and an information sheet about this is enclosed with this newsletter. Also enclosed is a sheet outlining all our contact details - you will see that there have been one or two changes so please take time to read this carefully.

Nuffield Collaboration

Our partnership with Nuffield Health and Wellbeing Centres has gone from strength to strength and some of you may have seen the short article in the Mail on Sunday. Nuffield continue to offer all our CF children free access to their gym facilities accompanied by one adult. They also offer highly competitive family membership rates. The GOSH physiotherapy team train the Nuffield trainers in the specifics of CF. Regular exercise is a vital part of CF care. All referrals need to be made via the physiotherapists at GOSH so if you would like further information on the Nuffield Collaboration please do not hesitate to contact us.

Website

This continues to be a 'work in progress' as we have not been able to dedicate as much time to it as we would like. However we very much hope to be able to work on it this year. It is already accessible and contains general information about the unit and some of our information leaflets are available to download.

Admissions to hospital

Another important project we are working on is the availability of beds for inpatient admission. Four of the beds on Badger Ward are now labelled as CF beds and this has alleviated the pressure on admissions to some extent. Badger ward will be moving toward the end of this year and we very much hope that our new location will have five CF beds with ensuite facilities.

We are also currently undertaking a big project on CF admissions to Badger ward – looking at how we might improve the admission process and experience. Ana Marote (Badger ward sister) and Denise Sheehan (CF Clinical Nurse Specialist) are working together on this project and some of you have already offered your help too. We will be canvassing opinion from families who are admitted to Badger to get your views on the admission experience and we will also be looking at our internal admission processes to see how these can be improved.

Newborn Screening Project

In the last Newsletter we wrote about a project that was being undertaken to evaluate the service we provide around the time of diagnosis (following the heel prick test). This project involved many of you giving up your valuable time to help us understand how you felt about the service and how we might improve it. We are very grateful to all of you who participated.

The findings indicated a few areas where we can improve:

- We are consulting with health visitors and other specialities to discuss how the first contact with families to inform them of the heel prick result is best made (recognising of course that this is never a "good" way of receiving this kind of news).
- The format of the very first visit to GOSH has been changed and families will in future meet the consultant and/or the clinical nurse specialist on arrival and before any tests are carried out.
- We are improving the written information we provide and will ensure that this is regularly updated. It will also be available on the GOSH website
- We are working towards changing the very first CF clinic appointment to ensure new families have longer time with the team.
- In future we will be mindful of the way we communicate information about research projects ensuring that this is done at a time point that is suitable and sensitive.

- Our weekly team meeting has been changed and in future the notes for children due to clinic that afternoon will be discussed to ensure that all team members are up to date.

There were also some things that were very much valued and we will ensure they continue:

- Our clinical nurse specialist and physiotherapist will continue to undertake appropriate home visits in the immediate period following diagnosis.
- The current Education Day (where families can gain information specific to their baby and ask questions they have) will not change.
- We will continue to ensure that all the CF team members have up to date knowledge of all CF treatments and new developments so that all the multidisciplinary team can provide appropriate support and advice.

RESEARCH PROJECTS

INSPIRE-CF

In May 2012, the CF Unit was awarded a GOSH Children's Charity Grant to fund a three-year research study, called INSPIRE-CF. This study will evaluate a new model of care for children with CF which incorporates regular exercise sessions. To date, 60 children aged six to 15 years have signed up to take part in the study. Half (50%) of the children in the study will be allocated to Group 1, the control group, and the others to Group 2, the exercise group. Children in Group 1 will continue to receive their usual programme of specialist CF care, while children in Group 2 will, in addition to this, receive a weekly supervised, 30-60 minute exercise training session. The study is in its early stages; however we will update you as the study progresses. The study is being led by Sean Ledger (Physiotherapist) and Dr Eleanor Main (Senior Lecturer in Children's Physiotherapy). Dr Paul Aurora is the Consultant supervising the research programme.

If you would like more information on the study you can contact the INSPIRE-CF team 0207 9052178 or 07500 918480.

Body Composition Study

Starting in October 2002 children between the ages of six and 12 were invited to take part in a study of their growth and body composition. One hundred children took part with measurements being done every two years using sophisticated techniques that are not generally available anywhere else in the UK.

The results of the first measurements have now been published and presented at several conferences. The findings are that as a group, the children with CF tend to be shorter than healthy children and the girls have less fat which does not seem to be related to delayed puberty. After two years, as a group, both boys and girls did not acquire as much lean tissue as children without CF and the girls still had less fat and slightly less bone mineral. This is the first time that deficits have be found in young children using a 'gold standard' technique - in the past we had to rely on simple measurements of weight, height and skin-fold and these do not really tell you how much overall fat and lean a child has. We hope that when the study has progressed further we will be able to identify how fat and lean are related to clinical outcome and therefore help treatment.

We would like to thank all the children and parents who have taken part and for continuing for as long as eight years – this study would not have been possible without your dedication. Another newsletter will be sent out in the near future with more details of the research but in the mean-time if you have any questions or would like a copy of the first paper please contact Jane Williams: jane.williams@ucl.ac.uk or 0207 9052743

London CF Collaboration Newborn Screening Study

Newborn screening (NBS) for CF was introduced across the UK in 2007, providing a much needed opportunity for early treatment to try and minimise the effects of the disease. However until we understand when the first signs of lung disease appear in NBS CF babies receiving the best routine care, it is difficult to design suitable clinical trials to see if new treatments are more effective.

Six hospitals form the London CF Collaboration and all were involved in this study, which was led from GOSH. Between January 2009 and December 2011, we invited all families with baby diagnosed with CF through NBS to take part in an observational study of lung function, lung structure (using chest CT) and lung infection (using bronchoscopy). Lung function was measured at GOSH/UCL Institute of Child Health at around three months, one and two years of age (we also measured lung function in healthy babies without CF from Homerton and University College London hospitals maternity units for a comparison). At one year of age the babies with CF also had a chest CT and bronchoscopy.

We have completed lung function measurements at three months and one year of age in 72 CF and 44 non-CF babies. When we compared the two groups, abnormalities of lung function were seen in almost half of the CF infants by three months. However, when the measurements were repeated at one year of age the babies with CF showed an improvement in lung function. Some aspects had improved such that it is no longer any different from babies without CF, while other aspects of lung function stabilised with no further deterioration. This is the first time any study has shown the benefits of CF newborn screening on lung function. Chest CT scans have shown very few structural changes and even when present, any changes are mild. These babies are currently being followed up to see whether these improvements are sustained throughout infancy and preschool years.

This study has enabled us to better understand the natural history of lung disease in CF babies diagnosed through newborn screening cared for with the best routine treatment at specialist CF centres. Through this, we may be able to identify babies most at risk of lung problems and who should be considered for more intensive treatment.

We would very much welcome your feedback on any of the projects above or on any general comments you have regarding the CF service at GOSH. Thank you again to everyone who has been involved in these projects and we very much look forward to working with you all further as the service moves forward. If you would like any more information on any of the issues raised in this newsletter please feel free to talk to us at your next clinic visit or contact us in the CF Office.

Ammani Prasad (CF Manager) & Ranjan Suri (Lead Consultant CF)
On behalf of the GOSH CF Team