This booklet is aimed to provide you with some information on Congenital Hyperinsulinism (CHI). We hope that by reading this, we will help you understand more about this complex condition. However, this booklet will not answer all your questions about CHI, so please feel free to talk to any member of our Northern Congenital Hyperinsulinism (NORCHI) team.

**What is CHI?**

Congenital Hyperinsulinism is a rare condition where the body makes too much of the hormone insulin from a gland in the tummy called the pancreas. CHI is the short term for Congenital Hyperinsulinism. The word ‘congenital’ means that the condition is present from birth. The word ‘hyperinsulinism’ means too much insulin production. The hormone insulin lowers sugar levels in normal people. That is, when the blood sugar levels are high, usually after a meal, insulin is made by the pancreas to bring down the sugar (glucose) level to normal. Normal levels are 4 to 6 mmol/l. However, children with CHI make too much insulin, causing blood sugars to drop to dangerously low levels. This is called hypoglycaemia. A blood sugar (or glucose) value of less 2.6 mmol/l is quite low. Such low sugar levels are not good for the brain, and may cause long term brain damage, particularly in babies and young infants. It is accepted that the blood sugar levels should be kept above 3.5 mmol/l to keep the brain in good condition. It is really important to recognise the diagnosis of CHI and correct the low sugars as soon as possible.

Children with diabetes also have a problem with insulin. In these children, insulin is low, so blood sugars are high. Therefore, diabetes is the opposite of CHI.
CHI is a rare condition, with 1 in 25,000 newly born babies being affected. These children often have very low sugar levels. Other children may have a milder type of CHI, which may even get better over time. However, at the time of diagnosis, it is not possible to predict which children get better and which children need more medication. Some children could have a very severe type of CHI. These children are often unresponsive to medication and the only treatment option is surgery to remove the pancreas.

What is the NORCHI service?

The Northern Congenital Hyperinsulinism (NORCHI) service was set up in the north of England to provide a highly specialised service for children with CHI. NORCHI is a joint service based in the two large children’s hospitals in Manchester (Royal Manchester Children’s Hospital) and Liverpool (Alder Hey Children’s Hospital). Your child has been referred to one of these hospitals for the treatment of low blood sugars and to find out why the pancreas is making too much insulin. He/she is likely to need some tests while the doctors and nurses
monitor the condition carefully. Your child may need a more concentrated sugar solution into the veins. Sometimes the extra sugar may be given as supplements in feeds. The NORCHI team will keep you updated every day about your child’s progress. However, please bear in mind that the treatment of CHI can be complicated and difficult. Your child may need to be in hospital for a long time, sometimes as long as several weeks. He/she is likely to need genetic tests to look for a ‘spelling mistake’ in the genetic code. Sometimes a special scan called a PET-CT scan may need to be done. The doctors will do their best to control your child’s condition without removing the pancreas. However, if the condition is not managed safely by medicine, surgery of the pancreas may have to be undertaken. All aspects of CHI will be discussed in detail with you. Please do not hesitate to ask for clarification about your child’s short term and long term care.

Why is it important for your child to receive prompt treatment for CHI?

Severe low blood sugars can cause many problems. In adults and older children, it causes tiredness and a feeling of restlessness. As the sugar levels drop to very low levels, the nerves in the brain are affected. Sugar is a crucial fuel for the brain. In the absence of sugars, for example when missing meals, the body is able to break down fat stores to form alternative brain fuels called ketones. However, in children with CHI, ketones are not formed. Therefore, in CHI both sugar and ketone levels are low, which means that the brain could be severely affected. This combination can cause fits and if left untreated, lead to coma and even death. If not treated correctly, the brain may be damaged in the long term. Such children may have fits on a regular basis (epilepsy). They could have learning problems and may also have problems with their eyesight. Therefore, early diagnosis of CHI is very important to prevent brain damage.
Team members and follow up clinics

The NORCHI team is multidisciplinary and consists of:

Consultant Paediatric Endocrinologists
Specialist Nurses
Psychologist
Dieticians
Speech and Language Therapists

The follow up clinics are also multidisciplinary and you may meet some or all the above members of staff in an out-patient clinic appointment.

Lindsey Rigby:
(0161) 701 2460

Louise Bowden:
(0161) 701 0518

Endocrine Department:
Monday – Friday 9 am to 5 pm

Manchester:
(0161) 701 1675

Alder Hey:
(0151) 252 5281

Dietitians and speech and language therapists:
Monday – Friday 9 am to 5 pm

Manchester:
(0161) 701 2640

Alder Hey:
(0151) 252 5231

Psychology:
Monday – Friday 9 am to 5 pm
(0161) 701 4514
No Smoking Policy

Please protect our patients, visitors and staff by adhering to our no smoking policy. Smoking is not permitted in any of our hospital buildings or grounds, except in the dedicated smoking shelters in the grounds of our Central Manchester site.

For advice and support on how to give up smoking, go to http://www.nhs.uk/smokefree.

Translation and Interpretation Service

It is our policy that family, relatives or friends cannot interpret for patients. Should you require an interpreter ask a member of staff to arrange it for you.

تُنصِّرنا سياستنا على عدم السماح لافراد عائلة المرضى أو اقاربه أو اصدقائهم بالترجمة لهم. إذا احتاجت الى مترجم فرجى اتصل بواحد العاملين ليرتب لك ذلك.

بمارى يه. باليسي بئ ک. خاندان ، رشت دار اور دوست مريضون کی لیے ترجمہ نہیں کرسکتے. اگرآپ کمترجم کی ضرورت بئ تو عملی کسی رکن سے یہ کہ بئ اپ کیے لیے اس کا بندوست کریے.

إِيْهَا أَئْمَادَٰرُ النَّبِيِِّ بِشَكَرٍ، اِلْكَانِشََِّ بِأَهْلِكَ مَا كَانَ أَنْبِيَادَكَ حَتَّى ضَرَبَهُمُ النَّبِيُّ نَبِيٗ.ً أَيْنَ مِنْ نَظَرٍ فِي عَالِمٍ إِلَّا ذَلِكَ.

اِلْهَمْ لَانَارَ اِلْكَانِشََِّ بِأَهْلِكَ مَا كَانَ أَنْبِيَادَكَ حَتَّى ضَرَبَهُمُ النَّبِيُّ نَبِيٗ.ً أَيْنَ مِنْ نَظَرٍ فِي عَالِمٍ إِلَّا ذَلِكَ.

آَمَّلُ أَنْ يَنِسَجَنَّ مَعْلِمَانِ عَلِيمَانِ نَجَّمِيِّ بِأَهْلِكَ مَا كَانَ أَنْبِيَادَكَ حَتَّى ضَرَبَهُمُ النَّبِيُّ نَبِيٗ.ً أَيْنَ مِنْ نَظَرٍ فِي عَالِمٍ إِلَّا ذَلِكَ.


Waa nidaamkeena in qoys, qaraaboamasaa xiiiboysanu tarjumikarina bukaanka. Haddii ay u baahatotarjumaanka codsoxubinka mid ah shaqaalahainaykuusameeyaan.

我们的方针是，家属，亲戚和朋友不能为病人做口译。如果您需要口译员，请叫员工给您安排。