Diagnosis and management of Cystic Fibrosis Related Diabetes (CFRD)

This guidance is recommended for use by staff looking after young people with CFRD or suspected CFRD. The following groups of children should be screened for CFRD using home glucose monitoring:

- All children over the age of 12 - screening to take place annually
- All children (of any age) where there is unexplained weight loss or loss of lung function
- All children (of any age) with symptoms of hyperglycaemia - tiredness, polydypsia, polyuria
- All children (of any age) with an HbA1c > 6.0%. HbA1c is measured in all children at annual review

Sometimes there is doubt about the need for insulin. This most often occurs when:

- HbA1c >48 mmol/mol (6.5%) without documented high BG levels on profiles
- HbA1c >42 mmol/mol (6.0%) with decreased lung function and/or weight loss and without documented high BG levels on profiles

In these circumstances continuous glucose monitoring (CGM) carried out in order to determine if insulin therapy is currently required and which regime would be appropriate.

Insulin therapy should be started when either of the following criteria is reached (on the glucose profile or CGM):

- HbA1c >42 mmol/mol (6.0%) with intermittent BGL >8.0 mmol
- 2 x fasting BG >8.0 mmol and/or 2 x post prandial BG >11.0mmol

A referral should be made to the diabetes team. Before insulin is started, a discussion **must** be had with the family by the CF consultant (preferably with the diabetes consultant) about the diagnosis of CFRD. If the young person is not already an inpatient when the decision to commence insulin therapy is made, they should be admitted to the ward to do this.

Insulin doses should be prescribed based on the following algorithm:

1. Is the fasting BG level above 8 mmol/l?
   - YES → Start Lantus 2 units (evening)
   - NO ↓
2. Are there post-prandial BG levels greater than 11 mmol/l? (apart from PEG feeds – see below)
   - YES → Start NovoRapid 2 units before relevant meal(s)
   - NO ↓
3. Is there a random pattern of BG levels >8 mmol/l during the day?
   - YES → Start Levemir 2 units (morning)
   - NO ↓
4. Are there BG levels above 8 mmol/l during or after PEG feeds?
   - YES → Start Humulin S 2 units pre PEG feed (Consider Continuous Glucose monitoring)
   - NO

Initial Education:

The diabetes specialist nurse will educate the patient and family in:

- Injection technique, site rotation
- BG (blood glucose) testing and recording. (They will be given a BG diary to complete -see 'monitoring' section.)
- Signs, symptoms and treatment of hypoglycaemia
- The family will be given the following targets:

Pre-meal BG level: 4-7 mmol/l
Post prandial BG level: 5-9 mmol/l
HbA1c: < 42 mmol/mol (6.0%)

Dietary advice

The dietician in the CF team should be made aware of the decision to start insulin therapy.
The high calorie diet should remain a priority and the patient should continue with regular meals and snacks. Considerations should be made with regard to timings of any supplement drinks. Encourage the young person to consume any full sugar/ energy drinks that are required with meals.

Monitoring

- BG testing:
  Once/twice daily insulin:
  - One day per week the patient should test regularly throughout the day.
  - If the young person is not managing to adhere to this, identify an area of the day where there is most concern and ask them to concentrate on testing at this point.

  Basal bolus:
  - The young person should test pre meal and before bedtime.
  - The young person may also need to do post-prandial testing periodically.

- HbA1c 3 - 4 monthly
- Eye screening and Microalbuminuria testing annually (at diabetes annual review clinic) if >12 yrs old.

Considerations with steroid use:

If the young person is prescribed steroids, they need to test their BG regularly during this period regardless of their insulin regime. Their insulin doses will need to increase to compensate for the rising blood glucose levels. If the decision is made to commence steroids, contact **must** be made with the diabetes team. The diabetes clinical guideline: 'Investigation and management of steroid diabetes' can also be used for further information.

Considerations for young people receiving gastrostomy feeds:

Young people who receive overnight gastrostomy feeds should have their BG checked pre-feed, during the feed and 1-2 hours post feed. If the BG levels are elevated during or post feed, Humulin S (soluble insulin lasting 8-12 hours) should be considered (see above).

Follow up and appointments

After initially commencing once daily insulin therapy, BG levels should be reviewed in CF clinic or phone call by CF nurse within 1-2 weeks. If BG levels and/or HbA1c are not within target range, adjust the insulin dose(s) as appropriate or liaise with the diabetes team. The diabetes team will then liaise directly with the young person/family.

Patients who are on a basal bolus regime with be contacted by the diabetes team directly within 1-2 weeks.

Adjust Doses as appropriate:
Lantus/Levemir by 1-2 units
Novorapid by 0.5-1 unit
Humulin S by 1-2 units

Routine 2 monthly CF clinics should continue and BG profiles/ HbA1c should be reviewed here.

All patients diagnosed with CFRD should be seen in a diabetes annual review clinic annually.

Young people who are on once/twice daily insulin and who can maintain their HbA1c level <53 mmol/mol (7.0%) can predominantly be managed by the CF team.

Young people who are on a basal bolus regime (either fixed dose or carbohydrate counting), need direct contact with the diabetes team including specialist nurse/dietician phone contacts and 3 monthly reviews in the diabetes clinic.

NB: Injection sites must be checked for signs of lipohyperatrophy. If this is suspected, the young person should be discouraged from using this site until it has resolved.

Transition to adult services

Young people who have maintained good control of their CFRD, who are on once or twice daily insulin should aim to be seen in the joint (adult) CF/diabetes clinic and transferred there by the CF team.

Those who have required more intensive input from the diabetes team should be transitioned via the diabetes transition clinic at OCDEM (or local equivalent). They will then move onto the young adult clinic and have appropriate follow up from there.

Contact information

Dr Julie Edge Dr Jeremy Hull	Paediatric diabetes consultant Paediatric respiratory consultant	
Hannah Powell Katherine Corkey	Paediatric diabetes specialist nurse Paediatric CF specialist nurse	X 28736 X 34087 bleep 6879
Gillian Reynolds Judy Wadsworth	Dietician (CF) Dietician (diabetes)	X 31234 X 31227
	Paediatric endo registrar	Bleep 1775

Responsibilities of each team:

Diabetes team	CF team
Prescribe initial doses of insulin (doctor)	Review BGL/ check HbA1c at clinic. Check injection sites (doctor)
To review insulin doses whilst YP is an inpatient (doctor)	Contact young person to r/v BGL in between clinic appointments where necessary if not being followed up by diabetes team (nurse/dietician)
Provide initial education (nurse)	Ensure/encourage adherence to treatment (doctor/nurse/dietician)
To see young people on basal bolus in diabetes clinic To decide if/when a new insulin regime is required (doctor/nurse)	Contact diabetes team where BGL and/or HbA1c are not within target range either at OPA or following phone call (doctor/nurse) Request prescription items to the GP (nurse)
To provide ongoing support with regard to dose changes for young people on OD/BD insulin (doctor/nurse)	
Educate young person/ family in CHO counting where required (dietician) Arrange diabetes annual review clinic appointment (nurse)	