Acute hyperglycaemia in Cystic Fibrosis Related Diabetes: The role of insulin pumps

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Background

- Cystic Fibrosis Related Diabetes (CFRD) is the commonest comorbidity in CF leading to increased mortality rates
- CFRD is associated with reduced lung function and poor nutritional status
- The pathophysiology includes pancreatic fibrosis, reduction in acell and B-cell mass, delayed insulin secretion and variable insulin insensitivity. Insulin production can fluctuate with progression over time to an insulinopenic state1 (figure 1)
- Clinical condition may also influence insulin sensitivity, leading to hyperglycaemia in acute respiratory episodes2
- Insulin is the recommended treatment for CFRD2
- Continuous subcutaneous insulin infusion (CSII) is increasingly used in children and young people with type 1 diabetes but there are few publications on CSII use in those with CFRD
- CSII has the advantage of fewer injections which is an important factor for CF patients who already have a heavy burden with their complex treatment regimes3

Case 1

- 15 year old female diagnosed with CFRD following positive OGTT and elevated HbA1c (7.1%)
- Commenced on Insulin detemir injection once a day (4 units)
- Within 4 months she presented to her local A&E with blood glucose >20.0mmol/L, polyuria, polydipsia and negative ketones indicating an insulinopenic state. Discharged home
- Admitted to GOSH and commenced IV insulin, requiring >2 units/kg/day
- Treatment options offered multiple daily injections (MDI) and CSII, patient opted for CSII
- Commenced insulin pump therapy (1.4 units/kg/day)
- Highly motivated, positive parental support and excellent self management

Case 2

- 15 year old female diagnosed with CFRD following positive OGTT and elevated HbA1c (6.5%)
- Commenced on Insulin detemir injection once a day (2 units)
- Two years later presented to local A&E with blood glucose >20.0mmol/L, polyuria, polydipsia and negative ketones indicating an insulinopenic state. Discharged home
- Admitted to GOSH and commenced IV insulin, requiring >2 units/kg/day
- Commenced on MDI with carbohydrate counting (1.5 units/kg/day)
- Initial improvement in BG control was followed by issues with adherence to insulin injections, low mood and suicide attempt
- Three and a half years after diagnosis a decision was taken to support her request to start insulin pump therapy
- Commenced insulin pump therapy (0.74 units/kg/day)
- Highly motivated in using pump therapy
- Overall engagement with CF MDT improved, however some ongoing non-compliance with CF treatment

Outcomes

- HbA1c improvement was seen at 6 months despite a reduction in insulin requirements
- The greatest improvement was seen in psychological wellbeing and subsequent engagement with all treatments

Case 1 comments

“First of all I had injections, but I had to have so many of them that they just thought I should just go on an insulin pump instead”

“The pump makes me feel better…I think it’s much better to be on the pump because now I don’t have to have any injections”

Case 2 comments

“I take insulin with everything I eat, but I’m on an insulin pump so I don’t have to do any injections”

“I type in my blood sugar level and then the number of carbs I’m eating then I press a button and it delivers the insulin…It makes me feel a lot better”

“The transformation was dramatic…she didn’t have to think about insulin which had removed a great burden” (Clinical psychologist)

Discussion

- These cases show that young people with CFRD can present in an insulinopenic state requiring as much insulin as a young person with Type 1 diabetes
- Children and young people with CF require a high calorie diet, with multiple meals and snacks. Therefore multiple injections are required to achieve euglycaemia in CFRD4
- Limiting carbohydrate intake or omitting insulin can effect nutritional status and growth. An insulin pump gives the ability to give insulin without multiple injections4
- When managing CFRD the goal of treatment is not necessarily reduction in HbA1c but supporting the young person to manage the burden of a second chronic disease
- In a cohort of patients with CFRD, insulin pump therapy has the potential to give these children and young people the best quality of life alongside improved glycaemic control

References


Figure 1 adapted from Moran 20101