

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

Samantha Drew, Rebecca Margetts, Hannah Gordon, Catherine Peters
Great Ormond Street Hospital for Children NHS Foundation Trust, London UK

Background

- Cystic Fibrosis Related Diabetes (CFRD) is the commonest co-morbidity 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 α cell and β 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

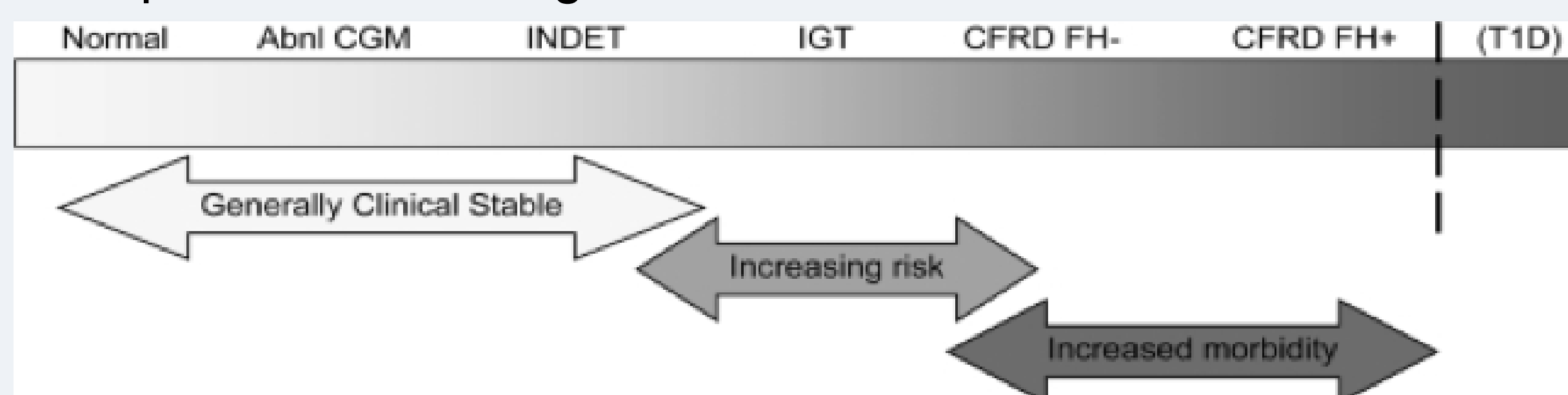


Figure 1 adapted from Moran 2010¹

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 on-going 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		Case 2	
	Pump Start	6 months	Pump Start	6 months
HbA1c	8%	7.70%	14%	8.60%
FEV1 (% pred.)	1.72 (56.3%)	1.88 (61.9%)	2.08 (63.6%)	1.88 (57%)
BMI sds	0.28	0.11	-2.44	-0.93
Insulin requirement (units/kg/d)	1.4	0.5	0.74	0.6

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

1. Moran A, Becker D, Casella SJ et al. Epidemiology, pathophysiology, and prognostic implications of cystic fibrosis-related diabetes: a technical review. *Diabetes Care* 2010; 33: 2677-2683.
2. ISPAD Clinical Practice Consensus Guidelines 2014. Management of cystic fibrosis-related diabetes in children and adolescents. Moran A; Pillay K; Becker DJ; Acerini CL; International Society for Pediatric and Adolescent Diabetes. *Pediatric Diabetes*. 15 Suppl 20:65-76, 2014 Sep.
3. Sulli N et al. Use of continuous subcutaneous insulin infusion in patients with cystic fibrosis related diabetes: Three case reports. *Journal of Cystic Fibrosis* 6 (2007) 237-240
4. Hardin DS et al. Use of the insulin pump in treat cystic fibrosis related diabetes. *Journal of Cystic Fibrosis* 8 (2009) 174-178