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CYSTIC FIBROSIS-RELATED DIABETES: A SMALL COHORT IN MALAYSIA

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INTRODUCTION/BACKGROUND

Cystic fibrosis (CF) has a high prevalence in Europe and America but is very rare in Malaysia. Here we report a small cohort of patients with cystic fibrosis-related diabetes (CFRD) from University Malaya Medical Centre (UMMC), a main tertiary centre in Malaysia. We searched through the registry of patients diagnosed with CFRD in our centre from 2019 to 2024. We compared their demographic, clinical and management data related to CF and CFRD.

CASES

There were four patients in our registry. Three of them were diagnosed with CF requiring pancreatic enzyme supplements and had positive sweat tests, but only one had a confirmed genetic test. The fourth patient was diagnosed with non-CF bronchiectasis and pancreatic insufficiency, given negative sweat tests and genetic tests. All of them developed complications related to CF.

The patients developed CFRD about 9 to 17 years after diagnosis of pancreatic insufficiency, mostly triggered by steroid medication. They had hyperglycemia, high HbA1c, hyperosmolar symptoms and poor body mass index (BMI) at diagnosis. All had normal C-peptide; none had developed ketoacidosis. The third patient was diagnosed with CFRD from an oral glucose tolerance test (OGTT). She had poor weight gain but no hyperosmolar symptoms.

All except the third patient were started on basal-bolus insulin during diagnosis of CFRD, and subsequently transitioned to mixed insulin or only bolus insulin. The third patient opted for a trial of metformin. We faced challenges with various insulin regimes. Most of them had high insulin requirements with a high-calorie diet and steroid treatment. However, they were also prone to hypoglycemia at fasting.

CONCLUSION

CFRD is a unique condition. Even though insulin is the mainstay of treatment, the best regime is not easy to determine, given the fluctuating nature of the disease, multiple comorbidities, and psychosocial issues. The physician should always explore the condition with sympathy, empathy and understanding.