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CHARACTERISTICS OF PATIENTS WITH TYPE 1 DM AND LADA IN A MALAYSIAN PUBLIC HOSPITAL: A CROSS- SECTIONAL STUDY

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INTRODUCTION
Type 1 diabetes mellitus (DM) occurs as the result of pancreatic beta cell destruction. Latent autoimmune diabetes in adults (LADA) is immunologically similar to T1DM but immune destruction progresses at a slower rate. The aim of the study is to identify the clinical characteristics of patients with T1DM and LADA in our clinic.

METHODOLOGY
This is a single centre cross-sectional study involving all 122 patients with T1DM and LADA. Information was obtained from patients' records and interviews during follow up.

RESULTS
There were 49 males (40.2%) and 73 females (59.8%) with a mean age of 35.3 (*SD* 14.9) years old. The mean duration of disease is 12.9 (*SD* 9.7) years. Ninety-five subjects (77.9%) have T1DM and 27 subjects (22.1%) have LADA. The most common complication was retinopathy (14.8%). Almost 2/3 of subjects (61.5%) reported having minor hypoglycemia and 15 (12.3%) had diabetic ketoacidosis in the past year. The most common co-morbid is dyslipidemia (45.9%). Eighteen percent of the subjects have other autoimmune diseases. Majority of the subjects were on at least one analogue insulin (93.4%) and on basal bolus regimen (89.3%). Only 6 subjects (4.9%) were on insulin pump. One hundred fourteen (93.4%) subjects performed self-monitoring of blood glucose (SMBG) and only 26 (21.3%) subjects have used continuous glucose monitoring systems (CGMS) at least once. The mean HbA1c is 8.91% (SD 2.2). The most frequent pancreatic autoantibodies detected were glutamic acid decarboxylase (GAD) (77.9%) and islet cell antibody (ICA) (77.7%).

CONCLUSION
Majority of our subjects with T1DM and LADA are on analogue insulin andon basal-bolus regimen with most of them performing SMBG. Despite this, the rate of hypoglycemia is high and control remains suboptimal. Increasing the use of technologies such as CGMS and insulin pumps which are not fully utilized at present, may improve outcomes.

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CASE REPORT: PRIMARY HYPERPARATHYROIDISM AND JAW TUMOUR SYNDROME WITH CDC73 GENE MUTATION IN A YOUNG PATIENT

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INTRODUCTION
Primary hyperparathyroidism (pHPT) occurs frequently in those over the age of 50 years. This condition is uncommon in young adults and are more likely to have an underlying germline mutation.

CASE
We present a case of a healthy 16-year-old male who was incidentally found to have an elevated calcium of 3.16 mmol/L. Family history revealed that his father, aunty and grandfather also had a history of hypercalcemia. No genetic study was done previously. The patient was diagnosed with iPTH-mediated hyperparathyroidism based on blood investigations. Localization scan revealed an overactive right parathyroid gland secreting excess iPTH. Subsequently, he was scheduled for right parathyroidectomy.

Histopathology report confirmed the diagnosis of a right superior parathyroid adenoma. His iPTH level declined from 968.5 pg/ml to 7.9 pg/ml after the surgery while calcium and ALP levels also normalized. He subsequently required calcium and activated Vitamin D supplementation.

The patient and his family were referred for further genetic assessment, revealing CDC73-related disorders, with a pathogenic mutation on CDC73 gene. The patient's father was found to develop a jaw tumour with histologic confirmation of invasive ossifying fibroma. Hence, tumour debulking was planned.

CDC73-related disorder is an autosomal dominant disorder resulting from the inactivation of the CDC73 tumor suppressor gene. The spectrum includes: Hyperparathyroidism jaw tumor (HPT-JT) syndrome, parathyroid carcinoma and familial isolated hyperparathyroidism (FIHP). Penetration of pHPT is as high as 80% to 95%, while parathyroid carcinoma may be found in more than 20% of patients. Lifelong surveillance is indicated for positive gene carriers to look for recurrent hyperparathyroidism, parathyroid carcinoma, renal and uterine tumour in females.