An MRI abdomen was normal. Octreotide study was negative for insulinoma. He had a normal response to IM glucagon, inferring normal glycogen stores. He was started on Diazoxide 160 mg thrice a day for recurrent hypoglycemia. An endoscopic ultrasound and DOTATATE scan were negative. He had no hypoglycemia for a few days, attributable to lingering effects of diazoxide. Eventually, his serum glucose was 52 mg/dL. Labs prior to glucose correction included an insulin level elevated at 1,000 (normal <3 mcIU/mL), c-peptide at 0.90 ng/mL, and proinsulin of 5.6 pmol/L. Given exceedingly high insulin levels, we measured an IAA level. This was >50 u/mL (normal <0.4 u/mL). With negative imaging and high IAAs, a diagnosis of IAS was made. **Discussion:** IAS or Hirata disease is a rare condition with hyperinsulinemic hypoglycemia and high titers of antibodies to endogenous insulin. The binding kinetics of endogenous insulin to these antibodies causes physiologically inappropriate levels of bioavailable insulin, causing either hyper- or hypoglycemia. IAA should be measured in patients with high insulin levels that are inconsistent with C peptide levels. We believe this to be the first African American patient to have been diagnosed with Hirata disease. Making a correct diagnosis may spare a hypoglycemic patient from unnecessary pancreatic surgical intervention.

## Diabetes Mellitus and Glucose Metabolism

### DIABETES CASE REPORTS

***A Novel Homozygous WRN Mutation Identified in a Middle Aged Man With Diabetes Mellitus Complicated By Multiple Features of Accelerated Aging***

*Batoul Jaafar, MD[1], Mona Nasrallah, MD[1], Junko Oshima, MD, PhD[2], Davor Lessel, MD, PhD[3].*

[1]American University Of Beirut, Department of Internat Medicine, Division of Endocrinology, Beirut, Lebanon, [2]University of Washington, Department of Pathology, Seattle, WA, USA, [3]University Medical Center Hamburg - Eppendorf, Department of Human Genetics, Hamburg, Germany.

**Background:** Werner syndrome (WS; MIM# 277700) is a rare genetic disorder characterized by accelerated aging and predisposition to cancers. The causal gene, *WRN*, encodes a multifunctional nuclear protein that belongs to the RecQ family of DNA helicases. The earliest sign of WS is short stature due to the lack of pubertal growth spurt. In addition to the aged-appearance, WS patients typically develop functional declines of multiple organs including the endocrine system in their middle ages. Frequent Endocrine disorders include Type 2 Diabetes (T2D), dyslipidemia (DL), hypothyroidism, hypogonadism and osteoporosis. Most common causes of deaths are myocardial infarction and cancers in their 50s. **Clinical Case:** A 54-year old man was admitted for right wrist septic arthritis. His medical history was pertinent for premature cataract diagnosed in his 2$^{nd}$ decade. He had uncontrolled T2D (peak HbA1C level 10%) since the age of 32 years, treated with oral hypoglycemics including glimepiride, sitagliptin and metformin then shifted to insulin over the past 2 years. He also had DL since the age of 41 years, characterized by hypertriglyceridemia (766 mg/dL) with very low HDL (7 mg/dL). At age 50, he was diagnosed with hypothyroidism and negative anti-TPO antibodies. Moreover, he had refractory bilateral leg ulcers that manifested at the age of 35 years requiring multiple debridments, premature coronary artery disease at the age of 38 years and ischemic cardiomyopathy. On examination, his height was 154 cm and weight 42 kgs. He had prominent eyes, a pinched nose, grey and sparse scalp hair with absence of eyebrows and eyelashes. Hoarse voice was easily noticeable. He had very thin limbs, flat feet, central fat distribution and muscle wasting. Skin was shiny, tight, and atrophic with overlying hypermelanosis. Old bilateral hyperkeratotic deep feet ulcers were noticed with variable necrosis. Blood chemistry testing showed: HbA1C 8.2%, disturbed lipid profile, TG 341 mg/dL, HDL 17 mg/dL, LDL 155 mg/dL, and TSH 21 μU/mL (0.2 - 4.2 μU/mL). MRI of the foot revealed Achilles' tendon calcification. His parents were 2$^{nd}$ degree cousins and had normal features. Three of his siblings had similar features and premature death. His diabetes was atypical due to his low BMI, high insulin requirements and the relatively early onset. WS was suspected based on the overall features of accelerated aging with characteristic deep leg ulcers and the family history. Genetic sequence analysis revealed a novel pathogenic homozygous nonsense variant mutation c.1111G>T, p.Glu371* of the *WRN* gene.

**Conclusion:** Werner syndrome is a rare genetic disease, but it carries high morbidity and mortality burden, in addition to impaired quality of life. It should be highly suspected in patients with atypical T2D, DL, premature coronary disease, refractory leg ulcers with features of accelerated aging and short stature.

## Diabetes Mellitus and Glucose Metabolism

### DIABETES CASE REPORTS

***A Rare Case of Metformin Associated Lactic Acidosis in a Type 2 Diabetic Patient With No Known Contraindications for Metformin Prescription - a Diagnostic Conundrum!***

*Rima Gandhi, MD[1], Randa Abdelmasih, MD[2], Alauddin El-Hag, MD[1], Elis Cruz Salcedo, MD[1].*

[1]University of Central Florida, Ocala, FL, USA, [2]UNIVERSITY OF CENTRAL FLORIDA, Ocala, FL, USA.

**Introduction:** Metformin is a biguanide drug primarily inhibits hepatic gluconeogenesis and improves insulin sensitivity. Lactic acidosis is a rare complication of metformin. The incidence of Metformin-associated lactic acidosis (MALA) is 6.3 per 100,000 patient-years. Metformin raises lactate levels by inhibiting the conversion of lactate and pyruvate into glucose, shunting towards anaerobic glycolysis. Although, MALA is a reported side effect, metformin is still identified as the drug of choice for Type 2 DM. Here we present a case of MALA in a Type 2 Diabetic patient to shed light on this controversial dilemma. **Case Presentation:** A 56-year-old African-American male with Type 2 DM and diabetic retinopathy presented after a fall and generalized weakness. Upon arrival, his blood sugar was 22 mg/dL. Patient was vitally stable with signs of dehydration. Home medications includes Metformin 1000 mg twice daily and Glipizide. Laboratory results showed an anion gap metabolic acidosis of 18 mmol/L, Lactic acid was 6.5 mmol/L