female case of NDI initially presenting and diagnosed in an adult woman.
Clinical Case:
A previously well 47-year-old woman of Italian descent underwent an elective laparoscopic repair of an abdominal hernia. Her medical history included obesity and migraine headaches. She was not taking any medications prior to admission. She had a bowel perforation 6 days after surgery, necessitating an emergency right hemicolectomy and small bowel resection. Upon instituting bowel rest with *nil per os* (NPO), she developed severe hypernatremia (Na+ 163 mmol/L) with polyuria (>6 L/day) and dilute urine (osmolality 174 mmol/kg). Further inquiry revealed that the patient routinely drank at least 10 L/day of fluids throughout her entire adult life. Her family history was remarkable for polydipsia affecting at least additional six people across three generations (including her son, her mother, 3 maternal uncles and 1 nephew). Following administration of desmopressin 1 ug subcutaneously, her urine remained inappropriately dilute (osmolality 160 mmol/kg) with no significant change in urine output (rate 350 mL/h for 3 hours). Her arginine vasopressin level was detectable (3.2 pmol/L, reference range 0.8–3.5 pmol/L), consistent with nephrogenic diabetes insipidus. Subsequent molecular analysis of the AVPR2 gene, located on chromosome Xq28, confirmed a pathogenic mutation (c.253G>A), consistent with a p.Asp85Asn substitution resulting in decreased binding affinity between the V2 receptor and arginine vasopressin. Thus, X-linked NDI was diagnosed according to the patient's presentation, compatible family history, and genetic analysis. When she was able to eat and drink *ad lib* again, a low-salt, low-protein diet along with a trial of a thiazide diuretic were recommended. The patient remained well with 3 years of follow-up.
Conclusion:
The diagnosis of congenital NDI may be delayed until adulthood because of a relatively mild phenotype and compensatory drinking behavior, so that the disorder will not be clinically apparent until a person is deprived of free water. Men and women alike can be affected by this X-linked dominant condition which should be considered in any polyuric, hypernatremic hospitalized patient.

Reproductive Endocrinology

CLINICAL STUDIES IN FEMALE REPRODUCTION I

Increased Occurrence of Anemia, Gastrointestinal and Liver Diseases in Women with Turner Syndrome - a Nationwide Registry Study

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SAT-025

Background: Gastrointestinal disorders, such as celiac disease, inflammatory bowel diseases and liver disease have previously been described with increased occurrence in women with Turner syndrome. However, evidence towards increased occurrence of bleeding disorders and anemia are sparse. Likewise, the impact of hormone replacement therapy on gastrointestinal disorders remains unknown. **Aim**: To investigate the risk of bleeding disorders, anemia, gastrointestinal and hepatological disease in women with TS compared with the female background population and to assess the effect of HRT on these conditions. **Design:** National cohort study **Method**: 1,156 females with TS diagnosed during 1960–2014 were identified using the Danish Cytogenetic Central Registry and linked with personal-level data from the National Patient Registry and the Medication Statistics Registry. Statistics Denmark randomly identified 115,577 age-matched female controls. Negative binomial regression was used to analyze hospital discharge diagnoses. Medical prescriptions, mortality and the effect of hormone replacement therapy were estimated using stratified Cox regression. **Results**: The risk of anemia, coagulation disorders and gastrointestinal hemorrhage were all increased three-fold in women with TS compared with controls. Gastrointestinal disorders were twice as frequent in TS individuals, with a three-fold increased risk of inflammatory bowel disease and a twelve-fold increased risk of liver disease and elevated liver enzymes. Both gastrointestinal and hepatological mortality were increased three-fold in TS women. **Conclusion:** Anemia, gastrointestinal hemorrhage, inflammatory bowel disease is more frequent in women with Turner syndrome compared with controls. The risk of liver disease may be higher than previously reported.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS I

A MEN-2a Syndrome Index Case Presenting with Adrenergic Crisis and Cardiogenic Shock Due to Bilateral Pheochromocytoma

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SUN-913

Background: MEN-2A syndrome is commonly asymptomatic at diagnosis. Withal, pheochromocytoma presenting as cardiogenic shock is a recognized but exceptional occurrence. **Case:** A healthy 26-year-old female presented to the emergency department with precordial discomfort, headache and shortness of breath, starting that morning. She had a gum corrective surgery in the day before, was medicated with ibuprofen, pantoprazole and amoxicillin/clavulanic acid, and had previous history of migraine and smoking. Her blood pressure was high, and she had pulmonary edema and respiratory failure. ECG: sinus tachycardia, left axis deviation, and negative T wave in aVL. Analytically: leukocytosis, elevated myocardial necrosis markers (troponin I 1.29 ng/mL, normal < 0 ng/mL), and hyperlactacidemia. Transthoracic echocardiogram: severe left ventricular dysfunction, akinesia of the basal