

each. Most TFTA (79%) occurred under pembrolizumab and nivolumab. Grade 2 CTCAE was the most frequently reported. None of the events led to ICI suspension.

Patients with TFTA underwent a significant higher number of ICI cycles than control group [median 11 cycles (IQR 20) vs 7 (IQR 11), $p=0.017$] and had a higher period under ICI (median of 7.6 months (IQR 13.8) vs 4.2 (IQR 7.7), $p=0.026$). Comparison between TFTA patients and control group did not reveal statistical differences in patients' age and sex, primary neoplasm, tumor staging and ICI.

Overall survival was significantly higher in patients that developed TFTA during treatment with ICI, comparing to the control group (mean OS 3.62 years vs 1.92 years, $p=0.033$). The risk of mortality was higher for the control group, approximately 3 times, considering the adjustment for the covariates (HR 2.94, 95%CI=1.18 to 7.34, $p=0.021$).

Overall survival was not affected by the covariates.

Our study shows that patients under ICI that develop primary or central thyroid dysfunction had an improved survival. In these patients, the occurrence of TFTA could be a marker of a better response to ICI.

Adrenal

ADRENAL CASE REPORTS II

A False Positive Result in Newborn Screening for Congenital Adrenal Hyperplasia (CAH) in a Girl with Beckwith Wiedemann Syndrome

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SUN-172

Background: Congenital Adrenal Hyperplasia (CAH) comprises a spectrum of autosomal recessive diseases, resulting in enzymatic defects in the cortisol secretion. CAH newborn screening can avoid neonatal mortality in children with the salt-wasting form and prevent incorrect gender assignments in females. The occurrence of false-positive results creates diagnostic difficulties presenting therapeutic implications. Beckwith Wiedemann Syndrome (BWS) is a congenital disease characterized by somatic overgrowth, increased risk of neonatal hypoglycemia, and development of embryonic tumors. BWS is due to (epi)genetic changes involving growth-regulating genes with good genotype-phenotype correlation. The adrenal gland is frequently involved and may present diffuse cytomegaly of the adrenal cortex¹. We reported a BWS newborn girl with a false-positive diagnosis of CAH in the screening. **Case report:** The patient was born at 39 weeks from an uneventful cesarean section, 5.6kg (>p97) and 52cm (>p97), referred to the Endocrinology service due to abnormal neonatal tests (neonatal 17-OHP: 96ng/mL) collected at 6 days old. At 14 days old, she was 6.3 kg (Z:+5.59), and 58cm (Z:+2.47), BMI: 18.7 kg/m² (Z:+4.45),

and with typical female external genitalia, ruling out the diagnosis of classic CAH. She presented some syndromic characteristics as macroglossia, ogival palate, orbital hypertelorism, hepatomegaly, and umbilical hernia. At 1 month and 14 days old, serum 17OHP was 7.4ng/mL, androstenedione: 6.1 ng/mL, total testosterone: 279ng/dL, 11-deoxycortisol: 2.11ng/mL, cortisol: 5.0ug/dL, and ACTH: 54pg/mL. At five months old she evolved with normalization of serum 17OHP, androstenedione and testosterone levels (1.36ng/mL, <0.50ng/mL, and 37ng/dL, respectively), but still with high DHEAS levels: 2913ng/mL. At 11 months old, DHEAS also normalized, confirming that it was transient hyperactivity of the *zona reticulata*. A molecular test was performed in a blood sample by MLPA, showing a gain of methylation in the imprinting control region 1 (ICR1) of chromosome 11p15, which controls two imprinted genes, H19 and IGF-2, confirming the clinical diagnosis of BWS. The hypermethylation of ICR1 is largely related to the Wilms tumor. The patient was diagnosed with bilateral Wilms tumor at 11 months old and undergone chemotherapy without adequate response requiring left nephrectomy at 1 year and 5 days old. **Conclusion:** We presented the first description of false-positive diagnosis of CAH in the newborn screening of a girl with Beckwith Wiedemann syndrome, probably due to a transient overactivation of the *zona reticulata*. **References:** 1.Brioude F, Kalish JM, Mussa A, Foster AC, Blik J, Ferrero GB, et al. Expert consensus document: clinical and molecular diagnosis, screening and management of Beckwith-Wiedemann syndrome: an international consensus statement. *Nat Rev Endocrinol.* 2018;14(4):229-49.

Healthcare Delivery and Education

EXPANDING CLINICAL CONSIDERATIONS FOR PATIENT TESTING AND CARE

Reducing Unnecessary Inpatient Adrenocorticotropin Stimulation Tests

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Background:

Outpatient adrenocorticotropin (ACTH) stimulation tests can be challenging to interpret due to heterogeneously reported cut-offs. Inpatient stimulation tests present additional challenges due to the presence of acute illness and unreliable coordination of dynamic function testing on a busy inpatient service. This study aims to characterize the use of ACTH stimulation tests in hospitalized patients to determine necessity of testing.

Methods:

We conducted an inpatient audit of ACTH simulation tests done to rule out adrenal insufficiency between April 2018 to March 2019 at our institution. Normal post-ACTH response was defined as peak cortisol ≥ 500 nmol/L. Testing was considered inappropriate in patients with normal