of steroid synthesis elsewhere. Extra adrenal sources for glucocorticoid production are known such as skin, gonads and thymus. However, the levels are insufficient to mount a significant stress response. There is evidence of adrenal regeneration in adrenalectomized animals. The regeneration is primarily of the adrenal cortex and does not involve the medulla. There has been one case report in literature of a 11 year old German boy who had adrenal regeneration detected on adrenal scintigraphy (Bilateral normal adrenal glands with normal activity) 13 years after adrenalectomy for Cushing's disease.

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Adrenal

ADRENAL CASE REPORTS

Cardiac Arrest in a Child with Non-classic Lipoid Congenital Adrenal Hyperplasia Associated with a New STAR Gene Mutation

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Introduction: Steroidogenic acute regulatory (STAR) protein regulates steroid hormone synthesis by transporting cholesterol into mitochondria. *STAR* gene mutations lead to lipoid congenital adrenal hyperplasia (LCAH), the rare but most severe form of congenital adrenal hyperplasia in children. We present an unusual case with an episode of cardiac arrest in a young girl during an acute febrile illness and later she was diagnosed with adrenal insufficiency secondary to a non-classic LCAH. **Case:** 2-year 11-month-old previously healthy white female was brought to an urgent care clinic due to severe lethargy and a seizure-like activity during a fever illness. She was found to have an undetectable blood glucose level and went into cardiac arrest shortly after arrival. CPR was performed for approximately 11 minutes. She then developed sever respiratory distress and was intubated. She was transferred to the PICU with IV sodium bicarbonate given en route. On admission, her body weight was 13.26 kg (36.80^{th} percentile), height 90 cm (17.56^{th} percentile), and BMI 16.17 (62.88^{th} percentile). Her physical exam revealed normal external female genitalia and normal skin pigmentation. Lab evaluation revealed normal sodium and potassium but elevated anion gap, hyperuricemia, elevated creatinine kinase, abnormal liver function tests and abnormal coagulation profile. Brain MRI revealed findings consistent with hypoxic-ischemic encephalopathy. Renal function improved within 24 hours and hepatic function returned to normal after 20 days. Due to her severe hypoglycemic event, a high-dose ACTH stimulation test was performed. The results were consistent with adrenal insufficiency: baseline cortisol level, 7.3 µg/dL; 30 minutes cortisol, 7.8 µg/dL; 60 minutes cortisol, 9 µg/dL (normal response, ≥18 mcg/mL at 30 or 60 minutes). The baseline ACTH level was significantly elevated, 1688 pg/mL (0–46) as well as the renin activity, 24.3 ng/hour (1.7–11.2). Genetic testing revealed a 46 XX karyotype. *STAR* gene analysis identified compound heterozygosity; a novel deletion (c.811delC, p.Leu271Cysfs*50) and a previously reported missense mutation (c.661G>A, p.Gly221Ser). The girl is now 11 years old and exhibits normal growth, normal cognitive development, and she has developed early signs of puberty (Tanner stage 2 for breast). She takes daily hydrocortisone, fludrocortisone and stress dose hydrocortisone as needed. **Conclusion:** In non-classic LACH, the onset is generally late or not acute. Initial clinical features are variable and nonspecific. For this reason, non-classic LCAH may be overlooked. Adrenal crisis is a life-threatening complication, and it is important that clinicians are aware of the clinical features of non-classic LCAH and consider it in the differential diagnoses. Genetic testing for *STAR* should be considered in individuals with non-autoimmune primary adrenocortical insufficiency.

Adrenal

ADRENAL CASE REPORTS

Careful Evaluation of Cosyntropin Dose in the Diagnosis of Adrenal Insufficiency

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Introduction: The use of the 250µg cosyntropin dose or otherwise called high-dose ACTH test is the gold standard test for diagnosis of primary adrenal insufficiency. The 1µg dose test or the low-dose test is mostly reserved for diagnosis of secondary adrenal insufficiency. Careful consideration of the results produced during the diagnostic process is imperative to avoid mislabeling of patients with a disease that requires lifelong treatment.

Case Report: This is the case of a 45-year-old female with a history of asthma and psoriasis who presented with emesis. Home medications included monthly TNF-alpha inhibitor injections for psoriasis, triamcinolone acetonide topical spray and budesonide-formoterol inhaler. On admission, she also had nausea, chills and diaphoresis, as well as palpitations, lightheadedness, and shortness of breath. When she arrived at the ER, vitals were remarkable for low blood pressure. Labs were unremarkable except for CMP concerning for anion gap metabolic acidosis, hyponatremia, and hypokalemia. A random serum cortisol was 6.4 mcg/dL, which was relatively low. ACTH was within normal range. Due to concern for adrenal insufficiency, a 1µg cosyntropin test was performed which showed a peak cortisol concentration of less than 18 mcg/dL. As the response was assessed as suboptimal, endocrinology was consulted to offer a treatment plan for steroids. However, the test was repeated using the gold standard 250µg cosyntropin dose and the patient then showed an adequate response and she was not started on steroids.

Conclusions: This is a case that demonstrates how the 250 µg ACTH or high-dose stimulation test should be used