

NEW TEST

NOTIFICATION DATE: December 20, 2016 **EFFECTIVE DATE:** January 4, 2017

11-Deoxycorticosterone, Serum

Test ID: DOCS

USEFUL FOR:

- Diagnosis of suspected 11-hydroxylase deficiency, including the differential diagnosis of 11 beta-hydroxylase 1 (CYP11B1) versus 11 beta-hydroxylase 2 (CYP11B2) deficiency, and in the diagnosis of glucocorticoid-responsive hyperaldosteronism
- Evaluating congenital adrenal hyperplasia newborn screen-positive children, when elevations of 17-hydroxyprogesterone are only moderate, suggesting possible 11-hydroxylase deficiency

METHOD: Liquid Chromatography-Tandem Mass Spectrometry (LC-MS/MS)

REFERENCE VALUES:

< or =18 years: <30 ng/dL >18 years: <10 ng/dL

SPECIMEN REQUIREMENTS:

Preferred: Red top **Acceptable:** Serum gel **Specimen Volume:** 0.5 mL

Collection Instructions: Morning (8 a.m.) specimen is preferred.

Minimum Volume: 0.4 mL

SPECIMEN STABILITY INFORMATION:

Specimen Type	Temperature	Time
Serum	Refrigerated (preferred)	21 days
	Ambient	7 days
	Frozen	21 days

CAUTIONS:

- At birth, the hypothalamic-pituitary-adrenal axis and the hypothalamic-pituitary-gonadal
 axis are activated and all adrenal steroids are high, including mineral corticoids and sex
 steroids and their precursors. In preterm infants, elevations can be even more pronounced
 due to illness and stress. In doubtful cases, when the initial test was performed on a justborn baby, repeat testing a few days or weeks later is advised.
- Adrenocorticotrophic hormone (ACTH)1-24 testing has a low, but definite risk of drug and allergic reactions and should, therefore, only be performed under the supervision of a physician in an environment that guarantees the patient's safety, typically an endocrine, or other centralized, testing center.

• Interpretation of ACTH1-24 testing in the context of diagnosis of congenital adrenal hyperplasia (CAH) requires considerable experience, in particular for the less common variants of CAH, such as 11-hydroxylase deficiency or 3-beta-hydroxysteroid dehydrogenase (3beta-HSD deficiency), for which very few, if any, reliable normative data exist. For the even rarer enzyme defects, such as deficiencies of StAR (steroidogenic acute regulatory protein), 20,22 desmolase, 17a-hydroxylase/17-lyase, and 17-beta-hydroxysteroid dehydrogenase (17beta-HSD), there are only case reports. Expert opinion from a pediatric endocrinologist with experience in CAH should, therefore, be sought.

CPT CODE: 82633

DAY(S) SET UP: Tuesday 10 am **ANALYTIC TIME:** 3 days

NOTE: The following referral test code will become obsolete.

Test Name	Test ID	Referral Lab Code	Referral Lab
Deoxycorticosterone (DOC), Serum	FDOC	500124	Esoterix

NOTE: The following MML test code will become obsolete.

11-Deoxycorticosterone, Serum	DCRN	Mayo Medical Laboratories
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