

Chapter 3

ACTH Stimulation Test for Late Onset (Nonclassic) 21-Hydroxylase Deficiency

Indication: To evaluate for androgen excess in diagnosing non-classic CYP21A2 deficiency. If Basal 17-hydroxyprogesterone (17 OHP) <2 ng/ml, diagnosis is unlikely and ACTH stimulation may not be necessary [1, 2].

Preparation: Women are best tested in the early follicular phase of the menstrual cycle. It is recommended to hold glucocorticoids for 24 h prior to testing to avoid any effect on 17 OH progesterone level.

Materials Needed: Two (2) gold top tubes labeled as baseline and 60 minutes

Cortisol:
Gold top tube

Cortrosyn 250 mcg

Syringes/needles

17 OH Progesterone:
Gold top tube

Assay for 17 OHP: Radioimmunoassay (RIA).

Interpretation: With late onset 21-hydroxylase deficiency, the absolute value of 17-hydroxyprogesterone at 60 min sample is > 10 ng/dl [2, 3].

Caveats:

- Baseline androgen levels return to baseline after 8 weeks of discontinuation of oral contraceptive pills (OCP) [4].
- There is not enough data in regards to the effect of OCP on 17 OHP levels after ACTH stimulation.

Procedure: Completed as outpatient.

1. Obtain baseline blood sample for (17 OHP) and cortisol.
2. Give cortrosyn 250 mcg IM.
3. At 60 min, obtain sample for (17 OHP) and cortisol.

Physician name and signature: _____

RN performing the procedure: _____

Additional orders by physician: _____

ACTH stimulation	Cortisol	17-OHP
Basal		
30 min		
60 min		

References

1. Azziz R, Zacur HA. 21-hydroxylase deficiency in female hyperandrogenism: screening and diagnosis. *J Clin Endocrinol Metab.* 1989;69(3):577–84.
2. Azziz R, Dewailly D, Owerbach D. Clinical review 56: nonclassic adrenal hyperplasia: current concepts. *J Clin Endocrinol Metab.* 1994;78(4):810–5.
3. New MI, Lorenzen F, Lerner AJ, et al. Genotyping steroid 21-hydroxylase deficiency: hormonal reference data. *J Clin Endocrinol Metab.* 1983;57(2):320–6.
4. Sanchez LA, Perez M, Centeno I, David M, Kahi D, Gutierrez E. Determining the time androgens and sex hormone-binding globulin take to return to baseline after discontinuation of oral contraceptives in women with polycystic ovary syndrome: a prospective study. *Fertil Steril.* 2007;87(3):712–4.