Test Definition: OHPG

17-Hydroxyprogesterone, Serum

Overview

Useful For

Screening for congenital adrenal hyperplasia due to 21-hydroxylase deficiency in patients with compatible clinical findings

As part of an evaluation for nonclassic 21-hydroxylase deficiency in patients with hyperandrogenic symptoms such as hirsutism, acne, menstrual irregularity, infertility, or premature pubarche

As one component of a broader steroid evaluation when another disorder of adrenal steroidogenesis is suspected. 17-Hydroxyprogesterone alone is not sufficient to distinguish all forms of CAH

Adjunctive evaluation of patients with an adrenal mass and clinical or imaging features suspicious for adrenocortical carcinoma (ACC), as part of a serum steroid panel

Biochemical follow-up of known ACC when serum OHPG was elevated at diagnosis, in conjunction with other hormonal markers and imaging

Testing Algorithm

For information see Steroid Pathways

Special Instructions

- Steroid Pathways

Method Name

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

NY State Available

Yes

Specimen

Specimen Type

Serum Red

Ordering Guidance

The preferred screening test for congenital adrenal hyperplasia caused by 21-hydroxylase deficiency is CAH21 / Congenital Adrenal Hyperplasia (CAH) Profile for 21-Hydroxylase Deficiency, Serum, which allows the simultaneous determination of 17-hydroxyprogesterone, androstenedione, and cortisol.

Necessary Information

Patient's age and sex are required.

Specimen Required

Collection Container/Tube: Red top (serum gel/SST are **not acceptable**)
Submission Container/Tube: Plastic vial
Specimen Volume: 0.6 mL Serum
Collection Instructions: Centrifuge and aliquot serum into plastic vial.

Forms

If not ordering electronically, complete, print, and send General Test Request (T239) with the specimen.

Specimen Minimum Volume

Serum: 0.25 mL

Reject Due To

Gross hemolysis	OK
Gross lipemia	Reject
Gross icterus	OK

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum Red	Refrigerated (preferred)	28 days	
	Ambient	7 days	
	Frozen	28 days	

Clinical & Interpretive

Clinical Information

17-Hydroxyprogesterone (17-OHP; OHPG) is an intermediate in adrenal and gonadal steroidogenesis. In the adrenal cortex, OHPG is converted by 21-hydroxylase (CYP21A2) to 11-deoxycortisol and subsequently by 11-beta-hydroxylase (CYP11B1) to cortisol. Deficiency of 21-hydroxylase impairs cortisol synthesis and, in classic salt-wasting disease, aldosterone synthesis. Loss of cortisol-mediated feedback increases adrenocorticotropic hormone (ACTH), causing adrenal hyperplasia and accumulation of steroid precursors, particularly OHPG, with increased androgen production.(1)

Congenital adrenal hyperplasia (CAH) comprises inherited disorders of adrenal steroidogenesis. More than 90% of cases are caused by 21-hydroxylase deficiency due to pathogenic variants in *CYP21A2*. Classic disease may present in infancy with adrenal insufficiency and salt-wasting crisis and, in 46,XX infants, prenatal androgen excess with virilization of the external genitalia. Nonclassic 21-hydroxylase deficiency is milder and may present later with premature pubarche, hirsutism, acne, menstrual irregularity, or infertility; some affected individuals are asymptomatic.(1,2)

17-Hydroxyprogesterone is produced by the adrenal glands and gonads and by the fetoplacental unit during pregnancy.

Serum measurement reflects total circulating OHPG. Concentrations vary with age, sex, time of day, menstrual-cycle phase, physiologic stress, and analytical method; these factors should be considered when interpreting results.(1,2)

The steroid pattern differs among CAH subtypes. In 11-beta-hydroxylase deficiency (CYP11B1), 11-deoxycortisol and 11-deoxycorticosterone are characteristically increased, and adrenal androgens and OHPG may also be elevated. In combined 17-alpha-hydroxylase/17,20-lyase deficiency (CYP17A1), OHPG and androgens are low or low-normal, whereas progesterone, 11-deoxycorticosterone, and corticosterone are increased. In 3-beta-hydroxysteroid dehydrogenase type 2 deficiency (HSD3B2), delta-5 steroids, particularly 17-hydroxypregnenolone, are disproportionately increased. A broader steroid profile is therefore preferred when a rare defect of steroidogenesis is suspected.(3-5) Measurements of 21-deoxycortisol and other steroid precursors may provide additional information beyond OHPG alone.(6)

Adrenocortical carcinoma (ACC) can cause elevations of serum OHPG and other steroid precursors because of disorganized steroidogenesis. European Society of Endocrinology/European Network for the Study of Adrenal Tumors guidelines include OHPG in the hormonal work-up of suspected ACC and suggest measurement of sex steroids and steroid precursors, preferably using liquid chromatography tandem mass spectrometry multisteroid profiling, when clinical or imaging findings raise concern for ACC. This is not recommended as routine testing for every adrenal incidentaloma.(7,8) ACC has heterogeneous secretory phenotypes, including cortisol-secreting, androgen-secreting, mixed cortisol/androgen-secreting, and nonsecreting tumors. Serum OHPG should be considered within a broader hormonal assessment.(7,9,10)

For more information see Steroid Pathways.

Reference Values

Children:
Preterm infants
Concentration may exceed 630 ng/dL, however, it is uncommon to see levels reach 1000 ng/dL.
Term infants
0-28 days: <630 ng/dL
Levels fall from newborn (<630 ng/dL) to prepubertal gradually within 6 months.
Prepubertal males: <110 ng/dL
Prepubertal females: <100 ng/dL

Adults:
Males: <220 ng/dL
Females
Follicular: <80 ng/dL
Luteal: <285 ng/dL
Postmenopausal: <51 ng/dL

Note: For pregnancy reference ranges see: Soldin OP, Guo T, Weiderpass E, et al. Steroid hormone levels in pregnancy and 1 year postpartum using isotope dilution tandem mass spectrometry. Fertil Steril. 2005;84(3):701-710

Interpretation

Marked elevation of 17-Hydroxyprogesterone (OHPG) is characteristic of 21-hydroxylase deficiency. In classic disease, concentrations are typically substantially above the age-appropriate reference interval. Androstenedione and other

adrenal androgens are often increased. Diagnosis should integrate the clinical presentation with the steroid pattern rather than rely on a single analyte.(1,6)

In 11-beta-hydroxylase deficiency, 11-deoxycortisol is the most informative biochemical marker; 11-deoxycorticosterone and adrenal androgens are also increased, and OHPG may be elevated. Hypertension and hypokalemia can occur because of excess mineralocorticoid precursors.(5)

In combined 17-alpha-hydroxylase/17,20-lyase deficiency, reduced cortisol and sex-steroid synthesis is accompanied by increased production of 17-deoxysteroids, especially 11-deoxycorticosterone and corticosterone. Patients commonly have hypertension, hypokalemia, and impaired pubertal/sexual development. An elevated OHPG is not expected, so OHPG alone is unsuitable for screening.(3)

In a patient with an adrenal mass, increased OHPG should be interpreted alongside DHEA-S, androstenedione, 11-deoxycortisol, and other clinically appropriate sex-steroid measurements, as well as adrenal imaging.(9,10)

For more information see Steroid Pathways.

Cautions

17-Hydroxyprogesterone (OHPG) concentrations are physiologically higher in newborns, particularly in premature, low-birthweight, ill, or stressed infants, contributing to false-positive newborn-screening results. Interpret results using gestational-age-, postnatal-age-, and method-appropriate criteria. Second-tier liquid chromatography tandem mass spectrometry (LC-MS/MS) steroid testing can improve screening specificity, and 21-deoxycortisol is a useful additional marker in this setting. Dried-blood-spot screening cutoffs should not be applied to serum results.(1,11,12)

17-Hydroxyprogesterone shows diurnal variation and is influenced by adrenocorticotropic hormone (ACTH) secretion. In menstruating patients, luteal-phase ovarian production can increase OHPG.

Results obtained by different analytical methods are not interchangeable. Immunoassays may show positive bias from cross-reacting steroids, whereas LC-MS/MS methods are more analytically specific and may yield lower values. Interpret results using method-specific reference intervals and diagnostic thresholds.(13-15)

Glucocorticoid exposure should be considered because it can suppress adrenal steroid concentrations and complicate diagnostic testing.(3,4,6)

An elevated OHPG is not specific for adrenocortical carcinoma (ACC), and a normal result does not exclude ACC. Do not use OHPG alone to distinguish benign from malignant adrenal masses.(9,10)

Clinical Reference

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3. Auchus RJ. Steroid 17-hydroxylase and 17,20-lyase deficiencies, genetic and pharmacologic. J Steroid Biochem Mol Biol. 2017;165(Pt A):71-78
4. Guran T, Kara C, Yildiz M, et al. Revisiting Classical 3beta-hydroxysteroid Dehydrogenase 2 Deficiency: Lessons from 31 Pediatric Cases. J Clin Endocrinol Metab. 2020;105(3):e1718-e1728

5. Khattab A, Haider S, Kumar A, et al. Clinical, genetic, and structural basis of congenital adrenal hyperplasia due to 11-beta-hydroxylase deficiency. Proc Natl Acad Sci U S A. 2017;114(10):E1933-E1940
6. Fiet J, Le Bouc Y, Guechot J, et al. A Liquid Chromatography/Tandem Mass Spectometry Profile of 16 Serum Steroids, Including 21-Deoxycortisol and 21-Deoxycorticosterone, for Management of Congenital Adrenal Hyperplasia. J Endocr Soc. 2017;1(3):186-201
7. Fassnacht M, Dekkers OM, Else T, et al. European Society of Endocrinology Clinical Practice Guidelines on the management of adrenocortical carcinoma in adults, in collaboration with the European Network for the Study of Adrenal Tumors. Eur J Endocrinol. 2018;179(4):G1-G46
8. Fassnacht M, Tsagarakis S, Terzolo M, et al. European Society of Endocrinology clinical practice guidelines on the management of adrenal incidentalomas, in collaboration with the European Network for the Study of Adrenal Tumors. Eur J Endocrinol. 2023;189(1):G1-G42
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11. Held PK, Bialk ER, Lasarev MR, Allen DB. 21-Deoxycortisol is a Key Screening Marker for 21-Hydroxylase Deficiency. J Pediatr. 2022;242:213-219 e1
12. Nordenstrom A, Wedell A, Hagenfeldt L, Marcus C, Larsson A. Neonatal screening for congenital adrenal hyperplasia: 17-hydroxyprogesterone levels and CYP21 genotypes in preterm infants. Pediatrics. 2001;108(4): E68
13. Bizzarri C, Chioma L, Bottaro G, et al. Diagnostic cut-offs of 17-hydroxyprogesterone by LC-MS/MS in children with non-classical congenital adrenal hyperplasia. J Endocrinol Invest. 2025;48(7):1623-1633
14. Chesover AD, Millar H, Sepiashvili L, Adeli K, Palmert MR, Hamilton J. Screening for Nonclassic Congenital Adrenal Hyperplasia in the Era of Liquid Chromatography-Tandem Mass Spectrometry. J Endocr Soc. 2020;4(2):bvz030
15. Nakhleh A, Saiegh L, Shehadeh N, et al. Screening for non-classic congenital adrenal hyperplasia in women: New insights using different immunoassays. Front Endocrinol (Lausanne). 2022;13:1048663

Performance

Method Description
Isotopically labeled internal standards are added to the samples followed by a protein precipitation step. The resulting supernatant is injected and enriched by online solid-phase extraction prior to liquid chromatography tandem mass spectrometry analysis.(Unpublished Mayo method)

PDF Report
No

Day(s) Performed
Monday through Friday

Report Available
2 to 5 days

Specimen Retention Time
14 days

Performing Laboratory Location

Mayo Clinic Laboratories - Rochester Superior Drive

Fees & Codes

Fees

- Authorized users can sign in to Test Prices for detailed fee information.
- Clients without access to Test Prices can contact Customer Service 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact Customer Service.

Test Classification

This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. It has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information

83498

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
OHPG	17-Hydroxyprogesterone, S	1668-3

Result ID	Test Result Name	Result LOINC® Value
9231	17-Hydroxyprogesterone, S	1668-3