Adrenal

ADRENAL – CLINICAL RESEARCH STUDIES

What Cut-off Value of 17-Hydroxyprogesterone Should Be an Indication to Perform a 250 µg Cosyntropin Stimulation Test When NCCAH Is Suspected? - a Retrospective Study.

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The Nonclassic Congenital Adrenal Hyperplasia (NCCAH) is a less severe form of CAH in which the activity of the 21-hydroxylase is estimated at about 20% to 50%. Cosyntropin stimulation test is the gold diagnostic standard used to test for this condition. The study was aimed at verifying the currently accepted threshold of 17-hydroxyprogesterone (17OHP) level (≥2.0 ng/mL) at which cosyntropin stimulation test should be performed. Material and methods. The study included 343 patients (328 females and 15 males) referred for a cosyntropin stimulation test due to suspected NCCAH. The median age at the time of evaluation was 27 years. Serum 17-OHP was measured with ELISA assay using Ledect96 Microplate Reader. The NCCAH diagnosis was made if cosyntropin-stimulated 17OHP level exceeded 10.0 ng/mL. The ROC curve was determined, and the cut-off point with the highest sensitivity and specificity was established. The study was approved by the Ethics Board of JUMC. **Results:**. Symptoms, which prompted testing for NCCAH, most often were: hirsutism in 187 patients, irregular menstrual cycles in 178 patients, and acne in 138 patients. A total of 79 patients (77 females and two males) were diagnosed with NCCAH based on cosyntropin stimulation test results. Seventy-one of them had baseline levels of 17OHP≥2.0 ng/mL. The mean age of patients with confirmed NCCAH was 28.86 years. The baseline 17OHP cut-off value that qualified patients best for testing was 2.79 ng/mL in our group, with sensitivity and specificity of 77.2% and 91.3%, respectively. The sensitivity and specificity for a guideline-recommended cut-off point (17OHP ≥2.0 ng/mL) was 86.1% and 76%, respectively. In five of six patients with secondary amenorrhea and a baseline level of 17OHP ≥2.0 ng/mL, NCCAH was confirmed in a cosyntropin stimulation test. **Conclusions:**. Our results suggest considering an upward shift in the 17OHP threshold at which patients suspected for NCCAH should be referred for further evaluation. This may reduce the number of unnecessary cosyntropin stimulation tests, mainly that patients with mild phenotype (or asymptomatic) frequently may not require any treatment. However, attention should be paid to patients with coexisting secondary amenorrhea and 17OHP levels ≥2.0 ng/mL, which may be a clinical predictor of NCCAH.

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ADRENAL CASE REPORTS

"Relieving the Pressure" With Unilateral Adrenalectomy in a Case of Chronic Resistant Hypertension From Primary Aldosteronism

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Introduction: Primary aldosteronism is an underrecognized cause of resistant hypertension that is associated with an increased risk of cardiovascular disease and mortality. While surgical intervention may not lead to complete resolution of hypertension, partial success (defined by reduction in blood pressure and/or medications) can be achieved and future cardiovascular risks can be minimized. We present a case of a patient with primary aldosteronism whose chronic resistant hypertension improved significantly after unilateral adrenalectomy. **Clinical Case:** A 54-year-old female with resistant hypertension for 17 years and hypokalemia was diagnosed with primary aldosteronism. The patient had uncontrolled hypertension despite atenolol 50 mg, nifedipine XL 60 mg, triamterene-hydrochlorothiazide 37.5-25 mg, lisinopril 40 mg daily.
Laboratory evaluation was significant for plasma aldosterone concentration (PAC) 26.8 ng/dL (reference: 0.0–30.0 ng/dL), plasma renin activity (PRA) 0.168 ng/mL/hr (reference: 0.167–5.380 ng/mL/hr), PAC/PRA ratio 159.5, and potassium of 3.2 mmol/L (reference: 3.5–4.5mmol/L). Other workup showed plasma normetanephrines 128 pg/mL (reference: 0–145 pg/mL), metanephrines 25 pg/mL (reference: 0–62 pg/mL), and two normal midnight salivary cortisol tests. Saline infusion testing confirmed primary aldosteronism with a non-suppressed aldosterone level of 15.6 ng/dL (normal <5 ng/dL). Abdominal imaging revealed two low-density right adrenal nodules consistent with adenomas and thickening of the left adrenal gland. The patient underwent adrenal vein sampling (AVS) with cosyntropin stimulation, which showed lateralization to the right adrenal. Despite adding clonidine 0.1 mg three times daily, hydralazine 25 mg three times daily and spironolactone 100 mg daily (which substituted triamterene-hydrochlorothiazide) after AVS, her blood pressure remained uncontrolled with blood pressure ranging from 150–180/90–110 mmHg. A laparoscopic right adrenalectomy was performed. Pathology revealed two adrenal cortical adenomas. At the one-month post-operative visit, her potassium was normal, PAC was 4.3 ng/dL, and blood pressure improved on a reduced regimen of atenolol, lisinopril, and nifedipine.
Conclusion: In patients with chronic uncontrolled hypertension due to primary aldosteronism, surgical intervention may not lead to complete resolution of hypertension but may lead to partial clinical success. Residual hypertension may be due to underlying vascular changes associated with long-term exposure to elevated aldosterone. The post-operative PAC in our case was less than 5 ng/dL suggesting biochemical cure and potentially reduced cardiovascular risk. This case emphasizes that adrenalectomy should be considered in resistant hypertension, even if complete clinical cure may not be attainable to reduce future cardiovascular events.