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Synacthen Stimulation of 17 α OHP

Adrenal glucocorticoid secretion is controlled by adrenocorticotrophic hormone (ACTH) released by the anterior pituitary. This test evaluates the ability of the adrenal cortex to produce cortisol after stimulation by synthetic ACTH (tetracosactide: Synacthen®). In subjects with enzyme deficiency in the steroid synthetic pathway, cortisol may, or may not, be adequately secreted. However, there is excessive secretion of the precursor steroids before the defective enzyme. The commonest form of CAH is due to deficiency of 21-hydroxylase and in these subjects increased secretion of 17 α OH-progesterone (17 OHP) can be detected.

Indications

This is performed for the investigation of congenital adrenal hyperplasia (CAH).

Contraindications

The Synacthen test gives unreliable results within 2 weeks of pituitary surgery.

Side Effects

There are rare reports of hypersensitivity reactions to Synacthen particularly in patients with a history of allergic disorders.

Preparations and precautions

- Prednisolone should be stopped 24 hours before the Short Synacthen test.
- Hydrocortisone should be omitted on the morning of the Short Synacthen test.
- Consider sending a random urine for urine steroid profile (10mL Monovette urine tube)

Requirements

- 6 x brown top serum tubes
- 250 microgram Synacthen (1 ampoule)

Procedure

- This test should be performed preferably in the morning between 0800 and 0900 hours but can be performed later in the day.

| Minutes | Procedure | Sample |
|---------|--|---|
| 0 | Take sample for Cortisol and 17 OHP and then administer 250 μ g Synacthen IV / IM | 2 x brown top serum tube (cortisol and 17OHP) |
| 30 | Take sample for Cortisol and 17 OHP | 2 x brown top serum tube (cortisol and 17OHP) |
| 60 | Take sample for Cortisol and 17 OHP | 2 x brown top serum tube (cortisol and 17OHP) |

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Interpretation of results

Cortisol normal response at 30 mins: Peak > 430nmol/L

17-OHP Unaffected adults and children usually have a basal 17-OHP of <6 nmol/L.

A minority of patients with non-classical CAH have a normal basal 17-OHP, even on early morning samples.

A normal response to Synacthen is a stimulated 17-OHP of <9.8 nmol/L at 60 minutes.

A stimulated 17-OHP between ≥ 9.8 but ≤ 30 nmol/L is an equivocal response and CAH is not excluded. Genotyping and/or a urine steroid profile is recommended.

A stimulated 17-OHP of ≥ 30 nmol/L is consistent with a diagnosis of CAH. Genotyping of the 21-hydroxylase gene and urine steroid profiling can be used to confirm the diagnosis.

Milder elevations of 17-OHP may be found in rarer forms of CAH: 11- β -hydroxylase deficiency and 3- β -hydroxysteroid dehydrogenase deficiency.

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