OBJECTIVE

Clinicians in Alberta provide urgent referral to an endocrinologist and optimize laboratory tests for investigation of adrenal insufficiency (Addison's disease)

TARGET POPULATION

Children and adults with signs or symptoms of primary or secondary adrenal insufficiency (Addison's disease)

EXCLUSIONS

None

RECOMMENDATIONS

- ✓ Request URGENT CONSULTATION with an endocrinologist
- ✓ Order a synthetic adrenocorticotropic hormone (ACTH) stimulation test to confirm diagnosis
- ✓ Administer glucocorticoids as soon as the stimulation test is completed (glucocorticoids can be a life saving measure)
- X DO NOT order serum cortisol levels. Low random serum cortisol levels have a poor predictive value and are not recommended. A serum cortisol with or without ACTH stimulation which exceeds 500-550 nmol/L excludes adrenal insufficiency[1]
- X DO NOT exclude secondary (pituitary) insufficiency as a cause with a normal response in the short ACTH stimulation test

Addison's Disease Clinical Features
• Loss of appetite and weight • Chronic worsening fatigue and muscle weakness • Low blood pressure decreasing further when standing, causing dizziness or fainting • Darkening skin on exposed and non-exposed areas of the body, particularly on skin creases and scars • Nausea, vomiting and diarrhea (occur in about 50% of cases) • Hypoglycemia (more severe in children than in adults)

PRACTICE POINT

If the diagnosis of Addison's disease is strongly suspected, treatment should be initiated immediately.

BACKGROUND

The most common cause of adrenal insufficiency is Addison's disease, a rare endocrine disorder that affects about one in 100,000 people. Addison's disease occurs in all age groups and afflicts men and women equally.

Disease occurs more frequently in families predisposed to autoimmune endocrinopathies, e.g., thyroid, Type 1 Diabetes Mellitus. Additionally, a normal response in the short ACTH stimulation tests does not exclude secondary (pituitary) insufficiency as a cause.

Addisonian crisis is a catastrophic complication of adrenocortical insufficiency which can develop rapidly. The patient usually develops adrenocorticol insufficiency symptoms followed by profound hypotension. The patient may remain alert. Because this crisis is life threatening, urgent **treatment and consultation cannot** await laboratory results.[1-4]

CAUSES

PRIMARY ADRENAL INSUFFICIENCY

- Seventy percent of reported cases are due to autoimmune disorders
- Tuberculosis accounts for about 20% of cases in developed countries
- Less common causes of primary adrenal insufficiency are chronic infections:
    - AIDS and fungal infections
    - Hemorrhage (secondary to anticoagulant therapy)
    - Cancer
    - Amyloidosis
    - Adrenalectomy

SECONDARY ADRENAL INSUFFICIENCY

- Lack of pituitary adrenocorticotropin (ACTH)

REFERENCES

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3. Chodosh LA, Daniels GH. Addison's disease. Endocrinologist, 1993; 3: 166-81.
4. Snow K, Jiang NB, Kao FC, Scherthauer BW. Biochemical evaluation of adrenal dysfunction: the laboratory perspective. Mayo Clinic Proc, 1992; 67: 1055-65.

SUGGESTED CITATION

Toward Optimized Practice (TOP) Endocrine Working Group. 2008 January. Laboratory endocrine testing: adrenal insufficiency (Addison's disease) clinical practice guideline. Edmonton, AB: Toward Optimized Practice. Available from: http://www.topalbertadoctors.org

For more information see www.topalbertadoctors.org

GUIDELINE COMMITTEE

The committee consisted of representatives of family medicine, medical biochemistry, pathology, internal medicine, endocrinology, laboratories and the public.

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