# Anaesthesia recommendations for patients suffering from Acute adrenal insufficiency

**Disease name:** Acute adrenal insufficiency  
**ICD 10:** E27.1-E27.4  
**Synonyms:** Addisonian Crisis

Acute adrenal insufficiency is a rare complication that can manifest in the perioperative period. The most common cause is exogenous steroid administration (tertiary adrenal insufficiency) resulting in atrophy of the adrenal cortex due to insufficient ACTH release from the pituitary gland, and the subsequent inability to produce endogenous glucocorticoids. Rarer causes of adrenal insufficiency are Addison's disease (primary adrenal insufficiency), which results in absence of all endogenous steroid production, as well as pituitary and hypothalamic tumors or lesions (secondary adrenal insufficiency). It should be noted that patients with Addison's disease are at higher risk for developing acute adrenal insufficiency due to a lack of both glucocorticoid and mineralcorticoid production.

Preoperatively, this is a diagnostic dilemma for clinicians. In cases of tertiary adrenal insufficiency, a history of exogenous steroid use within the past 3 months is a useful history to elicit. For primary and secondary presentations, the patient may manifest non-specific symptoms like nausea, vomiting, weight loss, and increased skin pigmentation. Electrolyte abnormalities like hypoglycemia, hyponatremia and hyperkalemia may be found on chemistries. However, most patients may be asymptomatic until the perioperative period when hypotension unresponsive to vasopressor and fluid support presents itself.

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**Medicine in progress**  
**Perhaps new knowledge**  
**Every patient is unique**  
**Perhaps the diagnostic is wrong**

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**Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net**
Disease summary

Patients with primary adrenal failure (for example, autoimmune Addison’s disease) are significantly more at risk of developing acute adrenal crisis because of the deficiency of both glucocorticoid and mineralocorticoid deficiency (in contrast to isolated glucocorticoid deficiency in cases of suppression of hypothalamus-pituitary-adrenal axis due to exogenous glucocorticoid treatment and other causes of secondary adrenal insufficiency, such as pituitary and hypothalamic lesions or surgery). It would be helpful to give cardinal clinical features and key biochemical findings (for example, hyponatraemia, hyperkalaemia and hypoglycaemia) of acute adrenal crisis to help clinicians to suspect the condition early.

Typical surgery

Not applicable.

Type of anaesthesia

Acute adrenal crisis manifests itself most often when the patient is under general anaesthesia. However, there have been cases of patients with persistent hypotension during regional and monitored anaesthesia care anaesthetics.

Necessary additional diagnostic procedures (preoperative)

In addition to the eliciting the history, physical exam findings, and symptoms listed in the disease summary, some clinicians may elect to perform an ACTH stimulation test. Although uncommonly done in the preoperative period, this allows assessing the degree of suppression of a patient's hypothalamic pituitary adrenal axis. The cortisol response to ACTH is usually similar to the maximal cortisol response that occurs after induction of general anaesthesia. An intact hypothalamic pituitary adrenal axis results in a cortisol level of greater than 19 micrograms/dL. Maximal cortisol response to general anaesthesia and surgery can range from 28 - 47 micrograms/dL.

Particular preparation for airway management

Not applicable.

Particular preparation for transfusion or administration of blood products

Not applicable.
Particular preparation for anticoagulation

Not applicable.

Particular precautions for positioning, transport or mobilisation

Not applicable.

Probable interaction between anaesthetic agents and patient's long-term medication

Not applicable.

Anaesthesiologic procedure

Refractory hypotension may increase risks of low perfusion events especially on induction of anaesthesia.

Particular or additional monitoring

May consider arterial invasive monitoring for refractory hypotension.

Possible complications

Perioperative Addisonian crisis is an extremely rare complication for anaesthesiologists, ranked in the literature in incidence from 0.01% to as high as 0.7%. It must be a consideration in patients with intraoperative or postoperative hypotension unresponsive to vasopressors and fluids, especially those with a known history of Addison's disease, pituitary/hypothalamic pathologies, and/or a known history of steroid use. If there is a suspicion intraoperatively, the patient must be immediately treated with stress dose steroids, which is traditionally hydrocortisone 100mg IV, and must be maintained every 8 hours for the first 24 hours and then can be tapered to the patient's maintenance dose or discontinued altogether.

In order to prevent this complication, anaesthesiologists must decide the necessity of steroid supplementation. This is a difficult decision as the literature in recent years has been unequivocal on prophylactic administration of stress-dose steroids versus waiting for the manifestation of clinically significant hypotension intraoperatively. However, it is universally accepted that patients with known Addison's disease or pituitary or hypothalamic processes should receive additional supplementation in addition to their maintenance dose depending on the stress of the procedure which will be discussed in the following paragraph.

In patients who are on chronic steroid supplementation for other pathologies, these are the general recommendations. Patients who receive Prednisone 5mg or less daily usually do not require additional supplementation, but this does not preclude them from developing from acute adrenal crisis. Those who receive higher dosages will be administered intravenous
steroids according to the stress of the procedure. For minor surgical or medical stress (GI or GU procedures, or those with mild illnesses like nausea or vomiting), the recommendations include hydrocortisone 25mg or methylprednisolone 5mg on the day of the procedure. For moderate surgical or medical stress (laparoscopic procedures or more severe illnesses like pneumonia), the recommendations include hydrocortisone 50-75mg or methylprednisolone 10-15mg on the day of procedure, with a taper to the expected dose of steroids 1-2 days after the procedure. Finally, for severe medical or surgical stress (cardiac, thoracic, hepatic resections, etc), the recommendations include hydrocortisone 100-150 mg or methylprednisolone 20-30mg on the day of procedure with a taper over 1-2 days to the patient’s normal dose.

Postoperative care

Addisonian crisis with hypotension also occurs in the postoperative period. In addition, it is imperative to ensure adequate steroid delivery, which may require prolonged intravenous dosing if oral route is not available.

Information about emergency-like situations / Differential diagnostics

cause by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease

Here is a case demonstrating acute adrenal insufficiency: A 61 year-old male presents for a muscle biopsy to help elucidate the cause of his new onset rhabdomyolysis. He has a past medical history of type I diabetes mellitus, kidney transplantation on low dose prednisone, coronary artery bypass grafting, hypertension, hyperlipidemia, and idiopathic hypertrophic subaortic stenosis. It should be noted he did take his Prednisone the day of the procedure. His case was performed under monitored anaesthesia care with stable intraoperative hemodynamics. However, in the recovery room, the patient remained persistently hypotensive, with transient responses to vasopressors and fluid boluses. After eliminating alternative causes of his shock state, hydrocortisone 100mg was administered with immediate stabilization of his hemodynamics. A post-operative cortisol level was obtained and was measured to be 4.0ug/dL, which was consistent with a diagnosis of adrenal insufficiency.

Ambulatory anaesthesia

See complications section.

Obstetrical anaesthesia

The obstetrical population must be considered under severe medical or surgical stress, and must be treated with an appropriate intraoperative steroid dose.
Literature and internet inks

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