Acute Pituitary Failure

Synonyms: pituitary apoplexy, hypopituitary coma

Hypopituitarism refers to deficiency of one or more hormones produced by the anterior pituitary or released from the posterior pituitary.[1] Acute pituitary failure is a rare medical emergency presenting with the sudden onset of headache (with or without neurological symptoms involving the second, third, fourth and sixth cranial nerves), vomiting, visual impairment and decreased consciousness.

It is caused by haemorrhage and/or infarction of the pituitary gland. Rapid replacement with hydrocortisone may be life-saving. Surgery is required urgently for patients with worsening neurological symptoms but it is otherwise currently unclear whether conservative or surgical management carries the best outcome. After acute pituitary failure, there needs to be careful monitoring for recurrence of tumour growth. [2]

Epidemiology[2]

- Acute pituitary failure occurs in a small but significant number of patients with a pituitary tumour, most often a clinically non-functioning macroadenoma.
- The incidence of acute pituitary failure in pituitary adenomas is between 2% and 7%.
- Acute pituitary failure usually occurs in patients with pre-existing pituitary adenomas and evolves within hours or days. In over 80% of patients, acute pituitary failure is the first presentation of the underlying pituitary tumour.
- Most cases occur during the fifth decade of life, with a predominance in males. [3]
- Precipitating factors for acute pituitary failure include:
  - Systemic hypertension.
  - Major surgery, in particular coronary artery bypass surgery.
  - Dynamic pituitary function tests with GnRH, TRH and CRH.
  - Anticoagulation therapy.
  - Coagulopathies.
  - Oestrogen therapy.
  - Initiation or withdrawal of dopamine receptor agonist.
  - Radiation therapy.
  - Pregnancy.
  - Head trauma.

Patients known to have a pituitary tumour must be observed for signs and symptoms of acute pituitary failure when performing pituitary stimulation tests, starting anticoagulation therapy or when having coronary artery bypass or other major surgery.

Postpartum pituitary necrosis (Sheehan’s syndrome)[4]

Sheehan’s syndrome is postpartum hypopituitarism caused by necrosis of the pituitary gland. It is usually the result of severe hypotension or shock caused by massive haemorrhage during or after delivery.

Patients with Sheehan’s syndrome have varying degrees of anterior pituitary hormone deficiency.

As a result of advances in obstetric care in developed countries, Sheehan’s syndrome is a rare cause of hypopituitarism. It remains common in underdeveloped and developing countries.

Sheehan’s syndrome usually presents progressively with a variety of developing symptoms (starting commonly with failure to lactate, breast involution and then amenorrhoea), rather than the more dramatic and acute presentation.

The more dramatic acute presentation of acute pituitary failure only occurs in a minority of patients with Sheehan’s syndrome.[5]

Presentation[6, 7]

A diagnosis of acute pituitary failure should be considered in all patients presenting with acute severe headache with or without neurological and ophthalmic signs. [2]

History

The onset of symptoms is sudden and may occur in patients known to have a pituitary adenoma.

- Headache occurs in 95% of patients. It often starts suddenly as a unilateral, retro-orbital headache before becoming more generalised. It is thought to be caused by stretching of the dura mater in the sella. [8]
- Vomiting occurs in 69% of cases. It often accompanies the headache and may be caused by elevation of intracranial pressure or irritation of meninges.
- Diplopia may occur in the conscious patient, from involvement of different cranial nerves.
Visual field defects occur in 64% of patients, by compression of the optic chiasm. Defects in acuity may result from optic nerve compression. Ptosis may occur.

Examination findings
There may be rapid progression to coma, suggesting stroke, subarachnoid haemorrhage and with accompanying meningism, even meningitis. Careful and early examination may detect a variety of findings. These may include:

- Bitemporal superior quadrantic field defect, which is classic. There may be a contralateral homonymous hemianopia if the optic chiasm is affected differently.
- Compression of the cavernous sinus may impair cranial nerves III, IV and VI, causing ocular paresis and hence diplopia (in the conscious patient).
- Horner’s syndrome, which may develop if sympathetic fibres are damaged.
- Stroke, which can occur if the carotid syphon is compressed against the anterior clinoid process. This may be associated with meningism, stupor and coma.
- Thermal regulation, which may be impaired by hypothalamic involvement. Hypothermia can result.
- Endocrine deficiencies, which can result from destruction of adenohypophyseal tissue.
- Complete ophthalmoplegia.

Differential diagnosis
As the patient is often not known to have a pituitary tumour, the diagnosis of acute pituitary failure can often be difficult and delayed with a wide differential diagnosis, including:

- Subarachnoid haemorrhage
- Meningitis
- Brain tumours
- Demyelinating disease
- Cerebrovascular disease

Assessment

- Initial assessment should include a detailed history focusing on symptoms of pituitary dysfunction followed by a thorough physical examination including cranial nerves and visual fields.
- Formal visual fields assessment must be undertaken when the patient is clinically stable, preferably within 24 hours of the suspected diagnosis.
- All patients with suspected acute pituitary failure should have urgent blood samples drawn to check electrolytes (hyponatraemia), blood glucose, renal function, liver function, clotting screen, FBC, and random cortisol, prolactin, TFTs (FT4 and TSH), insulin-like growth factor 1 (IGF-1), growth hormone, LH, FSH, testosterone in men, and estradiol in women.
- To confirm the diagnosis, urgent MRI scan must be done in all patients with suspected acute pituitary failure.
- A pituitary CT scan is indicated if an MRI scan is either contra-indicated or not possible.

Management

Initial management
The immediate medical management of patients with acute pituitary failure should include careful assessment of fluid and electrolyte balance, replacement of corticosteroids and supportive measures to ensure haemodynamic stability.

- Resuscitation and stabilisation of the patient, with particular attention to correction of fluid and electrolyte abnormalities.
- In haemodynamically unstable patients, intravenous hydrocortisone should be administered after baseline endocrine function tests.
- Patients who remain clinically and neurologically unstable require urgent transsphenoidal surgical decompression as definitive treatment.

Once the diagnosis has been confirmed, it is recommended that all patients should be transferred once medically stabilised to the local neurosurgical/endocrine team as soon as possible.

Steroid therapy

- Patients with acute pituitary failure, who are haemodynamically unstable, should be commenced on empirical steroid therapy. In adults, hydrocortisone 100-200 mg as an intravenous bolus is appropriate, followed either by 2-4 mg per hour by continuous intravenous infusion.
- Indications for empirical steroid therapy in patients with acute pituitary failure are haemodynamic instability, altered consciousness level, reduced visual acuity and severe visual field defects.
- Patients who do not fulfil the criteria for urgent empirical steroid therapy should be considered for treatment with steroids if their 9 am serum cortisol is less than 550 nmol/L.

Further management
Further neuro-ophthalmic assessments, to check the visual acuity, visual fields and ocular paresis must be undertaken, when the patient is clinically stable.
**Conservative management**

- Patients with acute pituitary failure, who are without any neuro-ophthalmic signs or who have mild and stable signs, can be considered for conservative management with careful monitoring.
- In patients with reduced visual acuity or defective visual fields, formal assessment of visual fields and acuity should be performed every day until there is a clear trend of improvement.
- Acutely unwell patients should have neurological assessment every hour and any deterioration in neurological status should prompt consideration to proceed with surgery.
- The frequency of neurological assessment could be gradually reduced to every four to six hours, when the patients are stable.
- Renal function and electrolytes should be checked daily.
- The presence of a new or deteriorating visual deficit or neurological deterioration should prompt further urgent imaging with a view to decompressive surgery (including external ventricular drain (EVD) placement in the presence of hydrocephalus).

**Surgical management**

- Patients with severe neuro-ophthalmic signs, such as severely reduced visual acuity, severe and persistent or deteriorating visual field defects or deteriorating level of consciousness, should be considered for surgical management.
- Ocular paresis due to involvement of III, IV or VI cranial nerves in cavernous sinus in the absence of visual field defects or reduced visual acuity is not in itself an indication for immediate surgery. Resolution will typically occur within days or weeks with conservative management.
- Surgery should be performed preferably within the first seven days of symptoms.
Long-term monitoring and surveillance

Patients should be followed up for regrowth of the tumour and endocrine abnormalities.

- All patients with acute pituitary failure should have an endocrine review at four to eight weeks after the event. Assessment should include pituitary function and formal assessment of visual acuity, eye movements and visual fields.
- Patients treated for acute pituitary failure should have an annual thorough assessment of pituitary function.
- Both conservatively and surgically treated patients need close radiological follow-up and, if residual tumour or recurrence is detected, additional treatment such as radiotherapy or further surgery should be considered.
- An MRI scan is recommended at three to six months after acute pituitary failure. Then an annual MRI scan should be considered for the following five years, then two-yearly.

Prognosis

- Acute pituitary failure is a potentially life-threatening condition with a high mortality.\[12\]
- It is difficult to diagnose and difficult to treat.\[13\]
- However, with rapid diagnosis, followed by prompt correct management, it is possible to achieve complete recovery.\[14, 15\]

Further reading & references

- The Pituitary Foundation
- Guidelines for the Management of Pituitary Apoplexy; Society for Endocrinology (January 2011)
- Weiss RE; Hypopituitarism: Emergencies. last updated April 2015.

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