Acromegaly

Excessive secretion of GH in adults often by adenoma in anterior pituitary resulting in skeletal, connective, and soft tissue overgrowth. In children excess GH is called gigantism.

ANESTHETIC CONSIDERATIONS:

- Potentially difficult BMV
- Potential difficult larygoscopy and intubation
  - Enlarged tongue and epiglottis
  - Prognathia
  - Subglottic diameter may be decreased (smaller ETT)
  - Recurrent laryngeal nerve palsy and vocal cord paralysis
- Potential for increased ICP (pituitary tumor)
- Altered respiratory function:
  - Lung volumes increased
  - V/Q mismatching increased
- Associated medical comorbidities:
  - HTN/LVH, diastolic dysfunction, CAD, valvular abnormalities
  - DM and impaired glucose tolerance
  - Hypothyroidism
  - Peripheral neuropathy and entrapment syndromes
- Skeletal muscle weakness and fatigue

ANESTHETIC GOALS:

1) Anticipate and appropriately manage possible difficult airway (consider awake fiberoptic intubation)
2) Maintain hemodynamic stability and optimize cardiac/ pulmonary comorbidities
3) Watch perioperative and intraoperative glucose levels
4) If concerned about ICP, anesthetic considerations and goals for increased ICP

HISTORY

- Manifestations of acromegaly reflect parasellar extension of the anterior pituitary adenoma and peripheral effects produced by the presence of excess growth hormone
- Generalized complaints of fatigue
- Symptoms of upper airway obstruction secondary to enlarged tongue and epiglottis
- Symptoms of sleep apnea (obstructive and central)
- Hoarseness and abnormal movement of the vocal cords secondary to thickened vocal cords or paralysis of a recurrent laryngeal nerve due to stretching by overgrowth of cartilaginous structures
- Involvement of the cricoarytenoid joints can result in alterations in the patient's voice due to impaired movement of the vocal cords
- Dyspnea or stridor secondary to subglottic narrowing
- Symptoms of CAD and heart failure (chest pain, dyspnea)
- Headache and papilledema reflect increased intracranial pressure due to expansion of the anterior pituitary adenoma
- Visual disturbances (bitemporal hemianopsia) are due to compression of the optic chiasm by the expanding overgrowth of surrounding tissues
- Peripheral neuropathy secondary to trapping of nerves by skeletal, connective, and soft-tissue overgrowth
- Artery entrapment eg. ~50% of patients have compromised ulnar artery flow in one or both hands– carpal tunnel syndrome
- Glucose intolerance/DM secondary to effects of GH on carbohydrate metabolism (polydipsia)
- Hypothyroidism
- Increased incidence of systemic hypertension, ischemic heart disease, osteoarthritis, and osteoporosis

PHYSICAL

- Thick and oily skin, weak skeletal muscles
- Airway exam – distorted facial and upper airway anatomy, increased length of mandible and enlarged jaw (macrognathia), polypoid masses, enlarged lips, tongue, epiglottis
- Cardiopulmonary exam
  - Blood pressure
  - Signs of LVH, diastolic dysfunction, murmurs for valvular abnormalities, heart failure
  - Lung volumes are increased, and ventilation-to-perfusion mismatching may be increased
- Skeletal overgrowth (prognathism)

INVESTIGATIONS

- Diagnosis of acromegaly:
  1) Plasma GH does not decrease 1-2 hours post-ingestion of 75-100g glucose
  2) GH concentrations higher than 3 ng/ml
  3) CT/MRI head
- Labs:
  - CBC (Hb, plts), lytes, glucose, thyroid function tests
- Imaging:
  - ECG for LVH
  - Echocardiogram
  - Lateral neck films or CT scans of the neck and direct or indirect visualization can identify patients with subglottic stenosis or an enlarged tongue, mandibles, epiglottis, or vocal cords
  - Skull radiograph and CT for detecting enlargement of the sella turcica (characteristic of anterior pituitary adenomas)
  - CT or MRI (not only for tumour but if there are concerns about increased ICP [suprasellar mass])
  - Nasal culture (to guide Antibiotic therapy in event of post-op infection)

OPTIMIZATION

- Profound hypocortisolism (and hyponatremia) should be corrected preoperatively
- Hypothyroidism should be sought and corrected preoperatively

- Pre-op: stress-dose steroids/steroid replacement if hypopituitarism (will need to continue steroids periop for pituitary surgery)
- Planning for difficult airway management plus discussion with the patient of possible awake fiberoptic intubation is necessary

TREATMENT

- Primary treatment is transsphenoidal resection of tumour
- If the pituitary tumor is not totally removed, patients are often offered external pituitary irradiation
- In the case of suprasellar extension, conventional transfrontal hypophysectomy is often performed
- When adenomas have extended beyond the sella turcica, surgery or radiation is no longer feasible; medical treatment with suppressant drugs (bromocriptine) may be an option. Bromocriptine (dopamine agonist) can lower GH levels
- Octreotide, a long-acting analog of somatostatin produces effective palliation in 50% of patients (depot injection monthly)
- Other medical therapies such as pegvisomant or somatostatin analogs are also medications that have been tried before surgery

ANESTHETIC OPTIONS

- GA
- Regional – Skeletal changes may make performance of regional anesthesia difficult
- Local

ANESTHETIC SETUP

Monitors:

- Standard CAS monitors
- Arterial line - If placement of an arterial line is necessary, a brachial or femoral site may be preferable to a radial site due to possible inadequate collateral circulation
- Nerve stimulator
- Visual Evoked Potentials (VEP) – compression of chiasm and perfusion to optic nerve
- Precordial Doppler and right atrial catheterization for transphenoidal surgery (air embolism)

MANAGEMENT OF ANESTHESIA

Induction

- The incidence of difficult intubation is 20 to 30%, and may be clinically unpredictable
- Consider awake fiberoptic intubation
- Possible difficult mask ventilation
- Narrowed glottic and subglottic opening may necessitate use of a smaller internal diameter tracheal tube than would have been predicted based on the patient's age and size
- Nasal turbinate enlargement may preclude the passage of nasopharyngeal or nasotracheal airways
- Induction as you would for a normal case since ICP usually not raised
- Alter your technique if concerns with ICP (awake art line, blunt sympathetic response to laryngoscopy, no use of drugs which raise ICP)

Maintenance:

- Monitoring blood glucose concentrations is useful if diabetes mellitus or glucose intolerance accompanies acromegaly
- Doses of nondepolarizing muscle relaxants are guided by the use of a peripheral nerve stimulator, particularly if skeletal muscle weakness exists before anesthesia induction
- There is no evidence that hemodynamic instability or alterations in pulmonary gas exchange accompany anesthesia in acromegalic patients
- Surgical preferences for CO_2 management vary:
    - Hypocapnia can reduce brain volume and minimize the degree to which the arachnoid bulges into the sella
    - If suprasellar extension of a tumor, normal or increased CO_2 helps to deliver the lesion into the sella for excision
    - Some surgeons have resorted to "pumping" of saline or air into the lumbar CSF space as an alternative

Emergence:

- Smooth emergence especially if CSF space opened and resealed with fibrin glue and sphenoid packed with fat
- Avoid valsalva (cough, vomit) – may re-open CSF leak and risk meningitis
- Clean airway (blood debris)
- Extubate fully awake and following commands (will be mouth-breathing since nasal packing)

DISPOSITION & MONITORING

- ICU
- Monitor for:
    - Blood loss/active bleeding
    - DI (careful attention to fluid balance) – commonly seen during first 12 hours post-op lasting 2-4 days)

COMPLICATIONS

- Post-operative respiratory complications
- Air embolism
- Blood loss
- Cavernous sinus (immediate lateral relation of the pituitary)
- Carotid arteries (pass through the cavernous sinuses)
- Some patients have a venous sinusoid that lies in front of the pituitary gland and connects the two cavernous sinuses
- Cranial nerve weakness due to trauma or stretching
- Visual complications (damage to optic nerve or chiasm)
- CSF rhinorrhea
- Hypothalamic injury/stroke
- Cerebral ischemia
- Meningitis

PATHOPHYSIOLOGY

- More than 99% of cases of acromegaly are attributable to pituitary adenoma (or use of recombinant GH for unapproved aging prevention)
- Generally, as a pituitary lesion expands and compresses the pituitary tissue, the sequence in which hormonal function is lost is first gonadotropins; second, growth hormone; third, ACTH; and fourth and last, thyroid-stimulating hormone

REFERENCES: Barash Pg. 1301; Miller Chpt 34, 35; Coexisting Chpt 16