

Acromegaly

Diagnosis, comorbidity management, surgery, medical therapy, radiotherapy, and special circumstances for adults with acromegaly.

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Acromegaly Clinical Practice Guideline
Katznelson et al. • JCEM 2014;99:3933–3951

GRADE

1 • Strong For

2 • Suggest For

2- • Suggest Against

3 • Strong Against

Ungraded

1 Diagnosis

1 Measure **IGF-1** in patients with typical clinical manifestations (acral/facial features). **Suggest (2)** IGF-1 even without typical features if sleep apnea, T2DM, debilitating arthritis, carpal tunnel syndrome, hyperhidrosis, or hypertension present.

3 **Against** relying on random GH levels to diagnose acromegaly; confirm elevated/equivocal IGF-1 with an **oral glucose load** — lack of GH suppression to $<1 \mu\text{g/L}$ confirms diagnosis.

1 After biochemical diagnosis, obtain a pituitary **MRI** (imaging of choice; CT if MRI contraindicated/unavailable) to assess tumor size and parasellar extent. Suggest formal **visual field testing** if the tumor abuts the optic chiasm.

2 Comorbidities

2 Evaluate all patients for **hypertension, diabetes, cardiovascular disease, osteoarthritis, and sleep apnea**; monitor longitudinally and manage rigorously (ungraded).

2 Screen for **colon neoplasia** with colonoscopy at diagnosis; obtain a **thyroid ultrasound** if palpable nodularity.

1 Assess for **hypopituitarism** and replace hormone deficits.

3 Goals of Management

2 Target an **age-normalized IGF-1** and random **GH $<1.0 \mu\text{g/L}$** to signify biochemical control. Use the **same GH/IGF-1 assay** consistently in a given patient throughout management.

4 Surgery

1 **Transsphenoidal surgery** is primary therapy for most patients; consider repeat surgery for residual intrasellar disease.

2- **Against** routine preoperative medical therapy to improve postop biochemical control — exception: SRL therapy for severe pharyngeal thickening/sleep apnea/high-output heart failure to reduce surgical risk.

2 **Surgical debulking** when parasellar disease makes total resection unlikely, to improve response to subsequent medical therapy.

2 **Postop**: measure IGF-1 and random GH ≥ 12 weeks after surgery (nadir GH after glucose load if GH >1); obtain imaging (MRI preferred) ≥ 12 weeks postop.

5 Medical Therapy

1 Medical therapy indicated for **persistent disease** after surgery.

2 Significant disease (moderate–severe symptoms, no mass effect) → **SRL or pegvisomant** as initial adjuvant. Modest IGF-1 elevation/mild symptoms → trial of a **dopamine agonist** (cabergoline).

2- **Against** routine abdominal ultrasound for gallstones on SRL therapy (only if symptomatic); add pegvisomant or cabergoline if inadequate SRL response.

2 On **pegvisomant**: serial MRI for tumor size; monthly LFTs for 6 months then every 6 months, consider discontinuing if transaminases $>3\times$ ULN.

2 **SRL as primary therapy** if not curable by surgery, extensive cavernous sinus invasion without chiasmal compression, or poor surgical candidate.

6 Radiotherapy

2 **RT** for residual tumor after surgery when medical therapy is unavailable/unsuccessful/not tolerated; prefer **stereotactic RT** over conventional unless significant residual burden or chiasm proximity (exposure $>8 \text{ Gy}$).

1 **Annual GH/IGF-1** reassessment off medication to monitor RT efficacy; annual hormonal testing post-RT for hypopituitarism/delayed effects.

7 Special Circumstances

1 **Gigantism** — use standard diagnostic/therapeutic approach as for adult-onset acromegaly.

2 **Pregnancy**: discontinue long-acting SRL/pegvisomant ~ 2 months before conception (bridge with short-acting octreotide if needed); withhold medical therapy during pregnancy except for tumor/headache control; serial visual field testing if macroadenoma; against routine GH/IGF-1 monitoring during pregnancy.