CPD PRESENTATION

TOPIC : **ACROMEGALY**

PRESENTER : Dr. **SANYU DONAH**
INTERNIST

WE WILL START WITH A CASE

CHIEF COMPLAINTS

32 years old lady complaining

- persistent headaches
- loss of vision
- facial and extremity deformity

History of present illness

- Headache since 1999 which was diffuse throbbing, not changing along the day, associated with blurred vision.

- Since 2006 started to have progressive loss of visual acuity and seeing her face and extremities enlarging gradually

HPI CON'T

- She consulted many times for headache and then in 20014 was referred for vision loss.

- Referred to ophthalmologist and CT brain was requested .

Review of systems

- She reports sweating, irregular menses, palpitations while sleeping, no dizziness, no voice changes, no heat and cold intolerance.

PMH, FH, SH

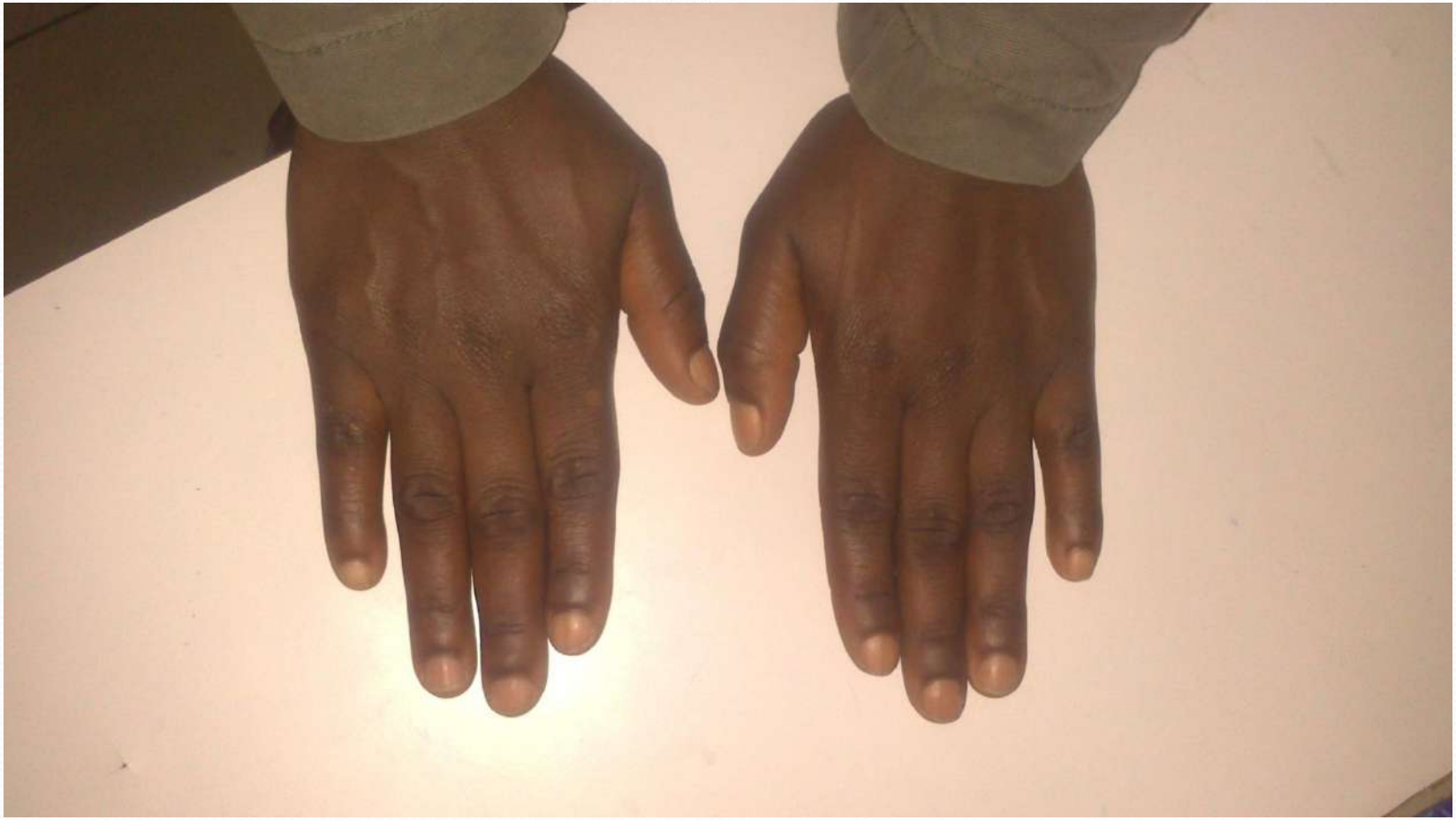
- No chronic illnesses
- Family history noncontributory
- She is an orphan lives with 2 sisters
- No smoking nor alcohol intake reported

Physical exam

- BP:128/86, pls:81, Oxy sat:98%, temp:36.8
- She is stable, no pallor, no clubbing, and no LN, no edema.
- Fully conscious, complete loss of vision in left, and temporal field defect in right eye.

Physical Exam (Nxt

- Appears with prominent skull growth and supra orbital ridges, enlargement of lips, nose and tongue.
- Enlargement of hands, fingers, feet and toes.
- no Galactorrhea
- The rest of the examination (CVS, Respiratory, GIT) was unremarkable.



Summary

- This 32 year old lady presented with persistent headache, progressive loss of visual acuity . P/E revealed facial bones and distal extremities enlargement, signs of optic chiasma compression and irregular menses.

INVESTIGATIONS

- **Lab work up:**
- -FBC: normal, fasting glycaemia; 7.5 mmol/l
- LFTs, U/E, TFTs normal
- - Growth hormone at 50, 23 ng/mL, prolactin 150 ng/dl
- **-Imaging:**
- EKG: LVH

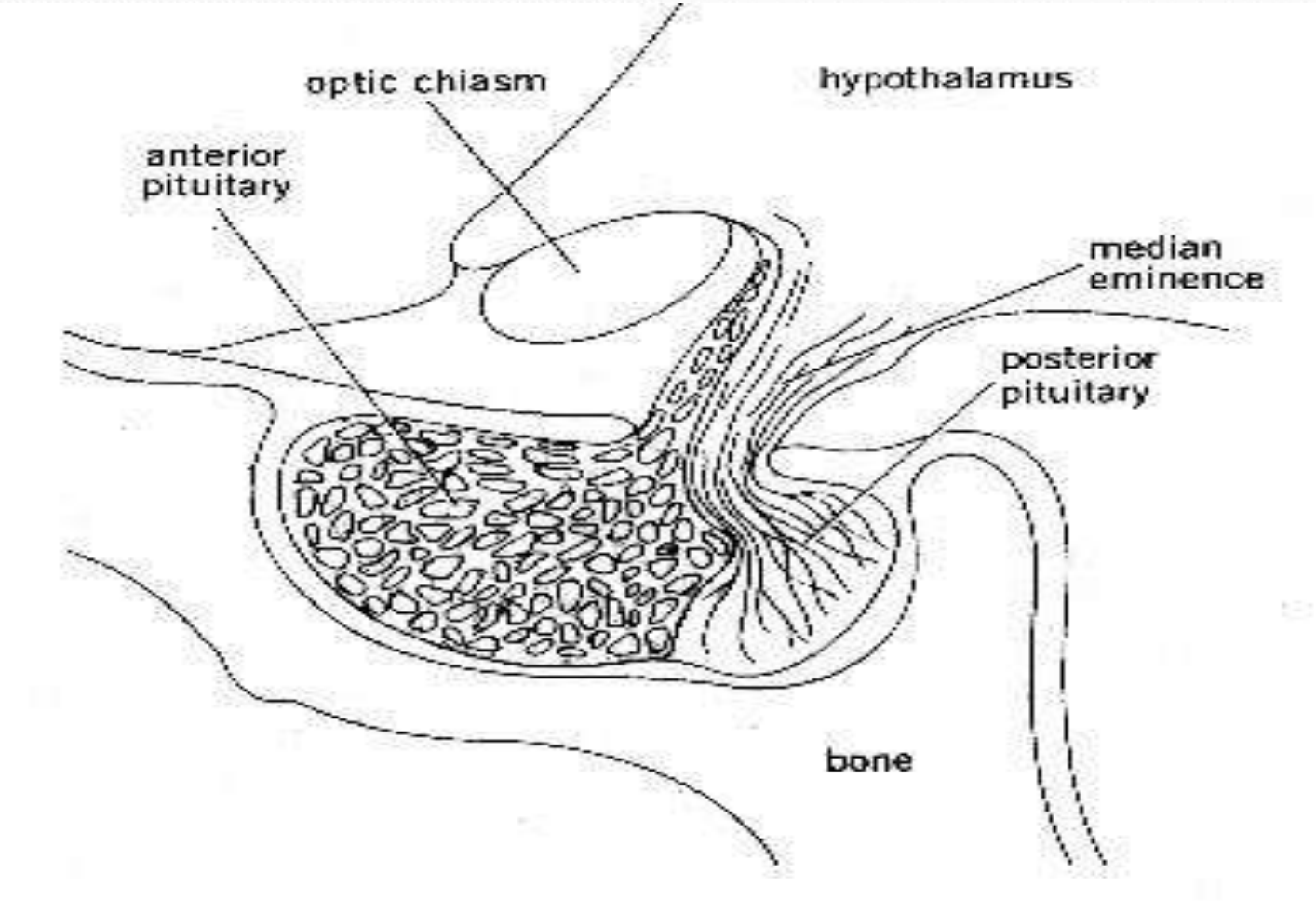
- Head MRI on 5/09/2013:
- Features suggestive of pituitary macro adenoma with no evidence of hemorrhage: There is a large sella tumor with suprasellar extension, which appears homogenous in nature & enhances moderately and continuously with no plateauing. The tumor measures about 8.0cms in depth, 2.0cms in width and about, 2.6cms in length. There is displacement optic chiasma, bilaterally. There is no adjacent bone erosion. There is mild displacement of vascular structures, bilaterally.

Acromegaly

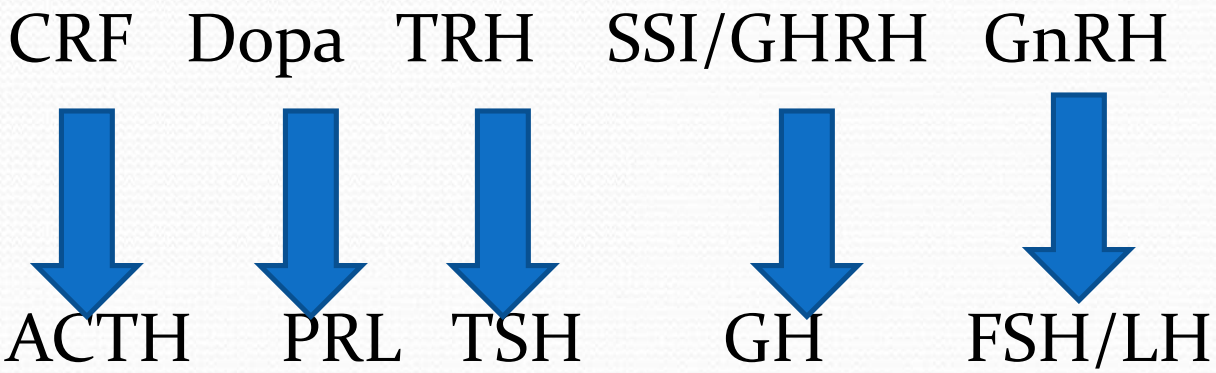
Objectives

- Introduction
- Incidence/epidemiology
- The role of IGH-1 and GH
- Approach to management
- Prognosis

Introduction: Anatomy



Introduction: 5 functional anterior hypothalamo-pituitary axis



Introduction cont

- This rare disease is due to hypersecretion of GH from a pituitary adenoma.
- IGH-I regulates the effect of GH in your body
- Its progression is usually very slow.
- Interval from the onset of symptoms until diagnosis is about 12 years

Incidence

- Male to Female ratio is 3: 2
- It usually presents between the ages of 30 and 50yrs old.
- Its annual incidence is 3-4 per million people per year world wide.
- Prevalence of 38-69 cases per million

Role of IGH-I and GH

- Growth hormone (GH) is produced by the pituitary somatotroph cells
- IGF-I that is synthesized in the liver and secreted into the blood is under the control of growth hormone (GH).

The role of IGH-I and GH cont

- GH stimulates the liver to produce IGH-I in blood.
- Excess GH and IGF-I have both somatic and metabolic effects
- The somatic effects include stimulation of growth of many tissues, such as skin, connective tissue, cartilage, bone, viscera, and many epithelial tissues.

- Random GH is not useful because it is pulsatile
- GH excess stimulates gluconeogenesis and lipolysis
- There is impaired glucose uptake in muscles in acromegaly
- The clinical features of acromegaly are attributable to high serum concentrations of both GH and insulin-like growth factor-I (IGF-I), which is GH-dependent.

Role of IGH-I and GH cont

- Clinical findings are due excessive soft tissue over growth and local mass effect.
- which include headache, sweating, skull growth, prominent supra orbital ridge, macro glossia, arthralgia, deepening voice ,Sleep apnoea, Enlargement of hands, fingers, feet and toes.
- Carpal tunnel syndrome, cardiomyopathy
- DM
- HTN

DIAGNOSTIC TESTS

- . Visual fields
- GH measurement
- serum insulin-like growth factor-1 (IGF-1
- Glucose tolerance test
- MRI (or CT) scan of pituitary fossa
- ECG. Echocardiogram

MEDICAL TREATMENT

3 classes of drugs

- Somatostatin analogues
- Dopamine agonists
- GH receptor antagonist

GHR antagonist

Pegvisomant

- Binds to peripheral GH receptors blocking function
- Daily injection

SRLs

Octreotide and Lanreotide

Target the somatostatin 2 and 5 receptors in the pituitary and directly on the liver to inhibitIGH-I synthesis.

75% will see 25% tumour shrinkage

Biochemical control in 34-45% of patients

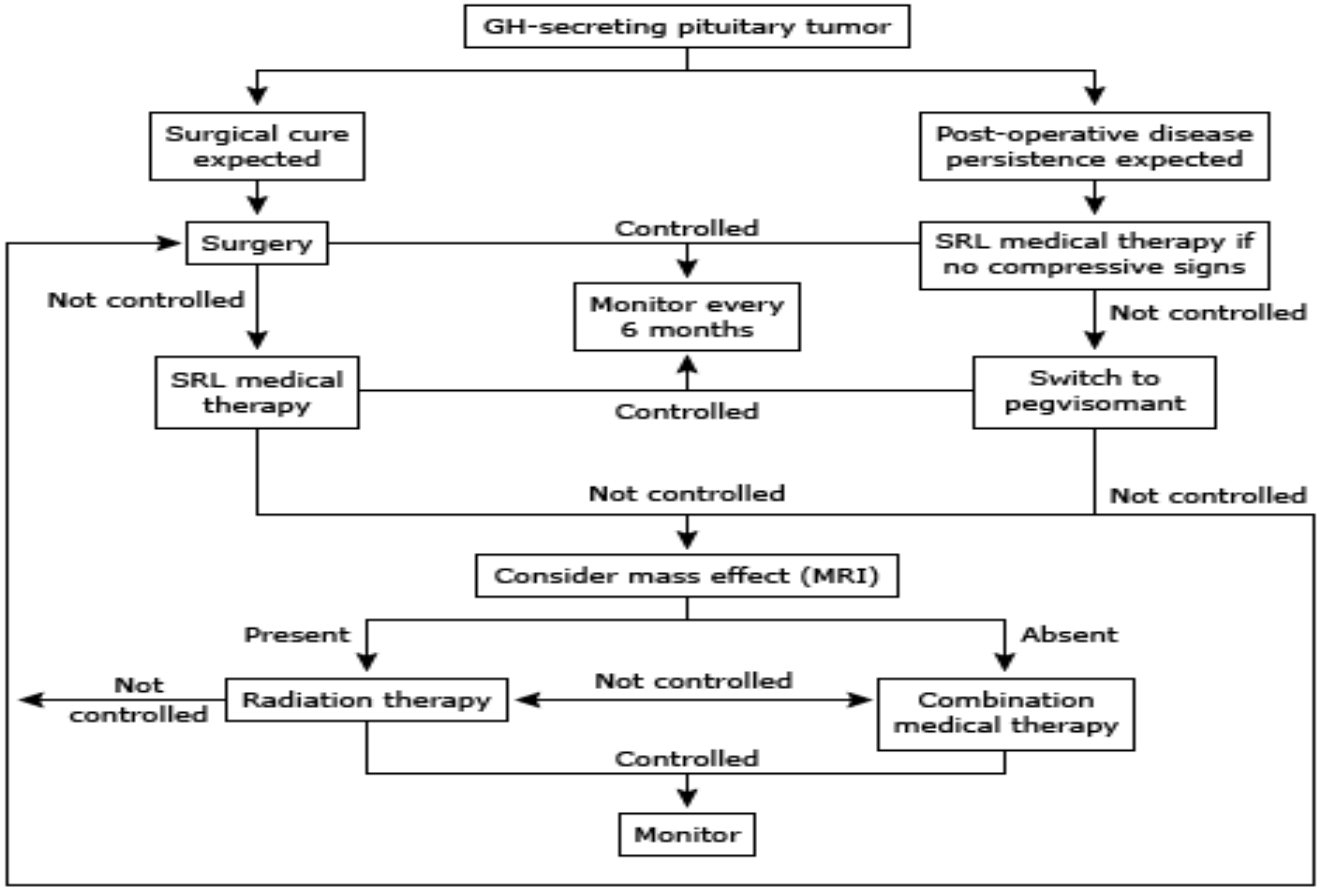
Dopamine agonists

- Bromocriptine
- Normally dopamine agonists stimulate GH secretion in normal individuals, yet suppress GH secretion in acromegaly patients
- A potent D2- dopamine agonist suppresses GH
- Studies show that D2 agonists have direct inhibitory effect on pituitary.

SURGICAL MANAGEMENT

- Trans sphenoidal surgery to debulk the tumour
- Radiotherapy

Summary of management strategy for patients with acromegaly



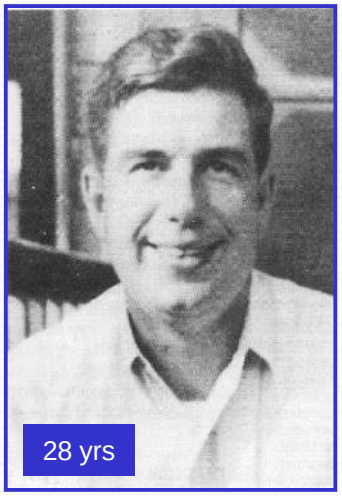
Summary of management strategy for patients with acromegaly. Control is defined by GH and IGF-I measurements.
SRL: somatostatin receptor ligands.
Adapted with permission from: Melmed S, Colao A, Barkan M, et al. Guidelines for acromegaly management: An update. J Clin Endocrinol Metab 2009; 94:1509.

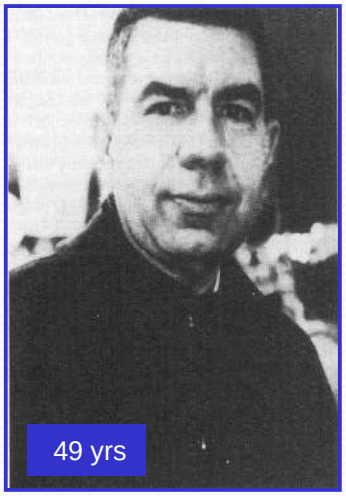
Prognosis

Survival determinants and causes of death in acromegaly

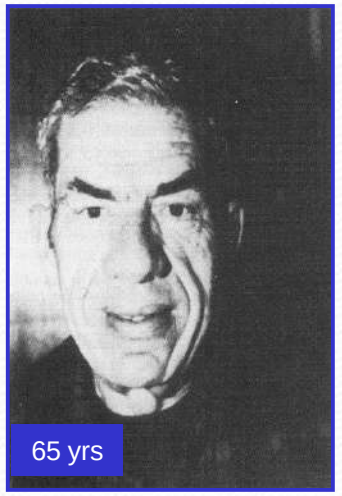
Survival determinants
Last known GH level
Hypertension
Cardiac disease
Diabetes mellitus
Symptom duration
Causes of death
Cardiovascular - 38 to 62 percent
Respiratory - 0 to 25 percent
Malignancy - 9 to 25 percent

- Has 2 -3 fold increase in premature mortality especially from hypertrophic cardiomyopathy.

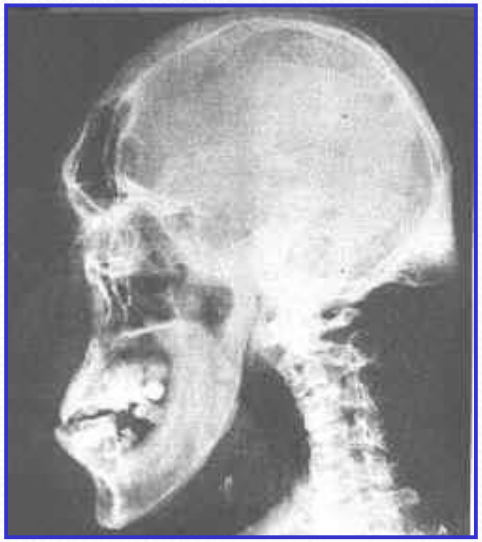

28 yrs


49 yrs


55 yrs


65 yrs

Frontal bossing, large jaw, dental maloclussion



THANK YOU