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Pituitary masses and malfunction - role of an endocrine nurse

Warrick Inder

Endocrinologist, Princess Alexandra Hospital
Associate Professor, University of Queensland
Brisbane



Early 1995...

- 14 year old boy referred with growth failure and delayed puberty
- Seen by the 3rd year advanced trainee
- GH stimulation tests arranged
- Endocrine nurse asks
- **“why doesn’t the boy just get some of his mother’s spare growth hormone?”**
- Patient has normal GH status and responds well to a short course of testosterone injections to initiate puberty



Later in 1995

- Index patient's mother referred to Endocrinology Department for assessment of hirsutism
- Seen by one of the senior consultants
- Acromegaly diagnosed...



Outline



- Too big
 - Non-functioning pituitary adenomas
 - Other sella and suprasellar lesions
- Too high
 - Prolactinoma, acromegaly, Cushing's
- Too low
 - Hypopituitarism



Endocrinology - principles



- 4 reasons why a patient presents to an endocrinologist
 - too much hormone
 - acromegaly
 - too little hormone
 - hypopituitarism
 - hormone not working
 - thyroid hormone resistance
 - a “lump”
 - mass effect e.g. pituitary tumours causing headache, visual field problems.



Pituitary disease - Background



- Pituitary adenomas are the commonest intracranial neoplasm
 - 10-15% of such lesions
- Clinically apparent tumours in 1 in 1000 people
- 35-40% non-functioning and prolactinoma
- 10-15% acromegaly (GH) and Cushing's (ACTH)
- <1% secrete TSH
- Other non-adenomatous lesions important to recognise



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Too Big



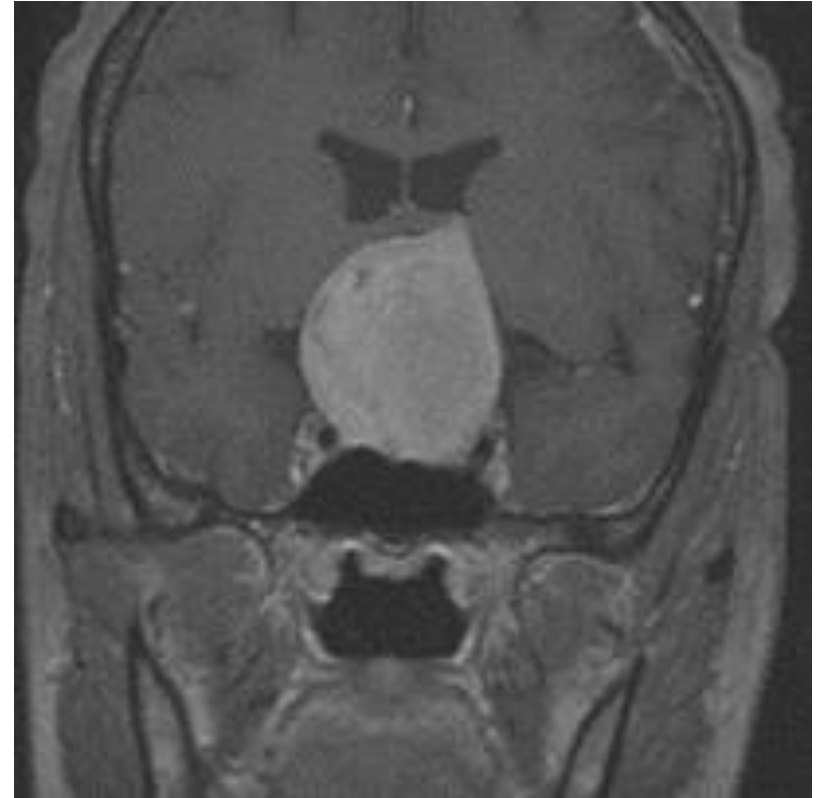
Pituitary/Suprasellar Masses

- Pituitary adenoma
- Rathke's cleft cyst
- Arachnoid cyst
- Craniopharyngioma
- Meningioma
- Glioma
- Germinoma
- Metastases
- Sarcoidosis
- Langerhan's Cell Histiocytosis
- Carotid artery aneurysm
- Lymphocytic hypophysitis
- Abscess



Non Functioning Adenomas

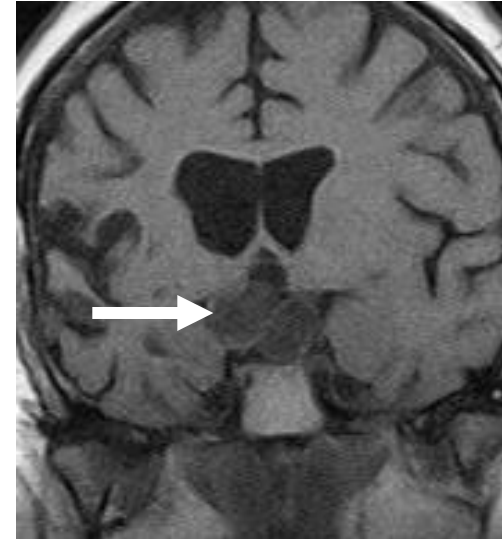
- Present most commonly with mass effect
 - visual loss, headache
- Hypopituitarism
 - Tumour causes loss of function of the normal pituitary gland
- “Incidentaloma” on scan for other reason
- “Apoplexy”
 - Sudden increase in size due to a bleed or infarct within the tumour
 - Causes severe headache





Craniopharyngioma

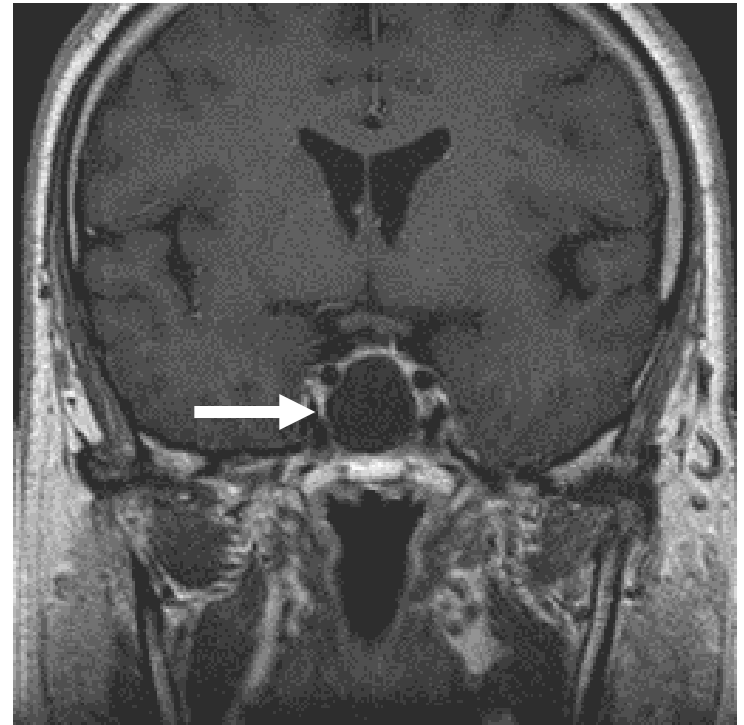
- 95% of adults have hypopituitarism
- Diabetes insipidus may occur
- Tumours often cystic and have calcification
- Tend to have worse long term outlook than pituitary adenomas
 - Increased mortality
- High recurrence rate.





Rathke's Cleft Cyst

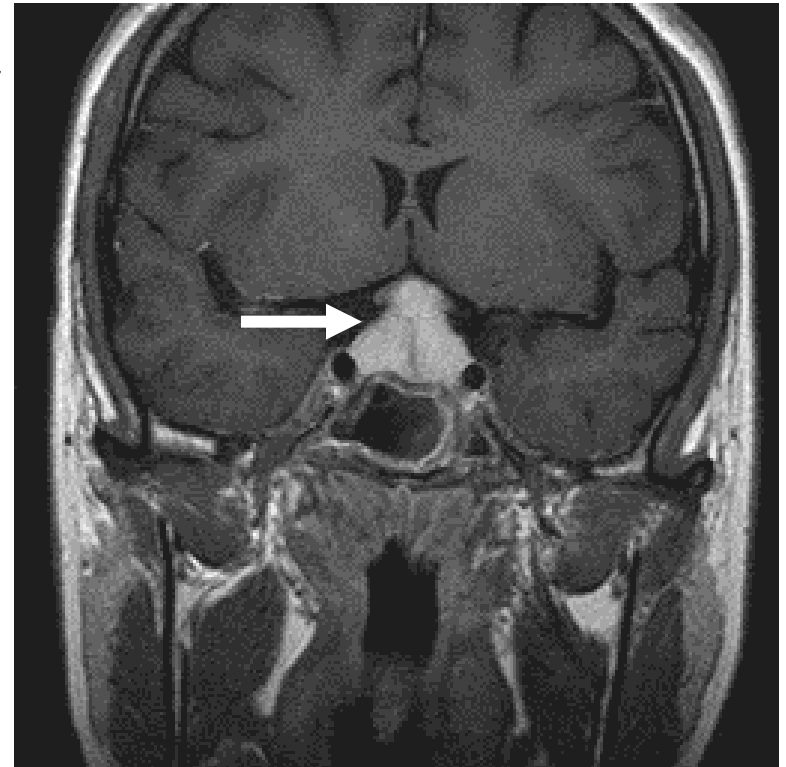
- Cysts that grow from cells originally from the roof of the mouth
- May present with visual loss and hormone deficiency
- Rarely, they can co-exist with pituitary adenomas





Lymphocytic Hypophysitis

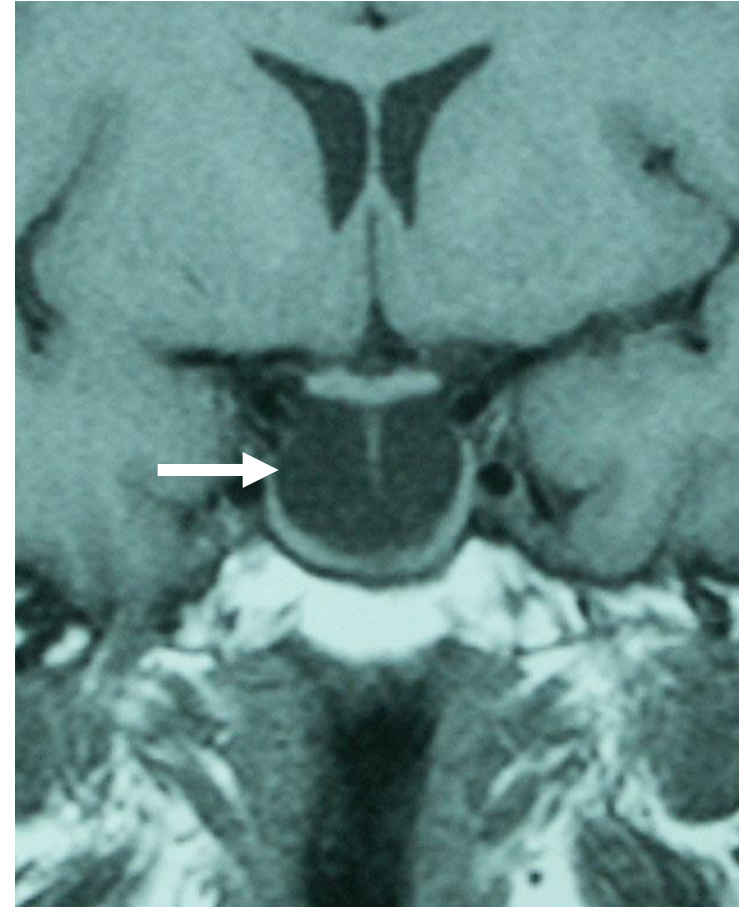
- Inflammation of the pituitary gland rather than a tumour
- Can occur in anyone but especially during pregnancy and in first 6 months post-partum
- Headache
- Hormone deficiency
 - especially cortisol deficiency (in isolation)
- Diabetes Insipidus can occur
- Prolactin high, normal or low





Empty Sella

- Usually not associated with endocrine deficit
 - But high prolactin and sometimes hormone deficiencies can occur
- Non surgical approach
- Associated with idiopathic intracranial hypertension
 - This can cause visual loss due to high pressure inside the skull
 - Tends to occur in women who are overweight





Pituitary Masses - Clinical features

Symptoms

- Headache, vision
- Loss of libido
- Erectile dysfunction
- Menstrual disturbance
- Galactorrhoea
- Fatigue, weakness
- Nausea
- Cold intolerance
- Arthralgia / Myalgia
- Postural symptoms
- Thirst / Polyuria

Signs

- Visual field examination
- Blood pressure
- Features of hormone excess
esp GH, Cushing's
- Breast examination
- Testicular size



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Too high



Prolactin

- Significant if elevated $>2x$ normal
 - Even lesser elevations may affect sensitive individuals
- Can be affected by many drugs and non-pituitary conditions
- Prolactinoma commonest secretory pituitary tumour (35-40%)



Galactorrhea is a common symptom!



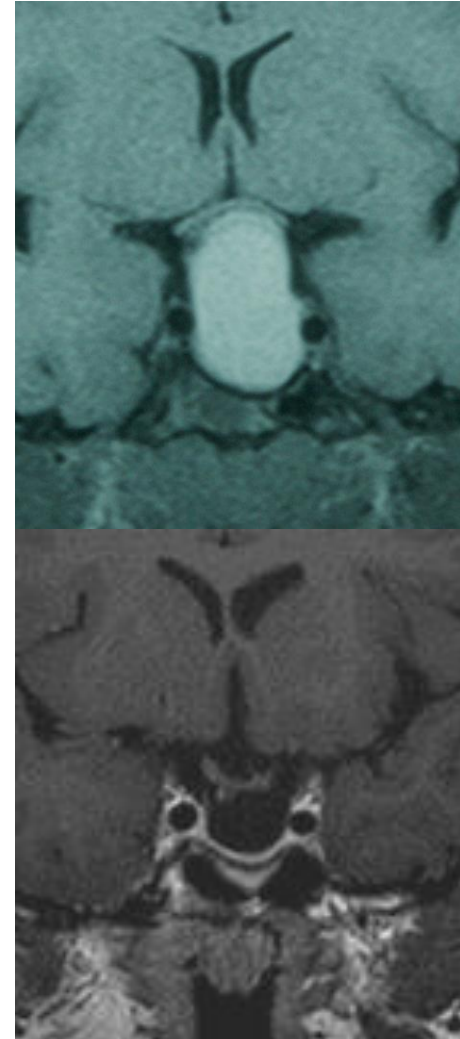
High Prolactin - Causes

- Pregnancy
- “Stress”
- Breast feeding/breast stimulation
- Chest wall lesions
- Hypothyroidism
- Kidney failure
- Cirrhosis
- Hypothalamic disease
- Non-functioning pituitary tumours
- Other sellar masses
- Mixed secretory tumours (making more than one hormone)
 - GH/PRL
 - ACTH/PRL
- Prolactinomas



Prolactinoma – Management

- Cabergoline drug of choice
- Dose 0.5 – 3.0 mg per week in 1 – 2 divided doses
- Restores normal prolactin and regular periods in 85 - 90% of women
- Tumour shrinkage in approx 80% of macroadenomas
- Successful drug withdrawal in >60% of well controlled pts after 3-5 years
- Usual to treat women until menopause, men life long





Acromegaly

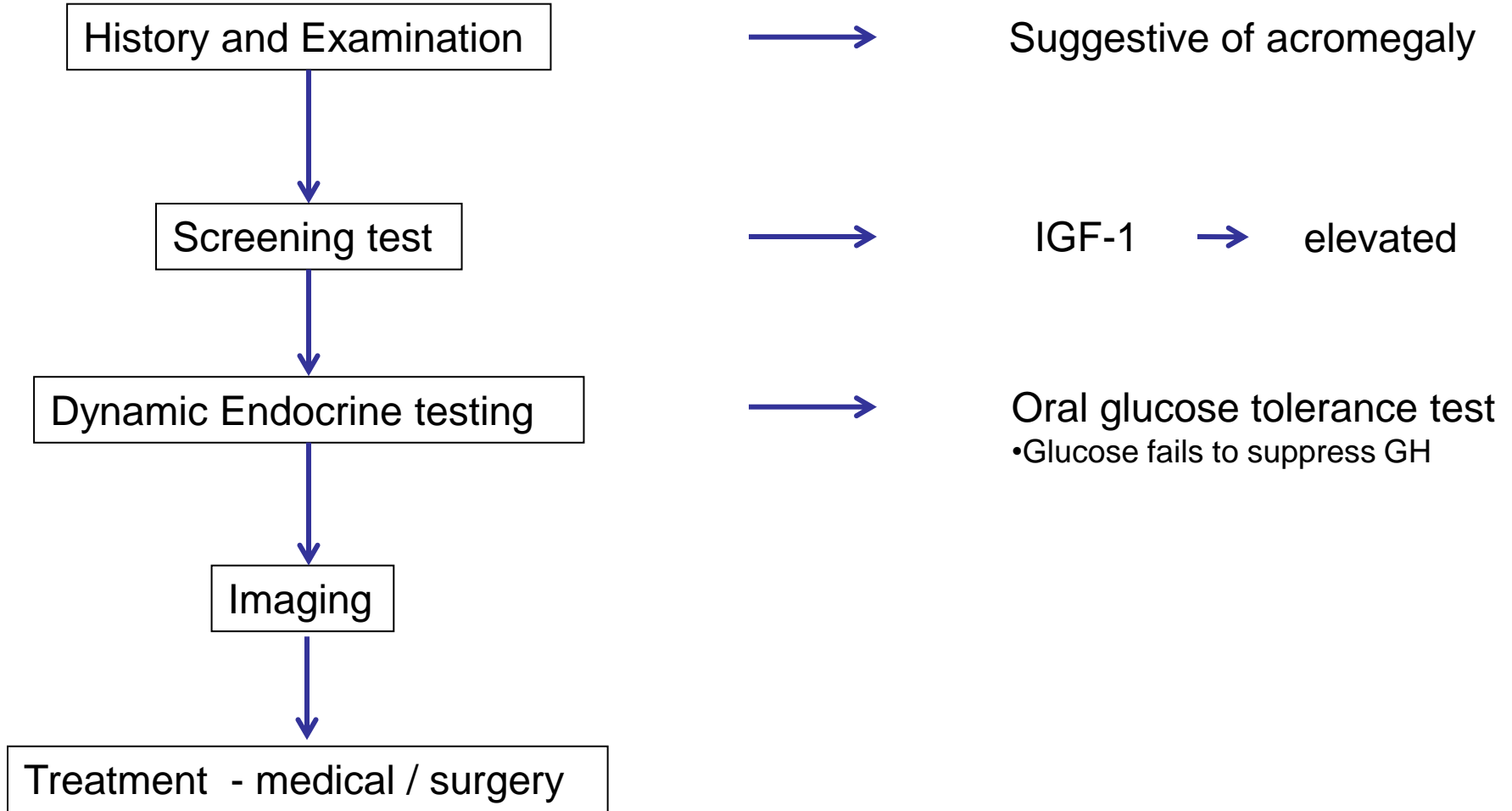
Symptoms and Signs

- Large hands and feet
- Frontal bossing
- Prognathism
- Interdental separation
- Prominent nose
- Large tongue
- Skin tags
- **Sweating**

Associated Conditions

- Arthritis
- Carpal tunnel syndrome
- Spinal stenosis
- Sleep apnoea
- Diabetes
- Goitre
- Colonic polyps

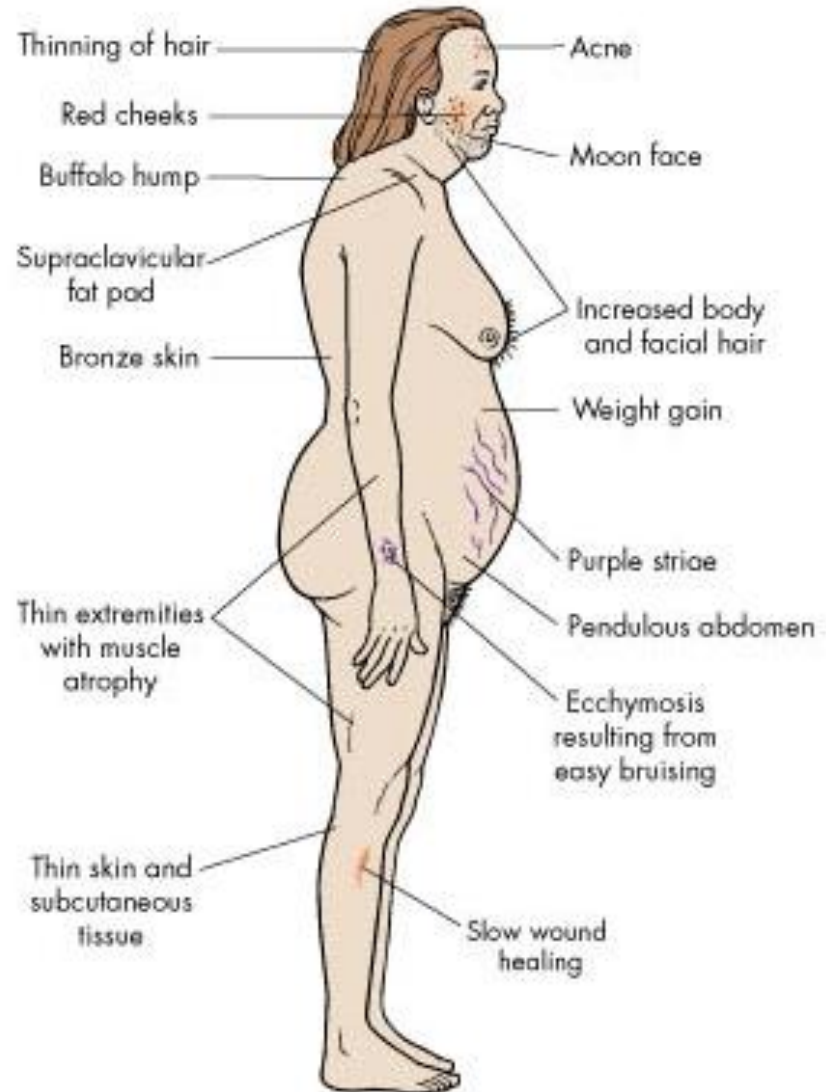


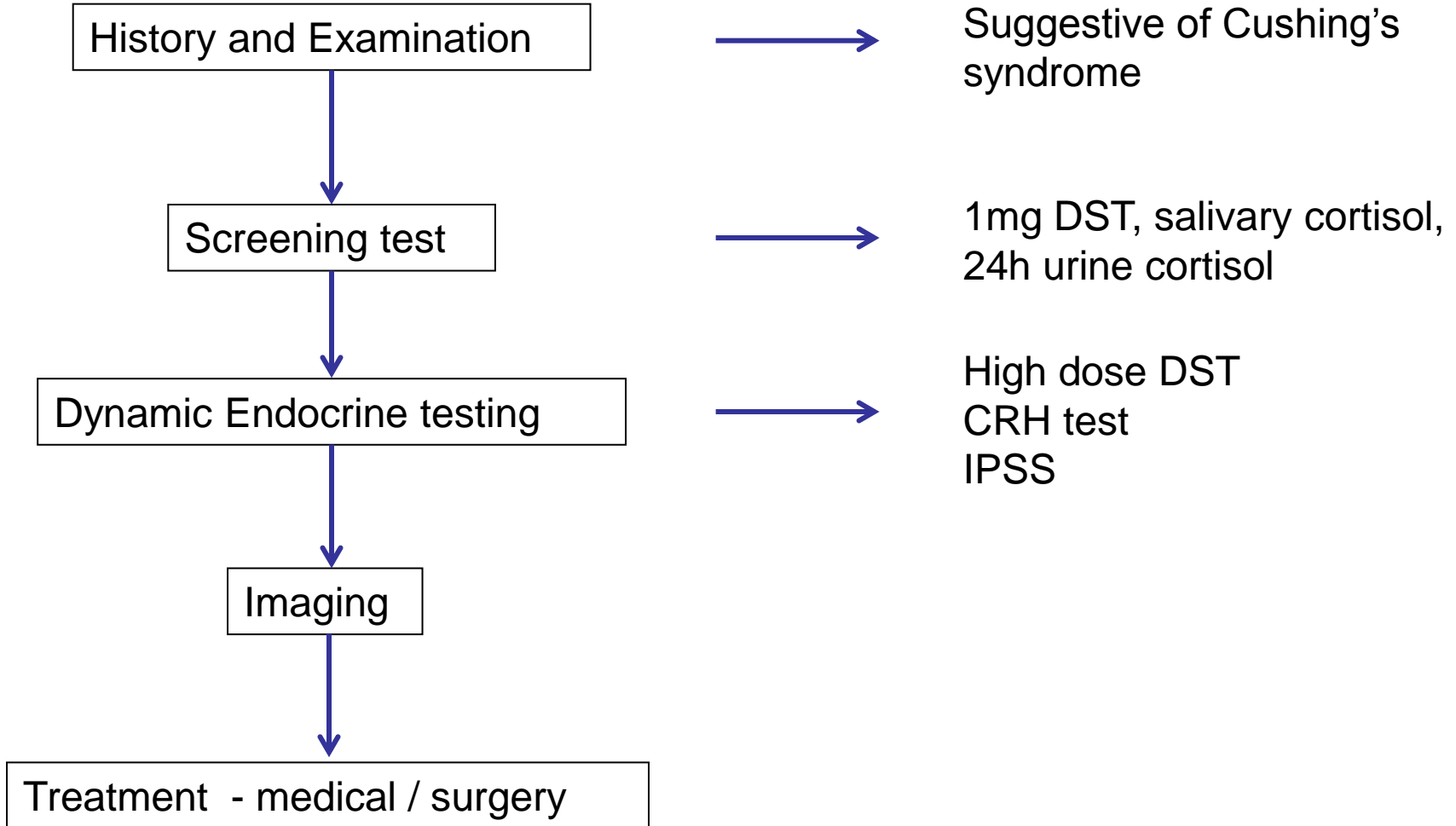




Cushing's

- Thin skin
- Spontaneous bruising
- Proximal myopathy
- >1cm purple striae
- Documented osteoporosis







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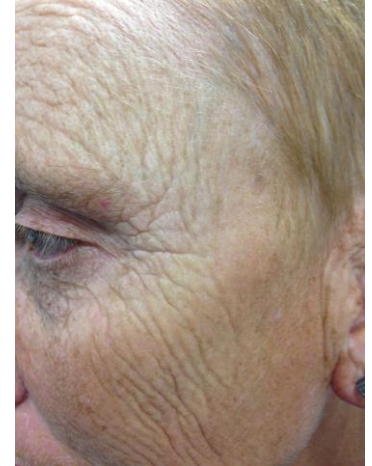
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Too low



Hypopituitarism

- Symptoms
 - Cold intolerance
 - Postural dizziness
 - Arthralgia/myalgia
 - Galactorrhoea
 - Hypogonadal symptoms
 - Especially ↓ libido
 - Headache
 - Visual loss
- Signs
 - Pale
 - Fine wrinkled appearance
 - Postural BP
 - Reduced androgen-dependent body hair
 - Visual fields





Clinical suspicion

- Suspect with new onset of reproductive dysfunction in either gender
- Some cases of “fatigue”
- Low testosterone/oestradiol with normal FSH and LH
 - Men and premenopausal aged women
- FSH and LH <10 in a post-menopausal aged woman
- Low T4 with normal TSH



Tests

- U+Es – hyponatraemia (Na <130 mmol/L)
- Morning cortisol (before 0900h)
 - <100 nmol/L – definitely abnormal
 - >350 nmol/L – probably normal
 - 100-350 nmol/L – needs further testing
- T4 and TSH
 - Low T4 with normal TSH
- Other pituitary function
 - Prolactin, FSH, LH
 - Testosterone or oestradiol
 - IGF-1



Borderline cortisol??

- Short Synacthen test
- 250 μg synthetic ACTH₁₋₂₄ (Synacthen)
 - im or iv
- Cortisol at 0, 30 and 60 minutes
- Normal >500 (550) nmol/L
- 1 μg Synacthen test (iv only)
- ITT, overnight metyrapone test, glucagon stimulation test



ITT – practical guidelines



Internal Medicine Journal 43 (2013)

HOW I TREAT

Selective use of the insulin tolerance test to diagnose hypopituitarism

S. Sarlos¹ and W. J. Inder^{1,2,3}

¹Department of Endocrinology and Diabetes, St Vincent's Hospital, Melbourne, Victoria, ²Department of Diabetes and Endocrinology, Princess Alexandra Hospital and ³School of Medicine, University of Queensland, Brisbane, Queensland, Australia

Key words

hypopituitarism, cortisol, growth hormone, insulin tolerance test.

Correspondence

Warrick J. Inder, Department of Diabetes and Endocrinology, Princess Alexandra Hospital, Woolloongabba, Qld 4102, Australia.
Email: warrick_inder@health.qld.gov.au

Received 27 May 2012; accepted 11 September 2012.

Abstract

The insulin tolerance test is considered the gold standard for assessing the hypothalamic-pituitary-adrenal and growth hormone (GH) axes, but its use varies considerably among different endocrine units. We recommend using the insulin tolerance test to assess the hypothalamic-pituitary-adrenal axis within 3 months of pituitary surgery, where adrenocorticotrophic hormone 1–24 testing is equivocal, and to assess for GH deficiency where the patient is being considered for GH replacement therapy. We also discuss safety issues, how to ensure adequate hypoglycaemia and possible alternative tests, such as the overnight metyrapone test and glucagon test.

Having established protocols and an experienced endocrine nurse has provided continuity (where registrars change annually) and prevented deskilling in the field of dynamic endocrine testing.



Diabetes insipidus

- Increased volumes of inappropriately dilute urine
- Pass 3+ litres of urine per day
- Thirst ++
 - Preference for cold water
 - Often abrupt onset
 - Normal glucose, Ca^{2+}
- Usually indicates something other than a pituitary adenoma
- Post-op following pituitary surgery





Water Deprivation Test

- Test for Diabetes insipidus
- Withhold fluid until urine is maximally concentrated or clinically dehydrated
- Measure plasma and urine osmolality before and after desmopressin

- In outpatient setting, can withhold fluids overnight and do a matched serum and urine osmolality
 - If polyuria is not too severe...



Water Deprivation Test (2)

- Urine should maximally concentrate after a serum osmolality of approx 291-3 mosmol/kg
 - Normal should be >750 mosmol/kg
- Urine osmol of <300 mosmol/kg = DI
 - Partial defects make concentrate partially
- Central DI
 - Inadequate AVP secretion
 - >50% increase in urine osmol after DDAVP
 - Partial >10% increase
- Nephrogenic DI
 - Resistant to action of AVP at level of V2 receptor at kidney
 - No increase in urine osmol after DDAVP



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Role of the Endocrine Nurse



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- Educating the registrar
 - Providing continuity
- Educating the patient
- Co-ordinating and undertaking dynamic endocrine tests
- Administering medication
 - Octreotide LAR, Lanreotide Autogel
- Research



Summary

- The pituitary is a vital little gland
- Controls functions which put joy into life
- Symptoms and signs may be hard to pick up
 - For patient, endocrine nurse and doctor!
- Pituitary disease can be treated
 - Surgery, medical therapy, radiotherapy
- The Endocrine Nurse plays a pivotal role at all steps along the diagnostic and management pathway.



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Acknowledgements



Metro South Health

- Elaine, Joce and Patrice
 - “Special Tests Sisters” Christchurch Hospital - early to mid 1990’s
- Reeta Singh, Maresa Derbyshire
 - St Vincent’s Hospital, Melbourne
- Jane Sorbello
 - Princess Alexandra Hospital, Brisbane
- Prof Eric Espiner, Prof Rick Donald