



# Programme

## Seventeenth Clinicopathological Conference on Pituitary Disease

Monday 2nd February 2015

Royal College of Obstetricians and Gynaecologists, London, NW1 4RG

Conference organiser: Dr Stephanie E. Baldeweg, UCLH/ NHNN

**A multidisciplinary approach to pituitary disease, with workshop discussions of cases by representatives from neurosurgery, endocrinology, ENT, paediatrics, radiotherapy, pathology and neuroradiology**

A number of medical companies have provided educational grants for this meeting and will have an exhibition stand present. They have had no input into the meeting agenda, organisation or selection of speakers

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**Pituitary**  
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# Faculty List



## **Dr James Ahlquist**

Consultant in Endocrinology and Diabetes  
Southend Hospital

## **Dr Stephanie Baldeweg**

Consultant Endocrinologist  
University College London Hospitals

## **Prof Pierre-Marc Bouloux**

Consultant Endocrinologist  
Royal Free Hospital, London

## **Prof Gerard Conway**

Consultant Endocrinologist  
University College London Hospitals

## **Mr Neil Dorward**

Consultant Neurosurgeon  
National Hospital for Neurology and  
Neurosurgery, London

## **Miss Catherine Gilkes**

Neurosurgery Consultant  
The Walton Centre NHS Foundation Trust,  
Liverpool

## **Dr Tony Goldstone**

Consultant Endocrinologist  
Imperial College Healthcare NHS Trust, London

## **Miss Joan Grieve**

Clinical Lead for Neurosurgery  
The National Hospital for Neurology and  
Neurosurgery, London

## **Prof Marta Korbonits**

Professor of Endocrinology and Metabolism  
Barts and the London Hospital

## **Dr Gordon Plant**

Consultant Neurologist  
National Hospital for Neurology and  
Neurosurgery, London

## **Mr Michael Powell**

Consultant Neurosurgeon  
National Hospital for Neurology and  
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## **Dr Andrew Toogood**

Consultant Endocrinologist  
University Hospitals Birmingham

## **Dr Mark Vanderpump**

Consultant Endocrinologist  
Royal Free Hospital, London

## Panel Members:

### **Endocrinologists:**

Dr Stephanie Baldeweg - London  
Dr Paul Carroll - London  
Dr Tony Goldstone – London  
Prof Ashley Grossman - Oxford  
Prof Marta Korbonits - London  
Prof Karim Meeran - London  
Dr John Newell-Price - Sheffield  
Dr Federico Roncaroli - London  
Dr Francesca Swords - Norwich  
Dr Andrew Toogood - Birmingham

### **Neurologists:**

Dr Gordon Plant - London

### **Neurooncologists:**

Dr Naomi Fersht - London

### **Neuroradiologists:**

Dr Katherine Miszkiel - London

### **Neurosurgeons:**

Mr Simon Cudlip - Oxford  
Ms Catherine Gilkes - Liverpool  
Mr Ramesh Nair - London  
Mr Saurabh Sinha - Sheffield  
Mr Nicholas Thomas - London

## Organising Committee:

Dr SE Baldeweg, Consultant Endocrinologist  
Miss Joan Grieve, Consultant Neurosurgeon  
Prof GS Conway, Consultant Endocrinologist  
Dr J Ahlquist, Consultant Endocrinologist

Dr M Vanderpump, Consultant Endocrinologist  
Prof PM Bouloux, Consultant Endocrinologist  
Mr Neil Dorward, Consultant Neurosurgeon  
Mr M Powell, Consultant Neurosurgeon

# Agenda

**08:30** **Registration**

**09:25** **Welcome and Introduction**  
Dr Stephanie Baldeweg, Consultant Endocrinologist  
University College London Hospitals

**09:30** **Key note lecture:**  
**The Pituitary in 2014**  
Dr Andrew Toogood, Consultant Endocrinologist  
University Hospitals Birmingham

**10:00** **Forum 1 – Case Presentations – Hypophysitis**  
Chairs: Dr Mark Vanderpump and Mr Michael Powell

**Case 1 – Hypophysitis and Aseptic Meningitis, an unusual combination: A Case report**  
Authors: Myuri Moorthy, Anukul Garg, Chidambaram Nethaji, North Middlesex Hospital

**Case 2 – 3-for-1: Pituitary apoplexy, hypophysitis and an adenoma – but which one is it?**  
Authors: K McCullough, N Hill, R Nair, N Mendoza, B Jones, A Mehta, A Pambakian, E Hatfield1, K Meeran, N Martin.  
Charing Cross Hospital, Imperial College Healthcare NHS Trust

**Case 3 – A case of recurrent lymphocytic hypophysitis**  
Authors: A Dimakopoulou, K Vithian,  
Colchester University Hospital

**Case 4 – Case report: Challenges of lymphocytic hypophysitis**  
Authors: Y. Ling, S. Seechurn, S. Zac-Varghese, M. Kostoula, B. Jones, A. Mehta, S. Robinson  
St Mary's hospital, Imperial College healthcare NHS Trust  
Charing Cross Hospital, Imperial College Healthcare NHS Trust

**11:00** **Coffee and Posters**

**11:30** **The pituitary gland and the eye**  
Dr Gordon Plant, Consultant Neurologist  
National Hospital for Neurology and Neurosurgery

**12:00** **Forum 2 – Case Presentations – Pituitary disease in pregnancy**  
Chairs: Dr James Ahlquist and Miss Joan Grieve

**Case 5 – Use of Medical Therapy in a Pregnant Patient with Acromegaly**  
Authors: P Jacob, NB Hashim, WM Drake

**Case 6 – Pituitary enlargement in pregnancy presenting with visual field defects and interesting pituitary imaging**

Authors: Hsiao L Yap, Edouard Mills, Maura Moriarty, Ana Pokrajac, Thomas Galliford  
Watford Hospital

**Case 7 – Pituitary apoplexy in early pregnancy resulting in partial loss of the pituitary function**

Authors: M Kostoula, S Robinson, A Lodhia, C Yu

**Case 8 – Interpretation of dynamic test results in secondary hypocortisolism post pituitary surgery and hydrocortisone replacement doses**

Authors: Dr Aikaterini Theodoraki, Mr Christopher McAlpine, Dr Anne Dawnay, Miss Joan Grieve, Mr Neil Dorward, Mr Michael Powell, Dr Stephanie Baldeweg  
University College London Hospitals, National Hospital for Neurology and Neurosurgery

**13:00 Lunch and Posters**

**14:00 The Pituitary Foundation**

**14:10 Forum 3 – Case Presentations – Pituitary**

Chairs: Prof Gerard Conway and Mr Neil Dorward

**Case 9 – Radiological Landmarks in Endoscopic Trans-sphenoidal Hypophysectomy**

Authors: Simon Carr, Vicky Twigg, Saurabh Sinha, Showkat Mirza  
Sheffield Teaching Hospital NHS Foundation Trust

**Case 10 – Not all Pituitary Tumours are Adenomas: A Rare Case of a Pituicytoma**

Authors: Archana Dhere, ST6 Diabetes & Endocrinology, Olaf Ansorge, Consultant Neuropathologist, Simon Cudlip, Consultant Neurosurgeon, Ashley Grossman, John Radcliffe Hospital, Oxford University Hospitals NHS Trust, Oxford

**Case 11 – A case of pituitary functional recovery in a patient with Langerhan's Cell Histiocytosis following chemotherapy with chlorodeoxyadenosine and Mercaptopurine.**

Authors: Andrew Ghabbour, Roselle Herring, David Russell-Jones  
Royal Surrey County Hospital NHS Foundation Trust

**Case 12 – Hypothalamic dysfunction associated with the novel anti-cancer agent programmed death ligand-1**

Authors: N.Tshuma, S.Dhalen, K.Constatinou, K.Gungunah, S.Jacobsberg, W.M. Drake, J . Evanson T.Powells  
Barts Hospital, London

**15:10 Pituitary papers that changed or will change my practice (not my CV)**

- Prof Marta Korbonits – Barts and the London Hospital
- Miss Catherine Gilkes – The Walton Centre, Liverpool

<b>15:40</b>	<b>Tea, Coffee and Posters</b>
<b>16:00</b>	<b>Key note lecture:</b> <b>Pituitary function after traumatic brain injury</b> Dr Tony Goldstone, Consultant Endocrinologist, Imperial College Healthcare NHS Trust
<b>16:30</b>	<b>Forum 4 – Case Presentations – Acromegaly</b> Chairs: Prof Pierre-Marc Bouloux and Mr Michael Powell  <b>Case 13 – An interesting case of Pituitary Adenoma with Neuronal Choristoma (PANCH); an uncommon pituitary tumour</b> Authors: S Hameed, N Mendoza, B Jones, A Mehta, B Field, K Meeran, N Martin, E Hatfield, F Roncaroli. Charing Cross Hospital, Imperial College Healthcare NHS Trust East Surrey Hospital, Surrey and Sussex Healthcare NHS Trust  <b>Case 14 – 17 years an acromegalic...</b> Authors: Olympia Koulouri, Andrew Powlson, Andrea Steuwe, Dan Gillett, Sarah Heard, Andrew Hoole, Neil Donnelly, Nagui Antoun, HK Cheow, Richard Mannion, Mark Gurnell, University of Cambridge & Addenbrooke's Hospital  <b>Case 15 – Complications of Long-Standing Uncontrolled Acromegaly and its Management</b> Authors: Ellenbogen JR, Daousi C, Gilkes CE.
<b>17:15</b>	<b>Poster and presentation prizes</b> Dr Stephanie Baldeweg, Consultant Endocrinologist University College London Hospital
<b>17:30</b>	<b>Close</b>

We would be most grateful if you could please complete your evaluation form and hand it in to a member of the CFS Events team as you leave

Thank you.

## Poster Presentations

No:	Title	Authors
<b>Poster 1</b>	An unexpected case of pituitary malignancy	S N Ali <sup>1</sup> , N E Hill <sup>1</sup> , K. Meeran <sup>1</sup> , B. Jones <sup>2</sup> , A. Mehta <sup>2</sup> , F. Roncaroli <sup>3</sup> , N. Mendoza <sup>4</sup> , M Williams <sup>5</sup> , N Martin <sup>1</sup> , E Hatfield <sup>1</sup> . 1Imperial Centre for Endocrinology, 2Department of Neuroradiology, 3Department of Neurohistopathology, 4Department of Neurosurgery. Charing Cross Hospital, Imperial College Healthcare NHS Trust, London.
<b>Poster 2</b>	'Silent' corticotroph in a macroadenoma – when to operate?	Tarig Babiker, Antonia Brooke
<b>Poster 3</b>	The Aggressive Clinical Course Of Silent Corticotroph Pituitary Adenomas – A case series	M Beebejaun, E Chinnasamy, P Rich, L Bridges, A Martin and G Bano St Georges Hospital NHS trust, Tooting, London
<b>Poster 4</b>	Non- Pituitary sellar masses – meningioma case series	E Chinnasamy, M. Beebejaun, P Rich, P Patel, A Martin and G Bano St Georges Hospital NHS trust, Tooting, London
<b>Poster 5</b>	A difficult case of Nelson's syndrome	Dr K Gunganah Professor J.P.Monson Dr N.Plowman Dr O.Koulouri Dr M.Gurnell Professor W.M.Drake
<b>Poster 6</b>	Resisting 'Resistance' in the Management of Prolactinomas	AS Powlson <sup>1</sup> , O Koulouri <sup>1</sup> , A Steuwe <sup>2</sup> , D Gillett <sup>2</sup> , S Heard <sup>2</sup> , A Hoole <sup>3</sup> , M Scott <sup>1</sup> , BG Challis <sup>1</sup> N Antoun <sup>4</sup> , RJ Mannion <sup>5</sup> & M Gurnell <sup>1</sup>  1Metabolic Research Laboratories, Institute of Metabolic Science, & Departments of 2Nuclear Medicine,, 3Medical Physics, 4Radiology & 5Neurosurgery, University of Cambridge & Addenbrooke's Hospital, Cambridge, CB2 0QQ, UK

<b>Poster 7</b>	A case of recurrent Rathke's cleft cyst: a treatment dilemma	R Ramli (1), A Mehta (4), B Jones (4), N Mendoza (2), F Roncaroli (3), K Meeran (1), E Hatfield (1), N Martin (1) 1 Imperial Centre for Endocrinology, 2 Department of Neurosurgery, 3 Department of Neurohistopathology, 4 Department of Neuroradiology Charing Cross Hospital, Imperial College Healthcare NHS Trust, London.
<b>Poster 8</b>	Bilateral adrenalectomy versus TSS for Cushing's disease	T Vakilgilani <sup>1</sup> , R Kadiyala <sup>1</sup> , N Mendoza <sup>2</sup> , R Nair <sup>2</sup> , B Jones <sup>3</sup> , A Mehta <sup>3</sup> , F Roncaroli, E Hatfield <sup>1</sup> , K Meeran <sup>1</sup> , N Martin <sup>1</sup> , F Wernig <sup>1</sup> 1 Imperial Centre for Endocrinology; 2 Dept of Neurosurgery; 3 Dept of Neuroradiology; Charing Cross Hospital, Imperial College Healthcare NHS Trust, Fulham Palace Road, London W6 8RF
<b>Poster 9</b>	Transient diabetes insipidus in pituitary apoplexy- complication in a late bloomer	Parvathy Valsalakumari, Jayashekara Acharya, Sriranganath Akavarapu, Jeannie F Todd Endocrinology, Hammersmith hospital, Imperial College Healthcare NHS Trust
<b>Poster 10</b>	Cabergoline – resistant hyperprolactinaemia: should we treat the asymptomatic male?	Lisa Yang, Daniel Darko

## Oral Presentation Abstracts

### **Case 1**

**Title: Hypophysitis and Aseptic Meningitis, an unusual combination: A Case report**

**Author(s): Myuri Moorthy, Aunkul Garg, Chidambaram Nethaji**  
**North Middlesex Hospital**

We are presenting an unusual case of Hypophysitis with aseptic meningitis.

A 49 year old female known to have Turners syndrome and primary hypothyroidism was found unresponsive at her home, 1 week after root canal treatment. Paramedics found her with profound hypoglycaemia and a low GCS. Her GCS remained low despite correction of the hypoglycaemia. Initial bloods showed sodium of 122mmol/L.

The patient was treated for a presumed meningo-encephalitis with IV ceftriaxone and acyclovir. She was also commenced on IV steroids for cortisol insufficiency because her admission cortisol was just 276nmol/L. A subsequent Lumbar Puncture showed a lymphocytosis: Gram staining, AFB, viral and fungal screen were all negative.

Unexpectedly, the patient's sodium rose to 159mmol/L despite adequate fluid replacement. Subsequent biochemical testing revealed plasma osmolality of 324mOsm/kg and urine osmolality of 419mOsm/kg. A water deprivation test proves Partial Diabetes Insipidus.

Following on from this, she had a Pituitary MRI which demonstrated a bulky pituitary gland with a normal pituitary stalk. A low baseline ACTH and a suboptimal response from a short synthacten test confirmed secondary adrenal insufficiency. The rest of her pituitary function was normal except for suppressed Gonadotrophins despite coming off HRT four weeks prior.

The patient's mental state spontaneously improved but she has ongoing problems with expressive dysphasia. This has been attributed to damage secondary to prolonged hypoglycaemia and is improving with rehabilitation.

When considering the diagnosis, we investigated and ruled out other inflammatory and infective causes such as Tuberculosis, Sarcoidosis, Fungal infections and abscesses. A pituitary biopsy was not performed because the patient recovered well and the test was felt to be unnecessary.

We highlight a difficult case of hypopituitarism and aseptic meningitis. In the absence of another unifying diagnosis we postulate an association between hypophysitis and aseptic meningitis.

## Case 2

**Title: 3-for-1: Pituitary apoplexy, hypophysitis and an adenoma – but which one is it?**

**Author(s): K McCullough<sup>1</sup>, N Hill<sup>1</sup>, R Nair<sup>2</sup>, N Mendoza<sup>2</sup>, B Jones<sup>3</sup>, A Mehta<sup>3</sup>, A Pambakian<sup>4</sup>, E Hatfield<sup>1</sup>, K Meeran<sup>1</sup>, N Martin<sup>1</sup>.**

**<sup>1</sup>Imperial Centre for Endocrinology; <sup>2</sup>Dept of Neurosurgery; <sup>3</sup>Dept of Neuroradiology; <sup>4</sup>Dept. of Neurology; Charing Cross Hospital, Imperial College Healthcare NHS Trust, Fulham Palace Road, London W6 8RF**

A 36 year old Bulgarian lady presented to her GP in the UK in April 2014 with a two month history of headache and reduced visual acuity. Pending investigations, she attended her local hospital in Bulgaria with sudden onset left sided headache, excessive thirst, double vision and subsequent loss of vision in her right eye. A MRI brain was reported as consistent with pituitary apoplexy. In addition, an intrasellar 12 mm pituitary mass was described with compression of the left optic chiasm. She underwent emergency transphenoidal surgery. A histology report described an acidophilic hypophyseal adenoma with diffuse acinar growth. Post-operatively, she developed diabetes insipidus requiring desmopressin and was informed that she needed thyroxine replacement, although the details of any post-operative pituitary function testing are unknown.

In August 2014, she travelled to Turkey to obtain a second opinion due to unresolved visual impairment. A pituitary MRI revealed persistence of the pituitary mass with minor post-operative changes. On return to the UK a month later, she requested referral to the Endocrinology team at our centre with thirst, polyuria and ongoing visual disturbances. Anterior pituitary function tests showed adequate replacement of thyroxine with no evidence of secondary hypoadrenalinism. A water deprivation test confirmed cranial diabetes insipidus. Review of pre-operative imaging at our Pituitary MDT meeting demonstrated an enlarged pituitary gland with a cystic central component which was clear of the optic tracts. However, the left optic nerve appeared bright and swollen consistent with an inflammatory process such as hypophysitis. A repeat MRI (5 months after her initial presentation) revealed a thickened infundibulum and a 12 mm peripherally enhancing lesion with mild chiasmatic elevation, representing either an inflammatory or granulomatous process. The Pituitary MDT decision was to proceed conservatively, with a further MRI three months later to establish whether surgical intervention was needed. Neuro-ophthalmology review confirmed a mild left visual field defect in a clover leaf pattern. An optical coherence tomography (OCT) scan was normal with no evidence of chronic optic nerve compression.

Our questions to the expert panel are:

Does the panel agree with our conservative approach or would they recommend a biopsy now to secure a diagnosis?

Are there any other imaging modalities or investigations that the panel would recommend to help confirm the diagnosis?

## Case 3

**Title: A case of recurrent lymphocytic hypophysitis**

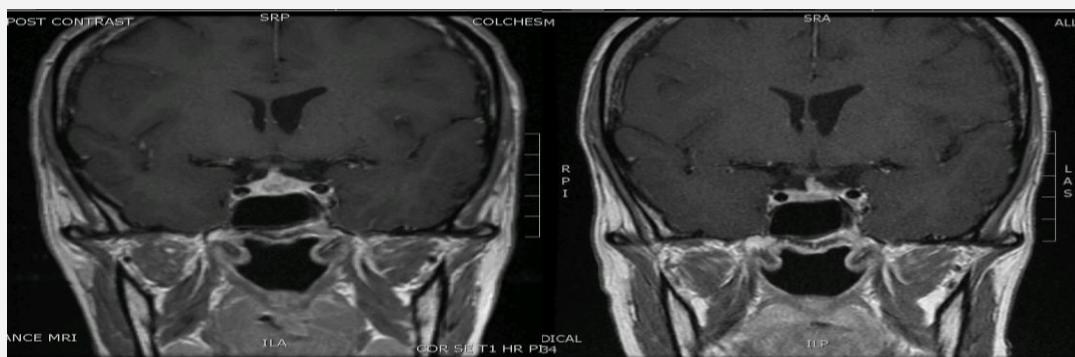
**Author(s): A Dimakopoulou, K Vithian**  
**Colchester University Hospital**

A 50 year old previously fit, District Nurse presented with a four month history of polyuria and polydipsia. She had to get up 3-4 times at night to pass urine; her 24 hour urine output was measured and was found to be 10L. Her alcohol intake was non-significant and she was a non-smoker. Examination was unremarkable with visual fields full to confrontation.

Baseline investigations revealed hypernatraemia (serum Na 152 mmol/L), with high Plasma osmolality (311 mOsmol/kg) and Urine osmolality of 152 mOsmol/kg. Endocrine testing demonstrated hyperprolactinaemia (3247 mu/L) and 9am cortisol was satisfactory at 208 mu/L. She was euthyroid with normal gonadotrophins.

MRI pituitary showed a poorly enhancing mass of 1.4 cm with some deviation of the pituitary stalk and some suprasellar extension. There was no optic chiasm involvement. MRI findings were suggestive of lymphocytic hypophysitis.

DDAVP and Cabergoline therapy was associated with clinical and biochemical improvement. Repeat serum Na in three months, was 141 mmol/L. A tapering course of Dexamethasone over four months was administered in addition to above therapy. One year after initial presentation, prolactin decreased to 12 mu/L and 9 am cortisol was 409 mu/L. An MRI scan which was performed at that time, showed good response to corticosteroid treatment with shrinkage of the pituitary gland to near normal proportions.



Eight months after completion of corticosteroid treatment, our patient presented to the Emergency Department with symptoms suggestive of mass regrowth; she complained of headache and an urgent CT head ruled out apoplexy. Dexamethasone was restarted and led to rapid relief of her symptoms.

Question: What is the experience of other centers with regard to duration of corticosteroid therapy in cases of lymphocytic hypophysitis, follow up intervals for monitoring of symptoms and MRI findings and finally, indications for surgery.

## Case 4

### Title: Case report: Challenges of lymphocytic hypophysitis

**Author(s): Y. Ling<sup>1</sup>, S. Seechurn<sup>1</sup>, S. Zac-Varghese<sup>1</sup>, M. Kostoula<sup>1</sup>, B. Jones<sup>2</sup>, A. Mehta<sup>2</sup>, S. Robinson<sup>1</sup>**

**<sup>1</sup>Imperial Centre for Endocrinology, St Mary's hospital, Imperial College healthcare NHS Trust**

**<sup>2</sup> Department of Neuroradiology, Charing Cross Hospital, Imperial College Healthcare NHS Trust**

We report a 33-year old lady (BMI 27) who presented with a 5 day history of generalised headache exacerbated by coughing. She was 7 months post-natal (P 1 +0) with an uncomplicated spontaneous vaginal delivery. On examination, she had bilateral papilloedema with normal visual fields. Neurological examination was otherwise unremarkable. Her CT head revealed a 17 x 20 mm pituitary macroadenoma which was confirmed on an MRI pituitary scan. The lesion was elevating the optic chiasm but had normal neural signal. Formal visual perimetry was normal.

Her pituitary profile was: free T4 7.8pmol/l; TSH 0.92 mU/l; prolactin 186 mU/l; Growth hormone 0.84 mcg/l; IGF-1 31.1nmol/l; FSH 1.1 IU/l; LH 0.8 IU/l; oestradiol 168 pmol/l. Short synacthen test showed 0 min cortisol 100 nmol/l; 30 min cortisol 383 nmol/l; 60 min cortisol 408 nmol/l. CSF opening pressure was raised at 27.2 cm H<sub>2</sub>O with normal analysis. Clinically, she had intracranial hypertension and an enlarged pituitary which was thought to be either a non-functioning macroadenoma or lymphocytic hypophysitis. She was commenced on acetazolamide and replacement hydrocortisone and thyroxine.

Further discussion at the pituitary MDT suggested that this was consistent with lymphocytic hypophysitis given the avidly enhancing lesion and dural involvement. We therefore treated her with pulsed methylprednisolone (1g) over a 6 week period. Sequential MRI following treatment showed a reduction and resolution by 4 months post-treatment.

This case highlights the challenges of managing lymphocytic hypophysitis which is a rare autoimmune condition. In this case, there was also evidence of intracranial hypertension which could be related to an inflammatory process affecting the optic discs or a chance association resulting from idiopathic intracranial hypertension. The treatment with pulsed methylprednisolone has been reported in literature with varying extent of success and here we report the resolution of pituitary lesion on MRI temporally associated with steroid use.

## Case 5

**Title: Use of Medical Therapy in a Pregnant Patient with Acromegaly**

**Author(s): P Jacob, NB Hashim, WM Drake**

We describe the case of a female acromegalic patient exposed to both pegvisomant and octreotide during pregnancy. Originally referred aged 31 with hirsutism and headaches, she was clinically acromegalic and had nadir serum growth hormone of 35 mU/L during standard oral glucose tolerance testing in association with serum IGF-I 733ng/ml (upper end of age-adjusted normal range 358). Anterior pituitary function was otherwise normal. She underwent uncomplicated trans-sphenoidal debulking of a large, locally invasive pituitary macroadenoma, followed by external beam radiotherapy. After suboptimal response with maximum dose long-acting somatostatin analogue therapy, biochemical control was achieved with pegvisomant 45mg twice weekly (serum IGF-1 112ng/ml). Disappointingly, her headaches persisted, necessitating subcutaneous octreotide therapy 100mcg 2-3 times daily.

Years later, she expressed a desire to conceive. Plans were made for stereotactic radiosurgery to the residual tumour tissue. The hope was to subsequently withdraw pegvisomant prior to pregnancy in the absence of its safety data. She proceeded with assisted conception at another centre whilst briefly absent from endocrine follow-up. Pegvisomant was discontinued immediately upon re-attendance at our centre at 20 weeks gestation. After counselling she elected to continue with subcutaneous octreotide therapy on account of disabling headaches.

The pregnancy was uneventful with no disturbance of glucose homeostasis. A healthy female infant weighing 3.2kg, was born with no complication. Aged 4 months, she was healthy with appropriate developmental milestones.

To our knowledge, this is the second described case of the use of pegvisomant in pregnancy (albeit up to 20 weeks gestation), and the first with dual use of octreotide and pegvisomant. Good maternal and fetal outcomes echo the observations made by Brian et al, J Clin Endocrinol Metab. 2007 Sep;92(9):3374-7.

There are numerous case reports of uncomplicated octreotide use in pregnant patients. We observed normal glucose tolerance throughout pregnancy and no side effect of octreotide therapy.

## Case 6

**Title: Pituitary enlargement in pregnancy presenting with visual field defects and interesting pituitary imaging**

**Author(s): Hsiu L Yap, Edouard Mills, Maura Moriarty, Ana Pokrajac, Thomas Galliford**  
**Watford Hospital**

We report a rare case of a patient presenting with headaches and almost complete bilateral hemianopia during her third trimester of pregnancy. This lady presented with headaches and vomiting 2 years prior, with a pituitary MRI initially reporting an acute pituitary haemorrhage. This time, due to progression of her visual field defects, it was decided in a multidisciplinary meeting to perform a Caesarean section at 37 weeks and start Cabergoline post-operatively, as the patient did not plan to breast-feed. She had a raised prolactin in keeping with pregnancy, but the rest of her pituitary profile was normal.

Upon reviewing her initial pituitary MRI and subsequent imaging at a later date, it was noted that the patient had a bulky pituitary gland, but the signal characteristics on MRI remained unchanged, which brought the original diagnosis of pituitary haemorrhage into question. It was therefore felt that this was either a proteinaceous developmental cyst or persisting blood products on serial imaging. This pituitary abnormality in combination with physiological pituitary gland enlargement in pregnancy is likely to have caused chiasmal compromise and visual field defects in this patient.

Since her delivery, she has had repeat MRI imaging, which showed regression in the size of her pituitary gland with no compression of the optic chiasm. She was gradually weaned off the cabergoline. There is no evidence of any endocrine dysfunction. However, the patient still reports some persisting visual defects, which are unaccounted for in view of her most recent pituitary imaging. She has been referred on for neuro-ophthalmic assessment.

This case also demonstrates the importance of multi-specialty management in a complex patient to achieve a satisfactory outcome. The patient has been strongly advised to avoid future pregnancies, as it is likely that she will get chiasmal compression once again.

## Case 7

**Title: Pituitary apoplexy in early pregnancy resulting in partial loss of the pituitary function**

**Author(s): M Kostoula, S Robinson, A Lodhia, C Yu**

Pituitary apoplexy is characterized by a sudden neurologic impairment and hormonal dysfunction due to acute haemorrhage or infarction of the pituitary gland, classically occurring peripartum with an existing pituitary adenoma. The pituitary enlarges in pregnancy with lactotroph hyperplasia. We present a case of pituitary infarction early in pregnancy.

A 39-year-old female with a history of hypothyroidism had hyperprolactinaemia with a 10 mm adenoma diagnosed during subfertility investigation and was treated with cabergoline for three months. She conceived with IVF and Cabergoline was discontinued. Her pregnancy was initially uncomplicated. At fourteen weeks of gestation, during a long haul flight, she developed severe temporo-frontal headache, associated with vomiting and photophobia. The symptoms persisted for a week, at which point the patient sought medical attention. She was given antibiotics, for a presumed otitis. With clinical deterioration she was admitted when pituitary MRI demonstrated haemorrhage. The patient was commenced on Hydrocortisone immediately. Symptoms and non-dynamic investigations suggested Diabetes Insipidus, and Desmopressin was introduced. The Levothyroxine dose remained unchanged. The patient remained on hormone replacement treatment throughout her pregnancy. Late second trimester imaging demonstrated resolution of the haemorrhage with a stable pituitary adenoma. She had a safe delivery by elective Caesarean section at 39 weeks of gestation as there was a fibroid uterus. She was able to breastfeed.

Pituitary apoplexy is a rare event in pregnancy, very rarely reported in early pregnancy. Management should be aimed at general stabilisation, protection of the peri-pituitary structures and hormone replacement. A multidisciplinary approach can minimise morbidity and mortality. Immediate cover with steroid replacement therapy is crucial.

## Case 8

### **Title: Interpretation of dynamic test results in secondary hypocortisolism post pituitary surgery and hydrocortisone replacement doses**

**Author(s): Dr Aikaterini Theodoraki, Mr Christopher McAlpine, Dr Anne Dawnay, Miss Joan Grieve, Mr Neil Dorward, Mr Michael Powell, Dr Stephanie Baldeweg**

**University College London Hospitals, National Hospital for Neurology and Neurosurgery**

**Background:** Following pituitary surgery, dynamic testing with insulin tolerance (ITT) or glucagon stimulation testing (GST) is used in order to identify patients with secondary hypocortisolism who require glucocorticoids. High glucocorticoid replacement doses are associated with adverse effects, and the optimal replacement dose is unknown.

**Aim:** We aimed to assess the interpretation of dynamic testing post pituitary surgery in clinical practice and its relation with the hydrocortisone replacement doses used.

**Methods:** Retrospective review of all dynamic testing results post pituitary surgery between 2000-2014 in a tertiary centre, and of the medical notes.

**Results:** 101 patients fulfilled the inclusion criteria, out of which 61 (60.4%) were tested with an ITT and 40 (39.6%) with a GST. 27 patients were prescribed glucocorticoids following ITT testing, and 26 were prescribed glucocorticoids following GST. The decision regarding the hydrocortisone dose was made by the trainee or Consultant who had requested the test from clinic. Patients that were prescribed hydrocortisone 20mg/day had significantly lower peak cortisol levels on dynamic testing compared to patients prescribed 10mg/day ( $252 \pm 142\text{mmol/L}$  vs  $356 \pm 95\text{mmol/L}$ ,  $p<0.05$ ) or emergency steroids only ( $484 \pm 45\text{mmol/L}$ ,  $p<0.005$ ), but no different peak cortisol levels compared to patients prescribed 15mg/day ( $313 \pm 149\text{mmol/L}$ ). Similarly, patients prescribed 10mg/day or 15mg/day had significantly lower dynamic testing peak cortisol compared to patients prescribed emergency hydrocortisone only ( $p<0.005$  and  $<0.05$  respectively). Administration of radiotherapy did not seem to influence the hydrocortisone dose used. Most patients continued the steroid dose recommended post dynamic testing and did not require dose changes in stable conditions with a mean follow-up of  $4.4 \pm 3$  years. On the day of dynamic testing, patients that failed an ITT had baseline cortisol of  $190 \pm 110\text{mmol/L}$ , and patients that failed GST had baseline cortisol of  $220 \pm 111\text{mmol/L}$ .

**Conclusion:** Patients diagnosed with hypocortisolism on dynamic testing post-pituitary surgery require variable dose or emergency only glucocorticoid replacement. The peak cortisol level on dynamic testing is useful when deciding the glucocorticoid replacement scheme

## Case 9

### **Title: Radiological Landmarks in Endoscopic Trans-sphenoidal Hypophysectomy**

**Author(s): Simon Carr, Vicky Twigg, Saurabh Sinha, Showkat Mirza**  
**Sheffield Teaching Hospitals NHS Foundation Trust**

#### **Introduction**

In order to perform transnasal endoscopic pituitary surgery safely and efficiently, it is important to identify anatomical and pituitary disease features on the preoperative CT and MRI scans; thereby minimising the risk to surrounding structures.

#### **Methods**

We retrospectively reviewed pre-operative CT and MRI scans of 100 adults undergoing transnasal endoscopic pituitary surgery.

#### **Results**

Scan findings and their incidence include deviated nasal septum (62%), concha bullosa (32%), bony dehiscence of carotid arteries (18%), sphenoid septation overlying the internal carotid artery (24% at the sella), low lying CSF (32%) and identifiable pituitary gland (45%). The distance of the sphenoid ostium to the skull base was a mean 10mm (range 2.7-17.6mm). We also describe the 'teddy bear' sign which when present on an axial CT indicates the carotid arteries will be identifiable intraoperatively.

#### **Conclusions**

There are significant variations in the anatomical and pituitary disease features between patients. We describe a number of features on preoperative scans and have devised a checklist including a new 'teddy bear' sign to aid the surgeon in the anatomical assessment of patients undergoing pituitary surgery.

## Case 10

### Title: Not all Pituitary Tumours are Adenomas: A Rare Case of a Pituicytoma

**Author(s): Archana Dhore, ST6 Diabetes & Endocrinology, Olaf Ansorge, Consultant Neuropathologist, Simon Cudlip, Consultant Neurosurgeon, Ashley Grossman, Professor of Endocrinology**  
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#### Abstract:

A 44 year old man presented to the Emergency Department with a severe sudden headache, photophobia and intermittent diplopia. He had sustained a minor head injury three days ago following a mechanical fall. An MRI head revealed a pituitary lesion which was referred for urgent discussion at the multidisciplinary pituitary clinic at the Oxford Centre for Diabetes, Endocrinology and Metabolism: he had a dense right temporal hemianopia. The hormonal profile showed low testosterone, low LH and FSH, but otherwise normal pituitary functions. The testosterone was replaced. The review of MRI head images confirmed a mass lesion above the pituitary gland arising from the stalk and compressing the optic chiasm. The appearance initially suggested a wide differential diagnosis including an 'ectopic' pituitary tumour, a pituitary stalk tumour or a craniopharyngioma. An urgent transphenoidal partial pituitary excision and biopsy was planned and successfully performed: the histopathology showed a benign neoplasm of TTF-1 positive cells representing specialised glia and designated as a *spindle cell oncocytoma or pituicytoma* (WHO grade 1). Tumour cell nuclei were strongly TTF-1 positive and the cells expressed annexin-1 in the cytoplasm, showed patchy GFAP positivity (5-10%) and faint focal EMA expression (<1%), with a low Ki-67 (1-2%). The visual fields improved post operatively and the patient has recovered well without complications.

#### Conclusion:

We present a rare case of a pituicytoma: less than 50 cases have been described in the literature. These are highly vascular tumours which preclude complete resection and have tendency to recur, and thus our patient remains under regular follow-up. In 2007, pituicytoma was named as a distinct entity according to the World Health Organization (WHO) classification of central nervous system tumours.

#### References:

Brat DJ, Scheithauer BW, Staugaitis SM, Holtzman RN, Morgello S, Burger PC. Pituicytoma: A distinctive low-grade glioma of the neurohypophysis. Am J Surg Pathol 2000;24:362-8.

## Case 11

**Title: A case of pituitary functional recovery in a patient with Langerhan's Cell Histiocytosis following chemotherapy with chlorodeoxyadenosine and Mercaptopurine.**

**Author(s): Andrew Ghabbour, Roselle Herring, David Russell-Jones**

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### Introduction:

Langerhans Cell Histiocytosis (LCH) is a rare multisystem disease. Pituitary manifestations include failure of the anterior and/or posterior pituitary, with diabetes insipidus and gonadotrophine deficiency being most prevalent. We present a case of a female who had pituitary failure yet conceived naturally following chemotherapy.

### Case description

A 24 year old female presented with polydipsia, polyuria and lethargy. A water deprivation test was diagnostic of cranial diabetes insipidus. MRI showed thickening of the infundibulum and an abnormal pituitary.

She initially responded well to DDAVP but following a miscarriage developed progressive anterior pituitary functional failure requiring thyroxine, hydrocortisone and GH replacement.

She had secondary amenorrhoea and did not tolerate HRT or OCP.

At age of 36 she developed new and increasing breathlessness, which prompted further investigation. HRCT confirmed progressive diffuse pulmonary shadowing and histology following VATS biopsy was consistent with Langerhan's Cell Histiocytosis presumed to be involving the pituitary gland and the lungs.

Due to increasing respiratory dysfunction she was being considered for lung transplantation and the decision was made to treat her with 4 cycles of chlorodeoxyadenosine (cladribine or 2-CDA) and this was followed with 1 year of 6 Mercaptopurine. Prior to chemotherapy she was referred for ovum harvest and possible IVF.

Following chemotherapy, her periods returned but in an irregular manner. One year later, she had a LHRH test with LH rose from 3.8 to 23 and FSH from 2.9 to 6.8. She is now 42 years old and has just found out that she is 26 weeks pregnant, something that she never thought possible.

### Discussion

The case demonstrates functional pituitary recovery post cladribine and 6 mercaptopurine in a patient with Langerhan's Cell Histiocytosis.

### References

- Modan-Moses et al; Hypopituitarism in Langerhans Cell Histiocytosis: seven cases and literature review. *J Endocrinol Invest*, 2001;24(8):612-7.
- Kaltsas GA et al; Hypothalamo-pituitary abnormalities in adult patients with Langerhans Cell Histiocytosis: clinical, endocrinological, and radiological features and response to treatment. *J Clin Endocrinol Metab*. 2000;85(4): 1370-6.

## Case 12

**Title: Hypothalamic dysfunction associated with the novel anti-cancer agent programmed death ligand-1**

**Author(s): N.Tshuma, S.Dhalen, K.Constatinou, K.Gungunah , S.Jacobsberg, W.M. Drake , J . Evanson T.Powels**

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**Background:** The immunomodulatory drug Ipilimumab, used for the treatment of melanoma, is known to cause hypophysitis with hypopituitarism. We report the development of a destructive inflammatory hypothalamic mass in association with programmed death ligand-1 (PDL-1) for the treatment of refractory metastatic bladder carcinoma.

**Case description:** A 74 year old female, previously well, was diagnosed with a T3b grade 3 bladder carcinoma. She received gemcitabine and cisplatin chemotherapy, followed by a radical cystectomy. Routine surveillance revealed multiple lung lesions (shown to be metastases on biopsy) and she was enrolled into a phase II clinical trial of a novel immunomodulatory drug, PDL-1. She received 12 cycles over 8 months with a single urinary tract infection, treated with trimethoprim, being the only complication. Cross-sectional imaging showed complete resolution of the pulmonary disease. A month later, she presented with confusion, reduced mobility and recurrent falls. On examination hypothermia was noted. A septic screen was negative. Cranial imaging demonstrated an unusual enhancing hypothalamic mass atypical for a metastasis treated, under oncological supervision, with dexamethasone with clinical improvement. Follow-up cranial imaging demonstrated reduced size and enhancement of the hypothalamic lesion. Some weeks later, she presented to hospital with adipsia associated with hypernatraemia (150 mmol/L) and a disordered sleep-wake cycle and referred for endocrine evaluation. She was clinically and biochemically hypothyroid and hypothermic. A diagnosis of hypothalamic dysfunction and hypopituitarism was made and a regimen of hormone replacement, fixed fluid intake, rewarming and supportive care initiated. The patient has improved clinically sufficiently to be discharged and continues rehabilitation at home. To our knowledge, this is the first case of hypothalamic inflammation and dysfunction secondary to PDL-1 and highlights the growing issue of novel cancer therapies causing, on occasion, significant endocrine dysfunction.

## Case 13

### **Title: An interesting case of Pituitary Adenoma with Neuronal Choristoma (PANCH); an uncommon pituitary tumour**

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A 67 year old woman presented with features of acromegaly. She had recently undergone a hemicolectomy for Dukes C colon carcinoma. The previous year, biopsy of a mediastinal mass had shown a benign spindle cell lesion. She was also under investigation for hypercalcaemia (2.64mmol/L) with elevated PTH (9.8pmol/L). Her son had recently developed bilateral carpal tunnel syndrome. On examination she had a visual field defect. Biochemistry confirmed the diagnosis (IGF-1nmol/L 110.4nmol/L (reference range 6.0-36.0nmol/L), random growth hormone (GH) 20.5 $\mu$ g/L). In view of her co-existent primary hyperparathyroidism, sequence analysis of the *MEN1* gene was performed, but this did not identify a mutation. DNA is also being analysed for *AIP* mutations. MRI demonstrated a large, partly cystic tumour compressing the optic chiasm and extending into the right cavernous sinus. She underwent transsphenoidal pituitary surgery. The postoperative MRI showed residual tissue in the right cavernous sinus. Biochemistry confirmed a reduction of the GH burden. Histology showed a somatotroph adenoma (Ki67 2%, no overexpression of p53). The adenoma cells were positive for synaptophysin and GH and neuronal cells and axons were intensely positive for neurofilament proteins and synaptophysin. The transcription factor Pit-1 was positive in both adenoma cells and neurones. These findings are consistent with a diagnosis of pituitary adenoma with neuronal choristoma (PANCH). PANCH is an uncommon tumour that consists of an adenomatous component almost always producing GH admixed with well differentiated neurones. The neuronal component is thought to derive from adenoma cells as a result of divergent differentiation. This hypothesis is supported in our case by the expression of Pit-1 in neuronal cells.

Our questions to the expert panel are:

Is there a unifying diagnosis that would bring together the PANCH, bowel carcinoma, mediastinal mass and raised calcium and PTH?

What further genetic analysis would be useful on this patient?

## Case 14

**Title: 17 years an acromegalic . . .**

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A 65 year-old man had a 17 year history of acromegaly requiring multimodal therapy. He presented in 1998 with a pituitary macroadenoma, IGF-1 2.4 xULN and GH nadir during OGTT of 11 mcg/L. Despite transsphenoidal surgery (TSS) on two occasions (1998, 1999), his acromegaly remained active (IGF-1 1.76 xULN, OGTT GH nadir 4.5 mcg/L), and he proceeded to radiotherapy (RT: 45 Gy, 25 #), which was completed in 2000. This resulted in complete anterior hypopituitarism. However, his acromegaly remained active, requiring continuation of somatostatin analogue (SSA) and dopamine agonist (cabergoline) therapy. Serial GH and IGF-1 levels over time were either just within, or marginally above, accepted targets. Pituitary MRI suggested a possible small right-sided remnant, but with the caveat of not being able to reliably differentiate from post-operative/post-RT scar tissue/change.

In 2013, the patient expressed a wish to discontinue depot SSA therapy and to explore other (preferably non-injection-based) treatment options. After detailed discussion, he requested further transsphenoidal (endoscopic) exploration.

*We would like to invite the panel to comment on whether they would have supported such an approach or suggested an alternative strategy.*

To provide added reassurance that the possible right sided remnant was indeed 'active' residual adenoma, and to screen for other possible sites of residual tumour, we proceeded to <sup>11</sup>C-methionine PET-CT coregistered with volumetric MRI. This revealed a single focus of 'activity' in the right side of the sella, with no other abnormal tracer uptake.

Based on these findings, the patient underwent a third TSS focusing on this area and residual GH-staining adenoma was resected. Post-operatively, the patient's acromegaly has been (for the first time in 17 years) in full remission off all medical therapy (IGF-1 0.74 xULN and GH nadir 0.47 mcg/L on OGTT – 8 weeks post-operatively and >12 weeks after last SSA injection).

## Case 15

**Title: Complications of Long-Standing Uncontrolled Acromegaly and its Management**

**Author(s): Ellenbogen JR, Daousi C, Gilkes CE.**

We present a case of aggressive acromegaly with a radiation induced brainstem cavernoma.

A 56 year old male was diagnosed with acromegaly in 1996 whilst working in Holland (GH 22.5 ug/l, IGF-1 186.3) secondary to a large, irregular pituitary macroadenoma, invading the right cavernous sinus. He underwent a transphenoidal hypophysectomy followed by radiotherapy. In 1997 he returned to the UK with active acromegaly despite optimum medical therapy, including bromocriptine and octreotide therapy. A small persisting nodule remained above the optic apparatus. The patient refused craniotomy to resect this, and stereotactic radiosurgery was deemed too high risk to vision. Up to 2011 he refused any further intervention but he developed sequelae of his disease including; joint pains, mood swings, dyslipideamia, renal calculi, colonic polyps, non-insulin dependent diabetes mellitus and benign prostatic hypertrophy. In 2011 he underwent a further endoscopic transphenoidal exploration of the intrasellar area. Histology confirmed growth hormone expressing adenoma. There was no post operative improvement in the IGF level. Pegvisomont funding was applied for, but refused.

In January 2012 he was presented with an acute internuclear ophthalmoplegia. MRI demonstrated a pontine cavernous haemangioma within the radiation field of his previous treatment. This was initially treated conservatively but he re-presented in March 2012 with a progressive headache, ophthalmoplegia and further visual deterioration. Imaging demonstrated progressive hydrocephalus secondary to haemorrhage from the cavernoma. An endoscopic third ventriculostomy was performed followed by a posterior fossa craniotomy and removal of the pontine cavernoma.

Currently his acromegaly remains active with an IGF1 of 31 (7-25).

Points for discussion:

Is the cavernoma radiation induced?

Should the patient move house?

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