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# Programme

## Eighteenth Clinicopathological Conference on Pituitary Disease

Thursday 25th February 2016

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



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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 Rene Bernays**

Consultant Neurosurgeon  
Neurosurgery Klinik Hirslanden, Zurich

**Prof Pierre-Marc Bouloux**

Consultant Endocrinologist  
Royal Free Hospital, London

**Dr Paul Carroll**

Consultant Endocrinologist  
Guy's & St Thomas' NHS Trust, London

**Mr Simon Cudlip**

Consultant Neurosurgeon and Spinal Surgeon  
John Radcliffe Hospital, Oxford

**Prof Mehul Dattani**

Consultant Paediatric Endocrinologist  
University College London Hospitals

**Mr Neil Dorward**

Consultant Neurosurgeon  
National Hospital for Neurology and Neurosurgery, London

**Dr Jane Evanson**

Consultant Neuroradiologist  
Barts and the London Hospitals NHS Trust

**Miss Joan Grieve**

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

**Mr Jonathan Pollock**

Consultant Neurosurgeon  
Queens Hospital, Romford

**Mr Michael Powell**

Consultant Neurosurgeon  
National Hospital for Neurology and Neurosurgery, London

**Dr Lisa Sze Rogdo**

Consultant Endocrinologist  
Hormon Zentrum Zürich

**Dr Mark Vanderpump**

Consultant Endocrinologist  
Royal Free Hospital, London

## Panel Members:

**Endocrinologists:**

Dr Paul Carroll - London  
Prof Ashley Grossman - Oxford  
Prof Marta Korbonits - London  
Prof Karim Meeran - London  
Dr Francesca Swords - Norwich

**Neuropathologist:**

Dr Federico Roncaroli - London

**Neurologists:**

Dr Gordon Plant - London

**Neurooncologists:**

Dr Naomi Fersht - London

**Neuroradiologists:**

Dr Katherine Miszkiet - London  
Dr Jane Evanson - London

**Neurosurgeons:**

Mr Simon Cudlip - Oxford  
Ms Katie Gilkes - Liverpool  
Mr Saurabh Sinha - Sheffield  
Mr John Pollock - Romford

## Organising Committee:

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

Dr M Vanderpump, Consultant Endocrinologist  
Prof PM Bouloux, Consultant Endocrinologist  
Mr N 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 2015**  
Mr Jonathan Pollock, Consultant Neurosurgeon, Queens Hospital, Romford
- 10:00**      **Forum 1 – Case Presentations – Pituitary all sorts**  
Chairs: Mr Michael Powell & Dr Stephanie Baldeweg
- Case 1 – ACTH dependent Cushing’s Syndrome – an unexpected source**  
Authors: Dhere A, Trott MJ, Farah G, Wang LM, Grossman A, Oxford Centre for Diabetes, Endocrinology & Metabolism
- Case 2 – Cushing’s disease in pregnancy: Treatment dilemmas**  
Authors: Karan Jolly, Wiebke Arlt, Niki Karavitaki, Shahzada Ahmed, University Hospital Birmingham
- Case 3 – An unusual Pituitary adenoma with doors cross checked and set to automatic**  
Authors: Zosanglura Bawlchhim, Isuri Kurera, Roselle Herring, David Russell-Jones, Royal Surrey County Hospital NHS Foundation Trust, Federico Roncaroli, Hammersmith Hospital
- Case 4 – A giant invasive prolactinoma**  
Authors: Muquit S, Daousi C, Gilkes CE, University Hospital Aintree, Liverpool
- 11:00**      **Tea, Coffee and Posters**
- 11:30**      **Inflammatory pituitary conditions**  
Dr Paul Carroll, Consultant Endocrinologist, Guy’s & St Thomas’ NHS Trust
- 11:45**      **Forum 2 – Case Presentations – Inflammatory pituitary conditions**  
Chairs: Miss Joan Grieve & Prof Pierre-Marc Bouloux
- Case 5 – Living with Lymphocytic Hypophysitis. A 10 year history**  
Authors: Ms Anouk Borg, University College London Hospital  
Mr. Jonathan Benjamin, Queen’s Hospital Romford & Dr James Ahlquist, Southend Hospital
- Case 6 – Hypophysitis: A single-centre case series**  
Authors: E Chinnasamy, P Rich, A Martin, L Bridges, L Seal and G Bano  
St Georges University Hospitals NHS Foundation Trust
- Case 7 – Recurrent autoimmune hypophysitis - a systemic disease?**  
Authors: S Hussain, K McCullough, N Mendoza, A Mehta, B Jones, A Pambakian, K Meeran, N Martin, F Wernig, Imperial College Healthcare NHS Trust  
F Roncaroli, University of Manchester
- 12:30**      **Imaging of the pituitary**  
Dr Jane Evanson, Consultant Neuroradiologist, Barts and the London Hospitals NHS Trust

- 12:50 Lunch and Posters**
- 13:50 The Pituitary Foundation**
- 14:00 Acromegaly and Klotho**  
Dr Lisa Sze Rogdo, Consultant Endocrinologist, Hormon Zentrum Zürich
- 14:25 Forum 3 – Case Presentations – Pituitary mix and match**  
Chairs: Mr Neil Dorward & Dr Mark Vanderpump
- Case 8 – Dual pituitary pathology in acromegaly – identifying the culprit lesion**  
Authors: A Powlson, O Koulouri, D Gillett, S Heard, N Antoun, HK Cheow, R Mannion, M Gurnell, Addenbrookes Hospital, Cambridge
- Case 9 – Changing pituitary structure associated with severe hyponatremia**  
Authors: Isuri Kurera, Zosangira Bawlchhim, Roselle Herring, David Russell-Jones, Royal Surrey County Hospital NHS Foundation Trust
- Case 10 – Temozolomide for Aggressive Pituitary Tumors**  
Authors: F. Solda', J Grieve, M. Powell, N Dorward, S.E. Baldeweg, N. Fersht, University College London Hospitals, J. Harney, City Hospital, Belfast, P. Bouloux, Royal Free London NHS Foundation Trust
- 15:10 Medical key note: Management of Congenital Hypopituitarism**  
Prof Mehul Dattani, Consultant Paediatric Endocrinologist, University College London Hospitals
- 15:40 Tea, Coffee and Posters**
- 16:00 Forum 4 – Case Presentations – Surgical Conundrums**  
Chairs: Miss Joan Grieve & Dr James Ahlquist
- Case 11 – Diagnostic challenge in a case of rapidly enlarging pituitary gland**  
Authors: R Mallik, A Rathore, J Pollock, U Pohl & J Ahlquist  
Southend Hospital, Westcliff on Sea & Queen's Hospital, Romford
- Case 12 – Cerebrospinal fluid rhinorrhea in a large multicystic invasive macroprolactinoma**  
Authors: A Dimakopoulou, D Choi, S Baldeweg, University College London Hospitals
- 16:30 Debate: The endoscope means the iMRI is obsolete in pituitary surgery**  
**For:** Mr Simon Cudlip, Consultant Neurosurgeon and Spinal Surgeon, John Radcliffe Hospital, Oxford  
**Against:** Prof Rene Bernays, Consultant Neurosurgeon, Neurosurgery Klinik Hirslanden, Zurich
- 17:15 Poster and presentation prizes**  
Dr Stephanie Baldeweg, Consultant Endocrinologist  
University College London Hospitals  
Miss Joan Grieve, Consultant Neurosurgeon, University College London Hospitals
- 17:30 Close**

## Oral Abstract Presentations

<b>Oral 1</b>	ACTH dependent Cushing's Syndrome – an unexpected source	Dhere A, Trott MJ, Farah G, Wang LM, Grossman A
<b>Oral 2</b>	Cushing's Disease in pregnancy: treatment dilemmas	Karan Jolly <sup>1</sup> , Wiebke Arlt <sup>2,3</sup> , Niki Karavitaki <sup>2,3</sup> , Shahzada Ahmed <sup>1</sup> <sup>1</sup> Ear, Nose and Throat Department, Queen Elizabeth Hospital Birmingham, University Hospitals Birmingham NHS Foundation Trust; <sup>2</sup> Institute of Metabolism and Systems Research, School of Medical and Dental Sciences, University of Birmingham; <sup>3</sup> Department of Endocrinology, Queen Elizabeth Hospital Birmingham, University Hospitals Birmingham NHS Foundation Trust
<b>Oral 3</b>	An Unusual Pituitary Adenoma with Doors cross checked and set to automatic	Zosanglura Bawlchhim <sup>1</sup> , Isuri Kurera <sup>1</sup> , Roselle Herring <sup>1</sup> , Federico Roncaroli <sup>2</sup> , David Russell-Jones <sup>1</sup> <sup>1</sup> . Royal Surrey County Hospital NHS Foundation Trust <sup>2</sup> . Hammersmith Hospital
<b>Oral 4</b>	A giant invasive prolactinoma	Muquit S, Daousi C, Gilkes CE
<b>Oral 5</b>	Living with Lymphocytic Hypophysitis. A 10 year history.	Ms Anouk Borg Mr. Jonathan Benjamin Dr. James Ahlquist
<b>Oral 6</b>	Hypophysitis: A single-centre case series	E Chinnasamy, P Rich, A Martin, L Bridges, L Seal and G Bano St Georges University Hospitals NHS Foundation Trust, Tooting, London
<b>Oral 7</b>	Recurrent autoimmune hypophysitis- a systemic disease?	Author(s): S Hussain <sup>1</sup> , K McCullough <sup>1</sup> , N Mendoza <sup>2</sup> , A Mehta <sup>3</sup> , B Jones <sup>3</sup> , A Pambakian <sup>4</sup> , F Roncaroli <sup>5</sup> , K Meeran <sup>1</sup> , N Martin <sup>1</sup> , F Wernig <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, Imperial College Healthcare NHS Trust, Fulham Palace Road, London W6 8RF <sup>5</sup> Wolfson Molecular Imaging Centre, University of Manchester
<b>Oral 8</b>	Dual pituitary pathology in acromegaly – identifying the culprit lesion	A Powlson, O Koulouri, D Gillett, S Heard, N Antoun, HK Cheow, R Mannion, M Gurnell

<b>Oral 9</b>	Changing pituitary structure associated with severe hyponatremia	Isuri Kurera <sup>1</sup> , Zosanglra Bawlchhim <sup>1</sup> , Roselle Herring <sup>1</sup> , David Russell-Jones <sup>1</sup>  Royal Surrey County Hospital NHS Foundation Trust
<b>Oral 10</b>	Temozolomide for Aggressive Pituitary Tumors	F. Solda <sup>1</sup> , J Grieve <sup>2</sup> , M. Powell <sup>2</sup> , N Dorward <sup>2</sup> , J. Harney <sup>3</sup> , S.E. Baldeweg <sup>4</sup> , P. Bouloux <sup>5</sup> , N. Fersht <sup>1</sup>  1 Department of Oncology, University College London Hospitals NHS Foundation Trust, London, UK 2 Department of Neurosurgery, National Hospital for Neurology and Neurosurgery, London, UK 3 Department of Clinical Oncology, Northern Ireland Cancer Center, Belfast City Hospital, Belfast, UK 4 Department of Endocrinology, University College London Hospitals NHS Foundation Trust, London, UK 5 Department of Endocrinology, Royal Free London NHS Foundation Trust, London, UK
<b>Oral 11</b>	Diagnostic challenge in a case of rapidly enlarging pituitary gland	A Rathore, R Mallik, J Pollock, U Pohl & J Ahlquist Southend Hospital, Westcliff on Sea & Queen's Hospital, Romford
<b>Oral 12</b>	Cerebrospinal fluid rhinorrhea in a large multicystic invasive macroprolactinoma	A Dimakopoulou, D Choi, S Baldeweg

# 1.

<b>Title:</b>	<b>ACTH dependent Cushing's syndrome – an unexpected source</b>
<b>Author(s):</b>	<b>Dhere A, Trott MJ, Farah G, Wang LM, Grossman A</b>
<p>A 34 year-old Japanese lady presented with facial swelling, lethargy, muscle weakness, sleep disturbance, and cognitive blunting since 2012. She had relapsing and remitting symptoms in the Winter of 2013 and 2014. She had skin dryness, pigmentation and acne when we saw her in March 2015.</p> <p>Investigations revealed low serum potassium of 3.2 mmol/L, 24h urinary cortisol 363 nmol/L (slightly elevated), serum cortisol 1469 nmol/L (elevated), incomplete suppression on overnight dexamethasone-suppression testing at 158 nmol/L. Due to the intermittent nature of her symptoms, two years lapsed prior to correct diagnosis of hypercortisolaemia. She had a midnight serum cortisol of 331 nmol/L, failed to suppress on low-dose dexamethasone suppression testing 484, 648 and 1536 nmol/L (0, 24, 48h). ACTH persisted at 680-760 ng/L and failed to rise after hCRH. Pituitary-MRI demonstrated a right 8mm lesion. At this stage we concluded she had periodic ACTH-dependent Cushing's syndrome. Inferior petrosal sinus sampling was delayed as she went into remission. CT scans of chest, abdomen and pelvis demonstrated bulky adrenal glands and a 24mm anterior mediastinal nodule suggestive of a thymoma. The cortisol levels did not rise over several months so we decided to proceed with bilateral inferior petrosal sinus sampling, which showed no central gradient. Octreotide scanning showed normal uptake.</p> <p>In view of thymic mass and patient's concern, we decided to proceed with thymectomy. She had complete resolution of symptoms with histopathology confirming a well-differentiated grade 1 neuroendocrine tumour positive for ACTH on immunostaining. Mutation testing for MEN-1 was negative.</p> <p>Thymic tumours may secrete ACTH ectopically, but cyclical Cushing's usually suggest Cushing's disease. Thymic ACTH-producing tumours are aggressive and often associated with MEN-1. This case is notable for the absence of MEN-1, along with the female gender and history of non-smoking, and the very variable ACTH secretion which initially suggested Cushing's disease.</p>	

## 2.

<b>Title:</b>	<b>Cushing's disease in pregnancy: treatment dilemmas</b>
<b>Author(s):</b>	<b>Karan Jolly<sup>1</sup>, Wiebke Arlt<sup>2,3</sup>, Niki Karavitaki<sup>2,3</sup>, Shahzada Ahmed<sup>1</sup></b>  <b><sup>1</sup>Ear, Nose and Throat Department, Queen Elizabeth Hospital Birmingham, University Hospitals Birmingham NHS Foundation Trust; <sup>2</sup>Institute of Metabolism and Systems Research, School of Medical and Dental Sciences, University of Birmingham; <sup>3</sup>Department of Endocrinology, Queen Elizabeth Hospital Birmingham, University Hospitals Birmingham NHS Foundation Trust</b>
<p>A 30-year old female was diagnosed with Cushing's disease attributed to microadenoma in 2008 (presenting manifestations: weight gain, easy bruising, fatigue, secondary amenorrhoea for at least one year) and underwent TSA. Histopathology confirmed corticotroph adenoma but she had persistent disease (morning serum cortisol 491 nmol/l). Repeat assessments in the following months revealed normal 24hr UFC (x3) with serum cortisol at 84 nmol/l on the oDST. Given the improvement in her clinical picture and the restoration of periods, monitoring was decided. In the following years, 24hr UFC remained normal (apart from one occasion) and serum cortisol on the oDST ranged between 119-148 nmol/l. Pituitary MRI in 2011 disclosed no adenoma. Her periods had remained regular, she had no typical Cushingoid appearances and her BMI had dropped from 40 to 37 Kg/m<sup>2</sup>; however, she had developed diabetes mellitus. In July 2013, 24UFC was high (x3) and in October 2013, she was referred 13 weeks pregnant. Fetal ultrasound at 21 weeks had revealed a congenital diaphragmatic hernia (contents: stomach, bowel, liver) and following discussion at the MDT and taking into account the risks of hypercortisolism for the mother and fetus, TSA was recommended. Pituitary MRI had shown a R-sided microadenoma and endoscopic TSA took place at the 23rd week. Pathology confirmed corticotroph adenoma and post-operatively she was put on Hydrocortisone. Her diabetes was controlled with metformin and insulin and at 33 weeks she was found hypertensive and was put on labetalol and nifedipine. At 38 weeks, she had emergency CS but the baby died 36 hours later. Assessment of cortisol status a few months after delivery, suggested disease remission (9.00 am serum cortisol 71 nmo/l).</p> <p>Pregnancy in the setting of Cushing's relates with significant adverse sequelae; optimal management remains a major challenge and depends on severity, stage of gestation, risks-benefits for maternal/fetal outcomes.</p>	

### 3.

<b>Title:</b>	<b>An Unusual Pituitary Adenoma with Doors cross checked and set to automatic</b>
<b>Author(s):</b>	<b>Zosanglura Bawlichhim 1, Isuri Kurera<sup>1</sup>, Roselle Herring<sup>1</sup>, Federico Roncaroli<sup>2</sup>, David Russell-Jones<sup>1</sup></b> <b>1. Royal Surrey County Hospital NHS Foundation Trust</b> <b>2. Hammersmith Hospital</b>
<p>We have previously presented the fourth reported case of silent ACTH/GH secreting adenoma, which went on to demonstrate the unique co-location of the 2 different granules within the same cell on electron microscopy (a reported first). However, the clinical management of this case has proven very difficult and we offer it for discussion at the meeting.</p> <p>A 34 year old male who worked as cabin crew was noted to be missing people on one side of the aisle and was asked to have an eye test by his seniors which showed significant visual field defect. The development of visual symptoms coincided with lack of libido. Examination revealed Bitemporal hemianopia and no obvious syndrome of hormone excess. Pituitary function profile was normal. MRI showed a large pituitary adenoma measuring 2.53x2.77x2.77cm displacing the optic chiasm. He underwent Trans-sphenoidal hypophysectomy and immune-staining showed tumour consisting of mixed ACTH and GH secreting adenoma. Electron microscopy showed co-location of secretory granules within the same cell. Postoperative visual field showed marked improvement as did MRI. Repeat pituitary function revealed normal levels with IGF-1 of 30nmol/L(13-50). Insulin Tolerance Test showed growth hormone response above 12ug/L and a flat cortisol response. He continued on hydrocortisone replacement. Repeat MRI showed expansion of the residual pituitary tumour without chiasmal compression. He underwent a second operation followed by radiotherapy. Four years after his last operation, MRI showed significant increase in size of the residual tumour. He went on to have his third operation complicated by prolonged CSF leak. Recent MRI has shown extensive enlargement of pituitary mass extending to optic chiasm. The neurosurgeons are hesitant to operate again.</p> <p>In conclusion we present an unusual pituitary adenoma which has been very aggressive and treatment options will be open for discussion. Should he be offered further surgery, octreotide or other chemotherapy?</p>	

## 4.

<b>Title:</b>	<b>A giant invasive prolactinoma</b>
<b>Author(s):</b>	<b>Muquit S, Daousi C, Gilkes CE</b>
<p>We present the case of a patient with an invasive giant prolactinoma with significant bone erosion of anterior skull base.</p> <p>A 70-year-old man presented with a short history of headaches, nausea, dizziness and right 6th nerve palsy. Over the previous month he had noticed a mild proptosis of his right eye. He was fit and well prior to the onset of his symptoms.</p> <p>Examination findings were of proptosis of his right eye and marked right 6th nerve palsy, but no other significant findings.</p> <p>CT and MRI revealed an extensive skull base tumour involving the sphenoid sinus, pituitary fossa, clivus and left occipital condyle as well as extending over the right posterior orbit and left orbital apex. There was erosion of the bone of most of the anterior skull base.</p> <p>Prolactin levels was 1,057,000 mIU/L. Testosterone level was low at 7.0 nmol/L. The remainder of his pituitary hormone profile was normal, as was the result from short synacthen test.</p> <p>Within a few weeks after presentation his 6th nerve palsy and resulting diplopia improved. His headaches and dizziness resolved.</p> <p>Points for discussion:</p> <p>The pros and cons of immediate medical management of such an extensive, essentially asymptomatic prolactinoma. Which DA agonist should we use and how? What is the risk of CSF leak? What is the most likely site of CSF leak and how would the panel manage?</p>	

## 5.

<b>Title:</b>	<b>Living with Lymphocytic Hypophysitis. A 10 year history</b>
<b>Author(s):</b>	<b>Ms Anouk Borg Mr. Jonathan Benjamin Dr. James Ahlquist</b>
<p>We present the case of a 41-year old lady with a delayed diagnosis of lymphocytic hypophysitis (LH). She presented at the age of 29 with amenorrhea. Visual fields and endocrine evaluation were normal except for a prolactin of 1831mIU/L. MRI brain revealed an enhancing sellar and suprasellar mass with compression of the optic chiasm.</p> <p>She underwent a transphenoidal subtotal resection in 2003. A firm pituitary tumour was encountered at surgery which did not curette easily. Histology showed non-specific changes and described as necrotic amorphous tissue with no evidence of neoplasia. The pituitary profile one month after surgery was normal. Her cycles became regular after her operation but she subsequently developed amenorrhea again a year later. She was found to have biochemical changes of hypopituitarism.</p> <p>The residual tumour was initially observed with interval scans and showed a slight increase in size over time. Due to the radiological recurrence and the fact that no histological diagnosis was made from prior surgery she was offered another resection. In 2014 she underwent a right lateral supra orbital craniotomy and debulking of the suprasellar element. Histology showed prominent fibrosis surrounding atrophic non-neoplastic anterior pituitary tissue as well as prominent lymphocytic infiltrate. These findings were consistent with chronic lymphocytic hypophysitis.</p> <p>At present, more than ten years since her first presentation, she remains on hormone replacement. Her vision is normal and the pituitary tissue shows stable appearances on surveillance scans.</p> <p>Our case demonstrates that LH can be an easily missed diagnosis due to its non specific changes on histology. In the presented case with long term follow up, it is demonstrated that the clinical course of LH can be managed with hormone replacement and the role of surgery being mainly for providing a tissue diagnosis. Surveillance imaging is still recommended due to the unclear natural history of this condition.</p>	

## 6.

<b>Title:</b>	<b>Hypophysitis: a single-centre case series</b>
<b>Author(s):</b>	<b>E Chinnasamy, P Rich, A Martin, L Bridges, L Seal and G Bano</b> <b>St Georges University Hospitals NHS Foundation Trust,</b> <b>Tooting, London</b>
<p>Hypophysitis is a complex clinicopathological spectrum and it can be classified in a number of ways, including histologically, anatomically, or aetiologically. Such a heterogeneous nature of this condition, poses a diagnostic challenge. Here we present a case series of hypophysitis from varied aetiology.</p> <p>Data are currently available for 11 patients, 3 confirmed lymphocytic hypophysitis (LyHy), 2 probable LyHy, 3 likely TB hypophysitis, an idiopathic hypophysitis and 2 related to Ipilimumab. Male to Female ratio is 2:9. Mean age: 37 years, excluding Ipilimumab cases. Among the LyHY group, 1 presented in third trimester of pregnancy and it was recurring in subsequent pregnancy, 2 presented after stopping OCPs. Headache was the predominant symptom in these patients. Hormonal deficiencies were DI in 4, secondary hypoadrenalism in 7/10 and hypothyroidism in 7, secondary hypogonadism in 8/10, GH deficiency in 5/6. Common MRI finding was contrast enhancing sellar and/or suprasellar lesions. 3/4 with DI had absent posterior bright spot. IgG subclasses were measured in 3, normal in 2, one had elevated IgG1 and she also has asymptomatic Sjogren's syndrome. TB test was done in 6 patients, 3 were positive. 2/3 positive TB patients had systemic symptoms. CT chest was done for 8 and 2 had findings suggestive of TB. Serum ACE was negative in all 6 patients measured. Among non- TB patients, 3 improved with high dose steroids and one patient with less active inflammation did not show any improvement radiologically or clinically. Two patients treated with Azathioprine, as steroid sparing agent had gradual improvement.</p> <p>Primary hypophysitis is diagnosis of exclusion and our question to the panel is about standard diagnostic approach in a patient presenting with hypophysitis and what is your threshold for biopsy? Our case series and the recent publication from German Endocrine society, highlight the need for a standardised diagnostic approach for this condition.</p>	

## 7.

<b>Title:</b>	<b>Recurrent autoimmune hypophysitis- a systemic disease?</b>
<b>Author(s):</b>	<p><b>Author(s): S Hussain<sup>1</sup>, K McCullough<sup>1</sup>, N Mendoza<sup>2</sup>, A Mehta<sup>3</sup>, B Jones<sup>3</sup>, A Pambakian<sup>4</sup>, F Roncaroli<sup>5</sup>, K Meeran<sup>1</sup>, N Martin<sup>1</sup>, F Wernig<sup>1</sup></b></p> <p><b>1Imperial Centre for Endocrinology; 2Dept of Neurosurgery; 3Dept of Neuroradiology; 4Dept. of Neurology, Imperial College Healthcare NHS Trust, Fulham Palace Road, London W6 8RF 5Wolfson Molecular Imaging Centre, University of Manchester</b></p>
<p>A 36-year old Bulgarian lady presented in April 2014 with headache, sudden deterioration of visual acuity, polyuria and polydipsia. She was diagnosed with pituitary apoplexy due to an intrasellar 12mm pituitary adenoma with left optic chiasm compression and underwent emergency transphenoidal pituitary surgery in Bulgaria. Anterior pituitary function tests demonstrated low levels of gonadotropins and no evidence of secondary hypoadrenalism. She developed diabetes insipidus and was commenced on desmopressin as well as a combined oestrogen and progesterone preparation. She had a prior diagnosis of Hashimoto's thyroiditis since 2010 for which she was taking levothyroxine.</p> <p>Due to on-going visual impairment, repeat MRI imaging was performed in Turkey in August 2014. This suggested an increase in size of her pituitary mass and interval imaging was advised. She was referred to the Imperial Centre for Endocrinology in October 2014 for on-going management. Pituitary function tests confirmed earlier findings from Bulgaria. Neurophthalmic investigations demonstrated a mild left visual field defect with no evidence of chronic optic nerve compression. Re-assessment of her previous imaging and repeat imaging suggested an inflammatory process with an enlarged pituitary gland, cystic central component, thickened left optic nerve and infundibulum without chiasmal involvement. The case was discussed at the CPC meeting in 2015 where several differentials were considered. Since then, review of her histology from Bulgaria at our centre confirmed a diagnosis of autoimmune panhypophysitis with negative IgG4 immunohistochemistry.</p> <p>In May 2015 she subsequently developed persistent derangement in her liver function tests. A full hepatitis screen including autoimmune serology and serum IgG4 antibody levels were negative. In December 2015, she developed a recurrence of her severe headache with diplopia. An MRI pituitary scan revealed an increase in size of her inflammatory pituitary lesion and derangement in her liver function tests. She was commenced on high-dose prednisolone with significant improvement of her headache, diplopia and liver function tests. Given the recurrent nature of her autoimmune panhypophysitis and systemic involvement, initiation of azathioprine as a steroid-sparing treatment and a liver biopsy are being considered.</p> <p>Our questions to the expert panel are:</p> <p>Does the panel agree with our approach and choice of agent to manage this lady long-term? Are there any investigations that may help us obtain a unifying diagnosis?</p>	

## 8.

<b>Title:</b>	<b>Dual pituitary pathology in acromegaly – identifying the culprit lesion</b>
<b>Author(s):</b>	<b>A Powlson, O Koulouri, D Gillett, S Heard, N Antoun, HK Cheow, R Mannion, M Gurnell</b>

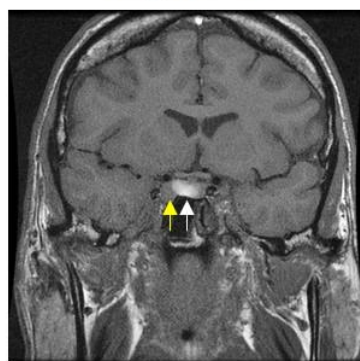
A 52-year-old man presented with classic acromegaly features. He had a history of hypertension, irritable bowel syndrome and rheumatoid arthritis, for which he was taking methotrexate.

His IGF-1 was raised [125 nmol/L (4xULN)] and an oral glucose tolerance test revealed a nadir GH of 3.3 mcg/L. His prolactin was also elevated at 595 mU/L (<360), and he had a mildly raised FT4 [20.8 pmol/L (10–19.8)] with a near fully suppressed TSH [0.03 mU/l (0.35–5.5)]. Examination demonstrated a small goitre, but no clinical features of thyrotoxicosis. He was otherwise eupituitary.

His pituitary MRI showed a hypointense lesion in the right sella extending towards the cavernous sinus (yellow arrow), and a second, apparently discrete lesion, in the centre of the gland which was of high signal intensity (white arrow), raising the suspicion of a Rathke's cleft cyst, or alternatively a cystic/ haemorrhagic pituitary adenoma (Figure 1). In order to establish whether one or both of the lesions corresponded to a 'functioning' adenoma, and represented a surgical target, the patient underwent <sup>11</sup>C-Methionine PET/CT. This revealed intense tracer uptake in the region of the right cavernous sinus (yellow arrow), but no uptake inside the cystic lesion (Figure 2).

This case illustrates the potential utility of functional pituitary imaging with <sup>11</sup>C-Methionine PET/CT in differentiating causative and coincidental pituitary lesions in patients with acromegaly and dual/multiple pituitary pathology on MRI.

**Figure 1**



**Figure 2**



## 9.

<b>Title:</b>	<b>Changing pituitary structure associated with severe hyponatremia.</b>
<b>Author(s):</b>	<b>Isuri Kurera<sup>1</sup> , Zosanglra Bawlchhim <sup>1</sup>, Roselle Herring<sup>1</sup>, David Russell-Jones<sup>1</sup></b>
<p>A 39 year old Texan oil man male presented in 2004 with severe headache, reduced level of consciousness and severe hyponatremia (sodium &lt;100mmol/L) . MRI showed an enlarged homogeneous pituitary gland. He was treated as an emergency with hydrocortisone and IV fluids. On ITU despite efforts, his sodium rose from &lt;100mmol/L to 128mmol/L over 4 days causing mild central pontine myelinosis. His pituitary profile showed random cortisol of 372nmol/L and TSH of 0.21mU/L((0.3-5 Mu/l) ) with low T4 of 5.8 pmol/L ((9-24pmol/L) ) and low T3 of 1.8 pmol/L (3.5-6.5pmol/L). He then recovered clinically and the MRI evolved with reduction of the enlarged pituitary. His adreno-cortical axis was demonstrated to have recovered. He was reviewed in the pituitary clinic and remained well until he represented again 4 years later with sudden onset headache and serum sodium of 106mmol/L. His random cortisol was 36nmol/L.. He was again managed with intravenous steroids and fluids. His pituitary profile showed TSH of 0.41mU/L with low T4 of 8.9pmol/L and low T3 of 2.9pmol/L. His FSH /LH and testosterone levels were undetectable with low prolactin of 30Miu/L(0-700Miu/L) and IGF-1 was on the lower end of normal 13.5nmol/L (13-50nmol/L). On recovery glucagon test revealed no growth hormone response. Clinically and biochemically he did not show any evidence to suggest diabetes insipidus.</p> <p>Interestingly one month after presentation, there was radiological shrinkage of the pituitary gland.</p> <p>He now remains on full pituitary replacement consisting of hydrocortisone, levothyroxine, nebido and growth hormone and there have been no further episodes. He managed to climb up to 5000m on Mount Kilimanjaro in 2012.</p> <p>MRIs continue to show progressive shrinkage of the pituitary gland. In conclusion, we present changing pituitary pathology associated with severe hyponatremia.</p> <p>The question for discussion: Q1. What is the underlying aetiology?</p>	

## 10.

<b>Title:</b>	<b>Temozolomide for Aggressive Pituitary Tumors</b>
<b>Author(s):</b>	<p><b>F. Solda<sup>1</sup>, J Grieve<sup>2</sup>, M. Powell<sup>2</sup>, N Dorward<sup>2</sup>, J. Harney<sup>3</sup>, S.E. Baldeweg<sup>4</sup>, P. Bouloux<sup>5</sup>, N. Fersht<sup>1</sup></b></p> <p><b>1 Department of Oncology, University College London Hospitals NHS Foundation Trust, London, UK</b>  <b>2 Department of Neurosurgery, National Hospital for Neurology and Neurosurgery, London, UK</b>  <b>3 Department of Clinical Oncology, Northern Ireland Cancer Center, Belfast City Hospital, Belfast, UK</b>  <b>4 Department of Endocrinology, University College London Hospitals NHS Foundation Trust, London, UK</b>  <b>5 Department of Endocrinology, Royal Free London NHS Foundation Trust, London, UK</b></p>
<p>Aggressive pituitary adenomas are challenging to manage and multiple treatment approaches are routinely offered. Owing to their invasive nature, repeated surgeries are not curative and adjuvant radiotherapy and medical therapies are unable to provide long-term control. Temozolomide (TMZ), an alkylating agent traditionally used in the treatment of brain gliomas, has shown promising results in the management of refractory aggressive pituitary adenomas. We present our experience with four patients treated with TMZ at further progression after conventional combined treatment modalities: three atypical adenomas (PRL adenoma - case 1, GH adenoma - case 2, silent ACTH adenoma - case 3) and a non-functioning pituitary carcinoma (case 4). Oral TMZ was given using the standard regimen of 150-200 mg/m<sup>2</sup>/day for 5 consecutive days every 4 weeks (mean 6 cycles, range 2-12). Treatment was well tolerated. Clinical and radiological control of disease was achieved for 36, 10 and 14 months from completion of treatment in cases 1, 3 and 4 respectively. Hormone level normalization was also observed in case 1. Tumour progressed rapidly after two cycles in case 2. At last follow up, 1 patient was alive and free from disease (case 4) and 1 patient was alive with recurrent disease treated with stereotactic radiotherapy (case 1) and he is currently being re-challenged with TMZ for further tumour progression. The other 2 patients (case 2 and 3) died from disease progression respectively at 16 and 29 months respectively after TMZ completion. The use and efficacy of TMZ in aggressive pituitary adenomas needs to be further evaluated in prospective clinical trials</p>	

## 11.

<b>Title:</b>	<b>Diagnostic challenge in a case of rapidly enlarging pituitary gland</b>
<b>Author(s):</b>	<b>A Rathore, R Mallik, J Pollock, U Pohl &amp; J Ahlquist Southend Hospital, Westcliff on Sea &amp; Queen's Hospital, Romford</b>
<p>A 33-year-old lady presented with polyuria and polydipsia for 2 weeks with headache and galactorrhoea. There was no relevant past medical history and she was not taking any regular medications. Pituitary function was normal except prolactin 1759 mU/L.</p> <p>MRI scan showed diffuse pituitary enlargement with inhomogenous uptake of gadolinium. Pituitary stalk was markedly thickened, enhanced diffusely and was touching the optic chiasm. A presumptive diagnosis of autoimmune hypophysitis was made. Diabetes insipidus was confirmed with a water deprivation test. She was treated with prednisolone 40 mg daily and desmopressin.</p> <p>Repeat blood tests a week later showed development of panhypopituitarism. A repeat MRI 2 weeks after presentation showed no shrinkage and slight enlargement of the gland. CT chest, abdomen and pelvis showed no evidence of a primary cancer, lymphoma or sarcoid. ANA, ANCA and serum ACE levels were normal.</p> <p>She represented 5 weeks after the initial presentation with acute onset of diplopia due to a right 6th nerve palsy. She had a right temporal hemianopia in the right eye and a left upper temporal quadrantanopia. MRI pituitary showed further increase in the size of the pituitary gland extending laterally into the right cavernous sinus and inferiorly also. The pituitary stalk had nearly doubled in size compared to the initial scan and was compressing the optic chiasm. She underwent urgent pituitary surgery for decompression of the optic chiasm and the cavernous sinus, and to achieve a tissue diagnosis. Histological analysis is underway: preliminary results indicate an unusual form of infiltrating malignant disease.</p> <p>This case is highly unusual in view of the rapid progression of the pituitary gland pathology and presented a diagnostic challenge. Multi-disciplinary management between endocrinologist, neurosurgeon, neuroradiologist and pathologist has been particularly important. The panel is invited to review the case and comment on diagnosis and management.</p>	

## 12.

<b>Title:</b>	<b>Cerebrospinal fluid rhinorrhea in a large multicystic invasive macroprolactinoma</b>
<b>Author(s):</b>	<b>A Dimakopoulou, D Choi, S Baldeweg</b>
<p>A 41 year old female patient was referred to Neurosurgery with nasal discharge on background of a macroprolactinoma.</p> <p>She originally presented with amenorrhoea and galactorrhoea. She was treated with bromocriptine, but changed to cabergoline due to intolerance. Prolactin levels settled from 1760 mu/L to 757 mu/L and delivered her first child by the age of 34. Her MRI head prior conception showed a large pituitary mass, extending into the sphenoid sinus with a large cystic component due to herniation of the arachnoid into the fossa. Two further successful pregnancies followed. Nasal discharge was noted whilst she was taking cabergoline. Prolactin levels were normal, at 422 mu/L. Symptoms of fluid leak were more prominent when she got up in the morning. MRI pituitary showed destruction of the pituitary fossa by a large multicystic mass protruding into the nasopharynx. There was a posterior soft tissue component which was invading into the clivus. On repeat imaging the macroprolactinoma has almost completely disappeared and the sphenoid was full of dura and cerebrospinal fluid. Skull base had to be repaired via trans-sphenoidal endoscopic approach. The procedure was uneventful and histology confirmed adenoma with co-expression of growth hormone and prolactin - Ki-67 was 3%.</p> <p>Our patient continued with cabergoline 0.5mg twice weekly after neurosurgery and her menses were regular. However, she developed a new episode of rhinorrhoea with prolactin levels of 922 mu/L. Pituitary appearances were stable on imaging but asialotransferrin was present on fluid analysis. She underwent a second transnasal endoscopic CSF leak repair with lumbar drain and fat graft. Cabergoline was stopped and most recent prolactin is 830 mu/L. Nasal discharge was re-sent for analysis; asialotransferrin was negative.</p> <p>It is reported that large invasive prolactinomas can reduce in size after treatment with dopamine agonists and CSF rhinorrhoea can occur as a result of tumour shrinkage. Case series data suggest that it is not necessary to stop dopamine agonist therapy in subjects with macroprolactinomas who develop CSF rhinorrhea. We would like to ask for the panel's expert opinion on use of Cabergoline in this context and discuss further neurosurgical management options.</p>	

## Poster Abstracts

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Poster 6	Tale of an Untreated MEN1 patient involving Pituitary & Parathyroid	Dr Yogesh Kalaiah, The Royal Oldham Hospital
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1.

**Title: Interesting case of pituitary tuberculosis**

**Author(s): H Rachabattula , A Crown, D Agranoff**

**Case History:** A 19 year old lady presented in June 2015 with left sided visual disturbance, reduced visual acuity and colour vision. She did not have any symptoms of Diabetes Insipidus. Her MRI Pituitary showed 15mm pituitary macroadenoma with uniform enhancement, extending into the suprasellar region and stretching the chiasm. Her blood results showed cortisol 462nmol/l, prolactin 831miu/l, FT4 12.4pmol/l, TSH-1.38mu/l, LH-3.2iu/l,FSH-5.9iu/l.

She had a background history of parapneumonic effusion diagnosed in Jan 2011 and her pleural fluid analysis showed reactive lymphocytes with no bacterial growth or acid fast bacilli (AFB). In April 2011, she was diagnosed with right sided psoas abscess extending into T12 extradural compartment with some cord displacement. She had her abscess drained and the pus was tested negative for AFB. She was diagnosed with tuberculosis based on clinical and radiological evidence. She was started on anti tuberculous treatment (ATT) following which she clinically improved and had completed 1 year course of ATT.

**Diagnosis and Treatment:** Her differential diagnosis for the pituitary adenoma was possible pituitary tuberculosis or non functioning pituitary adenoma. She underwent trans-sphenoidal resection of the pituitary mass. Histology of the resected tissue showed granuloma consistent with tuberculosis. She was started on ATT and prednisolone 40 mg post operatively to cover the initiation of ATT. Her Post-operative MRI Pituitary showed relatively normal looking pituitary gland. She clinically improved with all the treatment and the plan was to continue the ATT for 1 year.

**Discussion:** Pituitary tuberculosis is extremely rare. Pituitary tuberculosis could present with endocrine dysfunction or symptoms of mass effect. Imaging modalities are non- specific, can show thickening of pituitary stalk and intense post contrast enhancement. Diagnosis is therefore difficult and challenging. Trans-sphenoidal biopsy is considered essential to establish the diagnosis. These lesions tend to resolve with appropriate ATT. Our case highlights the significance of considering the tuberculosis as a differential diagnosis of suprasellar mass and emphasizes that the close collaboration between the Infection control Physicians, Neurosurgeons and Endocrinologists is vital in such cases.

## 2.

Hypopituitarism after radiotherapy for primary brain tumour: is radiotherapy the cause?

*Authors:* R Mallik, U Aziz, K Madhavan, J Ahlquist. Southend Hospital, Westcliff on Sea, Essex.

A 67-year-old lady was admitted with lethargy and drowsiness for 3 weeks, with headache, polyuria and polydipsia. She had a history of frontal lobe glioblastoma (WHO grade IV); in 2007 she had undergone excision followed by radiotherapy and concurrent temozolomide. Radiotherapy was delivered by intensity modulated radiotherapy (IMRT) in 30 fractions over 6 weeks, total dose 60 Gy. She remained well until July 2015 when she presented with tumour recurrence at the original site and underwent re-excision.

She was postmenopausal, had type 2 diabetes mellitus and also had a history of Graves' disease not currently requiring treatment. She was taking dexamethasone 4 mg twice daily and metformin 500 mg twice daily. On examination she was Cushingoid. Blood tests revealed plasma glucose 28.9 mmol/L, serum cortisol 4 nmol/L, TSH 0.07 mU/L, fT4 7.4 pmol/L, fT3 1.6 pmol/L, LH <0.1 U/L, FSH 1.2 U/L, prolactin 130 mU/L. There was no evidence of diabetes insipidus. MRI pituitary revealed a small pituitary gland, with no focal abnormality. The biochemistry suggests hypopituitarism 8 years after her first tumour removal and radiotherapy. She was treated with glucocorticoid, thyroxine, and gliclazide.

Hypopituitarism following radiotherapy for primary brain tumours has been attributed to the hypothalamus and pituitary gland being in the field of radiation. However, in this case the pituitary was not in the field of radiation. A dosimetric review indicated that the dose of total radiation to the hypothalamus and pituitary gland was negligible, being around 4% of the total dose; we believe this would not account for hypopituitarism. It is possible that a very low dose of radiation to the pituitary gland, outside the primary target volume, may lead to loss of pituitary function, though this seems unlikely. We suggest that other indirect mechanisms may be implicated.

### 3.

<b>Title:</b>	<b>SIADH AS THE FIRST MANIFESTATION OF A CHORDOID GLIOMA OF THE THIRD VENTRICLE</b>
<b>Author(s):</b>	<b>M. Calanchini, M. Hofer, S. Cudlip, A. Fabbri, A. Grossman</b>

A 48 year-old woman presented with polydipsia and polyuria. Diabetes mellitus was excluded, but a serum sodium level of 122 mmol/L was noted. She had no significant past medical history and was on no medication. There was no memory disturbance, nor visual problems, and she was euvolaemic.

Her laboratory results were consistent with diagnosis of SIADH. She underwent brain MRI which revealed a 2.3x1.6 cm avidly-enhancing mass involving the third ventricle, with low and high signal on T2-weighted images and areas of cystic change. It appeared to be separate from the pituitary. MDT meeting recommended biopsy, which showed a moderately cellular tumor with a prominent chordoid pattern of differentiation, with strong positivity for glial fibrillary acidic protein, CD34, TTF-1. Ki-67 was 1-2%. She underwent transcortical debulking, which confirmed the diagnosis of a chordoid glioma (CG). At 10 weeks follow-up, she was well with a serum of 128 mmol/L. At 12 months follow-up, MRI showed stable residual disease with normalisation of sodium and no evidence of endocrine defects apart from secondary hypothyroidism.

This is the first report of CG presenting with SIADH. CG of the third ventricle is a rare and recently described tumour, characterised by a unique histomorphology and exclusive association with the suprasellar/third ventricular compartment. Its histogenesis is still uncertain, but is probably of glial origin. The WHO 2007 classification assigned it as a grade II neoplasm. Clinical, radiological and histological features are pleomorphic and may mimic other types of lesions. The most common clinical presentations are headache, visual and memory disturbance; an endocrine presentation is rare. The treatment is not well established: a less aggressive surgical approach, with or without radiosurgery, may reduce surgically-induced morbidity, but a recurrence is more likely. We decided for a less aggressive surgical approach without radiotherapy, but therefore careful follow-up is mandatory.

4.

<b>Title:</b>	<b>A challenging case of dual endocrine pathology</b>
<b>Author(s):</b>	R Ramli, K. Steer, E. Hatfield, A Mehta, B Jones, N Mendoza, K Meeran, NM Martin. Imperial College Healthcare NHS Trust, London and London North West Healthcare NHS Trust.
<p>A 39-year-old man presented with thyrotoxicosis and was diagnosed with Graves' disease. Despite high doses of anti-thyroid medication for 18 months, he remained biochemically and clinically hyperthyroid. Therefore, a thyroidectomy was planned. Four days before surgery, he developed double vision and was referred for urgent Neurosurgical review at our centre. On further questioning, he reported a 12-month history of lethargy and low libido. On examination, he had a right 6th cranial nerve palsy and a partial right ptosis. A pituitary MRI showed a large suprasellar lesion with right cavernous sinus involvement. Biochemistry showed prolactin 37,384 mU/L (macroprolactin negative), testosterone 1.6 nmol/L, LH 1.5 IU/L, FSH 1.7 IU/L, T4 16.4 nmol/L, T3 7.2 nmol/L, TSH &lt;0.01 mU/L, cortisol 53 nmol/L and IGF-1 23.2 nmol/L (13-50 nmol/L). His case was discussed at the ICHNT Pituitary MDT meeting. His thyroidectomy was cancelled and he commenced cabergoline 0.5 mg/ week and hydrocortisone replacement. Serum prolactin fell quickly to 5456 mU/L after one dose of cabergoline 0.5mg. Both the right-sided ptosis and 6th nerve palsy resolved.</p> <p>With regards to his thyroid dysfunction, TSH receptor antibody level was raised at 2.1 u/ml (ULN 0.3) with a persistently elevated fT3 and undetectable TSH. He elected for radioactive iodine treatment rather than thyroidectomy as a definitive cure for his Graves' disease. Nine weeks following radioactive iodine, he commenced thyroxine replacement (fT4 9.3 nmol/L).</p> <p>A subsequent pituitary MRI showed a significant reduction in the size of the prolactinoma, albeit with persistent right parasellar extension to the cavernous sinus. Prolactin reached a nadir of 437 mU/L. However, he demonstrated low mood, with aggression and anger at out-patient review . We have concerns regarding continuing dopamine agonist therapy. His compliance with cabergoline has lessened with a resultant rise in prolactin (2872 mU/L).</p> <p>Questions for discussion:</p> <p>What are the options for this gentleman if we decide dopamine agonists are worsening his mental state? Will surgery alone be sufficient in view of probable cavernous sinus involvement?</p>	

## 5.

<b>Title:</b>	<b>A challenging case of acromegaly complicated by cardiac failure</b>
<b>Author(s):</b>	<p><b>R Ramli(1), S Mehta(2), P Valsalakumari(1), B Jones(3), A Mehta(3), N Mendoza(4), M Williams(5), K Meeran(1), NM Martin(1), E Hatfield(1)</b></p> <p><b>1. Imperial Centre for Endocrinology, 2. Ealing Hospital, London North West Healthcare NHS Trust, 3. Department of Neuroradiology, 4. Department of Neurosurgery, 5. Department of Oncology Charing Cross Hospital, Imperial College Healthcare NHS Trust, London.</b></p>
<p>A 43 year old lady presented to her local hospital with a 2 month history of dyspnoea, weight gain, amenorrhoea, visual disturbances and abdominal swelling. She had a past medical history of insulin-treated type 2 diabetes, hypertension and thyroidectomy for multinodular goitre. On admission she was noted to have features of acromegaly along with signs of congestive cardiac failure. Investigations confirmed markedly elevated growth hormone (mean GH 94.2ug/l) and hypogonadotropic hypogonadism with a prolactin of 105mIU/l. Echocardiogram showed a severely dilated LV with impaired global function (LVEF 25%). CT coronary angiogram and cardiac catheterisation studies demonstrated moderate stenosis in the mid LAD. MRI pituitary revealed a large pituitary macroadenoma compressing the optic chiasm and invading the right cavernous sinus. Formal visual field test results were unreliable. A diagnosis of acromegalic cardiomyopathy was made and she was started on Lanreotide autogel 120mg monthly. She was referred to our centre for urgent neurosurgical review. A cardiology opinion was obtained and it was felt that she was unsuitable for general anaesthesia due to the high mortality risk associated with cardiac dysfunction. After a pituitary MDT discussion, it was planned to optimise her cardiac failure medically, and to reduce her growth hormone burden with the addition of cabergoline and uptitration of Lanreotide. Despite this, her growth hormone burden remained high (mean GH 26.4ug/l, IGF-1 68.5nmol/l). Her cardiac function did not improve (LVEF 20%) so therefore she remained high risk for neurosurgery. Following further pituitary MDT discussion, she was referred for pituitary radiotherapy. Two months after completion of radiotherapy, her IGF-1 and random GH are 71.6 nmol/l and 17.9 ug/l respectively. She remains on weekly Lanreotide and Cabergoline, but has been difficult to follow up due to poor clinic attendance. Addition of Pegvisomant is planned.</p> <p>Our questions for the expert panel are:-</p> <ol style="list-style-type: none"> <li>1. What other options shall we pursue to treat her acromegaly or should we wait for the radiotherapy to take effect?</li> <li>2. Would any members of the panel opt for surgery despite her poor cardiac function?</li> </ol>	

6.

**Title: Tale of an Untreated MEN1 patient involving Pituitary & Parathyroid**

**Author(s):** Dr Yogesh Kalaiah

MEN 1 syndrome is associated with neoplasms of pituitary, parathyroid and pancreas. The order of presentation neoplasm varies and hence management earliest neoplasm becomes crucial. I report about 76 year lady diagnosed to have Growth Hormone producing Pituitary macroadenoma at the age of 53 years and refused surgery, radiotherapy & medical therapy due to fear of the side effects of the treatments. 20 years later she develops primary hyperparathyroidism with benign large multi-nodular goitre extending to mediastinum. At this stage she is unsuitable for parathyroid surgery due to cardiorespiratory sequel of Acromegaly. She was intolerant to cinacalcet due to gastrointestinal side effects but received IV bisphosphonates to bring down calcium.

Acromegaly sequel included cardiomyopathy with severe aortic stenosis (AS), Obstructive sleep apnoea, osteoporosis, over growth of hand & facial bones, osteoarthritis and T2 DM. The complications of PHPT included renal calculus and pancreatitis needing hospitalisation. She had kypho-scoliosis due osteoporotic spinal fractures, headaches with reduced visual acuity & field of vision but not significant enough to disable her. ACTH was raised with no signs of Cushing's.

Investigations:

IGF1	>999 ug/L
Prolactin	713 mu/L
ACTH	58 ng/L
SST	380/571/634 nmol/L (N)
FSH & LH	<1 u/L
FT4, TSH	19.4 pmol/L, 0.65mu/L, TPO Ab Neg
Ca, PTH	3.08mmol/L, 141ng/L
Spirometry	FEV1/FVC 59%
ECHO	mod to severe left atrial dilatation with severe AS & preserved LV function
SPECT CT	MNG with parathyroid adenoma
MRI Pituitary	Pituitary Macroadenoma

She had frequent admissions to hospital with pulmonary oedema due to severe AS. She had made decision not to have medical therapy for acromegaly even at this stage. This case reflects the importance of counselling such patients about the later presentation of MEN syndrome and their sequel of disabled complications if not treated early.

**Title: Multidisciplinary management of a patient with intractable headache and a pituitary adenoma**

**Author(s): Shapey J, Bouloux P-M, Martin J, Fersht N, Dorward N**

**Case history**

This 30 year old lady first presented in 2009 with an 18 month history of progressive generalised bifrontal headaches. She described no other neurological or endocrinological symptoms and her clinical examination was normal. An MRI scan demonstrated a pituitary adenoma and she subsequently underwent endoscopic transphenoidal resection of her tumour in December 2009. Histopathological analysis confirmed an atypical TSH-producing adenoma.

A post-operative MRI performed in 2010 demonstrated involution of the tumour but in 2012 she represented with further headaches and right-sided facial pain. A repeat MRI demonstrated tumour recurrence predominantly on the right side associated with parasellar extension into the cavernous sinus. A second operation was performed with a good surgical result; however, there remained some residual tumour within the right cavernous sinus so she then underwent a course of radiotherapy (RapidArc IMRT 50.4 Gy in 28 daily fractions).

Her symptoms dramatically improved following treatment but soon recurred and she continues to experience severe cluster headaches with intractable pain on the right side of her face, head and neck. Subcutaneous octreotide injections (50mcg tds) have been the most effective treatment to date but had to be stopped because she developed severe nausea and vomiting. Her pain continues to affect her sleep and she is distressed and low in mood.

**Treatments**

This patient has been reviewed in various specialist clinics and numerous different treatment modalities have been attempted in managing her headaches. Previous unsuccessful treatments include greater occipital nerve blocks, lidocaine infusions, various opioid analgesics (oromorph, oxycodone, fentanyl patches), methadone, lamotrigine, amitriptyline, and acupuncture. Her current medication includes hydrocortisone 30/15/10mg, paracetamol 1g qds, codeine phosphate 60mg bd (sometimes qds), MST 30mg bd, gabapentin 300mg tds and carbamazepine 200mg tds.

**Conclusion and points for discussion**

Cluster headache is the most severe form of primary neurovascular headaches. They are uncommon in patients with pituitary tumours occurring in just 4% of cases but they are associated with cavernous sinus invasion. This lady continues to suffer with daily headaches and current management is failing to address her symptoms. She wishes to undergo further surgery but should this be attempted given the small size of residual tumour evident on her MRI? Percutaneous ganglion blockade and trigeminal rhizotomy may be an alternative option for the treatment of severe headache in selected patients with pituitary adenomas but would such treatment be appropriate for this patient? Are her symptoms being compounded by opioid-induced hyperalgesia or could her medical management be optimised? This complex case demonstrates the importance of close-working multidisciplinary care.

**Contact:**

Rob Bullen  
CFS Events Ltd  
Mindenhall Court  
17 High Street  
Stevenage  
Herts, SG1 3UN

[www.cfsevents.co.uk](http://www.cfsevents.co.uk)  
Tel: +44 (0) 1438 751519

