

ESE Market 2012 Summer 2012 Summer 2012 Summer 2012

THE NEWSLETTER OF **THE EUROPEAN SOCIETY OF ENDOCRINOLOGY** – THE EUROPEAN HORMONE SOCIETY

ICE/ECE 2012



Giants should still fear body snatchers the history of acromegaly

Recent Advances in Adrenal Disease

Also in this issue:

Endo Explorer

visits the University of Cordoba

A Day-in-the-Life of...

A Dublin professor of Endocrinology

WIN WIN WIN!

with the Endo Crossword

In this issue

- 03 Society news **ICE/ECE 2012**
- 04 Society news Welcome to the new Executive Committee members ESE's new Open Access journal
- 05 ESE's Basic Science Committee & Clinical Committee ESE's Basic Science Committee and Clinical Committee EYES news
- 06 The history of Acromegaly Why giants should still fear body snatchers
- 08 Editor's Selection
- 09 Endo Explorer Featuring Spotlight on the Slovak **Endocrine Society**
- 10 Recent Advances in Adrenal Disease
- 11 Day-in-the-Life of... a Dublin Professor of Endocrinology
- 12 Coffee Break

This document is available on the ESE website.

The address lists used to mail this issue of ESE News were supplied by the member societies of ESE and are stored in BioScientifica's database for future use. If you do not wish to receive further mailings, please advise editorial@ endocrinology.org

...........

©2012 The European Society of Endocrinology The views expressed by the contributors are not necessarily those of ESÉ.

Editor: Professor Philippe Bouchard, France Email: philippe.bouchard@sat.aphp.fr

Deputy Editor: Professor Richard Ross, UK Email: r.j.ross@sheffield.ac.uk

Editorial Board: Justo Castaño, Spain Wouter de Herder, The Netherlands Hans Romijn, The Netherlands Dominik Schulte, Germany Leonie van Houten, The Netherlands

Website: www.euro-endo.ora

Newsletter designed by Qube Design Associates

ESE would like to thank its Corporate Members for their continued support: Amgen, CMM Global, DuoCort AB, Eli Lilly, Ipsen, Laboratoire HRA Pharma, Merck-Serono, Novartis, Novo Nordisk, Otsuka Pharmaceuticals Europe Limited, Pfizer, Roche, Sandoz International GMBH

Editorial

I am writing today as the new ESE President. Before presenting our activities and road map for future developments, I would like first to express my gratitude for the support of the ESE members. In addition to the role of President, three new ExecutiveCommittee members were also elected; further information on them can be found on page 2.

I preside over ESE at an exciting time for our Society; ESE has had a successful first five years and is noww in a financial position to be able to invest further in delivering public benefit in European endocrinology. I would like to thank all the previousExecutive Committee members and the two ESEPresidents. Steven Lamberts and, more recently, Ebo Nieschlag for their hard work, achievements, and their continuous commitment to the Society.

This plan is being refined and will be published on the ESE website in due course. ESE will have a special focus on:

• Supporting early-career basic and clinical endocrinologists so that they feel part of the European endocrine community and remain active and contributing members of this community as their careers develop



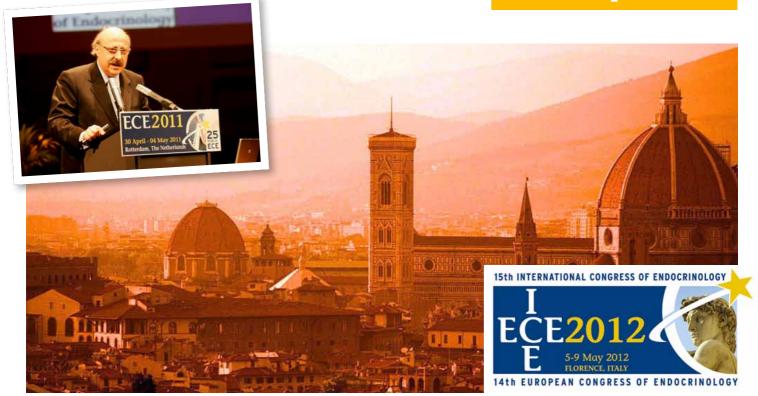
- Promoting equality of opportunity across Europe by providing a programme of support services, such as education, especially to clinicians and scientists in less advantaged countries
- Representing and integrating the full breadth of endocrinology throughout its activities, including the core endocrine diseases and also diabetes, metabolism & obesity, bone & calcium, reproduction, cardiovascular, and oncology; and balancing coverage of common and rare endocrine diseases
- Being the 'voice of endocrinology' in Europe, promoting ESE and understanding of endocrinology to endocrinologists, European legislative and funding bodies, and to the public, working in a collaborative way with national and specialist societies.

ESE will achieve these aims in a range of ways and has set key objectives for the five-year period; these will be shared with the ESE membership in due course. It is clear that the support of all members of the Society is needed. It is time that our Sub-committees recruit members outside our Executive Committee. Details of the Subcommittees, including their remit

NEWLY LAUNCHED

ESE has launched an exciting new-look website as part of the Society's aim to modernise and increase benefits to our members. New sections include: Career Opportunities, Special Interest Group information, a developed Affiliated Society Members section, Educational Videos, Patient materials and much more! You will find the new website at www.ese-hormones.org take a look and let us know what you think! info@euro-endo.org





ICE/ECE 2012

15th International Congress of Endocrinology & 14th European Congress of Endocrinology 5-9 May 2012, Florence, Italy

Lorem ipsum dolor sit amet, consectetuer adipiscing elit. Maecenas porttitor, justo a condimentum luctus, justo nibh tempor ligula, guis blandit metus metus non.

Lorem ipsum dolor sit amet, consectetuer adipiscing elit. Maecenas porttitor, justo a condimentum luctus, justo nibh tempor ligula, quis blandit metus metus non ipsum. Nulla et augue sit amet libero accumsan fermentum. Etiam sit amet enim id libero rutrum tempor. Fusce nec orci. Suspendisse potenti. Pellentesque molestie. Praesent mauris justo, rhoncus porttitor, pretium ac, faucibus eu, elit. Mauris facilisis justo sed ante faucibus gravida. Vestibulum ante ipsum primis in faucibus orci luctus et ultrices posuere cubilia Curae; Sed vulputate. Etiam arcu. Quisque quam sapien, dictum sit amet, tincidunt eget, auctor ut, massa.

Nunc eget sem non tellus porta laoreet. Pellentesque mauris eros, nonummy vel, scelerisque euismod, posuere quis, magna. Praesent tincidunt nonummy sem. In hac habitasse platea dictumst.

Suspendisse metus odio, euismod et, aliquet sed, laoreet in, dolor. Nunc vestibulum lorem eget leo. Proin massa orci, convallis id, malesuada sodales, dignissim a, ligula. Phasellus interdum aliquam sem. Proin ultrices dui. Nam tristique. Nulla sodales aliquam eros. Sed non leo.

Morbi molestie elementum arcu. Fusce quam mi, eleifend ultrices, sagittis nec, lobortis vitae, mi. Vestibulum eget dui. Phasellus mollis quam eu metus. Integer diam. Fusce vehicula, purus iaculis congue ultricies, enim mauris dapibus turpis, eu porttitor justo dolor et est. In varius pretium libero. Nam sed mi vitae diam adipiscing viverra. Sed lectus ipsum, placerat a, malesuada at, luctus at, nisl. Vestibulum libero augue, sagittis tincidunt, scelerisque eget, nonummy nec, velit. faucibus eu, elit. Mauris facilisis justo sed ante Integer id diam id velit feugiat feugiat. In iaculis placerat augue. Nunc nibh magna, volutpat eu, tristique ut, vestibulum vel, neque.

Aliquam convallis erat a mi. In adipiscing. Nulla eu tortor ac mauris placerat tristique. Donec fermentum nulla a risus. Nunc convallis eros sit amet

Congratulations to ICE/ **ECE 2012 Prize Winners!**

The Geoffrey Harris prize was awarded to Professor Jonathan Seckl (UK). ESE would like to thank the sponsor Ipsen for their continued and generous support of this prize.

The European Journal of Endocrinology prize was awarded to Professor Sadaf Farooqi (UK).

Six Young Investigator Awards were presented to:

Dr Francesca Marta F Elli (Italy) Dr Julien J Hadoux (France) Dr Marietta M Stadler (Austria) Dr Justyna J Janik (Poland) Dr Carmello C Quarta (Italy) Dr Immacolata Nettore (Italy)

Four Poster Prizes were also awarded to:

Name 1

Name 2

Name 3

Name 4

If you were not able to attend the ICE/ECE 2012 Congress, the Geoffrey Harris and European Journal of Endocrinology Prize Lectures and eight Plenary Lectures will be available for members to view on the ESE website in due course.

Many congratulations to all the prize winners!



We are pleased to announce that Endocrine Connections, ESE's exciting new Open Access journal is now open for submissions. Published as a

joint venture between the European Society of Endocrinology and the Society for Endocrinology, Endocrine Connections is an important new journal in our field, with a particular focus on rapidly publishing the very best research that has relevance across the many disciplines that intersect with endocrinology, helping to stimulate cross-discipline collaboration. Submit your article at www.endocrineconnections.com

Welcome to new Executive Committee member

Lorem ipsum dolor sit amet, consectetur adipiscing elit. Nunc consequat lobortis enim, sed interdum velit varius vitae. Vivamus venenatis sem quis velit luctus conque. Vestibulum vehicula tincidunt velit, eu cursus velit auctor posuere.

Quisque consequat, erat sed vulputate accumsan, est ipsum pharetra urna, ut ultricies leo mi ut libero. Integer eu ante eget felis scelerisque egestas aliquet sed mauris. Fusce pellentesque leo id sapien dignissim aliquet. Praesent lacinia placerat nisi, nec accumsan velit fringilla ac. Sed

Welcome to our new members

Over 845 new member applications were approved by the Executive Committee during ICE/ECE 2012. ESE's membership is growing each year and we currently have an impressive 2770 members from all over Europe, truly making us the European Hormone Society.

New Honorary members: ESE is pleased to announce that Honorary membership was presented at ICE/ECE 2012 to Professor Wilmar Wiersinga in special recognition of his services to ESE, and to Professor Pierre Chambon in special recognition of his contribution to science. Congratulations to them both.

AGM 2012

Lorem ipsum dolor sit amet, consectetur adipiscing elit. Nunc consequat lobortis enim, sed interdum velit varius vitae. Vivamus venenatis sem quis velit luctus congue. Vestibulum vehicula tincidunt velit, eu cursus velit auctor posuere. Quisque consequat, erat sed vulputate

accumsan, est ipsum pharetra urna, ut ultricies leo mi ut libero. Integer eu ante eget felis scelerisque egestas aliquet sed mauris. Fusce pellentesque leo id sapien dignissim aliquet. Praesent lacinia placerat nisi, nec accumsan velit fringilla ac. Sed posuere aliquet enim, vestibulum gravida est suscipit quis. Phasellus nec libero ut elit hendrerit placerat faucibus nec turpis. Fusce in ultricies eros. Proin

Grants and prizes

Nominations now open for the **European Journal of Endocrinology** Prize 2013 - the prize of a certificate and □10,000 will be awarded to a candidate who has significantly contributed to the advancement of knowledge in the field of endocrinology through publication. Nominations for the prize close on 8 June 2012.

ESE Meeting Grants - there are still 34 grants of up to €1400 available to ESE members! Don't forget to apply for grants for the upcoming Postgraduate course in Turkey and the Summer school in Bregenz.

NEW! Basic Science Meeting Grant – worth up to □450 each.

ESE JOE/JME Prize update Congratulations to Dr Li Chan, of the William Harvey Research Institute, London, on winning the 2012 JOE/JME prize for basic endocrinology.

ASPIRE Young Investigator Research Awards in Endocrinology 2012 For 2012 three awards of \$50,000 will be provided to research proposals that seek to explore basic, translational, or clinical research in the area of endocrine disease in adults and children. For full details please go to http://www.pfizer. com/research/investigator/investigator_ initiated_research.jsp

For full details of the criteria and how to apply for ESE grants and prizes, please see the Prizes Grants and Awards page of the ESE website www.ese-hormones.org

ESE ENDO International Endocrine Scholars Programme (IESP) - 2012

An exceptional training experience for young endocrinologists: the successful candidates receive counsel through a unique mentoring programme that helps them find financial support, advice and esteemed training opportunities.

ESE is pleased to announce that the 2012 IESP scholarships have been awarded to:

ENDO ESE International Endocrine Scholars Programme

Carmelo Nucera, Italy Sebastian Michael Schmid, Germany

ESE Travel Bursary Jovana Kaludjerovic, Serbia

Giampaolo Trivellin, Italy

Congratulations to you all!

Introducing ESE's Basic Science Committee and **Clinical Committee**

Both Committees have versatile action Ilpo and Pia's affiliations plans to achieve their goals with their aims detailed below.

ESE's Clinical Committee:

Advancing and harmonizing clinical practice and research in Europe

1. To play a leading role in the development of guidelines for optimal management of endocrine disorders, and to give guidance on matters in clinical endocrinology, including ethical issues and best practice.

Special Interest Groups have been initiated to establish recommendations for treatment and management in specific areas of expertise: 1) monogenic metabolic bone disorders (Chair: Prof. Östen Ljunggren, Sweden), 2) long-term monitoring of patients operated for pheochromocytoma/ paraganglioma – prognostic indices for tumor recurrence (Chair: Prof. Pierrre-Francois Plouin, France), and 3) management of polycystic ovary syndrome (Chair: Prof. Renato Pasquali, Italy).

The Special Interest Groups will present their recommendations in peer-reviewed publications, and at ESE-sponsored symposia 2013-2014.

- 2. To support all healthcare professionals (clinicians, specialist nurses and allied professions) delivering patient care.
- 3. To support clinical research in Europe, with a special focus on supporting endocrinologists in less economically developed countries, and to advance quality and equality of patient care in endocrinology across Europe.

Professor Pia Burman

ESE's Basic Science Committee:

Promoting basic endocrine science within Europe

- 1. To establish a European basic endocrine scientists' network and to recruit basic scientists as members of ESE
- 2. To establish a basic science training course programme

The Summer School on Endocrinology for young scientists will continue as an ESE-sponsored event (see Save the Dates!). We will also establish a circulating postgraduate course to be held according to topical needs.

3. To establish a high-quality basic endocrinology congress series in Europe

We have joined forces with the Mt St Odile Symposia on Hormones and Cell Regulation, held annually in Alsace, France. Travel grants for young ESE members will be available.

4. To establish a funding programme for basic scientists to attend the ESE congresses and courses

A total of 100 travel grants, □450 each, will be available for basic scientists to attend the European Congress of Endocrinology.

- 5. To develop a funding programme focused on networking and exchange visits within the European basic endocrinology community
- 6. To establish links with other European Societies in the field of endocrine sciences

Professor Ilpo Huhtaniemi

Introducing EYES:

European Young **Endocrine Scientists**



XIVth Annual Meeting, 12-14 October 2012. Dresden, Germany

The European Young Endocrine Scientists (EYES), founded in 2011, is a committee under the patronage of the European Society of Endocrinology (ESE) with a primary goal of increasing the mutual exchange of ideas and knowledge between endocrinologists – from basic researchers to clinicians – in the initial stages of their careers.

As an official committee of the ESE, EYES enables endocrinologists from all ESE member societies to actively contribute to the Society's activities and provides a platform to make young endocrinologists feel welcome at ESE. EYES therefore assists young scientists in developing and finding their own personal path through the different fields of endocrinology.

EYES emerged from a German initiative called Young Active Research in Endocrinology (YARE) and takes advantage of the existing structures of YARE, including the YARE webpage (www.young-activeresearch.eu) and its interactive forum. EYES holds annual meetings exclusively for young scientists where they can present their work, improve presentation skills and establish professional networks. This year, our annual meeting will take place in Dresden, Germany, in October 2012. Participation in EYES activities is free of charge.



Giants should still fear body snatchers

A recent BMJ article (BMJ 2011;343:d7597) made the case for the skeleton of Charles Byrne, "The Irish Giant", to be thrown into the sea, almost 230 years after his death.

It was Byrne's dying wish that his body should be buried deep into the ocean, safe from those 18th Century physicians who wanted instead to dissect and display him. Despite his wishes, Byrne's skeletal remains have been on display at the Hunterian Museum at the Royal College of Surgeons in London for two centuries. But in their BMJ article, Professor of Medical Ethics, Len Doyal, and Thomas Muinzer, a lawyer, argued that it wasn't too late to fulfil Byrne's request to rest in peace at the bottom of the sea. Their pleadings prompted lively debate, both in the BMJ and the world's media.

Charles Byrne suffered from acromegalic gigantism and reached a final height of 2.31 metres (7' 7") before his death at the age of 22 in 1783. Just a year earlier he was put on show in London as a 'curiosity' that Londoners were willing to pay to see. However, not long after his arrival in London, Charles Byrne was robbed of most of the money he had earned as a side-show exhibit and had contracted "consumption" (tuberculosis) and became an alcoholic. He realized that he was a dying man, but death was not his greatest fear, rather it was the physicians who were eager to obtain his body after death for dissection.

One of these surgeons was Dr John Hunter (1728-1793), the "Godfather of Modern Surgery". Charles Byrne was determined to stay out of the hands of Dr Hunter and arranged desperate precautions to avoid such a fate. Byrne instructed that, after his death, his body was to be sealed in a lead coffin. His loyal friends were to guard it day and night until such time that it could be sunk deep into the sea. Byrne pre-paid an undertaker to ensure that his will would be carried out, but alas it was all to no avail. After his death, the remains of Byrne came into the possession of Dr Hunter. Charles Byrne was put on display in Dr Hunter's museum 4 years after his death.

If Dr John Hunter had opened the skull of Charles Byrne he might have been the first to describe pituitary enlargement in gigantism/acromegaly. The famous American neurosurgeon, Dr Harvey Williams Cushing, also known as the "Godfather of Neurosurgery" (1869-1939) reasoned that Hunter never ventured so far because "his passion as a collector exceeded his thirst for knowledge". It wasn't until 1909 that Dr Harvey Cushing, together with Sir Arthur Keith, the curator of the John Hunter museum, opened the skull of Charles Byrne and demonstrated an enlarged sella turcica.

More recently, in 1980, Drs Alexander M. Landolt and Milo Zachmann estimated the "bone age" of Charles Byrne to be only about 17, indicating that he was still growing at the time of his death age 22, implying that he was suffering from (hypogonadotropic) hypogonadism.

Just two years ago, Dr Harvinder S. Chahal and his colleagues at the Charles Byrne's body has undoubtedly assisted the advancement of science, even though his wish was for a private end. He is not, however, the only acromegaly patient to involuntarily provide their body after death for medics and marauders; scientists and sightseers. The practice, or at least the posthumous fascination for giants, has never actually stopped.

Very recently the controversial German anatomist, Gunther von Hagens, tried and failed to gain access to a modern giant so that he could display his 'plastinated' remains after death in his travelling museum. The world's tallest basketball player, acromegalic giant Ukrainian, Alexander Alekseyevich Sizonenko, was 2.39m, 7' 10.1" when he died in St Petersburg earlier this year at the age of 52. Despite rapidly declining health and living on meager means, Sizonenko declined the offer by von Hagens of a monthly income in exchange for the right to posthumously exhibit his "plastinated" body.

Charles Byrne was determined to stay out of the hands of Dr Hunter and arranged desperate precautions to avoid such a fate.

Department of Endocrinology, Barts and the London School of Medicine, extracted DNA from a tooth of Charles Byrne. From this they identified a germ-line mutation in the 'aryl hydrocarbon-interacting protein gene' (AIP). Four contemporary Northern Irish families who presented with gigantism, acromegaly, or prolactinoma were found also to have the same mutation. Using coalescent theory, it was thus inferred that Charles Byrne and these four families with pituitary disorders shared a common ancestor who lived about 57 to 66 generations earlier.

It's clear that little has changed in more than 200 years - there's still an overriding and macabre interest in giants. Endocrinologists, therefore, should warn their acromegalic giant patients: "Watch out! There are still body snatchers about."

Professor Wouter de Herder

Professor of Endocrine Oncology, Erasmus MC, Rotterdam, The Netherlands



AIP mutations and sporadic pituitary macroadenomas

Only 5% of all pituitary adenomas are related to genetic causes, including MEN1, and familial isolated pituitary adenomas (FIPA). Mutations of the aryl hydrocarbon receptor interacting protein (AIP) account for 15-30% of the FIPA kindreds. This study indicates that germline AIP mutations occur in 11.7% of patients < 30 years with sporadic pituitary macroadenomas and even in 20.5% of such patients < 18 years of age, with different pituitary macroadenomas. Testing for germline AIP mutations should be considered in young patients with macroadenomas.

Tichomirowa et al. 2011. Read full article at doi: 10.1530/EJE-11-0304

Professor J A Romijn

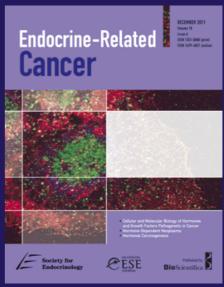
Department of Medicine, Academic Medical Centre, University of Amsterdam



Differentiation of nestin-positive cells

Major questions remain with respect to beta cell neogenesis, and the mechanisms by which b-cell mass is maintained in adulthood. This study reports an effective multistep protocol in a serum-free system, which could efficiently induce beta cell differentiation from multipotent nestin-positive bone marrow stem cells. The differentiated cells not only expressed insulin and glucose transporter 2, but also displayed glucose-responsive secretion of insulin. These results delineate a new model system to study islet neogenesis and possible pharmaceutical targets. Nestinpositive bone marrow stem cells may be therapeutically relevant for beta cell replacement in type 1 diabetes.

Milanesi et al. 2011. Read full article at doi: 10.1530/IOE-10-0344



Oxidative stress and thyroid cancer

Aerobic organisms have complex antioxidant systems that can counteract reactive oxygen species and free radicals. This study measured the total antioxidant status and total oxidant status in sera of patients with thyroid cancer, benign thyroid disease and controls to assess the oxidative stress status of a subject. The data suggest that oxidants are increased and antioxidants are decreased in patients with thyroid cancer, without correlations with thyroid hormone profiles. The clinical relevance of the observations in this cross sectional study should be assessed in prospective studies.

Wang et al. 2011. Read full article at doi: 10.1530/ERC-11-0230

SLC30A8 and type 2 diabetes

The mouse Slc30a8 gene encodes the zinc transporter-8 (ZnT-8). ZnT-8 is thought to be required for providing zinc to allow for proper storage and secretion of insulin. This study detected ZnT-8 in both alpha and beta cells in human pancreatic islets. It also documented that the human SLC30A8 genomic region located in intron 2

contains a conserved islet beta cellspecific enhancer. The authors speculate that it is possible that SNPs that affect SLC30A8 expression, rather than ZnT-8 function, may influence Type 2 diabetes risk.

Pound et al. 2011. Read full article at doi: 10.1530/JME-11-0055





Córdoba, located in the most Southern region of Spain -Andalucía-, has a rich cultural heritage and an ancient historical tradition.

There was a time when Córdoba was one of the most influential cities in Europe in terms of politics, culture and science; now many claim that the only remnants are a breath-taking Mosque and picturesque spots. I do not quite agree. Córdoba is well positioned in terms of scientific research, especially considering our size. Indeed, the University of Córdoba and the Córdoba University Hospital have ranked well in recent national evaluations, despite both institutions being relatively young.

Very much in line with the spontaneous, or even serendipitous, way in which many good things crystallized in Spain in the 1980s, the University of Córdoba and its Hospital witnessed the emergence of active groups working in Endocrinology and its related areas, such as nutrition. In basic endocrinology, two major nodes agglutinated: one in reproductive neuroendocrinology; the other in cellular (and later, molecular) endocrinology of the pituitary gland.

In the last decade, our group (departing from the pioneering work of Enrique Aguilar, Francisco Gaytán and Leonor Pinilla, who are still very active members of our team) has become increasingly interested in deciphering the neuroendocrine and molecular basis of mammalian puberty. Our growing interest partially stems from our involvement in the National Network for Research in Obesity and Nutrition, CIBERobn; a 2006 initiative of the

Spanish Institutes of Health. Similarly, we belong to the recently created Biomedical Research Institute of Córdoba (IMIBIC), named Maimonides after the famous philosopher and medical doctor born in Córdoba in the twelfth century; a liaison that has increased our interest for the implications of our work in translational medicine, as well as our collaborations with clinically-oriented groups.

Well reputed local endocrinology and metabolism groups include the teams of Justo P. Castaño (pituitary and metabolic neuroendocrinology), Mar Malagón (cellular and molecular endocrinology of the adipose tissue) and the clinical team headed by Francisco Perez-Jimenez and Jose López-Miranda at the Córdoba University Hospital (nutrigenomics and lipid metabolism). These groups, including ours, fall under an umbrella of institutions (University of Córdoba, IMIBIC and CIBERobn).

Facing pessimistic news about the funding of Spanish science, we trust that the scientific track record, institutional support and international connections of Córdoban endocrinologists will secure the continuation of our activities in the near future, thus pushing forward biomedical, specifically endocrine, research in Córdoba and making our brains heavier and more influential than our ancient stones.

Manuel Tena-Sempere

Professor of Physiology, Faculty of Medicine, University of Córdoba

The Slovak **Endocrine Society**

The Slovak Endocrine Society (SES) was founded in 1937 as part of the Czechoslovak Endocrine Society. Following the division of Czechoslovakia into the Czech and Slovak Republics, the SES started to work as an independent organization. Nowadays the SES, which is an affiliated member of the European Society of Endocrinology (ESE), has more than 300 members, 36 of whom are simultaneously members of ESE.

Despite the separation of Czechoslovakia, both the Slovak and Czech Endocrine Societies continued in the tradition of joint endocrine meetings, alternating every year between the Czech and Slovak Republics. When the meeting takes place in the Czech Republic, the SES organizes Slovak endocrine workshops on a specific topic. As a nonprofit organisation, the SES promotes research and training in endocrinology, supporting basic and clinical research by providing scientific grants and prizes to researchers, including travel grants to allow young endocrinologists to attend the European Congress of Endocrinology.

Professor Ivica Lazúrová, MD, PhD President of the Slovak Endocrine Society

Recent Advances in Adrenal Disease

In phaeochromocytoma the famous "10% rule" that 10 % of tumors are hereditary has been abandoned and a growing number of familial cases are observed.

The latest is MAX, the MYC associated factor X gene (Comino-Méndez et al., Nature Genetics 2011). In 1694 patients with phaeochromocytoma or paraganglioma mutations in the MAX gene were found in 1.12% (Burnichon et al., Clinical Cancer Research, 2012). This now brings up the number of phaeochromocytoma susceptibility genes to 10: RET, VHL, SDHA, SDHB, SDHC, SDHD, SDHAF2, NF1, TMEM127, and MAX. Thus, 30-40% of phaeochromocytomas or paragangliomas have germline mutations. A challenge in pheochromocytoma/paraganglioma is the diagnosis of malignancy prior to metastases. Using modern catecholamine metabolomic profiles Eisenhofer et al. (European Journal of Cancer 2011) demonstrated that plasma methoxytyramin holds promise as a biomarker for metastatic paragangliomas.

The power of exome sequencing was demonstrated in aldosterone-producing adenomas (APA). Sequencing of only four APA blood pairs revealed a mutation in the potassium channel KCNJ5 in 2 cases. Expanding the series to 22 human APAs and focussing exclusively on KCNJ5, mutations were found in 8 cases. Furthermore, a family with an inherited KCNJ5 mutation was described presenting with a severe form of aldosteronism and massive bilateral adrenal hyperplasia (Choi et al., Science 2011). These exciting findings were further validated by a comprehensive analysis of 380 patients with APA (Bulkroun et al., Hypertension 2012). Somatic KSNJ5 mutations were found in an impressive 34% of patients. Unfortunately, these advances do not yet impact on the difficult clinical management of patients with primary aldosteronism. However, it is predicted that modern sequencing will soon identify the genetic cause of bilateral hyperplasia which could greatly simplify the differential diagnosis between unilateral and bilateral aldosteronism.

"It is predicted that modern sequencing will soon identify the genetic cause of bilateral hyperplasia"

In adrenocortical cancer (ACC) Arlt et al. (JCEM, 2011) have demonstrated the power of steroidobolomics using GC-MS. This sensitive non-invasive tool may greatly facilitate the diagnosis of ACC



and early detection of tumor recurrence. Targeted treatments for ACC have been disappointing. However, mitotane, the standard drug for advanced ACC, has recently been found to massively induce CYP3A4 leading to profoundly increased metabolism of a large number of drugs, including most targeted therapies (van Erp et al., European Journal of Endocrinology 2011, Kroiss et al., Clinical Endocrinology 2011). Thus, insufficient drug levels may have played a major role in the negative trials using targeted treatments, as the effect of mitotane on CYP3A4 persists for many months after cessation.

Adrenal insufficiency is associated with impaired well-being and increased mortality. Despite patient education, adrenal crisis, occurs with an incidence of 8 per 100 patient years (White & Arlt; Hahner et al., European Journal of Endocrinology 2010). Improved treatment strategies are a major goal and a hydrocortisone dual-release formulation has been shown to allow administration once daily (Johannssen et al., JCEM 2012). However, a more physiological cortisol profile (with hydrocortisone release in the hours before waking) is probably needed to restore well-being and may be particularly important for patients with congenital adrenal hyperplasia.

Recent progress in adrenal disorders has been supported by strong European networking (e. g. European Network for the Study Adrenal Tumours, EURADRENAL consortium).

Professor Bruno Allolio,

University of Wuerzburg

1. I. Comino-Mendez et al., Nat Genet 43, 663 (Jul, 2011). 2. N. Burnichon et al., Clinical Cancer Research, (March 27, 2012). N. Burnichon et al., Clinical Cancer Research, (March 27, 2012).
3. G. Eisenhofer et al., Eur J Cancer, (Oct 28, 2011). 4. M. Choi et al., Science 331, 768 (Feb 11, 2011). 5. S. Boulkroun et al., Hypertension 59, 592 (March 1, 2012). 6. W. Arlt et al., J Clin Endocrinol Metab 96, 3775 (Dec, 2011). 7. M. Kroiss et al. Clin Endocrinol (Oxf) 75, 585 (Nov, 2011). 8. N. P. van Erp et al., Eur J Endocrinol 164, 621 (Apr, 2011). 9. S. Hahner et al., Eur J Endocrinol 162, 597 (March 1, 2010). 10. K. White, W. Arlt, Eur J Endocrinol 162, 115 (January 1, 2010). 11. G. Johannsson et al., J Clin Endocrinol Metab 97, 473 (February 1, 2012).



A Day in the life of...



A Dublin Professor of Endocrinology

06.30
Physiotherapy, following shoulder surgery. My physio described this as "voluntarily hurting yourself 270 times a day for the next three months".

11.00
Start morning tutorial. The junior doctors dread these interactive sessions but express gratitude afterwards. Like physiotherapy, tutorials are painful, but ultimately beneficial.

10.00
Diabetes clinic. Large queues dictate that we work fast; I hope the medical students absorb enough to keep them conscious. Between patients I field phone calls from those family doctors canny enough to realise I am a sitting duck. One of them simply wants tickets for Dublin's next hurfing match and through gritted teeth I concede that I'll do my best.

13.00
Clinic draws to an exhausted conclusion. Outside a crowd jostles for attention, my secretary prevails. I adjudicate on clinic defaulters – two strikes and they are discharged (along with the match tickets). A new neurosurgical referral is prolactin macroadenoma but VHI positive (VHI is our private insurance provider) and the surgeon is keen to operate. Firm diplomacy ensues, along with a prescription for cabergoline. I proceed to the neurosurgical ITU to review a patient with symptomatic hyponatraemia and negotiate a plan with the match tickets). Once out of earshot I encourage my SpR to double check them himself. Those anaesthetists will say anything but their prayers.

13.55
Review patient with septicaemia then bolt down a sandwich so fast that dyspepsia seems an inevitable consequence. They say that when God made time he made plenty, but I wish he'd sent a bit more my way.

14.00
The pituitary clinic. My second patient looks awful; an enthusiastic QP trainee has been gradually reducing her thyroxine dose and I continue to suppress thoughts of violence as I dictate the clinic letter. My next patient, an auld lad from the country, leaves a bag of spuds as a thank you, a practice officially discouraged and unofficially very much appreciated. Colleagues in country hospitals regularly

officially discouraged and unofficially very much appreciated. Colleagues in country hospitals regularly benefit from freerange eggs, salmon and, on occasion, live poultry!

I head up to review the septicaemic patient – the SHO has stayed well beyond the time that the lunatics in Brussels dictate that he should, and has done a fine job. I file his name in the dark recess of my brain marked "potential registrars".

Head off to Parnell Park and my duties as doctor to the Dublin Hurling team. I change into a tracksuit and enjoy the smell of liniment and the symphony of the rough banter between the players, the clack of studs on the dressing room floor and the barked commands of the trainers. Our captain has type 1 diabetes, and in a corner of the dressing room he checks his glucometer reading as a prelude to the decision to opt for novorapid or Lucozade Sports. Next to him is a hero who is infusing Factor V111; haemophilia has not prevented him from playing the fastest and most violent ball sport on the planet. Training is brisk, efficient and exhausting and I only have one significant injury – a dislocated pinkie which I put back into place and buddy-strap, pitchside. The stoic victim grunts thanks and rejoins the fray. These guys are tough.

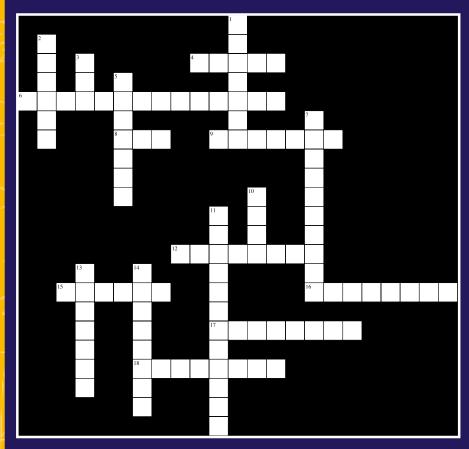
I decline post training hot stew and head home. My youngest lad is just back from training with our club U13 team. I inspect an oozing scrape on his knee; "The other fella was worse, Da'" he insists with pride. As I finish my salad I am happy to look on as my wife applies Savlon and dresses the wound.

After all, a Professor has to delegate sometimes...

J. Thompson

Academic Department of Endocrinology, Beaumont Hospital,





Endo Crossword

Across

- Nobel prizewinner "for the development of radioimmunoassays of peptide hormones", second name (5)
- 6. Male congenital birth defect (14)
- 8. Major risk factor for osteoporosis (3)
- 9. _____'s syndrome, post-partum necrosis of anterior pituitary (7)
- 12. The 'cuddle' hormone (8)
- 15. Element of the thyroid (6)
- 16. Cryptic clue: part of escort is older, shows stress (8)
- 17. Cryptic clue: Debate is raging to find cause of ulcer (8)
- 18. Trained iguanas (anagram): ESE ExCo member, second name (8)

Down

- 1. Described gonadal dysgenesis, second name (7)
- 2. See 18 across, first name (6)
- 3. A plenary speaker who can make you blush (3)
- 5. See 4 across, first name (7)
- 7. Erotic gain (anagram): disease that's your fault! (10)
- 10. See 1 down, first name (4)
- 11. Cryptic clue: The butterfly effect. A hormonal catastrophe? (7,5)
- 13. ______'s oil, possible preventative treatment for Adrenoleukodystrophy (7)
- 14. Virilising hormone (8)

Did you know?

According to a controversial 2004 study, in the Olympics of 2156 women will beat men in the 100-meter run*



*Tatem and colleagues extrapolated winning 100-metre times since 1900 to forecast how race times would change in the future. Tatem et al. (2004) Nature 30: 431(7008): 525. doi: 10.1038/431525a

endo Lingo

HASHIMOTO'S DISEASE

"Hashimoto's disease is an autoimmune disease which causes inflammation of the thyroid gland and results in a reduction in thyroid hormone levels." Also known as chronic thyroiditis. The symptoms of Hashimoto's disease vary, but usually include a swollen thyroid gland (goitre) and hypothyroidism. Hashimoto's disease causing hypothyroidism is found in just above 2% of the population.

Credit: www.yourhormones.info

Save the Dates

Summer School on Endocrinology

5-9 August 2012

Monastery Mehrerau in Bregenz, Austria.

37th Symposium on Hormones and Cell Regulation European Society of Endocrinology (ESE)

11-14 October 2012

Mont Ste Odile, France.

ESE Postgraduate Training Course in Clinical Endocrinology

18-21 October 2012

Antalya, Turkey.



ESE Clinical Update 2012

11-12 January 2013

Abu Dhabi, United Arab Emirates

Register your interest today by emailing info@euro-endo.org

ECE 2013



27 April-1 May 2013

Copenhagen, Denmark.

ECE 2014

3-7 May 2014

Wroclaw, Poland.

Invitation to host ECE 2017 now open

Proposals are invited from ESE Affiliated Societies and the Executive Committee to host the annual European Congress of Endocrinology in their country in 2017.

The ECE is organised centrally by ESE in close collaboration with the host national society. For more information please read the guidelines on the Congress Proposals page of the ESE website. Proposal deadline:

31 July 2012