Insights into MANaging Growth for Endocrine Nurses

October 9-10 2014, Prague, Czech Republic
A Springer Healthcare Medical Education Event for Paediatric Nurses
Dear Delegate,

Welcome to Prague.

The Insights into MAnaging Growth for Endocrine nurses (IMAGE) symposium, organised by Springer Healthcare, is an important opportunity for further training of nurses involved in paediatric endocrinology practice. The major theme of the symposium is related to the diagnosis and treatment of growth disorders. There will be plenary lectures, followed by generous time for discussion, and parallel break-out sessions dedicated to responses to growth hormone therapy in growth hormone deficiency, short stature related to birth weight small for gestational age, and Turner syndrome.

There will be an emphasis on informality, discussion and interaction between delegates and faculty. The IMAGE symposium offers an important opportunity for nurses to be updated on new developments in the growth field and to learn about the important opportunities that paediatric endocrinology can offer to nurses interested in becoming involved in this exciting branch of paediatrics.

We hope that you enjoy this highly informative symposium.

Kate Davies

Chair
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Organisers and CME

Organisers

Springer Healthcare

This educational activity has been planned and independently implemented by Springer Healthcare, with collaboration on programme development from Siyemi Learning.

Continuing Medical Education

This continuing education activity has been approved by the International Council of Nurses (ICN) for the award of 7.3 International Continuing Nursing Credits (ICNECs).
Programme

8 October

19:00 Welcome buffet

9 October

09:00 - 09:10 Welcome, Introduction and Objectives of the Symposium

Session One: Normal And Abnormal Growth
Chairs: Kate Davies & Martin Savage

09:10 - 09:30 Theoretical and practical aspects of anthropometric measurements
Lee Martin

09:30 - 10:00 What is the IGF system and why is it important?
Peter Bang

10:00 - 10:30 Genetic control of growth: Examples of abnormalities
Marie-José Walenkamp

10:30 - 10:45 General discussion

10:45 - 11:15 Coffee

Session Two: Hormone Therapy For Growth
Chairs: Pauline Musson & Marie-José Walenkamp

11:15 - 11:45 GH and other hormone resistance states: Implications for growth
Martin Savage

11:45 - 12:15 Are current indications for GH therapy justified?
Pierre Chatelain

12:15 - 12:30 General discussion

12:30 - 13:30 Lunch

Session Three: Parallel Break-Out Groups

13:30 - 15:15 Turner syndrome
Chairs: Pierre Chatelain & Susan Rybansky

A patient with a good response to GH therapy - Why?
Irena Hozjan

A case of late diagnosis and poor response to GH
Martin Savage

Summary by Chairs and Q&A
Session Three: Parallel Break-Out Groups (continued)

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<tr>
<td>13:30 - 15:15</td>
<td>Small for gestational age</td>
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<td>Chairs: Marie-José Walenkamp &amp; John Chaplin</td>
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<td>A patient with a good response to GH therapy - Why?</td>
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<td>Lee Martin</td>
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<td>15:15 - 15:40</td>
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Session Four: Short Stature

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<th>Time</th>
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<tr>
<td>15:40 - 16:10</td>
<td>A balanced view of idiopathic short stature management</td>
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<td>Martin Savage</td>
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<td>16:10 - 16:30</td>
<td>Brief summary and key messages for the day</td>
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<td>Kate Davies</td>
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<td>16:30</td>
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<td>19:30</td>
<td>Symposium dinner</td>
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10 October

Session Five: GH responses in GH deficiency: Examples and explanations for good and poor responses to GH

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<th>Time</th>
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<tr>
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<td>Case Study: A good response to GH therapy - Why?</td>
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<td>Case Study: A poor response to GH therapy - Why?</td>
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<td>Discussion</td>
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<td>09:40 - 10:10</td>
<td>Panel discussion on treatment of GH deficiency</td>
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<td>Chair: Pierre Chatelain</td>
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<td>Panel: Kate Davies, Pauline Musson, Marie-José Walenkamp</td>
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<td>10:10 - 10:30</td>
<td>Coffee</td>
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Programme

Session Six: Psychological Stress In Short Stature And Adherence To GH Therapy
Chairs: Kate Davies & Martin Savage

10:30 - 11:00  How stressful is short stature?  John Chaplin
11:00 - 11:30  Is poor adherence an issue in GH therapy?  Svante Norgren
11:30 - 11:50  Panel discussion on the psychological pressures of short stature and their potential effects on adherence in GH therapy
Chairs: Kate Davies & Martin Savage
Panel: John Chaplin, Svante Norgren, Irena Hozjan, Peter Laing

11:50 - 12:00  Summary of Symposium and Closing Remarks  Martin Savage
12:00  Close
Venue, Location and Map

Conference Venue

The meeting and delegate accommodation are in the Novotel Praha Wenceslas Square (38 Katerinska, 1200 Prague). Located in the historic part of Prague, the hotel is a 5-minute walk from the famous Wenceslas Square, National Museum, National Opera or Prague Convention Center.

Airport Transfers

Transfers to and from the airport will be provided for all delegates.
Please look for a driver displaying the meeting name in the airport arrival hall.
Information for Delegates

Registration

Delegates should register at the registration desk in the lobby of the Novotel Praha Wenceslas Square. Registration is open 8 October 16:00 - 22:00 and 9 October 08:00 - 09:00.

Symposium Dinner

The dinner will take place at Restaurace Kamenný most on 9 October. Delegates are asked to meet at 19:30 at the lobby of the Novotel Praha Wenceslas Square. Coaches will leave from the Novotel and will give delegates a short guided tour en route to the restaurant.

Luggage

Delegates have the option of either delaying checkout until 13:00 on 10 October or storing their luggage in the hotel luggage room until the end of the symposium. Please see the front desk of the hotel to make either arrangement.

CME Certification

Following successful completion of the provided Attendance Verification Form, delegates will receive certificates indicating that they have earned 7.5 International Continuing Nursing Education Credits (ICNEC) after the symposium by post.

Point of contact

If you have any problems or questions during the meeting, please contact the registration desk or a Springer Healthcare representative.

Filming and Photography

Springer Healthcare may film or take photographs at the symposium that may be used in publicity and marketing materials. Your attendance at the meeting may mean you are featured in such photographs and films and, unless you notify the registration desk of your objection to this, your consent will be implied.
Speaker Biographies

Kate Davies – Chair
Clinical Nurse Specialist
Great Ormond Street Hospital NHS Trust, London

Professional experience:
Kate Davies has spent 15 years working as a Clinical Nurse Specialist in Paediatric Endocrinology at Great Ormond Street Hospital, London, and has a thorough knowledge of all the growth hormone preparations on the UK market and their indications.

Martin Savage – Co-Chair
Emeritus Professor, Professor of Paediatric Endocrinology
Barts and the London School of Medicine & Dentistry, London

Professional experience:
Dr Savage is Emeritus Professor of Paediatric Endocrinology at Barts and the London School of Medicine and Dentistry and consults at The London Clinic Centre for Endocrinology. He has 30 years’ clinical experience in paediatric endocrinology and treatment with growth hormone and recombinant human IGF-1. He has written many publications on growth disorders and on growth hormone resistance states. He was a member of the Organising Committee for the 2008 Consensus Meeting on the management of idiopathic short stature.

Pierre Chatelain – Co-Chair
Professor of Pediatrics Hôpital Mère-Enfant de Lyon Service d’Endocrinologie & Diabétologie
Pédiatriques Groupement Hospitalier, Lyon

Professional experience:
Dr Chatelain is Professor of Paediatrics at Université Claude Bernard, Lyon, France. He is Chairman of the Collège de Pédiatrie and coordinator of the French National Reference Centre of Rare Diseases of Sex Differentiation and Development. He has authored or co-authored more than 170 publications in the field of paediatric endocrinology, diabetology and paediatrics.
Peter Bang

**Professor of Pediatrics and Senior Consultant Pediatric Endocrinology and Diabetes**

**Linköping University, Sweden**

**Professional experience:**

Dr Bang is Professor of Paediatrics and Academic Head of the Division of Paediatrics, Linköping University. He graduated from the University of Copenhagen Medical School and received his PhD from the Karolinska Institutet, Sweden. He has more than 25 years’ research experience in the GH/IGF field and has a publication list of more than 50 peer-reviewed papers.

John Chaplin

**Psychologist**

**Sahlgrenska Academy at the University of Gothenburg, Sweden**

**Professional experience:**

Dr Chaplin is a psychoendocrinologist with experience in treating paediatric patients in a growth research centre. He received his PhD from the City of London and is a chartered psychologist in the UK and Sweden, an Associate Fellow of the British Psychology Society, and an ambassador for epilepsy. For the past 10 years, his clinical and research experience has focused on working with children treated with growth hormone, and he has conducted cognitive and behavioural follow-up studies of children treated with growth hormone from pre-pubertal to final height.

Irena Hozjan

**Paediatric Endocrine Nurse Practitioner**

**Hospital for Sick Children “SickKids”, Toronto**

**Professional experience:**

Irena Hozjan has been a Nurse Practitioner in the Paediatric Endocrine Clinic at SickKids, Toronto, Canada, for over 10 years. She has co-edited a book entitled ‘Turner Syndrome: Across the Lifespan’ and authored or co-authored a number of chapters in this resource.
Speaker Biographies

Peter Laing
Advanced Paediatric Nurse Practitioner for Endocrinology
Alder Hey Children’s NHS Foundation Trust, Liverpool

Professional experience:
Peter Laing worked as a Clinical Nurse Specialist for 10 years and for the past 18 months as an Advanced Nurse Practitioner. He is an independent nurse prescriber, holds a professional qualification in the UK which meets accredited standards as an Advanced Nurse Practitioner, and provides a nurse-led clinic for children who are referred as new patients to the specialty.

Lee Martin
Clinical Nurse Specialist in Paediatric Endocrinology
Barts Health NHS Trust, London

Professional experience:
Lee Martin is a Clinical Nurse Specialist in Paediatric Endocrinology with over 10 years of experience. He is a contributor towards the RCN Competency Framework for Paediatric Endocrine Nurse Specialists and has worked with families of children requiring growth hormone treatment throughout this 10 year span.

Pauline Musson
Clinical Nurse Specialist - Paediatric Endocrinology
University Hospital Southampton NHS Foundation Trust, UK

Professional experience:
Pauline Musson is a RGN/RN Child with a Post Graduate Certificate in Advancing Professional Practice (Paediatric Endocrinology). She has been working as a Clinical Nurse Specialist in Paediatric Endocrinology since 1994 and during this time has continuously developed the service. Her role has evolved to include areas such as endocrine function testing, nurse-led clinics and nurse prescribing.
Svante Norgren
Professor and Director Children’s hospital
Astrid Lindgren Children’s Hospital, Stockholm

Professional experience:
Dr Norgren is a senior consultant in paediatric endocrinology. He received his MD and PhD from the Karolinska Institutet and his further clinical and scientific training at the Karolinska University Hospital and Harvard Medical School. He has served as secretary of the Swedish Paediatric Society and is presently chairman of the Swedish Paediatric Endocrinology Society and director of the Astrid Lindgren Children’s Hospital at the Karolinska University Hospital.

Marie-José Walenkamp
Paediatric endocrinologist
VU University Medical Center, Amsterdam

Professional experience:
Dr Walenkamp is a paediatric endocrinologist at the Vrije Universiteit Medical Centre, Amsterdam, the Netherlands, and Vice Director of the Paediatric Residency Programme. Since 2009, she has been the secretary of the Dutch Growth Hormone Advisory Group. She studied medicine at the University of Utrecht and specialised in paediatrics at Leiden University Hospital, the Netherlands. Her special interests and research focus on molecular defects in the growth hormone/IGF-I axis.
Lee Martin

Theoretical and practical aspects of anthropometric measurements

Auxology is an essential part of growth assessment in children and young people. Most frequently it is part of the nurse’s role to carry out the anthropometric measurements which make up auxological observations. While weight and height are the two most commonly performed measurements, other detailed anthropological techniques should also be considered such as: body mass index, height velocity, sitting height, mid-parental/target height and head circumference.

In order to perform these measurements accurately and reproducibly, it is essential for the nurse to have a good understanding of how to carry out these techniques precisely and also the theory behind them. This includes a solid knowledge of the normal parameters for age, sex and ethnicity so that those individuals who deviate from the normal patterns can be identified rapidly.

Marie-José Walenkamp

Genetic control of growth: Examples of abnormalities

Intrauterine growth is determined by maternal, placental and foetal factors. Of the foetal factors insulin, IGF-I and IGF-II are the main regulators of growth. Defects in the genes encoding these factors result in intrauterine growth retardation. The clinical consequences of defects in these genes will be demonstrated by presenting cases with mutations or deletions in the IGF-I and IGF-I receptor gene. Disturbances in the regulation of the IGF-II gene result in the Silver Russell syndrome, which is also characterised by intrauterine growth retardation. For normal postnatal growth an intact growth hormone–IGF-I axis is essential. Genetic disorders have been identified in various components of the growth hormone–IGF-I axis. These molecular defects and the clinical features of patients will be summarised in the presentation. Finally causes of disproportionate short stature will be presented.
GH resistance states: Implications for growth

Compared to GH deficiency, GH resistance (GHR) is relatively rare. But it remains an important cause of short stature. GH resistance is caused by a defect in GH action, i.e., effect. This contrasts with GH deficiency which is an abnormality in GH secretion, i.e., production. In GHR the GH–IGF-1 axis is disturbed. A number of primary genetic defects in this axis can cause GHR. The most common is a mutation in the GH receptor that gives rise to the classical GHR syndrome described by Laron in 1966 and known as Laron Syndrome. All forms of GHR are compared to Laron syndrome, which is characterised by extreme post-natal growth failure, mid-facial hypoplasia, hypoglycaemia and high GH levels, contrasting with very low levels of IGF-1, IGFBP-3 and ALS. GHR can also be caused by mutations of the STAT gene, the IGF-1 gene and the ALS gene. Milder phenotypes are associated with dominant negative and pseudo-exon mutations of the GH receptor gene. Most genetic defects are inherited as autosomal recessive traits.

A second form of GHR exists in association with primary inflammatory or nutritional diseases such as Coeliac disease, Crohn’s disease, cystic fibrosis and idiopathic juvenile arthritis. GHR is also recognised in critical illness states and following major surgery. In these disorders, the secretion of pro-inflammatory cytokines in addition to nutritional deficiency, if present, disturb the function of the GH receptor and the synthesis of IGF-1 and IGFBPs to decrease the circulating concentrations of IGF-1 leading to deficient linear growth and anabolic functions.

GHR, particularly in its primary form, can be successfully treated by replacement with rhIGF-1. This treatment is licensed by the FDA and EMA and is administered twice daily by SC injections in a recommended maintenance dose of 120 μg/kg/dose. Long-term benefit has been achieved with this therapy. Side effects in the Laron syndrome children are: hypoglycaemia, tonsillar enlargement and benign intracranial hypertension.
A balanced view of idiopathic short stature management

Idiopathic short stature (ISS) is not a specific diagnosis. It is a descriptive term for a heterogeneous collection of patients with short stature of undefined aetiology. The management of ISS is controversial and divides clinical opinions. The widespread availability of recombinant hGH (human growth hormone) after 1985 led to therapeutic trials in ISS subjects. After >5 years of placebo-controlled hGH therapy, growth was shown to be significantly increased in ISS subjects treated with hGH. Data from several trials were submitted to the FDA and in 2003 ISS received FDA approval as an indication for hGH therapy. In contrast, several applications have been made to the EMA, but none has been successful. Consequently, hGH is not approved for treatment of ISS in Europe. The short term studies have shown increased growth on hGH, but the long-term results are disappointing and demonstrate that only 3–5 cm are gained to adult height. Psychological stress has been proposed as an indication to treat ISS patients, but baseline evidence of psychological disturbance in normal short children is weak and long-term benefit has not been definitively shown in ISS subjects after hGH therapy. However some ISS patients do benefit from hGH therapy. These are likely to be those that have marginal GH deficiency and have been excluded from hGH therapy because of the relative unreliability of pharmacological GH stimulation tests. It is likely that ISS management will remain controversial.

How stressful is short stature

Short stature has been widely regarded to be a liability for the individual, but despite the importance ascribed to the psychological impact of physique in all cultures, methodologically sound research is still lacking. Many of the studies that have been conducted have been criticised for selection bias because the individuals studied are drawn from clinical populations. However, there is evidence of short-stature children being bullied in school because of their height and there is increasing evidence that early life stress may have profound long-term effects on the individual’s physiology and psychology which is are not evident during childhood. There is also some evidence to suggest that short-stature adults are disadvantaged both socially and economically. Added to this, there is increasing understanding of the effect of growth hormone deficiency on cognitive abilities and social interactions. In this presentation I will review the evidence on stress related to short stature, with reference to the recent literature and to my own research on this topic.
Abstracts

Svante Norgren

Is poor adherence an issue in GH therapy?

Improving adherence to treatment may have greater impact on treatment outcome and patient safety than improvement in specific medical treatments. There is accumulating evidence that adherence is a significant problem and limits the response to growth hormone treatment. Poor adherence may result from either practical or motivational barriers causing doubts about the necessity of medication or concerns about potential side effects. We must employ objective measures to score adherence and develop effective interventions to improve adherence. Forming quality relationships with the parents and the patients and specifically addressing these potential barriers are key issues in order to improve adherence and treatment outcome. By doing this, we have the potential to reach the full potential of growth hormone treatment.
Independent coverage of the latest clinical developments in Paediatric Endocrinology and Growth Disorders

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