conference syllabus

CPEG GCEP

Canadian Pediatric Endocrine Group
Groupe canadien d'endocrinologie pédiatrique

2013 Scientific Meeting

January 24-26, 2013

Québec City at the Delta Québec

690 Boulevard René-Lévesque Est

In cooperation with:

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Interprofessional Continuing Education

interprofessional.ubc.ca/CPEG2013

Welcome

Dear Delegates,

I would like to welcome you to the 7th Annual Scientific Meeting of the Canadian Pediatric Endocrine Group (CPEG). Our past meetings have been tremendously successful, and this year's meeting promises to offer our nurses, endocrinologists, and trainees another valuable educational experience. The Organizing Committee has worked hard to craft an agenda that highlights work in Quebec City, includes presentations by national and international experts, and emphasizes the work of our fellows.

I would like to thank our sponsors, who make this meeting possible, and in addition thank those companies who sponsor our fellowship awards. Those awards will be announced at this meeting, and they allow us to train future endocrinologists.

I look forward, with you, to an enjoyable and collegial meeting,

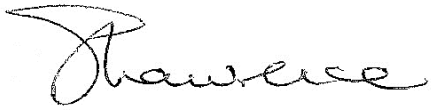
Bienvenue

Chers participants,

J'aimerais vous souhaiter la bienvenue à la 7ème réunion scientifique annuelle du Groupe Canadien d'Endocrinologie Pédiatrique (GCEP). Nos réunions précédentes ont été de belles réussites et cette année encore, notre réunion promet d'offrir aux infirmières (ers), endocrinologues, résidents et autres participants, une expérience éducationnelle de haut niveau. Le comité organisateur a travaillé ardemment pour vous offrir un programme présentant les présents travaux et réalisations du centre hôte, soit Québec, tout en incluant des présentations d'experts nationaux et internationaux en plus de mettre à l'avant-scène le travail de nos résidents.

J'aimerais remercier nos commanditaires qui rendent cette réunion possible ainsi qu'aux compagnies pharmaceutiques qui subventionnent notre programme de bourse aux résidents permettant ainsi la formation de futurs endocrinologues pédiatres. Les récipiendaires de ces bourses seront d'ailleurs annoncés lors de cette rencontre scientifique.

Je nous souhaite donc une rencontre plaisante et empreinte de collégialité!



Sarah Lawrence, MD, FRCPC
Scientific Chair, CPEG 2013 Scientific Meeting

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Fellowship Listing

The Canadian Pediatric Endocrine Group would like to acknowledge and thank the following organizations for their generous support in the form of fellowships:

YEAR	NOVO NORDISK	ELI LILLY	SERONO	HOFFMANN LA ROCHE	SANDOZ
1992-1993		*Lilly I* : M. Lawson			
1993-1994	S. Muirhead (Lawrence)	*Lilly I*: M. Lawson *Lilly II*: A. Simone			
1994-1995	S. Muirhead (Lawrence)	*Lilly II*: A. Simone *Lilly I*: S. Taback			
1995-1996	C. Vaz (50%-one yr)	*Lilly I*: S. Taback *Lilly II*: B. Cummings			
1996-1997		*Lilly I*: J. Hamilton E. Sellers *Lilly II*: B. Cummings			
1997-1998		*Lilly I*: J. Hamilton *Lilly II*: E. Sellers	B. Cummings		
1998-1999		J. Curtis (YR 1)	J. Hamilton		
1999-2000		J. Curtis (YR 2)	J. Hamilton		
2000-2001		C. Panagio-topoulos (YR 1)	C. Huang		
2001-2002		C. Panagio-topoulos (YR 2)		S. Stock	
2002-2003		P. Krishna-moorthy (YR 1)	P. Zimakas	R. McEachern	
2003-2004		P. Krishna-moorthy (YR 2)		H. Bui	
2004-2005		M. Nakhla (YR 1)		J. Simoneau-Roy	
2005-2006		M. Nakhla (YR 2)	I. Chapados	M. Jetha	
2006-2007		BA Wicklow (YR 1)		S. Amed	
2007-2008		BA Wicklow (YR 2)	T. Pinto B. Babic	J. Deladoey	
2008-2009	AM Sbrocchi	P. Olivier (YR 1)		T. Pinto	
2009-2010	R. Shulman	P. Olivier (YR 2)	T. Édouard	S. Runge-Wildi	C. Saaman
2010-2011	E. Bassilious	J. Wasserman (YR 1)		Y. Yeshayahu	S. Tsai
2011-2012	M. Millete	J. Wasserman (YR 2)		C. Zuijdwik	M. Cohen
2012-2013	J. Harrrington	T. Oron	P.Luca	M.Nour	D. Manouski

Program

Please note: 25% of the scientific program will be interactive.

Thursday, January 24, 2013

Time	Session & Presenter
14:00	Fellows Symposium (for CPEG Fellows only) (Room: Lauzon) Part I: Meet the Professor with Dr. Cheri Deal
16:00	Part II: Associate Members' Business Meeting

16:00	CPEG 2013 Registration Opens (Foyer)
17:00	Welcome Reception & Exhibits (Foyer)
19:00	Adjourn

19:00 Satellite Symposium (Room: Duquesne)

Friday, January 25, 2013

Time	Session & Presenter
07:00	CPEG 2013 Registration Re-opens (Foyer)
07:15	Breakfast (Room: Lauzon)
08:00	Opening Remarks & Welcome (Room: Duquesne/Jonquiere) *Dr. Mark Palmert, Dr. Dardye Eugène, Ms. Irena Hozjan, and Ms. Marie Dorion*
	<u>Theme I: Obesity (Room: Duquesne/Jonquiere)</u> Moderators: Dr. Dardye Eugène and Dr. Julie Gagné
08:30	Medical Management of Childhood Obesity: What Can We Learn from Clinical Trials? (Including Q&A) *Dr. S John Weisnagel, Quebec City, QC*
09:15	Obesity Surgery in the Adolescent (Including Q&A) *Dr. Laurent Biertho, Quebec City, QC*
10:00	Refreshment Break & Exhibits (Foyer)
	<u>Theme II: Genetics (Room: Duquesne/Jonquiere)</u> Moderator: Constantin Polychronakos
10:30	Genomic Technologies to Advance Clinical Care in the Pediatric Endocrine Clinic (Including Q&A) *Dr. Kym Boycott, Ottawa, ON*

11:20 Inherited Syndromes of Reduced Sensitivity to Thyroid Hormone (Including Q&A)
Dr. Samuel Refetoff, Chicago, IL

12:10 Lunch (Room: Lauzon) & Exhibits (Foyer)

12:40 Posters and Desserts (Foyer)

13:40 Split Rooms*

Theme III: Biochemistry/Ch (Room: Duquesne/Jonquiere)
Moderator: Dr. Katherine Morrison

13:40 Closing the Gaps in Pediatric Laboratory Reference Intervals: The CALIPER Initiative
Dr. Khosrow Adeli, Toronto, ON

14:10 Endocrine Epiphanies: Laboratory Testing Dilemmas and Evolving Solutions
Dr. Jon Nakamoto, San Juan Capistrano, CA

14:40 Joint Q&A Session

15:00 Refreshment Break & Exhibits (Foyer)

15:30 Abstracts (6) (Room: Duquesne/Jonquiere)
Moderators: Dr. Danièle Pacaud and Dr. Susan Sanderson

17:00 Adjourn

***Nursing Program for Friday, January 25 (Room: Crémazie/Garneau)**
Moderator: Ms. Nicole Kirouac

13:40 Continuing Education Updates
Nicole Kirouac, Winnipeg, MB ICE/ECE 2012 & PENS 2012
Peggy Kalancha, Calgary, AB ESPE 2012

14:30 Topical Anesthetics
Marie Dorion, Quebec City, QC

15:00 Refreshment Break & Exhibits (Foyer)

15:30 Interesting Cases
Sara Chang, Ottawa, ON & Brenda Fraser, Ottawa, ON
Lina Moisan, Montreal, QC

16:00 Round Table Nursing Discussion

17:00 Adjourn

FRIDAY NIGHT DINNER (18:00)
Musée National des Beaux-Arts du Québec, 1 rue Wolfe Montcalm, Québec City
(Bus transportation will be provided. First bus will leave the hotel at 17:45. The second and last bus for the dinner will depart at 18:15. Please see one-pager in your delegate package for detailed information.)

Saturday, January 26, 2013

07:15 Breakfast (Room: Lauzon)

08:00 Business Meeting (Room: Duquesne/Jonquiere)

10:00 CPEG Fellowship Awards (Room: Duquesne/Jonquiere)
Presented by *Dr. Mark Palmert*

10:10 Refreshment Break & Exhibits (Foyer)

10:30 Split Rooms**

10:30 Abstracts (6) (Room: Duquesne/Jonquiere)
Moderators: Dr. Cheril Clarson and Dr. Celine Huot

12:00 Lunch (Room: Lauzon) & Exhibits (Foyer)

12:30 Posters and Desserts (Foyer)

Theme IV: Critical Care (Room: Duquesne/Jonquiere)
Moderator: Dr. Jill Hamilton

13:00 Adrenal Dysfunction in Pediatric Critical Illness: Differing Perspectives (Including Q&A)
Dr. Kusum Menon, Ottawa, ON

13:40 Thyroid Hormone Deiodination in Injury and Nonthyroidal Illness (Including Q&A)
Dr. Stephen Huang, Boston, MA

14:20 Tight Glucose Control in Pediatric Critical Care: Where Do We Stand? (Including Q&A)
Dr. Michael Agus, Boston, MA

15:00 Refreshment Break & Exhibits (Foyer)

15:30 Presentation of Dr. John Bailey Resident Research Award (Room: Duquesne/Jonquiere)

Clinical Debate (Room: Duquesne/Jonquiere)
Moderator: Dr. Rose Girgis

15:35 Debate: Thyroid Nodules: To Remove or Not Remove. That is the Question.
Pro: Dr. Celia Jane Rodd, Montreal, QC
Con: Dr. Johnny Deladoey, Montreal, QC

16:35 Closing Remarks & Evaluation
Presented by *Dr. Mark Palmert*

17:00 Adjourn

****Nursing Program for Saturday, January 26, 2013 (Room: Crémazie/Garneau)**

10:30 CPEN Nurses AGM

12:00 Lunch (Lauzon)

Fellow Abstract Schedule

Time	Title	Presenter	Oral Abstract #	Page
Friday, January 25				
15:30	Predictors of BMI Reduction in the SickKids Team Obesity Management Program (STOMP) for Adolescents with Severe Obesity	Paola Luca	1	17
15:45	Phenotypic Variability in Familial GLI2 Deletion	Despoina Manousaki	2	18
16:00	Congenital Adrenal Hyperplasia due to 11β-hydroxylase Deficiency in a 5-year-old Boy	Karine Khatchadourian	3	19
16:15	hCG Induced Thyrotoxicosis in a 14-year-old Girl.	Shira Harel	4	20
16:30	Post-operative Hypocalcemia - A Case Series and Proposed Approach to Care Following Thyroidectomy	Andrea R. Ens	5	21
16:45	Impact of Elective Hospital Admission on Glycemic Control in Adolescents with Poorly Controlled type 1 Diabetes	Maude Millette	6	22
Saturday, January 26				
10:30	A Short 17-year-old with Primary Amenorrhea: When There is More than Turner Syndrome	Lyne Chiniara	7	23
10:45	A Novel Heterozygous Mutation in the Glucokinase Gene (GCK) with Exercise-induced Symptomatic Hyperglycemia Responsive to Sulfonylurea	Marwa S.E. Ebrahim	8	24
11:00	Growth Failure in Hunter Syndrome - A Contrasting Tale of Two Patients	Munier A Nour	9	25
11:15	Iliac Bone Histomorphometry in Children on Glucocorticoids for the Treatment of Rheumatic Disorders	Jennifer Harrington	10	26
11:30	Type 1 Diabetes and Celiac Disease: Associated Comorbidities and Complications.	Alexandra Tsouka	11	27
11:45	Pde11a and Adrenal Disease. Characterization of a Genetically Manipulated Pde11a-/- Mouse Model	Isaac Levy	12	28

Poster Abstract Listing

Title	Presenter	Abstract #	Page
Discordance Between Perceived and Measured Glycemic Control in Young Adolescent Males with Type 1 Diabetes	Jennifer Harrington	1	29
Process Evaluation of the Living Green, Healthy and Thrifty (LiGHT) Virtual Child Obesity Management Program: Combining Health Promotion with Ecology and Economics	Maria Jogova	2	30
Efficacy of Finasteride for Treatment of Hidradenitis Suppurativa: A Case Series in the Pediatric Population	Sareea Al Remeithi	3	31
A Case of Hypothalamic Hamartoma Associated with Central Precocious Puberty and Growth Hormone Deficiency	Isabelle Rousseau-Nepton	4	32
Impact of the Pediatric Residents' Initiative For Healthy Active Living In Youth (RHALY): A Prospective Cohort Study	Reem A AL Khalifah	5	33
Esophageal Stricture Following Severe Diabetic Ketoacidosis in a 12-year-old Girl	Brenden E Hursh	6	34
The Truthful Teenager: A Rare Case of an Insulinoma as a First Presentation of MEN-1 in a Teenage Male	Karin Winston	7	35
Recombinant Parathyroid Hormone Therapy in Severe Hypoparathyroidism.	Betty Messazos	8	36
Long-Acting Release (LAR) Octreotide in Congenital Hyperinsulinism (CI)	Betty Messazos	9	37
Prolactinoma-The Role of the Endocrinologist in Pituitary Tumors	Betty Messazos	10	38

Program Organizing and Scientific Committee

Robert Barnes
Johnny Deladoey
Marie Dorion
Dardye Eugène
Celine Huot
Nicole Kirouac
Sarah Lawrence
Danièle Pacaud
Mark Palmert
Constantin Polychronakos
Wendy Schwarz
Karin Winston

Dr. John Bailey Resident Research Award

The John Bailey Award is named after Dr. John D. Bailey. Dr. Bailey graduated from the University of Toronto Medical School in 1949, did his training in pediatrics at the Hospital for Sick Children, Toronto and the University of Manitoba in Winnipeg. By the mid-1950s he was firmly established as a major contributor to pediatric care and education at the Hospital for Sick Children and University of Toronto. His major area of interest was in understanding normal and abnormal growth patterns in childhood and adolescence. He was the first chief of the Endocrine Division at the Hospital for Sick Children and an important contributor to the development of the Canadian national growth hormone treatment program. He was the consummate academic: clinically fastidious, intellectually challenging and constantly learning and sharing.

Credits

This event is an Accredited Group Learning Activity (Section 1) as defined by the Maintenance of Certification Program of the Royal College of Physicians and Surgeons of Canada and approved by the Canadian Paediatric Society.

Learning Objectives

The overall learning objective of this meeting is to present the current state of knowledge of topics in pediatric endocrinology and diabetes.

Session Learning Objectives:

Theme I: Obesity

Medical Management of Childhood Obesity: What Can We Learn from Clinical Trials?
John Weisnagel, MD FRCPC, Chief of Endocrinology, Centre Hospitalier Universitaire de Québec CHUL-CHU, Department of Medicine and of Paediatrics; Adjunct Professor, Université Laval, Département des Sciences des Aliments et de Nutrition, Québec, QC
1: Assess the clinical data on lifestyle-based approaches to manage overweight/obesity in the pediatric population
2: Review published trials on pharmacological treatments for overweight/obesity in this population
3: Discuss the clinical management guidelines for obesity proposed by leading Societies/Associations

Obesity Surgery in the Adolescent
Laurent Biertho, MD, Clinical Professor, Department of Surgery, Laval University, Quebec, QC
1: Understand the rationale of obesity surgery in the adolescent
2: Be able to recognize potential candidates for bariatric surgery
3: Understand the risks and benefits of surgery in the morbidly obese adolescent

Theme II: Genetics

Genomic Technologies to Advance Clinical Care in the Pediatric Endocrine Clinic
Kym Boycott, PhD, MD, FRCPC, FCCMG, Clinician Investigator, Children's Hospital of Eastern Ontario; Associate Professor, Department of Pediatrics, University of Ottawa, Ottawa, ON
1: Summarize current and emerging genomic technologies for clinical care
2: Recognize the clinical utility of each technology in the context of the pediatric endocrine clinic
3: Demonstrate when to use and how to interpret these new genetic methodologies

Inherited Syndromes of Reduced Sensitivity to Thyroid Hormone
Samuel Refetoff, MD, Frederick H. Rawson Professor in Medicine; Professor of Pediatrics, The Committees on Genetics and Molecular Medicine; Director of the Endocrinology Laboratory, University of Chicago, Chicago, IL
1: Recognize resistance to thyroid hormone
2: Recognize resistance to thyroid hormone cell transport defects
3: Recognize congenital defects of thyroid hormone metabolism

Theme III: Biochemistry/Ch

Closing the Gaps in Pediatric Laboratory Reference Intervals: The CALIPER Initiative
Khosrow Adeli, PhD, FCACB, DABCC, Head and Professor, Clinical Biochemistry, The Hospital for Sick Children, University of Toronto, Toronto, ON
1: Review the current major gaps in pediatric laboratory analysis and the significant risk this poses to diagnosis and monitoring of pediatric disease
2: Describe the objectives of the CALIPER study and the development of a comprehensive database of pediatric reference intervals to the pediatricians across Canada

3: Discuss the CALIPER data for endocrine hormones and the influence of age, gender, and Tanner age. Discuss the current laboratory issues that complicate interpretation of endocrine test results including lab to lab and method to method variability

Endocrine Epiphanies: Laboratory Testing Dilemmas and Evolving Solutions
Jon Nakamoto, MD, PhD, Laboratory Medical Director, Quest Diagnostics Nichols Institute; Associate Clinical Professor (Voluntary) of Pediatrics, UC San Diego, San Juan Capistrano, CA
1: Explain how "antibody interference" can produce falsely elevated or depressed immunoassay results
2: Recite at least two reasons why assays with the same name can give widely different results
3: Describe in broad terms the difference between "bottom-up" and "top-down" mass spectrometric approaches to measuring peptides and proteins

Theme IV: Critical Care

Adrenal Dysfunction in Pediatric Critical Illness: Differing Perspectives
Kusum Menon, MD, MSc, FRCPC, Associate Professor, Department of Pediatrics, University of Ottawa, Children's Hospital of Eastern Ontario, Ottawa, ON
1: Compare the methods by which intensivists and endocrinologists diagnose adrenal dysfunction in critically ill children
2: Describe the critically ill patient population in which intensivists believe adrenal dysfunction occurs
3: Describe the intensivists approach to management of adrenal dysfunction in the critically ill pediatric population

Thyroid Hormone Deiodination in Injury and Nonthyroidal Illness
Stephen A. Huang, MD, Director, Thyroid Program, Division of Endocrinology, Children's Hospital Boston, Boston, MA
1. Understand the pathophysiology of nonthyroidal illness
2. Understand the concept of local vs. systemic thyroid status
3. Recognize the role of deiodination in tissue injury

Tight Glucose Control in Pediatric Critical Care: Where Do We Stand?
Michael Agus, MD, Pediatric Endocrinologist & Intensivist; Director, Medicine Critical Care Program, Boston Children's Hospital; Assistant Professor of Pediatrics, Harvard Medical School, Boston, MA
1: Understand the evidence for and against controlling glucose in the pediatric ICU
2: Recognize the value of specific components of a glucose control protocol
3: Understand the risks of insulin-induced hypoglycemia

Clinical Debate
Thyroid Nodules: To Remove or Not Remove. That is the Question
(Pro) Celia Rodd, MD, FRCPC, Associate Professor of Pediatrics, McGill University; Associate Director, Division of Endocrinology and Metabolism - MUHC, Montreal Children's Hospital; Program Director, McGill Fellowship Training Program, Pediatric Endocrinology, Montreal, QC
(Con) Johnny Deladoey, MD, PhD, Assistant Professor, Department of Pediatrics and Biochemstry, University of Montreal, Montreal, QC
Common debate learning objectives:
1. Thyroid nodule: question to ask and initial work-up at 1st visit
2. Thyroid US: malignancy criteria, their sensitivities / specificities
3. Utility of FNAC in children: evidence (pro and con)
4. Thyroid nodules in children: epidemiology / risk factors

Biographies

Dr. Khosrow Adeli

Dr. Adeli is currently the head and full professor of Clinical Biochemistry at the Hospital for Sick Children and the Departments of Biochemistry, and Laboratory Medicine & Pathobiology at the University of Toronto in Toronto, Canada. He is the Director of Point of Care Testing program at the Hospital for Sick Children in Toronto. Dr. Adeli is a fellow of the Canadian Academy of Clinical Biochemistry and a diplomate of the American Board of Clinical Biochemistry. Dr. Adeli served as the Editor-in-Chief of the Clinical Biochemistry journal for 7 years (1999-2006). He is an editorial board member of the Clinical Biochemist Reviews. He served (2006-2010) as the President of COMACC , the Commission on Accreditation in Clinical Chemistry, a North American organization responsible for accreditation of clinical chemistry training programs in the USA and Canada. He currently serves as the Vice-Chair of Publications and Communications Division of the International Federation of Clinical Chemistry (IFCC), as well as the Public Relations Coordinator for the IFCC organization.

Dr. Michael Agus

Dr. Michael Agus is pediatric endocrinologist and pediatric intensivist, Director of the Medicine Critical Care Program and Medical Director of the Medicine ICU and Intermediate Care Unit at Boston Children's Hospital. He is an Assistant Professor of Pediatrics at Harvard Medical School. His research focus has been in the realm of endocrinologic disturbances in the critically ill child. Most recently, he completed a large clinical trial of tight glycemic control in cardiac surgical babies in the Cardiac ICU in Boston and Michigan, recently published. He is currently conducting a 2nd large trial in non-cardiac patients in 20 PICUs across the country, and in engaged in clinical research on cerebral edema in diabetic ketoacidosis and on closed loop insulin delivery systems in the ICU and for outpatients.

Dr. Laurent Biertho

Dr. Biertho completed his "candidat" in Biomedical Sciences and Medical training at the University of Liege in Belgium. At the end of his surgical residency in Belgium, he completed a one year Research Fellowship at the Mount Sinai School of Medicine in New York. He then trained in Minimally Invasive Surgery at McMaster University. At the end of his fellowship in 2005, he was appointed Assistant Professor at McMaster University. He was then offered a position as an Associate Professor of Surgery at Laval University in 2006, to develop Minimally Invasive Bariatric and Metabolic Surgery at the Institut Universitaire de Cardiologie et Pneumologie de Quebec, in particular to perform a certain type of bariatric surgery (Duodenal Switch). Currently, the vast majority of the 500 bariatric surgeries performed each year at the IUCPQ use minimally invasive approaches. In addition to developing these clinical activities Dr Biertho contributes to the dissemination and publication of knowledge related to bariatric surgery mostly through clinical studies.

Dr. Kym Boycott

Dr. Kym Boycott is a Clinical Geneticist at the Children's Hospital of Eastern Ontario (CHEO) and Investigator at the CHEO Research Institute. She is an Associate Professor and holds a Tier II Research Chair in Neurogenetics from the Faculty of Medicine at the University of Ottawa. She completed her PhD, MD and FRCPC training in Medical Genetics at the University of Calgary. Dr. Boycott's research, bridging clinical medicine to basic research, is focused on elucidating the molecular pathogenesis of rare inherited diseases using next-generation sequencing approaches. She is the Lead Investigator of the Genome Canada, CIHR funded 'Finding of Rare Disease Genes in Canada' (FORGE Canada) project which is investigating the molecular etiology of almost 200 rare pediatric diseases. Dr. Boycott is also interested in translation of next-generation sequencing approaches to advance diagnostics and patient care.

Ms. Sara Chang

Sara Chang graduated from McMaster University in 2003 with a BScN. She began nursing at Toronto's Hospital for Sick Children in general pediatrics and went on to case management in its metabolic genetics clinic. After moving to Ottawa she worked in pediatric palliative care at Roger's House, and

joined the endocrine team at the Children's Hospital of Eastern Ontario in 2009.

Dr. Cheri Deal
Dr. Deal obtained her PhD (Experimental Medicine), funded by the Medical Research Council of Canada (CIHR), at McGill University, followed by her MD degree at the University of Montreal. She is Canadian and American board certified in Pediatrics (CHU Sainte-Justine/Montreal Children's Hospital) and in Pediatric Endocrinology. She obtained a Medical Research Council Scholarship to pursue a research fellowship in the laboratory of Dr. Ron Rosenfeld at Stanford University in Pediatric Endocrinology from 1989-1991. Dr. Deal has been on staff with the Endocrine Service at CHU Sainte-Justine since 1992, and is tenured Full Professor with the Department of Pediatrics at the University of Montreal as well as an Associate Member of the Faculty of Medicine (Division of Experimental Medicine) of McGill University. Her administrative experience includes the position of past President of the Canadian Pediatric Endocrinology Group (1999-2001), President of the Canadian Society of Endocrinology and Metabolism (2009 – 2011) as well as a member and secretary of the Board of Directors of the Fonds de la recherche en santé du Québec (FRSQ)(1999-2006). She has served as site leader for the Canadian Institutes of Health Research (C.I.H.R.) Canadian Clinical Scientist Training Program in Pediatrics, and is currently Program Director for Pediatric Endocrinology and Metabolism at the University of Montreal.

Her major research interest is in the contribution of the growth hormone-insulin-like growth factor (GH-IGF) axis to the regulation of human growth. One of her laboratory focuses has been the genetic and epigenetic regulation of IGF2. She received support from the Canadian Breast Cancer Research Initiative to study the role of IGF2 and its receptor in breast cancer. Her secondary field of focus concerns the regulation of various other members of the GH-IGF axis in post-natal life, their role in carcinogenesis, within the context of pre-pubertal obesity, and as a target for clinical intervention in children with Turner Syndrome. Dr. Deal has a strong commitment to patient-driven research, and has helped to elucidate the molecular defects associated with a wide range of rare pediatric endocrine disorders including Type 1 polyglandular autoimmune syndrome (APECED) and congenital hypopituitarism and, as well as participated in clinical studies aimed at ameliorating outcomes in a variety of pediatric endocrine diseases such as congential hypothyroidism, precocious puberty, Prader-Willi syndrome and Turner syndrome. She has over 100 publications**.**

Dr. Johnny Deladoey
Dr. Johnny Deladoey is a pediatric endocrinologist at the CHU Sainte-Justine and an investigator at the CHU Sainte-Justine Research Center. He completed his PhD, medical degree and pediatrics residency in Bern, Switzerland and then undertook a 3-year clinical and research fellowship in pediatric endocrinology and diabetes at the CHU Sainte-Justine. He is currently an Assistant Professor of Pediatrics and an Affiliated Professor of Biochemistry at the School of Medicine of the University of Montreal. His research is currently focused on the epidemiology and genetics of congenital hypothyroidism.

Ms. Marie Dorion
1976 to 1979: Neonatal intensive care unit (NICU), and during this period, at times, supervision of nursing students.

1979 to 2008: Assistant head-nurse on a pediatric hematology-oncology unit
1984: Head-nurse on a NICU for one year
2006: six months as nurse navigator on a pediatric oncology unit
2007 to 2009: 2 years as research nurse on a pediatric oncology research unit
2009 - : Ambulatory care in pediatric endocrinology

Ms. Brenda Fraser
Brenda is a pediatric endocrinology nursing case manager at the Children's Hospital Of Eastern Ontario (CHEO). She received her diploma in Nursing in 1989 and completed her degree in 2007 at Charles Sturt University in Australia. She has been at CHEO since 1994, her experience includes a case management position in the Neurooncology service and outpatient Oncology clinic, staff nursing positions with the IV team, and the Emergency dept. She joined the Division of Endocrinology in 2006.

Dr. Stephen A Huang
Stephen A. Huang, MD, obtained his medical degree from the University of Pennsylvania School of Medicine. Following his residency training in pediatrics at Harvard Medical School, Dr. Huang completed pediatric endocrinology and thyroidology fellowships at Children's Hospital Boston and the Brigham and Women's Hospital, respectively. Dr. Huang is involved in both clinical practice and laboratory research, with a special emphasis on pediatric thyroidology. Dr. Huang serves as the director of the Thyroid Program at Children's Hospital Boston and director of thyroid research in the hospital's Division of Endocrinology. Dr. Huang's research is in the field of thyroid hormone metabolism and relates to the modulation of local thyroid status during injury. Because of clinical interests in the long-term care of childhood thyroid cancer, he maintains active appointments in the Brigham and Women's Hospital and the Dana Farber Cancer Institute.

Ms. Peggy Kalancha
Peggy is a graduate from the University of Saskatchewan college of Nursing with a Bachelor of Science in Nursing. She has worked in pediatrics at the Alberta Children's Hospital for a number of years as a Clinical Resource Nurse, specializing in the areas of Perinatology, Develomental Clinic, Genetics, Neuromuscular and Neurology Clinics, Gastrointestinal and Rheumatology Clinics. The past 4 years she has been working in the areas of Endocrinology and Gynecology where she has found a happy home!

Ms. Nicole Kirouac
Nicole is a graduate from the University Of Manitoba Faculty Of Nursing Baccalaureate Program as well as the Pediatric Endocrinology Nursing Society Research Fellowship Program. She has been the Pediatric Endocrinology Nurse Clinician at Winnipeg Health Sciences Centre for the past 15 years. Nicole has been a leader in her field as current director and past President of the Canadian Pediatric Endocrinology Nurses Group and past chairperson of the Evidence Based Nursing Steering and Nurse Clinician committees in her centre. Nicole has also been a volunteer Nurse Coordinator for camps for Children with Diabetes and Cancer for over 20 years. She has helped plan and execute various family information sessions for her patients as well as provided ongoing education for Nurses around Pediatric Endocrine Emergencies. Nicole has recently presented a poster on Bone Health in Children at the Pediatric Endocrinology Nursing Society meeting in Florida as well as the Joint International and European Congress in Endocrinology (ICE/ECE) meeting in Florence, Italy.

Dr. Kusum Menon
Dr. Kusum Menon graduated from the University of British Columbia, Vancouver, British Columbia in 1989 and received her training in Pediatrics and Pediatric Critical Care at the Universities of Ottawa and British Columbia in 1994 and 1996 respectively. She then completed a research year at the University of Western Ontario from 1996-97 and her Masters in Epidemiology at the University of Western Ontario in 2000. She is currently an Associate Professor of Pediatrics at the University of Ottawa and a Senior Investigator at the Children's Hospital of Eastern Ontario Research Institute. She sits on the Executive of the Canadian Critical Care Trials Group. She has won numerous awards for research mentorship, and her areas of interest include adrenal insufficiency in pediatric critical care as well as the consent process in pediatric critical care research. She is funded by the Canadian Institutes of Health Research for work in these areas.

Ms. Lina Moisan
Graduated from Université of Montréal in 1980

Work experiences:
Hotel Dieu de Montreal
Centre Hospitalier Universitaire Vaudois à Lausanne

Montreal Children's Hospital since 1982 in different positions (ward nurse, assistant head-nurse and acting head nurse)

In the present position since 1997; in endocrinology nursing and other activities part of the Clinical Investigation Unit.

She is a member of CPEG and CPEN. She is also a member of AIPI.

Dr. Jon Nakamoto
Dr. Nakamoto (M.D., Yale; Ph.D. in Biological Chemistry, UCLA) completed residency training in pediatrics at the University of California, San Francisco, and endocrinology fellowship at UCLA under the auspices of a Physician Scientist Award from the National Institutes of Health. After joining the Nichols Institute laboratory in 2002 as an Endocrine Medical Director, he served as Managing Director of the Nichols Institute from 2003-2009 and now serves as the general Laboratory Medical Director. His interests include new assay development, informatics, and ways to increase collaboration between clinicians and the clinical laboratory. He is an active member of the Endocrine Society, Pediatric Endocrine Society, American Association of Clinical Chemistry, and is incoming Chair of the Pediatric Endocrinology Sub-board of the American Board of Pediatrics. Dr. Nakamoto is certified in pediatric endocrinology and maintains an appointment as Associate Clinical Professor (Voluntary) of Pediatrics and Endocrinology at the University of California, San Diego.

Dr. Samuel Refetoff
Samuel Refetoff is known for the 1967 discovery of resistance to thyroid hormone, also known as Refetoff syndrome, and elucidation of its genetic and molecular basis (1989/92);. His work has guided investigation of the mechanism of thyroid hormone action and serves as model for characterization of other nuclear hormone receptor-mediated diseases. Devoted to the study of inherited thyroid disorders, his laboratory first identified mutations in: serum thyroid hormone transport proteins [TBG (1989) and albumin

(1994)]; the thyrotropin receptor producing resistance to TSH (1995); and two defects combining neuropsychological and thyroid abnormalities caused by mutations in the TTF1 (2002) and MCT8 (2004) genes. His laboratory identified a defect of thyroid hormone metabolism caused by mutations in the SBP2 gene (2005). He has elucidated two long-sought regulatory mechanisms: that required for synthesis of a biologically active dual oxidase (2006) and the other controlling thyroid hormone secretion (2010).

Dr. Celia Jane Rodd
Celia Rodd is currently an Associate Professor in Pediatric Endocrinology and in the School of Dietetics and Human Nutrition at McGill University. She completed her medical degree at the University of Toronto, her pediatric residency at the Montreal Children's Hospital, and then undertook a year of clinical pediatric endocrine training at the Hospital for Sick Children in Toronto. Celia subsequently completed a research fellowship with Dr. Jack Oppenheimer at the University of Minnesota, exploring the ontogeny of thyroid hormone receptors. She returned to Montreal in 1993 to join the McGill faculty. Celia recently completed her Masters in Epidemiology from the London School of Public Hygiene and Tropical Medicine in 2010.

Celia's research interest include vitamin D adequacy at the individual and population level, bone health issues in children with chronic diseases and thyroid conditions.

Dr. John Weisnagel
Dr. S. John Weisnagel, M.D., FRCPC, is an endocrinologist who practices at Laval University Health Center (CHU) de Québec both in adult and pediatric endocrinology He directly follows children and adolescents in the diabetes and endocrinology clinics at the Centre Mère Enfant. He is also a researcher at the Centre de recherche du CHU de Québec and is an adjunct professor in the Department of Food Science and Nutrition, Université Laval. He has extensive clinical experience working both with type 1 and 2 diabetes, obesity and metabolic complications related to diabetes. He is recognized by his peers for his expertise in glucose metabolism and factors that influence it's regulation, particularly insulin sensitivity and secretion, glucose tolerance, nutrition, physical activity and obesity. He is a principal investigator or co-investigator in many studies funded by public organisations and the pharmaceutical industry. He is the author and co-author of more than 100 publications and supervised many graduate students.

Disclosure of Conflict of Interest

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Oral Abstracts

Oral Abstract 1

Predictors of BMI Reduction in the SickKids Team Obesity Management Program (STOMP) for Adolescents with Severe Obesity

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Department of Paediatrics[1], Division of Endocrinology[2], Department of Psychology[3], Division of Paediatric Medicine[4], The Hospital for Sick Children, University of Toronto, Toronto, ON

Background/Aims: Combined behavioural lifestyle programs have been shown to be effective in treating youth who are overweight, however, patients' responses to a given intervention vary significantly. Our aim was to compare baseline characteristics of adolescents with severe obesity who reduced versus increased their BMI after 6 months in the STOMP program and to identify predictors of BMI reduction.

Methods: We divided the patients into two groups: those who reduced their BMI and those who increased their BMI after 6 months. Independent t-tests and chi-square/Fisher exact tests were used to compare baseline anthropometric, metabolic, and psychological characteristics. For the entire study population, Pearson correlation coefficients were calculated between change in BMI and baseline variables. Significant variables were entered into a multi-linear regression model with change in BMI as the outcome.

Results: There were no differences at baseline between adolescents who reduced versus increased their BMI at 6 months. A greater reduction in BMI at 6 months correlated with older age, lower weight-specific quality of life (QOL) measurements (indicating a poorer quality of life), and a higher teen Readiness-to-Change (RTC) score. The strongest predictors of BMI change were baseline scores on the Impact-of-Weight-on-Quality-of-Life-Kids Physical Comfort questionnaire ($t=3.5$, $p=0.001$) and baseline BMI ($t=3.0$, $p=0.004$).

Conclusions: Although older age, increased RTC and a greater impairment of weight-related QOL at baseline correlated with a BMI reduction after 6 months, only lower QOL (specifically physical comfort) and lower initial BMI remained as predictive factors for BMI reduction in multivariate analysis. Assessment of QOL should be part of the intake process of paediatric obesity programs and may guide the intervention and treatment goals.

Oral Abstract 2

Phenotypic variability in familial GLI2 deletion

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Ste Justine, Université de Montréal, Montréal, QC

Introduction: Diminished Sonic Hedgehog (SHH) signalling is associated with the most common forebrain defect in humans, holoprosencephaly (HPE). The SHH protein acts as a crucial factor that patterns the ventral forebrain and is required for the division of the primordial eye field and brain into two discrete halves. GLI2 is a transcription factor implicated in SHH signal transduction. Loss-of-function mutations in the human GLI2 gene are associated with a distinctive phenotype (within the HPE spectrum), whose primary features range from defective anterior pituitary formation and panhypopituitarism, with or without overt forebrain cleavage abnormalities and HPE-like midfacial hypoplasia, to milder presentations with pituitary involvement.

Case report: We report a family with four members having a microdeletion encompassing GLI2 on chromosome 2 (2q14.2). The index case is a 2-year old girl, known for several congenital malformations (atrial septal defect, interruption of the inferior vena cava, situs ambiguous, polysplenia, postaxial polydactyly, cleft lip and palate). Using Comparative Genomic Hybridization and FISH, a microdeletion encompassing GLI2 was found. Cerebral MRI showed an ectopic posterior pituitary and a hypoplastic anterior pituitary. Because of these IRM findings, she was referred for endocrinological assessment. Initial IGF-1 was low (5.2 nmol/L; normal range 7,0-42,0 nmol/l), thyroid function tests and morning cortisol were normal; the growth hormone response to a stimulation test with arginine was low (1.38 µg/L). The father and the two brothers (aged 3.5 yrs and 10 months) of the index case carry the same microdeletion (confirmed by FISH), which confirms dominant transmission from the father. The older brother has normal growth but mild hypertelorism; he has a normal cortisol response to stimulation with 1 mcg ACTH (1-24), normal IGF-1 and normal thyroid function tests. The younger brother has normal growth, no dysmorphic feature and no symptom suggesting hormone deficits. Cerebral MRIs have been scheduled for both brothers and the father's endocrine functions are currently assessed.

Conclusion: We report a family with three sibs, who have a GLI2 deletion transmitted from their father. These cases present a wide phenotypic variability, confirming the incomplete penetrance of deletions of the GLI2 gene.

Oral Abstract 3

Congenital adrenal hyperplasia due to 11β-hydroxylase deficiency in a 5-year-old boy

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A 5-year 4-month old Caucasian boy presented initially to the Dermatologist for a one-year history of body odour, acne on the ears and cheeks, and rapid growth over the last 6 months. Work-up by the dermatologist revealed elevated 17-hydroxyprogesterone (17-OHP) at 14.2 nmol/L, performed by immunoassay at a commercial laboratory.

On physical exam at our centre, the patient was normotensive. Other than the mild acne, height was at 120.7 cm (97%ile, +1.9 SD for age) and the patient had increased penile length at 7 cm (>2 SD), with absence of pubic or axillary hair.

The bone age was advanced at 9 1/2 years. Biochemical analysis of specific adrenal steroids by tandem mass spectrometry was recently initiated at our centre on baseline and cosyntropin-stimulated plasma. To our surprise, basal 17-OHP was 2.5 nmol/L, increasing to 15.5 nmol/L at 60 min. The basal 11-deoxycortisol was 63.5 nmol/L (normal, 0.2–6.0) and increased to 351.4 nmol/L (normal 2.8–10.4) at 60 min. Baseline samples revealed mildly elevated testosterone (1.6 nmol/L), androstenedione (8.5 nmol/L) and DHEAS (2.8 μmol/L).The high-dose cosyntropin stimulation test also demonstrated a suboptimal cortisol response (194 nmol/L at baseline, 345 nmol/L at 60 min).

The patient was started on hydrocortisone replacement, and the molecular genetic analysis of the *CYP11B1* gene is pending.

This case highlights the advantageous use of tandem mass spectrometry for rapid detection and quantification of steroids related to rare forms of congenital adrenal hyperplasia beyond its use for newborn screening for this condition.

Oral Abstract 4

hCG induced thyrotoxicosis in a 14 year old girl

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A previously healthy 14 year old girl was admitted for elective termination of pregnancy. She reported a single incident of sexual activity two months prior to the positive pregnancy test. An ultrasound, in conjunction with high hCG levels, lead to the diagnosis of a molar pregnancy. However, a repeat ultrasound demonstrated a mass superior to the uterus. She was noted to be tachycardic, raising a concern for thyrotoxicosis.

Retrospectively, the patient reported palpitations, heat intolerance and weight loss of 20 lbs over the last month. Treatment was started with Lugol's iodine solution, Tapazole and Atenolol. Laboratory studies demonstrated TSH < 0.02 mU/L, fT4 51 pmol/L and fT3 14.9 pmol/L. hCG was 630,000 IU/L. MRI scan demonstrated a large mass, arising from the left ovary and pathology was compatible with a choriocarcinoma. PET CT scan demonstrated metastases to lungs and spine. Chemotherapy was started and hCG levels fell to 680 IU/L within a week. Treatment with Tapazole was required for 4 months until tumor resection was performed, due to residual hCG secretion. hCG secreting gynecologic tumors include gestational trophoblastic disease and some of the primary germ cell ovarian tumors. The mechanism of thyrotoxicosis is through elevated hCG levels, stimulating the thyroid gland's TSH receptor. Our patient presented with a primary ovarian choriocarcinoma, which is highly suspicious for nongestational origin. There are approximately 60 published cases of non gestational choriocarcinoma (NGCO) and the mean age of patients is 13-14 years. To date, thyrotoxicosis has not been reported in any female patient with NGCO. In gestational choriocarcinoma, the youngest thyrotoxic patient reported was 17 years of age. Our patient is therefore the youngest with a choriocarcinoma to develop thyrotoxicosis and her case illustrates the diagnostic and therapeutic challenges associated with this rare condition.

Oral Abstract 5

Post-operative hypocalcemia - A case series and proposed approach to care following thyroidectomy

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Introduction: Total thyroidectomy is the standard of care for suspected thyroid carcinoma and is an option for definitive treatment of Grave's disease in children. Hypocalcaemia (HypoCa) due to hypoparathyroidism is a potential complication. Rates of HypoCa range between 0.3-49% in the published literature; it most commonly presents within 2 days of surgery. Approaches to monitoring and treating post-op HypoCa vary. Biomarkers predictive of HypoCa are poorly described in pediatrics. Currently a uniform approach to the management of post-op HypoCa in the pediatric population is lacking, which may lead to confusion among team members, delays in treatment and prolongation of stay. As part of a QI initiative we performed this analysis to gain insight into rates of HypoCa and outcomes at our institution, and to devise a consistent approach.

Methods: A retrospective review determined rates and predictors of HypoCa post-thyroidectomy at our institution over a 6 year period.

Results: From 2005-2011 there were 34 patients who underwent total/completion thyroidectomy. Post-operatively 14 had mild HypoCa, and 8 had moderate to severe HypoCa. Four patients (11.8%) required IV calcium (Ca) infusions and 9 (26.5%) were discharged on Ca supplements ± calcitriol. Three patients (8.8%) developed permanent hypoparathyroidism. Patients who required Ca supplementation had significantly more Ca checks (12.9 vs. 6.2, $p<0.01$) and there was a trend towards a longer length of stay (3.89d vs. 3.03d, $p=0.06$) compared to patients who did not require supplementation. Eight (23.5%) patients received written information on symptoms of HypoCa. Of the 9 patients discharged on Ca supplements 3 received written instructions on the treatment.

Conclusions: Together with the surgical team, nurses, laboratory and pharmacists we developed a protocol incorporating risk stratification based on Ca and PTH for post-thyroidectomy Ca monitoring and intervention. Our aims are to streamline our approach, optimize communication between team members, prevent episodes of HypoCa, decrease length of stay, and to consistently provide written information on HypoCa and treatment to patients and families at discharge. We will re-evaluate this protocol at 6-12 months.

Oral Abstract 6

Impact of elective hospital admission on glycemic control in adolescents with poorly controlled type 1 diabetes

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Objective: In order to prevent the risk of acute and long term complications, different treatment strategies have been used to manage adolescents with poorly controlled type 1 diabetes. We investigated whether a brief elective hospital admission improves HbA1c over 12 months.

Methods: We studied a retrospective cohort of hospitalized adolescents with poorly controlled type 1 diabetes attending a diabetes clinic in a tertiary care pediatric hospital in Montreal, Canada between January 2005 and December 2010. Hospitalized adolescents (admitted group) were matched with control adolescents (non-admitted group) according to age and baseline HbA1c. HbA1c values at baseline, 6 and 12 months were obtained from the clinic database.

Results: Thirty patients aged 12 to 17 years with a first elective admission for poor metabolic control were paired with thirty non-admitted patients. At baseline, HbA1c was 12.2 ± 1.6% in admitted and 12.0 ± 1.2% in non-admitted patients. There was no improvement in the primary outcome as assessed by the change in HbA1c at 12 months in the admitted group (-1.3% ± 2.3) compared with the non-admitted group (-2.1% ± 1.7). No improvement in intermediary measures of glycemic control was observed (HbA1C at 6 months or change at 6 months). After 12 months, HbA1c values were higher in the admitted group (10.9 ± 1.9%) versus the non-admitted group (9.9 ± 1.4%, p = 0.016).

Conclusions: The study indicates that elective hospital admission for adolescents with poorly controlled type 1 diabetes does not seem to be an effective strategy to improve HbA1c over 12 months.

Oral Abstract 7

A short 17-year old with primary amenorrhea: when there is more than Turner syndrome

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We present the case of a 17-year-old girl, who was referred to our pediatric endocrinology clinic at the age of 12 for short stature. She was known for pes equinovarus and multiple ear infections, and had been operated for bilateral inguinal hernias. Family history was negative for endocrinopathies. She presented spontaneous thelarche. Her physical examination showed a high-arched palate, bilateral cubitus valgus, wide-spaced nipples, and a Tanner stage M2P1. A 46 XX/Del(X)(p11.23p22.1) karyotype confirmed the suspected Turner syndrome. She had neither thyroiditis nor heart or renal anomalies and her serum FSH and LH concentrations were normal (5.33 UI/L and 1.57 UI/L). Although she progressed spontaneously through puberty to Tanner stage M5P5, she had had no menarche at the age of 17. The report of a pelvic ultrasound done at age 16 mentioned a small unstimulated tubular uterus and two normal-sized ovaries with follicles. FSH and LH levels remained normal, estradiol was 900 pmol/L, and AMH was 1.43 ng/L (N 2.00-6.80 ng/L). A gynecological examination revealed a blind vaginal pouch. The upper third of the vagina and the uterus were not seen on MRI. Thus, our patient was diagnosed with Rokitansky syndrome as well. Rokitansky syndrome (OMIM #277000) originates from the abnormal regression of the Müllerian ducts in females with a normal 46,XX karyotype. Some cases have been shown to be associated with WNT4 mutation (OMIM #158330), SHOX (Xp22) duplication (Sandbacka 2011) chromosomal imbalance at 1q21.1, 16p11.2, 22q12.2 (Cheroki 2008) and Xp21.31. To this day, the only case of a girl with 45 X/46 XX Turner syndrome and Rokitansky syndrome was published in 1987. With our patient's consent, we will try to obtain a more detailed genetic analysis, including array comparative genomic hybridization, which may help to identify candidate genes for this exceptional combination of syndromes.

Oral Abstract 8

A novel heterozygous mutation in the glucokinase gene (GCK) with exercise-induced symptomatic hyperglycemia responsive to sulfonylurea

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Background: Inactivating heterozygous mutations in the glucokinase gene (GCK) are usually associated with asymptomatic, non progressive, mild elevations of fasting glucose that do not require treatment.

Objective: To describe the atypical phenotype and genotype of an adolescent girl with a novel GCK mutation.

Design/methods: Chart review, gene sequencing, and blinded continuous glucose monitoring (CGM with iPro2) were used to characterise the case.

Results: A 14 year old elite-level competitive athlete developed shakiness, tremors and extreme fatigue with mild hyperglycemia (6.8-13 mmol/L). Symptoms and hyperglycemia were exacerbated with exercise. The severity of symptoms required her to cease competitive sports. Family history was positive for type 2 diabetes in father, paternal uncle and grandmother. Investigations: A1C 6.7% (nondiabetic range 4-6.2%). Fasting venous BG 6.8 mmol/l; 2 hour OGTT glucose 7.8 mmol/L. Negative anti GAD and islet cell antibodies. Normal fasting insulin and c-peptide. HNF1A and HNF4A mutation analyses were negative. A novel heterozygous mutation p.Q219x (c.655C>T) in exon 6 of GCK was confirmed in the patient and father. Clinical Course: Initiation of Gliclazide 20 mg bid was associated with resolution of symptoms and normalization of A1C (5.6%). Patient reported additional beneficial effect of starting high protein and low carbohydrate diet, and returned to competitive sports. Blinded CGM demonstrated significantly less time spent in the hyperglycemic range (sensor glucose (SG) > 8.0 mmol/L) when on twice daily Gliclazide vs intermittent or no Gliclazide (mean minutes/day > 8 mmol/L: 53.6 +/- 90.0 vs 307.9 +/- 246.6; p=0.02).

Conclusions: This novel mutation in the GCK gene led to atypical symptomatic exercise-induced hyperglycemia that was responsive to low dose sulfonylurea with self-reported additional benefit after reduction of carbohydrate intake. We postulate that her atypical clinical presentation was related to the intense elite-level physical activity combined with carbohydrate loading before exercise.

Oral Abstract 9

Growth Failure in Hunter Syndrome - A Contrasting Tale of Two Patients

MUNIER A NOUR, DAVID K STEPHURE, ANEAL KHAN
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Background: Hunter syndrome is an X-linked mucopolysaccharide storage (MPS) disease due to deficiency of iduronate 2-sulfatase activity caused by mutations in the IDS gene. The presentation typically occurs in early childhood with short stature, coarse facial features, joint contractures, hepatosplenomegaly, valvular heart defects, and - in some children - developmental delay.
We contrast two cases of Hunter syndrome evaluated for growth failure following discovery of abnormal pituitary anatomy.

Case Presentations: Patient 1, a 10 year old boy with Hunter syndrome (MPS type IIB), presented for evaluation of impaired growth following MRI identification of a J-shaped sella and an ectopic posterior pituitary gland. Comprehensive functional endocrine testing revealed growth hormone deficiency, secondary adrenal insufficiency, and tertiary hypothyroidism. He was treated with recombinant human growth hormone with subsequent improvement in growth velocity (3.7 cm/year (-2 S.D.) pretreatment to 9.3 cm/year (+5.6 S.D.) over the next 12 months).

Patient 2, a 13 year old also diagnosed with Hunter syndrome, was evaluated for growth failure. He was identified to have a large empty sella with posteriorly displaced pituitary gland. Complete functional endocrine testing revealed normal pituitary functioning and growth hormone therapy was not instituted.

Conclusions: We present 2 patients with Hunter syndrome and abnormal pituitary imaging. Patient 1 was found to have anterior panhypopituitarism while patient 2 had intact pituitary function. To our knowledge, these anatomical anomalies of the pituitary gland have not been previously associated with anterior panhypopituitarism in Hunter Syndrome. We conclude that anterior panhypopituitarism associated with abnormal neuroradiologic imaging of the sella and suprasellar areas may occur in some patients with Hunter syndrome. Further investigation in a larger sample is required to determine the frequency of this association in Hunter syndrome.

Oral Abstract 10

Iliac bone histomorphometry in children on glucocorticoids for the treatment of rheumatic disorders

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Department of Endocrinology[1], Rheumatology[2], Hospital for Sick Children, Samuel Lunenfeld Research Institute[3], University of Toronto, ON

Context: Rheumatological conditions are associated with an increased fracture risk. The tissue level characteristics of the bone involvement in children have not been well elucidated.

Objective: To describe the bone micro-architectural characteristics as measured by histomorphometry, from a cohort of children with rheumatological disorders treated with glucocorticoids, and to determine associations between micro-architectural findings with clinical and radiological variables.

Results: 15 children (14.0±3.2 years) with either systemic lupus erythematosis or idiopathic juvenile arthritis had a trans-iliac bone biopsy as part of an assessment for symptomatic osteoporosis. Mean duration of glucocorticoid exposure was 6.2±4.1 years with a cumulative prednisone dose of 0.32±0.23 mg/kg/day in the 12 months prior to the biopsy. All were on calcium and vitamin D supplementation. 25-hydroxy vitamin D at time of biopsy was 73.2±21.0 nmol/L and median number of vertebral compression fractures since glucocorticoid commencement was 3.1(1 to 12).

Histomorphological analysis demonstrated significant decrease in trabecular number ($p=0.01$), and increase in trabecular separation ($p=0.04$) and osteoid thickness ($p=0.01$) compared to published age, sex-matched normative data. Severity of the trabecular deficit correlated to cumulative glucocorticoid dose ($p=0.02$) and growth velocity ($p<0.01$), but not glucocorticoid treatment duration, or erythrocyte sedimentation rate. While there were no correlation with lumbar-spine BMD Z score (mean Z score -3.4±1.3) at time of biopsy, change in BMD over the preceding year correlated with bone volume on histomorphometry ($p=0.04$).

Conclusion: Impairments of bone micro-architecture relate to cumulative glucocorticoid dose in children with rheumatic conditions. Ongoing analysis of bone quality and architectural parameters will allow further insight into the mechanisms underlying these changes.

Oral Abstract 11

Type 1 Diabetes and Celiac Disease: Associated Comorbidities and Complications

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Objective: To evaluate the rates and outcome of complication screening in a population with type 1 Diabetes Mellitus (T1DM) and Celiac disease (CD).

Study Design: We retrospectively reviewed the health charts of 42 children with T1DM and biopsy proven CD diagnosed between 2005 and 2011 to evaluate associated comorbidities, changes in metabolic control and anthropomorphics before and after diagnosis of CD, in addition to frequency and results of complication screening within 2 years from CD diagnosis.

Results: CD was observed in 5% of established patients with T1DM. Mean age at the time of CD diagnosis was 8.82 + 3.4 years (range 3-16 yr) with a male: female ratio of 0.68. Autoimmune thyroid disease was observed in 12.5%. Family history was positive in 22.5%. Symptoms were present at 47.5%. Good to excellent adherence to a gluten-free diet (GFD) was observed in 69% of patients. Metabolic control before and 6 months post diagnosis revealed an increase in HbA1c by 4.97% in the adherent group versus a decrease by 5.45% in the non adherent group ($P<0.05$). A trend towards lower LDL levels was seen in the adherent group (Mean= 2.34 vs 2.77 mmol/L, $P=0.06$). No differences were observed in height, weight and BMI z scores assessed at diagnosis and 1 year later. Complication for anemia and liver function testing was tested for 29/41 & 15/41 children respectively with abnormalities observed in 2 patients. Vitamin D status and bone mineral density was available for 10/41 & 4/42 children respectively with >50% having a VitD level of <70nmol/L.

Conclusions: In patients with T1DM & CD, we observed significant rates of comorbid thyroid autoimmunity and CD history in families. Discordant changes in HbA1c were observed 6 months post diagnosis on the basis of GFD adherence. Complication assessment was variable and negative for the majority of the patients. Rates of bone health assessment were suboptimal.

Oral Abstract 12

Pde11a and adrenal disease. Characterization of a genetically manipulated Pde11a-/- mouse model

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Phosphodiesterases catalyze the hydrolysis of cyclic nucleotides and maintain physiologic levels of intracellular concentrations of cAMP and cGMP. Increased cAMP signaling has been associated with genetics disorders that lead to adrenocortical tumors and Cushing syndrome. Genetics defects in the phosphodiesterase 11A (PDE11A) are associated with increased levels of cAMP and bilateral adrenal hyperplasia; they also contribute to the development of adrenocortical, prostate, and testicular tumors in the general population. The aim of the present work is to characterize the genetically manipulated Pde11a mice and study the physiologic, cellular and molecular effect in the adrenal glands. A colony of Pde11a-/- (KO), Pde11a+/- (Het) and wild type (WT) mice was studied in a complete phenotypical gross and microscopy pathological study. Molecular, immunohistochemical and biochemical studies were done to determine the function and expression of Pde11a in the tissues of normal and mutant mice.

Pde11a RNA and protein expression is reduced but still present in Pde11a-/- mice. Although Pde11a activity is reduced and cAMP levels are higher in adrenals and testes of Pde11a-/- mice, Protein Kinase A (PKA) activities is not different in both genotypes. Although adrenal corticosterone levels are similar in both genotypes, low-dose dexamethasone do not significantly suppress adrenal corticosterone secretion in Pde11a-/- mice. Pde11a knockout mice show adrenal subcapsular hyperplasia and a zonation defect phenotypes while foamy cells are more frequent in wild type animals. Eosinophilic cells emerging from the innermost area (X-Zone) of the cortex is predominant in female Pde11a-/- adrenals when compared with the wild type. Other interesting phenotype is related to the body weight. Mice were consistently leaner than Pde11a+/- (29.03± 5.35 gr, $p=0.03$) and Pde11a+/+ (31.21 ± 6.36 gr, $p=0.04$). In addition, defect in Pde11a is associated with reduced deposits of fat in liver. In Summary, these data support the involvement of PDE11A in the pathogenesis of adrenocortical hyperplasia, the role of the cAMP pathway in steroidogenesis and metabolism, but also provide significant insight into a previous reported mouse model.

Poster Abstracts

Poster Abstract 1

Discordance between perceived and measured glycemic control in young adolescent males with type 1 diabetes

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Objective: To compare subjective perception of glycemic control with objective measure of glycated hemoglobin (A1c) in adolescents with type 1 diabetes (T1D), and identify discrepancies that could be targeted in interventions designed to improve outcomes.

Methods: From October 2011 to January 2012, 6-18 year old children with T1D or their parents attending diabetes clinic at the Hospital for Sick Children completed a questionnaire, to rate how they perceived the patient's current diabetes control. A1c measured at the clinic visit and clinical variables were also recorded.

Results: Mean A1c of the adolescents was 8.9±1.5% (n=177) compared to 8.5±1.1% (n=88) in the 6-11 age group (p=0.02). Early adolescence was associated with an increase in A1c in boys, but not girls (12-15 year olds: boys 9.4±1.5% v.s. girls 8.6±1.5%, p=0.005). Elevation of A1c in the young adolescent boys was greater in those where the questionnaire was completed by the patient, rather than the parent (p=0.01).
Overall in adolescents, lower A1c was associated with better perceived diabetes control (r=-0.52, p<0.001). However in the 12-15 year old boys, where the adolescent completed the questionnaire, there was less concordance: 45.5% of them rated their control better than it was ("over-raters"), compared to 26.7% of other adolescent respondents (p=0.04). In these boys, a greater misperception of diabetes control was associated with higher A1c ("over-raters" A1c 9.9±0.8% v.s. "non over-raters" 9.1±1.7%, p=0.04).

Conclusions: In our clinic, young adolescent males have both the highest A1c levels and greatest discordance between perceived and measured glycemic control. Interventions designed to improve education around A1c targets and goal setting within this group may improve clinical outcomes. Moreover, A1c levels were highest and impressions of glycemic control most inaccurate in cases where the questionnaire was completed by the adolescent male rather than the parent. Further investigation is needed to determine whether this suggests that less parental involvement in diabetes care within these cases is a causal factor, a potential additional area of intervention.

Poster Abstract 2

Process Evaluation of the Living Green, Healthy and Thrifty (LiGHT) Virtual Child Obesity Management Program: Combining Health Promotion with Ecology and Economics

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Insufficient access and high attrition rates are affecting the outcomes of pediatric weight management programs. Here, we describe Phase 1 (program development and process evaluation) of the LiGHT project, which aims to help youth and their families adopt a healthy lifestyle. The two novel aspects of LiGHT are: 1. web-based concept to reduce geographic barriers to accessibility; 2. demonstration of the environmental and personal financial benefits of a healthy lifestyle. The curriculum consists of 11 modules designed by a multi-disciplinary team of experts. Each module contains a questionnaire on personal habits, information on the topic from health, environmental and financial perspectives, homework, and practical goals to be achieved within a month. Process evaluation integrated feedback from focus groups through the 12 months of program development. A multi-phase qualitative data collection was conducted with 12 families to inform program development. 17 families participated in 4-week program pilot and provided qualitative and quantitative feedback in the form of knowledge tests and program evaluation. Transcripts and written evaluations were reviewed and coded for themes by two independent reviewers. Responses to knowledge surveys and multiple-choice questions were evaluated using descriptive statistics. Interviews and the focus group revealed that electronic communication, site legitimacy, child and youth engagement, a sense of community, and cultural relevance and financial feasibility of nutrition recommendations are essential for program participation. Participants wanted for the program to address psychological issues related to obesity. Program strengths included its online format, multidisciplinary approach to weight management, level of support, facilitation of parent-child communication, and feasibility of suggestions for lifestyle changes. Website design, program duration, complexity of information and online community support were indicated as areas for program growth. Phase 2 of the project will focus on implementation and outcome evaluation.

Poster Abstract 3

Efficacy of Finasteride for Treatment of Hidradenitis Suppurativa: A case series in the pediatric population

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Background: Hidradenitis suppurativa (HS) is a chronic, recurrent inflammatory disease of the apocrine gland bearing areas including axillae, perineum, and inframammary regions. The cause of HS is largely unknown but may relate to altered 5α reductase sensitivity at the follicular epithelium. Finasteride, a selective inhibitor of type II 5α reductase, has been found to be effective in small case series in treating adults with refractory HS. To date, finasteride as a therapy for HS has not been reported in the pediatric age group. We present three females patients with refractory HS who were treated successfully with finasteride after failing to achieve clinical remission while on other medical and or surgical therapies. Patient 1 had extensive HS lesions overlying her axillae, flanks and groins that were noted at age 7. She had partial improvement reported while on oral antibiotic, isotretinoin, oral contraceptive (OCP), and spironolactone. She had no improvement with photodynamic therapy. Localized surgical resection of the lesion was partially beneficial in the short term, but did not alter the frequent flare up of the disease. She was started on oral finasteride 5 mg daily in combination with OCP which resulted in markedly fewer and less severe flares. She has remained on finasteride for five years. Patient 2, a 15 year with PCOS who had severe HS lesions in the groin region and on the posterior neck required surgical drainage and intravenous antibiotics. She had demonstrated poor therapeutic response while on isotretinoin and OCP. A trial of triple therapy with daily oral erythromycin, OCP and oral finasteride was very effective in decreasing the frequency and severity of her flares over the subsequent two and a half years. Patient 3 was a prepubertal, 7 year old, girl with significant recurrent perianal, inner thigh and axillary HS. She had frequents flares despite being compliant with oral and topical antibiotics. Oral finasteride was initiated in combination with oral trimethoprim. She had remarkable improvement with minimal flares subsequently. She has continued on this treatment for three years.

Conclusion: Finasteride was very effective in controlling the HS and reducing disease flares in our patients during the 2-5 year length follow up period. It was tolerated well with no significant side effects. Further prospective and randomized controlled studies are needed to determine optimal candidates and to provide additional safety and efficacy data.

Poster Abstract 4

A case of hypothalamic hamartoma associated with central precocious puberty and growth hormone deficiency

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Introduction: It is known that hypothalamic hamartomas (HHs) are associated with central precocious puberty (CPP). The pathophysiology of the hypothalamic-pituitary-gonadal axis activation is not perfectly understood. When the endocrine system is involved in HHs, it is most often with excess gonadotropin secretion or, in rare cases, excess growth hormone secretion. Here, we present a case of HH associated with CPP and growth hormone deficiency (GHD).

Case: A 14 3/12 year-old girl was referred for evaluation of short stature and primary amenorrhea. She was followed previously in another center for idiopathic CPP and was treated with a gonadotropin-releasing hormone analogue (GnRH) between the ages of 6-4/12 and 9-9/12 years. On examination, her height was well below the 3rd percentile and her weight was above the 97th percentile. She had acanthosis nigricans on the nape of her neck. Pubertal assessment revealed Tanner 5 estrogenized breasts. Vaginal mucosa appeared estrogenized. She had no acne or hirsutism. Review of the patient growth curve showed that she never had a growth spurt, even after cessation of the GnRH agonist. Baseline hormone levels, including thyroid function, prolactin and androgen levels, were normal. She subsequently failed two GH stimulation tests, with peak GH levels of 0.5 µg/L and 2.8 µg/L after clonidine and L-arginine respectively (normal > 3.4 µg/L). MRI of the brain was done (even though the original CT scan was negative) and revealed a 5 mm hypothalamic hamartoma posterior to the pituitary stalk and attached by a small pedicle to the hypothalamus. A provera challenge was done, at which point she had a positive withdrawal bleed.

Discussion: Central precocious puberty is often a manifestation of HHs. Without treatment, there is a risk of early growth acceleration, bone age advancement, premature closure of growth plates and potential for compromise in adult stature. GnRH agonists are the treatment of choice. They are known to be safe, effective and their effect is reversible. Jaruratanasirikul et al., in a longitudinal observational study, showed an increase of 5 to 8 cm in final adult height after treatment of CPP with GnRH agonists.

To our knowledge, this is the first case of HH associated with CPP and GHD. The association of CPP and GHD has been described in cases of arachnoid cysts, septo-optic dysplasia, cerebral tumors, after radiotherapy, in few cases after cerebral trauma and in four patients who had nonacquired GHD associated with CPP and were discovered to have a developmental defects in the hypothalamic-pituitary area. This association had also been known in Pallister-Hall syndrome.

Conclusion: In conclusion, we report a unique case of a girl with HH associated with CPP and growth hormone deficiency. This case highlights the importance of following growth velocity even after the completion of treatment for CPP and to consider GH stimulation testing in the absence of a growth spurt.

Poster Abstract 5

Impact of the Pediatric Residents' Initiative For Healthy Active Living In Youth (RHALY): A Prospective Cohort Study

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Background: To determine 1) the prevalence of sub-optimal healthy habits in children followed at the Residents Continuity Clinic (RCC) at the Montreal Children's Hospital; 2) the effect of structured counselling and dissemination of healthy active living (HAL) educational tools on the child's physical activity, sedentary behaviour, nutrition and sleep.

Methods: Didactic teaching sessions on childhood obesity screening and management were incorporated within the residents teaching curriculum. HAL educational tools (brochures, pamphlets) developed in part by the RHALY group were disseminated to residents. Our prospective study included 5-16 year-old children. The intervention included the pediatric residents providing to RCC patients/families: 1) structured counselling, including the 5,2,1,0 guidelines (Daily: 5 portions of fruits, vegetables; < 2 hr screen time; >1 hr physical activity, zero caloric drinks); 2) educational materials; and 3) personalized health habits prescription. Recruited patients and families completed a physical activity log and a questionnaire about the health habits of their child and their family members. The questionnaire data will be collected at 6 months and one year.

Results: Recruitment and follow-up of patients is currently underway.

Conclusion: Our study will be the first to assess the impact of a resident-led initiative for healthy active living in youth. We anticipate that our initiative will empower children and their families to improve their health habits.

Poster Abstract 6

Esophageal stricture following severe diabetic ketoacidosis in a 12 year old girl

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A 12 year old girl presented in severe diabetic ketoacidosis with pH 6.81, blood glucose 36.2 mmol/L, and large urine ketones. At presentation, she had altered mental status with concern for cerebral edema, her blood pressure was initially unobtainable, and she was toxic-appearing. Additionally, in her initial course she had coffee-ground emesis. Her acute presentation was complicated by influenza A illness with pleural effusion and pneumonia, and she required chest tube placement. After hospital discharge she subsequently went on to develop progressive dysphagia. Her oral intake decreased over the next month, and she had an 8.7 kg weight loss. Evaluation by endoscopy revealed esophageal stricture, and despite multiple esophageal dilation procedures she continues to have a worsening stricture of her distal esophagus. She has ongoing difficulty tolerating oral intake of solids and liquids, and she has suffered significant morbidity as a result of this rare complication of DKA.

Acute esophageal necrosis can occur in the setting of acute illnesses associated with poor perfusion. Diabetes is thought to be a risk factor, but that is typically in the setting of adult patients with multiple other medical co-morbidities. We are aware of nine cases in the literature associated with DKA, and no cases previously reported in children with DKA. The initial presentation of acute esophageal necrosis can include evidence of upper gastrointestinal bleeding, as was seen in this case. The initial endoscopic appearance is a circumferentially black-appearing and friable esophageal mucosa, and this gives way to thick white exudate with underlying friable pink mucosa. Severe complications can include stricture formation or perforation. We will present a literature review of acute esophageal necrosis, describing the management and prognosis of this rare condition. We will also discuss this patient's difficult course, and her ongoing management with regards to esophageal stricture, nutrition, and insulin management.

Poster Abstract 7

The Truthful Teenager: A rare case of an insulinoma as a first presentation of MEN-1 in a teenage male

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We report a case of a 14-year-old male, known to experiment with illicit drugs, who was brought to the emergency room with altered mental status. Full toxicology screen was negative. He was found to be hypoglycemic and on further investigation had two pancreatic insulinomas which required resection. Despite having no relevant family history, genetic evaluation showed a mutation consistent with Multiple Endocrine Neoplasia type 1 (MEN1). Insulinomas in children and adolescents are exceedingly rare in general and even less common as a first presentation of MEN1. This diagnosis carries implications for potential future neoplasms, both benign and malignant. Ongoing surveillance and management is extensive and lifelong. Management of hypoglycemia due to insulinoma can be accomplished temporarily through medication or more definitively through surgery, although recurrences are common.

While intoxication is a more common cause of altered mental status in adolescents, clinicians must maintain a high index of suspicion for organic disease as this may dramatically alter the course of treatment.

Poster Abstract 8

Recombinant Parathyroid Hormone Therapy in Severe Hypoparathyroidism

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This case describes the effective short and long term treatment of recombinant parathyroid hormone (rPTH) in a three year old child with hypoparathyroidism-retardation-dysmorphism (HDR) syndrome, known at Sanjad-Sakati Syndrome, a rare autosomal recessive disorder characterised by hypoparathyroidism, growth failure, developmental delay and characteristic facies. In the neonatal period the patient sustained refractory hypocalcemia requiring high dose calcitriol 800ng/kg/day (usually 90ng/kg/day). Treatment over twelve days with subcutaneous rPTH was an effective alternative to conventional oral therapies and produced predictable effects of calcium homeostasis during a period of severe feeding intolerance and malabsorption. rPTH was reconsidered when on conventional therapy the patient had been very unstable, since August 2009 and March 2011 there were a total of 21 hospital admissions with gastroentestinal (GI) disturbance and hypocalcemia requiring calcium/phosphate stabilisation. After approval from the Drug Committee at The Children's Hospital Westmead, rPTH therapy was restarted daily and this showed a significantly reduced number of hospital admissions for calcium stabilisation. rPTH is not usually considered in the treatment algorithm for hypoparathyroidism in the pediatric population. However it may have a role in the management of severe hypocalcemia in children with associated GI abnormalities with the potential for long term treatment of hypoparathyroidism.

Poster Abstract 9

Long-Acting Release (LAR) Octreotide in Congenital Hyperinsulinism (CI).

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This case describes the effects of monthly intramuscular LAR Octreotide for the management of congenital hyperinsulinism in a 16 month old girl with pseudohypoparathyroidism (PHP) and Albright Hereditary Osteodystrophy (AHO). Due to other significant co morbidities this patient required lengthy periods of hospital admissions with a complex medical background of severe developmental delay with a tracheostomy, a seizure disorder and a Sturge Weber syndrome variant. Persistent hypoglycemia as a result of hyperinsulinism was however the primary reason for her prolonged admission. She was diazoxide unresponsive and medically too unstable to transfer from Sydney to Brisbane for pancreatic imaging with PET scan facilities. Genetic studies were undertaken for investigation of defects in SUR1 and Kir6.2 proteins but there were no genetic mutations in ABCC8 and KCNJ11 genes found. Further analysis of genes are underway at Exeter. We communicated with Dr Arnoux and his team in France, who had experience in use of LAR Octreotide in CI, as our daily regimen was not sustainable long term since the primary carer, the mother, had needlephobia. Our patient was started on once monthly IM octreotide whilst weaning off daily Octreotide over a further 3 month period in hospital. This case adds to the reviewed current literature and highlights the overall improvement in management of CI with significant enhancement in the quality of life for the patient and family.

Poster Abstract 10

Prolactinoma -The Role of the Endocrinologist in Pituitary Tumors.

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A 13 year old boy presented with abdominal pain, was diagnosed with appendicitis and underwent an appendicectomy. As part of his preoperative work up, a six month history of headaches was elicited. The day before admission he had an optometrist review which showed a temporal field defect and visual loss. An endocrine consultation revealed decreased growth velocity with weight gain, mild diabetes insipidus and a pre-pubertal status. The MRI findings showed a large lobulated supracellar and intrasellar mass measuring 4.4x3.3x4.6cm. The neurosurgeons reviewed the MRI images and planned a date for surgery with the presumed diagnosis of craniopharyngioma. Baseline endocrine bloods performed showed a level of prolactin of >3,000 mIU/L, and on dilution 20,288 mIU/L (normal <760mIU/L). There was a difference in opinion between the endocrinology and neurosurgical team about management. The endocrinologists requested a trial of Cabergoline for 4-6 weeks with repeat MRI images and visual field testing, since extreme elevation of prolactin was virtually diagnostic of prolactinoma and there was no change in the 6 month symptoms, no acute pituitary apoplexy or raised ICP indicating immediate surgery. The case describes the outcome in detail and reviews the literature, presenting the debate on surgical versus conservative management of prolactinomas with recommendations by the Endocrine Society 2011 on Management of Hyperprolactinemia.

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Mark Your Calendar

The next CPEG Scientific Meeting will be held on
February 20-22, 2014 at **Le Centre Sheraton Montréal Hotel, in Montréal, QC**

Important Dates:
November 22, 2013: Abstract submission deadline
January 10, 2014: Early-bird deadline for registration
January 18, 2014: Early-bird deadline for hotel
($149/night + applicable taxes, based on availability)