CPEG GCEP

Canadian Pediatric Endocrine Group
Groupe canadien d'endocrinologie pédiatrique

February 25-26, 2021

Digital Conference

15th Annual

CPEG Scientific Meeting

Hosted in Toronto by:Division of Endocrinology
Hospital for Sick Children

Official Program

Welcome to Toronto (virtually!)

We are delighted to be hosting CPEG 2021. What a year! At the start of the pandemic we were unsure as to whether we would be hosting virtually or an in-person conference. There was discussion of cancellation or postponement. At a certain point we decided to host the CPEG meeting virtually, and then…. Jo informed us that our longstanding meeting organizing company was closing. This left us scrambling to find a replacement, and fortunately, the University of Toronto Continuing Professional Development group stepped in. Fast forward 6 months and we are proud to have persevered! We have created a scientific program that covers topics requested from previous CPEG evaluations and highlights renowned speakers in their field. Our conference platform is interactive, so please take advantage of all the features. Network with colleagues and visit our industry partners who support our Scientific Meeting.

Thank you for joining us. Welcome to CPEG 2021!

Jill, Diane, Farid, Mark, Jonathan, Julia, Etienne and Rayzie

Dear Delegates,

Welcome! I would like to extend to you a warm welcome to the 15th Annual Scientific Meeting of the Canadian Pediatric Endocrine Group (CPEG). Our past meetings have provided a wonderful opportunity for the Canadian pediatric endocrine community to come together to learn, network, share ideas, visit old friends and make new ones. Of course, this year's meeting will be a bit different but hopefully still offer each of you these same opportunities, just in a different format. Planning this year's meeting was complicated by the COVID-19 pandemic, which forced us to go to a virtual format, and the folding of our longtime conference organizing partner, University of British Columbia Interprofessional Continuing Education. However, with our new partner, the University of Toronto Continuing Professional Development group, the organizing committee has developed a wonderful virtual program. As in past years, the meeting highlights work from our local hosts, this year "virtual" Toronto, and also includes presentations by other national and international experts. It was important for us to continue to provide the opportunity for our learners to present their work in scheduled oral and poster abstract sessions. In addition, we will continue to enjoy high level scientific symposia and the now infamous CPEG debate. We hope that our efforts and attempts to adapt will produce a virtual program that meets the needs of all attendees including our nurses, scientists, endocrinologists and trainees.

I would like to thank our sponsors who continued to support us this year and made this meeting possible. This year's virtual format allows for a unique interaction with exhibitors and I encourage you to explore the virtual exhibits. I would also like to thank those sponsors who also support our CPEG Fellowship Awards allowing us to train our future endocrinologists. This year's awardees will be announced at this meeting.

I wish you all a stimulating and collegial meeting and look forward to seeing and hearing you all on my computer.

Sincerely,

Seth Marks MD, MSc, FRCPC
Scientific Chair
CPEG 2021 Scientific Meeting

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Program Overview

The 15th Annual Scientific Meeting of Canadian Pediatric Endocrine Group (CPEG) includes a program of current and high-level content in pediatric endocrinology. The meeting also provides an opportunity for the Canadian pediatric endocrine community to come together, network and share ideas.

Of course, this year's meeting will take a different format and be done virtually due to the worldwide COVID-19 pandemic. The organizing committee has worked hard to adapt the annual meeting to this virtual format and still provide an exceptional educational experience.

The program includes theme-based symposia, an annual debate, oral abstracts, and poster presentations. Presenters include national and international experts. The meeting also provides a forum for trainees to present their work.

We have an exciting program planned for this year that should meet your educational needs as it has in past years.

Program Learning Objectives

At the conclusion of this conference, the participants will be able to:

1. Define neonatal hypoglycemia and its proper management specifically in hyperinsulinism
2. Differentiate and employ advanced diabetes technology to improve management in patients living with type 1 diabetes
3. Recognize potential adjunctive therapies to use along with insulin in patients living with type 1 diabetes
4. Utilize current approaches in the management of lipid disorders in children

Session Learning Objectives

Symposium I: Hyperinsulinism

SickKids Experience with Hyperinsulinism – *Jennifer Harrington*
Objectives:
1. Review the current challenges in the clinical management of children with hyperinsulinism
2. Explore presenting features that can help streamline clinical decisions
3. Discuss possible opportunities for future improvement in the management of children with hyperinsulinism

Clinical Approach to the Management of Hyperinsulinism – *Paul Thornton*
Objectives:
TBA

Genetics of Hyperinsulinism – *Sarah Flanagan*
Objectives:
1. Describe the most common genetic causes of congenital hyperinsulinism.
2. Explain the genetic mechanism of focal and diffuse hyperinsulinism.
3. Explain why a genetic diagnosis is clinically important for individuals diagnosed with congenital hyperinsulinism.

Symposium II: Diabetes

Adjunctive Therapy in Type 1 Diabetes - *Bruce Perkins*

Objectives:

1. Explain the rationale and efficacy of adjunctive-to-insulin therapy in type 1 diabetes, such as Sodium Glucose-Linked Transporter Inhibition (SGLTi)
2. Recognize putative renal and cardiometabolic advantages to SGLTi in T1D
3. Identify key findings from clinical trial programs and regulatory discussions of SGLT inhibition in type 1 diabetes

Advances in Diabetes Technology - *Gregory Forlenza*

Objectives:

1. To understand the importance of continuous glucose monitoring in control of people with type 1 diabetes
2. To review the pivotal trial results for existing and emerging hybrid closed loop artificial pancreas systems

Prevention of Type 1 Diabetes - *Diane Wherrett*

Objectives:

1. Review recent developments in identifying those at risk of developing type 1 diabetes
2. Examine current trials of interventions to delay/prevent the development of type 1 diabetes and to preserve insulin secretion in recent onset diabetes
3. Discuss challenges and future directions in progress toward prevention of type 1 diabetes

Symposium III: Lipid Disorders

Approach to Pediatric Lipid Disorders - *Katherine Morrison*

Objectives:

1. Develop an approach to the diagnosis and management of lipid disorders in children
2. Understand the mechanism and potential use of lipid lowering therapies

Gut Peptide and Neuroendocrine Regulation of Hepatic Lipid and Lipoprotein Metabolism in Health and Disease - *Khosrow Adeli*

Objectives:

1. Understand the pathophysiology of diabetic dyslipidemia in insulin resistant states
2. Learn the key underlying mechanisms particularly the role of the gut-brain-live axis
3. Identify critical neuroendocrine pathways that regular hepatic and intestinal lipid absorption and hepatic lipid and lipoprotein metabolism

Lipodystrophies - *Abhimanyu Garg*

Objectives:

1. To recognize rare disorders of lipodystrophies.
2. To learn about metabolic complications associated with lipodystrophies.
3. To recognize various types of genetic and acquired lipodystrophies.
4. To learn about targeted therapies for patients with lipodystrophies.

Debate

Be it resolved that all children with papillary or follicular thyroid cancer be treated with radioactive iodine
PRO: ***Sarah Lawrence***
CON: ***Jonathan Wasserman***

Objectives:
1. List and describe the indications for radioactive therapy in children with differentiated thyroid (papillary and follicular) carcinoma
2. Decide whether RAI is most appropriately administered on a universal or selective basis to affected children with differentiated thyroid carcinoma

CPEN Symposium

Genetic Counselling & Testing in Endocrinology - ***Lucie Dupuis***

Objectives:
1. Compare the utility of cytogenetic and molecular testing
2. Case examples – selecting a genetic test and interpreting results
3. Review the role of genetic counselling

Hyperinsulinemia: The highs and the lows - ***Eileen Pyra***

Objectives:
1. Understand the pathophysiology of diabetic dyslipidemia in insulin resistant states
2. Learn the key underlying mechanisms particularly the role of the gut-brain-live axis
3. Identify critical neuroendocrine pathways that regular hepatic and intestinal lipid absorption and hepatic lipid and lipoprotein metabolism

How Low Can you Go? Features & Management of Hypoparathyroidism in the Clinical Setting - ***Julia Sorbara***

Objectives:
1. To review the underlying physiology of calcium homeostasis
2. To identify the biochemical and clinical features of hypoparathyroidism in children and adolescents
3. To describe the principles of management of hypoparathyroidism in children and adolescents

Subcutaneous Lupron: A Novel Approach to Precocious Puberty Management - ***Mabel Tan***

Objectives:
1. Identify available treatments for precocious puberty in Canada
2. Discuss advantages or disadvantages to available treatments for precocious puberty in Canada
3. Describe patient teaching considerations for use of subcutaneous Lupron therapy

Interactive Learning

Each presentation will include a 25% (minimum) of interactivity from a combination of audience submitted questions throughout the presentations via the chat and audience polling questions.

Accreditation

Royal College of Physicians and Surgeons of Canada - Section 1

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 Post MD Education – Continuing Professional Development Temerty Faculty of Medicine, University of Toronto. You may claim up to a maximum of **8.0 hours** (credits are automatically calculated).

American Medical Association – AMA PRA Category 1 Credit™

Through an agreement between the Royal College of Physicians and Surgeons of Canada and the American Medical Association, physicians may convert Royal College MOC credits to AMA PRA Category 1 Credits™. For more information on the process to convert Royal College MOC credit to AMA credits please see: https://www.ama-assn.org/education/earn-credit-participation-international-activities.

European Union for Medical Specialists (EUMS) EC-MEC® Credit

Live educational activities recognized by the Royal College of Physicians and Surgeons of Canada as Accredited Group Learning Activities (Section 1) are deemed by the European Union of Medical Specialists (UEMS) eligible for ECMEC®.

Faculty Disclosure

It is the policy of the University of Toronto, Faculty of Medicine, Continuing Professional Development to ensure balance, independence, objectivity, and scientific rigor in all its individually accredited or jointly accredited educational programs. All speakers, moderators, facilitators, authors and scientific planning committee members participating in University of Toronto accredited programs, are required to disclose to the program audience any real or apparent conflict(s) of interest that may have a direct bearing on the subject matter of the continuing education program. This pertains but is not limited to relationships within the last FIVE (5) years with for-profit organizations, not-for-profit and public sector sponsors and donors, biomedical device manufacturers, or other corporations whose products or services are related to the subject matter of the presentation. The intent of this policy is not to prevent a speaker with a potential conflict of interest from making a presentation. It is merely intended that any potential conflict of interest should be identified openly so that the listeners may form their own judgements about the presentation with the full disclosure of facts. It remains for the audience to determine whether the speaker's outside interests may reflect a possible bias in either the exposition or the conclusions presented.

Scientific Committee

- Seth Marks (Chair)
- Jill Hamilton (Local Chair)
- Shazhan Amed
- Barbara Butler
- Farid Mahmud
- Munier Nour
- Colleen Nugent
- Rebecca Perry
- Wendy Schwarz
- Bailie Tabak
- Diane Wherrett
- Brandy Wicklow
- Zachary Zytner

Invited CPEG Speakers

Khosrow Adeli PhD FCACB DABCC FACB
Professor
Biochemistry and Laboratory Medicine & Pathobiology
University of Toronto
Division Head, Clinical Biochemistry
The Hospital for Sick Children
Toronto, ON

Sarah Flanagan PhD
Associate Professor
Genomic Medicine
Sir Henry Dale Fellow
University of Exeter Medical School
Exeter, UK

Gregory P. Forlenza MD
Assistant Professor
Pediatrics
Barbara Davis Center for Childhood Diabetes
University of Colorado
Aurora, CO

Abhimanyu Garg MD
Professor & Chief
Division of Nutrition and Metabolic Diseases
Distinguished Chair, Human Nutrition Research
UT Southwestern Medical Center
Dallas, TX

Jennifer Harrington MBBS PhD
Assistant Professor
Pediatrics
University of Toronto
Staff Endocrinologist
Hospital for Sick Children
Toronto, ON

Sarah Lawrence MD FRCPC
Associate Professor & Chief
Division of Endocrinology
University of Ottawa
Division Head, Pediatric Endocrinology
Ottawa, ON

Katherine Morrison MD
Associate Professor
Pediatrics
Co-director, Metabolism and Childhood Obesity Research Program
McMaster University
Hamilton, ON

Bruce A. Perkins MD MPH FRCPC
Professor & Clinician-Scientist
Medicine
University of Toronto
The Sam and Judy Pencer Family Chair in Diabetes
Director of the Diabetes Clinical Research Unit
Leadership Sinai Centre for Diabetes
Sinai Health System
Toronto, ON

Paul Thornton MB BCh MRCPI DCh
Medical Director
Endocrine and Diabetes Program
Cook Children's Health Care System
Fort Worth, TX

Jonathan Wasserman MD FRCPC
Staff Physician, Endocrinology
Project Investigator, Genetics and Genome Biology
Hospital for Sick Children
Associate Professor, Paediatrics
Toronto, ON

Diane Wherrett MD FRCPC
Professor
Endocrinology
University of Toronto
Staff Endocrinologist
Hospital for Sick Children
Toronto, ON

Invited CPEG Speaker Biographies

Khosrow Adeli
Dr. Adeli is the Division Head of Clinical Biochemistry at SickKids and a Professor at the University of Toronto. For the past 30 years, he has been actively involved in molecular and clinical laboratory research, and is well known for his contributions to the field of metabolic health and disease.

Sarah Flanagan
Sarah's research focuses on understanding the genetic basis of congenital hyperinsulinism and neonatal diabetes with her work contributing to the discovery of 17 disease genes. With the team in Exeter Sarah has helped to provide genetic testing for over 6000 patients affected by these conditions from over 100 countries worldwide.

Gregory Forlenza
Dr. Forlenza is a pediatric endocrinologist at The Barbara Davis Center at the University of Colorado Denver. He has been caring for children with type 1 diabetes for over 15 years. His research is focused on technology to improve the health and lifestyle of people with type 1 diabetes.

Abhimanyu Garg
Abhimanyu Garg, M.D. is Chief, Division of Nutrition and Metabolic Diseases at UT Southwestern. He has carefully characterized the clinical and metabolic features of lipodystrophies for over 30 years, including reporting of novel syndromes. He has discovered many novel lipodystrophy genes, such as, AGPAT2, PPARG, ZMPSTE24, PSMB8, ADRA2A, and PRRT3.

Jennifer Harrington
Jenny joined the Hospital for Sick Children's faculty in 2015. Her clinical and research interests include the management of children with bone and mineral disorders as well as hyperinsulinism. Just recently she has returned to Adelaide, Australia to take up a new position at the Women's and Children's Hospital.

Sarah Lawrence
Sarah Lawrence is currently the Division Head for Pediatric Endocrinology at CHEO and has been practicing at CHEO since 1995 with clinical interest in general endocrinology and a focus on neuro-oncology, growth and puberty, and has been Medical Director for Camp Banting for 20 years. Her academic career has been focused in medical education, having served as Pediatrics Program Director from 2004 to 2011. She is a longstanding member of RCPSC Pediatric Specialty and Examination committees, including a term as Director of the SAQ Committee.

She was the recipient of the 2014 Canadian Pediatric Society Michael Weber Education Award which recognizes a CPS member whose work in medical and/or inter-professional education has had a significant and positive impact on learners in child and youth health. Within endocrinology, she chaired the CPEG Growth Charts Working Committee which developed a new growth chart for use in Canada and contributed to the redesign of the national WHO Growth Charts Adapted for Canada.

Katherine Morrison
Dr. Katherine Morrison is a pediatric endocrinologist, Professor and Associate Chair -Research in the Department of Pediatrics at McMaster University. Dr. Morrison is also the Co-Director of the Centre for Metabolism, Obesity and Diabetes Research at McMaster. She is the Medical Director of the Growing Healthy Pediatric Weight Management clinic and the Pediatric Lipid Clinic at McMaster Children's Hospital. She completed her training at the University of Calgary and Stanford University, has held a research appointment at Ludwig-Maximilian University of Munich (Germany) and faculty positions at the University of Manitoba and McMaster. Dr. Morrison's research is centered upon the etiology, consequences and treatment of obesity and lipid disorders in children. Dr. Morrison's work is supported by the Heart and Stroke Foundation of Canada, the Canadian Institutes of Health Research and the Ontario Ministry of Health.

Bruce Perkins

Bruce A Perkins, MD MPH is Professor, Endocrinologist and Diabetes Complications Clinician-Scientist at the University of Toronto appointed to the Temerty Faculty of Medicine and to the Institute of Health, Policy, Management and Evaluation. He holds the Sam and Judy Pencer Family Chair in Diabetes Clinical Research. He obtained his MD and Internal Medicine training at the University of Toronto, his endocrinology subspecialty training at Harvard University, his Masters of Public Health in Epidemiology at the Harvard School of Public Health, and a research fellowship in epidemiology at the Joslin Diabetes Center.

Using longitudinal cohort methods as well as clinical trials, his research work has focused on early biomarkers of diabetes complications, and interventions for the prevention of complications, including artificial pancreas technologies and disease-modifying adjunctive-to-insulin pharmacotherapies. In 2015 he was awarded the Canadian Diabetes Association/CIHR Young Scientist Award for his research. Among other projects funded by the NIH, JDRF, and Diabetes Canada, he leads an Innovations in type 1 Diabetes group within Diabetes Action Canada, a national patient-oriented research strategy.

Paul Thornton

Paul Stephen Thornton, MB BCh, is Medical Director, Distinguished Consultant of the Endocrinology/Diabetes Clinic and the Hyperinsulinism Center at Cook Children's Medical Center, in Fort Worth, Texas.

His recent awards include the Cook Children's Clinical Scholar Award 2012-2014 and recipient of the 2012 Cook Children's Health Care System Endowed Chair Award.

As an expert in the evaluation and management of congenital Hyperinsulinism, Dr. Thornton is frequently invited to lecture on congenital hyperinsulinism and hypoglycemia in infants and children. He has coauthored more than a dozen book chapters and over 60 journal articles. He presents his research at international and national meetings. Paul is the lead author of the recent Pediatric Endocrine Society's Recommendations and Management of Persistent Hypoglycemia in Neonates, Infants, and Children. He also leads the Pediatric Endocrine Society's Rare Disease working group on Hyperinsulinism.

Diane Wherrett

Dr. Wherrett is a pediatric endocrinologist at the Hospital for Sick Children and Professor, Department of Pediatrics, University of Toronto. She completed medical school at Queen's University in Kingston, Ontario and paediatrics and pediatric endocrinology training at the Hospital for Sick Children. She completed a research fellowship in the immunology of type 1 diabetes at Stanford University. She has been a faculty member at Sick Kids since 1995.

Her major research focus is in interventions to prevent beta cell loss in type 1 diabetes. She is a member of the Steering Committee of the NIH-sponsored multi-centre clinical trials group, Type 1 Diabetes TrialNet, chairs its largest study and is the director for the Canadian Clinical Centre for this study group. She was a member of the Expert Panel for the 2008, 2013 and 2018 Diabetes Canada Clinical Practice Guidelines and was lead author of the chapter on Type 1 Diabetes in Children and Adolescents for the 2013 and 2018 guidelines. She is the medical lead of the Hospital for Sick Children's clinic for children with disorders of sex development.

Jonathan Wasserman

Jonathan is born and bred Torontonian. He pursued his undergraduate studies at McGill before heading to the University of Cambridge where he completed a PhD in Developmental Genetics. He then undertook an MD and peds residency at Boston Children's, subsequently returning to SickKids where he pursued a Peds Endo fellowship prior to joining the staff in 2012. His clinical focus is in Endocrine Neoplasms and endocrine care of Childhood Cancer Survivors.

Invited CPEN Speakers

Lucie Dupuis MSc MS CGC
Genetic Counsellor
The Hospital for Sick Children
Division of Clinical and Metabolic Genetics
Department of Genetic Counselling
Lecturer, Dept of Molecular Genetics
University of Toronto
Toronto, ON

Eileen Pyra RN MN
Clinical Nurse Specialist
Endocrine Clinic
AB Children's Hospital
Calgary, AB

Julia Sorbara MD MSc FRCPC
Staff Physician, Pediatric Endocrinology
The Hospital for Sick Children
University of Toronto
Toronto, ON

Mabel Tan RN MScN AP-PEN
Nurse Clinician, Endocrinology
BC Children's Hospital
Vancouver, BC

Invited CPEN Speaker Biographies

Lucie Dupuis
Lucie Dupuis is an ABGC certified genetic counsellor who completed her MS in Genetic Counselling at Brandeis University. In 1998, she joined the Department of Genetic Counselling at the Hospital for Sick Children. Her primary interest is in skeletal dysplasias. Lucie is a lecturer and clinical supervisor at the University of Toronto MSc Program in Genetic Counselling.

Eileen Pyra
I became a registered nurse in 1973 and worked on a pediatric ward in a general hospital for 6 years. I graduated from the University of Calgary with a Bachelor of Nursing in 1980 and a Master in Nursing in 2007. I have worked in the Diabetes and/or Endocrine Clinics at Alberta Children's Hospital since 1980 where I am currently a Clinical Nurse Specialist. I have had the opportunity to participate in research projects, give presentations at ESPE, CPEG, CPEN and PENS and am a chapter contributor in a published textbook, Pediatric Endocrinology. I have 3 amazing daughters and most importantly, I am the Nana for 2 Grandsons and soon to be 1 Granddaughter.

Julia Sorbara
Julia Sorbara trained in Paediatrics and Paediatric Endocrinology at The Hospital for Sick Children. She joined the Division of Endocrinology at SickKids in 2019. Dr. Sorbara's clinical and academic interests include disorders of calcium and bone metabolism as well as the medical care of gender-incongruent youth.

Mabel Tan
Mabel Tan is a nurse clinician at BC Children's Hospital's Endocrine and Gender clinic. She received her Bachelor of Nursing degree from the University of British Columbia and completed her Master of Nursing degree in advance practice nursing at the University of Victoria. She also is the first nurse to receive the designation of Advance Practice- Pediatric Endocrine Nurse. She has been in her role for 20 years providing teaching and support to families and children/youth that are gender diverse and families and children/youth with endocrine conditions.

Fellowship Listing

1992-1993	M. Lawson
1993-1994	S. Lawrence M. Lawson A. Simone
1994-1995	S. Lawrence S.Taback A. Simone
1995-1996	C. Vaz S.Taback B. Cummings
1996-1997	J. Hamilton E. Sellers B. Cummings
1997-1998	J. Hamilton E. Sellers B. Cummings
1998-1999	J. Curtis J. Hamilton
1999-2000	J. Curtis J. Hamilton
2000-2001	C. Panagiotopoulos C. Huang
2001-2002	C. Panagiotopoulos S. Stock
2002-2003	P Krishnamoorthy P Zimakas R. McEachern
2003-2004	P Krishnamoorthy H. Bui
2004-2005	M. Nakhla J. Simoneau-Roy
2005-2006	M. Nakhla I. Chapados M. Jetha
2007-2008	B. Wicklow T. Pinto, B. Babic J. Deladoey
2008-2009	A.M. Sbrocchi P Olivier T. Pinto
2009-2010	R. Shulman P Olivier T. Edouard S. Runge-Wildi C. Saaman
2010-2011	E. Bassilious J. Wasserman Y. Yeshayahu S. Tsai
2011-2012	M. Millete J. Wasserman C. Zuijdwijk M. Cohen
2012-2013	J. Harrington T. Oron P. Luca M. Nour D. Manousaki
2013-2014	K. Winston C. Leblicq A. Ens B. Hursh I. Rousseau-Nepton
2014-2015	I. Levy D. Manousaki
2015-2016	L. Chiniara S. Basak K. Verbeeten
2016-2017	C. Nugent K. Pundyk N. Coles
2017-2018	C. Nugent S. Fuchs
2018-2019	J. Sorbara
2019-2020	A. Chesover B. Navabi
2020-2021	A. Marr M. Lautatzis J. Ladd H. Geddie

Within the last 5 years, the CPEG Fellowship Program was and/or is supported by:
Eli Lilly, EMD Serono, Novo Nordisk, Pfizer, Sandoz, and Ultragenyx.

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.

Below is a list of the recipients of the Dr. John Bailey Resident Research Award:

2007 Meranda Nakhla
2008 Meranda Nakhla
2009 David Saleh
2010 Brandy Wicklow
2011 Jonathan Wasserman
2012 Jennifer Harrington
2013 Karine Khatchadourian
2014 Akash Sinha
2015 Rayzel Shulman
2016 Sanjukta Basak
2017 Stephen Zborovski
2018 Marie Eve-Robinson
2019 Julia Sorbara
2020 Christine Tenedero

CPEG Distinguished Service Award

The CPEG Distinguished Service Award will be awarded periodically (not annually) to a member who has shown exemplary service to the organization or to the discipline of pediatric endocrinology in Canada. The award will be focused on work that furthers the aims of CPEG and can be in one or more of the following areas: administration, teaching, research, clinical service. Nominations will be solicited by the CPEG Executive Committee every 2- 4 years. CPEG members can put forward a name for nomination at any time. The nomination should include a letter signed by two CPEG members in good standing describing the contributions of the nominee. The award will be presented at the annual CPEG business meeting. The awardee will receive a certificate and a $500 donation to a charity of their choice.

Below is a list of past recipients of the CPEG Distinguished Service Award:

2017 Daniel Metzger
2019 Denis Daneman

Program Thursday, February 25, 2021 (agenda times listed in EST)

0945	Platform Orientation	
1000	Welcome & Opening Remarks	
	Symposium I: Hyperinsulinism ***Chairs:*** *Chelsey Grimbly, Edmonton & John Mitchell, Montreal*	
1015	SickKids Experience with Hyperinsulinism	*Jennifer Harrington, Adelaide/Toronto*
1045	Clinical Approach to the Management of Hyperinsulinism	*Paul Thornton, Fort Worth, TX*
1115	Genetics of Hyperinsulinism	*Sarah Flanagan, Exeter, UK*
1145	Break & Exhibits	
1200	**Oral Abstracts I** ***Chairs:*** *Rebecca Perry, Calgary & Katie Pundyk, Winnipeg*	
OR1	Treatment practices and confidence in the management of pediatric metabolic bone disorders: a survey of pediatric endocrinologists in North America	*Emma McCutcheon*
OR2	A Novel Longitudinal Diabetes Educational Program (the Diabetes Learning Centre) to Improve Confidence in Type 1 Diabetes Self-Management Skills in Adolescents	*Kristina Pabedinskas*
OR3	QTc Intervals in Gender Diverse Youth on Leuprolide Acetate	*Richelle Waldner*
OR4	Pediatric thyroglossal duct cyst carcinomas: what to do once the cat is out of the bag?	*Patricia Diaz Escagedo*
OR5	Persistently Hypoglycemic and Sometimes Hypophosphatemic: The Variable Presentation of HNF4A Variants	*Zachary Zytner*
OR6	Implementation of a Quality of Life and Mental Health Screening Tool in a Tertiary Care Pediatric Diabetes Clinic	*Zoyah Thawer*
1330	Self-Care/Nutrition Break & Exhibits	

1445	**Poster Viewing I**	
P01	A structural equation model of factors associated with prevalent albuminuria in youth with type 2 diabetes in the iCARE national cohort	*Melissa Gabbs*
P02	Incidental vertebral fractures in patients with Duchenne muscular dystrophy on intravenous bisphosphonate treatment	*Kim Phung*
P03	De novo mutation of IGF2 gene in a patient with short stature	*Kelly Milton*
P04	Assessing the need for teaching of Tanner stages of breast development using 3D breast models	*Rachel Parker*
P05	Severe Hypertriglyceridemia Complicating Pediatric Acute Lymphoblastic Leukemia Treatment: A Need for Monitoring Guidelines	*Carly Baxter*
P06	Exploring Medical Students' Perceived Competence and Comfort Performing Physical Examinations on Patients with Obesity: A Qualitative Study	*Rachel Parker*
P07	Are you sure that it is a Pituitary Macroadenoma?	*Funmbi Babalola*
P08	Use of Lanreotide in Infancy for Severe Hyperinsulinism	*Jennifer Ladd*
P09	Flip-Flopping Function in a Transplanted Teen with a Tricky Thyroid	*Krista Oei*
P10	A novel germline GNAS variant causes Nephrogenic Syndrome of Inappropriate Antidiuresis as part of an emerging multisystem phenotype	*Ashlee Yang*
P11	Congenital lipoid adrenal hyperplasia: a novel StAR mutation and atypical presentation	*Alexander Chesover*
P12	Could Multiple Pituitary Hormone Deficiencies Be Within the Spectrum of Disease in Aicardi Syndrome?: A Case Report	*Kriti Kumar*
P13	Antibody-negative insulinopenic diabetes mellitus following CAR T-cell therapy for pediatric acute lymphoblastic leukemia	*Maria-Elena Lautatzis*
P14	A Rare Presentation of New Onset Hypoglycemia in an Adolescent Male	*Shelby Thompson*
P15	A Rare Case of Differences of Sexual Development	*Noor Gazzaz*
P27	Diabetes Management Order Set (DEMOS): Patient Safety in Admitted Children and Youth with Diabetes	*Manasi Parikh*
1545	Break & Exhibits	
1600	CPEG Business Meeting	
1800	Adjourn for the Day	

Program Friday, February 26, 2021 (agenda times listed in EST)

0945	**Platform Orientation**	
1000	Welcome & Announcements	
	Symposium II: Diabetes ***Chairs:*** *Andrea Ens, London & Beth Cummings, Halifax*	
1015	Adjunctive Therapy in Type 1 Diabetes	*Bruce Perkins, Toronto*
1045	Advances in Diabetes Technology	*Gregory Forlenza, Aurora, CO*
1115	Prevention of Type 1 Diabetes	*Diane Wherrett, Toronto*
1145	**Break & Exhibits**	
1200	**Oral Abstracts II** ***Chairs:*** *Ereny Bassilious, Hamilton & Carol Lam, Vancouver*	
OR07	Elevated TSH in Children and Adolescents with Obesity: Retrospective Chart Review at Children's Hospital of Eastern Ontario's Centre for Healthy Active Living	*Behdad Navabi*
OR08	Impact of the COVID-19 pandemic on pediatric new-onset type 1 diabetes rates and severity of presentation in Montreal	*Kim Phung*
OR09	Patient and Family Perspective of a Pre-Transition Visit in a Pediatric Tertiary Care Diabetes Clinic	*Alexa Marr*
OR10	Evaluation of virtual visits for the routine clinical care of transgender youth during the COVID-19 pandemic	*Carolina Silva*
OR11	Understanding the Role of Maternal Vitamin D supplementation in Early Infantile Rickets in Dhaka, Bangladesh	*Maria-Elena Lautatzis*
OR12	Adrenal Insufficiency among Children treated with Hormonal Therapy for Infantile Spasms	*Gabrielle Dore-Brabant*
1330	**Self-Care/Nutrition Break & Exhibits**	

1445	**Poster Viewing II**	
P16	The Impact of the COVID-19 Pandemic on Adolescents and Young Adults with Type 2 Diabetes	*Marylin Carino*
P17	Insulin reactions: what do you do when your treatment's the trigger?	*Madeline Edwards*
P18	Growth response to human growth hormone in two patients with IGF1 receptor defects	*Sarah Riedlinger*
P19	Type 2 diabetes off-label treatment: Retrospective review of children and adolescents followed at Children's Hospital of Eastern Ontario's Type 2 Diabetes Clinic 2014-2020	*Behdad Navabi*
P20	A Novel STAT3 Gain-of-Function Mutation as a Cause of Neonatal Onset Polyendocrinopathy	*Christine Tenedero*
P21	Minimum Incidence of Optic Nerve Hypoplasia and Septo-Optic Dysplasia in Canadian Children: A CPSP Study	*Reem Alfattouh*
P22	Don't be chicken, it's just thyroiditis!	*Tracey Dyer*
P23	Permanent Versus Transient Hypothyroidism In Children With Congenital Hypothyroidism (CH) Identified Through The Newborn Screening Ontario (NSO) Program	*Alexa Marr*
P24	Illness Management Advice in Congenital Adrenal Hyperplasia – Is an Update Needed?	*Katie Ross*
P25	Successful Improvement of Severe Hypertriglyceridemia in Congenital Lipodystrophy type 4 with Diet and Icosapent Ethyl	*Funmbi Babalola*
P26	Variable Presentations in the Diagnosis of Complete Androgen Insensitivity Syndrome	*Matthew Feldman*
P27	A Rare Etiology of Clitoromegaly in Neurofibromatosis Type 1	*Richelle Waldner*
P28	Bilateral gynecomastia due to intake of sweet potatoes: a case report	*Noor Alhuda Sawalha*
	Symposium III: Lipid Disorders ***Chairs:*** *Melanie Henderson, Montreal & Karine Khatchadourian, Ottawa*	
1545	Approach to Pediatric Lipid Disorders	*Katherine Morrison, Hamilton*
1615	Gut Peptide and Neuroendocrine Regulation of Hepatic Lipid and Lipoprotein Metabolism in Health and Disease	*Khosrow Adeli, Toronto*
1645	Lipodystrophies	*Abhimanyu Garg, Dallas, TX*
1715	Break & Exhibits	

	Debate ***Chairs:*** *Mark Inman, Saskatoon & Munier Nour, Saskatoon*	
1730	Be it resolved that all children with papillary or follicular thyroid cancer be treated with radioactive iodine	***Pro:*** *Sarah Lawrence, Ottawa* ***Con:*** *Jonathan Wasserman, Toronto*
1830	John Bailey Award, CPEG Fellowship Awards, & Closing Remarks	
1900	Meeting Adjourns	

CPEN Program Thursday, February 25, 2021 (agenda times listed in EST)

1415	CPEN Business Meeting
1545	Meeting Adjourns

CPEN Program Friday February 26, 2021 (agenda times listed in EST)

	CPEN Symposium ***Chair:*** *Bailie Tabak, Toronto*	
1015	Genetic Counselling & Testing in Endocrinology	*Lucie Dupuis*
1100	Hyperinsulinemia: The highs and the lows	*Eileen Pyra*
1200	How Low Can you Go? Features & Management of Hypoparathyroidism in the Clinical Setting	*Julia Sorbara*
1245	Subcutaneous Lupron: A Novel Approach to Precocious Puberty Management	*Mabel Tan*
1330	Rejoin CPEG Group	

Oral Abstracts

OR1

Treatment practices and confidence in the management of pediatric metabolic bone disorders: a survey of pediatric endocrinologists in North America

Emma McCutcheon (1), Sasigarn A. Bowden (2), Halley Wasserman (3), Alicia Diaz-Thomas (4), Laura Bachrach (5), Marie-Eve Robinson (1,6); and the Pediatric Endocrine Society Bone and Mineral Special Interest Group

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Pediatric bone diseases encompass a wide variety of disorders with challenging diagnostic approaches and management due to disease rarity, or inadequate dedicated training. As practice variation and confidence level may impact clinical outcome, we sought to assess physician confidence level in management of pediatric bone disorders as well as treatment practices among North American pediatric endocrinologists and pediatric endocrinology fellows through an online survey distributed through the Pediatric Endocrine Society (PES) and the Canadian Pediatric Endocrine Group (CPEG). Two hundred and forty-four surveys were completed. Variations were observed amongst the respondents' confidence in the management of bone disorders and in the criteria used to initiate/discontinue intravenous bisphosphonates or prescribe burosumab therapy (Table 1). Over 50% of respondents reported not feeling confident with tumor-induced osteomalacia, chronic recurrent multifocal osteomyelitis, osteonecrosis and osteopetrosis (Table 1). Physicians working in a bone clinic had greater comfort in prescribing burosumab for the treatment of X-linked hypophosphatemic rickets compared to those not working in a bone clinic (65% versus 47%, $p = 0.03$). Most respondents reported having received inadequate training in the field (52%). Common barriers to providing care included rarity of bone disorders (69%), paucity of clinical guidelines (53%), and personal lack of knowledge in the field (47%). The most commonly identified needs were: online education for physicians (89%), a consultation platform with bone experts (80%), and an e-mail list of appropriate bone experts (65%). Dedicated training, knowledge acquisition and education resources are needed to increase confidence and standardize the use of bone-targeted therapies.

OR2

A Novel Longitudinal Diabetes Educational Program (the Diabetes Learning Centre) to Improve Confidence in Type 1 Diabetes Self-Management Skills in Adolescents

Kristina Pabedinskas (1,2), Jennilea Courtney (3), Nick Barrowman (3,4), Christine Richardson (5), Liz Stevens (5), Ellen B Goldbloom (3,4,5), Sarah E Lawrence (3,4,5), Caroline Zuijdwijk (3,4,5), Margaret Lawson (3,4,5), Sarah Zankar (4), Alexandra Ahmet (3,4,5).

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Background: During transition to adulthood, many adolescents with type 1 diabetes (T1D) demonstrate insufficient knowledge for continued self-management and experience deterioration in glycemic control. Higher confidence in T1D self-management skills (self-efficacy) has been shown to predict better self-reported adherence and glycemic control. Educational interventions can expand diabetes knowledge and self-efficacy in youth; however, in most T1D centres, implementing structured and continuous education, in keeping with the recommendations in international guidelines, has been challenging. To address this, a working group with representatives from each discipline within the CHEO Diabetes Team developed a novel longitudinal educational program. The Diabetes Learning Centre (DLC) was designed to improve adolescents' self-efficacy through confidence in T1D self-management skills.

Objectives: To describe the conception and implementation of the DLC. To evaluate adolescents' confidence in T1D self-management skills and predictors of confidence prior to their first DLC visit.

Methods: 13 to 17 year old youth rated their confidence in overall T1D management and individual T1D self-management skills on a 5-point Likert-scale prior to attending the DLC. Baseline characteristics were collected and summarized using frequency and percentage for discrete variables and median and interquartile range (IQR) for continuous variables. Spearman's correlation coefficient was used to estimate association between ordinal and continuous characteristics.

Results: 232 eligible youth were approached and 215 (92.7%) consented and completed their questionnaire: 97 (45.1%) females, age 14.9 (IQR 13.9, 15.9) years, duration of diabetes 5.8 (IQR 2.9, 8.9) years, A1C 7.9% (IQR 7.1, 8.8), 110 (51.2%) on insulin pumps, 47 (21.9%) with parents involved "always" or "very often" in management. Median overall confidence in diabetes management on a 5-point (0-4) Likert-scale was 3, representing "quite confident". This correlated with mean ratings of individual self-management skills ($r=0.54$). Higher confidence in overall diabetes management was associated with lower A1C ($p<0.001$). There was little evidence of association between confidence and other baseline characteristics ($p>0.05$).

Conclusions: Adolescents reported being quite confident in T1D self-management skills prior to the DLC, which aims to further improve confidence in diabetes related skills. Assessment of this novel educational program is ongoing, including evaluation of its impact on confidence, glycemic control, and patient satisfaction.

OR3

QTc Intervals in Gender Diverse Youth on Leuprolide Acetate

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Background: Puberty suppression is a standard of care for gender affirming therapy in gender diverse youth. Leuprolide acetate is a gonadotropin releasing hormone agonist (GnRHa) commonly used for pubertal suppression. There are concerns that GnRHa agents prolong the rate-corrected QT interval (QTc) when used as androgen deprivation therapy in management of prostate cancer. There is a paucity of literature regarding leuprolide acetate and QTc intervals in gender diverse adolescents. Our study aimed to determine the proportion of gender diverse youth that had QTc prolongation, defined as greater than 460 milliseconds (ms), while on leuprolide acetate.

Methods: We performed a retrospective chart review of gender diverse youth who received care at the Stollery Children's Hospital Endocrinology Gender Clinic between July 1, 2018 and December 31, 2019. Youth aged 9-18 years were included if they had a 12-lead electrocardiogram (ECG) obtained after initiating leuprolide acetate. If available, baseline or pre-leuprolide acetate, ECGs were also reviewed. The ECGs were read by a pediatric cardiologist who calculated the QTc values. Data on concomitant medications and/or gender affirming therapy was also collected. QTc values and continuous variables were analyzed by determining the mean, standard deviation, minimum and maximum values.

Results: A total of thirty-three pubertal adolescents were included. Our cohort had a mean (SD) age of 13.7 (2.1) years and 69.7% identified as male (assigned female at birth). Nineteen youth had a baseline ECG. The mean (SD) QTc at baseline was 413ms (18). The mean (SD) post-leuprolide acetate QTc was 415ms (27), with a minimum of 372ms and maximum of 455ms. Eight adolescents (24.2%) on leuprolide acetate had a QTc between 440ms and 460ms, categorized as borderline prolongation. None of the 33 adolescents on leuprolide acetate demonstrated QTc prolongation greater than 460ms.

Conclusion: In this study, we defined QTc prolongation conservatively at 460ms and no gender diverse youth on leuprolide acetate demonstrated clinically significant QTc prolongation. QTc prolongation may not be a clinical concern in healthy adolescents pursuing puberty suppression for gender affirmation.

OR4

Pediatric thyroglossal duct cyst carcinomas: what to do once the cat is out of the bag?

Patricia Diaz Escagedo (1), Céline Huot (1), Guy Van Vliet (1), Noémie Rouillard-Bazinet (2), Dorothée Dal Soglio (3), Sophie Turpin (4), Despoina Manousaki (1)

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The thyroglossal duct is a remnant of the embryonic migration of the thyroid from the tongue to the anterior neck and usually contains degenerated thyroid follicular cells. In 7% of the population, a cyst develops along the duct (Thyroglossal Duct Cyst, TGDC). An asymptomatic cervical mass is the most typical presentation. The Sistrunk procedure (SP), which consists of excision of the cyst, the central portion of the hyoid bone, and the soft tissues around the duct, is the most common surgical approach. Carcinomas in TGDC (TGDCCa) are found in 1-3.9% of resected cases. TGDCCas can be papillary, mixed (papillary and follicular), squamous, or anaplastic, with the majority being papillary, in adult women, and with a good prognosis. Preoperatively, TGDCCas are often indistinguishable from benign TGDC, and the diagnosis is based on histology. SP is recommended for adult patients < 45 years without any risk factors; otherwise, a more extensive approach is considered. TGDCCa are rare in childhood and their management remain unclear. Among the 59 reported cases of pediatric TGDCCa, all were papillary and underwent a SP, while thyroidectomy was also performed in half and radioiodine treatment given to a third.

We present four pediatric cases with TGDCCa identified in our center over 10 years: patient 1 presented with a papillary thyroid carcinoma without any risk factor. Patient 2 also had papillary thyroid carcinoma but with extensive local disease. Patient 3 exhibited a poorly differentiated follicular carcinoma. Patients 2 and 3 had SP, total thyroidectomy, and radioiodine, while patient 1 underwent SP with favorable evolution. Patient 4 was recently diagnosed with a papillary carcinoma and a post-operative suspicion of lymph node invasion. The details are shown in the Table.

In summary, TGDCCas in children present a management challenge. The decision to complete their surgical resection with thyroidectomy and radioiodine should be made by an interdisciplinary team.

OR5

Persistently Hypoglycemic and Sometimes Hypophosphatemic: The Variable Presentation of HNF4A Variants

Zachary Zytner (1), Dror Koltin (1), Jennifer Harrington (1)

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We present four children (2 females, 2 males) who presented with hyperinsulinemic hypoglycemia secondary to HNF4A pathogenic variants in the neonatal period, and describe their variable course.

All children had birth weight greater than 4.0 kg. They required high maximum glucose infusion rates to treat hypoglycemia (12–26.4 mg/kg/min) and all except Female 2 required treatment with glucagon infusions. All had critical hypoglycemic samples which supported the diagnoses of hyperinsulinism, with serum glucose 2.2–2.6 mmol/L and serum insulin 35–393 pmol/L. Diazoxide was started within the first month of life with maximum doses 10–15 mg/kg/day. All were diazoxide-responsive and discharged from hospital on diazoxide.

Genetic testing for monogenic forms of hyperinsulinism revealed pathogenic variants in the hepatocyte nuclear factor 4a (HNF4A) gene. The two male subjects manifested with hyperinsulinism alone, with Male 1 having a heterozygous HNF4A c.114T>A (p.Cys-38Ter) pathogenic variant and Male 2 found to have a heterozygous HNF4A pathogenic deletion of Exons 6 and 7. The two female subjects were found to have HNF4A pathogenic variants also associated with Fanconi syndrome (Female 1: heterozygous c.187C>T (p.Arg63Trp) pathogenic variant and Female 2 heterozygous c.253C>T (p.Arg85Trp) pathogenic variant). Two of the subjects had an inherited variant from a parent, while the other two were de novo variants. Resolution of the hyperinsulinism occurred at 4 and 5.5 years in two subjects, while the other subjects remain on medication at 2.5 and 6 years of age.

The HNF4A gene controls expression of genes important for glucose-stimulated insulin secretion of the pancreatic beta cell. HNF4A variants cause macrosomia and congenital hyperinsulinism, which all of our patients had. These same HNF4A variants can cause maturity-onset diabetes of the young (MODY). Previous reports have described p.Arg63Trp HNF4A pathogenic variants in patients with hyperinsulinism and Fanconi syndrome, a generalized dysfunction of the renal proximal tubule. We suggest that clinicians be aware of the broad range of presentations of HNF4A gene variants.

OR6

Implementation of a Quality of Life and Mental Health Screening Tool in a Tertiary Care Pediatric Diabetes Clinic

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Psychosocial risk assessment in youth with type 1 diabetes (T1D) is recommended by both The International Society for Pediatric and Adolescent Diabetes and Diabetes Canada, however, there are no clear guidelines detailing which tools to use or how frequently to screen.

We describe the implementation of the Mind Youth Questionnaire (MY-Q), a validated psychosocial screening tool encompassing 36 items over several domains that assess quality of life, mood and eating disorders in youth with type 1 diabetes in our Pediatric Tertiary Care Centre.

Thirty-nine patients aged 13 to 18 completed the MY-Q from October 2019 to March 2020. The mean age was 15 years and 56% were female. 45.9% were on injection versus 54.1% on pump therapy; 50% were on continuous or flash glucose monitors. The mean A1c was 8%. 17.9% of patients had a pre-existing mental health disorder.

Seventy-five percent of patients debriefed with a physician and 25% with a social worker. Social workers completed the debrief if there was a flag on the mood domain. Mean time to complete debrief was 31.2 minutes overall, 21 for physicians and 57.3 for social workers.

Overall, 71.8% of patients flagged on at least one domain; the mean number of domains flagged was 3.43. Of patients who were flagged, 25% had pre-existing mental health diagnoses. "Family" and "body and weight" were most commonly flagged, each by 20 patients. Following debrief, no patients who flagged in the "body and weight" section were suspected to have an eating disorder. Patients who screened positively on the mood domain completed a validated depression screen (PHQ9A). Of the five patients who flagged on the mood domain, 40% scored positively on the PHQ9A. 50% patients were scheduled for a follow up social work visit and 22.2% were referred to outpatient services following debrief.

Psychosocial screening has not been widely adopted in youth with T1D, despite national and international recommendations. Ambiguity in the ideal screening tool, required resources and logistics of implementation are barriers. Based on our experience, implementation is possible without additional clinic resources and allows for the recognition and management of psychosocial issues of youth with T1D.

OR7

Elevated TSH in Children and Adolescents with Obesity: Retrospective Chart Review at Children's Hospital of Eastern Ontario's Centre for Healthy Active Living

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Subclinical hypothyroidism in children with obesity has been a growing area of interest as thyroid studies are frequently part of clinical assessments. The mechanism of thyroid stimulating hormone (TSH) changes in obesity remains elusive. Weight loss has been associated with normalization of circulating TSH. This study primarily aimed to determine the prevalence of elevated TSH among children and adolescents followed at the Children's Hospital of Eastern Ontario's Centre for Healthy Active Living (CHAL) from April 1, 2014 to March 1, 2020.

The medical records of a total of 391 (mean age 12.56 ± 5.57) cases were retrospectively reviewed. Those with preexisting underlying thyroid disease (primary, n= 12; secondary, n= 5) were excluded from the study population. Elevated TSH (> 3.5 mIU/L) was noted in 21.9% (n= 82) of the study population either at baseline (17.1%) or at any other time point over the course of 24 months of follow up.

Baseline BMI Z-score was not significantly different between those with elevated TSH (2.77 ± 0.65) (mean ± SD) versus study population (2.77 ± 0.69). No baseline difference in age (11.8 ± 3.7 vs 12.7 ± 2.1) or baseline BMI Z-score (2.82 ± 0.75 vs 2.66 ± 0.39) was noted among girls (n=64) and boys (n=28) with elevated TSH, respectively. To assess the course of thyroid function over 24-months of follow up, we examined TSH, fT4 and BMI z-score of those with baseline TSH > 3.5 at 12 months and 24 months (Table 1). Those with baseline elevated TSH demonstrated a significant drop in TSH at 12 and 24 months, while their fT4 and BMI remained unchanged (Table 1). Finally, only 2.4% (n=2) of the study population were treated for subclinical hypothyroidism.

In conclusion, moderate elevation of TSH is not an uncommon finding in children and adolescents with obesity and this study suggests that it does not require treatment in the majority of cases.

OR8

Impact of the COVID-19 pandemic on pediatric new-onset type 1 diabetes rates and severity of presentation in Montreal

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Background: Since the onset of the COVID-19 pandemic, anecdotal reports and a small number of published studies have documented a decrease in the number of patients with newly diagnosed type 1 diabetes (T1D), with a concurrent increase in the rates of diabetic ketoacidosis (DKA) at presentation.

Objectives: To determine rates and severity of DKA at diagnosis of new-onset T1D during the COVID-19 pandemic in Montreal and surrounding areas.

Methods: Retrospective review of pediatric new-onset T1D patients who presented or were referred to the Montreal Children's Hospital and CHU Sainte-Justine between 03/2019-08/2020. We documented age, presence and severity of DKA, DKA duration, and socio-demographic and anthropometric data. Rates and severity of DKA were compared between 03-08/2020 (pandemic) and the same period in 2019 (pre-pandemic).

Results: 81 and 72 patients were included from the pandemic and pre-pandemic periods, respectively, of whom 33 (41%) and 29 (40%) presented in DKA (p= 0.95), and 14 (17%) vs. 10 (14%) in severe DKA (p=0.56). In patients with DKA, during the pandemic vs. pre-pandemic, sex was male in 64% vs. 55%, mean age at diagnosis 9.1±4.2 vs. 10.1±5.3 years, mean weight Z-score -0.18±1.8 vs. -0.24±1.14, mean pH 7.14±0.12 vs. 7.18±0.11, and mean DKA duration 12.5±5.4 vs. 10.0±5.4 hours (all p>0.05). Sub-analysis comparing the strict confinement period (03-04/2020) and the post first-wave months (06-08/2020) to 2019 showed 15 vs. 27 new cases during 03-04/2020 vs. 03-04/2019. Of these, 6 (40%) vs. 11 (41%) presented in DKA (p= 0.96),and 4 (27%) vs. 5 (19%) in severe DKA (p=0.44). In 06-08/2020 vs. 06-08/2019, 55 vs. 34 new cases were diagnosed. Of these, 21 (38%) vs. 14 (41%) were in DKA (p=0.78), and 8 (15%) vs. 4 (12%) in severe DKA (p= 0.71).

Conclusion: DKA and severe DKA rates in the Montreal area are unacceptably high but did not significantly increase during the COVID-19 pandemic. During the first two pandemic months, half as many new pediatric T1D diagnoses were made as compared to 2019, followed by a surge of new diagnoses after the first wave. Fear of the virus and lockdown regulations may cause delays in new pediatric T1D diagnoses.

OR9

Patient and Family Perspective of a Pre-Transition Visit in a Pediatric Tertiary Care Diabetes Clinic

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Introduction: The need for an improved approach to transition from pediatric to adult care for youth with type 1 diabetes is evident. As part of a regional quality improvement initiative, a novel Pre-Transition (Pre-T) Visit was developed and piloted at a pediatric tertiary care centre in January 2018 for patients aged 15-18 years to capture the status of their self-management skills, introduce transition tools, and identify self-care goals and knowledge gaps to be addressed prior to transition.

Purpose: To evaluate patient and family satisfaction with, visit relevance of, and patient engagement with a novel Pre-T visit

Methods: From May 2019 to March 2020 a survey was offered to all youth who attended a Pre-T Visit and their parent(s)/guardian(s). Patient and family satisfaction with, relevance of and engagement with the Pre-T Visit were evaluated using a 5-point Likert scale. Multivariable regression was used to assess patient factors associated with patient level satisfaction.

Results: Of the 63 youth who participated in a Pre-T Visit, 60 completed the survey. Mean age (SD) of participants was 16.7 (0.8) years; 46.7% were female. Mean (SD) hemoglobin A1C (A1C) was 8.2% (1.8). Patients reported high levels of satisfaction (95% quite or extremely satisfied) that were consistent across age, A1C, gender and disease duration. Visit relevance and engagement were also rated highly by youth. Parent participants (n=27) also reported high levels of satisfaction and relevance.

Conclusions: Pre-T Visits were rated highly by patients and their parents. Their impact on glycemic control and health outcomes following transition requires further study.

OR10

Evaluation of virtual visits for the routine clinical care of transgender youth during the COVID-19 pandemic

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Background: During the current pandemic, continuing to deliver gender-affirming care has required innovative solutions. With the outbreak of COVID-19, our clinic rapidly shifted all routine visits to virtual care. We set out to evaluate the usability of these visits among transgender youth and to assess their interest in receiving ongoing virtual care.

Methods: an online survey was sent to trans youth and families who participated in a virtual gender visit between March and August 2020. It was developed by a multidisciplinary gender team and piloted with both trans youth and parents. The survey included the Telehealth Usability Questionnaire (Parmanto, 2016), and also asked about differences between in-person and virtual visits and preferences for the future. Consent to link responses with basic demographic and clinical information from medical records was obtained.

Results: 87 families completed the survey (39%). Mean patient age was 15.7 (IQR 13.7-17.7) years. 24% were receiving puberty blockers, and 52% hormone therapy. The vast majority were follow-up visits [median duration of follow-up 20 months (IQR 5-35)], while it was the first gender clinic visit for 15% of the participants. Most visits included both family members and transgender youth, and only 4% involved the parents alone. Median scores for all usability items ranged between "quite a bit" and "completely". 40% of participants were "quite a bit" and 48% "completely" satisfied with virtual visits. 41% felt that virtual appointments were safer than in-person, and 59% that these were equally safe, while none described virtual care as less safe. Building a relationship was reported as the most common challenge of using virtual care. 94% would like to have virtual visits continued after the COVID-19 pandemic; the majority would prefer 1 in-person and 2 virtual visits per year with a multidisciplinary gender team.

Conclusion: Virtual gender visits had impressive usability. Trans youth and their families are overall satisfied with virtual gender care, and they perceive this as safer than in-person. Virtual visits should continue to be a valuable tool for the provision of gender care in the future, with most families wanting a combination of virtual and in-person appointments.

OR11

Understanding the Role of Maternal Vitamin D supplementation in Early Infantile Rickets in Dhaka, Bangladesh

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Background: Vitamin D deficiency remains a global health concern, even in tropical regions where sun exposure is presumed adequate; however, controversy remains around its role in rickets in low-middle income countries. In a secondary post-hoc analysis of a large maternal vitamin D supplementation trial in Bangladesh, we aimed to estimate the effect of a range of maternal vitamin D supplementation doses, compared to placebo, on risk of infantile biochemical rickets at ages 6-12 months.

Methods: In the Maternal Vitamin D for Infant Growth (MDIG) trial in Dhaka Bangladesh, 1300 women randomized to one of five groups of vitamin D supplementation: placebo, 4200 IU/week, 16800 IU/week, or 28000 IU/week from 2nd trimester to delivery and 28000 IU/week prenatally and 28,000 IU/week 0-6 months post-partum. The primary outcome of this sub-study was 'biochemical rickets', defined as either 1) ALP >450U/L OR 2) ALP >350U/L and one of: calcium <2.2mmol/L or phosphate <1.6mmol/L or PTH >6.9pmol/L. Infants were included if they had alkaline phosphatase (ALP) measured between ages 6-12 months (n=790). For each vitamin D group versus placebo, we estimated the relative risk (RR) and 95% confidence interval (95%CI) of rickets.

Results: Overall, 39 of 790 infants had biochemical rickets. The highest risk was in the placebo group (7.8%), but prevalence among infants whose mothers received only pre-natal weekly vitamin D supplementation (4200 IU, 16800 IU and 28000 IU) were not significantly different: 3.8% (RR:0.48, 95%CI: 0.19,1.22), 5.8% (RR: 0.74, 95%CI: 0.33,1.69), 5.7% (RR: 0.73, 95%CI: 0.32, 1.65) respectively. However, infants whose mothers received weekly 28000 IU vitamin D3 prenatally and postnatally had a significantly lower risk of developing rickets compared to placebo (1.3%; RR:0.16, 95%CI: 0.03,0.72). Vitamin D deficiency (defined as 25-hydroxyvitamin D concentration less than 30nmol/L) was observed in 30/39 of infants with biochemical rickets on at least one occasion between birth and 12 months of age.

Conclusions: The risk of biochemical rickets was significantly lower in infants of mothers receiving both high dose maternal prenatal and postnatal vitamin D supplementation, versus placebo. Further research is needed to define optimal postpartum vitamin D supplementation dosing in lactating mothers in this population.

OR12

Adrenal Insufficiency among Children treated with Hormonal Therapy for Infantile Spasms

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Background: Hormonal therapy is a standard treatment for children with infantile spasms. However, the high doses given and long treatment duration exposes patients to the potential risk of AI. There is no literature recommending any test or hydrocortisone replacement following the hormone treatment even if we don't have any data on the occurrence of adrenal suppression following that treatment and even if the consequences of that adrenal suppression can be severe. A case of an adrenal crisis at our center motivated us to look further into the risk of developing AI in such patients.

Objective: To quantify the incidence of adrenal insufficiency (AI) among children with infantile spasms treated with high-dose corticosteroids and/or adrenocorticotropic hormone (ACTH).

Design and Method: A retrospective chart review of patients treated for infantile spasms was performed between January 2009 to March 2020 in our hospital. Variables collected included sex, age, etiology, hormonal treatment, adrenal function testing and signs of AI. Analysis included descriptive statistics such as incidence.

Results: 29 patients met the inclusion criteria and received hormonal treatment (19 received corticosteroids [prednisone/prednisolone], 6 received ACTH and 4 received both). Post-hormonal treatment testing of adrenal function was obtained in 26/29 (90%) patients. AI occurred in 20/26 (77%) of children who had testing. One patient presented to the emergency room with an acute adrenal crisis, the day following the weaning off of hormonal treatment.

Conclusion: Our study suggests that adrenal suppression is frequent after standard hormonal therapy regimen for infantile spasms. This can lead to serious complications, such as adrenal crisis, if not supplemented. A routine laboratory assessment of adrenal function should be considered after hormonal therapy for all patients. We suggest hydrocortisone supplementation therapy should be given at the end of hormonal therapy and until testing results for adrenal function are obtained.

Poster Abstracts

P1

A structural equation model of factors associated with prevalent albuminu-ria in youth with type 2 diabetes in the iCARE national cohort

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Introduction: Youth living with type 2 diabetes (T2D) are at high risk of early renal injury. We evaluated interrelationships between biological, psychological, socioeconomic factors and preva-lent albuminuria in youth with T2D in the Improving renal Complications in Adolescents with type 2 diabetes through REsearch (iCARE) cohort.

Methods: We performed a cross-sectional structural equation model (SEM) analysis on 319 youth with T2D 10-24 years old from 8 Canadian sites involved in the iCARE cohort. The main outcome measure was non-orthostatic albuminuria (urine ACR >2mg/mmol). Main factors of interest (indi-cator variables) were blood pressure (daytime and night systolic and diastolic loads), urine cy-to-kines (IL-6, RANTES, Fractalkine, ENA78, TNF□, IL-1□B, sTNFRI51, sTNFRII53, MCP1), systemic inflammatory markers (c-reactive protein (CRP), erythrocyte sedimentation rate (ESR), fibrinogen), mental health (Resiliency Scales for Children and Adolescents, Kessler-6, Perceived Stress Scale-14), and the Canadian Index of Multiple Deprivation (CIMD) scores. Additional covariates were hemoglobin A1c (HbA1c) and duration of diabetes. Multiple imputation addressed missing data.

Results: Youth were 15 ± 2.3 years old, lived with diabetes for 2.3 ± 1.9 years, 66% were female, and 33% had albuminuria. Factors significantly associated with albuminuria were blood pressure (standardized coefficient, b=0.31), urinary cytokines (IL-6, RANTES, Fractalkine, ENA78, TNFa, IL-1□) (b=0.34) and HbA1c (b=0.32); Residential Instability (b=0.21) and ethnic composition (b= -0.36). The factor mental health was positively correlated with systemic inflammation (CRP, ESR, fibrinogen), which was predictive of albuminuria (b=0.29). Diabetes duration was not associated with albuminuria. The model fit well per the CFI (0.988), RMSEA (0.012), and SRMR (0.057) statis-tics.

Conclusions: The presence of albuminuria in youth with T2D is associated with blood pressure, HbA1c, renal and systemic inflam-mation and residential instability. Mental health was indirectly associated with albuminuria via systemic inflammation. Albuminuria is complex and requires mul-ti-pronged approaches for prevention.

P2

Incidental vertebral fractures in patients with Duchenne muscular dystrophy on intravenous bis-phosphonate treatment

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Background: Children with Duchenne muscular dystrophy (DMD) have an increased risk of verte-bral and long bone fractures due to long-term corticotherapy, delayed puberty, linear growth sup-pression and immobilization. Intravenous bisphosphonates are used to treat osteoporosis in these patients, but the evidence supporting their efficacy on the prevention of incidental or recurrent fractures in DMD patients remains scarce.

Objectives: To describe the progression of vertebral fractures (VF) and bone mineral density (BMD) in a cohort of osteoporotic DMD treated with intravenous bisphosphonates.

Methods: Retrospective review of a cohort of DMD patients treated with zoledronic acid (ZA) for at least 12 months at CHU Sainte-Justine since 2008. Age at ZA initiation, ambulatory status, cor-ticosteroid duration and doses, ZA doses and durations (total and until first incidental VF), other medications, yearly anthropometric data, BMD, bone biomarkers and VF assessed by Genant clas-sification were documented.

Results: Twenty-one boys were included. Prior to the initiation of ZA, the median age was 13.5 years (10.6– 14.5), five (24%) were ambulant, median corticotherapy duration was 6.1 years (3.8- 6.9), median lumbar BMD Z-score was -2.4 (-3.2- -1.4), and 16 (76%) had at least 1 prevalent VF. Median total duration of ZA therapy was 44 months (31.0- 65.0). Median change in BMD lumbar Z-score at the end of ZA treatment was -0.2 (-0.9- 0.7). In 11 boys (52%), 22 incidental VF (either in previously normal vertebral bodies or worsening of pre-existing VF) were detected, occurring 21 months (10- 39) after ZA initiation. Thirteen of these VF occurred in 7 boys (63%) beyond 1 year of ZA treatment.

Conclusion: We observe a high incidence of new or worsening VF in DMD patients beyond the first year of ZA treatment, suggest-ing that bisphosphonate therapy does not eliminate the risk of VF when treatment is started after clinical evidence of osteoporosis.

P3

De novo mutation of IGF2 gene in a patient with short stature

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Background: Historically, IGF-2 was thought to drive fetal growth, while IGF-1was largely responsible for postnatal growth. Newly discovered mutations in IGF2 have been implicated in both prenatal and postnatal growth restriction, however the cases are few in number with varying phenotypes. There is significant overlap with Russel Silver syndrome and both map to 11p15.5, a known imprinted region. We present a case of a de novo IGF2 mutation in a patient with short stature, microcephaly, developmental delay, and dysmorphic features.

Case Presentation: An 8 year old male presented for assessment of short stature. He was born at 36 weeks gestational age via induced vaginal delivery for poor in utero weight gain with a birth weight of 1.9 kg and required NICU admission for nutritional support. In early childhood, he had developmental delay in speech and gross motor function. He has academic challenges in school and was diagnosed with ADHD on stimulant therapy. There was a long-standing history of poor appetite. He had orchidopexy for right cryptorchidism. Historical screening for chronic disease and endocrinopathies was otherwise negative.

Midparental height was between the 25th-50th percentile. He was proportionate with height 122.1 cm (4.30 percentile, Z -1.72) and weight 18.7 kg (0.12 percentile, Z -3.03). Exam was significant for dysmorphisms including microcephaly (Z -2.9), prominent eyebrows, mild synophrys, flattened nasal bridge, and pointed chin. His bone age was concordant. Growth velocity was 3.97 cm/year at first reassessment and between 4-5 cm/year with later assessments. Work up including chronic disease screen, endocrinopathy screen, and growth hormone stimulation were negative. Chromosome microarray and specific Russell Silver testing was normal. Whole exome sequencing showed a de novo paternally derived pathogenic variant in the gene IGF2: c.1456G>A, p.Gly49Ser. He has since entered puberty and achieved a growth velocity of 9.76 cm/year, without growth hormone therapy.

Conclusions: We present a newly described sporadic mutation in IGF2 in a patient with prenatal growth restriction, and an element of post natal growth restriction. This adds to existing case reports of patients with mutations in IGF2 to further characterize the variety of phenotypes.

P4

Assessing the need for teaching of Tanner stages of breast development using 3D breast models

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Tanner staging of breast development is a standardized method that allows practitioners to define the stages of puberty and help identify pubertal progression. This can impact clinicians' decision making and their path of investigations. In medical education, it is challenging to determine the best modality of training to ensure both accuracy and confidence when performing sensitive examinations. Currently there are no standardized simulation sessions for tanner staging of breast development in the medical curriculum for residents at the University of Ottawa. The goal of this study was to assess the confidence and accuracy of tanner staging of breast development of cur-rent pediatric, obstetrics and gynecology (OBGYN), pediatric endocrinology, reproductive endocri-nology and infertility (REI), and pediatric and adolescent gynecology (PAG) residents at the Univer-sity of Ottawa. Secondly, we aimed to identify the need for a formalized simulation session using 3D models as representations of the tanner stages. A needs assessment survey of current resi-dents in the aforementioned programs was completed between October and December 2020 using RedCap software. Statistical analysis looked at proportions of total and subgroup respons-es. Twenty-one pediatric, 5 OBGYN, 4 pediatric endocrinology, 1 REI and 1 PAG residents complet-ed the survey. The current confidence level in performing tanner staging was highest amongst pe-diatric endocrinology residents (50% confident, 25% very confident, 25% somewhat confident, 0% not confident), who also use tanner staging most frequently (75% weekly, 25% daily). Only forty percent of respondents accurately identified a tanner stage based on a written description. Eighty-eight percent of participants thought it would be beneficial to received further training. Sixty-three percent of respondents' preferred method of training would be simulation-based, 25% didactic, 47% online modules, 66% bedside teaching and 9% textbook learning. Eighty-eight and 90% of re-spondents felt that a simulation using 3D breast models would improve their accuracy and confi-dence, respectively, of tanner staging of breast development (86% pediatrics, 100% OBGYN, 75-100% endocrinology, 100% REI, 100% PAG). It appears that there is a need for further teaching of tanner staging of breast development using 3D models for current residents at the University of Ottawa, to improve both accuracy and confidence with these skills.

P5

Severe Hypertriglyceridemia Complicating Pediatric Acute Lymphoblastic Leukemia Treatment: A Need for Monitoring Guidelines

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Severe hypertriglyceridemia is rare in the pediatric population however, the incidence is higher in pediatric oncology patients. Severe hypertriglyceridemia is defined as a triglyceride level >11 mmol/L and is associated with an increased risk of pancreatitis. Although, the etiology of hypertriglyceridemia is multifactorial several chemotherapeutic agents including PEG-asparginase and glucocorticoids have been reported to cause elevations in triglyceride levels. However, the hypertriglyceridemia is often transient and mild, requiring no specific treatment.

We present two children who experienced severe hypertriglyceridemia, a rare complication of chemotherapy during treatment for acute lymphoblastic leukemia (ALL). As there are no established guidelines with regards to the recommended management of severe hypertriglyceridemia in pediatric or pediatric oncology patients, pediatric endocrinologists must rely on anecdotal experiences in making treatment decisions.

The first patient, an 11-year-old female with T-cell ALL receiving induction chemotherapy presented to a clinic appointment with a one-week history of feeling unwell, increased fatigue, weakness, abdominal and back pain. She was admitted to hospital and found to have lipemic serum and an elevated triglyceride level of 80.5 mmol/L.

The second patient, a 7-year-old male with relapsed B-cell ALL receiving maintenance chemotherapy, was found on routine investigations for follow-up of hemophagocytic lymphohistiocytosis (HLH) to have elevated triglycerides of 26 mmol/L.

Both patients were admitted to hospital for acute management of their hypertriglyceridemia. This included several days of fasting, in addition to insulin and dextrose containing intravenous fluids, given their increased risk of pancreatitis. They were both then transitioned to a low-fat diet and started on fibrate therapy as well as omega-3 fatty acids. However, there were concerns in starting fibrate therapy given both patients had evidence of transaminitis. Both patients experienced weight loss and faced nutritional concerns given the restricted diet, which complicated their therapy. Neither patient experienced pancreatitis and with dietary changes and triglyceride lowering medications both patients continued chemotherapy with triglycerides below 9 mmol/L.

These two cases highlight a rare complication of chemotherapy in pediatric patients, as well as additional considerations that complicate the management of severe hypertriglyceridemia in pediatric oncology patients. Guidance regarding monitoring and management of hypertriglyceridemia in this context is needed.

P6

Exploring Medical Students' Perceived Competence and Comfort Performing Physical Examinations on Patients with Obesity: A Qualitative Study

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Background: There has been a dramatic increase in the prevalence of obesity within the last few decades, and currently one third of Canadians meet criteria for being overweight or obese. Despite the rise in demand, studies have shown that many physicians express low competency in the skills needed to appropriately assess and manage patients with obesity. The physical exam is a key component of any clinical assessment and an important diagnostic tool for detecting pathology but can present a particular challenge in these patients. This study set out to explore medical stu-dents' experiences and feelings of comfort and competency in conducting physical examinations on patients with obesity.

Methods: This was a qualitative, mixed-methods study using questionnaires and semi-structured focus groups. Study recruitment occurred via a convenience and snowball sampling method at The Hospital for Sick Children. Four focus group interviews were conducted from August 2018 to Janu-ary 2019, with a total of 23 undergraduate medical students participating. Medical students were stratified based on their year of training. Interviews were transcribed verbatim and thematic analy-sis was performed, accord-ing to Braun & Clark's six-phase framework.

Results: Medical students across all years of training described low levels of confidence and com-petence in examining patients with obesity, compared to patients with normal body mass in-dex. Important challenges identified by trainees included modifying physical exam maneuvers, identifying abnormal findings, and sensitive communication around patients' body habi-tus. Participants identified lack of exposure to patients with obesity, lack of instruction by pre-ceptors, and lack of curriculum focus as key barriers to developing these skills. Furthermore, medical students' discomfort with such physical exams was perceived to have a negative impact on patient care.

Conclusions: This study shows that adequate training on the physical examination and over-all approach to care of patients with obesity is currently lacking. Given the increasing rates of obesity in both adult and pediatric populations, developing curricula and including more formal teaching around these key competencies within medical education is necessary.

P7

Are you sure that it is a Pituitary Macroadenoma?

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The most common causes of large sellar masses are pituitary adenomas and craniopharyngioma. Some rare etiologies can have a similar presentation, resulting in diagnostic challenge.

A 16-year-old female presented with a 5-month history of daily headaches, low energy and 9 month history of secondary amenor-rhea. She did not have visual changes, polyuria or polydipsia. MRI showed a 1.5 cm pituitary mass expanding the sella and ex-tending into suprasellar cistern with mild mass effect on optic chiasm, felt to be a pituitary macroadenoma. Investigations showed deficiency of all anterior pituitary hormones. The patient was started on hydrocortisone, thyroxine and estrogen. As she had already reached final adult height, growth hormone treatment was not initiated. 2 months later, she noted polyuria and polydipsia. Diagno-sis of central diabetes insipidus was confirmed. With the additional diagnosis of central diabetes insipidus, the diagnosis of cranio-pharyngioma was considered to be most likely. CT scan showed no calcifications. Decision was made to proceed with transsphenoi-dal surgical resection. Pathology showed granulomatous hypophysitis with normal IgG levels. Secondary causes of granulomatous hypophysitis were ruled out. Repeat MRI post debulking surgery showed decrease in size of pituitary mass and she had resolution of headaches. Anterior and posterior pituitary deficiencies persisted post-surgery.

Hypophysitis is a rare cause of hypopituitarism with an incidence of approximately 1 in 9 million, of which <1% of cases are in the pediatric population. Granulomatous hypophysitis classically presents with headache and a range of pituitary hormone deficiencies with majority of cases having central diabetes insipidus. MRI finding is similar to adenoma and craniopharyngioma with homoge-nous pituitary enlargement and symmetrical suprasellar expansion. This case illustrates a rare etiology of sellar mass and highlights the diagnostic challenge radiologically, where it can easily be misdiagnosed as a macroadenoma. In cases with atypical presentation of a sellar mass, other rare etiologies should be considered and surgery for diagnostic purposes may be required.

P8

Use of Lanreotide in Infancy for Severe Hyperinsulinism

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Introduction: Pancreatectomy is often recommended for severe congenital hyperinsulinism unresponsive to diazoxide and glucose-enriched feeds. We present a unique case of medical management of severe hyperinsulinism in an infant with Beckwith-Wiedemann syndrome (BWS) and mosaic genome-wide paternal uniparental disomy (GWpUPD).

Case: A 3.44kg female infant was born at 38 weeks to a G4P2 mother. Exam was notable for hemihypertrophy of the right arm, leg and labia. Bloodwork revealed hyperinsulinemic hypoglycemia, and abdominal ultrasound showed hepatomegaly with right adrenal gland enlargement. Targeted molecular testing revealed UPD of chromosome 11, consistent with BWS.

A glucose infusion rate (GIR) of 15 mg/kg/min was required to maintain normoglycemia, and oral diazoxide 15 mg/kg/day was started at 4-days-old. Diazoxide was stopped after four weeks due to a pulmonary hypertensive crisis. Subcutaneous octreotide 20mcg/kg/day was trialed at 1.5 months of age with no effect. After extensive discussion, the parents declined transfer to a specialized center for pancreatectomy. Subcutaneous lanreotide (60mg) was started at 2.5 months of age, allowing slow weaning of intravenous fluids, and continued every four weeks. The infant was discharged home at 5-months-old on continuous fortified gastrostomy feeds (GIR 8.6 mg/kg/min).

A genetic panel for other causes of hypoglycemia revealed a likely pathogenic variant in PGM1, consistent with a congenital disorder of glycosylation, which prompted addition of enteral galactose 0.5 g/kg/day to her regimen. Additionally, mosaic GWpUPD was detected, suggesting the possibility of other autosomal recessive or imprinted disorders in addition to BWS.

Currently, the infant is 9.5-months-old and developmentally appropriate, with weight-for-length at the 99th percentile. She remains on lanreotide, galactose, and fortified feeds. Based on a continuous glucose monitoring system (CGMS), her time spent in hypoglycemic range (less than 3.0) is three percent. She has not had any rehospitalizations or gastrointestinal side effects from lanreotide.

Discussion: This is one of the youngest reported cases of lanreotide use in severe hyperinsulinism and suggests that lanreotide may be a safe and effective alternative to surgery in selected cases. CGMS may be a useful adjunct in the care of these infants. Close monitoring for adverse medication effects and neurodevelopmental sequelae of hypoglycemia remains necessary.

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P9

Flip-Flopping Function in a Transplanted Teen with a Tricky Thyroid

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A 15-year-old female, two years status-post hematopoietic stem cell transplant for aplastic anemia, presented to the emergency department with suicidal gestures and tachycardia. Laboratory investigations were consistent with thyrotoxicosis (TSH 0.01 mIU/L, free T4 32 pmol/L [10.0-17.6 pmol/L]). Past treatment exposures included cyclosporin, prednisone, antithymocyte globulin and alemtuzumab, a monoclonal antibody directed against CD52+ cells. Thyroid Stimulating Immunoglobulins (TSIg) were borderline positive (143% [< 140%]), and TSH Receptor Antibody (TRAb) titres were subsequently strongly positive, consistent with Graves' disease. Methimazole was initiated. Two weeks after initiating therapy, symptoms resolved and free T4 had decreased from

32 pmol/L to 9.5 pmol/L, while TSH remained suppressed. Over the next two months, hypothyroidism ensued (TSH 51 mIU/L, free T4 8.3 pmol/L), despite weaning and ultimately discontinuation of methimazole. Thyroid Blocking Immunoglobulins (TBIg) were then found to be elevated (66% [< 34%]) and TSIg were normal (81% [< 140%]), confirming that she had both stimulating and blocking antibodies. As such, the conversion from Graves' hyperthyroidism to hypothyroidism was likely mediated by a change in the proportion of stimulating to blocking TRAbs. She has since remained hypothyroid with a 36-month follow-up duration requiring levothyroxine supplementation.

We speculate that this young woman's prior exposure to alemtuzumab was etiologically related to her autoimmune thyroid dysfunction, as has been previously reported among individuals receiving alemtuzumab for treatment of multiple sclerosis. We will discuss this in the context of existing literature describing this effect in adults and children.

This case highlights the functional interplay of TSH receptor activating and blocking antibodies in mediating thyroid function and stresses the importance of monitoring thyroid function in patients treated with alemtuzumab.

P10

A novel germline GNAS variant causes Nephrogenic Syndrome of Inappropri-ate Antidiuresis as part of an emerging multisystem phenotype

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Introduction: GNAS mutations cause a spectrum of phenotypes from hyperfunctioning endocrinopathies in McCune-Albright Syndrome to hormone resistance in Albright's Hereditary Osteodystrophy. Lately, a new condition with hyponatremia, precocious puberty and skeletal abnormalities has been de-scribed (Bieberman et al, 2019) as well as a nephrogenic syndrome of inappropriate diuresis (NSI-AD; Miyado et al, 2019) due to germline GNAS dominant mutations. We describe a three generation family with this newly proposed genetic condition due to a novel GNAS variant, expanding on the variability of the phenotype.

Case Presentation: The index case, 9-year-old male was referred for subclinical hyperthyroidism. Past medical history included speech delay and early childhood hyponatremic seizures consistent with NSIAD (low/normal ADH with high urine osmolality), responsive to fluid restriction. Family history in-cluded hyponatremic seizures and tall stature in his mother and neonatal hyponatremia and speech delay in his younger brother. Height was >99th percentile (z=+3.33) and weight was at the 98th percentile. Thyroid was unremarkable, and he was not in puberty. There was one hyperpig-mented patch on his left chest. TSH was 0.08 (0.3-5 mU/L) with free T3 of 8.0 (4.1-7.9 pmol/L) and free T4 of 15.3 (8.0-20.0 pmol/L). TSH receptor and TPO antibodies were negative; AVPR2 testing was normal. Tc-99m-pertechnetate thyroid scintigraphy demonstrated diffuse uptake. Skeletal survey identified bilateral acro-osteolysis in the distal phalanges of the feet and hands and subtle erosive changes along the medial metaphysis of the distal left femur. Whole exome se-quencing identified a new variant in the GNAS gene, shared with the affected brother, mother, and apparently healthy maternal grandfather.

Discussion: The GNAS gene encodes the stimulatory G protein alpha-subunit (Gαs), which mediates the sig-nalling of G protein-coupled receptors, including arginine vasopressin receptor 2 (AVPR2). Our case highlights a family with a new variant in GNAS associated with NSAID, non-autoimmune sub-clinical hyperthyroidism, and skeletal abnormalities. The variable intrafamilial phenotype in mem-bers sharing the same genotype expands on the clinical features previously described in the litera-ture. Importantly, this case further refutes the widely believed notion that germline GNAS gain-of-function mutations are lethal and contributes to our understanding of this emerging GNAS-mediated multisystem disease.

P11

Congenital lipoid adrenal hyperplasia: a novel StAR mutation and atypical presentation

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Background: Congenital lipoid adrenal hyperplasia (lipoid CAH) is a severe, rare defect in steroid biosynthesis caused by mutations in steroidogenic acute regulatory protein (StAR) - necessary for cholesterol transport into mitochondria. Patients have adrenal and gonadal steroid deficiencies and present phenotypically female with adrenal crisis typically in the neonatal period.

Case: A previously well 6-month-old female, born at term, presented with vomiting, weight loss, lethargy and dehydration. Parents were non-consanguineous with no family history of adrenal disease or early infant mortality. Initial investigations: sodium 122 mmol/L (133-142), potassium 7.7 mmol/L (3.5-6.0), glucose 2.5 mmol/L (3.9-6.0), creatinine 83 μmol/L (8-49). Further investigations: random cortisol 20 nmol/L, ACTH >330.3 pmol/L, DHEAS 0.1 μmol/L (0.2-4.8), renin >550 μIU/ml (5.3-99.1), 11-deoxycortisol <0.6 nmol/L, 17-hydroxypregnenolone <0.6 nmol/L, 17-hydroxyprogesterone <0.6 nmol/L, pregnenolone <0.6 nmol/L. Ultrasound: normal adrenal glands. Karyotype: 46, XX. Genetic panel (Blueprint Genetics, Finland) showed two pathogenic mutations in STAR: c.824T>C p.(L275P) and c.144G>A p.(Trp48*) for which her parents are carriers. Current treatment: 0.1 mg/day fludrocortisone and 8 mg/m2/day hydrocortisone.

Discussion: We describe a female with genetically confirmed lipoid CAH, who presented with adrenal crisis, acute kidney injury, but atypically delayed presentation, normal ultrasound findings, and novel nonsense StAR variant (Trp48*). Ultrasound showing adrenal cholesterol accumulation is a characteristic feature of lipoid CAH; however, is not a universal finding. Patients typically present within one month of birth but can present up to age 14 months.

L275P is previously described and causes impaired binding of cholesterol to StAR leading to decreased steroid biosynthesis. Homozygous L275P patients have presented at 4.5 and 11 months, suggesting some residual enzyme activity. This variant has never previously been reported along with a nonsense variant. The fact that our patient's phenotype is similar to the homozygous patients suggests that less residual enzyme activity is needed to delay presentation timing from neonatal to infantile, than previously thought.

Lipoid CAH has a broad clinical spectrum. Minimal residual enzyme activity likely explains her delayed presentation. A normal adrenal ultrasound could represent early lipoid CAH pathophysiology when intracellular lipid droplets have not yet accumulated, and mitochondrial cholesterol transport is facilitated by StAR independent mechanisms.

P12

Could Multiple Pituitary Hormone Deficiencies Be Within the Spectrum of Disease in Aicardi Syndrome?: A Case Report

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Aicardi syndrome is a presumed X-linked disorder that presents in females with the triad of infantile spasms, dysgenesis or agenesis of the corpus callosum, and chorioretinal lacunae. Although children with dysgenesis or agenesis of the corpus callosum in septo-optic dysplasia can have multiple pituitary hormone deficiencies, the corpus callosum abnormality itself has not been shown to predict hypopituitarism. In Aicardi syndrome, there is limited information regarding pituitary abnormalities. Precocious puberty has been reported. We present a case of a 9-year old girl with Aicardi syndrome who initially presented with hypotension, hypothermia, and respiratory failure. She was enrolled in a clinical trial comparing placebo with glucocorticoid, but she had refractory hypotension and a cortisol level of 36nmol/L. She was withdrawn from the study and started on intravenous hydrocortisone; her hypotension resolved. Thyroid function tests were normal at discharge. She continued on physiologic hydrocortisone with a plan to taper it, however, within 2 months, she presented with seizures, hypothermia, hypotension and polyuria. Her sodium was 169mmol/L, serum osmolality was 327mOsm/kg, and urine osmolality of 98mOsm/kg, consistent with diabetes insipidus. She responded well to DDAVP. Free T4 was 7pmol/L (13-22) and TSH was 1.03mIU/L (0.6-4.84). Levothyroxine was initiated for treatment of central hypothyroidism. Given this clinical picture, a diagnosis of presumed central adrenal insufficiency was given. Magnetic resonance imaging of her head demonstrated a normal pituitary gland and infundibulum. The patient has progressed from Tanner stage 2 to 3 breast development over the 9 months since presentation. Luteinizing hormone (LH) and Follicular Stimulating Hormone (FSH) were 2.7IU/L and 6.7IU/L at initial presentation, and 9 months later were 1.24IU/L and 6.3IU/L, respectively. Estradiol has been consistently undetectable. Her growth velocity has been 3.7cm/year with a normal IGF-1 of 212ug/L (62-504), indicating potential growth hormone deficiency. Prolactin levels have been normal. She remains on maintenance hydrocortisone, levothyroxine, and DDAVP. Multiple pituitary hormone deficiencies may be within the spectrum of disease for girls with Aicardi syndrome. It is important to monitor for and recognize signs of pituitary dysfunction in children with Aicardi syndrome to ensure prompt assessment and management.

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P13

Antibody-negative insulinopenic diabetes mellitus following CAR T-cell therapy for pediatric acute lymphoblastic leukemia

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A previously normoglycemic 3-year-old boy undergoing treatment with CAR (chimeric antigen receptor) T-cell therapy for multiple-relapsed ALL presented with diabetic ketoacidosis on day 5 of therapy. Glycemic control had been normal until polydipsia and mild polyuria were noted the day prior; random blood glucose at the time was 12mmol/L and normalized without intervention to 7.5mmol/L. Otherwise, there was no notable prodrome. Within 24 hours, he developed hyperglycemia (glucose 22.9mmol/L) and acidosis (pH 7.15, HCO3 13 mEq/L) with large ketonuria. DKA management was initiated. He was briefly admitted to the PICU, as cytokine release syndrome was also suspected. He recovered swiftly, and transitioned to subcutaneous insulin therapy, as hyperglycemia persisted.

Past medical history is remarkable for infantile ALL, diagnosed at 10 months and subsequent relapse treated with allogeneic stem cell transplant. There is neither history of maternal gestational diabetes, nor a family history of diabetes. Investigations revealed undetectable pancreatic autoantibodies (GAD-65, IAA, ICA) and undetectable C-peptide in the context of hyperglycemia. An abdominal ultrasound obtained 2 weeks after this presentation showed a structurally normal pancreas. In the preceding months, there had been no steroid exposure. He received chemotherapy (fludarabine and cyclophosphamide) in preparation for CAR T-cell therapy; however, these agents are not associated with glucose excursions. Six months following the onset of hyperglycemia, he continues to require insulin (~0.8u/kg/day).

CAR T-cell therapy has been approved for the treatment of childhood ALL since 2017; it is a form of targeted immunotherapy using the patient's T-cells, genetically altered to target CD19 antigen expressed on leukemic cells. Cytokine release syndrome (systemic inflammatory response), transient neuropsychiatric effects and B-cell aplasia are the most common side effects. Organ damage could hypothetically occur when CAR T-cells cross-react with an antigen expressed on normal tissue that is similar to the target antigen; however, this has not been reported in clinical trials to date. To our knowledge, antibody-negative diabetes mellitus requiring insulin therapy following CAR T-cell therapy has not previously been reported. As this is a relatively novel therapy in pediatrics, clinicians should be aware of this possibility in these fragile patients, particular as metabolic decompensation progressed rapidly.

P14

A Rare Presentation of New Onset Hypoglycemia in an Adolescent Male

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Introduction: Functional insulinomas are a type of pancreatic neuroendocrine tumor rarely encountered in the pediatric population. They may occur as sporadic, solitary lesions or in the setting of Multiple En-docrine Neoplasia Type 1 (MEN-1).

History: A 13-year-old Caucasian male presented to the emergency room with a first episode of hypogly-cemic seizure. There was no history of ingestion, intercurrent illness, previous episodes of docu-mented hypoglycemia or seizures. He had last eaten 13 hours prior to the episode. There was no access to oral anti-glycemic agents and the child was otherwise healthy. The family did, however, describe an approximately 2-month history of morning nausea, anxiety and pallor, which always improved with food. A critical sample obtained after 6 hours of fasting revealed endogenous hy-perinsulinemia: venous blood glucose 2.2 mmol/L, insulin level 120 pmol/L, C-peptide 781 pmol/L, B-OH-butyrate negative. Growth hormone, cortisol, and additional metabolic screening labs were within normal limits. A urine toxicology screen and insulin-antibodies were negative. An abdominal MRI was read as unremarkable. He was transferred to a tertiary care pediatric center for further work up of hyperinsulinemia. At the receiving hospital, a mixed meal tolerance test did not reveal post-prandial hypoglycemia and he passed a safety fast of 10 hours. Screening for evidence of other endocrinopathies (Prolactin, IGF-1, PTH and calcium) was negative. He underwent a 111indium pentetreotide scan (OctreoScan) that revealed focal uptake within a well-defined 1.8 x 2.5 cm ovoid lesion in the distal tail of the pancreas. A laparoscopic distal pancreatectomy was performed and surgical histopathology confirmed the diagnosis of a well-differentiated insulin-producing neuroendocrine tumor. Post-operatively our patient has had no hypoglycemia and we are awaiting the results of MEN1 genetic testing.

Discussion: The differential diagnosis of new-onset endogenous hyperinsulinemia in a well adolescent in-cludes sulfonylurea ingestion, insulinoma, late-onset congenital hyperinsulinemia, insulin-autoimmune hypoglycemia and non-insulinoma pancreatogenous hypoglycemia syndrome (NIPHS). Hyperinsulinemia confirmed on a critical sample combined with Whipple's triad is highly suggestive of an insulinoma, however, there remain challenges in confirming this diagnosis due to the relative unavailability of sensitive, specific functional imaging, particularly when conventional cross-sectional imaging is non-diagnostic.

P15

A Rare Case of Differences of Sexual Development

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Introduction: Persistent Mullerian duct syndrome (PMDS) is a rare difference of sex development characterized by persistence of Mullerian duct derivatives in a genotypic (46 XY) and phenotypic normally virilised male. PMDS is transmitted in an autosomal recessive manner, caused by mutations in the anti-Mullerian hormone gene or in the gene encoding the AMH receptor. In 12% of the cases, no mutation of AMH or AMHRII has been detected, suggesting a disruption of other pathways involved in mullerian regression. Only a few cases have been reported in the literature.

Case report: A 9-days-old phenotypic male presented with left scrotal swelling at day 8 of life. The scrotum was firm, tender, and erythematous with palpable masses extending from the left inguinal canal to the left scrotum. He has normal stretched penile length 3.6 cm, complete fusion of the labioscrotal folds, bilateral cryptorchidism and a urethral opening at the tip of the penis. He had no other dysmorphic features. He was admitted for 24 hour for observation and evaluation. The ultrasound showed 2 gonads (presumed testis) and a uterus herniating into the left scrotum. He had a normal newborn screen, including 17-OHP. He also had FISH for X and Y probe. All of the cell lines are consistent with 46,XY complement. On day 10 of life he had the following: LH 1.4 IU/L, FSH 1.2 IU/L, Testosterone 5.37 nmol/L, Estradiol <15 pmol/L, and low AMH 19 pmol/l. Based on the above, he was diagnosed with persistent Mullerian duct due to AMH deficiency. The microarray is normal. Single gene testing showed heterozygous mutation in the AMH gene c.1518C>G. There is no history of consanguinity. The older brother has a hydrocele.

Conclusion: PMDS is a rare disease and is often misdiagnosed due to a lack of familiarity with the condition. In cases of unilateral or bilateral cryptorchidism associated with inguinal hernia, as in our patient's case, the possibility of persistent Mullerian duct syndrome should be kept in mind. Genetic testing could help screening family members who are mildly affected. In our case his brother who had hydrocele might be more subtly affected, so knowing the mutations might help with screening the family.

P16

The Impact of the COVID-19 Pandemic on Adolescents and Young Adults with Type 2 Diabetes

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Background: The coronavirus disease (COVID-19) pandemic has had a widespread impact. The ob-jective of this study was to assess the impact of the pandemic on adolescents and young adults living with type 2 diabetes (T2D) involved in the national Improving Renal Complications in Adolescents with type 2 diabetes through REsearch (iCARE) study.

Methods: The Environmental influences on Child Health Outcomes (ECHO) COVID-19 Question-naire developed by the National Institutes of Health ECHO COVID-19 Task Force was administered to participants (n=82) from the iCARE study in Canada between June 4, 2020 to October 31, 2020 on children aged 12 years old (via parent report), and adolescents and young adults aged 13 years and older (via self-report). The questionnaire was administered over the phone by trained research personnel. The questionnaire assessed the impact of the COVID-19 pandemic on healthcare ap-pointments, lifestyle, social connections, mental health, school, internet usage, and food security.

Results: Participants were 17.8 ± 3.3 (12-27) years of age and predominantly female (62.2%). The cohort had a mean HbA1c of 9.6 ± 2.7% prior to the pandemic. None of the participants had tested positive for COVID-19 or had close contact with a case of COVID-19. However, 12.2% had experi-enced symptoms suspicious of COVID-19 and 2.5% received testing with negative results. During the pandemic, 35.3% were unable to see their healthcare providers, 61.0% of youth spent less time outside, and 64.6% felt less socially connected. Stress and anxiety were also reported by the par-ticipants, with 46.1% experiencing a mild level of severity; 43.4% reported the COVID-19 pandemic as having a negative impact. Due to school closures, 81.4% of youth were offered online learning and 42.7% spent more time on the internet for educational purposes. Additionally, 19.0% experi-enced a worsening in food insecurity.

Main findings: The COVID-19 pandemic has had a negative impact on adolescents and young adults with T2D in multiple spheres. This includes a decrease in access to healthcare, negative mental health impact, and increasing concerns related to food insecurity. This information is nec-essary in order to develop strategies to mitigate these effects.

P17

Insulin reactions: what do you do when your treatment's the trigger?

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We present the case of a 17yo female patient with Type 1 Diabetes who developed subcutaneous reactions to her insulin. The patient was diagnosed with diabetes at age 4y and has a history of both lipohypertrophy and lipoatrophy. After 11 years of insulin therapy, she started to develop painful, erythematous nodules at the sites of her insulin injections. These reactions gradually worsened over a two-year period before she brought them to medical attention. Reactions initially were only related to rapid insulin but progressed to also occur with long-acting insulin. Allergy and Dermatology teams were consulted. Work-up including patch testing did not reveal a specific trigger. Multiple different insulin types were trialed without improvement in her symptoms. Alternative delivery methods were trialed including the Medtronic i-Port injection port and pump therapy, both of which resulted in the same painful nodular lesions after a brief reprieve lasting <24h. Oral antihistamines did not alter the reaction severity. Anti-inflammatory medications did not reduce the pain. Topical treatments - including ultra-potent steroids, tacrolimus, EMLA and ice - applied to the skin prior to injection were ineffective. A biopsy of a nodule revealed septal panniculitis (inflammation of the subcutaneous fat), consistent with an immune-mediated reaction to insulin or its components. She was started on hydroxychloroquine with some improvement in the severity of the reactions but not the associated pain; hydroxychloroquine dosing has been recently optimized. The patient, who has a longstanding history of poor metabolic control has experienced worsening of her glucose control and increased diabetes distress. While cutaneous insulin reactions are described in the literature, there is a paucity of information about the presentation, risk factors and effective management strategies of insulin-induced panniculitis – an entity that has detrimental medical and psychosocial consequences.

P18

Growth response to human growth hormone in two patients with IGF1 re-ceptor defects

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BACKGROUND: The growth promoting effects of human growth hormone (GH) are mainly mediated through the action of in-sulin like growth factor-1 (IGF-1) on the IGF-1 receptor (IGF-1R). GH itself has little effect on height velocity, as shown in patients with GH receptor defects and low circulat-ing IGF-1. Interestingly, an average gain of 1.9 height SDS has been observed in patients with IGF-1R defects receiving GH.

OBJECTIVES: To describe the genetic characteristics and the changes in height and height velocity during GH therapy in two patients with IGF-1R defects.

RESULTS: Patient 1 initiated GH therapy (0.26 mg/kg/w) at 8.1 years and height increased by 0.8 SD after 2.2 years. Bone age at 9.3 years was 7 years 10 months. Patient 2 initiated GH therapy (0.2 mg/kg/w) at 1.4 years and height increased by 2.0 SD after 1.6 years. In both patients, GH treat-ment caused an increase in IGF-1 concentrations to supraphysiological levels.

DISCUSSION AND CONCLUSION: GH therapy caused an increased in HV and height SDS in both pa-tients. This effect was more pronounced in patient 2 who was started at a younger age. Consistent with other studies, our patients were given a dose of GH higher (0.2-0.26 mg/kg/w) than what we use for GH deficiency (0.18 mg/kg/w). The effect of GH in IGF-1R defects is un-clear; proposed mechanisms include paracrine activity of GH or supraphysiological concentrations of IGF-1 that stimulate partially ineffective receptors (although correlation between IGF-1 levels and growth re-sponse has not been evaluated). One study showed that stimulation with recombinant IGF-1 in fibroblasts containing IGF-1R defects resulted in a normal amount of mutated IGF-1R protein, but downstream signaling was impaired. The definitive mechanism by which patients with IGF-1R de-fects respond to GH, and the impact on final height, are unknown.

P19

Type 2 diabetes off-label treatment: Retrospective review of children and adolescents followed at Children's Hospital of Eastern Ontario's Type 2 Diabetes Clinic 2014-2020

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Despite a plethora of approved medications for adults with type 2 diabetes (T2DM), non-research based medication management of T2DM in the pediatric population is limited to metformin and insulin. At the same time, evidence suggests youth with T2DM are at greater risk of treatment failure. This study aimed to determine the frequency of off-label medication use among children and adolescents followed at the Children's Hospital of Eastern Ontario (CHEO) T2DM clinic from April 1, 2014 to March 1, 2020. A total of 205 children and youth with T2DM were followed by the CHEO T2DM clinic during the study period.

Off-label treatment was initiated in 18.5% (n=38) of the study population (mean age: 16.34±1.79; min 10.48, max 19.18). In this group, T2DM management prior to off-label medication included metformin in 97.4% (n=37) plus insulin in 73.7% (n=28). There was no difference in duration of T2DM or HbA1C at initiation of off-label management among girls (n=28) and boys (n=10). How-ever, HbA1C was significantly higher both at onset of T2DM (9.17±1.98 vs 7.67±1.17) as well as at initiation of off-label treatment (9.86±2.99 vs 7.5±2.17) among those who were on insulin prior to off-label treatment. Improvement in glycemic control was the most common indication (84.2%) for off-label treatment. Basal insulin only (75%), followed by MDI regimen (14.29%) were the most common method of insulin use prior to off-label treatment.

Sitagliptin (78.95%), followed by empagliflozin (13.16%) and liraglutide (5.26%) were the most commonly used off-label medications in the study population. An upward trend in off-label use with increased duration of T2DM was noted, and 44.74% youth with 2-4 years history of T2DM were on off-label medication. A second off-label medication was used in management of 4 youth (1.9%). With the exception of one local reaction to liraglutide at the injection site, there were no other reported adverse effects.

In conclusion, off-label treatment use is not an uncommon practice in management of youth with T2DM to improve glycemic control. National and international pediatric T2DM centers' experience with off-label treatment may facilitate the introduction of additional therapeutic options for youth with T2DM.

P20

A Novel STAT3 Gain-of-Function Mutation as a Cause of Neonatal Onset Polyendocrinopathy

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A term newborn male presented on day one of life with neonatal goiter. Mom had been diagnosed with hypothyroidism late during pregnancy and was treated with levothyroxine. Thyroid function tests done on first day of life showed overt hypothyroidism, with elevated TSH > 500 and low free T4 5.9. Anti-TPO antibodies were strongly positive. He was started on levothyroxine therapy and discharged home with follow-up to titrate doses.

He presented at 3 weeks of age with a 4 day history of vomiting, lethargy, and tachypnea. Labs drawn indicated severe diabetic ketoacidosis (DKA), with glucose 43 mmol/L, pH 6.94 and bicar-bonate 5 mmol/L. He was admitted and managed as per DKA protocol. He transitioned to subcu-taneous insulin using a multiple daily injection regimen, with Lantus and diluted Humalog. He was also empirically started on glyburide, but found to be sulfonylurea non-responsive and was sub-sequently managed on insulin alone. Anti-GAD and anti-insulin antibodies were strongly positive. In the months following diagnosis, he had persistent hyperglycemia, with insulin requirements much higher than expected for his age (>1.5 units/kg/day).

Due to his atypical presentation of congenital goiter and early-onset diabetes, whole exome se-quencing was performed which revealed a variant of unknown significance in the STAT3 gene (c.1847A>G, p.Glu616Gly). STAT3 is an important transcription factor involved in JAK-STAT sig-naling pathways. Functional assays confirmed this variant to be pathogenic, resulting in very high STAT3 gain of function (GOF). Immunologic studies showed elevations in IgG and multiple cyto-kines suggesting immune dysregulation. Monitoring for other autoimmune conditions revealed development of intermittent neutropenia at 6 months of age.

Discussion: The majority of neonatal diabetes mellitus (NDM) involve isolated diabetes. However, NDM may also be associated with other pathological conditions and autoimmune diseases. In these cases, more comprehensive genetic testing may be required to identify a unifying diagnosis. STAT3 GOF disease is a rare cause of NDM, often associated with lymphoproliferation and early-onset autoimmunity affecting multiple organs. Confirming this genetic diagnosis is essential as management includes immunosuppressive and immunomodulatory therapy, which might lower insulin requirements; as well as close monitoring by a multidisciplinary team for other immune-related complications.

P21

Minimum Incidence of Optic Nerve Hypoplasia and Septo-Optic Dysplasia in Canadian Children: A CPSP Study

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Background: Isolated optic nerve hypoplasia (ONH) is a non-progressive congenital abnormality associated with a reduced number of axons in one or both optic nerves. Previously believed to be a rare condition, studies have found isolated ONH to be relatively common with an estimated incidence of 1 in 2,287 live births. ONH can be associated with other developmental abnormalities resulting in a spectrum of clinical phenotypes from isolated ONH to include disrupted pituitary gland development and/or development of the midbrain. Septo-optic dysplasia (SOD) is defined as the presence of at least two of the following findings: ONH, pituitary hormone dysfunction (PHD), or midline abnormalities of the brain. There are few studies reporting the incidence rates of SOD, with rates reported between 2.4/100,000 (<19 years) and 53.3/100,000 (<18 years) but many suggest the incidence rates are rising. Several studies have identified prenatal risk factors for developing ONH/SOD including primiparity, prematurity, young maternal age, and alcohol or cocaine exposure. More research is needed to better understand the association of prenatal risk factors to the development of SOD and associated timing of PHD, which can be associated with significant morbidity if not detected early.

Objectives: We aim to determine: (i) the minimum incidence of ONH and SOD in Canadian children <18 years of age; and (ii) the minimum incidence of PHD at the time of diagnosis of ONH and the associated clinical phenotypes.

Methods: In this study, pediatricians and pediatric sub-specialists will report new cases of ONH and SOD in Canadian children <18 years of age over a 12-month period using the surveillance methodology and infrastructure available via the Canadian Pediatric Surveillance Program. We will also collect demographic and clinical characteristics, age at diagnosis, associated PHD at the time of diagnosis and key investigations.

Conclusion: Our study will report prenatal and perinatal risk factors for ONH/SOD, and associated clinical phenotypes that will inform the development of screening guidelines for PHD. This research has the potential to reduce the morbidity associated with these conditions with early identification and timely intervention.

P22

Don't be chicken, it's just thyroiditis!

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Background: Thyroiditis is a rare complication of blunt and penetrating trauma to the neck and can lead to thyroid storm due to release of thyroid hormone into the bloodstream. Management may include medical and surgical options. There have been a few case reports of emphysematous thyroiditis due to infection. Here, we report a case of emphysematous thyroiditis due to trauma which has not been previously reported to our knowledge.

Case: A 15 year old female presented with a 3 day history of throat pain after eating chicken. X-ray and CT of the neck revealed a 2x0.4cm ossified foreign body lodged in the retro-esophageal soft tissues anterior to C6 vertebra. After urgent endoscopic removal, barium studies revealed an esophageal perforation at the level of C6. As the patient was febrile on presentation she was treated with antibiotics. A second CT scan 3 days after removal of chicken bone demonstrated an increased amount of air lucencies around and within the thyroid, consistent with emphysematous thyroiditis, with no evidence of surrounding abscess.

Furthermore, a non-occlusive thrombus of internal jugular vein was also demonstrated. FT4 and Tg were elevated (35.7 pmol/L and 67.8 ug/L) with suppressed TSH. All thyroid antibodies were negative. The patient remained clinically euthyroid, and heart rate remained below 90/min. Thyroid function tests normalized over several weeks.

Discussion: We report a rare complication of esophageal trauma, emphysematous thyroiditis, which resolved with conservative management. Previous case reports of thyroiditis secondary to trauma have required more active management due to clinically significant hyperthyroidism. There have also been reports of neck trauma complicated by thyroid hemorrhage and suppurative infection of the thyroid. Our case demonstrates the importance of considering thyroid involvement in neck injuries due to both blunt and penetrating neck trauma.

P23

Permanent Versus Transient Hypothyroidism In Children With Congenital Hypothyroidism (CH) Identified Through The Newborn Screening Ontario (NSO) Program

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Background: The reported incidence of CH has increased in recent years, and is thought to be largely related to the identification of mild, transient thyroid dysfunction. Due to concerns for permanent neurodevelopmental sequelae from undertreated CH, many with transient CH (T-CH) are treated with thyroxine for 3 years, leading to possible unnecessary medicalization, healthcare use and parental stress. We sought to determine the frequency of T-CH vs permanent CH (P-CH) in a cohort of children, and to identify potential early predictors to guide management decisions.

Methods: A retrospective chart review identified patients with CH from the five main NSO retrieval centres from 2006-2015. Data were analyzed by descriptive statistics.

Results: In total, 499 cases (57.5%) were included for analysis: 360 (72.1%) had P-CH, 109 (21.8%) had T-CH and 30 (6%) were indeterminate.

Conclusions: Analysis of descriptive data demonstrates several key data points that diverge between patients with T-CH versus P-CH. Analysis is underway using multivariable logistic regression to develop a predictive tool for earlier differentiation between these groups and will be presented.

P24

Illness Management Advice in Congenital Adrenal Hyperplasia – Is an Update Needed?

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Background: Congenital adrenal hyperplasia (CAH) is a family of autosomal recessive disorders that impair cortisol biosynthesis. In the severe or 'classic' form of the disease, physical stress like illness or injury can lead to life-threatening hyponatremia, hypoglycemia, and adrenal crisis, espe-cially in children. Without proper treatment, patients are at risk for seizure, coma, and even death – with rapidly progressing adrenal crises significantly related to hypoglycemia. As a result, it is es-sential patients and their families have a thorough understanding of how to manage physical stress to avoid serious adverse outcomes.

Methods: Clinical practice guidelines from the Endocrine Society (ESCPG) and four recent review papers were assessed. All patient education handouts regarding adrenal insufficiency posted on the Canadian Pediatric Endocrine Group (CPEG) website from pediatric health centres across Cana-da were also reviewed. Items noted were the described elements of adrenal crisis and illness man-agement recommendations during periods of stress.

Results: The ESCPG and all review papers described hyponatremia, hypoglycemia, and death as possible consequences of adrenal crisis. Additionally, all recommended doubling or tripling the dose of hydrocortisone during periods of stress and immediate parenteral administration of hy-drocortisone when patients are unable to tolerate doses orally. Lastly, 4/5 papers recommended increasing consumption of fluids and 3/5 recommended frequent carbohydrate consumption dur-ing periods of stress – including the ESCPG.

Of the 9 available patient education handouts, 3 mentioned hyponatremia, 6 mentioned hypogly-cemia, and 3 mentioned death as possible consequences of adrenal crisis. All handouts provided individualized instructions for oral and parenteral hydrocortisone administration during periods of stress and adrenal crisis respectively. However, only 1/9 handouts recommended increased fluid intake and none recommended increased carbohydrate consumption.

Conclusion: During periods of stress, in addition to increased glucocorticoid dosing, recent published recommendations suggest increased fluid and carbohydrate intake for CAH patients, especially children. However, these non-pharmacological recommendations are lacking in current educational handouts found on the CPEG website. Given that recent literature reports hypoglyce-mia as an important cause of morbidity and mortality in classic CAH, handouts should be revised to specifically recommend regular consumption of simple and complex carbohydrates to help avoid potentially fatal adrenal crises.

P25

Successful Improvement of Severe Hypertriglyceridemia in Congenital Lipodystrophy type 4 with Diet and Icosapent Ethyl

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Congenital Lipodystrophy type 4 (CLD4), is a rare autosomal recessive condition secondary to CAVIN1 gene mutation. It prevents formation of caveolae, a cell membrane structure important in lipid transport, thereby leading to severe hypertriglyceridemia and insulin resistance. CLD4 is also associated with arrythmia, myopathy and early mortality.

We present a unique case of a 17-year-old patient who presented with hyperglycemia, polydipsia, polyuria, and a hemoglobin A1C of 13.9%. She had coarse facial features, lack of adiposity, acanthosis nigricans, hepatosplenomegaly, left leg amputation, right leg lymphedema and failure to thrive with weight z score of -3.28. Investigations revealed negative anti-GAD, islet cell and anti-insulin antibodies. On oral glucose tolerance test, blood glucose increased from 10 to 17.9mmol/L and insulin from 809 to 1,450, with C-peptide of 5197 in keeping with insulin resistance and type 2 diabetes. She was started on insulin and metformin. Genetic testing subsequently confirmed a diagnosis of CLD4.

Her triglycerides (TG) were initially 4.48mmol/L but increased with improved nutritional status and weight gain. She required multiple hospital admissions for severe hypertriglyceridemia with TG ranging between 13.52 to >17.79 (<1.02 mmol/L). Xanthomas were present. Lipase levels were normal. Her abdominal ultrasound showed hepatosplenomegaly and non-alcoholic fatty liver disease. She had one admission for severe septic shock of unknown etiology. Due to other known association of CLD4, further investigations revealed CK levels of 1000 - 1400s (45 – 230u/L).

Her Holter study showed frequent ventricular ectopy, and nadalol was started.

The treatment for CLD4 is metreleptin, which is unavailable in Canada. She was started on a strict low carb and low-fat diet. Due to her elevated CK and concern for rhabdomyolysis with fenofibrates, icosapent ethyl, an omega 3 fatty acid was initiated. Clinically, the patient is doing well and has not required further hospital admission. Hemoglobin A1C improved to 5.7% and TGs to 4 mmol/L. However, TG continue to fluctuate with diet and medication compliance.

This is a unique case report of a patient with congenital lipodystrophy and significantly elevated triglycerides who responded well to strict diet, tight glycemic control and icosapent ethyl.

P26

Variable Presentations in the Diagnosis of Complete Androgen Insensitivity Syndrome

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Complete androgen insensitivity syndrome (CAIS) is a disorder of sexual development (DSD) resulting in the failure of virilization in genetic (XY) males. This is characterized by external female genitalia and male gonads in the absence of a uterus and ovaries along with levels of testosterone and DHT that are of a normal male. The detection, presentation and impact of this diagnosis on the patient and their family is highly variable as the diagnosis usually results secondary to investigations for other clinical concerns.

We present a case series of 3 very different patients assessed at 3 different stages in development and with 3 different presenting concerns. An ex-32-week premature infant was diagnosed at 2 weeks of life following a finding of XY karyotype on microarray which was done for genetic assessment of congenital limb anomalies and dysmorphism. Elevated testosterone and pelvic ultrasound showing absence of a uterus and ovaries with bilateral masses in the inguinal canals further supported the diagnosis. A 4-year-old diagnosed at 2 years of age with XY karyotype following microarray for concerns of developmental delay and autism was found to have elevated anti-Müllerian hormone and testicles in the inguinal canals bilaterally on ultrasound. Finally, a 16-year-old teen was referred to Endocrinology at 16 years of age following concerns of primary amenorrhea discussed in Neurology Clinic where she is followed for generalized epilepsy. She had a microarray done in March 2019 for a genetic basis of her learning disability and epilepsy, which identified XY karyotype. A left inguinal hernia correction 11 years earlier in 2009 identified a gonad in the inguinal canal but follow-up could not be located in her medical records.

The variable age at presentation of these patients and associated clinical questions that arise surrounding gonadectomy, pubertal induction and gender identity provide a complex and challenging foundation upon which to manage and counsel patients and families. In presenting this case series, we hope to highlight important lessons for the medical practitioners involved in the care of these patients and provide further exposure to the variability and complexity of CAIS diagnosis and management.

P27

DiabEtes Management Order Set (DEMOS): Patient Safety in Admitted Chil-dren and Youth with Diabetes

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Background: Standardized physician order sets may reduce medication errors and incorporate best available ev-idence. However, limited data exists regarding their impact on pediatric inpatient care. Type 1 Dia-betes (T1D) is a relatively common chronic health disorder among Canadian children. Inpatient di-abetes care (including insulin administration) is a frequent source of medication errors and pre-ventable adverse outcomes. To address this, we developed a pediatric inpatient DiabEtes Manage-ment Order Set (DEMOS).

Objectives: To determine pre-DEMOS implementation adherence to quality improvement (QI) standards in in-patient diabetes orders and compare this with post-implementation adherence.

Methods: We will perform a before-and-after study of children (0-18 years) admitted to McMaster Children's Hospital (MCH) with diabetes requiring insulin. We excluded patients admitted for new diabetes presentations and patients admitted to mental health units.

We selected a random sample of children registered with the MCH Pediatric Diabetes Clinic who matched our inclusion and exclusion criteria. Patient demographics and inpatient physician orders were extracted from charts. Our "before cohort" included children hospitalized between Jan 2019 – Jun 2020 (pre-DEMOS introduction) and the "after cohort" between Aug 2020 – January 2021. Our target sample size was 20 children in each cohort, to facilitate future rapid cycle improvements after initial data collec-tion.

We will measure the uptake of DEMOS and other selected QI outcome measures (the proportion of physician orders that include orders for hyper- and hypoglycemia management, insulin-carbohydrate ratios, insulin sensitivity factor calculation and length of stay). Additionally, PDSA cycles will be used to evaluate uptake of the DEMOS.

Results: We have analysed data from our baseline cohort of 20 children. Prior to DEMOS introduction, the two most common omissions in physician orders were not including an age/weight-based oral dextrose dose for hypoglycemia and not including orders for hyperglycemia management. Fur-thermore, 70% of initial physician orders included an insulin-carbohydrate ratio and only 65% in-cluded an insulin correction factor. The average length of stay was 2.45 days.

Conclusion: This QI study will provide additional information regarding the impact of standardized physician order sets on inpa-tient pediatric care and local adherence to established standards of care for in-patient diabetes management at MCH.

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P28

A Rare Etiology of Clitoromegaly in Neurofibromatosis Type 1

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Introduction: A 3-year-11-month old female with known neurofibromatosis type I (NF1) presented for evalua-tion of clitoromegaly. She was experiencing new urinary frequency, dysuria, and urge incontinence, having previously been toilet trained and continent during the day.

Case Description: The patient had no other symptoms of virilization. On examination, she was pre-pubertal. Her clitoris measured 5cm (length) by 1.5cm (width) and had an irregular, firm texture. There was no posterior labial fusion, labial rugae, or palpable gonads to suggest a difference of sexual differen-tiation. The left labia was enlarged compared to the right. Initial investigations revealed undetect-able testosterone, androstenedione, and 17-hydroxyprogesterone levels. Karyotype was 46XX. Pel-vic ultrasound demonstrated bilateral lobulated retro-vesicle masses which were 4x3x2cm (left) and 2x2x1cm (right). Pediatric urology advised urgent biopsy and excision. Laparoscopy revealed a diffuse sheet of suspected plexiform neurofibromatosis (PN) adhering to bladder and throughout the pelvis. Pathology confirmed the diagnosis of PN. Magnetic resonance imaging (MRI) demon-strated inferior extension of the PN to the perineum and clitoris. The mass surrounded the urethra and rectum and demonstrated intraspinal extension through the sacral foramina bilaterally. There was also left sided obturator muscle and gluteal medius involvement.

The PN was unamenable to surgical resection given its complexity and large size. Pediatric oncol-ogy advised treatment with Trematinib, a MAP-K kinase enzyme (MEK) inhibitor, to reduce or sta-bilize the PN. A follow-up MRI after 5 months of treatment demonstrated partial radiographic re-sponse with reduction of the bladder mass measurements. She demonstrated clinical response with resolution of her urinary symptoms within 3 months of treatment.

Discussion: Clitoromegaly can be classified as hormonal and non-hormonal. Features that suggested a non-hormonal etiology in this case include the absence of other virilizing signs, asymmetrical en-largement of left labia and perineum, irregular texture of clitoris and undetectable serum andro-gens. Genitourinary PN is a rare etiology of non-hormonal clitoromegaly that is important to con-sider in those with known diagnoses of NF1 or clinical features of same. It is imperative to evalu-ate the extent of involvement with diagnostic imaging because genitourinary PN can pose signifi-cant risk of morbidity with involvement and compression of critical pelvic structures.

P29

Bilateral gynecomastia due to intake of sweet potatoes: a case report.

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Gynecomastia is a common benign breast condition in males, typically occurring in neonates and during puberty. It can result from various mechanisms, but is generally due to increased action of exogenous or endogenous estradiol relative to endogenous production of androgens (in particular testosterone). It is important for endocrinologists to be aware of environmental causes of gyne-comastia, given they are preventable and easily treatable.

We report a 4-year-old male with cerebral palsy who was referred to our clinic for evaluation of bilateral breast budding of several months duration. He had no other signs of precious puberty. His history was not suspicious for pathologic causes of gynecomastia, and he was on no relevant medications. However, due to his multiple food allergies, his diet was highly reliant on sweet po-tato. His parents would feed him multiple mashed sweet potatoes per day to the extent that his skin pigment was notably orange. His physical examination showed a bilateral Tanner II gyneco-mastia with Tanner I testicular volume and pubic hair. Otherwise, his physical examination was unremarkable. Consequently, we recommended eliminating sweet potato from his diet completely. Upon re-evaluation four months later, his gynecomastia had completely resolved.

Our hypothesis is that in this case the patient's gynecomastia occurred due to high intake of sweet potato (ipomoea batatas) which contains phytoestrogen, an estrogen-like compound that is structurally similar to 17B-estradiol. There have been similar case reports in the literature of gy-necomastia or precocious puberty due to exposure to lavender, tea tree oil and soy. However, to our knowledge this is the first case report related to sweet potato intake. We present a discussion of the pathophysiology, and brief review of the literature on gynecomastia due to environmental exposure.