

February 8-10, 2024

Delta Hotels Saskatoon Downtown

18th Annual

CPEG Scientific Meeting

Hosted in Saskatoon by:

Division of Pediatric Endocrinology & Diabetes at the University of Saskatchewan

Full Digital Program



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Welcome to Saskatoon!

On behalf of the Division of Pediatric Diabetes and Endocrinology at the University of Saskatchewan, we are pleased to welcome you to Saskatoon!

We are privileged to host our colleagues from across Canada in our beautiful city of Saskatoon — Treaty 6 Territory and the Homeland of the Metis – for our 18th annual Scientific Meeting of the Canadian Pediatric Endocrine Group (CPEG). For many of you, this may be your very first visit to our province of Saskatchewan so we hope you enjoy your stay and have the opportunity to experience the wonderful things we have to offer.

The scientific program for CPEG 2024 is inspired by what is new and upcoming in the field of Pediatric Endocrinology. Based on feedback from our recent conferences, our invited symposia speakers and debaters will share their expertise in novel understandings of clinical diseases, diagnostics, and therapeutics. Furthermore, we are excited to hear from our array of oral and poster presenters who will be showcasing their research.

As we cannot hold such a conference without the support of our sponsors, we encourage you to visit our industry booths in the conference foyer. Furthermore, as this conference provides an optimal way to network and socialize, we hope to see you all at our Friday evening event to be held within the Remai Modern — one of Saskatoon's newest attractions situated on our picturesque riverfront.

As your hosts, we would be pleased to share any local information with you — whether it be suggestions for where to eat, grab a coffee, or engage in outdoor activities.

We hope you have an engaging and memorable experience at CPEG 2024

Mark Inman & Raelynn Friesen (CPEG 2024 local organizing committee)

Dear Attendees,

On behalf of the Scientific Committee and the Canadian Pediatric Endocrine Group (CPEG) Executive, I would like to welcome you all to the 18th Annual CPEG Scientific Meeting.

With our partners at the University of Toronto Continuing Professional Development, I hope that you will find that, the Scientific Committee has developed a fantastic meeting and program. In CPEG tradition we will continue to enjoy the habitually dynamic CPEG debate. As always, learners and others will have the opportunity to present their research in the oral and poster abstract sessions. The catchy one-minute poster highlights will also return this year. We hope that the program will meet the expectations of all attendees including nurses, scientists, trainees, endocrinologists, and other care providers..

I would like to thank the Scientific Committee for their hard work in planning this meeting with a special acknowledgement to Mark Inman and Raelynn Friesen, our local hosts.

Finally, a big thank you to our sponsors whose continued support makes this meeting possible, and I encourage you to explore their exhibits. CPEG would also like to thank those sponsors who also support our CPEG Fellowship Awards allowing us to train future endocrinologists. This year's awardees will be announced on Saturday...

I hope that you all have a motivating, fun, and collegial meeting.

Rebecca Perry

Chair, 2024 CPEG Scientific Committee

Scientific Committee

Rebecca Perry (Chair) Rose Girgis

Mark Inman (Local Jana Haylor

Chair) Melanie Henderson

Funmbi Babalola Paola Luca

Sharon Costantini Mallory McNiven

Manpreet Doulla Kate Potter

Raelynn Friesen Andrea Ens (Ex Officio

Patricia Gallego Member)

Full Conference Program

Wi-Fi Internet Access

Network: **Delta_Conference**Password: **saskatoon2024**

You can download the full conference program from the link in your conference reminder email or from the conference home page at www.cpd.utoronto.ca/cpeggcep/

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, approved by Continuing Professional Development, Temerty Faculty of Medicine, University of Toronto. You may claim a maximum of 10.0 hours (credits are automatically calculated).

Session Polling and Q&A

Visit slido.com and enter the code CPEG (not case-sensitive) or scan the QR code on the right to participate in polling questions and to submit your question during each session. The moderator will review the questions and ask the speaker during Q&A.



Faculty Disclosure

It is the policy of the University of Toronto, Temerty 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.

The 18th 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.

Having returned to in-person meetings in 2023, we are pleased to gather again for our 2024 CPEG Meeting for the very first time in the province of Saskatchewan. The scientific committee has worked hard to build on the successes of both the past in-person and virtual meetings. We hope to provide attendees with an exceptional meeting experience. Please note this year's meeting will commence midday on Thursday and conclude mid-day on Saturday, enabling attendees to return home on Saturday.

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. We look forward to visiting and learning with you.

Program Learning Objectives

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

- 1. Utilize knowledge of newer therapeutic and diagnostic options to optimize the management of pediatric pituitary disease
- 2. Recognize the efficacy of newer therapeutic agents in the prevention of Type 1 Diabetes Mellitus
- 3. Utilize clinical and technical knowledge to optimise management of youth living with Type 1 Diabetes Mellitus on advanced pump therapy
- 4. Formulate a comprehensive approach to assessment and management of pediatric obesity
- 5. Identify children and adolescents at higher risk of future infertility who would benefit from timely referral to specialized fertility services
- 6. Compare different treatment options in youth with Type 2 Diabetes Mellitus

Session Learning Objectives

Symposium I - Pediatric Fertility Preservation

Perspectives from Reproductive Endocrinology and Fertility - Shu Foong

Objectives:

- 1. Describe normal physiology as it relates to the female reproductive system
- 2. Identify fertility preservation options currently available for the post-pubertal patient with ovaries
- 3. Discuss emerging fertility preservation options for the pre-pubertal patient with ovaries

Fertility Considerations and Preservation in Klinefelter Syndrome - Trustin Domes Objectives:

- 1. Explore the challenges, implications and controversies surrounding fertility preservation in patients with Klinefelter syndrome
- 2. Discuss the options, strategies and technologies available for patients with Klinefelter syndrome wishing to pursue fertility preservation
- 3. Appreciate the different ethical considerations that may influence decision making when discussing fertility preservation with pediatric patients and their family

Symposium II - Pediatric Obesity Guidelines

Review of Literature in Pediatric Obesity - Stasia Hadjiyannakis, Kathy Morrison, Melanie Henderson, Krista Oei Objectives:

- 1. Utilize knowledge of pharmacotherapeutic options in the management of the pediatric patient with obesity
- 2. Be familiar with the literature highlighting the efficacy of pharmacotherapy in the management of obesity in childhood
- 3. Incorporate knowledge of health outcomes of relevance in the assessment and management of a child or youth with obesity
- 4. Examine the evidence related to behavioral and psychological interventions that will inspire the upcoming Canadian guidelines for the management of pediatric obesity
- 5. Identify gaps and shortcomings of available evidence

Symposium III - Hot Topics in Pituitary Disease

Long-acting GH - Preetha Krishnamoorthy

Objectives:

- 1. To discuss the rationale: Why even consider LAGH?
- 2. To describe the prolongation technology and pharmacologic properties of the 2 LAGH approved by Health Canada
- 3. To review the trials to date with a focus on the safety and efficacy data
- 4. To discuss our experience using LAGH in patients at the Montreal Children's Hospital

Clinical Utility of Co-Peptin in the Diagnosis of Diabetes Insipidus - Greg Kline

Objectives:

- 1. Participants will be able to list the potential study design problems for any test pertaining to the diagnosis of diabetes insipidus
- 2. Participants will be able to describe the physiology and interpretation of copeptin measurements
- 3. Participants will be able to incorporate a new stimulated copeptin test protocol into their diagnostic pathways for polydipsia/polyuria syndromes

Symposium IV – Diabetes

Advancements in Loop Technology - Laurent Legault

Objectives:

- 1. Identify different characteristics of current diabetes technologies
- 2. Describe strategies to alleviate carb counting in a closed loop system

Emerging Treatments to Prevent Beta Cell Loss in Type 1 Diabetes - Diane Wherrett Objectives:

- 1. To review evidence for treatment of recent onset type 1 diabetes from recent trials
- 2. To review the use of teplizumab to delay the onset of type 1 diabetes in those at high risk

Debate

Be It Resolved That a GLP-1 Agonist Should Be Used As 2nd Line Treatment, Instead of Insulin, in Youth-Onset Type 2 Diabetes - Sanjukta Basak, Brandy Wicklow

Objectives:

1. Compare different treatment options in youth with Type 2 Diabetes Mellitus

CPEN Symposium

To Grow or Not to Grow? Understanding Growth Hormone and GnRH Analogues - Kate Davies Objectives:

1. To understand the pharmacodynamics of growth hormone and GnRH analogues

What's Amazing for the Adrenals? New Treatments and Novel Monitoring Methods in CAH - Kate Davies Objectives:

1. Appreciate alternative methods in managing children and young people with congenital adrenal hyperplasia

Fellows' Symposium

Pediatric Hypoglycemia - Daphne Yau

Objectives:

- 1. Discuss the biochemical features used to diagnose congenital hyperinsulinism
- 2. Provide an overview of the causes of congenital hyperinsulinism through cases illustrating some of the more common persistent/genetic causes
- 3. Discuss the management of congenital hyperinsulinism and provide an overview of the outcomes

Academic and Community Pediatric Endocrinology - Zoya Thawer

Objectives:

- 1. To explore the subjective and objective benefits of a academic and community practices
- 2. Reflect on how you can build a career that embodies what you want (or at least how to start this reflection process!)
- 3. To leave you with some food for thought regarding career trajectories in Pediatric Endocrinology

Invited CPEG Speakers

Sanjukta Basak MD CM FRCPC MSc

HPTE

Associate Program Director, Department of Pediatrics, Division of Endocrinology Clinical Assistant Professor, University of British Columbia BC Children's Hospital Vancouver, BC

Trustin Domes MD MEd MCPM

FRCSC

Associate Professor, Department of Surgery Director of Admissions, College of Medicine University of Saskatchewan Saskatoon, SK

Shu C. Foong MD FRCSC

Clinical Assistant Professor, Department of Obstetrics & Gynecology University of Calgary Section Lead, Reproductive Endocrinology & Infertility Medical Director, Regional Fertility Program Calgary, AB

Stasia Hadiivannakis MD FRCPC

Assistant Professor, Director of Centre for Healty Active Living University of Ottawa Pediatric Endocrinologist Children's Hospital of Eastern Ontario (CHEO) Ottawa, ON

Mélanie Henderson MD PhD FRCPC

Pediatric Endocrinologist Associate Clinical Professor Division of Endocrinology and Diabetes University of Montreal Centre Hospitalier Universitaire Sainte-Justine Montreal, QC

Greg Kline MD FRCPC

Clinical Professor, Medicine/ Endocrinology Cumming School of Medicine University of Calgary Calgary, AB

Preetha Krishnamoorthy MDCM

FRCPC

Associate Professor McGill University Pediatric Endocrinologist The Montreal Children's Hospital Montreal, QC

Laurent Legault MD FRCP

Associate Professor
Department of Pediatrics
Division of Pediatric Endocrinology
McGill University
Montreal, QC

Katherine Morrison MD FRCPC

Professor, Department of Pediatrics Co-Director Centre for Metabolism, Obesity and Diabetes Research McMaster University Pediatric Endocrinologist, Children's Exercise and Nutrition Centre McMaster Children's Hospital Hamilton, ON

Krista Oei MD MSc FRCPC

Pediatric Endocrinologist The Hospital for Sick Children Toronto, ON

Diane Wherrett MD FRCPC

Professor, Department of Pediatrics Hospital for Sick Children University of Toronto Toronto, ON

Brandy Wicklow MD FRCPC

Associate Professor, Rady Faculty of Health Sciences University of Manitoba Pediatric Endocrinologist Winnipeg Children's Hospital Winnipeg, MB

Invited CPEN Speaker

Kate Davies RN(Child) DipHE MSc PGCert PGDip
Associate Professor, Paediatric Prescribing & Endocrinology
Children's Advanced Nurse Practitioner
Honorary Clinical Research Fellow in Paediatric Endocrinology
QMUL/Barts and the London NHS Trust/University College
London NHS Trust
Division of Advanced Clinical Practice and Non Medical
Prescribing
School of Nursing and Midwifery
London South Bank University
London, UK

Invited Fellows' Symposium Speakers

Zoya Thawer MD FRCPC

Clinical Instructor, Faculty of Pediatrics University of British Columbia Pediatric Endocrinologist, Vancouver Island Health Authority Pediatric Endocrinologist, West Coast Pediatric Endocrinology Group

Victoria, BC

Daphne Yau MD FRCPC MSc

Assistant Professor, Department of Pediatrics University of Saskatchewan Pediatric Endocrinologist Jim Pattison Children's Hospital Saskatoon, SK

Invited CPEG Speaker Biographies

Sanjukta Basak

Dr. Sanjukta Basak is a pediatric endocrinologist and clinical assistant professor at the University of British Columbia and BC Children's Hospital. She completed her medical degree at McGill University, followed by a Paediatric residency and Paediatric Endocrinology fellowship at the Hospital for Sick Children in Toronto. Sanjukta has a keen interest in medical education also has completed a Masters of Community Health with a focus on health practitioner education at the University of Toronto. She is the medical lead of pediatric insulin resistance and Type 2 Diabetes program at BC Children's hospital. She is also currently the associate pediatric endocrinology fellowship program director and has a research interest in health equity in diabetes and obesity care.

Trustin Domes

Trustin Domes is an Associate Professor of Surgery (Urology), practicing male reproductive health specialist and the current Director of Admissions for the USASK College of Medicine. He is passionate about leading transformative changes that improve processes and outcomes in both medical education and healthcare.

Shu Foong

Dr. Foong completed fellowship in Reproductive Endocrinology and Infertility at the Mayo Clinic, USA. She is the Medical Director of Oncofertility and the Regional Fertility Program in Calgary. Her clinical interest in Oncofertility spans over two decades. Dr. Foong is responsible for starting both the oocyte cryopreservation and ovarian tissue cryopreservation programs in Calgary.

Stasia Hadjiyannakis

Stasia Hadjiyannakis is a pediatric endocrinologist and the Medical Director of CHEO's Center for Healthy Active Living. She is an associate Professor of Pediatrics at the University of Ottawa and has been an active member of the Department of Pediatrics at CHEO in the division of endocrinology since 2002. Her clinical, advocacy and research interests are in the area of pediatric obesity assessment and management, with a focus on the development and evaluation of clinical tools such as the 5As of Pediatric Obesity Management and the Edmonton Obesity Staging System for pediatrics.

Mélanie Henderson

Dr Henderson's research focuses on pediatric cardiometabolic health and its epidemiology, particularly the influence of lifestyle habits (physical activity, sedentary behavior, diet and sleep) on metabolic health in various population, including healthy children and those living with Type 1 and Type 2 Diabetes, in order to identify the best preventive strategies.

Greg Kline

Dr. Kline joined the faculty of medicine in 2001 and has been the Medical Director of the Endocrine Hypertension Clinic since 2005 and of the Dr. David Hanley Osteoporosis Centre since 2015. He has supervised over 50 medical trainees in research programs and lists over 150 peer reviewed publications pertaining to adrenal disease, electrolytes, and osteoporosis/metabolic bone disease.

Preetha Krishnamoorthy

Preetha Krishnamoorthy completed her pediatrics residency and fellowship in Pediatric Endocrinology at the Montreal Children's Hospital and has been an attending staff there since 2003. She is the Director of Pediatric Undergraduate Education at McGill. She won the Paige and Bernard Kaplan Award for excellence in teaching in 2004, the Osler Award by McGill Medicine's graduating class of 2012 of 2022 and was named to the Faculty Honour List for Educational Excellence. She is one of the recipients of the 2016 CAME Certificate of Merit Awards. In addition to her medical-related teaching, she loves to teach Zumba!

Laurent Legault

Dr Laurent Legault is a graduate of Université de Montréal.

He is an associate professor at the department of pediatrics at McGill University. His research interests are in the fields of diabetes technology, diabetes prevention as well as severe complex obesity management.

Katherine Morrison

Dr. Katherine Morrison is a pediatric endocrinologist, Professor and Co-Director of the Centre for Metabolism, Obesity and Diabetes Research at McMaster University. Having attended the University of Calgary for medical school and pediatric training, she undertook her pediatric endocrinology training at Stanford University. She has subsequently worked at the University of Manitoba and Ludwig-Maximilians University in Munich before moving to McMaster. She is a clinician researcher and is active clinically in the Growing Healthy Pediatric Weight Management and Pediatric Lipid Clinics at McMaster Children's Hospital. Her research is centered around the etiology, consequences and treatment of obesity and lipid disorders in children.

Krista Oei

Krista Oei is a pediatric endocrinologist at the Hospital for Sick Children. She completed her pediatric residency at BC Children's Hospital and fellowship in pediatric endocrinology at the Hospital for Sick Children. Her clinical and research interests include pediatric obesity, type 2 diabetes, calcium and bone disorders and quality improvement. She has been involved in the update to the Canadian Clinical Practice Guidelines for Managing Pediatric Obesity — in particular the bariatric surgery systematic review.

Diane Wherrett

Dr. Wherrett is a pediatric endocrinologist at the Hospital for Sick Children and Professor, Department of Pediatrics, University of Toronto. Her research focuses on interventions to prevent beta cell loss in type 1 diabetes. She is a member of the Steering Committee of Type 1 Diabetes TrialNet, chairs its largest study, Pathway to Prevention, and is the director for the Canadian Clinical Centre for this study group. She is the Principal investigator for Canadian Population Screening for Risk of Type 1 Diabetes Research Consortium.

Brandy Wicklow

Brandy Wicklow is a pediatric endocrinologist, head of the section of pediatric endocrinology and clinician researcher at the Children's Hospital Research Institute of Manitoba. Her clinical research focuses on the intergenerational and early life determinants of type 2 diabetes (T2D) and renal disease in children and adolescents.

Invited CPEN Speaker Biography

Kate Davies

Kate has been a children's nurse for 30 years, with 15 years working as a clinical nurse specialist in paediatric endocrinology. She now lectures in this field and in teaching healthcare professionals principles of paediatric prescribing. She is currently in her 3rd year of her PhD in Nursing, looking at alternative methods of monitoring in young people with congenital adrenal hyperplasia.

Invited Fellows' Symposium Speaker Biographies

Zoya Thawer

Dr. Zoya Thawer is a Pediatric Endocrinologist with the West Coast Pediatric Endocrinology Group in Victoria, BC and a clinical instructor at the University of British Columbia. She completed medical school at McMaster, pediatrics residency at Queen's and her fellowship in pediatric endocrinology at the University of Ottawa. Before settling in Victoria, she completed locum positions both at the University of Alberta Hospital in Calgary and the Stollery Children's Hospital in Edmonton. In her spare time, Dr. Thawer enjoys knitting, doing ceramics, baking and running - all things she has been able to make time for in her blended community and academic practice!

Daphne Yau

Dr. Daphne Yau is a pediatric endocrinologist at Jim Pattison Children's Hospital and an assistant professor at the University of Saskatchewan. Prior to her current appointment, she pursued post-fellowship training in congenital hyperinsulnism at the Children's Hospital of Philadelphia as well as at the Northern Congenital Hyperinsulinism Service through Royal Manchester Children's Hospital in the UK.

Awarded Fellowship Listing

1992-1993	M. Lawson		T. Pinto, B. Babic	2018-2019	J. Sorbara
1993-1994	S. Lawrence M. Lawson A. Simone	2008-2009	J. Deladoey A.M. Sbrocchi P Olivier	2019-2020	A. Chesover B. Navabi
1994-1995	S. Lawrence S.Taback A. Simone	2009-2010	T. Pinto R. Shulman P Olivier	2020-2021	A. Marr M. Lautatzis J. Ladd H. Geddie (declined)
1995-1996	C. Vaz S.Taback B. Cummings		T. Edouard S. Runge-Wildi C. Saaman	2021-2022	F. Babalola M. Jiang
1996-1997	J. Hamilton E. Sellers B. Cummings	2010-2011	E. Bassilious J. Wasserman Y. Yeshayahu S. Tsai	2022-2023	K. Oei T. Dyer K. Pabedinskas M. Feldman
1997-1998	J. Hamilton E. Sellers B. Cummings	2011-2012	M. Millete J. Wasserman C. Zuijdwijk	2023-2024	J. Stanley S. Rengan
1998-1999	J. Curtis J. Hamilton	2012-2013	M. Cohen J. Harrington		
1999-2000	J. Curtis J. Hamilton		T. Oron P. Luca M. Nour		
2000-2001	C. Panagiotopoulos C. Huang	0010 0014	D. Manousaki		
2001-2002	C. Panagiotopoulos S. Stock	2013-2014	K. Winston C. Leblicq A. Ens		
2002-2003	P Krishnamoorthy P Zimakas		B. Hursh I. Rousseau-Nepton		
	R. McEachern	2014-2015	I. Levy D. Manousaki		
2003-2004	P Krishnamoorthy H. Bui	2015-2016	L. Chiniara		
2004-2005	M. Nakhla J. Simoneau-Roy		S. Basak K. Verbeeten		
2005-2006	M. Nakhla I. Chapados M. Jetha	2016-2017	C. Nugent K. Pundyk N. Coles		
2007-2008	B. Wicklow	2017-2018	C. Nugent S. Fuchs		

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

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	2013	Karine Khatchadourian	2019	Julia Sorbara
2008	Meranda Nakhla	2014	Akash Sinha	2020	Christine Tenedero
2009	David Saleh	2015	Rayzel Shulman	2021	Richelle Waldner
2010	Brandy Wicklow	2016	Sanjukta Basak	2022	Funmbi Babalola
2011	Jonathan Wasserman	2017	Stephen Zborovski	2023	Tracy Dyer
2012	Jennifer Harrington	2018	Marie Eve-Robinson		

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 1 - 3 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 \$1,000 donation to a charity of their choice.

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

2017	Daniel Metzger	2022	Cheril Clarson	2023	Sarah Lawrence
2019	Denis Daneman	2023	Heather Dean		

Day 1 Agenda (All times are listed in local time)

Fellows' Symposium Thursday, February 8, 2024 (Room: Michelangelo C)

0830	Fellows Welcome & Breakfast	
	Fellows' Symposium Chair: Mallory McNiven (Edmonton)	
0850	Pediatric Hypoglycemia	Daphne Yau
0950	Coffee Break	
1020	Academic and Community Pediatric Endocrinology	Zoya Thawer
1120	Fellows Closing Remarks	Mallory McNiven

CPEG Program Thursday February 8, 2024 (Room: Michelangelo AB)

Each presentation will include a 25% (minimum) of interactivity comprised of audience response questions (polling) and audience submitted Q&A via Slido.com.

1100	On-Site Registration & Lunch	
1230	Opening Remarks & Thursday Poster Highlights (Odd Numbered Posters)	Mark Inman, Rebecca Perry
	Symposium I: Pediatric Fertility Preservation Chairs: Daphne Yau (Saskatoon), Richelle Waldner (Edmonton)	
1300	Perspectives from Reproductive Endocrinology and Fertility	Shu Foong
1345	Fertility Considerations and Preservation in Klinefelter Syndrome	Trustin Domes
1430	Break & Exhibits	
1500	Poster Viewing I (Odd Numbered Posters)	
P1	PROGRES, A Multi-country, Non-interventional, Prospective Study of Patients Receiving Human Growth Hormone Treatment Under Routine Clinical Care: Study Update	Carol Huang
P3	6-Month Subcutaneous Leuprolide Acetate Achieved and Maintained Hormonal and Clinical Suppression in All Weight Groups of Children with Central Precocious Puberty	Rebecca Perry
P5	Long Term Effects of Pediatric Hematopoietic Stem Cell Transplant on Endocrine Function	Ali Alghamdi

Day 1 Agenda (All times are listed in local time)

CPEG Program Thursday February 8, 2024 (Room: Michelangelo AB)

P11	Evaluation of a Multidisciplinary Pediatric Insulin Resistance and Type 2 Diabetes Program: The BC Children's Hospital Experience	Gonzalo Dominguez Menendez
P13	Integrating Exome Sequencing Into a Pediatric Endocrinology Clinic: A Genetics and Endocrinology Collaborative Quality Improvement Initiative	Duha Hejla
P15	Comparison of Characteristics of Children With Diabetes and Their Outcome in Haiti Compared to Countries With Different Economic Background	Regina Duperval
P17	Pheochromocytoma and Transient Central Hypothyroidism	Marie Edelyne St Jacques
P19	The Risk of Growth Hormone Therapy in Patients with ROHHAD Syndrome	Gabrielle Scantlebury
P21	Application of DKA Risk Mitigation Algorithm for ATTEMPT Clinical Trial Evaluating SGLT2i in Youth with T1D	Aliya Allahwala
	Symposium II: Pediatric Obesity Guidelines Chairs: Marina Ybarra (London), Jean-Pierre Chanoine (Vancouver)	
1530	Review of Literature in Pediatric Obesity	Stasia Hadjiyannakis, Katherine Morrison, Melanie Henderson, Krista Oei
1700	Welcome Reception & Exhibits	
1900	Adjourn for the Day	

Day 2 Agenda (All times are listed in local time)

CPEG Program Friday, February 9, 2024 (Room: Michelangelo AB)

Each presentation will include a 25% (minimum) of interactivity comprised of audience response questions (polling) and audience submitted Q&A via Slido.com.

0730	Registration & Breakfast	
0830	Opening Remarks & Friday Poster Highlights (Even Numbered Posters)	Rebecca Perry, Mark Inman
0900	Oral Abstracts I Chairs: Karen McAssey (Hamilton), Danya Fox (Vancouver)	
OR1	Hyperglycemia in Acute Lymphoblastic Leukemia: Evaluation of Risk Factors for Insulin Requirement During Induction Chemotherapy	Kristina Pabedinskas
OR2	Short-term Impacts of a Gluten-Free Diet on Children and Adults with Type 1 Diabetes and Celiac Disease: Findings from the CD-LiFE Study	Daniel Weiman
OR3	Type 1 Diabetes Genetic Risk Score 2 to Estimate the Proportion of Non-Autoimmune Diabetes in Autoantibody Negative Patients	Jacob Nicodemo
OR4	Diabetic Kidney Disease Screening and Management Practices Amongst Canadian Paediatric Endocrinologists: A Nationwide Survey	Paul Ryan
OR5	Fertility Preservation for Transgender and Gender Diverse Youth: Development and Evaluation of a Patient-Centred eLearning Module	Joshua Stanley
OR6	More Than Just Hypercalcemia: A Case of Neonatal Severe Hyperparathyroidism treated with Cinacalcet Monotherapy	Emma Metivier
1030	Break & Exhibits	
	Symposium III: Hot Topics in Pituitary Disease Chairs: Andrea Ens (London), Mark Palmert (Toronto)	
1100	Long Acting Growth Hormone	Preetha Krishnamoorthy
1145	Clinical Utility of Co-Peptin in the Diagnosis of Diabetes Insipidus	Greg Kline
1230	Lunch & Exhibits	
1330	Poster Viewing II (Even Numbered Posters)	
P2	Impact of Transition From Symptomatic to Routine Celiac Disease Screening in a Pediatric Tertiary Care Type 1 Diabetes Clinic	Gabrielle Doré-Brabant

Day 2 Agenda (All times are listed in local time)

CPEG Program Friday, February 9, 2024 (Room: Michelangelo AB)

P4	Central Versus Peripheral? A Diagnostic Challenge in Precocious Puberty	Stephanie Lenet
P6	Measuring Adherence to Clinical Practice Guidelines for Screening of Complications and Comorbidities in Children and Youth Living With Type 1	Fahd Alshammri
P8	Bridge Over Troubled Water; Diagnostic Uncertainty in a Patient With Transient Diabetes Insipidus	Cillian Lineen
P10	Primary Hypothyroidism Presenting with Ovarian Hyperstimulation	Elise Martin
P12	Trends and Risk Factors for Severe Hypoglycemia in Type 1 Diabetes in a Pediatric Diabetes Clinic	Saheba Bajwa
P14	Pituitary Stalk Interruption Syndrome: A Rare Case Presenting with Arthrogryposis and Type 1 Diabetes Mellitus	Duha Hejla
P18	Early Onset Obesity and Adrenal Insufficiency Caused by POMC (Proopiomelanocortin) Deficiency	Hamad Haidar
P20	Hyperosmolar Hyperglycemic Syndrome and Diazoxide: A Case Report in a Child With Kabuki Syndrome	Harsh Kahlon
1400	Oral Abstracts II Chairs: Liz Rosolowsky (Edmonton), Celine Huot (Montreal)	
OR7	Use of Glucagon-like Peptide 1 Receptor Agonists in Adolescents with Prader-Willi Syndrome: A Case Mini-series	Paul Ryan
OR8	A Single-Center Experience With Pediatric Thyroid Nodules	Mallory McNiven
OR9	Hypothyroidism in Congenital Nephrotic Syndrome: A Case Series	Rachel Parker
OR10	Short Stature in Glucose Transporter 1 Deficiency Syndrome	Sruthi Thomas
OR11	Caregiver Knowledge of Hypoglycemia and Hyperglycemia Management in Children and Adolescents With Type 1 Diabetes	Matthew Feldman
OR12	Assessing Bone Mineral Accrual Rates in Non-binary Adolescents on Prolonged Course of GnRH Analogues	Poonam Jariwala
1530	Break & Exhibits	

Day 2 Agenda (All times are listed in local time)

CPEG Program Friday, February 9, 2024 (Room: Michelangelo AB)

	Symposium IV: Diabetes Chairs: Manpreet Doulla (Edmonton), Seth Marks (Winnipeg)	
1600	Advancements in Loop Technology	Laurence Legault
1645	Emerging Treatments to Prevent Beta Cell Loss in Type 1 Diabetes	Diane Wherrett
1730	Adjourn for the Day	
1800	Conference Social Event [pre-registration required] Join us for a drink and casual dinner at the Remai Modern Art Gallery. Door open at 6:00 PM - Dinner will start at 7:30 PM	Remai Modern, 102 Spadina Crescent E, Saskatoon Located an 8-minute walk from the Delta hotel

CPEN Program Friday, February 9, 2024 (Room: Michelangelo C)

	CPEN Symposium Chairs: Raelynn Friesen (Saskatoon), Sharon Costantini (Ottawa)	
0900	To Grow or Not to Grow? Understanding Growth Hormone and GnRH Analogues	Kate Davies
1030	Rejoin CPEG Group	
1400	What's Amazing for the Adrenals? New Treatments and Novel Monitoring Methods in CAH	Kate Davies
1530	Rejoin CPEG Group	
1600	CPEN Business Meeting [members only]	
1730	Adjourn for the Day	

Day 3 Agenda (All times are listed in local time)

CPEG Program Saturday, February 10, 2024 (Room: Michelangelo AB)

0730	AGM Check-In & Breakfast
0830	CPEG Business Meeting [members only]
1030	Poster Viewing III (All Posters)
1045	Break & Exhibits
1100	Debate Chairs: Munier Nour (Saskatoon), Mark Inman (Saskatoon)
	Be It Resolved That a GLP-1 Agonist Should Be Used As 2nd Line Treatment, Instead of Insulin, in Youth-Onset Type 2 Diabetes Pro: Sanjukta Basak Con: Brandy Wicklow
1200	John Bailey Award, CPEG Fellowship Awards & Closing Remarks
1230	Meeting Adjourns

Oral Abstracts

OR1

Hyperglycemia in Acute Lymphoblastic Leukemia: Evaluation of Risk Factors for Insulin Requirement During Induction Chemotherapy

Kristina Pabedinskas (1), Melissa Braschel (2), Amanda M. Li (3), Danya A. Fox (1)

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- (2) BC Children's Hospital Research Institute, Vancouver, BC. (3) Division of Hematology, Oncology, and BMT, Department of Pediatrics, University of British Columbia, Vancouver, BC.

Background: Hyperglycemia is a common complication of induction chemotherapy in pediatric patients with acute lymphoblastic leukemia (ALL); however, only some patients with hyperglycemia require insulin. Though risk factors for hyperglycemia have been proposed, risk factors for insulin requirement are not clearly defined.

Objective: To identify risk factors for hyperglycemia requiring insulin during induction chemotherapy in pediatric patients with ALL.

Methods: All patients between the ages of 1 and 18 years who were diagnosed with ALL at BC Children's Hospital between April 2007 and March 2022 were identified. Patients were excluded if they had pre-existing diabetes or had received high dose glucocorticoids within 2 weeks of diagnosis. Patient demographics, glucose values, and treatment data were collected retrospectively from medical records. Hyperglycemia was defined as two instances of fasting glucose ?7.0 mmol/L or random glucose ?11.1 mmol/L. Patient level data were summarized, and potential risk factors were examined using univariate and multivariate binomial and multinomial logistic regression.

Results: 443 patients met inclusion criteria: 193 (43.6%) female, median age 5 (IQR 3-9) years, 95 (21.4%) overweight or obese, 11 (2.5%) with Trisomy 21, 175 (39.5%) with high risk ALL. Sixty-four (14.4%) had hyperglycemia and 38 (8.6%) required insulin. In multivariate analysis, older age (odds ratio [OR] 1.30 per year, 95% confidence interval [CI]: 1.17-1.43), female sex (OR 1.89, 95% CI: 0.99-3.62), higher BMI (OR 3.11, 95% CI: 1.53-6.30), high risk ALL classification (OR 3.90, 95% CI: 1.51-10.08), and comorbid Trisomy 21 (OR 6.22, 95% CI: 1.33-29.12) were associated with hyperglycemia. Older age (OR 1.24 per year, 95% CI: 1.11-1.39), female sex (OR 2.41, 95% CI: 1.13-5.14), higher BMI (OR 2.52, 95% CI: 1.11-5.72), and high risk ALL classification (OR 5.18, 95% CI: 1.69-15.92) were associated with insulin requirement. Prednisone (versus dexamethasone) was associated with insulin requirement only in univariate analysis.

Conclusions: Older age, female sex, higher BMI, and high risk ALL were all associated with insulin requirement in our cohort. This could be used to inform a formal hyperglycemia screening protocol for children with ALL and may also help avoid delays in prescribing insulin after the onset of hyperglycemia in high-risk groups.

OR2

Short-term Impacts of a Gluten-Free Diet on Children and Adults with Type 1 Diabetes and Celiac Disease: Findings from the CD-LiFE Study

Daniel I. Weiman (1), Margaret A. Marcon (2), Antoine B.M. Clarke (1), Keiran Pace (1), Esther Assor (1), Charlotte McDonald (3), Fred Saibil (4), Heather A. Lochnan (5), Zubin Punthakee (6) & Farid H. Mahmud (1)

(1) Division of Endocrinology, Department of Pediatrics, The Hospital for Sick Children, University of Toronto. (2) Division of Gastroenterology, Hepatology and Nutrition, Department of Pediatrics, The Hospital for Sick Children. (3) Division of Endocrinology and Metabolism, St. Joseph's Health Care, Western University. (4) Division of Gastroenterology, Sunnybrook Health Sciences Centre, University of Toronto. (5) Department of Endocrinology, The Ottawa Hospital. (6) Department of Endocrinology, McMaster University.

Background: Evidence is limited on longitudinal health outcomes with a Gluten-Free Diet (GFD) in patients with type 1 diabetes (T1D) and Celiac Disease (CD).

Methods: 2-year follow-up study from the CD-DIET Clinical Trial (NCT01566110). Anthropometrics, glycemic management, gastrointestinal symptoms, health-related quality of life as well as nutritional intakes and celiac serology were evaluated annually in children and adults aged 8-45 years with established T1D and newly screen-detected CD.

Results: 62 participants (48 adults 14 children) were evaluated over the course of 166 study visits. 51.6% (n=32) were GFD adherent at study baseline and GFD adherence (assessed using gluten intake) remained high (n=24; 72.7%) after 2 years. GFD transition to a regular, Gluten-Containing Diet (GCD) occurred in 15.2% of participants while 10.3% elected to adopt a GFD. No differences were observed in HbA1c between individuals reporting a GFD and GCD (?=0.1%; 95%CI: -0.6 to 0.4; P=0.613) over the study period. No differences were seen in weight (?=0.2kg; 95%CI: -2.3 to 2.8; P=0.848) and BMI among adults (?=-1.1kg/m2; 95%CI: -0.4 to 0.6; P=0.149) nor BMI Z-Scores among pediatric participants (?=0.91; 95%CI: -0.34 to 2.16; P=0.136). No differences were observed in the number of GI symptoms reported by the GFD group relative to the GCD group (IRR=0.95; 95%CI: 0.6 to 1.5; P=0.810) nor was there a difference in the odds of reporting any GI symptom (OR=1.1; 95%CI: 0.5 to 2.2; P=0.846). Lastly, no longitudinal differences were observed between those in the GFD and GCD groups with respect to generic (?=-0.4; 95%CI: -4.0 to 3.3; P=0.840) nor diabetes-specific health-related quality of life (?=1.5; 95%CI: -2.9 to 5.9; P=0.505).

Conclusions: 2-year longitudinal follow-up of adults and children with T1D and asymptomatic, screen-detected CD demonstrates that adherence to a GFD does not impact glycemic control, anthropometrics, GI symptom development, nor quality of life. These results highlight that the GFD did not negatively impact important clinical outcomes, and dietary transitions were common.

OR3

Type 1 Diabetes Genetic Risk Score 2 to Estimate the Proportion of Non-Autoimmune Diabetes in Autoantibody Negative Patients

Jacob Nicodemo (1,2), Constantin Polychronakos (1,2)

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Background information: Diabetes mellitus is a common disease with diverse etiologies. Accurate diagnosis of a specific form is critical for appropriate treatment. Type 1 diabetes (T1D) is an autoimmune condition for which there is no definitive test. Islet autoantibodies can be used to confirm a T1D diagnosis, but their absence cannot exclude autoimmune etiology. As much as 10-15% of T1D patients are negative for all known islet autoantibodies. It is believed that most autoantibody negative patients do in fact suffer from autoimmune diabetes; however, many have been misdiagnosed. The exact proportion of autoantibody negative patients clinically diagnosed with T1D who suffer from non- autoimmune diabetes is unknown.

Purpose of the study: This study aimed to use the autoantibody status of patients and their family members as well as T1D Genetic Risk Score 2 (T1DGRS2) to estimate the proportion of autoantibody negative clinically diagnosed T1D patients who suffer from non-autoimmune diabetes, an important clinical unmet need, as many of these cases can respond to treatments other than insulin injections.

Methods: Test group patients were sourced from the T1D Genetics Consortium. Where possible, single-nucleotide polymorphisms (SNPs) considered in T1DGRS2 that were not directly available in the consortium databases were imputed using the T0PMed Imputation Server. Patients were then classified according to their autoantibody status and that of their families. The presence of an autoantibody positive family member in autoantibody negative patients was considered proof of autoimmune etiology. Non-T1D controls were sourced from the UK BioBank.

Results: A partial T1DGRS2 considering all HLA haplotypes, HLA classical alleles, and 22 of 37 intergenic-HLA and non-HLA SNPs was shown to be highly discriminative of T1D compared to non-T1D controls. Among autoantibody negative patients, 251 had no autoantibody positive relative (negative-family children) and 371 had at least one autoantibody positive relative (positive-family children). A frequency distribution of negative family children showed a bimodal distribution, with the area under the curve of the minor mode corresponding to undoubtably non-autoimmune diabetes (2.3%). The major mode of this distribution showed an increase in frequencies of scores below 11.25. The difference in the area under the curve for negative family children and positive family children below this threshold gave an estimate for the maximum proportion of non-autoimmune cases (9.8%).

Conclusion: Between 2.3% and 9.8% of autoantibody negative clinically diagnosed T1D patients have been misdiagnosed and suffer from non-autoimmune diabetes. The T1DGRS2 score can be used to target them for genetic testing.

Diabetic Kidney Disease Screening and Management Practices Amongst Canadian Paediatric Endocrinologists: A Nationwide Survey

Paul M Ryan (1), Edgar Delbert (2), Melissa Del Vecchio (2), Carrie Costello (3), Jill K Hamilton (4), Brandy Wicklow (2), Allison Dart (2), on behalf of the iCARE Study Team

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Background: Adolescents with type 2 diabetes (T2D) are 2-4 times more likely to develop renal failure than their type 1 diabetes counterparts. Although routine diabetic kidney disease (DKD) screening is therefore recommended, the current practice of Paediatric Endocrinologists has not been assessed.

Purpose: To establish the current DKD screening practice in paediatric T2D, determine comfort of Paediatric Endocrinologists in interpreting and acting on markers of kidney function, and determine barriers to uptake of a novel, paediatric T2D-specific estimated glomerular filtration (eGFR) formula as a means of DKD surveillance.

Methods: The anonymous REDCap survey was distributed to Canadian Paediatric Endocrinologists via the Canadian Paediatric Endocrinology Group in October 2023, with one reminder circulated in November 2023.

Results: Twenty-four Paediatric Endocrinologists (75% female), with a mean of 12.7 (SD 7.8) years of independent practice, responded. Most practiced in a tertiary setting (87.5%). All responders routinely screen for albuminuria at diagnosis and annually thereafter. Most would consider Nephrology referral with a confirmed albumin:creatinine ratio ?3mg/mmol (58%). A minority routinely use eGFR in their assessment of patients (17%), although there was no consensus on the formula applied. The reasons cited for not using eGFR included that it was not standard of care within their site (67%) and lack of comfort in interpreting (42%), calculating (33%) or acting upon an abnormal result (29%). When posed with a hypothetical case vignette of hyperfiltration, a quarter of responders recognized this as concerning, while over half were unsure of how to interpret such a trend (54%). Responders reported that they would be likely to use a novel, paediatric T2D-specific formula for GFR estimation if there was departmental uptake (71%) and it was endorsed by Diabetes Canada (62.5%) or CPEG (42%). Barriers to uptake included efforts to integrate it into their current electronic medical record system (62.5%), availability of a current serum creatinine (42%), and time required for assessment (17%).

Conclusions: Canadian Paediatric Endocrinologists demonstrated high fidelity to DKD screening guidelines in T2D. However, comfort with interpretation of eGFR values was limited. Efforts should be targeted towards education and automation of eGFR calculation with associated normative values aiding interpretation.

OR5

Fertility Preservation for Transgender and Gender Diverse Youth: Development and Evaluation of a Patient-Centred eLearning Module

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Background: Fertility preservation (FP) counseling is recommended for transgender and gender diverse (TGD) youth before initiating hormone blockers and gender-affirming hormones. However, rates of FP and counseling practices vary across centers, and information regarding relevant patient-centred education materials is limited.

Objectives: 1) develop a patient-centred eLearning module on FP for TGD youth; 2) evaluate the module's impact on youths' knowledge of and confidence in decision-making around FP; 3) assess youths' perceptions of the module.

Methods: Three-phase study involving 10-18-year-old pubertal TGD youth from SickKids Transgender Youth Clinic (TYC).

Phase 1: A cross-sectional needs assessment of TYC patients, semi-structured interviews with gender care and fertility medicine experts, and input from community stakeholders and patient- education experts.

Phase 2: Development of the online module using Phase 1 input with subsequent piloting of this module with TGD youth and healthcare providers. Revisions were based on feedback regarding content, language and length in preparation for Phase 3.

Phase 3: Completion of pre- and post-module questionnaires by 20 TGD youth prior to gender- affirming medical intervention.

Phase 3 outcomes: youths' knowledge of FP, confidence in informed decision-making regarding FP consultation, and perceptions of the module.

Results: Phase 1: Twenty-one youth (12 assigned females [AF], 9 assigned males [AM]). Learning preferences: written text (15/21), supplemental resources (14/21), and photographs (7/21).

Phase 2: Of 13 participants (7 youth and 6 healthcare providers), 12 reported the module content was clear and easy to understand. All 13 reported the module was acceptable and inclusive.

Phase 3 (Preliminary): 16/20 participants enrolled to date (14 AF and 2 AM). Mean age was 15.6 ± 0.8 years. After module completion, all 16 youth felt more knowledgeable about FP and 13/16 reported increased self-confidence in informed decision-making around FP consultation (3 neutral responses). All 16 were satisfied with the module and 15/16 reported that the content was informative (1 neutral response).

Conclusions: This module addresses the lack of patient-centred FP education resources for TGD youth and improves FP knowledge and self-confidence in decision-making around FP consultation. Broad dissemination will help to promote FP education across Canada, enhancing FP-related informed decision-making among TGD youth.

More Than Just Hypercalcemia: A Case of Neonatal Severe Hyperparathyroidism treated with Cinacalcet Monotherapy

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A term female infant presented with respiratory distress requiring positive pressure support secondary to perinatal depression at birth. Her chest x-ray showed a bell-shaped thorax and multiple bony abnormalities. Subsequent investigations were consistent with primary hyperparathyroidism: total calcium 3.38mmol/L (2.08-2.64mmol/L), PTH 155ng/L (6-89ng/L) and phosphate 1.22mmol/L (1.63-2.83mmol/L). Over the first few days, calcium remained elevated, PTH increased to >2500ng/L. Skeletal survey showed multiple bony abnormalities including suspected pathologic fractures with periosteal reactions. Hypercalcemia was initially managed with hyperhydration, calcitonin and bisphosphonates. The infant was noted to have hypotonia, tachypnea, respiratory distress and bicytopenia (anemia and thrombocytopenia). Whole exome sequencing revealed a homozygous mutation, variant of unknown significance (VUS), in the calcium sensing receptor gene (CASR: c.2333G>T(p. GLy778Val)), suggestive of a diagnosis of neonatal severe hyperparathyroidism (NSHPT). NSHPT is a rare disease due to inactivating mutations of the CASR gene. NSHPT presents with severe hypercalcemia, markedly elevated PTH, lethargy, hypotonia, respiratory compromise and fractures.

Ultimately this patient was treated with Cinacalcet monotherapy, however has required high doses, 9 mg/kg/day (range 0.4-11.5mg/kg/day), to maintain normalization of calcium and PTH. Despite resolution of hypercalcemia, chronic respiratory distress and hypoventilation persisted, requiring home oxygen and nocturnal BiPAP. She is now 2.5 years old and stable on cinacalcet monotherapy. Bicytopenia resolved by 4 months and respiratory support weaned off at 15 months.

This case emphasizes several important clinical features of NSHPT. While the degree of hypercalcemia was mild, the clinical presentation of NSHPT was multisystemic and severe. The VUS was eventually re-classified as a likely pathogenic mutation. Long-term monotherapy with cinacalcet has been safe and effective, although high doses have been required likely due to the location of the genetic mutation. Unlike previously reported cases, the respiratory distress did not improve shortly after normalization of calcium and its persistence was thought to be multifactorial, with hypotonia, neuromuscular weakness and bell-shaped thorax contributing. Finally, this case highlights the impact of PTH on the bone marrow. Unique to this case is the bicytopenia; despite extensive hematologic work-up, a specific etiology was not identified and was attributed to a leukoerythroblastic response secondary to hyperparathyroidism.

OR7

Use of Glucagon-like Peptide 1 Receptor Agonists in Adolescents with Prader-Willi Syndrome: A Case Mini-series

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Background: The glucagon-like peptide 1 receptor agonist (GLP-1RA) liraglutide has been effective in addressing weight gain in a small cohort of children with Prader-Willi Syndrome (PWS). However, just one adult case report is available on use of the more recently developed semaglutide in the syndrome.

Research Question: What is the experience of GLP-1RA use in adolescent patients with PWS?

Methods: A list of all patients prescribed semaglutide was autogenerated from the electronic medical record (n=71), while a staff-curated list of patients known to be prescribed a GLP-1RA within the Healthy Active Living Clinic was also reviewed (n=23). All patients with PWS were included, while other forms of genetic obesity, hypothalamic obesity or exogenous obesity were excluded. Data on body mass index (BMI), BMI z-score, transaminases, glycated haemoglobin (HbA1c), and triglycerides at initiation of therapy and at follow-up appointments around 3, 6, 12, 24 and 36 months thereafter were extracted. Clinic notes were reviewed for tolerance and adverse effects.

Results: Four cases of GLP-1RA use in PSW, three females and one male (mean: 17 years, SD:1.4), were identified. Two patients had 24 months follow-up, one had 6 months, while the final patient was excluded due to lack of follow-up. Liraglutide was prescribed initially in two cases, one of which was later transitioned to semaglutide, while one was GLP-1RA-naïve at the time of semaglutide initiation. Mean age at initiation was 16 years (SD:1.7), while mean BMI z-score was 3.4 (SD:0.8). Of the three patients with follow-up, the two prescribed semaglutide demonstrated decreases in their BMI-z scores (2.84 to 2.39 and 3.07 to 2.86), while the patient prescribed liraglutide experienced an increase (4.28 to 4.87). Alanine aminotransferase levels were decreased in all three patients for whom follow-up was available. In the patient with a diagnosis of T2D and follow-up, HbA1c decreased from 13.5% at baseline to 5.5% and 5.6% at 12 and 24 months, respectively.

One patient experienced nausea with liraglutide. There were no serious treatment-emergent adverse effects.

Conclusion: GLP-1RAs, and in particular the newer agent semaglutide, may be useful adjunct therapies in the global health management of children with PWS.

A single-center experience with pediatric thyroid nodules

Mallory McNiven (1); Andre Isaac (2); Safwat Girgis (3); Susan Chafe (4); Rose Girgis (1)

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Background: Pediatric thyroid cancer is a rare diagnosis; however, there is evidence that the incidence has been increasing in recent years. The majority of cases in pediatrics are due to papillary thyroid cancer, with follicular and medullary thyroid cancer being much less common. Most pediatric patients present with an asymptomatic thyroid nodule. Papillary thyroid cancer is often aggressive in the pediatric population, presenting with metastatic disease more often than adult patients. In comparison, follicular thyroid cancer is rarely invasive with a low risk for metastatic disease.

Study: We describe the clinical presentation, investigations and management of thyroid nodules seen from 2019 - 2023 at the Stollery Children's Hospital. We witnessed an increased incidence of thyroid cancer. 16 pediatric patients, aged 4-17 years old, presented with suspicious thyroid nodules on ultrasound. 7/16 patients were seen within the last year alone. They all underwent an ultrasound guided fine needle aspiration (FNA) biopsy. 8/16 cases were positive for malignancy. 7/8 cases had papillary carcinoma, while 1/8 had follicular neoplasm. 4/7 had metastatic disease at diagnosis. Out of the benign nodules, one patient presented with a DICER1 mutation, and another was found to have a PTEN mutation.

The patient with the indeterminate pathology of follicular neoplasm underwent molecular studies which were positive for a high risk ??PAX8/PPARG mutation. Total thyroidectomy was performed, with final pathology revealing follicular carcinoma. Surgical management included hemi or total thyroidectomy by 2 dedicated ENT surgeons. Post-operative complications included hypoparathyroidism and dysphonia. One patient developed airway obstruction a few days postoperatively due to abscess formation in the operative bed. All patients with malignant nodules received radioactive iodine ablation postoperatively and subsequently were started on Levothyroxine.

Conclusion: While pediatric thyroid cancer is a rare diagnosis, the incidence is increasing. Thyroid nodules require prompt recognition and investigation for malignancy. Molecular studies are valuable in further classifying indeterminate nodules and determining malignancy risk.

Hypothyroidism in Congenital Nephrotic Syndrome: A Case Series

Rachel Parker (1), Jonathan D. Wasserman (1), Farid H. Mahmud (1).

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Background: Congenital nephrotic syndrome (CNS) presents with proteinuria, hypoproteinemia and edema in patients less than 3 months old and is commonly associated with underlying genetic defects leading to disruption of the renal glomerular filtration barrier. Hypothyroidism is a common feature of CNS related to urinary loss of proteins including thyroxine (T4), triiodothyronine (T3) and thyroid binding globulin (TBG).

Rationale: We present 3 patients with CNS and hypothyroidism. Biochemical testing to optimize thyroid hormone replacement was monitored including mode of levothyroxine administration (IV versus oral), frequency of administration (daily versus twice daily (BID)) and dose (mcg/kg) required to normalize serum thyroid hormone levels. We hypothesized that due to the persistent and rapid protein loss requiring BID albumin infusions, IV and/or BID dosing of levothyroxine may help normalize thyroid function.

Cases: All patients carried pathogenic genetic variants identified: NPHS1 (1) variant which is the most common, and LAMB2 (2 and 3) associated with Pierson syndrome. Patients 1 and 3 had elevated TSH with low FT4, while patient 2 had inappropriately normal TSH with low FT4. All patients received IV levothyroxine due to NPO status (3) or lack of response to high oral doses (1 and 2). The maximum dose of levothyroxine for all patients ranged from 20-25mcg/kg/day orally and from 7.4-18.4mcg/kg/day IV which is higher than typical dosing for congenital hypothyroidism (10-15mcg/kg/day oral).

Findings: There was less need for dose adjustments while on IV levothyroxine, potentially reflective of poor absorption of oral levothyroxine due to hypoalbuminemic gut edema. It was unclear whether this was significantly different in daily versus BID administration. Confounding factors include non- thyroid illness, concomitant use of medications such as furosemide and steroids which can lower FT4 levels, timing of blood draws and frequency of monitoring which varied for each case (every 1-2 weeks).

Conclusions: IV levothyroxine could be considered in patients with CNS if FT4 does not normalize with oral dosing. It is unclear whether BID dosing was beneficial in patients with poorly controlled CNS. Higher levothyroxine dosing should be considered at initiation of management. The timing of blood draws at nadirs should be consistent to facilitate interpretation.

Short stature in Glucose transporter 1 deficiency syndrome

Sruthi Thomas (1), Laura Stewart (1)

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Glucose transporter 1 deficiency syndrome (GLUT1DS) is an autosomal dominant disorder, with 90% of individuals developing the disorder as a result of a de novo mutations in the SLC2A1 gene encoding the glucose transporter 1 (GLUT1). The transporter is ubiquitous in blood tissue barriers supplying most tissues energy demand, and mediates glucose transport across the blood-brain barrier.

An 18 month old with known developmental delays and abnormal eye movements noted at 4 months was referred to our clinic. He measured at the 2nd percentile (SD -2.05), with mid-parental height closer to the 50th percentile. There was no family history of short stature. A growth hormone stimulation test done at 2 years demonstrated a peak growth hormone (GH) level of 4.0 ug/L. He continued to track just below the 3rd percentile and was noted to have a normal IGF-1. A drop in growth velocity was noted at 11 years and he was started on growth hormone therapy. His growth velocity increased from 3.5 cm/year to 12 cm/year and achieved a final adult height of 177 cm placing him at the 54th percentile (SD +0.1). Additionally, he had improvements in neurocognitive and neuromuscular skills.

One possible pathophysiological link between GH deficiency and GLUT1DS is that GH has a role in the counter-regulatory hormonal response to hypoglycemia. This counter-regulatory response is blunted in patients with recurrent severe hypoglycemia. It has been suggested that given the chronic hypoglycemic state of the neural cells in GLUT1DS could cause relative GHD. This could explain the lack of robust growth in childhood, but no ongoing GH need in adulthood. In terms of cognitive function, IGF-1 does upregulate GLUT1, which might explain the neurological improvement. It is possible that earlier GH initiation during the critical time for brain development would have improved the patient's outcomes.

Since neurological symptoms often lead to the diagnosis, pediatric neurologists should be alert to the monitoring for short stature and pediatric endocrinologists should monitor for motor and cognitive developmental delay, seizures, deceleration of head growth, movement disorder with ataxia, dystonia, and spasticity in the context of short stature to suggest GLUT1DS.

Caregiver knowledge of hypoglycemia and hyperglycemia management in children and adolescents with type 1 diabetes

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Objective: To determine the current knowledge of the assessment and management of acute diabetes-related complications including severe hypoglycemia and diabetic ketoacidosis (DKA) among caregivers of patients with type 1 diabetes (T1D).

Methods: A cross-sectional survey was completed at a tertiary care pediatric hospital diabetes clinic from November 2021 to July 2022. Data was analyzed from all completed survey responses and included both Likert and evaluation-style questions.

Results: A total of 152 surveys were collected and 96 were included in the analysis. Caregiver knowledge on management of hypoglycemia was significantly better than their knowledge on hyperglycemia management (median score 100% vs 60%, p-value <0.001). There was no difference in caregiver knowledge around assessment of hypoglycemia or hyperglycemia (median score 88.9% vs 88.9%, p- value 0.909). There was no significant difference in overall knowledge as determined by percentage of questions correct between those on different blood glucose monitors (p-value 0.184) or whether the parent/caregiver or the patient were primarily responsible for T1D management (p-value 0.964).

There was no difference in total score by management type when adjusted using the Bonferroni method. There was also no significant interaction by level of caregiver involvement.

Conclusions: Caregiver knowledge of management of hypoglycemia was significantly better than their knowledge of management of hyperglycemia in pediatric T1D. Blood glucose monitoring or T1D management type was not found to be associated with any differences in caregiver knowledge in the assessment or management of hypoglycemia or hyperglycemia.

Assessing bone mineral accrual rates in non-binary adolescents on prolonged course of GnRH analogues

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Introduction: Transgender adolescents undergoing pubertal suppression therapy are known to be at an elevated risk of impaired bone accrual due to the absence of endogenous sex hormones. Gender-affirming hormone therapy offers protection for bone health in binary transgender individuals. However some adolescents who identify as nonbinary do chose prolonged courses of unopposed puberty blockers. As a result, they may experience suboptimal bone gain or worse experience bone losses, potentially detrimental to their overall bone health. BMC and aBMD-Z change may serve as valuable indicators not only of fracture risk but also of the trajectory of peak bone mass development. In contrast, BMC and aBMD velocity-Z scores are more indicative of the impact of recent events and changes that occurred within the previous year on bone accrual(1). This distinction is essential in clinical practice, as it allows for a more comprehensive understanding of a patient's bone health status.

Objectives: The primary objective of this case series is to study bone mineral accrual trends in non-binary transgender adolescents. Secondary objectives include evaluating the BMC and aBMD velocity-Z scores to assess the bone health accrual patterns in non-binary transgender adolescents who have not undergone gender-affirming hormone therapy.

Methods: This study is a retrospective, observational case series involving three non-binary adolescents aged 13 to 17years. Participants underwent assessments of bone health at baseline before starting pubertal suppression through dual-energy X-ray absorptiometry (DXA) scans and data was collected at subsequent intervals over one to three year period. BMC z- scores were calculated using LMS values and percentiles(2). The application of BMD and BMC velocity-Z calculation equations will offer a snapshot of the impact of recent events on bone accrual(1).

Results: Preliminary results indicate that non-binary adolescents on prolonged course of GnRH analogue monotherapy experience suboptimal bone accrual. The application of BMD and BMC velocity-Z calculation equations will capture more subtle variations in bone accrual patterns. The study's findings can play a role in guiding healthcare providers and transgender individuals, especially those who identify as nonbinary, in making informed choices about their healthcare, with a focus on maintaining and enhancing their bone health while respecting their gender identity and healthcare needs.

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Poster Abstracts

P1

PROGRES, a multi-country, non-interventional, prospective study of patients receiving human growth hormone treatment under routine clinical care: Study update

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Background and aims: Children with growth hormone deficiency (GHD) are usually treated with once-daily injections of recombinant human growth hormone (rhGH). Somatrogon is a long-acting rhGH approved in several countries for once-weekly treatment of children with growth failure due to GHD. The Pfizer Registry of Outcomes in Growth hormone RESearch (PROGRES) study will assess the long-term safety and effectiveness of once-weekly somatrogon and once-daily rhGH preparations under routine clinical care; PROGRES will also enable comparisons across different once-daily rhGH preparations.

Methods: This non-interventional, prospective, phase 4 study aims to enroll eligible patients (male/female; any age) from >20 countries. Enrollment is from September 2021-October 2029, with data collected until October 2030 (minimum 1-year follow-up). Study inclusion criteria include prescription of once-weekly somatrogon or one of the specified daily rhGH preparations (Genotropin, Norditropin, Humatrope, or Omnitrope) and provision of informed consent. Somatrogon-treated patients are eligible for inclusion if somatrogon is approved and commercially available in their country. Primary safety outcomes include adverse events (AEs), serious AEs, and AEs of special interest. Primary effectiveness outcomes include annualized height velocity (HV) and change in HV standard deviation scores (SDS) from baseline.

Results: As of July 2023, 496 patients have been enrolled in PROGRES, with 386 in the full analysis set (FAS). The data from the FAS are presented here. Idiopathic GHD is the most common primary diagnosis overall (81.2%) and, in each treatment group, most patients are non-naïve to treatment. The countries with the largest number of patients are the USA (162; 42.0%), Japan (83; 21.5%), and France (43; 11.1%); in all 3 countries, most patients are receiving Genotropin. Canada has 12 patients, with 2 receiving Genotropin, 3 receiving somatrogon, and 7 receiving another daily rhGH. In the FAS, the mean (SD) age of patients at baseline is 11.6 (3.11) years and 27.2% are female; mean height SDS, weight SDS, and BMI SDS (SD) at baseline are -0.9 (1.14), 0.9 (1.61), and 0.0 (1.15), respectively.

Conclusions: Currently, the FAS for PROGRES consists of 386 patients from 11 countries, most having a primary diagnosis of idiopathic GHD.

Impact of transition from symptomatic to routine celiac disease screening in a pediatric tertiary care type 1 diabetes clinic

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Background: Prevalence of celiac disease (CD) is approximately 5 times higher in children with type 1 diabetes (T1D) than the general population. While typical CD symptoms are gastrointestinal, clinical presentation is variable and often asymptomatic. In T1D, the only sign may be glycemic variability or hypoglycemia. Despite an increased clinical awareness, approach to screening in T1D is controversial and CD remains undiagnosed in many individuals with T1D if routine screening is not performed. After reviewing literature showing an association of CD (including asymptomatic) with osteoporosis, iron deficiency, growth failure, and potential increased risk of retinopathy and albuminuria for individuals with T1D, the CHEO diabetes team decided to change from symptomatic to routine screening for CD in all patients in their pediatric T1D clinic population of approximately 890 children and youth.

Objective: To describe the change in celiac screening and gatroenterology referral frequency in a pediatric tertiary care T1D clinic following a change from symptomatic (per Diabetes Canada 2018 guidelines) to routine (per ISPAD 2022 guidelines) CD screening in all children with T1D.

Design & method: In this first phase of a quality improvement initiative, data was retrospectively acquired from our electronic health record. Analysis compared the number of CD screening test results (Tissue Transglutaminase antibodies (TTG)) and gastroenterology referrals in the year before (August 10, 2021-22) and the year after (August 10, 2022-23) implementation of routine CD screening.

Results: The number of TTG results rose from 108 to 396 in the year pre- versus post-implementation of routine CD screening, an increase of more than 3.5-fold. Similarly, the number of referrals to gastroenterology increased by more than 2.5-fold, with a rise from 12 to 33 for the year pre- versus post-implementation of routine CD screening.

Conclusion: Changing from symptomatic to routine screening for CD in a pediatric T1D population generated a significant increase in the number of screening tests and gastroenterology referrals. Our next step is to evaluate the impact of this practice change on the incidence of CD diagnosis (symptomatic and asymptomatic) in our clinic and the cost of this intervention.

6-Month Subcutaneous Leuprolide Acetate Achieved and Maintained Hormonal and Clinical Suppression in All Weight Groups of Children with Central Precocious Puberty

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Background: Childhood obesity poses an increased risk for central precocious puberty (CPP). Data on whether weight affects the treatment of CPP would be helpful to optimize management. We present secondary analyses of pubertal hormone suppression, and height velocity (HV) data from the pivotal trial of the first small-volume, long-acting, subcutaneously administered gonadotropin- releasing hormone agonist (GnRHa) for CPP, with the objective to determine if study drug, ELIGARD® adequately suppresses puberty in overweight and obese children.

Methods: 62 children with treatment-naïve CPP received 2 doses of 45 mg subcutaneous leuprolide acetate (LA) at 24-week intervals over 48 weeks. The BMI percentile was calculated based using Center for Disease Control growth charts, accounting for height, weight, age, and gender. GnRHa- stimulated luteinizing hormone (LH) concentrations were assessed with a lower limit of detection of 0.100 IU/L. Estradiol (E2) concentrations were assessed with a lower limit of detection of 10 pg/mL. HV was calculated as the change in height between visits/[(number of weeks between visits)/52]. Results: Mean GnRHa-stimulated LH concentrations at screening, week 24, and week 48 were 24.5, 2.1, and 2.5 IU/L in non-overweight children, 10.5, 3.2, and 2.2 IU/L in overweight children, and 31.5, 4.8, and 1.9 IU/L in obese children, respectively. In these same groups, mean E2 concentrations at screening, week 24, and week 48 were 28.2, 10.5, and 10.3 pg/mL in non-overweight children, 17.8, 10.4, and 10.4 pg/mL in overweight children, and 25.9, 11.0, and 11.1 pg/mL in obese children, respectively. Mean HV at week 4, week 24, and week 48 was 10.2, 5.6, and 5.7 cm/year in non-overweight children, 7.7, 5.5, and 6.2 cm/year in overweight children, and 8.2, 4.9, and 6.6 cm/year in obese children, respectively.

Conclusions: A 45 mg dose of 6-month subcutaneous LA, ELIGARD suppressed LH and E2 to very low concentrations and reduced HV in all BMI groups, regardless of patient weight, without dose adjustment. This is consistent with results from previous studies of GnRHa efficacy in obese and normal-weight children with CPP. Clinicians should consider providing counseling and interventions that support healthy diets and lifestyles to children with above-normal BMI and their caregivers.

Central Versus Peripheral? A Diagnostic Challenge in Precocious Puberty

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Introduction: Precocious puberty, defined in males as the development of secondary sexual characteristics before the age of 9 years old, can have central or peripheral causes, and is rare in infants. Hypothalamic priming can also occur from exposure to high levels of sex steroids. We present a case of precocious puberty in a 9-month-old boy that posed a diagnostic challenge.

Case: A 9-month-old boy known for mild hypospadias was referred to Pediatric Endocrinology for presence of public hair. Pubarche occurred at age 7 months. The parents reported multiple daily erections since age 4 months and onset of body odor at age 7 months. Examination showed increased testicular volumes of 3 mL on the left and 2-3 mL on the right, scrotal laxity and rugation, an enlarged penis (6.5 cm in length), duvet public hair, incomplete glans, and presence of hypospadias. Laboratory investigations showed pubertal gonadotropin and testosterone levels (luteinizing hormone 5.5 IU/L, follicle-stimulating hormone 2.8 IU/L, total testosterone 14.28 nmol/L (compatible with Tanner stage 4-5)). Dehydroepiandrosterone sulfate was pre-pubertal. Electrolytes, 17-hydroxyprogesterone, and random cortisol levels were normal. Tumor markers of alpha-fetoprotein and human chorionic gonadotropin were negative. There was bilateral testicular enlargement (1.4 mL and 1.6 mL on the right and left, respectively) without any focal mass on testicular ultrasound. There was no suprarenal mass on abdominal-pelvic ultrasound. Brain MRI revealed a pedunculated non-enhancing mass in the left-sided suprasellar cistern arising from the hypothalamus consistent with a hypothalamic hamartoma, and a normal appearing pituitary gland. Genetic testing is pending. Treatment with monthly leuprolide depot injections was started.

Discussion: This case presented a challenge to identify a central versus peripheral cause of precocious puberty. The examination findings of extensive external genitalia androgenization relative to the testicular size (less than 4 mL; the volume indicative of true puberty), led us to believe a peripheral cause with secondary priming of the hypothalamus was most likely. However, a hypothalamic hamartoma, which is a central cause of precocious puberty, was found with imaging.

This patient, symptomatic from 7 months old, is one of the youngest infants with hypothalamic hamartoma amongst those reported.

Long Term Effects of Pediatric Hematopoietic Stem Cell Transplant on Endocrine Function

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Background: Advances in the treatment of childhood and adolescence cancer treatment have led to significant increase in survival rate. Current 5-survival rate of childhood cancer is nearly 80%. Hematopoietic stem cell transplant (HSCT) is treatment of choice in many clinical conditions including malignant and non-malignant hematological diseases, solid tumors and immunodeficiency diseases. Children receiving HSCT are prepared with different pre-transplant conditioning regimens. These regimens expose the recipients to potent cytotoxic chemotherapy and sometimes to fractionated or non- fractionated total body irradiation (TBI). Adverse effects on endocrine function and bone health are the most commonly found sequelae affecting 20%-50% subjects.

Method: We did a retrospective chart review of children age less than 18 years at the time of HSCT at King Faisal Specialist Hospital and research Centre Jeddah. They received conditioning regimen prior to HSCT either myeloablative (MAC) or reduced intensity (RIC) regimen and in few case no conditioning regimen was given. All participants were followed for a median period of 5 years. Endocrinology assessment carried out periodically including thyroid, adrenal, pituitary glands function and gonadal assessment upon expected physiological age of puberty.

Results: 141 children (75 males and 66 females) had HSCT at median age of 7 years. 128 patients had allogenic while13 patients had autologous HSCT. The conditioning regimen include MAC in 110 (78%), RIC in 28 (19.9%) and no condition in three (2.1%) children. Only 17 children received radiation; 13 had fractionated and four had non-fractionated. Nearly half of the patients (65/141, 46%) developed one of endocrine dysfunction. Girls are more effected than boys (48 vs 17). Growth hormone deficiency in 4 children (1 male and 3 females), adrenal insufficiency in 13 children (6 males and 7 females), thyroid deficiency in 23 children (13 males and 10 females) and gonadal failure in 36 children all females. Endocrine outcomes are compared against the primary disease and conditioning regimen. Gonadal failure has significant association with MAC (p-value 0.0002) however; other endocrine dysfunctions do not have any significant association with conditioning regimen or underlying disease.

Conclusion: Current treatment strategies for childhood cancers and hematological diseases have significantly improved long-term survival. However, this survival is associated with treatment associated adverse effects, which have an impact not only to physical health but also on psychosocial wellbeing of survivor. Our data suggests further improvement in conditioning regimens to avoid endocrine dysfunction.

Measuring adherence to clinical practice guidelines for screening of complications and comorbidities in children and youth living with type 1

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Objective: To assess adherence to the 2018 International Society for Pediatric and Adolescent Diabetes (ISPAD) screening guidelines for complications and comorbidities in children and youth with type 1 diabetes (T1D) in a tertiary pediatric diabetes clinic in Vancouver, British Columbia (BC).

Methods: Data were extracted using the BC-Pediatric Diabetes Registry (BC-PDR) that prospectively collects visit-level data on consented patients receiving care the BC Children's Hospital Diabetes Clinic. The BC-PDR includes demographic (i.e., date of birth, sex, ethnicity) and clinical data (i.e., date of diagnosis, body mass index (BMI), hemoglobin A1C) as well as screening metrics such as blood pressure (BP), urine albumin to creatinine ratio (ACR), thyroid stimulating hormone (TSH), lipid profile, and IgA + anti-tissue transglutaminase (TTG) antibodies. To descriptively assess adherence to the 2018 ISPAD guidelines, visit-level data were analyzed annually from 2017-2022 for patients with clinically diagnosed T1D between 2-18 years old with at least one clinic visit between May 1, 2021 – May 1, 2023.

Results: Among 355 patients, 56% (n=199/355) were male, and 59% (n=196/333) were White. At the most recent clinic visit, median age was 13 years [IQR 9.9 - 15.4] and median A1C was 7.6% [IQR 6.9% - 8.5%]. From 2017-2022, adherence to hypertension screening annually was 90.6%, 96.1%, 94.6%, 42.3%, 7.7%, and 54.9% (Figure 1), whereas nephropathy screening rates were 42.9%, 48.4%, 58.7%, 42%, 42.3%, and 28.8% (Figure 2). From 2018-2022, adherence to screening for autoimmune thyroid disease with TSH only was 81.9%, 81.5%, 72.2%, 73%, and 74.8% (Figure 3), whereas dyslipidemia screening rates were 0%, 8.6%, 11.8%, 9.3%, and 7.4% (Figure 4). In patients without prior celiac disease screening, 93.5% (n=174/186) had at least one celiac screen from 2017 - 2022.

Conclusion: Adherence to the 2018 ISPAD screening guidelines between 2017 – 2022 was highest for celiac disease and lowest for dyslipidemia. Understandably, hypertension screening during the COVID-19 pandemic declined; however, screening for nephropathy and dyslipidemia was low prior to the pandemic and remained low or declined further during the pandemic. Our results highlight the need to understand patient/family, healthcare provider and health system barriers that can impact screening.

Teens at MUG clinic: a study describing clinical and psychosocial characteristics

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Introduction: Disorders of sexual development (DSD) are a heterogenous group of rare diseases with atypical development of chromosomal, gonadal, or anatomic sex. This study aims to describe outcomes in patients with DSDs.

Method: In this cross-sectional study, we used chart review and validated questionnaires to describe medical, surgical and psychosocial data of patients aged 12-18 who attend the multidisciplinary DSD clinic of The Hospital for Sick Children. Psychosocial data included emotional/behavioural problems, gender identity/dysphoria, body image, resilience and Quality of Life.

Results: To date, we have collected complete data on 18 patients. Baseline medical and surgical characteristics are in Table 1. Initial results are in Table 2. Of 17 participants, only one respondent reported feeling uncertain of their gender.

Conclusion: Our data shows a diverse population, with good rates of social support and resilience, medium rates of body image satisfaction, and low rates of gender dysphoria. This study can assist Multidisciplinary Teams provide optimal patient-centered care.

Table 1: Baseline Characteristics - Data are n (%)	
Sex of Rearing	
Male	7 (26%)
Female	20 (75%)
Diagnosis	
Congenital Adrenal Hyperplasia	10 (37%)
Gonadal Dysgenesis	7 (26%)
Androgen Insensitivity	2 (7%)
- Partial	1 (4%)
- Complete	1 (4%)
Maternal Luteoma	1 (4%)
Hypogonadotropic Hypogonadism	1 (4%)
No Diagnosis Found	6 (22%)
Gonadectomy	
Yes	8 (30%)
Masculinising Surgery	
Yes	6 (86%)
Feminising Surgery	
Yes	11 (55%)

Table 2: Results – Data are n (%)	
I feel comfortable talking to my friends about	N 18
why I come to MUG clinic	
Strongly agree	2 (12%)
Mostly agree	4 (25%)
Mostly do not agree	5 (31%)
Do not agree at all	6 (25%)
Prefer not to answer	1 (7%)
My family stands by me during difficult times	N 18
A lot	13 (69%)
Quite a bit	4 (25%)
Somewhat	1 (6%)
I feel supported by my friends	N 18
A lot	11 (56%)
Quite a bit	3 (19%)
Somewhat	2 (13%)
A little	2 (13%)
I am satisfied with my body appearance	N 18
Very satisfied	3 (6%)
Satisfied	8 (50%)
Neutral	4 (25%)
Dissatisfied	1 (6%)
Very dissatisfied	1 (6%)
Prefer not to answer	1 (6%)

Bridge Over Troubled Water; Diagnostic uncertainty in a patient with transient diabetes insipidus.

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Introduction: Diabetes Insipidus (DI) can be a challenging diagnosis to establish; however delineating Central DI (CDI) from Nephrogenic DI (NDI) is typically straightforward. Here we present a case of transient DI which was initially managed as NDI, yet developed features overlapping with CDI. The underlying cause remains uncertain.

Case Description: We present a case of a 15yo boy with a background of Myelodysplastic Syndrome (MDS), on chemotherapy (Fludarabine, Cyclophosphamide, and Treosulfan) awaiting Bone Marrow Transplant (BMT). He was admitted with fever and hypotension due to sepsis requiring multiple fluid boluses. He was diagnosed with a hepatic abscess. Within two days of admission, he developed polyuria and polydipsia, with serum osmolality 308 mmol/kg H2O, serum sodium 154 mmol/L, and urine osmolality 136 mmol/kg H2O. Urine output was 7900ml in 24 hours. He was initially assigned a diagnosis of NDI, managed with fluid restriction, however this failed to suppress his urine output. He was also started on amphotericin B (a known cause of NDI) although after the onset of DI symptoms. Endocrinology was consulted and initiated desmopressin therapy. He required high doses, up to 900 mcg oral tablets per day, to maintain normal urine output. A copeptin of 3.9 pmol/L [2.4-8.6] was non- diagnostic. MRI sella turcica identified mild pituitary stalk thickening and an absent bright spot.

Anterior pituitary function was normal. He underwent successful BMT two months later, and his DDAVP requirements decreased. 9 Months later, he successfully discontinued DDAVP and demonstrated normal urine concentrating ability.

Conclusion: We present a diagnostic conundrum; a case of DI with multiple potential etiologies. CDI has been described as a rare complication of MDS due to lymphocytic infiltration of the pituitary stalk. ADH resistance (NDI) can also be induced with renal fluid overload, known as medullary washout.

Amphotericin B has also been described in case reports to cause NDI. Our patient successfully weaned from DDAVP, although whether this was due to CDI that was treated with BMT or due to transient NDI remains elusive. Establishing the underlying cause early has important implications for management as CDI requires ADH replacement, whilst NDI management varies by cause.

Primary Hypothyroidism Presenting with Ovarian Hyperstimulation

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Background: Prolonged hypothyroidism in children often leads to short stature with delayed puberty and bone maturation. However, the contradictory occurrence of peripheral precocious puberty in protracted juvenile hypothyroidism was reported in 1960. The initial hypothesis suggested TRH stimulation led to excess FSH and prolactin. Now, it is postulated that cross-activation occurs as LH, FSH and TSH share an alpha subunit with amino acid overlap.

Case 1: A 7-year-old female presented to the ED with 6 days of abdominal pain, anorexia, and emesis and was treated for constipation. She returned with worsening pain with an acute surgical abdomen. Ultrasound revealed a mass-like structure (6.6x3.4x6.7cm) with enlarged follicles. She underwent right ovarian laparoscopic detorsion. She re-presented 5-days later requiring surgical drainage of a 6-cm right complex hemorrhagic cyst. Further history revealed fatigue, cold intolerance and poor growth. Investigations: TSH >1000 mU/L, fT4 3.1 pmol/L, TPO antibodies 202 kIU/L, estradiol <30 pmol/L, LH <0.3 IU/L, FSH 1.7 IU/L, LDH 388 U/L, AFP < 9 ug/L, CA-125 57, and no bone age delay. Her height was 108.8cm (1%ile), weight 16.2 kg (<1%ile), and she had thelarche (B2) but no goiter.

Case 2: A 6-year-old female was assessed as an outpatient for 5 months of hematuria. Pelvic ultrasound showed bilateral ovarian cysts with septations, follicles and a pubertal-appearing uterus. She was later referred to pediatric gynecology from the ED for 3 months of vaginal bleeding. History revealed 3-years of growth deceleration, weight gain, fatigue, cold intolerance and constipation.

Investigations: TSH >1000 mU/L, undetectable fT4, TPO antibodies 44 kIU/L, prolactin 155 ug/L, FSH 7.8 IU/L, LH <0.3 IU/L, estradiol 235 pmol/L, elevated ALT, dyslipidemia and delayed bone age (2-years 6-months). Height was 100.5 cm (<1%ile) and BMI 23.46 kg/m2 (>99 %ile). She had a puffy face, no goiter, and was prepubertal.

Discussion: These cases highlight that despite hypothyroidism being clinically obvious, the diagnosis was initially missed as the unusual symptoms of ovarian hyperstimulation took precedence. Although rare, a raised awareness of this clinical entity amongst family physicians and other specialists such as ED physicians and surgeons, would be desirable to avoid compromised final height and/or ovarian complications.

Evaluation of a Multidisciplinary Pediatric Insulin Resistance and Type 2 Diabetes Program: The BC Children's Hospital Experience

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Background: Recent Canadian surveillance data demonstrated incidence of pediatric Type 2 Diabetes (T2D) has increased by 60% over the last decade highlighting the need for dedicated care in this population.

B.C. Children's Hospital has provided multidisciplinary care for children and adolescents living with insulin resistance (IR), prediabetes and T2D since 2003. Our study aims to assess outcomes in participants within this specialized program.

Methods: We conducted a retrospective review (n=250 patients) who completed an initial assessment between 2014-2023 and followed for a minimum of 6 months. Information was gathered on (1) patient characteristics (2) clinical variables (initial presentation, presence of other diabetes related diagnoses, anthropometrics, HbA1C, rates of complications and remission) (3) health care utilization and (4) psycho-social factors. Analysis will be divided in 4 distinct time periods of 2014-2018 (CDA 2013 guidelines), 2018-2020 (CDA 2018 guidelines), 2020-2021 (COVID telemedicine only) and 2022-present (ISPAD 2022 guidelines).

Results: Thus far, we have completed a preliminary analysis of 58 participants, enrolled between January-November 2023 with median age of 13.6 years (IQR: 12.4 - 14.5) with 57.5% having T2D, 35% prediabetes and 7.5% with insulin resistance; 52.5% of the cases were diagnosed in context of obesity related screening. The average time between diagnosis and initial consult was 13.1 weeks (IQR: 0 - 21.9).

In the prediabetes cohort, initial HbA1c was 6.1% (5.9-6.3), with average change of HbA1c during the first year -0.25% (-0.6 - -0.1); 64.3% managed with lifestyle education alone, and 35.7% receiving metformin.

Among the T2D cohort, initial HbA1c was 8.5% (7-11.1), and the average change of HbA1c during the first year was -2.5% (-3.9 - -0.8). 95.7% were prescribed metformin, and 3 cases discontinued in context of diabetes remission; 39.1% were prescribed on basal insulin, and 26.1% on rapid-acting insulin. 100% discontinued rapid-acting and 56.6% basal insulin. In other medications, 34.8% was prescribed on GLP1 agonist. Data collection and analysis is ongoing for additional outcomes.

Conclusions: This preliminary data shows the benefit of a dedicated multidisciplinary program in improving the care of youth living with T2D and IR, with positive outcomes in HbA1c and implementation of current pediatric diabetes care guidelines.

Trends and Risk Factors for Severe Hypoglycemia in Type 1 Diabetes in a Pediatric Diabetes Clinic

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A known barrier for children with Type 1 diabetes (T1D) achieving glycemic targets is the concern and risk for hypoglycemia, including severe hypoglycemia. Risk factors for severe hypoglycemia include prior episodes of hypoglycemia, lower hemoglobin A1c (A1C), hypoglycemia unawareness, long duration of insulin therapy (long duration of diabetes), preschool age, and adolescence. There is some evidence that increased use of and advancements in diabetes-related technology, including continuous and flash glucose monitors (CGM, FGM) and insulin pumps, have demonstrated reductions in hypoglycemic events in youth with T1D. We reviewed the frequency of severe hypoglycemic events in our pediatric diabetes clinic over a four-year period and its association with known risk factors for severe hypoglycemia including age, diabetes duration, prior hypoglycemic events, A1C, frequency of clinic attendance, insulin regime and modality of glucose monitoring.

Severe hypoglycemia was defined as measured hypoglycemia (< 4 mmol/L) associated with decreased level of consciousness, necessitating the assistance of another person. Data from clinic visits was extracted from electronic medical records, organized in Microsoft Excel and analyzed using SPSS.

In 2019, 17 hypoglycemic events occurred in 14 patients, with 3 patients experiencing 2 events each.

In 2020, there were 23 hypoglycemic events in 17 patients, with 2 patients each experiencing 2 events and another 2 each experiencing 3 events. In 2021, there were 26 hypoglycemic events in 20 patients, with 1 patient experiencing 2 events, 1 experiencing 3 events and 1 experiencing 4 events.

In 2022, there were 10 hypoglycemic events in 9 patients with 1 patient having 2 events (Table 1). Between 2019-2021, severe hypoglycemic events per 100 patient years ranged from 3.09-4.73, but decreased in 2022 to 1.82.

Overall, there is variability among risk factors for severe hypoglycemia in patients experiencing severe hypoglycemic episodes from 2019 through 2022. Corresponding with a decrease in severe hypoglycemic events in 2022 is an increasing trend in the proportion of patients utilizing CGM/FGM. This is consistent with overall clinic trends. It is unclear if this association is significant. Further assessment over the next several years is required, particularly as the use of insulin pumps and CGM/FGM continues to increase.

Integrating exome sequencing into a pediatric endocrinology clinic: A Genetics and Endocrinology Collaborative Quality Improvement Initiative.

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Background: Exome sequencing (ES) has emerged as a fast and efficient tool for detection of human genetic disorders. In the field of pediatric endocrinology, the adoption of ES will become a routine diagnostic. The Provincial Medical Genetics Program (PMGP) and the Division of Genome Diagnostics (DGD) are leading a quality improvement (QI) initiative with the pediatric endocrine clinic at BC Children's Hospital to streamline access to WGS for eligible patients and to improve the diagnostic access and care.

Methods: Patients with differences of sexual development (DSD), short stature, and hypopituritism were screened using a simple questionnaire to identify those eligible. Eligible patients were scheduled for an appointment in the endocrine clinic in collaboration with the Provincial Medical Genetics Program (PMGP). During the clinic visit, patients were assessed by an endocrinologist and reviewed by a geneticist. In the initial phase, a genetic counselor and the endocrinologist counseled the patient and obtained informed consent. Subsequently, this counseling process was conducted by the endocrinologist with the aid of an online genetic education tool. When the WGS results became available, either a geneticist or genetic counselor and the endocrinologist discussed the findings with the patient and the family.

Results: This initiative allowed interested endocrinology providers to assess the eligibility of their patients for ES and appropriately to counsel and to order testing. Eligible patients avoided the consultation with the PMGP and thus accelerated access to testing.

Conclusions: This collaborative approach educated endocrinology providers and thereby empowered them to make informed decisions regarding ES for their patients leading to more efficient access to genetic testing and diagnosis.

Pituitary Stalk Interruption Syndrome: A Rare Case Presenting with Arthrogryposis and Type 1 Diabetes Mellitus

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Introduction: Pituitary stalk interruption syndrome is a rare congenital abnormality of the pituitary gland. It is characterized by a classic triad, including an interrupted pituitary stalk, absent or ectopic posterior pituitary, and anterior pituitary hypoplasia or aplasia. Clinical presentation varies with age, often appearing as short stature and anterior pituitary hormone deficiency in adults. Notably, there are limited cases in the literature of co-occurring arthrogryposis and hypopituitarism. Recent research has unveiled variable hypopituitarism associated with arthrogryposis, linked to mutations in genes such as MAGEL2 and L1CAM, which code for endosomal processing and cell adhesion, respectively.

Case: A 17-year-old female with severe amyloplastic arthrogryposis was referred to the BCCH ER with concerns of a new diagnosis of diabetes mellitus. This followed a routine blood test that revealed a random blood glucose of 20.7 mmol/L. Notably, blood testing conducted three months earlier by her GP had shown an A1C of >14.0%, but no action had been taken, and the family was unaware of this result. Initial blood work in the ED was consistent with new diagnosis of diabetes without ketoacidosis. The patient was subsequently diagnosed with type 1 diabetes, based on the presence of positive GAD65 autoantibodies (489.8 IU/mL, reference range (RR) <5), and she was started on insulin therapy. During her assessment, she exhibited Tanner stage 3 breast development, but she had not had menarche. Further pituitary screening revealed multiple pituitary hormone deficiencies: low free T4 (6.8 pmol/L, RR 8.0–15.0), low morning cortisol (134 nmol/L, RR 240–618), low IGF-1 (<7 μ g/L, RR 155–485), low LH (<0.1 IU/L), low FSH (<0.3 IU/L), and low estradiol (<60 pmol/L); prolactin was 11.0 μ g/L (RR 0–24). A brain MRI showed a small anterior pituitary gland, markedly hypoplastic infundibulum, and an ectopic posterior pituitary bright spot, consistent with a diagnosis of pituitary stalk interruption syndrome. Exome sequencing was obtained, and results are pending.

Conclusion: This case highlights the rare co-occurrence of arthrogryposis, type 1 diabetes, and pituitary stalk interruption syndrome. This case underscores the importance of considering multiple medical conditions when evaluating patients with complex clinical pictures. It is likely that a genetic cause underlies this complex presentation. Therefore, the pending exome sequencing result holds great potential for shedding light on the underlying etiology in this case.

Comparison of Characteristics of Children with Diabetes and their Outcome in Haiti Compared to Countries with Different Economic Background

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Background: According to the World bank classification, 34% of children under 15 years old living with type 1 diabetes are from low- and lower-middle income countries. In Haiti, both economic as well as political / security topics heavily impact the outcome of pediatric diabetes care.

Objectives: To investigate differences in demographics and outcomes among patients with T1D aged <25 years, with diabetes duration >3 months, treated between 2021 and 2022, and compare Haiti centres to low-income, lower-middle income, upper-middle income, and high-income countries.

Methods: We analyzed the 07/2023 SWEET database and compared average age, presentation at onset, gender, insulin regimen, total daily insulin doses, HbA1c, BMI and rate of hypoglycemia between Haiti and 4 groups of countries, adjusted for age, sex, duration and CSII use.

Results: Haitian patients are predominantly females (58.2%) unlike the other groups (50.1/47.1/47.5 and 47.3% for countries with increasing GDP). Use of prandial analog insulin is extremely low among Haitian patients (1.3%) vs 70.8 vs 93.0 vs 99.6 and 99.9 for the four other groups. Daily insulin administered was lowest in Haiti (0.75 U/kg) compared to 0.86, 0.78, 0.81 and 0.84 for the other groups. Based on WHO reference, BMI-z-score was lowest in Haitian patients (-0.46) compared to 0.19, 0.54, 0.72 and 0.82 in the 4 groups of countries with increasing GDP.

Average HbA1c achieved was 12.2% in Haiti compared to 8.1/7.5/7.8 and 8.3% in the four comparison groups. Rate of severe hypoglycemia (definition: help required) was higher in Haiti (0.84 events/person-year) compared to 0.08, 0.05, 0.03 and 0.03 for the 4 comparison groups with increasing GDP (all p<0.0001).

Conclusion: Even though organizations such as Life for a Child work hard to reduce the gaps in T1D care and outcome between countries, results from the SWEET registry suggest that there are still huge differences in outcome for children with T1D around the world, and those differences are partially associated with the economic background / GDP of the countries. The closest linear relationship was present between BMI-z-score and GDP quartile.

Pheochromocytoma and transient Central Hypothyroidism

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Pheochromocytoma is a rare catecholamine-secreting adrenal tumor characterized by severe episodic hypertension, associated with significant morbidity and mortality. Clinical features related to hypertension include headaches, palpitations, and sweating. However, manifestations can mimic other conditions, leading to erroneous diagnoses such as anxiety, thyrotoxicosis and intracranial lesions.

We present the case of an 11-year-old girl, previously healthy, who presented to the emergency department with intermittent headaches. She was referred to endocrinology for possible central hypothyroidism with normal TSH:0.91 mIU/L (0.34-5.60) and low thyroxine:5.10mIU/L (8.0-18.0) after ruling out a brain mass on imaging. Resting blood pressure was elevated at 160/100. She was admitted for severe hypertension management. Further imaging of the adrenal gland revealed a 5 cm left adrenal mass, suspicious for a pheochromocytoma given the elevated plasma norepinephrine level 27400pg/ml (<3600). Management involved blood pressure stabilization before surgery, utilizing continuous IV Nicardipine, PO Amlodipine, and hydralazine as needed with continuous blood pressure monitoring. After this medical treatment was initiated, her diastolic blood pressure decreased to 80, and she continued with alpha-blocker (Doxazosin). Repeated Thyroxine 5 days later showed a normal value: 9.60 (8.0-18.0). After blood pressure stabilization, surgery to remove the pheochromocytoma followed, involving a multidisciplinary team of specialists in nephrology, pediatrics, endocrinology, and surgery. Her blood pressure normalized with no further anti hypertensive medications.

This case underscores the intricate interplay between pheochromocytoma and central hypothyroidism. It has been reported in the medical literature transient elevation of Thyroid Stimulating Hormone (TSH) or decrease of Thyroxine (FT4) during acute non-thyroidal illness. Prompt recognition of pheochromocytoma presentation, coupled with comprehensive biochemical and radiological assessments, is crucial for accurate diagnosis and effective management.

Keywords: pheochromocytoma, headache, hypertension, central hypothyroidism

Early onset obesity and Adrenal Insufficiency caused by POMC (Proopiomelanocortin) Deficiency

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A case of POMC deficiency: challenges and opportunities for treatment

The POMC gene encodes the preproopiomelanocortin (POMC) protein, which is sequentially cleaved to generate several active biopeptides, including adrenocorticotrophin (ACTH), melanocyte- stimulating hormones (MSH), and the opioid-receptor ligand beta-endorphin. Proopiomelanocortin (POMC) deficiency is an extremely rare disorder characterized by early-onset obesity, adrenal insufficiency, red hair, and decreased skin pigmentation. Hyperphagia, cholestasis, exponential weight gain and adrenal insufficiency are typically observed during the first months of life.

We describe a case of suspected POMC deficiency with early onset extreme obesity, hypothyroidism and adrenal Insufficiency in a five-year-old girl who is a recent newcomer to Canada.

Although prior growth charts were not available, the patient's anthropometrics are as follows: weight: 64 kg (>99th percentile), height: 134 cm (>99th percentile), BMI: >99th percentile. Our patient was born to consanguineous parents and has 5 siblings without any known genetic disorders. She experienced prolonged hospitalization in the first year of life and was started hydrocortisone ~10 mg/m2/day for adrenal insufficiency and Levothyroxine when she was one year old. In the first few years of her life, she experienced multiple hospitalizations for unclear cause, and developed a pattern of rapid and uncontrolled weight gain despite lifestyle intervention. She has not had any pharmacologic treatment. Genetic testing was completed prior to arrival in Canada according to parents, however there is no formal or translated documentation available.

Genetic confirmation and offering treatment with Setmelanotide are the next steps. Setmelanotide is a medication that acts on the melanocortin-4 receptor (MC4R) for the treatment of severe obesity caused by genetic disorders, including deficiency of proopiomelanocortin (POMC), proprotein convertase subtilisin/kexin type 1 (PCSK1), or leptin receptor (LEPR). We present a brief summary of evidence supporting the use of Setmelanotide in this population and our specific plans for coverage via the "Uncovering Rare ObesityTM" program sponsored by Rhythm Pharmaceuticals, Inc

The Risk of Growth Hormone Therapy in Patients with ROHHAD Syndrome

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Background: Rapid-onset obesity with hypothalamic dysfunction, central hypoventilation and autonomic dysregulation (ROHHAD) syndrome, is a rare disorder with no known etiology but devastating morbidity. Patients present in early childhood with rapid onset obesity followed by hypothalamic dysfunction including disorders in growth, hypothyroidism and dysnatremia associated with impaired fluid balance. They can also develop autonomic dysregulation with ophthalmologic, cardiac and gastrointestinal effects. Many of these patients develop neuroendocrine tumors (ROHHAD-NET).

Case: A 4 year old previously healthy girl presented with hyperphagia, pedal edema and rapid onset obesity with an 18kg weight gain over 8 months. She had central hypoventilation and was biochemically but not clinically growth hormone deficient. At the age of 9 her mother started to limit her fluid intake due to pedal edema despite normal serum electrolytes. At age 10 she had an incidental finding of a ganglioneuroma at the level of T4 to T5 which was inoperable. Six months later she was commenced on GH therapy due to a steadily declining growth velocity and GH levels. After 6 months of GH therapy she presented with agitation, raised ICP with dysnatremia. She had a subarachnoid hemorrhage and initially assessed as having diabetes Insipidus which was ruled out and GH was stopped. A diagnosis of paraneoplastic encephalopathy secondary to ROHHAD (NET) was made and she was started on immunotherapy and symptoms improved.

Discussion: GH therapy is proven and effective in the management of growth hormone deficiency. It is associated with rare but serious side effects including fluid retention, raised ICP and increased risk of tumor growth. GH increases IGF-1-mediated production of CSF from the choroidal plexus, leading to increased ICP. Patients with ROHHAD syndrome have abnormal fluid balance and dysnatremia. GH therapy could exacerbate this and increase their risk of cerebral edema and raised ICP. The development of neuroendocrine tumors in ROHHAD syndrome is common. There is the possibility of increased risk with GH therapy but there is not much research in this area. In her case, the tumor did not grow. Growth hormone is a treatment that can be commenced in these children but should initiated with caution.

Hyperosmolar Hyperglycemic Syndrome and Diazoxide: A case report in a child with Kabuki Syndrome

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Background: Diazoxide is a commonly used first- line pharmacologic therapy for treatment of hyperinsulinism. While hyperglycemia may occur with the use of diazoxide, Diabetic Ketoacidosis (DKA) and Hyperglycemic Hyperosmolar State (HHS) secondary to diazoxide is an exceedingly rare, but potentially life-threatening, adverse effect of diazoxide. We present a case of a 2-year-old female with hyperinsulinism in the context of Kabuki syndrome who presented with HHS, describing the diagnostic evaluation and treatment course.

Case presentation: The patient is a 2-year-old female with Kabuki Syndrome who was diagnosed with hyperinsulinism at 1 month of life and had been managed with diazoxide (3.75 mg/kg/day). She presented with 4 days of fever, cough and lethargy, requiring oxygenation and fluid resuscitation. Her parents had not monitored her blood glucose levels at home in the days prior to her presentation.

Initial diagnostic workup indicated HHS, with her biochemical profile demonstrating a serum glucose of 47.1 mmol/L, corrected sodium of 166 mmol/L, serum osmolality of 357 mmol/kg H2O, and no metabolic acidosis (venous blood gas with pH of 7.34, bicarbonate of 36 mmol/L) or ketosis (negative urine ketones). She was also positive for influenza B. She was admitted under general paediatrics with endocrinology, nephrology and critical care consultation. Her management involved discontinuation of diazoxide and IV hydration at 1.5 times maintenance rate. She did not require insulin therapy. Her course was complicated by an acute kidney injury (peak creatinine of 83 umol/L) but she did not experience venothromboembolism, rhabdomyolysis or cerebral injury. Her hyperosmolarity and hyperglycemia resolved approximately 36 hours after her initial presentation.

Upon resolution of HHS and after reintroduction of her home feeds, she successfully passed a 6-hour fast while off of diazoxide and was discharged home off this medication.

Discussion: HHS is associated with high morbidity and mortality rates. We present a case of HHS in a child on diazoxide therapy. This is an exceedingly rare entity with only five prior reports in the literature. This case highlights both the need for early recognition and prompt management of diazoxide-related complications, including DKA and HHS.

Application of DKA Risk Mitigation Algorithm for ATTEMPT Clinical Trial Evaluating SGLT2i in Youth with T1D

Aliya Allahwala (1), Antoine Clarke (1), Cheril Clarson (2), Petter Bjornstad (3), Samantha Anthony (4), Lynne McArthur (4), Farid H. Mahmud (1,4) on behalf of the ATTEMPT Writing Group

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Background: ATTEMPT (Adolescent Type 1 diabetes Treatment with SGLT2i for hyperglycEMia & hyPerfilTration Trial) is designed to evaluate the impact of Dapagliflozin (a sodium glucose cotransporter 2 inhibitor (SGLT2i) on renal and glycemic outcomes in adolescents with Type 1 Diabetes (T1D), given the benefits of this medication class on renal, metabolic, and cardiovascular outcomes in adults with Diabetes. However, there is an association between SGLT2i use and risk of ketosis and euglycemic Diabetic Ketoacidosis (eDKA), related to increased fatty acid mobilization and volume depletion.

Objectives: To describe the development and operationalization of a risk mitigation strategy through an early recognition, monitoring, and actionable intervention approach.

Methods: Existing DKA mitigation protocols were reviewed and the STOP-DKA protocol was adapted with permission due to its defined, tiered approach based on blood ketone measures and blood sugars that allowed for clear instructions for insulin, fluids and carbohydrate ingestion. Focus groups of research staff, physicians, educators and the trial research Patient and Family Advisory Committee (rPFAC) provided feedback that tailored the protocol to include separate injection and pump therapy guides with additional graphics, plain language, and clearer instructions. As part of the study, participants and families are provided a ketone meter, strips and a fact sheet outlining the symptoms of DKA, situations associated with elevated risk of ketosis and the ATTEMPT STOP-DKA protocol that was reviewed at study visits. Additionally, a wallet card was provided to share with health care providers informing them that they are enrolled in this drug trial and may present with an atypical DKA.

Results: ATTEMPT has completed enrollment and will report objective outcomes as well as patient- centered, qualitative outcomes related to ketosis, DKA risk and lifestyle factors.

Discussion: Clinical trials to evaluate the efficacy of SGLT2i in addition to details regarding their safe and appropriate use in the T1D population are important. Knowledge dissemination from the ATTEMPT trial will be valuable as to the potential implementation of a practical risk mitigation strategy using the ATTEMPT STOP-DKA approach in youth with T1D.

Optional Non-Accredited Industry Symposia (All times are listed in local time)

Thursday, February 8, 2024 (Room: Michelangelo AB)

Lunch Session: 1145-1215

Tech-Tastic: Transformation of Pediatric Diabetes Care Through Digital and Data **Driven Innovation**

IMAGINE... an online platform that easily and seamlessly integrates data from clinical encounters and data generated by patients, including CGM and insulin data, in a SINGLE, SECURE, TRUSTED DIGITAL SPACE accessible to patients, caregivers and their health care team.

The BC Childrens CareHub provides continuous data synchronization into a dashboard viewable by the patient/family and HCP. In this pre-CPEG bonus session, Dr. Shazhan Amed will describe how the CareHub digital platform works and share preliminary feedback from HCP and family/patient users as well as planned system improvements.





Dr. Shazhan Amed MD FRCPC MSc.PH Head, Division of Endocrinology Department of Pediatrics, Faculty of Medicine Clinical Professor, University of British Columbia

Friday, February 9, 2024 (Room: Michelangelo AB)

Lunch Session: 0745-0815

A Practical Approach to Identifying Rare Genetic MC4R Pathway Diseases of Obesity

In this session, participants will learn:

- How the melanocortin-4 receptor (MC4R) pathway regulates satiety in our brain
- How early-onset obesity and hyperphagia can lead to a diagnosis of MC4R pathway diseases, including Bardet-Biedl syndrome
- How you can set up a multidisciplinary pediatric obesity clinic and integrate genetic testing to identify MC4R pathway diseases
- How setmelanotide can help with weight management in patients living with obesity due to POMC, PCSK1, LEPR deficiency, or Bardet-Biedl syndrome





Dr. Marina Ybarra MD MSc Director, Pediatric Weight Management Program **London Health Sciences** Centre - Children's Hospital Assistant Professor, Dept. of Pediatrics, Western University

Optional Non-Accredited Industry Symposia (All times are listed in local time)

Friday, February 9, 2024 (Room: Michelangelo C)

Lunch Session: 1245-1315

(Engage the Speaker) Long-Acting Growth Hormones in Pediatrics: What Do YOU Think?

Explore the first clinical experience with long-acting growth hormones in children with GHD alongside Dr. Preetha Krishnamoorthy, and voice your opinions on where pediatric once-weekly growth hormone replacement therapy fits in your clinical practice. This is an interactive symposium where audience discussion is highly encouraged.





Dr. Preetha Krishnamoorthy MDCM FRCPC Associate Professor, McGill University Pediatric Endocrinologist The Montreal Children's Hospital

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