CPEG GCEP

Canadian Pediatric Endocrine Group
Groupe canadien d'endocrinologie pédiatrique

2020 CPEG Scientific Meeting
Official Program

February 20-22, 2020

The Fort Garry Hotel
Winnipeg, MB

In cooperation with

THE UNIVERSITY OF BRITISH COLUMBIA
Interprofessional Continuing Education

Welcome

Dear Delegates,

I would like to extend to you a warm welcome to the 14th Annual Scientific Meeting of the Canadian Pediatric Endocrine Group (CPEG). Our past meetings have provided a wonderful opportunity for the Canadian pediatric endocrine community to come together to learn, network and share ideas. The organizing committee has worked hard to craft a program that highlights local Winnipeg work and includes presentations by national and international experts. It also provides a forum for our fellows to present their work. We have an exciting program that we hope will meet the educational needs of our nurses, scientists, endocrinologists and trainees.

I would like to thank our sponsors, who make this meeting possible. I would also like to thank those companies who sponsor our CPEG Fellowship Awards and allow us to train endocrinologists for the future. We look forward to the award announcements at this meeting.

I wish you a stimulating and collegial meeting.

Sincerely,

Bienvenue

Chers délégués,

Je tiens à vous accueillir chaleureusement à la 14ème réunion scientifique annuelle du Groupe canadien d'endocrinologie pédiatrique (GCEP). Nos dernières réunions ont été d'excellentes occasions, pour la communauté canadienne d'endocrinologie pédiatrique, pour se réunir afin d'apprendre, de réseauter et de partager nos idées. Le comité organisateur a travaillé fort pour concevoir un programme qui met en lumière les travaux des gens de Winnipeg ainsi que ceux d'experts nationaux et internationaux. Il fournit également un forum pour que nos « fellows » aient l'occasion de présenter leurs travaux. Nous avons un programme captivant qui, nous l'espérons, répondra aux besoins éducatifs du personnel infirmier, des chercheurs, des endocrinologues et des étudiants du domaine de l'endocrinologie.

Je tiens à remercier nos commanditaires, qui rendent cette rencontre possible. Je tiens aussi à les remercier pour le soutien financier qu'ils offrent à notre programme de bourses CPEG; un programme qui nous permet de former les endocrinologues de demain. Nous attendons d'ailleurs avec impatience l'annonce des récipiendaires de cette année lors de ce congrès.

Je vous souhaite une réunion agréable et stimulante.

Bien cordialement,

Seth Marks, MD, FRCP(C)
Scientific Chair
CPEG 2020 Scientific Meeting

Welcome to Winnipeg

We are delighted to host CPEG 2020 and hope you have a great visit and productive meeting. CPEG is a unique mixture of science and socializing! We look forward to hosting you Friday night at the Manitoba Museum where we have been granted access to the Nonsuch exhibit.

We have a few suggestions below for dinner after the opening reception on Thursday night or for Saturday night if you are staying over.

Thanks for joining us and we hope you have a great time at CPEG 2020. Welcome to the center of Canada!

Kristi, Liz, Brandy, Harpreet, Celia, Katie and Seth

Dining Suggestions – walkable from the Fort Garry (or a short taxi ride)

The Commons – at the Forks – this is a open market style area – common seating in licensed area with many excellent eateries to choose from – casual but fun! $$ *
Passero – more of a fine dining choice at the forks adjacent to the "Common" – small plates $$$
Peasant Cookery –283 Bannatyne Ave -"Farm to Fork" locally source ingredients - $$$
Hermano's Restaurant and Wine Bar – 179 Bannatyne - South American $$$
Deer and Almond – 85 Princess Street – small plates $$$
Kevin's – 141 Bannatyne Ave -comfort food $$
Hy's Steak house – 1 Lombard Place—classic steak house $$$$
BluFish – Asian - $$-$$$ 179 Bannatyne
La Carnita —456 Main Street—Mexican $$-$$$
The Capital Grill – 275 Broadway - casual dining $$-$$$

Dining Suggestions – within 10-15 mins by taxi **

Inferno's Bistro – 312 Des Meurons - French $$-$$$
Gusto Pizzeria – 404 Academy Academy -Italian- $$$
Maque – 909 Dorchester-Asian Fusion $$$
Nonsuch Brewing Co—125 Pacific Ave—Brewery and Shareable plates $$-$$$
Cibo 339 Waterfront Drive—Mediterranean $$$

* this is where the cool people will be Thursday night
**please note UBER is not available in Manitoba

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Financial Contributors

We would like to acknowledge with great appreciation the financial contributions through unrestricted educational grants from the following organizations:

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GOLD

EMD SERONO

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SANDOZ A Novartis Division

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SILVER

ALEXION

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IPSEN Innovation for patient care

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omnipod INSULIN MANAGEMENT SYSTEM

TANDEM DIABETES CARE

Fellowship Listing

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

The CPEG Fellowship Program was and/or is supported by the following:
Eli Lilly, EMD Serono, Hoffmann La Roche, Novo Nordisk, Pfizer, and Sandoz

Program

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

Thursday, February 20, 2020

Time	Session
08:00	CPEN Executive Business Meeting (Room: Salon B)
12:00	CPEG Executive Business Meeting (Room: Salon A)

	Fellows Symposium
13:00	Welcome & Lunch (Gateway)
13:20	What a General Endocrinologist Needs to Know about Bone Health *Marie-Eve Robinson*
14:20	Refreshment Break
14:50	Navigating the Transition to Practice: Thoughts and Perspectives *Harpreet Gill*
15:50	Conclusion

16:00	CPEG 2020 Registration Opens (Crystal Ballroom Foyer)
17:00	Welcome Reception & Exhibits (Room: Crystal Ballroom)
19:00	Adjourn

Friday, February 21, 2020

Time	Session
07:00	Registration (Room: Crystal Ballroom Foyer) Breakfast & Exhibits (Room: Crystal Ballroom)
08:00	Opening Remarks: The Land Acknowledgement & Welcome (Room: Concert Hall) *Seth Marks, Kristi Bell, Brandy Wicklow* Friday Poster Highlights *Each odd-numbered poster presenter will give a 1-minute & 1-slide presentation.*
08:30	THEME I: Endocrine Stimulation Testing (Room: Concert Hall) *Moderator: Wendy Schwarz* Growth Hormone Stimulation Testing: A Standard of Care That Is Anything but Standard *Adda Grimberg*
09:30	GnRH Stimulation Tests in the Diagnosis and Management of Precious Puberty: One Stimulus Too Many? *Jennifer Harrington*
10:00	Break and Exhibits (Room: Crystal Ballroom)
10:30	THEME II: Puberty (Room: Concert Hall) *Moderator: Colleen Nugent* Constitutional Delay of Growth and Puberty and Isolated Hypogonadotropic Hypogonadism: Two Sides of the Same Coin? Can We Tell Heads from Tails? *Mark Palmert*
11:15	Estrogen Replacement in Girls with Turner Syndrome *Laura Stewart*

12:00 Poster Viewing & Highlighted Poster Walks (x3) (Room: LaVerendyre)
Moderator: Brandy Wicklow

Highlighted Poster #1:
The Impact of Hospital Surgical Volume on Healthcare Utilization and Surgical Outcomes for Pediatric Thyroidectomy
Jonathan Wasserman

Highlighted Poster #3:
A Simulation-based Intervention Teaching Illness Management Skills to Caregivers of Children with Adrenal Insufficiency: a Randomized Controlled Study
Heidi Virtanen

Highlighted Poster #5:
Did Pamidronate Cause Pulmonary Edema in a Neonate treated for Hypercalcemia Secondary to Subcutaneous Fat Necrosis?
Rosalie Cavin

12:30 Lunch & Exhibits (Room: Crystal Ballroom)

13:30 THEME III: Social Determinants of Health & Indigenous Health (Room: Concert Hall)
Moderator: Denis Daneman

Rights-Based Reconciliation: A Pre-Requisite for Child Health
Marcia Anderson

14:15 Renewing Relationships with Indigenous Families: Our Path Towards Reconciliation at the Children's Hospital
Melanie Morris

15:00 Break & Exhibits (Room: Crystal Ballroom) & Poster Viewing (Room: LaVerendyre)
Nurses split, see page 11

15:30 Oral Abstract Presentations (6) (Room: Concert Hall) – see page 12
Moderator: Rebecca Perry & Annie Sbrocchi

17:00 Adjourn

FRIDAY NIGHT EVENT

Dinner and Entertainment at **The Manitoba Museum** featuring the **Nonsuch Gallery** 190 Rupert Ave, Winnipeg, MB R3B 0N3: 1.7km walk (transportation will be provided). If you have purchased a ticket to this event during the registration process, please refer to your blue folders as the ticket will be placed there with further instructions. If you have not pre-purchased for this event and would like to come, please see Jo at the registration desk.

18:00 – 18:45	Cocktail Reception
18:45 – 19:30	Dinner
19:30	Legacy – The Bunkowsky Family Band & other entertainment

Saturday, February 22, 2020

Time	Session
07:30	Breakfast (Room: Crystal Ballroom)
08:00	Business Meeting (Room: Concert Hall)
09:55	Fellowship Awards *Presented by Dr. Carol Huang*
10:00	Break and Exhibits (Room: Crystal Ballroom)
	Nurses split, see page 11
10:30	THEME IV: Diabetes (Room: Concert Hall) *Moderator: Mark Inman* Diabetes and Technology – Help or Hindrance? *Lori Berard*
11:15	Post-Transplant Diabetes in Children After Organ Transplant and Approaches to Mitigate Risk *Tom D. Blydt-Hansen, Shazhan Amed*
12:00	Saturday Poster Highlights Each even-numbered poster presenter will give a 1-minute & 1-slide presentation.
12:15	Poster Viewing & Highlighted Poster Walks (x3) (Room: LaVerendyre) *Moderator: Caroline Zuijdwijk* Highlighted Poster #2: Safety and Efficacy of Subcutaneous Insulin for Treatment of Diabetic Ketoacidosis in Children – A Systematic Review *Noemie Pothier* Highlighted Poster #4: Let's Go to Camp: Type 1 Diabetes Innovations - From the Lab to the Field *Emilie Palisaitis* Highlighted Poster #6: Hypoglycemia with Acute Lymphoblastic Leukemia Treatment in Pediatric Patients: A Novel Case Series *Mary Jiang*

12:45	Lunch & Exhibits (Room: Crystal Ballroom)
	Nurses split, see page 11
13:30	Oral Abstract Presentations (6) (Room: Concert Hall)– see page 12 *Moderator: Paola Luca & Stasia Hadjiyaanakis*
15:00	Break & Exhibits (Room: Concert Hall) & Poster Viewing (Room: LaVerendyre)
15:30	John Bailey Award (Room: Concert Hall) *Presented by Shazhan Amed*
15:35	Debate: Growth Hormone Therapy in Russell - Silver Syndrome (Room: Concert Hall) *Moderator: Liz Sellers* *PRO: Rose Girgis* *CON: Danièle Pacaud*
16:35	Closing Remarks & Evaluation (Room: Concert Hall)
16:45	Adjourn

*Nursing Program for Friday, February 21 & Saturday, February 22 (Gateway)

Moderators: Ms. Kristi Bell & Ms. Alice Boland

*Nursing Program for Friday, February 21

15:30	CPEN AGM
17:00	Break & Exhibits (Room: Crystal Ballroom)
17:30	Re-join CPEG group

*Nursing Program for Saturday, February 22

10:30	Jordan's Principle - Improving Patient Outcomes for Children in Manitoba First Nations Communities *Nicole Kirouac*
11:15	CAPTH 2019 Update *Peggy Kalancha*
11:35	Learning from ESPE 2019 Vienna *Rebecca Brooke*
12:00	Lunch & Exhibits (Room: Crystal Ballroom)
13:30	Current Considerations for Pediatric DSD Surgery *Darius Bägli*
14:15	"Life-Cycle" Healthcare Transitions with Canadians Experiencing Differences in Sex Development: What Do We Know and How Do We Move Forward? *Caroline Sanders*
15:00	Break & Exhibits (Room: Crystal Ballroom)

Fellow (Oral) Abstract Schedule

Time	Title	Presenter	Abstract #	Page
Friday, February 21 *Moderators: Rebecca Perry & Annie Sbrocchi*				
15:30	Reducing Unnecessary Thyroid Function Testing: A Quality Improvement Initiative	Christine Tenedero	1	27
15:45	Comparison of 1- and 3-Month Lupron Depot®(LD) for Pubertal Suppression in Transgender Youth	Zoyah Thawer	2	28
16:00	Infantile Hypoinsulinemic Hypoketotic Hypoglycemia: An AKT2 Gene Mutation	Sarah Ames	3	29
16:15	Transient Hyperinsulinism: Off to a Bad Start?	Ravit Regev	4	30
16:30	How Does Access and Cost of Gender-Affirming Surgery Differ for Adolescents and Adults Across Canada?	Zachary Zytner	5	31
16:45	Predicting Gestational Age Improves Newborn Screening for Congenital Adrenal Hyperplasia	Danny Jomaa	6	32

Saturday, February 22 *Moderators: Paola Luca & Stasia Hadjiyaanakis*				
13:30	An Incidentally-Discovered Paraganglioma	Marian Thorpe	7	33
13:45	Socioeconomic Disparities in PUMP Uptake Among Canadian Children and Adolescents With Type 1 Diabetes: Comparison of Two Provincial Programs	Jennifer Ladd	8	34
14:00	Influence of Pre-Diabetes on Respiratory and Nutritional Status in the Montreal Cystic Fibrosis Cohort	Kate Potter	9	35
14:15	Cyclical Cushing Syndrome in a Patient with Carney Complex: a Rare Association and a Management Challenge	Carolina Silva	10	36
14:30	Perspectives on a Type 1 Diabetes Pediatric-to-Adult Transition Program in Vancouver, Canada	Catherine Lim	11	37
14:45	Frequency of Contacts with the Diabetes Team in Pediatric Patients with Type 1 Diabetes on an Insulin Pump and Glycemic Control	Funmbi Babalola	12	38

Poster Abstract Listing

All odd-numbered posters can be viewed on Friday, February 21 from 1200-1230 & 1500-1530. Posters 1, 3, 5 will be highlighted at 1200.

All even-numbered posters can be viewed on Saturday, February 22 from 1215-1245 & 1500-1530. Posters 2, 4, 6 will be highlighted at 1215.

Title	Presenter	Abstract #	Page
The Impact of Hospital Surgical Volume on Healthcare Utilization and Surgical Outcomes for Pediatric Thyroidectomy	Jonathan Wasserman	1	39
Safety and Efficacy of Subcutaneous Insulin for Treatment of Diabetic Ketoacidosis in Children – A Systematic Review	Noemie Pothier	2	40
A Simulation-based Intervention Teaching Illness Management Skills to Caregivers of Children with Adrenal Insufficiency: a Randomized Controlled Study	Heidi Virtanen	3	41
Let's Go to Camp: Type 1 Diabetes Innovations - From the Lab to the Field	Emilie Palisaitis	4	42
Did Pamidronate Cause Pulmonary Edema in a Neonate treated for Hypercalcemia Secondary to Subcutaneous Fat Necrosis?	Rosalie Cavin	5	43
Hypoglycemia with Acute Lymphoblastic Leukemia Treatment in Pediatric Patients: A Novel Case Series	Mary Jiang	6	44
Combined Indeterminate and Impaired Glucose Tolerance is a High Risk for Cystic Fibrosis-Related Diabetes in the Montreal Cystic Fibrosis Cohort	Kate Potter	7	46
A New Twist on a Rare Form of Rickets	Zachary Zytner	8	47
~~Psychosocial and Socio-Economic Well-Being as Determinants of Quality of Life and Glycemic Control in Haitian Youth With Diabetes~~	~~Rosemary Vincent~~	~~9~~	~~48~~
Strange Behaviour: A Case Report of an Adolescent With an Insulinoma	Carly Baxter	10	49
Idiopathic Infantile Hypercalcemia - Clinical Spectrum of Milder Phenotypes	Nina Lenherr-Taube	11	50
Improving Sick Day Management among Patients and Families with Type 1 Diabetes	Hannah Geddie	12	52
Successful Management of T1D in an Infant with Insulin Pump Therapy and Diluted Insulin	Emma McCutcheon	13	53
Rickets Complicating Pseudohypoaldosteronism Type 1	Ravit Regev	14	54
Awareness of Primary Care Physician That Lavender Oil Causes Prepubertal Gynecomastia or Premature Thelarche	Rana Yassa	15	55
Vitamin D Deficiency Is Associated With Higher Systolic Blood Pressure in Adolescents with Type 1 Diabetes Compared to Healthy Controls	Nina Lenherr-Taube	16	56
Growth Hormone Deficiency in Patient with Megalencephaly Capillary Malformation Syndrome (MCAP)	Reem Alfattouh	17	57
Can Rickets Be Transient Without Treatment?	Patricia Diaz Escagedo	18	58
Reversal of Precocious Puberty with Letrozole in McCune-Albright Syndrome	Samantha Gerber	19	59
A Curious Case of Hypertension	Maria-Elena Lautatzis	20	60
The Development of Standard Operating Procedures (Sops) for the Use of the Edmonton Obesity Staging System for Pediatrics (Eoss-P) in Community Practice in the Management of Patients With Obesity.	Matthew Feldman	21	61

Large-Scale Genome-Wide Association Study for Vitamin D Levels and Their Effect on Risk of Type 1 Diabetes and Autism Spectrum Disorders	Despoina Manousaki	22	62
Lessons learned regarding "transient" hyperinsulinism: A case of diazoxide responsive hyperinsulinism secondary to ABCC8 mutation	Noor Sawalha	23	64
MYRF Variant as a Cause of 46,XY Disorder of Sexual Development	Nour Gazzaz	24	65
Case Report of a 14 Year Old Female with Primary Burkitt Lymphoma of the Thyroid Gland	Alexa Marr	25	66
A Three-Year Review of the Night Monitor Protocol: A Camp Banting Quality Improvement Initiative	Alexa Marr	26	67
Challenges in the Treatment of GACI Patients: Promising Future Therapeutic Directions	Kim Phung	27	68
The Conundrum with Early Diagnosis of 46, XY Disorder of Sexual Development	Trisha Patel	28	69

Program Organizing and Scientific Committee

Shazhan Amed
Kristi Bell
Alice Boland
Farid Mahmud
Seth Marks
Susan Murphy
Jo Nam
Munier Nour
Colleen Nugent
Elizabeth Sellers
Bailie Tabak
Zoyah Thawer
Brandy Wicklow
Caroline Zuijdwijk

Credits

This event has been approved by the Canadian Paediatric Society for a **maximum of 11.25 credit hours** as 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. The specific opinions and content of this event are not necessarily those of the CPS, and are the responsibility of the organizer(s) alone.

Dr. John Bailey Resident Research Award

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

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

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

CPEG Distinguished Service Award

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

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

- 2017 – Daniel Metzger
- 2019 - Denis Daneman

Learning Objectives

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

Session Learning Objectives:

Fellows Symposium

What a General Endocrinologist Needs to Know about Bone Health

Marie-Eve Robinson, *M.D., C.M., MSc., FAAP, FRCPC, Pediatric Endocrinologist, Children's Hospital of Eastern Ontario; Assistant Professor of Pediatrics, University of Ottawa; Clinician Researcher in Pediatric Bone Health, CHEO Research Institute, Ottawa, ON*

1. Develop an approach to the screening, diagnosis and treatment of osteoporosis in children
2. Describe a new classification for the differential diagnosis of rickets and how it impacts treatment
3. Discuss an approach to screen and treat osteopenia of prematurity

Navigating the Transition to Practice: Thoughts and Perspectives

Harpreet Gill, *MD, FRCPC, Pediatric Endocrinologist, Health Sciences Center – Children's Hospital; Assistant Professor, Department of Pediatrics, the University of Manitoba, Winnipeg, MB*

1. Discuss important considerations during fellowship training to optimize future career opportunities
2. Analyze key factors that may impact your practice choices after evaluating the available opportunities
3. Increase preparedness for transition to practice by reviewing common themes in advice shared by recent graduates

Theme I: Endocrine Stimulation Testing

Growth Hormone Stimulation Testing: A Standard of Care That Is Anything but Standard

Adda Grimberg, *MD, Scientific Director, Diagnostic and Research Growth Center, Children's Hospital of Philadelphia; Professor, Dept. of Pediatrics, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA*

1. Describe the rationale behind provocative growth hormone testing in the diagnosis of growth hormone deficiency
2. List the limitations of provocative growth hormone testing in the diagnosis of growth hormone deficiency
3. Endorse the importance of harmonized assays for measuring growth hormone and insulin-like growth factor-I

GnRH Stimulation Tests in the Diagnosis and Management of Precious Puberty: One Stimulus Too Many?

Jennifer Harrington, *Training Program Director, Division of Endocrinology, Hospital for Sick Children; Assistant Professor, Department of Paediatrics, University of Toronto, Toronto, ON*

1. Discuss the role of GnRH/GnRH agonist stimulation tests in the diagnosis and ongoing management of children with central precocious puberty (CPP)

2. Compare the utility of other investigations in CPP, including basal gonadotropins
3. Describe an approach to the diagnosis and management of a child presenting with CPP

Theme II: Puberty

Constitutional Delay of Growth and Puberty and Isolated Hypogonadotropic Hypogonadism: Two Sides of the Same Coin? Can We Tell Heads from Tails?

Mark Palmert, MD, PhD, Associate Chair of Pediatrics (Ambulatory Care), Hospital for Sick Children; Professor of Pediatrics and Physiology, University of Toronto, Toronto, ON

1. Discuss the pathophysiology of Constitutional Delay of Growth and Puberty (CDGP) and Isolated Hypogonadotropic Hypogonadism (IHH), with focus on areas of overlap
2. Recognize clinical features that may allow for discrimination of one condition from the other
3. Discuss utility of diagnostic laboratory tests

Estrogen Replacement in Girls with Turner Syndrome

Laura Stewart, MD, Clinical Associate Professor, Program Director, Pediatric Endocrinology, BC Children's Hospital, Vancouver, BC

1. Review the normal changes in estrogen production during infancy, childhood, & puberty
2. Review the literature with respect to pubertal induction in Turner Syndrome
3. Review pubertal hormone replacement options for girls with Turner Syndrome

THEME III: Social Determinants of Health & Indigenous Health

Rights-Based Reconciliation: A Pre-Requisite for Child Health

Marcia Anderson, MPH, MD, BSc, Executive Director, Indigenous Academic Affairs in the Ongomiizwin Indigenous Institute of Health and Healing, Rady Faculty of Health Sciences, University of Manitoba Winnipeg, MB

1. Compare and contrast a rights-based approach to reconciliation and a benevolent approach to reconciliation
2. Describe multi-level racism and its impact on Indigenous children's health
3. Identify and commit to individual and collective action that will contribute to closing the gaps in Indigenous children's health

Renewing Relationships with Indigenous Families: Our Path Towards Reconciliation at the Children's Hospital

Melanie Morris, PhD, Assistant Professor, Section of Pediatric General Surgery, University of Manitoba; Indigenous Lead for Children's Hospital, Winnipeg, MB; Medical Director & Assistant Professor, Department of Surgery, Global Surgery Office, Vancouver, BC

1. Describe some of the historical reasons influencing the health status indicators, particularly those relating to Indigenous children
2. Discuss the importance of engaging Indigenous communities in developing initiatives for the children's hospital journey toward reconciliation
3. Recognize the complexities of Social Determinants of Health and discuss a working framework for including the various determinants of Indigenous pediatric health

Theme IV: Diabetes

Diabetes and Technology – Help or Hinderance?

Lori Berard, Nurse Consultant, Diabetes Management and Clinical Trial Operations, Winnipeg, MB

1. Summarize recent advances in technology design to improve diabetes management
2. Reflect on the patient perception of technology in their diabetes self-management
3. Facilitate understanding of the role of the clinician in promoting self-management skills through technology
4. Discuss practicalities of technology burnout – for the clinician and the client

Post-Transplant Diabetes in Children After Organ Transplant and Approaches to Mitigate Risk

Tom D. Blydt-Hansen, MDCM, FRCPC, Associate Professor, Department of Pediatrics, University of British Columbia; Director, Multi-Organ Transplant Program, BC Children's Hospital, Vancouver, BC

Shazhan Amed, MD, Clinical Associate Professor, Division of Endocrinology & Diabetes, Department of Pediatrics, Faculty of Medicine, University of British Columbia, Vancouver, BC

1. Identify children at risk for post-transplant diabetes based on their pre- and peri-operative clinical findings
2. Employ a management approach to mitigate the long-term risk for diabetes

Debate:

Growth Hormone Therapy in Russell - Silver Syndrome

PRO: Rose Girgis, MBBCh, MSc, FRCPC, Associate Professor, Pediatric Endocrinology, Divisional Director, Director, Clinical Fellowships, Postgraduate Medical Education, Faculty of Medicine & Dentistry, University of Alberta, Edmonton, AB

CON: *Danièle Pacaud, MD, FRCPC, Professor, Pediatrics, University of Calgary; Pediatric Endocrinologist, Alberta Children's Hospital, Calgary, AB*

1. Review the diagnosis of Silver-Russell syndrome
2. Review to benefits of growth hormone therapy in children with Silver-Russell syndrome
3. Review risk of growth hormone therapy in children with Silver-Russell syndrome

Nursing Session:

Jordan's Principle - Improving Patient Outcomes for Children in Manitoba First Nations Communities

Nicole Kirouac, RN, BN, Community Nurse Consultant, St. Amant, Winnipeg, MB

1. Gain an understanding of the history and scope of Jordan's Principle across Canada
2. Know how and when to access Jordan's Principle services for children and families you support

CAPTH 2019 Update

Peggy Kalancha, RN, BSN, Clinical Resource Nurse, Pediatric Endocrine, Gynecology and Metta Gender Services clinics, Alberta Children's Hospital, Calgary, AB

1. Update by presenting some of the academic learning presented at CPATH
2. Review changes to best practise in gender affirming care
3. Contrast the provision of gender affirming care to children and youth, with the concept of a gender affirming world

Learning from ESPE 2019 Vienna

Rebecca Brooke RN BSN, Endocrine Clinic Nurse, BC Children's Hospital, Vancouver, BC

1. Puberty Induction in Girls with Turner Syndrome: A Swedish Approach
2. Not to be 'Googled" at Work: Tips from the British Nurses

Current Considerations for Pediatric DSD Surgery

Darius Bägli, MDCM, Hospital For Sick Children research institute, University of Toronto; Department of surgery and physiology, Institute for medical sciences, Toronto, ON

1. Recognize the spectrum of disorders of sexual development
2. Recognize surgical approaches to disorders of sexual development
3. Appreciate controversies and timing of surgery decisions for disorders of sexual development

"Life-Cycle" Healthcare Transitions with Canadians Experiencing Differences in Sex Development: What Do We Know and How Do We Move Forward?

Caroline Sanders, PhD, MBE, RN, Associate Professor, School of Nursing, University of Northern British Columbia, Prince George, BC

1. Identify current transition hurdles for individuals with DSD, applying this knowledge to your local contexts
2. Discuss the variation between 'transition times' comparing adolescent-young adult to older-adult experiences
3. Apply health promotion and prevention strategies, describing successful approaches focused on empowerment and self-care
4. Recognize opportunities to explore service framework planning both locally and across Canada

Biographies

Shazhan Amed

Dr. Amed is a pediatric endocrinologist at BC Children's Hospital, a Clinical Associate Professor at UBC, an Associate Clinician Scientist at the BC Children's Hospital Research Institute, and Head of the Division of Endocrinology at BC Children's Hospital. Dr. Amed is a health services and population health researcher. Her research is focused on the prevention of childhood obesity and youth-onset type 2 diabetes as well as population-level surveillance of childhood type 1 and type 2 diabetes and the design and evaluation of quality improvement initiatives related to health service delivery for children and youth with diabetes.

Marcia Anderson

Dr. Marcia Anderson is Cree-Anishinaabe and grew up in the North End of Winnipeg. Her family roots go to the Norway House Cree Nation and Peguis First Nation in Manitoba. She practices both Internal Medicine and Public Health as a Medical Officer of Health with the Winnipeg Regional Health Authority. She is the Executive Director of Indigenous Academic Affairs in the Ongomiizwin Indigenous Institute of Health and Healing, Rady Faculty of Health Sciences, University of Manitoba. She serves as the Chair of the Indigenous Health Network of the Association of Faculties of Medicine of Canada. She is a Past President of the Indigenous Physicians Association of Canada and Past Chair of the Pacific Region Indigenous Doctors Congress. She was recognized for her contributions to Indigenous peoples health with a National Aboriginal Achievement Award in March 2011. In 2018 she was named one of the 100 most powerful women in Canada by the Women's Executive Network.

Darius Bägli

Dr. Bägli is a world expert in interdisciplinary pediatric urology and translational science. He has been an academic member of the division or urology at the University of Toronto and Sickkids Hospital since 1995 and a Full Professor in the departments of Surgery & Physiology since 2012.

He is certified in Urologic Surgery by the Royal College of Physicians and Surgeons of Canada. As an experienced hypospadiologist, Dr. Bagli is at the surgical epicenter of the debates regarding timing of genital surgery in children, and the controversies surrounding surgical indications and outcomes in this field. He is one of the few paediatric urologists with an independent cell and molecular biology program in North America. His interests include biology, technology, innovation, bioethics, and clinically in hypospadias and genital reconstruction. Dr. Bagli has published works in urological molecular biology, authored numerous papers on hypospadias addressing surgery, outcomes, indications, management, and the physics of flow dynamics.

Lori Berard

Lori Berard is Diabetes Educator with an expertise in diabetes education, management and clinical research. She has over 30 years' experience primarily as the Nurse Manager for the Health Sciences Centre Diabetes Research Program and a Faculty Member at the University of Manitoba Department of Medicine Section of Endocrinology. Currently she is working as a consultant in diabetes management and clinical research operations. Lori has been a professional member and major volunteer of Diabetes Canada for more than 25 years and has extensive experience with the Clinical Practice Guidelines. She has received many honors and awards related to her work in diabetes.

Tom Blydt-Hansen

Dr. Tom Blydt-Hansen completed his MD at McGill University, and his Pediatric and Nephrology specialization at the Montreal Children's Hospital, followed by further specialization in transplantation at the University of California, Los Angeles. He began his Nephrology career at the University of Manitoba in 2001, where he was Division Head of Nephrology from 2005-2014. He then moved to BC Children's Hospital in 2014, where he is Director of the Multi Organ Transplant Program and Senior Scientist at the BCCH Research Institute. He is a past-President of the Canadian Society of Transplantation, and is a Board member of the NAPRTCS Registry. His research program is focused on identification and development of relevant non-invasive urine biomarkers of allograft injury in pediatric kidney transplant recipients using metabolomic and proteomic methods. He is also engaged in collaborations to identify urinary biomarkers (metabolomics) associated with chronic kidney disease, acute kidney injury, cisplatin toxicity, type 2 diabetes and cisplatin nephrotoxicity; and is co-investigator on several nationally funded transplant research studies including VIRTUUS, TAKE-IT TOO, CKiD, iCARE, CAN-RESTORE and CNTRP.

Rebecca Brooke

Rebecca is an Endocrine Clinic Nurse at BC Children's Hospital in Vancouver BC. Rebecca has worked with the Endocrine and Gender Team at BCCH since February 2016. Prior to taking on this role, she worked in Diabetes Care. She started her diabetes career after being invited to Diabetes Camp in the summer of 2003. Following this she was head nurse at diabetes camp on the Sunshine Coast for 10 years. She left her Adult Acute Medicine job to do contract work with clients and families using Insulin Pumps and Continuous Glucose Monitoring. In 2011, she was part of a small, tight knit team who started up a Community Pediatric Diabetes Clinic in Surrey BC. Currently she is happy to serve her CPEN colleagues as Treasurer. She is also a member of CPEG and PENS.

Harpreet Gill

Dr. Harpreet Gill is a pediatric endocrinologist at the Children's Hospital of Winnipeg and an assistant professor at the University of Manitoba. She completed her undergraduate degree at McMaster University and medical school at the University of Western Ontario. After completing her General Pediatric residency at McMaster Children's Hospital, she pursued her Endocrinology fellowship at the Children's Hospital of Eastern Ontario. Dr. Gill is currently pursuing a distance-based Master's degree in Quality Improvement and Patient Safety at the Johns Hopkins Bloomberg School of Public Health. In addition to clinical and research pursuits, Dr. Gill is passionate about teaching and the delivery of healthcare in resource-poor settings.

Rose Girgis

Dr. Rose Girgis is an Associate Professor of Pediatrics at the University of Alberta, Edmonton, Alberta, Canada. Dr. Girgis first came to Canada from Egypt in 1987, an MD credential in hand; she completed a Master of Sciences in Biochemical Genetics in the Department of Pediatrics, University of Alberta. Then she went on to do residency training in General Pediatrics and Endocrinology. She completed a research fellowship in "The Immuno-pathogenesis of Type 1 Diabetes" at the University of Alberta and a clinical fellowship year at Baylor College of Medicine, Houston Texas.

In 1998 she joined Dr. Bob Couch on staff at the University of Alberta. She was the Pediatric Endocrinology Residency Program Director from 2003 – 2012. She is currently the Divisional Director of Pediatric Endocrinology at the University of Alberta. She is involved in Medical Education and in 2016 was appointed the Director of Fellowships, Postgraduate Medical Education, Faculty of Medicine & Dentistry, University of Alberta. She is proud to be one of the founding members of CPEG since its

inception. In the late nineties early 2000s, Dr. Girgis participated in the monumental Canadian randomized controlled trial looking at the impact of growth hormone supplementation on adult height in Turner syndrome. Over the years, she has also been involved in multiple growth hormone clinical trials.

Adda Grimberg

Adda Grimberg, MD, is professor of pediatrics at the Perelman School of Medicine and senior fellow of the Leonard Davis Institute of Health Economics, both at the University of Pennsylvania, and Scientific Director of the Diagnostic and Research Growth Center at the Children's Hospital of Philadelphia.

Jennifer Harrington

Dr. Jenny Harrington received her medical degree at the University of Adelaide and undertook her pediatric residency, endocrine fellowship training and PhD through the Women's and Children's Hospital in Adelaide, Australia. Following a clinical and research pediatric endocrine fellowship at SickKids she joined the Endocrine faculty in December of 2015. Jenny's clinical and research interests include the management of children with disorders of calcium homeostasis and bone metabolism. In addition she has co-authored several papers and chapters on the diagnostic utility of tests in children with disorders of pubertal timing.

Peggy Kalancha

Peggy is a Clinical Resource Nurse in the Pediatric Endocrine, Gynecology and Metta Gender Services clinics at the Alberta Children's Hospital. Peggy has worked in this area for approx. 12 years but has also worked in a number of other areas in Pediatrics subspecialties. These include Neonatal Follow-up, Developmental, Genetics, Rheumatology, Neuromuscular, Neurology, and a few others thrown in! At this time she has taken on a partial leadership role in the development of a Gender Program at ACH. Peggy is a member of CPEG, CPEN, CPATH and NASPAG.

Nicole Kirouac

Nicole is a graduate from the University of Manitoba Nursing. She was the Pediatric Endocrine Nurse at the Children's Hospital of Winnipeg for 19 years. Nicole was the founding leader of this CPEN Group and also held the seat of president with PENS.
In 2017 Nicole moved over to St. Amant Inc. to work as a Nurse Consultant. She is present in First Nation Reserves for assessments, advocacy and education to support children with unique medical needs. This Jordan's Principle – Child First Initiative Program is funded by FNIHB.

Melanie Morris

Dr. Melanie Morris is an indigenous pediatric surgeon and urologist at the Children's Hospital in Winnipeg. Melanie has recently been appointed the Inaugural Lead on Indigenous Health at the Children's Hospital in Winnipeg. Dr. Morris founded with the Department of Surgery the Winnipeg Global Surgery Office and currently serves as the medical director. Melanie has supported teaching and training in the area of Pediatric Surgery in Kenya and Uganda and remains an active board member of The College of Surgeons of East, Central and Southern Africa. Melanie is an instructor for the University of British Columbia, College of Medicine's Masters in Global Surgery program on Indigenous Health. . Dr. Morris has been working with Ongomiizwin to create outreach clinics with pediatric surgery in Nunavut. Her most recent interest is in developing culturally safe spaces for Indigenous children receiving care at the Winnipeg's Children's Hospital For this work, Dr. Morris is engaging Indigenous parents, children and Elders in priority-setting, designing and evaluating all phases of the transformation of the Children's Hospital.

Danièle Pacaud

Dr. Danièle Pacaud, MD, FRCPC is a Professor of Pediatrics at the University of Calgary and Pediatric Endocrinologist working at Alberta Children's Hospital Diabetes and Endocrine Clinics for more than 20 years. She trained in Montreal at the Hôpital Ste-Justine, Université de Montréal and at McGill University. She is a founding member of CPEG and has served as an executive member in various role including as president years. She has ongoing involvement in clinical research in pediatric endocrinology with a focus in pediatric diabetes.

Mark Palmert

Mark Palmert is a Professor of Pediatrics and Physiology at the University of Toronto. Mark graduated from the Medical Scientist Training Program at Case Western Reserve University, Cleveland, Ohio, with a MD and PhD in 1992 and then completed his pediatrics and pediatric endocrinology training at Children's Hospital, Boston. After training, Mark held faculty appointments in Boston and at Rainbow Babies and Children's Hospital in Cleveland before he moved to Toronto in 2007 to become Head of the Division of Endocrinology at SickKids. In 2017 Mark began a new role at SickKids as Associate Chair of Pediatrics (Ambulatory Care). Mark has a long-standing clinical and research interest in the regulation and disorders of pubertal timing. He has conducted clinical studies of precocious and delayed puberty and has directed a laboratory-based program designed to identify and understand genetic factors that regulate the onset of puberty.

Marie-Eve Robinson

Dr. Robinson is a pediatric endocrinologist specialized in pediatric bone health and assistant professor at the University of Ottawa. She completed her pediatric endocrinology training at the Montreal Children's Hospital, McGill University. She then completed a two-year pediatric bone fellowship, during which she spent one year at the Shriners Hospital for Children's – Canada and one year at the Children's Hospital of Eastern Ontario, University of Ottawa. Dr. Robinson has a master's degree in epidemiology and biostatistics from McGill University. She is a clinician researcher affiliated with the Children's Hospital of Eastern Ontario Research Institute, her main research interest being in genetic and metabolic pediatric bone diseases.

Caroline Sanders

Dr. Sanders, MBE is an Associate Professor and Coordinator of Graduate Programs in the School of Nursing at UNBC. Caroline moved to UNBC from the UK in 2016 where she undertook a postdoctoral National Institute Health Research (NIHR) fellowship. Research has focused on qualitative and mixed method approaches specifically, phenomenology, narrative enquiry, and participatory action research often framed within patient-oriented research. Clinical-academic research methods focus on rare disease to include differences in sex development specifically focusing on the development and translation of knowledge across healthcare and community boundaries.

Laura Stewart

Laura Stewart a pediatric endocrinologist working at BC Children's Hospital. She was involved in the Canadian Growth Hormone Trial for girls with Turner Syndrome. She is the Program Director for the Pediatric Endocrinology and Metabolism Fellowship Program at the University of British Columbia.

Conflict of Interest Disclosures

All speakers and committee members must disclose whether they do or do not have a relationship with a commercial entity such as a pharmaceutical organization, medical device company or a communications firm.

Committee Members

Shazhan Amed
- I am a member of an Advisory Board with Lilly Insulet Dexcom.
- I have received payment from Lilly Insulet Dexcom for attending advisory board meetings.
- I have received a grant or an honorarium from Lilly for a transition study

Kristi Bell
- No affiliation

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- Not disclosed

Carol Huang
- No affiliation

Farid Mahmud
- I am a member of an Advisory Board with Lilly, Insulet, Novo Nordisk.
- I am currently participating in or have participated in a clinical trial within the past two years with AdDIP/Attempt.

Seth Marks
- I am currently participating in or have participated in a clinical trial within the past two years for Boehringer Ingelheim as a co-investigator in DINAMO Study

Susan Murphy
- I was a member of the Nursing Advisory Board with EMD Serono and Sandoz in 2019.
- I am currently participating in or have participated in a clinical trial within the past two years for Levo as an Interviewer/Rater for the LV-101-3-01 CARE PWS Study

Jo Nam
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- I was a member of an Advisory Board with Lilly in May 2017
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Colleen Nugent
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- I am currently participating in or have participated in a clinical trial within the past two years for Boehringer Ingelheim as a co-investigator in DINAMO Study

Ballie Tabak
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Zoyah Thawer
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Brandy Wicklow
- I am a site co-investigator for Boehringer Ingleheim DINAMO study.

Caroline Zuijdwijk
- I was a one time member of the advisory board for Eli Lilly Canada (honorarium directed to Diabetes Educator fund).
- I am currently participating in a clinical trial for Medtronic - CHEO co-investigator for multi-centre RCT re: home use of hybrid closed loop system. No monetary gain.

Speakers

Marcia Anderson
- No affiliation

Darius Bägli
- I am a member of an advisory board with Advances in Urology as an associate editor

Lori Berard
- No affiliation

Tom Blydt-Hansen
- I have received payment from Astellas Canada for speaker fee at a national conference.
- I have received grant from Astellas Canada for investigator initiated grant.
- I am currently participating in a clinical trial for Astellas Canada to investigate once-daily dosing of immunosuppressant drugs

Harpreet Gill
- No affiliation

Rose Girgis
- I am currently participating in a clinical trial for Ascendis for long acting Growth Hormone Clinical Trial

Adda Grimberg
- I am a member of an advisory board with Pfizer & Sandoz

Jennifer Harrington
- No affiliation

Peggy Kalancha
- I have attended advisory board with Serono, Lilly, Sandoz, Pfizer in the past

Nicole Kirouac
- No affiliation

Melanie Morris
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Danièle Pacaud
- I have received a grant from Eli Lilly Canada
- I am currently participating in a clinical trial for Versatis Inc., Novo Nordisk/Quintiles Canada Inc./Tolmar Inc./Sandoz Canada Inc.
- I intend to make therapeutic recommendations for medications or medical devices that have not received regulatory approval.

Mark Palmert
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Marie-Eve Robinson
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Caroline Sanders
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Laura Stewart
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Oral Abstracts

Oral Abstract 1
Reducing Unnecessary Thyroid Function Testing: A Quality Improvement Initiative
Are intra-articular injections safe?

Christine Tenedero (1,2), Leah Abitbol (1,2), Lusia Sepiashvili (3), Benjamin Jung (3), Jonathan D. Wasserman (1,2), Mark R. Palmert (1,2)
(1) Division of Endocrinology, The Hospital for Sick Children, Toronto, ON. (2) Department of Pediatrics, University of Toronto, The Hospital for Sick Children, Toronto, ON. (3) Department of Clinical Biochemistry, The Hospital for Sick Children, Toronto, ON.

BACKGROUND: The American Thyroid Association and Choosing Wisely Canada recommend measuring exclusively thyroid stimulating hormone (TSH) to screen for and monitor treatment of primary hypothyroidism. Health care providers frequently order free thyroxine (fT4) and triiodothyroxinine (T3) measurements when not clinically indicated. These unnecessary tests can lead to excessive use of health- care resources and compromise quality of care, by increasing parental and patient anxiety, misinterpretation of results and unnecessary referrals and interventions.
This quality improvement initiative aimed to promote resource stewardship and decrease unnecessary thyroid hormone (fT4, T3) testing at our institution.

METHODS: Quality improvement strategies were used to generate three change ideas that were implemented simultaneously: (1) the introduction of a laboratory "reflex fT4" system (whereby fT4 is automatically measured and reported if the TSH is outside the reference range), (2) a "forced function" for thyroid hormone orders within the hospital's electronic medical record (EMR) (compelling the user to indicate rationale for ordering fT4/T3) and (3) built-in educational messaging provided at the time of ordering thyroid hormone tests, stating the recommendations regarding use of TSH. Laboratory data were audited to determine the mean number of TSH, fT4, and T3 tests performed per week as well as indications for testing. The main outcomes were change in number of tests performed per week during two 20-week periods, pre- and post-intervention, and the cost-savings associated with any reductions in testing.
RESULTS: The mean number of fT4 and T3 tests processed per week decreased from 153.7 ± 20.9 and 12.5 ± 7.2, respectively, in the pre-intervention period, to
107.3 ± 11.5 (30% reduction) and 5.1 ± 3.7 (60% reduction) post-intervention. These reductions were sustained for the full 20-week assessment period. Approximated cost savings were $38,000 per year. There was no statistically significant difference in the mean number of TSH measurements in the pre vs. post intervention periods (217.2 ± 28.1 vs. 203.6 ± 9.0 per week).

CONCLUSIONS: Introduction of a reflex fT4 and forced function for ordering thyroid hormone tests resulted in a significant decrease in fT4 and T3 testing. EMR-based interventions can create sustained improvements in health-care utilization, leading to significant cost-savings.

Oral Abstract 2
Comparison of 1- and 3-Month Lupron Depot®(LD) for Pubertal Suppression in Transgender Youth

Zoyah Thawer(1), Zoé Nicolas-Pelletier(2), Vid Bijelic (3), Jemila S. Hamid (3), Scott Somerville (1), Margaret Lawson(1,4), Karine Khatchadourian(1,4).
(1) Department of Pediatrics, Division of Endocrinology and Metabolism, University of Ottawa, Ottawa, ON. (2) Faculty of Medicine, University of Ottawa, Ottawa, ON. (3) Clinical Research Unit, CHEO Research Institute Children's Hospital of Eastern Ontario, Ottawa, ON. (4) CHEO Research Institute, Children's Hospital of Eastern Ontario, Ottawa, ON.

Background: Lupron Depot® (LD) is the preferred method of hormone suppression for transgender youth wanting to stop or delay onset of menses, or decrease erection frequency and intensity. Most transgender youth start 7.5mg monthly LD with stimulated bloodwork at the 3rd injection for LH and sex steroids. If clinical information and hormone levels confirm pubertal hormone suppression, they are switched to 11.25mg 3-monthly. In the last year, we have started 31 youth on 3- monthly LD.
Objectives: To characterize clinical and biochemical pubertal hormone suppression after transgender youth start 7.5mg LD monthly or 11.25mg LD 3-monthly.
Methods: A retrospective chart review of 247 transgender youth seen in our Endocrine Diversity clinic between January 1st, 2014 and May 31, 2019.
Results: 176/247 youth (71%) were natal females, >90% Tanner 4-5; 71 (29%) were natal males, >84% Tanner 4-5. 216 (87%) started on 7.5 mg monthly LD compared to 31 (13%) on 11.25mg 3-monthly. Preliminary results show that natal females' mean LH levels on 3-monthly LD (4.22±4.31 IU/L,n=8) were higher compared to monthly LD (1.78±2.31 IU/L,n=137). Mean estradiol levels were also higher in natal females on 3-monthly LD (98.6±29.36 pmol/L) compared to monthly LD (63.67±24.82 pmol/L). Menses stopped within one month in 87% (39/45) of natal females who started monthly LD compared to 83% (10/12) who started 3-monthly LD. One natal female who started 3-monthly LD continued to have menses 4 months after their first injection.
Mean LH levels of natal males on 3-monthly LD (9.12±5.18 IU/L,n=4) were higher compared to monthly LD (3.07±3.61 IU/L,n=57). Mean testosterone levels were also higher in the 3-monthly group (3.70 ± 4.25 pmol/L) compared to the 1-month group (1.66±2.74 pmol/L). Due to persistent erections or worsening dysphoria, 3 of 9 (33%) transgender females on 3-monthly LD were changed to a higher dose of 3-monthly LD (22.5mg) or 7.5mg monthly LD.
Conclusion: Monthly LD leads to greater suppression of LH and sex steroid levels than 3-monthly LD. Time for menses to cease was not significantly different between the groups, however limited data about menses were collected amongst those started on 3-monthly LD.

Oral Abstract 3
Infantile Hypoinsulinemic Hypoketotic Hypoglycemia: An AKT2 Gene Mutation

Sarah Ames (1), Mark Inman (1), Marisa Chard (1), Munier A. Nour (1)
(1) Department of Pediatrics, University of Saskatchewan, Saskatoon, SK.

Background: Pediatric hypoglycemia is a common clinical entity, yet hypoketotic, hypoinsulemic hypoglycemia without evidence of a disorder of fatty oxidation is rare. Current literature reports only five cases of a novel AKT2 mutation resulting in this rare, but important diagnosis.
Case: A 6-month-old female presented to our center with a history of recurrent, undefined seizures and was found to have severe hypoglycemia (blood glucose of
1.8 mmol/L). Her history was significant for macrosomia, coarse facial features, hypotonia and dysmorphology at birth (including coarse facial features, proptosis, and facial asymmetry with left-sided facial hemihypertrophy) that was undergoing evaluation. Mild, uncomplicated, 'transient' hypoglycemia was noted postnatally but resolved with establishment of feeds by day 3 of life. 'Critical sample' laboratory evaluation at 6 months of age demonstrated hypoketoic hypoglycemia with undetectable insulin levels, appropriate fatty acid elevation, normal liver enzymes, and normal metabolic testing (including acylcarnitine profile and urine organic acids). Glucagon stimulation test resulted in blood glucose rise of 2.2 mmol/L. Recurrent asymptomatic hypoglycemia occurred despite high glucose infusion rates (> 15 mg/kg/min). Treatment with both high dose diazoxide (>15 mg/kg/day) and octreotide were unsuccessful. Genetic testing sent upon clinical suspicion revealed a pathogenic heterozygous mutation in AKT2, c.49G>A, p.(Glu17Lys).

Discussion: This AKT2 gain-of-function mutation has been reported in three previous papers totaling to five reported cases worldwide. This mutation leads to an activation of insulin mediated glucose uptake (via the SLC2A4/GLUT4 transporter), stimulation of glucose storage as glycogen, cell proliferation, and protein synthesis. This has promoted recurrent, severe, fasting as well as non-fasting hypoglycemia and has been associated with dysmorphisms and overgrowth with mainly left-sided hemihypertrophy. Responses to conventional treatments is poor, often
necessitating frequent bolus or continuous feeds. While never reported, there also may be a theoretic role for therapy using mTOR inhibitors.

Conclusion: We present the sixth reported case of hypoketotic, hypoinsulinemic hypoglycemia due to an AKT2 mutation in the world. Due to the downstream nature of this defect, treatment options are limited. In the event of hypoketotic hypoinsulinemic hypoglycemia, especially with associated dysmorphisms and overgrowth, an AKT2 gain-of-function mutation should be considered

Oral Abstract 4
Transient Hyperinsulinism: Off to a Bad Start?

Regev R.[1], Harrington J[1]. Falzone N.[2], Fuchs S. [1,3]
(1) The Hospital for Sick Children, University of Toronto. (2) University of Toronto. (3) Lunenfeld Tanenbaum Research Institute, Mount Sinai Hospital.

Background: Hyperinsulinism is the most common cause of recurrent hypoglycemia in neonates and infants. Hyperinsulinism can be transient (e.g. secondary to perinatal stress or maternal diabetes), or persistent (monogenic and syndromic forms). Transient hyperinsulinism typically resolves within 6 months. Currently, there are no effective tools to predict severity and time to resolution in infants with transient hyperinsulinism.
Study aim: To assess whether early glucose trends predict disease duration and secondary outcome measures in transient hyperinsulinism.

Method: A retrospective cohort of infants admitted with hyperinsulinism, was phenotyped for demographic, clinical and laboratory parameters, treatment and follow-up post-discharge.
Blood glucose (BG) values were collected from the first documented hypoglycemia for 120 hours (5 days). BG data was analyzed for mean BG, cumulative BG AUC (area under the curve), mean daily BG, daily BG variability, time and AUC above, below and within the normal range (defined as 3.5-7.0 mmol/L).

Results: Of 96 patients with hyperinsulinism, 53 infants had transient hyperinsulinism (defined as normoglycemia off all treatment within the first year of life). Thus far we have conducted a preliminary analysis of the initial 5-day BG trend for 10 patients with hyperinsulinism ranging in duration from 6 to 241 days.
Of 10 infants with transient hyperinsulinism, 6/10 had hypoglycemic CHI i.e. all BG values between hypo and normoglycemia. 4/10 had labile CHI identified by BG values ranging from hypo to hyperglycemia, while receiving clinical standard care. Notably, in the infants with labile hyperinsulinism, 5-day BG measures positively correlated with the primary outcome, increased disease duration [time above range ($p=0.012$ $R2=0.97$), mean BG ($p=0.029$ $R2=0.947$) and cumulative BG AUC ($p=0.0009$ $R2=0.98$)].
This pilot data will be expanded upon to include analysis of the entire cohort of infants with transient hyperinsulinism.

Discussion and impact: Preliminary pilot data suggests that for a subgroup of patients with transient hyperinsulinism and labile glycaemia within the first 5 days, BG measures may allow prediction to time of resolution of the hyperinsulinism. If validated, this will benefit clinical decisions as well as offer insight into the heterogeneity underlying the clinical course of infants with transient hyperinsulinism.

Oral Abstract 5
How Does Access and Cost of Gender-Affirming Surgery Differ for Adolescents and Adults Across Canada?

Zachary Zytner (1,2), Karine Khatchadourian (1,3), Kevin Cheung (1), Ken Tang (3), Margaret L. Lawson (1,3). (1) Children's Hospital of Eastern Ontario, Ottawa, ON. (2) Northern Ontario School of Medicine, Sudbury, ON. (3) Children's Hospital of Eastern Ontario Research Institute, Ottawa, ON.

Introduction: Gender dysphoria is defined as distress caused by an incongruence between a person's gender identity and gender assigned at birth. Gender-affirming treatment to alleviate gender dysphoria may include gender-affirming surgery. To ensure equitable and timely access to care, it is essential to understand if barriers to surgical care exist. We hypothesize that conditions and cost of gender-affirming surgery vary significantly within and between the provinces and territories of Canada. Some individuals may accrue significant personal expense ranging from transportation cost to direct costs for surgery. Data regarding access and cost of gender-affirming surgery are needed so individuals can have the information they need to make informed decisions about their health.

Objective: The primary objective is to describe wait times, personal cost and criteria for gender-affirming surgery within and between provinces and territories.

Methods: A cross-sectional survey was distributed electronically by nine Canadian medical specialty associations to physicians, surgeons and nurse practitioners across Canada who care for transgender and non-binary youth and adults.

Results: A total of 314 physicians and nurse practitioners across Canada responded to the survey, 143 of whom indicated they provide gender-affirming care. Survey questions were related to top surgery and to bottom surgery. There was no difference in wait times for appointments between provinces ($p=0.981$), though wait times differed between specialties for adolescents ($p=0.003$) and adults ($p<0.001$). Mean wait times for surgery were not significantly different between provinces ($p=0.094$). Personal cost of top surgery in transgender males varied between no cost in British Columbia, Quebec and Saskatchewan up to $9,000 in other provinces including Ontario. A majority of practitioners indicated minimum age of 16 for referral for mastectomy and chest contouring, though 45% of plastic surgeons and 22.5% of medical practitioners indicated younger minimum ages of 14 or 15. The stability of mental health conditions was the single most important criterion for surgery.

Conclusions: Wait times for appointments and surgery are similar across Canada, though wait times differ between specialties. Personal cost of top surgery differs across Canada, so advocacy for full coverage under provincial health plans across the country may be warranted.

Oral Abstract 6
Predicting Gestational Age Improves Newborn Screening for Congenital Adrenal Hyperplasia

Danny Jomaa (1), Matthew Henderson (2), Steven Hawken (3), Pranesh Chakraborty (2).
(1) School of Medicine, Queen's University, Kingston, ON. (2) Newborn Screening Ontario, Children's Hospital of Eastern Ontario, Ottawa, ON. (3) Clinical Epidemiology Program, Ottawa Hospital Research Institute, Ottawa, ON.

Newborn screening for congenital adrenal hyperplasia (CAH) is performed using a two-tier approach. The first tier involves comparing neonate 17- hydroxyprogesterone levels to gestational age (GA)-based thresholds. When GA is unreported, which occurs in approximately 5% of births, birth weight (BW)-based thresholds are the only available option. However, these have a lower specificity and result in more false positive results. Recently, a predictive model was developed to estimate GA based on newborn demographics and screening analytes measured in a blood sample. The objective of this study was to determine whether supplying a predicted GA to newborns with unreported GA, and subsequent GA- based screening, has a higher positive predictive value (PPV) than BW-based screening.
Screening data was obtained for approximately 700,000 births that occurred in Ontario between 2011 and 2015. Predicted GA was calculated using the model described above. The PPV of BW- and predicted GA-based screening was calculated for newborns with unreported GA. A sequential approach was then developed whereby newborns with unreported GA were first screened by BW-based screening. Newborns that screened positive were supplied with their predicted GA and screened using GA-based thresholds.
First-tier CAH screening using GA-based 17-hydroxyprogesterone thresholds had a higher PPV than using BW-based thresholds (1.30% vs. 0.82%). In the study time period, 3.61% of newborns had unreported GA. For these newborns, predicted GA- based screening had a higher PPV than BW-based screening (0.83% vs. 0.76%) and correctly identified the 2 infants with CAH and unreported GA. A sequential screening approach was then used: BW-based screening then, for the screen positive population, predicted GA-based screening. This further increased the PPV compared to BW-based screening (0.95% vs. 0.76%), reduced the false positive rate, and identified true positive cases. Reducing the false positive rate of CAH screening is important to prevent unnecessary second-tier screening and referrals. For newborns with unreported GA (4-5% of births), BW-based screening is the only available approach. However, this has poor specificity and a high false positive rate compared to GA-based screening.

This study is the first to demonstrate an alternative screening strategy with a higher PPV for newborns with unreported GA.

Oral Abstract 7
An Incidentally-Discovered Paraganglioma

Marian Thorpe (1), Rebecca Mitchell (2), Manpreet Doulla (1)
(1) Department of Pediatrics, Division of Pediatric Endocrinology, University of Alberta, Edmonton AB. (2) Department of Medicine, Division of Endocrinology, University of Alberta, Edmonton AB

Background:
Pheochromocytomas and paragangliomas are rare neuroendocrine tumors arising from chromaffin cells of the adrenal medulla and extra-adrenal cells. Functional tumors often cause clinical symptoms related to catecholamine release, however some children can be asymptomatic. We describe a patient presenting with acute appendicitis, in whom severe hypertension following appendectomy led to the discovery of an incidental paraganglioma. We highlight the clinical and imaging features of paragangliomas and discuss the importance of appropriate medical therapy with alpha blockade.

Case Presentation:
A previously healthy 17-year-old male presented with abdominal pain secondary to acute appendicitis and was treated with an uncomplicated open appendectomy.
Post-operatively, he had episodic hypertension with systolic blood pressures up to 300mmHg. He was admitted to the pediatric intensive care unit and started on nitroprusside and beta blocker therapy. While on Esmolol, he had dramatic fluctuations in his blood pressure with corresponding severe bradycardia and hypotension. A clinical diagnosis of catecholamine secreting tumor was made, and he was initiated on alpha blockade with Doxazosin, resulting in rapid stabilization in both blood pressure and heart rate. Subsequent investigations revealed elevated plasma and urine catecholamines, with norepinephrine and normetanephrine predominance. CT scan showed a 6cm posterior mediastinal mass adjacent to the descending aorta with strong uptake on MIBG-scintigraphy and limited uptake on FDOPA- PET imaging, which were both negative for metastases. Genetic investigations revealed an SDHB gene mutation.

Conclusion:
This case describes the clinical presentation of an incidentally-discovered paraganglioma and highlights the factors involved in false positive and false negative findings on MIBG-scintigraphy and FDOPA -PET imaging of paragangliomas. The importance of alpha blockade prior to beta blocker therapy is highlighted for the medical management of these tumors, and pitfalls of beta-blockade therapy are reviewed.

Oral Abstract 8
Socioeconomic Disparities in PUMP Uptake Among Canadian Children and Adolescents With Type 1 Diabetes: Comparison of Two Provincial Programs

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Objectives: Even with universal healthcare, previous studies have shown that disparities in socioeconomic status (SES) exist in uptake of insulin pump therapy by patients with type 1 diabetes. We hypothesized that SES disparities would be reduced in Quebec, where pumps and all associated supplies are fully funded by the Insulin Pump Access Program, as compared to Manitoba, where the cost of additional supplies is only partially covered by Pharmacare.

Methods: We conducted parallel population-based retrospective cohort studies using multiple linked health administrative data. Cohorts were defined as follows: in Quebec, children ages 1 – 17 years diagnosed with type 1 diabetes from 1996 to 2016 were identified using a validated definition for use within administrative data, and in Manitoba, children under age 18 years diagnosed with type 1 diabetes between 1996 and 2017 were identified using a provincial clinical registry. Our primary outcome was pump uptake, identified by specific physician billing codes or clinical data from 2011 to 2017 (Quebec) or 2012 to 2017 (Manitoba). Our primary
exposure was SES using a validated area-based deprivation index to assign material and social deprivation quintiles. We used multivariable logistic regression analysis to determine the association of SES with pump uptake, adjusted for age, sex, and rurality.

Results: In both provinces, as material deprivation increased from least deprived to most deprived, the odds of initiating pump therapy decreased (odds ratio (OR) 0.88, 95% confidence interval (95% CI) 0.83-0.93 (Quebec); OR 0.68, 95% CI 0.57-0.81 (Manitoba)). Further, on comparison of the provinces, the effect of material deprivation on pump uptake in Manitoba had a more significant effect than in Quebec.

Conclusions: While SES disparities exist in both provinces, comprehensive government financial support in Quebec seems to mitigate the impact of material deprivation on pump uptake as compared to limited support in Manitoba. Further efforts should focus on exploring other drivers of SES disparities in pump therapy uptake.

Oral Abstract 9
Influence of Pre-Diabetes on Respiratory and Nutritional Status in the Montreal Cystic Fibrosis Cohort

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Background: A landmark study by Lanng et al. (1991) demonstrated clinical deterioration over 6 years prior to the diagnosis of cystic fibrosis-related diabetes (CFRD). The aim of this study was to determine the impact of a pre-diabetic state on the nutritional and respiratory status in a more recent generation.

Methods: We performed a retrospective analysis of respiratory function (FEV1%) and body mass index (BMI) in subjects from the Montreal Cystic Fibrosis cohort. Analysis was restricted to those who maintained normoglycemia (NG, n=37) and in patients who developed de novo CFRD at their last visit (n=49). Subjects were further grouped according to pancreatic sufficiency (PS) or insufficiency (PI).

Results:
At the last screening visit, subjects had had a mean age of 31.7 ± 8.1 years, BMI of 23.0 ± 4.0 kg/m2, FEV1% of 70.1 ± 24.2%, and 81% had PI. Patients were divided intro four groups as follows: NG+PS (N=16), NG+PI (N = 21), CFRD+PS (n=3), and CFRD+PI (n=46). We excluded the CFRD+PS group from analysis. Post-OGTT 2h- glucoses were increased in the CFRD+PI group (2h-post PGTT, NG+PS, 5.2 ± 1.1 mmol/L; NG+PI, 5.2 ± 1.1 mmol/L; 12.8 ± 1.1 mmol/L, p<0.0001).

The NG+PS group had significantly greater BMI and FEV1 at their most recent screen, as compared to NG+PI and CFRD+PI groups (BMI, 26.2 ± 3.6 kg/m2 vs 22.6 ± 4.2 kg/m2 vs 22.1 ± 3.5 kg/m2, p = 0.0016; FEV1,(91.5 ± 16.8% vs 67.8 ± 25.3% vs 63.5 ± 22.2%, p = 0.0002).

We compared the rate of change in BMI and in FEV1 over 6 years prior to the most recent visit (NG+PS, NG+PI groups) or prior to the diagnosis of de novo CFRD. The rate of decline in BMI (p = NS) and in FEV1 (p = NS) were similar between the 3 groups.

Conclusion: A pre-diabetic state has no clinical impact prior to the onset of CFRD, while pancreatic insufficiency adversely effects BMI and FEV1 in all patients with CF. Early diagnosis of CFRD is critical to minimize the decline in nutritional and respiratory status that after progression to overt diabetes.

Oral Abstract 10
Cyclical Cushing Syndrome in a Patient With Carney Complex: A Rare Association and a Management Challenge

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A minority of patients with Cushing syndrome (CS) exhibit periods of hypercortisolism interspersed with periods of normal secretion; a phenomenon known as cyclical Cushing Syndrome. Etiologies include pituitary or ectopic adrenocorticotropic hormone (ACTH) secretion and primary adrenal disease. Carney complex (CC) is a rare multiple endocrine neoplasia syndrome characterized by distinctive skin and mucosae lesions, primary pigmented nodular adrenal disease (PPNAD) and increased risk of different tumors.

Case: 20-year old male, who presented at age 3 years with weight gain, poor energy, hypertension and cushingoid facies. He had similar symptoms 6 months before, which resolved spontaneously. Past personal and family history were unremarkable. Initial workup showed elevated 24-hour cortisol excretion (998 mmol/d) and serum cortisol post 0.375 mg dexamethasone (772 nmol/L); ACTH <0.1 pmol/L. There was no evidence of adrenal adenomas on imaging. He underwent a left adrenalectomy, and pathology showed multifocal adrenal microadenomas. Following the surgery, symptoms resolved. He had bluish-black lentigines on his face and oral mucosa, raising the possibility of CC. Confirmatory genetic testing identified a mutation in PRKAR1A gene. After seven years of remission, he showed activation of his right adrenal gland, with growth deceleration, muscle weakness and hypercortisolism. Since then, he has had flares every 1-3 years, lasting a few weeks. These are followed by adrenal suppression, leading to two hospitalizations for adrenal crisis. He receives hydrocortisone during stress, but not for daily needs. The possibility of right adrenalectomy was discussed, but not chosen due to relatively short and infrequent flares of CS and good quality of life. He undergoes regular screening with no other findings of CC (normal echocardiograms, normal thyroid and pituitary function, no testicular masses). He has osteopenia, with no evidence of vertebral fractures [BMD Z score 1.8-2.1].

Conclusion: We present a patient with PPNAD-associated Cyclical CS and his challenging management. CC should be considered in the differential for ACTH- independent CS, particularly if there are characteristic skin lesions. Timely diagnosis and surveillance of both CS and CC can help prevent complications. Even though bilateral adrenalectomy is the recommended treatment, this should be individualized.

Oral Abstract 11
Perspectives on a Type 1 Diabetes Pediatric-to-Adult Transition Program in Vancouver, Canada

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Background: Type 1 Diabetes (T1D) is a chronic disease characterized by autoimmune destruction of insulin-secreting pancreatic beta cells. Recently, there has been increasing recognition of the specific health risks associated with adolescents (ages 10-18) and emerging adults (ages 18- 25) with T1D. Deterioration in metabolic control and disengagement with healthcare have been well documented at the time of transition to adult care. This may lead to increased microvascular complications, hospitalizations, and mortality. To address these issues, numerous T1D pediatric-to-adult transition models have been proposed, however the most effective model remains unclear.

Objective: To obtain perspectives on the pediatric-to-adult transition in T1D care from adolescents, young adults, and parents in BC and to provide evidence for formulating and implementing a T1D transition clinic affiliated with BC Children's Hospital (BCCH).
Methods: In this mixed methods study, we conducted eight focus groups with adolescents and young adults aged 16-35 (n=38) and four focus groups with parents (n=17). All participants completed a demographics form and a Transitions Intervention Survey, where they rated different transition interventions on a 10- point Likert scale. Audio recordings were transcribed and survey results were tabulated. Thematic content analysis of qualitative data is being analyzed using the grounded theory approach.

Results: Among the 55 participants, the five most appealing interventions identified in the Transitions Intervention Survey were (mean score ± SD): identifying an adult care provider suitable for the patient (9.05±1.63), good communication, including written communication between pediatric and adult care providers (9.05±1.80), ability to rebook appointment if appointment is missed (8.87±1.54), mental health resources (8.87±2.25), and discussion with the adolescent in advance as to the best time for transition (8.80±2.15). Preliminary findings of qualitative data reveal that stigma, discrimination, and mental health are issues that need to be addressed in this vulnerable population.

Conclusion: Transition models that consider practice patterns of the adult provider may be important in helping adolescents with T1D transition to adult care. Interventions to address stigma, discrimination, and mental issues such as counselling, educational workshops, and mentorship may be required in order to facilitate a more successful transition to adult care.

Oral Abstract 12
Frequency of Contacts with the Diabetes Team in Pediatric Patients with Type 1 Diabetes on an Insulin

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Background:
Type 1 diabetes (T1D) management has become technologically advanced with the use of insulin pumps, continuous glucose monitoring and diabetes data management software. Although, these technologies exist, research shows they are underutilized by patients. Several studies have shown improvement in A1C when adult patients review their blood glucose trends with their diabetes team between clinic visits. This study aimed to assess if these findings could be replicated in a pediatric population.
Objective:
The primary objective was to assess if there was a correlation between A1C within target (</=7.5%) and frequency of contact with the diabetes team between clinic visits. Secondary objectives included determining barriers to contact and the change in A1C from baseline to end of study.
Methods:
Patients with T1D (1 – 19 years) using an insulin pump for one year or more were approached to participate. Patients completed a questionnaire detailing their diabetes management over the past year. A1C and diabetes team contacts for insulin adjustments between clinic visits were collected for the past year.
Results:
176 participants were enrolled, 46% female, with a mean age of 12.9 years. Mean duration of T1D was 6.7 years with a mean duration of pump use of 4.5 years. Mean A1C at baseline was 8.1%. 66% of patients contacted the diabetes team for insulin adjustments between clinic visits with a mean of 1.2 contacts. 90% of contacts were by e-mail, 1% by phone and 10% combined phone and e-mail. There was no significant relation between target A1C and frequency of contact ($p>0.05$). There was a significant negative correlation ($r=0.20$, $p = 0.009$) between age and contact. In addition, longer duration of pump use was associated with decreased frequency of contact ($r=0.17$, $p = 0.022$). Barriers to contact included too busy (41%), and technical problems with software (39%).
Conclusion:
There was no significant relationship between frequency of contact with the diabetes team and A1C. There was a low frequency of contact with the diabetes team for insulin adjustments in between clinic visits. This raises the urgency of exploring strategies to engage pediatric patients with T1D.

Poster Abstracts

Poster Abstract 1
The Impact of Hospital Surgical Volume on Healthcare Utilization and Surgical Outcomes for Pediatric Thyroidectomy

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Introduction: Children are more likely to experience surgical complications of thyroidectomy than adults. Higher surgeon volume is linked to better surgical outcomes in children and adults. Hospital volume (HV) and healthcare utilization outcomes are less often reported, particularly in pediatrics, and we lack Canadian data. We investigated associations between pediatric thyroidectomy HV with surgical and healthcare utilization outcomes in Ontario using population-level data. Methods: Retrospective analysis of Ontario's administrative health data, 1993-2017; a province-wide single-payor insurance system permits sampling the entire population. A cohort was established of 1908 thyroidectomies – excluding repeat procedures – in patients under 18 years. HV was defined per-case as the number of thyroidectomies at a site in the preceding year. Thyroidectomies were divided into HV quartiles. The healthcare utilization outcomes included: length of stay (LOS), readmission, emergency department (ED) visits. The surgical outcomes were: hematoma, disease-free survival. Multivariate analysis adjusted for demographics, diagnosis, and thyroidectomy type.

Results: Mean age was 10.4 ± 5.4 years, 61% female and 20% had a cancer diagnosis. The lowest HV quartile performed 0-1 thyroidectomies annually and 30% of thyroidectomies overall. Most patients are referred out of region; one region performed 36.4% of thyroidectomies province-wide. However, patients from regions with a low-volume hospital are more likely to be treated at a low-volume hospital.

LOS was 0.64 days longer in the highest, versus the lowest, HV quartile. HV was not associated with readmission or ED visits. Male sex, increased socioeconomic deprivation, and rurality were associated with increased ED visits. Hematoma risk increased by 2.88 in the highest, versus the lowest, HV quartile (overall event rate 1.7%). HV was not associated with disease-free survival from cancer (overall 76.3%).

Conclusion: Higher HV associated with increased hematoma and LOS and may reflect patient complexity at these centres. Low HV was not linked to poor outcomes; these data support that surgeon, rather than hospital, volume impact thyroidectomy outcomes. Further study of groups disproportionately accessing ED may help direct resources to these populations.

Poster Abstract 2
Safety and Efficacy of Subcutaneous Insulin for Treatment of Diabetic Ketoacidosis in Children – A Systematic Review

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OBJECTIVES: Standard treatment of diabetic ketoacidosis (DKA) uses intravenous (IV) insulin infusion which carries significant risks without sophisticated equipment and high-dependency units. Alternative treatment protocols are needed for low- resource settings.

METHODS: We systematically searched PubMed (MEDLINE), EMBASE, Cochrane and the grey literature for studies evaluating subcutaneous (SC) or intramuscular (IM) insulin administration for the treatment of pediatric DKA before 04/06/2018. Two independent reviewers screened abstracts and full text papers, extracted data and assessed risk of bias.

RESULTS: Of 2934 abstracts, 137 qualified for full text review, and 23 for data extraction, including five randomized controlled trials, 14 cohort studies and 4 case reports/series, for a total of 402 episodes of DKA. Based on a historical shift in DKA management from high-dose to low-dose insulin, studies were grouped into publication before 1990 (n1=19 [79%], "past") vs. thereafter (n2=5 [21%], "recent"). Age and sex were no different, GCS <12 was more common in past (37/68 episodes) vs. recent (14/90 episodes) studies (p=0.01). Of 245 (61%) episodes where DKA severity was reported, DKA was mild, moderate and severe in 12 vs. 30%, 28 vs. 59% and 60 vs. 12%, p=0.002. All past vs. 3/5 (60%) recent studies used Regular, while 2/5 recent studies used Ultra-Rapid. Thirteen past vs. no recent studies combined IV+IM or IV+SC administration for the initial bolus. Bolus dose and frequency ranged widely, especially in past studies. Death (n=10) and cerebral edema (n=2) occurred in 3 studies from India, Ethiopia and Sudan in severely ill patients at presentation. DKA recurred in 1 recent episode, severe hypoglycemia occurred in 5 past episodes. In the 12 (52%) studies that had a control group, rates of death, cerebral edema, severe hypoglycemia, and DKA recurrence were no different in SC/IM treated cases (n=169) vs. IV treated controls (n=143). Hypokalemia differed marginally (24% vs. 6% in case vs. control, p=0.05). Risk of bias was overall assessed as high, mainly as a result of study design and limited outcome reporting.

CONCLUSION: Emerging evidence suggests that low-dose SC insulin may be a safe and effective treatment of all severities of pediatric DKA. More high-quality studies are needed.

Poster Abstract 3
A Simulation-based Intervention Teaching Illness Management Skills to Caregivers of Children with Adrenal Insufficiency: a Randomized Controlled Study

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Background: Permanent adrenal insufficiency (AI) is an uncommon but potentially life-threatening condition in children. Patients are at particular risk during times of stress. Thus, caregivers should have good illness management skills. Simulation (SIM) is increasingly used in medical education, but its use in teaching illness management to caregivers of children with AI has not been evaluated.

Objectives: To compare the impact of illness management teaching delivered using SIM or traditional teaching on caregiver knowledge, confidence and ability to manage illness in a child with AI. 2) To determine retention of caregiver knowledge, confidence and ability 6 months post- teaching.

Methods: Subjects were randomly assigned to SIM or traditional teaching. Participants completed knowledge/self-confidence questionnaires and performance assessments using SIM scenarios at their baseline visit (before and after teaching) and 6-months later.

Results: 41 caregivers of mean age (SD) 40.6 (8.7) years, of children with AI of mean duration (SD) of 6.0 (4.7) years; were randomized to receive SIM-based teaching (n=20) or traditional teaching (n=21). 61% of participants were female. Knowledge Scores (max:10) increased in Traditional and SIM groups, respectively, from pre-teaching 7.2(2.1);7.8(1.7) to post-teaching 8.3(1.6);p=0.005*;8.4(1.0);p=0.08 and 6mo visits 8.6(0.9);p=0.43;8.7(1.0);p=0.37. Confidence Scores (max:40) increased in Traditional and SIM groups, respectively, from pre-teaching 29.6(6.9);31.5(3.0) to post-teaching 38.1(2.9);p<0.001*;38.8(1.6);p<0.001*) and 6mo visits 37.3(4.0);p=0.34;35.2(7.8);p=0.05. Sim-Scenario Scores (max:26) increased in Traditional and SIM groups, respectively, from pre-teaching 18.6(5.2);16.8(5.6) to post-teaching 23.5(1.6);p<0.001*;21.7(2.8);p=0.003* and 6mo visits 21.4(2.9);p=0.09;22.5(1.7);p=0.15. Change from post-teaching to 6mo visit in Traditional and SIM groups, was -1.96(3.47);p=0.09 and +1.23(2.86);p=0.15 respectively, with difference between groups: p=0.03*.

Conclusion: Caregiver performance was sub-optimal at baseline and highlights the need for on- going teaching of caregivers of children with established AI. Caregiver knowledge, confidence and performance improved significantly post-teaching in both arms with no difference observed between SIM and traditional teaching. However, caregivers randomized to SIM teaching performed better at 6 months during SIM scenarios of adrenal crisis than those with traditional teaching. No significant differences were observed in caregiver knowledge or confidence between the baseline assessments and on follow-up at 6-months. Our findings suggest SIM teaching in this context is more effective than the current standard of care.

Poster Abstract 4
Let's Go to Camp: Type 1 Diabetes Innovations - From the Lab to the Field

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Background: Type 1 diabetes technologies such as continuous blood glucose monitoring and artificial pancreas systems are rapidly being developed while translation to the real-world setting is slower. Camps for youth with diabetes both benefit from the use of new technologies and provide a real-world setting for research translation.

Objective: To develop an innovation loop that translates new diabetes technologies to camps for youth with diabetes and feeds real-world experiences back to the research lab.

Methods: A collaboration between Montreal artificial pancreas research groups and Camp Carowanis, a camp for youth with diabetes in Quebec, Canada, was established in 2014. Four novel advancements for the treatments of type 1 diabetes have been developed and tested at the camp: a dual-hormone artificial pancreas, a basal rate and carbohydrate learning algorithm for the artificial pancreas, an optimization algorithm for multiple daily injection users, and an overnight wireless glucose sensor monitoring system.

Results: Between 2014 and 2019, over 160 campers and counselors at Camp Carowanis from ages 7 to 21 have participated in these four projects. (i) In 2014, the dual-hormone artificial pancreas led to a median of 0% (IQR 0.0–2.4) of nighttime spent below 4.0 mmol/L compared to 3.4% (0–11.0) with conventional pump therapy (p=0.0048). (ii) In 2018, the mean time campers spent between 3.9 and 10mmol/L using the artificial pancreas was 55% (SD=15). (iii) The multiple daily injection learning algorithm's changes to carbohydrate ratios and basal injections were approved by physicians 92% of the time. (iv) Finally, the overnight wireless glucose sensor monitoring system was effective in detecting nocturnal hypoglycemia events and intervening when necessary. The data and experience from the field was then brought back to the lab to further improve the technologies. In turn, lessons learned from these studies was presented to the camp team in order to potentially modify camp practices.

Conclusion: The success and recurrence of these projects have exemplified the effective innovation pathway that is possible between research teams and diabetes camps for the improvement of type 1 diabetes treatments.

Poster Abstract 5
Did Pamidronate Cause Pulmonary Edema in a Neonate treated for Hypercalcemia Secondary to Subcutaneous Fat Necrosis?

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Intravenous Pamidronate has been successfully used to treat neonatal hypercalcemia secondary to subcutaneous fat necrosis (SCFN). Pamidronate, a bisphosphonate, acts by binding to hydroxyapatite on bone mineral surfaces where it is selectively internalized by osteoclasts, inhibiting their activity and thus suppressing bone resorption. Side effects of pamidronate include an acute flu-like reaction with fever, myalgia, bone pain, vomiting, and hypocalcemia. However, case reports have shown that this treatment is well tolerated in neonates even when they are given three to four doses (0.5-1.0 mg/kg/dose), suggesting that pamidronate may be used as first-line treatment for severe hypercalcemia in SCFN. The purpose of this report is to describe a case of a neonate with hypercalcemia secondary to SCFN who was treated with a second dose of intravenous pamidronate and subsequently developed pulmonary edema requiring intubation. The patient was born at 38 weeks via emergency C-section, suffered asphyxia and developed SCFN post therapeutic hypothermia. At 4 weeks of life, he presented with a history of 3-4 days of irritability and extensive SCFN lesions including along his mandible and on his gums. His ionized calcium was 1.85 mmol/L, prompting immediate admission for hyperhydration and treatment with furosemide, calcitonin, followed by 1 dose of IV pamidronate 0.5 mg/kg. The dose of pamidronate was divided into 2 due to loss of the intravenous line mid-infusion. He developed a mild fever after the first half-dose of pamidronate. After his second half-dose, he had increasing peripheral edema and exhibited signs of respiratory distress, and clinically remained intravascularly deplete. After receiving albumin, he developed flash pulmonary edema requiring intubation and admission to the intensive care unit as his blood gas revealed a pH of 6.9 (pCo2 69.5 mm Hg, HCO3 12 mmol/L, lactate 6.6 mmol/L). We hypothesize that the pulmonary edema was due to a systemic inflammatory response caused by the IV pamidronate, which has been previously reported as an adverse side effect, and that the albumin may have exacerbated the clinical picture. To our knowledge this is the first case of pulmonary edema reported to occur post administration of pamidronate in the context of hypercalcemia due to SCFN.

Poster Abstract 6
Hypoglycemia with Acute Lymphoblastic Leukemia Treatment in Pediatric Patients: A Novel Case Series

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Introduction:
Acute lymphoblastic leukemia (ALL) is the most common pediatric cancer. One of the mainstays of chemotherapy is the enzyme L-asparaginase which is known to improve outcomes. When L-asparaginase is linked to polyethylene glycol (PEG- asparaginase) it decreases immunogenicity of the enzyme and prolongs half-life. Hyperglycemia is a known side effect of L-asparaginase, however, there have been recent case reports of hypoglycemia associated with this medication. Other case reports also describe severe hypoglycemia associated with 6-mercaptopurine (6- MP), but it is unclear if effects are independent of, or connected to PEG- Asparaginase.

This case series illustrates a temporal relationship between PEG-asparaginase, 6-MP administration and hypoglycemic events and describes possible mechanisms. To our knowledge, this is the first case series that demonstrates an association between PEG-asparaginase and hypoglycemia.

Methods:
We performed a retrospective chart review of six children with documented hypoglycemia who were being treated for ALL at the Children's Hospital of Eastern Ontario from May 2016 to August 2019. The definition of hypoglycemia was a serum blood glucose of <3.0 mmol/L. The timing and duration of hypoglycemia relative to the administration of PEG-asparaginase, 6-MP, corticosteroids, and other chemotherapy agents was determined. When available, laboratory values of the critical sample were collected.

Results:
The average age of the cohort was 3.6 years (8 months to 11 years). Three patients had trisomy 21. Timing of hypoglycemia onset ranged from 7-21 days after the most recent PEG-asparaginase administration. Critical samples were drawn in 5/6 patients with 14 episodes documented. Six events occurred while patients were on high dose corticosteroids. Venous blood glucose values ranged from 1.9-3.0mmol/L with inappropriately low beta-hydroxybutyrate (<0.10- 0.77mmol/L) and inappropriately detectable serum insulin (10-74pmol/L). Two patients who were receiving 6-MP concurrently also had ketotic hypoglycemia.

Conclusion:
Patients undergoing ALL therapy are at risk of both non-ketotic and ketotic hypoglycemia. Hypoglycemia associated with PEG-asparaginase occurred later and lasted longer than previous reports with L-asparaginase. Hyperinsulinism is the most likely mechanism of PEG-asparaginase related hypoglycemia. A non-ketotic etiology may be associated with 6-MP. An understanding of these risks

will allow clinicians to develop strategies to appropriately investigate and prevent episodes of hypoglycemia.

Poster Abstract 7
Combined Indeterminate and Impaired Glucose Tolerance is a High Risk for Cystic Fibrosis-Related Diabetes in the Montreal Cystic Fibrosis Cohort

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Background: Cystic fibrosis-related diabetes (CFRD) occurs in up to 50% of patients with CF and is associated with increased morbidity and earlier mortality. Annual oral glucose tolerance testing (OGTT) is recommended annually. Early dysglycemia (post-OGTT 1h-glucose > 11.1 mmol/L; INDET) and impaired glucose tolerance (post- OGTT 2h-glucose between 7.8 -11.1 mmol/L, IGT) are independently associated with the risk of progression to CFRD. In this study, we sought to determine whether patients meeting criteria for both INDET and IGT might have a higher risk of diabetes.

Methods: The Montreal CF cohort was established in 2004 for prospective observational yearly assessment of pulmonary function (FEV1%), oral glucose tolerance testing (OGTT), and anthropometry. We analyzed diabetes risk according to initial OGTT classification.

Results: A total of 293 patients with CF were included in this study, with a mean age of 25.5 ± 7.7 years, a BMI of 21.7 ± 3.0 kg/m2, and FEV1% of 73.2 ± 22.1%. Eighty percent of the patients had pancreatic insufficiency. At initial screening, subjects met the following classifications: normal glucose tolerance (NGT, 39.2%); indeterminate glycemic status alone (INDET, 16.4%), impaired glucose tolerance alone (IGT, 13.3%), combined indeterminate and impaired glucose tolerance (INDET+IGT, 16.4%), or de novo CFRD (14.7%). We performed Kaplan-Meier analysis of progression to CFRD in 198 patients who did not have diabetes at their initial visit and who were followed serially (mean 6.9 ± 3.8 years). Subjects in the IGT and INDET+IGT rates had a significantly higher probability of developing CFRD over the first 7 years, while the NGT group had the lowest risk ($p = 0.0043$). The proportion of patients who developed diabetes were as follows: NGT, 17.2 ± 3.8%; INDET, 20.0 ± 4.1%; IGT, 31.0 ± 4.7%; and INDET+IGT, 41.6 ± 5.0% ($p = 0.02$). There was no difference between BMI ($p = 0.3799$) or FEV1% (0.2410) between groups and between initial and final screening visits.

Conclusion: Patients who fulfill criteria for both indeterminate and impaired glucose tolerance have a higher risk of developing CFRD and at younger age.

Poster Abstract 8
A New Twist on a Rare Form of Rickets

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A previously well 14-month-old boy presented to medical attention for concerns of bony lesions over the ribs. Investigations revealed hypocalcemia with a total calcium of 1.61 mmol/L (2.25 – 2.75) and the patient was directed to the Emergency Department. There, hypocalcemia was confirmed with ionized calcium 0.85 mmol/L (1.22 – 1.37), total calcium 1.56 mmol/L (2.22 – 2.54) and albumin 49 g/L (35 – 47). Past medical history was significant for isolated gross motor delay. Diet was appropriate for age with no restrictions. Family history was unremarkable and parents were of Indian descent and non-consanguineous. Physical exam demonstrated frontal bossing and prominent costochondral junctions. Additional investigations included elevated parathyroid hormone of 380 ng/L (12 – 78), elevated alkaline phosphatase of 1,677 U/L (143 – 318), sufficient total 25- hydroxyvitamin D of 85 nmol/L and low 1,25-dihydroxyvitamin D of 22 pmol/L (48 – 190). Knee and wrist radiographs identified fraying and cupping of the distal metaphyses, consistent with rickets.

The patient was admitted to hospital and started on intravenous calcium gluconate, enteral calcium carbonate, cholecalciferol, and alfacalcidol. By the eighth day of admission, calcium infusion was discontinued, and calcium levels remained stable on alfacalcidol 0.25 mcg/kg/day, vitamin D 3,000 units daily and elemental calcium 200 mg/kg/day. Targeted genetic testing revealed two variants in the CYP27B1 gene: 1) a novel, likely pathogenic frameshift variant in exon 1 (c.144delT, p.L49WfsX29) predicted to result in protein truncation and 2) a variant of unknown significance in exon 8 (c.1243A<C, p.T415P).

Vitamin D-dependent rickets type 1 (VDDR 1A), also known as 1-alpha-hydroxylase deficiency, is an autosomal recessive disorder characterized by the inability to convert 25-hydroxyvitamin D to 1,25-dihydroxyvitamin D secondary to inactivating mutations of CYP27B1. Given the degree of clinical, biochemical, and radiographic similarity, VDDR 1A should be considered if there is a concern for nutritional rickets in the absence of 25-hydroxyvitamin D deficiency. This is the first report of a pathogenic effect of the CYP27B1 variant c.144delT. Furthermore, as our patient's clinical presentation was in keeping with VDDR 1A, we propose c.1243A<C as a novel pathogenic CYP27B1 variant implicated in VDDR 1A.

Poster Abstract 9
Psychosocial and Socio-Economic Well-Being as Determinants of Quality of Life and Glycemic Control in Haitian Youth With Diabetes

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Background: Youth with diabetes in low- and middle-income countries have poor glycemic control even when medical care, insulin and diabetes supplies are provided free of charge. In Haiti, health-related quality of life (HRQL) among youth with diabetes is suboptimal.
Objective: To assess psychosocial and socio-economic factors as determinants of glycemic control and HRQL in Haitian youth with diabetes in Haiti.

Methods: Cross-sectional study of patients with type 1 diabetes aged 0 to 25 years old at the Kay Mackenson Clinic (KMC) in Montrouis, Haiti from 07-12/2017. Demographic and socio-economic status (SES) variables and most recent hemoglobin A1c (HbA1c) values were recorded. We administered the Diabetes Quality of Life for Youth questionnaire to assess HRQL and life satisfaction, patients ranked their perceived health and subjective social status, and responded to the Patient Health Questionnaire depression (PHQ-9), Rosenberg Self-Esteem (self- esteem) and Perceived Diabetes Self-Management (self-management) questionnaires. Confirmatory factor analysis and Cronbach's alpha were used to confirm validity and reliability. We used linear regression to evaluate determinants of HbA1c, HRQL and life satisfaction. Statistical significance was $p < 0.05$.

Results: 85 participants (59%F, mean age 17.5±5.0y, mean diabetes duration 3.7±3.5y, mean HbA1c 11.3±2.6%, mean HRQL 61±16/100, mean life satisfaction score 64±20/100) were included. Scale validity was adequate. Depression was absent, mild, moderate and moderate-to-severe in 34%, 38%, 21% and 7%, respectively. On a scale of 0 to 100, mean scores were 61±12 for self-esteem, 45±17 for self-management and 79+24 for perceived health. Subjective social status ranking was 8±2 out of 10. Neither quality of life measure nor any of the psychometric scores predicted HbA1c ($p>0.05$ for all). When adjusted for age, sex and diabetes duration, higher self-esteem ($p=0.003$), higher subjective social status ($p=0.04$) and lower depression score ($p<0.05$) predicted HRQL, while higher objective SES ($p=0.01$) and lower depression score ($p=0.04$) predicted life satisfaction.

Conclusions: Rates of depression, low self-esteem and low perceived self- management skills are high among Haitian youth with diabetes but do not predict their ubiquitously poor glycemic control. Depression and objective socio-economic well-being emerge as intervention targets while subjective psychosocial well-being emerges as a resilience factor for improved HRQL and life satisfaction.

Poster Abstract 10
Strange Behaviour: A Case Report of an Adolescent with an Insulinoma

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Insulinoma, is a rare diagnosis in the pediatric population. Given the low incidence rate of these pancreatic neuroendocrine tumors, Pediatric Endocrinologists may encounter very few cases in their clinical practice. This case report reviews the clinical presentation and management of an insulinoma in an adolescent.

A 13-year-old male was referred with a history of two years of what the family referred to as unusual episodes. During these episodes he would become acutely upset, with palpitations, tremors, and complain of hunger, and each episode would resolve after eating. He was not fasting during any of the episodes described and did not have any upon waking in the morning. He had been previously healthy. He was referred to neurology and the diagnosis of focal seizures was made and he was started on anti-seizure medications prior to seeing endocrinology. Given the history, a fasting challenge was organized, which was suggestive of hyperinsulinemia with two elevated insulin levels at the time of hypoglycemia. The patient was started on dextrose containing intravenous fluids and oral diazoxide. Further work-up included an MRI which showed evidence of a large mass in the head of the pancreas. An octreotide scan demonstrated uptake within the pancreatic mass. The patient went on to have a pancreaticoduodenectomy. Pathology revealed a well differentiated insulinoma of 6.5cm x 4.7cm. Post-operatively the patient had no further episodes of hypoglycemia, his diazoxide was discontinued and he successfully completed a prolonged fast. Genetic testing was also sent for possible association with multiple endocrine neoplasia type 1, which was negative. His anti-seizure medications were also discontinued.

This case highlights the diagnostic difficulty associated with a first presentation of insulinoma. In addition, it provides the opportunity to review the diagnostic pathway and subsequent imaging options prior to final surgical management.

Poster Abstract 11
Idiopathic Infantile Hypercalcemia - Clinical Spectrum of Milder Phenotypes

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Background: Idiopathic Infantile Hypercalcemia (IIH) is an uncommon disorder with variable clinical presentations. Classical cases present with failure to thrive, vomiting and renal calcification, whereas milder phenotypes may present with feeding difficulties or seizure-like-symptoms. Biochemically, IIH is characterized by elevated serum calcium, elevated 1,25 (OH)D and low PTH. Mutations in the CYP24A1 or SLC34A1 genes are causal in many classical cases of IIH. However, characterization of milder phenotypes remains less clear. Study aim: To describe milder phenotypes using biochemical measures, dietary assessment, renal ultrasound and expanded genetic testing.

Methods: Patients between 6 months and 17 years who were followed long term in our Calcium Clinic with clinical and biochemical characteristics of IIH were approached to participate. Eligibility criteria included an elevated serum calcium, 1,25(OH)D level elevated or in the upper normal range, PTH level reduced or in the lower normal range and urinary calcium/creatinine-ratio increased or borderline high. Subjects were excluded if another cause of hypercalcemia had been identified. Detailed dietary assessment, renal ultrasound and relevant biochemical measures were obtained. Stepwise molecular genetic testing was performed with Sanger sequencing of CYP24A1, and if negative, followed by Nephrocalcinosis Sequencing Panel (including 30 genes causing hypercalciuria). Serum levels of vitamin D metabolites were analyzed using LC-MS/MS.

Results: To date, 14 patients with IIH have been involved. Median age at initial presentation was 3.5months where most patients presented with failure to thrive, poor feeding or seizure-like symptoms. All patients are managed with low calcium diet (median DRI 45%). 6 have radiological evidence of renal calcification and 5 had family history of renal stones and/or hypercalcemia. Median serum calcium 2.81 mmol/l, urine Ca:Crea ratio 1.49 mmol/mmol, 1.25(OH)D 178 nmol/l and PTH 11ng/L at initial presentation. Genetic testing came back positive for 9 patients. In addition to CYP24A1 and SLC34A1 mutation, we identified mutations in the genes CLCN5, SLC34A3 and SLC4A1 as possible cause of IIH. 25(OH)/24,25(OH)D-ratio was elevated in patients with biallelic CYP24A1 mutation.

Conclusion: The clinical, biochemical and genetic spectrum of IIH is broader than previously appreciated. Heterozygous mutations may play a role in milder phenotypes and elevated 25(OH)/24,25(OH)D-ratio seems to be specific for biallelic CYP24A1 mutations.

Poster Abstract 12
Improving Sick Day Management among Patients and Families with Type 1 Diabetes

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Background: For patients with Type 1 Diabetes, sick day management is a critical component of diabetes education. Complications such as hypoglycemia or ketosis are more likely to occur during times of illness, contributing to more frequent emergency department visits and hospital admissions. Sick day management is often challenging for families, requiring advanced insulin dose calculations and problem- solving, using knowledge that is less frequently used. The goal of this study is to develop an educational intervention on sick day management for patients and families with Type 1 Diabetes, and evaluate its impact on patient satisfaction, knowledge retention and behavior change.

Methods: Our study population includes patients with Type 1 Diabetes followed at our centre, aged 0-18 years and diagnosed for >/= 1 year. We assessed baseline knowledge of sick day management among patients/parents via a pre-test questionnaire, which was developed using a standardized diabetes knowledge assessment tool. Our educational intervention will adhere to the 2019 Ontario Pediatric Diabetes Network provincial guidelines for sick day management, and will be delivered at routine clinic visits, with a didactic and interactive component. Satisfaction and knowledge retention will be assessed via questionnaire at 3-month clinic visit post educational intervention. Behavior change will be assessed via questionnaire at 6 months, in addition to chart review to identify phone calls to diabetes team, emergency department visits and DKA episodes.

Results: Of the 24 pretests collected to date, results indicate general knowledge of sick day management is high. Knowledge gaps include whether to check for ketones with a normal blood sugar, optimal fluid intake during illness and whether to skip rapid insulin at meals when unwell (20-25% answered correctly). Results suggest that our intervention must not only address general knowledge but also the practical application of sick-day calculations, decision-making, and target areas of common misunderstanding. Our next step is to develop and pilot our educational intervention.

Poster Abstract 13
Successful Management of T1D in an Infant with Insulin Pump Therapy and Diluted Insulin

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Type 1 diabetes (T1D) rarely presents in infancy, but when it does, there are unique challenges and considerations in the management of these very young patients. Factors contributing to this include: unpredictable eating behaviours; unknown carbohydrate intake if breast feeding; and extreme insulin sensitivity. Our current approach to T1D management, including indications for continuous glucose monitoring (CGM) and continuous subcutaneous insulin infusion (CSII), are based on research and experience with older children. Although some reports suggest benefit of CSII in infants and very young children with T1D, there are currently no guidelines that address the specific challenges of CSII initiation and therapy in this population. We present the case of an infant diagnosed with T1D at 8 months of age. As evidenced by her CGM data, initial management with multiple daily injections (daily insulin glargine and mealtime diluted (U10) insulin lispro) was suboptimal, with significant evening (7pm - midnight) and mid-day hyperglycemia, as well as difficulties correcting for high blood glucose due to very small insulin dose requirements. At 14-months, she was started on CSII therapy using diluted (U25) insulin lispro. We discuss the positive impact this change in management had on our patient's glycemic control, with illustrative CGM data. This data is also used to discuss the benefits of early CGM in identifying the unique insulin needs that may be observed in infants and very young children with T1D. Finally, we present a practical and safe method of using diluted insulin in CSII, including the rationale for and preparation of a U25 insulin dilution, and an approach to initial pump settings and adjustments.

Poster Abstract 14
Rickets Complicating Pseudohypoaldosteronism Type 1

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Pseudohypoaldosteronism type 1 (PHA1) is an electrolyte disorder characterized by resistance to aldosterone. An association between PHA1 and rickets has not been described.

We present non-identical twins, born 32+5 weeks. Antenatal and birth history were unremarkable. Hyponatremia, hyperkalemia and metabolic acidosis were noted on day three of life. The clinical diagnosis of PHA was made based on severe salt wasting, hyperkalemia and very high aldosterone levels. Genetic testing confirmed the diagnosis showing a homozygous mutation in the SCNN1A gene (systemic form of PHA1). Because of feeding intolerance, a G-tube was placed. The PHA was managed with kayexalate and sodium supplementation. They were discharged at 2 months of age. Na, K and serum calcium were within the reference range. However, 1,25(OH)2D was significantly increased (786pmolL). At one year of age Twin A presented with multiple fractures. Skeletal survey (Twin A) showed multiple fractures, demineralization and signs of rickets. Skeletal survey (twin B) showed rickets, demineralization and bowing of the legs, but no fractures. Both twins had similar blood work results. Twin A showed low ionized calcium (1.19mmol/L) normal total calcium (2.41mmol/L) and phosphate (1.44mmol/L). PTH was elevated (128ng/L) 25(OH)D normal (67nmol/L) and 1,25(OH)2Dwas elevated (1,146 pmol/L)

A provisional diagnosis of calciopenic rickets was made based on the potential for calcium malabsorption related to the G-tube, secondary hyperparathyroidism and the1,25(OH)2D. They were managed with increased enteral calcium supplementation and alfacalcidol. Over the following year, the secondary hyperparathyroidism, rickets and severe demineralization resolved and there were no further fractures.

To date there are no case reports of psueodohypoaldosteronism and rickets. Mutations in the SCNN1A gene are associated with severe renal sodium wasting and potentially renal calcium wasting as well. The rickets in our cases is calciopenic in nature and likely due to calcium malabsorption but the contribution of renal wasting of calcium needs further study. Given our experience, we recommend monitoring for disturbances in calcium, phosphate metabolism and rickets in infants with PHA.

Poster Abstract 15
Awareness of Primary Care Physician That Lavender Oil Causes Prepubertal Gynecomastia or Premature Thelarche

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Background: Prepubertal gynecomastia and premature thelarche warrant thorough evaluation for the pathological source of estrogens from the testes or adrenal glands and precocious puberty in girls. If serum concentrations of sex steroids and gonadotropins are normal, an exogenous source of estrogens should be considered.

Aim: We present a case series of prepubertal gynecomastia and premature thelarche from 2 different families who have been exposed to lavender products. A survey was also conducted of community pediatricians and family doctors about their awareness and practise around counseling around endocrine disruptors such as lavender and tea tree oil.

Case Presentations: Brothers aged 6 and 5 years presented with bilateral breast development of 2-4 weeks duration. The older sibling had bilateral breast Tanner stage 3 while the younger sibling had left sided breast Tanner stage 2. The genitalia were prepubertal (testes 3cc). Both these children were using Aveeno Lavender lotions on a regular basis. Siblings 7 years old brother and 4 year old sister both presented with six month history of bilateral breast development with tenderness. Both children were bilateral Tanner Stage 2 at time of initial assessment by family doctor which resolved at time of endocrine evaluation. The family was using a lavender oil diffuser (20 drops/day) during the period of breast development. In both families, laboratory evaluations showed normal thyroid function, adrenal profile and gonadotropins in prepubertal range. Both families expressed that were not aware about the impact of lavender products on breast development.

Survey Results: A survey was distributed to greater Toronto area family doctors and pediatricians during the Scarborough Health Network Pediatric Pearls conference (response rate 40.7%). Results showed 86 % of family physicians were not aware about the impact of lavender products while 35% of pediatricians were not aware. Furthermore, only 12% of pediatricians routinely counsel the patients to monitor usage of these products.

Discussion: Estrogenic properties of lavender oil have been well described in the literature. It sold over the counter in many forms, primary care physicians should be aware of this possibility of endocrine disruption by these essential oils and caution parents and patients about its over-usage.

Poster Abstract 16
Vitamin D Deficiency Is Associated With Higher Systolic Blood Pressure in Adolescents with Type 1 Diabetes Compared to Healthy Controls

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Background: Cardiovascular disease is the major cause of mortality in Type 1 Diabetes (T1D), but underlying pathophysiology is not fully understood. In young patients with T1D, blood pressure (BP), arterial stiffness (AS) and endothelial dysfunction have been described as cardiovascular risk factors, beside well known factors such as hypercholesterinemia, BMI and poor glycemic control. There is also some evidence that vitamin D deficiency (VDD) may be pro-atherosclerotic in T1D. The aim of this study is to investigate a possible association between VDD and blood pressure as well as other cardiovascular risk predictors in adolescents with T1D.

Methods: 106 adolescent patients with T1D between 11-16 years where recruited as part of the Adolescent Type 1 Diabetes Cardio-Renal Interventional Trial (AdDIT) at The Hospital for Sick Children, Toronto as well as 106 healthy controls. Biochemical measures included HbA1c, creatinine, urinary albumin-creatinine ratio, cholesterol, 25-OHD, calcium, phosphate and PTH. Comprehensive cardiovascular assessment included BP, pulse wave velocity as marker of AS, flow-mediated dilation as maker of endothelial function and global longitudinal strain as a marker of left ventricular function. 25-OHD level above 50nmol/l was defined as sufficient, below as deficient. Descriptive statistics and ANOVA with pairwise comparison was performed and results adjusted for age, sex, ethnicity, BMI and cholesterol.

Results: Mean age, sex and ethnicity distribution were similar in both groups. 25- OHD was lower in T1D (mean 56.9 versus 88.4nmol/l, $p < 0.001$). 53% of T1D versus 31% of controls had low 25-OHD levels. Calcium was lower (2.39 versus 2.42mmol/l, $p=0.02$) and PTH higher (36.4 versus 24.1ng/l, $p<0.001$) in T1D. Higher systolic and diastolic BP (114.7 versus 111.4mmHg, $p=0.027$ and 66.7 versus 63.6mmHg, $p<0.001$), higher BMI (22.1 versus 20.8kg/m2, $p=0.12$) and higher cholesterol level (4.3 versus 4.1mmol/l, $p=0.025$) were found in T1D. Other cardiovascular results will be presented. Using 2 way ANOVA, patients with T1D and VDD had significantly higher systolic BP when compared with those that had sufficient vitamin D and when compared with controls, even after adjusting for potential confounders.

Conclusion: VDD is more prevalent in T1D when compared with controls. VDD is associated with higher systolic BP in adolescents with T1D.

Poster Abstract 17
Growth Hormone Deficiency in Patient with Megalencephaly Capillary Malformation Syndrome (MCAP)

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Megalencephaly capillary malformation syndrome (MCAP) is a rare overgrowth disorder characterized by brain abnormalities including progressive ventriculomegaly and polymicrogyria, somatic and cerebral asymmetry, cutaneous vascular malformations, digit anomalies and developmental delay. It is known to be associated with mosaic mutations in PIK3CA. Cases are usually sporadic. Patients with MCAP were noticed to have poor linear growth with case reports of associated hypoglycemia and confirmed growth hormone (GH) deficiency. The proposed etiology of GH deficiency includes hypopituitarism secondary to brain anomalies or hydrocephalus, down-regulation of GH and IGF-1 production and augmented clearance of GH, IGF-1, and IGFBP-3.

We here report a 5-year-old boy with a complex medical history who was born late preterm, had macrosomia, lax skin, dislocated left hip, brachydactyly, small ears, extensive cutaneous vascular stains as well as hypotonia. During infancy, he developed hydrocephalus requiring ventriculostomy. He also has developmental delay, hemihypertrophy and poor linear growth. Genetic testing confirmed the diagnosis of MCAP syndrome, due to the presence of a low-level mosaic mutation in PIK3CA, which is a heterozygous pathogenic variant in PIK3CA gene (p.G914R). He had a fluctuating growth velocity. He had no history of hypoglycemia. His IGF-1 level was low normal at 18 ug/L (range: 18-176). He was proven to be GH deficient via stimulation test with a peak of 1.69 ug/L in glucagon test and 3.07 ug/L in arginine test (cut off: > 5.6 ug/L). Other pituitary hormone screening was unremarkable. The pituitary gland was normal in MRI. We opted to start our patient
on recombinant growth hormone therapy when his growth velocity decreased to 2.3 cm/year and his height z-score was -2.32.

Initiation of GH therapy in such patients can be a challenging decision due to the risk of aggravating hemihypertrophy as well as the theoretical risk of tumorigenesis. Patients with MCAP are at higher risk of developing malignancies including Wilms tumor and meningiomas. To date, data is lacking consensus regarding GH initiation in such patients. Close follow up was organized with different specialties to monitor for side effects.

Poster Abstract 18
Can Rickets Be Transient Without Treatment?

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An 18-month-old boy was referred to the Bone Clinic for suspicion of rickets. A chest X-ray initially done to rule out pneumonia revealed incidental findings of widening of humeral metaphysis and cupping of anterior costo-chondral junctions suggesting rickets. Past medical history was non-contributory: born at term with normal weight/height (50th percentile) and psychomotor development was appropriate for age. He had recurrent asthma-like episodes but no bone pain or fractures. He had a normal diet and was supplemented with 400IU/d of choleclaciferol. Physical examination revealed decreased height velocity with a height at the 3rd percentile, below his family target (172 cm +/- 8.5, 25th percentile), bilateral femoral and tibial bowing, rachitic rosary, enlarged wrist metaphyses and non-ossified anterior fontanelle. Initial laboratory findings revealed eucalcemia, euphosphatemia with suppressed parathyroid hormone (PTH), and normal calciuria and tubular reabsorption of phosphate. Low serum ALP activity for his age suggested hypophosphatasia. However, serum levels of PTH and ALP normalized after one month of follow-up. 25 (OH) vitamin D status was sufficient. Serum levels of 1,25 (OH) vitamin D, vitamin B6 and FGF23 were all in the normal range. A rickets sequencing panel revealed a heterozygous variant of unknown significance in the DMP1 gene, known to be pathogenic only in the homozygous state. A metaphyseal dysplasia genetic panel was then ordered revealing a heterozygous variant of unknown significance in the MMP13. MMP13 gene encodes a matrix metalloproteinases specially expressed in hypertrophic chondrocytes and osteoblasts of growing bones and has an important role in bone formation and growth. Heterozygous MMP13 pathogenic variants are implicated in metaphyseal anadysplasia (MAD) [Lausch E, 2009] which presents with short stature, rhizomelic micromelia, varus deformity of lower extremitie and severe metaphyseal changes. The prognosis is favourable with the dysplasia (and radiological findings) lasting exclusively during growth whereas final adult height may be below the expected genetic potential [Bonafé L et al, 2013; Dong Li, 2015]. We report a novel variant in MMP13 associated with MAD in a young boy that presented with a phenotype initially mistaken for rickets. Genetic testing for skeletal dysplasias should be considered in children presenting metaphyseal anomalies mimicking rickets that have an aberrant clinical course.

Poster Abstract 19
Reversal of Precocious Puberty with Letrozole in McCune-Albright Syndrome

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Introduction: McCune-Albright syndrome is characterized by cafe-au-lait macules, polyostotic fibrous dysplasia and precocious puberty due to recurrent estrogen secreting ovarian cysts.
Case: This is the case of a 3 year old girl who presented following an initial episode of painless vaginal bleeding lasting two days and transient breast budding. There was no significant past medical history and no family history of precocious puberty. On exam, her height and weight were both at the 90th percentile. Breast development was Tanner 1, genital examination revealed pubic hair Tanner 1, and no hyper-estrogenization of the vaginal mucosa. There was extensive patchy pigmentation in dermatomal distributions on the right upper back and lumbar region. Investigations: bone age 5 years, estradiol 257pmol/L, LH < 0.2IU/L and FSH 1.3IU/L. A clinical diagnosis of McCune-Albright syndrome was made. Mutation analysis of the GNAS gene in the blood did not identify any pathogenic variants. The family has not consented to further testing with skin biopsy. At age 4.5 years, there was a second episode of vaginal bleeding with breast development (Tanner stage 2-3). Her annual growth velocity was 10 cm. Investigations: estradiol 313pmol/L, LH < 0.2IU/L, FSH < 0.2IU/L, bone age 6 years 10 months to 7 years 10 months, pelvic ultrasound showed a right ovarian cyst (3.4 x 2.8 x 1.6 cm) likely representing an estrogen-secreting cyst. She was started on a trial of the aromatase inhibitor Letrozole. Less than two months later, her breast development had regressed and she had no further vaginal bleeding. On pelvic ultrasound, the ovarian cyst had resolved. She has continued on Letrozole 2.5mg daily (0.1mg/kg) with no adverse effects and no further pubertal development.

Conclusion: In summary, this is a case of a 3 year old girl with a clinical diagnosis of McCune-Albright syndrome who presented with precocious puberty secondary to an estrogen-secreting ovarian cyst. Treatment with Letrozole has been successful in resolving the cyst and clinical symptoms. The duration of therapy will be predicated on chronologic age and bone age with the aim of optimizing final adult height.

Poster Abstract 20
A Curious Case of Hypertension

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A 4-year-old previously well girl presented to the emergency department with a 2- month history of 2.5kg weight loss, fatigue, diaphoresis and intermittent bilateral lower limb pain. She was admitted due to persistence of symptoms and a new erythematous rash, hyperreflexia and hypertension (144/63 mmHg). Assessment for rheumatologic and malignant conditions was negative. A detailed history revealed that a home sphygmomanometer had broken, spilling mercury onto the carpet, which was then vacuumed. Continued use of the vacuum released mercury vapours into the home. Testing confirmed mercury toxicity with elevated serum (24.4 nmol/L [RR <8.6nmol/L]) and urinary (197.9 nmol/L, [RR <20nmol/L]) levels, and 21 days of chelation therapy was initiated with dimercaptosuccinic acid. The clinical course was notable for refractory hypertension that required multiple antihypertensive agents (amlodipine, metoprolol and doxazosin) to achieve normotension. Other investigations revealed elevated plasma catecholamines and metanephrines (norepinephrine 9.4 nmol/L [RR 0.8 – 3.4nom/L]), epinephrine 1.1 nmol/L [RR <0.8 nmol/L], normetanephrine 1.71 nmol/L [RR <0.89nmol/L], metanephrine 0.44 nmol/L [RR <0.49 nmol/L]). Urinary free cortisol was also elevated (584 nmol/L [RR 10 – 160 nmol/d]). These results normalized within three months, coinciding with her becoming normotensive off all medications.

A literature search identified rare reports (n = 6) with elevated catecholamines secondary to mercury toxicity. Mercury toxicity is thought to bind and inactivate S- adenosylmethionine (SAM), a co-enzyme required to convert norepinephrine to epinephrine, leading to elevated catecholamines and metanephrines, findings that can mimic pheochromocytoma. Similar to our patient, biochemical profiles normalized in the published cases within a few weeks up to 2 – 3 months. Elevated urinary free cortisol has not been reported in previous cases. However, animal studies have reported that mercury can affect cortisol production, including one study that described elevated serum cortisol with acute mercury toxicity in rainbow fish.

Conclusion: Although rare, mercury toxicity can cause refractory hypertension and pheochromocytoma-like biochemical profiles. Despite its favourable prognosis, clinicians should be aware that symptoms and laboratory abnormalities may persist up to 3 months after completion of chelation therapy.

Poster Abstract 21
The Development of Standard Operating Procedures (Sops) for the Use of the Edmonton Obesity Staging System for Pediatrics (Eoss-P) in Community Practice in the Management of Patients With Obesity.

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Pediatric obesity is a chronic condition that requires ongoing interdisciplinary management. In 2016, Hadjiyannakis et al published a paper in Pediatric and Child Health proposing a modification of the Edmonton Obesity Staging System to be tailored towards pediatric care. The Edmonton Obesity Staging System for Pediatrics (EOSS-P) is a clinical staging system developed to assist healthcare professionals in the assessment of health risk in children with obesity and identify management needs. The primary objective of this research is to develop standard operating procedures (SOPs) for the assessment and management of pediatric obesity in primary care practice.

The Delphi method was used in this study to aggregate data collected from stakeholder communities in the development of the SOPs. Key stakeholders were identified based on authorship of the original EOSS-P framework paper published in 2016. These individuals were integral to the development of the EOSS-P framework and are a diverse team of multidisciplinary health professionals. The peripheral stakeholder members were identified through the Canadian Pediatric Weight Management Registry (CanPWR) as well as the Ontario Pediatric Bariatric Network. These individuals work directly with or very closely in pediatric weight management and their input will be from direct observation and experience.

As the management of pediatric obesity is multidisciplinary in nature across many allied health professionals, the objective of our research will be to engage individuals from backgrounds that align most directly with the objectives of the EOSS-P – physicians, including Family Physicians, Pediatricians and Pediatric Endocrinologists; psychologists; social workers; dietitians; and exercise specialists – in the development of SOPs for pediatric obesity management. As rates of obesity continue to rise and management needs become increasingly complex, these guidelines and their use in primary care practice will assist practitioners in the overall care of this important patient population. There is opportunity to not only impact local communities in Ontario but to reach national and international communities as well.

Poster Abstract 22
Large-Scale Genome-Wide Association Study for Vitamin D Levels and Their Effect on Risk of Type 1 Diabetes and Autism Spectrum Disorders

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Objective: To increase our understanding of the genetic determinants of vitamin D levels by undertaking a genome-wide association study (GWAS) of serum 25 hydroxyvitamin D (25OHD) levels and test the effect of these genetic determinants on risk of type 1 diabetes and autism spectrum disorders (ASD).
Methods: We conducted a GWAS using imputed genotype data from 401,460 white British individuals from UK Biobank with available 25OHD levels. After quality control, we retained single nucleotide polymorphisms (SNPs) with minor allele frequency (MAF) > 0.1%, and imputation quality score > 0.3 from the autosomes and the X chromosome. We performed a linear mixed model GWAS on standardized log-transformed 25OHD levels, adjusting for age, sex, season of measurement and vitamin D supplementation. We next combined these results with results from a previous GWAS meta-analysis including 42,274 Europeans, and performed a conditional analysis to identify independent signals. We then used 57 lead independent common SNPs as instrumental variables in two-sample Mendelian randomization (MR) studies testing whether 25OHD has causal effects on type 1 diabetes and ASD.
Results: The SNP heritability of vitamin D levels in the UK Biobank GWAS was estimated to 16.1%. We observed 138 independent 25OHD-associated SNPs (pre and post-conditioning p-value< 6.6 x10-9) among which 53 had MAF<5%. These SNPs map in 69 distinct loci, among which 63 are novel. The 53 alleles with MAF
<5% conferred an average absolute effect of 0.23 standard deviations on standardized log-transformed 25OHD, compared to 0.03 of the 85 alleles with MAF

>5%. Using 69 lead independent common SNPs in separate two-sample MR studies, we found no evidence of a causal association between low 25OHD levels and type 1 diabetes or ASD (inverse-variance weighted MR OR=1.09, 95% CI: 0.86-1.40 and OR=0.98 , 95% CI: 0.85-1.13 per 1 SD decrease in standardized log–transformed 25OHD, respectively). Results were similar in sensitivity analyses.

Conclusions: Through the largest GWAS of 25OHD levels to date, we identified 63 novel 25OHD associated loci. MR studies did not provide evidence supporting a causal association of low 25OHD with risk of type 1 diabetes or ASD.

Poster Abstract 23
Lessons Learned Regarding "Transient" Hyperinsulinism: A Case of Diazoxide Responsive Hyperinsulinism Secondary to abcc8 Mutation

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Our patient was born at term following healthy pregnancy. Birth weight 3.2 kilograms. No gestational diabetes or perinatal stress. The infant was jittery on second day of life with a venous blood glucose of 1.7mmol/L and insulin level of 55pmol/L. Her hypoglycemia was attributed to transient hyperinsulinism, and she was weaned off glucagon and polycal. She was discharged home within 48hours of weaning.

She presented to our centre at 6 months with tonic/clonic seizure, BG of 1.6mmol/L and insulin level 43pmol/L. Parents noted episodes of decreased tone with sweating and lethargy since birth. She required IV Dextrose, followed by Diazoxide and Octreotide due to ongoing hypoglycemia. Genetic testing revealed a mutation in ABCC8 gene: c.4306C>T; p.Arg1436*, heterozygous, paternally inherited. Due to her mutation, we attempted to wean Diazoxide however it was resumed due to worsening hypoglycemia. Maximal treatment to maintain euglycemia included Diazoxide 20mg/kg/day, Octreotide 24mcg daily and GIR 10mg/kg/min. L-DOPA scan revealed 9mm focal lesion in the tail of the pancreas which was removed operatively. This resolved her hypoglycemia, and Diazoxide and Octretide were discontinued.

Discussion:

Although inactivating mutations of the K-ATP channel are commonly associated with diazoxide unresponsive hyperinsulinism, there is a spectrum of clinical presentation. In focal lesions, this may be due to therapeutic effect on normal islet cells and/or partial response of K-ATP channels within the lesion. Our case reinforces the importance of genetic testing, and imaging for paternally inherited defects. Our case also suggests the need for a trial of fasting in cases of suspected "transient" hyperinsulinism, particularly with no clear precipitant.

Poster Abstract 24
MYRF Variant as a Cause of 46,XY Disorder of Sexual Development

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Introduction: The myelin regulatory factor gene (MYRF) encodes a transcription factor that is widely expressed. There is increasing evidence that heterozygous loss-of-function variants in MYRF can lead to abnormal development of the heart, genitourinary tract, diaphragm, and lungs. A novel syndrome has recently been described.

Case presentation: We report a 3-year-old, 46,XY, boy with nonconsanguineous parents of Chinese descent. The patient had an antenatal diagnosis of aortic arch hypoplasia, coarctation with bicuspid aortic valve and multiple muscular ventricular septal defects. At birth, he presented with bilateral intraabdominal testes, hypospadias, a single perineal opening, non-fused labioscrotal folds. He also had no Mullerian structures on urogenitogram. Due to the cardiac and urogenital findings, whole-exome sequencing was performed, and showed a heterozygous variant for c.1702del (p.Asp568Metfs*37) in the MYRF gene. This variant is predicted to cause loss of normal protein function and classified as likely pathogenic. Parental testing is pending.

Conclusion: Heterozygous loss of function variants in MYRF have recently been shown to cause a constellation of abnormalities that is now described as Cardiac- Urogenital syndrome (MIM #618280). Other phenotypes identified in these subjects include scimitar syndrome, pulmonary hypoplasia, congenital diaphragmatic hernia, cleft spleen, thymic involution, and thyroid fibrosis. We present a case of a child who was identified as having this syndrome based on whole exome sequencing.
Including our patient, there are now 17 reported cases of individuals carrying variants in MYRF with structural birth defects. Consideration should be given to screening individuals with cardiac and urogenital defects for pathogenic variants in MYRF given its future clinical implication, as those with mutation in this gene have been shown to develop developmental delay and intellectual disability in the future.

Poster Abstract 25
Case Report of a 14 Year Old Female with Primary Burkitt Lymphoma of the Thyroid Gland

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Lymphomas of the thyroid gland are rare, accounting for 1-2% of all thyroid malignancies. Of these, Burkitt lymphomas account for likely less than 1%. This case report describes a 14-year-old girl who presented with a rapidly enlarging goitre, asymptomatic hyperthyroidism, and progressive tracheal compression secondary to primary thyroid Burkitt lymphoma (PTBL). She initially presented with an asymptomatic goiter and suppressed thyroid-stimulating hormone (TSH) with mildly elevated free T3 and free T4, positive anti-TPO antibodies and a thyroid ultrasound and uptake scan confirming thyroiditis. Within the next 3 weeks the neck mass rapidly grew with new biochemical hypothyroidism and was started on Levothyroxine. In spite of treatment, the mass continued to enlarge, causing progressive dysphagia and dyspnea. An urgent CT angiogram showed a heterogeneous thyroidal mass with mediastinal extension, encircling and shifting the trachea and esophagus. A second mass was identified in the pelvis. A core needle biopsy of the thyroid mass was consistent with an aggressive, mature B cell lymphoma; cytogenetic analysis was most consistent with Burkitt lymphoma, stage III. Following definitive pathologic diagnosis, she was started on induction chemotherapy for Group B Burkitt's lymphoma according to ANHL1131 with an excellent response. Her hypothyroidism, which is suspected to be due to thyroid destruction from the lymphoma rather than solely autoimmunity, continues to be managed with Levothyroxine. This case illustrates that although it is rare, lymphoma of the thyroid gland, should be considered in pediatric patients presenting with a rapidly enlarging goitre even in the presence of biochemistry and imaging consistent with thyroiditis.

Poster Abstract 26
A Three-Year Review of the Night Monitor Protocol: A Camp Banting Quality Improvement Initiative

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Objective: To evaluate the impact of the existing night monitor protocol on nocturnal hypoglycemia.

Background: Night monitor (NM) protocols have been in place at diabetes camps nationally as a safety precaution against nocturnal hypoglycemia, however, there is no consensus protocol. Camp Banting's guidelines combine clinical experience with components of other camps' protocols. Campers on injections are treated with Glucerna if their blood glucose (BG) is <6mmol/L at evening snack (ES), midnight, and/or 3am (injections intervention) and are placed on the NM list if <8mmol/L (6-7.9 = Injections at risk). Campers on pumps run a temporary basal at 50% for 2 hours if their BG is <5 mmol/L (pump intervention) and are placed on the NM list if <6mmol/L (5-5.9 = pump at risk)

Methods: Retrospective review of the night monitor data from 3 consecutive summers (2017-2019). Data was analyzed using descriptive statistics.

Results: Of 1335 overnight observations, 477 (36%) were injections and 858 (64%) pumps. For the injections intervention group at ES, 39% on BID and 36% on MDI required an intervention at midnight; 35% of those on BID and 20% on MDI at required an intervention 3am. In the ES injections at risk group, 28% of those on both BID and MDI required an intervention at midnight; 38% of those on BID and 18% on MDI required an intervention at 3am. In the ES pump intervention group, 13% required an intervention at midnight, and of that group, 36% required an intervention 3 am. In the ES pump at risk group, 18% required an intervention at midnight. Of the campers whose bedtime snack BG was "normal" (above at-risk level), 14% on BID, 8% on MDI and 18% on pump were checked clinically and needed intervention at midnight.

Conclusions: The percentage of campers requiring intervention at midnight and 3am is higher than desired with existing protocols. The threshold for routine monitoring should be raised above 8mmol/L(injections) and 6mmol/L (pumps), and the threshold for ES intervention should be raised above 6mmol/L (injections) and 5mmol/L (pumps). Strengthening of the midnight intervention is required. Ongoing quality improvement efforts will modify and re-evaluate the protocol.

Poster Abstract 27
Challenges in the Treatment of GACI Patients: Promising Future Therapeutic Directions

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Mutations in the gene of ectonucleotide pyrophosphatase/phosphodiesterase 1 (ENPP1) have been linked to generalized arterial calcification of infancy (GACI), a rare autosomal recessive disorder characterized by extensive medial calcifications of large- and medium-sized muscular arteries. Treatment with bisphosphonates has been reported to improve patient prognosis by reducing arterial calcifications in these patients. ENPP1 mutations have also been found to cause hypophosphatemic rickets through an unknown mechanism. We discuss here the case of a young girl with a homozygous ENPP1 mutation diagnosed with GACI prenatally. She was treated with etidronate disodium in infancy, which allowed resolution of her arterial calcifications by 2 years of age. The patient was found to have mild hypophosphatemia (0.9-1.1mmol/L) at the age of 2 years, but only showed clinical and biological signs of rickets at the age of 5 years. She was diagnosed with hypophosphatemic rickets and treatment with phosphate supplements and calcitriol was initiated. She responded well without relapse of her arterial calcifications until the age of 8 years, when progression of vascular and aortic valve calcifications was noted. This called for gradual weaning of her rickets treatment due to concerns regarding future surgical challenges associated with worsening arterial hypermineralization. However, phosphate supplements were restarted at the age of 9 years due to increasing bone pain causing significant functional impairment. The delicate balance between therapeutic approaches for GACI and hypophosphatemic rickets will be discussed as well as new upcoming treatment modalities.

Poster Abstract 28
The Conundrum with Early Diagnosis of 46, XY Disorder of Sexual Development

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Introduction: 46XY disorder of sexual development (DSD) may occur for a number of reasons, including a defect in gonadal development, androgen production, or androgen action. Typically, 46XY DSD presents with variable degrees of undervirilization and a molecular diagnosis is determined in about 35% of cases. We present a case of antenatally diagnosed 46XY DSD due to a SRY gene mutation.

Case Presentation: A gay couple achieved this pregnancy by letrozole and intrauterine insemination. The biological father was healthy. The biological mother had a complex mental health history. Serologies were protective. Non-invasive prenatal testing showed a male fetus with a low probability of trisomy 13, 18, 21 or sex chromosome aneuploidy. However, fetal ultrasound at 21 weeks' gestation showed female genitalia. Amniocentesis was offered to investigate this discordance. The karyotype showed a normal male complement, 46XY. Thus, a gene panel for 46XY DSD was arranged. This testing identified a mutation in the SRY gene, resulting in an amino acid substitution at position 113 from alanine to valine. This amino acid is highly conserved, and this mutation is likely pathogenic. The pregnancy progressed normally with spontaneous labor at 40 weeks' gestation. Due to meconium and non-reassuring fetal heart rate, the newborn was delivered by urgent caesarean section. The newborn required brief respiratory support. The birth weight was 3959 g. Examination revealed normal female genitalia and the neonate was sex assigned female. At two weeks of age, hormonal testing showed: LH 3.8 IU/L, FSH 20.9 IU/L, and testosterone 0.67 nmol/L. At six weeks of age, abdominal ultrasound demonstrated normal kidneys and bladder, a normal uterus, and no gonads. The multi-disciplinary DSD team suspected the gonads were streak and too small for visualization, so additional imaging may be required. Nonetheless, prophylactic gonadectomy was recommended because of the risk of gonadal germ cell tumors.

Conclusion: The SRY gene is critical for testis determination. Inactivating mutations in the SRY gene account for 10-15% of 46XY DSD cases. Adolescent amenorrhea is a frequent presentation, but prenatal diagnosis is an increasing possibility. Germ cell tumors commonly occur in patients with 46XY gonadal dysgenesis but the timing of gonadectomy remains unclear.

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