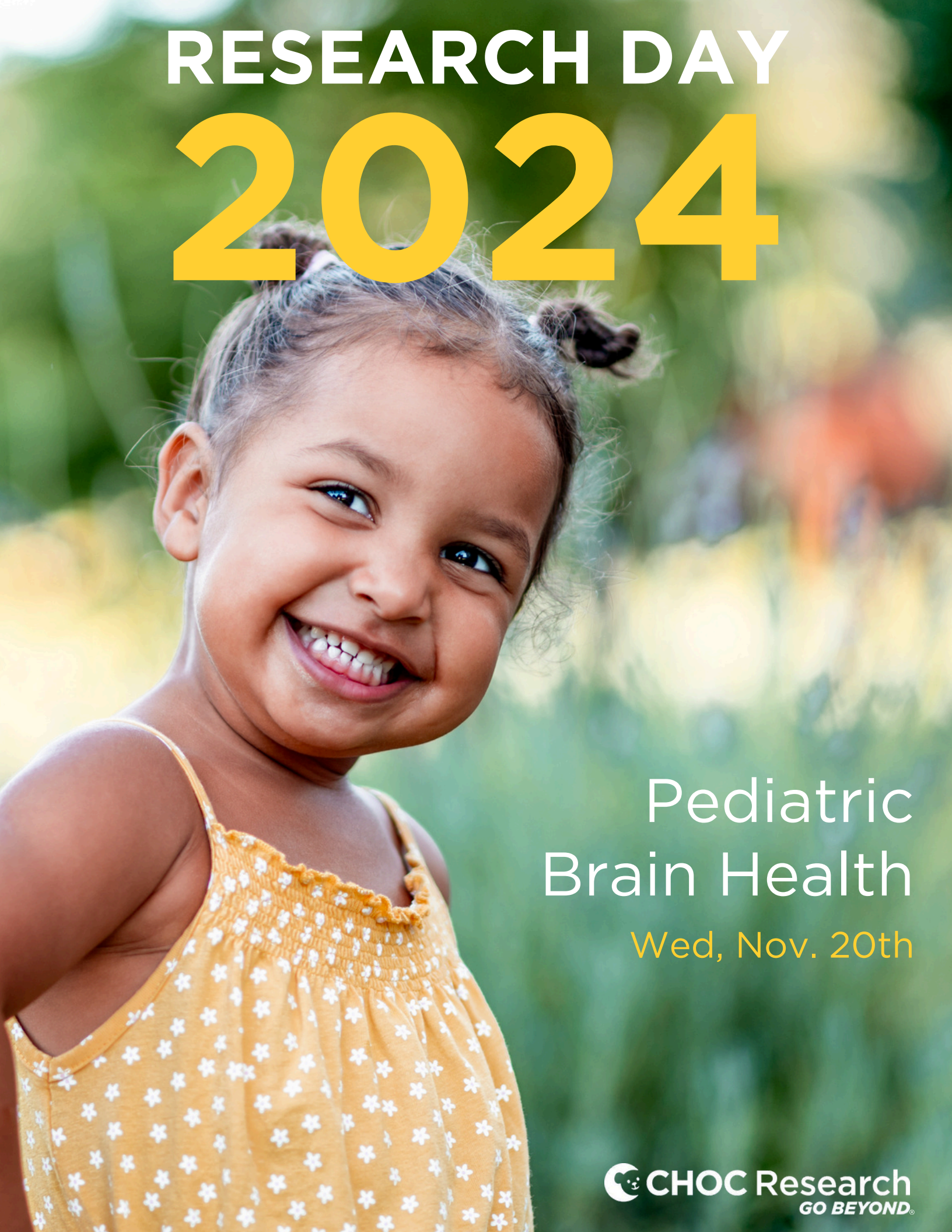

RESEARCH DAY
2024
Pediatric
Brain Health
Wed, Nov. 20th
CHOC Research
GO BEYOND®

A Message from Kimberly Chavalas Cripe

President & Chief Executive Officer, CHOC

As President and CEO, I have long believed that research is pivotal to CHOC's mission and to our vision to be the leading destination for children's health by providing exceptional and innovative care. I frequently share with people that the use of the word "the" in our vision statement is intentional. We do not aspire to be "a" leading destination for children's health, rather, "the" leading pediatric healthcare destination for children and families across the country and around the world. Investment in research is essential to realizing our vision.

On CHOC's 60th anniversary, we come together to celebrate research and the importance of ongoing discovery to the future of pediatric healthcare. I want to extend my sincere thanks to our CHOC researchers and research support teams. I am incredibly grateful for your commitment and perseverance to advancing clinical, basic, and translational science in our never-ending quest to improve the lives of children and families.

I hope you leave feeling supported, energized, and inspired to ask the tough questions that lead to research breakthroughs to secure brighter futures for our children, adolescents, and young adults everywhere.

Be bold in your curiosity and joyful in your pursuit of answers!

Kimberly Chavalas Cripe
President and Chief Executive Officer
CHOC

"On CHOC's 60th anniversary, we come together to celebrate research and the importance of ongoing discovery to the future of pediatric healthcare."

CHOC Research GO BEYOND.

RESEARCH INSTITUTE AT A GLANCE

146 Employees

116 Interns

24 Visiting Scientists

624 Active Studies

89 CHOC Staff with UCI Faculty Appointments

214 Active Principal Investigators

3,524 CHOC Patients Enrolled in Research

1,987 Publications to Date

A Message From Dr. Terence Sanger

Vice President & Chief Scientific Officer, CHOC

On behalf of myself and the entire Research Institute, it is my pleasure to welcome you to Research Day 2024! This year's November celebration continues over a decade of tradition by recognizing the many contributions of research to CHOC's mission – "to nurture, advance and protect the health and well-being of children." While CHOC Research has experienced tremendous growth over the past few years, we remain a family. A family that is committed to asking the hard questions, working together to overcome barriers to "find the answers that each child needs to thrive."

Research Day celebrates the spirit of curiosity and discovery. While we highlight research across the organization, this year's theme focuses on advances in pediatric brain health and the diversity of CHOC's research efforts to find novel approaches to treatments for intractable childhood diseases with devastating effects on the brain.

I want to thank the organizers of today's event; our researchers and support teams who have worked tirelessly to make today possible. I invite each of you to get out of your comfort zone, make new introductions, invite unexpected collaborations, and most of all, celebrate our extraordinary opportunity to leverage research to make life better for the many children and families we are privileged to serve.

"I invite each of you to celebrate our extraordinary opportunity to leverage research to make life better for the many children and families we are privileged to serve."

Go Beyond!

Terence Sanger, MD, PhD
Vice President, Chief Scientific Officer, CHOC
Professor of Electrical Engineering and Computer Science, UCI
Dept. Pediatrics Vice Chair for Research, UCI School of Medicine
Child Neurology and Movement Disorders, CHOC

TABLE OF CONTENTS

Keynote Speaker

Dr. Eric James Benner

Assistant Professor of Pediatrics
Duke University

As a neonatologist, my research interests revolve around improving the survival and quality of life of high-risk neonates cared for in Neonatal Intensive Care Units. My primary interest is perinatal brain injuries impacting both full-term infants and those born prematurely. One of the most common forms of perinatal brain injury involves damage to white matter (myelin). My laboratory has developed models of perinatal brain injury to investigate how the endogenous neural stem cell responds to myelin injury. Our hope is to develop innovative strategies to successfully redirect stem cells into the oligodendrocyte lineage and promote myelination after injury. In order to successfully restore myelination after injury, we want to better understand the molecular mechanisms governing 2 important aspects of myelin development.

First, we must understand the molecular signals that drive neural stem cells to differentiate into oligodendrocytes (oligodendrogenesis) and how brain injury impacts this process. This interest has led my laboratory to investigate intracellular and extracellular changes that occur in the neural stem cell niche following injuries that lead to white matter damage.

Secondly, after stem cell commitment to the oligodendrocyte lineage has occurred, we must understand the ongoing signals from the neural environment that influence oligodendrocyte maturation. For this work, my laboratory has developed an innovative technology to remotely control ion channels non-invasively using magnetic fields. Using this technology, we are developing strategies to alter the activity of targeted neural circuits both in utero as well as postnatally to understand the impact of altered activity on myelin maturation. Members of my laboratory are also currently using this technology to understand how altered temperature-gated channel activity in utero may contribute to birth defects associated with maternal fevers.

Eric James Benner, MD, PhD
Assistant Professor of Pediatrics
Member of Duke Molecular Physiology Institute

AGENDAS

Time	Morning Virtual Sessions	Location
8:00-9:00	Opening Remarks & Keynote Address	Virtual | Streaming in the Wade Center
9:15-10:30	Panel 1 - Perspectives on Neonatal Brain Health through Three Lenses	Virtual | Streaming in the Wade Center
10:45-12:00	Panel 2 - Pediatric Brain Health: Perspectives from Neurosurgery, Oncology, and Neurology	Virtual | Streaming in the Wade Center
Time	**Afternoon In-Person Sessions**	**Location**
12:00-2:00	Poster Session	2nd Floor Bill Holmes Tower
2:00-2:20	Welcome & Research Patient Story	2nd Floor Bill Holmes Tower
2:25-3:45	Podium Poster Presentations	2nd Floor Bill Holmes Tower
3:50-4:00	Awards Ceremony & Closing Remarks	2nd Floor Bill Holmes Tower
4:30-5:30	Networking Reception	2nd Floor Bill Holmes Tower

Kids Cracking Science Quest

Time	Morning Sessions	Location
10:00 - 10:10	Welcome from CHOC Scientists	Seacrest Studios
10:15 - 10:30	Mini-Lecture 1: Sleep - What happened while you were asleep last night?	Seacrest Studios
10:35 - 10:50	Mini-Lecture 2: Mobile Aquarium from the Discovery Cube	Seacrest Studios
11:05 - 11:20	Mini-Lecture 3: Robotics. “Hi, I’m PiMICS, your personal multispectral imaging camera system.”	Seacrest Studios
11:25 - 11:40	Live In-Studio Demonstration, CHOC Scientists	Seacrest Studios
Time	**Afternoon In-Person Sessions**	**Location**
12:30-2:00	Hands-On Experiments & Meet Choco!	2nd Floor Bill Holmes Tower
4:00-4:30	Egg Drop!	2nd Floor Bill Holmes Tower
4:30-4:45	Egg Drop Judging	2nd Floor Bill Holmes Tower

PANEL DESCRIPTIONS

Panel 1: Perspectives on Neonatal Brain Health through Three Lenses

This panel discussion showcases the collaborative efforts of diverse experts from CHOC and UCI specializing in pediatric neurology, neonatology, and neuropsychology to explore opportunities in neonatal brain health research for optimizing pediatric neurodevelopmental outcomes through the lifespan. Each panelist offers a unique perspective rooted in their extensive experience and expertise in both clinical practice and research, contributing to a compelling conversation on innovations in neonatal brain health.

Featured speakers:
Peter Anderson, PhD (CHOC, UCI)
John Crawford, MD, MS (CHOC)
Terrie Inder, MD (CHOC)

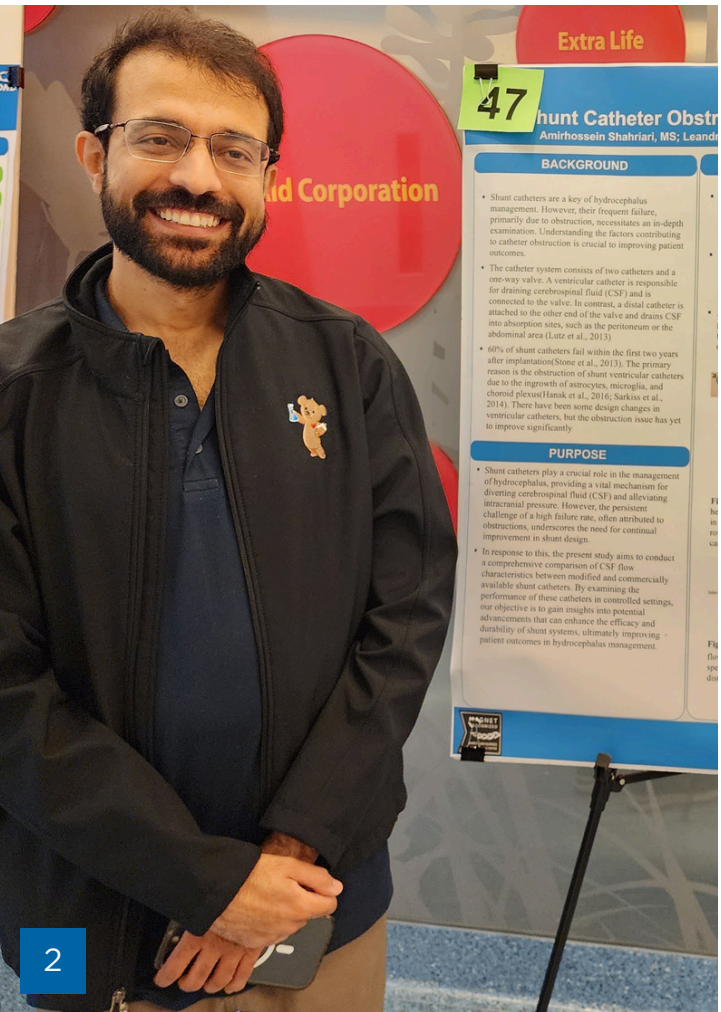
Panel 2: Pediatric Brain Health: Perspectives from Neurosurgery, Oncology, and Neurology

This panel brings together a diverse group of experts from CHOC specializing in pediatric neurology, neurosurgery, and neuro-oncology to explore various aspects of brain health in children. Each panelist offers a unique perspective based on their extensive experience and research, contributing to a broad-ranging conversation on neurological and brain disorders, treatments, and innovations in pediatric care.

Featured speakers:
Maija-Riikka Steenari, MD (CHOC)
Mariko Sato, MD, PhD (CHOC)
Michael G. Muhonen, MD (CHOC)


keep swimming, swi

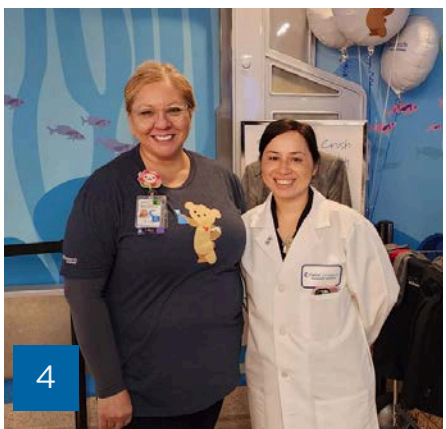
1


Extra Life
47
BACKGROUND
PURPOSE

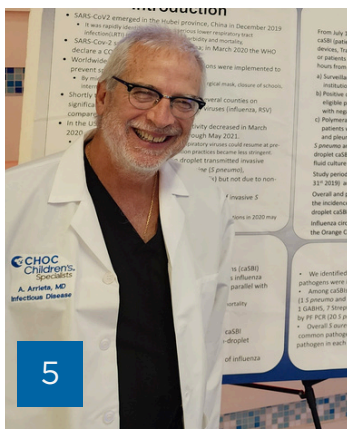
2


CHOC Mi4

3

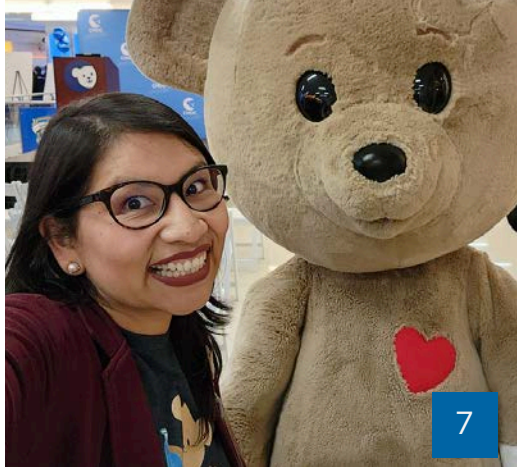


4


CHOC Children's Specialists
A. Arrieta, MD
Infectious Disease

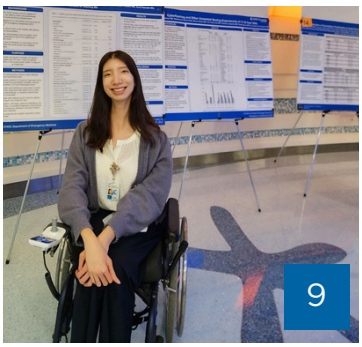
5

6



7

8



9

11

10

RESEARCH DAY 2023

1 Left to Right: Caitlin Calhoun, *Staff Scientist I, PharmD, Research Administration*; Perla Heckman, *Laboratory Technician, Research Administration*; Ofelia Vargas-Shiraishi, *Manager, Clinical Research Programs*; Stephanie Osborne, *Supervisor, Clinical Research Nurse, Research Administration*; Harriet Chang, *Clinical Research Coordinator II, Research Administration*

2 Amirhossein Shahriari, *Laboratory Technician, Research Administration*

3 Left to right: Keri Zabokrtsky, *Manager, Health Sciences Administration*; Shannon Tanaka, *Manager Research Programs, Research Administration*; Sindhu Agnihotri, *Clinical Research Coordinator II, Research Administration*

4 Perla Heckman, *Laboratory Technician, Research Administration*; Nerida Guerrero, *Clinical Research Coordinator II, Research Administration*

5 Antonio C. Arrieta, *MD., Medical Director, Pediatric Infectious Disease*

6 Jennifer Barrows, *Nurse Scientist, Nursing Research & Innovation, PhD, RN*

7 Left to right: Diana Zuniga, *Supervisor, Clinical Research Coordinator, Research Administration*

8 Left to right: Madison Olson, *Research Compliance Specialist, Research Administration*; Krista Larson, *Sr. Research Compliance Specialist, Research Administration*; Amy Thai, *Sr. Research Quality Specialist, Research Administration*

9 Helen Lee, *Clinical Research Coordinator Assistant, Research Administration*

10 Carolina Amaya, *Clinical Research Coordinator I, Research Administration*

11 Pornchai Tirakitsoontorn, *MD, Pediatric Pulmonology, PSF*

PODIUM PRESENTATIONS

2:25-3:45PM

Moderator: Carol Davis-Dao PhD

Title	Speaker
Opening	*Carol Davis-Dao*
Tetrahydrobiopterin Metabolism is Impaired in DARS2 Deficiency	*Alexandra Latini*
A Machine Learning Approach for Predicting Care Transition Risk	*Radha Nagarajan*
Impact of Social Determinants of Health on Development of Children with Down Syndrome	*Minodora Totoiu*
Effects of Antisense Oligonucleotide for Treatment of VCP Disease	*Pallabi Pal*
Pediatric Injuries from Electric-Bicycles: Analyzing the Role of Speed in Injury Patterns	*Zoe Flyer*
High Risk, Low Compliance: Surprising Post-Pandemic Influenza Vaccine Trends in a Children's Hospital	*Brianna Leyden*
Unlocking Relief: Efficacy of PENFS in Alleviating Long COVID Symptoms in Children - A Pilot feasibility study	*Jamie Janchoi*
Nutrient Composition of Defatted Human Milk for Chylothorax and Very Long-Chain Acyl-CoA Dehydrogenase Deficiency	*Kristin Cheng*
Closing Remark	*Carol Davis-Dao*

SPEAKER PROFILE

Dr. Alexandra Latini is an Associate Professor of Biochemistry at the Universidade Federal de Santa Catarina, Brazil, where she coordinates the Laboratory of Bioenergetics and Oxidative Stress (LABOX). For over a decade, LABOX has been dedicated to studying tetrahydrobiopterin (BH4) metabolism and mitochondrial energy production under both physiological and pathological conditions. By combining expertise in genetic metabolic disorders, state-of-the-art cellular and molecular technologies, and insights into cytotoxic pathways, LABOX aims to understand how modulating BH4 production may trigger or treat diseases related to energy metabolism.

Dr. Radha Nagarajan is a Senior Data Scientist at CHOC, where he leads the implementation and deployment of machine learning and artificial intelligence models to improve healthcare outcomes. He collaborates with diverse teams, including the A2B Care Transition Team, to integrate advanced analytics into clinical workflows.

Dr. Minodora Totoiu, is a board-certified child neurologist and the Director of the Down Syndrome Clinic at CHOC. She is the lead investigator in a national multi-center consortium funded by an NICHD grant, which studies the development of children with Down syndrome. Dr. Totoiu is also an Associate Clinical Professor of Pediatrics at UCI, where she actively engages in research on adult Down syndrome.

Dr. Pallabi Pal is a Project Scientist in Dr. Virginia Kimonis's laboratory at UCI. Her research focuses on therapeutic strategies for genetic neuromuscular disorders, particularly hereditary inclusion body myopathies associated with Valosin Containing Protein (VCP) and Heat Shock Protein B8 (HSPB8). Her work aims to elucidate the underlying mechanisms and treatments for these disorders, which lead to muscle atrophy and early demise.

Dr. Zoe Flyer is a General Surgery resident currently completing a research fellowship in pediatric surgery at CHOC. She completed her first three years of residency at Community Memorial Hospital in Ventura, California, before transitioning to CHOC to pursue clinical and basic science research. Dr. Flyer works alongside esteemed faculty and mentors as she strives to specialize in pediatric surgery.

Dr. Brianna Leyden is a second-year Pediatric Hospital Medicine fellow and a part-time hospitalist attending at CHOC. She earned her medical degree at the University of Kansas School of Medicine in Kansas City, KS, and completed her pediatric residency at Phoenix Children's Hospital in Phoenix, AZ. Her specialty interests include inpatient pediatric vaccination trends and improving communication with families.

Jamie Janchoi is a Clinical Research Coordinator in gastroenterology at CHOC Children's Hospital. She holds a background in biochemistry, molecular biology, and public health from UC Davis and leverages this foundation to manage pediatric research projects. Her current interests include improving pediatric and community health through innovative therapies such as percutaneous electrical nerve field stimulation (PENFS), as well as utilizing virtual reality (VR) and artificial intelligence (AI) to enhance health outcomes for children and their families.

Kristin Cheng is a Metabolic Dietitian at CHOC Children's Hospital and a graduate student at Chapman University. She earned her B.S. in Nutritional Sciences and Dietetics from UC Berkeley and completed her dietetic internship at UCSF Medical Center, followed by her pediatric residency at CHOC. Currently, she is part of Dr. John Miklavcic's research group at Chapman University, where she is conducting human milk research.

ABSTRACTS FOR PODIUM PRESENTATIONS

PODIUM PRESENTATION #1

Tetrahydrobiopterin Metabolism is Impaired in DARS2 Deficiency

***Alexandra Latini*[1,2]**, *Luisa Cruz*[1], *Milad Gazanfari*[2], *Wei-Lin Huang*[2], *Thomas Creczynski Pasa*[1], *Alexander Stover*[2], *Philip Schwartz*[2], *Jose Abdenur*[2]

[1]*LABOX, Biochemistry Department, Universidade Federal de Santa Catarina, Florianopolis, SC, Brazil;* [2]*Laboratory for Energy Metabolism, Division of Metabolic Disorders, CHOC Children's Hospital of Orange County, Orange, CA, United States*

Background: Tetrahydrobiopterin (BH4) metabolism has emerged as a promising metabolic pathway that can be positively modulated to inhibit deleterious signaling pathways that may result in cell death.

Aim: Considering that mitochondrial diseases are characterized by energy deficits, oxidative stress and inflammation, this work aimed to investigate whether BH4 metabolism is compromised in the primary mitochondrial disorder, DARS2 deficiency. DARS2 deficiency is characterized by leukoencephalopathy with brainstem and spinal cord involvement, as well as lactic acid elevation assessed by magnetic resonance spectroscopy. Since there is no effective treatment for the disease, we also tested whether enhancing cellular BH4 levels increases mitochondrial function.

Methods: Fibroblasts were established from skin biopsies from two affected siblings (Patient-1: 8y and Patient-2: 4y) and from an asymptomatic brother, who is a non-carrier (Control). Mitochondrial physiology was investigated by assessing cellular respiration and the activities of the mitochondrial complexes. BH4 metabolism was characterized by measuring the gene expression and activity of the main BH4 biosynthetic enzymes. Differential gene expression was assessed by analyzing the transcriptome. Exposure to 100 μM sepiapterin was used to increase intracellular levels of BH4.

Results: DARS2 deficient fibroblasts showed reduced mitochondrial respiration and activities of complexes I and IV. Transcriptome analysis showed downregulation of several mitochondrial proteins involved in energy production. Downregulation of the enzymes involved in BH4 biosynthesis were also downregulated, which were confirmed by assessing *GCH1* and *SPR* gene expression, BH4-link genes. Furthermore, the activity of GTPCH (*GCH1*encoded) was not enhanced when stressed with lipopolysaccharide (24h; 2 mg/mL). Finally, when fibroblasts were exposed to increasing concentrations of sepiapterin for 24h, cellular respiration and ATP generation efficiency were enhanced, leading to increased mitochondrial metabolism with reduced lactate production.

Conclusion: The administration of BH4 may result in a new approach for treating primary and acquired mitochondrial disorders, including DARS2 deficiency.

PODIUM PRESENTATION #2

A Machine Learning Approach for Predicting Care Transition Risk

*Reny Partain, LCSW, MPH, **Radha Nagarajan, PhD**, Wendy N. Gray, PhD, Erin Benekos, FNP-C, Kenneth Grant, MD, Steven Martel, MD, & Michael Weiss, DO*

Background: Bob's Level of Social Support (BLSS) has been historically used to predict pediatric care transition risk. The BLSS assumes an underlying linear structure in its scoring, with pre-defined thresholds categorizing patient risk level into Low Risk (BLSS Score: ≤ 7), Moderate Risk (Score: 8–10) and High-Risk (Score: 11–15). However, as the BLSS is unvalidated, it is unclear to what extent these groupings represent increased risk.

Methods: BLSS data for 495 adolescent and young adult (AYA) patients with chronic health conditions were collected as part of routine care at a children's hospital. A data-driven unsupervised machine learning approach (PAM: partitioning around medoids) was used to group patients into distinct profiles. A chi-square test (a = 0.05) assessed the similarity between the BLSS approach (Low/Moderate/High) and our data-driven clustering approach.

Results: The clustering approach grouped the AYA patients (N = 495) into three groups. Rather than depict risk linearly (Low/Moderate/High), as the BLSS is currently used, results suggest non-linear, qualitatively different profiles. Specifically, patients in the BLSS Moderate group (N = 140) had higher behavioral/mental health risk. The BLSS High group (N = 231) contained patients that were medically-complex/likely requiring lifelong dependent care. Dendrogram from clustering also revealed hierarchical grouping of the patients within each of the risk groups. Chi-squared test revealed significant overlap (p-value < .05) between the approaches.

Conclusion: The proposed study uses a data driven multivariate approach for pediatric care transition risk prediction. The approach determines the risk groups based on patient similarity rather than pre-defined thresholds currently proposed by the BLSS. These subgroups also reflect nuances such as high behavioral/mental health needs seen in the "Moderate Risk" group that need more investigation. Such an understanding may encourage targeted intervention and disease management of care transition subjects.

PODIUM PRESENTATION #3

Impact of Social Determinants of Health on Development of Children with Down Syndrome

Minodora Totoiu, *Christy Hom, Virginia Allhusen, Mark Mapstone, Linda Do, Caitlin McIntyre*

Purpose: Children with Down Syndrome (DS) experience varying abilities in cognitive, language, behavior functioning. Assess the impact of residential location-based child opportunity, as measured by the Child Opportunity Index (COI), on verbal intelligence (KBIT-2 VIQ), cognitive function (RADD-2), and head circumference in children with DS.

Methods: This retrospective cohort study analyzed the relationship between head circumference, cognitive development, and COI, adjusting for age and sex, in 50 children ages 6-17 years with DS, evaluated between May 2022 and October 2023.

Results: The median COI value was 71 distributed across categories (8% low, 28% moderate, 32% high, 32% very high). In 42 patients completing KBIT-2 testing, the raw verbal IQ scores varied with COI and age, showing significant association starting at age 10 ($p=.004$). Scores were lower in lower COI areas (10.9±4.1) compared to higher (20.1±4.9) and very high areas (22.9±4.4). In 33 children able to complete RADD-2 test, average total scores were significantly higher in high, very high COI areas across all ages. Non-verbal KBIT-2 and RADD-2 scores did not vary by COI. Head circumference varied with COI in females, where high COI areas correlated with larger measurements than lower and very high areas ($p<0.05$).

Conclusions: Social determinants of health (SDH) are significantly related to development of verbal intelligence in DS. Consistent with literature, development of language skills and crystallized intelligence appear to be associated with home environment or quality of education. Based upon the relationship between COI and head circumference, females may be more affected by SDH than males.

Funding: PSF TITHE Award # 16979008 and CSO Award # 16979013

PODIUM PRESENTATION #4

Effects of Antisense Oligonucleotide for Treatment of VCP Disease

Pallabi Pal[1], *Michele Carrer*[2], *Olga G. Jaime*[3,4] *Marwan Youssef*[1], *Danae Bosch*[1], *Alyaa Shmara*[1], *Lan Weiss*[1], *Cheng Cheng*[1], *Yasamin Fazeli*[1], *Michael R. Hicks*[3,4], *Paymaan Jafar-nejad*[2], *Virginia Kimonis*[1]
[1]*Division of Genetics and Genomic Medicine, Department of Pediatrics, UC Irvine*
[2]*Ionis Pharmaceuticals, Carlsbad, CA,*
[3]*Department of Physiology and Biophysics, School of Medicine, UC Irvine*
[4]*Sue and Bill Gross Stem Cell Research Center, UC Irvine*

Valosin-containing protein (VCP) disease is an autosomal dominant disease caused by gain-of-function pathogenic variants in the VCP gene. The disease is associated with inclusion body myopathy, with early-onset Paget's disease of the bones, frontotemporal dementia, and familial amyotrophic lateral sclerosis, also known as multisystem proteinopathy 1 (MSP1). There is currently no treatment for this progressive disease. We hypothesize that regulating mutant VCP hyperactivity to normal levels can reduce the disease pathology. We propose this could be achieved through the reduction of expression in VCP with the use of antisense oligonucleotides (ASOs). In this study, we assessed the effect of ASOs specifically targeting the human VCP gene in the patient (R155H) iPSC-derived skeletal muscle progenitor cells (SMPCs) and the transgenic mouse model of VCP disease which overexpresses the humanized VCP gene with the severe A232E mutation. ASO2 significantly reduced mRNA and protein expression in the SMPCs. After establishing the validity of the mouse as a model of the human disease, we treated the VCP A232E mice with ASO2 starting from 6 months of age for 3 months and performed monthly motor tests. ASO2 demonstrated tolerability and showed over 50% knockdown of VCP at the mRNA level in mice treated weekly for 8 weeks. We also found a significant reduction in VCP at the protein level upon treatment with ASO2 as compared to control ASO. We found improvement in the autophagy markers and TDP-43, hallmarks of VCP disease. ASO2-treated VCP A232E mice showed significant improvements in the inverted screen compared to mice treated with control ASO. These results suggest that targeting VCP in SMPCs and mice with the VCP A232E variant could be beneficial in preventing the progression of the myopathy, and holds promise for treatment of patients with VCP MSP1.

PODIUM PRESENTATION #5

Pediatric Injuries from Electric-Bicycles: Analyzing the Role of Speed in Injury Patterns

Zoe E. Flyer DO[a,c] *John Schomberg PhD MPH,*[a,b] *Andreina Giron MD,*[a,e] *Mary Maginas MSN,*[a] *Jeffrey Nahmias MD MHPE*[b]*, Yigit S. Guner MD,*[a,b] *Romeo Ignacio MD,*[d] *Troy Reyna MD,*[a,b] *David Gibbs MD,*[a,b] *Laura F. Goodman MD MPH.*[a,b]

[a]*Division of Pediatric Surgery, Children's Hospital Orange County, Orange, California, USA*
[b]*Department of Surgery, University of California Irvine, Orange, California, USA*
[c]*Department of Surgery, Community Memorial Hospital in Ventura, California, USA*
[d]*Division of Pediatric Surgery, Department of Surgery, Rady Children's Hospital San Diego*
[e]*University of California San Diego School of Medicine, San Diego, CA*
[f]*Department of Surgery, Montefiore Medical Center, New York City, New York, USA*

Background: Electric bicycles (e-bikes) are becoming increasingly popular, offering higher speeds compared to traditional pedal bicycles. Despite their growing use, there is limited data on the epidemiology of e-bike related injuries in the pediatric population. Specifically, previous studies have not adequately explored the injury circumstances regarding e-bikes, particularly concerning loss of control due to speed. This study aims to assess the patterns and outcomes of e-bike injuries in children, hypothesizing that speeds higher than 20 miles per hour (MPH) result in more internal injuries necessitating hospital admission.

Methods: This retrospective cross-sectional study analyzed data from the National Electronic Injury Surveillance System, specifically targeting pediatric ages 0-18 e-bike injuries recorded between 2019 and 2023. We utilized natural language processing techniques to extract narratives from the database, identifying words related to the circumstances of injury, and distinguishing between speed-related incidents vs. non-speed-related incidents. The cohort was divided into two groups based on the identified cause: injuries due to increased speed and injuries attributed to other causes. We then conducted bivariate analyses to compare the characteristics and outcomes between these groups, focusing on the type of injury, its severity, and the need for hospital admission.

Results: The study cohort consisted of 15,121 pediatric patients with injuries related to e-bikes (79.7% males and 71.3% adolescents aged 13-18). Injuries attributed to speed were associated with a higher incidence of head, neck, or facial injuries (49.1% vs 28.7%, $p<0.001$) compared to those resulting from other causes. A greater proportion of children with speed-related injuries sustained internal organ injuries (24.1% vs. 10.4%, $p<0.001$) and were admitted to the hospital (7.3%vs.4.7%, $p<0.001$). Of those injuries specified as "internal" 96.7% were head and neck injuries compared to 3.3% other anatomic sites ($p<0.001$). Over the five-year study period, the frequency of e-bike injuries showed a sharp increase, with 4.18% occurring in 2019 and 49.8% in 2023 ($p<0.001$).

Conclusion: Pediatric e-bike injuries have increased in frequency and can be severe, requiring hospitalization. The findings highlight the risks associated with speeds higher than 20 MPH on e-bikes and the need for targeted safety measures and legislation especially related to head injuries. Future research should focus on the effectiveness of safety interventions, including helmet usage and speed control features on e-bikes.

PODIUM PRESENTATION #6

High Risk, Low Compliance: Surprising Post-Pandemic Influenza Vaccine Trends in a Children's Hospital

Brianna Leyden, MD; *Wendi Gornick, MS, Infection Prevention; Tuan Tran, PharmD; Jasjit Singh, MD,*

Infectious Disease, Children's Hospital Orange County, CA, USA

Background: Post-pandemic, the CDC reports a decline in influenza vaccinations in both children 6 months to 17 years by 6.3% from 2019-2020 to 2022-2023, and in those who take care of them, with hospital healthcare personnel vaccination rates declining from 95.2% to 85.7%.

Purpose/Aims: We describe concerning widespread declines in our institution in inpatient influenza vaccinations, outpatient oncology influenza vaccine rates in those receiving active chemotherapy, and employee vaccinations in the context of local influenza epidemiology, influenza admissions, and reasons for refusal.

Methods: Our institution collected inpatient influenza vaccinations based on calendar years 2019-2023 and influenza admission rates over winters 2018-2019 to 2022-2023. Additionally, employee vaccination rates, including reasons for refusal, were collected 2018-2019 to 2023-2024. Lastly, oncology clinic influenza vaccination rates among the high-risk patients receiving active chemotherapy were collected from Sept-Dec 31 2020-2023.

Results: Although influenza admissions plummeted from 302 in winter 2019-2020 to 0 in winter 2020-2021, these admissions have since rebounded. While inpatient influenza immunizations have continued to decline to a low of 0.0029 vaccines/patient day, they are now outpaced by influenza admissions at 0.0037 admissions/patient day. This trend in decreased vaccines is echoed in oncologic patients on active chemotherapy, with non-medical refusals increasing from 5% to 16%, and in employees, with refusals increasing from 1.9% to 5.3%. Although employee rates for medical exemption are consistent, non-medical reasons for refusals have increased 6-fold.

Conclusions: We saw a decline in influenza vaccine rates for hospitalized patients, immunocompromised outpatient oncology patients on active chemotherapy, and employees, despite increased influenza burden in the community post-pandemic. The latter were almost exclusively due to non-medical reasons for refusal. Further study needs to be done to elucidate post-pandemic vaccine trends and identify missed opportunities for immunization, particularly in high-risk patients and their contacts, with emphasis on bolstering influenza vaccine confidence in not only patients, but staff.

PODIUM PRESENTATION #7

Unlocking Relief: Efficacy of PENFS in Alleviating Long COVID Symptoms in Children - A Pilot feasibility study

Jamie Janchoi[1], *Grace Mucci, Tammy Tran*[1], *Alda Taube*[1], *Ashish Chogle*[1]
Department of Pediatric Gastroenterology, CHOC Children's, Orange, CA

Background: Long COVID presents persistent symptoms like fatigue, cognitive dysfunction, functional disability, anxiety, depression, and gastrointestinal (GI) issues in children. Traditional treatments for chronic GI disorders often fail to address the complexities of Long COVID. Percutaneous electrical nerve field stimulation (PENFS) targets the auricular branch of the vagus nerve and has shown promise in treating GI and non-GI symptoms in similar conditions like irritable bowel syndrome. This study examines the efficacy of PENFS in children with Long COVID.

Methods: Children aged 11-18 years, with Long COVID symptoms persisting for over three months, were enrolled in the CHOC GI clinics. The PENFS device was applied weekly for six weeks. Parents and children completed questionnaires on GI and non-GI symptoms, including fatigue, cognition, functional disability, somatization, quality of life, anxiety, depression, and sleep. Data were collected at baseline, during treatment, and at 1-week and 1-month follow-up.

Results: Fifteen children (10 females, 5 males, from ages 12-18 years). Significant improvements were observed in functional disability, somatic distress, and depressive symptom scores, with reductions from baseline to 1-month follow-up. Abdominal pain frequency and intensity also decreased significantly. Sixty percent of children showed significant improvement in psychomotor functioning and attention. Parent reports showed larger improvements in global health and depressive symptoms compared to child self-reports, suggesting potential perceptual differences.

Conclusions: PENFS significantly improves both GI and non-GI symptoms in children with Long COVID. The device effectively reduced psychomotor functioning, attention, functional disability, somatic distress, and depression, offering a potential therapeutic option for managing Long COVID in pediatric patients. Further studies are needed to assess long-term outcomes and understand parent-child reporting discrepancies better.

Funding organization: CHOC CSO Small Grants Program

PODIUM PRESENTATION #8

Nutrient Composition of Defatted Human Milk for Chylothorax and Very Long-Chain Acyl-CoA Dehydrogenase Deficiency

Kristin Cheng, *RD, CLEC[1,2]; Gina O' Toole, RD, MPH, CLEC, CSPCC[1]; Stephanie Chang, MS, RD, CSPCC[1]; Richard Chang, MD, FACMGG, FACPM[3], John Miklavcic, Ph.D.[4,5]*

[1]Clinical Nutrition and Lactation Department, CHOC; [2]Schmid College of Science and Technology at Chapman University; [3]Division of Metabolic Disorders, CHOC Children's Hospital; [4]Schmid College of Science and Technology, Chapman University; [5]School of Pharmacy, Chapman University

Background: Human milk feeding is the gold standard for infant nutrition but is usually discontinued for six weeks in chylothorax and permanently in severe forms of very long-chain acyl-CoA dehydrogenase deficiency patients to restrict fatty acid intake. Defatted human milk is an alternative option for feeding but may be inadequate for providing essential micronutrients to support growth and development.

Purpose/Aim: The purpose of this study was to evaluate the full nutrient content of defatted human milk.

Methods: Human milk samples were obtained from Mommy's Milk Human Milk Research Biorepository (San Diego, CA) from morning (n=10), afternoon (n=10), and evening (n=10) expressions. Upon receipt, samples were thawed, pooled, and centrifuged (2 °C, 835 x *g*, 15 min) before a syringe connected to a nasogastric tube was used to obtain the aqueous fraction for nutrient analysis (Eurofins Nutrition Analysis Center). Individual nutrients were compared between defatted and control milk groups. Statistical analysis was performed using Wilcoxon Rank Sum Test.

Results: Defatted human milk had lower mean amounts of digestible carbohydrate (7.24$\pm$0.38% vs. control 7.72$\pm$0.14%), cis-polysaturated fatty acids (0.05$\pm$0.0% vs. control 0.24$\pm$0.01%), cis-monounsaturated fatty acids (0.08$\pm$0.01% vs. control 0.57$\pm$0.02%), total fat as triglycerides (0.23$\pm$0.03% vs. control 1.75$\pm$0.04%), total saturated fat (0.12$\pm$0.01% vs. control 0.83$\pm$0.02%), iodine (0.1$\pm$0.0ppm vs. control 0.17$\pm$0.0ppm), choline (5.5$\pm$0.26 mg/100g vs. control 6$\pm$0.10 mg/100g), retinol (30$\pm$0.0 IU/100g vs. control 168.67$\pm$8.14 IU/100g), total vitamin A (30$\pm$0.0 IU/100g vs. control 168.67$\pm$8.14 IU/100g), pantothenic acid (0.37$\pm$0.01 mg/100g vs. control 0.40$\pm$0.01 mg/100g), folate (0.0067$\pm$0.0 mg/100g vs. control 0.01$\pm$0.0 mg/100g), total tocopherols (0.1$\pm$0.0 mg/100g vs. control 0.49$\pm$0.09 mg/100g), and zinc (1.7$\pm$0.10ppm vs. control 2.07$\pm$0.15ppm), as compared to the control ($p \leq 0.05$).

Conclusions: Defatted human milk had lower amounts of key macro- and micronutrients that support infant growth and development. Results of this study can inform supplementation guidelines for defatted milk in patients with chylothorax and very long-chain acyl-CoA dehydrogenase deficiency.

Funding Sources: CHOC CSO Small Grant Program

Poster Presentation Abstract Index

#	Presenting Author	Poster Title
BASIC and TRANSLATIONAL RESEARCH		
1	Caitlin Calhoun	iPSC-Derived, Human Neural Stem Cell Therapy in the Brains of MPS I Mice
2	Allisandra Rha	Exo-editing: patient-independent genome editing strategy optimization using exogenous DNA
3	Chloe Martin-King	Multiple instance learning for classification of histological features in pediatric inflammatory bowel disease
4	Makenzie Ferguson	Empowering Mothers to Prevent Window Falls in Children: A Campus-Community Partnership
5	Jerry Harb	Exploring Pompe disease: Insights into the natural history of novel Gaac.1826dupA knock-in murine model
6	Tatiana Moreno	Oracle Health Real-World Data (Cerner Real-World Data, CRWD) for Research
7	Wei-Lin Huang	Aminolevulinate/iron exposure elicited Nrf-2-mediated cytoprotection in DARS2 deficient fibroblasts with impaired energy and antioxidant metabolisms
8	Chloe Christensen	Adenine base editing in a murine model of infantile-onset Pompe disease: Preliminary results
9	Lan Weiss	Rescuing lethality in the Homozygote VCPR155H/R155H mouse model provides a robust disease model for testing AAV gene therapy
10	Audrey Kao	Serial Brain MRI Volumetrics and Tractography of Atypical CLN2 patients receiving ICV cerliponase alfa treatment
11	Alexander Stover	iPSC-derived Motor Neurons for Mitochondrial Disease Modeling and Drug Testing
12	Calvin Li	A Tunable Platform Technology for Early Detection of Traumatic Brain Injury
13	Shih-hsin Kan	Improvement of hypertrophic cardiomyopathy in Gaac.1826dupA knock-in murine model with neonatal gene therapy
14	Allen Murray	Identification of the Storage Products and Assessment of Enzyme Replacement Therapy for Pompe Disease
15	Jennifer Barrows	Feasibility of a 12-Week Yoga Intervention for Adolescents with Chronic Health Conditions
16	Jennifer Barrows	Asthma Control Phenotypes Predict COVID-19 Illness Severity in Children
STUDENT RESEARCH		
17	Ethan Rutherford	Smooth Sailing or Rocky Road in Navigating the Ureteral Orifice: Does Preoperative Tamsulosin Improve Successful Ureteroscopy in Children?
18	Ryan Quinn	"Dealers Choice": Univalve Location Has No Effect on Skin Surface Pressures in Long Leg Casts
19	Emily Campi	Mom's developing too: The reciprocal effects of infant sensory reactivity and maternal sensitivity
20	Sara Alshehabi	Predictors of Risk of Subsequent Urinary Tract Infection in a Multicenter Cohort of Patients with Primary Vesicoureteral Reflux: Preliminary Insights on Risk Prediction

21	Jackson Helms	Cast Art Analgesia
22	Gabriela De Lara	Assessment of Bladder and Bowel Dysfunction among Spanish Speakers Presenting to a Pediatric Urology Outpatient Clinic
23	Adam Lee	OPTIMIZING VORICONAZOLE DOSING IN PREMATURE INFANTS WITH PRIMARY CUTANEOUS ASPERGILLOSIS: WHEN LARGE DOSES ARE NECESSARY FOR TINY BABIES
24	Kelly Bauer	Procedural Anxiety in Pediatric Emergency Care: Incidence and Predictors of Nonresponse to Intranasal Midazolam in Children Undergoing Laceration Repair
25	Kevin Kwan	Smartphone Digital Radiograph Review Prior to Transfer to a Tertiary Pediatric Trauma Center: A Picture is Worth 1000 Words
26	Britanny Winckler	Variation in Admission and Treatment Decisions for Previously Immunocompetent Pediatric Patients Presenting to the ED with Febrile Neutropenia
27	Brianna Leyden	Project POOP: Preventing Opioid-Originated Problems, a Quality Improvement Initiative
28	Allen Nguyen	Variation in Urinary Tract Infection Risk by Race/Ethnicity among Patients with Vesicoureteral Reflux: A Multicenter Analysis
29	Vanessa Vargas	Who, why, and how to screen: Community perspectives on early life adversity screening and research in primary care settings
30	Michael Nguyen	Longitudinal Case Study of Sleep Architecture in Patient with Miller-Dieker Syndrome
31	Divya Hadavale	Physical Activity Is Associated With Maternal Metabolic Profile and Inflammation in Pregnant Mexican American Women
32	Sophie Alter	Efficacy of a dietary buttermilk powder enriched in milk fat globule membrane concentrated with gangliosides for treatment of pediatric IBD depends on SNPs in ganglioside metabolism enzymes
33	Angela Martin-Rios	Expanded phenotypes of beta-mannosidosis: a new association with white matter disease
34	Peter Dinh	Age and Sex on Psychosocial Outcomes in Children with Cleft/Craniofacial Differences
CLINICAL and SOCIAL SCIENCES RESEARCH		
35	Shifa Shaikh	Unusual Presentation of Retroperitoneal Teratoma
36	Van Huynh	Incidence and Risk Factors for Hypersensitivity Reactions in Pediatric and Adolescent/young adult Patients Receiving Premedication for Pegaspargase: a Prospective Multicenter Study
37	Carolina Amaya	Identification of Patients Experiencing Child Trafficking in the Pediatric Emergency Department
38	Kellie Bacon	Achieving Fast Assessment and Stroke Triage in Kids (FAST-Kids)
39	Helen Lee	Pediatric Pain and Maladaptive Behavioral Changes Following Laceration Repair Procedures in the Emergency Department
40	Shelby Shelton	Age, Adverse Childhood Experiences, and Healthcare Utilization
41	Shelby Shelton	Implementation of a Child Maltreatment Screening Tool in the Prehospital Setting
42	Zoe Ta-Perez	Climate Anxiety in Young People: at the Generational Crossroads of Environmental and Mental Health
43	Jamie Janchoi	Enhancing Medical Device Care Education via Virtual Reality: A Paradigm Shift
44	Anahita Darabpour	Utility of the Prehospital Pediatric Assessment Triangle

45	Wendy Grey	Comparing Pediatric Care Transition Risk Predictors: BLSS and the TRxANSITION Index™
46	Shasta Erickson	Optimal Agent and Timing of Surgical Antibiotic Prophylaxis and Surgical Site Infection in Pediatric
47	Michelle Hough	Sutured versus Sutureless Closure in Neonatal Gastroschisis: A Two-institution Study
48	Michelle Hough	Deaths due to isolated extremity gunshot wounds in children and young adults
49	Alice Martino	Pre-hospital Tourniquet Use in Adolescent and Pediatric Traumatic Hemorrhage: A National Study
50	April Carlson	Disparities in Access to Neonatal and Pediatric ECMO Services in the United States
51	April Carlson	THE DEVELOPMENT OF A MULTIDISCIPLINARY FETAL CENTER INCREASES INSTITUTIONAL CONGENITAL LUNG MALFORMATIONS (CLM) VOLUME
52	Andreina Giron	Fetal Care Deserts: Disparities in Access to Expert Care for Congenital Anomalies
53	Andreina Giron	Cancer and Trauma Collide: Injury Patterns and Outcomes in the Pediatric Trauma Patient with Cancer
54	Ashley Bone	Distress among parents and caregivers of patients with Anorectal Malformation and Hirschprung Disease
55	Peter Yu	EMS Pediatric Trauma Transport: Do Disparities Exist?
56	Peter Yu	Racial Disparity Among Youth Decedents of Legal Intervention Trauma
57	Christopher Matsko	Thoracic Endovascular Aortic Repair in Pediatric Patients After Blunt Trauma: A National Analysis, 2018-2022
58	John Schomberg	High Volume, Pediatric Trauma Centers are Associated with Improved Mortality in severe Pediatric TBI
59	John Schomberg	Association of Traumatic Abdominal wall Hernias and Hollow Viscus and solid Organ Injury in Pediatric Patients
60	Andreina Urrutia Gonzales	PTSD among patients with Anorectal Malformation and Hirschsprung Disease
61	Andreina Urrutia Gonzales	Development of a Patient and Peer Advocacy Program in Pediatric Colorectal Surgery
62	Zoe Flyer	Ketorolac: Safety and Efficacy in the Management of Pain for Post-Operative Pediatric General Surgery Patients
63	Zhou Chen	Impact of Hospital Volume on Mortality in Pediatric Patients with Severe Blunt Abdominal Trauma
64	Zhou Chen	Variability in EMS Management of Venomous Snake Bites
65	Melanie Patterson	Enhancing Leadership Well-Being: Outcomes of a 4-Day Workweek Pilot
66	Alice Martino	Age and Resource Utilization in Pediatric Falls: a study of the National Trauma Data Bank
67	Virginia Kimonis	Correction of the common splice site variant associated with mitochondrial complex I deficiency caused by recessive mutations in the NUBPL gene
68	Katherine Simon	Relationships between age, REM sleep, and sleep disordered breathing across the first 24 months of life

69	Kathryn Gasperian	What's the plan? A single center's experience navigating through the challenges of newborn screening identified attenuated MPS-I cases
70	Jessica Ardo	Patient Demographics, Medication Adherence and Self-Management as Reported by Caregivers of Children with Epilepsy: An analysis of the baseline CHOC data from the multi-site Epilepsy Adherence in Children and Technology (eACT) study
71	Jessica Ardo	The Family-Based Crisis Intervention-Inpatient at CHOC Mental Health Inpatient Center: Participant demographics and methods
72	Erica Navarro	Effectiveness of FIT Brain, a virtual-based cognitive-behavioral therapy program in addressing long-term psychological distress among childhood cancer survivors.
73	Vanesa Perez	Evaluation of the School Reintegration Program for Youth Recovering from an Acute Crisis: A mixed-methods approach

POSTER #1

iPSC-Derived, Human Neural Stem Cell Therapy in the Brains of MPS I Mice

Caitlin Calhoun[1], Shih-hsin Kan[1], Alexander E. Stover[1], Jerry F. Harb[1], Evelyn Torres[1], Edwin S. Monuki[2], Philip H. Schwartz[1]

[1]CHOC Children's Research Institute, Orange, California, USA; [2]Department of Pathology and Laboratory Medicine and Sue and Bill Gross Stem Cell Research Center, University of California, Irvine, Irvine, California, USA.

MPSI is a severe genetic disease caused by deficient α-L-iduronidase (IDUA) enzyme activity, which impairs degradation of complex glycosaminoglycans (GAGs) and triggers tissue damage and organ dysfunction, including neurodegeneration. Despite existing FDA-approved treatments such as hematopoietic stem cell transplantation (HSCT) and enzyme replacement therapy (ERT), their effectiveness in slowing neurodegeneration is limited due to the inability to penetrate the blood-brain barrier (BBB).

Previously, we successfully generated human neural stem cells (NSCs) through the differentiation of induced pluripotent stem cells (iPSCs) derived from reprogrammed human cord blood cells. These NSCs were further purified using fluorescence-activated cell sorting and then transplanted into neonatal immunocompromised MPS I ($Idua^{-/-}$) mice based on a NOD/SCID/Il2rγ (NSG) background into both lateral ventricles and the cerebellar parenchyma. Eight months post-transplantation, these cells exhibited migration and partial restoration of IDUA activity, resulting in a reduction in β-hexosaminidase levels within the brains of the transplanted MPS I mice.

In this study, we performed comprehensive histopathological analyses to elucidate the distribution of transplanted human NSCs in MPS I mouse brains with human-specific antibody, STEM121. The staining highlighted a widespread migration of transplanted cells, a critical characteristic ensuring broad distribution of IDUA into the $Idua^{-/-}$ brain. Co-staining of STEM121 with GFAP and Olig2 antibodies demonstrated the differentiation of many transplanted human NSCs into astrocytes and oligodendrocytes. Notably, the presence of engrafted cells correlated with a reduction in certain elevated biomarkers in MPS I mouse brains, including CD68 and LAMP1. These findings suggest a promising therapeutic impact of the transplanted cells in addressing the deficiencies associated with MPS I.

Further examinations involving IDUA-overexpressing NSCs will be conducted to evaluate their efficacy in vivo within our platform, paving the way for potential therapeutic applications.

Funding: This work was supported by the California Institute for Regenerative Medicine (CIRM; TR3-05476), Strategic Grant Program, and the Glass Slipper Guild through the CHOC Foundation, Susan Scott Foundation, CHOC CSO Small Grants Program, Advancing Pediatric Stem Cell Research from the Hoag Foundation (9415), and the Campbell Foundation of Caring.

POSTER #2

Exo-editing: patient-independent genome editing strategy optimization using exogenous DNA

Allisandra K. Rha[a], Chloe L. Christensen[a], Perla Andrade-Heckman[a], Jerry F. Harb[a], Max C. Chen[a], Shih-Hsin Kan[a], Raymond Y. Wang[b,c]

[a]Research Institute, Children's Hospital of Orange County, Orange, CA 92868, United States
[b]Division of Metabolic Disorders, Children's Hospital of Orange County Specialists, Orange, CA 92868, United States
[c]Department of Pediatrics, University of California-Irvine School of Medicine, Irvine, CA 92697, United States

The development of genome editing strategies for disease modeling, molecular engineering, and the correction of pathogenic variants has revolutionized medicine and biomedical research in the last decade. Typical genome editing approaches for pathogenic variant correction rely on the derivation of patient cells or genomic installation of targets in cellular or animal models. However, the availability of tissue harboring target genomic variants has precluded or delayed patient access to precision therapeutics. While some genetic diseases have a single or few causative variants, others have hundreds to thousands, necessitating patient-specific therapeutics requiring patient-specific genomic material. Additionally, patient cells can be difficult to transfect or transduce in culture and can delay important therapeutic milestones. Here, we describe the establishment of exo-editing, a platform for the optimization of genome editing strategies using exogenous DNA. Exo-editing streamlines strategy optimization by bypassing the initial need for patient cells and can be used for the evaluation of prime editing (PE) and base editing (BE). While protocols relying on patient cells may take 6-8 weeks, exo-editing can complete optimization within one week, accelerating therapeutic timelines. To optimize genome editing strategies using exo-editing, the genomic variant and ~200bp of surrounding genomic context are cloned to a plasmid that also expresses the sgRNA (BE or PE) or pegRNA (PE). This plasmid, alongside the editing enzyme, and/or other plasmids expressing additional strategy components, is transfected to HEK293-T cells and sustained in culture for four days. Sequencing reveals that editing efficiencies are strategy-dependent and comparable to efficiencies achieved in patient cells or cellular models harboring the equivalent genomic site. With this system, it is now possible to optimize and evaluate editing strategies for most pathogenic variants without requiring patient cells or genomic DNA, test approaches for the installation of specific DNA elements, and identify top strategies for use in disease modeling.

POSTER #3

Multiple instance learning for classification of histological features in pediatric inflammatory bowel disease

Chloe Martin-King[1], Ali Nael[2,3], Louis Ehwerhemuepha[1,4,5], Blake Calvo[1,4], Quinn Gates[1,4], Jamie Janchoi[1], Elisa Ornelas[1], Melissa Perez[6], Andrea Venderby[1,4], John Miklavcic[4,7], Peter Chang[8,9,10], Aaron Sassoon[2], Brian Rubio[11], Ghislaine Barragan[12], Kenneth Grant[6,13]

[1]Research Institute, [2]Department of Pathology Children's Health Orange County (CHOC); [3.]Department of Pathology, University of California-Irvine (UCI) Medical Center; [4]Schmid College of Science and Technology, Chapman University; [5]Department of Statistics, UCI Donald Bren School of Information and Computer Sciences; [6]Department of Gastroenterology and Nutrition, CHOC; [7]School of Pharmacy, Chapman University; [8]Center for Artificial Intelligence in Diagnostic Medicine (CAIDM), UCI; [9]Department of Radiological Sciences, UCI School of Medicine; [10]Department of Computer Science, UCI Donald Bren School of Information and Computer Sciences; [11]College of Natural Sciences and Mathematics, California State University, Fullerton; [12]Department of Neuroscience, Cognition and Behavior, College of Arts and Sciences, University of San Diego; [13]Department of Pediatrics, UCI School of Medicine, Orange, CA, USA

Background: Histopathology from endoscopic biopsies is a standard component of inflammatory bowel disease (IBD) diagnostic evaluation and certain features may be characteristic of either Crohn's disease (CD) or ulcerative colitis (UC)[1–3]. Many IBD studies that utilize whole slide images (WSIs) present promising results[4–7].

Aims: The aim of our study is to assess whether artificial intelligence (AI) can be used to improve pediatric IBD patient outcomes by providing pathologists with accurate detection of abnormal tissue sections in WSIs. A set of AI models were developed for detecting abnormal tissue, as well as important phenotypical features: active inflammation and chronic changes/architectural distortion.

Methods: CHOC IBD patients under 22 years of age, diagnosed endoscopically between 2014 and 2022, were considered for this study. All tissue sections from all slides were scanned. 25 patients were included resulting in a dataset of 1302 tissue section scans from 229 whole slides. Due to the large size of each scan, scans were sub-divided into patches then resized to 128x128 pixels. After removal of uninformative patches, 24,372 patches remained. Three convolutional neural network (CNN) multiple instance learning (MIL) models were developed. The first model distinguishes between normal and abnormal sections. The second and third models indicate whether a tissue section contains active inflammation and chronic changes, respectively.

Results: The abnormal vs. normal classification CNN MIL model achieved an accuracy of 0.84, AUC-ROC of 0.91, and F1-score of 0.79. The accuracy for predicting active inflammation was 0.85, AUC-ROC was 0.92, and F1-score was 0.78. The accuracy for predicting chronic changes/architectural distortion was 0.86, with an AUC-ROC of 0.93, and F1-score of 0.76

Conclusions: The findings resulting from this study are significant primarily because they indicate that there is strong AI-interpretable signal present in endoscopic WSI, even with the necessary, weakly supervised method of MIL.

POSTER #4

Empowering Mothers to Prevent Window Falls in Children: A Campus-Community Partnership

Makenzie Ferguson, Jennifer Barrows

Background: Falls are the leading cause of pediatric injury, and window falls can lead to particularly severe injuries. Children aged 2 to 5 years have the highest risk of window falls, and those younger than 5 are more likely to be hospitalized or die from a window fall-related injury. Window falls can be prevented through strategies such as moving furniture away from windows and installing safety devices. Yet, a gap remains in translating evidence into practice, as many parents remain unaware of the risk or lack resources to modify their home environment. The purpose of this study was to evaluate the effect of a community-based intervention on mothers' window safety knowledge, self-efficacy, and behavior.

Methods: Community health workers (CHW) provided window safety education and resources to low-income mothers during 4-month postpartum home visits. Mothers completed a baseline survey, received educational materials, and were provided with window safety devices. Mothers completed a follow-up survey during the 8-month postpartum visit. CHW participated in a focus group to evaluate implementation.

Results: 146 mothers received the intervention. They had 349 children living at home including 239 children aged ≤5 years. Most identified as White (70%) and/or Hispanic (84%). More than half were Spanish- (41%) or Vietnamese-speaking (10%). 787 window safety devices were distributed. At baseline, 22% falsely believed that window screens could prevent falls compared to 9% post-intervention ($p=.009$). Before education, 84% felt confident teaching others about window safety compared to 100% after ($p<.0001$). The proportion of homes with safeguarded windows increased from 25% to 83% ($p<.0001$). Mothers and CHW expressed interest in additional home safety topics.

Conclusion: A brief educational intervention delivered by CHW empowered mothers to protect their children from window falls and prevent future tragedies. A range of home safety topics could be addressed using this approach to childhood injury prevention.

POSTER #5

Exploring Pompe disease: Insights into the natural history of novel *Gaa*$^{c.1826dupA}$ knock-in murine model

*Jerry F. Harb[1], Shih-hsin Kan[1], Nancy D. Dalton[1], Alejandra Padilla[1], Chloe L. Christensen[1], Allisandra K. Rha[1], Perla Andrade-Heckman[1], Yunghang Chan[1], Evelyn Torres[1], Dwight D. Koeberl[2,3], Raymond Y. Wang[4,5]**

[1]CHOC Children's Research Institute, Orange, California, USA; [2]Division of Medical Genetics, Duke University School of Medicine, Duke University Medical Center, Durham, NC; [3]Department of Molecular Genetics and Metabolism, Duke University Medical Center, Durham, NC; [4]CHOC Children's Specialists, Orange, CA; [5]Department of Pediatrics, University of California-Irvine, Irvine, CA

Background: Pompe disease (PD) arises from a deficiency in lysosomal acid α-glucosidase (GAA), resulting in glycogen buildup within lysosomes, notably in skeletal and cardiac muscles.

Purpose/Aims: In this study, we established and characterized a novel PD murine model, *Gaa*$^{c.1826dupA}$, which parallels human GAA mutation c.1826dupA (p.Y609*). *Gaa*$^{c.1826dupA}$ mice exhibit markedly decreased GAA expression and enzymatic activity compared to wild-type (*Gaa*$^{+/+}$) mice.

Methods: *Gaa*$^{c.1826dupA}$ mice have undergone comprehensive study up to 72 weeks, with planned future assessments. Investigation involved two cohorts, *Gaa*$^{+/+}$ (n=18; 9 ♂, 9 ♀), and *Gaa*$^{c.1826dupA}$ (n=18; 9 ♂, 9 ♀) mice, that underwent catwalk gait analysis, grip strength testing, rotarod performance evaluation, and echocardiography. Commencing at 12 weeks, mice were subjected to these evaluative assays at regular intervals of 12 weeks thereafter.

Results: Premature mortality was not observed in either cohort. Compared to *Gaa*$^{+/+}$ mice, *Gaa*$^{c.1826dupA}$ mice exhibit weight loss from 48 weeks (mean difference: [48wk, -6.5g], [60wk, -8.6g], [72wk, -10.9g]). Gait assessment demonstrates significant differences in temporal and interlimb coordination parameters, including decreased base of support from 12 weeks, and increased max contact at (% of stand time) from 36 weeks. Furthermore, *Gaa*$^{c.1826dupA}$ mice exhibit compromised skeletal muscle strength, with progressive deficits in grip strength beginning at 12 weeks (mean difference: [12wk, -0.14N], [24wk, -0.24N], [36wk, -0.22N] [48wk, -0.28N], [60wk, -0.27N], [72wk, -0.34N]), and impaired coordination determined by rotarod performance from 36 weeks (mean difference: [36wk, -35.6s], [48wk, -83.7s], [60wk, -81.0s]). *Gaa*$^{c.1826dupA}$ mice manifest early-onset and progressive cardiac hypertrophy, with increases in diastolic left ventricular wall thicknesses; interventricular septum, left ventricular posterior wall, and left ventricular mass index observed from 12 weeks.

Conclusions: Altogether, this study illustrates that *Gaa*$^{c.1826dupA}$ murine model recapitulates many aspects of human infantile-onset PD and serves as a valuable model for assessing therapeutic approaches, including genomic correction strategies.

POSTER #6

Oracle Health Real-World Data (Cerner Real-World Data, CRWD) for Research

Tatiana Moreno, Don Wen, Louis Ehwerhemuepha

Children's Hospital of Orange County, Orange, CA, USA

Background: Oracle EHR Real-World Data (formerly known as Cerner Real-World Data, CRWD) serves as a comprehensive, de-identified big data repository derived from multicenter electronic health records. Cerner Corporation has diligently obtained data use agreements and permissions from over 100 U.S. health systems contributing to this extensive and secure clinical database. This rich dataset encompasses information on patient encounters, diagnoses, medications, procedures, vaccinations, clinical results, and social determinants of health. Researchers can leverage this comprehensive resource to analyze health trends across diverse populations.

Methods: Oracle EHR Real-World Data amounts to over a remarkable 1.8 billion patient encounters. These encounters provide a window into real-world patient experiences, allowing for comprehensive longitudinal analyses. Researchers can harness this vast dataset to explore disease trends, evaluate treatment efficacy, and expose disparities in health outcomes.

Results: As of 2024, Oracle EHR Real-World Data remains on an upward trajectory, establishing a robust foundation for evidence-based research. Researchers have skillfully utilized this dataset to investigate sepsis, mental health, precision medicine, and other critical areas in healthcare. The substantial number of patient encounters within this dataset facilitates clinical trials and outcomes research, leading to transformative discoveries.

Discussion: Oracle EHR Real-World Data opens new horizons for research. By fostering collaboration across institutions, researchers can tackle critical inquiries related to health equity, therapeutic approaches, and overall population health. This dataset serves as a centralized platform for informed decision-making and enhanced patient outcomes in a dynamic healthcare landscape.

POSTER #7

Aminolevulinate/iron exposure elicited Nrf-2-mediated cytoprotection in DARS2 deficient fibroblasts with impaired energy and antioxidant metabolisms

Wei-Lin Huang[a], Tuany Eichwald[a,c], Alexander Stover[a], Milad Gazanfari[a], Philip H. Schwartz[a], Alexandra Latini [a,c] , Jose E. Abdenur[a,b]

[a] Division of Metabolic Disorders, CHOC Children's Health Orange County, Orange, California, United States
[b] Department of Pediatrics, University of California Irvine, Orange, California, United States
[c] Laboratório de Bioenergética e Estresse Oxidativo – LABOX, Departamento de Bioquímica, Centro de Ciências Biológicas, Universidade Federal de Santa Catarina, Florianópolis, Santa Catarina, Brazil

Background: Leukoencephalopathy with brain stem and spinal cord involvement and lactate elevation (LBSL) is caused by mutations in the mitochondrial aspartyl-tRNA synthetase gene *DARS2*. Clinical presentation varies from severe infantile to slowly progressive deterioration in adolescents / adults with a distinctive brain MRI. There is no effective treatment.

Aims: To study the effect and mechanism of action of aminolevulinate plus ferrous iron (ALA/Fe) on energy metabolism and antioxidant defenses.

Material & Methods: DARS2 deficient fibroblasts were exposed to ALA/Fe (100 μM/50 μM) for 14 days to evaluate mitochondrial oxygen consumption, glycolysis, respiratory-chain enzyme activities, mitochondrial morphology, reduced and oxidized glutathione (GSH/GSSG), activities of catalase (CAT), glutathione peroxidase and superoxide dismutase (SOD), and production of reactive oxygen species. The activation of Nrf-2 pathway, which controls cell antioxidant defenses, was also investigated using known inhibitors and activators.

Results: Our previous result have shown energy impairment, altered mitochondrial dynamics, and increased oxidative stress in DARS2 deficient cells. ALA/Fe exposure rescued these deficiencies and increased the antioxidant status by enhancing the GSH/GSSG ratio and the activities of SOD and CAT, suggesting the Nrf-2 pathway was activated. Supporting this, the main Nrf-2-regulated downstream proteins, HO-1 and NOQ1 were increased in ALA/Fe-treated cells. Furthermore, the levels of bilirubin, a product of HO-1, was increased, as well as the expression of PGC-1a, a transcription coactivator that plays a central role in the regulation of mitochondrial biogenesis and function. Additionally, mtDNA copy number was also increased in ALA/Fe-treated cells. The use of dexamethasone, a known Nrf-2 inhibitor, blocked the positive effects of ALA/Fe treatment.

Conclusion: We demonstrate that the mitochondrial dysfunction and oxidative stress in DARS2 deficient fibroblasts can be ameliorated by ALA/Fe through the activation of Nrf-2 pathway, providing a better understanding of cellular pathways that can be targeted in the treatment of mitochondrial diseases.

Financial support: This work was supported by Sue and Ralph Stern Family Grant (16984011 & 16984017).

POSTER #8

Adenine base editing in a murine model of infantile-onset Pompe disease: Preliminary results

Christensen, Chloe L.[1]*, Harb, Jerry F.*[1]*, Kan, Shih-Hsin*[1]*, Andrade-Heckman, Perla*[1]*, Rha, Allisandra K.*[1]*, and Wang, Raymond Y*[2,3]

[1]*CHOC Children's Research Institute, Orange, CA 92868;* [2]*Division of Metabolic Disorders, CHOC Children's Specialists, Orange, CA 92868;* [3]*Department of Pediatrics, University of California-Irvine School of Medicine, Irvine, CA 92697*

Infantile-onset Pompe disease (IOPD) manifests in cardiac and skeletal myopathy and, without treatment, leads to death within the first two years of life. Our recent *in vitro* work demonstrated that adenine base editing (ABE) corrects transition variants in *GAA*, which encodes acid α-glucosidase (GAA), a lysosomal enzyme required for glycogen catabolism. We hypothesize that ABE will restore GAA function *in vivo* and lead to improvement in heart and skeletal muscle function. To test this hypothesis, we generated a mouse model of IOPD using CRISPR/Cas9 via pronuclear injection to introduce *Gaa*$^{c.2227C>T}$ and backcrossed this model to C57BL/6J mice. Cardiac pathology, skeletal muscle weakness, GAA enzyme activity and glycogen accumulation are currently being assessed in this mouse model.

Adeno-associated virus (AAV)-9 has broad tropism, including for heart and skeletal muscle, but the high doses required to transduce skeletal muscle may result in severe immune-mediated toxicity and undesired liver transduction, leading to reduced treatment efficacy. Since the ABE cassette exceeds the maximum AAV cargo capacity (~5 kb), we adopted a split-intein approach to divide the N- and C-terminal halves of ABE to two viral vectors, which results in trans-splicing of the expressed proteins *in vivo*. We sought to determine the lowest AAV9-Split-ABE dose required to improve heart pathology (lowest effective dose) in neonatal *Gaa*$^{c.2227>T}$ mice (n=4 mice/dose). Mice received one low (2E10 vector genomes (vg)/g), mid (1E11 vg/g), or high dose (2E11 vg/g) via retro-orbital injection at 12 days of age. Sanger sequencing shows 21%±6% editing in heart and 56%±6% editing in liver at a dose of 2E11 vg/g. Evaluation of enzyme activity and glycogen storage in various tissues is on-going. Future work will focus on treating *Gaa*$^{c.2227>T}$ mice with AAV9- and MyoAAV-Split-ABE, a myotropic and liver de-targeted AAV capsid, at the lowest effective dose established through this pilot study.

This research was supported by CHOC (CSO263), the Campbell Foundation of Caring, and the National Institutes of Health (1-R01AR079223-01, 2021).

POSTER #9

Rescuing lethality in the Homozygote VCP$^{R155H/R155H}$ mouse model provides a robust disease model for testing AAV gene therapy

Lan Weiss[1], Alyaa Shmara[1], Victoria Boock[1], Pallabi Pal[1], Cheng Cheng[1], Virginia Kimonis[1]

[1]Division of Genetics and Genomics Medicine, Department of Pediatrics, University of California, Irvine, CA.

Pathogenic variants in Valosin Containing Protein (*VCP*) gene cause a unique autosomal dominant multisystem proteinopathy (MSP1) characterized by inclusion body myopathy, Paget disease of bone and frontotemporal dementia, and amyotropic lateral sclerosis. The mechanism has been shown to be gain of function of the *VCP* missense variant. The knock-in VCP mouse model carrying the common *R155H* mutation includes several of the features typical of the human disease. The VCP $^{R155H/R155H}$ homozygous mice exhibit progressive weakness and accelerated pathology prior to their early demise, typically before 3 weeks of age. While homozygous VCP $^{R155H/R155H}$ mice die before the weaning age when kept on the normal chow diet, lipid-enriched chow can rescue lethality by 50% and over 50% of mice can survive into young adulthood.

We have also been able to rescue lethality of *the* homozygous mice by removing healthy litter mate pups and delayed weaning of the VCP$^{R155H/R155H}$ homozygous pups. This strategy results in improved survival from 3 weeks to over 20 weeks of age. Using this robust mouse model, we tested the muscle specific AAV variant, AAVMYO as a vector for delivery of gene payloads to muscle tissue. Treatment with AAVMYO vector carrying micro-RNA targeting VCP, with or without wild-type VCP replacement, demonstrates improvement in the muscle pathology in VCP $^{R155H/R155H}$ mice.

Conclusion: Our strategy of high fat diet and delayed pup weaning and removing littermates is helpful in providing a useful disease model. Downregulating the VCP allele with AAVMYO vector carrying anti VCP micro-RNA is associated with improvement in the muscle pathology in the VCP$^{R155H/R155H}$ mice.

Success in these preclinical AAV studies holds potential for promising therapeutic benefits in patients with VCP disease and related diseases.

POSTER #10

Serial Brain MRI Volumetrics and Tractography of Atypical CLN2 patients receiving ICV cerliponase alfa treatment

Audrey Kao[1,2,5], Harriet Chang[1,2], Igor Nestrasil[3], Raymond Wang[1,2,4]

[1]Research Administration, Children's Hospital of Orange County, Orange, CA
[2]Division of Metabolic Disorders, Children's Hospital of Orange County, Orange, CA
[3]Department of Pediatrics, University of Minnesota, Minneapolis, MN
[4]Department of Pediatrics, University of California - Irvine School of Medicine, Irvine, CA
[5]California Health Sciences University College of Osteopathic Medicine, Clovis, CA

Ceroid Lipofuscinosis Type 2 (CLN2) is caused by deficiency of tripeptidyl peptidase 1 (TPP1) enzyme. The standard of care treatment for CLN2 is a biweekly intracerebroventricular enzyme replacement therapy (ICV-ERT) of recombinant human TPP1, or cerliponase alfa. The disease progression of classical CLN2 patients is relatively uniform, with motor function loss, ataxia, seizures, cognitive dysfunction, and vision loss beginning at ages 3-4 years. However, atypical CLN2 patients demonstrate later onset of clinical symptoms, follow a more heterogeneous disease course, and under-explored response to treatment. Thus, this study aimed to better understand the disease course of treated atypical CLN2; brain imaging results are reported here.

Five atypical CLN2 subjects (ages 14-18) were enrolled in the study. Four of the subjects (subjects 1 and 2, subjects 4 and 5) were sibling pairs. Each subject underwent brain magnetic resonance imaging (MRI) before starting ICV-ERT and each year after for up to 5 years. Post-imaging analytics for structure volume and tractography are presented as mean ± standard deviation.

Corpus callosum volume increased (+1.8±5.4%) but subject 1 had a decline of 5.1%. Total cerebellar volume decreased (-1.2±7.0%). Cerebellar gray matter volume decreased (-1.7±6.8%). while cerebellar white matter volume increased (+1.4±8.4%). Cortical volume decreased (-2.7±7.4%). Additionally, Subjects 1 and 2 underwent diffusion tensor imaging (DTI). Fractional anisotropy (FA) and radial diffusivity (RD) values were analyzed. The corpus callosum body FA increased (+2.7±0.6%) and RD decreased (-3.9±0.2%). Total brain white matter demonstrated reduced FA (-0.5±0.1%) and RD (-0.3±0.4%).

Overall, despite treatment, the subjects demonstrated reduced total cerebellar volume, cerebellar gray matter, and cerebral cortical volumes. For subjects 1 and 2, tractography showed myelination improved in corpus callosum myelination and declined total brain white matter.

POSTER #11

iPSC-derived Motor Neurons for Mitochondrial Disease Modeling and Drug Testing

Alexander Stover [1], Hafiz Muhammad Umer Farooqi [1], Wei-Lin Huang [1], Alexandra Latini [1,2], Philip Schwartz [1], Jose E. Abdenur [1]

[1]Laboratory for Energy Metabolism, Division of Metabolic Disorders, CHOC Children's, Orange, CA, USA; [2]Laboratório de Bioenergética e Estresse Oxidativo – LABOX, Departamento de Bioquímica, Centro de Ciências Biológicas, Universidade Federal de Santa Catarina, Florianópolis, Santa Catarina, Brazil

Background: Progressive neurological decline is a hallmark of many primary mitochondrial disorders (PMD). Traditionally, characterization of the molecular and functional abnormalities of PMD has involved the use of skin biopsy-derived fibroblasts. However, fibroblasts do not always express the expected cellular abnormalities, limiting our ability to explore therapeutic targets. Patient-derived induced pluripotent stem cells (iPSCs) can now be generated to improve tissue–specific manifestations of the disease and explore drug discovery. These iPSCs can accurately reproduce the disease phenotype and provide an ethical source of disease-specific differentiated cells for drug discovery.

Purpose: To model a mitochondrial disease, leukoencephalopathy with brain stem and spinal cord involvement and lactate elevation (LBSL), we are using patient-derived iPSC-based motor neurons (MNs) to understand the molecular basis of LBSL in neuronal tissue and screen therapeutic agents.

Methods: Two patient-derived and two isogenic control iPSC lines from LBSL patient blood were treated with dual SMAD inhibition and ventral patterning to differentiate the iPSCs into mature MNs. Immunofluorescent staining, flow cytometry, and microelectrode arrays were utilized for phenotypical and functional characterization. Cellular respiration was tested using a Seahorse bioanalyzer. Additionally, lactate levels were assessed to test efficacy of 5-Aminolevulinic acid plus iron (ALA/Fe) treatment.

Results: Tubb3+, CD25+, CD184-, CD44-, and CD15- staining confirmed the successful differentiation into MNs of both patient and isogenic control iPSC lines. The MNs were found to be electrically active. ALA/Fe treatment lowered lactate levels and improved cellular respiration.

Conclusion: We have successfully established patient-derived iPSC lines-based MNs for LBSL disease, The MNs exhibited decreased energy productions and increased lactate production, which improved after ALA/Fe treatment. This model has significant potential to study mitochondrial disorders that mostly affect the brain. Further studies are underway to investigate the mechanism behind the alleviation of oxidative stress in LBSL MNs using ALA/Fe and other drugs.

POSTER #12

A Tunable Platform Technology for Early Detection of Traumatic Brain Injury

S. Calvin Li, PhD; Zhongping Chen, PhD; John F. Zhong, PhD; Hong Zhen Yin, MD; Steven S. Schreiber, MD-PhD; John H. Weiss, MD-PhD; Mustafa H. Kabeer, MD; William G. Loudon, MD-PhD*

Background: Traumatic brain injury (TBI) presents a significant health threat. The inflammatory response to TBI involves infiltrating leukocytes, such as macrophages (MA), and neural stem cells (NSCs), which can either promote repair or exacerbate tissue damage, depending on the TBI-microenvironment. Currently, no method exists to differentiate between these outcomes. Therefore, a tunable tissue-engineered graft (ETG) model is needed to better understand the initiation of TBI.

Purpose/Aim: This study combines the TBI/ETG model with multiphoton microscopy (MPM) and single-cell transcriptomics to identify biomarkers that reflect the interactions between TBI, MA, NSCs, and potential treatments. Our goal is to enable the early, rapid, and non-invasive detection of TBI and to track treatment-driven subclonal evolution at the single-cell level.
Methods: We employed a TBI ETG model (referencing PLoS ONE 10: e0120336 and PNAS 108:12793-8), single-cell transcriptomics (PubMed PMID: 32952853), and in vivo multiphoton microscopy (MPM) (Stem Cell Rev. 6:317-33).

Results and Discussions: Here, MPM imaging was used to fluorescently tag macrophages (MA) and neural stem cells (NSCs) associated with TBI-injured tissue. These tagged cells were then subjected to single-cell transcriptome profiling using our innovative microfluidic devices. Molecular differences between normal and injured cells exposed to various drug treatments were evaluated to identify optimal therapeutic combinations. We show that multiphoton microscopy (MPM) offers several key advantages. First, unlike other imaging techniques, MPM provides a unique window into cellular and subcellular processes in both in vivo and ex vivo ETG models. Second, MPM's capacity to capture multiple fluorescent signals from a single sample allows for robust correlative analyses by characterizing up to three parameters within a single volume. Our tumor models show that MPM achieves high-resolution imaging of tumor invasion zones and adjacent normal tissues down to the single-cell level in unprocessed living tissue. Third, MPM can utilize multiphoton excitation and four-dimensional microscopy to create fluorescence lifetime maps of ETG anatomy.

Conclusions: The integration of MPM with high-resolution multi-parameter imaging and single-cell transcriptomics provides a tunable system for individualized early diagnostics in traumatic brain injury. By profiling MA-NSC interactions on TBI-ETGs, we can advance the understanding of injury progression and inform personalized treatment strategies.

POSTER #13

Improvement of hypertrophic cardiomyopathy in *Gaa*$^{c.1826dupA}$ knock-in murine model with neonatal gene therapy

*Shih-hsin Kan[1], Jerry F. Harb[1], Songtao Li[2], Nancy D. Dalton[1], Alejandra Padilla[1], Chloe L. Christensen[1], Allisandra K. Rha[1], Perla Andrade-Heckman[1], Yunghang Chan[1], Evelyn Torres[1], Dwight D. Koeberl[2,3], Raymond Y. Wang[4,5]**

[1]CHOC Children's Research Institute, Orange, California, USA; [2]Division of Medical Genetics, Duke University School of Medicine, Duke University Medical Center, Durham, NC; [3]Department of Molecular Genetics and Metabolism, Duke University Medical Center, Durham, NC; [4]CHOC Children's Specialists, Orange, CA; [5]Department of Pediatrics, University of California-Irvine, Irvine, CA

Pompe disease (PD) is caused by deficiency of lysosomal acid α-glucosidase (GAA), leading to the accumulation of intra-lysosomal glycogen, particularly within skeletal and cardiac muscle tissues. We generated and characterized a novel PD murine model, *Gaa*$^{c.1826dupA}$, orthologous to the human *GAA* mutation c.1826dupA (p.Y609*). *Gaa*$^{c.1826dupA}$ mice display significantly reduced levels of *GAA* expression and enzymatic activity relative to wild-type (WT) mice.

Gaa$^{c.1826dupA}$ mice exhibit early-onset, progressive cardiac hypertrophy, demonstrating significantly increased diastolic left ventricular wall thicknesses, interventricular septum (IVS_d), left ventricular posterior wall ($LVPW_d$), and left ventricular mass index (LVMI) from 12 weeks of age compared to age-matched WT controls. *Gaa*$^{c.1826dupA}$ mice also display impaired skeletal muscle strength and coordination compared to age-matched WT controls, with deficiencies becoming evident at 12 and 36 weeks of age for grip strength and rotarod performance, respectively. Moreover, *Gaa*$^{c.1826dupA}$ mice exhibit discernible differences in gait parameters from 12 weeks of age, as assessed through catwalk performance analysis.

To elucidate the impact of gene therapy in *Gaa*$^{c.1826dupA}$ mice, AAV2/8-LSPhGAA (2E9 vg/g body weight) was administered retro-orbitally to 12-day-old mice. Twelve weeks post-treatment, *Gaa*$^{c.1826dupA}$ mice demonstrated supraphysiological GAA enzyme activity in heart (588% of WT levels) and liver (967% of WT levels), and remarkable glycogen reduction in heart (95% lower than untreated *Gaa*$^{c.1826dupA}$ mice).

Gene therapy-treated *Gaa*$^{c.1826dupA}$ mice showed substantially reduced IVS_d, $LVPW_d$, and LVMI, to levels comparable to WT mice. However, no improvement in grip strength was observed in treated mice.

Altogether, this study illustrates that the *Gaa*$^{c.1826dupA}$ murine model recapitulates many aspects of human infantile-onset PD and serves as a valuable model for assessing therapeutic approaches, including genomic correction strategies.

POSTER #14

Identification of the Storage Products and Assessment of Enzyme Replacement Therapy for Pompe Disease

Allen K. Murray[1,2] and Virginia Kimonis[2]

[1]HIBM Research Group, Inc., Glycan Technologies, Inc., and [2]Dept of Pediatrics, UC Irvine

Background: Pompe disease is an inherited glycogen storage disease caused by a deficiency of the lysosomal a--glucosidase (GAA). The treatment by enzyme replacement therapy (ERT) is helpful but it is not as successful as anticipated.

Aim: The original aim was to assess the actions of enzyme replacement therapy on stored glycogen in tissue. However, in the course of the investigation we have discovered that the storage product is not only glycogen but includes a major fraction of soluble oligosaccharides that contain glucose. The role of these glucans must be identified and their origin determined.

Methods: Glycogen determination involved KOH/HCl extraction, ethanol precipitation of the glycogen, reduction of the ethanol fraction and amyloglucosidase degradation followed by enzymatic glucose determination.

Results: Work in this laboratory involves glycogen determination by two different methods extraction o with KOH and precipitation of glycogen in ethanol followed by amyloglucosidase degradation and enzymatic glucose determination and direct amyloglucosidase degradation followed by enzymatic glucose determination. In untreated Pompe mice the KOH extraction and ethanol precipitation method detected significantly less glycogen that the direct amyloglucosidase method. Amyloglucosidase will cleave any a-1,6- and a-1,4- linkages. The ethanol fraction was taken down in a Speed Vac and the material was first analyzed by the Phenol Sulfuric Acid method for carbohydrate followed by amyloglucosidase degradation and enzymatic glucose determination. We believe this to be the first report of oligosaccharide storage glucans in Pompe mice. A review of the literature going back to the development of the Pompe mouse model for the disease did not include verification of glycogen as the only storage product.

Conclusion: Identification of the glucan oligosaccharides and determination of their origin is important for our understanding of Pompe disease

Funding: NIH/NIAMS R21AR080972

POSTER #15

Feasibility of a 12-Week Yoga Intervention for Adolescents with Chronic Health Conditions

Jennifer Barrows, PhD, RN; Linda Mendoza, DNP, CPNP-PC, AE-C; Clarisse Casilang, MD; Olga Guijon, MD; John Schomberg, PhD, MPH; Paulina Schuhler, BSN

Background. Obesity is one of the most common health conditions, affecting nearly one quarter of U.S. adolescents aged 12 to 19 years. Yet only one in five adolescents get enough physical activity to reduce the risk of obesity and other health conditions. Physical activity levels decline with age: 42% of children aged 6 to 11 meet the guidelines of 60 minutes per day compared to 15% of adolescents aged 12 to 17. A community survey of CHOC adolescents found that most had health goals to be more physically active (62%) and maintain a healthy body weight (62%) despite having conditions such as asthma (100%) and overweight or obesity (62%). Many were interested in trying a new type of activity such as yoga to improve their health (46%). Thus, the purpose of this pilot study was to evaluate the feasibility of a 12-week yoga intervention to increase physical activity in adolescents with health conditions such as asthma and obesity.

Methods. CHOC Wellness on Wheels (WoW) partnered with Be the Change Yoga to offer a summer yoga program for adolescents. Participants were WoW patients aged 13 to 18 years who were overweight, obese, and/or underactive. They attended 60-minute small group yoga classes twice weekly for 12 weeks. Physical activity, body mass index (BMI), self-efficacy, social support, and health-related quality of life were measured with surveys, accelerometry, and health record review at baseline and post-intervention.

Results. 10 adolescents (mean age = 15 years) enrolled with a retention rate of 100%. Mean attendance was 43% (10/24 classes). 75% of classes were attended via zoom and 25% were attended in the studio. Post-intervention data collection is ongoing. Two participants experienced a reduction in BMI reflecting a shift from overweight to normal body weight.

POSTER #16

Asthma Control Phenotypes Predict COVID-19 Illness Severity in Children

J. Barrows[1], M. Baxi[1], L. Ehwerhemuepha[1,2], S. P. Galant[1,3]

[1]Children's Hospital of Orange County, Orange, CA, [2]Chapman University, [3]University of California, Irvine

Rationale. Evidence is mixed on the relationship between pediatric asthma and severe COVID-19 infection, and less is known about the influence of asthma control phenotypes on COVID-19 severity in children. The purpose of this study was to determine the effect of asthma control phenotypes on COVID-19 hospitalization in children.

Methods. A retrospective analysis was conducted on COVID-19 encounters from March 2020 to 2022 of children aged 3 to 18 years with clinician-diagnosed asthma. Asthma control phenotypes were defined by: (A) ≥2 oral corticosteroid courses and (B) any hospitalization(s) for asthma in the previous year. Children were categorized into two groups: (1) well controlled (WC) (neither A nor B) and (2) not well controlled (NWC) (A and/or B). Causal inference techniques were employed to develop the following models: 1) asthma vs. non-asthma, 2) WC vs. NWC asthma, and 3) non-asthma vs. WC vs. NWC asthma.

Results. 618,675 records were included. Children with asthma had 27% lower odds of COVID-19 hospitalization compared to non-asthmatic children (OR= 0.73, CI=.71-.76, p<.001). However, the effect varied depending on asthma control. Children with NWC asthma were nearly twice as likely to be hospitalized compared to WC children (OR=1.98, CI=1.77-2.23, p<.001). Further, WC asthma reduced the odds of hospitalization by 32% compared to no asthma (OR=.68, CI=.66-.69, p<.001). Conversely, NWC asthma increased the odds of COVID-19 hospitalization by 32% compared to no asthma (OR=1.32, CI=1.21-1.43, p<.001).

Conclusions. COVID-19 hospitalization in children is significantly influenced by asthma control levels, highlighting the importance of asthma management in mitigating risk.

POSTER #17

Smooth Sailing or Rocky Road in Navigating the Ureteral Orifice: Does Preoperative Tamsulosin Improve Successful Ureteroscopy in Children?

Ethan F. Rutherford[1,2], Carol A. Davis-Dao[1,2], Kathy H. Huen[3,4], Anthony Schaeffer[5,6], Kiarad Fendereski[5,6], Vi Nguyen[7,8], Sarah Marietti-Shepherd[7,8], Renea Sturm[3,4], Elias J. Wehbi[1,2], Kai-Wen Chuang[1,2], Antoine E. Khoury[1,2], Heidi A. Stephany[1,2]

[1]*Children's Hospital of Orange County, Orange CA*
[2]*University of California-Irvine School of Medicine, Orange CA*
[3]*UCLA Mattel Children's Hospital, Los Angeles CA*
[4]*David Geffen School of Medicine at UCLA, Los Angeles CA*
[5] *University of Utah School of Medicine, Salt Lake City UT*
[6]*Primary Children's Hospital, Salt Lake City UT*
[7]*Rady Children's Hospital, San Diego CA*
[8]*University of California-San Diego, San Diego CA*

BACKGROUND: Preoperative tamsulosin (Flomax®) efficacy has not been well established for use in pediatric ureteroscopy (URS) when aiming to improve ureteral access. We hypothesized that tamsulosin use in unstented pediatric patients improves URS ureteral access success.

METHODS: A retrospective, multi-institutional cohort study of patients aged 3-18 years who underwent primary URS from January 2017-December 2023 was conducted. Exposure was preoperative tamsulosin use. Patients with a ureteral stent placed prior to URS, history of URS within the past year, ureteral reimplant, or ureterovesical junction (UVJ) obstruction were excluded. Demographic, preoperative, and intraoperative data were recorded. The primary outcome was success with primary URS—ability to pass a ureteroscope into the ureter. The secondary outcome was use of additional intraoperative techniques for ureteral access. Univariate and multivariable logistic regression analyses were performed, adjusting for potential confounders.

RESULTS: 115 patients from 4 institutions were included: median age at surgery was 15 years, with a median BMI of 20.4. Primary surgical indication was nephrolithiasis (92%). No significant differences in preoperative and intraoperative characteristics between the two cohorts were observed. Pre-operative tamsulosin was administered for median 14 days. Overall success of URS was 84%. On univariate analysis, URS success did not differ between cohorts (tamsulosin 86% vs. non-tamsulosin 83%, p=0.66). Tamsulosin use likewise did not affect success when additional intraoperative techniques for ureteral access (coaxial dilators, balloon dilators or ureteral access sheaths; 43% vs. 44%, p=0.97). When adjusting for age, sex, BMI, ureteroscope type and ureteral access techniques, tamsulosin use did not significantly improve successful URS odds (OR 1.5, CI 0.44-5.1, p=0.51).

CONCLUSIONS: Tamsulosin was not associated with improved odds of ureter access in pediatric patients undergoing URS. Potential limitations include sample size and varied Tamsulosin administration protocols. Future, larger series are needed to further understand the utility of tamsulosin for pediatric URS.

POSTER #18

"Dealers Choice": Univalve Location Has No Effect on Skin Surface Pressures in Long Leg Casts

Ryan Quinn, DO[1,2]; Gian Ignacio, MD[2,3]; Hayley Ditmars, BS[2,4]; Christopher Baker, BS[2,5]; John A. Schlechter, DO[1,2]

[1]Department of Orthopaedic Surgery, Children's Hospital Orange County, CA, USA
[2]Department of Orthopaedic Surgery, Riverside University Health System, CA, USA
[3]The Warren Alpert Medical School of Brown University, RI, USA
[4]University of Kansas School of Medicine, Lawrence, KS, USA
[5]Philadelphia College of Osteopathic Medicine – South Georgia, Moultrie, GA, USA

Background: Fiberglass long leg cylindrical casting can limit the ability of the extremity to swell, increasing chances of compartment syndrome of the leg. Casts are frequently univalved to accommodate for soft tissue edema. Still, there remains a paucity of information analyzing the effect of univalve location on skin surface pressures (SSP) in patients with long leg casts.

Purpose/Aims: By comparing the laterality of cast univalves, our research aims to better inform clinical practice techniques and optimize the management of trauma patients at risk of lower extremity compartment syndrome.

Methods: A 100-mL saline bag attached to a pressure transducer was placed along the anterior or posterior compartment of a single adult volunteer underneath 20 and 26 long leg casts, respectively. The casts were randomly assigned to receive either a lateral or medial univalve. The bag was insufflated with water to approximately 100 mmHg, and the change in SSP was recorded with a univalve (stage I), 3 mm cast spacer (stage II), 6 mm cast spacer (stage III), and bivalve (stage IV) in both groups. The study was powered sufficiently to detect a SSP difference of 10 mmHg.

Results: In the anterior and posterior compartments, when comparing medial versus lateral univalves, there were no significant differences in the change of SSP within any given stage (I through IV). Comparing stage I and stage IV, there was a significant difference in the change of SSP across all anterior and posterior compartment groups ($p < 0.001$, 95% confidence).

Conclusions: There were no statistically significant differences in anterior and posterior SSP in long leg casts with either a medial or lateral univalve. Our data supports a "dealer's choice" that the practitioner may select either a medial or lateral univalve to reduce anterior and posterior SSP

POSTER #19

Mom's developing too: The reciprocal effects of infant sensory reactivity and maternal sensitivity

Emily Campi,[1] Vincent Berardi,[1] Elysia Poggi Davis,[2,3] Curt A. Sandman,[4] Laura M. Glynn[1]

[1]Department of Psychology, Chapman University; [2]Department of Psychology, University of Denver; [3]Department of Pediatrics, University of California, Irvine; [4]Department of Psychiatry and Human Behavior, University of California, Irvine

Background: Caregiver sensitivity to infant cues is a well-established predictor of child development (Mesman et al., 2012; Madigan et al., 2019; Vallotton et al., 2017). Infants also actively influence their caregivers, even from their earliest days (Fantasia et al., 2019; Wu, 2021). Sensory reactivity (SR; hypo- [under] and hyper- [over] responding to stimuli) is one domain of development that may influence caregiver-infant interaction, due to its role in regulating emotions and responses to physical and social stimuli. Although SR could be an important target for improving caregiver-child interaction, relations between infant SR and caregiver sensitivity are largely unknown.

Purpose: To examine the longitudinal, reciprocal associations between infant SR and maternal sensitivity across the first year of life.

Methods: In a community sample (N=252), we measured infant SR on the novel Sensory Subscales of the Infant Behavior Questionnaire-Revised (Gartstein & Rothbart, 2003) and maternal sensitivity via an observational coding scheme (NICHD, 1999) at 6 and 12 months. We used cross-lagged panel models (CLPM) to test longitudinal associations.

Results: Results indicated that maternal sensitivity decreases infant hyperreactivity (β = -.21; p = .002), while infant hyporeactivity increases maternal sensitivity (β = .14; p = .02).

Conclusions: The results of this study provide initial evidence for the mutually influential nature of infant sensory reactivity and maternal sensitivity over time and suggest potential new intervention targets for improving the quality of parent-infant interaction to support optimal developmental outcomes.

Funding: National Institutes of Health P50 MH096889, MH86062, HD51852, HD40967, MH73136

POSTER #20

Predictors of Risk of Subsequent Urinary Tract Infection in a Multicenter Cohort of Patients with Primary Vesicoureteral Reflux: Preliminary Insights on Risk Prediction

Sara Alshehabi BS[1], Carol A. Davis-Dao PhD[1], Elias J. Wehbi MD[1], Cameron J. Hinkel MD[3], Allen Nguyen BS[1], Kai-Wen Chaung MD[1], Heidi A. Stephany MD[1], Ala'a Farkouh MD[2], Ruby Kuang[2], David A. Chamberlin MD[2], Emily S. Blum MD[3], Joshua D. Chamberlin MD[2], Christopher S. Cooper MD[4], Andrew J. Kirsch MD[3], Antoine E. Khoury MD[1]

[1]Division of Pediatric Urology, Children's Hospital of Orange County and University of California, Irvine, Orange, CA, USA
[2]Division of Urology, Loma Linda University Health, Loma Linda, CA, USA
[3]Department of Pediatric Urology, Children's Healthcare of Atlanta, Atlanta, GA, USA
[4]Department of Urology, The University of Iowa, Iowa City, IA, USA

Background: Risk Stratification has become an attractive clinical management strategy in patients with primary vesicoureteral reflux (VUR)**.** However, little multicenter data has been gathered to study the modern VUR population. We have developed a large, multicenter cohort of VUR patients to further study the relationships between urinary tract infection (UTI) risk factors in a diverse group of patients.

Methods: Retrospective data on patients with primary VUR from 2010-2021 were collected from four academic centers. Children with secondary VUR, ureterocele, multicystic dysplastic kidney, less than 3 months of follow-up or incomplete data were excluded. Primary outcome was UTI during two years of follow-up following diagnosis. Multivariable Cox regression analysis was used to estimate adjusted UTI risk.

Results: Among 848 patients with primary VUR, 781 were included. Median age at diagnosis was 0.58 years, median follow-up time was 1.2 years and 33% (257/781) of patients had high grade VUR (grades IV-V). Males comprised 41% (311/781) of patients and 63% of males (196/311) were uncircumcised. A total of 140 patients (18%) developed a UTI during two years of follow-up and median age at UTI was 1.3 years. In multivariable Cox regression models, several key UTI risk factors were identified. The predictors of 2-year UTI risk in this cohort were: female gender, presence of foreskin, high grade VUR and Hispanic ethnicity (compared with non-Hispanic whites).

Conclusions: In a large analysis of VUR patients, females, uncircumcised males (particularly those under one year old), patients with high grade VUR, and patients of Hispanic ethnicity were at highest risk of UTI development within two years of diagnosis. These preliminary findings provide additional data on important factors to consider for risk stratification. Next steps involve refining a risk prediction model using a combination of traditional statistics and machine learning models.

POSTER #21

Cast Art Analgesia

Jackson Helms, DO[1,2]; Hayley Ditmars, BS[2,3]; Christina Ayoub, MD[1]; John A. Schlechter, DO[1,2]

[1]Department of Orthopaedic Surgery, Children's Hospital Orange County, CA, USA
[2]Department of Orthopaedic Surgery, Riverside University Health System, CA, USA
[3]University of Kansas School of Medicine, Lawrence, KS, USA

Background: A multidisciplinary approach to pain management has been described as a means of mitigating stress for pediatric patients undergoing a painful procedure. One method of non-pharmacological support is cast art. No study to date has demonstrated the effect that cast art has on pain perception.

Purpose/Aims: By evaluating the outcomes of cast art, our research aims to inform alternative methods of non-pharmacologic pain management.

Methods: A retrospective chart review was performed on 68 patients from a single institution who underwent closed reduction with percutaneous pinning (CRPP) of supracondylar humerus (SCH) fractures. Patients were separated into two cohorts: cast art versus control. Visual analog scale (VAS), Face, Legs, Activity, Cry and Consolability (FLACC), and medication administration records (MAR) scores while each patient was in the post-operative anesthesia care unit (PACU) and on the medical-surgical floor were collected. Statistical analysis was performed to evaluate the differences in perceived pain and analgesic requirement.

Results: There was no significant difference in laterality of injury, BMI, sex, or fracture type. The control group had statistically significant shorter surgery times (20.1 min ± 5.6 vs 24.6 min ± 8.4, p=0.012) and time under anesthesia (49.9 min ± 9.9 vs 59.9 min ± 19.4, p=0.01). The cast art group had significantly lower VAS (p=0.009) and FLACC (p=0.004) scores, a significantly lower number of patients who required acetaminophen (p=0.048), and lower dose of ibuprofen given to patients on the floor (p=0.014). Most importantly, the number of cast art patients who required narcotics during admission was approximately four times less than in the control group (p=0.003).

Conclusions: Patients undergoing CRPP for SCH fracture were found to have lower subjective pain scores and decreased analgesic requirements on the medical-surgical floor if they received cast art. This supports the use of cast art as an effective non-pharmacologic pain management tool.

POSTER #22

Assessment of Bladder and Bowel Dysfunction among Spanish Speakers Presenting to a Pediatric Urology Outpatient Clinic

Gabriela De Lara[1,2], Sara Alshehabi[1,2], Carol A. Davis-Dao[1,2], Timothy Boswell[1,2], Elias J. Wehbi[1,2], Kai-Wen Chuang[1,2], Heidi A. Stephany[1,2], Antoine E. Khoury[1,2]

[1]Division of Pediatric Urology, Children's Hospital of Orange County, Orange, CA
[2]Department of Urology, University of California-Irvine School of Medicine, Orange, CA

Background: Bladder and bowel dysfunction (BBD) presents significant challenges in pediatric urology in both English speaking and non-English speaking populations. This study aimed to evaluate a newly adapted Spanish-language survey and assess the characteristics of children with and without BBD from Spanish-speaking families attending a pediatric urology clinic.

Methods: A culturally adapted Spanish-translated Vancouver Symptom Score for Bladder and Bowel Dysfunction was administered to 159 Spanish-speaking pediatric patients aged 3 to 21 years or their caregivers at an outpatient urology clinic. Data were collected on demographics, symptoms, and survey results, with analysis focusing on differences in symptom prevalence by patient demographics and survey accessibility. Patients with survey scores of 11 or higher were defined as BBD patients, while patients with scores 10 or below were defined as non-BBD patients.

Results: Most of the surveyed patients were male (68%) and identified as Hispanic/Latino (92%). Commonly reported BBD symptoms were urinary incontinence, frequent urination, and constipation. Females reported significantly higher BBD symptom scores than males (median score 9.0 vs. 5.5; $p=0.005$). Notable reasons for urology visits among BBD patients were lower urinary tract symptoms (52%) and other urologic problems (32%), while the main reasons for visit for non-BBD patients were penile or scrotal complaints (43%) and other urologic problems (45%) ($p<0.0001$).

Conclusions: This Spanish language survey effectively identified BBD symptoms in a pediatric population, highlighting reasons for presentation and variation in symptom prevalence by gender. The study underscores the need for targeted diagnostic tool catering to the linguistic needs of diverse populations.

POSTER #23

Optimizing Voriconazole Dosing In Premature Infants With Primary Cutaneous Aspergillosis: When Large Doses Are Necessary For Tiny Babies

Adam Lee,[1,3]; M. Tuan Tran,[2] ; Anne Denslow; Beverly Walti; Kushal Bhakta; Antonio C Arrieta,[1,3]

[1] Department of Infectious Diseases, Children's Hospital of Orange County, Orange, California, USA
[2] Department of Pharmacy, Children's Hospital of Orange County, Orange, California, USA
[3] Department of Infectious Diseases, University of California Irvine, Orange, California, USA

Introduction: Primary cutaneous aspergillosis (PCA), increasingly identified in preterm infants (preemies), often results in dissemination. Voriconazole is the preferred antifungal to treat Aspergillus infections. Pediatric patients require larger per kilogram doses than adults. Limited dosing information in preemies exist; free drug exposure (*f*AUC)/MIC > 25 or trough 1-5 mcg/mL are associated with efficacy. We report voriconazole use in preemies with PCA using therapeutic drug monitoring (TDM) for dosing optimization.

Methods: We conducted a retrospective review of preemies with PCA admitted to Level 4 Neonatal ICU (2021 to 2023). Information on demographics, clinical course, pathology and culture results, voriconazole dosing, drug concentrations, and toxicity data (complete blood count, liver chemistries) were obtained. Voriconazole was initiated at 3-4 mg/kg/dose every 8 hours intravenously (IV). Using TDM, doses were adjusted to obtain targeted $AUC_{(0\text{-}24)}$/MIC or trough levels.

Results: Three preemies (<25 weeks gestation) with PCA survived to discharge with resolution of infection (a fourth still undergoes treatment). Three patients underwent debridement (two had biopsies with clear margins). Biopsies demonstrated invasion of hyphae into subcutaneous tissues; cultures identified Aspergillus spp. Three had antifungal susceptibilities. All patients were initiated on IV voriconazole and switched to enteral route when possible. 73% of trough levels while on IV voriconazole were between 1-5 mcg/mL, and AUC ranged 96-173. Trough levels on patients receiving enteral voriconazole were consistently < 1 mcg/mL even after dose escalation (range 9.2-19 mg/kg/day) (Table 1). No hematological, hepatic, or cutaneous adverse events were identified.

Conclusions: Parenteral voriconazole for treatment of PCA in preemies at 3-4 mg/kg/dose q8h, with TDM to ensure adequate drug exposure, was well tolerated; combined with surgical debridement, it showed consistent therapeutic success. Enteral dosing failed to produce trough levels in therapeutic range.

POSTER #24

Procedural Anxiety in Pediatric Emergency Care: Incidence and Predictors of Nonresponse to Intranasal Midazolam in Children Undergoing Laceration Repair

Kelly Bauer[1], Jenny Zhu[1], Theodore W. Heyming[2, 3], Helen Lee[2], Zeev N. Kain[4-7], Sarah R. Martin[2, 4, 5]

[1] University of California Irvine School of Medicine; [2] Emergency Medicine, Children's Hospital of Orange County; [3] Department of Emergency Medicine, University of California Irvine; [4] Department of Anesthesiology and Perioperative Care, University of California Irvine School of Medicine; [5] Center on Stress & Health, University of California Irvine; [6] Children's Hospital of Orange County; [7] Yale University Child Study Center

Background: Pediatric laceration repairs are common in the emergency department (ED) and often associated with procedural anxiety. Despite the increased use of intranasal midazolam (INM) prior to pediatric ED procedures, there is limited, low quality data on the effects of INM on anxiety.

Purpose/Aims: This study described the proportion of children who were nonresponsive to INM (i.e., exhibited extreme anxiety) and identified factors associated with INM non-response in children undergoing laceration repair.

Methods: This cross-sectional study included 102 children (ages 2–10 years) who received INM prior to laceration repair in the ED. Procedural anxiety was scored using the modified Yale Preoperative Anxiety Scale (mYPAS). Children scoring >72.91 (extreme procedural anxiety) were considered INM non-responders. Bivariate and multivariable logistic regression analyses explored associations between age, temperament, laceration location, sedation, time from INM administration, and likelihood of INM non-response.

Results: In this sample, 45.1% of the children were classified as INM nonresponders. Nonresponders were younger (p=.050), had lower sociability temperament (p=.006), longer delay between INM administration and the procedure (p=.035), lower sedation levels (p<.001), and were more likely to have extremity lacerations (p=.003). In multivariable regression, extremity lacerations (OR=8.66, 95%CI [1.72,43.67], p=.009) and lower sociability temperament (OR=0.30, 95%CI [0.13,0.71], p=.006) were associated with likelihood of INM nonresponse.

Conclusions: Nearly half of the children in the sample exhibited extreme procedural anxiety despite receiving INM. This finding has important clinical implications, suggesting that pharmacological interventions alone may not be sufficient to manage all pediatric procedural anxiety in the ED. Findings highlight a need for further research examining multimodal strategies to manage procedural anxiety in the pediatric ED, particularly for children with extremity lacerations or low sociability temperament.

Financial support: National Institutes of Health National Institute for Child Health and Human Development (K23HD105042, PI: Martin)

POSTER #25

Smartphone Digital Radiograph Review Prior to Transfer to a Tertiary Pediatric Trauma Center: A Picture is Worth 1000 Words

Kevin Z Kwan, DO[1]; Hayley Ditmars, MS[2,3]; Amirhossein Misaghi, MD[2]; John A Schlechter, DO[1,2]

[1]Department of Orthopaedic Surgery, Riverside University Health System, CA, USA
[2]Department of Orthopaedic Surgery, Children's Hospital Orange County, CA, USA
[3]University of Kansas School of Medicine, Lawrence, KS, USA

Background: Orthopedic injuries in children are one of the most common reasons for a higher level of care transfer to a pediatric trauma center. When transfers are warranted, the care of these children can often be expedited via images sent to the smartphones of the consulting orthopedic surgeon. Given the increasing reliance on advanced practice providers (APPs) in referring institutions as well as outside radiologists who are often less experienced with evaluating musculoskeletal injuries in children, this can lead to discrepancies in diagnosis that may affect patient care.

Purpose/Aims: This study aims to evaluate the accuracy and significance of evaluating digital radiographs for children with isolated orthopedic injuries prior to transfer.

Methods: A retrospective chart review was performed on all children that were transferred for isolated orthopedic injuries to a pediatric tertiary referral center from September 2023 – May 2024. All children that were transferred from outside hospitals had accompanying deidentified images which were sent to the on call orthopedic surgeons smartphone and outside radiologist's reports were included. Demographic information was collected. All radiographs were reviewed by a pediatric fellowship trained orthopedic surgeon and corresponding diagnosis given and compared to the outside radiologist's read for corroboration.

Results: 27 children were included, 24 of which underwent surgical intervention. 10/27 (~37%) reads from the outside radiologists were not in agreement with the diagnosis assigned by the reviewing orthopedic surgeon. The mean time to intervention once arriving to our institution for reduction or surgery was 364 minutes (82-911).

Conclusion: The utility of receiving images prior to transfer is underscored by these findings allowing on call orthopedic surgeons who will ultimately be providing care for these children to expedite triage, more accurately know what to expect prior to arrival and better prepare to care for these injuries after evaluating radiographs through their smartphones.

POSTER #26

Variation in Admission and Treatment Decisions for Previously Immunocompetent Pediatric Patients Presenting to the ED with Febrile Neutropenia

Britanny Winckler, MD, Brianna Leyden, MD, Ricardo Aguilar, MS, Jennifer Lusk, MD

Children's Hospital of Orange County

Background: Patient volumes limit studies of immunocompetent pediatric patients with febrile neutropenia; no guideline consensus exists. Extrapolating serious bacterial infection (SBI) risk from other populations leads to resource overutilization.

Purpose/Aims: We quantified practice variation in emergency department (ED) disposition for immunocompetent pediatric patients with febrile neutropenia. We evaluated which factors increase admission odds and the association of disposition and SBIs.

Methods: We performed a multi-center retrospective study using Cerner Real-World Data. ED patients ages 2 months – 18 years from January 2016-June 2023 with an absolute neutrophil count (ANC) < 1500 and temperature > 38° C were included. Encounters resulting in transfer or death or with diagnoses of neoplasms, immunocompromised status, or implanted devices were excluded. A logistic regression mixed effects model determined the variance partition coefficient (VPC), the proportion of variance explained by health systems differences, and what factors increase admission odds. Univariate analysis (chi-squared and Fisher's exact tests) examined the association of ED disposition and rates of sepsis/SIRS or SBIs (bacteremia, bacterial meningitis, or urinary tract infection) within 5 days of presentation.

Results: There were 5,554 qualifying encounters. Estimated VPC is 8% (Table 1). Diagnoses of sepsis/SIRS ($p<0.001$), bacteremia ($p<0.001$), and bacterial meningitis ($p = 0.002$) were more likely in admitted encounters. Only bacteremia was associated with discharged encounters (Table 2). Multiple factors increased admission odds: diagnoses of sepsis/SIRS (OR 14.02), pancytopenia (5.49), thrombocytopenia (5.13), and anemia (5.06); if antibiotics were given (4.75); system characteristics of children's hospital encounter; after the COVID-19 pandemic (Table 3).

Conclusions: Our study showed that variation in practice is relatively low. Patients with SBIs are more likely to be admitted, suggesting providers correctly identify at-risk patients. Further studies should elucidate the association between ED discharge and bacteremia. With validation of factors associated with increased admission, a safe clinical practice guideline to optimize outcomes and decrease overutilization is possible.

POSTER #27

Project POOP: Preventing Opioid-Originated Problems, a Quality Improvement Project

Brianna Leyden, MD, Katherine Stringer, RN, PEP Consultant.

Children's Hospital of Orange County

Background: Opioid-induced constipation (OIC) is a well-known complication of pain management in pediatric populations, affecting significant numbers of patients with chronic pain, critical illness, and acute post-operative pain. Utilizing bowel regimens can be one tool to both treat and prevent constipation.

Purpose/Aims: Improving bowel regimens initiated early in patients on opiate prescriptions will lead to decreased lengths of stay, decreased future ED visits or readmissions, and decreased cost burden on the healthcare system. This project aimed to increase the prescription of bowel regimens at discharge for patients on opioids from 24% to 40% by July 2024.

Methods: Stakeholders were assembled including hospitalists and surgeons, pain and palliative team representatives, nursing, and members of the Opioid Stewardship Task Force. The team discussed the current state of OIC on the medical-surgical units including complications, lack of standardized protocols, and barriers to implementing changes. Using Plan-Do-Study-Act (PDSA) methodology, the primary outcome of percent of patients on opioids prescribed bowel regimens at discharge was obtained pre- and post- interventions via chart review, also broken down to individual services of medical, surgical, oncologic, and ED. Choice of bowel regimen was also collected. A balancing measure of follow-up gastrointestinal complaints was also measured via chart review. Interventions included education via email, group lectures, and committee meetings, and creation of a bowel regimen order set in the Electronic Medical Record.

Results: Bowel regimen prescription at discharge increased overall from 24% October 2023 to 67% August 2024. Prescription by hospitalist services increased from 67% to 100%, 16% to 36% by surgical services, and 43% to 100% by oncologic services, with no consistent trend in the balancing measure post-discharge.

Conclusions: Increasing bowel regimens in patients discharged with opioids has not led to increased gastrointestinal complications. Additional work to increase prescription of regimens across all services and increase utilization of the standard order set remains.

POSTER #28

Variation in Urinary Tract Infection Risk by Race/Ethnicity among Patients with Vesicoureteral Reflux: A Multicenter Analysis

Allen Nguyen BS[1], Elias J. Wehbi MD[1], Carol A. Davis-Dao PhD[1], Cameron J. Hinkel MD[3], Sara Alshehabi BS[1], Kai-Wen Chaung MD[1], Heidi A. Stephany MD[1], Ala'a Farkouh MD[2], Ruby Kuang[2], David A. Chamberlin MD[2], Emily S. Blum MD[3], Joshua D. Chamberlin MD[2], Christopher S. Cooper MD[4], Andrew J. Kirsch MD[3], Antoine E. Khoury MD[1]

[1]Division of Pediatric Urology, Children's Hospital of Orange County and University of California, Irvine, Orange, CA, USA
[2]Division of Urology, Loma Linda University Health, Loma Linda, CA, USA
[3]Department of Pediatric Urology, Children's Healthcare of Atlanta, Atlanta, GA, USA
[4]Department of Urology, The University of Iowa, Iowa City, IA, USA

Introduction : Changing criteria for the diagnostic workup of children with urinary tract infections (UTI) has impacted patients with vesicoureteral reflux (VUR). In parallel, the Hispanic population has increased dramatically in the United States. Our objective was to investigate the risk of urinary tract infection by race/ethnicity in a contemporary group of patients with VUR with a focus on Hispanic patients.

Methods: Patients with primary VUR diagnosed by voiding cystourethrogram from 2010-2021 were collected from four academic centers. Children with secondary VUR, less than 3 months of follow-up or incomplete data were excluded. Primary outcome was confirmed urinary tract infection during two years of follow-up following diagnosis. Multivariable Cox regression analysis was used to estimate adjusted UTI risk.

Results: Of 848 patients, 781 met inclusion criteria. Median age at diagnosis was 7 months and 40% were male. Of those patients with self-reported race/ethnicity, 43% were Hispanic. Non-Hispanic white patients had a 13% UTI rate, while Hispanic patients, had a 23% UTI rate. Among 52 Asian-American patients and 12 African American patients, the UTI rate was 17% and 0% respectively. In multivariable Cox regression analysis, the effect of race/ethnicity was analyzed with adjustment for significant UTI risk factors. In comparison with non-Hispanic White patients, Hispanic patients had a significantly increased risk of UTI ($p = 0.03$).

Conclusions: In a large multicenter group of patients with VUR, we found that Hispanic patients had a significantly increased UTI risk compared with non-Hispanic white patients. We also confirm that the rate of UTI among African American patients with VUR is low. The risk of UTI among Hispanic patients is noteworthy, as this is a rapidly expanding population in the U.S. The reasons for this difference in UTI risk and the possible impact of healthcare disparities among this population of patients requires further investigation.

POSTER #29

Who, why, and how to screen: Community perspectives on early life adversity screening and research in primary care settings

Vanessa M Vargas[1], Sabrina R Liu[2], Candice Taylor Lucas[3], Megan Maxwell[1], Natasha G Lindert[1], Dan M Cooper[3], Charles V Golden[4], Michael Weiss[4], Laura M Glynn[1]

[1]Department of Psychology, Chapman University, Orange, CA USA
[2]Department of Human Development, California State University, San Marcos, San Marcos, CA USA
[3]Department of Pediatrics, University of California, Irvine, Irvine, CA USA
[4]CHOC Children's Primary Care Network, Orange, CA USA

Background: Early life adversity (ELA), in the form of adverse childhood experiences and exposure to unpredictability, are associated with detrimental mental and physical outcomes in later adulthood (Centers for Disease Control, 2016, Felitti & Anda, 2010, Davis & Glynn, 2024; Glynn et al., 2019; Lindert et al., 2022). The SoCal Kids Study's purpose is to assess the effects of ACEs and unpredictability on child neurodevelopment. Given the sensitive nature of this topic, it is important to conduct research that is transparent and guided by community members to promote uptake of clinical practices in real-world settings. A way to ensure that this research study is ethical and inclusive is by holding community engagement studios (CES). Purpose/Aims: To engage community experts in the planning phases of the SoCal Kids Study to guide screening and research practices.

Methods: The research team held four separate CES: pediatricians (*n*=9), non-physician medical providers (*n*=5), and English (*n*=7) and Spanish (*n* = 3) preferring parents. These were moderated by an impartial third party unaffiliated with the project. A member of the research team provided an overview of the study, discussed ELA and its impacts on long-term health and development, and described study procedures. Afterwards, participants provided feedback on the feasibility, acceptability, and importance of the study's methodology and the ACEs and unpredictability screening tools. Transcripts of the CES were analyzed using rapid qualitative analysis.

Results: Five themes emerged: trust, benefits to future generations, procedural recommendations and concerns, perspectives on questionnaire responses, and self-reflection. Participants also endorsed the importance of engaging minoritized communities, screening for and scientific study of ELA, and the value of partnering with primary care physicians.

Conclusions: From these results, the researchers adopted recommendations including the use of visual aids for enhancement of recruitment and health literacy, and involving study team members that speak the same language as the participants. The study shows the advantage of partnering with community experts in the early stages of research and provides a framework for doing community-informed research on ELA.

This research was supported by the California Initiative to Advance Precision Medicine.

POSTER #30

Longitudinal Case Study of Sleep Architecture in Patient with Miller-Dieker Syndrome

Michael Nguyen, Kalysa Bui, Brandon Dang, Lia Galut, Spencer Polk, Neal Nakra, and Katharine Simon

University of California, Irvine, Irvine, CA USA

Introduction: Miller-Dieker Syndrome (MDS) is a rare genetic condition that involves microdeletion of chromosomal region 17p13.3 and is characterized by Type 1 lissencephaly, facial dysmorphic features, intellectual disability, and seizures (Baker et al., 2022). How MDS affects sleep architecture longitudinally given underlying brain structural abnormalities is unknown. One prior case study demonstrated surgical intervention improved obstructive sleep apnea (Baker et. al., 2022). In non-MDS patients with lissencephaly, few to no sleep event markers were present and they had larger slow wave amplitude (Falsaperla et al., 2018).

Methods: We retrospectively evaluated three clinical sleep studies from a female patient with MDS. Sleep studies consisted of three nocturnal sleep studies. Age of sleep records were 8 months, 8 months, and 20 months. We evaluated sleep macroarchitecture changes, including total sleep time, sleep stages, onset latency, and apneas.

Results: Across the studies, the total sleep time ranged 438.5 to 194.5 to 299 minutes. Slow wave sleep latency ranged from 7 to 40.5 minutes and for REM, ranged from 187.5 to 451 minutes. The proportion slow wave sleep ranged from 50.1% to 97.5%, while the proportion of REM ranged from 0.2 to 3.6%. The apnea index declined across studies from 1.4 to 0 and the hypopnea index varied from 0.8 to 2.3 to 0.2.

Discussion: Our study longitudinally evaluates sleep architecture changes in a patient with MDS. Thus far, in comparison with reported normative sleep, this patient has greater amounts of slow wave sleep. Further, PAP intervention was successful across the sleep studies, however, intervention did not impact overall sleep architecture. Ongoing analyses will be conducted to determine spectral power in slow wave (1-4Hz) and sigma (12 to 15Hz) ranges and characterize slow oscillation (.03-1Hz) and spindles (12 to 15Hz). Our results may point to novel avenues of intervention for these youth.

Funding: Pulmonology Department

POSTER #31

Physical Activity Is Associated With Maternal Metabolic Profile and Inflammation in Pregnant Mexican American Women

Hadavale D, Rasmussen JM, Buss C, Entringer S, Wadhwa PD, Gyllenhammer LE

BACKGROUND: Physical activity (PA) during pregnancy is a subject of growing interest, as it can influence maternal metabolic health outcomes and, consequently, fetal development. Engaging in regular PA during pregnancy is associated with improved glucose metabolism and reduced inflammation. This research aims to address this gap by investigating the associations between PA and maternal metabolic and inflammatory markers among pregnant Mexican American women, utilizing gold standard objective PA measurement.

METHODS: In 162 pregnant Mexican American individuals, PA was measured using the ActiGraph GTx3 accelerometer during mid- (26.1±2.6 wks) and late (34.5±2.0 wks) gestation. Vector magnitude counts per minute (CPM) was quantified as a measure of general PA. Fasting plasma glucose, insulin, total cholesterol (TC), HDL-cholesterol (HDL-C), adiponectin, IL-6, IL-10, TNF-alpha, and CRP were measured during mid- and late gestation. HOMA-IR was calculated with the following formula: fasting insulin (microU/L) x fasting glucose (nmol/L)/22.5. Associations between mean PA and maternal metabolic and inflammatory markers were assessed using bivariate correlations and linear regressions.

RESULTS: Mean PA, as measured in vector magnitude counts per minute, was positively correlated with adiponectin levels (p=0.018) and negatively correlated with HOMA-IR (p=0.008) and IL-6 levels (p=<.001). These relationships were found to be independent of maternal pre-pregnancy BMI.

CONCLUSIONS: In a cohort of pregnant Mexican American individuals, PA associates with improved maternal metabolic and inflammatory profiles, independent of maternal BMI. These results suggest that regular PA may support maternal health during pregnancy with important implications for both maternal and fetal health outcomes. Further exploration of these associations is needed to provide targeted PA guidelines among this underserved population, particularly at risk for pre-existing and gestational diabetes.

Funding: This research was supported by Grant 5R01MD010738-05 from the National Institute on Minority Health and Health Disparities.

POSTER #32

Efficacy of a dietary buttermilk powder enriched in milk fat globule membrane concentrated with gangliosides for treatment of pediatric IBD depends on SNPs in ganglioside metabolism enzymes

Sophie Alter[1], John Miklavcic PhD[1], Kenneth Grant MD[2], Michael Wang DO[2]

[1]Schmid College of Science and Technology, Chapman University; [2]Gastroenterology and Nutrition, Children's Hospital of Orange County

Background: IBD is a condition affecting about 0.5% of the western world and prevalence is rising globally and it can be debilitating for pediatric patients. Dietary gangliosides have been proposed as a new potential treatment for IBD. However, human sialidase enzymes may modify the treatment efficacy by catabolizing ganglioside GD3 to GM3 and then into lactosylceramide by the successive removal of sialic acid.

Purpose/Aims: To assess the feasibility of a dietary buttermilk powder treatment for IBD and to explore how endogenous ganglioside metabolism alters the efficacy of dietary buttermilk powder.

Methods: This double-blind randomized placebo-controlled clinical study involved patients (n=11) aged 9 – 21 years with mild-moderate IBD from June 2022 to February 2023. Participants assigned to the treatment group consumed 5 grams of buttermilk powder daily enriched in milk fat globule membrane. Participants were assessed for well-being using the IMPACT-III questionnaire, complete metabolic panel, and SNPs in genes for ganglioside metabolism: NEU3 (544115), NEU4 (rs2293761).

Results: Compared the patients with the wildtype (TT) genes, the average well-being domain score calculated from the IMPACT-III was 23% lower in carriers of a minor allele (C) in NEU3. At baseline, the average albumin level for patients with wildtype NEU4 (AA) was 4.83 (+/-0.44) g/L, compared to 4.04 (+/-0.35) g/L for carriers of the minor allele (G). Treatment with dietary ganglioside reduced albumin level by 16% in the wildtype group, but albumin was unchanged in carriers of the minor allele in NEU4.

Conclusions: These findings suggest that genetic variation in ganglioside metabolism pathways may influence the efficacy of dietary interventions with milk fat globule membrane for pediatric patients with IBD. Further research is needed to better understand how SNPs in genes for ganglioside metabolism may alter the efficacy of dietary buttermilk powder for clinical outcomes including disease activity index and physician global assessment.

Funding: This research was funded by the CHOC PSF grant and Chapman University Institutional Funds.

POSTER #33

Expanded phenotypes of b-mannosidosis: a new association with white matter disease

Angela Martin-Rios[1], Karolina M Stepien[2], Liliane H. Gibbs[3], Katherine Hall[1,4], Patricia L. Hall[5], Gisele Bentz Pino[5], Raymond Wang[6,7], Nishitha R. Pillai[8], Troy C. Lund[9], Paul J. Orchard[9], Virginia Kimonis[1,4,10,11]

[1]Division of Genetics, Department of Pediatrics, University of California - Irvine; [2]Adult Inherited Metabolic Diseases Department, Salford Royal Organization, Northern Care Alliance NHS Foundation Trust, Salford, United Kingdom; [3]Department of Radiological Sciences, School of Medicine, University of California, Irvine; [4]CHOC Children's Hospital; [5]Biochemical Genetics Laboratory, Division of Laboratory Genetics and Genomics, Mayo Clinic, Rochester; [6]Division of Metabolic Disorders CHOC Children's Hospital; [7]Department of Pediatrics, University of California-Irvine School of Medicine, Irvine, , USA; [8]Division of Genetics and Metabolism, Department of Pediatrics, University of Minnesota; [9]Division of Blood and Marrow Transplantation, Department of Pediatrics, University of Minnesota; [10]Department of Neurology, University of California - Irvine; [11]Department of Pathology, University of California - Irvine

Background: β-mannosidosis is a lysosomal storage disorder caused by the deficiency of β-mannosidase, which catalyzes the last step of degradation of N-glycans. The reported incidence of this condition is 0.12 to 0.16 per 100,000 live births. Descriptions of brain imaging in this condition is scarce, and there are no pathology reports of patients. Also, given the low number of patients reported, little information of the natural history of b-mannosidosis is available.
Purpose: To expand the spectrum of clinical phenotypes and brain imaging features of individuals with b-mannosidosis.
Methods: We describe the clinical features of six patients with b-mannosidosis, the biochemical profiles including enzyme activity and urine oligosaccharides, their findings on brain magnetic resonance imaging (MRI) and the changes over time in two of them. We also review previously reported patients, analyze the variants in *MANBA,* the first symptom and spectrum of symptoms, and the ages of onset and of diagnosis of the disease.
Results: Forty-four patients have been reported to date, including our patients. The mean age of diagnosis is 12.8 years, and the age of onset of symptoms is 2.4 years. Hearing loss is the most frequently reported initial symptom and intellectual disability is the most frequent symptom overall. Erythromelalgia, nystagmus, macrocephaly and obsessive-compulsive-like behavior are newly described features associated with b-mannosidosis. 40% of the patients have abnormal brain imaging. Brain MRI showed hypomyelination in one patient and dysmyelination in another patient. Twenty-nine pathogenic variants in *MANBA* have been reported among the 44 patients; 60.6% of patients have private variants, and 39.4% have the recurrent variant c.2158-2A>G.
Conclusion: Due to disease heterogeneity, establishing a genotype-phenotype correlation remains challenging in b-mannosidosis. Two patterns of brain MRI were described as manifestations of this condition: hypomyelination and dysmyelination. Additional studies are needed to delineate the pathophysiology of this condition.

POSTER #34

Age and Sex on Psychosocial Outcomes in Children with Cleft/Craniofacial Differences

Peter Dinh, MA[1,2], Jacob Paredes, BS[1], Parsa Khawari, BS[1], Raj Vyas, MD[1,2]

[1]Children's Hospital Orange County, CA, USA
[2]University of California-Irvine, CA, USA

Introduction: Children with craniofacial abnormalities (CFA) such clefts (lip/palate) and Hemifacial Microsomia may have poorer social and mental well-being. Poor social and mental well-being may stem from poor interpersonal and intrapersonal experiences related to the children's craniofacial abnormalities. We aim to identify these outcomes in our patients. This study examines the interaction of age and sex with psychosocial outcomes of anxiety, depression, stress, and anger among patients at CHOC with craniofacial abnormalities.

Methods: Our study examined patients (N=862) whose age ranged from 5-21 who presented at CHOC with CFA who had received or were being evaluated for surgical treatment. Mixed methods modeling was utilized to assess the relationship of age and sex on psychosocial outcomes. Patients were assessed using the NIH Patient-Reported Outcomes Measurement Information System (PROMIS) measures of anxiety, depression, stress, and anger. Additional t-test analyses were utilized to compare craniofacial patients and controls on their psychosocial outcomes.

Results: Our final analysis included 862 craniofacial, jaw, and speech patients. Our results indicate a significant inverse relationship between age, anxiety and anger, such that as age increased, anxiety ($\beta = -.02991$, $p < .0001$) and anger ($\beta = -.01168$, $p < .0001$) scores decreased. Furthermore, craniofacial patients had significantly higher anxiety ($p = .0464$), depression ($p = .0009$), and stress ($p = .0160$) compared to their control counterparts.

Discussion/Conclusion: Our results indicate a significant inverse relationship between age and anger/anxiety. Furthermore, psychosocial differences in anxiety, stress, and depression were observed between CFA and control patients. These results support the need for targeted social interventions for younger CFA patients. Specifically, to promote appropriate coping skills to manage negative emotions and stress. Additionally, there is a need for interventions that support the development of proper interpersonal skills that teach social cohesion of peers who may appear and sound different.

POSTER #35

Unusual Presentation of Retroperitoneal Teratoma

Shifa Shaikh MBBS, Hemanshi Shah MCH, Neha Sisodiya MCH, Vini Joseph MCH

Department of Pediatric Surgery, TNMC and BYL Nair Hospital, Mumbai, India

Objective: We present a case of a 1-year-old female child who had a rapidly growing retroperitoneal mass causing respiratory distress, which proved to be a retroperitoneal teratoma.

Background: Teratomas are tumors that contain tissues derived from all three germinal layers (ectoderm, mesoderm, and endoderm). Retroperitoneal teratoma are rare and are tumors of childhood. Primary retroperitoneal teratoma in children accounts for 3.5–4 % of all germ cell tumors and 1–11% of primary retroperitoneal neoplasms, although these tumors are silent in nature, especially in young children, they may present with abdominal pain, flank pain, or back pain which are nonspecific. However, it can also cause abdominal swelling, palpable mass or obstructive gastrointestinal/ genitourinary symptoms along with lymphatic obstruction. Rarely they may progress to an abscess formation due to secondary infections or an acute peritonitis due to rupture, or with hemorrhage within tumor and even malignant transformations. Here we are discussing an unusual presentation of retroperitoneal teratoma.

Results: In view of the rapidly progressing nature of the lesion, causing respiratory distress and with suspicion of intratumoral hemorrhage, a decision was taken to perform an emergency exploratory laparotomy. Histopathology study of the excised specimen revealed that it was a cystic structure measuring 10*7*4cm and was well encapsulated. The vertical cut section showed solid and large cystic areas with fluid. The absence of vertebral column bone, cartilage, and other anatomical structures on microscopy favored the diagnosis of Mature Cystic Teratoma over Fetus in Fetu.
The patient was followed up with an ultrasound and α-fetoprotein levels at 3 months, 6 months, and then yearly for two years. After 2 years of follow-up, there are no signs of recurrence and other complaints and the patient is doing well.

Conclusions: The main step of treatment for retroperitoneal teratoma is complete surgical resection and the prognosis depends primarily upon the adequacy of surgical resection. Detailed gross and microscopic histopathological examination of the excised tumor stands as the gold standard for diagnosis. Surgical excision of retroperitoneal teratoma in toto is a curative and definite treatment modality in such patients.

POSTER #36

Incidence and Risk Factors for Hypersensitivity Reactions in Pediatric and Adolescent/young adult Patients Receiving Premedication for Pegaspargase: a Prospective Multicenter Study

Van T. Huynh, MD[1], Sonia Morales, MD[1], Anurag K. Agrawal, MD[2], Christine L. Phillips, MD[3], Beth A. Winger, MD, PhD[4], Catherine Aftandilian, MD[5], Janel R. Long-Boyle, PharmD, PhD[6], Keith J. August, MD, MS[7], Erin Guest, MD, MS[7], and Carol H. Lin, MD[1]

[1]Division of Oncology, Children's Hospital of Orange County (CHOC), Orange, CA; [2]Division of Oncology, Department of Pediatrics, UCSF Benioff Children's Hospital Oakland, Oakland, CA; [3]Division of Oncology, Department of Pediatrics, Cincinnati Children's Hospital, Cincinnati, OH; [4]Division of Hematology-Oncology, UCSF Benioff Children's Hospital, San Francisco, CA; [5]Division of Pediatric Hematology/Oncology/Stem Cell Transplant and Regenerative Medicine, Lucile Packard Children's Hospital, Palo Alto, CA; [6]Division of Pediatric Allergy, Immunology, and Bone Marrow Transplantation, UCSF Benioff Children's Hospital, San Francisco, CA; [7]Children's Mercy Kansas City, Kansas City, MO

Background/Aims: Asparaginase therapy for patients with acute lymphoblastic leukemia/lymphoma (ALL/LBL) can be limited by hypersensitivity reactions (HSR). The objective of this study was to determine: 1) whether premedication with histamine-1 (H1) and histamine-2 (H2) blockers prior to pegaspargase (PEG) decreases HSR, and 2) the incidence and risk factors for HSR.

Methods: From 2019 to 2022, prospectively enrolled patients (n=144) with T or B-ALL/LBL at six centers in the United States received H1 (diphenhydramine) and H2 blockers (ranitidine or famotidine) prior to PEG. Patients were monitored for HSR and therapeutic drug monitoring (TDM) was performed.

Results: The mean age at diagnosis was 8.6 years (range 1.1-23.9). There were 61.8% males, 86.8% B-ALL, 52.5% Hispanic, 36.1% NCI High Risk (HR), and 19.4% obese patients. Twenty-eight patients (19.4%) experienced grade ≥2 HSR. The mean age of patients with HSR was 10.5 years, 15 (53.6%) were Hispanic and 10 (35.7%) were obese. The incidence of HSR was increased with obesity (p=0.020), older age ≥10 years (p= 0.036), NCI HR (p=0.013) and non B-ALL (p=0.048). In patients ≥10 years, being non-obese was protective against HSR (OR 0.23, 95% CI[0.05, 0.96], p=.045); whereas obesity amplified their HSR risk, (OR 12.50, 95% CI[2.68, 58.82], p=.002). In younger patients, the impact of obesity on risk of HSR was not significant (p=.629). Six of the 10 patients who experienced HSR were successfully rechallenged with PEG, through desensitization. TDM detected silent inactivation in 4 (2.8%) and accelerated clearance in 3 (2.1%) patients.

Conclusion: We observed that 19.4% of patients experienced HSR, indicating premedicating with H1 and H2 blockers may not be beneficial in lowering HSR. There was a tendency for those with HSR to be older, obese, NCI HR and non B-ALL. Although desensitization is one reasonable approach to allow continuation of PEG, alternative methods are needed to mitigate HSR.

This study was funded by a grant from Servier Pharmaceuticals.

POSTER #37

Identification of Patients Experiencing Child Trafficking in the Pediatric Emergency Department

Carolina Amaya[1], Shelby Shelton[1], Brooke Valdez[1], Kellie Bacon[1], Lisa Chambers[1], Jennifer Hayakawa[2], Theodore Heyming[1]

[1]Children's Hospital of Orange County Department of Emergency Medicine
[2]Children's Hospital of Orange County Department of Nursing & Innovation

Background: Child trafficking is the recruitment of a child for labor or services using force or coercion. Child victims in the healthcare system often remain unidentified. As frontline care providers, Emergency Departments (ED) present an ideal setting for identifying and intervening in child sex (CST) and labor trafficking (CLT).

Purpose: To review retrospective pilot data from a new child trafficking screening tool. Methods: Retrospective review of patients aged 12+ with risk factors who received the screening tool in an urban pediatric ED with over 100,000 yearly visits. Descriptive statistics were used to describe the cohort.

Results: We analyzed 412 patient surveys completed 2/8/23-12/5/23. Most patients (66.7%) were female, mean age was 14.7±1.8 years; most patients (57.3%) were Hispanic ethnicity, and white race (64.1%). The most frequent chief complaint was mental health complaint (45.1%), followed by ingestion of drug/alcohol (14.6%); 28.9% of patients visited the ED for a medical complaint. We found that 35.2% of patients reported ever having sex, 53.4% experimented with drugs or alcohol, 13.1% experienced intimate partner violence, 12.4% had been raped, 3.6% traded sex for money, 1.9% had worked without being paid, and 3.4% felt they could not leave the place they worked. As a result of the screening, patients received consults from Mental Health Evaluation Services, social work consults, and Child Protective Services was notified for three patients. Neither age, gender, nor mental health vs. medical complaint were associated with being raped, trading sex for money, or working without being paid.

Conclusions: These findings indicate that there is no distinct demographic group more likely to screen positive for the at-risk experiences evaluated through this screening tool, highlighting the need for a broad screening approach for CST and CLT in the pediatric ED.

POSTER #38

Achieving Fast Assessment and Stroke Triage in Kids (FAST-Kids)

Kellie Bacon, MPH; Rachel Pearson, MD; Nancy Hills PhD; Shelby Shelton, MPH; Tatiana Moreno, BS; Maria Kuchherzki; Theodore Heyming, MD; Carl Schultz, MD; Heather Fullerton, MD, MAS

BACKGROUND: Pediatric stroke, while a top 10 cause of mortality among U.S. children, remains underrecognized. Identification of pediatric stroke starts with the community and emergency medical services (EMS) recognizing acute neurological symptoms (ANS) in a child as a medical emergency and appropriately triage to facilitate urgent imaging.

PURPOSE: In this study we examined timing and predictors of neuroimaging among children with EMS activation for ANS, and arrival modality and symptoms of children with acute stroke presenting to the emergency department (ED).

METHODS: This is a retrospective study of patients 1-14 years with an EMS activation for ANS who were transported to CHOC ED from 1/2019-6/2023. We identified all out-of-hospital strokes admitted to CHOC and determined the proportion of stroke cases presenting via EMS.

RESULTS: Of 3888 children with EMS activations for ANS, 695 (17.9%) had neuroimaging: 570 (82.0%) had CT scan only; 125 (18.0%) also had MRI. The median times to CT and MRI were 2.29 (1.56, 3.21) and 26.8 (16.3, 43.8) hours, respectively. Factors associated with neuroimaging included age ≥5 years ($p<0.001$) and known medical conditions associated with increased stroke risk ($p<0.001$). All patients with an EMS primary impression code of stroke had neuroimaging. We identified 43 admissions for out-of-hospital acute stroke. Those presenting as EMS activations had primary symptom and impression codes as seizure, altered mental status, loss of consciousness, and headache and those who presented as a walk-in, the most common chief complaints were head injury and general neurologic complaint.

CONCLUSIONS: Most EMS activations for ANS were not imaged; of those who were, many had only CT, not MRI, the preferred modality for diagnosing pediatric ischemic stroke. Our findings call for more education of the public and prehospital/ED providers on pediatric stroke, as earlier consideration may expedite diagnosis and increase opportunities for intervention.

POSTER #39

Pediatric Pain and Maladaptive Behavioral Changes Following Laceration Repair Procedures in the Emergency Department

Helen Lee BA[1], Sarah R. Martin PhD[1,2,3], Theodore W. Heyming MD[1,4], Kellie Bacon MPH[1], Shelby Shelton MPH[1], Carolina Amaya[1], Hailey Worden MPH[1], Anahita Darabpour MPH[1], Zoe Ta-Perez[1], Michelle A. Fortier PhD [1,3,5], Zeev N. Kain MD, MBA[1,2,3]

[1] Children's Hospital of Orange County, Orange, CA
[2] Department of Anesthesiology and Perioperative Care, University of California, Irvine, CA
[3] Center on Stress & Health, University of California, Irvine, CA
[4] Department of Emergency Medicine, University of California Irvine, CA
[5] Sue & Bill Gross School of Nursing, University of California, Irvine, CA

Background: Each year, millions of children sustain injuries and undergo laceration procedures in the emergency department (ED). Post-discharge recovery following laceration procedures is understudied, but previous perioperative research indicates that maladaptive behavioral changes are common following such stressful and painful episodes.

Purpose: This study examined post-discharge pain and maladaptive behavioral changes following pediatric laceration repair in the ED.

Methods: Participants included 102 children 2-12 years old (M_{age}=5.72±2.64; 35.3% female, 51% Latinx) undergoing laceration repair. Demographics, treatment data (e.g. anxiolytic medication), and caregiver-reported child pre-procedural and procedural pain (numeric rating scale [NRS]) were collected. On post-discharge days 1, 3, 7, and 14, caregivers reported children's pain and new onset behavioral changes (e.g., separation anxiety) via the Post Hospitalization Behavior Questionnaire-AS. Descriptive, correlational, and mean difference analyses were conducted.

Results: 47.1% of children exhibited new onset maladaptive behavior changes over the two-week post-discharge period endorsed clinically significant levels of anxiety in the ED. Child pain and behavior change were positively correlated on post-discharge days 1, 3, and 7 (r=.37, r=.39, r=.20, p's<.01). Children exhibiting maladaptive behavioral changes during the post-discharge period had higher pain on post-discharge days 1 and 3 (Z=-3.68, Z=-2.94, p's<.01), and reported higher post-procedural pain in the ED (Z=-2.22, p<.05). Higher pain on post-discharge day 1 was the only variable independently associated with an increased likelihood of maladaptive behavioral change (OR=1.61, p=.04). No other variables, including demographics, anxiolytics medication, or laceration size, were associated with post-discharge outcomes.

Conclusions: A considerable portion of children experienced pain and maladaptive behavior changes following laceration repair in the ED. Post-discharge pain was positively associated with maladaptive behavioral changes, with pain on the day following discharge being a key predictor. Further examination of multilevel correlates of pain and post-discharge recovery is needed to improve outcomes following ED procedures.

POSTER #40

Age, Adverse Childhood Experiences, and Healthcare Utilization

Shelby K Shelton MPH[1], Charles V. Golden DO[1], Raymen R. Assaf MD[1,2], MPH, Ricardo Aguilar MS[1], Tatiana Morena BS[1], Heather Huszti PhD[1], Michelle Fortier PhD[1,3], Theodore Heyming MD[1,2], Uma Rao MBBS[1,2], Louis Ehwerhemuepha PhD[1,2], Michael Weiss DO[1]

[1]Children's Hospital of Orange County
[2]School of Medicine, University of California, Irvine
[3]Sue and Bill Gross School of Nursing, University of California, Irvine

Background: Adverse childhood experiences (ACEs) are events occurring in a child's social or physical environment that increase the risk for physical, psychological, or developmental harm. ACEs include (but are not limited to) physical, sexual, or emotional abuse; neglect; and household challenges, such as caregiver mental illness and substance misuse. Both pediatric and adult studies demonstrate an association between high ACEs and utilization of acute health care resources, such as sick visits, urgent care, emergency department (ED) use, and hospitalization.

Purpose: To assess whether the association of ACEs with pediatric health care utilization differs by age.

Methods: In this retrospective cohort study, we included patients completing primary care ACEs screening between January 2020 and September 2021. Pediatric ACEs and Related Live Events Screener Part 1 scores were categorized to 0 (none), 1 to 3 (low), or 4 (high). Two multivariable logistic regression models assessed ED and inpatient utilization across all ages six months after screening.

Results: Among 37,315 patients, 15.7% visited the ED and 2.5% were hospitalized within six months of ACEs screening. Using no ACEs as the reference variable, infants and toddlers with any ACEs had lower odds of ED and inpatient utilization, whereas older children with any ACEs had higher odds of ED (age-low ACEs: 0.04, $p<0.001$; age-high ACEs: 0.15, $p<0.001$) and inpatient (age-low ACEs: 0.06, $p<0.001$; age-high ACEs: 0.15, $p<0.001$) utilization and increased with each successive year of age.

Conclusions: The association of ACEs with health care utilization is dependent on age and is more complex than previously described. These trends may inform specific therapeutic strategies for pediatric patients by age.

POSTER #41

Implementation of a Child Maltreatment Screening Tool in the Prehospital Setting

Shelby Shelton MPH[a], Makenzie Ferguson RN, BSN[a], Shelley Brukman RN, MSN[a], Kim Zaky RN, DNP[a,b], Bryan A. Lara[a,c], Chloe Knudsen-Robbins MD[d], Carolina Amaya[a], Theodore Heyming MD[a,e]

[a] Children's Hospital of Orange County (CHOC Children's), Department of Emergency Medicine; Orange, CA; [b] University of California, Irvine; Department of Surgery, Division of Trauma, Burns, Critical Care, and Acute Care Surgery; Irvine, CA; [c] Johns Hopkins University, School of Nursing, Baltimore, MA; [d] Department of Emergency Medicine, University of Cincinnati College of Medicine, Cincinnati, OH; [e] University of California, Irvine; Department of Emergency Medicine; Irvine, CA

Background: Emergency Medical Services (EMS) clinicians are in a unique position to screen for child maltreatment as they are often the first point of contact with the healthcare system. However, EMS training regarding the signs and symptoms of child maltreatment is lacking. Although several child maltreatment screening tools have been developed, there are no studies describing a prehospital screening aid.

Purpose: The objective of this study was to pilot the implementation of a prehospital child maltreatment screening tool and investigate EMS perceptions surrounding its use.

Methods: We completed a mixed methods convergent study design, with data collection spanning June 2021-June 2022. We developed a child maltreatment screening tool "Shield" for the prehospital setting by adapting P-CAST, a screening tool developed for use in the ED. Shield was integrated into the EMS electronic Patient Care Report (ePCR) for participating agencies. Demographics, ED evaluation, and outcomes, were collected for patients who underwent Shield evaluation. Additionally, EMS clinicians completed self-assessment surveys and participated in focus groups to provide feedback on their experience using Shield.

Results: Participating EMS agencies evaluated 1,054 eligible patients during the study period, June 2021-June 2022. Of these, Shield screenings were initiated on 948 patients and completed on 753. Among patients for whom a Shield evaluation was started, 32 (3.4%) screened positive. Of these, 20 patients were transported to the primary study institution; in the ED 10 patients underwent additional child maltreatment evaluation. Pre-implementation surveys suggested a majority (77.2%) of EMS clinicians desired a child abuse screening tool integrated into the ePCR and post-implementation focus group data demonstrated EMS clinicians found Shield to be well integrated into their ePCR.

Conclusion: Shield screens were initiated on nearly 90% of pediatric patients evaluated by EMS during the study period, suggesting that this tool has the potential to help standardize child maltreatment screening in the prehospital setting.

POSTER #42

Climate Anxiety in Young People: at the Generational Crossroads of Environmental and Mental Health

Zoe Ta-Perez[1], Rammy Assaf[1], Shelby K. Shelton[1], Kellie Bacon[1], Jun Wu[2]

[1]Children's Hospital of Orange County Department of Emergency Medicine
[2]University of California, Irvine School of Public Health

Background: Healthcare providers are expected to address climate change as a social determinant of health, as outlined by the American Academy of Pediatrics. Climate change can impact psychological well-being through perceived threats from climate events and concerns about future risks.

Purpose: This preliminary analysis explores the relationship between generalized anxiety and climate anxiety among young people and their caregivers in a pediatric emergency department (ED).

Methods: This study involved patients aged 12-17 and their caregivers at CHOC's ED. Patient-caregiver surveys include the Climate Change Anxiety Score (CCAS), which assesses cognitive-emotional and functional impairment subscales, and the Generalized Anxiety Disorder-7 (GAD-7).

Results: Data from 169 adolescents and 144 caregivers (124 dyads) were analyzed. Most participants were Hispanic ethnicity (39%) or White race (17%); many participants (32%) preferred not to disclose their race. Adolescents were 52% female and 48% male, with a median age of 15.2 years (mean=15.1, SD=1.7, range=12-17.9). Six adolescents had moderate to high climate anxiety (CCAS score ≥3). Median CCAS scores showed no significant differences between adolescents and caregivers or among dyads ($p=0.40$). Significant differences were observed in GAD-7 scores between adolescents and caregivers ($p<0.001$) and among dyads ($p=0.002$). CCAS and GAD-7 scores were significantly correlated within both groups ($p<0.001$).

Conclusion: This study is the first to use the validated CCAS in a pediatric ED, showing that climate anxiety is a component of generalized anxiety. Our findings may support the integration of climate-related mental health issues into routine pediatric screenings.

POSTER #43

Enhancing Medical Device Care Education via Virtual Reality: A Paradigm Shift

Jamie Janchoi, BS; Ashish Chogle, MD, MPH; Wanda Rodriguez, RN; Nicole Callas, RN; Tammy Tran, PNP Jennifer Hayakawa, DNP; Sandeep Godambe; Chloe Thomas, PA-C , Elisa Ornelas, BA

Children's Hospital of Orange County (CHOC), Orange, CA

Background: Each year, approximately one million medically complex children, dependent on devices such as gastric tubes, tracheostomy tubes, and central venous lines, transition from US hospitals to home care. A significant percentage of these children face unnecessary emergency visits and readmissions due to complications with their medical devices. A core issue identified is the lack of parental self-efficacy in handling their child's medical devices. Currently, parents are taught about medical device use and care by bedside nurses in most hospitals. Parents are then sent home with printouts of instructions. This educational strategy usually leads to inconsistent information being provided to patients and their families. Parents may be overwhelmed, stressed, or fatigued, making it difficult to absorb and retain information about medical device care. To solve the problem with informed consent and adequate medical education for parents, we created a virtual reality-based education platform (CareXR) for children with medical devices. This platform includes procedure videos, educational videos by nurses, and virtual simulation modules where parents can practice the skills in a virtual environment.

Objectives: Our study aims to determine if the application of a novel virtual reality (VR) platform, designed for educating parents about medical device care, has a transformative impact on several outcomes in parents of children who had percutaneous endoscopic gastrostomy (PEG) tube placement. These include parental anxiety levels, competency, transition from hospital to home, post-discharge coping, and the incidence of device complications, compared to our current standard of care.

Methods: Parents of children between ages 0 and 21 years undergoing a PEG tube placement at CHOC Children's Hospital participated in the study. Following the procedure, parents were randomized to either an experimental group (device care education via nurse instruction and VR platform) or a control group (standard nurse-led device care education). A series of questionnaires and scales were utilized at various time points pre- and 1-month post-discharge to measure parental anxiety, competency, coping difficulty, ease of transition to home from the hospital, and the incidence of PEG tube complications.

Results: So far 42 parents (22 experimental and 20 control) have completed the study. These were parents of 23 male children and 19 female children, mean age of 24.03 months. Significant improvement was observed in the experimental group compared to the control group, specifically, reduction in anxiety levels ($p=0.01$). The experimental group experienced heightened competency, smoother transitions from hospital to home, and fewer instances of device complications and post-discharge health care utilization.

Conclusions: Initial results signify that our VR intervention offers a transformative approach to inpatient education. It enhances parental capabilities in managing their child's medical devices, reduces anxiety, improves discharge experiences, and decreases complication rates, thus constituting a potential paradigm shift in pediatric care. Further participant recruitment is in progress to fortify our conclusions.

POSTER #44

Utility of the Prehospital Pediatric Assessment Triangle

Anahita Z. Darabpour[1], Shelby K. Shelton[1], Kellie Bacon[1], Shelley Brukman[2], Theodore Heyming[1], Zeev N. Kain[3,4]

[1]Children's Hospital of Orange County Department of Emergency Medicine
[2]University of California, Irvine Department of Emergency Medicine
[3]University of California, Irvine Department of Anesthesiology and Perioperative Care
[4]University of California, Irvine Center on Stress and Health

Background: Emergency Medical Services (EMS) providers often report a lack of training and experience with children, leading to discomfort with assessment and treatment. The Pediatric Assessment Triangle (PAT) was designed to provide a rapid and standardized method of pediatric assessment that is completed based solely on visual and auditory observation without additional tools. Despite widespread adoption, literature remains limited, and only one study has investigated the use of the PAT in the field.

Purpose: We aimed to address this gap in the literature by further investigating the use of PAT in the prehospital setting

Methods: We completed a retrospective chart review of patients aged 1 day-14 years transported to the CHOC Emergency Department (ED) between 10/1/22-11/7/23 who had all three PAT variables collected. Descriptive statistics, independent sample T-test, and Chi-square tests were used to describe the cohort.

Results: We reviewed 3,148 runs from nine different 9-1-1 Paramedic Providers in the county. 61.8% of the 9-1-1 calls came from a residential home and the mean age was 14±5.4 years. The most frequent abnormal PAT observation was appearance (23.8%), 13.9% of the patients were abnormal for breathing, and 5.2% were abnormal for circulation. Combined, 5.7% of patients were abnormal for at least two variables and 1.8% for all three variables. Patients with younger age were more likely to have an abnormal appearance (p<0.001), breathing (p<0.001), and circulation (p<0.001). EMS providers spent an average of 2.5 more minutes at the scene with patients who had abnormal breathing (p<0.001) and 8.3 minutes with abnormal circulation (p<0.001), but there was no difference between patients with abnormal vs. normal appearance (p=0.118).

Conclusions: These preliminary data indicate that the PAT variables may support EMS provider decision-making, primarily with younger patients. Future analyses will include ED variables and a granular analysis for each item on the PAT scale.

POSTER #45

Comparing Pediatric Care Transition Risk Predictors: BLSS and the TRxANSITION Index™

Reny Partain, LCSW, MPH, Radha Nagarajan, PhD, Wendy N. Gray, PhD, Erin Benekos, FNP-C, Kenneth Grant, MD, Steven Martel, MD, & Michael Weiss, DO

Background: Bob's Level of Social Support (BLSS) is a measure of social complexity that has been used in clinical practice to identify families in need of care management support. It has also been informally used to assess risk factors that may negatively impact a patient's transition to adult care. Although used clinically, the tool itself lacks clinical validation. In this study, we examine the validity of the BLSS in predicting transition readiness.

Methods: Using our hospital's transition registry, we searched for adolescent/young adult patients who had both a completed BLSS and TRxANSITION Index™ in their medical record. Data from 413 patients were extracted and examined. Linear regression and robust linear regression examined the relationship between the total scores of these measures.

Results: The BLSS Total Risk Score was negatively associated with and the TRxANSITION Index™ Total Score (slope = -0.39, $p < 10^{-3}$, $R^2 = 0.27$), suggesting that transition readiness is lower in youth (Age:19±2.8yrs) with higher identified risk. Robust linear regression confirmed the decreasing trend (slope = -0.41, $p < 10^{-3}$). While the relationship between the BLSS and TRxANSITION Index™ was significant, the general effect sizes were small and the scatter plot of the association between these variables had considerable noisiness.

Conclusion: Social complexity, as measured by the BLSS, is negatively associated with transition readiness. While the BLSS offers valuable insight, the noticeable variability in this association suggests the need for careful consideration when using BLSS as a surrogate for TRxANSITION Index™ in predicting care transition risk. This provides opportunities for refining how we evaluate and utilize these measures to enhance predictive accuracy and support effective transitions in care.

POSTER #46

Optimal Agent and Timing of Surgical Antibiotic Prophylaxis and Surgical Site Infection in Pediatric

Shasta Erickson, PharmD; Negar Ashouri, MD; Melissa Powell, MSN; Laurie Moore, BS; M. Tuan Tran, PharmD

Background: Surgical site infections (SSI) make up ~20% of all healthcare-associated infections (HAI). Beta-lactams (BL) including cefazolin (CFZ) and cefoxitin (CFO) are recommended as surgical antibiotic prophylaxis (SAP). Cephalosporins are often avoided in patients with reported BL allergies despite cross-reactivity with penicillin at <1%. Guidelines recommend giving SAP 60 min prior to incision. CFZ and CFO pharmacokinetics (PK) exhibit rapid time to peak serum level and short elimination half-life. We aim to evaluate differences in SSI rate in patients who received CFZ or CFO vs. non-BL alternatives (NBL) and administration of preop BL dose at different time interval prior to incision.

Methods: Retrospective observational study included patients aged 0 – 17 years who received SAP at CHOC from Jul 2021 – Jun 2023. Outcomes include differences in SSI rate within 90 days of surgery between patients receiving recommended BL (CFZ, CFO) vs NBL (clindamycin, gentamicin, metronidazole, vancomycin). Rate of SSI in patients who received pre-op CFZ or CFO at different time interval (0-20 min, 21-40 min, 41-60 min) will be examined.

Results: Median age was 9.0 years [IQR: 3.0, 14.0], 58.8% males. Wound classification is as follows: 37.6% (Class I), 34.1% (Class II), 22.9% (Class III), and 5.1% (Class IV). Overall SSI occurred in 1.0% (92/9054) of procedures. Beta-lactams were used as SAP in 96% of procedures, with 89.8% receiving pre-op dose within 20 min of incision. Age <1 year, use of NBL SAP and administration of BL SAP >20 min from incision was associated with significant increase in SSI ($p<0.05$, Table 1). No difference in SSI rate observed in patients who continued receiving post-op SAP for class I-II procedures.

Conclusions: Our study supports the continued use of CFZ, CFO compared to NBL for SAP. Pre-op doses of CFZ or CFO within 20 min of incision time were associated with decreased risk of SSI, likely due to their PK characteristics.

POSTER #47

Sutured versus Sutureless Closure in Neonatal Gastroschisis: A Two-institution Study

Hough M[a,b], Giron A[a], Martino A[a,d], Gonzalez A[a], Dinh P[c], Schomberg J[c], Flyer Z[a], Aziz H[a], Hoang V[a], Yu P[a,b], Goodman L[a,b], Ahmad I[a], Uy C[b], Guner Y[a,b], Awan S[a,b].

[a]Division of Pediatric Surgery, Children's Hospital Orange County, Orange, California, USA
[b]Department of Surgery, University of California Irvine Medical Center, Orange, California, USA
[c]Children's Hospital Orange County (CHOC) Research Institute, Orange, California, USA
[d]Department of Surgery, Montefiore Medical Center, New York City, New York, USA

BACKGROUND: The gastroschisis defect has historically been closed with fascial sutures after bowel reduction. Sutureless repair utilizing the umbilical cord has gained popularity since its introduction in 2004. Current literature is ambiguous regarding in-hospital outcomes such as time to full feeds and length of stay (LOS).

AIMS: We sought to compare sutured versus sutureless closure, hypothesizing lower rates of intubation, shorter LOS, and quicker advancement to feeds in sutureless closure.

METHODS: All cases of gastroschisis were examined within a single surgeon group at two institutions from 2014-2022. Complex gastroschisis was excluded. Sutured versus sutureless closure were compared using bivariate analyses.

RESULTS: Out of 59 repairs, 37 (62.7%) were sutured and 22 (37.3%) were sutureless. The median LOS was 30 days for sutured closure versus 33.5 days for sutureless closure (p=0.735). Median days to first enteral feed was 11 in both groups (p=0.531), and 11 versus 12 days (p=0.265) to reach full feeds (120 Cals/kg/d) in sutured vs sutureless groups. 100% of sutured closures required intubation versus only 10 (45.4%) of the sutureless closures (p<0.0001). The indication for intubation in the sutureless group was respiratory failure.

CONCLUSIONS: This nine-year review of neonates with gastroschisis demonstrated no significant differences in LOS, days to first feeds, or days to full enteral feeds in sutured versus sutureless closure. All sutured closures require intubation for general anesthetic administration, whereas sutureless closures have the potential to avoid intubation. Future research is needed to determine if long term functional and cosmetic outcomes differ between the two closure methods.

POSTER #48

Deaths due to isolated extremity gunshot wounds in children and young adults

Michelle Hough MD[a,b], Alice M. Martino MD[a,b], Andreina Giron MD[a,d], John Schomberg PhD[c], Jeffry Nahmias MD MHPE[b,c], Peter T. Yu MD MPH[a,b], Laura F. Goodman MD MPH[a,b].

[a]Division of Pediatric Surgery, Children's Hospital Orange County, Orange, California, USA
[b]Department of Surgery, University of California Irvine Medical Center, Orange, California, USA
[c]Children's Hospital Orange County (CHOC) Research Institute, Orange, California, USA
[d]Department of Surgery, Montefiore Medical Center, New York City, New York, USA

BACKGROUND: Isolated extremity gunshot wounds (GSW) cause a significant number of deaths annually despite exsanguinating extremity hemorrhage being potentially treatable, and thus the deaths preventable.

AIMS: We sought to compare differences between children and young adult decedents of isolated extremity GSW (E-GSW) to decedents from other site GSW (O-GSW) in terms of demographics, incident circumstances, and disposition.

METHODS: A retrospective analysis of the National Violent Death Reporting System (NVDRS) database was performed for years 2012-2021. E-GSW decedents were compared to O-GSW decedents in patients 0-25 years old. Variables analyzed included race, sex, incident, EMS (Emergency Medical Services) response, transportation to the emergency department (ED) and survival times. Analysis was conducted using bivariate inferential statistics: chi square and Wilcoxon rank-sum.

RESULTS: Of 40,746 firearm injuries, 39,878 (97.9%) were O-GSW and 868 (2.1%) were E-GSW. African Americans comprised the majority of both cohorts but more commonly E-GSW (75.3% vs 66.6%, $p<0.0001$). More E-GSW were male (90.3% vs 87.1%, $p=0.006$) and in single homicides (89.9% vs 86.1%, $p=0.0001$). Both groups were admitted to the hospital at similar rates (12.1% vs 12.8%, $p=0.34$). 72.0% E-GSW survived minutes after injury and 20.9% of E-GSW decedents survived hours. 66.1% of E-GSW was transported to the ED compared to 53.4% of O-GSW. A subgroup analysis was performed for decedents <18 years old and mirrored these trends.

CONCLUSIONS: This national analysis demonstrated disparities in race and sex among young decedents of isolated E-GSW compared to O-GSW. One-fifth of decedents with E-GSW survived hours and had higher transport rates to the ED, representing a window of intervention to prevent deaths. Targeted intervention programs such as Stop the Bleed training may help improve survival after isolated extremity gunshot wounds.

POSTER #49

Pre-hospital Tourniquet Use in Adolescent and Pediatric Traumatic Hemorrhage: A National Study

Alice M. Martino MD, Andreina Giron MD, John Schomberg PhD, Makenzie Ferguson, Jeffry Nahmias MD MHPE, Sigrid Burruss MD, Yigit Guner MD, Laura F. Goodman MD MPH

Background: Tourniquet placement (TP) is a crucial intervention to control hemorrhage, but limited literature exists for use in children. This study aimed to evaluate the effectiveness of tourniquet application by different providers (Emergency Medical Services (EMS), police, and bystanders), hypothesizing equivalent impact on outcomes for pediatric trauma patients for all providers.

Methods: Data from the National EMS Information Systems (NEMSIS) 2017-2020 was used to examine patients 0-19 years old and assess the outcomes of tourniquet application. We considered demographics, procedure success, timing of TP relative to EMS arrival, revised trauma score (RTS), and improvement in acuity. Multivariable logistic regression models were employed to predict initial acuity and likelihood of acuity improvement after TP, while accounting for patient and provider-related variables.

Results: 301 patients were included with a median age of 17 and 86.7% male. TP by any provider before EMS transport arrival was associated with reduced odds of critical acuity upon EMS arrival (OR=0.84, CI=0.76-0.94, p=0.003). After EMS arrival, bystander- and police-placed tourniquets were associated with increased odds of improved acuity compared to EMS-placed tourniquets (OR=1.90, CI=1.06-3.41, p=0.03). There was only one TP failure (0.43%) in the EMS group.

Conclusion: Early TP for pediatric traumatic hemorrhage is crucial. Failures were rare. Placement by bystanders and police were associated with improved acuity when controlling for other factors including RTS and EMS arrival time. These findings emphasize the importance of training on TP for all providers in prehospital settings.

POSTER #50

Disparities in Access to Neonatal and Pediatric ECMO Services in the United States

April Carlson MD[a,b], Andreina Giron MD[a,c], Zoe E. Flyer DO[a,d], Habiba Aziz MS[a], John Schomberg PhD[a], Yigit S. Guner MD[a,b], Laura F. Goodman MD[a,b], Peter T. Yu MD MPH[a,b].

[a]Division of Pediatric Surgery, Children's Hospital Orange County, Orange, California, USA
[b]Department of Surgery, University of California Irvine Medical Center, Orange, California, USA
[c]Department of Surgery, Montefiore Medical Center, New York City, New York, USA
[d]Department of Surgery, Community Memorial Hospital in Ventura, California, USA

Objective: This study assesses geographic and socioeconomic disparities in the accessibility of ECMO (Extracorporeal Membrane Oxygenation) centers across the United States, focusing on neonatal and pediatric populations, particularly in rural and low-income areas.

Methods: We analyzed data from the U.S. Census and location data of ECMO centers from the Extracorporeal Life Support Organization (ELSO). Population densities of children aged 0-19 were mapped against locations of ECMO centers to identify coverage gaps. Socioeconomic variables were assessed using the American Community Survey, focusing on income levels and rural versus urban residency.

Results: Approximately 31.4% of the population resides ≥ 50 miles from an ECMO center with a racial distribution 69.95% Caucasians and 79.9% non-Hispanic ($p<0.0001$). ZIP codes ≥ 50 miles have lower median incomes ($59,767 vs. $62,066, $p<0.0001$), higher prevalence of poverty (12.90% vs. 12.41%, $p<0.0001$) and unemployment (2.90% vs. 2.70%, $p<0.0001$). Area Deprivation Index (ADI) significantly worsens with increasing distance from ECMO centers (mean ADI 60 vs. 56, $p<0.0001$). Higher education levels, including Bachelor and Graduate degrees, are predominantly located within 50 miles of ECMO centers ($p<0.0001$). Mapping shows more areas without proximate to ECMO services in the midwestern and southwestern U.S.

Conclusion: disparities exist in the availability of ECMO services for neonates and children in the U.S., predominantly affecting rural and economically disadvantaged communities. These findings suggest a need for strategic placement of new ECMO facilities to ensure equitable health care access.

POSTER #51

THE DEVELOPMENT OF A MULTIDISCIPLINARY FETAL CENTER INCREASES INSTITUTIONAL CONGENITAL LUNG MALFORMATIONS (CLM) VOLUME

April Carlson[4,6], Andreina Giron[4,7], Shelby Daughetee[1], Peter T. Yu[4,6], John Schomberg[5], Martha A Monson[2], Anne M. Kennedy[3], Paula J. Woodward[3], Janice LB Byrne[2], Katie W. Russell[1], Stephen J. Fenton[1]

[1]Department of Surgery, University of Utah School of Medicine, Salt Lake City, Utah, USA
[2]Department of OB/GYN, University of Utah School of Medicine, Salt Lake City, Utah, USA
[3]Department of Radiology, University of Utah School of Medicine, Salt Lake City, Utah, USA
[4]Division of Pediatric Surgery, Children's Hospital of Orange County, Orange, California, USA
[5]Division of Trauma, Children's Hospital of Orange County, Orange, California, USA
[6]Department of Surgery, University of California Irvine Medical Center, Orange, California, USA
[7]Department of Surgery, Montefiore Medical Center, New York City, New York, USA

Introduction: Congenital lung malformations (CLM) are often diagnosed on prenatal ultrasound. In 2016, a multidisciplinary fetal center was developed at a freestanding Children's Hospital with the initiation of clinic including Maternal Fetal Medicine physicians later that year. The purpose of this study was to review the impact of this center on the diagnosis and treatment of CLMs within the referral region.

Methods: Patients with CLM at Primary Children's (2012-2022) and CHOC (2013-2023) were stratified into two groups: those diagnosed before establishment of an institutional fetal center and those after. Kruskal-Wallis and Chi-squared tests were used for statistical analysis.

Results: 227 children underwent CLM resection. Median maximum CVR was 0.79 [0.38,1.4]. 84.1% were found prenatally with 68.7% evaluated in the fetal center. Median gestational age was 37.5 weeks [39,3.2] and birth weight 3109 grams [2729,3289]. Resection occurred at 472 days [168,1077]. 76.9% underwent thoracoscopy with LOS of 6.9 days [3,15], 2.19% required transfusion, 11.4% experienced post-operative complications, and 97.8% survived. After its development, mothers were more likely to be seen in the fetal center (82.8% v. 13%, $p<0.0001$), have a calculated CVR (63.5% v. 15.2%, $p<0.0001$), and receive betamethasone (30.9% v. 2.17%, p=<0.33). Births per year increased three-fold (16 v. 5, $p<0.0001$) and resections per year increased five-fold (14 v. 3, $p<0.0001$).

Conclusions: Development of a multidisciplinary fetal center led to a significant increase in volume for CLM at two separate institutions. Prenatal care was more uniform and surgical outcomes were excellent.

POSTER #52

Fetal Care Deserts: Disparities in Access to Expert Care for Congenital Anomalies

Andreina Giron MD[a,e], Habiba Aziz MS[a,b], John Schomberg PhD MPH[a,b], Zoe E. Flyer DO[f], Laura F. Goodman MD[a,b], Yigit S. Guner MD[a,b], Erin E. Perrone MD[c,d] , Peter Yu MD[a,b].

[a]Division of Pediatric Surgery, Children's Hospital Orange County, Orange, California, USA
[b]Department of Surgery, University of California Irvine Medical Center, Orange, California, USA
[c]Division of Pediatric Surgery, C.S. Mott Children's Hospital, Ann Arbor, Michigan, USA
[d]Department of Surgery, University of Michigan, Ann Arbor, Michigan, USA
[e]Department of Surgery, Montefiore Medical Center, New York City, New York, USA
[f]Department of Surgery, Community Memorial Hospital in Ventura, California, USA

Objective: This study investigates socioeconomic disparities in access to fetal care for congenital anomalies in the United States.

Methods: Employing American Community Survey data, ZIP codes of women of childbearing age (WOCBA) (15-50 years) were heatmapped using ArcGIS, a secure mapping and spatial analysis platform. Using NAFTNet centers as proxies for expert fetal care, ZIP codes were stratified by distance. Descriptive statistics and Chi-square tests examined socioeconomic factors for distance stratification (<50 miles vs. >50 miles). Area Deprivation Index (ADI)for each ZIP code was analyzed using the Wilcox rank sum test. Lastly, using the product of WOCBA * fertility rate for each ZIP code, a rank order of fetal center need (FCN) for fetal care deserts was created.

Results: 43.16% of WOCBA live <50 miles from a NAFTNet center, while 56.84% live farther. Median income was $76,706 for areas <50 miles vs. $53,971 for areas >50 miles (p<.0001). Poverty, unemployment, uninsured status, and ADI were higher for areas >50 miles (p<.0001). Lakewood, NJ, Antioch, TN, and Lawrenceville, GA scored highest in FCN.

Conclusion: Socioeconomic disparities and geography are barriers to accessing expert fetal care. Strategic expansion of fetal centers may improve equitable access for expecting families.

POSTER #53

Cancer and Trauma Collide: Injury Patterns and Outcomes in the Pediatric Trauma Patient with Cancer

Andreina Giron[2], Zoe Flyer[2], Brittany Sullivan[1], John Schomberg[2], Josephine Haduong[3], Sourav K. Bose[4,5], Yigit S. Guner[1,2], Maryam Gholizadeh[1,2], Peter T. Yu[1,2]

[1.] University of California Irvine Medical Center, Department of Surgery, Orange, CA
[2.] Children's Hospital of Orange County, Division of Pediatric Surgery, Orange CA
[3.] Children' Hospital of Orange County, Division of Oncology, Hyundai Cancer Institute, Orange, CA
[4.] Center for Fetal Research, Division of General, Thoracic and Fetal Surgery, Children's Hospital of Philadelphia, Philadelphia, PA
[5.] Leonard Davis Institute for Health Economics, Philadelphia, PA

Purpose: There is a lack of data regarding pediatric trauma patients with a cancer diagnosis. The purpose of our study is to analyze demographics, ED admissions, procedures and characteristics pertaining to this population.

Methods: The NTDB database was queried from 2007 to 2015. This study collected 1,146,422 trauma admissions under the age 19. Of these patients, 176 were identified with a cancer diagnosis, of which 152 (86.3%) had received chemotherapy within 30 days of their trauma admission.

Results: The mean age of trauma patients with cancer was 9.1 years old vs. 10.5 years old for non-cancer patients ($p<0.05$). Injury severity in cancer patients was not significantly different from non-cancer patients ($p=0.42$), but cancer patients had a longer length of stay compared to patients without a cancer diagnosis (3.2 +/- 8.3 vs. 3 +/- 5.5, $p<0.05$). There was a significant difference in the frequency of vertebral/spinal injuries in patients with versus without cancer diagnosis (32.3% vs 42.03% respectively, $p<0.05$). Finally, cancer patients were more likely to receive transfusion during admission versus non-cancer patients (2.27% vs 0.37%, $p<0.05$).

Conclusion: We conclude that pediatric trauma patients with a cancer diagnosis had a longer length of stay despite a similar injury severity score, were less likely to sustain vertebral/spinal cord injuries, but more likely to receive transfusions when compared to non-cancer trauma patients. There is need for further research regarding traumatic injuries in this patient population.

POSTER #54

Distress among parents and caregivers of patients with Anorectal Malformation and Hirschsprung Disease

Zoe Flyer DO, Andreina Giron MD, John Schomberg PhD MPH, Ashish Chogle MD MPH, Donald Shaul MD, Hira Ahmad MD

Purpose: Hospital anxiety and depression occurs commonly among famiSocely members who care for patients with congenital anorectal disorders. Better understanding of the prevalence is needed.

Methods: After IRB approval, a validated DT-P (Distress thermometer for parents) was given to 98 caregivers of the above patients at the Pull-through Network (PTN) conference, asking them to rank their degree of distress from 0 (no distress) to 10 (extreme distress) within the past week in a variety of areas including practical, emotional, family/social, physical, support, and communication categories. Scores greater than 4 strongly correlate to hospital anxiety, depression and parenting stress index. Diagnosis and demographic data were also collected. Participants were also asked to rank their most significant stressors with 1 being the most significant stressor and 5 was least significant.

Results: Among 98 caregivers that completed the survey, 90% of respondents were parents, 68% were female, and 61% were 35-54 years old. The diagnosis groups included isolated ARM (n=61), Cloaca (n=11), HD (n=9), and VACTERL (n=17). The DT-P scores over 4 occurred in 50% of ARM/cloaca, 35% of VACTERL and 77% of HD caregivers, p=0.03, among those completing the scoring. High proportions of surveyors reported high stress (score of 1-2): anal invasive treatment such as dilations, enemas, irrigations (36%) and unexpected diagnosis (46.9%).

Conclusions: Anxiety and/or depression was present in over half of caregivers of congenital anorectal disorders even though respondents were asked to simply quantify their stress over the past week. Unexpected ARM or HD diagnosis in the newborn period was a source of a significant stressor in almost half of the caregivers. These findings suggest that mental health interventions should be utilized early, perhaps as soon as the diagnosis of an ARM or HD is made.

POSTER #55

EMS Pediatric Trauma Transport: Do Disparities Exist?

Andreina Giron MD[a,d], John Schomberg PhD[a], Peter N. Dinh MA[a], Zoe E. Flyer DO[c], Laura F. Goodman MD[a,b], Yigit S. Guner MD[a,b], Peter T. Yu MD MPH[a,b].

[a]Division of Pediatric General & Thoracic Surgery, Children's Hospital Orange County, Orange, California, USA
[b]Department of Surgery, University of California Irvine Medical Center, Orange, California, USA
[c]Department of Surgery, Community Memorial Hospital in Ventura, California, USA
[d]Department of Surgery, Montefiore Medical Center, New York City, New York, USA

Introduction: Trauma is the leading cause of disability and death among children in the United States. Optimal outcomes are achieved at pediatric trauma centers (PTCs), which are specialized to address the unique needs of this population. Disparities in trauma have been reported, affecting optimal delivery of care. This study aims to investigate disparities in Emergency Medical Services (EMS) transport of pediatric and adolescent trauma patients and the factors influencing destination decisions.

Methods: This study utilized data from the National Emergency Medical Services Information System (NEMSIS) from 2017 to 2019, including EMS, sociodemographic, clinical, and decision-making variables. Patients aged 0-18 years that required EMS transport were included. Descriptive and regression analyses were conducted to identify factors influencing EMS transport destinations.

Results: 896,881 pediatric and adolescent patients requiring EMS transport were identified. After adjusting for trauma severity using NEMSIS Revised Trauma Score, 129,627 pediatric trauma patients were found. Most (89.4%) were transferred to a non-trauma center (NTC). Transport times to trauma centers (TCs) were slightly longer than NTCs (16.1 vs. 14.0 minutes, $p<.0001$). Transports to TCs were more likely to be associated with a clinical reason ($p<.001$); transports to NTCs were more likely to be associated with geography ($p<0.001$). Younger patients ($p<.0001$) and African Americans ($p<.0001$) had reduced odds of transport to TCs, even after adjusting for geography as the destination reason.

Conclusions: Racial disparities exist in EMS transport of pediatric and adolescent trauma patients. Addressing these disparities is critical to the equitable delivery of healthcare for children in the United States.

Support: This work was supported by the 2022 Pediatric Trauma Society Research Grant.

POSTER #56

Racial Disparity Among Youth Decedents of Legal Intervention Trauma

Annie Giron, MD[a,d] , Laura F Goodman, MD, MPH[a,b], John S Riley, MD, MS[c], Yigit S Guner, MD, MS, FACS[a,b], William H Peranteau, MD[c], John Schomberg, PhD, MPH[e], Peter T Yu MD, MPH.[a,b]

[a]Division of Pediatric General and Thoracic Surgery, Children's Hospital of Orange County, Orange, CA
[b]Department of Surgery, University of California Irvine Medical Center, Orange, CA
[c]Division of Pediatric General, Thoracic, and Fetal Surgery, Children's Hospital of Philadelphia, Philadelphia, PA
[d]Department of Surgery, Montefiore Medical Center, The Bronx, NY
[e]Trauma and Nursing Administration, Children's Hospital of Orange County, Orange, CA

Background: Legal intervention trauma (LIT) is defined as injury due to any encounter with law enforcement. This study investigates associations between demographics, violent status, and law enforcement tactics among youth decedents of LIT.

Study Design: Decedents of LIT age ≤ 26 years were identified using the CDC's National Violent Death Reporting System from 2003-2018. Decedents were classified as "violent" if they possessed a weapon, were committing a violent crime, or if law enforcement reported justified use of force. All others were classified as "nonviolent." Law enforcement tactics were stratified into "lethal" (firearm with standard ammunition) or "less-lethal" (any other) force. Differences in the racial distribution across these classifications were assessed using chi-square tests of proportions.

Results: We identified 1281 youth decedents of LIT. 92.5% met violent criteria. African Americans were less likely than Whites to possess a weapon (71.6% vs. 77.4%, p=0.02) and were not more likely to be committing a violent crime (63.6% vs. 60.4%, p=0.27). They were, however, more likely than Whites to experience force reported as justified by law enforcement (89.9% vs. 82.4%, p=0.002) and to experience exclusively lethal force not preceded by less-lethal tactics (94.0% vs. 88.7%, p=0.001). Among the subset of 85 cases where law enforcement reported justified use of force despite the decedent not possessing a weapon or committing a violent crime, the precipitating event was more often a traffic stop for African Americans than for Whites (28.5% vs. 6.66%, p=0.02).

Conclusions: These findings indicate a racial disparity among youth decedents of LIT.

POSTER #57

Thoracic Endovascular Aortic Repair in Pediatric Patients After Blunt Trauma: A National Analysis, 2018-2022

Andreina Giron, MD[a]; Christopher Matsko, MPH[c]; Bima J. Hasjim, MD, MSc[b]; Jeffry Nahmias, MD, MHPE[b]; John Schomberg PhD, MPH[a]; Laura Goodman, MD, MPH[a,b.]

[a]Children's Hospital of Orange County Division of Pediatric Surgery, Orange, CA, USA.
[b]University of California Irvine Department of Surgery, Orange, CA, USA.
[c]Nova Southeastern University Dr. Kiran C Patel College of Allopathic Medicine, Fort Lauderdale, FL, USA.

Introduction: Blunt thoracic aortic injury (BTAI) is relatively rare in children but lethal without appropriate and timely intervention. A prior national analysis from 2007-2015 found that Thoracic Endovascular Aortic Repair (TEVAR) made up 78% of all operative interventions compared to open thoracic aortic repair (OTAR). This study aims to provide an updated analysis of BTAI management in pediatric patients, hypothesizing that TEVAR was associated with better outcomes and fewer complications compared to OTAR.

Methods: This is a retrospective study of the National Trauma Data Bank 2018-2022. Patients <21 years-old with BTAI. Cohort was divided into TEVAR and OTAR. The primary endpoints were mortality, hospital and ICU length of stay, discharge disposition, and unplanned return to the OR. Descriptive statistics and bivariate comparisons of TEVAR vs OTAR were performed.

Results: Of 1,914 pediatric BTAI patients, 520 (27.2%) were managed operatively. Of these, 476 (91.5%) underwent TEVAR and 44 (8.5%) underwent OTAR. The OTAR group had a lower median age (17 years (14-19 years) vs. 19 years (17-20 years), P<0.001) but a higher rate of pancreatic injury (11.3% vs. 3.15%, p=0.02) compared to TEVAR patients. TEVAR patients more commonly sustained rib fractures (47.2% vs. 22.7%, P=0.002) compared to OTAR. There was no significant difference in mortality (p=0.63), hospital and intensive care unit length of stay (p=0.56 and 0.23, respectively), or discharge disposition (p=0.32). OTAR patients were more likely to return to the operating room (11.3% vs. 4.41%, P<0.001).

Conclusion: The proportion of TEVAR compared to OTAR has increased over the past decade. However, there were no differences in ISS, mortality, or discharge disposition. OTAR patients were younger and more often underwent unplanned return to the OR. Future research needs to be conducted on the use of TEVAR in younger pediatric patients.

POSTER #58

High Volume, Pediatric Trauma Centers are Associated with Improved Mortality in severe Pediatric TBI

Andreina Giron MD[a,d], John Schomberg PhD[a], Amy Lawrence[a,c], Alice Martino[c], Danh Nguyen[c], Brian Hanak MD[b], Joffre Olaya MD[b], Jeffry Nahmias MD MHPE[c], Laura F. Goodman MD[a,c], Peter T. Yu MD MPH[a,c], Yigit S. Guner MD[a,c].

[a]Division of Pediatric Surgery, Children's Hospital Orange County, Orange, California, USA
[b]Division of Neurosurgery, Children's Hospital Orange County, Orange, California, USA
[c]Department of Surgery, University of California Irvine Medical Center, Orange, California, USA
[d]Department of Surgery, Montefiore Medical Center, New York City, New York, USA

Background: Traumatic brain injury (TBI) is a leading cause of death among injured children. High case volume centers are linked to improved outcomes. This study evaluates the effect of hospital case volume on pediatric severe TBI (PSTBI) mortality rates, hypothesizing decreased mortality at high-volume centers.

Methods: We analyzed patients under 19 with PSTBI (Glasgow coma score ≤8) from the National Trauma Data Bank, 2017-2019. Hospitals were considered high volume if annual median case volume was greater than 18.3 (n=4432) and low-volume (n=8178) if otherwise for Bivariate analysis. Multivariate mixed effect logistic regression was conducted where facilities were divided by volume tertiles.

Results: Of 12,268 PSTBI patients, 24.7% died. Cohort populations were comparable in demographic and clinical parameters. High volume hospitals had decreased mortality rates (23.2% vs 25.7%, p=0.002), resulting in a modest but significant mortality risk reduction (OR=0.97,0.95-0.99, p=0.006) when compared to low volume hospitals. Also, patients <10 years-old (OR = 0.92, 0.90-0.96, p<0.001) and abuse victims (OR=0.85, 0.82-0.88, p=0.03) had decreased associated risk of mortality. Lack of VTE prophylaxis (OR=1.31, 1.29-1.34, p <0.001), blood product use (OR=1.28, 1.25-1.30, p=0.002), injury severity (OR=1.20, 1.18-1.22, p <0.0001), treatment at adult-only facilities (OR=1.02, 1.00-1.03, p=0.01), and high-volume centers without pediatric designation (OR: 1.06, 1.04-1.09, p<0.001) were associated with increased mortality.

Conclusion: In-hospital mortality was lower for PSTBI patients treated in high-volume centers with pediatric center status. There may be benefits to concentrating TBI patient care in these centers.

POSTER #59

Association of Traumatic Abdominal wall Hernias and Hollow Viscus and solid Organ Injury in Pediatric Patients

Andreina Giron, MD[a]; Zoe Flyer, DO[a]; John Schomberg PhD, MPH[a]; Jeffry Nahmias, MD, MHPE[b]; Laura Goodman, MD, MPH[a,b.]; Hira Ahmad, MD[a,b.]

[a]Children's Hospital of Orange County Division of Pediatric Surgery, Orange, CA, USA.
[b]University of California Irvine Department of Surgery, Orange, CA, USA.

Purpose: Traumatic abdominal wall hernias (TAWHs), though rare in pediatric trauma patients, may indicate a more severe underlying injury particularly to hollow viscus and solid organs. This study aimed to evaluate the incidence of hollow viscus injury (HVI) in the setting of TAWH versus an abdominal wall hematoma hypothesizing increased HVI with TAWH.

Methods: A retrospective study of the National Trauma Data Bank (NTDB) years 2018-2022 was conducted. Patients aged ≤18 years diagnosed with TAWH or abdominal wall hematoma/ecchymosis without TAWH were included and compared with bivariate statistics and a multivariable logistic regression evaluating risk of HVI, controlling for age, sex, race, ethnicity, insurance type, systolic blood pressure (SBP) on presentation and the Injury Severity Score (ISS).

Results: From 13,261 included patients 414 (3.1%) had TAWH and 12,847 had abdominal wall hematomas. The median age was similar between cohorts (TAWH: 11 years vs abdominal wall hematoma: 12 years, p=0.08). However, more males comprised the TAWH cohort (63.5 vs 55.6%, p=0.01). The ISS was lower in the TAWH group (median 5 vs. 9, p<0.0001), however TAWH patients had an increased rate of HVI (10.3% vs. 2.5%, p<0.0001). Logistic regression model showed that the presence of TAWH was associated with an increased odds of HVI (OR 1.08, p<0.0001).

Conclusions: TAWHs are associated with a higher risk of HVI compared to abdominal wall hematomas or ecchymosis alone in pediatric trauma patients. Thus, heightened clinical suspicion and thorough evaluation is necessary in this subgroup.

POSTER #60

PTSD among patients with Anorectal Malformation and Hirschsprung Disease

Andreina Giron, MD[a]; Zoe Flyer, DO[a]; Ashley Bone, MSN, CPNP[a]; John Schomberg, PhD, MPH[a]; Andreina Urrutia Gonzales, BS[a]; Ashish Chogle MD, MPH[c]; Elicia Wartman, PhD[c]; Peter T. Yu, MD, MPH [a,b]; Donald Shaul, MD[a,b], Hira Ahmad, MD[a,b].

[a]Children's Hospital of Orange County Division of Pediatric Surgery, Orange, CA, USA.
[b]University of California Irvine Department of Surgery, Orange, CA, USA.
[c]Children's Hospital of Orange County Department of Gastroenterology, Orange, CA, USA.

Purpose: Management of patients with anorectal malformations (ARM) and Hirschsprung Disease (HD) requires multiple interventions and lifelong follow up. We aimed to determine the prevalence of Post-Traumatic Stress Disorder (PTSD) among patients with ARM and HD.

Methods: After IRB approval, a validated self-rating *DSM-IV* PTSD (PCL-5) checklist was administered to 30 patients with ARM and HD to assess the presence and severity of PTSD symptoms. The PCL-5 is a 20-item questionnaire, corresponding to the DSM-5 symptom criteria for PTSD. The survey (likert scale) is a provisional diagnostic tool to connect patients to appropriate mental health care resources. PCL-5 cutoff score between 31-33 is indicative of probable PTSD.

Results: All 30 patients completed the survey; the participants had even gender distribution, a median age of 12-64 years and majority were White (76.6%). Education levels varied, with 30% holding a Bachelor's degree and 20% having less than a high school education; employment status varied across as well, with only 3 (10%) reported being unable to work. 27 patients had a diagnosis of ARM (90%). Among the 30 respondents, 12 (44.4%) scored >31 on the PCL-5 score; 17 (62.9%) of patients reported "moderate" or greater in all PCL-5 domains. When asked about highest stressors, 13 (43.2%) ranked anal dilation/enema treatment as their highest stressor, while 12 (39.9%) reported effect of diagnosis on body image and social life as the highest stressor.

Conclusions: The high prevalence of PTSD among patients with ARM and HD underscores the need for mental health support. About half of the patients met the provisional PTSD diagnosis based on PCL-5, with a significant proportion in the moderate to severe category. These findings highlight the critical role of early psychological involvement and availability of mental health care resources for these patients.

POSTER #61

Development of a Patient and Peer Advocacy Program in Pediatric Colorectal Surgery

Zoe Flyer, DO[a]; Andreina Giron, MD[a]; John Schomberg, PhD, MPH[a]; Eliad Amini, BS[c]; Hira Ahmad, MD[a,b]; Donald Shaul, MD[a,b.]

[a]Children's Hospital of Orange County Division of Pediatric Surgery, Orange, CA, USA.
[b]University of California Irvine Department of Surgery, Orange, CA, USA.
[c]University of California Irvine, Irvine, CA, USA.

Purpose: Patients, and their families, who were born with Anorectal Malformations (ARM) and Hirschsprung disease (HD) often worry about what the future holds and struggle with decisions regarding major elective surgical interventions. Providing a peer resource may provide perspective on the future and facilitate decision making.

Methods: A patient and peer advocacy program was established wherein a young adult patient with a history of ARM and a Malone conduit (MACE) volunteered in our colorectal center. That individual met with each family and patient who had questions about their disease and/or about potential future surgeries. Surveys were administered prior to, and after meeting with the advocate, or retroactively, if that meeting had occurred in the past. A series of 24 questions were asked with a 5-point Likert scale response option.

Results: 4 surveys were completed prior to, and 11 surveys were completed after meeting the advocate, and these responses were compared. There was almost no change in median and modal responses as to how families answered regarding how well they understood the patient's condition, how well they managed it, and how much they worried about their child's overall health or social future, among other questions. There was a decrease in how much they worried about their child's academic future and a considerable improvement in their understanding of the Malone concept. 11 of 11 families felt that meeting with the advocate would benefit other families, and 9 of 11 felt that meeting with the advocate had a positive impact on their family.

Conclusions: Families who met with a patient and peer advocate (who has a MACE) improved their understanding of the MACE concept considerably, and worried less about their child's academic future. Nearly all felt that the meeting was valuable and should be offered to other families.

POSTER #62

Ketorolac: Safety and Efficacy in the Management of Pain for Post-Operative Pediatric General Surgery Patients

Zoe Flyer DO[1], Andreina Giron MD[1,2], Rebecca John MSN CPNP[1], John Schomberg PhD[1], April Carlson MD[2], Laura Goodman MD MPH[1,2], Yigit Guner MD[1,2], Peter Yu MD MPH[1,2]

[a]Children's Hospital of Orange County, Division of Pediatric Surgery, Orange California USA
[b]University of California, Irvine, Division of Pediatric Surgery, Irvine California USA

Purpose: Ketorolac is a nonsteroidal anti-inflammatory drug used for pain management with an adverse effect of bleeding. The safety and efficacy of this medication has not been examined in children who have undergone general surgery procedures.

Methods: This is a single institution retrospective cohort study examining pediatric patients with and without Ketorolac treatment <24 hours after surgery. Healthy children ages 0-18 from 2017-2022 who underwent one of the following operations were included: laparoscopic appendectomy (LA); laparoscopic cholecystectomy (LC); laparoscopic inguinal hernia repair (LIHR); open inguinal hernia repair (OIHR); umbilical hernia repair (UHR); and ventral hernia repair (VHR). The distribution of patient demographic and clinical characteristics were reported using bivariate inferential statistics. A Cox proportional hazards model was used to identify associations between Ketorolac and time to pain score ≤3 after adjusting for surgery type and patient demographics. A logistic regression model was used to measure the odds of opioid use in morphine equivalents 24-hours after surgery.

Results: 5761 patients were identified. Children who received Ketorolac after surgery versus those who did not were more likely to be older (11.3 vs. 6.6 years, p<.0001). Whites were more likely to receive Ketorolac after surgery, whereas Asian Americans, African Americans, and those with Hispanic ethnicity were not. 4303 (75.8%) of patients underwent LA; 478 (8.4%) LC; 147 (2.5%) LIHR; 559 (9.8%) OIHR; 150 (2.6%) UHR; and 34 (.005%) VHR. Blood transfusions were rare but were less common in the cohort that received Ketorolac (0.4% vs. 0.98%, p=.004). Patients receiving Ketorolac were less likely to have a higher pain score (>5) after surgery (59.8% vs. 85.7%, p<.0001). Patients receiving Ketorolac had a 10% decreased odds of receiving morphine equivalents after surgery (OR=.90, 95% CI: .87-.94, p<.0001) when compared to patients that did not.

Conclusion: Ketorolac is safe to use in healthy pediatric patients who have undergone pediatric general surgery procedures. It does not appear to be associated with increased bleeding risk in the post-operative period, even less than 24 hours after surgery. Ketorolac is efficacious in this patient population either alone or in combination with opioids and can decrease the amount of narcotic received.

POSTER #63

Impact of Hospital Volume on Mortality in Pediatric Patients with Severe Blunt Abdominal Trauma

Zoe Flyer DO, Andreina Giron MD, Zhuo Chen PhD, Jeffry Nahmias MD MHPE, Laura F. Goodman MD MPH, Yigit S. Guner MD, Peter T. Yu MD MPH

Background: Blunt abdominal trauma is a significant cause of mortality in pediatric patients. This study evaluates the impact of hospital volume on mortality rates in trauma patients <18 years with an injury severity score (ISS)> 25 and blunt abdominal trauma, hypothesizing that lower volume centers are associated with increased mortality.

Methods: We analyzed severely injured pediatric patients suffering blunt abdominal trauma (ISS ≥ 25) within the National Trauma Data Bank (NTDB) 2017-2019. Using the median of the total volume distribution of abdominal trauma (ISS ≥ 25) cases per year, hospitals were categorized into higher-volume centers (HVCs) (≥ 8 cases/year) and lower-volume centers (LVCs) (< 8 cases/year) for Bivariate analysis. Multivariate mixed effect logistic regression was also conducted where facilities were divided by volume tertiles.

Results: Of 6499 included patients, 3252 (50.1%) were treated at HVCs and 3247 (49.1%) at LVCs. HVC patients were younger (mean: 12.36 vs. 13.91 years, p<0.0001) and had less males (63.8% vs. 67.4%, p=0.002). HVCs had increased rates of intubations (31.0% vs. 24.1%, p<0.0001) and ICU admissions (53.3% vs. 49.8%, p<0.0001). Additionally, patients at HVCs were more often treated in verified American College of Surgeons Level I trauma facilities (55.0% vs. 34.8%, p<0.0001) and verified pediatric Level I facilities (47.7% vs. 9.6%, p<0.0001). The in-hospital mortality rate at LVCs was higher (15.8% vs. 17.5%, p<0.001) compared to HVCs. Multivariable analysis showed that treatment at low-volume centers was associated with a significant increase in mortality risk (OR=1.29, 95% CI 1.06-1.68, p=0.01).

Conclusion: Pediatric patients with severe blunt abdominal trauma treated at LVCs had a significantly higher in-hospital mortality rate. Future research should focus on identifying and standardizing best practices to improve outcomes, morbidity and mortality across trauma centers of varying patient volumes.

POSTER #64

Variability in EMS Management of Venomous Snake Bites

April Carlson, MD[b], Zoe Flyer, DO[a], Andreina Giron, MD[a], Peter T. Yu MD, MPH[a,b], Laura F. Goodman, MD, MPH[a,b], John Schomberg PhD, MPH[a].

[a]Division of Pediatric Surgery, Children's Hospital of Pediatric Surgery, Orange, CA
[b]Department of Surgery, University of California Irvine Medical Center, Orange, CA

Background: A combination of opioids, epinephrine, diphenhydramine, and Non-Steroidal Anti-Inflammatory Drugs (NSAIDs) are frequently used by Emergency Medical Services (EMS) when treating venomous snake bites. However, the efficacy of diphenhydramine and NSAIDs in this context remains controversial. This study aims to assess the impact of diphenhydramine on the clinical acuity of venomous snake bite victims, hypothesizing that it does not contribute to clinical improvement.

Methods: this retrospective cohort study was conducted utilizing data from the National Emergency Medical Services Information System (NEMSIS) spanning 2017 to 2020. Cohort was stratified by treatment received after venomous snake bites. Descriptive statistics were conducted.

Results: 1,269 pediatric patients were included. Median age was 9 (5-14, $p<0.001$), with no differences in gender ($p=0.62$) or bite by snake species distribution ($p=0.99$). 57 (4.49%) patients were treated with diphenhydramine/NSAIDs, 102 (8.93%) with opioids alone, and 1100 (87.47%) with neither diphenhydramine/NSAIDs nor opioids. On initial assessment 17.99% of diphenhydramine/NSAIDs group, 17.7% of opioids alone group, and 6.94% of neither diphenhydramine/NSAIDs nor opioid group were considered critical ($p < .0001$). On critical status reassessment the diphenhydramine/NSAIDs treatment group had 31.2% improvement in acuity, opioids alone treatment group had 9.23% improvement in acuity, and neither diphenhydramine/NSAIDs or opioids treatment group had 9.33% improvement in acuity ($p=.002$). Administration of epinephrine was higher in the diphenhydramine/NSAIDs treatment group (26%) vs. opioids alone treatment group (0%), and neither diphenhydramine/NSAIDs nor opioids treatment group (0.26%, $p < .0001$).

Conclusion: The study highlights variability in the treatment of venomous snake bites by EMS across the United States. Despite the lack of robust scientific evidence, we found that diphenhydramine and NSAIDs are frequently administered alongside epinephrine. It is difficult to assess if the improvement is due to diphenhydramine/NSAIDs use or epinephrine use alone. Further research is needed to align clinical practices with established best practices.

POSTER #65

Enhancing Leadership Well-Being: Outcomes of a 4-Day Workweek Pilot

Melanie Patterson, Jennifer Hayakawa

This study evaluates the implementation of a 4-day workweek among healthcare leaders as a strategy to enhance work-life balance, alleviate burnout, and promote workforce stabilization. High leadership turnover rates in healthcare significantly impact associate retention and care quality. Work-life balance, emotional health, and job satisfaction are critical issues for the retention of nurse leaders (AONL, 2023). Implementing effective strategies such as flexible scheduling, increased recognition, and additional time off is essential for improving staffing and retention.

In June 2022, a 4-day workweek was implemented for inpatient nurse and respiratory leaders across our two hospitals. A cohort of 38 leaders transitioned to this compressed schedule while maintaining their responsibilities. The objective was to achieve a 20% reduction in self-reported burnout and a 20% increase in job satisfaction by June 30, 2023. Quantitative data was collected biweekly using the Mini-Z survey to measure changes in burnout, joy in work, and job satisfaction.

Self-reported burnout decreased from 61% to 14%, joy in work increased from 34% to 86%, and job satisfaction rose from 71% to 96%. Job-related stress decreased from 76% to 14%. Participants reported better management of work and personal responsibilities, enhancing their mental and physical well-being. Qualitative analysis provided insights into the barriers and highlighted the positive effects of the new schedule on participants' well-being, work-life balance, and overall morale.

Implementing a 4-day workweek in patient care leadership roles requires clear guidelines on leader availability and maintaining continuous accountability, including 24/7 coverage for urgent matters. Efficient workload distribution across teams and well-defined evaluation criteria are essential for monitoring performance, providing feedback, and addressing concerns while ensuring seamless operations. This study adds to the growing evidence on alternative work schedules in healthcare settings and offers practical insights for administrators and leaders seeking strategies to enhance staff well-being and job satisfaction.

POSTER #66

Age and Resource Utilization in Pediatric Falls: a study of the National Trauma Data Bank

Alice M. Martino, MD[b], Laura F. Goodman, MD, MPH[a,b], Andreina Giron, MD[a], Ellie Gibbs, BA[c], John Schomberg PhD, MPH[d], Elizabeth Wallace MPH[d], Peter T. Yu MD, MPH[a,b], Troy Reyna, MD[a,b], Maryam Gholizadeh, MD[a,b], Mustafa Kabeer, MD[a,b], Saeed Awan, MD[a,b], Yigit S. Guner, MD, MS[a,b]

[a]*Division of Pediatric Surgery, Children's Hospital of Pediatric Surgery, Orange, CA*
[b]*Department of Surgery, University of California Irvine Medical Center, Orange, CA*
[c]*Wellesley College, Department of Biological Sciences, Wellesley, MA*
[d]*Children's Hospital of Orange County, Orange, CA*

Background: The aim of this study is to elucidate trends in resource utilization among pediatric fall-from-building patients on a U.S. national scale based on patient demographics.

Methods: We used the National Trauma Data Bank (NTDB) from 2007-2015 and identified pediatric (1-5, 6-10 years) falls patients using ICD-9 E882 and ICD-10 W13.9 codes. Resource utilization was represented by admission to the operating room (OR) directly from the emergency department (ED), ventilator use, radiology exams, invasive procedures, and admission to the intensive care unit (ICU). Descriptive analyses and a Cox proportional hazards model were used, along with logistic regression and Poisson regression.

Results: 8,362 patients were studied and divided into two age groups. Younger children (ages 1-5 years) compared to older children (6-10 years) were more likely to use hospital resources due to a traumatic fall. Children in the 6-10 age group had higher OR utilization than those in the 1-5 age group (12.0% vs 5.8%, $p<0.01$). Children between 1-5 years had higher ventilator utilization compared to children 6-10 years of age (10.5% vs 6.8%, $p<0.01$). In addition, children in the 1-5 years group were more likely to require ICU admission (39.4% vs. 27.0%, $p<0.01$).

Conclusion: Pediatric falls are the leading cause of injury for children under 10 years old and treatment can require considerable resources. Younger children are likely to have more severe falls and therefore have increased resource utilization. Prevention efforts should focus on standard window guards and locks, as well as education for families to reduce falls particularly amongst children ages 1-5 years.

POSTER #67

Correction of the common splice site variant associated with mitochondrial complex I deficiency caused by recessive mutations in the *NUBPL* gene

Virginia Kimonis

Children's Hospital of Orange County, Orange CA
Division of Genetics and Genomic Medicine, Department of Pediatrics, UC Irvine, Orange CA

Mitochondrial Complex I deficiency caused by recessive mutations in the NUBPL gene is an early onset mitochondrial disorder, most commonly presenting with cerebellar dysfunction (e.g., ataxia, dysarthria, nystagmus, and tremor). Patient brain MRIs show progressive cerebellar atrophy. Presently, no specific curative treatment exists for patients with this rare progressive disease. We have identified respiratory chain respiratory bioenergetics, and RNA splicing variants in fibroblasts from patients, versus controls.

Utilization of patient versus control fibroblasts for translational studies is in progress with novel antisense oligonucleotides (ASOs). We will study the effects of ASOs in correcting splicing and improving mitochondrial function in three individuals versus controls fibroblasts and iPSCs derived neurons.

POSTER #68

Relationships between age, REM sleep, and sleep disordered breathing across the first 24 months of life

Neal Nakra, Tricia Morphew, Spencer Polk, Lia Galut, Lois Sayrs, Kevin Blaine, and Katharine C. Simon

Introduction: Infant airway anatomy and immature central respiratory systems place infants at risk for sleep disordered breathing. Further, infant sleep is comprised of higher amounts of rapid eye movement (REM) sleep, with a steady decline across the first twenty-four months. Compared to the controlled breathing rate during non-REM (NREM), REM sleep has an erratic rate. The combination of immature anatomy and increased REM amount predispose infants to greater apnea risk, yet there is a poor characterization of age- and REM-associated changes in disordered breathing across the first twenty-four months.

Methods: We retrospectively analyzed 95 clinical sleep studies from 50 infants (M=10.5 months, SD = 7.6., F=24), each had one to three records, who were being evaluated for sleep disordered breathing. We investigated the average number of combined central and obstructive apnea events per hour, controlling for age of infant, and percentage of total sleep time in both NREM and REM, respectively, using a Poisson regression within the Generalized Linear Mixed Model (GLMM) framework.

Results: Infants slept on average 371 minutes (SD=121), of which approximately 33% (SD=12%) was spent in REM. We observed an average of 5.3 apnea events per hour (SD = 5), with a median of 3.5 [IQR: 1.7, 7.9], unadjusted, which we found varied by age ($p<.05$). During the first year, higher apnea event rates were observed at one and six months, while the lowest apnea rate occurred at three months of age. Between 12 and 24 months, rates stabilized, mirroring those observed at three months, but with a noticeable increase at 18 months. There was a strong relationship between age and REM sleep, with 3.2 times more apnea events per hour during REM versus nREM. This increased rate during REM was consistent across age, including when adjusted for the percentage of REM sleep and sex (RR=3.18, 95% CI 2.61, 3.89, $p<.001$).

Conclusions: We observed a dynamic relationship between age, REM sleep, and total apnea events across the first twenty-four months of life. As infants are at higher risk for apnea, evaluating age- and REM-associated apnea changes may provide new assessment and intervention targets in sleep disordered breathing.

POSTER #69

What's the plan? A single center's experience navigating through the challenges of newborn screening identified attenuated MPS-I cases

Kathryn Gasperian[1], Rebecca Sponberg[1], Laura Pollard[3], Patricia Hall[2], Marzia Pasquali[4], Raymond Wang[1]

[1]Children's Hospital of Orange County, Orange, CA, USA, [2]Department of Laboratory Medicine and Pathology, Mayo Clinic, Rochester, MN, USA, [3]Greenwood Genetic Center, Greenwood, SC, USA, [4]University of Utah School of Medicine/ ARUP Laboratories, Salt Lake City, UT, USA

Mucopolysaccharidosis Type I (MPS-I) is characterized by the deficiency of alpha-L-iduronidase (IDUA) due to pathogenic variants in the *IDUA* gene leading to accumulation of glycosaminoglycans (GAGs) in various tissues and organs. There is a wide phenotypic spectrum associated with MPS-I ranging from a severe form (Hurler syndrome), which is treated by hematopoietic stem cell transplant, to an attenuated form (Hurler-Scheie/ Scheie syndrome), treated by enzyme replacement therapy (ERT), with many shared features between them. California began screening MPS-I in 2018 by utilizing a two-tiered system, IDUA enzymatic analysis and DNA Sanger sequencing of *IDUA* gene, which has been instrumental in early identification and treatment of newborns with severe form. This approach has also detected infants with unique molecular and biochemical profiles more likely consistent with an attenuated phenotype. Due to the absence of longitudinal data on the clinical outcomes and treatment course, and uncertain molecular and biochemical profiles, there is no consensus regarding management of these patients who are likely to have an attenuated form of MPS-I. This makes it challenging to standardize care across centers. A retrospective case series was completed on five patients identified from California's newborn screening at a single pediatric hospital, who likely have attenuated MPS-I. Data was collected on each patient's genotype, biochemical findings, clinical evaluations, treatment, and outcomes. Patients' genotypes include p.Trp402Ter/p.Ala513_Ala514insGlyArgValAlaAla, p.Gln70Ter/p.Ser423Arg, p.Arg89Trp/p.Arg628Pro, p.Phe248del/p.Lys153Ter, and p.Leu526Pro/p.Leu18Pro. We have observed decreased IDUA enzyme activity levels (5/5 patients), abnormal (1/3 patients) or essentially normal blood GAGs (2/3 patients), abnormal (1/3 patients) or essentially normal urine GAGs (2/3 patients), and abnormal urine NREs (Sensi-Pro®) [2/2 patients]. Only three patients are receiving ERT. This study confirms that longitudinal studies and collaboration with other centers across states completing newborn screening for MPS-I need to further advise management of patients with an attenuated phenotype.

POSTER #70

Patient Demographics, Medication Adherence and Self-Management as Reported by Caregivers of Children with Epilepsy: An analysis of the baseline CHOC data from the multi-site Epilepsy Adherence in Children and Technology (eACT) study

Jessica Ardo, MSW[1], Erica Navarro, BA[1], Phuoc (Jenny) Vo, BS[1], Stacy Buschhaus, BA[2], Shanna Guilfoyle, PhD[2], Janelle Wagner, PhD[3], Anup Patel, MD[4], Heather Huszti, PhD[1], and Avani Modi, PhD[2]

[1]Children's Hospital of Orange County, [2]Cincinnati Children's Hospital Medical Center, [3]Medical University of South Carolina, [4]Nationwide Children's Hospital

Background: Effects of site for the Epilepsy Adherence in Children and Technology (eACT) multi-site study on demographics and baseline Pediatric Epilepsy Medication Self-Management Questionnaire (PEMSQ) are explored for the CHOC study participants. Characterizing the site-level demographic and PEMSQ data is helpful to identify the specific strengths and needs of one hospital's patient population and allows for better allocation of resources.

Purpose/Aims: A mHealth intervention was used to address patient medication non-adherence in children with epilepsy. This poster will describe the baseline characteristics of the participants enrolled at CHOC. The broader purpose of the Epilepsy Adherence in Children and Technology (eACT) study is to test the effectiveness of a mHealth behavioral intervention on pediatric patient medication adherence.

Methods: The eACT study is a four-site clinical trial which enrolled 466 participants across all sites. The trial examined the efficacy of a mHealth intervention delivered at stepped levels to pediatric patients with epilepsy on medication adherence. Participants were asked to choose one of two electronic medication devices to monitor adherence (bottle or pill tray) and to complete survey measures. After a two-month baseline period, those participants with ≤95% adherence were randomized to control or treatment (education modules and reminders; or education modules, reminders and feedback). After three months, participants with adherence at ≤95% were re-randomized to one of two groups: continue with same treatment or offered two sessions of individual telehealth with a specialist. While analysis is ongoing, study-wide demographic and baseline characteristics have been explored and are presented. In this study, the baseline demographic data and the correlation between baseline adherence and PEMSQ sub-scales among CHOC participants is reported.

Results: Demographics study-wide demonstrate among 466 participants enrolled, 400 completed the Baseline timepoint, 268 of whom had adherence ≤95% and were randomized. At CHOC, the study team enrolled 134 participants and 107 completed the Baseline timepoint, 77 of whom had ≤95% adherence and were randomized. Baseline adherence, child and caregiver demographics, and baseline PEMSQ scale scores were analyzed for correlation.

Conclusions:
The demographics of the CHOC participants (child and caregiver), medication adherence at end of the two-month Baseline period and scale scores for the four PEMSQ domains, as well as implications for CHOC patients are discussed. **Funding:** This work was funded by NIH grant 5R01NR017794.

POSTER #71

The Family-Based Crisis Intervention at CHOC Mental Health Inpatient Center: Participant demographics and methods

Jessica Ardo, MSW[1], Erica Navarro, BA[1], Sharonne Herbert, PhD[1], Francesca Bahn, PhD[1], Gabrielle Hernandez, LCSW[1], and Abigail Ross, LICSW, MPH, PhD[2]

[1]Children's Hospital of Orange County, [2]University of Pennsylvania Perelman School of Medicine, School of Social Policy and Practice

Background: Demographics of the participants in the Family-Based Crisis Intervention - Randomized Controlled Trial at CHOC's Mental Health Inpatient Center are presented. Youth participants included those admitted to CHOC's MHIC with suicidal ideation and/or suicide attempt, who consented and agreed to participate and were eligible. The methods used for the study are described and discussed.

Purpose/Aims: A randomized controlled trial of the Family-Based Crisis Intervention-Inpatient as developed by Dr. Abigail Ross was conducted at CHOC's MHIC. This poster will describe the baseline characteristics of the participants (youth and their caregivers), the methods used in the study in the context of the MHIC and barriers and lessons learned.

Methods: FBCI is a single-session intervention designed to be used with suicidal adolescents and their family in the emergency department setting. CHOC implemented FBCI in the ED in 2019. As part of two of the author's ongoing work, the intervention was adapted for use with inpatient adolescents (FBCI-IP) and a randomized controlled trial comparing it to treatment as usual (TAU) was done in CHOC MHIC. Three CHOC MHIC clinicians were trained to use the FBCI-IP intervention, and the remaining therapists were not trained. Participants were enrolled and randomized at baseline to either FBCI-IP or TAU. Those randomized to FBCI-IP were assigned to a therapist trained in the intervention and those randomized to TAU were assigned to an untrained therapist. All participants were asked to complete surveys at baseline, discharge, and at one week, one month and six months post-discharge.

Results: CHOC enrolled 26 participants and their caregivers in the FBCI-RCT study. Of those 26, 22 completed the surveys at baseline and discharge, and 19 completed at least one follow-up. 12 were randomized to FBCI-IP and 13 were randomized to TAU (one participant withdrew prior to randomization). Baseline demographics are analyzed and methods are described.

Conclusions: Implementing a randomized controlled trial in a very busy 18-bed inpatient mental health setting, study methods, and the demographics of the 26 participants (youth and caregiver), as well as lessons learned are presented.

Funding: This work was supported by a grant from the American Foundation for Suicide Prevention (#YIG-1-140-19).

POSTER #72

Effectiveness of *FIT Brain*, a virtual-based cognitive-behavioral therapy program in addressing long-term psychological distress among childhood cancer survivors

Erica Navarro[1], Jenny Vo[1], Grace Mucci, Ph.D.,[1,2] Jessica Ardo,[1] Crystal Yu,[1] & Tricia Morphew,[1,3]

[1]CHOC Children's Hospital, United States of America; [2]University of California, Irvine, United States of America; [3]Morphew Consulting, LLC, United States of America

Background: As the number of childhood cancer survivors is growing as researchers develop increasingly aggressive treatments, more and more children experience iatrogenic effects, including detriments to neuropsychological functioning (Moore, 2005) and psychological distress (Cox et al., 2018). In response to this growing need, we developed a virtual group program using cognitive-behavioral strategies to help improve coping skills and self-esteem of survivors.

Methods: In this pilot study, we enrolled a total of 35 patients from 9/19/2019 to 11/15/2023. At the time of analysis, a total of 18 patients had complete assessment data at baseline and one-month follow-up. To assess changes over time we implemented repeated measures analysis using mixed effects modeling that included a random intercept term for patients utilizing the Behavior Assessment System for Children (BASC-3; Reynolds & Kamphaus, 2015).

Results: Among the 18 patients with one month follow-up data, a substantial portion exhibited psychological challenges: 44.4% had psychological maladjustment present, 38.9% experienced anxiety and developmental disabilities with 27.8% having cognitive disability. Significant improvements after one month of intervention were noted for personal adjustment from 42.88 (SE=3.38) to 46.91 (SE=3.25) (baseline-one month Δ=-4.03, 95% CI -8.05, -0.01, p=.050), interpersonal relations from 41.38 (SE=3.11) to 45.74 (SE=2.96) (Δ=-4.36, 95% CI -8.53, -0.19, p=.041), and self-reliance from 40.84 (SE=3.14) to 46.60 (SE=2.93) (Δ=-5.76, 95% CI -10.69, -0.82, p=.024). However, after adjusting for multiple comparisons (significance threshold p<.001), these improvements should be interpreted with caution. No enhancements were observed in the remaining BASC-3 scales.

Discussion: Utilization of innovative virtual-based programs, such as *FIT Brain*, with pediatric cancer survivors may be instrumental in improving overall psychological functioning. The applicability of *FIT Brain* and other similar programs may be beneficial within clinical settings. Future research is needed to further explore the effectiveness of virtual-based programs targeting both the psychological and cognitive difficulties survivors often experience.

POSTER #73

Evaluation of the School Reintegration Program for Youth Recovering from an Acute Crisis: A mixed-methods approach

Vanesa Perez, PhD; Gabriela Hernandez, LCSW; Francesca Bahn, PhD

Background: Youth returning to school following psychiatric hospitalization are at increased risk for mental health crises and rehospitalization. Aligned with the goals of AB 2246 & 1746, facilitating school reintegration during youth's hospitalization may reduce this risk and support positive mental health outcomes. The School Reintegration Program (SRP) was piloted at CHOC's MHIC and focused on partnering with schools during hospitalization to better support reintegration.

Aims: This study aimed to 1) assess and evaluate local schools' experience with SRP; 2) identify and explore areas for improvement.

Methods: A mixed-methods approach was used to address study aims. To address Aim 1, a survey was sent out to school district leads to evaluate factors related to program dissemination and implementation (n= 26 accessed the survey; n= 15 completed the survey; n= 11 chose not to participate). District leads (n= 3) participated in a semi-structured interview to address Aim 2.

Results: Results from the survey indicate hospital communication is widely used by the schools (93%), followed by the safety plan (73%), treatment team meetings (53%), support in the school re-entry meetings (53%), and the school reintegration form (53%). Acceptability of the program ranged from 4.1-4.5, appropriateness ranged from 4.2-4.3, feasibility ranged from 4.1-4.5, and satisfaction ranged from 3.7-4.3. Analysis of the semi-structured interviewed revealed various program strengths, as well as areas for improvement.

Conclusions: SRP is a highly valued program from the perspective of school-based stakeholders. School leads identified several strengths, expressed desire for more services, and for greater presence of the program in schools. Areas for improvement aligned with current program adaptations that are in development. Future directions will assess caregiver perspectives and intervention-based outcomes.

Contact Us

CHOC Research Institute | 1201 W La Veta Ave.,
Orange, CA 92868 | 714.509.4341 | research@choc.org

CHOC Research
GO BEYOND®

CHOC Research Changes Lives.

One Child at a Time.