http://www.upstate.edu/gch/academics/newsletter.php

The APPD Report

We were very well represented at the APPD conference in March. Holly Stacey, Alex Ramirez, and Asmita Sharma went as our present and future chiefs. Holly tells the Crier, "This year APPD was in Atlanta, Georgia and we had an awesome time. The sessions were inspiring and motivating. I had the opportunity to meet with graduating chiefs across the nation and we were able to debrief our year; the ups and the downs. We then met with rising chiefs from across the country and had the chance to pass on our wisdom and guidance! We attended a phenomenal session focusing on simulation in pediatric training and it was awesome to see what other programs are doing! It gave us a ton of new ideas for coming years. Asmita, Lauren T and I also went to the World of Coca-Cola which was so much fun - we were able to try different coke products from across the world (some were certainly better than others). We walked a lot, ate a ton and learned so much! It was an awesome week and I can't wait to go back next year!

Lauren Takes AIMS

Lauren Thomas went as a member of the AIMS cohort. She shared a few of her impressions of the experience with the Crier: "Being a part of this year's APPD's AIMS cohort was invaluable! To be surrounded by other URIM residents that have the same common goal of advancing health equity in underserved communities was encouraging. Everyone that was apart of the AIMS organization was supportive and genuinely wanted us to win. It's hard to put to words what the conference meant to me. I only wish it was longer!"



And Dr. Nelsen was there both as program director and presenter.

Peds Research Day Winners

Your vote counted! The annual pediatric research platform/poster session was held on Wednesday, April 12th. Several members of the department gave oral presentations (Sarah Sloan, Jenny Black, Teaghen Buscemi-Kimmins, Heather Ross, Danielle Daniels, and Elizabeth Nelsen), and an online poster session with 23 posters was available for two weeks. Viewers were encouraged to submit their vote for the People's Choice award. Congratulations to everyone who participated in either the platforms or poster session for the annual Pediatric Department Research Day. But special congratulations go to the three winning posters:

Best QI Poster

Michelle Jeski, MSN, RN, PCNS-BC: "Nurse Driven Diabetic Ketoacidosis Fluid Titration Protocol Outside the ICU"

Best Research Poster

Danielle Daniels, MD: "Epidemiology of RSV bronchiolitis among young children in central New York before and after the onset of the COVID-19 pandemic"

People's Choice Award:

Jennifer Black, MS, CGC: "Deletions of 14q32.2 result in severe neurodevelopmental outcomes and multiple congenital anomalies: three affected males and review of the literature"

In case you missed them, the winning posters are attached.

Open House!

In just two months, UPAC will be making the official move to the new Nappi Wellness Institution. The moving date is now set for the first week in July (after the holiday). In the meantime, you are all invited to the open house on Friday, June 2^{nd} from Noon -2 PM.



They-PAS

The Pediatric Academic Society held its annual conference in Washington, DC from April 27-May 1 (in fact it is happening even as we go to press). Our department was once again extremely well represented including posters, presentations, oral abstracts, and workshops. We hope to have more details next month but congratulations to



Drs. Clarke, Nelsen, Schafer, Shaw, Sura, Teelin, Wasik, Janovicova and El Nakib for their participation!

(Picture is

enroute to PAS with the famous Dr. Blatt airport poster in the background).

Turkish Delight

A delegation from Turkey came to visit the Upstate Golisano Center for Special Needs in February to glean information for improving care for people with autism back in their country. According to our very own Dr. Roane, the delegation was in New York City at the headquarters of Autism Speaks when members asked to view a program firsthand. Autism Speaks sent them to Upstate the next day.

https://www.upstate.edu/news/articles/20 23/2023-03-30-turkey.php

Program Social Worker Jessica Giannino described the visit to the Crier saying, "My main takeaway from this was that it was eye opening to see that the obstacles we face at GCSN are not only ones faced within our immediate community but nationally and internationally. It validates the importance of the work GCSN is committed to every day and makes me appreciative of the strong, innovative team we have." The other social worker who met with the delegates, Corinne Dilaura added, "It was a pleasure meeting the Turkish delegation and being able to share about the things we do and resources and services within CNY and New York State."

Pediatric Clerkship Team

DUCATION CORNER

Many thanks and appreciation to our educator volunteers!!! We are extremely grateful for the following physicians who dedicated their time to teach and guide our students this past year. We couldn't have done it without you, and we know our students hold you in high esteem.

Formative Standardized Patient Exam: Faculty: Anjali Sura, Gloria Kennedy, Joan Pellegrino, Elizabeth Lodge, Tyler Greenfield, Rhonda Philopena

Residents: Colleen Feeney, Simi George, Andrew Brooks, Ayat Siddiqui, Adil Siddiqui, Lauren Thomas, Saber Jan, Rida Sherwani, Laaibah Ejaz, Clara El Nakib, Akilah Evans, Sarah Hendrix, Holly Stacey, Alex Ramirez, Ahmed Eltayeb

Fellows: Danielle Daniels, Alex Dang, Mitchell McKinnon

Evidence Based Medicine Discussion: Faculty: Anjali Sura, Andi Dvorak, Jenica O'Malley, Kim Rush, Stuart Trust, John Andrake, Mide Ajagbe, Elizabeth Lodge, Jennifer Myszewski, Nusrat Zaman, Andrew Osten, Kerry Biggs, Matt Mittiga, Melissa Schafer

Fellows: Danielle Daniels, Heather Ross, Alex Dang

History and Physical Exam Skills Session (Sim Center): Holly Stacey, Alex Ramirez, Ahmed Eltayeb, and Alan Blayney

Volunteering is a great way to become involved in medical student education and is always a wonderful addition to CVs! If you are interested in learning more about our clerkship volunteer opportunities, then please feel free to reach out to one of us!

-Jen Nead (director), Leah Bennett (associate director), and Chris Kuehnle (coordinator)

In the News: Cycle of Health

Several of our Peds faculty appeared on on WCNY-TV's Cycle of Health in April. On 4/20 the topic was Organ Donation and featured Upstate's own Drs. Dorgam Badram and Angela Wratney. If you missed it you can live stream all their previous shows at https://video.wcny.org/show/cycle-health/episodes/season/15/

CNY Central: "Doctors Say..."

Our very own Dr. Jenica O'Malley was quoted in an article entitled, "SCSD looks to hire 50 teaching assistants to support special needs students" in CNY Central.

https://cnycentral.com/news/local/scsd-looksto-hire-50-teaching-assistants-to-supportspecial-needs-students#



A Cochrane Review

Dr. Aamer Imdad was featured on Upstate Online for his new Cochrane Review entitled, "Upstate-led study finds stool transplants more effective than antibiotics for treating recurring, life-threatening gut infections" https://www.upstate.edu/news/articles/2023/2 023-04-25-fecal1.php

Where Are They Now? Dr. Marvin Mata

Marvin Mata (2010) shared a fascinating story that made national news of one of his patients where he works in Peds Critical Care in Alexandria, LA.



https://www2.cbn.com/article/miracle/boy-recalls-being-held-jesus-during-pool-drowning

Dr. Amy Reiss

We were excited to hear from former resident Dr. Amy (Goldberg) Reiss who has been practicing in New York City since graduating the program in 2002. Amy shared with the Crier that her daughter Ariel (pictured below) is going to apply to Upstate's new Early Decision Program for Medical School. Amy and Ariel are both advocates for advocating for JDRF and affordable insulin for all.



(Amy pictured with Congressman Nick Lalota).

Ariel is a Type 1 Diabetic and is pictured on JDRF's promotional materials.



She is heading to Washington DC to advocate with congress. We are proud of Amy and her daughter and would be delighted if Ariel were to end up in our program someday!

Congratulations Jason!

Based on results of campus-wide SUNY Faculty Council Elections, our very own Jason Zeleny who was elected to the Council Operations Committee.

78 Baskets!

Lest you thought there was no such thing as an Easter Bunny, the Maureen's Hope Easter Bunny delivered 78 Spring Baskets to Upstate Golisano Children's Hospital and placed them in the rooms of sleeping pediatric patients so all children spending Easter in the hospital woke up to a spring basket that morning. Thank you to Maureen's Hope, and all the volunteers and donors including Zulma Tovar-Spinoza (for her new book "Now that you are Here").

Zoo Not Forget!

On Monday, May 8th, the Department is hosting an evening at the Rosamond Gifford Zoo from 5:30 – 8:00 PM. It is a private event for all who previously registered. No tickets are needed at the gate.

MAY BIRTHDAYS

5/2 Matt Mittiga

5/5 Ellen Schurman

5/8 Heather Wasik, Asalim Thabet

5/9 Ghina Fakhri

5/10 Andrea Dvorak

5/11 Chris Lopez

5/14 Zafer Soultan

5/16 Gina Romero

5/17 Aubrey Mariani

5/18 Andrew Osten, Debby Carlson,

Katy Scott

5/20 Lou Pellegrino

5/27 Jaclyn Sisskind

5/31 Kerry Biggs



PLEASE JOIN US

FRIDAY, JUNE 2 | NOON - 2 PM 725 East Adams Street, Syracuse



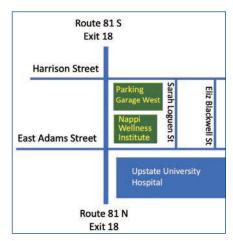
The Nappi Wellness Institute is a 200,000 square foot outpatient facility across the street from Upstate University Hospital.

The innovative space is designed for collaboration, offering primary and specialty health care services under one roof. Our providers bring a focus on wellness to all practices and resources for healthy aging and brain health, including Alzheimer's disease research and care. The Nappi Wellness Institute supports preventive health, which can help reduce hospitalization and emergency treatment and lead to better health for all.









The Nappi Wellness Institute is easily accessible off Route 81 in Syracuse. Event parking is in garage west.

Deletions of 14q32.2 result in severe neurodevelopmental outcomes and multiple congenital anomalies: three affected males and review of the literature

Jennifer Black¹, Maayke de Koning¹, Robert Roger Lebel¹, Scott Smith¹, Melissa Byler¹, Arie van Haeringen¹, Claudia Ruivenkamp², Himanshu Goel^{3,4}

 Medical Genetics Section, SUNY Upstate Medical University, Syracuse, NY, USA. 2. Department of Clinical Genetics, Leiden University Medical Centre, Leiden, the Netherlands. 3. Hunter Genetics, Hunter New England Local Health District, Waratah, NSW 2298, Australia. 4. University of Newcastle, Callaghan, NSW 2308, Australia

INTRODUCTION

Deletions of 14q32.2 have been reported, and mostly involve the imprinted region resulting in paternal/maternal UPD(14)-like phenotypes. Few individuals with deletions sparing the imprinted region have been reported, and deletion size/gene involvement is highly variable. We report three males with similar deletions (2.5 to 3.9 Mb), two of which spare the imprinted region, with similar phenotype including severe developmental disability, feeding problems, cryptorchidism, and dysmorphic features.

CASE REPORTS

Patient 1

Born at 39 weeks by C-section due to fetal distress, to a 21-year-old primigravid woman; pregnancy complicated by cerebral ventriculomegaly, hydronephrosis, and IUGR. Birth weight <3rd percentile, length 5th, head circumference 4th. Fetal anomalies confirmed by ultrasound. Neonatal period: feeding problems, hypoglycemia, bilateral cryptorchidism, hypotonia, cephalohematoma. Newborn screen positive for SCID, not confirmed by diagnostic testing.

Medical history: t-cell lymphopenia with recurrent URIs, dysphagia, constipation, FTT, esotropia, hyperopia, dextroscoliosis, positional plagiocephaly, and global developmental delays. Brain MRI: slightly prominent ventricles. Could track, smile, and roll over at 7 months but achieved no other milestones. At 6.5 years: intellectual disability, non-verbal, non-ambulatory, and incontinent. He has dysmorphic features (figure 1 and table 1).

SNP microarray: de novo 3.125 Mb deletion of 14q32.2 (98,029,521-101,154,362)arr[hg19].









Figure 1: (From left to right) Patient 1 at 1 month, 13 months, 4 years and 6.5 years.

Patient 2

Born at 41 weeks by C-section due to fetal distress, to a 42-year-old G2P1>2 woman; pregnancy complicated by IUGR. Birth weight 1st percentile, length 50th, head circumference 50th. Neonatal period: feeding problems requiring NG tube, bilateral cryptorchidism, glandular hypospadias, hypotonia.

Medical history: severe intellectual and developmental disability. Brain MRI: nonspecific mild ventricular enlargement and increased CSF space. At 20 years old: described as non-verbal but had first words at 6 years and occasional two-word sentences at 8 years. Can walk with a frame but mostly non-ambulatory, and incontinent. He has dysmorphic features (table 1).

SNP microarray: de novo 3.9 Mb deletion of 14q32.2q32.31 (98,890,898-102,783,552)arr[hg19].

Patient 3

Born at 36 weeks by spontaneous vaginal route to a 31-year-old primigravid woman; pregnancy complicated by premature rupture of membranes. Birth weight <3rd percentile, length and head circumference unavailable was SGA. Neonatal period: feeding problems requiring NG tube, hypoglycemia, bilateral cryptorchidism, and hypotonia.

Medical history: recurrent URIs, pyelectasia, nephroliths, dysphagia, FTT, constipation, strabismus, horizontal nystagmus, scoliosis, craniosynostosis of all sutures, hyperthyroidism, type 1 diabetes mellitus, three seizures starting at 17 years, intellectual and developmental disability. Brain MRI normal. At 21 years old: can roll over but achieved no other milestones. Non-verbal, non-ambulatory, and incontinent. He has dysmorphic features (figure 2 and table 1). SNP microarray: de novo 2.5 Mb deletion of 14q32.2 (98,534,687-101,064,183)arr[hg19].









Figure 2: (From left to right) Patient 3 at 12 months, 9 years, 14 years, and 19 years.

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Patient 1		-	BCLI18 CONV EML1 VÝ2 WAŘS1		анистиз тесяна
	Patie	nt 2:			

Figure 3: Location and gene content of 14q32.2 deletions identified in patients 1, 2, and 3; minimal overlapping region highlighted in red.

	Patient 1	Patient 2	Patient 3
Age at observation	6.5 y	8 y	21 y
Sex	M	M	M
Height	7 th %ile	10 th %ile	<1st %ile
Weight	<1 st %ile	<3rd %ile	32 nd %ile
Head circumference	6 th %ile	2 nd %ile	47th %ile
Low-set ears	*	+	
Wide nasal bridge	*	+	**
Small nasal passages	+	+	- *
Microstomia	+	+	
Micrognathia	+	+	1.0
High arched palate	+	+	
High prominent forehead	+	+	
Upward slanting palpebral fissures	+	~	+
Small/short palpebral fissures		+	
Epicanthal folds	2	+	+
Prominent smooth philtrum	+	+	
Micropenis	+		+
Pes planus	+	+	-
Spasticity	+	+, later onset	- (motoric restlessness)
Other	- Hypotelorism - Hypoplastic nipples - Tense limbs - Long feet - Toe camptodactyly - Toe overlap - Positional plagiocephaly	- Sparse eyebrows - Thick eyelashes - Anteverted nares - Depressed nasal bridge - Abnormal hair whorls - Occipital flattening - Large big toe - Long fingers and toes	Overarching big toe Left single transverse palmar crease Valgus deformity of ankles Hemangioma of neck and heel Metopic ridge

Table 1: Findings from the physical exams of patients 1, 2, and 3. (+) present, (-) absent.

CONCLUSIONS

All three patients had prenatal and postnatal growth restriction, feeding problems, severe global developmental disability, cryptorchidism, hypotonia, and similar dysmorphic features (table 1). The two whose deletions spare the imprinted region also shared recurrent infections, strabismus, and kidney problems. These deletions overlapped for 13 different genes, three of which are associated with autosomal dominant conditions: BCL11B, CCNK, and YY1 (figure 3).

Other reported patients with similar 14q32.2 deletions show some clinical overlap with our patients, but none have the same combination or severity of features. Those with smaller deletions involving BCL118 and CCNK had global developmental disability but no congenital abnormalities¹, while others with deletions involving only YY1 had mild to moderate neurodevelopmental problems and minor dysmorphic features². We propose that deletions involving these three genes result in a discrete clinical entity entailing a severe neurodevelopmental phenotype, characteristic facial features, and multiple congenital anomalies. We propose this be referred to as 14q32.2 deletion syndrome.

METHODS

Deletions were identified by clinical SNP microarray and analyzed according to the GRCh37 genome build.

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Nurse Driven Diabetic Ketoacidosis Fluid Titration Protocol Outside the ICU



Michelle Jeski MSN, RN, PCNS-BC, Emilie Hess MS, Margaret Anderson B Pharm, BCPPS, Alana Guidetti BSN, RN, David Hansen MD, MPH, Abeir Mohamed MD, Melissa Schafer MD, Angela Wratney MD, Roberto Izquierdo MD

Introduction

- Treating diabetic ketoacidosis (DKA) requires careful titration of a 2 bag IV fluid system, continuous insulin delivery, and hourly glucose monitoring.
- Current practice: Nursing notifies provider with blood glucose hourly, receives written order for 2 bag IV fluid rate change.
- At baseline, this process took an average of 16 minutes to complete.
- Our team identified an opportunity to reduce the time between blood glucose collection and fluid rate change.
- Nurse driven fluid titration is common in the ICU, but has not been reported outside the ICU in pediatric literature

SMART aim: Decrease the time from blood glucose to rate adjustment from 16 minutes to <5 minutes within 3 months in pediatric patients in DKA admitted to 12E.

Methods

- A multidisciplinary team was formed: Nursing, Quality,
 Pediatric Endocrinology, Pediatrics, and Pharmacy
- An RN driven protocol was developed to allow nursing to titrate the rates of the 2 bag system. Insulin was not titrated.
- The focus population was patients with DKA, aged 2-18 years on 1.5x maintenance fluids outside of the ICU.
 Children with a mixed pictures, concerns for cerebral edema or requiring critical care were excluded.
- Using our existing DKA policy and guideline, a calculator was built into the EMR which identifies the rate change needed based on the nurse entered blood glucose.
- Time to rate change was collected through EMR report, and complications assessed through a unit-based tracking system.

INCLUSION CRITERIA Pediatric patient (age 2-18 years) with Primary diagnosis of DKA Diabetes Type I Admitted to inpatient pediatric medicine On 1.5x total maintenance fluids EXCLUSION CRITERIA Age <2 years

Presence of severe headache or concern

On maintenance or 2x maintenance fluids

for cerebral edema upon presentation

Admitted to PICU or in PED

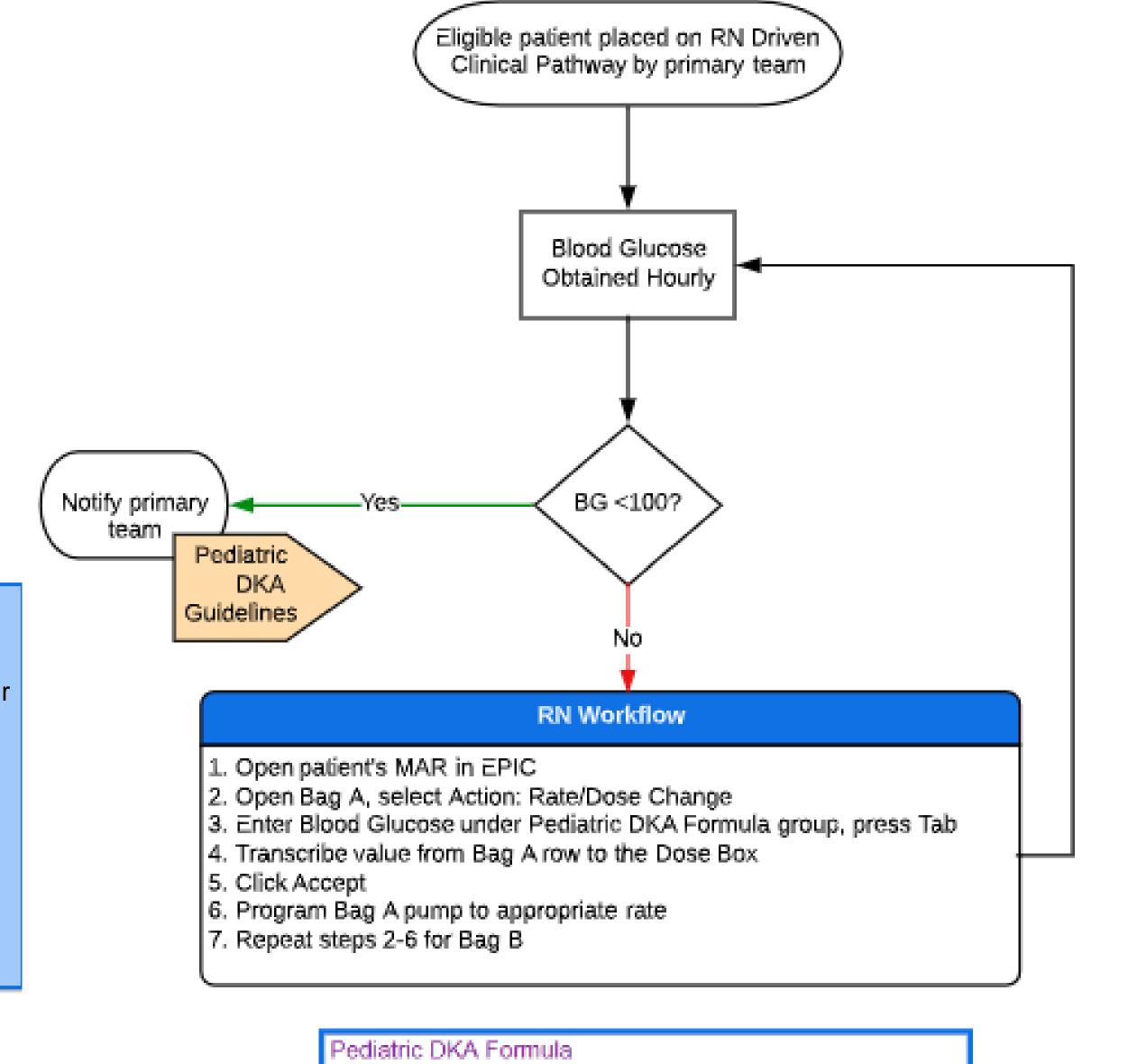
PRIMARY TEAM NOTIFICATIONS:

BG <100 mg/dL

DKA with HHS mixed

Diabetes Type II

- BG increase by >50 mg/dL/hour within 1 hour
- BG decrease by >100 mg/dL/hour within 1 hour
- Abrupt decrease in heart rate ≥ 20 BMP
- Diastolic BP >90 mmHg
- Insulin infusion interruption for any reason Mental status changes, recurrent vomiting, worsening/severe headache, age inappropriate incontinence



Blood Glucose (mg/dL)

1.5X Maintenance Infusion

NaCl 0.9% base fluid - BAG A

NaCl 0.45% base fluid - BAG

Results

- Twelve patients have been treated on the protocol with an average time to rate change of 5 minutes.
- No complications have been reported.
- Time on insulin has remained unchanged at 14.4 hours.
- Nursing and residents stated increased satisfaction with the RN driven protocol as care is more efficient.

Discussion

This nurse driven protocol has been effective in streamlining care for our patients with DKA outside of the ICU. Fluid titration has been described in the literature as an ICU based skill, pediatric floor nurses were able to accomplish this easily and achieved improved care for their patients. Using a EMR calculator facilitated safe and efficient care. Providing the RN with the ability to measure the blood glucose hourly and perform the fluid rate adjustment in real-time maximizes the patient time spent on appropriate fluid rates before the next hourly blood glucose collection. Future steps include expanding the protocol to other units and patients with DKA on alternative fluid regimens, as well as assessing other factors that influence time on insulin.

Acknowledgments

We would like to thank our EPIC team, Lindsay Bugge and Nancy Miller for their safe, innovative build to fuel the success of this QI project. Danielle Daniels¹, MD, Manika Suryadevara¹, MD, Zachary Wolf², MBA, MS, Joseph Domachowske¹, MD

¹Department of Pediatrics,, SUNY Upstate Medical University, Syracuse, New York; ²Clinetics, Durham, NC

Background

JPSTATE.

MEDICAL UNIVERSITY

- Respiratory syncytial virus (RSV) is the greatest contributor to lower respiratory tract infections (LRTI) in young children
- Non-pharmacologic interventions enacted to slow transmission of SARS-CoV-2 ignited a global disruption to RSV circulation with patterns that remain disrupted today

Objective

To describe differences in testing patterns and clinical characteristics for RSV bronchiolitis among young children in central New York before and after the onset of the COVID-19 pandemic.

Methods

A retrospective, cohort study was conducted using data collected from the EMR of SUNY Upstate Medical Center. Clinical and sociodemographic data were collected between October 2015 and January 2022 for children < 5 years with medical encounters associated with a bronchiolitis ICD-10 code and available testing results for RSV. Weekly data was electronically collected, graphically displayed, and reviewed using the Clinetic NOWcasting platform.

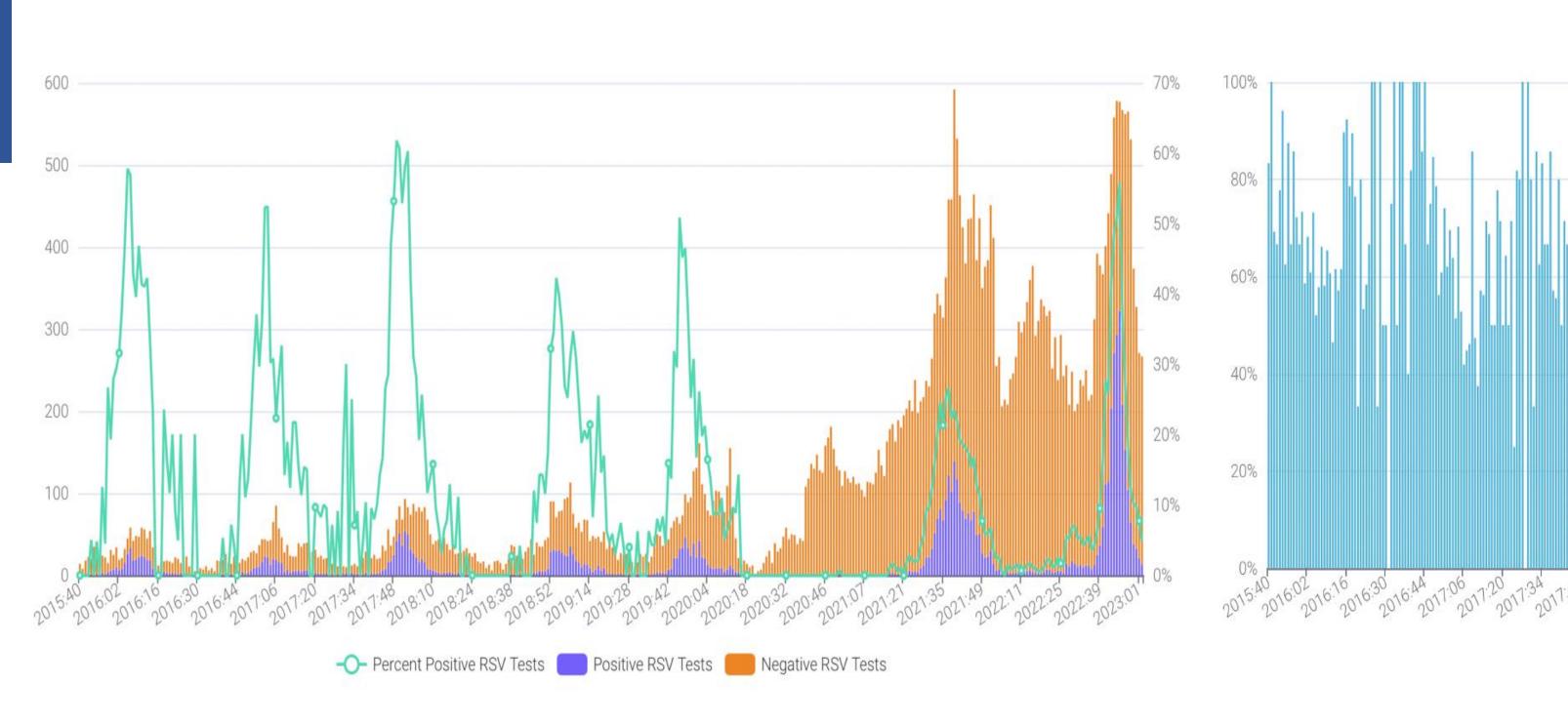


Figure 1. RSV testing patterns among children < 5 years before and after the onset of the COVID-19 pandemic

-O- Bronchiolitis Cases -O- Positive RSV Tests Among Bronchiolitis

Figure 3. Total bronchiolitis cases and positive RSV

pandemic. There was a significant difference in the

bronchiolitis cases between 2021 and 2022 (43.3%

number of RSV+ bronchiolitis cases for each season

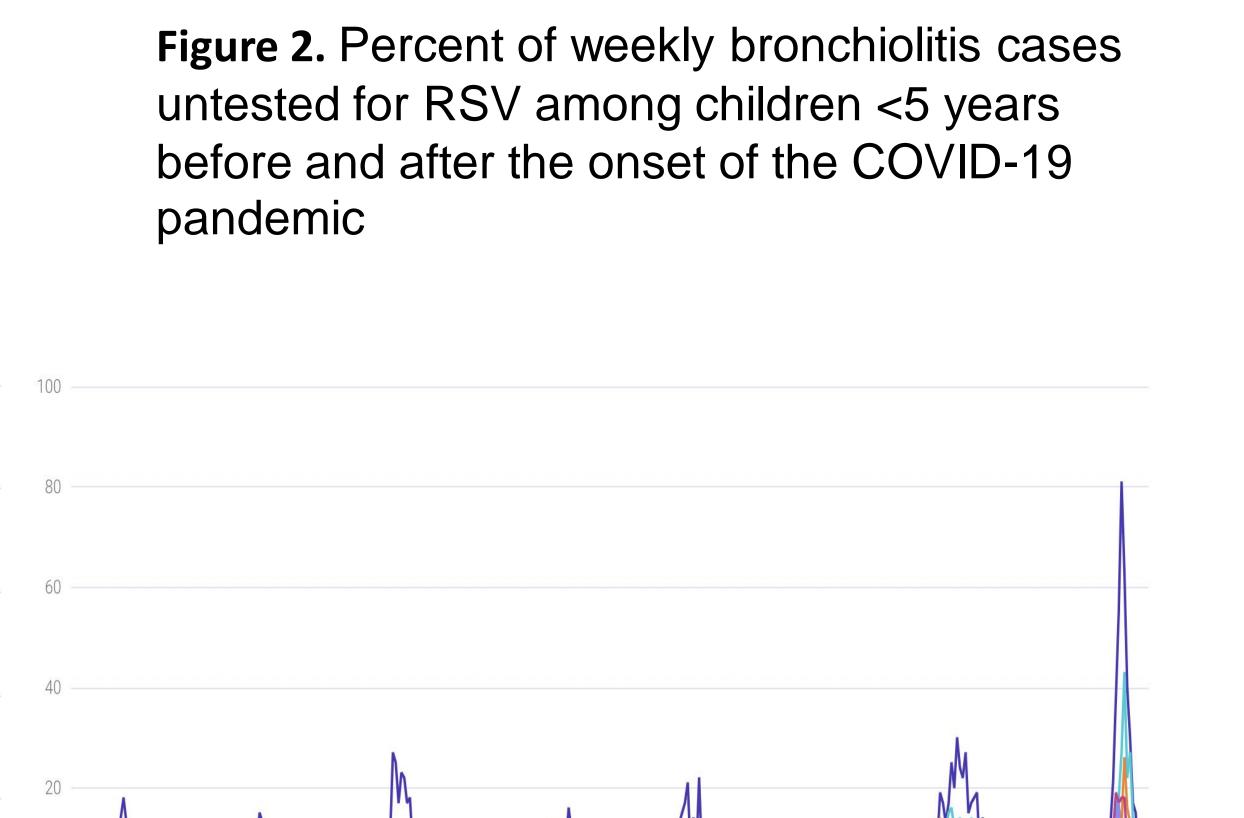
in 2021 vs. 69.5% in 2022, p<0.05). The absolute

tests among bronchiolitis for children <5 years

percentage of positive RSV tests among total

was 993 in 2021 and 1,141 in 2022

before and after the onset of the COVID-19



Percent Untested Among Bronchiolitis Cases

Figure 4. RSV+ Bronchiolitis cases by age cohort before and after the onset of the COVID-19 pandemic. Cohorts are reported in months

-O- 0-11 **-O**- 12-23 **-O**- 24-35 **-O**- 36-47 **-O**- 48-59



Golisano Children's Hospital

Following the onset of the COVID-19 pandemic, RSV testing increased (figure 1). To date, a sustained increase in RSV testing is observed among children with bronchiolitis (figure 2). Even among inpatient encounters, the average percentage of bronchiolitis cases untested for RSV decreased from 25% (pre-pandemic) to 8% (post-pandemic).

During the 2022 bronchiolitis peak:

- RSV predominated, more so than during previous seasons (figure 3)
- The proportion of children ≥12 months old with RSV bronchiolitis increased, with the largest increase observed in the older age cohorts (figure 4)
- RSV+ bronchiolitis hospitalizations increased by 124%, ICU stays by 246%, and mechanical ventilation by 186% compared to pre-pandemic peaks
- RSV+ bronchiolitis hospitalizations increased by 63%, ICU stays by 50%, and mechanical ventilation by 140% compared to the 2021 peak

Conclusions

- Increased testing practices have unveiled the true burden of RSV disease
- The disease burden is greatest among children <12 months, however extension beyond the first year is increasingly recognized and deserves ongoing surveillance
- Indices of severe disease suggest higher acuity of illness during the 2022 RSV bronchiolitis season
- Ongoing surveillance to identify vulnerable populations and shifting patterns is necessary to ensure appropriate distribution of preventative agents once available