Nicklaus Children's Hospital

The 2021 VIRTUAL 12th Annual Scholarship Week

Nicklaus Children's Hospital
Research Institute

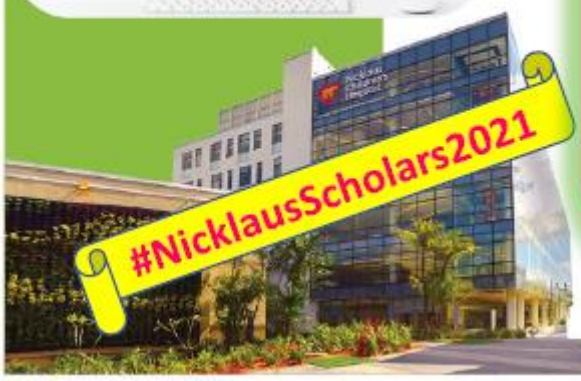
Organized by

Nicklaus Children's Hospital
DEPARTMENTS OF
MEDICAL EDUCATION & NURSING
&
THE RESEARCH INSTITUTE

The Week of May 31-June 4th, 2021

EVENT HIGHLIGHTS

- E-Posters, Digital Abstracts, Poster Summary Formats
- Scheduled author presentations by themes/ specialty (Schedule to follow)
- June 4th 2021 8:00am-9:30am – WINNING ABSTRACTS VIRTUAL ORAL PRESENTATIONS



12th ANNUAL SCHOLARSHIP WEEK

(Virtual)

May 31st - June 4th 2021

Underlined Author: Denotes a "Trainee"
** - Denotes a Winning Peer-Reviewed Abstract
* - Denotes a Presenting Author
§ - Denotes Late Breaking / Non-Rated Abstract

ORIGINAL RESEARCH

I- RESIDENTS ABSTRACTS:

Abstract #1: (RES-001-OR)
Lopez Gonzalez MS*; Fonseca AG; Colon Guzman S; Castro Rivadeneira M; Felipez LM; Caicedo Oquendo LF. ***"Impact of Telehealth in Our GI Practice in South Florida".***

OR-Introduction

In December 2019, clusters of patients in health centers in Wuhan, China, were found to have pneumonia of unknown cause, linked to a seafood market. A novel, highly infectious coronavirus, SARS-CoV-2, was isolated and by July 2020 became responsible for more than 13 million cases worldwide. As of July 9, 2020, 17000 pediatric cases were reported in FL, USA, with numbers rising. Throughout the COVID-19 pandemic, telemedicine (TM) has proved an innovative tool and has displayed a vital role in the continuity and sustainability of patient care. Individuals with chronic conditions are particularly susceptible to COVID-19 (C19), and medication compliance and disease optimization are essential ways to mitigate the severity. Our gastroenterology team at Nicklaus Children's Hospital (NCH) have used multiple platforms, including our hospital's NCHS Anywhere® (NCHA), Doximity®(DO), FaceTime®(FT), and Zoom®(ZM). We present a retrospective observational study in which we measure TM benefits and barriers for both patients and providers.

OR-Hypothesis/Study Objectives

OR-Study Design, Methods, and Statistical Analysis

A retrospective observational study was performed. A chart review of a database of all visits made via telehealth from March 27, 2020 to June 1, 2020 by 9 specialists. Data obtained included: patient demographics, diagnosis, follow up visit (FU) or new to the service, platform used for the encounters, type of insurance, RVU evaluation and

technical issues during the visit. Data from the same timeframe in 2019 was used for comparison. For IBD patients assessed by Improve Care Now (ICN): status of disease, flare during quarantine and steroid use. A survey was sent to all 9 GI specialists (GIS), consisting of 10 questions asking satisfaction rate and difficulties with use of TM.

OR-Main Results

A total of 1052 charts, of patients from 0 to 23 years old (y/o). Mean age of 8.78 y/o. 47.9% were female, 52.1% male. The most common diagnosis was constipation (20%) followed by GERD and IBS. Platforms used: NCHS®86%, DO®13.5%, uncharted 0.5%.When comparing data from 2020 to 2019 in the same timeframe/same GIS, RVUs were found to go from 2970 to 1473, number of visits from 2265 to 1052, FU vs new patients appointments had a proportion of 63.8 and 36.2%, vs 84% and 16%. Insurance use in 2019 was 45% commercial and 55% Medicaid, compared to this year 49.5% and 49% with 0.5% self-paid.

A total of 82 IBD patients were analyzed as a subgroup. Of those, 70% had Crohn's Disease, 29% Ulcerative Colitis, and 1% indeterminate IBD. 2020 to 2019 data: remission(R)rate:37% vs 77%, active disease(AD) rate:63% vs 23%, prednisone free rate: 96% vs 91.4%.

Within the GIS survey: 5/9 reported they were very satisfied with TM. 5/9 felt comfortable with patient communication. How often they plan to use TM in the near future, 1/9: 75% TM, 25% in-person encounters; 1/9: 50%-50%; 4/9: 25% TM, 75% in-person encounter; and 1/9 only do in-person encounters. 4/9 thought technical difficulties and lack of physical examination were the most problematic aspects of the practice. The most frequently used platforms in order were: 1) NCHA®, 2) DO®, 3) ZM®. GIS found easier to use DO®. Patient population thought by GIS to benefit the most with TM: 1.wheelchair or technology-dependent patients (TDP), 2.IBD patients, 3.post-procedural follow-ups. 5/9 believed TM helps with time management of the practice. 9/9 agreed that patient health status is more reliable with follow-up. 4/9 had no trust on vital signs taken at home.

OR-Conclusion and Significance

The Covid 19 pandemic changed the modern world and medical practices. With the requirement of quarantine and social distance, an urgent need for alternative means of attending medical appointments arose. From having a minor role in patient care, TM has gained an essential role. We concluded: most encounters were FU, likely due to difficulty to assess a new patient w/o an in-person meeting, ratified by the GIS perception through the survey. NCHA was the most used platform, but not the most user friendly according to our GIS, which could be taken into consideration for improvement. 50% RVUs

reduction represented an important financial impact. Most of the IBD FU encounters were of patients with AD, which contrast with NCH-ICN data from 2019 were most were in R, but explained by the fact that patients with AD needed closer follow up and benefited from not going to the hospital and choose TM as a useful tool. A 4% increase in steroid use was observed, possibly from a more aggressive use of steroids treatment to avoid in person visits and flares by the GIS.
As described by the GIS in the survey, TM still has multiple barriers in terms of technology, communication and reliability.TM is possibly a convenient method to provide healthcare possibly for: wheelchair or TDP due to difficult transportation logistics (during COVID-19 Pandemic and potentially after) and/or IBD patients on biologics who could be consider to have higher risk of SARS-CoV-2 infection and required a closer follow up.

Abstract #2: (RES-002-OR)

Napky Raudales P*, Ness-Cochinwala M, Sendi P, and Totapally BR. ***"Epidemiology and outcomes of necrotizing enterocolitis in preterm neonates".***

OR-Introduction

Necrotizing enterocolitis (NEC) is a common gastrointestinal emergency in neonates. Historically, it has been associated with lower birth weight and gestational age (GA). Despite advances in treatment strategies, case fatality remains high, stressing the need to identify risk factors with poor outcomes.

OR-Hypothesis/Study Objectives

To evaluate the epidemiology and outcomes of NEC in preterm neonates.

OR-Study Design, Methods, and Statistical Analysis

A retrospective analysis of the Healthcare Cost and Utilization Project 2016 Kids Inpatient Database was performed. The database was filtered using respective ICD-10 codes to identify preterm neonates (<37 weeks of GA) with NEC. GA were grouped into extreme prematurity, very premature, and moderate to late prematurity. Various risk factors associated with mortality in preterm neonates with NEC were identified and chi-squared test was used to compare categorical variables. Results were presented as Odds ratio with 95%CI. Multivariable binary regression analysis was done for etiological factors associated with mortality. Continuous variables are presented as medians with interquartile range (IQR).

OR-Main Results

Among a total of 365,314 preterm neonates, 4,734 (1.3%) had the diagnosis of NEC. Among preterm neonates with NEC, males were 54.5% and whites were 37.7% (Table 1).

The prevalence of NEC was highest in the black race. The median length of stay was 54 days (IQR 27-96), and median charges was $414,250 (IQR 186,844-903,484). The prevalence and case fatality rate of NEC increased with increasing in prematurity (Table 2). Overall case fatality rate with NEC was 15.2%. The case fatality rate was significantly higher when associated with acute kidney injury, intraventricular hemorrhage (IVH), sepsis, disseminated intravascular coagulation (DIC), invasive mechanical ventilation, arterial line, CPR, congenital heart disease (CHD), and invasive abdominal procedures (Table 3). However, on multivariable regression analysis, the presence of IVH and CHD did not change mortality (Table 4). The proportionate mortality from NEC was highest in very premature group compared to the other two groups (Figure 1).

OR-Conclusion and Significance

NEC is common in extreme premature neonates and in black race. Case fatality rate decreased with increase in GA, however, proportionate mortality rate is highest in the very premature group compared to extreme and late premature groups.

Table 1. Demographic characteristics of preterm neonates with NEC.

Variable	Number (95% CI)	Distribution % (95%CI)	p-value
Sex			0.385
Male	2579(2361-2797)	54.5(52.7-56.2)	
Female	2155(1978-2332)	45.5(43.8-47.3)	
Race			0.032
White	1541(1379-1703)	37.7(35.0-40.6)	
Black	1332(1148-1519)	32.6(29.7-35.6)	
Hispanic	722(604-841)	17.7(15.2-20.5)	
Other	491(408-574)	12.0(10.3-14.0)	
Payer			0.476
Government	2939(2669-3208)	62.1(59.6-64.6)	
Private	1558(1403-1712)	32.9(30.6-35.4)	
Other	234(186-281)	4.9(4.0-6.0)	
Income			0.183
Quartile 1	1761(1563-1959)	37.6(35.0-40.2)	
Quartile 2	1167(1052-1288)	25.0(23.2-26.8)	
Quartile 3	1024(912-1136)	21.9(20.1-23.7)	

Quartile 4	730(629-831)	15.6(13.7-17.7)	
Location			0.292
“Central" counties of metro areas of >=1 million population	1736(1501-1972)	36.8(33.0-40.7)	
"Fringe" counties of metro areas of >=1 million population	1096(932-1260)	23.2(20.4-26.3)	
Counties in metro areas of 250,000-999,999 population	879(724-1035)	18.6(15.7-21.9)	
Counties in metro areas of 50,000-249,999 population	423(350-496)	9.0(7.6-10.6)	
Micropolitan counties	350(280-421)	7.4(6.2-8.9)	
Not metropolitan or micropolitan counties	236(191-280)	5.0(4.2-6.0)	

Table 2. Prevalence, case fatality, and proportionate mortality rate of NEC in prematurity groups.

Prematurity Groups	Total NEC N (95% CI)	Prevalence % (95% CI)	Mortality N (95% CI)	Case Fatality Rate (95% CI)	Proportionate Mortality %
All Preterm	4734 (4324-5145)	1.3(1.2-1.4)	717(624-809)	15.2(13.7-16.7)	7.1
Extreme Preterm	2096 (1881-2311)	7.9(7.3-8.6)	501(433-569)	24.0(21.7-26.4)	6.8
Very Preterm	1418 (1277-1558)	3.5 (3.2-3.8)	134(106-162)	9.5(7.8-11.5)	11.6
Moderate to Late Preterm	1242 (1109-1375)	0.4 (0.4-0.5)	85(59-111)	6.9(5.1-9.2)	5.3

Figure 1. Prevalence and proportionate mortality percentages for preterm neonates with NEC by preterm categories.

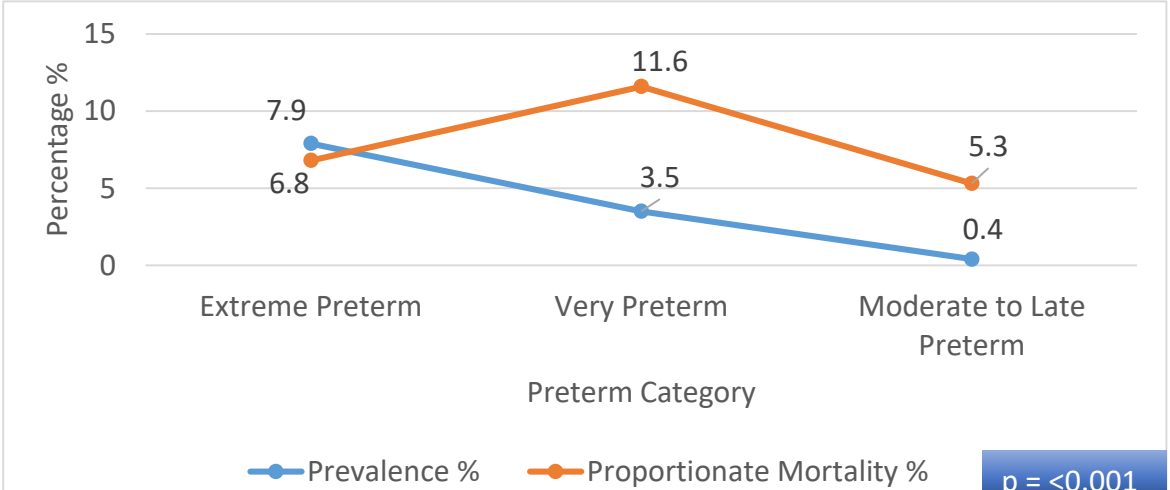


Table 3. Univariate analysis of risk factors associated with mortality in preterm neonates with NEC.

Organ Dysfunction	Mortality % (95% CI)	OR (95% CI)	p-value
Intraventricular Hemorrhage	22.1(19.3-25.2)	1.9(1.6-2.4)	<0.001
Congenital Heart Disease	17.3(15.3-19.4)	1.5(1.2-1.8)	<0.001
Invasive Mechanical Ventilation	21.0(19.0-23.1)	4.5(3.4-5.9)	<0.001
Cardiopulmonary Resuscitation	63.9(53.5-73.2)	10.9(7.0-17.1)	<0.001
Arterial Line	23.9(20.5-27.7)	2.0(1.6-2.5)	<0.001
Acute Kidney Injury	37.3(31.9-43.0)	3.8(3.0-5.0)	<0.001
Invasive Abdominal Procedure	30.3(27.0-33.7)	3.7(3.0-4.5)	<0.001
Sepsis	29.1(25.4-33.1)	2.9(2.3-3.6)	<0.001
Disseminated Intravascular Coagulation	49.0(42.0-56.1)	6.1(4.5-8.3)	<0.001

Table 4. Multivariable binary regression of risk factors associated with mortality in preterm neonates with NEC.

Organ Dysfunction	OR (95% CI)	p-value
Intraventricular Hemorrhage	1.0(0.8-1.2)	0.808
Congenital Heart Disease	0.8(0.6-0.9)	0.008
Invasive Mechanical Ventilation	2.5(2.0-3.2)	<0.001
Cardiopulmonary Resuscitation	8.7(5.6-13.3)	<0.001
Arterial Line	1.4(1.1-1.8)	0.001
Acute Kidney Injury	2.0(1.6-2.6)	<0.001
Invasive Abdominal Procedure	1.7(1.4-2.1)	<0.001
Sepsis	1.6(1.3-1.9)	<0.001
Disseminated Intravascular Coagulation	3.2(2.3-4.4)	<0.001

Abstract #3: (RES-003-OR)

Sanchez-Solano N*; Bermudez Chicango D; Frade-Garcia A; Cruz H; Diaz-Barbosa M; Vallarino D; Totapally B; Llanos-Martinez A. ***"A Ten-Year Experience in the Management of Congenital Diaphragmatic Hernia in a Freestanding Children's Hospital".***

OR-Introduction

The overall survival for infants with congenital diaphragmatic hernias (CDH) has gradually improved. Outborn is recognized as a clinical predictor of mortality. Few studies have reported the survival of infants with CDH transfer to a freestanding Children's Hospital.

OR-Hypothesis/Study Objectives

The aim of this study was to describe the patients' characteristics and risk factors, management and outcomes of CDH in a Freestanding Children's Hospital.

OR-Study Design, Methods, and Statistical Analysis

This retrospective chart review study included all infants transferred to our institution within the first 3 days of life with a diagnosis of CDH between 2008 and 2018. The data collected included demographic variables; known predictors of mortality on admission; medical, surgical, and extracorporeal membrane oxygenation (ECMO) management; hospital survival and length of stay. Severity of CDH was calculated with the clinical predictor model developed by the CDH Study Group (CDHSG). We compared the results to data published by the CDHSG, using Epi Info TM 7.

OR-Main Results

A total of 34 infants were included in the analysis. Delivery occurred in a birthing center (n=2), at home (n=1) and in 18 different hospitals (only 2 of those performed CDH repair). The patients' characteristics, clinical predictors of mortality and risk category, medical, surgical and ECMO management, hospital survival and length of hospital stay for survivors are shown in the Table 1 and 2. Compared to the CDHSG, we have a lower rate of prenatal diagnosis (20.6 vs 66.8%), higher rate of non- left sided defects (38.2 vs 16.7%) and infants in the low severity group (50 vs 33%). ECMO support was started in 7 hours (20.5 %) at a median age of 14 hours. Surgical correction was done in 31 infants (3 died before surgery) at a median age of 9 days. Patch repair was conducted in 13 (42%). The overall hospital survival was 82.5 %, and 92.6% in those not requiring ECMO support.

OR-Conclusion and Significance

Most infants with CDH transferred to our institution were not diagnosed prenatally, and delivery occurred mostly in hospital with no surgical capabilities. This regional variation may be due to preferential delivery of prenatally diagnosed CDH infants in hospitals with surgical capabilities. Despite the lack of prenatal diagnosis, the survival and length of stay are comparable to that reported in the literature. Having feto-maternal center attached to a Freestanding children's hospitals might improve the care of infants with severe CDH.

Abstract #4: (DRES-004-OR)

Acloque S*; Miranda P; Gupta A; Saman DM. ***"Oral health practices, knowledge, and attitudes among Hispanic immigrant parents of pediatric dental patients in South Florida"*.**

OR-Introduction

OR-Hypothesis/Study Objectives

Purpose: To measure oral health practices, dental knowledge, and attitudes of Hispanic immigrant parents of children receiving dental care at Nicklaus Children's Hospital dental clinic by education level.

OR-Study Design, Methods, and Statistical Analysis

This exploratory cross-sectional study conducted between September-December 2020 had a convenience sample of 141 immigrant parents of Latin descent who completed a paper questionnaire.

OR-Main Results

A higher proportion (86%) of mothers participated in the study. When exploring immigrant Hispanic parent's practices, 87% brushed their children's teeth two or more times a day and 65% gave only water after brushing their children's teeth at night. Nearly 70% of parents routinely gave their children bottled water. Parents with at least some college scored significantly higher in in oral health knowledge scores than those with a high school education or less (61% vs 50%, respectively, P=.0043). Pertaining to parents' attitudes, 94% thought it was somewhat or very important to fix baby teeth, 62% thought it was very or somewhat easy to address their own dental needs, while 59% thought it was very or somewhat difficult to limit sugar in their child's diet.

OR-Conclusion and Significance

Parents' oral health knowledge and practices differed by education. These results may help in informing and tailoring education intervention programs, potentially leading to reductions in early childhood caries among children of immigrants.

Abstract #5: (DRES-005-OR)

Cardenas MB*; Meincken M; Saman D; Patel P. ***"Motivational Interviewing to Improve Pediatric Dental Visits after General Anesthesia".***

OR-Introduction

Explore the impact of motivational interview (MI) on parents of patients who have undergone comprehensive dental treatment and oral rehabilitation under general anesthesia (GA) to improve their dental return visits.

OR-Hypothesis/Study Objectives

Questions include: Would a MI phone call after GA increase attendance to the 6 month recall? Would a second MI phone call after the first recall increase attendance to the 12 month recall? We hypothesized that parents of children who underwent oral rehabilitation under GA who participated in a MI would have an improvement in attendance to the 6 month and 12 month recall visits.

OR-Study Design, Methods, and Statistical Analysis

An intervention based prospective case control study was conducted and data gathered from February 2019 to October 2020. 100 randomly selected parents of patients who underwent oral rehabilitation under GA at Nicklaus Children's Hospital during February 2019 - July 2019 where consented to participate. An 18 item questionnaire recording demographics was collected. 3 months after GA parents participated in an MI phone call. Phone interviewer followed a 5 minute script and encouraged patients to schedule an appointment for their 6 month recall. Attendance of patients at the 6 month recall appointment was recorded whether they attended the NCH dental clinic or another dental home. A second MI phone call occurred some time after the initial recall following the same 5 minute script and encouraged patients to schedule an appointment for their 12 month recall. Attendance of patients at the 12 month recall appointment was recorded whether they attended the NCH dental clinic or another dental home. Data was compared to a control group (n=172) of participants who underwent oral rehabilitation at Nicklaus Children's Hospital under GA during the dental date range but did not receive MI.

OR-Main Results

Our data suggests the incorporation of an MI was associated with an increase in attendance compared to the control. At the 6 month recall, 61.1% attended with MI compared to 38.4% in the control which had no MI. At the 12 month recall, 50.9%

attended with MI compared to 30.2% in the control which had no MI. MI was significantly associated with attendance to the 6 month recall visit when compared to the control [OR=2.52; 95% CI (1.43, 4.44); *P*=.001]. MI was significantly associated with attendance to the 12 month recall visit when compared to the control [OR=2.40; 95% CI (1.27, 4.54); *P*=.006]. A second MI does not drastically increase the number of attendances compared to one MI, but 85% who attended the 12 month recall also attended the 6 month recall.

OR-Conclusion and Significance

Motivational Interview showed promising results as a behavioral intervention and led to an improvement in attendance to the 6 month and 12 month recall visits. A second MI did not drastically increase the number of attendances compared to one MI. Further research will need to be investigated to determine long-term effects of MI counseling sessions to measure sustained behavior change.

Abstract #6: (DRES-006-OR)

Chery J*; Freitas V; Saman D; Gupta A. ***"Comparing Residents' Cavity Using Er,Cr:YSGG Laser Versus Conventional High-Speed".***

OR-Introduction

Minimally invasive dentistry is becoming more popular when treating children. One of alternatives to providing minimally invasive treatment in pediatric dentistry is the use the hard tissue laser.

Er,Cr:YSGG laser allows the clinician the capability of providing selective and precise caries removal that will preserve sound enamel and dental tissue, without the need of drilling with high-speed handpiece. Laser has the advantage of bactericidal effects, providing decontamination of the treated area. Besides, the use of laser has been associated with analgesic effects, that would decrease the need of local anesthetic, providing more comfort to the patient. The disadvantages in regard to lasers usage are up-front costs, high maintenance cost, lack of familiarity in hard tissue applications, the need for specialized training and slower cavity preparation.

The primary aims of this study are to compare pediatric dental residents' preparation times using Er,Cr:YSGG laser relative to convention high-speed preparations and assess differences in overall comfort level.

OR-Hypothesis/Study Objectives

We hypothesize that the operator (pediatric dental residents) preparation times will be significantly shorter than traditional preparation times in the long-run. Although the

cavity preparation with laser may take longer than conventional high-speed preparation, the total procedure time needed for cavity preparation with Er,Cr:YSGG may be comparable to conventional high-speed preparation.

OR-Study Design, Methods, and Statistical Analysis

A cross-sectional study to observe the difference between high speed and laser total preparation time and comfort level was conducted at the Department of Pediatric Dentistry at Nicklaus Children's Hospital. The study was carried out among five (5) pediatric dental residents in their second year of residency. The residents performed Class I or Class II composite restorations on patients with Frankel behavior score F4. A total of 131 cases were divided into two groups:

- Group I - High Speed : Conventional cavity preparation with high-speed handpiece in diamond/carbide burs under water cooling
- Group II - Laser: Cavity preparation with Er,Cr:YSGG laser Waterlase IPlus (Waterlase MD, Biolase World Headquarters, Irvine, CA, USA) at a wavelength 2780 nm. Laser energy was delivered through a turbo handpiece and MX7 fiber tip with adjustable air and water.

The total time of tooth preparation was recorded at each case. At the end of each procedure, the resident also documented both the difficulty level and comfort level.

Additional data collected included the preparation type (Group High Speed – Conventional high-speed handpiece/Group Laser – Er,Cr:YSGG laser), the location of the prepared tooth (maxillary/mandibular, right/left side), type of cavity preparation (Class I/Class II), depth of preparation (enamel, enamel/dentin 1/3, enamel/dentin 2/3), use of nitrous oxide, use of high-speed to refine the preparation and use of local anesthetic.
All the variables were observed descriptively. Chi-square or fisher exact test was used to determine the differences between the categorical variables. Median total time and median comfort level was calculated for each operator separately.

Adjusted random intercept model (adjusted for nitrous oxide, high speed to refine preparation, and anesthesia) to determine average difference between time and comfort level between group I (high speed) and Group II (laser). $P<0.05$ was considered to be significant.

OR-Main Results

A total of 131 cases were included in this study, 79 cases in Group I (high speed) and 52 cases in Group II (laser). The cases were completed among five (5) pediatric dental residents. At baseline, none of the participants reported a history of dental laser experience and all indicated a level of not comfortable with it use.

The statistical analysis showed there was an average additional preparation total time per tooth of 1.92 minutes with the use of laser in comparison to conventional high-speed, 95% CI (1.54,2.39). The participants reported 26% less comfort when using laser in

comparison to high speed, 0.74 95% CI (0.61, 0.91). Also, there was an increased need of local anesthetic and nitrous oxide usage during procedure in Group I when compared to Group II (p-value <0.0001).

OR-Conclusion and Significance

There was a difference observed in the total preparation time required to complete restorative treatment using Er,Cr:YSGG laser in comparison to the time taken to complete treatment using a conventional handpiece. Therefore, it is important that further comprehensive training amongst pediatric dental residents be performed in order to ensure confidence in lasers hard tissue applicational use.

Abstract #7: (DRES-007-OR)

Hui N*; Arevalo O; Gupta A; Saman DM; Wilson S. ***"Assessing the Current Research Infrastructure of Pediatric Dentistry Residency Programs: An Update for 2020"*.**

OR-Introduction

Learning the basic principles of scientific research and participating in scholarly activity is paramount in a pediatric dentistry residency experience. Quality evidence that bridges gaps between research and clinical practice often arises from residency research endeavors.

Nainar evaluated all the literature published in two prominent pediatric dental journals between 1969-1998. He found that nearly three-fourths of published evidence was from the weakest hierarchical level (i.e., descriptive studies) and suggested the need to improve the quality of evidence in the literature. According to Loevy et al., only 48% of abstracts and poster presentations by American Academy of Pediatric Dentistry (AAPD) members at two consecutive annual American Association of Dental Research (AADR) meetings in 1989 and 1990 became full-length publications. In a 2004 study by Rhodes et al., over a five-year timeframe, 7% of programs reported no resident publications and only 12% had more than 10 publications.

In 2004, there were 55 pediatric dentistry residency programs—each averaging four residents per year. Given the number of total pediatric dental residents, a large discrepancy exists between the amount of research projects being conducted each year and the number of resulting publications. Today, over 15 years later, there are 94 pediatric dentistry residency programs in the United States—a 71% increase compared to 2004. The aim of this study is to understand the characteristics of pediatric dentistry residency programs and evaluate factors associated with increased scholarly activity. The results will allow programs to examine their research curriculum, infrastructure, and resources, and make adjustments to close the aforementioned discrepancy between required resident scholarly activity and the number of resulting publications.

OR-Hypothesis/Study Objectives

To assess the research infrastructure of pediatric dentistry residency programs in the United States and to determine the variables associated with increased scholarly activity.

OR-Study Design, Methods, and Statistical Analysis

A 21-item questionnaire assessing program characteristics, components of research infrastructure, publication output and grants secured, perceptions on barriers to scholarly activity, and satisfaction with current research infrastructure was developed and administered to residency program directors in the United States from March 2020 through June 2020. All the variables in the study were analyzed descriptively. Chi-square or Fisher's exact test was used to assess differences in various program characteristics and program director perceptions treated as categorical variables between hospital-based, combined hospital-university-based, and university-based program types. Fisher's exact test was only used when cell counts were less than five. The number of residents, faculty, publications, and grants were analyzed as continuous variables, utilizing Kruskal-Wallis one-way ANOVA to report differences between the three residency program types. Simple logistic regression models were created to calculate estimated odds ratios (**OR**) and 95% confidence intervals (**CI**) for three dependent variables: faculty and resident publications (dichotomized as greater than or equal to 5 versus less than 5 publications from 2015-2019) and grants (dichotomized as at least 1 grant versus 0 grants from 2015-2019). Independent variables regarding program characteristics were included in the simple logistic regression models. Differences were considered statistically significant if $P<.05$. All analyses were completed in SAS (SAS Institute Inc., Cary, NC, USA) statistical software.

OR-Main Results

Of 94 pediatric dentistry residency programs, 43 (46%) responded to the survey; 22 (51%) were hospital-based (**H**), 15 (35%) combined hospital-university-based (**C**), and 6 (14%) university-based (**U**). Only two-year residency programs responded. The majority of the responding programs (81%) have been in existence for over ten years. Overall, 86% and 83% of the respective full-time and part-time faculty were board-certified, and 47% and 34% of the respective full-time and part-time faculty had advanced degrees. Combined hospital-university-based and U exhibited a higher number of median full-time faculty with board certification compared to H ($P=.004$). University-based programs had a higher median number full-time faculty with advanced degrees (e.g., Master's and/or PhD) than the other two program types ($P=.001$).

Most programs (77%) offered protected research time to residents with varying time allocation depending on program type ($P<.001$). Forty-four percent of programs did not offer any protected time for faculty ($P=.001$). In terms of grant procurement, 49% of the programs, only H and C, reported no funding sources for research and programmatic activities ($P=.007$). The remaining programs secured funding from various sources, including intramural, HRSA, NIH, foundations, and other private grants.

Median number of faculty publications, resident publications, and grants were highest for U (16, 5, and 5, respectively) and lowest for H (0, 0, and 0, respectively) (P<.001; P=.03; P<.001). Programs were 92% more likely to have ≥5 publications over the five-year period for each additional 1.0 full-time faculty member (95% CI [1.26-2.93]). A similar finding exists for full-time board-certified faculty and full-time faculty with advanced degrees. The only statistically significant variable associated with total number of resident publications were number of full-time faculty with advanced degrees (OR 1.88, 95% CI [1.22-2.90]) and part-time faculty with advanced degrees (OR 1.52, 95% CI [1.07-2.17]). Total full-time faculty (OR 1.9, CI [1.26-2.93]), full-time faculty with board certification (OR 2.1, CI [1.31-3.42]), and full-time faculty with advanced degrees (OR 2.3, CI [1.35-3.82]) were also associated with securing at least one grant versus none. With the exception of part-time faculty with advanced degrees and resident publications, part-time faculty were not statistically significant predictors of publications or grant procurement.

Regarding the perceived faculty expertise in research mentorship, U reported the highest percentage in the expert/skilled category (66.7%), followed by C (53.3%), and H (22.7%). Nearly 25% of the total program directors report a weak expertise among their faculty's ability to mentor residents on research methods. Eighty-six percent of H, 60% of C, and 50% of U program directors reported feelings ranging from neutral to high dissatisfaction with their current research infrastructure, and U were the most satisfied (P=.046). In terms of the importance of resident research, 73% of H, 87% of C, and 100% of U perceived it to be crucial or important (P=.001). Regarding perceived importance of research for clinical practice, 64% of H, 80% of C, and 100% of U felt that research for clinical practice was crucial or important; 12% of the responding programs reported that research was unimportant for clinical practice, and all of these were H (P<.001). Although 30% of program directors, comprised of H & C, perceived residency applicants' prior research experience or publications when considering them for the interview and match to be unimportant or irrelevant (P<.001), 50% of U perceived this experience to be crucial or important, while the other half exhibited a neutral opinion. Interference with revenue-generating clinical time, insufficient faculty time to mentor, and lack of protected research time for faculty were the largest barriers to publication.

OR-Conclusion and Significance

1. The number of total full-time faculty, full-time faculty with board certification, and full-time faculty with advanced degrees were associated with an increase in faculty publications and grants secured.
2. The number of full-time faculty with advanced degrees and part-time faculty with advanced degrees were associated with an increase in resident publications.
3. The number of full-time faculty with advanced degrees (e.g., Master's and/or PhD), along with the number of resident publications have decreased over the past 15 years.
4. Although university-based programs exhibit the infrastructure most conducive to increase scholarly activity, our findings suggest pediatric dentistry residency programs may lack resources and infrastructure to generate high quality research.

Abstract #8: (DRES-008-OR)**

Oliva M*; Aponte-Rodriguez S; Saman D; Gupta A. ***"Association Between Parental Acculturation and Children's Oral Health".***

OR-Introduction

Acculturation is a continuous sociocultural process that varies in rate and degree among individuals.[1] It is the change in behaviors, attitudes and values that occurs when an individual is continuously exposed to a new cultural system differing from their native cultural system.[1,2] It was first described by American anthropologist Otis Tufton Mason in 1895.[3] Acculturation can have beneficial effects on health care utilization and detrimental effects on diet.[3,4]

Differences in acculturation can play a role in children's overall and oral health.[2,6] Children of immigrant parents often experience more caries and barriers accessing care.[5] Acculturation has been shown to be predictive of oral health behaviors.[3]

OR-Hypothesis/Study Objectives

This study aimed to evaluate the association between parental acculturation and their children's oral health practices and decayed, missing, filled teeth in primary, permanent and mixed dentition

OR-Study Design, Methods, and Statistical Analysis

- A prospective, cross-sectional study was conducted at the Department of Pediatric Dentistry at NCH in Miami, FL, from May 2020 to August 2020.
- Inclusion criteria
    1. Foreign-born parents/legal guardians
    2. ASA I or II children
- Exclusion criteria
    1. Children with special needs
- Parents completed two questionnaires: one for acculturation and a second one on their children's oral hygiene/dietary practices (composite score). We recorded decayed, missing, and filled teeth during initial or recall examinations
- Pearson correlation coefficient was used to observe if patients standardized composite score and standardized acculturation score were associated with dmft index (primary dentition), DMFT index (permanent dentition), dmft/DMFT (mixed dentition)

OR-Main Results

- The final sample size consisted of 106 children
- Mean dmft/DMFT was 4.38, 1.86 and 2.63/0.54 in primary, permanent, and mixed dentition, respectively
- Most parents spoke Spanish at home (86.8%), worked full-time (44.3%) and lacked dental insurance (60.8%), (Table 1)

- Most children brushed their teeth twice a day (70.8%), used a fluoridated toothpaste (87.7%), and flossed less than once a week (50%). (Table 2)
- Most children had sweets (85.8%) and juice (78.3%) one to twice a day (Table 2)
- Overall, there was a significant negative association between acculturation and dmft/DMFT in primary and permanent dentition (p=0.0442, p=0.0126) (Figure 1 & 2)
- The more adequate OH/Dietary practices the higher dmft in primary dentition (p= 0.3423) (Figure 1)
- The less adequate OH/Dietary practices the higher DMFT in permanent dentition (p= 0.9573) (Figure 2)
- Overall, the higher the acculturation level the more adequate OH/Dietary practices (p=0.904)

OR-Conclusion and Significance

Our study found that high parental acculturation was associated with lower decayed, missing, filled teeth in children with primary, permanent and mixed dentitions. The association was statistically significant for primary and permanent dentition groups. Children in primary dentition with more adequate oral hygiene and dietary practices had greater dmft than those with less adequate practices. In contrast, the more adequate oral hygiene and dietary practices the lower the DMFT in permanent dentition group. These associations were not statistically significant. This study did not differentiate between untreated and treated decay. Another limitation is social desirability bias. These could explain the difference found between both groups.

To date limited studies have been conducted evaluating the association between acculturation and oral health. A study by Tiwari and Albino (2017) found that children of less-acculturated parents may experience poor health outcomes and a lower health care utilization. Similarly, Johansena et al. (2019) found an association between acculturation and increased consumption of sugar-sweetened beverages. These studies supports the higher dmft/DMFT in children of lower acculturated parents finding in our study

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RESULTS

Table 1 Demographics	N (%)
Person completing the questionnaire	
Mother	94 (88.7)
Father	12 (11.3)
Language spoken at home	
Spanish	92 (86.8)
English and Spanish	9 (8.5)
English	5 (4.7)
Highest grade or level of school completed by parent/legal guardian	
Some college or 2-year degree (College)	36 (34)
High school or equivalent	32 (30.2)
Less than high school	20 (18.9)
More than 4-year college degree	10 (9.4)
4-year college graduate	8 (7.5)
Dental Insurance Coverage	
No	64 (60.4)
Yes	41 (38.7)
no response	1 (0.9)
Parent/Legal guardian most recent dental visit	
0-12 months	69 (65.1)
over 24 months	28 (26.4)
12-24 months	7 (6.6)
no response	2 (1.9)
Parent/legal guardian employment status?	
Work Full Time	47 (44.3)
Work Part Time	35 (33)
Unemployed	24 (22.6)
Parent/Legal guardian annual household income	
20,000 – 40,000	56 (52.8)
less than 20,000	38 (35.8)
more than 40,000	8 (7.5)
no response	4 (3.8)

Table 2 Oral Hygiene & Dietary Habits	N (%)
Child's gender	
Female	57 (53.8)
Male	49 (46.2)
Frequency of sweets	
once to twice a day	91 (85.8)
two to four times a day	6 (5.7)
more than four times a day	2 (1.9)
no response	7 (6.6)
Frequency of juice	
once to twice a day	83 (78.3)
three to four times a day	7 (6.6)
more than four times a day	7 (6.6)
no response	9 (8.5)
Number of times parent brushes child's teeth	
twice a day	75 (70.8)
once a day	18 (17)
more than twice day	9 (8.5)
less than once a day	4 (3.8)
Number of times parent flosses child's teeth	
less than once a week	53 (50)
two to four times a week	39 (36.8)
more than four times a week	14 (13.2)
Usage of Fluoridated Toothpaste	
Yes	93 (87.7)
No	13 (12.3)

Figure 1

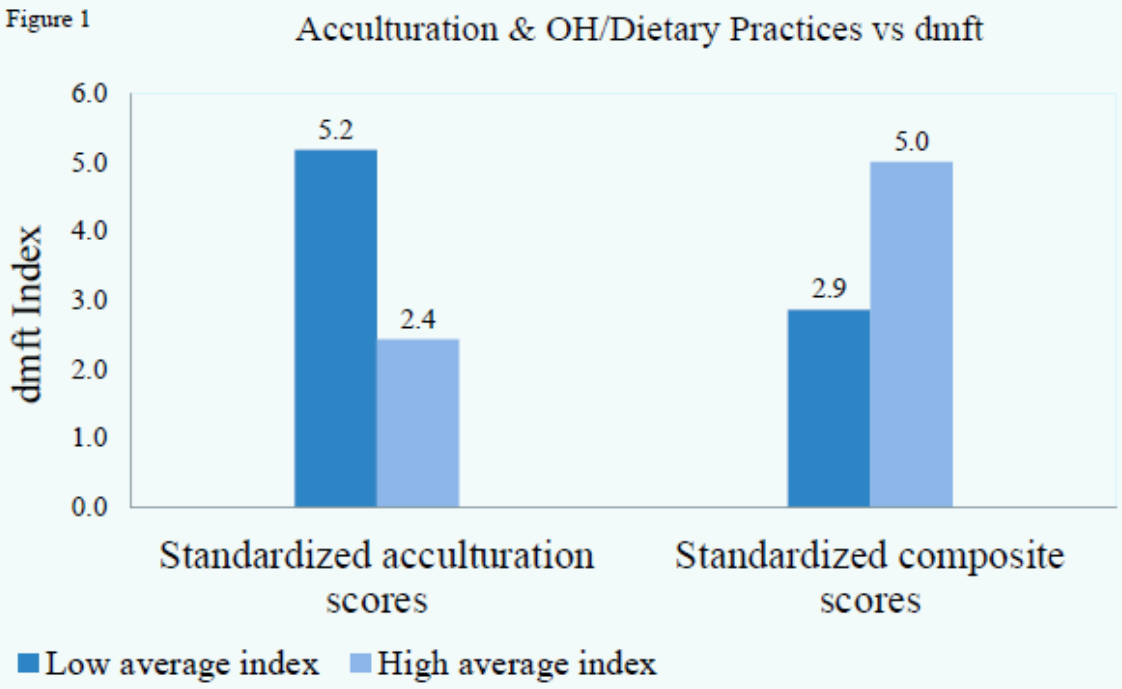
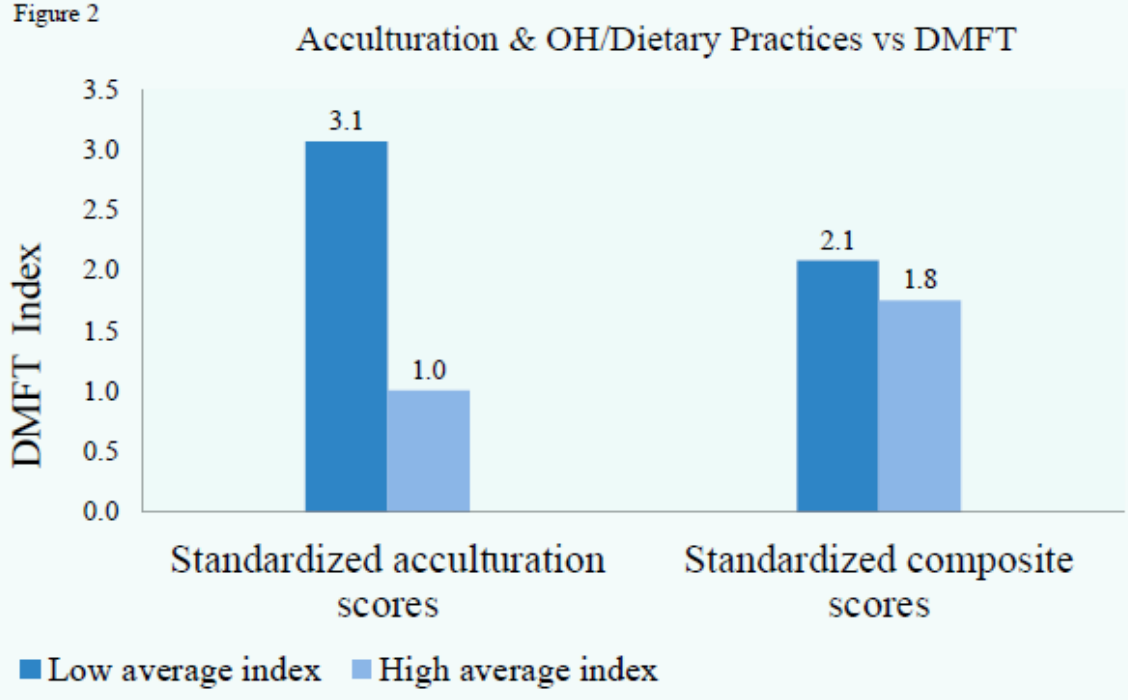


Figure 2



Abstract #9: (PHARM-009-OR)

Alvira-Arill G*; Chang N; Totapally B; Mulas N; Sotto I; Lee H. ***"Incidence of hyponatremia with the use of vasopressin in pediatric patients".***

OR-Introduction

Vasopressin, or antidiuretic hormone, is indicated for vasodilatory shock unresponsive to fluid resuscitation and catecholamines. Vasopressin stimulates V1a smooth muscle receptors to induce arterial vasoconstriction and V2 renal receptors to promote water reabsorption from the renal tubule. A potentially serious adverse effect of vasopressin use is hyponatremia which may result in altered mental status, brainstem herniation, and other neurologic injury.

OR-Hypothesis/Study Objectives

- Assess the incidence of hyponatremia and severe hyponatremia in children who received vasopressin for shock management or post-operative cardiac support
- Determine the association of vasopressin dose and infusion duration with incidence of hyponatremia
- Evaluate sodium trends after vasopressin discontinuation

OR-Study Design, Methods, and Statistical Analysis

- Single-center, retrospective chart review of < 18-year-old ICU patients who received vasopressin for shock management or post-operative cardiac support between August 2012 to August 2016
- Patients with cerebral edema, diabetes insipidus, or gastrointestinal bleeds were excluded
- Baseline characteristics recorded include age, gender, height, weight, and indication for use
- Serum sodium concentrations at baseline, lowest level during therapy, and last level taken within 24 hours of vasopressin discontinuation were recorded
- Hyponatremia and severe hyponatremia were defined as sodium levels less than 135 mmol/L and 130 mmol/L, respectively
- Additional clinical parameters recorded include fluid and sodium intake, highest BUN, lowest GFR, and urine output
- Other treatment-related parameters collected include lowest and highest dose of vasopressin used, length of vasopressin infusion, mortality while on vasopressin, and vasoactive-inotrope score (VIS)

OR-Main Results

- Incidence of hyponatremia was significantly higher during vasopressin infusion compared to baseline and after
- Longer duration of vasopressin infusion was significantly associated with higher incidence of hyponatremia
- Highest dose of vasopressin used showed no association with incidence of hyponatremia
- Sodium did not return to baseline 24 hours after infusion; however, a trend towards baseline was observed

OR-Conclusion and Significance

Abstract #10: (PHARM-010-OR)

Kuchnik M*; Sotto I; Vuong M; Rodriguez Y; Lee H. ***"Evaluation of advancing chemotherapy administration times and the incidence of hemorrhagic cystitis/hematuria"*.**

OR-Introduction

Hemorrhagic cystitis/hematuria is a known complication of high dose cyclophosphamide (>1gm/m^2) and ifosfamide and can cause significant morbidities.

The prophylactic use of Mesna and adequate hydration are essential in order to prevent bladder toxicity.

Daily rescheduling of chemotherapy to an earlier administration time is a practice amongst some institutions in an effort to decrease hospital length of stay.

OR-Hypothesis/Study Objectives

1. Evaluate the impact of advancing chemotherapy administration times during multiple-day regimens and the incidence of hemorrhagic cystitis/hematuria.
2. Determine if decreased hydration during chemotherapy leads to increased hemorrhagic cystitis/hematuria.
3. Evaluate the impact of advancing chemotherapy administration on the length of stay.

OR-Study Design, Methods, and Statistical Analysis

Retrospective, cohort chart review of pediatric patients (≤21 years) previously admitted to Nicklaus Children's Hospital between August 1, 2015 to August 31, 2020 who have received cyclophosphamide or ifosfamide as part of a chemotherapy regimen.

Patients admitted to the Hematology/Oncology or Pediatric Intensive Care Units who have received cyclophosphamide (>1gm/m^2) or ifosfamide (all doses) for multiple days of therapy.

Patients diagnosed with BK virus, autoimmune diseases such as systemic lupus erythematosus, rheumatoid arthritis or Crohn's disease, and received pelvic radiation or cyclophosphamide for non-oncology indications were excluded.

The study reviews 125 scheduled cycles of ifosfamide and 23 scheduled cycles of cyclophosphamide.

Wilcoxon rank sum test, Mann-Whitney test and Chi-Square test were used to analyze the statistical significance of categorical data using Microsoft Excel/REDCap.

Hematuria was defined as having either small, medium or large amounts of blood reported in urinalysis during the patients stay.

Baseline characteristics collected include age, gender, ethnicity, race, body surface area, and oncology diagnosis.

Treatment-related parameters collected include chemotherapy dose, cycle number, protocol name, time of administration, platelet count, occurrence of hemorrhagic

cystitis/hematuria, urinalysis, readmissions, urine output, Mesna and hydration administration methods, and the occurrence of early discharge.

OR-Main Results

Table 1. Baseline Characteristics

Baseline Characteristics	Chemotherapy Time Advanced (n=63)	Chemotherapy Time As Per Protocol (n=85)	P Value
Age (years) [median (IQR)]	13 (6)	10 (7)	0.003
Male [n (%)]	30 (47.6)	38(44.7)	0.83
Weight (kg) [median (IQR)]	58.88 (46.4)	27.73(28.4)	0.003
Height (cm) [median (IQR)]	158.4(42)	134.3 (40.7)	<0.001
BSA (m^2) [median (IQR)]	1.61 (0.9)	1.02 (0.7)	<0.001
Platelet Count (10k/µL) [median (IQR)]	232 (193)	208.5 (158)	0.157

Table 2. Oncology Diagnosis

Oncology Diagnosis [n(%)]	Chemotherapy Time Advanced (n=63)	Chemotherapy Time As Per Protocol (n=85)
Ewing's Sarcoma	25 (39.7)	16 (18.8)
Osteosarcoma	7 (11.1)	9 (10.6)
Synovial Sarcoma	3 (4.8)	3 (3.5)
Carcinoma of Choroid Plexus	0	7 (8.2)
Pineal Choriocarcinoma	3 (4.8)	0
Rhabdomyosarcoma	10 (15.9)	5 (5.9)
Large Cell Lymphoma	2 (3.2)	5 (5.9)
Extragonadal Germ Cell Tumor of Mediastinum	0	3 (3.5)
Embryonal Sarcoma	6 (9.5)	3 (3.5)
Medulloblastoma	4 (6.3)	15 (17.6)
Primary Undifferentiated Sarcoma	0	6 (7.1)
Neuroblastoma	2 (3.2)	5(5.9)
Germ Cell Tumor of the Brain	0	3 (3.5)
Retinoblastoma	0	2 (2.4)
Atypical Teratoid Rhabdoid Tumor	1 (1.6)	3 (3.5)

Table 3. Treatment Parameters

Treatment Parameters	Chemotherapy Time Advanced (n=63)	Chemotherapy Time As Per Protocol (n=85)	*P* Value
Actual Amount of Hydration [n (%)]			
<3000 mL/m^2/day	28 (44.4)	30 (35.3)	0.183
≥3000 mL/m^2/day	35 (55.6)	55 (64.7)	0.002
Urine Output [n (%)]			
<3 mL/kg/hr	49 (77.78)	49 (57.65)	0.012
≥3 mL/kg/hr	14 (22.22)	36 (42.35)	0.067
Discharged Early [n (%)]			
Yes	57 (90.48)	1 (1.18)	<0.001

Table 4. Incidence of Hematuria

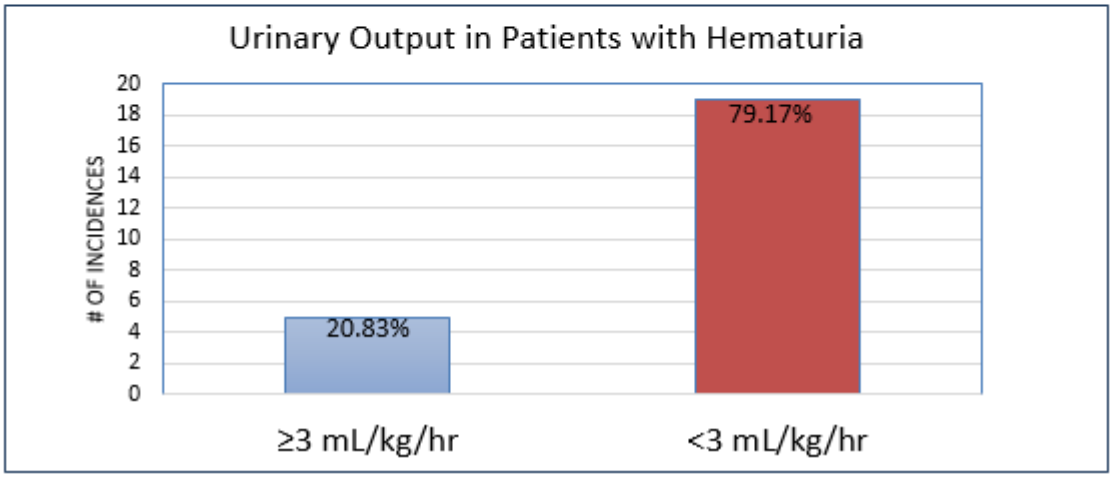
Clinical Outcomes	Chemotherapy Time Advanced (n=63)	Chemotherapy Time As Per Protocol (n=85)	P Value
Hemorrhagic cystitis	0	0	0.934
Hematuria	10 (15.9%)	14 (16.5%)	

Table 5. Incidence of Hematuria based on Hydration

Clinical Outcomes	≥3000 mL/m²/day (n=90)	<3000 mL/m²/day (n=58)	P Value
Hemorrhagic cystitis	0	0	0
Hematuria (n=24)	11 (12.2%)	13 (22.4%)	0.107

Table 6. Incidence of Hematuria based on Urine Output

Clinical Outcomes	<3 mL/kg/hr (n=98)	≥3 mL/kg/hr (n=50)	*P* Value
Hemorrhagic cystitis	0	0	0
Hematuria (n=24)	19 (19.4%)	5 (10%)	0.206



OR-Conclusion and Significance

- No statistically significant difference seen in advancing chemotherapy administration times and the development of hematuria/hemorrhagic cystitis.
- There was no statistically significant difference in the incidence of hematuria and fluid hydration.
- Patients who were advanced were more likely to be discharged early than the per protocol group.

Abstract #11: (RES-011-OR)**

Saini A*; Cavalcante R; Aquino Crisanto L; Sasaki J. ***"Outcomes of catheter-related arterial and venous thrombosis after enoxaparin therapy in neonates and infants with congenital heart disease".***

OR-Introduction

Central arterial and venous catheters are frequently associated with thrombosis in the cardiac intensive care unit (CICU). Enoxaparin is the treatment of choice for catheter-related thrombosis.

OR-Hypothesis/Study Objectives

The purpose of this study is to report the epidemiology and outcomes of central catheter-related arterial and venous thrombosis after enoxaparin therapy in neonates and infants with congenital heart disease following cardiac surgery & cardiac catheterization.

OR-Study Design, Methods, and Statistical Analysis

All patients less than one year of age cared for in CICU from January 2015 to January 2019 and treated with enoxaparin for central catheter-related thrombosis were included in the study. Treatment-related outcomes were stratified by age at diagnosis into two groups: 1) neonates < 1 month & 2) infants, 1 month - 1 year.

OR-Main Results

A total of 156 events in 122 patients were included in the study, 109 (69.6%) arterial and, 47 (30.1%) venous. The median age at diagnosis was 54.5 days. 103 events (66%) occurred after cardiac surgery, while 53 (34%) followed cardiac catheterization. Femoral catheters, both arterial & venous, accounted for 138 (88.5%) of the events. The median duration of central catheters preceding the diagnosis was 1 and 7 days for arterial and venous catheters, respectively. Occlusive thrombus accounted for 116 (74.4%) of the events, the rest 40 (25.6%) being non-occlusive. Therapeutic dose of enoxaparin was achieved after a median of 5 doses or 2 days. The therapeutic dose of enoxaparin was significantly higher in neonates compared to infants (1.8 Vs. 1.6 mg/kg, p=0.004). A comparatively higher number of infants were discharged home on enoxaparin (64.4% Vs. 29.1%). Treatment follow up was available for 106 patients of which 92 (86.8%) demonstrated either partial or complete clot resolution. The rate of resolution and duration of therapy were comparable between the two groups.

OR-Conclusion and Significance

Disproportionately high incidence of thrombosis with femoral catheters emphasizes the importance of utilizing alternative sites. Higher than recommended doses of enoxaparin is consistently required to achieve therapeutic Anti Xa levels in both neonates and infants. Thrombus resolution is achieved in most patients with enoxaparin treatment.

II- FELLOWS ABSTRACTS:

Abstract #12: (FEL-011-OR)

Abdul Kayoum A*; Rivera E; Reyes M; Almasarweh S; Ojito J; Burke R; Sasaki J.

"Outcomes Of Bloodless Congenital Cardiac Surgery On Cardiopulmonary Bypass".

OR-Introduction

Bloodless cardiac surgery defined as blood transfusion-free open-heart surgery, where cardiopulmonary bypass (CPB) circuits primed with crystalloid only and no intraoperative blood transfusion. Limited data have been published in this field.

OR-Hypothesis/Study Objectives

We asked whether blood conservative surgery is feasible in congenital heart disease.

OR-Study Design, Methods, and Statistical Analysis

We retrospectively reviewed patients who underwent bloodless cardiac surgery for congenital heart disease on CPB between January 2016 and December 2018. Our unique CPB system utilizes assisted venous drainage, bioactive coating, and reduced tubing size to decrease priming volume, and complement activation.

OR-Main Results

A total of 164 patients were reviewed (86 male and 78 female) at a median age of 9.6 years (range, 13 months-55 years), weight of 32 kg (IQR, 16-55), preoperative hemoglobin 13.7 g/dl (IQR, 12.6-14.9), and preoperative hematocrit of 40.3% (IQR, 37.2-44.3). Median CPB time was 81.5 minutes (IQR, 58-125), and median hematocrit coming off CPB was 26% (IQR, 23-29.7). Congenital Heart Surgery risk (STAT) category distributed in STAT 1 for 70 (43%), STAT 2 for 80 (49%), STAT 3 for 9 (5%), and STAT 4 for 5 (3%) of the patients. The majority (95%) of patients were extubated in the operating room with low complications rate during the hospital stay (7%). Only 6 (4%) patients needed a blood transfusion in the postoperative period with higher incidence of complications during the hospital course (LR 14.9; $p<0.001$). The median length of hospital stay was 3.6 days (IQR 2.6-5.6). There was no in-hospital mortality or 30 days mortality after surgery.

OR-Conclusion and Significance

Bloodless congenital cardiac surgery has a high success rate in selected low to medium surgical risk and even higher risk patients (STAT 3 and 4). Our patients had a low rate of complications and short hospital course. The blood product transfusion correlated significantly with a higher rate of complications during the postoperative course.

Abstract #13: (FEL-012-OR)

Girgis S*; Seitz A; Runyon J; Gupta A; Sasaki N. ***"Incidence of post-operative left outflow tract obstruction in patients with juxtaposed atrial appendages".***

OR-Introduction

Juxtaposition of the atrial appendages (JAA) is a rare congenital malformation in which the both atrial appendages are located either to the right or left of the great arteries. It is more commonly associated with tricuspid atresia (TA), transposition of the great arteries (TGA), double outlet right ventricle (DORV). Long-term hemodynamic consequence associated with JAA has not been well evaluated. We hypothesized that patients with JAA may develop left ventricular outflow tract obstruction (LVOTO) secondary to right or leftward deviation of conotruncus by JAA.

OR-Hypothesis/Study Objectives

Our aim was to review post-operative echocardiograms of patients with and without juxtaposed atrial appendages who had undergone surgery for congenital heart defects to evaluate for the development of LVOTO.

OR-Study Design, Methods, and Statistical Analysis

Patients with JAA (cases) from June 1995 to June 2016 were identified from our institutional echocardiographic database on patients who had associated lesions (TGA, TA or DORV) who had pre and post-operative echocardiograms performed at our institution. Systematic sampling was done to select control patients with associated lesions during the same period. The echocardiographic reports and surgical documentation for the identified patients were reviewed. Bivariate analysis including chi-square test was used to observe the association between JAA and development of gradient between cases vs control. P- value of <0.05 determined significance.

OR-Main Results

There were 20 cases with JAA, 18 of which were leftward juxtaposed and 10 of which were female. Ninety controls were included in the study for a case-control ratio of approximately 1:5. Of patients with JAA, the most common lesion with JAA was double outlet right ventricle- transposition type, followed by transposition of the great arteries with ventricular septal defect (Table 1). Defining left outflow tract obstruction as

gradient of ≥20mmHg found by Doppler interrogation, a total of 5 patients (25%) in the JAA group were found with obstruction, and 6 in the control group (6.7%) (Table 2), resulting in a significant difference in development of LVOTO (p=.013). Two patients with JAA underwent subaortic membrane resection following development of obstruction with complete alleviation of gradient in one patient and continued increasing gradient in the other. Three patients in the control group had subaortic membrane resection

OR-Conclusion and Significance

Although JAA has not been associated with additional hemodynamic burden, our study suggests that there may be increased risk of LVOTO after surgical repair in patients with JAA. This finding may provide guidance to long term post-operative care.

Figure 1: Demographics of Cases versus Controls

		Juxtaposed Atrial Appendage Orientation	*Normal Atrial Appendage Orientation*
Gender	Male	10	63
	Female	10	27
Cardiac Lesion	DORV Pulmonary Atresia	1	4
	DORV, TGA Type	6	30
	DORV, Uncommitted VSD	3	6
	Tricuspid Atresia	4	20
	TGA with IVS	1	5
	TGA with VSD	5	25
	Total	20	90

*DORV = Double Outlet Right Ventricle, TGA = Transposition of the Great Arteries, VSD = Ventricular Septal Defect, IVS = Intact Ventricular Septum

Figure 2: Number of Patients with Left Ventricular Outflow Tract Obstruction defined as left ventricular outflow tract gradient ≥20mmHg post-operatively

	Juxtaposed Atrial Appendage Orientation	*Normal Atrial Appendage Orientation*
DORV Pulmonary Atresia	0	0
DORV, TGA Type	1	4
DORV, Uncommitted VSD	3	0
Tricuspid Atresia	1	0
TGA with IVS	0	0
TG with VSD	0	2

*DORV = Double Outlet Right Ventricle, TGA = Transposition of the Great Arteries, VSD = Ventricular Septal Defect, IVS = Intact Ventricular Septum

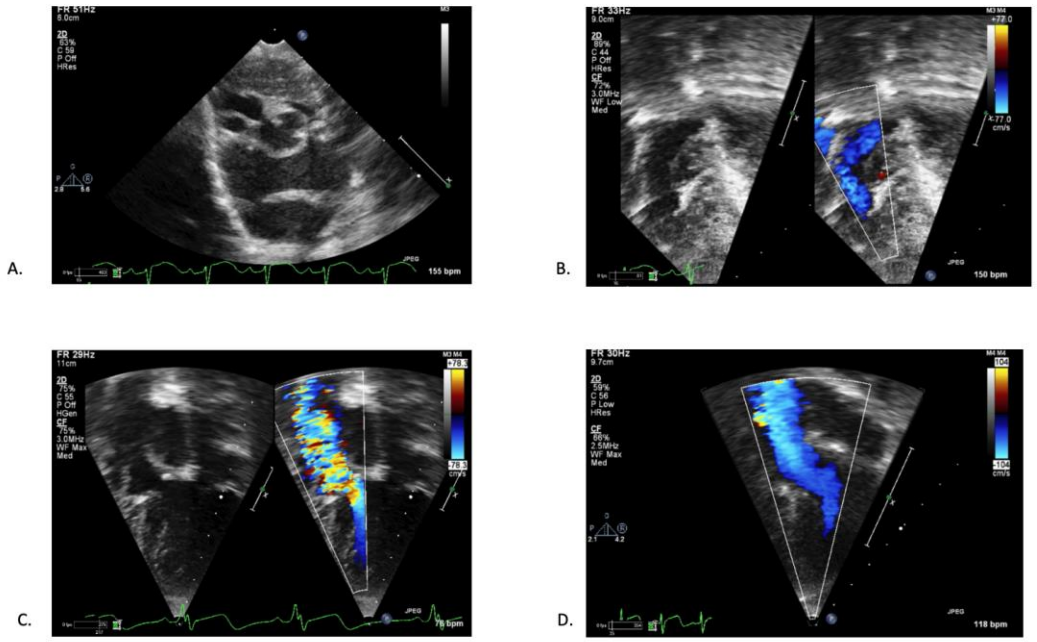


Figure 3: Echo images from a 5-day-old patient with Double Outlet Right Ventricle-Transposition of the Great Arteries type with juxtaposed atrial appendages. (A) Parasternal short axis view - posteriorly there is leftward juxtaposition of the right atrial appendage. (B) Apical 4 chamber view. No left ventricular outflow tract obstruction (LVOTO) noted at this time. (C) Images now at 2 years of age, post-operatively from Arterial Switch Operation. In Apical 4 chamber view, noted is LVOTO secondary to fibromuscular ridge at the crest of the muscular septum with a maximum instantaneous gradient of 75-80mmHg with a mean gradient of 35-40mmHg. (D) Apical 4 chamber view status post subaortic membrane resection with no residual gradient or turbulent flow seen across the left ventricular outflow tract.

Abstract #14: (FEL-013-OR)

Ness-Cochinwala M*; Kobaitri K; Sendi P; Martinez P; Totapally BR. ***"Alteplase Use in Pediatric Necrotizing Pneumonia"*.**

OR-Introduction

Necrotizing pneumonia is an uncommon but serious form of pneumonia in children that is often associated with empyema. The use of alteplase for pediatric necrotizing pneumonia is not well studied.

OR-Hypothesis/Study Objectives

This study's objective is to describe and evaluate treatment strategies for necrotizing pneumonia, specifically alteplase and antibiotic usage.

OR-Study Design, Methods, and Statistical Analysis

After IRB approval, the Pediatric Health Information System was queried for patients, ages 30 days to 19 years, with lung necrosis (ICD-10 code J85.0) from January 2016 to December 2019. Demographic characteristics, therapies utilized, and outcomes were compared in children who received alteplase with those who did not receive alteplase. Chi-squared, student's t-test, binary logistic regression and propensity matching were used for analyses. Children requiring alteplase were matched 1:1 using a correlative propensity score for age groups, gender, ethnicity, emergency department admission, presence of a complex chronic condition (CCC), mechanical ventilation, ECMO, anti-staphylococcal antibiotics used, pleural effusion and bacterial pneumonia and their outcomes were compared to controls.

OR-Main Results

A total of 2,177 patients with necrotizing pneumonia were analyzed. CCC was present in 1,119 (51.4%), 867 (39.8%) received alteplase and 98 (4.5%) underwent video assisted thoracoscopic surgery (VATS). The most common interventions and complications in children with necrotizing pneumonia are pleural effusion (59.1% of patients), intensive care unit admission (44.8% of patients), chest tube placement (42.1 of patients) and mechanical ventilation in 28.3% of patients. The mortality rate for pediatric necrotizing pneumonia is 3.9%. Both alteplase use (OR 3.86; CI 2.99-4.98) and VATS (OR 2.24; CI 1.4-3.6) were associated with an increased incidence of pneumothorax compared to patients who did not undergo these interventions in univariate analysis. The most commonly utilized classes of antibiotics were cephalosporins (82.6%) and anti-staphylococcal (Vancomycin, Linezolid and Ceftaroline) antibiotics (62.1%). On univariate analysis, alteplase was used more often for patients with a CCC and those who required mechanical ventilation or ECMO. Multivariable analysis showed that the top three risk factors associated with mortality are mechanical ventilation (OR 15.9; CI 7.1 – 35.8), ECMO (OR 7.5; CI 4.27 – 13.0) and presence of a CCC (OR 3.9; OR 1.7- 9.1). TPropensity score matched (442 pairs) analysis showed that alteplase use was not

associated with an increased risk of bronchopleural fistula or mortality but was associated with increased charges and longer length of stay.

OR-Conclusion and Significance

Alteplase is commonly used in pediatric necrotizing pneumonia. The rates of bronchopleural fistula and mortality are similar with or without use of alteplase. However, alteplase use is associated with increased charges and longer length of stay.

Abstract #15: (FEL-014-OR)

Ness-Cochinwala M*; Totapally BR. ***"Risk Factors for Pediatric Pulmonary Embolism: A National Database Study".***

OR-Introduction

Pediatric pulmonary embolism (PE) is a rare but potentially fatal event with a significant paucity of literature. The need for identification of risk factors for PE occurrence in hospitalized children is of paramount importance.

OR-Hypothesis/Study Objectives

OR-Study Design, Methods, and Statistical Analysis

A retrospective analysis of the Healthcare Cost and Utilization Project (HCUP) 2016 Kids Inpatient Database (KID) was performed. The database was filtered using ICD-10 code I26.0x and I26.9x for pulmonary embolism in children aged 28 days to 20 years. Complex analysis using chi squared and multivariable regression analysis were done to determine risk factors for developing pulmonary embolism. Sample weighting was employed to produce national estimates.

OR-Main Results

Of the 2,296,220 non-neonatal discharges, 3,172 patients (14/10,000 hospital discharges) had a PE with a mortality rate of 3.8%. Children over 12 years accounted for 89.8% of cases, with a prevalence of 24/10,000 hospital discharges as compared to 3/10,000 hospital discharges in those less than 12 years. Multivariable regression analysis showed that presence of liver disease (OR 1.57; 95% CI 1.18-1.95), history of DVT or PE (OR 3.54; 95% CI 2.96-4.24), mechanical ventilation (OR2.99;95% CI 2.54-3.50), central venous line (OR 1.45;95% CI 1.10-1.93), obesity (OR3.25;95% CI 2.93-3.61), hypercoagulable condition (OR 9.94; 95% CI 8.32-11.88), DVT (OR 82.89; 95% CI 74.08-92.75), sickle cell disease (OR 2.14; 95% CI 1.76-2.61), acute renal failure (OR 2.30; 95% CI 1.98-2.67), nephrotic syndrome (OR 2.42; 95% CI 1.82-3.22), congenital heart disease (OR 2.76; 95% CI 2.34-3.26), systemic lupus erythematous (OR 1.84; 95%

CI 1.42-2.37), malignancies (OR 2.59, 95% CI 2.30-2.92), and age >12 years (OR 8.47; 95% CI 7.40-9.69) were associated with an increased risk of developing pulmonary embolism. Systemic thrombolytic and catheter directed thrombolytic therapy were used in 160 patients and 35 patients respectively and neither had significantly increased risk of bleeding complications.

OR-Conclusion and Significance

This study provides a list of risk factors for developing pulmonary embolism in hospitalized children. The findings of this study can be used to select target population for PE-prophylaxis.

Abstract #16: (FEL-015-OR)**

Payson A*; Etinger V; Napky P; Montarroyos S; Ruiz-Castaneda A; Mestre M. ***"Risk of Serious Bacterial Infections in Febrile Infants With COVID-19".***

OR-Introduction

The diagnostic evaluation and management of young febrile infants continues to be a highly debated topic within the pediatric literature. Fever is a nonspecific symptom in this population that is frequently due to a benign viral process but is feared to be the initial presentation of serious bacterial infections (SBI) associated with high rates of morbidity and mortality. Reported rates of SBI in this population vary due to differences in age groups and definitions amongst studies, but generally range from 7.0-12.5% of low risk infants with high risk infants having a rate as high as 21%. Febrile infants with respiratory viral infections, such as Respiratory Syncytial Virus (RSV) and influenza, have significantly lower risks of SBI with rates of bacteremia of <2% and rates of urinary tract infections (UTI) ranging from 1.1-5.4%. Therefore, the presence of a concomitant viral infection can aid in the workup and management of these febrile infants.

Severe Acute Respiratory Syndrome Coronavirus 2 (SARS-CoV-2) is a novel single stranded RNA virus spread by respiratory droplets that originated in Wuhan, China and has caused a global pandemic of Coronavirus Disease 2019 (COVID-19). There have been case reports of COVID-19 infections in neonates and infants illustrating a spectrum of disease from asymptomatic positive cases to severe illness requiring endotracheal intubation and mechanical ventilation. Several case series have described the generally benign course of COVID-19 in febrile infants and observed a low rate of SBI in this population.

OR-Hypothesis/Study Objectives

The purposes of this study were to describe the clinical characteristics of febrile infants <90 days of age with SARSCoV-2 infections, to investigate the prevalence of SBI in these

infants, and to compare the risk of SBI in SARS-CoV-2 positive febrile infants with gender-and age-matched SARS-CoV-2 negative febrile infants.

OR-Study Design, Methods, and Statistical Analysis

This was a retrospective cohort study conducted from March to November 2020 in a tertiary children's hospital. Patients were identified by ICD-10 codes and included if age was <90 days, infant had fever >100.4F at home or in the Emergency Room, a SARS-CoV-2 PCR test was performed, and at least 1 bacterial culture was collected. Positive cases of SARS-CoV-2 were age- and gender-matched to negative controls for analysis. SBI was defined as a urinary tract infection (UTI), bacterial enteritis, bacteremia, and/or bacterial meningitis. We compared the demographics, clinical characteristics, laboratory results, and rates of SBI between infants with and without COVID-19. Controls consisted of infants who were gender- and age-matched (within 14 days of age) who presented with fever during the same time period but had a negative SARS-CoV-2 PCR test. Student's *t* tests were used to analyze continuous variables, Fisher exact tests were used to analyze categorical data, and the Wilcoxon rank-sum test for ordinal data. Relative risk ratios and 95% confidence intervals were calculated between the two populations of interest. All statistical tests were 2 tailed. Statistical significance was designated at a p-value of $\leq$ 0.05.

OR-Main Results

A total of 220 charts were identified based on ICD 10 codes and a total of 141 patients met the inclusion criteria. (Fig 1) 53 SARS-CoV-2 positive infants were age- and gender-matched to 52 SARS-CoV-2 negative infants. Both groups primarily identified with White race, Hispanic ethnicity, and had Medicaid insurance; less parents of infants with COVID-19 identified as primary English speakers (45%) than parents of controls (62%). There was no statistical difference in rates of admissions, median length of stay if admitted, or treatment with antibiotics between the two groups. (Table 1) Infants in the two groups had similar clinical presentations, but infants with COVID-19 identified with a higher rate of respiratory symptoms and lower white blood cell and C-reactive protein values than their SARS-CoV-2 matched controls. (Table 2) In the COVID-19 group, there were 4 cases of SBI (8%). There were 3 (6%) UTIs caused by *Escherichia coli* (*E. coli*), *Enterococcus faecalis*, and *Klebsiella aerogenes*, all occurring in males <60 days of age, and 1(2%) bacterial enteritis due to *Salmonella* species. There were no cases of bacteremia, bacterial meningitis, or additional respiratory infections in this group. In the control group, there were 18 cases of SBI (34%).

There were 12 (23%) UTIs, 4 (8%) cases of bacterial enteritis, and 2 (4%) cases of bacteremia. UTIs occurred in 8 males and 4 females; all 12 of the UTIs were caused by *E. coli.* 1 case of bacteremia was due to *Group B streptococcus* (GBS) in an 18 day old male born to a mother with unknown GBS status. The other bacteremia infection was due to *E. coli* in a 23 day old male with a negative urine culture who was born to a GBS positive

mother adequately treated with intrapartum antibiotics. All 4 cases of enteritis were due to *Salmonella* species.

There were no cases of meningitis. There was a significant difference in the rate of any SBI between the two groups (8% vs 34%) and the relative risk of any SBI between the two groups was 0.22 (95%CI: 0.1-0.6; p-value 0.001). (Table 3) There was also a significant difference in the rates of UTIs between the two groups (6% and 23%) with a RR of 0.25 (95% CI: 0.08-0.76; p-value 0.023). There was no significant difference in invasive bacterial infections (IBI) between the groups with only 2 cases of bacteremia found in the control group and no cases of meningitis in either group.

OR-Conclusion and Significance

To our knowledge, our study is the largest case series of COVID-19 infections in febrile infants <90 days of age and the first study to specifically investigate the rates of SBI in these infants compared to age-and gender-matched SARS-CoV-2 negative febrile infants. Although our overall rates of SBI were higher than previous studies addressing the risk of SBI in infants with respiratory viral infections, there remained a statistically significant difference in the rates of SBI between SARS-CoV-2 positive and negative febrile infants with a relative risk of 0.22. Similar to published literature, the most common infections in both groups were UTIs, and there was a statistical difference in UTIs between the two groups with a RR of 0.25. There were very few cases of IBI and no statistical difference between the two groups; the 2 cases of bacteremia occurred in <28 day old infants who are generally considered a high risk population, and frequently receive a complete evaluation and empiric antibiotic treatment solely based on age. These results suggest that febrile infants <90 days of age with COVID-19 have lower rates of SBI than their matched SARS-CoV-2 negative controls. This data is consistent with previous studies describing lower risks of SBI, particularly invasive bacterial infections, in febrile infants with concomitant viral respiratory tract infections. While larger studies on this population are needed, this study can aid in the diagnostic workup and treatment plan for young febrile infants with this novel virus.

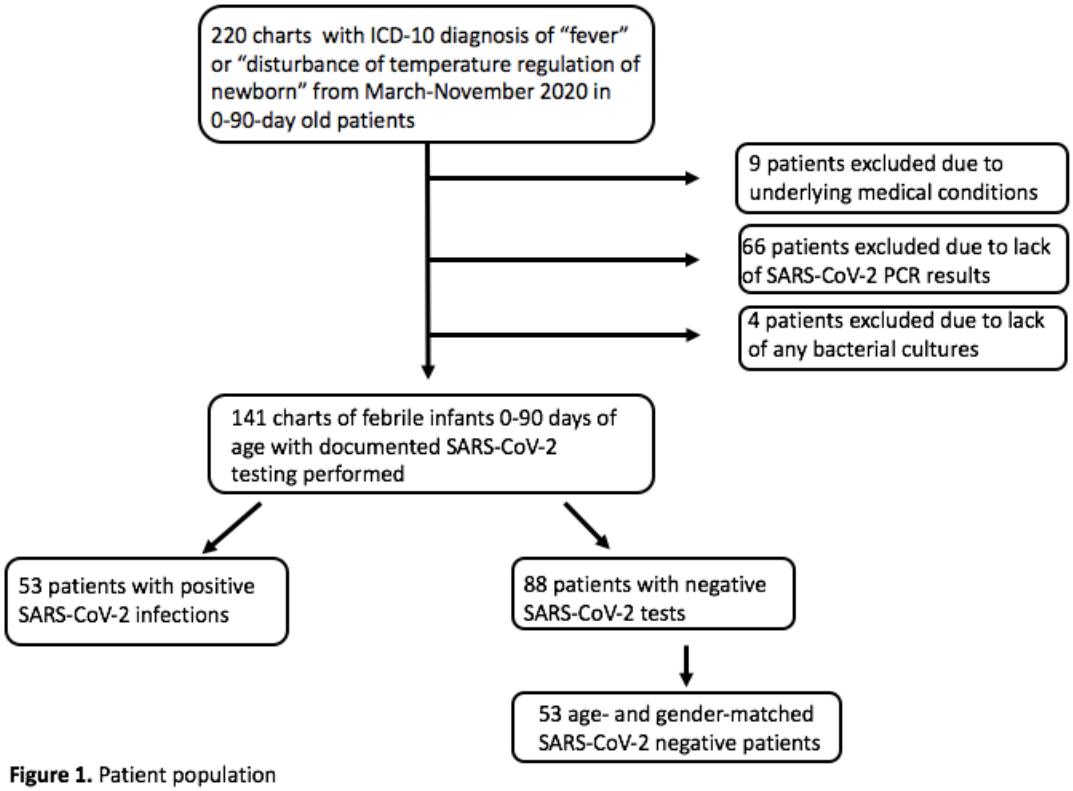


Figure 1. Patient population

Table 1. Patient Demographics According to COVID-19 Status

Variable	COVID-19 Positive (N=53)	COVID-19 Negative (N=53)	Risk Difference (95% CI)	p-value
Mean Age, days	46.5 ± 20.1	46.4 ± 21.6	0.15 (-8.2 to 7.9)	0.97
White Race	49 (92%)	45 (85%)	7.5% (-6.4 to 21.6)	0.36
Hispanic Ethnicity	47 (89%)	46 (87%)	1.9% (-12.5 to 16.3)	>0.99
English as Primary Language	24 (45%)	33 (62%)	-17.0% (-1.5 to 37.2)	0.12
Medicaid Insurance	43 (81%)	36 (68%)	13.2% (-4.7 to 30.2)	0.18
Chest Radiography Obtained	31 (58%)	11 (21%)	37.7% (17.9 to 53.9)	0.0001
Urine Culture Obtained	50 (94%)	52 (98%)	-3.8% (-7.3 to 14.2)	0.62
Blood Culture Obtained	48 (91%)	50 (94%)	-3.8% (-8.9 to 16.2)	0.72
CSF Obtained	19 (36%)	23 (43%)	-7.5% (-11.3 to 27.2)	0.55
Admitted to General Inpatient Hospital Unit	36 (68%)	33 (62%)	5.7% (-13.5 to 24.3)	0.68
Median Length of Stay, Hours	42.3 (29.5-62.1)	45.6 (38.5-66.5)	-3.3 (-15.6 to 4.2)	0.28
Patients Receiving Treatment with Antibiotics	25 (47%)	34 (64%)	17.0% (-1.4 to 37.1)	0.12

Values are mean ± SD, median (IQR), or number (%)

Table 3. SBI According to COVID-19 Status

Variable	COVID-19 Positive (N=53)	COVID-19 Negative (N=53)	Relative Risk (95% CI)	p-value
Any SBI	4 (8%)	18 (34%)	0.22 (0.08 to 0.57)	0.001
UTI	3 (6%)	12 (23%)	0.25 (0.08 to 0.76)	0.02
Enteritis	1 (2%)	4 (8%)	0.25 (0.04 to 1.6)	0.36
IBI (Bacteremia/Meningitis)	0	2 (4%)	0	0.5

Values are number (%)

Abstract #17: (FEL-016-OR)**

Urschel D*; Hernandez-Trujillo VP; Calderon J. ***"Panel of Unintended Consequences".***

OR-Introduction

Food allergy panels test a variety of food allergens at once. Indiscriminate food allergen testing is not sensitive. Inaccurate diagnosis of food allergy results in a variety of adverse consequences. The misapplication of these values as standardized criterion for food allergy diagnosis and management leads to both over- diagnosis of food allergy and management confusion of IgE mediated allergy.

OR-Hypothesis/Study Objectives

Our study compared unnecessary testing, unnecessary food avoidance, and epinephrine prescriptions between patients referred with food panel testing versus individual food testing. We hypothesized that compared to individual tested patients, food panel patients are more likely to have unnecessary testing, food avoidance, and less likely to have epinephrine prescription following an acute food reaction.

OR-Study Design, Methods, and Statistical Analysis

We conducted a retrospective review of 6675 patient's initial visits at Nicklaus Children's Hospital Allergy and Immunology Clinics from 2012-2020. Patients referred with food panel testing were compared to those with individualized testing prior to appointment. The number of patients with food panels via serum and/or skin testing, individual food testing or no testing, diagnosis, history of clinical food reactions, food avoidance history, and Epinephrine auto-injector prescriptions prior to appointment were documented. Patients with prior Allergist evaluation or insufficient encounter documentation were excluded.

OR-Main Results

5000 initial patient encounters were included. Food allergy panels comprised 961 (86.4%) of 1111 food allergy tests. Panels were more likely to be ordered than individual tests without a history of reacting to a food within two hours of ingestion (66% vs. 37%,

p<.001). The most common reasons for testing for food panels without history of acute reactions were rhinitis (45.4%), atopic dermatitis (18.6%), and urticaria (10.9%). Panel patients were more likely to be avoiding a food based on test results without a clinical reaction to a food than individual tested patients (46.5% vs. 25%, p<.001). Patients with a food reaction within two hours of ingestion were more likely to be prescribed epinephrine if they were tested to individual allergens rather than panels (62% vs. 43%, p=.001).

OR-Conclusion and Significance

Compared to individual tested patients, food panel patients are more likely to have unnecessary testing, food avoidance, and less likely to have epinephrine prescription following an acute food reaction. We suspect many food allergy panels were ordered for rhinitis and asthma because they also included inhalant allergen panels which would be more consistent triggers of these conditions. Given the large volume of patients undergoing unnecessary food testing when inhalant allergens alone would be more appropriate, our findings make a strong case for not automatically ordering these tests together. These results suggest a need for increased education regarding appropriate food allergy testing, interpretation, and management with ordering providers.

Abstract #74: (FEL-017-OR) §

Hassor S*; Payson A; Dantas M; Clemente M; Sanchez-Vegas C. **"*Identification of Pneumonia-Causing Pathogens using Karius Testing in Pediatric Patients*".**

OR-Introduction

Pneumonia is a leading cause of hospitalization in pediatric patients and can lead to significant morbidity and mortality if not appropriately treated. Historically, it has been difficult to identify pneumonia-causing pathogens via culture, leading to empiric antimicrobial use. A new diagnostic test known as, The Karius® Test, sequences microbial cell-free DNA in blood to identify over 1,000 pathogens including bacteria, viruses, fungi, and parasites, offering a less invasive method of identification. The clinical usefulness of this new test is still unknown and no guidelines are available to dictate when this test may be of value to patients hospitalized with pneumonia

OR-Hypothesis/Study Objectives

To investigate the association between pneumonia- causing pathogens, as identified by Karius testing, in pediatric patients hospitalized with pneumonia and their clinical course.

OR-Study Design, Methods, and Statistical Analysis

We conducted a retrospective, descriptive chart review at a tertiary care pediatric hospital of patients hospitalized with pneumonia in 2019, aged 0-21, who underwent Karius testing during hospitalization.

OR-Main Results

Of the 28 patients that were included, 54% (15) were male, 86% (24) were Caucasian, and 68%(19) were Hispanic, with an average age of 8.8 years (6 months-21 years). 89% of patients had significant co-morbidities, most commonly (14 patients) hematologic/oncologic diagnoses undergoing chemotherapy, bone marrow transplant, or on immunosuppression. 82% (23) of patients met SIRS criteria, 43% (12) required non-invasive ventilation, 7% (2) required invasive ventilation and 7% (2) patients required chest tubes. Karius testing was positive in 71% (20) and resulted in a change of management in 70% (14) of these positive cases including de-escalation of therapy in 6 cases and changing medications in 8 cases. Patients with a positive Karius had longer average lengths of stay (22 days) compared to negative tests (12 days) and longer total days of antibiotics per patient (26 days vs 17 days).

OR-Conclusion and Significance

The identification of a pneumonia-causing pathogen via Karius changed management in 70% of positive cases, indicating that initial empiric treatment was inadequate. Our patients are a small subset of those hospitalized with pneumonia, as most do not undergo Karius testing. Therefore, this study is skewed towards those meeting SIRS criteria and those immunosuppressed, who frequently require broad-
spectrum coverage. While clinically useful, the Karius is an expensive test that should be reserved for those not improving on traditional therapies or at risk for uncommon pathogens.

III- NURSING ABSTRACTS:

Abstract #18: (NUR-017-OR)

Bandin A*; Roberts K; Valiente A. ***"Pedaling for Air: A Pediatric Inpatient Approach to Enhance Lung Function".***

OR-Introduction

The purpose of this project was to demonstrate how inclusion of aerobic activities in a plan of care improves overall patient outcomes in Cystic Fibrosis (CF) hospitalization. Literature supports that the increase of aerobic stimulation in a patient with CF improves lung quality and function (O'Connor, 2017). However, CF population tends to have about a 2 week sedentary inpatient length of stay due to the complexity of their treatments and infection prevention strategies. These circumstances have empowered nursing to get

creative in their care. To address this gap, clinical nurses embedded portable physical pedal exercise into the plan of care to increase aerobic stimulation.
As key care providers, nurses are enabled to encourage CF patients to take an active role in the management of their illness. The implementation of pedal therapy for these patients boosts lung function. Guidelines are set to ensure safety and sustainability. Providing these patients with aerobic stimulation in the safety of their own hospital room, enhances their impatient treatment while positively impacting physical and emotional state. This improves overall lung function and promotes an active lifestyle outside the hospital.

OR-Hypothesis/Study Objectives

The objective of this project was to evaluate the efficacy and impact in lung function of the addition of aerobic exercise to the daily plan of care of pediatric Cystic Fibrosis patients.

OR-Study Design, Methods, and Statistical Analysis

An assessment of the availability for CF patients to participate in aerobic exercise during hospitalization was conducted. Evidence showed barriers and limited access to methods of physical activity. In collaboration with pulmonology and respiratory therapists, unit nursing leaders and clinical nurses created a guideline for portable pedal exerciser use. The pedal exerciser is easily transported to the patient's room, meets falls precaution, and infection control guidelines.
By way of a provider communication order, pedal therapy was initiated. Clinical nurse then educated the CF patient and caregivers on the opportunity for aerobic exercise, encouraging them to participate for a minimum of 20 minutes daily post airway clearance therapy. Pre- and post-exercise patients' tolerance is noted and exercises are modified as needed.

OR-Main Results

Study was conducted from January-December 2019 due to inpatient availability. For patient 1, pre implementation force expiratory volume (FEV1) increased by 31%. Implementation month demonstrated a 25% increase in FEV1s. Post implementation FEV1s improved 63.3% and 78.57% on 2 different admissions. For patient 2, pre implementation FEV1s increased by 12.6%, post implementation 27.1%

OR-Conclusion and Significance

Aerobic activities plays a key role in the enhancement of lung function. The addition of aerobic stimulation to the plan of care for CF population has shown to make a positive impact in the 2 patients evaluated as evidence of the FEV1s improved by 31% and 12.6%

respectively. Implementing this pedal therapy with a portable pedal exerciser created an opportunity for nurses, patients and caregivers to team up on creative ways for additional interventions within the barriers of a hospital stay. This enhanced patient care and maximized their inpatient stay and improved their lung function all while influencing future self-care routines to improve their quality of life, and promoting an active lifestyle.

Abstract #19: (NUR-018-OR)

Burke ML*; and Taylor-Amador S. ***"Parent Education Discharge Support Strategies".***

OR-Introduction

One of the most significant life changing events is a major medical illness, such as the diagnosis of cancer. A new diagnosis of a childhood cancer affects the patient and the entire extended family, including parents, grandparents, siblings, classmates, and friends. The initial phases of the cancer journey can be overwhelming for families. Parents are expected to learn about side effects, treatment regimens, and when to seek medical care This study evaluated an effectiveness of two different early – discharge educational tools to improve the parent's ability to care for their child during the transition from hospital to home.

OR-Hypothesis/Study Objectives

Objectives:

1. Explore the effects of the Parent Education Discharge Support Strategies (PEDSS) on childhood cancer symptoms and parents' perception of their ability to care for their child.
2. Determine if PEDSS decreases unplanned utilization of healthcare services and preventable toxicity among children with cancer.
3. Examine the feasibility and fidelity of implementing PEDSS at the initial hospital discharge among parents of newly diagnosed children with cancer.

OR-Study Design, Methods, and Statistical Analysis

The design of the study was a cluster randomized control trial involving 16 Magnet recognized hospitals with pediatric oncology programs. The interventions were a symptom management educational intervention and a parent support and coping intervention. Nicklaus Children's Hospital was assigned the support and coping intervention. Parents of newly diagnosed patients were recruited on the inpatient unit during the initial admission in which the diagnosis was cancer. Parents would receive the traditional discharge education and if they agreed and consented to the study the additional PEDSS intervention would be provided. The surveys were completed at 3 time

points: 1) initial diagnosis before discharge, 2) at 1 month and 3) at 2 months. There was a total of 283 subjects and statically analysis revealed that a standardized approach to discharge education for this patient population is successful across all institutions participating.

OR-Main Results

Results concluded that the symptom management group had a better outcome in pain control compared to the psychosocial group. In both groups symptoms of nausea and appetite alterations were experienced equally. Also, fatigue and sleep alterations decreased over time for both study groups. The most significant finding is that the standardization of the education was significant for parents that reported feeling confident in their ability to care for their child with cancer after hospital discharge.

OR-Conclusion and Significance

PEDSS INTERVENTION

Support for Parents and Caregivers

pedss

When a child is diagnosed with cancer, families deal with many new things. This worksheet may help you manage events and care for yourself during this time. If you have concerns, please let us know and we can contact professionals to help you.

Topic...	Other families found it helpful to...
Expect changes	
Daily life often changes after a cancer diagnosis. These changes may include time away from work and changes in schedules for your other children.	Talk to your other children about changes Talk to a social worker about leave of absence from work and other available resources Ask relatives and friends to help you
Help from the cancer team	
Common responses to a cancer diagnosis may include feeling upset and worried. This is natural. Most people find it helpful to share their feelings. Experts can teach new skills, such as relaxation and stress management.	Write out your questions for the doctor or nurses Talk about your concerns with nurses, doctors, social workers, chaplains, child life specialists, psychologists, and/or psychiatrists Consider attending parent support groups
Take care of yourself	
Parents and caregivers often handle their feelings better when they take care of their own needs. Taking care of yourself also helps you care for your child. Keeping up with routines and normal activities reassures your child and give your child a sense of safety.	Consider some form of exercise, such as walking Eat three meals every day Continue to take your medications See your doctor(s) as indicated Participate in relaxing activities, such as meditation
Involve your child	
Communicating with your child is important to maintain an open and honest relationship. In general, secrets tend to shut down communication.	Explain the diagnosis and treatment in words your child can understand Encourage your child to share his/her feelings by talking, writing, or playing Answer questions when your child is asking for information
Involve others	
Do not keep information from family members. The health care team can help you share news in a way that is sensitive to any special issues of a family member. Good communication and working together are especially important at this time.	When you are ready, share information with others Encourage family members to express their thoughts and emotions Ask for help

pedss

Parent education discharge support strategies

Abstract #20: (NUR-019-OR)**

Bandin A; Canizares A; Dieguez R; Klareich J*; Mirabal C. ***"There's No Place like Home: A Standardized Discharge Plan to Reduce Hospital Readmission Rates".***

OR-Introduction

The purpose of this evidence-based project was to identify causes of patient readmissions, and implement interventions to help decrease patients returning to the hospital. Readmission is defined as an unplanned return to the hospital for any cause within 30 days. The top 5 diagnosis identified for readmission at Nicklaus Children's Hospital are epilepsy, sepsis, dehydration, major depressive disorders, and pneumonia. Prior to this project, there was no standardized process to discharge planning, thereby creating challenges at time of discharge, and leading to higher risk of potential readmissions.

OR-Hypothesis/Study Objectives

The objectives of this project was to define readmissions to the hospital, state the reasons for preventable and non-preventable readmission, and discuss standardized interventions utilized to decreased readmission rates.

OR-Study Design, Methods, and Statistical Analysis

An interprofessional readmission taskforce was created to focus on decreasing readmission rates on 5 Tower and 3 Northeast, 2 of the highest units for readmissions. A comprehensive literature review was conducted with evidence showing a standardized discharge plan will reduce readmission rates. The biggest challenge identified in the taskforce, was communication among all providers. To help with communication, an interprofessional huddle was standardized on 5 Tower and 3 Northeast each morning with key stakeholders to discuss each patient and the plans for the day. Discharge planning is the focus and identifying anything that can be accomplished prior to rounding is to be completed. This taskforce also identified the need for a common area within the electronic medical record (EMR) for all discharge needs. The taskforce collaborated with nursing informatics to develop a discharge landing page for all providers to see where the patient is in the discharge process. Prior to this landing page, the healthcare team member would have to navigate through multiple areas of the chart to identify what was needed for discharge. By using the landing page, it created a one stop shop for all providers to assess what has been completed, and what is still pending for discharge. Along with the landing page, a written checklist was also utilized on each patient to ensure nothing was missing from the landing page and all needs were being met. Another topic identified was the importance of using teach-back on all patients and families to ensure discharge instructions were understood. All disciplines were educated on how to use teach-back through videos and role playing. Lastly, when a discharge was known, a “time out for discharge” was implemented and conducted with all healthcare providers. A few moments of pausing and focusing on all items related to discharge ensured everything was finalized before initiating the discharge for a patient. Interprofessional collaboration included identifying whether all patients received case management initial assessment, social work pre discharge assessments, nursing education, teaching of medication, disease process for home, physician discharge medication reconciliation as well as follow-up medical and post discharge tests/labs appointments prior to discharge.

A pilot study was completed from October 2020-December 2020 utilizing these standardized interventions to reduce readmission rates. Chart reviews were completed on all patients that were readmitted to find out the specific reason in order to identify other interventions needed.

OR-Main Results

From October 2020 through January 2021, 5 Tower had a downward trend in readmissions consistently. The percentage ranged from 9.4-7.5% of readmissions occurred on 5 Tower during the pilot phase. After the pilot phase, a downward was sustained. 3 Northeast also had a downward trend in percentage of patients readmitted. The percentage ranged from 11.9-6.9% of patients readmitted within 30 days. Ninety-three patients returned to the hospital within 30 days to 5 Tower or 3 Northeast during the pilot. Of the 93 readmissions, 81% were concluded to be non-preventable, while the other 19% could have been prevented.

OR-Conclusion and Significance

Standardized discharge planning proved to reduce readmissions by utilizing the standardized interventions implemented. An interprofessional approach is key to successfully reducing preventable readmissions. Future implications include electronic integration of the written checklist into the electronic medical record through rounding documentation and expand this standardized process to all inpatient nursing units. Another conclusion and future implication is the importance of collaborating with all specialties and community care coordinators to ensure a safe home transition and resources for home care.

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CASE REPORTS

I- RESIDENTS ABSTRACTS:

Abstract #21: (RES-001-CR)

Aquino Crisanto L*; Napky Raudales PJ; Girgis S; Jayakar P. ***"Complex combined Trisomy 14 and 21 Mosaicism and Multisystem Involvement in a Newborn"*.**

CR-Introduction/ Background

Trisomy 14 mosaicism is a rare chromosomal anomaly of variable phenotype with more than 50 cases reported to date. The frequency of trisomy 21 mosaicism has been estimated to range from 1 in 16,670 to 1 in 41,670 livebirths. However, cases reported with both trisomies 14 and 21 are not well documented. Growth delay, dysmorphic craniofacial features, congenital heart disease (CHD) and genitourinary (GU) abnormalities are features of both mosaic trisomy 14 and 21. We report a case of complex mosaicism with both trisomy 14 and trisomy 21 presenting with multiple congenital anomalies.

CR-Case(s) Presentation(s)

Patient was born full-term with normal anthropometric measurements aside from macrocephaly (Birth weight 3480g, length 48cm, head circumference 37.5cm) to a 32-year-old mother followed prenatally for suspected CHD. Pregnancy and delivery were uneventful. Parents denied consanguinity. Following birth, physical exam was notable for frontal bossing, bilateral temporal narrowing, low-set folded ears, small palpebral fissures, left iris coloboma, retrognathia, high-arched palate with soft palate cleft, short neck, heart murmur, ambiguous genitalia, sacral dimple with hair tuft and hypotonia. Abdominal/pelvic imaging detected undescended gonads with no Müllerian structures and enlarged bilateral adrenal glands. Endocrine workup was compatible with hypogonadotropic hypogonadism. Neuroimaging showed bilateral choroid plexus cysts and a low conus medullaris at L4. Echocardiography confirmed unbalanced right ventricle dominant common atrioventricular canal defect with left atrioventricular valve atresia and double outlet right ventricle with pulmonary stenosis, partial anomalous pulmonary venous return and right aortic arch with patent ductus arteriosus. Karyotype analysis revealed an abnormal mosaic complex with 3 cell lines: 47,XY,+14[12]/48,XY,+14,+21[6]/46,XY[27].

CR-Diagnosis and Discussion

Unlike patients with both mosaicism trisomy 14 and 21, our patient lacked growth retardation and had macrocephaly. He had no skin pigmentation anomalies which is characteristic of patient with mosaic trisomy 14. Our patient had abnormal craniofacial

features (low-set ears, small palpebral fissures, retrognathia, and short neck) and ambiguous genitalia, which are characteristics of both trisomy 14 and 21. Ambiguous genitalia are more commonly a consequence of primary hypogonadism in trisomy 21, as opposed to our case, which was due to secondary hypogonadism. Eye abnormalities have been described in both trisomies, with iris coloboma being reported in patients with trisomy 14. Common CHD described in mosaic trisomy 14 include atrial and ventricular septal defects, and patent ductus arteriosus. Aorto-pulmonary window has been recently reported as an associated lesion. Remarkably, our patient had a complex congenital cardiac lesion predominated by unbalanced common atrioventricular canal defect. Atrioventricular canal defects is the main cardiac abnormality occurring in 40% of patient with trisomy 21, which could explain the cardiac lesion in our patient.
Trisomy 14 mosaicism resulting from trisomy 14, translocations, uniparental disomy and isochromosomes has been reported in the literature. Trisomy 14 and trisomy 21 has been reported associated with non-Down syndrome acute megakaryoblastic leukemia.. This is a case of trisomy 14 mosaicism due to the coexistence of 2 abnormal cell lines, one with trisomy 14 and another with trisomy of both chromosomes 14 and 21. The variability in the severity of clinical characteristics could be explain by trisomy rescue. Our case is unique with clinical phenotype suggestive of both mosaic trisomies 14 and 21.

CR-Conclusion and Significance

- Trisomy 14 and trisomy 21 has been previously reported associated with non-Down syndrome acute megakaryoblastic leukemia cells.
- This is a case of trisomy 14 mosaicism due to the coexistence of 2 abnormal cell lines, one with trisomy 14 and another with trisomy of both chromosomes 14 and 21.
- Our case has unique clinical features suggestive of both mosaic trisomies 14 and 21.
- Despite normal birth anthropometrics (except for macrocephaly), at 3 weeks of age, weight and length fell off the growth curve
- Trisomy rescue could explain the variable phenotype.

Abstract #22: (RES-002-CR)

Cavalcante RC*; Dantas MA; Perez-Burnes L. ***"Go Where The Blood Is"*****.**

CR-Introduction/ Background

Salmonella is a gram-negative bacilli that most commonly causes gastroenteritis and enteric fever, as well as meningitis on infants, and osteomyelitis on asplenic patients. However, another extraintestinal manifestation of Salmonella is urinary tract involvement causing cystitis or pyelonephritis. We report case of a 7-year-old female with subacute generalized abdominal pain, and fever, associated with hematuria.

CR-Case(s) Presentation(s)

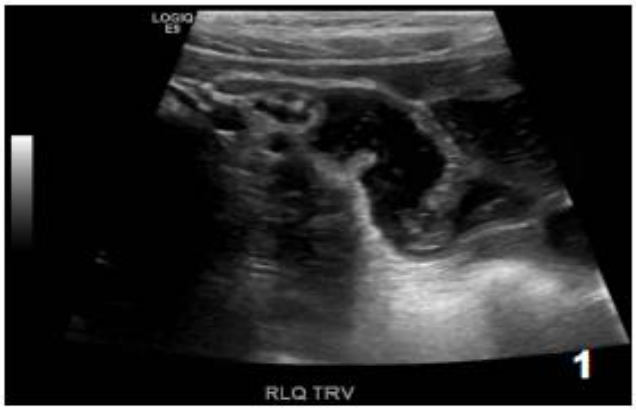
7-year-old previously healthy female presented to the ED with a 2-week history of intermittent colic-like abdominal pain, moderate, initially described as diffuse, non-radiating, associated with constipation, and resolved after a bowel movement. She also had a 5-day history of fever, rhinorrhea, and sore throat. Initial diagnosis was a viral syndrome. Returned in the following day due to worsening of abdominal pain, now localized to right lower quadrant (RLQ) and hypogastrium. She had one pet dog, no history of recent travel, or ingestion of uncooked foods. Abdomen was nontender to palpation on the first presentation, but had RLQ and hypogastrium tenderness on the second. Initial laboratory studies showed 6% atypical lymphocytosis, microscopic hematuria, and negative tests for influenza, monospot, and rapid strep. Appendix US with normal appendix and fluid-filled hyperperistaltic bowel loops in the RLQ suggesting enteritis/colitis (image 1). On the second day of admission, patient presented an episode of macroscopic hematuria. Renal-bladder US showed horseshoe kidney and bladder debris with wall thickening (Images 2 and 3). Further results were negative for adenovirus PCR and positive for gram-negative rods in urine culture. Blood culture was negative. Ceftriaxone was started empirically. Urine culture was then confirmed to be Salmonella sp. Given that Salmonella was positive in the urine and initial presentation of RLQ pain with description of enteritis/colitis, stool culture and GI panel were collected, both returning positive for Salmonella species.

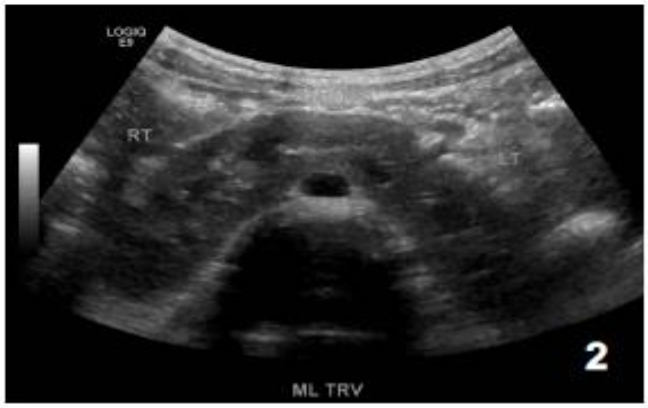
CR-Diagnosis and Discussion

Microscopic hematuria prevalence ranges from 0.5-2% in pediatric population. The most common causes of transient microscopic hematuria in children are fever, exercise, UTI, and trauma. When persistent, the most common causes of hematuria are glomerulonephritis, hypercalciuria, and nutcracker syndrome. During hospitalization, our patient presented macroscopic hematuria, which is found to be secondary to UTI in about half of the patients. Reported causes of infectious gross hematuria include Schistosoma haematobium, Mycobacterium tuberculosis and Adenovirus. Salmonella is a rare etiology for UTI and is almost always associated with abnormalities in the urinary tract. One case report described a Salmonella gastroenteritis complicated by acute interstitial nephritis, which presented with hematochezia and gross hematuria. However, there was no other report with findings of simultaneous positive culture and radiological findings suggestive of acute infection on both urinary and gastrointestinal tract.

CR-Conclusion and Significance

Salmonella, as well as Yersinia enterocololitica, and Campylobacter, are causes of fever with acute/subacute generalized abdominal pain that migrates to the RLQ, with or without diarrhea, mimicking acute appendicitis. In the light of the exposed case, urinary tract infection should be remembered as an extramanifestation of Salmonella.





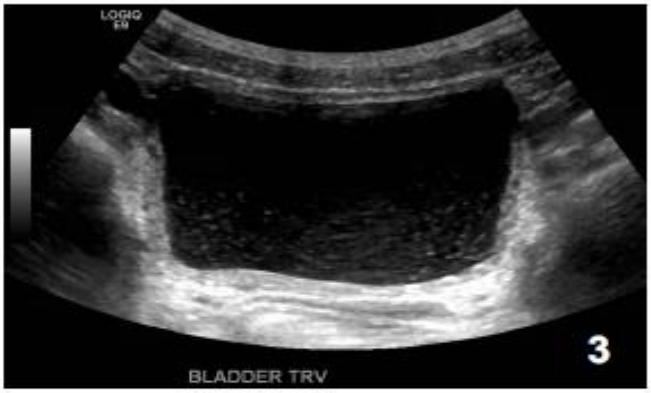


Image 1– Appendix ultrasound showing fluid-filled hyperperistaltic bowel loops in the right lower quadrant suggesting enteritis/colitis; **Image 2** – Renal bladder ultrasound showing horseshoe kidney; **Image 3** – Renal bladder ultrasound showing bladder debris with wall thickening suggesting cystitis;

Abstract #23: (RES-003-CR)

Colón Guzmán S*; Roberts A; Sunny J; Bujarska M; Gonzalez-Vallina R. ***"A Forgotten Diagnosis: A Case About Anterior Cutaneous Nerve Entrapment Syndrome".***

CR-Introduction/ Background

Chronic abdominal pain is a common finding in the pediatric population. When abdominal pain is severe and localized, entrapment of the cutaneous branches of the thoracoabdominal intercostal nerves in the rectus abdominis muscle should be considered. Anterior Cutaneous Nerve Entrapment Syndrome (ACNES) has a very particular presentation. The pain is often localized to a small area of the abdominal wall, less than 2cm in diameter, and characteristically in the right side of the rectus abdominis muscle. Characteristically, and unlike visceral abdominal pain, the discomfort is exacerbated by tension of the abdominal muscles (Carnette's sign). Failure to differentiate visceral from abdominal wall pain leads to extensive and invasive work-up and subjects patients to the stigma of chronic abdominal pain without a source. ACNES can be easily diagnosed and treated with local injection of an anesthetic and should be present in clinician's differential diagnoses when presented with localized abdominal pain. We present a case of a 13-year-old female with a characteristic presentation of ACNES that, despite delay in diagnosis, had complete resolution of symptoms with the first-line treatment.

CR-Case(s) Presentation(s)

A 13-year-old female initially presented to the ED with right lower quadrant abdominal pain that was sharp, constant, worsened by activity and coughing, and relieved by rest. Pain had been present for more than 8 months. She had been seen at outside hospital four days prior where ultrasound and CT had ruled out appendicitis. On arrival to the ED, ultrasound and CT were repeated and again ruled out appendicitis or ovarian abnormalities. CBC, CRP, and urinalysis were within normal limits for age. She was referred to Gastroenterology who recommended MR enterography which was normal. Based on the characteristics of the pain, she was diagnosed with abdominal wall pain, likely from entrapment syndrome. Anesthesiology became involved and recommended a Lidocaine 5% patch, which temporarily relieved pain. However, pain returned, and decision was made to treat with abdominal nerve block by Anesthesiology. Injection of Bupivacaine and Methylprednisolone to area of maximum pain relieved discomfort completely. On follow up one month after procedure, patient remained symptom free.

CR-Diagnosis and Discussion

Patients with ACNES often present with chronic, debilitating, abdominal pain and delayed diagnosis. The absence of radiographic or laboratory evidence of this type of chronic abdominal pain may cause patients to be misdiagnosed with disorders such as Functional Abdominal Pain, delaying appropriate treatment. Patients may also be submitted to unnecessary tests and polypharmacy in their search for relief. We present this case with the goal of describing a classic case of ACNES, with positive Carnett's sign, and which responded to conservative management.

CR-Conclusion and Significance

Recognizing a different presentation of right lower quadrant pain can save patients from years of abdominal pain and discomfort, can reduce health care costs and length of stays, and thus improving quality of life of a vulnerable patient population.

Abstract #24: (RES-004-CR)

Colon Guzman S*; Prieto P; Gamboa H. ***"Acute Management of a Suspected Case of Cannabis Induced Cyclic Vomiting Syndrome".***

CR-Introduction/ Background

Cannabis induced CVS, currently diagnosed with Rome IV criteria, is a disorder of disabling and stereotypical nausea, vomiting, and abdominal pain associated to prolonged and excessive use of cannabis. As this is a new diagnosis by Rome IV and because of poor follow up of patients and stigma associated to cannabis use, patients with this diagnosis frequently are undertreated, most times not even getting the minimum treatment of CVS. We present a case of a 17-year-old male with suspected Cannabis Induced CVS and propose a standard protocol that can be implemented in patients with similar presentation.

CR-Case(s) Presentation(s)

17 years old male with depression transferred from an outside hospital for acute worsening of abdominal pain, intermittent emesis, and weight loss. Previous work up included ultrasound of the appendix and CT of abdomen, both of which were normal. Cell count, electrolytes, liver enzymes, and lipase also normal for age. Due to severe epigastric pain unresponsive to H2 blockers, he was transferred for further management. During admission, labs were remarkable for urinalysis with 80 ketones, negative H. pylori antigen in stool, negative COVID19, normal thyroid function, negative Celiac panel, normal CRP, ESR, and stool calprotectin, and positive urine toxicology for cannabis. He was initially treated with parenteral re-hydration, PPI, Ondansetron, and Carafate. After 4 days of no improvement of symptoms noted, GI service was consulted. EGD was done which showed mild distal esophagitis and gastritis. Due to history of cannabis use >2 times per week for great than 1 year he was diagnosed with presumed Cannabis induced Cyclic vomiting syndrome and started on CVS protocol of D10, Ketorolac for pain, Lorazepam around the clock for 24 hours, and Cyproheptadine at night. Nausea and vomiting resolved within 24 hours; abdominal pain resolved within 48 hours. Behavioral medicine and Nutrition were also involved for optimization of his depression and nutrition. He began tolerating his diet and was discharged home on Cyproheptadine for prophylaxis. Since his discharge he has done well with no further symptoms. He continues on Cyproheptadine, ceased use of cannabis, and follows with Gastroenterology.

CR-Diagnosis and Discussion

CVS is a debilitating disease characterized by recurrent bouts of nausea, vomiting, and abdominal pain. When associated to heavy cannabis use, patients face an additional barrier of stigma which may cause them to receive sub-optimal treatment, especially when no standard protocol currently exists. The 2008 NAPGHAN Consensus statement gives recommendations for recognition and treatment of CVS. Treatment is divided into the acute or episodic phase and the well phase between episodes. Treatment during acute attack includes supplemental carbohydrate for increased energy demand such as IV dextrose. Cyproheptadine as an antihistamine and serotonin receptor antagonist has been identified as the preferred choice in children for abortive treatment as well as prophylaxis between episodes. Early intervention of pain control with NSAIDs is also recommended. Ondansetron in higher doses is beneficial for abortive treatment of nausea and vomiting and when anti-emetics alone don't control nausea and vomiting, combination of ondansetron and lorazepam has been described as most effective option. We propose a standard protocol when patients meet criteria of CVS associated to cannabis use in order to decrease severity of symptoms and length of stay in the hospital. As seen with this patient, when the appropriate protocol was implemented, patient demonstrated improvement of symptoms in less than 24 hours and complete resolution in 48 hours.

CR-Conclusion and Significance

For cannabis induced CVS, a standard treatment protocol does not exist, making management decisions subjective and varied among medical professionals and causing delays in appropriate treatment. The case presented demonstrates a first-time episode of nausea, emesis, and abdominal pain, not meeting the time criteria for CVS. However, we present this case as an example, due to the extensive work-up done to rule out other organic causes of his presentation. Symptoms were observed to significantly improve with CVS protocol and the patient has not had recurrence after cessation of cannabis use and treatment with Cyproheptadine. We propose that patients presenting with suspected cannabis induced CVS be treated with a CVS protocol as described in this case report.

Abstract #25: (RES-005-CR)

Dantas M*; Cavalcante R; Hassor-Rivero S; Clemente M. ***"An Uncommon Cause of Fever of Unknown Origin".***

CR-Introduction/ Background

Fever is one of the most common pediatric presentations. Fever of unknown origin (FUO) refers to a temperature higher than 38.3oC at least once per day for equal or more than 8 days. FUO is a clinical challenge and impose a high level of stress to families.

CR-Case(s) Presentation(s)

The patient was a 6-year-old male who presented with a 21-day history of fever. Patient had emesis and URI symptoms that resolved but continued to present daily intermittent fevers and decreased activity level with no other localizing symptoms. There was no

history of travel outside of the country and no exposure to animals. Immunizations were up to date. Patient underwent extensive workup with no positive findings and was discharged home for outpatient follow-up. One week later, he was re- admitted with self-limited urinary retention, and discharged after negative workup. Two weeks later, patient presented with loss of balance and somnolence.

On 1st admission, physical exam showed an innocent murmur. On 2nd admission, patient had patellar hyperreflexia and lower extremity weakness that were associated with ataxic gait on 3rd admission.

Initial labs showed WBC 16,2K/uL, CRP 1.8mg/dL, ESR 37 mm/ hr, negative UA, CXR, and CMP. EBV panel was positive for recent infection. CMV PCR, QuantiFERON, Mycoplasma PCR, Bartonella titers, RPP, Influenza PCR, Karius, urine and blood cultures were negative. ASO, C3 and C4 levels, rheumatoid factor, lupus panel, LDH, uric acid, ferritin and CT of abdomen and chest, ECHO and EKG were also negative. On 2nd admission, a spine MRI and an RBUS were done with no abnormalities. On 3rd admission, brain and spine MRI showed multiple areas of demyelination (images 1- 3). Lumbar puncture showed 12 WBC (78% lymphocytes), 27 RBCs, normal protein and glucose levels. Meningitis encephalitis panel, auto-immune encephalitis panel, CSF IgG Index, oligoclonal bands, MOG-Ab, anti-aquaporin 4 Ab were negative. Patient was started on IV steroids with improvement.

CR-Diagnosis and Discussion

Given the presence of first polyfocal CNS findings, encephalopathy manifested by persistent somnolence and abnormal brain MRI during acute phase, patient's clinical picture is consistent with Acute Demyelinating Encephalomyelitis.

ADEM is not frequently considered as an FUO etiology since clinicians expect neurologic deficits to develop earlier than in the case presented. However, atypical ADEM presentations should be considered, especially when patients present significant sleepiness, which may be an early sign of encephalopathy. Furthermore, abnormalities were not identified on the initial spine MRI on our patient, something previously reported by literature and that physicians should be aware of the possibility. Imaging and clinical follow-up is required to classify ADEM subtypes and rule-out other demyelinating syndromes such as Multiple Sclerosis.

CR-Conclusion and Significance

Physicians should be aware of atypical presentation of ADEM and consider the condition in the differential diagnosis of FUO.

Abstract #26: (RES-006-CR)

Diaz M*; Torres D; Murphy K; Pulido A. ***"A Lump In My Chest, Will You Be Able To Guess?"*****.**

CR-Introduction/ Background

Osteomyelitis is a relatively common infection in children that predominantly affects the metaphysis of the long bones. Osteomyelitis in the chest wall, however, is a rare occurrence in the pediatric population, with only 58 cases of rib osteomyelitis reported in the literature. Most cases of rib osteomyelitis result from penetrating trauma, spread from adjacent tissue, or hematogenous seeding.

CR-Case(s) Presentation(s)

A 10 month old, previously healthy, fully vaccinated, boy presented to the ED with a 10 day history of daily fevers (Tmax 103.3 F) and left anterolateral chest swelling. Associated symptoms included rhinorrhea, cough, mild decrease in activity level, decreased oral intake and urinary output. No history of trauma. On arrival, the patient was febrile and tachycardic. On physical exam, a firm tender mass (measuring approximately 5 cm x 3 cm) was felt on the left anterior lower chest, with no overlying erythema or skin changes. Initial work up showed a white blood cell count of 24.4 10K/µL with a low hemoglobin of 9.6 gm/dL and elevated platelets of 532 10K/µL. C-reactive protein was elevated at 5.6 mg/dL. Complete metabolic panel, lactate dehydrogenase, and uric acid were within normal limits. Rapid influenza test was negative. Chest X- ray was negative except for soft tissue mass, and chest ultrasound revealed a complex solid, avascular oval anterolateral left chest wall soft tissue mass. The patient was started empirically on clindamycin and piperacillin/tazobactam. Characterization of the mass with computed tomography (CT) later showed a complex cystic mass with minimal nodular peripheral enhancement, associated soft tissue swelling extending from the lower chest down the flank to the level of the spleen. The 11th rib was engulfed and had mild destructive changes. These findings were consistent with osteomyelitis. The patient was taken to the operating room for surgical exploration and drainage. Cultures grew methicillin-sensitive Staphylococcus aureus (MSSA). Thus antibiotics were switched to cefazolin. Allergy and Immunology was consulted to rule out an immunodeficiency and work up was found within normal limits. The patient improved clinically and was discharged with a peripherally inserted central catheter line to complete 28 days of cefazolin.

CR-Diagnosis and Discussion

Rib osteomyelitis is uncommon in children, and in this case the source of the infection was not clear and was presumed to be secondary to hematogenous spread. With dissemination, the most common sites are near the costochondral junction and the costovertebral angle with the most common pathogen being Staphylococcus aureus.

CR-Conclusion and Significance

Rib osteomyelitis is a rare finding in the pediatric population, therefore it is important to have a high index of suspicion in patients presenting with fever and tender chest wall swelling in order to provide appropriate treatment.

Abstract #27: (RES-007-CR)

Fallatah E*; Chang Y; Calderon JG; Hernandez-Trujillo VP. ***"DiGeorge Syndrome and COVID-19 in Two Pediatric Patients".***

CR-Introduction/ Background

Immunocompromised patients, including those with primary immunodeficiency, may be expected to have more severe COVID-19 disease. However, the evidence is unclear as limited cases have been reported. Hispanic ethnicity and obesity also appear to confer risk of more severe disease in adults.

CR-Case(s) Presentation(s)

Case 1:
A 12-year-old female with DiGeorge syndrome with hypogammaglobulinemia on monthly immunoglobulin replacement and T cell lymphocytopenia with normal mitogens, congenital heart disease, and VP shunt, presented to the ED with headache and an episode of emesis. Brain CT and shunt series were obtained to rule out shunt malfunction, both normal. Immediate family members were diagnosed with COVID-19 and our patient also tested PCR positive. She was treated with supportive care. Repeat COVID-19 PCR testing 3 weeks later was still positive, although asymptomatic.

Case 2:
A 13-year-old Hispanic male with DiGeorge syndrome, obesity, and congenital heart disease with immediate family members diagnosed with COVID-19 also tested PCR positive. Three family members required hospital admission, one of which required ICU admission and remains on supplemental oxygen. He had history of normal T lymphocytes, normal mitogens, and low IgM. Despite a complex medical history, he remained asymptomatic. Both patients had history of normal mitogens.

CR-Diagnosis and Discussion

It is well documented that excessive production of proinflammatory cytokines is associated with worse severity and outcome of COVID-19.

- PIDD patients may have a mild disease course and favorable outcome as a result of a weak immune system, as seen in our patients. This was supported by a systematic review of immunocompromised children and adults with cancers, transplantation and immunodeficiency with COVID-19.
- Another systematic review and meta-analysis of immunosuppressed patients with COVID-19 concluded that immunodeficient and immunosuppressed patients are at increased risk of severe COVID-19 disease. However, the results were not statistically significant.

- Studies of PIDD patients and COVID-19 are limited. To our knowledge, no reports of pediatric patients with DiGeorge syndrome and COVID-19 have been reported. Hence, more cases and studies are needed.
- In addition, obesity and Hispanic race were found as potential risk factors for a severe disease course in adults studies, however, here we describe a Hispanic and morbidly obese patient who was completely asymptomatic.

CR-Conclusion and Significance

Pediatric patients with DiGeorge syndrome may not, necessarily, be at increased risk of severe COVID-19 disease. More cases are needed to better assess the association and understand risk factors in pediatric patients with PIDD and COVID-19.

Abstract #28: (RES-008-CR)

Fonseca A*; Sunny J; Muniz Crim A; Felipez LM. ***"Anti-Tumor Necrosis Factor Alpha (TNF-A) Infliximab-Induced Pleural Effusion and Pericarditis In Crohn's Disease".***

CR-Introduction/ Background

Crohn's disease (CD) is a chronic inflammatory disease that is associated with intestinal and extraintestinal manifestations. Rarely, inflammatory bowel disease (IBD) involves the pleural space and pericardium. Infliximab therapy is an anti-tumor necrosis factor alpha (TNF-α) blocking therapy associated with an increased likelihood of achieving and maintaining remission, preventing recurrence, and improving quality of life in Crohn's disease. However, 6% of the patients with CD experience serious adverse effects, including infections and immunogenicity in the form of seroconversion, systemic lupus (SLE), and drug-induced lupus following treatment with infliximab. We report a case of 19-year-old Hispanic male with CD that acquired anti-TNF-α induced lupus after infliximab therapy presenting with pleural effusion and pericarditis.

CR-Case(s) Presentation(s)

A 19-year-old Hispanic male with stenosing ileocolonic Crohn's disease on infliximab 10mg/kg presented with a 2-week history of pleuritic chest pain. The pain was located at the mediastinum, radiating to the shoulder, worse with deep breaths and lying flat, and improved by standing up or siting. Laboratory workup was remarkable for a leukocytosis of 13.2 10K/uL, CRP of 8.2 mg/L, rheumatoid factor of 12.5 IU/mL, and calprotectin of 480 mcg/gm. EKG showed non-specific ST abnormalities. CT of the chest showed mild pericardial thickening with pericardial effusion and minimal bibasilar pleural thickening with bibasilar pleural effusions. Echocardiogram showed small circumferential pericardial effusion. Cardiology recommended colchicine 0.6mg PO TID for 6 weeks. Rheumatological workup results obtained remarkable for negative ANA titers, positive ANA IFA, and increased anti-histone antibodies (1.7 U). Pain improved after discharge but resurfaced

six days later, on day of infliximab infusion. Chest x-ray showed blunting of the posterolateral left lateral costophrenic angle, suggestive of a small pleural effusion. EKG showed diffuse abnormal T waves. Echocardiogram showed interval decrease in pericardial effusion. Ultrasound of the chest showed right and left simple pleural effusions, measuring 34mL and 150mL, respectively. Ultrasound-guided thoracentesis was done and removed 430cc of clear amber fluid. Body fluid cultures were negative and body fluid chemistry was consistent with an exudative pleural effusion. He was continued on colchicine 0.6mg BID and discharged with a weaning schedule of prednisone 20mg PO daily. Infliximab was discontinued and he was started on an induction of ustekinumab 390mg with a maintenance regimen every 8 weeks. Serum ustekinumab (UST) concentration was 13.9 μg/mL and antibodies to ustekinumab (ATU) concentration was <1.6 U/mL. Currently, the patient denies any chest pain or shortness of breath and continues with the previously mentioned treatment.

CR-Diagnosis and Discussion

Anti-TNF-α therapy is effective in patients with IBD and other chronic inflammatory diseases. However, this therapy may be associated with the induction of autoimmunity including the formation of anti-nuclear antibodies (ANA), double stranded (ds) DNA antibodies, anti-histone antibodies, and others, thus developing as a drug-induced lupus. Drug-induced lupus is a syndrome similar to idiopathic systemic lupus erythematous (SLE). Sulfadiazine, infliximab, etanercept, and others are among the drugs implicated in a drug-induced lupus. Only a few cases in IBD patients have been reported. Rare manifestations are myalgias, pericardial/pleural effusion, glomerulonephritis, valvulitis, pneumonitis, deep vein thrombosis, and oral ulcers. Infliximab-induced pericarditis and pleural effusions are reported infrequently in the literature. Drug-induced SLE only has been associated with infliximab therapy for CD in 0.6% to 1.6% of cases. It has been theorized that infliximab may have pro-inflammatory activity in certain tissues or have a delayed type III hypersensitivity reaction. The main approach in the treatment of anti-TNF-α induced lupus is withdrawal of the offending drug. In some patients, corticosteroids and immunosuppressive agents might be required to achieve full recovery.

CR-Conclusion and Significance

Our patient is unique as few cases of anti-TNF-α infliximab induced pleural effusion and pericarditis in patients with Crohn's disease are reported. After discontinuation of the offending drug and start of ustekinumab, the patient's chest pain has slowly improved. Anti-histone antibodies have returned to normal.

Abstract #29: (RES-009-CR)

Fonseca A*; Sunny J; Muniz Crim A; Felipez LM. ***"Very Early Onset IBD (VEOIBD) Presenting With Recurrent Leukocytoclastic Vasculitis Preceded by Streptococcal Pharyngitis".***

CR-Introduction/ Background

Inflammatory bowel disease (IBD) that presents in children <6 years of age is known as very early-onset IBD (VEO-IBD), most of the time refractory to treatment in comparison with older children and adults. Extra-intestinal manifestations (EIMs) of IBD are common, more frequently in Crohn's disease (CD) than in ulcerative colitis (UC). The most common cutaneous manifestations of IBD are erythema nodosum (EN) and pyoderma gangrenosum (PG). Other less common manifestations reported includes psoriasis, Sweet's syndrome, dermatitis herpetiformis, epidermolysis bullosa acquisita, necrotizing vasculitis, and leukocytoclastic vasculitis (LV). Association between LV and UC is uncommon and in most cases cutaneous LV proceeds the intestinal symptoms. We report a case of 6-year-old female that was diagnosed late with VEOIBD-UC phenotype and has had multiple episodes of leukocytoclastic vasculitis each preceded by Streptococcal pharyngitis.

CR-Case(s) Presentation(s)

A 6-year-old Hispanic female with previous self-resolving episodes of bloody stools and recurrent gastrointestinal infections presented with bilateral swelling of the hands and lower extremities, inability to bear weight, petechial rash of the palms, soles, and legs, and watery diarrhea with specks of blood. The patient had 6 similar previous episodes, each preceded by Streptococcal pharyngitis and treated with antibiotics and steroids. Skin biopsy had shown leukocytoclastic vasculitis with negative IgA. During initial presentation she had anti-strep O and anti-DNase B increased. Rheumatoid factor and serum Ig A level normal. *Clostridium difficile* PCR was positive for *Clostridium difficile* toxins A and B. Treatment for LV consisted of methylprednisolone IV 20mg for four days with a weaning schedule of prednisolone for two weeks and naproxen 250mg BID for three days. *Clostridium difficile* was treated with metronidazole 250mg TID for ten days. She remained stable for three years; then presented with anterior chest pain with radiation to the left shoulder, shortness of breath and continuous bloody stools. CT angiogram was normal. Abdominal MRE showed thickening at the transverse colon, distal colon, sigmoid colon, and rectal wall with chronic inflammation at the sigmoid colon and distal colon. Endoscopy and colonoscopy showed mild chronic gastritis at the antrum and body of the stomach with moderate chronic active colitis at the distal colon, sigmoid colon, and rectum. Due to previous history of bloody stools since the age of 5, a VEOIBD ulcerative colitis phenotype diagnosis was made. She was thus started infliximab 10mg/kg induction. Initial induction Anser IFX level of 12.4 ug/mL, ATI <3.1 unit/mL. Infliximab frequency was decreased to every 4 weeks. Currently, the patient remains on clinical and biochemical remission and no recent episodes of LV.

CR-Diagnosis and Discussion

Leukocytoclastic vasculitis is a disease with inflammation of the postcapillary venules with neutrophilic infiltration and nuclear debris. It is believed to be triggered by various drugs, infections, malignancies, and systemic and autoimmune disorders. Association between LV and UC is uncommon and in most cases LV proceeds the intestinal symptoms and is seen in older males. It's believed the inflamed colonic mucosa in UC allows fecal antigen exposure to submucosal lymphoid tissue, resulting in the formation of immune complexes. The deposition of immune complexes in the vascular wall of small dermal vessels leads to complement activation, leukotaxis, release of lysosomal enzymes, and destruction of the vascular wall, resulting in erythrocyte extravasation and tissue necrosis. Treatment has included steroids, mesalamine, dapsone, colchicine, and infliximab.

CR-Conclusion and Significance

Our patient is unique as no case report has been published with multiple episodes of leukocytoclastic vasculitis in association with VEOIBD-UC phenotype.

Abstract #30: (RES-010-CR)

Fonseca A*; Roberts A; Barry R; Meyer K; Malvezzi L; Tackett J; Koyfman S. ***"Pediatric Patient Presenting with Upper Gastrointestinal Bleed Following Consumption of Whole Sunflower Seeds".***

CR-Introduction/ Background

Seed bezoars can form as a result of accumulated indigestible vegetable or fruit seeds in the intestinal lumen. Formation of bezoars from sunflower seeds can be seen in children or preteens who are unable or unwilling to spit out shells, particularly when the shells are flavored. This bezoar can become further dehydrated and harden as it moves through the colon towards the rectum resulting in fecal impaction. Sunflower seed impaction in the rectum has been reported in literature with the majority of cases requiring manual disimpaction under general anesthesia for resolution of symptoms. However, GI bleeds related to seed bezoars are extremely rare and have not yet been reported in literature. In this report, we examine the case of a 5-year-old male who presented with an upper GI bleed 3 days after consuming whole sunflower seeds with their shells intact.

CR-Case(s) Presentation(s)

The patient is a previously healthy 5-year-old male who initially presented to the Emergency Department with fatigue, syncope, hematochezia, and hematemesis 3 days after consuming an entire bag of sunflower seeds with the shells intact. On the day of admission, the patient had 3 episodes of syncope and one episode of emesis containing dark blood and shelled sunflower seeds. His mother also reported an episode of dark

bloody stools containing shelled sunflower seeds one day prior to presentation. He was found to have a microcytic anemia with a hemoglobin level of 6.4 g/dL (hematocrit 20.7%) on complete blood count. Coagulation studies including prothrombin time, international normalized ratio, and partial thrombin times were within normal limits. Complete metabolic panel showed a low albumin level of 2.9 g/dL and total protein of 5.3 g/dL. Otherwise ALT, AST, alkaline phosphatase, amylase, lipase, and total bilirubin levels were within normal limits. C-Reactive Protein was less than 2.9 mg/L. Computed tomography (CT) scan of the abdomen and pelvis revealed mild thickening of the jejunum in the left upper quadrant without colonic thickening or obstruction and no evidence of a gastrointestinal bleed. Abdominal X-Ray showed a nonobstructive gas pattern with mild gaseous distention of the small bowel loops. Chest X-Ray was unremarkable and without pleural effusions or consolidation. Patient was transfused 1 unit of packed red blood cells prior to being transferred to the Pediatric Intensive Care Unit (PICU).

While in the PICU, the patient required a second blood transfusion (hemoglobin level 6.7 g/dL). His hemoglobin levels remained stable following the second transfusion. He was placed on a cleanout regimen with polyethylene glycol and continued to pass sunflower seeds and black stools in the PICU. Gastric lavage revealed no evidence of active upper gastrointestinal bleed. CT enterography showed no acute abdominal or pelvic pathology or signs of bowel wall thickening. Esophagogastroduodenoscopy showed some small ulcers but no active bleeding. A capsule endoscopy (CE), which did not pass the pylorus, was placed during the upper endoscopy and visualized mild gastritis, post-biopsy bleeding, and no source of active bleeding. During the colonoscopy, sunflower seeds mixed in with stool were manually disimpacted from the rectum. The colonoscopy was limited due to large amounts of black stool. No active bleed was identified. A Meckel's scan was performed and was found to be normal. The patient clinically improved and hemoglobin levels stabilized. Therefore, it was decided not to pursue further investigation into the source of the GI bleed.

CR-Diagnosis and Discussion

Seed bezoars can result in fecal impaction and, rarely, an upper GI bleed. However, despite undergoing diagnostic testing with esophagogastroduodenoscopy and colonoscopy, GI bleeds will not be identified approximately 5% of the time. One explanation may be that the source is located in the small intestine, which is relatively less accessible than the stomach or colon and more difficult to identify. CE, which has a diagnostic yield of 70%, can be considered in these cases.

CR-Conclusion and Significance

In this case report, we report a rare case of seed bezoars causing a GI bleed that was not identified despite evaluation with esophagoduodenoscopy, colonoscopy, CT enterography, and nuclear medicine scan. Due to significant clinical improvement and stabilized hemoglobin levels, further investigation with CE past the pylorus was not pursued. Accordingly, the small intestines were not examined and may have been the source of the

bleed. Evidently, it is still important to maintain a high index of suspicion of the cause of bleeding, as with our patient, especially when diagnostic testing doesn't reveal a clear answer.

Abstract #31: (RES-011-CR)

Gawron Roberts A*; Colon Guzman S; Bujarska M; Sunny J; Gonzalez-Vallina R.

"Pediatric Patient Presenting with Diphyllobothrium Latum Infection Following Consumption of Raw Fish at a Local Sushi Restaurant".

CR-Introduction/ Background

Diphyllobothrium latum (*D. latum*) is a cestode that can cause an intestinal parasitic infection following the ingestion of raw or poorly cooked fish containing larvae. It is often referred to as the "fish tapeworm" or the "broad tapeworm" and is the longest known human tapeworm. The adult tapeworms can reach a length greater than 10 meters. *D. latum* can be found in both endemic and non-endemic regions and outbreaks have been reported in South America, Japan, Siberia, Europe, and North America. Cases of *D. latum* have recently increased in parts of the developed world presumably due to the increased popularity of raw fish dishes including but not limited to Japanese sushi, sashimi, tartare maison, and ceviche. Diphyllobothriasis occurs after the tapeworm matures within the host's intestines. Infected humans begin to pass eggs in their stools approximately 15 to 45 days following ingestion of larvae.

Mild gastrointestinal symptoms are often seen with diphyllobothriasis but patients may also be asymptomatic. More severe cases can present with significant anemia or luminal obstruction. Studies suggest that twenty-five percent of patients infected with *D. latum* will present with symptoms of abdominal pain, fatigue, headaches, diarrhea, or pernicious anemia. The majority of identified cases of *D. latum* has been reported in middle-aged men. Here we present the case of a 7-year-old Hispanic male who passed a 2.2 meter tapeworm after consuming raw fish at a local sushi restaurant.

CR-Case(s) Presentation(s)

The patient is a previously healthy 7-year-old Hispanic male who initially presented to the gastroenterologist's office after passing a tapeworm measuring 2.2 meters in his stool. One week prior to his presentation, the patient had eaten raw fish at a local sushi restaurant with his parents. Two days later, he began with diffuse abdominal pain, nausea and vomiting during his soccer practice. No household members or close contacts reported similar symptoms. There was no recent travel history. Over-the-counter bismuth subsalicylate was tried with little relief. Later that day he had 5 episodes of diarrhea. During one of these episodes, the patient noticed a tapeworm projecting from his anus. He called out for his mother, reporting that he had "a piece of spaghetti" protruding out of his anus. His mother manually extracted the parasite. The extracted parasite and stool were sent for analysis. The

laboratory confirmed the diagnosis of *D. latum* proglottids and eggs in the stool. Serum iron and ferritin levels were found to be within normal limits at 75 ug/dL and 30 ng/mL respectively. Further workup was not obtained and the patient was treated with a single dose of praziquantel 300mg and lansoprazole 15 mg daily for two weeks. Family members were also treated with a one time dose of praziquantel 10 mg/kg. Repeat stool cultures following treatment were negative and the patient's abdominal symptoms quickly resolved. The patient did not have any recurrence of symptoms and resumed his normal activities following treatment.

CR-Diagnosis and Discussion

Diphyllobothriasis should be considered in the differential of patients presenting with abdominal pain, fatigue, headaches, and diarrhea regardless of travel history especially in the setting of recent raw fish consumption. Although *D. latum* was rapidly identified and treated in our patient without the need for extensive laboratory workup, clinicians should consider obtaining a workup that includes complete blood count and vitamin B12 levels in suspected cases. These studies may reveal eosinophilia, megaloblastic anemia, and vitamin B12 deficiency which would be suspicious for infection with *D. latum*. Diagnosis is determined by the presence of proglottids or eggs in the patient's stool. Treatment involves a single dose of praziquantel 5 - 10 mg/kg taken orally for both adults and children. Niclosamide is an alternative treatment option. There are currently no formal recommendations regarding the treatment of household members of patients with *D. latum*. However, given the fact that humans may remain asymptomatic despite being infected, physicians should use their clinical judgement in determining whether close contacts or household members should also be treated.

CR-Conclusion and Significance

Abstract #32: (RES-012-CR)

Gawron Roberts A*; Bujarska M; Bauer M; Brathwaite C; Pelaez L; Reeves-Garcia J.

"Gastric Adenocarcinoma and Proximal Polyposis of the Stomach in a Hispanic Pediatric Patient with APC Gene Variant c.-191T>G".

CR-Introduction/ Background

Gastric adenocarcinoma and proximal polyposis of the stomach (GAPPS) is a rare autosomal dominant gastric polyposis syndrome linked to a variant in the promoter 1 B region of the APC gene (3,5). It is defined by the presence of numerous polyps (>100 polyps) in the body and fundus of the stomach with sparing of the antrum, small intestines and colon (1,3,6,9,11). The sparing of the antrum, colon and intestines distinguishes it from familial adenomatous polyposis (FAP) syndrome, an autosomal dominant polyposis syndrome characterized by multiple adenomatous polyps affecting

the colon and rectum also linked to variants in the APC gene (3,4). FAP may occasionally present with extensive gastric fundic gland polyposis especially in individuals with large deletions in the promoter 1B (3,4,8,10).

There have been few reported cases of GAPPS in the literature and all reported cases have identified variants in the promoter 1B of the APC gene (2,3,6,7,11). Li et al. reported three different point mutations (c.-195A>C, c.-191T>C, c.-192A>G) that affect the expression of the promoter 1 B of APC by interruption of the Ying Yang 1 (YY1) binding motif (3). Deletions in the promoter 1B have been observed in some cases of FAP, however, point mutations in promoter 1B rarely lead to FAP and are more closely associated to GAPPS (3). The APC gene promoter 1B is known to be highly transcribed in the gastric mucosa compared to the intestinal mucosa (2,3). Furthermore, it is believed that the colon and intestines are protected by the promoter 1A of the gene, which accounts for the sparing of the intestinal mucosa in patients with GAPPS (2,3).

The age of onset is highly variable among individuals with GAPPS and is likely influenced by multiple factors including genetics, environment, and lifestyle habits (2). In those with extensive polyposis in the setting of a promoter 1B APC variant, the risk of developing gastric adenocarcinoma is significant and ranges between 12-25% based on current studies (2,6,7). In a study done by Foretova et al. at the Masaryk Memorial Cancer Institute (MMCI) in Brno, Czech Republic, 6 out of 24 individuals carrying the APC gene promoter 1B point mutation (variant c.-191T>C) were diagnosed with gastric cancer (2).

The earliest age at diagnosis reported in their study was 22 years (2). In the patients observed by Repak et al., one female (age 26) developed poorly differentiated adenocarcinoma with metastasis despite endoscopic surveillance with multiple biopsies every 18-24 months (6). She died shortly after beginning palliative chemotherapy (6). The transition from dysplasia to gastric cancer can be rapid and endoscopic surveillance often misses tumors or dysplastic polyps making endoscopic surveillance a poor option for early cancer detection (2). Therefore, the MMCI recommends prophylactic total gastrectomy with D2 lymphadenectomy consistent with the standard of care for stomach malignancies to all patients identified with extensive gastric polyposis secondary to GAPPS (2). Currently, non-gastric manifestations of GAPPS are poorly defined in literature (5). Despite relative sparing of the antrum, intestines, and colon, colonic polyps have been reported in literature in patients with the GAPPS phenotype (5). As a result, Martin et al. advocates for colonoscopic surveillance in the routine management of patients with confirmed GAPPS (5).

Furthermore, genetic testing should be recommended to the relatives of a GAPPS affected individual according to the National Comprehensive Cancer Network (NCCN) clinical practice guidelines for gastric cancer. The MMCI currently recommends genetic testing to all relatives of patients with confirmed GAPPS (2). GAPPS has been identified in people of Australian, white American, white Canadian, Czech, and Japanese descent (2,3,6,11). There has not been a case report or study published to date identifying GAPPS in a Hispanic patient. This report examines the case of a 16-year-old Hispanic female of

Cuban descent diagnosed with GAPPS after endoscopy revealed extensive fundic gland polyposis. Genetic testing identified an APC gene promoter 1B point mutation, variant c.-191T>G. Varsome classifies this variant as a variant of "uncertain significance." While similar variants linked to GAPPS have been reported in literature (i.e c.-191T>C, c.-195A>C, c.-192A>G), variant c.-191T>G, has not (2,6).

CR-Case(s) Presentation(s)

A 16-year-old Hispanic female of Cuban descent was referred to our gastroenterology clinic following several months of intermittent epigastric pain, dyspepsia, nausea, and vomiting. Her past medical history was notable for clinically diagnosed dyspepsia and irregular menses secondary to polycystic ovarian syndrome (BMI in the 55th percentile). Her only home medication was a daily oral contraceptive pill.

Notable family history included her father who died of metastatic hepatocellular carcinoma at age 68, a maternal grandmother who died of colon cancer at age 60, and a paternal grandmother who died of leukemia at age 65 (Figure I).

The patient's physical exam and vitals were found to be unremarkable during the initial visit. Lab workup with complete blood count, urinalysis, and comprehensive metabolic panel were all normal except a lipid panel with mildly elevated cholesterol level. Stool culture, stool *Helicobacter Pylori*, and stool ovum and parasite testing were all negative. Patient was sent for an endoscopy which detected over 150 fundic gland polyps (Figure II). No polyps were seen in the antrum, pylorus, or small bowel (Figure III, IV). The esophagus was normal. Four polyps were removed in toto by polypectomy and sent to pathology.

Microscopic examination of the fundic gland polyps showed distorted glandular architecture composed of irregular and cystically dilated glands with stellate configuration lined by normal gastric body-type epithelium (Figure V). No atypia or dysplasia was reported. One week later, the patient returned for a complete workup with colonoscopy, capsule endoscopy, and repeat endoscopy. This time, two polyps were removed in toto by polypectomy with similar findings on pathology. The mucosa of the esophagus, antrum, duodenum, and colon were normal. Colonoscopy did not detect any polyps in the colon and biopsies showed normal colonic mucosa without evidence of aberrant crypt foci. Video-Capsule Endoscopy revealed gastric fundus polyposis and lymphoid hyperplasia of the terminal ileum. No small bowel polyps were identified.

The patient was referred for full genetic workup and genetic counseling to a clinical geneticist at our institution with experience in the clinical management and genetic workup of patients with suspected underlying hereditary cancer syndromes. Given the extent of polyposis in our patient with a strong family history of malignancies, our geneticist selected the OncoGeneDx Colorectal Panel by GeneDx, an OPKO Health company located in Gaithersburg, MD. The rationale for the selection of this gene panel was its ability to examine 20 different genes involved in hereditary colorectal cancer and

other gastrointestinal cancers. Buccal mucosa DNA was used for this panel and informed consent was obtained from the patient's mother prior to genetic testing.

Next generation sequencing using the OncoGeneDx Colorectal Cancer Panel identified a variant in the promoter 1B region of the APC gene, c.-191T>G, a nucleotide substitution 191 base pairs upstream of the ATG translational start site in the 5' untranslated region. This variant was considered "likely pathogenic" in the genetic report. The patient was also positive for a POLE gene variant, c.2510T>C, at the cDNA level and p.Phe837Ser at the protein level. This variant was considered a variant of uncertain significance. The patient was negative for PTEN, STK11, BMPR1A, SMAD4, ATM, AXIN2, CDH1, CHEK2, EPCAM, MLH1, MSH2, MSH6, MUTYH, NTHL1, PMS2, POLD1, SCG5/GREM1, and TP53 gene variants. Genetic testing was recommended to the patient's mother and relatives. The mother's genetic testing did not reveal any APC gene mutations, but she did share the same variant of uncertain significance on the POLE gene. The patient's father's other four offspring all of whom live in Cuba were contacted and informed of their risks but they were not interested in genetic testing or further workup. We advised our patient to undergo a prophylactic total gastrectomy. At first, the family refused the procedure. The mother later sought out the National Institute of Health (NIH) who became interested in her case and also recommended total gastrectomy. She is now scheduled for prophylactic total gastrectomy in June of 2021. Given the novelty of our variant, c.-191T>G, the variant was reported by our team to the LOVD3 database for APC mutations.

CR-Diagnosis and Discussion

To date, there have been few studies reporting GAPPS none of which describe GAPPS in a person of Hispanic descent. Furthermore, c.-191T>C, c.-192A>G, and c.-195A>C point mutations in the promoter 1B of the APC gene have been identified, but the gene variant exhibited in our patient, c.-191T>G, has not (2,5). The APC variant c.-191T>G is also unreported in multiple genetic databases making it unique. In Varsome, it is classified as a "Variant of Unknown Significance." This case introduces a new variant in a unique patient population and highlights both the importance of early detection of GAPPS with endoscopy and early intervention. After taking into consideration the risk of gastric cancer associated with promoter 1B APC gene variants and the limitations of endoscopic surveillance, a prophylactic gastrectomy was recommended to our patient who ultimately agreed to the procedure.

CR-Conclusion and Significance

Abstract #33: (RES-013-CR)

Gawron Roberts A*; Bujarska M; Muniz Crim A; Felipez LM. ***"A Case of Very Early Onset Inflammatory Bowel Disease (VEOIBD) Presenting with Salmonellosis".***

CR-Introduction/ Background

A 3-year-old Hispanic female presented initially to the Emergency Department (ED) with 3 days of high fevers (Tmax 104F) and 10 days of loose, bloody stools. Stool culture was positive for Salmonella species. Subsequent blood cultures returned positive for Salmonella species. The patient was diagnosed with Very Early Onset Inflammatory Bowel Disease (VEOIBD) Ulcerative Colitis (UC) type following a colonoscopy that depicted severe colonic friability and edema and biopsy findings of mild crypt distortion and chronic inflammation in the colon. This case highlights the possible association between acute Salmonella infection and the onset of VEOIBD.

CR-Case(s) Presentation(s)

A 3-year-old Hispanic female with a past medical history of milk protein allergy and gastroesophageal reflux presented to the ED with 3 days of high fevers (Tmax 104F) and 10 days of loose, bloody stools. Travel history was significant for cruise travel to Belize, Honduras, and Mexico 1 month prior to presentation. Her parents denied any sick contacts. There was no reported history of IBD in the family. Complete Blood Count showed mild leukocytosis. Stool culture was positive for Salmonella species. She was initially treated with probiotics. Two weeks later, she returned to the ED with persistent bloody stools and 3 days of lower abdominal pain. Labs were remarkable for an erythrocyte sedimentation rate of 67 mm/hr and moderately elevated liver enzymes (ALT 33 U/L, AST 65 U/L). Stool calprotectin was 615.5 mcg/gm and her Inflammatory Bowel Disease (IBD) Panel was positive for pANCA and elevated anti-A4-Fla2, anti-CBir1, and anti-FlaX. ATG 16L1 SNP (rs2241880) variant was detected. Blood cultures were obtained prior to beginning treatment and returned positive for Salmonella species, serotype subspecies IV determined by Whole Genome Sequencing (WGS). The patient was admitted for treatment with IV antibiotics. Due to persistent bloody diarrhea, an endoscopy and colonoscopy were performed following adequate treatment of Salmonellosis with IV antibiotics. The endoscopy report was consistent with mild gastritis. The colonoscope did not extend past 40 cm due to severe colonic friability and edema. Pathology report indicated mild crypt distortion and chronic inflammation in the colon concerning for VEOIBD UC type. The small bowel appeared normal on CT Enterography. Genetic testing in the patient was negative. The EGL Genetics Early Onset Inflammatory Bowel Disease (EOIBD) Deletion/Duplication Panel which uses comparative genomic hybridization (CGH) array was negative. The EOIBD Sequencing Analysis which uses whole genome sequencing found no pathogenic variants. The Prometheus IBD sgi Diagnostic test resulted in a pattern consistent with IBD but inconclusive for Crohn's Disease vs. Ulcerative Colitis. Immunological workup proved to be unremarkable and negative for underlying immunological disorders. Neutrophil oxidative burst assay showed 99.2% positive neutrophils within the normal range and the patient was noted to have elevated B and NK lymphocyte subset count on the

Lymphocyte Subset Panel. The patient was started on immunosuppressive therapy with mesalamine 1.125g daily. After initiation of therapy, she has achieved clinical and biochemical remission. Her most recent stool calprotectin level was 60 mcg/gm.

CR-Diagnosis and Discussion

IBD is a condition of the gastrointestinal tract caused by an abnormal immune response in genetically susceptible hosts resulting in intestinal tissue damage and inflammation2,3. The etiology of IBD is thought to be multifactorial in nature and influenced by environmental and genetic factors4. Very Early Onset Inflammatory Bowel Disease (VEOIBD) has been described as IBD diagnosed in children less than 6 years of age1. The frequency of IBD is increasing worldwide with the greatest increase in incidence occurring in young children1,2.

One of the many factors believed to play a role in the development of IBD is infection with pathogenic microorganisms such as Campylobacter, Escherichia coli, Clostridium difficile, or Salmonella enterica serovar Typhimurium (S. Typhimurium)2,3. These organisms may trigger IBD development through the disruption or alteration of the intestinal epithelial barrier, intestinal microflora, and intestinal immune response2,3. Particular attention has been placed on S. Typhimurium infection and its relationship to IBD3,4. Infection with S. Typhimurium is believed to provoke chronic inflammation in patients with IBD3,4. In both humans and in mice, S. Typhimurium has been shown to cause persistent and relapsing infection despite treatment with antibiotics4. Furthermore, patients with IBD may have increased susceptibility to infection by S. Typhimurium due to genetic factors and altered microbiota3,4. This report highlights the case of a 3-year-old Hispanic female who presented with salmonellosis and was later diagnosed with VEOIBD UC type after her symptoms did not improve following adequate treatment of Salmonellosis with IV antibiotics. She was found to have improvement in her symptoms and biochemical markers only after starting aminosalicylate therapy. Given her negative family history and genetic and immunological workup, it is believed that the Salmonella infection may have precipitated the development of IBD in this patient. Infection with S. Typhimurium may play an important role in the development of IBD in some individuals3,4. IBD should be considered in the differential for young children presenting with persistent bloody stools, abdominal pain, and positive stool cultures for Salmonella despite adequate and appropriate antibiotic treatment.

CR-Conclusion and Significance

Abstract #34: (RES-014-CR)

Graneiro A*; Chang Y; Hernandez Trujillo V. ***"Chronic chough in a patient with CVID and GLILD".***

CR-Introduction/ Background

Common Variable Immunodeficiency (CVID) is a primary immunodeficiency that results in variable presentation. Patients are prone to infections, malignancies, and autoimmune disorders. They are also prone to developing Granulomatous Lymphocytic Interstitial Lung Disease (GLILD) and bronchiectasis, which can manifest as chronic cough and shortness of breath (1). These require early and aggressive detection and treatment to prevent morbidity and mortality. In addition, this group of patients can also be more sensitive to certain environmental products. For example, antidegradration agents that are used to enhance rubber products—including paraphenylenediamine (PPD)—are collectively known as black rubber mix and can be found in many hair products. The rationale for the use stems from the fact that when oxidized, PPD turns hair a darker color, hence its popularity (2).

CR-Case(s) Presentation(s)

This patient had a history of CVID on subcutaneous gamma globulin replacement, GLILD on infliximab, congenital heart disease, allergic rhinitis, chronic sinusitis and contact dermatitis, who presented with a dry cough for months. One day, the patient went to the hair salon to have her hair colored and she had an acute worsening of her cough. She had not dyed her hair in over one year.

During this time, she was followed by multiple specialties including pulmonology for her history of GLILD, who suspected her cough was upper respiratory in nature. Skin prick testing was positive only to feathers. A CT scan of the sinuses was normal. She was evaluated by ENT with rhinoscopy that was normal. She was evaluated by Cardiology and noted to have elevated Fontan pressures, though unlikely the cause of her cough. She had a bronchoscopy to evaluate her cough, with biopsies and cultures, which were all normal.
Treatment for allergic rhinitis and asthma was optimized, including Albuterol, Inhaled Fluticasone, Fexofenadine, and Azelastine. She was treated with Cefdinir for three weeks for suspected sinus infection. She also was treated with a short course of oral steroids. Over a course of several weeks, the cough slowly improved. She continued with her baseline cough however.

Of note, in the preceding months she had been developing contact reactions to medical tape, jewelry and EKG leads. Patch testing was positive to nickel, gold, Cl+Me-Isothiazolinone and black rubber mix.

CR-Diagnosis and Discussion

GLILD has been reported in 8-22% of patients with CVID (3). As our patient had known history of GLILD, bronchiectasis, and congenital heart disease, these were all evaluated.

We then addressed the more common causes for dry cough such as chronic sinusitis, allergic rhinitis with post nasal drip, and asthma.
Our patient presented with an acute exacerbation which worsened her baseline cough. Due to her history of multiple medical conditions requiring procedures, it is likely she became sensitized to contact allergens over the years. As we use more chemicals in our foods, soaps, and beauty products, we must acknowledge their potential impact on our health as well.

Contact dermatitis is estimated to occur in 20% of the general population and PPD is one of the more common culprits (4). It is thought to be present in over 70% of hair products worldwide and can contribute to local reactions such as dermatitis or systemic reactions including cough and anaphylaxis (5). Given its long-lasting nature, it is a common additive to hair dye products and it is regarded as an extreme sensitizer (5). Patients should read labels carefully, eliminate hair products with black rubber mix/PPD or opt for natural hair products.

CR-Conclusion and Significance

Patients with primary immunodeficiency and chronic lung disease with persistent cough and acute exacerbation, who have eliminated pertinent environmental allergens and are on appropriate treatment, should be evaluated for infection, as well as contact allergens. We therefore recommend that, in patients with primary immunodeficiency and chronic lung disease presenting with acute exacerbation of cough, unexpected causes such as contact allergy, to common sensitizers should be considered.

Abstract #35: (RES-015-CR)

Graneiro A*; Mandel G; Laufer M. ***"What to consider when things start to heat up: A rare cause of FUO".***

CR-Introduction/ Background

Fever of unknown origin is a common conundrum for the pediatric specialist. Differential diagnosis includes infectious and non-infectious causes. We present an unusual cause of FUO.

CR-Case(s) Presentation(s)

A 15-year-old female with no significant past medical history presented with 7 weeks of daily high fever, intermittent rash, vomiting, and malaise. She was treated with Augmentin for one week at the beginning of her illness and symptoms subsided, but returned shortly thereafter.
Physical exam was unremarkable. Bloodwork was remarkable for leukocytosis with left shift, microcytic anemia, elevated LDH and ferritin above 20,000 ng/ml. A CT of the abdomen was ordered and we obtained the following result:

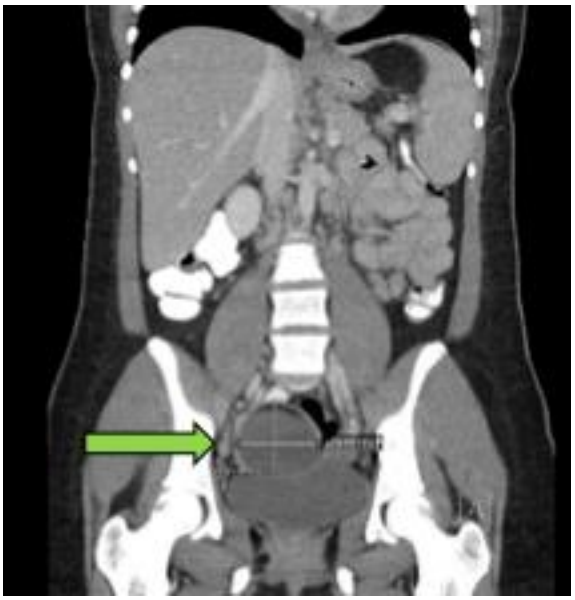
(image at the end of the document)

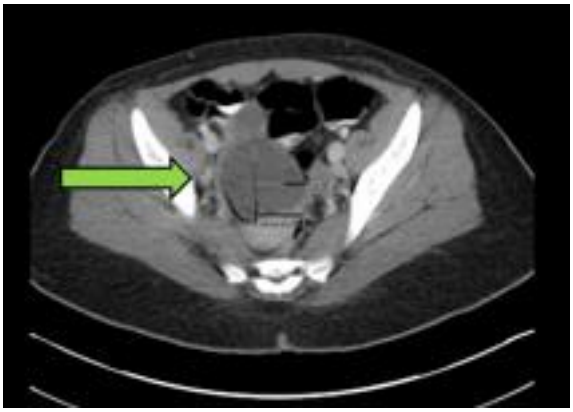
Patient underwent laparoscopic excision, removing purulent and foul-smelling mass which subsequently grew pansensitive pseudomonas aeruginosa on tissue culture; Biopsy confirmed mature cystic teratoma. Patient's course was further complicated by hemorrhagic ascites and anemia. Patient was stabilized and fevers resolved with removal of mass and course of antibiotics. Upon 18 month follow-up, she was doing well without continued medical issues.

CR-Diagnosis and Discussion

CR-Conclusion and Significance

In females with chronic fever of unknown origin, when other sources of fever have been ruled out, it is important to consider rare causes that can include ovarian masses, even when physical examination is unremarkable.





Abstract #36: (RES-016-CR)

Lopez Gonzalez MS*; Caicedo Oquendo LF. ***"Dorsal Agenesis of the Pancreas: A Rare Entity".***

CR-Introduction/ Background

Agenesis of the dorsal pancreas is a rare entity, resulting from the defective development in utero of the organ. This malformation was first described in 1911 during an autopsy and was associated with diabetes, approximately ten years before the discovery of insulin. It can either be asymptomatic and incidentally found through imaging or may present with mild symptoms to more severe pathology such as diabetes mellitus, acute or chronic pancreatitis. The dorsal pancreatic agenesis is described in two forms, the partial and the complete form.

Until today, around 100 cases of dorsal agenesis of pancreas have been reported in the world literature and we report one case that was incidentally detected by imaging at our institution in 2019.

CR-Case(s) Presentation(s)

15-year-old male, PMHX (-) presented with 6 month history of bloody stool, calprotectin 203.8 mcg/gm, negative Infectious stool studies, s/p EGD and colonoscopy at OSH: Colonoscopy: macroscopic (+) ulcers cecum, microscopic: (+) chronic active colitis / crypts abscesses. Diagnosed with possible IBD, came to us for second opinion. During IBD w/u:

- MRE: *normal small/large intestine, however there was an incidental visualization of only the uncinate and a small portion of the pancreatic head most in keeping with complete dorsal pancreatic agenesis.*
- MRCP: *short pancreas, with pancreatic head and neck present and body and tail absent, with a nondilated remnant of the pancreatic duct, likely draining into the major papilla but the confluence with the common bile duct not well defined. The remnant pancreatic duct was nondilated measuring 2 mm.*
- Calprotectin: 209 mcg/gm, s/p Flagyl trial, f/u calprotectin : 438mcg/gm. Repeat EGD/Colonoscopy/Secretin Stimulation test: mild duodenitis, mild colitis ascending/cecum, active/chronic colitis rectum and sigmoid. Negative SST, Normal Fecal elastase, Normal Alpha-1-Atitripsine stool, Negative reducing substances stool. (+) P-ANCA/ASCA.

Dx with Mild Ulcerative Colitis, started on Rowasa 4gr pr/day x 4 weeks and Apriso 0.375mg tab 4tabs PO BID, repeat Calprotectin : 94 mcg/gr.

Currently on clinical and biochemical remission.

Treatment plan for this patient includes monitoring for signs /symptoms of malabsorption, diarrhea, weight gain, yearly alpha 1 antitrypsin in stool, fecal elastase yearly and MRCP every 1-2 years

CR-Diagnosis and Discussion

The formation of the pancreas is a relatively complex process, consisting of the generation of 2 pancreatic buds by the septum transversum, composed of ventral and dorsal endoderm at the foregut/midgut junction. The dorsal bud forms the upper part of the head, body and tail of the pancreas. The ventral bud forms the major part of the head and uncinate process It culminates at 6-7 weeks of gestation, with the fusion between the two. Complete agenesis of the pancreas and agenesis of the ventral pancreas are not compatible with life. In dorsal pancreatic agenesis, the body and tail are absent and the pancreatic bed anterior to splenic vein is filled with stomach and bowel loops described as dependent stomach and dependent intestine sign. The clinical presentation of dorsal pancreatic agenesis can range from complete absence of symptoms to non specific abdominal pain, diabetes mellitus, pancreatitis and sometimes exocrine pancreatic insufficiency. In the case of the patient we report, the symptoms he presented with are likely unrelated to the dorsal pancreatic agenesis and more related to an infectious or inflammatory process. There is no established guideline for treatment of this anomaly, and approach is made on a case to case basis.

CR-Conclusion and Significance

Dorsal agenesis of the pancreas is a rare entity, of which around 100 cases have been described. In the last decade, more cases have been found, likely due to improvement in diagnostic methods and increased availability of cross-sectional imaging. The most accurate imaging method that can accurately depict both the pancreatic ducts and parenchyma is MRI with MRCP, as was done with the patient in this case. It can present with a multiple array of symptoms, from none to severe pathology. Treatment guidelines are not available however a consensus found in multiple case reports, is that only symptomatic management is to be provided.

Abstract #37: (RES-017-CR)

Lopez Gonzalez MS*; Fonseca AG; Sunny J; Caicedo Oquendo LF; Felipez LM.

"Inflammatory Bowel Disease and Hidradenitis Suppurativa: Is the Association Real?".

CR-Introduction/ Background

Crohn's disease (CD) and ulcerative colitis (UC) are the two predominant types of chronic relapsing inflammatory disorders of the gastrointestinal tract, collectively referred to as inflammatory bowel disease (IBD). Accumulating evidence indicates that genetic susceptibility to dysregulated inflammatory reaction and altered microbiota may play crucial roles in the pathogenesis of IBD. Extra-intestinal manifestations (EIMs) include: musculoskeletal, hepatopancreatobiliary, ocular, renal, and pulmonary systems, as well as the skin. Different dermatological manifestations may arise during the course of IBD including pyoderma gangrenosum, psoriasis, Sweet's syndrome, aphthous stomatitis, hidradenitis suppurativa (HS), and erythema nodosum.

Hidradenitis suppurativa (HS) or acne inversa is defined as a chronic, inflammatory, recurrent, debilitating skin disease of the hair follicle that usually presents after puberty with painful, deep-seated, inflamed lesions in the apocrine gland–bearing areas of the body, most commonly the axillary, inguinal, and anogenital regions. The etiology is largely unclear, but dysregulated inflammatory response of cytokines, follicular occlusion, obstruction and dilatation of the pilosebaceous unit, and altered microbiota may be involved in the pathogenesis of HS.

CR-Case(s) Presentation(s)

We present a series of 5 cases seen in our institution with IBD and HS:

15 y/o Hispanic male with stenosing CD, with ileocolonic and upper GI involvement . Initially treated with 6 mercaptopurine (6MP) then switched to infliximab 5 mg/kg, presented with recurrent purulent lesion in his left axilla requiring 2 incisions and drainages (I/D) and multiple antibiotic courses. Resolution of symptoms after increase of infliximab dose to 10mg/kg. Last IFX level 12 ug/ml.
20 year old Hispanic male with ileocolonic CD anti-TNF non-responder, currently on ustekinumab 90 mg. Required multiple I/D for axillary and inguinal abscesses, and still has incomplete resolution of disease after optimizing biologics.
23 year old African-American female with CD, presented with skin lesions in the bilateral axillary and groin region. Lesions were small, firm, tender, dark brown papulonodules. Patient reported chronic history of similar lesions in the axillary, groin and perianal regions for which she previously had not received treatment. Started on adalimumab 40 mg weekly with subsequent improvement of skin lesions. Last ADA level was 31 ug/ml.
19 year old Hispanic male with ileocolonic CD on Infliximab 10 mg/kg, presented with skin lesion in right inguinal area, with purulent drainage, treated with multiple oral antibiotics. Improvement after changing treatment to ustekinumab 360mg IV and maintenance of 90 mg every 4 weeks .
14-year-old African American male with left sided UC with upper GI involvement, on adalimumab, presented with two right axillary circumferential lesions and one left axillary circumferential, open, and non-actively draining lesion. Biopsies confirmed hidradenitis suppurativa. Draining resolved but lesions persisted after I/D and doxycycline tx. ADA was increased to 80mg biweekly. ADA level 11.8 ug/ml with no antibodies. Non-draining lesions are present.

CR-Diagnosis and Discussion

HS diagnosis is based on the following clinical criteria: typical lesions, characteristic sites, chronic course and recurring flares. Our patients meet all of the criteria. All of them have different stages of the disease and had improvement or recurrences after aggressive treatment. HS shares common clinical manifestations, genetic susceptibility, and immunologic features with IBD. Both diseases have similar clinical manifestations in the skin and gut, characterized by sterile abscesses in the perineal and inguinal areas, scarring, and sinus tract formation. Smoking and obesity are known common risk factors for HS and IBD. None of our patients are smokers but 3/5 are overweight/obese.

4/5 of our patients with HS have CD, both characterized by scarring and sinus tract formation.

CR-Conclusion and Significance

Even though there appears to be an association between IBD and HS, their true relationship is still poorly understood. The involvement of certain cytokines, T helper type 17 cells, IL-23, and tumor necrosis factors (TNF) appears to be the key to the relationship. However, even when some patients' gastrointestinal disease is under control with monoclonal antibodies, HS seems to be harder to lead into remission. Obesity is a well characterized risk factor that most of our patients share. In our cases, Hispanic ethnicity tends to be a common factor as well. However, it is unclear if this is a true risk factor or a confounder secondary to our studied patient population. More studies between the association of IBD and HS are needed to understand better the pathogenesis and treatment modalities.

Abstract #38: (RES-018-CR)

Mandel G*; Riera Canales C; O'Farrell C; Fader M. ***"Methotrexate Use in End-Stage Renal Disease for Treatment of Osteosarcoma".***

CR-Introduction/ Background

Methotrexate is critical to many chemotherapy regimens, however it is eliminated renally and therefore in patients with renal failure there are concerns that it could lead to unacceptably high blood levels and consequently high toxicity.

CR-Case(s) Presentation(s)

A 17 year old female with a history of hemodialysis dependent lupus nephritis was diagnosed with localized osteoblastic type osteosarcoma of the right femur WHO grade II. Treatment per protocol AOST0331 with adjusted doses for renal failure was begun. Regimen began with two consecutive days of half-dose cisplatin (30 mg/m^2/dose) and regular dose doxorubicin (37.5 mg/m^2/dose) and dialysis began 24 hours after medications. On week 4 of therapy methotrexate was administered at 25% of protocol dose (3 g/m^2), she received daily hemodialysis, and cleared methotrexate at 120 hours. The frequent hemodialysis put her at risk for disequilibrium syndrome which was mitigated with intradialytic mannitol. She developed mucositis, necessitating intradialytic TPN and febrile neutropenia without growth in blood cultures.
For week 5 of therapy methotrexate was maintained and hemodialysis was begun 24 hours after MTX and continued every 12 hours and she cleared MTX at 108 hours, completing her first cycle.

Cycle 2 was delayed by 1 week due to mucositis and thrombocytopenia .Once begun she tolerated chemotherapy well. Methotrexate was cleared at ~110 hours for both weeks 9 and 10 of cycle 2. She subsequently developed febrile neutropenia, found to have HSV

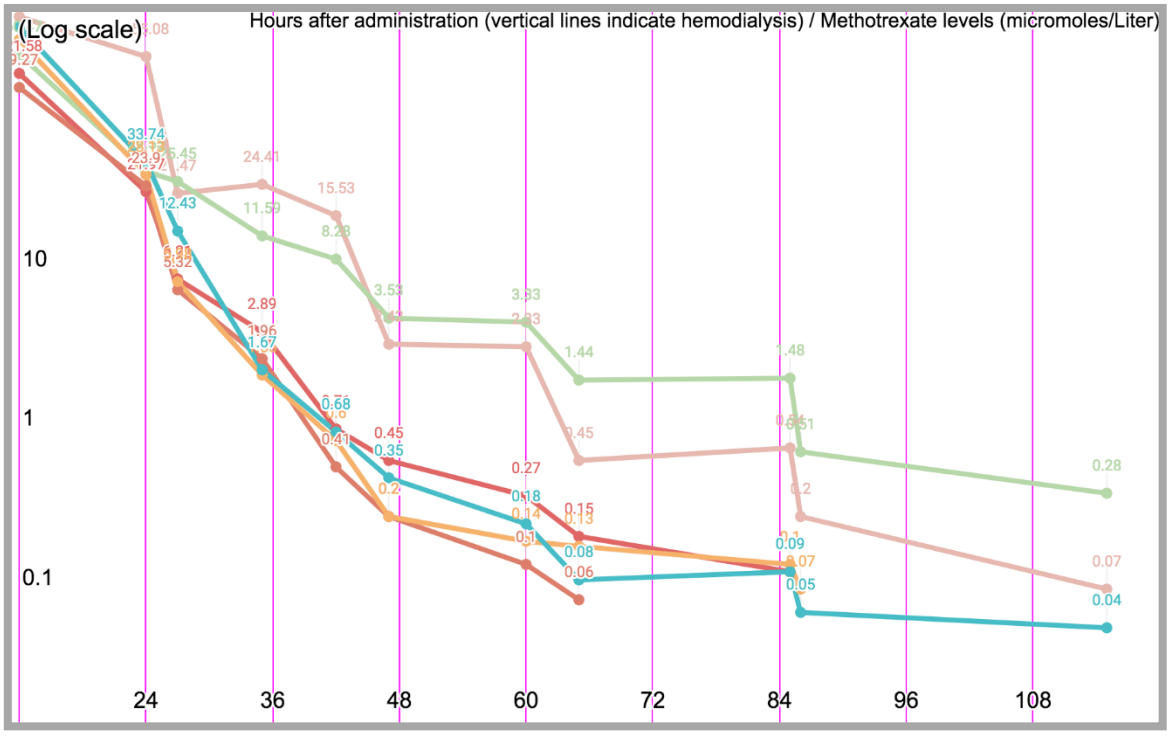
viremia and clostridium difficile colitis that resolved with antimicrobials. Limb salvage surgery was performed. Chemotherapy was then delayed due to development of pericardial effusion and persistent thrombocytopenia. Next course of cisplatin and doxorubicin were given and she subsequently developed febrile neutropenia and a pseudomonal skin infection. Methotrexate was given with hemodialysis every 12 hours and she cleared MTX after 114 hours. Her next dose was given with a similar regimen, however with a clearance goal of less than 0.01 micromoles/L due to previous toxicities, which she achieved at 148 hours.
Due to the extensive toxicities, the family elected not to continue chemotherapy. She has remained in remission upon 4 year follow up.

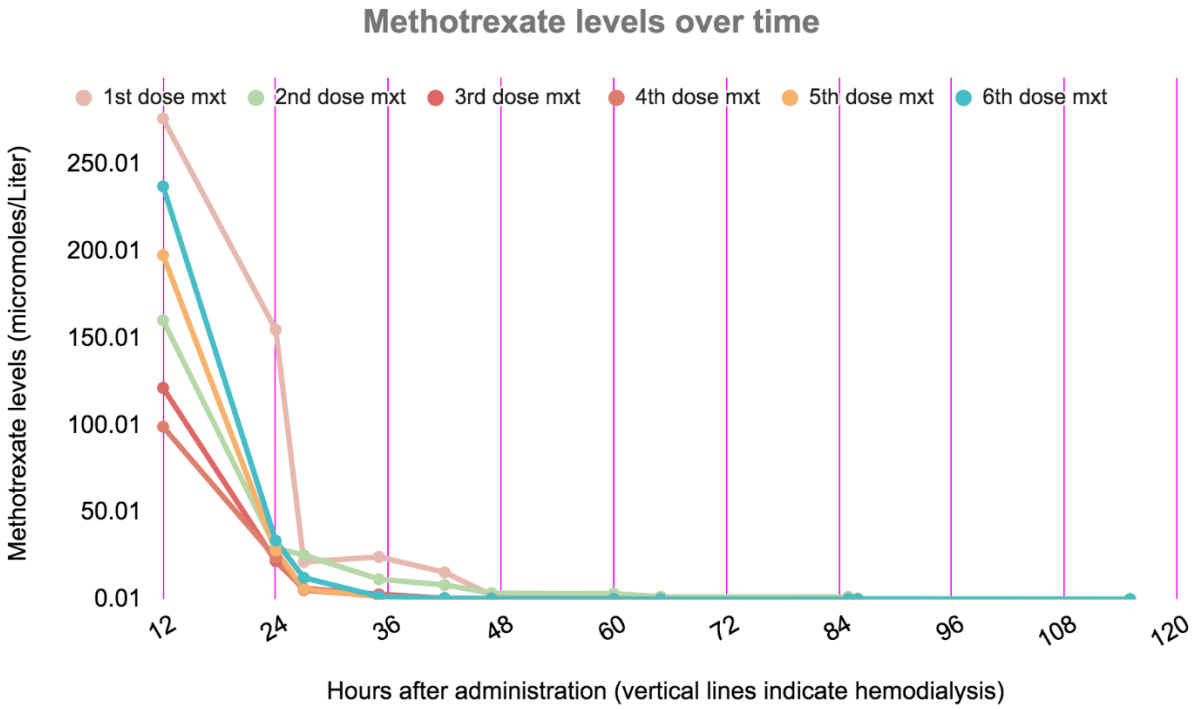
CR-Diagnosis and Discussion

Osteosarcoma is a malignant tumor of the osteoblastic cell lineage. Treatment protocols per COG include methotrexate and cisplatin, highly toxic medications that are cleared via the renal system. Methotrexate in OS patients Is administered as a single dose intravenously and methotrexate levels are measured by serial lab monitoring until clearance of drug, generally defined as <0.1 umoles/L. Published dosing recommendations recommend decreasing methotrexate by 75% and cisplatin by 50%. High flux hemodialysis is effective at removing methotrexate from circulation. Doxorubicin is excreted via the hepatic system and does not need to be corrected for in ESRD.

CR-Conclusion and Significance

Methotrexate can safely be used in renal failure with a multi-disciplinary approach.





Abstract #39: (RES-019-CR)

Mandel G*; Pelaez L; Hogan A; Riera Canales C; O'Farrell C; Fader M. ***"All that grows isn't malignant: The Hibernoma".***

CR-Introduction/ Background

Pediatric chest masses are a concerning finding. The differential is wide and a broad index of suspicion must be maintained. In this report we present a case of chest mass that resulted with benign course.

CR-Case(s) Presentation(s)

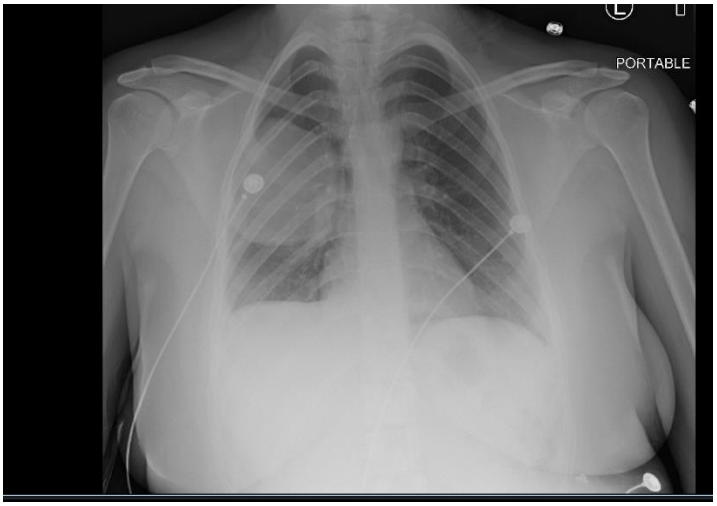
A previously healthy 15 year old female presented to hospital due to acute onset right sided chest pain, shortness of breath that progressed to episode of syncope. She was found on chest x-ray to have right lung mass concerning for Ewing sarcoma. CT revealed a large pleural mass in the right chest measuring 8.3 cm with posterior right pleural effusion. COVID PCR resulted positive. Ultrasound-guided biopsy was performed and resulted with coarsely granular to multi-vacuolated eosinophilic to pale cytoplasm and large mature univacuolated adipocytes, without atypia or mitosis. Immunohistochemical analysis was immunoreactive for S100, negative for Desmin, with presence of CD34 and CD31, highlighting a vascular network. These findings were consistent with hibernoma. Excisional surgery was performed and she did well with no recurrence of symptoms upon 2 month follow-up.

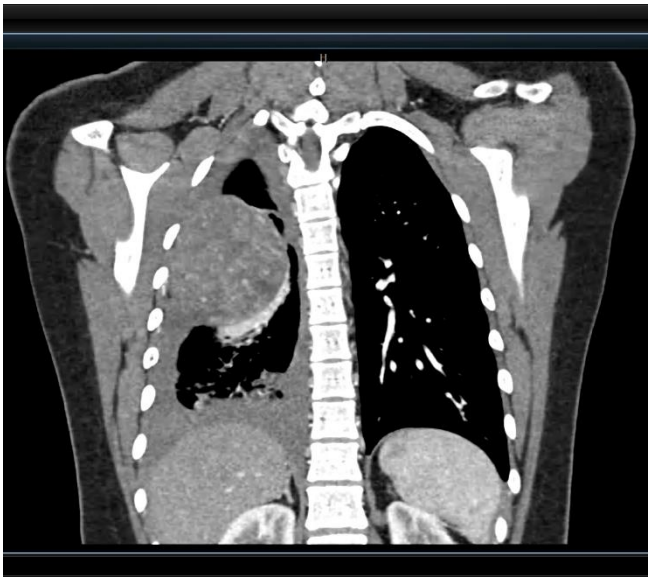
CR-Diagnosis and Discussion

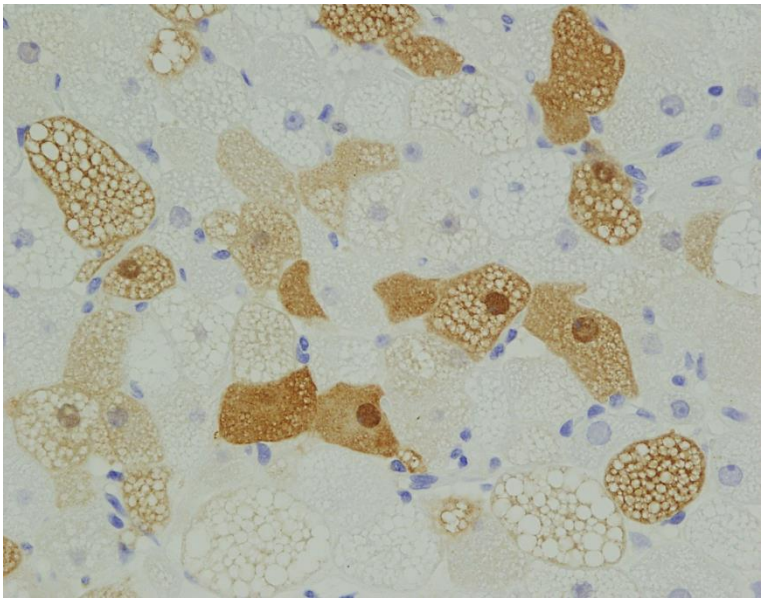
Chest masses in the pediatric population are a rare, concerning finding. They can be divided to congenital, infectious, neoplastic, inflammatory, and vascular causes. Neoplastic can be subdivided into benign vs malignant etiologies. Any mass, whether found incidentally or due to workup for symptoms deserves a closer evaluation. Hibernomas are rare, benign growths of neonatal fat tissue that can present in any period of life, but most frequently occur in the 3rd and 4th decades, about 5% of cases are seen in pediatric population, in children they have most often been found in the neck. There have been no reported cases of recurrence after excision.

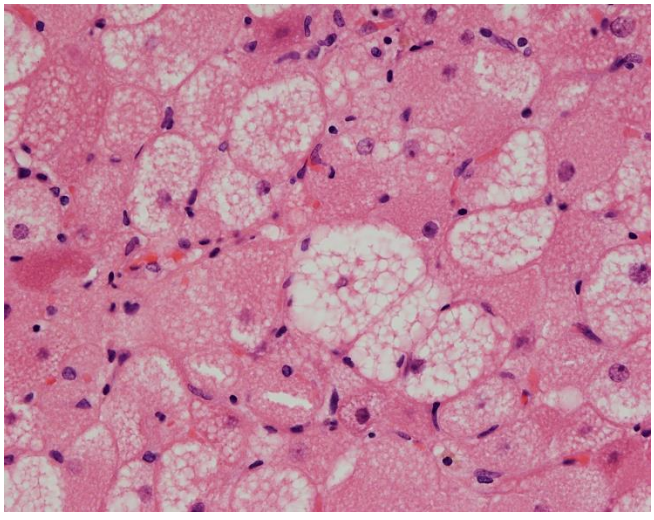
CR-Conclusion and Significance

Pediatric chest masses are most often secondary lesions or benign. Hibernoma is a rare cause of benign chest mass with no reported recurrence after excision.









Abstract #40: (RES-020-CR)

Napky Raudales P*; Barry R; Dantas M; Fader M. ***"More than meets the eye: Juvenile xanthogranuloma in a child with hyperlipidemia and bicytopenia".***

CR-Introduction/ Background

Juvenile xanthogranuloma (JXG) is a non-Langherhans cell histiocytosis of unknown incidence and pathogenesis. It is usually a self-limited cutaneous disease of childhood presenting with a single raised, rubbery lesion; cases of disseminated skin and systemic involvement (including bone marrow) have been described in the literature. Although rare, an association between JXG and certain blood disorders (such as juvenile myelomonocytic leukemia, acute lymphoblastic leukemia, autoimmune lymphoproliferative disorder, and myelodysplastic syndrome) has been reported.

CR-Case(s) Presentation(s)

Our patient is a 6-year-old female with past medical history of hyperlipidemia, referred by her pediatrician due to an abnormal routine CBC showing anemia (8.9 g/dl), thrombocytopenia (13,000/µl), and giant cells on PBS. Mother reported that anemia and thrombocytopenia had been noted during her WIC appointments. Denied fever, weight loss, bone pain, easy bruising, or bleeding. Initial exam revealed multiple soft nodules on bilateral auricles, elbows, and knees, previously attributed to her hyperlipidemia, but was otherwise unremarkable.

CR-Diagnosis and Discussion

Institutional CBC confirmed bicytopenia. Reticulocyte count (2.01%), triglycerides (276 mg/dl), and LDL cholesterol (222 mg/dl) were elevated. Infectious workup, iron studies, thyroid function tests, and Hemoglobin electrophoresis were unremarkable; lupus, acute leukemia, and FISH MDS panels were negative. Bone marrow aspiration (BMA) showed occasional histiocytes with phagocytosis of fragmented cells, but no evidence of malignancy. Chromosome analysis of the bone marrow was unremarkable. Punch biopsy of the nodules showed foamy histiocytes (CD4, CD68, ALK, and FXIIIA positive, and S100, CD1a, and langerin negative) and scattered Touton cells with extensive fibrosis consistent with JXG. Although a specific trigger for her bicytopenia was not identified, she remained asymptomatic and with improved cell counts during the hospital course.

CR-Conclusion and Significance

The presence of hyperlipidemia can make the distinction between JXG and tuberous xanthomas challenging. Although the diagnosis is usually made clinically, biopsy can be used to distinguish them (with the latter lacking Touton cells). Our patient had bicytopenia, giant cells on PBS, and hemophagocytosis on BMA, suggestive of possible bone marrow involvement. We highlight the importance of performing a thorough workup including BMA when abnormal blood cell counts are found in patients with JXG, as cases of bone marrow involvement and other associated blood dyscrasias have been reported.

Abstract #41: (RES-021-CR)

Napky Raudales P*; Barry R; Shariati F; Sanchez E; Fonseca A; Niazi T; Maher O; Khatib Z. ***"Acute psychosis as an initial manifestation of a pineal germinoma in an adolescent male".***

CR-Introduction/ Background

Intracranial germ cell tumors can be classified as either germinomas or non-germinomatous germ cell tumors. Although psychiatric symptoms are known manifestations of central nervous system tumors, very few cases have been described in pineal germinomas, more so in the pediatric population. These symptoms can be present at onset, during the course or after completion of treatment. Proposed mechanisms include mass effect from the tumor itself or from the secondary obstructive hydrocephalus, presence of anti-brain antibodies, circadian rhythm disorders, and medication induced psychosis from hormone replacement therapy.

CR-Case(s) Presentation(s)

Our patient is a previously healthy 19-year-old male who presented with a 1-month history of behavioral changes (auditory and visual hallucinations, disorientation to place, and insomnia), with poor response to outpatient psychiatric management. Initial exam revealed flat affect, confused conversation, repetition, disorientation to time and place, and Parinaud syndrome.

CR-Diagnosis and Discussion

Neuroimaging revealed a pineal mass with associated hydrocephalus for which he underwent external ventricular drain and subsequent ventriculo-peritoneal shunt placement. Biopsy confirmed diagnosis of pineal germinoma (CD117, PLAP, and Oct 3/4 positive, hCG, AFP, and CD30 negative, and a Ki-67 high proliferation index). He underwent chemotherapy with carboplatin and etoposide per protocol ACNS 1123, and after 2 cycles had significant decrease in tumor size, and resolution of his psychiatric symptoms. He completed a total of 4 cycles of chemotherapy, followed by whole ventricular proton radiation; to date he has had no recurrence in his symptoms.

CR-Conclusion and Significance

Psychiatric symptoms may be the only presenting feature of a pineal germinoma in the pediatric population. A thorough history, physical exam, and high index of suspicion are required for early diagnosis and intervention. Imaging should be considered in patients with focal neurologic findings or atypical psychiatric symptoms. The timeline of symptom onset (initial presentation) and the fact that our patient's symptoms improved after initiation of treatment and decrease in mass size, suggest these behavioral changes were likely related to the tumor itself. Further studies are required to determine the exact mechanism by which pineal germinomas produce psychiatric symptoms, and could

provide insight into the pathophysiology and treatment opportunities for psychiatric disorders.

Abstract #42: (RES-022-CR)
Navarro R*; Etinger V. ***"Don't be Fooled! The Many Faces of Abdominal Pain".***

CR-Introduction/ Background

Acute abdominal pain is one of the most common complaints encountered by pediatricians and it accounts for 5-10 % of visits to the emergency department.[1] It can be caused by a variety of conditions ranging from mild to more serious diseases. Although most children with acute abdominal pain have self-limited benign conditions, pain may be a manifestation of an urgent surgical or medical condition. An early and accurate diagnosis results in a more appropriate management and, subsequently, leads to better outcomes and lower risk of morbidity.

We present a case of abdominal pain in a 6-year-old previously healthy girl admitted for treatment of constipation.

CR-Case(s) Presentation(s)

A 6-year-old previously healthy girl presented to the urgent care for acute onset abdominal pain, which started four hours prior to arrival. Pain was localized to the left lower quadrant and constant, worse with meals, and had no associated relieving factors. Last bowel movement was on the day prior to presentation. Bowel movements are non-bloody, non-mucosy, and described as a Bristol type 1. Mother reported she becomes occasionally constipated but usually passes bowel movements every day or every other day. Other than one week of poor appetite, the patient has not had any recent weight loss, fatigue, chills, fevers, emesis, diarrhea, rashes, urinary complaints, or recent upper respiratory tract infections. No recent travels, sick contacts, or change in diet. No surgeries, active medications, or drug allergies. She was born full term via vaginal delivery with no complications.

On physical exam she had vital signs within normal limits. She was noted to have tenderness to palpation in the left lower quadrant and increased firmness that was attributed to stool burden. Xray of the abdomen revealed moderate stool burden with no obstruction or fecal impaction. A children's enema was administered, and the patient passed two large bowel movements. After a PO trial, she was discharged home with instructions to begin a stool softener.

The following day, the patient presented to our emergency department due to persistent abdominal pain and new onset emesis. Abdominal pain continued to be localized to the left lower quadrant but was now described as crampy, intermittent, with episodes of hip

flexion. Upon exam, she had diffuse tenderness to palpation of the lower abdomen, including the right lower quadrant. An ultrasound of the appendix was obtained that poorly visualized the appendix. An abdominal X-ray showed moderate stool but no free air, obstruction, or fecal impaction. Labs including a CMP, amylase, lipase, and urine analysis were all normal. TSH was low at 0.4 with a normal free T4 consistent with subclinical hyperthyroidism but the patient denied any symptoms of rapid or irregular heartbeat, sweating, or irritability. She was given a second children's enema with failure to pass stool. Due to persistent abdominal pain and failure to respond to treatment, patient was admitted overnight for a bowel cleanout with polyethylene glycol.

Upon evaluation the following morning, mom reported the patient was unable to sleep overnight due to severe and recurrent episodes of abdominal. One of those pain episodes were witnessed during morning rounds prompting further evaluation with an urgent CT scan of the abdomen and pelvis that revealed a heterogenous mass with cystic components, fat, and calcifications in the pelvis consistent with a teratoma causing mass effect upon the sigmoid colon. Pediatric surgery was immediately consulted who recommended evaluating for co-existing ovarian torsion with an ultrasound of the pelvis and obtaining tumor markers. Ultrasound showed a normal uterus and right ovary but was unable to rule out left ovarian torsion. Tumor markers (AFP, Uric acid, B-hcg, CA-125, LDH, and Inhibin B) all returned within normal limits, making malignancy less likely. She was taken to the operating room that same day where she underwent laparoscopic cystectomy and detorsion of the left ovary. Pathology report confirmed the diagnosis of a mature ovarian teratoma. The patient was discharged the following day in stable condition.

CR-Diagnosis and Discussion

Ovarian tumors are extremely rare in children. A study revealed that about 9.6% of all ovarian masses are diagnosed between the ages of 0-19 years old. Of those diagnosed with ovarian mass, 65.7% were found to have a germ cell tumor, the most common being mature teratoma.[2] Ovarian teratomas are usually asymptomatic up until they start causing mass effect upon other organs. Patient's may also present with symptoms of ovarian torsion, as our patient did, or frank peritonitis from acute rupture and hemorrhaging. Definitive diagnosis is made using pathology but initial imaging modalities such as pelvic ultrasound are used to aid in the diagnosis. Obtaining tumor markers to rule out malignancy is necessary to begin proper treatment. Surgical resection of the mass with preservation of the ovary is usually suffice and results in complete resolution. If torsion is present, detorsion or removal of the ovary may also be necessary depending on the viability of the ovary during surgery. Prognosis for these patients is excellent and reoccurrence is rare.

Ovarian torsion is an important differential diagnosis to keep in mind during the work up of a child presenting with acute onset abdominal pain. Given the importance of early surgical intervention in order to restore ovarian blood flow and ensure ovarian

preservation, it is vital that physicians consider this diagnosis early on. Ovarian torsion most commonly occurs in women of childbearing age, with a peak incidence at around the second decade of life. Prevalence in the pediatric population is about 4.7 per 100,000 females.[3] Typically, patient's present with acute onset moderate to severe pelvic pain, nausea, and emesis. Pelvic ultrasound with doppler is used to aid in the diagnosis. It is important to note that the presence of blood flow on doppler does not rule out ovarian torsion as there can be only partial vascular obstruction. Diagnosis is clinical and treatment requires immediate detorsion. Removal of the affected ovary is determined during surgery.

Although the admitting diagnosis was acute constipation the patient's inability to pass bowel movements and her colicky abdominal pain were secondary to sigmoid compression from a left ovarian teratoma, complicated by ovarian torsion.

This case serves as an opportunity to bring awareness of cognitive biases and how these biases can lead to medical errors. Anchoring bias, which is described as focusing on one particular piece of information or symptom early in the diagnostic process and failing to make adjustments, is one of the most common biases amongst physicians. In this patient's case, the focus given to acute constipation as the cause of the acute abdominal pain almost led to a delay in diagnosis and surgical intervention, which could have increased the chance of complications and ultimately compromise future fertility. Two strategies that have proven effective in combatting these biases are reflection (in which a medical provider engages in discussion and thoughtful reasoning with a mentor) and cognitive forcing (conscious consideration of other diagnosis by taking a systematic approach with every case).

CR-Conclusion and Significance

This case serves as an example of the importance of keeping a high index of suspicion for ovarian torsion in female children presenting with acute abdominal pain and the importance of adjusting medical management based on clinical response and information gathered during each part of the medical encounter. It also describes an uncommon gynecological tumor presenting with a very common chief complaint in the pediatric population.

Abstract #43: (RES-023-CR)

Riera-Canales C*; O'Farrell C; Mandel G; Abdella H. ***"Late relapse in Wilms Tumor: Does intense chemotherapy improve survival?".***

CR-Introduction/ Background

Wilms tumor (WT) is the most common primary pediatric kidney cancer. Five year survival approaches 90%. High-risk features include: age >2 years, higher stage, poor differentiation, anaplasia, and molecular findings like loss of heterozygosity in

chromosome 16p, 1p, and 11p15. Relapse occurs in 50% of these high-risk patients, with most common sites being the lungs, original tumor bed, and liver. Among those who relapse, 95% do so within two years of initial diagnosis. Late recurrence (LR) is a relapse after five years remission and is extremely rare for patients with WT, with only a few cases reported. Due to the rarity, there are no current treatment recommendations. (107)

CR-Case(s) Presentation(s)

Describe the case of an eighteen-year-old male diagnosed at three-years-old with stage IV WT who relapsed fifteen years after diagnosis. (20)

CR-Diagnosis and Discussion

Eighteen-year-old male, diagnosed at three-years-old with stage IV WT (liver and lung metastasis). Pulmonary nodule biopsy showed favorable histology, and received neoadjuvant therapy for ten weeks under NWTS Regimen DD4A with vincristine + doxorubicin. Then, he underwent left nephrectomy, partial hepatectomy, and lymph node biopsies. Pathology showed WT with rhabdomyomatous differentiation and diffuse anaplasia, with tumor extension into the renal sinus, but no lymph node or liver margin infiltration. Due to unfavorable histology, treatment was changed to NWTS5 relapse protocol stratum C alternating cyclophosphamide/carboplatin with etoposide, plus total lung irradiation. End-of-treatment scans showed subcentimeter bilateral pulmonary nodules, which remained stable for several years. Surveillance was followed per NWTS5 protocol. He remained disease-free for 15 years, until he presented with a two-week history of progressive shortness of breath. CT showed several lung nodules, left-sided complex pleural effusion, and possible liver metastasis. Pulmonary nodule biopsy confirmed recurrence of WT with rhabdomyomatous differentiation but no anaplasia. He has received therapy alternating between ICE, vincristine + irinotecan, and vincristine + cyclophosphamide + doxorubicin. PET scan after 7 cycles showed decreased disease burden, yet still significant pleural-based nodules, making future gross total resection challenging. (189)

CR-Conclusion and Significance

LR in WT is rare. Primary high-risk patients receive more aggressive chemotherapy. Since time to relapse is prolonged, it is unclear if treatment with similar previous drugs would be effective or if typical more intense relapse regimen is necessary. Local control can also be challenging as utility of radiation is limited when many received radiation previously. Further research is needed to identify and improve treatment options for these patients. (69)

Abstract #44: (RES-024-CR)

Saini A*; Kana SL; Visona C; Diaz A; Jayakar P. ***"Hypermethioninemia due to Adenosine Kinase Deficiency: Novel Mutations in ADK Gene diagnosed by Rapid Whole Genome Sequencing"*.**

CR-Introduction/ Background

Adenosine kinase (ADK) deficiency is a rare autosomal recessive metabolic disorder of methionine metabolism due to variants in *ADK* (OMIM: #614300). Adenosine is synthesized during hydrolysis of S-adenosylhomocysteine (AdoHcy). It is phosphorylated to adenosine monophosphate (AMP) by the enzyme adenosine kinase, the main pathway of adenosine metabolism. As a result, deficiency of ADK causes accumulation of AdoHcy, S-adenosylmethionine (AdoMet) and methionine, and results in inhibition of methylation reactions. *ADK* is located at chromosome 10q22.2.

CR-Case(s) Presentation(s)

In this report, we describe a patient with two novel pathogenic mutations in the *ADK* gene. She presented at three months of age with persistent cholestatic jaundice and elevated liver enzymes, including gamma-glutamyltransferase (GGT). She was born at full term to non-consanguineous Hispanic descent parents with an unremarkable family history. Birth weight was 3.97 kg (93rd %), and the length was 53.3 cm (98th %). The physical exam showed frontal bossing, sparse hair, and macrocephaly. Hypotonia and delayed motor development were observed at seven months of age. In addition, she had asymptomatic hypoglycemia. The ocular exam was normal.

The biochemical evaluation showed evidence of hepatitis, cholestasis, and mildly deranged coagulation profile, but normal plasma carnitine, very-long-chain fatty acids, acylcarnitine profile, karyotype, and microarray. Plasma amino acids showed methionine levels of 1236 µmol/L (NL 12-50 µmol/L) and homocysteine levels of 13.8 µmol/L (NL <10.4 µmol/L). Imaging of the liver demonstrated diffuse hypoechoic echotexture with increased periportal echogenicity suggestive of hepatitis. Echocardiogram and ultrasound of the gallbladder and kidneys were normal. Liver biopsy showed signs of chronic active hepatitis with confluent lobular necrosis and extensive fibrosis. An exhaustive infectious workup was negative.

CR-Diagnosis and Discussion

Initial genetic evaluation with a cholestasis panel was positive for heterozygous variants of *PKHD1* (pathogenic) and *PEX6* (unknown significance), implicated in autosomal recessive polycystic kidney disease and peroxisome biosynthetic defects, respectively. Since the patient was heterozygous, and the clinical phenotype did not corroborate with either condition, rapid whole genome sequencing (rWGS) was performed. Two heterozygous, novel and pathogenic mutations were identified in *ADK*. These include a paternally-inherited deletion, c.642_645del (p.Ser215HisfsTer12), in exon 8 of 12 and a maternally-inherited c.916C>T (p.Gln306Ter) variant in exon 11. Both variants result in

a premature stop codon, which is predicted to result in loss of normal protein function through protein truncation or nonsense-mediated mRNA decay, and were confirmed by Sanger sequencing.

CR-Conclusion and Significance

ADK deficiency is a rare metabolic condition with only 20 cases reported in the literature. The presenting symptoms are cholestatic jaundice and hepatitis with subsequent global developmental delay, early-onset seizures, hypotonia, and hyperinsulinemic hypoglycemia. Nonspecific dysmorphic features can be present as well. Eleven pathogenic variants of the *ADK* gene have been reported to date. Our patient was heterozygous for two pathogenic variants that have not been previously described in the literature. Therapeutic options include a methionine-restricted diet and diazoxide for hypoglycemia, and both were utilized in our patient. After initiating dietary treatment with low methionine medical formula, methionine levels decreased to 286 μmol/L (NL 12-45 μmol/L), and homocysteine levels normalized. Liver functions and hepatic echotexture improved as well. There was no immediate change in hypotonia and motor developmental delay on short-term follow-up. ADK deficiency has only been recently identified, and the clinical phenotype mimics that of numerous other conditions. Therefore, it should be considered in the differential diagnosis of infants presenting with cholestatic jaundice, hypoglycemia, developmental delay, hypotonia, and epilepsy to avoid delayed diagnosis. Rapid whole-genome sequencing enables prompt diagnosis and the institution of appropriate treatment.

Abstract #45: (RES-025-CR)

Saini A*; Almasarweh S; Acosta S; Sasaki J. ***"Microphthalmia Syndrome 9: Novel mutations in STRA6 Gene".***

CR-Introduction/ Background

The association of Anophthalmia/Microphthalmia, Pulmonary agenesis/hypoplasia, Cardiac defects, and Diaphragmatic hernia/eventration is known as Matthew Wood syndrome/ PDAC syndrome or, Syndromic Microphthalmia 9 (MCOPS9, OMIM 601186). It presents in the neonatal period as severe & refractory pulmonary hypertension with an unfavorable prognosis.

CR-Case(s) Presentation(s)

We describe a female neonate, born at 40-week gestation in a non-consanguineous family to a 30-year-old primigravida mother. Birth weight was 3.0 kg (11th %), length 49.5 cm (41st %), and head circumference 33.5 cm (14th %). The patient had bilateral anophthalmia on physical exam. Echocardiogram demonstrated levocardia, an anomalous venous connection of the left pulmonary veins to the persistent left superior vena cava, atrial septal defect, dilated main pulmonary artery with severely hypoplastic branch pulmonary arteries, large patent ductus arteriosus, and mild hypoplasia of distal transverse arch and aortic isthmus. Cardiac CT showed hypoplastic right lung, absent

right upper lobe bronchus, and severely hypoplastic branch pulmonary arteries (Z score < -2.5). Malrotation of the right kidney and uterus didelphys were also present. The patient died on day 12 of life from complications of severe pulmonary hypertension.

CR-Diagnosis and Discussion

MCOPS 9 is a rare and fatal condition. Hypoplasia of lungs and pulmonary vessels entails severe pulmonary hypertension posing considerable challenges in management. In addition to ocular, pulmonary, and cardiac affection, malformations affecting the kidneys, brain, gastrointestinal, and genitourinary systems are frequently seen. The condition is inherited as an autosomal recessive condition caused by mutations in STRA6 (Signaling Receptor and Transporter of Retinol) gene on chromosome 15. We identified two novel pathogenic mutations in the STRA 6 gene: 1) paternally inherited c.1418-6T>A, which introduces an abnormal splice site upstream from intron 17, 2) maternally inherited frameshift mutation c. 464del, which generates a premature stop codon in exon 9a. Of the fifty cases with this condition, only seven alive patients have been reported to date, thus highlighting the prognostic significance of this condition.

CR-Conclusion and Significance

Ocular abnormalities should prompt evaluation for congenital cardiac and pulmonary defects. Rapid genome sequencing is a robust tool to guide prognosis and management.

Abstract #46: (RES-026-CR)

Sanchez-Solano N*; Laufer M; Della Volpe A. ***"Unexpected Case of Pulmonary Nodules"*.**

CR-Introduction/ Background

Whipple's disease (WD), a rare and hard to diagnose illness, is caused by the *tropheryma whipplei (TW)*, an intracellular organism that was successfully isolated in the early 1990s with developing PCR techniques. Although usually presenting in its classical form with gastrointestinal involvement, acute localized forms such as pneumonia/pulmonary nodules have also been described, posing a harder diagnostic challenge. We present a case of a patient with history of Systemic Lupus erythematous, presenting with cough and dyspnea, found to have pulmonary nodules.

CR-Case(s) Presentation(s)

A 16-year-old Vietnamese female, with history of Systemic lupus erythematous (SLE) on cyclosporin, presented with 1 week of dysphagia, mouth ulcers, subjective fever, maculopapular rash, cough and new onset dyspnea. Physical exam remarkable for tachycardia, tachypnea, fever (103F), mouth ulcer, and bibasilar hypoventilation. Diagnostic workup showed CBC with leukopenia (3.5), and thrombocytopenia (149). Elevated ESR (42) with normal CRP (< 0.5), low C3/C4 levels with elevated dsDNA consistent with lupus flare. Respiratory pathogen panel negative. Unremarkable EKG and echocardiogram. Chest x-ray showed bibasilar airspace and nodular opacities and small bilateral pleural effusions. She was started on methylprednisolone, for suspected lupus

flare, as well as ceftriaxone for multifocal pneumonia. Chest computed tomography (CT) revealed confluent infiltrates with diffuse multiple round opacities, as well as hilar lymphadenopathy. CT angiogram negative for pulmonary embolism. Infectious workup remarkable for Aspergillus, galactomannan, and T2 Candida panel, PCP PCR, chlamydia pneumoniae/trachomatis titers, mycoplasma, HSV, sputum, urine and blood culture all negative. Bronchoscopy was performed for bronchoalveolar lavage (BAL) with karius, and fungal culture negative. However, Bacterial 16srDNA positive for TW and Acid fast bacilli culture positive for nontuberculous mycobacteria (NTM). She completed 26 days of ceftriaxone and then switched to oral Bactrim, with clinical improvement and resolution of fever and dyspnea.

CR-Diagnosis and Discussion

Although TW poses a diagnostic challenge, it can be eradicated with appropriate antibiotic therapy. Given new PCR techniques its prevalence has been increasing, allowing for expansion of knowledge of its pathogenicity. Not only does it presented with chronic gastrointestinal involvement, but can also present in isolated forms such as encephalitis, endocarditis, bacteremia, and pneumonia/pulmonary nodules. Host factors need to be taken into consideration for predisposition of infection such as genetic and immunologic, given immunosuppressed state has been associated with increased risk for infection. However, multiple studies have also shown that there can be a chronic carrier state. More research is needed to characterize the spectrum of this disease, diagnosis and management.

CR-Conclusion and Significance

More research is needed to characterize the spectrum of this disease, diagnosis and management.

Abstract #47: (RES-027-CR)

Smith J*; Behnam-Terneus M. ***"Rub-a-dub-dub Pathogens In The Tub".***

CR-Introduction/ Background

CR-Case(s) Presentation(s)

Patient is a 19-year-old male college football player with a history of ulcerative colitis, on azathioprine and mesalamine, who presented with 4 days of fever of 102.3F, fatigue and generalized myalgia. Seen at an urgent care where he tested negative for flu and strep and discharged on amoxicillin. Patient reported a persistence of symptoms with fever of 104F and presented to an OSH. He was flu negative but sent home on Tamiflu. Later that week, he came to our ED and was found to have WBC of 14.7, CRP 15.6 mg/dL and

total CK 478. He was diagnosed with myositis in the setting of a viral illness and sent home on naproxen. He returned to the ED with persistent fever and a new productive cough. He had associated NBNB vomiting, sweating and chills. Denied diarrhea, constipation or abdominal pain. He disclosed travel to Pittsburgh to play in a football game and used communal showers. He had non-labored respirations, with crackles of the right lower lung field. Patient had significant chest wall tenderness, but no pleuritic chest pain or wheezing. He also had malaise and weakness.

CR-Diagnosis and Discussion

Admission labs showed a WBC 12.6, CRP now 32.8, CMP normal and CK 255. CXR showed a complicated right lower lobe pneumonia with a small pleural effusion. Despite broad-spectrum treatment of complicated CAP, he developed hemoptysis. Repeat CXR showed worsening pneumonia and effusion. Further work up with Legionella Ag, Urine, Ab, and sputum, C. pneumoniae, C. psittaci, and C. trachomatis, Mycobacterium and acid-fast bacilli were ordered. Legionella urine Ag, acid-fast culture, and respiratory lower culture with Gram stain were negative. Legionella pneumonia antibody was found to be positive. PFTs obtained which showed early obstructive pulmonary impairment. The patient was diagnosed with legionnaire's disease with a positive antibody test in the setting of worsening complex pneumonia, non-responsive to empiric antibody therapy.

CR-Conclusion and Significance

Legionnaire's is a very rare cause of interstitial pneumonia, affecting around 10-15 people per million, seldom in the pediatric population. It is caused by Legionella pneumophila and found in artificial bodies of water including air conditioning units and communal showers. Infection occurs via inhalation of aerosols. Immunosuppressed individuals are at higher risk. Our patient displayed the classical course of disease including rapidly deteriorating clinical status within the first week past incubation. Correct identification and treatment are pertinent as the death rate in immunosuppressed patients can be as high at 80%.

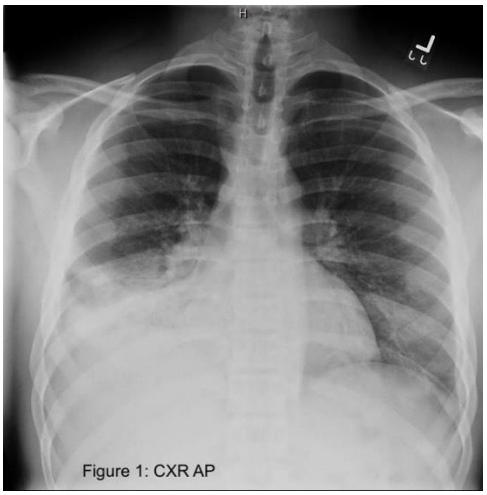


Figure 1: CXR AP

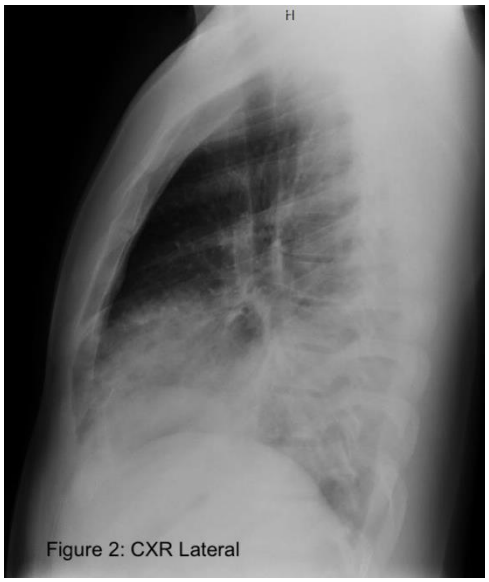


Figure 2: CXR Lateral

Figures 1 & 2: CXR AP and Lateral show a complicated right lower lobe pneumonia with small associated pleural effusion.

Abstract #48: (RES-028-CR)**

Smith J*; Behnam-Terneus M. ***"Vaping Associated Lung Injury: A Case of Rapid Respiratory Decompensation".***

CR-Introduction/ Background

Vaping and E-Cigarette use is rapidly becoming the next avoidable health crisis among the pediatric population. In 2018, 25% of 12th graders were using vaping products. Teens who vape are 3.5 times more likely to start conventionally smoking, leading to addiction and long-term health consequences. Moreover, E-Cigarette or Vaping Associated Lung Injury (EVALI) and subsequent ARDS is on the rise. We present a case of a 16-year-old male diagnosed with vaping induced lung injury after the insidious onset of systemic symptoms and ultimately respiratory distress.

CR-Case(s) Presentation(s)

Patient is a 16-year-old male with a history of increased liver enzymes, presenting with 2-weeks of cough and diffuse pleuritic chest pain. Symptoms progressed to exercise intolerance, increased dyspnea, decreased oral intake, weakness, and a 10lb weight loss with vertigo. He presented to the ED with altered consciousness, fever of 104F, congestion and diarrhea for two days. He endorsed 2-3 drinks of hard liquor per month, LSD use one month ago and smoking marijuana 1-3 times per week. Patient vaped 4-5 times per week, both nicotine and THC cartridges. On presentation he had a respiratory rate of 18, saturating 100% on room air. Physical exam was significant for decreased air entry bilaterally with no lung sounds from the mid to lung bases. With further clinical progression he was tachypneic at 31 breaths per minute and displayed increased work of breathing. He was subsequently transferred to the PICU due to the concern for acute

respiratory decompensation. His blood venous gas displayed respiratory acidosis, pH 7.25, PCO2 51.7 and HCO3 22.7. CBC showed a WBC of 18.9 10K/uL, BMP and procalcitonin were normal, CRP was significantly elevated at 21.6 mg/dl. His respiratory pathogen panel and mycoplasma antigen were negative. Patient had a negative blood culture, HIV was non-reactive, gonorrhea and chlamydia negative. Urine toxicology was positive for marijuana. CXR indicated left lower lobe consolidation (Figure 1). Ultimately, he did not need intubation, but was monitored in the PICU and discharged after respiratory stabilization.

CR-Diagnosis and Discussion

The patient was diagnosed with E-Cigarette or Vaping associated Lung Injury (EVALI) vs. community acquired pneumonia (CAP). Common pathogens were ruled out, while his clinical appearance was not fully consistent with CAP. Respiratory symptoms in conjunction with systemic findings on initial presentation fit the diagnosis of vaping related lung injury. Physical exam corroborated the diagnostic results, displaying significantly decreased bilateral lung sounds and rapidly deteriorating respiratory status. Blood gas indicated poor gas exchange.

CR-Conclusion and Significance

Vaping related lung injury is a novel diagnosis affecting the pulmonary parenchyma. The scientific community has yet to elucidate the pathophysiology behind injury, although many studies point to acute lung injury second to lipoid pneumonia or vitamin E acetate inhalation. There has been a surge of cases within the last year leading to ARDS and ultimately respiratory failure. Inclusion criteria are the use of vaping related products in the last 90 days, lung opacities on either CXR or CT and the exclusion of lung infections. This was consistent with our patients' presentation.

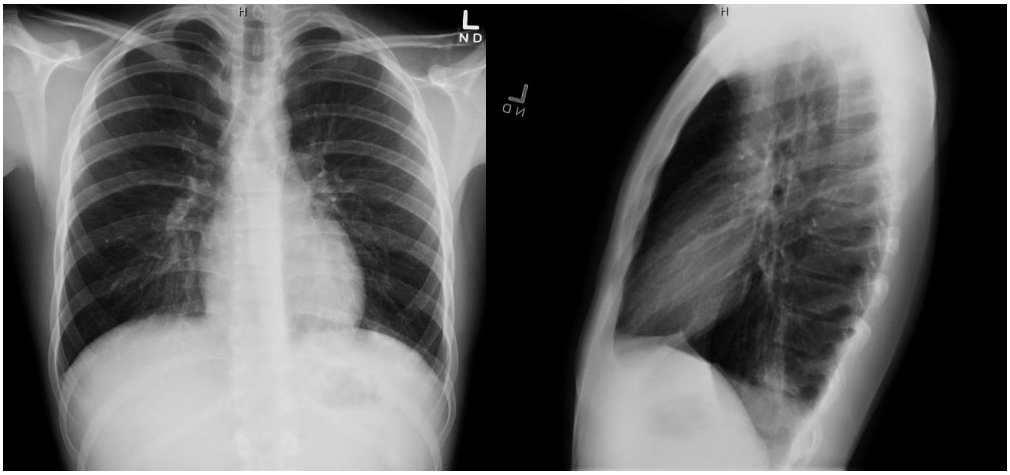


Figure 1: CXR AP and lateral indicating left lower lobe consolidation.

Abstract #49: (RES-029-CR)

Smith J*; Saini A; Brathwaite C; Koyfman S. ***"A Case of Refractory Diarrhea and Malabsorption Secondary to Autoimmune Enteropathy in a 10-Year-Old Child".***

CR-Introduction/ Background

Autoimmune enteropathy (AE) is a rare condition characterized by refractory diarrhea, histologic changes and extraintestinal manifestations. The diagnostic criteria include; intractable diarrhea, villous atrophy and autoantibodies without known immunodeficiency. The presence of autoantibodies such as anti-enterocyte and anti-goblet cell antibodies are supportive of diagnosis but not pathognomonic. In the pediatric population AE is rare, affecting infants up to 6 m/o with an incidence less then 1/100000 children. We describe a case of AE, elucidated by treatment refractory diarrhea in a 10 y/o.

CR-Case(s) Presentation(s)

10 y/o Bahamian male with a PMH of eczema and food allergies presenting with periumbilical pain, NBNB vomiting and profuse non-bloody diarrhea for 6 weeks, diagnosed with Crohn's disease (CD). Pre-arrival course included a 2-week hospitalization with treatment for malnutrition, diarrhea and hypokalemia. CT abdomen showed mural thickening of the rectum, sigmoid and descending colon, with mesenteric lymphadenopathy. EGD/colonoscopy were remarkable for thickened nodular folds of the duodenum and diffuse erythema of the rectum without ulcers. Biopsy indicated chronic active inflammation with cryptitis and non-caseating granulomas in the small intestine. Colonic mucosa noted crypt destruction and abscess, with no granuloma. MRE noted pancolitis, terminal ileitis and areas of active inflammation in the jejunum. Capsule endoscopy revealed terminal ileitis. Results corroborated the likely diagnosis of CD. Treatment with infliximab 10mg/kg, TPN and steroids was started with no resolution of symptoms. A second EGD/colonoscopy performed for persistent symptoms showed an extremely dilated colon. Histopathology showed multiple foci in the GI tract with lymphocytic infiltration of the lamina propria, acute cryptitis, absence of goblet and Paneth cells and villous blunting on low power, with relatively scant intraepithelial infiltration on high power. No granulomas, parasitic organisms or viral inclusions were identified. Antienterocyte IgA was positive. Octreotide, loperamide, cholestyramine, ganciclovir and tacrolimus were started with symptomatic resolution.

CR-Diagnosis and Discussion

AE is a rare disease, which frequently masquerades as other intestinal disorders. Pathophysiology is attributed to self-activated CD4+ T cells attacking the intestinal barrier, seen in IPEX syndrome, a severe form of immunodeficient AE with systemic involvement. Alternatively, autoantigens may incite an immune response weakening integrity of the mucosa. Histopathology in the literature describes disease involving multiple foci in the GI tract, villous blunting in the small bowel, crypt abscesses with lymphocytic infiltration although scant intraepithelial infiltration, CD4/8 T cells in the

mucosa and absence of goblet and Paneth cells. Acute management starts with addressing malnutrition and electrolyte imbalances. Mainstay of pharmacologic intervention is steroids. In case of refractory AE, immunosuppressive therapy may be initiated with 6-MP, tacrolimus and vedolizumab among others. Resolution is seen in 60% of patients, incomplete in 20% and refractory AE in 20%. Bone marrow transplant may be utilized.

CR-Conclusion and Significance

This report illustrates a case of severe AE, now responsive to tacrolimus therapy. There is limited existing literature on disease process, further research is needed to elucidate pathophysiology and treatment protocols. Our case report furthers knowledge of this rare disease and illustrates diagnostic work up and treatment in a pediatric patient, who did not respond to infliximab therapy.

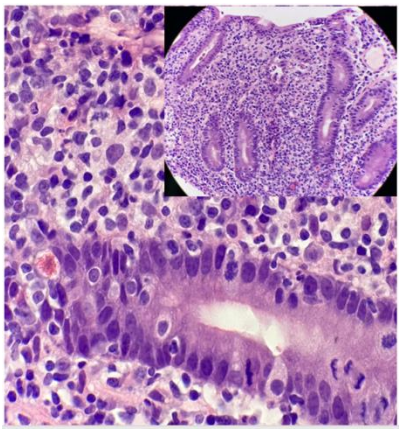


Figure 1: Biopsy of the duodenum showing lymphocytic infiltration of the lamina propria, acute cryptitis, absence of goblet and Paneth cells and villous blunting on low power with scant intraepithelial infiltration on high power

Abstract #50: (RES-030-CR)

Smith JD*; Maher O; Galvez Silva J; Khatib ZA. ***"Complete Response of Therapy Related Myelodysplastic Syndrome to Alisertib"*.**

CR-Introduction/ Background

Therapy related myelodysplastic syndrome (tMDS) is a secondary malignancy after treatment of a primary oncologic process. Prevalence is 5-11% after pediatric solid tumor therapy. Treatment guidelines focus on induction chemotherapy for cytoreduction and subsequent allogeneic hematopoietic stem cell transplant. Overall survival for tMDS is dismal. Atypical teratoid rhabdoid tumor (ATRT) is a highly aggressive class of CNS embryonal tumors. Treatment with surgery, chemotherapy and radiation is standard and may be associated with tMDS. Alisertib is an Aurora A Kinase inhibitor in phase II clinical trials used in the treatment of ATRT, now investigated for tMDS as well as AML. We

report a 3 y/o female patient with tMDS after undergoing treatment for ATRT, with complete response to alisertib therapy.

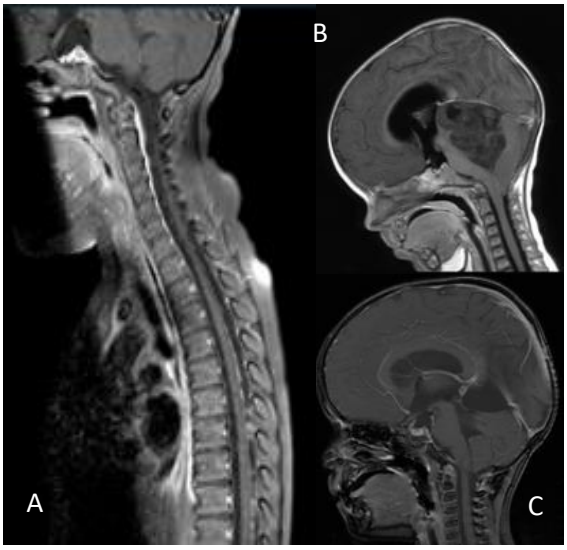
CR-Case(s) Presentation(s)

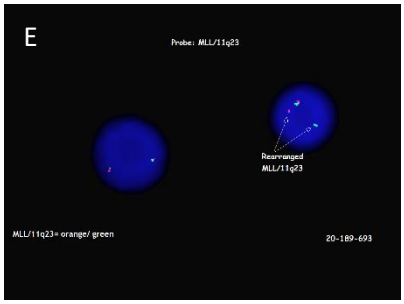
Patient is a 3 y/o female, diagnosed with a posterior fossa ATRT and leptomeningeal spinal disease. Diagnosis was at 12 months of age after 2 weeks of progressive ataxia, lethargy and emesis. MRI showed a posterior fossa tumor without metastasis (figure B). A gross surgical resection was performed (figure C) with ATRT histology (figure D). Molecular testing confirmed a SMARCB1 mutation in tumor tissue and did not show germline mutation. Cytology on LP displayed tumor cells. She achieved complete remission after treatment as per the Dana Farber ATRT regimen and proton radiation. On re-evaluation 9 months off of therapy, new spinal lesions with leptomeningeal coating were observed (figure A). Intrathecal thiotepa/etoposide/topotecan via an Ommaya reservoir was started along with alisertib, as well as metronomic chemotherapy. Follow up bone marrow aspiration for prolonged thrombocytopenia showed MDS with excess blasts, 3.4% on flow cytometry. Cytogenetic studies showed t(11,16)(q23;p13.3) confirming tMDS (figure E). She underwent two rounds of 5-azacytidine treatment at 75mg/m^2/day x 7 days, after which bone marrow aspirate showed 0.1% residual blasts but 86% r-MLL/11q23 by FISH. Alisertib was re-started for recurrent spinal lesions, 3 cycles 40 mg/m^2/day x 7 days q21 days. Subsequent bone marrow aspirate showed an MRD down to 0%, and no r-MLL/11q23 rearrangement identified on FISH (figure F). There was a progressive improvement in blood counts with resolution of cytopenia and transfusions, with no major side effects.

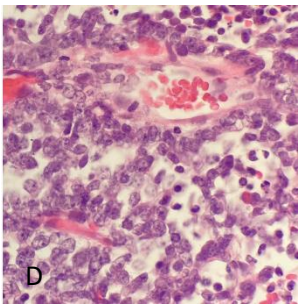
CR-Diagnosis and Discussion

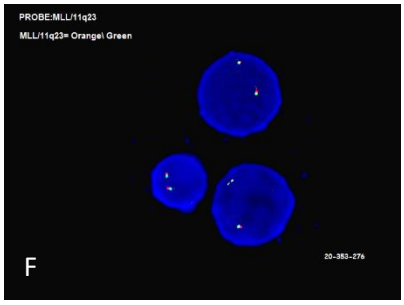
CR-Conclusion and Significance

Alisertib has been investigated for the treatment of ATRT, targeting propagation of the cell cycle. Response of tMDS to chemotherapy has been poor. We report a case of tMDS with complete response to alisertib. This could pose as a novel treatment in the arsenal against tMDS, improving morbidity and mortality. Further investigation is warranted.









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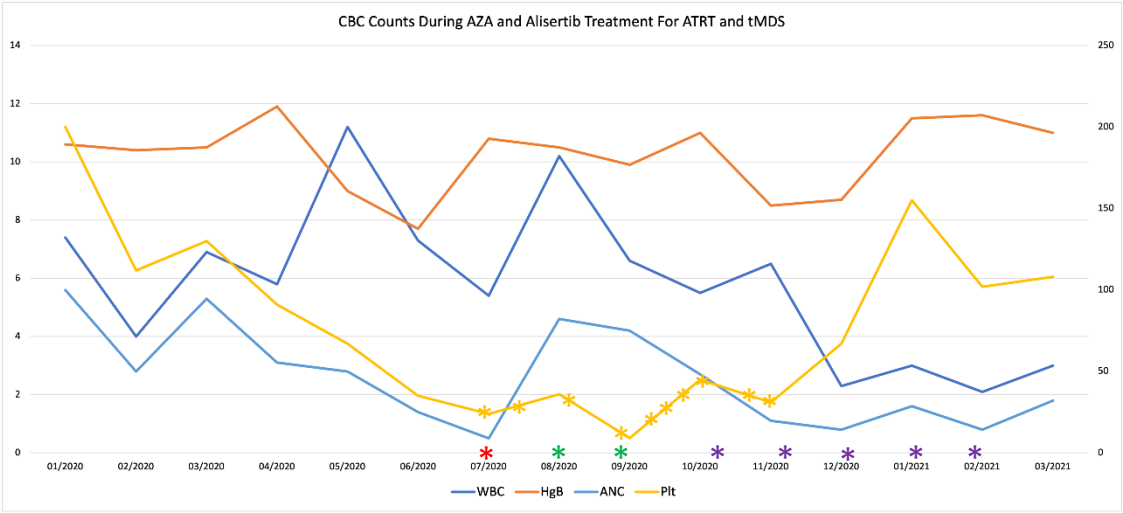


Figure A: MRI showing new spinal lesions with leptomeningeal coating.

Figure B: MRI showing a posterior fossa tumor without metastasis.

Figure C: Post-surgical MRI showing a gross surgical resection.

Figure D: High power resolution displaying ATRT histology.

Figure E: Cytogenetic studies showing t(11,16)(q23;p13.3) confirming tMDS by FISH.

Figure F: Bone marrow aspirate showing no r-MLL/11q23 rearrangement identified on FISH.

Figure G: CBC counts over time trending WBC (10K/uL), hemoglobin (gm/dL), ANC (10K/uL) and platelets (10K/uL) during treatment with 5-Azacitidine (AZA) and Alisertib for ATRT and tMDS. Red star indicates tMDS diagnosis on

7/7/2020. Green stars indicate AZA treatment from 7/30/2020-8/3/2020, 8/27/2020 and 9/1/2020-9/6/2020. Purple stars indicate Alisertib treatment on 10/12/2020, 11/11/2020, 12/7/2020, 1/6/2021 and 1/27/2021. Yellow stars indicate Plt transfusions.

Abstract #51: (RES-031-CR)

Smith J*; Behnam-Terneus M. ***"More Than Just a Rash: A Case of Multisystem Langerhans Cell Histiocytosis".***

CR-Introduction/ Background

CR-Case(s) Presentation(s)

Patient is a 9-month-old male, with a past medical history of eczema, presented with 4 month history of worsening skin lesions, starting on the groin and spreading to the head and back. The lesions were flat, erythematous and desquamating but evolved to be maculopustular and pruritic. He had a swollen hard palate with white plaques. This was diagnosed as oral thrush with an eczema exacerbation, although neither resolved with appropriate therapy. Initial treatment for these inferred diagnoses included fluconazole, cephalexin, amoxicillin, ketoconazole shampoo, triamcinolone cream, Derma-Smooth, mupirocin and cortisone cream, with no resolution. The patient presented to dermatology where a skin biopsy was done and he was admitted for treatment of superinfected eczema herpeticum. On evaluation, there was an erythematous, maculopustular desquamating rash, affecting the inner bilateral thighs and arms. Scaling on the scalp with lesions in multiple stages of healing and dry desquamation was visualized. There was significant swelling of the mandibular gingiva and an erosive, soft mass with white plaques on the hard palate (Figure1). A CBC indicated microcytic anemia with an elevated platelet count (414 10K/uL) and low ferritin (4ng/mL). CMP was normal excluding elevated protein (8.7 gm/dL). Uric acid, ESR, CRP and LDH were within normal limits. MRSA and HSV cultures were negative, subsequent skin cultures were positive for MSSA. A tissue biopsy done on the hard palate mass resulted CD1A positive, indicating the diagnosis of Langerhans Cell Histiocytosis (LCH). A positron emission tomography (PET) scan was performed to elucidate system involvement. Results revealed multifocal, multisystem LCH of the skin, bone and hard palate (Figure 2).

CR-Diagnosis and Discussion

Due to the persistent and systemic nature of the patient's rash, further work up was pursued to rule out superinfected eczema herpeticum vs. LCH. On exam the patient displayed characteristics of systemic LCH including severe intertriginous and scalp rash. Biopsy was consistent with the diagnosis. PET scan corroborated these results, indicating multifocal systemic disease. Langerhans Cell Histiocytosis is a rare inflammatory neoplasm, affecting 4-8 children per one million. It stems from Langerhans cells, myeloid in origin with characteristic markers of Da1+/CD207+. Presentation is varied from

isolated lesions to fulminant multisystemic disease. Prognosis varies dependent on system involvement, from resolution to a fatal course. Initial treatment is a combination of vinblastine and prednisone. Our patient was started on this regimen according to the LCH III protocol for multisystem, multifocal involvement.

CR-Conclusion and Significance

Prompt recognition and diagnosis of Langerhans Cell Histiocytosis is important for optimal clinical outcome. In cases with multisystem involvement, reactivation rates may be as high as 30% with possible long-term sequelae. In this case, warning signs of a persistent systemic eczematous rash that did not respond to treatment, along with significant hard palate mass led to the correct etiology.

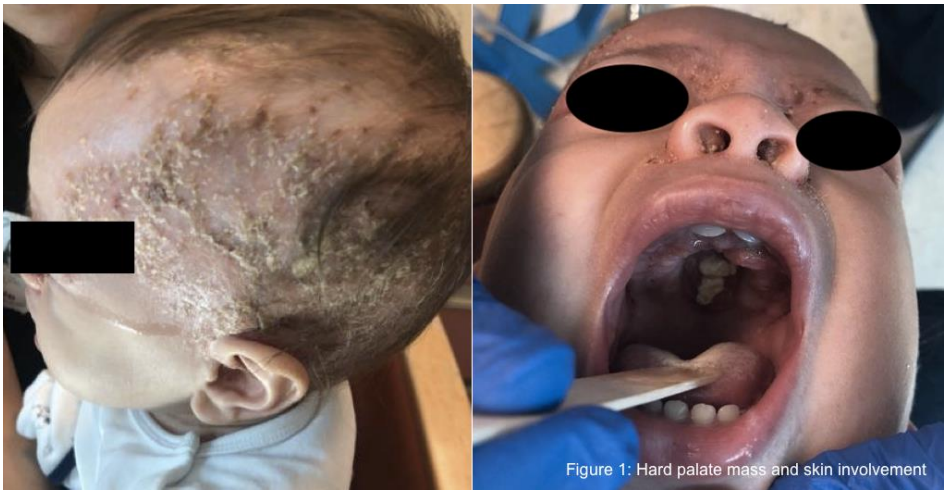


Figure 1: Hard palate mass and skin involvement

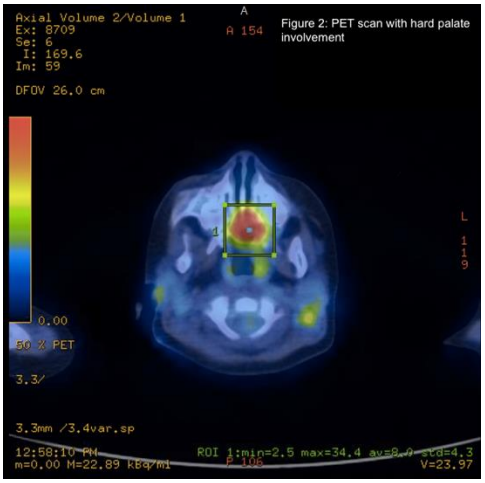


Figure 2: PET scan with hard palate involvement

Abstract #52: (RES-032-CR)**

Suaris M*; Sunny J; Colon Guzman S; Johnson N; Arboleda R; Felipez LM. ***"A Case Series of Non-Bacterial Osteomyelitis Associated with Inflammatory Bowel Disease".***

CR-Introduction/ Background

Approximately 20 percent of the diagnosis of inflammatory bowel disease (IBD) in the United States is comprised of children and adolescents with increasing incidence. Diagnosis may be complicated due to initial presentations involving atypical extra-intestinal symptoms which can occur in 25-40% of IBD patients. A rare manifestation is chronic non-bacterial osteomyelitis (CNO). This autoinflammatory condition presents as multiple or as a single lesion and may be seen clinically as bone pain, arthralgias, and functional disability. The current literature studying the relationship between CNO and

IBD is limited in the pediatric population. We are reporting three cases of CNO between the ages of 7 and 20 and will discuss their presentations and diagnosis.

CR-Case(s) Presentation(s)

Case 1:

A 7-year-old female with a past medical history of very early onset IBD (VEOIBD) managed with infliximab for one year who presented with left clavicular, right ankle, and left shoulder pain. Initial laboratory work-up was unremarkable other than an elevated sedimentation rate and stool calprotectin. Her pain was initially managed with Tylenol. Due to progressive refusal to bear weight a full body MRI was completed. The results showed several focal areas of increased T2 signal particularly in the left clavicle, several thoracic vertebral bodies, right tibia, and left calcaneus. In order to treat both her IBD and newly diagnosed CNO, a higher dose of infliximab was started along with methylprednisolone, which resulted in significant clinical improvement.

Case 2:

A 20-year-old female with history of Ulcerative Colitis, on Vedolizumab, presented with 1.5 weeks of lower back pain that radiated to her calves and thighs. With her history of IBD, difficulty ambulating, and mildly elevated inflammatory markers, a full body MRI was done. Results showed evidence of CNO with lesions in the right clavicle, first right rib, sternum, and spine. To address both her IBD and CNO, she was switched from Vedolizumab to Adalimumab along with Methotrexate and IV steroids. She showed clinical improvement and was discharged with a steroid taper.

Case 3:

A 11-year-old with Crohn's Disease managed with Ustekinumab presented with 3 months of right mandibular swelling and tenderness. A CT scan revealed a poorly defined lytic focus of the right mandible concerning for infectious osteomyelitis versus CNO versus bone tumor. She was empirically treated for osteomyelitis with ampicillin-sulbactam but extensive infectious work up was negative. Bone biopsy showed acute on chronic osteomyelitis and total body MRI showed expansion along the right mandibular ramus, bone marrow edema along L2, S4, and metatarsal edema - suggestive of CNO. She was treated with bisphosphonates with improvement of her joint and muscle pain.

CR-Diagnosis and Discussion

Chronic non-bacterial osteomyelitis is a non-pyogenic autoinflammatory condition. Clinical manifestations include gradual bone pain (most commonly the mandible, clavicle, and pelvis), fever, malaise, and swelling which is similar in presentation to that of bacterial osteomyelitis or arthritis. Approximately 25% of individuals with CNO may have an underlying inflammatory disorder but this relationship is not yet established in the literature, particularly in the pediatric population.

There is no widely accepted diagnostic criteria for CNO leading to possible delay in diagnosis and treatment. In our series, diagnosis was confirmed with the use of a whole-body MRI which showed evidence of a sclerotic/lytic lesions and cortical thickening. Bone biopsies have also been utilized in other studies for confirmation of disease. However, in two of our cases the invasive procedure was not needed due to high index of suspicion. Although there are no standardized guidelines, long term NSAIDS have been traditionally used as first line therapy. Two clinical presentations in our series involved difficulty ambulating secondary to pain and showed prompt clinical improvement with steroids and TNF-alpha inhibitors +/- Methotrexate.

CR-Conclusion and Significance

The aim of our case series was to contribute to the literature on the relationship between CNO and IBD in the pediatric patient not only by presentation but also diagnosis and response to treatment. For our patients with more advanced presentation findings such as inability to bear weight and multiple bone lesions, the use of steroids with a taper along with modulating the TNF-alpha inhibitor yielded a prompt recovery. In our third case, we also documented clinical improvement with bisphosphonate use.

We suggest for future studies to investigate the utility of Methotrexate and bisphosphonates in the treatment of CNO in addition to determining which immunomodulators show increased risk for development of the disease. By having an increased clinical suspicion when evaluating IBD patients with a chief complaint of bone pain and swelling, long term sequalae of recurrent CNO disease can be avoided.

Abstract #53: (RES-033-CR)

Sunny J*; Fonseca A; Rios M; Felipez LM. ***"Acute Pancreatitis in the setting of Lipoprotein Lipase Deficiency: A Familial Case".***

CR-Introduction/ Background

Lipoprotein lipase deficiency is characterized by the deficiency of the enzyme lipoprotein lipase (LPL) which prevents proper breakdown of certain lipids and subsequently leads to accumulation of chylomicrons in the circulation. This subsequently leads to an increase in the concentration of triglycerides (TG) in the plasma.
Hypertriglyceridemia is defined as fasting plasma levels above the 95th percentile adjusted for age and gender1. Affected individuals can experience abdominal pain, pancreatitis, eruptive xanthomas and hepatosplenomegaly2.

We present the case of a 22 -month old with lipoprotein lipase deficiency with severe hypertriglyceridemia who presented with acute pancreatitis.

CR-Case(s) Presentation(s)

A 22 -month old previously healthy Hispanic female presented with fever, abdominal distention, persistent emesis and inability to tolerate oral intake. Initial evaluation was positive for elevation of pancreatic lipase to 985 IU/L and amylase of 144 IU/L. She had associated transaminitis with AST of 77 IU/L. CT abdomen showed pancreatitis with surrounding liquid and inflammation. Lipid panel found to have elevated triglycerides of 1,165 mg/dL, cholesterol of 222 mg/dL, high density lipoprotein (HDL) of 14 mg/dL and low-density lipoprotein of 33 mg/dL. The remainder of initial evaluation including thyroid hormones, respiratory pathogen panel, celiac disease panel, cystic fibrosis panel and amino acid, urine organic acid and acylcarnitine profiles were within normal limits. She was started on aggressive intravenous hydration at 1.5 maintenance rate and began fasting. Within two days of treatment, there was resolution of transaminitis with down-trending lipase and hypertriglyceridemia (ALT of 24 IU/L, AST 56 IU/L, Lipase 512 IU/L and Triglycerides of 627 mg/dL). Genetic studies were positive for a compound heterozygous for a likely pathogenic variant in the Lipoprotein lipase (LPL) gene consistent with autosomal recessive lipoprotein lipase deficiency. She was started on a low-fat diet with nutritional management to restrict dietary fat to 20 grams per day or less (15% of total energy intake). She has been able to maintain goal of triglycerides less than 400 mg/dL with strict dietary management and growth velocity has been normal (BMI of 15.93 kg/m2, 51.08% percentile) at evaluation at 34-month of age. Her younger 2 -month old brother was screened and also found to have hypertriglyceridemia of 669 mg/dL at time of evaluation with cholesterol of 148 mg/dL. Liver function tests and thyroid studies were within normal limits. He was also started on a low- fat diet with Enfaport formula diluted to 20kcal and solids. Most recent laboratory findings revealed triglycerides of 355 mg/dL which is below his targeted goal of less than 400. Growth velocity has been normal for him with weight to length ration of 53.25% with a zscore of 0.08. No adverse events including pancreatitis at this time. Targeted testing of LPL gene is pending.

CR-Diagnosis and Discussion

LPL is an enzyme that degrades circulating triglycerides to fatty acids and glycerol. It is an extracellular enzyme primarily found on the vascular endothelial surface (capillaries), adipose tissue, muscle and the heart3. LPL breaks down triglycerides that are carried by very low-density lipoproteins (VLDL) and chylomicrons. Fat absorbed from the intestines (from dietary consumption) are transported to the bloodstream by chylomicrons and VLDL carries triglycerides from the liver to the bloodstream4. The fatty acids from triglyceride breakdown are used by the body as energy or stored in adipose tissue. Subsequently, any mutations in genes that encode for this enzyme and its cofactors (such as apolipoprotein-CII) will lead to impairments in the lipolytic cascade and accumulation of chylomicrons. Patients with hyperchylomicronemia will present with recurrent pancreatitis, hepatosplenomegaly and xanthomas1. Acute management involves fasting which results in decline in triglyceride levels and allows for gradual clearance of existing chylomicrons. Our patient had a significant drop in TG level after 24 hours of fasting with adequate hydration. Other acute management may involve insulin

(which is an activator of LPL) and plasmapheresis and exchange transfusion, if required1. The long-term treatment for this disorder is a very low-fat diet with close management of nutrition and growth, as seen to be beneficial in our patient.

CR-Conclusion and Significance

Our case is unique in that it highlights the importance of further genetic screening of siblings and other family members of children who present with hypertriglyceridemia. Early screening, identification of deficiency and initiation of proper management will reduce the rate of associated complications, as evidenced by the younger sibling in this case.

Abstract #54: (RES-034-CR)

Sunny J*; Lairet S; Muniz-Crim A; Goyal A; Felipez LM. ***"Hypersensitivity reaction to Ustekinumab in Pediatric Inflammatory Bowel Disease Patients."***

CR-Introduction/ Background

Ustekinumab is a human IgG1K monoclonal antibody that binds to the p40 protein subunit used by cytokines IL-12 and IL-23. These cytokines have been implicated in the chronic inflammation associated with Inflammatory Bowel Disease and Ustekinumab targets this pathway. While hypersensitivity reactions to biologics have been reported, cases regarding Ustekinumab in particular are lacking. We report a case series of five patients who developed hypersensitivity reactions after their initial dose of Ustekinumab, ranging from allergic reactions to anaphylaxis.

CR-Case(s) Presentation(s)

Case 1:
An 18- year old Caucasian female with Ulcerative Colitis (UC) developed an allergic reaction during initial Ustekinumab 390 mg dose. Within 30 minutes of infusion onset, she developed chest tightness, shortness of breath, cough, lip tingling and tachycardia. IV diphenhydramine was given with immediate resolution of symptoms. She has received subsequent subcutaneous injections of Ustekinumab without any further reactions. Previous medical treatments for UC included steroids, Azathioprine, Infliximab, Vedolizumab and an antibiotic cocktail.

Case 2:
An 11- year old Caucasian female with ileocolonic Crohn's disease developed an anaphylactic reaction during Ustekinumab infusion of 390 mg. Despite premedication with Methylprednisolone, she developed abdominal and chest pain, pruritus, cough, shortness of breath, nausea, dizziness and a sensation of throat tightening within 30 minutes of infusion. She was treated with IV diphenhydramine, methylprednisolone and epinephrine with resolution of symptoms. She has continued to receive subcutaneous

therapy with Ustekinumab with no further reactions. She was previously on steroids, Azathioprine, Methotrexate, Infliximab and Adalimumab.

Case 3:
An 11- year old Hispanic female with Inflammatory Small Bowel Crohn's disease experienced an anaphylactic reaction during induction Ustekinumab infusion of 390 mg. She was premedicated with Acetaminophen and Diphenhydramine and within 2 minutes of infusion onset, developed an erythematous rash, cough and appeared flushed. Infusion was immediately stopped, and methylprednisolone was administered with resolution of symptoms. She is currently in clinical remission with Infliximab. Prior to Ustekinumab, she was trialed on Budenoside and Adalimumab.

Case 4:
A 14 - year old Caucasian male with Crohn's disease involving the colon and upper GI tract developed an anaphylactic reaction with induction Ustekinumab dose of 390 mg IV. After 12.1 mL of infusion was administered, he began to cough, turned red and had difficulty breathing. Infusion was immediately stopped, and symptoms resolved immediately after receiving Solumedrol 60 mg IV. No antibodies to Adalimumab or Ustekinumab were detected afterwards. He is currently on Vedolizumab. Previous medical treatments include adalimumab.

Case 5:
A 22- year old Caucasian male with history of stenosing small bowel Crohn's disease requiring ileal resection with primary anastomosis for an acute small bowel obstruction developed an allergic reaction to induction Ustekinumab dose of 390 mg IV. Immediately after initial dose was started, he began to report a "funny feeling" in his chest with associated flushing of the face. Infusion was discontinued and he was given Solumedrol 60 mg IV with immediate resolution of symptoms. Due to acute allergic reaction with Ustekinumab, he was subsequently started on treatment with Adalimumab. Previous medical treatments include Infliximab.

CR-Diagnosis and Discussion

The safety of Ustekinumab was evaluated in 1407 patients with moderate to severe active Crohn's and Ulcerative Colitis disease in 3 randomized, double blind, placebo controlled, parallel-group, multicenter studies. Less than 3% of patients treated with Ustekinumab developed antibodies to the drug and less than 1% reported hypersensitivity reactions in research studies, including signs and symptoms consistent with anaphylaxis. While the drug studies do not indicate a high risk of a hypersensitivity reaction, the cases above demonstrate the possibility of such a reaction.

CR-Conclusion and Significance

Our cases are unique as there have been no previous case reports regarding hypersensitivity reactions including anaphylaxis to Ustekinumab, in the setting of the

first loading dose and no antibodies detected. In some cases, patients were able to continue with subcutaneous Ustekinumab with no further reactions while others were switched to another biologic in other cases due to the hypersensitivity reaction. We hypothesize the reaction is due to EDTA which is only present in the intravenous preparation which should be treated by supportive measures. Patients who have reactions during infusion may still be able to safely receive subcutaneous preparations. Further studies looking at why reactions occur only to IV form of Ustekinumab and not the subcutaneous form are needed.

Abstract #55: (RES-035-CR)

Sunny J*; Bujarska M; Gawron A; Colon-Guzman S; Gonzalez-Vallina R. ***"Orientia Tsutsugamushi and subsequent development of auto-immune hepatitis in a liver transplant Patient".***

CR-Introduction/ Background

Orientia tsutsugamushi (*O. tsustsugamushi*) is an obligate, intracellular bacterium which commonly causes scrub typhus in humans. Genus Orientia belongs to the order rickettsiales within the family Rickettsiaceae. Trombiculoid mites are thought to be the vectors and reservoirs for *O. tsutsugamushi.* Transmission is through bites from these mites. Human diseases caused by agents within this order can range from mild to lethal. Symptoms generally tend to be flu-like (fever, headache and myalgias). In severe cases, meningitis, intravascular complications, severe pneumonitis/peritonitis and cardiac distress have occurred. It is primarily distributed throughout the Asia Pacific rim and scrub typhus is endemic in regions of Korea, China, Taiwan, Japan, Pakistan, India, Thailand and Malaysia. We present a case of a 36 -year old patient who is 23 years post liver transplantation who acquired an infection with *O. Tsutsugamushi* and subsequently developed autoimmune hepatitis.

CR-Case(s) Presentation(s)

A 36 -year old Hispanic female who is status post liver transplantation at the age of 13 years secondary to liver failure of unknown etiology presented with symptoms of abdominal pain, distention and fever two weeks after moving to Indonesia. She was doing very well clinically after transplant and did not have any previous symptoms concerning for rejection prior to presentation, tolerating treatment with Cyclosporine. At time of presentation, laboratory values were significant for elevated total bilirubin and transaminitis in the high thousands. She was found to have *O. tsutsugamushi* in the stool and was subsequently started on management with prednisolone and doxycycline 100 mg daily for 14 days. She completed treatment with resolution of symptoms and improvement of transaminitis. Four weeks afterwards she began to develop symptoms of abdominal distention and pain once again. Liver biopsy that was subsequently performed was suggestive of autoimmune hepatitis and she required further treatment

with prednisolone and mycophenolate mofetil. Our patient has been in clinical remission from infection and autoimmune hepatitis after proper treatment.

CR-Diagnosis and Discussion

Manifestations of *O. tsustsugamushi* may begin with headache, anorexia, malaise, chills or fever and may last for prolonged periods in untreated patients (around 14 days with a range of 9-19). Common presenting laboratory values may include leukopenia, leukocytosis, thrombocytopenia, elevations in hepatic enzymes, creatinine and bilirubin. Mainstay of diagnosis can be with PCR or serology with indirect fluorescent antibody test. Treatment includes doxycycline 100 mg orally or IV twice daily. Azithromycin is also an accepted alternative agent.

CR-Conclusion and Significance

The aim of our report is to bring this unique case of developing *O. tsutsugamushi* and subsequently developing auto-immune hepatitis in a patient who was otherwise doing very well after liver transplantation to focus. Our patient has been in clinical remission after treatment with mycophenolate mofetil and steroids. It is important to consider infectious etiologies in the differential diagnosis for such patients as the clinical course can be very severe. It is also prudent to be aware of potential future complications, including of an autoimmune etiology.

Abstract #56: (RES-036-CR)

Sunny J*; Fonseca A; Muniz Crim A; Felipez LM. ***"Pyoderma Gangrenosum presenting after changing from Anti-TNF to Ustekinumab".***

CR-Introduction/ Background

Extra-intestinal manifestations are common with Inflammatory Bowel Disorders (IBD) including pyoderma gangrenosum (PG)1. PG is a neutrophilic dermatosis that affects the skin and the lesion usually begins as a papule or pustule at a site of trauma with a surrounding violaceous undermining border and subsequent necrosis of the dermis which results in deep ulcers. It is most commonly seen on the lower extremities, but these lesions may present anywhere2. We present two cases of patients with Crohn's disease (CD) who were transitioned from anti- TNF therapy to Ustekinumab and subsequently developed pyoderma gangrenosum.

CR-Case(s) Presentation(s)

Case 1:

An 18-year old Hispanic female with fistulizing Crohn's disease with ileocolonic involvement was initially on Infliximab, treatment discontinued due to antibody

formation. She transitioned to Ustekinumab and received 390 mg IV initial infusion. She was also started on oral Tacrolimus 4 mg twice daily as a bridge. Tacrolimus levels were therapeutic at 13.9 ng/mL. Two weeks later, she developed severe epigastric pain and emesis in the setting of a small bowel obstruction and required hospitalization. She was also found to have multiple skin lesions in the right axillary and right pre-tibial region that were clinically consistent with pyoderma gangrenosum. Biopsy findings were confirmatory with infiltrates of neutrophils within the epithelium and occasional necrotic keratinocytes. Inflammatory markers were elevated (ESR 38 mm/hr, CRP of 5.6 mg/dL) with elevated stool calprotectin of 92.3 mcg/gm. She was treated with two doses of IVIG of 500 mg/kg and a prednisone taper with significant improvement of skin lesions afterwards.

Case 2:
A 15 -year old Hispanic female with Crohn's disease of the upper GI tract with ileocolonic involvement with thickening of a long segment of distal ileum up to the ileocecal valve treatment was initially started on Infliximab. She had worsening inflammatory markers (CRP of 41.5 mg/dL, Platelets of 391 10K/uL) and persistent right lower quadrant abdominal pain with repeat EGD/Colonoscopy revealing terminal ileum stenosis with possible stricture. She was subsequently started on Ustekinumab, 390 mg IV infusion. She subsequently developed a generalized skin rash with plaques of psoriatic disease with biopsies revealing dermatitis. A month after Ustekinumab therapy initiation, she began to develop subcutaneous lesions in her lower extremities along with abdominal pain in the right lower quadrant and periumbilical region. Biopsies were taken from site and confirmed to be pyoderma gangrenosum. Inflammatory markers were elevated (ESR of 13 mm/hr, CRP of 7.5 mg/dL. She was started on prednisone 40 mg with a taper with improvement of symptoms afterwards.

CR-Diagnosis and Discussion

Both of our patients had severe Crohn's disease which required transitioning medical management from Infliximab to Ustekinumab and they both subsequently developed pyoderma gangrenosum within two-four weeks. Both patients were symptomatic with abdominal pain and elevated inflammatory markers at time of presentation with lesions and they both eventually required ileocolonic resection due to severity of disease. There is no targeted or specific therapy for PG but the goals are to reduce inflammation of the lesion and promote healing. Common treatment regimens include antimicrobials and anti-inflammatories, immunosuppressants such as methotrexate, immune modulators such as IVIG and steroids3. Both of our patients responded well to treatment with IVIG and steroids.

CR-Conclusion and Significance

While pyoderma gangrenosum is a known extra-intestinal manifestation of IBD, our cases are unique in that there are no previous case reports that depict the development

of pyoderma gangrenosum immediately after the transition from Anti-TNF treatment to Ustekinumab.

II- FELLOWS' ABSTRACTS:

Abstract #57: (FEL-037-CR)

Chang Y*; Lairet S; Calderon JG; Hernandez-Trujillo VP. ***"A Case of Drug Allergy –Or is it?".***

CR-Introduction/ Background

The Jarisch-Herxheimer reaction is a systemic inflammatory reaction secondary to the release of endotoxins. It results in high levels of TNFα, IL6, and IL8. Symptoms are similar to anaphylaxis and occur immediately after administration of antibiotics, thereby often being confused as a drug allergy. Here we present a child with E. coli urosepsis and likely Jarisch-Herxheimer reaction.

CR-Case(s) Presentation(s)

To describe a case of systemic cytokine release and septic shock not responding to treatment for anaphylaxis.

CR-Diagnosis and Discussion

This patient initially presented with 2 days of fever, back pain, and dysuria. She was found to have left pyelonephritis and treated with cefotaxime. Soon after antibiotic administration, she developed tachycardia, difficulty breathing, rash, and hypotension. She was given multiple doses of epinephrine and transferred to the PICU. Given concern for drug allergy, she was changed to meropenem and developed hypotension requiring epinephrine. She was subsequently labeled as allergic to penicillins, cephalosporins, and meropenem. When consulted, the allergy team diagnosed septic shock and suspected Jarisch-Herxheimer reaction, as the patient was not improving despite aggressive treatment for anaphylaxis. She improved clinically once treatment for septic shock was initiated.

CR-Conclusion and Significance

In this age of growing antibiotic resistance, a misdiagnosis of drug allergy, especially in a child, can greatly affect and restrict patient care. It can turn simple outpatient treatments into long inpatient stays. We believe that in patients not responding to appropriate treatment for anaphylaxis, septic shock and Jarisch-Herxheimer reaction should be considered. The patient should be followed closely with consideration of antibiotic testing and/ or reintroduction.

Abstract #58: (FEL-038-CR)

Chang Y*; Urschel D; Hernandez-Trujillo VP; Calderon JG. ***"A Case of Mild COVID-19 in a Teenager with Common Variable Immunodeficiency and Granulomatous Lymphocytic Interstitial Lung Disease on Replacement Immunoglobulin and Infliximab".***

CR-Introduction/ Background

Common variable immunodeficiency (CVID) is a disorder of the immune system. Patients are generally diagnosed in their adolescence and require regular immunoglobulin replacement. Due to their immune dysregulation, they are also at increased risk of infection, autoimmune disease, and cancer.

CR-Case(s) Presentation(s)

To describe a case of mild COVID-19 in a patient with CVID and granulomatous interstitial lung disease on replacement immunoglobulin and infliximab.

CR-Diagnosis and Discussion

The patient is a 15 year old female with CVID, granulomatous interstitial lung disease, and hepatosplenomegaly with pancytopenia and portal hypertension. She receives weekly SCIG and monthly infliximab. In July, an uncle who visits frequently tested positive for COVID-19. Mother, father, and brother subsequently had mild nasal congestion and tested positive for COVID-19. Our patient developed symptoms of headache and nasal congestion, for which Azithromycin was prescribed and symptoms resolved. No fever, cough, shortness of breath, or loss of sense of smell or taste was reported. No hospitalization was required. Her most recent labs with lymphocyte subsets revealed normal CD4 and CD8 T cell levels, low CD19 B cell levels, and normal immunoglobulins. Follow up COVID-19 PCR testing 1 month later remains positive.

CR-Conclusion and Significance

There are concerns regarding persons with immunodeficiency being at higher risk of serious illness from COVID-19. However, our patient's clinical course suggests that certain types of immunodeficiency or immunomodulators may potentially limit the cytokine storm which causes some COVID-19 complications. Further studies are needed.

Abstract #59: (FEL-039-CR)

Urschel D*; Hernandez-Trujillo VP; Calderon J. ***"Medication Induced Agranulocytosis".***

CR-Introduction/ Background

A diverse array of congenital and secondary conditions are responsible for neutropenia, leukopenia, and agranulocytosis in infants and children include. These include primary immunodeficiencies, genetic abnormalities, infections, malignancies such as leukemias,

and autoimmune disorders. A variety of medications have known associations of agranulocytosis including Dipyrone (Metamizole).

CR-Case(s) Presentation(s)

An eleven-month-old previously healthy vaccinated male traveled with family to Cuba. He developed fever and oral lesions after arrival. Patient was diagnosed with viral stomatitis and treated with an over-the-counter antipyretic. Symptoms worsened to include frequent drooling, decreased oral intake, cough, and intermittent stridor leading to multiple ER visits and admission after returning to the United States. Evaluation showed thrush, stomatitis, stridor at rest. Initial laboratory evaluation at admission demonstrated neutropenia (ANC 294) and leukopenia (WBC count of 1400), with normal hemoglobin and platelet counts. Concerning imaging included unilateral infiltrate/pneumonia on chest x-ray and epiglottitis on CT of neck. Immunologic laboratory evaluation pertinent for borderline low IgG for age, normal IgM and IgA, significant T cell lymphopenia, normal B cell and NK cell lines, normal antibody/antitoxin titers S.Pneumoniae, H.Influenzae B, Diphtheria and Tetanus.

CR-Diagnosis and Discussion

The initial concern in our case of an infant presenting with severe illness, leukopenia, T cell lymphopenia and neutropenia was for a primary immunodeficiency such as severe combined immunodeficiency. Identifying the underlying cause of these lab abnormalities in a severely ill infant without a significant medical history is challenging. Infections, primary immunodeficiencies, malignancies, idiosyncratic drug reactions, autoimmune and genetic conditions must be considered. The patient's symptoms in conjunction with recent medication history, rapid clinical improvement, resolution of leukopenia and neutropenia following medication discontinuation solidified the diagnosis. This represents the importance of thorough history taking and awareness of secondary immunodeficiencies especially given the severity illustrated in this case.

CR-Conclusion and Significance

Dipyrone is a medication commonly used analgesic, antipyretic, antispasmodic, and anti-inflammatory throughout the world. It has well documented association with agranulocytosis. Following the availability of non-steroidal anti-inflammatory medications, it was removed from United States, British, and Swedish markets. The mechanism of agranulocytosis secondary to Dipyrone is not fully understood although it is believed to be immune mediated via anti-neutrophil antibody production. Illness secondary to agranulocytosis can be severe and life threatening. Treatment includes withdrawing the offending medication and secondary infections. Granulocyte colony-stimulating factor is a treatment option. Studies from several countries have demonstrated wide ranges of variation in incidence. Further research is necessary to confirm relative risk and whether certain HLA alleles may predispose to higher risk.

Abstract #60: (FEL-040-CR)

Urschel D*; Cardenas M; Hernandez-Trujillo VP. ***"Chronic Variable Immunodeficiency and Coronavirus".***

CR-Introduction/ Background

Common variable immunodeficiency is one of the most common humoral immunodeficiencies. Many risk factors were identified early in the 2020 COVID-19 pandemic. Evidence of severe morbidity and mortality risk in specific immunodeficiencies was not initially clear. Our case report adds to the limited documentation of COVID 19 in common variable immunodeficiency patients.

CR-Case(s) Presentation(s)

29-year-old with common variable immunodeficiency, lymphangiomatosis, portal hypertension status post TIPS procedure was lost to follow up. He was off gamma globulin and prophylactic antibiotics for fourteen months prior to presentation secondary to insurance issues. Early February 2020 patient presented with abdominal distension and hemoptysis. He denied fever, shortness of breath or additional respiratory symptoms. Initial CT of chest showed basilar cavitary infiltrate. Coronavirus PCR testing was positive, presumed SARS-CoV2. Immunoglobulin G level was 467.

CR-Diagnosis and Discussion

The patient was started on empiric broad spectrum antibiotics, antifungals and immunoglobulin replacement. He developed fever, worsening respiratory symptoms, and lower lobe infiltrates soon after admission. Bronchoscopy was performed, and culture was positive for Aspergillus. Blood culture was positive for Streptococcus Agalactiae. Paracentesis, TIPS check with balloon dilation, and splenic artery aneurysm coiling were performed. He developed thrombosis of greater saphenous vein and was started on anticoagulation. He required oxygen supplementation, but not intubation. Patient gradually improved and was discharged after seventeen days hospitalization. He was continued on antibiotic, antifungal, and anticoagulation at discharge. Outpatient follow up was arranged with Immunology, subcutaneous immunoglobulin was resumed, and he continues to clinically improve to baseline three months after discharge

CR-Conclusion and Significance

The knowledge of SARS-CoV2 infections in patients with underlying Immunodeficiency is rapidly evolving. Immunodeficiency has been speculated to protect against cytokine storm and hyper inflammation from COVID-19. Markers of cytokine storm were not measured during our patient's admission. However, despite coinfections and additional sequelae, our patient had no evidence of ARDS or requirement for prolonged respiratory support.

QI PROJECTS

I- RESIDENTS ABSTRACTS:

Abstract #61: (RES-001-QI)

Gawron Roberts A*; Fonseca A; Colon Guzman S; Vidal G; Rios M; Reeves-Garcia J; Farias A; Behnam M. ***"Improving Adherence to the Management of Acute Pancreatitis in the Pediatric Population."***

(QI) Introduction

In 2018, the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) Pancreas Committee released recommendations for the management of Acute Pancreatitis (AP) in the pediatric population. These are the first recommendations of their kind for this population. Our team wanted to determine how well our hospital follows these recommendations and then improve our adherence to them by providing an educational intervention, to ultimately improve management of AP in our institution.

QI-Plan

Our quality improvement plan was to inform stakeholders of and measure the adherence to the updated NASPGHAN guidelines for the management of uncomplicated acute pancreatitis in the pediatric population in order to better adopt these recommendations in our practice and to ultimately improve the management of patients with acute pancreatitis at Nicklaus Children's Hospital.

Our SMART aim was to improve adherence to NASPGHAN recommendations by specifically targeting 4 main criteria: initiation of enteral feeds within 24-72 hours, avoiding prophylactic antibiotics when not clinically indicated, providing appropriate maintenance IV hydration (1.5 -2 times maintenance), and optimizing pain management with the appropriate use of first and second line pain medications. We focused our interventions on 3 different stakeholders: pediatric residents, hospitalists, and gastroenterologists.

(QI)- Do

Our first intervention was intended to inform stakeholders of the updated NASPGHAN recommendations through the use of a short in-person or virtual presentation. Our second intervention consisted of creating and disseminating a flowchart of the summarized NASPGHAN recommendations for acute pancreatitis to be placed in

physician work stations. Our final intervention was to create an acute pancreatitis PowerPlan in Cerner to be utilized when patients with acute pancreatitis are admitted on our floor in order to make adherence to recommendations simple and effortless.
We were able to determine our current adherence to the updated NASPGHAN recommendations by chart reviewing patients previously admitted for the management of acute pancreatitis. We then administered our intervention and measured the validity of educational intervention with pre- and post-intervention tests to assess short-term retention. Finally, we did a second post-intervention Cerner chart review to determine whether our management in adherence to the updated NASPGHAN recommendations improved following our intervention.

QI-Study

A significant improvement was noted in 3 of the 4 management recommendations. We also found that more patients were diagnosed appropriately with acute pancreatitis using the diagnostic criteria as highlighted in the updated NASPGHAN recommendations. Finally, although the overall percentage of cases who followed early enteral feeding recommendations decreased, over 90% of cases were still managed correctly with early initiation of enteral feeds within 24 to 72 hours.

(QI)- Act

Prior to our intervention, gaps in knowledge regarding the NASPGHAN recommendations for management of acute pancreatitis were identified, specifically regarding optimization of pain control and adequate maintenance IV fluid rates. After our short educational intervention, significant improvement was seen in 3 out of 4 variables in the management of acute pancreatitis. Furthermore, the correct diagnosis of acute pancreatitis using NASPGHAN criteria was also improved. SMART aims were reached as adherence in >90% of cases was achieved in all variables. We are currently working on approving and launching our new Cerner PowerPlan for the management of acute pancreatitis in accordance to the NASPGHAN recommendations to improve long-term adherence to the guidelines.

Abstract #62: (RES-002-QI)

Graneiro A; Fallatah E; Mandel G*; Hernandez-Trujillo VP. ***"Increasing Awareness of the Montelukast Black Box Warning".***

(QI) Introduction

In March 2020 the FDA issued a black box warning to montelukast in order to raise awareness regarding its potential neuropsychiatric side effects. Montelukast has been associated with worsened and new-onset suicide, depression, irritability, among other neuropsychatric side effects.

QI-Plan

Global Aim- Improve both patient and prescribers knowledge of the new montelukast black box warning on neuropsychiatric events and increase adherence to the black box warning in primary care clinic and allergy and immunology clinics at NCH.
Smart AIM: Improve questioning and documentation of questioning of pediatric patients of all ages on montelukast side effects from 0% to 50% in one year.
Stakeholders: Clinic attendings, pediatric residents, clinic nursing staff.

(QI)- Do

Intervention 1 (7/8/20): Education via presentation
Intervention 2 (8/3/20, 9/1/20): Whatsapp reminders
Intervention 3 (9/2/20- present): PCC cards
Intervention 4 (11/25/20, 1/11/21): Social media presentation
Measures: 50% increase in documentation of teaching and questioning on montelukast use. 50% improvement on black box warning quiz
Methods: 120 charts chosen from a pool of patients with asthma and/or allergic rhinitis from PCC. Pre-intervention charts reviewed between 3/13/2020-7/7/2020. Post-intervention reviewed 7/8/20-1/15/2021. Physician surveys administered pre and post interventions.

QI-Study

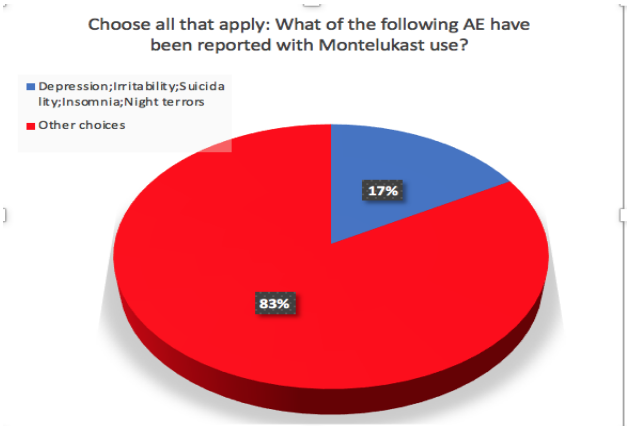
Side effects documented increased from 0/29 to 2/29
Awareness increased from 39% to 98%
Percent of providers who never discussed side effects decreased from 69% to 39%
Knowledge of side effects increased from 17% to 48%

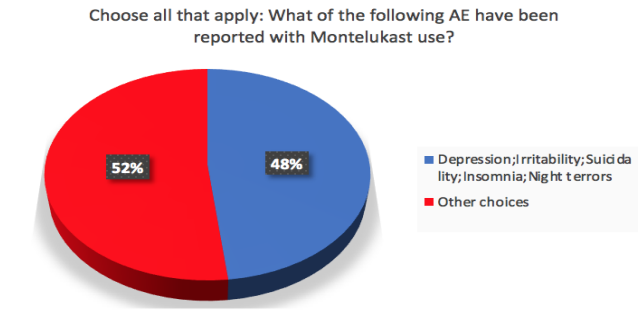
(QI)- Act

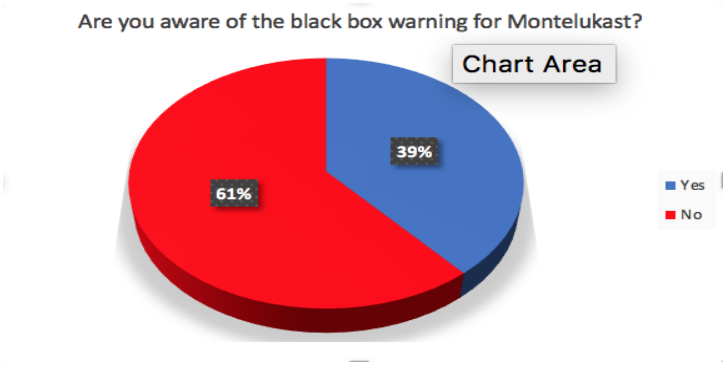
Ultimately the QI project was successful in increasing awareness of montelukast black box warning. The project used innovative social media methods including whatsapp and TikTok to engage and educate the audience which increased awareness.

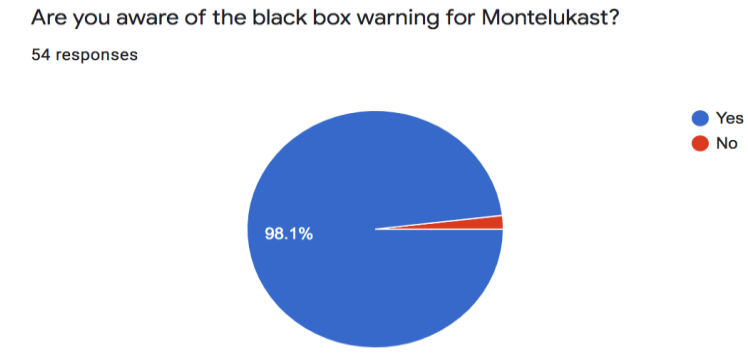
The number of charts with improved documentation did not significantly increase. This was likely due to several factors including poor access to charts, general documentation on montelukast adherence, decreased patient census due to COVID-19 pandemic.

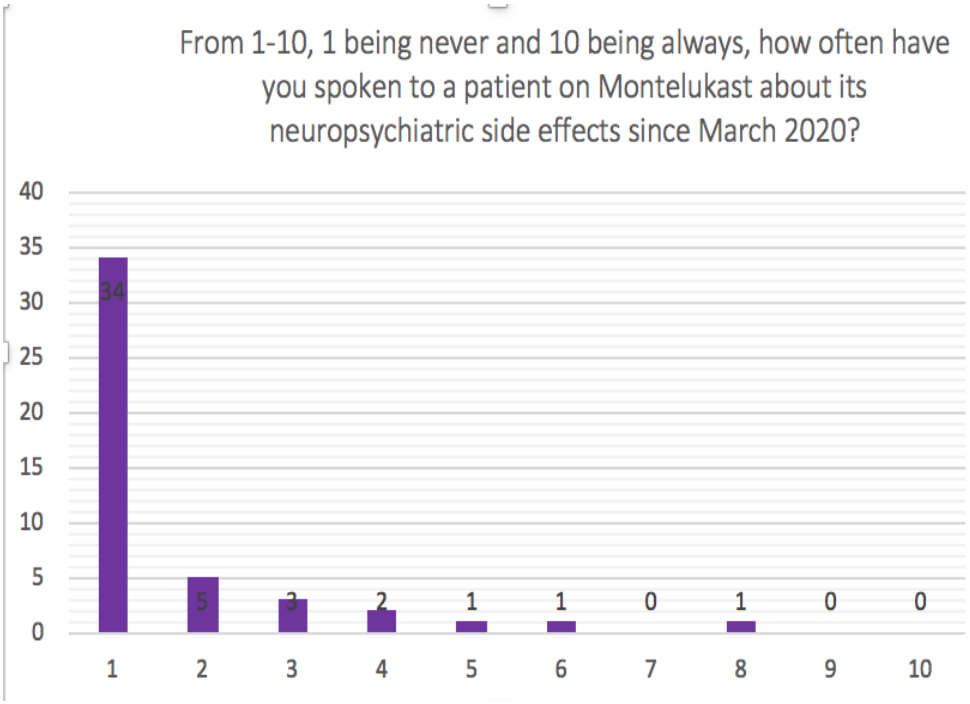
Increased education for patients and providers is warranted. This will lead to appropriate counseling and better outcomes for our patients.


Choose all that apply: What of the following AE have been reported with Montelukast use?
Depression;Irritability;Suicidality;Insomnia;Night terrors
Other choices
17%
83%


Choose all that apply: What of the following AE have been reported with Montelukast use?
52%
48%
Depression;Irritability;Suicidality;Insomnia;Night terrors
Other choices

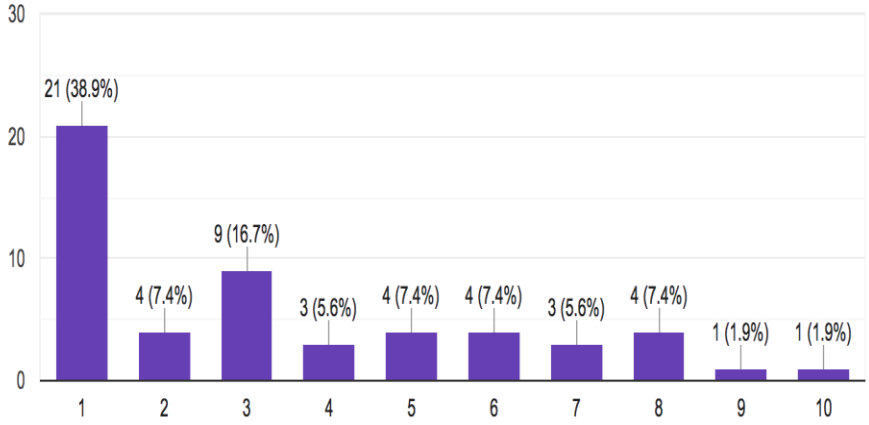

Are you aware of the black box warning for Montelukast?
Chart Area
39%
61%
Yes
No


Are you aware of the black box warning for Montelukast?
54 responses
98.1%
Yes
No



From 1-10, 1 being never and 10 being always, how often have you spoken to a patient on Montelukast about its neuropsychiatric side effects since March 2020?

54 responses



Abstract #63: (RES-003-QI)

Urschel D*; Mendoza M*; Lairet S*; Riefkhol G; Llopiz F; Milla M; Hernandez-Trujillo V. ***"Improving Asthma Control Test Use in the Pediatric Care Center".***

(QI) Introduction

Poor asthma control is a risk factor for further exacerbations and impacts greatly patients' quality of life. Current asthma recommendations include assessing patient's level of control. The Asthma Control Test (ACT) is one of several validated asthma questionnaires to evaluate control in pediatric and adult patients. This questionnaire has been validated in patients over the age of four with the aim to identify poorly controlled individuals in need of treatment escalation. The established cutoff value of ACT scores of 19 or less, identifies patients under poor control. Additionally, these patients would benefit most from having a formal Asthma Action Plan (AAP).

QI-Plan

The goal of the project is to increase rate of use of ACT during PCC visits in patients with asthma diagnosis. We aim to increase the frequency from 15% to 25%. Residents are the main stakeholders in this projects and were educated on the importance of ACTs and how to properly document them in the EMR. Additional visual cues were used as reminders to use ACT in the resident PCC clinic.

(QI): Do

Baseline data was obtained from the electronic medical record (EMR) from Pediatric Care Center (PCC) appointments. The aim was to identify patient's with a diagnosis of asthma who have presented for PCC visits from January-December 2019.

Our first intervention involved educating residents during noon conference and using visual cues in the PCC regarding the ACT questionnaire.

Our second intervention involved educating and making clinic team leaders aware of ACT questionnaire twice per block to encourage them to help enforce ACT use during well-child visits with asthma diagnosis.

QI-Study

Pre-intervention data:

1. 231 charts were collected from 01/2019-12/2019 of only Well Child visits in patients with asthma diagnosis. Of 231 patients, we identified 35 (15%) patients who had ACT questionnaires documented.

Post-1st intervention data:

2. 56 charts were collected from 02/05/2020- 07/20/2020 of only Well Child visits in patients with asthma diagnosis. Of 56 patients, we identified 8 (14%) patients who had ACT questionnaires documented.

Post-2nd intervention data:

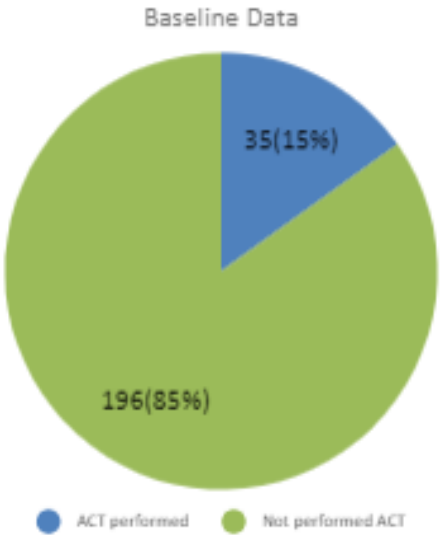
3. 143 charts were collected from 07/21/2020- 02/09/2021 of only Well Child visits in patients with asthma diagnosis. Of 143 patients, we identified 81 (57%) patients who had ACT questionnaires documented.

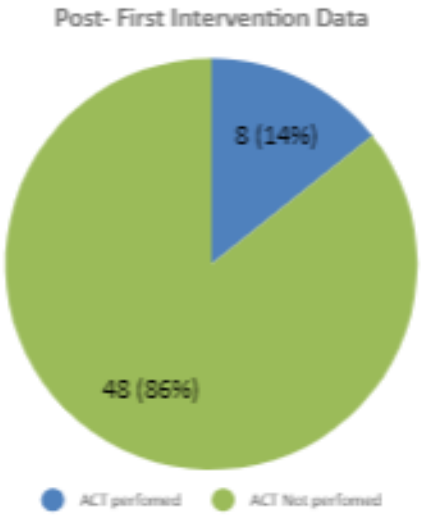
(QI)- Act

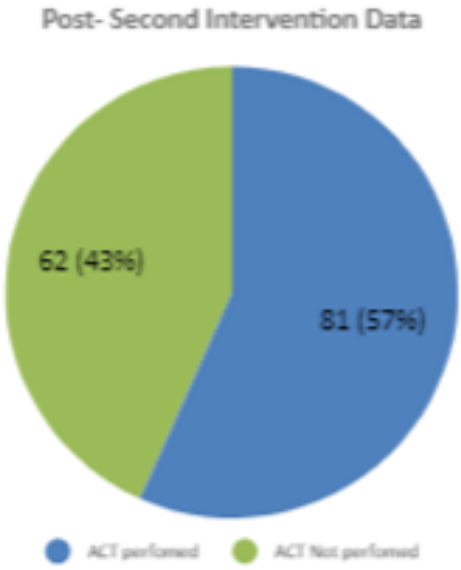
In conclusion, we initially observed a mild decrease from baseline ACT use from 15% to 14% after our first intervention. However, the sample size was much smaller (56 vs. 231), thus only capturing a small portion of the population. Additionally, our patient population was limited due to the COVID pandemic during the first intervention period.

In our second cycle, which began July 21 2020, we shifted from educating residents in noon conference and using visual cues in the PCC to focus on educating clinic team leaders twice per month to aid in distributing ACT questionnaires. The team leaders helped enforce ACT questionnaire distribution to patients that qualified in the PCC. After this second intervention, we observed a significant increase in ACT questionnaire implementation from 14% to 57%. This finding surpassed our goal of 25%. We believe that making the process more systematic was essential for it to be incorporated more seamlessly into a busy clinic workflow.

In the future, making the intervention even more automatic would greatly benefit outcomes. For example, having clinic staff distribute the questionnaires prior to patient encounters.







<u>Abstract #64: (RES-004-QI)</u>
<u>Deiros G</u>; <u>Frias J</u>; <u>Modir P</u>*; <u>Ramos-Garcia A</u>; Milla M. ***"Improving HPV Vaccination Rates at NCH PCC".***

(QI) Introduction

HPV is a very important vaccination that has been continually questioned given that it has not been a mandatory vaccine for school requirements. HPV is a very prevalent disease and is currently the most STI recorded. The HPV vaccine is also the only cancer preventable vaccine. The importance of this vaccine and the gap between parental knowledge regarding this vaccine was a big gap that we wanted address and change.

QI-Plan

Improve HPV vaccination rates in clinic patients between 11-18 years of age that are due to receive the vaccine per CDC guidelines (and have not completed the cycle) from 59% to 65% within 3 months.

(QI)- Do

Pre intervention survey to assess resident comfort level
Educational video with resources to address parental rebuttal
Post intervention survey to measure improvement in resident comfort approaching vaccination discussion with parents

QI-Study

Total number of patients=268
- Overall vaccination rate=83%
- 54% Pt were already vaccinated
- 29% were due for the vaccine and received it
- 17% were due for the vaccine and refused it
- Of those who were not fully vaccinated, 62.6% were due for the vaccine and received it.

(QI)- Act

The vaccination rate increased from 59 to 62.6% compared to baseline data of 2019.
Many pts had incomplete records, making vaccination rates seem lower.
Resident knowledge was the almost the same after intervention (77% and 78%).
Resident confidence level is weakest with vaccine rebuttal.
Next Goal: Simulated cases to help improve resident approach to vaccine rebuttals.
Streamline vaccination documentation to improve accuracy.

Abstract #65: (RES-005-QI)

Liberti A; Montarroyos S*; Sanchez Solano N, Etinger V; Reyes M. ***"Implementation of Asthma Clinical Pathway".***

(QI) Introduction

The purpose of NCH's asthma clinical pathway and corresponding power plan is to ensure standardized, evidence-based and cost-effective care for the treatment of asthma on the general medical floor. The initiative was launched in January 2020. To date, other outcome metrics such as length of stay, CXR use and charges have not been investigated.

QI-Plan

GLOBAL AIM:
Standardize the care of pediatric patients admitted with Asthma exacerbation via the implementation the Asthma Clinical Pathway
SMART AIM:

Continue to achieve >70% resident adherence to "PULM Asthma Floor Admission" power plan for patients admitted with primary diagnosis of asthma exacerbation through 2/2021. In addition, reduce cost and length of stay (LOS) by 20% on 2N and 3S inpatient floors for patients admitted with the diagnosis of asthma exacerbation for a three-month period prior to power plan launch compared to the same time period after launch (10/2019-12/2019 vs 10/2020-12/2020)

STAKEHOLDERS:
ED, Hospitalist and Pulmonology attendings
Pediatric residents
Clinical nursing staff
Respiratory therapists
IT department

(QI)- Do

INTERVENTION #1
Continue ongoing educating residents, nursing and RT staff on proper use of asthma clinical pathway and order sets, through one-on-one educational meetings, "asthma pathway videos", and interactive virtual presentations during noon and other resident conferences.
MEASURES
Outcome Measures:
Resident adherence to "PULM Asthma Floor Admission" power plan
LOS in hours
Charges per patient encounter
METHODS
-To investigate resident adherence to "PULM Asthma Floor Admission" power plan, charts of all patients who received albuterol on 2N and 3S medical floors from 10/2020-2/2021 will be reviewed to assess whether the power plan was utilized and initiated for those patients meeting inclusion criteria for power plan use
-To investigate outcome measures relating to LOS, charges and CXR use, chart reviews of all patients admitted to hospital with with primary ICD-10 code relating to acute asthma exacerbation from the time period 10/2019-12/2019 (n=~73) will be compared using the same criteria for the time period 10/2020-12/2020)

QI-Study

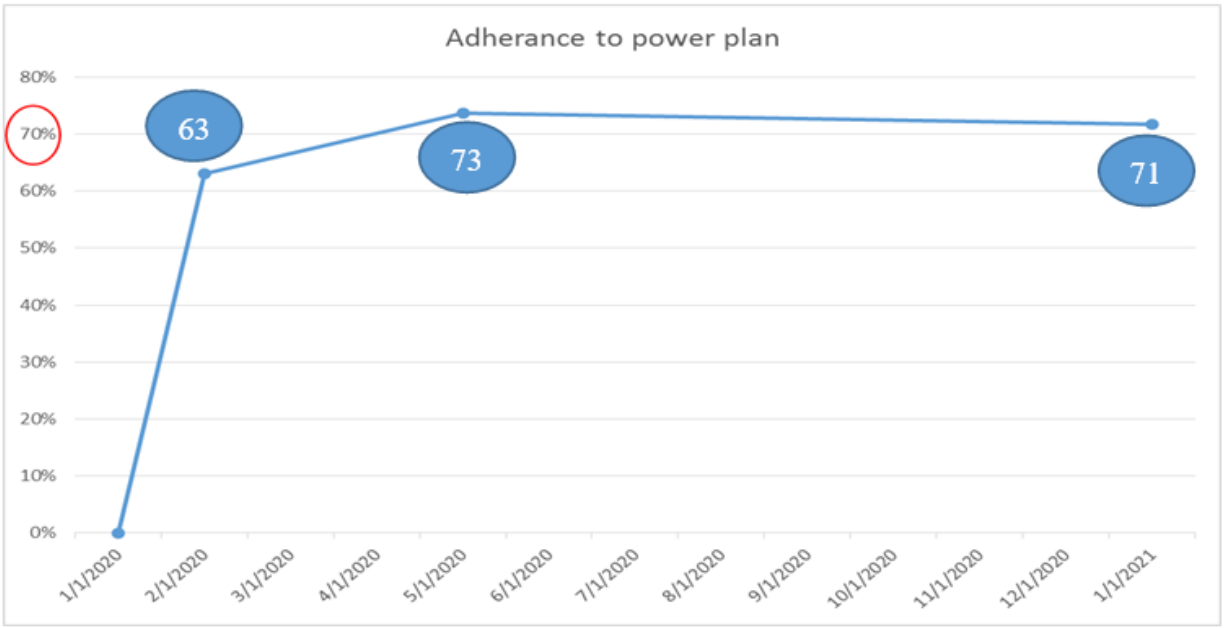
See tables attached to end of document

(QI)- Act

Despite the COVID pandemic which greatly impacted our hospital, we were able to continue to achieve our goal for adherence to the asthma power plan use. However, we did not see a reduction in LOS or total hospital charges as we expected. We believe it may be due to the COVID pandemic, given increase in admissions for more severe cases of asthma, requiring more aggressive treatment and higher level of care. In addition, changes in nursing staff and work flow of 3S/2N secondary to the pandemic could have

led to more changes in personnel leading to a need for more frequent education. The plan is to continue to provide education to medical/nursing personnel as well as expand use of the asthma power plan to 5 Tower.

	Length of stay (hours)			**Total Charges (USD)**		
	Mean	95% CI		Mean	95% CI	
2019	37.84	31.79	43.9	19,599.6	1,6334.9	22,864.2
2020	41.59	34.96	48.22	26,961.8	2,1880.1	3,2042.6
Difference	-3.748	-12.616	-5.12	-7,361.8	-1,2241.4	1,382.1
P value	0.7981			0.9917		



Abstract #66: (RES-006-QI)**

Riera-Canales C*; O'Farrell C; Zaidi Z; Gabay M; Felipez L. ***"Increasing Depression Screening In Admitted Patients 12-21 Years Old With Inflammatory Bowel Disease".***

(QI) Introduction

Inflammatory bowel disease (IBD) affects 100-200 per 100,000 children in the United States (US) and its incidence is increasing. Depression incidence is also increasing in adolescents across the US. Depression prevalence is higher in patients with chronic medical conditions. Particularly in pediatric patients with IBD, depression rates of up to 25% have been described, much higher than the 9-11% prevalence described in healthy kids. However, as many as 75% of cases of depression in adolescents go undiagnosed, with only 25-35% receiving appropriate treatment. Depression in patients with IBD is associated with nonadherence, risk of relapse, worsened disease activity, and higher health care costs. Routine screening strategies can lead to early identification of depression and timely treatment. The PHQ-9 is a self-administered, well-validated questionnaire shown to have good sensitivity and specificity for detecting major depression among adolescents.

Baseline data from NCH showed depression prevalence of 7% in patients 12-21 years old with IBD. (152)

QI-Plan

GLOBAL AIM: increase early detection of depression in patients 12-21 years old with IBD admitted to NCH under the GI PSA service.

SMART AIM: increase early detection of depression in patients with IBD by applying a validated screening questionnaire (PHQ-9) to at least 50% of patients 12-21 years old with IBD admitted to NCH under the GI PSA service between 09/25/20 and 01/25/21.

STAKEHOLDERS:
Nursing staff
PL-1s and PL-3s
GI PSA attendings (72)

(QI)- Do

INTERVENTION #1: educated the medical and nursing teams on the increased incidence of depression in IBD patients and its effects such as poor disease outcomes, increased mental health comorbidities, increased medical costs and poorer quality of life.

INTERVENTION #2: provided all the documentation needed including a cover sheet for parents and patients explaining the reason of the questionnaire, questionnaires in English and Spanish, and the grading scale. Also provided a bin to collect answered questionnaires and reminders of our project were placed in the work rooms

MEASURES: total number of patients with IBD ages 12-21 admitted under the GI PSA service during 09/25/20-01/25/21 and then calculated the percentage that received screening with PHQ-9.

METHODS: an excel sheet database was built including all admissions fitting our study population between 09/25/20-01/25/21 including PHQ-9 scores, actions taken, previous mental health diagnosis and IBD-specific data (age at diagnosis, phenotype, current treatment, etc.) (148)

QI-Study

Total IBD PSA admissions from 9/25/20-1/25/20: 26

Total patients screened: 8 (30.8%), found to be depressed: 6 (23%)
Mild-to-moderate depression: 4 (50%)
Moderately severe-to-severe depression): 2 (25%)

Prior depression diagnosis in patients screened: 0 (0%)

Prior depression diagnosis in all admissions: depression 2 (7.7%)

Depression dx in all admissions (abnormal PHQ-9 or previous dx): 8 (30.8%)

Patients admitted for treatment failure were more likely to have depression than patients admitted for other reasons (infections or new diagnosis). The types of IBD, severity or location were similar in both groups however the length of stay (LoS) was almost double in patients with depression (16 vs. 9 days). (106)

(QI)- Act

Routine depression screening increases detection of depression.

Lower prevalence of depression seen in baseline data likely due to underdiagnoses. Depression may be more prevalent in pediatric patients with IBD at NCH than in other pediatric patients with IBD.

Specific types of IBD, sites of involvement and phenotypes do not seem to correlate with increase risk of depression.
Admission for treatment failure seems to be associated with increased risk of depression. LoS is significantly longer in patients with depression. Does depression have a direct impact on the LoS or depressive symptoms develop with longer inpatient stays?

Screening for depression is a valuable intervention in admitted patients with IBD and can have a positive impact on the patient's illness course. (118)

Abstract #67: (RES-007-QI)**

Alruwaili A; Bujarska M; Lopez Gonzalez M; Sunny J*; Felipez LM; Behnam-Terneus M. ***"Appropriate Thromboembolic Prophylaxis Therapy in IBD Patients Admitted to the Hospital: A Proposed Risk Stratification Algorithm".***

(QI) Introduction

Inflammatory bowel disease is high risk for development of both venous and arterial thromboembolism (TE). The risk of TE in IBD vs non-IBD patients is 6-fold higher overall and can rise as high as 15-fold during disease flares. The increased risk is thought to be due to the chronic inflammatory state, extensive mucosal damage impairing physiological anticoagulation, and long-term steroid use increasing fibrinogen and clotting factors. When combined with other risk factors such as immobility during hospitalization and insertion of central lines, the need for prophylaxis is evident. Adult patients already use Lovenox as a standard of care during inpatient care of IBD flares but this is not seen frequently in pediatric medicine. Certain pediatric centers such as Boston and Cincinnati have begun using algorithms for TE prophylaxis in children. Our QI project intended to create a TE prophylaxis algorithm at Nicklaus Children's Hospital for IBD patients and consisted of two PDSA cycles.

QI-Plan

GLOBAL AIM:
To improve the practice of appropriate TE (thromboembolic) prophylaxis therapy in pediatric IBD patients
admitted to the general floors at Nicklaus Children's Hospital.

SMART AIM:

1. Determine percentage of admitted pediatric IBD patients who receive appropriate TE prophylaxis therapy based on a risk stratification algorithm.
2. Measure percentages before and after secondary intervention consisting of educational sessions, risk stratification questionnaire.
3. Our goal is to increase percentage of IBD patients receiving appropriate prophylactic therapy measured over a 24-week intervention period.

STAKEHOLDERS:
1. GI attendings, both PSA and PGA groups
2. Pediatric Residents (interns and team leaders)

(QI)- Do

INTERVENTIONS:

1. Provide education of residents regarding the importance of risk assessment about correct documentation
2. Create a DVT risk stratification questionnaire for easier risk categorization
3. Educate both GI practice attendings about goal of QI

MEASURES

1. DVT Risk Stratification Questionnaire usage
2. Percentage of patients who received appropriate therapy from chart review
3. Percentage of attendings who reported receiving/reviewing current data

METHODS

1. Implement interventions and then conduct a chart review of a minimum of 50 patient charts of IBD patients admitted from August to February to assess risk categorization of patients and receipt of appropriate therapy.
2. Assess for use of risk stratification questionnaires.
3.

QI-Study

Baseline Data: 30% of inpatient IBD patients received appropriate prophylaxis.

Cycle 1: 14% of high-risk patients received Lovenox (3 out of 21)

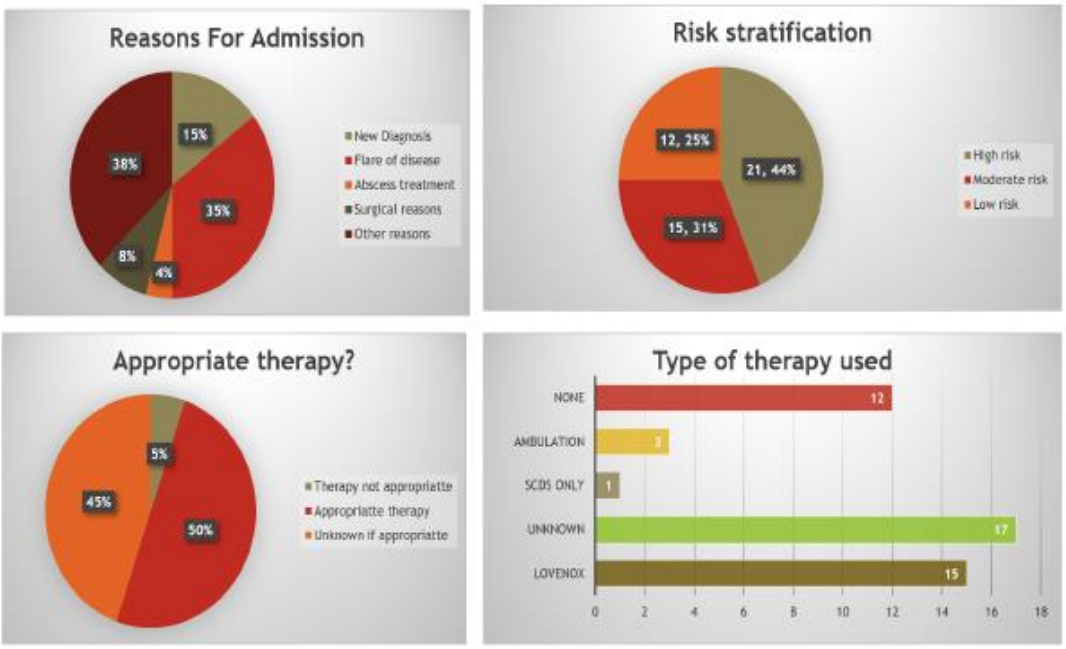
Cycle 2:

- 48 patients were admitted from 08/2020-02/2021 under Gi PSA/GI PGA.
- 50% received appropriate therapy, 45% unknown due to lack of documentation of intervention and 5% did not receive appropriate therapy.
- 70% of high-risk patients received Lovenox (15 out of 25)

(QI)- Act

1. 50% of inpatient IBD patients received appropriate DVT prophylaxis compared to 30% from baseline data. The actual percentage may be higher, but this remains unknown as documentation for moderate risk group (SCDs) remained unclear.
2. Only 5 risk assessment forms were filled out in total indicating that the forms had minimal contribution to the increased percentage of patients with appropriate therapy noted during PDSA cycle 2.
3. No patient received Lovenox inappropriately (all patients were in High risk group)
4. There was no significant increase in GI bleeding reported in any patient receiving therapy with Lovenox. The most common adverse side effect was injection site discomfort.
5. 57% of PSA and 40% of PGA attendings confirmed review of current data. Continuing to discuss current data with IBD patients admitted under other independent practices and Baptist GI attending may contribute to increased incorporation of risk stratification algorithm.

Study/Results:



<u>Abstract #68: (RES-008-QI)</u>
<u>Rezk M</u>; <u>Thompson J</u>*; Comkornruecha M; and Gray S. ***"Improving Screening for Child Abuse in Adolescent Medicine Clinic".***

(QI) Introduction

Screening for abuse in adolescents is important due to the severity of its consequences. Our QI project will bring awareness to this subject and encourage providers to have a discussion with patients to screen for abuse so that resources can be provided if abuse is disclosed.

QI-Plan

In the Adolescent Medicine clinic, each patient encounter gets a regular note written and a HEADS note should be written for any confidential HEADS information that a patient may not want their guardians to see. Many times a HEADS note is forgotten about. Our plan is to implement a didactics session for pediatric residents into their adolescent medicine rotation curriculum discussing screening and management of abuse. We also encouraged providers to document screening in regular notes instead of in the HEADS notes.

(QI)- Do

Baseline Data: There were a total of 1345 patient encounters in the Adolescent Medicine clinic from 9/1/2018 to 12/31/2018. Only 7 of the 1345 patient encounters had a HEADS note in this 4 month period. Of the 7 HEADS notes, 4 of them screened for abuse. Therefore only 4 out of the 1345 patient encounters were screened for abuse, which is 0.29%.

PDSA #1 Data: There were a total of 1489 patient encounters in the Adolescent Medicine clinic from 1/1/2019 to 4/30/2019. There were 69 of the 1489 patient encounters that had a HEADS note in this 4 month period. Of the 69 HEADS notes, 14 of them screened for abuse. Therefore only 14 out of the 1489 patient encounters were screened for abuse, which is 0.94%.

PDSA #2 Data: There were a total of 1492 patient encounters in the Adolescent Medicine clinic from 6/19/2019 to 10/18/2019. There were 43 of the 1492 patient encounters that had a HEADS note in this 4 month period. Of the 1492 regular notes, 342 of them screened for abuse, which is 22.92%.

QI-Study

	Baseline 9/1/2018 – 12/31/2018	PDSA #1: 1/1/2019 – 4/30/2019	PDSA #2: 6/19/2019 – 10/18/2019
Patient Encounters	1345	1489	1492
HEADS Notes	7	69	43
HEADS Notes Screening for Abuse	4	14	N/A
Total Screened for Abuse	4/1345 (0.29%)	14/1489 (0.94%)	342/1492 (22.92%)

(QI)- Act

- Increased number of patients screened for abuse from 14 to 342
- Overall percentage of patients screened improved, from 0.29% to only 22.92%
- Opening 2 notes per patient (regular note and a HEADS note) increases work for providers, therefore screening was less likely to be done.
- Having providers screen for abuse in regular notes improved screening rates more than if HEADS notes were used for screening.
- We will continue our current intervention with the didactic session and using regular notes with embedded questions.

Current intervention of embedding a screening question in regular patient notes rather than in a HEADSS note has improved screening rates and our SMART goal was reached.

II- NURSING ABSTRACTS:

Abstract #69: (NUR-009-QI)

Augustin W*. ***"Empowered to Discharge: Development of Nurse-led Clinical Guidelines to Decrease Length of Stay and Cost".***

(QI) Introduction

The purpose of this evidence-based practice project was to create a nurse-driven in-patient plan of care (IPOC) for postoperative appendicitis care and discharge planning. An opportunity was identified to create nurse-led criteria and guidelines to expedite patient discharge after admission to a short-stay observation unit.

QI-Plan

In pediatric patients with acute appendicitis, the emergence of laparoscopic surgery has significantly decreased the postoperative length of stay (LOS). This has the potential to reduce hospital costs and improve patient experience by providing a coordinated transition to the community and recovery at home. However, the inpatient care process could be further streamlined by empowering nurse-led care and discharge planning. Allowing nurses to initiate discharge orders and planning based on IPOC criteria was hypothesized to decrease LOS in the pediatric population admitted for laparoscopic appendectomy.

(QI)- Do

In 2020, working in collaboration with pediatric surgery, a nurse-led team developed IPOC guidelines based on clinical expertise and published literature on best practices for discharge. A literature search and comprehensive review informed the development of

discharge criteria. In-service education was provided to nursing staff on the IPOC, and it was uploaded into the electronic medical record system in November 2020. LOS for patients undergoing appendectomy in quarter one of 2020 (January-March) was compared to LOS for patients post-practice change (Dec 2020-Feb 2021).

QI-Study

The median LOS for 2020 was 0.86, 0.84, 0.66 days for January, February and March 2020, respectively. After IPOC implementation median LOS was 0.74, 0.68, and 0.79. The overall mean LOS pre-and post-implementation was 0.79 days vs. 0.74 days. Nurses reported increased autonomy and high satisfaction with the IPOC guidelines and nurse-led discharge process.

(QI)- Act

Creation and implementation of the IPOC guidelines for appendicitis empowered nurses to initiate the discharge process for patients undergoing appendectomy. There was a decrease in average LOS (0.79 vs. 0.74 days) however a longer data collection period is needed. Further staff education for nurses will be performed to sustain the implementation of nurse driven IPOC utilization.

Abstract #70: (NUR-010-QI)

Burke ML* & Taylor-Amador S. ***"Screening for Malnutrition in Oncology".***

(QI) Introduction

Many pediatric oncology patients suffer from side effects on all body systems related to their treatment for cancer. Protocols for most childhood cancers involves chemotherapy, biotherapy, radiation, surgery and other innovative treatments. Most of these therapies and disease related side effects are known to cause adverse symptoms that affect the child's nutritional status. Some of the common gastrointestinal (GI) alterations may include: changes in taste, nausea, and vomiting, poor appetite, aversion to food, weight loss / gain, and abdominal discomfort. The results of these GI disturbances may lead to malnutrition which can contribute to poor treatment outcomes.

QI-Plan

Objectives

1. The development of a standardized tool the Pediatric Oncology Nutrition screen (PONS) for early identification of malnutrition.
2. Determine if the PONS tool provided an early indication of malnutrition in the pediatric oncology patient based on the measured variables.

3. Prevent delays in treatment regimens due to poor nutritional status

(QI)- Do

The PONS tool was developed to assess if the if child's cancer treatment influenced the degree of malnutrition. A 7- item tool was developed by the Clinical specialist and the Oncology Dietician to evaluate the patient's nutritional status. Initial a small pilot test was completed and then the tool was presented to be added to the oncology documentation in PEDS. Nurses would complete the first five items of the tool and the dietician would complete the last two items to tabulate a total score to determine the degree of malnutrition. After an IRB approval a total of 414 patients were evaluated utilizing the PONS tool from 2015 – 2016. Statistical analysis was completed and overall the mean total score for patients with a nutritional deficiency was significantly higher than patients who did not have a nutritional deficiency.

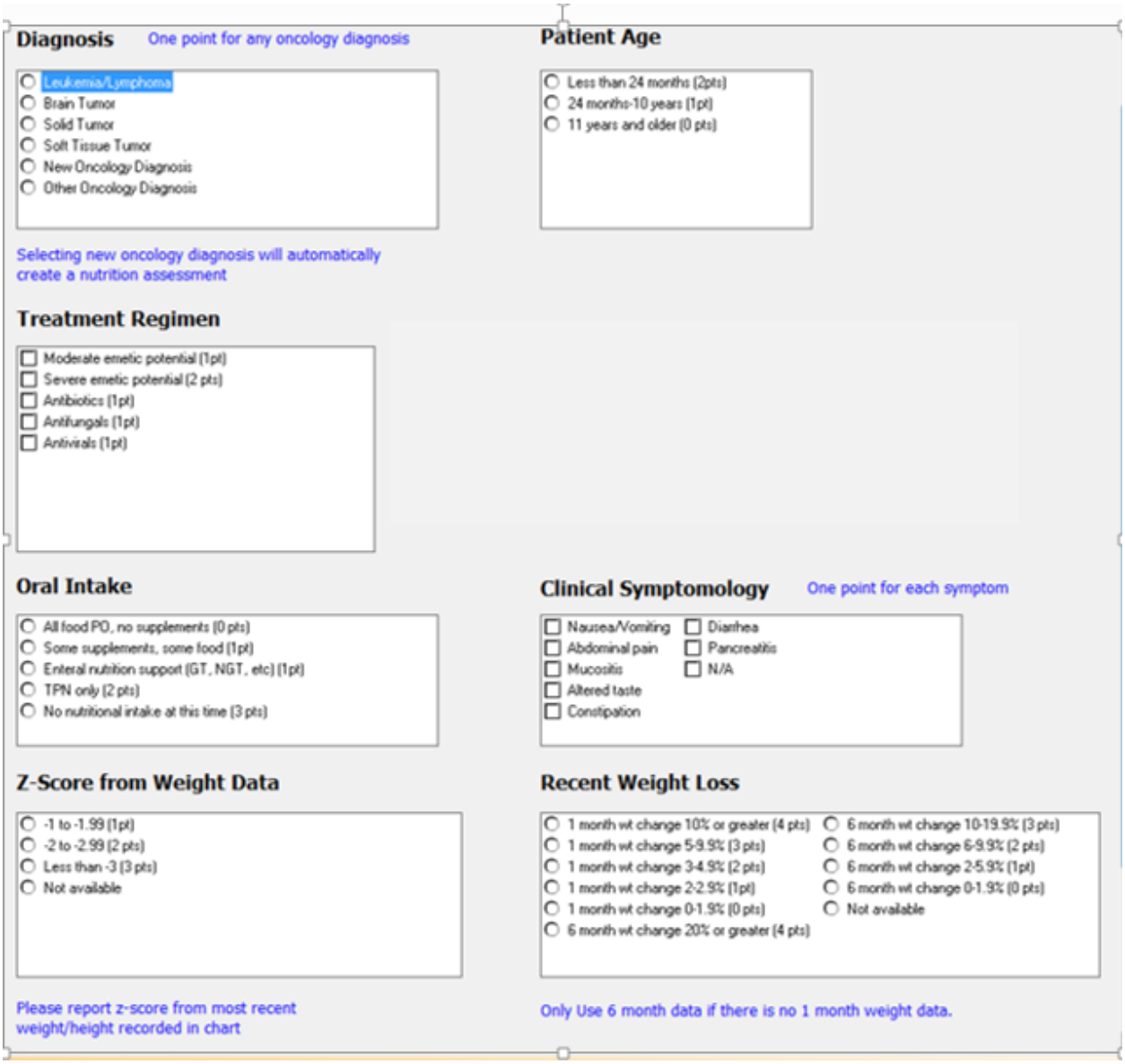
QI-Study

The significance of the PONS tool was that patients could be identified and evaluated on their specific treatment regimen for risks of malnutrition. Based on the total score the patient would be identified as mild, moderate, or severe risk for malnutrition.

(QI)- Act

The collaboration of nursing and the dietician provided a good method to proactively provide nutritional interventions when the risk for malnutrition was identified early in the patient's treatment regimen. The PONS tool could identify patients at risk and in the future could influence better outcomes in care if the patient's nutritional status is optimal during cancer treatment.

Oncology Nutrition Screen



<u>Abstract #71: (NUR-011-QI)</u>

Cruz D*. ***"Pain Management Protocol for Vaso-Occlusive Crisis in Children with Sickle Cell Disease in the Emergency Department at Nicklaus Children's Hospital".***

(QI) Introduction

Vaso-occlusive crisis (VOC) is the most common complaint for pediatric patients diagnosed with Sickle Cell Disease (SCD) in the Emergency Department (ED). Patients that present with the hallmark symptom of severe pain require aggressive and timely

administration of analgesia, such as parenteral nonsteroidal anti-inflammatory drugs (NSAIDs) and opioids. The National Heart, Lung, and Blood Institute (a subdivision of the National Institute of Health [NIH]) created an expert panel report in 2014 with recommendations targeting rapid assessment and initiation of opioids, effective opioid doses at frequent intervals, and a SCD-specific pain protocol. However, studies show that pediatric patients wait approximately 65 to 90 minutes for the first dose of parenteral analgesics when presenting to the ED with VOC despite NIH guidelines and best practices.

QI-Plan

Implement a pain management protocol for participants experiencing vaso-occlusive crisis (VOC) in the Emergency Department (ED) to improve patient self-reported outcomes (e.g., pain relief) and standardization of medication administration.

(QI)- Do

In October 2018, a clinical pathway for patients with SCD experiencing acute pain (≥5 on 0-10 pain scale) was created for the ED at Nicklaus Children's Hospital.

QI-Study

This quality improvement project analyzed medication administration times and patient self-reported pain scores pre-/post-implementation of the clinical pathway. The aim was to support the implementation of the clinical pathway by demonstrating improvements on the self-reported pain scores and standardization of medication administration including timely administration of the first analgesic.

(QI)- Act

From June 2018-September 2019, there was a total of 75 patient encounters, with patient's age ranging from 13 months to 20 years (mean: 12 years). When analyzing data from June-December 2018 with data from January-September 2019, parenteral administration of NSAID and opioids within 60 minutes of triage increased. Timely administration of the first parenteral analgesic improved self-reported pain scores. Also, there was a notable decrease in other important metrics such as length of stay and 30-day readmissions. This is significant because it supports the clinical pathway for pediatric patients diagnosed with SCD experiencing VOC in the ED.

Abstract #72: (NUR-012-QI)

Ferreyra G*; Perdomo M. ***"A Quality Improvement Project to Reduce Mother's Own Milk Administration Error Risk in the NICU".***

(QI) Introduction

Introduction of Mother's Own Milk (MOM) is a very important key component of an infant's nutrition, providing essential nutrients, antibodies and other factors important for growth and development. The NICU at Nicklaus Children's Hospital, is committed to continuously improve our qualitative measures to provide safely administration of Mother's Own Milk. However, during October 25th 2016, Nicklaus Children's Hospital NICU moved to the Advanced Pediatric Care Pavilion. The move presented some challenges, including but not limited to an expanded floor plan, new configurations for supplies, storage, and preparation of Mother's Own Milk (MOM) and lack of a defined and standardized process to handle Expressed Breast-Milk in the new setting. Without a process on how to handle Human Milk in the new setting, the neonatal population was at risk for breast milk errors.

QI-Plan

Aim:
A process that satisfies the target conditions, that is: A standardized high quality and safe process that is efficient and simple for the nurses to carry out, thus reduce risk for MOM administration errors by 20%.

(QI)- Do

Methods:
To address MOM with Fortification Preparation, the largest risk, according to the Failure Modes and Effect Analysis (FMEA) were:

- Standardized process for ensuring safe MOM handling
- Standardized preparation set up area that is conducive to the safest process possible
- Standardize Recipe guide

Key Driver Changes

Standard Set-Up

- Standardize supplies of each work zone station (most used items in counter tops)
- Quite environment signs
- Penguin Warmers in each station
- Labeling of refrigerators
- Delegation of stocking rooms to (CA)
- Standardization of recipe guide in Hospital Drive for easy access.
- Visual management of cabinets

- Bins/Bottles/Supplies for freezer allocated in freezer
- Educating staff of New Women and Infant Electronic System for MOM handling
- Limiting staff in rooms
- Zone identification on refrigerators

Standard Process for Safe Administration

- Computers for verification of orders in nourishment rooms
- Dedicated Milk Technician coverage (Began January 2018 for 4 days a week. Expanded to 7 days a week, 24-hour preparation coverage Mid-2019).
- New Electronic System for Breast Milk Handling (Integrated Mid-2018).

QI-Study

Results:
During Mid-2018, Quantitative Error Risk Reports identified four types of prevented errors; Expired bottles scanned, previously disposed bottles scanned, wrong patient scanned, and wrong fortifier scanned.

During 2018 (August through December)
Expired bottles scanned 2.7%
Previously disposed bottles scanned 13.10%
Wrong patient scanned 15.50%
Wrong fortifier scanned 0

2019
Expired bottles scanned 5%
Previously disposed bottles scanned 18%
Wrong patient scanned 23%
Wrong fortifier scanned 1%

2020
Expired bottles scanned 3%
Previously disposed bottles scanned 9.8%
Wrong patient scanned 8.5%
Wrong fortifier scanned 0.15%

These results demonstrate since beginning phase of the quality improvement project on 2018 until the end of the improvement on 2020, there was a 40% decrease for Expired bottles scanned risk, 45.5% decrease in previously disposed bottles scanned risk, 63% decrease in wrong patient scanned risk, 85% decrease in wrong fortifier scanned risk.

(QI)- Act

Based on these results, Integration of a Standardized Nourishment Room Set-Up, Utilization of an Electronic System for Breast Milk Handling, and a Dedicated Milk Technician, are powerful interventions to reduce breast milk administration error risk in a NICU. All infant healthcare settings would benefit in adoption of these practices to provide safe administration of Mother's Own Milk.

Abstract #73: (NUR-013-QI)

Lima-Keller N*; Suarez R*; Sztokfisz I; Sánchez A; Perez J; Cincotta C; Cortes I; Simon-Mengana K. ***"Horizon SharePoint Page".***

(QI) Introduction

Utilizing information technology is increasingly becoming a common practice in the healthcare environment. SharePoint sites are an excellent tool that enable nurses prompt access to valuable and up-to-date information at any time. Throughout the course of the Horizon Nurse Residency Program, different approaches have previously been used to distribute necessary information to the incoming nurse residents. Orientation schedules, information regarding the program, the organization, and unit specific curriculum and guidelines make up the necessary onboarding resources given to each new nurse resident. Although informative, previous methods were not the most efficient way to disseminate said information. In order to overcome the obstacles met with the previous methods utilized, the nurse residents of cohort 18 found it essential to work on a process improvement project that would eliminate cost and improve accessibility to pertinent information, resources, outcome data, and much more.

QI-Plan

- Create an easily accessible format for Horizon nurse residents to access curriculum, documents, and necessary resources when onboarding into the Horizon Nurse Residency Program
- Sustain an interactive SharePoint page where past, current, and future nurse residents can participate in open dialogue via a Horizon Chat platform, access current Horizon course schedules, review latest announcements, outcome metrics, current news, recognitions of their colleagues, images, educational videos, program overview, and much more
- Implement a Lean process to access pertinent and necessary information with long term organizational cost savings

(QI)- Do

- Ask
    - First critical step
    - Using PICO to formulate a clinical question

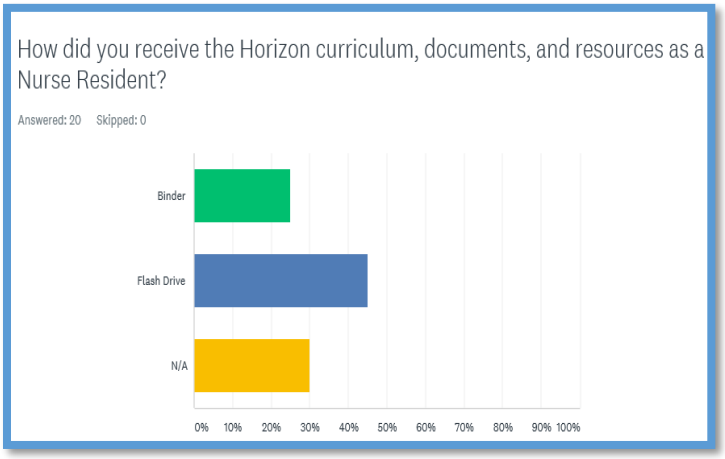
        - Population and Problem: Horizon Nurse Residents and the distribution method of the program training materials
        - Intervention: Cultivating the most efficient and effective method to distribute these materials via a SharePoint page
        - Comparison: Binders and flash drives given to previous cohorts
        - Outcome: Easily accessible information readily available at the fingertips of all nurse residents
- Acquire
    - This process involved performing a literature review of the distribution methods of other programs and the possible use of a SharePoint page as a method of distribution in these other programs
- Appraise
    - Appraising the evidence gathered for validity and practicality, while focusing on the outcome and impact it had on the recipients of these materials
- Apply
    - This step included brainstorming ideas as well as filtering through the materials previously given
    - Applying this information into a SharePoint page that would be easily accessible and clearly constructed to allow its users to find information in an organized manner
- Assess
    - In order to assess the previous steps and the outcome of our clinical question, a survey was distributed to current and past cohorts, focusing on comparing this method to previous methods of distribution

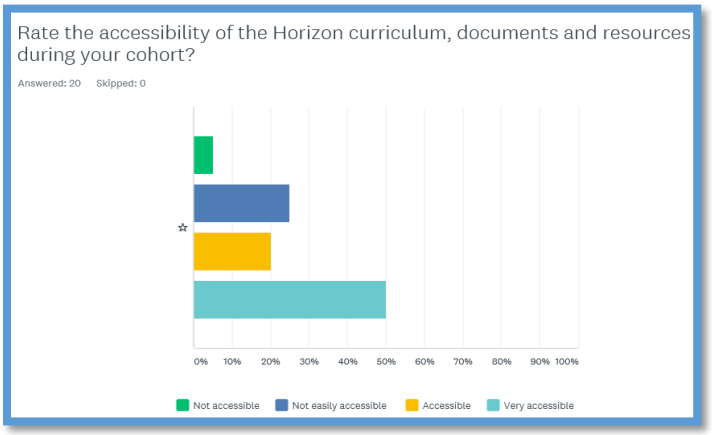
QI-Study

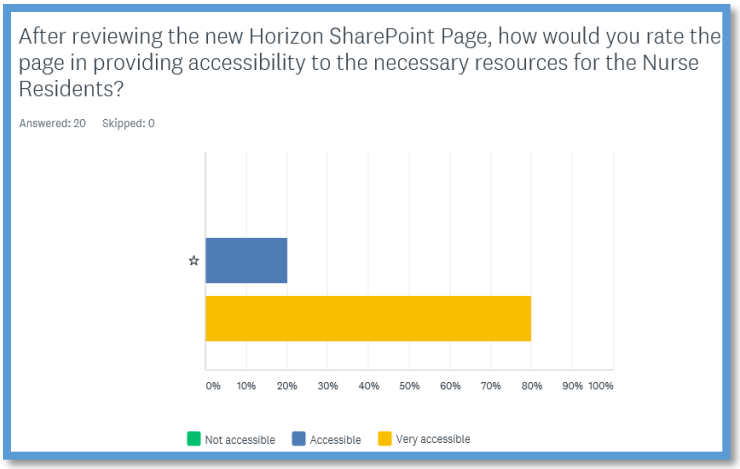
The tools that had been used previously to provide Horizon nurse residents with the necessary information were a binder, which later evolved to a flash drive. Prior barriers encountered with the binder, was the size and weight of it and the USB's were blocked, unable to access on hospital computers. Another disadvantage of the use of USB's, was the cost. There is an estimated expense of $700.00 for every 200 USB's purchased. Therefore, Cohort 18 decided to develop a cost effective and sustainable method of providing all the necessary information for future Horizon nurse residents. In order to evaluate the effectiveness of the new proposed Horizon SharePoint page, a survey was distributed to past and current nurse residents, as well as content developers, such as faculty and program leadership. This survey consisted of questions that identified each individual's form of receiving information during their nurse residency cohort, accessibility to the information via the format they were provided during their onboarding phase, along with weighted answers of how accessible they perceived the information to be for the current and future cohorts utilizing the newly developed Horizon SharePoint page. Positive feedback regarding the new Horizon SharePoint page was overwhelmingly provided by the survey participants as well. After carefully analyzing the responses, the data supports that the new Horizon SharePoint page is a more accessible resource for the program content.

(QI)- Act

Nursing residency programs are transforming nursing practice by arming new graduate nurses with the tools and education necessary to provide safe, evidence based, high quality nursing care. Easy access to education and training materials is a key part of ensuring new hires are able to take full advantage of the program. Constructing a SharePoint page that allows nurse residents access to information instantly at home or while at work will bridge the informational gap that previously existed related to receiving pertinent program information via a binder or flash drive. The site also serves as a useful resource for the float pool staff, whom can now easily review unit specific policies, procedures, and curriculum content when floated to various units throughout their orientation. Although previous resources provided similar information, nurses encountered the obstacles discussed when trying to access them. A collaborative effort between leadership, previous cohorts, and current nurse residents to create a SharePoint site for training materials has shown to be an effective, cost-saving, LEAN solution to eliminating previous barriers encountered with information distribution within the Horizon Nurse Residency Program.







Nicklaus Children's Hospital

HORIZON
Nurse Residency Program

ACCREDITED PRACTICE TRANSITION PROGRAM WITH DISTINCTION AMERICAN NURSES CREDENTIALING CENTER

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Annnouncements

Welcome Horizon Nurse Residents - Cohort 19!

Cohort 18 graduation and Poster Presentations - October 19, 2020

Program Metrics and Outcomes

Horizon Calendar