The Perspective

A quarterly newsletter published by the National Med-Peds Residents’ Association in collaboration with the Med-Peds Program Directors Association & the AAP Section on Med-Peds

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Dear Med-Peds family,

With spring fast approaching, we are well on our way into the heart of 2022, and it is hard to believe that means the end of this academic year is visible on the horizon. In a year full of many successes, we have also faced many setbacks which, when coupled with the undue stresses of medical school and residency during a seemingly never-ending global pandemic, unfortunately make this particular time of year weigh heavily upon many in our community. The physical and mental exhaustion of our careers, the difficult balance between professional and personal life, and the tragedies of suffering and oppression experienced by so many around the world all too often seem unbearable. Our mission as the leaders of NMPRA is to prioritize the wellbeing of our members and offer support in whatever ways may be possible. We encourage you, as always, to reach out to any one of our Board members with questions or concerns, and to take advantage of support services offered by your individual programs and institutions. We also want to highlight the Physician Support Line, a national, psychiatrist-supported resource which offers free and confidential peer support to physicians and medical students across the country. You can access their services simply by calling 1-888-409-0141 or by visiting physiciansupportline.com.

Despite the innumerable obstacles placed in our paths over the past several years, our community has demonstrated great resilience in the face of adversity, and this strength is something we strive to highlight and celebrate. In fact, this year marks the 55th Anniversary of our founding as a specialty, and we can think of no better time to reflect on and take pride in our unique history. Originally approved as a combined specialty by both the ABP and ABIM in 1967, Med-Peds has since grown to represent over 79 residency training programs and over 8,000 graduates working in a variety of healthcare settings across the country. We have national representation not only in our own organization for residents and medical students, but also through the Med-Peds Program Directors Association, the American Academy of Pediatrics’ Section on Med-Peds, and the American College of Physicians-American Society of Internal Medicine. We care for patients across the life spectrum and bring unique perspectives to the fields of primary care, hospitalist medicine, subspecialty care, and our particular niche of transitional care. We look forward to celebrating our specialty and its achievements throughout the year and with the culmination of our festivities at the annual National Conference in Anaheim, CA this fall. We hope you can join us!

Our Board of Directors has been hard at work to bring you lots of new content focused on medical education, professional development, and national advocacy. Keep an eye on your inboxes for the latest information on the Educational Webinar Series, Career Fair, and Advocacy Lectures all happening this spring. We are also excited to be bringing you new merchandise for purchase through our official online shop hosted on the Bonfire platform; please check in regularly at www.bonfire.com/store/nmpra-online-shop to ensure you do not miss out on the latest apparel and accessories for yourself and your Med-Peds colleagues. Bulk orders are also available for all items – please email president@medpeds.org with your requests!
This time of year also represents a natural transition in our seasons which is reflected in the annual transitions which occur in our own organization. Our work on a national scale would not be possible without the dedication of an incredible Board of Directors, and we have been so lucky to have an amazing legacy of hard-working residents who strive to make NMPRA better with each passing year. Applications for the Board of Directors positions, both Directorships and Executive Board memberships, will be opening in the next several months and dispersed via our listserv. We encourage you to consider getting involved in the Med-Peds community on a national scale by serving in a NMPRA position. All of our current members are available via email (found on our website) to answer questions about their positions and share with you their experiences in NMPRA. We look forward to reviewing your applications!

Yours in Med-Peds,

Sophia Urban
NMPRA President 2021-2022
president@medpeds.org
Internal Medicine/Pediatrics PGY-4
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2022 is a new year. It presents with similar challenges, but hopefully renewed optimism.

We are starting to see in-person events. This includes ACP 2022 which will be in Chicago, Illinois. The Section on Med-Peds Session will be highlighting “Transitions of Care in Sickle Cell Anemia”. This will be followed by a Med-Peds reception from 6-7 PM in the Marriott. The Section on MedPeds and myself are so excited to be able to meet with everyone in Chicago and to chat about what you are doing. If you have not already done so, please register at https://annualmeeting.acponline.org. In the Fall (hopefully in-person) during AAP National Conference October 7-11, 2022 in Anaheim, CA we are offering a 3-hour section program plus a reception and as well as cosponsoring with the Counsel on Children with Disabilities on their topic of “Transitions of Care”. I hope you will attend these sessions, which are focused for MedPeds physicians.

The Section on Med-Peds scientific abstract call will open February 25 and closes April 22. You can submit a case or quality improvement abstract. All abstracts are graded by a panel of judges comprised of members from the Section. You can earn an invitation to show a poster of your work and maybe even win a $500 prize. The call will be sent via the SOMP list serv or check the AAP National Conference website.

During the upcoming months, residency programs will have completed fellowship matches, interviewed and ranked applicants for their programs, and prepared program evaluations and improvements. Well-being remains a priority for the Section of Med-Peds. If you have not already done so, the tips for wellness and preventative care can be found on Med-Peds AAP collaborative site (Section on Medicine-Pediatrics (Med-Peds) Home [aap.org]). I have been following a couple of the domains of wellness, including trying to read more. A book that I found inspirational was “When Breath becomes Air” by Paul Kalanithi, which highlights his journey with Stage 4 lung cancer and the meaningful or virtuous aspects of life. I always welcome other suggestions!

As part of our mission, the Section is working with the AAP to find ways to engage our future Med-Peds graduates as well as current Med-Peds members. We want all Med-Peds to value membership and to have access to the Section; we want to be a place to bring ideas and to discuss topics important to members. This also includes our continuing discussions with NMPRA for DEI initiatives and mentorship for underrepresented students and Med-Peds residents. Let us know some of your ideas for educational webinars, virtual gatherings, or other activities.

I always think that it is important to disseminate medical knowledge and interesting case reports. For those looking to publish scholarly activity consider a submission to Cureus and the Med-Peds Academic Channel (www.Cureus.com). The submission process is free if you submit to the channel as opposed to the general Cureus journal.
Stay safe! As always, if you have any topics that you would like the SOMP to address, please feel free to reach out.

Jayne

Jayne Barr MD MPH FAAP FACP FHM
Chair, AAP Section on Med-Peds
Introducing the new Med-Peds Academic Channel (MPAC)!

- Forum created to promote scientific advancement and dissemination of knowledge in the field of Med-Peds

- Peer-reviewed publications on various topics unique to the practice of Med-Peds

- Open to submissions including original articles, review articles, case reports, technical reports, editorials, and posters!!

Check out more details on the website:

https://www.cureus.com/channels/med-peds

**Please submit under “academic channels” to submit to MPAC rather than to cureus.com**
Lessons Learned From My Experience With Street Medicine

Richa Bisht, MS III
USF Health Morsani College of Medicine

When I joined Tampa Bay Street Medicine (TBSM) two years ago as a first-year medical student, I never could have imagined its profound impact on my life or how it would shape the type of physician I want to be. From the first time I did patient intake at our downtown clinic for the homeless in February 2020 to the hygiene kit distribution I helped organize last week, partaking in street medicine has challenged my preconceived notions about homelessness and reaffirmed my personal mission of increasing access to care for underserved populations.

It can be frighteningly easy to become desensitized to the hardships faced by the homeless, which makes it all the more common to believe in stereotypes and pass quick judgements. Street medicine consists of providing medical care to people on the sidewalk, in open parks, and other non-traditional settings, which forces us to see the difficulties our unsheltered patients encounter in their everyday lives. Working as a clinic coordinator taught me that our patients are not the negative labels that surround them – they are people with stories, challenges, and dreams just like everyone else. Many have overcome difficult odds and still find themselves unhoused, not because of moral failings or bad choices but instead due to mental and physical health problems beyond their control.

Street medicine has also made me reconsider previous assumptions about what homelessness “looks” like: some patients I’ve met look younger than me, and some blend perfectly into the crowd of people enjoying the sunset at a park. Many patients are dealing with crippling mental health issues, disabilities, or language barriers on top of the stigma of homelessness, which makes it harder to navigate a healthcare system that’s already tough to access.

This heavily marginalized group needs targeted assistance and yet is all too often forgotten, demonstrating the need for street medicine as a tool to close the gap between our community’s most vulnerable members and crucial medical services. By easing the burden of illness, we can remove an obstacle on their way to achieving stability, housing, or employment. When incorporated into medical schools, street medicine can teach lessons that aren’t always covered in class, including harm reduction techniques and how to work in low-resource settings. Additionally, it can broaden perspectives and empower students to become more empathetic, compassionate, and caring physicians.

Through TBSM, I’ve had the honor of working with an amazing team that inspires me every day, faculty advisors who are incredibly dedicated to our shared cause, and community
partners whose steadfast support has enabled us to continually expand our reach. My favorite part of volunteering is getting to connect with patients and hear their stories. I love being able to help them regain a sense of comfort and dignity through something as simple as providing a clean pair of socks or facilitating a clinic encounter where they can finally receive the medications that help them manage an otherwise debilitating condition.

Despite the limitations to medical student volunteering during the COVID-19 pandemic, we saw over 1000 patients, logged more than 1500 service hours, and supplied countless masks in the first year of the pandemic. We’re leading the second needle exchange program in Florida and have distributed hundreds of Narcan kits designed to reverse opioid overdoses. We also expanded our goals as well as our team by starting a new clinic serving the refugee population of Tampa.

Although the COVID-19 pandemic has exposed the weaknesses in our socioeconomic structure, it has proven the strength of street medicine as a way to increase access to medical care for the homeless, especially during times of crisis. I am hopeful for the future and look forward to seeing how street medicine can be used as a tool to develop innovative solutions that can help us meet our patients’ changing needs.
“So, this will make him better?” asked Mrs. Smith* with tears in her eyes. She was surrounded by the large care team outside of her fifteen-year-old son’s room in the PICU during morning rounds. Carson had been diagnosed last year with a rare Stage IV cancer. His treatment options had been limited due to hypersensitivity reactions to the chemotherapy and the complicated anatomical distribution of the tumor. Despite continuous efforts to investigate other treatment options, there was nothing else that could be done at this point to reverse the progression of his illness. Carson was on a ventilator and surrounded by a sea of IV poles. “This will make him more comfortable”, the pediatric oncologist gently replied. I could tell this physician was carefully considering her word choices in response to this mother’s question. Mrs. Smith blinked several times and looked down at the floor. Today’s discussion focused on the balancing act of titrating Carson’s medication regime to manage his increasing pain. Mrs. Smith was very resistant about making him too sedated. She stated she did not want to lose the ability to communicate with him.

This tragic situation had been further complicated by Carson being unaware of the current extent of his terminal illness. Due to his endotracheal tube, Carson’s communication was limited to slight nods and blinking through the alphabet to spell out words. He had been wanting to know if his tumor had changed. However, Mrs. Smith did not want him to know that all treatment options had been exhausted. She continued to tell Carson that his lungs were just sick, and they would be going home soon. She believed it was “cruel” to tell someone they were dying when they could not do anything to improve their outcome. Nurses and techs continued to come in and out of Carson’s room throughout the day and night. It is heartbreaking that all these interventions were being done for him, but he was not able to really know what was going on. Mrs. Smith said that Carson had said he did not want to die. However, this was before he knew the most recent extent of his tumor progression.

From my perspective, it seems cruel to not let someone know the truth about their own body, especially when they have continued to ask questions about what is going on. Since Carson was under the age of eighteen, Mrs. Smith oversaw all his healthcare decisions. Despite Carson’s worsening symptoms and increasing pain, Mrs. Smith had maintained his full code status and wanted to continue all supportive measures. “We just need to hang on until there is a cure. And then, he can live a normal teenage life.”

It is understandable to try to reach for any glimmers of hope in this devastating situation. However, it felt like we were losing sight of the patient we were supposed to be treating. Instead of focusing on Carson, Mrs. Smith was center stage. This experience emphasized to me the nuances that exist within the field of medicine. The exact word choices can be so important in hard conversations with family members. It would be nice if there was a magical standard protocol for how to handle all the facets of end-of-life care. Just follow this algorithm and everything works out well every single time. Unfortunately, things are not black and white in medicine. It can be a very gray world.
Being on the healthcare team allows our primary focus to be the objective data. We can talk about trending lab values and repeat X-rays and CT scans. However, the family members must balance the slew of medical words and acronyms with their own collection of memories of their loved one. This is Mrs. Smith’s child. She had soothed him in the night after a bad dream and dropped him off at school. She knew his favorite movie and his favorite pair of shoes.

Determining how long to fight and when to let go is unfathomable for a parent to have to endure. As a future physician, I want to be a supportive partner for families when they are having to make these decisions. We must be patient throughout the process. We can take a deep breath and focus on sharing evidence-based practices documented in the medical literature. Our duty is letting the family know that our priority is taking the best care of their loved one.

*Names and identifying details changed to protect patient confidentiality*
A letter to the girl who lost her sister and father

Megan Barnes, PGY-2  
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Dear sister,

I'm sorry.

This isn't fair.

I wish you had gotten to say goodbye to your seven-year-old little sister and your dad. I can't imagine the heartbreak you are experiencing, and I know you're in pain. You're probably overwhelmed with so many emotions right now, and it's okay if you feel confused, lost, sad, fearful.

We never got the chance to meet. I was one of the doctors taking care of your sister in the hospital for three months. Throughout all of her chemo and complications, she was a fighter. I wanted to share the glimpses of beauty she shared with us at the hospital.

You probably know this--she had the cutest smile and laugh. She had such a creative side too! She made bracelets for some of the staff and colored pictures when she felt well. She filled her room with art! And she was so strong! Despite hit after hit on her body, she fought to stay alive. And by her side, the entire time, were her parents. They came in every day, and even if it was a bad day, they spoke of hope and the drive not to give up. And they were indeed some of the most compassionate people I've met in the hospital. Your mother and father thanked us profusely every day for even the smallest of acts. I left the room feeling uplifted and hopeful when I thought that was my job for families.

One of the most brutal consequences of this pandemic is that while your father was visiting her, he unknowingly passed on Covid virus to her. Seeing her was an act of love; a seven-year-old shouldn't be alone in the hospital. He was a good father. I am furious and sad about this, and I want you to know it was no one's fault. Sometimes bad things happen, and we can't predict them; we learn to cope with them.

I am sorry you never got to say goodbye to her or your dad. I am sorry your dad never got to see his children one last time. I have cared for many people with covid these past few years. I can confidently say that both of them were surrounded by caring people who helped them peacefully go when their time came. Our hearts ache for them and for you.

Now let me tell you about your mom. Your mom is one of the most extraordinary women I've had the pleasure of meeting. Her hope, resilience, compassion, and fortitude have shown in the darkest of circumstances. She's going to fight for you and your three siblings, and she'll never give up on you. I am sure you will inherit some of her loving compassion and grow to be a remarkable woman like her. Be kind to her.
This is going to hurt. Recovery will be slow, and you will not function normally right now. Eventually, you will recover and experience joy and happiness again. This pandemic will end, and we will all breathe deeply again.

I will never forget your sister’s smile and laugh. I hope you hold on to her memory of her smile and laughter and remember her as a fighter. Take this time to draw close to your family. Grieve together, love together, and, one day, slowly move towards feeling okay together. You are courageous, and you come from a stunning family. Know that your family has touched so many lives here at the hospital.

Peace,

A friend
To My Cousin

Irene Martinez, MS IV
Rutgers New Jersey Medical School

I always thought I would see your face in the inpatient psych unit. Every tall, handsome, young, tan-skinned man became you, for just a second. I would double-take every time to see if that new admit was you and my heart would sink every time it wasn’t. If you were there, it meant you were off the streets and out of harm’s way. We would know where you were, we would have an answer. And yet, seeing you there meant you would need to face your demons all at once. The hallucinations, the psychosis, the addiction, the withdrawals. You were sick, but no one was ever able to tell you or help you in the way you needed.

You were “the bad one” growing up - the one who got into trouble, who couldn’t follow directions. You stayed out late and got into fights. You fell into alcohol just like your father, just like so many of our family members whose struggles we still refuse to acknowledge to this day. The drugs and crime followed soon thereafter. But who is really to blame? You were just a child struggling with your inner demons, screaming for help. The only ears that heard you were the streets’ and they took you in, as they do with so many other lost kids.

When I got the call saying you had been taken to the hospital, I felt the adrenaline kick in. I was in doctor mode: locate the patient, stabilize them, send them home. "He had a heart attack." My mind immediately created differentials: overdose, cocaine-induced vasospasm, drug withdrawal. I listed all the diagnoses that could present with chest pain, but I refused to consider anything that would kill you. You always felt immortal. As we scoured the ED track board, the unnamed 30- yo in trauma bay 05 called to me, CC: cardiac arrest. And then we saw the note, intubated on the field, multiple rounds of CPR, asystole, dead upon arrival. My heart sank, just as it had all those times on the psych ward. If it was you, you would be off the streets, out of harm’s way. We would know where you were, we would have an answer. And yet, if it was you, it meant facing the harsh reality that the addiction had finally won. I pulled every string, found anyone and everyone in the ED to check on you. The tunnel vision set in; I had a new task: identify this body. The nurse said no one ran to your bed because you were already dead. The tech said that the tattoos we described matched the ones on your arms, she thought anyway. The resident said that unidentified bodies are processed by the police, not the hospital. The student who has found the morgue for us said it was closed on the weekends.

I hadn’t seen you in years, 5 to be exact. I scour my memory to think of how you were that day, and nothing comes to mind. I don’t know whether to be relieved or more hurt. You were always chill, so cool, calm and collected. I knew what you could do and what you had done. I heard the whispered stories from my Abuela, and other family. And yet, when you were standing in front of me, none of that mattered. All that mattered was the massive buffet of Thanksgiving dinner in front of us, the usual banter between the adults that we had all seen growing up, your dad’s meatloaf, and the same holiday stories that were shared every year.

And now, you’re gone. Lying in a dark, quiet morgue. Alone. Alone, just as I’m sure you’ve felt many times before, for years. I pray you have peace, that the voices in your head are no longer tormenting you, that the grapples of addiction are no longer suffocating you. We always talked about all your mistakes, but did we ever really consider what you were facing.
When I go back to the hospital, and I see another human struggling with addiction, with their demons, lost to drugs and alcohol, will I think of you? Will I still double take at every tall, handsome, young, tan-skinned man who comes in with hallucinations? With drug-use? With chest pain? Will I finally have the guts to stand up to those around me when they call them "just another druggie", "good for nothing", "a complete psycho". Will I be able to remind them, that this is someone’s child, someone’s best friend, someone’s cousin? Because I admit, I haven’t been able to so far. So now that you’re gone, does that change anything? I don’t know.

I don’t know anything.
All I can do is hope and pray you’ve found peace. That you are no longer hurting.
And I’m so so sorry.
I love you; we all do.
Signs: Not Superstitious, just a Little-stitious

Sarah Calvert, PGY-3
Prisma Health Internal Medicine-Pediatrics

During the course of our medical training, we are instructed in a litany of ominous signs-- the mottled extremities that portend imminent passing, the triad that precedes cerebral herniation, the sudden crop of seborrheic keratoses that heralds a GI malignancy. Each of these appropriately increases the tone of certain sphincters for any provider who should happen across them. So too, there are signs that we learn after leaving the classroom. Included among these are the “crossed leg sign.” This semiotic tidbit was taught to me by an ED attending who, gesturing across the ED towards our next patient, noted, “No one in any significant distress sits with their legs crossed. I would bet we will be sending her home.” Interestingly, a recent study suggests this may actually be an evidence-based statement.¹

Along a similar vein, the “Chick-fil-A (CFA)” sign is a positive prognosticator that has been added to my repertoire since starting residency. The CFA sign holds that any pediatric patient with an empty box of Chick-fil-A nuggets bedside is soon to be homeward bound. I am skeptical that the studies for this sign will be as soon forthcoming, but have, anecdotally, found this to be a trustworthy indicator.

However, what deserves a few words are the signs in medicine to which we subscribe despite the fact that they have no scientific basis whatsoever.

The senior resident has worn his lucky socks for call day. With scrub leg hiked up, he shows them off in the workroom. The team nods with approval at this talisman to ward off a deluge of admissions. The attending pokes her head in for a final word before stepping out for the evening. Looking around, she wishes all a “quiet night.” Said senior resident offers a side eye to the door as it closes behind her. “The audacity-- using the ‘Q’ word!” each of the residents think to themselves as the keyboard clicking resumes.

A code is called overhead.

"Let’s hope that’s not one of our guys." The resident remarks as he and the intern stand, slinging their stethoscopes around their necks. As she trails the resident out of the workroom, the intern raps her knuckles against the table. She frowns to herself as she realizes the tabletop is not actually made of wood.

Hours later, the resident is leaving from his fourth code of the night. He walks out of the ICU beside the pharmacist who dutifully pushes her code cart ahead of them. "Should have known with this full moon," she offers, as if it is the most logical thought in the world. Post-call the following day, the resident takes his less-than-lucky socks out of the dryer and shoves them to the back of his drawer.

Bedside an ICU patient, I am reviewing our care plan with my patient’s primary nurse. Thankfully our patient is much recovered from her initial presentation, which featured profound bradyarrhythmias in the setting of acute myocardial infarction. As I lean up against the portable computer in her room, reminiscing about how much she has improved, I note a
syringe taped to the monitor. A premeasured push of atropine stands at the ready, like a good soldier. Over the preceding week, this syringe had been plucked from the monitor and nearly pushed into her IV on countless occasions. Each time her own sympathetic system had rallied just enough to stave off the severe bradyarrhythmia and cardiac arrest that lurked just beyond it. Thumbing the syringe, I smile. "I think we can get rid of this now." Somewhat sheepishly, her nurse replies, "Let’s leave it just a little longer."

Despite all the medical training, still we pick up found pennies. Across all shifts, around every unit, our humanity takes the liberty of poking its head out of our tempered academic armor to offer its two cents. An evidence-based care plan is wonderful, but the addition of a little "good juju" surely could only help, right?

Just ask any resident what color their cloud is. Shortly into their training, they will have been assigned a hue along the grayscale. Deny as they might, a resident known to bring a black cloud will likely never live down this association. The presence of a resident with a white cloud will inevitably bring a sigh of relief to their co-resident on call that night. Though no studies have demonstrated statistically significant variability in the volume or acuity of admissions between call shifts, the cloud color of a resident will remain a well-accepted predictor of the quality of a call day.

Though we love our double blinded trials, still our intensive care unit lacks a room number 13. How can we, as providers, reconcile this paradox? Though we do not consciously assess the weight of these less-than-scientific signs and wonders, still my day-to-day experience informs me these superstitions remain alive and well.

The fact is that we remain human. As humans, we long for control. In a wildly unpredictable workplace where morale and acuity can swing like the tides, we yearn for a tide table. If we participate in these practices, are we morally culpable? Have we thrown our sound academic practices to the winds? Perhaps this sense of agency amidst the hurricanes that blow in unexpectedly really is contributing to our resilience. Though we consciously fool ourselves into this sense of control, judge us not until you have been rained upon with admissions. We cope. We remain, if not superstitious, at least a little-stitious. After all, watching the phases of the moon is healthier than many other alternatives.

In Parallel
Tessa Adzemovic

71 year old Veteran with COVID-ARDS admitted, intubated, sedated
His course complicated by septic shock and renal failure
Treated with steroids, remdesivir, tocilizumab.
A persistent pressor requirement led to goals of care,
After a lifetime of fighting, he’s DNAR/DNI.

Family finally allowed to come to the hospital, They say he wouldn’t want to live like this.
Comfort measures begin, but the only thing that looks comfortable,
Is his patchwork quilt.
His breathing slows, the rattling begins, then
No heart, nor lung sounds, no response to pain.
Time of Death is 13:15, two days before the year turns.

4 year old with neuroblastoma on her fifth round of dinutuximab
Her course complicated by febrile neutropenia and refractory pain
Treated with vanc, pip-tazo, a morphine PCA.
Her hair falling out led to online blogs, a community,
A lifetime of fighting, she’s a little weaker each round.

Family always at the hospital, wondering if her brother feels abandoned.
Everyone calls this “aggressive care”, but the only other option,
Feels like giving up.
It’s her last round, and she’s doing better, They say
No longer tachycardic, nor tachypneic, the pain fades away.
Then the fever finally breaks,
two days before she turns five.

The long awaited words are spoken,
"she can be discharged".

Taps is performed,
the age-old trumpets sound.

Pom Poms shake,
a guitar is played.

Civilians stand
with their hands on their hearts,

Nurses and residents
make a tunnel with their arms,

Veterans give the salute,
And sing their goodbyes,

His tired body put to rest.
To this 'patient', finally a child.

It's time to go home, for good this time.

It’s time to go home, for good this time.

Author’s Note:

In this piece, I used a form of the contrapunto structure to contrast the story of a Veteran who passed away from COVID-19 in the early days of the pandemic with the story of a child who had completed months of chemotherapy. As a resident in internal medicine and pediatrics, I have had the great privilege of taking care of a diverse array of patients. From month to month, a Med-peds resident can go from the adult liver service to inpatient pediatric pulmonology to ambulatory care. What accompanies this wide spectrum of pathology is a wide variety of patient outcomes. We spend our days and nights with families who are grieving, celebrating, saying goodbye, or welcoming a new normal. This poem describes two rare instances where teams of providers were able to come together and say goodbye to the patients described. In both scenarios, the patients left the hospital with significant “processions”. To bear witness to both was extraordinary, and to be a small part of their lives, an honor.
Breaking Bones and Breaking Biases: A Case Report and Quick Review of Hypophosphatasia

Hannah Shin, MD
University of South Florida

Introduction

Hypophosphatasia (HPP) is a rare genetic disorder caused by loss-of-function mutation(s) of the ALPL gene that encodes tissue-nonspecific isoenzyme of alkaline phosphatase (TNSALP), a ubiquitous enzyme in the alkaline phosphatase family. There is variable clinical presentation, age of onset, and severity. We present a case of a patient with previously diagnosed odontohypophosphatasia, an isolated dental form of HPP, with concern for worsening disease. We discuss the clinical presentation, diagnosis, and management.

Case Description

A 15-year-old female presented for second opinion of previously diagnosed odontohypophosphatasia after undergoing recent surgery for a left femoral neck stress fracture. She was active since childhood and she recently joined the cross-country team running 20-30 miles a day. She was born term with no complications, with normal growth and development, except premature teeth loss at 18 months which prompted her workup. She had a nondisplaced ankle fracture at 9 years old while performing hurdles that required no surgical intervention. Permanent teeth erupted at 7 years old and remained. She took a daily multivitamin. There was no family history of recurrent fractures or bowed legs. Physical exam showed benign findings except residual tenderness with range of motion of her left hip.

Childhood work-up showed alkaline phosphatase (ALP) levels ranging between 77-101 U/L from 3-12 years old (110-150 U/L depending on age). Calcium, phosphate, magnesium, parathyroid hormone (PTH), and 25 and 1,25 vitamin D were normal. X-rays showed no skeletal deformities. Repeat workup initially showed a normal ALP 101 (35–104 U/L). Serial monitoring showed consistently low ALP average 31 U/L. Calcium was low-normal 9.0 mg/dL (8.9-10.4 mg/dL), B6 levels were elevated 75.8 µg/L (2-30.8 µg/L). All other labs, including PTH, 1,25 vitamin D, thyroid stimulating hormone (TSH), free thyroxine (T4), sedimentation rate, anti-nuclear antibody, and urine calcium, were normal. Dental films of alveolar bone and tooth appearance were normal. DEXA scan of the spine showed lumbar spine Z score -0.8 and normal bone mineral density for age. Skeletal dysplasia genetic panel revealed a variant of uncertain significance (VUS) in the ALPL gene - c.1208G>A(p.Ser403Asn).

Discussion

HPP is a multisystemic disease with dental, skeletal, muscular/rheumatologic, and renal manifestations. There is heterogenous clinical presentation and symptoms can be nonspecific
such as delayed developmental milestone and fatigue. There is a wide spectrum of severity, from transient to life-threatening disease. It is classified by age of onset: perinatal, infant, childhood, and adult. Milder forms often present with premature teeth loss as was the case with our patient. Serum ALP levels will be consistently low. Normal ranges must be adjusted for age and sex; levels can be elevated and appear inappropriately normal in pregnancy, fracture, and liver disease as seen in her initial normal ALP level after orthopedic surgery. TNSALP inactivity leads to accumulation of pyridoxal 5'-phosphate (PLP), the principal circulating form of vitamin B6.

In this patient, HPP was suspected due to her premature tooth loss, multiple fractures, consistently low ALP levels, and elevated B6 levels. There is a new recombinant enzyme replacement therapy called asfotase alfa (Strensiq™) that has shown to improve quality of life and increase survival in pediatric HPP, which is more often severe. Common side effects include injection site reactions and lipodystrophy which can be dysfiguring. Given this patient’s intense exertion in the setting of a growth spurt and otherwise good functional status, the family decided to continue serial monitoring and supportive care.

Conclusion

This case raises awareness of the importance of recognizing low ALP in the right clinical setting as it does not raise the same level of alarm as high ALP. In addition, premature teeth should prompt HPP screen using ALP and/or serum B6 levels. Early diagnosis and multidisciplinary care may improve quality of life of patients of all ages.

Take Home Points

HPP is a rare genetic disorder in TNSALP that causes a wide spectrum of clinical variability with dental, skeletal, muscular/rheumatologic, and renal manifestations. Symptoms with a low serum ALP warrants further workup to discuss need for treatment and family planning.
More Than Meets the M.A.I.

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Introduction

E-cigarette induced lung injury is now a well-known risk to the multitude of vaping products that rapidly rose to popularity over the past few years, but what is still not well known, is the other potentially dangerous outcomes of e-cigarettes and vaping.

Case

A 31-year-old woman with past medical history significant for sickle cell disease with frequent acute pain episodes, chronic pain on oxycodone with indwelling chest port, and recent hospitalization for severe acute pain episode and pseudomonas bacteremia complicated by pneumonia who was instructed to present to the emergency room by her primary care physician after a blood culture drawn during the last hospitalization resulted positive for acid-fast bacilli (AFB) on fungal culture. Social history significant for five to nine cigarettes a day and daily vaped marijuana. Exam was significant for a temperature of 38.2 °C, heart rate of 115, and blood pressure of 96/58. On physical exam the patient was anxious and fidgeting due to pain. Blood cultures on admission grew Pseudomonas aeruginosa and the fungal cultures from the last admission speciated into Mycobacterium avium-intracellulare (MAI). Her chest port was removed and she was started on an extensive antibiotic regimen through a peripherally inserted central catheter placed after bloodstream sterilization.

Discussion

Since MAI infection is not a reportable disease, exact incidence and prevalence is not accurately known. However, one study reviewing medical claim diagnostic codes across the United States, found the prevalence of MAI annual infection rate increased from 2008 to 2015. The most common patient presentations for MAI lung infections are in those with underlying lung pathology and in elderly females. However, our patient is a young female without underlying lung disease and a history of vaping. We hypothesize the patient’s vape usage damaged her lungs and increased her susceptibility to this rare infection.

Not much is still known about vaping induced lung injury. Studies have unveiled the link between the pathological chemical-induced pneumonitis and the presence of vitamin E acetate in THC-containing e-cigarette oils suggesting marijuana oil vapor is the main culprit. However, the exact mechanism is still yet to be uncovered. What we can say for certain is that inhaled vapor does cause pulmonary injury as well as immunological dysfunction. A study using human neutrophils found that exposure to e-cigarette vapor inhibited their antimicrobial function, decreasing their effectiveness in preventing infection. There is more to be investigated in the link between e-cigarette use and lung infections. As we learn more about vaping induced lung injury, we may discover that the pathological damage
to lung tissue makes a fertile nesting ground for opportunistic pathogens, calling for vape injury patients to have routine testing for AFB disease.

References


**New Initiative!** Calling all interested residents and medical students!

All residents and medical students interested in advocacy are invited to join NMRPA's Advocacy Task Force as we work to develop and execute advocacy projects on issues relevant to Med-Peds providers and our patients.

Please reach out to nicole.danit.damari@gmail.com with any questions or for additional information.
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