



THE Journal

OF THE ARKANSAS MEDICAL SOCIETY

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AUGUST 2019

The Not-So-Free World of Prison Medicine

*A Look at the Challenges and Rewards
of Caring for Patients From the Inside*

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WHAT HAVE WE DONE FOR YOU LATELY?

The Eyes Have It – The Fight is On



DAVID WROTEN
EXECUTIVE VICE PRESIDENT

It makes me proud to see a group of physicians stand up for what's right. I'll be even more proud if you stand with them! Many would argue that eyesight, of all the senses, may be the most precious. No offense if you believe otherwise. One thing we should all agree on is that eye surgery should be performed by the most well-trained and experienced professional. That would be, of course, an ophthalmologist, right?

Not if Act 579 of 2019 goes into effect. Act 579, pushed forward by the state's optometrists, would legislate that optometrists – without attending medical school or a surgical residency – could use lasers, needles, and scalpels. Thankfully, our state's ophthalmologists, through the Arkansas Ophthalmological Society (AOS), are standing up and saying enough is enough.

Hopefully, by the time you read this, a signature-gathering campaign will have gathered enough signatures (approximately 54,000) to put Act 579 on the ballot for the 2020 election. If polling done during the 2019 legislative session is any indication, the voters of Arkansas will, in effect, veto Act 579. A rarely used provision in the Arkansas Constitution allows the public to submit a referendum on enacted legislation if you can gather enough signatures prior to the effective date of the Act (July 23rd). If successful in gathering the signatures, the Act cannot go into effect unless approved by the people of Arkansas.

The last time the referendum process was used was 1994, and the soft drink bottling industry used it in an unsuccessful attempt to overturn legislation creating the "soft drink tax." This tax, to this day, provides critical funding for Arkansas's Medicaid program.

I won't use this space (at least not this month) to give you the details and background on just how this bad legislation (Act 579) became law. Suffice it to say that our Legislature was overwhelmed by optometrists who outnumber ophthalmologists four to one and are involved in every local election.

The General Assembly passed Act 579 in spite of overwhelming public opposition.

That's a very important point. The public strongly believes that only physicians who have been residency-trained in eye surgery should be allowed to operate on our eyes. Only three states – Oklahoma, Kentucky, and Louisiana – allow optometrists to do these procedures, and in each of those cases, the battle was won through the legislative process and not because the public wanted it. And for the record, no one should downplay the important role that optometrists play in vision health. But ... and this is a big but, they are not eye surgeons.

During every legislative session, AMS faces a barrage of scope-of-practice bills. Most of those involve APRNs and/or CRNAs. They all argue, with sketchy evidence or proof, that their bills will increase access to care and that they are well-trained to do what they want to do. But never has the issue been more clear-cut than who can perform surgery! This is an issue for your profession, not just for ophthalmologists.

This will be expensive. The signature-gathering campaign must be completed in less than five weeks. The AOS, its individual members, and its national organization have stepped up strong and raised enough to cover the \$700,000 price tag. Once they gather the signatures and get the referendum on the ballot, "Katy bar the door" because you are now looking at a financial cost with seven digits to the left of the decimal sign. The optometrists aren't going to just sit back. They can be expected to put up a monumental fight.

The implications of winning this battle are monumental and far-reaching. Most importantly, it is the right thing to do. Standing up to protect the safety of medical practice, and in this case surgical procedures, is an obligation of the profession that each of you hold so dear.

When the time comes, I hope you will stand with our state's ophthalmologists and help overturn this unwise legislation. **AMS**

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Issam Makhoul, MD

Is Physician Burnout a Self-Inflicted Wound?

Frozen in his chair and exhausted, staring at his computer and dreading the rest of his evening that will stretch way into the night to finish charting, making phone calls, and signing the stacks of forms that his assistant has left for him, Dr. Clarity feels that nothing is clear in his life anymore. Fifteen years ago, when he signed up for medical school, he thought that he would be making a difference in the lives of every patient he sees. Even though every patient gets out with their prescriptions in hand and their paperwork signed on time, he feels that he barely knows them and they barely know him. He feels fake, like an impostor. Dr. Clarity is suffering from the most dangerous epidemic that the medical profession has seen in its history: burnout.

The number of physicians with at least one symptom of burnout (emotional exhaustion, feelings of cynicism, detachment from work, and a sense of low personal accomplishment) was an alarming 54% in 2014. In 2018, it reached a record high of 78%, based on the 2018 Survey of America's Physicians conducted by Merritt Hawkins for the Physicians Foundation.¹ The majority of physicians are working at full capacity or are overextended (80%), and the majority of them are employed by a hospital or an organization (the number of independent physicians in the U. S. dropped from 48.5% in 2012 to 31% in 2018). Forty-six percent (46%) consider changing their career paths, and 49% would not recommend the profession to their children.

Attempts at diagnosing the root cause of the problem led some to blame the physicians. Hence, solutions were proposed to increase their resilience and improve their well-being – getting

more sleep, changing their diet, doing yoga and mindfulness exercises – and several companies were formed to administer these recipes. What we can see from the burnout trends is that these solutions were no more than a bandage and could not even scratch the surface.

It is fair to say that burnout is a byproduct of the modern transformation of medicine. Indeed, organizational and environmental factors are the main causes for burnout. Due to the introduction of technology requiring huge investments in every aspect of medicine, the fragmentation of medical practice into an enormous number of subspecialties, and the large demand for health care, the profession changed from being a calling or a mission to being a business where profit became a major impetus. This was associated with the rise of a large, bureaucratic army of health administrators that increased by 3,200% between 1975 and 2010 compared to a modest growth of 150% for the population of physicians. The growth of the health administration was a cause and consequence of the increased complexity of the regulations and reimbursement rules. Administrative costs went up along with the cost of medical equipment and drugs. The time consumed to fulfill the bureaucratic tasks imposed on physicians started occupying an increased percentage of their day, leading to long work hours. The modernization of medicine culminated in the universal adoption of electronic health records (EHR), which became a problem by itself. It is estimated that every physician spends up to 23% of his or her time documenting in the EHR.

» *Many accuse physicians of having been passive during this transformation and not standing up enough for their profession to preserve its soul.*

The global effect of this change is a loss of autonomy and control, with slow erosion of respect and self-respect. If this trend continues, the physicians – who used to be special individuals endowed with healing power and shrouded by a halo of respect for their dedication, skills, altruism and sense of service – may become completely alienated parts in a large factory, a part of an assembly line where productivity is the metric by which they are assessed. Many accuse physicians of having been passive during this transformation and not standing up enough for their profession to preserve its soul.

The next horizon is the introduction of artificial intelligence (AI) into medicine. This revolution is coming and is unstoppable, in my opinion. And here again, physicians will be facing a more fundamental change in their identity and role. Looking at the fields that will be overtaken by AI, it is likely that physicians will practice medicine in two or three decades in a completely different way than now. Many of the cognitive processes that physicians used to spend years to acquire will be relegated to AI platforms and will surpass human abilities. The only solution to the current epidemic of physician alienation and burnout is that physicians confront this upcoming revolution head on and thereby direct it to a place where it will have at its center the well-being of patients and physicians.

Physician burnout is not a self-inflicted wound. It is one of the unintended consequences of huge societal and professional changes that happened despite physicians' will or awareness. Its reversal, however, depends on their awareness of its underpinnings and coalescing with their peers and society as a whole to recreate a humanistic medicine for our time.

¹<https://physiciansfoundation.org/wp-content/uploads/2018/09/physicians-survey-results-final-2018.pdf> AMS



The Not-So-Free World of Prison Medicine

*A Look at the Challenges and
Rewards of Caring for
Patients From the Inside*

“ tell new hires or interviewed applicants that it’s like working in a third-world country,” said Nan-

nette Vowell, MD, of her work as medical director of the Ouachita River Correctional Unit’s Special Needs Hospital. “It’s basic medical care, with basic equipment and dedicated staff. If you approach it that way, it’s doable because you sort of know what you’re getting into.” It’s challenging, rewarding, and a specialty she invites more physicians to discover.

Located in Malvern, ORCU is the state’s only prison-based hospital. “It’s a two-man, two-bed-per-cell barracks that we’ve turned into a hospital,” said Dr. Vowell. “Picture a horseshoe grouping of rooms with a nurse’s station in the center. We don’t have x-ray, in-house lab, or ultrasound. We keep a small stock of certain medicine; beyond that, we’re required to go to a pharmacy that ships us overnight the medicine that we need.”

ORCU is also the state’s male intake unit. (McPherson Unit in Newport is the female intake unit.) All inmates moving from an Arkansas jail to another Arkansas (or out-of-state) prison stop first at ORCU. There they are assessed by a dental, mental health, and medical team before being assigned to a parent unit such as Cummins, East Arkansas, or another Arkansas Department of Corrections facility.

A Glimpse Inside

About 1,800 inmates reside at ORCU, and there are 27 hospital beds to go around. Divided into non-hospital care and special-needs care (hospital), the facility’s medical staff includes three physicians, two nurse practitioners, one physician assistant, and a plenteous nursing staff.

One physician works the non-hospital side, which sees the healthier patients – treating chronic health issues like diabetes and hypertension in addition to acute medical illnesses. In contrast, Dr. Vowell and her staff service roughly 900 special-needs patients, who vary in severity from severe psychological illnesses to total care for all activities of daily living – and all points in between.

While prison physicians treat the same maladies as other physicians, their circumstances around providing that care are quite different. There are many misconceptions about working

in the prison system, according to Dr. Vowell, and generally, much to share with her “free-world” colleagues.

She would point out first that physicians that choose to work in corrections are generally trained in family practice or internal medicine. “There aren’t a lot of physicians looking to work in corrections,” said the internal medicine physician who – training aside – has developed quite a passion for her job caring for incarcerated patients. “It’s a different world, but I enjoy it. I love having a wide variety of patients to care for in a single day. It’s a blending of hospital and office work all in one setting.”

(Correctional medicine *is* gaining a foothold as a recognized medical specialty, particularly on the osteopathic side. David Thomas, MD, JD, was instrumental, with Anthony J. Silvagni, DO, PharmD, in the American Osteopathic Medicine’s first approved designation of correctional medicine as a medical specialty. That designation happened in 2012. In 2014, the AOA awarded the first board certification for correctional medicine.*

Much like serving on a medical mission trip, Dr. Vowell treats ORCU patients who are suffering from a variety of disease in all its stages. More self-neglect is seen initially with these patients. “A patient may come in with a large cancer on his neck that has gone untended for a period of years,” she elaborated. “Another may have rheumatoid arthritis so severe that he is completely debilitated.

“The [doctor-patient] relationship is established relatively quickly and can provide a positive influence. It’s rewarding to educate these patients to take better care of themselves. There’s no doubt we’re making a difference. In prison, they have access to health, dental, and mental health care. Some of the patients have never taken the time to consider their own health. These guys use the spiritual, mental health, and medical support to really make changes in their lives.”

Addressing a variety of malaise without the benefit of specialist consult is one unique challenge her job setting creates. On the one hand, it has strengthened her ability to sort out answers alone. “I’m a little more multi-faceted, as

I have had to learn much about post-op orthopedics, pulmonary, and other specialties,” she said. On the other hand, it can feel isolating at times. “There isn’t much access to colleagues here. There are not opportunities for sidewalk consults. I’m fortunate that I have a couple of advisors that I can bounce ideas off. There are prisons out there with a single MD or APN that never sees another colleague in his day’s work.”

Who Controls Whom?

A typical 10-hour workday at ORCU finds Dr. Vowell close to the patients she cares for (more so than if she worked in an outside clinic). Still, she has no control over them. “My patients are wards of the state, with individual rights,” she noted. “That’s important for [outside] physicians to understand. The medical staff cannot give consent for the patient. The patient can refuse treatment, medication, dialysis, surgery, etc., just as any other patient can. Of course, if it is life-threatening, and the patient can’t communicate at all, we do everything [we can] to save a life.”

It’s frustrating, she shared, seeing a patient continually refuse life-preserving medication, particularly when you work in close quarters with that patient. “All providers have non-compliant patients,” she noted, “but with my office in the middle of the room and the patients walking up and down the halls to and from the chow, pill call, and classes, I see these guys more frequently than 30 minutes in an exam room one time a month. So, I feel it hits home just a bit more. We get everyone involved to be sure the patient is making a fully-informed decision – the chaplain, mental health staff, officers, and sometimes his family. But ultimately, these guys make their own medical decisions.

“We are all in the business of good health care for patients,” she said. “I think the biggest thing for me is for the ‘free world’ providers to understand that we on the ‘inside’ need more communication. We implement the plan that the consulting physician recommends. We can’t make the patient compliant, but we can be sure that the tools and motivation are there for the plan to be successful. A good discharge summary, a doc-to-doc call, and good consulting notes with detailed treatment plans are best.”

> Continued on page 32.



Nanette Vowell, MD

The Medical Goal

“The medical goal at ORCU is to keep inmates’ footprint in the community to a minimum,” said Dr. Vowell. “It would be startling for most people to see a shackled man flanked by officers sitting in the waiting room. Instead, we’re trying to establish more on-site clinics, so we can get physicians to come to the unit or participate in telehealth. We want to keep the patient here and bring here everything that he needs.”

Surgeon Robert Breving, MD, visits ORCU’s special-needs clinic every other weekend to help with upper endoscopy and colonoscopy patients. While he doesn’t do operative procedures on site, he does carve out time when needed for on-site general surgery consultations. He will recommend the appropriate course of action. After that, it is up to Wellpath to determine where they will send the patient for definitive care. (ADC contracts with Murfreesboro-based Wellpath to provide health care services to prisoners in the state of Arkansas.)



Robert Breving, MD

➤ ***“It’s a different world, but I enjoy it. I love having a wide variety of patients to care for in a single day. It’s a blending of hospital and office work all in one setting.” – Nannette Vowell, MD***

“If we can bring services to them, it does a couple of things,” said Dr. Breving. “It lessens chances of prisoners escaping, but also it cuts costs significantly. A study done in another state** estimated that a round-trip visit to take an inmate to a hospital or clinic and back – transportation and medical guard costs alone – could be as much as \$2,000. So, if I can see 10-15 patients a day there, that’s a significant cost savings to the state.”

There are some real challenges that physicians should be aware of when coming on-site. Malpractice coverage – and the threat of frivolous lawsuits – can be an issue. “I started going out there seven years ago and had to purchase a different malpractice policy,” said Dr. Breving.

“The policy I had would not cover the prison. I could only find two companies nationally that would write a policy covering that. Malpractice alone for me to go out there is around \$1,300 per day.”

Depending on the specialty they’re providing, some outside specialists may only need to come once per month or even less often. “We have a nephrologist that comes once a month and manages our dialysis patients, but we could benefit from having pulmonary, urology, endocrinology, and neurology,” said Dr. Vowell.

Still, at times it’s simply necessary to take patients to an outside clinic or hospital for treatment. This is no small chore. So many people must know of the pending appointment, from the warden to the corporal who will drive the van. “Sometimes the consulting office clears the office for the patients to be seen – it is quicker in and out for everybody,” she explained. “These trips out of the unit require security to be on high alert to protect the community and the inmate. Offices and hospitals that are willing to cooperate with the extra complications and drama of providing care to the correctional patient are really appreciated. We have limited medical runs daily, and they are planned several weeks in advance. We try to have all the information needed to have a complete visit every time we send a patient out. We try to use our resources wisely.”

Even though not on-site, physicians who are willing to see patients in their clinics despite the real scheduling and patient issues are doing a good service. Speaking to those who might be considering such, Dr. Vowell attempted to lessen potential fears. “Sometimes, physicians are understandably hesitant about having shackled patients visit their clinic for fear that [those patients] may frighten or even harm their staff or established patient population,” she said. “Most of the time, these guys are respectful and kind. They want their health issues treated. I often get comments from consulting physicians like, ‘He was really nice!’ or ‘He hasn’t been a problem here at all!’”

As for getting started receiving patient visits, there isn’t as much red tape as one might expect. “They simply agree to do it, and they

understand that there will be two officers and a prisoner in shackles in their facility,” said Dr. Vowell. “Payment is through Arkansas Blue Cross Blue Shield of Arkansas, which is essentially a Medicaid plan.”

Heeding the Call

While specializing in correctional medicine is probably not a path most doctors have given much thought, many who have tried it believe it’s worth considering. In a January 2016 article from the American Osteopathic Association,*** veteran correctional medicine physician John G. Mills, DO, MPH, shared the plights and rewards associated with the job. In speaking to the qualities a physician needs to succeed in correctional medicine, he said, “You need to be positive and have an upbeat personality. The issues you encounter, such as addiction and homelessness, can be hard on your psyche. I like to tell myself, ‘We did the best we could today. We’re going to come back tomorrow to do more.’”

“I never imagined I would be in correctional medicine,” summed Dr. Vowell. “I have been here seven years. I learn something new every day. I’m grateful to Wellpath for the opportunity to continue to work in this system. I don’t think it’s for every physician, but I do think that every physician has something to offer this unique medical practice.”

Physicians who would like to learn more about correctional medicine in Arkansas or how to get involved, please contact Dr. Vowell at nvowell@wellpath.us or Dr. Breving at bigbertmd@md.

*<https://thedo.osteopathic.org/2013/02/when-patients-are-behind-bars-new-specialty-challenges-physicians/>

**The State Health Care Spending Project, an initiative of The Pew Charitable Trusts and the John D. and Catherine, reports, “Expenses add up quickly when inmates must travel long distances to see specialists or stay overnight in hospitals. The Legislative Analyst’s Office in California, for example, reported that medically related guarding and transportation costs for one inmate can exceed \$2,000 per day.” <http://www.pewtrusts.org/~media/assets/2014/07/stateprisonhealth-carespendingreport.pdf>

***<https://thedo.osteopathic.org/2016/01/working-with-inmates-a-correctional-medicine-physician-shares-insights/> AMS



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Behavioral Modifications (BM) – Headache Management

Jhablall Balmakund, MD

Professor of Neurology, Department of Pediatrics
University of Arkansas for Medical Sciences, Lowell

Abstract

Management of migraine and tension-type headaches (TTH) can be chal-

lenging. Medications may or may not be helpful, may cause side effects, or may worsen headaches. Behavioral Modification (BM), with or without medication, will expand treatment options. Care providers are frequently reminded about the risks and benefits of medications. Patients and caregivers are becoming increasingly wary of medication side effects and would like to avoid them. It is recommended that care providers encourage patients to decrease their dependence on medication and increase BM therapy. Caregivers are reassured that medication is an option if BM is ineffective.

Introduction

BM may play a role in headache management. It has been empirically validated for migraine and tension-type headache (TTH). Meta-analyses yielded a 37-50% reduction in tension-type headache, compared to 33% using amitriptyline.¹ Penzen DB et al concluded that modifications in TTH management would make standard behavioral treatments available and conducive to primary care settings where most patients receive treatment.¹

In November 2015, the Food and Drug Administration issued strong caution to manufacturers of over-the-counter acetaminophen about potential liver injury when the drug is used in larger quantities.² The FDA has also warned that non-aspirin, Nonsteroidal Anti-Inflammatory Drugs (NSAIDs) increase the chance of a heart attack or stroke.³ A discussion of non-medicinal therapies can be challenging when medication is available that may work almost immediately. This challenge may be less daunting with the use of motivational decision making:

- 1) Use open communication and coordination between patients and care-providers.
- 2) Encourage patients/caregivers to come up

with their own approach after agreeing on the goals of management.

- 3) Discuss these therapies as part of the management.
- 4) Set realistic goals.
- 5) Take into account the patient's comorbidities and medications.
- 6) These modalities of care may require a trial of weeks or months.
- 7) Use medication after these therapies have not met the expected treatment goals or when appropriate.
- 8) A tincture of time for some symptoms, including headache, may suffice.
- 9) Many of these modalities of care are beneficial to health and general wellbeing. It may be helpful if insurance companies suggest an initial trial of non-medicinal therapy in the appropriate headache patient before medication use.

Patients may prefer non-medicinal interventions for a variety of reasons including poor tolerance or response to medication, contraindications to medication, pregnancy, planning to become pregnant, and/or breastfeeding. Stress or deficient coping skills may also make a patient better suited for non-medicinal therapies.⁴ BM may be used in conjunction with medicine, devices, medical personnel, etc.

Behavioral Modification Therapies

The U.S. National Sleep Foundation recommendations for sleep practices and tips for children include the following: maintain a regular circadian rhythm (**a sleep routine**) appropriate for age in a comforting environment; enforce bedtime; avoid naps, activities, devices and anything that may delay or disrupt sleep;⁵ understand that too much or too little sleep are considered triggers for migraine, so the bedroom should be primarily for sleeping (TV, video games, cell phones, etc. should be removed to avoid temptations).

The human body is about 60-65% water. A consistently pale yellow to colorless urine may indicate **appropriate fluid** intake. To avoid disruption of sleep, most of the fluid should be drunk earlier in the day. It has been speculated that water deprivation may play a role in migraine, particularly in prolonging attacks.⁶

A **well-balanced diet** at the appropriate time and the avoidance of “high sugar foods” may help prevent “hunger headaches.”⁷ Minimize processed, fatty foods, fasting, or skipping meals. There are a variety of chemicals and food additives that may trigger a headache. Higher levels of lipids and free fatty acids increases platelet aggregability associated with decreased serotonin and heightened prostaglandin levels. This combination may provoke vasodilatation, the precursor of migraine headaches.⁸

Both the U.S. Department of Health and Human Services and the American Academy of Pediatrics recommend a combination of aerobic and anaerobic **activity** for 60 minutes daily.⁹⁻¹⁰ This should be age-appropriate and tailored to the individual's ability to strengthen muscle and bone.¹⁰ Randomized controlled and non-controlled trials have demonstrated that exercise may be associated with reduction of migraine intensity and migraine-related disability. Darabaneanu et al found that the mean number of headache days per month, duration, and intensity decreased in the group that exercised compared to control after 10 weeks of exercise.

Triggers provoke the cascade of events that culminate in a headache. They may be inconsistent, require a “cofactor,” dose dependent, or have a delayed effect making avoidance or identification a challenge. These triggers may have an additive effect, which is especially seen in female headache patients during their menses.⁴

Participation in activities that are **enjoyable and relaxing**, while not being harmful or destroying property, may help decrease the focus on headaches.

Body Mass Index (BMI) between 85th and 95th percentile is overweight and above the 95th percentile is **obesity** for children and teens of the same age and sex, per Centers for Disease Control. Obesity is felt to increase the risk of migraine by 40-80%, with the higher risk for the more obese patients, especially in those of reproductive age. Dr. B. L. Peterlin, director of headache research at Johns Hopkins University School of Medicine in Baltimore, feels that care providers should **promote healthy lifestyles and exercise** for patients with migraine. Obesity is associated with increased headache frequency and disability regardless of headache type. Conservative therapies for migraine seem to be more effective in children younger than six years than in older children.

Complete resolution of headache is a bonus. Help patients manage their symptoms without too much dependence on medication. If the headache remains the same with decreased use of medicine, that is an improvement. Some patients may not consider the long-term effects and opt for the much simpler, quick relief of medication. While others may not respond well or have comorbidities that preclude the use of medications. More than 12 million Americans visit their doctor complaining of headaches yearly. The cost to society is greater than \$31 billion dollars. A Beth Israel Deaconess Medical Center study suggests some of that cost could be offset by physicians ordering fewer tests and an increased focus on counseling about lifestyle changes. Long term, and within year one, Minimal Contact Behavioral Treatment was found to be the least costly approach to migraine prevention. In the CHAMP Study, there were no significant differences in reduction of headache frequency or headache-related disability in childhood and adolescent migraine with Amitriptyline, Topiramate, or placebo over a period of 24 weeks. The active drugs were associated with higher rates of adverse events. Medication Overuse Headache (MOH) may decrease quality of life, cause daily and incapacitating headaches, insomnia, and non-restorative sleep, as well as psychological distress and reduced functioning. MOH is associated with biochemical, structural, and functional brain changes.

Behavioral Modification may help with maintenance of good health, wellbeing, and management of life's stresses including headaches.

Long term goals of BM include reduction of frequency and severity of headaches, headache-related disability, reliance on poorly tolerated or unwanted medications, headache-related distress and psychological symptoms, and the feeling of not being in control of their treatment.⁴

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Following Children with Developmental Disorders in Primary Care

ANGELA SCOTT, MD, PHD, FAAP; MAYA LOPEZ, MD, FAAP; JILL FUSSELL, MD, FAAP; and JAIMIE FLOR, MD, FAAP

GD, a 37-month-old boy with Autism Spectrum Disorder returns for his three-year well-child check. Last year you referred him to the Arkansas Autism Partnership (AAP) Waiver program for evidence-based behavioral interventions, and to First Connections for speech-language and occupational therapy at his Head Start. His parents say “everything is going well” but can’t give details about progress or goals. Annual occupational and speech therapy assessments indicate slow progress. Your clinical observations indicate very limited social engagement.

The importance of early access to developmental therapies in children with developmental delays is well established.¹

How does a primary care physician (PCP) assess the quality of a child’s developmental services? Progress may be difficult to appreciate during a PCP visit. Updated therapy goals or annual testing results may not be readily available.

While detailed assessment of progress and outcome may not be possible in a brief visit, it is incumbent upon the child’s PCP to follow up with families to ensure

that developmental services are optimizing outcomes. These key questions help assess quality of services during follow-up visits.

What progress has he made? Parents of children with developmental delays watch their child’s progress closely. It is important to give them a chance to celebrate hard-won improvements. Start with general questions and then ask detailed ones, such as *What has improved since I last saw you? What has not? Where had you hoped to see more progress?* Parents may have unrealistic expectations for progress and/or may not recognize clinically specific gains. This provides an opportunity for counseling and education.

What therapies is he getting? Is he receiving services and therapies appropriate for his developmental needs? Therapies often take place within complex systems that are difficult to navigate. Provider availability, transportation and competing priorities can get in the way. The National Autism Center maintains a reliable list of clinically appropriate, evidence-based interventions, ranked according to strength of evidence. The *National Standards*

Project is available online at www.nationalautismcenter.org.

What are his therapy goals? Do his current goals match the family’s most pressing concerns? Are the goals clinically appropriate to his diagnosis? A child with autism should have at least some goals that target foundational communication skills. In GD’s case, his progress in all other areas will be slow until his social engagement skills improve. His treatment plan should include evidence-based strategies, such as applied behavioral analysis (ABA) to target the core social-communication deficits associated with autism. The AAP’s MCD Waiver program provides these and other autism-specific interventions. Additional information at <https://uofapartners.uark.edu/projects/autism-partnership/>.

Who is on his team? Effective developmental services must happen within a coherent team of adults caring for the child. The team should include parents and family caregivers, therapists, teachers, clinicians and everyone who interacts with the child. Framing the relationship between caregivers and professionals in this way highlights the impor-

tance of family engagement and effective communication.

How is each team member helping meet his goals? If team members communicate well and work closely together, parents and teachers understand what therapists are working on and can provide opportunities to practice those skills throughout the day. This therapeutic “carryover” not only multiplies exposure to intervention, but also ensures the child is practicing skills in different contexts. Skill “generalization” can be especially difficult for children with autism. It is important that all team members realize this to avoid mistrust among caregivers who may doubt each other’s versions of the child’s abilities.

How does the team communicate? Do parents communicate directly with the therapists and therapists with teachers? How often? Does the family feel like an important part of the therapeutic team? Do they feel welcome observing therapy sessions or asking questions? Effective team communication is a key marker of service quality. It is necessary for therapeutic carryover and integration of skills across settings. Asking about and providing suggestions to improve team communication is one of the most important things a PCP can do in a follow-up visit. Some teams communicate well through a long-standing email conversation. A low-tech option is a communication notebook that travels with the child across activities and providers. However, written communication cannot take the place of face-to-face meetings where parents, teachers and therapists develop trusting relationships.

Where does he spend the day and what’s that like for him?

Services, both at home and in a group setting, should align with the Individuals with Disabilities Education Act’s (IDEA) guidance to provide a “free and appropriate education in the least restrictive environment possible.”² To a physician, appropriate services should provide necessary supports for the child to successfully engage in their environment. Services must include targeted, evidence-based interventions to improve the child’s delays.³ Intensity of intervention frequently requires a higher adult-to-child ratio and higher provider expertise than is found in a typical daycare or classroom. The challenge is always to balance effective intervention and adequate support with the benefits of a naturalistic, inclusive setting. Therapy services integrated into daily home and classroom routines enhance therapeutic carryover, build effective team relationships, decrease behavioral problems associated with transitioning in and out of therapy settings, and effectively utilize naturalistic, child-led intervention strategies.

After discussing these issues with GD’s parents, you arrive at a shared plan for them to set up a team meeting with his therapists and teachers. You help them list topics for that meeting, including a communication notebook and specific things they can do at home to improve his social engagement. You encourage them to apply for the AAP Waiver program. They have not done this because they’ve heard the waiting list was too long. You provide contact information and encourage them to speak directly with program personnel. You reinforce that his progress in other

areas is limited by his poor social-communication skills. You emphasize that the interventions provided by the AAP, such as ABA, have been shown to improve the core social engagement delays associated with autism. Acknowledge that it is ultimately the family’s decision but strongly recommend this program. The family agrees to this plan and a follow-up visit with you in three months.

Parents understandably worry about doing everything they can to optimize their child’s future. It’s important for PCPs to regularly check with families about their concerns, and be a knowledgeable and trustworthy ally in the family’s efforts. Reviewing the child’s service plan and reassuring the family that it seems appropriate, or offer suggestions for how to intensify it, benefits the child and the whole family. ▲

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Iron Deficiency Anemia for the Pediatric Primary Care Provider

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Abstract

Iron deficiency anemia (IDA) is prevalent in the pediatric population in the U.S. Early identification and initiation of treatment is crucial in preventing adverse outcomes associated with IDA. Hence, it is critical that providers are able to identify at-risk patients, obtain and interpret the appropriate laboratory studies and prescribe oral iron therapy in the primary care setting. Referral to a pediatric hematologist is indicated if the etiology of anemia is unclear or if the patient remains resistant to oral iron despite adequate dosing and administration.

Introduction

Iron deficiency is the most common nutritional deficiency worldwide. Fortunately, the addition of iron-fortified infant formulas and foods has decreased the prevalence of iron deficiency in the U.S.¹ The 2007 to 2010 National Health and Nutrition Examination Survey estimates that in the U.S., 2.7% of children ages 1 to 2 years and 2.4% of adolescent females ages 12 to 21 years have iron-deficiency anemia.^{1,2}

Iron is an essential ingredient for effective red cell production, myoglobin formation, collagen and neurotransmitter production, immune function, and numerous other biochemical pathways.³ Seventy percent of the body's iron stores are used to manufacture hemoglobin.⁴ A microcytic, hypochromic anemia develops from insufficient iron delivery during heme synthesis.⁵ Anemia impairs oxygenation of tissues and can manifest as poor energy, pallor, headache, dizzi-

ness, and shortness of breath. In severe anemia, children may have tachycardia; a systolic flow murmur; or conjunctival, palmar, or oral mucosal pallor. However, physical signs of anemia typically do not present until hemoglobin drops significantly.

Several observational studies suggest that iron deficiency, even without anemia, is associated with neurocognitive and behavioral delays in younger children.¹ Iron-deficient adolescents may also experience negative effects on cognition, audiovisual reaction time, and physical endurance.² Therefore, it is critical that primary care providers identify at-risk children, perform appropriate diagnostics, and initiate treatment to prevent poor health outcomes associated with severe and chronic IDA. Uncomplicated IDA can be managed effectively by the primary care provider.

Risk factors

In children, IDA presents in a bimodal distribution occurring in the late infant/toddler population and then later in adolescents.¹ These two stages of rapid growth have increased physiologic iron requirements and are frequently periods of poor nutrition.³

In infants/toddlers, iron deficiency most commonly develops following the transition to

cow's milk. Infants who drink cow's milk before the age of 1, and those who consume more than 24 ounces per day, are at increased risk. Cow's milk is a poor source of iron, and high volumes of milk often replace other iron-rich foods in the diet.⁴ Cow's milk may also inflame and damage the intestinal mucosa, leading to microvascular bleeds and chronic blood loss.³

There are multiple other risk factors for iron deficiency: a history of prematurity (80% of iron stores are transferred in the third trimester of gestation), low birth weight, lead exposure (causes iron malabsorption), exclusive breastfeeding beyond 4 to 6 months of age, low socioeconomic status, children of Mexican-American descent, and prolonged bottle feeding.^{1,3} The American Academy of Pediatrics (AAP) recommends a complete blood count (CBC) for all children at 1 year of age. Additional screening should be repeated as needed throughout childhood if risk factors or symptoms are present.¹ The prevalence of IDA peaks again during adolescence. Additional risk factors in this age group include obesity, alternative diets (i.e. vegetarian), eating disorders, new onset chronic illnesses, strenuous athletic training, and menarche. Anovulatory cycles leading to dysfunctional uterine bleeding are common within the first two years of menarche.⁴ IDA is more common in females due to this chronic blood loss. Although the AAP does not recommend routine screening of all adolescents, the Centers for Disease Control recommends that all women of childbearing age be screened every five to 10 years.²

Diagnosis of IDA

A CBC provides valuable information on the etiology of anemia. Anemia is defined as a he-

» **Along with initial CBC and reticulocyte count, the newborn screen should be reviewed since the presence of hemoglobin Barts in a neonate suggests alpha thalassemia.**

moglobin that is two standard deviations below the mean of the population of the same age and gender.⁶ Other cell lines are typically in normal ranges in IDA, although thrombocytosis can be present.⁷ Evaluation of red blood cell (RBC) indices reveals critical information that can suggest iron deficiency as a cause of anemia. The RBC count is decreased due to insufficient substrate for red cell production. Mean corpuscular volume (MCV) is decreased as cells become smaller due to decreased iron delivery for heme synthesis.⁵ The Mentzer index can be calculated to help differentiate iron deficiency from the second-most-common microcytic anemia, thalassemia. The index is equal to the MCV (in fL) divided by the RBC count (in millions of red cells per microliter). An index greater than 14 is consistent with IDA. A value less than 13 is consistent with thalassemia.⁶ This calculation highlights the difference between the elevated RBC count characteristic of thalassemia and the depressed RBC count seen in IDA.

Another finding on CBC consistent with IDA is an elevated red cell distribution width (RDW), which signifies a wide variation of cell size (anisocytosis). Findings on peripheral smear (Figure 1) parallel those of the CBC, with small, pale RBCs that demonstrate increased central pallor. There is typically significant size variation among red cells, and unusual forms – such as pencil cells – may be seen.

In addition to a CBC, a reticulocyte count should be obtained to determine whether the anemia is a result of inadequate production or destruction of red cells. Critically, the reticulocyte count will be inappropriately low in IDA despite the degree of anemia since red cell production is defective.⁷ If the reticulocyte count is significantly elevated, red cell destruction or acute blood loss should be considered.

Along with initial CBC and reticulocyte count, the newborn screen should be reviewed since the presence of hemoglobin Barts in a neonate suggests alpha thalassemia. Beta thalassemia will typically present around 3 to 6 months of age once fetal hemoglobin decreases.⁶ Family history and ethnicity should be reviewed to evaluate for other hemoglobinopathies.

In patients with clinical histories and lab values that are consistent with IDA, a thera-

peutic trial of oral iron can be initiated without further studies. If the diagnosis remains unclear, the total iron-binding capacity (TIBC) and serum ferritin should be obtained. The TIBC is typically

elevated as a response to maximize iron absorption. Ferritin is a measure of total-body iron stores, making it more reliable than a random

> Continued on page 40.

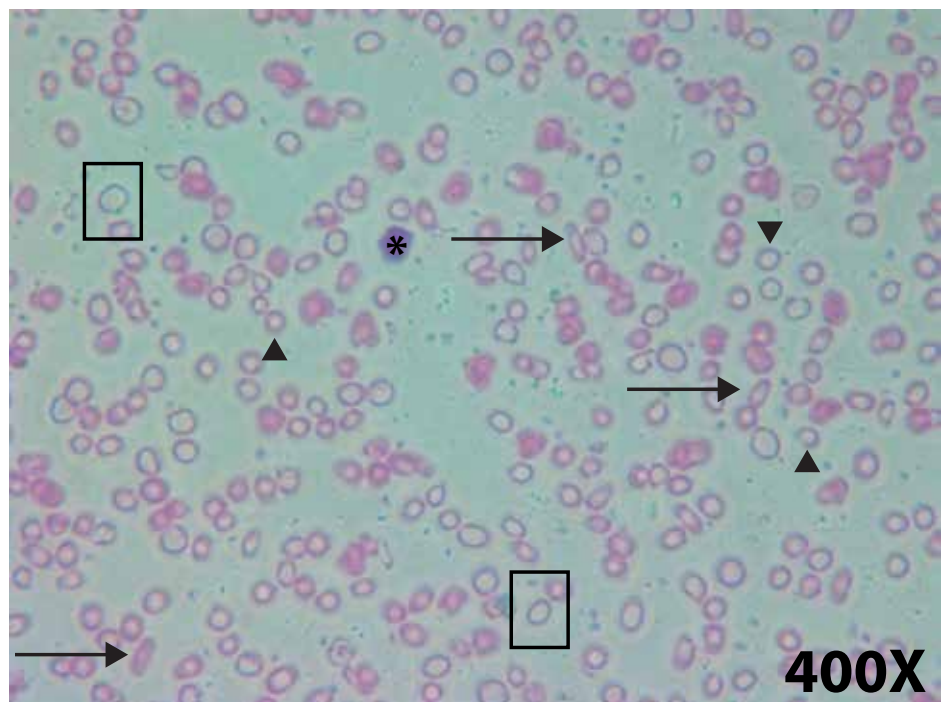
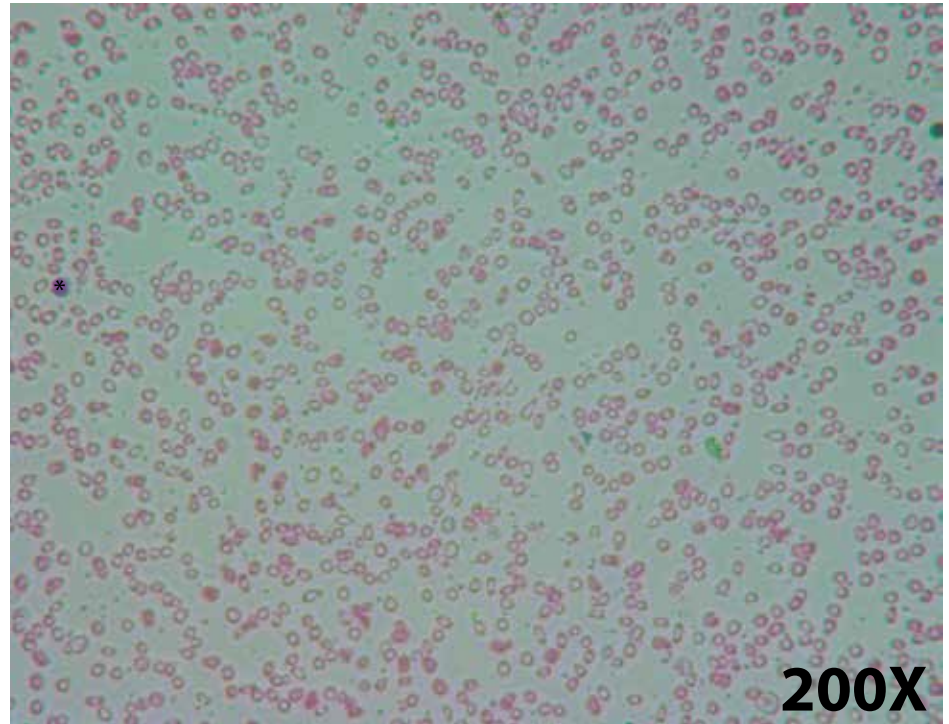


Figure 1: A peripheral smear of moderate to severe IDA shows microcytosis, with the average RBC smaller (arrowhead) than a lymphocyte (*), hypochromia (central pallor > 1/3 total cell diameter, examples boxed) and anisocytosis (varied cell size, which correlates with elevated RDW). Thin, elongated cells known as pencil cells are also commonly seen (arrows).⁵ The smear illustrates findings from a teenager with menometrorrhagia. The child was symptomatic, presenting with a hemoglobin concentration of 3.8 grams/dL, hematocrit 15.1%, MCV 60.4 fL and RDW 20%. Serum ferritin was 1.2 ng/mL and TIBC was 405 µg/dL.

Table 1: Common laboratory findings in iron deficiency anemia, thalassemia and anemia of chronic disease.^{4,6}

Lab Values	IDA	Thalassemia	Anemia of Chronic Disease
Hgb	Anemia is defined as two standard deviations below the mean of the population of same age and gender.		
RBC count	Decreased	Increased	Decreased
MCV	Decreased	Decreased	Decreased
Mentzer Index (MCV/RBC)	>=14	<= 13	--
RDW	Increased	Normal/increased	Normal
Ferritin	Decreased	Normal	Increased
TIBC	Increased	Normal	Normal/Decreased

(non-fasting) serum iron. Hypoferritinemia is diagnostic for IDA if the value is less than 12 ng/mL in children ages 1 to 5 years or less than 15 ng/mL in children older than 5 years of age. However, since ferritin is an acute-phase reactant, it may be elevated and thus not reflective of tissue iron stores during periods of inflammation or illness.⁸ Measurement of serum iron is not a reliable indicator of iron status as there is natural diurnal variation, and the value may be elevated after iron intake despite depletion of body iron stores.⁵

If iron studies are unremarkable, other causes of microcytic anemia in children need to be considered including thalassemia, anemia of chronic disease, and lead poisoning. Table 1 compares the laboratory findings amongst these etiologies.

Treatment

Once IDA is diagnosed, oral iron therapy should be initiated. A packed red blood cell transfusion is only recommended if anemia is severe and the child demonstrates clinical signs of distress: tachycardia, chest pain, shortness of breath, dizziness, poor feeding, or poor growth. If required, pRBCs must be transfused slowly to prevent overload and heart failure. In addition to transfusion, oral iron supplementation is required since the iron in the donor red cells is not available for red cell production.³

The standard dosing recommendation for oral iron in IDA varies widely from 3 to 6 mg/kg/day of *elemental* iron divided in one to three daily doses.⁷ There are multiple oral iron preparations available, and the amount of elemental iron

varies by formulation. For example, in ferrous sulfate, elemental iron comprises 20% of the total dose, so 15 to 30 mg/kg/day of ferrous sulfate is recommended. A recent, randomized clinical study suggests that ferrous sulfate results in a greater increase in both hemoglobin and iron indices when compared to another formulation, iron polysaccharide complex.⁹

In another study comparing a single daily dose versus three divided daily doses, both regimens showed a comparable increase in hemoglobin and ferritin.¹⁰ Several trials have suggest-

ed that lower, once-daily doses may be as effective as high doses since the absorptive capacity of the duodenum appears to be saturable.^{5,7} For children who poorly tolerate oral iron, once-daily dosing given every other day can also provide good effect.⁷ For moderate-to-severe IDA, we typically give ferrous sulfate at 20-30 mg/kg/day divided twice daily and capping at one 325 mg tablet two or three times daily in larger children and adolescents. Whichever iron formulation is chosen, it is critical that the prescriber be aware of the proportion of elemental iron present in the chosen iron preparation and dose appropriately. In our experience, many children referred for hematology consultation of poor response to iron therapy have been under-dosed.

Poor compliance with treatment is common due to dosing frequency, grey discoloration of teeth, constipation, nausea, and poor taste.⁴ Families should be counseled to wipe the teeth after administration to prevent discoloration and start a stool softener, if needed, for constipation. The iron supplement should be given one to two hours before or after meals with water or juice. It is important to avoid milk products with iron administration since this can decrease

Table 2: Etiologies of poor response to oral iron treatment.⁸

Common Etiologies of Treatment Failure with Oral Iron:	Recommendations:
Inadequate Dosing	Prescribe 3-6 mg/kg/day of elemental iron (e.g. 15-30 mg/kg/day of ferrous sulfate).
Inadequate Duration of Treatment	Treat for a minimum of 12 weeks or until ferritin and CBC normalize.
Improper Administration	Administer on an empty stomach with water or juice. Do not give with milk. Avoid antacids, H2 blockers, and PPIs if possible.
Poor Compliance	Prescribe once-daily dosing. Prescribe slow-release formulations. Give at bedtime to help minimize stomach upset. Counsel on and treat constipation.
Poor Diet	Educate on proper nutrition. Encourage less than 20 ounces of cow's milk daily. Increase iron-rich foods.
Ongoing Blood Loss or Malabsorption	Evaluate for chronic bleeding or malabsorption (e.g. menorrhagia or metrorrhagia in females, inflammatory bowel disease, Celiac disease, high stomach pH)
Incorrect Diagnosis	Other microcytic anemias to consider include thalassemia, anemia of chronic disease, sideroblastic anemia, lead poisoning.

» Families should be counseled to wipe the teeth after administration to prevent discoloration and start a stool softener, if needed, for constipation.

absorption. Iron given on an empty stomach at bedtime may enhance absorption since GI motility is decreased during sleep.⁷ Absorption may also be enhanced in the presence of ascorbic acid, hence some clinicians recommend co-administration with vitamin C supplements or fortified juice.⁷ More generally, oral iron requires an acidic environment for optimal absorption. Children who require daily antacids, H₂ blockers, or proton pump inhibitors may be resistant to oral iron due to impaired absorption.⁷

A CBC and reticulocyte count should be repeated four weeks after the initiation of treatment. It is critical to evaluate a patient's compliance before interpreting results. If there is good compliance and IDA is the correct diagnosis, the reticulocyte count should be elevated and hemoglobin should have increased by at least 1 g/dL.¹ If there is a positive response to treatment, oral iron should be continued for at least three months to replenish tissue iron stores and minimize the possibility of recurrence of IDA. Repeat labs should be drawn prior to discontinuing treatment to ensure normalization of the hemoglobin concentration, MCV, and serum ferritin level. If hemoglobin does not respond as expected, several etiologies should be considered, as described in Table 2.

For patients who have impaired absorption, intolerance, or remain resistant to oral supplementation, there are several newer and safer formulations of intravenous iron available. A referral to hematology should be made if IV iron therapy is indicated, the etiology of anemia is unclear, or if the anemia remains refractory despite adequate dosing and administration.

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Malignant Hyperthermia: When You Least Expect It



Benjamin K. Woodle, MD; Daniel Bingham, MD;
Tolga Suvar, MD; Jill Ramsey, MD; Alvin Stewart, MD

Background

Malignant hyperthermia (MH) is a rare genetic condition that predisposes 1:15,000 pediatric patients and 1:40,000 adult patients to a hyper-metabolic state resulting in abnormal muscle contraction predominant after exposure to an offending agent such as volatile anesthetics or succinylcholine.

Other manifestations of MH include rising EtCO₂, acidosis, hyperkalemia, rhabdomyolysis, myoglobinuria (“Coca-Cola colored urine”), and hyperpyrexia. Onset of symptoms vary, from immediate to several hours after exposure.

Early efforts attempting to determine the mechanisms of MH focused on the gene for the ryanodine (Ryr1) receptor. This particular receptor is an ion channel responsible for calcium release from the sarcoplasmic reticulum, and it plays an important role in muscle depolarization. Subsequent genetic studies in humans have revealed at least five different chromosomes and more than 180 individual mutations associated with MH. Most patients with an episode of MH have an indicative family history that includes a relative who has an “anesthetic allergy” or possible death resulting from a reaction to anesthesia. Thus, a pre-op evaluation consisting of a family history

» **There are not many side effects to treating MH with dantrolene; whereas, the end result of not treating MH promptly is death.**

of anesthetic complications is always important prior to surgery. With high clinical suspicion, a proper diagnosis can be made with a muscle biopsy and a halothane-caffeine contracture test.

In many of the early reported cases, both succinylcholine and a halogenated anesthetic agent were used. Of note, and interestingly, nearly 50% of patients who experience an episode of MH have had at least one previous uneventful exposure to anesthesia, during which they received a recognized triggering agent. MH is most commonly triggered by inhaled, halogenated volatile anesthetics such as sevoflurane, isoflurane, desflurane, and halothane; but it can also be triggered by the depolarizing muscle relaxant succinylcholine. A halogenated anesthetic agent alone may trigger an episode of MH; however, the prevalence of MH with exposure to succinylcholine only, has been controversial. There has been doubt that succinylcholine, alone, causes MH crisis in susceptible individuals.^{1,2} However, data from the MH registry and new data from the Canadian MH experience show that MH crisis can be induced by succinylcholine administration alone.³⁻⁵

Case Report

A 20-year-old Caucasian male, with an unknown past medical history, was a restrained driver involved in a MVC. Upon EMS arrival, his GCS was 5, interosseous access was obtained, and the patient was intubated with succinylcholine and ketamine for airway protection at the scene. A level-one trauma was activated, and the patient was transported to the trauma center via med flight. On arrival to the trauma bay, vital signs were GCS 6T, HR 163, BP 70/32, spO₂ 99% and EtCO₂ 45, with no obvious injuries on pri-

mary or secondary survey. Vital signs were Temp 40.8C, HR 130, BP 144/34, spO₂ 100, etCO₂ 53. iSTAT showed a pH of 7.30, PCO₂ 28, and HCO₃ of 14.3. The patient was given 650mg Tylenol via NG tube and started on Propofol and fentanyl for sedation. A CT scan was then obtained and showed no intracranial or intraabdominal injury and was only significant for a transverse fracture of L1 vertebrae. Patient was taken back to the trauma bay, where a Foley catheter was inserted revealing dark urine. The patient’s temperature continued to rise to 41.3C. The first set of labs returned and were significant for potassium of 6.0, Cr 2.2, Lactate 6.0, CK 1688, and troponin 2.99. ABG 7.06/58/92/16.8 on (SIMV 90% 600/20/11). A cooling blanket was placed over the intubated patient, with ice packs placed around his head and extremities; cold intravenous fluids were started. The patient continued to have hypercarbia, with EtCO₂ peak at 100. The decision was made to initiate dantrolene therapy for suspected malignant hyperthermia. The patient received a total of four doses of dantrolene, dosed at 2.5mg/kg, with improvement in both temperature and etCO₂. The patient was transferred to the ICU, where dantrolene administration was continued at 1mg/kg/hr. The patient later developed multi-system organ failure due to rhabdomyolysis and severe acidosis. The family elected to make the patient DNR, and he was pronounced shortly thereafter.

Discussion

Attributing to the rarity of malignant hyperthermia, most health care providers do not have a high clinical suspicion to make this diagnosis. However, in the presence of an offending agent such as succinylcholine and/or halogenated-vol-

atile anesthetics, it is important to always keep in mind the diagnosis and a promptness to treat. This case is exemplary of a situation where succinylcholine is administered to facilitate endotracheal intubation without exposing the patient to anesthetic gases. Recognition of this clinical syndrome cannot be undermined.

In addition, prior exposure to an offending agent without signs or symptoms of malignant hyperthermia does not rule out the diagnosis. Furthermore, when a patient presents with the above signs and symptoms, it is important to recognize them and treat early. There are not many side effects to treating MH with dantrolene; whereas, the end result of not treating MH promptly is death. Early recognition and treatment of MH is critical to patient survival and is a necessary skill for first responders, emergency personnel, and trauma staff. While more studies are needed to elucidate the actual magnitude risk of MH associated with succinylcholine administration alone, it is becoming clear that organizations where succinylcholine may

be used with or without triggering inhalational agents should have both the MH antidote (dantrolene) and a clearly outlined approach for the management of an MH crisis readily available in case an event occurs.

Once the diagnosis of MH is made, the timely response to treatment is potentially life-saving. Emergency treatment of MH consists of discontinuation of volatile agents and succinylcholine. The crash cart and dantrolene should be at bedside. Hyperventilation with 100% oxygen at flows of greater than 10L/min should be aimed at achieving lower levels of EtCO₂. Simultaneously, dantrolene should be given at a bolus dose of 2.5 mg/kg through a large-bore intravenous access. Cumulative doses of greater than 10 mL/kg may be given until the symptoms of contracture or rigidity resolve. Blood gases may be measured to guide therapy and monitor the degree of metabolic acidosis and hyperkalemia. The patient's core temperature should be cooled if rapidly rising >39°C. The Malignant Hyperthermia Association of the United States (MHAUS) should be contacted

for incidence report and further guidance of this life-threatening disorder.

Background

1. Iazzo PA, Wedel DJ. Response to succinylcholine in porcine malignant hyperthermia. *Anesth Analg* 1994;79:143-51.
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Blueberry Muffin Rash in a Neonate

David N. Matlock, MD;^{1,2} Franscesca Miquel-Verges, MD^{1,2}

¹UAMS, Little Rock

²Arkansas Children's Hospital, Little Rock

A 36-week newborn, followed prenatally for an enlarged liver, microcephaly, and Dandy-Walker malformation, was delivered by caesarean section due to non-reassuring fetal heart tones to a 25-year-old G1 mother without significant past medical history. The mother had appropriate prenatal care, and the pregnancy had been complicated only by the abnormal ultrasound findings and an afebrile exanthema, which occurred during the first trimester. The infant was noted to have jaundice, a purpuric rash with petechiae (Panels A and B), and hepatosplenomegaly.

Cranial sonography revealed punctate foci in the periventricular white matter. Echocardiogram revealed a moderate pericardial effusion. She had hepatitis

with a direct bilirubin of 19, disseminated intravascular coagulopathy with severe thrombocytopenia (platelet count was 21,000), and respiratory failure. She had elevated serum IgG and IgM titers as well as a positive urine polymerase chain reaction for cytomegalovirus.

Magnetic resonance imaging of the brain revealed microcephaly with diffuse cerebral volume loss, periventricular calcifications, and neuronal migration abnormalities. Hearing screens were failed bilaterally. Thrombocytopenia responded to multiple platelet transfusions. Neutropenia developed during antiviral treatment and required multiple doses of granulocyte-monocyte colony stimulating factor.

The infant completed a six-week course of intravenous ganciclovir before transitioning to oral valganciclovir. She was discharged from the neonatal intensive care unit on room air, taking full oral feedings at seven weeks of life. She is enrolling in developmental therapies and will require close follow-up of her growth and development.

While usually asymptomatic, 10% of newborns with congenital cytomegalovirus infection will have manifestations at birth. Among symptomatic patients, multi-organ involvement is common and a small subset present with life-threatening disease. The preferred diagnostic tests are viral culture or polymerase chain reaction in urine. Treatment with ganciclovir (IV) is



recommended. Once stable, treatment can be provided orally (valganciclovir). Mortality remains high at 3-10% for newborns with symptomatic disease. Sequelae are common, specifically sensorineural hearing loss and neurodevelopmental disability.¹

1. American Academy of Pediatrics. Cytomegalovirus Infection. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. Red Book: 2018 Report of the Committee on Infectious Diseases. American Academy of Pediatrics; 2018; 310-317. **AMS**

>> Magnetic resonance imaging of the brain revealed microcephaly with diffuse cerebral volume loss, periventricular calcifications, and neuronal migration abnormalities.

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