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Arkansas Blue Cross and Blue Shield Begins VBCI Implementation

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by BRENT DAVIS

ON THE COVER

Arkansas Blue Cross and Blue Shield Begins VBCI Implementation

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COMMENTARY

Sandy Johnson, MD

Quality Versus Quantity

My dad has and continues to have a very big influence in my life. He has many sayings that replay themselves in my mind. Some of these include "When the blind lead the blind, they both fall in the ditch," "Knowing is doing," "The only way to do great work is to do what you love," "Your life is God's gift to you and what you do with your life is your gift to God," "Judge and you will be judged" and "Make sure the door does not kick you in the $$ on your way out." He likes to work hard and play hard. He also has overcome a lot in life including being injured in battle in Vietnam. He wants me to make sure to live every moment to the fullest because you never know if this moment may be your last moment. I know he repeatedly says these things to remind me to make good choices, give my best, treasure every moment, and to love everyone in my life.

While my dad takes life by the horns and lives every minute to the fullest, my mom believes more in moderation. She believes in "quality over quantity." I have been blessed to have two wonderful parents who teach me various great values, love each other, and love me.

I have been thinking about their sayings recently when it comes to the practice of medicine. Our clinic, Johnson Dermatology, has a saying also: "We strive to provide the most effective, efficient, empathic, and empowering medical care." We have opted to participate in all of the quality improvement programs that we can. We participate in MOC, MIPS, MACRA, PQRS, Choosing Wisely, MORE, etc. We have our own internal reviews and quality assurance programs. We have recently learned that BCBS is now rolling out its Value Based Compensation Initiative as a way to "reward value over volume." At first glance, this sounds skintastic. I believe quality always wins. Quality definitely wins in a free market society. However, as some people say, the devil is in the details. I am interested to know how many physicians were involved with creating this program. One of my concerns is who is excluded from the program. If I understand it correctly, non-physician providers, NPPs (nurse practioners and physician assistants), are not included. At Johnson Dermatology, I work very closely with two NPPs. They went through a rigorous training program with my husband and myself after their schooling. We work under the same roof. We follow the guidelines of working with NPPs closely. I think NPPs are an important part of the medical team. I am concerned why they would be excluded from this program. I think our NPPs provide high quality care.

That being said, I think we all know of NPPs who choose to work independently. There is even legislation and movements to allow NPPs to work without the collaboration or supervision of a physician. I have – and I am sure many of you have – seen patients who have not received high value or high quality care from some of these independent NPPs. My concern is that if BCBS and other organizations are going to be judging physicians on quality, shouldn't they also be judging NPPs? If NPPs are wanting to be treated the same as physicians, shouldn't they be held to the same standards as physicians? I think we all would agree that the training of a NPP is much different than medical school, residency, and board certification. I think we all would agree that we all want to provide the latest and greatest care; that is why we participate in MOC, read journals, do CME, attend conferences, chat with our peers, participate in medical societies, etc. I think we all would agree we strive to provide high quality care. I am excited to participate in these programs. Like my mom, I believe in quality over quantity. Like my dad, I believe that the only way to do good work is to do what you love. I also believe; however, that when the blind lead the blind, they both fall in the ditch.

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by BRENT DAVIS

Arkansas Blue Cross and Blue Shield Begins VBCI Implementation

The winds of change blow constant in the health care industry and a new breeze is forming. Arkansas Blue Cross Blue Shield has begun the process of switching payments to medical providers from the longstanding Fee-for-Service (FFS) structure to a new systems called the Value-Based Compensation Initiative, or VBCI. While some see the change from FFS as inevitable, others seek answers and details of the new program.

Steven Spaulding, executive vice president and chief health management officer at Arkansas Blue Cross and Blue Shield, explains how VBCI will work.

"A portion of the payment to providers will be directed to a value pool." says Spaulding. "Funds in that pool will be distributed each quarter to providers based on their RowdMap score. Providers who score well on that system will get a greater portion of that distribution than those who score poorly. This redistribution is intended to compensate providers at a higher level for providing high value care as informed by RowdMap using Dartmouth Atlas and Choosing Wisely criteria."

RowdMap is a company that has studied the work of Choosing Wisely, an organization comprised of over 70 physician specialty societies, and the Dartmouth Atlas of Health Care that has done work for many years with participation by physicians to identify variation in cost and quality of care. RowdMap has built algorithms using the work of these organizations as a provider-informed source of information and is able to score individual providers based on the overall value of services on a comparative basis in defined geographic areas.

So how will Arkansas Blue Cross and Blue Shield determine "low-value" versus "high-value" treatment? The major insurance provider in the state will use its claims data to create "value" scores for providers that measure low- versus high-value care based on evidence-based medicine from well-respected resources within the medical community, including national provider associations. These value scores will be used to distribute value payments to providers.

In 2019, Arkansas Blue Cross and Blue Shield will begin stepping down fee-for-service compensation over a four-to-five-year period until base fee-for-service compensation approximates Medicare. The funds from this step down of fee-for-service monies will be placed into three separate value pools — hospital, specialist and primary care. These value pools make up the value-based compensation component.

On a quarterly basis, 100% of the value-pool funds will be distributed to providers based upon a set of value-based performance metrics using the most recent and available 12-month performance period, which will roll forward every quarter. These performance metrics, as stated earlier, will be based primarily upon a claims-based scoring methodology developed by RowdMap, with sources from Choosing Wisely (www.ChoosingWisely.org), the Dartmouth Atlas (www.DartmouthAtlas.org), and other evidence-based clinical guidelines and research studies.

Steven Spaulding

During 2018, Arkansas Blue Cross and Blue Shield has committed to continuing communications with stakeholders, releasing information on the specific metrics and measures by provider specialty type by which providers will be compared to their peers (cohorts), releasing shadow reporting that lets providers know how they're scoring based upon historical claims, and providing information to help them improve their scores.

In the Winter edition of "Blue & You," the Arkansas Blue Cross and Blue Shield publication, the reason for the change to VBCI is explained. "For decades, doctors and hospitals received payment based solely on volume — how many tests that they ran, how much bloodwork they collected, for example. We realize this fee-for-service model will no longer work in the future if we want to maintain affordability and sustainability for our members. And we do."

Additionally, the publication notes, "Low-value care raises health care costs for everyone. The average American family spends more than $4,000 annually on direct and indirect costs driven by low-value care. An example of low-value care is having surgery for a condition that can improve through physical therapy. The surgery might fix the problem, but physical therapy is less risky for you (going under anesthesia can have complications) and less expensive."

Spaulding adds, "I want to be clear in saying that in no way are we judging the competency of providers in this process. We are not saying these providers are good or bad from a clinical perspective. The scores will reflect the difference in the way providers provide services to similarly situated patients which most times lead to very similar outcomes, but can vary significantly from a resource use perspective to get to those similar outcomes. There is general agreement in the industry that there is currently as much as 30% of health care spend that supports low value care. This program is focused on rewarding not the provision of the service itself, but the value of the service to the patient."

Lonnie Robinson, MD, FAAFP, is a physician at the Regional Family Medicine in Mountain Home, Ark., who has seen his fair share of change in the health care field.

"I completely understand and, to a large degree, embrace the fact that FFS is going away. Change, particularly change in our current health care environment, is inevitable." says Robinson. "I am interested in seeking out new payment models that will redesign the way we are paid for care delivery, but I feel strongly that they should also be developed in a way that they do not present unintended consequences for providers, specifically those practicing in primary care. That being said, I have several concerns about the VBCI."

His concerns are highlighted below:

Perpetual Losers Based on Relative – Rather Than Absolute Performance. "Based on the understanding I have about the program, it seems to

indicate that there will always be losers. If we are measured relative to our peers and split into quartiles, there will always be relatively low performers, thus there will always be penalties, no matter the level at which the entire pack is performing. If the goal is to increase overall quality, why not decide on a reasonable standard to which the program might aspire to raise the quality bar, and reward all providers who perform above the mark? As planned, if there were a theoretical situation in which all providers were providing a high level of care, there would still be providers who would be penalized despite their collectively high level of performance. I would suggest that setting a mark of acceptable performance (above which there would be no penalty) would be a way to raise quality while allowing for the (albeit unlikely) scenario in which all providers could avoid penalty if meeting an acceptable level of performance. This approach has already been utilized successfully with the pattern seen in APII, both for the Episodes of Care and PCMH programs, in which BCBS already participates. It is my understanding that the only way to continue to receive the traditional/historic rate of reimbursement from BCBS is to be a top performer. This is quite concerning, since the costs involved in clinical practice for all providers have not decreased and are, in fact, rising."

Lonnie Robinson, MD, FAAFP

No Relief of Administrative Burden (Prior Authorization). "As you present VBCI as a way for BCBS to gain control of rising costs, the administrative burden on practices continues to rise as well. We have already had discussions about removal of prior authorization and other utilization-management strategies employed by payers that present an excessive administrative burden for practices as we move increasingly away from FFS, and I have heard the argument by BCBS that studies demonstrate a rise in utilization with removal of the PA process. I would argue that none of those studies took place in an environment in which providers were ultimately held accountable for both quality (adherence to Choosing Wisely and other guidelines) and cost.

"The AAFP agrees with me … in their Principles for Administrative Simplification, they state: 'Physicians strive to deliver high-quality medical care in an efficient manner. The frequent phone calls, faxes, and forms physicians and their staff must manage to obtain prior authorizations (PAs) … impede this goal.' and 'PA for imaging services should be eliminated for physicians with aligned financial incentives (e.g. shared savings, etc.) and proven successful stewardship.' "

"If you are going to hold providers accountable on the tail-end (threat of reduced reimbursement if not performing well re: cost and quality) then the reasonable trade-off should be removal of the onerous process of prior authorization on the front-end. Failure to do so is the health care analogy of double-jeopardy: requiring us to deal with administrative headaches and paying someone in our offices to jump through hoops on the front end, then being penalized on the tail end with lower reimbursement."

Threat to Primary Care. "Value-based programs should bolster primary care, not present a threat to them. The threat of narrowing the already thin margins under which many of our rural primary care physicians operate does not incentivize improvement as much as it presents a threat to practice viability. I believe that the constantly rising cost issues being faced by BCBS ARE NOT significantly impacted by the habits of primary care providers … we are not the 'big spenders' in this situation. And yet my colleagues feel this initiative is targeting the physicians who bring more value to the table than any other specialty. As you probably know, despite all its short-comings, FFS income continues to be the 'bread and butter' of primary care practices, since we are charged with providing E/M-dependent care to our populations, rather than procedures and surgeries.

"As Kent Moore, senior strategy analyst for the AAFP recently said, 'While payment is moving toward value-based care, fee-for-service (FFS) remains the dominant method of payment. Family physicians provide high quality, cost-effective care but are financially dependent on the thin margins associated with current FFS payments to pay for the increased administrative and clinical personnel needed to transition to and be successful in value based contracts. To reduce payment in any amount for the services done by primary care physicians is detrimental to their ability to provide high-quality, low-cost care in the current FFS care environment.' An initiative that seeks to decrease reimbursement in such a manner (i.e., there will always be losers) is a threat to primary care practice viability. Unfortunately, those who are most vulnerable are going to be those in small practices in rural parts of our state, and this represents a significant portion of the Arkansas primary care workforce."

Alignment. "We are bombarded with new programs and new ways in which we are measured. We have literally dozens of disparate quality metrics by which we are evaluated. What reassurance do we have that there will be near-perfect alignment with existing programs (CPC+, PCMH, MIPS, etc.)? All of the programs have some measures in common, but there are always some/many that are different. This too, creates additional administrative burden the practice must bear [while] at the same time figuring out a way to staff adequately despite declining reimbursement.

On a quarterly basis, 100 percent of the value pool funds will be distributed to providers based upon a set of value-based performance metrics using the most recent and available 12-month performance period, which will roll forward every quarter.

"According to the AAFP Principles for Administrative Simplification, 'Quality measures have proliferated in the past 15 years, leading to a significant compliance burden for physicians. Most of the measures are disease-specific process measures, rather than more meaningful evidence-based outcomes measures. With many family physicians submitting claims to more than 10 payers, the adoption of a single set of quality measures across all public and private payers is critical,' and similarly, 'All payers … should implement the core measure sets developed by the multi-stakeholder Core Quality Measures Collaborative to ensure parsimony, alignment, harmonization, and the avoidance of competing quality measures.' "

Data. "Every program in which we participate has a secure website at which we may obtain access to our performance data. Every one of those sites requires a secure password and often asks us to change that quarterly. The CMS CPC+ portal takes me at least five minutes to login and get to the information I need. The AHIN site makes me change my password often, I typically don't have ready access when I need it. Same story with the Arkansas PDMP. I know that there will be a similar process for this program, one that presents data in the way YOU want me to see it, not necessarily in the way I need to have it. I don't really need another login. If we are being asked (forced?) to participate in a program such as VBCI, real data access and transparency is a must. Additionally, we need the opportunity and a reasonable period of time in which to review and validate the data by which we are being measured before the financial implications are applied. I have

> Continued on page 248

not been made aware of [such] a process in the context of VBCI."

Jason Wilson, CPA, FACMPE, is the chief executive officer of Medical Associates of Northwest Arkansas (MANA), an independent physician group with more than 80 physicians. His concerns focus on the impact VBCI may have on groups such as MANA.

When asked what impact he foresees VBCI will have on various patient populations — specifically if it might have any negative effect on the chronically ill patient or those with severe disabilities/conditions — Wilson responds, "I think this is hard to determine and one that I really hope would not be a concern. There is a concern that it could incent physicians to focus their practice on the healthier patients that are seen less and therefore do not negatively impact the metrics as much. I worry that doctors with older patient populations may score worse than doctors with younger populations. I've also heard from groups like GI that feel that the metrics could be a deterrent in finding issues during screening exams. For instance, the metrics may incent doctors performing colonoscopies to not have the additional charges that come with finding polyps during a colonoscopy or may reward doctors who have low detection rates vs doctors with higher detection rates. Typically, the Adenoma Detection Rate (ADR) is a measure of quality in GI practices for colonoscopy, but this model does not measure that. Instead it measures the number of times the CPT code is used indicating that polyps were detected. I would like to believe that which patients a doctor sees is not influenced by the economics of a model, but it is something to think about. By the way, I have also had this same concern with some of the PCMH measures from Medicaid, ABCBS and others that incent lower total cost of care."

Jason Wilson, CPA, FACMPE

Wilson also has concerns regarding the various metrics/measures upon which physicians will be scored. "Some of the metric trees that ABCBS has released contain as many as 35 to 45 different metrics that are being measured in this program," he says. "These individual metrics are then rolled up into subscores and rebalanced, rolled up into another subscore and then rebalanced before ultimately being rolled up into the physician's overall score. Each physician's overall score is then ranked and the physician given a score of 1 to 5. Even if a physician can determine that one metric is out of line, will the data be detailed enough for the physician to determine how to modify their practice style to influence that metric (i.e. if the metric for procedures per patient are out of line, will the physician be able to determine which procedures are out of line?)"

Wilson adds a concern about the depth of control a physician may have over scoring, "Several of the metrics are based upon referrals or hospital measures. For instance, a *referral* is defined as any specialist visit occurring within 30 days of a primary care visit. A patient who self refers to a dermatologist within 30 days of an unrelated primary care visit will count as a *referral* on the primary care physicians score card. Regarding the hospital numbers, some of the scores for 'Optimal Hospital' are based on 'Average Readmission Rate to your Referral Hospitals' or 'Average Length of Stay of your Referral Hospitals.' These measures may be controllable if a physician has multiple options of hospitals to refer to, but in many places in Arkansas, the physician does not have this luxury. As an independent medical group, it is important that we receive accurate payment for the services that we provide. Currently, we are able to track the number of visits or services we provide and then ensure that we are paid the correct fee schedule amount for each service. Under the VBCI model, it will be much more difficult, if not impossible, to accurately monitor that payment. In this new model, each payment will be reimbursed at a reduced amount and then we may receive a payment for the difference in the future. Depending on how this quarterly payment is reported, we may or may not be able to audit that back to the *withhold* amounts that occurred each month. It will be even more difficult to reconcile if the quarterly payment is 50 percent, 150 percent or 200 percent of the withheld amounts.

"To complicate matters more, ABCBS controls the data that is used to determine how a physician is scored, whereas, in the current model, physicians have all of the information to ensure that they are paid correctly. The measures used are not typically tracked by physicians and even if they were, physicians do not have the comparable data needed to determine whether or not they are being scored correctly when compared to their peers. How transparent will the data be?"

Will the proposed method fairly measure a physician's performance? Wilson isn't so sure: "The VBCI system will be graded on a bell curve. To my knowledge, ABCBS has not released the projected percentages in each of the one-to-five categories; therefore, it can only be assumed, based on a standard bell curve, that the distributions will something close to the following: 1) 0-10%; 2) 10-15%; 3) 65-75%; 4) 10-15%; and, 5) 0-10%. Regardless of how poorly the best physician scores or how well the worst physician scores, using a bell-shaped curve methodology, there will always be physicians graded in each of the five categories, and there will always be winners and losers. Even if all physicians were to outperform some national benchmark, there would still be physicians who lose in this system.

"In addition, this is a moving target. The score a physician receives not only depends on how that physician performs but also on how the physician's peer group performs. A physician who is rated a 5 in one quarter and makes improvements may still be rated a 5 in the next quarter if the entire group of physicians shows improvements. Presumably, all physicians will be trying to improve their numbers and it may be difficult to move from one ranking to the next. To further complicate matters, if physicians who score a 4 or a 5 leave Arkansas or are forced out of business, all remaining physicians will naturally move down the scale with some 3s becoming 4s or 5s OR some 3 scores becoming 4 scores or 5 scores."

Regarding the timing of payments and cash flow, Wilson shared additional concerns, "Most independent physician practices in Arkansas practice cash-based accounting. In a cash-based system, revenue is recorded when the payment is received. In the VBCI system, the practice will receive a reduced amount each month and then a settlement at the end of the quarter that could return anywhere from 0-200% of the withholds from the clinic. Most practices look at financials monthly. With the fluctuations that will occur in monthly cash flow, will these monthly financial statements accurately show how the practice is performing or will that only be determinable on a quarterly basis?

"In several years, when the payment reductions from ABCBS reach 25%, physicians in Arkansas will be receiving monthly payments roughly equal to Arkansas Medicare reimbursement. Arkansas currently has one of the lowest Medicare reimbursement rates in the country. As the payment withholds from ABCBS increase, will practices be able to pay all of their monthly bills on the reduced rates? Do some of the quarterly payments need to be held and used for overhead payments in future quarters? What if there are no quarterly payments?

"Many practices pay physicians monthly or quarterly based on the practices' cash flow. The change to the VBCI payment methodology will cause large fluctuations in monthly cash flow. Practice changes could include the timing of bonuses, holding some of the funds in the months the quarterly bonus pool is received to help in the months that reimbursement is cut, etc."

Wilson was asked how the practice will make decisions and prepare for the VBCI. He responded, "ABCBS is planning to start the program in 2019; however, since the program is using a rolling 12 months of data in order to determine the physician scores, the work that physicians are doing right now will influence their score in the first quarter of 2019. ABCBS has not yet fully rolled out the program to Arkansas physicians and hospitals; therefore, physicians are going to be measured on data that was generated before knowing the impact of their decisions or before physicians/hospitals have been able to make changes in processes. Since the program is not yet fully developed, physicians also have not yet received any preliminary data to identify areas of potential improvement. ABCBS has stated they hope to have preliminary 'shadow' reports out by Summer 2018. By that point, physicians will be six months into the measurement period for the first quarter of 2019.

"ABCBS has talked about adding a 'Trendline' to the data starting in 2020; however, this has not yet been defined to my knowledge. ABCBS has stated that there will be some sort of 'Reasonable Trend' added to the overall calculation in the future. ABCBS has stated that if physician utilization decreases over time, the addition of this *trendline* could result in more funds being available than were actually withheld. ABCBS has also stated that if overutilization were to occur, that could reduce the funds withheld therefore reducing the payout to all physicians. How is this *trendline* determined? What happens if the "overutilization" is a valid increase in utilization much like we saw this year when flu activity pushed volumes much above normal trends? Will valid fluctuations in utilization be taken into account in the trend or will the physicians be taking all of the risk for valid increases in utilization? Based on the trendline implications and questions above, is ABCBS still taking normal insurance risk or is that being passed along to the physicians/hospitals? Will the ABCBS fee schedules change in future years? How will fee schedule changes or other significant changes (such as the recent pharmacy reimbursement issues) be factored into the trendline?"

What are the implications on Ancillary services? Wilson has a unique perspective on this point. "Many physicians offer in-house lab and x-ray services," he begins. "Larger clinics may also offer other services such as MRI or CT. Technical reimbursement in most of these areas has been reduced to the point of these services having marginal profitability already. With the further cuts that will come from the VBCI, many of these services may become unprofitable when actual collections are compared to actual expenses. Will it be possible to determine how much of the quarterly payments are allocable to the ancillary services to determine the actual profitability of these services? What if none or only 50% of the bonus pool is earned? Will the ancillary service be profitable?

"Practices have to allocate professional and technical payments using different methods. Professional payments may be allocated directly to the physician generating the service, while technical payments have different rules that must be followed to properly allocate those payments without violating Stark and Anti-Kickback regulations. If the quarterly bonus pool that is paid to a physician includes both professional and technical payments, what steps must a practice take in order to remain compliant with the law? Will the practice be able to determine how much of the payment is professional versus technical? What if the bonus pool is 150% or 200% of the withhold? Does some of the excess need to be allocated as technical to remain compliant?"

One point upon which both providers and ABCBS agree is [that there] is the potential to reduce administration costs through the elimination of prior authorization programs.

Spaulding is open to the prospect. "ABCBS would rather not maintain prior authorization programs, they are administratively costly and physicians do not like them. They are effective in reducing the number of services that do not meet appropriateness criteria. Our goal is to be able to eliminate these utilization management programs when we see that other incentives have caused the prevalence of those services to be well managed, not by the prior authorization process, but in the decisions of those who are providing services. It is quite likely that we will find a way to eliminate that requirement for certain providers who have shown a history of the provision of high value care which would include appropriate use of those services which now require prior authorization. We are working to find a way to do that."

AMS Executive Vice President David Wroten has received multiple calls from across the state with questions about the new program. He feels the Society and physicians should have greater input into the structure and implementation of VBCI.

"We have set up a committee (headed by Dr. Amy Cahill) with the intention of meeting with Arkansas Blue Cross and Blue Shield to express our concerns and to offer suggestions that we are hearing. We are most concerned with the Bell Curve and how physicians, hospitals, and groups on the back end of the curve will be, in essence, penalized. We are doing our best to advocate for inclusion in the decision-making process."

From any perspective, it seems clear that the winds of change are gathering strength. AMS

CASE STUDY

Aspergillosis Appendicitis in an Immunocompromised Patient: A Case Report and Review of the Literature

Sara McKeever DO; Emily Barrett BA; Jason Mizell, MD, FACS, FACSRS

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Abstract

Appendicitis is a common condition caused by inflammation of the vermiform appendix. Fungi are a rare cause of appendicitis, but critical to recognize and treat. Disseminated fungal infections can be lethal, particularly in immunocompromised patients. Early diagnosis, surgery, and appropriate antifungal regimen are imperative for survival. We review a case of a 55-year-old man with multiple myeloma (MM), who presented with fever and right lower quadrant (RLQ) pain; the diagnosis of appendicitis was made. Microbiology revealed fungal hyphae and Aspergillosis index and immunoassay were positive, establishing the diagnosis of aspergillus appendicitis. He was treated with laparoscopic appendectomy and antifungal medications.

Introduction

One major cause of mortality in immunocompromised patients is disseminated fungal infections. With various presentations and difficulty in obtaining a definitive diagnosis, fungal infections must remain on the differential in susceptible patients with a fever of unknown origin. We present both a case report and literature review of fungal appendicitis.

Case Report

A 55-year-old male presented in January of 2013 with a two-month history of severe weakness and fatigue. He was found to have elevated serum protein, and with further work-up a diagnosis of MM was made. On October 5, 2013, he received his first stem cell transplant. Nine days later, he developed fever and RLQ pain and was admitted to the hematology/oncology service. On physical exam, he was tender to palpation of the RLQ without guarding or rebound. CT scan revealed appendicitis. Relevant lab values were as follows: WBC 0.04, hemoglobin 8.7, hematocrit 24.2, platelets 28, and CRP 190. He was taken to the operating room, where suppurative appendicitis was confirmed, and an appendectomy was performed without any perioperative complications. Pathology report was positive for numerous fungal hyphae within the wall of the appendix and vessels. His Aspergillosis index was positive. After an Aspergillosis immunoassay returned positive, he was started on antifungal treatment. CTs of the chest and sinuses were negative for fungal infection, and blood and fungal cultures were negative. He completed three months of Voriconazole, Amphotericin, and Micafungin. He is currently being re-staged and has plans to restart chemotherapy.

Literature Review

Appendicitis is one of the most common causes of an acute abdomen, and it's currently approximated that males and females have a lifetime incidence of 8.6% and 6.7% respectively.[1] The classic pathogenesis of appendicitis begins with luminal obstruction, which leads to increasing intraluminal pressure, occlusion, and thrombosis of the small vessels. The resulting inflammation engorges the appendix creating a susceptible environment for microbial invasion by gut flora. Common causes for the inciting obstruction include fecaliths, calculi, lymphoid hyperplasia, recent gastrointestinal infections, and tumors. The common inhabitants of the GI tract that are frequently associated with appendicitis include *Escherichia coli*, Peptostreptococcos, *Bacteroides fragilis*, and Pseudomonas.

Fungal organisms are far less commonly found in appendicitis. There have been increasing reports of fungal appendicitis presenting with acute abdomen, and these have largely been attributed to invasive fungal infections. This increase is suspected to be due to the increasing use of antineoplastic and immunosuppressive agents, antibiotics, and prosthetic devices.[2] The prevailing theory for the development of fungal appendicitis is the hematogenous spread of fungus in immunocompromised patients This etiology is better described as a manifestation of invasive infection, rather than a predilection of fungus for the appendix. The most common pathogens isolated in ill patients with disseminated fungal infections are *Candida, Cryptococcus, Aspergillus, Fusarium*, and *Scedosporium.* Presentation involves a variety of non-specific symptoms and signs of sepsis, most commonly involving the pulmonary or gastrointestinal tract.[2] There is little data regarding fungal appendicitis, making an accurate estimation of incidence difficult. In a review of autopsy cases with systemic mycosis in Thailand, fungal forms in the appendix were found in 0.65% of the patients.[3]

Fungal Appendicitis: Presentation and Incidence

The presentation of fungal appendicitis mirrors bacterial appendicitis: RLQ pain, anorexia, nausea, vomiting, and fever. Labs and imaging are typically unable to determine the infectious agent, and fungal cultures are classically negative early in disease.[2] The initial step in management of fungal appendicitis is identical to bacterial appendicitis: appendectomy. Diagnosis of fungal etiology is either through positive fungal cultures, increasing indexes in antigen assay, or histopathological demonstration of fungal elements. If these means confirm or increase suspicion for diagnosis, the patient is treated for a disseminated fungal infection. There is no absolute treatment recommendation for invasive fungal infections, so it is dependent upon both the fungal form suspected and the treating institution.

In a review of appendices removed from June 2010 to June 2011, three out of 262 resections were identified as fungal appendicitis.[4] All three patients were immunocompromised and had a presentation consistent with acute appendicitis requiring appendectomy.[4] Another patient with AML developed appendicitis secondary to aspergillus thrombosis of the appendicular artery.

Despite prolonged therapy of Amphotericin B, the patient's small bowel perforated and he soon passed.[5] Although rare, the immunocompetent patient is not without risk.

Fungal Infections in the Immunocompromised

Classically, *Candida* has caused the majority of infections. With the increasing use of antifungal prophylaxis, like Fluconazole, *Aspergillus* is increasingly becoming the dominant agent. *Candida* was seen in 8-25% of patients with hematologic malignancies when patients were not on antifungal prophylaxis.[6] *Aspergillus* has been observed in 2 to 28% of hematologic malignancy patients.[7] In Invasive Aspergillus, the respiratory tract is the most common site infected due to inhaled spores. Other organs involved in 10-25% of cases are the liver, kidney, brain, GI, and skin.[8] Focal extrapulmonary IA is very uncommon.[9]

Invasive Aspergillosis

Aspergillus spores are aerosolized and inhaled. In an immunocompromised patient, inhaled conidia colonize the respiratory system and penetrate the bloodstream causing disseminated infection. In pulmonary aspergillosis, the classic triad of symptoms are fever, pleuritic chest pain, and hemoptysis in the neutropenic patient. The most common presentation is simply a prolonged fever of four days in the setting of neutropenia. The ingestion of *Aspergillus* spores could be an alternate route of infection.[10] Typically, *Aspergillus* cannot infect the mucosa of the GI tract, but several chemotherapeutic agents increase the risk of mucositis, which provides a favorable environment for spore invasion.

The gold standard for diagnosis of hematogenous *Aspergillus* infections is a fungal blood culture. *A. fumigatus* infections, the most common, are often culture negative, especially early in the disease.[8] Direct microscopy sensitivity is quite variable ranging from 0 to 90%.[8] Histopathology of infected tissues will show only fungal forms and are not diagnostic of the specific fungal species. Nonculture methods such as antigen assays are required to diagnose IA.[8]

Treatment is based on three components: antifungal therapy, immunosuppression, and surgery. Initially, a Voriconazole regimen is recommended (Grade 1A). Voriconazole has a greater likelihood of response, low mortality, and less adverse reactions.[9] There is some debate on the addition of echinocandins, so the choice is institution dependent. Alternatively, Amphotericin B can be used, as Voriconazole is often limited by hepatotoxicity. Duration of therapy is dependent on location of infection, underlying disease process, and amount of immunosuppression required during therapy. There is no recommended regimen for extrapulmonary IA due to the rarity of presentation.[9] Voriconazole is the recommended primary treatment, but therapy is often modified for a salvage approach. Typically, antifungal therapy will continue for many months.

The degree of immunosuppression should be decreased during treatment of the fungal infection. Fewer patients with severe suppression had a response to therapy when compared to less suppression.[9]

Surgical debridement is dependent on location of the infection, amount of tissue involved, and the patient's ability to tolerate surgery. In cases where necrotic tissue limits antifungal efficacy or local vasculature is threatened, surgery is indicated. In this case, surgical therapy was necessary to prevent hemorrhage and perforation. Outcomes for IA are variable in the literature, with mortality ranging from 35% to over 94%.[8]

Conclusion

Focal extrapulmonary IA is uncommon and only appears in the setting of case reports. It must be managed with antifungals, surgery, and lessening of immunosuppression. We presented an immunosuppressed patient with clinical findings consistent with appendicitis. Following appendectomy, a diagnosis of IA was made, and he was treated with antifungal therapy. The patient recovered well and has continued MM treatment.

Physicians should be aware of the possibility of fungal appendicitis in the immunosuppressed, and of appropriate therapy due to the high potential for poor outcomes.

» *Physicians should be aware of the possibility of fungal appendicitis in the immunosuppressed, and of appropriate therapy due to the high potential for poor outcomes.*

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Next Steps After a Positive Developmental Screen

BY MAYA LOPEZ, MD; JILL FUSSELL, MD; ELDON SCHULZ, MD; and CHAD RODGERS, MD

Case: *GD, a 22-month-old healthy boy is in your clinic for his 18-month visit. Mother reports, "He's got to have his Thomas movie going over and over. He's very stubborn, won't talk, won't mind. But his granddaddy was like that too." Parents say he does not point, "he tends to fall out upset when he wants something" and ignores other children, "unless they have something he wants."*

Health surveillance, conducted by primary care providers (PCPs) during patient encounters, has a low yield when screening for developmental delays. To identify children at high risk for developmental delay, the American Academy of Pediatrics recommends using standardized developmental screening tools at 9, 18, and 24 or 30 months; and a standardized screen for Autism Spectrum Disorders at 18 and 24 months.[1]

While many of us would correctly conclude that GD needs a developmental evaluation, we may struggle with what and how to tell the family or knowing what else GD needs. Aside from referring appropriate children for developmental evaluation and services, the PCP is tasked to:

1) present screening results to the family using a culturally sensitive, family-centered approach
2) determine the cause of delays, including hearing and vision screens
3) maintain links with community resources and coordinate care with them.[2]

EFFECTIVE COMMUNICATION

When you must deliver bad news, using effective communication skills helps families share personal information, become active participants in treatment, and cope better with medical issues and grief. Families need to provide information about symptoms, and feel that we are listening and are empathetic.[3] Parents' satisfaction about receiving testing results or news about their child's medical condition is affected by the directness with which the news is given, if they feel the clinician understands their concerns, the clinician's communication skills and if they received a lot of information.[4]

Parent focus groups for typically developing children preferred a non-alarmist style of sharing the news (e.g., "I think most children should be doing this by now.") Parents of children receiving early intervention (EI) services preferred an honest, straightforward approach.[5] Parents also recommended that clinicians emphasize the need for action by families and care providers, and parents be given an opportunity to prepare themselves to accept the news. Families expressed frustration on the timeliness of their care provider's response to their concerns. Clinicians who make direct and specific statements regarding their concerns, despite being uncertain, may be more helpful to the family than false assurances.

GD's mother shares her concerns but also subtly suggests these may be unfounded as "GD is just like his granddaddy" or these symptoms are part of GD's personality. It is important to assess parents' level of developmental awareness. If they are aware, be direct but gentle and avoid sugar-coating your concerns. If parents are not ready to proceed to an evaluation, prepare them by providing developmental charts and schedule a follow-up visit in one to two months for discussion. Parents may also benefit from hearing a description of a developmental evaluation.

INITIAL MEDICAL EVALUATION

GD's initial medical evaluation should include a careful review of his medical history, including family

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history, and a physical examination with focus on dysmorphology and neurologic findings. If there are clues of possible medical etiology such as specific genetic conditions, neurologic, metabolic and/or mitochondrial issues, further work up may be considered. Note that children with communication delay or possible autism should have a formal audiological evaluation, even if they passed their newborn hearing screen. Periodic lead level checks should be done for children with active pica symptoms.[6] In the absence of specific clues, chromosomal microarray is recommended. For males, DNA testing for Fragile X is recommended.[7]

COMMUNITY RESOURCES

Pediatric subspecialists (such as developmental and behavior pediatricians (DBP), neurology and psychiatry) and experienced PCPs can perform diagnostic evaluations in conjunction with child psychologists, educators, speech-language pathologists, occupational and physical therapists.[1] In Arkansas, diagnostic evaluations led by DBP physicians are accessed primarily at the UAMS Dennis Development Center in Little Rock. The UAMS Schmieding Center in Lowell provides diagnostic evaluations led by neuropsychologists. Information for both programs are found on **http://pediatrics.uams.edu/clinical-programs-affiliates/**. Alternatively, consultation for child neurology and child psychiatry are also available within the UAMS and Arkansas Children's Hospital system. Information regarding these programs may be searched for at **http://www.archildrens.org/a-to-z-services-list**.

Subspecialty care in Arkansas is very limited, tending to cluster in metropolitan areas. To address this gap, the UAMS Developmental Behavioral Pediatrics section is working with the state to extend developmental specialty care into the community through the DBP Outreach Clinics and Arkansas Co-BALT program. For outreach clinics, developmental teams (DBP, social worker and pediatric nurse) conduct quarterly clinics in underserved areas around the state. The team conducts diagnostic interviews and follow-up with children needing developmental monitoring. For outreach clinic appointments, PCPs may send referrals to the Dennis Developmental Center.

The Arkansas Co-BALT program trains and mentors mini-teams of community-based clinicians to provide developmental evaluations. Mini-teams include a PCP and either a speech-language pathologist or nurse. These teams receive an intensive three-day training course on conducting diagnostic interviews and specific developmental assessments from the Co-BALT home team in Little Rock. Schedule appointments by contacting teams directly at **www.cobaltar.org**.

The Individuals with Disabilities Education Act (IDEA) requires each state to provide a free and appropriate public education, in the least restrictive environment, to all eligible children with disabilities from birth to 21 years. Part C of IDEA pertains to EI services for children birth to 2 years, 11 months. The Arkansas EI program is called First Connections, (**https://dhs.arkansas.gov/dds/Firstconnectionsweb/#fc-home**). Referrals may be completed online or by phone 1-800-643-8258 or 1-501-682-8158; fax referrals to 1-501-683-4745.

As GD and his family pursue services, his PCP stands as the center of his medical home, providing referral to community resources, helping explain evaluation results, coordinating services and monitoring GD's progress. ▲

Drs. Lopez, Fussell and Schulz are faculty in the Section of Developmental Behavioral Pediatrics at UAMS; Dr. Rodgers is Chief Medical Officer for AFMC.

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Amebic Abscess In a Patient Presenting With Abdominal Pain and Weight Loss

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ABSTRACT

A 40-year-old male with no significant past medical history presented to our clinic with cough, right upper quadrant pain, fatigue and weight loss over a period of eight months. Magnetic Resonance Imaging (MRI) demonstrated a mass in the liver. Amebic liver abscess was confirmed serologically. Patient responded well to antibiotics and the post hospital follow-up in clinic correlated with a good clinical outcome. Amebic liver abscess should be suspected in patients presenting with unexplained right upper quadrant pain (with no definitive etiology identified after the initial evaluation). Since it is quite uncommon in the United States (U.S.), there is a tendency to miss the diagnosis. High index of suspicion is needed in endemic areas of the US and in risk groups.

INTRODUCTION

Amebic liver abscess (ALA) is the commonest extra-intestinal manifestation of Entamoeba histolytica, affecting greater than 50 million people worldwide with highest prevalence in Central and South America, Mexico, India, South West Asia, Africa.[1] The incidence of ALA has decreased in the United States. Young hispanic males in the southwestern states are mostly affected amongst other risk groups. Decreasing incidence thought to be due to shifts in migration to or from endemic countries, better outpatient management in the United States leading to earlier diagnosis and a decreasing burden in countries like Mexico.[2] ALA remains a serious but treatable disease if identified early. Disease typically affects men aged between 40-50 years.[3] Transmission is through fecal-oral route, most common symptom is abdominal pain presenting along with other constitutional symptoms.

CASE REPORT

A 40-year-old male presented to our clinic with a three-month history of non-productive cough, fatigue, right upper abdominal episodic pain, subjective fever, unintentional weight loss of 60 pounds over eight months. He had visited a physician about a month prior to presentation and was diagnosed with gastro-esophageal reflux disease. He was started on a proton pump inhibitor that did not alleviate his symptoms. Past medical history was significant for syphilis treated a year prior to presentation. The patient smoked one pack of cigarettes per day for the last 20 years, consumed 24 ounces of beer daily for 20 years, and smoked marijuana and cocaine for five years, having quit three years prior to presentation. He had three heterosexual partners over the past two years and denied intravenous drug abuse. He has never traveled outside the United States, but he works outdoors everyday as he owns a landscaping company. Patient was not taking any medications at the time of presentation. Physical examination revealed a cachectic looking middle-aged man and abdominal examination was significant for right upper quadrant tenderness, without guarding or rebound tenderness. There was no abdominal distension or a palpable mass. Chest exam was significant for decreased breath sounds on the right middle and lower lung zones. During his initial visit in the clinic, he was febrile with temperature of 100.7ºF, tachycardic with a heart rate of 120 beats per minute, hypotensive with a blood pressure of 98/54 mm Hg, and his respiratory rate was 18 beats/minute. Liver function tests (LFTs) were unremarkable except for an elevated aspartate transaminase (AST) level of 308. Hepatitis panel was negative except for a positive hepatitis B surface antibody. Human Immunodeficiency Virus (HIV) was negative and thyroid-stimulating hormone (TSH) was within normal limits. Chest x-ray showed a right hemi-diaphragm with bibasilar atelectasis. Computer tomography abdomen revealed 15 x 13 x 16 cm mass in the right hepatic lobe. Patient was brought back to clinic for a follow-up visit. On the second visit, a week later, his symptoms persisted and hence gastroenterology was consulted over the phone. They considered hydatid cyst as a likely diagnosis and suggested that patient should not be subjected to diagnostic paracentesis given the risk of anaphylaxis. Infectious disease team was also contacted who considered amebic liver abscess as the most likely diagnosis and recommended admission to the hospital for further evaluation and diagnostic paracentesis. Laboratory investigations showed hemoglobin of 9.7 g/dl, white blood cell count of 11.32 k/ul, and mild monocytosis of 1.1%. Prothrombin time/international normalized ratio was 17.2 sec /1.4. Stool was negative for ova and parasites. Serum cancer antigen (CA) 19-9 was elevated at 68.8 U/mL while serum carcinogenic embryonic antigen (CEA) was normal. Rapid plasma regain (RPR) was reactive with a concentration of 1:16. Histoplasma urine antigen was negative. Serum erythrocyte sedimentation rate (ESR) was elevated at 140mm/hr. Blood cultures were negative and tuberculosis spot test (T-Spot TB) was unremark-

> Continued on page 256.

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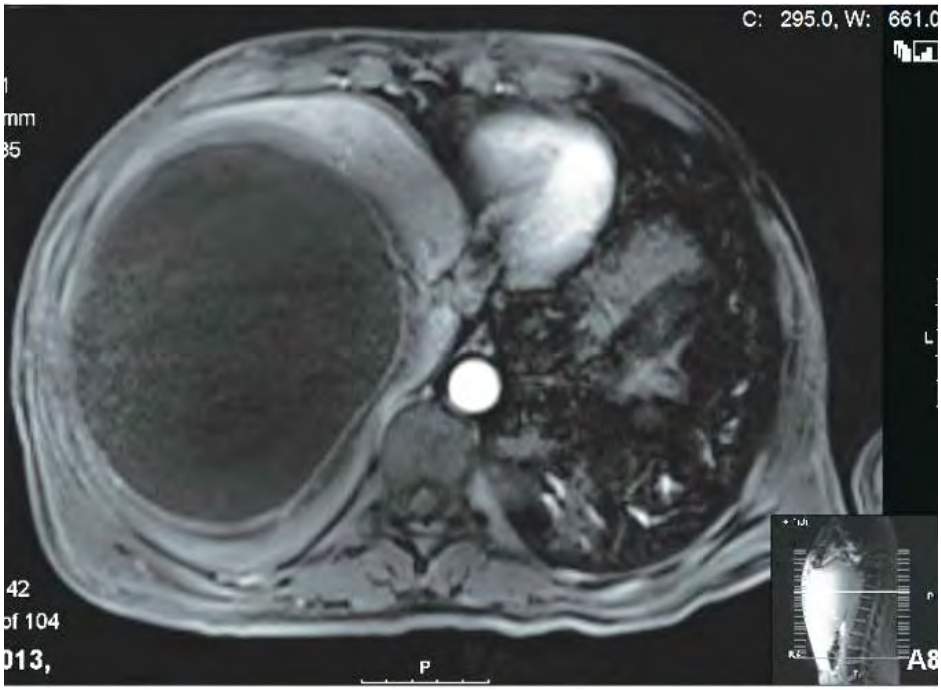

Figure 1. T1 image. MRI showing large hypodense abscess prior to drainage.

able. Serum alpha-fetoprotein (AFP) was normal. Indirect hemagglutinination (IHA) serology was positive for immunoglobulin antibody G (IgG) to entemoeba histolytica with a titer of 6.66 IU. Stool microscopy, antigen detection, and molecular testing were not done. Magnetic Resonance Imaging (Fig. 1) done to characterize the CT findings showed a 15.4 x 12.9 x 16.3 cm mass with no septations consistent with CT findings. He subsequently had CT-guided drainage and removal of 1.7 Liters of pus from the liver. He was started on IV metronidazole and levofloxacin empirically and levofloxacin was discontinued on day two once serology was suggestive of Entamoeba histolytica. Gram stain of abscess fluid revealed moderate polymorphnuclear cells and culture yielded no growth. AFB smear was negative for acid-fast bacilli by fluorescent microscopy and culture was negative for mycobacteria after six weeks of incubation. Fungal culture was negative and pathology report was also negative for any malignancy. Infectious Disease team was involved in managing patient and they suggested 10-day course of oral metronidazole 750mg three times daily (TID) and subsequently paramomycin 750mg TID for seven days, which was completed by patient. He showed remarkable improvement on oral metronidazole while in the hospital. He was discharged with the aforementioned antibiotics. A pigtail drain was left on discharge and he was to follow up with his primary care physician (PCP) in two weeks. The CT scan at discharge revealed significant decrease of abscess cavity to 12.5 x 9.2 x 11.8 cm. He had two follow-up appointments within two months of discharge with his PCP, by which time his symptoms had resolved and the patient had gained a remarkable 23 pounds.

> *Physicians should be aware that amebic liver disease could be seen in patients who have not traveled outside of United States to endemic regions.*

DISCUSSION

Amebic liver abscess is caused by ingestion of infected Entamoeba histolytica cysts through fecal-oral route. Humans ingest infected food or water; cysts degrade in the small intestine releasing trophozoites that invade the mucosa causing disease. Clinical features include fever, chills, right-sided abdominal pain and tenderness, nausea, vomiting, right-sided pleuritic chest pain, weight loss, and anorexia. ALA is ten times more common in males than females, possibly due to hormonal factors and sex differences in the background of liver disease.[4] High occurrence in men is also believed to be associated with increased alcohol intoxication leading to impaired kupffer cell function resulting in immunosuppression.[5] Travel to endemic regions plays a major part in development of the disease but in the United States, a study revealed that almost one-third of U.S. patients diagnosed of amebic liver abscess have not visited endemic areas.[5] Amebic liver abscess in this population is more prevalent in immigrants from endemic region, immunosuppressed individuals including people with HIV, people who are malnourished with severe hypoalbuminemia, or the ones with a history of alcohol abuse.[5]

The most common complication is hepatic rupture. Other possible complications are peritonitis, paralytic ileus, and toxic megacolon.[6] The right lobe of the liver is more affected than the left due to blood supply from superior mesenteric vein. Mild to moderate leukocytosis, elevation of liver enzymes can be seen. Since 70% of patients with amebic liver abscess do not have detectable parasites in the stool, serological or antigenic testing should be used for confirmatory purposes. Serology may be negative in the first week of infection but usually is positive at presentation in most cases.[7] This does not differentiate between current and past infections and thus remains unreliable in endemic areas. Enzyme immunoassay becomes a valuable test in diagnosing amebic infection in non-endemic areas. Other diagnostic options are liver exudate antigen assay or polymerase chain reaction (PCR) on material from liver abscess. Ultrasound, CT, MRI, nuclear hepatic scan are various imaging options for evaluating suspected patients, but only the nuclear hepatic scan can differentiate amebic liver abscess from pyogenic abscess.[8]

Treatment of choice is metronidazole 750mg three times daily for 10 days, followed by luminal agents like paramomycin, iodoquinol, or diloxanide fumigate. Patients usually respond to medical therapy. Therapeutic aspiration is considered when abscess is greater than 5 cm in size, if abscess is located in the left lobe of liver or in cases that fail to respond to medical management in 5-7 days.[9] Imaging-guided percutaneous needle drainage combined with antibiotics is considered standard modality of treatment and surgery remains an option only after failure of such initial treatment. Early recognition and treatment is associated with better outcomes. Poor prognostic factors include albumin less than 2.0mg/dl, bilirubin level greater than 3.5mg/dl, and patients with encephalopathy.[10] Radiologic disappearance of abscess can take between 3-19 months; hence repeat imaging is unnecessary if clinical improvement is discernible.[11]

Our patient was noted to have clinical signs and symptoms of the disease. Given the history of chronic alcohol consumption, supported by imaging and positive serology in an individual from a non-endemic region, we were able to make a diagnosis and manage the patient accordingly, resulting in an appropriate therapeutic response.

CONCLUSION

Physicians should be aware that amebic liver disease could be seen in patients who have not traveled outside of the United States to endemic regions. With one-third of patients in the United States diagnosed of amebic liver abscess not having visited endemic areas, high index of suspicion should be considered in men with history of alcohol abuse, features of immunosuppression, or unexplained weight loss presenting with unexplained right-sided abdominal pain. This is likely to be missed in patients who have not visited areas with high prevalence such as the patient in our discussion. Early imaging should be considered in patients with concerning symptoms to facilitate a diagnosis and to prevent severe complications like hepatic rupture and peritonitis. These complications may require immediate surgical intervention and have an increased risk of mortality.

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Rheumatic Heart Disease in Marshallese Youth in Northwest Arkansas: A Case Series

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Abbreviations

RHD, rheumatic heart disease; ARF, acute rheumatic fever; AR, Arkansas

Abstract

Rheumatic heart disease (RHD) is the leading cause of pediatric acquired cardiac disease worldwide. It is now rare in developed countries, but still prevalent in lower income countries with some of the highest rates in Pacific Island nations. The Marshallese population in northwest Arkansas has significant risk factors for RHD coupled with financial and cultural barriers to health care access.

This paper describes the clinical course of three Marshallese adolescents with RHD, all of whom presented in critical condition and developed heart failure requiring surgery. We review their risk factors and discuss ways to identify children earlier in the disease course.

Introduction

Rheumatic heart disease (RHD) is the leading cause of pediatric acquired cardiac disease. It is a long-term sequela of acute rheumatic fever (ARF), a multi-system inflammatory disease following group A streptococcal pharyngitis. ARF is diagnosed using the Jones criteria summarized in Table 1.[1] Between 40-80% of children with ARF have cardiac involvement during the initial illness. Children with recurrent ARF or with moderate to severe valve disease on initial presentation can progress to chronic RHD. World Heart Federation echocardiographic criteria for RHD are summarized in Table 2.[2]

Of the estimated 336,000 cases of ARF and 282,000 cases of RHD each year, 95% occur in less developed countries. In developed countries, immigrants and indigenous minority groups are disproportionately affected.[3,4] In our institution, we recently treated three adolescent patients with RHD. All presented in critical condition requiring surgical intervention, and all were of Marshallese origin.

Historical Background

After WWII, the United States administered the Marshall Islands as part of the UN Trust Territory of the Pacific Islands. Several atolls were used as nuclear testing sites from 1946-1958. In 1982, the U.S. government and the new Republic of the Marshall Islands signed the Compact of Free Association, which provides compensation for claims related to the effects of the nuclear testing and allows Marshallese citizens to lawfully reside, work, and study in the United States without a visa.[5] Since then, Marshallese have been immigrating to northwest Arkansas, drawn by jobs in the poultry industry and the relatively low cost of living. An estimated 4,000+ Marshallese live in Arkansas, an increase of 250% from 2000-2010. Sixty-two percent were born overseas. Many face significant challenges after immigrating, with lower socioeconomic status, living conditions, and educational achievement compared to the state average.[6,7]

Cases

Patient 1. A 15-year-old Marshallese boy presented with fever and arthralgia, progressing to tachypnea, orthopnea, and chest pain. Exam included bounding pulses, jugular venous distention, pericardial friction rub, and respiratory distress. Echocardiogram showed a large pericardial effusion, moderate mitral insufficiency with a normal-appearing valve, and mild-moderate aortic valve insufficiency. Relevant laboratory studies are presented in Table 3. He was diagnosed with ARF with carditis, requiring drainage of the pericardial effusion. He completed a course of penicillin and was discharged with furosemide, lisinopril, and aspirin. Follow-up was arranged with monthly penicillin injections for secondary prophylaxis. He was subse-

Table 1. Revised Jones Criteria for Diagnosis of Initial ARF [1]

Diagnosis requires laboratory evidence of recent streptococcal infection, plus 2 major criteria or 1 major and 2 minor criteria.
Major criteria
• Carditis
• Polyarthritis
• Chorea
• Erythem marginatum
• Subcutaneous nodules
Minor criteria
• Polyarthralgia
• Fever > 38.5ºC
• Peak ESR>60 mm/hr and/pr CRP greater than upper limit of normal for laboratory
• Prolonged PR interval, if carditis is not a major criterion

quently lost to follow-up despite multiple attempts to contact the family. Nearly two years later, he returned to cardiology clinic with worsening exercise intolerance and had not taken his diuretics or penicillin in several months. Repeat echocardiogram showed worsened panvalvular insufficiency with moderately thickened mitral valve, severe mitral regurgitation, and left ventricular dilation. He underwent mitral valve leaflet extension with annular ring placement, posteromedial commissuroplasty, and aortic valve repair with cusp augmentation. At routine follow-up six months afterwards, he was found to be in asymptomatic atrial fibrillation with 3:1 conduction. He required synchronized cardioversion and is maintained on digoxin and flecainide in addition to his diuretics.

Patient 2. A 15-year-old Marshallese girl presented with several weeks of progressive cough, fatigue, exercise intolerance, lower extremity edema, and orthopnea. She was tachycardic and hypotensive with widened pulse pressure. After fluid resuscitation she developed pulmonary edema requiring intubation. Echocardiogram showed severe left atrial dilation, severe mitral valve insufficiency, and severe aortic valve insufficiency with rolled edges of valve leaflets. The mitral valve had a shortened posterior leaflet and a "hockey-stick" anterior leaflet. Laboratory studies were consistent with prior streptococcal infection (Table 3). She underwent mitral valve replacement and aortic valve replacement with posterior annular enlargement. Postoperatively she had left vocal cord dysfunction requiring thickened liquids. She was discharged on warfarin, furosemide, and ranitidine. Follow-up with INR checks and monthly penicillin injections was also arranged. Three months after discharge her INR was subtherapeutic. Multiple unsuccessful attempts were made to contact the family. They did, however, come to her cardiology appointment several weeks later, where she was found to have severe stenosis of her artificial aortic valve with peak gradient 65.6 mmHg. She reported poor compliance with her warfarin. After heparin anticoagulation, her repeat echocardiogram was improved, with moderate residual stenosis and peak gradient 51.3 mmHg.

Patient 3. A 15-year-old Marshallese girl presented with several weeks of worsening dyspnea and exercise intolerance, and 3-4 days of nausea, vomiting, abdominal pain, and chest discomfort. On exam she had a hyperdynamic precordium, 3/6 systolic murmur at the apex and left sternal border, and hepatomegaly. Echocardiograms showed moderate tricuspid valve insufficiency with redundant valve leaflet edges, severe mitral valve insufficiency with annular dilatation of 40-42mm, severe left atrial dilation, and moderate left ventricular dilation. The mitral valve was thickened with redundant valve edges, abnormal coaptation, and shortened posterolateral valve leaflet. Laboratory studies were consistent with prior streptococcal infection. She underwent mitral valve repair with ring placement, annuloplasty, and valvuloplasty, and tricuspid valve repair with ring placement, valvuloplasty, and anterior leaflet cleft repair. Postoperative echoes showed worsening aortic valve insufficiency and diminished ventricular function. She was discharged on aspirin, furosemide, spironolactone, lisinopril, omeprazole, iron supplements, and monthly penicillin VK. At her cardiology clinic follow-up two months later, her symptoms had improved and she was nearly back to her baseline activity, but imaging showed severe aortic insufficiency, moderate mitral and tricuspid insufficiency, and moderately diminished ventricular function.

Table 2. World Heart Federation criteria for echocardiographic diagnosis of RHD in individuals aged 20 years or younger [2]

Definite RHD (A, B, C, or D)	
A) Pathological MR an at least two morphological features of mitral valve RHD B) MS mean gradient >4 mmHg C) Pathological AR and at least two morphologic features of aortic valve RHD D) Borderline diseage of both the aortic and mitral valve	
Borderline RHD (A, B, or C)	
A) At least two morphological features of mirtal valve RHD without pathological MR or MS B) Pathological MR C) Pathological AR	
Doppler echocardiographic criteria for pathological regurgitation (must meet all four)	
Pathological MR	Pathological AR
1. Seen in two views	1. Seen in two views
2. Jet length >2 cm in at least 1 view*	2. Jet length >2 cm in at least 1 view*
3. Velocity >3 m/s for 1 complete envelope	3. Velocity >3 m/s in early diastole
4. Pan-systolic jet in at least one envelope	4. Pan-diastolic jet in at least one envelope
Morphologic features of RHD	
Mitral valve features	Aortic valve features
1. Anterio leaflet thickening >3 mm†	1. Irregular or focal thickening
2. Chordal thickening	2. Coaptation defect
3. Restricted leaflet motion	3. Restricted leaflet motion
4. Excessive leaflet tip motion during systole	4. Prolapse

MR, mitral regurgitation; MS, mitral stenosis; AR, aortic regurgitation
*A regurgitant jet length should be measured from the ven contracta to the last pixel of regurgitant color (blue or red)
† Leaflet thickness should be measured during diastole at full extension on a framce with maximal separation of the chordae from the leaflet tissue. Measurement should be taken at the thicket portion of the leftlet including focal thickening, beading, and nodularity.

Marshallese population in general has a cultural distrust of Western governments and health care systems stemming partly from the effects of the nuclear testing of the 1940s-50s.

Discussion

All three patients are from a highly specific group — Marshallese youth in northwest Arkansas. Several factors contribute to this population's increased risk for RHD.

Both ARF and RHD are endemic in the Pacific Islands with some of the highest rates in the world. Surveillance studies and retrospective reviews show them to be more common in patients of indigenous descent.[10-14] Studies from other regions

> Continued on page 260.

Table 3. Laboratory results at presentation (normal range)	Patient 1	Patient 2	Patient 3
Erythrocyte sedimentation rate, mm/hr (0-10)	>73	>73	6
C-reactive protein, mg/L (0-10)	195	167.7	18.4
Anti-streptolysin O antibody (<200)	1600-2400	800-1200	200-400
Anti-DNAse B, U/mL (<170)	757	576	559
Streptozymw	positive		
Pro-B type natriuretic peptide, pg/mL (0.2-1318)		7850	3660
Rheumatoid factor, IU/mL (0-12)	<8.6		
ANA Screen	not detected		
Anti-ds-DNA IgG	not detected		
HIV 1 & 2 antibody	nonreactive		nonreactive
T-spot TB	negative	negative	
M. tuberculosis IgG (<0.424)	0.297		

have suggested an association of RHD with specific HLA alleles,[15] although to our knowledge this has not yet been studied in the Pacific Island nations. Socioeconomic factors appear to play a role as well: household crowding, low maternal education, and maternal unemployment have all been associated with ARF and persistence of RHD.[8,16] In addition to limited health literacy and financial and language barriers to health care access, the Marshallese population in general has a cultural distrust of Western governments and health care systems stemming partly from the effects of the nuclear testing of the 1940s-50s.

Primary prevention of rheumatic fever by treating group A streptococcal pharyngitis is the ideal method of reducing RHD. Failing that, the recognition of children with ARF allows secondary prophylaxis to be initiated. But in low-resource settings the initial episode of ARF may often go undiagnosed as it did in two of our patients, making it reasonable to screen high-risk patients for RHD. Even children with "borderline" echocardiographic changes have an eight-fold increased risk of recurrent ARF and progression to definite RHD.[17] Secondary prophylaxis for patients with subclinical RHD reduces this risk and may even cause minor valve lesions to regress[8,17,18] Screening for asymptomatic RHD has traditionally relied on auscultation of pathologic murmurs, which fails to detect most RHD — in one high risk Australian group, the sensitivity ranged from 18-47%.[19]

Echocardiography is the gold standard in detecting RHD. Field studies in Uganda using handheld echo (with modified diagnostic criteria) show it to be a promising screening tool in low-resource settings.[20-23] In higher-income countries, standard portable echo-based screening is still likely cost-effective.[24,25] An echo-based screening approach in our setting would need to incorporate culturally appropriate outreach efforts for the Marshallese population, possibly church-based community health screenings. In the meantime, we suggest health care providers working with these patients should maintain an increased index of suspicion for ARF and RHD. When applicable, efforts should be made to educate about the importance of diagnosing and treating strep throat.

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OBITUARIES

POCAHONTAS – **Thomas Bernard DeClerk, MD**, passed away January 10, 2018. He received a Bachelor of Science degree in Chemistry and a Doctor of Medicine degree from the University of Arkansas and interned at St. Vincent Infirmary in Little Rock. He enter the Army and served in the Army Air Corps and United States Air Force, where he graduated from the School of Aviation Medicine and became a flight surgeon during World War II. He later earned an underwater medicine certification. After leaving the service, Dr. DeClerk returned to Pocahontas, where he practiced general medicine, surgery, and obstetrics for 37 years, delivering approximately 2,500 babies. Dr. DeClerk served as president of the Randolph County Medical Society and was a member of the Arkansas Medical Society and a member of the Fifty Year Club. He also served as chief of staff of Randolph County Medical Center.

MOUNTAIN HOME - **Maxwell G. Cheney, MD**, 84, passed away February 8, 2018. Dr. Cheney was a well-known physician in Mountain Home for over 52 years. He was instrumental in improving health care in Mountain Home. Some of his accomplishments include starting the second medical clinic in Mountain Home, starting a renal dialysis unit, placing the first pacemaker at Baxter Regional Medical Center, serving as Chief of Staff several times at BRMC, and recently being honored by the Mountain Home Education Foundation for dedicated community service to the sports program. Dr. Cheney enjoyed hunting — especially bow-hunting — and was an avid skeet shooter. He was a member of the Arkansas Medical Society and a member of the Fifty Year Club.

LITTLE ROCK – **James Clay Wellborn, Jr., MD**, 64, passed away January 23, 2018. Dr. Wellborn attended UAMS for medical school, where he was a Barton Research Scholar, a member of Alpha Omega Alpha Honor Society, and a recipient of the Lange Book Award and the E. Forest Ellis Award for Outstanding Surgical Student. After completing a general and vascular surgery residency at UAMS, he established his surgical practice in Stuttgart, Arkansas, where he continued growing his family and made lifelong friends. He moved his practice to Little Rock in 1988, and has been performing general and bariatric surgery for the past 30 years. Dr. Wellborn was the first surgeon in the state to perform laparoscopic gallbladder surgery. He was a member of the Arkansas Medical Society.

LITTLE ROCK – **Gilbert S. Campbell ("Gil"), MD**, passed away January 9. He was 94. From 1949 to 1950, he was a basic sciences instructor and head of the experimental laboratory at Walter Reed Army Medical Center in Washington, D.C. From 1950 to 1951, he served as captain and surgeon in the 3rd Battalion of the 7th Infantry Regiment of the United States Army Medical Corps in Japan and Korea, where he earned two Silver Stars, two Bronze Stars and a Purple Heart. He was highly regarded by his fellow infantrymen as a fearless and highly competent surgeon. In 1965, Campbell became a professor and chairman of the Department of Surgery at UAMS where he served for 18 years. He remained as a professor emeritus until his retirement in 2000. Among his numerous departmental accomplishments, his greatest joy was the recruiting, training, and mentoring a legion of outstanding residents who went on to successful medical careers of their own, mostly in Arkansas. He initiated the first thoracic surgery residency at UAMS and was inducted in 2004 into the inaugural class of the UAMS Hall of Fame. He was a member of the Arkansas Medical Society and the Fifty Year Club.

LITTLE ROCK – **Walter S. "Walt" Mizell, MD**, 103, passed away February 25, 2018. As a young man, he joined the Marine Corps, as he did not weigh enough to get into the U. S. Army. He was a Marine for five years, and one of the ships to which he was assigned was the Battleship USS Arkansas. A degree from Ouachita Baptist University followed. The patriot served his country as an infantry officer during World War II, followed by carrying out his ambition to go to Medical School at the University of Arkansas for Medical Sciences. For the next 20+ years, he dedicated himself to medicine, serving as a physician and as medical director of the Arkansas State Hospital in Benton. AMS

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