December 2022
Volume 75
Number 12

South Dakota medicine

THE JOURNAL OF THE SOUTH DAKOTA STATE MEDICAL ASSOCIATION

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December 2022
Volume 75 | Number 12

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The Journal of the South Dakota State Medical Association

Office of Publication
2600 W. 49th Street, Suite 100
Sioux Falls, SD 57105
605.336.1965
Fax 605.274.3274
www.sdsma.org

Editor
Keith Hansen, MD

SDSMA President
Lucio N. Margallo II, MD

Chief Executive Officer
Barbara A. Smith

Staff Editor
Elizabeth Reiss, MS
–ereiss@sdsma.org, 605.336.1965

Advertising Representative
Elizabeth Reiss, MS
–ereiss@sdsma.org, 605.336.1965

South Dakota Medicine
(ISSN 0038-3317)
Published 12 times per year with one special issue by the South Dakota State Medical Association.

The SDSMA thanks the University of South Dakota Sanford School of Medicine for its financial contributions toward the production of *South Dakota Medicine*.

Subscription price: $50 per year domestic
$65 per year foreign, $8.95 for single copy

Periodicals postage paid at Sioux Falls, South Dakota and additional mailing offices.

Postmaster: Send address changes to
South Dakota Medicine
2600 W. 49th Street, Suite 100
Sioux Falls, SD 57105

SDSMA Home Page: www.sdsma.org

AMA Home Page: www.ama-assn.org

Printer:
Walsworth
P.O. Box 370
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RECENT CHANGES IN FEDERAL STUDENT LOAN PROGRAMS

KADIN WHITE, CFP®, CHFC® *Lead Advisor*

As physicians, student debt is likely no stranger to you! For borrowers who have federal student loans, there has been a string of updates over the last couple of years. Below, I have provided the most recent headlines that might impact you or your family.

Pause on Payments
On August 24, the US Department of Education announced one final extension on student loan repayments. This final extension will run through December 31st, 2022. Payments will be set to resume on these loans starting in January of 2023.

Student Loan Forgiveness
In addition to the student loan repayment pause, the department of education announced student loan forgiveness for borrowers that meet certain eligibility requirements. Under the forgiveness plan, borrows who have income under $125,000 or married individuals with income less than $250,000 with Pell grants would be eligible for forgiveness of up to $20,000. Borrowers without Pell Grants who meet the same income thresholds will be eligible for forgiveness of up to $10,000.

How to Apply for Debt Forgiveness
While nearly eight million borrowers will have their debts relieved without filing an application, the department of education recommends that borrowers complete and submit applications for this debt relief by mid-November of this year. However, borrowers may continue to apply for debt relief even after the student loan pause expires at the end of 2022. Borrowers who file formal applications can expect debt relief within six weeks.

Public Service Loan Forgiveness (PSLF)
The Public Service Loan is a program that allows borrowers a path to forgiveness for time spent working in the public/non-profit sector. Borrowers become eligible for forgiveness after a specific number of "qualified" payments. In late 2021, changes to the PSFL program allowed borrowers to receive credit for prior payments that, otherwise, would be deemed ineligible. However, these temporary changes expired on October 31st.

If you have any questions on how these changes might impact, you or your family please reach out to our team. We would love to be part of the conversation.

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The Striking Down of *Roe* (and Physicians' Judgment)

Lucio N. Margallo II, MD
President, South Dakota State Medical Association

In this month's issue, we feature a point-counterpoint discussion on the *Dobbs* decision.

The U.S. Supreme Court's opinion in *Dobbs v. Jackson Women's Health Organization* in June overturned the nearly 50-year precedent set by *Roe v. Wade*. The *Dobbs* decision has led to changes in state laws across the nation, and more are expected.

Two historic U.S. Supreme Court rulings, *Roe v. Wade* (1973) and *Planned Parenthood of Southeastern Pennsylvania v. Casey* (1992), established and affirmed a constitutional right to obtain an abortion. The ruling in *Roe* based the right to abortion on the right to privacy which the Court found to be implicit (implied) in the "liberty" guarantee of the due process clause of the Fourteenth Amendment ("... nor shall any state deprive any person of life, liberty, or property, without due process of law"). Both the right to privacy and the right to abortion are however nowhere mentioned in the text of the U.S. Constitution.

Roe had recognized a constitutional right to obtain an abortion before approximately the end of the second trimester of pregnancy (which the Court construed as the usual period of fetal viability).

Casey affirmed the "essential holding" of *Roe*, which it had described in part as "a recognition of the right of the woman to choose to have an abortion before viability and to obtain it without undue interference from the State." *Casey* held that a state unduly interferes in the right to pre-viability abortion if its restrictions "impose ... an undue burden on a woman's ability to make this decision" or present "a substantial obstacle to the woman's effective right to elect the procedure."

Casey modified however *Roe*'s pregnancy trimester framework in favor of a fetal viability standard, typically 23 or 24 weeks into pregnancy (rather than at the 28 weeks as previously understood by the Court in *Roe*) while affirming that the right to abortion is not absolute, must be balanced with possible government interest, and may be affected by scientific and medical advancements that allow premature babies to survive at younger gestational ages.

Mississippi passed a law banning most abortions after the 15th week of pregnancy. The state in the case of *Dobbs v. Jackson Women's Health Organization* claimed that laws banning pre-viability abortion aren't unconstitutional. The Court in *Dobbs* agreed with Mississippi which in June 2022 overturned the nearly 50 years of judicial precedent first established by *Roe*, thereby allowing states to regulate and restrict abortion or even to ban it completely. *Dobbs*' entrenching of state's power has the outcome of retrenching a physician's ability and judgment that *Roe* has secured.

While the *Roe* and *Casey* rulings were in effect, some states passed what are known as trigger laws. Thirteen states including South Dakota enacted trigger laws that automatically banned abortion in the event *Roe* being overturned. South Dakota's trigger law (22-17-5.1) language is as follows: Procurement of abortion prohibited – Exception to preserve life of pregnant female – Felony. Any person who administers to any pregnant female or who prescribes or procures for any pregnant female any medicine, drug, or substance or uses or employs any instrument or other means with intent thereby to procure an abortion, unless there is appropriate and reasonable medical judgment that performance of an abortion is necessary to preserve the life of the pregnant female, is guilty of a Class 6 felony. (Section 7 of SL 2005, ch 187, as amended by SL 2005, ch 188, § 1, provides: "This Act is effective on the date that the states are recognized by the U.S. Supreme Court to have the authority to prohibit abortion at all stages of pregnancy.")

Editorial

Joy, Gratitude, Vitality

Jason J. Kemnitz, EdD

The University of South Dakota Sanford School of Medicine (USD SSOM) is now moving into the third year of our strategic plan. If you are not familiar with our strategic plan, it can best be described as a five-pointed star with kindness at the center. Each point represents an area of emphasis that we would like to strategically focus on to improve. The star includes Learners, Research and Scholarship, Relationships and Engagement, Diversity and Inclusion, and faculty and staff. Each topic area also contains a bevy of sub-goals and strategic initiatives. (If you'd like to see the plan you can simply search for USDSSOM Strategic Plan on a search engine and find the basic outline). For today's purpose we will specifically look at Goal II of the faculty and staff portion: Increase institutional vitality for staff, residents, faculty, and administration. I have thought a great deal about this goal, and I always come to the same questions. What is vitality and how do we increase our personal vitality?

Academic medicine has long sought to define what vitality means to faculty members. A quick PubMed search offers no less than 2,000 articles devoted to faculty vitality and while all mention the term few attempt to define it. We have defined that vitality is important for faculty to learn new skills and knowledge and that faculty who feel vital produce at a higher rate. We have further defined that vitality can identify an entire faculty body as well as a single faculty member. We have even operationalized it into an algorithm, $A_{cp}+F_1+S_c+AR_1....n=FV$. The most comprehensive list of definitions of Faculty Vitality for academic medicine lists fifteen separate definitions.[1] The most recent definition includes items like professional fulfillment, engagement, personal growth, and positive feelings. All of these items appear to have the same common denominator, they make us feel good or bring us joy. So, are we actually asking to increase joy? If so, how do we do that? As an institution, I am still working on it! Maybe we start as individuals, and once we have that sense of joy, we share how we achieve that feeling with others. In a recent interview author and researcher Brene Brown states "In the research, we learned that the most effective way to cultivate joy in our lives is to practice gratitude. The key word here is practice. It's not just about *feeling* grateful, it's about developing an observable practice. So often we think that joy makes us grateful when in reality it's gratitude that brings joy." So, as we hurtle into the holiday season where joy is the word of the day; perhaps, we do need to stop and practice intentional gratitude. I'll start.

I'm grateful for a faculty that is willing to help when needed. The SSOM put out a call for volunteers to help with mock interviews and were overwhelmed with the response from our faculty. Our faculty spend countless hours teaching our learners, often without reward or acknowledgment. Thank you.

I'm grateful for a student population that is challenging, flexible, and willing to teach me as much as I hope to teach them. Our students not only achieve academically but are good people as well. Most of our students have multiple community volunteer encounters to make a difference.

Finally, I'm grateful for my support system. Sarah, Zach, Aidan, Kevin, Megan, Carmen, and Mel. Without these folks, I'd probably be a hot mess. Not only do they keep me on track, they support some of my crazier ideas, act as sounding boards, and ground me when I need it.

Now it's your turn. I challenge you to take a moment in this busy season and sit quietly and name what you are grateful for. Perhaps it will increase your feeling of joy and you will feel more vital.

We say a fond farewell to Michael Koch, MD, who will be retiring as the department chair of Pathology after 20 years of service. We also welcome DesiRae Muirhead, MD, who will begin her term as chair on Dec. 19!

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About the Author:

Jason J. Kemnitz, EdD, Associate Dean, Academic Development and Faculty Affairs; Assistant Professor, Family Medicine, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

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Point-Counterpoint: *Dobbs* Decision

Point: The State of Abortion Care in South Dakota: Past, Present, and Future

Amrita Bhagia, MS II; and Henry Travers, MD

In January 2022, I met a 31-year-old pregnant Native American woman who had been diagnosed with severe fetal growth restriction early in her pregnancy. Hoping for a child, she continued her pregnancy. However, now in the second trimester of her pregnancy, it was clear that the fetus' kidneys were failing to develop, a lethal condition. This pregnancy would not be viable, and the offspring would either be stillborn or die shortly after birth.

As was the standard of care, the physician I was shadowing as part of my medical school training presented the option of terminating the pregnancy, which the patient agreed would be the best course of action for her. Naively, I assumed it would be a straightforward process to receive a medically-indicated procedure in the case of a non-viable pregnancy. However, due to the Hyde Amendment, Indian Health Services would not cover the cost of her abortion.[1] Planned Parenthood in Sioux Falls was no longer an option, as the doctor mentioned that they were only offering medical abortions up to 10 weeks at this point, and these fetal abnormalities had been detected later in pregnancy. While the clinic she was receiving care at could perform the procedure, South Dakota Medicaid would not pay for an abortion unless the pregnancy endangered the mother's life[2], and this woman did not have the funds to pay for a multi-thousand-dollar surgery out of pocket. Traveling out of state was also not possible based on her limited transportation and financial resources. With all options exhausted, this woman was denied any choice but to carry her non-viable pregnancy for another 25+ weeks to term.

As a medical student shadowing in a maternal-fetal medicine clinic, my scientific curiosity was piqued by the complexity of this case, but more than anything, the story of this woman invoked a deep sense of rage within me. This woman was not only grieving the loss of a child she had wanted but had been informed that she would be forced to carry this non-viable pregnancy to term because of restrictive federal laws, a statewide lack of access to care, and systemic disparities related to socioeconomic status and race/ethnicity. This woman lost both her child and her choice, and perhaps most painful of all, such injustice was not uncommon. Rather, the case of this 31-year-old woman and those like her demonstrates that abortions, even in South Dakota, have been and will continue to be medically necessary. As such, especially now that *Roe v. Wade* has been federally overturned, healthcare providers across the state must be aware of current legal and socioeconomic barriers to abortion and how they can continue to advocate for their patients despite these limitations.

As the above story illustrates, even before June 2022, abortion access in South Dakota was extremely limited. Although abortion was federally protected, the state had a strong anti-choice sentiment, which manifested in limited funding for organizations providing abortion care. As such, there was only one abortion clinic in the state, Planned Parenthood in Sioux Falls. Additionally, this Planned Parenthood clinic did not have a full-time abortion provider and instead transported a physician from Minneapolis twice a week, causing limited appointment availability.[3] Further restrictions imposed by the state made the process of getting an abortion much harder, including a 72-hour waiting period that "necessitat[ed] two trips to the facility[4]" for pregnant individuals as well as a ban on telemedicine and mail delivery services seeking to provide prescription drugs to South Dakotans for the use of medical abortions.[5]

Now, in late 2022, the federal protections that guaranteed the right to abortion, as established by *Roe v. Wade* and *Casey v. Planned Parenthood*, have been overturned by the *Dobbs v. Jackson* decision. In anticipation of this decision, Sioux Falls Planned Parenthood stopped scheduling abortions on June 15, 2022.[6] South Dakota's trigger law came into effect on the day of the Supreme Court decision itself, June 24, 2022. The trigger law prohibits individuals from administering, prescribing, or procuring medicines or instruments with the intent of using them for an abortion unless "an abortion is necessary to preserve the life of the pregnant female.[7]" This law seems to apply to *all* pregnancies, including non-viable pregnancies such as ectopic and molar pregnancies as well as

those with severe fetal abnormalities. Furthermore, while the law allows abortions *if the pregnant individual's life is threatened*, in line with the federal Emergency Medical Treatment and Active Labor Act, the most accepted meaning of this phrase suggests that this only applies to emergent conditions that cause acute, severe symptoms.[8,9] This has the potential to force pregnant individuals – even those with non-viable pregnancies - to wait until hemorrhage, septic shock, or other life-threatening consequences develop before they would be allowed an abortion. The law also has no exceptions for cases of rape or incest, and Governor Noem has indicated that she does not support the addition of any such exceptions to the law.[10] Healthcare providers failing to adhere to the trigger law can be charged with a Class 6 felony, which is punishable by two years imprisonment and/or a fine of $4000.[11] Alongside the trigger law, the ban on abortion-related telemedicine/mail delivery services remains intact through Governor Noem's Executive Order 2021-12, which prevents out of state individuals or companies from prescribing medication abortions to South Dakotans.[5]

While some aspects of the South Dakota laws around abortion remain vague and in need of additional legal analysis, these restrictions negatively impact the ability of healthcare providers to refer, advise, and perform necessary, time-sensitive medical care based on their best judgment. Both the American Medical Association and the American College of Obstetricians and Gynecologists have urged legislators to allow abortion to remain unrestricted on national and state levels and in the hands of trained healthcare providers and their patients.[12,13] Noting the past and current limitations on abortion care as well as the fact that abortion is still necessary in the state - both in cases where the pregnant individual chooses it for personal reasons as well as in cases in which the pregnancy is non-viable and/or poses a non-emergent threat to the pregnant individual's life or health – what can healthcare providers in South Dakota do to support their patients who are seeking an abortion?

First, remind your patients that while abortion is banned (except in very limited cases in South Dakota), it is legal to travel out of state for abortion care. As of this writing, in the region, abortion remains legal in Iowa, Nebraska, Minnesota, Colorado, and Montana - and all these states can provide appropriate abortion care. Abortion Finder (www.abortionfinder.org/) provides patients with a list of abortion clinics near them and information on state laws regarding abortion pertinent to each clinic (including waiting periods and other restrictions). Since deciding between clinics may be overwhelming for patients, individuals can search the Women's Reproductive Rights Assistance Project (WRRAP) (https://wrrap.org/assistance- services/find-clinic-abortion-provider/) for tips on appropriate questions to ask clinics and how to avoid crisis pregnancy centers – fake abortion clinics that often provide inaccurate information about reproductive care and are not subject to the same privacy and regulatory laws as healthcare facilities.[14] Because out-of-state travel and abortion care can be expensive, healthcare providers should also inform their patients that financial resources are available to help. WRRAP provides a list of funding sources for individuals who need assistance to obtain an abortion (https://wrrap.org/assistance-services/find- abortion-funds/) and can direct patients to relevant funds from state-specific organizations like South Dakota Access for Every Woman, the Midwest Access Coalition, and the Justice through Empowerment Network.

As you think about referring your patients to these resources, it is important that you also protect yourself from legal risk. While several legal experts interpret referring your patients to abortion-specific resources as within the confines of the law, because the waters remain untested, it is strongly suggested that you ask your institution/hospital how they are protecting doctors who share these resources.

Second, regardless of medical specialty or the age or gender of your patients, take a moment in subsequent clinic visits to ensure your patients are informed about their reproductive health. Do they have access to short and long-term contraceptive care if they desire? Do they know how to reliably use their birth control to effectively prevent pregnancy? Third, when political actions interfere with healthcare practice, healthcare providers take political action to change public policy. Share your expertise as a medical provider regarding how restrictions on abortion negatively affect your patients. Write to your legislators, testify to your colleagues, and vote in a way that honors the importance of this issue.

Overall, it is clear that abortion is severely restricted in our state. When regulations interfere with the best interests of our patients and our ability to provide essential services, it is the duty of healthcare providers to find new ways to help their patients. I began this essay with the story of a 31-year-old woman who was forced to carry her pregnancy to term despite having a non-viable pregnancy because she couldn't access abortion care in the state. I felt great sadness and pain for her, but I didn't do anything to help her situation. At that moment, I failed to take action. Yes, I am a medical student,

and yes, it was my first time encountering such a situation. Nonetheless, I let this woman continue without the support she needed. I can't turn back time, but next time maybe the outcome will be different. Maybe if she chooses you as her provider, she will find an advocate beside her.

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Please note: Due to limited space, we are unable to list all references. You may contact South Dakota Medicine at ereiss@sdsma.org for a complete listing.

About the Authors:

Amrita Bhagia, MS II; University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Henry Travers, MD, FACP, Clinical Professor of Pathology, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Counterpoint: Protecting South Dakota Women

Patricia K. Giebink, MD

The book *Unexpected Choice: An Abortion Doctor's Journey to Pro-Life* details my time spent working on both sides of the divisive issue of elective abortion. Because of my experiences not only as an abortion doctor but also as an obstetrician committed to both my patients – mother and child – I can offer a unique perspective on the effect that the recent Supreme Court decision in *Dobbs v. Jackson* will have on physicians and our patients. Since the day I started practicing medicine, I have seen the topic of abortion go from a near-taboo to the cause of a public and unapologetic shouting match. But the medical community's abortion discourse should not be about which abstract ideology is right, but rather about how can we show compassion, empathy, respect and understanding, and how we can find truth and balance in this post-*Roe* world.

When I first started working at the Sioux Falls Planned Parenthood in 1995, I felt it was important that women have access to legal abortions. After all, I remembered the horrific numbers circulating in the media about women hurt or dead from illegal abortions every year prior to *Roe v. Wade*. Thousands of deaths and hundreds of thousands of serious injuries, I was told. Preventing those deaths by offering women safe abortions seemed like a noble cause. Today, I know that those morbidity and mortality numbers were grossly inflated. In fact, Dr. Bernard Nathanson, cofounder of the National Abortion Rights Action League and pivotal figure in the pre-*Roe* fight for abortion, eventually admitted that the "thousands of deaths per year" figure was simply made up.[2] In reality, a CDC report shows that the number of deaths in the year prior to *Roe* was actually around 24.[3] Of course, 24 deaths is far too many – but the promulgation of this false information by major media outlets exemplifies the way in which proponents of certain ideologies garnered sympathy for their ideas in the nation's medical discourse. Through these manufactured numbers and other false narratives, medical professionals like me whose careers center around a passion for women's healthcare are made to feel that providing abortions is essential to serving their patients.

Organizations such as the American Medical Association (AMA), and the American College of Obstetricians and Gynecologists (ACOG) have staunchly advocated for unrestricted elective abortion access. But they do not speak for all doctors – especially obstetricians, 75-93 percent of whom do not perform elective abortions in their practice.[4] I know physicians who have left professional organizations over their increasingly extreme abortion views. The American Association of Pro-Life Obstetricians and Gynecologists (AAPLOG) was formed by OB-GYNs who disagreed with ACOG's abortion views, and today boasts a membership of over 7,000 medical professionals of all specialties.[5] Over the years, AAPLOG members have published evidence-based research and developed practice guidelines, available at www.aaplog.org, defending a life-affirming philosophy of medical care.

Having served patients facing a wide swath of pregnancy complications in South Dakota, and having treated patients in underserved countries such as Pakistan, I empathize with obstetricians facing challenging circumstances. And I empathize with the common concern that South Dakota's abortion laws may restrict doctors' ability to treat potentially life-threatening pregnancy complications such as miscarriage, ectopic pregnancy, and chorioamnionitis. Some of the confusion is over the definition of abortion, since "abortion" has about 10 different medical definitions. However, our state's legal definition is fairly straightforward, and it hinges on intent: abortion is the intentional ending of a human life in utero.[6]

In contrast, treating a miscarriage involves removing tissue from a pregnancy that has already been confirmed by two reference points or medical tests to be nonviable. There should be no confusion about whether this is the same as an elective abortion; at no point does the physician end the life of a fetus or embryo. Ending a pregnancy to save the mother's life should not be confused with elective abortion, either, nor does our state law recognize it as such.[7] For example, molar pregnancies and chorioamnionitis can be life-threatening, requiring treatments that result in pregnancy loss. However, the loss of life is not a goal of these treatments; the physician's only aim is to save the life of the mother. In such cases, a choice is being made between saving mom but losing baby or losing both; making the right choice is not a felony. This means that, in South Dakota, physicians can still end pregnancies under most circumstances; the only thing that's no longer legally permissible is ending a pregnancy with the intent of ending the fetus's life, unless necessary to save the mother's life.

To some physicians, this begs the question, how do we know when a mother's life is in sufficient danger to warrant termination? To offer support to those who may have this question, AAPLOG published guidelines outlining some criteria to determine when terminating a pregnancy would be acceptable under state abortion laws' life-of-mother exceptions. But no list can account for every possible medical case. This is where our training comes in. Determining when to end a pregnancy isn't the only time physicians need to rely on their own sound medical judgement. It's part of our job: making reasonable calls based on the information we have to maximize chances of good outcomes for our patients.

So, the narrative that pro-life laws will harm patients by forcing doctors to let them bleed out or die of sepsis while they wait for approval from the hospital's lawyers is entirely false. It is entirely possible to offer the highest quality medical care without abortion; in fact, withholding lifesaving care from patients amounts to malpractice.

A similar concern about abortion restrictions is that parents will be harmed by not being able to terminate pregnancies after receiving a fatal fetal diagnosis. A few years ago, I spoke with a Sioux Falls perinatologist who is not unsympathetic to pro-life issues but felt that doctors needed something to offer parents carrying a baby with a terminal fetal diagnosis. In addition to carrying the baby to term or near term, they wanted to offer timely termination to shorten the period of grief. Historically, we have assumed that the answer to a fatal diagnosis is termination, under the assumption that it's the only way to prevent the fetus's suffering, or that a mother shouldn't carry a pregnancy that, because of the child's short predicted lifespan, we deem "unviable". But is that in these parents' or their children's best interest?

For one, we could be wrong about the child's prospects. We've all treated patients whose health flourished beyond our expectations, either due to misdiagnosis or just luck. A recent study reported by the New York Times uncovered that many parents are inadequately informed about the staggeringly high rate of false positives in prenatal genetic screening tests, which may have led them to abort healthy children.[8]

And with the scientific developments we have seen in the past several decades that make ever more obvious the humanity and value of human beings in the womb, it may be time to reconsider the way we respond to the news of a life-limiting diagnosis. The most up-to-date research demonstrates that fetuses may be able to feel pain as early as 12 weeks' gestation.[9] And as medical technology advances, the gestational age at which they are able to survive outside the womb moves earlier and earlier. At the time of the *Roe* decision, the age of viability stood at 28 weeks; today, it's closer to 22-24 weeks.[10] From 4-D ultrasounds providing detailed models of our babies' faces to fetal surgery treating medical conditions such as spina bifida in utero, the scientific advancements that we have achieved since 1973 allow us to interact ever more closely with embryonal and fetal humans. Our medical practices should reflect today's science. Our society can only be uplifted when we decide to treat the most vulnerable among us with the dignity and respect that they deserve. Recognizing that deliberately and unnecessarily ending their lives violates this dignity is only one necessary step we need to take.

For families of children with fatal prenatal diagnoses, this might mean offering them the same kind of support that we give families with born children who are nearing the end of their lives: palliative care. Perinatal hospice is a service offered by hospitals across the country.[11] It allows parents to be with their child in their final moments before natural death prior to or shortly after birth, to say goodbye, and to fully grieve. Studies show that this service yields similar rates of complex grief but lower rates of anxiety for parents than termination.[12] Perinatal hospice offers many advantages over termination, such as a more natural grief process and closure, removing the potential guilt of having to make the decision to terminate their baby's life, and an option that involves a baby to hold and love. More stories are being revealed of families who chose perinatal hospice with a support team of people to guide families through this process to a more natural conclusion, and a birth that involves all family members to come together and participate in the life of this new little person, however brief it may be. These are the stories that break your heart in a good way.

Our state's new abortion ban brings home the fact that elective abortion is not healthcare; it treats no medical conditions because pregnancy is not a disease. Poverty, abusive relationship, mental illness, a lack of social and emotional support are all reasons that pregnant women may feel unprepared to have children, but abortion does nothing to solve these underlying problems. Instead, it only allows our society to avoid dealing with them. Once we stop relying on ending lives in utero as a way out of addressing these women's burdens, we can find real solutions for them. For policymakers, this may mean finding solutions to make housing, healthcare, and childcare more affordable. For nonprofits, it may mean creating more resources for women in crisis and offering help to empower them to feel confident as parents.

One type of nonprofit that offers such resources is pregnancy resource centers, which have been widely misrepresented in the media. The truth about these centers is that they largely provide compassionate information on abortion alternatives and available community resources. Centers like the Alpha Center in Sioux Falls and Black Hills Pregnancy Center in Rapid City have trained professionals to perform ultrasounds, to counsel, and provide other needed services. They offer parenting classes and programs, as well as material support. There is no arm-twisting and if the client chooses abortion, they also provide post-abortion counseling. Having served at one of these organizations, I can attest to the powerful ways in which they have helped vulnerable women confidently carry their pregnancies to term.

My passion has always been caring for underserved women. During my 20-plus years of delivering babies in Chamberlain, most of my patients were from Crow Creek or Lower Brule Indian reservations. I'm well aware of the socio-economic problems encountered on the rez, such as transportation and accessibility. But these problems are not insurmountable, and there are resources available not only through community workers, IHS, the tribe and rural hospitals. Ensuring that patients have access to these resources is absolutely worth the effort if it means offering pregnant women and their babies life-affirming care rather than using abortion to avoid dealing with these problems.

It is an unreasonable fear to believe that one's ability to practice medicine is unnecessarily limited by not being able to provide elective abortion on demand. Instead consider the hidden cost of abortion: the risks and potential complications often not considered, many permanent effects physically and psychologically. A woman's circumstances may change as the pregnancy progresses. A significant number of people wish they had known all the risks, immediate and delayed, before making such an important decision. We need to return to the practice of valuing and protecting both our patients. It's what they deserve.

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About the Author:

Patricia Giebink, MD, Chamberlain, South Dakota.

The views expressed in this Point-Counterpoint feature do not necessarily represent the views of the journal or of the SDSMA.

PREVENT CONGENITAL SYPHILIS

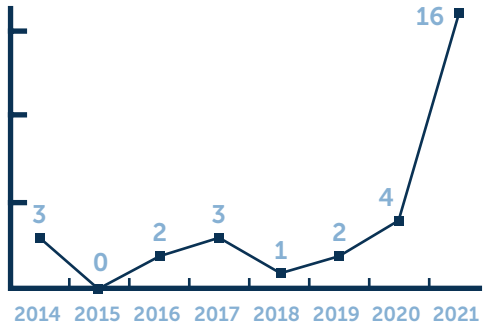
doh.sd.gov/std/

SOUTH DAKOTA DEPARTMENT OF HEALTH

16 Congenital syphilis cases reported to SD-DOH in 2021.

This represents a **700%** increase from the 5-year median.

2021 Provisional Data, as of March 1, 2022



PREGNANT MOTHER

TEST

TEST all pregnant women for syphilis at their first prenatal visit.

RE-TEST

RE-TEST all pregnant women at risk at 28 weeks AND again at delivery, including births, stillbirths, or terminations.

TREAT

TREAT* all women with diagnosed or suspected syphilis ***immediately*** using long-acting benzathine penicillin G; test & treat sex partner(s).

***CDC.gov/std/treatment**

NEONATE

TEST

TEST and examine all neonates born to mothers who have reactive nontreponemal and treponemal test results. Neonate's serum should be used for quantitative nontreponemal test.

RE-TEST

RE-TEST and examine all neonates with reactive nontreponemal tests every 2-3 months until the test becomes nonreactive.

TREAT

Maternal history of syphilis infection and treatment should be considered when **TREATING the neonate**. If mother's full history is unknown work with your local Disease Intervention Specialist to gather additional history.

Local disease intervention specialists can **SUPPORT YOU** with **gathering patient testing** and **treatment history** and with **partner notification**.

This publication was supported by Centers for Disease Control and Prevention (CDC) grant number, 6 NH25PS005161-03-04 as a part of an award totaling $1,000,000.00 with $0.00 state funding. The views expressed in this written material or publication do not necessarily reflect the official policies of the Centers for Disease Control and Prevention (CDC), Department of Health and Human Services.

Thumb and Index Finger Amputation after Radial Artery Occlusion in a Patient with COVID-19

Brandon T. Fisher, MD; Kelly McKnight, MD; Benjamin Jorgensen, MD; and Robert E. Van Demark Jr., MD

Abstract

The pathophysiology and predictability of radial artery thromboembolic events in patients with COVID-19 is not fully understood. We report a case of thumb and index finger gangrene and multiple digit amputations secondary to digital artery occlusion after radial artery cannulation in a patient admitted with COVID-19 pneumonia and encephalopathy. The exact association, causality, and potential hand manifestations in this patient population is unclear at this time, but is of particular interest in the current state of the pandemic.

Introduction

Radial artery occlusion (RAO) and embolization are rare complications of radial artery cannulation and transradial coronary procedures.[1] Clinical studies recognize a varying incidence that ranges from 2-10 percent, although this may be underreported.[2] Hand and finger ischemia secondary to RAO or embolization is rarely clinically relevant, as natural collateral perfusion and contribution from the ulnar artery are usually sufficient to overcome the deficit, although recent studies have shown the radial artery to have potential to be of dominant diameter at the level of the wrist.[2]

Devastating clinical consequences involving local digital ischemia and necrosis have potential to result in major morbidity and have been described in the literature since the 1970s.[1,3,4] Valentine et al[3] examined 8 patients over a five-year period that had radial artery thromboembolic events. All patients in their series had continued ischemia that eventually resulted in digital gangrene or amputation. Mangar et al.[5] describe a case requiring amputation of the hand after radial artery cannulation.

The incidence and clinical consequences of RAO/embolization in the setting of COVID-19 are not yet fully understood. There has been a growing acceptance of known association of hypercoagulability in these patients.[6] In this case, the patient had underlying risk factors for complications, but also had symptomatic COVID-19 upon initial hospital presentation.

We present a case of digital artery occlusion after radial artery cannulation in a patient admitted for COVID-19 pneumonia that resulted in multiple digit amputations. Given the current state of the pandemic, we find this case to be of interest for increasing awareness of hand manifestations due to pro-thrombotic events in these patients.

Case Report

The patient is a 62-year-old right hand dominant female with a past medical history that includes coronary disease, congestive heart failure, rheumatoid arthritis, and a 7.5 pack year smoking history that initially presented to the hospital for acute encephalopathy in the setting of COVID-19 pneumonia. She was admitted to the general COVID unit upon which time a radial artery cannula was placed secondary to borderline hypotension. She was placed on Enoxaparin for venous thromboembolic prophylaxis. The arterial line was in place for nine days, after which it was removed due to improving medical condition. The patient developed right thumb and index finger ischemia with

Figure 1. (A) Angiogram of the patient's right upper extremity showing avascularity of the radial artery terminal branches and insufficient ulnar artery collateral perfusion. (B) Angiogram with normal ulnar artery, radial artery and palmar arch.

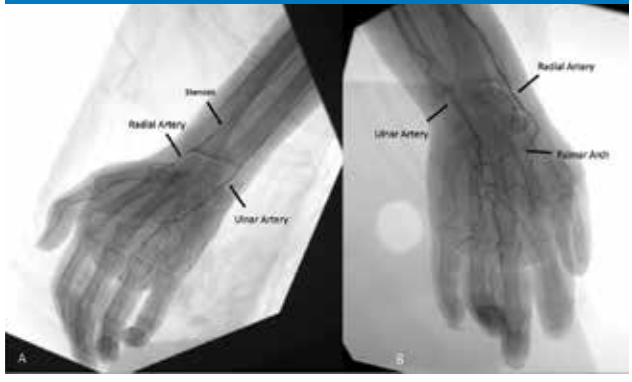


Figure 2. Clinical photographs 21 days post-admission.

Figure 3. Intraoperative photos of initial amputation.

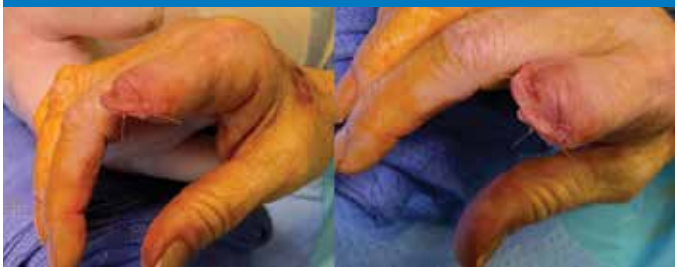


Figure 4. Clinical photographs three weeks following initial amputation.

Figure 5. Intraoperative photographs of index finger amputation revision and primary thumb amputation.

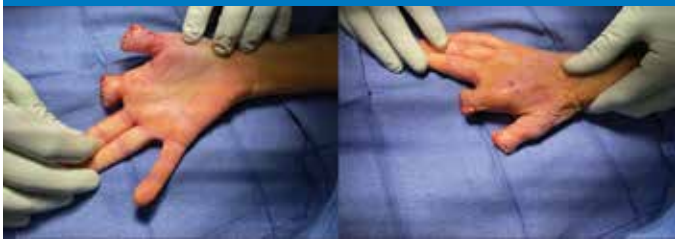


Figure 6. Final clinical photographs 11 weeks following initial procedure.

pain by hospital day 14. Noninvasive embolic workup was negative. Right upper extremity arterial and venous duplex showed no abnormalities with triphasic radial artery flow distally. Over the next 24 hours, the patient's thumb and index fingers clinically worsened. Bedside ultrasound was done that showed lack of continuity in the palmar arch and lack of flow to the index and thumb fingers of the right hand. She subsequently underwent right upper extremity angiogram demonstrating distal RAO and lack of an intact palmar arch (Figure 1A shows the abnormal angiogram and Figure 1B demonstrates a normal angiogram of the wrist). Tissue plasminogen activator was instilled for 20 minutes followed by suction thrombectomy and balloon angioplasty. There was a widely patent radial artery with perfusion of thumb and index finger following the angioplasty. She maintained adequate perfusion at the radial artery site of repair throughout the immediate postoperative period. However, the patient worsened clinically and orthopedic surgery was consulted for management of dry gangrenous fingers on post-admission day 21 (Figure 2). The patient subsequently underwent her first orthopedic procedure involving a right index finger amputation at the distal level of the middle phalanx under local anesthesia (Figure 3). The patient was discharged as an outpatient and was seen in follow up three weeks later with progression of tissue necrosis involving the index finger amputation site and the thumb tip (Figure 4). Therefore, the patient underwent revision amputation of the index finger stump to the level of the proximal interphalangeal joint and primary amputation of the thumb through the interphalangeal joint under local anesthesia (Figure 5). She was seen in the office approximately 11 weeks following her initial procedure. The wounds were well healed with minimal pain (Figure 6). Follow up arterial duplex showed widely patent radial artery. Her encephalopathy had resolved and she was living independently in her home.

Discussion

Over the last decade, the radial artery has become a preferred site of access for interventional cardiac and vascular procedures throughout the world due to superficial access, decreased major bleeding, and decreased overall cost.[2] Radial artery access is often utilized for cannulation by general surgeons, vascular surgeons, interventional radiologists, critical care physicians, registered nurse anesthetists, and/or residents alike. The rare, but devastating, consequences that lead to digital necrosis often go unmanaged by these primary proceduralists, as they are frequently managed by vascular and orthopedic surgeons.

The Society for Coronary Angiography and Interventions discusses an ideal patient for radial artery cannulation to be of hemodynamic stability, age <70, no history of prior ipsilateral brachial or transradial procedure, and a palpably large radial artery with a strong pulse in the presence of a normal Allen's test.[7] Contraindications listed include an absent palpable pulse, abnormal Allen's test (defined as reperfusion of the hand within 7 seconds), underlying vasospastic conditions (Raynaud's e.g.), planned or present arteriovenous shunt for dialysis, and potential use of radial artery as a conduit for aortocoronary bypass. The patient in this case met all ideal candidate criteria other than hemodynamic stability (which was borderline based on documented blood pressure readings). There was no documentation of Allen's testing prior to initial cannulation in this patient, but there was documentation of a strong palpable radial pulse. There are previous studies that question the validity of Allen's test. Jarvis et al[8] examined sensitivities and diagnostic accuracies of Allen's test in 93 patients with a mean age of 64, and reported findings that suggest a specific cut-off point for discriminatory/satisfactory testing does not exist, and as such recommended replacing this test with more objective measures such as Doppler ultrasound.

There are several suggested risk factors for hand ischemia after radial artery cannulation that include prior arterial injury, female gender, use of vasopressor support, hypotension, cannulation longer than seven days, malignancy, heparin-induced thrombocytopenia, disseminated intravascular coagulation, and hypercoagulable states.[3,9] The patient in this case report met several of the known risk factors, with perhaps the most interesting variable being hypercoagulability in the setting of COVID-19 infection. There are several recent studies regarding the effects of this novel virus and induced hypercoagulability, with many noting high frequencies of thrombosis and arterial thrombotic events secondary to proposed perpetuated thromboinflammation.[6] Kichloo et al.[6] and Singhania et al[10] describe an ill-defined pathogenesis in combination with Virchow's triad as a likely mechanism for the thrombotic events in patients with COVID-19. Although this patient had underlying risk factors for complications secondary to an indwelling catheter, we find this case to be of interest by drawing attention to the potential confounding effects of COVID-19 and the timing of initial clinical consequences (two to three weeks post-diagnosis) in this patient's clinical course and secondary outcomes.

In most cases, patients remain asymptomatic after RAO occlusion or thrombosis, but in rare cases (and in this case), ischemia and necrosis can occur.[1,4] There is some thought that nonoperative management prior to onset of gangrenous digits may be effective. Valentine et al.[3] utilized vasodilators and found their use to be equally effective in treating cannula-induced radial artery injuries in some patients. Garg et al.[1] recognized surgical intervention for radial artery occlusion to be successful, particularly in patients that are not critically ill. The patient in this case progressed to necrosis after radial artery revascularization, which again raises the question of pro-thrombotic effects of COVID-19.

Regardless of etiology, this patient sustained life-altering complications to her dominant hand that involved multiple subspecialty services, prolonged medical care, and increased hospital costs. She had documented risk factors for complications, but also met proper criteria for catheter placement. The confounding effects of COVID-19 hypercoagulability must not be ignored in the current state of the pandemic, and this case should raise awareness of this complication in this patient population.

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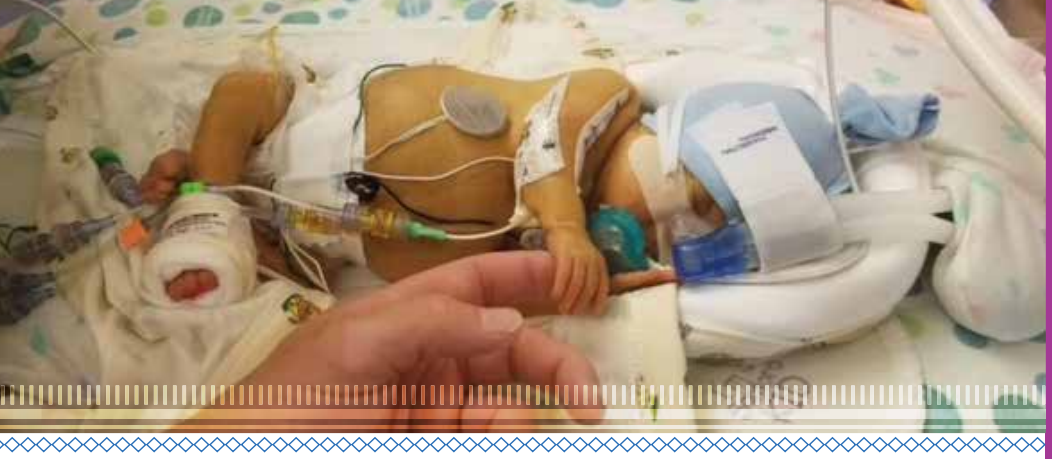
About the Authors:

Brandon T. Fisher, MD, Sanford Orthopedics and Sports Medicine, Sioux Falls, South Dakota; Orthopedic Residency Program, University of North Dakota School of Medicine and Health Sciences, Fargo, North Dakota; Sanford USD Medical Center, Sioux Falls, South Dakota.

Kelly McKnight, MD, Sanford Orthopedics and Sports Medicine, Sioux Falls, South Dakota; Orthopedic Residency Program, University of North Dakota School of Medicine and Health Sciences, Fargo, North Dakota; Sanford USD Medical Center, Sioux Falls, South Dakota.

Benjamin Jorgensen, MD, Sanford USD Medical Center, Sioux Falls, South Dakota.

Robert E. Van Demark Jr., MD, Sanford Orthopedics and Sports Medicine, Sioux Falls, South Dakota; Orthopedic Residency Program, University of North Dakota School of Medicine and Health Sciences, Fargo, North Dakota; Sanford USD Medical Center, Sioux Falls, South Dakota.



Premature baby beats the odds at Level IV NICU

In September 2020, Kristi and Ty Sabo learned to take things day by day – because, for nearly four months, they didn't have any other option. Their daughter, Lennon, was born prematurely at just 28 weeks.

"Being born this early, she didn't have time to develop all her organs and her anatomy, and everything in general. Those were a lot of the issues that we were dealing with. And, with being so small, she was catching infections left and right," Kristi Sabo said.

Lennon was born in Rapid City, South Dakota, where the Sabos call home. After eight days, Lennon and Kristi flew to the Sanford Children's Hospital in Sioux Falls, which is home to the Sanford Boekelheide Neonatal Intensive Care Unit (NICU), a Level IV NICU.

Then, Lennon received a life-threatening diagnosis: hydrops, a condition where fluid accumulates in different tissues and organs in an infant's body, resulting in dangerous swelling. After providers drained one pound of hydrops fluid from her abdomen, she weighed less than two pounds.

Suzy Reuter, MD, a neonatologist at Sanford Health in Sioux Falls, said she and other providers were concerned Lennon wouldn't make it.

"Hydrops can reach up to 90% mortality. We were absolutely concerned that was a real outcome," she said.

A team dedicated to Lennon's treatment

An entire team of specialists was working with Lennon during her NICU stay.

"It's hard to fully quantitate how many departments within Sanford helped Lennon," Dr. Rueter said. "Pediatric urology, pediatric nephrology, pediatric cardiology, pediatric infectious disease and a pediatric radiologist reading all of her imaging were all involved. Within the NICU, we have occupational therapists, physical therapists and case managers. All of these people were integral to the care of this complex baby."

It gave Sabo comfort knowing there were so many experts on her side.

"When I need to message her doctors, it is really interesting because I have to scroll through about 20 different people," she said.

A team dedicated to Lennon's treatment

The Sanford Boekelheide Neonatal Intensive Care Unit is the region's only Level IV NICU. This certifies that the hospital can provide the hightest level of care for sick and premature infants—even the most complex and critically ill newborns like Lennon.

Call (605) 333-4455 to refer a patient to Sanford Health.

>>> Lennon goes home

Sabo said Lennon continued to improve and get stronger. Then finally, after 118 days in the Sanford Children's Hospital, Lennon got to go home.

"We always said that we wanted to see Lennon run up and down our hallways, and after everything we'd been through, we just never thought it was going to happen. When it finally happened, it was one of those things where we asked, 'is this real life?'

"We looked at each other and said, 'this is our life now.' This is happening. We've made it this far. She's a miracle," Sabo said.

Lennon is currently a happy and healthy two-year-old girl with a full life ahead of her. The Sabos, and their entire expert team at Sanford Health, couldn't be happier.

SANFORD Children's

808-561-905 10/22

Presentation and Management of a Traumatic Right Sided Diaphragmatic Hernia: A Case Report

Austin Benson, MD; Luke Hurley, MD; and Michael Bauer, MD

Abstract

Traumatic diaphragmatic injury (TDI) is a rare and dangerous sequela of trauma. Right sided TDI is even more uncommon due to the usual protection of the diaphragm by the liver. TDI can present in a delayed fashion, making diagnosis difficult to obtain. In any case TDI needs to be taken very seriously as it can lead to bowel strangulation and require emergency surgery. Multiple approaches have been described in order to definitively repair diaphragmatic defects. This report describes a patient who developed a delayed onset right sided diaphragmatic hernia after experiencing blunt trauma.

Introduction

Traumatic diaphragmatic injury (TDI) is a relatively uncommon sequelae of trauma. The incidence of TDI has been reported to be between 0.8-5 percent following thoraco-abdominal trauma.[1] The method of injury can contribute to injury incidence, as TDI in the setting of a penetrating trauma has been more commonly reported on multiple occasions compared to blunt traumas.[1-3] Although less common, blunt traumas that result in a TDI have been shown to have a significantly higher mortality rate.[4] Furthermore, when comparing the location of a TDI, the incidence of a left TDI is far more common compared to right TDI.[4] Right TDI has been reported to account for only 5-19 percent of all TDI.[5] The discrepancy between right and left TDI has been attributed to the presence of the liver, which is less likely to herniate into the chest.[4] Additionally, it is possible for a TDI to be delayed in presentation. Delayed TDI is thought to occur secondary to the herniation of abdominal contents into the thorax.[6] These diaphragmatic hernias have been shown to present late in up to 30 percent of TDI cases.[7] Given the potential for delayed TDI, it is essential that a high index of suspicion is present in the setting of a high energy trauma.

Case Report

A 65-year-old man originally presented to an outside hospital in April 2020 after experiencing a fall from 3-4 feet off the back of a trailer. He fell on his right side and did not experience head trauma. CT of the chest, abdomen, and pelvis revealed fractured ribs at levels three through 10 on the right side, as well as a small right hemothorax. The patient was subsequently admitted for pain management. On day six of hospitalization, the patient began to experience severe lower abdominal pain and noted increasing abdominal distention. Subsequent CT of the abdomen and pelvis demonstrated further displacement of the inferior rib fractures with an enlarging right hemothorax and pleural effusion. There was also a newly developed hematoma in the soft tissues overlying the right hip that was concerning for possible hemorrhage. The patient was subsequently transferred to our facility for further workup and management.

Upon arrival, the patient underwent video-assisted thoracoscopic surgery (VATS) with hemothorax removal and placement of a chest tube. Following the procedure, the patient reported decreased pain and improved shortness of breath. He remained under inpatient care for the following week for aggressive pulmonary toilet and chest tube output monitoring before being discharged.

Despite the inpatient management, the patient continued to have ongoing shortness of breath and right sided chest pain over the next several months. He returned to the clinic four months later after experiencing minimal improvement. At this time, the patient was offered a referral to pain management for possible regional nerve block or ablation, but this was declined by the patient. A chest X-ray once again showed displaced rib fractures and persistent hemidiaphragm elevation (Figure 1). To further evaluate, a CT chest was ordered. The results showed multiple right sided rib fractures of the third through tenth ribs in various stages of healing. Displaced rib fractures included right sixth through ninth which were fractured in multiple places,

Figure 1. AP chest at four months following the initial trauma demonstrating concern to diaphragmatic hernia and displaced rib fractures. Black arrows point to the displaced right sided rib fractures. Blue arrow points to the elevated right hemidiaphragm.

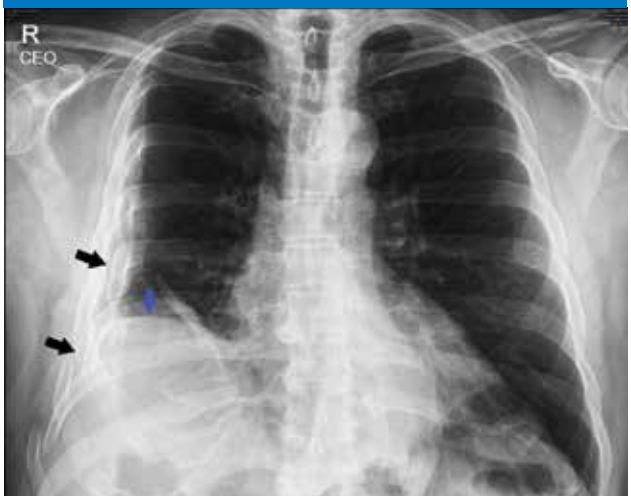
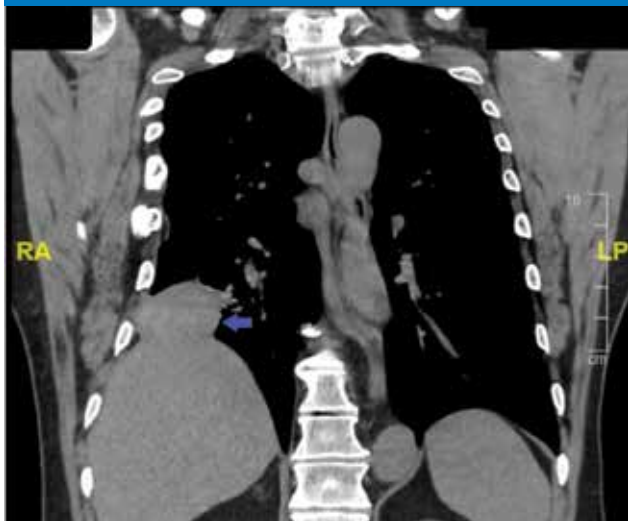


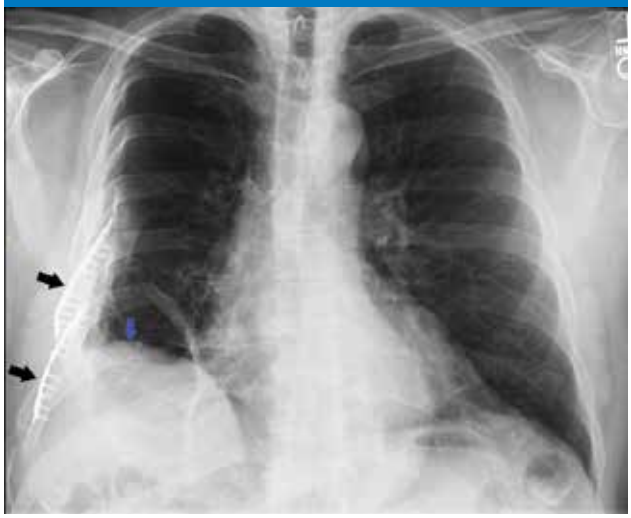
Figure 2. CT chest slice shows the portion of the liver herniating through the diaphragm. Blue arrow points to the portion of the liver herniating through the diaphragm into the chest cavity.



similar to prior examinations. Findings also revealed there was a new diaphragmatic defect on the right with a portion of herniated liver into the chest. The defect measured 4.2 cm. After reviewing the imaging, the patient elected to trial conservative management in an effort to resolve the pain and shortness of breath without further interventions. Three months later, the patient returned to the office after seeing minimal clinical improvement. Repeat CT scan of the chest revealed similar findings to his scan three months prior (Figure 2). At this visit, surgical management was presented as a treatment option, which the patient elected to pursue after a thorough discussion about the potential risks and benefits. In order to provide adequate relief for the patient's pain and shortness of breath, it was determined he should undergo a thoracotomy with diaphragmatic hernia repair and rib fixation.

Seven months after the initial trauma, the patient underwent the thoracotomy with diaphragmatic hernia repair with rib fixation. An incision was made over the lateral right chest that allowed exposure of the most displaced rib fractures and hernia defect. There was a clear diaphragmatic hernia with an approximately 6-7 cm diameter defect and a portion of the liver protruding into the chest cavity. The herniated portion was mobilized and reduced, and the defect was closed using a 2-0 Prolene suture. Attention was then turned to the fixation of the fractured ribs. The sixth and seventh ribs had the most displacement and these were realigned utilizing rib plates and self-tapping screws. Following the procedure, the patient remained inpatient for approximately one week for pain management and chest tube output monitoring.

Figure 3. Post op AP chest demonstrating resolution of the diaphragmatic hernia and reduction and fixation of the displaced rib fractures. Black arrows point to the plate fixation of the fractured ribs. Blue arrow demonstrates the improvement of the elevated right hemidiaphragm.



One month following the procedure, X-ray showed multiple partially healed rib fractures and a repaired diaphragmatic hernia (Figure 3). The patient reported improved shortness of breath and decreased pain levels, although he reported persistent rib pain located outside the operative field area. He underwent a myofascial injection of this area, located inferior to his incision. Overall, the patient reported his pain to be well-controlled, with remaining tenderness to palpation

in mid-back and inferior to his incision. He has also had improvement of his shortness of breath.

Discussion

Although uncommon, TDI is a dangerous potential sequelae following blunt thoracoabdominal trauma. The combination of delayed presentation and associated injuries can make a treatment plan difficult to generate. Conservative measures were initially utilized for this patient to allow the rib fractures to heal. Additionally, there was no apparent TDI at the time of initial evaluation. Even after the TDI was identified, the defect was thought to be small enough with no evidence of bowel herniating through and no obvious strangulation, allowing a watch-and-wait approach to still be utilized. The concern with TDI is the potential for progression of the defect, allowing more abdominal contents to herniate into the thoracic cavity; thus, increasing the risk for strangulation and emergency surgery. Due to concern for this progression, as well as the displaced non healing rib fractures, it was decided to proceed with surgery for definitive management.

Multiple approaches can be utilized in the surgical management of TDI including a minimally invasive approach through the abdomen or chest as well as open procedures. Each approach has its own advantages and disadvantages that should be considered depending on the defect and associated injuries.[8] Our patient was having continued shortness of breath, pain, and displaced rib fractures several months after the initial trauma. Due to these factors, it was thought a thoracotomy would allow for the repair of the diaphragmatic hernia as well as rib fixation.

Intraoperatively, the size of the liver herniating into the thoracic cavity was larger than what had been seen on previous imaging, further confirming the need for surgery. The use of mesh was considered to help with closure of the diaphragm but was ultimately not utilized as the defect was closed primarily without excessive tension on the suture.

Postoperatively, the patient noted improvement of his shortness of breath and pain, although he had not fully returned to pre-trauma baseline. The diaphragmatic hernia repair appears to remain intact. It was discussed with the patient once again that the rib pain may be chronic in nature due to the initial trauma. He has begun following with pain medicine to pursue additional treatment options for further pain management.

Delayed right sided TDIs are extremely rare and can be difficult to diagnose and treat. The choice to treat conservatively versus surgically depends on a variety of factors, but the risk for bowel strangulation should always be considered. A variety of surgical methods have been described, and the final treatment plan should be based on the specific needs of the patient.

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About the Authors:

Austin Benson, MD, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Luke Hurley, MD, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Michael Bauer, MD, Surgical Institute, Sioux Falls, South Dakota; Associate Professor, Department of Surgery, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

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Mesenteric Solitary Fibrous Tumor with High Risk for Aggressive Behavior: A Case Report

Anja Cucak, MS IV; Kaia Erickson-Adams, MD; and Matthew Sorrell, MD

Abstract

A 64-year-old female presented to the emergency department (ED) with complaints of two days of intermittent fever and chills, progressively worsening back pain, and hematochezia. Initial evaluation and computer tomography (CT) imaging work up revealed a hypervascular and necrotic appearing pelvic mass, measuring 11.7 x 7.8 x 9.7 cm, closely associated with the inferior mesenteric vein (IMV) in conjunction with portal venous gas. Flexible sigmoidoscopy with biopsy was performed to identify the etiology of the lesion and revealed an ulcerated non-obstructing mass in the recto-sigmoid colon measuring 3 cm in length and involving 1/3 of lumen circumference with oozing present. Interventional radiology (IR) embolization of the feeding vessels was done pre-operatively due to high vascularity of the mass. Pathology of the mass was consistent with a malignant solitary fibrous tumor.

Introduction

Solitary fibrous tumors (SFTs) were first described in 1931 by Klemperer and Rabin as a type of pleural spindle-cell neoplasm, which they named a hemangiopericytoma (HPC).[1] A rare type of tumor, arising from fibroblastic mesenchymal cells in soft tissue, was classified on a spectrum of solitary fibrous tumors/hemangiopericytomas. These tumors are most likely to arise in the thoracic cavity, affecting structures such as the lung pleura, parenchyma, mediastinum, and diaphragm; however, they can occur at virtually any anatomical site.[2] The name hemangiopericytoma was given due to their histological appearance, with 'stag-horn' thick-walled vessels composed of smooth muscle pericytes, separated by fibrous stroma,[1] but it was later realized these features were not specific to HPCs. Over the years, categorization of soft tissue tumors has advanced to be more precise and an identifying feature for SFT classification is presence of a NAB2-STAT6 gene fusion product on immunohistology.[3-5]

Like other soft tissue tumors, SFTs are often asymptomatic and can be found incidentally on imaging. Symptoms usually arise due to mass effect on the surrounding organs. These tumors occur equally in men and women in a wide range of 20-70 years of age.[2-7] Rarely, a paraneoplastic syndrome described as a non-islet cell hypoglycemia can arise with SFTs due to production of insulin-like growth factor-II (IGF-II).[5] Tumor size is variable and often depends on location. Typical classification criteria are used to identify malignant behavior of tumors, such as mitotic activity, cellularity, nuclear pleomorphism, and presence of necrosis.[5-6,8-9] Computer tomography (CT) scan is particularly useful in diagnosis, usually demonstrating a well-circumscribed, hypervascular mass with areas of necrosis.[2, 5-8] This case highlights a rare occurrence of a highly aggressive, extrapulmonary SFT of the mesentery that grew to be 15cm in size before symptoms presented.

Case Presentation

A 64-year-old female with past medical history significant for right breast ductal carcinoma in situ status post lumpectomy, radiation, and anti-hormonal therapy (2015), melanoma in situ, laparotomy for repair of perforating vessel injured during hysteroscopy (1998), hysterectomy with bilateral salpingo-oophorectomy and cystoscopy for treatment of abnormal uterine bleeding and fibroid uterus (2009), laparoscopic cholecystectomy (2014), hypertension, and hyperlipidemia, presented to the ED with complains of intermittent fever, chills, worsening back pain, abdominal pain, and hematochezia. The day prior she had no symptoms and denied any recent weight loss, chest pain, shortness of breath or changes to her bowel or bladder function. Her last colonoscopy in 2017 was unremarkable, except for presence of sigmoid diverticulosis. Vital signs were stable upon presentation to ED and physical exam was significant for lower left abdominal tenderness and positive digital rectal exam showing dark blood.

Labs at presentation showed leukopenia, neutropenia, and an elevated lactic acid. CT imaging revealed a 1.4 cm left hepatic lobe lesion, a 11.7 x 7.8 x 9.7 cm hypervascular

and necrotic pelvic mass (Figure 1) closely associated with the inferior mesenteric vein (IMV), and venous portal gas. Given the appearance of the pelvic mass, presence of portal venous gas, and elevated lactic acid, she was started on broad-spectrum antibiotics (vancomycin, metronidazole, and ceftriaxone) and intravenous (IV) fluids for treatment of neutropenic fever. Further workup included a flexible sigmoidoscopy with biopsy to identify etiology of the pelvic mass. Findings revealed ulcerated non-obstructing mass in the recto-sigmoid colon measuring 3 cm in length involving 1/3 of lumen circumference with oozing present. IR guided angio-embolization of the mass was performed prior to exploratory laparotomy with lysis of adhesions, en bloc resection of sigmoid colon and large mesenteric mass, stapled anastomosis, mobilization of the splenic flexure, and placement of a diverting loop ileostomy with transversus abdominis plane (TAP) nerve block. A loop ileostomy was performed due to a positive leak test secondary to a hole in the posterior mid portion of the anastomosis that was successfully repaired. The ileostomy was later reversed without complications. Her postoperative course was unremarkable, and she was discharged home on postoperative day seven with oral antibiotics for seven days due to positive fusobacterium blood culture. At the time of publication, patient continues to follow with oncology as well as is currently under active surveillance requiring abdominal and pelvic CT scans every three months during the first year. There has been no evidence of recurrence or metastasis 8 months post-surgery.

Pathology revealed a 15.0 x 12.5 x 9.5 cm, well-circumscribed mesenteric mass that ulcerated and involved the sigmoid colon. Sectioning of the mass demonstrated an 80 percent solid and 20 percent cystic architecture that had a variegated tan-pink to tan-white and fleshy cut surface with the cystic spaces releasing red-brown and hemorrhagic fluid. Microscopic examination revealed a spindled cell tumor with large pleomorphic cells (Figure 2), prominent staghorn vasculature (Figure 3), necrosis (Figure 4), and at least 1

Figure 1. Computer tomography (CT) scan showing mesenteric mass.

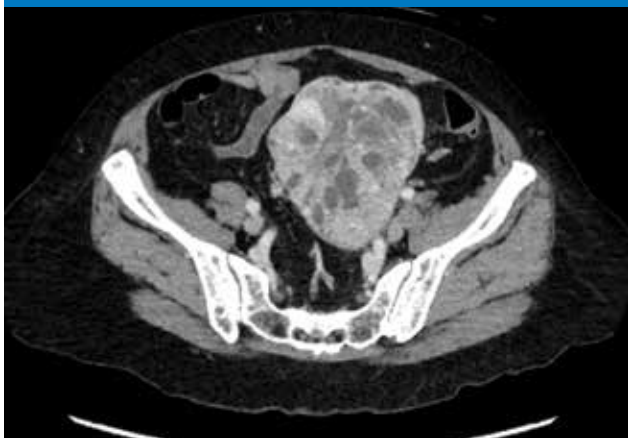


Figure 2. Spindled cell lesion with large pleomorphic cells (→), 20X (H&E).

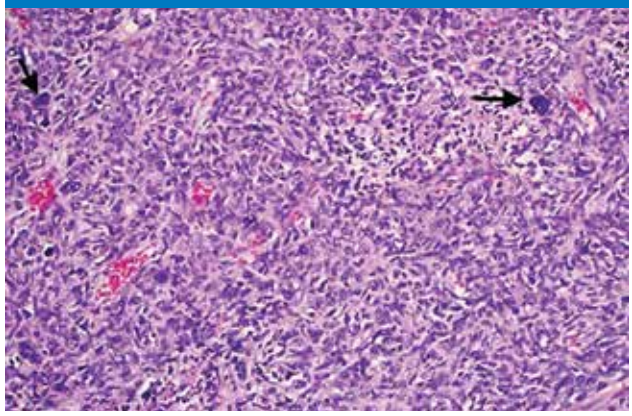


Figure 3. Prominent staghorn vasculature (←), 4X (H&E).

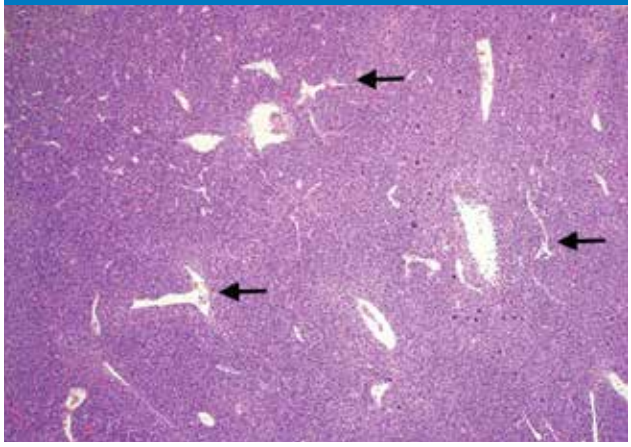
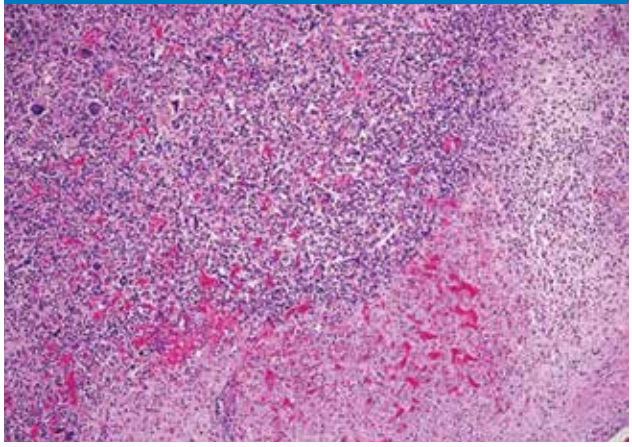


Figure 4. Lesion with necrosis (lower right/bottom), 10X (H&E).



mitotic figure per 10 high power fields. Immunochemical stains including STAT6, vimentin, S100, smooth muscle actin, Oscar, CD117, Dog–1, CD34, and CD10 were performed. The neoplastic cells were positive for STAT6 and vimentin and focally positive for CD34. The remaining stains were negative. The diagnosis of solitary fibrous tumor (SFT) was given. The presence of necrosis, increased mitoses, and large tumor size are features described in SFTs that exhibit malignant behavior.[10-11] Based on a modified four-variable risk stratification model for the development of metastasis in solitary fibrous tumor,[12] the tumor received the following score for each variable: 1 of 1 points for age (more than 55 years), 3 of 3 points for tumor size (15 cm or larger), 1 of 2 points for mitotic activity (1-3 mitotic figures per 10 high power fields), and 1 of 1 point for necrosis (involving 10 percent or more of the tumor). Therefore, this places the tumor (with a total score of 6 of 7) within a high-risk category for development of metastasis and creates a diagnosis of malignant solitary fibrous tumor.

Discussion

Solitary fibrous tumors (SFTs) are rare neoplasms that most commonly affect serosal surfaces of pleura and lung but are increasingly being identified in extrathoracic structures such as soft tissue in the thigh, pelvic fossa, retroperitoneum, as well as in the central nervous system (CNS). These tumors typically present as a single painless, enlarging mass that is sharply demarcated and homogenous in composition.[2,5-9] Often, SFTs follow a benign course and are asymptomatic at presentation in extrathoracic sites until their size reaches one that affect surrounding organs, creating a mass effect.[1-2,6-8] Rarely, hypoglycemia may occur due to a paraneoplastic syndrome secondary to increased insulin-like growth factor-II (IGF-II) expression.[5] Other symptoms reported include arthralgias and clubbing.

Due to their discrete nature, these tumors frequently are first identified incidentally on X-ray or CT imaging.[2,9] After identification of the mass, morphology, immunohistochemistry, and structural analysis is used to confirm the diagnosis of an SFT.[8] These multimodal diagnostics are crucial in identifying and predicting clinical behavior as there are many variants of SFTs and these tumors have been known to simulate other round cell, epithelioid, giant cell, adipocyte or myxoid tumors. Histologically, most tumors look benign, with spindle nuclei and ovoid-shaped growth patterns with minimal cytoplasm and surrounding fibrocollagenous tissue. Cases often display 'stag-horn' blood vessels, due to hemangiopericytic growth patterns, as well as perivascular sclerosis. Specifically, the presence of the NGFI-A-binding protein 2 (NAB2)-STAT6, which are both present on chromosomal region 12q13, fusion gene product support classification of an SFT.[2,4,13] While NAB2-STAT6 was not specifically tested for in this case, other histological features served as adequate support for the classification of SFT.

As mentioned previously, SFTs often present on serosal surfaces or soft tissue. Sigmoid colon presentation is extremely rare and very few reports in English literature have been documented. Cases concerning malignant SFTs may or may not present with invasion of surrounding tissue, specific criteria for which have been previously outlined in this report, and often go unidentified until growth reaches a large size. Regardless of classification, thus far, treatment has often best been achieved by full surgical resection with adequate negative margins.[8,14-16] Treatments using radiation therapy[17] and chemotherapy[18] with antiangiogenics have been limited due to the relatively low incidence of SFTs. Some small-scale retrospective studies[4] have shown promise in these therapies as adjuvant to surgical resection but with variable results. Currently, the accepted recommendation is to incorporate a multidisciplinary, case-by-case consideration when choosing treatment strategy of malignant SFTs.

Conclusion

This report features a rare presentation of a malignant solitary fibrous tumor of the mesentery involving the sigmoid colon. Our patient had an extensive past medical history that included prior malignancy and yet presented acutely to the emergency department with abdominal pain and hematochezia. Her CT scan revealed a large hypervascular pelvic mass that was further explored with flexible sigmoidoscopy, which then lead to a diagnostic and exploratory laparotomy with therapeutic en bloc resection of the sigmoid colon and tumor. This case further confirms the insidious nature of SFTs and how until they exert effects on surrounding tissues may be asymptomatic, therefore difficult to identify. Luckily, after identification and surgical resection, the prognosis for even malignant SFTs is favorable.

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Please note: Due to limited space, we are unable to list all references. You may contact South Dakota Medicine at ereiss@sdsma.org for a complete listing.

About the Authors:

Anja Cucak, MS IV, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Kaia Erickson-Adams, MD, Pathology Residency Program, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Matthew Sorrell, MD, Sanford Health Surgical Associates, Sioux Falls, South Dakota; Clinical Professor, Department of Surgery, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

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Netherton Syndrome in a Mother and Her Two Children

Jack DeMoss, MD; Leslie Cooper, MLS(ASCP)CMSBB(ASCP)CM; Colby Felts, MD; and Gregory Wittenberg, MD

Abstract

Netherton syndrome (NS) is a rare autosomal recessive condition caused by mutations in the serine peptidase inhibitor, Kazal type 5 (*SPINK5*) gene which encodes for a serine protease inhibitor, lymphoepithelial Kazal-type-related inhibitor (LEKT1). NS is characterized by a triad of ichthyosiform erythroderma, trichorrhexis invaginata, and atopic diathesis with elevated IgE levels. The syndrome typically presents in infancy, where life-threatening complications are frequent, and evolves into a less severe condition with milder clinical symptoms in adulthood. This case report details the clinical history and genetic testing of a mother and two children with clinically symptomatic and genetically proven NS.

Introduction

Netherton syndrome (NS) is a rare autosomal recessive condition characterized by a triad of ichthyosiform erythroderma, trichorrhexis invaginata, and atopic diathesis with elevated IgE levels. It is caused by a mutation in the Kazal type 5 (*SPINK5*) gene leading to a truncated lymphoepithelial Kazal-type-related inhibitor (LEKT1).[1] The syndrome initially presents in infancy with diffuse erythroderma. In severe cases, NS can have life threatening complications including hypernatremic dehydration and failure to thrive.[2] The syndrome transitions in adolescents and adults, primarily presenting with a swirling pattern of scaling called ichthyosis linearis cirumflexa. At any age, patients with NS can have trichorrhexis invaginata where their hair is fragile and easy to break. Finally, they have a predisposition to allergies, atopic dermatitis, and asthma, termed atopic diathesis, along with elevated IgE levels. Although a rare disease, it is thought the incidence of NS is under reported due to the difficulty in diagnosis.[3] This report describes a woman with a suspected history of NS who has two children presenting with NS-like symptoms which was subsequently genetically confirmed in all three individuals. This highlights the importance of genetic screening to assist with this difficult and under reported diagnosis.

Case Report

A 33-year-old gravida 1, para 0 woman at 30 weeks gestation with a history of clinically diagnosed NS presented to clinic with debilitating disease. Genetic testing had not been completed and she reported no erythema, xerosis, or severe allergies in childhood. Throughout early adult life to present, she reported intermittent ichthyosis, alopecia, and worsening xerosis. Her chief complaint on presentation was ichthyosis of her face and body. Physical exam revealed near complete alopecia with only sparse, fine hairs present. Additionally, there was obvious erythema of the face. Her ichthyosis and xerosis were well controlled using silver sulfadiazine cream, triamcinolone, and Domeboro soaks.

As her pregnancy progressed, her symptoms were stable until the third trimester, when she developed a sudden, mildly pruritic eruption on her bilateral anterior thighs. The confluent, sharply demarcated, erythematous plaques covered the entire surface of the anterior thighs. A potassium hydroxide (KOH) preparation was negative. No photographic documentation or biopsy was completed at the time, but likely differentials included contact dermatitis or a manifestation of NS, specifically ichthyosis linearis circumflexa.

Following the birth of her first child, genetic testing was more strongly considered given her child's subsequent dermatologic symptomatology. Her 6-week-old male child presented to clinic with mild to moderate diffuse erythema, most prominent on the anterior scalp. The scalp and upper chest also had scattered areas of light scaling. He was diagnosed with both seborrheic dermatitis and atopic dermatitis and subsequently prescribed pimecrolimus. At this time, discussion with his mother regarding genetic testing for the *SPINK5* gene occurred with plans to test the child for the mutation.

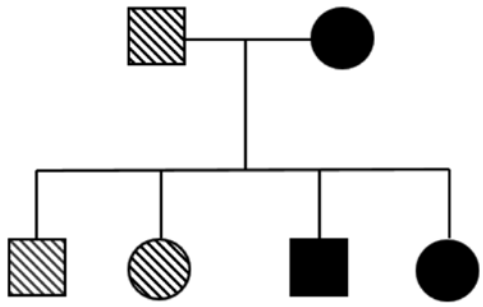
Following discussion with a geneticist, the new recommendation was to delay testing of the child, and instead obtaining serum IgE levels and radioallergosorbent (RAST) testing of the mother. If these results were elevated

and diffusely positive, respectively, the next step would be *SPINK5* testing of the mother followed by her child. Her IgE level was 2,106 kU/L (reference range 2-214) and RAST testing revealed allergy to peanuts, corn, walnuts, shrimp, various other foods, and environmental pollens. Laboratory results in combination with her clinical presentation and history justified *SPINK5* gene testing.

Genetic test results were consistent with compound heterozygous NS with a single nucleotide deletion (417delC) in exon 6 and nucleotide transition (1048C/T) in exon 12. Both mutations can be predicted to result in synthesis of a severely truncated or absent LEKT1 polypeptide, thus confirming the diagnosis of NS. This result prompted the genetic testing of her son, whose results also confirmed the diagnosis of NS. The records detailing his specific mutation were not retrieved.

In 2006, the now 35-year-old patient had her second child, a female. There were no exacerbations of her dermatologic symptoms during this pregnancy, compared to her first pregnancy. Starting at one week old, the female child had mildly erythematous, eczematous patches involving the scalp and upper back. The symptoms were reportedly less severe than her older brother's symptoms, but given the proven genetic history, genetic testing was performed which confirmed NS. It is reported that the father underwent genetic testing which showed heterozygous changes for a *SPINK5* gene mutation, but there is no documentation of the specifics. The family was counseled on the probability of future children having NS and informed any children would at least be a carrier. They were also informed about options such as preimplant genetic testing and diagnosis.

Figure 1. Example of autosomal recessive inheritance where one parent is affected, and one parent is a carrier. 50% of their children will be carriers, and 50% of their children will be affected.



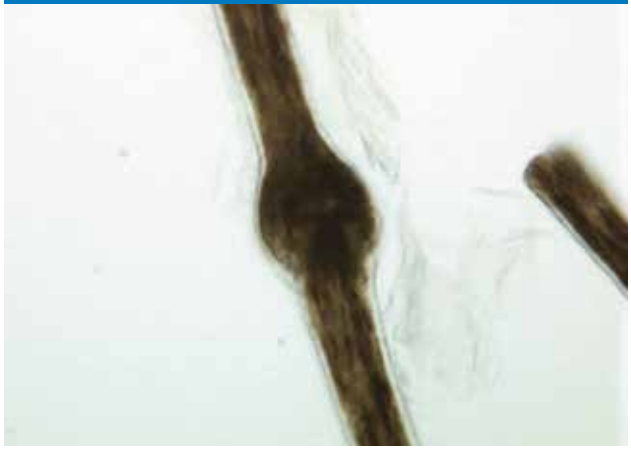
Based on the confirmed diagnosis of NS in the mother, and two children, Figure 1 illustrates the most likely inheritance pattern for this case study. In this figure, the mother has NS and the father is a carrier. This results in a 50 percent chance of any individual child having NS, and 50 percent chance of any individual child being a carrier of a NS gene mutation.

Discussion

This case report is the first to follow a woman with suspected NS who subsequently has two children also with suspected NS followed by genetic testing of the entire family. Despite NS having an estimated incidence of 1 in 200,000, less than 200 cases have been reported. It is thought the incidence may be underestimated and has a true rate closer to 1 in 50,000 births.[3] This underestimation may be due to the difficulty of diagnosis and low occurrence of genetic testing. The importance of confirmatory genetic testing of suspected NS to make informed decisions about family planning is highlighted in this case report.

NS initially presents in infancy with diffuse erythroderma and, less commonly, a diffuse collodion membrane.[4] Infants and young children may be born prematurely and can be more susceptible to recurrent skin infections. Clinicians should carefully monitor for life-threatening complications from NS which include failure to thrive and hypernatremia dehydration due to excessive loss of fluid through the inappropriate skin barrier. In adolescents and adults, the syndrome transitions to a primary presentation of ichthyosis linearis cirumflexa. At any age, patients with NS can have trichorrhexis invaginata. Finally, they will have a predisposition to allergies, atopic dermatitis, and asthma, termed atopic diathesis, along with elevated IgE levels. The

Figure 2. Light microscopy of hair shaft demonstrating trichorrhexis invaginate.[5]



differential diagnosis of NS most commonly includes diffuse atopic dermatitis, congenital ichthyosiform erythroderma, and X-linked ichthyosis.

Establishing the diagnosis of NS varies on patient age, but always starts with clinical suspicion. In adults, the triad of ichthyosiform erythroderma, trichorrhexis invaginata, and atopic diathesis seen over time should be followed by light microscopy of hair follicles. In infancy, clinicians should monitor patients for not only skin manifestations, but also the life-threatening manifestations of hypernatremic dehydration and failure to thrive.

Diagnosis of NS is confirmed by genetic testing patients with clinical manifestations for mutations in the *SPINK5* gene, which encodes for the serine protease inhibitor LEKT1. To date, approximately 80 different mutations of the *SPINK5* gene have been associated with NS. These mutations are located in both exonic and intronic regions and include 18 nonsense mutations, two non-synonymous mutations, 34 insertion and/or deletion mutations, and 24 mutations which disrupt the normal splicing pattern.[6]

One of the complexities with NS is the significant differences between mild and severe clinical presentations of patients. These variations are due to the direct effect of the patient's genotype on their phenotype. Clinical phenotypes among patients with NS appear to have a potential correlation between the mutation in the *SPINK5* gene, and where the mutation impacts the LEKT1 molecule domains.[1] Mutations located upstream in the LEKT1 molecule present with a more severe phenotype than mutations located downstream due to more domains being affected by early truncation of the protein.

Genetic testing for suspected NS can impact family planning, as well as neonatal care. Heterozygous carriers of a NS mutation lack clinical manifestations of the disease.[4] Identifying individuals with homozygous mutations via genetic testing could allow the family time to plan or seek genetic counseling for current or future pregnancies. Genetic testing in infants is also important to be adequately prepared for the more serious complications of NS in early life. Clinicians should also inform patients about preimplantation genetic testing and provide a referral if necessary.

The management of NS is complex, and symptoms often persist despite best management practices. The most critical aspects in management are oral antihistamines in addition to regular and copious use of emollients. It is important to note topical medications can be dangerous in patients with severe NS due to a compromised skin barrier allowing easier access to the bloodstream. In particular, topical tacrolimus should be used with great caution due to reported risk of systemic toxicity. Nevertheless, topical or systemic antibiotics as needed remain crucial in the treatment of recurrent skin infections and possible septicemia. Additional therapies may include topical steroids and dandruff shampoo for scalp scaling, oral retinoids for scaling, and topical tacrolimus. Finally, a 2011 case report detailed the use of Infliximab therapy to successfully manage the inflammatory components of NS. Infusions at a dose of 5 mg/kg were given in weeks 0, 2, and 6 and continued every other month. Inflammatory lesions were improved by the second infusion and entirely absent by the twelfth. Xerosis, ichthyosis, and IgE levels were unchanged, however.[7]

Conclusion

Netherton syndrome (NS) is a rare autosomal recessive condition with a complex presentation which can be mistaken for other dermatological conditions. Symptoms widely vary based on location of a SPINK5 gene mutation relative to location of the LEKT1 molecule. NS is likely underdiagnosed, and clinicians suspicious of a patient's symptoms should have a low threshold to order genetic testing. More liberal use of genetic testing for suspected NS will aid in genetic counseling and family planning.

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About the Authors:

Jack DeMoss, MD, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Leslie Cooper, MLS(ASCP)CMSBB(ASCP)CM; Avera Institute for Human Genetics, Sioux Falls, South Dakota.

Colby Felts, MD, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Gregory Wittenberg, MD, Clinical Professor, Department of Internal Medicine, University of South Dakota Sanford School of Medicine; Rapid City Medical Center, Rapid City, South Dakota.

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Esophageal Hemangioma: A Rare Cause of Chronic Gastrointestinal Blood Loss

Christian Tobin, MS IV; Christopher Van Hove, MD; Lauren Van Hove, MD; and Steve Gutnik, MD, FACP

Abstract

Esophageal hemangioma (EH) is a rare, benign tumor that is most often asymptomatic but may present insidiously with dysphagia and blood loss anemia. We report a case of a 70-year-old male with symptomatic anemia who underwent a full gastrointestinal evaluation and was found to have an EH. We review the classification of benign esophageal neoplasms and discuss the characteristics, imaging, interventions, and surveillance specific to EH.

Introduction

EH is a benign tumor of the esophagus with fewer than one hundred reported cases in the literature.[1,2] Analysis of 19,982 autopsies performed at the New York Medical College over a 10-year period revealed only three cases of EH.[1,3] The rarity of these tumors makes identifying demographic variables and risk factors challenging. The majority of reported cases occur in patients over the age of 40, with a slight increased incidence in men.[1,2] Hemangiomas occur throughout the length of the esophagus with a hypothesized slight preponderance for the upper 1/3.[4,5] On upper endoscopy, these tumors appear as soft, bluish-red lesions with a complex submucosal vasculature that predisposes them to bleeding from endoscopic trauma.[6] Endoscopic ultrasound with doppler is the best imaging modality to evaluate a suspected EH.[7] When symptoms necessitate intervention, several approaches have demonstrated effectiveness to excise or ablate these tumors. Our case highlights the insidious clinical presentation of a hemangioma of the esophagus as well as the consequent risk of hemorrhage from endoscopic investigation and excision.

Case Description

This is a 70-year-old male on rivaroxaban for paroxysmal atrial fibrillation who presented with an asymptomatic iron deficiency anemia. His hemoglobin was 10.5 g/dL without evidence of gross gastrointestinal bleeding. Workup with esophagogastroduodenoscopy (EGD), colonoscopy, CT enterography, and pill camera were unable to identify a clear cause of the anemia. The patient was started on oral iron and his hemoglobin improved to near normal levels.

Two years later, the patient presented with progressive fatigue and a hemoglobin of 9.5 g/dL. Once again, no gastrointestinal symptoms were noted. Colonoscopy was notable only for diverticulosis and repeat EGD revealed a 1.2 cm submucosal reddish colored lesion in the proximal third of the esophagus (Figure 1). The lesion was excised in its entirety by endoscopic biopsy utilizing a jumbo forceps. At the completion of the procedure, the base of the excised polyp bled freely. Endoscopic tamponade was utilized with the tip of the scope for 15 minutes. The lesion continued to bleed. Two hemoclips were placed at the site to achieve hemostasis. It should be noted that the patient held his anticoagulant for 3 days prior to the procedure. At discharge from the ambulatory surgical area, the patient was instructed to continue oral iron and resume rivaroxaban in 7 days.

Interestingly, the pathology revealed the lesion in the proximal esophagus to be a capillary hemangioma (Figure 2). At one month post-excision, the patient's hemoglobin had normalized. At eight months post-excision, an EGD showed a residual 4 mm hemangioma without evidence of recurrent bleeding. Given the patient's maintenance of normal hemoglobin and absence of concerning findings on upper endoscopy, no further therapeutic intervention was necessary. We will follow the patient's hemoglobin intermittingly and plan to perform a surveillance EGD at 2 years post-excision to visualize the growth pattern of the residual hemangioma.

Discussion

Benign esophageal tumors can be classified by appearance as raised, flat, or cystic (Table 1).[4,8] EH is classified as a raised (intraluminal), nonepithelial neoplasm that arises from hypertrophied blood vessels. The majority are cavernous (ie, have cavernous spaces seen histologically), although capillary lesions are also known to exist, as is the case in our patient.

Figure 1. Top – pre-excision: EGD demonstrating red-colored 1.2 cm, polypoid lesion 26 cm from the incisors and 5-6 cm below the upper esophageal sphincter. Bottom – post-excision: two hemoclips applied to base of excised hemangioma.

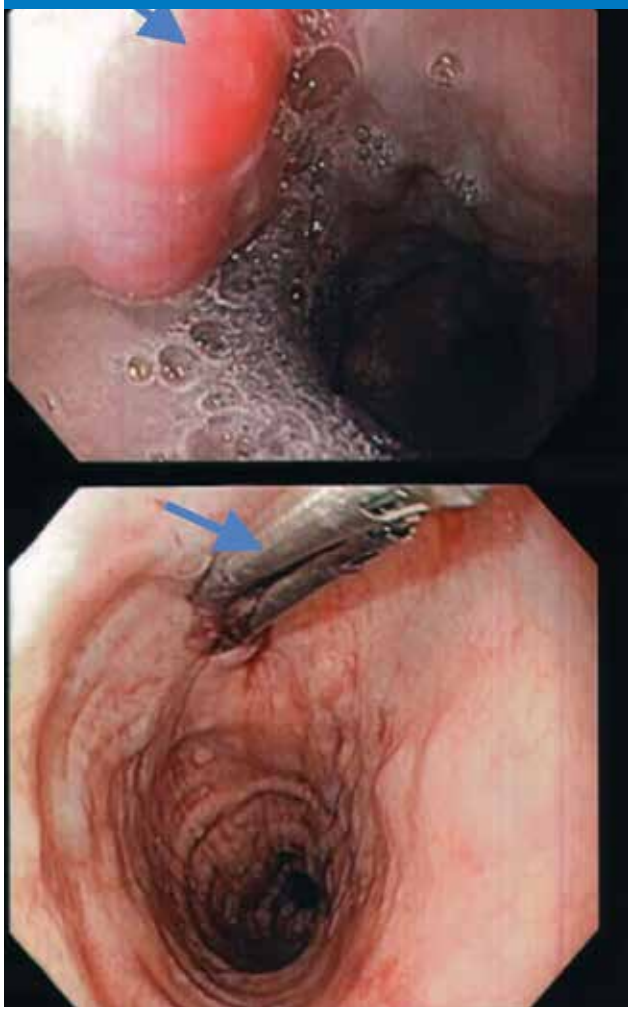
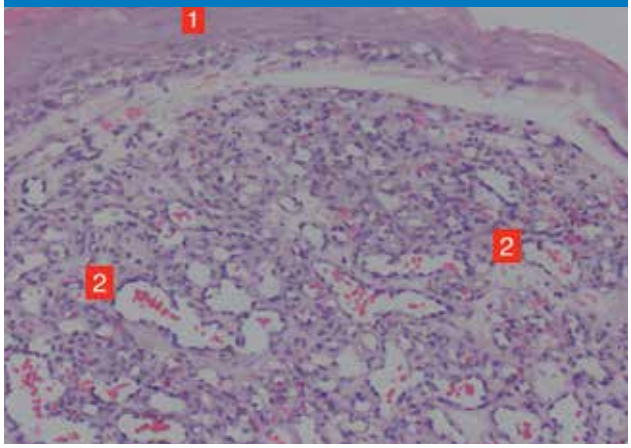


Figure 2. Capillary hemangioma at 100x. (1) Mucosa. (2) A solid growth of small, thin-walled capillaries arranged in a lobular pattern. These capillaries are lined by a single layer of endothelial cells.



An EH is most often singular but can present as multiple lesions in patients with Rendu-Osler-Weber syndrome, Klippel-Trénaunay syndrome, or congenital blue rubber bleb nevus syndrome.[6,7,9]

Hemangiomas are most often discovered incidentally in asymptomatic patients. They are discovered less often on screening endoscopy for obstruction (dysphagia) or occult blood loss anemia. In a small analysis of 29 cases of EH, Araki and colleagues reported the most common symptoms were dysphagia (45.2%), hematemesis (25.8%), melena (12.9%), and retrosternal pain (12.9%).[2] Spontaneous bleeding upon insult to the overlying mucosa is a major complication associated with an EH. Massive hemorrhage can occur from endoscopic trauma due to the complex submucosal vasculature of these neoplasms.[5]

Multiple imaging modalities can be utilized to evaluate esophageal lesions. Barium esophagography, CT, and MRI are relatively non-specific for EH as they show a well-defined, soft tissue mass in the esophageal wall.[5] Endoscopic ultrasound is the most sensitive and specific imaging study for EH because it can readily differentiate between benign and malignant

Table 1. Benign esophageal neoplasms classified as raised, flat, or cystic.[8]

Raised lesions
Leiomyomas and leiomyosarcomas
Gastrointestinal stromal tumors
Schwannomas
Lymphangiomas
Hemangiomas
Fibrovascular polyps
Fibromas, fibrolipomas, myomas, lipomas
Inflammatory fibroid polyps
Hamartomas, inflammatory pseudopolyps, eosinophilic granulomas
Granular cell tumors
Adenomas
Papillomas
Flat lesions
Heterotopic sebaceous glands
Glycogen acanthosis
Inlet patch
Parakeratosis
Esophagitis dissecans superficialis
Cystic lesions
Duplications and cysts
Bronchogenic and enterogenous cysts

lesions and assess pertinent characteristics such as location, size, and depth.[6,7] The addition of doppler ultrasonography can precisely demonstrate the submucosal vascular channels to help determine the feasibility of surgical excision.[5,7] EH must be differentiated from varices and Kaposi sarcoma, as they may appear similarly on endoscopy.[10] If present on CT, phleboliths (calcified venous clots) are considered a specific finding for hemangioma.[7] Visualization of hemangiomas with EGD will reveal a nodular, soft, bluish-red, polypoid submucosal lesion that may blanch when pressed with biopsy forceps.[6,11] The majority of the literature supports avoiding tissue biopsy due to risk of massive hemorrhage.[1,5,6,11]

Asymptomatic tumors are typically monitored without intervention, but as demonstrated by our patient, symptomatic anemia from presumptive esophageal bleeding warrants intervention. For small tumors such as ours, endoscopic excision is the most appropriate therapy of choice. In cases of large tumors unsuitable for endoscopic excision, video-assisted thoracic surgery (VATS) resection is preferred.[12] Several other novel techniques have also been proposed in the literature including sclerotherapy, radiation, laser fulguration, esophagectomy, and tumor enucleation.[1,6,13]

The literature does not report surveillance guidelines for cases of EH. From a malignancy standpoint, the risk of transformation is likely too low to warrant surveillance. However, the risk of bleeding with increasing tumor size does provide grounds for surveillance. In our case, it is likely the residual 4 mm hemangioma observed with EGD at 8 months post-excision will remain asymptomatic. However, if the lesion enlarges to greater than 1 cm or symptoms develop, it would be best to intervene with endoscopic methods. We plan to perform an additional surveillance EGD at two years post-excision to reevaluate the residual hemangioma. The infrequency and variability of these tumors makes a single treatment and surveillance recommendation difficult. We advocate for a multidisciplinary approach that considers tumor size, location, and clinical symptoms to devise the most appropriate interventional strategy with appropriate post-excisional surveillance.

In summary, we described a case of an EH in a 70-year-old male who presented with symptomatic iron deficiency anemia. EH is a rare, benign tumor that is most often asymptomatic but may cause obstruction and bleeding. The best imaging modality is endoscopic ultrasound with doppler to discern pertinent characteristics. Caution should be exercised when performing EGD with or without biopsy to prevent trauma and hemorrhage from the tumor. If excision is indicated, endoscopic or VATS resection are effective interventions.

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About the Authors:

Christian Tobin, MS IV, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Christopher Van Hove, MD, Internal Medicine Residency Program, Department of Graduate Medical Education, Abbott Northwestern Hospital, Minneapolis, Minnesota.

Lauren Van Hove, MD, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Steve Gutnik, MD, FACP, Clinical Professor of Medicine, Department of Gastroenterology, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Propofol Use in Pediatric Sedations: Will a Lesser Dose be Effective? A Quality Improvement Project

Mir H. Ali, MD; Michael Vlach, MS IV; Ethan Leif, MS IV; and Madisyn Braun, MS IV

Abstract

Background: Propofol provides sedation in the pediatric setting for MRIs, ensuring minimal movement from patients and high-quality pictures. Sanford Children's outpatient sedation clinic currently does not have a standard protocol for using propofol for sedation. The purpose of the project was to determine if we could decrease the dose of propofol while maintaining adequate sedation during MRI imaging.

Methods: The study had three phases of retrospective chart review. The first phase was a six-month review of propofol dosing. The second phase introduced a goal propofol drip dose of 200-300 mcg/kg/min with reviewing success of sedation for six months. Lastly, the third phase introduced a goal propofol drip dose of 175-200 mcg/kg/min with reviewing success of sedation for four months. A successful sedation was determined by completing the imaging study without the child awakening.

Results: A total of 181 patients ranging in ages 6 months to 16 years were recruited. Percentage of successful sedations in phase 2 and phase 3 were 83 percent and 84 percent, respectively. The average total propofol dose used in sedations decreased from 15.43 mg/kg in phase 1 to 12.31 mg/kg in phase 3. On average of the three phases, the mean arterial pressure (MAP) overall was below the normal range in 60 percent of sedations.

Conclusion: We conclude that creating a protocol with a baseline propofol drip rate of 175-199 mcg/kg/min for pediatric sedations would allow for successful sedations and prevent unnecessary excess dosing.

Introduction

Propofol is an IV anesthetic used in the setting of pediatric MRIs to maintain patient comfort and stillness via deep sedation to ensure completion of the procedure. Deep sedation ensures procedures are successfully completed while the pediatric patients remain comfortable without the risks of general anesthesia. Propofol is an IV anesthetic that exerts its effect via potentiation of the GABA-A receptor complex. Propofol is normally used to induce and maintain anesthesia. Also, it is often used for sedation in the ICU, rapid anesthesia induction, and deep sedation for short procedures. Higher doses of propofol use cause vasodilation, resulting in hypotension that routinely requires a saline bolus.[1] There is also data suggesting neurotoxicity associated with propofol use in pediatric patients.[2] If we can achieve the same anesthetic effect in pediatric patients with a lower dose of propofol, this will lead to a reduction in the risks of neurotoxicity, lower costs associated with saline boluses, and improve patient care.

Propofol is used as a sedative at Sanford Children's Hospital in Sioux Falls, South Dakota, to obtain successful magnetic resonance imaging (MRI) in children. Historically, the propofol drip dosing for pediatric MRI sedation at Sanford Children's Hospital is a 2 mg/kg bolus followed by a 200-400 mcg/kg/min drip, starting with 400 mcg/kg/min and titrating down as tolerated. This dosing is simply a range of values that the intensivists had agreed upon many years ago and which has been used since. Every patient also receives a bolus of normal saline due to expected hypotension with propofol use. We hypothesize that our institution may be using higher than needed doses of propofol. This quality improvement (QI) project aims to reduce total propofol dosing in MRI sedations by 5-10 percent. Results from this study are intended to establish current propofol dosing practices and will help guide new procedure protocols and future research to ensure minimization of sedative use. A lower standard of propofol dosing would lower healthcare costs. Additionally, while propofol is known to be a very safe medication, using decreased doses limits the possibility of

any dose dependent side effects.

Methods

This research was approved by both Sanford Health in Sioux Falls, South Dakota, and the University of South Dakota institutional review boards. Sanford accepted the work as non-human research according to the Protocol Template for Record Review (HRP-503a). The Plan Do Study Act model for healthcare quality improvement was followed.

The primary outcome is to lower the propofol drip dose by 5-10 percent while maintaining successful sedation. We performed a retrospective chart review in phase I of the study and created goal propofol drip rates in phases 2 and 3. Our primary outcome was measured using information collected from the sedating physician's procedure note in the EMR. A successful sedation was defined by using the goal propofol drip dose range for each phase of the study. If the intensivist titrated up the propofol drip dose outside the desired range to maintain adequate patient sedation, the sedation was deemed unsuccessful. Success of sedation was determined by reading the reported propofol drip rate from the templated anesthesia procedure note. The reported drip dose could also be cross-checked by reviewing the pediatric procedure flow sheet completed by nursing staff during the procedure. If the drip rate reported in the note exceeded the goal range, the sedation was recorded as unsuccessful.

Our secondary outcome aims to calculate mean arterial pressure (MAP) during sedation to determine if the routine saline bolus is necessary with lower doses of propofol. Blood pressure is recorded in the pediatric procedure flow sheet by nursing staff throughout the procedure. This data was used to calculate MAP and compared to the normal MAP for each age group reported by Pediatric Advanced Life Support (PALS).

Participants

The inclusion criteria to participate in the study were dependent on the population and the type of sedation used for specific pediatric procedures. First, the participants were pediatric patients aged 6 months to 16 years who underwent a procedure requiring sedation. All pediatric patients were seen at Sanford Children's Hospital, Sioux Falls, South Dakota. Secondly, the pediatric patient must have received a propofol drip during the procedure. The most common procedure for requiring a propofol drip was an MRI (magnetic resonance imaging). In addition to MRIs, other procedures like auditory brainstem response and renal scans require a propofol drip. Patients with a contraindication to the sedation method or those who had previously experienced an adverse reaction to sedation were excluded from the study. Lastly, patients who could undergo procedures like an MRI without sedation were excluded from the study. Sanford Hospital utilizes a propofol drip for certain procedures in up to 160 pediatric patients annually (internal data recorded over the last year) and there is no specific protocol for how much propofol to give. This study aimed to decrease the amount of propofol used in the propofol drip over the course of a procedure while still maintaining success of sedation.

Procedure

The study was divided into three phases of data collection. During phase 1, a retrospective chart review of patient medical records was used to collect data regarding the hospital's historical propofol dosing during MRI with sedation. Phase 1 data was collected for MRIs with sedation in the six-month period of September 2020-February 2021. The usual propofol drip dose was 400 mcg/kg/min. During phase 2 from March 2021-July 2021, a goal propofol drip dose of 200-300 mcg/kg/min was suggested to the pediatric intensivists. Finally in phase 3 from August 2021-November 2021, the suggested dose was 175-200 mcg/kg/min. The pediatric intensivist performing sedations was informed and consistently reminded of our goal propofol drip dose during each phase of the study. The sedation protocol was followed by physicians and nursing staff, including monitoring of vital signs in the electronic medical record during the MRI procedure. We elected to start at the highest dose for the phase and titrated it down to the lowest allowed dose before the procedure ended. If the goal propofol drip dose in each phase did not provide adequate sedation to the patient, the physician increased the dose as necessary to maintain MRI imaging satisfaction. In this case, the sedation would be termed unsuccessful. Data was collected by the student research team in all three phases via retrospective chart review of patient medical records. Data was stored anonymously without patient identifiers directly into our data collection sheet in RedCap, a secure, browser-based data collection system requiring authorized users. Accuracy of data collection was ensured by student researchers having a uniform template for data collection and rechecking outliers in the data. There was no missing data for patients who qualified to participate in our study based on inclusion criteria, as patients who underwent sedation always have the intensivist note available for review with appropriate propofol dosing, blood pressure, and demographic information. Data was analyzed using Microsoft Excel.

Results

Data was collected over a 15-month period from Sept.1, 2020, through Nov. 13, 2021, at Sanford Children's Hospital. The data collected contained no patient identifying information.

Over the collection period, a total of 181 patients with an age range of 6 months to 16 years old participated in the study using the exclusion criteria described above. The average age was 5.1 years. Phase 1, 2, and 3 had 73, 61, and 47 patients respectively. The percentage of successful sedations in each dose range is shown in Table 1. A successful sedation is defined by using the goal propofol drip dose range for each phase of the study. If the intensivist titrated up the propofol drip dose outside of the desired range to maintain adequate sedation for the patient, the sedation was deemed unsuccessful. However, the intensivists ensured the patients could complete the MRI satisfactorily by increasing the dose if needed. When a propofol drip of 200-400 mcg/kg/min, 201-300 mcg/kg/min, and 175-199 mcg/kg/min was administered, the sedation success rate was 100 percent, 83 percent, and 84 percent, respectively.

The average propofol dose was 15.15 mg/kg. When a propofol drip of 200-400 mcg/kg/min, 201-300 mcg/kg/min, and 175-199 mcg/kg/min was administered, the average propofol dose was 15.43 mg/kg, 15.55 mg/kg, and 12.31 mg/kg respectively. This results in a 20 percent decrease in total propofol dose between phase I and phase III of the study.

Table 1. Sedation completion by drip rate.

Propofol drip rate (mg/kg/min)	# of successful sedations	# of failed sedations	Percent successful
200-400	70	0	100%
201-300	50	10	83%
175-199	41	8	84%

Table 2. Average total propofol by drip rate.

Propofol drip rate (mcg/kg/min)	Average total propofol (mg/kg)
200-400	15.43
201-300	15.55
175-199	12.31

Table 3. Effect on MAP with varying propofol dose.

Phase	Total sedation	MAP within normal range	MAP below normal range	Percent below normal range
Phase 1	73	28	45	62%
Phase 2	61	23	38	75%
Phase 3	47	22	25	64%

Regarding our secondary outcome, patients receive a routine normal saline bolus prior to sedation. We aimed to determine the mean arterial pressure during sedation retrospectively for each patient to ascertain if a saline bolus is necessary with lower doses of propofol. The PALS determination of mean arterial pressure (MAP) was used as the goal normal range for MAP in each age group. MAP was attained after each sedation during chart review and was calculated based on the documented blood pressures throughout the sedation. In phases I, II, and III of the study, the percent of patients with a MAP below the normal range for the patient's age group was 62 percent, 75 percent, and 64 percent, respectively.

Discussion

This quality improvement project was intended to evaluate the success of pediatric sedation using lower doses of propofol to decrease risk of neurotoxicity. The results indicate that lower doses of propofol can be used for successful pediatric sedation. A sedation success rate of 84 percent at the 175-199 mcg/mg/min of propofol was higher than expected and exemplifies that patients can often tolerate lower doses of propofol without arousal. Based on the data, it is reasonable to create a protocol that defines 175-199 mcg/kg/min as the baseline propofol drip rate for pediatric sedation. The intensivists will start at 199 mcg/kg/min and titrate down to 175 mcg/kg/min as allowed to achieve adequate sedation and successfully complete the procedure. Regarding our secondary outcome, saline boluses are given routinely prior to each sedation. The amount given is based on the patient's weight. Lowering the propofol drip rate in phases 2 and 3 did not improve the MAP of sedated patients. In result, the data does not support discontinuation of standard IV saline boluses because the MAP was below the normal range in 60 percent of total sedations recorded.

Limitations

The primary limitations of this study are the small patient sample size. The small sample size makes the data difficult to generalize to the population. Additionally, the timing of the study, less than six months for phases I-III, limited the sample size for our data analysis.

Future Work

We aim to continue collecting data on successful sedations using lower doses of propofol for miscellaneous pediatric procedures. Increased data will improve the power of our study and therefore its generalizability to the larger population. Future studies could expand upon dosing in other commonly used sedatives (midazolam, nitrous oxide) to

ensure patients are receiving the lowest effective dose. Future studies could further analyze patient vitals during propofol sedation to determine if IV saline boluses are necessary as standard protocol.

Conclusion

When evaluating the dose of propofol as a sedating agent for imaging intervention at Sanford Children's, a lower dose of propofol was 84 percent successful compared to a higher dose. Overall, we conclude that lowering the propofol dose is still successful and prevents the chance of overdosing. We conclude that creating a protocol with a baseline propofol drip rate of 175-199 mcg/kg/min for pediatric sedations would allow for successful sedations and prevent unnecessary excess dosing.

Acknowledgements: Thank you to the University of South Dakota Sanford School of Medicine faculty and staff for the opportunity to participate in quality improvement projects. We would like to especially thank those who provided support toward the completion of this project. Also, a special thank you to the pediatric physician intensivists and the entire outpatient nursing staff at Sanford Children's Hospital, Sioux Falls, South Dakota, for their commitment to patient care and their ongoing support of healthcare quality improvement work.

This project was supported by T35 NIH grant 5T35HD088383-04 funding to EPL, MNB, and MTV.

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About the Authors:

Mir H. Ali, MD, Pediatric Critical Care Intensivist, Sanford Children's Specialty Clinic, Sioux Falls, South Dakota; Associate Professor, Department of Pediatrics, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Michael Vlach, MS IV, University of South Dakota Sanford School of Medicine.

Ethan Leif, MS IV, University of South Dakota Sanford School of Medicine.

Madisyn Braun, MS IV, University of South Dakota Sanford School of Medicine.

Bouveret Syndrome: A Rare and Dangerous Cause of Gastric Outlet Obstruction

Derek Rohlf, MS IV; McKayla Wieczorek, MD; and Tyler Bergstrom, MD

Abstract

Bouveret syndrome is a rare and dangerous complication of cholelithiasis in which a gallstone lodges in the distal stomach or proximal duodenum, resulting in gastric outlet obstruction. We report the case of an 85-year-old female that presented without many of the clinical symptoms and complaints characteristic of gallstone ileus, further complicated by significant cardiac pathology. We review the existing studies of this uncommon disease and discuss its clinical presentation, diagnosis, and therapeutic options.

Introduction

Bouveret syndrome is a rare and dangerous complication of cholelithiasis. Chronic inflammation of the gallbladder creates a biliary-enteric fistula through which a gallstone lodges in the distal stomach or proximal duodenum. This results in gastric outlet obstruction.[1] Clinical presentation is characteristic of gallstone ileus, including nausea and vomiting, epigastric pain, abdominal distention, fever, and chills. The typical patient is elderly with multiple comorbidities. This, combined with the relative rarity of the disease and frequent delays in diagnosis, results in significant morbidity and mortality. Estimates of mortality are as high as 24 percent.[1,2] Diagnosis is typically made with CT or endoscopy, the latter providing an opportunity for removal of the stone and resolution of the obstruction.[1,4] In the majority of cases, definitive treatment is surgical enterolithotomy without repair of the biliary-enteric fistula and without cholecystectomy.[1,5,6] This study describes a case of Bouveret syndrome unique for the initial absence of key physical exam findings and complaints that are otherwise typical of this condition. It was further complicated by significant cardiac comorbidity. The patient was treated successfully with surgery and discharged without complication.

Description

An 85-year-old woman with a past medical history of moderate carotid artery stenosis, non-ST segment elevation myocardial infarction, and cerebrovascular accident presented to the emergency department complaining of nausea, vomiting, and decreased appetite of one week duration. She denied any abdominal pain, chest pain, hematemesis, or constipation. Physical exam was negative for abdominal distention, rebound tenderness, or guarding. Bowel sounds were normal. High-sensitivity troponin I was significantly elevated to 1,025.7 pg/mL (normal values ≤ 14.9 pg/mL), and ECG showed ST segment depression in leads I, II, and V4 through V6. The patient's abdominal symptoms were thought to possibly be the atypical presentation of a myocardial infarction. It was decided to first perform cardiac catheterization, and then proceed to abdominal and pelvic CT should cardiac intervention prove inconclusive.

Cardiac catheterization revealed a mostly stenotic left anterior descending artery. Two drug-eluting stents were placed. Despite cardiac intervention, the patient continued to report significant nausea postprocedurally with several episodes of vomiting and an inability to tolerate any oral intake. Over the following days the patient developed moderate abdominal distention. On hospital day five an abdominal CT was ordered, revealing a severely dilated stomach and proximal duodenum with a calcified foreign body in the second portion of the duodenum (Figure 1). Imaging also showed mucosal hyperenhancement involving the common bile duct indicating cholangitis. These findings suggested a gastric outlet obstruction secondary to an impacted gallstone that had eroded through a biliary-enteric fistula. Gastroenterology was consulted and a nasogastric tube was placed for gastric decompression. Placement of the nasogastric tube resulted in significant output.

On hospital day six endoscopy was performed, confirming the presence of a large, impacted gallstone (>3 cm) in the duodenal bulb. Extraction of the foreign body was attempted using a snare, roth net, trapezoid basket, and talon grasper (Figure 2). Removal was unsuccessful due to limited mobility

Figure 1. a) Transverse view. b) Coronal view. c) Sagittal view. CT scan was completed on hospital day five after no improvement in nausea and vomiting. Imaging shows a calcified mass (arrow) in the second portion of the duodenum with a severely distended, fluid-filled stomach (*).

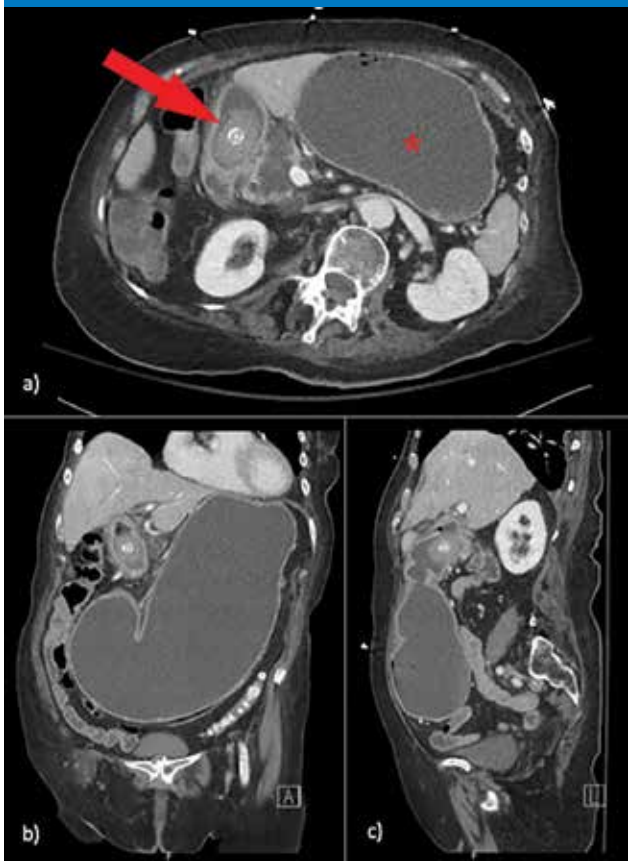

Figure 2. Images taken while attempting to remove the stone endoscopically.

Figure 3. The gallstone. Ruler markings indicating centimeters.

and a failure to adequately grasp the stone.

On hospital day eight exploratory laparotomy was performed. Laparotomy was chosen over laparoscopy given the predicted large gastrotomy that would be required to remove the stone and for better control of any intraoperative spillage from the gastrointestinal tract into the abdominal cavity. The mass was palpated in the proximal duodenum. The surgeon was able to manually dislodge and manipulate the stone into the stomach. No significant ulceration or perforation of the bowel was identified. Gastrotomy was performed along the greater curvature of the stomach and the gallstone was extracted (Figure 3). A Jackson Pratt drain was placed in the right upper quadrant prior to closure and the nasogastric tube was left in place.

Post-operatively, the patient was placed on total parenteral nutrition. Nasogastric tube output decreased after surgery and it was clamped on postoperative day two. The patient began a liquid diet, and peripheral parenteral nutrition was continued while the patient made the transition to oral nutrition. Return of bowel function occurred on postoperative day five. The patient was discharged to a transitional care facility with a swing bed on postoperative day 11.

Discussion

Cholelithiasis is a common finding in the U.S. and Western countries. It's estimated that approximately 6 percent of men and 9 percent of women have gallstones, the majority of which do not cause symptoms and are detected incidentally on imaging.[7] Gallstone ileus — the impaction of a stone in the bowel after passing through a biliary-enteric fistula — is a rare complication of cholelithiasis, occurring in 0.3-4.0 percent of patients with gall stones.[1] The majority of gallstone impactions in the bowel occur at the terminal ileum (60-70 percent). An extremely rare variation of gallstone ileus occurs when a stone lodges in the duodenum and causes an obstruction of the gastric outlet. This is termed Bouveret syndrome, and is estimated to account for 1-3 percent of cases of gallstone ileus.[1] A 2019 case report on Bouveret syndrome stated that only 315 cases of this condition had been documented in the literature between the years of 1967 and 2016.[2]

A comprehensive review of 128 cases of confirmed Bouveret syndrome determined the average age of affected patients to be 74.1 years old, with a female-to-male ratio of 1.86.[6] This review identified nausea and vomiting as the most common presenting symptoms (86 percent of patients), consistent with

the typical findings of an upper gastrointestinal obstruction. The next most commonly reported symptom characteristic of this syndrome is abdominal pain (71 percent of cases), which was notably absent in our patient. Other common findings described in the review are abdominal tenderness and abdominal distention, which were not observed in our patient on the initial exam. Bouveret syndrome is often preceded by a case of acute cholecystitis, the clinical signs of which (abdominal tenderness, right upper quadrant pain, fever) were not demonstrated in this patient.[8-10] This likely explains why ultrasonography, the preferred diagnostic tool for the detection of cholecystitis and gallstones, was not initially performed.[10]

Radiologic imaging such as CT is useful in determining both the cause and location of a bowel obstruction. Although ultrasound remains the imaging modality of choice for diagnosing uncomplicated cholelithiasis, CT is helpful in the evaluation of more acute complications such as gallstone ileus and gastric outlet obstruction as seen with Bouveret syndrome.[11] Magnetic resonance cholangiopancreatography (MRCP) is non-invasive and beneficial for both evaluating the biliary tree and identifying the location of the biliary-enteric fistula. Endoscopy is advantageous in that it can provide confirmation of the diagnosis and offer a therapeutic modality.[1-3] Endoscopic intervention is often unsuccessful due to problems with visualization, inflammation of the surrounding tissue, and immobility of the obstructing stone, as was demonstrated in this case. Over 90 percent of patients require surgery for definitive treatment. An estimated 58 percent of surgical patients underwent a previous failed endoscopic procedure before surgical resolution of the obstruction.[1,6,8] Despite therapeutic success in less than 10 percent of cases, endoscopy is the preferred first-line treatment given that the usual Bouveret syndrome patient is elderly with multiple comorbidities.[6,8]

For the majority of patients, surgical treatment consists of open or laparoscopic removal of the stone without subsequent cholecystectomy or repair of the fistula due to widespread inflammation and increased risk to the patient. Existing studies of gallstone ileus don't provide conclusive evidence that removal of the inflamed gallbladder or repair of the fistula provides a comparative improvement in outcomes.[1,12] A nationwide, retrospective review of surgical treatment for gallstone ileus concluded that enterotomy with stone extraction alone was associated with lower mortality and shorter hospital stays than more involved procedures.[5] This finding may be attributable to spontaneous closure of the fistula and shrinkage of the diseased gallbladder. It might also be explained by surgeons electing more conservative treatment for higher-risk patients. Routine follow up is not indicated in these patients provided they remain asymptomatic as in the majority of cases.[12] Recurrent gallstone ileus is seen in only 6 percent of patients that undergo enterolithotomy alone, and other persistent biliary symptoms (i.e. cholangitis, cholecystitis, cholangiocarcinoma) require treatment in less than 10 percent of cases.[12,13] In situations for which the surgeon elects to remove the gallbladder and repair the fistula, this can be performed during the initial enterolithotomy or as a second, separate procedure. Generally, the one-step approach is reserved for patients with fewer comorbidities that are deemed healthy enough to withstand a longer, more involved surgery.[1,13]

Conclusion

Bouveret syndrome is a rare and dangerous complication of cholelithiasis. As with all forms of gallstone ileus, presenting symptoms are nonspecific, and comorbidities in an elderly patient may obscure exam findings. A high degree of clinical suspicion is required for diagnosis. Imaging with CT and/or endoscopy is usually required for confirmation. Given the advanced age and relative poor health of the typical patient, endoscopy should be the first line treatment despite its low rate of success. Surgical treatment is required for removal of the gallstone and resolution of the obstruction in the majority of cases, with no definitive evidence to suggest that the gallbladder needs to be removed or the biliary-enteric fistula repaired.

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About the Authors:

Derek Rohlf, MS IV, University of South Dakota, Sanford School of Medicine, Rapid City, South Dakota.

McKayla Wieczorek, MS IV, University of South Dakota, Sanford School of Medicine, Sioux Falls, South Dakota.

Tyler Bergstrom, MD, Rapid City Medical Center, Rapid City, South Dakota.

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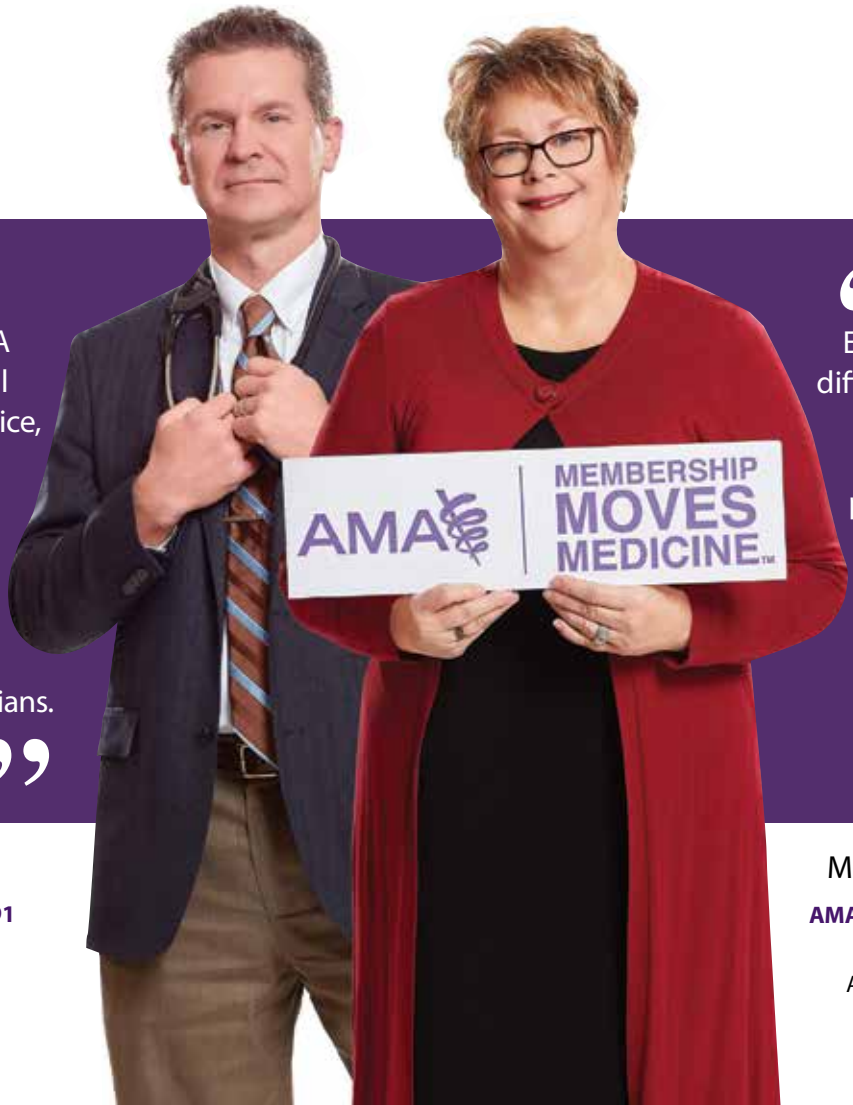


"The AMA and SDSMA help me "filter out" all the noise in my practice, which allows me to focus on the issues that are most important to me—my patients, the practice of medicine and my fellow physicians."

Robert Allison, MD
AMA MEMBER SINCE: 1991
South Dakota
AMA House of Delegates

"Every locality has a different perspective on what is most important for providing excellent health care. Everyone must be represented when policy is being crafted."

Mary Carpenter, MD
AMA MEMBER SINCE: 1982
South Dakota
AMA House of Delegates

Approach to Solitary Thyroid Nodules in Adults

Cody Ness, MD; Brian Duxbury, DO; Lauren Van Hove, MD; Wesley Halseth, MD; and Elizabeth Tacl, MD

Abstract

Thyroid nodules are exceedingly common in the general population and their increasing incidence appears to be secondary to incidental finding on imaging. Nonetheless, due to the potential for malignancy and thyroid dysfunction, most thyroid nodules require further investigation. Although there are no current guidelines for thyroid cancer screening in asymptomatic patients, a thorough history and physical which focuses on risk factors can serve as a good starting point during the evaluation of a thyroid nodule. This is followed by diagnostic analysis with thyroid-stimulating hormone; as well as thyroid scintigraphy, T4, and T3 when indicated. Ultrasound is the gold standard diagnostic imaging modality for suspicious thyroid nodules and can provide further information on malignancy potential and the need for fine need aspiration (FNA). Thyroid nodules can then be further classified on a spectrum ranging from benign to malignant based on a combination of ultrasound and FNA findings. Patients with thyroid nodules that are malignant, suspicious for malignancy, or intermediate lesions should be referred on to a surgeon for potential operative intervention. It is important for primary care providers to be well versed in the work-up and initial evaluation of thyroid nodules as they are often the first provider a patient will present to. This review article serves to refresh and guide the primary care provider through the initial evaluation and management of thyroid nodules.

Introduction

Thyroid nodules are discrete lesions within the thyroid gland that are radiologically distinct from the surrounding thyroid parenchyma.[1] These common entities are detected in 50 to 60 percent of healthy individuals at some point in their lifetime, and 25 percent of thyroid nodules are asymptomatic upon their discovery.[2,3] The rising identification rates of asymptomatic thyroid nodules is largely attributed to the progressively increased use of diagnostic imaging. 67 percent are found on ultrasonographic (US) imaging, 15 percent with computed tomography (CT) or magnetic resonance imaging (MRI) of the neck, and 1-2 percent with fluorodeoxyglucose (FDG) positron emission tomography.[4] Fortunately, most thyroid nodules are benign (90 percent); however, nodules may also harbor malignancy (10 percent) or result in thyroid dysfunction (5 percent).[3] The most commonly identified benign neoplasm of the thyroid gland is a follicular adenoma. Conversely, the most frequently encountered malignant neoplasm is a papillary thyroid carcinoma; followed by follicular carcinoma, medullary thyroid carcinoma, and undifferentiated carcinomas (also known as anaplastic or pleomorphic). Most malignant neoplasms of the thyroid have excellent long-term outcomes with appropriate surgical referral and management.[5] A particularly important consideration is that it is often impossible to distinguish benign from malignant thyroid nodules upon initial identification. Thus, all thyroid nodules > 1 cm warrant further investigation and potentially surgical referral. This article serves to refresh and guide the primary care provider through the evaluation and management of thyroid nodules.

Diagnostic Approach

History and Physical

Thyroid nodules may present in a variety of different ways including a palpable lump, symptomatic or cosmetic complaints, or as an incidental finding on diagnostic imaging. While most patients with an incidentally found nodule are asymptomatic, some present with pain or local symptoms such as globus sensation or compression (dysphagia, odynophagia, dyspnea, stridor, cough, dysphonia). Thyroid nodules can also contribute to aberrations in thyroid hormone; therefore, it is important to inquire about symptoms of hyper- and hypothyroidism. Thyroid nodules are more common in females (4 times more than men), those with iodine deficiency, increasing age, and increasing body mass index. History of radiation exposure, rapid growth of the nodule, family history of thyroid cancer, or hereditary syndromes (multiple endocrine neoplasia 2a or 2b, familial adenomatous polyposis, Cowden's disease) can all increase the risk for thyroid cancer and their presence should prompt workup and appropriate referral.[1,3]

Physical exam of the thyroid gland should include inspection

and palpation of both the thyroid and surrounding cervical lymph nodes.[1] The provider should pay close attention to location, size, and character as a large, firm, fixed, or rapidly growing mass would be immediately concerning.[6] The U.S. Preventative Services Task Force recommends against screening for thyroid cancer in asymptomatic adults; however, this does not apply to those at increased risk as above.[7]

Diagnostic Analysis

The first laboratory test to obtain when evaluating a thyroid nodule (>1 cm) is a thyroid-stimulating hormone (TSH) level. If TSH is high or normal this indicates a hypofunctioning "cold" nodule and should be further evaluated with FNA if it meets clinical and ultrasound (US) criteria that will be discussed below.[1,8] If TSH is low, this indicates a hyperfunctioning "hot" nodule or another thyroid pathology all together as the source of hyperthyroidism. In these instances, a thyroid scintigraphy/radionucleotide iodine-123 scan, as well as free T3 and T4, should be obtained to determine if the thyroid nodule is hyperfunctioning. Hyperfunctioning nodules are rarely malignant and do not require cytologic evaluation with fine need aspiration (FNA). Conversely, if thyroid scintigraphy demonstrates a hypofunctioning thyroid nodule in the setting of another thyroid pathology, such as Grave's disease, as the etiology of low TSH this thyroid nodule should be further evaluated with FNA if it meets clinical and U.S. criteria. While an elevated TSH can be associated with higher incidence and stage of thyroid cancer, most patients with thyroid nodules (and thyroid cancer) are euthyroid.[1] Current guidelines do not recommend routine measurement of serum thyroglobulin and calcitonin during the initial evaluation of a thyroid nodule.[1]

Ultrasound Imaging

Diagnostic thyroid US with survey of cervical lymph nodes should be performed in all patients with known or suspected thyroid nodules.[1] Ultrasound is a high yield imaging study that can answer many questions about thyroid nodules including location, size, involvement of cervical lymph nodes, and specific US characteristics (Figure 1), all of which increase or decrease suspicion for malignancy. Due to the high prevalence of thyroid nodules and variable US characteristics, the ACR Thyroid Imaging, Reporting and Data System (TI-RADS) was created to provide standard terms for US reporting and guide clinical decision making based on US characteristics.[9] These US characteristics include composition, echogenicity, shape, margin, and echogenic foci. Points are awarded for each characteristic and the sum determines the nodule's TI-RADS level (benign, not suspicious, mildly suspicious, moderately suspicious, and highly suspicious) as shown in Figure 2. A combination of TI-RADS level and size can provide further recommendations for the need to perform FNA or follow-up US (Figure 2).[1,9] Patients with multiple thyroid nodules have the same risk of malignancy as those with solitary thyroid nodules, however, each nodule > 1 cm carries an independent risk of malignancy and should be evaluated similarly to a solitary

Figure 1. American Thyroid Association thyroid nodule sonographic patterns and associated risk of malignancy.[1]

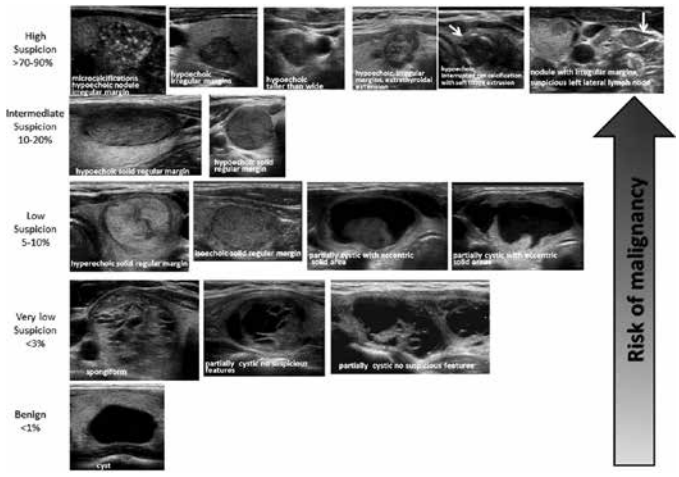
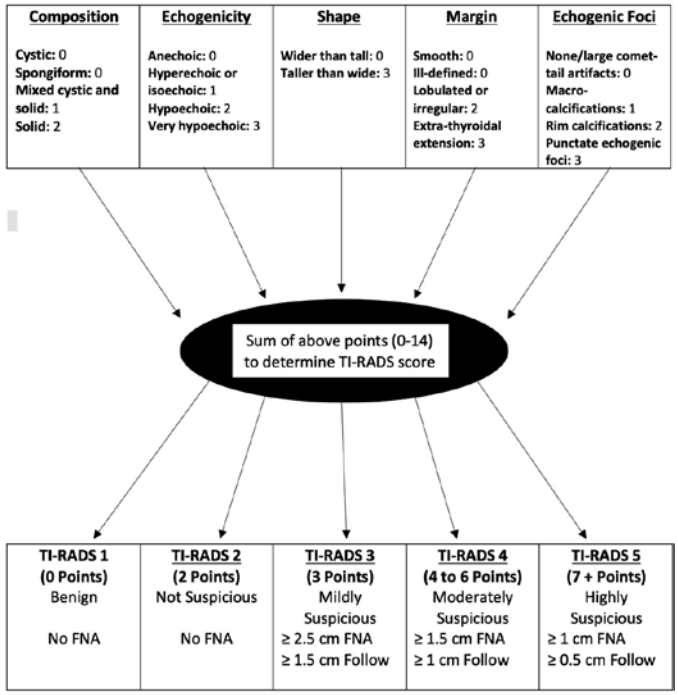


Figure 2. American College of Radiology (ACR) Thyroid Imaging, Reporting and Data System (TI-RADS).[9]



thyroid nodule.[1] Lymph nodes in either the central or lateral neck compartments that are suspicious for thyroid cancer on ultrasound require an FNA with cytology and possible thyroglobulin washout, discussed below.

Cytologic Evaluation

Thyroid nodule FNA offers accurate and definitive diagnostic information.[1] FNA is cost-effective, low risk, and usually performed at bedside under US guidance by an experienced physician. Information obtained from FNA is categorized by the Bethesda System for Reporting Thyroid Cytopathology (BSRTC), which is a standardized reporting system for thyroid FNA specimens widely accepted across the U.S.[10] BSRTC contains six categories with a respective malignancy risk for each category, and effectively guides clinical management (Table 1). It is sometimes difficult to extract enough cells with FNA for adequate cytological analysis, in these situations thyroglobulin washout is used to measure the concentration of thyroglobulin in the FNA washout, improving diagnostic accuracy.

Table 1. The Bethesda System for reporting thyroid cytopathology, associated risk of malignancy, and recommended clinical management.[10]

Bethesda category	Malignancy risk	Management
Category 1 (nondiagnostic or unsatisfactory)	0-5%	Repeat FNA with ultrasound guidance
Category 2 (benign)	0-3%	Clinical and sonographic follow-up
Category 3 (atypia of undetermined significance or follicular lesion of undetermined significance	10-30%	Repeat FNA or molecular testing or lobectomy
Category 4 (follicular neoplasm or suspicious for a follicular neoplasm	25-40%	Molecular testing or lobectomy
Category 5 (suspicious for malignancy)	50-75%	Total thyroidectomy or lobectomy
Category 6 (malignant)	97-99%	Total thyroidectomy

Molecular Testing

Molecular testing, including both mutational and gene expression analyses, of FNA specimens has become increasingly popular in the U.S.[11] Thyroid molecular testing was originally developed to reduce unnecessary diagnostic thyroid surgery for indeterminate thyroid nodules (Bethesda categories 3 and 4) but can also guide extent of surgery. The American Thyroid Association recommends molecular testing to further stratify indeterminate nodules after consideration of both clinical and sonographic features.[1] Some centers routinely obtain molecular testing on cytologically indeterminate thyroid nodules, while others have a more tailored approach. Patients with indeterminate thyroid nodules should be referred to Endocrinology or a high-volume thyroid surgeon for consideration of these additional diagnostic tests, based on institutional practice.

Overview of Operative Management

Referral to a surgeon for operative management of a thyroid nodule is absolutely indicated after a cytopathologic designation of Bethesda 5 (suspicious for malignancy) or Bethesda 6 (malignancy) on FNA. When an indication for thyroidectomy is present, multiple factors are considered prior to determining the extent of resection, including symptoms, presence of contralateral nodular disease, thyroid functional status, comorbidities, family history, surgical risk, and patient preferences. According to the American Association of Endocrine Surgeons, in general, most patients with differentiated thyroid carcinoma (DTC; papillary and follicular histologies) with tumors ≥ 4 cm, evidence of local invasion, nodal or distant metastases, multiple bilateral nodules, or evidence of medullary thyroid carcinoma should undergo total thyroidectomy. Patients with DTC 1 to 4 cm in size without aggressive cytologic or US features and no other identified reason for initial total thyroidectomy, may be offered ipsilateral lobectomy and isthmusectomy and are less likely to require postoperative radioactive iodine (RAI); however, such patients should be informed that close follow-up of the contralateral lobe and cervical nodal basins are indicated. In patients undergoing thyroid lobectomy, thyroglobulin measurements will be difficult to interpret (due to remaining thyroid tissue), and if RAI is indicated, the patient will first require completion thyroidectomy. Finally, patients with isolated DTC ≤1 cm or minimally invasive follicular thyroid carcinoma and no other worrisome radiographic or clinical features, such as lymph node metastasis or local invasion, are usually treated with lobectomy alone.[13] There is a growing role for active surveillance of subcentimeter papillary thyroid cancer in select centers, though this is beyond the scope of this paper and not included in the most recent ATA guidelines.

Patients with indeterminate lesions, including follicular lesions/atypia of unknown significance (FLUS/AUS; Bethesda 3) and follicular neoplasms (Bethesda 4), should also be considered for molecular testing or diagnostic thyroidectomy and referral to Endocrinology. Patients with a Bethesda 3 designation may also be offered the option of repeat FNA to potentially better stratify their risk of malignancy. For these individuals, clinical factors, radiographic findings, and patient preference should be included in the informed decision-making process.[13] Patients with indeterminate nodules who elect nonoperative management should be

followed with serial exams and ultrasound. Finally, patients with benign nodules but significant compressive symptoms are candidates for thyroid surgery.

Conclusion

Thyroid nodules are exceedingly common in the general population and a majority remain asymptomatic. Most of these nodules are benign, however may be malignant or have an effect on thyroid function. It is impossible to differentiate a benign from malignant thyroid nodule based on a history and physical alone, therefore further investigation is warranted. It is important for the primary care provider to be familiar with the presentation, work-up, management, indications for surgical referral, and surveillance of thyroid nodules given increasing incidence of this already common diagnosis.

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About the Authors:

Cody Ness, MD, General Surgery Residency Program, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Brian Duxbury, DO, General Surgery Residency Program, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Lauren Van Hove, MD, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Wesley Halseth, MD, General Surgery Residency Program, University of South Dakota Sanford School of Medicine, Sioux Falls, South Dakota.

Elizabeth Tacl, MD, Centracare Surgery, St. Cloud, Minnesota.

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The SDSMA is looking for volunteers for the association's Doctor of the Day Program.

This volunteer commitment involves one day at the State Capitol in Pierre during the state legislative session, by providing basic medical assistance to elected representatives and staff as needed. South Dakota's 2023 legislative session opens Jan. 10.

- **See a listing of available dates and learn more about the program at sdsma.org.**

Doctors of the Day have the unique opportunity to interact with legislators and get a first-hand look at the legislative process and how it affects the practice of medicine. Your presence at the Capitol shows legislators not only your expertise, but also your concern for the health of South Dakotans.

It is critical that volunteer physicians are serving each day of session. Every year we receive requests from PAs and NPs who wish to participate in the program.

Questions? Contact Justin Ohleen, SDSMA Director of Advocacy and Policy, at 605.336.1965 or johleen@sdsma.org.

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Renew your membership for 2023

Thank you for your membership in the SDSMA! Membership renewal for 2023 is now available — if you haven't yet or aren't signed up for auto-renew, please renew soon to ensure your benefits are not interrupted (students, residents and honorary life members do not need to act).

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Your involvement and advocacy strengthens physician voices, and provides a strong avenue for physicians to be advocates for patients.

SDSMA Representatives Attend AMA Meeting in Hawaii

SDSMA Delegates Mary S. Carpenter, MD; Robert L. Allison, MD, and Alternate Delegate Robert J. Summerer, DO, along with SDSMA President Lucio N. Margallo II, MD; and CEO Barb Smith attended the American Medical Association Interim Meeting Nov. 11-15 in Hawaii.

The meeting was filled with activities and policy debate that will help shape the future of health care in the nation. Catch up with the news and other key moments from the AMA House of Delegates meeting at ama-assn.org and in the next issue of South Dakota Medicine.

In addition, the North Central Medical Conference held its meeting; the NCMC is made up of the SDSMA and the medical societies of surrounding states.

Robert J. Summerer, DO; Mary S. Carpenter, MD; Robert L. Allison, MD; and Lucio N. Margallo, MD, attended the AMA Interim Meeting in Hawaii.

AMA House of Delegates.

The Issue Is...

SDSMA introduces the South Dakota Coalition for Excellence in Patient Care to safeguard physician-led health care and stop scope expansions that threaten patient safety

Across the U.S., non-physicians are attempting to expand their scope of practice. Now, the SDSMA is announcing the launch of the South Dakota Coalition for Excellence in Patient Care, an alliance of the SDSMA, district medical societies, and specialty societies with a mission to ensure that health care in South Dakota continues to be delivered with a physician-led, team-based approach with a focus on patients.

During the last several state legislative sessions, special interest groups have attempted to change state law to remove physician collaboration or oversight of non-physician health professionals. A bill that would remove physician supervision of physician assistants is expected to be proposed and debated in the upcoming 2023 session.

"While all health care professionals play an essential role in providing care for patients, and PAs are important members of the care team, their skillsets are not interchangeable with that of fully trained physicians. The high-stakes field of medicine demands the leadership that comes with the education, experience and acumen of a fully trained physician," said Lucio N. Margallo II, MD, president of the South Dakota State Medical Association. "I work closely with PAs and rely on them to help care for my patients. However, a team-based model is vital for high-quality patient care."

The coalition will work to block legislation proposing inappropriate expansion of medical services and procedures non-physicians are allowed to perform, and will help educate legislators about the pitfalls of scope of practice expansion legislation.

Scope expansion does not equal expanding access to care

A review of the geographic distribution of the healthcare workforce shows that removing physician supervision of PAs does not equal expanding access to care or more coverage in underserved geographic areas. In states that changed laws to allow non-physicians to practice without physician involvement, healthcare workforce shortages still persist.

Changing state laws to expand scope of practice can lead to increased healthcare costs

There is strong evidence that removing physicians from the care team has resulted in increased health care costs for patients. This is due to over-prescribing, overutilization of diagnostic imaging, and other services. This increased utilization has important ramifications on patient safety, quality of care, and the cost of healthcare in South Dakota.

Healthcare teams need leaders

Patients are concerned that allowing non-physicians to provide physician-level care is a step in the wrong direction. Healthcare is best delivered with a team-based approach. This includes physicians and other health professionals working together, sharing decisions and information for the benefit of the patient.

SDSMA and coalition members will receive updates and alerts from the coalition, including information about scope-related issues as the 2023 state legislative session nears. Stay tuned for more updates as the coalition prepares its efforts for January.

Legal Brief Highlight: Confidentiality of Patient Communication

Both South Dakota law and the HIPAA-mandated federal medical privacy rules generally prohibit the release of health care related information to third parties. However, this legal privilege against disclosure is not without exception. Federal privacy rules and South Dakota law permit, and in some instances even require, the disclosure of information to law enforcement officials and others in specified circumstances.

Individual healthcare related information is protected both under state and federal law, and may be disclosed to a third party only if permitted or required by law. Physicians should exercise care prior to disclosure of any healthcare related information, including knowledge of illegal drug activity. However, reporting the illegal use of drugs or diversion is required when done in compliance with a court order or court-ordered warrant, subpoena or an administrative request.

Disclosure of healthcare related information is permitted, but not required, in connection with other limited law enforcement related conditions, and is required in certain circumstances, including cases of abuse and neglect.

More information is available in the SDSMA legal brief *Confidentiality of Patient Communication* at sdsma.org. The SDSMA provides legal briefs for member physicians on a variety of rules and regulations that apply to the medical profession.

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PATIENT EDUCATION:

Germ Theory, Antibiotics, and our 21st Century Challenge

Kelly Evans-Hullinger, MD

As a lover of the history of science and medicine, one of my favorite topics to read and learn about is the discovery of germ theory. Up until the mid to late 1800s, diseases had numerous other theories, and the theory of *miasma* – meaning "bad air" – dominated as an explanation for cholera, plague, and other infectious outbreaks.

Bacteria themselves were seen and discovered with the development of the first microscopes in the 1600's. Dutch scientist Antonie Van Leeuwenhoek is credited as the father of microbiology, having created the early versions of our modern microscopes. Though he saw microbes with his inventions, the idea that these tiny organisms caused disease was yet to be discovered.

Germ theory, though it had beginnings smattered in earlier times, really did not take off until discoveries by 19th century thinkers including Louis Pasteur, Joseph Lister, and Robert Koch. This was an exciting time to be a biologist, and in my opinion these careers are all worthy of blockbuster movies. By the early 1900's an enormous shift had occurred, and the idea that microorganisms could cause disease was well accepted.

Initially the discovery of germ theory was most useful in prevention – sanitation of water and food went a long way toward decreasing outbreaks of previously common diseases. But another huge change occurred in 1928 with the discovery of penicillin. Penicillin was a chemical compound secreted by a type of mold which Alexander Fleming found killed bacteria. By the mid century many other antibiotics were discovered and ultimately used to treat bacterial infections.

Antibiotics are certainly one of the greatest advancements in the history of medicine and have saved countless lives worldwide. However, as our ability to treat them has advanced, bacteria have continued to evolve. By numerous processes

some types of bacteria have changed in ways to evade once-effective antibiotics. At the same time, development of new types of antibiotics have slowed to a trickle in the 21st century. Life-threatening bacterial infections for which we have no or limited ability to treat are a real concern of experts in infectious disease.

The challenge of our era, I think, is mitigating the danger posed by antibiotic-resistant infections. The most important step we can all take is to reduce the use of antibacterial medication when it is not necessary. Challenges for our hospital teams include looking critically every day to see if and which antibiotics can safely be stopped in hospitalized patients. Preserving the efficacy of this precious resource will be the work of all of us.

Kelly Evans-Hullinger, M.D. is part of The Prairie Doc® team of physicians and currently practices internal medicine in Brookings, South Dakota. Follow The Prairie Doc® at www.prairiedoc.org and on Facebook featuring On Call with the Prairie Doc® a medical Q&A show providing health information based on science, built on trust, broadcast on SDPB and streaming live on Facebook most Thursdays at 7 p.m. central.

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