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Doulas in the delivery room

A look at economics and outcomes



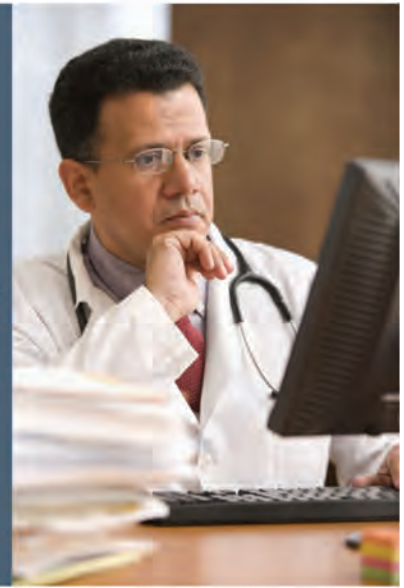
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COVER THEME

Doula in the delivery room: A look at economics and outcomes

Research has shown doula support to be associated with a wide range of positive effects on birth outcomes and maternal emotions and self-esteem. A study in this issue of *WMJ* also explores the economics of professional doula support and suggests the potential to lower costs associated with low-risk deliveries.

Cover design by
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Absence of Laboratory-confirmed Failures of High-dose Influenza Vaccine in an Elderly Population

Estimates of the limited effectiveness of conventional influenza vaccine in individuals 65 years of age and older have raised interest in the possibility of superior clinical protection with high-dose influenza vaccine.¹ We had an experience with high-dose vaccine in the 2010-2011 season in this population that offers some hope.

Taking into account the superior hemagglutinin antibody levels of high-dose vaccine² and the continuing stream of evidence correlating hemagglutinin antibody levels with clinical protection,³ we made high-dose vaccine the routine vaccine for the elderly in the Veterans Health Administration's Nebraska-Western Iowa (NWI) Health Care System. Colleagues responsible for similar patients at other sites in our region's Veterans Integrated Service Network (VISN 23), however, overwhelmingly chose standard dose vaccine.

Our laboratory identified no positive tests for influenza among 7575 elderly who received high-dose vaccines in NWI. The 8 laboratories of our VISN 23 colleagues in Iowa, Minnesota, and the Dakotas documented 22 positive tests among 36,565 vaccines. The Minneapolis site did note that one of its positive tests occurred in a high-dose recipient. Chi-square calculations comparing positive lab tests in our setting (0 of 7575 vaccines) and our colleagues' use of standard-dose vaccine (21 of 36,565) gave 3.43 (one-tailed P 0.03). This report was reviewed by the institution using a process that determined that this did not require approval by our institutional review board.

These results suggest clinical benefit of high-dose vaccine. Nonetheless, this data set has limitations. Laboratory methods varied, as did decisions to order tests. Geographic variation in the impact of influenza may occur in VISN 23. Because this was an informal review of extant data, not a prospective research study, our data

set is subject to errors in recording vaccination status and laboratory results. Conventional statistical methods, moreover, rest on the assumption of independence. This may not be valid in a community where vaccine-induced protection of 1 patient can influence the risk of influenza in an acquaintance.

Nevertheless, a Bayesian sensibility⁴ would consider the prior expectation of superiority of a high-dose vaccine that produces higher levels² of antibodies correlated with protection³ and give credence to our trend.

Our experience bolsters the view that, absent a randomized clinical trial, high-dose influenza vaccine is preferred for those 65 and older. Additionally, it hints at the value of conducting extensive observational studies, particularly in view of the similarity of results of observational studies and randomized clinical trials.⁵

Marvin J. Bittner, MD; Gary L. Gorby, MD; Chase C. Parks; Medical Service, Omaha Veterans Affairs Medical Center, VA Nebraska-Western Iowa Health Care System, Omaha, Neb

Acknowledgment: This letter does not represent the views of the Department of Veterans Affairs.

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Commentary: Solid Evidence Is Lacking, But Signs Are Hopeful

As evidenced by the recent seasonal (2012-2013) outbreak of influenza, elders bear the brunt of this pathogen, both in terms of hospitalizations and death. Sixty-five percent of Wisconsin's influenza hospitalizations were for individuals aged 65 years or older. Despite widespread use of inactivated influenza vaccine (IIV) by this group (62% coverage rate for 2012-2013 in Wisconsin), recent estimates of vaccine efficacy are disappointingly low.¹

High dose IIV was licensed in 2009, based on non-inferiority of the resulting antibody concentrations. However, we still lack good clinical evidence showing the anticipated superiority of this presentation. This is due in part to the very low prevalence of seasonal influenza in the wake of the 2009 influenza A (H1N1) pandemic, forcing extension of definitive effectiveness studies.

While awaiting the results of well-designed clinical trials, evaluations of experiences such as that presented by Bittner et al² are most welcome. Such reports provide some guidance to clinicians. Since adoption in 2010, the US Advisory Committee on Immunization Practices is increasingly using an evidence-based approach for vaccine recommendations using GRADE.³ This report would likely be scored as level 4 evidence (very low quality). This simply means that conclusions likely are to change as more evidence is gathered. Science is an iterative process. As best said by Jacob Bronowski, "We are always at the brink of the known; we always feel forward for what is to be hoped."⁴ Given the low efficacy of the current IIV in elders, we hope that Dr Bittner's conclusions are correct.

Jonathan L. Temte, MD, PhD, Professor of Family Medicine, University of Wisconsin School of Medicine and Public Health; Chair, US Advisory Committee on Immunization Practices

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An Alternative Form to Determine Patient Satisfaction with Medical Treatment

John R. Thurston, PhD

In their concern about patient satisfaction, many clinics/hospitals have come to rely on expensive multiple-choice questionnaires such as those available from Press Ganey (Press Ganey Associates Inc, South Bend, Indiana) to assess patient reactions. An important question needs to be asked and answered: to what extent have the results of these questionnaires proven useful, unsatisfactory, or irrelevant to the needs and interests of the users?

If contentment prevails, play on. If not, it's time to consider alternatives.

As a patient who has taken the Press Ganey (PG) several times, I've found completing this instrument to be long, boring, and difficult if not impossible. At no time did I ever feel I was providing information that truly reflected any of my highly personal feelings about my hospitalization. As a clinical psychologist, I once was asked by a hospital department head to help him contend with the blizzard of statistics that constitutes a PG report. He felt he had been "low-balled" when some of his department's activities were assigned a mere 30th percentile rating. As a result, there had been considerable unsuccessful soul searching by him and his staff in an effort to identify a modifiable basis for this perceived failing. They knew virtually nothing about statistics. The end result of all this was a vague feeling of guilt on their parts without a scintilla of a suggestion as to what occasioned the low

score and what might be done to improve the situation.

Alternatives to traditional questionnaires could include a patient council made up of ever-changing, recently-discharged patient volunteers and chaired by a sociable, retired physician, nurse, or other health care professional who interacts informally with patients and/or staff. Their goal would be to identify what bothered and what benefited their clientele. Ethical problems exposed should be addressed and resolved.

Another approach would involve the use of a sentence completion form. Such a form would consist of a number of sentence stems or beginnings related to a patient's experience during a hospital or clinic visit. In completing these, the patient is able to express highly personal attitudes and emotional reactions regarding what happened during the course of that visit.

A sentence completion form has many advantages. It's inexpensive and short, with a 15-20 minute administration time. It can be completed anonymously. Its instructions are simple and easily understood. It's inherently more interesting than the PG. It emphasizes expression of both personal complaints and compliments, a feature that receives only a perfunctory nod on the PG. The sentence completion form is flexible in that the hospital or clinic can evaluate special areas of concern simply by creating new, relevant sentence stems.

The patients' completions may be taken at face value as they describe personal problems and/or good experiences in their treatment. There is no need for special staff training, although an experienced reader will undoubtedly develop a greater understanding than a novice. Deeper interpretations may be undertaken by professionals if there is need.

The sentence completion form may be administered individually or in groups. Appointment of a special administrator and any en masse testing decisions should be deferred until the hospital/clinic has become thoroughly familiar with the test and what it has to offer. The determination of which patients supply this information is a matter of judgment on the part of hospital/clinic staff. It could be administered individually or to a group of patients, to recently discharged patients, or to a group of volunteers with instructions that they are to fill it out on the basis of their experiences. This and other determinations would depend upon the purpose of the survey.

I have developed a prototype sentence completion form that could be used to obtain a deeper, more individualized identification and understanding of patient satisfaction and dissatisfaction. There's nothing sacrosanct about any aspect of this form; flexibility is the name of the game. The accompanying instruction sheet with its guarantee of anonymity is designed for group administration. Users should feel free to invent new sentence stems that relate to special interest areas. They may be tacked on to the existing form or inserted in place of sentence stems that hold less interest. The last part provides an additional opportunity for specific compliments and complaints.

Hospitals/clinics have many unrecognized and unrealized resources that could supply them with information they seek. It might require a bit of creativity and "outside the box" thinking, but the issue remains a very important one.

Editor's Note: A sentence completion form prototype developed by Dr Thurston is available online at <http://bit.ly/11gpkRP>.

• • •

Dr Thurston is a clinical psychologist and professor emeritus of psychology at the University of Wisconsin-Eau Claire residing in Eau Claire, Wis. He can be reached at thurstjr@charter.net.

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Obstetric Teaching and Practice, Past, Present, and the Future

JP McMahon, MD, Milwaukee

Editor's note: The following is excerpted from WMJ, Volume 11 (No. 6), p. 178-180, published in November 1912.

The term "obstetrics" means to stand before; and, when it was adopted by our forefathers, this was as strong a term as they could rightfully employ, because all they pretended to do was to stand before the mother, with or without the perineum exposed, and wait for nature to do all but to tie and to sever the cord. Fortunately for the patients of those days, more was rarely required.

To us of the twentieth century, the term "obstetrics" should and does mean infinitely more. It means that we, as obstetricians, should be familiar with normal and all abnormal conditions of mother and child alike; that we should be acquainted with the exact physical and social condition of each mother during her entire pregnancy; that we should make observations as to position and presentation at stated intervals from the fifth month until term and frequently during abnormal or protracted labors; that we should have knowledge of just what to do and when to do it in abnormal cases; and last, but not least, that we should know when not to interfere, and thereby insure against injuring the mother and perpetrating serious and irreparable damage upon the innocent and defenseless offspring...

...All of the tragedies of pregnancy and labor, however, may not be charged to sentiment. Many of them are due to ignorance, lack of judgment and selfishness on the part of those to whom the expectant mothers have for the time being intrusted [sic] their care, welfare, and even life itself...The ignorance and lack of judgment are the result of improper, incomplete and generally inefficient instruction and clinical training...

...Inefficient training dates back to the first obstetric course offered in this country. The inadequate teaching of obstetrics was and still is part of America's poor medical educational system. The only difference between the obstetric courses and other courses is that the obstetric teaching has been and still is less efficiently done due to the following causes: incomplete preliminary and professional education on the part of the teachers, lack of laboratory and clinical facilities, failure of scientific and research men to become interested in this branch with consequent scarcity of ideal instructors to properly impress students, practitioners and the professors of the future...

...The general practitioner will undoubtedly continue to manage the greater number of labors presided over by physicians, at least until the present generation shall have retired...The least that those who intend to continue accepting the responsibilities of this work should do is to avail themselves of the advanced instruction afforded in these larger lying-in hospitals. This instruction will imbue them with the fact that obstetric practice is surgical practice, and that the more difficult obstetric operations belong to major surgery. They will also become imbued with the tact that all cases require surgical technique, skill and experience; that some cases require, in addition, muscular power; that the

management of the latter should not be attempted by the enfeebled or superannuated; that the direction of the hygiene of pregnancy and the prenatal care of the fetus are important branches of obstetric work; and that the present unnecessary sacrifice of the infants' eyes and lives is criminal.

Medical faculties must be made to realize that obstetrics is one of the most important subjects in the curriculum, that better facilities should be furnished, and that more abundant clinical material must be provided. The faculties must also be made to realize that the ideal obstetrician is necessarily a profound student of medicine and surgery, more profound than the ideal gynecologist, and a man no longer to be looked upon condescendingly by any confrere but rather a man to be looked up to as an advanced physician whose counsel should and will be sought. When colleges bring about the necessary re-adjustment, not only in the obstetric department but in all departments, the study and practice of medicine will become elevated to the plane upon which they justly belong; and their graduates will be men of only the highest ideals, who will be able to appreciate that the practice of medicine is not and should not be considered a commercial pursuit, that it is one of the three great professions whose privilege and duty it is to serve humanity, and that the only remuneration worthwhile comes from an honest effort to conscientiously perform in the best manner the tasks encountered.

The obstetrical work in all schools and hospitals should be intimately associated with and should cooperate with the work of the gynecological department. The practice of combining the chairs as followed in Europe should be universally adopted in the country under the caption of obstetrics and gynecology. A man becomes a good gynecologic surgeon but he cannot be a competent obstetrician without at the same time being a competent gynecologist...

...Future progress can be made to approach perfection by raising the scholastic qualification and the general standard of those entering upon the study of medicine, by improving didactic and clinical teaching facilities, by awarding professorships to only those who have properly qualified to become ideal instructors, by extending the lying-in hospitals so as to afford accommodations for all who can be induced to enter them, by maintaining outpatient services extensive enough to have all prospective mothers under the care of the competent obstetricians as soon as they become aware of an existing pregnancy, by teaching the laity that most of the ills of women, except those due to tumors and specific infections, are the result of poor obstetric management and that the way to prevent them is to avail themselves of the services of only those who can qualify under the standard here outlined.

Context Is Everything: How to Decide if a Journal Article Is Useful

John J. Frey, III, MD, Medical Editor

Reviewers and editors use the generalizability of a study as a major factor in deciding on the proper fit between a submitted manuscript and their journal. The more narrow the scope of a journal, the easier it may be for editorial boards, since the field may be circumscribed or, in the case of surgical journals, a procedure is less connected to the specific population and more technique or technology driven. A general medical journal such as the *WMJ* draws a wider range of manuscripts. Also, as the *WMJ* has tried to connect with a broader regional readership in the upper Midwest over the past 2 years, a question for readers might be, “How is Wisconsin similar to or different from where I work?” So, reading an article raises questions for readers but also poses challenges for editors and editorial boards.

Usefulness has to be driven by context. One of my favorite research projects carried out by one of my fellows almost 30 years ago was on the incidence of postpartum “baby blues.” She had two problems carrying out her research – there was no universal agreement about the definition of “baby blues”¹ and, as she dug deeper and deeper to find the origin of the oft-quoted study that showed 50% of women experienced baby blues, she found textbooks repeatedly cited a study from the late 1970s which, when she looked at the original research, involved a population of Latina women at a Los Angeles County hospital – hardly a useful comparison to her practice population in North Carolina.

Context often dictates whether one decides to use a study for guidance or not. In primary care, in particular, we see studies on common problems carried out in subspecialty clinics and

have to extrapolate whether they apply to our practice or not. Context includes, among other things, culture, populations, the organization of care, clinical personnel, technology, and payment systems. Authors should strive to discuss and reflect on context as they write, and we should do the same as we read.

being in an academic health center affect the availability of consultants, the universe of patients, and the method of payment?

The article by Wichmann and colleagues⁴ uses data from the Beaver Dam study, a 25-year long model of community-based research in a stable population in a small town in South

The local usefulness of studies in biomedical journals depends on the reader’s understanding of their own populations and communities, the nature of the systems in which they work, and the financial and organizational incentives that support patient care.

An example from this issue of an article where generalizability might not be as important would be the article by Kanth and colleagues² on surgical readiness. The growing literature on the value of checklists and rigorous routines in hospitals tries to “standardize” processes for maximal efficiency and quality. The important contextual issue for surgeons who read this article is whether their surgical team includes this checklist, as well as the authors’ recommendation to add preoperative assessment of cardiovascular fitness to the checklist. On the other hand, the report by Saha and colleagues³ describing a consult clinic for pregnant patients is very contextually dependent: how is the health system in which the clinic in the article operates similar to where others work; ie, does the clinic’s

Central Wisconsin,⁵ to look at changes in tobacco smoke exposure in older patients. Their finding that older members of the study population had significant decreases in environmental exposure bodes well for decreasing the risk of tobacco-related illness in that population. But of course, many communities do not have the population profile of participants in the Beaver Dam study, even in other smaller rural communities. Another contextual issue the authors raise is whether there would be differences in communities where state or local policies about public smoking are different from Wisconsin. So populations and policies affect results and perhaps generalizability.

Finally, the economic model developed by Chapple and colleagues⁶ on the potential economic effects of doula support in low risk preg-

nancies may be more generalizable to other states or regions. The authors use guidelines from the Cochrane database, which is one of the most scientifically rigorous international review systems, to frame their analyses and apply them to the statewide total statistics on birth outcomes with a series of inclusion and exclusion criteria in their formula. Wisconsin is not, for example, New Mexico, but the process and the analyses the authors use in this study are quite applicable to other populations. The absolute cost reduction would vary widely, but the study methods are easily replicable. However, to truly understand and generalize this model would require a level of granularity in the data that is not a part of their study. The readiness for a state or sub-population in a state to adopt universal or selected access to doula support in pregnancy, or a state or regional health system's ability to implement it on a wide scale, would require a more focused view of what cost savings and other outcomes might be expected.

The local usefulness of studies in biomedical journals depends on the reader's understanding

of their own populations and communities, the nature of the systems in which they work, and the financial and organizational incentives that support patient care. The best case for providing population and community health training in health sciences schools is that, without it, clinicians may change their behavior based solely on the results of a study rather than the appropriate application of those results to differing health systems and populations of patients.

Clinical research needs to increase the reliability of findings by repeating studies in larger and more representative populations that allow more generalizability.⁷ To not do so invites, at best, ineffectiveness, and at worst, clinical tragedy, by applying study results inappropriately. The context both of the study and the application of that study will often determine the likelihood of success. An example of how to understand generalizability is that, although horticultural research may demonstrate how to grow better bananas, we don't grow them in the Midwest. It would be a waste of time and money to try. So it also is for much medi-

cal research that fails to explain the context in which it takes place.

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Changes in Environmental Tobacco Smoke Exposure: The Beaver Dam Experience

Margarete A. Wichmann, BA; Karen J. Cruickshanks, PhD; David M. Nondahl, MS; Richard Chappell, PhD; Barbara E.K. Klein, MD, MPH; Ronald Klein, MD, MPH; Mary E. Fischer, PhD

ABSTRACT

Objectives: Environmental tobacco smoke (ETS) exposure has been associated with adverse health outcomes. Our goal was to determine if ETS exposure changed between 1998-2000 and 2003-2005 among participants in the population-based Epidemiology of Hearing Loss Study.

Methods: ETS exposure was ascertained using a cotinine-validated questionnaire at the 5-year (1998-2000) and 10-year follow-up examinations (2003-2005). Non-smoking participants with data from both visits were included ($n=1898$; ages 53-96 years at 5-yr follow-up). McNemar's test was used to test differences in ETS exposure overall and in 3 settings: home, work, and social settings. Generalized estimating equations (GEE) were used for multivariate logistic regression models of exposure.

Results: The proportion of nonsmokers with no or little ETS exposure increased from 80% to 88% ($P<0.0001$). The percent living in a home with no indoor smokers increased from 94% to 97% ($P<0.0001$). The percent reporting no exposure at work increased from 91% to 95% ($P<0.0001$). The percent reporting the lowest frequency of social exposure increased from 65% to 77% ($P<0.0001$). In the GEE model, age was inversely associated with overall ETS exposure (Odds Ratio [OR] per 5 yr = 0.87, 95% CI = 0.81, 0.94), as was education (OR for college vs < high school = 0.25, 95% CI = 0.16, 0.37), female gender (OR = 0.41, 95% CI = 0.33, 0.51), and later examination period (OR = 0.62, 95% CI = 0.53, 0.73); current employment was positively associated with exposure (OR = 1.44, 95% CI = 1.14, 1.83).

Conclusions: Between the late 1990s and the mid-2000s, ETS exposure in older adults decreased. Decreasing exposures suggest there may be future declines in ETS-related adverse health outcomes.

INTRODUCTION

Exposure to environmental tobacco smoke (ETS) has been associated with adverse health outcomes in nonsmokers. In 1992, the Environmental Protection Agency (EPA) classified ETS as a known human carcinogen.¹ The effects on the cardiovascular

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system are estimated to be substantial, on average 80% to 90% as large as that from active smoking (averaged across mechanistic effects, eg, endothelial dysfunction, arterial stiffness).² ETS exposure occurs in a variety of settings, including home, work, and social settings. Exposure in the home and workplace may be particularly harmful, as many people spend much of their time there. ETS exposure in the workplace previously was estimated to result in about 1710 excess ischemic heart disease deaths per year among 35- to 69-year-old nonsmoking US workers.³ A meta-analysis of 17 studies estimated a relative risk for coronary events of 1.25 (95% CI = 1.17-1.33) among never smoking spouses married to smokers, compared to those with nonsmoking spouses.⁴ In recent decades, increasing awareness of the harms of ETS has led to attempts to reduce or avoid exposure, and some cross-sectional population-based stud-

ies have reported declining temporal trends, as did 1 longitudinal study which excluded women.⁵⁻⁸

We aimed to determine if ETS exposure changed among non-smoking older adult participants in a longitudinal population-based study during the 5-year period between 2 examinations: 1998-2000 and 2003-2005. In particular, we investigated whether later time period was associated with ETS exposure in a variety of settings, after taking into account important temporal changes including age of the participants, retirement, and spousal death.

METHODS

Study Population

The Epidemiology of Hearing Loss Study (EHLS) is a population-based longitudinal study of sensory loss and aging in Beaver Dam, Wisconsin (1993-present). A private census was conducted in 1987-1988 that identified 43- to 84-year-old residents of the

Table 1. Participant Characteristics by Time Period (n=1898)

	1998-2000 n (%)	2003-2005 n (%)
Sex (Female)	1151 (60.6)	Time invariant
Age (Years)		
53-59	442 (23.3)	55 (2.9)
60-69	671 (35.4)	762 (40.2)
70-79	566 (29.8)	606 (31.9)
80-89	204 (10.8)	401 (21.1)
90-100	15 (0.8)	74 (3.9)
Education (Years)		
<12	300 (15.8)	Time invariant
12	921 (48.6)	—
13-15	330 (17.4)	—
≥16	345 (18.2)	—
Marital Status		
Never married	66 (3.6)	64 (3.6)
Separated/divorced	136 (7.3)	130 (7.2)
Widowed	370 (20.0)	471 (26.2)
Married	1280 (69.1)	1133 (63.0)
Number of Others Living in Home		
None	452 (24.0)	516 (28.4)
1	1198 (63.6)	1165 (64.1)
2	179 (9.5)	99 (5.4)
≥3	54 (2.9)	38 (2.1)
Employed		
Full-time	476 (25.1)	254 (13.4)
Part-time	257 (13.5)	250 (13.2)
Not employed	1165 (61.4)	1394 (73.4)

city or township of Beaver Dam (n = 5924). In 1988-1990, 4926 of the 5925 eligible (83%) participated in the first examination phase of the Beaver Dam Eye Study (BDES).⁹ Those who participated in the baseline BDES and were alive on March 1, 1993 (n = 4541) were eligible for the EHLS, and 3753 (82.6%) participated in the baseline EHLS examination phase from 1993-1995.¹⁰ The EHLS 5-year follow-up examinations took place from 1998-2000 and, of the 3407 EHLS participants alive as of March 1998, 2800 (82.2%) participated.¹¹ During the 10-year follow-up examination phase in 2003-2005, 2395 (82.5% of survivors) participated.¹²

Participants who provided ETS questionnaire data at both the 5-year (1998-2000) and the 10-year (2003-2005) follow-up interviews were included in this study (n = 2231). Analyses were limited to participants who were nonsmokers at both time points (n = 1898), 87.1% and 89% of all participants at the 5- and 10-year follow-up examinations, respectively. This study was approved by the Health Sciences Institutional Review Board of the University of Wisconsin and informed consent was obtained from each participant.

Data Collection

At both time points, participants provided information on ETS exposure in 3 settings: home, work, and social activities. Those not

working were coded as having no workplace exposure. Cotinine data on a subset of the cohort (n = 643) from the 5-year follow-up examinations previously were used to develop a 3-level cotinine-validated ETS exposure scale that uses only the self-reported questionnaire data, allowing estimation of ETS exposure among the entire cohort.¹³ The scale conveniently categorizes overall ETS exposure, combining information about work, home, and social ETS exposure, using 1 questionnaire item from each type of setting: the number of hours per day exposed to ETS at work (none, <1h/day, 1-4h/day, or >4h/day), exposure at home (number of housemates who smoke in the home), and the frequency of exposure in social settings (seldom, once a week, several times a week, or daily). Participants were considered to have high ETS exposure if they: (1) had >4h/day of exposure at work; (2) were living with a person who smokes in their home; or (3) were exposed daily to ETS in social settings outside the home. Participants not fitting those criteria were categorized as having moderate exposure if they had 1 to 4 h/day of workplace exposure or social ETS exposure several times a week. Finally, participants not categorized as having high or moderate exposure were considered to have little or no ETS exposure.

Because small numbers of participants reported higher ETS exposure levels, the greater categories of exposure were combined into 1 category (exposed) and compared to the lowest category (little or no exposure) as follows: for overall ETS exposure, “moderate” and “high” vs “no/little;” for exposure in social settings, ≥once/week vs seldom; for home exposure, ≥1 smoker in home vs no smokers; for work exposure, any time exposed at work vs never exposed.

Statistical Analyses

Statistical analyses were conducted using SAS software (version 9.2; SAS Institute, Inc., Cary, NC). McNemar’s test was used to test for change between the 2 time points in ETS exposure (lowest category vs higher categories), overall and in each setting. Generalized estimating equations (GEE) logistic regression was used to investigate factors associated with ETS exposure, overall and in each of the 3 specific settings. The GEE models were generated with the GENMOD procedure, using the REPEATED statement and specifying “unstructured” as the working correlation matrix to account for the correlation of responses from the same participants at 2 time points.

Models for each specific setting and overall ETS exposure included age, sex, education, and time period (2003-2005 vs 1998-2000). Additional factors that would be likely to influence ETS exposure (ie, number of other people living in the home, employment status and change in employment status, change in marital status, etc) were considered as potential confounders. Workplace exposure models were restricted to those working at both time points (n = 350). The main independent variable of

interest in all models was time period, to determine whether exposure was decreasing in this community over the 5-year period, other than through major age-related life changes such as retirement or death of a spouse.

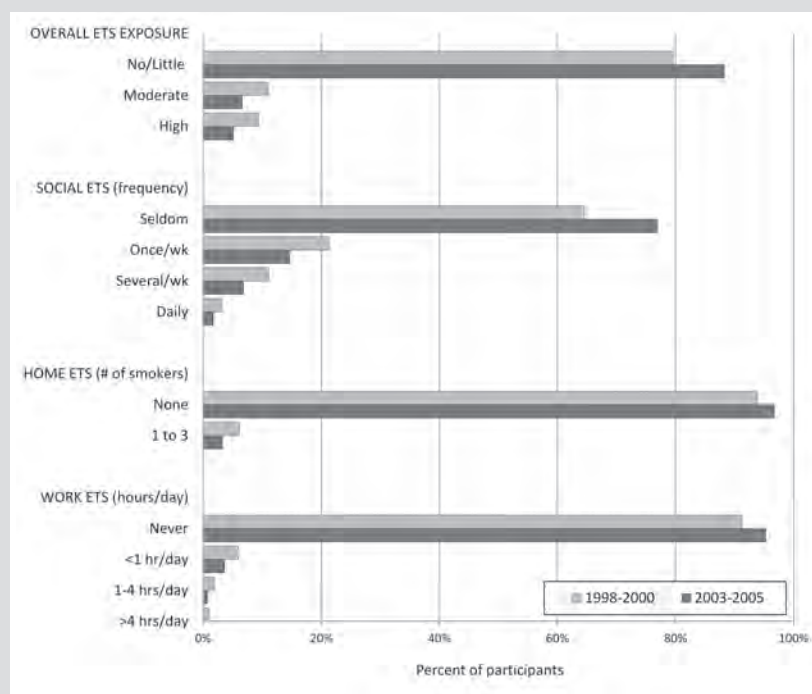
RESULTS

This sample of nonsmokers was 61% female. Fifty percent had a high school education and 35% had more than a high school education. During the period between the 5-year and 10-year follow-ups, the portion employed decreased from 39% to 27% and the portion widowed increased from 20% to 26% (Table 1).

Between the 5- and 10-year follow-up examination phases the proportion with decreased ETS exposure was significantly greater than the proportion with increased ETS exposure, overall and in each specific setting ($P < .0001$ for each). For overall ETS exposure, participants were more likely to be classified as having “no/little ETS” at the 10-year follow-up (88%) than at the 5-year follow-up examination (80%) (Figure 1). By specific setting, during the 5- and 10-year follow-up examinations, 65% and 77%, respectively, reported “seldom” being exposed to tobacco smoke in social settings outside their own home; 91% and 95%, respectively, reported no workplace ETS exposure; 94% and 97%, respectively, lived in a home with no indoor smokers (Figure 1). Among the 116 participants whose workplace exposure decreased to none, 61 had retired, 14 had changed from full-time to part-time employment, and 1 became unemployed/disabled. Of the 69 participants whose home ETS exposure decreased to none, 11 were widowed and 2 divorced during the interval.

To determine whether retirement or transitions to part-time employment were driving the decrease in workplace ETS exposure, we examined exposure change among the 350 participants who were working full-time at both points or part-time at both points (ie, those who changed from full-time to part-time or vice versa, or were not working at either or both points, were excluded). Among this subset, 78% reported no exposure at the 5-year follow-up, compared to 80% at the 10-year point ($p = 0.47$). Similarly, ETS exposure in the home was investigated amongst the subset with no change in marital status ($n = 1610$) and found 94% with no exposure at the 5-year follow-up and 97% with no home exposure at the 10-year follow-up ($P < .0001$). Among the subset who lived with at least 1 other person at both

Figure 1. Unadjusted Environmental Tobacco Smoke (ETS) Exposure (%) by Time Period, Overall and in Each Setting.



time points ($n = 1255$), the difference was slightly less (93% vs 95% with no exposure; $P < .0001$).

In a multivariate GEE model, factors associated with lower odds for overall ETS exposure were greater age, female sex, more education, and later time period (2003-2005 vs 1998-2000) (Table 2). Current employment was associated with increased odds of overall ETS exposure. For ETS in social settings, the same factors were associated with lower odds of exposure, except current employment, which was not part of the model. Fewer factors were associated with lower odds of ETS exposure at home, including later time period and having a college education (vs less than high school). Not surprisingly, the odds of exposure in the home also were increased with each additional person living in the home ($OR = 1.5$ for each person). The analysis of ETS exposure in the workplace was limited to those participants who were employed at both time points. Greater age, female sex, and a college education (vs less than high school), were associated with lower odds of exposure at work. Time period was not significantly associated with workplace exposure.

DISCUSSION

Over a 5-year period, ETS exposure declined in this population of older adults. Decreases in exposure were observed for all settings examined (home, work, and social settings). Controlling for several factors including age, later time period was associated

Table 2. Adjusted Odds Ratios for Environmental Tobacco Smoke (ETS) Exposure by Relevant Participant Characteristics [OR (95% CI)]

	Overall ETS	Social ETS	Home ETS	Work ETS ^a
Later time period	0.62 (0.53, 0.73) ^b	0.69 (0.61, 0.78) ^b	0.65 (0.51, 0.83) ^b	1.15 (0.77, 1.72)
Age (per 5 years)	0.87 (0.81, 0.94) ^b	0.24 (0.18, 0.31) ^b	0.62 (0.34, 1.13)	0.23 (0.07, 0.77) ^b
Female sex	0.41 (0.33, 0.51) ^b	0.52 (0.44, 0.62) ^b	0.71 (0.48, 1.05)	0.18 (0.11, 0.29) ^b
College (vs < high school)	0.25 (0.16, 0.37) ^b	0.57 (0.42, 0.77) ^b	0.40 (0.19, 0.87) ^b	0.20 (0.09, 0.45) ^b
Some college (vs < high school)	0.65 (0.46, 0.92) ^b	0.85 (0.63, 1.15)	1.32 (0.71, 2.44)	0.46 (0.21, 0.98) ^b
High school (vs < high school)	0.53 (0.40, 0.71) ^b	0.95 (0.74, 1.22)	0.78 (0.44, 1.37)	0.52 (0.26, 1.05)
Currently working	1.44 (1.14, 1.83) ^b	—	—	—
Lives with others (per each)	1.12 (0.99, 1.26)	—	1.53 (1.08, 2.18) ^b	—

Abbreviations: OR, odds ratio.

^a Analysis for work ETS was restricted to those working part- or full-time at both time points (n=350).^b $P < 0.05$

with reduced odds of exposure at home and in social settings, but was not significantly associated with odds of exposure at work, although temporal shifts in workplace ETS exposure may have occurred prior to 1998 as some employers instituted smoke-free policies. Older age and female sex were each associated with decreased odds of exposure at work and in social settings, but not associated with home exposure. College education, compared to less than high school education, was associated with reduced odds of exposure in all 3 settings.

In this cohort of older participants, we expected retirement and death among smoking spouses would explain some of the decreased ETS exposure. Among those working at both time points, the percentage reporting no workplace exposure was similar, but slightly greater at the later time period (80% vs 78% at the earlier time period) and later time period was not associated with reduced odds of exposure at work in the multivariate GEE model. Thus, among this older population, retirement and reduced work hours appear to account for a portion of the small decrease in prevalence of workplace exposure that was found in the whole sample. Regarding exposure in the home, we observed the same decrease as in the whole sample when we looked at the subset with no change in marital status, suggesting that death of smoking spouses was not a major reason for the small decrease noted in the prevalence of ETS exposure at home.

These results are consistent with reports of decreased ETS exposure among nonsmokers in the US population, based on several analyses using National Health and Nutrition Examination Survey (NHANES) cotinine data.⁷⁻⁸ One study reported that serum cotinine levels in nonsmokers decreased by 70% between 1988 and 2002.⁸ Another analyzed NHANES cotinine data collected from 2001 to 2006 and concluded that the previously observed declining trend in ETS exposure had leveled off.⁷ A Centers for Disease Control and Prevention report stated the prevalence among nonsmokers of serum cotinine levels ≥ 0.05 ng/mL decreased from 52.5% in 1999-2000 to 40.1% in 2007-2008, and that the greatest change was observed between the

periods 1999-2000 and 2001-2002.¹⁴ The current study included this time period in which greatest change was observed in the NHANES data.

In general, reductions in ETS exposure are thought to be due to decreases in the prevalence of smoking, implementation of nonsmoking policies and laws for workplaces and public indoor spaces, individual household bans on indoor smoking, and a general shift in social norms towards the unacceptability of exposing nonsmokers, especially children or frail individuals, to ETS.¹⁵ In the present study, the decreased odds of exposure at the later time period, independent of age and other covariates, likely reflects such changes in cultural norms in this population from south central Wisconsin, although we had no data to test this directly. It is likely that ETS exposure levels, especially in public places and workplaces, have decreased after this period due to a Wisconsin statewide ban on smoking in enclosed public places and workplaces, which took effect on July 5, 2010. Supporting this possibility, a recent study from the Survey of the Health of Wisconsin reported that participants interviewed after July 5, 2010 were less likely to report ETS exposure and more likely to report smoking bans in their households, compared to those interviewed before July 5, 2010.¹⁶ In particular, they observed greater differences among older participants, for both home and work ETS, suggesting that policy changes may have led to subsequent reductions in ETS exposure among older Wisconsin adults. Our longitudinal data, with repeated assessments in the same people, are consistent with their findings.

Interpretation of the current results should take into account some strengths and limitations of the study. A potential limitation is that exposure data was self-reported. Cotinine measures would have provided an objective measure. However, previous analyses comparing self-reported exposure and cotinine levels in a subset of this cohort indicated that self-reported ETS exposure was useful for categorizing relative exposure.¹³ In addition, cotinine level represents exposure over only the previous 2 to 3 days, whereas the participants in this study self-reported on their usual

exposure, which is more relevant for long-term health outcomes such as cancer and heart disease. Cotinine levels also are unable to provide information about the specific settings in which ETS exposure occurs, which is important for potential interventions, and is attainable through self-reported questionnaires. Our cotinine-validated questionnaire and classification scheme for ETS exposure accounted for exposure in each of 3 specific settings, which is important because neglecting exposure outside the home may lead to underestimation of total ETS exposure.¹⁷ This is a large, prospective, population-based cohort with consistently high participation rates. The current analyses were limited to those with data at both time points in order to examine change in ETS among the same group of individuals, unlike the NHANES design which utilizes serial cross-sectional samples. However, the vast majority of this population is non-Hispanic white, and results may not be generalizable to minority groups.

CONCLUSIONS

Self-reported ETS exposure in this population-based cohort of older adults decreased between 1998-2000 and 2003-2005. For exposure in social settings and the home, later time period was associated with lower odds of exposure, adjusted for age and other covariates. Decreased ETS exposure among older adults may contribute to declines in ETS-related adverse health outcomes such as heart disease.

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An Economic Model of the Benefits of Professional Doula Labor Support in Wisconsin Births

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ABSTRACT

Introduction: The purpose of this study is to estimate the immediate cost savings per delivery with in-hospital professional doula labor support in Wisconsin. This is the first study that calculates the estimated cost savings of professional doula labor support specific to Wisconsin.

Methods: This analysis used results presented in and derived from the Cochrane Review of continuous labor support to estimate procedure reduction and cost savings in Wisconsin using birth statistics from 2010. The delivery outcomes included were cesarean deliveries, instrumental deliveries, and regional analgesia use. To accurately reflect published studies on labor support, only low-risk deliveries were used for intervention reduction calculations.

Results: For 2010 data, estimated savings of \$28,997,754.80 could have been achieved if every low-risk birth were attended in-hospital by a professional doula. A professional doula providing only in-hospital labor support would yield an estimated cost savings of \$424.14 per delivery or \$530.89 per low-risk delivery.

Conclusion: A system-based change in how laboring mothers are supported would be an innovative step that would put Wisconsin at the forefront of cost-effective health care, reducing interventions while improving outcomes. It is recommended that Wisconsin insurers consider reimbursing for professional doula labor support. It is also recommended that pilot programs be implemented in Wisconsin that can better assess the implementation of professional doula labor support services.

INTRODUCTION

Labor support is defined as offering a woman comfort and encouragement during the active phase of labor, birth, and the immediate postpartum period. A doula is “a woman who provides physical, emotional, and informational support to the laboring mother throughout her entire labor.”¹ The emotional

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support skills of doulas have been shown to be a sophisticated mix of counseling skills and have not been duplicated in studies of nursing support.² Informational support refers to repeating medical information in terms the mother in labor can understand, assisting in communication with medical care providers, and explaining the reasons for using certain physical support strategies or pain coping techniques. Doulas also are experts in positioning to enhance fetal descent and lessen maternal discomfort. Individualizing comfort measures appropriately for each woman is a part of the doula's support role.³

In the United States and Canada, the common practice is for a doula to join the mother and her partner (or other friends or relatives) at the beginning of active labor and remain with her until several hours after the baby is born.¹

Research has shown doula support to be associated with a wide range of positive effects on birth outcomes, maternal emotions, and self-esteem during the postpartum period. One of the key components of the effectiveness of labor support is the continuous uninterrupted care of the mother by 1 doula.⁴ Except for toileting, the doula does not leave the mother no matter how long or short the labor. This is made exceptionally clear in a meta-analysis of 11 published clinical trials.⁴ Continuous labor support was significantly associated with shorter labors, and decreased need for the use of any analgesia, oxytocin, forceps, and cesarean sections. Intermittent support was not significantly associated with any of these outcomes. While several hypotheses have been proposed to explain the reason for the doula effect on obstetrical and neonatal outcomes,³ a specific mechanism has not been identified.

In North America, a professional doula refers to a woman who is trained and experienced in meeting the emotional needs of women and their families during the perinatal period. Almost all

professional doulas have an independent practice (IP) where they conduct 1 to 3 prenatal visits and 1 postpartum visit, usually of 2 hours duration.³ Based on personal correspondence with practicing doulas, IP doulas in Wisconsin may charge \$200 as novice doulas to \$900 for those who have attended 100 or more births.

In 2012 Hodnett et al updated the Cochrane Review that analyzed 22 trials and 15,288 women regarding continuous labor support.⁵ In the majority of randomized trials concerning labor support, the doula or caregiver meets the patient for the first time at the start of labor in the hospital, rather than building a relationship with the patient before labor begins. In addition, most studies included only full-term, singleton, and low-risk births. The review also noted that no adverse effects have been identified from the use of doula support during labor. The Cochrane Review included studies with interventions ranging from untrained companions, trained doulas, nurses, and midwives as labor support providers. With this in mind, the review's results showed that labor support is an effective option for improving many outcomes (Table 1). Most studies concerning labor support have been done outside the United States within nonwhite and low-income populations,⁵ which does not reflect the American middle class. A 2008 study⁶ addressed the concern of generalizability with a randomized study on labor support in Cleveland, Ohio. This study showed a 46.4% reduction in cesarean delivery rates and a significant decrease of epidural analgesia use in the doula group, demonstrating that the benefits of doula support for laboring women are present across countries, ethnicity, and socioeconomic status. More notably, it showed the benefits of doula support for women who also had paternal labor support.

The purpose of this cost-effectiveness analysis is to estimate the immediate cost savings per delivery in Wisconsin with professional doula labor support, compared to delivery without such support. The results may be used to guide insurance reimbursement policies to include cost-saving strategies that improve birth outcomes and patient satisfaction. The researchers hypothesize that insurance reimbursement toward the cost of professional doula support would increase access, lower cost, and improve outcomes.

METHODS

To accurately reflect the inclusion criteria of studies on labor support, high-risk births were excluded from the total number of births in Wisconsin for 2010 before calculating cost savings. The remaining low-risk births were defined as singleton and full-term births. In addition, low-risk cesarean births included adjustments for the excess risk of the interventions from gestational diabetes (high blood glucose or diabetes during pregnancy), gestational hypertension (high blood pressure during pregnancy), vaginal birth after cesarean (VBAC) success rates and feto-pelvic disproportion disorder. Then, the estimated reduction in delivery

Table 1. Birthing Outcomes Measured in Cochrane Systematic Review⁵

Outcomes Included in This Analysis		
Variable	Relative Risk	95% CI
Regional analgesia	0.86 ^a	0.80 to 0.92 ^a
Instrumental vaginal birth	0.90	0.84 to 0.96
Cesarean delivery	0.69 ^a	0.56 to 0.83 ^a
Outcomes Not Included in This Analysis		
Variable	Relative Risk	95% CI
Patient satisfaction	0.69	0.59 to 0.79
Shorter labor	Mean difference -0.58 hours	-0.31 to -0.85
Intrapartum analgesia	0.90	0.84 to 0.96
Spontaneous vaginal birth	1.08	1.04 to 1.12
Low 5 minute Apgar Score	0.69	0.50 to 0.95

^aRR and CI presented are from Table 2. (See page 60.)

interventions with professional doula care was calculated from low-risk births based on risk reduction presented in and derived from the updated 2012 Cochrane Review on continuous labor support.⁵ Finally, the cost of the interventions was applied to the number of reduced interventions to give an estimated cost savings of professional doula labor support.

The total estimated cost savings was divided among all deliveries in Wisconsin for 2010 to give the average cost savings per delivery. The total cost savings also was divided among the low-risk deliveries to give the cost savings if only low-risk deliveries were covered. Thus, the analysis presents the cost savings of professional doula labor support for in-hospital deliveries, and in-hospital low-risk deliveries. Confidence intervals (CI) were calculated in the same way using the 95% CI from Table 1 for each of the birth outcomes.

Delivery Outcomes

The main delivery outcomes included were regional analgesia use (epidural injection), cesarean delivery, and instrumental vaginal birth (Table 1). The relative risks (RR) for cesarean delivery and regional analgesia reported in the Cochrane Review were not used in this study (RR=.78 and .93 respectively) because the Cochrane Review combined studies using professional doulas and studies using non-professional labor support. When these studies were separated the outcomes were substantially different. The risk reductions reported in our study were recalculated from studies included in the Cochrane Review that utilized professional doula care. Thus, the derived relative risk of cesarean delivery (RR=.69 or a 31% reduction) and the derived relative risk of regional analgesia use (RR=.86, or a 14% reduction) were used in this analysis (Table 2).

Some outcomes from the Cochrane Review were not used in this analysis because of the difficulty in applying a direct measurable cost or measuring its prevalence (Table 1).⁵ While these measures did not have an assigned monetary value in this analysis,

Table 2. Cesarean Delivery Rates and Regional Analgesia Use in Studies Using Professional Doula Labor Support from Cochrane Systematic Review⁵

Studies	Cesarean Delivery		Regional Anesthesia ^a		Participant Description ⁵
	Doula Care	Control	Doula Care	Control	
Kennel 1991 ⁷	17/212	26/200	47/212	94/200	At Jefferson Davis Hospital in Houston, Texas. Nulliparous women age 13-34, single gestation, term, noncomplicated pregnancies in active labor. Excluded were women with breech presentation, hypertension, history of alcohol or drug abuse, gestational diabetes, or other complications.
Klaus 1986 ⁸	12/168	46/249			At the Social Security Hospital in Guatemala, full-term healthy primigravitus women in early labour who had a cervical dilatation of 3 cm or less and were without medical problems.
Langer 1998 ⁹	85/357	97/356	295/335	302/346	In Mexico City, singleton fetus, no previous vaginal delivery, < 6 cm cervical dilatation, and no indications for an elective caesarean delivery.
McGrath 2008 ⁶	30/224	49/196	145/224	149/196	Nulliparous women between the ages of 18 and 41 years in the 3 rd trimester of an uncomplicated pregnancy who expected to be accompanied during labor by their male partner and planned to deliver at University Hospitals in Cleveland, Ohio.
Total	144/961 (14.98%)	218/1001 (21.77%)	487/771 (63.16%)	545/742 (73.45%)	
Relative risk	RR: 0.69 (95% CI: 0.56-0.83) P-value <0.001		RR: 0.86 (95% CI: 0.80-0.92) P-value <0.001		

^a Empty cells indicate areas without data.

they influence other health and economic outcome factors and should be included in discussions of doula labor support or when interpreting results from this study. Quality of life measurements also were not considered in this analysis.

Criteria for the Inclusion of Low-Risk Cesarean Deliveries

First, high-risk cesarean deliveries must be excluded and adjustments made for VBAC success rate, gestational diabetes, gestational hypertension and fetopelvic disproportion disorder. The number of cesarean deliveries among women with full-term singleton births was calculated using Wisconsin Interactive Statistics on Health database (WISH) for 2010.¹⁰ The number of repeated cesarean births available for intervention reduction from doula labor support was decreased due to a 74% estimated VBAC success rate.¹¹ This VBAC success rate, with a sample size of 9437 deliveries, was reported in a secondary analysis of a retrospective cohort study conducted in 16 community and university hospitals. While other studies have reported VBAC success rates similar to the one used in this analysis,¹² a large sample size and the high quality of the study warranted its inclusion.

The number of deliveries in Wisconsin for 2010 with the primary diagnoses of gestational hypertension, gestational diabetes, and fetopelvic disproportion obstruction was obtained from the US Department of Health and Human Services' Healthcare Cost and Utilization Project (HCUP) online query system¹³ (3746, 1289, and 1186 respectively). The risk of cesarean delivery for these conditions was applied to the HCUP data. The relative risk of cesarean delivery for these variables was 2.23¹⁴ and 1.16¹⁵ respectively, with none of the fetopelvic disproportion cases considered low-risk (Figure 1).

Calculating Low-Risk Births for Instrumental Deliveries

HCUP reported 2987¹³ instrumental births (forceps, vacuum-assisted and breech deliveries) in Wisconsin for 2010. HCUP did not have data on instrumental deliveries among singleton full-term births and WISH did not include instrumental deliveries other than forceps deliveries. The reduction in forceps deliveries among singleton full-term deliveries compared to all forceps deliveries reported by WISH was used to estimate the number of singleton full-term instrumental births if they had been reported by HCUP. WISH reported 543 forceps deliveries and 487 (10.31% less) forceps deliveries among singleton full-term births in Wisconsin for 2010.¹⁰ The estimated number of instrumental births among singleton full-term births was 89.69% of 2987 births, or 2697 births.

Calculating Low-Risk Births for Regional Analgesia

Regional analgesia (epidural injection) use was not reported in WISH or HCUP and was estimated using a 2008 National Vital Statistics report¹⁶ of the epidural rate for over 1.8 million singleton vaginal deliveries among women of white, black, Hispanic and Native American ethnicities in 27 states. While Wisconsin was not included in the analysis and the rates varied by state, the application of rates by ethnicity provides a close estimate. The rates per ethnicity were applied to the WISH statistics by ethnicity for singleton full-term vaginal births in Wisconsin for 2010. It was estimated that 28,082 total epidural injections were administered.

Cost Estimates

The mean hospital charge for vaginal and cesarean delivery was taken from Wisconsin Price Point System, which presented data

Figure 1. Calculating Low-Risk Cesarean Deliveries

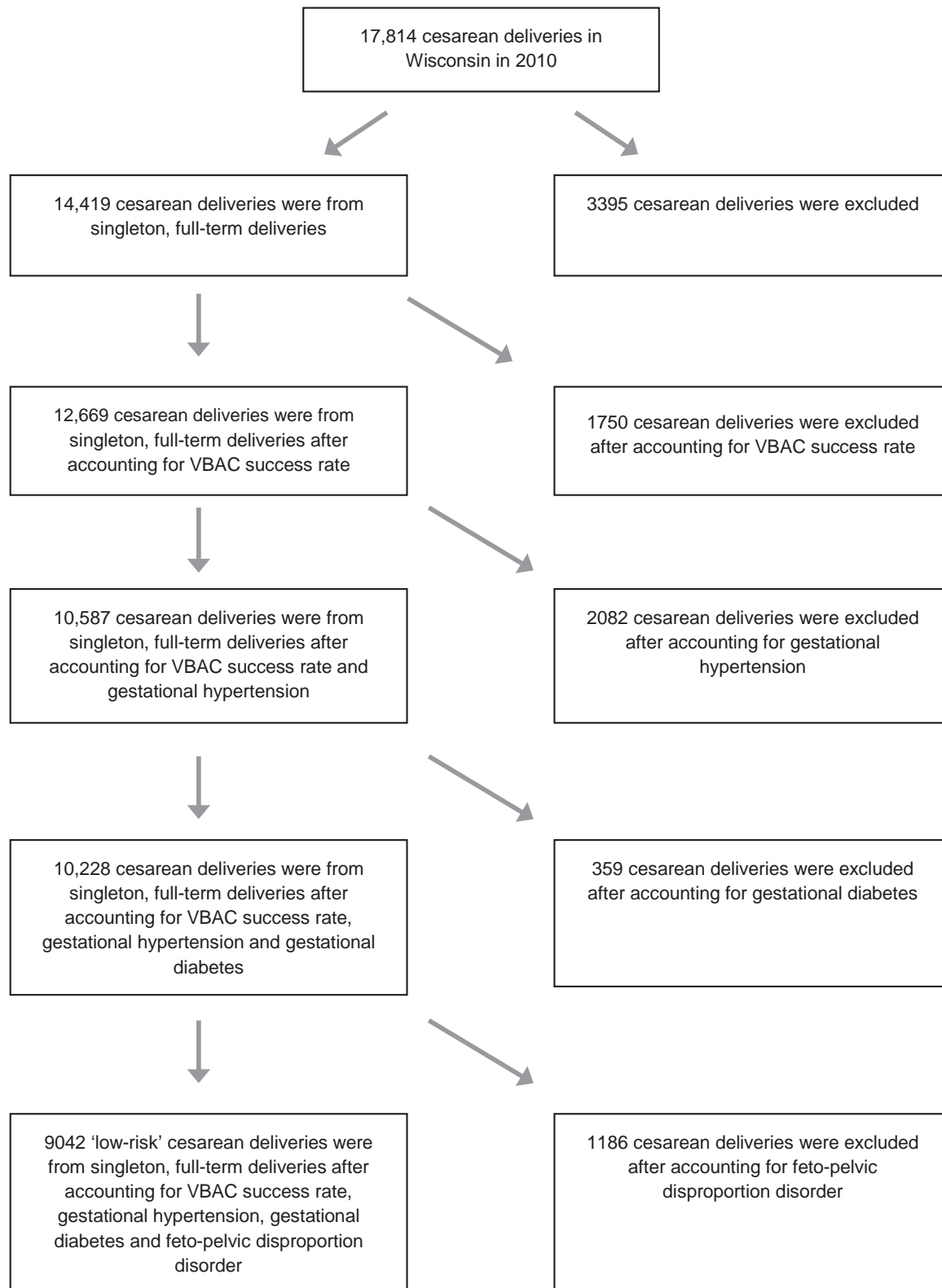


Table 3. Cost Savings if Professional Doula Labor Support Were Used in Wisconsin (2010)

	Difference in the Number of Interventions (95% CI)	Excess cost per Intervention. In US Dollars, FY 2010	Total Savings for In-hospital Service Only In US Dollars, FY 2010 (95% CI)	
Cesarean deliveries	2803 (1537-3978)	\$9337	\$26,171,611	(\$14,350,969–\$37,142,586)
Instrumental deliveries	286 (107-431)	\$711	\$203,346	(\$76,077–\$306,441)
Epidural use	3934 (2246-5616)	\$666.7	\$2,622,797.80	(\$1,497,408.2–\$3,744,187.2)
Total savings			\$28,997,754.80	(\$16,051,723.2–\$41,193,214.2)
Savings per delivery (68,367 ¹⁰)			\$424.14 per delivery	(\$234.78–\$602.53)
Savings per low-risk delivery (54,621)			\$530.89 per delivery	(\$293.87–\$754.16)

from October 2010 through September 2011.¹⁷ The mean hospital charge in Wisconsin for a cesarean delivery was \$16,048. The mean hospital charge for a vaginal delivery was \$7907. The physician fees were taken from Healthcare Blue Book.¹⁸ The physician fee for vaginal birth was \$1634. The physician fee for a cesarean delivery was \$2014 plus an anesthesiologist fee of \$816. There was a cost savings of \$9337 for vaginal deliveries compared to cesarean deliveries. Estimated costs of instrumental deliveries for Wisconsin in 2010 were obtained from the HCUP online query system.¹³ The mean hospital charge for an instrumental delivery was \$8618, or \$711 more than a vaginal delivery.

Epidural analgesia injection costs were estimated at \$325 per patient for intermittent anesthesiologist staffing and \$728 for around-the-clock anesthesiologist staffing in the year 2000.¹⁹ The average of \$526.50 in 2000 yielded the equivalent of \$666.70 in 2010 via the US Department of Labor's Consumer Price Index inflation calculator.²⁰

Based on personal correspondence with practicing doulas in Wisconsin, the cost of doula services ranges from \$200 to \$900 per birth. This fee most often includes 3 prenatal visits and 1 postpartum visit. Each visit lasts about 90 minutes to 2 hours. Labor support begins when the mother desires the doula's presence, usually at the onset of active labor or the initiation of an induction.

Ethics Statement

Institutional review board approval was not needed since this study used only aggregated, unidentifiable published data.

RESULTS

Studies included in the Cochrane Review that utilized the intervention of a professional doula for labor support were applied to eligible Wisconsin births in 2010. There were large estimated reductions in the number of cesarean deliveries, use of regional anesthesia and instrumental deliveries. In addition, there were extensive cost savings related to the lowered rate of these birth outcomes (Table 3).

In-hospital only professional doula labor support for all deliveries had a cost savings of \$424.14 per delivery (95% CI: \$234.78–\$602.53). The cost effectiveness ratios increased in a

linear trend with the cost for hiring professional doula labor support. For instance, the estimated cost effectiveness ratio was 0.47 (95% CI 0.33-0.85) if the cost for hiring professional doula labor support was \$200; it increased to 1.17 (95% CI: 0.83-2.13) if the cost for hiring professional doula labor support increased to \$500, and it further increased to 2.12 (95% CI: 1.49-3.83) with a cost of \$900 for hiring professional doula labor support.

Similarly, in-hospital only professional doula labor support for low-risk deliveries had a cost savings of \$530.89 per delivery (95% CI \$293.87–\$754.16). There was a linear trend in cost effectiveness ratios as the cost for hiring professional doula labor support increased. The estimated cost effectiveness ratio was 0.37 (95% CI: 0.26-0.68) if the cost for hiring professional doula labor support was \$200, and it increased to 0.94 (95% CI: 0.66-1.70) if the cost for hiring professional doula labor support increased to \$500, and it further increased to 1.69 (95% CI: 1.19-3.06) with a cost of \$900 for hiring professional doula labor support.

DISCUSSION

Cost savings were calculated from low-risk deliveries and applied to all births in Wisconsin. This method was used on the basis that every woman may desire doula labor support. However, this assumes that there is no cost benefit of labor support for births that are not full-term, singleton, and low-risk. In addition, the birth outcomes that were not included in this study (Table 1) would likely raise cost savings and patient satisfaction with the birthing experience (and presumably satisfaction with the insurer and hospital). Furthermore, the studies on labor support largely assign a doula after labor has started and professional doulas in Wisconsin often give care before, during, and after labor. No studies have evaluated whether this difference in care increases the effectiveness of professional doula labor support to reduce delivery interventions. If it does have a positive effect on outcomes, then the cost savings would be greater than projected in this study.

Another limitation of this study is that it is time sensitive. Doula labor support and obstetrics care and procedures are constantly improving. However, the Cochrane Review has illustrated the effectiveness of labor support during delivery since its first analysis in 1995, and there are many randomized trials show-

ing the continued effectiveness of doula labor support. Hospital charges, insurance reimbursements, and physician fees also constantly change. This study aims to set a monetary starting point that can be adjusted in the future.

Many statistics used in this study were easily accessible, such as singleton full-term cesarean and vaginal births, singleton full-term vaginal births per ethnicity, and instrumental births. However, other factors such as possible VBAC success, low-risk instrumental births, epidural rates, cesarean deliveries with gestational hypertension, and cesarean deliveries with gestational diabetes were estimates from the literature. To address the problem of having more than one possible source for many of these estimates, they were taken from studies with consideration of 3 criteria: how closely the studies matched Wisconsin's population, how recently the studies were published, and sample size. In addition, high-risk deliveries beyond singleton and full-term births were not accounted for in regional analgesia and instrumental births calculations. This study also assumed that the effect of doula care is the same for vacuum-assisted delivery as for forceps delivery, which has not been studied. Finally, the cost estimates reflect average costs and may vary by location, hospital, and time.

Implementation

A concern for any program that tries to apply evidence-based medicine to real life situations is the disconnection between the sterile research environment and health care settings. However, labor support studies were completed in hospital settings in randomized clinical trials where mothers in labor were given regular care vs labor support.⁵ Furthermore, studies addressed the external validity of labor support by showing similar results within different ethnic and demographic makeups, some of which are similar to Wisconsin.^{5,6} Therefore, it is likely these results would easily translate to Wisconsin health care settings.

Another concern when implementing evidence-based medicine is the differential use of the intervention. Would those women who are less likely to use regional analgesia and less likely to be at risk for a cesarean delivery be more likely to use doula labor support? Would those women who are more likely to use analgesia and have a higher risk of cesarean delivery be less likely to use doula labor support even if it is covered through insurance? These are important questions that would likely depend on the implementation and availability of doula care. If professional doula care were promoted by insurance companies and obstetrical health care providers, then this could greatly increase the use of doula labor support, positive outcomes, and cost savings. Furthermore, the professional doula can have an important role in the era of shared cost savings and health care teams. This would eliminate revenue struggles and keep the focus on overall savings to the health care system.

An important consideration is the cost of doula care in

Wisconsin. Would the insurance reimbursement be enough? If reimbursement is too low then usage also would be very low and the benefits might be hard to identify. Currently, doula care is generally provided in at least 3 separate visits: before birth, during labor, and postpartum. Could hospitals provide staff doulas that provide care to birthing mothers in labor at insurance reimbursement cost for those who cannot pay beyond the reimbursement? Columbia Center in Mequon, Wisconsin has doulas that work for the birthing center, which pays 75% of the \$300 fee for the doula.²¹ Hennepin County Medical Center in Minnesota offers doula labor support from staff doulas at no cost to all birthing patients.²² Providing doula care is an ongoing cost and there are many payment models that exist. These are important considerations for administrators and doulas in Wisconsin. Finally, questions about quality assurance and doula training capacity in Wisconsin must be addressed but are outside the scope of this article.

Clinical Implications and Recommendations

It is recommended that Wisconsin insurers consider reimbursing professional doula labor support, which has an estimated cost savings of \$424.14 per delivery for in-hospital only professional doula labor support without prenatal and postpartum visits (\$530.89 per low-risk delivery). In regard to doula pilot programs, the majority throughout the country have been focused on high-risk groups, minority populations, or the use of nonprofessional volunteer doulas. There are no pilot program results that have been published that could capture the effects of a system-based change of professional doula labor support use in Wisconsin. Thus, it is recommended that a pilot program be designed to assess implementation in Wisconsin. Future studies on the cost-effectiveness of doula labor support should also include quality of life measurements.

CONCLUSION

Professional doula labor support is a safe option to improve birth outcomes and decrease medical expenses for the US health care system. Large-scale insurance coverage for doula labor support has not been implemented in the United States despite a wealth of evidence that supports its benefits. A system-based change in how laboring mothers are supported is very much needed in modern health care. Implementing this change would be an innovative step that would put Wisconsin at the forefront of cost-effective health care: reducing interventions while improving outcomes.

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Patient and Referring Provider Satisfaction with a Gastrointestinal Consultation Clinic for Pregnant Patients

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ABSTRACT

Background: To improve multi-disciplinary care in pregnancy, a gastrointestinal (GI) disorders in pregnancy clinic was created. Patient and referring provider satisfaction with this service was assessed.

Methods: The first 100 patients and their referring providers were surveyed. Survey scores > 3 on a 5-point Likert scale were considered favorable. Descriptive statistics were calculated and open-ended items were analyzed.

Results: Fifty-four percent of patients and 32% of providers returned questionnaires. All satisfaction items received an average patient score of >3.6 and provider score of >4.1, demonstrating overall satisfaction with the clinic. Referring providers were particularly satisfied.

Conclusion: Patients and providers, in particular, report a high level of satisfaction with a GI pregnancy clinic.

for subspecialist input for both pregnancy-unique and non-pregnancy-unique GI conditions such as hyperemesis gravidarum, chronic viral hepatitis, and gastroesophageal reflux disease (GERD) has been demonstrated.⁷ To address the need for high-quality care for women with GI and liver disorders who are considering pregnancy or are currently pregnant, a GI disorders in pregnancy clinic was established in 2008 at the University of Wisconsin (UW) Hospital and Clinics. This paper discusses the creation of the clinic and patient and referring provider satisfaction with this new service.

INTRODUCTION

Normal physiologic changes produce dramatic modifications to the gastrointestinal (GI) tract during pregnancy. Changes occur in small bowel motility,¹ esophageal sphincter pressure,^{1,2} gallbladder contractility,³ and bile composition.⁴ In addition, immunological changes occur that may lead to exacerbations of autoimmune diseases.^{5,6} As a result, GI disorders represent some of the most frequent complaints during pregnancy, with some women experiencing the first occurrence of a disorder and others worsening of pre-existing disease.

Although many GI disorders in pregnancy can be managed effectively by obstetricians and primary care providers, the need

METHODS

Creating the Clinic

Faculty in the gastroenterology division at the UW School of Medicine and Public Health with expertise in treating women's GI health issues assembled in July 2008 to determine the objectives of a GI pregnancy program. Meetings were held concurrently with providers of prenatal and obstetric care within the UW Health system to determine the need and desirability of such a program and its potential offerings. Other gastroenterologists also were queried to determine the conditions for which they would refer to a pregnancy-specific clinic.

Assessing Satisfaction

In September 2010 the first 100 consecutive patients referred to the GI disorders in pregnancy clinic were identified using the Epic Resolute application (Epic Systems Corp, Verona, Wisconsin). Patient and referring providers were mailed a 22-item questionnaire, cover letter, and return envelope. Providers were asked to consider all patients they had referred to the clinic, if more than 1, when completing their surveys. The patient survey included 9 items assessing satisfaction using a 5-point Likert scale (1 = strongly disagree to 5 = strongly agree) and 2 open-ended questions; provider surveys had 10 satisfaction items. Survey scores greater than 3 were considered favorable. The surveys were reviewed by 2 gastroenterologists and a gastroenterology clinical nurse manager for

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Table 1. Respondent Characteristics: Patients

Characteristic	Total No. (%)
Mean patient age + SD (years)	30.3 (4.5)
Median gravida (range)	2 (0-5)
Median parity (range)	1 (0-3)
Referring source	
Primary care provider	13 (24.1)
Obstetrician/gynecologist	17 (31.2)
Gastroenterologist	11 (20.4)
Self-referred	6 (11.1)
Other (eg, midwife, emergency department provider)	3 (3.8)
More than one	4 (7.4)
Main reason for referral	
Worsening of pre-existing GI symptoms in pregnancy	19 (48.1)
New GI symptoms in pregnancy	13 (24.1)
New problem that developed after delivery	1 (1.9)
Prenatal counseling and/or treatment prior to pregnancy	12 (22.2)
More than one	1 (1.9)
Diagnosis^a	
Inflammatory bowel disease	39 (39.0)
Gastroesophageal reflux disease	15 (15.0)
Abdominal pain of unknown etiology	11 (11.0)
Nausea and vomiting of pregnancy/hyperemesis gravidarum	9 (9.0)
Irritable bowel syndrome	6 (6.0)
Hepatitis	6 (6.0)
Constipation	6 (6.0)
Diarrhea	4 (4.0)
Other	6 (6.0)
Number of visits to GI Pregnancy Clinic	
1	19 (35.2)
2 to 5	29 (53.7)
>5	6 (11.1)
Change in diagnosis as a result of consultation (yes)	8 (17.5)
Change in treatment as a result of consultation (yes)	34 (63.0)

^aPercentages do not total 100 as patients could have more than 1 diagnosis
Abbreviations: GI, gastrointestinal.

Table 2. Respondent Characteristics: Providers

Characteristic	Total No. (%)
Field of practice	
Primary care (internal medicine, family practice, other)	7 (35.0)
Obstetrics/gynecology	10 (50.0)
Gastroenterology	3 (15.0)
Other	0
Degree	
MD or DO	17 (85.0)
PA or NP	2 (10.0)
CNM	1 (5.0)
Other	0
Number of referrals to GI pregnancy clinic	
1	2 (10.0)
2 to 5	6 (30.0)
>5	12 (60)

Abbreviations: MD, doctor of medicine; DO, doctor of osteopathy; PA, physician assistant; NP, nurse practitioner; CNM, certified nurse midwife; GI, gastrointestinal.

content validity and by an ambulatory nursing director for face validity. These individuals were chosen based on their experience with patient satisfaction surveys in the outpatient setting.

Patient nonresponders were contacted by phone and given the option of completing the questionnaire by phone or receiving a second mailing. Patients who could not be reached by phone and all provider nonresponders were mailed a second questionnaire.

Descriptive statistics, including means and standard deviations for continuous variables and frequencies for categorical variables, were calculated. Open-ended items were analyzed independently by 2 researchers for positive, negative, or neutral responses.

This study was approved by the Health Sciences Institutional Review Board at the University of Wisconsin-Madison.

RESULTS

Clinic Objectives and Logistics

The following objectives were established for the clinic: (1) to work closely with referring providers to offer individually tailored preconception, intrapartum and post-partum care to women with existing or pregnancy-related GI disorders; (2) to create a training site for the care of pregnant women for medical residents and fellows; and (3) to create a registry of pregnant patients with GI disorders for future research studies.

The clinic began in December 2008. Registered nurses with extensive GI experience triaged all consultation requests to ensure that referrals were appropriate.

Satisfaction Survey

One hundred patients and 62 referring providers were mailed questionnaires. Fifty-four patients (54.0%) and 20 providers (32.3%) completed the surveys. Thirteen patients (24.1%) completed the questionnaire by phone. The remainder of patients (n = 41, 75.9%) and all providers returned it by mail.

Patient and Provider Characteristics

Patient and provider characteristics are summarized in Tables 1 and 2, respectively. The most common reasons for consultation were conditions that are not unique to pregnancy such as inflammatory bowel disease (IBD), GERD, and abdominal pain of unknown etiology. Pregnancy-unique conditions such as nausea and vomiting of pregnancy/hyperemesis gravidarum were present in 20.4% of referrals. Although most patients did not undergo a change in diagnosis as a result of their consultation, 63.0% experienced a change in treatment, most commonly medication initiation or change.

Patient and Provider Satisfaction

Mean patient scores for all 9 satisfaction items were greater than 3.6 (range 3.6 to 4.7) (Table 3). Satisfaction scores were not significantly different between patients who did and who did not experience a change in treatment as a result of the consultation.

Mean provider scores for all 10 satisfaction items were greater than 4.1 (range 4.1-4.8) (Table 3).

Open-ended Feedback

Inter-rater reliability for feedback analysis was 100%. Twenty-two patients (40.7%) provided feedback under “suggested improvements,” of which 13 were considered positive statements, thematically linked by their satisfaction with the expert advice they received regarding their specific GI issues. Eight patients provided suggestions that were coded as negative. These comments included a recommendation for more direct communication between clinic and referring provider, reducing patient wait times, and performing recommended testing in clinic. One patients’ suggestion was considered neutral.

With regard to other open-ended feedback, 10 patients provided positive comments, all of which focused on the quality of care provided by the clinic team. A representative statement was, “I think this is a great clinic [and] much needed while women are pregnant [as] they have lots of unexpected GI symptoms/problems and you need expert advice [on] what is safe to use.” Two patients commented on their experience negatively. One patient cited rudeness of the receptionist and another stated that the visit was unhelpful. The remaining 4 patients left neutral comments, 3 of whom reported feeling fine at the time of their consultation and thus stated the visit was neither helpful nor unhelpful.

Eleven providers (55%) left written feedback. Six comments were positive, 3 were neutral, and 2 were negative. The positive comments addressed the usefulness of having a GI pregnancy clinic within their referral network. One obstetrician commented, “Very positive to have this resource available to our expectant patients. [The clinic is a] great addition to [the] comprehensive care of patients.” Another provider wrote, “As a GI physician ... in central Wisconsin, I feel this will comfort many women during their pregnancy. The most common scenario we come across in this area [is] IBD and pregnancy. I think a second opinion from pregnancy experts [will go a] long way in alleviating some of the fears many women have during their pregnancy.” The negative comments both referred to the limited availability of appointments.

DISCUSSION

Caring for pregnant women who have chronic medical conditions or who develop medical problems during pregnancy can be challenging. Given that traditional teaching in obstetrics and midwifery in the past has concentrated on obstetric matters, a collaborative effort of clinicians from different specialties has

Table 3. Patient and Provider Satisfaction

Statement	Patient Mean Score ^a (SD)	Provider Mean Score ^a (SD)
The consultation helped in the diagnosis of my/my patient(s)’ problem	3.5 (1.3)	4.1 (0.7)
The consultation helped in the treatment of my/my patient(s)’ problem	4.1 (1.2)	4.8 (0.4)
My doctor/I agreed with the diagnosis and treatment plan(s) which resulted from the consultation(s)	4.5 (0.9)	4.7 (0.5)
Considering the treatment(s), explanations and/or education I/my patient(s) received, the consultation improved my/my patient(s)’ quality of life	4.1 (1.2)	4.6 (0.5)
The problem for which I/my patient(s) were referred to the clinic improved as a result of the consultation	3.9 (1.3)	4.6 (0.5)
Overall the consultation(s) was/were helpful	4.6 (0.8)	4.8 (0.4)
I am satisfied with my/my patient(s)’ experience in this clinic	4.6 (0.8)	4.8 (0.4)
I would recommend/refer other patients to this clinic	4.7 (0.7)	4.8 (0.4)
I would recommend this clinic to other providers	N/A	4.8 (0.4)
This clinic is a valuable addition to the offerings at UW Health	4.7 (0.7)	4.8 (0.4)

^aAll items scored with 5-point Likert scale (1=strongly disagree to 5=strongly disagree)

been recommended for women with medical illness in pregnancy in order to achieve the best possible outcomes through the reproductive years.⁸

In 2008 we created a GI disorders in pregnancy clinic to extend the comprehensive care offered to pregnant women in the UW Health system, as GI disorders make up some of the most common medical illnesses in pregnancy. To our knowledge, it is the second of its kind in the country. We found that GI conditions that are not unique to pregnancy were the largest reason for referral. Patients with IBD, in particular, were most frequently seen in this clinic, which likely reflects the expertise of the clinic director (Saha). Among the pregnancy-unique GI conditions, nausea and vomiting of pregnancy/hyperemesis gravidarum was most frequently encountered.

We found a high level of satisfaction with the clinic among patients and referring providers with provider satisfaction being particularly high. Based on written feedback, this may be the result of providers being pleased to have a new resource for complicated patients they would otherwise have either managed on their own or managed without expert advice. The greatest concern for providers was the inability of the program to handle the demand. Patients also were very satisfied. The majority experienced a change in treatment for their GI symptoms after being seen in the clinic and reported that the problem for which they were referred improved as a result of the consultation. Ninety-one percent reported that they would recommend the clinic to other women.

Even patients who did not undergo a change in treatment reported a high level of satisfaction. Some of these patients were seen for prenatal counseling and were not experiencing symptoms at the time of their visit. Written feedback suggests that having a specialist available to discuss the natural history of their disease

during pregnancy and create a plan should the disease become active was reassuring and helpful.

This study may be limited by several factors. First, the response rate for providers was low at 32%. Although this is comparable to the response rate reported in other mail survey studies of physicians,⁹ it raises the possibility of nonresponse bias. In addition, the response rate of 54% for patients raises similar concern. As nonrespondents have in prior studies been shown to have worse health status than the population average and be less satisfied with medical care,¹⁰ the results of this study may not be generalizable to the entire clinic population. Second, recall bias may have influenced the study's results, as the variation of time from consultation to survey completion might have influenced survey responses. Lastly, we have no data regarding satisfaction with pregnant patients referred to gastroenterology prior to the establishment of this clinic. Although our discussion with our referring providers suggested this clinic filled an unmet need within our health care system, we were unable to show that this service provides substantively different or more satisfactory care. Future assessments of the service will include pregnancy outcome measures.

CONCLUSION

We found that patients with new GI symptoms or established GI disorders during pregnancy are satisfied with a GI pregnancy specialist consultation. Providers also are highly satisfied with the service. Given the prevalence of GI disorders during pregnancy, academic gastroenterology programs should consider building expertise in this area and offering specialized service to pregnant patients.

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Hemodynamic Optimization in High-Risk Mitral Valve Repairs as a Key Component in Surgical Readiness

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ABSTRACT

Ensuring optimal readiness for surgery using a preoperative checklist has been shown to reduce perioperative morbidity and mortality in both elective and urgent surgeries. We recently introduced hemodynamic optimization as part of our preoperative preparedness strategy for cardiothoracic surgery. Here we describe the case of a patient with severe mitral regurgitation and suboptimal hemodynamics that was optimized preoperatively with nesiritide to reduce pulmonary hypertension. Postoperatively, the patient had an improvement of his heart failure from New York Heart Association functional class 3 to class 1. Without hemodynamic optimization the patient may have been considered too high-risk to undergo mitral valve repair. This case report illustrates the importance of a systemic approach with high-risk surgery, and the use of strategies that optimize key patient factors, including hemodynamics, prior to all elective and urgent procedures.

INTRODUCTION

Recognizing the value of surgery as an integral part of global health care and the need for standardized surgical practices to avoid unnecessary surgical complications, the World Health Organization¹ adopted a 19-item surgical safety checklist in 2009. The Safe Surgery Saves Lives Study Group² published data suggesting that implementation of this surgical safety checklist reduced the rates of deaths and complications in patients undergoing noncardiac surgery. Building on the idea of a general surgical readiness checklist, the Northern New England Cardiovascular Disease Study Group³ bundled several evidence-based, preoperative strategies shown to improve outcomes for patients following coronary artery bypass surgery (CABG) and demonstrated that implementation of these strategies reduced time to extubation and length of

stay after surgery.⁴ Prior to cardiothoracic surgery, we use a “Readiness for Cardiac Surgery Checklist” based on the checklist developed by the Northern New England Cardiovascular Disease Study Group’s checklist for CABG³ to optimize a patient’s surgical experience during any cardiac surgery (Table 1). Unique to our readiness checklist is the preoperative hemodynamic optimization bundle with a particular emphasis on pulmonary hypertension.

Patients with pulmonary hypertension undergoing mitral valve surgery are at increased risk for perioperative mortality.⁵⁻⁷ As a result, strategies for effectively reducing pulmonary hypertension in the perioperative period have been developed. The majority of studies have focused on postoperative management with pulmonary vasodilators, but several preoperative interventions also have been explored, including inhaled nitric oxide and prostacyclin, phosphodiesterase inhibitors, and recombinant brain natriuretic peptide (BNP).⁸ The addition of hemodynamic optimization to the cardiac surgery readiness checklist makes surgery safer for another group of patients that might otherwise be considered ineligible. Here we present a case of mitral valve repair (MVR) in a patient with pulmonary hypertension that clearly illustrates the value of preoperative hemodynamic optimization.

CASE PRESENTATION

A 72-year-old man who was followed in the heart failure clinic with stable heart failure symptoms developed progressive shortness of breath and dyspnea on exertion. These changes were associated with a marked reduction in exercise capacity over the previous 7 weeks. The patient also complained of fatigue, orthopnea, paroxysmal nocturnal dyspnea, new onset of bilateral lower extremity edema, ascites, and weight gain. His medical history was remarkable for an inferior wall myocardial infarction in 1987 with subsequent quadruple CABG surgery in 1993. At the time of surgery, bilateral internal thoracic arteries were used and configured as a composite Y-graft. The left internal mammary artery was left in situ and anastomosed as a sequential graft to the diago-

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Table 1. Readiness for Cardiac Surgery Checklist

Expectations	Yes	Fill in the interventions to be addressed	Initials
Patient taking ASA		Contraindicated:	
Creatinine at baseline: _____		Cr: _____ eGFR: _____	
Fasting glucose <125 mg/dl		Orders to address	
In diabetic patients, HgB A1c: _____			
Off clopidogrel 5 days		5th day: _____ (date)	
Greater than 3 days status post MI unless chest pain persists		MI: _____ (date)	
Hematocrit (HCT) greater than 30		Assess transfusion consent and GI consult for active bleeding or note contraindication: <30, epigen 500 units/kg, 200 mg IV iron- day before	
TRUST score: _____			
HR less than 80		Beta block orders to address	
LV function status assessed		EF% ____ by Echo. LV Gram or nuclear test (circle one)	
Eptifibatide off for 6 hours prior to surgery		Ordered off at: _____	
Carotid Doppler for history of PAD, bruit, or aortic stenosis if not done in last 6 months		<input type="checkbox"/> N/A ordered for: _____ last done: _____	
Vein mapping with marking if history of vein stripping, severe PAD, conduit assessment		<input type="checkbox"/> N/A	
Pre-op statins		Reason contraindicated:	
Lipitor 40-80 mg every day			
Bilateral arm blood pressure		Right: _____ Left: _____	
Pulmonary assessment		<input type="checkbox"/> N/A PFT ordered, smoking cessation. FEV-1 ____	
Chest pain free and/or no ECG changes		Notify both: Cardiologist Cardiac Surgeon	
Patient CHF managed BNP less than 500		<input type="checkbox"/> N/A	
Assess risk score on page 2 ^a			
Standard prep			
Preoperative hemodynamic optimization			

^aRefers to the Society for Thoracic Surgeons (STS) risk score available at <http://riskcalc.sts.org/STSWebRiskCalc273/>. Accessed March 11, 2013.

Abbreviations = ASA, aspirin (aceylated salicylate); Cr, creatinine; eGFR, estimated glomerular filtration rate; HgB A1c, hemoglobin A1c; MI, myocardial infarction; GI, gastroenterology; TRUST, transfusion risk understanding scoring tool; IV, intravenous; LV, left ventricular; EF, ejection fraction; Echo, echocardiogram; PAD, peripheral arterial disease; N/A, not applicable; PFT, pulmonary function test; FEV-1, force expiratory volume in 1 second; EKG, electrocardiogram; CHF, congestive heart failure; BNP, brain natriuretic peptide.

nal branch of the left anterior descending artery and to the left anterior descending artery proper. The right internal mammary artery was constructed as a Y-graft from the left internal mammary as a sequence graft to the obtuse marginal and the right posterior descending artery.

In 2010, the patient presented with a syncope episode and a non-ST elevation myocardial infarction. He underwent a left heart catheterization and was found to have a total occlusion of the right coronary artery, yet both left and right internal mammary arteries were patent. A dual-chamber pacemaker was placed for an intermittent high-grade atrioventricular block. The patient had ischemic cardiomyopathy with an ejection fraction (EF) of 30%. He underwent right thoracocentesis on multiple occasions for transudative effusion. Additional relevant medi-

cal issues included hypertension, type 2 diabetes mellitus, stage 3 chronic kidney disease, and benign prostatic hyperplasia. The patient's social history was remarkable for a remote history of tobacco use (quit 18 years prior) and a history of alcohol abuse. Medications at presentation included amlodipine (10 mg), aspirin (325 mg), diltiazem SR (180 mg), and furosemide (80 mg 3 times daily).

On examination, the patient was afebrile with a regular pulse of 79 beats per minute (BPM), respiratory rate of 24 breaths per minute, and blood pressure of 114/65 mmHg. Oxygen saturation was 84% on 3 liters nasal cannula. No jugular venous distention was noted. Cardiac exam revealed regular rhythm and the presence of 3/6 systolic murmur best heard at the left lower sternal border. Right basal lung sounds were decreased and bilateral lower extremity pitting edema was present. Other exams were unremarkable. A chest radiograph showed bilateral pleural effusion greater on the right than the left (Figure 1). An electrocardiogram (ECG) showed sinus rhythm at 80 bpm with a first degree atrioventricular (AV) block. On echocardiogram the patient's left ventricle and left and right atrium were enlarged (Figure 2A). His EF was 30%, left atrium diameter was 5.2 cm, left atrium volume/body surface area was 54 ml/m², and left ventricular end-diastolic diameter (LVED) was 5.8 cm. Mitral regurgitation (MR) was present, and quantitative analy-

sis of MR with grading assessed by the proximal isovelocity surface area (PISA) and 2D volumetric methods showed severe MR based on standard criteria.⁹ By the PISA method, regurgitant volume (RV) was 91 ml, regurgitant orifice area (ROA) was 57 mm², and regurgitant fraction (RF) was 54%. Similarly, by the 2D volumetric method, RV was 127 ml, ROA was 74 mm², and RF was 79%. Transesophageal echocardiogram revealed severe MR type 1 or central regurgitation. Laboratory values were as follows: hematocrit 31.4%, sodium 137 mmol/L, potassium 3.5 mmol/L, urea 30 mg/dL, creatinine 1.8 mg/dL, and BNP 2260 pg/mL. The patient underwent cardiac catheterization, and the findings were consistent with pulmonary hypertension and patent bypass grafts. He was referred for possible surgery.

The cardiothoracic surgeon deemed the patient to be a high-

risk candidate for MVR. His Society of Thoracic Surgeons (STS) risk for mortality was 9.6%,¹⁰ and his EuroScore perioperative mortality risk was 14.86%.¹¹ The decision was made to mitigate his perioperative hemodynamics with a series of strategies to optimize surgical readiness (see checklist, Table 1). He was admitted to an intensive care setting, and a pulmonary artery catheter was placed in order to effectively treat his pulmonary hypertension with the goal of reducing pulmonary artery pressures from near systemic to two-thirds systemic. A nesiritide infusion was started using the infusion protocol described and justified by Salzberg et al.⁸ The patient received a loading dose of nesiritide of 2 µg/kg over 30 minutes and then 0.01-0.02 µg/kg/min for 42 hours before surgery. Nesiritide administration was continued postoperatively at 0.01 µg/kg/min for 24 hours. His pulmonary artery pressures successfully dropped from a systolic pressure of 70 mmHg to 45 mmHg, meeting the aforementioned goal. Other hemodynamic parameters measured by pulmonary artery catheter and values at baseline, during the nesiritide drip, and at the end of the nesiritide drip (postoperative stage) are shown in Table 2. Following the nesiritide drip, the patient's STS score decreased from 9.6% to 6.3%, and his EuroScore decreased from 14.86% to 10.16%, indicating a reduction in mortality risk. During the preoperative period of nesiritide infusion, the patient had a total fluid intake of 2371 ml and output of 3245 ml. Once optimized, the patient underwent surgery.

A re-do sternotomy was performed without difficulty, and an intraoperative echocardiogram confirmed severe MR with a central jet and EF of 30%. The patient's mitral leaflets were normal on direct inspection of the valve, and the mitral annulus was symmetrically dilated. MVR was performed using a 28 mm Life Science GeoForm ring. A postoperative transesophageal echocardiogram showed no MR and no gradient across the valve (Figure 2B). The postoperative course was uneventful. The patient's home dose of furosemide 80 mg 3 times per day was continued at discharge with the addition of spironolactone 25 mg once per day. On postoperative day 8, the patient was discharged home without oxygen. By the fourth week, the patient had lost more than 30 pounds and was able to perform his usual daily living activities without shortness of breath.

DISCUSSION

Mitral valve regurgitation is classified into 3 functional types according to leaflet mobility. Type I MR has normal leaflet

Figure 1. Pre- and Post-surgical Chest Radiograph.

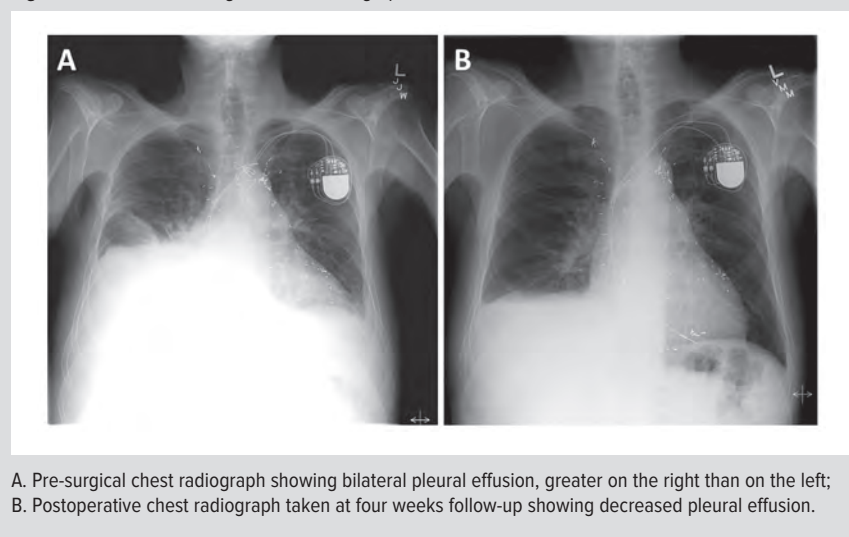
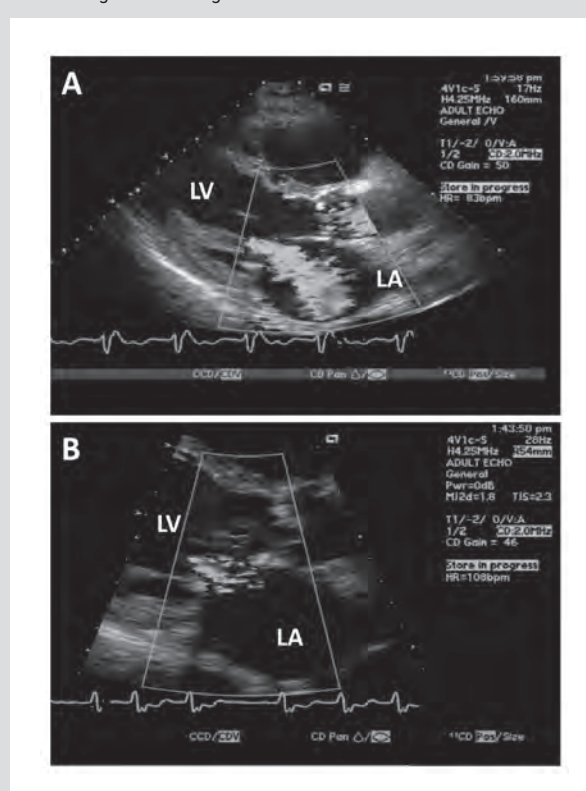


Figure 2. Transthoracic Echocardiogram, Parasternal Long Axis View Echocardiograms Showing Left Ventricle and Left Atrium.



motion, type II is characterized by excessive leaflet motion, and type III by restricted leaflet motion.¹² The patient described here had severe type 1 MR caused by a dilated ventricle resulting in symmetrically dilated mitral valve annulus. The papillary muscles were displaced inferior and laterally resulting in central

Table 2. Hemodynamic Changes During Treatment With Nesiritide

	Baseline	During Nesiritide Drip	Postoperative ^a
PAS (mmHg)	70	45	42
PAD (mmHg)	30	37	27
CVP (mmHg)	20	8	10
CO (L/min)	4.4	5.2	7.5
CI (L/min/m ²)	2.2	2.6	3.7
SVR (dynes·s/cm ⁵)	1144	1040	597
HR (bpm)	75	80	90

^aFollowing nesiritide drip in the postoperative stage, patient was on an epinephrine, norepinephrine, and dopamine drip (0.06 µg/kg/min norepinephrine, 0.03 µg/kg/min dopamine)

Abbreviations: PAS, pulmonary artery systolic pressure; PAD, pulmonary artery diastolic pressure; CVP, central venous pressure; CO, cardiac output; CI, cardiac index; SVR, systemic vascular resistance; HR, heart rate

severe MR. A successful MVR was performed using a Geoform ring, which remodeled the mitral valve annulus and provided a medium that helped to change the shape of annulus, thereby bringing the papillary muscles medial and superior. This patient's severe pulmonary hypertension in addition to other factors put him at particularly high risk for perioperative morbidity or mortality. The addition of hemodynamic optimization to our cardiac surgery readiness checklist and preoperative mitigation of his pulmonary hypertension made MVR surgery safer for this patient, who might otherwise be considered ineligible, although he clearly benefited from the procedure.

Nesiritide is a recombinant BNP that was FDA approved in 2001 for the treatment of heart failure.¹³⁻¹⁵ Increased cardiac filling pressure is a potent stimulus for BNP release,^{16,17} which then binds to the guanylate cyclase receptor on vascular smooth muscle and endothelial cells, increasing intracellular cyclic GMP and resulting in smooth muscle cell relaxation. The potent vasodilator effect of BNP results in a decrease in right and left ventricular filling pressures with a decrease in pulmonary capillary wedge pressure and an increase in cardiac output and diuresis without a reflex tachycardia.^{13,14} Nesiritide use is controversial in the outpatient setting, where post market analysis has shown it to be associated with a higher mortality rate.¹⁸ We have used nesiritide in the preoperative inpatient setting without incident.

As demonstrated by the Northern New England Cardiovascular Disease Study Group,^{3,4} preoperative optimization of patient factors is important for surgical readiness and improved perioperative outcomes in cardiac surgery patients. Our patient was hemodynamically optimized preoperatively with nesiritide infusion. Hemodynamic optimization with nesiritide has been described in high-risk mitral valve repair/replacement patients.⁸ The authors have adopted the above-described strategy to improve hemodynamic factors as a part of our bundle for preoperative readiness for cardiac surgery in elective and urgent patients. The above-described patient underwent MVR without

any perioperative complications. Postoperatively, the patient's symptoms and exercise tolerance improved significantly, potentially due to correction of the underlying problem.

CONCLUSION

A systemic approach to preoperative patient optimization with a checklist and bundles is important for managing the perioperative risks of surgical procedures. Modifying patient factors prior to surgery can result in improved outcomes for high-risk patients as demonstrated in the case presented here where pulmonary hypertension was managed with perioperative nesiritide infusion to safely decrease pulmonary hypertension. Hemodynamic optimization is a key component for cardiothoracic surgical readiness.

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The Wisconsin Chapter of the American College of Physicians held its annual meeting in Wisconsin Dells, September 9-11, 2012. Internal medicine residents from each of Wisconsin's 5 residency programs presented their research and/or unusual clinical experiences via posters and vignettes. Vignettes will be published in the next issue of *WMJ*.

DISPLAYED POSTERS

Assessing Efficacy of Nonpharmacological Intervention in Older

Hospitalized Patients with Insomnia

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Introduction: Insomnia is a common problem affecting 67% of hospitalized seniors. In acute care settings, 60% of patients receive sedative hypnotic drugs for insomnia. Sedatives may have serious adverse effects on older adults. Many nonpharmacologic sleep interventions have been proposed but have not been researched in depth. The goal of our study was to evaluate the efficacy of nonpharmacological vs pharmacological interventions for seniors hospitalized with insomnia.

Methods: We conducted a quality improvement project with 2 phases to adults 60 years and older admitted to our facility November 2009 to March 2011. We included all patients with acute sleeping problems without prior diagnosis of insomnia or patients with chronic insomnia on pharmacological treatment. Excluded were those who required frequent nursing monitoring due to acute illness, severe pain/immediate postoperative, unable to follow direction due to delirium/dementia, anosmia, hearing deficits, sleep apnea on continuous positive airway pressure (CPAP), allergies to lavender oils. After the completion of phase I with hypnotics/sedatives as standard of care, the second phase used the

sleep kit, which included massage therapy, lavender aromatherapy, relaxing music, and warm blankets as standard. Outcome measurements were total hours and quality of sleep, wake-times after sleep onset, feeling after waking up in the morning. T-test, chi-square or Fisher exact test was used for univariate analysis. Multivariable logistic regression analysis was used to assess the association between sleep quality (good/very good/excellent vs fair/poor) and sleep intervention (sleep kit vs sleep aid), while adjusting for age, sex, race, difficulties falling asleep, difficulties staying asleep, duration of sleep problem, and nap during the day. Statistical analyses were performed using SAS 9.2.

Results: There were 64 patients in the sleep aid group and 60 patients in the sleep kit group, and no significant difference in age, sex, race, and sleeping problems between the groups. Following intervention, there was no difference in hours of sleep, however the group with sleep kit (78%) reported significantly better quality of sleep than the sleep aid group (52%). Multivariable regression showed that sleep kit was significantly associated with better quality of sleep (adjusted odds ratio [OR]=3.72, $P=0.002$).

Conclusion: By using the nonpharmacological intervention, patients can improve their sleep quality while avoiding adverse effects from using sleep aids and thus enhance rest and sleep in hospital setting.

Infective Endocarditis Caused by *Cardiobacterium Valvarum*

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Introduction: *Cardiobacterium valvarum* is a newly recognized human pathogen related to infective endocarditis (IE). However, *cardiobacterium* species are rarely the etiology of IE. We present a case of *C valvarum* IE affecting normal native mitral valve in a patient with no history of recent dental procedure that was detected by broad-range polymerase chain reaction (PCR) and 16S rRNA gene sequencing.

Case: A 49-year-old woman presented with sudden loss of left central vision, 2 months history of fever, chills, and 40-lb weight loss. Temperature was 102°F, blood pressure (BP) 110/80, heart rate (HR) 72. Physical examination showed loss of left central visual field, with signs of retinal artery occlusion, and 2/6 systolic murmur at the apex. Laboratory data showed hemoglobin (Hb) 9.2, white blood cell (WBC) 7.1, C-reactive protein (CRP) 7.6 mg/dl. Transthoracic echocardiography (TTE) revealed vegetations on the posterior mitral valve (MV) leaflet. Blood culture grew *C valvarum* that was confirmed by 16S rRNA gene sequencing. Patient was treated with ceftriaxone for 4 weeks. A month later, he presented with severe right leg pain, fever 100.3°F, night sweats, orthopnea, and dyspnea on exertion (DOE). Exam showed 3/6 holosystolic murmur. The right calf was tender, with absence of dorsalis pedis pulse. Computed tomographic (CT) angiogram showed intraluminal filling defects within the right common femoral artery. Patient underwent MV replacement and right femoral endarterectomy with a vein patch angioplasty. Patient was continued on ceftriaxone and did well postoperatively.

Discussion: *C. valvarum* is a newly proposed species and, like *Cardiobacterium hominis*, is a rare cause of endocarditis. Since 2004, a total of 9 cases of *C. valvarum* IE have been described, including this case. Like other cases, the onset in our case was insidious, with low-grade fever and extensive valve tissue destruction. Four cases had bicuspid aortic valve (AV), one had tricuspid AV, one case had moderate MV prolapse and mitral insufficiency, and one case had bioprosthetic AV. Our case had a completely normal MV with no history of recent dental procedure, and no further focus was known. Two of the 9 cases were complicated by neurological events: subarachnoid hemorrhage secondary to a mycotic aneurysm and ischemic stroke. Our patient had extensive retinal artery and common femoral artery distal embolization. All patients were treated with β -lactam antibiotics. In summary, this is the 9th case of IE due to *C. valvarum*, the 3rd case affecting a native valve, the 2nd case affecting a low jet flow valve, and it is the 1st case affecting completely normal native valve with extensive septic distal embolization. Further advances and widespread use of molecular techniques will likely reveal more cases. Physicians should be aware that *C. valvarum* is a potential agent of IE.

Spontaneous Rectal Perforation Presenting as Necrotizing Fasciitis of the Lower Limb

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Introduction: Necrotizing fasciitis (NF) is a life-threatening soft tissue infection associated with high mortality and morbidity. We report an unusual case of lateral lower limb NF following spontaneous rectal perforation.

Case: A 68-year-old man with a history of type 2 diabetes mellitus (DM), T3 N2 M0 colorectal carcinoma S/P rectal resection, colorectal anastomosis, and chemotherapy presented with a 2-week history of left-sided back pain radiating to his hip, for which he was started on steroids. He had a number of small-bowel obstructions and underwent enterolysis along with abdominal washout. He also had fever and chills, then became lethargic and confused. At presentation, he

was toxic looking, afebrile, and hypotensive. Physical examination revealed left hip swelling, tenderness, and subcutaneous crepitus of the left lower limb. Initial labs revealed WBC of 30,700 with left-sided shift. His creatinine was up to 2.50. Lumbar spine magnetic resonance imaging (MRI) showed no evidence of diskitis. CT scan of the abdomen and pelvis showed rectal perforation with large amount of gas extending from the rectum laterally to the left, following muscle planes and bundles down into the left upper leg. Patient underwent sigmoid resection with end colostomy, decompression fasciotomy of the left thigh, and irrigation and debridement of a deep thigh abscess that required further wound debridements. He was started on broad-spectrum intravenous (IV) antibiotics and hyperbaric oxygen treatment. Wound cultures grew *E. coli* along with *Pseudomonas* and *Bacteroides*. The patient improved significantly.

Discussion: NF is a rare complication associated with colorectal malignancy. The majority of cases reported involve spontaneous perforation due to colorectal malignancy, with infection limited to the perineum, such as Fournier's gangrene, with the exception of 2 cases that presented as direct tumor invasion into the abdominal wall, and psoas abscess. There was 1 case of NF of the lower limb following traumatic rectal perforation in a patient with history of rectal cancer 5 years prior to the presentation. Our case appears unique, as we found no case reports of a bowel perforation causing NF of the lateral lower limb following spontaneous rectal perforation in a patient with history of treated colorectal cancer with no evidence of cancer recurrence, direct trauma, abdominal signs at presentation, or preceding changes in bowel habit. With regard to its etiology, we postulate systemic immunosuppression secondary to the cumulative effects of steroids therapy and DM in our patient. Treatment involves the use of high-dose antibiotic therapy, systemic support, and prompt and radical surgical debridement of the infected tissues. High index of suspicion and prompt surgical intervention are the cornerstone of treatment for improving the disease outcome.

Patient with Cystic Fibrosis-related Diabetes Presenting with Diabetic Myonecrosis

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Case: This case involves a 32-year-old white man with cystic fibrosis (CF) and 2 delta F508 mutations, status post lung transplant in 2008 and uncontrolled cystic fibrosis related diabetes mellitus (CFRD). He presented with worsening pain in the left thigh and calf of 3-week duration, of a magnitude preempting the leg from bearing weight. While examination of the right lower extremity was nontender and showed a normal range of motion, the patient was unable to flex the left lower extremity beyond 20 degrees. The posterior thigh and the calf were mildly tender to palpation. Erythema, swelling, and deformity were absent. Pulses were positive bilaterally. MRI showed extensive edema with inflammatory changes involving several muscles in the left thigh and calf on T1-weighted images suggestive of diabetic myonecrosis. Treatment included complete bed rest, analgesics, antiplatelet agent, aspirin 81 mg/day, and adequate glycemic control. The patient's condition improved and he was discharged 3 days later.

Discussion: Diabetic myonecrosis is an uncommon complication of poorly controlled diabetes mellitus. Numerous case reports/case series have been reported, but to our knowledge this is the 2nd published case of a CFRD patient presenting with diabetic myonecrosis. The only other report we found was a very recent abstract of a case presented by Dopp et al, "Sugar Pains: Novel Diabetic Myonecrosis in a Cystic Fibrosis Patient." CFRD is a common comorbidity in patients with CF, with prevalence in adult patients as high as 50%, increasing with age. Typical clinical presentation of diabetic myonecrosis consists of sudden onset of pain in the affected muscle, in association with swelling with thigh muscles most commonly affected, followed by calf muscles. The imaging modality of choice for soft tissue evaluation is MRI, which shows findings of increased signal on T2 imaging in patients presenting with edema. Diabetic myonecrosis is normally self-limiting, and

good glycemic control with supportive care is the mainstay of treatment. A high index of suspicion for diabetic myonecrosis should accompany a patient with CFRD presenting with lower extremity pain.

Unusual Presentation of a Rare Cancer: Histiocytic Sarcoma in the Brain 16 Years After Treatment for Acute Lymphoblastic Leukemia

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Case: A 44-year-old man presented to the emergency department (ED) with unsteadiness, loss of balance, and left-sided weakness for 2 weeks. A CT scan and MRI of the brain showed 2 intra-axial enhancing lesions: one in the corpus callosum of 3.5 cm and the other adjacent to the right lateral ventricle of 2.6 cm, both associated with vasogenic edema. The patient's history was significant for T lymphoblastic leukemia (T-ALL) with cerebrospinal fluid (CSF) involvement 16 years prior for which he received craniospinal irradiation and intrathecal chemotherapy for approximately 26 months with no additional significant health problems. A biopsy of the right parietal brain tumor was done. The final pathological diagnosis was atypical histiocytic infiltrate consistent with histiocytic sarcoma (HS). The patient was treated with 2 doses of intravenous methotrexate, but his condition continued to decline. He then was treated with 2600 cGy of whole brain radiotherapy, with an additional 2000 cGy boost to each lesion. He received temozolomide 150 mg/m² for 5 consecutive days during the whole brain radiotherapy. As there was no improvement clinically, he declined further treatment. He was enrolled in hospice and died 4 months later, 27 weeks after initial presentation.

Discussion: Histiocytic sarcoma is a very rare hematopoietic neoplasm that has been reported in association with other hematological malignancies. Presentation of HS in the central nervous system (CNS) is even less common. Diagnosis of HS requires the presence of histiocytic markers and the systematic

exclusion of markers of other cell lineages. Primary HS CNS tumors are aggressive and generally have poor outcomes. There are no standard treatment guidelines due to lack of clinical trials and a limited number of case reports.

Flaccid Paralysis of Lower Extremities in a Young Man with Uncontrolled Hyperthyroidism

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Introduction: Thyrotoxic periodic paralysis (TPP) is a syndrome of episodic muscle weakness accompanied by severe hypokalemia in patients with uncontrolled hyperthyroidism. It occurs more commonly in Asian males with hyperthyroidism in their 3rd or 4th decades. Early recognition is crucial for providing life-saving therapy.

Case: A 31-year-old Hmong man with a history of hyperthyroidism presented with an acute onset of bilateral leg weakness for 1 day. Over the previous 3 to 4 months, he lost 75 pounds and had frequent night sweats, diarrhea, and heat intolerance. He was prescribed, but had not been taking, methimazole since the diagnosis with hyperthyroidism 4 months prior. For 2 weeks he had intermittent painful bilateral leg cramps for which he took ibuprofen. He also reported going through a recent divorce and had a high carbohydrate meal for dinner 1 day prior to admission. On exam, he was alert and oriented. He had no lid lag, proptosis, or periorbital edema. His thyroid exam was unremarkable. His neurological exam was notable for fine bilateral hand tremor and marked weakness in his lower extremities (1/5) with decreased reflexes despite normal sensation. His labs were significant for a potassium of 1.2 mg/dl, magnesium 1.5 mg/dl, phosphorus 1.1 mg/dl, thyrotropin (TSH) 0.02 uIU/ml, free thyroxine (T4) of 3.6 ug/dl, and free triiodothyronine (T3) of 8.9 ug/dl. He was admitted to the intensive care unit (ICU), and his electrolytes were aggressively supplemented. Following their normalization, his leg weakness improved. Subsequent thyroid ultrasound showed a diffusely heterogeneous and hypervascular thyroid, and thyroid-stimulating immunoglobulins were severely elevated.

These were consistent with Grave's disease. A diagnosis of thyrotoxic periodic paralysis was made, and he was given methimazole along with propranolol. He was subsequently discharged home on day 3 of admission.

Discussion: TPP typically is triggered by a large carbohydrate meal, stress, or vigorous exercise. The underlying cause appears to be the increased activation of the Na/K-ATPase pump by excessive thyroid hormone, leading to an influx of potassium into the intracellular space. Thyroid hormone also enhances beta adrenergic receptors on muscle cells to stimulate Na/K-ATPase pump. TPP is diagnosed by the presentation of flaccid paralysis, hypokalemia, suppressed TSH, and elevated T4 and T3 levels. It is important to distinguish TPP from FHPP (familial hypokalemic periodic paralysis) because TPP is responsive to treatment with oral/IV propranolol. Screening with TSH therefore can be very helpful for patients presenting with hypokalemia and paralysis. Since hypokalemia is due to an intracellular shift of K ions rather than from actual loss, the hypokalemic state is only transient and often can resolve without interventions. However, life-threatening arrhythmia as well as respiratory failure from muscle weakness has been reported due to severe hypokalemia. In addition, 1 study showed that giving IV potassium shortened the recovery time by about half. There were studies that showed efficacy of using a large dose of oral or IV propranolol in treatment of TPP. If potassium is given, there often is a risk of rebound hyperkalemia, which has not been observed in the treatment with propranolol alone. Propranolol also has been found to decrease future attacks of TPP; however, the most effective prevention for recurrence lies in the treatment of the underlying hyperthyroidism.

Coronary-Pulmonary Fistula: Case Report and Brief Review of Existing Literature

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Case: An 86-year-old woman with history of hypertension, diabetes, hyperlipidemia, and coronary artery disease, admitted with

new-onset congestive heart failure and pneumonia, underwent cardiac catheterization after suffering a non-ST elevation myocardial infarction. Coronary angiogram revealed severe coronary artery disease and coronary pulmonary fistulas involving proximal right coronary artery and a branch from left main coronary artery. Coronary artery fistulas are abnormal communication between one or more coronary arteries and great vessels or a cardiac chamber. We reviewed 15 cases of coronary-pulmonary fistulas published in PUBMED and studied the clinical features of coronary artery fistulas.

Scientific Evidence Underlying National Comprehensive Cancer Network Guidelines for Supportive Care

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Introduction: The National Comprehensive Cancer Network's (NCCN) guidelines for supportive care of cancer patients have not been systematically investigated. Our objective was to describe the distribution of categories of evidence and consensus (EC) among the 10 available supportive care guidelines with regards to screening, treatment, and follow-up.

Methods: We obtained the latest versions (January 18, 2011) of relevant supportive care guidelines from the NCCN website (www.nccn.org). The definitions for various categories of EC used by NCCN panel members were as follows: Category 1 (high level evidence such as randomized controlled trials with uniform consensus), Category 2A (lower level of evidence with uniform consensus), Category 2B (lower level of evidence without a uniform consensus but with no major disagreement), and Category 3 (any level of evidence but with major disagreement).

Results: 680 guidelines were available (140 for screening, 394 for treatment, 146 for follow-up). The proportions of category I, IIA, and IIB were 5%, 92%, and 3% respectively. Guidelines with the most category I recommendations were cancer-related infection (14%) and cancer-related fatigue (12%), antiemesis (7%), venous thromboembolism

(4%), and distress management (2%). Nine percent of all therapeutic recommendations were category I and were found in prevention and treatment of cancer-related infections (63%), myeloid growth factors (11%), venous thromboembolism (8%), antiemesis (6%), cancer-related fatigue (6%), and distress management (6%). Category I guidelines were not available for palliative care, senior adult oncology, cancer and chemotherapy-induced anemia, and adult cancer pain. Category I guidelines also were not available for screening or follow-up.

Conclusion: Almost all of the NCCN supportive care guidelines are based on lower level of evidence but with uniform expert consensus. Huge opportunity exists for research to make recommended guidelines more evidence-based.

Attitudes and Behavior of Nursing Staff Towards In-patient Rounding by Hospitalists

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Introduction: Hospital medicine by design necessitates interaction of providers with nursing staff, specialists, and other clinical and nonclinical staff on a daily basis to care for patients with complex medical conditions.

Methods: Attitudes and satisfaction of nursing staff towards hospitalist rounding and communication were assessed before and 3 months after implementing a "patient-centered inpatient rounding" model.

Results: Three months after implementation, we noticed significant improvement in staff attitudes and behaviors. Compared with 3% satisfaction before implementing the model, 42% of staff were "completely satisfied" by our rounding; 44% reported improved communication with hospitalist staff vs 6.5% before implementation; 57% indicated they felt valued as a health care team member and their job satisfaction improved to 63%; and 53% reported a positive impact on their workflow.

Discussion: Patient-centered inpatient rounding is a great way to take patient care to the patient's bedside. It improves communication

between physicians and nurses, a cornerstone of cost-effective and safe patient care.

Senior Care in the ED:

A Qualitative Study

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Introduction: Senior citizens account for 11.7 million US emergency department visits annually; however, EDs are not equipped to adequately and safely care for seniors. Potential hazards in the ED and atypical presentation of illnesses make assessments and care more challenging. We sought to learn the difficulties and unmet needs of seniors in the ED to inform improved care at community hospitals.

Methods: Qualitative exploration using semi-structured key informant interviews (n=39) and "shadowing" observation sessions (n=32). Analysis of written text was performed using an editing style format by 2 authors who did not participate in data collection.

Setting: One central city and 1 suburban community hospital ED in Southeastern Wisconsin.

Subjects: Fourteen physicians, 8 physician assistants, 14 registered nurses, 2 emergency medical technicians, and incidentally encountered patients.

Results: Frustrations, challenges, and opportunities expressed by interviewees (227 listed items) were separated into 3 broad themes: complexity of the geriatric ED patient (98 items), including difficulty obtaining the history and its complexity and impaired patient communication; issues of patient flow and transitions (70 items), including perceived need for more and specialized staff, inadequate triage and barriers to disposition, system inefficiencies, and lack of continuity and access to care; and need for a senior-friendly environment (59 items) including physical space and amenities, and safety issues.

Observations (115 listed items) revealed positive (22) and negative (23) staff behaviors involving courtesy and safety, system innovations (4) and inefficiencies (28), unpleasant environmental features (29), and positive attributes of patients and families (9).

Conclusions: Our synthesis of staff interviews and observations suggest numerous opportunities to improve care of seniors in EDs. Quality improvement programs should focus on system efficiencies, use of nonclinical staff, environmental modifications, and improved communication during transitions.

Advancing Bedside Procedure Education with a Simulator-Based Workshop

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Introduction: In 2007 the American Medical Association (AMA) repeated its 1986 survey of numbers and types of procedures performed by general internists, which clearly demonstrated a decreasing trend in general internists' procedural practice. The American Board of Internal Medicine (ABIM) no longer requires proficiency in the majority of bedside procedures. Contrary to these regulatory changes, many ICUs are run by hospitalists and general internists. In the ICU setting, clinicians must be capable of performing many bedside procedures; so improving internal medicine training in this area is essential.

Methods: Fifty-two first-year internal medicine residents went through a 1-day, simulator-based procedure workshop. Bedside procedures included arterial line placement, central venous line (CVL) placement, lumbar puncture (LP), arthrocentesis, and advanced cardiac life support (ACLS) leadership training. Residents completed a survey of their confidence performing these procedures on a 10-point scale: 1 being least confident, 10 being most confident. Given the small sample size ($N=52$), we were unable to assume the probability distributions of variables. Hence, significance was calculated using Wilcoxon signed-ranked test.

Results: Our survey showed statistically significant increases in confidence in all procedures: arterial line placement (before:

median 5, standard deviation (SD) = 2.39; after: median 7, SD = 1.82; percent increase in median = 40%; $P<0.001$), CVL (before: median 3, SD = 2.30; after: median 7, SD = 1.90; percent increase in median = 133%; $P<0.001$), LP (before: median 5, SD = 2.35; after: median 7.5, SD = 1.87; percent increase in median = 50%; $P<0.001$), arthrocentesis (before: median 5, SD = 2.38; after: median 8, SD = 1.73; percent increase in median = 60%; $P<0.001$), and ACLS (before: median 4, SD = 2.09; after: median 6, SD = 1.90, percent increase in median = 50%, $P<0.001$).

Conclusion: Education in bedside procedures is becoming increasingly difficult in the modern health care system. The traditional teaching style of "see one, do one, and teach one" is becoming obsolete in today's higher acuity clinical environment with increasing time demands on trainees. Our simulator-based procedure training successfully increased residents' confidence in performing bedside procedures. Future studies will need to focus on demonstration of improved patient care outcomes.

A Case of Endobronchial Granular Cell Tumor: A Rare Entity

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Introduction: Granular cell tumors (GCT) are benign neural tumors first described in 1926. Mostly located in the head and neck region, skin, and subcutaneous tissues, their presence in the endobronchial region is uncommon, with less than 100 cases reported.

Case: A 54-year-old woman was seen in consultation for chronic obstructive pulmonary disease exacerbation. Chest radiograph showed mild apical fibro nodular scarring. Despite appropriate treatment, the patient continued to have shortness of breath, cough, and wheezing along with increasing oxygen requirements. A high resolution CT scan showed a wedge-shaped opacity in the medial left upper lobe. Bronchoscopy was performed revealing an endobronchial whitish growth at the subcarina. Biopsy was obtained. Microscopic examination showed bland appearing eosinophilic cells within the endobronchial submucosa. Cells had intact

nuclear to cytoplasmic ratio and immunohistochemical stains were positive for CD56, S-100 and vimentin. A diagnosis of endobronchial GCT was made.

Discussion: Granular cell tumors of the lung are rare. They comprise only 0.2% of all intrapulmonary neoplasms. Two percent to 6% of GCTs occur in the lung, and of these, 90% are endobronchial. It has been established now that they arise from Schwann cells. Patients with benign endobronchial tumors may present with cough, dyspnea, wheezing, hemoptysis, and post obstructive pneumonia. Chest radiographs may be completely normal. Depending on the size of the endobronchial mass, there may be signs of distal pneumonia, atelectasis, mucoid impaction, bronchiectasis, and air trapping. In some instances, malignant GCTs also have been reported. Endobronchial ablation using argon plasma coagulation is the current treatment of choice with special emphasis on bronchoscopy for follow-up due to the risk of recurrence.

Thrombocytopenia as the Initial Presentation Angioimmunoblastic T-Cell Lymphoma: A Case Report

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Introduction: Angioimmunoblastic T-cell lymphoma (AITL) is a peripheral T-cell lymphoma that typically presents with generalized lymphadenopathy and systemic B symptoms. We describe a patient with AITL who had an unusual presentation with only thrombocytopenia on a routine complete blood cell count (CBC).

Case: An 85-year-old woman presented with complaints of knee pain and generalized weakness for a week. Platelet count was 10,000, which led to her admission. Physical exam was unremarkable. No lymphadenopathy appreciated. Platelets and hemocrit continued to fall, and she subsequently received intravenous immunoglobulin, dexamethasone, platelets, and blood transfusions. CT abdomen showed extensive retroperitoneal, pelvic, and mesenteric lymphadenopathy. Bone marrow biopsy showed normocytic

anemia, severe thrombocytopenia, and mild immature myeloid shift. A right axillary lymph node biopsy showed AITL.

Discussion: AITL is a type of peripheral T-cell non-Hodgkin lymphoma that commonly presents with systemic B symptoms and generalized lymphadenopathy. One study found that 99% of patients with AITL had peripheral lymph node enlargement on initial presentation and 91% had involvement of at least 2 or more lymph node groups, which was absent or at best very obscure in our patient. Seven percent of patients had idiopathic thrombocytopenic purpura. Bone marrow was infiltrated in 60% of the cases in the study. In other studies, thrombocytopenia was present in 30% and 18.5% of the cases. In another study that examined bone marrow involvement in AITL, only 1 out of the 6 patients with AITL who had uninvolved bone marrow had platelet count below $150,000 \times 10^9/L$.

Conclusion: Although thrombocytopenia is rarely the initial presentation, AITL must be kept in mind in the setting of unexplained thrombocytopenia even if the typical features of a lymphoma are absent.

Medullary Renal Cell Carcinoma in a Patient with Sick Cell Trait

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Case: A 57-year-old African-American woman presented with a past medical history significant for morbid obesity, hypertension, and sickle cell trait. She was diagnosed with large right-sided renal mass as a result of workup of recurrent urinary tract infections. Subsequent MRI of the abdomen confirmed this finding, and further demonstrated invasion of the inferior vena cava (IVC) and adjacent lymph nodes. No mass was seen on imaging studies 1 year prior. Patient underwent nephrectomy, and pathology revealed renal medullary carcinoma. Subsequent workup showed multiple pulmonary lesions consistent with metastatic disease. Patient was initiated on bortezomib.

Discussion: Medullary renal carcinomas are rare, rapidly progressive cancers of the kidney, and primarily affect young black patients

with sickle-cell disease or trait. Survival is very poor, even with aggressive treatment, including surgical resection and various chemotherapeutic regimens. Currently, no effective therapy has been reported for this disease. Chemotherapy is based largely on very limited published data. Despite its close association with sickle cell trait and cytogenetic abnormalities, no specific genetic abnormalities were identified. Due to its rapid development, this cancer often is metastasized at the time of diagnosis. Early diagnosis and treatment will be the key to improve survival. Therefore, identification of the disease-specific genetic abnormality will be extremely helpful, and if a patient with sickle cell trait or disease presents with urinary symptoms, an imaging test is strongly indicated and should be considered early on, as renal medullary carcinoma is one of the differential diagnoses.

Atypical Presentation of Extra Adrenal Malignant Pheochromocytoma

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Introduction: It is believed 85% of pheochromocytomas are adrenal origin and 15% are extra adrenal paragangliomas. Metastatic pheochromocytomas are more common in extra adrenal abdominal pheochromocytomas in about 36% to 50%.

Case: A 51-year-old woman with past medical history of hypertension and migraines presented to the ED with 2-week history of generalized weakness, dull abdominal pain, nausea, and vomiting. Patient was hypercalcemic and was diagnosed with primary hyperparathyroidism. Due to hypertension, patient urine metanephrines and catecholamines were performed. She was discharged in stable condition after treatment for primary hyperparathyroidism and was readmitted within 2 weeks with worsening abdominal pain. Results of the previous tests were concerning for significant catecholamine release. Patient had a CT of abdomen, which showed no abnormalities of adrenal glands but a necrotic mass within the pancreas as well as multiple metastasis within the liver. At that time, she started to develop palpitations and signifi-

cant diaphoresis. She was started on alpha blockade with no significant improvement of symptoms. First biopsy of liver lesion was not indicative of pheochromocytoma. Due to worsening symptoms, a second biopsy of liver lesion was performed, confirming metastatic malignant pheochromocytoma. Patient was scheduled for embolization of liver metastasis, but decided not to follow up with treatment as well as chemotherapy.

Discussion: Case depicts an atypical presentation of a malignant extra adrenal pheochromocytoma. Even if a patient does not present with typical symptoms of palpitations, diaphoresis, or episodic hypertension, pheochromocytoma should be kept on the differential in a patient with hypertension. Absence of adrenal mass does not exclude pheochromocytoma; if there is a clinical suspicion screening should be performed.

A Case of Extraskkeletal Calcification-Calciophylaxis

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Introduction: Calciophylaxis is a rare, often fatal disorder characterized by systemic medial calcification of the arterioles that leads to ischemia and subcutaneous necrosis. It is seen almost exclusively in patients with end-stage renal disease (ESRD) and hyperparathyroidism, affecting 1% to 4% of the population with ESRD.

Case: A 48-year-old woman with history of ESRD due to focal sclerosing glomerulonephritis on hemodialysis and history of failed kidney transplant presented with a very painful skin rash over the lower abdomen and chest. There were no vesicles or ulcerations, but lesions were progressively darkening and enlarging in size over 3 weeks. There was no fever, joint pain, or sore throat. On physical examination, pulse rate 106/min, temperature 37.8°C, respiratory rate 18/min, blood pressure 90/60mmHg. She had marked areas of necrotic papulomacular lesions over lower abdomen, bilateral breast fold areas, and right upper thigh. There was bilateral pitting edema, but peripheral pulses were palpable symmetrically. The rest of physical examination was unremarkable. Laboratory

workup showed blood urea nitrogen 31mg/dl, creatinine 9.8mg/dl, parathyroid hormone (PTH) 63 pg/ml, calcium 7.5mg/dl, phosphorous 8.4mg/dl, hematocrit 29.6%, WBC 6800/mm³ and international normalized ratio (INR) was 1.0. Autoimmune work up for vasculitis was negative. Skin biopsy revealed intimal calcification in the small- and medium-sized blood vessels with necrosis of the overlying skin consistent with calciphylaxis. She was treated with phosphate binders, short interval dialyses, and sodium thiosulfate. The patient developed ulcerative lesions 3 months after discharge and is receiving outpatient wound care and pain control.

Discussion: Calciphylaxis is characterized by areas of painful ischemic necrosis that usually develop on areas with greatest adiposity including abdomen, buttock, and thigh. These ischemic changes lead to livedo reticularis and violaceous plaque-like subcutaneous nodules that progress to necrotic ulcers that often become super infected. There are no specific diagnostic laboratory tests for calciphylaxis. Skin biopsy with strong clinical suspicion is helpful in confirming diagnosis. Treatment is mainly supportive. There are no controlled prospective studies that compare different treatment strategies. Both medical and surgical interventions can be tried. Correcting PTH using cinacalcet or parathyroidectomy in refractory cases and normalizing serum calcium and phosphate abnormalities using noncalcium-containing phosphate binders is recommended. In patients with debilitating necrotic lesions, treatment with sodium thiosulfate has shown significant reduction in pain and skin lesions.

Record-Breaking Vancomycin Level Causes Kidney Catastrophe

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Introduction: Vancomycin-induced nephrotoxicity is a well-known condition affecting 5% to 7% of treated patients. Our case represents the highest measured serum vancomycin level ever recorded in the medical literature to cause acute renal injury.

Case: A 52-year-old man with past medical history of newly diagnosed type 2 diabetes mellitus presented with left hand cellulitis

and abscess. The patient denied taking any medications prior. He was started on IV vancomycin and piperacillin-tazobactam for broad spectrum coverage. He then underwent incision and drainage of the abscess with cultures of the wound positive for methicillin-resistant staphylococcus aureus. On day 7, he was discharged home on vancomycin 1.75 grams intravenously every 8 hours. One week later, his routine vancomycin trough level was 145.1 mcg/mL (therapeutic 10 - 40 mcg/mL). He was readmitted, vancomycin was discontinued, and repeat levels rose to 177.7 mcg/mL. His only complaint was a 3-day history of weakness and fatigue. Physical exam revealed a well-healing left hand wound and was otherwise unremarkable. Further workup uncovered his serum creatinine had climbed to 5.60 mg/dL from his baseline of 0.9 mg/dL 9 days prior. Urinalysis showed 1-5 white blood cells and no eosinophils. His fractional excretion of sodium was 6.3%, indicating intrinsic renal injury. Bilateral renal gallium scan demonstrated no increased uptake of contrast. Renal biopsy was not done. The etiology of the patient's injury likely was acute tubular necrosis due to the direct renal toxicity from vancomycin. After 4 days his symptoms resolved and he was discharged home. Follow-up labs demonstrated gradual improvement in renal function, but 19 weeks later the serum creatinine was 1.65 mg/dL.

Discussion: Vancomycin-induced nephrotoxicity is known to cause acute tubular necrosis or acute tubular interstitial nephritis. Treatment is simply discontinuation of vancomycin with close monitoring. To date, this is the highest known serum vancomycin level ever recorded in humans. Our case further exemplifies the importance of clinically managing vancomycin through careful dosing and drug level monitoring.

A Unique Presentation of Clostridium difficile Colitis

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Introduction: The classic presentation of *Clostridium difficile* colitis includes symptoms of watery diarrhea in the context of recent antibiotics while hospitalized. We present a

case of fulminant pseudomembranous *C difficile* colitis in a patient who did not fit this classic paradigm.

Case: An 82-year-old man was admitted to our institution with progressively worsening bilateral lower quadrant abdominal pain 1 month in duration. Midway through this course he was evaluated in the ED and found to have CT evidence of inflammatory changes about the lower left colon concerning for diverticulitis. He was prescribed oral ciprofloxacin and metronidazole therapy, along with close outpatient follow-up. However, his symptoms of severe "waves of pain" worsened, prompting hospital admission. Review of systems was negative for fevers, chills, weight loss, nausea, vomiting, diarrhea, constipation, or gastrointestinal (GI) blood loss. He had no sick contacts, recent travel, or mitigating factors. Pertinent past medical history included severe diverticulitis 10 years prior that led to a partial sigmoid resection with anastomotic repair subsequent to a ruptured diverticulum. Two years later he had required balloon dilation at the anastomosis secondary to a localized narrowing.

On exam, patient was afebrile and hemodynamically stable. He appeared uncomfortable but had a nonsurgical abdomen. Bowel sounds were present with moderate tenderness to palpation of the bilateral lower quadrants. Laboratory analysis included normal hemogram, electrolytes, and creatinine but with C-reactive protein (CRP) elevation to 4.5 (normal 0.0-0.8). Repeat CT scan revealed similar inflammatory changes as prior but dilation of the colon proximal to the inflammation also was noted. A gastroenterology consult was obtained with recommended colonoscopy to further define the inflammation. Interestingly, a stool sample obtained prior to colonoscopy tested positive for *C difficile* toxin. Endoscopic evaluation revealed fulminant, pseudomembranous colitis proximal to a tight stricture. Unfortunately, despite aggressive medical therapy, our patient developed toxic megacolon with subsequent rupture a few days later. He had an emergent total colectomy followed by a prolonged ICU stay but eventually passed away.

Discussion: *C difficile* infection is on the rise.

Heightened awareness of its presence and its potential to cause significant morbidity and mortality is needed. This case is unique in that our patient's lack of diarrhea was secondary to his colonic stricture, which was presumably worsened by surrounding inflammation.

Blast From the Past

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Case: A 54-year-old man living in Wisconsin, with no medical problems started experiencing numbness over his abdomen and back. Over the next 3 months, he developed leg weakness and difficulty walking. He was evaluated at a local ED where he had a normal head CT, chest x-ray, CBC, urine analysis, TSH, creatine kinase (CK), and basic metabolic panel. Lumbar spine x-ray revealed mild degenerative joint disease. He was referred subsequently to a free clinic due to lack of insurance.

The next month he was evaluated in our ED due to persistent symptoms and was found to have a stiff-legged gait but otherwise unremarkable neurological exam. Erythrocyte sedimentation rate (ESR) and B12 were normal. Patient was asked to follow up with neurology.

At his neurology clinic visit a month later he was noted to have 3/5 strength and hyperreflexia in his legs along with symmetric loss to pinprick sensation caudal to T4 and positive Romberg's test. He also had developed issues with bladder incontinence. MRI spine revealed destructive mass lesion at C7-T2 with epidural involvement and cord compression suggestive of TB, fungal infection, or neoplasm. Fine needle aspirate was nondiagnostic. Open biopsy specimen showed granulomatous inflammation with *Blastomyces dermatidis* organisms. Patient received induction liposomal amphotericin B, which ultimately led to renal failure. He was switched to oral voriconazole, which is to be continued for several months. He underwent physical therapy and was showing gradual improvement in strength when seen at 3-month follow-up in clinic.

Discussion: Vertebral blastomycosis is a rare, potentially fatal fungal infection of the spine

with myriad presentations. It often leads to delayed diagnosis, such as in our patient, with resulting neurological deficits and deformities. There are no pathognomonic findings on imaging studies. For patients who live in endemic areas who present with epidural masses/abscesses, the differential should include blastomycosis. Definitive diagnosis includes culture of this dimorphic fungus or direct visualization of the broad-based, budding yeast on histopathology. Amphotericin B is the first line treatment for life-threatening or CNS infections. Therapy can be switched to an oral azole once the disease is under control. Surgical management is reserved for those who do not respond to medication or those with progressive/severe neurologic deficits, spinal deformity, or instability.

Overlap Syndrome in the Presence of Renal Cell Carcinoma

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Introduction: Connective tissue disorders have been associated with malignancies. We describe a patient with an overlap syndrome whose rapid disease progression coincided with the discovery of a renal tumor.

Case: A 75-year-old woman presented with a 3-month history of progressive difficulty grasping objects, unsteadiness, dyspnea, xerostomia, xerophthalmia, and a 35-pound weight loss. She also described a several-year history of gastroesophageal reflux and Raynaud's phenomenon. Physical exam revealed facial telangiectasias, bibasilar inspiratory rales, sclerodactyly, and absent pinprick and vibratory sensation in her toes. Several metacarpophalangeal joints, proximal interphalangeal joints, and both ankles were swollen and tender.

Laboratory tests showed ESR 79, estimated glomerular filtration rate (eGFR) 39.9, ANA HEp-2 1:160, RF 80, SSA > 8.0, positive antineutrophil cytoplasmic antibody (c-ANCA), and proteinase 3 (PR3) > 8.0. Urinalysis revealed 3+ hematuria. Nerve conduction studies and electromyography (EMG) revealed a length-dependent, predominantly axonal, mixed sensorimotor neuropathy. CT of the chest and abdomen revealed a patu-

lous esophagus, pulmonary fibrosis, and a left renal mass. A left heminephrectomy was performed; tissue biopsies confirmed renal cell carcinoma, plus polyangiitis and granulomatosis. The patient's presentation is consistent with an overlap syndrome of limited scleroderma, Sjögren's syndrome, and Wegener's granulomatosis. Treatment with methylprednisolone and rituximab improved her sclerodactyly, polyarthritis, dyspnea, and renal function (eGFR 61.0) and stabilized her neuropathy.

Discussion: Patients with rheumatic disease are at increased risk for malignancy. Conversely, connective tissue disorders may manifest as rheumatological paraneoplastic syndromes, appearing at cancer diagnosis or earlier. Primary and secondary presentations are indistinguishable. Renal cell carcinoma has been reported independently to occur simultaneously with scleroderma and Wegener's granulomatosis, suggesting a common pathogenesis. Partial remission of symptoms has been achieved in some patients following treatment for a co-existing renal tumor.

Conclusion: Diagnosis of an autoimmune disorder in the presence of a malignancy should prompt the clinician to consider a secondary etiology. Eradication of the tumor may reduce the amount of immunosuppressive therapy required to treat the rheumatological disease.

Pyoderma Gangrenosum as the First Manifestation of Essential Thrombocythemia

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Introduction: Pyoderma gangrenosum (PG) is a rare disorder that physicians should consider in patients presenting with nonhealing ulcers. PG has been associated with inflammatory bowel disease, arthritides, and hematological conditions. We report the rare case of PG in a patient with essential thrombocythemia (ET).

Case: A 78-year-old man was admitted for IV antibiotic therapy for a nonhealing ulcer. On admission, he had elevated WBC of 24,000/ul and platelet count of 900,000/ul, which initially was thought to be reactive. After

48 hours of IV antibiotic therapy there was no clinical improvement. Initial and repeat cultures from the wound were negative for all organisms. Skin biopsy revealed intense neutrophilic dermatoses consistent with PG. Bone marrow biopsy was consistent with essential thrombocythemia showing increased megakaryocytes. He was treated successfully with hydroxyurea and prednisone.

Discussion: PG was first described in 1916 by Brocq. Studies have reported a low incidence ranging between 3 million and 10 million per year. It occurs in patients 20 to 50 years of age with a slightly female predominance. The pathogenesis is not clearly understood. Although it is idiopathic in 50% of the cases, it can be associated with other systemic illnesses. Multiple theories have been postulated including abnormal neutrophil trafficking, dysregulation of innate immunity, pathergy, and cytokine release. Rarely familial forms have been described.

PG is associated more commonly with inflammatory bowel disease and less commonly with arthritis, malignancies, and paraproteinemias. It starts as a painful nodule most frequently occurring on lower extremities. There are 5 subtypes, with bullous being the most common form associated with hematologic malignancies.

PG is a diagnosis of exclusion. Skin biopsy shows intense neutrophil dermatoses. The mainstay of treatment is corticosteroids. Cyclosporine or other immunosuppressive agents can be used. Biologic therapy with anti-tumor necrosis factor- α (TNF- α) have been used for refractory PG. Emerging research suggests granulocyte apheresis and phosphodiesterase-4 inhibitor may have a role in treatment.

“Blessing for the Bleeder”:

Bevacizumab in Hereditary Hemorrhagic Telangiectasia—A Novel Therapeutic Approach

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Introduction: Hereditary hemorrhagic telangiectasia (HHT) is a vascular disorder charac-

terized by cutaneous and mucocutaneous telangiectases resulting in severe and recurrent epistaxis and gastrointestinal hemorrhages. Role of bevacizumab in HHT has been reported to show improvement in epistaxis, telangiectasias, and hemoglobin stabilization. However the optimal dose and schedule of administration in HHT is unknown.

Case: A 56-year-old woman diagnosed with HHT at age 35 was on oral iron supplementation and maintained normal hemoglobin until June 2007 when her hemoglobin declined to <10 gm/dl and she was started on parenteral iron therapy. Because of persistent melena she was started on ethinyl estradiol. She developed dyspnea when her hemoglobin was <8.5 gm/dl, requiring 2 to 4 units packed red blood cell (PRBC) transfusions every month to maintain hemoglobin >8.5 gm/dl. In 09/2011, bevacizumab was started at 10mg/kg at 2 weekly intervals. Melena resolved within a week, hemoglobin rose to 14.2 g% within 4 weeks; serum ferritin increased (from 28 ng/ml to 246 ng/ml) within 6 weeks, with no further parenteral iron supplementation and marked decrease in episodes of epistaxis. She received total of 9 doses of bevacizumab at 10mg/kg body weight (3 doses each at increasing intervals) and 2 doses at 7.5 mg/kg body weight (every 4 weeks, still ongoing). Her hemoglobin continues to remain stable with negligible epistaxis, without the need for blood/iron infusions.

Discussion: Because of the molecular mechanisms involved in both angiogenesis and HHT, a vascular endothelial growth factor inhibitor such as bevacizumab may be an effective treatment for HHT. Prior studies used bevacizumab in doses ranging from 10 mg/kg to 5 mg/kg body weight every 2 weeks. We attempted to find a schedule minimizing side effects without compromising therapeutic benefits. Our observations suggest that bevacizumab at dose of 7.5 mg/kg every 4 weeks is efficacious in controlling symptoms in HHT.

Cervical Osteomyelitis After Repeated Esophageal Dilation for Dysphagia Associated with Chemoradiation-induced Esophageal Strictures

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Introduction: Dysphagia secondary to esophageal strictures is a common complaint of patients with head and neck cancers following treatment with chemoradiotherapy. We report a rare case of cervical osteomyelitis after multiple endoscopic dilations for chemoradiation-induced esophageal strictures.

Case: A 69-year-old man with stage IVa squamous cell carcinoma of the larynx was treated with chemoradiotherapy with prophylactic placement of a percutaneous endoscopic gastrostomy (PEG). He developed dysphagia 6 months after chemoradiation for which he underwent esophagogastroduodenoscopy via his PEG tube and was found to have high-grade proximal esophageal stenosis. After 6 endoscopic dilations to 20 mm over a 4-month course, his dysphagia gradually resolved and the PEG tube was removed.

He presented to the ED 6 weeks after his last dilation, complaining of progressive neck pain for 1 month. On examination he appeared healthy overall, except for a temperature of 103°F and marked tenderness on paravertebral cervical spine muscles. Cervical spine MRI studies demonstrated C6 and C7 pathological fractures, osteomyelitis at the C6-7 level, and an esophageal-spinal fistula. His blood culture was positive for peptostreptococcus micros. Given the absence of neurological deficits, he was treated conservatively with intravenous ertapenem for 10 weeks, followed by 6 weeks of augmentin, and cervical spine stabilization. He received nothing by mouth and a PEG tube was re-placed for nutritional support. Sequential MRI studies of the cervical spine showed progressive resolution. Over the course of 4 months, he remained neurologically stable.

Discussion: Esophageal strictures are a common sequela after chemoradiation therapy for head and neck cancers and can lead to significant dysphagia. Cervical osteomyelitis is a rare complication of multiple esophageal dilations in these patients. Its major clinical manifestation is neck pain without neurological deficits. A high index of suspicion is, therefore, necessary in all patients having neck pain post esophageal dilation for malignant esophageal strictures.

Erythromycin in Acute Upper Gastrointestinal Bleeding: A Meta-Analysis

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Introduction: Emergent upper endoscopy is the standard of care in patients with upper gastrointestinal bleeding (UGIB). Adequate visualization of the GI tract is important for diagnosis and therapy. Several studies have evaluated the role of erythromycin before endoscopy with mixed results. We performed a meta-analysis of randomized controlled trials (RCTs) to assess the efficacy of erythromycin for acute UGIB prior to endoscopy.

Methods: MEDLINE, Cochrane Central Register of Controlled Trials and Database of Systematic Reviews, PubMed, and recent abstracts from major conference proceedings were searched (through June 2012). RCTs evaluating the role of erythromycin in acute UGIB in adult patients were included. Two independent reviewers extracted data using standard forms. Data regarding the following outcomes were extracted: visualization of the mucosa, need for repeat endoscopy, blood transfusion, and length of stay. Summary statistics were computed using Comprehensive Meta-analysis software. Publication bias was assessed by funnel plots. Heterogeneity was assessed.

Results: Seven studies met the inclusion criteria (n=657). Patients with both variceal and nonvariceal bleeding were included. Mean age ranged from 56 to 64.5 years. Dose of erythromycin administered varied from 125mg to 3mg/kg. Endoscopy was performed 20 to 120 minutes after administration of erythromycin. Erythromycin group showed improvement in mucosal visualization (relative risk [RR] 1.6; 95% CI 1.14-2.35, $P=0.008$), number needed to treat (NNT) was 4 (95% CI, 2-11), decreased need for repeat endoscopy (RR 0.52, 95% CI 0.31-0.89, $P=0.02$, I² =18, NNT 11). The need for blood transfusion was lower (weighted mean difference) -0.52; 95% CI -0.957 to 0.08, $P=0.02$) and length of hospital stay was shorter (mean difference: 1.56,

95% CI 0.6-2.5) in erythromycin group.

Conclusion: Erythromycin prior to endoscopy in adult patients with upper GI bleed improves visualization of the mucosa and decreases the need for repeat endoscopy, hospital stay, and the need for blood transfusion.

Cytomegalovirus Colitis During Mycophenolate Mofetil Therapy for Anti-synthetase Syndrome

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Introduction: Mycophenolate is an immunosuppressive medication that inhibits purine synthesis and is often used in solid organ transplant patients. There is hardly more than 1 case report in the literature where there is a suggestion that mycophenolate on its own has the immunomodulatory power to cause cytomegalovirus (CMV) reactivation.

Case: A 49-year-old Caucasian woman who was receiving mycophenolate mofetil for anti-synthetase syndrome presented with a 2-week history of watery diarrhea, nausea, and vomiting. This was associated with right upper quadrant abdominal pain, low grade fevers, and malaise. Preliminary stool studies, cultures, and *C difficile* work-up were negative. Liver chemistries were slightly deranged in a noncholestatic pattern. Viral hepatitis panel was normal. Chest radiograph was consistent with patchy bilateral infiltrates suggestive of pneumonitis. CMV serology was positive. Colonoscopy was done for tissue diagnosis, light microscopy revealed CMV inclusions, and immunostains were positive confirming diagnosis. Mycophenolate therapy was discontinued and the patient was started on valgancyclovir for 3 weeks. Her symptoms resolved and imaging and laboratory markers improved.

Discussion: Colitis is a frequent manifestation of acute CMV infection. While there is evidence that mycophenolate may have increased risk of CMV disease in solid organ transplant patients, there are very few reported cases of CMV disease in patients receiving mycophenolate immunosuppressive therapy for other causes.

Triple Antiviral Therapy in Hepatitis C-Induced Cryoglobulinemic Vasculitis and Lymphoproliferative Disorder

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Introduction: Mixed cryoglobulinemia (MC) and B-cell lymphoma are known complications in patients with hepatitis C due to antigenic stimulation by hepatitis C E2. There is emerging evidence suggesting that combination therapy with ribavirin and interferon alfa is beneficial in patients with hepatitis C-induced cryoglobulinemia. The effect of protease inhibitors on cryoglobulinemia has not been studied yet. We present a case of mixed cryoglobulinemia secondary to HCV genotype 1 that presented with clonal B-cell proliferation and was successfully treated with triple antiviral therapy using protease inhibitors.

Case: A 58-year-old-man with history of intravenous drug abuse was hospitalized for maculopapular rash and severe Raynaud's phenomenon. Physical examination revealed massive splenomegaly and severe digital cyanosis. Laboratory tests were significant for pancytopenia, normal aspartate aminotransferase (AST) and alanine aminotransferase (ALT), positive rheumatoid factor and cryoglobulins, and low complement levels. Serology was positive for hepatitis C and negative for lyme, parvovirus, and human immunodeficiency virus (HIV). RNA amplification studies isolated genotype 1b. Imaging studies revealed a normal liver size and echo texture, massive splenomegaly, normal portal pressure, and retroperitoneal lymphadenopathy. Flow cytometry revealed a polyclonal B-cell proliferation. The patient was treated with peginterferon, telaprevir, and ribavirin (triple antiviral therapy) for HCV. There was marked clinical improvement with symptom resolution, nondetectable viral load and significant decrease in the cryocrit levels 4 weeks after treatment.

Discussion: Pathophysiology behind hepatitis C virus infection and the development of lymphoma is still under debate. However it is likely that mechanisms involved in mixed cryoglobulinemia and B-cell lymphoma share

similar features. Standard treatment for MC vasculitis has included pegylated interferon and ribavirin, reserving rituximab for severe cases. Introduction of new NS3/4A serine protease inhibitor like telaprevir and boceprevir has markedly improved sustained virological response. Combination of new NS3 serine protease inhibitors with peginterferon and ribavirin has not been studied yet.

Association of Nursing Home Regulatory Compliance and Emergency Department and Inpatient Admissions for Fall-related Injuries

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Objective: To evaluate the relationship between nursing home regulatory compliance and staffing with the occurrence of falls requiring ED or hospital inpatient care.

Methods: A county-level analysis of Wisconsin nursing home, ED, and inpatient data from the Wisconsin Hospital Association, Center for Medicare Services, and Wisconsin Department of Health Services was performed. There were 59,186 Wisconsin nursing home residents 65 years of age and older in 2007-2008; hospital inpatient and ED discharge records documented 715 admissions from skilled nursing facilities (ie, nursing homes). These admissions were reported in 56 of the 71 counties with nursing homes. A multivariate analysis of these 56 counties was performed using negative binomial regression to analyze the association between nursing home staffing, demographics, federal violations, and ED/inpatient admissions for fall-related injuries.

Results: Residents from counties with nursing homes having a greater number of federal violations had a significantly greater risk of ED/inpatient admissions for fall-related injuries, particularly with violations in the "Quality of Life" category. Counties with a fewer number of nursing homes per county and in large fringe metropolitan counties also had a significantly greater risk of ED/inpatient admissions for fall-related injuries.

Conclusion: Increased compliance with federal nursing home standards may decrease ED/inpatient admissions from nursing homes for fall-related injuries.

Pure Red Cell Aplasia and Anti-erythropoietin Antibodies—Not Only in Chronic Kidney Disease Patients

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Introduction: Acquired pure red cell aplasia (PRCA) is a rare condition causing severe anemia and characterized by a low reticulocyte count, absence of red blood cell precursors, and normal leukocyte and platelet morphology. PRCA, often idiopathic, has been linked to several medical conditions, such as thymomas, hematologic malignancies, autoimmune disorders, and a variety of drugs and viral infections. Review of the literature does show reported cases linking PRCA to the use of recombinant human erythropoietin (EPO). We present the case of PRCA secondary to anti-EPO antibodies.

Case: A 64-year-old man with cirrhosis secondary to hepatitis C and pancytopenia was admitted for anemia with hemoglobin of 5.5 g/dL. Six months prior he was started on treatment for hepatitis C with telaprevir, ribavirin, and peginterferon. He had baseline hemoglobin of 11.5 g/dL. Four weeks later, he started procrit injections (epoetin alpha, 40,000 units weekly) due to decrease hemoglobin to 7.0 g/dL. He responded to treatment and dosing was continued. He completed 12 weeks of treatment with telaprevir. Ribavirin and peginterferon were discontinued due to new onset of decompensated cirrhosis. Despite stopping the hepatitis C treatment and continuing epoetin injections, his hemoglobin decreased to 5.5 g/dL. On the day of his admission, his reticulocyte count was undetectable raising concern for PRCA. He was given blood transfusions, and EPO injections were discontinued. A bone marrow biopsy showed "markedly hypocellular bone marrow with red cell aplasia." Further work-up was negative for parvovirus, CMV, and Epstein-Barr virus (EBV). Chest CT did not show a thymoma or lymphadenopathy. Anti-EPO antibody titers were positive.

Discussion: The majority of documented cases of PRCA occurred in patients with chronic kidney disease (CKD). Our patient illustrates that this phenomenon is not limited to this population. Most cases of anti-EPO antibodies have occurred with use of Eprex,

an epoetin alpha product that is no longer manufactured. PRCA associated with anti-Procrit antibodies has been reported in only 6 cases. Data guiding management is limited. Treatment consists of blood transfusions, stopping EPO injections, and immunosuppressive therapy. Steroids alone or in combination with cyclophosphamide, and intravenous immunoglobulin have been used most widely. PRCA resolves spontaneously in 20% to 30% of cases associated with anti-epoetin antibodies. Rechallenge to EPO injections is contraindicated.

IIB or Not IIB? That is the Question

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Case: A 61-year-old man presenting with ST elevation infarct developed respiratory distress, hypotension, and thrombocytopenia to 26 post procedure. Eptifibatide and heparin were stopped and he received supportive measures including mechanical ventilation and pressure support. Initial work-up on chest x-ray and bronchoscopy revealed diffuse alveolar hemorrhage (DAH). Thrombocytopenia was negative for disseminated intravascular coagulation (DIC) and heparin-induced thrombocytopenia (HIT), but positive for eptifibatide antibodies. Platelets initially improved to 65, and 2 units of platelets were transfused with increase to 103, but again fell to the fifties. Repeat DIC panel gave a DIC score of 5 with low fibrinogen and elevated D dimer with schistocytes on smear. Ultimately the patient developed multi-organ failure and received comfort care.

Discussion: Glycoprotein IIB-IIIa inhibitor-induced thrombocytopenia is a rare, serious complication, occurring in roughly 0.3% to 0.7% of patients. Onset of eptifibatide-induced thrombocytopenia (EIT) tends to occur within minutes to hours of administration (usually within 24 hours) and can be severe, with platelet counts dropping below 30,000. Other causes of acute thrombocytopenia should be excluded including pseudo-thrombocytopenia, DIC, and HIT. Treatment includes discontinuation of the offending agent, platelet and red blood cell transfusions in the case of significant bleeding, and other supportive measures. Platelet count should

improve over the next 3 to 6 days. DAH is also a complication of eptifibatide use first reported in 2004, with a growing number of case reports highlighting this emerging risk. Recent retrospective analysis has found DAH rates to be from 0.2% to 0.3%. Symptoms can include hypoxia, anemia, hemoptysis (though one-third do not have this symptom), and new chest infiltrates. Treatment includes discontinuation of IIB-IIIa inhibitors and other anticoagulation and supportive treatment to maintain O₂ saturations. This is a unique case in that our patient developed not only EIT with DAH, but subsequently developed DIC within hours leading to persistent thrombocytopenia and multi-organ failure. Thus, even in the setting of possible EIT, one must monitor platelet counts after stopping the medication and, if not improving, consider alternative or co-commitment pathology.

The Case of Cancer Incognito

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Case: A 59-year-old man with a history of hypertension, hyperlipidemia, and recent onset of partial seizures presented with a 2-day history of fluent aphasia and right hemiparesis. After admission to another hospital, he suffered a complex partial seizure followed by simple partial status epilepticus with a focus in the left temporoparietal lobe. He was transferred to our facility for further evaluation after being started on antiepileptic drugs (AEDs). Brain MRI and CT angiography were negative for cerebral infarct and aneurysm, but his MRI displayed abnormal fluid-attenuated inversion recovery (FLAIR) signal involving the left temporal lobe and peri-insular cortex, of concern for herpes simplex virus (HSV) encephalitis. He underwent lumbar puncture and was started empirically on acyclovir. CSF analysis revealed an elevated protein of 145 mg/dL, normal glucose of 65 mg/dL, and no leukocytes. HSV nucleic acid amplification test (NAAT) was negative, but CSF HSV IgG and IgM from CSF came back elevated. He was continued on acyclovir and had no further seizures. His fluent aphasia improved markedly over the next 5 days though he continued to have word-finding

difficulty and some intermittent weakness on his right side with numbness and tingling. He was discharged home, and 1 month later had a repeat brain MRI that revealed a new necrotic lesion in the same location in the left temporal lobe. Magnetic resonance spectroscopy (MRS) was performed to better characterize the lesion and was found to be most consistent with glioblastoma multiforme (GBM). Resection confirmed GBM, and the patient has since begun chemoradiation therapy.

Discussion: GBM presenting as acute encephalitis syndrome (AES) is rare; it is the cause of encephalopathy in 1.5% of patients in a few case series. Brain MRI reveals geographic ring enhancing lesions of highly dense neoplastic cells with a hypodense core on T1 images representing necrosis. T2 and FLAIR reveal a surrounding zone of vasogenic edema. Our patient's clinical picture, MRI, and response to antivirals were suggestive of HSV encephalitis despite a negative PCR, which can occur early on in HSV encephalitis. Follow-up MRI in HSV encephalitis is suggested to monitor for late sequelae including hemorrhage and necrosis. The new necrotic lesion on our patient's follow-up MRI was concerning for post HSV sequelae, but MRS was performed to better characterize the lesion. MRS in GBM demonstrates increased choline to creatinine peak ratio, increased lactate peak and decreased N-acetylaspartate (NAA) peak, which our patient had. This case stresses the importance of including tumors such as GBM in a differential diagnosis of AES.

Paraproteinemia-associated Polyarteritis Nodosa

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Introduction: Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis of medium-sized vessels with clinical manifestations resulting from inflammation and ischemia of affected organs. PAN has been described in association with viral infections and various lymphoproliferative disorders. Few case reports exist describing PAN associated with multiple myeloma.

Case: A 44-year-old woman presented with worsening neck and back pain of 7 months

duration associated with right lower extremity pain, paresthesia, and weakness with ankle dorsiflexion that started 3 months prior to admission. She also complained of severe fatigue and a recent 10-pound weight loss. On admission the patient was afebrile, hypertensive, and tachycardic. Exam revealed weakness with right ankle dorsiflexion and diminished sensation to light touch on the dorsum of the foot. Laboratory studies showed a creatinine of 1.75 mg/dL, normocytic anemia, positive antinuclear antibody (ANA) (1:80 titer), and elevated ESR and CRP. Urinalysis also showed 3+ protein and blood with a protein to creatinine ratio of 1.34. Serum protein electrophoresis revealed monoclonal paraproteinemia. The patient underwent kidney biopsy showing medium-sized vessel vasculitis consistent with PAN. Bone marrow biopsy and skeletal survey were completed and the patient was subsequently diagnosed with stage II IgG lambda multiple myeloma.

Discussion: While the association between vasculitis and cancer has been well described in the literature, there are only a small number of case reports describing paraneoplastic vasculitis in association with multiple myeloma. Hematologic malignancies, most frequently lymphomas, are the most commonly described malignancies associated with PAN. Clinical findings associated with PAN are nonspecific and, in general, no different than those seen in patients without underlying malignancy. The most common clinical features include fatigue, weight loss, and fever. Diagnosis requires vigilance and the integration of patient history, clinical findings, and biopsy data.

Respiratory Failure in Cryptogenic Organizing Pneumonia

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Introduction: Cryptogenic organizing pneumonia often presents with persistent coughing of 1 to 2 months duration, dyspnea on exertion, and weight loss in the setting of failed treatment for community acquired pneumonia. In many cases, this disease is managed on an outpatient basis after a lung biopsy confirms the findings of intraluminal

inflammation of alveoli including alveolar ducts. Treatment largely consists of steroids, which are slowly weaned over months.

Case: A 30-year-old man with a history of cognitive delay and CKD was brought by his mother to his primary care physician for a new-onset cough 5 days prior to hospitalization. His mother reported that aside from the new cough, he was well and continued to participate in daycare 3 days per week. He was placed on a 5-day course of azithromycin for atypical pneumonia but did not improve. They returned for reevaluation of his cough as well as a 1-day history of increased agitation. Initial workup was suggestive of pneumonia on clinical exam and chest radiograph, along with worsening kidney function. While in the ED, he developed increasing respiratory insufficiency and required intubation. He was started on broad spectrum antibiotic therapy and underwent diagnostic bronchoscopy on the first day of admission. Upon admission, he had leukocytosis that continued to rise despite therapy. After 7 days of IV antibiotic therapy and a large workup that remained negative, a lung biopsy showed evidence of organizing pneumonia.

Discussion: This case of biopsy-proven cryptogenic organizing pneumonia is unique because it presented in a 30-year-old man, and it was associated with respiratory failure requiring prolonged intubation. His initial chest radiograph showed bilateral pleural effusions with alveolar infiltration that could suggest a multifactorial cause as the reason for the respiratory failure. His leukocytosis continued to rise despite antibiotic therapy. Bronchoscopy with bronchoalveolar lavage (BAL) was performed on initial presentation to ICU and 5 days later, but both sets of cultures remained negative leading us to consider cryptogenic organizing pneumonia. Steroid treatment was the only therapy to improve his clinical status and ultimately allowed him to be extubated.

Still a Challenging Diagnosis

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Case: A 35-year-old man with a past medical history significant for juvenile idiopathic arthritis presented with 2 days of sharp, substernal chest pain preceded by 3 days of

pharyngitis, fevers, and diffuse myalgias. Tachycardia, tachypnea, proximal muscle weakness, and white tonsillar exudate were present on physical examination. Initial diagnostic studies were significant for a leukocytosis of 13,000/uL, slightly elevated aminotransferase levels, an elevated troponin level, and PR segment depression in the inferior leads on electrocardiogram (ECG). A TTE was unremarkable. An initial diagnosis of myopericarditis was made, and treatment with colchicine and aspirin was initiated. Cardiac MRI showed changes consistent with myocarditis. Tests for HIV, parvovirus B19, coxsackie A and B viruses, cytomegalovirus, and hepatitis A and B were all negative. Multiple urine, respiratory, and blood cultures were also negative. The patient continued to spike high fevers each night, even after resolution of the chest and pleuritic pain, and his leukocytosis persisted. A ferritin level was checked and found to be significantly elevated at 3440 ng/mL. Based on the Yamaguchi criteria, the patient was diagnosed with adult-onset Still's disease.

Discussion: Adult-onset Still's disease (ASD) is an inflammatory disorder characterized by high daily fevers, arthralgia, and an evanescent rash. It is a rare condition, occurring in less than 1 per 100,000 people with a bimodal age distribution peaking between ages 15-25 and 36-46. Although an infectious origin is suspected, the etiology remains unproven. There is no definitive test or laboratory value to diagnose ASD. The Yamaguchi criteria are used to establish the diagnosis. The 4 major criteria are persistent high fever, leukocytosis, arthritis or arthralgia, and a skin rash that is usually present during the febrile episodes. The minor criteria include a sore throat, organomegaly, elevated liver function tests, lymphadenopathy, and normal ANA and rheumatoid factor. Five of these features must be present, including two of the major criteria. The serum ferritin level is also markedly elevated in the majority of these patients. Several treatment options are available, including nonsteroidal anti-inflammatory drugs, glucocorticoids, disease-modifying antirheumatic drugs, and biologic immunomodulatory agents. The prognosis in ASD is generally favorable.

Raynaud's to Renal Crisis: An Atypical Scleroderma Presentation

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Introduction: Scleroderma renal crisis is a well-known severe complication of systemic sclerosis. However the renal crisis is only uncommonly the presenting feature of scleroderma without prior disease manifestations and requires a high degree of clinical suspicion to diagnose.

Case: A 25-year-old Hispanic woman with a history of Raynaud's phenomenon was hospitalized after her outpatient nephrologist noted her creatinine steadily rising over a week in conjunction with elevated blood pressures and new thrombocytopenia. An admission 1 month prior for hypertension and tachycardia resulted in an exclusionary diagnosis of constipation-induced hypertension. On present admission, examination was negative except for elevated blood pressures and trace pedal edema. Thrombotic thrombocytopenia was excluded, as were pheochromocytoma and renal artery stenosis. The patient's renal function continued to worsen, and a kidney biopsy was performed showing thrombotic microangiopathy. Hemodialysis was initiated. The patients' blood pressures remained elevated despite multiple medications, however no angiotensin-converting enzyme (ACE) inhibitor was initially begun. An autoimmune workup revealed a positive ANA and, despite no synovitis or skin abnormalities, a rheumatology consultation was obtained. The consulting service suggested the early presentation of systemic sclerosis based on the overall clinical picture. A subsequent esophagram revealed a widely patent gastroesophageal junction consistent with the patulous gastroesophageal (GE) junction often seen in patients with scleroderma. The patient was begun on an ACE inhibitor with improvement in her blood pressure, and eventually was discharged with continued hemodialysis and close nephrology and rheumatology follow-up.

Discussion: Though commonly diagnosed by rheumatologists, scleroderma can present atypically with minimal clinical evidence and requires a high degree of clinical suspicion to diagnose. A usual presentation of scleroderma

would be in a 30- to 50-year-old woman with Raynaud's, skin tightening, and intestinal symptoms. True Raynaud's phenomenon is not normal and can represent the early pathological immune infiltration and microvascular damage of scleroderma.

Tricuspid Regurgitation: Valvular Dysfunction on the Rise

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Case: A 75-year-old man with a history of coronary artery disease, congestive heart failure and second-degree atrioventricular heart block status-post pacemaker presented from an outside hospital for progressive weight gain and increasing abdominal girth over a year. Patient denied shortness of breath, alcohol or drug use, or recent travels. On labs, AST, ALT, and alkaline phosphate were mildly elevated with a normal bilirubin, INR, and albumin. He also had acute kidney injury and thrombocytopenia, but the remainder of the CBC and complete metabolic panel were normal. Abdominal ultrasound revealed findings consistent with cirrhosis (portal hypertension, a moderate ascites, and bidirectional flow within the portal vein). On admission to our hospital, viral serologies for hepatitis were negative. His pacemaker was interrogated and revealed new atrial fibrillation. A repeat echocardiogram showed a left ventricular ejection fraction (EF) of 45% to 50%, normal right ventricular function, severe biatrial dilation and pinning of the posterior leaflet of the tricuspid valve by the pacemaker lead with wide-open tricuspid regurgitation. Patient had a normal pulmonary artery systolic pressure. Patient was aggressively diuresed, amiodarone was initiated, and patient was cardioverted with restoration of sinus rhythm. His ascites, peripheral edema, acute kidney injury, and thrombocytopenia improved. On discharge he was scheduled for a repeat echocardiogram to reassess his tricuspid regurgitation.

Discussion: Pacemakers and implantable cardioverter-defibrillators (ICDs) are important medical devices used in the treatment of a variety of cardiac diseases. With the aging population and an increase in life expectancy, utilization of these devices is expected

to continue to rise. As such, it is important that general internists appreciate not only the indications, but also the complications that can occur secondary to these devices. Tricuspid regurgitation is one such known but underappreciated complication. Limited data on the frequency of tricuspid regurgitation related to endocardial lead implantation is conflicting, but the importance and clinical impact is not. Severe tricuspid regurgitation is known to be associated increased mortality. Early diagnosis of and intervention is critical for addressing an iatrogenic cause of valvular dysfunction.

Methanol Toxicity and a Normal Osmolar Gap

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Introduction: In patients presenting with volatile alcohol toxicity, early diagnosis and treatment is critical to prevent organ damage and death. The goal standard for diagnosing methanol and ethylene glycol (EG) toxicity is gas chromatography. Results can take up to days and many hospitals are not equipped to provide such test. Clinicians must rely on the clinical presentation and other laboratory tests such osmolar gap (OG) to make a diagnosis. An OG of >10 indicates the presence of other osmoles in the blood such as methanol or EG. We present a case of methanol toxicity and a normal osmolar gap.

Case: A 48-year-old Caucasian man was admitted to a community hospital 20 minutes after ingesting > 500 tablets of extra-strength acetaminophen. N-acetylcysteine infusion was initiated. He was mechanically ventilated due to impending respiratory failure. Initial laboratory revealed acetaminophen level of 86 mg/L, high anion gap metabolic acidosis (AGMA), negative urine analysis and serum drug screen. The patient was transferred to our facility (tertiary center) for further management. Upon arrival, acetaminophen level was >800 mg/L and lactic acid was 10.5 mmol/L. Serum osmolality was 296 mOsm/Kg with a normal osmolal gap (OG). Later, he developed acute hepatic and renal failure. Despite appropriate management, he had persistent metabolic acidosis, thus a vola-

tile gas screen was ordered. The results were available within 3 hours and were positive for methanol intoxication. The patient was started on renal replacement therapy and fomepizole infusion. The remaining of the hospital course was significant for development of cerebral edema. Hypothermia protocol was initiated. He then had an upper GI bleed that was likely secondary to caustic ingestion.

Discussion: An elevated OG is highly sensitive for volatile alcohol intoxication, but a normal OG does not necessarily rule it out. Once methanol is converted to its toxic metabolite—formic acid, this compound is no longer osmotically active. In the case of our patient, the OG may have been normal because it was checked several hours after the ingestion. Methanol may have been converted to formate, which causes AGMA. In addition, some patients may have an OG in the negative range at baseline (children have an OG of -2 +/- 6), making an OG <10 abnormal in such cases.

Lupus-Related Pulmonary Hemorrhage

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Introduction: Systemic lupus erythematosus (SLE) is an autoimmune disorder that can affect any organ system. Pulmonary hemorrhage (PH) is a rare (<2%-5.4%) but catastrophic complication (mortality 26%-92%) of SLE.

Case: A 29-year-old African-American woman with SLE managed with prednisone and mycophenolate presented with hemoptysis. Her initial exam revealed tachypnea and diffuse right-sided rales. Labs showed a normal white cell count, hemoglobin at baseline of 10.1 g/dL, platelets of 127,000, normal basic metabolic panel, elevated CRP of 1.0 mg/dL, low C3 of 74 mg/dL, and normal C4. She was intubated after several episodes of hemoptysis and increasing oxygen requirements. Chest radiograph revealed patchy infiltrates throughout the right lung, which rapidly progressed to diffuse bilateral alveolar infiltrates. Broad spectrum antibiotics were initiated and bronchoscopy revealed bloody secretions from her bilateral bronchi. High-dose IV methylprednisolone was initi-

ated, and she was continued on oral mycophenolate. After 3 days of IV steroids without improvement, plasmapheresis was initiated, resulting in significant improvement and extubation after the first of 5 sessions. At discharge, she had complete resolution of pulmonary hemorrhage and stable blood counts.

Discussion: Due to varied clinical presentation and nonspecific radiological findings, the diagnosis of PH may be challenging. No clear predictors of patients at increased risk of developing PH exist. Bronchoscopy with BAL is reliable in diagnosing PH and identifying possible infection. This is critical as treatment of PH requires high-dose immunosuppression. Broad spectrum antibiotics are recommended while awaiting culture results. After PH diagnosis, high dose IV steroids and cyclophosphamide should be initiated. Due to previous treatment failure with cyclophosphamide, our patient was treated with high dose IV steroids and mycophenolate. However, plasmapheresis is effective in patients who fail initial therapy, as seen in our patient.

Doubly Uncommon

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Introduction: *Fusobacterium nucleatum* is an anaerobic gram-negative bacillus best known for its role in causing periodontal disease. This is a rare case of a patient who presented atypically with infective endocarditis, found to be secondary to *F nucleatum*.

Case: A 71-year-old woman presented with a 1-week history of right index finger pain. Physical exam was significant for fever, cyanosis of the right distal phalanx, and a new III/VI systolic murmur. Blood cultures were drawn, and she was started on empiric antibiotics for concern of endocarditis. TTE did not identify any masses or vegetations, but transesophageal echocardiography (TEE) revealed a small, serpiginous echodensity in continuity with the posterior leaflet. Over 48 hours later, one blood culture returned positive for *F nucleatum*, and the patient was treated with ertapenem for endocarditis. At subsequent follow-up, she had improvement in her right index finger pain and resolution of her murmur.

Discussion: Although documented cases of *F nucleatum*-causing endocarditis are quite rare, many of the features of this patient's case are consistent with prior known cases. Thromboembolic phenomena, the presenting feature in this patient, have been frequently associated with *F nucleatum*. Additionally, this patient had no underlying valvular disease, which was also the case in a high proportion of previously documented cases of *F nucleatum* endocarditis. Although *F nucleatum* bacteremia can be fatal, its presentation often is insidious, as it was in this patient. Diagnosis of IE relies heavily on the use of blood cultures and echocardiography. In this patient, a new onset murmur was the most useful tool in establishing the diagnosis, serving as a reminder that while blood cultures and echocardiography are important, overutilization of them should not serve as a substitute for a thorough history and physical examination.

Thrombolytics and Hypothermia Protocol in Early Gestation Pregnancy with Pulseless Electrical Activity Arrest

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Introduction: The hypothermia protocol was developed for patients who underwent ventricular fibrillation arrests in the field, but it is being utilized more widely.

Case: A 34-year-old woman with 13-week pregnancy was found down by her husband. Cardiopulmonary resuscitation (CPR) was started 10 minutes later with emergency medical services (EMS). She regained pulses after 7 minutes of compressions. Upon arrival, she was nonreactive and subsequently underwent CPR twice more. Bedside echocardiogram noted right ventricular strain and CT scan showed massive bilateral pulmonary embolism with thrombus extending to the segmental arteries. She was treated with thrombolytics, and hypothermia protocol was initiated. Rewarming was completed without acute events, with pressor support discontinued and extubation the day after. She did return to the medical intensive care unit (MICU) for respiratory distress secondary to blood clot found in the airway, which was removed without issue. She was discharged

with normal neurologic exam. Follow-up as outpatient showed normalization of her right and left ventricular function. Unfortunately, the fetus shows evidence of fetal hydrops but persistent cardiac rhythm, and the family plans to complete to term.

Discussion: Cardiopulmonary arrest occurs in 1:30,000 pregnancies. There have been several case reports of successful resuscitation in pregnant women, with varying viability of the fetus. The International Liaison Committee on Resuscitation and the American Heart Association advocate delivery within 5 minutes of loss of maternal circulation for best chance of survival. However, in the field this is often impractical. This particular case illustrates the importance of timely diagnosis and intervention in that the administration of thrombolytics and hypothermia likely had significant impact on the return of the mother's neurologic status.

Mixed Picture – A Probable Case of NSAID-induced Hepatotoxicity

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Introduction: Sulindac is the nonsteroidal anti-inflammatory drug (NSAID) most frequently associated with hepatotoxicity at a rate about 5 to 10 times that of other NSAIDs. The pattern of injury is usually mixed, representing both hepatocellular and cholestatic injury with an idiosyncratic mechanism of action.

Case: A 53-year-old man with a past history of hepatitis B infection and recent colitis presented with nausea, abdominal pain, and fatigue of 1 day's duration. He described the pain as dull in quality, diffuse in location, and exacerbated by eating, but not accompanied by vomiting, diarrhea, bloody or black stools. His medications included sulindac, cyclobenzaprine, doxazosin, omeprazole, and tramadol. His initial vital signs were normal, and his physical exam was significant for diffuse abdominal tenderness on palpation with voluntary guarding most pronounced in the right upper quadrant. His initial laboratory studies were significant for a leukocytosis with total bilirubin 1.4 mg/dL, alkaline phosphatase 347 U/L, AST 199 U/L, ALT 198 U/L, and γ -Glutamyltransferase 1589 U/L. His sulindac was discontinued due to the

possibility of NSAID hepatotoxicity. A conservative approach of bowel rest, pain control, and hydration was taken while awaiting further laboratory tests, including infectious and metabolic causes of mixed hepatotoxicity. The patient improved clinically over the next 4 days without further imaging or intervention, and several weeks later his hepatic function studies had nearly normalized.

Discussion: This case reinforces the importance of medication reconciliation during history taking, critical thinking, and the value of conservative medical management in patients with abdominal pain. NSAID-induced hepatotoxicity is relatively common and well-documented in the literature as the cause for about 10% of overall drug-induced liver injury. It is associated with moderate morbidity, but low mortality rates as it rarely leads to fulminant hepatic failure. Most patients don't even require hospital admission; therefore, early addition of this diagnosis to the differential in a patient on NSAIDs with acute abdominal pain can reduce unnecessary imaging, laboratory workup, and potentially days of hospital stay.

First Do No Harm: Corticosteroids and Recurrent Pericarditis

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Introduction: Recurrent pericarditis is a particularly troublesome complication of acute pericarditis and is seen in up to 30% of patients. Corticosteroid therapy is an independent risk factor for development of recurrent pericarditis. We present a case of recurrent pericarditis due to inappropriate use of corticosteroids to treat presumed viral bronchitis.

Case: A 43-year-old man without any past medical history presented to the urgent care clinic with cough, subjective fever, and wheezing. He was diagnosed with acute bronchitis and given tapering dose of prednisone and doxycycline. He came to the ED 1 week later with c/o substernal chest pain, fever, tachypnea, tachycardia, and pulsus paradoxus of 40 mmHg. CT showed significant left pleural effusion and large circumferential pericardial effusion. A diagnosis of symptomatic pleuropericarditis with effusions

was made. Echocardiogram confirmed large pericardial effusion. Pleural fluid studies were consistent with a transudate. Repeat echocardiogram showed improvement in pericardial effusion and patient was discharged home on ibuprofen and colchicine. He presented to ED 1 week later with worsening shortness of breath due to reaccumulation of pericardial effusion. A pigtail drain was placed in his pleural space with removal of over 1 liter of fluid. At this point, corticosteroids were introduced to treat his recurrent pericardial effusion. Autoimmune testing was equivocal. He was discharged on colchicine, ibuprofen, and prednisone.

Discussion: Recurrent pericarditis is a troublesome complication of acute pericarditis and occurs in 15% to 50% of cases. While corticosteroids traditionally have been used to treat acute pericarditis, it is now believed that treatment with corticosteroids during the index attack is an independent risk factor for development of recurrent disease. Corticosteroids can be used to treat refractory, recurrent, autoimmune, and uremic pericarditis.

Gemcitabine: A New Cause of Veno-Occlusive Disease

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Introduction: Veno-occlusive disease (VOD) is a rare complication of high-dose chemotherapy with significant mortality, most commonly seen in patients undergoing allogeneic stem cell transplantation. Gemcitabine is a common chemotherapy agent used in the treatment of various cancers, like non-small-cell lung cancer, pancreatic cancer, metastatic breast cancer, and in salvage therapy for lymphoma. Common side effects include myelosuppression, metabolic, pulmonary, and cardiac complications. This case illustrates 2 rare complications of gemcitabine: thrombotic thrombocytopenic purpura (TTP) and VOD.

Case: A 65-year-old woman with history of peripheral T-cell lymphoma was admitted with weakness and falls. Patient was noted to have progression of disease despite treatment with other chemotherapy regimens and was started on gemcitabine and dexamethasone.

On day 3 after administration, patient developed elevated liver enzymes, doubled total bilirubin, and elevated direct bilirubin, and chemotherapy was held. She soon developed worsening renal function and thrombocytopenia with schistocytes, indicative of TTP. Unfortunately patient's medical condition continued to worsen and bilirubin continued to trend up. Liver biopsy showed VOD. Patient received comfort care and died 2 days later.

Discussion: VOD is believed to be related to endothelial injury in liver venules; initially presenting with weight gain, ascites, tender hepatomegaly, and elevated bilirubin levels; and associated with renal failure. This case represents non-bone marrow transplant patient who developed rare side-effects of VOD and TTP in the setting of gemcitabine. This case demonstrates an established chronological relationship with gemcitabine and biopsy-proven VOD along with gemcitabine-related TTP diagnosed simultaneously. It is important to recognize VOD in the context of gemcitabine exposure, especially in patients with symptoms suggestive of VOD, even if they are not stem cell transplant recipients, as they must be managed aggressively given severe mortality associated with the disease.

Critical Illness Polyneuropathy

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Introduction: Development of neuromuscular weakness is a clinical finding often seen in patients admitted to the ICU. In these patients, the diagnoses of either critical illness myopathy (CIM) or critical illness polyneuropathy (CIP) must be considered. In certain cases, both of these diagnoses may be present.

Case: A 46-year-old woman presented with a 2-week history of dyspnea, cough, and fever. Her past medical history was remarkable for non-Hodgkin's lymphoma diagnosed 13 years ago and treated with radiation, chemotherapy, and bone marrow transplant. She subsequently developed renal failure and underwent renal transplant 3 years ago, for which she was currently on immunosuppressive therapy. In addition, she had a history of interstitial lung disease and had developed nontuberculous mycobacterial disease

6 months prior to admission. Initial evaluation demonstrated a systemic inflammatory response syndrome without a clear infectious source. Treatment in the ICU initially consisted of broad-spectrum antibiotics for presumed hospital-acquired pneumonia as well as stress dose steroids and IV fluid resuscitation, resulting in improvement in the her clinical status. Subsequent negative bacterial cultures and positive respiratory syncytial virus (RSV) NAAT led to discontinuation of antibiotics. Despite identification of the patient's infectious disease and improvement in other clinical parameters, her respiratory status worsened, leading to intubation with mechanical ventilation. Broad-spectrum antibiotics and IV methylprednisolone were initiated again with little to no improvement in her respiratory status and failure to wean mechanical ventilation over the next 9 days. She subsequently developed generalized flaccid weakness as well as absent reflexes. Nerve conduction studies (NCS) and EMG findings, along with the aforementioned clinical features, were consistent with a diagnosis of critical illness polyneuropathy. Continued attempts to wean mechanical ventilation failed, and the patient's clinical status continued to deteriorate. On hospital day 25, she had cardiac arrest and died.

Discussion: The diagnosis of critical illness polyneuropathy is made when a patient has signs, symptoms, and test results consistent with both CIM and CIP. CIM, which is often associated with IV glucocorticoids, presents with flaccid paralysis of all 4 extremities with preservation of sensation. In CIP, which is associated with severe sepsis, the patient has loss of sensory function in addition to muscle weakness and absent reflexes. EMG and NCS are used to confirm each diagnosis respectively. Muscle biopsy showing myosin loss in the setting of electrophysiologic evidence of axonal motor and sensory polyneuropathy is helpful in confirming the diagnosis. Recent studies indicate that an acquired sodium channelopathy may be the underlying cause for critical illness polyneuropathy. Management is aimed at the diagnosis responsible for the critical illness. Resolution of symptoms, if it occurs, takes weeks to months.

Dermatomyositis with Absent Skin

Findings

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Introduction: Dermatomyositis is an idiopathic inflammatory myopathy characterized by distinctive dermatological findings such as shawl sign, Gottron's papules, and heliotropic rash. Antisynthetase syndrome is a condition that presents with interstitial lung disease, arthritis, fever, Raynaud's syndrome, and myositis with anti-Jo1 antibodies. Diagnosis is established by elevated muscle enzymes, electromyography, and muscle biopsy.

Case: We report the case of a 38-year-old woman who presented with complaints of worsening cough, muscle pain, and weakness that began 1 week prior to admission. She had had previous multiple admissions for similar symptoms with uncertain diagnosis. Examination showed decreased strength in the proximal muscles, as well as diffuse muscle tenderness. Examination was negative for skin rash, Raynaud's phenomenon, or mechanic's hands. Initial chest CT showed severe scarring in bilateral lung bases. Serology was positive for ANA antibodies and anti-Jo1 antibodies, suggesting the diagnosis of inflammatory myopathy associated with interstitial lung disease. BAL was negative for infectious pathology. Muscle biopsy performed on day 4 of admission illustrated a clear inflammatory response, with variable fiber size, myophagocytosis, and perivascular muscle atrophy. The latter finding is considered to be pathognomonic for dermatomyositis. She was started on both prednisone and tacrolimus. Her condition improved significantly with this regimen, in addition to physical and respiratory therapy. She was discharged to acute rehabilitation therapy and scheduled for outpatient malignancy screening.

Discussion: Dermatomyositis without any of the characteristic skin manifestations is an uncommon finding. Patients with antisynthetase syndrome can present with myriad symptoms; however, these symptoms are not all required or frequently seen, posing a challenge for the clinician to diagnose. This diagnosis is crucial, however, as approximately 15% of patients with dermatomyositis have

an associated malignancy. Treatment and prognosis of patients with antisynthetase syndrome varies greatly depending on the type, severity, and progression of the lung disease. For patients such as ours with severe interstitial lung disease, systemic glucocorticoids with the addition of a second immunosuppressive agent are the treatment of choice.

Certolizumab-induced Cardiotoxicity in the Treatment of Ulcerative Colitis

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Case: A 28-year-old woman with a 13-year history of ulcerative colitis (UC) with significant clinical symptoms despite multiple conventional medical regimens was placed on infliximab therapy for 3 years with initial good response. Later, due to development of serum sickness, infliximab was replaced by adalimumab for 4 months without response, then certolizumab was used. However, her symptoms remained poorly controlled with persistent active pancolitis, elevated ESR (average 6.35 mg/dL), and CRP (average 42.6mm/hr). Over the following 6 months, she underwent 4 hospitalizations, including hospitalization for gram-negative bacillary sepsis and *C difficile* colitis. Finally, colectomy was performed. Fifteen months after initiation of certolizumab, during a hospitalization, she developed intermittent dizziness, palpitations, and left-sided chest pain unrelated to exertion. She had no orthopnea or dyspnea with exertion. Echocardiogram showed borderline enlargement of the left ventricle with severe systolic dysfunction (EF 30%-35%). She had no prior cardiac history and other causes of cardiotoxicity were excluded. Certolizumab was stopped due to the concern it may be the cause of cardiotoxicity. Seventeen months after stopping certolizumab, repeat echocardiogram showed a normal-sized left ventricle and only mild systolic dysfunction (EF 45%-50%).

Discussion: One of the most significant developments in the treatment of moderate to severe inflammatory bowel disease has been the class of biologics that are therapeutic antibodies against TNF-alpha. Certolizumab is a humanized pegylated anti-TNF-alpha antibody Fab fragment with clear efficacy in the

treatment of Crohn's disease with closure of draining fistulas, reduction of chronic glucocorticoid medication, and lasting remission. Recently, certolizumab also has been used in the treatment of UC. In addition to common side effects associated with immunomodulatory agents, cardiotoxicity has been reported with certolizumab. Although this complication is rare, it can be fatal. Therefore, monitoring cardiac function is critical when using any anti-TNF agent.

The Effects of Allergy Medications on the Evaluation and Diagnosis of Eosinophilic Esophagitis

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Introduction: Eosinophilic esophagitis (EoE) is an increasingly recognized cause of dysphagia and food impactions in adults. EoE is a chronic immune, antigen-mediated, esophageal disease characterized histologically by eosinophil predominant inflammation. EoE patients have atopy/allergy predisposition and frequently are treated with allergy medications prior to EoE diagnosis. We sought to evaluate the effect allergy medications have on diagnosis of patients with EoE.

Methods: A retrospective cohort of 51 patients diagnosed with EoE was enlisted over a 3-year period from dysphagia clinic. Use of allergy medication prior to EoE diagnosis was recorded. Each patient's endoscopic severity was graded on a 10-point scale. A pathologist blinded to the results recorded max number of eosinophils and histologic severity. Patients taking allergy medications (inhaled and nasal steroids, antihistamines, leukotriene antagonists) were compared to those not taking medications.

Results: Of the 51 patients in the study, 20 (39%) were on at least 1 allergy medication at the time of their diagnosis. There was a trend toward patients on allergy medications having a lower max eosinophil count (24.5 vs 31.5) on biopsy, although this did not reach statistical significance. There were more patients with <15 eosinophils on biopsy in the medication group compared to the non-medication group (35% vs 16%). There was a trend towards decreased endoscopic severity

in the medication group, but this difference did not meet statistical significance (3.2 vs 2.95). Subgroup analysis of patients on steroids also did not show significant differences in eosinophil count or endoscopic severity.

Acute Mitral Regurgitation: Another Great Masquerader?

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Introduction: Asymmetric or unilateral pulmonary edema is a rare cause of focal abnormalities on chest imaging, and it is frequently mistaken for more common conditions such as infection or malignancy. Acute mitral regurgitation is an important etiology of pulmonary edema and should be considered in any patient presenting with respiratory symptoms and abnormal lung imaging.

Case: An 87-year-old woman presented with dyspnea and hypoxia. Her chest CT revealed extensive consolidation and ground glass opacities predominantly in the right lung. She was treated with antibiotics for a presumed atypical infection without significant improvement. TTE showed mitral valve prolapse but no regurgitation. However, TEE demonstrated an extremely eccentric (explaining the lack of TTE findings), severe mitral regurgitation with a flail anterior leaflet due to idiopathic chordae rupture. The regurgitant jet generated more significant flow reversal in the right pulmonary veins than it did in the left, which could explain the unilateral chest findings. Myocardial infarction as an etiology was excluded. Other less common conditions associated with ruptured chordae, such as IE, blunt chest trauma, acute rheumatic fever, extensive mitral annular calcification, hypertrophic cardiomyopathy, and myxomatous disease, were unlikely. Due to advanced age and comorbidities, our patient chose medical management with afterload reduction rather than surgical valve repair.

Discussion: This case highlights the importance of recognizing the variation in clinical manifestations of acute mitral regurgitation. Acute mitral regurgitation is commonly misdiagnosed on presentation because the history and imaging findings may mimic an acute pulmonary process such as infection, acute pneumonitis, or acute respiratory distress

syndrome. Physicians may need to include acute valvular regurgitation in the differential diagnosis of any patient presenting with pulmonary manifestations, even with focal findings on chest imaging.

Hungry Bone Syndrome: How Much Calcium Is Enough?

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Introduction: After surgical correction of tertiary hyperparathyroidism with subtotal parathyroidectomy, aggressive bone remineralization and functional hypoparathyroidism can cause severe prolonged hypocalcemia known as "Hungry Bone Syndrome." As the following case illustrates, the duration of hypocalcemia is difficult to predict in this setting. Safe titration of calcium supplementation requires early identification of appropriate treatment targets.

Case: A 28-year-old woman with history of chronic dialysis-dependent ESRD presented with 4 days of weakness, dizziness, and paresthesias. She had undergone a subtotal parathyroidectomy 20 day prior due to severe tertiary hyperparathyroidism. Peak PTH prior to surgery was 3752 pg/ml; postoperatively it fell to 15-17 pg/ml. On admission she was noted to have tetany with serum calcium level of 5.2 mg/dL (ionized calcium 3.15 mg/dL). PTH on admission was 98 pg/ml. She was diagnosed with Hungry Bone syndrome and treated with a continuous infusion of IV calcium gluconate, oral calcium carbonate, and calcitriol. After 3 weeks, she was discharged on oral calcium and calcitriol; however, 48 hours later, she was readmitted with hypocalcemic tetany requiring reinitiation of continuous calcium infusion. After 3 more weeks of treatment with IV calcium infusion, she was discharged home successfully on oral therapy without recurrence of symptomatic hypocalcemia.

Discussion: Standard goals of therapy in Hungry Bone syndrome are to relieve symptoms and maintain low-normal serum calcium concentrations, typically 7.5-8.5 mg/dL. Rapidly progressive or symptomatic hypocalcemia after parathyroidectomy should be treated with 1-2g IV calcium gluconate followed by continuous infusion

of 0.5-1.5mg/kg/hr elemental calcium and 2-4g/day of elemental calcium orally. Serum phosphorus and magnesium should be monitored and replenished aggressively. Vitamin D deficiency should be corrected. Calcitriol is necessary in ESRD because of impaired 1-alpha hydroxylase activity. To prevent cardiac and neurologic complications, intensive monitoring and treatment must continue until symptoms are controlled and calcium levels are stable.

Dyspnea on Exertion and Cardiac Sarcoid

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Introduction: The lifetime risk of developing sarcoidosis ranges from 0.85% to 2.4%. Approximately 5% of patients with sarcoidosis develop clinically relevant myocardial involvement.

Case: A 59-year-old white man presented to his primary care provider (PCP) to discuss a 6-month history of worsening DOE. The patient's only medical history included hypertension (HTN). ETT was negative. He reported running 5 miles per day previously, but now had difficulty climbing 1 flight of stairs. TTE showed depressed left ventricular ejection fraction (LVEF) 40%, severely reduced RV systolic function with dilation and apical wall motion abnormality, severely dilated right atrium, and a small pericardial effusion. The PCP told the patient to present to the ED for pulmonary embolism (PE) evaluation. In the ED, he complained of worsening DOE and new left-sided chest pain radiating down his left arm and lasting a few seconds. An ECG noted normal sinus rhythm, 1AVB, RBBB, and anterior ST-elevation with inverted T waves. He had no acute complaints and his vital signs were within normal limits. A CT angiogram (CTA) chest ruled out PE. Basic labs were drawn in addition to a troponin, which was positive at 0.27ng/ml. The patient was admitted to the cardiology ward for further workup. He was stable overnight and his troponin peaked at 0.30ng/ml. The next morning, coronary catheterization showed nonocclusive coronary artery disease and cardiac MRI was consistent with cardiac sarcoid. The CT angiogram chest showed mediastinal and bilateral hilar lymphadenopathy consis-

tent with sarcoidosis. An ACE level was positive. The patient was discharged on steroids.

Discussion: This case illustrates the varied presentations of sarcoidosis and the importance of a thorough evaluation of new onset heart failure.

Autoimmune Lymphoproliferative Syndrome: A Case Report

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Introduction: Discriminating between self and foreign antigens is an integral component of immunity. One mechanism by which lymphocytes accomplish this is through FAS-mediated apoptosis. Ineffective apoptosis results in a rare genetic condition known as autoimmune lymphoproliferative syndrome (ALPS).

Case: J is a 26-year-old man whose clinical picture is consistent with this rare disorder. He first came to medical attention at age 5, when he presented multiple times with epistaxis, petechiae, and mucosal bleeding secondary to thrombocytopenia. He was found to have an autoimmune hemolytic anemia. These episodes responded to high-dose steroids. By age 14 he had had 3 episodes of shingles and an episode of severe onychomycosis. Subsequent T-cell and immunoglobulin analysis showed low absolute lymphocytes and IgA deficiency.

He remained well from age 16 to 23, when he was hospitalized with a subdural hematoma 3 days following a snowboarding accident. He was found to have a hemoglobin of 3 and a massive spleen. He was treated with splenectomy, rituximab, and a steroid taper with good response. In 2011 he presented with diffuse lymphadenopathy, leukocytosis, and fevers. Malignancy workup was negative. He had numerous hospitalizations in 2011-2012 for bleeding from immune thrombocytopenia and Coombs-positive hemolytic anemia. He became resistant to high-dose steroids, rituximab, and mycophenolate and required greater than 90 units of red blood cells. Bone marrow biopsies showed erythroid hyperplasia and a complete absence of megakaryocytes.

J meets the 2 required diagnostic criteria and 2 of the secondary accessory criteria for

ALPS based on the 2009 National Institutes of Health international workshop, giving him a diagnosis of probable ALPS. FAS mutation testing done by NIH was negative, placing him in the category of probable ALPS-U. Since diagnosis was made, he has been treated with antithymocyte immunoglobulin and cyclosporine and is again in a clinical remission. His history and negative FAS mutation studies raise the question to the existence of alternative mediators of apoptosis and self-recognition.

The Budd-Chiari Syndrome: An Important Consideration in New-Onset Ascites

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Introduction: New-onset ascites can be a diagnostic challenge, especially in previously healthy patients with no risk factors for liver disease. As this case demonstrates, the Budd-Chiari syndrome should be considered in any patient with new ascites and portal HTN as early recognition has important implications for treatment and prognosis.

Case: A 29 year-old woman presented to her PCP with 2 months of increasing abdominal distention and fatigue. She was concerned about possible pregnancy but home tests were negative. Physical exam was notable for marked abdominal distention, significant lower extremity edema, and spider angiomas with massive ascites confirmed by ultrasound. Upon admission, laboratory evaluation revealed total bilirubin of 3.1, mild transaminitis, an INR of 1.7 and a hemoglobin of 16.8. Ascitic fluid analysis showed serum-ascites albumin gradient of 3.25 consistent with portal HTN. Additional history obtained was negative for alcohol use, hepatitis risk factors, or family history of liver disease; serologic testing was unrevealing. CTA revealed splenomegaly, nodular liver suggestive of cirrhosis, and diffuse heterogeneous hepatic enhancement with non-visualized hepatic veins. Doppler ultrasound showed hepatic congestion with absence of hepatic venous flow, leading to a diagnosis of Budd-Chiari syndrome. Genetic testing confirmed the presence of the JAK2 (V617F) mutation and heterozygosity for Factor V Leiden. Subsequent bone marrow biopsy

revealed markedly hypercellular marrow (80%-90%) with panhyperplasia indicative of a myeloproliferative process and consistent with polycythemia vera. Treatment was initiated with anticoagulation, diuretics, and phlebotomy, with referral for consideration of liver transplantation given her advanced liver disease.

Discussion: Budd-Chiari syndrome results from any process that causes disruption of blood flow from the liver, but most commonly refers to thrombosis in the hepatic veins or inferior vena cava. It is a rare but important cause of ascites and liver disease. Myeloproliferative disorders are associated with up to 50% of Budd-Chiari syndrome cases, such as in this patient with previously undiagnosed JAK2-mutation polycythemia vera and Factor V Leiden heterozygosity. Treatment of underlying disorders is essential, highlighting the value of performing a comprehensive diagnostic evaluation for patients presenting with new-onset ascites and cirrhosis.

Emphysematous Pyelonephritis in the Setting of Poorly Controlled Diabetes Mellitus

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Introduction: Emphysematous pyelonephritis (EP) is a life-threatening necrotizing kidney infection characterized by intrarenal or perinephric gas seen primarily in diabetics.

Case: A 56-year-old man with non-insulin-dependent diabetes mellitus presented with a 2-week history of fever, chills, and right flank pain. Exam revealed an ill-appearing man in moderate distress with BP 109/49 mmHg, temperature 37°C, HR 85/min, respiratory rate of 25/min, and SaO₂ 99% on FiO₂ of 0.28. Abdominal examination revealed right-sided costovertebral angle tenderness. Laboratory studies showed WBC 35.8 × 10⁹/l, a serum glucose level of >750 mg/dl, pH 7.24, HCO₃ of 12 mmol/l, pCO₂ of 41 mmHg, potassium of 7.7 mmol/l, anion gap of 24, a creatinine level of 9.90 mg/dl and serum ketones were mildly elevated. Urine cultured *Escherichia coli*. CT showed gas in the right kidney, extending into the retroperitoneum, consistent with EP. Initial

treatment consisted of large-volume fluid resuscitation, insulin infusion and broad-spectrum antibiotics. An emergency nephrectomy was performed. A significant amount of purulent fluid was noted in the retroperitoneum and the kidney was partly necrotic. Postoperatively the patient required short-term dialysis but then improved and was off ventilator and vasopressors 36 hours from surgery.

Discussion: EP is an acute infection of the renal parenchyma that is most often observed in patients with poorly controlled diabetes mellitus (~95%) and is usually caused by *E coli* (70%). Depending on the severity of the infection, medical management plus either percutaneous drainage or nephrectomy is recommended. Risk factors associated with increased mortality include acute renal failure, thrombocytopenia, altered level of consciousness, and shock. CT is the diagnostic method of choice but a plain radiograph detects renal gas in the majority of cases. Even with early diagnosis and aggressive management, mortality remains high (~14%).

Myocardial Infarction, Deep Vein Thrombosis, Pulmonary Embolism, and In-stent Restenosis in a Patient with Heparin-induced Thrombocytopenia

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Introduction: Most commonly in heparin-induced thrombotic events, deep vein thrombosis (DVT) and PE comprise 25% of life-threatening events. Less commonly, arterial thrombotic events occur which can cause stroke, myocardial infarction, and limb ischemia.

Case: A 61-year-old man was hospitalized and placed on heparin for DVT prophylaxis. Ten days later, he had a ST-segment elevation myocardial infarction (STEMI) to his PRCA requiring a BMS. Two days later, he developed right lower extremity swelling. Doppler study showed extensive clot extending from the popliteal vein to the common femoral vein and patient was started on a heparin infusion (platelets 194,000). The next evening, he became bradycardic, tachypnic, and hypoxic. CTA chest revealed extensive acute bilateral pulmonary embolus (plate-

lets 41,000). The thrombocytopenia with advent of clotting led to the suspicion of heparin-induced thrombocytopenia (HIT). Therefore, heparin infusion was stopped, HIT antibody sent (which was later positive), and tissue plasminogen activator (tPA) was given followed by argatroban. Patient was intubated and hypoxia briefly improved after tPA administration. Later that night, hypoxia and hypotension worsened, requiring multiple vasopressors. Patient was sent to the cardiac catheterization lab for hemodynamic support; PRCA was 100% reoccluded. An aspiration thrombectomy was performed. Intraprocedurally, the patient went into complete heart block, which required placement of an intravenous pacer. Based on the angiographic appearance, the right ventricle and inferior base were not contracting. Therefore, an intra-aortic balloon pump (IABP) and a right ventricular assist device (RVAD) were placed. The right ventricle recovered enough to allow removal of the RVAD and IABP 3 days after its placement. Head CT 8 days after initiation of argatroban revealed an intraventricular hemorrhage. Argatroban was discontinued due to the intracranial hemorrhage and IVC filter was placed. The patient exhibited gradual improvement and was discharged home.

Discussion: This case demonstrates the morbidity of HIT, and reminds clinicians to have a high suspicion when thrombosis develops with thrombocytopenia.

A Case of Severe Anemia with Low Reticulocyte Count and Hemolysis

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Introduction: Anemia can result from blood loss, destruction of cells, or impaired production of cells. The first 2 causes often are accompanied by reticulocytosis, whereas the third is characterized by reduced reticulocyte count. We present the case of an anemic patient with a low reticulocyte count and evidence of hemolysis due to a unifying cause.

Case: A 48-year-old African American woman with a history of tricuspid valve repair presented with 1 month of generalized fatigue. Laboratory evaluation showed a hemoglobin 5.5 g/dL, mean corpuscular volume 109

fL, platelet count 80K, and normal WBC. Her reticulocyte count was 0.5%, and she had undetectable haptoglobin with LDH of 5439 U/L and total bilirubin of 1.8 mg/dL. B12 was 553 pg/mL (normal), and folate was also normal. Peripheral smear showed occasional fragments, ovalocytes, and teardrop cells. This constellation of findings raised concern for bone marrow dysfunction. Bone marrow biopsy revealed hypercellularity and megaloblastic red cells. Further lab testing showed significantly elevated homocysteine and methylmalonate (MMA) levels. She was treated with intramuscular cyanocobalamin and responded with a reticulocytosis and resolution of her macrocytosis and thrombocytopenia. She was found to have intrinsic factor blocking antibodies and was diagnosed with pernicious anemia.

Discussion: Pernicious anemia is a common malabsorptive cause of cobalamin deficiency. Cobalamin deficiency disrupts folate metabolism, affecting DNA synthesis and, consequently, cell division. Peripheral smear shows macrocytic ovalocytes and hypersegmented neutrophils, which are evidence of impaired hematopoiesis. Due to destruction of immature erythrocytes in the marrow, laboratory analysis may show elevated iron levels and evidence of hemolysis.

Cobalamin deficiency is distinguished from other types of megaloblastic anemia by measurements of the serum cobalamin, homocysteine, and MMA levels. Cobalamin levels above 300 pg/mL usually exclude deficiency, while levels below 200 pg/mL suggest deficiency. When levels are indeterminant, elevated homocysteine and MMA levels confirm deficiency, and high MMA level distinguishes B12 deficiency from folate deficiency. In this patient, cobalamin level was normal, but further testing was pursued given megaloblastic changes on biopsy. Treatment of pernicious anemia (diagnosed with anti-intrinsic factor antibodies) is lifelong B12 replacement.

Dress Syndrome in a Patient Undergoing Treatment for Prostate Cancer

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Introduction: Differentiation of Drug

Reaction with Eosinophilia and Systemic Symptoms (DRESS) from severe sepsis is essential due to the mortality associated with both syndromes. Yet definitions of DRESS remain in flux. We present a case of a patient with DRESS by the RegiSCAR scoring system currently undergoing treatment for prostate cancer to illustrate the difficulty in recognizing the syndrome, especially when a drug not usually associated with DRESS is the culprit.

Case: A 74-year-old man with prostate cancer was transferred for severe sepsis without identifiable source and worsening pancytopenia. The patient had received 2 leuprolide injections, with the second 2 months prior to presentation, and undergone a week of radiation 1 month prior. Three weeks later, he developed a confluent, blistering, pruritic rash over his entire body, including palms and soles. He subsequently developed nausea and vomiting, productive cough, fever, rigors, dizziness, dyspnea on exertion, fatigue, and tongue soreness. He was admitted and treated for sepsis and acute kidney injury. Overnight, he spiked fevers to 39.4°C with tachycardia and hypotension requiring pressor support. Following transfer, CBC showed pancytopenia with 24% eosinophils, CT suggested colitis, and skin biopsy demonstrated spongiotic dermatitis. Epstein-Barr virus was positive. Bone marrow biopsy showed hypercellular marrow with trilineage hematopoiesis. The patient required multiple transfusions for thrombocytopenia and anemia, and improved only with high-dose steroids. The patient had been taking allopurinol, a well-known cause of DRESS, for 4 years without incident, so suspicion turned to leuprolide, with case reports supporting this.

Discussion: DRESS is a life-threatening syndrome thought to be mediated by CD8+ T-cells and often including reactivation of a herpes virus. It can present with fever, rash, lymphadenopathy, eosinophilia, atypical lymphocytes, and involvement of the liver, kidney, heart, or other organ. With these symptoms, once infection has been ruled out, DRESS should be considered as a potential diagnosis.

Heparin-induced Thrombocytopenia and Thromboembolism in Lung Transplantation

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Introduction: Heparin-induced Thrombocytopenia (HIT) results in thrombotic complications in up to 50% of patients. We are not aware of previously reported cases of HIT-associated venous thromboembolism (VTE) in lung transplant recipients and present 3 patients who developed life-threatening VTE associated with HIT following successful lung transplant.

Cases: A 38-year-old man with idiopathic pulmonary fibrosis (IPF) who received a bilateral lung transplant was given routine postoperative DVT prophylaxis. He suffered cardiac arrest on postoperative day 12 and immediately received CPR but was asystolic for 32 minutes. Emergent pulmonary embolectomy was performed while receiving cardiopulmonary bypass. It became apparent that he had developed bilateral lower extremity DVT and a massive saddle pulmonary embolus (PE), and it was noted that the platelet count had fallen significantly over 2 days. Antiheparin antibody was positive, non-heparin anticoagulation was given, and an IVC filter placed. He completely recovered without sequelae.

A 51-year-old man underwent bilateral lung transplant for sarcoidosis. On postoperative day 14, he developed extensive PE in the left segmental pulmonary arteries and an upper extremity DVT that prompted the initiation of heparin therapy. He had a history of prior exposure to heparin, and a fall in platelet count prompted testing for antiheparin antibodies, which was positive.

A 64-year-old man with IPF underwent single lung transplant. He developed neutropenic fever and was found to have pulmonary aspergillosis at 1 year post-transplant and was admitted to the ICU. He developed bilateral lower extremity DVT and was started on IV heparin. An internal jugular vein thrombosis was detected 5 days later, and antiheparin antibody testing was positive.

Discussion: VTE can be a life-threatening postoperative complication in lung transplant recipients, and VTE/PE associated with HIT may occur. HIT should be considered in lung transplant recipients who develop VTE, especially if associated with a drop in platelet count.

Soft Tissue Presentation of Thyroid Cancer

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Introduction: Thyroid cancer is one of the most common, and often curable, malignancies. Atypical manifestations of thyroid cancer delay diagnosis and treatment, affecting prognosis. We present a case of metastatic follicular thyroid cancer, presenting as a nodule on the back.

Case: A 70-year-old woman presented with an asymptomatic “lump” on her back. CT chest showed a 9.3x4.7x5.4 cm lesion, extending from the right T8-T9 neural foramen to the posterolateral chest wall and involving the right ninth rib and T9 vertebra, consistent with a schwannoma, or neurofibroma. She was monitored by neurosurgery and later developed right paraspinal pain. Repeat MRI showed tumor growth causing new central canal stenosis. Biopsy was consistent with follicular thyroid cancer. Patient subsequently underwent thoracic laminectomy, corpectomy, and tumor removal with fusion of T6-T11 vertebral bodies. Pathology showed a solid pattern of growth in most of the tumor with rare colloid-filled follicles. TSH was normal with markedly elevated thyroglobulin and negative thyroglobulin antibody levels. Ultrasound revealed multiple thyroid nodules. She had a total thyroidectomy with pathology showing only a 10x7x7 mm focus of follicular carcinoma with capsular and vascular invasion limited to the thyroid. She underwent thyroid hormone withdrawal and radioactive iodine therapy. Post-treatment scan revealed iodine avid metastatic lesions in the spine, chest wall, and ileum. Patient was monitored with plans for radiation therapy should her disease progress.

Discussion: Thyroid cancer can have atypical manifestations, with a 4% incidence of

distant metastases in differentiated thyroid cancer at initial presentation. This is least common with papillary thyroid carcinoma (10%) and most common with Hurthle cell variants (33%). The overall long-term survival in patients presenting with distant metastases is 50%. Thyroid cancer is a common disorder, and we should have a low threshold to consider this disease in our differential of abnormal soft tissue lesions in the appropriate clinical context.

An Unusual Presentation of Merkel Cell Carcinoma

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Introduction: Merkel cell carcinoma (MCC) is a rare malignant neuroendocrine tumor of the skin. We describe a highly unusual initial presentation of MCC leading to acute fulminant liver failure and death in an elderly patient.

Case: An 87-year-old man with a history of prostate cancer treated only with the biologic agent bicalutamide for relief of lower urinary tract symptoms and no history of bony metastasis was admitted to the hospital after presenting with acute worsening of chronic low back pain, subjective lower extremity weakness, and dyspnea on exertion for the past several weeks. He notably had lost 10 pounds in the preceding 2 months, and had tender hepatosplenomegaly on examination. Labs were significant for new mild anemia, thrombocytopenia (96K), elevated liver transaminases, and marked hypoalbuminemia. Ultrasound of the right upper quadrant showed abnormal hepatic echotexture, concerning for an infiltrative disorder. Infectious workup was negative. Over the next several days, the patient's anemia and thrombocytopenia worsened, and peripheral blood smears showed leukoerythroblastosis, suggestive of a myelophthistic process in the bone marrow. A bone marrow biopsy demonstrated marrow involvement by metastatic carcinoma. The patient's liver and kidney function rapidly declined, and he began to bleed from his bone marrow biopsy site and upper GI tract. Intravenous steroids were given for symptom palliation. Due to his rapid clinical deterioration and previously stated wishes, his fam-

ily transitioned him to palliative measures only, and he died 6 days after admission. Immunohistochemical evaluation of the bone marrow biopsy demonstrated that the malignant cells coexpressed cytokeratin 20, neuron specific enolase, and synaptophysin without cytokeratin 7, CD117, or TTF-1, consistent with MCC. The primary site of disease was not identified.

Discussion: MCC usually presents as an asymptomatic, rapidly expanding pink or red tumor on the sun-exposed skin of elderly Caucasians and demonstrates a high propensity for recurrence and metastasis. While still considered rare, its incidence is increasing rapidly. Recent studies have implicated a newly identified Merkel cell polyomavirus in most MCC cases. The clinical presentation of fulminant hepatic failure and pathologic findings of marked bone marrow involvement without an identifiable primary site make this case an unusual presentation for MCC.

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Quality Improvement Organization

The Sunshine Act: It's For Real Now

Alyce C. Katayama, JD

The Sunshine Act, which became law on March 23, 2010,¹ requires manufacturers of drugs, devices, biologics and medical supplies to report to the US Department of Health and Human Services (HHS) certain payments and “transfers of value” to physicians and teaching hospitals (“covered recipients”). Payments to residents are not included in the reporting requirement. The Act also requires applicable manufacturers (AMs) and group purchasing organizations (GPOs), including those owned by physicians, to submit reports regarding ownership/investment interests held by physicians or their immediate family members. Final rules implementing the Act were published February 8, 2013.²

When Does It Become Effective?

The rules will be effective April 9, 2013. AMs and GPOs must begin gathering reportable information August 1, 2013. The first reports (covering August-December 2013) are due at HHS March 31, 2014. Reports will be processed, compiled, and placed in a searchable database for public view by September 30, 2014.

How Does My Name Get in the Database?

It's easy. All you need to do is receive, directly or indirectly, a single payment/transfer worth \$10 or more, or cumulative payments/transfers of \$100 or more in a calendar year, and your

name will appear in the database. Your principal practice address and specialty will appear as well. Both the form (cash, in-kind, stock, etc) and nature of the payment/transfer will be in the database. The dollar amount of ownership or investment interests will be reported.

The nature or purpose of the payment must be described in one of 16 mutually exclusive categories, including consulting fees, compensation for services other than consulting, honoraria, gift, entertainment, food and beverage, travel and lodging, education, research, charitable contribution, royalty/license, current or prospective ownership or investment interest, and grant.

What if I Speak at a Medical Education Program?

You may or may not end up in the database. Compensation for serving as a faculty member/speaker for an unaccredited/noncertified continuing medical education (CME) program is reportable, as is speaking at an accredited/certified CME program (in 2 separate nature of payment categories—to distinguish between accredited and non-accredited CME).

However, the compensation need not be reported if it fits into the exception created in the final rules. To make the payment nonreportable, all 3 requirements in the exception must be met: (1) the CME program is accredited by the Council for Continuing Medical Education, American Academy of Family Physicians, American Dental Association, American Medical Association, American Osteopathic Association (2) the AM does not pay the speaker directly; and (3) the AM does not select the covered

recipient speaker, nor give the program organizer a list of specific individuals to consider as speakers. Since these requirements are consistent with many professional societies' approach to industry support for CME programs, it is likely that a lot of that support will not be reportable.

Will I Be Listed Just for Taking Part in the Pharma-sponsored Coffee Break at a CME Program?

The final rule creates a special provision for allocating the cost of food and beverages where it is not possible to separately identify the individuals who participated. So, for example, if the drug rep brings a platter of bagels to your office, the value per person (for determining whether reporting is required) is calculated by dividing the cost of those bagels by the total number of participants, including not only the physician-covered recipients in the office, but also individuals who are not covered recipients, such as office staff. If a physician in the group did not actually share in the food offered (eg, because he was out of town that day), he will not be counted in the denominator for that calculation. Also, AMs are not required to report or track buffet meals, snacks, soft drinks, or coffee made generally available to all participants at a conference or similar large scale event. Even if the total value exceeds the \$100 annual aggregate reporting threshold, those transfers of value are not reportable.

What If I Don't Want to Be in the Database?

Payments/transfers below the \$10/\$100 thresholds are exempt from reporting, and these

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numbers will be adjusted for inflation. Further, no reports are required because you have ownership or investment interests in a publicly traded security or mutual fund and interests that arise from a retirement plan offered by a manufacturer to you (or member of your immediate family) through employment. Transfers of product samples (including coupons and vouchers) and educational materials intended to benefit patients, short-term loans of medical devices, educational services to patients, rebates, discounts, and items provided under warranty also are not reportable. Additionally, if you provide consulting or expert witness services to an AM in connection with a legal proceeding (civil, criminal or administrative), it is not reportable.

What Should I Do Now?

Review all of your relationships with AMs and GPOs. Make sure they will pass legal muster in the light of day. You can be sure the Office of the Inspector General and other regulators are

looking forward to mining this database.

If you receive payments/transfers from AMs or GPOs, keep track of them. You can then take advantage of your opportunity during the 45-day review period to make sure the information to be reported about you is accurate. You can review this information using online posting and notifications on CMS's list server, or you may register with CMS to receive notification about the review process. You will be able to login to a secure website to view information that has been reported about you. It is important to do this in a timely way since data corrected after the 45-day period by the AM/GPO will not be changed in the database until the following year. If your dispute on the accuracy of a report is brought to CMS's attention in a timely way, then even if it is not resolved before publication of the reports, CMS will mark the report as disputed. Unfortunately, if you have a dispute with the AM/GPO, it is yours to pursue and CMS will not assist.

It is likely that in the early implementation

of this massive program, there will be many glitches and hiccups. So you should be particularly vigilant starting this August and continuing at least through the publication of the first few annual data dumps.

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Editor's Note: *Wisconsin Medical Society policy ETH-004: The Relationship of the Profession to the Health Product Industry addresses the acceptance of gifts from those who provide health products prescribed by physicians, including the pharmaceutical and device industries. To read the policy, visit <http://bit.ly/Zsldlj>.*

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Jonathan Jaffery, MD



Robert N. Golden, MD

Accountable Care Organizations:

What are they? Where are they coming from? Where will they lead?

Jonathan Jaffery, MD; Robert N. Golden, MD

Accountable Care Organizations (ACOs) are receiving an increasing amount of attention, yet remain shrouded in mystery for many physicians and their patients. “No different from an HMO,” say some. “The answer to misaligned incentives inherent in the fee-for-service system,” say others, while a few compare an ACO to the Loch Ness Monster—everyone knows what it looks like, but no one has actually seen one.

Created under Section 3022 of the Affordable Care Act (ACA), the Medicare Shared Savings Program is a voluntary mechanism through which providers can contract with the Center for Medicare and Medicaid Services (CMS) to provide high-quality, cost-effective care to Medicare fee-for-service beneficiaries.

While the ACA clearly remains a political lightning rod, the ACO program has nonpartisan roots. In fact, Section 502 of the Patients' Choice Act of 2009 (HR 2520 and S1099), a bill introduced in the 111th Congress by Representative Paul Ryan (R-Wis) and Senator Thomas Coburn, MD (R-Okla), would have authorized a Medicare ACO Demonstration

Program that is remarkably similar in design to the actual legislation adopted in the ACA.

But what is an ACO? In some ways, the description is straightforward. An ACO is a group of providers who agree to accept responsibility for overall quality and cost of

care is less than projected, the ACO can share in a percentage of the savings.

Why would providers choose to join this program? One reason is to prepare for the future. While fee-for-service reimbursement, which rewards a high volume of activity irrespec-

We are deploying a new care model that we hope will deliver the most effective and efficient care for patients and populations, without requiring their physical presence in the clinic for all services.

the Medicare fee-for-service beneficiaries for whom they provide care. This “group” includes primary care physicians, at a minimum, but also may include specialists, hospitals, and other types of providers.

In the Medicare Shared Savings Program, quality is measured by 33 metrics defined by the Centers for Medicare and Medicaid Services (CMS), and cost is defined as the total cost of care to CMS. This last factor is key, and it points to a fundamental change from the status quo. Under this model, providers are at risk for all of the health care costs of their patients, not simply the costs for the services that they, themselves, provide. Throughout the contract period, CMS continues to pay fee-for-service claims as usual, but if the average total cost of

tive of quality or overall costs, has been the predominant payment mechanism for years, payers (including the government, commercial insurers, employers, and individual consumers) increasingly are insisting on value-based payments, which reward the highest quality and service and the lowest costs. While some may prefer the traditional payment system, most experts believe that maintaining the status quo is not a viable, long-term option. Early movement toward and engagement in value-based care may create the differential advantage that helps providers remain competitive in the marketplace.

Perhaps more importantly, this is an opportunity to add new capabilities to our practices. While critics of our country's health care system

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often emphasize discrete problem areas—such as inappropriate emergency department use, low generic prescription drug utilization, and redundant high-cost imaging studies—that hopefully are isolated and uncommon, some fundamental changes in how we approach health care delivery are necessary and underway.

The US health care system provides some of the best acute episodic and high-end specialty care in the world, but care coordination—especially for high-risk patients with chronic diseases or combinations of medical diseases, behavioral health issues and substance abuse—is largely lacking. Emerging payment mechanisms, such as ACOs, allow providers to invest in resources and activities that provide the full spectrum of care to their patients and re-orient their practices toward managing populations.

We know that employing nurses and social workers to help with care management can provide better patient-centered care while


reducing avoidable high-cost health care utilization for some high-risk patients. However, this costly investment results in decreased downstream revenue under the fee-for-service reimbursement model. By creating incentives to invest in care coordination services, shared savings models or other risk-sharing payment mechanisms, providers can make these fundamental changes to their practices without putting themselves out of business.

At UW Health, we think these investments are both the smart thing and the right thing to do. In 2012, UW Health established its own ACO (UW Health ACO, Inc) and—following a successful application in the most recent round of CMS competition—it now is an official Medicare Shared Savings Program.

To prepare for the launch of our ACO and for the “brave new world” of health care delivery, we restructured our primary care compensation models away from a system that rewards only volume of service, to a panel-based plan that encourages optimizing care for a popula-

tion. We are deploying a new care model that we hope will deliver the most effective and efficient care for patients and populations, without requiring their physical presence in the clinic for all services. Additionally, we are hiring a cadre of nurses and social workers who will coordinate and manage care for patients with complex, chronic diseases and high-risk social needs.

Of course, no one knows exactly what the future holds. But, currently, a number of new value-based payment mechanisms, including ACOs, are being tested. We predict that some of these will prove successful, others will fail to achieve their goals, and newer models will evolve. The importance lies not in any individual pilot, program, or payment mechanism, but rather in the hope that this evolutionary process will lead to a sustainable approach for providing the highest-quality, most cost-effective care to the patients and populations whom we serve.



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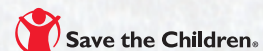
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