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Non-Clinical Aspects of Medical Practice:

A Pilot Project, Longitudinal Didactic Curriculum to Enhance Preparedness in the Practice of Medicine

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ON THE COVER

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Winner of the ASAE Excellence in Communications Award

THE Journal OF THE ARKANSAS MEDICAL SOCIETY

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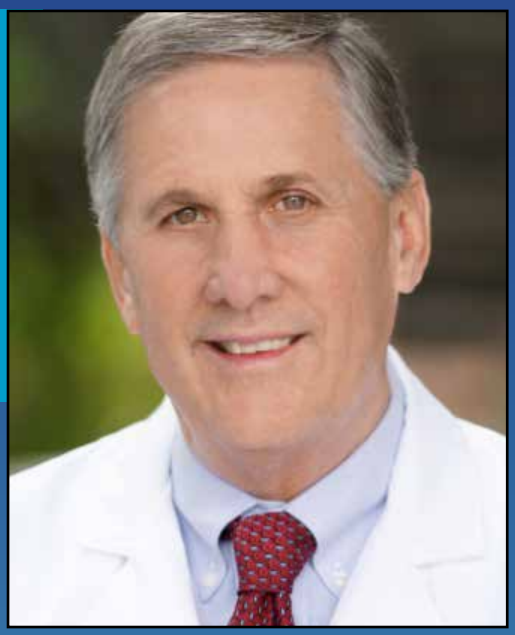
IABC arkansas BRONZE QUILL AWARD WINNER excellence Feature Articles

The Arkansas Medical Society would like to congratulate Scott Ferguson, MD

on his election to the Board of Trustees of the American Medical Association. Dr. Ferguson, West Memphis, has been an Arkansas delegate to AMA since 2009 and now serves as chair of the delegation. He is also Chairman of the AMA Council on Legislation and serves as vice chair of the AMA Radiology Section Council.

"I have spent my career fighting to preserve physician choice and autonomy, patient protections and the physician-patient relationship," said Dr. Ferguson. "As a member of the Board of Trustees, I can continue that fight and serve as an advocate for the AMA House of Delegates."



THE Journal
OF THE ARKANSAS MEDICAL SOCIETY

Tim Paden, MD

COMMENTARY

Count Your Blessings

Health Care, EMR, Outcomes, Data, Reimbursements

All those issues and others consume our days and our efforts. But have you been overseas, on mission trips? I joined a medical mission group from Texas to Haiti in 2016. Our mission field was located two hours up the coast from Port-au-Prince to a medical clinic serving the people of Gonaives and those in an orphanage in the desolate landscape. Flying into Haiti and traveling cross-country in an open-air bus; living behind concrete, guarded walls; and working at the orphanage and clinic with poverty-stricken, destitute patients ... is overwhelming at best. After adjusting to the environment and climate, you provide care at the best level you can with the supplies you brought, taking into account the ability of the patient. There are many of us in the AMS who have gone on similar journeys. The common thread among all these endeavors, I believe, is how it changes your perspective on your own life and your medical practice. When you consider the vast array of issues we deal with on a daily basis, it still comes down to a single point of focus: our patients' needs. Without the patient as the primary focus, we will lose our direction in this complex process we call health care.

I want us to consider two main points. First, are we holding to our values in the medical profession despite the pressures of society, legal issues, the health care climate, or the government influences? I do not know of any other profession with such high standards as the one in which we practice. As a new member to the AMS, my interest in joining centers on the direction our profession is likely to take if we do not stay engaged and apply our own pressure to the system.

Second, I ask us to count our blessings. If you have not taken a mission journey, I encourage you to sign up for one. In this country or another, there are multiple resources and locations that would gladly accept your contribution. The benefits far outweigh the time or commitments. Do you have EMR difficulties? Experiencing burnout like a large portion of physicians? Is the pay diminishing? Take a mission trip. It only takes a week.

I recall some patients in Haiti. One patient was an infant who suffered from microcephaly (felt to be a result of the zika virus). The mother came to the clinic every day for exam and nutrition advice. The life span of the infant was expected to be short, but the encouragement and advice were lifelong and priceless to this mother. Another patient, an American male, had multiple, recurring skin abscesses. He typically lanced them himself but one developed in the scrotum region so we assisted him on this one. I used local xylocaine 1%, which pleased him greatly as he usually did not have this available. We also provided him with sterile scalpels for him to use in the future on himself. Another patient, with multiple skin lesions simply required a large tub of Vaseline to hydrate his skin daily. If used sparingly, it would last him at least three months. This would likely impact his quality of life more than anything he had been given in the past several years.

When you look at your long list of patients to see today, or the long call hours, or the system of documentation required, just remind yourself of the resources available to you. It may be restricted or even denied in some cases, but we usually have some other avenue to pursue. I have a new perspective on medical practice with this experience under my belt ... and I don't plan on stopping. I need this again. AMS

Non-Clinical Aspects of Medical Practice:

A Pilot Project, Longitudinal Didactic Curriculum to Enhance Preparedness in the Practice of Medicine

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Objective

To develop a structured, broad-based longitudinal curriculum during neurology residency training to address the non-clinical aspects of medical practice (NCAM).

Methods

We developed and implemented an NCAM curriculum as part of the neurology residency didactic lectures. Residents were surveyed before and after the curriculum implementation.

Results

The mean readiness level across all domains and all years of training demonstrated a trend of increase after the introduction of the curriculum (3.08 SD 0.93 vs 3.3 SD 0.606, p= 0.46).

Conclusion

An organized curriculum focusing on NCAM may improve the level of resident trainees' preparedness in practice of medicine post-residency.

Introduction

The practice of neurology, and medicine in general, is rapidly changing. Besides medical knowledge, physicians now must also have knowledge of modern practice strategies. At the same time, contemporary training may not focus on the non-clinical aspects of medical practice. Often, new physicians struggle to find balance between functioning independently, medical school debt, and personal finances. According to a recent study by AMA Insurance, only 5% of young physicians consider themselves "very knowledgeable" about personal finance issues.[1] Fifty-five percent of nationwide graduating neurology residents in 2014 reported no business training during residency, and 53% felt unprepared for practice management tasks of their future jobs.[2] These lacunae are not only restricted to business aspects of medicine but also to domains such as professionalism, leadership skills, personal well-being, managing student-loans, etc.[2-5] There is limited data on how residency programs teach non-clinical aspects of medical practice (NCAM). There have been a few initiatives in different residency specialties.[6-10] Such initiatives have had restricted scope, not infrequently limiting themselves primarily to the business of medicine.[11,12] We identified the lack of a structured, broad-based longitudinal curriculum to address the NCAM comprehensively. In 2016, we instituted a structured, longitudinal curriculum that provides basic knowledge on the most important, what we called *non-clinical*, issues that physicians face in new practice. Physician burnout has come to the forefront lately, affecting 1 in 3 physicians in U.S.[13] Lack of work-life balance, regulatory burdens, and lack of autonomy seem to be important factors contributing towards physician burnout.[14] A well-structured program such as NCAM may contribute to fostering resilience and helping future residents develop strategies that will prevent burnout. Although our curriculum was geared primarily toward future neurologists, it can be tailored to any medical specialty.

Methods

We identified the subdomains of NCAM to be included in our longitudinal curriculum (Tab1). They were identified based on The Accreditation Council for Graduate Medical Education (ACGME) competency measures, non-structured department faculty interviews, structured anonymous resident survey with open ended questions, other models described in literature, and our own assessment of the needs of our trainees. The curriculum was executed as a part of the pre-existing, didactic lectures. These lectures were given by locally available experts in respective subject matters. The lecturers were provided a brief, summative overview of the desired scope and content but were allowed to design the lecture according to their expertise. Using established residency protocols, we ensure an ag-

gregate attendance rate of more than 80% of available residents during the didactic lectures.

This is an ongoing, educational-quality improvement project. Before the start of the curriculum, in October 2016, neurology residents (PGY2-4) were surveyed regarding their perceived preparedness for entering practice (supplement). The residents identified readiness in individual subdomains on the Likert scale ranging from 1 ("not prepared") to 5 ("very prepared"). The survey was repeated in March 2017 to follow up on the curriculum's impact. Statistical analysis was performed using GraphPad Instat software.[15]

The UAMS IRB Board assessed the project and determined it not to be a human subjective research, and exempted it from IRB review.

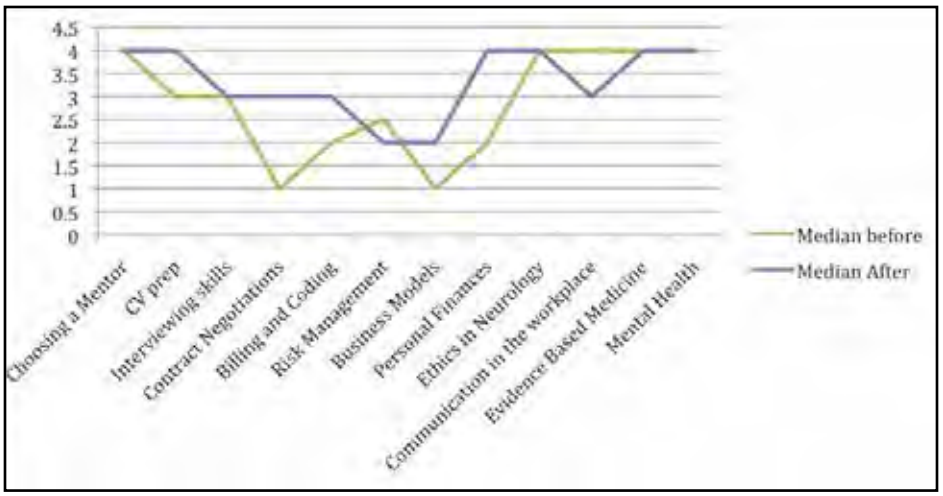
Results

Eleven neurology residents were approached to complete the survey. Pre-implemen-

> Continued on page 32

Supplement 1 Table. Summative results of Pre and post-implementation survey

	Mean	SD	n	25th Quartile	75th Quartile	Min	Median	Max	PGY-2 Mean	PGY-3 Mean	PGY-4-Mean
PRE-Mentor	4.00	0.82	7.00	3.00	5.00	3.00	4.00	5.00	3.67	4.00	5.00
POST-Mentor	4.20	0.45	5.00	4.00	4.50	4.00	4.00	5.00	4.00	4.50	4.00
PRE-CV PREp	3.71	0.95	7.00	3.00	5.00	3.00	3.00	5.00	3.00	4.00	5.00
POST-CV PREp	3.60	1.14	5.00	2.50	4.50	2.00	4.00	5.00	2.50	4.50	4.00
PRE-Interview skills	3.29	0.49	7.00	3.00	4.00	3.00	3.00	4.00	3.33	3.00	4.00
POST-Interview skills	3.40	1.14	5.00	2.50	4.50	2.00	3.00	5.00	2.50	4.00	4.00
PRE-Contract Negotiations	1.71	0.95	7.00	1.00	3.00	1.00	1.00	3.00	1.67	1.67	2.00
POST-Contract Negotiations	2.80	1.10	5.00	2.00	3.50	1.00	3.00	4.00	3.00	2.00	4.00
PRE-Billing & Coding	1.86	0.69	7.00	1.00	2.00	1.00	2.00	3.00	1.67	1.67	3.00
POST-Billing & Coding	2.80	0.84	5.00	2.00	3.50	2.00	3.00	4.00	2.50	2.50	4.00
PRE-Risk Management	2.67	0.82	6.00	2.00	3.25	2.00	2.50	4.00	2.33	2.50	4.00
POST-Risk Management	2.60	0.89	5.00	2.00	3.50	2.00	2.00	4.00	2.00	2.50	4.00
PRE-Business Models	1.43	0.79	7.00	1.00	2.00	1.00	1.00	3.00	1.00	1.67	2.00
POST-Business Models	2.20	1.30	5.00	1.00	3.50	1.00	2.00	4.00	1.50	2.00	4.00
PRE-Finances	2.86	1.77	7.00	1.00	5.00	1.00	2.00	5.00	1.67	3.67	4.00
POST-Finances	3.60	1.14	5.00	2.50	4.50	2.00	4.00	5.00	3.00	4.00	4.00
PRE-Ethics	3.57	0.53	7.00	3.00	4.00	3.00	4.00	4.00	3.67	3.33	4.00
POST-Ethics	3.80	0.84	5.00	3.00	4.50	3.00	4.00	5.00	3.50	4.00	4.00
PRE-Workplace communication	3.86	0.69	7.00	3.00	4.00	3.00	4.00	5.00	4.00	3.67	4.00
POST-Workplace communication	3.00	1.58	5.00	1.50	4.50	1.00	3.00	5.00	2.50	3.00	4.00
PRE-EBM	3.86	0.69	7.00	3.00	4.00	3.00	4.00	5.00	3.67	4.00	4.00
POST-EBM	3.80	0.84	5.00	3.00	4.50	3.00	4.00	5.00	3.00	4.50	4.00
PRE-Mental Health	3.71	0.95	7.00	3.00	4.00	2.00	4.00	5.00	3.33	4.00	4.00
POST-Mental Health	3.80	0.84	5.00	3.00	4.50	3.00	4.00	5.00	3.00	4.50	4.00
PRE-Aggregate									2.75	3.10	3.75
POST-Aggregate									2.75	3.50	4.00



Supplement 2 Graph: Median level of readiness across tested sub-domains of NCAM
KW = 15.896, p=0.1450

tation survey was completed by 7 (63.6%), and post-implementation survey was completed by 5 (45.5%). Results are presented in supplement 1, graph 1. The mean readiness level across all domains and all years of training demonstrated a nonsignificant increase after the introduction of the curriculum (3.08 SD 0.93 vs 3.3 SD 0.606, p= 0.46). Before the introduction of the curriculum, the various subdomains demonstrated statistically similar readiness levels. Exceptions included Contract Negotiations and Business Models (which demonstrated significantly lower median readiness levels than Choosing a Mentor, Evidence Based Medicine, and Communication in the Workplace, respectively (Graph 1). Post implementation, no significant difference was found in the readiness levels of the different subdomains. After introduction of the curriculum, there was a statistically nonsignificant trend towards increase in readiness level across most subdomains and along the year of training (Supplement 2). Within each subdomain, too, there was a similar trend towards increase in the readiness level after introduction of the curriculum (Supplement 1).

This pilot study was aimed at introducing a structured, longitudinal curriculum focused towards aspects of practice of medicine that are not routinely or usually covered during residency training consistently across the country.

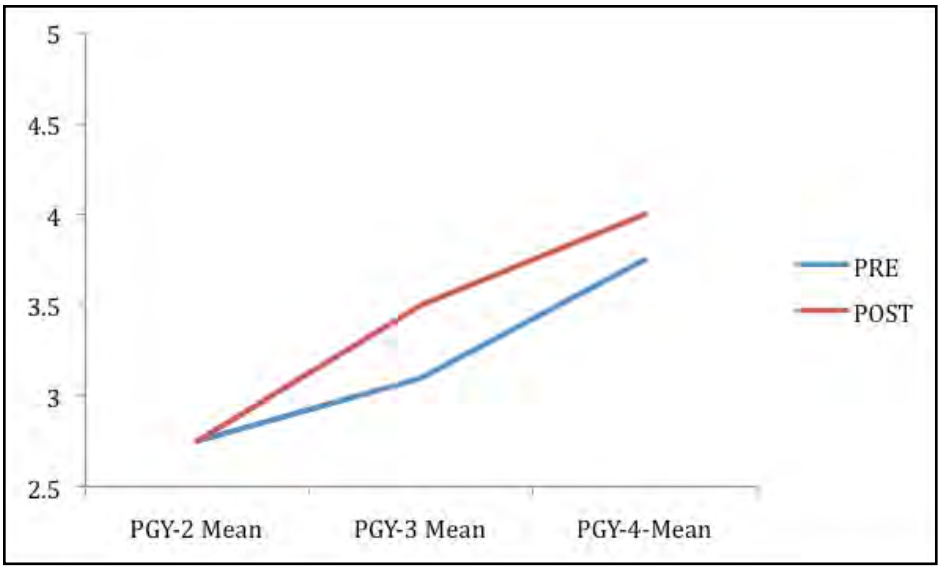
Discussion

This pilot study was aimed at introducing a structured, longitudinal curriculum focused on aspects of practice of medicine that are not routinely or usually covered during residency training consistently across the country. With the recent emphasis on physician burnout, especially in neurology, it is imperative that our trainees are equipped with tools for achieving a work-life balance in practice.[14,16] For the ease of communication, we chose the phrase *non-clinical* to describe these aspects, but they might not be entirely non-clinical.

The confidence level, and perhaps the knowledge level, of graduating residents in issues such as managing finances, starting a practice, and contract negotiation may not be as high as it should be. Several workers have tried to introduce various curriculum of somewhat restricted scope, commonly but not exclusively focusing upon business of medicine, with variable results.[6,8,10-12] We designed and implemented the curriculum change with broader scope, focused on practice in "real life." We identified the subdomains of interest by various strategies including, but not limited to, the survey of the residents, thus also incorporating the felt-need of the trainees. Before the implementation of the curriculum, most of the different subdomains included had similar perceived levels of readiness. At the same time, residents felt significantly lower readiness levels in a few subdomains (Business Models of Practice and Contract Negotiations) not routinely taught during residency. Although we were not able to demonstrate a statistically significant improvement in readiness levels by the introduction of our curriculum, there appeared to be a trend towards improvement (Graph 1, Supplement 2). The trend was seen across several subdomains. As might be expected, there was also an increase in aggregate readiness level with increasing level of training (Supplement 3). Our curriculum demonstrated a trend towards improvement in this aggregate readiness level in each level of training (Supplement 3).

Our project also has some limitations. This is a pilot project with a very small sample size limited to only one residency program. Our response rate was also low. As the surveys were anonymous, and not all residents participated in both the surveys, a true repeated measure analysis was not feasible. One may expect the level of readiness to be proportional to the level of training. This was also seen in the pre-implementation survey; hence, the increased readiness levels may simply be reflective of the general effect of residency training. At the same time, we saw a trend towards increase in all three levels of training; hence, the curriculum might have had a real impact on the level of readiness. We also did not have a control group. A bias may be introduced by the implementation of the curriculum itself as it may have led the residents to pay more attention to the NCAM. The survey instruments were subjective responses by the participating residents and no objective measures of improvement in skill were used. Such objective measures are neither widely available, nor easy to test.

To summarize, it seems that introduction of a structured, longitudinal curriculum may help in improving the readiness of resident trainees, but further research is warranted in designing such comprehensive curriculum, its implementation and ultimate effect on comprehensive practice



Supplement 3 Graph: Mean summated level of readiness per year of training

of medicine, and achievement of work-life balance in the real world.

Conclusion

An organized, longitudinal curriculum comprehensively focusing on NCAM may improve the level of resident trainees' preparedness in practice of medicine after residency. Further research in design, implementation and influence of such a curriculum is warranted.

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CASE STUDY

Youth-Onset Type 2 Diabetes

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Abstract

Youth-onset Type 2 diabetes is increasing at an alarming rate, in part due to the obesity epidemic. Type 1 diabetes and youth-onset Type 2 diabetes differ vastly in nature, and appropriate treatment regimens are necessary to preserve beta cell function and prevent complications. Guidelines set forth by experts should be followed for screening, diagnosis, and treatment. Children found to be at risk for, or have youth-onset Type 2 diabetes should be managed by a pediatric diabetes care provider. A multidisciplinary approach is necessary to provide complete, appropriate education.

Manuscript

Youth-onset Type 2 diabetes is more common in the pediatric population as obesity rates are on the rise. Due to the prevalence of obesity, differentiating Type 1 diabetes from youth-onset Type 2 diabetes is becoming increasingly difficult. Treatment regimens differ vastly, so appropriate recognition and management is crucial. Type 2 diabetes has been described as insulin resistance and non-autoimmune beta cell dysfunction leading to hyperglycemia. In contrast, Type 1 diabetes results from beta cell destruction, generally leading to absolute insulin deficiency.

For the overweight child, the American Diabetes Association recommends screening with any of the two following risk factors: family history of Type 2 diabetes in a first or second degree relative, race, or ethnicity with increased risk (Native American, African American, Latino, Asian American, or Pacific Islander), signs of insulin resistance or conditions associated with insulin resistance (hypertension, dyslipidemia, acanthosis nigricans, polycystic ovarian syndrome, small for gestation birthweight, or maternal history of gestational diabetes during the child's pregnancy). It is important to note that youth-onset Type 2 diabetes can exist in the non-obese child. Screening should begin at the age of 10 or at the onset of puberty (the earlier of the two) and be carried out every three years unless signs present sooner. However, documented cases have occurred at a much younger age. Diabetes should be suspected and ruled out in the primary care setting in children who have any of the following complaints, regardless of weight: increased thirst (polydipsia), increased urination (polyuria), nocturia, enuresis, blurry vision, persistent headaches, unintentional weight loss/failure to gain weight, recurrent yeast infections (diaper candidiasis, vaginal candidiasis or thrush), or recurrent abscesses.

Table 1

Diabetes Diagnostic Criteria	Prediabetes (Increased Risk for Diabetes)
FPG=/> 126 mg/dl	FPG =/> 100mg/dl-125mg/dl
2HR OGTT =/> 200 mg/dl	2HR OGTT 140 mg/dl-199 mg/dl
A1C =/> 6.5%	A1C 5.7%-6.4%
Random glucose + Sx =/> 200 mg/dl	

The first step in diagnosis should include a thorough history and physical assessment with attention to the above signs and symptoms, followed by an appropriate laboratory work up. The initial analysis should include finger-stick glucose for immediate evaluation (diagnosis must be based on venous stick levels unless there are overt symptoms), comprehensive metabolic panel, Glycosylated Hemoglobin A1C, and urinalysis to assess for ketones and glucose. Of note, a random blood sugar level > 200 mg/dl in an asymptomatic child should be repeated immediately prior to the diagnosis of diabetes. If these levels are suspicious for diabetic ketoacidosis (DKA) or if the child appears sick, the child should be sent to a local emergency room for further management.

Diagnostic evaluation in the stable patient can be performed in the following ways: fasting plasma glucose (FPG), random venous glucose, two-hour oral glucose tolerance test (2HR OGTT) with 75g carbohydrate load and venous A1C levels. Table 1 outlines diagnostic criteria for both Type 1 and youth-onset Type 2 diabetes, as well as 'Prediabetes.' Testing for prediabetes, or increased risk to develop diabetes should be considered in patients meeting the criteria to screen for youth-onset Type 2 diabetes.

Any child who is found to have youth-onset Type 2 diabetes with a random glucose of 250 mg/dl or greater or an A1C of 9% or greater should immediately be started on insulin under the supervision of a specialist and care should be transferred to a pediatric diabetes care provider. Children who will require multiple daily injections should be admitted to a pediatric inpatient setting to receive appropriate, pediatric fo-

cused, multidisciplinary care, and education. In the child who is not symptomatic or significantly hyperglycemic with a lower A1C (<9%), in whom Type 1 diabetes can be reliably excluded; it is acceptable to start an alternate mode of therapy such as oral medication or to consider a 24-hour insulin with close follow up with the pediatric diabetes care provider. These cases should be discussed with the specialist for management and dose recommendations. In this situation, the family must be reliable and have had education on blood glucose monitoring, insulin administration (if prescribed), and assessing the urine for ketones and hypoglycemia management including glucagon administration when insulin is utilized. Mildly elevated A1C levels may benefit from Metformin or lifestyle modification alone. It should be noted that any child in whom Type 1 diabetes cannot be ruled out should be admitted to the pediatric hospital under the care of a pediatric diabetes provider for multiple daily insulin injection initiation, glucose monitoring, and comprehensive diabetes education.

Differentiating Type 1 diabetes from youth-onset Type 2 diabetes can prove difficult for the primary care provider. Laboratory analysis to distinguish between the two types should include C-peptide and pancreatic auto-antibodies. Although C-peptide levels may be low in very late progression of Type 2 diabetes, presentation should yield normal to high levels in this population. Diabetic ketoacidosis with associated kidney injury can falsify C-peptide results. Approximately half of C-peptide is excreted by the kidneys and a decrease in kidney function would yield falsely elevated results. Positive pancreatic auto antibodies, which can take weeks to result, strongly suggests a diagnosis of Type 1 diabetes and in most cases can rule out youth-onset Type 2. History, initial presentation, physical exam, and immediate lab values should help with the initial diagnosis of Type 1 vs Type 2. Clinical course and response to treatment should guide ongoing care. Any uncertainty in plan of care should be directed immediately to a pediatric diabetes care provider. In comparison with adults, children with youth-onset Type 2 diabetes have a more rapid decline in beta cell function as a result of hyperglycemia, which emphasizes the need to start insulin in a timely manner for those meeting the criteria.

Insulin and Metformin remain the only two FDA-approved drugs for youth-onset Type 2 diabetes. Metformin should be started after acceptable kidney function has been documented with normal-for-age blood urea nitrogen and creatinine levels. Dosage should begin at 500mg once or twice daily, with meals and increase to a goal of 1 gram twice daily as tolerated. Families should be educated on potential side effects and to stop if the child becomes dehydrated, has ketones, or is in need of a radiologic or surgical procedure. Children requiring insulin may benefit from the addition of Metformin to their regimen-especially if there is overt insulin resistance. DKA is present at the time of diagnosis in approximately 6% of children found to have youth-onset Type 2 diabetes. For the child who presented in the setting of DKA, ketones should be cleared before starting this medication.

Type 2 diabetes is insidious in onset; therefore, some patients already have comorbid conditions at the time of diagnosis. Children should be screened for hypertension with routine, accurate blood pressure readings at diagnosis and at each visit thereafter. Near the time of diagnosis, they should also be evaluated for dyslipidemia, nonalcoholic fatty liver disease, random urine for microalbumin to creatinine ratio, dilated eye exam to rule out diabetic retinopathy, and a comprehensive foot exam. Screening should take place yearly. Females of child-bearing age should be strongly cautioned of fetal anomalies that can develop with uncontrolled hyperglycemia. Sleep disturbances should also be assessed and referral to a sleep specialist may be necessary. Children with youth-onset Type 2 diabetes are not excluded from developing metabolic syndrome and each disease process should be meticulously managed.

Routine follow up with a pediatric diabetes care provider should take place every three to four months with close monitoring of the A1C level. A goal A1C level of 7.5% or lower is recommended for all ages, and of course this should be individualized to each patient. Care should be taken to avoid severe hypoglycemia in those receiving insulin. Children with youth-onset Type 2 diabetes who are being treated with lifestyle modification or Metformin alone are at a much lower risk for hypoglycemia; therefore, a lower A1C level is acceptable. The pediatric population requires a multidisciplinary approach including the diabetes care provider, certified diabetes educator, registered dietician, physical therapist, social worker or psychologist and child life specialist where available. Considering the child will spend several hours of the day at school, the family should be provided guidance on communicating needs with the school and the school nurse should be involved in care. Once the patient is nearing adulthood, transition to an adult diabetes care provider should be a structured process.

> ***The first step in diagnosis should include a thorough history and physical assessment with attention to the above signs and symptoms, followed by an appropriate laboratory work up.***

In summary, youth-onset Type 2 diabetes is increasing in incidence as obesity is more prevalent. Appropriate management is critical to preserve beta cell function and reduce diabetes related complications. Each regimen should be individualized to meet the needs of the patient and the family. This is a complex and difficult process even for the most experienced program, and new approaches are published frequently. Therefore, education and support should be provided with an experienced and multidisciplinary approach.

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Infant Hearing Loss: A Neurodevelopmental Emergency

BY ALAN MEASE, MD, FAAP

Two or three of every 1,000 children in the United States are born with some degree of hearing loss. Even more will lose their hearing during infancy or childhood. The most important period of speech and language development occurs during the first three years of life, when the brain is constructing nerve pathways necessary for understanding speech.

Hearing is a brain function and a baby's brain is "pre-wired" to accept and process sound. Babies with typical hearing begin hearing before birth, at 20 weeks' gestation. At birth, babies prefer listening to their mother's voice, their native language, human speech rather than noise, and songs or stories heard before birth.

Hearing loss in babies is a "neurodevelopmental emergency." The brain is the true organ of hearing; the ears only transmit sounds to the brain. Babies born with hearing loss are not starting from the same point as a baby with normal hearing. They have missed 20 weeks of typical development of their auditory pathways before birth. They will miss the auditory neural development that occurs after birth, before hearing loss is diagnosed. They will miss the typical development of auditory brain pathways that could have occurred after birth, until the child begins hearing sounds consistently by wearing hearing aids during all waking hours.

A baby's brain must be exposed to meaningful sounds consistently for auditory neural pathways of multiple, spiral, ganglion neurons in the brain to develop.[1] If a baby does not hear sounds well or is exposed to only a little sound or speech during his or her early years, then a permanent re-assignment of the child's auditory brain cells occurs. If the brain is not stimulated by sound, it will reorganize itself through synaptic pruning to maximize processing through other senses, primarily vision. After about three-and-a-half years of age, the brain has considerably less flexibility to develop effective skills to process auditory information. This is why children with hearing loss will experience difficulty learning to listen and speak proficiently.[2]

In 1999, the Arkansas General Assembly passed Act 1559, implementing early detection of hearing loss at all birthing facilities. This law resulted in Arkansas' Early Hearing Detection and Intervention (EHDI) program. EHDI is responsible for successfully screening 98.4 percent of all babies born in Arkansas. Screening identifies about 50 babies per year with hearing loss. Unfortunately, only 15 percent of them receive early intervention services by six months of age. The Centers for Disease Control and Prevention (CDC), American Academy of Pediatrics, EHDI and the Joint Commission on Infant Hearing (JCIH) created consensus guidelines to have all babies receive early intervention by six months of age.[3]

Timing is critical to optimize outcomes. Babies with hearing loss identified in the first weeks of life, and who begin hearing optimally no later than six months, have a good chance of developing neural connections in their auditory brain pathways necessary to lay the foundation for spoken language development. This is especially true if they are provided with enhanced listening experiences. The best predictors of verbal language skill development are the child's age when full-time hearing aid use started, the degree of hearing loss and the amount of exposure to meaningful listening experiences. Hearing aids, FM systems and cochlear implants are "brain access" tools. To take advantage of the critical period of optimal auditory brain development, the ability of the brain to perceive as much sound as possible must be provided as soon as possible after birth.

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Using technology, hearing ability must be provided as close as possible to typical hearing level, if the child is to learn to listen and use spoken language. The child's family may choose one of five language education programs:

- **Auditory-oral:** teaches child to use residual hearing by using hearing aids or cochlear implants plus speechreading (lip reading); no sign language
- **Auditory-verbal:** includes above plus teaching parents to help child become auditory communicator; no speechreading or sign language
- **Bilingual-bicultural:** teaches child to use American Sign Language (ASL) as first language and English as second; deaf culture is taught with ASL as common language
- **Cued speech:** teaches child how to see and hear spoken language
- **Total communication:** teaches combination of all methods plus ASL

Parent involvement is critical to finding the best choice for each child. Language education programs must start by six months to maximize a child's ability to learn language. (**https://www.cdc.gov/ncbddd/hearingloss/freematerials/Communication_Brochure.pdf**)

To accomplish the goal of early intervention by six months, we must:

- Screen all babies for hearing loss before one month of age
- Ensure that babies who do not pass the screen receive an audiologic evaluation no later than three months (**https://www.cdc.gov/ncbddd/hearingloss/screening.html**)
- Enroll babies with confirmed hearing loss in early intervention services no later than six months

A major obstacle to these goals is the delay caused by repeating the failed screening exam after one month of age, before receiving complete audiologic evaluation. Any baby older than one month who has failed the hearing screen should have an expeditious audiologic evaluation to confirm or rule out hearing loss. This will allow early diagnosis and intervention services to begin no later than six months. Delays in early intervention result in permanent spoken language delays. Evidence indicates that many children with sensorineural hearing loss experience improved language abilities with early intervention.

Children with hearing loss are at risk not only for lifelong deficits in speech and language acquisition, but poor academic performance, personal-social maladjustments and emotional difficulties.

Children with hearing loss are at risk not only for lifelong deficits in speech and language acquisition, but poor academic performance, personal-social maladjustments and emotional difficulties. In addition to developmental delays, these children may have behavioral problems such as attention deficit/hyperactivity disorder, autism or learning disabilities. They should have regular surveillance of developmental milestones. The CDC's Learn the Sign Act Early (LTSAE) materials and smart phone Milestone app are useful tools to teach parents how to monitor their child's development. (Free download: **www.cdc.gov/MilestoneTracker**) Patients may find this simpler tool (**https://afmc.org/product-category/practices/epsdt-well-child-practices/**) more helpful. When delays are detected in any domain, the child's primary care provider (PCP) should do a complete developmental screening with a tool like the Ages and Stages Questionnaire.

While virtually all babies born in Arkansas are screened for hearing loss, those who fail the screen are not being diagnosed in a timely manner. We are failing to provide early intervention services by six months for all babies diagnosed with hearing loss. Delays are never acceptable. All PCPs, otolaryngologists, audiologists and early intervention providers must work together to be sure that all of Arkansas' babies with hearing loss get timely, early-intervention services.

Hearing loss in babies is a neurodevelopmental emergency. Any delay in auditory stimulation, or a reduced auditory signal during the optimal developmental stage, may cause permanent, irretrievable reassignment of auditory brain cells. There is a limited window of time during which babies can catch up to their normal-hearing peers. ▲

Dr. Mease is Medical Director, Child and Adolescent Health, Arkansas Department of Health.

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An Unusual Stroke-Like Presentation of HSV Encephalitis

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Keywords: encephalitis, stroke-like, HSV, frontal lobe.

Abstract

HSV (Herpes Simplex Virus) encephalitis is a potentially life-threatening illness that can affect neonates as well as adults.[1] Despite improvement in diagnostic techniques such as magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) examination, challenges and pitfalls remain in the diagnosis of this condition.[1] We are reporting a case of HSV encephalitis that presented like a stroke with atypical MRI and CSF findings. The possibility of HSV encephalitis in a patient with fever and focal neurological deficit should always be kept in mind because a full-blown picture such as seizures, abnormal behavior, confusion, disorientation, etc., may not be seen in every patient.[2]

Case Report

A 69-year-old woman, otherwise healthy, presented to the emergency department with a sudden onset of headache, numbness, and weakness in the left upper extremity. Her presentation was suggestive of an acute ischemic stroke, but intravenous thrombolysis was not considered as she arrived outside of the window period. Neurologic examination revealed weakness and hyperreflexia in left upper extremity. MRI of the brain (stroke protocol) demonstrated hyperintensity on diffusion-weighted imaging (DWI) in the right frontal lobe corresponding with the area of weakness (Figure 1). On hospital day two, she developed a fever followed by urinary incontinence. Urinalysis revealed leukocyturia (50 white blood cells) and dipstick was positive for leukocyte esterase. Intravenous ceftriaxone was initiated empirically to treat urinary tract infection. On hospital day three, she had a generalized tonic-clonic seizure followed by recurrent focal seizures involving the right side of face and arm. Over the next few hours, her mental status worsened and she was barely responsive to stimuli. Her EEG showed bilateral independent periodic lateralized epileptiform discharges (BIPLEDs). At this point, it was decided to repeat brain imaging and perform a lumbar puncture.

> ***Initially, HSV encephalitis affects one hemisphere and involves the contralateral side once it has extended in the initially involved hemisphere.[4]***

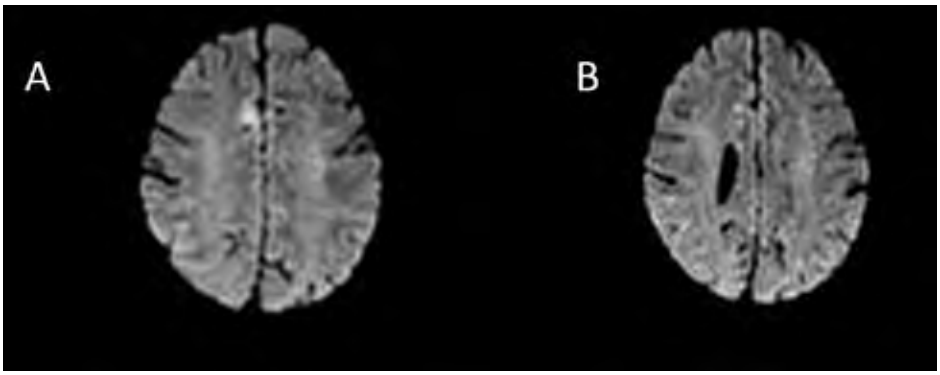

Figure 1. *Demonstrates hyperintensity on DWI sequence (A and B) in the right parafalcine region.*

CSF showed three white blood cells/µl, protein was 39 mg/dl, and glucose was normal. Cultures were negative for bacteria, fungi, and mycobacteria after six weeks of incubation. The detection of herpes simplex virus (HSV) type 1 DNA in CSF using polymerase chain reaction (PCR) confirmed the diagnosis of HSV encephalitis. Brain MRI showed the new development of bilateral fluid attenuated inversion recovery (FLAIR) hyperintensities involving the parafalcine frontal, temporal, anterior cingulate, and insular region (Figure 2). Complete blood count, lipid profile, hemoglobin A1c, blood culture, urine culture, and echocardiogram were either normal or negative. Autoantibodies in the serum against N-methyl-D-aspartate (NMDA) receptor, alpha-amino-3-hydroxy-5-methyl-4 isoxazolepropionic acid (AMPA) receptor, gamma-aminobutyric acid (GABAb) receptor, leucine-rich glioma inactivated-1 (LGI-1), and contactin-associated protein-like 2 (CASPR-2) were negative. After treatment with IV acyclovir, 10mg/kg every eight hours for 21 days combined with IV methylprednisolone, 1000 mg a day for five days, she improved dramatically, with only residual bilateral weakness in lower limbs and occasional partial seizures.

Discussion

This patient presented with a sudden onset of focal neurological deficit without any change in the mental status. Her initial MRI and CSF picture was not typical for HSV encephalitis. In this case, the diagnosis of HSV encephalitis was

> Continued on page 40.

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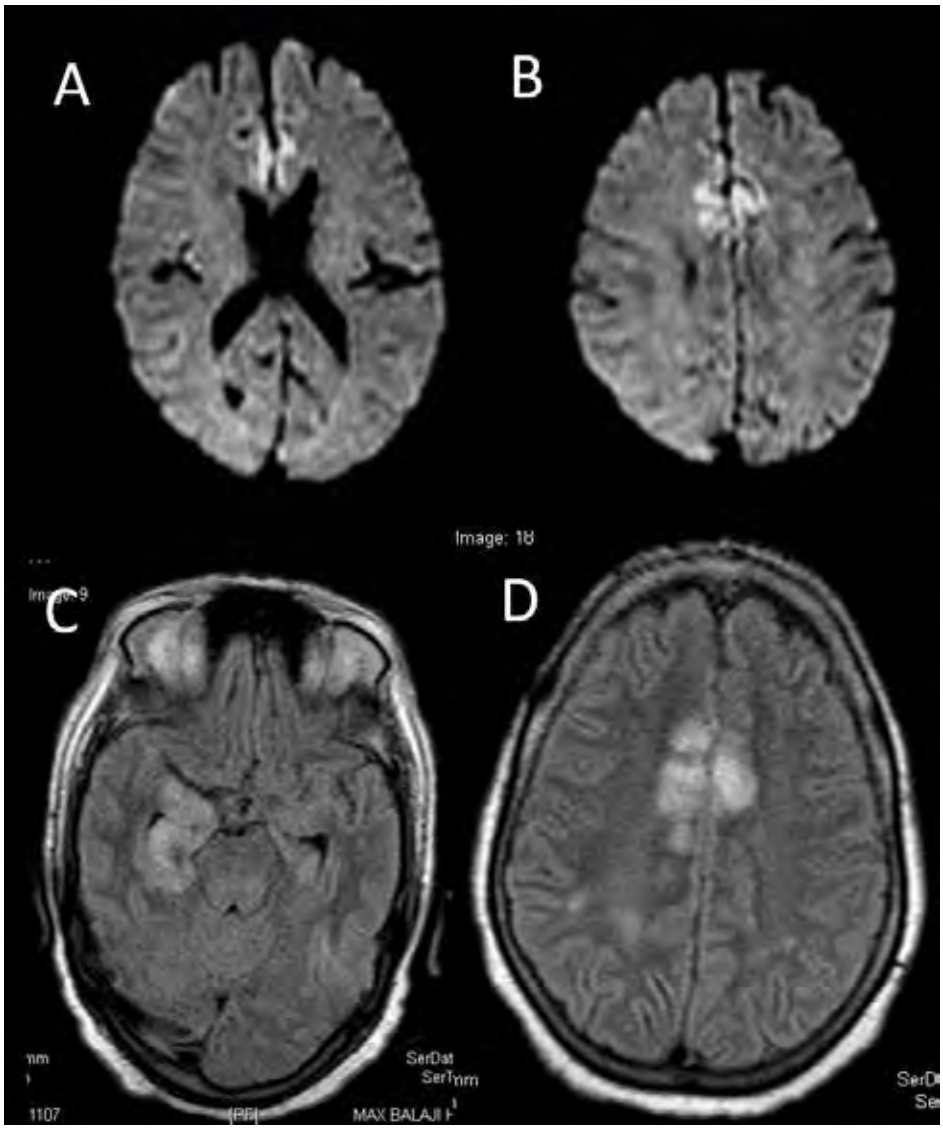

Figure 2. *Demonstrates hyperintensity on DWI sequence (A and B) in bilateral parafalcine area. FLAIR hyperintensity is also seen in bilateral temporal and parafalcine areas (C and D).*

clinched by the amplification of HSV type 1 DNA in CSF and improvement with Acyclovir. Earlier, brain biopsy was considered to be the gold standard for the diagnosis of this condition, but this has now been replaced by HSV PCR detection in CSF.[3] Despite being very sensitive, HSV PCR can be negative if CSF is obtained very early in the disease course; the testing should be repeated if the suspicion is very high.[3] The measurement of intrathecal synthesis of immunoglobulin (IgG) after one week into the disease course has been suggested if the HSV PCR in CSF continues to remain negative.[3] Despite the fact that brain imaging can be initially normal in 5-10% of cases[1], Renard et al showed that DWI (diffusion-weighted imaging) identified more areas of involvement than FLAIR sequence when MRI is performed early in the disease course.[4] Initially, HSV encephalitis affects one hemisphere and involves the contralateral side once it has extended in the initially involved hemisphere.[4] Brain MRI is not helpful to assess the response to therapy as the lesions can progress despite treatment.[1] The temporal lobe is most commonly as well as initially affected with HSV encephalitis but isolated extra-temporal forms of HSV encephalitis have been reported.[5] The frontal or parietal lobe can also be initially involved with HSV encephalitis, which can potentially delay the diagnosis of this catastrophic condition.[5] EEG when used alone is not helpful for making a diagnosis as non-specific changes are seen.[1] HSV encephalitis can present with a sudden onset of symptoms which can mimic a stroke.[6] It is possible that sudden deficits represent an ictal phenomenon or hypoperfusion of the involved area.

Early recognition of HSV encephalitis is important as it can unleash a variety of complications such as cognitive deficits, seizures, etc.[1] It is one of the neuroinfectious diseases where timely institution of treatment can prevent morbidity and mortality. There are several learning points in this case, namely the sudden onset of symptoms, delayed appearance of fever and partial seizures, initial frontal lobe involvement on brain imaging, and atypical CSF findings. The combination of brain MRI, CSF, and EEG should be used in cases of diagnostic confusion because it improves the sensitivity of diagnosis.[1]

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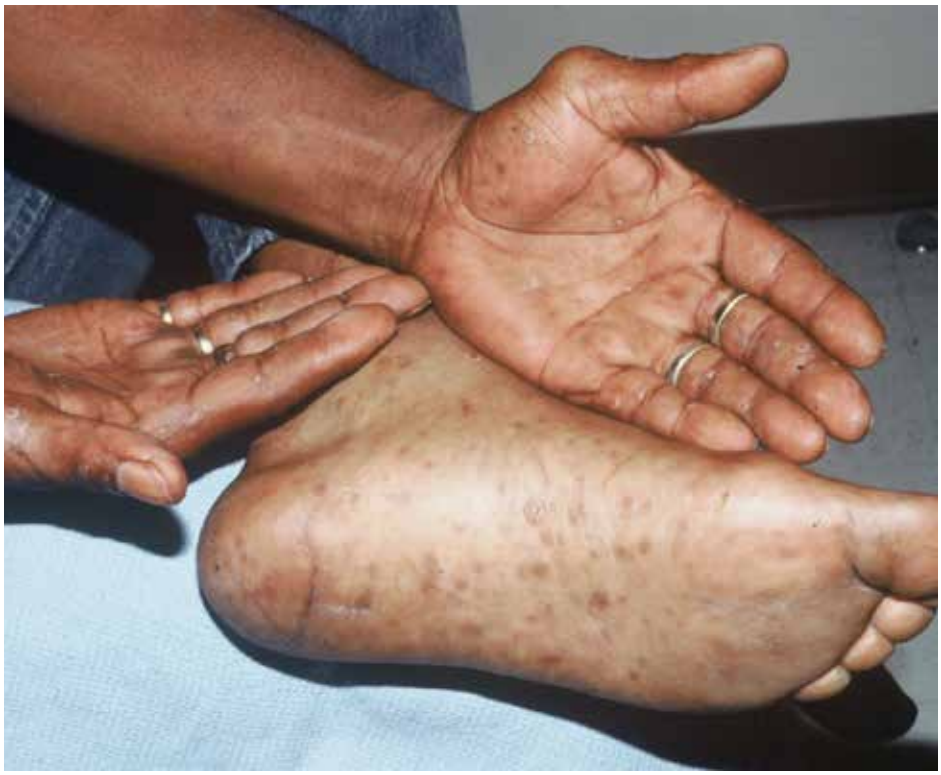
Derm Dilemma

Blake St. Clair, M3; Kevin St. Clair, MD

A 45-year-old man presents with a six-week history of mild malaise, arthralgias and myalgias, and a generalized skin eruption. The rash is characterized by 5-10mm red-brown macules involving the trunk symmetrically, but also noted on the palms and soles. He has also experienced patchy hair loss on the scalp. The only current medication is lisinopril, prescribed for hypertension 10 months ago. His mother suffers from lupus erythematosus.

Physical examination also reveals generalized adenopathy and moist white papules and plaques on the penile shaft and scrotum.



Initial steps in evaluation should include which of the following?

A. Immediate discontinuation of lisinopril and evaluation of hepatic transaminases and peripheral eosinophil count as this patient most likely is developing DRESS syndrome (drug rash with eosinophilia and systemic symptoms)

B. Autoimmune serologies, especially ANA (antinuclear antibody) as this man likely has acute lupus erythematous

C. Viral culture of the mucosal lesions of the groin, Herpes Zoster IgM titer, assessment for underlying immunosuppression, and empiric intravenous acyclovir as the patient most likely suffers from Varicella

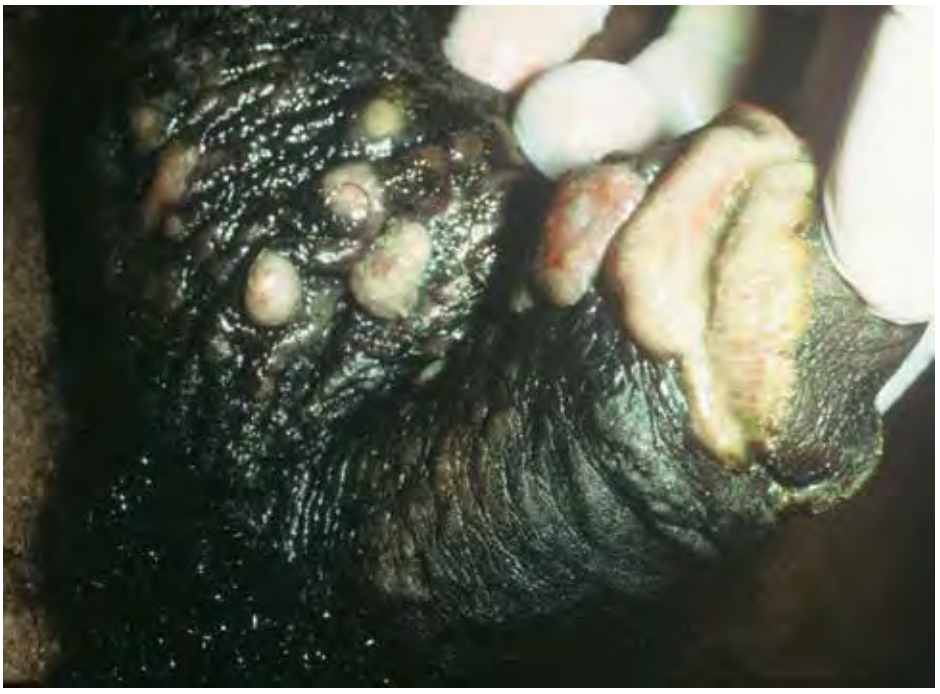
D. Nontreponemal serological screening with the Venereal Disease Research Laboratory (VDRL) or rapid plasma regain (RPR) assays as the patient most likely has syphilis

E. Reassurance that all symptoms will resolve spontaneously, as this patient probably has pityriasis rosea

Answer: D

Although the potential manifestations of the diseases listed as choices are protean, the signs and symptoms exhibited by the described patient are most consistent with secondary syphilis. If the result of the screening nontreponemal serology is positive, then confirmatory treponemal tests such as fluorescent treponemal antibody absorption (FTA-ABS), Treponema *pallidum* particle agglutination (TPPA), or T. *pallidum* hemagglutination (TPHA) should be performed.

Clinical presentations of syphilis are extremely variable, and the disease has been appropriately nicknamed the "Great Imposter." Sir William Osler has aptly stated that "the physician who knows syphilis knows

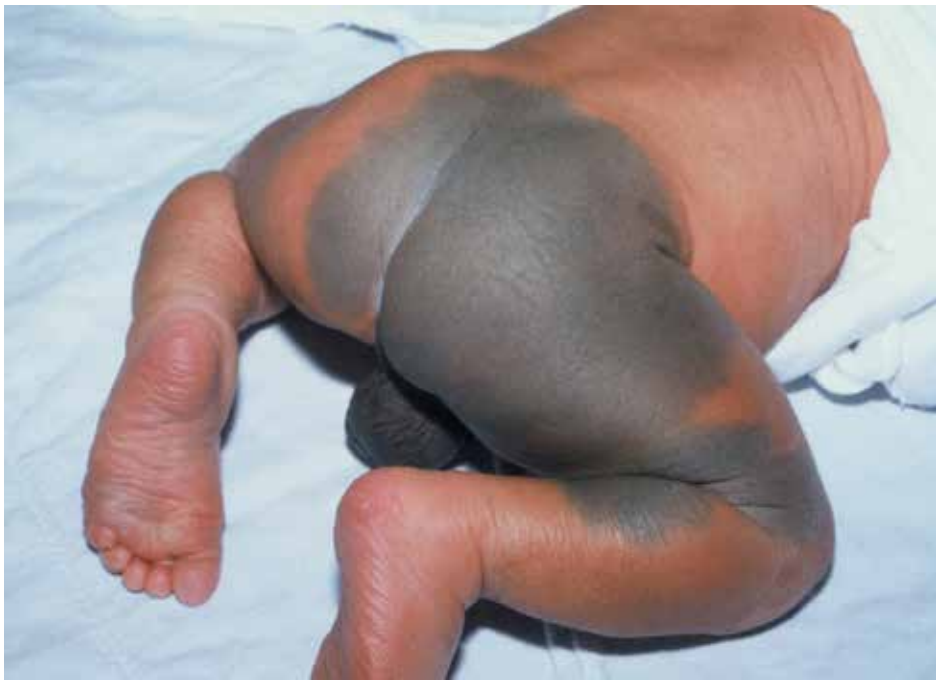


medicine." A complete description of this affliction is beyond the scope of this feature; the reader is referred to the many excellent resources for review of this topic, which is important to almost any discipline within the house of medicine. AMS

Derm Dilemma

Rachel White, M3; Kevin St. Clair, MD

A 5-week-old baby of American Indian descent presented at birth with an extensive hyperpigmented patch involving the sacrum, buttocks, scrotum, and right lower extremity. No other overt clinical abnormalities were noted by the examining pediatrician. A skin biopsy demonstrated banal appearing melanocytes involving the deep dermal appendages and neurovascular structures, as well as extension deep into the subcutaneous adipose. Subsequent appropriate step(s) in the evaluation or management of this child should include:



A. Magnetic resonance imaging of the central nervous system

B. Reassurance to the parents that no further evaluation is necessary, as this represents only a "Mongolian spot"

C. Urgent referral for Erbium-YAG laser therapy

D. Urgent referral to pediatric surgeon for staged excision

E. Genetic counseling for the parents

Answer: A

MRI of the CNS to exclude the presence of neurocutaneous melanosis

The described histologic findings indicate that this lesion is a congenital melanocytic nevus (CMN), which is present in an estimated 1-2% of newborns. These lesions are classified into three groups: small (< 1.5 cm greatest diameter), medium (1.5-19.9 cm), and giant (> 20 cm). Giant CMN are associated with an approximately 5-7% risk of transformation to melanoma by age 60, whereas risk of smaller lesions is not quantified but considered to be much less. Posterior midline location, as in this patient, or association with multiple satellite nevi are indications of potential underlying neurocutaneous melanosis, which portends a poor prognosis. Staged excision and laser therapy are eventual treatment options for CMN, but MRI imaging to rule out neurocutaneous melanosis should be obtained by age four months.

1. Congenital dermal melanocytosis (Mongolian spot) generally presents as localized blue-grey pigmentation of the sacrum, particularly in American Indians, Asians, and Hispanics. Spontaneous resolution usually occurs in the first few years of life. In uncommon situations where distinction from CMN is clinically difficult, biopsy allows differentiation. AMS

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OBITUARIES

LITTLE ROCK - **Edgardo "Eddie" Angtuaco, MD**, passed away on May 28, 2018. He is survived by his wife, Dr. Teresita "Terry" Angtuaco; his children, Dr. Michael Angtuaco (Melissa) and Christine Clarkson (Robert); and his granddaughters, Gabriella, Bella Maria, and Katarina Angtuaco, in addition to Christine's baby due later this year. Dr. Angtuaco proudly served on the faculty at the University of Arkansas for Medical Sciences for 39 years as a neuroradiologist and had been the division chief of neuroradiology for many years. He was also honored to have played a role in the education of hundreds of resident and fellow physicians at UAMS and will be remembered through this professional legacy.

LITTLE ROCK - **Charles Allen McKnight, MD**, passed away June 6, 2018. He is survived by children Carla Buchanan (Allen) of Orange, Cal., Charlotte Cook (Clay) of Little Rock, Cherie McKnight of Bryant, Connie McKnight of Little Rock, Charles McKnight (Robert) of Santa Monica, Cal., and Carson McKnight of Little Rock; grandchildren, Blake Buchanan, Michael Buchanan, Paige Harper, Scott Cook, Lance Cook, Allen Cook and Grant McKnight; and four great-grandchildren. Dr. McKnight was a graduate of the University of Arkansas, where he was a Kappa Sigma and a Razorback cheerleader. He was an OB-GYN for fifty years, delivered thousands of babies, and established Central Clinic for Women in Little Rock. In the late 1960s, he invented the amnihook, a medical instrument which is still in worldwide use today. He assisted in the founding of Pulaski Academy, of which all six of his children were graduates. He was a member of the Arkansas Medical Society Fifty Year Club.

LITTLE ROCK - **Eldon Gerald Schulz, MD**, passed away June 2, 2018. He is survived by wife Margaret Ann Brink Schulz, daughter Kestin Margaret Schulz and son-in-law John Christie, and daughter Kari Loretta Schulz. Dr. Schulz received his Bachelor of Science from the University of Idaho, Moscow, his medical degree from the University of South Dakota, his Residency of Pediatrics from the University of California, San Diego, and his Developmental/Ambulatory Pediatric Fellowship at the University of Massachusetts Worcester. He spent six years with Margaret in Heidelberg, Germany, working with the Exceptional Family Member Service as a civil servant in the military. He and his wife moved to Little Rock in 1991. Here, he became the professor of developmental pediatrics at UAMS. During his time at UAMS, he supported numerous programs including the James L. Dennis Developmental Center, CoBALT, LEND,Easter Seals, the State's Human Development Centers, and DHS. In 2008, he received the Rockefeller Endowed Chair for Children with Special Needs. AMS

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