

Sickle Cell Today

USA Comprehensive Sickle Cell Center

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Myths and Facts about Birth Control in Sickle Cell Disease

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The concept of developing methods to prevent pregnancy has been around for thousands of years. Ancient Egyptian scribes have described men wearing condoms made out of animal membranes. In the fourth century B.C. Aristotle, a Greek philosopher, recommended the use of olive oil as spermicides. It was only in 1960 when the first hormonal formulation for birth-control pills (called Enovid) was approved by the U.S. Food and Drug Administration. Over the past decade various hormonal preparations and combinations have been developed.

Women with sickle cell disease (SCD) have an increased risk of pregnancy complications, such as premature rupture

of membranes, preterm labor, and post partum infections. Women with SCD who want to avoid pregnancy need to have appropriate and reliable counseling regarding methods of birth control. Unfortunately, there are no universal guidelines established. In this article, we will try to talk about myths and facts in the minds of both doctors and patients regarding birth control options in SCD.

Myth: The “pill” is just the pill with different names

Combination Birth Control Pills (eg: Microgynon):

The combination pills (CP) contain two sex hormones called progestogens and estrogen. The CPs come in 21-day and 28-day packs. They are easy and effective if taken properly and most women are able to conceive soon after they stop taking the pills. They also help prevent cancers of the ovaries and uterus; improve acne and make periods regular, lighter and less painful. None of the pills protect against infections. The drawbacks of CPs include increased risk of heart attacks, strokes, and blood clots that start in leg veins but travel to lungs and other parts of the body. The more recent (new generation) CPs contain a much smaller dose of estrogen and different forms of progestogen that reduce the risk of clot formation. The risk of heart attacks is 20 times greater for women who take CPs and smoke cigarettes. Other important risk factors include SCD,

Continued on next page

obesity, high blood pressure or having a central venous access device (also called port).

The World Health Organization (WHO) currently classifies sickle cell anemia as a Category 2 for CPs, meaning that the benefits of using CPs generally outweigh the risks. However, some of the complications associated with SCD such as structural heart disease, history of strokes and high blood pressure are listed as Category 4 (refrain from use) and gall bladder disease as Category 3 (exercise caution). In a large study conducted in the United Kingdom, women with SCD were asked about complications while using different birth control methods. About 6% of women using CPs reported an increase in pain crisis. The bottom line is that careful screening and counseling is warranted before prescribing the CPs to patients with SCD.

Myth: Combined hormonal birth control pills are contraindicated in all women with SCD

Progestogen-Only Birth Control Methods:

This is a safer group of hormonal birth control methods. The WHO classifies SCD as Category 1 for all progestin only birth control methods, meaning no restrictions to use. They come in different varieties and are listed as follows:

1. Progestin only Pills (POP or mini-pills):

Unlike CPs, the POPs do not contain estrogen and the progestin dose is lower than the progestin dose in a CP. The POPs are taken daily at the same time every day, even during the period. They are considered safe in SCD patients and the WHO has

listed history of stroke as Category 3 meaning exercise caution.

2. Implantables (e.g.: Norplant)

These are capsules containing a form of progestogen, inserted under the skin in the upper arm by creating a small incision. They start working within 24 hours of insertion and last up to 5 years. A few clinical research trials outside of the United States have been performed on a small number of women with SCD. No serious or unexpected side effects have been reported. One of the studies from Brazil showed that the use of Uniplant caused a rise in fetal hemoglobin levels leading to decreased sickling and pain crisis.

3. Injectables (Depot)

The depot medroxyprogesterone acetate (DMPA) or Depot shots are given 4 times a year in the thighs, buttocks or upper arms. A clinical trial performed at the WHO Contraceptive Clinic in Panama compared the effects of DMPA to CPs. No serious side effects were reported in either of the groups. Interestingly 9 out of 13 patients receiving DMPA were pain free by 12 months. Another well conducted study has shown that the use of DMPA resulted in an increase in fetal hemoglobin levels, significantly decreasing frequency of pain crisis and improvement of anemia and jaundice.

Fact: Women with SCD may note a decrease in sickling or painful crises with Depo-Provera use

4. Progestin-Releasing Intrauterine System (e.g: Mirena)

This is another option for women in whom CPs are not safe and want a long-term birth control. It can be used for 5 years and may reduce painful menstrual bleeds in women with heavy periods.

Non Hormonal / Barrier Birth Control Methods:

These methods provide barriers between the sperms and egg. Common examples are male and female condoms, diaphragms, contraceptive sponge and vaginal spermicides. The copper-releasing Intrauterine Devices (IUD) can be used for 10 years before replacement. Some women experience painful periods. The IUDs can fall off requiring reinsertion by a doctor. The WHO classifies SCD Category 1 for use of all barrier methods, meaning there is no restriction for their use.

Fact: Male condoms provide best protection against sexually transmitted disease

Clinical Recommendations

The WHO has recommended that sickle cell anemia be classified as Category 1 for progestin only birth control methods, meaning that there are no restrictions for use.

The CPs are not contraindicated for use in SCD but must not be taken without consulting a doctor. We recommend all prescribers to strongly encourage all women to stop smoking and adapt healthy living. They should obtain information about family history for blood clots, past history of strokes, heart disease or presence of a central line, in order to optimize individual counseling and the achievement of an acceptable benefit to risk ratio. Women must know about symptoms of venous blood clots and remember to inform the doctor at any encounter about current use of birth control pills.

SICKLE TRAIT IN ATHLETES and SUDDEN DEATH: An Update

Johnson Haynes, Jr., MD

Professor of Medicine • Director, USA Comprehensive Sickle Center

In April of 2010, the National Collegiate Athletic Association (NCAA) Division I Legislative Council passed a ruling that all division I student-athletes must be tested for sickle cell trait (SCT), show proof of a prior test or sign a waiver releasing an institution from liability if they decline to be tested. The basis for this ruling stems from a law suit settlement between Rice University and the family of a football player with SCT that died while training in 2006. This ruling took effect for the 2010-2011 academic year. As of yet, the NCAA has not expanded this ruling to division II or III athletes, nor has the NCAA shared its data with the medical community. The NCAA maintains that athletes with SCT can participate in college sports but that certain training precautions should be implemented as suggested by the National Athletic Trainer's Association (NATA; <http://www.nata.org/sites/default/files/SickleCellTraitAndTheAthlete.pdf>). NATA and the NCAA promote education to create an environment that encourages trainers and athletes with SCT to immediately recognize and report any symptoms potentially caused by exertional sickling. Such symptoms include leg or low back pain, cramping, fatigue, and shortness of breath. Trainers and athletes are also educated on more effective interventions aimed at avoiding situations which may predispose to exertional sickling such as heat stress, dehydration, asthma, and intercurrent illness. Extended recovery periods between repetitive sprints are encouraged and supplemental oxygen should be available when training or competing at high altitude.

Following the NCAA ruling, the National Sickle Cell Disease Association (SCDAA) of America issued its Executive Summary on "Sickle Cell Trait and Athletics" (<http://www.sicklecelldisease.org/index.cfm?page=news&id=29>). This document reports that sudden death during military training in individuals with SCT "can be reduced in all recruits by avoiding dehydration and overheating during training". Of note, no similar studies have been done in athletes. Thus, the SCDAA raises concern as to the true vulnerability of athletes with SCT to exertional sickling/rhabdomyolysis during strenuous exercise and questions whether SCT is causal or just an association with sudden death. The SCDAA's greater concern is that, "Screening athletes for SCT and subjecting them to alternative training regimens, as recommended by the NCAA and NATA, has not been demonstrated to reduce the incidence of training-related deaths." Concerns of stigmatization and discrimination against athletes with SCT without adequate assurance of the privacy of genetic information have not been effectively addressed by the NCAA. Furthermore, the SCDAA supports the implementation of universal, safe training guidelines for all athletes.

In a July 25, 2012 press release, Barry J. Maron, MD director of the Hypertrophic Cardiomyopathy Center at the Minneapolis Heart Institute Foundation report findings for U.S. Sudden Death in Athletes Registry showing there is "convincing evidence

of a causal relationship between the SCT and deaths of young black competitive athletes, especially football players." This registry contains 2,462 athlete deaths of which 23 occurred in association with SCT: 21 were male and all were African American. Most deaths occurred in college age athletes (19- 23 years) during football conditioning drills, early in the season, while exposed to high environmental temperatures. Maron goes on to say, "To not acknowledge this link between SCT and sudden death creates the possibility of a failure to fully protect the athlete community." This study will be published in the October issue of the American Journal of Cardiology.

As more data is provided, more questions must be answered, i.e., why is the NCAA stance on SCT screening limited to Division I athletes? Is a retrospective registry data base analysis adequate enough to assign SCT as a cause of sudden death in athletes? Will the NCAA and SCDAA join forces to actually provide the resources necessary to prospectively determine event triggers and mechanisms of SCT as being causal in sudden death or not? Does hydration, avoidance of overheating during training and longer periods of rest between sprints protect the athlete with SCT from risk of sudden death? Until we have more information, implementation of the NATA recommendation for all athletes, at all levels, seems reasonable.

Make Plans to Attend the 2012 USA ANNUAL SICKLE CELL CONFERENCE SEPTEMBER 7-8, 2012



The 2012 Sickle Cell Conference, Practical Issues XII: Casting the Net II, will be held on Friday, September 7, 2012 from 8:30am until 3:30pm and Saturday, September 8, 2012 from 8:00am until 1:00pm at the University of South Alabama Mitchell Center Globe. The Keynote speaker will be Dr. Robert “Bob” Adams from the Medical University South Carolina. Dr. Adams will be featured on Friday, September 7, 2012 and the topic will be “Primary Stroke Prevention: The Falling Rates of Childhood Strokes in Sickle Cell Disease”.

For the first time conference participants will have the choice of attending the one day session or two day session for continuing education units or continuing medical education credits.

This year’s conference will feature Dr. Felicia Wilson, Professor in the Department of Pediatric Hematology/ Oncology University of South Alabama as the luncheon speaker on Friday, September 7, 2012. Dr. Wilson will speak on the topic “Newborn Screening in Sickle Cell Disease”.

Other speakers for Friday, September 7, 2012 are University of South Alabama faculty, Dr. Thomas Butler, (Platelet Disorders in Sickle Cell Disease); Dr. Susan Baker, (Sickle Cell Disease in Pregnancy), and Elise Labbe-Coldsmith, PhD (Psychosocial Issues in Sickle Cell Disease). All lectures will be 45 minute presentations with 15 minutes allotted for questions.

Speakers for Saturday, September 8, 2012 are Alan Sanders, PhD from Newtown Square, Pennsylvania (Ethics in Pain Management, Fears, Realities, and Needs); University of South Alabama faculty: Dr. Juvonda Hodge (Cholecystectomy Indications and Special Considerations

in Sickle Cell Disease), Dr. Johnson Haynes, Jr. (Sickle Cell Disease and Air Travel), and Dr. Anthony Martino (Moya/Moya / Encephaloduroarteriosynangiosis Intervention).

A light continental breakfast and lunch will be provided on Friday, September 7, 2012. A light continental breakfast will be provided on Saturday, September 8, 2012.

Reservations can be made by calling the USA Sickle Cell Center Administrative Office at (251) 470-5893 or online at www.usa-cme.com.

The program has been approved for 7.5 CEUs on day 1, 5.7 CEUs on day 2 and 13.2 for participants who attend both days. The University of South Alabama Medical Center Staff Development is accredited by the Alabama Board of Nursing as a provider of continuing education in nursing. Nurses licensed in the state of Alabama are required to bring their current Alabama license to the conference to scan for CEU credits each day. This is a mandate of the Alabama Board of Nursing. A \$5.00 fee each day will be applied to nurses without their current Alabama license.

The University of South Alabama College of Medicine is accredited by the Accreditation Council for Continuing Medical Education to provide continuing medical education for physicians.

The University of South Alabama College of Medicine designates this live activity for a maximum of 10 *AMA PRA Category 1 Credit(s)*TM.

Physicians should claim only credit commensurate with the extent of their participation in the activity.

“Funding for this conference was made possible (in part) by P20MD002314 from the National Institute on Minority Health and Health Disparities. The views expressed in written conference materials or publications and by speakers and moderators do not necessarily reflect the official policies of the Department of Health and Human Services; nor does mention by trade names, commercial practices, or organizations imply endorsement by the U.S. Government.”

PEDIATRIC TO ADULT CARE PROGRAM (PACT) PROMOTES HIGHER EDUCATION

Brittany Brown, RN, BSN • PACT Coordinator

It's that time of the year again! Returning for fall semester college classes or venturing into college life for the first time. For most students, this is a time of excitement and joy, meeting with old friends or embarking on a whole new adventure of leaving home for school. However, for those with SCD, this may be a time of anxiety and worry. One of PACT's main goals is to help prepare young clients for such changes through education.

Whether you are attending college locally or away from home, caring for yourself is a top priority. Listed below are a few tips for college life and maintaining good health with sickle cell disease (SCD).

- **Know your history-** No one knows your medical history better than yourself. Have your medical information documented in case your memory fails you. You should document the type of SCD you have (HbSS, sickle-C, or sickle- thalassemia) your transfusion history, current medications, and pertinent contact information.
- **Keep in touch with your home-based physician-** Doctors and nurses who have been caring for you over the years are the best people to manage your overall health. Arrange in advance to get regular check-ups during school breaks. Let your home care team know whenever you get medical care at school and have your school-based physician send a copy of any lab results or records back to your home-based physician.
- **Have your home-based physician copy your records and prepare a travel letter for you-** This information will help any medical professional determine appropriate care for you. Current records will allow physicians to decipher what medical treatment works best to manage your care.
- **Find a local physician in the college student health department or community-** Do this as soon as you arrive at college so you have a plan in case of an emergency. Inform the staff on your health situation and give emergency contact information as well as home physician information.
- **Find out if there is a day treatment center for SCD near where you will be living-** Some large cities have specialized sickle cell treatment centers. Because of their expertise, they may be able to help control any pain crises faster than a regular emergency room.

- **Ask for special housing if you need it-** If the climate where you are going gets very hot or cold, you'll need to have appropriate accommodations. These things are required under the Americans with Disabilities Act, so contact your schools disability services.

- **Sign up for a 504 plan-** Usually this process is handled during registration. The 504 plan is a law that states all federally funded schools must make reasonable accommodations for students with disabilities. In this case, SCD, a chronic medical condition falls under this disability category. The 504 plan will allow excused absences, make up work/tests, and private testing if needed.

- **Study hard-Try to stay ahead in your assignments in case unexpected pain crises occurs.**

Finally, make friends, call home to speak with mom and dad, and have fun! College life is a wonderful experience. Build memories that will last a lifetime. Enjoy the process of learning new things.

Listed are a few college scholarships dealing with SCD specifically that can be found on the internet at <http://www.collegescholarships.org/health/sickle-cell.htm> Most scholarships are based on grade point average, SAT scores, community involvement, and how SCD has impacted your life.

- Kermit B. Nash Academic Scholarship
- The International Association of Sickle Cell Nurse and Physicians Assistants, Inc.
- Ohio Sickle Cell and Health Association
- The Jacqueline M. Kidd Foundation



photo L to R: Dr. Hafeez Siddiqui, Dr. Hamayun Imran, Dr. Johnson Haynes Jr., Brittany Brown RN, Ardie Pack-Mabien CRNP

Never let a defined “disability” stop you from being the best you can be.



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U.S. Centers for Disease Control and Prevention recommends a yearly flu vaccine for everyone as the first and most important step in protecting against the flu

Ardie Pack-Mabien, CRNP



The Food and Drug Administration, World Health Organization, U.S. Centers for Disease Control and Prevention, and other institutions identify the influenza viruses most likely to cause illness during the upcoming flu season. While there are many different flu viruses, the flu vaccine is designed to protect against the three main flu virus strains. The 2012-2013 flu vaccine will protect against three different flu viruses: H3N2 virus, influenza B virus (from the B/Yamagata lineage of viruses), and the H1N1 virus. The vaccine is different from the 2011-2012 influenza vaccine for the Northern Hemisphere. The vaccine is chosen to maximize protection against the viruses most

likely to spread and cause illness among people during the upcoming flu season.

What actions can I take to protect myself and my family against the flu this season?

Getting the flu vaccine as soon as possible after it becomes available each year is always a good idea, and the protection you get from vaccination will last throughout the flu season. For information about

vaccine supply this season, please visit <http://www.cdc.gov/flu/about/qa/vaxsupply.htm>.

Contact your primary care provider or local health department for the availability of the flu vaccine and an appointment.

In addition to obtaining your flu vaccine annually, you can take everyday preventive steps like staying away from sick people and washing your hands to reduce the spread of germs. If you are sick with the flu, stay home from work or school to prevent spreading influenza to others.

Who should receive the flu vaccine?

Adults and children who have chronic disorders requiring medical follow-up or hospitalization due to kidney disease, hemoglobinopathies (sickle cell disease), or conditions that compromise lung function should receive the flu vaccine annually. The flu vaccine promotes immunity to the influenza virus by stimulating specific antibody production.

When should I receive the flu vaccine?

The optimal time to receive the flu vaccine is October – November and prior to exposure to the influenza virus. The flu vaccine can be given through the month of December and later as long as the vaccine is available. The flu season may last as long as May. There have been shortages of the flu vaccine in the past and it is strongly recommended you do not delay obtaining your flu vaccine.

Visit the Comprehensive Sickle Cell Center website at:
<http://www.usahealthsystem.com/sicklecellcenter>