8QLYHUVLW\ RI 6RXWK $ODEDPD

Sickle Cell Today

FROM THE DIRECTORS DESK:

ONE BAD APPLE DOESN'T SPOIL THE WHOLE BARREL

It had been an exceptionally warm fall for the month of November in Alabama. Everyone was still running around in VKRUWV DQG VKRUW VOHHYHV , ZDV MXVW OLNH DQ\ RWKHU GRFWRU LQ WKH HPHUJHQF\ URRP what an unusually quiet month it had been except for the routine chest pains, chronic "lungers," diabetics, and the "frequent À\HUV´ ZLWK VLFNOH FHOO GLVHDVH WKDW VHHPHG WR OLYH LQ WKH HPHUJHQF\ URRP PRVW GD\V RI WKH PRQWK ,W ZDV D VWDQ amongst the doctors how two patients with sickle cell disease, Margaret and Bobby, were in the emergency room for their daily shots of demerol and how they were just addicts. Every time I heard this it would make me so angry. I would often wish they could just walk a mile in Margaret's and Bobby's shoes. Mary and I were the only ones who were willing to treat them appropriately and respectfully when they came to the emergency room. Some of my colleagues would even try giving them sterile water for pain. All I knew was as doctors, we had no means to objectively measure pain and it always boiled down to believing the patient. It often seemed so unfair. Maybe they were not having a pain crisis and maybe they were just asking for help. All I knew was if it were me hurting, I would be very angry if I had to beg for the treatment I deserved if I had to convince someone I was in pain.

MARK YOUR CALENDAR

THE 2014 USA ANNUAL SICKLE CELL CONFERENCE:

Practical Issues XIII-The Many Faces of Pain

The USA Comprehensive Sickle Cell Center is in the final stages of planning its 13th dynamic and informative Annual Regional Sickle Cell Conference. The focus this year will be on "Pain in Sickle Cell Disease and Pain Management."The conference is scheduled for Saturday, May 3, 2014 from 8:00am – 3:45pm in the School of Allied Health auditorium on the main campus of the University of South Alabama. National and local experts will be presenting the most current and practical issues experienced by healthcare providers caring for those affected by sickle cell disease and pain management. The target audience is physicians, physician-assistants, nurse practitioners, nurses, and allied health professionals. This conference is supported by the Dr. Cecil L. Parker Jr. Sickle Cell Disease Lectureship Endowment. The purpose of the Endowment is to provide support for the Annual Regional Sickle Cell Conference and educational needs of the clients and health care providers of the Gulf Coast community.

Register early to enter for a chance to win complimentary admission to the 2015 Annual Regional Sickle Cell Conference. Early bird registration deadline is April 11, 2014. For additional conference information visit http://www.usahealthsystem.com/newspecial-events or call (251) 470-5893.

References continued from page 3

Charache, S., Dover, G.J., Moore, R.D., Eckert, S., Ballas, S.K., Koshy, M.,…Platt, O.S. (1992). Hydroxyurea: effects on hemoglobin F production in patients with sickle cell anemia. Blood, 79(10), 2555-2562.

Charache, S., Terrin, M.L., Moore, R.D., Dover, G.J., Barton, F.B., Eckert, S.V., …Bonds, D.R. (1995). Effect of hydroxyurea on the frequency of painful crises in sickle cell anemia. New England Journal of Medicine, 332, 1317-1322.

Hankins, J.S., Ware, R.E., Rogers, Z.R., Wynn. L.W. Lane, P.A., Scott, J.P., & Wang, W.C. (2005). Long-term hydroxyurea therapy for infants with sickle cell anemia: the HUSOFT extension study. Blood, 106(7), 2269-2275. doi: 10.1182/blood-2004-12-4973.

http://www.report.nih.gov/nihfactsheet. Sickle Cell Disease: Yesterday, Today, & Tomorrow. Released March 29, 2013.

McGann, P.T., & Ware, R.E. (2011). Hydroxyurea for sickle cell anemia: what have we learned and what questions still remain? Curr Opin Hematol., 18(3), 158-165. doi: 10.1097/MOH.0b013e32834521dd.

Nottage, K.A., Hankins, J.S., Smeltzer, M., Mzayek, F., Wang, W.C., Aygun B., & Gurney, J.G. (2013). Hydroxyurea use and hospitalization trends in a comprehensive pediatric sickle cell program. Plos One, 8(8), e72077. doi:10.1371/jounal.pone.0072077.

Platt, O.S., Orkin, S.H., Dover, G., Beardsley, G.P., Miller, B., Nathan, D.G. (1984). Hydroxyurea enhances fetal hemoglobin production in sickle cell anemia. J Clin Invest, 74, 652-656.

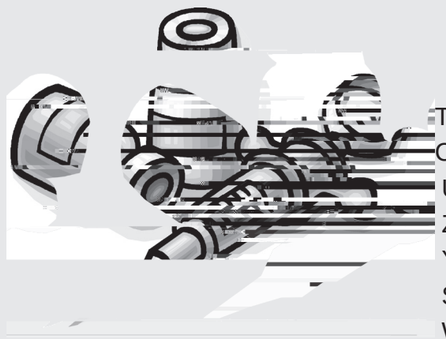
Quinn, C.T., Rogers, Z.R., McCavit, T.L., & Buchanan, G.R. (2010). Improved survival of children and adolescents with sickle cell disease. Blood, 115(17), 3447-3452.

Sickle Cell Disease and Hydroxyurea: Yesterday, Today, and Tomorrow

Submitted by
Ardie Pack-Mabien, CRNP

Mandatory newborn screening in the United States has resulted in early identification and medical access by those newborns affected with sickle cell disease (SCD). The addition of penicillin prophylaxis and pneumococcal immunizations as preventive measures, technological advancements in the development of diagnostics and screening tools and innovative medical management of the pediatric population have resulted in decreased morbidity and mortality for individuals living with SCD. The improvement in survivorship from childhood to adulthood has been estimated at 94% for individuals with HbSS and HbS $`^{0}$ Thalassemia and 98% for individuals with HbSC and HbS $`^{+}$ Thalassemia (Quinn, et. al, 2010).

What role has hydroxyurea played in the medical management of individuals with SCD? The utilization of hydroxyurea as a potentially useful drug therapy in the management of individuals with SCD was first reported in 1984. Additional multi-center studies sponsored by the National Institutes of Health were conducted and found that hydroxyurea reduced the frequency and severity of sickle cell related pain crisis, number of acute chest syndrome episodes, frequency and number



PLAYING IT SAFE

The Center for Disease Control and the Advisory Committee on Immunization Practices are now UHFRPPHQGLQJ WKDW LQGLYLGXDOV \HDUV RU ROGHU ZLWK VLFNOH FHOO GLVHDVH UHFHLYH WKH 3UHYQDU YDFFLQH 3UHYQDU ZDV LPSOHPHQWHG LQ WKH SHGLDWULF SRSXODWLRQ LQ :KLOH 3QHXPRYD[KDV EHHQ IRXQG WR EH HIIHFWLYH DJDLQVW SRWHQWLDOO\ OLIH WKUHDWHQLQJ LQIHFWLRQV GXH WR WKH SQHXPRFRFFDO EDFWHULD 3UHYQDU YDFFLQH KDV WKH DELOLW\ WR SURYLGH additional protection. Please see the sickle cell nurse or contact your healthcare provider for information on the vaccine.

Submitted by: Brittany Brown, BSN, RN

“Decline” continued from page 1

Between January 2006 and June 2012, a study led by Dr. Abdul Hafeez Siddiqui was conducted at the University of South Alabama Children’s and Women’s Hospital which reviewed 456 hospitalizations in 133