Sickle Cell News Update for September 2007 from
aplatt@EMORY.EDU and www.scinfo.org/news.htm

We Need To Make Preventing and Curing Sickle Cell Disease A National Priority

By Senators Thomas R. Carper (D-DE) and Benjamin L. Cardin (D-MD)

In recent years, a lot of attention has focused on a few debilitating diseases that affect millions of Americans. Coordinated public and private efforts -- including targeted biomedical research, awareness campaigns, and patient advocacy -- have enabled us to make remarkable progress toward conquering many of these diseases.

But other, less prevalent diseases that are just as serious have received scant attention. Sickle cell disease – a devastating genetic disorder that affects red blood cells -- is such a disease, and **greater awareness and more resources are required if we are to defeat it.**

September is National Sickle Cell Awareness Month, and many Americans have never heard of the disease or know little about it. They are unaware that this disease – which creates oxygen-depleted red blood cells that become "sickle"-shaped and block small blood vessels – causes pain so severe that a flare up is often referred to as a "crisis." These sickle cell "crises" can lead to stroke, organ failure, and death.

Experts estimate that between 70,000 and 80,000 Americans have sickle cell disease, and more than 1,300 babies are born with the condition each year. Although sickle cell disease is rare, it is by far the most common genetic blood disorder in this country.

Unfortunately, the treatment of sickle cell disease has been hampered by too few resources and lack of information.

Health care providers must be familiar with the symptoms of sickle cell and be able to properly diagnose and treat the disease – especially in hospital emergency departments where patients frequently seek treatment during a pain crisis. However, too many health care professionals have not received in-depth training about sickle cell disease.

In Delaware, for example, the Alfred I. duPont Hospital for Children is the only hospital in the state that serves the pediatric sickle cell population. In Maryland, Johns Hopkins houses the state's only clinic dedicated specifically to the medical care of adults with sickle cell. There needs to be greater availability of comprehensive health care services for both children and adults suffering from sickle cell disease.

The lack of public information about the disease is also a problem. Although virtually every state in America has screened newborns for sickle cell since 1986, **most adults do not know their sickle cell trait status.** Individuals with the sickle cell trait do not have the disease, but they carry a gene that increases their children's chances of having it. In cases where both parents have the sickle cell trait, there is a 1 in 4 chance – with each pregnancy – that their child will have sickle cell disease.

Many sickle cell patients also require blood transfusion, and most patients do best if they can receive blood transfusions from genetically similar donors. Yet too few individuals of African, Latino, Mediterranean, and South Asian heritage – groups with the highest incidence of sickle cell disease – are aware that they can help sickle cell patients by donating their blood.

It is difficult to outline a complete list of challenges with respect to sickle cell disease without addressing the issue of money. The reality is that treatment, prevention and advocacy efforts all require funding. Both public and private funding is needed to provide the resources for fighting this disease.

According to a 2006 Pediatrics article, only $1,000 per patient per year is spent on sickle cell disease research. This pales in comparison to the $9,000 per patient that has been raised for other diseases, like cystic fibrosis. The largest disparity was not in federal funding; rather, it was in private funding – an indication the public is still largely unaware of the disease.

Research supported by the National Institutes of Health (NIH) has led to the development of more effective newborn screening tools, the identification of blood transfusion as a way to reduce the risk of strokes, and an FDA-approved treatment that aids in the prevention of painful sickle cell episodes.

Scientists believe that we are on the cusp of developing more accurate screening tools for sickle cell disease, personalizing therapies according to individuals' profiles, and discovering actual preventions.

We must build on the steps taken in previous Congresses and ensure that the NIH has the resources to build on promising sickle cell research projects.

During National Sickle Cell Awareness Month, we should begin to bridge the gap between need and results. It is time to promote increased attention to sickle cell disease in our nation's medical schools and academic medical centers so that health professionals are trained appropriately to diagnose and treat sickle cell patients. We also need to put our money where it will get the best results – into more research and education about this devastating disease.