National Sickle Cell Awareness Day Bill

Second Reading—Debate Adjourned

Hon. Jane Cordy moved second reading of Bill S-211, An Act respecting National Sickle Cell Awareness Day.

She said: Honourable senators, I'm pleased today to speak to Bill S-211, An Act respecting National Sickle Cell Awareness Day. I introduced a similar bill in the previous Parliament, and as I did at that time, I must begin by thanking Ms. Lanre Tunji- Ajayi, President of the Sickle Cell Disease Association of Canada. This bill would not be before you today without her passion and dedication to improving the lives of Canadians living with sickle cell disease. I'm honoured to work with her to introduce Bill S- 211, An Act respecting National Sickle Cell Awareness Day.

Honourable senators, when I spoke to the previous incarnation of this bill, I mentioned taking part in the Sickle Cell Disease Association of Canada's advocacy day on Parliament Hill. I was honoured to meet with Dr. Robert Klaassen from CHEO, Princess Sanusi, whose 16-year old son has sickle cell disease, and Kemoh Mansaray, a nurse from Edmonton who has friends and family who carry the sickle cell gene. Along with other senators and MPs from all parties, I had the pleasure of attending the Sickle Cell Disease Association's reception on Parliament Hill. During the reception, several people told their personal stories about dealing with the disease. It was a moving experience to hear about the challenges of living with sickle cell. It was also touching to see the optimism in the room that life could improve for those with sickle cell because so many people were working together. Honourable senators, we all know that when people work together, good things can happen.

One mother at the reception told about her experience with her young son, who was frequently crying because of extreme pain in his arm. She and her husband thought he must have injured it in a fall, but they couldn't find any signs of bruising or swelling. They brought him to the hospital for examination by the doctor. An X- ray came up negative, and the doctor told the parents to take their son home and give him Tylenol. After three days with their child continuing to cry in pain, they returned to the hospital, and this time they were blessed. The doctor on duty that day had just learned about sickle cell disease. Because of this new knowledge, the doctor ordered a blood test. The boy's test came back positive for sickle cell. The parents were both carriers of the sickle cell gene, but had not even heard of the disease before that day. Now that the child's doctors and parents are aware of his condition, a suitable treatment and pain management plan has been put in place. This doctor has changed the life not only of this young boy but also of his family. How fortunate they were that this particular doctor, who was knowledgeable about sickle cell disease, was on duty that day. But, honourable senators, health care should not depend on being lucky.

We also heard from Adeniyi Omishore. Adeniyi is only 16 years old, and he was waiting for a hip replacement as the bones have been damaged because of sickle cell. His mobility has been so diminished that he needs a walker or wheelchair to move. He is a very courageous young man, who spoke openly about the challenges of being a teenager affected by this disease. You can't play soccer, hockey or baseball. You miss a lot of school because of the intense pain and time spent in the

hospital for treatment. Yet this young man remains upbeat and positive when telling of his challenges of living with sickle cell. He is truly an inspiration.

We met Adeniyi last spring, and he is still awaiting a hip replacement. He is unable to socialize much due to reduced mobility and constant hospitalizations. He is in Grade 12 but is unfortunately absent on quite a number of school days and is unable to do exams because of sickle cell disease. The good news is that all the surgical papers have been signed, and his family is now just waiting for a surgery date.

Honourable senators, these are just two Canadian stories of many who are living with sickle cell disease. According to the Sickle Cell Disease Association of Canada, over 5,000 Canadians live with debilitating and sometimes life-threatening sickle cell disease. Although the numbers will vary from one province and territory to another, the association reports that one in every 2,500 children in Canada will be born with this condition. In the United States, sickle cell disease is actually the most common genetic disease.

Honourable senators, I would like to take a moment to talk a bit about what sickle cell disease is, who has the disease, and how one gets sickle cell disease. Sickle cell disease or sickle cell anemia refers to a group of inherited red blood cell disorders. The three most common forms of sickle cell disease in North America are hemoglobin SS, or sickle cell anemia; hemoglobin SC disease; and hemoglobin sickle beta thalassemia.

Sickle cell disease is caused by an abnormal form of hemoglobin, the molecule in red blood cells that carries the oxygen throughout the body. With sickle cell disease, the red blood cells become deformed and the abnormal hemoglobin is unable to work properly. Normal red blood cells are doughnut- shaped and move easily throughout the body's blood vessels to deliver oxygen to the organs. In patients with sickle cell disorder, the red blood cells become stiff and sickle-shaped.

The sickle-shaped blood cells do not function like healthy red blood cells. A deformed cell does not flow easily through the blood vessels and can get caught up in the vessels and break apart. This can result in clogged blood vessels and low red blood cell count, or anemia. A normal, healthy red blood cell can carry out its job for 120 days, whereas a sickle-shaped cell has a lifespan of only 20 days. The compounding problem of clogged blood vessels and low red blood cell count drastically hampers the body's ability to deliver adequate oxygen to the organs. The continued starvation of oxygen to the body's systems most commonly manifests itself as severe pain, especially in the bones. It can also cause damage to shoulder and hip joints or cause chest pain from acute chest syndrome. This is why Adeniyi, the 16-year-old boy I spoke about earlier, requires a hip replacement.

There can also be damage to the lungs and heart — such as heart failure or pulmonary hypertension — and damage to the kidneys, liver and eyes. Sickle cell can also cause stroke, leg ulcers and infections. Because it can damage just about every organ in the body, sickle cell disease is known as a multi-system disorder.

Anyone can have sickle cell disease but, for an unknown reason, it is drastically more prevalent in people descended from Africa, the Mediterranean, the Caribbean, the Middle East, Southeast Asia, the Western Pacific Region, South America and Central America. In Canada, the Sickle Cell Awareness Group of Ontario conducted a study that showed 32 out of 40 African-Canadians are carriers of the sickle cell trait. Honourable senators, this is 80 per cent who carry the trait. This does not mean that

they have sickle cell disorder, but they can pass it on to their children if both parents are carriers of the sickle cell trait.

Sickle cell disease is not contagious. You cannot catch it; you inherit it from your parents. To have sickle cell disease a person must inherit one sickle cell gene from one parent and one sickle cell gene from the other. If a child inherits a sickle cell gene from one parent and a healthy gene from the other, they will be a carrier of the sickle cell trait and may pass it on to their children but will never have the disease themselves. As I stated, it is not contagious but, rather, hereditary.

At this time there is no known cure for sickle cell disorders. Treatment consists of managing symptoms of the disease through penicillin to fight infections in children, blood transfusions and a drug called hydroxy-urea. Both transfusions and hydroxy-urea can have serious side effects. However, it is encouraging to learn that research is finding that changes to a healthier lifestyle and diet have a positive effect on patients' quality of life. Because of the lack of oxygen travelling throughout the body, including to the brain, children with sickle cell disorders often struggle in school with fatigue, loss of concentration and memory lapses. It is important for educators to be aware of these symptoms so they can tailor their teaching accordingly.

Infections can also be a major complication of sickle cell anemia, especially during childhood. Early diagnosis is extremely important so that children can be closely monitored by family and medical personnel. Because of its relative rarity, most clinicians have limited experience and expertise with sickle cell disorders. When patients come in with conditions associated with sickle cell disease, doctors may not think to test for the disease.

This was the case for the parents of the young boy who experienced extreme pain in his arm on his visit to the hospital. The lack of awareness of the disease among medical staff can lead to misdiagnosis and ineffective treatments. In some cases, because of the persistent pain and need and desire for pain management medications, it is not uncommon for doctors to dismiss a patient as someone just wanting painkillers — especially if the patient is a teenager or young adult. Something as easy as a simple blood test at birth would help prevent misdiagnosis and would provide medical personnel with the information needed to properly treat the patient.

Universal screening for sickle cell disorders now occurs in every state in the United States. In Canada, newborn screening is available in Ontario, British Columbia, Yukon, New Brunswick, Prince Edward Island and Nova Scotia. Right now, newborn screening is taking place in hospitals in the Greater Montreal area and Laval, with commitments to expand screening across Quebec by March.

Honourable senators, shouldn't every newborn in Canada have access to screening? The screening provides so much information to health care providers patients and their families. Early diagnosis would mean ongoing care from birth. The Sickle Cell Disease Association of Canada is aggressively advocating for a national newborn screening program. A national approach to the disease is something that is sorely missing in Canada. Guidelines for universal screening would identify the disease and other blood disorders. The screening program would also identify carriers. Optimal treatment and management of sickle cell disease requires knowledge and understanding of the disorder, not only by medical personnel but also by the patient

and their family. Managing sickle cell disease is a lifelong process; the logical first step is early and proper diagnosis.

When a child is born with sickle cell disease it is impossible to predict which problems will develop, when they will start, or how bad they will be. During the first six months of life, infants have a high level of fetal hemoglobin in their blood which protects them from red-blood-cell sickling. The dangerous complications of sickle cell disease may quickly develop between ages six months and five years, after levels of fetal hemoglobin decrease. Infection is a major concern for children with sickle cell disorders, and an immediate regimen of daily penicillin is required to manage infection.

Older children and adults with sickle cell disease may have few problems or may have a pattern of ongoing complications such as organ failure or stroke, which can shorten their life. Stroke affects around 10 out of 100 children who have sickle cell disease. Screening all newborns would provide families and their doctors with the information needed to develop a plan of action to manage the disease.

Honourable senators, newborn screening can save lives and improve quality of life. Universal screening for sickle cell disorders of all Canadians will also provide doctors and researchers with the ability to track the disease and, honourable senators, because it is genetic, it can be tracked. It will also provide those planning a family with valuable information about their risk of having children with sickle cell disease. Screening of newborns will also eliminate the mystery surrounding patients with the disorder and will establish early recognition and management of the disease. The longer a child goes undiagnosed, the greater the chance of organ damage, episodes of severe pain or stroke, or possibly death. Early diagnosis means better care and a better health management plan.

Honourable senators, June 19 of each year is recognized as World Sickle Cell Day by organizations such as the African Union, the United Nations Educational Scientific and Cultural Organization, the World Health Organization and the United Nations to promote awareness of the disease around the world. Bill S-211 would add Canada's voice to this important cause by marking June 19 as "National Sickle Cell Awareness Day" in Canada.

We are a diverse country, and many Canadians can trace their roots to Sub-Saharan Africa, India, Saudi Arabia and the Mediterranean, all countries where the sickle cell trait is common. It is important for Canada to recognize this and develop strategies and policies which reflect the ever-evolving health care needs of our citizens.

The Sickle Cell Disease Association of Canada is doing great work to further the awareness of the disease, particularly among parliamentarians provincially and federally. The people I have met and had the pleasure to get to know within the Sickle Cell Disease Association of Canada are very positive, upbeat people. They just want Canadians to be more aware of sickle cell disorders. They want newborn screening to detect sickle cell at birth; they want treatment that will improve the quality of life for those with sickle cell and their families; and they would like to see a national strategy for sickle cell disease. Honourable senators, Bill S-211, which would mark "National Sickle Cell Awareness Day," is just a small step, but it is a positive step in the right direction.

Honourable senators, I am hopeful that you will see the positive change this bill can make in the lives of those Canadians living with sickle cell disease, and I am hopeful

that you will help to make "National Sickle Cell Awareness Day" a reality with the passage of this bill.

Thank you.