

**David C. Dale, MD**  
Professor of Medicine  
**Audrey Anna Bolyard, RN, BS**  
Research Nurse

January 25, 2011

The Honorable Klaus Botzet  
Consul General and Legal Advisor  
Embassy of the Federal Republic of Germany  
4645 Reservoir Road NW  
Washington, DC 20007-1998

Michael Regan  
Director, Office of Children's Issues  
U.S. Department of State  
220 C Street N.W., SA-29, 4th Floor  
Washington, D.C. 20520

RE: Ariana-Leilani King-Pfeiffer  
German Passport No: 875289379 (per mom, Dr. Ariel King)

Dear Mr. Botzet and Mr. Regan:

I am writing on behalf of Ariana-Leilani King-Pfeiffer. The Severe Chronic Neutropenia International Registry (SCNIR) has received the laboratory reports for Ariana-Leilani King-Pfeiffer. The graph of the blood work showing the neutrophil count has been included. The median neutrophil count from the 27 blood counts supplied to the Registry from June 20, 2005 to November 30, 2010 is 518 /cmm (the range is 96 to 3526 /cmm). Neutrophil counts would normally range from 1800 to 7000, the median normally around 4000.

Severe chronic neutropenia is a rare condition wherein the bone marrow doesn't produce sufficient neutrophils to keep the levels in the blood above 500/cmm, resulting in a level of neutrophils too low to fight infection. The diagnosis of benign neutropenia is an older medical term, benign meaning not malignant. The current terminology is mild neutropenia (referring to patients that have an ANC between 1000 to 1500 /cmm), moderate neutropenia (patients that have an ANC between 500-1000 /cmm), and severe chronic neutropenia (patients having neutrophils that are consistently below 500 without G-CSF treatment).

Neutrophils are very important because they fight infection. When bacteria invade the body a chemical signal is sent out and the neutrophils, like fire fighters responding to a blaze, rush to the site of infection. The bone marrow also responds by speeding up its production of neutrophils to replace those involved in fighting the infection. If, however, production of new neutrophils is suppressed or slowed down, a shortage may develop, and any infection can overwhelm the few neutrophils available. Therefore, a person with only a few neutrophils is at particular risk for developing a serious bacterial infection.

Because mortality rates associated with untreated bacterial infections in neutropenic patients are high, the rapid treatment of these patients is critical. Severely neutropenic patients may not always be able to mount a fever, inflammation or the typical response to an infection. It is important to stress that while fever often is the sole sign of severe infection in the neutropenic patient, because of this blunted fever response in some infected patients, fever may be totally absent and they may instead exhibit a severe drop in core body temperature, low blood pressure, listlessness, or confusion. Thus, infection must be considered and treated empirically if any signs of clinical deterioration are present in a neutropenic child, regardless of the recorded temperature.

Many SCN patients are treated with G-CSF, a hormone that increases the neutrophil level. This medication helps fight infection by raising the neutrophil count. However, even with the administration of G-CSF, the neutrophils may still drop to critical levels, and while G-CSF allows the patient to fight disease-causing microorganisms better than the untreated patient, infection is still a constant concern. The treated patient will continue to experience infections, but hopefully not life-threatening infections. The neutropenic person's life may be greatly affected by her/his inability to fight infections.

The SCNIR follows over 1300 SCN patients. We have tracked each of these patients, gathering medical information over the last 16 years for the Registry and 7 years before that in clinical trials.

Our mission is to follow closely the health of neutropenic patients and to continue to research the mechanisms causing this condition. The SCNIR is actively distributing information regarding SCN to doctors and patients. The goal is to help the local physician become more knowledgeable about this rare and difficult condition, and to prevent the severe consequences of untreated SCN: Toxic shock, loss of limbs, and loss of life.

If you have any questions regarding neutropenia and/or the work of the SCNIR, the SCNIR web site is very helpful: <http://depts.washington.edu/registry/>

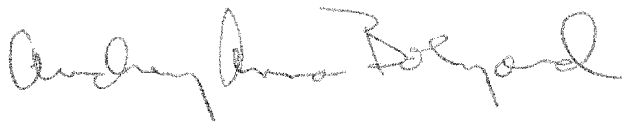
Please feel free to contact me directly at 1-800-726-4463.

Thank you.

Sincerely,



David C. Dale, MD  
Professor of Medicine



Audrey Anna Bolyard, RN, BS  
Clinical Manager SCNIR

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### Ariana-Leilani King-Pfeiffer

