



Cystic Fibrosis in Japan

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Cystic fibrosis (CF) has historically been recognized as a genetic disease that primarily affects people of Caucasian background. As a half Japanese person with CF, my twin and I always wondered why my Japanese mother carried the CF gene. As kids I even remember a nurse who told me, “You can’t possibly have CF, you’re not white!”

We are now learning that CF affects all ethnic groups, including, rarely, those of Asian descent. Many non-Caucasian patients with CF continue to be under-recognized, under-reported, and as a result, under-served. There is no greater truth that the situation for CF patients in Japan. Cystic fibrosis, known in Japan as the complicated condition *suinouhouseisennishou*, remains an unfamiliar condition.

My twin sister, Isabel, and I recently had the unique experience of visiting Japan and meeting other CF families for the first time. One CF mother, Rumiko Aoyama, had lost her child at the age of 18; the other parent has a son who is waiting for a lung transplant; and the other parent has an infant who was just diagnosed at the age of 18 months despite being born with meconium ileus. This was immensely meaningful for us, since we had never met anyone of Japanese ancestry with CF in our entire 37 years of life.

Our memoir, *The Power of Two: A Twin Triumph over Cystic Fibrosis*, which was published in the USA in 2007, was translated into Japanese by Iwanami Shoten Publishers in September, 2009 under the revised title of *Miracle Twins: Overcoming Serious Illness through a Twin Bond*. We spent the month of October 2009 in Japan promoting our book and, more importantly, raising awareness about cystic fibrosis and organ donation throughout Japan. In 25 days, we travelled to 9 cities and gave 20 speeches in Japanese to medical professionals, students and the general public.

In Japan, approximately 1 out of 350,000 babies is born with cystic fibrosis. There are less than 30 CF patients living in Japan, and all are geographically dispersed throughout Japan. There is no centralized CF care specialist or fundraising organization. The CF Japan Network, headed by Rumiko Aoyama, is a CF family support group that provides support and information for families whose child has received the diagnosis.

On October 8, 2009, Isabel and I participated in a CF awareness concert in the northern city of Sendai, Japan. Miyagi Women’s University and the Committee to Enable CF Treatment hosted the concert. This Committee was founded in June, 2009 by Dr. Tomoaki Adachi, whose 21 year old son has CF. The moving concert featured the talented folk singer Mitsutoshi Ambe and raised over \$10,000(USD) for CF advocacy in Japan. Video footage of this concert can be seen at:

<http://www.youtube.com/thepoweroftwomovie#p/u/3/Z1kvA984iYQ>.

Japanese CF patients are often diagnosed late and do not have access to standard life sustaining CF medications such as enteric-coated pancreatic enzymes for digestion (for example, Creon™ or Ultrase™), inhaled antibiotic medication (TOBI™ and Colistin™), and inhaled mucolytic enzymes (Dornase alfa-Pulmozyme™). Importing these critical CF medications into Japan is forbidden by the Japanese government because of concerns about the lack of clinical trials on Japanese patients.

As a result, in this wealthy nation, the current median life expectancy in Japan is only about 15 years of age. Moreover, Japanese CF children can receive a government



subsidy to pay for medications through a special program for those with pediatric chronic illnesses only until the age of 20. However, this supplemental government subsidy is cut off when the CF patient reaches 20 years of age, resulting in exorbitant medical bills for families of adult CF patients.

Some CF families are going through extraordinary means to obtain medications, such as seeking CF care abroad or paying significant fees for enteric-coated enzymes through special clinical trial programs.

In addition, sociocultural views towards the disabled and chronically ill limit opportunities for people with CF in Japan. The rigors of the Japanese education system make it very difficult for children with CF to keep up, yet there are no formal home-school programs to help children meet their educational needs. Frequent and lengthy hospitalization is not uncommon, and only recently has home intravenous antibiotic therapy been offered to CF families. Issues of shame and stigma towards difference can lead to loneliness and isolation. A rigid medical hierarchy makes it difficult for CF parents to question the physician and make demands for better treatment. All these factors can impact quality of life for CF patients and their families.

During our visit to Sendai, I was struck by the injustice of the situation. Why should our country of birth determine our access to care and in essence, our life expectancy? Japan is a highly developed, medically advanced country that is capable of providing quality medical care for its few CF patients. We feel strongly that Japan must narrow the gap of CF treatment, in comparison to Europe and the U.S.

We believe that the first critical step is through medical education about CF to Japanese healthcare providers, especially pulmonologists. Very few healthcare providers that we met in Japan had ever heard of cystic fibrosis. Yet, ironically, there are several Japanese scientists performing basic CF research on CFTR within Japan.

While we were in Japan, we hoped that by speaking to numerous medical schools and health care professionals, more people would become aware of CF as a rare but very possible diagnosis for children with frequent pneumonias, malabsorption or other CF symptoms. It is our wish that the publication of our book in Japan will increase public awareness about cystic fibrosis and organ donation in Japan.

Through the efforts of the Committee to Enable CF Treatment in Japan and the CF Japan Network, volunteers made up of CF parents are lobbying the Japanese government to allow CF medications into the country. The current law requires clinical trials, but this is impossible with so few patients. They are also urging the government to amend the healthcare coverage situation for CF adults in Japan. The need is urgent, as patients living with CF are becoming sicker each day without proper medications. This appeal may also impact the quality of lives of several foreigners living in Japan with CF.

In addition, lung transplantation in Japan is very limited due to cultural beliefs against organ donation. We learned that there is no universal acceptance of brain death as death in Japan, and despite the valiant efforts of a few strong transplant advocates, Japan continues to have the lowest rate of organ donation in all industrialized countries. In 2009, there were only 6 brain dead donors in all of Japan. Therefore, the prospects of lung transplantation for patients with end stage CF are slim, with an average wait of approximately 3-4 years.



We would like to appeal to the international CF community to understand the difficult situation for cystic fibrosis patients in Japan who lack adequate medication and treatments. The Committee to Enable Cystic Fibrosis Treatment in Japan would also like support from the international community. If you are interested in helping this situation by signing an online petition prior to Jan. 15, 2010, please see <http://www.PetitionOnline.com/cf2010/petition.html>

For more information about our book tour in Japan, and the documentary film about our CF and organ donation awareness efforts in Japan, please see: www.thepoweroftwomovie.com.