Cystic fibrosis

Danielle

Danielle did it,
Said, "You here, you there, and me
in my Daddy's arms
in my Mama's hands."

She said, "Bloom!" to the amaryllis,
ordered Stevie Wonder's "Overjoyed,"
the clothes for her casket,
a rose, a sand castle,
Gilbert bear,
the autopsy.
She made scrapbooks of her life
and a will.
Told people off,
gave away her toys,
said last things.

"Don't go! Don't go! I'm going to die today!"
Her doctor was driving back from New Brunswick
we post his picture on the IV pole, eye-level.
Wait for him. Wait.

But of course, she'd never died before, she didn't know. It would take
10 more days.
"Read me the *Velveteen Rabbit*."
We had already worn her away
to a taut salty skin
stretched in the dusk
she'd gathered to, closing the curtains.
She laboured for breath and sleep
leaning on the bed table,
a swagger of thick brown healthy hair
on the white pillows.
She needed crushed ice,
cherry Popsicles at midnight, her head held in our hands, her cheek in
the cup of my palm, so close I could breathe the leftovers from her
mask.
But she does it: the boss
calls the 14 angels
to keep her round
and talks so familiarly of heaven
we see everyone clustered there
at the gate and we're the sorry ones
soulless, left behind
right here on earth
listening to her shout into the stethoscope "Pray for me. Pray for
me."

—*Susan Kerslake*

Ms Kerslake is a published novelist and poet. She has been a volunteer in the care of children with cystic fibrosis for more than 30 years at the IWK Hospital for Children in Halifax, NS. Her poem "Danielle," written in the early 1980s, is a lucid, euphonic tribute to a child's wonderment and indomitable spirit.

The science

In cystic fibrosis, gene mutations lead to defects or deficiencies in cystic fibrosis transmembrane conductance regulator protein. This causes problems in salt and water movement across cell membranes, resulting in abnormally thick secretions in various organ systems and critically altering patient lung function.[1]

Treatment includes pancreatic enzymes, caloric and fat soluble vitamin supplements, airway clearance therapy, aggressive antibiotic use, and medication to improve regulator protein function. Current areas of research include gene therapy.[1]

Several decades ago cystic fibrosis was fatal in childhood. The median age of survival for Canadians with cystic fibrosis is currently estimated to be 50.9 years of age.[2]

References

1. Federico MJ, Baker CD, Balasubramaniam V, Deboer EM, Deterding RR, Halbower A, et al. Respiratory tract & mediastinum. In: Hay WW Jr, Deterding RR, editors. *Current diagnosis and treatment. Pediatrics.* 22 ed. New York, NY: McGraw-Hill Education; 2014. p. 534-87.
2. Cystic Fibrosis Canada. *2013 Annual report. The Canadian Cystic Fibrosis Registry*. Toronto, ON: Cystic Fibrosis Canada; 2015.

SICKBOY PODCAST

Sickboy

In *Sickboy Podcast* (**www.sickboypodcast.com**), Jeremie Saunders, a 27-year-old with cystic fibrosis, and his friends Brian Stever and Taylor MacGillivary and their guests use explicit humour and frank self-revelation to discuss what it is like to live with cystic fibrosis and other chronic illnesses, in an effort to reduce the stigma associated with such conditions. Listener discretion is advised.

Salty girls

Photographer Ian Pettigrew, based in Hamilton, Ont, is a 46-year-old living with cystic fibrosis. In his book, *Salty Girls*, he strives to create awareness and put an end to body shaming by photographing women with cystic fibrosis exposing their postsurgical scars, ports, and other physical marks of the condition (**www.ianpettigrew.com/salty-girls.html**). The project speaks to the strength and resilience of many living with chronic conditions.