King firefighters step up to help Vaughan family

By Mark Pavilons

King firefighters are joining forces to help raise money for a Vaughan child fighting a rare disease.
Rebecca Grace Silva daughter of Vaughan firefighter Mark Silva, is fighting through Spinal Muscular Atrophy (SMA) Type 1.
Fundraising efforts have been ongoing to raise enough money to get a very expensive drug in the United States. Zolgensma, not available in Canada, costs $2.124 million U.S. It's heralded as the most expensive treatment in the world. The one-time treatment will extend Rebecca's life.
Children with this condition often don't make it to their second birthday. Rebecca is just over 8 months old.
The King City Firefighters Association have lined up some candidates who are getting an ?epic mustache shave??in support of Rebecca. High-ranking personnel at the station will be saying good-bye to their staches, one of which was 20 years in the making!
The fundraiser will be held at Rockford's Bar &?Grill in King City Saturday, Feb. 1, starting at 6 p.m. Along with the shaving, Rockford's is donating $1 per drink to the cause.
The diagnosis of Spinal Muscular Atrophy (SMA) for Rebecca Grace has been devastating. A healthy body contains two copies of the Spinal Motor Neuron (SMN) 1 gene and two copies of the SMN 2 gene in each cell, however, the number of copies of the SMN 2 gene varies. Some people have up to eight copies. Rebecca has 0 copies of the SMN 1 gene and 2 copies of the SMN 2 gene.
This disease affects Rebecca's motor nerve cells in her spinal cord, taking away her ability to walk, eat, and breathe. SMA does not affect Rebecca's mental or cognitive abilities. Sadly, 95% of babies with SMA type 1 do not reach their 2nd birthday and it is the number one genetic cause of death in infants.
Rebecca received her first dose of Spinraza. Spinraza is a Spinal Motor Neuron (SMN) enhancing therapy that works by targeting the SMN2 gene, causing it to make more complete protein.
The family is very thankful that this treatment is available in Canada and hopeful that Spinraza can slow the progression of the disease in Rebecca's body, until they can raise enough money for Zolgensma.
Zolgensma is a single one time infusion therapy given intravenously which only takes about 60 minutes. It targets the genetic root cause of SMA by replacing the function of the missing or non-working survival motor neuron gene (SMN 1). The new gene tells motor neuron cells to produce more survival motor neuron (SMN) protein. Motor neuron cells need this SMN protein to survive and support muscle function.
Mark Silva pointed out this drug is still new, being out only three years now. There's still a lot of unknowns about the long-term success for the medication. But there is hope. The manufacturers holding ?draws??twice a month, to offer patients the drug, in countries where it's not approved. The Silvas have filled out all the applications and have their fingers crossed.
But time is of the essence for Rebecca.
She was born May 30, 2019 and was diagnosed with SMA Type 1 in November.
Her parents point out this is already a long time to have lived with the SMA disease without treatment. Awareness of this disease is lacking. SMA is not on the radar for most pediatricians. Rebecca has been misdiagnosed since birth. Rebecca is not able to sit on her own or keep herself supported. Her movement in her arms and legs are minimal. As time passes, she will continue to lose these functions.
With each day that passes without treatment for Rebecca, her motor neuron cells will continue to become weaker and weaker and eventually die. Once her cells stop working, they cannot be brought back.
Mr.?Silva pointed out one of the drawbacks with Canada's health system is we lag behind other countries, particularly the U.S. in getting drugs to market in a timely fashion.
The family is hosting a sold-out fundraising dinner on Valentine's Day.
Donations are being accepted through their page at gofundme.com/f/hope-for-rebecca-grace