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Let's turn grief for 'HD Angels' into new impetus for the Huntington's cure

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At Risk for Huntington's Disease

HD is a genetically caused brain disorder that causes uncontrollable bodily movements and robs people's ability to walk, talk, eat, and think. The final result is a slow, ugly death. Children of parents with HD have a 50-50 chance of inheriting the disease. There is no cure or treatment.

TUESDAY, JANUARY 17, 2012

Let's turn grief for 'HD Angels' into new impetus for the Huntington's cure

The Huntington's disease angels are sending us all an urgent message: we must redouble the effort to find effective treatments and a cure for this devil of a disease.

In the past few days, two more HD angels – children who have succumbed to this disease – have passed on.

On January 11, nine-year-old Kathleen Edward died while surrounded by loved ones in her Wyandotte, Michigan, home.

Kathleen and grandmother Rebecca (family photo)

On January 15 another child, twelve-year-old Olivia Ruggiano, died in Philadelphia.

While HD affects people of all ages, the ten percent of cases classified as juvenile Huntington's disease (JHD) wrenchingly dramatize the disease's crippling effect. Children and teenagers afflicted with JHD never experience a normal life. As in Kathleen's and Olivia's cases, some don't even reach adulthood.

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Huntington's Disease Society of America
International Huntington Association
Huntington's Disease Drug Works
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HD Blogs and Individuals

Chris Furbee: Huntingtons Dance
Angela F.: Surviving Huntington's?
Heather's Huntington's Disease Page

Together with Kathleen's and Olivia's families, the HD community grieves deeply: two young lives ended hopelessly.

Their deaths provide a startling reminder of the lack of treatments.

These HD angels want us all to cry out for increased funding for HD research – including the understudied juvenile onset – and a commitment from drug developers to broaden and speed up the search for treatments and a cure.

Two brave girls

In life, both Kathleen and Olivia had received an outpouring of sustenance from the HD community and beyond.

In 2010 a hateful neighbor, upset over a misunderstanding about a children's birthday party, started feuding with Kathleen's family. The neighbor bullied Kathleen on Facebook by posting a photo of the girl positioned over a set of crossbones. Another photo showed Kathleen's HD-stricken mother in the arms of the Grim Reaper.

News coverage of the incident spurred donations to the family and expressions of support from around the world. Thanks to many generous individuals, Kathleen had the opportunity to go on a shopping spree – but only after first choosing gifts for her family members. (Click here to read more about this incident.)

"Olivia was a normal child who loved to wear frilly dresses with dirty knees while digging for worms," Olivia's mother Jennifer wrote on a fundraising site for the Delaware Valley Chapter of the Huntington's Disease Society of America (HDSA). "Strong willed but always ready with a smile, giggle or hugs. She began showing developmental delays at 4 1/2, a slight lisp, uncoordinated, tripping a lot, dropping things."

Olivia and friend at HDSA fundraising walk

"Then the focal seizures came, the first one being discovered the last day of day care before starting kindergarten," Jennifer continued, referring to

how Olivia would lean back and prop up her head, and then let her head drop into her lap. "We thought she was just being difficult or having selective hearing. Then they developed into stronger more intense seizures. Late in December 2007, right before Christmas she went into status (non-stop seizures).... Since then she has been on a spiral downhill."

On a Facebook page titled "We All LOVE Olivia Ruggiano," supporters and members of the HD community left Olivia a constant stream of encouraging messages as she struggled against the disease. Jennifer and other family members read the messages out loud to Olivia and, holding up a laptop, showed her the pictures people had posted.

Words of support

At Kathleen's funeral on January 14, so many mourners turned out that the funeral home ran out of the flags fastened to cars in the procession to the cemetery, Rebecca told me via Facebook. Kathleen was buried next to her mother Laura Edward, who died of HD in 2009.

The *Detroit Free Press* ran two articles about Kathleen.

"Those who knew 9-year-old Kathleen Edward will never forget her infectious smile, one seen throughout her battle with juvenile Huntington's disease," one of the articles began.

The paper quoted grandmother Rebecca: "She suffered with this disease for a while, and she never complained," Rebecca told the paper. "She was always happy, always smiling."

You can watch a tribute to Kathleen in the video below.

After the two girls passed, scores of Facebook members expressed their condolences.

"We are all in mourning over Olivia leaving us," one HD activist wrote on Olivia's Facebook page. "Heaven is rejoicing to receive angel Olivia where she can be closer to everyone's heart."

"Know that we are with you in spirit," another supporter wrote. "It's going to be hard, but as Olivia was strong, you are also. Smile thru your tears. She is free. Love to all of you."

News of the girls' deaths and condolences also went out on other HD Facebook pages, including two dedicated to HD angels, Rebecca's page, and Olivia's mother's page.

Viewing of Olivia will take place at Stolfo Funeral Home in Philadelphia 7-9 p.m. on January 19 and 8:30-10:30 a.m. on January 20, followed by a funeral Mass at 11 a.m. at Stella Maris Parish. In lieu of flowers the family requests donations be made to the Philly HERO Trust, P.O. Box 18008, Philadelphia, PA 19147.

Remembering the Ruggianos' fight

In a phone conversation last night, Jennifer graciously recalled Olivia's and her family's fight against HD.

Jennifer started dating Ron Ruggiano in 1994, the same year his mother died of HD. Jennifer and Ron married in 1996.

As in many HD families, the disease was "a taboo subject in his family," Jennifer said. But his mother's death alerted Jennifer to the existence of the disease in the family. She contacted HDSA and learned what she could about the disorder, including the fact that a male could pass on a far worse form of the disease than he himself has.

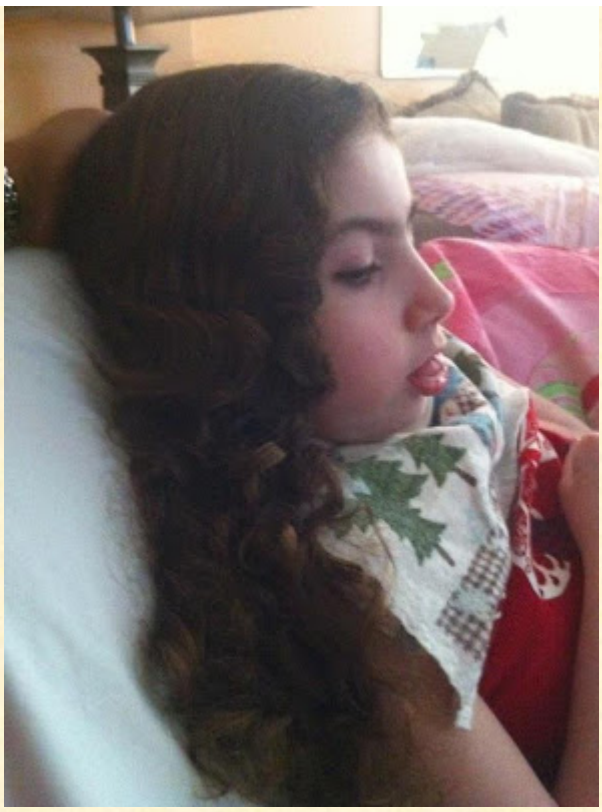
Ron had not been tested for HD, so the couple knew that having gene-negative children was "a roll of the dice," with a 50-50 chance of their children inheriting the defective gene, Jennifer recounted. In 1997 they had a daughter, Rania, who has not demonstrated symptoms of JHD (but has not been tested). Two years later, Olivia arrived.

Ron started to show the behavioral difficulties that often occur in the early stages of HD. In 2000, he was clinically diagnosed with the disease.

An astounding level of disease

On that fateful last day of daycare before kindergarten when Olivia had seizures, "she was speaking gibberish and not making any sense at all," Jennifer remembered. "It just mushroomed from there. She was clumsy. She would fall down and trip a lot."

Nevertheless, Olivia entered elementary school, where she participated in a small life skills class for the severely disabled. She stayed almost through the end of fifth grade.



Olivia (family photo)

"Sometimes she didn't want to go to school," Jennifer said, chuckling, "but she went. Sometimes she would fake her seizures. She was a smart little cookie."

In 2007 both Olivia and her father finally underwent HD testing at Johns Hopkins University in Baltimore. Whereas a normal huntingtin gene has only ten to 29 repeats, Ron's had 50 – ten repeats beyond the level that causes HD. Olivia had an astounding 109 repeats – a number that doomed her to childhood onset and an early death.

Olivia's joys

Despite this terrible fate, Olivia strived to live like any child.

"She loved to dance," Jennifer reminisced. "She loved to sing. She loved to watch musicals: of course, the *Wizard of Oz*, *Grease*, any kind of musical, *Hairspray*, all the Disney movies."

Olivia also loved to help her mother cook and care for the home. She wouldn't miss a chance to play in the pool at her cousin's house or visit the New Jersey shore with her family.

Olivia was a "little devilish" in everything she did, Jennifer said. And she had a fascination with bugs. "She could spot an ant ten feet away," Jennifer said, laughing.

With Ron unable to work, he received Social Security disability payments. Olivia further supported the family with her salary as a legal clerk in the Pennsylvania courts. A heavy emotional burden also fell onto Rania, as she watched her mother pay ever greater attention to Olivia.

The family dealt with HD as proactively as possible, Jennifer explained. In 2008 doctors gave Olivia a feeding tube to keep her properly nourished.

Meanwhile, in February 2010, Ron entered a nursing home, where, at 43, he struggles against HD.

A turn for the worse

Olivia's health worsened dramatically in December 2010. She spent nearly the entire month hospitalized.

Just ten days after returning home, she developed an infection. "That's when I made the decision not to take her back to the hospital," Jennifer said. She decided that Olivia would live at home until she died.

Olivia continued to decline throughout 2011.

"She was on so many medications," Jennifer recalled. "She was taking 40 milligrams of valium every four hours."

Along with Olivia's nurses, the family kept Olivia as comfortable as possible. On Thanksgiving Day, for example, they dressed her up in a pink dress and sweater and placed a pink flower in her hair. "Her nurses absolutely spoiled her," Jennifer said.

Olivia at home on Thanksgiving Day, 2011 (family photo)

Olivia's legacy

I asked Jennifer about Olivia's legacy.

"She's just another bright light," she said, "another child that just fought … and fought … and fought."

As an HD angel, Olivia will inspire others to fight – and will also contribute to the search for treatments and a cure. Months after undergoing genetic testing, both she and Ron donated cells for research. As Jennifer explained, researchers were able to make the cells from Olivia's forearm reproduce and are hoping to induce them into becoming stem cells. Olivia's cells could eventually end up in labs around the world that focus on developing treatments for HD.

Time for treatments

After learning about Kathleen and Olivia, my wife and I became saddened and distraught. Once again we relived the painful moments of 1999 and early 2000, when I tested positive for HD and we subsequently tested our daughter in the womb.

Our "miracle baby" tested negative, but had she inherited the HD gene, my wife would now face the terrible prospect of caring for two HD patients.

I became angry and depressed that HD had once again victimized families, and I feared that my own symptoms might start soon, leaving my unable to work and to enjoy my own family.

I also felt the urge to fight back.

"We must find treatments and a cure so that no more children suffer with Huntington's disease," I wrote on Olivia's page.

The angels have fought bravely, but our community wearies as it sheds yet more tears of sorrow.

We need treatments *now*.

Posted by Gene Veritas at 8:05 AM

Labels: CAG repeats , cure , death , gene , gene-negative , genetic , genetic testing , Huntington's , Johns Hopkins University , juvenile Huntington's , research , seizure , stem cell , symptoms , treatments

1 comment:

Kate's Kronies said...

Gene,

your blog has touched our hearts.. Landon was so scared and so sad about these two very brave girls.. He woke up this am and looked at me and said mommy cant we tell the dr that these kids are dying? cant they find a cure mommy please.. than he he turned around and said "mommy what about baby Noah?? Kate baby mommy will he have JHD?? I said we don't know... He looks at me and said mommy there is cure.. we just have to find it.. I don't want to loss any more..

He took a bag of buttons and key chains to school and I received a call this noon. Landon is mad and to tell you the truth so am I! I am fighting and days get long when there seems to be little roward progress but I beileve that a cure is out there. God please be with both these families, offer them comfort and hope. AMEN.

and to both families, Thank you for sharing your children with us.

Tara and KAtelyn and Landon and the rest of Kate's Kronies

11:54 AM, January 17, 2012

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