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A twinkle of hope

Family in New Albany marking fight against disorder one light at a time

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THE COLUMBUS DISPATCH

Tom Mouat drove home from Children's Hospital yesterday afternoon to string up another 500 Christmas lights. Christmas might have come and gone, but his holiday lighting display is only getting larger.

Mouat and his wife, Jennifer, have been adding a Christmas light for every dollar donated to the Propionic Acidemia Foundation, which funds research into the rare genetic disorder that prohibits the body from breaking down certain proteins.

Their 10-month-old daughter, Gwen, has a severe case of the disorder.

Mr. Mouat started with 435 lights Nov. 28. After last night's addition, there are more than 5,000 lights twinkling on the house, porch, garage, bushes and trees at their New Albany home.

The "Lights of Love," as the Mouats call the display, is the latest in the fundraising efforts of the Mouats and of Dave and Michelle Ellis, of Lewis Center. One-year-old Allison Ellis also has propionic acidemia. The two families met through Children's Hospital and in August began working together to raise money for the PA Foundation, of Highland Park, Ill.

Michelle Ellis said she has been selling lights to everyone she knows.

"We're just trying to help their house look like the Griswolds'," she said, referring to Chevy Chase's brightly illuminated home in the movie Christmas Vacation.

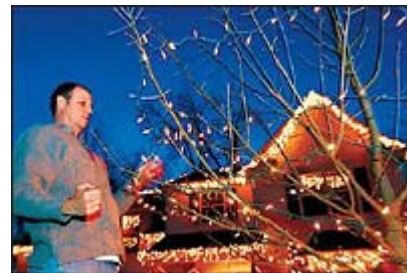
In five months, the two families have raised about \$27,000 for the PA Foundation. That is more than half of what the foundation typically raises each year, said its president, Jill Franks.

"They are amazing," Franks said. "They have embraced their situation and are making a huge difference."

With the infusion of money raised in central Ohio, the foundation is going to fund three research grants this year instead of two, Franks said. The small grants allow researchers to gather data that allow them to chase large National Institutes of Health grants.



Gwen Mouat



Tom Mouat is running out of places to add lights. His fundraising efforts have made new research grants possible.



JAMES D. D

DISPATCH PHOTOS

Jennifer Mouat adds strings of holiday lights to the roof of the family home in New Albany. A new light is added for every dollar raised for the Propionic Acidemia Foundation. More than 5,000 bulbs now adorn the home, shared with Gwen, a 10-month-old afflicted with the genetic condition.

It's difficult to place a life expectancy on infants being treated for propionic acidemia because for so long the disorder had gone undiagnosed. Dr. Britton Rink, a fellow in genetics at Children's Hospital, said some cases of sudden infant death syndrome likely were the result of propionic acidemia. Babies who are not treated for the disorder stop breathing, usually in their sleep.

More than a year ago, Ohio mandated a new batch of genetic tests for newborns, and one of those tests checked for propionic acidemia.

The Mouats had never heard of propionic acidemia despite both being recessive carriers of the mutated gene before Gwen was diagnosed.

Since then, their life has been full of learning what proteins can and can't go into their daughter's body. They've learned how to mix formula from five ingredients, all meticulously measured on a gram scale. Gwen is fed every three hours with that formula, and at night she receives a continuous pump of formula through a gastrointestinal tube.

"Anything that upsets that caloric intake can send her into crisis mode," Mrs. Mouat said.

It was a viral infection that sent her to the hospital Thursday. She should come home today or Sunday, Mrs. Mouat said.

Since Ohio started testing for the disorder, four cases of propionic acidemia have been diagnosed at Children's, Rink said.

"These kids get sick quickly, and you don't always have a lot of evidence that something bad is going on," she said.

Doctors manage the disorder mainly through diet and close monitoring, Rink said. They just now are learning how it affects other parts of the body. For instance, Gwen also has acid reflux, something likely attributed to propionic acidemia, Rink said.

"There's a lot we don't know because the kids didn't live this long," Rink said.

Despite Gwen's trip to the hospital, Rink said she looked good. She started crawling two days before Christmas, Mr. Mouat said. Yesterday, after babbling at the ceiling and chewing on the plastic IV tube that was flowing fluids into her chunky body, she rolled over and started crawling on the mattress.

"What are you doing, Peanut?" Mrs. Mouat asked.

She scooped her out of the crib and held her on her knee. Gwen smiled. The only proof she wasn't like most children was the IV tube snaking out of her pink outfit and running to the pump beside her crib.

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