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# The blue light kids

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Across the moonless dark of Lancaster County, where horse-drawn buggies clatter along dusty roads and many families shun electricity, a strange blue light cuts through the night.

Over the cornfields it beckons, like some otherworldly force, beaming from the bedroom window of a 100-year-old Mennonite farmhouse.

Downstairs, flaxen-haired girls with braids read to younger children…a mother in a traditional long print dress and white organdy cap rocks a slumbering child…a father returning from the fields pulls up a chair to the coal-fired stove. The scene is bathed in the glow of a single gas lamp.

Upstairs, a baby sleeps in another kind of light, in a very different world.

High-intensity blue electric rays burn down upon his crib. The lights are suspended from a heavy stainless steel canopy just inches above the child.

The baby wears only a diaper and has no blankets, just starched white sheets. Mirrors are built into one side of the crib. Fans hum loudly to keep him cool.

With his chubby cheeks and bleached blonde hair, 15-month old Bryan Martin looks like an angel in his luminous cocoon. But Bryan is a very sick child.

The whites of his eyes are yellow and his skin is an unnatural gold. The blue lights are saving his life.

**Sign of the disease**

In the lush, green pastures of Pennsylvania Dutch country, where life revolves around the one-room schoolhouse, the farm and the church, and locals speak a distinctive German dialect, the strange blue lights beam from a handful of homes.

To the Amish and Mennonites they mean one thing — the presence of an extraordinarily rare disease that seems to cruelly target their communities, forcing afflicted children to spend 10 to 12 hours a day, undressed, under lights.

The children suffer from a genetic disorder that causes high levels of a toxin called bilirubin to build up in their bodies, resulting in severe jaundice that, if untreated, causes brain damage and death.

Bilirubin, a natural waste product from worn-out red blood cells, is normally broken down by an enzyme in the liver. If the enzyme is missing, bilirubin can be checked only by the wavelengths of blue lights. Levels must be monitored constantly. Even minor injuries or infections can cause them to rise dramatically.

The disease is Crigler- Najjar syndrome, named for two doctors who identified it 55 years ago. There are about 110 known cases of Crigler's worldwide, including about 35 in the U.S. About 20 are among the Amish and Mennonite in Pennsylvania.

There is no cure; Bryan's only hope for long-term survival is a liver transplant.

**Unusual treatment**

As a Mennonite, Katie Martin embraces the teaching of her church, that sick children are gifts from God, born to foster compassion and understanding.

But nothing prepared her for the news that her firstborn, Derick, had Crigler's. Several years earlier, a nephew had suffered brain damage and died of the disease at age 3.

"I thought it was a death sentence," she said.

In the past, it usually was. But in 1990 a new clinic had just opened in Strasburg specializing in children with rare diseases. There, the Martins met a doctor who had once studied with Dr. John Crigler, who first described the disease with Dr. Victor Najjar in 1952. The doctor told them about bilirubin levels and the dangers of kernicterus, the brain disorder that killed their nephew.

Bring the baby back for blood tests every month, the doctor told them.

And keep him under blue lights.

So the Martins — who are unrelated to Bryan Martin — took their yellow baby back to their 140-acre dairy farm in Mifflinburg and embarked on a life of testing, monitoring and lights.

Floyd, who also works as a welder, fashioned a stainless steel-framed canopy to hold the lights over his son's bed. He learned all he could about phototherapy, as the blue light treatment is called. As the boy grew, Martin made bigger, more sophisticated frames. When his next child, Amy, was born, he made another set of lights. When their three cousins across the hill were stricken, he made more.

Today, Floyd Martin's blue light beds, which cost about $1,000, are sought by Crigler families all over the world.

The Martins, old-order Mennonites, have electricity and a phone, but there is no computer, television or radio in their house. They travel by horse and buggy, except for emergencies when they hire a driver.

They had no moral qualms about using electric lights, as some more conservative families do.

But the disease forced other compromises, like accepting state insurance for their sick children, even though church rules forbid any form of government help. Generally, the church pays for all medical care.

"The hardest thing," says Katie Martin, a slender woman of 37 with a pale, thin face and dark brown eyes, "was to hear them cry on cold winter nights and not just be able to wrap them in a blanket or curl up in our bed."

She is standing in the brightly lit cow barn, overseeing the noontime milking of 65 Holsteins. Derick, now a strapping young man of 17, hauls long milking tubes along a motorized pulley. Amy, 15, attaches them to the cows.

The teenagers radiate sturdiness and health. Still, their mother eyes them nervously.

For years she has worried about bilirubin levels.

Now she has another worry: liver rejection. Both children have had transplants in the past three years, and for Amy recovery was complicated. Ulcers. Lesions. Diabetes. High doses of steroids and anti-rejection therapy. Months of hospitalization. Martin estimates that Amy's total medical costs have amounted to more than $1 million.

And yet, Martin knew her daughter had no choice. Bilirubin builds up dangerously in adolescence as skin gets

more dense. And the psychological toll can be devastating.
For years, Martin received sad, lonely letters from a woman in England who survived Crigler's into early adulthood. In 2004, at the age of 30, she smashed her bed of lights. The disease killed her within a few weeks.
Martin tells this story to panicked parents who call from around the world when they have a yellow baby and they don't know where to turn.
She tells them exactly how to set the lights. She tells them what drugs to use and what to avoid. And she tells them about a special place and a doctor who is an expert on the disease.
"Go to Dr. Morton," she says. "He can save your child."

**'God sent Dr. Morton'**

On a dewy spring morning in Strasburg, strains of Bach drift from a post-and-beam building on a hill overlooking an alfalfa field. Inside, rays of light wash down from rafters, silhouetting the doctor-musician as he cradles his cello.
For Holmes Morton, his daily dawn "concert" is a rare escape from the sadness of sick children and the desperation of parents who come to him for miracles.
There are no miracles, he tells them. For many rare diseases there are no cures.
To the families who travel from miles around, Morton's Clinic for Special Children is itself a miracle.
Here, on what was once an Amish farmer's field, in a building erected in traditional barn-raising fashion by 70 local men, some of the world's rarest diseases are identified. Children who would never have survived in the past are treated with special formulas and dietary regimens tailored solely to their needs. And be-
cause the local community helps pay for the nonprofit clinic through annual auctions, costs are far less than at a regular doctor's office.
Geneticists have long studied the Amish and Mennonites, descendants of Swiss and German Anabaptists who settled in Pennsylvania in the 1700s. Forbidden to marry outside their religion, the Plain People, as they are known, have a relatively high risk of being carriers of a rare disease.
But research generally takes place in university laboratories, far from actual patients and their illnesses.
At the Clinic for Special Children, laboratory director Erik Puffenberger studies a mass spectrometer and DNA sequencing machine in one room, while across the corridor an Amish family clusters around Morton to discuss their sick child. Heirloom quilts decorate the walls. A horse and buggy is tethered to a hitching post outside.
And new genes are being identified all the time.
"The real frontier of genetic medicine is in the everyday practice," Morton says, as he bounces over hilly back roads in his silver Jetta, waving at bearded farmers and straw-hatted boys.
With his thinning hair, walrus mustache, starched white shirt and bow-tie, Morton looks every inch the genial country doctor. But the 56- year-old Harvard-trained pediatrician is far more. In 1989 he gave up a promising academic career to pursue his vision of the clinic, believing that the only way to understand rare diseases was to live in the communities where they occurred. Today, the clinic, which is run by Morton's wife, Caroline, treats about 600 Amish and Mennonite children. Morton's work is recognized around the world.
"These children are living with the sword of Damocles," Morton says. "They need treatment, not just research."
Morton is speaking not just of Crigler-Najjar syndrome, but of the many other rare disorders seen in the clinic.
Maple syrup urine disease. Glutari aciduria. Pigeon breast disease. Pretzel syndrome.
Many of the disorders can be fatal — or cruelly disfiguring — if undetected. Like Crigler's, many are so unusual they are simply not recognized by general pediatricians.
"God sent Dr. Morton to us," says Norman Burkholder, after leaving his mules and plow one day to bring in his 9-year-old son. The child is dizzy and coughing, and he complains, in Pennsylvania Dutch, that his stomach hurts.
Later the boy will be admitted to Lancaster General Hospital where he will spend days on a special formula prepared by Morton's clinic. The child has maple syrup urine disease, a rare enzyme deficiency that causes his urine and ear wax to smell like maple syrup. If he had not been properly diagnosed and the formula had not been available, he could have slipped into a coma and died.
Like Crigler-Najjar, there is no cure. The boy will eventually need a liver transplant.

**The power of light**

John Crigler remembers being baffled by the jaundice disease he encountered among Amish newborns when he was a young pediatrician working with Najjar at Johns Hopkins Hospital in Baltimore. The babies all died.
"There wasn't any treatment, any hope, any cure," said Crigler, now 87. "We were just spectators. There was nothing we could do."
Patients began living longer in the 1970s when doctors realized that the wavelength and energy of blue light changes the nature of the bilirubin, allowing it to be excreted from the body.
There was even a brief time, in the late 1990s, when a cure seemed imminent. Experiments in rats suggested that chimeraplasty, a form of gene therapy, could also succeed in humans. The therapy is based on the use of a molecule called a chimeraplast, a synthetic blend of DNA and the related molecule RNA, that would induce a patient's own cells to repair themselves.
At a conference of Crigler families in July 1999 Morton announced that the first human trials would begin on three of his patients in Lancaster General Hospital that fall.
"There was such excitement," recalls Katie Martin, who was pregnant with her third child at the time. "We thought that soon we would get rid of the blue lights forever."
Two months later an 18- year-old Arizonan named Jesse Gelsinger died during an unrelated gene-therapy trial at the University of Pennsylvania. Funding for human trials dried up.
So did hope for Derick and Amy Martin.
Liver transplants are expensive and invasive and bring their own share of heartache and fear. Rejection can be especially hard for teenagers like Amy, craving normality after years under lights.
Amy hated the treatment, hated having to sleep without a blanket, hated the flies that crawled under the glass. Most of all she hated her eyes.
When she woke up after her transplant, she begged for a mirror.
Carefully, she scanned her eyes for any trace of yellow.
"Wow," she thought. "They're so blue."
And then she thought, "I'm not a Crigler's child anymore."
Amy's uncle and aunt, John and Miriam Martin, have witnessed her trials even as they contemplate three transplants in their own family.
John is Katie Martin's brother; he has the same warm brown eyes and kind smile. His three eldest children have yellow eyes and honeyed skin.

Dawn, 12, is the eldest, a serious child with a mothering streak. Nine-year-old Eric is lanky and shy. Joyce, 8, is the mischievous one with the big imagination.

At their Mifflinburg farmhouse, Martin has built them a huge sunroom, all windows and light. He has fixed up one of Floyd Martin's inventions in the living room—a 6-foot-high box of blue lights and mirrors with a door that the children climb into after school, their heads popping out of the top. They call it "the shuttle" He has taken them on vacation to Florida, to a family with a Crigler's child who let them borrow blue lights.

But the 33-year-old father cannot escape the agony of having cursed his children with his genes.

The new baby, Joel, doesn't have Crigler's. Nor does 20- month-old Johnny. When they were born, John says, it seemed like a miracle.

Now the family prays for another miracle—a cure.

"Now I lay me down to sleep…I pray the Lord my soul to keep …"

Lying on their stomachs on their double bed, Dawn and Joyce chant their prayers in singsong unison. They are in their underwear, covered by a sheet. A heavy stainless steel canopy of lights hangs above them.

Their father kisses them good night in the dark. He cannot bear to turn on the blue lights or pull off their cover while they are still awake.

Later, he will creep back into their room and press a switch.

Outside, from far across the fields, a strange blue light will beckon in the dark.