

We will fight every inch of the way to give our two beautiful children a future

By David Cohen, Evening Standard
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Tina and Mark Harris face an unimaginable nightmare – their children are unlikely to live beyond the age of nine. Yet their courage will inspire you.

It is around midnight when Tina Harris slips silently into her super-king-sized bed and cuddles up alongside her son, Jordan, and her husband, Mark, her cheeks wet with tears. For a while, she observes their sleeping son, thinking, "You look so handsome, so peaceful, like a normal little boy. Oh why," she agonises, "couldn't you be"?

And then, holding her son tightly to feel his warmth, Tina tries to grab a few hours sleep, not knowing whether his lopsided smile will greet her in the morning. "I go to bed with the fear that when I wake, Jordan will be dead," she says. "He's six-and a-half and he's entered the zone where death could happen anytime. I just want to freeze time until they discover a cure for this wretched disease."

Two years ago, Jordan, who for the first years of his life developed as a regular little boy, was diagnosed with late-infantile Batten's disease, a rare inherited genetic disorder that affects about 500 children worldwide.

As yet, there is no treatment for this degenerative condition that gradually kills off the brain cells, causing blindness, deafness and loss of muscle control - and the prognosis is death, typically between the ages of six and nine.

As they watched their son's development halt and inexorably slide into reverse, the heartbroken couple from south-east London were overtaken by one all-consuming thought: did their gorgeous second child, Jasmine, then aged one, also have Batten's?

The doctors advised that Jasmine had a 75 per cent chance of being normal, and that they should not traumatise themselves by having her tested. For the next year, as Jordan progressively lost his ability to talk, walk or eat independently, Jasmine was, in contrast, bursting with new life, reaching every milestone ahead of time - walking at 10 months, talking at a year. They clung to the hope that their daughter would be normal.

But then Tina heard about pioneering research to find a cure for Batten's at Cornell University in New York. If the worst came to the worst and Jasmine did have Batten's, she wanted her, as well as Jordan, to have a chance for survival and to be included in the clinical trial. It was crunch time: Jasmine's blood was sent to three separate laboratories for testing. On these three test tubes hinged everything.

This is the story of how Tina, 42, and Mark, 44, a maintenance engineer working for a bank in the City, are coping with an unfolding tragedy that few of us could imagine. But the inspirational way they are doing it – refusing to give up, raising £160,000 in a critical race against time to help fund Cornell's groundbreaking research (which has already proceeded to the clinical trials of four children) - is nothing short of remarkable.

Recently scientists at Cornell have identified the specific enzyme, tripeptidyl peptidase-1 (or TPP1), whose absence is the cause of Batten's and which leads to the cells being unable to break down certain proteins, gradually killing off brain cells.

Their research has focused on two possible remedies: gene therapy, where the normal gene is implanted back into the cells; or enzyme therapy, where the missing enzyme is reintroduced. If either works, the Harris's prayers will be answered.

I meet Tina, snake-hipped, blonde and blue-eyed, on a beautiful sunny spring afternoon, pink blossoms swirling in the breeze. She is sitting in her Toyota people-carrier, parked up outside Jordan's Cherry Garden Special School in Bermondsey.

Jasmine is wriggling like a hyperactive puppy on her lap and playing, all at once, with the steering wheel, gear stick, windscreen wipers and radio.

"Mummy, mummy, look!" she says.

"You clever little girl!" Tina coos, endlessly patient, as she waits to attend Jordan's afternoon assembly and begins to tell me her story.

Tina and Mark met in their twenties while on holiday on the Greek island of Syros. From the start, they were an unlikely match: she was the daughter of a middleclass factory owner in Denmark who had enrolled for a university law degree, he the working-class son of a bricklayer from Elephant and Castle. But the holiday romance endured and the couple married in 1989, unaware that they were both carriers of Batten's and that the consequences if they started a family could be lethal for the children.

FOR 10 years, the couple travelled, partied and pursued their careers, until, in their thirties, they decided to start a family. "When Jordan was born, I thought I was the luckiest person alive," recalls Tina. "I didn't know you could love a person so much. For the first two years he was so active, so sociable, so healthy - he didn't have allergies or even suffer colds like his friends. I felt lucky my child was the healthy one."

But then, when Jordan was two and a-half and sleeping in his car seat, Tina turned around to find him in the middle of a seizure. "His arms and legs were jerking wildly, his eyes were swimming in their sockets and he was foaming at the mouth. I thought he was dying and started screaming and dialled 999."

Their GP referred them to Guy's Hospital where a specialist diagnosed Jordan with febrile convulsions, a condition not uncommon in infants. But one month later, Jordan suffered his second seizure. This time they ran a battery of tests and he was diagnosed with epilepsy. It would be another two years before the Harris's would discover the full extent of their son's illness.

Meanwhile, Jordan started falling over, and his vision and speech started to go. Not long after he turned four, he was falling with such regularity that he stopped trying to walk. Six months later, he was in a wheelchair.

"We were annoyed with the doctors because nobody could tell us what was wrong," says Tina. "I felt sure that if only they knew what it was, they could fix it."

But a skin biopsy taken in early 2003 showed that Jordan had something neither Tina nor Mark had ever heard about - Batten's disease, a genetic disorder named after the British paediatrician, Frederick Batten, who first described the condition in 1903.

For the disease to be triggered, both parents must not only carry the defective gene, but must also pass it on, theoretically only a 25 per cent risk.

Neither Tina nor Mark knew of any infant deaths in their families.

"It was just disbelief," says Tina, unable to hold back the tears. "I remember looking at Jordan and thinking, my God, we're going to lose him, he's going to die. For two weeks, I was in a daze, just crying, overwhelmed by sadness."

In the midst of their grief, and the physical exhaustion of managing Jordan's decline from a normal lad to an almost totally incapacitated boy, their hope turned to Jasmine. After holding their breath for a year, Jasmine was tested - and the Harris's were called in by their specialist.

Tina will never forget that moment.

"He looked very sad, then he said, 'This doesn't get any easier as you get older - it is Batten's'. I just fell in a heap on the floor, crying. Mark, too. I felt all my hope evaporate. I just felt so sad, so frightened. Jasmine, too! It was too much to bear."

Since early 2004, the Harris's have clung to the hope that Cornell will find a cure. Last September, they met the directors of the £4 million programme.

"They told us that their method was to drill six holes in the skull and inject the functioning gene to replace the defective one," says Tina.

They said it had worked in animal testing and that they were about to start clinical trials on humans. We got down on our knees and begged them to take our children."

So far, Cornell have given four children gene therapy. The first three are reported to be doing well, says Tina, and an MRI scan shows that it appears to have stopped, though not yet reversed, the spread of the disease. But there was a severe setback late last year when a fourth child died weeks after receiving therapy, halting the programme while further tests are carried out.

"It is early days," says Tina, "but Jordan is seventh in line to receive the treatment, and if they resume and are even able to keep Jordan the way he is, it would be an honour for me to look after him for the rest of my life." The couple have been told that Jasmine - who isn't showing symptoms yet - is still too young to receive treatment, but any breakthrough could save her the awful fate that has befallen her brother.

Later, I watch as Tina strokes Jordan's head, holds his hand and lifts him out of his wheelchair buggy into the car. Jasmine reaches out to touch him, and to pass him a toy.

"Neither of them know what they have," says Tina, who has been crying throughout most of our interview.

Back home, it is time for Tina to give Jordan physiotherapy, to massage him, and to feed him through a tube that goes directly into his stomach. "It's relentless," she says. "Sometimes I say to Mark, 'I love you, but I wish we'd never met. Anyone else and we both would have had normal children'. But then he puts his arm around me and says, 'Don't be silly, if we'd never met, we'd never have known Jordan and Jasmine'.

"Just as I am feeling the greatest despair, Jordan will smile at me. Or laugh. And then I think - I must carry on. For the children."