Our IMPOSSIBLE CHOICE

A Perth couple have been granted access to a drug that could save their sick boy—but there's a heartbreaking catch

After learning her firstborn had Hunter syndrome, "We cried for two days," says Michelle (with husband Robert and their children, from left, Abigail, Christian and Joel in Perth on Nov. 6).

For her third child, Michelle (with Christian at home on Nov. 6) used IVF and genetic screening to ensure the child would not have HS.

LOOKING FOR A CURE

There are 20–30 children with HS in Australia and their parents "are all desperate for clinical trials," says Dr Kaustuv Bhattacharya, of the Children's Hospital at Westmead. "If your child is going to die, you are desperate. There can't be anything more desperate than that."

Two weeks after the birth of their first child in 2007, Michelle and Robert Dierkx were called into the Children's Hospital at Sydney's Westmead for the results of a genetic test. Aware she was a carrier of the gene for Hunter syndrome, a rare and deadly enzyme deficiency that causes physical and cognitive damage, Michelle knew her beautiful baby boy, Christian, had a 50 per cent chance of inheriting the genetic disorder. Sitting before a grim-faced doctor in the hospital, Michelle was consumed with thoughts of the years-long nightmare she had endured in her native Jamaica, witnessing her three brothers, who had been born with HS, suffer slow, excruciating deaths.

"I was cradling Christian in my arms and I kept thinking, 'It *had* to be OK,'" recalls Michelle, who was then living with Robert in Sydney. "But I could tell from looking at the doctor's face that the news wasn't good." When the doctor confirmed Christian had HS, "I just remember screaming and going, "No, no, no!'" says Michelle, 37. "It was the most devastating news."

And the beginning of a long battle to keep their son alive. More than seven years after that heartbreaking diagnosis, Michelle and Robert, who met in Jamaica in 2001 where Robert, a civil engineer, was working, are fighting the Australian government for the right to place Christian on a US drug trial to treat the cognitive effects of HS. Currently, Christian is on the government-subsidised drug Elaprase, which treats the physical effects of HS. Should he take part in the US trial, the government will withdraw the family's access to Elaprase, leaving them to foot the $500,000 annual bill.

Without Elaprase, which is administered intravenously, Christian will suffer physically, and without the drug trial treatment, "he will have brain disease," says Newcastle-born Robert, 41, a senior manager at a WA construction company. "We're fighting for our son to have a normal life."

Sitting in the living room of their home in South Perth, Michelle and Robert, who had two children after Christian, gaze lovingly at their 7-year-old son as he plays with his beloved iPad. Siblings Joel, 6, and Abigail, 3 months, do not have HS (Joel, born premature, is deaf in one ear), but Christian does not talk, is deaf in one ear and partially deaf in the other, and suffers "slight deformity" in his face, says Robert. "This is the worst thing any parent could face—that they must watch their child decline."

While Christian underwent early intervention, including speech therapy, Michelle, a primary-school teacher, noticed that at 18 months he "was looking kind of bent over, almost like an old person," she says. They sought help and Christian became the youngest child in Australia to begin the Life Saving Drugs Programme, which involved Elaprase, one of the world's most expensive medications. After the treatment began, "he was doing really well," says Robert. But "mentally, he was delayed. We knew this treatment wasn't improving his brain."

During painstaking research, Michelle discovered US biopharmaceutical giant Shire was conducting a trial for another treatment (Intrathecal Idursulfase-IT), which targets the cognitive affects of HS. Christian qualifies for the trial, but the government dropped a bombshell: if Christian took part, access to his current medication would cease because, say the guide-lines, "The child cannot receive the subsidised treatment while they participate in a trial."

Christian, who has Hunter syndrome, with brother Joel. "Joel says, 'Mummy, I don't want Christian to die,'" says Michelle.

The couple refuse to make a choice; they want access to both treatments. "We have spoken to other parents whose children were on the trial, and they are living a normal life," says Michelle. So, says Robert, "We're now fighting for that for our son."

> "It was the most devastating news"
> —Michelle Dierkx

So far, the government has stood firm. The couple have reached out to federal health minister Peter Dutton, who "just regurgitated the guideline," says Robert. Says their local federal MP, Steve Irons: "I am working to achieve a mutually beneficial solution for all parties."

Meanwhile, the couple are in an exasperating state of limbo. "We've gone from a child with no hope two weeks after birth to now thinking, 'You know what, we can have almost a normal child.' That's what's at stake here." Says Michelle: "All I want to do is give my baby the best opportunity in life. Please give him a fair go. Let him live."

■ *By Melenie Ambrose and Michael Crooks*
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MAKEUP: STEPHANIE LAW; HAIR: DOREEN CHATFIELD; INSET: COURTESY OF DIERKX FAMILY

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