

# Sunday Insight

## All she wants for Christmas is a smile

While most of the world's children can crack a grin without thinking twice, one young Kiwi would give anything to be able to smile.

**Abby Gillies** reports.

**L**ITTLE MIA CRONJE has never smiled. And unless, by a miracle, treatment is found for her rare genetic disorder, she is unlikely to without a risky operation.

The Waiuku 4-year-old was born with the rare neurological disorder, Moebius syndrome, which causes facial paralysis. As a result, Mia's facial expression is blank. She cannot smile, frown, grimace or break into a laugh. Instead she must find other ways to express her feelings and in her short life, she has done just that.

Mia's mother Lielze Cronje, 40, a teacher at nearby Pukekohe Hill School, says her daughter "smiles with her eyes".

"She jumps up and down when she's happy or she'll say, 'I'm happy,' and pull her face up."

And if people ask Mia if she is happy, Lielze tells her to say "yes, I smile on the inside".

There is a chance that Mia could gain some facial expression if Lielze and her husband Mark, 40, agree to surgery. But Mark, who runs his own business installing security systems, is worried about the risks of the two 12-hour operations, performed three months apart, during which nerve and muscle from the upper leg would be transferred to the corners of the mouth.

Lielze is in favour of Mia having the \$5000-plus smile operation, which will also improve mouth movement, making speech easier. Afterwards Mia would have to learn to use the new muscles to smile at the right time.

"They have to think, 'I'm happy, I need to smile'," Lielze says.

But Mark is worried that the surgery might further deform his daughter's face. "I don't know if it's a good thing or not. Of course it would be good for her to be able to smile but I'm a bit worried that if she does have the operation it might change the look of her face."

For now Mia and her parents are on a waiting list to see an Auckland surgeon to discuss the operation and ongoing new developments which

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**SMILING ON THE INSIDE:** Mia Cronje has a rare genetic disorder which makes it impossible for her to smile. Her mother Lielzel hopes an operation may help.



JASON DORDAY

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may improve Mia's condition.

To see her child smile "would be huge for any parent", Lielzel says. But whether or not to have the operation will be Mia's decision to make in the future, say her parents.

Surgeon Jeremy Simcock, who performs the smile operation at Christchurch Hospital, says the operation is most successful on young children because of their ability to heal quickly and learn to use the muscles in a new way. "The younger the better, within reason," he says.

**E**VEN BEFORE Mia was born at Middlemore Hospital in September 2006, Lielzel knew something was different about her second baby. She had heavy bleeds at eight weeks — the time that the cranial nerves are formed in the foetus. "From that day on I thought I wasn't sure if everything was fine. I had this feeling. I prepared myself."

When Mia was born under weight and cross-eyed, the neonatal doctor at first thought she had Down syndrome. A paediatrician later confirmed a diagnosis of Moebius and Mia spent her first five weeks in the neonatal unit to monitor her feeding and growth.

There were other differences. Her cries were unusually soft and because of the facial paralysis she could not close her lips around the bottle, making sucking difficult. To solve the problem, Lielzel and Mark used a specialised feeder that allowed them to control the flow of milk.

As she grew, the disorder caused Mia, born with club feet, problems with balance. Because she cannot blink or squint, her eyes are sensitive to light. To protect them, she rolls her eyes back in her head when necessary. She has also had difficulty swallowing and struggles with her speech.

Getting support for Mia has been a frustrating process, Mark says. It was difficult to get Mia the care she needed as a baby, and the physiotherapists who work with her are constantly changing. "It's very hard to have to fight for everything. It's an ongoing struggle."

The lack of information and support, compared to more well-known conditions, has also been a

## Creating a smile

Surgeons remove a muscle, along with its blood and nerve supply, from the inner thigh.

The muscle is then joined to the nerves and muscles in the face: the muscle to the cheekbone and corner of mouth, and the nerve to the chewing muscle.



Source: Dr Jeremy Simcock

challenge, he says.

"If you compare [Moebius] with Down syndrome, for example, they have a lot more things available that we don't have available to us. We don't enjoy the same sort of things from a funding perspective."

This year Mia has had three operations — two on her knees after her legs started to bow out and a third to have a plate removed after a hip operation last year. She will need further operations on her legs and feet. But her parents say Mia is resilient and plucky.

In the backyard of the family's home, Mia plays with her sister Daneka, 10, like any 4-year-old — energetic, fearless, noisy, happy.

She is looking forward to starting school and can't wait until Christmas. Her wish list this year includes ballerina clothes, and princess and mermaid dolls.

But when Mia excitedly unwraps her presents under the Christmas tree, the neutral expression on her pretty face will not change. That's the most heart-breaking part for Lielzel and Mark — watching Mia struggle to communicate, impaired by speech difficulties and the lack of expression in her face.

The challenges for Mia and the Cronjes are ongoing as the family continues to learn how the syndrome

will affect her as she grows.

Mark says the emotional toll has been high.

"For example, seeing other parents with the same age kids and some of the things she can't do that they can. It's emotionally draining and we don't know what's going to be next."

To cope, the family takes each day at a time and they often head to the

**"There's nothing they can't do except smile on the outside."**

Vicki McCarrell

beach or take long walks to relax.

"We just hope she'll have a normal life and treat her like a normal child."

Suzanne Woon, a disabilities services co-ordinator from Hastings, has lived with Moebius syndrome for 41 years.

Woon was diagnosed a few days after being born with club feet and problems feeding. She still has difficulty saying some letters but has learned to manage her speech.

Four years ago she underwent the smile operation, but sadly she did not get the results she hoped for.

"I ended up with a dimple but no

smile, which is disappointing."

The operation wasn't available when Woon was younger and she believes those who have it early have a greater chance of success.

Like Mia, Woon has created her own ways of communicating her feelings, using gestures, laughing or smiling with her eyes.

Her teen years were difficult as she was often bullied for being different. Having a supportive family helped and her advice to others with the syndrome is to accept themselves and ignore those who don't.

"You have to be able to tell yourself 'I'm not the one with the problem, it's them,'" she says.

While there is no official support group in New Zealand — Mia and Woon are two of six known cases of Moebius syndrome in the country — the Cronjes have received support and information from the Moebius Foundation in the United States. The foundation's 1600 members are worldwide and include teachers, doctors, librarians, lawyers, professors and a CNN producer.

The lack of a local support group when a condition is so rare can make it difficult for families to cope, says John Forman, executive director of the New Zealand Organisation of Rare Disorders. "They are on their own in the community with no

source of information other than their own research on the internet. The isolation can be a really, really tough thing for people."

His organisation can help families connect with a Moebius syndrome support group in Australia, he says. "We can help them get some basic resources like a website with information and contact details."

The Cronjes already have plans to connect with other families by setting up a New Zealand support group next year.

Most of all they want to help new parents of children with the condition — "to let them know that life does go on", Lielzel says.

Next month the Cronjes will mark the first Moebius Awareness Day, held in the US by making bracelets and T-shirts saying "smiling from within".

In the meantime they hang on to the hope of new research into the cause and treatment of the syndrome. US-based Moebius Syndrome Foundation president Vicki McCarrell says the research is encouraging.

A new method that treats club feet by setting them in casts has been developed and researchers at the University of California are looking at artificial eyelids that blink.

Major progress has been made in developments in speech therapy, McCarrell says.

"There's nothing they can't do except smile on the outside."

While they wait for a decision about Mia's smile operation, the Cronjes are concentrating on developing their daughter's speech and fine motor skills.

She is undergoing intensive physiotherapy to increase mobility after her recent operations. Regular speech therapy in the past year has meant Mia has progressed from saying single words to full sentences.

As she runs around the house playing or into the kitchen to get a snack, Mia often falls because of problems with her balance.

But, says Lielzel, her daughter finds her own way to cope and rarely complains. It's that resilience that makes the Cronjes most proud.

"She's a gutsy little girl," Lielzel says. "Children will take a situation and carry on. Her strength is what I find amazing."