

Danilo and Mary Giribaldi were still immersed in the joy of welcoming their third baby when Bernadette, their second child, suddenly complained of feeling sick. The little girl with soft brown eyes and a cheeky smile was a couple of months shy of her third birthday and had just spent a busy day with her grandparents.

When Danilo, a civil engineer, arrived to take her back home to Castle Hill in Sydney's northwest, she was burning up. He noticed a strange new patch like a bruise on her temple. "Did you bang your head?" he asked. She nodded, but it was a sign of something far more serious than a backyard scrape.

While Mary was busy with their other two children, Danilo put Bernadette to bed. Several times throughout the night he checked on the restless toddler. He gave her paracetamol and then ibuprofen, but her temperature still raged. At 5am, Danilo checked her again and this time noticed that Bernadette had developed thousands of tiny red and purple dots across her trunk.

He called out to Mary. Even blurry-eyed, she recognised the rash immediately: she'd seen the distinctive markings of meningococcal disease on a TV show years before. So while Mary stayed behind to watch the other two children, Danilo bundled Bernadette

in the car and set off for the nearby Children's Hospital at Westmead. He thought they'd give her some medicine and he'd be back home in an hour or so. But within moments of walking through the doors of the emergency department, everything changed.

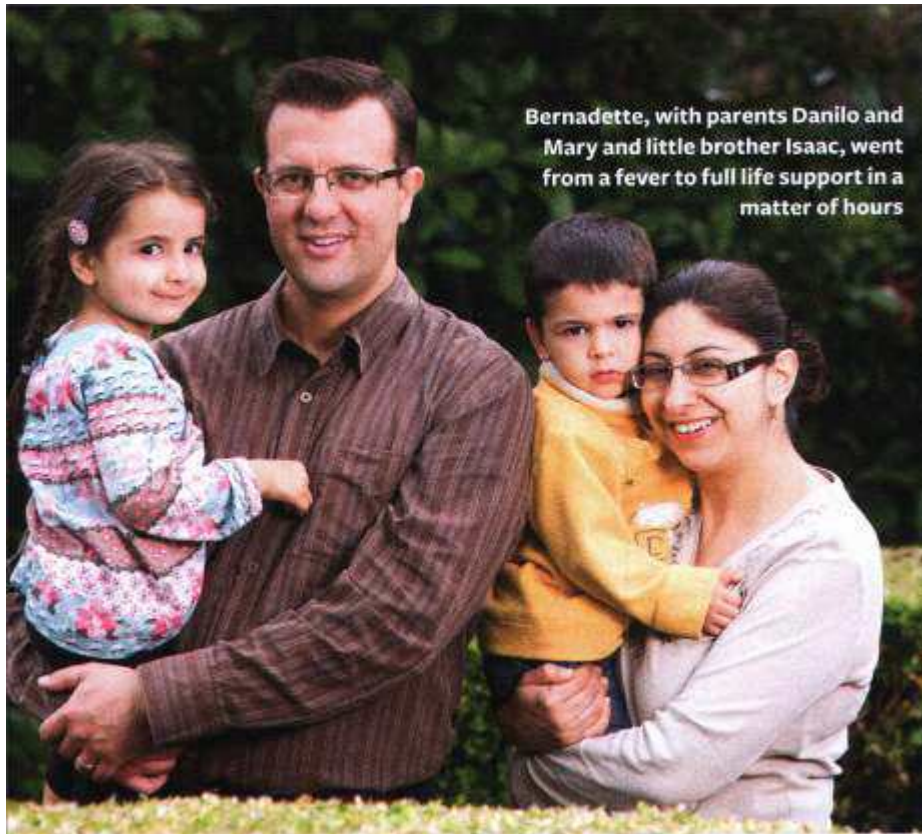
Meningococcal B is caused by the bacteria *Neisseria meningitidis*. Many of us carry this bug in the back of our throats or noses without any problems. An estimated one in four teens, for instance, are carriers at any one time.

However, on rare occasions the bacteria can enter the bloodstream and overwhelm the body's defences. If this happens, one in 20 victims dies, yet doctors aren't sure exactly what it takes for a body to succumb. "You're weakened by a viral infection," suggests Professor Robert Booy of the National Centre for Immunisation Research and Surveillance. "You might have been exerting yourself a bit more. Maybe your immune system is subtly different to everyone else's."

Once meningococcal B enters the blood, it multiplies rapidly and releases toxins, causing a range of life-threatening conditions including septicaemia (blood poisoning) or meningitis. The red or purple spots signal leaking blood vessels. To fight the bug, the body

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automatically recalls blood from the extremities to preserve vital organs. With no blood supply, irreversible damage to arms and legs occurs within hours. As flesh dies, gangrene sets in.

It was barely 6am when Danilo rushed his little girl into emergency. In minutes, a medical team had gathered around Bernadette to figure out what to do first. Intravenous lines were set up in each arm to infuse antibiotics. But as they watched, blotches spread all over her body, and “tears” of blood started leaking from her eyes.

By 10am, after Bernadette had been transferred to the intensive care unit,

her blood pressure had dropped to dangerous levels, and she was placed into an induced coma to allow her body to concentrate every last drop of energy on survival.

Bernadette’s blood pressure didn’t rally, and despite a raft of drugs for boosting heart rate and restricting blood vessels, her heart was in danger of stopping. Her kidneys had failed and she needed to be connected to a dialysis machine to filter her blood of wastes. Amid these dangers, her little body had started to swell. Twice during the day, the team cut into her limbs to ease the swelling and attempt to save her arms and legs.

What you need to know

Meningococcal disease is most common in children under five and in young adults aged 15 to 24. Symptoms include fever, pains in the legs and a rash of red-purple pinpoint spots or larger bruises. If meningitis develops, there may be vomiting, aversion to light, drowsiness, headache and a stiff neck. Simple checks include:
Does normal medication bring the fever down? If not, seek help.
Hold a glass against the rash and press firmly. If you can still see the spots despite the pressure, this could be meningococcal.
If in doubt, seek medical attention - swift treatment is critical.

By 7pm Bernadette was in septic shock, so she was moved into theatre. Her chest was opened up and her heart moved to one side to make room for tubes connected to an ECMO (extra-corporeal membrane oxygenation) machine - an external pump that would artificially circulate and oxygenate her blood. When Bernadette was wheeled back to the ICU, where her still stunned parents were waiting, she was surrounded by machines, all needed to keep her alive.

Mary and Danilo could barely contemplate how their little girl, who just 16 hours previously had been eagerly looking forward to being a flower girl at her aunt's wedding, was now on full life support.

That night, Bernadette's kidneys

failed. She was connected to a dialysis machine to filter her blood of wastes, extra salt and water. Her future looked bleak, but with her parents by her side the little girl fought on.

The ICU team began days of around-the-clock monitoring. They reopened Bernadette's chest to clean blood leakage from tubing connected to the ECMO pump and kept up a supply of fresh blood for transfusion. Two weeks in, with her condition stable, doctors decided the dangers of taking her off the ECMO pump were probably lower than the risks of infection if she stayed connected to it.

Still she fought.

At the three-week mark, the results of insufficient blood supply to the extremities were obvious. Gangrene had crept up Bernadette's foot and ankle. When the doctors couldn't feel a pulse below her knees, plastic surgeon Dr Peter Haywood explained to Danilo and Mary that they were at a critical point: to keep the limbs would be life threatening, as infection from the dead tissue would inevitably spread. There was no choice but to amputate. Doctors removed both legs below the knee, most of the fingers on the left hand, and parts of the fingers on the right.

A week later, Bernadette began to rouse. Distressed and uncomfortable, it wasn't clear whether she'd sustained brain damage (she hadn't), or whether she would have permanent kidney problems (she did).

After five weeks in intensive care, Bernadette was stable enough to be

transferred to the ward. Finally, in mid-June, after turning three in hospital, she was released. "It was like bringing home a newborn," says Mary. For months she needed to be fed via a nasogastric tube. Then as she gathered more strength and her limbs healed sufficiently, she could be fitted with her first artificial legs.

As her baby brother Isaac learned to crawl, Bernadette learnt to walk all over again. "At first all she could do was stand," says Danilo. "Then she'd take one or two steps, then she could walk with a frame. When we finally saw her after a few months walk with her legs all by herself, it was the greatest feeling any parent could have." ■

Getting on with living

When Bernadette learnt she'd lost her legs, she said to her parents brightly, "It's OK, we can go to the shop and get some new ones."

That's basically what Mary and Danilo did. Now, along with piles of toys and clothes, they're left lying in a heap in a corner of the playroom, pink shoes still attached to the feet.

Bernadette's too young to fully appreciate what has happened. Yet she's coping. And how. In just a few months she's learned to walk again. She draws and colours like any other toddler, the stumps of her fingers expertly curling around the pencil. And today she's giggling as she snuggles up to Marty Mayberry, a trainee doctor from The Children's Hospital at Westmead.

Marty is perhaps one of the few medical staff who can truly empathise with

her: a decade ago he, too, was struck down by meningococcal B.

These days the fit 25-year-old strides around the wards, but beneath his trousers are two prosthetic legs. He pulls off his socks to show them to Bernadette: "See, this is how this works, this is how the ankle bends."

Refusing to allow the disease to limit him, Marty is a Paralympic skier with a bright future in medicine. In fact, it was his brush with the deadly disease at 16 that convinced him to become a doctor. Now he's helping with research at Westmead into the social effects of meningococcal in the community.

While Bernadette and Marty share an incredible optimism, doctors are also very optimistic that they can stop the disease in its tracks. A vaccine that

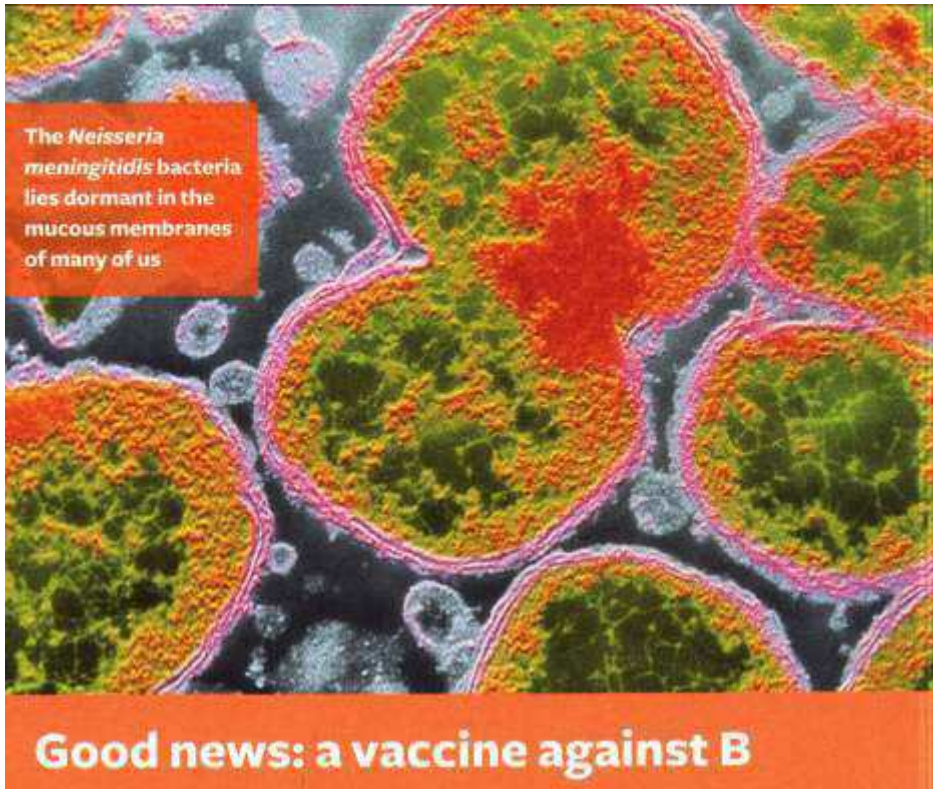
works to prevent the B strain has now been developed and children around the world could start receiving their vaccinations this year.

With three other kids to care for (the Giribaldis have since had a fourth) Mary and Danilo juggle appointments with doctors and rehab specialists. Bernadette's prosthetic legs, which cost about \$15,000 each, need upgrading as she grows; the house needs modifying to boost her self-sufficiency; and, of course, there's been the psychological strain.

"Immunisation is the only way to prevent all this," says Danilo.

For more information, visit meningococcal-australia.org.au.

For more about Bernadette, email howisbernadette@gmail.org



There are a good line of meningococcal bacteria, with B and C the most common. Vaccination against the C strain, introduced to Australia in 2002, has reduced the number of C cases by more than 90%.

Today, the vast majority of cases are caused by the B strain – about 5% of these prove fatal – but until now there's been no effective vaccine against the disease.

The problem for researchers lay in the great diversity of meningococcal B strains around the world – and previous vaccines have only prevented a small percentage of cases.

There was another problem, too. The outer coating of the meningococcal bug has exactly the same chemical structure that's found in the brain of newborn babies. Develop a vaccine to create

antibodies to this structure and you risk having the body attack itself via an auto-immune response. In a medical breakthrough though, international researchers have now produced a vaccine that breaks through the troublesome coating of the bug and targets the proteins just below the surface.

In late 2010, Swiss-based pharmaceutical company Novartis submitted a vaccine for regulatory review in Europe, and it's expected that it may be licensed later this year. In Australia, too, an application has been lodged with the Therapeutic Goods Administration and is under review.

The Australian Government has yet to consider whether it will fund universal vaccination with the new vaccine.