

Complex Surgery Saves Young Twin

A minuscule hole between a toddler's eyes was a direct line for bugs to her brain. But a complicated surgical procedure has given her a much safer passage through childhood. Kelly Andrew reports.

NAYYEREH and her sister Sohaila are 18-month-old identical twins, but despite their matching dimples and curly hair, Nayyereh looks a little different.

Her nose is broader across the bridge with a small scar and some swelling, and she has surgical tape on her neck, a reminder of a serious operation performed two weeks ago. Hidden under her hair is a neatly stitched scar from ear to ear.

Holding tightly to a well-loved blue toy rabbit, "Rabba", that has kept her company during stays in Hutt and Wellington hospitals over the past few weeks, Nayyereh is closely watched by her relieved parents Suzannah and Siavash Jandeh-Razmi.

She had surgery at Wellington Hospital to remove a mid-line nasal dermoid. If left untreated, this rare condition could have caused a life-threatening infection in her brain.

It affects only two or three children a year in New Zealand, and occurs because of a glitch in the first four weeks of development in the womb.

In the developing embryo, the brain is immediately under the tissue

that eventually becomes the nose. A nasal dermoid forms when the brain moves backwards in the skull and leaves a tunnel, or tract, of skin.

In Nayyereh's case, this tunnel was connected to the outer layers of her brain, and exposed to the outside world through a tiny indentation or pit at the bridge of her nose.

This one-millimetre opening, which had hairs growing from it, allowed bacteria to get inside, and potentially travel to the brain. The dermoid also pushed apart the nasal bones, broadening her nose.

It was when the pit began to swell a few weeks ago that her parents started to worry. The family's GP prescribed antibiotics for a suspected sinus infection. But the pills did not help and the swelling got worse. The GP referred the Jandeh-Razmis to Hutt Hospital's paediatrics department.

Nayyereh was hospitalised for a few days and then discharged, but a CAT scan was ordered of her brain. Initially her parents were told the scan showed everything was fine, and they were sent home.

But later, after the scan was checked by plastic surgeon Charles Davis, the seriousness of the condition became clear.

"Alarm bells started to go off," Siavash says.

Nayyereh was put on intravenous antibiotics to control the abscess which had formed around the pit between her eyes, and she spent nearly two weeks in Hutt Hospital.

Mr Davis recommended surgery, but it had to wait till the abscess had reduced in size. Operations were performed to drain fluid from it and release pressure. Siavash was stunned by how quickly something that seemed simple became complicated and life-threatening.

"For about three weeks we thought 'Oh God, are we going to lose her?' It was difficult going through all this with her but all the medical people we were dealing with were fantastic."

Siavash and Suzannah took it in turns staying overnight at Hutt Hospital, with one of them looking after their other two children, Sohaila and two-year-old Kurosh, at their Upper Hutt home.

Friends and family gave their support, contributing meals to their fridge and offering to babysit.

As a worried, protective father, Siavash grilled Mr Davis in detail about the planned operation. "I wouldn't trust just anybody to put a knife in my kid. I asked some rough questions but he's so professional about it. He explained as much as possible about what it was."

"At the end I just realised that I had 200 per cent trust in him."

When he realised there was a 95 per cent chance of his daughter dying without the surgery, he decided it was the only option.

The couple spent a nervous five hours waiting for news of their daughter during the procedure.

It involved making an incision from ear to ear so her scalp could be folded forward and part of her skull and nasal bones removed to expose

► REMOVING A DERMOID FROM THE BRAIN

The five-hour surgical procedure performed by Wellington plastic surgeon Charles Davis and neurosurgeon Agadha Wickremesekera to remove a mid-line nasal dermoid – an abnormal tunnel of tissue between the nose and the brain.

1 An incision is made from ear to ear



across Nayyereh's scalp to limit scarring around the nose, peeling forward a flap of skin to expose her skull and the top of her nasal passage.

2 A piece of frontal bone and the nasal bones are removed to give surgeons access to the dermoid attached to brain.



3 Dermoid separated from brain and front of skull replaced. Nasal bones narrowed to normal width.



Dermoid exiting through opening in nasal bones which have been played apart.



4 The frontal bone is placed back into position.

5 The wound is closed. A small scar on the nose remains where the pit has been excised.



Source: CHARLES DAVIS Graphic: RICHARD PARKER



Rabbit run: Nayyereh Jandeh-Razmi revels in her freedom with parents Siavash and Suzannah after a harrowing stay in hospital.
Picture: MAARTEN HOLL

the dermoid. Working in partnership with neurosurgeon Agatha Wickremesekera, Mr Davis carefully removed the tunnel of skin between Nayyereh's nose and brain and disconnected it from the outer layers, or dura, of her brain. The section of skull was replaced, and her nasal bones narrowed to normal width before her scalp was sewn closed.

Despite having confidence in the surgeons, Nayyereh's parents still feared the worst.

"It was the most harrowing, awful day of our lives," Suzannah says.

were told the operation had gone well.

After the toddler woke up the first thing she wanted to eat was strawberries. "That's when I knew she was getting better," Suzannah says. "And because she threw her toy rabbit out of the cot."

Both parents say the experience has changed their outlook on life and their family.

"Each day that a parent wakes up and sees their children happy and healthy they should be thankful," Siavash says. "When you are in hospital you see other people's misery.

noticed it but didn't think anything of it, and our doctor said we could do something when she got a bit older. If we had known we would've acted much, much sooner."

Mr Davis says though nasal dermoids are rare, their complications can be life-threatening and doctors need to be aware of the warning signs.

"It's very important for all doctors to remember that any mid-line swelling of the nose could potentially be connected to the brain because during development the brain is directly underneath there."

Nasal dermoids are not genetic and are "just one of those random events that can occur", he says.

He says Nayyereh was lucky the infection did not reach her brain, potentially causing meningitis.

"Most of these are picked up before it gets to the stage of an abscess. Hopefully, her case will raise the awareness of it."

"With any swelling from the base of the nose from the eyes down to the tip of the nose in a child you have to investigate the possibility that it might be connected to the brain before you do anything about it."

Relaxing at home just weeks after their daughter's operation, Siavash and Suzannah are still recovering from the traumatic experience.

Suzannah says: "We want to say thank-you to all the staff and specialists for giving us the best Christmas present ever."

"If parents see a little pit like a pore with a hair growing out of it on their child they should check with their GP. If we had known we would've acted much, much sooner."

"Taking her down there and having to let her go and not knowing what was going to happen — were we going to have a child afterwards and if we did, was she going to walk and talk and be the child that we had."

"The wait was incredibly long, we were reading old *National Geographic* magazines."

"They said if there were any complications they would phone through, so every time the phone rang I would feel tense."

The relief was overwhelming when they saw Nayyereh afterwards and

You never realise there are so many needy people in New Zealand when you are sitting in the comfort of your own home."

Suzannah agrees. "In hospital we met children who aren't here today. We feel incredibly lucky that we've got Nayyereh and incredibly grateful to Mr Davis and the others involved in her care."

She wants other parents to be aware of the condition. "If parents see a little pit like a pore with a hair growing out of it on their child they should check with their GP. We had

BRIEFLY

Vitamins heal

■ A vitamin A compound commonly used to treat acne improves the healing of foot ulcers in patients with diabetes. The study, of 24 patients, reported in the *Archives of Dermatology*, found that topical Retin-A (tretinoin) helped wound-healing.

Breath of air

■ High doses of an inexpensive drug may help people desperately short of breath due to idiopathic pulmonary fibrosis, an incurable illness that causes a hardening of lung tissue, a study has shown. But the drug, acetylcysteine — which is available without prescription — may simply be neutralising the side effects of standard therapies, according to an editorial in the *New England Journal of Medicine*, which published the study.

In the Belgian study, volunteers who took 600 milligrams of acetylcysteine three times a day on top of receiving standard care could exhale 9 per cent more air after a year than patients who received standard care alone.