

When the Pain of Words Bites deep

Eight-year-old Jack has a genetic condition called Opitz syndrome, characterised by widely spaced eyes and a broad nose. So hurtful was the taunting Jack got at school about his appearance that his parents resolved that he should have plastic surgery to correct his facial features — and save his youthful confidence.
By Kelly Andrew.

Rare surgery gives boy fresh chance

CHILDREN can be devastatingly cruel to a boy who looks different. South Canterbury couple Karen and Brian were horrified when they discovered their eight-year-old son Jack had been subjected to name-calling and teasing by older children at his school.

Teachers were unaware he had become a target, and Karen found out what was happening only when she was told by the mother of one of Jack's friends. "She said she thought it was terrible that all the kids were calling him names. I had no idea. It's the older kids, the ones that don't know him so well."

Jack has a rare genetic condition called Opitz syndrome which affects only about 100 people worldwide. A range of physical abnormalities are associated with the syndrome, but it is Jack's unusual facial features that have attracted unwanted attention. His wide-spaced eyes — a condition called hypertelorism — and extremely broad nose drew stares and, more harshly, name-calling such as "fish face".

Karen says Jack is too young to understand exactly why he has been singled out. "He doesn't realise that kids are teasing him because of the way he looks, he just thinks they're being mean. We don't think he realises that he looks a bit different."

Realising that their son's confidence and enthusiasm for school could be irreparably damaged by the prejudice he encountered because of his appearance, Karen and Brian made the difficult decision that he should have surgery that would help him fit in.

They flew from Timaru to Wellington two weeks ago and plastic surgeon Charles Davis performed an operation at Hutt Hospital that has moved Jack's eye sockets closer together and narrowed his nose into a more normal shape.

Until recently, the couple had hoped the operation would not be necessary or that it was a choice they could allow Jack to make for himself.

"Originally, we were going to leave it to him to make the decision," Karen says. "But in the last year, things had changed at school and with people's reactions. He used to be the kind of kid who would go and play with anyone in

the playground. But in the Christmas holidays, he wouldn't go near the other kids. If he lost his confidence through his appearance, I'm sure that would affect everything. He wouldn't want to go to school any more."

Brian says a family friend who is a teacher warned them Jack would be "taken down" at high school because of the way he looked. It is the reaction of strangers that they are most worried about, rather than the people who know his bright, cheeky personality.

"People who know him are used to him and they don't see him for what he looks like but for what he is."

Brian feared that if they did not act and Jack was unhappy later in life, he would come back to them and ask why they hadn't done anything to help him.

Now, just days after the operation, Jack is sitting in a motel room with his parents, talking about how much he is looking forward to going home and playing with the new Lego he has been given.

His face is bruised and swollen around the eyes and he has a small plaster cast across his nose held in place by bandages.

But his parents are hugely relieved that he is all right. Both they and Jack were anxious in the weeks leading up to the surgery. Karen says he was not sleeping properly and had started playing up at school.

They had to explain to him what was going to happen during the operation to ease his concerns, though they tried to play down the more frightening details of the procedure. "He thought they were going to take his nose off and his eyes would bleed. There were a lot of things that went through his head that we hadn't thought of. He kept saying that people would think he was scary," Karen says.

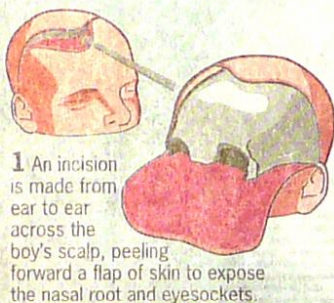
But the surgery went smoothly and it is hoped Jack will have fully recovered in about six to eight weeks.

THE five-hour operation involved an incision across the skull that is hidden by hair and avoids scarring around the eyes. The face is peeled forwards to expose the eye sockets underneath.

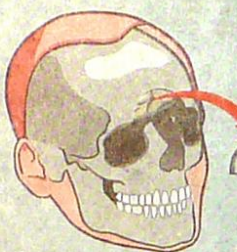
The central part of the nasal bone is

► FACING THE FUTURE

A five-hour surgical procedure by plastic surgeon Charles Davis moved a boy's eye sockets closer together.



1 An incision is made from ear to ear across the boy's scalp, peeling forward a flap of skin to expose the nasal root and eyesockets.



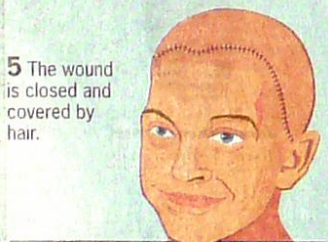
2 A piece of bone on the forehead and front of nose is removed. Sinuses in front of brain and between eyesockets are removed.



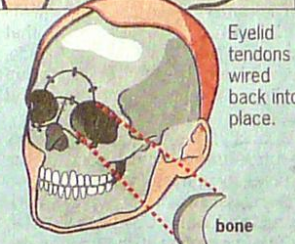
The eight-year-old boy, born with Opitz syndrome, has an extensive wide space between the eyes and a very broad nose.

Frontal bone
Central nasal bone
(18mm wide)

3 Eyesockets are moved 2cm closer and nasal bones narrowed to normal width and wired.



5 The wound is closed and covered by hair.



Eyelid tendons wired back into place.

4 The frontal bone is plated back into position. A piece of bone from the hip is fashioned to the shape of the eyesocket.

Source: CHARLES DAVIS Graphic: RICHARD PARKER

'People who know him are used to him and they don't see him for what he looks like but for what he is.'

It is not the first time Jack has been in hospital. He had surgery as a child to correct craniosynostosis, where the plates of the skull are fused together, and problems with his urethra. Both are related to Opitz syndrome.

KAREN says they feel lucky that the recent operation went ahead, considering recent changes to waiting lists. The surgery date had been postponed till August but at the last minute, was moved forward. Strangely, Brian says, Jack became quite excited in the days before he went into hospital after the family flew up to Wellington. "Even on the morning of the operation he was quite chatty going in."

Now the family is relieved that the worst is over.

"It's just the anticipation of the whole thing and torturing yourself about whether you've made the right decision to have it done. But I think we've done the right thing, definitely," Karen says.

However, it has been a traumatic process. The couple were awake for two days when they stayed with Jack in hospital after the operation, dozing in armchairs in the intensive care unit.

And after Jack woke up, he asked his parents why they had "made him" go through the operation.

"We just said that he had too much bone between his eyes, and so 'you'll look like dad'," Karen says.

Their experiences have made them more aware of the way society judges people based on their appearance.

Karen says parents need to be careful what they say in front of their children, "because kids will pick up on it. They've got to get these ideas that kids are different from somewhere."

Brian says having Jack has made him more tolerant and understanding. "People do see skin deep till they get to know people. We all do it."

"It's not till it affects you that you realise how cruel society is, and how much emphasis is put on appearance."

Names have been changed by request to protect the family's privacy.

removed and the brain is lifted up before air sinuses, which are not needed, are taken out.

The remaining two sections of bone in the nose are joined together with wire, making them a centimetre closer on each side.

Normally the nose is 1.5cm wide at the bridge but in Jack's case, it was 4cm across because of extra tissue that had formed between his eyes. Opitz syndrome affects the normal development of the eyes in the womb, preventing them moving from the side of the head to the front.

Only the inner half of Jack's eye sockets were moved closer together, making the eye wider.

A very thin piece of bone removed from Jack's hip was moulded into shape and used to create a new roof and floor of the sockets.

Eyelid tendons were wired together

behind the nose and a new tear duct drainage hole was created to allow tear ducts to drain into the nose.

Bone around his nose was sculpted into shape, but extra skin was left where tissue has been removed. The cast was put on to contract the excess skin, and his skull was stapled back together — a technique which allows for better hair regrowth than stitching.

Mr Davis had seen only one previous case of Opitz syndrome, in Australia, but he performs several operations a year to correct hypertelorism.

Most children affected by the condition have normal intelligence but are often treated as being intellectually impaired because of their facial appearance, he says. Jack was a suitable age for the surgery because his adult teeth had moved down into his jaw. Younger children still have their teeth behind their eye socket.

Mr Davis is pleased with the result of the operation and believes it will markedly improve Jack's quality of life. "I think he will look significantly improved. I think we've made a tremendous difference to his facial appearance. I don't think you'd notice him if you saw him on the street, which is the main test of whether this type of surgery has been successful or not."

One consequence of the procedure can be double vision, but he says there is no sign of this problem.

Jack's parents were worried that after the operation, he would not look like himself any more.

"He does look different, but he still looks like him. We were a bit scared that he was going to look totally different but he still looks like Jack, though it's hard to tell with all the swelling still there," Karen says.