

TRIAL HOPES | Researchers work on 'Flightless' protein

Skin disease therapy gets its wings

IMAGINE having skin as delicate as a butterfly's wings.

Blisters and wounds form at the slightest touch. Bathing and dressing is a three-hour ordeal.

Shoes, seams and even tags on clothing irritate the skin to the point of scarring.

Kate Turner, 21, of Victor Harbor, is one of 1000 Australians with the rare condition called epidermolysis bullosa, or EB for short.

She wants to raise awareness and promote research, even though she doesn't like to think too much about a cure.

"I just live my life, doing what I do every day," she said.

"I don't want to get too hopeful, just in case."

Scientists in the Wound Healing Laboratory at the Women's and Children's Health Research Institute are conducting research into Kate's condition with the aim of "working towards stronger skin".

The team, led by Associate Professor Allison Cowin, are studying a protein called "Flightless", so named as it was first identified in fruit flies.

The protein is essential to normal development but the team found it plays a role in wound healing. It turns out patients with skin blisters have higher levels of the protein.

Unfortunately, it does more harm than good.

"When we can get rid of it, wounds repair much better," Associate Professor Cowin said.

Antibodies can be used to mop up and neutralise excess protein, so there is potential for a new therapy. If all goes to plan, clinical trials will begin within the next five years.

But first, the scientists have to show their approach works in animals, specifically with mice and pigs, as pig skin is the most similar to humans.

University of Adelaide PhD student Zlatko Kopecki says preliminary results are promising.

"Reducing Flightless levels improves wound healing," he said. "We see a smaller wound area and increased cellular mi-



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gration and proliferation to close the wounds."

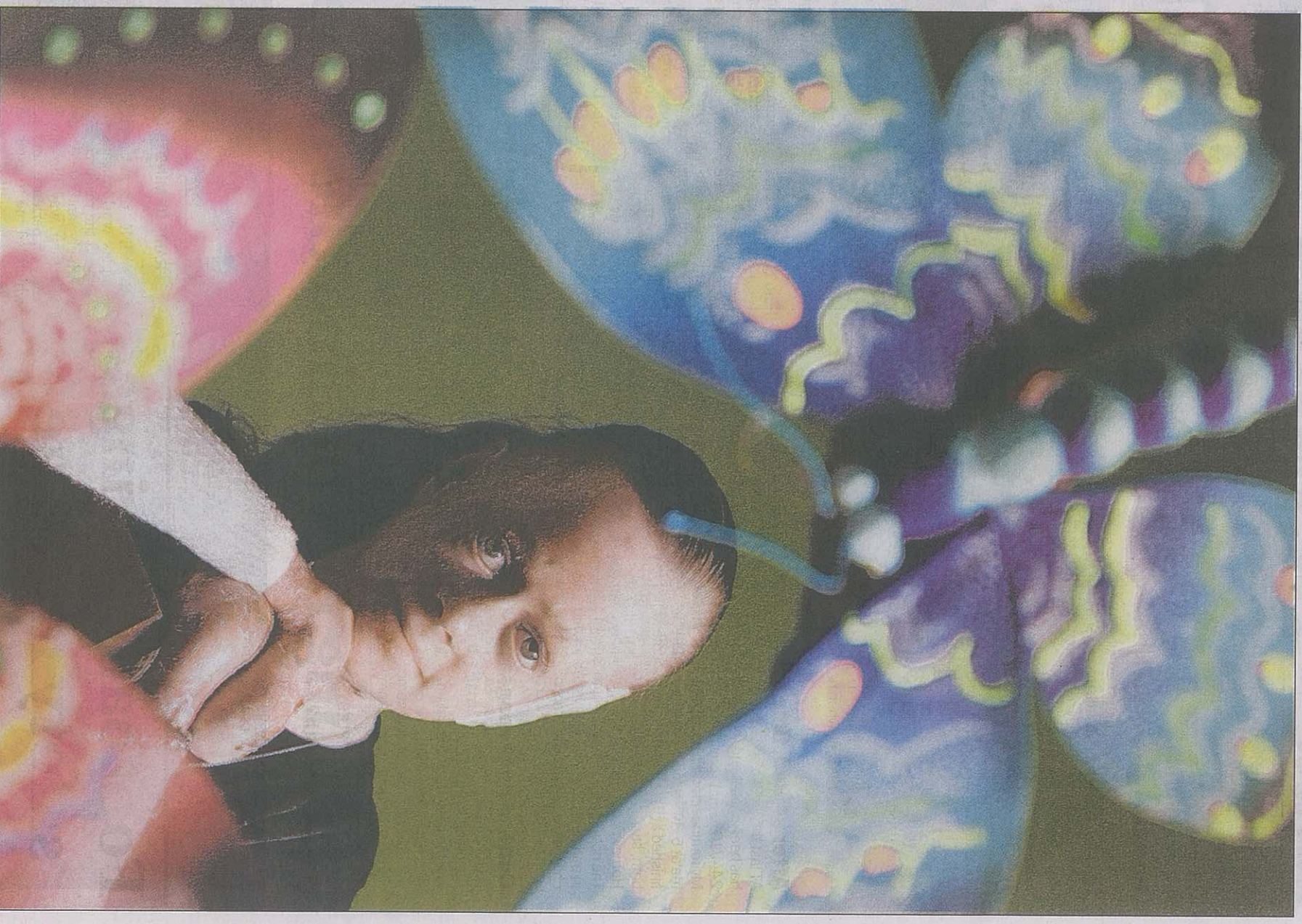
Targeting the problem protein can also help in the treatment of burns and chronic wounds.

The scientific community has welcomed the news. Mr Kopecki won a prize for his presentation at the Australian Wound and Tissue Repair Society Conference in Perth earlier this year.

His work has benefited from a travel fellowship offered by the university during his final year of study. This money is given to students who wish to travel overseas and present their research or do experiments in another laboratory.

"Last year, I went to international conferences in France and Hungary where I presented my research in plenary (keynote) talks and also spent some time in Freiburg, Germany at the Centre for Fragile Skin," he said. "I had the chance to use the best available mouse model of dystrophic EB and get trained by specialists in the field."

A development grant from the National Health and Medical Research Council worth \$529,450 will go towards further research. "This is a genetic disease," Mr Kopecki said. "But each patient would have



GETTING ON WITH LIFE: Sufferer Kate Turner and, below, PhD student Zlatko Kopecki.

Main picture: TAIT SCHMAAL

cure the disease, but it would improve the wound healing and quality of life."

Mr Kopecki, now South Australian president of the Dystrophic Epidermolysis Bullosa Research Association of Australia, said EB patients like

Kate were a constant source of inspiration. It was "very rewarding" to take research all the way "from the laboratory bench to bedside in the hospital".

"Having this interaction with patients has helped me realise the potential of our work and how important this therapy would be to improve the quality of life," he said.

Kate's mother, Linda Turner, said it was exciting to think that "things are really happening" for people living with EB and for future patients too, children yet to be born.

"I always thought that, hopefully in my lifetime, they'd come

up with something," she said. "Lots of things have happened in the last 21 years and it's amazing where medical science does go."

Meanwhile, Kate is simply getting on with life.

"A typical day used to be getting up, getting ready for school, but I don't go to school any more because I've finished," she said.

"Every Thursday, I go to my art class... I like drawing, painting, doing things with animals, watching TV and going to the movies."

Many people are surprised to learn she spent five months travelling overseas in 2007, but Kate says she can go anywhere if she takes loads of bandages and has time for the daily routine.

"No matter where you are, you still have to do it," she said.

"I got a voucher for my 18th birthday to go around the world and we met a lady at the (international) conference with EB. She's 42 and she's got the same degree as me."



a different gene mutation for the same protein anchoring the skin. It's very hard to develop patient-specific gene therapy.

"Our aim is to develop a therapy that would improve wound repair, so it wouldn't necessarily