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Boy cured of rare illness after two stem cell grafts

Two-year-old Donovan developed weepy, flaky skin when he was two months old. His skin became so inflamed that it turned black. After numerous tests, doctors discovered he had Omenn syndrome, a very rare disease and a severe form of immunodeficiency. After two stem cell transplants, he is now on the road to recovery. With him in the picture are his parents, Mr Tay (left) and Ms Lee, and Dr Soh, who diagnosed his condition. PHOTO: MATTHIAS HO FOR THE STRAITS TIMES

PUBLISHED 4 HOURS AGO

Doctors in Singapore have cured a baby of Omenn syndrome, the first known case of the extremely rare disease here. Two-year-old Donovan Tay is now thriving, after a rocky start in life. The Straits Times talks to the people who made this possible.

Chang Ai-Lien **(mailto:ailien@sph.com.sg)**

It took nine months before Ms Jace Lee saw her baby smile.

What had been an uneventful pregnancy turned into a nightmare for new parents Ms Lee and her husband, Mr Keith Tay, when Donovan was just two months old.

His nappy rash spread across his entire body, leaving him red, flaky and very itchy. "Every inch of his body was inflamed and we became really worried when his ears and neck got sticky and smelly," said Ms Lee, 34, a housewife.

Doctors initially thought he had developed an infection due to severe eczema and allergies, and started him on an intensive skin care regime. This included moisturising his body every two hours, and bathing him in a chlorine bath to help ease the symptoms.

"He was scratching non-stop by rubbing his head and body against the mattress," said Ms Lee. "We had to put his hands in silk mittens and put gauze all over him or his skin would become really weepy and bleed from the scratching."

Even a simple diaper change took 45 minutes because it involved changing all his dressings.

"He didn't have time to play and his hands were bound all the time," she added.

Then Donovan started vomiting, and did so for a month despite medication and constant care. But he stopped scratching, and his parents thought he was finally on the mend.

A visit to the doctor soon set them straight. Donovan had developed a dangerously high level of sodium in the blood, a consequence of proteins not playing their part in stopping water from leaking through the skin.

After several weeks at the National University Hospital Paediatric intensive care unit, doctors managed to bring his sodium levels down, but he remained small and very thin.

It was during his hospital stint that he came under the care of Dr Soh Jian Yi, a consultant at the NUH Division of Paediatric Allergy, Immunology and Rheumatology, and a key member of the medical army who would eventually save his life.

Faced with his small patient's unexplained fever, unusually severe eczema and surprising abnormal blood test results, Dr Soh turned medical detective to try to diagnose Donovan's mysterious illness.

His first clue: The baby had no detectable antibodies in his blood - the body's weapon against bacteria and viruses. Even a cold could have killed him.

"Every disease has its own time line," explained Dr Soh. "There tends to be a very specific sequence in which the features of each disease occur."

Dr Soh recalled his readings some years earlier on rare diseases, one of which resembled Donovan's situation. He pored over the medical literature and consulted local and international experts. The consensus: It all pointed to Omenn syndrome, a condition so rare that it has never before been reported in Singapore.

It is caused by mutations in genes that are critical to the immune system, which protects the body from infections. If not treated, Omenn syndrome is fatal.

Said Dr Lee Bee Wah, a visiting consultant at the NUH Department of Paediatrics: "To our knowledge, he is the first case that has been diagnosed in Singapore."

Added Dr Soh: "It's very, very rare; it would be easier to strike the lottery. But looking at all of the clues in Donovan, nothing else fitted the entire picture."

The only treatment was a transplant of blood-forming or cord blood stem cells.

Noted Dr Tan Poh Lin, a senior consultant with the hospital's Division of Paediatric Haematology and Oncology: "Without a transplant, it would have just been supportive care till death, which would be any time, such as when the most severe infection struck."

But Donovan was in such bad shape that he could not have survived the procedure then.

Said Dr Soh: "It was difficult but we didn't have a choice."

There were more hurdles to come. On New Year's Eve in 2014, Donovan collapsed, his heart stopped and his organs began to shut down. Doctors revived him 18 minutes later. His parents kept vigil at his bedside.

"We kept on getting 'the talk' from the doctors," said Ms Lee. "Be prepared, be prepared, they said. But on the third day, he opened his eyes."

At the time, he was on dialysis as his kidneys had failed, on a respirator to help him breathe and on 20 different types of medication. His skin was so damaged and inflamed that it had turned a black hue.

"There were IV (intravenous) plugs everywhere. He was connected to a whole 'garden' of syringes and tubes," said Mr Tay, 34, a regional risk manager with a Japanese trading house.

Although he was still very sick, doctors went ahead with the transplant of his father's stem cells in January last year after discussing it with his parents, as they felt it was his best chance of survival.

But, said Mr Tay, "after 14 days, his own cells came back and killed Daddy's cells".

His father's cells did, however, buy him some time, by rescuing him from severe infections and allowing for a more intensive preparation for his body to receive a second stem cell graft.

The next month,after chemotherapy again to kill off Donovan's own defective immune system, Ms Lee was the next donor. This time, the transplant was a success.

Said Mr Tay: "Visually there wasn't any improvement at first. But then his skin stopped peeling for the first time, and slowly he began to need less medication and less support. Slowly he moved from intubator to gas mask to a tube to breathe. I would like to think that with my transplant, I cleared the way for Mummy's cells."

Added Ms Lee: "He was still so weak, we had not seen him move for such a long time. But we celebrated all the little victories, such as one fewer antibiotic. When he peed, we jumped because it meant his kidneys were working."

As Donovan's blood tests began to show more normal results, his parents allowed hope to creep in.

He was still "a bag of bones", without the strength to lift his arms. And they were uncertain if he had suffered brain damage.

"The heartache was to see my baby not being able to move," said Ms Lee, breaking down for the first time during the interview.

"But his eyes were very expressive. We believed that he was in there somewhere. We kept believing he was in there."

Gradually, their boy was able to turn his head and move his arms. And, best of all, he began to smile and laugh. Two months ago, he no longer had to be tube-fed and could finally eat on his own.

These days, there is no stopping Donovan, whose skin is now smooth and itch-free. At the family's MacPherson flat, he is always on the go, and interested in everything.

His overjoyed parents reiterated how grateful they are to the doctors, nurses, surgeons and therapists for saving Donovan, be it from the ICU, renal, oncology, immunology or infectious diseases teams.

Said Ms Lee: "For a very long time, he was on the edge. We cannot express how happy we are now seeing him grow and learn, just like any normal child. Our hope for him is one that every parent wants, which is for Donovan to be happy and healthy."

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Singapore doctors cure baby of rare immune disorder

From the age of two months, baby Donovan Tay developed unexplained fever, unusually severe eczema that covered his entire body and blood abnormalities. His skin was also turning black from what was later diagnosed as Omenn syndrome, an extremely rare immune disorder. But he turned the corner after a successful stem-cell transplant in February last year. Now a bright and lively two-year-old, he is pictured here with his parents, Mr Keith Tay and Ms Jace Lee (above). PHOTOS: KEITH TAY, MATTHIAS HO FOR THE STRAITS TIMES

PUBLISHED 6 HOURS AGO

Boy with first known case of Omenn syndrome here saved by stem-cell transplant

Chang Ai-Lien Assistant News Editor

Doctors here have diagnosed and cured a baby of Omenn syndrome, the first known case of the extremely rare disease in Singapore.

Donovan Tay began to get very ill from the age of two months, plagued by unexplained fever, unusually severe eczema that covered his entire body, making him itch incessantly, and blood abnormalities.

It was Dr Soh Jian Yi, a consultant at the National University Hospital's paediatric allergy, immunology and rheumatology division, who was finally able to pinpoint what he was suffering from.

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"It is so rare that I had never seen it before, but in the end, it could only have been that," he said.

Omenn syndrome is caused by mutations in genes that are critical to the immune system, which protects the body from infections. It is thought to occur in fewer than one in a million people, and is fatal if not treated.

But Donovan could be cured if a transplant of blood-forming or cord blood stem cells worked.

Time was running out, though, as he became critically ill.

On New Year's Eve in 2014, the tiny baby's heart stopped beating for 18 minutes before doctors were able to resuscitate him.

But he turned the corner after a successful stem-cell transplant two months later - in February last year - from his mother, housewife Jace Lee, 34.

Now a bright, lively boy, he is unrecognisable from the baby whose organs had begun to shut down and whose skin was turning black from the immune disorder.

As their son turns two on July 10, his parents are looking forward to taking him to pre-school and other activities, such as baby gym and swimming, which he has missed out on so far.

Said Ms Lee: "He's very sociable, but he's been kept away from his peers, he's been a bubble boy for so long. Now we have the chance to take him out, especially to places where he can be with other kids."

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