



Meet **Adam**

**He looks like
an average
10-year-old
except that he
travels to the US
every month for
life-saving
treatment**

STORY PAGES 2&3

This Life Independent *Life* 19.7.05

Adam Costello-Doherty has to travel to a special clinic in the US every three weeks to bolster his immune system, otherwise a simple infection could take his life. Lisa Jewell reports

Adam: the smile that shows he's winning



Looking good and feeling better: Adam Costello-Doherty with his mum, Judi and dad, Tom. Pictures by Frank McGrath

Most of us regularly catch a cold or the flu and even occasionally contract more serious infections like pneumonia. When we are ill, the one thing we count upon is our body's immune system, which fights off viruses and bacteria and gets us back to good health. But for children like Adam Costello-Doherty bouncing back from a common cold or infection is not a straightforward matter.

Adam, 10, suffers from primary immunodeficiencies (PIDs), which means his body is more likely to contract infections and has a reduced ability in fighting them off once they take hold. Looking at the youngster, however, you have no idea that he has any health problems. An active boy who is mad about helicopters and Harry Potter, Adam is living a normal life thanks to treatment he receives every three weeks at the Paediatric Centre for Rare and Complex Disorders in the United States.

Adam was diagnosed with his deficiencies three years ago. Finally getting a name for his condition came as a great relief to his family. "At last, we knew what was going on," says Judi. "There's definitely some truth to the saying 'better the devil you know' because now we could get some treatment. Up until then, it was very stressful — you had no idea what was going to be thrown at you next."

Adam had medical problems since he was only a few days old but at the age of seven he began to show behavioural and neurological problems. "He had autistic and Tourette's Syndrome tendencies," explains Judi. "He would shout, shake his head and blurt out things. He also had Obsessive Compulsive Disorder and would jump over cracks and obsessively count numbers."

Just one of these behavioural problems would challenge any parent but tackling all three was extremely difficult to deal with. "He constantly screamed and couldn't speak properly. At one stage, I had to wear earplugs because he was so loud. It wasn't a nice time for us and it was getting to be unbearable," says Judi. It was uncharacteristic behaviour for the bright, outgoing boy and Judi knew there had to be some underlying cause. Adam also suffered from persistent bouts of pneumonia and common viruses. She decided to obtain his medical records under the Freedom of Information Act, and sent them off to a large teaching hospital in the States.

"They arranged all the consultants and we flew over for him to be assessed. They left no stone unturned and the blood tests showed that he had these immune problems." And this is how he ended up as a regular patient at the Paediatric Centre for Rare and Complex Disorders, based at the University of Medicine and Dentistry in New Jersey. He is treated there because he suffers from both an immune deficiency and an overproduction of immune cells in response to streptococcus infection (PANDAS syndrome).

Instead of fighting the infection, his antibodies mistakenly attack healthy cells in the brain, which can lead to respiratory illness, pneumonia and other infections. "It's quite rare to have both conditions," explains his mum Judi, who accompanies Adam on his trips from Dublin to New Jersey. Primary immunodeficiencies are relatively rare; about 70

Irish people are diagnosed with the conditions. There is evidence to suggest that more people suffer from a PID but are putting up with repeated infections instead of getting to the root of the problem.

Having an immune deficiency can affect almost every part of a sufferer's life — their work life, family and personal development. Minor ailments that wouldn't be of concern to other people have to be properly treated so they won't develop into serious life-threatening conditions. As well as taking medication here in Ireland, Adam goes to the States to receive IVIG blood infusions. They effectively give his body what his immune system cannot naturally provide — proteins in blood and tissue fluids called immunoglobulins, which protect the body against viruses and bacteria. Within a day or two of receiving the infusions, his mum notices a huge difference in Adam. "He's as right as rain when he comes back from the States," she says. "The infusions have had a profoundly good effect on him. He's become more active since he started the treatment three years ago and the behaviour difficulties have all gone away, except for some occasional tics when he is feeling unwell."

Adam receives treatment in the US because specialist care is needed for the administration of his infusions and at present there is no paediatric immunologist working in Ireland. There are plans, however, to recruit such a specialist in this country and if it goes ahead it will mean the end of tiring transatlantic journeys for Judi and her son. Adam also receives general paediatric care at Our Lady's Hospital in Crumlin, which Judi highly praises. "The communication between Our Lady's and the American hospital is excellent. With Adam's illness, it's been one of those things that you don't understand how terrible it is unless it touches you and then you suddenly realise how great a place like Our Lady's is and how worthwhile it is to support it."

Support is also on hand from the Primary Immunodeficiency Association of Ireland (PIAI), which was set up in 1997. "Our purpose is to create awareness of these deficiencies, especially amongst GPs, but also amongst the general public," says the group's secretary, Maria Nolan. "A lot of people aren't being diagnosed with the condition when getting proper treatment could give them a much better quality of life."

At present, Irish children with immune problems are treated at the Rainbow Clinic at Our Lady's Hospital for Sick Children, Crumlin. But the PIAI is concerned that those children are coming into contact with children suffering from infectious diseases. "We feel that the two groups sharing a waiting room is totally wrong," says Maria Nolan. "Children with a PID are supposed to be cared for in a way so they don't come in contact with infectious diseases as their immune systems don't have the mechanisms to fight infections."

The hospital, however, says it is following what is accepted as best practice in other countries such as Britain. At present PIDs are incurable but fortunately there has been a lot of new research into immune deficiencies and treatments. At last count, the World Health Organisation had identified 70 different deficiencies and new types are still being identified thanks to genetic research. One of the rarest kinds is Severe Combined Immune Deficiency (SCID), which usually results in the onset of serious infections within the first few months of a baby's life. Illnesses can include life-threatening bouts of pneumonia, meningitis or bloodstream infections and babies have to be kept in a sterile space, commonly called a 'bubble'.

SCID is often called 'bubble boy disease' because of a famous case in the US. A baby called David Vetter was born with the disease in Texas in 1971 and was immediately placed within a sterile plastic isolator because of the threat of fatal infection. Doctors assured his parents that this would be a temporary measure until a cure for SCID was found — but David lived in a bubble home for the next 12 years. Although he adjusted as best he could, it was a lonely existence for the child without physical contact with others or personal freedom. His story became a source of fascination for the media and members of the public. In 1983, a bone marrow transplant was arranged for David and in the months afterwards it looked like he might be able to leave the bubble and lead a normal life. Sadly, complications set in and he died in 1984, aged 12.

Even though the image of the boy in the bubble is still associated with primary immunodeficiencies, the treatment options have evolved since the 1980s and bone marrow transplants are proving to be very successful in treating SCID. One of the biggest factors in combating immune problems, whether they are severe or manageable with medication, is to keep the mind focused on positives rather than negatives. It's the approach that Judi Costello and Adam take with his PIDs.

"Adam knows he is the most important member of the team fixing him," she says. "He's very proactive about doing things like washing his hands when he comes in from playing, eating the right foods and keeping his happiness levels up." Despite missing some school because of his condition, the boy is catching up quickly. "The whole experience has had a profound effect on us but my family have been really supportive and Adam's dad is phenomenal," says Judi. "Other parents may have a child who is constantly ill and feel like they don't know what to do for them. I'd advise them to find out more about immune deficiencies and make sure they get help for it."