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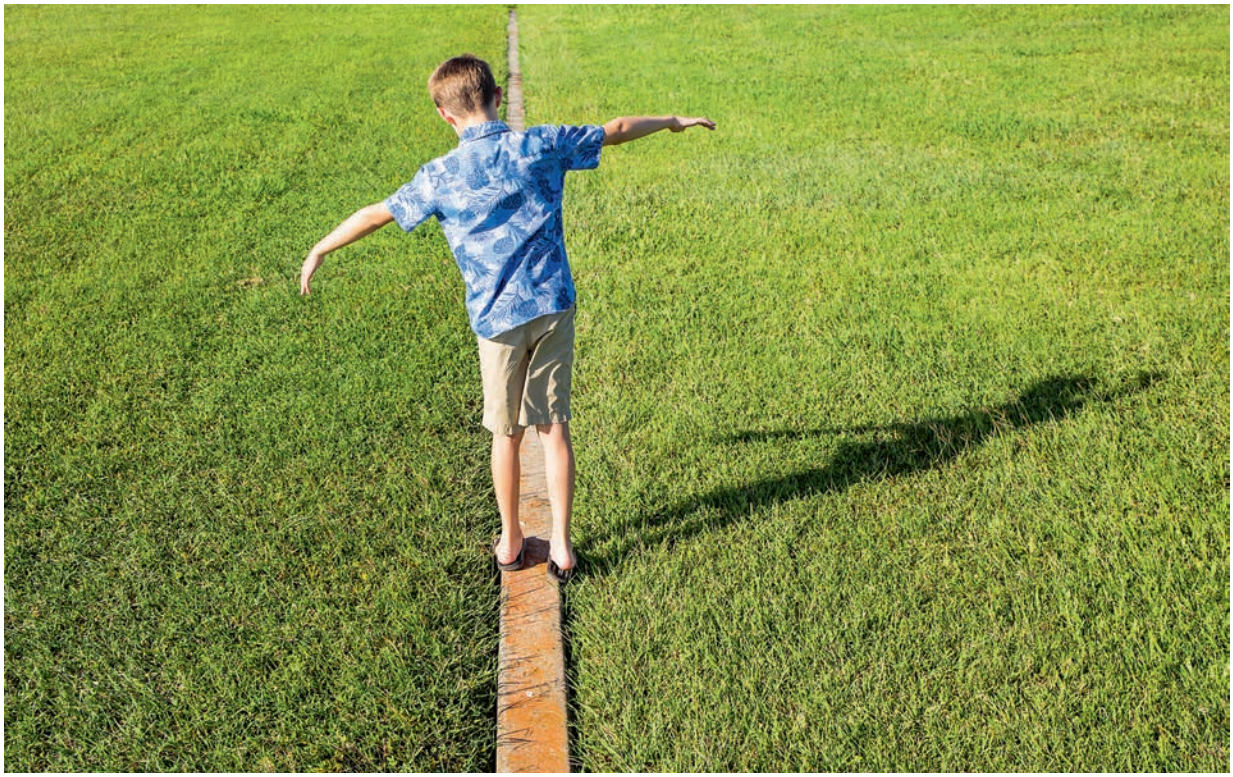
Team Ethan

The network within, the network without

Ethan Deviney was born without a cerebellum. The outside world stepped in to take its place



HONOLULU

"He shot a six-year-old in the eye!"

There's trouble in the playground on Ford Island. A handful of Navy kids have spent the afternoon scrambling up green slides and over climbing frames that are coated with smooth brown plastic, firing on each other with outsized Nerf guns. The mug of the Honolulu evening is tightening, and proceedings have turned dramatic.

Ethan Deviney, the accused, trots out to the pavement and states his case to his mother. To the extent, he insists, that he may have shot someone in the eye, he is vindicated through being the prior recipient of a foam bullet to the tooth.

Heather Deviney marches her son back into the playground. A stout boy in an Indianapolis Colts jersey, not the least bit hurt, stands at the apex of a protective huddle of children. "Can you say you're sorry, Ethan?" Heather asks. He can, and he does, and then he goes one better. "Let's hug it out," he says, grinning and flinging his arms wide. Colts jersey hugs gingerly. All is well.

Ethan is a skinny 11-year-old boy with a broad smile. He says hello to nearly everyone he sees in public and gets uncontrollably excited in the presence of new sets of Lego. His father, Jeff Deviney, is a captain in the US Navy's Civil Engineer Corps. Heather is a graphic designer. Ethan has two older brothers, Jared, 16, and Corbin, 19. And he is one of just ten people known to have been born without a cerebellum.

Put your hand on the back of your head, where the skull curves down to the neck. Your cerebellum is an inch or two beneath your palm, a dense knot of brain tissue which sits slightly apart from the rest. In just 10% of the brain's volume, the cerebellum contains half of all its cells. You would think that would mean it was vital. But though Ethan's life without one has its challenges, it is in most ways pretty close to normal—because what his absent cerebellum cannot do, the rest of his brain and the rest of his family can.

Equating family ties with a missing piece of the brain sounds odd. But maybe it, too, is close to normal. Maybe Ethan's unusual circumstances simply reveal a general but easily overlooked truth: that there is a fungibility between what bodies and minds can do, and the minds involved do not have to share one body.

Ethan was born in the early morning of November 28th 2006, a Tuesday, in San Diego. He was a healthy baby; the knot in his umbilical cord seemed inconsequential. But by the end of his first year, Heather no- ▸▸

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ticed that Ethan was not sitting up or crawling at the same stage as her first two boys. Photos from the time show him lying on his belly, playing intently with toys.

At 18 months, Ethan was referred to a development specialist at Rady's Children's Hospital in San Diego. The doctor found him an odd mixture of progress and lag. His cognitive development was right on track: he knew how to find things that had been hidden; he was able to solve puzzles. Socially and physically he was normal for his age, and his fine motor skills, tested by playing with blocks, bottles and toy cars, were OK. But his gross motor skills—his ability to hold his body steady or to crawl—were a long way behind. His language abilities were delayed, too.

Tough love

What was going on was not clear. Forms of autism, or epilepsy, or cerebral palsy were considered, but none provided a satisfactory explanation. What Ethan needed was easier to work out. He had session after session of physical and speech therapy. Supporting his development, Heather says, became the family's primary mission. "There was a lot of tough love to encourage walking. It was 'Off the floor, off the floor, off the floor'. It was our whole life." They had pushed their first two boys hard, Jeff says. They would not treat Ethan any differently.

With regular therapy and intensive family support, Ethan's movements and speech became stronger, but he still couldn't stand or walk unaided. When Heather took Ethan to a new paediatrician in Los Angeles the doctor warned that there was a high likelihood that Ethan would never be able to move around without assistance. The Devineys redoubled their efforts, still not knowing what they were helping him overcome.

Then, in summer 2009, aged three and three-quarters, Ethan stood up. The family was at a baseball game, watching Jared play. Ethan was mucking around in the grass in front of the stands, a few feet from his parents. Heather looked around at the precise moment that Ethan pulled himself to his feet and stood upright for a few seconds, while everyone else's attention was focused on the game. "It was monumental," she says. "I was moved to tears."

Since antiquity the cerebellum—"little brain" in Latin—has been understood as an anatomically, and presumably functionally, discrete part of the brain. Galen, a Roman physician, thought it the valve between brain and body through which "animal spirits" flowed. It would take another 1,400 years for Constanzo Varolio, an Italian anatomist who pioneered the dissection of the brain's soft tissue, to reveal the regularly repeating tight-packed whorls of tissue of which it is made.

Varolio decided that the cerebellum must be the brain's seat of taste and hearing. In the 18th and 19th centuries, though, anatomists observed the effects of its removal or damage on balance and movement. A goat with half its cerebellum removed, for example, subsequently swayed and fell but suffered no loss of consciousness. Neuroscientists noticed that patients who had suffered damage to their cerebellum often walked awkwardly or slurred their speech.

Today, the cerebellum is understood to be the brain's quality-assessment centre, ensuring that actions the higher brain has decided on go according to plan. The electrical signals that travel from the upper brain out to, say, the arm, to tell it to type, pass through the cerebellum, which takes down a copy of the plan. When information describing the movements in progress comes back from the arm muscles, the cerebellum's tightly meshed network of billions of nerve cells compares the situation described to the original plan, detects any deviations, and sends a better-honed signal back out to the arm. At the same time, the network of cells adjusts its internal wiring so as to make sure any error is less likely to happen again. As well as supervising the smooth running of the body through endless cycles of error correction, the cerebellum helps the brain and the body learn from their mistakes and miscommunications.

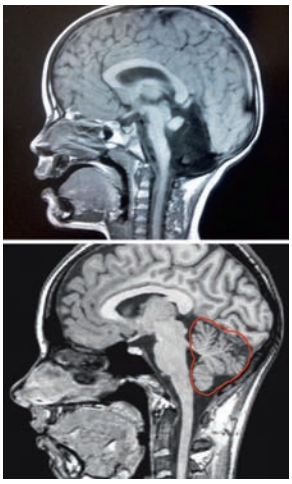
Until recently, this quality-assurance infrastructure was thought to be limited to the oversight of physical movements. But brain scans now suggest that the cerebellum's remit is much broader. "The cerebellum does the same thing to emotion and cognition that it does for motor control," says Jeremy Schmahmann, a neurologist at Harvard Medical School. Just as physical movements are monitored, controlled and regulated by the cerebellum, so too, it turns out, are other sorts of thought, including speech and emotional work. The set of mental actions required to remain calm under pressure, for instance, runs through the cerebellum.

Flipping through old pictures, Heather is particularly keen on the ones that show Ethan standing, running, or jumping

All this takes place far below the threshold of consciousness. It is a sort of internalised automation, allowing processes to go on in the background without any attention. You don't know that you are doing it. But you would notice very quickly if you stopped.

Ethan had his first brain scan on October 15th 2010, just before his 4th birthday, at Le Bonheur Children's Hospital in Memphis. By this stage, his doctors had begun to suspect a damaged cerebellum. But that was not what the scan (reproduced on this page, with a scan of a normal brain below for comparison) showed. His cerebellum was not damaged. It was absent; there was a featureless dark hole where the cerebellum would be. His brain—or his body, depending which way round you look at it—lacked an autopilot.

Heather says the scan "opened up a whole new world". The family had felt alone, isolated by a condition that no doctor quite understood. Now Heather was finding Facebook groups devoted to cerebellar issues and learning the right keywords to put into search engines. The term "cerebellar hypoplasia", meaning underdevelopment of the cere-



With cerebellum (below) and without

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bellum, led her and Ethan to Dr Schmahmann's office in Boston the following year.

Learning that her son's brain was not so much damaged as simply missing a part also allowed her to see her and her family's supporting role more clearly. "If you were born without a leg, you would put on a prosthetic and that would be it," she says. Being born without a cerebellum was more complicated. But prosthesis was still possible. It just had to be provided by people.

The person doing the most work is Ethan. Unfortunately, he cannot tell you much about how he does it. When you ask Ethan how it feels to do things that the cerebellum is supposed to govern, such as walking, he has trouble answering, and that is fair enough. To him, walking—something he does with a loose, slightly swaying gait—just feels like walking. But he is doing it in a very different way to most people.

Mind the gap

"We think the neural circuits are learning this somehow," says Dr Schmahmann, "but without the smoothness and automaticity of the cerebellum." One possibility is that the basal ganglia, parts of the brain responsible for the formation of habits, are taking up at least some of the slack. This would mean that Ethan remembers how to walk in the same way that you remember morning rituals, or your preferred route to work. You might think of those as being things you do "on autopilot", but the process is not quite the same. A habit does not provide the same sort of self-regulating feedback that an aircraft or cerebellum's autopilot provides. It is more a stereotypical sequence of actions. To make good the absence of a cerebellum with circuits designed to encode habits is to bridge a gap that is not meant to be bridged. If that is what Ethan's brain is doing, it is something that takes a lot of support.

Of his handful of patients without cerebellums, Dr Schmahmann says: "What we're finding is that with intensive rehab, bringing things to conscious awareness and making [patients like Ethan] conscious about what they need to focus on, you allow them to improve." Those without such intense rehab learn to walk and talk much later than Ethan.

The Devineys have, without really knowing it, built some of the functions of Ethan's cerebellum into his life, and their lives, through other means. When he was younger, they had to show him how to walk, how to control his body, over and over. As she flips through old pictures, Heather is particularly keen on the ones that show Ethan standing, running, or jumping, all things he learned to do against the odds. Her efforts were Ethan's efforts. The Devineys, says Dr Schmahmann, are Ethan's exoskeleton, an external support system shouldering the load of the one that is missing inside.

Ethan's family still regularly plays that role. During a day at the beach, as Ethan comes up from the ocean to the gazebo the family is sitting under, he walks over a patch of ground carpeted with small, spiky pine cones. Walking on them is painful, and Ethan doesn't really adapt to it. He loses control a little, his smooth temperament upset along with his balance. His mother goes over and picks him up, carrying him the few feet to the chairs. He starts saying that he wants to go home: "I think I'm done now, Mom." The hiccup makes him want something familiar, something that can guide him along, because his cerebellum is not there to help him.

Heather is having none of it, though, and soon enough Ethan is back to normal, chatting and asking questions.

"Why are you writing a story about me, Mr Reporter?" he asks.

"Because you and your family are interesting, Ethan, and I think people who read the story can learn something from you."

The answer is acceptable, and Ethan goes back to the Lego figures he brought with him from the car.

Science tends to treat the brain as the be-all and end-all of mental life—what it is constrains what people can think and do. Philosophers speculate about how people can know that they are not simply isolated brains in a vat, hooked up to input and output mechanisms that lie to them. But watching Ethan and his family deal with the absence of a chunk of brain makes you re-examine what you think you know about the organ. Some of what keeps Ethan going is thanks to other bits of his brain pitching in to take up the slack. But some is due to other people's brains. In this sense "Team Ethan" may be a special case of a broader principle: that all people rely on an external support network to be the people that they are.

Ethan, Incorporated

This idea that being a person is something distributed beyond a specific body drives the work of Hélène Mialet, a French philosopher and anthropologist. Her book "Hawking Incorporated", published in 2012, was an anthropological examination of the late Stephen Hawking and the human and mechanical aids which made it possible for him to navigate the worlds of knowledge and everyday life. The people and machines that surrounded him choreographed Hawking's genius, Ms Mialet wrote in his obituary.

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She is currently applying a similar analysis to people with diabetes, trying to understand how the people and objects around a type I diabetic stand in for the pancreas within them that does not work. A doctoral student of hers has studied Lance Armstrong, trying to understand the network of people, objects and materials which supported his cycling prowess.

This blurring of a biomolecular perspective with that of the social world and its man-made objects is a deliberate strategy. Ms Mialet argues that humans are "distributed" across these worlds; that what it is to be someone extends beyond a single body. But because such distribution is fundamental, it is also hard to see—except in exceptional circumstances. It is conceptually quite easy to study the cellular networks within skulls and immune systems, even if the actual experiments are hard. It is more difficult to get to grips with the networks beyond the body. They are so much a part of life that, like the action of the cerebellum, they are accepted automatically. But when the external networks start taking the place of an internal one—of a missing cerebellum, or an unbalanced insulin metabolism, or a nervous system raddled by amyotrophic lateral sclerosis, like Hawking's—their role is made more apparent. At least, it is to the theorised, anthropological eye.

This general idea of making up on the outside what is missing on the inside makes sense to Andy Clark, a cognitive scientist and professor of logic and metaphysics at the University of Edinburgh. "Brains don't care whether stuff gets done on the inside or on the outside, just as long as it gets done," he says. If using a prosthesis allows a more efficient use of resources, the brain jumps at the chance. Using calculators or search engines does not leave the human mind uncomfortable; it is just a convenient outsourcing. Ethan's replacement of his entire missing cerebellum with services from other parts of his brain and family may not even be that extreme, Mr Clark suggests. "We don't think of the successful poet or artist, 'Oh, poor thing, they can't do that in their heads, they need pens and paper and software'," he says.

How might one prove that this is more than a figure of speech—prove it in the way a brain scan proves things? Not by measuring which data from the environment are linked to what process in the brain, Mr Clark says, at least not with today's technology. But there is other corroborating evidence for the distributed self. Look at the way people with retrograde amnesia, who cannot remember events before an injury, use their phones to carry on healthy social lives. "They rely on trails they lay down in software space to remind themselves of what they're doing and why they care." Their lives are shaped by a structure they have built outside their brains.

Taking care

The load on Ethan's external networks is now shifting. Heather no longer has to think much about Ethan's physical co-ordination. Now she worries about him making friends. "I'm a really social person, so it's hard for me to see him being uninterested in that," she says. Friendship, like other sorts of balance, can be hard. Understanding what another person wants, or what they are talking about as they explain some event over which the two of you might bond, is the kind of function the cerebellum takes care of without you having to think about it. Ethan will have to practise. "We're in a more cognitive chapter," says Heather.

It will be one that the family will write for itself. Heather spends less time on the cerebellar Facebook groups these days. Almost all the children on there have far more serious disabilities than Ethan, with walking or talking being difficult. They all have cerebellums that are damaged in one way or another, not completely non-existent ones like Ethan. Somehow, not having a cerebellum at all is better than having a damaged one. Dr Schmahmann says it is likely that Ethan's cerebellum was destroyed by the equivalent of a freak accident, one that left the rest of the brain untouched (possibly a stroke of some kind, due to his knotted umbilical). Other children's cerebellar issues are just one of an array of confounding factors. Ethan's are more discrete—and thus more susceptible to distributed replacement by other parts of his brain, by his family, by objects in his life.

With his Lego, he sits back on his ankles and leans forward at the waist to pore over the field of components, deep in concentration, in thrall to the instructions. He is not his chattiest self when in Lego mode. But watching him and helping him (help from an amateur is clearly not making up any deficit, but it is politely accepted nonetheless) it feels like his brain is leaning on the design behind the Lego set. The logic of its structure frees him from the load of having to use the missing part of his brain designed to automate the fitting together of patterns.

At Honolulu Zoo, he sounds out the names of exotic birds as he and his mother wander past their enclosure. Each animal, whether lion or lemur, gets about 15 seconds of attention before Ethan announces that he is "moving on". He explores the zoo voraciously, padding along every branch of the path, as thorough and explicit in mapping it out as he is in playing with Lego. Peering into the elephant enclosure, Ethan's foot slips off the path. It's a tiny thing, but it bothers him disproportionately, just like the pine cones. He starts asking to go home. Heather reaches out, steadies his shoulders, and Ethan races on to the next attraction. ✱