YOUNG, FIT AND DROPPING DEAD

In 1947 US doctor Claude Beck used a defibrillator he had designed to revive a 14-year-old boy who went into ventricular fibrillation during open-heart surgery.

Health Sudden cardiac arrest kills five Australians aged under 35 a week. But we can reduce the toll, writes Sophie McBain in AFR on 3 Aug 2019.

On a mild spring morning in 2017, Clare Sutton, a 31-year-old biology teacher, a single mother and one of my closest friends, was running a 10-kilometre race in the village of Poynton, close to Manchester, when she collapsed 100 metres or so short of the finish line. One of her friends, a nurse called Helen Thompson, had just finished the race. Assuming that Clare had fainted, she grabbed a bottle of water and a banana and rushed to help. As soon as Clare came into view, Thompson realised things were much, much worse.

“Clare was completely lifeless,” Thompson tells me.

Another fellow runner – an adult critical care consultant called Jon Bannard-Smith – was already crouched down and checking Clare’s pulse. She had none. Together, Bannard-Smith and Thompson began performing cardiopulmonary resuscitation (CPR). Thompson had given CPR more times than she can remember, but when patients arrived at the emergency room still without a pulse their chance of surviving was minimal, and she had never revived someone. She’d give it her best shot, and then her role would shift to supporting distraught friends and family members. It helped her maintain her professional distance if she didn’t look too closely at her patients’ faces while she compressed their chests and filled their lungs with air and fought against all the odds to save them.

This time things were completely different. Thompson had never performed CPR on a friend. She thought of Clare’s young daughter, who mercifully was no longer watching the race, and remembers screaming at Clare not to give up.

Nobody knows why Clare’s heart stopped that day. She was slim and fit and ate healthily. She didn’t smoke or take drugs; she seldom drank more than a glass or two of wine. But without warning her heart had gone into ventricular fibrillation, a dangerous

rhythm that causes the chambers of the heart to quiver uselessly instead of pumping blood. She had suffered a sudden cardiac arrest. By the time Thompson reached her, just a minute or two after she fell, Clare was clinically dead.

Cardiac arrest is responsible for around a fifth of all deaths in developed countries. Each year it kills 100,000 people in the UK (in Australia the figure is 30,000). Around half of those who experience cardiac arrest have no prior warning that there is anything wrong with their heart. Of those cases that occur outside hospital, almost 90 per cent are fatal.

A cardiac arrest is not the same as a heart attack. The latter is the result of an arterial blockage to the heart, starving it of oxygen; the former is an electrical malfunction that stops the heart pumping. (A heart attack can cause cardiac arrest, but doesn’t always.) Most people who suffer cardiac arrests are elderly and/or have heart disease, but it can also affect the young and apparently fit, and hearts that appear structurally healthy.

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Sometimes a young person collapses from a cardiac arrest because of an undiagnosed heart disease. The then 23-year-old Bolton Wanderers footballer Fabrice Muamba, who collapsed during a match against Spurs at White Hart Lane in 2012 and who was clinically dead for 78 minutes before doctors managed to restart his heart, was later diagnosed with hypertrophic cardiomyopathy, a congenital disease in which the heart muscle becomes thickened.

In the 2014 City2Surf fun run in Sydney, 27-year-old Chris Head collapsed just metres from the finish line and died in hospital. He was a fitness enthusiast.

A drug overdose or huge blood loss can trigger cardiac arrest, as can a number of electrical heart diseases, and genetic conditions such as Brugada syndrome or Long QT syndrome. But, according to Dr Arthur Wilde, a professor of cardiology at the University of Amsterdam, around 30 to 40 per cent of cardiac arrests in the young are unexplained. In the UK, 12 people under 35 die of a sudden cardiac arrest every week (in Australia five people in this age group die each week). The social impact can be devastating, and not just for friends and family members. When so few medical conditions are completely unexplained, we retain a special horror for these stories of teenagers who go to sleep and never wake up, or of promising athletes who die mid-race. They raise the disturbing, unanswered question: why do the young and healthy sometimes drop dead?

Clare survived. Thompson and Bannard-Smith continued CPR for 15 minutes until another bystander retrieved an automated external defibrillator (or AED) from the nearby civic centre. The AED delivered an electric shock to Clare's heart, and it began beating again. An ambulance arrived shortly after, and she regained consciousness on her way to hospital in Manchester. Her recovery was long and at times painful, but also complete and therefore close to miraculous: it is unusual not to sustain some degree of brain damage after a cardiac arrest. She now has a cardioverter defibrillator implanted in her chest, which would deliver another life-saving electric shock should her heart enter a dangerous rhythm again. So far, it hasn't had to. Clare thinks of it as her "internal ambulance".

I could list many reasons why I am so lucky to count Clare as a friend, but I will give just one. On the morning of her 10-kilometre race, I messaged Clare on WhatsApp to wish her a happy Mother's Day and ask her something about breastfeeding. My daughter, my first child, was six days old, and we had been messaging a lot about the logistics of breastfeeding and the bloody, unglamorous aftermath of childbirth. Clare has an endless capacity for chatting about gross medical stuff, and an unfailing ability to cheer me up. I found it odd, and then annoying, that she didn't reply.

Three days later, Clare sent my husband a private message on Facebook explaining what had happened and asking for his advice: she felt uncomfortable lying to me, but she also didn't want to ruin my precious first weeks of motherhood by telling me that she was critically ill. He sat me down and told me right away.

"I'm sorry it's taken me a while to reply to you. It's been difficult to find the right words. I love you so much and I just don't want you to have to think about anything except [your baby]," she wrote to me after I got in touch. I now know that she was in a lot of pain then, that she was still suffering from amnesia and that she knew she faced at least one more heart operation, but her focus was on reassuring me.

"I am remarkably OK, really I am," she wrote. When the doctors suspected she had Brugada syndrome (she doesn't), she advised me: "It's not too scary to Google." She frequently asked for updates on my daughter and my breastfeeding worries in messages that were sometimes hilariously garbled because of her strong pain medication (a favourite: "I am experiencing some very interesting altered planes of consciousness! How are your boobs? Are they comfortable?"). It is typical of Clare that during the most difficult period of her life, her focus would be on protecting other people from her suffering.

Until the middle of the last century, the chance of surviving a cardiac arrest was zero. To understand how to restart a human heart, you need to understand why it beats, and this question puzzled physicians for centuries. As Leonardo da Vinci accurately observed in the early 1500s, the heart is a muscle, but unlike other muscles, it contracts spontaneously. So what animates it? In the 18th century, when scientists began experimenting with the effect of electricity on the human body, it became increasingly apparent that it might play an important role. As Thomas Morris recounts in his fascinating history The Matter of the Heart, several serendipitous cures hinted at the potential for electricity to disrupt, and fix, problems with the human heart.

In 1757 John Wesley, the founder of Methodism and an enthusiast for electric medicine, passed an electric shock through the chest of a 48-year-old teacher who had been suffering heart palpitations for 17 years. The palpitations stopped. Two decades later, an amateur scientist is recorded as having used an electrostatic generator to revive a three-year-old girl whose heart stopped beating after she fell from a first-floor window.

The spread of electric power to homes and cities added new urgency to investigations into how electricity affects the human heart, as scientists grappled to understand why electrocution kills. In the 1890s, two

researchers at the University of Geneva studying the effects of electrocution on dogs made a startling discovery: after a first electric shock stopped a dog's heart, a second one could sometimes restart it.

The American doctor Claude Beck successfully applied this principle to revive a human heart. He had a defibrillator built, with metal paddles to deliver the shock, and in 1947 he used it on a 14-year-old boy who went into ventricular fibrillation during open-heart surgery. The boy made a full recovery. Beck described his mission as saving "hearts too good to die". As the medical professor Mickey S Eisenberg wrote in the Scientific American in 1998, "a heart that fibrillates is like a million-dollar piece of equipment failing because of a 20-cent fuse".

Beck's device only worked if applied directly to the heart, but in 1956 a researcher at Harvard Medical School performed the first successful external defibrillation. In the years that followed, defibrillators became lighter and more portable, enabling them to be used where most cardiac arrests occur: outside hospitals. The first automatic electronic defibrillator, the kind that saved Clare, was manufactured in the late 1970s. The first implantable cardioverter defibrillator (ICD), Clare's "internal ambulance", was fitted in 1980.

The brain cannot survive without oxygen for more than a few minutes, and so the manufacturing of portable defibrillators would have been of little use were it not for the development of CPR – the external chest compressions and mouth-to-mouth resuscitation that helps keep a person's brain oxygenated even if their heart has stopped. Surprisingly – given that the firstaid method requires no special technology, and mouth-to-mouth resuscitation is mentioned in accounts of medieval midwifery – it was only in the late 1950s that researchers at John Hopkins University in Baltimore created modern CPR techniques.

In 1960, a Norwegian toy maker made the first Resusci Anne, the blank-faced dummy that has been used to teach hundreds of millions of people how to save lives (and that entered pop culture via Michael Jackson's 1988 hit Smooth Criminal: "Annie are you OK?").

Despite this, in England only 55 per cent of people who suffer a cardiac arrest in public are given CPR by a bystander (40 per cent in Australia), and only 2 per cent are treated with a defibrillator before the ambulance arrives – even though this offers the best chance of survival. As a result, you're much more likely to die of an out-of-hospital cardiac arrest in England than in Norway, where a quarter of victims survive, or in Seattle or the Netherlands, where survival rates are 21 per cent. Research by the University of Warwick suggests that if England's emergency medical services began operating on a par with that of Norway, this would save 4500 lives a year.

Improving public awareness will help: after years of campaigning, CPR and other basic first aid will become part of the national school curriculum from next year. Several people I spoke to for this piece wanted me to underline that if someone is unconscious and either not breathing or not breathing normally, there is absolutely nothing to lose by attempting CPR and everything to gain. Without CPR, they will die. Public access defibrillators are also now designed so that they can be used by people with no medical training – the machine will talk you through what to do, and won't deliver a shock unless the person needs it.

To decrease the death rate, however, scientists also want to get better at predicting who might be at risk. This poses a huge challenge, particularly when it comes to identifying the risks among those such as Clare, whose heart is structurally normal; who has tested negative for the known conditions that cause cardiac arrest; who has not been diagnosed with any further illnesses; and who does not recall any potentially suspicious events, such as unexplained spells of fainting, dizziness or chest pain, in the run-up to her collapse. What else could cause a perfectly healthy heart to stop?

Dr Sumeet Chugh, the medical director of the Heart Rhythm Center at the Cedars-Sinai Smidt Heart Institute in Los Angeles, has been trying to answer this question for almost three decades. In 1993, when he was still in medical training, he treated a 19-year-old woman who had suffered a sudden cardiac arrest while dancing. She died. Her death left an enduring impression on him, inspiring him to specialise in cardiology and to devote a career to attempting to predict the seemingly unpredictable.

Since 2002 he has headed the Oregon Sudden Unexpected Death Study, which collects extensive genetic and medical data on anyone who suffers a sudden cardiac arrest from a community of around one million in Portland, Oregon. His "most precious subjects" – a small but growing cohort – are the people who

survive cardiac arrest and who he can continue to monitor. He gathers all their medical data, from their head circumference at birth to recent electrocardiogram (ECG) readings – which give an electrical recording of the heart: who knows what this information could yield?

When we speak on the phone, Chugh says that when he first began the project, people thought he was "crazy", but now the scientific community is embracing the potential for big data to uncover hidden medical conditions and identify common patterns, and this kind of research is expanding. He has launched a second such study in California, and large-scale registries of sudden cardiac arrest have been created in several other parts of the world, including Europe and Taiwan.

In 2017, the EU launched a new initiative, known as European Reference Networks, to share information about rare diseases. One of its projects is ERN GUARD-heart, a collaboration between 24 specialist centres from 12 European countries to share information about rare heart diseases, including unexplained cardiac arrests (known in the medical community as idiopathic ventricular fibrillation). The group is headed by Wilde, the University of Amsterdam cardiologist. It too aims to expand databases of patients with unusual heart conditions, in order to make it possible to spot common trends or genetic causes.

While even big national research centres may only have access to 15 or 20 patients who have suffered an unexplained cardiac arrest, Wilde hopes eventually to build up the database to include a few thousand such patients from around the world. (One challenge he now faces is maintaining the UK's involvement, since after Brexit it will no longer be eligible for EU funding for such initiatives.)

Such databases offer our best hope of understanding cardiac arrest, but Wilde tells me that he isn't "optimistic that we are going to solve this very soon". He thinks it unlikely that scientists will uncover some new genetic disease or any single underlying cause. Chugh, too, believes that the future of our understanding of sudden cardiac arrests depends on our ability to identify and understand a whole range of risk factors acting in combination, some physical, some environmental, some genetic.

One way of thinking about it, Wilde suggests, is to consider a freak accident that sometimes strikes during a baseball or hockey match. Players have died because a hard ball or a puck hit them in the chest and stopped their heart. Had the object struck a millisecond or two later or earlier the event would have been nothing more than a painful inconvenience, but there is a tiny window during each heart cycle when it is especially vulnerable, and an external blow to the chest can trigger fatal ventricular fibrillation. "That's a very rare condition," Wilde says, "but given that this can happen, you can imagine other factors that are unknown as yet, that can lead to the same combination of things, the same problem."

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I wondered what drew Wilde to study a problem that seems so insoluble, and he points to two things. First, he notes that in the past 20 years the proportion of cardiac arrests that remain unexplained has shrunk significantly, thanks in large part to advances in genetics. Second, even small findings can have a remarkable impact on individuals.

He describes how one patient was referred to him because several of his relatives had died of unexplained cardiac arrests. This troubling history suggested that the problem was genetic. The Amsterdam group eventually uncovered the genetic cause and used the information to implant several other family members with preemptive internal defibrillators. Several of them subsequently suffered a potentially fatal arrhythmia that was treated by the ICD, saving their lives.

These decisions are seldom easy, however. Chugh describes agonising over whether to implant an ICD into a child whose identical twin had suffered a fatal cardiac arrest. If the dead twin's cardiac arrest was genetic in origin, the surviving twin would also be at risk – but the heart surgery could prove medically unnecessary. (Patient confidentiality means he couldn't disclose what decision was reached.)

Another thorny issue, given our patchy knowledge of what causes cardiac arrest, is whether people should be routinely screened for the conditions we know about. The public health service offers screening to anyone with a relative who has suffered a cardiac arrest aged 35 or younger, and Wilde also suggested that anyone

who faints while exercising should consult a cardiologist, as this could be evidence of an inherited cardiac arrhythmia.

But should young people with no symptoms or family history of cardiac arrest be screened? The British charity Cardiac Risk in the Young thinks they should. It screens more than 20,000 young people a year for free, using a health questionnaire, an ECG and sometimes an ultrasound scan of the heart. One study has suggested that when Italy introduced mandatory heart screening for athletes, rates of sudden cardiac death among them dropped by 90 per cent.

The National Health Service does not recommend population-level screening, however, in part because the anxiety caused by false positives – results that wrongly indicate a condition is present – could put people off exercising and damage their health. False negatives pose a problem, too. Had my friend Clare been screened, it seems nothing untoward would have been detected.

Clare's near-death experience did not involve bright tunnels or flashbacks or otherworldly encounters with dead relatives or spiritual beings, just the sudden dark of lost consciousness followed by the halfremembered haze of her first few days in hospital. Now 33, she is not the kind of person to commemorate a "second birthday". A few times she referred to the incident by text using the skull-andcrossbones emoji, and when I told her I wasn't ready for that kind of flippancy she laughed but apologised.

The uncertainty around why her heart stopped still haunts her, however – in part because she wants to be absolutely certain her daughter is not at risk, but also because she is a scientist, seeking explanations. She is participating in the UK's 100,000 Genomes Project, which is sequencing the genomes of people suffering from rare diseases and cancer, in order to aid future research. One genetic screening for known heart conditions revealed that she has a genetic variant of "unknown clinical significance", and so Clare regularly scans Google Scholar for any new research into the mutation, sometimes discussing her findings with her cardiologist and her brother, a biochemist.

Mostly though, she'd prefer not to think too much about the day her heart stopped. "I feel like there's no point surviving if you're just going to live your life framed by the fact that you nearly died, and I think there's no point worrying about dying," she says. In the weeks after her collapse she felt she had a new sense of perspective, a clarity over what mattered to her. But soon after she felt drawn back into the rhythm of everyday life, its simple pleasures and trivial annoyances. "I don't think I can say I'm a totally changed person, or that I live my life totally differently because of what happened," she says. "Because actually things have just gone back to – well, I don't think normal is the right word – but life goes on."

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