

I'm glad I was a guinea pig!

Marfan's syndrome meant Emma Barker, 25, could die at any moment. Then along came a new invention...

**W**alking into the classroom at Archer's Court Secondary School, Dover, Kent, I heard a voice shout: 'Oi, milk-bottle eyes! Nice glasses!' Like I haven't heard that before, I wanted to scream. That's what my sister Helen would have done if the taunt had been targeted at her. But I wasn't as confident as my little sis. Instead, I put my head down and hoped the ground would open up and swallow me. My classmate was right about my specs, though. The heavy pink frames and thick lenses were awful. Without them, though, I couldn't see a thing. I'd been wearing glasses since the age of five, when the lenses in my eyes had been removed after the retinas had grown too big and snapped. Helen, a year younger than me, had also undergone surgery, our bad vision a symptom of a shared genetic condition — Marfan's syndrome. I didn't know much about Marfan's back then, except that our dad, Peter, had it too. Once a year, the three of us visited a doctor, Anne Child, at St George's Hospital, Tooting, London. She'd examine us and we'd have a full body scan. It was only when I turned 14, in 1999, that I discovered what having Marfan's really meant. 'I need you to stop doing cross-country running,' Dr Child told me. 'And trampolining.' I was horrified — I loved running at school. 'Why?' I asked. For the first time, I learnt how the

Me, aged 10, with my sister Helen. Inset: Tal Golesworthy who invented the life-saving valve repair kit

# They wrapped my heart IN A BANDAGE

gene defect could cause abnormal growth of bones, including fingers and toes, damage to eyesight and weakness of connective tissue. Worst of all, it could result in the aorta — the main artery of the heart — splitting, causing instant death. At first I was

very scared. Then I looked over at Dad. He was OK and lived a completely normal life. *Nothing's going to happen to me, I decided.* I was more concerned about getting rid of my hated glasses than I was about dying, and was ecstatic when, a few months later,



Me, with Helen, in the glasses I loathed



Mum, me and Helen when I was nine



At 12, I was tall and got teased as school



I got contact lenses.

My confidence soared and I started to enjoy being a teenager. I didn't even mind being teased about being one of the tallest people in my class, another sign of Marfan's syndrome. 'What's the weather like up there?' the other kids would ask. 'Lovely, thanks,' I'd reply with a smile. After leaving school, I did media studies at South Kent College, Dover. At 18, I went to work as a stewardess on the ferries. Apart from my annual trip to St George's Hospital, Marfan's didn't factor in my life. At each appointment, Dr Child would measure the valve in my, Helen's and Dad's hearts to make sure they weren't enlarged. In 2004, Dr Child discovered that my valve was almost the same measurement as Dad's. 'In fact, your dad's is a little smaller,' she told me. 'What does that mean?' I asked

her, shocked. 'It means you both need surgery.' A feeling of dread crept over me. Dad's brother, Martin, had already undergone an operation during which his natural valve had been replaced by a tin one. As a result, he would have to take a blood-thinning medication, Warfarin, for the rest of his life. It left him feeling permanently tired and ill. 'He's got no quality of life,' I told Dad. 'I don't think I can do it.' 'I don't think I can, either,' Dad agreed. Our decision not to have the op, however, weighed heavily on our minds. We knew our hearts could fail at any moment. It was my mum, Christine, who kept our spirits up and supported us. Then, just before my 20th birthday, I met Paul, then 48. Like me he worked on the ferries and we shared a love of Caribbean food, reading and taking long walks. He reminded me that there was life beyond heart problems and hospitals. Maybe that's why I didn't tell him about Marfan's. It already dominated so many of my thoughts, I wanted to keep it away from our relationship. *I'll tell Paul when I need to, I decided.* Having passed Marfan on to his two children, Uncle Martin was determined to keep up to date with all the latest research into the syndrome. In 2004, he learnt of Tal Golesworthy, a Marfan's sufferer and engineer, who'd made his own aorta repair kit. 'Instead of replacing the valve, he made a polyester sleeve to go around it and stop it splitting,' Uncle Martin told Dad. 'That sounds like a brilliant idea,' Dad said. Eager to learn more, the brothers went to a seminar to listen to Tal Golesworthy explain his invention. Dad returned home impressed. 'I want to have it done,' he told us — and immediately e-mailed Professor Pepper at the Royal Brompton Hospital & Harefield NHS Trust, who'd carried out the procedure on Tal. When the professor agreed that Dad could have the surgery, I was eager to join him. 'Let me go through it first,' Dad said. 'If everything's fine, you can go ahead too.' In June 2004, Mum and Dad travelled to the Royal Brompton for the surgery. It was a worrying time, but Mum rang us every day with a progress report. Immediately after the surgery, Dad was in intensive care. Then he was moved to a high-dependency ward. 'Your dad's in really good spirits,'

you all the way,' Paul said. 'It is,' I told him. If I didn't, my heart would, quite literally, break. That didn't mean I wasn't afraid, however. Although Dad had seemingly sailed through his experience, for me, the op was still a leap into the unknown. *What if I don't wake up?* I thought. Still, I knew it was a chance I had to take. Not just for my future, but for Helen's. I went for an MRI and a special bespoke sleeve, like a sort of bandage, was made for my aorta. A few weeks later, in November 2005, I travelled to London for surgery. Paul wasn't with me — I knew I'd get emotional and didn't want him to see me like that. Instead, Mum was at my bedside at the Royal Brompton. She put up with me lashing out after the op, in pain and afraid, and consoled me when all the morphine I was taking reduced me to tears. 'I want to go home,' I sobbed. Despite suffering a slight allergic reaction to a blood transfusion, I was on my feet and walking around the ward the day after surgery. 'Make sure you tell Dad I beat his record!' I laughed to Mum. When I got home, however, I had a bone to pick with Dad. 'You said the surgery didn't hurt!' I told him. 'You lied!' 'I didn't want to scare you,' he said. When Helen, who by then was considering having the op, asked me about the pain, I did



I told my partner Paul about my condition, now we're busy building a future together



L-r: my aunt Valerie, uncle Martin and Dad



Helen and I are lucky to have great support around us. L-r: Helen's boyfriend Darren, Helen, Dad, me, Paul and my nephew Caide

Mum told us. In fact, just two days after the op, he was up and walking. After five days in hospital, Dad came home and was his usual, happy, self. 'Was it very painful?' I asked him. 'Not at all,' he reassured me. 'So is it my go now?' Satisfied that I would come to no harm, both Mum and Dad agreed that I could go ahead. But there was one other person I had to tell — Paul. We'd been together less than six months, but I had no choice. I had to explain about Marfan's and the operation. He listened quietly. 'So what do you think?' I asked. 'If it's what you want, I'll support

just as Dad had done. 'You barely feel a thing,' I told her. In fact, it wasn't until January 2006 that I started to feel like myself again, and even then I was exhausted. It was another six months until I could return to work. Later that year, Paul and I opened a restaurant together, Hudson's. It was our baby, especially as we'd decided not to have any of our own. The 50/50 chance I could pass on Marfan's was just too big a risk to take. Paul, now 53, already has four children from a previous relationship, who often visit us at our home in Oxford Avenue, Newtown, Southampton, and that's enough for me. Helen, however, went ahead with the operation after me and is hoping to start a family of her own. Paul and I sold the restaurant in January 2010 and, inspired by how Tal Golesworthy made a difference to my life, I decided I wanted to do the same. Now I'm working with adults who have autism and learning difficulties. For so long, Marfan's was a shadow over my life. Now, I'm determined to make every second count.

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I live life to the full these days