



Breakthroughs in children's medicine

heart and lung



More

Every year, we need to raise £50 million to help rebuild and refurbish Great Ormond Street Hospital, buy vital equipment and fund pioneering research. As well as the developments in this guide, amazing things happen at Great Ormond Street Hospital every day. With your help we can keep the magic alive for our very ill children and their families.

Our website has more information about the specialists, patients and treatments you've read about in this guide, as well as the pioneering research that happens in the hospital.

If you'd like to find out more, or you have your own stories that you'd like to share with us, please visit gosh.org/breakthroughs



Keep the magic alive



Welcome

From when it first opened as a ten-bed ward on 14 February 1852, Great Ormond Street Hospital has always been a very special and crucial place. Over the years, breakthroughs both big and small offer a chance of life to some very sick children.

This guide takes you through some of the dramatic milestones – from the very first heart-lung bypass machine for open-heart surgery in children, to pioneering research into how new stem cell treatments might be a lifeline for those with heart failure.

Ours is a story of world-class doctors, surgeons, nurses and researchers, whose vision for the sick children in our care has helped us treat the untreatable. The passion we feel drives us to go the extra mile for the children

in our care and through research we aspire to help children we will never ourselves meet. Both help to explain why we are one of the foremost children's hospitals and research facilities worldwide.

But we would be nothing without our patients and their families. Their bravery is truly inspirational and is the 'fuel' that energises our staff. There's always more we can do to help them. It's why we keep striving to give the children the best possible chance to lead happy, healthy lives.

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Yesterday ❤️

How it all began

On Valentine's Day in 1852, Britain's first children's hospital opened on Great Ormond Street. One of the first in-patients was three-year-old Eliza Armstrong, who was suffering from consumption (pulmonary tuberculosis).

Medicine was very different back then. Cures were in short supply, with doctors treating patients as best they could given the limited resources. As a result, little Eliza was given rest and a diet that would have included milk, beef tea and even wine. It must have worked, as after three weeks she was sent home to attend the hospital as an out-patient. Records show her condition had been 'relieved'.

Other children with severe heart and lung conditions weren't so lucky; life was grim and often short. Medical knowledge hadn't progressed to the point where doctors specialised very much, and there was a lack of reliable surgical techniques. In fact, for the first 100 years of Great Ormond Street Hospital's existence, little could be done for children with a serious heart and lung condition. The doctors did what they could to lessen the suffering, but tragically, the vast majority of their patients died very young.



1852 – The hospital opens, but few children with heart and lung conditions survive infancy.



Doctors did what they could with limited knowledge and resources



Mr David Waterson

A pioneering partnership

It wasn't until 1947 that the first glimmers of hope came for children suffering from heart and lung conditions, thanks to the efforts of two pioneering individuals.

Mr David Waterson was one of the hospital's leading paediatric surgeons, who saw that children with heart and lung problems were in beds all over the hospital. Shouldn't they be together, receiving specialist care and attention? Perhaps then a vision of the future might emerge.

So he formed a partnership with Dr Richard Bonham-Carter, a brilliant but unassuming cardiologist. They set up the heart and lung unit; the first dedicated place in the UK where

surgeons and doctors could collaborate to diagnose and treat children with heart and lung disease. Their type of work was often referred to as 'fixing the plumbing', so the new team approach meant it wasn't long before they were affectionately known around the hospital as 'the plumber and his mate'.

It may have started as just a ten-bed ward, but the heart and lung unit was proof of the two men's unique vision: that by combining disciplines and skills, treatment could be much more effective. The model was far ahead of its time, and set the precedent for how we still work today.



1947 – The hospital's heart and lung unit is launched: the first of its kind in the UK.



1954 – Catheters allow more precise diagnosis of heart and lung conditions. The first non-invasive heart procedure comes soon after.



Dr Richard Bonham-Carter

Early surgery and an early

breakthrough

When the unit first opened, heart and lung problems were usually diagnosed using a stethoscope. If that was inconclusive, the traumatic alternative was for patients to have their chests opened in surgery to determine what was wrong with them. The tragic reality was that three out of every four children admitted to the hospital's new heart and lung unit didn't survive.

A big leap forward came in 1954, when Dr Gerald Graham – another heart specialist – came to work with Dr Bonham-Carter to launch a cardiac catheter laboratory. By feeding thin tubes through blood vessels in the leg, doctors could use these 'catheters' to measure blood pressure in the

chambers of the heart. Now they could better diagnose problems without opening the chest, and even perform non-invasive surgery on some heart conditions, using the catheter as a tiny tool.

These new procedures helped many infants live into childhood. But keeping them alive for more than just a few years was a tougher proposition. To repair more complex heart and lung conditions, surgeons would need something that could keep young patients alive while they carried out challenging operations. Something that would keep the body working while the 'plumber' did his surgical work inside the heart.



1962 – The hospital pioneers the first heart and lung bypass machine for children, to help repair heart problems.

Heart and lung surgery was very different in the 1960s

How a little machine had a

By the mid-1950s, the heart and lung bypass machine had made open-heart surgery possible in adults. By pumping oxygenated blood round the body, it did the work of a patient's heart and lungs while surgeons worked on the heart. But adult machines couldn't be used on little children, because they removed too much blood from the child as they tried to pump it round. Considering how sick the children were, it was all too risky.

Until then, the hospital could only offer basic operations that at best relieved some of the suffering of the children on the heart and lung unit. But David Waterson wanted to take things further, so in 1957 he set up the heart and lung unit's first research programme. Funded

with a charitable donation of £25,000, its aim was to design a bypass machine that could be used in babies and infants, to give children born with severe heart and lung conditions the chance of a longer life.

After five years of painstaking design and testing, the first ever paediatric heart and lung bypass machine was ready to use. A year later five children's lives had been transformed, thanks to surgery which tackled their previously-untreatable heart conditions. By 1964, more than 50 children a year were benefitting from the bypass machine, and by 1967, 60 per cent of infants with severe heart and lung problems were surviving – thanks to surgery that would have been unthinkable less than a decade earlier.



1964 – One patient a week is admitted for life-saving surgery on the bypass machine.

huge impact



Image: Alan Knight, 1967

An early heart and lung bypass machine

Richard and Carol, the 'blue babies'



Richard and Carol's surgery was big news

Richard Carey and Carol Barclay were among the first children to have life-saving open-heart surgery at Great Ormond Street Hospital. They're both now in their forties; but without improvements led by the surgeons here, things might have been very different.

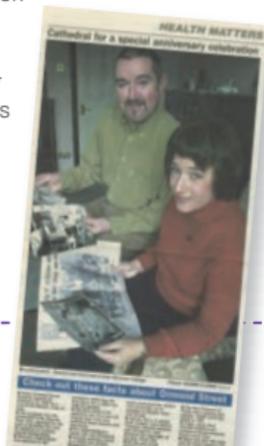
They were born with transposition of the great arteries, a condition which affects one in 5,000 babies. It means the heart's main blood vessels come off the 'wrong' pumping chambers, so only blue blood (which hasn't been oxygenated) circulates round the body. Babies born with this condition have a blue skin tone, but successful surgery means they literally change from blue to pink right there on the operating table.

Before the bypass machine enabled him to undergo surgery, Richard didn't just look blue. His condition meant he couldn't walk, and had spent much of his life stuck in an

oxygen tent. But after the operation he really made up for lost time, playing football and running cross-country.

Carol was four when she had surgery, having survived up to this point thanks to a hole in her heart that allowed red (oxygenated) and blue blood to mix. Luckily for Carol, this hole occurred naturally. However, without further surgery, the best that Carol's parents could have hoped for was that she lived to her teens.

Mr Waterson's operation fully corrected the heart's plumbing and changed her life – her mother still remembers Carol's fingernails being pink for the first time when she returned from the operating theatre.



1967 – 60 per cent of children admitted to the heart and lung unit now survive.

♥ Today

New techniques mean a new era

By the 1980s, specialist techniques and equipment such as catheters and the bypass machine had revolutionised the way we treated heart and lung conditions. Most patients were now surviving surgery in early childhood and many looked set to enter adulthood. Never before had children with congenital heart disease prepared to face adult life along with their friends.

The task for the team now was to make sure their operations were setting up patients well into their adult lives. Heart surgery at the hospital was led by Professor Jarda Stark and Professor Marc de Leval, who collaborated with specialists across Europe to find better ways to treat children with transposition – like Richard and Carol.

In 1986 a revolutionary new surgical technique called the 'switch' was adopted. The transposed arteries were disconnected and literally 'switched' over. The tiny coronary arteries also had to be switched, and the operation performed within a couple of weeks of birth – all amounting to a huge technical challenge.

As with the original bypass operations in the 1960s, there was a steep learning curve. But within a couple of years, switch surgery became standard. The knock-on effect was that instead of being a fatal condition, children with transposition could now expect to live for an average of 62 years.

In 1988, Professor de Leval set up the transplant unit, thanks to a £200,000 fundraising appeal. It was one of the first centres in the UK to carry out life-saving transplants on children with heart failure.

Chances of survival for babies and small children born with cardiomyopathy (disease of the heart muscle) are very poor, so a transplant can be a lifeline. Today the programme is one of the largest in the world, performing around 30 heart and lung transplants a year.



1971 – Survival rates for heart and lung conditions increase to 80 per cent.



1974 – Echocardiogram technology allows surgeons to capture images of the heart in real-time so they can diagnose heart disease in children without performing surgery.

Professor de Leval at work



Meet some patients

Hannah's extraordinary story

Hannah is a remarkable girl. She suffered heart failure as a baby, and was only three when a donor heart was grafted onto her diseased heart at Harefield Hospital. It's a technique called 'piggybacking', where the new organ supports the original.

But it wasn't easy. The immunosuppressant medicines Hannah needed so her body wouldn't reject the donor heart had a terrible side effect; they caused cancer. Hannah needed chemotherapy and yet more drugs to fight it, while the immunosuppressants also had to be greatly reduced.

But this reduction meant that Hannah's body started to reject the donor heart. At one point, she was even rushed back to Great Ormond Street Hospital suffering from seizures, because of a cancerous growth pressing on her spinal cord. Thankfully, the cancer was eventually eradicated.

Ten years after the original piggybacking operation, in an incredible – and unique – turn of events, Hannah's donor heart was removed at Great Ormond Street Hospital because her own heart had recovered sufficiently to work on its own.

Hannah today

“I've got a normal life just like all my friends. I've just done my GCSEs, and I've now got a Saturday job looking after animals, which I couldn't have done before. I'm really glad that I don't have to rely on life-saving drugs anymore.”

Image: Felix Clay/Guardian News and Media Ltd 2009.

Tineke: from patient to expert

Tineke was born with two holes in her heart, a defect which developed as she was growing up into a rare condition called Eisenmenger syndrome. This eventually caused her lungs to fail, and at 16 she finally became so ill that she was listed for a transplant.

“It was the only hope I had of any kind of life,” she said. “I can still remember being on the cardiac ward and hearing my parents' cries of anguish coming down a corridor as the doctors told them I had only a few months left.”

Her heart and lung transplant was such a success that Tineke could go on to complete a degree in medical physics and a PhD at Addenbrooke's Hospital, itself one of the UK's pioneering transplant hospitals. But disaster struck again, as

she'd inherited a congenital kidney disease from her father. Thankfully, she received a replacement from her mother, and her career is back on track.



Tineke on her wedding day

Tineke was only the third person to have a heart and lung transplant at Great Ormond Street Hospital, and is now in her 21st post-transplant year.



1980s – The life expectancy of children born with transposition rises by an average of 46 years.

When Anthony met ECMO

16-year-old Anthony had just returned from a school trip to Disneyland Paris when he collapsed and was rushed to Great Ormond Street Hospital. When he woke, he didn't know where he was. Thankfully, his mother was standing over him – she told him he was in hospital with pneumonia before he lost consciousness again.

But Anthony had three other infections too, and over the seven weeks he spent in the hospital, his heart stopped a terrifying twelve times.

Fortunately, the hospital had adopted the revolutionary new ECMO (Extra Corporeal Membrane Oxygenation) machine. Similar to a bypass machine, ECMO acts as a child's artificial heart and lungs if theirs are failing while they wait for a transplant or further treatment. This new technology kept Anthony alive through his ordeal – his nurses called him 'the big miracle'.

Since 1992, ECMO has kept over 400 children alive against all the odds.



1983 – Over 1,000 children a year are admitted following an assessment by echocardiography.



Anthony didn't just meet ECMO – but also his hero, musician Pharrell Williams

Heart repairs... without surgery?

Open-heart surgery is invasive and traumatic, whatever your age. But since 2001, thanks to a radical technique introduced to the hospital by Professor Philipp Bonhoeffer, we've been able to replace defective heart valves without resorting to surgery to open a child's chest.

The new valve is placed inside a 'stent' – an ultra-thin, collapsible, platinum mesh cylinder – then mounted on a tiny, deflated balloon on the end of a catheter. It's then placed in a blood vessel and pushed into place in the heart. As the balloon inflates, the stent and valve expand to the required width. Then the balloon is deflated and the catheter can be withdrawn, leaving the valve in place.

This technique has many more potential uses, but most importantly, it eliminates the need for open-heart surgery. Valves may need to be replaced as patients grow up, but it's hoped that they can undergo this procedure three or four times before they need more invasive surgery.

Patients undergoing this treatment have usually already had open-heart surgery at least once. Each time their chest is opened, operations are trickier and recovery takes longer. By avoiding all that, children can leave hospital within 24 hours and avoid intensive care. Since it was introduced, more than 200 children have been treated at the hospital, with 100 per cent survival at the time of the procedure. We're proud of the breakthrough, but we're even prouder of the statistic.

Professor Bonhoeffer's technique uses a 'stent' to put new heart valves in place



1986 – The new 'switch' operation is introduced; we estimate that children born with transpositions now have a life expectancy of up to 62 years.

Breathing easy

Imagine trying to run a 100-metre sprint and then breathing through a straw. It wouldn't be easy, would it? But for children born with very narrow windpipes, every single breath is that difficult. So the same year that the non-invasive technique to replace heart valves was introduced, Professor Martin Elliott launched a tracheal service to save the lives of children born with this condition.

Operating on tracheas is difficult and not always successful. So true to the spirit of David Waterson and Richard Bonham-Carter, the approach we took was multi-disciplinary. The team

included specialists in ENT (ear, nose and throat), interventional radiology, intensive care, physiotherapy, and heart surgery.

They developed a surgical technique which involves making cuts into the narrow part of the windpipe, then sliding the two sections over each other until they reach a part with normal width. Since its launch in 2000, the team has initiated an international training programme for hospitals around the world. Our tracheal service has become one of the largest and most successful in Europe – and a world leader in its field.



Today 

Professor Martin Elliott launched the hospital's tracheal service



1988 – Marc de Leval sets up the transplant unit.



2000 – The hospital launches its programme to replace heart valves without open-heart surgery.

Harvey's Berlin Heart

As heart transplants have become a more effective way to treat children with serious heart conditions, the waiting lists have grown too. The sad fact is that up to a fifth of all children who are heart transplant candidates die while waiting for a suitable donor organ.

So in 2004, a portable new machine was introduced which keeps children alive while they wait for a heart donor. It's called the Berlin Heart and, like an ECMO machine, it helps pump blood around the body. And because it's so compact, the patient isn't confined to bed and can move around.

Harvey is one of our Berlin Heart record-breakers. He has a rare metabolic condition, and at just six months old a scan showed that he might need a heart transplant. By the time he was 15 months

old his condition had drastically declined, and he had difficulty breathing. He was taken to Great Ormond Street Hospital and placed on an ECMO machine in the hope his heart would recover. But two weeks later his heart was still failing and a transplant seemed his only option. The team decided to trade his ECMO circuit for two Berlin Hearts to pump the blood around his body so he could be a proper toddler while he waited.

The Berlin Hearts kept Harvey alive for an incredible 167 days, after which a donor heart finally became available. Within five hours he was undergoing transplant surgery; removing the two Berlin Hearts and introducing the new one took nine hours. He's now doing remarkably well; enjoying being outdoors like any other toddler, and really loves playing football.



2003 – The hospital starts an electrophysiology programme to electrically stimulate the heart and cure children with irregular heartbeats (arrhythmias).



Harvey, aged four, in his favourite football kit

Saving Sarah

Sarah was only six days old when she became lethargic and stopped eating. She had viral meningitis, and the virus went on to attack her heart. Her condition was serious.

She was rushed to the cardiac intensive care unit at Great Ormond Street Hospital, but after a few days the team felt she only had 24-48 hours to live. So for the first time since little Sarah arrived at Great Ormond Street Hospital, her mother, Jennie, was

allowed to pick up her daughter and cuddle her.

Miraculously, a donor heart was found the next day, a very rare occurrence for someone so young. So at just a few weeks old, Sarah became one of the UK's youngest ever heart transplant patients. Now she's the youngest surviving one.

She's three years old now, and still comes into the hospital for follow-up treatment. She loves everyone here, and they love her back.



Sarah gives her teddy care too



2005 – The hospital performs its 100th lung and 200th heart transplant.

Meet the team

None of the breakthroughs – or indeed the care – we’ve told you about would be possible without the people who work in our heart and lung unit. As one doctor says, “Working here is like working for the United Nations. People travel from all over the world to work here.”



“ Nobody wants heart surgery, but if something goes wrong with your heart valve, you might want to replace it. Rather than us opening it up, changing it, closing it up, and spending a week in recovery, you come in, we feed a catheter into the heart, blow up something that fixes the valve, come out, and you can go home. The research isn’t about ‘is it a good idea’, it’s ‘what can we do to make it happen?’ **Here at Great Ormond Street Hospital, we often say ‘if I don’t know, there’s someone down the corridor who does’.**”

Martin Elliott, chairman of cardiac services



“ I’m inspired by the people that work here; there are so many world leaders across the hospital. But the real key is the range, severity and volume of heart disease that’s referred here – it’s quite unique, both in the UK and internationally. **I feel that I am working at the leading edge of paediatric medicine, and that has to be helpful to the children we treat.**”

Mike Burch, paediatric cardiologist and director of cardiothoracic transplantation

“ I first encountered children with cystic fibrosis nearly 30 years ago. **I was always impressed with their courage to live life to the full, in spite of severe disease.** At that time, it was considered lethal. There’s still no cure but current therapies have changed the outcomes significantly, and now many children with cystic fibrosis live into adulthood – although not without arduous treatment programmes. I hope the breakthroughs we make now will benefit children with cystic fibrosis for generations to come. ”

Colin Wallis, consultant respiratory paediatrician



2009 – Approximately 30 heart and lung transplants are carried out each year.

♥ Tomorrow





Giving today's children

The breakthroughs we've outlined came about as the result of years of painstaking research. As we move into the future, that desire for progress still fuels our work every day. The passionate individuals here are constantly refining the techniques we use, developing new ones, and improving the ways we care for children whose problems still thwart

us. But we're determined they won't thwart us for long.

We have been working in partnership with the UCL Institute of Child Health since its foundation in 1946, and together we are the UK's leading centre of excellence in research into childhood illnesses. When the new heart and lung unit opens in 2012, the spirit of

a brighter future

innovation – envisaged by Mr Waterson and Dr Bonham-Carter – will take us to an amazing new level of childcare. Thanks, of course, to the generosity of our supporters.

The work we do covers everything from pre-natal problems to the beginning of adulthood. And because we have a dedicated clinical research facility in the hospital, children can benefit from

research that is fully integrated into their treatment, in a way that's designed to minimise stress and keep them happy.

Most importantly, by sharing our findings with other leading hospitals, our research benefits children around the world. When you consider that childhood problems determine adult health too, it's an incredible responsibility. That's why we're here.

Could ten-pin bowling beat cystic fibrosis?

Cystic fibrosis is the most common genetically-inherited disorder in the UK. A steady decline in lung function is just one of the symptoms that cause premature death in affected children – so stopping that decay is vital if we're to help them grow up and lead healthy adult lives.

Over the years we've led the way in finding ultra-sensitive ways to measure lung function in infants, and detect early lung disease before other symptoms become visible. This has led to a unique programme to monitor and treat children with cystic fibrosis from birth, generously funded by supporters of Great Ormond Street Hospital.

It's led by Professor Janet Stocks and Dr Colin Wallis. But where does bowling come into it?

"Until now, it's been hard to get children under six to co-operate with tests to measure their lung volumes" says Dr Wallis, "but hook the output of the test up to a virtual ten-pin bowling game and it's hard to get them to stop!"

As yet, there is no cure for cystic fibrosis, but Dr Wallis's programme will help us intervene in the crucial early years, so these children can enjoy productive adult lives. "We need to pick up the differences our treatments are making before the disease takes hold. Waiting for these children to become unwell is waiting too long."



Ten-pin bowling helps us measure lung volumes



The right surgery at the right time

Many 'blue babies' have a condition called tetralogy of Fallot, another congenital heart condition which reduces the heart's ability to circulate oxygenated blood. When their apparently normal baby suddenly turns blue and floppy for no apparent reason, parents are understandably terrified.

Since the 1960s, the team has operated on more than a thousand children born with this condition. These days, the great majority experience a normal childhood, but their baby operations leave a leaky valve on the right side of their hearts. What we don't currently know

is whether it's worth putting in a new valve as an investment for their future.

At present, it's very difficult to know the best time to offer any new valve implantations. Too soon and the child may outgrow the valves; too late and the heart muscle may get too tired with years of extra work. By examining different patients across the whole age spectrum between one and 50, Dr Kate Bull is

determined to find the optimum times to perform surgery.

Dr Bull's team is also working with experts who are looking at ways to avoid the need for surgery altogether. In addition, they're putting together a DVD of patients' experiences. Learning from people who've lived through heart disease themselves will help patients come to terms with their own troubles.

“ I was born in 1955, and my first operation at Great Ormond Street Hospital was when I was just 18 months old. I had another one at the age of nine as my health was deteriorating fast.

I have treasured memories of my time there and despite being born with heart problems I have no regrets. I'm glad to be involved in Dr Bull's study, as I can never repay Great Ormond Street Hospital for what they have done for me. ”

Steve Weston, former patient

Dr Bull with Steve Weston, our 14th patient to have surgery to repair tetralogy of Fallot.

Turning virtual hearts into real improvements

When the hospital launched its programme to replace faulty heart valves without open-heart surgery (see p22-23), it changed the lives of hundreds of patients. But as everyone's heart is unique, the technique can only be used on 15 per cent of the children who need our help.

Now, Professor Andrew Taylor and his team are using the latest cardiac

imaging and computer modelling to potentially remove the need for open-heart surgery for thousands more patients every year.

"Advances in computer modelling mean we can design and test implants virtually", says Professor Taylor. By using MRI scans to create 3D models of patients' hearts, his team can build and test bespoke valves before they're implanted. The use of these devices in the future could prevent up to 10,000 patients a year around the world from having open-heart surgery.

As we move into tomorrow, Professor Taylor's team are taking another pioneering step into the unknown, using CT scans to monitor the valves after they've been inserted. "Every patient is different, and we're taking huge steps to make sure their treatment is tailored precisely to their needs."



Professor Taylor, and a 3D model of one of his virtual hearts

No more transplants?

Thanks to the specialist cardiac surgeons and the transplant team here at Great Ormond Street Hospital, nearly 400 children with heart failure have been given the chance of life. But for children in need of a transplant, both time and donor hearts are in short supply. What if that need could be removed?

As we've shown earlier in this guide, devices such as Berlin Hearts can keep children alive while they wait for a transplant, but Dr Mike Burch and his team have a longer term hope. They're exploring how using a patient's own stem cells could help the heart heal itself while a donor is being looked for. And one day, they hope to remove the need for transplants altogether.

As you can see, everything our specialists are doing for tomorrow is based on the same principles that inspired Mr David Waterson and Dr Richard Bonham-Carter all those years ago. By using our unique skills to work together, we can help more children than ever before. And as technology develops and our knowledge increases, who knows what is possible?



By developing more breakthrough treatments, we can help thousands more children like Scott and Harvey.