

Find out more

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

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**

To continue the legacy of breakthroughs at Great Ormond Street Hospital, we need to raise £50 million every year. This helps to rebuild and refurbish the 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.

©2007 Great Ormond Street Hospital Children's Charity. Registered charity no. 235825.



Great Ormond Street Hospital Charity

Breakthroughs in children's medicine

Neuroscience



Welcome

Welcome to our breakthroughs in children's medicine neuroscience guide. Great Ormond Street Hospital was the first of its kind in the UK to have a paediatric neurosurgery department, and has grown from a unit with a single surgeon to one in which we now operate on over 800 cases per year.

As technology has improved, what was once an area of fear and uncertainty is now one of confidence and understanding. Most of our patients are treated in multi-disciplinary teams and the depth of experience we can draw upon for the benefit of our patients is unparalleled. The department of neurosurgery has led the way in many different surgical fields but is particularly prominent in craniofacial, epilepsy and complex spinal surgery. We are constantly evolving new surgical techniques to improve patients lives.

We know we can always get better at what we do, so collaboration with our colleagues in the UCL Institute of Child Health is a crucial part of our work. By bringing scientific research to the bedside we're able to offer our patients and their families constant clinical improvement.

I hope you enjoy hearing about our department's past, present and future and that it will inspire you to support our work. Our children deserve to get better!



Mr William Harkness,
Consultant Neurosurgeon

Contents

Yesterday

Charles West's vision to transform children's medicine page 9
A time of little relief from neurological disease page 11
Pioneering child-friendly equipment page 12
The UK's first children's neuroscience unit page 14
The parent, the engineer and the pioneering paediatrician page 16
My grandfather the inventor by Sophie Dahl page 19

Today

Incredible advances in visualising the living brain page 23
A touch of superglue to cure Druhan page 24
Better surgery, long-term collaboration and much-needed expansion page 26
Ground-breaking treatment to shrink Madison's brain tumour page 29
Learning from our patients and sharing our discoveries page 30
Joni's decade of up to 30 seizures an hour page 32
Meet the team page 34

Tomorrow

A brighter future page 39
Relearning how we learn page 40
Ensuring visually impaired children get the best start in life page 42
Tracing the living pathways of the brain page 44
Thank you page 46



Yesterday





Charles West, Great Ormond Street Hospital's founder.

1842 7,500 children in London die from diseases of the nervous system.

Charles West's vision to transform children's medicine

In the 19th Century sick children had little hope for the future, with only one in three surviving to the age of five. For those with neurological diseases the situation was just as bleak. In 1842, over 7,500 children in London died from diseases of the nervous system and over half were under five years old.

Back then patients with epilepsy, the most common childhood neurological disorder, were often locked away in lunatic asylums or insane wards, while other nervous diseases such as paralysis were considered untreatable.

Great Ormond Street Hospital's visionary founder, Dr Charles West, was acutely aware of the urgent need to help children suffering from neurological illnesses. In 1848, Dr West published a series of 39 landmark lectures, having meticulously observed the fate of over 14,000 children. For the next 50 years, this publication became the authoritative reference for childhood diseases. Neurological disease was such a concern to Dr West that he devoted the first 12 chapters to illnesses such as hydrocephalus (literally meaning 'water on the brain'), meningitis (an infection of the brain) and disorders of the spinal chord and muscles.

Four years later, Dr West opened the Hospital for Sick Children at Great Ormond Street, later to become Great Ormond Street Hospital. For the first time children had a place where they could receive basic medical attention, shelter and nourishment. Dr West's compassionate vision had set the scene for some remarkable breakthroughs.

1848 West publishes landmark studies of childhood disease – neurology is a key focus.



A ward from the late 1800s.

A time of little relief from neurological disease

In 1860, the National Hospital for Paralysed and Epileptic – now the National Hospital for Neuroscience and Neurosurgery (NHNN) – opened around the corner from Great Ormond Street on Queen Square. It was the first institution in the country to offer care specifically to patients with neurological diseases. In the years to come, collaboration between the two hospitals meant Great Ormond Street Hospital could apply the skills of the NHNN's doctors, surgeons and researchers into breakthroughs for its young patients. However, these early days offered only the faintest hope to most children with neurological conditions.

In 1872, five-year-old Jesse Weatherseed was admitted to Great Ormond Street Hospital with epilepsy and diabetes. His doctor's notes revealed that he:

"First had a fit at two months old. Has had them at intervals ever since, the longest interval having been about a fortnight. Convulsions generally [on] one side, and on that side [he] is paralysed. Very sharp boy between the fits – talkative."

Within ten days his condition had worsened: "Another fit this morning, after which he was much exhausted – appears to be slowly sinking." Though there were days when he "rallied somewhat", Jesse's fits increased and he developed a severe chest infection. Tragically, his doctor recorded that "...three weeks after admission [Jesse] died from exhaustion."

Jesse's case was not uncommon. Without adequate medical knowledge, children with neurological diseases faced a bleak outlook.

Pioneering child-friendly equipment

Continuing Dr Charles West's vision to advance children's medicine, over the following decades, leading doctors and surgeons from the National Hospital for Neurology and Neurosurgery offered their expertise to help Great Ormond Street Hospital's patients.

However, diagnoses and treatments were limited. Operations were hampered by the lack of effective anaesthetics, which meant children faced a tortuous experience during surgery. Poor infection control also reduced the chances of successful treatment. In these cases children like Jesse paid the price.

In 1922, Denis Browne joined Great Ormond Street Hospital. A passionate believer in developing skills to perform surgery on children with birth defects, within six years he became the UK's first children's surgeon. He recognised the fragile state of children's nervous systems and the potentially fatal risk of shocks caused by poor anaesthesia, especially in those already weakened by a neurological disease. So he set about inventing a child-friendly mechanism to deliver anaesthetics.

The resulting cylinder-shaped device became known as the 'top hat'. It allowed surgeons to deliver sufficient anaesthetic to carry out operations on the child, while crucially not covering the patient's eyes. By allowing children to see out while gently putting them to sleep, the 'top hat' prevented them from panicking, enabling thousands of patients to benefit from life-saving head and neck surgery. Tailoring medical care to children's specific needs would be the driving force for significant further breakthroughs over the years.

 1928 Denis Browne appointed as UK's first consultant paediatric surgeon.

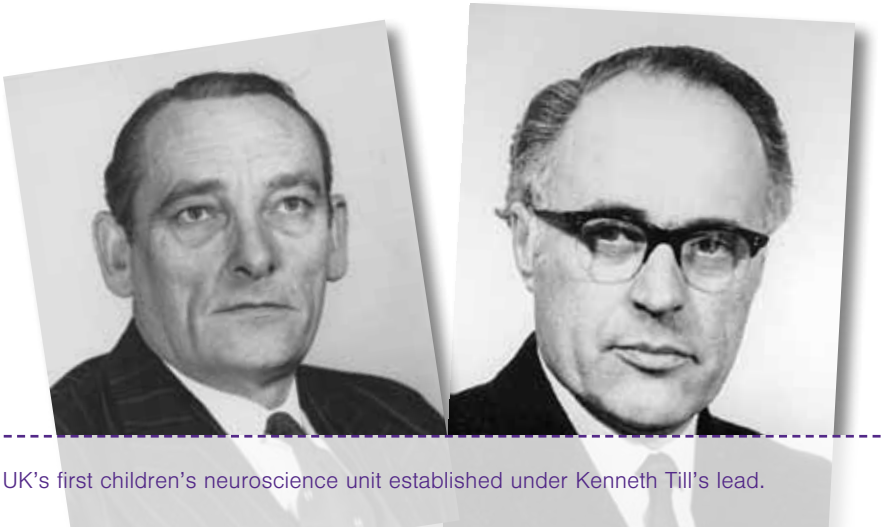


Denis Browne, regarded as the father of paediatric surgery in the UK.

The UK's first children's neuroscience unit

During the 1930s and 40s, improvements in the developing field of neurosurgery had begun to offer a lifeline to patients needing brain surgery. Recognising the critical lack of specialist care for children in this area, Wylie McKissock, one of the UK's leading neurosurgeons, introduced a young surgeon called Kenneth Till to Great Ormond Street Hospital in 1953.

Till was described by colleagues as a kind and compassionate man whose enthusiastic passion for neurosurgery was unquenchable. He recognised that children were not just small adults – they had their own special problems, and needed special treatments.



 1959 UK's first children's neuroscience unit established under Kenneth Till's lead.

Kenneth Till took the challenge of establishing paediatric neurosurgery to heart. By 1959, he had become the UK's first full-time neurosurgeon devoted to children, and established the country's first ever children's neurosurgery unit at Great Ormond Street Hospital.

Thanks to Till's commitment – until the 1970s he was always on call – children with brain tumours, lesions, severe epilepsy or hydrocephalus could receive life-saving surgical care for the first time, and the chance of a healthy childhood.

Till also became renowned for his skills in developing new surgical techniques for children with deformities of the face and skull. His lightning-fast surgery, combined with being completely ambidextrous, meant hundreds of patients with previously terminal illnesses could benefit from the specialist treatments on offer.

Great Ormond Street Hospital quickly grew into its role as the leading centre for paediatric neurosurgery. Till's dedication to pioneering new and better treatments formed the basis for a legacy of future breakthroughs.

Wyllie McKissock (far left), who mentored Kenneth Till (left), founder of the UK's first children's neurosurgery unit.

1960s One-third of children undergoing epilepsy surgery have life-threatening complications.

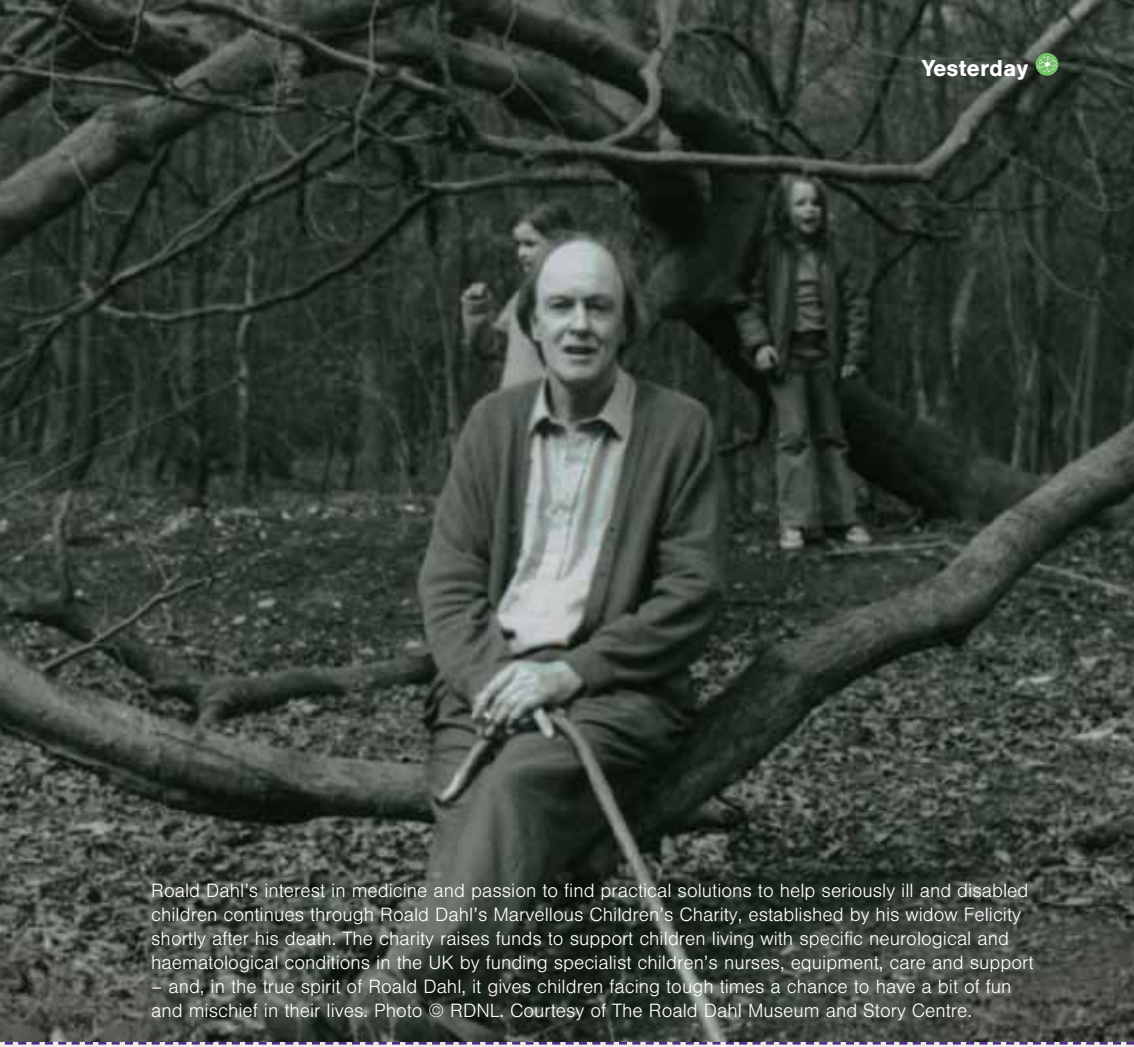
The parent, the engineer, and the pioneering paediatrician

Everyone knows the fantastic stories of Roald Dahl. But perhaps less well-known is the remarkable outcome of a devastating road accident in 1960, which left Dahl's son Theo with hydrocephalus at the age of four months.

Hydrocephalus causes a build-up of fluid in the skull, putting potentially lethal pressure on the brain. Treatment for the condition involved inserting a 'shunt' – a valve attached to a tube – which drained the excess fluid from the skull and relieved pressure on the brain. Unfortunately, the standard Holter shunt used by surgeons at the time, jammed regularly causing headaches, nausea and at worst extreme pain and even temporary blindness. Replacing it required more invasive surgery, and Theo spent the next three years undergoing operation after operation.

Determined to improve his son's condition, Dahl teamed up with an old friend and hydraulic engineer, Stanley Wade. Charged with the task of helping Theo, they approached Kenneth Till with a challenge to invent a new and better shunt valve.

By 1962, they had not only successfully designed and built a new device, but one which was robust, easy to sterilise and, crucially, didn't jam. Thanks to their collaboration it was patented and rolled out at one-third of the cost of other commercially-available valves. Thousands of children benefited immeasurably thanks to this inspiring and extraordinary collaboration.



Roald Dahl's interest in medicine and passion to find practical solutions to help seriously ill and disabled children continues through Roald Dahl's Marvellous Children's Charity, established by his widow Felicity shortly after his death. The charity raises funds to support children living with specific neurological and haematological conditions in the UK by funding specialist children's nurses, equipment, care and support – and, in the true spirit of Roald Dahl, it gives children facing tough times a chance to have a bit of fun and mischief in their lives. Photo © RDNL, Courtesy of The Roald Dahl Museum and Story Centre.



Image courtesy of McVirn Etienne.

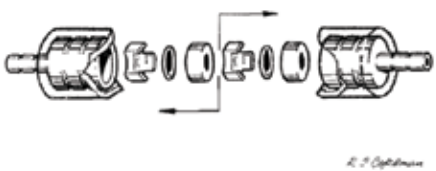
My grandfather the inventor

by Sophie Dahl

“My grandfather liked to know what made people and things tick. He had a relentlessly curious nature. It is no surprise that he collaborated on such an innovative invention, as he was always searching for answers.

“The previous shunt that had been used to treat children with hydrocephalus, like his son Theo, was clumsy at best. As a parent of a child with brain injuries, my grandfather had witnessed first hand the devastation they could wreak. The shunt had awful side effects and watching his baby (who had already been through so much) suffer them must have been excruciating. I can picture my grandfather now, thinking: ‘There must be a better way than this’. I imagine the eureka moment when he, Stanley Wade and Kenneth Till realised there was.

“I find the story incredibly moving. An engineer, writer and neurosurgeon, each used their specific skills to create a device that would improve life for children with neurological problems. The collaboration on the shunt shows that with imagination we are limitless. It’s an awe-inspiring, magical story.”



Left: Sophie Dahl.
Right: the Wade-Dahl-Till shunt valve.



Staff on our neurosciences wards.



Today



Incredible advances in visualising the living brain

Over the course of the 1970s, the emerging technology of computed tomography (CT) scanning was allowing researchers to capture detailed images of the living brain. Having historically worked on a combination of blind guesswork, invasive injections and often inconclusive x-ray images, neurosurgeons could now map the brain accurately before surgery. They could identify the position and size of tumours and lesions and plan operations in advance.

Kenneth Till was aware of the vital impact this new imaging could have on children's neurosurgery. He was quick to engage the support of philanthropists such as Sir William 'Billy' Butlin, who gave a £205,000 donation which made Great Ormond Street Hospital the first in the UK to have its own CT scanner suite, opened by the Queen in 1977.

The success of operations for hydrocephalus and brain tumours rocketed. In a short space of time, Great Ormond Street Hospital's neuroscience unit grew from carrying out 300 to 800 operations a year. For the first time, children with complex neurological conditions could be given a real chance of successful surgery.

Thanks to these developments, today Great Ormond Street Hospital is a national specialist referral centre for neuromuscular diseases such as Duchenne muscular dystrophy as well as craniofacial surgery and disorders of the blood vessels such as vein of Galen.

Left: The Queen looking at an image of the brain on the hospital's CT scanner with Sir Billy Butlin.

1977 Billy Butlin donates funds to purchase the UK's first paediatric CT scanner.

A touch of superglue to cure Druhan

Druhan was born with a condition called vein of Galen malformation. Because of missing capillaries, which normally slow blood flow, blood is pumped at high pressure from the arteries into the veins. As the veins aren't built to carry such pressure, this can result in blood leaking into the brain. If left untreated the condition is fatal, causing heart failure or brain damage.

Shortly after Druhan's birth, Consultant Neuroradiologist Stefan Brew carried out a number of CT scans on Druhan's brain. He recommended Druhan undergo immediate surgery to relieve the pressure arising from the build-up of blood. The operation involved sealing off the leaking blood vessels with a special kind of superglue. This would be fed through fine tubes directed into the brain via blood vessels using advanced imaging. Great Ormond Street Hospital was the first hospital in the UK to develop this technique for children – called interventional radiology.

The technique allowed Stefan to carefully navigate a path into the brain from a blood vessel in the leg, avoiding opening Druhan's skull. The procedure lasted several hours, but was a complete success.

Druhan now has speech therapy and is doing very well. His father Andrew says: "We're hugely grateful to Great Ormond Street Hospital and to Stefan Brew. He put us at ease, answered every question we threw at him and he was honest. We felt we'd made the right decision going to him. That man saved our son's life."



Druhan today.

Better surgery, long-term collaboration and much-needed expansion

We've come a long way from the days of having only one neurosurgeon permanently on call. Today at Great Ormond Street Hospital, five full-time consultants lead a team of over 25 doctors, clinical nursing specialists and other nurse and support staff. We have the largest paediatric neurosciences centre in the UK, treating over 8,000 children from across the world every year.

Research to apply new technologies such as CT and, more recently, magnetic resonance imaging (MRI), has completely revolutionised the accuracy and success of our operations. On average, a patient's stay at the hospital is less than half that of twenty years ago. Despite this, many more children need our help and we have an urgent requirement to increase capacity.

Thankfully, with the ongoing generosity of Great Ormond Street Hospital Children's Charity supporters, a new children's neurosciences centre is due to open in 2012. The centre will bring together the neurology and neurosurgery teams and provide new beds and state-of-the-art operating theatres.

Our surgeons are also at the forefront of developing strong working links with specialists outside of the neuroscience team. Such collaborative models of working have been crucial to improving patients' care. For example, our craniofacial team, joined with the plastic surgery team to plan and deliver vastly improved surgery for children with deformities of the face and skull.

 1994 The hospital pioneers new investigation of brain damage, using MRI, ultrasound and spectroscopy.



The neurosciences centre will be based in the Morgan Stanley Clinical Building, part of the Mittal Children's Medical Centre.



Ground-breaking treatment to shrink Madison's brain tumour

Madison was just a week old when she was admitted to Great Ormond Street Hospital with a large benign tumour in her brain.

It was a midwife who first noticed something might be wrong, as little Madison was unusually drowsy and her head was swollen and spongy. She suggested that the family get Madison checked. "When the doctors at Great Ormond Street Hospital said she had a tumour, it felt like it wasn't happening to me. I felt like the world had ended. It's something you don't ever want to hear," her mother Charlene recalls.

Madison's condition was very unusual, as babies are very rarely born with tumours and only a very few present in the first month of life. Left untreated, Madison would certainly have died, but doctors at Great Ormond Street Hospital undertook a risky, but ground-breaking procedure known as an embolisation. The treatment involved injecting arteries with glue to restrict the blood supply to the brain, killing the tumour whilst keeping blood flowing to the healthy regions of her brain.

Despite having seven operations in seven weeks, Madison responded well to the treatment and was able to go home for Christmas at just eight weeks old.

Left: Madison after the pioneering operation to treat her brain tumour.

Learning from our patients and sharing our discoveries

We've come a long way since our early days as the first and only provider of children's neurological care in a medically-limited field. Our teams now treat a full spectrum of complex and often life-limiting neurological illnesses – from spina bifida to facial reconstruction surgery; head trauma to hydrocephalus; brain tumours, epilepsy, and neuromuscular disorders. But we're always looking for ways to improve the care we provide.

Research by the hospital's surgical team showed certain children with a deeply-buried brain tumour – called a craniopharingioma – could benefit from less aggressive surgery and a course of radiotherapy. It meant those at risk of complications from surgery could be spared from severe personality disorders such as stealing and obsessive eating, while still removing the risk of the tumour spreading.

Meanwhile, in 2008 our epilepsy team announced the results of a landmark clinical trial of a carefully-controlled high fat diet to reduce the number and severity of children's seizures. This 'ketogenic' diet – named after the chemicals it causes the brain to release when starved of carbohydrates – had been known about since the 1920s. However, the team at Great Ormond Street Hospital showed that in children with drug-resistant epilepsy the diet could reliably reduce seizure frequency by up to a third.

 2008 The team demonstrates ketogenic diet effective in treating certain drug-resistant epilepsies.



A patient undergoing MRI scanning in the hospital's imaging suite.

Joni's story

Joni had her first seizure when she was just six months old. Her worried parents didn't know what was happening. They thought she was choking and called 999, and she was diagnosed with epilepsy immediately.

For the next nine years, Joni tried countless drugs and drug combinations with varying levels of success. At its very worst she was having 1,000 seizures a month. She had countless CT and MRI scans, which eventually revealed a lesion on the frontal lobe of her brain. Her consultant referred Joni to the epilepsy team at Great Ormond Street Hospital, to learn more about the exact nature of her condition.

Joni's consultant used a combination of specialist electrical monitoring and imaging equipment available at Great Ormond Street Hospital to pinpoint the precise region of the brain causing the huge numbers of seizures. During a five-hour operation he removed the lesion and Joni was discharged after five days.

Joni is now doing well and has only had one seizure since her surgery. She loves school, riding her bike and skiing. Her parents say: "She is so much happier and talkative since her operation. We are so grateful to the team of geniuses who helped make our daughter better."

Right: Joni experienced up to 1,000 seizures a month. She is now on the road to recovery.



Meet the team



“I arrived to learn paediatric neuroscience 21 years ago. The unique nature of patients referred here inspired me to learn more, to pursue better treatments and improve outcomes for the complex group we treat. I feel it a privilege to work here, both with regard to the fellow professionals with whom I work as well as the families we meet. The patients and families continue to inspire me – they often come to the hospital as a last resort but it is our role to help them. We have a unique range of patients who teach us more about their conditions day by day, and further our knowledge. As a result of the work we do, we can diagnose more children whose epilepsy was previously unknown and offer them previously unavailable treatments, such as surgery and the ketogenic diet. We have built up international expertise in treating the most complex epilepsies, and through that have advanced medicine for generations to come.”

Helen Cross, The Prince of Wales Chair of Childhood Epilepsy



There is a wide spectrum of tumours that affect the central nervous system in children. Many can be successfully treated with excellent prospects for healthy survival into adulthood. However, some tumours can be difficult to treat and a cure for all remains an elusive goal. Caring for the child and family through their illness is never the responsibility of an individual specialist but rather a carefully co-ordinated effort drawing upon expertise from a wide range of disciplines. The breadth and depth of expertise on hand at Great Ormond Street Hospital makes it a privilege to be a part of the team. *We aim not only to provide a world-class service to our patients, but also to share and learn from our experience through research, in an attempt to improve the lot of future generations.*

Dominic Thompson, Consultant Neurosurgeon

It must be a parent's nightmare to find themselves on our neurosurgical ward, suddenly surrounded by other sick children in head bandages and hearing new and confusing medical terminology describing their own child. Their bewilderment and fear can be partly overcome by the expert advice, support, education and care provided by the neurosurgical team. Nurses are at the forefront of this care – being part of a dynamic nursing team means we can work alongside our children and families and provide support, kindness, humour and outstanding care.

Lindy May, Nurse Consultant






Tomorrow



A brighter future

With the increasing specialism of today's medicine, a vital ingredient of Great Ormond Street Hospital's success has been to constantly investigate the best medicines, techniques and how to support our patients at every stage of their illness.

We're committed to improving our understanding of neuromuscular diseases and epilepsy, as well as less common neurological conditions such as spina bifida which affect children from birth. As the UK's largest centre for treating paediatric neurological disease, our teams are well-placed to remain at the forefront of medical advancement, constantly improving the care we offer.

As one of our senior clinical researchers puts it: "Carrying out thoughtful patient-focused research at the cutting edge of medicine is frighteningly expensive. But even if the work we do only helps a handful of patients, we have to be the ones to take the step into the unknown – it's what we do here."

Only with the ongoing generosity of our supporters can we continue our mission to advance research that will improve the lives of children with neurological diseases across the UK and beyond. Through the research we carry out today, we're investing in the lives of tomorrow's patients and advancing medicine for the benefit of generations to come.

Left: Great Ormond Street Hospital patient, Jaya.

Relearning how we learn

Some children exposed to low oxygen levels at birth or due to surgery early in their lives, have brain damage so severe that they can't create memories. Making sense of their own identity becomes an enormous challenge.

Since the hospital's pioneering use of imaging techniques to map the structure and activity of the brain in detail, there has been a revolution in the breadth and variety of questions we can answer about the workings of the human mind. Despite this, we still have much to learn about how the brain forms memories and are a long way from being able to cure children with these severe forms of brain damage.

Determined to help, Professor Faraneh Vargha-Khadem is leading research to support children with these severe and chronic forms of memory disorder, known as developmental amnesia. Her work aims to develop special educational materials and learning techniques to help these children form lasting memories, and gain a degree of independence as adults.

> *"It's the children who inspire us to gain a better understanding of their difficulties, so we can devise methods of overcoming them and help improve their quality of life. It's so important for children to construct a rich tapestry of their life events and to learn from their past experiences. We hope that our research will eventually provide children who have memory problems with the necessary tools to form an autobiography, and to capture their memories to share with others."*
>
> Professor Faraneh Vargha-Khadem (pictured, right)



Ensuring visually impaired children get the best start in life

Across the UK, over 300 children each year are born with problems affecting the regions of the brain which process visual information. Sight forms such a crucial part of our brain's function, that these visually-impaired children are at a risk of severely delayed development. This can impact hugely on the way they learn later in life and how they interact with their parents and other children.

A major part of Great Ormond Street Hospital's work is to provide support to help children outside of its clinics, and prevent unnecessary setback wherever possible. To achieve this, our neuroscience team have just begun a unique research project. With joint funding from Great Ormond Street Hospital Children's Charity, Fight for Sight and the Royal National Institute for the Blind, they are evaluating a tailor-made package of learning and psychosocial support tools for children with visual impairment.

If proven effective, this package will help parents and clinicians outside the hospital keep track of their child's development and flag issues early enough for action to be taken. It will help children to have a chance of keeping up with their peers, reduce parental stress and build confidence, ultimately changing the way visually-impaired children are supported across the UK.

Right: Louie, one of the early participants in research to help children with visual impairment.



Louie's mum talks about her experience of being a part of this research initiative:

> *"We found the research programme to be a huge help. The support we've had as a family has played a big part in Louie's development and we are very grateful for everything that we have received."*

Tracing the living pathways of the brain

Through a history of innovating and applying the latest technological advances to their work, our neurosurgeons are able to map, plan and perform their operations more accurately than ever before. However, the brain is by far the most complex organ in the body and there is still huge room for improvement if children having surgery for brain tumours and epilepsy are to be guaranteed a healthy outcome.

Dr Chris Clark is applying the latest techniques in magnetic resonance imaging (MRI) to document the natural history of the brain in ways as yet unexplored in a paediatric setting.

By building up a picture based on the living connections of nerves, his research uses a technique called tractography to map regions of the brain connected by the web of nerves responsible for carrying signals around the body. These maps provide a unique patient-specific insight into how the 'thinking' brain works. Armed with this invaluable information, surgeons can begin to know whether their operations will damage key pathways in the brain.

As well as revealing new insights into the biology of the body's least understood organ, it could make the difference between successful brain surgery and life-long paralysis.

Right: Mapping the brain's living connections, such as the optic nerve (shown here in fire colours) allows neurosurgeons to avoid damage when carrying out delicate operations such as those to treat epilepsy.

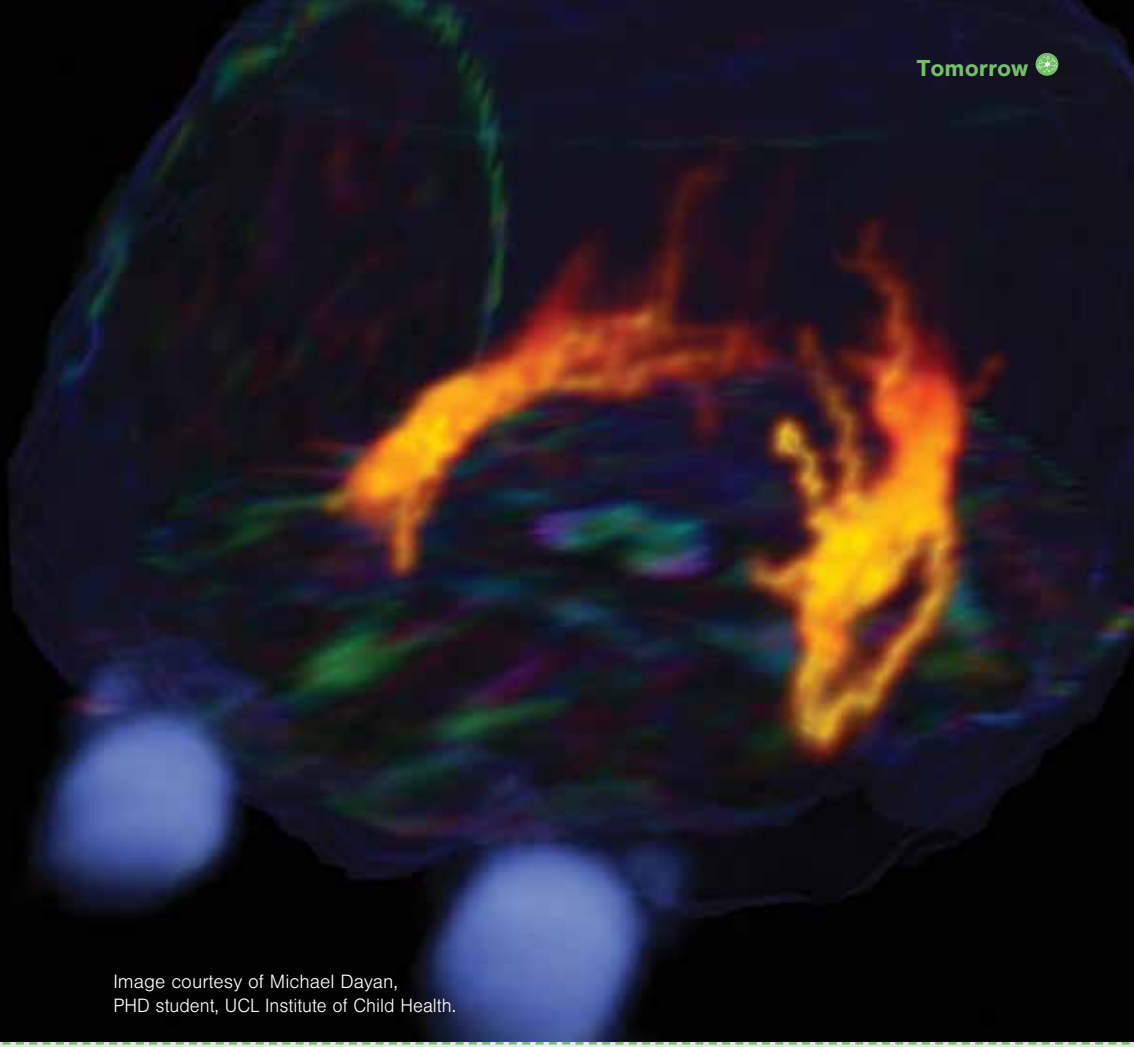


Image courtesy of Michael Dayan,
PHD student, UCL Institute of Child Health.

Thank you

We have come a long way in the past 150 years. Our dedicated and passionate staff have pioneered many new, and better ways of treating some very sick children. But we are yet to cure all the patients we see.

Throughout our history, it has been the continued and generous support of our donors who have helped to fund the research that provides new breakthroughs. With their ongoing generosity we seek to give hope to every child that needs our specialist help.

Right: Louie, who was born with hydrocephalus.

