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Chapter 1. The Charlie Gard case

"Begin at the beginning and go on till you come to the end; then stop."

Alice in Wonderland

Shortly after three-o'clock on 28th July 2017, in a private room of a specialised hospice in suburban London, staff disconnected the breathing machine that was keeping alive 11-month old infant, Charlie Gard. Lying on either side of him, his parents Connie Yates and Chris Gard held his hands, leaned close, and talked to him. They spoke to him of how proud they were of him. They saw him open his eyes briefly, and close them. Minutes later Charlie's heart stopped, and he was gone.[1]

This was the conclusion - quiet, private and fleeting, to a dispute about medical treatment that had been none of those things. In the preceding four months, a protracted series of court hearings around disputed treatment for Charlie Gard had yielded global media attention, and an outpouring of sympathy from onlookers around the world. It had attracted statements of support from many public figures, including US President Trump, and the Pope.

Charlie's parents had been seeking experimental treatment that might improve his rare genetic condition. They had raised funds for him to travel to the US, where a medical specialist had offered to provide treatment. Many observers could not understand why the doctors at London's Great Ormond Street Hospital opposed the requested treatment. Many found it hard to see why the UK and European courts[2] had decided against Charlie's parents and authorised withdrawal of life support.

In the final stages of the court hearing, three days before he died, Chris and Connie Yates accepted the view of experts that medical treatment could no longer help him and it was time to allow him to die. Their own medical experts from overseas had examined Charlie and concluded that his condition was now too far advanced for experimental treatment to work. The family agreed to a plan to withdraw life support, though bitterly regretted what they perceived as a lost chance to help Charlie.

1 Smith-Squire, A. (2017) Our last hours with our son. *Daily Mail*. accessed from http://www.dailymail.co.uk/ news/article-4762390/Charlie-Gard-s-parents-hours-son.html .

2 We will refer in this book to 'UK courts', but it is worth acknowledging that there are some differences between courts in different parts of UK. Most cases of disputed medical treatment relating to children have been referred to the courts of England and Wales.

But how had it reached this point? Where did the disagreement come from, and why had this decision been a matter for the court in the first place?

In this first chapter, we will describe the background to the Charlie Gard case and how it played out over the first half of 2017. We will look at how decisions about medical treatment are normally made and the role of the court in decisions. We will outline some of the important ethical questions raised by the Gard case, that will form the basis for the rest of this book.

The Charlie Gard Story

Charlie Gard was born, apparently healthy, in West London in August 2016. His parents, Chris and Connie, enjoyed a normal first few weeks with him at home. However, at a few weeks of age his parents started to notice that he wasn't able to lift his head as well as other children, an early sign of muscle weakness. By a couple of months of age, he was losing weight and muscle strength, and Charlie was admitted to the specialist children's hospital at Great Ormond St where soon afterwards he was put on a breathing machine in intensive care because he was not strong enough to breathe for himself. Doctors suspected that he had a genetic condition affecting his muscles and brain. A biopsy of his muscle, and later on, whole genome sequencing, identified the cause as an extremely rare condition with a long technical medical name: "infantile onset encephalomyopathic mitochondrial DNA depletion syndrome" ('MDDS' for short).

There are a number of genetic disorders that can cause worsening muscle weakness in babies. Some of these are problems with the structure of muscles. Others are problems with nerves, nerve signals or the brain.

MDDS is a condition that affects the basic energy structures of the body, the mitochondria. Mitochondria are found in most cells in the human body. There are several hundred in each human cell, and perhaps as many as 10 quadrillion in the whole human body. The energy released by these mitochondria fuels much of the activity of cells. They are often compared to batteries, but mitochondria are much more complicated than batteries. One little-known feature is that they contain their own genetic material or DNA. One theory is that they were originally bacteria captured by our primitive ancestor cells. Mitochondrial DNA is separate from the main chromosomes that we inherit from our parents (which live in the nucleus of the cell). It is much smaller than nuclear DNA. Nevertheless, these mitochondrial genes have essential roles in energy production and in regulating other important cellular processes.

Normally, mitochondria are continuously making more mitochondrial DNA. They do this by synthesising and by recycling the basic building blocks of DNA (nucleosides) and then assembling them in a way that copies the existing mitochondrial genes ("replication"). This continuous process of replicating mitochondrial DNA is essential for the mitochondria to work properly. In some very rare genetic conditions, however, there are mistakes (mutations) in the instructions that tell the cells how to make the mitochondrial DNA building blocks or in how to put them together. This leads to the cells having much lower amounts of mitochondrial DNA than normal – this is the problem that affected Charlie Gard.

There are different forms of MDDS, but all typically cause progressively worsening problems of energy production in the body. As an analogy, it is a little bit like a problem with your mobile phone battery. Your phone might start out working fine, however, over time, the charge seems to run out very quickly, and the battery does not last long before running out of power. In the case of a mobile phone, you might be able to get a new battery – but of course it isn't possible to replace billions of mitochondria.

The specific genetic form of MDDS in Charlie Gard (affecting a gene called "RRM2B") had previously only ever been reported in 15 cases in the world. All of these babies had developed early onset of muscle weakness, rapid progression of symptoms and death within a few months.[3] This form of MDDS is called 'encephalomyopathic'. The muscles do not work properly because of a lack of energy, while the effect on the brain leads to a lack of brain growth and seizures.

By the time that doctors had diagnosed Charlie's condition, he was paralysed and unable to breathe without a machine. He was found to have congenital deafness, and his heart, liver and kidneys were affected by the disorder. As this diagnosis became clear, doctors at Great Ormond Street felt that the outlook for Charlie was extremely poor and started to talk to his parents about whether continuing to keep him alive on life support was the right thing to do. In November, there was a meeting of the clinical ethics committee at Great Ormond Street hospital to discuss whether Charlie should have surgery to perform a tracheostomy. This is a procedure that creates a hole in the front of the neck so that a breathing tube can be passed directly into the windpipe (trachea) rather than being inserted through the nose or mouth. This surgery is performed for patients who are receiving long-term invasive breathing support from a machine. At that meeting, the ethics committee heard evidence from doctors. They felt that Charlie's quality of life was so poor that "he should not be subject to long term ventilation".[4] The ethics committee supported the medical team's assessment that Charlie should not have a tracheostomy. There is no cure for MDDS, and no proven treatment for Charlie's severe form of the illness. However, Charlie's parents, understandably, found this news very difficult to accept. His mother took to the internet to research possible treatments that might be in development or used in other parts of the world. In that research, she found mention of a treatment that had seemed to help some children with a less severe form of MDDS. She was in contact with a parent in the US whose child had received that treatment and Connie Yates asked the specialists at Great Ormond St Hospital if they would try it for Charlie.

The problem for Charlie's mitochondria was a lack of the building blocks that he needed to make new DNA inside his mitochondria. But what if he were given a supplement containing those building blocks? Could that bypass the problem and help his cells to make more mitochondrial DNA?

That is the basic idea behind the treatment that Connie Yates found on her search of the internet. 'Nucleoside therapy' (also called 'nucleoside bypass therapy') has been used in children with a different genetic form of MDDS. "TK2" is another gene that causes depletion of the mitochondrial DNA. The TK2 form of MDDS is also rare, but it has been described in more than 50 children.[5] In this condition, the problem isn't in making new DNA building blocks nucleosides – it is in the genes that recycle old nucleosides. Because cells are continuously making mitochondrial DNA, they need to be very efficient at recycling and reusing the building blocks from old strands of DNA. There are two basic types of nucleosides called purines and pyrimidines. In TK2, there is a problem with recycling the pyrimidine nucleosides. This causes less severe problems than the RRM2B form – children develop weak muscles in their first 1 or two years of life (rather than in the first weeks or months as in Charlie's illness). They too can have problems with muscle strength affecting their ability to move, feed and breathe. However, it does not usually affect the brain as much. Children with the TK2 form of MDDS have in the past usually died in early childhood, though some have survived longer, even into adulthood.[6]

Arturito Estopinan is one of those older survivors. In 2012, when Arturito was age one, doctors in the US diagnosed him with the TK2 form of MDDS. His parents were told that there was no medical treatment and that he would likely die within months.[7] However, just like Charlie Gard's parents, the Estopinans combed the internet and contacted experts across the US looking for something that might help him. They spoke with one

3 El-Hattab, A. W. & Scaglia, F. (2013) Mitochondrial DNA depletion syndromes: review and updates of genetic basis, manifestations, and therapeutic options. *Neurotherapeutics*, 10, 186–98. PubMed PMID: 23385875.

4 para 59. Details here and below are taken from the High court judgment, with paragraph numbers cited for specific quotations. The full judgment is available at http://www.bailii.org/ew/cases/EWHC/Fam/2017/972.html Great Ormond Street Hospital v Yates & Ors [2017] EWHC 972 (Fam) (11 April 2017)

5 El Hattab 2013

6 ibid

7 Gebelhoff, R. (2015) Family carves out its own path in fight against 4-year-old's rare disease. *Washington Post*. accessed from < Available at: https://www.washingtonpost.com/national/health-science/family-carves-out-its-own-path-in-fight-

expert, Professor Michio Hirano, a neurologist in New York who had been experimenting with a new treatment in mice who had a disease like TK2. Arturito was the first person to receive nucleoside treatment, and a year later he was able to come home, while still receiving intensive medical care and breathing support. In July 2017, Arturito's father described some of the improvements in muscle strength they had seen with the nucleoside therapy: "They have allowed him to get stronger by moving his arms, his fingers, his legs. He's even trying to move his hips".[8] Arturito remained severely disabled, and dependent on a ventilator to breathe. But he was alive, and rather than worsening, he had improved to some degree. This improvement has also been seen in research studies. In mice, genetically engineered to have a form of TK2, early supplementation with the pyrimidine nucleosides apparently led to higher mitochondrial DNA levels and less severe clinical features.[9] It wasn't a cure, but it seemed to help.

Connie Yates had found Arturito's story when she was researching Charlie's condition. She contacted Arturito's parents, who put her in contact with Dr Hirano. If nucleoside therapy could help in TK2 forms of MDDS, could it work in Charlie's form – RRM2B? Dr Hirano couldn't give a definite answer. Nucleoside treatment hadn't been tested, either in animals or in humans with RRM2B. The genetic defect in this condition is more severe, and giving the same pyrimidine nucleosides that help in TK2 wouldn't be enough. In theory though, giving a combination of three different nucleosides might work. Dr Hirano told Connie that could be worth a try given how serious Charlie's condition was. At the request of Charlie's parents, Dr Hirano corresponded at the end of December with the mitochondrial specialist at Great Ormond St. The specialist asked Dr Hirano a series of questions, which were later reported in one of the court judgments:

> *Question 1: "What is the evidence that this treatment might help?"*
>
> *Dr. Hirano's response: "There is no direct evidence, but there is a theoretical scientific basis for saying it could."*
>
> *Question 2: "Could the drugs cause toxicity?"*
>
> *Dr. Hirano's response: "The only toxicity seen is dose related diarrhoea".*
>
> *Question 3: "As the drugs do not cross the blood/brain barrier, is there any possibility of efficacy in a child with an epileptic encephalopathy?"*
>
> *Dr. Hirano's response: "This had been previously suggested in published research, but there is theoretical and anecdotal evidence that the drugs could in fact cross this barrier and, therefore, have effect on the brain. In particular, TK2 patients who have been treated have not developed seizures or encephalopathy's as had those who were not treated."*
>
> *Question 4: "If we were to embark on a clinical trial, how long would you suggest and what outcome measures?"*

against-4-year-olds-rare-disease/2015/09/04/7d18d8b0-2c91-11e5-a5ea-cf74396e59ec_story.html?utm_term=.53490173263c>.

8 Knight, V. (2017) His son tried the experimental treatment Charlie Gard's parents want. *CNN*. accessed from < Available at: http://edition.cnn.com/2017/07/18/health/american-baby-treatment-charlie-gard-case/index.html>.

9 Lopez-Gomez, C., Levy, R. J., Sanchez-Quintero, M. J., Juanola-Falgarona, M., Barca, E., Garcia-Diaz, B., et al (2017) Deoxycytidine and Deoxythymidine Treatment for Thymidine Kinase 2 Deficiency. *Annals of neurology*, 81, 641–652. PubMed PMID: 28318037.

Dr. Hirano's response: "A three month trial should be sufficient" He suggested a range of non-invasive outcome measures."[10]

The third question was important to the doctors at Great Ormond Street because there were worrying signs that Charlie's brain was affected by MDDS. In the middle of December, Charlie had started to have epileptic seizures. Since he was paralysed, there weren't always outward signs of these seizures. However, electrical tracings of his brain (EEG) had shown a combination of frequent seizures as well as abnormal background electrical activity. These findings together indicated a brain that was very sick ("epileptic encephalopathy"). Moreover, The Great Ormond Street doctors were concerned that nucleoside therapy would not reach the brain because of the protective "blood-brain barrier".

Dr Hirano was supportive of trying nucleoside therapy. Given that, the Great Ormond Street specialist started to arrange for Charlie to receive the treatment. This was an untested and unlicensed medicine, and to be able to provide it would require special approval. The decision was referred back to the ethics committee at Great Ormond Street, with a meeting planned for 13th January. There was a provision for Charlie to have a tracheostomy on 16th January.

However, the meeting, and the surgery, did not go ahead. In early January, the doctors arranged further tests of Charlie's brain. This included a magnetic resonance imaging (MRI) brain scan on the 6th of January. This was essentially normal and showed only subtle changes, a finding that is apparently typical in MDDS because the basic problem is at the microscopic level (and so isn't visible on scans). A repeat EEG, though, on the 10th of January, was felt to again show signs that Charlie's brain was severely affected by MDDS – with frequent electrical seizures and a pattern of severe epileptic encephalopathy.

On the 13th of January, doctors at Great Ormond Street hospital met with Charlie's parents and told them that the trial of nucleoside therapy would not go ahead. All of the medical teams at the hospital involved in Charlie's care had agreed that this treatment would be "futile and would only prolong Charlie's suffering".[11]

There had been some disagreement between Charlie's parents and the doctors at Great Ormond Street since November, but by this meeting in January, it seemed clear that the medical team and Charlie's parents had very different views about what would be best for him. Charlie's parents were determined that life support should continue, and that Charlie should receive a trial of nucleoside therapy. His doctors, though, felt that it was wrong to continue to keep Charlie alive. They felt that he was suffering by being kept alive by machines in intensive care, and that nucleoside treatment had no hope of helping him.

How often do disagreements like this occur in intensive care units? How are they usually resolved?

There are approximately 2000 children who die in paediatric and neonatal intensive care units each year across the UK.[12] Some of these children die suddenly, with little warning. In many cases though, a child is seriously ill and, sadly, death is expected. In paediatric and neonatal units, somewhere between 2/3 and three quarters of deaths follow decisions to either stop life-support or not to attempt resuscitation if a child deteriorates (we will refer to these together as "end of life decisions").[13] There are different reasons why treatment might not be provided, but fundamentally, the concern is that continuing treatment or trying to resuscitate would not help,

10 Para 76, High court judgment

11 Para 83

12 In England and Wales, there are approximately 650 deaths in paediatric intensive care units a year, Plunkett, P. & Parslow, R. (2016) Is it taking longer to die in Paediatric Intensive Care in England and Wales. *Arch Dis Child.*. There are another 1400 newborn infants who die each year across the UK MBRRACE-UK. (2016). 'Understanding babies' deaths in the UK: 2014.' from < Available at: https://www.npeu.ox.ac.uk/downloads/files/mbrrace-uk/reports/MBRRACE-UK-PMS-Report-2014-Infographic-poster.pdf>.. A high proportion of these deaths are likely to occur in neonatal units.

and would do more harm than good. That might be because even with treatment, the child would survive for only a short time ("quantity of life"). Or it might be because the child could survive, but would do so with a very reduced "quality of life". We will return to discuss those reasons in more detail in chapters 4 and 5.

Across the country, then, as many as three or four times every day, there are decisions made about life support for children or babies. Those decisions, in the vast majority of cases, are made jointly by families and health professionals. Parents or carers, together with the doctors and nurses looking after the child, come to a shared view that certain types of potentially life-prolonging treatment are not right for the child. That is not to say that these decisions are easy, or are made lightly. Quite the opposite: these are often agonising decisions, made only after much soul searching, and after all other options have been considered and rejected. The point is rather that conflict and disagreement are not an inevitable or even a usual part of end-of-life decision-making for children. In many cases, in most cases, while decisions are difficult, and distressing – they are able to be reached with agreement of all of those caring for the child – both professionals and family.

But how often is there disagreement? It is hard to know. One study in the Netherlands interviewed health professionals caring for 147 newborn infants who had died after an end-of-life decision. In about one in ten cases (18 infants), the doctors reported that there had been conflict between the professionals and parents.[14] Many of those cases were ones where the infant was predicted to be severely disabled if they survived (these are 'quality of life' reasons – see chapter 5). However, in all of the 18 cases, agreement was eventually able to be reached. Sometimes, that was after the parents obtained a second opinion. Sometimes it was after the child's condition worsened and parents changed their mind. In some cases, a compromise was reached, where treatment wouldn't be withdrawn, but resuscitation would not be provided if or when the child's heart stopped. A recent Canadian study of 942 babies who died, asked doctors about end of life decisions that had been made. From almost 800 cases where there had been discussions about end of life treatment, doctors reported that it had been "difficult" to reach agreement in only 12% of cases.[15]

A study of neonatal units in London provides some additional insight. They reported the outcome for 68 seriously ill infants, where there were discussions about end of life decisions. In half of these cases (34 babies), parents and professionals reached a decision to stop treatment or not resuscitate after an initial meeting. In the other half, treatment continued, and there were further discussions, often including second opinions. In 16 of these cases there was eventually agreement to limit treatment, while in the other half treatment continued.[16]

Protracted disagreement is the exception rather than the rule. There were no cases in any of those studies where doctors were reported to have obtained legal advice or gone to court. But where parents and health professionals cannot reach agreement there is the option of going to the court to resolve the impasse. One report suggests that in 2016 English courts were asked to make decisions about medical treatment for a child in 18 cases.[17] However,

13 Aladangady, N., Shaw, C., Gallagher, K., Stokoe, E., Marlow, N. & for Collaborators, G. (2017) Short-term outcome of treatment limitation discussions for newborn infants, a multicentre prospective observational cohort study. *Arch Dis Child Fetal Neonatal Ed*, 102, F104–F109 PubMed PMID: 27852667.. Hellmann, J., Knighton, R., Lee, S. K., Shah, P. S. & Canadian Neonatal Network End of Life Study, G. (2016) Neonatal deaths: prospective exploration of the causes and process of end-of-life decisions. Ibid. 101, F102–7, James, J., Munson, D., DeMauro, S. B., Langer, J. C., Dworetz, A. R., Natarajan, G., et al (2017) Outcomes of Preterm Infants following Discussions about Withdrawal or Withholding of Life Support. *J Pediatr*, 190, 118–123 e4. PubMed PMID: 28647272.

14 Verhagen, A. A. E., de Vos, M., Dorscheidt, J. H. H. M., Engels, B., Hubben, J. H. & Sauer, P. J. (2009) Conflicts about end-of-life decisions in NICUs in the Netherlands. *Pediatrics*, 124, e112–9. PubMed PMID: 19564256.

15 Hellmann, J., Knighton, R., Lee, S. K., Shah, P. S. & Canadian Neonatal Network End of Life Study, G. (2016) Neonatal deaths: prospective exploration of the causes and process of end-of-life decisions. *Arch Dis Child Fetal Neonatal Ed*, 101, F102–7. PubMed PMID: 26253166.

16 Aladangady, N., Shaw, C., Gallagher, K., Stokoe, E., Marlow, N. & for Collaborators, G. (2017) Short-term outcome of treatment limitation discussions for newborn infants, a multicentre prospective observational cohort study. Ibid. 102, F104–F109.

not all of those cases will necessarily have been about end of life treatment. (For example, some may have cases where parents disagreed between themselves about treatment, or where parents wished to refuse treatment – eg blood transfusions for religious reasons). From published court records, it appears that there were 5 end of life treatment disagreement cases in the first half of 2017. They included:

1. An 11 year old boy with advanced bone cancer and spread to his lungs. Doctors wished to provide with palliative care; parents did not agree with his diagnosis and were opposed to him being provided with palliative care. (Court decided in favour of providing palliative care)[18]
2. An 8-month old girl with inoperable complex congenital heart disease. Doctors did not wish to provide resuscitation (including inserting breathing tubes, and other invasive measures). Parents accepted that her life was limited, but wished her to receive resuscitation. (Court decided in favour of withholding resuscitation)[19]
3. An 13 year old child with severe multiple disabilities (severe cerebral palsy, scoliosis intellectual disability, epilepsy, poor lung function). Doctors did not wish to provide invasive life-prolonging treatment (including resuscitation, mechanical ventilation and renal dialysis) and wished to provide palliative care; mother was opposed to any limitation of treatment. (Court decided in favour of providing palliative care).[20]
4. An emergency court hearing about a 3-month old infant who had deteriorated after surgery for a large subdural haematoma and was thought to have very poor prognosis. Parents sought a court order to prevent doctors from limiting treatment; doctors wished to not provide further neurosurgery, resuscitation or escalation of treatment. (Court decided in favour of no further surgery or resuscitation).[21]

The fifth case was Charlie Gard.

After the meeting in January, it was clear to Charlie's parents that Great Ormond Street would not provide him with nucleoside treatment. Connie and Chris started to look at other options, and on 30th January they started an online appeal to raise funds for him to travel to the United States for treatment. They estimated that it would cost £1.2million for Charlie to be treated in the US.

Meanwhile, Charlie's doctors obtained a series of second opinions from intensive care specialists, neurologists, lung specialists and mitochondrial specialists at other hospitals in the UK. They obtained an opinion from a mitochondrial research team in Barcelona. with specific expertise in MDDS.

On the 24th February, lawyers for Great Ormond Street Hospital applied to the Family Division of the High Court seeking the court's permission to withdraw the artificial breathing support keeping Charlie alive, provide him with palliative care, and not to provide nucleoside treatment. There was a brief hearing in early March, which was adjourned to allow Charlie's parents to gather evidence from overseas experts and present them to the court.

17 Doward, J. & Robertson, H. (2017) Ten cases like Charlie Gard's heard in English courts this year. *The Observer*. accessed from < Available at: https://www.theguardian.com/uk-news/2017/jul/29/ten-cases-like-charlie-gards-heard-english-courts-this-year>.

18 An NHS Trust v SK (Best Interests Decision -Palliative Care) [2016] EWHC 2860 (Fam) (04 November 2016).

19 Great Ormond Street Hospital for Children Foundation NHS Trust v NO & KK & Ors [2017] EWHC 241 (Fam) (14 February 2017)

20 A Local Authority & Anor v MC & Ors (Care proceedings)(Inherent Jurisdiction) [2017] EWHC 370 (Fam) (24 February 2017)

21 An NHS Hospital Trust v GM & Ors [2017] EWHC 1710 (Fam) (30 June 2017)

In the first week of April, the court heard evidence from Charlie's parents, from experts at Great Ormond Street, and from the US expert Dr Hirano, who gave evidence by telephone. As is usual in such cases, there was a court-appointed guardian who was tasked with representing Charlie's interests in court. The following week, on the 11th April, Justice Nicholas Francis gave his decision.

We will summarise here the basic elements from that judgement, since they set out issues that we will return to in later chapters.

Legal principles

The first part of Justice Francis' judgment set out the legal principles underpinning his decision. As noted in the preface, our focus in this book is on ethical questions rather than on legal questions. Nevertheless, an appreciation of the key legal principles will help understand the decisions that were made for Charlie Gard. They will also provide a starting point for some of the ethical questions that we will return to in later chapters.

1. The court in the UK has legal authority to make decisions in a child's best interests.

   The judge noted:

   "Some people might ask why the court becomes involved at all, why should the parents not be the ones to decide? A child's parents having parental responsibility have the power to give consent for their child to undergo treatment, but overriding control is vested in the court exercising its independent and objective judgment in the child's best interests." (para 11)

   Obviously most of the time, and for most medical decisions, courts are not part of treatment decisions for children. The court is asked to make a decision either if there is no one to make a decision for a child (eg if the child is orphaned, or if parents are unable or unwilling to decide) or if there is disagreement about the decision. As indicated above, in rare instances health professionals and families cannot agree on what would be best for a seriously ill child. In that circumstance, the UK approach is neither to assume that the doctors know best, nor that parents are making the right decision. As in many other areas of life, when there is intractable disagreement between different parties about something important, there is a need for an impartial arbiter - the court is asked to decide. The court is then asked to determine what would be best for the child. This is sometimes referred to as the "best interests" of the child.

2. What is best for a child does not always mean prolonging life.

   Courts in the UK, as in other parts of the world, have traditionally been very cautious about making decisions that would lead to the death of a patient. The judge quoted a decision in the case of Charlotte Wyatt (another young child whose parents and doctors disagreed about treatment).[22]

   "There is a strong presumption in favour of a course of action which will prolong life, but that presumption is not irrebuttable." (para 13)

   In other words, prolonging life is important, but it is not always the right thing to do. This vital interest could be outweighed

   "if the pleasures and the quality of life are sufficiently small and the pain and suffering or other burdens of living are sufficiently great" (para 39)

22 Portsmouth NHS Trust v Wyatt [2005] 1 F.L.R. 21.

3. The best interests of a child include more than just medical factors.

   Medical evidence about illness and treatment is important, but court decisions in past cases have understood a child's (or older patient's) interests to incorporate a range of elements. For example:

   "These include … medical, emotional, sensory (pleasure, pain and suffering) and instinctive (the human instinct to survive) considerations." (para 39)

4. The views and opinions of parents should be carefully considered

   Parents are important because of their knowledge of the child "Where, as in this case, the parents spend a great deal of time with their child, their views may have particular value because they know the patient and how he reacts so well" (para 39)

   They are also important because of the child's interest in their relationship with their parents. Their wishes "may illuminate the quality and value to the child of the child/parent relationship" (ibid)

   However, courts in the UK have not considered parental wishes to be directly relevant to the child's best interests: "Their own wishes, however understandable in human terms, are wholly irrelevant to consideration of the objective best interests of the child" (ibid)

We will return shortly to the evidence heard by the court, but it is worth noting here that the first three principles are not particularly controversial, and are reflected (to a varying extent) in legal decisions about medical treatment for children in many different countries.

However, the last of the above four points is more distinctive, in that it identifies a distinctly limited role for parents in assessing the best interests of a child. It is this factor that perhaps explains why the UK is relatively unique in its legal approach to disputed treatment for children. As noted in the quotation from the judgment, parental wishes do not carry much weight for the court in consideration of the best interests of the child. It has meant that, unlike elsewhere, the English courts have been prepared to make decisions to authorise withdrawal of medical treatment against the objections of parents.[23]

In the US, by contrast, courts have been very reluctant to override parents who desire potentially life-prolonging medical treatment for their child.[24] This has proved true even in extreme cases such as that of Baby K, (an infant with anencephaly who received mechanical ventilation for a prolonged period)[25], or that of the Californian girl, Jahi McMath, in which a court had ruled the child was 'legally dead.'[26]

After summarising the legal basis for the decision, Justice Francis summarised evidence presented by the hospital and by Charlie's parents. That evidence related to two issues relevant to the decision for Charlie: his current quality of life and the benefit of nucleoside treatment. A third important issue, that of the cost of treatment, was considered not to be relevant.

23 This might also explain the role of the court-appointed guardian in Family Court proceedings. In cases like Charlie Gard's, the court appoints a separate independent party for the child who has a specific role to safeguard the child's interests. For an older child, the guardian has an independent role in assessing the wishes of the child.

24 Paris, J. J., Ahluwalia, J., Cummings, B. M., Moreland, M. P. & Wilkinson, D. J. (2017) The Charlie Gard case: British and American approaches to court resolution of disputes over medical decisions. *J Perinatol*.

25 New York Times News Service. (1994) Court says doctors must treat Baby K, infant born without most of brain. *Chicago Tribune*. accessed from < Available at: http://articles.chicagotribune.com/1994-02-20/news/9402200448_1_periodic-respiratory-crises-anencephalic-babies-active-labor-act>.

26 Luce, J. M. (2015) The uncommon case of Jahi McMath. *Chest*, 147, 1144–1151. PubMed PMID: 25846530.

A. Quality of life

The court heard evidence from various medical experts about Charlie's current medical state. They indicated that he was dependent on a mechanical ventilator as he had no ability to breath on his own. He could no longer move his arms or legs, and had no spontaneous movement of his fingers, hands, toes or feet. He was no longer able to open his eyes enough to be able to see. Because of his genetic condition, he was deaf. The intensive care consultant at Great Ormond St indicated that repeated EEG tests had shown that he was "persistently encephalopathic" – he was not brain dead, but there was no sign of normal brain responsiveness.

One of the doctors who examined Charlie reported seeing no movement apart from a flicker of Charlie's lips in response to pressing on his fingers. A nurse from the intensive care unit, who had spent over two hundred hours at Charlie's bedside over several months, reported that she had not seen him grasp fingers. Because of his lack of movement and response, she felt that it was impossible to tell if Charlie was awake or asleep.

Charlie's parents had a different view about Charlie's brain function. They felt that he knew when his parents were present, and could experience pleasure (for example tickles). They reported that he sometimes tried to hold their hands and open his eyes. They could tell the difference between when he was awake and asleep.

There were different views about whether, or how much Charlie was experiencing pain or discomfort from the current treatment he was receiving. The bedside nurse who gave evidence reported that it was "impossible to know" if Charlie suffered pain, or experienced pleasure. His parents felt that there were things that he did not like (such as nasal suction, or a blood test), which they could tell because his heart rate would go up. The medical team gave evidence that the treatments that Charlie required to stay alive (being on a ventilator, having his airway suctioned) could cause pain. The mitochondrial expert at Great Ormond Street testified that it couldn't be demonstrated that Charlie experienced pain, but that it was possible. The medical team apparently believed that he was suffering. "She said that she did not regard his pain of being of a low level of suffering, but something more significant." (para 114)

Despite their different perspectives, both the medical experts and parents appeared to agree that Charlie's current quality of life was poor. The medical team had apparently felt, even before Charlie developed seizures that "his quality of life was so poor that he should not be subject to long term ventilation". His parents indicated that "We would not fight for the quality of life he has now… This is not the life we want for Charlie" (para 110)

B. Nucleoside treatment

Given apparent agreement that Charlie's current state was not one that should be prolonged with life-sustaining treatment, the key issue was whether the proposed nucleoside treatment would or could change that.

The court heard evidence about nucleoside treatment in patients with TK2. Dr Hirano reported that a group of international experts (including himself) had treated 18 patients with the TK2 form of MDDS with nucleoside treatment. Patients had gained weight on treatment. All were still alive. Some had improved in strength.

The nucleoside treatment had never been tried in animals or in humans with RRM2B, so its effect could not be predicted. There was a scientific rationale for the treatment, and (in the patients with TK2) it appeared to have few side effects. The only side effect was diarrhoea if given in too high a dose. It was also very cheap. Because it was relatively easy to administer, and low risk, the central question was whether it would work (and whether this would justify continuing intensive care for a period of months more).

Could nucleoside treatment work in Charlie? One important difference between the two forms of MDDS is that RRM2B affects the brain, while TK2 affects (mostly) the muscles. Dr Hirano indicated that nucleoside treatment could cross the blood/brain barrier, and potentially work in the brain. This was based on previous research experience in TK2. The Great Ormond Street expert, in contrast indicated that there was no evidence in humans that the nucleoside treatment could enter the brain.

The other central issue for Charlie was whether he already had sufficient brain damage that the nucleoside treatment could not help him. When he had first spoken with the medical team at Great Ormond St, Dr Hirano had indicated that Charlie should have an MRI brain scan as "severe brain involvement" would mean that treatment shouldn't be tried. As noted earlier, that MRI had not shown any major changes, however, the development of very abnormal electrical activity had led doctors at Great Ormond Street to believe that he had severe brain involvement by MDDS. After reviewing recent EEGs, Dr Hirano indicated to the court that "I can understand the opinion that he is so severely affected by encephalopathy that any attempt at therapy would be futile. I agree that it is very unlikely that he will improve with that therapy." However, he also felt that he couldn't rule out some benefit. The probability was low, but not zero. "I think to a large extent it is irreversible, but I cannot say it is completely irreversible" (para 118).

C. Costs of treatment

One issue, raised briefly in the judgement, but dismissed, was the question of the cost of treatment. Charlie's parents had sought funding to allow Charlie to travel to obtain nucleoside treatment overseas, since it was clear to them that the public health system in the UK would not provide treatment. By the time of the court hearing in April, their fundraising campaign had reached its target of £1.2million. However, the judge indicated that "funding was never an issue". Great Ormond Street had been prepared to provide nucleoside treatment to Charlie, but had changed their mind once he deteriorated and they felt it would not help him. The judge wrote:

"It is imperative that I make clear that this case is not about money and, if anyone were to suggest that Charlie would have nucleoside treatment but for the cost, they would be completely wrong."

(We will later respectfully disagree, at least in part, with this statement. See Chapter 6).

One of the important roles of ethical analysis in complex cases, is to separate out questions of fact, from those of value. The factual questions are going to be based on evidence, and scientific and medical experts are important in determining those. However, questions of value aren't going to be determined by evidence in the same way. To address those we will need careful ethical and philosophical analysis.

What were the key questions of fact in the Charlie Gard case? The table below summarises the main ones, and identifies areas of disagreement.[27]

27 We divide the opinions into the hospital and the family. The legal guardian, appointed to represent Charlie's interests largely concurred with the medical opinion about facts and ethical questions and concluded that "it is not in Charlie's best interests to travel to America to receive nucleoside therapy". For simplicity, we will not discuss separately the Guardian's perspective.

"It is with the heaviest of hearts but with complete conviction for Charlie's best interests that I … rule that Great Ormond Street Hospital may lawfully withdraw all treatment, save for palliative care, to permit Charlie to die with dignity." (para 23)

Charlie's parents cried out in dismay, and wept as the judge read out his decision in open court. The judge provided a detailed justification for his conclusion. He had been convinced by the medical experts at Great Ormond St that nucleoside treatment would be futile. By this he meant "pointless or of no effective benefit". He noted that the US expert Dr Hirano had conceded that the treatment was very unlikely to benefit Charlie. Justice Francis observed that there was agreement from all parties, including Charlie's parents that his current quality of life was so poor that it was not justified to prolong it without hope of improvement. He ruled that nucleoside treatment should not be provided, and that doctors could proceed with their plans to withdraw life-sustaining treatment from Charlie.

Legal appeals

After the April decision in the High Court, there were a series of legal appeals. Figure 1 shows schematically the current available options for resolving a disagreement about treatment for a child in the UK.

In most cases, a single court might be involved. In the Gard case, all possible legal appeals were pursued. The decision was reviewed in the Court of Appeal (initial decision upheld, 23rd May), Supreme Court (declined permission to appeal 8 June) and European Court of Human Rights (endorsed approach of UK courts 20 June). All of those courts ultimately supported the initial decision that it would be in Charlie's best interests to stop life support and not provide nucleoside treatment.

We are not going to review those court judgements in detail here. Much of the evidence presented at those court hearings repeated what had been discussed at the High Court. (In the UK, appeals courts typically allow appeal on the basis of claims of errors of law or of the judicial process. They do not usually re-assess the factual evidence presented and debated at the lower court. The Supreme Court will usually only allow appeals if there is a legal principle of general public importance at stake.) Much of the legal appeals therefore turned on points of law, for example questions of the jurisdiction of the court, or questions whether the High Court decision contravened elements of the European Convention on Human Rights. We will set aside those questions for legal scholars to debate elsewhere.

However, there was some discussion of two questions that are of clear ethical relevance.

Harm threshold

One of the grounds for the first legal appeal focused on when courts may override parental wishes. We noted above that the UK court becomes involved in decisions about medical treatment if parents and doctors cannot agree on what would be best for the child. However, Charlie's parents argued in the Court of Appeal that this is the wrong standard. Their lawyers claimed that:

"the court may not interfere with a decision by parents in the exercise of their parental rights and responsibilities with regard to their child's medical treatment, save where *there is a risk the parents' proposed course of action may cause significant harm*."[28] Para 54 [emphasis added]

One of the arguments that the legal team drew on in support of this claim drew an analogy with the legal approach to placing children in care. The UK 'Children Act' sets out when public authorities can remove a child from their parents. Social services in the UK can obtain a court order only if:

28 Yates & Anor v Great Ormond Street Hospital For Children NHS Foundation Trust & Anor (Rev 1) [2017] EWCA Civ 410.

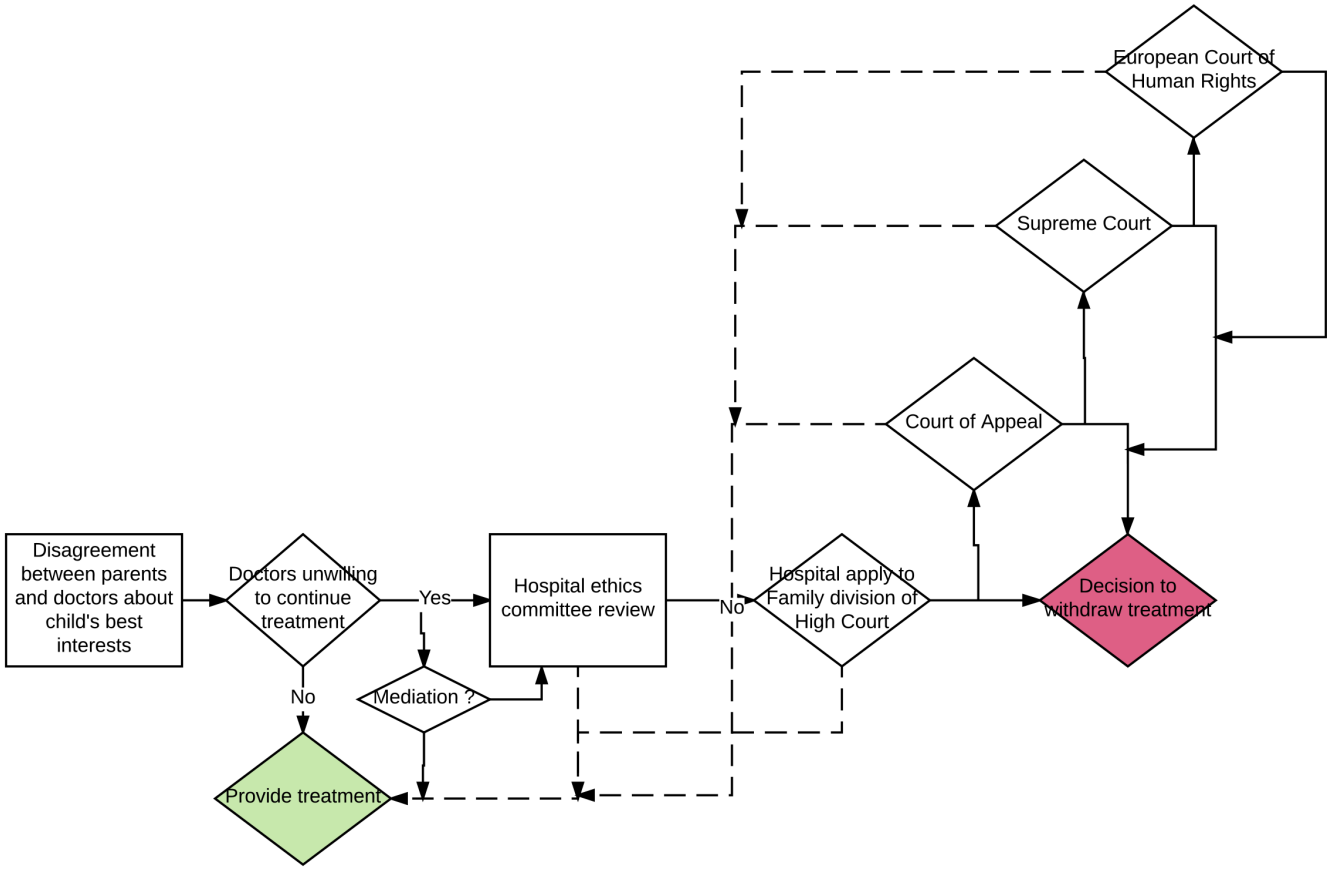

Fig 1.1. Current approach to resolving disagreement about medical treatment for a child in the UK.

"the court is satisfied that the child concerned is suffering or is likely to suffer significant harm, and that the harm, or likelihood of harm, is attributable to parental care or the child being beyond parental control"[29]

The idea is that public authorities can't intervene for a child any time that they feel that parents are doing less than the best for their child. All of us who are parents know, if we are honest, that we are not perfect, and there will be some times and some decisions where we do less than the best possible for our children. Perhaps that is because we have several children, and doing the best for one child means doing less than the best for another. Perhaps it is because our own work or our own interests are important too, and sometimes they take priority. Perhaps it is just because we all have times when we are tired, stressed, overworked, distracted, or just human, and fail to live up to the high standard of 'doing the best' for our children – much as we might aspire to that.

The invocation of 'significant harm' in the Children Act means that the state gives parents some leeway – it acknowledges that they have a right to care for their children in the way that they choose – within limits.

But if the 'harm threshold' is the standard for the state intervening in parental everyday decisions – shouldn't that also apply to medical decisions? Charlie's parents' legal team suggested that in cases where parents have an alternative viable treatment option to that proposed by doctors, the test shouldn't be whether one of these is 'better'. Instead, the parents' wishes should be overridden only if their preferred treatment plan would be harmful.

29 The Children Act, 1989. Available at: https://www.legislation.gov.uk/ukpga/1989/41/section/31

That was the argument put to the Court of Appeal. The judges in that court ultimately rejected that legal argument. As a point of law in the UK, medical treatment cases are treated differently from decisions about when local authorities can take over the care of a child.

That still leaves the ethical question unanswered – *should* we use the harm threshold for deciding when parents' wishes about medical treatment should be respected or refused? If we think the answer to that is right, it may be that the law should change. We will argue in chapter 8 that the harm threshold *is* the right way to set out the boundaries of parents' discretion about medical treatment.

As an aside, the judges in the Court of Appeal indicated that even if Justice Francis had been required to use a 'significant harm' test rather than a best interests test, that he would likely have reached the same decision.

Freedom to travel

One of the arguments discussed in the European court hearing, was whether the previous court decisions not to allow Charlie's parents to take him overseas for treatment deprived him of his liberty. Article 5 of the European Convention on Human Rights grants everyone a right to liberty and security. It gives individuals a right not be arrested or detained without an appropriate legal basis. In the press, it was claimed that the court decisions had effectively "imprisoned" Charlie in Great Ormond Street hospital.[30]

The specific legal claim about imprisonment was rejected by the European Court. (In essence, the convention doesn't stop countries from detaining individuals – it stops countries from detaining people without good legal reason, process and safeguards. The court felt that the reasons, process and safeguards articulated and followed by the UK courts meant that Charlie's Article 5 rights weren't being violated). However, the wider ethical question is one that we will return to. Particularly for cutting edge treatments, it is not unusual for treatment to be available in some countries but not others. Travel for treatment (so called 'medical tourism') is becoming increasingly common. When should, or shouldn't this be permitted?

The final appeal

After the European Court rejected his parents' appeal, it appeared that the final legal avenues for Charlie's parents had been exhausted. It seemed to all external observers that it was no longer a question of *if* treatment would be withdrawn from Charlie, but *when*.

However, the public and media attention to Charlie's case did not wane. In the last week of June, and first week of July, there were countless headlines, opinion pieces, comments as global attention focused on Charlie's plight. There were several very high profile statements of support. On the 2nd of July, Pope Francis released a statement indicating that he hoped that "their desire to accompany and care for their own child to the end is not ignored".[31] A day later, US President Trump tweeted "If we can help little #CharlieGard, as per our friends in the U.K. and the Pope, we would be delighted to do so." Later that week, a number of international medical and scientific experts came forward offering to provide treatment (in hospitals in New York or Rome) and offering apparently new evidence allegedly increasing the chance of benefit from nucleoside treatment. On the 10th July, Great Ormond Street Hospital elected to bring this evidence back to the High Court.

Over a series of hearings from 10th July until the 24th, Justice Francis again considered Charlie's case. The court heard further testimony from Dr Hirano by videoconference, and the judge then arranged for the US specialist

30 Mendick, R. (2017) Charlie Gard is being held 'captive' by the NHS, complains the family's spokesman. *The Telegraph*. accessed from < Available at: http://www.telegraph.co.uk/news/2017/07/12/charlie-gard-held-captive-nhs-complains-familys-spokesman/>.

31 Vatican Radio. (2017, July 3, 2017). 'Pope Francis expresses closeness to Charlie Gard's parents.' from < Available at: http://en.radiovaticana.va/news/2017/07/03/pope_francis_expresses_closeness_to_charlie_gard's_parents/1322731>.

to review Charlie in person in London as well as for further tests. The next week, several specialists (including Dr Hirano, but also other international experts) took part in a multi-disciplinary meeting. Following that meeting further tests were arranged to assess the severity of Charlie's illness including a full body MRI. On 24th July, Connie and Chris appeared in court and withdrew their appeal against the medical and legal decisions; they now accepted that further treatment could not help Charlie. Treatment was withdrawn later that week.

Cases like Charlie's offer an insight into the ethical dilemmas facing parents, doctors and courts. We have summarised in some detail the Charlie Gard case to provide the basis for further ethical discussion. We have explained some of the legal process and reasoning (though the law is not our main focus). We have also tried to put this extraordinary case into the broader context of most end of life decisions, as well as other cases of disagreement between parents and doctors.

Our aim in the next section of the book is to move beyond the specific details in Charlie's case. In doing so, our analysis will shed some light on disagreement itself – on its moral significance in culturally and ethnically diverse societies, and its implications for policy and practice. It is not surprising if difficult cases about providing or withholding life prolonging treatment divide experts and communities.

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