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CHAPTER 8

Pediatric Brain Tumors: Narrating Suffering and End-of-Life Decision-Making

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Abstract: When talking about decisionmaking for children with a life-threatening condition, the death of children with brain tumors deserves special attention. The last days of the lives of these children can be particularly harsh for bystanders, and raise questions about the suffering of these children themselves. In the Netherlands, these children are part of the group for whom a wide range of end-of-life decisions are discussed, and questions raised. What does the end-of-life for these children look like, and what motivates physicians and parents to make decisions that may affect the life and death of these children? This article highlights the story of the parents of the sisters Roos and Noor. When both their daughters were diagnosed with a hereditary brain tumor, they had to make similar decisions twice. Their story sheds light on the suffering of children in the terminal phase, and how this suffering may motivate parents and physicians to make decisions that influence the end of life of these children's lives.

We argue that complete knowledge about suffering in the terminal phase of children with brain tumors is impossible. However, by collecting experiences like those of Roos and Noor, we can move toward an experienced-based understanding and better guide parents and physicians through these hardest of decisions.

Introduction

No one is, or will ever be an expert when it comes to children dying. Physicians, no matter how medically skilled, will always lack significant intimate knowledge; for parents, these deaths are once-in-a-lifetime events. However, even lacking expertise, decisions need to be made that might influence life expectations.

The end of life for children with brain tumors deserves special attention. Their last days can be particularly harsh for bystanders, who feel powerless in the face of watching them display increasingly severe symptoms without being able to offer any good options. Moreover, the situation raises important questions about quality of life and suffering of these children, and the decisions that have to be made.

In the Netherlands, children with brain tumors are a significant part of the group of patients for whom end-of-life decisions¹ (ranging from withholding or withdrawing aspects of treatment to active life-ending) are being made and discussed.

Although there is a growing body of knowledge on how such decisions are made,^{2,3} what is needed is insight into what the experience means for those involved in making these decisions. The narrative side of pediatric end of life is a way to bridge that gap and provide insight into what it is like to live through these life-altering periods, and provide some understanding of how parents and healthcare professionals perceive the suffering of these children, and how this perception motivates them to make the decisions that affect their death.

Suffering for children with brain tumors

In the context of a more extensive study into decisions and care for children with a life-threatening condition in the Netherlands that will soon be published,⁴ we interviewed 64 parents of 44 children that lived or had lived with a life-threatening condition. Amongst those were the parents of 15 children who died from a brain tumor. Typical for children with brain tumors was the way these parents described suffering that occurs in the last phase of their life, when the mass effect of the tumor creates various symptoms, ranging from aching, nausea, and impaired

consciousness to epileptic seizures. While parents sometimes also saw other kinds of suffering in their children, it was this specific process in the terminal phase that motivated a significant number of interviewed parents to discuss end-of-life decisions. In several cases the decision was made to start terminal sedation. In a few occasions parents requested to actively end the child's life; but these requests were all denied.

Notably amongst them were the parents of two sisters: Roos and Noor. To provide insight into this unique perspective on suffering and the way it motivates decisionmaking, we used their story in this article to give insight into the death of children with neurological tumors. The reason why the story of Roos and Noor is of such importance is threefold: first, their story exemplifies the way parents see suffering in children with neurological tumors. Second, it is representative of how these perceptions motivate decisionmaking. And third, although their story is representative of the way parents see suffering, this story is even more poignant because for these parents, the death of a child was not a once-in-a-lifetime event. They had to face the same suffering, and the same decisions, twice, and their story gives much needed, intimate insight into the relation between the suffering of children with neurological tumors, and end-of-life decisionmaking.

The Story of Roos and Noor

Upon hearing the verdict that their youngest daughter Noor was suffering from a brain tumor with a very limited prognosis, her father expressed the following: "Doctor, we were thinking about Wednesday." The physician was baffled. "Wednesday, what?" he asked. "To let her die, on Wednesday," the father answered.

The painful story of this family begins with Roos, the elder sister. When a prenatal scan of Roos showed slightly enlarged brain ventricles, the suspicion of an ominous medical condition was an immediate concern. Three weeks after her birth, the cause was determined: a malignant and aggressive atypical teratoid rhabdoid brain tumor.

For a year, Roos underwent surgery and chemotherapy to which she responded very well; and, for a while the tumor seemed to be successfully removed. The parents recall her during this period as a happy child, who practically hopped into hospital for her check-ups. Up until that point, her parents remembered her life as one with relatively little pain, although her mother underlines that it is very difficult to know for certain, as Roos was a child who would never cry or complain when hurt:

“She never cried from the time she was operated on at three months old. Since crying is the only thing babies can do, I’m not sure why Roos couldn’t or wouldn’t cry any more. You could theorize that maybe she felt that she had been through so much already, there was just no point in crying. I have no idea whether children can have such realizations, but we never found an explanation as to why she did not cry.”

However, after several months, brain scans revealed that the tumor has relapsed. Seeing how well Roos had responded to the first treatment, and because at that time, there were no signs of the tumor spreading, treatment was started again, this time with the addition of radiation therapy. Looking back, her parents say that there was hardly any discussion about this decision: “Everyone agreed that we should give her this chance, seeing how well she had responded to her first chemo treatment.”

Three weeks after the final day of her radio- and chemotherapy, Roos’ condition suddenly deteriorated. The first symptoms of her discomfort were fatigue, nausea and an unwillingness to eat. Suspecting a case of influenza, Roos was admitted to the local hospital. But her symptoms turned out to be the terminal stage of Roos’ illness.

The final stage of Roos’ life is not what her parents would have hoped. During the second night in the local hospital, her symptoms worsened, and Roos unexpectedly became unresponsive. She was transferred to the Intensive Care Unit of the academic hospital, where initial brain scans indicated the possibility of metastatic tumors in the brain ventricles and spinal cord.

Before a final scan the next day could confirm this suspicion, the mass effect of the metastatic tumors had started to put pressure on nerves and brain tissue and cause seizures. Roos was screaming and over-arching her entire body as a side effect of the seizures. Throughout this

traumatic time, her parents were unable to comfort Roos, as it was impossible to hold or make contact with her:

“We didn’t know what to do, we were sitting at her bed, and we were panicking because she kept screaming, and we couldn’t calm her down. So the nurse suggested to wait in the hallway, but every time the door opened, we heard her screaming again. [...] and at that time I didn’t understand why they wouldn’t do anything and kept thinking, How is it possible that we are on an ICU, and no one does anything?”

Finally, physicians decided to temporarily sedate Roos and make further scans to confirm the suspected metastases, but Roos did not wake up from her coma. At age two, she died in the ICU.

After receiving several reassurances that the condition was not hereditary, the parents welcomed a second child, Noor. Sadly, during the pregnancy, ultrasound scans showed that Noor had similarly enlarged brain ventricles, suggesting the same brain tumor as Roos. Due to growing concerns about her health, a caesarian section was performed at 34 weeks. Just one day after her birth, scans confirmed that Noor had inherited the same condition as her sister. The mother remembers lying in bed, still exhausted from the caesarian section, when a delegation of doctors walked in to deliver the bad news. The parents were devastated.

Soon after, there was further confirmation that due to the spreading of the tumor, there was no chance that Noor would survive. The parents, having lived through the suffering of their eldest daughter, decided to ask that Noor be spared the suffering they saw in Roos’ final days. This is when the father said: “Doctor, we were thinking about Wednesday.”

Father: “I think the physician was a bit shocked by our bluntness. But we had the history with Roos [...] and with the prognosis being as it was, we wanted to spare Noor that suffering.”

Mother: Why do you have to watch a little girl’s condition deteriorating, if you know she will die? Why can’t you grant her a soft and dignified death? Because that feels like the only thing you can still do for her.”

This story shows the importance of the narrative behind the parents' request, and their existential understanding of the unbearable suffering that motivated their wishes. Without context, the parents' appeal for euthanasia for Noor may have seemed abrupt—perhaps even inconceivable—but from a view of their experiences, we can understand what they hoped to avoid for Noor.

At the time of these events, unlike her older sister Roos, Noor was less than a year old. For children between 1 and 12, actively ending life, euthanasia,⁵ is not legalized.⁶ However, for younger children like Noor, active life-ending is permitted under strict circumstances first described in the Groningen Protocol.^{7,8,9}

In Noor's case, thinking euthanasia might be illegal, the physicians denied the parents' request. Instead, in an effort to prevent the parents' fear of witnessing another traumatic death, they turned to a plan to provide Noor with palliative sedation.

Noor's life was short, she lived to be almost 8 weeks old. With the help of the physicians her parents tried to create good memories at home during this time, even though they had initially wanted to stay with her in the hospital:

“Out of fear of what was to come, I was so scared to take her home, but the hospital staff said: ‘We’re not saying that you should, but please think about it a bit longer. [And our pediatrician] said: ‘If you want to go home, I will come with you, to make sure you feel safe enough to take her home.’” And we responded ‘Wow, you would do that, for us? It made us feel so understood.’”

Her parents are now grateful to have those memories of Noor in their family home. But as effects of her tumor became visible, (and, although her parents remember her as looking comfortable) the tumor caused hydrocephalus, and her mother questions if this may have caused Noor to suffer:

“At some point, her head had grown so much, that it was difficult to hold her, she couldn't wear normal clothes because they wouldn't fit over her head, and she couldn't turn or lift her head very well... So those were the things that made us think: ‘that must be terrible for her. But of course that is debatable, because she did not know any better.’”

When increasing symptoms signaled that Noor's life was coming to a close, parents and physicians together agreed to begin terminal sedation.

Mother: "We took her home for one last night, and she slept between us. And the next day we brought her to the hospital. [...] They started the sedation, and she fell asleep peacefully. And then we waited, laying her on my chest, and her dad's.

The pediatrician had told us what the signs of dying would be. There were several hours of gasping, and moments when we thought it was over. But finally, at 20:30 she passed away.

Father: "And then you take your deceased child home, in the same stroller that you brought her in that morning, you take her home in the evening."

End-of-life with a brain tumor: Suffering for a child?

The end of life for Roos, with her impaired consciousness, seizures, and related signs of discomfort, is unfortunately not uncommon for children with neurological tumors.¹⁰ Although the experience was clearly traumatic for the parents, the question remains as to what we can say about the suffering of these children.

In the final stages of a neurological tumor, various symptoms can arise, ranging from headaches and nausea, to seizures and over-arching, as was seen with Roos.¹¹ These symptoms are caused by the mass effect of the tumor, pressing on nerves and brain matter. The general consensus amongst physicians is that in the final stages, when children are having seizures, children lack the awareness to actively experience suffering.¹²

From a philosophical point of view, however, that is not the end of the question. In both the medical and the philosophical contexts, the concept of suffering is somewhat underdefined (it is, for example, notably lacking an entry in the *Stanford Encyclopedia of Philosophy*¹³). In the medical setting, the commonly used definition dates back to the 1990s, when Eric Cassell described suffering as: 'the state of severe distress associated with events that threaten the intactness of a person.'¹⁴ Although the literature has since both criticized and refined this definition,^{15,16,17,18} Cassell's definition remains the one most often used and cited.

With the phenomenological experience (the state of distress) as one of the defining qualities of suffering, we have to keep in mind that we can never have complete access to the qualitative experiences of these children. Although there is evidence that children do not suffer from their seizures since they do not actively experience them,¹⁹ authors such as Thomas Nagel and David Chalmers argue that, even if we could have complete and perfect access to the brain activity of these children in their final stages (which in itself would present significant ethical problems), their qualitative experience would still elude us.^{20,21} Therefore, the conclusion that children themselves do not actively suffer from the brain tumors, is never a fact we can take for granted.

However, suffering is not merely a question about if these children actively have these phenomenological experiences. For the parents of Roos and Noor, as it is for many of the interviewed parents, suffering does not only consist of the possibility that the children might actively experience the symptoms caused by the mass effect of the tumor, it also consists of the absence of a dignified death, or the ‘futility’ of a child having to go through those symptoms at the end of their life. As the mother stated earlier: “Why do you have to watch a little girl’s condition deteriorating, if you know she will die? Why can’t you grant her a soft and dignified death? Because that feels like the only thing you can still do for her.”

But is this suffering? We might argue that an undignified death as a form of suffering does tie in with the second defining feature of Cassell’s concept of suffering: the intactness of the person.²² However, there is no definitive answer to that question. Suffering is not merely a factual question, it has semantic aspects as well, i.e., how to demarcate suffering. That can only be decided by consensus. The fact that parents perceive it as a form of suffering that even motivates them to consider end-of-life decisions, is an indication that we at should least debate the possibility.

Decisions at the end of life: A matter of regulations?

We have chosen these parents and their children to address the importance of narrative with regard to pediatric neurological suffering. For the parents of Roos and Noor, their experiences pushed them toward a request that goes against every instinct and wish of any parent: “Can you

end the life of our daughter Noor?’ Their appeal went against their own heartfelt wish to keep their daughter with them just a little bit longer; but came from wanting to protect Noor from the suffering they saw in their eldest daughter, Roos.

Children with neurological tumors constitute a significant number of the children for whom end-of-life decisions are often discussed, but perhaps what we really should talk about is: What is suffering for these children (not to be confused with the equally important debate on how parental suffering should be regarded in decisionmaking)? For suffering lies at the heart of pediatric palliative care; all definitions of pediatric palliative care and end-of-life decisionmaking focus on the relief and ending of suffering.^{23,24}

Decisionmaking, whether it is about withholding or withdrawing kinds of treatment, terminal sedation, or euthanasia, is not merely a case of regulations. In Noor’s case, euthanasia might have been permitted in cases of hopeless and unbearable suffering,²⁵ but the parents’ request was denied by the physician. It is possible but unlikely that the physician was not aware of the regulation. It is much more likely—and this is something that we often come across in our interviews with physicians—that the physician had difficulties in weighing Noor’s suffering, or demarcating it as unbearable and hopeless. We speculate that this might also play a factor in the fact that since installment of the formal regulations on active life-ending for newborns in 2007, only three cases have been reported.²⁶

From the Dutch pediatric point of view, any kind of end-of-life decisionmaking is not merely a matter of whether it is permitted by law, but also of a weighing a child’s suffering and quality of life against the possibility of relieving this suffering in any way.²⁷ In the Dutch context, it is emphasized that this is a decision that should always be made with parents.^{28,29}

It is the contention here that rather than merely legislating and leaving physicians alone with the question of how to use the law, we need to widen the discussion to include a closer examination of what suffering actually entails, when it becomes unbearable—and for whom? More often than not, the kind of suffering that occurs in situations where parents and physicians have to make these decisions, is not clear-cut. It is the kind of suffering we saw in Roos and

Noor: suffering that is not merely about pain, but also about dignity, social aspects, and lack of quality of life in all its facets.

The evidence-based scientists in us may want to wish for more absolute protocols, and find that these do not exist when it comes to suffering; the philosophers may want to wait for more information before drawing conclusions, and find that reality does not allow us that luxury of time. We have to give more meaning to the concept of suffering by seeing the various ways that children can suffer when confronted with a terminal illness.

Although a perfect understanding of suffering in children will be impossible, an experience-based conceptualization can help with making better decisions. Although regulations on decisionmaking can aid physicians with not standing empty-handed in cases of severe suffering, they need to be accompanied by guidance on how to interpret the suffering of these children. The ‘suffering’ of children with brain tumors might not fit our traditional understanding of the concept. We need to have conversations about what suffering is before we can even discuss legalizing end-of-life decisions such as euthanasia. By collecting and analyzing more stories like those of Roos and Noor, we gain more insight into the variety of situations that parents see as suffering, and the how it determines their decisionmaking.

Endnotes

1. End-of-life decisions are understood here as medical decisions that may have life-limiting consequences; either intentionally (active life-ending), or as an anticipated but not intended effect (such as DNR, withholding or withdrawing treatment, or terminal sedation).
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5. In the Dutch context, the term ‘euthanasia’ only applies to cases where the patient (i.e., the child) would make the request. When parents make the request, it is referred to as active life-ending.
6. While there is no regulation permitting active life-ending or euthanasia for children between ages 1 and 12, there might be a legal possibility for active life-ending for these children in the form of grounds for exemption from criminal liability. One of these grounds is ‘emergency,’ specified as the conflict of obligations. The conflict that a doctor might experience between his duty to relieve the suffering of a child and the law, might be such a conflict. There have been successful appeals based on this, with adult patients. These grounds might apply to cases with children as well, but so far no one has openly reported a case.
7. Government of the Netherlands. Euthanasia and newborn infants. 2017; available at <https://www.government.nl/topics/euthanasia/contents/euthanasia-and-newborn-infants> (last accessed 23 Dec 2018).
8. Verhagen EEE. The Groningen protocol—Euthanasia in severely ill newborns. *New England Journal of Medicine* 2005;352(10):959–62.
9. Despite fears that the legalization would lead to a slippery slope, since the installment of the formal regulation in 2007, only three cases have been reported, none of them for children with brain tumors. See: Beoordelingscommissie LZALP. Jaarverslag Beoordelingscommissie Late Zwangerschapsafbreking en Levensbeëindiging bij Pasgeborenen 2017. [Annual report on late termination of pregnancy and life-ending in newborns, 2017] available at <https://www.rijksoverheid.nl/ministeries/ministerie-van-volksgezondheid-welzijn-en->

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 12. See note 11, Goldman et al. 2012.
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 22. See note 14, Cassell 1991, at 33.
 23. WHO. Definition of Palliative Care for Children; available at <https://www.who.int/cancer/palliative/definition/en/> (last accessed 30 Dec 2018).
 24. Nederlandse Vereniging voor Kindergeneeskunde (NVK). Richtlijn Palliatieve Zorg voor Kinderen [Guideline on Palliative Care for Children], 2018; available at http://richtlijndatabase.nl/richtlijn/palliatieve_zorg_voor_kinderen (last accessed 12 Mar 2019).
 25. Unbearable and hopeless suffering is one of the five due care criteria in the Dutch regulation on active life-ending in newborns. See: Governemnt of the Netherlands Euthanasia and

Newborn infants. 2017; available at <https://www.government.nl/topics/euthanasia/euthanasia-and-newborn-infants> (last accessed 12 Mar 2019).

26. Beoordelingscommissie LZALP. Jaarverslag Beoordelingscommissie Late Zwangerschapsafbreking en Levensbeëindiging bij Pasgeborenen 2017 [Annual report on late termination of pregnancy and life-ending in newborns, 2017]; available at <https://www.rijksoverheid.nl/ministeries/ministerie-van-volksgezondheid-welzijn-en-sport/documenten/rapporten/2018/10/02/jaarverslag-beoordelingscommissie-late-zwangerschapsafbreking-en-levensbeëindiging-bij-pasgeborenen-2017> (last accessed 8 Mar 2019).
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28. See note 24, NVK 2018.
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