ABSTRACT

Title of Dissertation: VULNERABLE PATIENTS, AUTONOMY, WELL-BEING, AND DEATH

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At the beginning of our lives and often at the end, we have important medical decisions made for us by proxy consenters including family, legal guardians, and/or medical professionals. This places us in particularly vulnerable and dependent positions that essentially 'bookend' our lives. As a bioethicist, I view as among my duties working to improve the experience of medicine for vulnerable populations as well as advocating for protections for such patients against the poor decision-making of others (and, in rare exceptional cases, even themselves).

I've opted for a 'covering concept' model for my dissertation, which consists of three sizeable papers on related topics. The vulnerable populations I focus on in this project are children, the mentally ill, and the elderly. All three of these papers touch on issues surrounding the authenticity and limits of informed consent, tensions between respecting patient autonomy and promoting patient well-being, and how best to face death.

In *How to Face the Future: A Model for Delayed Disclosure of Incidental Findings from Pediatric Whole Genome Sequencing*, I argue that in cases of widely-focused pediatric genetic testing, consent for release of a limited class of incidental findings should be delayed until the pediatric patient or research subject reaches the age of majority. I also propose a model for delayed disclosure in such cases.

In *Early Palliative Sedation Therapy and the Challenge of Psychological Suffering*, I make the case that current end of life palliative care practices in the United States rationally commit us to the moral permissibility of palliative sedation to alleviate refractory psycho-existential suffering, even in cases where death of the patient is far from imminent.

In *Cardiac Pacemakers and Withdrawal of Care at the End of Life*, I make the case that deactivation of cardiac pacemakers is morally distinct from typical instances of withdrawal of care at the end of life. I argue that in highly dependent patients, pacemaker deactivation is morally akin to voluntary active euthanasia, while in non-highly-dependent patients, pacemaker deactivation only serves to lessen the patient's quality of life unnecessarily.

VULNERABLE PATIENTS, AUTONOMY, WELL-BEING, AND DEATH

by

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Dedication

Dedicated to J.B. for his unequaled ability to boost morale and inspire.

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Table of Contents

Introduction

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HOW TO FACE THE FUTURE: A MODEL FOR DELAYED DISCLOSURE OF INCIDENTAL FINDINGS FROM PEDIATRIC WHOLE GENOME SEQUENCING

1. Introduction

Whole genome sequencing (WGS) is rapidly becoming more affordable for use in clinical medical settings. At a price of around $1000, WGS is now barely more expensive than more targeted testing and is only growing cheaper as technology advances.[1] Further, while the genetic bases for many diseases currently remain opaque, it is not difficult to imagine a future where we can predict with great accuracy an individual's likelihood of developing a wide range of diseases based on personal genome analysis. It is the likelihood of this future that makes it necessary to figure out how to deal with such a substantial influx of information and, in particular, determine who ought to receive it.

In this paper, I advocate for delayed disclosure of incidental findings which may arise in broad pediatric genetic testing such as WGS, concerning a particular subclass of conditions. This subclass of conditions contains incurable or highly unlikely to be cured adult-onset disorders which cannot be prevented or mitigated by action in childhood (i.e. prior to clinical presentation of the disorder). I will argue that delaying disclosure of this specific range of incidental findings until pediatric patients reach the age of majority is more consistent than

[1] See Dewey et al. 2014

alternatives with what we morally owe to children, in terms of respecting their autonomy and promoting their well-being.

An adult facing the decision of whether to undergo WGS has difficult questions and trade-offs to consider during the consent process. There may be some very good reasons to undergo testing. Learning of one's carrier status for particular genetic mutations can help to inform reproductive decisions, and opting to be tested on the basis of knowing that a family member is a carrier of a particular disease can serve to alleviate the anxiety of uncertainty. However, if one undergoes WGS and discovers a high likelihood of developing a specific fatal genetic disease, this saddles one with the knowledge of how and even when one will likely die, often with an accompanying prediction of future suffering. This knowledge has the potential to impose a substantial psychological burden on those who possess it. Going through life with the knowledge of even a relatively low likelihood of developing any number of horrific diseases has the potential to cast a pall over years of perfect health. Aside from concerns regarding the psychological well-being of the tested patient, there is the sobering possibility that unanticipated genetic information might lead to employment discrimination or a denial of insurance coverage.

There may be many reasons to opt for WGS to intentionally answer some specific genetic questions, but what are we to make of *incidental* findings? Incidental findings are those that are not of primary or direct relevance to the purpose for which testing was undergone. Whether findings are incidental will be dependent upon the broadness of the aims of a given instance of testing. If

someone wants her genome mapped simply in order to obtain her genetic information in its entirety, no findings would be incidental. If someone were getting tested for BRCA mutations (mutations associated with an increased risk of breast and ovarian cancers) specifically, then any findings not directly related to the presence or absence of such mutations would be incidental. And, even if whole genome sequencing or other broadly-targeted genetic testing were undergone *as a means to identify the presence of relevant BRCA mutations*, any findings unrelated to BRCA mutation would be incidental, regardless of the fact that a large amount of data was generated. This is because that data was generated with the specific aim of identifying a small range of genetic anomalies. With WGS, the sheer volume of information generated means that the likelihood of incidental findings (if the data generated is analyzed) approaches 100%. If these findings have serious clinical import and are disclosed to the tested patient, she may be confronted with knowledge regarding her own genetic makeup that is unprecedented – as such information would have been impossible to obtain in even the quite recent past – and potentially life-altering. Determining appropriate rules of disclosure for incidental findings is thus one of the pressing problems raised by the growing prevalence of WGS.

WGS is now seen as a viable and exciting option for use in pediatric clinical practice and research.[2] While it is still far from common in clinical

[2] For example, pediatric genomic sequencing programs are in place at Boston Children's Hospital, NewYork-Presbyterian Morgan Stanley Children's Hospital, The Children's Hospital of Philadelphia, Children's Hospital of Wisconsin, and

practice, it is not difficult to imagine a future where WGS for fetuses and/or neonates becomes the norm. My concern in this paper is *not* with how best to disclose incidental findings to *adult* patients and research subjects. This is because adults can give genuinely informed consent for release of such findings, and so answering the question of which method of disclosure is best will be basically a matter of determining how to weigh the significance of findings and then effectively communicate the potential costs and benefits of disclosure to the patient. This may be a challenging and interesting endeavor, but I want to focus here on a different question: that of how best to disclose incidental findings which arise in the course of *pediatric* WGS. In such cases, parents or guardians must act as proxy consenters on behalf of their children. This leads to some unique challenges when it comes to determining how best to disclose incidental findings. Consider the following (hypothetical) motivating case:

> Five-year-old Ari is set to begin peewee soccer in the fall. Having heard about recent sudden cardiac deaths of child athletes, his concerned parents enroll him in a cohort study in which children of his age will undergo whole genome sequencing to search for the genetic mutations associated with long QT syndrome and related conditions which can lead to sudden death. Ari's parents give informed consent on his behalf, and Ari gives his assent to participate in the study. In the course of the study, researchers discover genetic mutations associated with Huntington's disease: a fatal

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Children's Mercy Hospital: Kansas City.

> degenerative disorder that will first manifest in mid- to late adulthood. There is no way to prevent or pre-emptively ameliorate this disease and it is unlikely that a cure will be developed within Ari's lifetime.[3]

This exemplifies the sort of situation with which I am concerned: WGS of a child incidentally reveals strong indicators of an adult-onset disease which is incurable or highly unlikely to be cured and of which there is no way to prevent or preemptively ameliorate the effects (medically speaking). Rosamond Rhodes (2006) provides us with a representative list of such diseases: Huntington's disease (HD); early-onset Alzheimer's disease; and the dominant variant of Charcot-Marie Tooth disease.[4] All of these diseases can be identified through genetic testing, first present in adulthood, are incurable or highly unlikely to be cured, and have symptoms that can only be managed or ameliorated once these disorders present clinically.

The question here is not whether to reveal incidental findings to Ari but rather whether to reveal such findings to Ari's parents. Ari's parents would then decide whether and when to reveal the findings to Ari. Is there moral reason to disclose this incidental finding to – or withhold it from – Ari's parents? Should Ari's parents be the ones tasked with deciding whether to have such findings revealed? If not, what then? These are questions I aim to answer in this paper.

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[3] Recent advances in the treatment on Huntington's disease are quite promising (see, e.g., Gallagher 2017), but for the purposes of this example, let us suppose that a cure does not seem likely to be developed within Ari's lifetime.

[4] (Rhodes 2006:210)

I will make the case that delaying the disclosure of incidental findings concerning many incurable or highly unlikely to be cured adult-onset diseases until the pediatric patient reaches the age of majority and can decide for herself whether to receive them is the best way to respect the autonomy and promote the well-being of children like Ari. However, delay does come with the risk of potentially being unable to re-contact patients and potentially poses a burden on our healthcare system as re-contacting may prove expensive, time-intensive, and logistically-challenging. These are concerns (among others) which I will be addressing in this paper.

I am preceded in this effort by the excellent work of Abdul-Karim, Berkman, Wendler, and colleagues. I take their ethical framework and guidelines for when to *immediately disclose* incidental findings produced from pediatric genetic testing to parents to be correct. However, I will here expand upon two main elements left underdetermined by their account: (1) in-depth theoretical justification for the importance of the moral considerations they appeal to in support of their account, and (2) an account of *which* incidental findings ought to be released to pediatric patients once they reach the age of majority, and *how* these findings ought to be released.

Abdul-Karim and colleagues appeal to the importance of these three moral considerations in arguing for a certain class of incidental findings to be immediately disclosed to the parents of pediatric patients: benevolence, duty to warn, and autonomy. However, they do not explicate these concepts or explain in detail their importance to the problem at hand. I will here explicate two relevant

moral considerations: promotion of well-being and respect for autonomy. I take these to contain the moral considerations Abdul-Karim and colleagues appeal to, as benevolence and duty to warn have fundamentally to do with promoting patient well-being. In what follows, I will first argue in favor of my account of delayed disclosure on the basis of its being most consistent with respecting the autonomy and promoting the well-being of pediatric patients. I will then propose a unique framework for delayed disclosure of incidental findings from pediatric genetic testing which indicate the tested patient will develop adult-onset disorders like Huntington's disease, early-onset Alzheimer's, and the dominant variant of Charcot-Marie Tooth disease.

2. Promoting Well-Being

Let's first consider how unexpected genetic information might compromise a child's well-being. If releasing incidental findings to children or their parents would harm the children (either directly or indirectly through their families), this is a consideration that should be taken seriously and weighed against the potential benefits of disclosing such findings. Of course, failing to release certain incidental findings has the potential to cause significant harm to pediatric patients. This is why it is important to take into account the importance of a duty to warn, which may pull us toward disclosing such findings. Broadly construed, the duty to warn applies when there is a foreseen harm or likelihood of some harm that will come to an individual. In medical cases, the duty to warn applies to risks of drugs and interventions, as well as discoveries regarding the patient which may potentially significantly affect the patient's well-being. The

notion of a duty to warn regarding genetic information normally has to do with the question of whether medical professionals have the obligation to warn potentially affected relatives of patients who have tested positive for a hereditary genetic disorder.[5] Generally, in cases where a patient opts to not warn a family member, patient autonomy and confidentiality trump the importance of the risk to family members.[6] In the case of pediatric WGS, the situation is a bit more complicated. This is largely due to the nature of proxy consent. If the patient is not able to give informed consent to receive her own medical information, then does the duty to warn the patient simply become a duty to warn the parents? This depends upon the nature of the information to be disclosed, but generally the duty to warn will apply to parents acting on behalf of their children.

So what type of information merits warning parents in cases of incidental findings which arise from pediatric WGS? This will be determined by the sort of findings discovered in the course of testing. If by taking immediate action a potential medical harm could be prevented or ameliorated, then the healthcare workers involved have a duty to warn the parents or guardians of the child in question. For instance, if a genetically-rooted disease could be prevented by prophylactic treatment and/or lifestyle changes, then it would be negligent of the healthcare workers involved to fail to warn the parents of this. In cases like Ari's, however, it seems that the duty to warn would only apply if there were something

[5] For information on relevant court cases dealing with this issue, see McAbee and Sherman 1998; Offit et al. 2004

[6] See, e.g., Dugan et al. 2003; Falk et al. 2003

that could be done medically to prevent or ameliorate the effects of Huntington's disease. In his case, given that no cure seems forthcoming within Ari's lifetime, a duty to warn seems to drop out of the moral equation.

There are essentially two scenarios for someone like Ari if his parents were given the information regarding his definite likelihood of developing Huntington's disease. Either his parents don't tell Ari, ever, or they choose a certain time to tell him. Either way it's not difficult to imagine how this would have a detrimental effect on Ari as he grows up. If his parents never tell him and instead quietly save up for his future medical expenses, there is still the likely chance that they will treat him significantly differently in light of knowing that he'll almost certainly die prematurely. Now, this might be a good thing. After all, perhaps knowing his life will be cut short will give Ari's parents a greater sense of every moment being precious and will help them pay better attention to Ari and appreciate his company more than they would have otherwise. It is just as likely – and I think probably more likely – that this knowledge would lead to significant familial stress and over-protectiveness or over-indulgence. One might respond that there are certainly parents who will be able to keep their emotions in check and raise a child like Ari as they would otherwise, but I am skeptical that this would be the norm.

If Ari's parents do opt to tell him at some point, Ari will have to face the fact of his premature mortality and find a way to psychologically adjust to the fact that his life will fall far short of the standard lifespan. Ari will likely either adjust his expectations regarding his future, or willfully ignore the fact that he won't be

able to have the same sort of lifespan and life narrative that most other people are able to experience. These imagined effects assume that Ari's parents will have waited to tell him until he's old enough to process the information in a generally rational manner. If his parents were to choose to tell Ari at age five that he'll eventually develop and die from Huntington's disease around middle age, it's unclear how this would affect him, although I would wager that, to the extent he could understand it, this information would be traumatic to Ari.

There is a serious lack of solid empirical literature regarding how children and family units react to unexpected genetic information.[7] However, we can tentatively extrapolate from the literature we do have to make a reasonable guess as to how incidental genetic findings of a serious sort might impact pediatric patients and their families. There are two main concerns here that must be examined: concerns regarding adverse *psychological* effects on the child and concerns regarding adverse *social* effects on the child. In reality, these will likely only rarely come apart. After all, social difficulties will often have a negative impact on psychological well-being and vice versa. However, it is worth considering these concerns each taken on their own, although there will inevitably be some overlap in terms of effects.

[7] I agree with the following evaluation of the empirical landscape: "Currently, there is insufficient evidence to inform a nuanced understanding of how children respond to genetic testing. This suggests a strong need for further research that uses rigorous approaches to address children's emotional states, self-perception, and social wellbeing." (Wade et al. 2010)

Let us first consider how children themselves might respond to results of predictive genetic testing. While we may not have information on how children as young as Ari might respond to such information, there have been studies regarding how adolescents respond to the results of such testing. However, empirical research on how adolescents react to the results of predictive genetic testing is messy and inconclusive. It seems that there are well-being related benefits that come about from learning about one's carrier status for particular diseases, but this can also negatively impact one's well-being. Finding out one's status can impact how one relates to family, plans for the future, and can substantially alter one's self-conception. Responses to the results of genetic testing tend to be neither straightforwardly positive nor entirely negative. This is true whether or not the patients who opted for predictive genetic testing turned out to be carriers for, e.g., Huntington's disease or BRCA mutations.[8] And further, these studies concern adolescents who are at or near the age of majority and who themselves opted for predictive testing. Oftentimes predictive testing is undergone because a relative is a carrier for a particular genetic disorder or testing seems indicated for some other clinical reason. In such cases, what is being tested for is known, and the main psychological difficulties which come about often involve interpersonal and identity issues (e.g. relating to family members who are carriers). Opting for predictive testing seems to set some clear expectations in place, and even if one reacts to results differently than anticipated, there is still

[8] See Duncan et al. 2008; Duncan et al. 2007

some base level of preparation and setting of expectations that seems to temper the experience of receiving the results of predictive genetic testing.

It is unclear, however, what impact the unanticipated discovery of incidental findings would have on these same adolescents who chose predictive testing for a specific disorder. After all, it seems that wholly unanticipated genetic information would be far more shocking and generally less welcome than genetic information that was specifically sought out. And, further, when we turn to the sort of cases I am concerned with, those which involve a child with parents acting as proxy consenters, the situation becomes even more complicated. This is because we must take into account both how the information might impact parents and how it might impact children if those parents opt to disclose the findings to them. It is not an impossibility to think that empirical research might come out showing that receiving incidental findings from genetic testing which indicate a disorder like Huntington's disease or early-onset Alzheimer's in a child would somehow make the child's and the family's lives as a whole demonstrably better in terms of well-being than they would have been without such knowledge. However, this doesn't seem likely. If responses to specifically sought-out genetic information may be extremely mixed, complex, and involve a lot of ambivalence on the part of many of those tested, there is reason to think that unanticipated and unrequested genetic information would be even more difficult to process, and I would posit, would probably have a more straightforwardly negative effect on the

well-being of the parents and patients who receive it.[9] However, more empirical research is needed to establish this with certainty.

Since it is the parents who would receive incidental findings in cases like Ari's, it is worth additionally considering how knowledge of Ari's eventual Huntington's disease might affect the dynamics of the family as a whole. One study of particular relevance to the question of how incidental findings of conditions like Ari's might impact a family unit concerns how parents (and the children themselves) cope with the prospect of a child dying prematurely. This study, conducted by Green and Solnit in 1964, focused on so-called "Vulnerable Child Syndrome" and looked at the behaviors of both parents of children expected to die prematurely and of the children themselves. This was a highly-subjective observational study and the sample size was small, but it's nonetheless important to consider since it is so specifically focused on the element of anticipated premature death of a child and its impact on family dynamics. Unsurprisingly, anticipating one's child's premature death is correlated with anxiety and depression on the part of parents. Fear of one's child dying was also correlated with overprotective behaviors on the part of parents as well as social and

[9] See, e.g., Duncan et al. 2007: 1988 and Duncan et al. 2008:53 for discussion of how predictive genetic testing seems to have a mixed impact on adolescents; solving some existing problems and alleviating some existing anxieties while at the same time the testing itself may generate new problems and anxieties for the patients who undergo it.

behavioral problems for the children.[10] This study was performed quite a while ago and relies quite a bit on personal evaluations (both on the part of researchers and participants) but the results line up with common sense in a way that seems plausible.

And, although there is clearly a difference between present chronic illness and anticipated illness, the literature on the impact of chronic illness of a child on families is worth considering, since we don't have much to refer to when it comes to the very specific and quite new issue of unanticipated discovery of an adult-onset genetic disorder in a child. Chronic illness is associated with stress in families across the board.[11] I posit that anticipating the difficulties that come along with degenerative genetic illnesses would also place stress on parents who know that their children will develop such an illness.

There is a serious lack of research on the impact of receiving incidental findings concerning adult-onset disorders like Huntington's Disease on the patient and her family. So, let's focus on one sort of degenerative disorder regarding which we have many years of data and which may also be identified in childhood and before symptoms of the disorder manifest, Cystic Fibrosis, "a serious life-threatening disease, leading to malnutrition and chronic lung infection."[12] Cystic Fibrosis (CF) is similar to other degenerative disorders in that it characteristically

[10] See Green and Solnit, 1964.

[11] See, e.g., McClellan and Cohen 2007; Holryod and Guthrie 1986; Hamlett et al. 1992; Holden et al. 1996

[12] Mérelle et al. 2003:346

starts out asymptomatic to mild and progresses in severity over time.[13] Neonatal screening often catches Cystic Fibrosis before the patient manifests any symptoms.[14] This leaves parents with the task of planning how to manage symptoms as they develop and monitor their child's progression and health state closely.

Knowing that a child will develop CF is similar to knowing that a child will develop Huntington's Disease (HD) in some relevant respects, as both are degenerative disorders that (in HD cases and for some forms of CF) eventually lead to death in middle age. Although patients with CF characteristically begin manifesting symptoms in childhood and patients with HD do not manifest symptoms until the disease presents in middle age, there may still be a long period of time between a diagnosis of CF and manifestation of symptoms. This period of time before symptoms present in some CF cases is roughly analogous to the

[13] "Most patients with cystic fibrosis have gradual but progressive deterioration in pulmonary and gastrointestinal functions." (Cowen et al. 1986: 745)

[14] Neonatal screening for Cystic Fibrosis is, unfortunately, not always the norm. Kharazzi and Kharazzi explain: "Cystic fibrosis (CF) is not always readily diagnosed without newborn screening (NBS). It has few unique features, it is uncommon, and it varies in its presentation. In the United States, half of all persons with CF were diagnosed after 6 months of age. The median delay in diagnosis is well over 1 year in parts of the United States where CF NBS is not used." […] "[S]tudies show that misdiagnosis leads to increased anxiety, guilt and anger, and mistrust of the medical profession." (Kharazzi and Kharazzi 2005: S22)

period of time between parents like Ari's learning that their child will develop HD and his first presentation of clinical symptoms. This time period for CF is much shorter, but if we can show that the anticipation of the child developing CF has a negative effect on family functioning, then we have at least preliminary reason to extrapolate to cases of childhood HD diagnoses and conclude that anticipation of this sort spread over many more years would likewise negatively affect a family like Ari's. Ultimately, what we're trying to determine is how the anticipation of the development of HD might impact the family dynamic and the development of the child.[15] To this end, I will examine the effects of having a child with CF on family functioning. While doing so, I will try to tease out the specific effects of anticipating one's child will develop CF in order to make an educated guess

[15] It is important to note that this import may vary depending on whether or not parents decide to tell their child about her HD diagnosis. It seems that whether a child knows she will develop a degenerative disorder would clearly have an impact on family dynamics. Unfortunately, there is no clear way to distinguish between studies where the pediatric patient herself knew of her CF diagnosis and studies where she did not. I think the most reasonable thing to assume is that parents of CF children will tell their child about her diagnosis in proportion to the sophistication of her capacity for understanding it. So, a very young child might only be told that she must go to the doctor regularly in order to make sure she stays healthy, and an older child would be told that she has a medical issue that needs monitoring in order to manage it, and an adolescent would likely be told exactly what disorder her parents were worried about what to expect in the future. This would be the most reasonable way to release information of this import to a child under such circumstances.

regarding how parents knowing their child will develop a disorder like HD might affect family functioning.

While recent research seems to show that the impact of chronic disease on the family is not as dire as previously supposed, having a child who is diagnosed with CF does seem to have a substantive negative impact on family functioning.[16] McClellan and Cohen concluded in a review of the relevant literature that families of a child with CF score lower than control families in "domains of communication, interpersonal involvement, affect management, behavior control, and role allocation" and that parental stress was significant in CF families.[17] Further, a diagnosis of CF seems to explicitly affect the way that parents treat their children: "Parents of CF children report that they tend to be excessively protective and indulgent with their children, and they acknowledge that such an attitude may be wrong."[18] It's not clear whether in the documented CF cases coddling began before or after the children began to exhibit symptoms, and so it's likewise unclear whether this coddling would extend to children who will eventually develop HD. It's also unclear whether the negative impact of a CF diagnosis on family functioning applies in cases where a child receives a diagnosis but does not yet exhibit any symptoms of the disorder. In order to draw

[16] For evidence that having a child with CF is associated with increased family stress, see e.g., McClellan and Cohen 2007, Spieth et al. 2001, Bouma and Schweitzer 1990, and Holyrod and Guthrie 1986.

[17] McClellan and Cohen, 2007:222.

[18] Perobelli et al 2009: 1932

conclusions about how parents who know their child will develop HD might treat their child differently than they would otherwise and how the family as a whole might be affected by such knowledge in terms of family functioning, we must determine whether anticipation of a child developing CF has a negative impact on family functioning. Given the similarities between CF and HD, if it can be established that family functioning is negatively impacted by a diagnosis of CF before the child begins to exhibit any symptoms, this will be at least *prima facie* evidence that the families of children whose parents know those children will develop HD will be negatively impacted as well.

It's hard to tease apart what portion of familial stress may be due to the *present challenges* of dealing with CF and what is due to the *anticipation* of a child developing CF, but a study by Perobelli et al. concerning uncertain diagnoses of CF may give us a clue as to how to do so. An uncertain diagnosis of CF means that the available testing for CF was inconclusive and so "a diagnosis of CF cannot be made by the current standard diagnostic criteria […] The long-term phenotypical consequences may be highly variable and some of these children might over time develop CF, others could never have any symptoms." (2009:1927) This study supports the claim that having a child with CF is associated with significant parental emotional disturbance, but also shows that *anticipating the possible development of CF* is also associated with significant emotional disturbance on the part of parents:

> [W]hen parents were asked if they believed that their child's health status was causing them any emotional disturbances, there was no differences between parents of CF children and parents of children with an uncertain diagnosis, [AD] and a very significant difference between the latter and parents of control healthy children [HC] ($p = 0.0003$). When parents' answers were processed separately, a significant difference between group AD and HC persisted for mothers ($p = 0.02$), but not for fathers. (Perobelli et al. 2009:1931-1932)

This seems to show that anticipating one's child will develop a degenerative disorder like CF, even when this is uncertain, places an emotional burden on parents. It is reasonable to think that anticipating one's child will develop HD, when this is certain, would impose at least as much if not more stress on the family of the diagnosed child.

The main point of difference between the CF and HD for our purposes is the time of onset. I posit that we can expect anticipating a child will develop any serious degenerative disorder at any point in the future to be a source of familial stress. Actual manifestation of symptoms will likely impose more stress than the simple anticipation that the child will develop this illness, but anticipation will still have a negative impact on the family. One might question here whether the stress I attribute to anticipation in cases of ambiguous diagnosis for CF really just amounts to the stress of having to cart a child to appointments and monitor her health state closely to see if she manifests signs of developing CF. Perhaps the stress in question is wholly or mostly a product of having to *do such things*, rather

than of *anticipation of illness*. To this I can only respond that it seems clear from the standpoint of empathic perspective-taking and common sense that anticipation of one's child developing a degenerative disorder will cause one stress, which will in turn affect one's family in a negative way. In the case of parents of children with an ambiguous diagnosis for CF, the stress reported was likely a combination of both anticipating potential illness and also having to monitor their children and bring them in for checkups, but it stands to reason that the anticipation would play a substantial part in this, especially given that (a) children with an ambiguous diagnosis were not yet manifesting symptoms of CF and (b) even healthy children need preventative care and regular checkups. It's unclear exactly how burdensome monitoring the child was on the parents, but it seems that further data from the Petrobelli study supports the supposition that at least at the time of ambiguous diagnosis and shortly thereafter, parents did not perceive their children to be any sicker than parents of control children did.[19] This seems to be another point in favor of attributing stress to anticipation of future illness, since there didn't seem to be immediate health concerns on these parents' minds.

Another significant difference between CF and HD is that receiving a diagnosis of CF early has substantial medical value, which is why we perform newborn screening in the first place.[20] This medical value justifies subjecting the

[19] In general, parents' anxiety about their child's health is reported as stronger in group CFD than in group AD ($p < 0.05$) and HC ($p < 0.001$), whereas groups AD and HC are not different. (2009:1931).

[20] For evidence that newborn screening for CF has medical value in terms of

family to the stress associated with anticipation, even when the diagnosis is uncertain. For CF there is tangible medical benefit from knowing early, while for HD there is not. If one is going to potentially impose a substantial psychological burden on patients and/or patients' families, there ought to be good reason for doing so. One might say that there are plenty of good reasons to let patients' parents know about an incidental finding of HD before that disorder manifests clinically, in the same way that one might argue that there could be financial and life planning utility to Ari's family if they knew about his diagnosis. However, as I will argue in section 3, concerns about preserving the child's right to an open future to the best of our ability, along with the fundamental uncertainty of whether such financial and life-planning-related benefits would accrue from knowing push us toward discounting such considerations from our moral calculus. If something could be done medically to prevent or ameliorate an illness, then there is tangible utility to the family knowing. In the absence of such benefit, one should not impose psychological distress on one's patient and/or her family. Imposing inevitable stress on the patient's family is thus warranted in the case of CF but unwarranted in the case of HD and similar adult-onset disorders for which there is no cure and for which nothing medical can be done in childhood in order to prevent or ameliorate future effects of the disease.

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generating better clinical outcomes and reducing morbidity of pediatric patients with CF, see e.g., Southern et al. 2009; McKay, Waters, and Gaskin 2005; Sims et al. 2005; Mérelle et al. 2003.

3. Respecting Autonomy

Autonomy is, simply put, the ability to make free and reasoned choices and (at least within certain bounds) to act on those choices. Being able to act autonomously requires that one possess the cognitive capacities required to make rational choices in accordance with one's values and ends. Autonomous action also requires that one possess (at least minimally) consistent and stable values and ends. Access to accurate information and the availability of an appropriate range of choices are also prerequisites for autonomous action and interfering with these can compromise one's ability to act autonomously. So, autonomy is a matter of capacities on the part of the agent coupled with external circumstances conducive – or at the very least not obstructive – to the agent being able to carry out her freely-chosen plans.

We can see how this notion of autonomy may come to bear in medical contexts, giving rise to certain obligations for medical professionals. For example, failure to make a patient aware of all reasonable treatment options for breast cancer would compromise her ability to make an autonomous choice, because the patient would necessarily be acting without relevant information. Perhaps the option offered to the patient is one she would have chosen anyway, but in the absence of other reasonable options (when there are other reasonable options) it cannot be said that the patient acted fully autonomously. This is a case where the patient's autonomy has been diminished by a lack of salient information. Additionally, being unable to understand and rationally choose between available options may compromise a patient's autonomy. If a patient doesn't understand a

consent form laden with complex medical terminology or cannot make a rational choice to due to the influence of drugs or simply not being far-sighted enough to reasonably weigh the costs and benefits of the available options, then that patient will be unable to make an autonomous choice due to being unable to choose rationally. Autonomy can also be diminished in more extreme ways, by closing off choices to an individual through coercion – physical or otherwise. If someone physically incapacitates me, then a substantial set of options have been closed off to me, namely all actions that require ambulation. Likewise, verbal-emotional coercion may compromise one's ability to view choices which ought to be open to one as genuinely viable options. One representative instance of this is when someone in an abusive relationship doesn't consider leaving to be a genuine option due to coercion on the part of the other partner. Being able to act autonomously requires that one can, rationally and free from coercion, choose from an appropriate range of options.

Children are not yet fully autonomous, which is why parents or guardians characteristically act as proxy decision makers in medical contexts. The threshold for being able to give informed consent for medical treatment is typically set at the age of majority. The age of majority thus serves as a heuristic for identifying whether a person is able to act autonomously. This is because the psychological capabilities associated with being able to make reasoned decisions in line with one's own (stable) values and aims are taken to be present by this point in most of the population. It is important to note that, in developing guidelines for disclosure of incidental findings, we're developing guidelines to cover entire populations, so

approximating a level of decision-making competence is the best we can do in terms of developing broad recommendations. It is, of course, possible that in a particular case a patient may be able to act autonomously regarding medical decision-making before the age of majority (I have in mind here an unusually mature older adolescent). When I talk about people being autonomous in this paper, this will serve as shorthand for saying that the individual has the cognitive faculties necessary to give genuinely informed consent to medical interventions.

What exactly does it mean to respect the autonomy of pediatric patients? Should we mete out respect for autonomy on the basis of the progression of a child's ability to make autonomous choices? This is what parents ideally do as their children mature and are given progressively more freedom in their progress to adulthood. And a child's maturing ability to make rational choices on her own behalf should surely be fostered by allowing her to have a say in decisions of great import which involve her. But this notion of autonomy as a developing skill doesn't seem to offer guidance in regard to the question of whether to inform the parents of a child like Ari of incidental findings indicating Huntington's disease. This is because the problem at hand isn't to what extent a child should be included by her parents in cases of medical decision-making, but whether parents should be made privy to this information to begin with. The question is precisely whether to disclose *at all* before the child can consent for herself, and the extent to which the child approaches the ability to give informed consent is largely beside the point. Ari may be able to understand things better at age 10 than he would at age 5, but regardless of his capacity for comprehension, Ari's parents as

proxy consenters would typically be the ones tasked with making the choice of whether or not to have such incidental findings disclosed and then, if they do opt for disclosure, of whether and how to disclose this information to Ari.[21]

What *is* relevant to the release of incidental findings in cases like Ari's is a respect for the child's *future autonomy*. We should, to the best of our ability, avoid compromising the autonomy of the patient as a future adult. By this I mean that we ought not to compromise adults' abilities to make autonomous choices by making certain decisions on their behalf in childhood, in the absence of some compelling and overriding justification. By opting for release of incidental findings which could have been delayed until the child reaches the age of majority with no foreseen adverse physical or psychological consequences, one runs the risk of unjustly compromising the autonomy of the patient as an adult. This is because un-consented-to information may shape the child's life in such a way that her choices are needlessly constrained or she is saddled with choices she ought not to have without choosing to open up those routes herself.

Certain things which happen in childhood clearly have the potential to constrain a child's future autonomy as an adult. By "constraining autonomy" I

[21] If future research reveals that older adolescents are fully capable of providing informed consent in advance of the current age of majority, then the age at which individuals can give informed consent should be lowered, and concerns regarding the legitimacy of proxy consent will apply to a smaller range of children (but still, notably, Ari).

mean limiting the child's future choices, or more precisely, a particular type of the child's future choices. While it could be argued that any choice made on behalf of a child strictly speaking constrains that child's future autonomy, I want to focus on a limited class of constraints on future autonomy. It is not inherently bad to cut off future choices from a child, but it is often bad to make decisions of great import for the life plan and life narrative of the child, when those decisions could have been made by the child herself once she reaches adulthood. This is why things like childhood betrothals and failing to sufficiently educate a child are wrong. They unjustly constrain the choices available to that child when she reaches adulthood: the opportunity to choose a life partner in the former case, and a whole range of employment, educational, and social opportunities in the latter. While it is likely that the children in both such cases would be worse off than they would be otherwise as a result of this constraint on their autonomy, merely being worse off (in terms of well-being) is not the most important aspect of the situation. Rather, it can be wrong to constrain the future autonomy of a child in this manner, even if so constraining would lead that child to have an overall happier or more successful life (as unlikely as this may seem in the aforementioned cases).

It makes good intuitive sense to think that there are life choices of greater and lesser import, some of which may be made in childhood. And, it also makes sense to think that among the most important choices are those which play a central role in the life narrative of a person. Choices that substantially impact the self-conception of the person as well as choices that significantly impact that

person's future are just such choices. And notice that these choices often come together. The choices that substantially shape one's future also often shape one's self, and how one conceives of one's self in terms of identity, character, and values will greatly impact how one chooses. But this is all very abstract. What exactly it means to substantially shape the life of the pediatric patient in terms of life narrative and personal identity and sense of self is in need of explication. To this end, I will consider two illustrative examples of cases where such choices are wrongly made on behalf of children: childhood betrothals, and surgical sex assignment of infants born with ambiguous genitalia.

Let's consider more closely the example of childhood betrothals. I am limiting this example to cases where two families decide that their children of a similar age will be married upon reaching adulthood (or a bit later after finishing school, or whenever). This analysis does not apply to marriages which take place in childhood or betrothals of children to adults, which are both clearly unethical for a whole host of reasons. So, let us consider the more philosophically interesting (and less repugnant) case of children who are betrothed to each other through an agreement between their families. We know that, in general, people who choose their own partners aren't particularly skilled at selecting partners who will last for the ideal lifelong commitment of a marriage.[22] It could very well be the case that childhood betrothals would produce more successful marriages. This might be the case because the partners would likely be compatible for believable

[22] "Current estimates of divorce indicate that about half of first marriages [in the United States] end in divorce" (Copen et al., 2012:1).

(albeit unromantic) reasons. Two families who decide together on a childhood betrothal are likely to be similar in terms of wealth, social status, and perhaps even values. Their offspring will probably have much in common and be able to relate fairly well to each other since they will start off on a basis of familiarity with each other and each other's upbringing and mode of life. Perhaps this is precisely the sort of relationship which would have a high statistical probability of lasting and growing into a deep affection or even love. At the very least, families would likely exert pressure which – while ideally not coercing anyone to stay in an abusive marriage – might incline the spouses to try harder to resolve issues which arise in the course of their marriage than they might otherwise do in the absence of such pressure. Perhaps similarity and familiarity, while not exactly the Platonic ideal of passion, might foster stability and affection better than going off into the world and choosing someone based on a romantic spark or the excitement of new and novel background and features would.

However, even if it were the case that childhood betrothals more successfully predicted future happiness and longevity of marriages than choosing one's own partner as an adult, it still seems like something important would be lost. There is something inherently worthwhile about choosing one's own life partner, even if one does a poor job of it. Choosing a life partner is a decision of immense import for the way one's life will go and having the freedom to self-directedly choose a partner is of the sort of special autonomy-based, life-defining importance which makes being able to make such a decision worthwhile, even if one will be less happy than one might be otherwise. To choose a partner for one's

child when she is very young, even if done from a reasoned belief that doing so will make her happier in the long run, would be to violate the future autonomy of one's child in an unacceptable and unethical manner. The case of childhood betrothals is thus one where autonomy considerations appear generally to trump simple considerations of well-being.

Consider now the more complicated example of surgically assigning an intersex infant (i.e. an infant with ambiguous genitalia) a gender at birth. This example is illuminating regarding the problem we are considering precisely because it is less clear-cut than examples such as childhood betrothals. The question is whether, for intersex children, it is better to choose on their behalf (making the best guess possible) which sex to assign and take surgical means to conform the child's genitalia to this assigned sex or whether it is better to delay this and wait to see which physical and personality traits emerge, ultimately allowing the child to choose a sex once they are able to do so. It is important to be clear that the decision under scrutiny here is not simply one to raise the child either as a boy or a girl. I hold that, for pragmatic purposes, it likely makes the most sense to *socially* assign a gender upon birth.[23] The decision under scrutiny is whether to take cosmetic *surgical* steps to either feminize or masculinize ambiguous genitalia in infants.[24] Such situations may be challenging to resolve in

[23] I here accord with recommendations given by Diamond and Sigmundson, 1997.

[24] In practice, feminization has served as a sort of default for surgeons, since the procedure itself is much simpler than attempting to construct a penis would be. (Beh and Diamond, 2000:3.)

terms of weighing costs and benefits and giving appropriate respect to the patient's autonomy, especially given the serious lack of empirical data available comparing outcomes between intersex children surgically assigned a sex in infancy and those who were not.

Those in favor of surgical sex assignment in infancy typically argue that it would be socially, psychologically, and sexually damaging for an intersex child to go through childhood and adolescence with ambiguous genitalia.[25] They even claim that a child having ambiguous genitalia may interfere with parental bonding and undermine the stability of that child's family relationships.[26] However, I maintain that sex assignment in infancy is a case where, generally, the importance of preserving the child's future autonomy ultimately outweighs such concerns. Surgical sex assignment in infancy and the culture of secrecy built around it seriously impede the child's ability to make a choice of immense import regarding

[25] "Some children with intersexuality require genital surgery for acute medical reasons. In the majority of cases, however, genital surgery has been performed for psychosocial reasons in order to confirm the assigned gender by genital appearance and, thereby, to facilitate gender-appropriate rearing, help develop a gendertypical body image, and avoid social stigma" (Meyer-Bahlburg, 2008: 347).

[26] E.g., "Prolonged periods of nondecision are thought to run the risk of chronically ambiguous or inconsistent sex typing by the family, or of rejection of the child altogether" (Meyer-Bahlburg, 2008: 346). And, "[i]t is generally felt that surgery that is carried out for cosmetic reasons in the first year of life relieves parental distress and improves attachment between the child and the parents. The systematic evidence for this belief is lacking" (Hughes et al., 2006: 557).

a feature that greatly shapes his or her life narrative and identity. Not only is the child surgically assigned a sex that might turn out not to coincide with their self-conception later on, but parents have historically been encouraged to keep the fact that the child was born intersex secret from everyone except the child's medical professionals.[27] This means that the child is growing up without crucial information that may help to open up choices and options regarding their life plan and identity that would not otherwise present themselves. If an individual cannot act on adequate information regarding their biological sex (and medical status more generally) then their autonomy has been seriously compromised. Choosing sex assignment surgery for a child in infancy additionally renders certain choices – such as the choice to live as a man even after undergoing feminization surgery as an infant – much more difficult to make than they would otherwise be. Even if proponents of surgical sex assignment in infancy were correct that having ambiguous genitalia would distress and negatively impact the social and sexual development of intersex children, there would still be value to delaying decisions regarding sex assignment surgery until the child themself is capable of having a substantive say regarding whether they identify as a boy, a girl, or neither.[28] In

[27] See Beh and Diamond, 2000: 50-55.

[28] I agree with Beh and Diamond that "[m]edical uncertainty, the infant's inability to consent to this life-altering treatment and the child's right to an open future suggest that a "moratorium" on infant surgery is the best course when surgery is solely intended to cosmetically change ambiguous genitals" (Beh and Diamond 2000:59).

having a sex surgically assigned at infancy, the child's future autonomy has been significantly and wrongly constrained.

This concern for future autonomy largely coheres with caring about what Joel Feinberg first termed the "right to an open future" for children.[29] While I'd like to avoid being prodigal in the assigning of rights, I do think that the "right to an open future" corresponds to some important moral considerations, considerations which generate substantial duties on the part of parents, medical providers, and even the state. The right to an open future consists of what Feinberg labels "rights-in-trust." Joseph Millum explains what "rights-in-trust" amount to for Feinberg:

> [F]or each autonomy right held by autonomous adults, there exists a corresponding right-in-trust held by children who are not yet autonomous, but are expected to become so. These autonomy rights are, by definition, rights whose exercise depends on the bearer having the capacity for autonomous action, and therefore cannot be exercised by a child. However, they can be violated before the bearer acquires this capacity.[30]

This means that for rights that belong to adults such as a right to privacy and a right to bodily autonomy, there are corresponding rights-in-trust for children. These are rights that do not belong to the child yet (or do not belong to the child in the fully developed forms that belong to adults), but nonetheless may be

[29] See Feinberg, 1980.

[30] Millum, 2014:524.

preemptively violated. There is an obvious question of scope which must be addressed when characterizing such rights. Indeed, one of the main criticisms of the concept of a right to an open future as Feinberg characterizes it is that it is unclear how demanding the standard for respecting a child's right to an open future should be.[31] If there is indeed a right-in-trust which corresponds to *each and every* autonomy right held by an autonomous adult, then it seems that depending on how we conceptualize rights we may end up with a right to an open future that precludes an absurd amount of parenting decisions from being made without thereby violating a right-in-trust. An adult, after all, has the right to make a substantial range of choices for herself, so long as those choices don't violate the rights of others. But, merely by making decisions on behalf of their children in childhood, parents are necessarily closing off certain future decisions from their children, and thus potentially violating rights-in-trust concerning those decisions. However, it would be ridiculous to say that parents are morally required to keep a maximal set of options open to their children in order to avoid violating their children's right to an open future. It is well within the autonomy rights of an adult to decide to make a living as a trapeze artist, but parents aren't morally required to force children to do gymnastics in order to keep this option open for them. Further, parents are not even required to *allow* their children to do gymnastics in order to keep the option of future trapeze artistry open for those children. It seems that for plenty of options an adult may have the right to autonomously select,

[31] See Millum, 2014.

parents have no positive or negative duties to keep those options open for their children.

I propose that we here focus on a limited class of rights-in-trust which concern one's ability to make particular decisions for oneself, perhaps framed in terms of a more general "right to self-direction" under the conceptual umbrella of which are more specific rights and corresponding obligations. For the project at hand I hold we ought to limit the scope of a right to an open future to certain pivotal and self-defining life decisions. The special importance of such life-shaping, identity-establishing choices is – at least in part – what makes childhood betrothals and surgical sex assignment at birth wrong. Choosing one's own life partner is a choice which will have an immense role in shaping one's life. And surgical sex assignment at birth greatly impacts the identity and sense of self of the child. In light of this, to choose a life partner for one's child or opt for surgical sex assignment if one's child is born with ambiguous genitalia would be to violate the child's right to an open future.

However, it seems that there are many life- and identity-shaping choices which would be totally legitimate for parents to make on behalf of their children. Take the example of letting a child know that she stands to receive a sizeable inheritance upon graduating from college. This knowledge would likely have a substantial impact on that child's sense of self and life plan. For instance, if the child is at all prudent, she will incorporate going to college into her life plan. Knowing that she has an inheritance waiting for her will likely influence what she decides to do with her life in terms of a career and life projects. Not having to

worry about financial hardship means that she will not be pushed into any particular career route out of desperation. And, in terms of self-conception, she will be unlikely to identify as proletarian. A child's knowledge that she will receive a sizeable inheritance will shape her life and likely identity in profound ways, but it seems well within the rights of parents to disclose this information to their child. Further, the child would not have been wronged by having a decision to disclose made on her behalf. Bestowing a sizeable inheritance has the effect of opening up a child's future in terms of her range of choices rather than closing it off.[32] This decision does not constrain the child's autonomy in the way that an arranged marriage or surgical sex assignment would. When it comes to making life-shaping decisions on behalf of children, whether those decisions constrain a child's future autonomy matters. Arranged marriages and surgical sex assignment are wrong not only because they make a life- and identity- shaping choice of great

[32] It is worth here asking whether *knowing about* a future inheritance would itself open up a child's range of future choices. After all, plenty of individuals who grow up knowing they have a substantial inheritance to look forward to end up leading lazy and dissolute lives. And, not only that, but these individuals seem to end up leading very similar sorts of lazy and dissolute lives. Perhaps knowing about a sizeable inheritance actually has the effect of closing off one's life in such a way that one is likely to become a feckless social parasite. Despite the seeming ubiquity of this sort of life, I hold that knowledge of a future inheritance would in fact have the effect of opening up one's future, even if some of the options it makes available are not ideal in terms of a standard of objective meaning or goodness in life. Further, it seems that the way in which one was raised would affect one's future plans to a greater extent than the knowledge that one will have the expanded range of options afforded by wealth as an adult would.

import for a child, but also because they have the effect of greatly narrowing the child's range of future life options, and very important options at that.

Rights in trust are thus rights to certain decisions. These decisions are those that will substantially shape the life and identity of the pediatric patient. Who to marry and what gender to live as (if one is born intersex) are examples of such decisions. Choosing whether or not to be made aware of potential incidental findings concerning incurable or highly unlikely to be cured adult-onset disorders is also a decision of this sort. However, it is not enough to simply say that all such choices must be left open to children, or else those children will have been morally wronged. In the same way that sometimes the rights of individuals may be justifiably violated to protect those individuals or others, sometimes the rights in trust of children may be rightly overridden by other moral considerations.

Violating a child's right to an open future ought not be done unless there are weighty countervailing moral considerations, such as serious harm or the imposition of physical, mental, and/or social deficits on the child. Such countervailing moral considerations are even weightier when they concern the sort of thing that would have the effect of closing off the future of the child. Further, certain decisions which would have the effect of opening up a child's future and thus enhance her future autonomy rather than constrain it may be morally permissible to make on behalf of a child, even if to do so might strictly-speaking violate a particular right-in-trust of that child. For example, taking action to prevent one's child from losing a leg would have the effect of opening up the child's future, even though to do so would mean that she will never have the

opportunity to participate in the Paralympics. This might strictly-speaking violate a right-in-trust of the child concerning self-direction, but such a violation would be clearly justified. Appropriate respect for the rights-in-trust of a child does not require that maximal choices be left open, only that particularly important self- and life-narrative-defining choices be left open to the child.

4. Criteria for Delayed Choices

I hold that the choice of whether to have incidental findings of adult-onset, incurable or highly unlikely to be cured, un-mitigatable (at least until clinical presentation) illnesses disclosed to parents of children who undergo WGS falls within the class of choices which ought to be delayed until a child reaches the age of majority and can decide for herself. I propose the following criteria for determining that a choice should be delayed until a child reaches the age of majority and can make the decision herself: (a) the choice concerns something that has the potential to substantially shape the life of the pediatric patient – both in terms of life narrative and personal identity/sense of self; (b) persons could reasonably disagree as to what the correct choice to make would be; (c) it's unclear what the pediatric patient would choose for herself, if she were competent to choose; and (d) the choice can be delayed without medical harm to the patient. These are meant to be jointly sufficient criteria for refraining from releasing incidental findings to parents of pediatric patients, but this is not to deny that there may be situations where withholding incidental findings from parents would be justified on different grounds. With regard to the topic at hand, I hold that (a) the choice of whether to release incidental findings to parents concerns something

that has the potential to substantially shape the life of the pediatric patient – both in terms of life narrative and personal identity/sense of self; (b) persons could reasonably disagree as to what the correct choice to make would be – both concerning release of findings to parents as well as whether to opt for release of findings oneself; (c) in most cases, it will be unclear what the pediatric patient would choose for herself; and (d) the fact that the choice concerns disorders for which nothing can be done medically in childhood or before clinical presentation means that the choice can be delayed without medical harm to the patient.

I have argued that criterion (a) is met and motivated why criterion (a) matters morally as it relates to respect for the patient's future autonomy. As I have argued, releasing such incidental findings to a child's parents has the potential to shape that child's life and future, even if the parents opt to not disclose to the child. But the content of the choice itself is also of great import here. Knowing that one will develop such a disorder has the potential to profoundly shape one's life and identity. Pivotal life choices such as choosing a long-term partner, deciding whether to have children, which career to pursue, what personal projects to pursue, and so on will be shaped by either knowing or not knowing that one will develop such an illness. The child should be able to choose for herself whether she wants to risk a future constrained by such knowledge, or whether she wants to live her life in the absence of such certainty. While it may be difficult to determine exactly how to weigh potential constraints on autonomy imposed by knowing on the one hand and not knowing on the other, the choice of *whether to risk receiving such knowledge in the first place* is one that should be left up to the

child herself. This is reason to take the choice of disclosure out of the hands of parents and preserve it for the pediatric patient once she is able to make it for herself. I will now motivate the importance of criteria (b)-(d) and argue that they, too, are met regarding the choice of whether or not to receive incidental findings of incurable or highly unlikely to be cured adult-onset disorders produced in the course of pediatric WGS.

Criterion (b): *People can reasonably disagree as to what the correct choice to make would be.*

If persons can reasonably disagree as to what the correct choice to make would be, then it follows that, in general, a patient herself could reasonably select either alternative. This fact alone could support either disclosure or nondisclosure. However, if there is room for reasonable disagreement *and* criteria (a), (c), and (d) are also met, then a choice ought to be delayed until the patient herself is competent to choose. I will argue that the choice of whether or not to receive findings of incurable or highly unlikely to be cured adult-onset disorders from pediatric WGS is one where persons could reasonably disagree as to what the right decision to make would be. This is because a preference to either possess or not possess such information is eminently reasonable, and additionally, it is possible to have an extremely strong preference one way or the other and to have even such strong preferences be reasonable. Such preferences will be rooted in what the individual values and subjective features of the individual will greatly shape what the individual would choose. Depending on one's outlook on life and happiness, and the projects and aims one cares about, it is totally coherent to

prefer not to find out about any lethal and incurable or highly unlikely to be cured disorders one will develop or to prefer to know maximal information in order to plan for the future. Depending on the value one places on, e.g., reducing anxiety vs. having control over one's future, holding either preference could make perfect sense. Further, holding either preference *strongly* could make perfect sense. After all, knowledge that one will develop an incurable or highly unlikely to be cured, ultimately fatal, adult-onset disorder has the potential to significantly shape one's life and sense of self. There is no fact of the matter as to whether it is objectively better to go through life blissfully ignorant of disease in one's future or with full knowledge of such disease. What is best will depend upon features of the individual and her subjective values and preferences.

There's no perfect algorithm to use here, so we ought to bear in mind that both giving and withholding incidental findings has the potential to compromise patient autonomy. We should then integrate harms, benefits, and the inherent value of possessing certain choices, such as the choice of one's own life partner, into account when determining whether autonomy has been unjustly constrained. In this way, we can evaluate and choose between two courses of action, both of which have the potential to unjustly constrain a patient's autonomy. Precisely because weighing considerations in this manner is so difficult, reasonable people may disagree regarding whether opting for disclosure and risking the possibility of receiving knowledge of an incurable or highly unlikely to be cured adult-onset disorder is the best choice, or whether it would be best to live one's life in ignorance of any such information. And, because what is right *for the individual*

may vary dramatically from individual to individual, the choice of whether to opt for disclosure ought to be delayed until the individual herself is able to make it. In such a case, there is no right decision to make on the patient's behalf, and any decision made will reflect the proxy consenter's idea of what would be best. There is no decision that is objectively best and a very real possibility that a proxy consenter would be choosing against what the patient herself would view as best, could she rationally understand and weigh her options.

Criterion (c): *It's unclear what the pediatric patient would choose for herself, were she competent to choose.*

While it may be possible for reasonable people to disagree regarding the correct course of action in many cases, it might nonetheless be clear what the patient herself would choose, were she competent (e.g. through advance directives and/or persistent and consistent stated preferences). Criteria (b) and (c) go hand in hand. But, criterion (c) might not apply to some circumstances to which criterion (b) does apply. That is, even if reasonable disagreement is a possibility, it might be the case that the preferences of the patient are clear or easily-inferred. This is less of a possibility in the case of children than it would be in certain cases where, e.g., the elderly need the help of a proxy consenter. In such cases there might be many prior preferences and stated desires to draw upon in making a decision on the patient's behalf, whereas children in general do not have the decision-making and reasoning capacity required in order to form and maintain rational and stable preferences. When an elderly patient is unable to provide informed consent due to dementia, there may be a whole lifetime of stated and persistent preferences to

draw upon in inferring what the patient would want, were she competent to decide on her own behalf. With pediatric patients, there is no such repository of rationally-endorsed and stable stated preferences. The cognitive limitations of children which preclude them from being able to provide informed consent, combined with uncertainty regarding what preferences children would endorse were they competent (or may come to endorse as adults) gives us good reason to hold that – in the absence of countervailing considerations – criterion (b) will be met in the case of choosing whether or not to be made aware of incidental findings of incurable or highly unlikely to be cured, adult-onset disorders from pediatric WGS.

Criterion (d): *The choice can be delayed without medical harm to the patient.*

The choice of whether to be made aware of the sort of incidental findings that this project concerns could be delayed without medical harm coming to the patient. This is because the range of relevant findings is limited to those which concern disorders that are incurable or highly unlikely to be cured and for which nothing medically can be done to ameliorate the effects before the disorder presents clinically. The necessity of criterion (d) being fulfilled has thus been already built into my account.

An Objection: What About Planning for the Future?

The natural objection to press here is this: even if nothing can be done *medically* to prevent, cure, or mitigate a child like Ari's condition by taking action in childhood, surely being made aware of his eventual development of

Huntington's disease could help Ari's parents to plan financially for his medical care and perhaps might help them to organize their life priorities in such a way that Ari has as good of a life as possible, in spite of this life inevitably ending prematurely. And it does seem that one way in which parents' knowledge that their child will develop such a disorder might be beneficial is that it might enable them to better plan financially for their own future and the future of their child. If parents know that their child will develop a degenerative disorder in middle age, this will enable them to plan on better information than they would otherwise, and hopefully to save up for future medical expenses. Additionally, this could have a substantial impact on future planning for events like retirement. If parents know that there is a very real possibility that they will be supporting a child – at least in part – through the course of a fatal degenerative disorder, parents' priorities in old age may look a lot different from those of individuals who do not have such considerations to take into account.

If the child herself is told at some point that she will develop an adult-onset incurable or highly unlikely to be cured degenerative disorder, this information would likely have an impact on her career planning as well as general life structuring. In fact, this information may prove a great boon in terms of adjusting expectations and setting realistic life goals that can be accomplished within a necessarily truncated timeframe. Clearly considerations such as saving for retirement will be less salient to someone who knows that she will likely die before reaching retirement age. And decisions such as whether to have children will likely be informed by such information as well. One might decide against

having children, opt to have children earlier in life than one would otherwise, or simply opt to have fewer children than one would otherwise if one knows that one won't be around for a good portion of their lives and/or to help one's partner raise them. Additionally, for the patient (as we established earlier regarding her parents), financial planning will likely be greatly affected by her possessing such information. Priority-setting in general will be affected as well. There is, after all, not much point in putting off fun things for the future when that future is itself unlikely. So, in light of all this, how can I justify limiting considerations here to those involving *medical* harm and benefit rather than including those considerations which may affect planning of both financial and a more general life-trajectory-related sort? I will here make two arguments to support my position: that the importance of an open future overrides the importance of such planning, and that there is no guarantee that such benefits would accrue from knowing.

The importance of an open future overrides the importance of such planning.

The main tension here is between the utility of planning and the value of an open future for the child. It's already been established that the choice of whether or not to be made aware of incidental findings of incurable or highly unlikely to be cured adult-onset disorders from pediatric WGS is one of those potentially life- and identity- shaping choices which the right to an open future is meant to preserve, in the absence of overriding moral considerations. And, it's been established that persons could reasonably disagree as to whether it would be better to have such information and be able to plan, or not to have such

information and thus avoid having one's life shaped by such information and the anxiety of knowing. So, the question here is, does the utility of planning for Ari's parents (and Ari if they decide to tell him) outweigh Ari's right to an open future? It does not, I contend. Knowing who one's child would marry would surely aid parents in planning for their child's future, financially and otherwise, but the utility of this knowledge does not override the child's right to choose her own partner. In the same way, the importance of the pediatric patient herself being able to decide whether or not to be made aware of any available life- and identity-shaping incidental findings overrides the importance of financial and life-planning on the part of her parents or the patient herself, if the parents opt to disclose this information to her.[33]

There is no guarantee such benefits would accrue from knowing

The contention that the utility of financial and general life-planning should be accounted for when determining whether parents should be given the option to have incidental findings released immediately relies upon a rose-tinted view of how most parents would react to such a situation. This is not to say that there aren't plenty of parents who would responsibly plan for the future of a child who they know will develop Huntington's disease and try their best to ensure that he

[33] One might ask at this point whether Ari's parents might not provide Ari the option of receiving his findings once he reaches age 18. This would, presumably, still leave Ari's future open. However, as I argued in section 2 of this paper, I hold that disclosing to Ari's parents is, in itself, the sort of thing that could potentially compromise Ari's and his family's well-being in a troubling manner.

has the best short life possible. What I mean to say is that there is no saying how all or even most parents would react to knowing that their child's life will necessarily be cut short by a painful degenerative disorder.

It's not outside the realm of possibility that Ari's parents might treat him more coldly than they would otherwise in an effort to not get too attached and insulate themselves emotionally from the pain of loss. Or, perhaps Ari's parents are the cool, calculating types who don't see much point in enrolling Ari in a private kindergarten rather than the abysmal public school in their district. After all, if solid elementary education correlates to later earnings and Ari's life will likely be cut off in his prime working years, what's the point of investing in expensive primary education? Or, perhaps more realistically, his parents will simply not want to deal with thinking about the fact that Ari will die prematurely and would make whatever life and financial decisions they would anyway, with no tangible benefit to Ari of them having this information. Perhaps Ari's parents would live their lives as they would have done anyway, only sadder.

The scenarios outlined above may express an unrealistically pessimistic view of how Ari's parents might respond, but they underscore the point that there is a vast range of possible responses to learning that one's child will develop an incurable adult-onset genetic disorder. Once non-medical circumstances and conjecture regarding priorities which may differ greatly from family to family are brought into a determination of whether certain considerations override a child's right to an open future, determining potential harms and benefits becomes a

deeply uncertain endeavor. Moreover, in light of this uncertainty, we should opt for delayed disclosure.

5. Taking Stock

In light of the importance of respecting the future autonomy of children like Ari, coupled with the unclear and likely negative psychological effects on Ari's family (and families like his) of receiving incidental findings of Huntington's disease (and similar genetic disorders), we have substantial reason to delay release of *at least a certain class of incidental findings* from pediatric WGS until the patient herself reaches the age of majority. While I have gone into greater detail in examining the relevance and importance of respect for autonomy and promotion of well-being, Abdul-Karim and colleagues precede me in pointing to these considerations as favoring a tiered consent process for release of incidental findings from genetic testing on children. However, they only make explicit recommendations for which type of findings from broadly-focused pediatric genetic testing warrant immediate release. This leaves it open for me to build on their account and provide a model for findings which warrant delayed disclosure. Before extending Abdul-Karim et al.'s account, I will briefly outline the model they provide, note what remains to be done, and then outline my unique proposal for return of certain incidental findings which may arise from pediatric WGS.

6. Abdul-Karim et al.'s Proposal Regarding Which Incidental Findings to Release to Parents Upon Discovery

Abdul-Karim and colleagues put forward an account of what sort of incidental findings ought to be disclosed to the parents of pediatric patients immediately upon discovery:[34]

> [I]nvestigators should, at a minimum, disclose incidental findings of genetic variants with known, urgent clinical significance for the children enrolled in the study. We propose the following criteria for evaluating whether a finding has "known, urgent clinical significance": 1. Knowledge of the finding must have a clear and direct benefit that could be lost if the disclosure was postponed until the child reaches the age of majority, such as information that could substantially alter medical decisions in the short term. 2. The potential benefit of knowing the information must clearly outweigh the potential risks of anxiety and other psychosocial harms that could result from this knowledge. 3. Genetic variants related to multifactorial conditions that also have strong environmental components, such as heart disease or diabetes, should be disclosed only if they indicate a substantial increase in risk.[35]

[34] Abdul-Karim et al. also leave it open that a sufficiently cognitively-advanced child like an adolescent might properly receive such findings along with their parents.

[35] Abdul-Karim et al., 2013:567

This is a *minimal* account of disclosure, and it is left open that some findings which do not meet these criteria might be appropriately disclosed immediately. For instance, in some cases where a child may prove to be a carrier for an incurable and impossible to mitigate (through action before clinical presentation) adult-onset condition, future reproductive considerations on the part of the parents may be weighty enough to override reasons for delaying return of such findings until the child reaches the age of majority. By this I mean that if the child will almost certainly develop disease X, and further that any siblings of this child will almost certainly develop disease X as well, we may be justified in immediately disclosing these findings to the child's parents (assuming that the parents are able and might potentially decide to have more children). In a range of cases where there is a real and imminent risk of harm to future children, countervailing reproductive considerations for the family of the child test subject may motivate immediate disclosure. If, for instance, medical providers had reason to believe that Ari's parents were planning on having future children, or even that they *might* have future children, this might warrant immediate disclosure (to his parents) of the incidental finding of HD. Abdul-Karim and colleagues maintain, and I agree, that in certain situations, disclosure may be warranted, even if a finding does not fall within the guidelines laid out by this account. However, this should be left up to the discretion of the relevant healthcare providers.

I will adopt Abdul-Karim and colleagues' criteria for which incidental findings from pediatric WGS warrant immediate disclosure to a child patient's proxy consenter. In what follows, I will extend this account and provide my own set of guidelines for which incidental findings ought to be disclosed to patients upon reaching the age of majority (rather than being disclosed immediately or not warranting disclosure at all). I will also develop guidelines for what procedure ought to be followed in re-contacting patients and potentially disclosing such findings.

7. Specifying Which Findings Ought to Be Subject to Delayed Disclosure and How to Disclose Them

Abdul-Karim and colleagues have laid out a solid account of which incidental findings ought to be disclosed immediately, but it is still an open question whether any remaining findings ought to be disclosed once the patient reaches the age of majority and, if so, how**.** I hold that the information that should be released upon the patient reaching the age of majority is information of clear medical import, but for which nothing could have been done of clinical import before the patient reached the age of majority. Further, the information should concern adult-onset disorders which are incurable or highly unlikely to be cured. However, there may be further considerations (e.g. sufficiently weighty social, reproductive, or financial concerns) which tip the scales in favor of immediate release to proxy consenters. Whether such considerations reach the threshold of significance required to prompt immediate disclosure should be determined by a panel of experts. Further, any findings of unclear clinical import or of marginal

import should not even be considered for release at any point. These are the sort of findings of which the patient need never even be made aware. Receiving findings of unclear or marginal clinical significance would likely be distressing for the patient, as well as clinically useless.

Next, the method for disclosure should be established. There are better and worse ways to go about releasing incidental findings to a patient once she reaches the age of majority. Simply providing a patient with a printout of her genome and wishing her luck would be far from the ideal method of disclosure. The ideal method of disclosure ought to run the least risk of harm to the patient while at the same time leaving open a route to obtaining incidental findings if the patient wishes to do so. Additionally, as always, it is imperative that the patient understand what is going on and the potential consequences of having distressing findings disclosed to her. The patient should be able to provide genuinely informed consent on the basis of the information given.

The ideal model would look something like this: upon reaching the age of majority, everyone who underwent WGS as a child is contacted and told that there may or may not be incidental findings of clinical import related to childhood WGS. The subject is also told that everyone who was tested is being contacted, to prevent her from concluding that something of clinical import must have been found in order for her to be contacted. The person contacting the patient must be wholly agnostic as to the existence of clinically significant findings. At this point, the subject may opt to pursue this further or not. If she opts out, or ignores repeated re-contacting attempts, then the process for disclosure will not even

begin. If she opts in, and there are no clinically significant findings, she'll be told that there are no clinically significant findings, and the process will end. If she opts in, and there are findings of clinical significance, then she will go through the same consent process that she would in the case of any incidental findings for adults.

It is true that contacting each and every former pediatric patient who undergoes WGS will mean contacting a large number of people, but if an efficient, standardized protocol for re-contacting is established it needn't pose too burdensome a pragmatic challenge for clinicians and researchers. After all, e.g., subjects in cohort studies and individuals who donate bone marrow are contacted on an extended basis, and so it seems like there are ample precedents for figuring out a method of patient tracking and re-contacting.

8. A Model for Delayed Disclosure

I will now outline a model for delayed return of a limited class of incidental findings from pediatric WGS. This model will specify the sort of genetic disorders and diseases to which it applies (Conditions on Disease), circumstances necessary in order for the model to hold (Conditions on Circumstances), and what release of incidental findings will look like after the patient reaches the age of majority (Conditions on Release).

i. Conditions on Disease

The sort of diseases and medical conditions to which this model applies are lethal, manifest clinically in adulthood, and are either incurable or have a very

low likelihood of cure.[36] It must also be the case that there is no way to mitigate – in medical terms – the effects of the disease by taking action in childhood, or before the disorder presents clinically. Candidate disorders for currently incurable disorders include Huntington's Disease, Early-onset Alzheimer's, and the dominant variant of Charcot-Marie Tooth Disease. Additionally, since I am primarily concerned with protecting the pediatric patient from being saddled with unasked-for information regarding a truly bleak medical future, disorders with a very low likelihood of cure will also be included in my delayed return model. Genetically-based disorders like Familial Adenomatous Polyposis Coli "(an inherited disorder of the bowel associated with a very high incidence of early onset bowel cancer)"[37] and Early-onset Breast or Ovarian Cancer "(an inherited disposition to develop breast or ovarian cancer in early adulthood)"[38] do not have good prognoses for 5-year survival, even with treatment and would thus fall under the umbrella of disorders included in my delayed return model.[39]

There is a clear concern here as to how we are to determine which disorders to include; arbitrarily choosing a percentage of probability of cure and

[36] It is important to note here that what a 'cure' amounts to may be disease-dependent. Some diseases may be eliminated entirely from the patient's body, while the progression of others may be merely delayed or slowed to a sufficient extent (I have in mind here certain cancers which may be eliminated from the patient's body but are likely to recur at some point within the patient's lifetime.)

[37] Rhodes 2006:210

[38] Ibid.

[39] Ibid.

making that the cutoff for delayed disclosure would be far from ideal. In order to determine whether a particular disorder is unlikely enough to be cured to be included in my delayed disclosure scheme, I propose that we once again rely upon the notion of reasonable disagreement. Specifically, we should take a large representative sample of the population and present them with different candidate disorders and likelihood of successful treatment, and the point at which a plurality of people say that they either would not want to know or would be indifferent to knowing about developing a disorder far in advance of clinical presentation would serve as the threshold for determining whether or not delayed disclosure is a solid option for findings concerning that particular disorder.

This is an admittedly rough sketch of how things would work, but I have in mind a preference elicitation method where people are presented with cases in which the likelihood of successful treatment grows steadily higher. Then, for each case, they would be asked if they would opt for disclosure significantly before the disorder presents clinically. If there are limited enough candidate disorders, all of the disorders under consideration could be included. And, if there are many candidate disorders, then disorders with similarities in terms of symptoms, morbidity, and time of onset could be sorted into bins and individuals being surveyed could be presented with representative disorders from each bin. When it comes to disorders which are currently incurable but for which a treatment is likely to be developed, the preference elicitation method should be adjusted for the possibility of such a treatment being developed, perhaps even building in both the likelihood of a treatment being developed as well as the projected likelihood

of success of that treatment. In this way, which disorders should be included in my delayed disclosure model could be determined in a rigorous and empirically-grounded manner.

ii. Conditions on Circumstances

This account only applies to situations where there is enough stability and bureaucratic competence in institutions that re-contact is likely to be possible. Additionally, there must exist sufficient information security to maintain patient privacy and prevent genetic information from falling into the wrong hands. Maintaining data privacy is a problem that is not unique to this context. Even if incidental findings were to be immediately disclosed to parents, they would presumably be stored in the same electronic format as findings for delayed release. It is sufficient that genetic information is stored at the same level of security as are other medical records.[40] Finally, in order to keep contact information for patients up to date, their parents or guardians should be regularly contacted every couple of years or so to confirm that information is accurate and current.

[40] The only way that security would absolutely not be a concern would be if one either disclosed findings to parents without recording the findings, or didn't disclose findings to parents and also didn't record the findings. In certain exceptional cases, perhaps telling the parents and not recording findings might make sense, but this is not the sort of case with which this model is concerned.

iii. Conditions on Release

I think it best to do a blanket re-contacting of all test subjects, rather than just focusing on those who have incidental findings which may be of clinical import. When the patient reaches the age of majority, she will be re-contacted and informed that she participated in genetic testing, that there may or may not be incidental findings of clinical import for her from that testing and asked whether or not she is interested in being made aware of such findings. The patient should also be told that *everyone* who participate in testing of the same sort is being re-contacted. By which I mean, the patient will be reminded or made aware of the fact that she underwent WGS as a child, and then told that there is a chance of incidental findings of clinical import for all subjects. There may or may not be any incidental findings of clinical import in her particular case, and she may opt in or out of finding this out, once she has an idea of the risks she runs either way. This will avoid the situation where a patient is re-contacted, told that something of import was found, and then asked whether she wants to be made aware of those findings. This is saddling the patient with a choice she may very well prefer to be exempted from, given that a finding of import is probably a finding with bad implications. To call someone up and ask "We found some genetic information we thought you might want to know. Do you want to know?" would essentially amount to presenting her with the information that something is significantly worrisome about her genetic makeup, which could be as troubling and have as much of a negative impact on her life as knowledge of a specific genetic propensity might be.

If the patient simply ignores attempts to re-contact, then her decision to do so should be respected. Re-contact should not be attempted more than 2-3 times, after which failure will be taken to indicate a lack of interest on the patient's part. If the patient opts in to begin the disclosure process she will have to meet with a medical professional who can explain the sort of potential findings which might be revealed and give her an idea of the import of such findings. She will thus have to go through a process to ensure informed consent for the return of any incidental findings from her childhood testing. If the patient provides informed consent, and if there are findings of any substantial significance, she will then meet with a genetic counselor in order to determine both the potential clinical significance of and how to psychologically process the findings. The patient will be free to opt out at any stage in this process.

So, this process for re-contacting has the following steps (1) re-contact all subjects, explain there may or may not be incidental findings from their genome being mapped in childhood and that everyone who underwent pediatric WGS will also be re-contacted. Allow the subject to opt in or out of being made aware of such findings. (2) If the subject opts in, tell her whether or not there are findings of clinical import. (3) If there are findings of clinical import, have the subject meet with a professional to go through a consent process that gives her an idea of what the findings may be and the implications they may have for her life going forward. Given that these findings are limited to a relatively small set of incurable or highly unlikely to be cured adult-onset disorders for which nothing can be done during childhood, the subject should be able to have categories of options

presented to her in terms of general types of conditions of this sort, and these categories will be limited. Hopefully this will make the process minimally burdensome. At this point, the subject may opt in or out of receiving her specific findings. (4) If the subject opts in, a genetic counselor will reveal and run through the findings with her. Special attention should be paid to making the subject be aware of relative risks and how to interpret her findings in terms of risk.

Guidelines for Delayed Disclosure:

Conditions on Disease*:

D1. Adult-Onset. Clinical symptoms would not manifest before patient reaches age of majority.

D2. Incurable or highly unlikely to be cured. Disease in question is currently incurable or highly unlikely to be cured.

D3. Unmitigatable. Nothing medically can be done in childhood to affect the course of the disease.

**That all conditions are met should be verified by a board of medical professionals.*

Conditions on Circumstances:

C1. Significant Likelihood of Re-contact. Re-contacting patients at the age of majority will -- in all likelihood -- be possible.

C2. Sufficient Information Security. Genetic data will be stored at least as securely as other patient information.

C3. Regular Check-In. Check-ins should take place every couple of years in order to make sure that contact information stays current.

Conditions on Release:

R1. Re-contact at Age of Majority. Re-contacting will take place upon the patient reaching the age of majority.

R2. Patient Opts In. Once re-contacted, it will be up to the patient to opt in to release of incidental findings.

R3. Opt In Triggers Secondary Consent Process. The patient will provide informed consent to receive incidental findings.

9. Potential Challenges

9a. Parental Autonomy Concerns

One might object that my proposal restricts parental autonomy in a troubling manner. After all, parents have incredible latitude in how they raise their children and the decisions they may make on their children's behalf. Parents may choose not to vaccinate their children, they may choose to homeschool their children in accordance with strict Biblical principles, to raise their children on a macrobiotic diet or to consult a naturopath for all but the most dire medical conditions. To restrict parents from making such decisions, regardless of whether or not we agree with them, would be to perniciously undermine parental autonomy. And to challenge parental authority by restricting the sort of medical

information they are permitted to acquire on their child's behalf may even go so far as to undermine the family unit, its goals, and collective decisions.[41] In light of this, how can I justify restricting the range of incidental findings available to parents, subverting current parental autonomy in favor of the future autonomy of the child?

Parents rightly have many areas of discretion when it comes to choice-making on behalf of their children, but being a parent doesn't make someone a medical expert or even a particularly good reasoner. Parents aren't allowed to violate seatbelt laws because they think seatbelts harmfully place pressure on the body's essential meridians or completely substitute meditation practice for schooling because they think all knowledge is innate and must be drawn out rather than learned. It's not outrageous to limit the incidental findings made available to parents out of a concern for the well-being of the child, just as it's not outrageous to limit parental authority in any number of other ways.

Parents often have great latitude when it comes to making medical decisions on behalf of their children. This is evident from cases of parents who try – sometimes successfully – to bar their children from receiving conventional medical treatments and talk in the bioethical literature about how taking medical decisions out of the hands of parents threatens 'familial objectives' or fails to give

[41] See Schoeman, 1985; 1983 for a representative position on the importance of and rights that accrue to the family unit.

adequate weight to 'the familial perspective'.[42] The idea behind taking 'familial objectives' or 'the familial perspective' seriously is that the family is a largely autonomous unit that operates on norms that may differ from the norms of the rest of society, or even with basic liberal ideals. The importance of the family as something grounded in intimate relations and shared familial norms and interests is taken to be a weighty moral consideration when family norms and interests come into conflict with competing norms and interests. Ferdinand Schoeman explains:

> The state's relation with the child is formal while the parental relation is intimate, having its own goals and purposes. While the liberal canons insist on the incompetent one's best interest, parents are permitted to compromise the child's interests for ends related to these familial goals and purposes. Parents decisions should be supervened, in general, only if it can be shown that no responsible mode of thinking warrants such treatment of a child.[43]

The state encroaching on the autonomy of the family sphere is generally seen to be bad on this sort of view, although the badness of this may be overridden by other considerations like the welfare of children in extreme cases where "it can be

[42] See chapter 5 of Buchanan and Brock 1989 for further discussion of these notions. The importance of 'the familial perspective' and 'familial objectives' for medical decision making in pediatric cases was originally explicated by Schoeman 1985.

[43] Schoeman, 1985:45

shown that no responsible mode of thinking warrants such treatment of a child" as Schoeman puts it.

I do not agree that restricting the decisions parents are able to make on behalf of their children necessarily denies the importance of the family's perspective and objectives, nor do I entirely understand how appeal to the *familial* perspective amounts to much more than a more palatable presentation of the *parental* perspective. As Allen Buchanan and Dan Brock perceptively point out:

> Given the very great inequality of power between parents and children, reference to the family's interest or "familial objectives" is all too likely to serve as a cover for the parents' interests precisely in those cases in which the latter conflict with those of the child.[44]

The stance I am taking here is controversial, but I hold that there is good reason to restrict parental rights in many cases of medical decision making. This is due, at least in part, to widespread medical illiteracy.[45] It's surely unfair to expect everyone to have the same level of knowledge of medical data and how to interpret it as medical professionals ideally should, but this lack of understanding

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[44] Buchanan and Brock, 1989: 237

[45] For example, the 2003 National Assessment of Adult Literacy report from the U.S. Department of Health and Human Services the found that "[o]nly 12 percent of U.S. adults had proficient health literacy." and "[o]ver a third of U.S. adults—77 million people—would have difficulty with common health tasks, such as following directions on a prescription drug label or adhering to a childhood immunization schedule using a standard chart."

should entail restrictions on the types of decisions that should be made by those with little to no medical expertise on behalf of others for whom they are responsible.

In reality there may be a lot of freedom for parents to make poor medical decisions on behalf of their children, but I contend that there often shouldn't be. While the question at hand is whether my model for delayed disclosure of incidental findings unjustly constrains parental autonomy, I hold that it does not, and that there are likely other cases where similar restrictions on parental decision-making may be desirable. I will here endorse some general rationality and expertise-based criteria for restricting parental autonomy in some cases of medical decision-making. So, a parent may choose a cancer treatment for her child from among the available options presented by a competent physician, but she may not choose to treat her child's cancer solely with sage oil or coffee enemas. In the same way that a parent cannot refuse a life-saving blood transfusion on behalf of her child, a parent cannot make decisions on behalf of her child that are so misguided as to bring about the same outcome, even indirectly.

It is clear that a respect for and prioritization of parental autonomy ought not extend to situations where a parent is inclined to make a decision that will kill her child. But how does justification for such a restriction on parental autonomy relate to whether or not parents ought to have the authority to receive incidental findings of incurable or highly unlikely to be cured adult-onset diseases for their children? While the decision to receive conventional cancer treatment or a life-saving blood transfusion is a matter of life and death, I maintain that a similar

justification for restricting parental autonomy applies to less extreme, but nevertheless quite important, medical decisions. Sometimes adequately respecting a child's future autonomy entails restricting the autonomy of parents. And, in cases where there exists both the potential to seriously constrain the future autonomy of the child coupled with serious concerns regarding the impact of a decision on the child's well-being, parental decision-making authority ought to be constrained if possible in favor of allowing the child to make her own choice once she is competent. Whether to be made aware of incidental findings of incurable or highly unlikely to be cured adult-onset disorders is just such a case. Our status as autonomous agents confers upon us a right to make poor decisions on our own behalf, but not to make poor decisions on behalf of others, in the same way that a right to make our own decisions does not entail that we have a right to make decisions that seriously harm others without some overriding justification for doing so.[46]

[46] A reasonable question to ask here is whether *medical professionals* have a right to make poor decisions on behalf of their pediatric patients. Would medical professionals be less likely to make poor decisions than parents would? I hold that, on the whole, medical professionals would be less likely to make poor decisions on behalf of their patients. This is not to say that there are no deeply incompetent medical professionals, but I do think that the general populace is more likely to be deeply incompetent in regard to medical decision-making than medical professionals would be. If this were not the case, it would be unclear what purpose going to school to obtain specialized medical knowledge would serve.

9b. Potential Restrictions on Testing

This challenge to parental autonomy additionally points to an issue beyond the matter of incidental findings -- that of how to handle the widespread genetic testing of children and neonates which looms on the horizon. My position might theoretically commit me to endorse placing limitations on the sorts of conditions that parents may test their children for in the first place. If it is the case that being saddled with the knowledge of incurable or highly unlikely to be cured adult-onset conditions is damaging to the well-being of children and having such knowledge forced on them does compromise these children's autonomy, then it seems that preventing parents from having targeted testing done on their children for such conditions might be morally justified on the same grounds.

I hold that the way in which we handle incidental findings of the sort with which my model is concerned should also be the way in which we address the prospect of generating non-incidental findings of the same sort. That is, either certain conditions (incurable or highly unlikely to be cured, unmitigable, adult-onset conditions) simply should not be tested for, or, if such testing is conducted, the findings should be withheld until the patient reaches the age of majority and can provide informed consent to either receive or not receive the findings. In general, genetic testing shouldn't be performed in the first place unless it is medically indicated and/or necessary for medical research. In some research contexts, the issue of return of incidental findings is precluded by the design of the project itself. For example, in cases of biobanking, full anonymization may be a prerequisite of donating one's genetic material. In such cases, parents consent

on behalf of their child to this anonymization, which entails that any information gleaned from sequencing the child's genome cannot be returned. In this sort of case, WGS is undergone to benefit medical science rather than the individual whose genome is being sequenced. In the case of non-anonymized medical research, results should simply be disclosed or released to the child upon reaching the age of majority in the manner I have suggested. In cases of clinical testing which may produce incidental findings, my model for delayed disclosure should be followed.

The growing and projected availability of commercial genetic testing complicates this issue as parents may someday be able to test their children to see if they are carriers for diseases like early-onset Alzheimer's or Huntington's disease without having to go through a medical professional. However, this would be wrong of the parent to do. There are many adults with a family history of genetic disorders like Huntington's who opt not to be tested.[47] It seems wrong to deprive the child of being able to make that choice for herself once she's competent to do so. The position put forward in this paper regarding proper limitations on proxy consent for return of certain incidental findings will likewise constrain the reach of proxy consent in cases of non-incidental findings of the same sort.

[47] See, e.g., Abdul-Karim et al. 2012, Marteau and Richards 2000

9c. Re-contacting

The biggest pragmatic challenges to this proposal are the logistical problems of re-contacting and the possibility of failure to re-contact. It may be difficult to keep track of children until they reach the age of majority, especially if they are very young when the testing occurs. This is of particular concern in the United States, where medical records have yet to be standardized across institutions. Unless their parents are very conscientious about informing the hospital or research institution about address and phone number changes, it would be easy for individual patients or subjects to fall through the cracks.

The best way to make sure that contact information remains current would be to check in with the parents regularly (perhaps every couple of years). There is some precedent for this, particularly in cohort studies and other situations where information on particular patient populations is collected over time. However, this leaves a lot of latitude for non-compliance or simple mistakes interfering with one's being able to effectively contact the now-adult subject once she reaches the age of majority. The parents could simply forget to return calls when these periodic check-ins occur, and the research institution might give up after a couple of attempts to establish contact.

Fortunately, there is some hope that patients could soon be tracked from institution to institution, without necessarily needing to re-establish contact to determine if information is current. If a patient moves from one state to another, and their new primary care physician shares electronic records with the old one, and if there were a way to search the entire system for a given patient, then –

assuming the family hasn't gone off the grid entirely and has current contact information listed with some institution in the system – it will be possible to keep track of the patient even over many years. At the moment, we are in the midst of a difficult transition to electronic record-keeping and sharing of medical information.[48] There are still many problems to resolve in order to ensure that hospitals and other medical research institutions can share patient records across different platforms. However, once medical records are shared between all or nearly all hospitals (at least in the United States), it should be much easier to keep track of patients over a prolonged period of time.

10. Conclusion

I have made the case that delaying the disclosure of incidental findings concerning incurable or highly unlikely to be cured adult-onset diseases for which nothing can be done medically in childhood until the pediatric patient reaches the age of majority and can decide for herself whether to receive them is the best way to respect the autonomy and promote the well-being of children like Ari. I have expanded upon Abdul-Karim, Berkman, Wendler, and colleagues' ethical framework and guidelines for when to *immediately disclose* incidental findings produced from pediatric genetic testing and developed and motivated my own account of *which* incidental findings ought to be released to pediatric patients

[48] For a thorough treatment of medicine's difficult transition to digitization, see Wachter, Robert M. *The Digital Doctor: Hope, Hype, and Harm at the Dawn of Medicine's Computer Age.* McGraw-Hill Education: New York. 2015.

once they reach the age of majority, and *how* these findings ought to be released. I maintain that the model I have built here for delayed disclosure of incidental findings is most consistent with respecting the autonomy and promoting the well-being of pediatric patients.

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EARLY PALLIATIVE SEDATION THERAPY AND THE CHALLENGE OF PSYCHOLOGICAL SUFFERING

Introduction

Recently, a physically-healthy 24-year-old Belgian woman named Emily was granted medical assistance to end her life due to unbearable psychological suffering as the result of persistent and severe depression.[49] Many agree that we should provide medical aid in dying (e.g. physician-assisted suicide, active voluntary euthanasia, withdrawal of care necessary to sustain life) to terminally ill patients who find themselves in excruciating and intractable pain.[50] But cases like that of Emily raise some difficult ethical questions. If individuals may opt for physician-assisted dying due to unremitting psychological pain in the absence of terminal illness, this leaves us with some troubling epistemic uncertainty, especially regarding questions of how to determine the severity of the patient's suffering, how to determine the refractoriness of pain, how to decide whether a particular treatment ought to be tried before labeling a patient's suffering refractory, and which psychological factors preclude competent medical decision-making.

In the United States, physician-assisted suicide (PAS) is currently legal in only a handful of states. How exactly PAS is regulated varies from state to state. For instance, according to Oregon's Death With Dignity Act (DWDA), on which

[49] See O'Gara, 2015 and The Economist, 2015.

[50] See, e.g., Dworkin et al., 1997; Brock, 1992; Dworkin 1994.

physician-assisted suicide legislation in other states has been based, in order to receive assisted suicide, a person must have a terminal illness, a prognosis of 6 months or less to live, approval from at least one physician that she is competent to make such a decision, and approval from a psychiatrist or psychologist if her decision-making competence is unclear.[51] This system precludes the sorts of situations which generate the difficult ethical questions that arise from physician-assisted dying to relieve psychological suffering. By limiting the availability of physician-assisted suicide to those who are terminally ill and at the end of their lives, the question of whether to accommodate cases of intractable and severe psychological suffering in the absence of terminal illness by providing medical aid in dying does not even arise.

However, there is an end-of-life (EOL) intervention available throughout the United States which can be employed to relieve unremitting psychological suffering: palliative sedation therapy (PST). And, even though it currently only takes place in EOL contexts, I will argue that endorsing its moral permissibility for use at the end of life rationally entails that we endorse it for use in the absence of terminal illness and when death is far from imminent, if criteria of sufficiently severe refractory suffering and patient consent obtain. This is because if severe suffering, refractoriness, and consent are present in both cases, it is unclear why we ought to permit a patient to opt for PST in the EOL case but not in an earlier case which is identical in morally relevant respects. Thus, a commitment to the

[51] See Oregon Health Authority, 2018.

moral permissibility in EOL cases further rationally commits us to the moral permissibility of early PST.

I will first show how considerations of respect for patient autonomy and well-being (specifically in terms of the patient's dignity) ground the moral permissibility of PST. I will then argue for the rational entailment from EOL PST to early PST and consider the problems of how to determine sufficient severity and genuine refractoriness of psychological pain and how to determine reasonable means to take in establishing refractoriness outside of an EOL context (where this is much less clear than it is in temporally-limited EOL cases). I will further address the question of when psychological illness precludes competent decision-making and whether it might not in some cases be morally right to end the lives of incompetent psychiatric patients who have nonetheless exhibited an established and persistent desire to die, and whose suffering appears genuinely refractory (as Jukka Varelius argues).[52] Finally, I will sketch out a possible way to limit cases of early PST by explicitly endorsing a requirement for severity of pain that is inversely proportional to the amount of time a patient is expected to live.

PART 1: ESTABLISHING A RATIONAL COMMITMENT TO THE MORAL PERMISSIBILITY OF EARLY PST

1a. Defining Palliative Sedation Therapy

Palliative Sedation Therapy (PST), also referred to as "palliative sedation" or "terminal sedation", is an intervention that may be employed by physicians at

[52] See Varelius, 2015.

the end of a patient's life to relieve intractable pain by dropping the patient below the level of consciousness until she eventually passes away. PST is defined by Susan Chater and colleagues as follows:

> [D]eliberately inducing and maintaining deep sleep […] in very specific circumstances. These are: 1) for the relief of one or more intractable symptoms when all other possible interventions have failed and the patient is perceived to be close to death, or 2) for the relief of profound anguish that is not amenable to spiritual, psychological, or other interventions, and the patient is perceived to be close to death.[53]

PST may be used to treat both physical and psychological symptoms. The requirements for employing PST are that the patient be suffering from refractory physical or psychological symptoms, that death be close at hand, and that consent be obtained from the patient herself and/or her surrogate decision-maker. Refractory symptoms are those which cannot be adequately alleviated by any other means (e.g. psychotherapy, drugs, spiritual and/or familial support and so on). Patients are administered sedatives which keep them in a state of unconsciousness and food and fluids are often withheld. PST can last from hours to weeks, but typically a patient will die within the first few days.[54]

Although some object to the use of Palliative Sedation Therapy on the grounds that it is akin to euthanasia, it is widely employed and generally

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[53] Chater et. al, 1998: 257-8.

[54] See Morita, 2004:448; Rousseau, 2000:1065-6

acknowledged to be a beneficial and morally permissible medical treatment.[55] One of the main reasons that PST is legal throughout the United States while active euthanasia remains illegal in all but a handful of states is because PST is prima facie easily differentiated from active euthanasia in terms of the rule of double effect. I will rely here upon Timothy Quill, Rebecca Dresser, and Dan Brock's definition:

> According to the ethical principle known as the "rule of double effect," [when certain conditions are met] effects that would be morally wrong if caused intentionally are permissible if foreseen but unintended. […] Classic formulations of the rule of double effect emphasize four key conditions. [1] The first concerns the nature of the act, which must be good, such as the relief of pain, or at least morally neutral and not in a category that is absolutely prohibited, such as the killing of innocent persons. [2] The second concerns the agent's intention. The "good effect and not the evil effect must be intended." The bad effect, such as respiratory depression after the administration of opioids, may be "foreseen, but not intended." [3] The third condition is the distinction between means and effects. The bad effect, such as death, must not be a means to the good effect, such as the relief of suffering. [4] The fourth condition is the proportionality between the good effect and the bad effect.

[55] See, e.g., ten Have and Welie, 2013; Taylor and McCann, 2005; Chater et al., 1998.

> The good effect must outweigh the bad effect. The bad effect can be permitted only when there is "proportionally grave reason" for it.[56]

It is easy to see how the rule of double effect might justify PST but not active euthanasia.[57] PST satisfies all four of the criteria outlined above by Quill, Dresser, and Brock: (1) In PST cases, the act is good in that it relieves pain. (2) The agent (usually understood to be the relevant physician) intends to relieve the patient's pain through sedation. (2-3) Death in this case is foreseen but not intended, and it is not a means to the end of pain relief, but rather a side-effect of achieving that end. (4) And, in PST cases, the good effect of pain relief is proportional to the "evil" effect of death. This is because the patient is suffering greatly, to such an extent that permanent unconsciousness is desirable. Even if the patient were not sedated, this suffering would in all likelihood continue until the patient passes away from her underlying illness. If being unconscious for the remainder of one's life is preferable to being conscious given the degree of suffering consciousness entails, I think it's reasonable to say that the patient would be better off dead. If the patient's quality of life is so low that she prefers to be rendered unconscious, and if there's no reason to think the patient's suffering will eventually abate (as it

[56] Quill et al., 1997: 1786.

[57] One might make the case that withdrawing a patient's feeding tube while she undergoes PST demonstrates that the physicians intend the death of the patient (see, e.g., Jansen and Sulmasy, 2002). However, it is commonly the case that the withdrawal of food and fluids would have taken place regardless of whether the patient was sedated or not (see, e.g., Morita et al., 2005).

would if the patient were suffering from an illness from which she would be expected to recover, or temporarily sedated in order to let her heal from some sort of trauma) then the bad effect of death would, for that patient, be proportional to the good effect of relieving the patient's suffering. In cases of *active* euthanasia, on the other hand, death is a clear means to the end of relieving suffering. Ending the patient's life is itself the method employed to alleviate suffering rather than an anticipated side-effect of another method of pain relief.

Even if one holds that PST is morally permissible while active euthanasia is not, it is still the case that we can have reason to administer PST to the not-imminently-dying while maintaining accordance with the rule of double effect. This is because the intention to relieve severe and intractable suffering holds regardless of whether the patient is terminally ill or not. I would argue that, if the same degree of intractable suffering is present in a patient who is not imminently dying and one who is, there is no reason to limit the administration of PST to the former. If PST is the only means of relieving such pain, then it seems equally justified in both sorts of cases.

This will be so unless it is the case that death is a greater bad the earlier it occurs such that the bad effect of death will not be proportional to the good effect of suffering relief. The notion that death is worse if it occurs earlier in life, or that death is worst if it occurs during a certain pivotal time period in one's life, is put forward by philosophers such as Jeff McMahan, who argues that, from about age

10 onward, death is worse *for an individual* the more good life years are lost.[58] This is an intuitive notion. After all, a death after one has had the opportunity to live a long and full life seems like a much lesser evil than a death in one's prime, or even before one's life has had a chance to really begin.

However, it also seems to be the case that more suffering is worse than less suffering, and so the total badness of suffering is worse the longer it lasts. If this is true, then the necessity of relieving suffering may be greater if it is anticipated that the patient will suffer greatly for many years. Verhagen and Sauer, for instance, found in a sample of cases of neonatal euthanasia in the Netherlands under the Groningen Protocol, that long life expectancy was a consideration in favor of euthanasia in over 50% of the cases sampled. This was because "[t]he burden of other considerations is greater when the life expectancy is long in a patient who is suffering".[59] Temporal length of suffering seems to be an important consideration in determining whether medical aid in dying is warranted. How are we to weigh the increased badness of death earlier in life against the increased suffering that accrues to a longer timespan?

Many of the general assumptions that undergird accounts of the badness of death do not apply to the sorts of patients who would opt for PST when not-imminently-dying. The tragedy of dying early is the loss of good life years, and so if the life years lost do not meet some minimum threshold of goodness in quality

[58] McMahan, 2002.

[59] Verhagen and Sauer, 2005:960.

of life terms, it seems that death would be less bad for the person than it would be if she had life years of adequate quality to look forward to. If psychological suffering has made it such that a person won't be able to do the things that make human life valuable then, in such a case, an early death might not be any worse for that person than it would be if she were elderly. We might say that death is more tragic *in a sense*, but we wouldn't have real reason to say that death would be worse *for the individual* if she is, e.g., in her early twenties, given sufficient severity and persistence of suffering.

1b. Justification for the Moral Permissibility of PST in the Terminally Ill

I will here explicate two primary moral considerations by appeal to which PST is often justified: respect for patient autonomy and promotion of patient well-being (and specifically dignity related well-being considerations). First, it is important to take the importance of patient autonomy into account. Tom Beauchamp and James Childress provide a canonical notion of autonomy in biomedical ethics, which they summarize as follows:

> Personal autonomy encompasses, at a minimum, self-rule that is free from both controlling interference by others and from certain limitations such as an inadequate understanding that prevents meaningful choice. The autonomous individual acts freely in accordance with a self-chosen plan […] A person of diminished autonomy, by contrast, is in some respect

> controlled by others or incapable of deliberating or acting on the basis of his or her desires and plans.[60]

In order to be an autonomous person, one must at the very least be uncoerced by others and possess the cognitive capacities necessary in order to understand one's choices and make plans. I will add that in order to *act* autonomously, one must have enough relevant information to make a choice (as lacking such information can lead to an "inadequate understanding that prevents meaningful choice") as well as at least a minimal ability to act upon the world.

The notion of autonomy I endorse here is fundamentally individualistic rather than relational as it focuses on the agent herself as a discrete choice-maker rather than building how the agent is socially-situated explicitly into the criteria for autonomous action. However, it should nonetheless be able to withstand some common objections leveled by certain feminist philosophers against individualistic notions of autonomy. The central criticism of individualistic notions of autonomy by proponents of relational theories of autonomy often turns on the truth that sometimes things get in the way of patients being able to freely choose between the choices provided to them. In particular, autonomy may be diminished due to facts about how the individual is socially-situated on a personal and/or institutional scale interfering with an individual being able to know what's actually good for her. Susan Sherwin articulates an objection to individualistic

[60] Beauchamp and Childress, 2009:99

notions of autonomy based on their alleged insensitivity to important features of an agent's broader social context as follows:

> [W]e must pursue a more careful and politically sensitive interpretation of the range of possible restrictions on autonomy than is found in most of the nonfeminist bioethics literature. We need to be able to look at specific decisions as well as the context that influences and sometimes limits such decisions. [T]raditional conceptions [of autonomy] are inadequate to the extent that they make invisible the oppression that structures such decisions. By focusing only on the moment of medical decision making, traditional views fail to examine how specific decisions are embedded within a complex set of relations and policies that constrain (or, ideally, promote) an individual's ability to exercise autonomy with respect to any particular choice.[61]

Sherwin would presumably find the account of autonomy I endorse unsatisfactory. However, we need not think explicitly in terms of structures of oppression in order to "get the right answer" in the sort of cases with which Sherwin is likely concerned.

For example, suppose a heterosexual married couple comes into a plastic surgeon's office for a breast augmentation consultation and the husband is quite overbearing and insistent on his wife's viability as a candidate and the necessity of the surgery, while the wife herself appears squeamish and unhappy. Sherwin

[61] Sherwin, 2012:22.

would say that there is a structure of oppression here that the plastic surgeon ought to be sensitive to and so on. On my notion of autonomy, it may be the case that the wife is being coerced (financially or emotionally) to undergo the surgery, thus compromising her ability to make an autonomous decision. Or, what I think is more likely, sub-coercive pressure by her husband has skewed her conception of what is good for her. I believe that both Sherwin and I would agree that the physician should talk to the wife alone and try to determine whether or not the surgery is actually something she wants. If not, he should refuse to perform the surgery.

The main difference between the proponent of relational autonomy and myself here is that I hold that if the wife convincingly expresses that she is opting for surgery of her own volition and is committed to her choice, the physician should demonstrate respect for her autonomy and consent to perform the surgery. For certain proponents of relational autonomy, this particular choice might be the sort of thing that can never be autonomously chosen in virtue of the relation between its content and pernicious social pressures and norms.[62] That is, on a strongly substantive view of relational autonomy, no woman could ever autonomously consent to (at the very least) purely cosmetic breast augmentation surgery. However, I take this sort of view to be troublingly paternalistic and deeply problematic for reasons I am not able to adequately explore here. Suffice it

[62] See, e.g., Mackenzie & Stoljar, 2000.

to say that I am deeply skeptical that one's capacity to act autonomously is dependent in this way on the content of what one chooses.

Medical aid in dying is often given because the patient has autonomously chosen it or because she has lost her autonomy and it is clear (ideally as stated in an advanced directive) that she would not have wished to persist in her current state. PST is chosen either by the patient herself or her proxy consenters. Informed consent requires that the consenter can autonomously choose between her available options. Loss of autonomy is the top concern patients cite in opting for physician-assisted suicide in Oregon.[63] It is unclear exactly what the patients in Oregon understood autonomy as consisting in; we can probably assume that for most it was not rooted in some sophisticated philosophical understanding of voluntariness in action. However, all that is necessary to reasonably endorse the importance of autonomy in one's own life is an understanding of a commonsense notion of autonomy and the role it plays in persons' lives. And it seems that such a commonsense notion of autonomy would not very difficult to grasp, as it is clearly very important – in the absence of overriding considerations – that a person is able to make choices regarding how her life will go. A life without the capacity for self-direction is a life that is greatly depreciated in value in terms of the individual's happiness. A diminished ability to make choices and act on these choices contributes greatly to a patient's quality of life being severely compromised at the end of her life. PST is justified in part by the value of

[63] See Oregon Health Authority, Public Health Division, 2018:10.

autonomy because it can be autonomously chosen, and it is the sort of choice that can be made with the aim of preventing a loss of autonomy as well. Respecting the autonomous choice of PST by a patient is to respect and affirm an exercise of her autonomous will regarding an immensely important decision in the narrative of her life.

Further moral motivations for PST concern promoting the patient's well-being, specifically as this depends upon her sense of dignity. At the end of life, a patient's sense of her own dignity may be severely compromised by pain and dependence upon others, so it is worth considering dignity specifically in fleshing out well-being related factors which motivate PST. Autonomy and dignity clearly have much to do with the patient's well-being. Someone who is unable to make and pursue self-directed plans will likely have a life of seriously diminished well-being, and someone who takes herself to possess little dignity will likewise have her well-being negatively impacted. Often compromised autonomy and compromised dignity will come together, as a person's sense of her own dignity is often dependent upon what she is able to do for herself rather than depending on others. Here I am limiting well-being to terms having to do with quality of life (QOL) evaluations, where a life of low enough quality can be negative in terms of well-being and, if bad enough, simply not worth living. I take the notion of well-being to be most relevant here to be one that is understood in terms of health states and QOL evaluations / preference elicitations, as they are at least roughly measurable and quantifiable. While QOL understood in terms of health state preferences is not an infallible metric, a very low QOL in these terms is a reliable

indicator that a patient is doing poorly and, at least in EOL contexts, a poor enough quality of life ought to justify taking drastic measures to avoid a miserable death.

Dignity in the empirical sense (i.e. "empirical dignity") is a moral consideration and component of well-being that supports PST at the EOL. A patient's empirical dignity has fundamentally to do with her sense of self and maintaining a level of successful functioning both in terms of physical bodily functioning and in terms of social functioning (the two often go hand-in-hand). We may easily differentiate between the dignified and the undignified in everyday life, but it's difficult to determine what exactly we are identifying when we evaluate an individual as dignified or undignified. Dignity in the empirical sense is a person's view of herself as having a special status, one at least on par with the same sort of status as possessed by others. This sense of dignity is particularly dependent upon a person's conception of herself and her place in the world. Harvey Max Chochinov and colleagues worked to construct an empirical model of dignity in patients at the end of life (aptly titled the "Dignity Model") which I take to be adequate for present purposes. On this model, the following factors– divided into four relevant categories –may diminish a patient's sense of dignity:

> <u>Psychological</u>:
> Depression or anxiety
> Difficulty with acceptance [of own mortality]
> Inability to mentally fight
> Not being able to think clearly

Physical:

Experiencing distressing symptoms

Not being able to carry out usual routines

Not being able to carry out usual roles

Inability to attend to tasks of daily living

Inability to independently attend to bodily functions

Changes in physical appearance

Existential:

Thinking how life might end

Uncertainly about illness

Not having a meaningful spiritual life

Not feeling any longer like who you were

Feeling life has no purpose

Not feeling worthwhile or valued

Not feeling you have made a meaningful contribution

Feeling you do not have control over your life

Social:

Privacy concerns

Not feeling adequately supported

Feeling a sense of burden to others

Not being treated with respect or understanding[64]

It's not the case that we must take each and every one of the above factors into account when determining whether an intervention promotes this sort of dignity in a patient. However, when looked at as a whole, such a list provides us with a

[64] Chochinov et al., 2006:669.

fairly clear conception of what factors may impact an individual's sense of dignity. There will be variation between individuals when it comes to which specific factors contribute to their sense of dignity (e.g. certain factors may vary across cultures) as well as how important their own dignity is to them, but it is reasonable to hold that there are at least some broad unifying factors which most people take to be important to their dignity.

A protracted process of dying often involves having to experience the deterioration of one's mind and body until one is almost entirely dependent upon others for the necessities of daily life. No one wants to lose control of one's body, have one's activities constrained by chronic pain, or rely on others to take care of daily activities such as bathing and eating, to name just a few factors which might compromise one's sense of one's own dignity. When a patient's empirical dignity is compromised by illness this is a bad in itself and can greatly damage that patient's well-being and distress the patient, reducing their quality of life even further. States like being bedridden, having to be helped with basic tasks like using the toilet, bathing oneself, and eating, and a state of extreme dependence generally (and which, in EOL contexts, will often last indefinitely) all seriously compromise a patient's empirical dignity.[65] PST may be opted for out of a desire

[65] There are additional notions of dignity which I will not here consider, in particular a notion of dignity as social currency (i.e. a sort of dignity which essentially depends upon one's place in a society and which can be increased or diminished in proportion to others' opinion of one). This is a type of dignity which is determined from the perspective of one's society and particular individuals in that society, while the empirical notion of dignity I will consider here is

to either alleviate the individual's suffering when she is in that state, or to alleviate her distress at the prospect of falling into that state.

One requirement to receive euthanasia in accordance with the Dutch Euthanasia Act is that "the physician must be convinced that the patient's suffering is unbearable".[66] In section 2a. I will argue that, in general, unbearableness is too high a threshold to set for the severity of suffering that warrants medically-assisted dying. Nonetheless, it is clearly important that patients have, at the very least, a bearable level of well-being. Now, it is true that whether a patient's level of well-being is bearable from her point of view will depend at least partly upon subjective evaluations on the part of the patient. However, I think it is possible to sketch in broad strokes some general features that contribute to making one's life bearable. Being able to pursue activities and projects one enjoys without being racked with pain is important to having a bearable level of well-being. On a more fundamental level, being able to function

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determined from the perspective of the patient herself. Social notions of dignity will be bound up in empirical notions of dignity, and the way in which the individual views herself as possessing dignity will be affected by social factors to a greater or lesser extent (depending on the person and her society). However, I want to focus on empirical dignity from the perspective of the patient rather than from the perspective of society. It is true that myriad social factors may affect a patient's sense of her own dignity, but if those are only important insofar as they affect how the patient sees herself–as I take them to be–then I see no need to consider a separate, socially-oriented notion of dignity here. I will also set aside for the current project a Kantian notion of dignity as inherent worth in persons.

[66] Pasman et al., 2009:1.

at all without being racked with pain seems to be a prerequisite for having a bearable level of well-being. When pain interferes with basic functioning to a great extent, this will seriously compromise an individual's well-being. And, while the point at which life becomes intolerable in the face of that pain may differ from individual to individual, unbearable pain will usually or always be characterized by seriously impairing the individual's ability to function. It is also important that the patient either avoid compromised autonomy and/or empirical dignity if this is what she wants and freely chooses, or, if her autonomy and/or empirical dignity have already been compromised, that she needn't continue to endure living in a state she finds intolerable. PST enables the patient to escape from this intolerable state at the EOL, in cases where such suffering becomes both severe and refractory.

1c. Psychological Suffering

It's not just terminal biophysical illness that can render a patient's life intolerable to her. Psychological suffering can be at least as painful as physical suffering, and so we have good reason to take psychological suffering as seriously as we take physical suffering. In many real-life circumstances it is near-impossible to impossible to tease the two apart. Particularly in end-of-life situations where physical illness is often accompanied by psychological and existential distress, psychological problems like agitation and confusion may be directly caused by or bound up with physical suffering like dyspnea (shortness of breath) or even the disorienting effects of pain medication used to treat the patient's physical suffering.

Pain is complicated and may have vastly different causes and characters. Moreover, the fact that the root cause of psychological suffering may be "all in one's head" in no way renders it less painful or less genuine than something with clear biophysical correlates. This entails that the same obligations which accrue to medical providers in the face of straightforwardly biophysical pain equally apply in cases of psychological suffering. As I'll argue later in this paper, psychological illness poses some unique challenges when it comes to determining refractoriness of pain and decision-making competence on the part of the patient. However, these challenges do not lessen the importance of taking psychological suffering seriously.

1d. A Troubling Entailment

PST involves:

i. the presence of severe and refractory patient suffering (physical or psychological)

ii. chemical sedation to unconsciousness

iii. requested by the patient or proxy consenter

iv. taking place in an end-of-life context (and/or terminal illness)

Now suppose that genuinely refractory symptoms were present earlier in life. Chemical sedation (ii) is used to relieve severe refractory suffering (i) with the consent of the patient or proxy (iii) in order to protect and/or promote patient autonomy, but it is unclear as to why feature (iv) ought to be necessary at all. That

is, in the presence of genuinely refractory suffering, it is unclear what difference the patient being at the end of her life ought to make.

I take it to be the case that an endorsement of PST at the end of life leads us to further endorse PST far earlier in life and in the absence of terminal illness. (I will employ as shorthand "early PST" to refer to cases of PST where the patient in question's death is not imminent and "EOL PST" to refer to cases of PST which take place at the end of a patient's life.) If someone is not terminally ill, but nonetheless is experiencing severe and unremitting refractory suffering that will in all likelihood never be alleviated, the options available to treat that patient are greatly constrained. Here one might ask, what is to be done when a patient whose suffering is otherwise refractory refuses to undergo a possibly effective treatment? A good example of this would be someone who declines to undergo electroconvulsive therapy (ECT) for severe depression. Should the patient be denied medical aid in dying if she refuses to try ECT? Although she may have good reasons for declining – e.g. not wanting to deal with the pain and side-effects like memory loss – I hold that the patient should be denied medical aid in dying in such a case.

While her fear of the pain of treatment and its side-effects are warranted, if the patient is at the point where she's considering ending her life she ought to try all reasonable available treatment options. There is simply too much at stake not to, namely decades of life that could potentially be rendered bearable by ECT. If there is the chance of drastically improving the patient's life rather than ending it, this is something that justifies side-effects that might normally not be warranted.

The potential benefit is proportional to the risk, and while it would be perfectly justified for the patient to decline to undergo that treatment, it would also be perfectly justified to make ECT a requirement for providing medical aid in dying to someone who is, in temporal terms, far from her projected natural death. In the same way that a practitioner is not obligated to prescribe opiates to relieve obesity-related pain in a patient who is unwilling to undergo bariatric surgery out of reasonable concerns about long-term side-effects and the danger of the surgery itself, so a practitioner is not obligated to provide medical aid in dying to a patient who is unwilling to undergo ECT out of reasonable concerns regarding side-effects. This analogy might not be perfect, but the point is that one ought not to go straight to a drastic treatment for symptoms when there is an available – albeit risky – treatment with a reasonable likelihood of success to address the underlying cause. In both cases, the patient could potentially have years of low-quality life drastically improved, and in both cases the patient could still receive her preferred method of treatment if the recommended treatment fails.

In order to support the rational entailment from EOL PST to early PST, let us now consider the following hypothetical example of "Alice":

> Alice is a 35-year-old occupant of the United States who suffers from severe clinical depression. She is not terminally ill. Her depression started in puberty and has lasted for over twenty years. Alice cannot work or maintain friendships due to her total lack of motivation and is in a constant state of affective misery. Her family is supportive and loves her, but she cannot bring herself to feel happy about this, even though she appreciates

> their help and knows intellectually that she cares a great deal for them. Alice has tried every possible cocktail of psychiatric medication to treat her condition, but to no avail. Occasionally, a new medication will lead to a few days of lessened dysphoria, but these always give way to the same state of deep unhappiness as before. Alice has further tried talk therapy with many different professionals, but to no avail. She has tried to make lifestyle changes regarding diet and exercise, and even sampled different religious practices in an attempt to find something to make her feel better. None of these interventions worked either, and merely felt like going through the motions with little to no payoff. Finally, Alice tried electroconvulsive therapy (ECT) as a last-ditch effort to alleviate her depression. This also failed. Alice has considered suicide many times but could never find the motivation to go through with it, and further didn't want to cause her family any trouble or distress upon discovering her body. She has resigned herself to the fact that she will never be happy and over the years has persistently and consistently expressed a desire for her life to end. The psychiatric consensus is that Alice's depression is severe enough to seriously compromise her quality of life, as well as being genuinely refractory.

What should be done to help Alice? If her pain truly is refractory and she has exhausted all her treatment options, then it seems that medical professionals in the United States are left with the choice of either allowing her suffering to continue or sedating her to the point where Alice is no longer aware of her suffering. If

Alice is sedated and continues to receive food and fluids, then she will likely live for quite a long time, ultimately dying of some complication related to being hospitalized, which will likely take the form of an infection. If food and fluids are withheld, Alice will pass away sooner.

The case of Alice illustrates how all the central moral considerations behind EOL PST can apply far in advance of a person's projected natural death. Alice has expressed a persistent wish to die. If given this choice, she would autonomously consent to have her life ended with medical aid. The reason she wishes to die is that her subjective quality of life is so low as to be worse, on her own evaluation, than death. Her unremitting and severe psychological suffering is such that Alice cannot derive any joy or hope from life, and since it has been judged refractory, there's no prospect for that pain to be alleviated, apart from a completely unexpected breakthrough in treatment or change in the way her brain is 'wired'. In light of this, it seems that it would be justified to use PST to end Alice's suffering and the suffering of patients like her. This further shows that our commitment to the moral permissibility of PST at the end of life rationally commits us to endorsing the moral permissibility of PST earlier in life, when the conditions of very intense and unremitting refractory suffering and informed consent obtain.

While it may seem a bit far-fetched to imagine non-terminally-ill patients who are tired of life coming to the hospital in droves and requesting to be sedated below the point of consciousness, Julian Savulescu has posited that something

very similar could at least be possible.[67] Voluntarily stopping eating and drinking (VSED) can take place at any point of life by individuals who have decided it would be better to die for whatever (competent) reason. Savulescu argues that, since it would be immoral to compel a competent individual who has made the reasoned decision to refrain from eating to eat, and doctors have the obligation to care for patients regardless of whether harm is self-inflicted or not, a patient who voluntarily chooses to starve herself in order to die has the right to receive palliative care:

> [A]ny competent person has the right to refuse to eat and drink, leading to their death. And given that they will certainly die if they do not eat and drink, they are entitled to relief of their suffering as a part of medical treatment as they die. This can be achieved through palliative care involving sedation and analgesia, perhaps even so-called 'terminal sedation.'[68]

In such cases, PST (terminal sedation) would presumably take place when suffering from starvation becomes refractory. A patient who voluntarily stops eating and drinking and who receives such palliative care will thus be in the process of what Savulescu terms "Voluntary Palliated Starvation" (VPS).

[67] See Savulescu, 2014; 2015.

[68] Savulescu, 2014:111.

Now, while I think this is a novel and potentially useful solution to certain end-of-life problems in places where voluntary euthanasia is not yet legal, Savulescu and I disagree on a few grounds. Savulescu bases his claim that a competent person ought not be compelled to eat on a 'moral principle of inviolability of the person': "It is impermissible for one person, A, or several people B-D, to insert any part of their body, object or substance into the body of another competent person, X, without X's valid consent".[69] I am skeptical that such a principle applies to force-feeding under the wide range of circumstances which Savulescu takes it to, especially when it comes to his claim that "[s]ome anorexics, perhaps many, are competent" when it comes to a reasoned desire to starve.[70] There is a difference between using voluntary starvation as a means to die when life has become unbearable and assigning *not eating itself* such a high priority in and of itself that resulting death does not matter to one as much as the importance of refraining from eating. Savulescu wrongly conflates the two situations. As the Mayo Clinic characterizes it, anorexia involves "an abnormally low body weight, intense fear of gaining weight and a distorted perception of body weight."[71] If an anorexic person prioritizes not eating over living on the basis of her fear of gaining weight or inaccurate conception of her body as being too heavy, then she is making her decisions on the basis of a skewed perception of the world that is rooted in psychological illness, which would preclude her from

[69] Savulescu, 2014:111.

[70] Ibid.

[71] Mayo Clinic Foundation for Medical Education and Research, 2018.

being able to make a reasoned and competent decision. There is evidence that decision-making in general is impaired in people with anorexia nervosa, manifesting specifically in "a preference for immediate reward despite the long-term adverse consequences".[72] Additionally, "there is a broad consensus that involuntary treatment of patients with eating disorders is ethically and legally justifiable when the patient is at acute risk of death from the medical complications of his or her disorder".[73] So there is good reason to think that an anorexic patient cannot competently opt for VPS as a means to persist in a state of starving herself.

However, if an anorexic chooses to die rather than eat on the basis of the suffering generated by her psychological illness being unbearable to her, this might be a situation where she could make a competent decision to opt for VPS. After all, anorexia is a painful illness to suffer from, especially when one is in the latter stages of starvation. So, it might be the case that an anorexic could competently choose to end her life rather than eat because her illness itself has rendered life intolerable to her. And, if her suffering could be proven severe and refractory, then she would possibly be a candidate for early PST as well under the entailment I've built in this paper.

[72] Adoue et al. 2014:121

[73] Bryden et al. 2010:139

I take the motivation of anorexics who would prioritize not eating over remaining alive to show that the intention behind VPS matters when it comes to determining whether or not (a) the person making the decision is in fact competent to make that decision, and (b) whether a case of VPS is chosen with the aim of permanently relieving suffering (physical, psychological, or existential). It's not out of the question that an anorexic might judge that a world in which she must eat, given the pain of her psychological illness and little to no prospect of a cure in her particular case, would be worse than death, and rationally choose VPS as a means to death. However, this is importantly different from using VPS as a means to persist in a state of starving oneself with death as a mere side-effect (there is here a novel reversal of the sort of argument which applies the rule of double effect to distinguish between PST and euthanasia).

There would, of course, be a significant chance of error in determining the intentions of any particular anorexic. And this uncertainty alone might provide reason to judge anorexics incompetent in general to make such a choice. However, it might also be the case that, if the anorexic were suffering greatly and likely to die soon of her illness regardless of whatever intervention might be taken, granting her request might make the most sense morally, all things considered. However, at this point we come up against the problem – which will be discussed in section 2d – of when psychological illness precludes competent medical decision making and whether, in some cases, the presence of extreme suffering should override a requirement of competence in order to request aid in dying.

PART 2: CHALLENGES TO EARLY PST

Clearly, a lot rides on the severity and refractoriness of a patient's pain when determining whether PST is morally permissible, but how are we to determine genuine refractoriness of psychological pain? Whether or not pain is refractory will depend upon whether it is resistant to the available treatments, but it is not the case that literally all treatment options need be considered. Some treatments may be unproven and experimental, and some may constitute *heroic measures* – i.e. measures that are so burdensome or so risky as to fall outside the category of treatments that medical professionals are obligated to provide to their patient (when it is within their power to do so). How we answer the question of how to determine the genuine refractoriness of psychological pain in candidates for early PST will thus depend in part upon the answer to the following question: how are we to define heroic measures when it comes to treating psychological pain in candidates for early PST? Both questions grow more difficult to answer the further away the patient under consideration is from the end of life. I will consider these questions in sections 2a – 2c.

There are further questions regarding the potential effect of psychological illness on decision-making competence to consider here. As I will argue, decision-making competence is rooted in a person's ability to make a reasoned choice based on an at least minimally-consistent set of values or conception of the good and an accurate understanding of the facts of the situation at hand. But it seems that psychological suffering is the sort of thing that, at least under certain circumstances, could seriously undermine an individual's ability to choose

rationally and consistently. And, further, even if there is some straightforward way to make competence determinations in patients suffering from psychological illness, we have a related question of whether some cases where patients clearly cannot give consent but whose suffering is severe and who clearly desire to die warrant ending those patients' lives through medical means. I will consider these questions in sections 2d and 2e.

2a. Determining Severity

Before addressing the question of how to determine refractoriness of pain, it is worth saying something about the severity of pain that warrants PST. In the Netherlands, in order to obtain medical aid in dying, physicians must "be satisfied that the patient's suffering is unbearable".[74] However, opinions on what exactly constitutes unbearable suffering may differ from physician to physician and have been shown to differ significantly in situations of psychological rather than purely physical suffering.[75] It is difficult to know how exactly to understand what it means for a patient's suffering to be unbearable. Should a patient's suffering being unbearable mean something akin to its being intolerable to her, or should unbearableness be understood in a stronger sense, as something that would eventually drive the patient to madness or literally render her unable to function? One would rely largely on subjective QOL evaluations on the part of the patient in order to determine the former. If the latter sense were understood strictly enough,

[74] See van Tol et al., 2010.

[75] See Ibid; Rietjens et al., 2009.

it would mean that the patient would – due to physical and/or psychological suffering – be rendered either unremittingly hysterical, catatonic, or in a state resembling paralysis as a result of her suffering. Even understood more loosely, unbearableness in anything approaching a literal sense would set a very high bar for suffering. In practice, unbearableness seems to be understood more closely to the former more subjectively patient-dependent sense, although there is no concrete universally accepted definition of unbearable suffering.[76] There is significant variation in how unbearableness is interpreted by different medical providers. In one survey, for example, it was found that a patient's being able to read despite her pain was taken as evidence by her physician that this pain was not, in fact, unbearable to the extent that warranted medical aid in dying.[77] If there were interpretive agreement on what constituted unbearable suffering that accorded with this particular understanding, that would be very high bar to set for a patient's suffering to be unbearable.

I'm not convinced that an unbearable degree of suffering – especially if this is understood in anything resembling the strong sense sketched above – is required in order for one to properly be a candidate for medical aid in dying, regardless of one's age and proximity to death. I suspect the unbearableness requirement is present in the Netherlands' legislation because they do not have a requirement that the patient be terminally ill or at the end of her life, and so the degree of pain which may warrant ending a life ought to be of an extreme sort in

[76] See Dees et al., 2010.

[77] See Pasman et al., 2009.

order to be taken to warrant ending an otherwise sustainable quality of life (I will say more about this later).

In cases of PST, I hold that the pain necessary to warrant sedation needn't be strictly-speaking unbearable. But suffering does need to be sufficiently severe that the patient would want drastic measures to be taken in order to alleviate that pain. That is, suffering should generally be understood in terms of intolerability *to the patient*. If the patient is competent to opt for PST herself, then determinations of severity of suffering will depend upon her own stated evaluation of her suffering as intolerable to her. If the patient is not competent to consent to PST, then behavioral indicators of pain along with stated intolerableness or similar statements on the part of the patient (when possible) will be taken as evidence for the presence of suffering which either is or would be intolerable to the patient (if she were aware enough to process and/or articulate this). In practice, the determination of whether the patient's pain is severe enough to warrant PST is left up to the medical professionals involved. And, so, there may be much variation in practice in the sort of suffering that warrants PST. However, I think that understanding sufficient severity in terms of the sort of suffering which would bring about a state that the patient would find intolerable is the best way to go in moral terms. This leaves room for variation between patients in the sorts of states which might warrant PST, variation which would presumably align with the patient's values, preferences, and conceptions of the good.

However, such a patient-centered determination of sufficient severity should be tempered by professional and commonsense judgment. For example, a patient who is unusual to the point of not taking any degree of pain to be tolerable should probably not be able to opt for PST due to finding intolerable (or claiming to find intolerable) the minor stiffness that accompanies her daily walks around the hospital ward. In this case the absence of any objective level of pain that could reasonably warrant PST would weigh against the patient's judgment that the pain she does experience is intolerable. In the same way, a patient who is otherwise mostly content but who undergoes intermittent episodes of despair ought not to be able to opt for PST in the midst of one of these episodes. The fact that she is prone to such episodes might ultimately provide good reason to opt for PST, but the persistence and consistence of her preferences must be established before such a request can be carried out.[78] However, if the patient falls into an episode of despair and is likely to die before cycling out of it, PST would be warranted.

[78] One might here ask what should be done if the patient persistently and consistently requests PST while in the midst of such intermittent episodes. Perhaps every time the patient is in an episode of despair, she persistently and consistently requests PST, even saying something like "I don't care what I want or say when I *don't* feel like this; right now and every time I *do* feel like this, I would do anything to make it stop." Why should we give priority to the decisions she makes when she is not experiencing such suffering? One might say that in the midst of an episode she is not thinking as clear-headedly as she would were she not in pain, but perhaps she will have forgotten the severity of the pain when she is not actively suffering. So, it might be perfectly reasonable to want to die in order to avoid an episode the true severity of which is forgotten as soon as she

In determining that suffering sufficiently severe to warrant PST is present, expressed and/or apparent intolerableness of pain must be considered along with whether there is good reason to think that the patient's pain – both in terms of degree and persistence – is at all proportionate to this sort of appearance and/or expression of intolerableness. Here would ideally be where professional and commonsense judgment come into play. Relevant factors to account for in determining whether the stated intolerability of a patient's pain corresponds to reality should include the patient's level of functioning regarding basic physical and cognitive tasks, behavioral indicators of suffering, and the patient's general disposition and mental/emotional state. Of course, a default attitude of suspicion towards patients' expressions of intolerability doesn't seem as if it would be the best sort of orientation to take towards those to whom one has a professional duty of beneficence. And it is also clear that the attitudes of medical providers regarding medical aid in dying and the circumstances under which it is warranted may lead differing standards of proof regarding intolerability, in the same way that the standard of unbearableness that warrants medical aid in dying has been interpreted and applied quite differently across individual medical providers in the

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cycles out of it. It is difficult to say what to do under such circumstances, but I think it might be best to take into account both how much longer the patient has left to live and how much of that remaining lifespan is likely to be spent in the midst of an episode. If she will spend the majority of that time in the midst of an episode, then granting a request for PST ought to be considered. If episodes are likely to be infrequent, then the desire to stay alive of her non-suffering self ought to be prioritized.

Netherlands.[79] Some basic parameters should be put in place to ground a reasonable standard of intolerability. Defining such parameters will be a challenge, as they must be flexible enough to accommodate variation across patients while at the same time ruling out statements of intolerability that seem to accord not at all with reality. However, it is important to note that the timeframe in which PST typically takes place is quite short. The patient is usually imminently dying, and the fact that the patient is suffering greatly will usually be apparent. In such situations, in which the patient is already dying and clearly suffering, erroring on the side of pain relief makes sense, even when persistent preferences cannot be established, or the patient is not lucid enough to express whether or not she finds her pain intolerable. While determinations of severity may be complicated in practice, I maintain that the severity of pain which warrants PST is that which could reasonably (both in professional and commonsense judgment) be found intolerable by the patient.

2b. Determining Refractoriness

PST ought only to be employed in circumstances where the patient's pain is genuinely refractory. This is something that may be determined relatively easily at the end of a patient's life. The temporal limitations that imminent death places on the treatments available to medical professionals to relieve their patients' pain make it so that there are only a few options. This is why, for example, the

[79] See Rietjens et al., 2009; Pasman et al., 2009; Dees et al., 2010; van Tol et al. 2010

addictiveness of opioid pain relievers does not factor into the decision of whether to prescribe them to patients who will be dead within days to weeks. Of necessity, at the end of life, the emphasis is on relieving pain and facilitating a comfortable death rather than considering future consequences of treatment. This is so for psychological pain as well as physical pain.

As a rule, the further one is from death, the more difficult it becomes to determine whether one's pain is genuinely refractory. After all, sometimes time itself does heal certain wounds. Many biophysical ailments and disabilities may be adapted to psychologically.[80] However, when the illness itself is psychological, it makes at least intuitive sense to think that adaptation would be less likely. In fact, rather than adaptation, it seems that what would be required to alleviate pain from a psychological disorder would be a lessening in severity of the disorder itself. But major mental illness is notoriously treatment-resistant.[81] And, despite the seemingly intractable nature of many mental disorders, determining the refractoriness of suffering for psychological problems can be immensely challenging. This is because psychological suffering is often a multifactorial problem with unclear causes and even less clear solutions. It is a challenge to

[80] See, e.g., Menzel et al., 2002; Bagenstos 2007.

[81] Major depression is one representative mental illness that may cause refractory pain in those who suffer from it: "There are many individuals (up to 15% of patients [treated for major depression]) for whom multiple interventions will be unhelpful and who will have significant depressions despite aggressive pharmacologic and psychotherapeutic approaches" (Berlim and Turecki, 2007:47).

determine, for any particular patient with a major mental illness, what combination – if any – of medication, talk therapy, lifestyle, environmental, or social changes, and/or more extreme measures such as ECT will prove effective. It is thus difficult to successfully treat mental illness, and it is also difficult to determine when all reasonable treatment options have been exhausted in any particular case. This complexity leaves us with substantial uncertainty in making determinations of refractoriness in cases of mental illness, an uncertainty that compounds the further a patient is from her projected natural death.

2c. Defining Heroic Measures

Whether a patient's suffering is genuinely refractory can be reasonably thought to depend upon what treatments are available and whether all those treatments have been tried. However, it may be the case that some treatments fall under the heading of "heroic measures" which would be too uncertain and/or too burdensome to reasonably justify subjecting a patient to. For instance, it is clear that, although the River Ganges is taken by many to have extraordinary healing properties, it would be totally unreasonable to fly every patient for whom all other treatment has failed over to India to give bathing in the river a try. This would constitute an enormous investment of resources in the hope of bringing about a totally unproven and unlikely cure. However, it is difficult to know where to draw the line regarding what sorts of treatments would count as "heroic" when the suffering patient potentially has a substantial portion of her life left.

It is accepted that heroic measures are unnecessary (and often undesirable in terms of patient quality of life) in care for patients at the end of life. This is largely because experimenting to find the absolutely optimal treatment for a pain condition at the end of life will expose the patient to unnecessary stress and pain. However, what constitutes a heroic measure changes depending on how much time one has to work with when treating a condition. This is why long-shot surgeries ought not to be performed at the end of life; they are unlikely to help the patient and very likely to make the patient's last days or hours more uncomfortable than they would be otherwise. Not to mention, such surgeries are a substantial waste of resources.

It is difficult to determine which interventions ought to be characterized as "heroic" the further one is from the end of life. Recall the example of Alice, the 35-year-old woman who suffers from severe and unremitting major depression. She has struggled with this for about 20 years; it began roughly when she hit puberty. Alice has tried every conventional treatment for her condition: a wide range of medications and combinations of medications, lifestyle changes, talk therapy, cognitive behavioral therapy, even hypnosis and ECT. She has been conscientious about taking medications as prescribed and waiting an appropriate amount of time to see if an intervention worked. Nothing has worked to alleviate her depression. Considering this, she is tired of life and wishes for it to end. However, she can't work up the motivation to kill herself and doesn't want to impose the burden and legal risk of assisting in her death on any of her loved ones, so medical aid in dying is her only viable option to relieve her suffering.

In order to clarify the question of what constitutes a heroic measure in Alice's case, let's focus on one example of a candidate heroic measure: psychosurgery, specifically in the form of lobotomy. Were Alice hospitalized at the end of her life due to some biophysical comorbidity, she would likely be sedated, or at least persistently drugged to the point that her sense of self disappeared enough to make her psychological problems disappear along with it. Psychosurgery would be out of the question because the potential benefits wouldn't outweigh the risks, given the limited timeframe. But Alice is in perfect biophysical health. The conventional treatment options have been exhausted. Despite being perfectly healthy, she nonetheless wishes to die. Her depression and anxiety have proven genuinely refractory in conventional terms. The question here is whether, with at least 30 years more of expected life in good health, this patient would need to try psychosurgery in order to determine that her condition is absolutely refractory. Put another way, do 30 future years of life render otherwise heroic measures reasonable?

In general, I think they do, but much will depend upon the individual patient's particular circumstances. Given the current medical landscape, I would suggest as a rule of thumb that psychosurgery like lobotomy should only be tried if the patient expressly and persistently requests it and, even then, this request should be heavily scrutinized and might reasonably be denied. This is because, even after decades of research and practice, lobotomy remains a procedure that poses immense risk to the patient in terms of potentially altering her personality

and/or further decreasing her quality of life.[82] However, experimental treatments that would ordinarily be considered heroic and would pose less of a risk of further compromising the patient's well-being ought to at least be considered for patients like Alice.

2d. When Does Psychological Illness Preclude Competent Decision Making?

Another challenge regarding early PST for patients with psychological suffering is determining whether the patient is competent to rationally opt for PST. Cases of extreme psychological suffering pose a unique challenge to obtaining informed consent. Psychological illness doesn't necessarily preclude a patient from possessing the capacity necessary to give genuine informed consent, but it often may do so. Doernberg et al. explain that "although psychiatric diagnoses should not be equated with incapacity, some neuropsychiatric conditions are known to increase its risk. These include psychotic illnesses, neurocognitive disorders, some forms of depression, anorexia nervosa, and mental retardation".[83]

Decision-making competence in medical settings requires certain capacities. I will here adopt Buchanan and Brock's account of decision-making competence in medical setting wherein such competence requires "the capacity

[82] "Nowadays, lesions [caused by treatments such as lobotomy] should not be considered anymore except if no other alternative is available due to […] complications, low rates of success, and irreversibility." (Andrade et al., 2010: 573)

[83] Doernberg et al. 2016:557

for understanding and communication and the capacity for reasoning and deliberation".[84] In addition to these capacities, decision-making competence requires that the patient "have a set of values or conception of the good".[85] What the capacity for understanding and communication amounts to is prima facie straightforward (at least in theoretical terms; in clinical reality, as with most things, matters may be less clear cut); the patient must be able to understand the content being relayed to her and be able to express her preferences and decisions and ask questions when necessary. Understanding requires that the patient have the cognitive abilities required to take in and process information, along with the ability to do at least some basic perspective-taking and mental time travel when it comes to envisioning what the future might be like in the face of different treatment alternatives. The capacity for reasoning and deliberation again requires certain cognitive abilities. The patient must have at least some ability to draw inferences and reason probabilistically and must be able to retain information long enough to run through a process of deliberation. Finally, the patient's possession of some (at least minimally stable and consistent) set of values or conception of the good is required for decision-making competence because without these, the patient would be unable to reasonably and consistently assign weights to different considerations and options in terms of goodness or desirability. For example, the relative weights assigned to four additional weeks of life in great pain would be

[84] Buchanan and Brock, 1995:23

[85] Ibid.

radically different for a patient with a strong Protestant work ethic as opposed to one who greatly valued hedonic goods.

In order to rationally choose between options, a patient must first be able to evaluate those options in terms of her own values and the preferences which arise from those values. However, merely possessing a minimally consistent set of values and conception of the good seems not to be enough to ground a satisfying conception of decision-making competence. If one's conception of the good is warped by mental illness, for example, this may preclude one from being able to choose competently. Figuring out how to determine whether all of the criteria for decision-making competence are met by particular patients in a rigorous and standardized way is a daunting challenge (and one which physicians in the Netherlands seem largely to have failed at) but it is imperative we get it right, given the immense and irrevocable import of a decision to seek medical aid in dying. [86]

2e. Should Decision Making Competence Always Matter?

However, in requiring individual competence for medical decisions of great import, we come up against a dilemma. Sometimes the people who are suffering the most are also those who aren't competent to make their own decisions. Consider Jukka Varelius' example of Mary:

[86] See Doernberg et al., 2016 for evidence of inconsistency in the way that capacity determinations are made for patients requesting euthanasia or physician-assisted suicide.

> Mary is a psychiatric patient who has repeatedly tried to kill herself. Once again, her suicide attempt failed. […] Though they are unable to have meaningful contact with her, the mental health care providers are also convinced that, when she is not sedated to near unconsciousness, Mary is suffering unbearably. And they deem her condition incurable. Consequently, although this would be against the common psychiatric goal of suicide prevention to which they have adhered to so far, some of the mental health care providers treating Mary have started to wonder whether they should assist her in ending her life rather than aim to prevent her from killing herself.[87]

Mary is clearly suffering greatly, her condition has been deemed incurable, and although she does not possess the mental capacity to give genuine informed consent or to even make a request for physician-assisted suicide, Mary has clearly and persistently expressed a wish to die through her actions.

This presents the following problem: ending one's life early is an irrevocable decision of immense import, the sort of decision that seems to require that the person making it be able to do so rationally and in accordance with her own values and aims. Ending one's life early is also the sort of decision that requires weighty reasons in order to be justified. In medical contexts, extreme and intractable pain is often considered to constitute such a reason. However, in

[87] Varelius, 2015:2.

Mary's case, it seems that she has a very good reason to opt to end her life early in that she is suffering greatly, but the very thing causing her suffering also precludes her from being able to give informed consent to have her life ended.

It's unclear what we ought to do in a case like Mary's. On the one hand, she is clearly suffering greatly and wishes to die. This is apparent, even though she is incompetent to consent to medical aid in dying. And, it seems that medical professionals do have a duty of beneficence to Mary, a duty that would be best fulfilled in such a case by aiding her in dying, either through PST or more direct means. On the other hand, providing medical aid in dying to suicidal but incompetent psychiatric patients seems like an undesirable and even dangerous precedent to set. This is not to assume some nefarious motivations on the part of medical professionals. Rather, it is a hesitancy that is rooted in a concern for human fallibility. It would be unfortunate if patients whose suffering could have eventually been alleviated or who would in fact regain competence at some point were aided in dying before this change had the opportunity to take place. Note that this does not mean that competent patients who are able to autonomously consent to PST due to psychological suffering ought to be kept alive in the hope of some treatment, however unlikely, being developed to alleviate their suffering. In such cases, the patient is consenting not only to receive medical aid in dying, but also to receive such aid in dying while knowing that *there is the possibility, however slight, that her pain may someday be alleviated.* Because there is no way for an incompetent psychiatric patient to consent under such a caveat,

considerations of eventual expansion of treatment options should take a greater role in deciding whether aid in dying is warranted.

One might be inclined to say here that the problem could be solved in many cases by simply allowing proxy consenters to choose a medically-assisted death for patients like Mary. If such proxies are choosing out of a concern for Mary's interests and well-being, then they could make this choice on the assumption that it is best for those patients and in line with what those patients themselves would want, were they competent. (Although, if the patients were competent, they likely wouldn't have the same weight of circumstances pushing them towards choosing death, so this may be seen as a bit of a puzzling standard under the circumstances.) However, I do not think that allowing a proxy consenter to make such a decision solves the underlying problem, at least not entirely. This is so for a couple of reasons. First, it may be unfair to saddle proxies with such a decision. While making life and death decisions on behalf of loved ones is always difficult, doing the same in a situation where not only is no terminal illness present, but additionally there is so much epistemic uncertainty regarding refractoriness of suffering and what the patient would want were she competent seems even more difficult. Second, the special relationships between most patients and their proxies (who are often parents, guardians, or other family members) may seriously and troublingly complicate decision-making. For instance, the emotional and sometimes financial toll that severe mental illness can take on relationships and the lives of those who support mentally ill family members are the sort of things that might lead a proxy to rationalize aid in dying

in situations that might be borderline, or in which medical aid in dying might be inappropriate, out of sheer exhaustion or frustration. This is especially so in places like the United States where mental healthcare is inadequately funded and so the financial burden of illness must be largely shouldered by families. Such circumstances compound the unfairness of saddling proxies with making this decision.

As uncomfortable as I am with such philosophical positions in general, I am inclined to say that in this sort of case providing medical aid in dying to incompetent psychiatric patients whose suffering is refractory, severe, and continuous, and who have expressed a consistent, persistent, and unwavering wish to die through their actions and/or words might be the morally best choice on the part of *individual medical providers*. However, I am not sure I could accept the implications of legislating such aid and/or explicitly incorporating it into medical practice. Here I am endorsing something similar to something David Velleman proposed where medical euthanasia is permitted "by tacit failure to enforce the institutional rules that currently serve as barriers to justified euthanasia" rather than "an explicitly formulated permission" in the form of policy or law.[88] In Mary's case, her medical providers might be morally justified in assisting her suicide, but it might nonetheless set a dangerous precedent to incorporate such assistance into hospital or legal policy.

[88] Velleman, 1992:680

Concluding Remarks

I have argued that a moral commitment to the permissibility of PST to treat psychological suffering at the EOL further rationally commits us to the moral permissibility of PST to treat psychological suffering earlier in life and in the absence of terminal illness. However, this entailment raises a whole host of problems, among them how to determine sufficient severity and refractoriness of psychological pain, how to know which treatment measures to characterize as heroic and thus unwarranted, when and whether psychological illness precludes informed consent, and whether PST might ever be justified in the mentally ill who are suffering greatly but are not competent to give informed consent. In the face of such challenges, the solution is not to ban PST wholesale and thus break the entailment. This would cause great unnecessary suffering for some of our most vulnerable patients and would further make the already unpleasant process of dying even more arduous for patients in the United States for whom PST is often the only legal available option for medically-assisted dying.

Instead, a better route to take in resolving the difficulties posed in this project would be to compensate for the unique epistemological uncertainties that apply to cases of early, but not EOL, PST. Given that refractoriness of suffering especially is so much more difficult to determine with certainty when the patient is not imminently dying, it makes sense to approach determinations of whether early PST is warranted with more caution than we would in making such determinations at the EOL. One way to so this might be to simply to flesh out and

make explicit the application of different standards for the severity of pain that warrants medically assisted dying depending on the patient's proximity to death.

That is, we might require a higher severity of suffering (perhaps even approaching the level of literal unbearableness) to employ PST earlier in life in someone who is not terminally ill than we would apply to someone who is elderly and/or terminally ill and is expected to die within weeks or months (for whom discomfort judged intolerable would provide a sufficient standard of severity). This might in a way "correct" for the uncertainty issues that accompany determinations of refractoriness in non-terminal situations of severe psychological suffering.

Perhaps at a certain distance from death and/or in the absence of terminal illness, one's suffering would need to be the sort of thing that is the focal point of her entire life in order to warrant ending her life on that basis. This degree of suffering would seriously compromise the patient's autonomy and well-being to the point that she could not do much of anything aside from suffer. The importance of alleviating such debilitating psychological pain would be so great as to outweigh future-oriented considerations such as the possibility (which, in the cases under consideration where pain is judged to be refractory, would likely be exceedingly slight) of the discovery of a way to ameliorate the patient's symptoms or cure the underlying issue. That is, suffering so severe as to constitute the entirety of a person's perception of her existence would justify PST in cases where there is substantial uncertainty regarding the accuracy of refractoriness judgments due to an extended timeframe and the possibilities

contained therein. Further, it seems that the uncertainty surrounding determinations of decision-making competence would pale in importance when compared to a sufficiently high degree of suffering. If a patient is suffering to such an extent that she cannot focus on anything beyond her suffering, and if this suffering is unlikely to ever abate, then it seems that the possibility that this suffering might cloud the patient's decision-making judgment is less important than the fact that the patient's suffering ought to be relieved, even if to do so would necessitate ending the patient's life. In this way, the severity of pain required to justify early PST would compensate for the uncertainty inherent to judgments of refractoriness and decision-making competence outside of an EOL context.

But, the closer a patient gets to death, the laxer such standards for suffering should grow, until a patient who is expected to die within days or hours might be able to opt for PST due to any sort of discomfort. After all, if someone's imminently dying, what's the point of making things more difficult on her? The severity of suffering required to warrant PST will thus correspond inversely to a patient's expected remaining lifespan. It is of course difficult to determine the precise severity of suffering, but a patient's expressed level of pain combined with a questionnaire designed to measure health-related quality of life like the EQ-5D (and perhaps a variant designed to measure the impact of mental

disorders) could enable us to make some reasonable ballpark judgments regarding severity.[89]

A natural question to ask as this paper draws to a close is whether I think there is any reason to favor PST for patients who are experiencing severe and refractory suffering over voluntary active euthanasia (VAE). The answer to this question understood in general moral terms is no. I don't see any fundamental moral difference between PST and VAE, so in all of the cases where PST will be morally permissible, so will VAE. However, I think there may be good reason to favor PST over VAE under particular circumstances. That is, while PST and VAE may both be morally permissible, PST may be preferable in light of a situation's particulars (there will also be situations where VAE is preferable). I will give some examples of situations where PST may be preferable to VAE, although both would be morally permissible. Firstly, in EOL situations, if there is a real chance that the patient's condition may be reversible, it may make sense to employ PST rather than VAE in order to allow the time necessary to determine whether there is any way to solve the underlying problem and prolong the patient's life (assuming a resulting QOL that would make life extension desirable to the patient). For instance, suppose an experimental medication that might prove helpful to the patient may or may not be approved for use within the near future. In such a case, PST might be employed to relieve the patient's suffering temporarily while waiting to see if the medication will be made available. Further,

[89] See, e.g., Bognar and Hirose 2014:33-36 for information on the EQ-5D.

in situations where the patient or her family may be uncomfortable with the explicit idea of euthanasia or assisted suicide, PST may be a useful tool for alleviating the patient's suffering without causing guilt on the part of the patient or her family.

There may also be situations where (painlessly) prolonging death may be indicated for non-medical reasons. Perhaps opting for PST rather than VAE would allow family members to come say goodbye to a patient (although this would clearly only be valuable for those family members since the patient would be unconscious). Even if the patient herself is not aware that this is taking place, if before her deterioration she would have found it valuable for those family members to be able to say goodbye and get a sense of closure through this, perhaps we could assume that the patient would have consented – were she capable – to dying more slowly in order to allow this to happen. Perhaps this is even something that a particularly family-oriented patient would want included in an advanced directive (although given the reality of advance directives and the lack of fine distinctions included in most, I doubt this would actually become common in practice).

My aim in this paper has been to show that a commitment to the moral permissibility of PST to alleviate severe and refractory suffering in terminally ill patients further rationally commits us to the moral permissibility to alleviate severe and refractory psychological suffering in the absence of terminal illness. I take this to be a troubling entailment and explored exactly why it is troubling by looking at challenges to determining refractoriness, defining heroic measures, and

obtaining informed consent. In light of these challenges, I have sketched out a solution wherein the severity of suffering necessary to warrant PST should be inversely proportional to the estimated remainder of the patient's life.

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CARDIAC PACEMAKERS AND WITHDRAWAL OF CARE AT THE END OF LIFE[90]

1. Introduction

Cardiac implantable electronic devices (CIEDs) are a part of life for millions of patients.[91] Given the advanced age of most of this patient population, many individuals with CIEDs face end-of-life decision making in medical contexts. By this I mean that many patients with CIEDs (or their proxy consenters if patients are incompetent to make their own decisions) must make decisions regarding the circumstances under which they would like to receive or forego care, and circumstances under which they would like care to be withdrawn at the end of their lives. These decisions are ideally made while working out an advance directive covering such situations, such as a Do Not Resuscitate (DNR) order which stipulates that the patient does not wish to be resuscitated if he is found unconscious and/or undergoes cardiac arrest while in care. In practice, however, advance directives rarely mention CIEDs.[92]

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[90] Special thanks to Dr. Robert Gipe, MD, PhD for medical advising on this project. His input on drafts has been invaluable.

[91] "More than 4.5 million people worldwide live with an implanted pacemaker, including >3 million in the USA alone. Also, >0.8 million people in the USA have an implantable cardioverter defibrillator." (Benjamin and Sorkness, 2017: 157)

[92] See Pasalic et al., 2014; Buchhalter et al., 2014:5.

Compassionate end of life (EOL) care often necessitates foregoing or withdrawing treatments which become burdensome or simply unnecessary as the patient draws closer to death. As the body shuts down, burdens of many treatments become more pronounced and benefits become reduced to nonexistent. This paper deals with the ethical complexities surrounding one type of withdrawal of care: the deactivation of implantable cardiac electronic devices, specifically pacemakers, at the end of a patient's life. I hold, roughly, that pacemakers are unique in their function among CIEDS and this gives them a unique moral status when it comes to situations of withdrawal of care at the EOL.

I will argue that deactivating a pacemaker at the EOL is importantly morally different from, for example, deactivating the shocking function of an Internal Cardioverter Defibrillator (ICD) or removing a patient from a ventilator. Specifically, deactivating a pacemaker at the EOL will characteristically either impose additional quality of life related burdens on the patient or will have the same moral status as voluntary active euthanasia (VAE) performed at the EOL. This is so for a few central reasons: (1) There is good reason to conceive of pacemakers as "biofixtures" (in contrast to a feeding tube or – as I will argue – even an implantable cardioverter defibrillator). A pacemaker is more analogous to a biofixture like a porcine valve than it is to a ventilator. And, the fact that one (the pacemaker) is easier to "deactivate" than the other (the porcine valve) does not mean that the pacemaker should not be understood as a biofixture; (2) A pacemaker is a low-burden intervention that does not warrant removal in a comfort care situation; and (3) There is typically little benefit to deactivating a

pacemaker, unless the express goal is to bring about or hasten the patient's death. While it is true that the express goal in some standard cases of withdrawal of care may be to lead to the patient's death, what is different in the case of pacemakers is that there is no good reason for deactivation *aside* from hastening or bringing about death in a highly dependent patient. This is because considerations of the patient's discomfort that often drive withdrawal of care in standard cases do not apply in the case of pacemakers.[93] If a patient is highly dependent upon the pacemaker, he will pass away quickly after deactivation. And, if he is not highly dependent, deactivating the pacemaker is likely to adversely affect his quality of life. There seems to be little reason to deactivate a pacemaker at all for a patient who is not dependent upon it to live, and the motivation behind deactivating a pacemaker that a patient is dependent upon to live seems to be clearly to end that patient's life. If, as I will argue, deactivation of pacemakers at the end of life is typically either useless (and even harmful) or akin to VAE, this will have important implications for the conditions under which such deactivation is morally permissible and, ultimately, what guidelines ought to be put into place for dealing with pacemakers at the end of life. I will assume that most if not all cases of non-pacemaker-related autonomously patient (or proxy)-chosen withdrawal of

[93] It is true, of course, that other motivations may drive withdrawal of care. For instance, one may withdraw care at the end of life out of respect for a patient's autonomy in accordance with an advance directive, or with the sole aim of allowing the patient to die of his underlying disease. However, the burdens of treatment often do play a weighty role in determining whether withdrawal of care is warranted.

care at the end of life are morally permissible. I will also assume that there is a moral difference between withdrawal of care and VAE or physician-assisted suicide.

Suppose a patient's family requests that a physician deactivate their relative's pacemaker at the end of life. What should the physician do? Bioethicists have characteristically argued that deactivating a pacemaker at the end of a patient's life would be no more morally problematic than taking the patient off ventilator support or removing a feeding tube.[94] Moreover, a recent expert consensus statement from the Heart Rhythm Society (HRS) states that deactivation of CIEDs at the end of life is legally and morally permissible. They argue this on the basis of nine basic ethical and legal principles, most of which I find acceptable. However, this statement misguidedly lumps together cardiac devices with importantly different functions and relations to the patient's body. I take particular issue with two of the principles to which the HRS appeals, but only in their specific application to cardiac pacemakers as distinct from other CIEDs. I will briefly highlight central problems with these principles and then proceed to argue for the status of pacemakers as biofixtures, the uselessness of deactivating pacemakers in comfort care situations, and the inaccuracy (and even disingenuousness) of characterizing pacemaker deactivation as just another case of EOL withdrawal of care when their status as biofixtures makes pacemaker deactivation morally akin to removing a transplanted organ, which would seem to

[94] See, e.g., Zellner et al., 2009.

constitute VAE rather than withdrawal of care. Here are the two HRS principles under consideration and my preliminary responses to each:

- [a] Ethically and legally, there are no differences between refusing CIED therapy and requesting withdrawal of CIED therapy.[95]

I will challenge this premise by conceptualizing pacemakers as biofixtures. One can refuse an organ transplant but not have a transplant withdrawn. This is because the organ transplant, being a biofixture, is bound into and plays a constitutive replacement role in the patient's body. So, withdrawing that transplant (or even compromising the functioning of that transplant through medication), would be akin to introducing a new pathology rather than a simple case of withdrawal of care. If, as I will argue, pacemakers likewise have biofixture status and are related to the patient's body in the same way as an organ transplant would be, then withdrawing pacemaker therapy (through either deactivation or removal) would be likewise akin to introducing a new pathology. In this way, there is an important moral difference between refusing and withdrawing pacemaker therapy that is akin to the moral difference between refusing and withdrawing an organ transplant.

One of the possible concerns on the part of clinicians that the authors of the HRS guidelines address is the question of whether withdrawing a CIED therapy is akin to assisted suicide or euthanasia. As Lynn Jansen notes, many clinicians take there to be an important moral distinction between killing and letting die and thus euthanasia and physician-assisted suicide on the one hand and withdrawal of care

[95] Lampert et al., 2010:1009.

on the other; moreover, hospital policy and law often take seriously the moral importance of such a distinction.[96] Those who support the claim that pacemaker deactivation at the EOL is morally akin to withdrawal of care also typically assume that there is a morally important distinction between VAE and withdrawal of care and the authors of the HRS guidelines take such a distinction seriously. In this paper, I will operate under the assumption that this distinction is correct and will argue that pacemaker deactivation at the EOL is morally akin to VAE rather than withdrawal of care. The second of the HRS's principles under consideration here explicitly makes such a distinction:

- [b] Ethically, CIED deactivation is neither physician-assisted suicide nor euthanasia. When carrying out a patient's request for withdrawal of a life-sustaining treatment that a patient perceives as unwanted (including CIED therapies), the clinician's intent is to discontinue the unwanted treatment and allow the patient to die naturally of the underlying disease - not to terminate the patient's life.[97]

This principle appeals to the clinician's intent in order to differentiate CIED deactivation from euthanasia. And, intent is often what grounds a moral distinction between withdrawal of care and VAE.[98] But, it is important to also account for the wider landscape in which CIED deactivation takes place, namely when the patient is already at the end of his life. As I will argue, it doesn't make

[96] Jansen 2016, 106.

[97] Lampert et al., 2010:1009.

[98] See, e.g., McMahan, 1993.

any sense to say that deactivating the pacemaker would allow a highly pacemaker dependent patient to die naturally of the underlying disease *that is actually killing him*. This is because, in correcting for slow heart rate (bradycardia) the pacemaker serves to prevent heart failure from developing in the first place. In a highly dependent patient, deactivation would serve to cause the patient to die of a chronic underlying illness, but one that had been managed – often for years – by an implanted biofixture. In such a context, deactivation is more akin to the introduction of a new pathology which kills the patient than to allowing the patient to die of an underlying terminal illness. And, if the patient is not highly dependent upon his pacemaker, all that deactivation would accomplish would be to worsen the patient's quality of life unnecessarily.

2. Pacemakers vs. ICDs

It is important to be clear on what exactly is under discussion here. "CIED" is a blanket term which covers all implantable electronic cardiac devices. Both pacemakers and Implantable Cardioverter Defibrillators (ICDs) are implantable devices that help to control arrhythmias (abnormally slow, fast, or irregular heartbeat). Pacemakers provide a pacing function wherein they regulate slow heart rhythms. ICDs monitor the heart's rhythm and deliver a shock if the heart reaches a dangerously fast rate. Current ICD devices always have a pacing function as well, although pacing is neither used nor needed in most ICDs. In ICDs, the shocking mechanism and the pacing function can be deactivated independently. For the purposes of this paper, when I refer to pacemakers I will be talking about both the discrete implantable device that serves as a pacemaker

and the pacing function of ICDs. The conclusions I draw regarding pacemakers will thus apply equally to the pacing function of ICDs, although most ICDs do not perform an ongoing pacing function.

Much of the literature on the moral status of deactivating cardiac devices at the end of life has focused on the less controversial case of implantable cardioverter defibrillators (ICDs).[99] Deactivating an ICD is less morally fraught than deactivating a pacemaker because near the end of life ICDs can often give the patient a series of painful shocks which provide clear justification for deactivating the ICD out of a concern for the patient's comfort: "In the last weeks of their lives, twenty percent of ICD patients receive shocks which are painful and known to decrease quality of life, and which greatly contribute to the distress of patients and their families."[100] Understood in terms of burdens, the pain of shocks at the EOL overrides the potential benefits. This is especially true in the most common cases where the patient is shocked multiple times. Here the ICD attempts to correct for arrhythmias that cannot be permanently corrected for as the patient's heart is failing. This leads to a series of repeated and painful shocks.

Further, in cases where a patient has an advance directive with a DNR specifying that resuscitation be foregone in cases of cardiac arrest, it makes sense that this would apply to an ICD as well. If the patient wants to forego *external* defibrillation due to the potential burdens (especially pain of shock) of such, it

[99] See, e.g., Daeschler et al., 2015; Lampert, 2015; Strömberg et al., 2014; Svanholm et al., 2015

[100] Lampert et al., 2010: 1008.

makes sense that the patient would also want to forego *internal* defibrillation that would deliver a series of painful shocks to him. In both cases, patients do not want their lives extended by treatment of a cardiac arrest and would rather expire rapidly.

3. Pacemakers as Biofixtures

The moral status of pacemakers and the moral permissibility of deactivating them depends heavily upon the relation that these devices have to the patient. Assuming a moral distinction between killing and letting die, stopping a patient's heart with an injection at the end of life would be morally different from withdrawing care such as ventilator support. This is because the actions taken are different, the justification behind these actions are typically different, and (of special relevance to the argument at hand) the nature of the component being compromised in or removed from the patient is different. Withdrawal of ventilator support constitutes a situation where an artificial method of life support is discontinued in order to allow the patient to die of his underlying illness. Stopping the patient's heart is a situation where a native fixture of the patient's body is being actively compromised in order to cause the patient's death. It is clear in the latter case that such interference with the functioning of a bodily system would uncontroversially amount to euthanasia. It is further clear that stopping a *transplanted* heart through the same method would likewise be tantamount to euthanasia.[101] There is no morally significant difference between the heart a patient is born with and the heart a patient receives as a transplant when it comes

[101] I owe this example to Sulmasy, 2007:71.

to questions of cessation or withdrawal of treatment at the end of life. A transplanted heart is a paradigm example of a *biofixture* – something that has become a part of the patient such that the way we are morally permitted to treat that thing will not differ from the way in which we are permitted to treat other parts of the patient that are necessary to sustain life. In the same way as compromising the functioning of an organ in order to bring about a patient's death would amount to euthanizing the patient, so would doing the same to a transplanted organ. But what about deactivating a pacemaker? The pacing that would be stopped upon deactivation serves the same function that would be stopped by an injection to a healthy heart. In both cases, an abnormal heart rhythm is produced through intervention and, in the case of pacemaker deactivation, the patient will likely die of severe bradycardia (slow heart rate) or asystole ('flat line'), at least if the patient is highly dependent upon his pacemaker to live. Does this mean that a pacemaker is a biofixture like a transplanted heart? In order to answer this question, we must pin down what is required in order to count as a biofixture and determine whether pacemakers fulfill those criteria.

The notion of a biofixture was first put forward by Frederick Paola and Robert Walker.[102] Paola and Walker draw on the notion of property law to draw an analogy between fixtures of property and biofixtures in human beings. Daniel Sulmasy proposes criteria for determining whether a technological intervention is a part of the patient (i.e. a biofixture).[103] According to Sulmasy, for a

[102] Paola and Walker, 2000.

[103] Sulmasy, 2007.

technological intervention to be a part of the patient, it must be a *constitutive* therapy which is also a *replacement* therapy. A therapy being constitutive is best understood by contrast with regulative therapies. An ICD is a regulative therapy because it only operates intermittently to shock and reset the heart when necessary; in doing so the ICD "coax[es] the body back towards its own homeostatic equilibrium".[104] Constitutive therapies, by contrast, "take over a function that the body can no longer provide for itself".[105] A pacemaker is a constitutive therapy because it stimulates a continuous heart rhythm in essentially the same way the heart would; pacemakers replace the function of the conduction system of the heart in much the same way that a heart transplant replaces the overall function.

Under the heading of constitutive therapies, Sulmasy draws a further distinction between substitutive therapies and replacement therapies. He argues that for something to count as a part of the patient, it must constitute a replacement therapy, which he characterizes as follows: "The most important feature of a replacement therapy is that it provides the function that has been pathologically lost, more or less in the same manner in which the patient was once able to provide this function when healthy".[106] A substitutive therapy, by contrast, provides a substitute for some function in the body that does not resemble the way in which the body provides that substitute for itself. Hemodialysis for kidney

[104] Ibid:70.

[105] Ibid.

[106] Ibid: 71.

failure and insulin injections for diabetes are examples of substitutive therapies. Sulmasy provides the following indicators of whether something constitutes a replacement therapy:

> Additional signs suggestive of an intervention being a replacement therapy might include: (1) its responsiveness to changes in the organism or its environment, (2) properties such as growth and self-repair, (3) independence from external energy sources and supplies, (4) independence from external control by an expert, (5) immunologic compatibility, (6) physical integration into the patient's body.[107]

Sulmasy characterizes these properties as indicators and so it seems like they are meant to be used essentially as rules of thumb for identifying replacement therapies. Nonetheless, it is evident that HRS has adopted Sulmasy's indicators as *criteria* for replacement therapies.[108] Others in the debate (e.g., Kay and Bittner 2009; Zellner et al. 2009) also seem to understand Sulmasy's indicators as criteria.[109] In light of this misunderstanding, it is worth looking at the entailments

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[107] Ibid: 71-72.

[108] E.g., "A replacement therapy (e.g., kidney transplantation) literally becomes "part of the patient" and provides the lost function in the same fashion as the patient did when healthy. Replacement therapies also respond to physiologic changes in the host and are independent of external energy sources and control of an expert. Removing or withdrawing a replacement life-sustaining treatment has been characterized as euthanasia." (Lampert et al., 2010:1012)

[109] E.g., "[A] "replacement" must be capable of growth and self-repair and must be independent from external energy sources and expertise. Pacemakers are not capable of growth or self-repair. They rely on batteries that deplete. Pacemakers

of accepting these indicators as individually necessary and jointly sufficient criteria for pacemaker status. Moreover, even if these are just meant to serve as heuristics for identifying biofixtures, I take issue with the implications of indicators (2-5).

Indicator (2) basically amounts to a presupposition that a biofixture must be made of flesh, blood, and/or other organic materials (or something similar enough to them) to be integrated into the body in the same way that an organ, bone marrow, blood, or stem cells would. This is too restrictive a definition to be plausible and will not serve for the future. It seems merely to reinforce unfounded pre-theoretical intuitions about the importance of material over function and is not based in a reality where progressively better artificial therapies and materials are being developed. Both growth and self-repair serve as inadequate focal points when what is of far greater importance to biofixture status is the relation between the fixture in question and the surrounding body. Integration to the body is something that is universal to biofixtures, while growth and self-repair apply to only a limited class of biofixtures. Artificial joints cannot grow or repair themselves, but they are surely a part of the patient's body in the sense relevant to biofixture status.

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are subject to malfunction, often need expert intervention, and are subject to recall. Thus, pacemakers are not "replacements."" (Zellner et al., 2009: 339)

Now suppose that a permanent artificial heart developed out of entirely synthetic materials were approved for widespread use in patients in need of heart transplants. (Although artificial hearts are now mostly used as a temporary measure while patients await heart transplants, the technology is developing at such a rate that I think it would not be surprising to see permanent artificial heart transplants become widespread within our lifetimes.)[110] Suppose further that this artificial heart (being implanted only in adults) remained one size and, rather than repairing itself, if damaged had to either be repaired by the surrounding body or through surgery. It seems that this artificial heart would clearly be a biofixture, despite lacking the capacity for growth and self-repair. It would be bound into the body both by vascular pathways and scar tissue and would fulfill the same role in the body that a heart made of organic tissue would. The only substantive difference would be what the heart was made of, and this alone does not seem enough to call the biofixture status of this artificial heart into question.

Indicators (3-4) would require further specification in order to be genuinely helpful in identifying biofixtures. Depending on how dependence upon external energy sources and expert control are characterized, this could have the effect of ruling out many interventions that require charging and/or monitoring, even if this is infrequent and only takes place during regular check-ups. Of course, if there were some sort of device that required constant remote control by a professional or had to be plugged into a wall in order to function, this would seem to call into question its status as a replacement therapy. However, devices

[110] See Struber et al., 2009; Copeland et al., 2004.

with batteries can run for months or even years without needing to be recharged in some cases. Typical pacemakers, for instance, have 8-12 years of battery life and most pacemakers require very little adjustment after the initial three months.[111] And, some devices, while they may require regular calibration from a professional, may not need the sort of constant control or adjustment that would seem to make a fixture a substitutive therapy rather than a replacement therapy.

Further, indicator (5) immunologic compatibility, understood strictly, may rule out most organ transplants from counting as biofixtures. This would be an undesirable result since it seems that if anything should count as a biofixture, organ transplants should. However, with organ transplants, in all but the most exceptional cases of immunologic matching, the patient must take immunosuppressants for the rest of his life in order to keep his body from rejecting the transplant.[112] By contrast, something like a titanium knee replacement would, strictly speaking, be more immunologically compatible with a patient than would a standard organ transplant, since an artificial knee does not require that the patient take immunosuppressants in order to keep the body from rejecting it. The immunologically benign nature of such inorganic materials leads to the odd entailment that if we chose to adopt immunologic compatibility as one of the individually necessary and jointly sufficient criteria for something to count as a biofixture, and further construed this requirement in terms of *strict* immunologic compatibility, then immunologically benign fixtures made of metal

[111] See, e.g., Ganz and Hayes, 2018.

[112] See Enderby and Keller, 2015; van Sandwijk et al., 2013

or plastic would count as biofixtures while organic 'fixtures' like organ transplants would not count as biofixtures unless they were a near-perfect immunologic match to the patient.

I hold, like Sulmasy, that a biofixture must constitute a constitutive therapy and a replacement therapy. That is, a biofixture must fulfill a constitutive function and be a replacement that does so in roughly the same way the system it is replacing would. However, I disagree on how exactly replacement therapies ought to be characterized. I will here endorse a wider conception of what it is to be a replacement therapy. This conception neither implicitly nor explicitly implies that for something to count as a replacement therapy it must be constituted of organic material. To this end, I will adopt and expand upon two of Sulmasy's indicators: (1) "responsiveness to changes in the organism or its environment" and (6) "physical integration into the patient's body" as indicators of something counting as a replacement therapy. I will further stipulate that, in order to be *a biofixture worth having*, a fixture ought not impose burdens on the patient disproportionate to the burdens imposed by the systems or functions that it is replacing, and further that the burdens imposed by the biofixture ought not be disproportionate to the benefits of having that fixture in place.

Regarding indicator (1), I want to specify that for an object to count as a replacement therapy, it must respond to changes and it must respond roughly in the way that the function it is replacing would. It needn't be responsive to each and every change in the organism. Many systems operate more or less independently within the body, or at least can still function in the face of a failure

of other systems. A replacement therapy need only be responsive within the ordinary domain of the function it is replacing. This means that a knee replacement would only need to be responsive to changes in the adjoining bones and muscles (e.g. adjusting to shifting weight, moving with muscle tension, etc.) and a porcine valve would only need to be responsive to changes in the surrounding heart (e.g. accommodating higher or lower blood pressure). In the same way, a pacemaker replacing the pacing function of the heart would need to be responsive to fluctuations in heart rhythm and not (at least not in any direct way) to, e.g., changes in muscle tension in the patient's foot.

Regarding indicator (6), I hold that while it is the case that something needn't be strictly or wholly internal to the patient in order to count as a biofixture, it must be integrated with the patient in such a way that it is attached the patient's body in a semi-permanent to permanent manner and removal would be invasive. There are constitutive therapies that are more or less integrated into the patient's body. Hemodialysis or ECMO rely on machines that must be "hooked up" to the patient and which, aside from an access point, are entirely external to the patient. Something like a hip replacement, porcine heart valve, or pacemaker seems to be more integrated into the patient's body, and, further, they would be even if part of them were external. If there were some sort of pacemaker that had an external display that sat on top of the patient's skin or a line that connected the device to a small console (as ill-advised as this might be in light of infection concerns) the mere fact that the entire device is not internal to the patient would not change the device's status as a biofixture. It would be sufficiently

integrated to the patient and removing the part below the skin would be invasive, requiring surgery. Pacemakers sit below the skin and become further integrated into the patient's body by the encapsulation of the pacemaker itself and connected leads by scar tissue, so the integration requirement for counting as a replacement therapy has been met in this case.

Someone might argue that the reason one cannot request that an organ be withdrawn is that to do so would be expensive and difficult, which would render it untenable in practical terms.[113] However, deactivating a pacemaker is neither expensive nor difficult. So, biofixture or not, a pacemaker may be deactivated in situations where removing an organ would be pragmatically untenable. Regarding pacemakers, there is an important distinction to be made between the invasiveness and challenges of *removal* and those of *deactivation*. A pacemaker is easy to deactivate (although far more complicated to physically remove from the patient's body), and other candidates for biofixtures may be as well. However, when we're talking about something being integrated into the patient's body, the ease with which it can be made to quit working – as opposed to the ease of removal – is not a consideration relevant to the level of integration. Plenty of native body parts can be incapacitated and caused to fail by relatively easy and minor interventions like

[113] One representative example of such an argument: "Deactivating a pacemaker is non-invasive and does not introduce a new pathology. Removing an implanted porcine valve, however, is invasive and introduces a new pathology (i.e., a sternal wound). Thus, in this context, it is permissible to carry out requests to withdraw CIED therapies from patients who no longer want these therapies." (Lampert et al., 2010: 1012)

the administration of certain drugs. This does not mean that those organs and systems are less integrated into a patient's body than other less easily compromised organs and systems. In the same way, the fact that a pacemaker is convenient to deactivate and a porcine valve would require surgery either to remove or otherwise seriously compromise its functioning does not mean that the pacemaker is somehow less integrated into the patient's body than the porcine valve. The mere ease with which someone could stop a patient's heart or cause liver failure through drug-induced means does not mean that doing so would be different in moral terms from removing the patient's heart or liver surgically. In the same way, the mere fact that pacemaker deactivation is easy and cheap does not mean that it is different in moral terms from surgically or medically compromising the functioning of a patient's heart. The ease with which a pacemaker can be deactivated may make the decision to deactivate a pacemaker at the end of life less psychologically difficult than the psychotic-seeming removal of a porcine heart valve in the name of withdrawal of care would be, but this psychological phenomenon does not point to anything morally significant.

Comparable Burdens

I will now move from expanding upon two indicators I have adopted from Sulmasy as criteria for biofixture status to a criterion of my own. In the same way that, in order for a heart to be a heart worth having, it ought to fulfill the basic functions of a heart without imposing disproportionate burdens on the patient, so in order for a transplanted heart to be a heart worth having, it ought to do the same. To extend this analogy to biofixtures generally, in order for something to be

a biofixture worth having, it ought not impose disproportionate burdens on the patient.[114] If a native heart is failing, it ought to be either repaired or replaced. A biofixture that replaces the function of a failing heart should be at least competent to stave off death or serious suffering, and, ideally would bring the patient up to

[114] Now, it's important to note here that burdens can be understood in different ways. One straightforward way a device or treatment might be burdensome would be by inflicting pain on the patient. An intervention might also be burdensome in the way that it interferes with the patient's normal functioning - even if it does not inflict pain on the patient. A device that physically interferes with a patient's movements but does not actually cause the patient pain would fall under this heading. An intervention might likewise be burdensome in virtue of the time it requires from the patient in terms of maintenance and upkeep. If an intervention didn't have any effect on the patient's physical normal functioning but required the patient to travel to a clinic constantly for checkups, this would impose a burden on the patient. An intervention that required a patient to be hospitalized would likewise be burdensome, regardless of whether or not the intervention itself were cumbersome or painful to the patient. And, of course, there is the matter of financial burdens of treatment. While I will be largely bracketing financial issues in the current paper, it is important to bear in mind that financial considerations may weigh heavily in EOL decisions on the part of patients and their families. However, such financial burdens are imposed by an often dysfunctional health economic system rather than being a result of the intervention or treatment itself. While it may be totally rational to choose to withdraw care on financial grounds, explicit consideration of financial burden is best left out of the current project. Suffice it to say that a patient's being forced to choose between an intervention that is unobjectionable to her and the financial solvency of her family is an extremely important moral problem, but one that is tangential at best to the project at hand.

the quality of life he would have enjoyed with a healthy heart. Functioning well qua biofixture requires that such a fixture not be the sort of thing that the patient would be better off not having. Now, in the same way that a heart's functioning poorly and painfully in a patient doesn't undermine that heart's status as a body part, a biofixture that functioned poorly and/or painfully would not necessarily thereby lose its status as a biofixture. However, a biofixture that functioned poorly enough or imposed enough burdens on the patient might rightfully lead us to question whether it can be properly characterized as a biofixture at all. On the account I propose here, some devices might perform so poorly as to fail to qualify as biofixtures, and some devices, although they perform well enough to strictly-speaking qualify as biofixtures, will not perform well enough to count as biofixtures worth having.

The requirement that something reach a threshold of proper functioning in order to be properly considered a biofixture can be made sense of, at least in part, in light of the requirement (adopted from Sulmasy) that in order for something to count as a replacement therapy, it must serve the function of the thing it is replacing in roughly the same way that thing originally fulfilled (or would have fulfilled) the function. If a transplanted heart had a weird defect where it made an earsplitting and persistent whistling noise every time the patient's heart rate went above 80 bpm, one might be led to question how adequate a replacement that heart actually was, despite clearly seeming to be integrated into the patient's body and sensitive to the relevant changes in the body. The same might be asked of a transplanted heart that caused a sharp stabbing pain every time it beat or a

transplanted kidney which somehow produced urine that – despite causing no lasting physical harm to the patient – had roughly the consistency of wet sand. These sorts of shudder-inducing examples are admittedly fanciful, but the point here is that burdens of a thing may actually compromise that thing's functional status, despite the defects or additional burdens in question not strictly compromising the essential functioning of that thing. Crafting scissors that cut paper like a dream but also had a 40% chance of removing a fingertip or two might be very good qua paper cutting scissors but not qua scissors, period. And, a particularly creaky church pew might be good at holding one's body up in a sitting position, but one would probably call its fulfillment of its function into question while trying one's best not to shift one's weight around at a particularly solemn funeral. In the same way, a pacemaker that made an obnoxious noise or had flashing LED lights that constantly shone through the patient's skin might be good qua heart-rhythm-replacer but not necessarily qua biofixture. Presumably one important part of replacing the function of something in roughly the same way the original thing functioned is that the replacement doesn't come along with additional disproportionate or even intolerable drawbacks.

In the same way that there are better and worse functional objects of all sorts, there are surely better and worse biofixtures. This is evident from the fact that a highly immunocompatible organ transplant is surely better than a less immunocompatible organ transplant, in that the former will not impose the lifelong burden of immunosuppressant therapy on the patient. However, both of these are totally adequate biofixtures, precisely because they meet the basic

criteria of being constitutive replacement therapies and having benefits that clearly outweigh their burdens. I will here take no stance on just how poorly something must function in order to compromise its status as a biofixture. I will only state that a biofixture worth having ought not to impose burdens disproportionate to its benefits or disproportionate to the burdens that would be imposed by the system it is replacing. And, if something fulfilled its function poorly enough in terms either of its direct medical function or burdens imposed apart from its direct function, that thing might, as a result, not warrant the status of biofixture.

I hold that, for it to be worth incorporating into the patient's body, a biofixture must not be burdensome to the patient to an extent that is substantially greater than the burdens that the system or function it is replacing would impose. Further, a biofixture must at the very least not impose burdens on the patient that outweigh the benefits of having it. Now, depending on the status of the system or function being replaced, and thus the burdens imposed by the patient prior to implantation of a biofixture, the latter may be an easier or more difficult requirement to meet. After all, if a patient has a heartbeat so slow as to make even the ordinary activities of daily life a challenge, a device that imposed significant burdens but allowed the patient to perform the activities of daily life would be preferable to the status quo. Ideally, the biofixture will impose burdens not much more substantial than those imposed by a basically normally-functioning (and normally aging) instantiation of the system or function it is replacing. By this I mean that, in the same way that upkeep of normal bodily functions and systems

may require medication or supplementation or exercise or various lifestyle changes (especially as the patient ages), upkeep of a biofixture might require things to be done of a similar degree of burden or inconvenience. Of course, if the patient would receive only marginal benefit from having a fixture implanted, the risks of surgery should not be undergone. But this is a background consideration.

In order to illuminate this burdens-related requirement, let us return to the example of a transplanted heart. In cases of organ transplant, in all but the most exceptional cases, the patient must take immunosuppressants for the rest of his life. While this is a burden that the patient would not have if his organs functioned normally, the burden imposed here seems proportional to the benefit derived from the transplanted organ. Further, especially as we age, different bodily systems may develop dysfunctions that require medications and monitoring. The burden of having a heart that doesn't work perfectly (as happens with many aging hearts), which may require lifelong medication, seems comparable to the burden of taking immunosuppressants to keep one's body from rejecting a transplanted heart. Medications will have side effects, some more severe than others, but the benefit derived from having a bodily system that functions normally will outweigh these downsides. So, in the case of a transplanted heart, the requirement of immunosuppressants is comparable to the burdens imposed by the system it is replacing, and the benefits of having the transplanted heart outweigh the burdens it imposes. As will be made clear in the next section, pacemakers also impose burdens comparable to the system they replace and the benefits of having a pacemaker characteristically outweigh the burdens that device imposes.

A pacemaker is a low-burden intervention. When a pacemaker is implanted, the surgery itself presents risks to the patient. The healing period lasts about six weeks, in the course of which the patient may potentially dislodge leads or develop an infection. But these complications are rare and can be rectified, although this usually requires surgery.[115] There are risks to having a pacemaker, even once the healing process is over, but these risks are minimal.[116] A patient will have to return in 8-12 years once the pacemaker's battery runs out and the standard of care regarding regular monitoring is to check every three to four months to insure the pacemaker is working properly.[117] These checks can take place either at a clinic or in the patient's home (through a remote monitoring system). Ideally, patients will go through their daily lives forgetting that the pacemaker is even there.

Pacemakers clearly have benefits that outweigh their burdens. A patient characteristically opts for a pacemaker because he is suffering from symptomatic bradycardia, which may cause shortness of breath, loss of consciousness, severe lightheadedness, and fatigue.[118] These symptoms can have a substantial negative impact on the patient, and so the burdens imposed by surgery, recovery, and

[115] See Ellenbogen et al., 2000: 669-694 for a thorough treatment of potential complications of pacemaker implantation.

[116] One such risk, which is rare, is of the pacemaker eroding through thin skin and thus causing a situation where the pacemaker and leads must be removed and re-implanted. (Ellenbogen 2000: 673-4, 676)

[117] See Ganz and Hayes, 2018.

[118] See Mangrum and DiMarco 2000.

check-ups will be outweighed by the improvement in the patient's day-to-day functioning and quality of life. Moreover, these burdens, and especially the long-term burdens once the patient has healed from surgery (remote or in-person checkups every three months, replacement in 8-12 years) are comparable to and in some cases less than the burdens imposed by a normally aging heart. A pacemaker is thus clearly a biofixture worth having.

I have argued that pacemakers are biofixtures on the basis that they meet the criteria I've endorsed of responsiveness to changes in and physical integration into the patient's body. Additionally, pacemakers are clearly *biofixtures worth having* since they impose burdens comparable to the burdens imposed by the bodily function being replaced, as well as proportional to the benefit derived from the fixture. Now I will make the case that pacemakers additionally do not impose the sorts of burdens on the patient that, at least in part, motivate withdrawal of care in EOL situations. In order to support this claim, I will use as examples two common types of care that are typically withdrawn at the EOL: ventilator support and artificial nutrition and hydration (ANH).

4. Pacemakers vs. Other EOL Interventions

One of the main justifications for withdrawal of care at the EOL is the burdens imposed on the patient by such care. Simply put, if withdrawal of a treatment would make the patient less comfortable, this counts against withdrawal. And, if continuing a treatment would keep or put the patient in a state of discomfort, this counts in favor of withdrawal. These are not the only considerations in play when it comes to moral justification for withdrawal of care,

but the effect that the care is having on patients in terms of burdens and benefits is a weighty consideration. Moreover, if a treatment is substantially improving the quality of life of the patient and also sustaining the patient's life, it is clear that withdrawal cannot be justified on straightforward quality of life grounds. Rather, it seems that the primary motivation for withdrawal would be to cause the patient's death. Even if withdrawal of care were performed *by a clinician* out of respect for a patient's autonomy in accordance with an explicit request or advance directive, withdrawal of an extremely low-burden life-sustaining intervention would be the sort of thing that (assuming the patient understood what he was asking for) would be requested *by the patient* with the aim of having his life ended, because there would be no other reasonable rationale for making such a request.

Let's start by considering standard interventions which are withdrawn at the end of life in terms of the burdens they impose on patients that are alleviated when care is withdrawn. One common form of withdrawal of care at the EOL is removing a patient from ventilator support. In order to get an accurate idea of the burdens that a ventilator may impose on a patient, it is important to know exactly what being on ventilator support entails. There are both invasive and noninvasive methods of ventilator support. In cases of invasive ventilator support, the patient will either be intubated (if she is to be on ventilator support short-term) or have a tracheostomy put in (if she is to be on ventilator support longer-term). Usually an endotracheal tube is used for a maximum of 14 days, and then a tracheostomy will

be put for longer-term ventilator support.[119] Having an endotracheal tube inserted is painful, requiring sedation, and as such imposes a clear burden in quality of life terms on the patient. Further, the endotracheal tube is characteristically poorly tolerated by patients, there is a risk of dislodgement so heavy sedation is required, and the patient's communication is severely limited, both by the presence of the tube itself as well as the accompanying sedation.[120] Having a tracheostomy is less obviously a burden, once the healing process is complete. The patient is more mobile and can communicate much better than he would be able to if intubated. However, a tracheostomy still imposes burdens on the patient. Having a tracheostomy can be inconvenient in terms of speaking and can be a source of embarrassment to the patient. And there are some serious potential complications, including "misplacement or displacement of the tube, bleeding, infection, failure of the stoma to heal, and tracheal stenosis [narrowing of the trachea]."[121] Noninvasive ventilator support requires that the patient wear either a mask or a helmet and also imposes some significant burdens on the patient. Common problems associated with noninvasive ventilator support include discomfort to an extent that may require sedation in poorly tolerant patients, ulceration across the nasal bridge, unpleasant patient-ventilator dyssynchrony (where, simply put, the patient's breaths and the assistance from the mask or helmet don't 'sync up'

[119] See Shelly and Nightingale, 1999:1675.

[120] See Hasan, 2010: 305-341 for the full extent of complications from mechanical ventilation with an endotracheal tube.

[121] Shelly and Nightingale, 1999:1675.

correctly), and carbon dioxide rebreathing.[122] The burdens imposed by both invasive and noninvasive methods of ventilator support negatively impact patient well-being to an extent that may clearly warrant withdrawal of such support in many EOL cases.

Another example of EOL care which imposes substantial burdens on the patient is the administration of artificial nutrition and hydration (ANH). This is so regardless of method of delivery. Artificial nutrition and hydration can take place intravenously, through a tube inserted down the throat of the patient, or through a tube inserted directly into the stomach of the patient.[123] Intravenous nutrition and hydration runs a high risk of infection and requires surgical insertion of a port for long term care. Having a feeding tube inserted down one's throat is uncomfortable and may be painful, and is at best a temporary solution. A tube inserted directly into the stomach of the patient requires surgery, which runs a risk of infection, and leaves the patient with a stoma that is susceptible to further infection and can cause long-term discomfort. Further, "patients with advanced dementia who receive ANH through a gastrostomy tube are likely to be physically restrained and are at increased risk of aspiration pneumonia, diarrhea, gastrointestinal discomfort, and problems associated with feeding-tube removal by the patient" and "when a patient's renal function declines in the last days of life, ANH may cause choking due to increased oral and pulmonary secretions,

[122] See Esquinas, 2010: 107-117 for the full extent of complications from noninvasive pressure support ventilation.

[123] See Casarett et al., 2006.

dyspnea due to pulmonary edema [fluid in the lungs], and abdominal discomfort due to ascites [which cause swelling]."[124] These burdens on the patient, combined with the marginal to nonexistent benefit of (and sometimes active harm caused by) most artificial nutrition and hydration once the patient reaches an advanced stage of the dying process, justify foregoing or withdrawing artificial nutrition and hydration for many patients at the end of life.[125] There are no such considerations in cases of pacemaker deactivation. Pacemakers are extremely low-burden interventions; if anything, a pacemaker will improve the patient's QOL. A pacemaker will not impede a patient's ability to ambulate or communicate. It will not impose burdens on the patient in terms of pain or discomfort and has a low risk of infection once the initial healing process is complete. It is thus clear that the sort of burdens that typically drive withdrawal of particular therapies at the EOL do not apply to pacemaker therapy.

It is worth here considering whether a patient's already having the surgery for artificial nutrition via the stomach prior to being hospitalized at the end of life would change the moral status of withdrawing this care. Speaking in general terms, I believe that it would not. This is for a couple of reasons. First, even if the burdens imposed by artificial nutrition in this sort of case were minimal, there is still enough of a burden to justify withdrawal of care if the patient or his family requests it. There is a negative impact on the patient on a daily basis in terms of

[124] Casarett et al., 2006: 2608.

[125] See, e.g., Heuberger and Wong, 2018; Borasio and Jox, 2016.

discomfort and limited movement, along with continuous risk of infection.[126] Further, the fact that artificial nutrition is a usually futile intervention at the end of life makes imposing these burdens on the patient doubly unreasonable. Second, if the reason the patient received artificial nutrition were related to his terminal illness, such as dementia, debilitating stroke, or cancer, then removal of such nutrition would constitute removing an impediment to dying of that terminal illness rather than introducing or enabling a new (or otherwise well-managed and largely unrelated) pathology to end the life of the patient.

I have argued that a pacemaker is a biofixture because it is a constitutive replacement therapy that is integrated into the patient, sensitive to changes in the patient's cardiac state, and has burdens comparable to upkeep of a healthy (albeit aging) heart as well as benefits that far outweigh these burdens. Further, when compared to the burdens of other characteristic interventions that may be withdrawn or foregone at the EOL, pacemakers do not impose comparable burdens on the patient. I will now turn to argue that pacemaker deactivation at the end of a patient's life is either inconsistent with the purpose and aims of comfort care (in non-heavily dependent patients) or morally akin to VAE (in heavily dependent patients).

[126] Broadly speaking, whether burdens are substantial enough to justify withdrawal of care will have to do with proximity to death along with the patient's own perception of how heavily these burdens weigh on him. Generally, I hold that the closer the patient is to death, the less weighty burdens must be in order to justify withdrawal of care.

5. Pacemaker Deactivation and the Purpose of Comfort Care

The point at which interventions are withdrawn at the end of life is often when physicians determine that a patient should receive comfort care - that is, care in which the ultimate priority is to make the patient comfortable as he dies, even if doing so might potentially hasten the patient's death. In such situations, alleviating pain is the primary goal. However, there are two likely options regarding outcomes if a pacemaker is deactivated in the name of comfort care: either the patient will not die and feel worse as a result of losing the pacing function (in a patient who is not highly dependent upon his pacemaker, or at any rate less dependent than previously thought before deactivation) or the patient will die of an arrest shortly after having the pacemaker deactivated (in cases where the patient is highly dependent upon his pacemaker).[127] The decrease in quality of life

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[127] Freddy M. Abi-Samra explains in more detail:

> The benefits of antibradycardia pacing are threefold:
>
> 1. Preventing sudden cardiac death in patients with sinus node dysfunction (rarely) or complete heart block without any escape rhythm (pacer-dependent patients)
> 2. Preventing syncopal or near syncopal spells [loss or near-loss of consciousness] in patients who have existing but unreliable escape rhythms
> 3. Preventing general constitutional symptoms resulting from reliable but slow heart rates (fatigue, malaise, shortness of breath, etc)
>
> Patients who are completely pacer dependent make up a minority of patients receiving CIEDs. Deactivation in these patients would provide the intended result of shortening an uncomfortable dying process. Unfortunately, the consequences of deactivation in this scenario are so

in a non-highly-dependent case may be variable; a patient may feel significantly worse or only slightly worse, but regardless of the specific degree to which the patient's quality of life is lessened in particular cases, losing the pacing function will likely have a negative effect on the patient's well-being. It is important to emphasize that the reason the pacemaker was originally implanted was to correct for *symptomatic* bradycardia (slow heart rate), typical symptoms of which are shortness of breath, intermittent syncope (loss of consciousness), severe lightheadedness, and fatigue. These quality of life reducing symptoms would return upon cessation of pacemaker therapy.

When it comes to highly dependent cases, the patient will likely die shortly after deactivation, and the manner in which the patient will die is likely to be uncomfortable. The effect of cessation of pacemaker function is not to let the patient drift gently into that good night. On the contrary, the patient will likely die in distress, gasping for breath, and panicked from loss of pacing. Unless the patient is heavily sedated, this will be a deeply unpleasant death. Now, in all fairness, it is important to note that medication is often required to make patients

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> immediate that death would result in a matter of minutes, placing a great psychological burden on the provider, who must be completely at ease with the concept that his or her actions are not tantamount to assisted suicide or euthanasia.
>
> In contrast, deactivating pacing in patients whose conditions coincide with 2 or 3 above is problematic at best, cruel at worst, and in most cases would not seem to promote the goals of comfort care. (Al-Samra, 2011: 344).

comfortable in other cases of withdrawal of care at the end of life.[128] So, needing such palliation is not unique to death from loss of pacing. What is morally and substantively different about pacemaker deactivation is that, along with potentially rendering the dying process less comfortable, the underlying rationale for deactivation does not cohere with the typical, generally morally-acceptable rationale behind withdrawal of care.

Withdrawal of care in a comfort care situation is the sort of thing that ought to be undergone with the aim of facilitating a good death (or as good a death as possible) for the patient. Withdrawal of care may also be done out of respect for a patient's autonomous request. And, generally speaking, competent requests for withdrawal of care should be honored. But what I am trying to show here is that a request for pacemaker deactivation at the EOL is akin to a request for VAE in highly pacemaker dependent patients rather than a standard case of withdrawal of care. If a patient is competent to request withdrawal of care and does so, then the medical professionals involved have strong reason to honor this request out of respect for the patient's autonomy. However, in many EOL cases the patient will have deteriorated to the point that he cannot make such requests on his own. Under these circumstances (and in the absence of an advance directive or less formal knowledge of the patient's wishes), a concern for patient well-being comes to the fore and ought to serve as the primary consideration in EOL decision making. A patient's proxy consenters should make decisions which give great weight to minimizing the patient's suffering as his life draws to a close.

[128] See, e.g., Kompanje, van der Hoven, and Bakker 2008.

If the patient is suffering or simply not experiencing a quality of life worth having, then care may be foregone or withdrawn with the aim of allowing the patient to die of his underlying terminal illness. And, if the medical interventions which could sustain the patient's life would impose burdens deemed unacceptable to the patient, then this provides additional reason for withdrawing or foregoing such interventions.

As I have argued, having a pacemaker active does not in itself impose burdens on the patient in the same way that ventilator support or artificial feeding and hydration might. Often the only burden that could be reasonably attributed to a pacemaker at the end of life would be the fact that, in correcting for the patient's bradycardia, it contributes to sustaining a life that has itself become burdensome to the patient. And, perhaps, in conjunction with an autonomous request from the patient, deactivation would be justified on such grounds. Respect for patient autonomy may justify pacemaker deactivation under some circumstances. But, I suspect that a request for pacemaker deactivation would fall into one of the following two categories. Either the patient (or proxy consenter) will be confused about the details and reality of what pacemaker deactivation entails and thus not choosing based on the information required to make a genuinely informed decision in such a case, or he will be essentially requesting VAE.

I maintain that pacemaker deactivation would not constitute a standard case of withdrawal of care. If medication would be required to make a death from pacemaker deactivation comfortable (as it likely would be), and if deactivation is not of any quality of life related benefit in non-heavily-dependent patients, then

what justifiable purpose is being served by deactivation apart from causing death in heavily dependent patients?[129] And, if that is the goal, pacemaker deactivation seems to be a disingenuous and inefficient way to go about facilitating a good patient death. And, as I will now argue, deactivating the pacemaker is not consistent with an intention to allow the patient to die of his underlying terminal illness.

Are We Allowing the Patient to Die of Underlying Illness? Does this Matter?

One motivation for withdrawal of care at the EOL that pacemaker deactivation does *prima facie* cohere with is the desire to allow the patient to die from an underlying illness rather than unwantedly prolonging his life. In highly pacemaker dependent patients, deactivating the pacing function will most likely lead to the patient's death from his underlying conduction disorder. However, the terminal illness from which the patient was *actually dying* may not necessarily be at all related to the bradycardia being corrected for by the patient's pacemaker. It is important to bear in mind that pacemakers *only* correct for bradycardia. They cannot restart a stopped heart, prevent heart attacks, or correct for the underlying causes (commonly ventricular tachycardia, i.e. fast heartbeat) of the sort of

[129] It should be noted here that, while a death resulting from withdrawal of ventilator support might require medication in order to keep the patient comfortable, the relevant difference between that and pacemaker deactivation is that ventilator support characteristically imposes a substantial burden on the patient both in terms of pain and lack of mobility, whereas cardiac pacing does not.

cardiac arrest (ventricular fibrillation) that commonly kills people.[130] So, a pacemaker is not sustaining the patient's life or preventing a natural death apart from the very limited role it plays in correcting for bradycardia – a role it will have played for years in many patients.

In the absence of whatever terminal illness the patient is suffering from, he could go on to live comfortably (assuming no other QOL-reducing comorbidities are present) with the help of a pacemaker. Non-terminally-ill patients with otherwise lethal bradycardia often feel perfectly fine with the aid of a pacemaker. Further, deactivating a pacemaker in a highly dependent patient who was not dying of a different terminal illness would be correctly understood as an instance of killing that patient. So, if one wishes to justify pacemaker removal by appealing to the aim of allowing the patient to die of his underlying illness, this sort of justification will not fit the sort of situations under discussion. In these situations, in order to allow a patient who is terminally ill to die, his pacemaker is deactivated, thus removing vital treatment for *a chronic but not terminal* cardiac illness, which is ultimately what kills the patient. The patient's pacemaker is an impediment to his death by chronic and fatal-if-left-untreated bradycardia, which would be removed upon deactivation, but this does not equate to allowing the patient to die of the terminal illness that was already killing him and thus led to this EOL decision.

[130] See Snipes et al., 2014; Koplan and Stevenson, 2009.

Here I would like to call into question both the accuracy and importance of characterizing a death from pacemaker deactivation as 'dying naturally' as characterized in the HRS's principle [b]. It would make little sense to say that a patient is in fact terminally ill from cardiac disease (since the patient's cardiac disorder could indeed be fatal in the absence of treatment) when that patient had a pacemaker implanted to deal with the symptoms and underlying cause of bradycardia.[131] This is so in the same way that it would make little sense to say that a patient is terminally ill from diabetes when a regimen of insulin injections or an implanted insulin pump is managing the disease perfectly well. And it is not

[131] However, let us suppose that a patient's body is weakened by terminal illness to the point that the patient becomes heavily dependent on his pacemaker when he wasn't before the onset of his otherwise unrelated terminal illness. In reality, causation would be difficult to prove since patients generally become more pacemaker dependent as they age. Nonetheless, let us take as our example a patient where acute myocardial infarction takes out the atrioventricular node and causes that patient to become highly pacemaker dependent. This case is complicated. Would the fact that the pacemaker was implanted beforehand have any bearing on the moral status of deactivation? If a pacemaker were implanted after the onset of terminal illness for a reason related to that terminal illness, would this make it morally the same as a ventilator when it comes to withdrawal of care? I'm inclined to say that in this particular sort of case, if the patient's underlying terminal illness led to the patient becoming highly dependent on his pacemaker, then deactivation would be in accordance with the intention to allow the patient to die of his underlying illness. The fact that the pacemaker was implanted before this deterioration happened doesn't seem morally relevant to this case.

just the availability of these treatments that is relevant here, but also the role they play once integrated into the patient's life. Plenty of individuals live for many years in what seems on the surface to be perfectly adequate health while having chronic illnesses that, when well-regulated, don't actually pose a threat to that patient's life or even quality of life to any great extent. To characterize such chronic fatal-when-untreated illnesses as terminal would be disingenuous and even willfully obtuse.

It may be true in a sense that, in deactivating a patient's pacemaker at the EOL, "the clinician's intent is to discontinue the unwanted treatment and allow the patient to die naturally of the underlying disease."[132] But we ought not equivocate here between allowing a patient to die of an underlying disease which has been managed by a pacemaker, often for years, and thus in most cases need never have proven fatal, and allowing a patient to die of the effects of whatever terminal illness prompted a decision regarding withdrawal of care to begin with. The patient is at the end of his life due to a particular terminal illness that, unless it is tied to the patient's underlying bradycardia, will not be prevented from taking its course by the presence of a functioning pacemaker in the patient's body. So, while a death resulting from pacemaker deactivation may be strictly-speaking 'natural' in the same way that any death that results from bodily disfunction is 'natural', *it will not be the natural outcome of the patient's actual dying process* in the context of which the decision to deactivate was made.

[132] Lampert et al. 2010:1009

One might ask here whether this position I'm taking might have implications for more conventional instances of withdrawal of care. After all, couldn't the patient be on a ventilator for something unrelated to the underlying disease which is killing him? If so, wouldn't I then be committed to saying either that removing a patient from a ventilator under such circumstances would be akin to VAE, or that standard cases of withdrawal of care might involve allowing a patient to die of an underlying issue which was not actually killing him? There are a couple of ways to sidestep such an entailment. First, at the end of life patients are usually placed on ventilator support because their underlying terminal illness has rendered it a challenge to breathe on their own. Even if a patient's underlying terminal illness isn't explicitly respiratory in nature, bodily systems shutting down as a result of terminal illness at the end of life often necessitate ventilator support in order for the patient to go on living. This means that removing a patient from ventilator support will, in nearly all cases, amount to letting the patient die from his underlying terminal illness, even if the causal relation is a bit indirect. Second, the manner in which a pacemaker regulates an otherwise lethal illness is different from the way in which a ventilator would do so, and different in a way that might justify removing someone from ventilator support under circumstances where it would be unjustified to deactivate the patient's pacemaker. This is because, as I have argued, a pacemaker is a biofixture and thus part of the patient's body in a way that the ventilator is not.

I have argued that pacemaker deactivation at the EOL is inconsistent with the aims of comfort care. Deactivation will either lead to a decrease in the patient's quality of life or lead to the patient's death from a chronic but not terminal cardiac conduction disorder. In non-heavily-dependent patients deactivation will likely only serve to make the patient less comfortable, and in heavily dependent patients deactivation will likely directly lead to the death of the patient. However, this death is not consistent with a burdens-based justification typically given for withdrawal of care at the EOL. This is because, instead of allowing the patient to die of his underlying terminal illness, pacemaker deactivation leads to the patient's death from a chronic but not terminal cardiac illness, one that would likely be well-managed and never prove fatal were the pacemaker allowed to continue functioning. Because of this, I have argued that pacemaker deactivation is more akin to introducing a new pathology to end the life of the patient rather than removing an impediment to death from the patient's underlying terminal illness. And, as such, deactivation is inconsistent with an intention to allow the patient to die of his underlying terminal illness. While it is true that it allows a patient to die of *an* underlying illness, to say that this is a 'natural' death in the sense of facilitating or according with the death that patient is already experiencing would be disingenuous. If the patient were not already terminally ill, deactivating the pacemaker of a heavily dependent patient would be rightly seen as euthanasia or murder (depending on the intentions and circumstances involved), in the same way that refusing insulin to a diabetic or stopping a

transplanted heart through an injection would. This makes pacemaker deactivation at the EOL morally akin to VAE rather than withdrawal of care.[133]

It is possible that withdrawal of care may be justified on grounds apart from the aim to allow the patient to die of his underlying illness. For instance, withdrawal of care may be justified by appeal to the obligation produced by a patient's autonomous request for it. In such situations, is it the case that a patient's request for pacemaker deactivation should be honored? While there may be reason to honor the patient's request, I hold that to do so would nonetheless amount in moral terms to VAE rather than a standard case of withdrawal of care. This is because (as I argued earlier) deactivating a pacemaker, as it is a biofixture, would be akin to stopping a heart with an injection or removing a porcine valve rather than removing a patient from ventilator support. So, the patient's autonomous request should be taken seriously, but it should also be clear what the patient is requesting, namely a form of euthanasia.

6. Conclusion

Let us now return to the principles put forward by the HRS with which I take issue:

- [a] Ethically and legally, there are no differences between refusing CIED therapy and requesting withdrawal of CIED therapy.

[133] It is worth considering here whether refraining from giving a diabetic insulin because the diabetic has requested to have it withheld would be likewise morally akin to VAE. I maintain that it would not. This is because, while a pacemaker has the role of a biofixture in the patient's body, insulin, whether administered through a pump or injection, does not.

- [b] Ethically, CIED deactivation is neither physician-assisted suicide nor euthanasia. When carrying out a patient's request for withdrawal of a life-sustaining treatment that a patient perceives as unwanted (including CIED therapies), the clinician's intent is to discontinue the unwanted treatment and allow the patient to die naturally of the underlying disease - not to terminate the patient's life.[134]

While these may be true with regard to many CIEDS, I have called both of these principles into question as they relate to pacemakers specifically. On an understanding of pacemakers as biofixtures, there is a difference between refusing a pacemaker and requesting deactivation, in the same way that there is a difference between refusing a life-saving organ transplant and requesting that a transplanted organ be compromised by medication or removed entirely. This is because the pacemaker has become a part of the patient in a manner analogous to the way in which a transplanted organ is part of the patient. Both are constitutive replacement therapies that are bound into the patient's body and sustain the patient's life by replacing a function that would have been lost otherwise. Because of this role and integration in the patient's body, compromising the function of either would be akin to introducing a new pathology. A heart transplant may be refused and pacemaker implantation may likewise be refused by a patient. A patient may allow his native heart to fail and also may opt to allow the batteries on his pacemaker to run down and forego replacement. Both refusing a biofixture and allowing one to fail are importantly morally different from actively

[134] Lampert et al. 2010:1009

compromising the functioning of a biofixture. Correctly conceptualizing pacemakers as biofixtures works against the applicability of principle (a) to pacemakers. Further, when pacemakers are understood as biofixtures and we look at the underlying rationale behind deactivation at the EOL, deactivation of pacemakers does not just morally amount to another instance of withdrawal of care. While deactivating a pacemaker may allow the patient to die of *an* underlying disease, the disease in question is not what was actually killing the patient; the underlying rhythm disorder that would lead to a highly dependent patient's death upon deactivation is importantly unrelated to the terminal illness that actually prompted deactivation in order to allow the patient to die. Presumably, were the patient not already dying of some other illness, pacemaker deactivation would not have been considered. I have argued that, rather than being a standard case of withdrawal of care at the EOL, pacemaker deactivation in a highly dependent patient is akin to VAE. This is counter to principle (b) as it applies to pacemakers. Life-sustaining biofixtures such as pacemakers fall under the heading of treatments where discontinuation should be considered morally akin to VAE rather than standard withdrawal of care. Therefore, whatever ethical guidelines apply to VAE should apply equally to the deactivation of pacemakers.

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