III. Hutchinson-Gilford Progeria Syndrome—Questioning Assumptions of Age(ing)

People with the rapid aging disease Hutchinson-Gilford progeria syndrome (progeria), are not as visible in everyday media as centenarians are. Especially due to the low number of worldwide people living with the condition—as of January 2022, the Progeria Research Foundation (PRF) reports 132 known cases—the disease has only gained public interest during the last decade ("Meet the Kids"). Nevertheless, compared to the rarity of the disease, there is a rather high number of narratives that find their way into public media. There are newspaper articles, *YouTube* videos, social media performances, documentaries, and full length (auto)biographies, as well as a large variety of scientific publications. The scientific engagement with progeria seems to have two main aims: either to find a cure for the disease or to find a 'cure' for aging in general. While non-scientific discourses provide insights into life with the condition, the search for a cure is a strong presence within these narratives as well. Therefore, the topics of everyday life and scientific research are often intertwined in almost all progeria narratives.

Within the scientific discourse surrounding progeria, researchers ask questions about the connection between the condition and the average human aging process. Burtner and Kennedy point out that "[o]ne of the many debated topics in ageing research is whether progeroid syndromes are really accelerated forms of human ageing" (567). Progeria syndromes are thus researched in order to find out how understanding the disease can help unravel the mysteries of human aging in general. Burtner and Kennedy continue to explain that,

> [e]xciting recent findings regarding a severe human progeria, Hutchinson—Gilford progeria syndrome, have implicated molecular changes that are also linked to normal ageing, such as genome instability, telomere attrition, premature senescence and defective stem cell homeostasis in disease development." (567)

Hence, there is a similarity between the progeria body and the aging body which, medically speaking, makes progeria an interesting point of inquiry for biological research on age(ing). The theory inherent in this biological research is that, once there is a cure for progeria, this cure might also slow down the 'normal' aging process. This theory is used by the PRF to advocate for support in finding a cure for progeria. It says on the PRF homepage that "there is clearly a tremendous need for research in progeria. Finding a cure for progeria will not only help these children but may provide keys for treating millions of adults with heart disease and stroke associated with the natural aging process" ("Progeria 101"). The strategy of the foundation is thus clear: besides evoking compassion for children with progeria, the homepage also elaborates on how supporting progeria research will not only benefit people living with the disease, but also the general population. Here, the PRF ties into the discourse used by the SENS research foundation, advocating a 'cure' for aging.

When it comes to progeria, the scientific discourse of how to find a cure is always connected to the non-scientific one. Besides advocating the search for a cure and therefore dealing with the biology of the condition specifically, these non-scientific narratives also provide insight into the cultural construction of progeria as an extraordinary form of aging, as well as the social roles children living with the condition are presented in. Because awareness for the disease has emerged fairly recently, progeria, like centenarianism, used to be a cultural blank sheet that has only started being filled within the last decade.[1] Hence, the increasing number of recently emerging narratives has defining power over the way the condition itself as well as its implications about human age(ing) are presented to the world. The non-academic narratives dealing with the topic follow a limited number of individuals, as there are assumably only a very few people who are willing to make their lives with the condition public. The number, in fact, is so small, that the PRF introduces many of them on their homepage. The total of 26 children represented by the PRF, as of 2022, are introduced with short bios and are those who are more or less frequently presented in popular media ("Meet the Kids").

At first glance, texts on children with progeria appear to be fundamentally different to those on centenarians, however, looking closer reveals many commonalities between the discourses. For narratives about centenarians, the core question seems to be 'how can I become one of them,' whereas articles on progeria implicitly pose the question 'how can I help them?' A common reference in progeria discourses is, for instance, the lack of a cure. In an article for *ABC News*, Joseph Diaz writes that "there was no known treatment for progeria and no cure, leaving [families] to care for their beloved baby, whom they knew they would lose too soon." This rather emotional narrative may evoke sympathy, leaving the reader with the notion that these children and their families need help.

Although, both centenarian and progeria discourses pose different questions, they can be accused of deriving from the same interest in expanding the human life span. Both, the interest in imitating centenarians and the wish to help children with progeria, are related to a biological understanding of the aging body, asking how this biological process can be stopped or slowed down. Furthermore, narratives about centenarians, as well as about children with progeria relate to a certain amount of awe. Either, because a person has managed to live extraordinarily long or because a 'young' person lives in an extraordinary 'old' body.

Related to the search for ways to increase longevity, the direct connection between progeria and 'old' age is another common trope in discourses about the disease. Mia Graaf suggests in a *Daily Mail* article that there are new developments in

---

[1] The interest in centenarians emerged earlier, providing about two decades of narratives by now. Nonetheless, the phenomenon of centenarian narratives can still be regarded as a rather recent one.

age(ing) research owed to the efforts to find a cure for progeria. She writes that a "breakthrough came as a surprise to researchers who were investigating a cure for progeria, a genetic disorder that causes children to age rapidly and die before they reach their late teens." Graaf not only directly connects the progeria body to the aging body, she also refers to the benefits progeria research may have for the general population. Thereby she reveals a pitfall of the discourse: children with progeria are prone to be imagined as human guineapigs whose stories are only noteworthy, because they may ultimately lead to a 'cure' for aging.

Consequently, both, discourses of centenarians and children with progeria, tap into fears of the aging body and imagine people who live with it as inspiring. Both centenarian and progeria narratives connect the biological with the cultural and the social; yet, the two phenomena, represent different core assumptions. Centenarians are presented in light of an absence of disability and illness, suggesting that one can only be a star of aging when physical signs of 'old' age remain mostly absent; with progeria, age(ing) is the illness. Discussing progeria hence offers a new angle on questions concerning the way age is culturally, socially, and biologically framed in Western societies. One of the overarching questions of this study remains what constitutes a definition of 'old' age and which age subcategories are more dominant in which situations. With regard to progeria this question becomes especially interesting because age subcategories are juxtaposed to an extreme. This rare condition then offers insights into cultural constructions of age by juxtaposing its cultural, social, and biological dimensions.

Despite many differences, the connection between centenarians and children with progeria can be drawn with regard to the biological interest concerning the aging body: Both are studied in terms of their extraordinary aging process, in order to find answers for the biological aging process in general. Furthermore, all these bodies and stories are met with fascination and interest. Because they tell the story of processes that are considered to be extraordinary, the public is inclined to look. Therefore, despite all the differences, there is a point of comparison between centenarians and children with progeria as both present age(ing) in a way that challenges social roles, as well as cultural and biological imaginaries.

While the previous part focused on extreme longevity and the way people come to terms with the aging process in these extraordinary cases, this second section will deal with the way ageing is perceived when it is accelerated. What happens if one does not look back on an extremely long life but rather has to deal with the fact that life is considerably shortened? How does this altered temporality of a life influence the way the life-course is defined? What are the signifiers for age(ing) when the process denies every expectation fostered through public imaginaries? Finally: How do social, cultural, and biological norms work together in order to foster age(ing) imaginaries or even question them? Ultimately, a discussion of progeria, as an extraordinary form of age(ing) can contribute to the field of aging studies in general by making visible numerous aspects of the interplay of age subcategories in terms of culture, society and biology that work together in order to form a cultural understanding of what age(ing) is. Through this visibility, in turn, the condition allows a closer look at the contexts in which a certain category of age becomes important for the definition of age as a whole.

Progeria narratives are not only narratives of extraordinary age but also of illness. Ann Jurecic explains that what "medical humanities defines as 'illness narratives'" are "autobiographical accounts of illness spoken or written by patients" (2). Although the accounts looked at in this section are not strictly autobiographical, they are all narratives of and about people who can be defined as patients. Jurecic continues to explain that illness narratives are

> the consequence of a variety of changes in culture, medicine, media, and literacy over the past century, which include medical professionalization; the rise of modern health care; the emergence of the women's movement and the gay rights movement; the etiology of the AIDS virus; the inability of master narratives to give meaning to suffering in the modern era; and technological advances that promote self-publication and the global distribution of information. (10)

The most important factor here, I would argue, is the lack of master narratives dealing with illness. During the second half of the 20th century, society moved away from conformity and embraced individuality, a concept that also left room for imperfection. Being ill became something people could and should think about in order to try to make sense of suffering and death through writing. Although this movement, away from the norm, began decades ago, today, society still struggles with things that are outside of this norm, especially when it comes to the human body. In that connection, Elizabeth El Refaie argues that "[t]he very notion of 'normalcy' in relation to our embodied existence is problematic [as] it disregards the experiences of disabled people" (2). In other words, by propagating that the human body can have a state that is 'normal,' the experiences of those who are not considered to be part of said norm are left out of the picture and are simultaneously devalued.

Disease, according to El Refaie is one of the things that moves the human body suddenly far away from a constructed norm. She argues that "the experience of disease fundamentally unsettles our usual relationship with our bodies and in doing so changes our thought patterns" (5). Disease or disability then present a rupture in the way people regard themselves. Illness narratives, in turn, are a means to cope with this new sense of not belonging to a norm anymore and finding meaning within this new framework of life. In case of progeria, however, the children affected by the disease do not experience anything other than having a body with progeria. Here, the unsettling takes place for the parents who imagined having a child with a 'healthy' body. They then have to come to terms with the new information whereas the children themselves grow up with the experience of a deviating body. Therefore, this section will focus on the perspectives of the parents and how they come to terms with the new knowledge of having a child with progeria as well as with the perspectives of the children, who grew up with this knowledge but have to cope with the deviating body.

In that connection, it is fruitful to consider Elisabeth Kübler-Ross' "five stages of dying," or stages of grief— denial and isolation, anger, bargaining, depression, and acceptance (11). As progeria shortens the life expectancy of a person significantly, children and their families need to come to terms with the thought of a premature death. For parents, this awareness comes with the diagnosis, for the children it is established, when they are considered 'old' enough to cope with the information. Kübler-Ross' stages serve as a tool in order to navigate where the subjects of the individual narratives locate themselves in this process.

In light of the framework of illness narratives, this Part C will engage with questions about the interconnectedness of age and disease. Or more specifically, what happens when age is the actual disease? How do the narratives by children with progeria, who have lived with the condition their entire life, differ from the narratives by their loved ones?

Although this section deals with a fundamentally different form of extraordinary age(ing), mechanisms and topics such as age(ing) and gender, physical age and systems of care, or age(ing) as a phenomenon occurring within progeria narratives are very similar to those discussed with regard to centenarians. My analysis of progeria narratives will thus always be tied back to my readings of centenarian narratives in order to find similarities and differences. The aim is to reread the finding of both sets of narratives through each other and find out where an analysis of one can lead to a better understanding of the other.

Within this section, I will look at three narratives about and by children with progeria: the short documentary "Living With Progeria: Born Different," introducing three children with progeria of different ages and backgrounds, hence giving an introduction to the topic; the HBO documentary *Life According to Sam,* granting insights into the experience of one single child but also focusing on medical trials for a progeria treatment and therefore on the biological implications of the condition; and, finally, *Old Before my Time,* a full length written (auto)biography by Hailey Okines and her mother Kerry Okines, discussing the social implications of progeria for an individual as well as a family as a hole. The three examples are chosen to be somewhat analogue in terms of media format, to the examples used in part B. Thus, I am analyzing a widely seen medial example, fueling the phenomenon ("Living With Progeria: Born Different"), a documentary, presenting every-day life with an extraordinary form of age(ing) (*Life According to Sam*), and a co-written (auto)biography, giving the possibility to study a different genre and varying challenges of narrative structures (*Old Before My Time*).

The first chapter of this part will focus on the short documentary "Living with Progeria: Born Different." This section will deal with the condition as a phenomenon and the fact that popular videos, such as "Living with Progeria" tend to make a spectacle out of everything outside the norm. I will trace how children with progeria and their aging process are framed as 'different' by modern media and ask how they, themselves regard their aging process.

The second chapter, focusing on the HBO documentary *Life According to Sam* goes into detail about progeria as a medical issue. While it focuses on the biological, it narrates how the aging progeria body may affect social realms of a life and ultimately contribute to a cultural imaginary of the condition in particular and age(ing) in general. I will trace the meaning of age(ing) as a disease and the implications this narrative may have on the popular discourse around 'curing' the aging process. Furthermore, the chapter discusses age, disease, and meaning, as Sam Berns is not only known for this documentary but also for his TED talk, dealing with the recipe for being happy in life.

The final chapter of this section is the only one dealing with a full-length (auto)biography: *Old Before my Time: Hayley Okines' Live with Progeria* by Haley Okines and her mother Kerry Okines, co-written with Alison Stokes. Comparable to the discussion of Dawson's *Life is So Good!*, this chapter deals with the question of agency in co-written life narratives. Moreover, as this is the only narrative presenting a full account from birth over diagnosis into Okines' teenage years in detail, this last chapter focuses on how age in connection to illness is discussed from the very beginning of life.