

DiGeSt

Volume
4
Issue
2

Journal of Diversity
and Gender Studies

2017

**Special
issue**

Unruly Bodies: Gender \ Norms \ Resistance

Sarah Bracke, Lith Lefranc, and Anaïs Van Ertvelde (Eds.)



Reprint from DiGeSt 4.2 - Print ISSN 2593-0273 - Online ISSN 2593-0281 - © Leuven University Press, 2017

LEUVEN UNIVERSITY PRESS

2

Normalising What? About a GMO Body and Shan's Life

Nathalie Grandjean

Abstract

This paper questions the troubled links between norms, normality, and normalisation in the case of Shan, a young woman affected by Angelman syndrome and severely genetically disabled (absence of speech, disability, behavioural and sleep disorders). Firstly, it explains Shan's life, regulated by a classical approach to the normalisation of life (Nirje, 1969) and ethically guided by theories of care (Gilligan, 1982; Tronto, 1993; Kittay, 2011), leading to a conception of autonomy in high dependence. Secondly, it unpacks the controversy that occurred with the arrival of a medication that might "cure" Angelman's syndrome. Beyond analytical descriptions, the paper attempts to open the black box of Shan's body's materialities in order to show how several regulatory fictions, such as genetics and personalised care, are intertwined and correlated to different normative processes.

Keywords

body, materiality, genetically disabled, regulatory fiction, deinstitutionalisation

"(...) boundary creatures are, literally, monsters, a word that shares more than its root with the word, to demonstrate. Monsters signify." (Haraway, 1991, p.2)

The context and subject of this article is the case of Shan, a young woman affected with Angelman syndrome, who lives her disability in a deinstitutionalised way, at home, surrounded by personal assistants and "solidary" housemates.¹ Shan could

¹ Solidary here means a unity or commonality of interests, responsibilities, etc.

be described as a “genetically modified human”,² because her chromosome 15 is (partly) defective. Generally, Shan is first described by this defective gene as if it explains her identity: absence of speech, disability, behavioural and sleep disorders, etc. Her body is so out of the ordinary that it is the first thing summoned to describe it. Her genetic disability at first appears to represent her whole personality. Nevertheless, Shan’s non-standardised body tells other stories. It questions crucial ethical, political, and feminist issues: normalisation as both an integrating and stigmatising process, a possible freedom in dependence, and the possibility of envisaging Shan’s physical being beyond her bodily boundaries.

Giving an account of Shan’s “case” interests me as a feminist philosopher because of the consolidated links between feminist theory and disability theory regarding the subjection of bodies.³ As Rosemarie Garland-Thomson underlined: “Perhaps because women and the disabled are cultural signifiers for the body, their actual bodies have been subjected relentlessly to what Michel Foucault calls ‘discipline’ (1979). Together, the gender, race, ethnicity, sexuality, class, and ability systems exert tremendous social pressures to shape, regulate, and normalise subjugated bodies” (Garland-Thomson, 2002, p. 10). Both women and disabled people have been recognised as medically abnormal. They both undergo a politics of normalised appearance and a medicalisation of their subjugated bodies.

Conversely, feminist theorists like Alexa Schriempf (2001) note that even if disabled women and women share a similarly subjection of their bodies, they don’t undergo similar oppressions, due to a certain displacement in how their bodies’ identity is structured by a “regulatory ideal” (Butler, 1993) or a “regulatory fiction” (Haraway, 1991), such as sex in the case of gender identities. These regulatory ideals or fictions provide idealised and reified norms in which people are expected to live and struggle with. Thus categories are not biological facts; rather, they are created and recited through processes of performativity, expressed in discourses and gestures. Following Schriempf, it seems that disabled women are not defined principally as women but as disabled: “Michelle Fine and Adrienne Asch (1988) point

2 This is how the Shan Foundation website describes her (<http://www.fondationshan.be/syndromeangelman.html>). Strictly speaking Shan is not a genetically modified organism in the sense that her genetic heritage is not the result of deliberate genetic manipulations. If the Shan Foundation ironically names Shan as a GMO, it is to draw attention to the fact that Angelman’s syndrome is the result of a genetic accident. At the same time, calling Shan a “GMO-human” is a way to show the paradoxical contrast between “human”, designating a functional biological entity, closed and endowed with reason, and “genetically modified”, meaning that this entity can innately be genetically “modified”. This formulation is both a call to irony and a call to reconsider what a human and/or a human body is in biological discourse. I choose to keep this equivocal formulation in this paper.

3 Every Thursday, the Shan Foundation organises a team meeting with the assistants, housemates, and other people involved in Shan’s life. For the past few years, the Shan Foundation has devoted one Thursday every month to a philosophical seminar, animated by a professional philosopher. These seminars are aimed at “decolonising minds” so that the specific practices that enable Shan to live an autonomous life do not become dogmatic. The themes of the philosophical seminars are therefore always centered on the person of Shan, dealing with classical philosophical questions, such as time, love or happiness, through the specific prism of Shan, her daily life, and her future. My role as a supervisor of these seminars is simultaneously that of a professional philosopher, endeavoring to animate conversations and debates with philosophical contents, and that of a feminist philosopher, concerned with the promises of emancipation and the practices of the theories of care chosen by the Foundation, but also concerned with feminist philosophical practices through avoiding the universalisms and abstractions detached from the reality of Shan. I agree with Haraway when she expresses her need to think from the ordinary and the mud (2008, p. 3), as a plea of a radical and situated empiricism.

out that disabled women in general do not deal with the same oppressions that non-disabled women do primarily because disabled women are not seen as women in this society” (2001, pp. 53–4). It seems that for Shan’s body, the regulatory ideal/fiction is structured around her deficient gene or her particular genetic mapping and not around her sex or gender. Shan’s body and identity are in a primordial way marked by her genetic abnormality. Why do “genes” structure Shan’s body regulatory ideal, instead of sex? And how does this impact the question of Shan’s bodily materiality?

The first part of this article describes how Shan’s daily life is regulated by both a classical approach to the normalisation of life (Nirje, 1969) and ethically guided by theories of care (Gilligan, 1982; Tronto, 1993; Kittay, 2011), theories that have re-conceptualised the notion of autonomy, according to a critique of autonomy from the point of view of dependence. The second part of the article is devoted to the controversy that arose when researchers announced the arrival of a medication that could “cure” Angelman’s syndrome. The people surrounding Shan questioned the possibility of giving this medicine to Shan. Is it necessary to administer the medicine in order to ensure Shan a life as “normal” as possible? Or should we consider that Shan is happy as she is, and that not giving this medication follows the precautionary principle? And what does this imply with regard to Shan’s normalisation, normality, and “nature”?

More than proposing definitive answers, this article is an opportunity to discuss the linkages between normalisation/normalities (of body and life) and the materialities of bodies through Shan’s existence.

Shan and the Angelman Syndrome

According to the Angelman Syndrome Foundation, Angelman Syndrome (AS) is a “rare neurogenic disorder that occurs in one in 15,000 live births. Angelman syndrome is often misdiagnosed as cerebral palsy or autism due to lack of awareness. Characteristics of the disorder include developmental delay, lack of speech, seizures, and walking and balance disorders. Individuals with Angelman syndrome will require life-long care”.⁴ According to the Shan Foundation,⁵ Shan endures numerous consequences of Angelman’s syndrome:

[S]evere mental retardation, unsteady walking, easy laughter, hyperactivity, sleep disorders, and tendency to obesity. But the main disability is the absence of speech. Shan cannot speak. Her understanding of language is adequate but Shan has no access to speech. She must use other means to express herself. This is called Alternative and Augmented Communi-

4 <http://www.angelman.org/what-is-as/>; <http://cureangelman.org/understanding-angelman/causes/>
5 <http://www.fondationshan.be/syndromeangelman.html>. My translation.

cation (AAC).⁶ Shan uses all possible expressions and facial expressions. Non-verbal is her first, simple but very effective, way to communicate. Shan also uses gestures without using sign language, as the latter is too complex. Her preferred medium is the use of images that she points out to get what she desires. Shan uses several thousands of photo cards. She also has an iPad application that ‘makes images speak’ by speech synthesis.

Facing the effects of Angelman Syndrome on their daily life, Shan’s parents made the choice, very early on, to “deinstitutionalise” her. Several reasons guided this choice: first, they strongly believe that institutions should not be the only solution for a disabled person, as often is the case in Belgium. For them, institutions for the mentally disabled do not seem appropriate because the needs and desires of individuals are sacrificed for the goal of collective management; in that sense, institutions are more ghettos than homes. Shan’s parents believe in personal freedom as a key of autonomy’s fulfilment. Secondly, they feared that an institution would have difficulties in managing Shan’s behavioural disorders, and that restraint, confinement or strong medication would be considered as the solution to such troubles. Why would Shan not be able live at home in normal conditions while being disabled, they wanted to know. “Living at home, with her dog, going to the restaurant or the show, on vacation, receiving friends, going shopping. In a word, living free, among and with other citizens”.⁷

While the Shan Foundation is focused on Shan’s life and future, they consider their labour to have a collective dimension. Yet rather than promote a new model, they attribute the alternative that they are elaborating with the power to contaminate. For Shan’s parents, freedom is for everyone, with or without disabilities. They postulate that vulnerability is the normal state, which constitutes the human condition, and that everyone has the right to live in normal living conditions, even if he/she is “abnormal”. Shan’s parents are aware of the ethical issues that their decision raises, such as the questions of autonomy and freedom in high dependency contexts. They seek to engage the challenge of such questions, in order to think Shan’s life with more dignity. They are, for example, very sensible to theories of care, as these offer the possibility to think Shan as the premise of a philosophical reasoning, even if Shan herself does not speak and she is so vulnerable that she constantly needs someone to support her agency.

6 As with many children affected by Angelman Syndrome: “This investigation of children with Angelman syndrome (AS) examined reported uses of electronic augmentative and alternative communication (AAC) devices (i.e. VOs-CAs), including speech generating devices, in relation to other aided and unaided methods of communication. A total of 122 parents of children with AS, mostly from the USA, self-administered a survey over the Internet. Qualitative methods based primarily on thematic analysis were used to organise and examine data. Children relied heavily on unaided methods of communication such as natural gestures, regardless of their experiences with electronic communication devices. Parents cited various reasons for children’s acceptance or rejection of their most advanced devices.” See <https://www.ncbi.nlm.nih.gov/pubmed/23606637>

7 <http://www.fondationshan.be/avenir.html>. My translation.

A Foundation for Shan: Companionship, Prosthesis, and Theories of Care

Shan lives in her home, surrounded by a team of personal assistants who help her to live “normally”. Once Shan’s life plan was formulated in this way, her parents decided to establish a foundation, the Shan Foundation, to create a trustworthy and legal alternative to institutions. They chose a foundation because it is the only legal frame in Belgium that may be dedicated to the well-being of a single individual. The founders legitimise this position by explaining that Shan’s disability requires a personal and dedicated response to her needs, so that Shan may “become a person”.⁸

In order to fulfil these objectives, the Foundation investigates several possibilities, beginning with the experimentation of normal living conditions by an abnormal, genetically modified twenty-year-old young woman. A first response consists of the personal assistance surrounding Shan. The Foundation manages a team of personal assistants for Shan. As they state,

this very innovative approach stems from the movement for autonomous living and deinstitutionalisation in Scandinavia. Shan is accompanied in all her activities by a personal assistant. It is a form of pure companionship, without any educational or therapeutic concern. The assistant makes the gestures that Shan would have made even if she had not been disabled. She forms a real binomial with Shan. It is more about sharing life with Shan than caring. It is a highly skilled craft that requires more artistic than technical skills. The relationship of assistance is an artisanal product steeped in confidence, ethical considerations, and constant questioning.⁹

Shan therefore lives in her own home while being constantly surrounded by someone: personal assistants, solidary housemates, volunteers... The Foundation’s intention is that Shan may enjoy a personal space where her freedom can be exercised: “a home, a house with a living room, a bathroom, and a mailbox”¹⁰ – spaces as symbols of intimacy that Shan would not be able to benefit from if she lives in a mental health institution.

A second response consists of Shan sharing her house with solidary housemates. These two housemates rent a private room in Shan’s house for a reduced rent, in exchange for accompanying Shan during one night a week, and sharing some household tasks. Because of her sleep disorders, Shan often needs to be comforted if she wakes up, and sometimes she needs to be helped to wash, change, and put on new pyjamas. This solidarity agreement allows Shan to sleep safely at home.

⁸ <http://www.fondationshan.be/principe.html>. My translation.

⁹ <http://www.fondationshan.be/role.html>. My translation.

¹⁰ <http://www.fondationshan.be/philosophie.html>. My translation.

The last response resides in the use of “prostheses”. Shan uses two sorts of prosthetics, which enable her to facilitate her daily life and, as a corollary, the daily life of her assistants and relatives. We use the term “prosthesis” in a general and broad sense, defining it as a techno-scientific artefact allowing the repair or augmentation of bodily capacities seen as deficient. The prosthesis has a reversible character, in the sense that not using/wearing it implies remarkable effects in comparison with the reverse situation (analogue to wearing glasses if one is myopic). However, using a prosthesis modifies irreversibly – but heterogeneously – the image of the body as experienced and the identity representations of the user, with regard to herself and others. Shan’s first “prosthesis” is the use of images that she points to in order to get what she wants. This can be described as a “cognitive external prosthesis”. Shan uses and manages several thousand photo-cards. She also has an application on iPad that “makes images speak”, via a speech synthesizer. This allows her to “orally” address her interlocutors. The second “prosthesis” is regular medication to reduce behavioural disorders,¹¹ often socially invasive if not controlled. However, according to the Foundation, this medication is slight compared to the medication administered to people in institutions. The work of the assistants, following the normalisation principle, enables a reduction in the occurrence of these behavioural disorders. This second prosthesis is rarely qualified as such. Medication is first seen as a molecule that heals rather than repairs. In the case of Shan as in the case of other people with behavioural disorders, the question is more complex: does medication cure behavioural disorders? This would require, on the one hand, that the definition of normal behaviour is stabilised, and this is highly controversial; on the other hand, that this supposedly deficient “behavioural state” can be treated solely by molecules. The reality is infinitely more complex: what “behaviour” is, and what it represents, is maintained in a heterogeneous and moving multiplicity of meanings – social, political, psychological, identifying, and spiritual. In Shan’s case, this medication allows her to stabilise or reduce the impact of her behavioural disorders on her immediate relational environment, rapidly improving the agreeableness of her personality.

From the Normalisation of Life to the Materialities of Bodies

Normalisation as both an integrating and a stigmatising process

We emphasised that the Foundation experiments with some of the philosophical theories and debates that seem important to Shan’s well-being. One of them consists of living according to the principle of normalisation, that is, living with the most normalised daily life possible. Bengt Nirje (1969) described the principle of normalisation as follows: “making available to the mentally retarded patterns and

¹¹ The terminology “behavioural disorders” is very ambiguous. Concretely, in Shan’s case, it means aggressive behaviours, such as hair pulling, biting, hitting, pinching, pushing.

conditions of everyday life that are as close as possible to the norms and patterns of the mainstream of society” (p. 181). Concretely, according to Nirje (1994, p. 19),

Normalization means a normal rhythm of day for the retarded. It means getting out of bed and getting dressed even when you are profoundly retarded and physically disabled. It means eating under normal circumstances (...). Facilities must also give consideration to the individual’s need for a personal rhythm, allowing him to break away occasionally from the routine of the group.

The principle of normalisation also means that the choices, wishes, and desires of the mentally disabled person must be taken into consideration as accurately as possible and respected. Moreover, the prerequisite for persons with intellectual disabilities to obtain an existence as close as possible to normality is to provide them with normal incomes. The application of the principles of normalisation has profound implications not only for people with intellectual disabilities, but also for the public, for those working with people with disabilities, and for the parents of people with disabilities.

Against this model, which foregrounds liberation in the sense that it allows the deinstitutionalisation of disabled or impaired people, Ingunn Moser (2000) opposes a critical voice. In her view, the normalisation approach is constantly neutralised by processes that produce inequalities and reproduce exclusion: the main problem being the existence of a norm that places organised action, mobility, and subjectivity in a naturalised and determined human body. When measured against this norm, disabled persons will always be designated as the other, as deficient and dependent. They cannot be considered as competent and able people.

This critical stance resonates with the critique of the “social model” in disability studies. The British social model “claims that disabled people are an oppressed social group. It distinguishes between the impairments that people have, and the oppression, which they experience. And most importantly, it defines ‘disability’ as the social oppression, not the form of impairment” (Shakespeare and Watson, 2002, p. 2). What was a revolutionary vision for the disabled in society has turned into a “sacred cow” (Shakespeare and Watson, 2002, p. 5), that is to say, a strong ideology that resists being unpacked. The main critique of the “social model” revolves around the way it obscures the very materiality and diversity of disabled bodies. The absence or denial of disabled bodies brackets their very existence. Shakespeare and Watson compare this denial with the denial of sexual difference in feminist debates and women’s movements in the early 1970s. They argue that, following the feminist reclaiming of the body, one needs to understand and to take into account how bodies are interpreted through biology, but also how bodies – in their very materialities – affect lives in practice.

“Back to bodies”: interrogating materiality and normality

Let us take a first step back from these paradoxical conclusions about normalised life. On the one hand, in the beginning of our article, we deplore the fact that Shan is mainly defined by her disabled body, and on other hand, we’ve just recommended, following Moser, Shakespeare and Watson, to strive to return to the materiality of Shan’s body. This could be seen as a contradiction. Moreover, in our opinion, there’s a problem with the blind angle of the “return to the body”, because we have not bothered to open the black box of what a body is. The metaphor of the black box, often used to designate technological artefacts, is relevant to designate the body in Shan’s case. Opening the black box of the materiality of the body allows us to unfold different strata of the normativity and normality of bodies. Whose bodily materialities are we talking about?

In Shan’s case, we should hold on to a contradiction: on the one hand, her body needs to be considered in its effective impairments (such as: genetic, (in)visible, mental, physical, etc.), that is to say, in its difference. On the other hand, her body’s identity needs to be complicated by challenging the intricate sum of (biopolitical and normative) discourses, which usually define her as a disabled/GMO-human. In other words, we need to unfold *how* specific normativities, exerted on singular bodies, produce specific materialities.

In order to answer this question, we suggest following Butler’s rationale in *Bodies that Matter* (1993). Butler starts questioning what could link “the question of the materiality of the body to the performativity of gender” in order to understand why sexual difference is first invoked as material difference and not as a result of the reiteration of normativities and discourses (what Butler designates as performativity). This call for materiality/material difference is taken seriously by Butler and she suggests to go back to the Foucauldian concept of sex as a “regulatory ideal”:

‘sex’ not only functions as a norm, but is part of a regulatory practice that produces the bodies it governs, that is, whose regulatory force is made clear as a kind of productive power, the power to produce—demarcate, circulate, differentiate—the bodies it controls. Thus, ‘sex’ is a regulatory ideal whose materialization is compelled, and this materialization takes place (or fails to take place) through certain highly regulated practices. In other words, ‘sex’ is an ideal construct which is forcibly materialized through time. It is not a simple fact or static condition of a body, but a process whereby regulatory norms materialize ‘sex’ and achieve this materialization through a forcible reiteration of those norms. (Butler, 1993, p. 1)

Butler shows that “sex” as a regulatory ideal allows practices producing specific sexed/gendered bodies; and that such processes of “sexual” materialisations through bodies are never complete, always undone. Performativity, more than a discourse or a singular action, must be understood as “the reiterative and citation-

al practice by which discourse produces the effects that it names” (1993, p. 3). In Shan’s case, it seems that her body’s regulatory ideal is not “sex” but “genes”.

Shan’s body is firstly shaped by (bio)medical discourses and especially by her genetically failing body, causing a series of disabilities compared with the normal functioning of a human body. Medical discourse conceives bodies as closed units, composed of organs and functionalities; those closed entities firstly shaped and originally coded in a valid and specific genome. The “gene” performs the first materialisation of Shan’s body, as Shan’s ironic status of “GMO-human” shows.¹² In that sense, Haraway (1997) shows that there is a “gene fetishism” in our technoscientific culture that leads to consider “genes” or DNA as the origin of life itself (1997, p. 143). She also states that “gene mapping is a particular kind of spatialisation of the body” (1997, p. 141). Considering genes and DNA as the regulatory fiction that performs the genetic materialities of Shan’s body enlightens our vision. Within this regulatory ideal, genetically disabled people could be seen as in need of repairing or recovering some lost or missing abilities by directly intervening in the gene mapping. Providing gene therapy to Shan, as a cure, is exemplary of this paradigm, thus demonstrating blind trust in our gene fetishist culture.

Similarly to Butler’s rationale, if we follow Haraway, the material normality of Shan’s body is performed and shaped by a fetish, “a phantom object” (1997, p. 142). Regulatory fictions such as genes and gene maps “are ways of enclosing the commons of the body – of corporealising” (1997, p. 148). For Haraway, bodies are the “material-semiotic” products of the processes of corporealisation: “technoscientific bodies, such as the biomedical organism, are the nodes that congeal from interactions where all the actors are not human, not self-identical, not ‘us’” (1997, p. 142).

This finding allows us to consider bodies and processes materialising bodies as *interactions*, not “frozen things” (Haraway, 1997, p. 142). Those “interactions” shaping Shan’s body come from two other discursive regimes: one from a discourse on prosthetics, and another one from the outcomes of philosophical discussion.

Shan’s body is also shaped by a prosthetics discourse. It tells a very different story that positively includes new possibilities offered by modern information and communication technologies to create identities, to be an actor, and to occupy different positions as a subject.

As Moser explains, “the prosthetics discourse transfers metaphoric meanings from orthopaedic medicine and the field of rehabilitation to an entirely separate field, namely cyberculture, and puts it into action there. In this way, the rehabilitation discourse and the prosthetics discourse converge with regard to prosthetics – the idea of adding extensions and supplements to the human body and human nature” (Moser, 2000, pp. 213–4). In this paradigm, prostheses are not seen as nor-

¹² Another example of the gene as a regulatory ideal performing the materialisation of Shan’s body is the following: Shan’s parents went through a very long judicial process against the Belgian State in order to recognise Shan as genetically disabled. The Belgian social welfare wanted to recognise Shan as fully disabled since the moment the diagnostic of Angelman’s syndrome was made (at the age of 2,5 years old), while Shan’s parents wanted that recognition since the day she was born, because of the genetic origin of her condition. For them, this genetic origin means that the chromosomal deficiencies are inscribed since the first weeks of Shan’s conception.

malising artefacts, but rather as normalised, as they are the norm. In Shan's case, her body is extended by several prostheses (medication, photos, images and iPad) that expand her sphere of action and her agency. Even if the prosthetics discourse allows to enhance disabled bodies, the anthropological vision embedded within this discourse is inherited from a humanist and liberal stance, from what Bruno Latour calls "the modern constitution" (1993), that distinguishes nature and society, body, and subjectivity, and founds the idea of the independent, autonomous, rational, able, and valid human subject as the fictional and operational subject. The prostheses help to enhance this fictional subject of autonomy.

Finally, Shan's body is also formed by philosophical discussions, inspired by feminist theories of care. The Shan Foundation pleads for a vulnerable and dependent subject as an anthropological ground. Shan's daily life is led by this principle: surrounding and accompanying Shan, the personal assistants manage to exert Shan's freedom following her own agency. According to the Foundation, Shan can benefit from decisions, inevitably taken by the Foundation, but focused on her own freedom: to use her own body, to choose her own food, activities, etc. The Foundation works on the assumption that the feeling or the consciousness of freedom is dissociated from the ability to exercise it, and that an outside conscience can undertake this role. This assumption reverses the liberal subject and its bodily representation on at least three accounts: firstly, Shan possesses her body in an extended way. She extends her bodily abilities with the help of those of her personal assistant. Her ability to dispose of her body, her bodily agency, is enabled by those of each personal assistant. Secondly, it shows that high dependency situations are not only synonymous with a loss of autonomy or a lack of freedom. Extending the perspective of vulnerability and dependency as a paradigm for our very existence as beings, it demonstrates very pragmatically that the principles of autonomy and freedom as foundational features of the liberal and modern subject are merely fictional. Those principles function as fictional features, as they write stories for the fictional subject of the Modernist tradition. Thirdly, it questions the meanings of freedom, which is often considered as an individual ability and an individual fundamental right. Freedom can be exerted in an extended version of individuals, as in Shan's case or in the case of the relationship between a newborn and its caring parent. These layers of significations shape Shan's body in its very materiality. Her body is alternately genetically deficient and disabled, enhanced and extended, ruled by the "gene-regulatory-fiction" as well as the "theories-of care-fiction".

Towards a Material Normalisation?

Agilis, an (ir)reversible gene therapy

Yet another process of normalisation of Shan's body resides in the possibility to treat her body with a gene therapy in order to restore what should be its "nor-

mal” capacities. This possibility could have remained a mere fantasy about which to speculate, but in reality researchers are about to propose a gene therapy to treat Angelman’s syndrome, by administering daily medication, Agilis Biotherapeutics, LLC. As Greg Robinson and Jodi Cook point out, “This is gene therapy. A virus (adeno-associated virus = AAV) is used to carry the UBE3A into the nerve cells (neuron) of the brain and implant it into the nucleus. There it can make the protein UBE3A (E6-AP ubiquitin ligase) that is coded by UBE3A and is missing in those individuals with Angelman Syndrome (AS). This intrinsic, therapeutic procedure may be used for the treatment of the brain itself, or directly in the brain itself.”¹³

This promising medicine raises new issues, with respect to both the choice of a life led according to the principle of normalisation as well as the abnormal/deficient genetic nature of Shan’s body. The announcement of the possible administration of Agilis to Shan challenged the Foundation members and assistants: Should we give it or not give it? If we give it, is it for the good of Shan, changing her or transforming her? Is this the good we want for Shan?”. Moreover, they raise the fact that they already transform her (being mandated for that), by giving her daily medications to regulate her moods and behavioural disorders. The bioethical questions of consent, freedom, and autonomy are indeed central issues during the Foundation’s team meetings.¹⁴

But this time the impact would obviously be of a different kind. Agilis is a gene therapy, not a psychotropic. It is therefore necessary to distinguish it from Shan’s other prostheses already described above. Indeed, some authors (Folscheid, 2006) usefully distinguish internal and external aspects among prostheses. While the iPad and Shan’s photo database would be considered external prostheses, her medication would be considered an internal prosthesis, that is to say, more like an implant. The same goes for gene therapies, as promised by the Agilis molecule, which would be seen as a genetic implant. Gene therapy transforms the “substantial interiority” of the body. There is no longer any possible or predictable reversibility.¹⁵

13 <http://cureangelman.org/gene-therapy-for-the-treatment-of-angelman-syndrome-future-clinical-trials/>

14 Agilis raises typical bioethical issues, those of consent and autonomy. The values and principles that usually govern ethical thinking, such as consent and autonomy, are no longer appropriate or operational. As we have underlined, Shan cannot speak and thus is not recognised as a person of legal age. Legally and practically, she cannot consent, in any case – and her autonomy is very restricted, in the sense that she needs to be surrounded by her personal assistants to live a “normal” life. Moreover, she is not conscious of her a-normality; she lives in the innocence of her condition of mental deficiency. The question of consent in Shan’s case must therefore be enlarged, and a broadened consent should be considered. Extensively, it raises the question of the responsibility of those who make the decision for Shan. In reality, people surrounding Shan (the Foundation, the personal assistants) are already doing so. But administering a medicine for mood disorders is not comparable to giving a drug that would completely change Shan. It is a more invading decision, attacking Shan’s very identity. In the end, the main issue is not that of consent, but that of responsibility – response-ability – and responsivity engaged in the potential collective decision. Responsivity is an matter of relationality, not identity. As Oliver noted, we need to change our ethical standpoint and leave the ethics of identity and difference: “We can talk about both identity and difference without examining the relationship between them. What we need is to move from an ethics of sameness, through an ethics of difference, towards an ethics of relationality and responsivity.” (2010, p. 270)

15 The reversibility of Agilis is not clear: if we assume the fact that Agilis will genetically treat Shan, what would happen if the treatment is stopped? Will Shan’s body go back to its “normal” Angelman status? At present it is impossible to predict.

Normal or better?

This plausible possibility of genetically curing Shan also questions the linkage between normalisation and the materiality of the body, but in another way. It starts from the very materiality of Shan's body and supposes that a "normalised" flesh/DNA would lead to a normalised life, as if Agilis would repair the "natural" processes of the creation of Life itself (starting from a valid and normal genome to a valid and autonomous body, and then a "normal" life).

While discussing and debating, the people of the Shan Foundation and the personal assistants underline that they were struck by the social evidence in favour of giving Agilis to Shan. They feel socially pushed by the obligation of curing Shan so that she would have a normal body or a *better* body. The medical discourse does not convince nor reassure them: "Shan is not an Angelman mouse... we are not sure of the consequences." Let's suppose that Shan regularly takes Agilis, and that it renders her "normal". Several criteria should be assessed: first, the reversibility of Agilis. That is to say, if Shan stops taking Agilis, does she revert to her previous identity/status? Second, the unpredictability of the effects on Shan's body. Third, Shan's identity. What about her past, her memory? How could she confront her past as a disabled and speechless person? She risks to be deeply traumatised by the Agilis transformation of her identity. The risk of suffering might be higher than if Shan keeps living as she currently does – she risks to be reflexive, to be conscious of the world as it is. This consciousness frightens the people of the Foundation in a way that one might be frightened of leaving behind the Garden of Eden. Taking the risk of a normal(ised) body does not imply the insurance of a better body.

We retain from this iteration the possibility – even the freedom – of not giving Agilis to Shan. More than a spontaneous protection of Shan, this discussion shows two major points: 1) thinking from the point of view of vulnerability allows us to think vulnerability *not only* as a premise, but as a normal condition of existence; and 2) opening the black box of the materialities of Shan's body shows that several regulatory functions can be consciously intertwined; this possibility opens a way for "defrozen" identities.

References

- Butler, J. (1990). *Gender Trouble: Feminism and the Subversion of Identity*. New York, NY: Routledge.
- (1993). *Bodies that matter. On the discursive limits of "sex"*. New York, NY: Routledge.
- Calculator, S. N. (2013). Use and acceptance of AAC systems by children with Angelman syndrome. *Journal of Applied Research in Intellectual Disabilities*, 26(6), 557–567.
- Folscheid, D. (2006). Un implant paradigmatique. In: *Ethique et implant cochléaire. Que faut-il réparer?* J. Griot and L. Meurant (éds.). Namur: Presses Universitaires de Namur. Transhumances VII.
- Fraisse, G. (2007). *Du consentement*. Paris: Seuil.

- Garland-Thomson, R. (2002). Integrating Disability, Transforming Feminist Theory, *NWSA Journal*, 14(3), 1-32.
- Gilligan, C. (1982). *In a Different Voice: Psychological Theory and Women's Development*. Cambridge, MA: Harvard University Press.
- Haraway, D. J. (1991). *Simians, Cyborgs, and Women: the reinvention of nature*. New York, NY: Routledge.
- (1997). *Modest_Witness@Second_Millennium.FemaleMan©Meets_OncoMouse™: Feminism and Technoscience*. New York, NY: Routledge.
- (2008). *When Species Meet*. Minneapolis, MN: University of Minnesota Press.
- Hartssock, N. (2004). The Feminist Standpoint: Developing the Ground for a Specifically Feminist Historical Materialism, in S. Harding, (ed.), *The Feminist Standpoint Theory Reader: Intellectual and Political Controversies*. New York, NY: Routledge.
- Kittay, E. F. (2011). The Ethics of Care, Dependence, and Disability, *Ratio Juris*, 24, 49-58.
- MacKinnon, C. (1997). Rape: On Coercion and Consent. In K. Conboy, N. Medina & S. Stanbury. (eds), *Writing on the Body: Female Embodiment and Feminist Theory*. New York, NY: Columbia University Press, 42-58.
- McGuinness, K. (1993). Review: Gene Sharp's Theory of Power: A Feminist Critique of Consent. *Journal of Peace Research*, 30(1), 101-115.
- Moser, I. (2000). Against Normalisation: Subverting Norms of Ability and Disability, *Science as Culture*, 9(2), 201-240.
- Nirje, B. (1969). The normalization principle and its human management implications. In R. Kugel, & W. Wolfensberger (Eds.), *Changing patterns in residential services for the mentally retarded*. Washington, D.C.: President's Committee on Mental Retardation.
- (1994). The Normalization Principle and Its Human Management Implications, *SRV-VRS: The International Social Role Valorization Journal*, 1(2), 19-23.
- Oliver, K. (2010). Animal Ethics: Towards an Ethics of Responsiveness, *Research in Phenomenology*, 40, 267-280.
- Schriempf, A. (2001). "(Re)fusing the Amputated Body: An Interactionist Bridge for Feminism and Disability", in *Hypatia*, 16(4), 53-79.
- Seville, C. (2009). *The EU Intellectual Property Law and Policy*. Cheltenham & Northampton: Edwards Elgar.
- Shakespeare, T. and Watson, N. (2002). The social model of disability: An outdated ideology? In *Research in Social Science and Disability*, 2, 9-28.
- Shildrick, M. (2015). "Why Should Our Bodies End at the Skin?": Embodiment, Boundaries, and Somatechnics, *Hypatia*, 30(1), 13-29.
- Tronto, J. (1993). *Moral Boundaries. A Political Argument for an Ethics of Care*. New York, NY: Routledge.

About the author

Nathalie Grandjean holds a PhD in Philosophy and is a Lecturer in Ethics and Feminist Philosophy (University of Namur). Her research interests include the body and technology, gender and feminist studies, surveillance, STS, French Theory, and post-structuralism. She co-edited *Corps et Technologies. Penser l'hybridité* (with Claire Lobet, Peter Lang, 2012) and *Valeurs de l'attention* (with Alain Loute, Presses du Septentrion, 2017 – forthcoming).