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\*Corresponding author: Marie Cholley-Gomez, ACTES Laboratory. "Adaptations to Tropical Climate, Exercise and Health" (EA N°3596), Université des Antilles, Campus de Fouillole, Pointe-à-Pitre, BP - 250 97157, 97157, Guadeloupe  
E-mail: [marie.cholleygomez@hotmail.fr](mailto:marie.cholleygomez@hotmail.fr)

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Sandro Serpa, Sociology, University of the Azores, Ponta Delgada, Portugal

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## SOCIOLOGY | RESEARCH ARTICLE

# Ecological approach to sickle cell disease and environmental quality assessment in Guadeloupe (French West Indies)

Marie Cholley-Gomez<sup>1\*</sup>, Sébastien Ruffié<sup>1</sup>, Benoit Tressières<sup>2</sup>, Gaël Villoing<sup>1</sup> and Sylvain Ferez<sup>3</sup>

**Abstract:** Sickle Cell Disease (SCD) has many psychosocial impacts and affects social participation of young people. In Guadeloupe (French West Indies), where this blood disease is a major concern, young patients face a doubly constraining environment, on both social and physical aspects. Based on the *Disability Creation Process* (DCP), an ecological model of disability and health, this controlled study aimed to assess the perceived influence of social and physical factors. The study involves 80 young patients (Mage =  $11.25 \pm 3.23$ , 36 females, 44 males) with severe Sickle Cell Disease (Major Syndrome or S $\beta$ 0-thalassemia) and 45 healthy children and adolescents (Mage =  $10.55 \pm 2.93$  years; 22 females, 23 males). The *Measure of the Quality of the Environment* (MQE) was used, after adaptation of the scale to the socio-cultural context. Results show that environmental factors are more negatively perceived among SCD participants, physical factors constituting strong barriers, and social determinants being weaker facilitators. In the micro and meso-environment, SCD patients face more difficulty; finally, findings suggest that more inclusive spaces are needed in school settings, sports and leisure activities, as well as in the cultural offer.

**Subjects:** Disability Studies - Sociology; Public Health - Medical Sociology; Sociology of Health and Illness

**Keywords:** Sickle Cell Disease; environment; disability; youth; Caribbean studies

## 1. Introduction: conceptualization of terms

### 1.1. An ecologic-systemic explanatory model of diseases: the *Disability Creation Process* (DCP)

Understanding chronic diseases and disability situations, their production mechanisms and their consequences from a social and inclusive perspective remains a crucial challenge. Successive theoretical approaches have been proposed since the 1950s, and can be conceptualized in five major models: biomedical, economic, social, embodied, and lastly, human rights based (Fougeyrollas et al., 2019). Although a consensus has been reached in the international community stating that disability is the product of social and environmental practices, a strong conceptual and operational framework is still needed (*Ibid.*).

The Disability Creation Process model (DCP, Fougeyrollas et al., 1998, ; Fougeyrollas et al., 2019) has been a powerful tool in providing conceptual and operational definitions of participation -and disability situations. Used in a broader range of contexts, and on a practical level, it has also helped in the development of policies and guided service delivery (Boucher et al., 2018; Fougeyrollas &

Grenier, 2018). This model, based on the anthropological model of human development and disability, and on the human rights paradigm, explains the causes and consequences of disease, trauma, or other damage to the individual's integrity or development. It states that the disability process cannot be grasped without considering the dynamic and interactive process between personal (intrinsic) factors and environmental (extrinsic) factors. This process determines the situational result of social participation, operationalized as the degree to which life habits are performed, on a continuum going from disability situation (participation restriction) to optimal (full) participation. These life habits—current or daily activities and social roles—are related to the individual's age, gender, and sociocultural identity (Fougeyrollas & Charrier, 2013).

In the DCP, environmental components have a direct impact on, and are incorporated as a determinant of, social participation, similarly to intrinsic factors. Based on Bronfenbrenner's ecological theory (Bronfenbrenner, 1979, 1994) this conceptual field is divided in three special levels, from *micro* (intimate analysis of the immediate living environment), to *meso* (local or community analysis), and then *macro* (analysis of society as a whole) level. This spatial perspective is relevant for the modulation of intervention strategies (Edwards et al., 2014; Noreau et al., 2015).

Environmental components refer to social or physical dimensions that determine a society's organization and context in relation to the individual. Therefore, this classification is appropriate for every human, regardless of their level of disability, in different geographical or cultural contexts. In interaction with personal determinants, when it supports the accomplishment of life habits and provides optimal participation, an environmental factor is considered as a *facilitator*; in contrast, when contributing to disability situations or limiting the fulfillment of life habits, it is an *obstacle*.

### **1.2. Understanding Sickle Cell Disease: interaction between intrinsic and environmental factors**

Sickle Cell Disease (SCD) is an hemoglobinopathy that belongs to a group of inherited autosomal recessive disorders, characterized by the production of abnormal hemoglobin (Bonner et al., 2010). SCD is the most widespread genetic disorder in the world (Wastnedge et al., 2018), and in Guadeloupe (*French West Indies*, Saint-Martin et al., 2013) where it affects approximately 1 in every 300 newborns. Its prevalence warrants a systematic neonatal screening throughout this territory (HAS, 2010). The severity of symptoms alters daily life and has led to its recognition as a disability, in the broader category of “disabling diseases” (Lespinet-Najib & Belio, 2013).

SCD places a significant emotional and physical burden on patients (Osunkwo et al., 2021). To experience this disease is, first and foremost, to experience chronic pain (Taylor et al., 2010; Wilkie et al., 2010), with unpredictable, repeated and painful crises that lead to the interruption of everyday activities, with sometimes very long stays in the hospital (Panepinto et al., 2005). As SCD is a genetic condition present from birth, this disease is likely to interact at an early age with developmental and personal factors, which have implications for cognitive and psychosocial functioning (Bonner et al., 2010; Prussien et al., 2020. Tarazi et al., 2007). The disorder thus affects mental health and quality of life of young patients, as shown by the prevalence of depressive syndromes and anxiety disorders within this population (Jerrell et al., 2011; Moody et al., 2019), and more generally, a lower quality of life (Graves et al., 2016; Houwing et al., 2022).

The metabolic and physiological SCD complications (anemia, vaso-occlusive crises), and impairments (in motricity due to osteonecrosis of the femoral head), in relation to the psychological dimensions (stress adaptation strategies), play a role in the limitations of children's participation, and must be considered in connection with the individual's environment.

A complex, social approach to this disease is thus necessary as it has historically been investigated more from a biomedical perspective, with political or social considerations in studies on hemoglobinopathies being more recent (Dyson & Atkin, 2012; Lainé & Tchernia, 2010). The

theoretical and methodological anchoring of our study to a systemic and holistic approach such as the DCP model is thus relevant, as highlighted by our previous work (Ruffié et al., 2021; Cholley-Gomez et al., 2021).

Young SCD patients must face a doubly constraining environment, on both social and physical aspects. From an early age, medical recommendations are given to SCD children and their family, due to their vulnerability to, among various things, temperature variations, infectious risks, or physical effort (HAS, *ibid.*; American Society of Hematology—ASH, 2016; Tanabe et al., 2019), where environment plays a crucial role. In a daily life requiring constant monitoring, young sicklers face many activity restrictions. Limitations particularly concern sports and leisure activities, which have a strong social and cultural dimension, especially in the French Caribbean context (Pruneau et al., 2008). Analyzing the impact of SCD in a controlled study using the Life-H scale (life habits), we (Cholley-Gomez et al., 2021), highlighted a lower social participation score in young SCD Guadeloupians compared to controls. The accomplishment of daily activities and social roles was thus more impeded, in particular for leisure activities, community life, schooling and mobility.

Devoid of prospect of recovery, chronic illness involves, contrary to transitional acute illness, a constant management. It is not a rare life episode but a permanent state that merges with the life itself (Baszanger, 1986; Pierret, 1997). This feature—being a long-term or permanent component in an individual's life—invites a sociological analysis of this experience, with emphasis on its time-frame (Bury, 1982, 1991), from the “biographical disruption” caused by the announcement of the diagnosis (Bury, 1982; Corbin & Strauss, 1987). Sociological research has shown the impact of such a turmoil on the patient's experience and on his social interactions (Strauss & Glaser, 1975), according to the interactionist concept of “career”. It is thus possible to identify an “illness trajectory” characterized by organization of work, involving biographical and identity reorganizations—developed since the diagnostic, and then throughout the evolution of illness, to control it. In the case of SCD, the disruption is less experienced by the young sicklers, “born with illness” and diagnosed at birth, than by their parents, who face significant changes of their daily lives.

Negative social treatment of SCD also provides information on how patients face their social environment. Young SCD patients are caught between a familial micro-environment, which is protective, and negative social treatment of the disease. Works in the field of sociology and using an interactional approach offer a glimpse of the way in which SCD is experienced from a stigmatizing and exclusion angle, or of the self-exclusion from social spaces, by considering that the stigma is, within an interaction, what affects, by discrediting it, the social identity of an individual (Goffman, 1963).

Indeed, SCD is a stigmatized pathology (Bougerol, 1994; Bulgin et al., 2018; Lainé, 2009). Its categorization as “Blood disease”, by questioning heredity, conveys complex identity and symbolic meanings (Bonnet, 2001, 2009; Bonniol & Benoist, 1994). In the French Caribbean culture, traditional elements thus influence the social representation of this pathology, leading to a weak social visibility of it. In a previous work, Pruneau et al. (2009) showed that the content of the social representation of SCD in young Guadeloupians was far from the biomedical definition, and underlined by social and identity issues. The notions of exclusion and contagion are part of a complex perception of the disease, which carries contrasted elements and is the result of a conflict between bad “reception” and bad “perception” (Ibid). Stigmatization thus operates at the crossroads of social and identity-related processes, orienting interaction postures just as much as avoidance strategies.

As they face an asymmetric interaction (Goffman, 1963) young patients may suffer from exclusion from social spaces-sometimes leading to negative self-perception. Therefore, SCD children grow up “with and despite the disease” by implementing strategies depending on social context (Atkin & Ahmad, 2001; Ciribassi & Patil, 2016). Family environment appears to be resilient, protective, even overprotective (Atkin & Ahmad, 2000; Bonnet, 2004, 2009; Reader et al., 2020;).

In contrast, outside of the family unit, interpersonal relationships are more difficult and, sometimes ignoring recommendations, young patients with SCD try to be “secret keepers” (Héjoaka, 2012). This is particularly the case for physical and sport activities, where the challenge is “to restore a positive relationship with a bruised body” (Pruneau et al., 2008, p. 16). At school, they have a paradoxical desire for recognition, adjacent to a wish of normalization and “normal sociability” (*Ibid.*) -and often refuse to disclose their illness (Bibrac & Etienne-Julan, 2004).

## 2. Issue

Young children and teenagers with SCD thus evolve in a diversity of environments and various socializing contexts, which constitute barriers, or on the contrary, improve their social participation. These various settings—micro, family environment; meso, school and community environment; and macro-environment, at the level of society—in which they live with their functional limits or their particular capacities, shape the consciousness of their singularity.

What contextual determinants, social and physical factors of their environment, respectively, limit, and improve the accomplishment of their daily habits? How do youths with SCD perceive the Guadeloupean context, compared to their healthy peers?

## 3. Method

### 3.1. Study design and population

The study took place at the Sickle Cell Disease Transversal Unit (UTD<sup>1</sup>). The initial group of subjects included young patients (n = 102, from 6 to 16 years old, 49 females, 53 males), who suffered from severe forms of SCD (i.e., Major Sickle Cell Syndrome or S $\beta$ 0-thalassemia<sup>2</sup>) and were monitored at the UTD. Other inclusion criteria for the study were the following: having been diagnosed during the neo-natal or natal period and having provided a declaration of non-opposition to the study (also signed by their legal representative). Out of the total cohort of 102 patients, our sample included 80 participants (Mage = 11.25  $\pm$  3.23, 36 females, 44 males).

A control group, composed of healthy children (n = 45, age range = 6–16 years old, Mage = 10.55  $\pm$  2.93, 22 females, 23 males) who were randomly selected and did not present any chronic disease, was also integrated into the study. The aim was to neutralize any possible effect linked to the context, as no study using the DCP model and evaluating social participation, or the perception of social and physical environmental factors, has been conducted within the specific context of Guadeloupe.

### 3.2. Assessment tool

Environmental factors were estimated with the *Measure of the Quality of the Environment* (*Mesure de la Qualité de l'Environnement*, Fougeyrollas et al., 1999, 2008), using prior adjustments of items to the population and the socio-cultural context (details of this adaptation of the tool are presented in our work, Cholley-Gomez et al., 2020). Based on the DCP conceptual framework, the MQE assesses the influence, or the perceived quality, of environmental factors divided into two sub-categories: *social* and *physical* factors (Fougeyrollas, 2010). These factors are grouped into areas/domains of the environment: politico-economics and socio-cultural for the social factors, and nature and land-use planning for the physical ones. Their influence is assessed on a 7-point scale ranging from *major facilitator* (+3) to *major or complete obstacle* (–3).

For further analyses, in addition to the score calculation, each answer/factor can be coded into three categories: obstacle, facilitator or no influence (Fougeyrollas, 2010). Table 1 details, for each score on the assessment scale, its appreciation, and the corresponding definition.

Due to the complexity of some items (e.g., concerning political systems, social-health services), assessment sometimes required that a parent or a relative be present with the child, in case they were too young to fully understand the statement.

**Table 1.** Detailed presentation of the Measure of the Quality of the Environment (*MQE scale, adapted from Fougeyrollas, 2010*)

Scale point	Appreciation	Definition
-3	Major obstacle	Completely prevents the accomplishment of the life habit
-2	Moderate obstacle	Substantially interferes with the accomplishment of the life habit
-1	Minor obstacle	Interferes with the accomplishment of the life habit, or just slightly increases the difficulty
0	No influence	Situations or environmental factors having no effect on the performance of an activity
1	Minor facilitator	Makes up for deficiencies or disabilities and allows for a partial accomplishment of the habit, or only slightly reduces difficulty
2	Moderate facilitator	Moderately compensates deficiencies or disabilities, or allows to partially realize the habit, or with difficulty
3	Major facilitator	Compensates deficiencies or disabilities, and allows to fully accomplish the life habit without constraint or difficulty
N	Unknown	The effect of situations or environmental factors on an individual's life is unknown
P	Does not apply	Situations or environmental factors that do not exist in the context

Additional socio-demographic data included: place of residence (recoded into 3 modalities: *Grande-Terre, Basse-Terre, Central Zone*<sup>3</sup>), parents' occupation (recoded into 3 modalities: *Inactive, Worker-Employee, Intermediary-Upper level*; *Unknown-Deceased* was also available but was not included in the analysis, where the data was considered as [missing data]) and their marital situation (recoded into 2 modalities depending on the family-structure of the child's household: *single parent* vs. *parents together*, including new union situations), along with the number of siblings (including those from the previous or current union of each of the parents, recoded into 2 modalities: *only child* vs. *presence of sibling(s)*).

The main setting of the study was the UTD, within the University Hospital Centre (CHU) of Pointe-à-Pitre (n = 75) or, for patients who came from the Basse-Terre area, and depending on another hospital structure, the Basse-Terre Hospital Centre, CHBT (n = 5). For the control group, data was collected at convenient places for the subjects, mainly at home. Data was entirely collected using an eCRF.<sup>4</sup>

#### Analysis

Statistical analyses were conducted using Statistica and R 3.6.1 software. Discrete and continuous variables were compared using Khi2 test and Student's t-test (for two groups) or ANOVA (for more than 2 groups) respectively. Normality of distribution was assessed with Kolmogorov-Smirnov tests; group sizes allowed us to perform parametric tests (Student's t-test, ANOVA). Concerning the ANOVA, assumption of homogeneity of variance was examined with Levene's test.



## 4. Results

### 4.1. Participant data

The sociodemographic characteristics of participants are presented and compared in Table 2. Characteristics of the participants were described using means and standard deviations or frequencies and percentages according to the type of variable (continuous or categorical, respectively) and compared across groups.

### 4.2. MQE data analysis

Independent Sample t-test results (i.e., between SCD group, n = 80 and Control Group (CG, n = 45) are reported below, in Table 3. Mean scores ( $\pm$ Standard Deviation, SD) are presented for each contextual factor assessed by the MQE (n = 11 categories), including the *physical environment* subscore (comprising the following categories: traffic lanes, natural elements), the *social environment* subscore (social network, attitudes of entourage, sources of income, socio-health services, education services, public infrastructures, community-based organizations, social rules and political systems), and lastly, the *overall environment* score -i.e., the general perceived quality of the context.

Analyses showed a strong effect of the group variable on main outcomes: firstly, for global environment, i.e., the general perceived quality of the context, with a lower score in the SCD group compared to controls. Environmental factors were positively perceived by both, but lower in the SCD group. Physical environment was negatively rated by SCD participants, while it was positively assessed by controls, a statistically significant difference. Social factors were positively perceived by both groups, but the assessment score was significantly lower within the SCD group. Among the eleven categories composing the MQE, most of them were more negatively rated by SCD children than by their healthy peers (for the positive scores, perceived as being a weaker facilitator,

**Table 2. Comparison of participant characteristics between SCD group and control group**

	SCD Group N = 80	Control Group N = 45	p
<i>Continuous variable</i>	<i>Mean <math>\pm</math> standard deviation</i>	<i>Mean <math>\pm</math> standard deviation</i>	
<b>Age (in years)</b>	11,25 $\pm$ 3,21	10,55 $\pm$ 2,93	0,241
<i>Category variables</i>	<i>n (%)</i>	<i>n (%)</i>	
<b>Gender</b>			0,729
Female	36 (45%)	22 (48,9%)	
Male	44 (55%)	23 (51,1%)	
<b>Marital situation of the parents</b>			<0,001
Together	24 (30,7%)	32 (71,1%)	
Single parent	54 (69,3%)	13 (28,9%)	
Missing	2		
<b>Siblings</b>			0,005
Single child	29 (37,1%)	6 (13,3%)	
Siblings	49 (62,9%)	39 (86,7%)	
Missing	2		
<b>Occupation of parents</b>			<0,001
Intermediary-superior	17 (21,8%)	26 (57,8%)	
Worker-employee	32 (41,2%)	15 (33,3%)	
Inactive	29 (37%)	4 (8,9%)	
Missing	2		



**Table 3. Mean scores (SD) of MQE factors according to group and T-Student results (p value)**

	<b>SCD Group</b>	<b>Control Group</b>	<b>p</b>
MQE	<i>Mean ± standard deviation</i>	<i>Mean ± standard deviation</i>	
Social network	1,15 ± 0,93	2,01 ± 0,62	<0,001
Attitude of relatives	0,93 ± 0,72	1,37 ± 0,80	0,002
Sources of income	0,93 ± 1,41	2,00 ± 1,31	<0,001
Socio-health services	1,57 ± 0,78	1,65 ± 0,94	0,616
Educational services	0,45 ± 0,87	1,44 ± 0,71	<0,001
Public infrastructures	0,48 ± 0,67	1,64 ± 0,87	<0,001
Community organisms	0,72 ± 0,77	1,76 ± 1,15	<0,001
Traffic lanes	0,18 ± 1,08	1,09 ± 1,45	<0,001
Natural elements	-1,26 ± 0,67	0,43 ± 0,78	<0,001
Political systems	-0,05 ± 0,97	-0,02 ± 1,22	0,910
Social rules	-0,17 ± 1,08	0,62 ± 1,33	<0,001
<b>Social Environment</b>	<b>0,69 ± 0,48</b>	<b>1,52 ± 0,65</b>	<b>&lt;0,001</b>
<b>Physical Environment</b>	<b>-0,54 ± 0,67</b>	<b>0,76 ± 0,79</b>	<b>&lt;0,001</b>
<b>Overall Environment</b>	<b>0,46 ± 0,43</b>	<b>1,37 ± 0,61</b>	<b>&lt;0,001</b>

and for the negative ones, as being a stronger obstacle). There was an exception, however, for *socio-health services* and *Political systems*: indeed, results indicated a positive rating, and a lower negative assessment (i.e., close to a lack of influence), respectively, with scores being similar for both groups ( $p = .61$ , and  $p = .91$ , respectively).

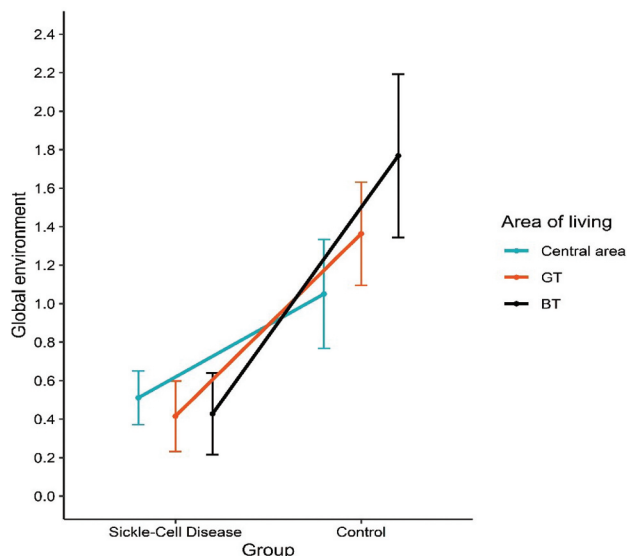
Other independent variables (socio-demographic data) were also statistically analyzed. Among them, gender had no influence on the quality of global environment ( $p = .51$ ), boys and girls having similar scores ( $M = 0.82 \pm 1.06$  vs  $M = 8.21 \pm 1.33$ ). It did not either have any impact on the perception of social ( $p = .32$ ) or physical ( $p = .47$ ) components.

To briefly summarize family configurations, as these specific analyses were detailed in a parallel work (Cholley-Gomez et al., 2021), results showed that marital situation did not have an impact on the perception of overall, physical, or social environments for the SCD Group ( $p = 0.4$ ,  $p = 0.1$ , and  $p = 0.7$ , respectively). It did, however, have an effect on the perception of overall environment, as well as on the social environment sub-category for controls (both for  $p < .05$ ). Presence vs absence of siblings did not make any significant difference within the SCD Group concerning the overall, physical or social environment ( $p = 0.3$ ,  $p = 0.9$ , and  $p = 0.3$ , respectively). The occupational category of the parents did impact the perception of environment. Indeed, the global environment score, as well as the social sub-category, were both significantly lower for SCD subjects with inactive parents, in comparison to those with parents who were workers or employees ( $p = 0.01$ ,  $p < .01$ ).

Contrary to expectation, the analysis of variance did not allow to conclude that the area of residence had a main effect on global environment perception, nor on the social or physical subcategories ( $p = 0.16$ ,  $p = 0.19$ , and  $p = 0.15$ , respectively), scores between Basse-Terre (BT), Grande-Terre (GT) and the central area of the island being equivalent.

However, the interaction term of the ANOVA 3 (area of living)-by-2 (group) was significant when assessing overall environment ( $F(2, 119) = 5.33$ ,  $p < .01$ ). As shown in following graphical representation (Figure 1), among the SCD group, all participants, regardless of their area of living, had a similar perception. Among the control group, residents of BT had a more positive perception

**Figure 1. Score of global environment quality depending on group and area of living.**



of their global context ( $M = 1.75 \pm 0.15$ ) than residents of GT ( $M = 1.36 \pm 0.10$ ) and of the central area ( $M = 1.05 \pm 0.14$ ), the difference between these two last subgroups being significant.

The interaction term was also significant on *physical* ( $F(2, 119) = 7.75, p < .001$ ) and, more moderately, on *social* ( $F(2, 119) = 3.54, p < .05$ ) factors. In CG, concerning *physical factors*, BT score ( $M = 1.17 \pm 0.21$ ) was significantly higher than those from GT ( $M = 0.88 \pm 0.14$ ) and central area ( $M = 0.18 \pm 0.19$ ), with the two latter significantly differing from each other. Regarding the social context, the pattern of results was different: still for CG, BT score of assessment ( $M = 1.92 \pm 0.17$ ) was higher than GT ( $M = 1.49 \pm 0.11$ ) and central area ( $M = 1.24 \pm 0.15$ ), the difference between the last two being significant.

#### Identifying key drivers and constraints in the micro and meso-environments

As previously mentioned, *social network* is perceived as a facilitator for the young SCD patients. In a previous work (Cholley-Gomez et al., 2021) concerning the social participation of Guadeloupian SCD participants vs control group, we found that, concerning the life habit related to *interpersonal relationships*, both had similar performance scores. However, further analyses helped to clarify that although the relations within close family and friends were similar between groups, the broader relations, with non-family and peers, were more difficult for the young with SCD.

In the present work, through analyzing items from MQE social factors, we aimed to identify which social spaces and social interactions could provide resources or facilitators in the social environment of SCD children. Results are outlined in Table 4.

Briefly, among items composing the *social network*, *support from family and loved ones* has the highest score. *Support from friends* was perceived as a facilitator, but a less important one among patients. Similarly, in the school context, *support from classmates* was a lower facilitator among the SCD group, and this pattern of results was also noted with *support from teaching staff*.

*Attitudes from entourage* provided, in the micro-environment of young SCD patients, a minor facilitator ( $M = 0.92 \pm 1.16$ ), but all its components were systematically and significantly lower rated by SCD respondents, except for *attitudes from the school doctor* and *attitudes from family and circle*, both dimensions being similarly perceived; and except for *religious beliefs*, which provided a stronger driver among the SCD group. Among the social circle, only items relating to

**Table 4. Mean score (SD) of MQE social factors according to group, and T-Student (p value)**

	SCD	CG	p
Factors	Mean ± SD	Mean ± SD	
Support from family and loved ones	1.85 ± 1.64	2.26 ± 1.19	0.02
Support from friends	1.34 ± 1.50	2.42 ± 0.86	< .001
Support from classmates	0.68 ± 1.56	1.88 ± 1.16	< .001
Support from teaching staff	0.75 ± 1.82	2.25 ± 1.05	< .001
Attitudes from school doctor	0.52 ± 1.44	0.78 ± 1.52	0.43
Attitudes from family and circle	1.43 ± 1.79	1.77 ± 1.66	0.29
Religious beliefs	1.77 ± 1.38	1.23 ± 1.21	< .05
Attitudes from persons in group	-0.46 ± 1.22	0.56 ± 1.46	< .001
Attitudes from unknown people	-0.64 ± 1.38	0.37 ± 1.39	< .001
Attitudes from friends	1.27 ± 1.34	2.11 ± 1.22	< .001
Attitudes from teaching staff	0.65 ± 1.92	1.66 ± 1.41	< .001
Attitudes from sports teacher	0.79 ± 1.84	1.94 ± 1.60	< .001

*attitudes from persons belonging to group, and unknown people* were negatively perceived. *Attitudes from friends, attitudes from teaching staff and attitudes from sports teacher* were more negatively perceived and assessed as weaker facilitators among SCD participants.

As previously noted, the *education services* score was lower among SCD respondents. In this category, except for the *orientation service* and *school catering service* showing no difference between groups ( $p = 0.12$ ;  $p = 0.48$ ), all the items were more negatively perceived by SCD participants. Moreover, two components were assessed as obstacles by young patients: *physical layout of school buildings* and *school hours* ( $M = -0.15 \pm 1.40$ ,  $M = -0.46 \pm 1.46$ , respectively).

On the larger scale of the environment, factors related to *cultural establishments* ( $M = 0.82 \pm 1.10$  vs  $M = 1.95 \pm 1.65$ ,  $p < .001$ ), recreational and sports organizations ( $M = 0.77 \pm 1.43$  vs  $M = 2.16 \pm 1.15$ ,  $p < .001$ ), and *public services* ( $M = 0.82 \pm 1.37$  vs  $M = 1.38 \pm 1.41$ ,  $p < .001$ ) provided less facilitators in the SCD patient's daily life. While *family incomes* were described as a major facilitator by controls, the score was lower (i.e., slightly negative) among SCD participants ( $M = -0.06 \pm 1.89$  vs  $M = 2.00 \pm 1.48$ ,  $p < .001$ ). In this regard, support provided by national Health Insurance & MDPH (*departmental center for persons with disabilities*) were strong facilitators ( $M = 2.04 \pm 1.58$ ) despite the negative perception of *administrative procedures* ( $M = -0.59 \pm 1.89$  vs  $M = 0.61 \pm 1.91$ ,  $p < .001$ ).

The macro-level of *political system* and its components related to public programs and policy were, in an equivalent manner for both groups, minor obstacles in daily life. *Media* was more positively perceived by controls ( $M = 0.77 \pm 1.63$  vs  $M = 1.97 \pm 1.09$ ,  $p < .001$ ).

Ultimately, although the analyses primarily focused on social contexts, results related to physical environments indicated that these factors restrain the daily activities of young patients with SCD, whereas they provide facilitators in controls' everyday life. This was particularly the case for

*climatic conditions* ( $M = -1.37 \pm 1.22$ ), *access to drinking water* ( $M = -1.13 \pm 1.35$ ), and *land-use planning* ( $M = -0.22 \pm 1.22$ ).

## 5. Discussion

As patients are faced with disease, environmental and psychosocial challenges, the significant impact of SCD on quality of life across a large group of diverse countries has been amply demonstrated (Graves et al., 2016; Sahil et al., 2019; Osunkwo et al., 2021; Houwing et al., 2022). However, to date, no previous study has investigated the social participation of SCD patients with an interactionist and systemic framework such as the Disability Creation Process. Yet a connection must be made between deficiencies, cognitive limitations or psychological determinants, and extrinsic factors, on different levels of the environment, in order to contextualize the mechanisms of disease and its consequences on the accomplishment of daily activities.

The aim of this study was to assess the perceived quality of environmental factors, both on their physical and social aspects and with a focus on social contexts, among young Guadeloupians with SCD, compared to a control group of healthy peers living in the same socio-cultural and geographical context. The main objective was to highlight the impact on social components, by stratifying them, e.g., family context, friend circle, school environment, or community level, and identifying in each setting the facilitators and obstacles to social participation. As we supported in a previous work (Cholley-Gomez et al., 2021) the hypothesis of an impact of living with SCD on the accomplishment of daily activities is evident, and we can here further understand the extrinsic limits to this participation.

Results emphasized that this restricted accomplishment occurs in a complex environment in which physical factors (e.g., land and infrastructure planning, natural elements) constitute strong barriers, and social determinants (e.g., support from friends, attitudes of schoolmates) are weaker facilitators. In short, key findings showed that all environmental factors assessed by the MQE were more negatively perceived among SCD participants, except for *socio-health services* (described as facilitators by both groups) and *political systems* (similarly rated as obstacles). Concerning this last macro-environmental factor, it must again be noted that due to the complexity of the items (e.g., public policy), assessment sometimes required that a parent or a relative be present with the child, if their age did not allow them to fully understand. Nevertheless, we observed the contextual effect that could be a potential bias and thus justified the control group in the study. Indeed, it appears that the perception of this contextual political factor was independent of health conditions, but rather should be replaced in the socio-cultural framework of Guadeloupe.

Concerning the positive perception of socio-health services, this result could reveal evidence of a social desirability bias among patients, as the assessment was made in a hospital context, in a specific care unit dedicated to SCD. In many cases, the investigator (social sciences researcher) was confusedly assimilated to a member of the medical staff. Data is not detailed in the current work, but our study design also included a qualitative axis with biographical interviews, often conducted at the home of patients and family, which provided more information, and qualified the quantitative data collected with questionnaires.

Results relating to socio-demographic characteristics also demonstrated that among the SCD group, only the occupational category of the family unit (and more precisely, the absence of a professional activity), had an impact on their perception of environmental factors, particularly social factors. Contrary to our previous findings showing the positive contribution of marital situation on patients' social participation, it appears here that single parenthood does not worsen the perceived quality of the environment. However, family support is a major facilitator, as is the support of friends although in a weaker manner. Sickle Cell Disease is a specific chronic condition, being more collective than personal, as it highly involves family (Mitchell et al., 2007; Reader et al., 2020). Indeed, this is firstly the case due to its genetic and hereditary nature, and secondly because of the daily and collective management it involves (frequent hospitalizations, knowledge

necessary to avoid and recognize vaso-occlusive crises or symptoms, reducing or ceasing professional activities).

In the micro and meso-environment, SCD patients face more difficulty in interpersonal relationships outside of their close social circle. Despite a larger media coverage at a local and national level, the disease is still stigmatized (Bulgin et al., 2018; Lainé, 2009; Pruneau et al., 2009, Pruneau et al., 2008), and more so in a school context. Indeed, in a previous work, our findings highlighted that participation in the school context was impacted by SCD status (Cholley-Gomez et al., 2021). In this setting, this limited accomplishment of their activity was, in addition, less compensated by positive environmental factors: schoolmates and teachers' attitudes, and more widely education services, were classified as resources, but less strongly compared to the perception of healthy participants. In this category, obstacles were non-human elements (i.e., physical disposition and schedule).

SCD patients' schooling is sometimes chaotic, with many gaps and absences, due to the frequency of crises and/ or hospitalizations (Dampier et al., 2010; Poku et al., 2018). At school, despite the progress made in terms of accompanying measures and information provided to education staff, some teachers can misunderstand the disease, believing that the child is lazy or plaintive (Pondarré et al., 2020). In a recent study investigating the relationships between cognitive and academic skills, (Pondarré et al., 2020), in line with previous work on cognitive functioning in SCD children (Bernaudin et al., 2000; Puffer et al., 2010; Schatz et al., 2002), noted that SCD does not systematically lead to visible difficulties or impairments, even though young individuals with SCD can easily tire, suffer from attention, concentration and memorization limitations, and thus should receive further attention from education staff.

Lastly, on a macro-environment level, public orientations and policies are negatively perceived by most participants, regardless of their health condition, and these factors do not improve the social participation of young patients with SCD. Disabilities generated by this condition make it necessary, on a local level, to create more inclusive spaces in school settings, professional training, sports and leisure activities, as well as in the cultural offer.

## 6. Conclusion

In applying the comprehensive DCP model to understand how these young patients interact with their family, and in educational or community-based settings, this work is in line with previous studies investigating multiple sclerosis, HIV or cystic fibrosis (Ferez et al., 2021, Ferez et al., 2015; Lacroix et al., 2016).

This study highlights the complex environment that young patients face, including physical and social components, and provides information for clinicians and researchers interested in understanding and improving quality of life and social participation in this young population. At a practical level, the DCP model (Badley, 2008; Whiteneck & Dijkers, 2009) has been widely used to guide policy development and service delivery within the province of Quebec, has supported the development of rehabilitation programs, in addition of having being a useful framework for aligning research with clinical practice (Fougeyrollas, 2010; Fougeyrollas et al., 2019).

Our findings have implications for practices of professionals and local stakeholders who work at the various levels and within the different spheres of a young patient (e.g., medical and educational professionals, local political authorities, members of local associations, relevant institutions). In particular, healthcare professionals play a crucial role for patients and families, as socio-health services are positively perceived and are a facilitator in the child's environment. School is also an important setting for the social participation of a child. In order to promote optimal relationship with peers, and academic functioning, teachers and school-based caregivers should provide more support for children with SCD (e.g., by allowing learning accommodations and recourse to education services).

Finally, additional analyses will complete this data by considering the dynamic interaction between environmental and individual factors (e.g., clinical manifestations of SCD, neuropsychological disorders) when examining the social participation of individuals with SCD. These conclusions will stimulate reflection concerning the challenges and opportunities offered by the transdisciplinarity of this project and its conceptual rooting in a complex model such as the DCP, issues that are the focus of an ongoing discussion (Ruffié et al., 2020).

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### Author details

Marie Cholley-Gomez<sup>1</sup>

E-mail: [marie.cholleygomez@hotmail.fr](mailto:marie.cholleygomez@hotmail.fr)

ORCID ID: <http://orcid.org/0000-0001-7555-6079>

Sébastien Ruffié<sup>1</sup>

E-mail: [sebastien.ruffie@univ-antilles.fr](mailto:sebastien.ruffie@univ-antilles.fr)

Benoit Tressières<sup>2</sup>

E-mail: [benoit.tressieres@gmail.com](mailto:benoit.tressieres@gmail.com)

Gaël Villoing<sup>1</sup>

E-mail: [villoinggael@gmail.com](mailto:villoinggael@gmail.com)

Sylvain Ferez<sup>3</sup>

E-mail: [sylvain.ferez@umontpellier.fr](mailto:sylvain.ferez@umontpellier.fr)

<sup>1</sup> ACTES Laboratory. “Adaptations to Tropical Climate, Exercise and Health” (EA N°3596), University of Antilles – Guadeloupe, Campus Fouillole, 97110, Pointe-à-Pitre, Guadeloupe.

<sup>2</sup> Data Manager, Centre d'Investigation Clinique Antilles Guyane, Inserm CIC 1424, CHU de la, Guadeloupe.

<sup>3</sup> Département Hérault, Université de Montpellier, 700, Avenue du Pic Saint-Loup, Lab. SanteSIH, 34090, Montpellier, France.

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### Notes

1. The *Unité Transversale de la Drépanocytose*—UTD, Reference centre for sickle cell disease, part of the Pointe-à-Pitre hospital, initiates a complex and diverse approach to care, through the combination of medical, psychological and social monitoring. Paediatricians, neuropsychologists and computer engineers of the Clinical Investigation Centre of the hospital also participated in this research.
2. Other major sickle cell syndromes present compound heterozygous forms in which haemoglobin is associated with certain abnormal haemoglobins; S/C, S/D Punjab, S/Lepore, S/E forms (Etienne-Julan & Saint-Martin, 2005) but are less symptomatic and are not taken into account in this project.
3. The Guadeloupean landscape presents various territorial, climatic and cultural specificities. The Northern or superior part, named Grande-Terre and more urban, is more exposed to the trade winds and is much drier than Basse-Terre due to the lower

elevation (maximum 125 metres above sea-level) and the geology. The surface hydrography there is low; the island of Basse-Terre, more rural and where the Soufrière volcano can be found, along with the humid tropical forest covering its mountainous heights, is marked by a higher altitude and more relief, as well as more frequent rain (Pagney, 1991). The part known as Central (including Baie-Mahault, Pointe-à-Pitre and the Abymes) constitutes the active heart, the industrial and commercial area of Jarry being considered the economic lungs of the territory.

4. An eCRF is a digital form, the goal of which is to obtain information useful to a clinical research project.

### Disclosure statement

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.2021

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