

# Self-management support for young people living with fluctuating chronic diseases

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## FLUCTUATING CHRONIC DISEASES

Unpredictable fluctuations, episodic deteriorations and flares are common in many paediatric chronic diseases, for example, asthma, type 1 diabetes mellitus, juvenile idiopathic arthritis (JIA), epilepsy, systemic sclerosis, cystic fibrosis, inflammatory bowel disease and sickle cell disease.<sup>1-4</sup> For many children and adolescents, their chronic disease is not a steady state of illness. On the contrary, a continuous ebb and flow of symptoms interferes with functional and social daily life.<sup>1-4</sup>

These fluctuations constitute a difficult challenge to patients' ability to self-manage their disease, fundamentally different from more predictable illness patterns. In this Viewpoint, we—three professionals and three young adult patients—argue that the existing self-management programmes for children and adolescents with chronic illness and their families do not sufficiently prepare them to respond to these mostly non-modifiable fluctuations. In our view, the existing focus of most self-management programmes on the acquisition of skills to control presumed continuous symptoms might even be counterproductive.

Young people with chronic disease have a strong desire to be like others, to fit in and to be accepted by their peers.<sup>1</sup> The fear of rejection and the stigma associated with their condition often prevents them from disclosing their illness. In 'good' periods, there seems to be no reason for disclosure. Periodically, however, they may experience temporal deteriorations of their disease. The worsening of symptoms and their incapacitating effects disrupt normality and their daily life, leaving them with less control over what activities they

can engage in. During these 'bad' periods, even the most skilled adolescents may be defaulted back to parental care and support.<sup>4</sup>

Being unable to anticipate these disease fluctuations and flares contributes to a broader perception that they lack control over their lives.<sup>2,3</sup> Young patients living with JIA, for instance, believe themselves to be "trapped in a perpetual limbo, oscillating between having a sense of control over their bodies and losing all control as moments of intense pain overwhelmed them".<sup>1</sup> They feel that others do not recognise the seriousness and unpredictability of their arthritis flares. Many patients with JIA are deeply frustrated and hurt by the lack of empathy and understanding from peers, school personnel, family members and team mates in sports. This is described as the worst thing about living with JIA.<sup>1</sup> The patient-authors of this Viewpoint endorse this from personal experience. It does not only apply for JIA (online supplemental material). Clearly, learning how to deal with mistrustful healthy peers is one of the most important self-management support needs for many young people suffering from fluctuating diseases.<sup>1,4</sup>

## CURRENT SELF-MANAGEMENT SUPPORT

Thus far, most self-management programmes for young people with chronic conditions tend to focus on medical management (ie, adherence to treatment and healthy living) alone.<sup>5</sup> However, two other aspects of self-management—role management (ie, social participation), and emotion or identity management (ie, emotional consequences of being ill)—are commonly seen as equally important for young people suffering from fluctuating disease.<sup>5</sup>

Many paediatric self-management programmes are based on a theoretical model.<sup>5-7</sup> The most commonly used models—for example, cognitive-behavioural therapy, social cognitive theory and the health behaviour model—have in common that they aim to

strengthen individual agency. The acquisition of skills to self-manage presumed continuous symptoms is at the heart of these theories. A strengthened sense of mastery is thought to lead to more self-efficacy.<sup>5-8</sup> Developing young people are thought to grow into self-management, and gradually take over parents' responsibilities, as they acquire more knowledge, skills and confidence.<sup>4,6</sup>

Fluctuations and their impact on daily social life are rarely addressed in these theories and programmes<sup>5-7</sup>. Consequently, many programmes recommend these young people to make a realistic estimate of what they typically can and cannot do, as a result of their illness. For fluctuating diseases, this recommendation just does not make sense. Interestingly, in one model the prevention of flares is explicitly presented as a self-management task.<sup>5</sup> This—however—is an impossible task, as most flares cannot be prevented. For programmes that are supposed to strengthen self-efficacy, making these young people responsible for the occurrence of flares is clearly counterproductive.<sup>3</sup>

Self-management support programmes seek to help adolescents in the development of a robust identity and self-story.<sup>6</sup> Keeping their illness hidden and striving for normality is regarded as a barrier for efficacious self-management.<sup>7</sup> In real life, however, fluctuating disease clearly hinders the development of a single identity.<sup>1-4</sup> Many of them seem to develop two separate roles or identities.<sup>2,3</sup> In good periods, they show control, autonomy and self-reliance. In bad periods, however, they suddenly lose control and may need assistance from their family and peers.<sup>4</sup> In our view, effective self-management support should recognise both alternating roles, for which the uncontrollably oscillating body sets the agenda.

## FUTURE SELF-MANAGEMENT SUPPORT

How can new self-management programmes really help these young people? Based on the literature and focus group interviews with young people suffering from several fluctuating chronic diseases, we suggest that insurmountable flares and fluctuations can no longer be neglected. Instead, dealing with and preparing for fluctuations should be at the heart of future self-management support.<sup>1-4</sup> As this aspect is central to many paediatric diseases, these programmes could very well be transcending disease-specific strategies.<sup>5</sup>

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For these new programmes, the commonly used theoretical models aimed at mastery and control may not suffice: elements of other models might be helpful. In self-management programmes based on the Acceptance and Commitment Therapy (ACT) model, for instance, participants are explicitly educated about the limits of control.<sup>8</sup> In a recent ACT-based programme for paediatric chronic pain, the overarching goal was “to decrease ineffective struggles for control of pain or distressing emotions”.<sup>9</sup> Furthermore, the involvement of social networks in self-management programmes seems to be a very promising approach.<sup>10</sup>

What are the implications for practice? The first thing young children with chronic conditions should hear from their doctor is that they are not responsible for their flares. Over the years, they should learn to accept the uncontrollable fluctuations and learn to adapt to new and unpredictable situations.<sup>3,9</sup> The allocation of responsibilities between adolescents and parents in co-management should be formulated flexible, depending on the current severity of the disease and the developmental stage of the adolescent or young adult.<sup>4</sup>

For these young people, learning how to deal with their healthy peers is a top priority.<sup>1-4</sup> As a result of their last-minute cancellations, they often feel ‘unreliable’ as a friend, co-worker or team mate.<sup>2</sup> They need to think very carefully about who to disclose to and when. In ‘good’ periods, striving for normality could be regarded a tricky but healthy reaction. In the light of possible flares, however, it is important to learn to expect and adapt to two separate roles. Furthermore, after each transition in their lives, they will need to readjust both roles, based on their own developmental stage and their new peers.

In conclusion, there seems to be a large gap between the literature based on the experiences of young people living with a chronic disease,<sup>1-4</sup> and most paediatric self-management literature.<sup>5-7</sup> In this Viewpoint, written by professionals and patients together, we made an attempt to close this gap. In future research,

incorporating young people’s experiences should be considered essential for co-producing new programmes that match the needs of all those involved.<sup>3</sup>

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### **Social consequences of unpredictable fluctuations in self-management interventions for young patients with chronic diseases: a rapid review**

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Unpredictable fluctuations, episodic deteriorations, and flares are common in many pediatric chronic diseases. For many children and adolescents, their chronic disease is not a steady state of illness. On the contrary, a continuous ebb and flow of symptoms interferes with functional and social daily life. These fluctuations constitute a difficult challenge to patients' ability to self-manage their disease, fundamentally different from more predictable illness patterns.<sup>1-3</sup>

Numerous self-management interventions for young people with chronic conditions have been developed. do these self-management interventions offer young patients guidance on how to deal with the lack of understanding and even mistrust in school or at work, as a result of fluctuating disease activity?

In 2015, Sattoe et al.<sup>4</sup> performed an extensive search for papers on self-management interventions for 7 to 25-year-olds with somatic chronic conditions or physical disabilities. They distinguished three tasks involved in self-management: medical management (re. treatment), role management (re. social participation), and emotion or identity management (re. emotional consequences of being ill)<sup>4</sup>. The unpredictability of fluctuating symptoms substantially *affects role-management* in school or at work<sup>1,3</sup>. In 34 of the 86 studies (40%) included in the review, some form of role self-management was addressed<sup>4</sup>.

Three reviewers (CS, DvV, SvG) independently assessed these 34 papers, to find clues on interventions dealing with social consequences of fluctuating disease activity in school or at work. One of the authors (DvV) emailed all researchers in order to increase sensitivity. We asked the authors to send us the manual or intervention description of their program. Furthermore, we asked them if they thought the theme of unpredictable fluctuations had been addressed in their intervention. Three reviewers (CS, DvV, SvG) independently assessed all received information. The findings were discussed and consensus was reached on potential disaccord.

We assessed the 34 papers in the Sattoe et al. review in which some form of role self-management had been mentioned<sup>4</sup>. In 30 papers (88.2%), the social consequences of unpredictable fluctuation were not mentioned at all. In four papers<sup>5-8</sup>, we found some minor clues that it might have been a topic in the studied programs. We thoroughly assessed these papers and the underlying materials.

In the introduction of a paper on a school intervention for children with Sickle Cell, Koontz et al. stated: "The unpredictable, episodic nature of the disease can be especially disruptive to maintaining a normal schedule"<sup>5</sup>. However, in the description of the intervention, this theme was not addressed; the authors confirmed it was not part of the intervention. In a cognitive-behavioral program for adolescents with chronic pain, there was mention of the complex social and psychological problems associated with chronic pain, among which school absenteeism and nuisance in the

adolescents' lives<sup>6</sup>. Yet, the overview of the content of the program did not refer to unpredictability or fluctuations. In a web-based intervention for adolescent with type 1 diabetes, psychosocial demands such as struggling for identity, achieving independence and gaining peer acceptance were all mentioned as factors affecting compliance and school participation<sup>7</sup>. The author sent the method section of her PhD-thesis: the theme of unpredictable fluctuations was not specifically addressed. In the description of a camp for young people with end-stage renal disease, we found some potential clues that the topic of unpredictability might be addressed<sup>8</sup>. However, the authors confirmed that the theme was not part of their manual. Still, in case the participants brought this theme up, it might have been discussed.

The authors of 16 papers (47%) responded to our inquiry. For 12 interventions, we received additional information, a manual or a protocol. In this extra information, there was no indication whatsoever that the programs dealt with the social consequences of unpredictable fluctuation of symptoms. None of the responding authors indicated that part of their intervention had been specifically aimed at this theme.

Based on this rapid review, we concluded that current self-management interventions for young patients with chronic diseases do not seem to target the social consequences of unpredictable fluctuations.

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