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Achieving self-management of prophylactic treatment in adolescents: The case of haemophilia

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ABSTRACT

Objective: Adolescents with a chronic disorder, such as haemophilia, need to attain responsibility for their disease. The aim was to gain insight into how adolescents achieve self-management of prophylactic treatment.
Methods: In three Dutch Haemophilia Treatment Centres, adolescents (10–25 years) received structured questions on treatment responsibility and self-management (pre-specified definitions) during routine nursing consultation.
Results: In total, 155 interviews were performed in 100 patients (median age 14.4 years). Self-infusion was initiated at a median age of 12.3 years (IQR 11.5–13.0) and self-management was achieved 9.6 years later, at a median age of 22.6 years. This process included three phases coinciding with known stages of adolescence. In early adolescence, patients acquired the technique of self-infusion (12.3 years) leading to independent self-infusion in middle adolescence (17.2 years). In late adolescence, patients demonstrated an increase in more complex skills, such as bleeding management and communication with the haemophilia physician (19.9–22.6 years).
Conclusion: Although, the first steps in self-management with regard to self-infusion are taken in early adolescence, complete self-management was achieved in late adolescence after almost 10 years.
Practice implications: Insight in this transitional process helps to provide individualized support and emphasizes the need for continued education with regard to self-management skills.

1. Introduction

Adolescents experience physical, cognitive and psycho-social changes, including many aspects such as maturing of the body, an evolving identity, increasing independence and intimacy [1]. In addition, adolescents with chronic illnesses are also confronted with the challenges in attaining responsibility for their disease and its treatment. The desire to be as others often leads to non-adherence, with concomitant increased risk for complications and deterioration of the illness [2,3]. Self-management skills are learned in this difficult age period, which is defined as 'the individual's ability to manage the symptoms, treatment, physical and psychosocial consequences and lifestyle changes inherent in living with a chronic condition' [4].

Patients suffering from the inherited bleeding disorder haemophilia have a lifelong risk of bleeding and subsequent arthropathy [5,6]. Standard therapy is intravenous clotting factor replacement therapy administered at established intervals per week, also referred to as prophylaxis [7,8]. In the Netherlands, the majority of patients with severe haemophilia practice self-infusion of prophylaxis [9]. Prophylaxis in the home setting has greatly improved quality of life in haemophilia patients [10]. Adolescents with haemophilia mostly start with learning self-infusion at the age of 13 years [11], this is a procedure involving complex self-management skills.

In current Dutch practice, young adolescents follow a short course to learn the technique of self-infusion and the theoretical background of their illness, symptoms and treatment [11,12]. It remains unknown when patients acquire the more comprehensive

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self-management skills and take full responsibility for their treatment. In a recent qualitative study it was identified that low self-management skills were an obstacle for optimal adherence to prophylaxis [13]. Little is known about the development of self-management skills in adolescents with haemophilia. The aim of this project was to gain insight in the process of achieving self-management in adolescents with haemophilia using prophylaxis.

2. Methods

This study comprised a cross-sectional, multicentre study in three Dutch haemophilia treatment centres (Utrecht, Amsterdam and Rotterdam). Adolescents (10–25 years) were interviewed about topics (Box 1) with regard to self-management and treatment responsibility. This structured interview was implemented in a routine nursing consultation. The research ethics committee of the University Medical Centre Utrecht, The Netherlands, approved this study (10/269).

2.1. Participants

All patients between the age of 10 and 25 years with a congenital clotting factor deficiency (moderate or severe haemophilia A or B and Von Willebrand disease type III) using prophylaxis with a minimum frequency of once per week, were eligible for inclusion. Convenience sampling was used: patients were interviewed during their regular clinic visit during the period September 2010 until December 2013.

2.2. Data collection

The following baseline patient characteristics were collected from patient files: age, diagnosis, the person performing the intravenous infusion, route of intravenous access (IV), date of exam self-infusion and current treatment (start date of prophylaxis, prescribed regimen and when applicable, age learning self-infusion). Furthermore, structured interviews were conducted during a nursing consultation including closed questions about aspects of the treatment and the individual who was responsible for performing the treatment (patient, parent or both) (Box 1). These treatment aspects involved: learning self-infusion, independent performance and remembering of self-infusion, use of infusion diary, stock management, treatment decisions (diagnosing bleedings and subsequent dosing), and communication with the physician. The questions were based on the Dutch educational guideline for learning self-infusion [12] and were consistent with daily practice. Complete self-management of prophylactic treatment was defined as: a patients' ability to act independently concerning self-infusion, handling during bleeding episodes, monitoring of stock, and communication with the physician. To verify answers and assess validity, parents were asked the same questions in a separate room. Repeated interviews were allowed with a minimum interval of 1 year.

2.3. Analysis

Internal validity between answers of patients and parents was assessed by Cohen's kappa. To study the potential bias introduced by repeated interviews in some patients, the analyses were repeated after exclusion of the second or third interviews in these patients. The same trends were observed ($p = 0.2–0.9$) and therefore ultimately all interviews were used in the analysis. Median ages (interquartile range) were calculated for each activity based on the cross-sectional measurement; in short the median age was based on which activity is the adolescent able to perform at that moment. In addition, proportions were calculated for three specific age groups: early adolescence (10–12.5 years), middle adolescence (12.5–17.5 years) and late adolescence (17.5–25 years) [19,20]. Differences according to age groups were assessed using the Chi square test and were considered statistically significant at a $p < 0.05$. All data processing and analyses were performed with SPSS® software, version 20 (SPSS Inc., Chicago, IL, USA).

3. Results

In total, 155 interviews were conducted in 100 Dutch patients (Utrecht $n = 70$, Amsterdam $n = 22$, and Rotterdam $n = 8$). Overall, the response rate was 67% of the adolescents using prophylaxis were questioned. The patient characteristics are shown in Table 1. Median age was 14.4 years (Interquartile range (IQR):12.5–18.1), diagnosed with severe haemophilia (87% A, 10% B) and Von Willebrand type III (3%). Two patients (2%) had a central venous access device. The majority of patients infused three times per week the prophylactic treatment (64%) with a median dose of

Table 1
Characteristics.

	Total interviews $N = 155$
Interviews patients*	100
2nd interview	35
3rd interview	20
*Interviews parents**	38
Age (med, IQR)	14.4 years (12.5–18.1)
Diagnosis	
A	135
B:	15
VWB	5
Type	
Severe	145
Moderate	5
Type III	5
Venous access	
Peripheral vein	153
CVAD	2
Frequency prophylaxis (med, range)	3.0 (2–7)
Dose prophylaxis (med, range)	1000 (500–2000)

Values are frequencies and medians.
*First and repeated interviews did not show systematic differences, therefore all interviews were used in further analyses ($p = 0.2–0.9$).

Box 1. Interview questions.

- Who performs the self-infusion?
- Who reminds you to self-infuse?
- Who completes the infusion diary?
- Who decides to infuse during a bleed? And who decides the dosing?
- Who takes care of the stock?
- Who takes the lead in the communication with the health care provider?