Transition readiness among adolescents and young adults with haemophilia in the Netherlands: nationwide questionnaire study

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ORIGINAL ARTICLE

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Transition readiness among adolescents and young adults with haemophilia in the Netherlands: Nationwide questionnaire study

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Abstract

Introduction: Care for adolescents with haemophilia is transferred from paediatric to adult care around the age of 18 years. Transition programs help to prepare adolescents for this transfer and prevent declining treatment adherence. Evaluating transition readiness may identify areas for improvement.

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Objective: Assess transition readiness among Dutch adolescents and young adults with haemophilia, determine factors associated with transition readiness, and identify areas of improvement in transition programs.

Methods: All Dutch adolescents and young adults aged 12–25 years with haemophilia were invited to participate in a nationwide questionnaire study. Transition readiness was assessed using multiple-choice questions and was defined as being ready or almost ready for transition. Potential factors associated with transition readiness were investigated, including: socio-demographic and disease-related factors, treatment adherence, health-related quality of life, and self-efficacy.

Results: Data of 45 adolescents and 84 young adults with haemophilia (47% with severe haemophilia) were analyzed. Transition readiness increased with age, from 39% in 12–14 year-olds to 63% in 15–17 year-olds. Nearly all post-transition young adults (92%, 77/84) reported they were ready for transition. Transition readiness was associated with treatment adherence, as median VERITAS-Pro treatment adherence scores were worse in patients who were not ready (17, IQR 9–29), compared to those ready for transition (11, IQR 9–16). Potential improvements were identified: getting better acquainted with the adult treatment team prior to transition and information on managing healthcare costs.

Conclusions: Nearly all post-transition young adults reported they were ready for transition. Improvements were identified regarding team acquaintance and preparation for managing healthcare costs.

KEYWORDS
Adolescent, Haemophilia A, Haemophilia B, Self-Management, Transition to Adult Care, Treatment Adherence and Compliance, Young Adult

1 | INTRODUCTION

The prevalence of chronic diseases is rising.[1,2] Especially in the treatment of chronic diseases, self-management skills are crucial.[3] Self-management is defined as the ability to manage clinical, psychosocial and societal aspects of illness and its care.[4] This includes the integration of treatment activities into everyday life and the ability to react to symptoms.[5] Self-management requires a high level of health knowledge and treatment adherence.[6] However, treatment adherence generally decreases during adolescence.[7] It is in this phase of gradually achieving independence that adolescents transfer from paediatric to adult care.

This decrease in treatment adherence is also observed among adolescents with haemophilia.[5,8] Haemophilia is an inherited deficiency of coagulation factor VIII (haemophilia A) or IX (haemophilia B). Based on the residual coagulant activity, the severity of haemophilia is classified as either severe (factor activity < 0.01 IU/mL), moderate (.01-.05 IU/mL), or mild (.05-.40 IU/mL).[9] People with a severe bleeding phenotype, as seen in the majority of people with severe and moderate haemophilia, have spontaneous joint and muscle bleeds resulting in joint damage. To prevent bleeds, these people repeatedly self-infuse with prophylactic injections of coagulation factor concentrates or non-factor replacement therapies. Treatment outcomes depend on treatment adherence.[10–12] A previous global study reported that 90% of children with severe and non-severe haemophilia under 12 years of age have high treatment adherence.[8,13] However, treatment adherence decreases to 54% among adolescents aged 13—18 years, and further to 36% among young adults aged 19—28 years who transitioned to adult care. Low treatment adherence can result in joint bleeds and reduced quality of life.[14,15]

To prevent decreases in treatment adherence and health outcomes, transition programs have been developed. Transition programs aim to prepare adolescents and their parents in a structured way for transferring to adult care. An individual transition plan is drafted to monitor the gradual introduction of new skills and responsibilities related to medical, psychosocial and societal aspects of care.[16] A national Dutch protocol specifies its contents (Box 1). Consequently, transition programs are highly similar among Dutch care institutions, although local differences exist (e.g. specialized nurses exclusively treat children, adults or both). Haemophilia quality accreditation requires the use of a transition program.

The World Federation of Haemophilia (WFH) has established guidelines for the transition from paediatric to adult care that closely resemble the Dutch guidelines.[37] Ultimately, a transition plan teaches adolescents to take ownership of their condition and their transition, and to speak up for themselves.

In 2003, one of the first transition programs in the Netherlands was implemented in haemophilia care.[17] Yet, knowledge on the impact of transition programs on how adolescents and young adults perceive transition is limited. Previous studies either assessed outdated Dutch transition programs, or recently-implemented international programs.[17–21] Although transition programs aim to improve treatment adherence, health-related quality of life, and self-efficacy, associations between transition readiness and these characteristics in haemophilia care remains unknown.[12] Yet, these insights are crucial to further improve current transition programs. Therefore, we aimed to assess the self-reported readiness to transition from paediatric to adult care among adolescents and young adults with haemophilia in the Netherlands, and to identify associations of transition readiness with socio-demographic and disease-related factors, and between transition readiness and treatment adherence, health-related quality of life, and self-efficacy. Finally, we aimed to identify areas for improvement of transition programs.

2 | METHODS

This cross-sectional study was part of 6th nationwide, multi-centre 'Haemophilia in the Netherlands' (HiN6) study. The methods of this study have been described in detail.[11]

2.1 | Participants

Between June 2018 and July 2019, all 2191 male people with haemophilia A or B treated in one of the haemophilia treatment centres in the Netherlands were invited to participate in the nationwide questionnaire study. All six centres deliver comprehensive medical and psychosocial care. Centres are required to treat at least 40 patients with severe haemophilia, of which 10 children.[22] For this study on transition readiness, adolescents aged 12—17 years and young adults aged 18—25 years were eligible.

2.2 | Data collection

The HiN6 questionnaire covered multiple aspects of haemophilia. Age-specific questionnaires were sent to adolescents and young adults. Patient-reported questionnaire data on diagnosis were compared to clinician-reported electronic health record data to improve data reliability. The latter were used in case of discrepancies. Participants who completed the questions on readiness to transition were included in this study.

2.3 | Outcomes

The primary study outcome was self-reported transition readiness, defined as the readiness to transfer to adult care. Secondary outcomes were transition preparation, defined as whether adolescents and their care providers had adequately discussed transition-related topics and identified areas that might require extra preparation before transitioning. Potential determinants of transition readiness were socio-demographic characteristics and disease-related factors. Treatment adherence, health-related quality of life, and self-efficacy were evaluated and compared in adolescents that were ready and not (yet) ready for transition.

BOX 1 Transition plan[39–41]

From the age of 10 years onwards, an individual transition plan is drafted. This plan helps the treatment team to guide and monitor the gradual introduction of new skills and responsibilities related to medical, psychosocial and societal aspects of care, including:

- Knowledge of haemophilia, its signs and symptoms, and risky behaviour (e.g., alcohol, drugs, extreme sports, piercings, tattoos)
- Knowledge of the treatment of haemophilia
- Proficiency with the self-administration of treatment and management of bleeds (e.g., contacting a haemophilia treatment centre). Based on their own needs, children generally start to participate in the self-infusing process from the age of 6 years onwards (e.g., cleaning the injection site). Gradually, they increase their participation, and learn to fully self-infuse around the age of 12 years.
- Responsibilities in how to use a (digital) treatment diary and related medication inventory management
- Worries about haemophilia (e.g., regarding sexuality, menstrual bleeding, inheritance)
- Knowledge of which healthcare providers adolescents will encounter in adult care
- Knowledge of the available guidance in issues related to work, school or money
- Other aspects of life, such as:
  - School, future plans, career planning, and work experience
  - Spare time, activities with friends, and relationships

2.4 | Measures

Both validated and newly-composed questionnaires were used for outcome assessment.

2.4.1 | Transition readiness

To assess self-reported transition readiness, adolescents were asked: 'Do you think that you will be ready to transfer to adult care when you

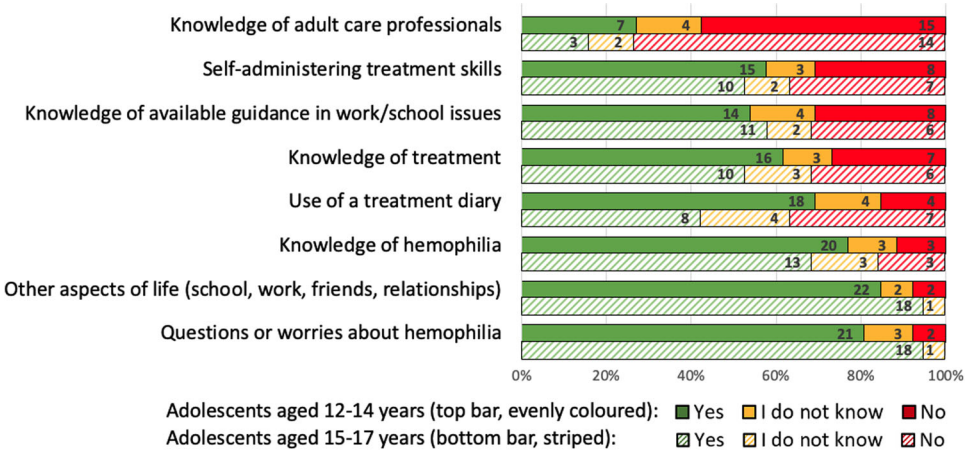


FIGURE 1 Transition preparation. Answers of 45 adolescents aged 12–14 years (top bar, evenly coloured) and 15–17 years (bottom, striped bar) on the question: 'Was [the respective transition preparation domain] discussed in an outpatient clinic visit?' Participants could answer 'yes' (in green), 'I don't know' (in yellow), or 'no' (in red). Numbers of respondents per answering option are shown within the bars.

turn 18?' Answer options included: 'ready', 'almost ready', 'not ready', or 'I don't know'. Young adults were asked: "Were you ready to transfer to adult care?" Answer options included: 'yes', 'no', and 'I don't know'. Both questions are newly composed.

2.4.2 | Associations of transition readiness with socio-demographic and disease-related factors, and with treatment adherence, health-related quality of life, and self-efficacy

Associations of socio-demographic and disease-related factors with transition readiness were assessed, including disease severity, haemophilia family history, and education level. Dutch versions of the VERITAS-Pro, CHO-KLAT, HSES questionnaires were used to assess treatment adherence, health-related quality of life, and self-efficacy respectively.

VERITAS-Pro

The Haemophilia Regimen Treatment Adherence Scale—Prophylaxis (VERITAS-Pro) was used to assess the adherence to prophylactic treatment in the past two weeks among adolescents with severe haemophilia.[13,23,24] It consists of 24 statements that are scored on a five-point Likert scale, with answer options ranging from never to always. The questions are split over six domains: 'Time' (adherence to prescribed schedule), 'Dose' (adherence to dosage), 'Plan' (inventory management), 'Remember' (forget infusions), 'Skip' (postpone/skip infusions) and 'Communicate' (contact professionals if needed). Both domain scores and overall VERITAS-Pro scores are presented as a standardized, continuous variable ranging from 0 to 100, with lower scores indicating better adherence. Good treatment adherence is defined as a standardized total score below 34.[25] VERITAS-Pro's reliability among Dutch boys with severe haemophilia using prophylaxis is good (Cronbach's α =0.70). Domain reliability varies from 0.39 ('Dose') to 0.73 ('Skip').[13]

CHO-KLAT

For assessment of health-related quality of life, the Canadian Haemophilia Outcomes–Kids Life Assessment Tool (CHO-KLAT) was used.[26,27] CHO-KLAT is a haemophilia-specific questionnaire for boys aged 4–18 years. CHO-KLAT consists of 35 statements regarding the past four weeks, which are scored on a five-point Likert scale, with answer options ranging from never to always. The total, standardized score is presented as a continuous variable ranging from 0 to 100, with higher scores indicating higher health-related quality of life. CHO-KLAT's reliability among boys with haemophilia aged 8–16 years is excellent (Cronbach's $\alpha = 0.81$-0.91), and its test-retest reliability in this group is high (intraclass correlation coefficient 0.74).[28]

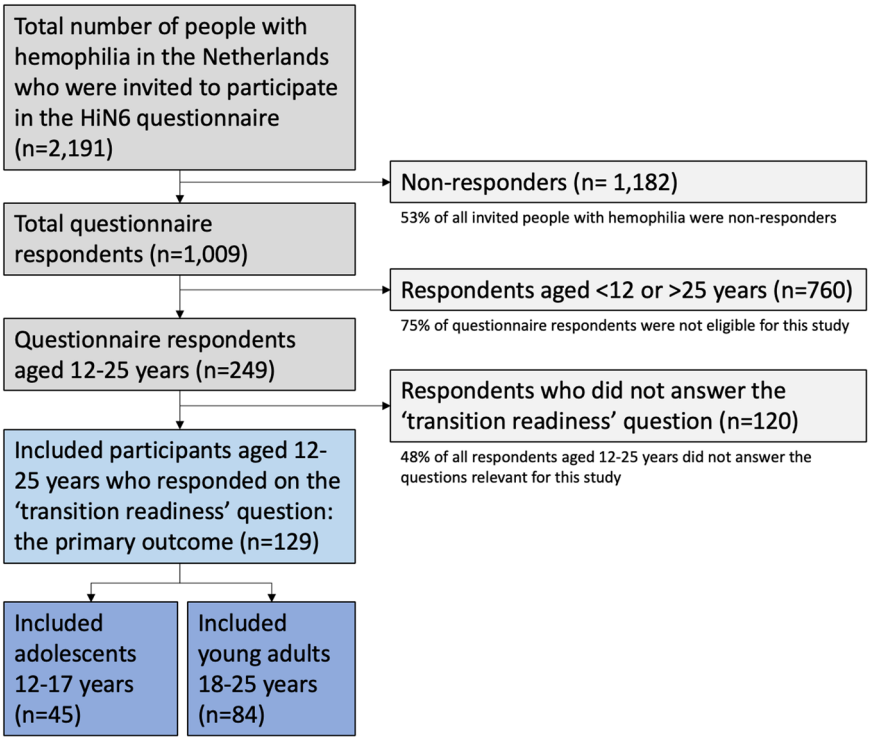
HSES

The Haemophilia-specific Self-Efficacy Scale (HSES) was used to assess self-efficacy.[29] Self-efficacy is an individual's belief he can successfully execute self-management behaviour; to cope with or manage clinical, psychosocial, and societal aspects of illness and its care.[30] The HSES consists of 12 statements that are scored on a five-point Likert scale, with answer options ranging from totally disagree to totally agree. The overall, non-standardized score ranges from 12 to 60, with higher scores indicating a greater self-efficacy. The reliability of the adolescent-reported HSES is excellent among Dutch boys with haemophilia using prophylaxis (Cronbach's $\alpha = 0.86$).[29]

2.4.3 | Transition preparation

To identify areas for improvement of transition programs we assessed transition preparation. Adolescents indicated which transition-related topics they had discussed with their healthcare providers. These eight newly-composed topics are listed in Figure 1, and match the topics of the Dutch haemophilia transition protocol (Box 1). Answer options included: 'yes', 'no', and 'I don't know'.

FIGURE 2 Participant flowchart.



To further explore areas for improvement, young adults were asked: 'What aspects of transitioning would you have liked to be better prepared for at the children's hospital?' Out of eight options, participants could select one or multiple aspects: knowledge of haemophilia, self-infusion skills, understanding treatment responsibilities (including ordering medication), understanding responsibilities in planning/keeping appointments, managing healthcare costs, guidance in work/school issues, and getting acquainted with healthcare providers in adult care. The eighth option was an open-ended answer choice.

2.4.4 | Data analysis

Descriptive statistics were used and reported as median values with interquartile range (IQR). To measure an increase in transition readiness, we analyzed adolescents aged 12–14 years and 15–17 years separately. For the total VERITAS-Pro, CHO-KLAT, and HSES scores, a minimum data completion rule of 75% of items was used. Standardization of the CHO-KLAT and VERITAS-Pro scores was done with this formula: [100%× ((raw score−minimal possible raw score) / possible range of raw scores]. To investigate factors associated with transition readiness, readiness was categorized into two groups: '(almost) ready' and 'unsure or not ready'. The former included the 'ready' and 'almost ready' answering options, the latter the 'not ready' and 'I don't know' options. Relative risks with 95% CI were calculated for categorical factors. Data were analyzed using SPSS 26.0. *p*-Value <.05 were considered statistically significant.

2.4.5 | Ethics

Ethical approval was obtained from the Institutional Review Board of the Leiden University Medical Centre (NL59114.058.17). Participants who completed the questionnaire were considered to consent.

3 | RESULTS

3.1 | Participant characteristics

The overall response rate of participants completing the full HiN6 questionnaire was 46% (1009/2191), as shown in the study flowchart (Figure 2). Respondents were similar to the overall Dutch haemophilia population regarding age and disease severity.[10] Of all respondents, 249 were aged 12–25 years. In total, 52% (129/249) completed the question on transition readiness and were included in this study. Of the 129 participants, 45 (35%) were adolescents and 84 (65%) young adults. Included participants were similar to all questionnaire respondents aged 12–25 years regarding the proportion with severe haemophilia (47% and 44%, respectively) and prophylaxis use (50%; 49%). Participant characteristics are presented in Table 1.

3.2 | Transition readiness

Of 45 adolescents, 49% (22) were '(almost) ready' for transition, and 51% (23) were 'not ready or unsure'. Of adolescents aged 12–14 years, 39% (10/26) were '(almost) ready', compared to 63% (9/19) of

TABLE 1 Baseline characteristics of included participants.

	All participants ($n = 129$)	Adolescents 12–17 years ($n = 45$)	Young adults 18–25 years ($n = 84$)
Age in years; median (IQR)	20 (15-22.5)	14 (13-15)	21 (20-24)
Haemophilia A, n (%)	111 (87%)	37 (84%)	74 (88%)
Severe haemophilia, n (%)	60 (47%)	27 (60%)	33 (39%)
Prophylaxis, n (%)	64 (50%)	27 (60%)	37 (44%)
Positive haemophilia family history, n (%)	99 (77%)	34 (76%)	65 (77%)
Any hospital admission in the past 12 months, n (%)	16 (12%)[a]	6 (14%)[a]	10 (12%)
Any joint bleed in the past 3 or 12 months[d], n (%)	23 (19%)[c]	3 (9%)[c]	20 (24%)
Any muscle bleed in the past 3 or 12 months, n (%)	31 (26%)[b]	8 (23%)[b]	23 (27%)
Higher education level[e], n (%)	77 (60%)	23 (51%)	54 (64%)

Abbreviation: IQR = interquartile range.
[a]unknown for 1 patient.
[b]unknown for 10 patients.
[c]unknown for 11 patients.
[d]Different time intervals for joint or muscle bleed occurrence were reported for adolescents and young adults: the past 3 months for adolescents, and the past 12 months for young adults.
[e]For adolescents, the higher education level includes current participation in: senior general secondary education (HAVO) and pre-university secondary education (VWO). For young adults, higher education refers to current participation in, or the highest degree that was obtained in: Bachelor and Master degree programmes at universities of applied sciences (HBO) and at research universities (WO); and doctoral degree programme.

adolescents aged 15–17 years. Of 84 young adults, 77 (92%) were ready, one was not ready (1%) and six did not know (7%).

3.3 | Associations of factors with transition readiness

In comparing adolescents who were '(almost) ready' for transition and 'not ready or unsure', age was slightly higher in the former (median (IQR) of 15 (14-16) years and 13 (13-15) years respectively), albeit non-significant. Other socio-demographic and disease-related factors did not differ significantly between groups, as shown in Table 2.

3.4 | Treatment adherence, health-related quality of life, and self-efficacy according to transition readiness

The median VERITAS-Pro treatment adherence scores among adolescents with severe haemophilia were worse in patients who reported not to be ready (17, IQR 9–29), compared to those who were ready for transition (11, IQR 9–16), indicating that patients who were ready for transition were more adherent (Table 3). This was largely due to a difference in the domain 'Planning' for which the median domain-specific scores were 19 (0-25) in the '(almost) ready' and 25 (25-47) in the 'not ready or unsure' group. Median standardized CHO-KLAT scores were similar in the '(almost) ready' group (86, IQR 78–88) and the 'not ready or unsure' group (81, IQR 70–89). Likewise, HSES scores were also similar (55 (50-58) vs. 53 (43-56)).

3.5 | Transition preparation

The transition preparation topics that were most frequently discussed with 45 adolescents were: other aspects of life (40; 89%), worries about haemophilia or its care (39; 87%), and knowledge of haemophilia (33; 73%), as shown in Figure 1. Least frequently discussed topics were: which healthcare providers adolescents will meet in adult care (10; 22%) and available guidance in work/school issues (22; 49%). These most and least frequently discussed topics were similar among adolescents aged 12–14 years and 15–17 years. Among adolescents aged 15–17 years, nearly all adolescents using prophylaxis (91%; 11/12) indicated to have discussed self-administering treatment, compared to 25% (2/8) of adolescents not using prophylaxis (supplemental figure S1).

The aspects on which young adults would have liked to be better prepared were: information on who will be their adult healthcare providers (13/84; 15%), managing healthcare costs (10/84; 12%), and understanding their responsibilities in planning appointments (8/84; 10%), as shown in supplemental Figure S2. Dutch healthcare costs are explained in Box 2.

4 | DISCUSSION

In this study, we described the perceived readiness to transition from paediatric to adult care among adolescents and young adults with haemophilia in the Netherlands, assessed potential factors associated with transition readiness, and identified areas of potential improvement in transition programs. We found that transition readiness

TABLE 2 Associations of transition readiness with socio-demographic and disease-related factors among adolescents, and with treatment adherence, health-related quality of life, and self-efficacy.

	(Almost) ready ($n = 22$) N (%)	Not ready or unsure ($n = 23$) N (%)	RR	95% CI
Age in years; median (IQR)	15 (14-16)	13 (13-15)		
Age 12−14 years	10 (38%)	16 (62%)	ref	0.91-2.98
Age 15−17 years	12 (63%)	7 (37%)	1.64	
Type of haemophilia				
Haemophilia A	17 (46%)	20 (54%)	ref	0.72-2.58
Haemophilia B	5 (63%)	3 (38%)	1.36	
Disease severity				
Severe haemophilia	15 (56%)	12 (44%)	1.43	0.73-2.79
Non-severe haemophilia	7 (39%)	11 (61%)	ref	
Prophylaxis				
Yes	15 (56%)	12 (44%)	1.43	0.73-2.79
No	7 (39%)	11 (61%)	ref	
Family history of haemophilia				
Positive	18 (53%)	16 (47%)	1.45	0.62-3.39
Negative	4 (36%)	7 (64%)	ref	
Any hospital admission in the past 12 months				
Yes	3 (50%)	3 (50%)	1.00	0.42-2.36
No	19 (50%)	19 (50%)	ref	
Missing	0	1		
Any joint bleed in the past 3 months				
Yes	1 (33%)	2 (67%)	0.71	0.14-3.68
No	15 (47%)	17 (53%)	ref	
Missing	6	4		
Any muscle bleed in the past 3 months				
Yes	3 (38%)	5 (63%)	0.65	0.25-1.69
No	15 (58%)	11 (42%)	ref	
Missing	4	7		
Education level[a]				
Higher	12 (52%)	10 (45%)	1.15	0.63-2.10
Lower	11 (48%)	12 (55%)	ref	

Abbreviations: CI, confidence interval; IQR, interquartile range; RR, relative risk.

[a]Higher education includes current participation in senior general secondary education (HAVO) and pre-university secondary education (VWO). Lower education includes current participation in prevocational secondary education (VMBO), and vocational and middle management training (MBO).

increased with age during adolescence, and nearly all post-transition young adults reported they were ready for transition. Aside from the VERITAS-Pro treatment adherence domain 'Planning', no other factors were found to be clearly associated with transition readiness. Still, young adults would have liked to get better acquainted with the adult treatment team prior to transitioning, and get better prepared for managing healthcare costs and planning and keeping appointments.

To interpret whether observed differences in median scores between the ready and non-ready groups may be clinically relevant, we considered the 'minimum clinically important differences' (MCID). The MCID indicates the smallest difference which is of clinical benefit to patients, taking both the magnitude of improvement and the value patients' place on the change into account.[31] In an international study among boys aged 7−18 years with haemophilia, the MCID of the CHO-KLAT was found to be 6.5.[32] Our observed difference in the median total CHO-KLAT scores (5.3) suggests that there is no clinically important association between health-related quality of life and transition readiness. For the HSES, no MCID has been determined, although our observed difference (2.5) is unlikely to indicate a meaningful

TABLE 3 Treatment adherence, health-related quality of life, and self-efficacy according to transition readiness in adolescents.

	(Almost) ready	Not ready or unsure	*p*-Value	Difference in median scores
Standardized VERITAS-Pro, total score[a]; median (IQR)	11.5 (9.4-15.6)	17.2 (8.9-27.9)	.17	5.7
Domain; Time	18.8 (3.1-28.1)	15.6 (0-23.4)	.45	3.2
Domain; Dose	0 (0-6.25)	0 (0-12.5)	.72	0
Domain; Plan	18.8 (0-25.0)	25.0 (25.0-46.9)	.02	6.2
Domain; Remember	12.5 (3.1-31.3)	15.6 (1.6-25.0)	.73	4
Domain; Skip	3.1 (0-7.8)	0 (0-12.5)	.52	3.1
Domain; Communicate	21.9 (6.3-32.8)	25.0 (0-75.0)	.36	3.1
Number of adolescents with non-adherence; n (%)[a]	0 (0%)	1 (4%)	NA	NA
Standardized CHO-KLAT score; median (IQR)	85.9 (78.4-88.0)	80.6 (70.0-89.2)	.19	5.3
HSES score median (IQR)	55.0 (50.0-58.0)	52.5 (43.0-55.8)	.25	2.5

Note: For standardized VERITAS-Pro scores, lower scores indicate better adherence. For standardized CHO-KLAT scores, higher scores indicate higher health-related quality of life. For non-standardized HSES scores, higher indicate a greater self-efficacy.
Abbreviation: IQR, interquartile range.
[a]Non-adherence was defined as a standardized VERITAS-Pro score of 34 or higher. The six VERITAS-PRO domains are: Time' (the adherence to the prescribed schedule), 'Dose' (the adherence to the prescribed dosage), 'Plan' (treatment planning and inventory management skills), 'Remember' (to not forget infusions), 'Skip' (to postpone or skip infusions), and 'Communicate' (to contact the haemophilia treatment centre if needed).

BOX 2 Healthcare costs in the Netherlands[42]

The Dutch healthcare system is characterized by a compulsory healthcare insurance.
Different healthcare insurers act in competition, and are obliged to accept anyone who applies for the standard insurance package. People may not be charged differently, and pay a monthly fee of approximately €130. Haemophilia care is part of standard insurance, as are medication costs, except from a yearly deductible fee of approximately €400. This fee is paid for by individuals themselves.
Children under the age of 18 years are insured through their parents and do not have to pay extra. When children turn 18 years, young adults have to choose between different insurers, take out standard health insurance on their own name, and have to pay a monthly fee. People who are on a low income are entitled to a financial healthcare contribution.

association with self-efficacy. The VERITAS-Pro MCID was presumed to be 5 in a study among Dutch adults with severe haemophilia.[33] The VERITAS-Pro scores were better for the '(almost) ready' group compared to the 'not ready or unsure' group with an observed difference of 5.7, which may indicate a meaningful difference. Of its domains, the difference for 'Planning' (6.2) suggests a meaningful association between treatment planning/inventory management skills and transition readiness. Yet, it is hypothesized that some patients are deliberately not strictly adherent to their prophylaxis schedule and adjust infusions according to their activities, like sports.[25] So, even though the VERITAS-Pro considers this 'low adherence', it could be considered a sign of good treatment insight. Thus, VERITAS-Pro scores should be interpreted with caution. Still, since this ambiguity does not hold for the 'Planning' questions, we hypothesize that treatment planning and inventory management skills are meaningfully associated with transition readiness.

4.1 | Comparison to earlier evidence

An interdisciplinary Canadian Delphi study among haemophilia care providers reported similar conclusions, and found that the most important indicators of a successful transition are: adherence and skills including ordering medication, self-infusing and planning appointments.[34] Additionally, a strong patient-professional relationship is conditional for good adherence.[35] Two other studies have reported on transition programs in haemophilia care. In a Dutch questionnaire study among adolescents, it was found that adolescents' illness-related distress did not differ before and after transition.[19] In a US questionnaire study among adolescents, 92% reported to be satisfied with the introduction of self-management skills.[20] The Canadian, US and Dutch transition programs are very similar.[36–38] Transition readiness among adolescents with haemophilia in our study (49% was (almost) ready) is similar to Dutch adolescents with any chronic condition (56%).[17]

The overall effectiveness of transition programs was reviewed by a 2016 Cochrane review.[21] Authors concluded that transition programs

slightly improve transition readiness, but do not affect health-related quality of life nor health status. In a 2017 systematic review with the same research question, it was found that 65% of studies reported positive effects on health outcomes, especially adherence.[12]

4.2 | Strengths and limitations

A strength of our study is the nationwide inclusion and the availability of a national transition protocol. Yet, several limitations are present. First, the study response rate of 46% was low, and was even lower for the questions concerning transition. Still, since the HiN6 cohort is very similar to the overall Dutch haemophilia population with respect to haemophilia severity and age, we expect this had limited effects on our results. Nevertheless, respondents might represent a subgroup with higher treatment satisfaction, adherence or better a priori self-management skills, resulting in potential selection bias and more positive findings. Second, participants were only asked if certain transition relation topics were *discussed* in outpatient clinic appointments. They were not asked whether these topics were *appropriately* addressed, nor to what extent they felt to have mastered these skills. Third, recall bias may have occurred, and may be more present among young adults who transitioned up to 7 years ago. Moreover, factors associated with transition readiness cannot be considered causal, since causal relationships cannot be established in a cross-sectional study. Finally, due to our small group size, potential differences between groups may not have been found. Since only few post-transition young adults reported to have not been ready for transition, we were unable to assess associations in this group.

4.3 | Practical implications

We recommend to continue to draft an individual transition plan based on the topics presented in Box 1, discuss with adolescents what aspects they find most challenging and/or important plan, and to place additional emphasis on the following aspects. Transitions programs could be further improved by facilitating personal introductions to the adult treatment team prior to transitioning (e.g. information evenings for adolescents); better explaining the practicalities of managing healthcare insurance as an adult; and emphasizing the skills required to manage and plan appointments. Focusing on treatment planning skills and inventory management skills (e.g. using a treatment diary) could further increase transition readiness.

5 | CONCLUSIONS

Self-reported transition readiness among adolescents with haemophilia in the Netherlands increased with age, and nearly all post-transition young adults reported they had been ready for transition. This may be ascribed to successful haemophilia-specific transition programs. Still, several areas of improvement were identified. We found that transition readiness was associated with better treatment adherence, especially treatment planning and inventory management skills. These insights may help to further improve and personalize transitioning.

AUTHOR CONTRIBUTIONS

S.C. Gouw coordinated the research project and supervised data collection. M.R. Brands analysed the data. M.R. Brands and S.C. Gouw drafted the manuscript. All authors provided feedback on the manuscript and approved the final manuscript.

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CONFLICT OF INTEREST STATEMENT

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DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

The study was conducted in accordance with the Declaration of Helsinki (version 2008, October 2008, Seoul, Korea). Ethical approval was obtained from the Institutional Review Board of the Leiden University Medical Centre (NL59114.058.17).

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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