

AIDS and Haemophilia

A Study among Dutch Haemophiliacs on the Psychological Impact of the AIDS Threat, the Prevalence of HIV Antibodies and the Adoption of Measures to Prevent HIV Transmission¹

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Abstract. More than 75% of all Dutch haemophiliacs participated in a mail survey that included a section on acquired immunodeficiency syndrome (AIDS). Although no cases of haemophilia-related AIDS had been reported in the Netherlands at the time of our survey, the threat of AIDS had caused considerable anxiety among Dutch haemophiliacs. This had led almost a third of the patients to change their therapy schedules. These changes were mostly not beneficial and possibly counterproductive, since they were not likely to reduce the risk of human immunodeficiency virus (HIV) infection. Measures that were advised to prevent HIV transmission, such as the use of condoms, were seldom carried out, even by the 17% seropositive individuals. We conclude that an increase in the informational efforts towards the patients is called for to avoid ill-advised treatment changes and to enhance preventive behaviour.

Introduction

In 1982 it became apparent that the acquired immunodeficiency syndrome (AIDS) can be transmitted by blood transfusions [1]. The first case of AIDS in a haemophiliac was reported in 1982 in the United States and since then more than 400 cases among haemophiliacs have been reported worldwide

[2-5]. Patients with haemophilia have been at high risk of contracting the human immunodeficiency virus (HIV) by infusions of coagulation factor preparations made from human blood. Recently, steps have been taken to reduce this risk: donors are asked to with-

¹ Grants were received from Het Praeventiefonds (No 28-1099) and De Stichting Haemophilia.

draw voluntarily if they belong to one of the risk groups for AIDS, blood donations are screened for antibodies to HIV and clotting factor concentrates are heat-treated. Since 1985, the Central Laboratory of the Dutch Red Cross also supplies heat-treated cryoprecipitate. The introduction of these measures has drastically reduced the risk of infection [6, 7], but unfortunately a number of haemophiliacs, in some countries up to 80%, has already been infected [8–12].

The availability of serologic tests for anti-HIV antibodies made it also possible to examine individuals from the risk groups on request or for medical or research reasons. The drawbacks of the test in an asymptomatic individual have been pointed out in several reports. The consequences of seropositivity are still uncertain [13] so that serious anxiety and stress may even be the only result. Stigmatization may evolve, with difficulties regarding occupation, schooling, insurance and social contacts [14, 15].

Since no therapy or vaccine against AIDS is available, the only possible action is prevention. In addition to the measures taken in the production of blood products, the haemophilia treatment centres in the Netherlands and elsewhere advocated a number of changes in individual therapy. At the same time the need to maintain adequate treatment was strongly emphasised. Haemophiliacs who are HIV carriers may transmit the virus to their sexual partners [9, 16, 17]. In order to reduce the risk of HIV transmission preventive measures were advised. The treatment centres and the Dutch Haemophilia Society cooperated to educate the patients on these issues and about AIDS in general. The treatment centres did this by organising meetings with their patients as well as informing them individually, while the pa-

tients' organisation regularly wrote about AIDS in its magazine and edited a brochure about AIDS [18].

The aim of our survey was to study the impact of the threat of AIDS on Dutch haemophiliacs. We compared the seropositivity rate in the Netherlands with other countries, and studied the attitude of the patients towards the test. Furthermore, we collected data on changes in therapy made both in accordance with and in contrast to the advice given by the haemophilia treatment centres, in relation with anxiety caused by AIDS. Finally, we examined compliance with measures to prevent HIV transmission.

Methods

The data were collected by standardised mail questionnaires sent to all 1,162 Dutch haemophiliacs who were registered at one of the treatment centres or the Dutch Haemophilia Society [19]. The questionnaire covered a broad range of topics concerning haemophilia, including AIDS. Closing date was January 1st, 1986. Forms from female carriers were included only when they had symptomatic haemophilia and had needed replacement therapy in the preceding year. Questionnaires that were completed inadequately were excluded. Data on type and severity of haemophilia were supplemented by information from the treatment centres.

The haemophilia treatment centres had agreed on the following adaptations in haemophilia therapy to reduce the risk of HIV infection: reevaluation of the need of prophylactic substitution therapy in all patients and a change to on demand treatment if possible; postponement of elective surgery until the availability of (proven) safe blood products; preferential use of desmopressin for patients with moderately severe or mild haemophilia A. The patients had been strongly advised, however, not to change their treatment regimens themselves. To prevent transmission of HIV to sexual partners or others the patients had been advised to use condoms in sexual intercourse, to postpone having a family and not to share toothbrushes or shaving utensils with other members of the

household. Patients on home treatment had (again) been instructed to use special waste disposal containers for used materials that might be contaminated. Parents assisting in the home treatment of their children had been advised to wear gloves when administering transfusions.

Seven items with a five-point scale were used to measure the psychological effect of the AIDS threat. A combined scale of 'AIDS-related anxiety' was constructed by scoring each item 0 to 4 points and adding the scores for each patient. With regard to the anti-HIV test we asked the patients whether they had been tested and whether the test result was known to them. The individuals who had not been tested could indicate whether they wished to be tested in the near future or not. We asked the patients to estimate the risk of a seropositive individual to develop AIDS, on a seven-point scale ranging from 'less than 5%' to '100%'. A list of seven items was included where patients could indicate any change in treatment because of AIDS. Information about current treatment regimens was obtained in the general medical section of the questionnaire. A list was included of the measures that had been advised to prevent HIV transmission where the patients could indicate whether they carried out these measures 'always', 'sometimes' or 'never'. We calculated compliance figures for each measure, for the patients to whom each measure applied, by only accepting 'always' as carrying out a measure in an effective way.

Statistical analysis was performed by using χ^2 -tests with Cramer's V as an indicator for the strength of relationships. Also used were product-moment correlations tests (Pearson's *r*). Significance was accepted at the 0.01 level. All results were analysed separately for severe, moderately severe and mild haemophilia, to avoid spurious relationships.

Results

Nine hundred and forty-seven (81%) out of the 1,162 questionnaires were returned, of which 12 had to be excluded from further analysis. The survey population of 935 patients included 801 patients with haemophilia A (86%), 132 with haemophilia B (14%), while 2 individuals had factor VII

and factor XIII deficiency, respectively. All combined, 384 (41%) had severe haemophilia (less than 1% clotting activity), 175 (19%) had moderately severe haemophilia (1-5% clotting activity) and 376 (40%) had mild haemophilia (more than 5% clotting activity). Substitution therapy consisted of cryoprecipitate (38%), Dutch factor VIII concentrate (18%), Dutch prothrombin complex concentrate (9%) and imported concentrates (3%), while 25% of the patients indicated not to receive substitution therapy regularly and 6% did not know the name of the product used. Six percent of the 484 patients with moderately severe or mild haemophilia A had received treatment with desmopressin (either exclusively or in combination with plasma products) in the year preceding our survey, while almost a third of them had received transfusions with plasma products during this period.

AIDS-related anxiety was significantly higher among patients with severe haemophilia than among those with moderately severe or mild haemophilia, but anxiety was frequently mentioned by these patients, too (table I). Sixty-five percent of all 935 patients mentioned preoccupation with AIDS, 44% a sense of lost future perspective, 31% feelings of depression, 21% feelings of tenseness, while 12% reported insomnia and 11% hypochondria. Social repercussions, i.e. being avoided, were mentioned only rarely.

Two hundred and seventeen out of the nine hundred and thirty-five patients had been tested for HIV antibodies and knew the result, while 90 had been tested but had chosen not to be informed about the result. The proportion of tested patients was similar for haemophilia A and haemophilia B. More than a third of all patients with severe haemophilia had been tested, as compared to 23

Table I. AIDS-related anxiety

	Respondents by severity, %		
	severe (n = 384)	moderate (n = 175)	mild (n = 376)
I think that I have AIDS or symptoms of AIDS	17	7	7
The future looks grim because of AIDS	55	49	31
It makes me feel depressed	40	33	21
I cannot sleep because of it	20	11	5
I feel tense	33	21	9
AIDS is on my mind	80	65	51
Other people avoid me	4	3	2

Anxiety scores by severity: $r = 0.27$; $n = 935$; $p < 0.001$. Indicated are those respondents who answered 'sometimes', 'regularly', 'often' or 'always'.

and 11% for moderately severe and mild haemophilia, respectively (table II). The overall seropositivity was 17% (36/217), with the highest prevalence of HIV antibodies among patients with severe haemophilia (21%). Seropositivity in haemophilia A and haemophilia B did not differ significantly.

Most patients were aware of the limited prognostic value of the anti-HIV test: only 7% out of 935 patients thought that seropositivity carried a 100% risk of developing AIDS, while almost half of the patients thought this risk to be less than 15%. More than 50% of the 628 patients who had not been tested indicated that they did not want to be tested. Many argued that they deliberately chose not to know whether they were seropositive or negative (35%), while particularly many of the patients with mild haemophilia indicated that they were sure to be seronegative (33% of all, 47% of those with mild haemophilia). We found no different anxiety levels between patients who wanted

to be tested in the near future and those who did not. The seropositive patients had the highest anxiety scores (mean score 5.6, $n = 36$). Surprisingly, the seronegative patients had an anxiety level that was slightly higher than that of the untested patients (mean scores 3.6 and 3.1 for severe haemophilia, $n = 107$ and $n = 183$, respectively), although this difference was not statistically significant.

Seven hundred and twenty-three patients received treatment regularly and 207 (29%) of them reported a change in their therapy schedules because of AIDS, evenly distributed over severe, moderately severe and mild haemophilia. Some patients reported several changes (table III). The majority (63% of 207 patients) reported ill-advised changes: waiting longer before treating a bleeding, using smaller doses (or the combination of these) or the complete abandonment of all treatment. The seropositive patients reported as many changes as the other patients. The patients who reported to wait

Table II. Test of HIV antibodies

Test group	Tested subjects	Seropositive, %	Untested subjects
Haemophilia A, severe	114	21	203
Haemophilia B, severe	20	20	45
Haemophilia A, moderate	34	15	116
Haemophilia B, moderate	7	0	18
Haemophilia A, mild	37	5	297
Haemophilia B, mild	3	0	39
All	217	17	718

Included among untested subjects are 90 patients who had been tested, but did not know the result of the test, included in the test group 'all' are the 2 persons with factor VII and XIII deficiency

Table III. Changes in therapy because of AIDS

	Respondents by severity, %		
	severe (n = 376)	moderate (n = 164)	mild (n = 183)
Stopped prophylaxis ¹	13	3	–
Postponed elective surgery	1	4	10
Changed to DDAVP	–	2	4
Changed to other blood products	7	2	2
Waited longer before treatment	14	20	15
Used smaller doses ²	7	–	–
Stopped all treatment	1	2	2

Patients who received treatment regularly were studied

¹ Proportion of patients on prophylaxis n = 183 and n = 18, for severe and moderate haemophilia, respectively

² Proportion of patients on home treatment n = 259 and n = 39, for severe and moderate haemophilia, respectively

longer had a significantly increased delay between the onset of a bleeding and the administration of a transfusion ($\chi^2 = 37.9$; 5 df; $V = 0.24$; $p < 0.001$). For patients with severe haemophilia on home treatment the average delay was 3.4 h for those patients who now waited longer, compared to 1.9 h

for those who did not; for the patients not on home treatment the corresponding delays were 4.9 and 3.1 h, respectively. Some patients reported a delay of more than 12 h. A similar trend existed for moderately severe and mild haemophilia (3.9–5.7 and 5.1–8.1 h, respectively). The patients who had made

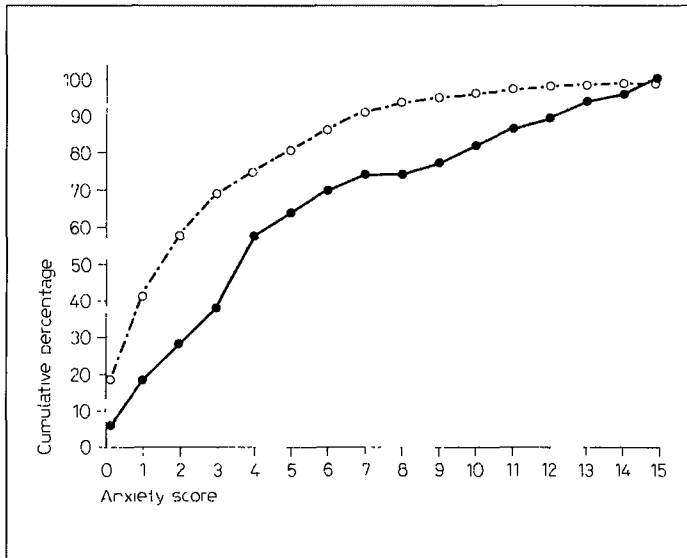


Fig. 1. Ill-advised changes in treatment and AIDS-related anxiety. Anxiety score for cumulative percentage of patients with severe haemophilia who made ill-advised changes in their therapy schedule (●, $n = 66$) versus those who did not make these changes (○, $n = 318$) ($r = 0.25$; $p < 0.001$).

ill-advised changes had significantly higher anxiety levels than the patients who had not made these changes (mean scores 5.5 and 3.1, for severe haemophilia, $n = 66$ and $n = 318$, respectively) (fig. 1).

In table IV we provide the data on measures to prevent HIV transmission. The great majority of the patients who were married or had a steady relationship did not use condoms, even if they were seropositive. None of the parents assisting their children in home treatment used gloves; several parents informed us that they thought the use of gloves to which they were not accustomed would increase the risk of needle pricking accidents. Only a minority of the patients on home treatment used special containers for possibly contaminated waste materials. Although the compliance with these measures was generally low, patients with severe haemophilia and the seropositive individuals complied significantly more often than the others. We also found that patients who con-

sidered themselves well-informed about AIDS were more likely to use condoms ($\chi^2 = 14.3$; 4 df; $V = 0.17$; $p < 0.01$). The compliance with measures to prevent HIV transmission was slightly higher among patients with more AIDS-related anxiety (controlling both for severity of haemophilia and the result of the test on HIV antibodies, use of condoms: $r = 0.10$, $n = 505$, $p < 0.05$; postponement of pregnancies: $r = 0.33$, $n = 80$, $p < 0.005$; use of containers: $r = 0.14$, $n = 302$, $p < 0.01$).

Discussion

The prevalence of haemophilia can be estimated at 7–9/100,000 [20, 21], which implies that we sent questionnaires to 90% of all Dutch haemophiliacs. This figure and the high response rate make us confident that our survey presents a reliable overview. Since only part of all patients had been

Table IV. Compliance with preventive measures

Measures	Respondents by severity and test status, %				
	severe	moderate	mild	seropositive	all
Use of condoms ¹	16	4	4	32	9
Postponement of pregnancies ²	50	13	0	(50)	16
Use of gloves ³	NC	NC	NC	0	0
Containers ⁴	NC	NC	NC	44	39

NC = No classification by severity of patients on home treatment.

¹ Use of condoms by patients who were married or had a steady relationship ($n = 505$) (severity $\chi^2 = 22.8$, 2 df, $V = 0.21$, $p < 0.001$, seropositivity $\chi^2 = 15.4$, 1 df, $V = 0.17$, $p < 0.001$)

² Postponement of pregnancies by couples who wanted children, patients married and younger than 40 years ($n = 80$) (severity, $\chi^2 = 13.1$, 2 df; $V = 0.40$, $p < 0.001$; seropositivity, this group consisted of only 2 individuals)

³ Use of gloves by parents assisting in home treatment of patients of 12 years and younger ($n = 50$).

⁴ Use of waste containers for sharp materials by patients on home treatment ($n = 302$) (seropositivity, not significant)

tested for HIV antibodies, however, the figures on seropositivity should be viewed with some caution. In severe haemophilia a fairly large sample of one third of the patients had been tested and the reported seropositivity rate does not differ much from that reported by Breederveld et al. [22] who found 19% seropositive individuals in a prospective study on 166 patients. Therefore, the number of seropositive patients in the Netherlands is low compared to the data from France (50%), Germany (53%), the United Kingdom (39%) and the United States (80%) [8–11]. It is higher, however, than the 5% seropositivity among Belgian haemophiliacs [12]. The low number of seropositive individuals in the Low Countries can be explained by the predominant use of plasma products from local nonpaid donors, and an early and effective voluntary self-exclusion of donors belonging to AIDS risk groups [Van der Poel, unpubl. observations]. This small use of commercial concentrates may

also explain the absence of a difference in the seropositivity rate in haemophilia A and B. Even though at the time of our study no cases of AIDS and only few cases of AIDS-related complex had been reported in Dutch haemophiliacs [4], the threat of AIDS has hit hard and caused much fear among haemophilia patients. The feelings of optimism that have surrounded the treatment of haemophilia during the last decades have made the impact of this sudden death threat only more devastating.

A third of the patients chose not to be tested for HIV antibodies. In the Netherlands, public health authorities have pointed out the drawbacks of the test [23], which evidently has had its influence on the population of haemophiliacs. We feel that considerations of public health or the protection of health care workers do not justify screening these patients for HIV antibodies, since the risk of nosocomial HIV transmission is extremely low [14, 17, 24–26]. One of the rea-

sons an individual may have for choosing to be tested is the hope for a negative and thus reassuring result. In patients with a low risk of seropositivity, e.g. those with mild haemophilia, one might offer the test for reasons of reassurance. It is obvious that patients need guidance and information when the test result is positive; however, our results indicate that this is also necessary for patients proven to be seronegative, since, surprisingly, their anxiety scores were not lower than the scores of patients who had not been tested.

Fear of AIDS has led to changes in therapy schedules that are mostly not beneficial and might also be considered counterproductive: use of smaller doses or waiting longer before treating a bleeding could well lead to an increase in the number of transfusions required. In contrast to the advice of the treatment centres to use desmopressin when appropriate, only very few of the patients had been treated with it. Even though new HIV infections from plasma products seem very unlikely, the risk of virus transmission by using human blood will remain. Therefore, we feel that a more widespread use of desmopressin is desirable.

The compliance with the advice for preventive measures is low, in spite of the information supplied by the haemophilia treatment centres and the Dutch Haemophilia Society. This finding has also been reported in two recent reports from the Netherlands and elsewhere [17, 27]. Presently, efforts should be aimed at the prevention of HIV transmission by infected haemophiliacs to their sexual partners and others. It is of vital importance that all haemophiliacs are instructed about the risk of HIV transmission and about the appropriate preventive measures. Only those known to be seronegative

may adopt a more liberal attitude towards these measures. The scarce use of condoms in a group that is clearly at risk of transmitting the virus, and that has received ample information about preventive behaviour justifies concern about the success of attempts to educate the heterosexual population at large about AIDS prevention.

Patients with moderately severe and mild haemophilia showed considerable anxiety and made just as many changes in their therapy schedules as patients with severe haemophilia, although their risk of infection is much smaller. Since most of these patients do not visit their treatment centre as often as patients with severe haemophilia, it will be difficult to reach them with information about AIDS other than through the popular press.

Future policy should be aimed at maintaining a high standard of treatment for haemophiliacs and prevention of further spread of HIV infection. The possible danger and uselessness of ill-advised changes in treatment and the need of practising safe sex will have to be emphasised again. We feel that the national patients' organisations and the World Federation of Hemophilia will be able to play an important role in this, but the treatment centres will have to make additional efforts to reach the nonmembers. Seropositive children entering adolescence deserve special attention. The further spread of AIDS among the population at large, the recent reports of an increasing risk of developing AIDS when seropositive and the great interest for AIDS in the press may well cause more anxiety among haemophiliacs. The only way to respond to this and to achieve the aforementioned goals is a further increase in information about AIDS towards haemophiliacs.

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