Coping with disability and co-morbidity: a patient's perspective

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Changing perspectives

In 56 years of living with severe haemophilia I have experienced a number of remarkable transitions in haemophilia health care. The first transition was the one from no treatment at all for haemophilia to treatment becoming available. This marked the start of the best of times for haemophiliacs of my age: the period from the mid-sixties of the last century to the early eighties. My perspective changed from the prospect of a difficult life with a lot of pain to the expectancy of a near normal life.

The worst of times came in the early eighties, when it became clear that recipients of blood and blood products, and especially haemophiliacs, were among the first known high risk groups for contracting HIV.[1] Later infection with the Hepatitis C virus also was seen to have serious and even life threatening consequences. When I found that I was indeed infected with HCV and HIV my life's horizon was shortened once more. The future brightened again as HAART therapy for HIV became available and even more so when antiviral therapy led to the disappearance of my Hepatitis C virus. Life expectancy was back to normal once more. And now I have actually reached that part of the human life cycle where I really am of an advanced age.

Persons with haemophilia as well-educated professionals

People with haemophilia, their partners and their families are mostly perceived as well-educated managers of their own disease. Nowadays in the Western world a child or young adult grows up with the availability of modern blood clotting products. Patients or their parents can administer these products intravenously at home on an 'on-demand' basis or on a 'prophylactic' schedule. This development led to an enormous improvement in quality of life for haemophiliacs.

The main difference with the older haemophiliacs is, however, that the younger generation of haemophilia patients has little experience with spontaneous and rare bleedings. And that their haematologists and haemophilia nurses also have no direct knowledge from the period that bleedings were quite common. The physicians from the early days of haemophilia treatment are growing older. Some of them have retired and some resigned because they blamed themselves for the viral transmission in their patients.

For the older haemophiliacs this implicates that he is the well-educated professional who now has to deal with a new generation of caregivers largely unfamiliar with his medical and social past and the old 'typicalities of haemophilia'.

My own haemophilia career

Due to the lack of treatment for haemophilia I spent a lot of time in hospital during the first twenty years of my life. About ten of these twenty years I lived in a hospital bed. In spite of this I never experienced that period in my life as a heavy burden. Contrary to general hospital policy in those days my friends were allowed to visit me. So like other youngsters, I played - whenever possible – with other children and enjoyed life at school. It was much later in life that I realized the exceptional and rather personalized care I had received from my physician and especially from the matron of the hospital unit where I stayed most of the time. Like my parents, who always stimulated me to be as independent as possible, this matron was also concerned about my long periods of absence from school and friends. During my final high school year I even had permission to receive the whole class and teachers in the hospital to prepare for school examinations. And when other non-haemophilia patients complained about my apparent preferential treatment, she told them that this was a necessary part of my medical treatment as well. Although she would never have put it that way, she perceived ‘a patient as more that his illness’.

From my early years in hospital I developed a strong aversion to the regular vena-punctures needed for the transfusion of clotting factor or – before that – fresh frozen plasma. Like many other haemophiliacs I developed ‘needle phobia’. At the age of fifteen I saw a psychologist to reduce this fear, however with little effect. It all disappeared the day that one of the physicians suggested that I should try to transfuse myself. It immediately went well and I have injected myself ever since. The phobia returns whenever I have to return for routine checks and blood sampling. However, I now have a special card indicating that I may and can inject myself. At the 2007 Global Nurse Symposium in Berlin a young adult with haemophilia from the UK told a similar story. The only difference was that his doctor suggested it when he was only three and a half years old!

A changing perspective

In the early seventies I left school and went to university in another city. Quite soon I experienced that the approach to which I was used, was not that common. After two years of negative experiences I finally found a hospital that offered a more personal contact between hospital staff, the patients and their parents.

At that time the Dutch National Haemophilia Patient Society (NVHP) was founded and from the United States the concept of home-treatment was imported. I was among the first patients on home-treatment. The future looked bright. Half joking, half serious the haematologist told my mother that she could stop being concerned about my health. He did not expect me to hospitalization for the next 25 years. Like all other men without haemophilia the biggest risk of hospitalization for me would be due to heart problems or other symptoms seen in hard working career persons. And indeed, I did not need hospital admission for the next 30 years. So in terms of ‘quality of life’ issues the perspective of haemophilia certainly has changed.

Co-morbidity

So this haematologist was right in his prediction. But what neither of us could have foreseen at the time were the consequences of viral infections. Morbidity and mortality from HIV and HCV became the dominant issue in the haemophilia population in the eighties and nineties. I have the fortune to be part of the one-third of HIV infected haemophiliacs with an infection survival of more than 25 years. It also was my fortune that the first manifestations of infection appeared when antiviral therapy had just been introduced. And by the time the complications became more serious, HAART therapy was available. I have needed hospital treatment for HIV twice. One episode was rather serious and had a tremendous impact on my life and that of my partner. While on holiday in France in 2001 I suffered acute renal failure due to one of the HIV inhibitors, Indinavir®. I became quite ill and needed about two weeks of intensive care at the hospital in Chalon-sur-Saone. Although I recovered rather well my general condition had deteriorated substantially. Due to the already existing bad condition of my knees and ankles a knee replacement seemed to be the next stop. However, thanks to intensive physical therapy and functional gait training in the swimming pool this could be avoided. My wife advised me to buy a tailor made cycle and so I further improved my condition through intensive cycling. All in all the developments of the last twenty five years have reduced the importance of haemophilia as a disease. For me the consequences of HIV infection have much greater significance. My greatest concern is the long-term effects of taking HIV inhibitors everyday and the progression of the infection.

Growing older

I entered into a long-lasting relationship rather late in life so I was used to managing my own haemophilia. I deal with home-treatment without help. Fellow patients have told me that their wives assist them or even inject them. I now realise that self-treatment may become a problem for me in the future, but I will continue it for as long as possible.

Shopping is one of the things older haemophiliacs do not like because of their severe arthropathy. Our weekly shopping expedition generally takes over two hours of constant walking and standing. There have been periods that my condition did not allow me to accompany my partner, but at the moment that is not a problem.

Things become more complicated when one’s partner falls ill. My wife once was unfortunate enough to break a leg. For three months I had to take care of the shopping, housekeeping and daily meals. The mere fact that I have haemophilia did not entitle us to home help services. We managed without, but it was very exhausting.

Because of my initial bad life expectancy, which did not improve in the period of the viral infections, I hardly gave matters like old age pension and financial future a thought. I never expected to reach retirement. When I was young my parents stressed the importance of good education in order to obtain a proper white-collar job. But once I entered university many things besides my study, e.g. the foundation of the Dutch National Haemophilia Patient Society, kept me from my studies. I gradually became the eternal student. With the occurrence of HIV, the last thing I worried about was my pension.

Unlike many other people with HIV I continued and could continue working. So I accumulated a small pension. I also received some financial compensation for HIV infected haemophilia patients through the Dutch government. With the money I bought my house. Since I have a partner, I feel a certain pressure to think about my pension. The only thing I have arranged is securing the house for my wife in case something happens to me.

Compliance

I absolutely comply with the necessary medical therapy for haemophilia and HIV. I am more than motivated to do physical therapy and exercise (swimming and cycling). Thanks to these activities I have avoided a planned knee replacement. I also strictly abstain from using alcohol, tobacco or other drugs. More or less from the moment I knew of my own HIV and HCV risk I gave up alcohol.

The perfect patient! That remains to be seen. I have to admit that I absolutely do not comply with the rules on registering home infusions and I definitely have a tendency to avoid my dentist. I was also very reluctant to start antiviral therapy for HCV. So, I am not the ideal patient who always meekly follows every medical advice.

The quintessence of this long personal story is that in more than fifty years of life, I became a person with multiple chronic diseases and with a broad range of health care contacts. It's my belief that although this is my own personal story, it is comparable with the experiences of many other people with chronic disease and co-morbidity. Details may be different, but the general picture will remain the same.

Three problems with co-morbidity

Three problems worry me specifically. The first problems is the lack of co-ordination between medical specialists, the second is polypharmacy – the use of multiple medications –and as a consequence of these first two problems a third occurs which I prefer to address as the 'fear' factor.

About polypharmacy, all in all I probably do not take that many different medications. But I already have experienced quite a few problems with the so far unexplainable complications occurring due to the interference of my HIV medication and haemophilia, predominantly bleeding complications. My acute renal failure was due to one of the HIV inhibitors.

Enter a third medical discipline not familiar with these two diseases and a disaster is waiting to happen. In such situations I – as a patient – have experienced a lack of co-ordination, willingness and time to discuss things before acting. Some specialists primarily act on what they perceive as the medical problem. After a number of these experiences as a patient I automatically developed the 'fear' factor.

The ‘fear’ factor

Like other persons with severe haemophilia I have a special type of fear. The ‘fear’ of a lack of co-ordination, a lack of control when you are hospitalized, when you need medical treatment from physicians who have no experience with haemophilia, or when you are involved in a serious traffic accident.

In all these cases you, who are a well-educated manager of your own disease, may not be able to influence or check the treatment you receive. The ambulance may take you to a hospital that has a bed available, but no experience with haemophilia, and then valuable time may be lost.

Part of the ‘fear’ of older people with haemophilia is that they are often not in good physical shape due to orthopaedic or viral complications. So when, for instance, minor or major surgery is proposed we not only fear the operation, but the consequences of this operation for total body functioning. To us our state of health is a delicate balance. ‘Vulnerable’ actually may be a better term. At the end of this chapter practical advice on how to deal with fear is given.

Improving health care settings for people with co-morbidity

In recognition of the expected growth of the number of people with haemophilia and co-morbidity and the problems that already exist, a way to adjust the health care system to the needs and wishes of patients, especially older patients, should be found. In a study NIVEL, RIVM and AMC discussed the co-ordination issue with several medical experts. They all held different opinions. At the end of the day no consensus for a solution had been reached.[2]

In the last couple of years I have regularly discussed the issue of co-morbidity and its co-ordination of care problems. One thing I have learned is that geriatric patients have problems which are similar to those perceived by people with co-morbidity; i.e. growing functional limitations, psychological symptoms and social and societal problems.

In rehabilitation, paediatric and geriatric care a holistic approach has been developed in which the patient is seen as a person in relation to his environment. Care encompasses not just the child, but the parents and family as well. This definitely compares with the way I grew up in the hospital setting where the matron regarded her patients as more than their illness. Although for haemophilia this concept already exists since the introduction of specialized comprehensive care centres, I wonder whether it is also possible to introduce a comparable hospital setting for patients with other multiple chronic diseases. This will be in sharp contrast with the way most hospitals are now organised, but it is worth the effort to start a number of experiments.

Of course, the approach cannot be too generalised. Co-morbidity problems are often so complex that highly specialized knowledge is absolutely necessary. A special clinical and outpatient facility for people with co-morbidity problems could be introduced.

References

[1] Rosendaal FR. *Haemophilia: the best of times, the worst of times*. Thesis. Leiden; 1988

[2] Heijmans MJWM, Rijken PM, Schellevis FG, et al. *Meer dan een ziekte: de gevolgen van comorbiditeit vanuit het perspectief van chronisch zieken en gehandicapten*. NIVEL/AMC/RIVM; 2003:78 pp.

How to avoid complications?
Practical advice for haemophilia patients with co-morbidity and their care givers

Advice for haemophilia patients with multiple chronic diseases and their partners may help to avoid come – maybe not all – complications. Useful suggestions from patients organisations or a nurse practitioner are mentioned below.

1. Become a member of one or more patients' organisations dealing with your major diseases, read their information materials and surf the internet.

2. It is advisable to stay in regular – at least once or twice a year – contact with medical and/or nursing staff at your haemophilia centre. Although there may be no acute medical reason, it is useful to know them and to be up-to-date informed about practical matters like changes in emergency telephone numbers.

3. Inform any physician or health care professional you have to deal with, about your haemophilia. Instruct them that your haemophilia treater should always be contacted to discuss the treatment regime. And for your own safety, personally call your specialist and check if everything is co-ordinated.

4. In case of emergency hospitalization or traffic accidents, always instruct your partner or family members that they should inform your haemophilia treater whenever you are admitted into another hospital. For these situations it is also sensible to wear a Medic Alert bracelet or pendant with your medical data and an up to date telephone number that can be contacted. Also file the telephone number of your haemophilia centre in your mobile phone. Carrying a USB stick - Medstick® - with your medical data is a modern sophisticated way to have all information with you. Your medical data can be available in various languages (Dutch, German, English, French) (www.cinsol.nl).

5. When you are admitted into another department that the one you're familiar with, be aware that the hierarchy of the hospital system may cause co-ordination problems. In most hospitals the doctor responsible for your daily care is a physician working in that particular department who may not be particularly familiar with your haemophilia and other diseases. Nurses and other health care professionals may lack experience in the daily treatment of your diseases. They may be surprised that you prefer to do some of the treatment, like intra-venous injections, yourself and cannot know that you are an expert in performing these.

   It is recommended to discuss this as soon as possible after admission and inform the physician in charge or the ward nurse that you are used to treating your own haemophilia. Also tell them in advance about any situations or eventualities that you are worried about.

6. It is always wise to determine in advance what you need to do in order to receive adequate medical care. It is advisable to tell your partner, friends or family members what you want them to do in case of emergencies. You may want to write down a short memo of these instructions as well.