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Haemophilia, aging and co-morbidity: an emerging problem

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Goal

To increase awareness of problems arising in elderly haemophilia patients



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- ” Life expectancy
- ” Treatment options
- ” Haemophilia related co-morbidity
- ” Non-haemophilia related co-morbidity
- ” Impact and quality of life

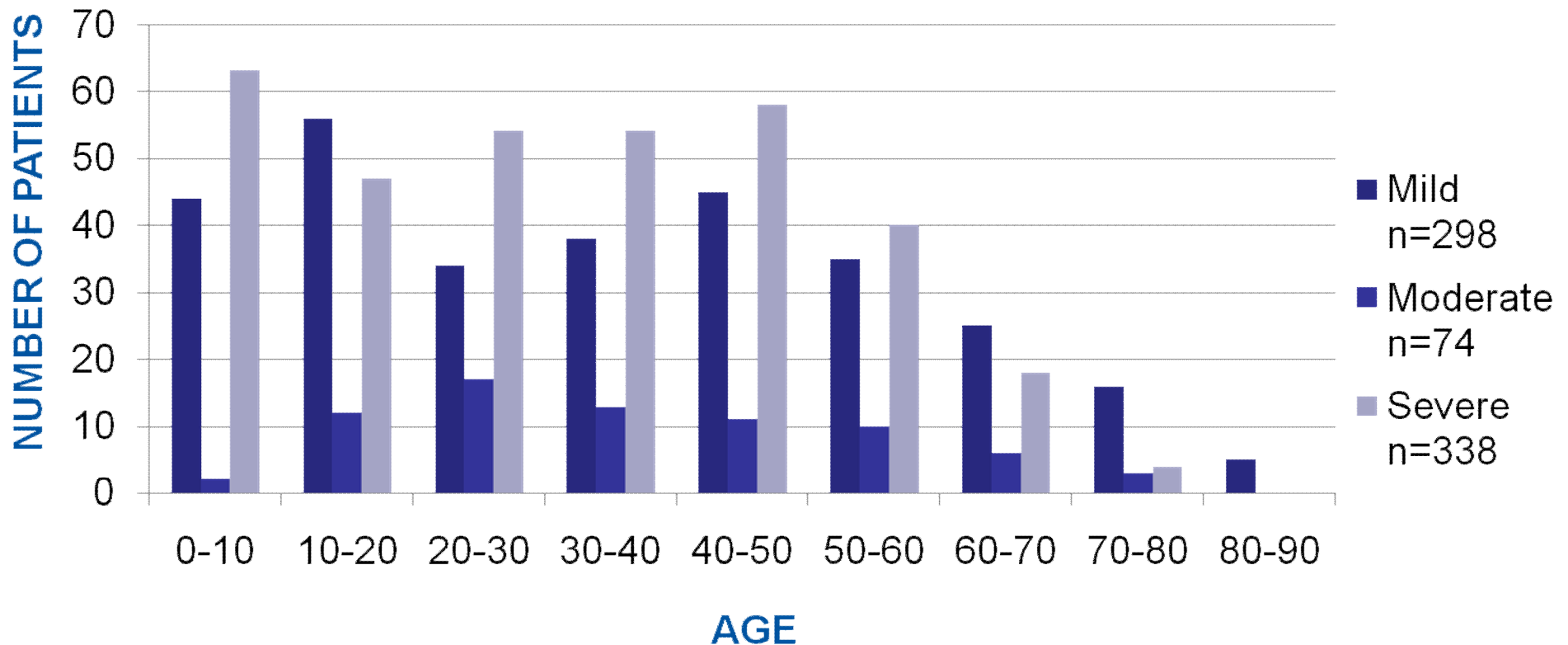


Life expectancy of haemophiliacs is increasing

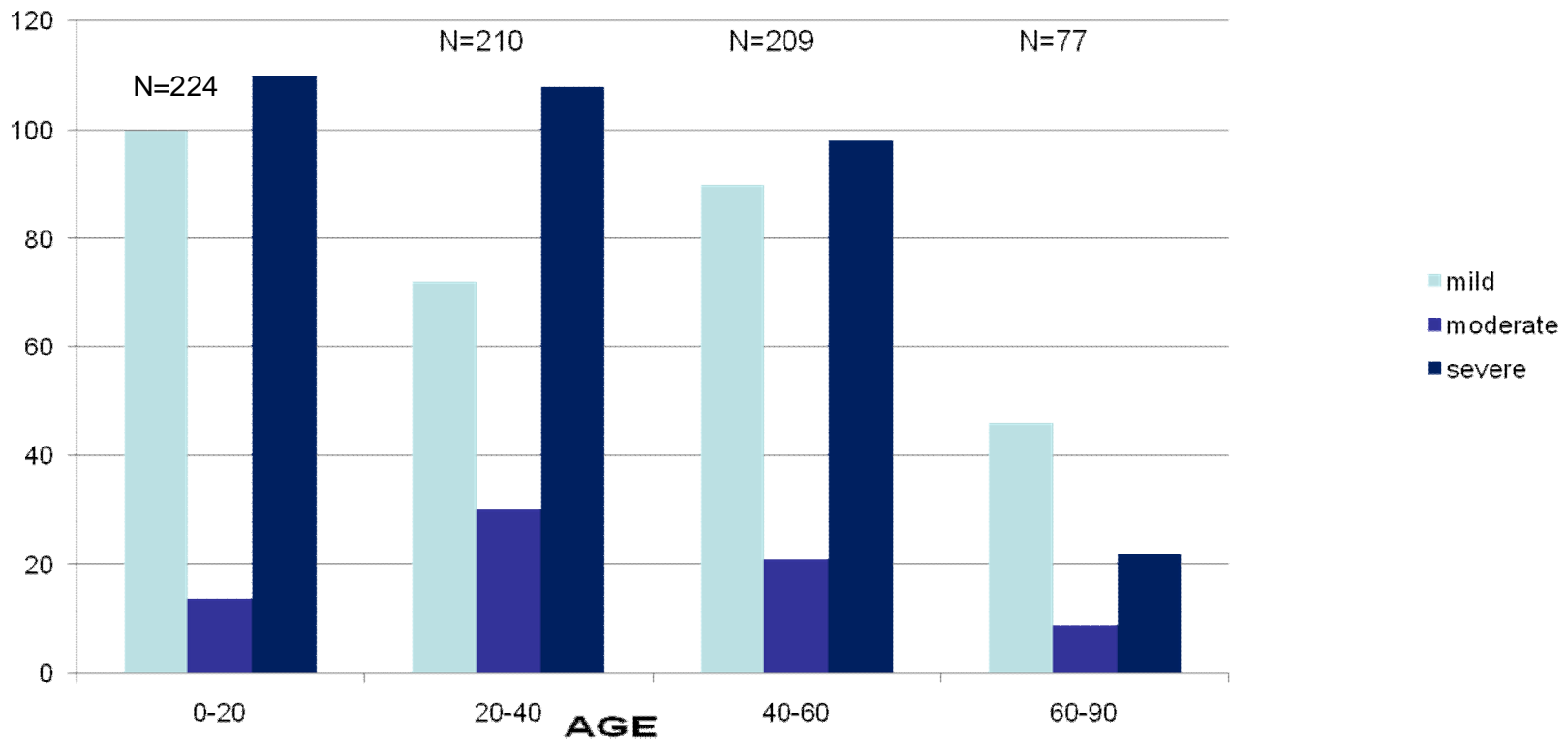
Life expectancy in years of Dutch haemophiliacs (I Plug et al)

	1985	1992	2001
Severe haemophilia HIV neg/HCV neg	63	61	59 71
Moderate HIV neg/HCV neg	65	65	67 75
Mild HIV neg/HCV neg		74	73 75
Dutch Males	71	74	76

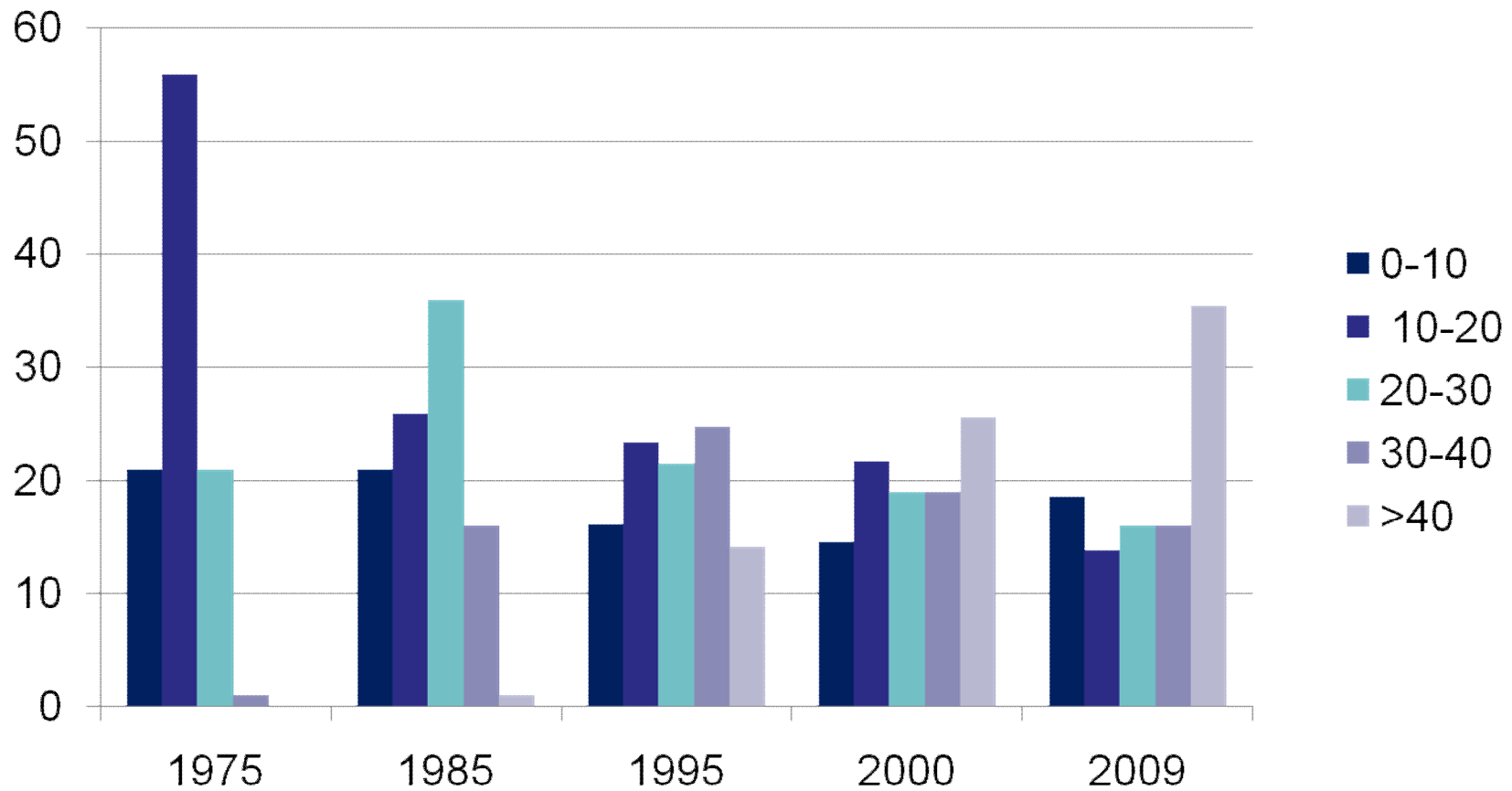
Number of patients according to age and severity of haemophilia, treated in the VCK at 01-01-2009



Patients according to age and severity treated at the VCK in 2009



Age of patients with severe haemophilia known at VCK



Age at death between 01-01-2000 and 01-01-09 of VCK pat's born before 1969

	Severe	Moderate	Mild	Total
Total number of patients	21	2	9	32
Median age (years)	59	72	75	64
Range (years)	38-73	71-73	60-89	38-89



Causes of death and age at death between 01-01-2000 and 01-01-09 of VCK pat's born before 1969

Causes of death	Severe	Moderate	Mild	Total
HIV related	1			1 (3%)
HCV related (carcinoma)	4 (2)	1 (1)		5 (3) (16%)
Malignancy (excl. hepatocellular ca)	5	1	8	14 (44%)
Intracranial bleeding	3		1	4 (13%)
Heart failure	1			1 (3%)
Other	7			7 (22%)
Total number of patients	21	2	9	32

Treatment options for haemophilia

- “ Until mid 60s: bedrest
- “ Now: prophylaxis
on demand

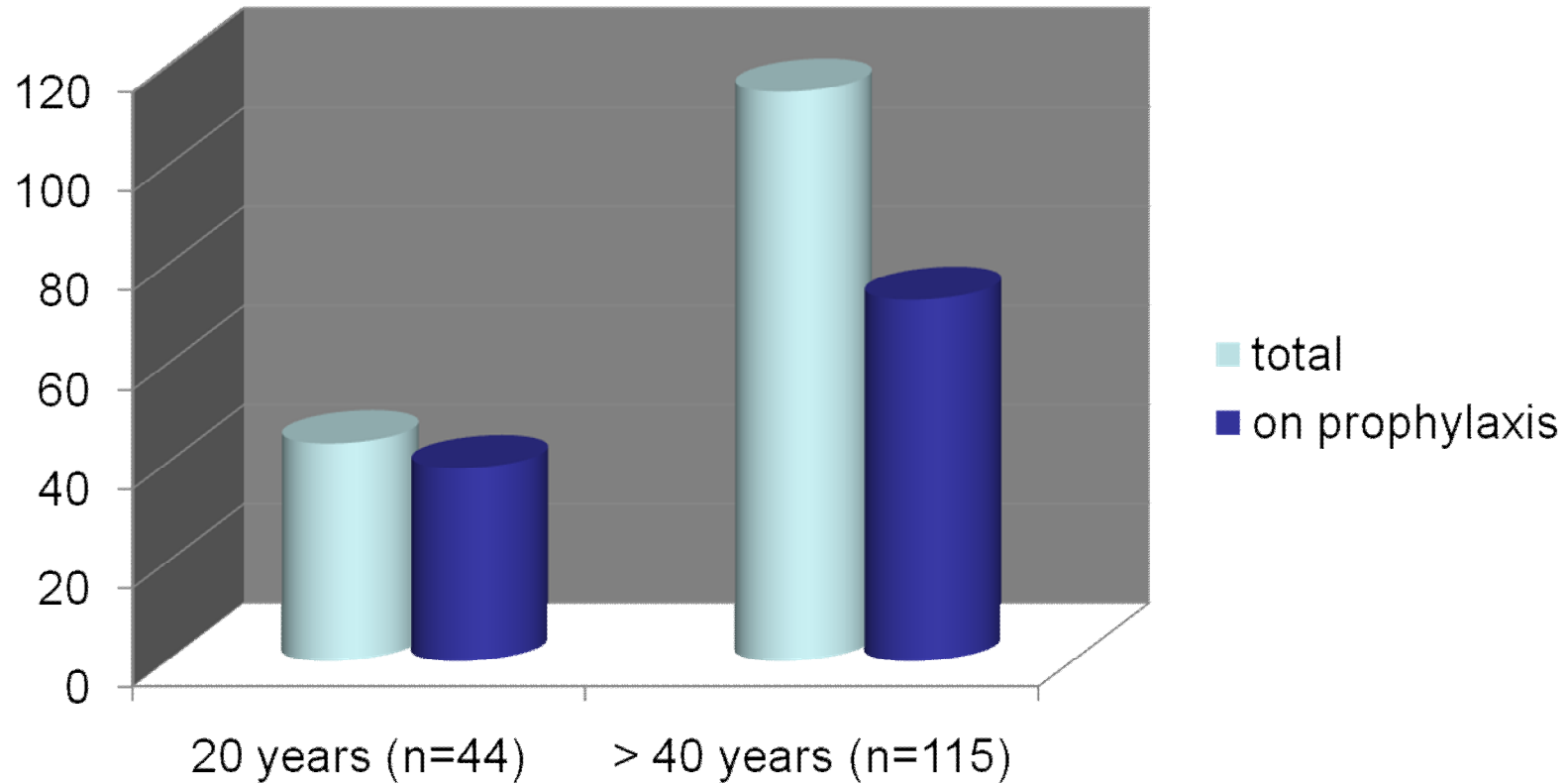


Prophylaxis

“ Prophylaxis: permanent
temporarily

number of bleeds
after severe / recurrent bleeding
medical intervention
rehabilitation

Number of patients with severe haemophilia on prophylaxis





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Criteria to temper or discontinue prophylaxis

- “ No bleeds and good clinical condition
- “ Complete recovery (temporarily prophylaxis)
- “ Lack of compliance
- “ Poor venous access
- “ Financial

Haemophilia treatment continuous infusion (CI)

- “ Severe bleeding (psoas muscle, intestinal, intracranial)
- “ Severe trauma
- “ Surgery

50 U/kg FVIII (100 U FIX) bolus
4 U/kg/ hr FVIII/IX CI



Haemophilia treatment specific problems in elderly pat's

- “ Poor venous access, especially in senior patients
- “ Improper use of clotting factor concentrate (as painkiller)
- “ Lack in knowledge of care giver
- “ Ignorance of bleedings by care giver or patient
- “ **Co morbidity**

With increasing age, haemophilia patients will suffer from haemophilia as well as age related diseases



Co-morbidity



Co-morbidity is defined as the effect of all other diseases an individual patient might have other than the primary disease of interest

Co-morbidity

Haemophilia related

Arthropathy

Chronic hepatitis C

HIV infection

Inhibitor development

not related

Internal Diseases

Cardiovascular

Malignancy

Surgery, tooth extraction

Urological (prostate)

Sexual problems

Psycho-social impact

Quality of life



Case history patient A

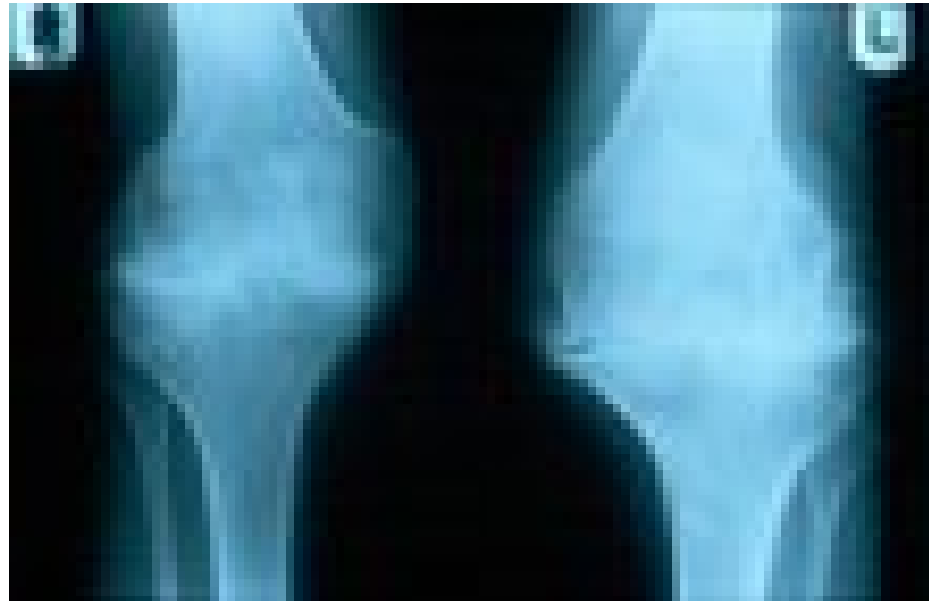
- “ 53 years severe haemophilia A
- “ Arthropathy of ankles, knee and elbows
- “ Chronic HCV
- “ HIV with HAART
- “ Diabetes Mellitus
- “ Hypertension
- “ Overweight
- “ Cardio Vascular disease with coronary bypasses

Number of elderly pat's with severe haemophilia and co-morbidity in the period 2000-2007 (n=145)

“ Severe arthropathy in 2 or more joints	145
“ HIV positive	17
“ HCV RNA positive	74
“ Number of patients known with one or more other diseases	65

Orthopedic problems → arthropathy

- “ Therapy:
- “ Conservative
- “ Surgery

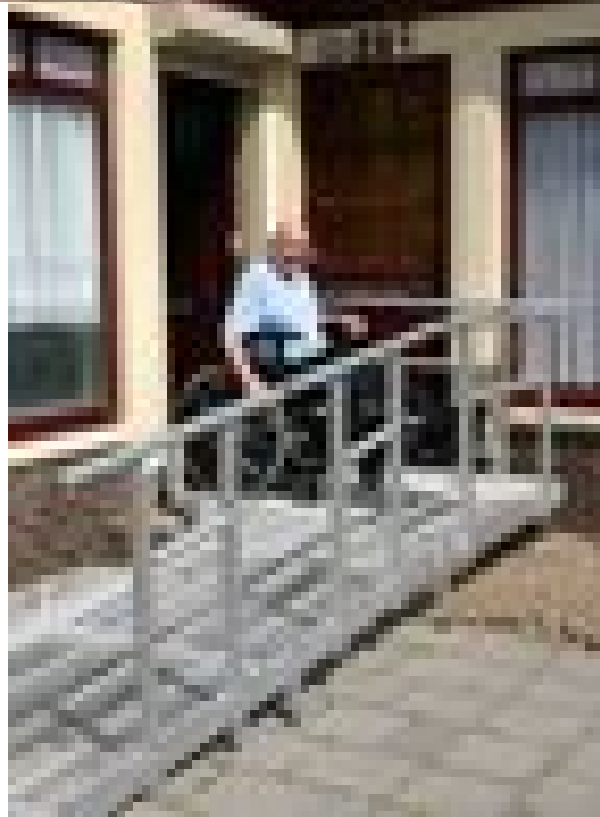


Orthopedic problems → arthropathy → therapy

- “ Conservative
 - physical therapy
 - pain medication
 - orthoses
 - adaptations
 - training for balance and falling



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Guideline for the use of analgesics for haemophilic arthropathy

- “ Paracetamol (500-1000 mg, max 6 times a day) is the initial medication of choice; if not effective:
- “ Paracetamol and a muscle relaxant (diazepam 5 mg, 1-3 times a day), or:
- “ Paracetamol and codeine (10-20 mg, max 6 times a day)
- “ Tramadol is indicated for very severe pain (50-100 mg, 3-4 times a day)
- “ Morphine (20 mg 2 x dd)

- “ TENS, hot packs

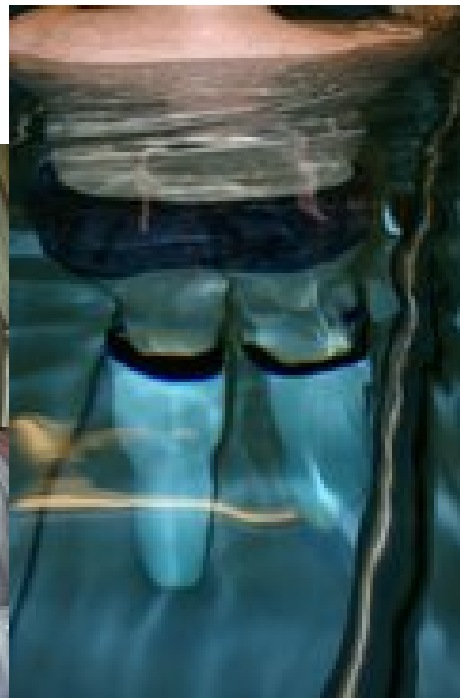
Orthopedic problems → arthropathy → therapy

“ Orthopedic surgery

arthrodeses

joint replacement

Followed by intensive rehabilitation



Orthopaedic problems

Adequate functional training, pain killing, adaptations and orthopedic intervention, can improve activities, participation and quality of life of elderly patients with painful arthropathic joints





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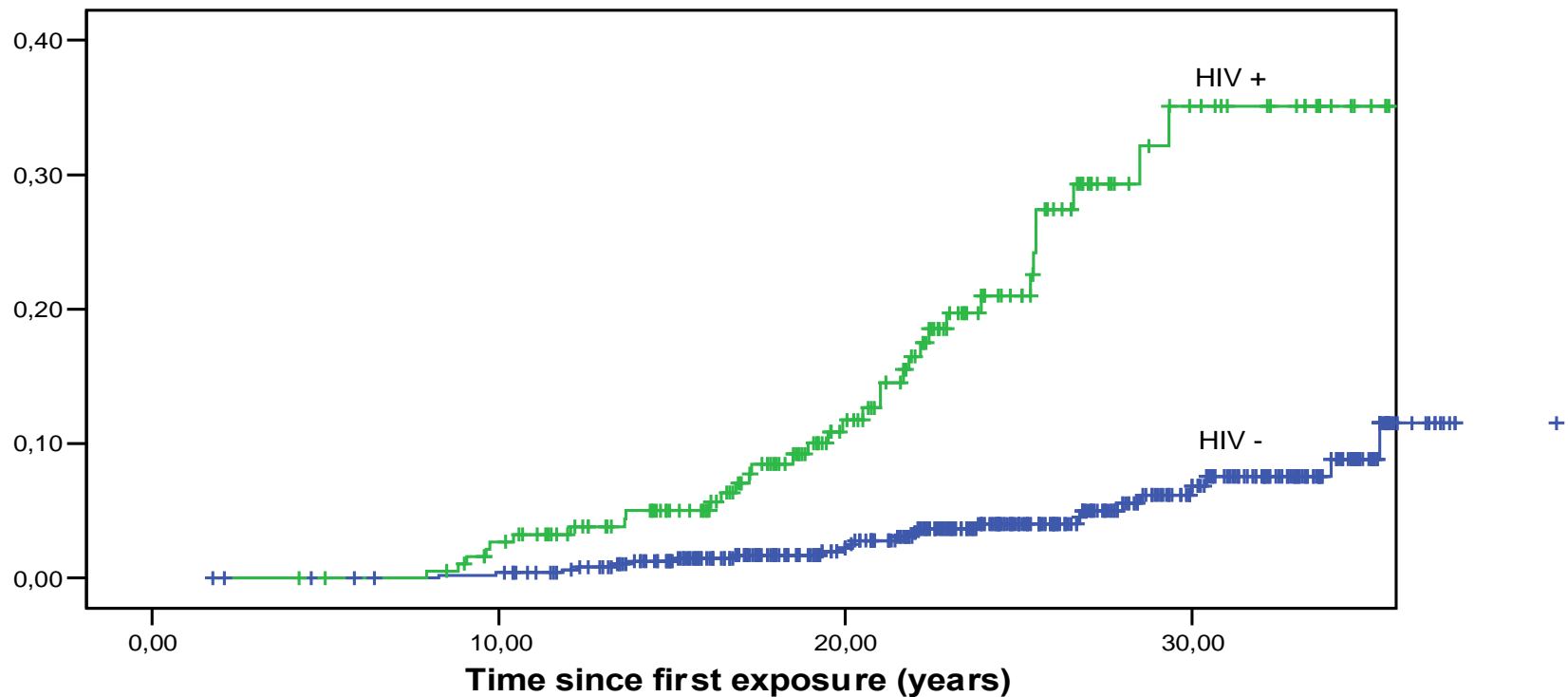
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When functional condition further deteriorates admission into a nursing home may be indicated. This requires good information and coordination by the hemophilia treatment center.

HCV infection

50 % of severe haemophiliacs > 40 years has chronic HCV infection

Cumulative incidences of End Stage Liver Disease in patients with chronic hepatitis C according to their HIV status (190 HIV+, 497 HIV-)





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HCV infection

- “ In HCV infected patients therapy with interferon should be considered
- “ Careful follow-up is required

HIV infection

- “ In 1985 16% of Dutch haemophiliacs was infected with HIV
- “ 67% of HIV infected patients has died

- “ HAART therapy may cause side effects:
 - . Increased bleeding tendency
 - . Metabolic complications: DM, hyperlipidemia, etc
 - . Hepato-toxicity: steatosis
 - . Nephro- toxicity

Non-haemophilia related Co-morbidity

- “ Internal diseases: Hypertension, Diabetes Mellitus, obesity
- “ Cardio-vascular diseases
- “ Urological problems
- “ Dental problems
- “ Malignancy
- “ Surgical interventions
- “ Sexual problems



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Cardiovascular disease in haemophilia

- “ Exact incidence unknown
- “ Lower mortality of ischaemic heart disease in haemophilia patients

Ischaemic Heart Disease

Prevalence 1993-1998 (Kulkarni *Am J Hematol* 2005)

< 30 years 0.05%

> 60 years 15.2 %

1972-2001 death caused by Ischaemic Heart Disease increased from

2 6 % (Plug *J Thromb Haemost* 2006)



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Indications for antithrombotic therapy

- “ Cardiovascular diseases
 - ischaemic heart disease



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Sexual problems

- Caused by pain
- Result of functional limitations
- Fear for viral transmission

- Side effects medication
- Internal diseases (high blood pressure)
- Cardio vascular disease

Co-morbidity requires extensive comprehensive care





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Pitfalls in patients with co-morbidity

- “ Haemophilia specialist pays no attention
- “ Patient does not mention (mild) haemophilia to other specialists
- “ Ignorance of haemophilia by specialist
- “ Fear of haemophilia by specialist
- “ Development of an inhibitor
- “ Bleeding tendency caused by medication

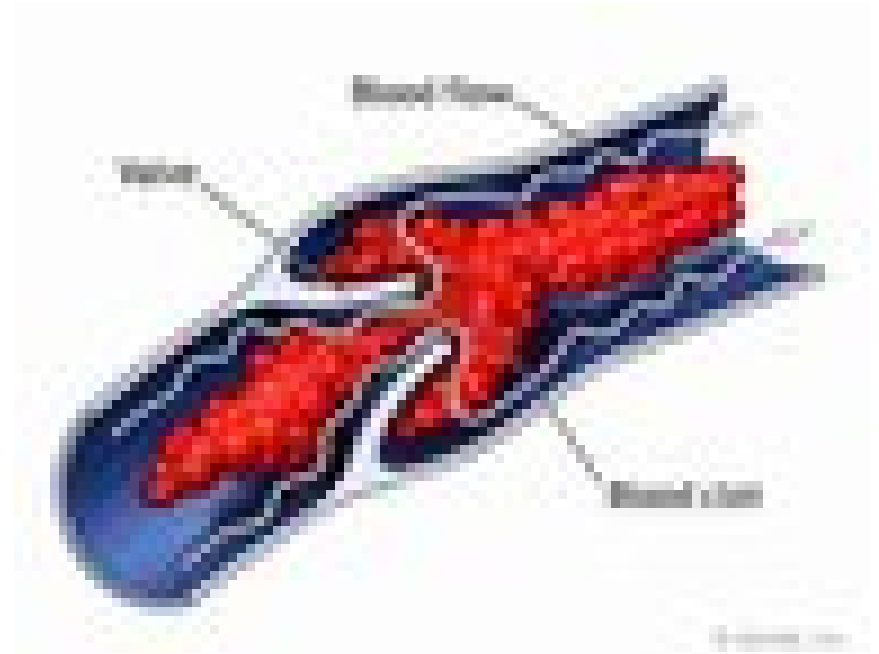
The tackle

- “ Education and information of patient
- “ Psycho-Social support
- “ Education and information of specialist and nursing staff
- “ Adequate care during surgical period or medical intervention
- “ Careful follow-up after surgery or intervention (inhibitor!)
- “ If indicated: thrombosis prophylaxis, early mobilization



Prevention of deep venous thrombosis

- “ **When?** during (orthopaedic, abdominal, malignancy) surgery when complete clotting factor correction is required



Prevention of deep venous thrombosis

“ How?

heparin, according to local protocols **after** bolus injection with factor VIII/IX continue until top levels of CF are < 0.5 U/L and mobilization

compression stockings peri-operatively until full mobilization

early mobilization



Problems caused by co-morbidity

<i>Functional limitations</i>	<i>Social problems</i>
Physical deterioration, fatigue	<ul style="list-style-type: none">- Lack of understanding- Reduction in social contacts
<i>Psychological complaints/ symptoms</i>	<i>Societal problems</i>
<ul style="list-style-type: none">- Fear and depression- Feeling of losing control- Lower capacity of self care	<ul style="list-style-type: none">- Reduced participation in labour and leisure activities- Increase in health expenditure

The fear factor

- “ Fear for lack of control and lack of coordination especially during hospitalization, when you are not able to check and control the treatment you receive
- “ Fear for venous access performed by others
- “ Emotional distress during hospitalization caused by negative experiences during childhood





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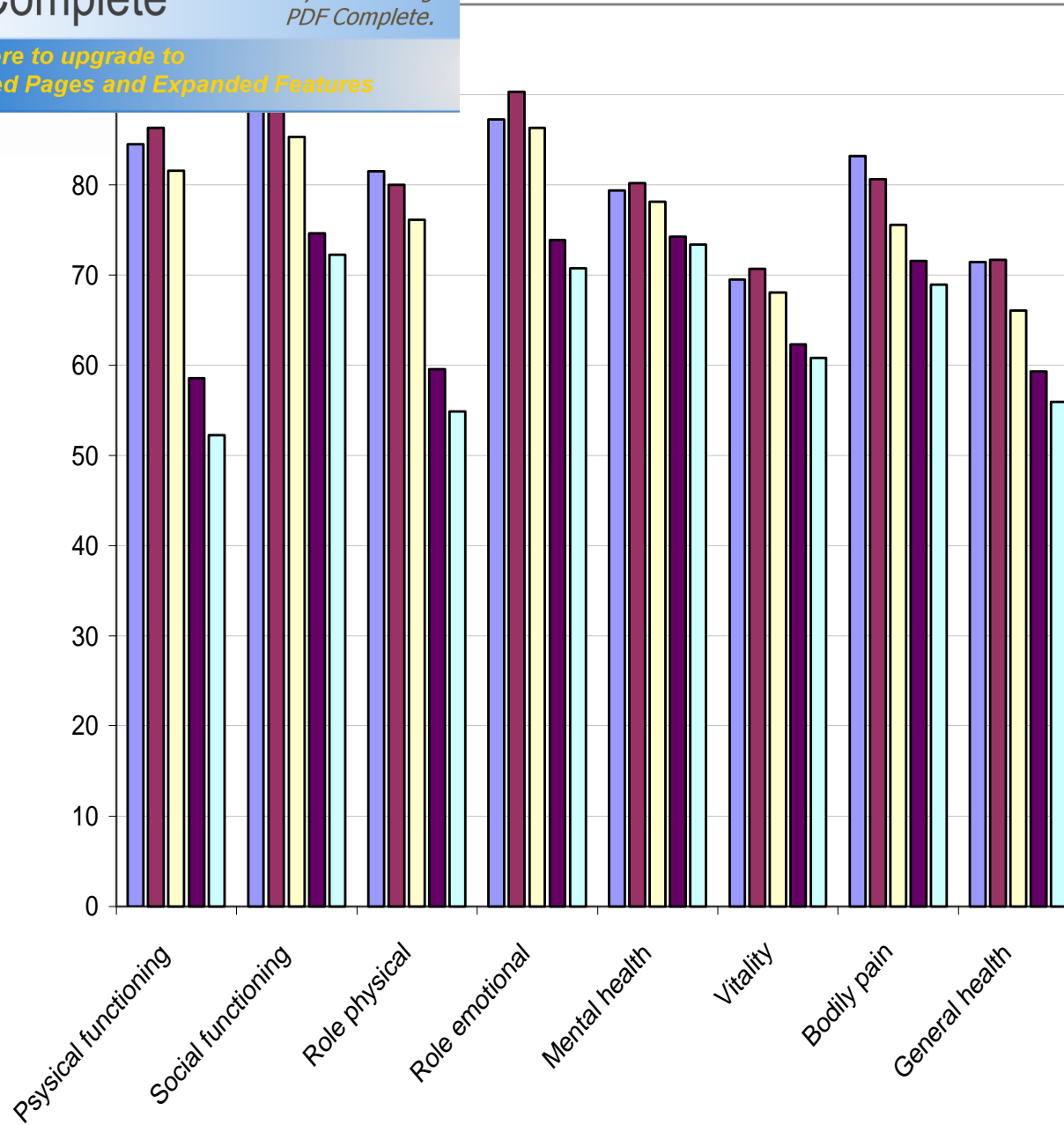
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Impact

Limitations have a great impact on quality of life of

- . Patients
- . Partners
- . Family members
- . Friends
- . Colleagues

RAND-36 score



Health related quality of life domain

- General population
- Haemophilia patients in general, aged <40 years (n = 279)
- Haemophilia patients with current HCV infection, aged <40 years (n = 129)
- Haemophilia patients in general, aged 40 years or older (n = 323)
- Haemophilia patients with current HCV infection, aged 40 years or older (n = 204)



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Challenge

- “ Adequate treatment of haemophilia
- “ Access to treatment for co-morbidity
- “ Improve quality of life of the elderly haemophilia patient

What a patient can do to avoid complications

- “ Regular (> 1 year) contact with haemophilia center
- “ Inform any physician you have haemophilia and that your haemophilia specialist should be contacted to discuss treatment regimen
- “ Check if care is coordinated (call HTC)
- “ Instruct partner and family
- “ Become a member of patient organizations
- “ Wear medic alert
- “ Save telephone number of HTC in your mobile telephone

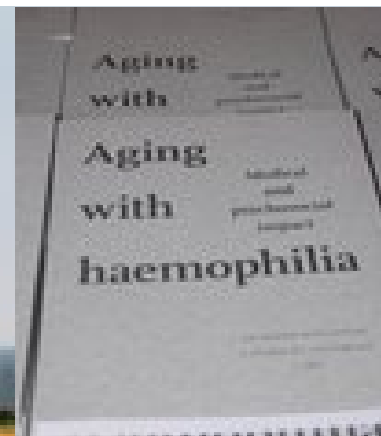
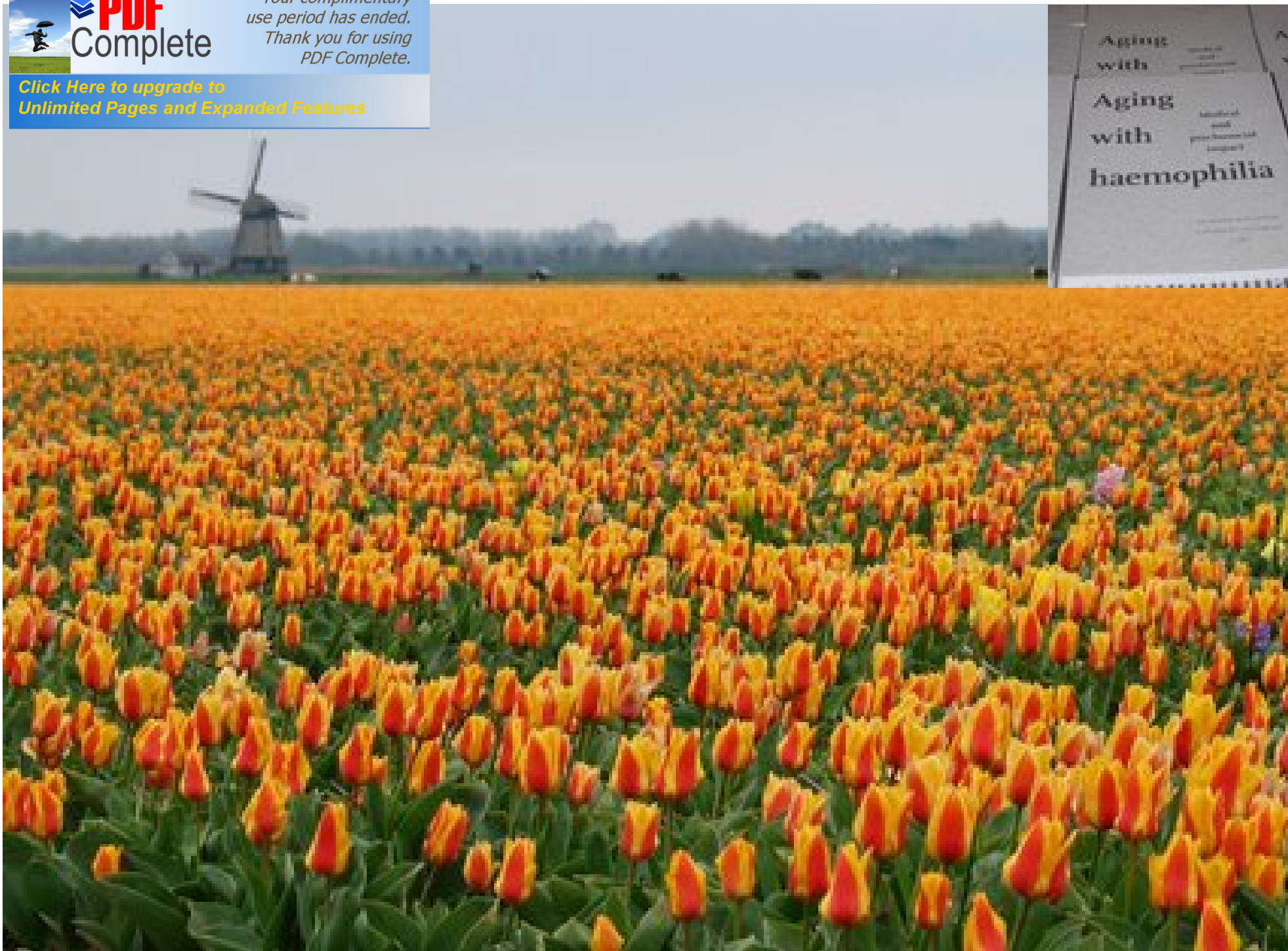
Conclusions

- “ With increasing age, patients with haemophilia will, apart from haemophilia related diseases, suffer from the same diseases as other seniors
- “ Quality of life in elderly patients decreases
- “ Hemophilia care givers should pay attention to the development of co-morbidity and to the well being of patients
- “ Haemophilia treatment centers should take the responsibility for coordination of care
- “ Patient should be alert too!



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