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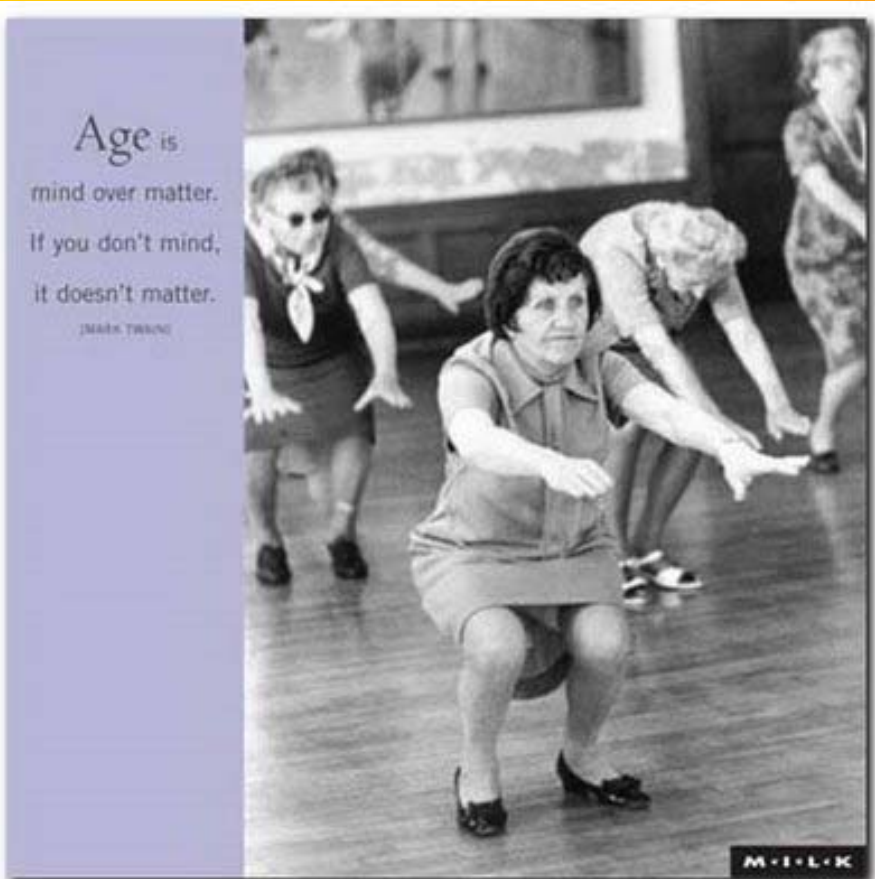
“You don’t look anything like the long haired, skinny kid I married 25 years ago. I need a DNA sample to make sure it’s still you.”



TheAlfred



Health Issues for an Ageing Haemophilia Population



Huyen Tran
Ronald Sawers Haemophilia Centre,
The Alfred Hospital,
Australian Centre for Blood Diseases,
Monash University

Case (1)

- 72-year old male
- Moderate haemophilia A (FVIII 3%)

Case (2)



Other medical conditions

- Chronic hepatitis C
 - Chronic liver disease
- Insulin requiring diabetes
- Hypertension
- Obesity
- Osteo-arthritis
- Cataracts
- COPD (ex-smoker)
- Bladder TCC
- Depression

Clinics

- GP
- Hepatitis
- Endocrinology
- Cardiology
- Rheumatology
- Ophthalmology
- Respiratory
- Psychology
- Social work

Potential medical illnesses for ageing haemophilia patients



Haemophilia related

- Chronic hepatitis
- HIV infection
- Haemophilic arthropathy

- Inhibitor development

Non-haemophilia related

- CVS disease
- Cancer
- Other
 - Hypertension, diabetes, lipids, obesity, kidneys, cataracts

Psycho-social

Quality Of Life

Sexual health

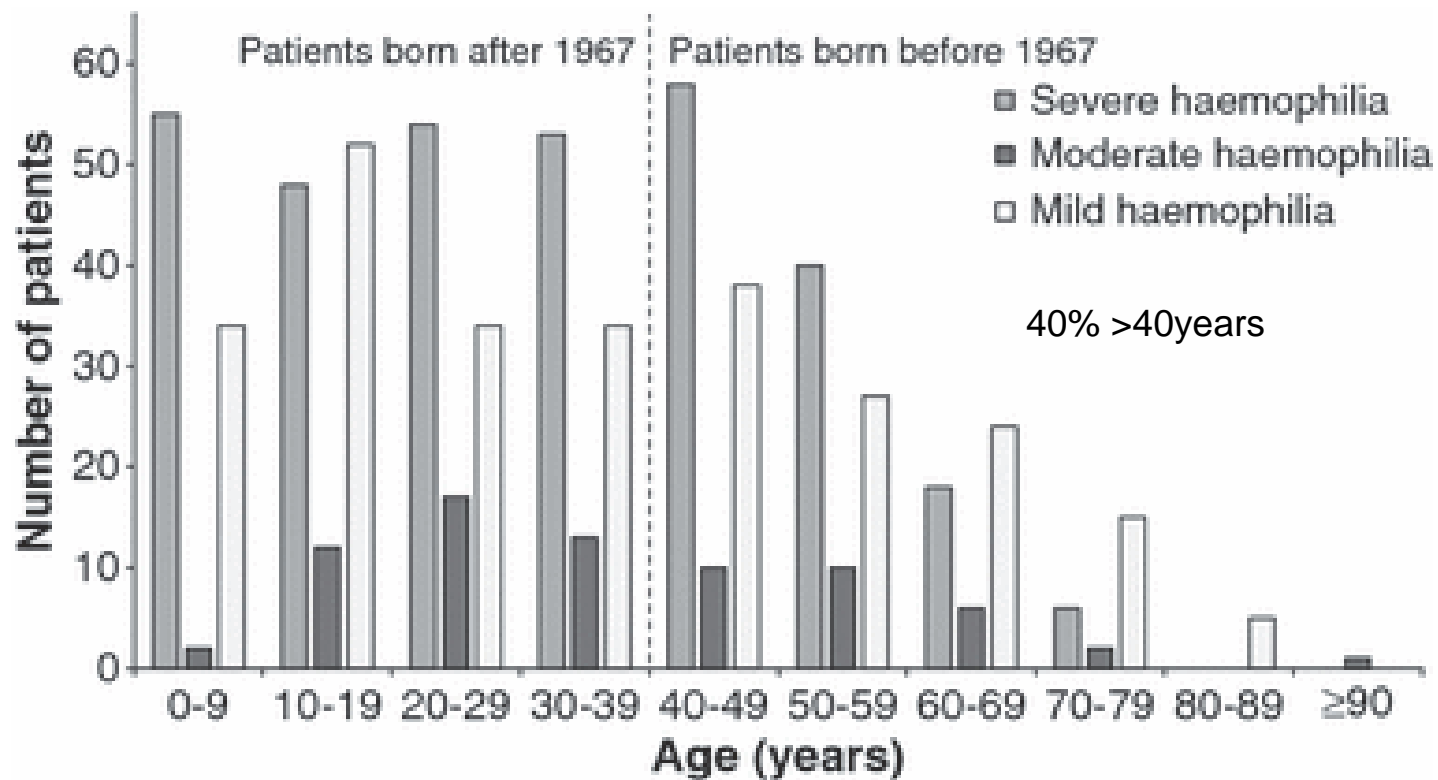
You can't help
getting older,
but you don't have
to get **old.**

(GEORGE BURNS)



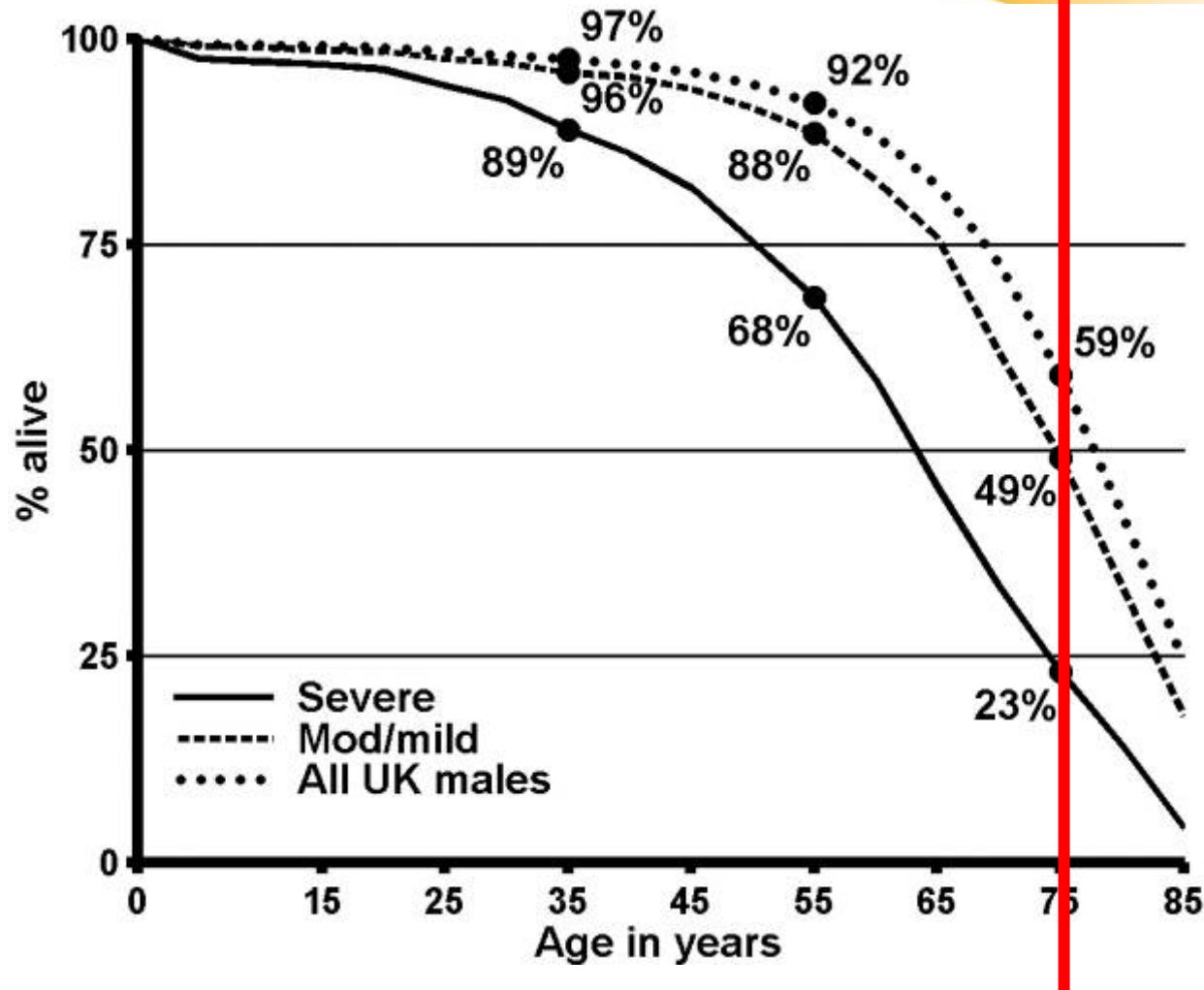
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Number of patients according to age and severity



van Creveldkliniek Centre, Netherlands. N=668

Survival of men in UK with hemophilia not infected with HIV and in the general male population, 1999



Darby, S. C. et al. *Blood* 2007;110:815-825

Growing **old** is inevitable, growing **up** is optional.

(ANON)



M·I·L·K



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Haemophilia related co-morbidities

- Haemophilic arthropathy
- Pain management
- Orthopaedic surgery
- Rehabilitation and physical therapy
- Balance dysfunctions & risk of falls
- Osteoporosis

Hepatitis C

Natural history

- Chronic infection, 80%
- End-stage liver disease, 17%;
35% if HIV co-infected*
- Death related to HCV, 6.5%
- Liver cancer, 1.5%

Treatment

- Peg IFN & ribavirin
- Potential AE
 - Flu-like symptoms
 - Weight loss
 - Psychological
 - Haematologic

- 10% haemophilia patients
withdraw from Rx

* 35 years FU

Inhibitor development

- Mild/moderate haemophilia A
 - Occurs later in life
 - **Medical & Surgical needs (age-related) requiring factor correction**
 - Careful monitoring

Factors influencing inhibitor development in patients with mild/moderate haemophilia A

Genetic risk factors

- Missense mutations in the A2 and C2 domains of factor VIII gene
- HLA-II polymorphisms

Non-genetic risk factors

- Immunological factors (T-cell response)
- Surgery and trauma (pro-inflammatory)
- Treatment related factors
 - Changes of FVIII product type
 - Modality of administration (continuous infusion)
 - Intensive treatment (factor replacement)

Management of inhibitors

Acute bleeds

- Recombinant FVII_a
- Activated prothrombin complex concentrate
- (DDAVP)

Immune induction tolerance

- Regular FVIII infusion, 25% success rate
- Rituximab
- Spontaneous resolution

Common Ageing related ailments

- Hypertension
- Dyslipidemia
 - lowest among severe haemophilic patients
- Diabetes
 - Higher prevalence among mild haemophilia?

Common Ageing related ailments

- Obesity

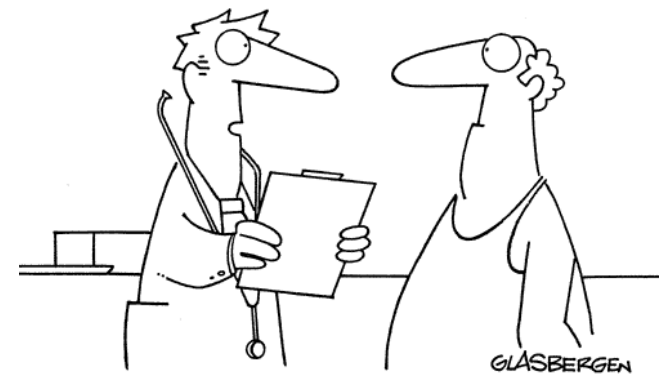
- Adult Dutch haemophilia patients (1992-2001)
 - > BMI 25–30 kg m² increased from 27% to 35%
 - > BMI > 30 kg m²) 4% to 8%
- Risk factor for diabetes, atherosclerosis, CVD, arthropathy

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“My doctor told me to start my exercise program very gradually. Today I drove past a store that sells sweat pants.”

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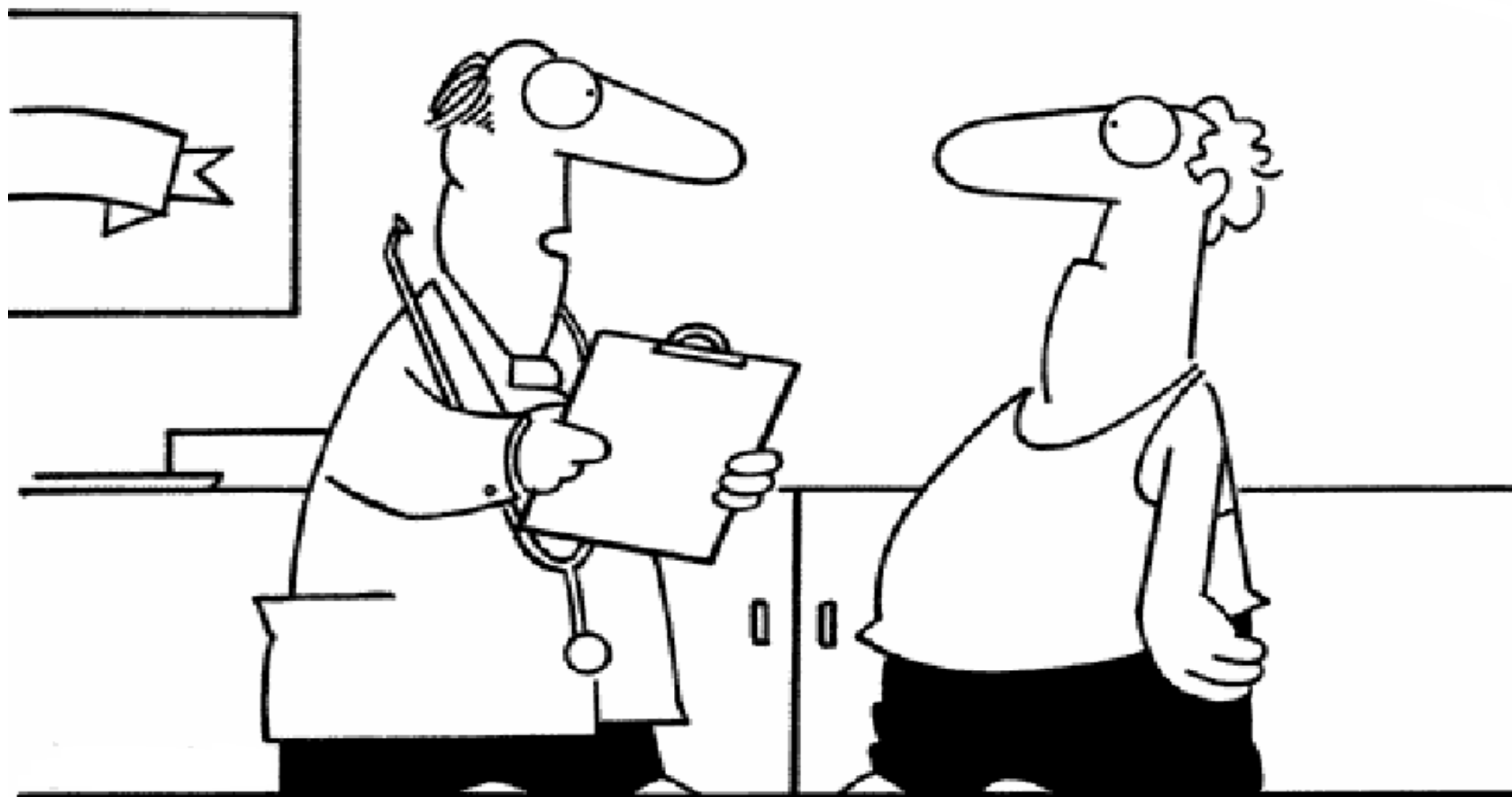
“The handle on your recliner does not qualify as an exercise machine.”

Cardiovascular disease

- Risk of death from IHD for haemophilia 62% less vs. general population
 - Rare MI among severe haemophilia
- Hypocoagulable state is protective?
 - Surrogate marker of intimal thickness is variable
 - (Temporal) Link between factor replacement and ACS
 - advanced atherosclerosis at autopsy
- No specific guidelines
 - Cooperation with cardiologist
 - Factor correction with tailored antiplatelet therapy



"An aspirin a day will help prevent a heart attack if you have it for lunch instead of a cheeseburger."



“An aspirin a day will help prevent a heart attack if you have it for lunch instead of a cheeseburger.”

Cancer



- Ageing haemophilia patients will be as susceptible to developing common cancers (prostate, colon, skin, lung)
 - Surveillance important
 - Mortality rates are same compared with general population
- Haemostatic support for diagnostic procedures & cancer surgery
- Risk of venous thrombosis
- Associated risk for haemorrhage
 - Thrombocytopenia & risk of bleeding with chemo-radiotherapy
- Liaison with oncology team

DEATHS, INCIDENCE AND SURVIVAL RATES FOR COMMON REGISTRABLE CANCERS

	Deaths (2004)	Incidence (2001)Five-year relative survival (1992-97)
Prostate	2,761	11,191
Colon	1,432	4,233
Skin	821	5,024
Lung(b)	4,733	5,384

Hospitalisation

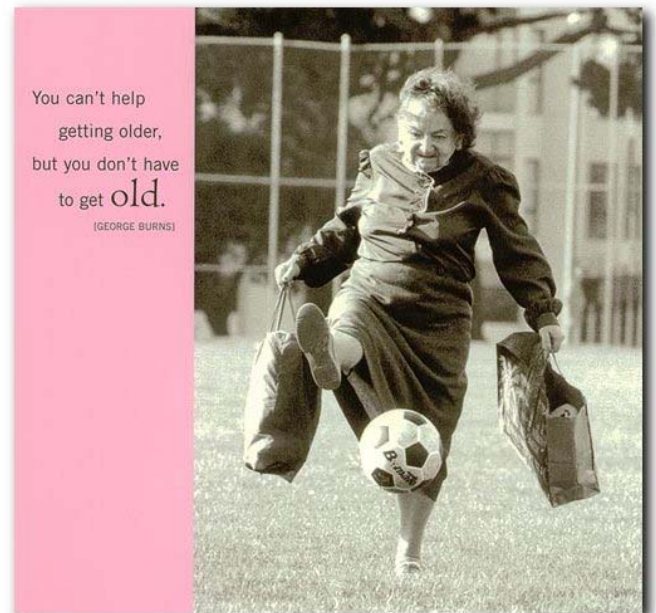
- Patients want to stay away from hospitals
 - Negative previous experiences
 - Aggregated by fear
 - Loss of control during admission can cause stress
 - > E.g., unable to self-infuse
- Good information and education by a haemophilia nurse is crucial

Sexuality

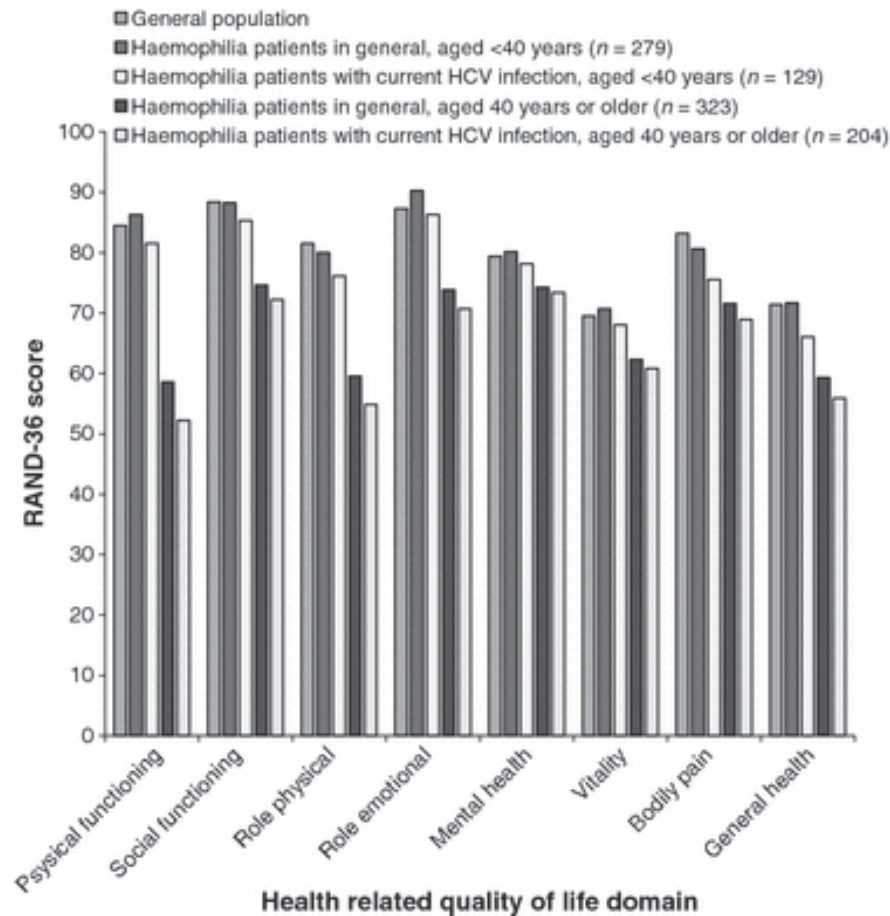
- Sexual dysfunction (lack of desire or response)
- Contributors
 - Pain
 - Haemophilic arthropathy
 - HCV HIV infection
 - Co-morbidities (HPT, DM, kidney disease)
- Counselling
- Pre-sex analgesia
- Position advice
- Medications to enhance erection
 - Viagra may cause bleeding

Psychological impact

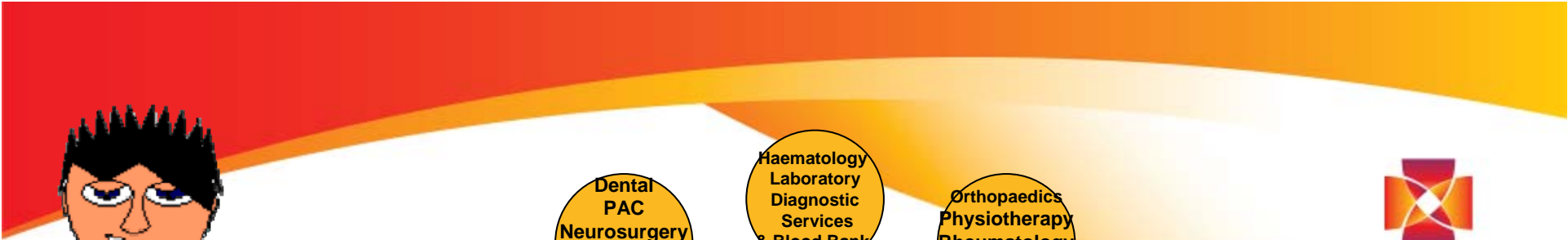
- Decline in health
 - Altered family dynamics
 - Loss of employment
 - Early retirement
- Positive aspects of ageing
 - Experience with overcoming haemophilia-related adversity during early years



Health-related quality of life among haemophilia patients >40-yrs vs. <40-yrs



RAND-36 questionnaire



Summary

- Adult haemophilia patients are susceptible to haemophilia-related AND non-haemophilia related co-morbidities in later life stages
- Aim prevent or reduce co-morbidities
- Haemophilia treatment centres play an integral role in co-ordinating care
 - Leading to improved QoL and maintain patient independence
- ***Continue to identify and improve gaps in care with ageing***

It's not the years
in your life that count,
it's the *life*
in your years.

(ABRAHAM LINCOLN)



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