Co-morbidity in the ageing hemophilia patient

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A multidisciplinary care approach for elderly haemophilia patients – How to develop optimal strategies to treat co-morbidities in older haemophilia patients?

Hemophilia

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- Hemophilia is a congenital sex-linked disorder
- Incidence: 1:10000 males

- Deficiency in clotting factor VIII (hemophilia A) or IX (hemophilia B)

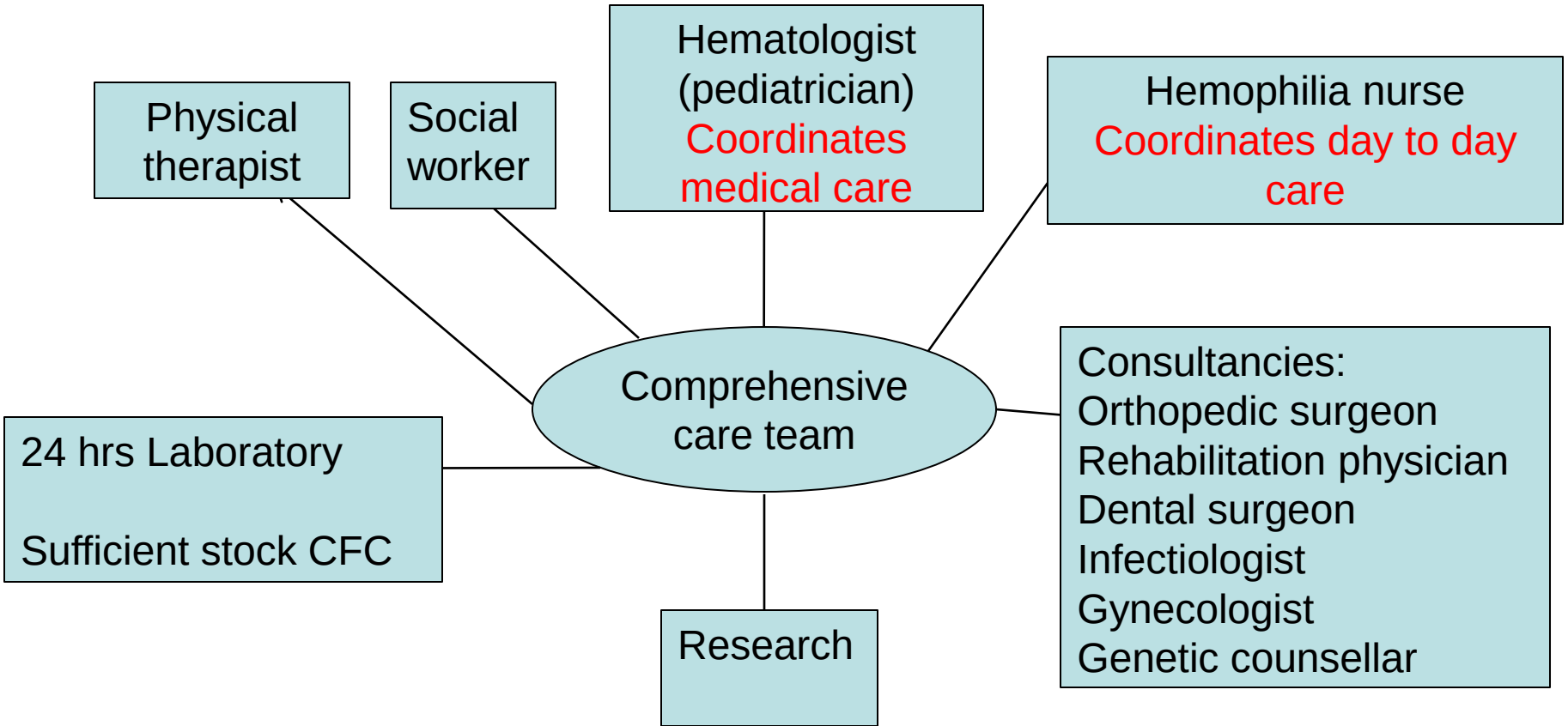
- Severity:
  - Severe (40%)
  - Moderate (20%)
  - Mild (40%)

Clinical expression

- Varies with severity
- Severe hemophilia: repeated hemorrhagic episodes, mostly in joints and muscles ⇒ arthropathy and functional limitations
- All patients with hemophilia: increased bleeding after trauma and medical interventions

Therapeutical setting

- All patient should be treated in hemophilia treatment centers in a comprehensive care setting



Therapy and life expectancy

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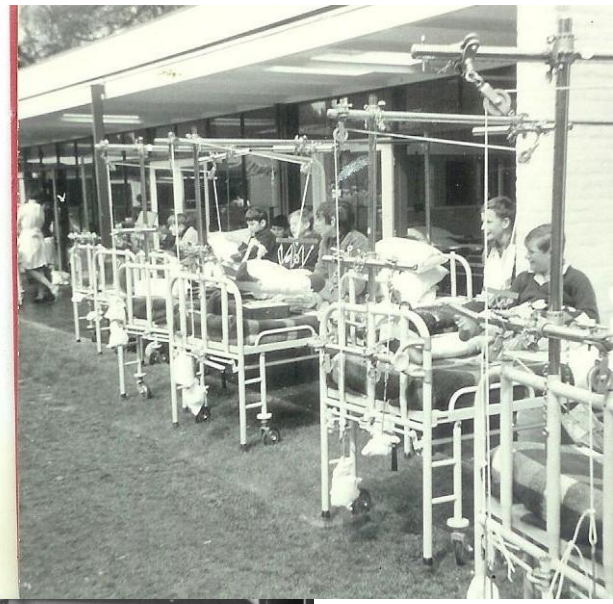
Until the 1960s

- Hospitalization with bed rest
- life expectancy 20 years
- poor quality of life



Therapy and life expectancy

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21st century in developed countries:

- Home infusion of clotting factor concentrates
- Life expectancy normal (when not infected with HIV or hepatitis C)
- Improvement of quality of life

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Life expectancy of hemophiliacs is increasing

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

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

Causes of death are changing

Cause of death in Dutch hemophiliacs (I Plug et al)

	1973-1986 N=43 (%)	1986-1992 N=45 (%)	1992-2001 N=94(%)
Hemorrhages	20 (47)	1 (2)	5 (5)
Aids	0 (0)	12 (27)	24 (26)
Hepatitis C related	0 (0)	5 (11)	21 (22)
Malignancies	13 (30)	7 (15)	12 (13)
Ischemic hart disease	1 (2)	0 (0)	6 (6)
Cerebrovascular disease	3 (7)	9 (20)	4 (5)
Other	6 (14)	11 (24)	22 (23)

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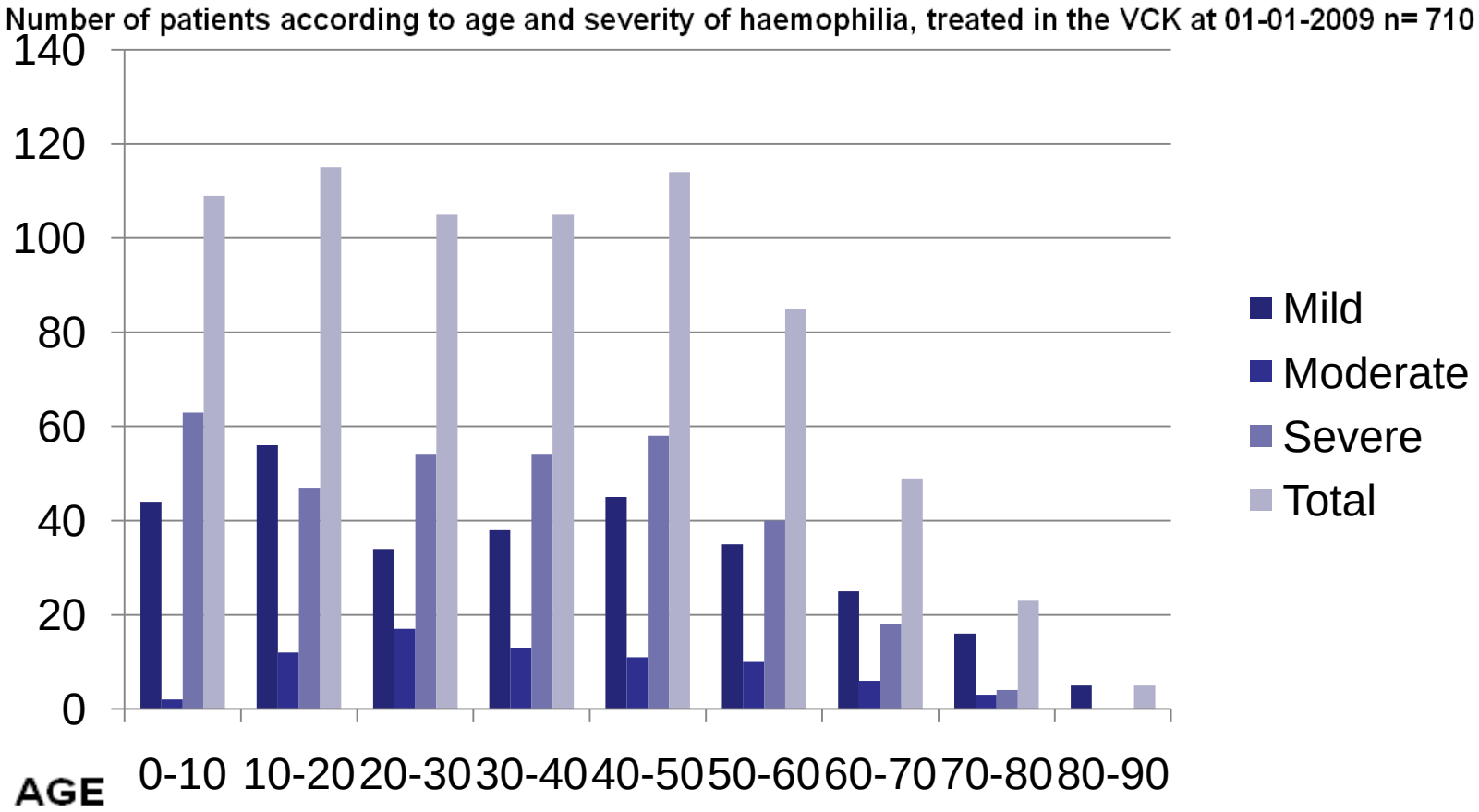
Age and number of patients with hemophilia treated at the VCK



Percentage and age of patients with severe hemophilia treated at the VCK

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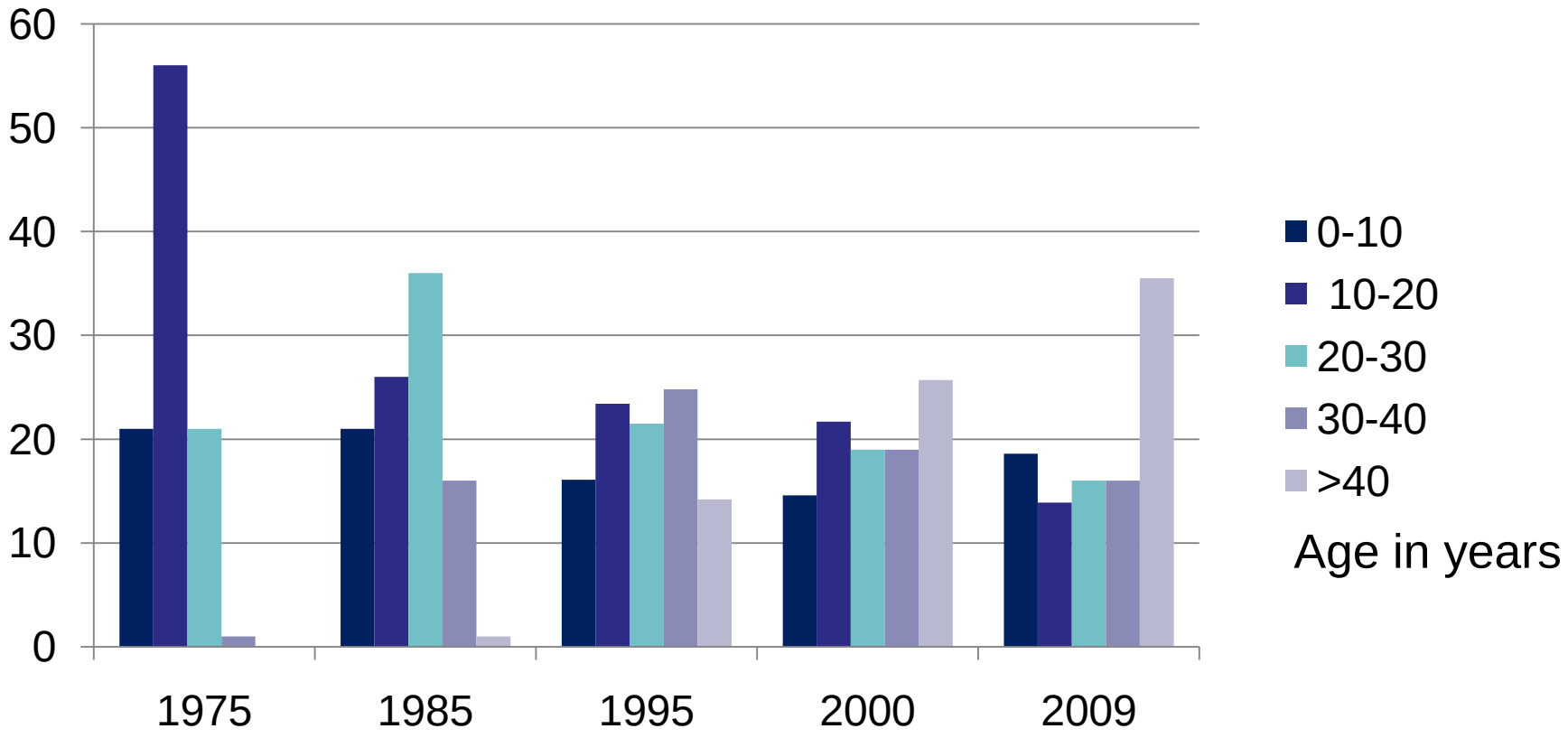

Impact

- > 35% of patients with severe hemophilia is > 40 years
- This number will increase over the years
- Older patients have different problems compared to the younger generation → co-morbidity

Co-morbdity

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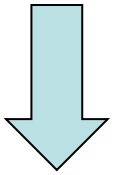
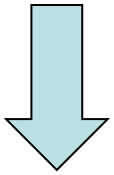
Co-morbidity is the result of the ageing process of cell systems



This process affects all systems and organs



It is an irreversable process occurring after the age of 40 years

Co-morbidity

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

⇩

Co-morbidity

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

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In the ageing hemophilia patients co-morbidity tends to become more of a problem than hemophilia itself

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Co-morbidity in hemophilia

Hemophilia related	not related
Arthropathy	Internal Diseases
Chronic hepatitis C	Cardiovascular
HIV infection	Malignancy
Inhibitor development	Surgery, tooth extraction
	Urological (prostate)

Osteoporosis

Sexual problems

Psycho-social impact

Quality of life

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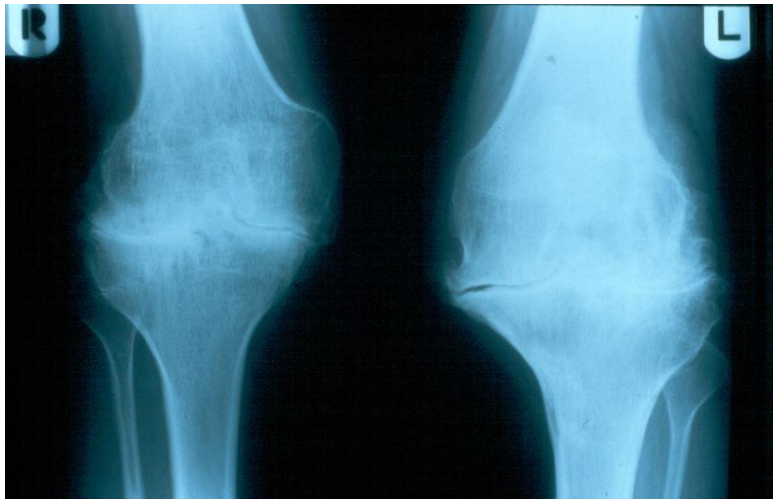
Incidence of hemophilia related

- Severe hemophilia
  - Arthropathy: 100% of patients > 40 years
  - Chronic Hepatitis C: 50% > 40 years
  - HIV 6%

- Mild hemophilia: inhibitors in elderly patients



Non-hemophilia related Co-morbidity incidence is mostly the same as in other males.
But not always!

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Ischemic Heart Disease

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

< 30 years 0.05%

> 60 years 15.2 %

1972-2001 death caused by Ischemic Heart Disease increased from

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

Ischemic Heart Disease

Management of Ischemic Heart Disease in hemophilia warrants close cooperation between cardiologist and hemophilia specialist

equilibrium

Clotting factor correction

Anti thrombotic therapy

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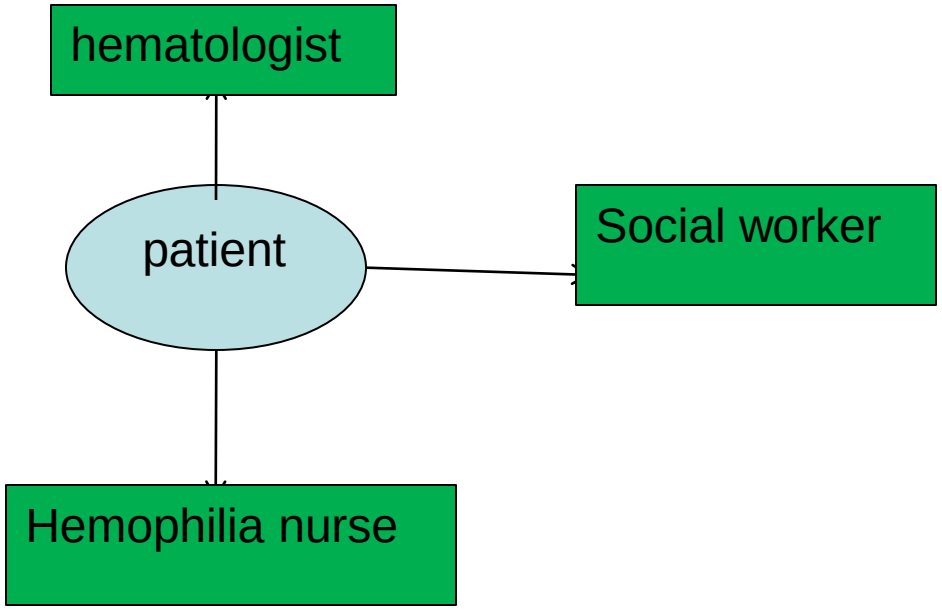
Young mild hemophilia patients and care givers



The senior severe hemophilia patient and the specialists around him

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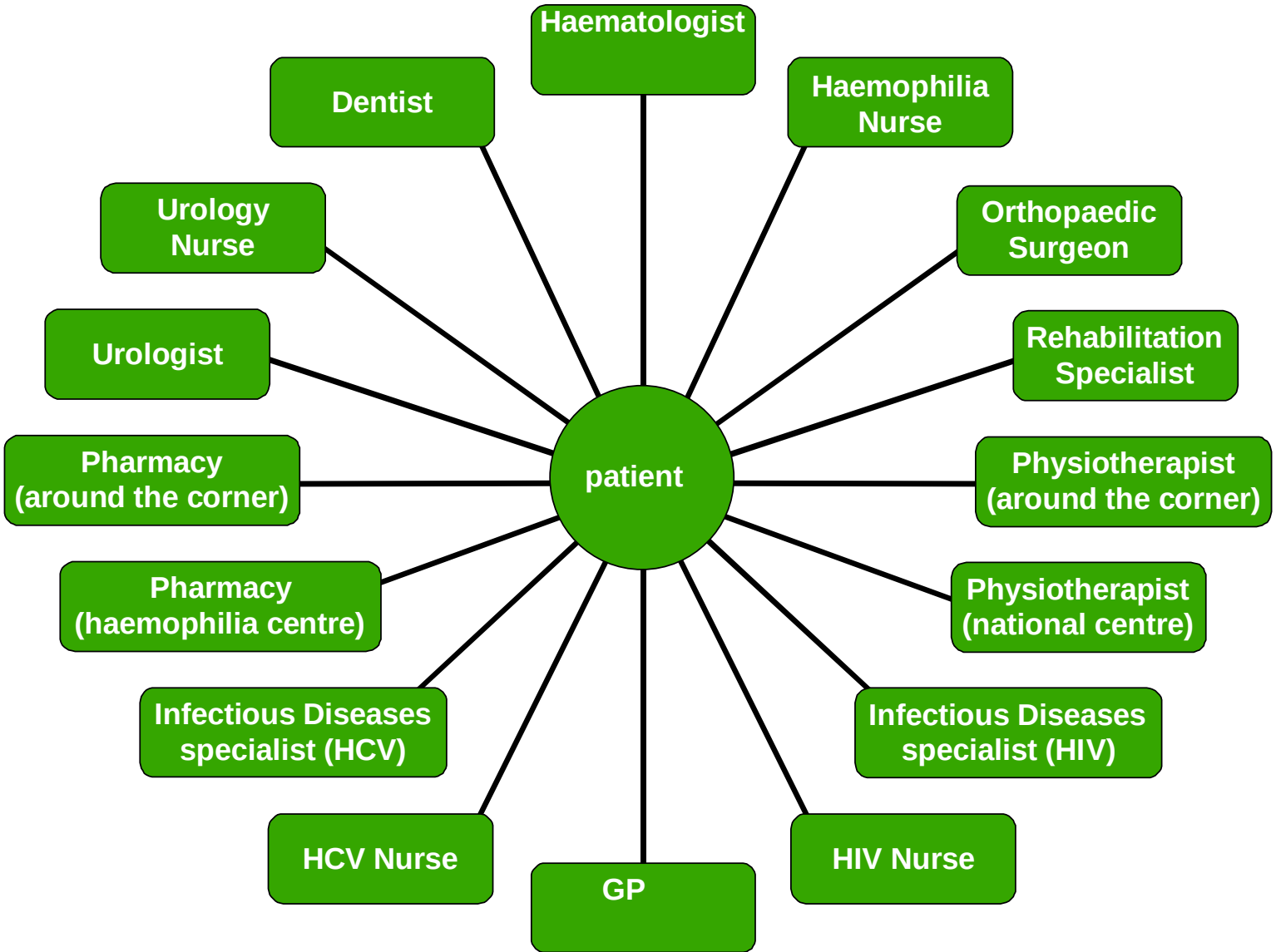

Co-morbidity requires extensive comprehensive care and a good coordination of care: the hemophilia specialist may also be the coordinator

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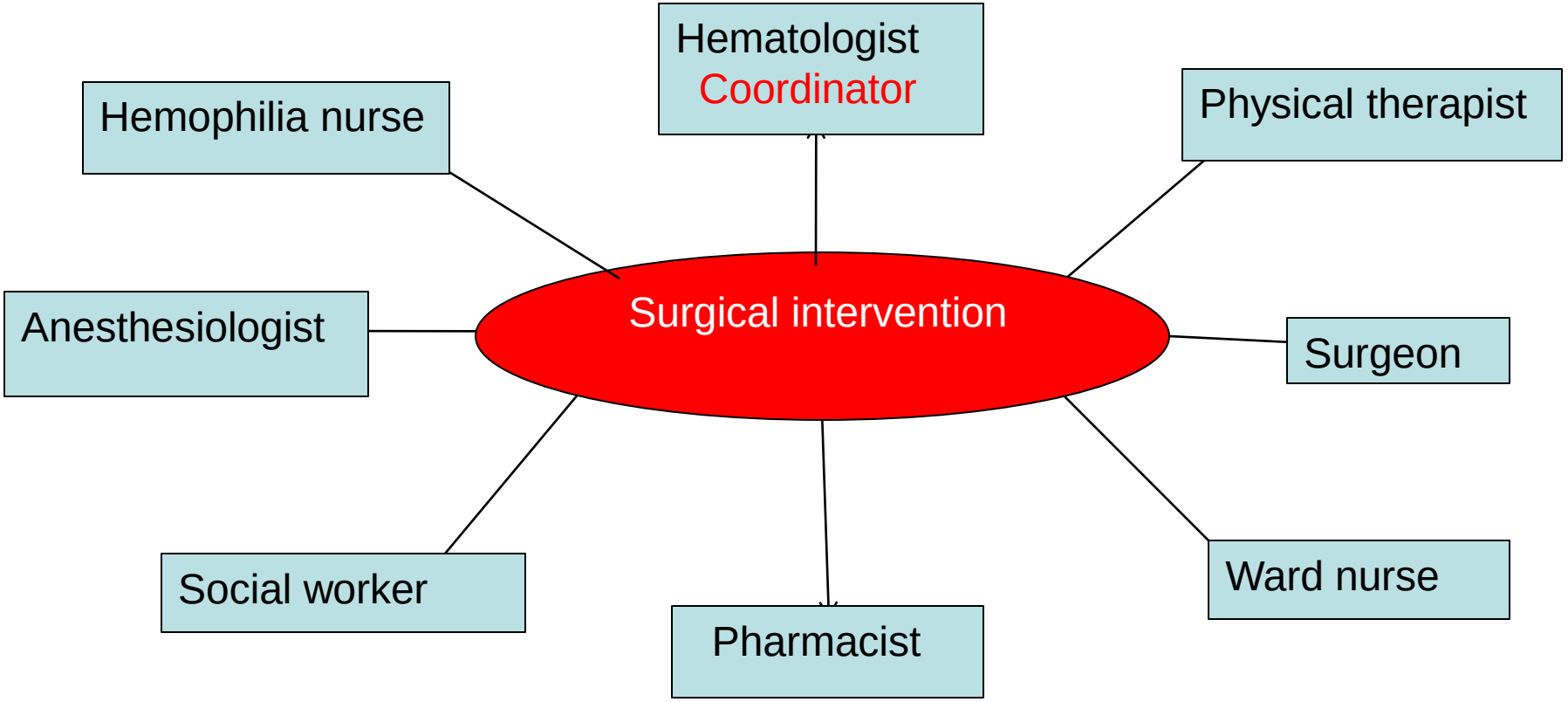
Comprehensive care for surgical intervention



Impact co-morbiditeit

Hemophilia and co-morbidity have an impact on

- Patiënt
- Partner
- Family members
- Friends
- Collegues

- **Quality of life and social functioning ↓**

Impact co-morbiditeit

Quality of life and social functioning ↓↓

Loneliness

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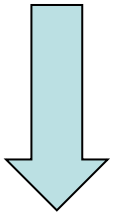
Challenge for care givers

- Adequate treatment of hemophilia
- Awareness and optimal treatment of co-morbidity



- Training in an interdisciplinary setting should focus on subject-specific medical issues

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Integrated care requires

- Timely identification of health risks related to co-morbidity
- Timely identification of age related health risks
- Management of integrated care for home dwelling elderly with co-morbidity
- Diagnostic and therapeutic advice by hemophilia specialists to support other specialists, general practitioners and home care nurses
- Application of scientific knowledge of complex co-morbidity within the clinical and extramural setting

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Intergrated care results in

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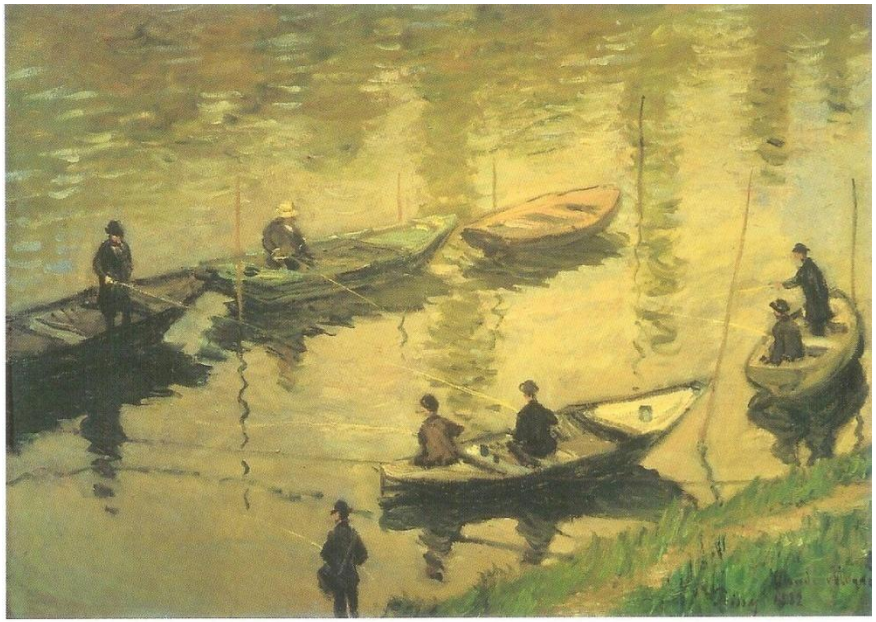
Improvement of quality of life of the ageing hemophilia patient



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Conclusions

- Hemophilia patients have an almost normal life-expectancy
- The number of ageing hemophilia patients is increasing
- With ageing hemophilia patients will increasingly suffer from co-morbidity
- Co-morbidity requires multidisciplinary and inter-disciplinary care

Centers of excellence are required to guarantee optimal care