



“Working together for
a green, competitive and inclusive Europe”

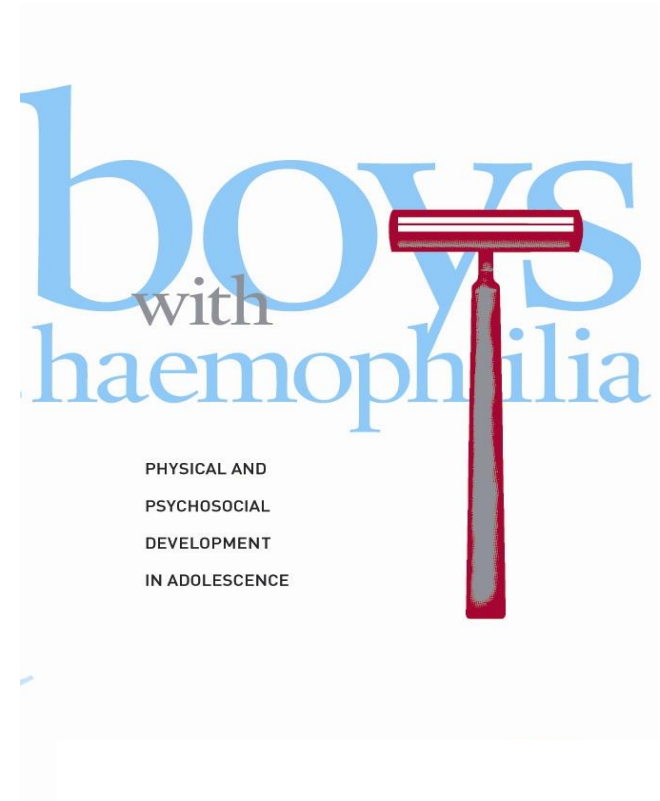
“HE-RO-IS strategic cooperation in hematology”

F SEE 2014-2021 No. 19-COP-0031

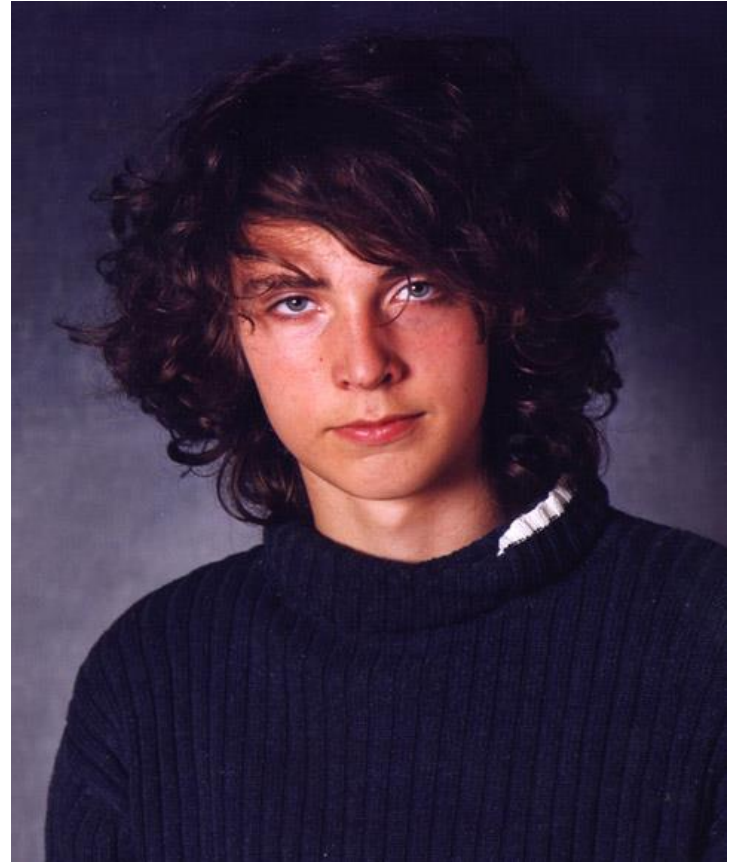
Curricula in hemophilia

Disclaimer: This curricula was realised with the EEA Financial Mechanism 2014-2021 financial support. Its content (text, photos, videos) does not reflect the official opinion of the Programme Operator, the National Contact Point and the Financial Mechanism Office. Responsibility for the information and views expressed therein lies entirely with the authors.

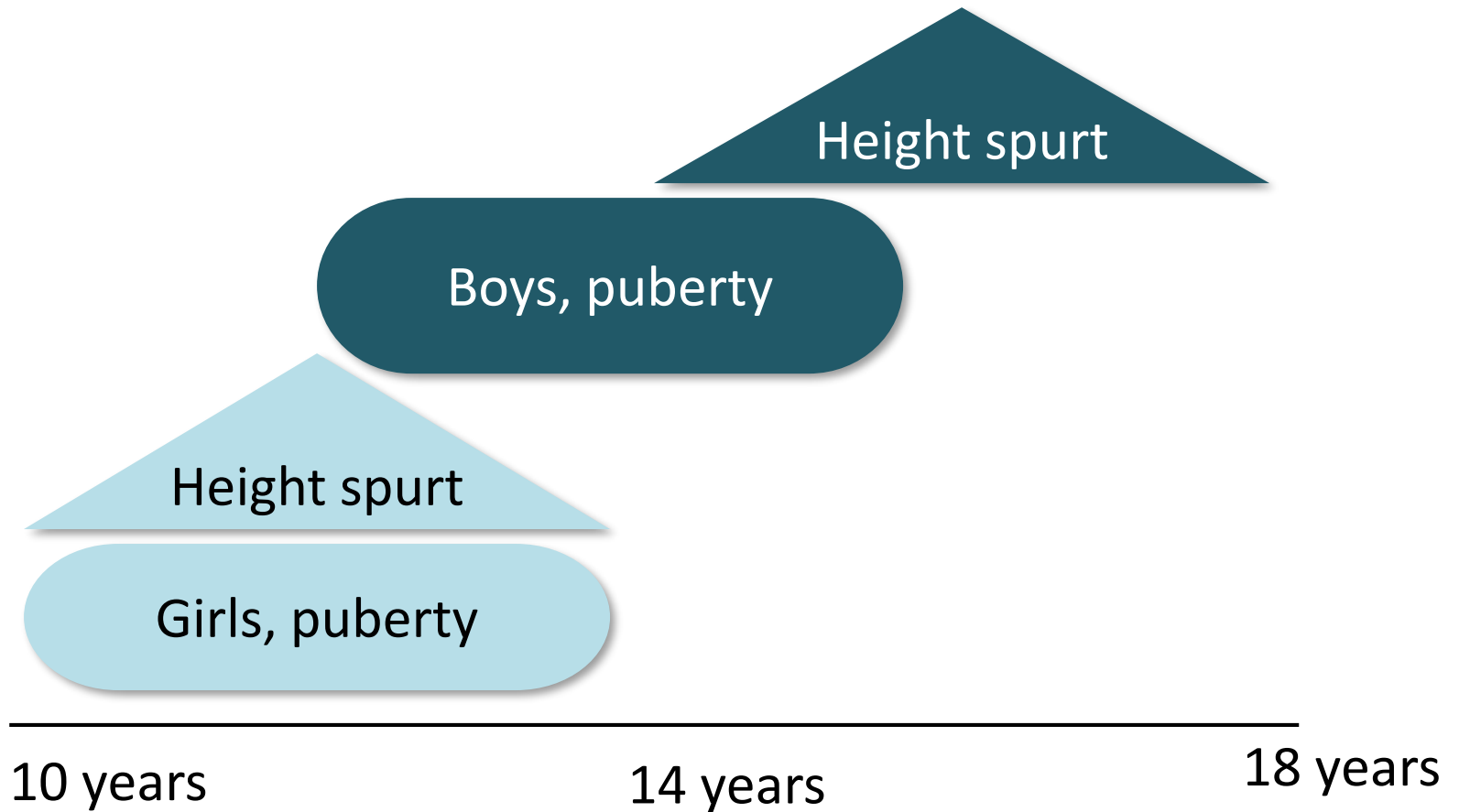
Haemophilia care in adolescents



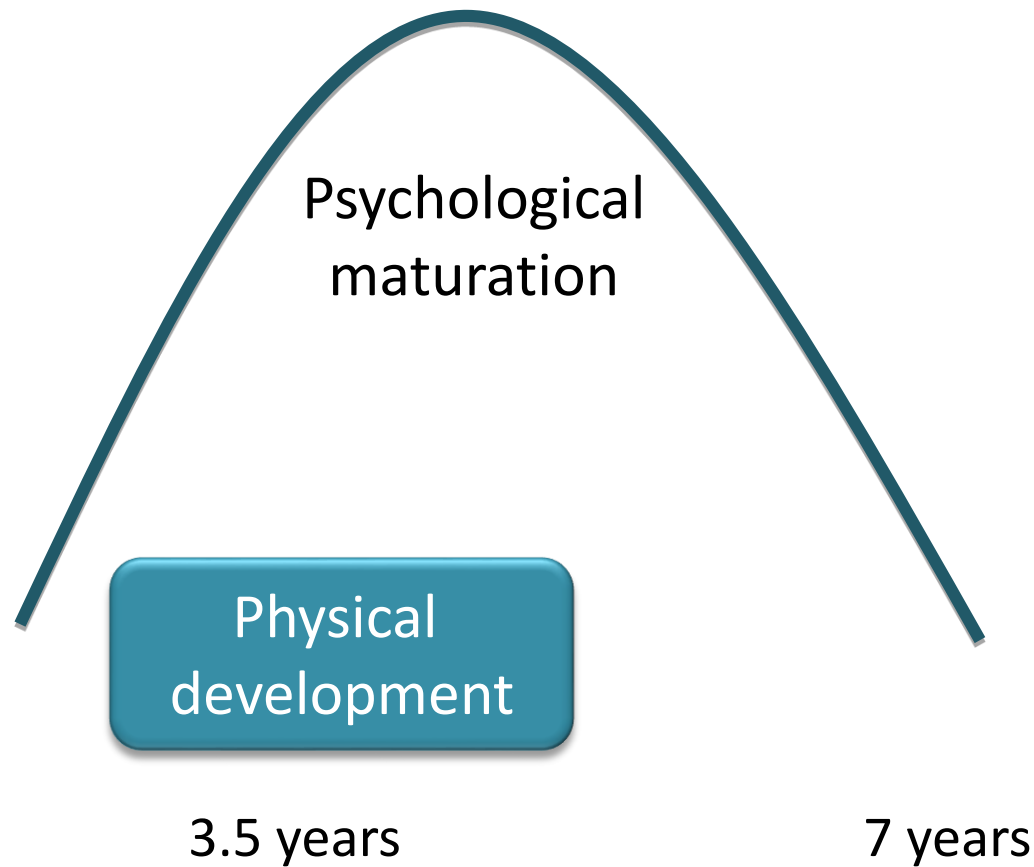
Where do all the boys go ?



Average development of growth and puberty among healthy girls and boys



Schematic presentation of the average duration and interrelation of pubertal development and psychological maturation



Psychosocial development

Psychosocial regression

Testicular growth

Penis

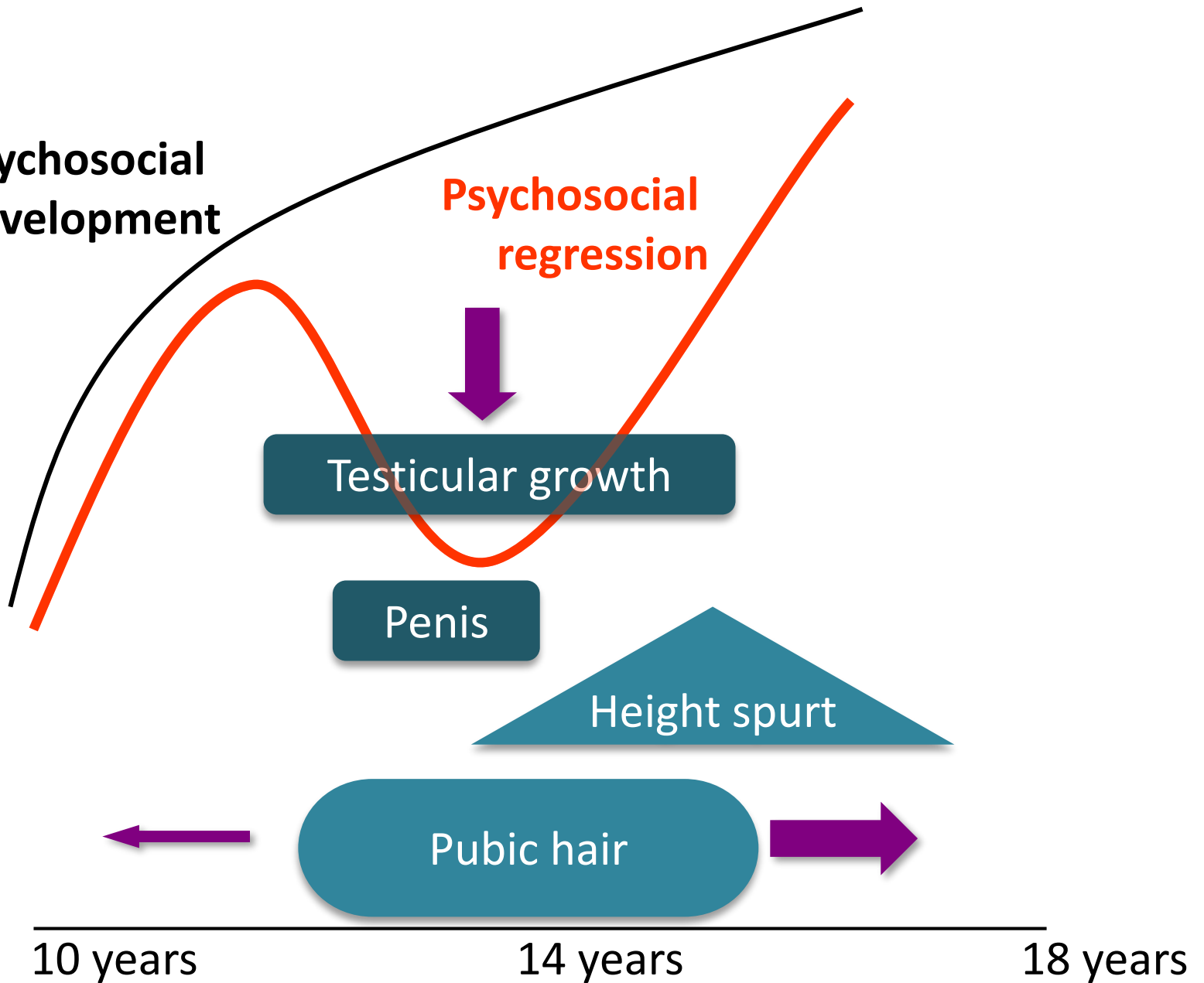
Height spurt

Pubic hair

10 years

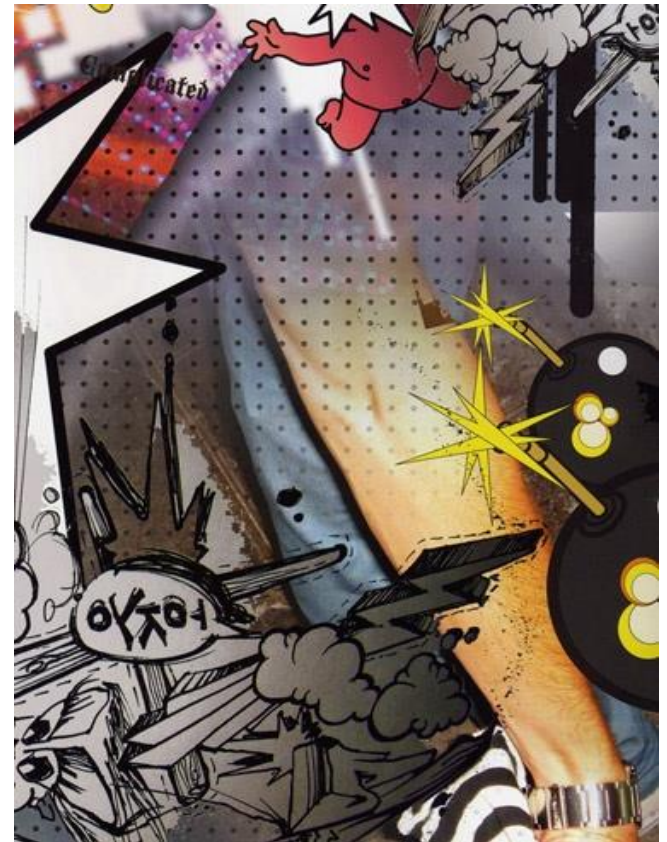
14 years

18 years



Psychosocial regression

- Childlike features are again emphasized
- The use of language changes
- Academic performance at school slows down
- Several consequences at home



Different and changing needs

Adolescents (age 10–24) are a diverse population group

- A time of rapid physical and psychological (cognitive and emotional) growth and development
- A time in which new capacities are developed
- A time of changing social relationships, expectations, roles and responsibilities

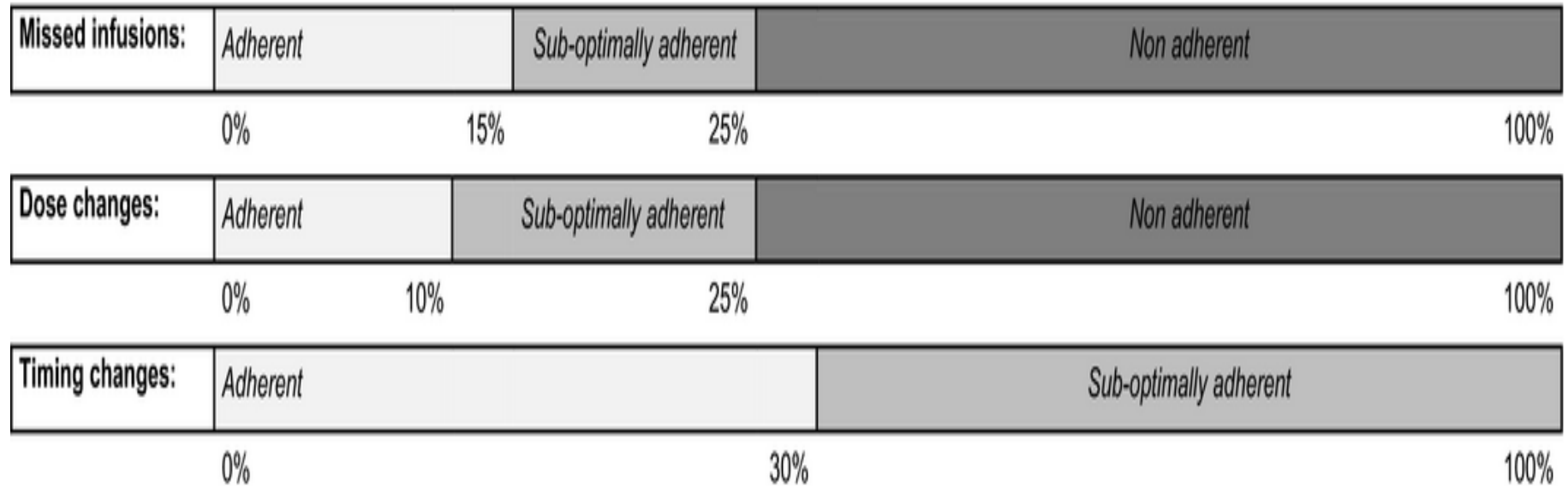


How do we measure treatment in "real life"?

Adherence - the obedience of the patient to the medical advice

Compliance - describes the degree to which a patient correctly follows medical advice. Most commonly, it refers to medication or drug compliance.

Defining adherence to prophylaxis in haemophilia



Assessment over a period of 4 weeks: prescribed versus taken infusions, extracted from infusion diary.

Voting question

Which of the below statements do you think best describes adherence to medication when a child with haemophilia becomes an adolescent?

A) Adherence improves

Getting older means you become more sensible and understand your disease better.

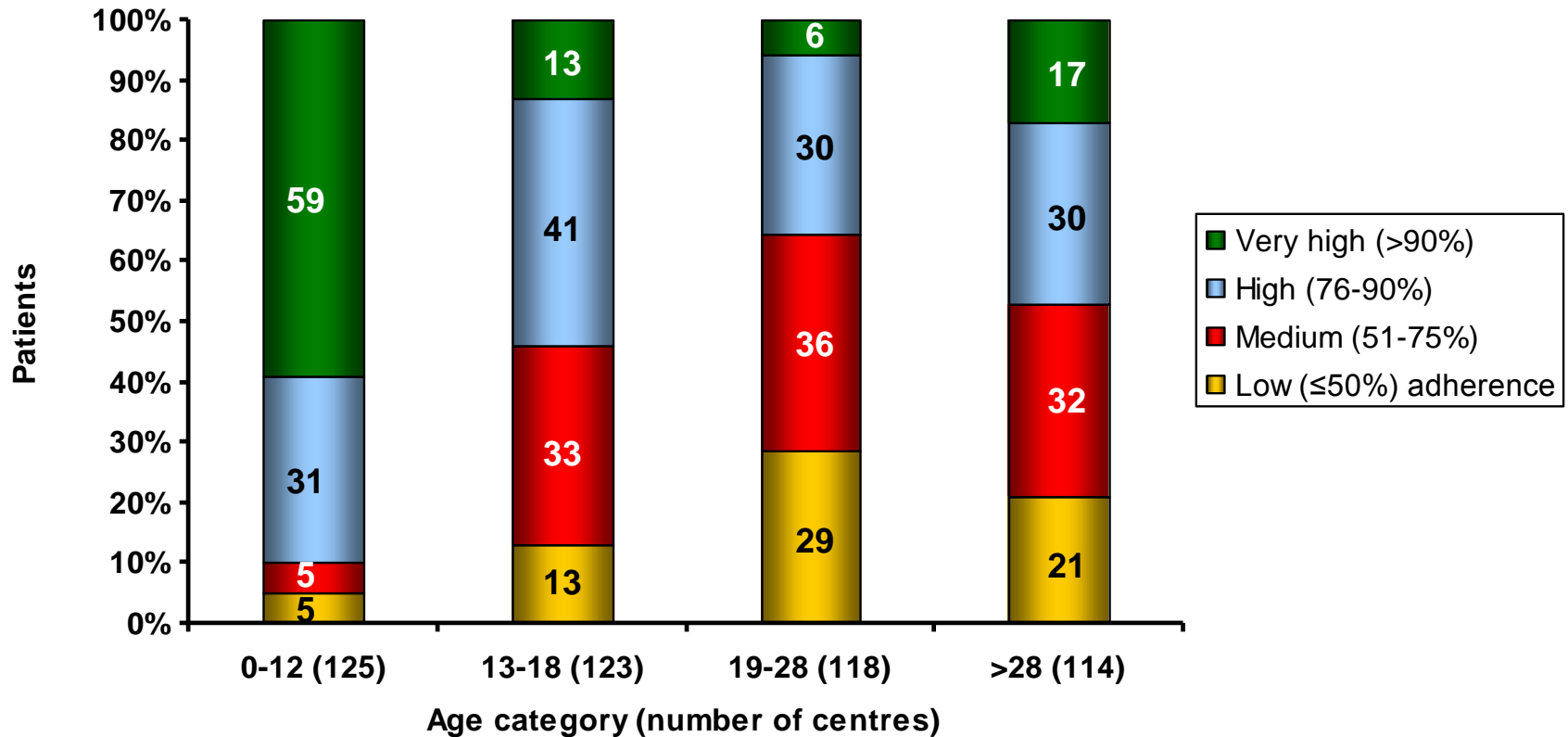
B) Adherence doesn't change

You have learnt from your parents how to manage the disease and therefore continue in the same way.

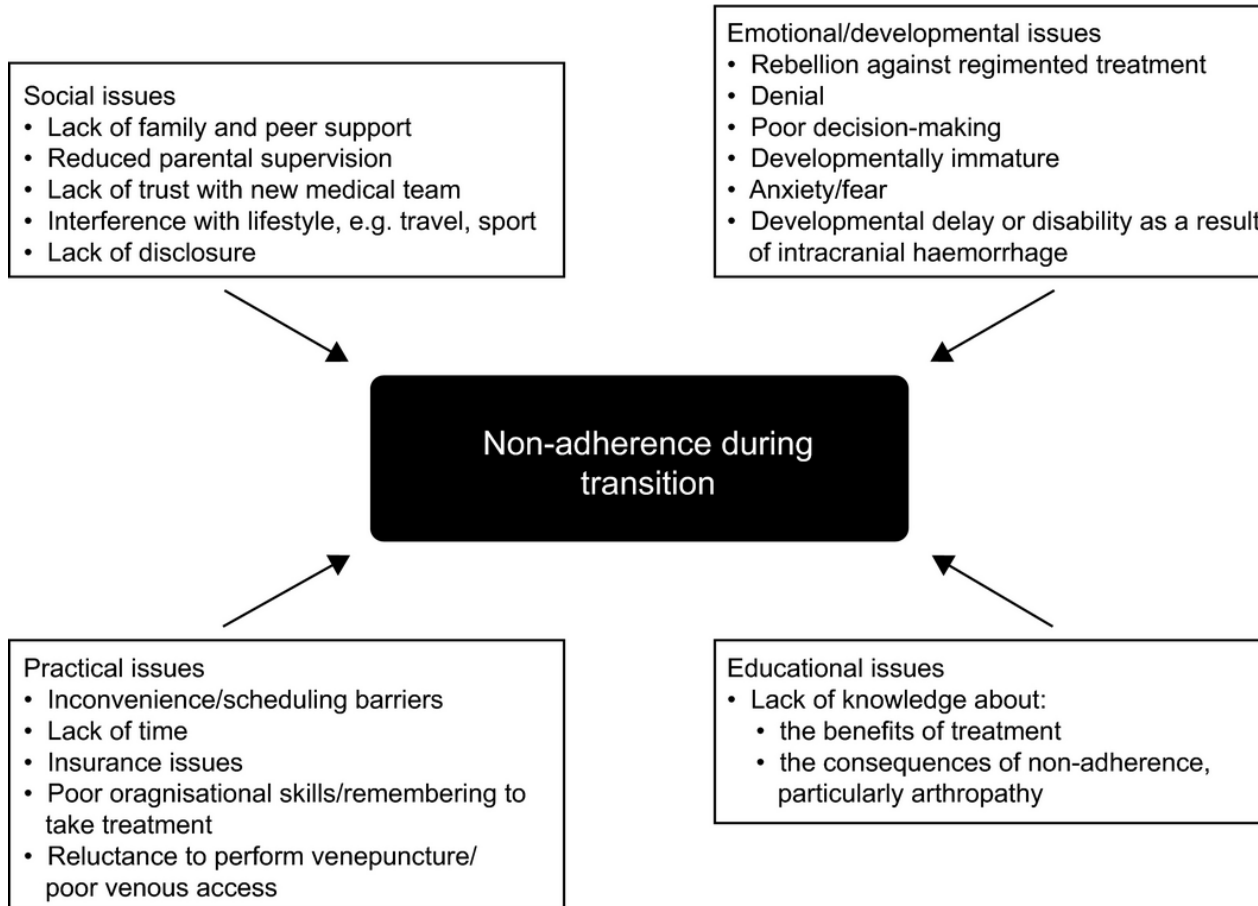
C) Adherence decreases

Physiological and psychological changes make it more difficult to maintain a demanding treatment regimen.

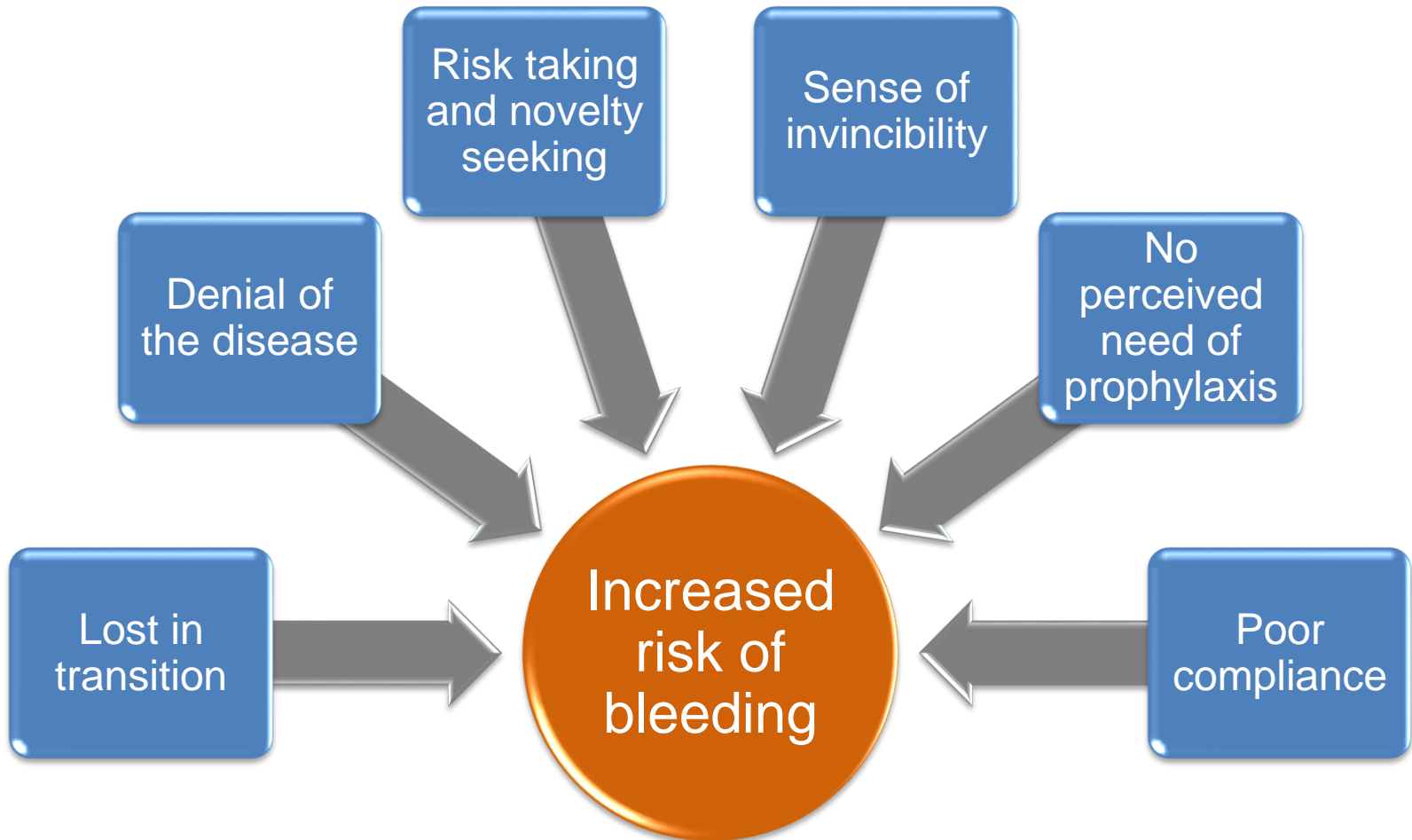
Adherence to therapeutic regimen by age category



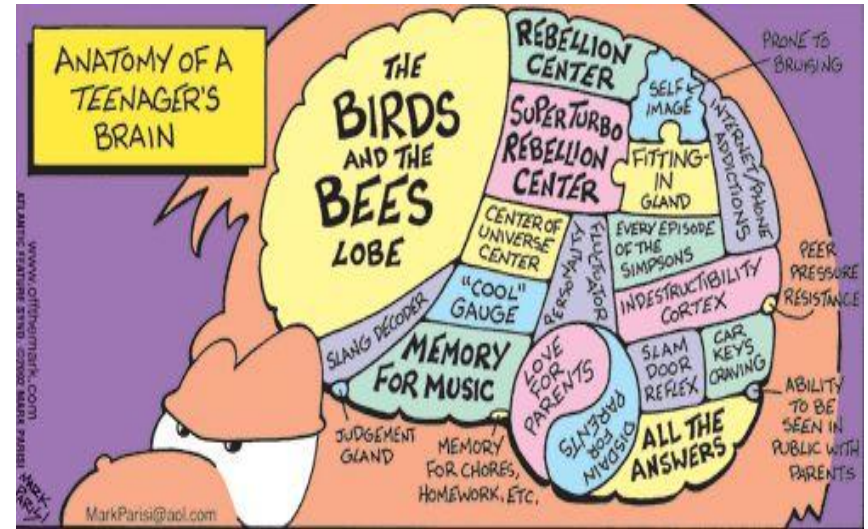
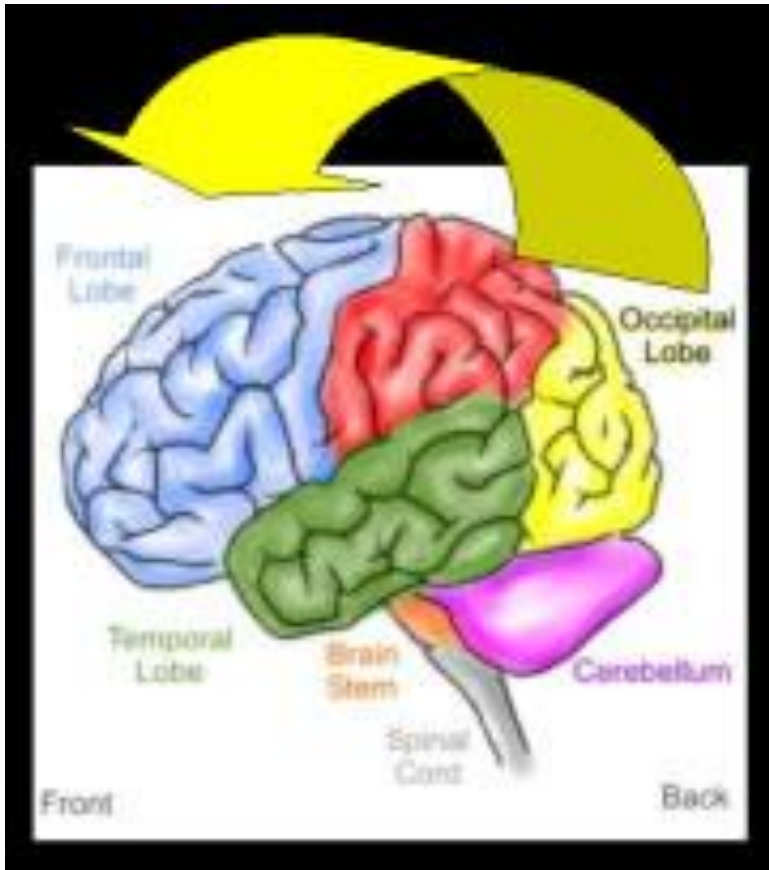
Challenges in the management of haemophilia on transition from adolescence to adulthood



Haemophilia in adolescents



Brain maturation during adolescence



M

(born 1989) – history

Diagnosis

- No family history of haemophilia
- Diagnosed after birth – subgaleal haematoma
- Blood transfusion
- FVIII <1%

Treatment

- Prophylaxis: 2 years
- Home treatment: 3 years, 500 IU 3x per week
- 0–2 ankle bleeds per year
- Self infusion: 15 years

Social and sporting activities

- Attended summer camp for PWH twice
- Gymnastics
- Some problems with bullying at school

18 years

Treatment

- Transition to adult centre at 18 years of age
- Prophylaxis treatment 1500 IU (25IU/kg) e.s.d
Reduced to 1–2x per week
- HJHS: 0

Social and sporting activities

- Lives with girlfriend
- Parcour

Follow-up Adult center

- Follow-up visits cancelled

Follow up – age 19 years

- Prophylaxis
- 2000 IU 1–2 times/week?
- 27 IU/kg
- No treatment logs
- 2 traumatic elbow bleeds
- 1 ankle bleeds
- 1 muscle bleed
- 2 visits to emergency department
 - no FVIII at home
- FVIII peak: 57%
- FVIII trough: <1%
- HJHS
- Ultrasound
- MRI
- QoL

Yes, I know I take risks!

HJHS? US? MRI? Nothing wrong with my joints!

QoL? Never been better!

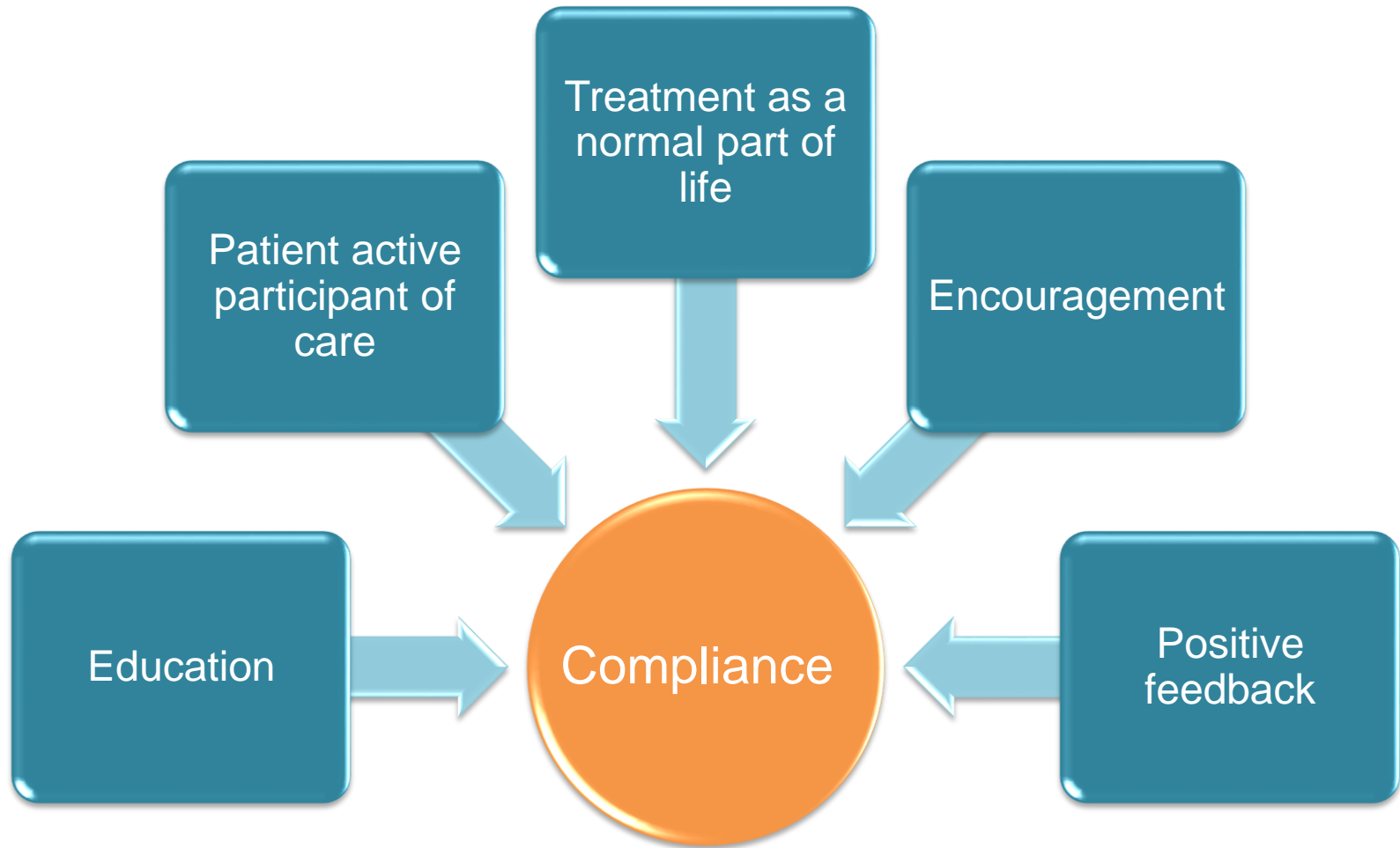


Adolescent behaviour

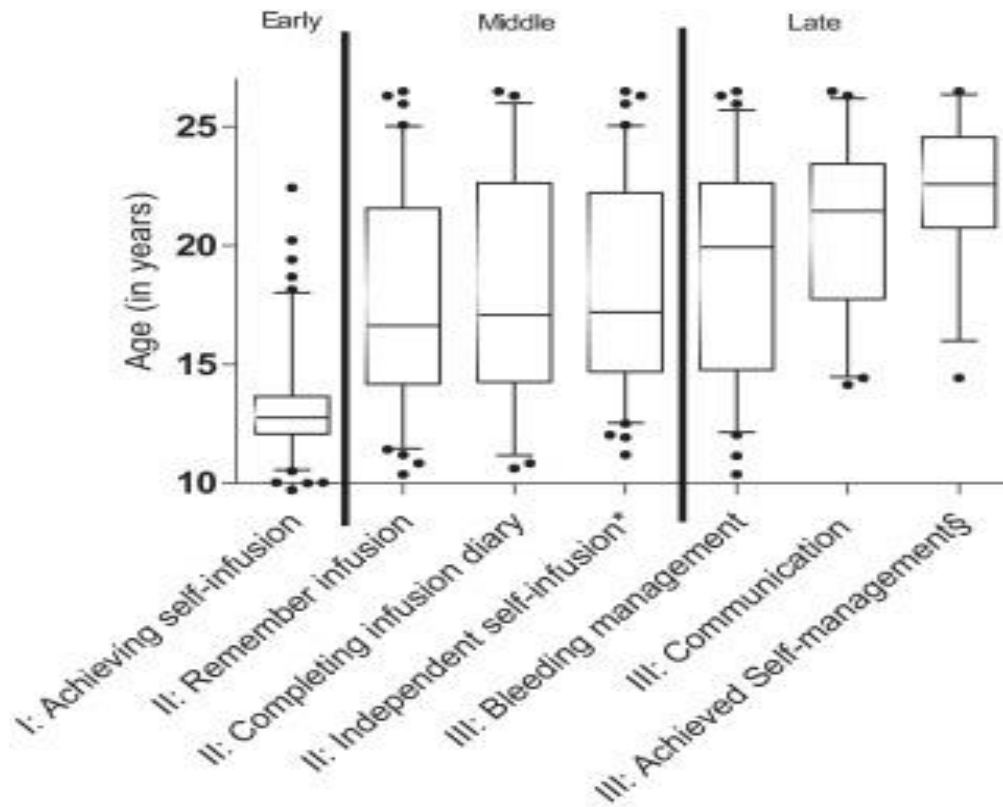
- Risk taking
- Novelty seeking
- Sensation seeking
- Emotionally-influenced behaviour

- Without consideration of future outcomes or consequences

What Can We Do To Support Compliance?



. Activities for self-management process according to age.



Voting question

A survey among persons aged 13–21 years with haemophilia asked how long they usually took before treating a bleeding episode. Approximately what percentage do you think replied that they treat bleeds within the recommended 1 hour?

A) 10%

B) 30%

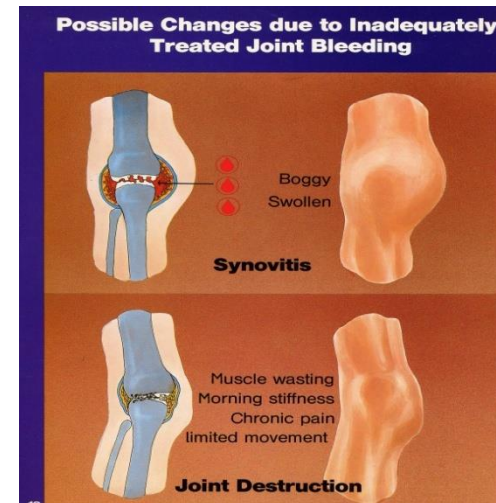
C) 60%

Barriers and perceived limitations to early treatment of hemophilia

- 31% of patients with hemophilia aged 13–21 years treated bleeding episodes within 1 hour
- 15% waited over 6 hours.
- 29% replied that they did not recognize the bleeding episode
- 11% did not think the bleed was serious enough to warrant treatment.
- likely delays arthropathic progression and disability
- improves efficacy at stopping bleeds
- results in the use of less product and is more cost-effective
- delays or prevents the need for more expensive interventions later.

[Saxena K Blood Med.](#) 2013

Centers for Disease Control and Prevention (CDC) 2009



Sports and physical activity in adolescence with Haemophilia

Pros

- Better health status
- Less risk of obesity
- Less risk of osteoporosis
- Better QoL
- Reduced number of bleeds??

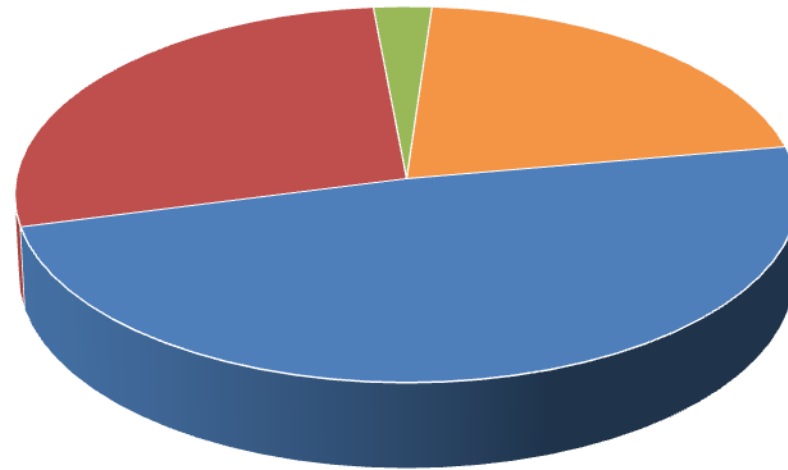
Cons

- Trauma / bleeds
- Costs / extra CFC



The impact of sport on children with haemophilia

- 84 boys aged 6–17 years with haemophilia A or B, any severity
- $\frac{2}{3}$ on prophylaxis



- 90.5% did sport
- Average 4.9 hours per week

■ once/week

■ twice/week

Organized sports participation and the association with injury in paediatric patients with haemophilia-

Retrospective single-centre cohort study from 2008 - 2010 in subjects ages 10–18 years with a factor VIII (FVIII) or FIX level <40%.

- 77 reported injuries in 36/48 (75%) subjects. (mean 1.6 ± 1.5).
- 40.% soft tissue,
- 40.% haemarthrosis,
- 14.% muscle haemorrhage
- 5.% head injury without intracranial haemorrhage.
- All subjects with severe haemophilia were prescribed prophylaxis
- There was no statistical difference in the mean number of injuries or target joint formation between the subjects who participated in organized sports compared to those who did not. during the study period.

1..	Risk category	Pat nr
Golf	1	2
Swimming	1	6
Rowing	2	1
Tennis	2	2
Baseball	2.5	7
Basketball	2.5	12
Martial arts	2.5	2
Gymnastics	2.5	1
Soccer	2.5	4
Track and field or cross country	2.5	9
Volleyball	2.5	1
Hockey (ice or street)	3	2

Diagnosis

- No family history of haemophilia
- Diagnosed at the age of 10, haematomas
- FVIII <6%

Treatment

- On demand
- DDAVP (nasal spray fVIII 25%) and rFVIII
- A few trauma related bleeds

Social and sporting activities

- Go secondary school which has a sport profile
- Wants to become a professional in handball player
- Lots of friends, most of them from the handball club

Treatment

- Major bleeding in right thigh
- Intensive FVIII treatment , rest and rehabilitation.
- Relapse after 3 months.
- Two more muscular bleeds within a year

What do you recommend ?

- He is devastated. Sports is his dream. Life is meaningless without it !

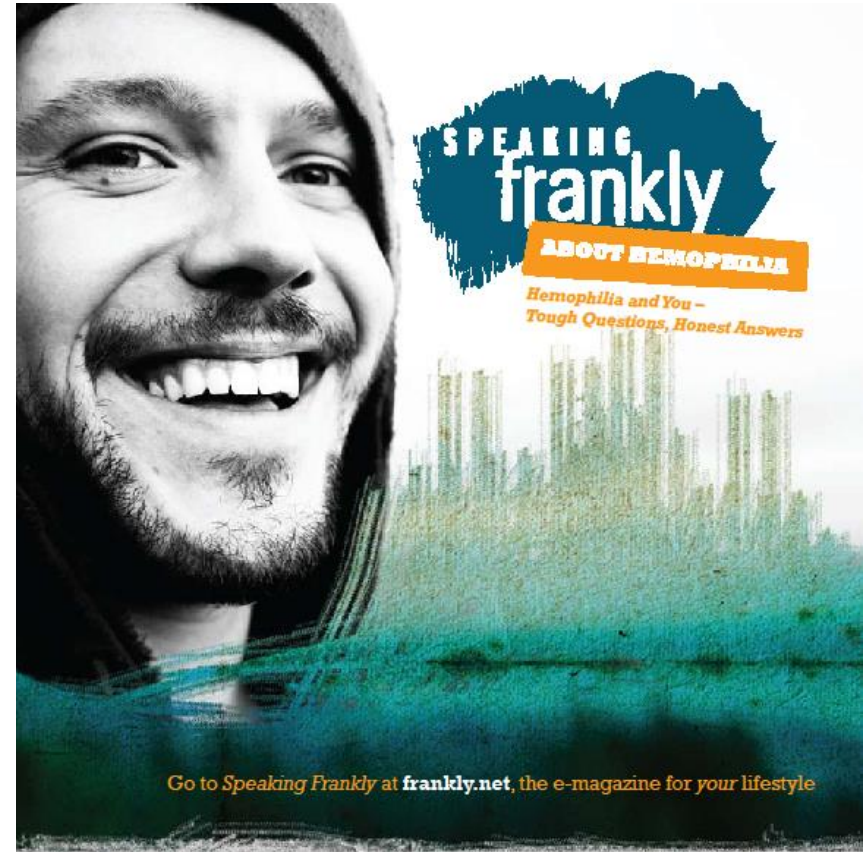
Voting question

If patients has access to CFC-
What do you recommend?

1. Stop handball playing
2. Recommend prophylaxis before sport activity
3. Recommend secondary prophylaxis

Support network of adolescents with a chronic disease

- Parents
- Peers –
Fellow sufferers
Healthy friends
- School
- Health care providers
- Internet



Transition is a process not an event!

- Assess in detail his knowledge about his disease and treatment (management skills, self-treatment and self-care).
- Assess the impact of haemophilia on his daily life and future plans in a personalized context
- Achieve his involvement in the control of haemophilia and the knowledge of his rights and responsibilities, stressing benefits and risks
- Promote effective communication of his demands and needs, so these can be adapted to an effective treatment
- Work with parents. They should understand that transferring responsibility does not mean fading into the background, but rather taking a secondary role for the benefit of their son

Nothing new !

- Aristotle: “youth are heated by Nature as drunken men by wine”
- Socrates: “inclined to contradict parents and tyrannize their teachers”
- *Apter : Adolescence is society's permission slip for*
- *combining physical maturity with psychological irresponsibility”*

