Patient with factor H deficiency associated hemolytic uremic syndrome looses fourth renal allograft –

What comes next?

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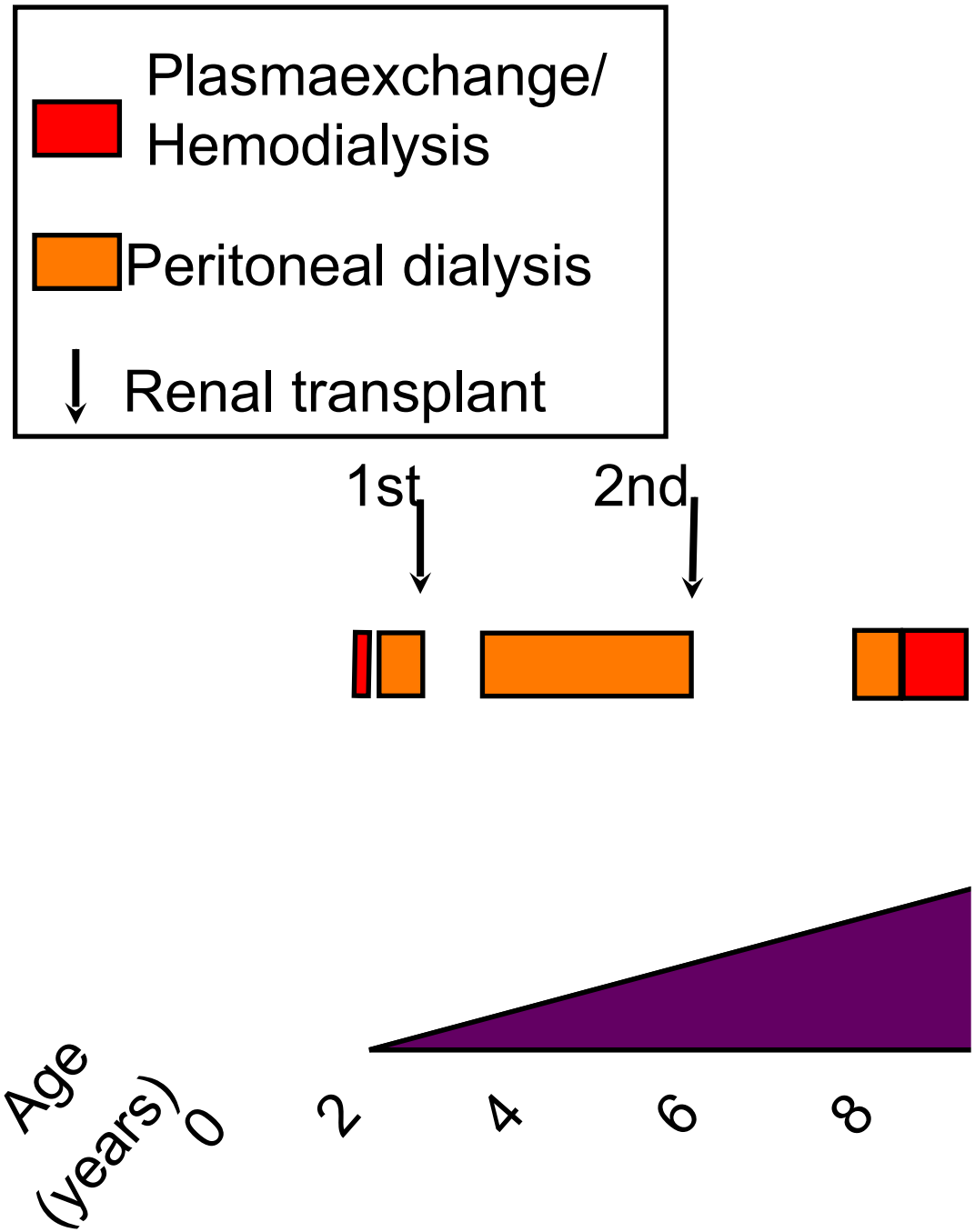
Introduction

- At 2½ years, patient M presented with D(+) hemolytic uremic syndrome (HUS)
- Shigatoxin negative

- microangiopathic hemolytic anemia
- thrombocytopenia
- renal impairement

- In most cases of D(+) HUS renal replacement treatment only needed short term

Case Presentation



- 1 yr old sister died from HUS when M. was 6,5 yrs old
- Heterozygous mutation in complement factor H gene → familial HUS

- recurrent peritonitic episodes (bacterial and fungal)

Factor H deficiency associated HUS

- Plasma protein made in the liver
- Involved in complement regulation
- Plasma exchange as treatment to replace mutated factor H only partially successful
- Renal transplant as treatment of ESRD but not HUS, high risk of disease recurrence

Case Presentation

Plasmaexchange/ Hemodialysis

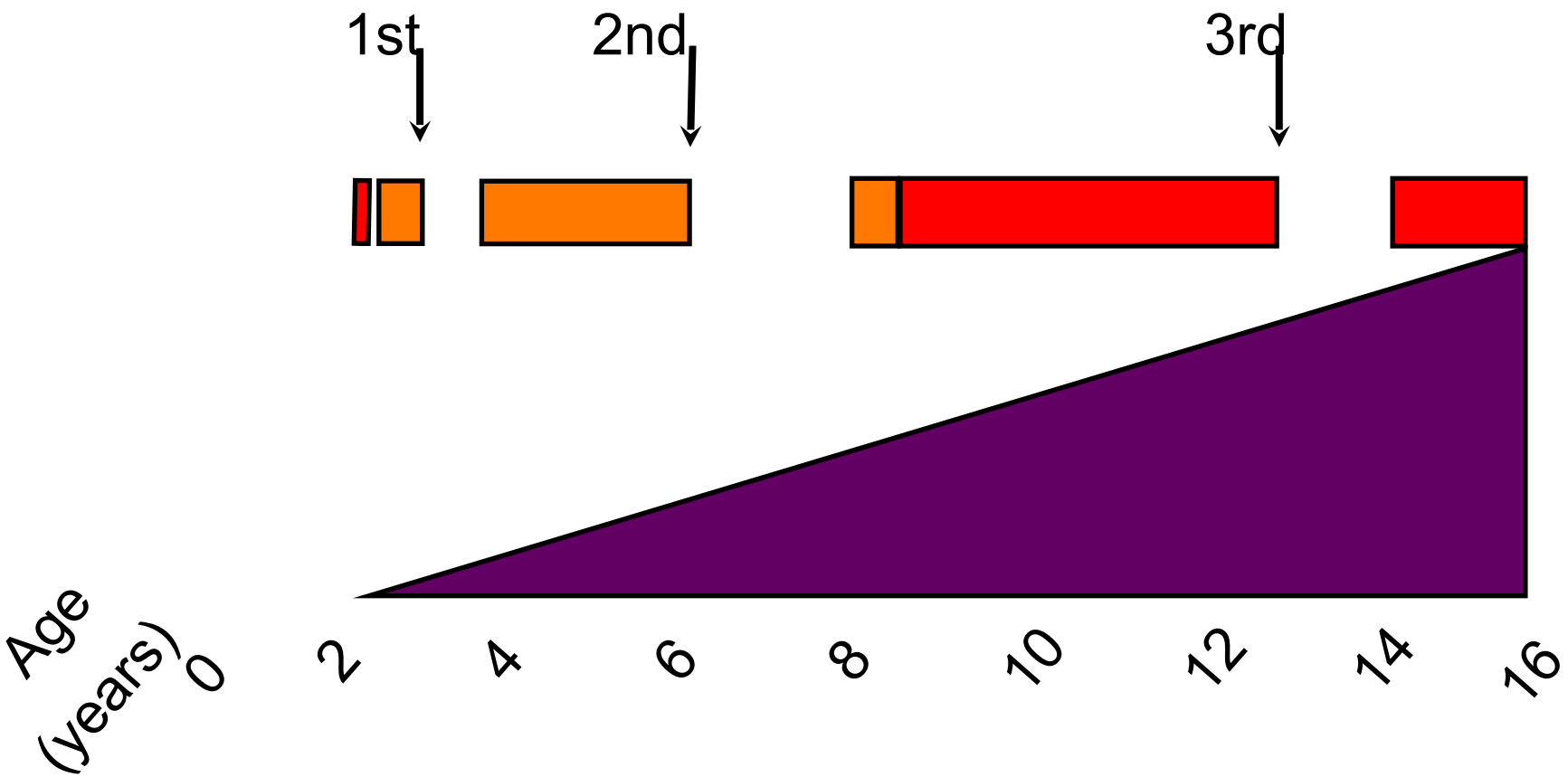
Peritoneal dialysis

Renal transplant

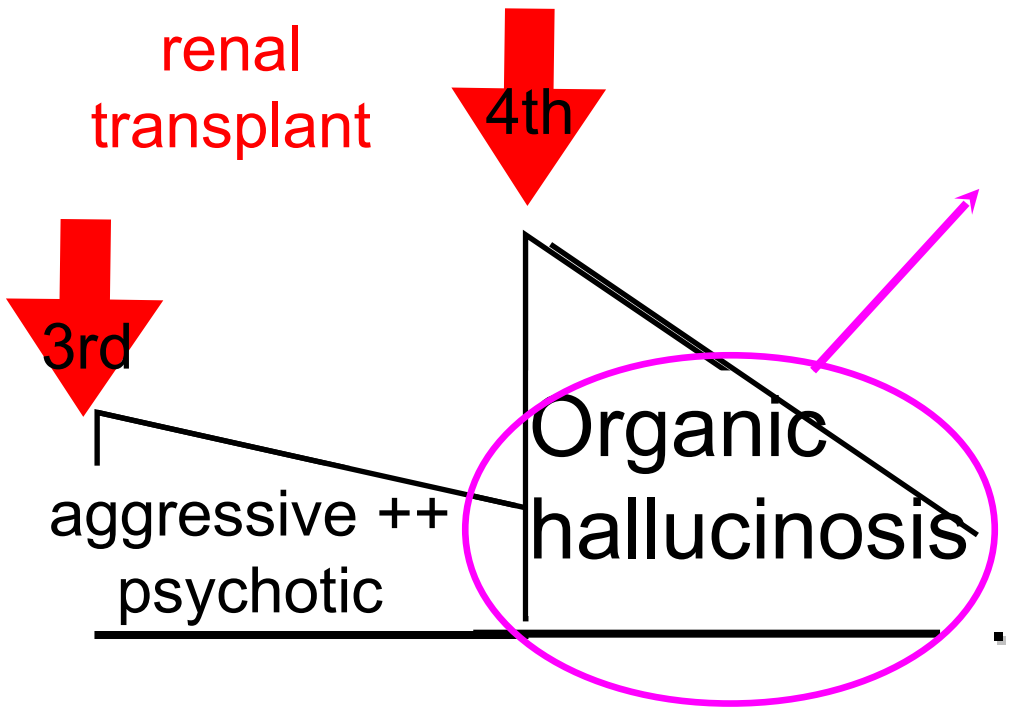
High allo-antibody levels

<u>After third renal transplant:</u>

- Deafness
- Change in behavior: ↑aggressive



Psychiatric complications



? Caused by:
- Prolonged ICU stay (sedatives/narcotics)
- Immunsuppressive treatment (steroids)
- HUS

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Magical Thinking (Mangas, computer)

Impulsive/ aggressive

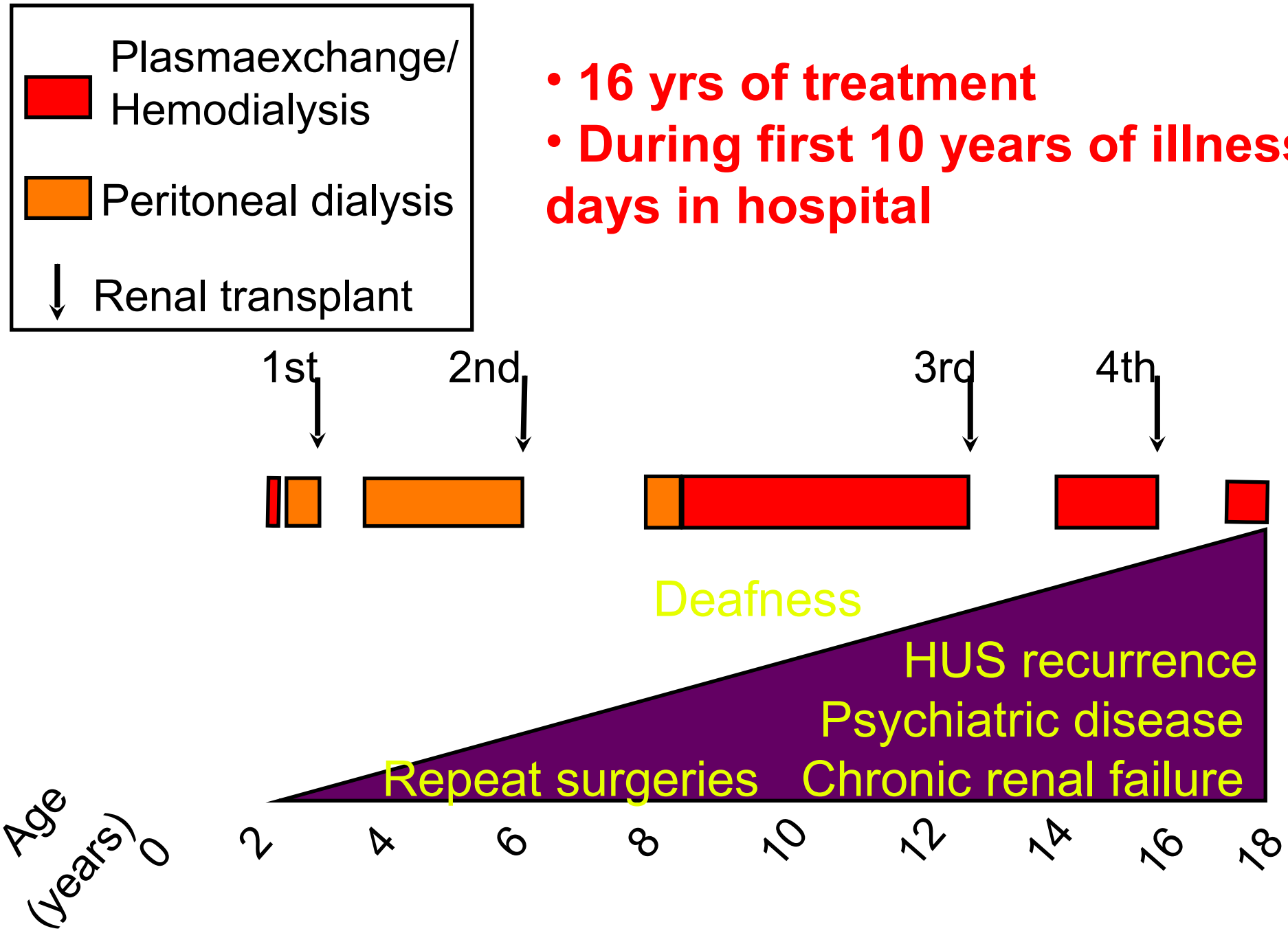
Family Background

- Working class family
- Eldest of three siblings (two sisters)
- Little died from same disease,
- Frustration and aggressions built up within family, hardly accessible by counseling
- Compliance difficult

Psychosocial Background

- Irregular schoolvisits, since the age of 13 school for deaf children
- Communicates mainly by sign language and written words
- Cognitive development delayed, is able to understand illness
- Suicidal tendencies (‘I dont want to live anymore’) with excessive drinking despite fluidrestriction since after 2nd transplant

Case Presentation



- **16 yrs of treatment**
- **During first 10 years of illness 700 days in hospital**

At present

- Living with parents and sister, living in own house
- Transferred to adult service
- Hemodialysis 3 sessions/ week
- School for deaf children
- Olanzapine
- No counselling

Conclusion

QUESTIONS

- Longterm hemodialysis as nocturnal home hemodialysis until other options available?
- Combined liver-kidney transplant?
- Is the additional psychiatric disease per se contraindication for another transplant?

QUESTIONS - **general**

- How many transplants should be performed in patients with recurrent disease (HUS/FSGS)?
- When does the child have the right to say ‘NO’ to have dialysis or another transplant?
- How can we optimize psychosocial care for children with recurrent diseases and their families?
- Are there special aspects of transfer to adult services in these patients?