

# CASE PRESENTATION

REHABILITATION CENTER PULDERBOS  
BELGIUM



Leen Govaerts  
CF NURSE

# CASE 1



# Medical history

Boy

Born at 37 weeks: BW 3,125 kg

48h after birth: meconium ileus, abdominal surgery in 2 times (abdominal stoma)

CF diagnosis by sweat test and genetics; homozygote dF508

Difficulty weight gain in the first month, mothermilk with additional calories

1 month

2 months

# Medical history

1st respiratory exacerbation  
oral antibiotics and every 1,5 hours feeding

Hospitalisation IV treatment AB 10 days

Hospitalisation intestinal  
obstruction (MIE)  
2nd respiratory exacerbation  
start oral AB

Hospitalisation for IV AB  
E coli, P putida

3 months

4 months

5 months

6 months

7 months

# Medical history

Question for submission inpatient rehabilitation

- Demand from CF center and parents
- Respiratory deterioration and difficult parent-child relationship
  - Mother : Fear of evolution to depression 'having a break down...'
  - Father : Engaged but not a lot actually present
  - Siblings: Sister of 4 years old with intermittent respiratory infections

Transfer from hospital to Pulderbos

7 months

8 months

9 months

# Inpatient rehabilitation - Pulderbos

1st day after admission

fever, cough +++, mucus+++, anorexia

After two days of intensive respiratory physiotherapy without improvement

Therapy changes

- ❑ Oral AB 14 days
- ❑ nasogastric tube
- ❑ mucusdrainage with PEP

Medication: Creons, Vitamines, Ursodeoxicholic acid

Inhalation therapy: Intermittent Salbutamol and Fluticason

Aerosols: 3 x mucoclear 7%, 1 x pulmozyme, 2 x colistineb

Respiratory physiotherapy 3 times a day

**Big therapy burden**

8 months

9 months

10 months

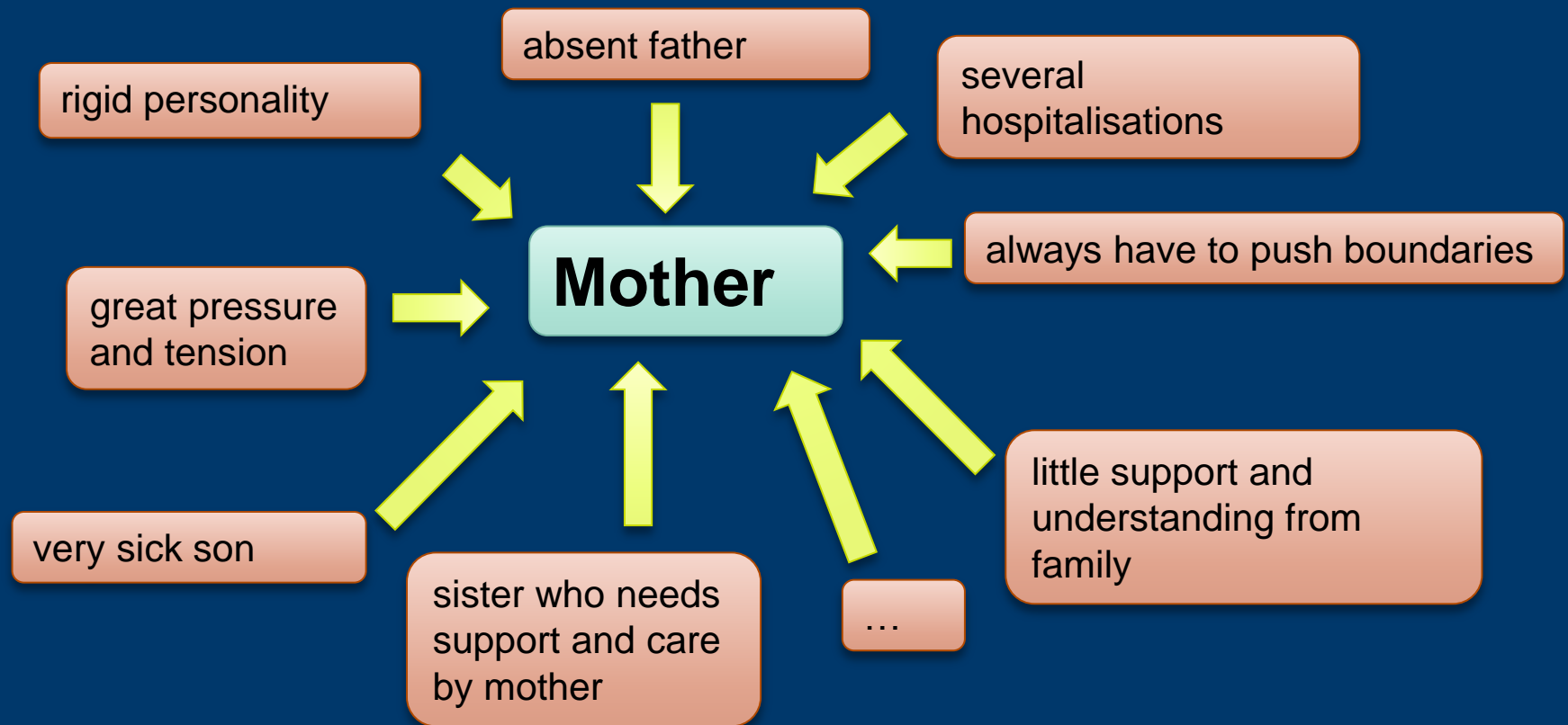
# Aims and goals of inpatient rehabilitation

## ➤ Optimisation therapy

- ❑ Optimize therapy and medical treatment
- ❑ Optimize and stimulate oral medication 'appelsauce'
- ❑ General and specific motor development
- ❑ Optimize hyper-caloric diet
- ❑ Reduce tube feeding

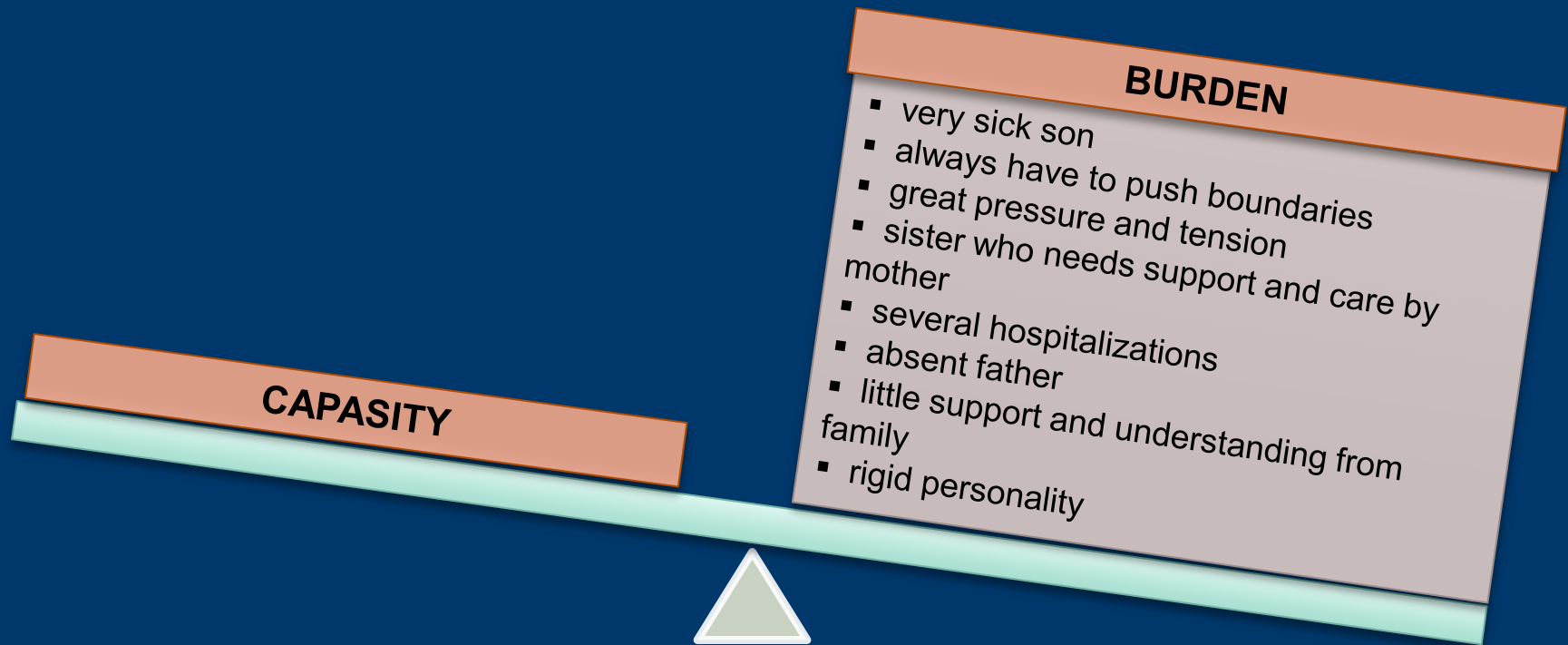
# Aims and goals of inpatient rehabilitation

- Assessing the social situation and possibilities for coping for the mother





# Aims and goals of inpatient rehabilitation



**Out of balance**

# Aims and goals of inpatient rehabilitation

- ❑ Optimisation therapy
- ❑ Psychosocial and familial aspects :
  - ❑ difficult parent-child relationship
  - ❑ **big therapy burden**

## Need for :

- Teaching parents
- supporting Parents
- Building a safety net, home guidance
- ...

# Points of discussion



- Psychological decompensation or are we overasking the parents?  
How hard is it to see everything in proper perspective?
- The need for education and observation of the child and parent in the diagnosis of CF

# CASE 2



# Medical history

## Data on admission in Pulderbos

- 9 year old girl
- CF, dF508, meconium ileus at birth
- Chronic bronchusobstruction and bronchiectasia
- Chronic colonisation Pa and multiresistant *Achromobacter* *Xylosodans*
- Frequente respiratory excacerbations
  
- Longfunction: FEV1: 25 – 35%
- Conditional decline
- Failure to thrive, anorexia and vomiting
  
- Cyanotic, O2 saturation 87%
- Clubbing, thoraxhyperinflation
- Dyspnoe, tachypnoe
- Diffuse crackles

# Aims and goals of inpatient rehabilitation

- respiratory rehabilitation
  - Intensifying respiratory physiotherapy
  - Optimize and fine tuning medication
  - Start oxygen therapy
- Approach to nutrition problems with failure to thrive
  - Optimize and fine tuning medication
  - Maximise calories
  - Tailoring schedule tube feeding
  - Stimulation of oral feeding
- Improvement of the physical condition

# Interdisciplinary approach

- Continuous nursing and pedagogic support
- Age appropriate self-reliance, taking into account her physical capabilities
- Providing school during the admission
- Guiding parents and other caregivers
  - Dealing with negotiation and behavior
  - Teaching
- Striving for reintegration at home

# Challenges and interventions in cf care Physiotherapy

## Challenges

### Respiratory physiotherapy

- Many viscous sputa
- Cough and vomiting mucus
- Dyspnoea
- Insufficient mucus draining through lack of strength

### Exercise physiotherapy

- Muscle mass and endurance

## Interventions

### Respiratory physiotherapy

- Intensive respiratory physiotherapy and drainage
- Change aerosol machine (Akita) → because of inefficient breathing
- Start Bibap during physiotherapy

### Exercise physiotherapy

- Controlled exercise with oxygen
- Use of wheelchair during (longer) distances
- Daily moments of rests with Bipap



# Challenges and interventions in cf care medication

## Challenges

- Continuous fine-tuning medication

## Interventions

- Continuously adjusting inhalation medication
- Decreasing corticoids
- Antibiotics systematic
- Intermittent IV AB therapy

# Challenges and interventions in cf care feeding

## Challenges

- Less appetite because of her CF
- Picky eater
- Many mucus and coughing while eating
- Fatigue during meal

## Interventions

- Increasing food supply, maximize calories
- Positive stimulation and no pressure on quantity
- Adjust nutrition and physiotherapy, search for the optimal moment
- Help with meals and custom food
- Fractionated administering tube feeding

# Challenges and interventions in cf care emotional

## Challenges

- Hard and time-consuming daily therapy
- Little understanding well being and health
- Enormous pressure on nutrition (from parents)
- Lots of negotiating about therapy and nutrition (mainly at home)
- Difficulty to recognize borders

## Interventions

- Take away the pressure of the meals
- Make therapy duration achievable
- Educating child and parents

# Challenges and interventions in cf care sleeping

## Challenges

- Dyspnoe and coughing at night + vomiting because of lots of mucus
- Enuresis nocturna

## Interventions

- Oxygen and intermittent Bipap during sleep
- Enuresis nocturna : no priority because of her medical conditions

# After resignation Pulderbos

- Further deterioration
- Admission to hospital
- Signed up for lungtransplantation
- Pretransplant she came on the intensieve care unit (on a ventilator and eventually ECMO)
- She has been transplanted at the age of 10

# Points of discussion



- You can not always achieve all goals and aims
- Continuously fine tuning goals and aims
- Sometimes it is a whole achievement to stabilize the condition



**Thank you for listening  
and  
enjoy the congress!**