

# „To treat or not to treat?“



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# „To treat or not to treat?“

- ★ About the many treatment decisions that have to be made in a life with EB and in a life as a carer or health professional for people with EB
- ★ About the difficulties to decide what is „the best“ thing to do next - and who is the one who judges what „the best“ means?

**„About the eternal search for balance  
between treatment and life“**

**(Ingrid Jageneau)**

# Treatment of EB

## ★ Requirements:

- Wound Care
- Nutritional therapy
- Physiotherapy
- Oral Hygiene
- Treatment of Pain/Pruritus
- Medications
- Psychological measures
- Operations
- ...

# Treatment of EB

## ★ Depends on the problem:

- Wound problems (infection, chronic wounds, pain, difficult area,...)
- Nutrition (Oesophageal structure, gastrostomic tube, dietary supplements,...)
- Operations (Fingers, Cancer, not EB related operations,...)
- Psychological problems (Depression, Fears, lack of self-confidence,..)
- ...

# Treatment of EB

## ★ Depends on circumstances:

- Type of EB
- Age
- Family dynamics
- Relationships
- Country/Region
- Public health System
- ...

# Treatment Plans

★ Dressing changes, wound care	2 hours
★ Nutrition	2 hours
★ Physiotherapy	1 hour
★ Oral hygiene	20 min
★ Medication	10 min
★ Psychological measures	1 hour
★ Sleep	9 hours
★ School/Work	8 hours
★ Total	23,5 hours
★ Friends, hobbies, fun, activities....	0,5 hours



# Quality of Life??

- ★ Treatment the whole day?
- ★ EB rules the whole life?
  - We all know: yes, sometimes it does
  - Effects not only the life of the people with EB, but also their families and friends
- ★ But life is (much!) more than this
- ★ Living requires time free of treatment

# Desire

- ★ To expand the living space -  
despite difficult circumstances !!

# Treatment of EB

- ★ Examples of daily life requirements
  - Wound care
  - Nutrition
  - Pain/Pruritus
  - Physiotherapy

# Wound care

- ★ Effective
- ★ As simple as possible
- ★ Few materials
- ★ Establish routines
- ★ Daily? Every second day? ...?

# Pain/Pruritus

- ★ Effective treatment tricky but essential!!!
- ★ To consider
  - Medication?
    - Be aware of side effects!
    - Dosage?
  - Non-medicinal options?
  - Role of Histamine?

# Dermasilk

- ★ Made of silk, coated with „AEGIS“
- ★ Reduces pruritus
  - EBS Dowling Meara
  - JEB
  - dDEB
  - Limited experience in rDEB
- ★ Available in different forms
- ★ Underwear makes it possible to wear nice clothes

# Nutrition

- ★ Requires time
  - to prepare meals
  - to eat/drink
  - to talk, to have fun
- ★ People with EB and their families and carers do their best, but nevertheless malnutrition is quite common
- ★ Stop feeling guilty

# Nutrition

## ★ Consider well:

- What is the attainable goal?
- Who might be able to help?
- Oesophagus constriction?
  - -> Dilatation?
- Gastrostomy?

# Physiotherapy

- ★ Professional advice very helpful and absolutely essential
- ★ Connection of therapy and fun/activity would be very welcome
- ★ Very often it is possible - even for a person with EB - to improve motoric skills with sport

# Sports/Physical training

- ★ Well known positive influence on
  - General health
  - Bone growth
  - Self-confidence
  - Fatigue
  - ....
- ★ Many objections can be found
  - Above all: risk of injury
  - Fear of overexertion

# Sports/Physical training

- ★ Consider well the pros and cons
- ★ Search for something interesting and available
  - Riding horses? Biking? Dancing? Cycle ergometer? Swimming? Dumbbell training?.....
- ★ Face the consequences!
- ★ For some it might stay unrealizable
- ★ But for many more it is possible
  - even with restrictions (such as no contact-sport, no ball-games)
  - Accompanied by someone who knows EB

# Sports/Physical training

- ★ Start low - go slow!
- ★ Stay realistic, but dare to try !

# Strategies for daily life

- ★ Establish routines
- ★ Accept imperfectness
- ★ Prioritize accordingly
- ★ Don't try everything

If you have found a good, suitable way for yourself/for your child/for your patient:  
Don't change it (without a good reason)

# Treatment challenges

## ★ Particularly difficult:

- JEB Herlitz
- Cancer
- Contracture and fusion

# JEB Herlitz

- ★ Exact diagnosis absolutely essential
- ★ Resources are limited
- ★ Time plays a special role
- ★ To consider:
  - Hospitalisation really necessary?
  - Can we make it possible for the family to be together at home, even if it is for a limited time?
  - Palliative approach from the beginning? High quality required!!

# Cancer

- ★ As far as we know at the moment it cannot be „prevented“
- ★ Our aim is to detect it as early as possible
- ★ Options
  - Operation?
  - Other options?
  - Palliative approach?
- ★ To consider:
  - Size, extent, localisation
  - Therapeutic options
  - Attitude towards life

# Contraction and fusion

- ★ Operation yes or no? When is the right time? Who can do it?
- ★ Prevention?
- ★ Other options?

# Treatment Decisions

- ★ Sometimes really difficult and tricky
- ★ Search for help
- ★ Not alone, look for a team!
  - Needed for both sides:
    - people with EB and
    - health care providers
- ★ Collect informations
- ★ Communication!

# Who decides?

- ★ Person with EB?
- ★ Doctors?
- ★ Health care professionals?
- ★ Family?
- ★ All together?

# Health care professionals

- ★ Desire to heal/help
- ★ Asked to use „Guidelines“
- ★ Should never make mistakes
- ★ Sometimes under pressure
- ★ Always expected to treat,  
to make things better
- ★ Not enough time?

# Patient

- ★ Insecurity- what will happen?
- ★ Searching for answers/solutions
- ★ Hoping for help
- ★ Full of expectations
- ★ Requires time
- ★ Wants to be seen
  - with his needs
  - AND with his strenghts

# Conflicts

- ★ Unavoidable
- ★ Between people and internal thoughts
- ★ Require:
  - Good communication!
  - Patience
  - „Good bye“ to perfectionism
  - Open eyes and ears to catch the critical moments

# Decisive factors

- ★ Health care professionals have to give clear information and facts
- ★ Professionals must take the responsibility for their advice and their decisions - but the person with EB has to live with it
- ★ Respect for autonomy

# Requires

- ★ Respect for both sides
- ★ Courage
- ★ Honest and sincere communication
- ★ „Mindful Awareness“ (D.Siegel)
  - „COAL-attitude“
    - Curiosity
    - Openness
    - Aceptance
    - Love/Kindness

# Common goals

- ★ Support in physical, psychological, social and spiritual aspects
- ★ Development of a reasonable yet creative life perspective
- ★ Respect for autonomy and self-determination
- ★ Discovery of useful coping strategies

# Good „copers“ ....

- ★ Are full of ideas
- ★ Are flexible
- ★ Don't deny
- ★ Act on the basics of reality
- ★ Re-define their problems to find solutions
- ★ Consider alternatives
- ★ Run an open communication
- ★ Search for help - and take it
- ★ Keep high moral standards
- ★ Don't give up hope

(Weisman 1979)

# Take time

- ★ Don't hurry when you have to make difficult treatment decisions
- ★ „People forget how fast you did a job - but they remember how well you did it!“  
(Howard Newton)

# „To Treat or not to treat“

- ★ Wrong question!
- ★ Better: „What is the suitable therapy for this special person with this special life in these special circumstances?“

**„Treat-Life-Balance!“**

# Creative life

★ „To live a creative life we must loose our  
fear of being (doing) wrong.“

(Joseph Chilton Pearce)

# Thank you!



Willkommen im  
*eb-haus Austria*

