

„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
 - Acceptance
 - 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
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