

Living with EB - Impact on Daily Life

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1. Introduction

EB is a lifelong challenge, varying not only in the severity of the disease but also by whether secondary complications are present; for example, musculoskeletal deformities or problems with food intake. Speaking of patients as having mild forms of EB is not meant to trivialize their disease. Rather it means only that those children or adults have problems that are oftentimes easier to solve. Problems caused by the disease can be small or very stressful, starting with anxious looks which are not meant to be mean or judgmental, but still are very hurtful and at times can be malicious. Such unintentional behavior by others can result in an EB patient becoming more and more subdued, avoiding contact with others. Information and explanation about the disease and problems associated with it can lead in many cases to respect and understanding for the person, which he or she rightfully deserves.

Living with a special condition like EB, yet still trying to live a normal life with its highs and lows, success and failures, wishes and dreams, strength and weaknesses, has its difficulties. Every form of EB has its own special problems. Just as every person has a different personality, one patient's disease cannot necessarily be compared to another's. In this chapter I will attempt to summarize the duties and challenges that we are confronted with in our clinic, including those that are not necessarily directly related to our patients' underlying disease.

EB patients have unexpected medical and surgical problems, as does everyone else. The presence of such illnesses, though, at times may exacerbate their underlying genodermatosis. Patients with EB, even those with relatively localized skin involvement, may experience moderate to severe pain, caused simply by standing, walking, lying down or even sitting. Symptomatic involvement within the oral cavity can impair eating, negatively impacting on overall nutritional status. EB patients can develop very painful blisters on their eyes which may prevent them from seeing for days. Of course this medical condition and its side effects have a great influence on daily life, preventing patients from participating in many activities that are usually taken for granted. Only being able to participate with limitations and often being absent from school or work can have a negative outcome. Another important factor is the length of time needed for life's daily activities, wound care, therapy and the preparation of meals, eating and bathing. Patients, therefore, must find the right balance between therapy and the care that is really needed. A daily time plan needs to be made to account for necessary therapy and hygiene, but oftentimes there is not enough time to accomplish everything that is needed. As a result, these patients and their caregivers need to learn to cut down on unnecessary actions and yet still perform the important tasks. Of course, parents and caretakers often find it difficult to make these decisions. Time needs to be found for dressing changes, as well as time for school, job, leisure time and friends, and all the little things that make one's daily life full. This is far easier said than done, but most EB families somehow find a way to meet these many needs, once they establish routines and prioritize accordingly.

2. General information

Adaptation of the home and the work place should be planned differently. In every case a quiet private space is desirable for dressing changes. This should ideally be in a bathroom with a bathtub, or shower. It is much easier and less painful for an EB patient to remove the bandages by first pre-moistening them with water. It can also reduce the amount of time required to perform this necessary daily task. The place where the person can lie down for bandage changes should be well cushioned. It also needs to be adjusted to the right height for the caregiver. To help distract the

person from this often painful and time consuming procedure, a TV, radio or CD player is often very helpful. Expensive adaptations in the home may be needed so as to accommodate the use of a wheelchair. Seating must be cushioned and skid proof padding is recommended. Other adaptative measures will need to be made according to individual circumstances.

2.1. Clothing and footwear

A very important issue is clothing. It must be very soft and have no pressure points from buttons or seams. Clothing that is too warm or too tightly fitted should definitely be avoided. Special materials that are particularly well tolerated by most EB patients are cotton, rayon and silk. Protective dressings can be placed under the clothing and over those areas that are easily irritated or traumatized, to prevent further rubbing and the formation of blisters. Unfortunately, with children it is often difficult to find clothing that is functional and yet also cool and in style.

It is particularly important to find shoes that fit properly; otherwise blisters will arise. Shoes should be neither too large nor too small, and they need to be made from a material that is both soft and sturdy. Sweating should be avoided at all costs, as it contributes to the development of blisters, especially in patients with Weber-Cockayne EB simplex. The sole of the shoe should be flat. Unfortunately, leather soles may transmit heat from pavement to the soles; they are also rather stiff until worn for prolonged periods of time. As such, they may facilitate blister formation in some children. It is oftentimes a financial burden for EB parents to find the correct type of shoes, since they are frequently quite expensive. Orthopaedic shoes are particularly good at providing support, and may be beneficial for those EB patients needing shoes, which have specially designed heels and inserts.

2.2. EB and sports

Sports and mobility are necessary for building muscle and bone, for the maintenance and efficiency of the body's functions, and for building self confidence and a positive body image. It is known that participation in sports is good for general health and has been proven to influence other coexisting illnesses. Naturally, this is also evident in people with EB. However, sports participation must be judiciously restricted, due to the risk of easy injury to the skin. Still, though, EB patients need to get enough daily exercise. This should begin in childhood, when natural mobility should be supported and encouraged. Bandages for the especially vulnerable areas should be applied, as they can be especially helpful at this time. The children should be allowed to play without too many restrictions, so that they can learn their boundaries and at the same time improve their coordination. The latter is important, since poor coordination leads to restricted mobility and can lead to insecurity and increased risk of injury.

Participation in sports is important not only in childhood. Exercise and sports should continue to be important throughout life. Which types of exercise are appropriate for someone with EB? This will depend on the extent and severity of skin involvement, as well as the child's overall health. Most types of sport can be practiced (sometimes even playing soccer and skiing), and it is appropriate to allow the child to try such an activity if it is physically reasonable and if the child can learn the limitations imposed by his or her disease. According to the severity of the disease an individual consultation is important and should be carried out.

2.3. House pets

When someone has EB there is no reason not to have a house pet (unless there is a co-existing allergy). On the contrary, the relationship with a house pet can have an extremely pleasant effect on one's overall self-esteem. The friendship, faithfulness, and love that an animal gives to his owner can help in making the difficulties of every day life more bearable. When time, money, space and patience, love and readiness for the responsibility are all available, consideration for having a house pet should seriously be considered. Particularly good is a trained guide dog, but almost every animal is a possibility.

2.4. Vacation planning

When planning a vacation it is essential to organise it properly. There are many possibilities, but ultimately one really needs to think about what would be best for both the EB child and his or her family. An ideal vacation might be one that offers something different from every day life and yet is also relaxing and stress-free. Of course this is also dependent on the financial resources available, the age of the children (both affected and unaffected), and other pertinent factors.

Of great importance is the weather. Conditions such as extreme dry heat or bitter cold can have a negative effect on people with EB. If the decision is made to spend the vacation in the southern regions or in warmer climates it is best not to go in the height of summer, when it is hottest, and to make sure that the room has climate control. A neutral climate is preferable for most EB patients although a visit with the sun, sand and ocean can help improve the health of the skin.

During city visits it is essential to find the correct transportation. Long walks are not advisable, which is why city and museum tours should be carried out only after careful planning. The time and extra help needed for the use of a wheelchair is unavoidable.

2.5. Hobbies

For those with EB whose life decisions and possibilities are restricted, hobbies may be especially important. Because of the need of a strictly organised day, a hobby offers a balance and a chance to relax. Art, drawing and painting, collecting, reading, playing an instrument, singing, and other recreational activities are all possible and are enjoyed with great pleasure.

2.6. Alternative medicine

As with other chronic illnesses that have no true cure, parents and relatives of an EB child may at one time or another look into alternative types of medicine, in the hope that they might help change the course of their relative's illness. There are indeed many unconventional approaches; which have achieved beneficial outcomes, both subjective and objective, in individual patients. In the absence of concerns over patient safety, medical and nursing personnel should be open to hearing of new therapies, including those which are currently viewed as alternative ones, to be willing to share with their patients and their families whatever information they may have regarding those modalities, and to be available to assist them in comparing the risks and potential benefits of new therapies with those currently being employed. Unfortunately, although many anecdotal reports of benefit exist,

most are based on observations made in only one or a few patients, making it impossible to critically assess their validity, efficacy, and safety. Whenever concerns arise, however, it still behooves the physician to follow the physician's creed of "first do no harm". Hopefully someday it may be possible to more rigorously compare data derived from much larger numbers of patients, to see whether any of these "alternative treatments" might significantly help in enriching the quality of life for patients with EB.

2.7. Costs

A sometimes overwhelming problem with a chronic illness is the financial burden that it brings. What resources are available to cover the costs related to the adaptation of living space, supplies, and the support given by care personnel? Where does the money come from to pay for the essential things in life or for the things that just make life easier and more pleasant? Fortunately health care by obligatory insurances is provided in Austria and is available for patients with EB. In other countries, however, this is where the problem begins, since the costs for dressing materials, frequent treatments and operations may not be fully covered by the respective health care systems. In many countries medical health coverage is not even available or obtainable. Even in countries in which the insurance system includes coverage for chronic illness, it may not cover all of the costs, forcing the family to finance at least part of it themselves. The special healthcare needs of EB patients, given how variable they may be, are not easy to incorporate into an overly structured insurance system. Therefore it is essential that insurance companies are willing to help the families in developing individualized solutions concerning both care and coverage. Costs related to enhancing the quality of life of an EB patient, as opposed to covering essential therapies, may not be covered by healthcare insurers. For instance, additional equipment needed for the household, suitable furniture, or special food and extra hours of therapy, will need to be financed by other means. Involved families need assistance in their search for all possible sources of financial help. These options may vary considerably, not only from country to country but also between one city and the other. Fortunately, in some countries, including Austria, financial help is available to EB patients and their families from local or national EB support groups. However, these organizations often still lack sufficient funds of their own to fully support the financial needs of all of their patients. It is, therefore, absolutely essential that a caring and highly trained social worker be integrated into the overall management plan of every EB patient.

3. Aged based stages

3.1. The newborn

When a baby is born and there is a suspicion that EB exists, it is a normal reaction that many questions will be asked. In the first days of the baby's life the parents will go through many hot and cold, emotional feelings. From the time the parents realize that something is not right with their baby until the time that the definitive diagnosis is made, feelings of uncertainty and anxiety will show, yet everyone reacts differently, according to the circumstances in which the child was born. During this particularly stressful period of time, the parents need to be given emotional support from everyone involved in the infant's care. As soon as the exact diagnosis is made the parents need to be informed about the special needs that the child is going to have as a result of having EB. The parents have a right to know what is going on with their baby, and details of the diagnosis should be given without delay, even if it is serious.

A proper place should be chosen by the physician to discuss the diagnosis, one in which there can be undisturbed discussion with the family. Enough time has to be planned so that all the questions that come up from the family can be answered in as thorough a manner as possible. It is likely, though, that this will most likely not happen after a single meeting. At the time when the parents first learn about the diagnosis, it is understandable that they will likely have a rather short attention span and be unable to absorb and understand all of the information that is being presented, even after they have asked and re-asked many questions. For this reason, it is very important to have more than one meeting with the family. The diagnosis that has been given to the baby, what the cause of it is, the symptoms that he or she will have and the problems that EB will likely create, should be given to the parents with empathy and honesty. At the same time it is crucial that the parents be told that it is never possible to accurately predict exactly how life will be for the particular baby, given the great variability in clinical course which is known to occur within a single EB subtype, let alone within even the same family. There should always be hope expressed! The parents must be treated with respect and given the emotional support that they will need in order to accept their child as he or she is. Comprehensive information is needed for the parents, to develop trust in their physicians, so that they can cope with the surprisingly new situation they are now in.

The baby that was born with EB or is suspected to have EB has needs like every other newborn. Feelings of closeness and security and a stimulating environment (colour; pictures; mobile) will help. Due to the vulnerability of their skin they also need meticulous medical and nursing care. Trained therapist and nursing care need to be organised. The parents need to learn as soon as possible how to care for the baby themselves, learning to give the baby physical closeness without inadvertently causing harm. An EB baby should never be lifted up from underneath the arms, as there the skin is very easily injured. It is best to use a pillow or a thick blanket to pick up or carry the baby. It is also important to avoid adherent materials. Parents will need to be educated about the most appropriate dressing materials that be used on EB skin. Adhesive electrodes should be avoided unless there is a serious need for their use. Usually, though, there are other ways to apply them without injuring EB skin.

3.1.1. Breast feeding

When the mother of an EB baby is able to breast feed it is really beneficial for the nutritional value to the child. At the same time it can also facilitate a positive bond between mother and child, which is otherwise often hindered by the stressful situation. With more severe forms of EB that are accompanied by involvement of the oral mucosa, breast feeding is the gentlest form for nutritional intake. If there is any reason that breast feeding is not possible, however, then the mother should not get stressed about not being able to directly provide breast milk to her child. A baby with EB can be fed with milk pumped mechanically from the breast. Alternatively, baby formula can be used. If the baby has symptomatic oral cavity involvement, then a special feeding nipple (Habermann Sauger®) can be employed, to reduce pain and trauma during feeding.

3.1.2. Discharge from the hospital

Discharge to the home is possible as soon as the parents feel confident in caring for the baby. It is necessary to organise transitional arrangements for them well in advance of the actual day of discharge. In the beginning the parents will need a contact person who is readily available for them at any time, should questions or problems arise. They will also need to establish a relationship with a family doctor or pediatrician in their hometown, who is willing to take over the day-to-day care of

the infant. It is important to remember that the care of a small baby with EB is labor-intensive and very time consuming. A proper place to provide optimal wound care and bandaging needs to be arranged. Whenever possible it would be helpful to have a home care nurse available to assist the parents at that particularly difficult time. Even if a nurse is available to the parents, though, close friends and relatives should all be familiar with the care needed by the child.

It is helpful for the parents to be able to contact and meet other parents of children who have EB as this is a good way to find support, both physical and emotional, during the first days at home. We have found that in these first days the greatest challenge that the parents encounter is trying to decide which problems are due to the special effects and characteristics of EB and which are simply a baby's normal needs. Good monitoring and assessment of the baby by the parents and caretakers, and communication between the parents and the doctor taking care of the baby, are essential in providing the most proper care.

The baby's bed should have a soft pad, like lambskin, to lie on, and the sides of the crib should be covered in a soft material, to prevent injury. The bed should not be used as a table for changing dressings, as this may be associated with painful, unpleasant procedures. The bed should instead become a safe restful place for the baby where later, as a small child, he or she can retreat to as a safe haven.

3.2. Preschooler

3.2.1. Motor skills development

When the child is of preschool age the parents and caregivers are in for a challenge. Early on he or she will be beginning to sit and trying to crawl. It is normal in this phase for minor or sometimes even more major accidents to occur. When an EB child has an accident it often involves the development of blisters and it is then followed by pain. Parents understandably want to protect their children against all accidents and pain, as they do not want to see them suffer. It is also important for the child to find out for himself what his physical abilities are and thus learn his or her own boundaries. A balance needs to be found between the parents' protection and the child's will, whereby they protect the child and yet allow him or her to explore new things. This is a major challenge for both sides of the family. The parents need to be advised that the relative vulnerability of their child's skin is often related to the type of EB they have and that all EB children should still be allowed some measure of freedom. Parents need to be informed about protective measures they need to enforce, such as bandaging techniques for the knees and elbows, so that the child can crawl without serious injury, and for the feet, when their child is first learning how to walk.

Early consultations with a physical and occupational therapist are recommended, to evaluate the kinetic motion problems that often lead to motor functional disabilities. Early actions can modify the restrictions and help with the physical development of the child.

In those children who have a type of EB that increasingly restricts the movements in the fingers, hands, and even the tongue and opening of the mouth, practice fun exercises should be pursued in a playful way, to help to delay or prevent the development of constrictions. Unfortunately, practice shows that it is not completely possible to stop this restrictive process in every severely affected child, yet from the exercises it is possible to maintain the movement they already have. Mouth

exercises, for example, can be done by making frowns and smiling in front of a mirror, blowing bubbles and best of all, singing, which most children enjoy doing.

3.2.2. Promotion of intellectual abilities

Making important decisions for the child`s future should help in the development of all of his or her senses and in the enhancement of physical and intellectual abilities. In later years the child`s talents and special abilities can at least partially offset the limitations caused by physical disabilities that may develop.

3.2.3 Information about the disease

It is helpful to teach the child in easy words about the vulnerability of skin. In kindergarten the child will be confronted with frequent questions and a lack of understanding. The more confidence he or she has in knowing about the disease, the easier it will be to cope in unaccustomed situations. At this stage it is important to have playmates of the same age, and they should be informed about this rare skin disease, so that parents and child can learn to deal with both the positive and negative reactions that they may receive. A basic requirement for life with a severe chronic disease like EB is positive self esteem. It is important for everyone involved in caring for EB children to give them the support they need to achieve this.

3.2.4. Dressing changes

During early childhood difficulties with bandage changes will undoubtedly develop, as it often hurts and takes a long time. Pain and discomfort can cause anxiety for the child and the parents. The feelings of helplessness and being at the mercy of someone can anger an EB child and drive him or her into a rage which may be very difficult to manage. Advice and support may be necessary during this particularly difficult period of life. It may not be physically possible for parents to always provide the most optimal wound care, following a sophisticated care plan, every day of the week, given the other issues and conflicts that may at times unexpectedly arise. Despite that, the parents need to be assured that they are doing the right thing. For the years ahead they need self assurance, since their child will need to have the bandages changed fairly often in the week. All the tricks to divert the child should be used. Proven successful techniques include telling stories or listening to them on CD or tape, or watching videos and DVDs. Moreover, the child needs to learn relaxation techniques (and parents too!) and there are many ways available to learn them.

3.3. School days

3.3.1. Beginning school

Numerous fears accompany the child (and the parents) in the first days of school, and they are not only in relation to the learning needs. Will their classmates accept them, will they find a friend, and is the school structurally safe? Will the child have a long way to walk between classrooms? Are there dangerous areas in the class where the child might get injured? Is there a quiet place for the child to rest? Will the child have a teacher who is empathetic, caring, and patient? Is there enough time in the teacher`s day, along with her/his normal teaching duties and commitments, to invest energy into the care of the special needs of an EB child? These questions and concerns can be answered only when there is a good relationship developed between the school and the parents. For

their part in this partnership, the parents need to provide information about EB to the school's teachers and staff, and about their child's extra needs while at school to both their classmates and the teachers. Because of the rareness of EB it is not unusual that an EB child is the first one with this disease to have entered the school.

Prior to that, the parents will also need to get sufficient information from school administration and other parents of EB children who reside in their hometown, so as to identify the best possible school for a child with this particular disease. And undoubtedly, more questions will still arise. For example, which school will be most appropriate for the child? Does the type of school wanted even exist within the community? Whenever possible, it is preferable that the child be integrated into a normal school. Unfortunately, for some children this is not possible. In that situation, a special needs school is an option.

Some children with EB can cope very well. Others need a little help, while some need to have a fulltime assistant with them, to cope with their daily and special medical needs. This is often related to which type of EB the child has. It will also be influenced by the child's own level of development, and lastly what type of personality the child has. It should also be ascertained whether the child will be able to eat at school. An EB child with mucous membrane involvement will need to have special meals provided. This may not be available in every school, as it involves the need for special equipment and individualized preparation of the meal. However, quite often even the children who have more severe forms of EB can still be integrated into a regular school without major problems arising.

Finding friends is not always easy, due to the obvious effect of the illness and the child's restrictions at recreational activities. Being labelled as a "special needs" child can also be an obstacle. How easy school life becomes for an EB child often relates to the personality of the child, and certainly whether other children try to avoid rather than accept them. The better other children are informed about the illness, the more success the EB child will have. As a result of great efforts on behalf of the child by both the parents and the teachers, most children with EB do manage to build good friendships.

Despite the fact that society is better informed, EB children cannot avoid receiving prying looks by others who are ignorant of the underlying disorder. In rare situations, EB wounds may be confused with child abuse, even by pediatricians who are unaware of the underlying diagnosis. Classmates and teachers can play a very important role in providing support and encouragement to the EB child.

Even after a successful school start, parents still need to pay attention to the problems that may later arise. Most of these problems are easy to take care of, others require a lot of effort to resolve. There are many ideas and answers to help to solve them. On a day where the student is having a lot of pain or is feeling too exhausted to attend school, a helpful teacher might be willing to organise a web camera in the classroom so that the child can also see what is being taught in class that day. A computer with voice recognition is helpful for the student who has mobility problems in his or her fingers and is unable to type fast enough to finish assignments or papers for school. Little things like special scissors or pencil adapters can make it much easier for the EB child to participate in the activities of the classroom.

Allowing the EB child to leave the classroom a few minutes earlier than the other children will provide an opportunity to avoid the rush of students during break, or when heading home, thus avoiding the chance of injury. The use of a lambskin padding on the child's seat will make it more

comfortable for sitting for prolonged periods of time. Whenever reasonable the EB child should be allowed to participate in gymclass activities, but with special attention paid to the extra needs and disease-mandated boundaries that the child may have. An EB child can become easily exhausted, for example, due to pain or anemia. Such a child should be able to decide what can and cannot be done. As a correlate, we have repeatedly seen that children who are allowed to make their own choices often make very good ones.

Small accidents are unavoidable in the school setting. It is important that the school has a person trained in first aid (and with sufficient knowledge about EB), and that first aid kits, stocked with EB-friendly dressings, be available for use. Most teachers and classmates learn quickly the “do’s and don’ts” related to having EB. As a consequence, the EB child experiences a normal school day with all of its highs and lows!

3.4. Adolescence

In this often times difficult phase of life, friends become very important. All adolescents want to fit in, and at the same time they want to develop their independence and experience things on their own. Thoughts about physical contact with the opposite sex start to form, especially during puberty, and are accompanied by body changes and mood swings. The family, parents, and child have much to cope with. Teenagers with EB go through this phase just like any other child, yet those with EB have the additional problems and burdens that accompany their disease. The problem of looking good becomes very important, physical limitations bother them more, and the constant appearance of new blisters, wounds, and scars come to the forefront. At this time, establishing contact with other teenagers who have the same illness can help, since it gives them a chance to see that they are not the only ones in the world with EB. But sometimes at this phase of development they may prefer to avoid contact with other affected teenagers, since being different has always been a part of their lives and they want to show that they have a right to be different and unique, and are yet still seen as a normal person. Parents need to help them gain their independence and autonomy and to become responsible for themselves and their actions. This is not only in reference to wound care, which has been taken care of by their parents up until then. This becomes increasingly important for their future development of friendships and for the pursuit of a satisfying way of life. Even though it may be difficult for parents of an EB child to distance themselves as the child becomes older and more self-sufficient, responsible teenagers need to be able to have progressive control over their lives.

The question of a future profession is also an issue at this time. The teenager should be advised, according to his or her impairment, as to which jobs might be most suitable, realistic, or attainable. It is important to get the assistance and expertise of job counsellors who help the teenager to identify all of the available possibilities, both locally and at more distant locations. Choices of the types of employment may be limited, due to the teenager’s illness. Abilities, talents and special interests of the teenager also need to be taken into account and can play a large role in what is finally chosen.

3.5. Adulthood

The most important issue for someone with EB at this stage is to be self-reliant and independent. Most patients normally wish to have his or her own apartment, a driver’s license, a job, a partner and a family. Wishes vary and there are different outcomes. There are many obstacles to overcome to fully achieve all of their dreams.

3.5.1. Choice of profession

When looking for a profession an adult with EB should not focus on the deficiencies that are present as a result of the disease, but instead look at all the possibilities that are available. The choice of profession will often depend on the type and severity of EB. For most forms of EB the potential range of choices may be unlimited, yet not all professions may be realistically possible. This, of course, applies for everyone, not just those affected by a rare disease such as EB. Factors influencing the choice of profession also include the educational level achieved, the personality of the EB person, and whether the individual has special skills or talents.

Once vocational training has been completed, a decision must be made as to whether full time or part time work will be pursued. If it becomes impossible to work part time or to work at all, due to the extent and severity of disabilities present, a disability pension should be taken into consideration. It is worth remembering that for most adults, their feeling of self-esteem is influenced by the choice and success of their careers. In the setting of a severely disabling disease like EB, however, even a relatively insignificant job might be satisfying. When employment is not realistic, a hobby can at least provide some balance in their life.

3.5.2. Operating motor vehicles

Having a disease such as EB does not exclude one's obtaining a driver's license. On purchasing a car several important adaptations may need to be made, particularly for those with severe generalized forms of EB. A large mirror should be installed so that the driver does not have to make excessive spontaneous movements while driving. A knob on the steering wheel will make it easier to turn the wheel, or to turn it using one hand. It would also be beneficial to have power windows, an automatic transmission, soft seats, automatic door locks and a well padded safety belt.

3.5.3. Relationships and sexuality

Very difficult and sensitive topics are personal relationships and sexuality. If in today's world, finding a partner who is "rich, young and beautiful" is difficult, then it must be even harder for an EB adult to fulfill such a dream.

Society norms are luckily not the major issue of one's whole life. It would be ideal if it were possible for people to look and see that there is a normal person in front of them, rather than one affected by EB, and that an individual with EB has the same strengths and weaknesses as others do. The way would then be freed to be candid with each other if and when the conditions fit, and then a relationship could be built based on equal footing. There are many encouraging examples of this that we have seen among our patients, although it is all too often that the wish for a serious relationship is not met. The need is always there and the longing for sexual contact is not so easy to put aside. Everyday answers are not always available and a sensitive discussion of these issues is necessary. It is important for both the adult with EB and the caretaker to take the person serious about his or her most personal needs. With creativity and an open mind, other ways might be found that can satisfy the need for sex and tenderness.

3.5.4. General physical examinations

As EB patients age, the more severely affected ones may develop increasing physical limitations, as the result of a worsening of extracutaneous complications. The most notable complication is squamous cell carcinoma, which can lead to death. For the overall well-being of the EB patient, a visit to a dermatologist at least twice a year is recommended, once adolescence is reached.

It also should not be forgotten that routine, EB independent medical check ups are still necessary, as it is possible for a person with EB to develop other unrelated health problems, which can be more difficult to treat in the setting of such a disabling disease.

4. Conclusion

An important goal for everyone with EB is to develop strong character and self confidence. This can be achievable with lots of effort and the love and support provided by family and friends. All patients with EB should have the opportunity to experience the many things that life has to offer and, even with their many disease-associated limitations, to be able to lead to a happy and fruitful life.

Regretably, reality often deviates from idealizations. Expectations may have to be modified on an individual basis, and with the efforts and cooperation of all participants. Every patient has his or her own EB. This means that every individual with EB has a unique history and personality. Therefore, experiences with the illness will differ from one patient to another. Age-dependent individualized care of this heterogeneous group of patients is necessary. Our experience with many EB patients over several decades shows that this approach will pay off.

5. Information

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