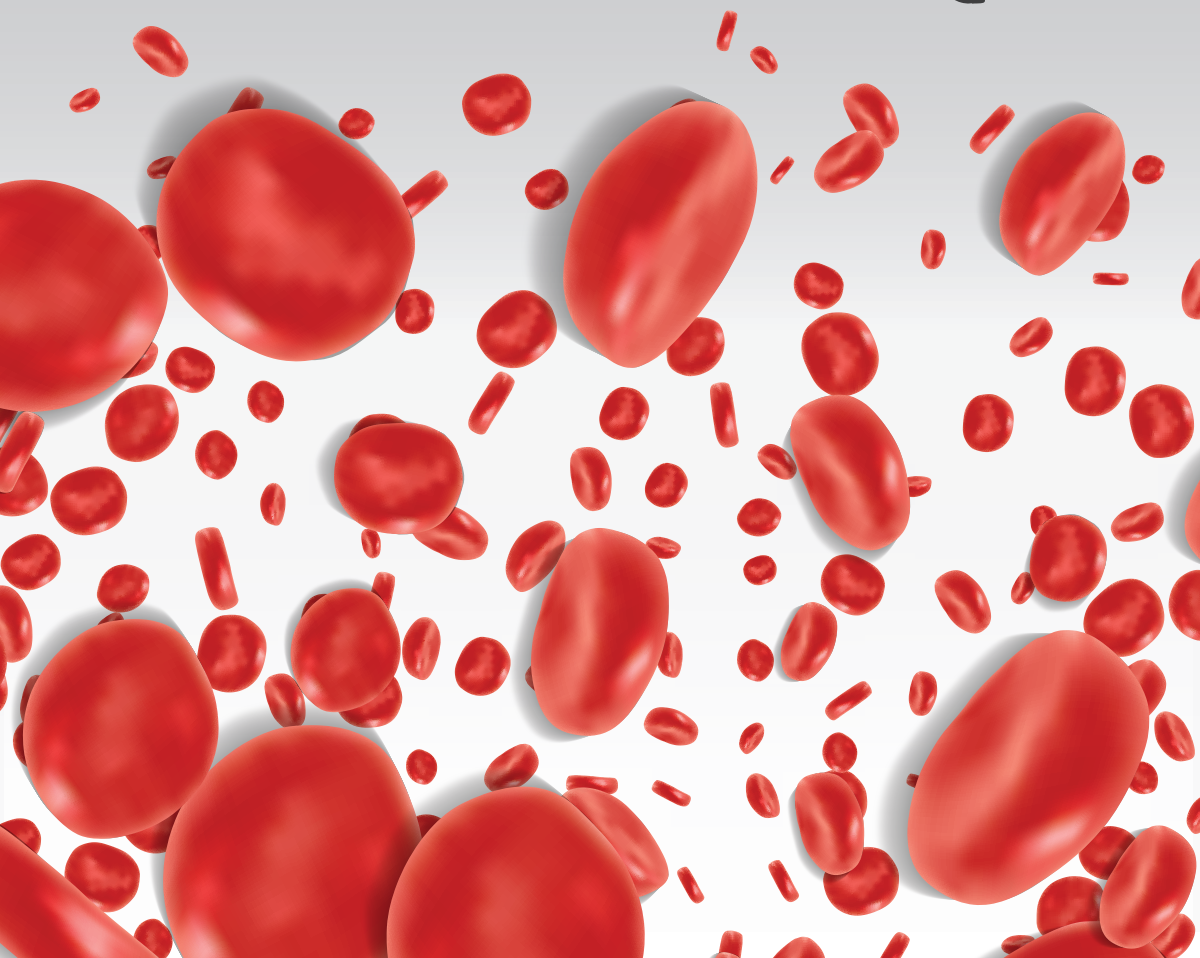


BERGIT KORSCHAN-KUHLE

out of **SHAPE**





FOREWORD

Bergit Korschan-Kuhle is a courageous woman. She has been living with the diagnosis of MDS since 2006. In “Out of Shape” she takes the reader on a journey from her first symptoms over a decade ago to the present day.

This book will help patients with MDS, because what the Author reveals about herself is honest and authentic, and the encouraging aspect of this book is therefore not made up and constructed, but convincing and leads to hope despite the condition.

Bergit speaks very openly about her fears, the chaos of emotions that the illness caused in her and the uncertainty about what is to come. She also does not hide the loss of friends who could not cope with her illness, or the friends she met through MDS but then lost through MDS. She lets MDS patients and their relatives share her knowledge about the disease and her perception of MDS. In my opinion, this is a very valuable addition to the medical perspective, which can help patients to better cope with the disease.

Over the years, Bergit’s fight against MDS has turned into a life with MDS. She lets her readers participate in this development. She reports on her involvement in patient advocacy, which in turn became her own support. She is even able to recognize happiness in certain aspects of her illness and to accept it for herself. She justifiably joins the demand “Patients In, Not Out”, in which she calls for involvement in all areas and with all stakeholders in our health system. Her story provides insights into a closed digital MDS Facebook group in Germany, which is also of great value to relatives. This encourages patients to work on their development, to accept information, but also to actively demand it.

With her book, Bergit offers her fellow patients and their relatives the wonderful opportunity to benefit from her valuable experience, e.g. in dealing with typical MDS problems such as anaemia, fatigue, transfusions, bone marrow punctures or port systems and iron overload. Bergit makes the development she has gone through—the change of perspective as she calls it—comprehensible. She thus creates a high degree of transparency, and it is transparency that takes away fear and helps in the acceptance of the unchangeable but without surrender or resignation.

In “Out of Shape”, the Author advises a realistic approach to hopes and expectations when participating in therapeutic studies. She calls for a stronger involvement of Patients in the design of these studies and a much higher weighting of Quality of Life as a therapeutic goal.

Bergit also deals with the topic of stem cell transplantation in an honest way. From her own experience, she is able to convey what it feels like to have waited for years for the right time for a transplantation, without letting it get you down.

Bergit directs her gaze inwardly with realism, the right dose of emotionality and sometimes with restrained humour, but also outwardly towards relatives, friends, the “Carers” and their worries, hardships and limitations on the one hand, and dependence on them on the other.

This book demands strength in reading, but also gives a lot; not least because the Author opens up and reveals a lot of personal experience.

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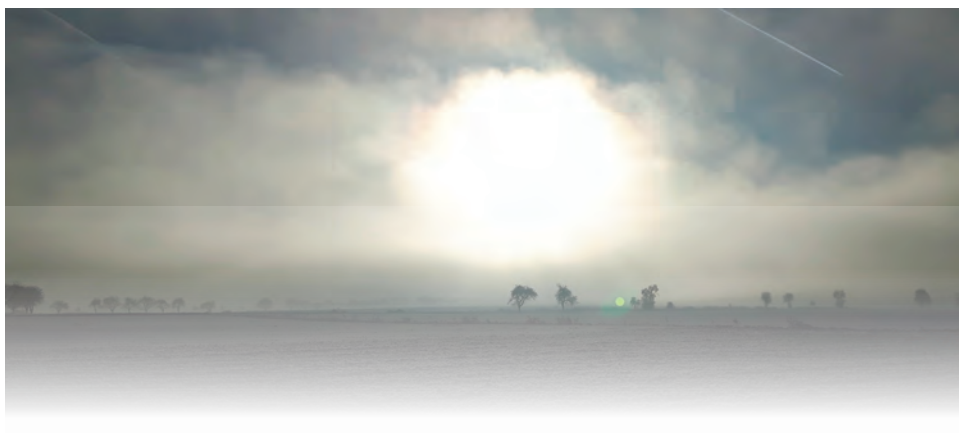
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1. BECAUSE IT CANNOT BE, WHAT CANNOT BE...

The family doctor says “There is nothing.”

The ultrasound shows nothing.

The palpations do not show anything.

Yet, the slight pain under the right ribcage is there for hours, almost every day.

Maybe a slight inflammation of the gastric mucosa?

“Do you have a lot of stress?”

At first, I am calm, but then there is an appointment with a foreign internist who is very thorough and checks everything.

Blood is drawn. I swallow a tube for a gastroscopy. The thyroid gland is examined...

Afterwards, I can relax.

“There is nothing” says the internist, just a slight redness of the stomach lining and two inconspicuous lumps in the thyroid gland.

Would the afternoon of February 16th be convenient to discuss the blood values?

That would be on my 49th birthday.

“I’m sorry I have to bring this up on your birthday, but I suspect a systemic blood disease”, the internist says without a pause.

He seems to be a perfectionist.

There is panic on my part: “Is that bad?”

“Well...”

I say the name of the only blood disorder I know: leukaemia.

“No, probably not. There are several possibilities.”

I start to feel hot.

“How can you be so sure? Maybe there’s a simpler explanation?”

He explains that he has already tested B12 and folic acid. These two values are fine, so they couldn’t explain my poor haemoglobin level. I did not understand the reason, so I suggested: “Then I should take iron supplements.”

I knew from my pregnancies that a poor haemoglobin value meant an iron deficiency, which could be corrected with oral iron therapy.

The doctor forces a smile and shakes his head. I don't think it's funny, and I don't like him either.

"I'll call my colleague, a haematologist, and he'll check everything else with you."

He looks at his watch. "Maybe he's still in the clinic."

He is. The internist makes an appointment for the subsequent telephone conversation between colleagues. My internist is very thorough, and he seems satisfied, but he is now finished with me.

I also have nothing more to say to him and never want to see him again.

Arriving home, I sneak across the terrace like a ghost in the dark. My husband later says that he saw me creeping across the terrace from outside through a window in the conservatory and suspected something was up.

The internist had told the hematologist on the phone, "The patient has no B symptoms."

On Google I find B symptoms:

Night sweats, lack of appetite, weight loss, pallor, exhaustion.

No, I don't have any of that.

And I no longer have pain in my right upper abdomen.

Back to the family doctor, who takes a look at the blood values faxed by the internist.

I pass them to him with reproachful words: "Can you please explain these blood values to me?"

The internist says I have a malignant blood disease, as if that were my family doctor's fault. In the face of my inference, my doctor seems insecure for a tiny moment, then he says what I want to hear:

"Oh, there's nothing, at least nothing bad."

My husband looks at me reassuringly.

The three of us show solidarity against the bad blood values. My family doctor ends with a convincing argument against young specialists who, unsolicited, pour out all their specialist knowledge about patients and unnecessarily panic them.

However, I could still pay a visit to the haematologist, a friend of his, by the way.

The thing is, if you go to a hematologist or oncologist, you don't just have a cold.

Do the receptionists at these places have the job of being particularly happy here because of the sad diagnoses they deal with?

The blue tumor booklet, which some patients hold in their hands, reminds me of the mother's passport from pregnancy, in which the haemoglobin value was entered every time they visited the doctor, in addition to the weight gain.

The haematologist appears for my consultation, radiating trustworthy charisma.

In my eyes and in my voice there is so much fear—how can he know about so many patients?

He totally glosses it over.

He explains to me that he will look at my blood cells under the microscope. He is quite satisfied with my blood values. All the values in question—and these are all those related to the red blood cells—are significantly better than those from the internist. My haemoglobin value is 11 g/dl where 12—14 g/dl would be normal.

If he should notice deviations from the norm under the microscope—he clears his throat at this point—then he would perform a puncture here, tomorrow, and then we would have clarity.

There is no disease name.

In the mean-time I research what a bone marrow puncture (BMP) is.

In many haematological diseases, a blood count alone is not sufficient for a diagnosis, so a puncture of the bone marrow is necessary. This is used to obtain a tissue sample (biopsy). The bone marrow is aspirated with a special hollow needle. Either some bone marrow is aspirated with a syringe or a small bone marrow cylinder is punched out. The tissue sample is examined microscopically in the laboratory.

The next day, the receptionists are just as loud and happy as the day before.

One of them is so cheerful that she calls to me from a distance: “So you are back for a puncture today?”

I hardly get a sound out.

“But I didn’t expect a puncture today.”

“Oh, it’s not that bad.”

I go to the patient toilet, stand in front of the mirror and watch myself cry.

Meanwhile, my husband is still looking for a parking space. I pull myself together and come out again when my husband, who is just arriving, is told that I will have a puncture done immediately. Since the waiting room is overcrowded, we can hardly talk about it during the waiting time, and both sit quite petrified for a while. After half an hour we are called into the consulting room.

For a few minutes we are alone in the consulting room, standing far apart and I say without any prompt: “Maybe we have to sell the house when I can’t work anymore.”

The doctor enters with a quick step and a lively “Good morning.”

Irritation on our part. He does not even give the impression that he has something serious to tell us.

He doesn’t either. Everything is fine.

“The blood cells look quite normal under the microscope. There is slight anemia, probably the result of a viral disease, but that can happen and that shows up in the blood count, even if you don’t notice it.”

“But the puncture, shouldn’t I be punctured right now?”

“I didn’t order that.”

The receptionist gets a rebuke; it is a mix-up.

It turns out that another woman from the same year of birth was sitting in the waiting room, now lulling herself into a false sense of complacency, believing she doesn’t need to be punctured—and yet she does and I don’t have to.

Not me!

The hematologist adds: “Why don’t you come back and do the blood test from time to time? With our heads raised, we stroll past the counter. I don’t have any business here anymore. I am healthy.

At my birthday party I tell the story in a snotty tone and with the arrogance of a healthy person: “That’s how it can go. I mean, theoretically you always know that, but now I have understood that anyone can get it any time. Lucky again.”

Very quickly I dive again into my normal everyday life of work, family, small and large pleasures and annoyances.

In the following weeks and months, I often remember those terrible weeks full of insecurity and fear, but I remember something that is over and has nothing to do with me anymore.



2. THINKING DESTROYS THE ILLUSION

The year 2006 had just begun. On my birthday in February, I had suffered the shock of possibly being terminally ill. I had stood close to the abyss; I had looked inside and was allowed to walk again. I was lucky!

I decided to definitely go back to the hematologist and take the blood test he had asked me to do, in summer or by the latest in autumn.

During the Easter holidays, my husband and I fly to Tenerife for two weeks. For the summer holidays I book two weeks of “hiking on Gomera” for myself without my husband; hiking is not for him.

On Tenerife we also rent a car, because we want to see more of the island than the monstrous hotel complexes. Our most spectacular route leads us directly to Pico del Teide.

At 3718 metres, Pico del Teide is the highest mountain in Spain. We start at 0 metres altitude. We can go up to the valley station by way of cablecar, with its restaurant which lies at 2356 metres.

From there, another cable car goes up another 1,200 metres in seven minutes and the last 150 metres are on foot.

We park the car at the parking lot and head for the cable car station, which is maybe 200m away from the parking lot.

I start at my usual speed but after a few meters I come to a stop. I can hardly get any air and my legs hurt. I am completely surprised, as the other visitors walk up the path quite easily, as well as my husband, who is heavily built and also a heavy smoker. He gives me a few vegetable cardiovascular drops on the back of my hands. That helps. He always has them with him, just in case...

We take a break, then walk very slowly and a little later we reach the centre. On the building there are several signs, which indicate in several languages that 2356m is already quite high and the air is thin; one should not exert oneself otherwise, under certain circumstances, this could result in breathlessness. On the way back downhill, I walk with a smaller step without the slightest pain in my legs. I have already forgotten the incident.

In August 2006 I am on Tenerife again, this time alone.

We start in Los Christianos to take the ferry from there to San Sebastian de la Gomera. There I meet the hiking guide and the remaining eight fellow trekkers of the small travel group. We will hike around Gomera for eight days, with hikes between four and six hours—and enjoy the second week in the Valle Gran Rey on a bathing holiday, as it says in the booking description.

Our hiking guide is a small wiry woman in her mid-30s, with a muscular build and well-trained calves. She leaves us in no doubt that she will “do” this, and that she has certain expectations of our fitness. On the stairs, as we head for our rooms after our first dinner together, we try to check with each other if we are in good enough physical condition for Gomera-Trekking. Most of the group say that they’re actually quite untrained, that they do not do much sport, and that they’d hiked before but along level ground. But if you can’t handle it, you can take a taxi ahead on the way out, or after the group on the way back. There is nothing dishonorable about it. At least that is what it says in the travel documents.

Gomera is a hiking paradise, but hiking on La Gomera is almost exclusively mountain hiking. It almost always goes up and down: 500 meters up, 500 meters down, 300 meters up... When I booked it was not so clear to me, or to the others either.

We start the first hiking day with a bus ride to our starting point at 900 m altitude.

Our hiking guide makes sure that we drink enough; we also have to eat a snack from the packed lunch at regular intervals and she sets the breaks. I don’t find the heat so stressful, but I would like to have more and longer breaks. I also notice that I am always slightly behind with my speed. But today is the first day and I have to get used to the walking pace. The descent to the south to the sea to Playa de Santiago takes hours. Although five hours walking time were indicated, we needed more than seven. Most of us plunge into the sea in the evening to cool down. It’s too exhausting for me, but I don’t waste any time thinking about it.

At dinner we discuss the next day, and the hiking guide suggests that those who want to hike a little less strenuously, can be taken by taxi to the first station in a small village, while the others make a steep ascent of about one hour. The subsequent longer part of the hike would be with the group together.

Another woman and I go with the taxi.

After meeting with the rest of the group, we climb a slope without a real path with lots of rubble, partly on all fours, where we have to hold on to plants. In order to overcome the gradient better, we walk in serpentine fashion. On our backs we have about four to five kg of day luggage.

After a few minutes, I’m at the back, everyone has passed me, and very soon nobody is in sight and I’m stuck on the mountain. I manage with my backpack to climb about five to six steps on this ascent, then I have to take a breather and then stumble further. I turn around several times, see the island of Tenerife and Teide in the distance, wide orange, lemon and almond groves below me. The view is indescribably beautiful. I can still hear the voices of my fellow hikers when they take a break on a rock ledge, whether planned or because of me, I don’t know. Two of the group smoke another cigarette. When I finally arrive there, they get up to leave, because now I am there. Now I was the only one who had no break at all. I feel exhausted.

When we reach the top, there is only a short stretch of road left to walk, not even steep, before we have lunch under shady trees in the Garajonay National Park. The sun is shining. Shortly before the last two or three steps leading up a small slope, I suddenly stop. I can hardly move anymore. There are worried voices, I get support, I have to sit down, drink something and eat. I can't talk, I can't put down my backpack alone. My battery is completely empty. Even during the lunch break it doesn't recharge properly.

When I descend, I am very slow, and when I see the new ascent, which we are supposed to go up again afterwards, I get scared and ask the hiking guide to order a taxi for me, although we only have about an hour left to walk. She is gruff; from her point of view I am now identified as a disruptive factor. I should not have registered for such a trip due to this lack of physical condition. For her, nothing is more damaging to her reputation than when tourists overestimate themselves and possibly have to be picked up by helicopter because of a circulatory collapse on the mountain. People would not want to hike with her anymore, and since she has only recently emigrated to Gomera, she is dependent on the earnings.

For the rest of the vacation, I used taxis more frequently, but despite this, I was enthusiastic about my vacation, particularly the time in Gomera.

I reported my fitness problems at home, but I didn't dramatize it; I accepted it afterwards without thinking about it any further.

The rest of 2006 was "trouble-free." I felt capable at work and in my free time. I barely thought of the occasional turbulence from the beginning of the year, and what the holidays on Tenerife and Gomera had put me through.



3. SHOWDOWN

On the penultimate day of 2006, I went to my family doctor and had blood taken for a checkup as scheduled. Although the blood values would have been available one day later, i.e. on New Year's Eve, I did not call the practice until the beginning of January 2007, completely relaxed.

"Yes," said the friendly receptionist, "I would like to tell you more, but the doctor sent the blood count back to the laboratory for examination, because one value was a little low."

"Which one?" I enquired, but I knew the answer right away.

"The haemoglobin value, I think. The doctor wrote it down—please have vitamin B12 and folic acid checked—the results will be there tomorrow."

There it was again, the abyss, and this time I fell into it.

The internist had already found out almost a year ago: "A lack of B12 and folic acid cannot be the reason why your hemoglobin level is so bad." I didn't ask the receptionist any more questions. After that I cried for a long time.

How immune can the mind sometimes be to clear physical signs? But thinking would have destroyed my illusion of being healthy.

It wasn't just the one value that was "a little low." All values related to erythrocytes, the red blood cells, were outside the norm – all of them.

Somehow, I hoped that the haematologist's receptionist—to whom I had given a devastating look when I left last year after she confused me with another patient—would not recognize me. Maybe she was not working there at all. But when I walked in the door, I saw her sitting behind the counter.

I don't think she remembered me and she was very friendly. She became my "favorite doctor's assistant" at the haematologist.

My red blood cells no longer looked so normal under the microscope, "I found a few too big," the haematologist said solemnly, adding "I would like to perform a puncture."

I was afraid of the puncture.

I got Midazolam and slept through the actual action, lasting about 15 minutes. Midazolam prevents you from remembering anything afterwards, so I didn't know if we had taken the stairs or the elevator down and I didn't remember the car ride either. I was just relieved to have it behind me. The next day I went back to work.

I did what the haematologist had strongly advised me not to do: I surfed the Internet. I combined blood values, the cases of normal puncture, symptoms, forms of anemia, and I came to an assumption. I found a name. I found out what it might be. A name for a malignant blood disease:

Myelodysplastic Syndromes or "MDS", also known by the outdated term "preleukaemia." I repeatedly found the phrase "a form of blood cancer."

No layperson who I spoke to about this condition knew about it.

The statistics show that at my age, only 4 out of 100,000 people suffer from MDS every year. The risk of developing MDS is unbelievably low. You'd be more likely to win the lottery. Maybe I had something much more harmless?

Once again I had the vague hope of getting away with it, as the haematologist had said: "maybe the puncture doesn't result in anything."

It took more than 14 days until the results of the puncture came. The tissue samples had been sent to special laboratories in Göttingen and Kiel.

A few days before my 50th birthday my husband and I sat in the haematologist's office.

After an initial exchange, he turned to his monitor and read it. He seemed businesslike and then looked at me: "My colleagues found signs of myelodysplastic syndromes."

At best I had only understood half of what I had read about it on the internet before, and it had frightened me.

I was willing to cooperate, to do everything for a successful therapy. Perhaps I had missed something important.

The haematologist really tried and tried to say everything positive that was possible: "You have the best MDS you can have. Only 6% chance of transformation to leukemia. If I could choose an MDS, I would choose yours. "You don't have any blasts"—he looked again very closely—"obviously only the red blood count is affected, leukocytes and thrombocytes are OK. However, a chromosomal defect has developed on the 8th chromosome, which is quite typical for MDS. This is called trisomy 8, an MDS type with intermediate risk. Prognostically it is neither favorable nor unfavorable; it is in the middle."

That was it. That was the diagnosis: It was lying on the table, ugly, naked and wrinkled like a little bird embryo. It would grow and become a bird of prey with sharp claws and a pointed beak. In my dreams over the next weeks, this bird hacked me again and again in the face and looked at me afterwards with its completely expressionless eyes, unimpressed and lurking.

The haematology specialist in the clinic commented on my prognosis as follows:

"You see, normally you are much older before you get MDS: 60, 70 or more. If I now classify you in the group of older MDS patients who, due to their age, have all possible pre-existing conditions, then your statistical prognosis is of course poor, even if your personal prognosis is much more favourable. Prognoses are very relative and say little about the individual case."

The only thing you understand as a patient in this situation is “your prognosis is bad”; you get stuck on it and all further explanations from the doctor end up evaporating from the mind.

There is the matter of “median life expectancy”; that was the number that always frightened me the most in the beginning. The median life expectancy of my MDS type RARS was 60 months. Trisomy 8 made me score 1 and only gave me 39 months. This is how it works: Median life expectancy means that statistically, 50% of patients with score 1 die after 39 months, the other 50% live longer. But for how long?

This is how MDS prognosis systems work.

I am entering the 15th year of my illness. If I enter my current parameters into prognosis scores, I would still be around three years old. Statistics are made for large groups however, and in order to represent an individual case, they are not suitable. Reaching this understanding cost me many sleepless nights, and my distrust of this insight has always remained.

“Maybe your haemoglobin value stays between 10 and 11 g/dl for a while, then we don’t have to do anything. You continue to live as before. If not, we can try different things. There are some drugs in the pipeline and blood transfusions. But we have not got there yet. You are doing well. And please don’t think of a transplant; you shouldn’t consider it at all. That is for when it’s a matter of life and death.”

My haematologist behind his desk was determined to downplay the situation. He wanted to help me and show me that my current stage of illness was not a big cause for concern. Given the background I already knew, and the expectations for the future, this was a rather hopeless undertaking. I was definitely worried. I was the personification of concern and despair.

The doctor tried to be gentle with me, and I appreciated his efforts. Although the waiting room was humming, he took over an hour for me and my husband. It’s just that after a while, I switched off and couldn’t absorb or process the information at all, because I had to keep staring at this grimace-like bird-thing that had been put on the table as a diagnosis.

On the way back home, my husband cried. I sat rigidly and stiffly beside him.

It was now up to me that my blood values should be closely monitored, at least every four weeks. The family doctor would fax the blood count to the hematological practice. At longer intervals I would have a conversation with the hematologist.

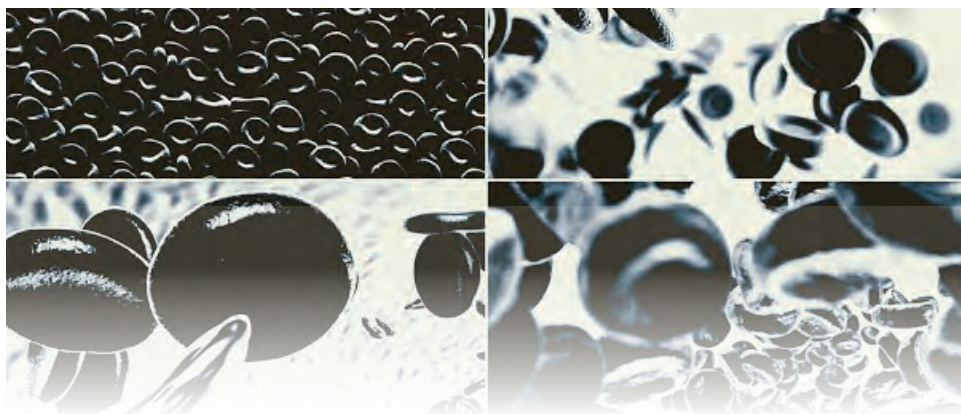
A new era had begun.

From now on, light-heartedness was seldom successful, and blood values took on an enormous importance. I made my well-being and my survival dependent on numbers—a typical beginner’s mistake.

Only one month later I celebrated my 50th birthday. This time the guests were divided in two: A few who knew, the others who didn’t. Unlike the year before, illness was not an issue. It was a beautiful evening and I was glad that I had been persuaded to celebrate and function normally.

What would have happened if I had gotten the blood test earlier, much earlier, perhaps three months after my first visit to the hematologist, and not almost a whole year later?

Maybe he would have noticed that my anemia was not harmless, but had a malignant origin. I would have lost a whole year in which I actually felt healthy, apart from the limitations of the Canary Islands. In the light of day, I was given this carefree year, because knowledge of the disease would have had no therapeutic consequences whatsoever. Not with every disease does early diagnosis make everything better, and especially not with those diseases for which there are no targeted drugs for the early phase.



4. DEFORMED

Myelodysplastic Syndromes (MDS) is a blood cancer—a disease characterized by the deformation or change in the appearance, size and the number of blood cells. All three types of blood cells can be affected. For example, the red blood cells (erythrocytes) are no longer smooth, round and concave discs of about the same size; instead, they appear as bizarre forms of crescents or various cloud shapes, which are clearly malformed when viewed under a powerful microscope. Medical specialists refer to these blood cells as “dysplastic.” The deformed cells often show additional abnormalities. Individual chromosomes can be defective. Pieces of the genetic material may be missing, or they might be malformed or rearranged. Additional material might also be present. Trisomy 8, for example, is one of the more frequent deformities. With Trisomy 8, the 8th chromosome forms a third “arm” instead of the normal two “arms.” During cell division, this error is copied because it is no longer eliminated by the normal cell destruction process. Finally, individual genes on the chromosomes may be mutated.

Depending on the stage of the disease, the quantity and quality of these deformations and disorders vary in severity and threat, especially if the white blood cells (leukocytes) and platelets (thrombocytes) show disorders in maturity. As far as the red blood cells are concerned, the miracle of continuous blood formation and oxygen transport to tissues and organs becomes permanently impaired.

It is not easy to diagnose MDS because other diseases of the blood have similar conspicuous blood counts. The diagnostic phase can take longer and is therefore very stressful for the person affected. Once the diagnosis of “myelodysplastic syndromes” has been made, however, it is clear that the person is seriously ill.

The Person is now a Patient, probably for a long time. The diagnosis is a shock; a deep wound for the patient and their close ones.

The course of the disease does not only reveal itself in internal changes to the blood cells; it changes the Patient’s entire life and identity from the time of diagnosis, the extent of this depending on the duration and severity of the disease.

A general attitude to life, self-perception, the ability to cope with crises, the willingness to learn, the ability to make decisions, relationships to other people, spiritual topics and finally questions of everyday life are inevitably put to the test. From now on, the topic of quality of life will play an important and, above all, highly conscious role. In addition to mental stress, there are physical problems in the areas of nutrition, excretion, exercise, sleep and pain management.

The aim is to integrate the disease into life, for which old patterns of behaviour and thinking may have to be abandoned. This is difficult, but there is no other choice but to accept, for example, a partial loss of physical mobility and restrictions in general well-being either in the short or long term, depending on the therapy options needed. Joy of life and satisfaction in everyday life must be given a different, sometimes completely new baseline. One sets out to endure the changes and integrate them into one's everyday life. This is a long process with mental successes and setbacks.

After many years with a chronic illness one forgets what it feels like to be healthy. "Do you remember what it was like when you woke up in the morning and it didn't hurt?" When friends tell us all too penetratingly about wonderful, extensive hiking tours, how many kilometres they made and how they felt at one with themselves and nature despite the great effort, then suddenly you remember very intensely about past healthy times. This is very painful.



5. NOTHING REMAINS THE SAME

Every time, it was a challenge to see how fast and with how few breaks I managed the climb. Every time, it was about the mountain; the rest was easy, but the mountain sometimes locked itself up. I could never be sure how good I would be. When I got to the top, I enjoyed the view every time, but in reality, I had to catch my breath.

The way through the rapeseed fields gave a marvellous view of villages, hills and open fields, then there was a second but shorter ascent into the forest, then finally downhill again and on country roads back to the starting point behind the village. It was a distance of approximately eight kilometres.

Our running group consisted of me and three to four women of my age, with an occasional addition who joined and then disappeared. We bought walking poles, better shoes, heart rate monitors—and developed into committed and ambitious runners who met two, sometimes even three times a week for running. The farmers who drove their tractors along the fields shook their heads, and we heard one or two stupid remarks from walkers. We didn't care.

In time, we managed the ascent without a break. Even at the top, in front of the entrance to the forest, we no longer stopped and we completed the route in considerably less than an hour. We felt in high spirits during the run and even better afterwards.

For me, running had welcome health effects. My tension headaches and back pain disappeared and I felt fit all week. It was just the right balance for a hard job and it was so easy to do. We also ran in the cold, rain and wind. We promised ourselves that we would keep running forever. That was in 2003. We thought we would still run together in 20 years. Running is a fountain of youth.

The physical effect was one side, but the other was the “mental purification.” When running, we talked easily and freely. There was hardly any topic, no matter how private, which was not examined in detail. We knew exactly where everyone was in their lives, we knew their biographies, the medical histories of the whole family, the conflicts with their neighbours, marriage problems and secret wishes from each other. We ran side by side and just talked. The words came of their own accord and unburdened us.

When I got the first slap in the face in February 2006 with the suspicion of a “malignant blood disease”, I had of course told my friends very little. While we were running, my problem was looked at from all sides and talked down until there was only one harmless thing left. That took away the pressure. After the all-clear, the topic was soon shelved. Nothing more stood in the way of our running motivation.

When I came back from my hiking holiday on Gomera, it was suggested that I was now in top form. I mentioned my fitness problems, but my friends couldn’t really imagine it.

After my diagnosis in February 2007, I went on, but the mountain gradually developed into my adversary. I would have liked to have taken a short break half way or at least at the top of the vantage point. I couldn’t be sure to make it every time without any problems. My calves usually hurt badly and I was completely out of breath when I reached the top. I began to fear the mountain, but I was not ready to give it up. The physical exertion I felt prevented relaxed conversations. I needed all my strength to master the mountain. And like on Gomera I was falling behind. That was new. So far, we had always run in threes or fours or pairs next to each other. Now I caught up only if the way went straight ahead or downhill.

In addition, we had a newcomer, who pressed hard on the tempo and was able to assert herself.

The change led to the others facing the faster pace ambitiously. Suddenly the total running time was measured, and we tried to undercut it each time. On occasion, I cried on my way home after these runs.

I had tried to make it clear that I couldn’t run faster because of my illness, but the story was lost: I could not expect the others to follow the pace set by my illness. They wanted a training result, and I simply couldn’t keep up. Of course, they didn’t say it like that. They didn’t really talk about it at all; what should they have said? I seriously considered whether I should ask for a head start on the mountain. As idiotic as it sounds, I was ashamed of my failure. I should have said: “I’ll stop.” But I wasn’t ready yet.

“Decision Time” came by itself on a rather warm day in April 2007. After about 350m uphill—about halfway up the mountain—I stopped. I couldn’t go any further. I was dizzy, ridiculously hot and I had a tachycardia. It was The End. Gomera 2.0. The other runners disappeared in the forest. Our era was finally over. I slowly made my way home.

Nobody called that afternoon to ask how I was doing.

It took me a very long time to cope with this forced end.

The ascent on the mountain was symbolic for me. It became clear to me that even my strong will could not bring my body to produce more power. I had to adjust to the illness and its symptoms. The mountain had shown me this, irrefutably and decisively.

At irregular intervals, I continued to run up the mountain at my own pace. Sometimes I managed it without a break, which was extremely rare. I tried to retain the enjoyment of the run, but I did not succeed. The running group split up; it did not recover from my retirement. Nothing remains the same.

Cancer is ruthless and frequent. Of the four women in our group who had run together for four years, two others fell ill with cancer apart from me in the following 11 years. The only thing was...the other two recovered.



6. THE FACTORY IS BROKEN

It is a great challenge to explain my complicated and complex blood cancer to others. Leukaemia is generally the only well-known term for the many different blood cancers. This term immediately triggers horror, and has such a negative impact on the listener that it is a matter of luck if the other person continues to hear anything further. I'm not saying that I have leukaemia, but I usually restrict myself to using the term "leukaemia-like." What this means exactly is rarely asked by anyone. On the other hand, irritated looks are frequent.

In 2007 I was in Heidelberg on my first patients' day for "Myelodysplastic Syndromes." The speaker described bone marrow as a factory for blood cells. He inspired me with the following idea:

Imagine your bone marrow in the large pelvic bones is a factory, a factory that has been producing healthy and functioning blood cells for decades; products of high quality in terms of size, shape and function.

Unexpectedly, an unpredictable event occurs in the production process, which can be triggered by environmental influences for example, or is simply due to the fact that the machines (stem cells) are too old and worn out. In any case, in addition to the still-high quality goods, defective products also emerge from the production line from time to time. This means that some products are sub-standard, defective and can no longer be used for further processing.

At first, the factory's own internal control system works quite well; it succeeds in correcting the errors and partially eradicating the defective products. In addition, there is still a sufficient quantity of quality goods to compensate for this, so that nobody really notices the growing malfunctioning processes, and the increasing number of defective products.

Over time, however, the factory's repair system becomes overwhelmed and the factory owner (the patient) notices that something is wrong.

After a time-consuming error analysis, immediate action is required, otherwise the existence of the factory is at stake. The factory director (the haematologist) sends his best engineers (the medical teams, laboratory staff and technical staff) to solve the problems.

Some of them take a more general approach, while others specialise in particular parts of the machines and work on them selectively.

At first, this strategy seems to work, although one does not know exactly which of the specialists was the decisive factor in the recovery of faultless production; but that is not so important; all the parties involved are celebrated.

However, the factory director (the haematologist) remains very cautious, and does not easily accept that the successful outcome will be permanent. He explains that certain errors in these highly complex machines cannot be corrected once they are there, and that it is very likely that there will be more production errors very soon. And that's unfortunately how it is! After a short period of success, production starts to falter again.

After a detailed analysis and evaluation of the situation, and after long discussions between the factory owner (the patient) and the factory director (the haematologist), it is decided to demolish the old ailing factory and equip it with new, state-of-the-art machines (stem cell transplantation), which promise the high-quality production of blood cells with a long service life.

A risky and expensive decision, but probably the only right one to stay in the business.



7. THE VALUE OF WORK

During the diagnostic period and for a year afterwards I was grateful for my full working week. I was able to switch back and forth between “being healthy and not being healthy” as needed. In addition, work distracted me and helped me to temporarily recover from my reflections because I had to devote all my concentration to my professional activities. In addition, my body got used to worse blood values and continued to function quite well for the time being. At the same time, treatment began to stabilize the blood values temporarily and prevent the disease from progressing.

My situation changed when the deterioration in blood values made blood transfusions necessary. My bone marrow no longer produced enough of its own red blood cells, so I needed a donor every now and then. One year after the diagnosis I was already in a 6-week rhythm.

Once in this state of dependency, I never got out of the transfusion requirement for a longer period of time, and by the summer of 2020, I had received over 650 blood transfusions, starting in 2010 once every 10–14 days.

The increasing dependence on transfusions gradually put me in checkmate in terms of work ability, stamina and concentration at work. It was a slow process, and more and more often I had severe fatigue and exhaustion episodes. They seemed to come out of nowhere and also disappeared again, if I was lucky. But it became more and more difficult for me to hide my illness from colleagues and staff. People talked. As head of the department I should have been the first to arrive and the last to leave. I was the last to arrive and the first to leave. Absenteeism became more frequent. The boss knew about it by now.

I was aware that others were doing part of my work. They probably noticed that I was slower than before. Ageing in fast motion, the new blood was used up quickly. Driving a car even for long distances became a great effort. Apart from work, I could no longer accommodate any mood-enhancing leisure activities due to constant exhaustion. This made me a little bit lonely, since most of my social contacts were through theatre, the choir and sport. Illness meant that these activities had to be foregone. That hurt.

I continued working on “three cylinders”, and after work I rested until it was time to go to work again. On the whole, it was not a satisfactory solution, and my family in particular watched me with a frown. It could not go on like this.

In the meantime, I had officially been assigned a degree of severe disability of 70%, which was supposed to reduce my workload by a number of hours. That sounded good, but by half past ten in the morning I would have liked to have gone home sometimes to lie down on the sofa. The willingness to continue working until 4 or 5 p.m. and perhaps have an official evening event is overly exhausting, and the only thing you can afford is to spend the rest of the day in a horizontal position, only to get up again in the morning. I was constantly looking for the next time-out: this way and that way until Christmas, until Easter, until then I could still make it. As the day of starting work came closer after the time-out, despondency and fear of being overtaxed became widespread. I needed psychological support.

In addition, the constant visits to the doctor, which had to be reported to the boss in advance, meant after a couple of hours of work, travelling 30 km, waiting at the doctor's for 30-60 minutes, having a blood test with the possibility of bad values or being subjected to another examination, the psychological processing of the results, the doctor's consultation itself, then driving back 30 km again. This required a seamless reintegration into the working world; out of the healthy world, into the sick and back into the healthy world in fast motion. All this in a period of time in which other colleagues also have the opportunity to briefly regenerate their energy or simply ‘just’ have to work. In addition, there was the postponement of meetings that had to be made up for, and material that had to be reworked. I withstood this for three more years.

Short sickness absences are not helpful, since all the work to be done is just dumped on your desk. Three months is better, then substitutes can be trained for ongoing projects. In the subsequent reintegration phase, however, it becomes apparent that things may have been decided differently than if you had driven the decision-making process yourself. Colleagues who have often wanted things to be different from you have used the time and asserted themselves. This may not affect the big concepts, but it does affect small ones, and you get an idea of how expendable you actually are. Some things work differently and the support of some employees dwindles. You are standing next to them, no longer in the middle of the work process and not yet completely outside. And you are still sick.

The employer wanted to reassign the management position and had sent clear signals that they would like to send me into early retirement. My boss asked for a prognosis of my illness, which the doctors were unable to provide. I was exhausted. I left in 2011. At the same time, I was wondering whether we as a family with two children would get by with correspondingly less money? I was 54 years old. This is not the usual time for people to leave work. But I had no choice.

My psychologist said: “I was afraid it would take even longer for you to let go.” She was pleased with me. I soon noticed that the pressure that had been weighing on me was clearly decreasing. I was relieved. As I said goodbye, I received a lot of genuine recognition and sympathy from colleagues and staff.

It was the right decision, also the right one for the whole family. At home, I was only busy resting in order to be able to work at all. My husband was the only one who looked after the children and the house. I had not been there for anyone anymore. Not even for me.



8. SADNESS AND HOPE

A small woman with drooping shoulders and a bowed head shuffled along an endless dusty road, somewhere in the middle of nowhere. Her head was covered in rags, her face was strewn with half-dried tears. As her walking became more and more tiring, she soon crouched down on a boulder at the side of the road to rest a little. After a while she saw—against the sun—a figure coming towards her that seemed to resemble her as it at first appeared. But when she approached, the little woman saw that the other woman, though not young either, was light-footed, with a straight back and a proud look. The little woman in her pitiful state slipped even more into herself and wished she were invisible.

The other bent down to her with a warm smile and asked, “Who are you?” “I, I am the Sadness”, but her voice was weak and hesitant, almost incomprehensible. “You know, I am so exhausted and discouraged because I am constantly confronted with people with incurable diseases. My sister, Fear, and I often accompany these people for many years. And because there are so many of them and they want to talk so little about their suffering, the burden is very heavy for us. My sister has gone a little further than I have, but I think she too can hardly walk any further because of exhaustion.” The voice failed with a suppressed sobbing.

“Oh, hello little Sadness, how nice to meet you,” the other woman replied as if she was greeting an old friend. “Do you know me?”, Sadness asked back suspiciously. “Of course I know you; I even know you very well. I am always near you when your sister, Fear, and you go your many tedious ways. You just don’t see me. If you’d pay more attention, we would have met more often.”

Sadness asked unbelievably “How is it that you do not run away from us? It is no joy to see us. Aren’t you afraid of me?”

“Why should I run away, you know very well that you can—if you want to—catch up with anyone, no matter if you are slow and weak or fast and strong. But tell me one thing—even if the question seems incomprehensible to you: why are you actually so sad?”

The answer came immediately: “Yes, isn’t it normal that people become sad and anxious when they are seriously ill?”

The other woman sat down next to the unfortunate little creature and put her arm around her.

The Sadness sighed deeply, she was glad that she had met someone who listened to her and she continued to say “I am so sad because no one likes me. When people see me, they are always very afraid, even more so when they see Fear who accompanies me. They try to banish me and pretend that I am not there, by pulling themselves together and by trying to be brave. They suppress their tears and suffer from all kinds of pain so that they can hardly breathe. Many of them take medication or drink alcohol so that they do not feel me. It is my job to be there for them and guide them through their illness. Only if they let me, they can look at their lives and their future. They see more clearly and will more easily say goodbye to what the illness denies them and will appreciate what they still have.



If they could accept me as a friend who comes and goes, I could help them on this path. But too many people don't see that, and nobody wants me. Too many of them end up in bitterness or give up. Only those who give space to the sadness and pain over what they have lost, and cry the suppressed tears will finally have the strength to face the illness. I can't stand being misunderstood and mistreated like this anymore,” Sadness concluded. Exhausted, she stopped speaking and sobbed loudly.

The other woman had listened to her silently, took her in her arms and comforted her gently: “Cry if you must and rest if you want to regain your strength,” whispered the woman lovingly. “From now on you and your sister will no longer be alone. I will accompany you on all paths.”

Sadness stopped crying, tried to wipe away her tears and stammered in surprise: “Who the hell are you? Tell me.” And when she looked directly at her new companion, she found that she had grown and become more beautiful. “Oh”, answered the other and smiled, “my name is Hope.”

The idea for this parable was given to me by Inge Wuthe “Das Märchen von der traurigen Traurigkeit” in: Körner, Heinz (Ed.) et al., “Alle Farben dieser Welt”, 1995.



9. I WILL CALL YOU

Recently a female friend “left” me, after more than 10 years of knowing her. To be honest, it never really worked out between us. I just always tried to overlook it. I hadn’t felt comfortable in her presence for a long time. Her readiness to accept and understand my illness was always nil, and the focus was always on her own sensitivities, which could hardly be better dramatized. Her withdrawal was not preceded by a dispute between us, only by growing estrangement.

This was the third time since I had blood cancer that people said goodbye in this way. Although each case was different, I could see a pattern. The break was not openly addressed, but rather it was pretended as if nothing was wrong—at first, they reported less and less often, eventually not at all. If one spoke directly to them about the reasons, they sidestepped the issue.

My first female friend left me in 2007—shortly after I was diagnosed with MDS—when, right at the beginning of my illness, I could no longer keep up with power walking on the mountain. I was no longer able to perform physically in the group and had to stay behind. Shortly afterwards she broke off contact completely, no phone call, no visit, nothing. The second impressive case concerned a good friend who seemed to be intensely interested in my illness, his sympathy was great, and his support was important to me. He was then himself in a very comforting phase, because his mother was also suffering from cancer. When she was healthy again, he hardly contacted me anymore. That was the beginning of the end between us. I had been familiar with both friends for years and had spent a lot of time with them before my illness.

You can make it easy on yourself and say that they were not real friends. But this is not true. There had been wonderful experiences with each other, clear mutual sympathy and selfless help over many years. That is what friendship is all about, isn’t it?

My mistake was to change the “Rules of the Game” for this unannounced: I couldn’t help it, but suddenly I confronted my friends with such a “hammer disease.” Somehow this was not agreed between us. The expectation that my illness would then be part of our relationship was obviously not fair to them. Cancer has a great intimidating power. Part of me understands that taking off might be the obvious thing to do.

Oh yes, even healthy people get dumped! And yet it is a common topic among chronically ill people. They often mourn the loss of friends. An aunt of mine, a sensitive alternative practitioner with a large circle of friends, larger than mine ever was, was seriously ill with lung disease for years before her death and lost almost all her friends during this time. The last two years she lived alone with her oxygen apparatus. This is the kind of loss that I mean.

Psychologically, the behaviour of friends is understandable: they leave because it is exhausting to be with sick people, because they often cannot be reliable, because there is little that can be done together, because they think they need to be comforted and somehow can't, because they think they can no longer communicate their own worries out of consideration, because they have the diffuse fear that something similarly threatening could happen to them one day. In this way, they put their stamp on the situation and the normal contact between two formerly friendly people is blocked.

Nobody intends to leave from the beginning. But somehow—and this is proportional to the duration of the illness—there comes a time when certain friends become more careless in their contact and eventually stay away. Most of the time, time pressure or your own health problems are the main reasons, but we all know that this is just a pretext, because a short phone call, an appointment for a meeting, a nice WhatsApp or a short email is always possible when you feel like it.

The breakaways don't feel comfortable with the "escape", and sympathy for the sufferer continues to exist in principle, but the manifold challenges of one's own everyday life gradually make contact and sympathies disappear more and more. Priorities are deliberately set differently and are vehemently defended.

How embarrassing an accidental reunion can be after weeks or months of silence. The former friend has nothing else in mind but to hurry on as quickly as possible, but pretends that everything is quite normal, possibly even a short, never-ending "I'll call you" comes about. At the next meeting by chance, it may even happen that the person in question passes by without saying hello.

When asked, the answer is "Oh, I didn't see you there." Eye contact is avoided. This is then the low point and you have to deal with unworthy and paranoid thoughts like "Maybe he or she really didn't see me?" It makes sense to really put the matter behind you.

What about me? Should I perhaps have been nicer, more generous, more lovable, more tolerant, more cheerful, bent over backwards to stop the deserters leaving? Certainly not, I had always lost at the very moment the other one slowly started their departure.

The nice thing is, there are friends who actually stay. Sometimes you're lucky enough to make new friends. Clinging is not a good option, anyway.



10. LOOKING BEYOND THE HORIZON

It took me at least a year before I had enough distance from my illness to talk about it with outsiders. Even longer before I was able to use the experiences of other patients for myself, to finally reach a point where I could become a support for other patients, which at the same time was also a help for me, up until today.

Many acutely ill patients do not even have the time to deal intensively with the disease and accept it, because it progresses rapidly and aggressive therapies must be used. Before some people have hardly understood what is actually happening to them, they may already be confronted with the decision to undergo stem cell transplantation.

I am lucky. For some medical reasons, which are beyond me and—yes—beyond my medical team, my progress is gradually slow and my status has been stable at a low level for several years. There are only theories as to why this is so, no evidence. Despite all the limitations, it is a great happiness without a net or false bottom, but isn't that the essence of happiness? At any time, a life-threatening condition can arise for me too. It seems to be part of my fate to have met many patients who were clearly better off than I was at the time of our meeting, but whose condition then suddenly or even gradually deteriorated. They passed me by and are no longer with us.

Contacts with other sick people take on a different character under the impression of my own serious chronic illness. Cancer and fear are connected. But when it comes to pain and deep sadness, everyone lies alone on the sofa at home and tries to get along somehow.

The certainty that one is not alone can bring relief. But the fear is not reduced by someone saying, "I have an illness as serious as yours." Anxiety is only reduced if one experiences the time, the individual therapy steps and pain with the realization "I can stand it, so I'll keep enduring it!" Overcoming part of the fear requires trust in oneself, in one's own path and in the doctors. The fear will never disappear completely. The matter of course with which one used to be healthy, so healthy that one did not even have to think about the fact that this condition could ever end, this matter of course will never return. I know from many successfully transplanted patients that the fear of a relapse is constantly breathing down their necks, and above all the fear of routine check-ups, the fear of having to deal with the question of whether and how long they will survive.

When I was still working, but was already ill, I once had a colleague sitting in front of me, who, due to an absence of several weeks due to illness, had ruined an important work assignment. It was a tricky personnel discussion, because with sufficient planning beforehand and, above all, with the possibility that other employees could have accessed the necessary data, there would have been no problem at all. The colleague, however, was in an unimaginably bad mental state, close to tears. It was not a smell of sweat: I could smell his fear. He had experienced something very bad in hospital. He was suspected to have leukaemia, he confessed to me. Seriously frightened, he had undergone a test and I asked him about it. No, the result was not yet available. He did not seem to be surprised that I asked him about the test, because as a person affected, I only knew recently that only this examination could clarify whether leukaemia was present or not. He was in the situation in which I had been immediately before the diagnosis, in which one can hardly hide one's psychological state, one walks around outside with tears in one's eyes, in which one tends to tell everyone who confronts one with a more insistent "How are you?", a phase in which one feels threatened by a life-threatening illness and in which one feels one's personal finiteness very intensely for the first time. I knew exactly where he was standing, namely where the pressure on his chest seems unbearable and it really hurts.

I took a deep breath and broke through my principle not to come out of my professional environment yet, and I told him about myself. He took up my words almost without comment; my story had no point for him now, even if it should have the purpose of uniting me with him. He collapsed even more and left my office very soon after we had clarified the problem. At my request, he promised to talk to me.

At my request he also promised to report his diagnosis in due course and to keep it confidential.

The result of his examination finally showed that there was no leukaemia, but in fact a treatable viral disease, which had temporarily severely affected the blood values. In addition to the sincere joy I expressed to him, I had an unpleasant feeling, a feeling that one is not usually very proud of: Envy, thick green envy.

In April 2010, a friend from a small leukaemia forum on the Internet died. She had had MDS for almost seven years and was only 51 years old, in itself a young age for this disease. Until December 2009 her blood values had always been better than mine. I had not only gotten to know her as a chat partner, but also in real life at several forum meetings. It had always been clear to me and her that not she but I would be the next one to need a transplant. But we were wrong. From autumn 2009 onwards she was getting worse and worse, not that you could see it in her blood values, but she had one infection after the other and was prescribed lots of antibiotics. Around Christmas her whole system suddenly collapsed, her blood values crashed completely, a high number of blasts in bone marrow and blood were detectable, and her condition had turned into acute leukaemia within a few weeks. She was in extreme danger of death. Within a few days she had to accept—struggling with pneumonia—that a transplant was the only possible immediate rescue.

Under a high dose of painkillers, she celebrated Christmas with her family. We spoke on the phone on 23 December. In the end she could not get rid of the infections and her organs were completely overloaded by the constant medication. The stem cell transplantation was carried out nevertheless, as there was no alternative. The transplantation was successful, the stem cells grew well, but my friend died after three weeks of artificially induced coma from multiple organ failure.

What had happened? Had the transplantation been delayed too long, or had the doctors chosen the wrong moment of intervention? Torturous questions. Or did she simply fulfil the statistics we all know, about which she and I had so often respectfully speculated? With fighting spirit, great hope and the counter-argument of individual fates, we had cheated ourselves out of the statistics every time. “We are young and our organs are healthy”, we had always assured ourselves, “this will not happen to us in the case of transplantation.” It happened to her. She’s dead.

For our little leukaemia forum, the agony of losing this beloved woman was a very big blow. We were all paralyzed for weeks. It was very difficult to find new words of sympathy over and over again. In the end only the word “miracle” was used, a miracle was supposed to happen for her. Even when she was put into an artificial coma and her kidneys were no longer working properly, we still hoped for a miracle that did not happen. It came to pass that her suffering was first presented by herself, then by her family in the forum in many shocking details. For example, it was described how she became temporarily blind under the high dose of chemotherapy. I was horrified and couldn’t escape the news; it was like an avalanche.

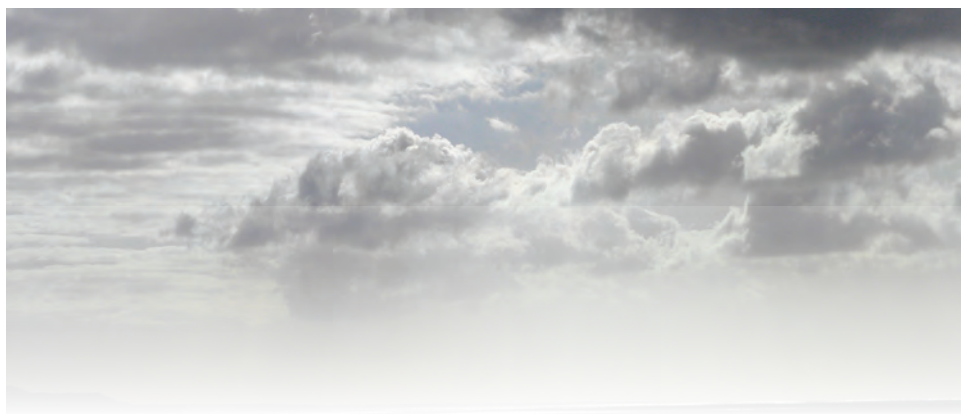
Today, a long time after the death of my friend, I find the bereavement voyeurism to which we were at the mercy of in her case, and which we had helped to promote, was ethically inappropriate, dignity-eroding for her, unnecessary and disrespectful of us; this is a danger of the digital opportunities now before us. Today, I am running a closed MDS forum on the Internet, and I would try to prevent such a vivid depiction of suffering at all costs.

There is always the first time that such an impact is being felt and for a while it covers up everyday life in a paralysing way. The first cut is the deepest. Such a death, such a loss, feeds the fear of possible similar experiences of one’s own. In the subsequent years until today, some losses were cut into my circle, which were very close to me.

In the beginning, the view beyond the end of one’s own nose serves only one purpose, namely to inform about one’s potential chances of survival. One wants to know, as a person who is ill themselves, how long the other person has been living with this illness, and then one concludes on one’s own chances. The question is all the more urgent if it is the same MDS subtype. The personal status is compared and one is very relieved if the other person has been living with the disease for longer than the length of time one has been suffering from it. This is then the proof that you still have at least this additional time. Only with time do you learn how different individual courses of events can be, and that comparisons of individual cases often have little meaning.

Of course, there have been some patients who are well known to me who could be cured by stem cell transplantation, although many of them with significant limitations.

What is really tragic, however, is that in the 15 years of my disease, the survival statistics have hardly improved at all and transplantation remains the only cure. The few therapies available are only temporarily effective and this at most for 2/3 of the patients or less. Whether this will change with the current knowledge that cancer apparently splits up into many diseases with the most diverse genetic mutations remains to be seen. We are currently experiencing a profound change in perspective. There are already promising immunotherapies for individual types of cancer, based on new findings. These personalised therapies are not only incredibly expensive, but some of them also have uncontrollable side effects. Nevertheless, there is a gold-rush atmosphere in the research community. As far as myelodysplastic syndromes are concerned, there have been breakthroughs in diagnostics, but not in the field of therapy. Not really.



11. MY SUPREME JUDGE

I can well imagine how it was in earlier times when medicine did not know the individual constituents of blood, but thought it was possible to determine that the life force—the blood—was diseased in certain patients and therefore had to be drained, in the same way that tumours are cut out of healthy tissue. For many centuries, bloodletting was one of the main procedures in medicine. Even today, bloodletting is still used for certain indications, for example to drain excess iron from the blood. For this to happen, however, blood formation must function properly, as the blood drawn off must be replicated by the body. This may be the case with various types of blood cancer, i.e. malfunctions in the bone marrow and/or blood cells, but is no longer always the case.

So, what can be done if the blood is sick and there are no effective drugs to treat it? One can supply healthy foreign blood (blood transfusions) or one has to renew the place where the blood is formed (through bone marrow transplantation).

In the beginning I did not understand much about the processes of blood formation and the importance of red blood cells, but as a newcomer to MDS, I was very quickly forced to get acquainted with the haemoglobin value (abbreviated Hb value). From this point on, this was my top guideline for my general condition and my psychological stability.

Haemoglobin is the red blood pigment, which is why blood is red. The haemoglobin in the red blood cells contains iron and is therefore capable of binding and transporting oxygen. It absorbs oxygen in the lungs and transports it via the bloodstream to the whole body: cells and tissues—especially muscles and organs—need to be supplied. On the way back, the haemoglobin ensures the removal of carbon dioxide, which is finally exhaled via the lungs.

If there are too few red blood cells, the haemoglobin level is low. The body is no longer supplied with sufficient oxygen. One shows obvious symptoms of anaemia, including weakness, tiredness and shortness of breath during exertion.

When I give a blood sample and look at the sheet with my lab results, my first glance always falls on the haemoglobin level and it subsequently defines much of my mood that day. There are many other values in a small or large blood count that reflect the general condition and the trend of the disease course, but I have been conditioned to rely on the

haemoglobin value since my diagnosis. For me it is a water level indicator of how full my power source is, whether I can get enough air, walk normally or even climb stairs. It is my number for quality of life. It largely determines when I receive a blood transfusion.

After so many years of experience with anaemia, however, it has become a rather reliable confirmation of my current condition and no longer represents a threatening surprise in itself. In most cases, I can predict the haemoglobin count to within 0.3 g/dl before the blood is drawn. I love to confuse young interns with this.

In the early days of my disease, when I was overwhelmed with all the new demands of being sick and at the mercy of the reference value of haemoglobin every time, I once stormed out of the haematologist's office crying and could not calm down at all. I continued to cry for hours at home as well. I had read my haemoglobin value from the display of the evaluation machine in the practice.

It had fallen off massively compared to the last time. In my anxiety this came close to a death sentence; I was lost until my husband called the haematologist to discover that it was not my haemoglobin value at all, but that of my predecessor. The machine had not yet adjusted to my values so quickly. My haemoglobin level was much higher. The power of a single number can be very great.

Today, my average haemoglobin level is much lower than the one I had mistakenly read then. Perspectives and feelings of security change over such a long period of illness. Nevertheless, my mood is always worse at 7.9 than at 8.1; a minimal difference, possibly only depending on the time of day or laboratory, but the 8 before the decimal point makes the decisive difference in my head.

What a miracle our body is! In healthy people the haemoglobin value is about 12-16 g/dl, but when it drops, the body adapts and the haemoglobin can still do its job to some extent. One should then simply not demand too much effort from the body. Only when the value drops below 6-8 g/dl does the oxygen supply slowly become critical. This is an enormously large and ingenious buffer that nature has developed for us.



12. YESTERDAY, YOU COULD WALK SO FAR

The expression in my friend's eyes was panic with a capital "P." Because of me, we were so slow and almost missed the train home from Berlin. We barely caught it, although "my" elevator simply wouldn't start on the platform. My girlfriend rushed up the stairs with luggage, hers and mine of course.

I felt so sorry for her because she was afraid I was going to collapse right away, and unfortunately, I shared her concern: Damned tachycardia and pudding legs. But then everything went well again.

I spent three days in Berlin with my two oldest friends. We, two women and a man, have known each other for almost 50 years and live in three different cities. It was a wonderful weekend, unusually sunny and warm for early November at Wannsee. I finally had some real fun again, profundity, irrepressible laughter and challenging conversations, combined with a quick walk through the last 50 years. We could not stop talking at all.

My friends know about my illness: "Can you still?" "Shall we sit down again?" "Would you like to rest?" "Give me your bag." "Should we take a taxi for this route?" caringly probing, almost to my boundaries of privacy. Why should I say "shame", why can't I still deal with it when others offer help because I need it?

In Berlin, travel is always hard work. Of course, we took taxis and buses and the S-Bahn, but not the U-Bahn because of the many non-functional escalators. But I couldn't admit that even switching between means of transport was hard work for me.

I felt like the Little Mermaid with her fish tail from the Andersen fairy tale. She was given legs to be with her human love, but only at the cost of unspeakable pain with every step. She endures and smiles. I had to stop, otherwise I would have had to leave to go to the hotel for hours or even until the next morning. After all, I rested for three hours in the afternoon on the second day, but unfortunately that was not enough to recover.

To explain: I took 50 spoons with me for the Berlin trip, where one spoon corresponds to one energy unit. Fifteen spoons per day plus five spoons of iron supply. I should have managed better, but could I? Taking a shower already took me two spoons, a 15-minute walk at least five, and a long conversation with concentrated listening definitely three. In

the evening before the journey home after a walk at the Wannsee, suburban train back and forth, going out for dinner, incessantly lively conversations, cinema and pub until 1.00 a.m. with two Campari Oranges, I had only two spoons left. Too little to walk from the hotel to the train station in the morning plus the stairs in the train station, a walk that by the way takes only 10 minutes for healthy people and has no gradient at all. I already suspected that my body would show me the red card on the evening of the strenuous day before, but I still had to hope to manage the circumstances of the trip.

“Yesterday you were able to run much longer and faster”, my friend wondered in shock. “Yes, and that’s exactly why I can’t do it today.”

The original spoon theory is found in the book “The Spoon Theory” by Christine Miserandino. Christine has lupus, an autoimmune disease. She invented the ingenious spoon comparison to describe this debilitating chronic disease to her friend. The name of Christine’s website alone made me grin, it’s www.butyoudontlooksick.com. This woman knows all about it. You don’t look sick, but you are.





13. THE WEEKEND TRAP

It is Friday afternoon and I am sitting in the ICE train on the way back from F. to G. with a haemoglobin value of 7.8 g/dl and. I barely made it into the train: exhausted, my pulse much too fast, all ways arduous, the air scarce and my muscles aching. Luckily, I only had my handbag as luggage, carrying anything heavier is now impossible.

Because of the upcoming carnival weekend, the train is overcrowded and I just save myself a seat in the dining car. Making a seat reservation is not possible on my trips to a clinic 300 km away for treatment with a study drug, because I never know how long the waiting time and treatment will take.

I hold on to my cup of coffee and hope that the trip will end soon so that I can get back to my sofa at home.

Friday. Only on Monday I can get a blood transfusion in G., that is two days and many hours left. For me this is a long time, unless I go to the emergency room, which would cost me many hours of waiting before I even get anywhere near my destination. I do not have the strength for that.

The haemoglobin level will continue to drop for the next two days. I know exactly how bad I'll feel over the weekend until Monday morning when I'll finally be able to drag myself to the outpatient day clinic to be transfused with two bags of red blood. With the time it takes the blood bank to prepare the transfusion for me, it will be Monday noon until the needle is in place and the blood is running. Until then, it's a matter of keeping your nerve and keeping the condition manageable. Unfortunately, sleeping a lot does not help. In this oxygen-deficient state I am sleepless and restless, highly nervous and irritable. I am a blood junky. Addicted. Every 10-15 days I need fresh blood, otherwise I feel bad.

Actually, I have been a professional in organizing my blood transfusions for many years. When I clearly feel the symptoms of anaemia, I call the day clinic in G., get an appointment immediately, go there and after 6-7 hours I usually leave the clinic freshly doped. But holidays and weekends are the enemy of all planning. The haemoglobin level and the state of health before that can be bearable, but if you are not careful, you will reach your low exactly on weekends and holidays. Christmas and Easter, for example, are major logistical

challenges. So you have to keep a close eye on the possible time for a follow-up transfusion and keep it on your calendar.

This time, the dilemma is beyond control and I end up exactly where I never want to be on the weekend, namely on hold. This is because I have my therapy appointment in F. today on Friday, but at the same time I should have had a transfusion because of my condition. A planning error, but difficult to avoid.

I thought that I could manage that and that I could also get a blood transfusion in F. if necessary. I miscalculated, my haemoglobin value is not where it should have been from experience, but even lower. My red blood cells, donated and supplied to me by strangers, have been used up faster than usual this time.

Since I am not an emergency case, it is not possible to order blood for me on an outpatient basis before the weekend on a Friday afternoon in F. in the clinic, because it takes between two to four hours to test whether the possible donor blood actually matches the recipient. After my long transfusion career, antibodies have been formed in my blood against certain components of donor blood that is actually suitable. This, too, must be meticulously tested in the blood bank to exclude these unsuitable donors for me. Otherwise a life-threatening allergic shock can occur. Once the blood bank has released the blood products, it takes another two hours for the blood to pass through my system. That means I would have to stay in the day clinic until 7 or 8 o'clock in the evening. But no staff would be there to look after me after 5 p.m. at the clinic in F.

So I fell into the famous weekend trap, and the trap snapped shut. The only other solution would be to have no other appointments on Fridays and Mondays in case I might need a transfusion. This time, however, there was an "overlap of interests" and I am now dealing with it.

The statements of the hospital doctor in F., which are all too familiar to me, accompany my train journey home. As if I didn't know that: "On Monday you absolutely must have a transfusion in G. On the weekend you may feel a little worse. In case of abnormalities (i.e. severe shortness of breath and heart problems), please go to the emergency room immediately, even on weekends. But don't worry too much, there is no immediate danger of a heart attack.



14. OBSTACLES

Three times a month I have to spend almost a whole day in the outpatient department of the day clinic of a university hospital for the purpose of receiving supportive therapy for my MDS disease. These therapies keep me alive. These therapies include transfusions of red blood cells, as my bone marrow no longer produces enough of them, and the administration of immunoglobulins to support my immune system so that not every germ or fungus can penetrate me unhindered and destroy me.

It goes without saying that I am not feeling very well in the mornings of these days. I have massive anaemia symptoms at this time, which are supposed to be corrected by the eagerly awaited transfusion. Driving with dizziness or unexpected microsleep is out of the question. So I need a chauffeur, my husband, whom I really do not envy on such days. Getting up early at 6:00 a.m. really gets on my nerves. Head and legs are rather leaden and during the 40-minute drive to the clinic, which is prone to traffic jams, I usually don't say a word because talking is much too exhausting for me.

When I do say something—my husband says—it is obviously too quiet and too squeaky. It may be that I acknowledge the question of what I have said with an irritated “nothing.” My thoughts are like thick mush to repeat themselves, and explaining something seems impossible. I am a real monster without enough oxygen for my brain cells.

Arriving at the university hospital, the first thing to do is to find a parking space as close as possible to the entrance so that I don't have to walk that far. Since we are there early, this is rarely a problem. Most of the time it is the parking lot where half of the car is on the rather high curb. I am pleased every time we come back from the clinic in the afternoon and the left mirror is still on. But I keep these thoughts to myself, because discussion is impossible and my husband would rightly ask me what I really want, a short way or a comfortable parking place? Of course, he's right, even though the tilting position of the car on the sidewalk makes it so difficult to open the passenger door that I can't hold it open with the little energy I have to get out. So my husband has to go around the car and literally pull me out, always with the same grip.

Once the parking issue is dealt with, there is still this and that to be removed from the car to make it through a long day at the day clinic. Freed from any carrying duties, I start wiggling towards the clinic entrance, past the still unoccupied ice cream stand and the stinking concrete ash pails for the smokers, until I reach the sign at the entrance door “If you suspect contagious diseases, please do not enter, but ring the bell.” Then I wobble slowly on, past the doorman, along the first of the three corridors. In the second corridor, directly opposite the prayer room, I usually need a little breather. If I have insufficient power to continue, there’s a bench right there, where I then sit down for a short time.

I have already overcome three obstacles typical for these days: getting up, driving and getting out of the car. Now I have to master the way to the day clinic until the end, where my husband has long since caught up with me and is waiting to help me. Afterwards, I have to pass the conversation at the admissions desk of the day clinic, and without talking, it is not possible.

No one who is healthy can really imagine how much I have already achieved by then.



15. FATIGUE

All well-informed cancer patients, doctors, psychologists and relevant literature give three main pieces of advice to combat fatigue in everyday life:

1. To be more in harmony with your body. To feel when you need rest.
2. To develop an active and positive attitude towards life.
3. Exercise.

This is good advice with great theoretical value. But often comparable to the advice to a calm person to be spontaneous. In fact, the suggestion to move outside in the fresh air is also of great practical value. I have to admit that it significantly reduces feelings of tiredness and exhaustion, at least for a while. Even resting afterwards feels better than if you had been lying down for a while.

Because I suffer from anaemia almost constantly, a large part of my fatigue is due to the fact that my muscles, organs and brain are not supplied with enough oxygen.

But fatigue also comes when I am well transfused and should be functioning. Chronic fatigue is an almost constant state of weakness that limits my energy and mental abilities. Fatigue therefore also affects my emotional and psychological well-being. With fatigue my body feels heavy, as if I'm hatching a cold, I feel dizzy more often and I can hear my heart beating in my ears.

With fatigue, I suffer from a partial performance weakness during a parlour game such as Scrabble. I'm still good at forming new words, but I have trouble just adding up higher double-digit numbers. I have to concentrate very hard and it takes a long time. I hardly retain foreign language vocabulary any more. In phases of severe fatigue, I can't express myself in a sophisticated way, I can't think of words, especially names. In such situations I don't like it at all when I have to explain something in a lengthy way.

Above all, the effects on my mental abilities change parts of my personality, which is hard for me to deal with. Fatigue often dominates my everyday life, and it disturbs my quality of life.

Because of fatigue, I had to retire early and accept limits on my travel plans. Because of fatigue, I often have to cancel an appointment because another appointment a few hours earlier has been so stressful that I can no longer keep another one. When time is short, I am not able to accelerate and hurry. Showering takes a long time and exhausts me. If I am on the road with other people, I should know the route beforehand and know if there is a bench on the way for safety reasons. Of course I prefer elevators and escalators to stairs. My family members know my unhappy look when the escalators in the Berlin subway are broken at some stations or there are none at all. Then they push me—one hand firmly on my back—up the steep stairs, with a break after the first landing.

I have survived my blood cancer for so many years. This is the second best solution after a cure for the time being. So with a certain equanimity I obey the rules of fatigue. I have to, because it has not only happened once that I have become physically completely overburdened. In such cases the body will completely stop after I ignore the warning signs and I cannot go one meter further; one or two days of absolute rest are then necessary to get back on my feet. Just like when I climbed a church tower with my mother, which did not have as many steps as it seemed to me. The ascent was arduous, but went quite well. With a certain elation I reached the top of the church tower and could enjoy the view. But the descent made me pay the price. Never before have my legs trembled so much and I had such painful cramps. I had to slide down the stairs on my backside as the last of the group, to questioning looks from others.

It took a long time for fatigue to be recognised as a “disease” by doctors and the health service. I cannot say that in the course of my illness this common “side effect” of cancer was often made a topic of discussion between my doctors and myself. Collateral damage. The blood count and the clinical picture are more important. To this day, fatigue does not count as a separate disease for recognition or upgrading of the degree of disability. Although it has been known for a long time that fatigue does not necessarily disappear after successful treatment of an underlying disease, but can become independent and may continue to torture those affected for years.

Consequently, there is almost always a dark guest at my table whom I have not invited. When I want to enjoy an evening with friends, I sometimes manage to scare him away. Faced with the choice between a chronic form of the disease—which is usually bearable and exhausting—and a form that overruns me and calls for aggressive, potentially life-threatening therapies, I calmly and submissively choose the first option, including fatigue.



16. THE THRILL OF VOLUNTEERING

At the time of my diagnosis in 2006/07, self-help groups in the blood cancer field were not very evident to those affected. They were not as conspicuously advertised in the waiting rooms as they are today. Only with the spread of social media has access to self-help groups for rare diseases become easy. Today, large self-help groups call themselves patient organisations and they are professional service associations with a budget, which are not only organised locally and regionally but are also members of federal associations. Often, they are also networked at European and international level.

With my myelodysplastic syndromes I was all alone at the beginning. My family doctor and specialist did not have any helpful addresses of leukaemia groups, let alone MDS. But I wanted contact with people who were affected in the same way, wanted to get to know comparable cases and hear the experiences of other patients. I wanted to share the pressure of the threat and not carry it alone.

Finally, after long research on the Internet, I found what I was looking for, but the appropriate contact was only established through someone who knew someone who knew where to turn.

For me, the entry into the world of self-help was a rescue from the overload caused by the diagnosis. Suddenly, my own illness was no longer just suffering, but valuable knowledge and usable experience that I could share with other patients and their relatives. I was also a source of information, because what I had to say about my MDS history really interested other patients and, above all, other players in the health care system. This in turn helped me, gave me the incentive to better understand the disease and to become a more empowered patient who would take responsibility for themselves and speak to the practitioners on an equal footing. It was easy for me to attract the attention of doctors and pharmaceutical representatives at patient days, because I succeeded not only in telling about my own experience of the disease, but also in lifting the anecdotal into the universal, in being an exemplary case and not just having a difficult fate. I became a data source that could be tapped.



The motives for getting involved in self-help are nourished by a combination of altruistic and selfish motives. For me, this combination has a healing effect. In the meantime, I am a trained patient advocate, who works on the level of general patient needs, which I support because they are or could be my own needs and interests. I understand patients from my own experience in all states of bearing illness. I know what makes them tick despite all the differences. The initial stigma of being reduced to “sick and disabled” has

dissolved, and I can help other patients to soften their own stigma by changing their perspective. This is good for them and me too.

Behind the conference booth, the association members provide interested people with information and materials about the disease, as well as self-help, networking with other patient representatives, further training at specialist congresses and finally the publication of these experiences in articles or on Internet platforms. All of this has taken away much of the uncertainty that the diagnosis and prognosis of the disease initially caused me. These new possibilities have rebuilt my deeply hurt self-esteem and closed psychological wounds: THIS is MY kick in volunteering.

I find many motives for voluntary work confirmed in me in many ways. It is my concern to help other patients and their relatives to cope better with their illness by providing them with the right information and the best possible care. I am generally happy to take responsibility for promising projects. With my commitment, I also fulfil a social task. I organise interests that are difficult to organise. Self-help is a necessary and support-worthy addition to the health system. In my case, however, selfish motives probably outweigh this. Voluntary work is meaningful for me, especially after the signature of the public health officer, who declared me, at 54, to be permanently too incapacitated to be able to work. Of course, my new voluntary work has given me a new sense of social integration. I have met interesting people from all over Germany and from many different countries, built up new and sustainable connections and have been able to acquire completely new knowledge and insights into other health systems, research and development and the different ways in which those affected deal with illness.

I was also able to contribute many skills and abilities from my earlier professional life.

I would like to do much more, more continuously, more intensively, more responsibly. But it is not possible. I am sick and I notice it—every day! Physical strain from travelling, long periods of sitting, and permanent concentration make me tired and weak. This is the stamp of real incapacity to work. But if you define my ability to work not only from an economic and performance-related point of view, but as a voluntary, sporadic and social commitment that has a positive effect on others, then I can say that I have actually worked a lot in the last 10 years and still do so.

Pure self-help, more care-oriented, and policy-oriented patient advocacy aspects overlap. The community of patient representatives is heterogeneous. Understandably, there are rather

few patient representatives in acute illness because they cannot cope with the job-like demands of patient representation due to their illness. A larger proportion are “cured” patients with varying degrees of general health, who use a lot of energy and ambition to change the inadequacies of the health care system that they have experienced first-hand. Relatives as experienced insiders of the current or former patients also more often find their way into such a commitment. The group of professionals who have studied health management, social work or health economics, for example, is becoming ever larger. This is about a job and career opportunities in non-profit organisations. If there is a lack of empathy, then the actual patients are no longer listened to properly and their real needs are sacrificed to economic and political interests.

Larger, professional patient organisations form stable structures due to their large number of members and their acquired financial strength and can afford to employ trained business economists or legally trained people as managing directors, secretarial administrators or public relations officers. These associations are heavyweights in the community because they not only “take care” of patient needs, but also represent political interests in important bodies of the health system and shape patient representation in an innovative way.

Smaller associations, on the other hand, find it harder to defend their existence and are heavily dependent on the leadership of the first chairman and the willingness of the board to work. Even though associations are considered the nucleus of the culture of democracy, the competition mechanisms are similar to those in “normal” professional life, especially when positions are not clearly defined and areas of work are not clearly demarcated. When the need of individuals for power and recognition is paramount, and apparent altruism is a particularly sophisticated strategy of selfish self-realization, it is difficult for newcomers to work in a result-oriented manner. In the beginning you are encouraged and challenged because you obviously have important things to contribute. But soon the competitive awareness of the top dogs awakens. Perhaps you are too fast, too good, too critical, too demanding, and that is disturbing. If you haven’t been able to build up your own power by then, you are in danger of being put out of business. It is a big mistake to assume that in a social organization with the motto “everything for the patients” you would find yourself in a cosy working atmosphere. In the case of a conflict it becomes clear that there is no “occupational safety” at all and the person affected has no rights, not even the right to demand a clarifying conflict discussion. Long live the works or staff council in companies and in the public sector! Long live conflict management like mediation. A club statute hardly helps there. In small associations the “Wild West” rules! One can only withdraw, change the club or found a new one. This often happens, fluctuation and personnel shifts in the organizations are frequent.

Almost everyone who has ever belonged to an association for a longer period of time knows such stories of overly dominant chairmen or board members and resulting conflicts: This is the other side of the shiny medal of honour. The currency of “recognition”, which is coveted by everyone, is highly competitive.

The “congress hopping” in an internationally networked patient organisation is another kick. All in English, of course: “Hi, you’re here too. How are you? Where did we actually meet the last time? I think it was in Vienna or was it in Copenhagen? No, it was in Madrid in October last year.”

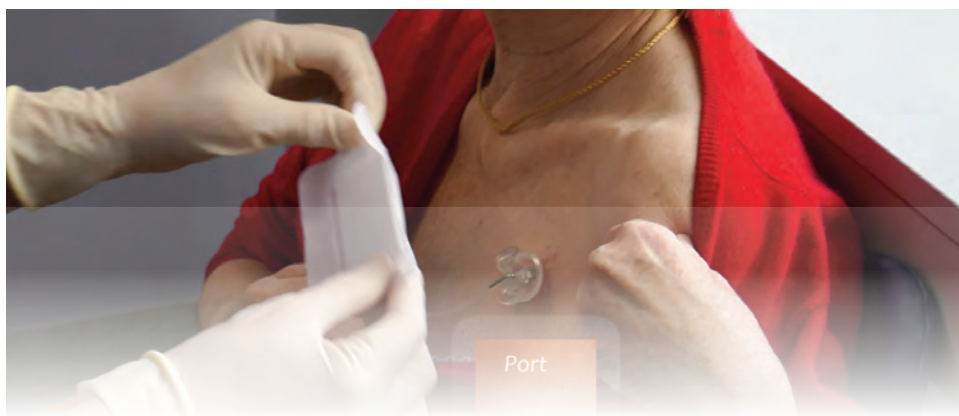


However, the meetings, lectures or seminars last all day long and usually take place in windowless, air-conditioned conference rooms of hotels, which all look the same. When one is done with it, the patient is often so exhausted that a trip to a sun terrace, sightseeing or a culinary highlight, which is often also “on business”, is too much, as one would prefer to simply relax in the hotel after the conference strain.

I often came back from a congress from abroad and afterwards, I had to lay down in bed for three days due to complete exhaustion. Apart from the fact that the relatives are not very enthusiastic about such consequences, as they had already advised against it before due to the effort, this price is

actually too high. This realization has led me to attend only a few well-chosen congresses a year. For some years now I have cut back my travel activities quite a bit, but I am always tempted by interesting external inquiries to go beyond my limits.

As a volunteer patient representative, you are the only one at the table of stakeholders who is not professionally trained for your activities. It is therefore more than logical to demand that patient representatives be equipped with the appropriate professional negotiating skills. One is also the only contributor who is not remunerated for his or her activities, which is what volunteering is all about. The only problem is that the same demands are made on the volunteers on the parquet of professional players in terms of time and performance as on the professionals employed in the health sector. This makes it clear once again how much we volunteer patient representatives actually do, and how necessary professional training is for us.



17. NEEDLES AND PINS

“Needles and Pins” is a song title from the 60’s, which has nothing to do with taking blood or illness, but with lovesickness. Nevertheless, the title fits as a symbol for incessant needle attacks on sensitive parts of the body: painful and distressing.

When my oldest son was six or seven years old, he was to have blood drawn for the first time in his life. Our family doctor, who normally left the blood tests to his receptionists, took care of it himself in this case, “so that the boy will not be afraid of it in the future.” I was thrilled by such empathy, although I now know from experience that nurses are often more skilled than doctors simply because they take blood more often than doctors.

Before I had Myelodysplastic Syndromes (MDS), taking blood was a walk in the park for me, even though I have quite deep veins. But after two or three years of regular close blood monitoring, my veins became more and more scarred and it became increasingly difficult to find a way into them. In addition, it usually really hurt most of the time, and the pain depended very much on the person who took the blood and was determined by how many attempts were necessary. An unwritten rule was that after two unsuccessful stabbing attempts, someone else was called for it. As time went by, an average of three nurses tampered with me one after the other, and sometimes a doctor who was under pressure in such a situation to bring the matter to a successful conclusion. This meant that the needle was often simply pounded in with a firm push, which caused me a brief outcry.

The whole procedure was accompanied by discussions of all participants about the strength and angle of the impact, including the question to me, where did it work last time? Depending on how the needle was applied, I usually knew beforehand whether it would work or not, but at this point the directional decision had already been made long ago and my know-it-all attitude was no longer effective.

Depending on my mood, the failures were dealt with in different ways. Most of them apologized for the pain they caused me, others blamed my supposed rolling veins or the fact that I did not keep still enough. Still others were so frustrated that I had to cheer them up. It was teamwork, with beads of sweat on all of us, only I was often close to tears.

When it finally worked, my forearms were punctured and bruised in several places and hardly recovered until the next procedure two weeks later.

I was not the only patient with these problems, as you could see from the number of bandages in the waiting room. A mutually understanding look and short conversations ensued. We described our lot with a mixture of whining and black humour.

Eventually it was suggested that I should have a port in my upper thorax through which blood could be drawn and given without any problems. The short outpatient procedure for this was quickly forgotten. Since then, my relationship to taking blood has relaxed again. There are experts who can hit the small membrane under the skin painlessly and better than others. Unfortunately, there are also pricks and punctures that hurt you, but on average, taking blood is no longer a big deal, especially on a haematology ward where a double-digit number of ports are punctured every day.

Unfortunately, the veins do not recover, and scarred is scarred. In foreign clinics the nurses often try to persuade me to take blood via the veins, because taking blood via a vein is less effort and less germs. If I let them, they usually give up after the second attempt and still use the port system.

Lack of hygiene is a dangerous trap. Although I always pay meticulous attention to the necessary hygienic measures of care myself, after five years of wearing a port in connection with a transfusion, a germ has crept into the system. Just two hours after therapy, I had a feeling of sore muscles at the port site: Saturday morning admission to the emergency room, with signs of inflammation and a high fever. The horror of all port wearers: a port infection. Operation on Sunday; the old port out, a new one on the other side of the chest in. A hospital germ, which fortunately responded well to antibiotics. After that, hospital for two weeks with six bottles of two different antibiotics per day. Not the ward doctor, but the director of the microbiological institute decided on my discharge. Temporary needles and pins into a vein, every morning for blood collection and without port, because the new one had not healed yet.

Painful memories and a surly mood. So “wearing a port” does involve some risk. Port infections are—unfortunately—not rare.





18. FOCUS ON THE PATIENT

What does patient involvement mean?

Nearly all actors in the health care system strive for the slogan “Focus on the patient.” It can be found in scientific publications, as a corporate philosophy of the pharmaceutical industry, especially in glossy brochures, as a congress title, as a title for national health initiatives, in explanatory texts on laws and in statements by politicians.

The 20-year-plus old story about “Patients in Focus”, an article in the German Medical Journal from 1999 with the title “Communication in Medicine: Patients in Focus”, should be mentioned as an example. This reported on the 1st Koblenz Patients’ Day in November 1999. The congress was organised by “Kirstin’s Weg—Verein zur Förderung der Krebsmedizin e.V.”, an association founded by a patient suffering from cancer in Neuwied. *The Deutsches Ärzte blatt* 1999; 96 (p.31-32) described the patient as follows: “Even as a patient, she never allowed herself to become an object, became active herself, took care of raising funds for cancer research and suggested the establishment of a support association.” This association has the statutory mission to support patient-oriented cancer research, the model of open medicine and programmes to improve the position, competence and orientation of patients. In this sense Kirstin Diehl was already a professional patient representative in 1999, as we would say today.

Back to the beginning: The slogan “the patient in the centre of attention” has catchword character. It is interpreted by all interest groups in their own way. Such slogans are simple and always have a positive connotation. Here, an end point of a development that has not yet ended is assumed.

The slow dissolution of paternalistic structures in the medical profession is only just taking place, due to generational factors, and is more hesitant in Germany than in many other European countries, where there are increasing health initiatives and projects in which all those involved in the health sector work together. In Germany, patients and the general public are increasingly being told “from above” that the patient, who was previously seen as a weak and passive recipient of health care, must be more closely involved in planning and decision-making processes by the professional players in the health care market.

However, it is seldom defined exactly what this should look like. The political will is seldom shown in a concrete form.

The most recent example of a possible future-oriented cooperation between partners in the health care system is the “National Decade against Cancer”, established by the Ministries of Education and Research and Health in January 2019. Two prominent patient representatives from self-help organisations are, after all, represented in the impulse-giving strategy circle. The strategy circle defines goals and fields of action and initiates corresponding activities of the “National Decade against Cancer.” The aim is to work together for strong cancer research that closely involves patients. This is not unlike the above-mentioned project at the “1st Koblenz Patients’ Day” in 1999. It is to be hoped that this renewed announcement for the 10-year project “National Decade against Cancer” will also be reflected in concrete results that bear the direct stamp of patient interests and needs.

It is not possible to do both: offer participation and at the same time retain authority to act and total control. Friction between the actors and the patient community is inevitable and win-win solutions must be found. “Nothing about us, without us!” and “Patients in—not out!” are combative statements that demand real patient participation. “Nothing about us, without us” aims at joint decision-making in medical treatment between doctor and patient (shared-decision-making), at having a say in health policy and at patient participation in research and drug development (R&D). “Patients in—not out” was a protest reaction of European patient representatives between 2013-2015 to repeated attempts to exclude patient representatives from the industrial exhibition at individual medical congresses, on the assumption that they would be influenced by visiting the exhibitions to buy and advertise certain medical products or services.

After 20 years of audible and readable appeals, the issue is no longer that the patient is at the centre of health systems, but to what extent the status shift is actually approved and recognised as added value by the powerful stakeholders of the medical profession, the pharmaceutical industry, politicians and the authorities regulating the pharmaceutical market.

Beyond all these exhortations, when it comes to implementing patient participation, the laws are often broader and clearer than the inner attitude of traditional stakeholders towards “the patients.”

I am thinking of many contacts with pharmaceutical representatives who are beaming with joy with “We put the patient at the centre of attention” [note the passive use of language], as if this were already a valid programme. However, they are very evasive on the issue of transparency of and access to study results, thus stonewalling on any issue that would constitute the actual patient involvement.

Why is the uncertainty of health care professionals in the health care system so great when dealing with “the” patients? Because when we talk about patients, we often vaguely mean three target groups: 1. The individual patients plus their relatives, who define themselves through their own involvement and through the level of information about their illness; 2. The patients who have developed into patient representatives through self-education and in individual cases also through training and; 3. The external patient representatives who represent their interests purely professionally.

The insecurity of other actors in dealing with patients and patient representatives is particularly evident when there is an overlap between their involvement and representation of interests. At all levels of committee work in the health care system, it would have to be ensured that the patient representatives involved are trained in such a way that they can advise and participate in decision-making at ground level. However, contact with the patients must be maintained at all times so that reliable feedback can be provided on the real needs of individual patients.

Three examples will show how differently the term patient participation is used. Patient participation from the patient's perspective is always understood in a broader and more equal way than it is in the prevailing health care system.

In cancer care, for example, physicians understand patient participation as participation in the best possible diagnosis and treatment using state-of-the-art technology, such as that provided in multidisciplinary cancer centres. In addition, patients should also have a say in their own treatment decisions, as is stipulated by law. Professional patient representatives, however, would go a step further and, for example, seek the participation of patient representatives in the tumour boards, in which the available cancer diagnoses are discussed holistically from a wide range of medical disciplines. In fact, such patient participation for breast cancer patients in the tumour boards of 18 tumour centres in North Rhine-Westphalia has been tested in a three-year study since 2017, against the resistance of some representatives from the medical profession. In the same way, patient participation in medical conferences could be established in general; in fact, this has been successfully carried out several times in clinical practice for HIV, breast cancer and rheumatology.

Since 2004, patient representatives have been involved in the Federal Joint Committee (the highest body that decides which medical services German citizens should receive from insurers), and from which the term patient participation from the patient perspective is always understood in a broader and more equal way in the health care system. They are allowed to submit applications and have a say. Professional patient representatives, on the other hand, strive for a real right to vote on decisions in the Federal Joint Committee and in the state committees.

In medical research and drug development, pharmaceutical companies that carry out research on clinical trial drugs are increasingly seeking contact with patient organisations in order to integrate real patient experiences into their study design, as they hope to achieve better study results. Professional patient representatives demand exactly this, but in addition they also demand that patient participation in all aspects of drug development takes place, that patients are not only questioned, but also have a say in research questions.

It is encouraging that the chairman of the European Patients Forum (EPF), as one of the two patient representatives, was elected to the board of the European Medicines Agency (EMA) in Amsterdam for three years starting in June 2019. The nomination of a former breast cancer activist as the new Commissioner of Health in the EU is another good example. THESE are the right signals for active patient participation.

**PATIENT
ADVOCATES** **IN**
NOT OUT



However, there are also setbacks on the stony path of co-determination by patient representatives. In June 2019, for example, the European Commission announced the top experts for the board of the mission “Cancer Research and Innovation” within the next EU research programme “Horizon Europe” (2021–2027). There was no patient representative among them. Why were we disappointed? Because in this case, patients are being researched and evaluated by third parties through us, without us being able to hear the voice of the patient. However, through various discussions with decision-makers and the input of influential patient representatives, it was finally possible to achieve a subsequent nomination of a patient representative from the melanoma sector. What an important breakthrough!

From the patient’s perspective, the phrase “putting the patient at the centre” does not mean initiating an ideological change from the conviction “the doctor knows best” to “the patient knows best”, but rather the aim is to achieve cooperation between the individual actors in the health care system in order to benefit from the different perspectives. In this way, demonstrably better and more patient-oriented health outcomes are achieved. In order for this to work, mutual recognition and appreciation of the different perspectives and competencies is a prerequisite.



19. CLOSED SOCIETY

When I did my first desperate internet searches in 2007 to find information about my rare “Myelodysplastic Syndromes” and people who were equally affected, Facebook was only three years old. The small leukaemia forum, which I came across by chance and which I joined immediately, had its provider in Luxembourg. The administrator paid for the provision of the service. As a special feature, the forum already had the opportunity for group chats, which we as administrators used once a month to discuss issues in the group. There were only a few new members, and our selection process included a phone call that decided whether to accept a new member. It was a small forum with about 50 members in its heyday. During the four years of my membership, between three and six leukaemia or MDS sufferers died each year due to transplantation or illness, and some members who were thought to be cured left us. With the Facebook experience of today, our mistake was not to constantly seek new members. We recognized this, but the opportunities to digitally network with potential members and attract their attention were still limited at that time. With Facebook, that works by itself. Anyone who enters the three letters MDS will find us now. Our Luxembourg forum, on the other hand, bled out over time — what a phrase in this context! We closed the forum in 2012 for lack of membership growth.

It wasn’t until the beginning of 2017 that I got back into the business and since then, with the help of two other administrators, I have been managing a closed Facebook group for MDS sufferers and their families.

Compared to its beginnings in 2004, statistics for the 2nd quarter of 2018 showed 30 million Facebook users in Germany. I have reservations about the global Facebook group because of the user benefits. I am well aware that Facebook is repeatedly criticized by European data protection and security experts for its poor data protection practices. I also know that we are only guests on Facebook. If Marc Zuckerberg decided to close Facebook, our whole community would disappear.

There are now similar restricted groups for many diseases. This is an ingenious communication tool with a self-increasing propagating effect. Self-promotion is not necessary. The functions that the closed group concept offers are tailored to the needs of a protected exchange between the group members.

In a sense, closed groups have a doorman, an administrator, who checks whether or not potential members are accepted into the group by looking at their profiles. An important rule in our group is that members who are admitted must introduce themselves with their MDS history in a timely manner. If this is not done, they will be dropped from the list. Meaningless combinations of letters as usernames will also be rejected. These rules are intended to establish a minimum level of commitment in a group dealing with serious, sometimes fatal illnesses and serious discussion of very sensitive content. There are “conversation situations”, where a person affected or a relative confides in the other group members with his or her innermost fears about their illness and is left vulnerable to the reactions expressed. Therefore, it is very important to take in members in a hand-picked way. It is clear that rules can always be circumvented by individuals. Nevertheless, I rely on the sense of responsibility. So far, we have been able to keep trolls away. Posts that are written in closed groups are not visible to non-members, so you are actually in a closed room in a virtual sense.

Myelodysplastic Syndromes are a rare form of cancer . The disease rate in Germany is on average 4-5 cases/100,000 inhabitants/year. Local or regional patient groups for leukaemia and lymphoma therefore have little or no MDS patients. In this respect, our digital Facebook group is so far the only platform in Germany that addresses affected people and their relatives nationwide without having to travel somewhere to meet. Because many of those affected are not well, or they do not travel long distances due to their advanced age or illness. Many commute constantly between doctors and hospitals anyway.

This results in several larger groups who are now based on the platform, which in summer 2020 has 325 members and is still growing.

First there is a large circle of relatives whose fathers, mothers or even grandparents are ill. We would never learn so much about the disease itself and its course in old age if not for the concerns and need for information expressed by relatives, because people over 72 years of age are unlikely to be online themselves. The stories that their relatives tell about them are often similar. Excluded from transplantation for age reasons, older high-risk patients in Germany only have access to an approved drug to slow down the course of the disease. All discussions and hopes on the net refer to this one substance, to the manifold side effects, to the actual effectiveness in individual cases, as well as to the duration of the response rate. At the end of this is a great silence.

The second largest grouping is younger sufferers, whereby younger means everyone aged 20 and over, with a clear majority between 50 and 70. Apart from very few low-risk patients who have not yet undergone transplantation, these are those who are eligible for transplantation, those in the process of transplantation or those who have already been transplanted with their aftercare concerns.

Finally, there is a smaller group whose children have MDS from infancy to adulthood. Again, most of them are allogeneically transplanted. The children, with their different degrees of disease, can hardly be summarized as a separate group. As a rule, those whose MDS has turned into acute leukaemia also stay with us. Finally, we have some patients with a cancer history in whom MDS has developed secondarily due to previous aggressive cancer therapies. Special forms of MDS and overlap syndromes with other blood cancers also occur.

In the group discussions it becomes clear that the majority of patients do not yet have access to the new world of the molecular genetic map. It is hardly mentioned in the diagnostic descriptions of the group members.

With a relatively small number of 325 members, this group surprisingly provides a “statistically” fairly accurate reflection of what is known about MDS in general, its courses and treatment options. This also applies to the topic of stem cell transplantation. There are a number of “marching-through” people who go home after four weeks. There are those who are heavily burdened by side effects and those who do not make it. One of the most anxious topics is recurrence, the possible relapse after a stem cell transplantation, the risk case, which in the case of MDS decreases in years after the transplantation, but is still quite high and remains so. Throughout the entire follow-up phase, we read the sentence “Hopefully my blood levels are okay.” Since blood values are known to fluctuate, the fear is often greater than the real danger, but it is dominant and does not let go. It multiplies itself before routine checks. There have been too many cases where a relapse has been mercilessly announced. We have also accompanied second transplants on several occasions and have sometimes had to endure the hopelessness after the failure of a second transplant.

Not only are fears and worries shared, but just as often exuberant joy about better laboratory results or successful treatments. Who, if not the fellow patients in the MDS group, could immediately grasp what is pleasing about certain laboratory values, in circumstances that require long explanations to real relatives and friends because they are not very familiar with the disease; and, more precisely, because they are not familiar with the feeling of having the disease. And when 50 comments in response express their shared joy about positive lab results, then, yes, the poster feels valued and understood in his group. He or she feels like a winner, and this sense of triumph can lessen the real threat of the complicated disease for a while.

What motivates patients and/or relatives to join a group with complete strangers? Since Myelodysplastic Syndromes is a rare disease, most new members are initially relieved and grateful when they meet people with similar problems and concerns, many of them for the first time. They gain more self-confidence in dealing with their own diagnosis and the course of their disease. They also gain an insight into possible treatment options, including the reassuring realization that they do not have to end their lives as quickly as they might have thought before. Newly diagnosed patients usually appear very anxious. The function of “older” group members can be seen in reassuring remarks such as: “In time, you will grow into this condition and be better able to tolerate it.” It is also important that patients or their relatives can get rid of conflicts that affect the way they deal with each other. For this reason, we recommend that either only the patient or the family member registers. Otherwise, there is a risk of limiting each other’s expressions of opinion and feelings. Other recurring themes in the group are problems with health bureaucracy and dissatisfaction with medical treatment, especially among registered doctors. We always recommend MDS patients to visit an MDS centre in a university hospital for a second opinion in order to exhaust all treatment options, including participation in clinical studies. There are enough experienced people in the group who recommend this to the new members; we administrators no longer have to do this ourselves. This means that this advice loses its instructive character and becomes much more effective.

Since the group is now large and also heterogeneous enough to cover all disease profiles, treatment stages and age groups, administrators hardly need to intervene with input. Together, a pool of knowledge is created, which covers most of the topics in the MDS environment. Knowledge and experience are increased and passed on together.

In addition to giving courage, showing solidarity and feeling that you are being taken care of, maintaining relationships with each other is of great importance. Mutual postings often lead to real relationships by telephone, followed by private meetings in the region or at patient days. A large proportion of the group members look into the group every day to see what's new.

The Facebook Messenger, on which private messages can be sent to each other as a couple or as a small group, is an additional tool that is used to "discuss" difficult, very personal or even conflictual topics that are too private even for a closed group.

Some group members confide in us that they cannot stand it when writing about dying. They then withdraw for a while. I can understand this well from my experience with my first forum. But if you exclude dying from a group dealing with an incurable disease, then you cannot offer such a group. That would be dishonest. You have to be very careful that descriptions in this context don't get out of hand, don't prolong themselves unnecessarily into inappropriate drama. Voyeurism is not allowed. This includes the administrators being careful in their choice of words and, if necessary, deactivating the comment function or deleting posts that do not comply with the group rules.

Some members leave the group again because they do not want to or cannot cope with the hard topic and the difficult fates. In particular, patients are most likely to leave the group if they have undergone a complication-free stem cell transplantation and can afford to keep their distance from the topic of cancer. Sometimes they read along for quite some time and then leave at some point. A new phase of life without the stresses and strains they have experienced should begin. This is more than okay.

Of course, there is no medical evaluation or even diagnosis of symptoms in such a group. In case of purely medical questions, the person is sent to the doctor for clarification. Experiential knowledge is offered and/or a serious information link is given. Sometimes we administrators have to intervene quickly so that a small thoughtless remark in a post does not turn into a major conflict. Social media is notoriously susceptible to shitstorms that seem to emerge from nowhere. Engaging in unobjective discussions will not help. Most posters are not willing to change their mind in an online discussion. So far, we have not experienced any serious problems and minor incidents could be easily dealt with by private messages on the messenger. Troublemakers have left the group by themselves.

I didn't want to interrupt or "disturb" the writing dialogues and the form of the exchange with factual information on the topic of MDS. On the other hand, I always had the ambition to provide not only an exchange platform, but also an information platform. Thus, the closed group is linked to a publicly accessible information platform to which I upload—with a short, summarizing comment—scientific articles, research results and blogs about MDS in the narrower sense and about blood cancer in the broader sense. Without claiming completeness, there is no standard for this. From experience, however, research and networking lead me to high-calibre sites that focus on this topic.

The number of subscribers to our info page exceeds the number of members in the closed group and from the comments I see that we have “external visitors.” My wish and that of many users would be to offer the mostly English texts in German translation. So far, my time is unfortunately insufficient. Common translation systems are hardly suitable. Most users understand the German medical terminology as poorly as English. There would be a great need for interested patients for a well understandable, but nevertheless high-quality “translation” of research results in German.

It requires an hour a day, sometimes more, to provide serious support to the group. After all, one has to scan every post. In order to have a lively and trust-based exchange, users must always feel that things are being taken care of. To achieve this, the administrators must show empathy in a careful but clear choice of words and appreciate questions at every level. As a self-involved person, I see many MDS outcomes and witness many stories.



20. HEAD IN THE SAND OR THE RIGHT NOT TO KNOW

In haematological practices, university clinics, study outpatient clinics and various therapy rooms, I constantly meet patients (and relatives) who have no idea what disease they have, what it does to them and who do not know which therapy or study drug they are actually receiving. I am irritated and disbelieving every time. What do you talk about with your doctors? What do the doctors tell them?

In the case of a study drug, they have been informed theoretically by a mandatory special interview and sign a multi-page Declaration of Consent! They are a large number of patients, but they seem to hardly notice or quickly forget about it. There are all kinds of carelessness in the health care system, but I cannot imagine that in specially set up study outpatient clinics the legally required clarification talks often do not take place or the patients are not given the necessary guidance. Why don't the contents of the conversation and the text reach the patients and why don't they memorize them? When I ask patients about it, they answer: "Yes, I have signed something. But I can't remember exactly what it was."

Overload.

We patients inevitably get into conversation in outpatient day clinics or practices. Where else do you lie so close to each other, as strangers, next to each other on beds or recliners, condemned to do nothing, listen to music, read or—if you are not feeling well—sleep? Most of the time, however, one waits for the therapy to begin or for it to be completed.

So, at some point I ask my bedfellows the curious and heretical question, "What is your illness?" And after a short break from listening and small talk, I immediately launch the next one, when I come across an unknowing person again: "Why don't you know? Most of them are willing to talk, even though I may be offending them. Their lives may be at stake. So, it would be good if they knew what illness they have and what therapy they're getting for it!

Accompanied by a shrug of the shoulders or a clear expression of a guilty conscience, I usually get the following answers: "You have to ask my wife / husband, she/he knows better." "I don't know, you're right, I should know "these things" and be more interested." "The doctors already know what's right for me." "I'd rather not know." "I don't understand any of this anyway."

If I may say anything else, I then ask: “All your life up to this point you have taken responsibility for your family, for your job, for your car... have done what you thought was best to keep everything as long as possible. Now the time has come when you have to take responsibility for your body. The doctors help you and you have influence on what is done with you. Understanding and accepting this is your new task since the diagnosis and at the same time your best chance to get well again. Ask and try to understand more and more about your illness over time. Don’t let the decisions get out of your hands.”

Sometimes I recommend patient-oriented brochures or contact with a patient organisation, but not always. I push enough. I don’t want to give the impression that I’m just trying to recruit club members.

No one has ever told me to leave him/her alone. Maybe I wake up one patient or another a little bit; if it is enough to change their attitude, I do not know. Probably rarely.

Recently I experienced a completely absurd situation on the couch between two men in the haematologist’s practice: Both men, about mid-60’s, knew neither the name of their disease nor that of their therapy, let alone how it works. It got to the point where I tried to guess what they had based on the symptoms and the colour of their transfusion bags and by mentioning the names of drugs. Both had been ill for several months. The result was a lively conversation about how to slip into it: “You may have this or that,” such a conversation is simply absurd. On the other hand, both were suddenly highly interested, as if we were playing a game. Meanwhile the assistant doctor came with the therapy bags for one of the two gentlemen. Then I heard both of them ask the haematologist in the choir: “Doctor, what’s wrong with me?” I held my breath. One had mantle cell lymphoma—I hadn’t guessed—the other had myelodysplastic syndrome—I was right about that. I have it too.

Head in the sand—the patient’s right not to know.

This can be threatening, especially in disease areas where more research is being done than treatment, i.e. where there is still a great deal of uncertainty about treatment and where patients are unwilling to test new drugs in studies.

I am convinced that most doctors are not aware of the ignorance of their patients. Discussions with doctors about this immediately lead to indignant self-defence. “I know perfectly well, but I always talk and inform my patients.” Yes, and I think so, I do believe them. Nevertheless, I wonder whether it would not be useful to check several times with the patient what he has understood, what he wants to know and what he should know. This is a process and it does not end with a signature under a form confirming that you have been informed.

I often think about whether I have the right to force fellow patients into their reality. In fact, I do not know exactly what the right way to deal with a life-threatening disease is. This is individually very different. However, my personal motto is: “mature patients survive longer,” and sometimes I go on a mission with this sentence.



21. CARPE DIEM

What a restless night with ringing ears, headaches and aching limbs! But tomorrow morning the taxi will come and by noon I will have received new donor blood. Then I will feel much better. But one worry remains: Can you trust your symptoms? Is the haemoglobin (HB) level really low enough to justify a blood transfusion in the eyes of your doctors? Because if not, you may be sent home and continue to suffer.

I have been a transfusion addict since 2008. In the beginning, the interval between two transfusions was six weeks, soon after only four weeks and after a serious infection in 2010 the interval shrank to two weeks. Since then I have been fighting every day for a longer time without transfusions without suffering too much. Currently, my transfusion frequency fluctuates between 9-14 days. This is quite often and considerably impairs my mobility, as I am always in an outpatient clinic for a whole day at this rhythm.

It is not advisable to be away from home for more than a week and it sometimes happens that I have to request a so-called “advance transfusion” even if my haemoglobin levels are tending to be higher. This is in order to manage the timing with weekends and my commitments on other days so that I do not suddenly run out of strength on a strenuous return journey, for example. This happened once on a planned return flight from Edinburgh, when I was unable to fly for three days due to a volcanic eruption in Iceland and an additional storm on site. Instead I was taken by ambulance from the airport to the clinic, where I learned about the free basic care of the English health system and received my blood transfusion there. Excitement and effort could have been avoided if they had filled me up “earlier” in Germany and not calculated so narrowly with the transfusion dose.

The routine procedure when I need blood is to have my blood drawn, wait for the lab results, order the transfusion from the blood bank, test and prepare the two bags of red cell concentrates from two suitable donors, administer the blood and finally be on site after the transfusion to rule out any allergic reaction. This can take up to eight hours in total. It’s like a working day, at the end of which I usually do nothing else but rest and give the donor blood the opportunity to mix with my remaining blood.

From the patient perspective, I have become a true expert on blood transfusions. I know all the procedures, all the regulations and all the contingencies involved at a local clinic. I can estimate my own haemoglobin level fairly well and know about all the daily growing physical and psychological stress factors that patients like me suffer when they urgently need a transfusion. I am a junkie. I'm addicted to fresh red blood. It has become part of my identity.

Internet forums are used to discuss the effects of blood transfusions based on personal experience. They deal with the importance of the haemoglobin value for the transfusion decision, the quality of one's own vein or port system, the point at which one actually feels better after a transfusion, and the hope that no major accidents will occur, for which blood is needed in the emergency room and operating theatres, because this considerably extends the waiting time for one's own supply.

The understandable principle is to transfuse as sparingly as possible.

Firstly, experiments with rats as well as the results of clinical studies suggest that recipients of frequent blood transfusions are suspiciously prone to certain types of cancer. Yes, there are reasons not to use transfusion therapy so generously. There can be dangerous allergic reactions and infections in the statistically minimal range. The development of antibodies during regular blood transfusions is likely, which limits the circle of donors. Iron overload is a major long-term problem. It is only partially possible to remove the iron attached to the haemoglobin, which damages the body tissue, by means of medication. In addition, each hospital has its own transfusion policy, which considers even a lower HB value to be sufficient or transfuses more generously.

Secondly, the patient's symptoms should show that he or she is severely anaemic—"the paler the better." Symptoms such as breathlessness on exertion, ringing in the ears, tachycardia, etc. are queried and documented. Unfortunately, inexperienced assistants often hardly look at the patient's face and do not pay attention to the mucous membranes in the eyes and mouth or the colour of the fingernails. Experienced nurses, on the other hand, immediately see my dilemma in the pallor of my lips and face. I never feel better in my neediness than when a receptionist at the counter of the day clinic compassionately looks at me with the words: "I'll go and get the order forms for the blood bank ready."

Thirdly, it is always up to the treating physician himself to decide how strongly he weighs the other two conditions. He has considerable discretion to decide for or against a transfusion. He makes the decision—especially if he is not so familiar with the individual case—strictly on the basis of laboratory values rather than the patient's condition, even though a documentation booklet is available which shows the course of the blood values of the last few months and, above all, the transfusion rhythm.

It is often only a few tenths that would prevent a transfusion in such a case, i.e. the difference between HB-value 8.0 and 8.4 g/dl for example. It is also important to know that the HB-value can be subject to daily fluctuations and that the mechanically collected laboratory values can vary from clinic to clinic by up to 0.5 g/dl. These values are actually only a guiding indicator for the clinical condition of a patient. Therefore, the symptoms of the patient should be the main focus. There is no rule that "one" only needs blood from an HB value of 8.0 g/dl. Some patients can tolerate even lower values, others feel weak, tired and without energy at much higher values. Concomitant diseases can also play a role in the weakness status.

My view, backed up by many years of experience, is that the need for regular blood transfusions must be based on improving the patient's quality of life and not on the long-term consequences or complications that may arise. With all due prudence and respect for regulations and guidelines, a personalized "Carpe-Diem policy" must take precedence!

The lower the HB-value at transfusion, the lower the increase through transfusion, because two bags of concentrated red cells generally "bring" only a maximum of 1–2 points in the unit of measurement g/dl. So the lower the value drops, the faster you fall below a critical value again, from which you no longer feel well. This can make the days immediately before the next transfusion a torture and every staircase a difficult obstacle to overcome.

Once the HB level is below a critical level, it seems to fall faster and may fall already on the next day by 3-4 tenths, for which you did not fit into the guideline the day before. So you would definitely have to go to the clinic a second time in the same week. Spending two days a week in the clinic is a considerable restriction of freedom for patients who—if well transfused—manage to organise a still largely normal everyday life and feel reasonably comfortable and efficient.

In addition to the laboratory values, transfusions should therefore always be based on symptoms and not strictly on numbers. The most important question remains always the threshold value below which a blood transfusion is actually administered. In my experience, at this point the patients' perspective and the doctors' perspective can differ considerably. Doctors must look at their patients closely for the decision and listen to them carefully. They must take into account the experience of patients who know their bodies well after a long transfusion history. A good quality of life is indispensable for the physical and psychological management of a severe chronic blood cancer. Symptoms of anaemia are treatable!

To transfuse or not truly needs to be a shared decision.



22. I'M GONNA LAY DOWN MY BURDEN

There is a gospel song with the refrain “I’m gonna lay down my burden on the rivers of Babylon.” A beautiful picture, to be allowed to lay down a heavy burden that you don’t want and can’t carry anymore.

With iron overload this is not possible. Our organism has no excretory route to discharge excess iron. Instead, the extra iron is accumulated in the liver, joints, pancreas, bone marrow and, in the worst case, in the heart, causing long-term organ and tissue damage. Here we are talking about iron overload due to chronic blood transfusions.

The main cause of iron overload is transfusion therapy. Each dose of erythrocytes contains 200–250 mg iron. After only 20 units, this is up to 5000 mg which is introduced into the body. At this point the so-called ferritin value is at about 1,000 µg/l, which is about 10 times the normal value. This means that the attending physician recommends iron chelation therapy (therapy for iron elimination) for transfusion-dependent patients like me.

I had no particularly detailed knowledge about iron overload. I had long since accepted the most important fact: Excess iron damages my vital organs, so the iron must be removed. Everything went according to plan. I had no complaints whatsoever, trusted the medication and no longer attached such great importance to the subject.

I tolerated the iron chelator since 2009 and without any side effects. Only at the beginning I suffered from an unpleasant rash, which disappeared after two weeks and never returned. My ferritin value reliably decreased and is still below 1000 µg/l to this day. I trusted the ferritin value and my haematologists were satisfied with 400–600 µg/l! Adherence to therapy was my top priority, I never forgot to stir my suspension or take my tablets, even when I was on the road. More than once I therefore disappeared into the washrooms of an airport in the evening. I definitely felt safe from life-threatening iron overload, just like the package insert said. I was free of side effects, while in MDS web forums and in my work with other patients I met quite a few people who suffered from all kinds of side effects, mainly in the gastroenteric area.

There were discussions in the patient community mainly because of the efficacy, the development of the ferritin value and because of the incompatibilities about the right time

to take the tablets, about the appropriate dosage or also about the reasons for therapy breaks. The discussion about the substance Deferasirox has calmed down somewhat since the switch from suspension to tablet form in 2016. The tablets are better tolerated because they no longer contain lactose, which apparently contributed to stomach and intestinal problems in many patients. In addition, according to the statistics, the tablets should be more effective. This means less medication for more iron removal.

Even with minor side effects, patients unfortunately stop taking the tablets carelessly. This is an illusion. Iron overload does not hurt until it hurts, because an organ is permanently damaged. It can take years before it gets there. Some patients are not aware of the connection between disciplined adherence to therapy and effectiveness. Some even stop taking the tablets because it takes too long for them for the ferritin level to drop. When the doctor asks his patients whether they regularly take their iron chelator, they confirm this. But in many cases, this is not true. Pharma surveys in several countries support this.

Although everything is explained in great detail in the package leaflet, patients often do not read or understand it. A package leaflet cannot be simplified or shortened significantly due to legal regulations. When on holiday, when other illnesses occur and especially when the daily routine is irregular, the tablets are often not taken. However, the pharmaceutical company is dependent on the patients' adherence to therapy. After all, proof of efficacy is what determines success on the drug market.

As a patient representative, I participated in national and international advisory committees of the pharmaceutical company, where we discussed additional brochures with patient-oriented texts, graphics and diagrams in order to better communicate the mode of action and form of metabolism of Deferasirox to patients. Our aim was to draw attention to the dangers of iron overload. In many countries in which Deferasirox is sold, the pharmaceutical company regularly organises training days on the drug, always with the message that only regular intake can guarantee the success of iron deficiency.

I had long since become chronically dependent on blood transfusions and between 2008 and 2020 I received more than 650 bags of red blood, in a relatively stable interval of 10-14 days since 2010. My equanimity ended abruptly in autumn 2017, when my treating professor sent me to Hamburg for a special MRI. There all organs were to be checked for iron overload. My heart, liver, pancreas and bone marrow were scanned. The results were devastating. My confidence in the iron drainage by Deferasirox shattered at the moment when the radiologist asked me incredulously why I didn't actually have diabetes with such an iron overloaded pancreas?

I have still not recovered from the results of the MRI: liver and pancreas are five times more iron-loaded, and the bone marrow is also full of iron. My heart is still free, which is no reason for long-lasting joy, because iron overload of the pancreas is an early and reliable marker for later iron overload of the heart.

It is not an option for me to increase the dosage of the iron chelator to possibly drain more iron—increasing the dosage would have a toxic effect on the kidneys, among other things—nor can I reduce my need for transfusions—this would destroy my quality of life, drive me even further into anaemia and also damage the organs in the long term. As so often in the course of my illness, as an MDS patient I have to accept with frustration that there is a diagnosis but no therapeutic option.

In my already quite extensive medical report, I now read “severe secondary haemosiderosis” (= acquired iron storage disease) in addition to all other signs of the disease. My liver values are completely fine. This means that the iron overload measured in the MRI does not (yet) show any damaging effect. It also shows that the internal organs seem to be able to cope with massive iron overload for a long time before they show their overload in poor values. Above all, freely circulating iron is toxic for the organism. Therefore, the iron stored in the cells is coated by a protein complex, ferritin. If the ferritin value rises above the norm, it is called iron overload.

Intensive research shows that the predictive value of ferritin is often not very meaningful. Two patients with the same iron overload can obviously have completely different ferritin levels. Many factors play a role in individual iron metabolism. It is known that the ferritin value fluctuates and also increases, for example, in the case of infections.

However, as the ferritin value generally correlates with both the level of the body’s own iron reserves and clinical results, it is usually used as a parameter to monitor the course of iron excretion. This is less expensive than an MRI.

The self-regulation (homeostasis) of body iron is subject to a complicated system of iron uptake, transport and cellular utilization. I am aware that this metabolic process is completely disrupted in the case of severe iron overload. It is also clear to me that this makes me very ill over time.

Nevertheless, I must stress that the positive effects of iron chelation therapy in chronically transfused patients have been proven by retrospective studies. There is no doubt that Deferasirox therapy is associated with higher overall survival. The transplantation results are also better after chelation therapy than without. Without Deferasirox, I might not be alive after years of foreign blood transfusions.

However, I doubt the further effect of the drug promised in the package leaflet. Obviously, the drug captures the free iron that can form the harmful oxygen radicals and effectively drains it through the stool. What the drug does not do, however, is to reverse the iron overload of the important organs such as the liver or prevent further iron accumulation.

Iron-overloaded patients urgently need new, innovative and effective drugs that are administered in parallel to transfusion therapy and at a relatively early stage. Waiting until after stem cell transplantation, when blood formation can be restored and blood cleansing is possible to reduce the iron overload, is not a good option. Often the blood formation after an allogenic stem cell transplantation is not (yet) sufficiently stable for phlebotomies.

Organ damage may already be present or may be exacerbated by an even greater need for transfusion during the aplasia phase—the phase immediately following transplantation, when the body has no immune system.

Iron overload remains a problem that must be solved medically even after stem cell transplantation.



23. IT IS WORTH A TRY

When you are in a drug trial, you want more than anything else for the drug to work on you. Family and friends who are informed tend to look on the bright side and keep their fingers crossed because they wish you the best, without having understood exactly what it is all about. Together you float on a pink cloud of hope. One should be careful with this hope, because anything between miracles and failure is possible at any time. The simple and exciting question is: Will this new drug work for me and if so, how long and with how few side effects?

Hope can be a deceptive currency in an incurable haematological disease. It sometimes obscures the view of the actual blood values. Figures or trends are the only thing the haematological investigator can rely on. To make positive predictions, he needs trends, i.e. values that increase over a longer period of time.

Rising trends, reliable statistics from earlier study phases and good clinical values entitle the haematologist to say “I think we can have hope that the drug will work for you.” There is real joy written all over the doctors’ faces and also a bit of incredulity as I drive home on my pink cloud. I well remember the warm feeling of gratitude that flowed through me on the long train ride: “There is a drug that will buy me time, maybe even a few years.”

I had to stop two other trials with other substances before due to serious side effects. With the first trial drug, I ended up in hospital after weeks of severe abdominal pain with acute kidney failure, and with the second drug, the white blood cell count dropped so low that the trial protocol required immediate discontinuation.

The now third study drug gave me no side effects whatsoever, only a long train ride to F. for a check-up every three weeks and to have my three injections administered there. I did this for almost four years, much longer than any effect on my transfusion frequency could be proven. But my doctors and I couldn’t say whether my blood values wouldn’t drop massively after I quit the study. The argument was that the drug could still have a stabilizing effect on blood formation. So we kept delaying the withdrawal. Until we finally tried it, with the result that my blood values remained stable despite the discontinuation of the study. I was lucky, because my return to the study would have been blocked for formal reasons.

The vertical distance between those who design and conduct studies and those who go through them is huge. According to my observation, very few patients ask about the scientific and clinical superstructure. They don't ask themselves at what stages of a drug trial they are involved in as patients and to what extent this might even make sense or even be logical. Very few are aware that they are not only objects but also subjects in medical research and that this alone gives them a moral right to have a say in the development of therapies. In this situation, most of the study patients that I met in the haematological field did not have the self-confidence and certainly not the willingness to understand a change of perspective from pure recipient to actor in the study development process. The "end users" or test persons of a drug talk among themselves primarily about the side effects of the study drug and whether, when and how long a substance succeeds in pushing back the disease. This is often a matter of life or death. However, I think it is likely that the younger generation of (blood) cancer patients under the age of 50 will be more critical and self-confident about their own role and its significance, not least because of the information available on the Internet, good work by patient organisations and new healthcare legislation.

Patients take part in a study because the doctor they trust has convinced them that an experimental drug gives them the chance to improve their symptoms in the long term despite the risk and hopefully tolerable side effects. The fewer alternative treatments are available, the greater the insight.

The bloated regulations, assurances, explanations and rules concerning the conduct of a study that are available to patients in writing are disconcerting. The language of the informed consent forms and the consent form is still complicated, although the physician and study nurse will answer any questions. Excessive demands due to a lack of transparency and thus disinterest is a bad combination to arouse the patient's awareness: "This is actually all about me all the time." Therefore, the contents are quickly forgotten. At best, they are interested in: "Can I continue to receive the drug if it helps me, but the study is terminated?", a question which in practice—despite ethical necessity—unfortunately cannot always be answered with "yes" without reservation.

It's like squaring a circle to assume that those existentially affected by a disease can stand up for themselves and the patient community in important committees with commitment, competence and the necessary distance, and exert co-determination in research and therapy development. I strongly support the idea of the informed and responsible patient, but the issue of patient participation in drug research, development and approval must be advanced by delegated patient representatives from patient organisations. These patient representatives must be sufficiently well trained and have communication skills that make them recognised players at the table with doctors, pharmaceutical representatives, regulators, evaluation institutions and policy-makers. A patient academy run by patient representatives and dedicated to these concerns deserves respect, recognition and encouragement from all stakeholders in the health care system.

What patient involvement has already been able to change is shown, for example, by the discussion about the so-called primary endpoint of a study, which is the primary goal of a trial, by which the additional benefit of the drug compared to other drugs is measured at the end. For a long time, the undisputed focus was usually on the overall survival rate of patients. By including the patient perspective, however, quality of life is increasingly becoming the indicator for the positive effect of a drug. At the end of their lives, cancer

patients are not interested in living three months longer with strong therapy side effects, but rather in the fact that the drug not only has a medical effect, but also benefits in everyday life, which has a positive effect on their satisfaction. The method of querying the criteria of health-related quality of life by means of patient questionnaires and evaluating them on the basis of evidence has now been refined. This makes it possible to counter the accusation that quality of life is a completely subjective and therefore unreliable variable.

The experience of patients and patient organisations must be recognised even more strongly as expertise. Patients must have a say in all areas of clinical research. This concerns the priorities of what research is to be done in a particular disease, the study design, the study commissions, the calls for tenders for studies, the complete implementation and execution and finally the publication culture of the study results. The reason for this is obvious. It is not only a matter of an interest-based involvement, but above all the fact that the study quality, drug safety and efficacy are higher due to the direct participation of patients in the study-related decision-making processes. Because it is not the doctors who ask us how we are doing and then pass this on, but we ourselves answer how we are doing, what needs we have and what gaps in patient care need to be closed.

Patients who look for a suitable study for themselves on their own initiative are disappointed by the Internet research. A keyword-supported register in understandable language would be patient-friendly. In this way it would be possible to search for an experimental therapy option within Germany or within Europe, to introduce oneself at the appropriate place and to receive medical advice about the possibility of inclusion in a study. Such a register does not exist in Germany, in the USA it does. Searching for studies in individual university hospitals or networks within Germany is laborious. You have to know exactly what you are looking for. It is not transparent why certain studies are offered at certain university hospitals and not at other study centres. One is lucky if the treating physician draws one's attention to a study.

The first two studies for which I was recruited ran at the local clinic, but the third study I looked for myself and only found it because I now knew enough people in the field who I could ask about it. This cannot be the basis for a concept to recruit more patients for studies. No wonder that there is a general lack of patients willing to be recruited.

Effective new drugs are at the end of a chain that can cost many millions of Euros and that leads from visions of a cure, through years of research work, to successful studies that eventually lead to the approval of drugs. However, in contrast to their actual value and claim, studies do not have a good standing, neither with healthy nor with sick people. Reservations, fears and distrust are great and are still burdened by historical memories. More educational work and complete transparency of study results for all target groups would be necessary.



24. AM I HYSTERICAL?

To have a life-threatening disease well under control means to integrate it into everyday life in such a way that a minimum of quality of life is provided. Although the disease is present in our thoughts and feelings, it is manageable despite all theoretical and visible threats. One accepts small attacks from The Monster. The really big attack has not yet happened to me.

However, there are one or two days a year when the superficial peace in me starts to falter. These are the days on which I am being punctured. If there is any suspicion of progress in my condition, the punctures are more frequent. One has to look into my bone marrow to see if and how my blood disease progresses. My doctors are relentless. During the diagnosis, the laboratory counts the blasts (immature blood cells), documents the dysplastic (malformed) shapes of the blood cells and searches for newly discovered chromosomal malformations or new gene mutations. In addition, the development of the connective tissue of my bone marrow is examined, i.e. whether it is becoming increasingly dense through defibrination, for example. These would all be signs of disease, which would increasingly restrict effective blood formation.

In order to obtain bone marrow, a drill is used to penetrate the inside of the pelvic bone and suck it out. That's it! This hurts very much despite external anaesthesia, at least for me. I know from numerous conversations with fellow patients and from discussions in Internet forums that it is not only me. Nothing is more fitting than the phrase: "The pain runs through my marrow (sic!) and leg."

It is a sudden sharp, pulling pain that races through the hip and sometimes also through the leg and foot. Depending on how many tubes have to be filled with bone marrow or the so-called fragments, the pain comes several times. If it came unexpectedly, it might be easier to bear. The combination of anxious expectation, cramping and pain is intense. I am hardly up to this. My blood pressure is already at 180 mmHg and higher at least an hour before.

It is even worse when a so-called puncture is made. Then a piece of bone is additionally milled out of the bone marrow to assess its density, which allows conclusions to be drawn about how well or poorly the blood formation is functioning.

It is theoretically possible to receive sedatives such as Midazolam or Propofol, as is the case in other procedures for example during a colonoscopy. However, sometimes I find that doctors do not think much of “sleeping through the procedure.” The reasons given for this are manifold: “too dangerous because of a possible circulatory collapse, too personnel-intensive, too costly” or it is simply said that sedation is not common at this or that clinic. I often give doctors the patient’s perspective in this area.

No patient-argument will dissuade an attending physician from their point of view. Sometimes there is a story that a doctor has already had himself punctured for study purposes and that this is not so bad. By God yes, the man is healthy! I, on the other hand, take my entire physical and mental illness package with me for a puncture. And when the senior physician leaves the room, more than one nurse has confided in me that she would never let herself be punctured without anaesthesia. Some patients would cope with it to some extent, but others would scream in pain.

Many men of the older generation pull themselves together during a bone marrow puncture and comment on this later with sayings such as: “If it doesn’t kill me, it makes me stronger” or “I have already had to endure so much pain in my life, it doesn’t matter anymore.”

When it starts, I lie on the table half-naked and listen to the doctor and nurse talking while preparing the operation and sterilizing the instruments, I am angry every time, because I feel like a victim. I feel abandoned. My dignity is violated. Pain is inflicted on me and it would not be necessary for me to endure it to this extent.

The worst sentence: “Now it’s about to get a little uncomfortable.” Nice paraphrase, especially if the puncher isn’t that skilled. It happens, unfortunately. Anyone who has already been bone-marrow punctured several times will rave about the scorers who had “good hands”, and swear at the “butchers.” So you have to wait until the external anaesthetic has worked optimally, the angle at which the drill sinks into the pelvic bone and the speed at which the bone marrow is aspirated (sucked out) etc. Good puncturists are experienced practitioners who have punctured frequently. I would never let a first-year resident touch my pelvis.

But everything can also be done differently. I know from clinics and especially from haematologists in private practice that patients can be sedated before a bone marrow puncture if they so desire, without much discussion. It can be observed that younger doctors are more willing to grant the patient’s wish than those of the “old school” who think that one can “grit one’s teeth.”

The sensation of pain is, without doubt, subjective. As a long-standing patient with an incurable blood cancer disease and massive physical and mental stress, do I have to be latently accused of being a “wimp”? Most likely I am hysterical, but I am not in a position to turn that off. I have already had too many painful experiences in this context. After a puncture it is very soon before another is performed. It is only weeks before I think about what is going to happen to me again.

I have been punctured about 25 times during my longstanding illness. Among them perhaps five or six times with the sleeping and sedative Midazolam. I really appreciate the difference.

I have already described my trauma compared to painful iliac crest punctures several times in blog articles, expressing my lack of understanding for the unwillingness to sedate the patient during the procedure, especially in university hospitals. This sedation is quite common e.g. during gastroscopy and colonoscopy in the established internists' practices and is comparable in intensity and duration.

The reason for this clinic policy seems to be the red pen, which has eliminated this service in outpatient clinic departments. The use of surveillance monitors and personnel, both mandatory for sedation, causes too many costs. However, instead, hospital physicians say that sedation is generally more dangerous for the patient than the pain of a puncture. This depends on the individual case.

During the first punctures at the beginning of my illness, the mercilessly dominating fear of pain was certainly greater than the pain itself at one time or another and the whole procedure was often over after 15 minutes. With the duration of the disease, however, my bone marrow becomes increasingly fibrous. This means that there is too little usable material in the syringe after the aspiration.

The peak of my suffering consisted of three punctures in a row, performed by three different doctors with a total duration of one hour. Between the individual punctures, a nurse was sent to the laboratory with the respective bone marrow fragments to ask whether the material would be sufficient for a finding. Each time she received the answer "Sorry, that's not enough." This is called a "punctio sicca," a dry puncture.

On the third attempt, the doctor did not suck in, but tore the material into the syringe with a strong jerk—at least that's how it felt. My cries of pain could be heard loudly and I almost crushed the hand of the nurse who had been assigned to me. I could tell from the looks of the people involved that things were going very badly and nobody could really look me in the eye during the short breaks. They knew what torture it meant for me and they did not feel good about it. "We'll try again and if it doesn't work out, we'll stop." And then?

But this time the lab was satisfied.

Still on the hospital bed, with tears in my eyes, I announced unequivocally that I would never again have a puncture done without sedation. Never again! This time there was no contradiction. Much earlier I would have had to be adamant about my demand. I could have spared myself great fears, pain and the feeling of diminished dignity if I had asserted myself.

At the next puncture half a year later—in consultation with me—great importance was attached to freedom from pain and I was already asleep before any hands were laid on me. The puncture itself had been just as difficult as the one before, but I only learned this from the stories of the people involved. Just like that!

I stoically endured the three plasters in the pelvic area and the pain "afterwards."

As a patient representative, I was able to express my views on how bone marrow punctures are generally felt by blood cancer patients in an interview in an issue of the journal *Oncology* 2016, which was accompanied by a survey of my own.

After a survey of more than 150 blood cancer patients, it became clear just how much psychological pressure is placed on the patient before and during a puncture. Will it hurt? Has the therapy worked? Am I still in remission or has my condition worsened? Was the stem cell transplantation successful or has the disease returned?

More than half of the respondents would prefer a short sleep (sedation) by infusion to local anaesthesia alone because of the pain they experienced. The majority of patients stated that they were afraid or very afraid of the pain caused by a bone marrow puncture each time and again more than half of the patients described the procedure as quite or very painful. This is especially true when punctures have to be performed repeatedly over the years.

In the meantime, there is a trend towards punctures only being necessary at the initial diagnosis and less often during the course of the disease. Nowadays, many changes in the blood can be detected in a simple blood smear using special procedures. This is still the theory, as many doctors continue to rely on the good old standard puncture. If one is included in a medical study, punctures are still required in a very narrow range. In order to assess the success of a stem cell transplantation, more rather than less punctures are also carried out for safety reasons.

I would have liked to have had the opportunity to speak about this topic at specialist congresses in order to reach as many doctors as possible with the patient perspective. I would have liked to have advocated that, in the case of bone marrow punctures, patients can generally be given sedation if they so desire, and that this right should also be incorporated into haematological guidelines. An article in a specialist journal is quickly forgotten, and the opportunity to get to a microphone was unfortunately not available for this.

A missed opportunity. At this point, one would have had to keep on “drumming” in order to sustainably enforce patient needs. But the fact was that patient interest was not sufficiently organized. It was therefore not possible to build up pressure in the necessary places and there were no financial means to conduct a campaign for general sedation for difficult punctures. This was quite a disappointment for my personal as well as for my advocate commitment.



25. TRANSPLANTATION

I'm an impatient person.

All my life I have been working on my often-destructive impatience. Waiting in general often makes me nervous and irritable. Of all people, fate has forced me to practice patient waiting in the context of my illness. Not only in waiting, but also in enduring the situation that the therapy decision made might be exactly the wrong one with regard to a further lifetime. If the successful handling of a life-threatening disease would only depend on one's own personality type, how much would I wish to be a pragmatic believer instead of a quick-tempered agnostic!

At the beginning of my blood cancer disease, my family and I calculated from appointment to appointment that my doctors would tell me if and when I needed a transplant. But every time I brought up the subject, it was "We'll wait and see. There is no immediate need for action. The risk of allogeneic stem cell transplantation far exceeds your current risk of disease." This refers to the risk of contracting and dying of acute myeloid leukaemia (AML). First, second and even third opinions at different centres in Germany were—fortunately—in agreement.

Not that the decision to undergo transplantation is completely out of the question. It is the phrase "not yet" that makes the uncertainty so heavy on the shoulders. As one MDS expert put it: "I could very well justify a stem cell transplantation in your case, but if I transplant you now, you could be dead in six weeks. But I am actually convinced that you will be alive for at least another year without a transplant." I hesitate to say that this conversation took place as far back as 2010, but I would like to point out that the experts did not dare to venture far ahead with a good prognosis back then.

To him, an almost-guaranteed year of survival compared to six uncertain weeks seemed to be a real gain, and rightly so. The fact that to date I have increased that prognosis more than eightfold was not in either of our wildest imaginations.

I'm sitting on a powder keg without a net or a double bottom. There are no more therapeutic options for me. I have tried everything that the scarcely existent market of MDS therapies has to offer me, plus three experimental drugs from different studies. There is



nothing but blood transfusions and waiting. On the other hand, there are low blood levels, iron overload, a chromosomal disorder, three blood cell mutations and a compromised immune system.

A drug for the high-risk stage is still available. This is an ambitious plan, because if the drug were to be used without need, there would be nothing left in case of emergency that would bring me into remission immediately before a transplant.

As far as the timing for a transplantation is concerned, I must rely on my MDS doctors. If my general condition remains good, the green light would be given immediately if the following three cases—individually or together—were to occur:

- Increase in blasts (= immature white blood cells). 1–2 % in the blood is normal. If more than 5% are present, the traffic light would be red.
- Increase in neutropenia (= deficiency of white blood cells, which regulate the majority of the immune system) or sudden drop in thrombocytes (blood platelets).
- Increase in cytogenetic disorders and mutations in the blood cells.

These would be clear signs of progression.

The prognosis systems in MDS are based on risk levels. With the above-mentioned contingencies, I would be in the highest risk level for leukaemia and imminent death. To prevent this, I would be transplanted at very short notice. I would literally have my back against the wall, there would be no alternative to stem cell transplantation (SCT).

Currently, and indeed since my diagnosis many years ago, I am classified with an intermediate risk, which means that everything remains open; the if and when of a transplantation is still subject to a “watch and wait strategy.”

Already twice during the course of my disease I came across a clear SCT recommendation. When my cell mutations could be made visible for the first time by new technical procedures, the indications looked bleak for a few days. With a targeted retrieval of my initial blood samples, which were lying dormant in a kind of evidence room in a laboratory of my university hospital, my attending professor succeeded in proving that exactly these mutations had already been present in the blood samples since my diagnosis 10 years earlier. So, these mutations did not seem to further promote my disease. The stem cell transplantation was therefore cancelled. I can't say whether it made it any easier for me at that time than if I had been glad to finally go on the ride through hell with the prospect of getting well again.

The second time I was advised by three university professors to have a transplant, I went through a period of fatigue that made me so depressed that I took every opportunity to lie down for exhaustion and tiredness during the day. I hardly benefited from blood transfusions. It finally turned out that my deplorable general condition was due to a total deficit of vitamin B12 and vitamin D. After replenishing my vitamin stores sufficiently, I reached my “old form” after a few weeks.

This time, due to my poor general condition, I had firmly reckoned with the transplantation and was already on my way to coming to terms with the fears and hopes that this step would unleash. I had informed my family and close friends of my decision and surrendered to a new era. Braking hard and turning back was hard for me. For the first time I had the impression that the doctors would have pulled through and transplanted me if I had insisted on it myself. The door to this is now much more open than at the time of my diagnosis, even though my MDS has been “stable” for years. The words of my transplant surgeon when I announced my decision against it because I felt much better again were: “You can’t escape us now anyway.” Now I am the one who is watching myself closely and continues to wait, knowing that the “right time” can be missed.

I must admit that I am greatly influenced by the survival statistics, prognosis scores and the many acquaintances I have made with patients before, during and after transplantation.

Due to the long duration of the disease, the iron overload and the many blood transfusions, a stem cell transplant is risky for me. There is a shaky 30-30-30 “drawer” for this: 30% of transplanted patients will recover—30% live with mild to severe limitations and 30% die. Although I have already beaten so many statistics, survival statistics of transplant patients still frighten me. After five years, about 40-50% of the transplanted people are still alive after a stem cell transplantation. The door is wide open to speculation. There are no guarantees in life, only chances!

It is not only the transplantation that has to be survived, but in the background, there is a risk of relapse that is progressive over time. High-risk patients are those with the highest risk of relapse according to studies. But the vast majority of patients are only transplanted when they are “high-risk.” Who can break this vicious circle?

In fact, the risk of a relapse is tragically high. Without wanting to stick to numbers too much, the risk is at least 40%, decreasing with the years after a transplant. This does not seem to be due to which donors are preferred, not to the type of chemotherapy and not to the drugs given before and after. Unfortunately, these instruments have not been able to achieve significantly better survival curves, according to significant studies.

It is the percentage of minimal residual disease (MRD) after stem cell transplantation that is decisive. This always occurs when the anti-leukaemic effect of the new stem cells, the so-called GvL (graft-versus-leukaemic effect), has not succeeded in flattening, eliminating, destroying the mutations that were already present before the therapy. This is a merciless revenge for the fact that there are no therapies available that can be used specifically against these mutations that are making people ill again.

In research into myelodysplastic syndromes, there has unfortunately been little progress in the therapeutic options, but considerable progress in prognosis. For example, the mutation burden before and after transplantation is increasingly establishing itself as a reliable prognostic marker for the probability of relapse (recurrence). There are “favourable and unfavourable” mutations, the latter more likely to suggest a relapse. This means that the sword of Damocles may be hanging over you before you have even had a transplant.

Stop!

Here is my best scenario: To live as an MDS patient for good years with a good quality of life BEFORE an allogeneic stem cell transplantation, then to catch the right moment for it, and AFTER that to get well again. Some patients actually succeed in this, the younger the better, the healthier the better, except for MDS.



26. CHRONICALLY ILL

Those who suffer from a chronic illness often do not show that they are ill, but often pretend to be healthy.

At the beginning I expressly rejected the term “chronic” for my case. Chronic illnesses are, for example, diabetes or rheumatism, which can be kept largely under control with medication, even if you have to take them for life. This is a status that is beyond all prospects in the case of myelodysplastic syndromes (MDS). To date, there is no therapy that is effective for longer than an undetermined, rather short time.

Moreover, the few drugs available are only ever effective for some MDS patients. The disease can quickly deteriorate dramatically, turning into leukaemia, so that a risky stem cell transplantation becomes necessary, but only if you are “still young and otherwise healthy enough.”

Otherwise, the only alternative is palliative therapy. In my case, “chronic” refers exclusively to the extremely slow course of the disease, for which my professors have been congratulating me for years after every status-enhancing bone marrow puncture.

The Internet is full of cancer stories. We are used to thinking in stories. Most stories have a beginning: the diagnosis, a turbulent middle part: the treatment, and a positively phrased “end”: the cure. Simply woven. Often the end of the story is inevitably “open”, but even then the focus is on the positive coming to terms with things that have happened worse. Readers love that. They are just stories.

But the average healthy person doesn’t really know what it means to be chronically ill in my sense and to wake up every morning with the knowledge that the condition will tend to get worse and a cure is in the stars.

A few years ago I broke my wrist. All my friends and acquaintances pointed to the cast and were full of compassion, concern and good wishes. Phone calls were initiated with the question about it. Visible, assessable, temporary, relatively harmless. In contrast, blood cancer is invisible, unpredictable, chronic, serious and therefore scary for the observer. As an outsider, one would rather not have anything to do with this.

Being chronically ill gives insights and experiences into the meaning of life and into all ephemerality. But the price for these valuable insights is high. Your entire life is taken out of your control and you fight ceaselessly to get at least a part of it back.

The vocabulary of serious illnesses contains a striking number of war metaphors, which are used by patients, doctors, but above all by the media to lend drama to the situation. “I am fighting cancer, the battle is hard, but I will defeat this enemy in the end. If I’m brave, I can do it.” In English-speaking countries, heroes are spoken of when patients have fought the battle and survived the cancer. Patients are called soldiers and warriors who do not allow defeat.

These metaphors can carry a burdening connotation for individual patients: “Did I lose the battle against cancer because I didn’t fight hard enough? Only those who fight hard enough will win.” These are fallacies which can put a patient under just as much pressure as the constant “positive thinking” which often only relieves the environment emotionally and not the patient themselves. Undisputedly, patients with stamina and an optimistic attitude towards life have a better prognosis, but there is no inherent link between “will to fight” and success.

The terminology of stress psychology knows three Fs for humans and animals to protect us in case of danger or threat: “Fight”, “Flight” or “Freeze”, means “fight”, “escape” or “play dead” (ignore, just do nothing). Which reaction promises the greatest success in the therapy against a life-threatening disease? Fighting as the unconditional will to live, yes, but please no warlike metaphors that cause further anxiety!

Over the years I had to revise my belief in the power of conventional medicine. At first I had the idea that the knowledge and experience of the doctors would bring me into a kind of safety zone where I would either be cured or die. I wanted to prepare myself for one or the other. However, I hadn’t realized for a long time that I would be hanging somewhere in between for years, and that this would be my greatest chance. In time I realized that there was no point in demanding reassuring words from my doctors about my condition.

In the meantime, I want the facts, and I’m getting them. And I have to be able to stand the fact that I’m not being spared anymore. The fact is that there are different degrees of difficulty for each case, including me. No doctor has the power to make the predictions that would comfort me.

There are a few ways that I can counter the sheer concept of struggle. Not that I can always manage to take them to heart, and what makes me feel relieved and upright does not have to apply to someone else.

At first, crying helps. You can also run into the woods or stand under a bridge and cry out loud. It is also okay to hammer with your bare hands on something, preferably something that is not dear to you. A punching bag, why not? Or chopping wood. Letting your anger out once in a while can help. But it is important to consciously take control again afterwards.

I mercilessly repress the illness if I cannot bear it at the moment. I simply block it out. I suppress negative feelings, I ignore my own prognoses and the medical histories of other MDS patients who have not progressed well. After some time I can let it all happen again, bit by bit, and that has to be the case. Dealing with reality is essential, but I know my limits of mental resilience and change approached when necessary.

I am always dealing with my illness, possible therapies, research results and the experiences of other patients. Information is the key. Contact with other MDS patients creates a connection that is important to me, that I nurture, personally, online and by phone.

I reject far-reaching future plans. To live in the present is not easy and can only be successful with discipline. I collect the memories of great loves, good conversations and overwhelming experiences of nature like pearls, ready to bring them out again in dark hours. I try to cultivate friendships and family relationships without using my illness as a pledge, without seeing myself as a victim to be taken care of because of my illness. I want to continue to be an equal friend and conversation partner who takes the ailments and small worries of others seriously. For this I sometimes have to hold myself back a lot. Painkillers are always in my handbag if needed.

Movement and distraction often help to bring about a positive change of mood. Often it is the quite stupid activities like weeding or ironing that get me back on track. Music can build solid bridges, while a spiritual framework can be a great calming and source of strength. Being able to eat and drink, to digest having no pain and a relatively restful sleep in addition to sufficient mobility define quality of life in the original sense. Only on the second level are activities such as travel, social life, pursuing hobbies, working, etc. In these areas, however, renunciations are constant companions.

As a chronically ill person, one must always plan realistically. You don't have to, and you can't visit all the sights of a city during a holiday that you can just about afford physically. If it is only the cathedral and a bench in the park, then it is still possible without any significant disappointment. It is the breaks that set the rhythm. The rest you have to let go!

You should have the living will and testament in the drawer.

More and more I realize that on average I feel more comfortable and freer when I am with people who know what illness and the blows of fate mean.



27. YOUNG. BEAUTIFUL. CANCER.

Young. Beautiful. Cancer: I recently saw this title of a photo collection somewhere and I haven't been able to get it out of my mind since then.

Three words in a row, two with positive connotations and then the shock. Many perspectives on the subject of cancer open up before me:

The dramatic fate of young people developing cancer before they have even lived. That young cancer patients can also be beautiful, as aesthetic photos of women with only one breast in black and white photo series have already shown. That many young women—to many men anyway—look attractive without hair.

I knew—coincidentally in parallel—two young women from a forum, both of them—only 25 and 27 years old—had a relapse relatively quickly after a stem cell transplant.

The leukemia was back and it was clear that they would not survive. I especially remember their unrestrained anger and sadness that they would not be allowed to experience anything of what a normal, healthy life is all about: family, work, travel and the fresh breeze on their skin. Both of them got married shortly before they died to at least still feel this experience.

But cancer as a fashion accessory? As a tolerably portrayed burden in youth? “I may have cancer, but at least I am young and beautiful. I will show the world how well I can handle it, how confident the disease has made me.”

Young. Beautiful. Cancer? A macabre way to make money with young, beautiful people who are treated as something special because they have cancer? You can only look at it with a certain fascination with morbidity. Really? Voyeurism of viewers and exhibitionism of models? Then cancer can't be that bad! In any case, the photographer has to be highly sensitive and gifted in order to depict the vulnerability of those photographed as well as the necessary distance to the models for the viewer.

Those who have survived are photographed. Whether they give courage to those who are still hanging in the loop may be true in isolated cases. However, the natural distinction from the majority of severely cancer patients is brutal.

The title “Young. Beautiful. Cancer” has alienated me.

“Old. Ugly. Cancer” — would be a horror title par excellence. No one would ever want to call a book or a film that. Who would want to read or watch that? A triple judgment, old, ugly, and then cancer? Cancer as a heightening of old and ugly. The combination of old and ugly alone is a no-go. An end of days feeling in contrast to the ever-present youth and beauty craze in our society.

How young, how old? As is well known, beauty is always in the eye of the beholder. Who draws the boundaries? Cancer is the only thing that disturbs such attempts at demarcation. Cancer wants to be overcome at any age, no matter whether the face is smooth or full of wrinkles. Do we need these external demarcation categories and drawers for pure advertising purposes?

No.

Not in that language.



28. RELUCTANT HEROES

This section is dedicated to the caregiving, supportive relatives of chronically and terminally ill people, some of whom swore at the beginning of their marriage to be their partner “in good times and bad.” As for the bad times, this applies no matter how hard it is and no matter how long it takes for death to put an end to it.

The Christian ethical values to which one is committed stand like a wall in our heads: charity, humanity, tolerance, loyalty, willingness to perform up to the (interpreted) categorical imperative that you should never do to a person what you do not want to be done to yourself.

This is how we were brought up, including a sense of duty.

When we step out of this restraint, we feel the pressure from outside as well as from inside. Society and your own guilty conscience will finish you off, with the exception of those who are immune to it.

You do not leave a family member who is terminally ill with cancer.

If the course of the disease requires years of care, then that’s just the way it is. One should be grateful to have a partner for such a long time, even if he or she is gradually getting sicker and sicker physically and perhaps completely changed in character. Only in the sheltered space of a patient support group or in a psychotherapy session is it permissible to say what a terrible burden it is that can make you ill yourself.

It is especially difficult for when the situation involves:

Partners who are impatient or irritable.

Partners who are not easily prepared to make restrictions and sacrifices in their own lives.

Partners with 60 hour working weeks or other high demands in their profession.

Partners whose strengths lie neither in taking care of each other without a break nor in scrupulous hygiene.

Partners who may be plagued by sporadic depression or a tendency to addictive behaviour, nothing really pathological, but who must make an effort to stay on track themselves.

Partners who seem to be uncomplainingly in danger of disappearing behind mountains of laundry and medicine.

Partners who are sick themselves and subordinate their own illness to the worse illness of the partner in the long term.

All of the above together are not a minority, are they?

Care, dependable, day-to-day running of the place: You can learn it all. Can you? Learn by doing it? Because love means being there for others without limits? I don't mean that you can because you've raised children. That's something completely different.

The time perspective plays a big part in caring. Short term, medium term a few weeks, a few months, even a few years, but year after year, getting worse, no end in sight? Mostly only on yourself and your sick partner? Purely excessive demands, even if you don't think you'll notice it for a long time.

The happiness of living in the German welfare state, where there are reliefs such as short-term care, etc., does not hide the long-term burdensome personal demands.

I am not one of those who want to transfer all private tasks to the state. Rather, this is first and foremost the content of a contract between two people. That was the deal when we got married. "Even in bad times", that was the agreement. Now it's time to pay the piper.

But what to do with the forbidden rage and the disillusioning feelings of being overwhelmed, the paralyzing helplessness? The pressure of expectation? The yawning lack of prospects? Only a few partners manage to jump into the gaps left to them by the care they receive and knit themselves into a self-determined, fulfilling life.

Then most of the time they do it alone? This requires strength, the will to resist, organizational talent, discipline and the gift of being able to enjoy the little things. One must either think about it for a long time or have the instinct to save oneself in this way, so that one can still enjoy life despite the burdens.

Family and friends praise "How brave you are, how much strength you radiate, how you manage all this...", they comfort "You grow with your tasks", advise "Try to relax in between" and between them, they say "How long can she/he keep this up? This is terrible. Hopefully it will be over soon! I couldn't do it!"

The best solution is to have someone else from the family who is reliable and helps out.

The price of being a hero is high. For there is often only the diffuse recognition of the immediate social environment, often not even from the patient themselves, who has enough to do with themselves and their illness.

It happens more often than one thinks that partners can no longer stand it at the side of their sick relatives. And then? Exclusion by society. Disguising the despair on the part of the affected person.

A grim picture. Do I not believe in the all-powerful power of love that overcomes all obstacles?

The more I wish that the often almost idyllic-looking tales and stories about the self-sacrificing care and constant emotional and mental support of sick partners are mostly true, the more skeptical I become. The reality is much more mixed and disturbed. A couple has achieved a great deal if they can manage to redefine the relationship, which requires a protracted serious chronic illness of one partner, and if they can manage to balance out lows for a while. However, there is no protection against the inevitable uncontrollable gradient that follows.

There were times when one did not live alone in pairs, especially not in old age. Caring and nursing were distributed on several family shoulders. Shared care was definitely the better care. For each carer there was still a piece of real life left.

In favour of individual autonomy, have we forgotten in recent decades that self-determination usually ends in serious illness and that we become dependent on our neighbours in the worst way? Today, everything focuses on the one neighbour, the partner, if you have one. Only when the partner himself is no longer able to do it, there may be children who intervene. But many are not prepared for this.

Today's isolation makes long-lasting care a great burden, only a few can carry it well. This burden can break up marriages and destroy joy of life, marriage vows or not.



29. FALLING DOWN THE CELLAR STAIRS

At the very top, on the first wide landing, the healthy people sit and enjoy themselves, sunlight and fresh air included. At the very bottom, on the last narrow and crooked steps in the dark and musty cellar, death and dying are lurking.

The order of the stairs is clear and simple. The better off someone is, the higher up they sit. The worse, the further down. There is constant movement on the stairs, people are walking, crawling or falling down stairs or moving up again. Sometimes just one step, sometimes several. It happens that someone falls down the whole staircase or someone slowly gasps from quite far down looking upwards.

You have less influence on how long you can sit on a step, however well you have prepared yourself for it. The watering can principle. There are, at best, probabilities. Illusory infinity—as is usual in the fantasy of the healthy at the top—has long since been abandoned anyway.

Who decides who must descend or who may ascend? Actually nobody. It just happens. One adapts to one's level until it is no longer possible, then one changes up or down, quite automatically. No one ever sits on the wrong step. Even the depressives and dementia sufferers always sit exactly right.

Each stair step is differently equipped, has different rules and defines itself above all by the degree of renunciation compared to the topmost wide step, where the healthy people cavort. The sicker, the greater the renunciation of quality of life, the lower the mobility, the larger the pill packs, the more frequent and intense the pain or weakness. The description of the differences between the levels could be continued endlessly.

However, it is not only the sick who sit on the stairs, but also special do-gooders, self-proclaimed moralists and insatiable encouragers have settled there to support them. Here the glittering commodity hope is trumps. Because very few sick people make their way back up to the light by themselves. The tireless troop of helpers can be recognized by their constant smiles, their stern looks or by their reassuring or encouraging gestures.

These people ceaselessly try to lift the sick to the next level. Often this succeeds temporarily. But hope alone is an unreliable commodity. Without physical strength and a lack of growth in real health, it is hardly a guarantee for a long-term return from the underworld. What counts much more are laboratory values, the number and/or size of metastases, the condition of the heart and circulation and/or other vital organs.

The majority of us, with a blood cancer that is actually incurable, go down the stairs. This can happen quickly, but it can also take some time. Sometimes there are breathers due to temporarily effective therapies. There are some patients who are given a new life through foreign stem cells. They climb up the stairs again a long way after their trip on the oxen in the cellar area and then remain sitting on a step in the upper third for at least a while. At least.

My current position is probably about in the middle third of the stairs, of which, by the way, nobody knows how many steps it actually has and certainly not where exactly the basement area begins. A steep descent and a bend block the view, fortunately.

A few weeks ago, I fell down a few steps from my stairs. How many, I do not know. It happened quickly, almost without warning, within a few hours. Suddenly I was lying there. Ugh!

After a few days, when I was able to think more coherently again and took stock of how I was doing, I looked around to see what the new staircase would offer me and what new restrictions I would have to expect here. Walking slowly upright a few steps was possible after some effort, but I saw my stair mates buckling so strangely in between. And when something deteriorated, I noticed it myself. I could no longer bend down, my body could not manage it, I was too weak. So there I stood with trembling knees desperately thinking of ways to manage my foothold. Even if I managed to pick it up, it fell out of my hand again and I had to try to bend my body towards the ground. The glass of water tipped over, the water ran onto the floor, my pills rolled under the bed, the jacket fell off the chair, I accidentally pushed the mobile phone off the bedside table, the toothbrush bounced onto the bathroom floor.

Swearing and crying? One of my smiling helpers and encouragers made it clear to me that cursing is a wild card for ascension: "For cursing you need strength, use it better." And yes, just a few days later I was able to bend down easier again to pick up things that had fallen down and not much fell down at all.

Now I am even back on the same level I was sitting on before my fall. That's great, because it's not uncommon that after a fall you have to settle for a step below the previous one. I have come back to where I know my way around. Despite some limitations, I've gotten quite used to this step for quite some time now and don't want to give it up so quickly. Maybe I could chain myself to the banister. But others have already tried that, it didn't help them.



30. A TOTAL CHANGE OF PLAN

In English, the term “Carer” is used to refer to a relative, friend or professional. It is not necessarily a third-party carer, but rather the person who reliably cares for the patient. There is no German word for it, and “carer” is a linguistically embarrassing solution, not really common and often still put in quotation marks, but without alternative in meaning.

My husband was my carer for years. Since my serious diagnosis in 2007, he and I had clearly agreed and repeatedly discussed that I would be the one to die first. The diagnosis, the probability of an incurable malignant disease, the statistics and our feeling for the situation had undoubtedly suggested this to us.

Not only were we completely sure of this, but also the rest of the family, friends and acquaintances, and everyone who knew about my illness.

And then one sunny September morning in 2018, my husband and caretaker simply left. His death was the biggest low blow of my life.

Although my cancer diagnosis many years before had also been a great shock, which took me months to somehow integrate into my life and my everyday life, my husband had been there for me at that time, had taken care of me and had gone through the mental turbulences as a result of the diagnosis together with me. Not only had he provided me with emotional support, but he had almost completely relieved me of physical demands such as household chores and had accompanied me for years in the marathon of doctors and therapists. Certainly, he had waited far more than 100 hours next to and with me for doctor’s consultations and therapies during all these years. He was always a willing chauffeur, no matter whether early in the morning, in the evening or on the weekend and no matter how far he had to drive. He was also a reliable prescription redeemer at the pharmacies, telephone buffer to the rest of the family, craftsman and of course buyer and cook.

He did what many marital carers do for their sick loved ones. This doing, the worrying and caring was comprehensive and available almost at any time. Sometimes even too intensively for me: “Don’t be such a mother hen!”

The caring of the relatives often remains unseen, because the focus is always on the affected patient. As psychological help is increasingly being set up in oncology wards and patient organisations are taking care of this issue, it is gradually becoming apparent that after a period of constant stress, support must also be offered to the relatives. All the more so, the longer the period of care or stress for them lasts.

In the waiting rooms of resident oncologists and clinics, the severity of the diseases prevalent there is clearly evident from the fact that most patients do not wait alone for their doctor's appointment, but together with their caregiver.

When, at an advanced age of patients, sometimes the caregivers themselves suffers from a serious illness and the mutual care and attention has become a very sensitive symbiosis, a functioning system breaks down emotionally and organisationally without replacement when one of the two no longer functions and dies.

If the person who is looking after the other suddenly disappears, in addition to the paralyzing sadness of having lost a partner who may have been with them for decades, a frighteningly large gap opens up with the question of who will take over the care, emotional support and other helpful services from now on? And the responsibility?

It is not yet a question of a stage of illness in which the degree of care has already been applied for and, under certain circumstances, external services may have to be bought in, but rather the stage before that in which necessary everyday activities and the few remaining leisure activities are nevertheless difficult and can only be carried out to a limited extent. This is due to frequent states of exhaustion, limited mobility or chronic pain of the patient's relatives. It is about the permanent settling down, e.g. with the cancer, about the real slowly creeping worsening and the fear of further aggravation.

All-round care from a former loved one is no longer available in the package. The loss and the admission that perhaps you did not appreciate this care enough when it was available in abundance is incredibly painful. Everyday life is now more strenuous, more difficult, and the fact that it is more self-determined again is little consolation. Soberly considered, this means: getting out of the comfort zone, finding out anew what still works alone and what no longer works. Depending on the form of the day, lack of physical strength, endurance and walking difficulties are the biggest opponents.

You have to ask your adult children and friends for help, distribute the order packages well and be generous in terms of timing and quality of help. If the adult children and friends are not available, you have to rent or buy what is necessary: taxi rides, drinks, help with the household, etc. It is obvious that under certain circumstances larger sums of extra money must be available. In any case, the wallet has to be pulled out more often now than in the days when a skilled husband assured "I'll do it."

An anxiety-ridden topic are situations in which one is caught up relatively suddenly by pain attacks, fever attacks or infections, felt mostly at the weekend or at night. How to assess the symptoms, should you go to the clinic or wait and possibly stay at home all night? That is hard! You get into a situation in which you can no longer trust yourself, are insecure, are at the mercy of pain.

Otherwise, the person in charge would be there to take you seriously, to calm you down and keep your nerves, who would then sometimes take full responsibility and call the ambulance on his own initiative, even though that is exactly what you didn't want to do

because of a misjudgment. How good it is when there is someone who takes control in an emergency so that you can simply let go. It becomes crystal clear now.

Neither of us expected a total change of plan. My husband and I never talked about that. I wasn't prepared for his sudden death: For this pain. And now? What now?

Other than emergency addresses of friends, I have no solution. Alone is alone for now. Over the years of experience with disease, it has proven best to think as little about such lousy scenarios as possible. All you have to do is look ahead to the next few days and weeks. After all, at some point there will be no getting around the installation of an emergency call button. An interim solution is to send my sons a smiley on WhatsApp every day, which doesn't need to be responded to, it just needs to be seen.

If it fails to appear, they will have to deal with it.



31. I WANT TO HAVE FUN!

I just want to have fun! Cooking together with good friends in the evening and talking about everything or sitting outside with a small clique in warm weather without cooking, with people who like me and whom I like, where one word results in another and we laugh a lot. Where you look at each other and the other one knows what you are about to say in a conversation that is carried by word wit and light irony. Lightheartedness and a slight buoyancy, not to be confused with superficiality. A very special atmosphere, which was taken for granted in past times and which you have contributed to yourself. With ringing glasses, candles on the table, almost like in a TV commercial for an alcoholic drink. What an illusory idyll! But something like this really has happened more often in my past. Everything negative could be faded out effortlessly for a while in such situations and it did so much good!

I want that back! It's not about drinking the world away but rather to endure it unclouded with loving irony. To feel comfortable, safe and at home. Harmony as an illusion? No, as a snapshot. Reasonable, realistic, rational we can be again tomorrow.

Where have these times gone, which often occurred in youth, study and during the first time as young parents? When did this lightness disappear? And where have the people gone with whom it was experienced? Some of them are still there, but—having become much older—they often behave differently. The retreat into the purely private has long since begun. It is only in private conversations that one learns about the many worries that individuals have. There's an "oh under every roof"; illnesses, separations, addictions, professional defeats often reduce the joy of contact by a considerable amount. Everyday life has become more strenuous for everyone. Lamentation on a high plane?

I want to have fun again... despite my incurable illness. It would do me so much good and temporarily get me out of the hamster wheel of preoccupation with my and others' illness. Distraction always gives you strength.

I have specifically asked a few friends about my selfish "fun problem", and instead of looking at me in disbelief and telling me that they didn't know what I meant, they immediately knew what I was talking about. "Yes, it's true, now that you say that. I miss that, too."



There was even an attempt between us to find explanations for this phenomenon for society as a whole: Interpersonal alienation under capitalism, a tribute to aging, the omnipresence of social media.

The problem cannot be solved by investigating the causes. It has become difficult and exhausting to find a common date in a small group. Only seldom is there unexpectedly one or the other pearl of entertainment. Nothing can be forced. I appreciate conversations in a cozy, informal atmosphere more than I used to. They resonate in me for a long time and warm my heart.



32. FUTURE EXPECTATIONS FROM THE PATIENT PERSPECTIVE

It is difficult to assess the prospects for new therapies in the field of myelodysplastic syndromes from the patient perspective. I am primarily a patient, I am neither a physician, biochemist nor geneticist, not even a scientist. I am a humanities scholar and the advanced course in biology was a very long time ago. So what can I write about current and promising MDS therapies that would not be written off somewhere and, to be honest, only half understood?

As a patient representative, I often have access to scientific MDS symposia and congresses as a listener, I have familiarized myself with many MDS-relevant topics, but I still have difficulties finding my way around the world of medical acronyms in many specialist lectures. I have to write down individual abbreviations each time and Google them later. The same is true for terms with endings like -ase, -ie or -ung (the endings are referring to German language). Behind them are connections and processes that I first have to crack for myself in order to grasp the step to their meaning for potential therapeutic approaches. This is sometimes unsatisfactory and above all amateurish.

I would wish for strongly interested patients like me, but especially for patient representatives, a descriptive “translation” of the correlations that is reduced in terms of complexity yet one can still call correct despite the reduction. This is the way cell structure and cell division are explained in biology textbooks for the 7th grade. This would be a reasonable basis on which one could build up and extend one’s understanding in a higher order if one so wished. As a patient representative you need to professionalise.

In order to expand my disease-specific knowledge, it is not enough to conquer a small island from time to time, but I would need systematic basics, especially in biochemistry, genetics and epigenetics. With the latter sub-science and here especially with molecular genetics, we have reached the centre of current research development for potential MDS therapies.

The only thing is that there are almost none. There are no new potent therapies for MDS, not a single effective drug for almost 12 years. Since 2008 (!) no drug for the treatment of MDS had been approved.

However, there has been some news from the low-risk sector for a long time. A drug that reduces the transfusion burden in transfusion-dependent MDS patients for a while in January 2020 indeed was approved by the FDA (Federal Drug Administration) and in June 2020 by the EMA (European Medicines Agency). This will mainly affect patients with a specific mutation or subtype. I have had this drug in a study for more than three years. In my case, the effect was hardly convincing after a few months, while a fellow patient became transfusion-free for years.

The experts are increasingly expanding their tools for diagnosis and more and more is understood about the disease: Early forms of myelodysplastic syndromes are unmasked. However, it is possible to go so far back in the history of the disease that it is not yet MDS and perhaps never will be. Here, new names have been “invented” to describe deviations in the genetic profile or in isolated blood values. They do not yet deserve to be called “disease” and are perhaps only an individual expression of the material examined or an acquired blood count change in old age.

The question as to which interacting factors actually cause MDS, an already malignant and progressive blood cancer, is being discussed and there is increasing agreement on certain diagnostic thresholds. Since these findings — due to a lack of available information — are systematized without any therapeutic consequences, this often seems like a mere playground in an ivory tower to us patients. The hope is that the more of the disease mechanisms are understood, the more likely therapeutic approaches become.

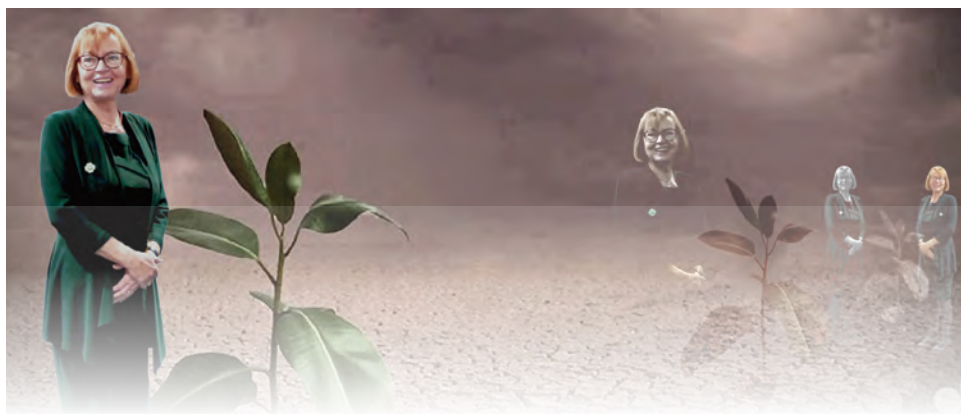
In the meantime, researchers and clinicians are scratching their heads and making it clear at specialist congresses. A physician at the 2019 “MDS Foundation International Symposium on MDS” in Copenhagen asked the audience the rhetorical question: “What do we still have if our only potent substance for high-risk patients substance Azacitidine, which was approved in 2008, no longer works in a patient? He answered the question himself by presenting a white slide: “NOTHING.”

The drug in question has a response rate of no more than 47% with potentially severe side effects and a statistical survival advantage of only nine months. This drug is only temporarily effective for high-risk MDS patients. Patients who do not receive a recommendation for stem cell transplantation, mainly due to age, are treated with this drug as standard. If the effect wears off, the only way to try to revive the effect is to combine it with other substances.

So-called combination therapies are being tested in a number of studies, but none of them are very promising because both substances are even more toxic, i.e. have more side effects, than one substance alone.

This is exactly where we currently stand in MDS treatment for high-risk patients. And this has been the case for years! Innovative monotherapies are missing. For this knowledge and its consequences, I don’t need a scientific study.

Cautious optimism is spread at specialist congresses. They say that there are young, motivated researchers with courageous questions, actually enough research funds and the willingness to de-bureaucratize medical studies. In addition, eight new effective drugs for acute leukemia have actually been approved in the last four years. These statements are intended to spread hope for early therapeutic breakthroughs in MDS. However, hope is usually not a category that is worked with in the research community. It is about evidence-based data, drug response rates and survival curves. Hope and optimism carry us through strokes of fate and crises, but they are not a good basis for research into MDS therapies.



33. YOU ARE STILL ALIVE!?

I wrote this article for the first time in March 2014. Several more years have passed since then!

Between 2005 and 2006 I suffered from a myelodysplastic syndrome. In 2007 I finally received the confirmed diagnosis and thus had my first contact with survival prognoses and statistical processes. Every time I visited the haematologist's practice, I suffered from racing heart, thoughts of flight and a constant urge to urinate for fear that my blood values would continue to drop. I was bothered by the information brochures about cancer that were lying around everywhere and the flyers advertising wigs that could be bought cheaply. I read everything I could get on the subject of blood cancer, but always with a queasy feeling in my stomach and with the actual goal of finding something that would prove to me that I was the big exception.

I wanted to find medical results and findings that would dissolve the feeling of threat in me.

Today, I am hardcore. When it comes to MDS, nothing shocks me so easily, neither for myself nor for others. To be honest, this is also due to the fact that my blood values and clinical chemistry as well as the complicated findings on cyto- and molecular genetics, have been reasonably stable for years. There are sometimes downward outliers in the blood values, but I have learned that it is never the individual value that matters, but always the trend. Only when the blood and other values fall over a longer period of time do you have to follow up and start the medical examination program. However, the possible buffer downwards is no longer large for me.

By July this year I have received 685 blood transfusions. Six hundred eighty five single bags of 250mg red blood each have kept me alive since 2008. That is 685 different donors, because the two bags that are transfused in one session are usually from two individual donors. Six hundred eighty five people and me in one room, what an image to realize how many blood donors and thus life savers I have had. My gratitude to these people knows no bounds.

Due to the many transfusions, iron has been deposited in my vital organs. Iron causes lasting damage to the tissue. Although there are tablets that are supposed to remove the iron

again, this is only partially successful due to the frequency of blood transfusions and the amount of iron that is supplied.

An important subspecies of the white blood cells has decreased so much in the meantime that I am very susceptible to infections. Some doctors go so far as to prescribe me a mouthguard for train journeys. I stay away from crowds as best I can. I get up immediately and disappear if someone near me coughs or snorts and I wash my hands all the time. My immune deficiency is regularly treated with so-called immunoglobulin infusions.

According to my haematologist at the time, my prognosis from the beginning was intermediate, i.e. between a slow and a fast course, with a medium risk of acute leukaemia. After all these years, I am still “intermediate” according to the factors that go into this prognosis, although my doctors tend not to try to categorize my case. I do not fit into any pigeonhole. I broke my first survival prognosis of three years in 2007 several times. However, when viewed in the light of day, my body is quite exhausted after many years of fighting this disease, and other aspects of the disease have been added, which make a normal and mobile life difficult for me. Nevertheless, my life is not directly threatened at the moment: that is the really decisive statement.

Already in 2007 I had told my family, relatives, friends and acquaintances that I had blood cancer and would probably not live much longer. This had been communicated to me by the doctors, unless I would be transplanted. Then I might still have a chance to even get well again.

However, because my risk profile has not increased, i.e. I am “not bad enough yet”, bone marrow transplantation, which is considered risky, is not yet an option and I am living with regular, supportive therapies in a state that can still be called “watch and wait”, although this term is reserved for an early phase of the disease in which no therapies are administered.

However, watching and waiting brings my situation right to the point. When I meet people I know, they are always visibly pleased that I look healthy or that I am “fine” to some extent. They often add that I am a little pale. By phone I only contact them when I am actually feeling quite well, translated: when I am well transfused.

It sometimes happens to me that, triggered by certain questions or remarks, I feel under “pressure to explain” why I am still alive. Following the dramatic diagnosis in 2007, no one expected that I would last this long. No one would have been surprised to learn of my premature demise. Everyone was prepared. So, did I exaggerate the drama of my illness in 2007 in the hours of phone calls and conversations with relatives and friends? No, unfortunately not! To survive eight years with MDS is quite a long time. Few patients manage ten, even less 20 years. At the time I finished my writing, I had survived for 15 years, calculated from the time the first symptoms of my disease appeared.

Occasionally I suggest to distant acquaintances the thought: “Then it can’t be that bad!” I have already been told similar things, e.g. from my former working environment. Such statements cannot be rectified, and in view of my gratitude for considerably more years of life than I ever expected, they play no role.

I am very vigilant with my future prospects. I do not waste my time and the view ahead is always a restrained one. It always only reaches a few months into the plus. I never promise that I will redeem anything in a year or more. I force myself into the now.

That’s what I’ve learned to do.



34. MDS & CORONA: A DOUBLE THREAT!

I thought that a chapter with the title “You are still alive!” would be an encouraging conclusion for my experiences and my life WITH myelodysplastic syndromes (MDS); a hopeful perspective for me and for other patients. This perspective—which I have been able to experience despite the threat for 15 years—has been possible by a stable inner frame of orientation, confirmed and strengthened by the fact that I was allowed to write the text for the first time six years ago, and I am willing to write again in the future, if I am asked to share my experiences with living with MDS.

Then came Corona.

At first rather quietly, still obscured by geographical distance, but then growing exponentially, relentlessly present everywhere and finally obtrusively loud, aggressively represented in all media and on the WWW: Corona Spezial, Corona Kompakt, Corona Live-Ticker, Robert Koch Institute, virologists, politicians, state of research, worldwide and German-specific disease rates, infection routes, deaths...phew!

An inner loop, which I felt inside, tightened imperceptibly. I registered strain, labored on unanswerable questions, felt nervousness and restlessness, the unconditional desire to somehow escape the offensive reporting. But I rarely managed to switch off; insomnia was the consequence. Somehow it took me some time to realize that I obviously belonged to the most endangered risk group because of my weak immune system. I was able to read this again and again in serious sources and this was also conveyed to me by my own doctors in tough formulations. I took this as an attack on my previously successful repression mechanism: I might not survive an infection with COVID-19 as an MDS patient.

In mid-February 2020, we celebrated my birthday with 15 guests at home and offered a night of films on March 11, 2020 as a women’s initiative on the occasion of International Women’s Day. A couple of days later, kindergartens and schools in our federal state closed. Contact barriers and distance regulations between people were issued; clinics were upgraded to be able to care for a possible wave of infected people; public life and the economy came to a standstill; basic rights were suspended.

All this is still a fact after more than a month and will remain so for a while. Even if foreseeable relaxations in the behavioural bans and corona requirements are implemented, the highest risk groups will continue to be threatened and at risk from the existence of the virus until a vaccine and medication against it are developed. For the time being, we high-risk patients will not be able to return to our previous lives, which were already limited by our disease status. For now means for at least a few months. Not an exit from, but a life with Corona. Our scope of movement, what we can and may do, what we dare to do, will be limited for a long time and we would do well to accept a certain degree of isolation in order to be protected from an infection with Corona.

Dealing with restrictions and unpredictable threats—I should be able to do that! After 15 years of cancer, I should know how to keep my head up and shift the value system so that, on the whole, you still have joy and satisfaction in your daily life when the quality of life is still reasonably good. As a patient representative, I have been passing on these “slogans” to other patients for quite some time. Since a few weeks ago, however, I have had difficulties in transferring my proven coping strategies for my blood cancer disease to the current, actually similar corona situation. I am insecure and disoriented. Telephone conversations with people who are equally affected show that I am not alone, not the only one who feels this way.

Why is this so? Why is my inner frame of orientation—the framework within which I had been assuming for years that I could cope with my illness, which I thought was fairly stable—slipping away at this very moment? The answer is not so difficult: It is because in the corona crisis not only the inner but also the outer frame of orientation is weakened. All the time with my blood cancer, I at least felt I could rely on external security, on a secure supply of medicines, on the secure availability of therapies, on my patients’ rights, even more, on my rights in general and on the fact that I have relatives and friends who come and look after me in an emergency, something that is now only possible to a limited extent because of the contact ban. I do not want to go into the conditions in many other countries that do not have such a well-functioning health system as Germany. I am more interested in the currently-felt personal perception of insecurity and difficult to bear open-mindedness, which one has unjustifiably ignored in one’s everyday life out of habit and convenience.

And so the Corona crisis teaches you a lot, namely that there are never guarantees in life on whether something will turn out for the better or worse and that you have to bear it somehow, at least the part you cannot influence yourself. This is an insight that you know very well from the course and prognosis of diseases. Obviously, I have to deal with it intensively again and work on my inner and outer frame of orientation to be able to deal with the double threat of MDS & Corona.

In any case, I will never forget anymore how fragile *is the subjective perception of security and stability* whether for sick or for healthy people.



BIOGRAPHY: BERGIT KORSCHAN-KUHLE

Bergit Korschan-Kuhle was born in 1957, and is the mother of two sons. She studied Geosciences, History, German Language and Literature in Darmstadt and Braunschweig, graduating with a Master's degree in Geosciences and passing the 1st and 2nd state examinations for grammar schools. Her career has led her into several professions, all of which were related to education, training, communication and editing. For the last 15 years of her active professional life, she worked as a coordinator at management level at a grammar school in Lower Saxony/Germany. At the age of 49, Bergit fell ill with the still incurable malignant blood disease, myelodysplastic syndromes (MDS). She had to take early retirement in 2011 because of this disease. In 2007, Bergit started to contact other patients and patient organisations via the Internet. Over the years, her growing number of contacts, her German, European and international networking, as well as her attendance at patient events and specialist congresses, have increased her desire and opportunities to become more professionally involved in patient representation. Today, Bergit is on the Board of Directors of the LHRM, a patient organisation for leukaemic patients and their carers (the German Leukämiehilfe Rhein-Main e.V.), and is the contact person of the German MDS-PAT-IG (www.mds-patienten-ig.org). Bergit is a blogger, article writer and guest speaker on topics such as blood cancer diseases from the patient perspective and patient participation. She is a Steering Committee and Founding Member of the MDS-Alliance, the global network of MDS patient organizations. The EUPATI (European Patients' Academy of Therapeutic Innovations) training course 2015/16 turned her perspective on drug research and development from head to toe and broadened her horizons, particularly with regard to the circumstances of rare and chronic diseases. This enabled her to systematically organize the experience and knowledge she had accumulated over the years from her own involvement and from her grassroots work with patients. In 2017, a well-connected MDS Facebook group was created from her commitment, which successfully promotes the exchange between patients and relatives, and passes on serious information about the disease.

MDS GLOSSARY OF TERMS

GENERAL

Acute: Sudden, such as a sudden onset of symptoms or diseases.

Acute Myeloid Leukemia (AML): A cancer of the blood cells. AML happens when very young blood cells (blasts) in the bone marrow fail to mature normally. More blastcells are produced than needed, so there is not enough room within the marrow for other normal blood cells to develop, such as red blood cells or platelets. Some cases of MDS may develop into AML. However, most do not.

Synonyms: acute myeloblastic leukemia, acute myelocytic leukemia

Anaphylaxis: A very severe allergic reaction to a foreign protein, such as in a bee sting, or to a medicine. This reaction causes the blood pressure to drop and may cause difficulty breathing. Emergency treatment is required to manage these symptoms. If very severe, anaphylaxis can progress to shock.

Synonym: anaphylactic shock

Anemia: A condition in which the number of red blood cells is below normal. This may result in fatigue, generalized weakness and shortness of breath.

Antibiotics: Medications used to treat bacterial infections and other similar microorganisms.

Antibodies: Proteins produced by plasma cells in response to foreign substances in the body.

Apheresis: A procedure in which blood is taken from a person, and part of that blood component (such as white blood cells, red blood cells, or plasma) is separated out, and the remaining blood components are reinfused back into the donor.

Aplastic Anemia: A rare and serious condition in which the bone marrow does not make enough blood cells: red blood cells, white blood cells, and platelets. The term aplastic is a Greek word meaning not to form. Anemia is a condition that happens when the red blood cell count is low. Most scientists believe that aplastic anemia happens when the immune system attacks the bone marrow stem cells. Aplastic anemia can be acquired (begin any time in life) or can be hereditary (less common, passed down from parent to child).

Synonyms: acquired aplastic anemia, hereditary aplastic anemia

Apoptosis: Death of a cell as a part of the normal lifecycle.

Autoimmune Disease: Any condition that happens when the immune system attacks the body's own normal tissues. The immune system is a complex organization within the body that is designed normally to "seek and destroy" invaders of the body, including infectious agents. Patients with autoimmune diseases frequently have unusual antibodies circulating in their blood that target their own body tissues.

Basic Research: The study of a subject to increase knowledge and understanding about it. The goal of basic research in medicine is to better understand disease. In the laboratory, basic research scientists study changes in cells and molecules linked to disease. Basic research helps lead to better ways of diagnosing, treating, and preventing disease.

Benzene: A chemical that is widely used by the chemical industry to make plastics, resins, nylon and synthetic fibers. Benzene is found in tobacco smoke, vehicle emissions, and gasoline fumes. Exposure to benzene may increase the risk of developing a bone marrow failure disease. Benzene can affect human health by causing bone marrow stem cells not to work correctly.

Biologic Agent: A substance made from a living system, such as a virus, and used to prevent or treat disease. Biological drugs include antibodies, globulin, interleukins, serum, and vaccines. Also called a biologic or biological drug.

Synonyms: biologic, biological drug

Blast Cells: Immature blood cells that would normally become fully functional mature red cells, white cells, or platelets. The number of blast cells in the bone marrow helps define how severe MDS is in a person. When 20 or more out of 100 cells in the bone marrow are blasts, this is considered acute myeloid leukemia (AML).

Synonym: precursor cell

Blood Clot: A clot or small cluster of blood cells that forms when platelets stick together. A combination of platelets and fibrin that form a mesh with the intention of preventing bleeding in response to an injury or illness. The term thrombus describes a blood clot that develops and attaches to a blood vessel. Blood clots are more common in Paroxysmal Nocturnal Hemoglobinuria (PNH) or in people with blood clotting disorders.

Synonym: thrombus

Blood Tests: Blood samples drawn from the arm that are evaluated for cell counts (red cells, white cells [and their subtypes], and platelets). The blood is also evaluated for the shape and size of the different blood cells and for how various organs are functioning such as the kidneys and liver.

Blood Thinner: A medicine used to treat or prevent blood clots. Also called anticoagulants or blood thinners. Some common blood thinners are enoxaparin or clexaine (Lovenox or Clexane), heparin (Calciparine or Liquaemin), and warfarin (Coumadin).

Synonyms: anticoagulant, anti-clotting

Blood Transfusion: A procedure in which whole blood or one of its components is given to a person through an intravenous (IV) line into the bloodstream.

A red blood cell transfusion or a platelet transfusion can provide temporary relief for some patients with low blood counts.

Bone Marrow: The soft, sponge-like tissue in the center of bones that functions like a factory to produce white blood cells, red blood cells, and platelets.

Bone Marrow Aspirate: The bone marrow aspirate is a sample of the liquid portion of the bone marrow. It is used to obtain spicules—a small collection of blood forming cells. This provides information about the shape of the cells (morphology), how the cells are maturing (differentiation) and the number of blasts (immature cells) in the bone marrow. The aspirate may also be used for additional testing that may help to determine the cause of the cytopenias, such as cytogenetics.

Bone Marrow Aspiration: The process of removing bone marrow from a specific area using a small needle and syringe. Used for diagnostic purposes. Tests may also be run on the bone marrow cells to look for any genetic abnormalities.

Bone Marrow Biopsy: The bone marrow biopsy is a small core (the shape and size of a medium pencil lead) of the spongy center of the bone marrow. It provides information about the cellularity of the bone marrow (crowded=hypercellular, empty=hypocellular). It will also provide useful information about iron storage, scarring (fibrosis), and the presence of any other abnormal cells.

Synonym: Bone Marrow Trephine Biopsy

Bone Marrow Failure: A condition that occurs when the bone marrow stops making enough healthy blood cells. The most common of these rare diseases are myelodysplastic syndromes (MDS), aplastic anemia, and paroxysmal nocturnal hemoglobinuria (PNH). Bone marrow failure can be acquired (begin any time in life) or can be hereditary (less common, passed down from parent to child).

Cellularity: How much of the bone marrow volume is occupied by various types of blood cells.

Chemotherapy: The use of medicines that kill cells (cytotoxic agents). People with high-risk or intermediate-2 risk myelodysplastic syndrome (MDS) may be given chemotherapy. Chemotherapy may also hurt healthy cells causing side-effects. If chemotherapy works in controlling abnormal cells, then relatively normal blood cells will start to grow again. Chemotherapy agents include: cytarabine (Ara-C) and hydroxyurea (Hydrea), daunorubicin (Cerubidine), idarubicin (Idamycin), and mitoxantrone (Novantrone).

Chronic Illness: A medical condition that lasts a long time. A chronic illness can affect a person's lifestyle, ability to work, physical abilities and independence.

Chromosomes: A structure that contains your genetic information, or DNA. Normally each person has 23 pairs of chromosomes.

Clinical Trial: A type of research study that tests how a drug, medical device, or treatment approach works in people. There are several types of clinical trials. Treatment trials test new treatment options. Diagnostic trials test new ways to diagnose a disease. Screening trials test the best way to detect a disease or health problem. Quality of life (supportive care) trials study ways to improve the comfort of people with chronic illness. Prevention trials look for better ways to prevent disease in people who have never had the disease.

Trials are in four phases:

- Phase I tests a new drug or treatment in a small group to see if it is safe.
- Phase II expands the study to a larger group of people to find out if it works.
- Phase III expands the study to an even larger group of people to compare it to the standard treatment for the disease.
- Phase IV takes place after the drug or treatment has been licensed and marketed to find out the long-term impact of the new treatment.

Clone: To make copies. Bone marrow stem cells clone themselves all the time. The cloned stem cells become mature blood cells that leave the bone marrow and enter the bloodstream. Abnormal clones are associated with cancers, such as MDS.

Coagulate: To thicken.

Normal blood platelets cause the blood to coagulate and stop bleeding.

Combination Chemotherapy: The use of more than one drug during cancer treatments.

Comorbidities: Additional diseases beyond MDS.

Complete Blood Count (CBC): The CBC measures the number of white blood cells (WBC) and the number and size of red blood cells, the total amount of hemoglobin, and the fraction of the blood made up of red blood cells.

Complex Karyotype: Three or more abnormalities in their chromosomes.

Conditioning Treatment: Used to kill all remaining cancer cells before stem cell transplantation.

Cytogenetics: Testing that is performed on bone marrow samples and examines the chromosomes of the cells. Your cytogenetic results are used to identify the type of MDS you have and to calculate the International Prognostic Scoring System (IPSS) and the revised IPSS

(IPSS-R) risk category.

Common abnormalities include:

1. Deletion 5q – deletion of chromosome 5
2. Deletion 20 – deletion of chromosome 20
3. Deletion Y – deletion of the Y chromosome
4. Monosomy 7 – loss of one of the two 7 chromosomes
5. Trisomy 8 – addition of a third chromosome 8

Synonyms: Chromosomes, Karyotype, DNA

Cytogenetic Remission: No sign of previously detected abnormal chromosomes are found. This represents a response to treatment. When a bone marrow test is performed on a patient with 5q deletion MDS, and there are no signs of an abnormal chromosome 5, then that patient has achieved a cytogenetic remission. Also called cytogenetic response.

Cytokines: Proteins

Cytopenia: A deficiency of (or too few) mature cells in the blood. Deficiencies can occur in red cells, white cells, and/or platelets.

Cytotoxic Agent: A medicine that kills certain cells. Chemotherapy for MDS patients often involves the use of cytotoxic agents.

del(5q): Deletion in the long (q) arm of chromosome 5.

De Novo: The original source of disease, something present at the start. MDS may be de novo, the original source of disease, or treatment related, caused from chemotherapy or radiation given for other forms of cancer.

Differentiation: The process of cells maturing to become healthy adult cells of a particular type (i.e. red cells, white cells, and platelets).

Dietary Supplement: Vitamins, minerals, herbs and other substances meant to improve your nutritional intake. Dietary supplements are taken by mouth in the form of a pill, capsule, tablet or liquid.

DNA Methylation: A process that helps control gene activity, resulting in blockage of cell growth.

Dysplasia: Abnormal shape and appearance or morphology, of a cell.

Synonym: dysplastic

Embolus: A blood clot or other foreign matter that gets into the bloodstream and gets stuck in a blood vessel.

Epidemiology: The study of patterns and causes of disease in groups of people. Researchers who study how many people have a disease, how many new cases are diagnosed each year, where patients are located, and environmental or other factors that influence disease, are known as Epidemiologists.

Erythroid Response

- In patients who have not received red blood transfusions—hemoglobin increase of 1.5 g/dl
- In those who have had transfusions—reduction in transfusions by at least four units of packed red blood cells over 8 weeks compared with the 8 weeks before treatment

Erythropoietin (EPO): A protein substance manufactured by the kidneys in response to low oxygen levels in body tissues. Erythropoietin stimulates the production of red blood cells in the bone marrow.

Erythropoietin-stimulating Agent (ESA): A medicine used to help the bone marrow make more red blood cells. Epoetin alfa (Epogen, Procrit) and darbepoetin alfa (Aranesp) are erythropoiesis-stimulating agents that can help boost the red blood cell count of some bone marrow failure patients. Also called red blood cell growth factor.

Etiology: The cause or origin of a disease.

FAB Classification: A criteria used for classifying different types of myelodysplastic syndromes (MDS). The FAB (French, American, British) Classification System was developed by a group of French, American and British scientists. This system is based on 2 main factors: the percentage of blast cells in bone marrow, and the percentage of blast cells in the bloodstream. The FAB system is somewhat outdated, but is still used by some doctors today. The World Health Organization (WHO) Classification System has largely replaced the FAB Classification System.

Fanconi Anemia: A rare inherited disorder that happens when the bone marrow does not make enough blood cells: red cells, white cells, and platelets.

Fanconi anemia is diagnosed early in life. People with Fanconi anemia have a high likelihood of developing cancer. Genetic testing is used to diagnose Fanconi anemia.

Fatigue: A feeling of low or no energy, general feeling of tiredness with normal activity. Rest does not necessarily resolve fatigue.

Synonyms: tired, exhaustion, lethargy, malaise

Ferritin: A protein inside of cells that stores iron for later use by your body. Sometimes ferritin is released into the blood. The ferritin level in the blood is called serum ferritin.

Fibrosis: Scarring of tissue. Fibrosis of the bone marrow is a feature seen in some types of unclassified myelodysplastic syndrome (MDS).

Flow Cytometry: A laboratory test that gives information about cells, such as size, shape, and percentage of live cells. Flow cytometry is the test doctors use to assess for specific proteins on the surface of blood cells. It is the standard test for confirming a diagnosis of paroxysmal nocturnal hemoglobinuria (PNH).

Synonyms: Flow, Immunophenotyping Fluorescence-activated cell sorting (FACS)

Fluorescence In Situ Hybridization (FISH): An important laboratory test used to help doctors look for chromosomal abnormalities and other genetic mutations. Fluorescence in situ hybridization, also called FISH, directs colored light under a microscope at parts of chromosomes or genes. Missing or rearranged chromosomes are identified using FISH.

Folate: A B-vitamin that is found in fresh or lightly cooked green vegetables. It helps the bone marrow make normal blood cells. Most people get enough folate in their diet. Doctors may have people with paroxysmal nocturnal hemoglobinuria (PNH) take a man-made form of folate called folic acid.

Gene Expression: The process that genes use to make their products, such as proteins.

Graft-Versus-Host Disease (GVHD): Attack by transplanted cells on the recipient's body in which the transplanted cells cause inflammation of some normal tissues.

- Acute: within 3 months of transplantation
- Chronic: starting more than 3 months after transplantation

Graft-Versus-Leukemia Effect: T cells (part of the immune system) in the donated stem cells can attack the remaining cancer cells.

Haploidentical Stem Cell Transplantation: The donor's blood markers match half the patient's markers.

Hematocrit (HCT): Percent of the total blood volume that is made up of red blood cells. In men a normal hematocrit is 40–52% while in women the normal is 36–46%. Hematocrit is part of a complete blood count. Also called HCT, packed cell volume, PCV. (see red blood cells)

Synonyms: packed cell volume, PCV

Hematologist: A doctor who specializes in the diseases and disorders of blood.

Hematopoiesis: The formation and development of blood cells.

Hemochromatosis: A condition that occurs when the body absorbs and stores too much iron. This leads to a condition called iron overload. In the United States, hemochromatosis is usually caused by a genetic disorder. Organ damage, particularly to the liver and heart, can occur if iron overload is not treated.

Hemolysis: The destruction of red blood cells.

Hypercellular: A condition in which there are too many cells within the bone marrow.

Hypocellular: A condition in which there are too few cells, within the bone marrow. Patients with aplastic anemia have hypocellular bone marrow.

Hypomethylating Agent: A hypomethylating agent is a drug that inhibits DNA methylation. Works by preventing certain genes involved in controlling cancer from being silenced, allowing for the normal functioning of the tumor suppressor genes.

Synonym: demethylating agent

Idiopathic: Usually refers to any condition with no known cause.

Immature Blood Cells: May be called stem cells, progenitor cells or blasts.

Immune Deficiency: A decreased ability of the immune system to fight infection.

Immune System: The complex group of organs and cells that defend the body against infection and disease.

Immunocompromised: Occurs when the immune system is not functioning properly, leaving the patient open to infection. A person can be immunocompromised due to low white blood cell count or due to some medicines.

Synonym: immune compromised

Immunosuppressive Drug: Drugs that lower the body's immune response in autoimmune diseases. These drugs may be used to allow the bone marrow stem cells to grow and make new blood cells. ATG (antithymocyte globulin) or ALG (antilymphocyte globulin) with cyclosporine are used to treat bone marrow failure in aplastic anemia. Immunosuppressive drugs may help some patients with myelodysplastic syndromes (MDS) and paroxysmal nocturnal hemoglobinuria (PNH).

Intravenous Infusion: A method of getting fluids or medicines directly into the bloodstream over a period of time. Also called IV infusion.

Synonym: IV infusion

IPSS/IPSS-R: An International Prognostic Scoring System – system for grading the severity of MDS. The system turns patient data into a score. The score tells how quickly a myelodysplastic

syndrome (MDS) case is progressing and helps predict what may happen with the patient's MDS in the future.

Iron Chelation Therapy: A drug therapy to remove extra iron from the body. Patients with high blood iron (ferritin) levels may receive iron chelation therapy. The U.S. Food and Drug Administration (FDA) has approved two iron chelators to treat iron overload in the U.S.: deferasirox, an oral iron chelator, and deferoxamine, a liquid given by injection, these may differ depending on which country you live in.

Iron Overload: A condition that occurs when too much iron accumulates in the body. Bone marrow failure disease patients who need regular red blood cell transfusions are at risk for iron overload. Organ damage can occur if iron overload is not treated.

Ischemia: Occurs when the blood supply to specific organ or part of the body is cut off, causing a localized lack of oxygen.

Lymphatic System: A network of organs, lymph nodes, lymph ducts, and lymph vessels that help keep the body's fluids in balance and help the body fight infection.

Minimal Residual Disease: Small numbers of cancer cells that stay in the body after treatment.

Monosomy 7: Describes the loss of one of the two number 7 chromosomes. "Mono" means one and "somy" comes from the word chromosome. Bone marrow samples are used to detect monosomy 7 and other genetic abnormalities. Monosomy 7 can occur in adult patients with MDS and can occur in childhood bone marrow failure diseases.

Morphology: The study of the structure and form of an organism or one of its parts.

Multilineage Dysplasia: Abnormalities in more than one type of blood cell

Mutation: Any change or alteration in a gene. A mutation may cause disease or may be a normal variation.

Myelo: A Greek word meaning marrow.

Myelodysplastic Syndromes (MDS): The Myelodysplastic Syndromes (MDS) are a group of bone marrow failure disorders. Myelo refers to the bone marrow. Dysplastic means abnormal growth or development. In MDS, the bone marrow does not make blood cells normally. The result is too few cells or low blood counts (cytopenias) and cells that do not function properly.

The most common cytopenias include:

Anemia: low red blood cells (oxygen carrying cells)

Thrombocytopenia: low platelets (cells that help to clot the blood)

Leukopenia: low white blood cells (WBC) (help to fight infection)

Neutropenia: low neutrophils (most important type of WBC for fighting infection)

Natural Killer Cells: A type of cell that lacks B-cell and T-cell receptors and attacks mutant and virus-infected cells.

Neutropenia: A deficiency (below-normal number) of mature white blood cells called neutrophils that assist in fighting bacterial infections.

Neutropenic Diet: A diet for a patient with very low white blood cell count. A neutropenic diet avoids the use of certain foods that can contain bacteria or fungus, such as raw meats, unpasteurized dairy products, aged cheeses, fermented drinks, and unwashed fruits and vegetables.

Oncologist: A doctor who specializes in the treatment and prevention of cancer.

Over-the-Counter (OTC) Medicine: A medicine that is available without a prescription from the doctor. Also called OTC medicine.

Packed RBCs: A concentrated blood product in which most of the plasma, the fluid part of blood, is removed to make red blood cell transfusions easier and faster.

Pancytopenia: A reduced number of all types of blood cells – red blood cells, white blood cells, and platelets.

Paroxysmal Nocturnal Hemoglobinuria (PNH): A rare and serious blood disease that causes red blood cells to break apart. Paroxysmal means sudden and irregular. Nocturnal means at night. Hemoglobinuria means hemoglobin in the urine. Hemoglobin is the red part of red blood cells. A person with PNH may have episodes of dark urine in the morning, but this symptom is not present in all PNH patients.

Pathophysiology: Abnormal function or processes that cause or are associated with disease or injury.

Pediatric MDS: MDS is rare in children; but it does happen. Most patients are 60 years old or older.

Peripheral Blood Stem Cell (PBSC) Transplant: A procedure where stem cells are collected from the donor's circulating (peripheral) blood. These stem cells are then given to the patient through an intravenous (IV) line. In time, donated stem cells start making new, healthy blood cells. Also called PBSC transplant.

Petechiae: Small, flat red or purplish spots caused by pinpoint bleeding into the skin. It is often a sign of a low platelet count.

Pharmacist: A highly trained and licensed professional whose job concerns the preparation, distribution, and use of prescription drugs. A pharmacist also advises patients, as well as physicians and other health practitioners, on the selection, dosages, interactions, and side effects of medications.

Placebo: A placebo is an inactive pill, liquid, or powder that has no treatment value. Placebo use in clinical trials is extremely uncommon today.

Platelets: Irregularly shaped, colorless cells that are present in blood. Their sticky surface lets them, along with other substances, form clots to stop bleeding. Also called thrombocytes.

Platelet Transfusion: A procedure in which platelets are given to a person through an intravenous (IV) line into the blood-stream. Platelets are more likely than red blood cells to cause an immune response, such as chills and fever. The use of platelets from one donor (apheresis) reduces the chance of reaction to transfused platelets. Transfused platelets increase the blood platelet count and help control bruising and bleeding.

Prophylactic: Something that prevents or protects. For example, blood thinners may be given as a prophylactic measure to prevent blood clots in high risk patients.

Pure Red Cell Aplasia (PRCA): A condition that occurs when the bone marrow stem cells do not make red blood cells. Red blood cell counts are low. White blood cell and platelet counts are normal.

Red Blood Cell (RBC) Transfusion: A procedure in which packed red blood cells are given to a person through an intravenous (IV) line into the bloodstream. Transfused red blood cells increase the blood count and help improve symptoms of anemia. Before transfused blood is given, donated blood is typed and cross matched to the recipient's blood. Also called RBC transfusion.

Refractory: Not responsive to treatment or cure. For example, refractory anemia is a low red blood cell count that doesn't respond to standard treatments.

Reticulocyte: An immature red blood cell. Reticulocytes are normally found in the bone marrow. They are present in the bloodstream only in very low numbers.

Remission: Disappearance of the signs and symptoms of cancer. A remission may be complete (CR) or partial (PR).

Revised IPSS (IPSS-R): Takes more information into account than the IPSS and categorizes patients into five risk groups instead of four.

Ring Sideroblast: A red blood cell that has too much iron. The iron typically forms a ring around the cell's nucleus.

Secondary MDS: A type of MDS that is caused by a previous treatment for another disorder or disease. Treatments typically associated with secondary MDS include radiation therapy and chemotherapy used to treat cancer. Also called therapy-related MDS, T-MDS.

Synonyms: T-MDS, therapy-related MDS

Serum Erythropoietin: Amount of erythropoietin that is present normally in an individual's blood.

Serum Sickness: An immune system reaction to foreign proteins in certain medicines. Serum sickness can be a side effect of ATG, a medication used in the treatment of aplastic anemia, causing fever, rash, joint pain, and muscle aches.

Single Lineage Dysplasia: Abnormalities in only one type of blood cell.

Somatic Mutation: Change in a gene that happens after conception in a patient's cells, is not inherited, and is not passed on to the patient's children.

Stem Cells: Cells in the body that develop into other cells. Adult stem cells in the body repair and maintain the organ or tissue in which they are found. Blood-forming (hemapoietic) stem cells are found in the bone marrow. These cells make copies of themselves and develop into red cells, white cells, and platelets. Embryonic stem cells come from human embryos and may be used in medical research.

Subcutaneous Injection: A method of giving medicine in the fatty tissue area under the skin using a short needle.

Synonyms: shot, injection

Supportive Care: Care given to improve the quality of life, or comfort, of a person with a chronic illness. Supportive care treats the symptoms rather than the underlying cause of a disease. The goal is to help the patient feel better. Patients with low blood counts may be given blood transfusions as supportive care to help manage the symptoms of their disease. Also called palliative care, symptom management.

Synonyms: palliative care, symptom management

Therapy Related MDS: See Secondary MDS

Thrombus: A blood clot that develops and attaches to a blood vessel.

Thrombosis: The process of forming a blood clot.

Thrombocytopenia: A condition in which the number of mature platelets, or thrombocytes, is below normal. When severe, the tendency to bruise and bleed more easily can occur.

Transfusion: Process by which blood or one of its components (e.g., red blood cells, plasma, platelets) is delivered directly into the bloodstream by vein (intravenous or IV), similar to other IV medications.

Transfusion Independence: No longer needing any type of blood transfusion.

Treatment Failure: Occurs when a patient does not respond to the treatment, responds only temporarily, or has to stop the treatment because of side effects.

White Blood Cells (WBC): Cells produced in the bone marrow and lymph nodes. White cells are key cells in the immune system that prevent or fight infection.

World Health Organization (WHO) Classification: The most current system for classifying leukemia and myelodysplastic syndromes (MDS), it was developed by the World Health Organization (WHO). This system is based on patient data from around the world and on the most up-to-date knowledge of MDS. WHO Classification of MDS consists of many subtypes based on tests of the blood and bone marrow.

RED BLOOD CELLS

Erythrocyte: A red blood cell. It carries oxygen to body cells and carbon dioxide away from the cells.

(See red blood cells)

Red Blood Cells (RBC): These are cells that carry oxygen to your tissues. *(See erythrocyte)*

WHITE BLOOD CELLS

ANC (absolute neutrophil count): A measure of the actual number of mature neutrophils in a given volume of blood.

Basophil: Type of white blood cell that plays a role in allergic reactions and asthma.

Eosinophil: Type of white blood cell that kills parasites and plays a role in allergic reactions.

Granulocyte: A term for any of the white blood cell types that have granules containing enzymes to help fight infection: neutrophils, eosinophils and basophils.

Lymphocytes: Small white blood cells produced in the lymphoid organs (the lymph nodes, spleen, thymus, and tonsils) or bone marrow that are essential for normal function of the immune system.

Monocyte: A white blood cell that helps the body fight infections from some bacteria such as tuberculosis.

Neutrophil: A type of white blood cell that functions to destroy bacteria. When the number of neutrophils is too low, the body is at greater risk for developing an infection.

BONE MARROW BIOPSY

Allogeneic Stem Cell Transplantation: A procedure in which matched bone marrow or peripheral blood stem cells from a donor (usually related) are collected, stored, and infused into a patient (recipient) following high-dose chemotherapy with or without radiation therapy. In time, donated stem cells given to the patient begin making new, healthy blood cells (known as engraftment).

Allograft: An allogeneic stem cell collection used for transplant.

Autograft: An autologous stem cell collection used for transplant.

Autologous Stem Cell Transplantation: A procedure in which a patient's own stem cells from bone marrow or peripheral blood are collected, stored, and reinfused following high-dose chemotherapy or radiation therapy. In time, donated stem cells given to the patient begin making new, healthy blood cells (known as engraftment).

Bone Marrow Transplant: A procedure in which high doses of chemotherapy or radiation therapy are used to eradicate disease in the bone marrow and lymphatic system and then are replaced with healthy bone marrow from a donor or the patient.

Cord Blood Transplant: A procedure where umbilical cord stem cells are given to the patient through an intravenous (IV) line. Stem cells are collected from an umbilical cord right after the birth of a baby. They are kept frozen until needed. In time, donated stem cells given to the patient begin making new, healthy blood cells.

Engraftment: Refers to how well the donor cells (graft) are accepted by the patient's immune system (host) after a bone marrow or stem cell transplant. Several factors contribute to better engraftment: physical condition of the patient, how severe the disease is, type of donor available, age of patient. Successful engraftment results in new bone marrow that produces healthy blood cells (new white blood cells, red blood cells, and platelets).

Graft-Versus-Host Disease (GVHD): GVHD is a common complication of allogeneic bone marrow/stem cell transplantation. It is caused when the donor's immune cells, now in the patient, begin to see the patient's body as foreign and mount an immune response. GVHD most commonly affects the recipient's skin, intestines, or liver. Severity can range from mild to very severe. In some cases, GVHD can be prevented or treated with specific drugs to suppress the body's immune cells (immunosuppressive drug therapy).

Human Leukocyte Antigen (HLA): One of a group of proteins found on the surface of white blood cells and other cells. These antigens differ from person to person. A human leukocyte antigen test is done before a stem cell transplant to closely match a donor and a recipient.

Matched Related Donor: Bone marrow/stem cell donor that is a sibling or another blood relative to the patient.

Mini-Transplant: *See Non-Myeloablative Transplant*

Myeloablation: The killing of bone marrow by radiation or chemotherapy. This term usually refers to the complete or near-complete destruction of the bone marrow.

Non-Myeloablative Transplant: Type of allogeneic stem cell or bone marrow transplant that uses lower doses of chemotherapy. This reduces side effects caused by chemotherapy, making it more tolerable for older adults. It does not reduce the risk of graft-versus-host disease. Also called nonmyeloablative transplant or reduced intensity transplant.

Reduced Intensity Transplant: Procedure similar to standard bone marrow transplant. The reduced intensity transplant uses a mild form of chemotherapy pre-treatment. This reduces side effects caused by chemotherapy, making it more tolerable for older adults. It does not reduce the risk of graft-versus-host disease. Also called nonmyeloablative transplant.

Unrelated Donor: A donor that is not a sibling or other familial relation of the patient (recipient).

MEDICATIONS

Adverse Event (AE): Any undesired actions or effects of a drug or treatment.

Synonyms: side effect, toxicity

Antibiotic Therapy (AB): Used to treat bacterial infections or prevent recurrence of bacterial infections.

Antithymocyte Globulin (ATG): An immunosuppressive medication that eliminates abnormally proliferating white blood cells called T lymphocytes which disrupt normal blood cell growth. This may restore normal production of red blood cells which may lead to transfusion independence. The three brand-name drugs are Thymoglobulin®, Lymoglobulin®, and Atgam®.

Colony-Stimulating Factor (CSF): Protein that stimulates the development and growth of blood cells; sometimes called growth factor. Granulocyte colony-stimulating factor is a CSF that is used to stimulate stem cells from the bone marrow into the bloodstream prior to apheresis.

Corticosteroids: Also called “steroids,” corticosteroids are powerful anti-inflammatory medicines used to treat many diseases and conditions. They are similar to a protein called cortisol that is made in the adrenal glands. Names of corticosteroids include prednisone and dexamethasone.

Synonym: steroids

Dacogen™ (decitabine): A medication used in treating some types of MDS and AML. Dacogen works by preventing certain genes involved in controlling cancer from being silenced, allowing for the normal functioning of genes within the body. It is a DNA hypomethylating agent that is administered intravenously (IV).

Desferal® (deferoxamine): A medication that binds to iron and promotes its removal from the body for treatment of transfusion-dependent iron overload. It is an iron-chelating drug that is administered subcutaneously (under the skin).

Erythropoietin (EPO): A “recombinant” form of a natural growth factor used to treat symptoms associated with anemia. It stimulates the bone marrow to produce red blood cells. The three brand-name drugs are Aranesp®, Epogen®, and Procrit®. These drugs are administered intravenously or subcutaneously.

Exjade® (deferasirox): A medication that binds to iron and promotes its removal from the body for treatment of transfusion-dependent iron overload. It is an iron-chelating drug that is administered orally.

Growth Factors (hematopoietic): A substance made by the body that stimulates the bone marrow to produce blood cells. Some growth factors are man-made in the laboratory and used for treating low blood counts. These include red blood cell growth factors called erythropoietin (EPO) and darbepoetin, and white blood cell growth factors called granulocyte colony stimulating factors (GCSF) and granulocyte macrophage colony stimulating factors (GMCSF). Also called cytokines.

Synonym: cytokine

Hycamtin® (topotecan hydrochloride): A chemotherapy agent that may result in remission of MDS. It is administered intravenously.

JADENU™ (deferasirox): A medication that binds to iron and promotes its removal from the body for treatment of transfusion-dependent iron overload. It is an iron-chelating drug that is administered orally. It is a new oral formulation of Exjade tablets. Whereas the Exjade tablet

must be mixed in liquid and taken on an empty stomach, Jadenu can be taken in a single step, with or without a light meal, simplifying administration of treatment for chronic iron overload.

Leukine® (sargramostim): A growth factor, granulocyte macrophage colony-stimulating factor (GM-CSF), used for the treatment of neutropenia. It increases white cell production, which may help to reduce the likelihood of additional infection. It is administered subcutaneously.

Neupogen® (filgrastim): A growth factor, granulocyte colony-stimulating factor (G-CSF), used for the treatment of neutropenia. It increases white cell production, which may help to reduce the likelihood of additional infection. It is administered subcutaneously.

Prednisone: A corticosteroid that is used for many reasons. It is prescribed when the body is not producing enough of this chemical on its own. It is sometimes prescribed with ATG treatment to reduce the risk of anaphylaxis or serum sickness. It helps by reducing the antibody production of the immune system and in treating various allergic conditions. There are many brand names of prednisone. (*See corticosteroid*)

Pyridoxine (Vitamin B6): A vitamin needed to make red blood cells. It can be useful in improving red blood cell counts in sideroblastic anemia by increasing the red blood cell production.

Reblozyl® (Luspatercept): Medication indicated for the treatment of anemia failing an erythropoiesis stimulating agent requiring 2 or more red blood cell units of 8 weeks in adults with very low to intermediate risk myelodysplastic syndromes with ring sideroblasts (MDS-RS) or with myelodysplastic/ myeloproliferative neoplasm with ring sideroblasts and thrombocytosis (MDS/MPN-RS-T). Luspatercept may restore differentiation and maturation of red blood cells (normal development) in the last phase of erythroid cell (red blood cell) development in some patients with lower risk MDS.

Revlimid® (lenalidomide): A medication that works by stimulating the immune system, preventing new blood vessel growth, and stimulating cell death. It is categorized as an immunomodulatory agent and is taken orally.

Telintra™ (TLK199): A medication that inhibits a key enzyme (glutathione S-transferase P1-1 or GST P1-1) involved in cell growth and proliferation; this results in normal blood cell production. It is given intravenously (IV).

Thalomid® (thalidomide): A medication that reduces the blood supply in the marrow, thereby working to limit the growth of abnormal blood cells. It also acts to interfere with other proteins (cytokines) that promote premature death of cells in the bone marrow. It is taken orally.

Trisenox® (arsenic trioxide): A medication that inhibits new blood vessel growth and stimulates cell death of abnormal cells. It may increase transfusion independence. It is administered as an intravenous infusion (IV).

Vidaza™ (azacitidine, 5-azacytidine): A medication that works by preventing a cellular process (methylation) that silences the genes involved in controlling the development of cancer. It may increase red blood cells, transfusion independence, hemoglobin, white blood cells, platelets, and/or decreases the amount of blast cells within the bone marrow. It is categorized as a DNA hypomethylating agent and can be administered intravenously (IV) or subcutaneously (under the skin).



**"Being diagnosed and treated for MDS is like
riding a physical and emotional rollercoaster.
It's not fun, but you will cope as you get
used to it and learn as much as you can about it."**

**Bergit Korschan-Kuhle
Myelodysplastic Syndromes Patient**