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Two conclusions out of two difficult articles

1 Chondroma, Enchondroma, Chondrosarcoma

In the discussion of the disease Ollier-Maffucci the word Chondroma is no longer in use, in both of the above given articles it appears only in quoted texts (Herget et al.) or even only in the title of listed works (in the sixties I knew the illness not as Enchondromatosis, but only as Chondromatosis – or as "Morbus Ollier" when I listened to my doctor while he dictated across the patient, during curetting = scraping the sick cartilage in the right hand, to his beautiful secretary; the whole time of surgery I kept an eye on her as she was holding her sheet over the head on the door frame and was looking back to the victim as encouragingly as possible). The term Chondromatosis now, read out of the two articles refers to a phenomenon without real medical significance, because it can appear anywhere and, where it appears, does not cause a deeper problem, at least not a problem with regard to cancer. If there is a piece of the body as Chondroma, this sick part of cartilage could be indeed excessively large (one knows the german word “Überbein” = grown over the edge = ganglion), but the Chondroma only wraps the bone, stays on its surface and keeps itself outside.

The prefix En- of the word Enochondroma means that the diseased parts of bone and cartilage not only cover the bone, but go into it, exist in it, or come out of it: instead of producing new bone, the diseased cartilage eats the bone and lets itself grow. (By the way: if it becomes possible to let grow cartilage in the pharmaindustry by eliminating the inherent process of making bone material out of itself after a certain time of growth, it will be possible to isolate the physiological instance of growth and, later on, to stop by that knowledge the process of overproduction of cartilage in Olliers. – No help at all for young patients who undergo the treatment of leg lengthening but nevertheless a gateway for research on Ollier-Maffucci anyhow.)

Whenever a part of the skeleton is called Enchondroma, it is not a malignant tumour. But an Enchondroma can mutate, degenerate, or become dedifferentiated into a malignant tumour. Then it's called Chondrosarcoma. Today, there are Chondrosarcomas in three grades with the values or dedifferentiation levels 1, 2 and 3. The values 2 and 3 are dangerous: the Chondrosarcomas 2 and 3 are both malignant and produce metastases, number 3 much faster than 2. A Chondrosarcoma in dedifferentiation grade 1 is exactly the same as an Enchondroma. But it is a growing, space-occupying and therefore pain-inducing Enchondroma that, if left untreated, will break both internal organ parts and the body surface. – It is easy to see that as the Enchondromas in a child with Ollier's disease grow parallel to normal biological growth until reaching adulthood, they require constant and special

monitoring. A young person with Ollier (and anyway with the Maffucci syndrome) must have a kind of personal doctor who organizes a constant check to respond in time if an Enchondroma grows faster than normally and mutates this way into a Chondrosarcoma. Worldwide, this life-threatening vulnerability in the healthcare system is anything but optimally organized. The Ollier-Maffucci disease is misclassified in the healthcare system if patients or their parents have to seek a competent doctor by themselves. (I can not say that this problem would be urgent in Switzerland or in the neighboring German-speaking countries: until the age of 20, I had seen my doctor at least once a year, and there were constant blood tests – with the later confession that there was a crisis in the year of 14 or 16. I did not know the type of test results, but the doctor told me that they were three times as high as normal, over twentyone instead of below seven. – This may be the reason for the absence of an Ollier-Maffucci Group in German language that the medical care here is very good, in Lucerne from 1962 to 1984 with Dr. med. Jost Zemp and at the Insel Hospital in Berne from 2002 up to now in any case optimal.)

2 Risk of Malignancy

The field of medical prognosis remains empty in Ollier's patient's case files. Nevertheless, one always has a sound like in a swarm of bees in the ears, if one is staying in hospital areas because of the illness. Medical staff is under pressure to make statements about a patient's future as if Fortune Telling were part of the therapy. But the numbers that are served are through the tape: Nonsense.

The text by Verdegaal et al. is shown in a dress of statistical analysis, but it is overall a presentation of event numbers in the form of statistical graphics and tables. That would be legitimate if one had omitted the worst embarrassments such as a death chart based on a selection of individual cases and the indication of the statistical mean of the mutation in children and older adults – such mutations have as much to do with each other as the famous comparison of apples and pears, where an average does not consist of oranges.

Speaking in the outdated language of Chondromatosis, the risk of mutation is extremely low. Doctors who are at this level of discourse then tell the parents of young Olliers, the disease is, apart from leg extensions and curettage, absolutely unproblematic and everyday life always painless. This is a medical negligence. For in the prognosis of malignancy the Chondroma has no business. If one limits oneself properly to the concept of Enchondromatosis, a global statistical risk analysis still remains problematic, if the data over the illness cases can not finally be collected and published globally. At the moment, the numbers look bleak and make fear in concrete life. In 100% of the children who have been diagnosed with Ollier and Ollier-Maffucci (or the even rarer special variants), the possibility can not be ruled out that at least one of the Enchondromas can mutate into a Chondrosarcoma. Because of the difficulty of determining the normality of growth externally (both by the doctor's or even the patient's eyes and by scanning the skeleton by the doctor's outer or topsided fist-bones), each child needs a monitoring program in which themselves and their parents can be assured that every anomaly is taken seriously. Limiting the number of examinations is justified only by setting the upper limits of radiological and nuclear exposure (mostly by medical standards, in part economically, too).

Also my own doctor said to me, at the end of the sixties: "If you survive the age of 16 without mutation, you will not have anything to do with the disease until the age of 45." However, over time, I forgot that he had added that I might then be confronted with a tumour: when my

shoulder began to hurt at the age of 44, I was convinced that I had become a patient of osteoarthritis. As we know in the group, there are also patients who survive the childhood with various surgeries, but without the transformation of an Enchondroma into a Chondrosarcoma, and are attacked by a mutation before 45 years old. The conventional statistical prognosis must therefore be revised: not only from 40 or 50 years there is an increased risk of dedifferentiation of an Enchondroma, but always earlier. Even though patient monitoring does not have to be as consistent as during childhood and adolescence, Ollier's patients need the ability to have a painful part of the body examined immediately – which they may not even know as an Enchondroma.

Now, should one even speak of a risk factor, that is, of a defined risk? – Yes. Human beings can not talk about happiness and the success of life differently than looking at the dead, but it has to be done in such a way that it only appears from a distance as far as possible. Every formulation has to be considered carefully, and it has to be kept in mind who is talking to whom. A child and its parents are quickly frightened and perceive the prognosis communicated by the doctor as a threat that the risk of death can occur at any time; conversely, it should relieve them in their everyday life when they learn that all encounters are just ordinary controls without assuming a hidden danger. The same problem with adult Olliers: when a person understands to be allowed to speak at any time, and when he or she knows where and to whom, there will be a different, more rational thinking on illness and life than if the patient would be told (in false confidence to prevailing scientific standards according to statistical probability) no mutation would be expected with him or her, because in this period too many mutations have already taken place, be it to other persons or also to him- or herself. What the doctor says to the patient is no ornament and are not accessory nice words, but one of his tasks and may be interspersed with positivistic, scientific content, here statistical values; but the doctor’s statements must never hide the existential contradiction that on the one hand the patient as well as the family environment, should not devote too much emotional attention and energy to the illness, but on the other hand should have the confidence to never be dropped medically. – According to my parents' story, the newly established Lucerne surgeon did not say farewell words after my first medical visit in 1962; we left the new office with its doctor bent over the first x-rays and a thick work of scientific medicine – he had yet to learn his language: the scientific, the medical-therapeutic and the communicative-advisory.

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