



Osteogenesis Imperfecta
Federation Europe

Newsletter

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Editorial

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Dear friends,

In this edition you'll find a report about what happened throughout OIFE's annual general meeting 2013. It also gives a little impression about OIFE itself.

We are very happy to present Marit's report about her individual experience with a Cochlea Implant because many people with OI do have hearing problems and might be interested in that topic.

All around the world there are people with OI. Unfortunately, many of them have to cope with very difficult living conditions as well. Some do ask OIFE for help because they are really desperate. As much as we'd like to, we are not in the position to give money. BUT we can share experience, connect with our helpful medical contacts in the OI-world and give advice. Additionally, we start a series about new OI initiatives in various countries, in this edition we start with Uganda.

Stefanie Wagner

OIFE AGM in Paris, 11th – 14th April 2013

By Stefanie Wagner

This year the French OI-Association (AOI) hosted the 17 delegates and guests from all over Europe who came to the OIFE AGM. AOI held both its national meeting and a meeting for medical professionals on the topic "Women with OI". On Saturday we all had a more festive evening to celebrate OIFE's 20th and AOI's 25th anniversary. Below our report about about OIFE's Annual General Meeting.

A different start of the AGM

Usually our AGMs start on Friday morning. The participants arrive one day earlier, on Thursdays, and have dinner together. This year we tried to make better use of our precious time together by having open-forum discussions. The outcomes of the very lively discussions about OIFE's future will determine the policy of OIFE during the next year.

20th anniversary of OIFE: a reason for celebration but also for change

Founded in 1993 by 7 countries, the OIFE now has 27 member organizations, mainly from Europe. Since its foundation, the tasks and projects of OIFE have multiplied, but the number of volunteers has not. They have a very heavy workload, which means that unfortunately many important tasks and

projects cannot be realised. In addition, OIFE does not receive any regular governmental or European funding, nor does it have regular donors. The realisation of very important projects such as the Topical Meetings (like the recent one in Lisbon 2012 on psychosocial aspects of OI) requires that OIFE puts a huge amount of time in fundraising FIRST. At some points OIFE was almost forced to cancel the TM because it would just have been too expensive for speakers and participants to attend! (unlike other organizations, OIFE is not in the position to pay for its speakers).

OIFE does not even have an office as such; the "headquarters" consist of a table and a private laptop, printer and telephone in a private apartment. The few active volunteers of OIFE's executive committee (most of whom work full-time in their "normal" lives) invest many extra hours per month (voluntarily!) for OIFE and it is very hard to find successors and replacements for them. Further hours are contributed by the secretary and few other volunteers.

So one of the main issues of this year's meeting was to find a solution for re-structuring OIFE.

Discussion points and outcomes of AGM in Paris

- presentation of preliminary results of the workshops (topics were all related to the current situation and the future of OIFE);
- agreement to find professional support for OIFE;
- presentation of Dutch OI foundation "Care4brittle bones" (see www.care4brittlebones.org) by Dagmar Mekking. Result: Care4brittlebones becomes new supporting OIFE member;
- discussion about inactive members / members that do not reply to emails;
- discussion about inactive delegates / replacement of long-term-delegates. general agreement that OIFE needs new, young, motivated delegates;
- presentation of RBU (Swedish member organization) by Asa Hedberg;
- due to the existence of new social media and the decrease of active participants: agreement to end the Making Friends Project;
- international youth weekends 2013 in Spain and 2014 in Germany;
- first International Adult meeting planned for 2015;
- planned: collection of national OI-literature (leaflets, brochures etc.) of OIFE members;
- planned: collection of data about national ongoing medical projects and treatment protocols



*From left to right: Åsa Hedberg, Sweden; Stefanie Wagner, Germany; Carina Svenheden, Sweden; Ute Wallentin, Germany; Taco van Welzenis, The Netherlands; Trond Gården, Norway; Rob van Welzenis, The Netherlands; Eero Nevalainen, Finland; Filip de Gruytere, Belgium; Lidy van Welzenis, The Netherlands; Vanja Živković, Switzerland; Céu Barreiros, Portugal
Missing on the photo: our delegates Maria Barbero, Spain and Laurette Paravano, France and our guests Meriem Benjelloun, Germany; Julia Pfister, Germany and Dagmar Mekking, The Netherlands*

What has been achieved since AGM in Lisbon 2012

Due to the short period between the two AGMs and vacation times as well as cases of illness, there has not been fulfilled very much, but still:

- thanks to Anna Rossi, our youth-coordinator, OIFE is now on Facebook;
- thanks to Taco van Welzenis for a document about OIFE to help members understand what OIFE does, what they can expect from it;
- another edition of the OIFE newsletter.

Silent rain and crackling paper

By Marit Heggelund, Norway

I am 57 years old and have used hearing aids since the age of 28. First a small "in the ear"- hearing aid in my right ear, and a couple of years later, in both ears. Gradually and almost unnoticeable my hearing worsened and I eventually had to change to "behind the ear"-hearing aids in both ears. Suddenly and without warning my capabilities to comprehend speech, especially in rooms with bad acoustics and background noise, went from bad to worse. I felt it happened very fast, but I didn't ponder on why it happened. I knew several persons who had been through inner ear prosthetic surgery with good results and wanted the doctors to consider this option. The doctor on Rikshospitalet University Hospital agreed to the surgery and I was put on a waiting list for the procedure. I waited and waited and waited for years. Nothing happened.

During this waiting period I had a femur fracture that would not heal and I spent months in hospital and rehabilitating institution. At this stage I had used Fosamax for a couple of years and was recommended to stop taking the drug until the fracture was fully healed.

When I finally came home, my patience regarding the inner ear surgery was non-existent and I contacted the doctor in Rikshospitalet University Hospital and got a new appointment. We agreed on advancing the surgery, I stressed my OI condition and referred to the Finnish thesis on OI and hearing, naturally

expecting that the doctor would update his knowledge on this subject before operating. Obviously he did not do so. Because after the surgery I became totally deaf ☹. Exactly what Kaija Kuurila-Svahn warned about in her thesis had happened. My doctor was not aware of the relatively large possibility for large amounts of fluid gathering in my middle ear. He felt really sorry and promised me CI-surgery (cochlea implant) if I wanted it.

I took all the tests, but found out I was too fed up with hospitals after spending the last eight months there, so I wanted to wait.

By this time I was on my feet again and started taking Fosamax again. Again my hearing loss quickly grew even worse and I started thinking that there might be a connection between the intake for Fosamax and my continuing hearing loss. Of course I can't claim that there is such a connection. The events may be coincidental. But I mentioned it to Kaisa and asked if there was seen such a connection. She denied it but at the same time said that there were thoughts on the issue and that a study might be done. Enough about that.

After four years of waiting I started thinking about CI again and mentioned it to the doctors at

Rikshospitalet University Hospital. The doctor that had performed the inner ear prosthetic surgery and promised me CI was unfortunately dead, and I was told that my hearing aid on the other ear was sufficient. CI was out of the question for me. I was of course in despair as I experienced it to be very problematic to be in a conversation with more than three persons present. I could never hear where the sound came from and verbal communication was difficult. My back is tense and painful because of spinal curvatures. My efforts to hear have not eased these tensions.

At my next appointment at Rikshospitalet University Hospital, I raised the question again. I argued among others that I knew with 100% certainty that my hearing would worsen and I therefore wanted CI before I became deaf on both ears. I also reminded them of the promise given to me and said I felt it to be strange that such a promise was attached to a person and not the institution he represented. This time it was another doctor than last time and she listened to my arguments and said she would consider the case once more. My joy was immense when I was told that my name was added to the waiting list.

In May 2012 I finally undertook the surgery. It was performed in the intervention-ward and I felt well cared for. Among other the anesthetic procedure was special because of my curved spine and squashed lungs. The surgery was successful in the sense that it went well. But the doctor said there was a possibility that I would not hear some of the lower tones. I left hospital the day after surgery, in relatively good shape. In comparison I spent several days in hospital after the failed prosthetic surgery. Now I had some exciting weeks ahead of me. After a surgery like this one will have to wait for some time (max 6 weeks) before switching on the sound. This is because the device that is implanted needs some time to "settle down". I had giant butterflies in my stomach when the day of activation finally came. I knew that the sound would be mechanic and was not at all surprised when the audio physician sounded like Donald Duck on helium☺.

The first week I went back for checkups twice every day to adjust the sound. Slowly it became better. But even today, half a year after the surgery, the voices are still a bit squeaky. But now I can distinguish male and female voices. And when I combine hearing aids and CI it sounds quite natural! And I can actual hear quite well! One of the first days afterwards I sat on my sofa hearing this strange buzzing noise. I could not comprehend what it was. But then I discovered that it rained outside and the noise was the trickling rain. My tears started trickling too as I realized that this was a sound I hadn't heard for at least 15 years, maybe even longer.

I attended a national OI-gathering last autumn and people told me that they had noticed how much easier it was to chat with me, even with background noise. At the movies I joyfully told my friend that I could hear the crackling of chocolate paper. "The rest of us finds it quite annoying" she replied in dry wit. When you have spent a long time in an all too quiet world, even the most annoying sound turns into music.

In Norway, after surgery like this, one will have 25 sessions with an audio pedagogue to learn to hear again. She and I agreed that since my hearing was so good, I would not need all of the sessions and we terminated after the 19th session. But I can of course go back if I want to.

My life has become much simpler. I can hear better in all situations and it feels so good. I will never recover the hearing I had as a child, but I don't need that. I have stopped using subtitles when I watch TV and I don't have to use extra equipment in small meetings. And a marvelous side effect which I haven't figured out until recently is that my back hurts less than before. I think it is because I no longer have to strain my body to hear what people are saying. And of course my spirit has been lifted.

I have joined a CI-group on Facebook and have read about persons with negative experiences. I think it is important to mention that not everyone is as happy with the result as I am. As already said before, my hearing is not normal and adequate in all situations. Recently I visited the theater and was placed in the back of the hall. Unfortunately I could hear almost nothing of the dialogue. But the most important thing for me is how it works in everyday life. And there it is better than in years.

If anyone wants more details, feel free to contact me on e-mail: maheggel@tele2.no.

Marit Heggelund

OI in Uganda

This is the story of Sula, who lives with his family in Uganda. All four siblings are affected by OI. They live with their mother in a small house about 25 kilometres from Kampala (the capital of Uganda). Their living conditions are very difficult for several reasons; the lack of accessibility of both their own house and local public places, the lack of equipment and wheelchairs that really fit them (Sula says he has had several fractures just because of his non-fitting wheelchair), lack of medical care (even the largest hospital was not able to give them information about OI), they cannot go to school and last but not least, the people in his village have a "negative attitude" towards people with a disability and their mothers. This is because it is always the mother who is blamed for having sick children; also the fathers often leave their families. Sula says that the family has already been threatened by others with the worst sort of harm. He has tried for eight years now to get support from the government and via the media, but without any positive results. Without doubt, the story of Sula and his siblings is just one of many similar ones of people with OI in Uganda. He has been asking us for help for his family but he is also willing to start an OI-group there.

Recently a special group "OI support Africa" was started (by Jo Ragen from Australia) on facebook and many so far unknown OI people have already joined.

So we ask our readers: do you have any idea how to improve the situation of Sula and his siblings? Do you have contacts to Uganda? Or know OI-people in or from other African countries? Please contact office@oife.org . Thank you very much!



Sula with two of his siblings; missing in the photo is the 15 year old sister who cannot sit upright

Dates to remember

2013, June 7th – 9th Switzerland: SVOI Symposium

2013, Nov 8th – 10th Spain: International youth weekend – contact: youth-coordinator@oife.org

2013, Nov 8th – 10th Spain: AHUCE OI Congress near Tarragona – guests welcome!

2014, June 19th – 22nd Germany: DOIG Meeting in Duderstadt – guests welcome!

2014, Aug 1st – 4th USA: National OIF Conference in Indianapolis

2014, Sept 18th – 21st Finland: OIFE Annual General Meeting + 35 years Finnish OI Society

2014, Oct 11th – 14th USA: International Scientific OI-Conference in Wilmington, Delaware