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A Norwegian population based study of adults with Osteogenesis imperfecta

TRS is a national resource centre that offers services to seven diagnostic groups of congenital disorders. Osteogenesis imperfecta is one of them. The centre is a part of Sunnaas Rehabilitation Hospital, and is located just outside Oslo, Norway.

The centre works to increase knowledge about these disorders and offers guidance and counselling on medical, psychological, social and educational issues related to them.

During the clinical work at our centre, we get a lot of questions from adults with OI, concerning to the process of getting older with this disorder. They want to know about the prognosis of the disorder; age-related changes in organs like lungs, heart, blood vessels etc., development of osteoporosis, pain and medical treatment? They also have a lot of questions about work and social issues. However, it is very difficult to find the answers to all these questions in the literature. Therefore we decided to make a study on the adult population, to try to find some answers.

Most of the people with OI in Norway are registered at TRS. That made it possible for us to make a population based study. All the adults from 25 years of age and upon were invited to participate in this study. We started in 2003, and all the investigations are now done.

The purpose of this study was to describe the medical changes according to fractures, bone mass, deformities, joints, lungs, heart, large blood vessels and different oral problems. We did also want to know how the adult persons with OI live their daily lives by asking them about function, work and leisure time.

We hope that this study will give us some information about the natural (clinical) history on OI, and increase the knowledge about how to prevent complications and different challenges related to age.

We have started to analyse the data, and I hope that they will be presented in different international journals during 2007 and 2008. So far it has been a real pleasure to work with this OI population.

*Lena Lande Wekre, MD
TRS National Resource Centre for Rare Disorders*



The OIFE

- *Represents their members on a European level
 - *Presents the problems and needs of people with OI to national and international organizations
 - *Collects and publishes information about OI
 - *Promotes research on all aspects of OI
 - *Supports member-societies by the exchange of information and experiences
- www.oife.org

Fighting for free drug access (March 07) - Osteogenesis imperfecta patients in Romania

(complete article to read: http://www.eurordis.org/article.php3?id_article=1373)

Off-label use, availability, and reimbursement of drugs are three issues commonly encountered by rare disease patients. In the case of OI patients (Osteogenesis Imperfecta), these problems collide to create an even greater hurdle, of which Romania provides a telling illustration.

(...)

In Romania, until the beginning of 2006, Pamidronate (Aredia®) could be prescribed by any family physician; it was available in pharmacies, free of charge, and administered to patients by infusion in hospitals. However, in March 2006, the NHIH (National House of Health Insurances, Romanian Ministry for Health) removed Aredia® from the free drug list and decided that it would only be available in hospitals. 'When we asked authorities why they took this action,' says Florin Dananau, father of a young girl suffering from OI, 'they said it was because the drug could only be administered by infusion in hospitals, and that no one should get the drug outside healthcare facilities.' The cost of the drug is so high that patients from Romania, where the average monthly salary is 200 euros, could not afford it. In Romania, until

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(...)

Ute Wallentin, the President of OIFE, had turned to Eurordis for help. Since the 1999 adoption of the EU Regulation on Orphan Medicinal Products, Eurordis has been advocating for availability and affordability of orphan drugs for all rare disease patients in Europe; it conducts a regular survey to assess and compare availability of orphan drugs to patients in the EU, which shows that the situation of patients in new and candidate Member States is particularly difficult. Eurordis wrote a letter to the Romanian Minister for Health, asking that Aredia® be made available to all patients and be reimbursed.

However, to this day, the situation of OI patients in Romania remains worrisome. 'We don't know if the BHIH will provide the necessary funds in 2007,' says Florin Dananau. 'There are also very few hospitals in Romania that have both skilled personnel and the drug to administer it to patients. Without mentioning the general lack of knowledge on OI!' 'But the situation of OI patients is just as bad in other countries,' says Ute Wallentin. Towards the end of 2006 we were saddened to hear that OI adults from Belgium had lost access to free treatments. And OI children in Germany are in danger of losing access to treatments too if the planned budget cuts are put into effect. Countries like Portugal or France are also facing a difficult situation!

(...)

Interview: Life and work of a woman with OI in Zimbabwe

Ute Barschdorf emigrated from Germany to Africa. Together with her daughter and her partner she is living in Zimbabwe. Both mother and daughter are affected by Osteogenesis imperfecta Type I.

OIFE: Since when do you live in Zimbabwe and what was the reason why you said "good-bye" to Germany?

Ute: I'm living in Zimbabwe for 3 ½ years now, and before that time I lived for almost 10 years in Ethiopia. I went to Ethiopia together with my former husband. He was working there with the German actor Karlheinz Boehm for his welfare organization "Menschen fuer Menschen" and built up a training centre there.

During those years I volunteered for different organisations as well: I worked with blind women; they made knitwear and we sold it at a bazaar. I also helped people suffering from leprosy, orphans and HIV-infected persons: I was portioning out food in areas where drought and hunger predominated. I really loved to do this work - even without being paid for. For: when a child smiles at you because it is no longer hungry, or when a child that was not able to walk because of malnutrition starts walking again after you infused food every two hours and massaged his legs – those are moments that nobody can replace with money.

But I also made some very sad experiences in Ethiopia: for seven years I raised two orphans (they were one week old when they came to our family) but they were not allowed to leave with us to Zimbabwe. Up to now this burdens me a lot.

OIFE: The political and economical situation is extremely difficult and unstable, life expectancy is meanwhile 35 years. What does all this mean for your everyday life?

Ute: Due to the economical situation it happens quite often that we can't get any sugar for several weeks and even for months there is no or only temporarily fuel. But I think this is only a problem for people that come "right away out of civilization". For me this doesn't matter that much, even if there is no fresh water or electricity for a couple of days – you are getting used to it. So you have to improvise.

Well, our oldest gardener is 84 years old! But it is sadly true, many many people die of AIDS

OIFE: How is the climate and does it have any impact on OI?

Ute: The climate in Zimbabwe is very good. But, I must admit, in Ethiopia it was even better for people with OI because the country is located higher and in general it was dryer. When I lived in Germany I continuously suffered from back pain – here in Zimbabwe I only sometimes have pain and the pain isn't that strong. In Ethiopia I didn't have any pain at all! My daughter Stefanie hasn't used neither her wheelchair nor her crutches since we left Germany. She can walk for miles!

(to be continued...)

Blackboard:

"Hello! I have OI type 1. My biggest problem is my hearing loss. It has been determined that I am a candidate for a cochlear implant but my doctor told me he has never had patients with OI. He also told me it's very rare for someone with OI to go completely deaf like me. Is there

anyone like me here, with total deafness? I would be very happy to hear from anyone! My
eMail-address: yana.domuschieva@gmail.com
Thanks a lot – Yana”