



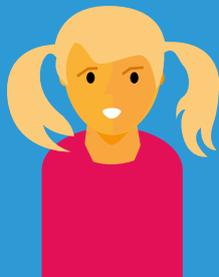
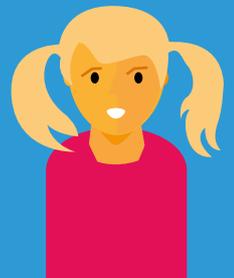
Living with OI



Insight into the psychosocial consequences of Osteogenesis Imperfecta (OI)



drs. C.J.C. Kalsbeek, drs. V.J.D. Platteel, mr. M.M. Wewer



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Introduction

Insight into living with Osteogenesis Imperfecta (OI)

If you have the brittle bones disease OI, you can encounter questions and challenges in every stage of your life. These are not only related to your physical condition, but to different aspects of your life. About how you feel, what emotions you have. They can be about school, work, sports; so, about participation. But also about social contacts such as friends, family etc. Everyone will eventually have to find a way to deal with these obstacles. This is regardless of whether you have a mild or a heavy form of OI. But you don't have to find out everything on your own; there is help. In your area, at the patient association VOI or at the expertise centers. From research that has been done, there is now a good overview of the consequences of living with OI. This is the public version of the research report.

#LevenmetOI (Living with OI) is the result of the collaboration project of the OI expertise centers [Het WKZ](#) and [Isala](#), the [OI Vereniging](#) and [Care4brittlebones](#) together with [Curias](#) within the programme Psychosocial consequences of major somatic diseases by [Zorginstituut Nederland](#).

Research

For the first time, the nature and extent of the psychosocial consequences of OI have been systematically mapped out. This was based on quantitative research, the survey and qualitative research among the target group.

The survey response was high, more than 20% to 30% of the total population.

20-30%
Response rate

252
People completed
the survey

of wich
164
adults with OI
107 women | 57 men

of wich
8
children with OI

42
Parents completed the survey on their
own behalf as a carer.

38
Parents completed the survey on
behalf their child
(for 27 children younger than 12 years and
for 11 children between 13 and 18 years)

The research questions:

- What is the extent of the psychosocial symptoms in people with OI?
- What is the nature of the psychosocial symptoms of people with OI?
- Is the care that the two expertise centers offer for psychosocial complaints also the care that patients with OI need?

Osteogenesis Imperfecta (OI)

Osteogenesis Imperfecta (OI) or 'brittle bones disease' is a rare, serious disorder of the connective tissue. The most striking feature of OI is the great fragility of the bones. The disease is therefore called Osteogenesis Imperfecta, which literally means 'imperfect bone production'. Other characteristics are slow growth, hearing loss, blue sclera and deformation of the bones.

The symptoms vary greatly from person to person and from one stage of life to the next. There are 5 different types of OI, which indicate the severity of the condition. OI cannot be cured. There is no remedy for OI. However, there are ways to manage the symptoms. It is a rare condition. Of every 15,000 to 20,000 people there is one person with OI, so in the Netherlands there are 800 to 1,200 people with OI. Every year, 10 to 15 children are born with OI. OI is equally common among men and women. OI is congenital. OI is caused by a small error (mutation) in the hereditary material (DNA). Someone with OI may have inherited it from their parents, but sometimes people without OI have a child with OI.

See also: <https://youtu.be/HZ2JsZ8r6UQ>

In the Netherlands there are two hospitals that are specialized in OI. These are expertise centers. The Wilhelmina Children's Hospital (WKZ) in Utrecht focuses on the treatment and care of children. The OI Expertise Center of the Isala Clinic in Zwolle focuses on adults with OI.

The VOI is an association for people with OI and their immediate environment. The VOI is active in the field of information provision, contact with fellow-sufferers and advocacy.

Care4BrittleBones is an organization that strives for a better quality of life for people with OI by means of research. It is an internationally oriented foundation with roots in the Netherlands. Founded in 2012 by family and friends of a child with OI (Dagmar and Peter Mekking and Jacqueline Hornman), the foundation has quickly grown into the largest fundraising organization for OI research in Europe.

At Curias it's all about experiential knowledge. Gathering this knowledge and using it effectively and efficiently requires a professional approach. Curias combines research with concrete advice and implementation. Curias carries out qualitative research and supervises improvement projects in healthcare.

In the Netherlands
→ ±1000
people with OI

Key findings and conclusions

Scope

More psychosocial symptoms

Psychosocial symptoms in people with OI are more common than are seen on average in other chronic conditions. For people with OI this is 40% to 60% compared to 35% for other chronic conditions.

Light form of OI

Psychosocial problems are type-independent. The psychosocial problems can also have a major effect on minor forms of OI. People with a light form of OI indicate more often that they experience obstacles in sports, exercise and work than people with a heavy form of OI.

How is that possible?

- The environment notices less that you have OI, and therefore expects full participation from you; there is less understanding.
- For people with a serious form of OI, it does not always feel realistic to have equal participation. Therefore, it may be that they accept sooner that it's not possible for them and therefore no longer experience it as a limitation.

Watch out!

By working hard on the image that you are just like all other people without OI, by compensating, there is a serious risk of overload that will manifest itself sooner or later.

Psychosocial symptoms

In people with OI:

40%-60%

In people with other chronic conditions:

35%

Nature

Age stage

In different stages of life, people with OI (in a heavy or light form) encounter psychosocial challenges, which require adjustment time and time again. They can be accompanied by (as yet unprocessed or new) emotions.

This concerns the following five age categories:

- Children (and their parents) up to primary school age (about 12 years)
- Young people (high school) of about 13-18 years and their parents
- (Young) adults (18-35 years)
- Adults (35-50 years)
- Older people (50+)



For each age group, specific obstacles are encountered at this stage of life. During the research, the solutions found were also divided into life stages.

Parents as carers

In addition to the five age categories, parents who are carers also experience obstacles.

Children

TOP 5

of activities in which children with OI, according to their parents, can experience obstacles:

63,2%

at school/studying

60,2%

when exercising or moving

39,5%

meeting up with friends

28,9%

at home

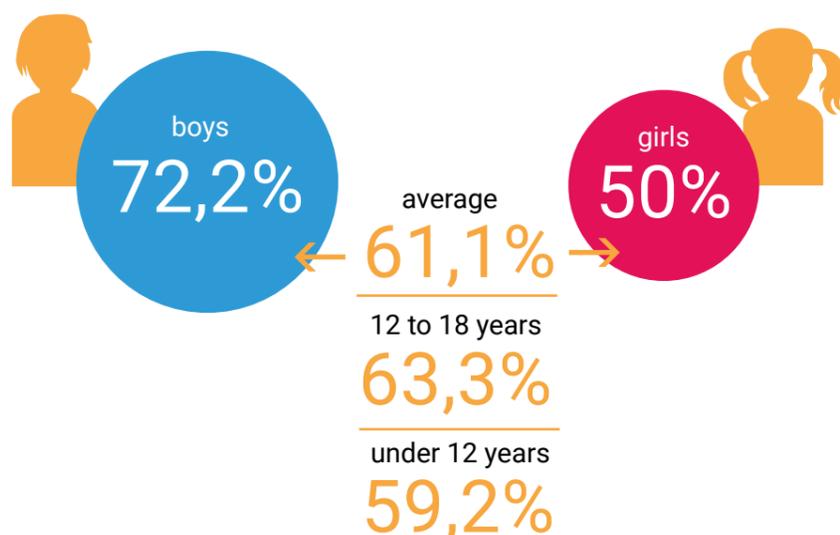
18,4%

doing hobbies

The main causes of obstacles at **school** are identified as:

- Fear of breaking bones
- Problems or limitations with movement

The obstacle to **exercising and moving** is mainly indicated by:



"You have a disability, but if you also act as if you have a disability, you'll feel completely restricted. So even if you have a severe form, of course you have to pay attention, but it's not that you have to be reminded every moment of your life that you have OI".



What the children say themselves about their emotions:

- Two thirds of the children who completed the survey themselves are sometimes angry or very angry because they have OI.
- Most children sometimes feel sad because they have OI, three of them feel very sad, and there is no one who never feels sad.
- Half of the children with OI sometimes feel lonely because they have OI.
- None of the children with OI feel guilty towards others because they have OI.
- Almost all children with OI say they cannot do everything they would like because they have OI.
- Two of the eight children experienced shame about their own body.

Adults

TOP 5

of activities in which adults with OI can experience obstacles:

67%

when exercising or moving

40%

at work

32%

at home

30%

meeting with friends

20%

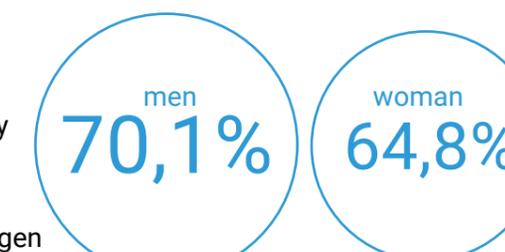
doing hobbies

When exercising or moving

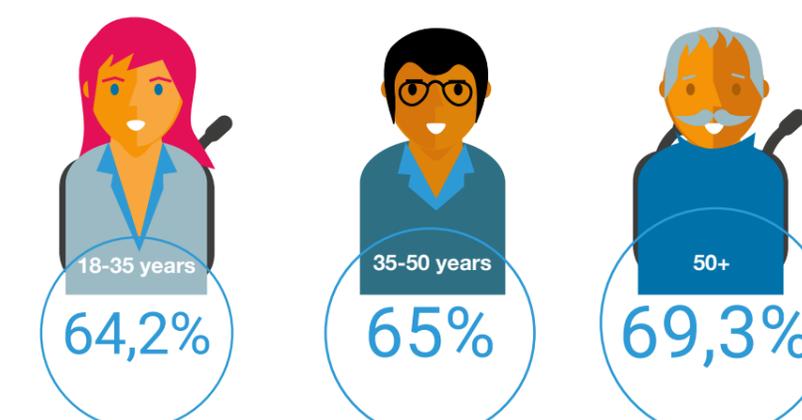
In the group that have a **light form** of OI, it is indicated that there are obstacles in sports and exercise more often than in the group with the **heavy form**



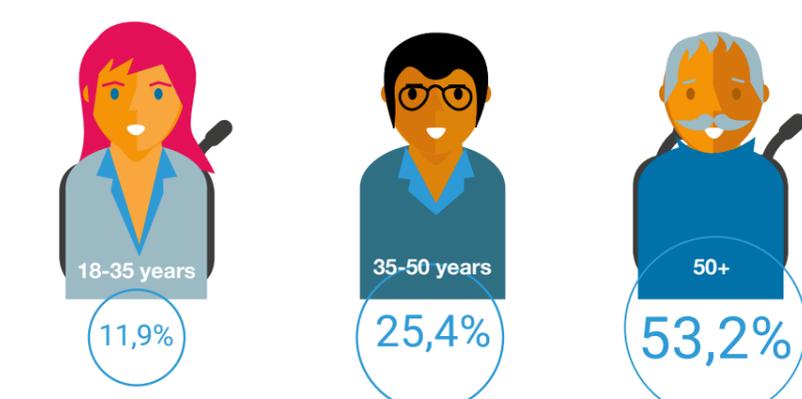
The percentage of **men and woman** that say they experience obstacles when exercising and moving



The percentage **in each age category** that say they experience obstacles when exercising and moving



It is striking that it's people above the age of 50 with OI in particular that indicate they experience obstacles **at home**

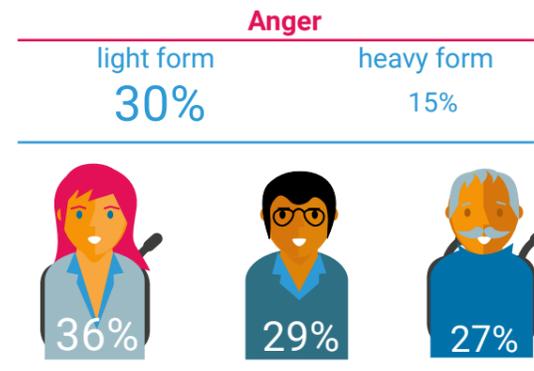


Adults continued

Emotions

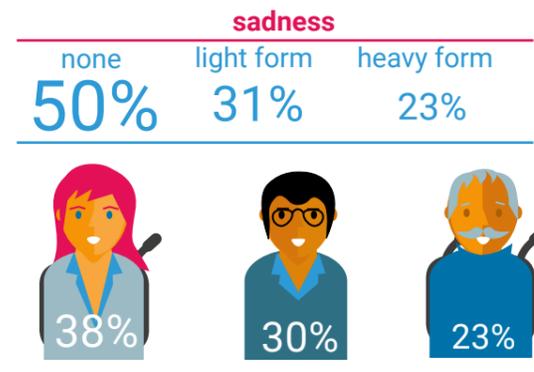
→ Anger

30% of the people with OI feels angry or very angry because they have OI. It is striking that people with a light type of OI suffer from this more than people with a heavy type of OI (30% versus 15%). The anger appears to diminish with age. 36% of the young adults versus 29% and 27% of the adults and those over fifty felt angry or very angry because of OI.



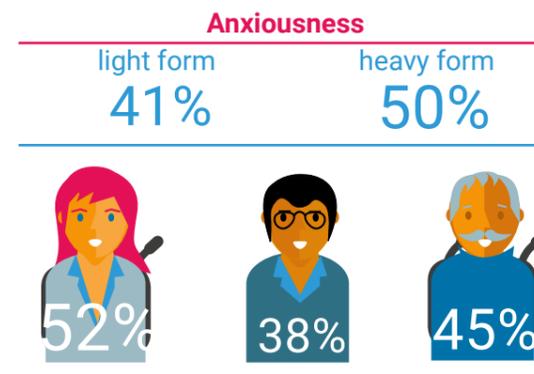
→ Sadness

50% of the people with OI don't (or never) feel sad because of OI. While 30% of the people with OI often or very often feel sad about it. It is striking that people with a light type of OI are sadder than people with a heavy type of OI, 31% versus 23%. The younger, the sadder. Young adults with OI feel sadder than adults and those over fifty, 38% versus 30% and 23%.



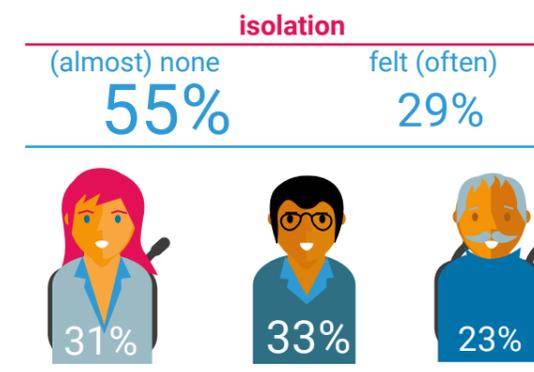
→ Anxiousness

45% of the people with OI often or very often feel anxious. People with the heavy type of OI feel more anxious than those with the type of light OI, 50% versus 41%. Young adults and those over fifty feel more anxious than adults, 52% and 45% versus 38%.



→ Isolation

55% of the people with OI don't or never feel isolated. 29% of the people with OI often or very often feel isolated because of OI. It is striking that people above fifty feel less isolated than young adults or adults, 23% versus 31% and 33%.



Parents as carers

TOP 5
of activities in which
parents experience
obstacles as carers:

50%
At home

36%
At work

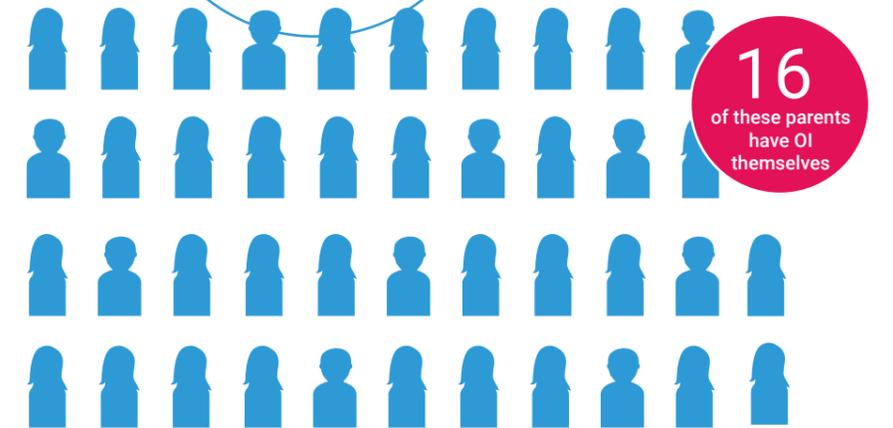
31%
Child care

31%
None

19%
During sport

42

Parents of children with
OI filled in the survey
32 woman | 10 men



Emotions

Emotions that carers struggle with:



31% of carers have a
different approach to caring
for their child with OI than
their partner.



Beyond the ages

Future prospects:

The respondents from the various phases of life are more or less concerned with the future in their own, individual and personal way. The teenagers have big plans, the young adults give substance to their lives and the over-50s are worried about getting older. The realization that not all dreams have come true or will come true is a painful fact for some respondents. In particular, having or not having children is accompanied by strong emotions

A 50-plusser with type OI I said:

"I'm a little worried about care, though. Because however it all declines, I still live with a stairlift, one floor up. And I find it complicated. Do I have to go and live somewhere else while I still live here to my liking? And in a pleasant neighborhood with benches everywhere, where you can sit. Relaxed cozy, not so much neighborly contact."

A 50-plusser with OI type II:

"I was divorced and then I met another partner and that was really my soul mate. We were together for years. Yet this relationship did not last. I've been so upset about that. But OI has never been an issue in relationships. I've always said, do you want to climb mountains, get up early, ski, just do it. And that people sometimes say, that's hard, isn't it, such a handicapped woman. And then he said, handicapped woman? Where? I don't see her like that, really. And I think she has a harder time with me than I have with her."

Advice from people with OI

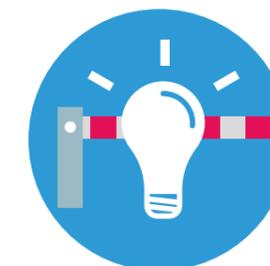
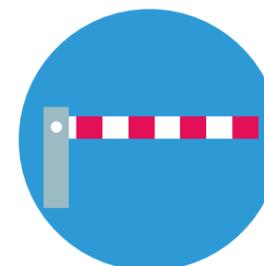
Advice

There is (as yet) relatively little you can do about the OI itself. You can do something about the psychosocial aspects.

- Prevent problems by knowing yourself well. Knowledge of the psychosocial consequences of the next phase of life can prevent you from getting stuck is important. Find your own way to YOUR happiness!
- Ask for help when you run into things. You can ask friends, other people from the OI world, family or care providers. Learn to put into words what you are going through, so that you can formulate a good request for help in time.
- There is room within the care sector for better guidance. Particularly in the field of screening and its follow-up.

A young adult with type OI III said:

"I learned from my psychologist that every 7 years you enter a new phase of your life and that every phase of your life involves changes. When you're 7 years old you're a child, and then you're 14 years old, then you're 21, then you're expected to do other things and by the age of 28 you're expected to settle down, socially speaking. But I'm past that age now. So there aren't really any more such changes coming up now. I know how to anticipate it better. Previously I didn't, and then I had a continuous rollercoaster that overwhelmed me. That's what I want to be for now. So I now ask for help."



Care

What is psychosocial care?

Psychosocial care is a collective term; it consists of psychosocial support and psychological care. Psychosocial support is mainly focused on emotional and practical support for (relatively) simple problems of a psychological and social nature. For the small group where psychological complaints such as anxiety, depression or psychological trauma occur, there is specialist care by a psychologist or psychiatrist.

Asking for help

It seems that people do not know that they can get psychosocial help or don't think that it exists. Those people who have asked for help have generally received it and are satisfied with it. So it is important that help is requested (correctly).

Many respondents (n=80) received psychosocial care. Some think that this should be offered more and that the care sector should pay as much attention to psychosocial components as to the physical components. Some respondents seek support from a mentor, parent or partner. Sometimes they have to deal with conflicting advice. However, not all respondents have or are open to a request for help.

Children

More than half of the children's parents have at one time sought psychosocial help for the children. Almost two thirds are satisfied with the help they have received. Of the children who did not seek help, three quarters also think that a care provider cannot help them. 59% of the parents think that the expertise center should pay more attention to the psychosocial complaints of their children.

A teenager said:

"If I go to the doctor, I only talk about the breaks with this doctor. Physicians never ask how it's going at school or at home."

Adults

Of the adults with OI (n=164), about a third has ever sought psychosocial support for the obstacles they encounter. Of the more than half who did not seek help, only 18% think that a care provider can offer psychosocial help. Only 38% of the adults have ever sought psychosocial help for the obstacles they encounter. Three quarters of them are satisfied with the help they have received. Of those who have not sought help, only 18% think that a care provider can help them with this. 45% think that the expertise center should pay more attention to psychosocial complaints.

164 adults



42 parents as carers



Parents as carers

Of the parents as carers who did not seek help, 63% also think that a care provider cannot help them with this. 44% of the carers think that the expertise center should pay more attention to the psychosocial symptoms of them as parents.

Medical care

People with OI often use medical care. Some respondents spent a large part of their youth in hospital because of fractures. Many of the older respondents suffered some form of (unprocessed) sadness, trauma or even a post-traumatic stress disorder. These experiences from their youth have an impact on how they make use of care today (avoidance of care provision, fear of hospitalization). In the case of children, more than 50% of the children are reluctant to visit the hospital.

36 children



Environment

Environment is important

OI creates physical barriers that affect the psychosocial domain, in which incomprehension, lack of understanding, ignorance and inadequacy of the environment evoke feelings of anger, sadness and fear, leading to feelings of isolation and doubts about the future. Good information provision to the environment as well as to supervisors within schools, employers, etc. could reduce the number of obstacles experienced

A young adult with OI type III said:

"I was raised very much with the thought that you're no different. So it was very difficult for me to say that I couldn't do something, working full-time, for example because of my OI. And that I was different, because that's not how I was raised."

Parental upbringing

The upbringing style of the parents of children with OI has an impact on the lives of these children as adults. The respondents from the study, from all age groups, talk about their upbringing, which in some cases was too strict, in other cases too protective, patronizing, or that it was a well-balanced compromise. That the parents prepare them for the upcoming obstacles and support them in these. The relationship with parents, siblings and partners seems to play a major role in the development of coping styles. Some respondents were strict with themselves or had feelings of guilt. Family members can also be a great source of support for the person with OI.

A young adult with OI type I said:

"My mother was really always extremely afraid that I would fall ever since I had my first bone fracture. And from the moment I was actually born, she had been very worried. She said, 'the child is not well'. And my doctor said, 'you don't have to worry, it's all right'. I only walked when I was already over two. And I was always a slider, I didn't crawl. I always sprained my ankles and at one point I had all sorts of fractures. My mother was so worried that from an early age I actually became super strong. Everyone around me was sympathetic, but actually out of pity, they all cried. And so I didn't cry and I was strong."

Background

Who and Why this research

Collaboration

With the project 'Psychosocial care for people with Osteogenesis Imperfecta', Stichting Care4BrittleBones joins forces with the two national expertise centers and the patient association (VOI) to map out the psychosocial aspects of OI, to make them part of the consultation process, and to be able to provide better care. Together they want to map out the psychosocial care needs of the patient and his or her next of kin, provide information about psychosocial consequences and find out if, and how, the existing care can be improved. This can be done, for example, by screening so that patients with OI in the different phases of life are able to limit these psychosocial consequences or "learn to live better" with them. The research and implementation of the project were carried out in collaboration with Curias.

particularly for the continuation of the project, for the development of various products and their application in practice

Positive health

The project is based on Machteld Huber's positive health model, which defines health as a state of physical, mental and social well-being – and not just the absence of disease or other physical defects – and the ability of people to adapt and have their own direction, in the light of physical, emotional and social challenges of life. The domains used in the model were used in the development of the survey. With the exception of the domain of meaning.

How was research done?

The survey was conducted via the VOI and the expertise centers. The results of the survey were analyzed, and four focus groups and three in-depth interviews were conducted to deepen the results of the survey. The results of the survey and the analysis of the focus groups and interviews are presented in a report. This report was discussed by the consultative group of healthcare providers and experience experts. They reflected on the results and interpreted them. The most important findings and conclusions were jointly sought in the discourse,



Bodily functions



mental wellbeing



meaning



quality of life



social participation



daily functioning

(Source: Institute for Positive Health, Machteld Huber)

Appendix I

Objective of the project

The research took place within the framework of the project 'Psychosocial care in people with Osteogenesis Imperfecta'. The project has four objectives:

1. The project contributes to clarifying the nature and extent of psychosocial problems by mapping out the symptoms and making them discussable. In doing so, the centers of expertise want to clarify whether the care they provide is also the care that patients need.
2. Raise awareness about the fact that OI can also have psychosocial consequences.
3. The project makes the provision of formal and informal care more accessible and more aligned, by making information available to everyone dealing with an OI patient (including regional care). A sub-objective is to strengthen the function of the expertise centers and the patient association collectively.
4. The aim is also to be able to provide better care in the expertise centers, by implementing screening instruments (better), so that problems can be identified quickly.

Part of Zorg Instituut Nederland (ZIN) program

Care4BrittleBones, together with the expertise centers and VOI, received a subsidy for this project from the Dutch institute for healthcare, Zorginstituut Nederland (ZIN). In 2017, the healthcare institute developed a special subsidy program to limit or prevent the psychosocial consequences of a major somatic disorder.

The annual theme (2017) of the subsidy scheme 'Transparency about the quality of care' is 'psychosocial consequences of major somatic disorders'. Having a somatic disorder can have major psychosocial consequences for the choices you make in life.

Vision for the future

The ultimate goal of the project is: By 2020, every child, (parent of children), adolescent, adult and elderly person with OI is aware of the different psychosocial consequences that can occur with OI and has access to appropriate care. Formal and informal care in this area has been sustainably altered/embedded.

Core team

Concerned parties are represented in the core team (in alphabetical order):

Arjan Harsevoort	nursing specialist Isala expertise center for adults with OI
Cecilia Kalsbeek	project manager Curias
Dagmar Mekking	project leader Care4BrittleBones
Iris van Wijk	Pediatric rehabilitation physician /researcher, WKZ expertise center OI
Bea Zoer	medical biologist, on behalf of patient organization VOI

Supported by *Margit Wewer*, researcher Curias

Consultative group

The consultative group consists of a number of people with OI (including a young person, an elderly person, a parent of a child with OI and an adult with OI) and a number of care providers with a psychology or social work background. Together they look at the results of the research and think about the different activities and products that we will develop. In this way, we ensure that people with OI remain involved in everything we undertake in this project and that it will therefore be well aligned with what is important for people with OI.

Participants of the consultative group

(in alphabetical order)

Danielle van der Grijn	Ergotherapist Isala expertise center OI
Danielle Guillaume	Orthopedagogue/ healthcare psychologist De Hoogstraat Revalidatie
Arjan Harsevoort	Nursing specialist Isala expertise center OI
Cecilia Kalsbeek	Project manager, Curias
Dagmar Mekking	Director Stichting Care4brittlebones, project leader
Jet Strijker-Kersten	Head of subsection Medical Psychology and Social work, Children's division, WKZ
Iris van Wijk	Pediatric rehabilitation physician /researcher, WKZ expertise center OI
Margit Wewer	Senior adviser/researcher Curias
Rik Withaar	Clinical psychologist-psychotherapist, RVEvz/ Head of Medical Psychology, Isala
Bea Zoer	Medical biologist, on behalf of patient association VOI
Stefan Buitenhuis	Experience expert
Chantal van Haaren	Experience expert / psychologist
Rebecca Meijer	Experience expert
René van der Rijst	Experience expert
Henk Slot	Experience expert

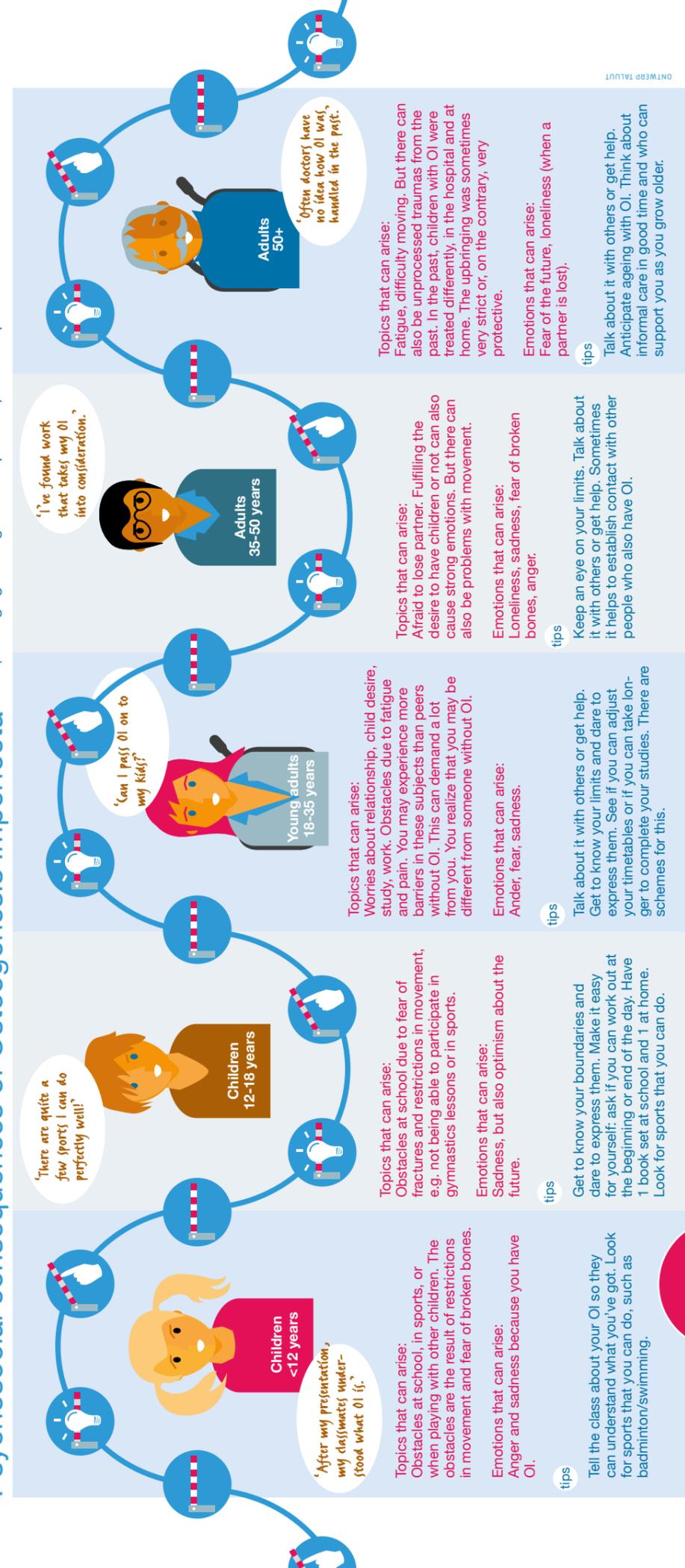
Appendix II

The infographic **Living with OI** Psychosocial consequences of Osteogenesis Imperfecta.

Living with OI

Psychosocial consequences of Osteogenesis Imperfecta

Even if you have OI, you can face questions and challenges at every stage of your life. Everyone has to find a way to deal with obstacles. It doesn't matter whether you have a mild or a heavy form of OI. You don't have to find out everything on your own, feel free to ask for help. In your environment, at the VOI (Vereniging Osteogenesis Imperfecta) or the expertise centers.



You don't have to do it alone

You can ask for help from: → Family/friends → Mentor at school → VOI → Expertise centres OI → Social worker → General practitioner → Mental health nurse → Occupational therapist → Psychologist → Mental health carer

Any questions? Go to www.oivereniging.nl

55% <18 years heeft hulp gezocht has sought help for psychosocial complaints

38% >18 years has sought help for psychosocial complaints

2018 Survey among people with OI **252** participants



Even if you have OI, you can face questions and challenges at every stage of your life. Everyone has to find a way to deal with obstacles. It doesn't matter whether you have a mild or a heavy form of OI. You don't have to find out everything on your own, feel free to ask for help. In your area, at the VOI (Vereniging Osteogenesis Imperfecta) or the expertise centers.

