

## Introduction

Shriners Hospitals for Children - Canada strives to transform children's lives by providing exceptional health care guided by high-quality, innovative research in a patient and family-centered environment. At Shriners Hospital for Children - Canada, we are committed to sharing our research with our patients and families. Together, we seek to discover new knowledge in Osteogenesis Imperfecta (OI) to improve their quality of care and quality of life. Understanding the current knowledge on quality of life will guide future research leading to improved care.

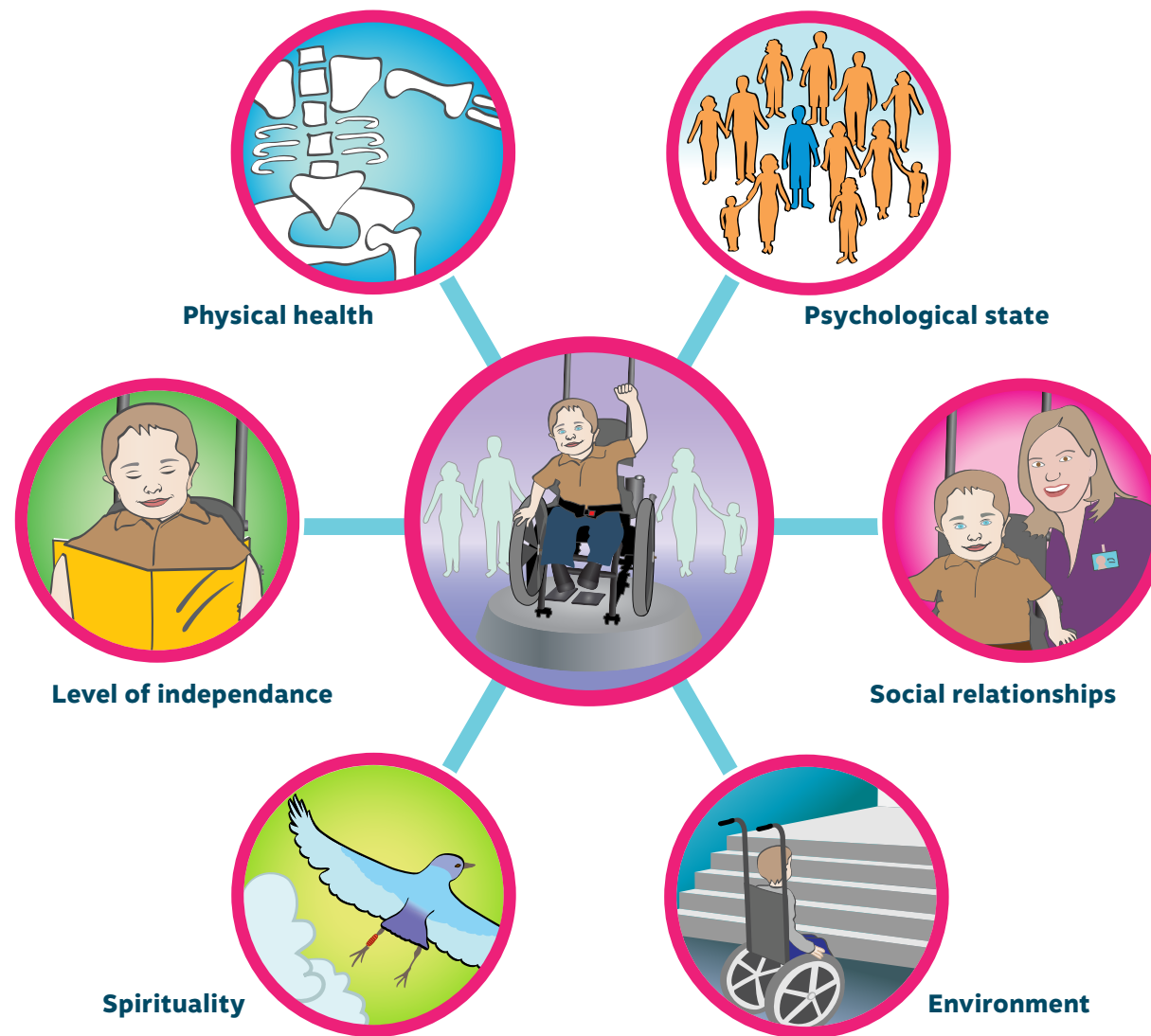
## What is Quality of Life?

“Quality of life” is a person's own view of their position in life or well-being.

There are six main aspects that contribute to one's quality of life: physical health, psychological state, level of independence, social relationships, personal beliefs/spirituality and their relationship to their environment.

There are many ways to improve quality of life. Treatments and medications are often given to enhance physical health, but we can do more! For example, having meaningful relationships with other people can also improve quality of life.

## Quality of Life



## What Have We Found?

We analyzed the findings of 43 research studies, which explored various components of quality of life among children with OI. These studies were conducted in 11 countries.

### There is a lack of high quality research.

The research on quality of life in children and adults with OI is of low to moderate quality. This means that the results from these 43 studies may not be 100% reliable. There is room for improvement for researchers who are conducting studies in this area.

### All aspects of quality of life have not been explored.

Many studies look at physical health and psychological components of quality of life. Sub-components of physical health such as experiences of pain have also been explored. Research on the contribution of the environment and personal beliefs has not been conducted to date. Little research has been conducted on the quality of life of parents and the quality of life of siblings and grandparents has not yet been explored.

### All types of OI need to be included.

Children and adults with mild types of OI are included more often than those with severe OI types, and more research needs to be done in this population. Studies should be consider both the child and parent's perception of quality of life, as they may be different. This means that the results from the 43 studies cannot easily be generalized to those with severe OI types.

## References

**Quality of Life in Osteogenesis Imperfecta: A Mixed-Methods Systematic Review**

Dahan-Oliel et al. 2016. *Am J Med Coenet A*, 170A (1), 62-76

**The psychosocial experience of individuals living with osteogenesis imperfecta: a mixed-methods systematic review**

Tsimicalis et al. 2016. *Qual Life Res*, 25 (8), 1877-1896

**Pain Experiences of Children and Adolescents with Osteogenesis Imperfecta: An Integrative Review**

Nghiem et al. 2017. *Clin T Pain* 33 (3), 271-280

## A Summary of the Experiences of Children with OI Related to Different Components of Quality of Life

### Psychological state

**Fear of fractures.** Both children and adults describe having a fear of fractures, which often prevents them from participating in certain activities.

**Coping with challenges.** Some children learn to overcome stressful situations by being positive and using humour. However, others have difficulties dealing with their everyday challenges and may need more guidance.

**Adapting by learning new skills.** Some children learn to adapt to their condition and maintain a “fighter” attitude. Many develop unique abilities in problem solving and protecting themselves.

### Social relationships

**Relationship with others.** Both children and adults are able to form positive and meaningful friendships with people “who understand OI.” They also describe the importance of having supportive relationships with their parents in both childhood and adulthood.

**Isolation and feeling different.** Children feel different and left out, especially from physical activities with their peers. They express a desire to belong and struggle with the judgments of their peers.

### Level of Independence

**Intellectual feats.** Accomplishments by children and adults with OI are often intellectual: there is a strong drive for achievements within their academic, extracurricular, professional, and community activities.

### Physical Health: Pain

**Pain is present and problematic.** Children and adults with OI can experience both short- and long-term pain. Pain can be localized in different areas of the body, differ in intensity from person to person, and be described as “annoying,” “discomfort,” or “aching.” Pain also can prevent children and adults from sleeping, attending school, participating in leisurely activities, and carrying out daily chores or tasks.

**Issues with pain management.** Not much is known about how children and adults with OI deal with pain and what they do to relieve their pain. Over-the-counter pain medications offer limited relief and children often use other coping strategies, such as distraction.

## Quality of Life of Children and Adults with OI in Comparison to the General Population

**Physical health** quality of life scores have been found to be **lower** when compared to the general population. This is because children and adults with OI report more pain and activity limitations.

Children and adults with OI score the **same or better** in the **psychological and social relationship** domains of quality of life when compared to the general population.

## What's next?

The introduction of bisphosphonate treatment and participation in physical activities have been shown to improve quality of life. However, many other strategies and resources need to be created, accessed and promoted.

We are currently working towards filling the gaps in OI research. We hope that you may want to participate in future studies to help us understand and improve your quality of life and care.

If you have questions about your own or your child's quality of life, talk to your doctor, nurse, physiotherapist, occupational therapist, social worker, and/or teacher. We want to help! You can also connect with OI communities such as:

- OI Foundation ([www.oif.org](http://www.oif.org))
- Brittle Bone Society ([www.brittlebone.org](http://www.brittlebone.org))
- OI Australia ([www.oiaustralia.org.au](http://www.oiaustralia.org.au))
- OI Federation of Europe ([www.oife.org](http://www.oife.org))
- Care 4 Brittle Bones Foundation ([www.care4brittlebones.org](http://www.care4brittlebones.org))

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## The Quality of Life of Children with Osteogenesis Imperfecta and their Families



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