Resilience in osteogenesis imperfecta

Understanding the occupational adaptation process of individuals with osteogenesis imperfecta and their families to promote participation and occupational engagement: a scoping review
Understanding the occupational adaptation process of individuals with osteogenesis imperfecta and their families to promote participation and occupational engagement – a scoping review

Occupational Therapy, Thesis I

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Abstract

Introduction: Osteogenesis imperfecta (OI) is a condition that affects the whole family on many levels and is important to consider the management of the disease and its impact on occupational participation and engagement in daily activities. Aim: To explore the natural occupational adaptation process (OAP) and its impact on daily activities among individuals and families with OI. Method: Using Arksey and O’Malley’s five-step Scoping Study Framework (2005), a scoping review of 24 articles from seven online databases and literature hand-searching was conducted. Data extraction was performed for all studies that met the eligibility and quality criteria. Five main themes containing several subthemes were emerged in qualitative inductive thematic analysis. Findings: Findings from 24 reports highlight the social and emotional impact along with the physical challenges of living with OI that direct into adaptation process. Different occupational adaptation strategies that emerged in the data help overcome physical deficits and psychosocial implications and facilitate occupational participation and engagement in meaningful activities. Conclusions: The study confirmed these individuals’ and families’ high coping skills, despite experiencing many obstacles in their daily life. Significance: To better understand the adaptation process in OI population and give practical implications for occupational therapists to offer a better support these families need.

Keywords
Brittle bone disease, literature review, occupational adaptation, occupational science, occupational therapy

**List of abbreviations**

<table>
<thead>
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<th>Abbreviation</th>
<th>Description</th>
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<tr>
<td>ADL</td>
<td>activities of daily living</td>
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<tr>
<td>DI</td>
<td>dentinogenesis imperfecta</td>
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<td>IBTs</td>
<td>Internet-based technologies</td>
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<tr>
<td>OI</td>
<td>osteogenesis imperfecta</td>
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<td>OAM</td>
<td>Occupational Adaptation Model</td>
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<td>OAP</td>
<td>Occupational Adaptation Process</td>
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<tr>
<td>ICF</td>
<td>International Classification of Functioning, Disability and Health</td>
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<tr>
<td>PRISMA</td>
<td>Preferred Reporting Items for Systematic Reviews and Meta-Analyses</td>
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Understanding the occupational adaptation process of individuals with osteogenesis imperfecta and their families to promote participation and occupational engagement – a scoping review

**Introduction**

Osteogenesis imperfecta (OI), also known as brittle bone disease, is a rare heritable connective tissue disorder characterized by low bone density leading to fractures; incidence of approximately one in 10,000 births (Glorieux, 2008). Clinical features of OI range from perinatal lethality through severe skeletal deformity, mobility impairment, and very short stature to nearly asymptomatic manifestation with mild predisposition to fractures, normal stature, and normal lifespan (Parker & Parker, 2007). Most disorders caused by gene mutations in collagen are expressed through the skeleton, but as OI is a systematic connective tissue disorder, manifestations like blue or grey sclera, dentinogenesis imperfecta (DI), joint hypermobility, hearing loss, muscle weakness, cardiovascular and respiratory problems in other collagen-containing tissues may also be involved (Forlino et al., 2011).

OI is a disease with varying severity affecting physical, social, and emotional well-being of the person and their family (Wekre et al., 2010). The most common consequence is fractures, easy to emerge, often even without major trauma (Stoicănescu et al., 2015). Physical limitations, fragile bones and re-occurring fractures have significant impacts on participation and occupational engagement. Mueller and colleagues (2018) found that impairments may lead to decreased occupational performance as well as restrict participation. Several papers
have shown that skeletal deformities, weak muscles and fractures might influence occupation and participation (Löwing et al., 2007; Wekre et al. 2010; Vans et al., 2015).

Occupational therapy considers humans as occupational beings (Pereira, 2017). Participation is defined in ICF as ‘involvement in a life situation’ (World Health Organization, 2001, pp. 14). Although occupational performance describes the active participation in an occupation, engagement refers to the broader emotional and cognitive factors that may be associated with occupation (Polatajko et al., 2007). Occupational adaptation is a substantial concept in occupational therapy and occupational science (Grajo et al., 2018; Johansson et al., 2018). According to the Occupational Therapy Practice Framework, adaptation is referred as simplified manner of modification of the task or environment to enable participation and support occupational engagement (AOTA, 2014). Nayar and Stanley (2015) considered occupational adaptation as a social process which emerge in people’s everyday lives and the ability to adapt in occupational challenge is influenced by personal factors as resilience, self-esteem, and relationships of interdependence with significant others (Palma-Candia et al., 2019). Resilience is an individual’s ability to overcome adversities (Muriithi et al., 2022).

Most research in the field of OI focuses on the medical, surgical, and orthopaedical management or on exploring the quality of life of OI population (Widmann et al., 2002; Biggin & Munns, 2014; Marom et al., 2020). Some studies have investigated activity limitations and participation restrictions (Harsevoort et al., 2020; Montpetit et al., 2011), but little is known about the OAP in this population. Therefore, the study also includes family’s perspective to note the importance of social environment and uncover the natural occupational adaptation process (OAP) to promote participation and engagement in meaningful activities (Nayar & Stanley, 2015).
Aim

This study aimed to explore experiences of OAP in individuals with OI as well as their families and how occupational adaptations influence participation and occupational engagement.

Research question:

- What are the experiences of OAP for individuals diagnosed with OI and their families?

Secondary questions:

- What occupational adaptation strategies are used by individuals with OI and their families?
- How do occupational adaptations influence participation and occupational engagement?

Method

The thesis presents a scoping review to explore an OAP in the OI population, which is an efficient and effective approach to collect, analyse, summarize and present available evidence and get a broad scope on the topic (McKinstry et al., 2014). Five-stage framework articulated by Arksey and O’Malley was guided the research (O’Brien et al., 2016). Qualitative data was captured from the perspectives of individuals and their families, including participants’ quotes, their experiences and perceptions. A decision was made include other publications and non-peer-reviewed journals in addition to peer-reviewed papers. The literature was also sought from reference lists of relevant papers.
The research question and search terms were defined utilizing the SPIDER tool (see Table 1); valuable for qualitative research exploring more about attitudes and individual experiences of the sample (Cooke et al., 2012).

[“Insert Table 1”]

The search terms (see Table 2) were formulated, combined and adjusted using Boolean operators as well as truncation and quotation marks where appropriate to ensure all relevant articles were detected.

[“Insert Table 2”]

Sources were selected by using inclusion and exclusion criteria (see Table 3). Year limits from 2000 to 2022 were applied only in PubMed database searching and considered relevant because of abundance of results. The research explored OI population, other similar conditions affecting connective or bone tissues likewise were excluded.

[“Insert Table 3”]

Participants
The chosen group were individuals diagnosed with OI and their families.

Data collection
Seven medical electronic databases (showed in Figure 1 PRISMA flow diagram) were searched between March to April 2022. Manual search of reference lists of included studies was also performed. The report was conducted according PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) statement guidelines for scoping reviews (see Figure 1 PRISMA flow diagram); during the search and data extraction phases, the author recorded studies found in each database, removed duplicates and refined number of sources into the screening and for inclusion (Page et al., 2021).
The selection process of the articles involved screening by titles and then abstracts to note potentially relevant sources, and finally reading the whole text to establish eligibility for inclusion (O’Brien et al., 2016). Additional studies were hand searched from reference lists of included freely available full texts. Literature that met the inclusion criteria and addressed the research questions were included for data analysis.

Data charting and analysis

Selected sources were read through several times to make sense of the data, extract results and descriptively map them (Peters et al., 2020). Data analysis included identifying firstly the key items of evidence received from the primary research reports (Arksey & O’Malley, 2005). Descriptive quantitative data as author(s), publication year, national context of the study, aim, study design, sample and important results were extracted (see Supplementary File Table 1). Data were charted using qualitative inductive thematic analysis to identify the main issues and major themes relevant to the aim of the study (Tong et al., 2012). Specifically, the qualitative data were read through and interpreted against themes and were sorted into different categories and subcategories related to the research question.

To facilitate the application of the report in occupational therapy theory and practice, elements of the OAM and OAP were applied in the data charting process. On that basis, the content areas of occupational challenge and adaptive response were used. Charted data were assembled in a form of table to create a descriptive summary of the results (see Supplementary File Table 2) and bring out the dominant areas of interests in research as well as significant gaps. (O’Brien et al., 2016; Stanley, 2014).
Ethical considerations

Several ethical aspects were taken into consideration to assure the quality of the study. Plagiarized material was avoided by acknowledging the author and using appropriate citations when referring to another people’s work and ensured trustworthiness using the existing evidence accurately and fairly by being unbiased and extracted the data precisely (Wager & Wiffen, 2011). The process of the research was fully detailed to ensure the transparency and replicability of the study (McKinstry et al., 2014). The supervisor helped to ensure that the thesis met the standards of the University and the academic discipline as well as provided independent review of the methods, especially search terms, data extraction and analysis.

In the view of the fact that the author has a personal interest regarding the research subject as being part of the OI population and thus having her own experiences, methods to ensure transparency of the study were adopted. The author tried to be as neutral as possible in the collection, interpretation and presentation of the data, keeping in mind the importance of the fact that there is no completely objective knowledge (Holmes, 2020). Author’s personal positionality were discussed with the thesis supervisor to contribute independence in data extraction and coding. The strengths and limitations of the study are considered in discussion section.

Findings

A total of 3628 records from databases searches and three additional studies were identified through reference lists. Ninety-nine full-text reports were reviewed against the inclusion criteria and 24 studies included for data extraction and synthesis. Methodological approaches included 16 qualitative studies, two mixed-method studies, two systematic reviews, one case report and three personal reflection reports from non-peer-reviewed journal were retrieved.
There were seven studies from Canada, five from Sweden, four from UK, three from USA and individual studies from Brazil, Portugal and Switzerland, published between 1989 to 2022.

All extracted data focused on the child, adolescent or adult with OI and/or caregivers’ experiences of living with OI. Two studies also collected health professionals’ observations among others about the impact of living with OI. Only six studies referred to the severity of the disease, specified as OI types I, III, IV, “mild to moderate OI” or “severe OI” and in 18 sources it was not specified. In qualitative studies, interviews and questionnaires or surveys were used as data collection method. Five main themes emerged containing several subthemes (presented in Table 4).

[“Insert Table 4”]

**Occupational performance challenge**

Physical and psychosocial limitations as well as pain affected engagement in daily activities.

**Physical and psychosocial limitations**

Young adults with OI reported difficulties in physical movements and activities like walking, running, climbing stairs, lifting heavy objects, managing personal care and participating in sport activities (Balkefors et al., 2013; Dogba et al., 2013; Michalovic et al., 2020). Reduced function was more evident during cast-wearing and recovery periods or after surgeries, which made engagement in ADL, educational activities and caring for the child challenging (Castro et al., 2019; Castro et al., 2020; Hill et al., 2014; Hill et al., 2022; Siedlikowski et al., 2021).

Children with OI perceived their differences from others and were not only limited by their own physical disabilities, but also influenced by others perception of their condition which culminated in them being excluded and bullied (Siedlikowski et al., 2021; Tsimicalis et al., 2016). Hearing loss drastically reduced the communication ability (Kasper, 2013). Living in
stress as well as fear of possible fractures reduced participation in different activities (Hill et al., 2014; Hill et al., 2022; Tsimicalis et al., 2016). Parents described difficulties trusting others to look and care for their child manifested the feeling of isolation (Castro et al., 2020; Hill et al., 2014; Hill et al., 2022).

Pain
Tiredness and pain reduced functionality and limited the types of activities performed (Hill et al., 2014; Hill et al., 2019). Most children experienced moderate to severe pain related to fractures, many reported nonfracture pain caused by a minor trauma, recent physical workout, prolonged poor posture position or other non-identified reason (Zack et al., 2005). Pain tolerance was mentioned to come with time and experience (Siedlikowski et al., 2021).

Experiences of participation

Engagement in activities
Children attended a play group or specialized school for physically disabled children, played with physically active peers and engaged in several various low-impact activities like swimming, boccia ball, artistic classes, and piano lessons or adapted sports and special Paralympic tournaments (Brodin & Millde, 1990; Castro et al., 2020; Cinman, 2001; Siedlikowski et al., 2021). An adult engaged in different activities as tubing down the river and horseback riding, although was mindfully prepared for the possibility to get seriously hurt (Ablon, 2003).

Occupational performance
Many children had constraints of outdoor activities and high impact sports like playing football, running fast, jumping on a trampoline and riding a horse (Hill et al., 2014; Paiva et al., 2018; Tsimicalis et al., 2016). Young children were restricted using slides at playground
or even playing with peers (Castro et al., 2020; Hill et al., 2014). Parents restricted their own activities and social relationships to stay close to their fragile child (Hill et al., 2019).

**Family interaction**

Having a child with OI affected family dynamics on many levels (Dogba et al., 2013).

**Everyday management**

Parents admitted that the child with OI received most of their attention compared to other siblings and their spouse (Dogba et al., 2013). Family plans were not discussed too far in advance to prevent disappointment in all family members (Wiggins et al., 2017). Families challenged to manage in everyday obligations and routines at times of fractures and hospital stays, while the whole family life was disrupted without mother being around (Hill et al., 2019; Hill et al., 2022; Santos et al., 2018). On the other hand, many children received treatment far from home and sometimes without a parent support (Siedlikowski et al., 2021).

**Responsibility and roles**

Parents’ role was being either a companion or manager to their child with OI, depending on the child’s age and their level of independence (Hill et al., 2019). Caregivers had also supportive role in achieving children’s independence and discussing the safety issues together (Hill et al., 2022; Wiggins et al., 2017). Being a “strong role model” was adopted to show that OI is not an obstacle (Hill et al., 2022, pp 4).

**Activities, risk and safety**

Finding suitable as well as enjoyable outdoor activities for their child with OI was complicated (Bernahäll Claesson & Brodin, 2002). Changes and additional planning in family activities were adopted to ensure that a child with OI is involved, but also meet the needs of other family members; low-impact family activities were preferred and sometimes
different types of activities were restricted or excluded (Castro et al., 2020; Hill et al., 2019; Dogba et al., 2013).

Proactive measures were used to ensure safety, manage everyday life as well as medical visits, although finding a balance between keeping safe and not expressing overprotectiveness was a major challenge for many parents (Castro et al., 2020; Hill et al., 2014; Hill et al. 2022). Children expressed discontent about parents’ attempts to protect them (Paiva et al., 2018; Siedlikowski et al., 2021). Excessive protection was often accompanied with restrictions in activities (Hill et al., 2014; Hill et al., 2022; Tsimicalis et al., 2016), but caregivers with OI did not apply as many restrictions as parents whose child was the only one in the family with this diagnosis (Brodin et al., 1990).

Occupational environment

Barriers

Lack of environmental adaptations in public areas were seen as hinders – activities were influenced by several obstacles located in playing fields and made it impossible to use with wheelchair (Bernahäll Claesson & Brodin, 2002). Concerns about physical accessibility in different areas and the absence of special transportation were pointed out (Cinman, 2001; Kasper, 2013).

Environmental modifications

Facilitative environment in home or in other frequently visited spaces as schools were highlighted to make the environment more suitable for the individual with OI (Brodin, 1989; Castro et al., 2020; Bernahäll Claesson & Brodin, 2002; Dogba et al., 2013; Paiva et al., 2018). The need of either home adjustments (see Table 5) or moving into more suitable environment, preferably in a one-level house with no stairs was mentioned (Dogba et al., 2020; Hill et al., 2022).
**Assistive devices and internet-based technologies (IBTs)**

The use of assistive devices (see Table 5) either standardized or individually modified were mentioned to promote functional, environmental and social management (Balkefors et al., 2013; Brodin, 1989; Hill et al., 2014; Kasper, 2013; Yochum et al., 2002). Mobility aids were the most mentioned (19 sources out of 24) assistive devices used in OI population within current data and were used to avoid pain or tiredness and promote participation in ADL as well as the way to ensure safety in a crowded environment (Hill et al, 2014; Michalovic et al., 2021; Millde et al., 1990; Wiggins et al., 2017; Yochum et al., 2002).

[“Insert Table 5”]

Castro et al. (2019) explored the use of IBTs among individuals with OI and identified benefits to manage physical (used as low-impact activity, against pain, during medical procedures, immobilisation and recoveries), social (enhance communication and reduce isolation), psychological (internet-based stress-management), environmental (e.g., avoid freshly washed slippery floors) as well as logistical (coordinating appointments, contacting clinicians, finding resources, product and services) challenges.

**Resilience**

In the theme of resilience, five subthemes emerged.

**Psychological, physical and psychosocial support**

Studies indicated patients’ and families’ positive outlook and optimistic mindset on life (Cinman, 2001; Hill et al., 2019) by enjoying simple things (Dogba et al., 2013), using humor and hoping for better tomorrow (Tsimicalis et al., 2016). Another way of thinking like “living one day at a time” (Dogba et al., 2013, pp. 6) and “seeing the challenges of life as relative” (Ablon, 2003, pp. 205) was adopted. Strong and “fighter” attitude, emotional talks and engagement in leisure activities helped managing stressful times (Brodin & Millde, 1990;
Hill et al., 2019; Tsimicalis et al., 2016; Wiggins et al., 2017). Families highlighted focusing on the strengths and achievements in social and academic performance over physical activities (Hill et al., 2019; Siedlikowski et al., 2021).

Sources of support included family, friends, health professionals and community resources as well as religious faith and other OI families (Hill et al., 2019; Michalovic et al., 2020). Firm friendships helped to break the feeling of being excluded, pets support was referred when reflecting “at least I have friends” (Siedlikowski et al., 2021, pp. 10). Physical help was needed to manage with physically difficult obligations like managing personal care, moving in community, shopping and reaching things above certain heights (Brodin, 1989; Dogba et al., 2013; Kasper, 2003; Michalovic et al., 2020; Paiva et al., 2018). Siblings were involved in supporting safety, avoiding rough play (Hill et al., 2019, Santos et al., 2018). Parents social interaction was promoted in caregivers’ support group (Hill et al., 2019; Millde & Brodin, 1990). Grandparents and friends were involved caring process to enable parenting breaks (Castro et al., 2020).

**Self-advocacy**

Several strategies were reported to develop self-protection measures and self-advocacy in view of the need for constant vigilence in everyday planning (Ablon, 2003). Children's developed self-awareness to discover about their physical boundaries and gain confidence in identifying environmental threat (Wiggins et al., 2017) and avoiding risks in activities with others (Hill et al., 2014). At an early age, children acquired specific survival skills as the ability to read X-rays, articulate about their bodies and needs clearly and accurately for self-defense in hospital setting as well as reduce risks by simply naming who and how to take care of them (Ablon, 2003; Tsimicalis et al., 2016). The ability to be especially attentive and attuned to own body as well as opting for less physically demanding positions were essential (Michalovic et al., 2020).
Educational and professional activities

School services, homeschooling and specialized transportation were used to support access to educational activities (Castro et al., 2020; Hill et al., 2022; Kasper, 2013). Alternative communication methods like lip-reading course and sign language lessons were acquired to compensate hearing loss (Kasper, 2013). Because of the limited access to specialized clinicians, caregivers learned about the condition and possible treatment options from websites and other parents (Castro et al., 2019; Dogba et al., 2015; Hill et al., 2019).

Ending employment to concentrate on children’s needs or working at home office either part time to manage several medical appointments and concerns about safe childcare were mentioned (Dogba et al., 2013; Hill et al., 2019). Choice of employment and leisure activities were influenced by the fear of fractures and safety concerns, which persists into adulthood and makes job changes sometimes inevitable (Michalovic et al, 2020; Tsimicalis et al. 2016). IBTs helped parents to continue with their professional activities (Castro et al., 2019).

Fracture-related

Parents took the lead in administering first aid by assessing the fracture, providing early pain relief and visiting hospital for clinical evaluation and having a cast applied, providing rest for the remainder of the day (Castro et al., 2020; Hill et al., 2014). Families were prepared by traveling with first aid supplies like fracture and splinting kits (Wiggins et al., 2017). Ice packs, forgoing painkillers and unnecessary movement prevention were used when family assessed the fracture as untreatable by medical intervention (Michalovic et al., 2020). Caregivers explained staying home to provide pain medications as well as social and psychological support after the surgery (Castro et al., 2020). Diverse situation and pain coping strategies were used depending on fracture-related or non-fracture pain (Tsimicalis et al., 2016), distraction and comfort was used to improve well-being (Santos et al., 2018).


**Preventive and adaptive strategies**

A preventive role was taken by all family members by using several proactive strategies to avoid bone and joint injuries and ensure safety (Hill et al., 2019; Paiva et al., 2018). Being extra careful, avoiding fast and unpredictable activities as well as the need of advance planning were noted by individuals with OI, while parents were staying close or forbidding their children to do certain occupations (Hill et al., 2014; Santos et al., 2018; Tsimicalis et al., 2016). Young adults described choosing low-impact activities, being cautious of tripping and other physical movements (Ablon, 2003; Michalovic et al., 2020; Paiva et al., 2018).

The need for adaptation was constant and inevitable (Carlson, 2007; Dogba et al., 2013; Hill et al., 2022; Paiva et al., 2018). After the diagnosis, parents redefined their “new normal” and educated children accordingly (Dogba et al., 2015), managing the uncertainty as part of daily life by being careful while also considering the possibility of getting hurt (Ablon, 2003; Wiggins et al., 2017). Childrens’ managed otherness by sharing their uniqueness and participating in activities as best as they could with peers (Siedlikowski et al., 2021); adjusting occupations by doing things in a slightly different way or choosing another field of action instead of missing out on activities (Castro et al., 2020; Hill et al., 2014).

Caregivers were amazed and inspired by their child’s coping ability (Hill et al., 2019). Castro et al. (2020) highlighted parents’ high adaptation skills and strategies as managing the environment, the importance of planning and routine, using medical resources and keeping their schedules flexible to respond possible injuries quickly.

**Discussion**

The review aimed to determine the factors and strategies involved in OAP and influence on participation and occupational engagement in individuals with OI and their families. An occupational therapy frame of reference focusing on the natural OAP which begins with
occupational challenges and takes place between the person and occupational environment was used to address relevant information (Nayar & Stanley, 2015).

Physical and emotional barriers (fear as well as over-protection) were seen to limit participation in daily occupations and social activities. Inclusive and carefully planned or adjusted environment make a great contribution to enable participation in activities, support independence and meet the needs of children with physical as well as visual impairments (AOTA, 2022; Doctoroff, 2001). Also, the influence of social attitudes on engagement in daily life cannot be ignored. In fact, Maguire et al. (2021) explored living with chronic illness and found that social appraisals had stronger associations with well-being than extent of social contact. Thus, societal expectations (environmental and attitudinal) may disable or enable participation and result accordingly to social inclusion or exclusion (Wong et al., 2021). Occupational therapist helps to develop and implement environmental modifications to create more supportive environment, which also have been shown to reduce caregiver burden (AOTA, 2022).

Overprotectiveness was found to be part of a preventive strategy in disease management and is common phenomenon when considering families with OI, but it’s implications may have a greater impact on child's development. Fingerman et al., (2012) found positive associations between intense parental support and adolescents’ psychological adjustment and functioning. By contrast, recent study showed that overprotection may lead to (mal)adaptive adolescent functioning (Arslan et al., 2023). Besides, children with OI appeared disturbed by their parents’ extreme protectiveness (Paiva et al., 2018; Siedlikowski et al., 2021) and further research is needed to determine the long-term effects on occupational performance and engagement of overcare on children with OI.

Families with OI reported living constantly in uncertainty and anxiety affecting mental health as well as family functioning (Hill et al., 2022). Lazow et al. (2019) revealed that higher
levels of pain and lower physical functioning in children with OI are associated with higher levels of parental stress and poorer quality of life. However, since it is a hereditary disease, many parents struggle also with the limitations of their own physical health (Hill et al., 2022). The adaptation process is thus multifaceted and there is a need to highlight family-centered care and adequate support to improve the well-being of both patients and caregivers (Kokorelias et al., 2019). Edwards and colleagues (2003) found that assisting caregivers to become more involved in the occupational therapy planning and implementation process while looking each family as a unique entity, also focusing on their strengths and individual needs is important in occupational therapy practice.

Hiding pain and covering it with overflowing positivity can be seen as a coping strategy and part of an adaptation process (Ablon, 2003). This is consistent with Wakefield et al. (2021) who found that adolescents with chronic pain use concealment because of desire to be treated as other peers, avoidance of being a social burden or judged by others. Likewise, individuals with OI have also mentioned the constant endeavor of normality to feel as equal as others (Hill et al., 2022; Santos et al., 2018; Siedlikowski et al., 2021). On the other hand, efforts to feel and be equal to peers as well as hiding real feelings may lead to the consequences of social isolation and emotional burden and therefore, symptomatic concealment and over joyfulness could prevent access to social support and impede adaptation process through it (Ross et al., 2018). Social support is an effective rehabilitation strategy can be used with individuals with disabilities to enhance motivation and ability to participate in meaningful occupations (Isaksson et al., 2007).

Based on current findings, the inclusion of animals and pets for the purpose of social support is rather low in families with OI, yet feelings of loneliness and exclusion were significantly expressed among children as well as parents. Notably, interactions with animals may provide benefits like increased physical activity, emotional support as well as pain reduction (Carey et
al., 2022; Gee et al., 2021). Service dogs could help OI patients with hearing loss to compensate for the physical deficit and reduce psychosocial challenges (Kasper, 2013; Kuurila et al., 2002; Pillion et al., 2011), Fairman & Huebner (2000) have found that service dogs offer help with self-care, and productive activities as well as play and leisure time (e.g., retrieve dropped or out of reach items as well as alert about different noises). In addition, animals can facilitate disabled people socializing with others – Hart et al. (2015) reported that there were more social approaches when an individual who moves with a wheelchair had the assistance dog around them. The potential use of pets or animals as social support among families with OI is unknown and needs to be explored.

Social support is seen as important part of an adaptation process and feelings of exclusion or being isolated/different from others were evident in the current findings (Dogba et al., 2013; Hill et al., 2022; Siedlikowski et al., 2021). In addition, Strom & Egede (2021) have found that high levels of social support have a positive influence on motivation and helps to cope with stressful situations in individuals with chronic diseases. Emotional and social support has a positive effect on adaptation mechanisms (Zhou & Lin, 2016), which emphasizes the need of a family involvement in therapy process.

Conclusions

This study provides more insight and deeper understanding into the OAP among individuals and families with OI. Occupational performance challenges like physical and psychosocial limitations as well as pain which activate OAP were identified. The findings suggest that individuals and families with OI use a wide range of adaptation strategies in response to the chronic illness and disability including environmental modifications, use of technology and assistive devices as well as different behavioural and psychological strategies when coping with disability.
Significance

The review creates important value for healthcare professionals as well as occupational therapy theory and practice. The study highlights the importance and need of family-centered practice and collaborative problem-solving. It has a practical implication on OI population, occupational therapists working in OI patients in rehabilitation services as well as policy makers to emphasize the accessibility of public environments. Occupational therapists can provide effective interventions supporting OAP and management with the disease to help people with OI and their families to enhance the participation and occupational engagement in daily activities with the use of various occupational adaptation strategies.

Methodological considerations

There are several limitations to this study, in particular the review was carried out by one author, although multidisciplinary team approach is recommended because of the complexity of the process (Arksey & O’Malley, 2005). The most information about technical aids were from 1989 and 1990, and few other studies that seemed important (also from similar time period), were not freely available. Thus, the information may be out of date and since new devices have been created over the years, there may be more recent innovations which needs to be explored in future studies. Further, the research from databases was undertaken a year ago, and quick search revealed new studies about pain management, quality of life of OI and “OI Splint Kit” for fracture management. Therefore, more recent studies and relevant papers may have been missed in this review and considered as a major limitation.

There are also strengths of the study. An in-depth review and analysis were conducted when searching from multiple databases and reference lists. The author provided new possible research themes in the discussion section.
References

References marked with an asterisk indicate studies included in the review.


Muriithi, B. A. K., Muriithi, J., Gupta, J., Radziak, J. & Story, A. (2022). Occupational resilience: construct and practice implications in occupational therapy. Research Square. [https://doi.org/10.21203/rs.3.rs-1786056/v1](https://doi.org/10.21203/rs.3.rs-1786056/v1)


Strom, J. L. & Egede, L. E. (2012). The impact of social support on outcomes in adult patients with type 2 diabetes: a systematic review. *Current Diabetes Reports*; 12(6), 769-81. [https://doi.org/10.1007/s11892-012-0317-0](https://doi.org/10.1007/s11892-012-0317-0)


Table 1. The SPIDER tool.

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<th>Design</th>
<th>Evaluation</th>
<th>Research Type</th>
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<td>Individuals diagnosed with osteogenesis imperfecta and their families</td>
<td>Lived experiences and perspectives of individuals and their families</td>
<td>All types</td>
<td>Participation, Occupational performance, Occupational roles</td>
<td>Qualitative or mixed methods, grey literature</td>
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Table 2. Search terms.

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<td>view*</td>
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<tr>
<td>person</td>
<td>experience*</td>
</tr>
<tr>
<td>child*</td>
<td>feel*</td>
</tr>
<tr>
<td>adolescen*</td>
<td>perce*</td>
</tr>
<tr>
<td>adult</td>
<td>attitude</td>
</tr>
<tr>
<td>parent*</td>
<td>opinion</td>
</tr>
<tr>
<td>mother</td>
<td>belie*</td>
</tr>
<tr>
<td>father</td>
<td>know*</td>
</tr>
<tr>
<td>family</td>
<td>understand*</td>
</tr>
<tr>
<td>‘Osteogenesis Imperfecta’</td>
<td>‘quality of life’</td>
</tr>
<tr>
<td>‘OI’</td>
<td>‘lived body’</td>
</tr>
<tr>
<td>‘Brittle Bone Disease’</td>
<td>challenge</td>
</tr>
<tr>
<td>‘fragile bone’</td>
<td>impact</td>
</tr>
<tr>
<td>‘Fragilitas Ossium’</td>
<td>daily</td>
</tr>
<tr>
<td></td>
<td>life</td>
</tr>
<tr>
<td></td>
<td>activ*</td>
</tr>
</tbody>
</table>
Table 3. Inclusion/exclusion criteria

<table>
<thead>
<tr>
<th>Inclusion criteria</th>
<th>Exclusion criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Individuals diagnosed with osteogenesis imperfecta and their families</td>
<td>Other similar conditions, e.g., Ehlers-Danlos syndrome (EDS) or osteoporosis.</td>
</tr>
<tr>
<td>English and Estonian language</td>
<td>Other languages</td>
</tr>
<tr>
<td>Full text available</td>
<td>Only abstract available</td>
</tr>
</tbody>
</table>
Table 4. Themes and subthemes emerged from data analysis.

<table>
<thead>
<tr>
<th>Theme</th>
<th>Subtheme</th>
</tr>
</thead>
<tbody>
<tr>
<td>Occupational performance challenge</td>
<td>Physical and psychosocial limitations</td>
</tr>
<tr>
<td></td>
<td>Pain</td>
</tr>
<tr>
<td>Experiences of participation</td>
<td>Engagement in activities</td>
</tr>
<tr>
<td></td>
<td>Occupational performance</td>
</tr>
<tr>
<td>Family interaction</td>
<td>Everyday management</td>
</tr>
<tr>
<td></td>
<td>Responsibility and roles</td>
</tr>
<tr>
<td></td>
<td>Activities, risk and safety</td>
</tr>
<tr>
<td>Occupational environment</td>
<td>Barriers</td>
</tr>
<tr>
<td></td>
<td>Environmental modifications</td>
</tr>
<tr>
<td></td>
<td>Assistive devices and internet-based technologies (IBTs)</td>
</tr>
<tr>
<td>Resilience</td>
<td>Psychological, physical and psychosocial support</td>
</tr>
<tr>
<td></td>
<td>Self-advocacy</td>
</tr>
<tr>
<td></td>
<td>Educational and professional activities</td>
</tr>
<tr>
<td></td>
<td>Fracture-related</td>
</tr>
<tr>
<td></td>
<td>Preventive and adaptive strategies</td>
</tr>
</tbody>
</table>
Table 5. Environmental adaptations and assistive devices emerged from data analysis.

<table>
<thead>
<tr>
<th>Environmental adaptations</th>
<th>Assistive devices</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wheelchair ramp</td>
<td>Mobility aids (wheelchair, walking frame, crutches)</td>
</tr>
<tr>
<td>An extra entrance with a lift</td>
<td>Adjusted car (with hand control, specially modified seat)</td>
</tr>
<tr>
<td>Functional elevator</td>
<td>Height-adjustable table and chair</td>
</tr>
<tr>
<td>Hoist</td>
<td>Chair with backrest and extra foot support</td>
</tr>
<tr>
<td>Wide doorways</td>
<td>Waterbed</td>
</tr>
<tr>
<td>Removed door thresholds</td>
<td>Chunky pen</td>
</tr>
<tr>
<td>Lowered or doubled light switches</td>
<td>Shoe insertions or specially made shoes</td>
</tr>
<tr>
<td>Lowered bathroom sink</td>
<td>Hearing aid</td>
</tr>
<tr>
<td>Shock-absorbent carpeting</td>
<td>Captioned phone</td>
</tr>
<tr>
<td>Removed physical obstacles (e.g falling objects)</td>
<td></td>
</tr>
</tbody>
</table>
Figure 1. PRISMA flow chart of Search and Screening.