Osteogenesis imperfecta (OI) is a genetic disorder with main clinical features that include bone fragility, frequent fractures, and varying degrees of physical limitations. Individuals with OI may experience pain in many forms including acute fracture pain and chronic non-fracture pain. Although pain is a recognized symptom in individuals with OI, there are no guidelines outlining how pain related to OI should be managed.

Objective

The objective of this integrative review was to:
1) determine the pain experiences of children and adults with OI;
2) assess the methodological quality of studies on OI related pain;
3) identify implications for future research and practice.

Methods

Table 1 Inclusion Criteria

<table>
<thead>
<tr>
<th>Type of Participants</th>
<th>Type of Outcome Measures</th>
<th>Type of Studies</th>
<th>Languages</th>
<th>Quality</th>
</tr>
</thead>
<tbody>
<tr>
<td>12-year-old children &amp; adults</td>
<td>Assessment of pain as a primary or secondary outcome</td>
<td>Published, qualitative, quantitative, mixed-methods</td>
<td>English, French</td>
<td>Studies with a score greater than 0%</td>
</tr>
</tbody>
</table>

Databases searched: CINAHL, Medline, EMBASE, PsycInfo, and Joanna Briggs Institute.

The Quality Assessment Tool (QAT) (Srinivay et al., 2012) was used to appraise studies. Scores range from 0 to 100.

Extracted data were analyzed using descriptive statistics and thematic analysis.

Results

Figure 1 Flow Chart of Study

Selection Process

Retrieved citations (n = 811)
Additional records identified (n = 0)
Titles and abstracts screened after duplicates removed (n = 607 (204 duplicates))
Records excluded (n = 485)
Full-text articles screened (n = 145)
Full-text articles excluded from review (n = 25)

Figure 2 Design and Quality

Number of Dimensions Assessed

- Type of Dimension
  - Sensory
  - Evaluative
  - Affective

Number of Dimensions Assessed

- One
- Two
- Three

Figure 3 Important Findings

Pain as an Outcome

57% 43%
Primary Secondary

Pain Management Method

42% 23%

Pain Intensity

50% 49.9%
Real Time Recall of the Last Day/Week/Month

61% 54.9%
Momentary Cross-section

15% 18%
Recall and Momentary

8% 8%
Hospital & Home

69% 8%
Not Stated

60% 2%
0% School & Work

Discussion & Conclusion

Future Directions

Future research would require longitudinal studies focused on assessing the multiple dimensions of pain related to OI in naturalistic environments and in real-time.

Future research with children diagnosed with OI must also focus on characterizing pain using valid and reliable pain assessment tools; the PediMPACT consensus statement has put forth evidence-based guidelines for the assessment of acute and chronic pain in pediatric populations (McGrath et al., 2008).

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References


Identified Themes

- Pain is Present and Problematic
  - Chronic pain related to OI was mild in intensity for children and mild to moderate in intensity for adults, located in several areas of the body (e.g., back, joints, limbs), and described as “annoying”, “discomforting”, and “aching”.
  - Acute fracture pain was reported to be of greater intensity from recall data in two pediatric studies.
  - Pain prevented individuals with OI from attending school, participating in leisurely activities, and carrying out daily activities, as well as interfered with sleep.
  - Experiences with pain management in children with OI suggest that standard doses of analgesics are not helpful.

Issues with Pain Assessment

- Pain was not always assessed using a multidimensional approach and appropriate tools.
- Pain related to OI was often assessed cross-sectionally or through recall memory in hospital settings.
- Assessing pain in younger children seemed to be a challenge.