# Osteogenesis Imperfecta in Childhood: Perceived Competence in Relation to Impairment and Disability

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ABSTRACT. Engelbert RH, Gulmans VA, Uiterwaal CS, Helders PJ. Osteogenesis imperfecta in childhood: perceived competence in relation to impairment and disability. Arch Phys Med Rehabil 2001;82:943-8.

**Objectives:** To examine the perceived competence of children with different types of osteogenesis imperfecta (OI) and to investigate the possible relationships between their perceived competence and impairment parameters.

**Design:** Cross-sectional study.

**Setting:** National referral center (hospital) for the treatment of children with OI.

**Patients:** Forty children with OI (type I = 17; type III = 11; type IV = 12) with a mean age ± standard deviation of 12.6 ± 3.2 years.

**Interventions:** Measured joint range of motion (ROM) in the upper extremities (UEs), and lower extremities (LEs), muscle strength, functional skills, ambulation, and perceived competence.

**Main Outcome Measures:** Joint ROM in UE and LE; muscle strength (using the manual muscle testing criteria of the Medical Research Council); functional skills using the Pediatric Evaluation of Disability Inventory in 3 domains (self-care, mobility, social function). Ambulation (according to Bleck and classified as nonwalking, therapy walking, household walking, neighborhood walking, community walking with or without the use of crutches), and perceived competence (using the Harter Self-Perception Profile for Children, which was cross-culturally validated for Dutch children).

**Results:** In children with type I, joint ROM and muscle strength were almost comparable to the healthy population. In children with type III, a severe decrease in joint ROM was measured, especially in the LEs, and muscle strength was severely decreased in the UEs and LEs. In children with type IV, joint ROM and muscle strength decreased, especially in the LEs. In all types, fairly to strongly positive perceived competence was measured except for fairly negative perceived competence in the athletic performance subscale in type I and a fairly negative perceived competence in the romance subscale in type III. No correlations were found between (1) joint ROM and athletic performance and physical appearance, (2) muscle strength and athletic performance or physical appearance, or (3) the functional skills, concerning self-care and mobility, with the subscales of the perceived competence.

**Conclusions:** Although joint ROM, muscle strength, and functional and walking ability were related to the severity of the disease and differed significantly between the different types of OI, overall perceived competence in children with OI was fairly to strongly positive, without significant differences between the different types of OI.

**Key Words:** Disabled children; Mental competency; Muscles; Osteogenesis imperfecta; Range of motion, articular; Rehabilitation; Walking.



OSTEOGENESIS IMPERFECTA (OI) is a congenital connective tissue disorder that probably affects 1 in 5000 to 10,000 individuals without racial or ethnic preference.[1] Major clinical characteristics of OI include fragility of bone, osteopenia, variable degrees of short stature, and progressive skeletal deformities. Additional clinical manifestations are blue sclerae, dentinogenesis imperfecta, joint laxity, and maturity-onset deafness.[2] OI is subdivided into 4 types (I, II, III, IV), based on clinical, radiographic, and genetic characteristics according to Sillence and colleagues.[3,4] The most common type, OI type I, is characterized by osteopenia leading to fractures, distinctly blue sclerae, and a high incidence of adult-onset conductive hearing loss. Two subgroups of type I OI exist, type IA with normal teeth and type IB with dentinogenesis imperfecta. OI type II is usually lethal in the perinatal period. OI type III, the most severe type compatible with life, is characterized by severe osteopenia leading to multiple fractures, progressive deformity of bones and spine, and severely decreased height. OI type IV is rare and is characterized by osteopenia leading to fractures. Sclerae are normal. Short stature and deformity of the long bones and spine tend to be more marked than in OI type I. Some patients have normal teeth (OI type IVA); others have dentinogenesis imperfecta (OI type IVB).

The functional consequences and aspects of quality of life (QOL) in children with OI can be displayed using the concept of the disablement process. The disablement process refers to the various impacts of chronic and acute conditions on the functioning of specific body systems, on basic human performance, and on people's functioning and roles in society.[5] Two important, leading models guide disablement research in the United States and throughout western Europe: the International Classification of Impairments, Disabilities, and Handicaps (ICIDH-2)[6] and the extended construct of Nagi.[7-9]

In our study, we used the extended construct of Nagi, who delineated this process in 5 domains: active pathology, impairment, functional limitation, disability, and QOL.[8,9] In a cross-sectional and follow-up study, the different domains regarding impairment (joint range of motion [ROM], muscle strength), functional limitation, and disability parameters (functional skills) were described in 54 children with OI.[10,11] Aspects of QOL in children with OI have not been studied before nor has the relationship with impairment, functional limitation, and disability parameters. Perceived competence is viewed as an

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aspect of QOL and defined as the operationalization of children's opinion with respect to physical, cognitive, and social domains, as well as the assessment of general feelings of self-esteem.

The purpose of our study was therefore to examine the perceived competence of children with different types of OI. Second, we investigated the possible relationships between the perceived competence of these children and impairment parameters, such as joint ROM and muscle strength, functional ability, and level of ambulation.

## METHODS

### Patients

This cross-sectional study was conducted in 1998 at the Wilhelmina Children's Hospital, Utrecht, The Netherlands, a national referral center for the treatment of children with OI. Ours is the only center for multidisciplinary treatment of OI children in the Netherlands. The diagnosis, classification of the severity of the disease, and development of treatment strategies were performed by a multidisciplinary team, comprised of a clinical geneticist, pediatric orthopedic surgeon, physiatrist, pediatric physical therapist, and pediatrician. Diagnosis and classification were based on the medical history, radiography, physical examination, and more extensive research as bone mineral density and collagen research from skin biopsy when indicated.

Forty children (20 boys, 20 girls) with OI (type I = 17, type III = 11, type IV = 12) with a mean age ± standard deviation (SD) of 12.6 ± 3.2 years (range, 8–16yr) participated. Children were included if the diagnosis OI was definite and the type of OI was classified according to Sillence.[3,4] All parents of the children in this study gave informed consent.

### Design

All measurements were performed by one of the authors (RHE). Joint ROM (impairment domain) was measured in a standardized way with a standard 2-legged 360° goniometer,[a] using the anatomic landmark method and compared with reference values for children (table 1).[10-12] The values of joint ROM were based on a single trial.[10] To measure the total ROM in the upper (UEs) and lower extremities (LEs), the Joint Alignment and Motion (JAM) Scale[13] was used (table 1).

Muscle strength (impairment domain) was measured according to the manual muscle testing criteria of the Medical Research Council using a 6-point scale (range, 0–5) (table 1).[14,15] Muscle strength was scored as the best performance of 3 measurements. To measure the total muscle strength in the UEs and LEs, the muscle strength of all muscles were added and divided by the number of muscles. Because no significant

**Table 1: Tests Conducted to Measure Impairment, Disability, and QOL**

| Test | Test Sites or Parameters | Scoring |
|---|---|---|
| Impairment | | |
| Joint ROM | | |
| UE | Shoulder: flexion, abduction<br>Elbow: flexion, extension<br>Wrist: dorsal and palmar flexion<br>Second MCP: flexion, extension | JAM Scale: grades 0–4, with 0 = no decrease in joint ROM, 4 = excessive decrease |
| LE | Hip: flexion, extension, abduction, rotation (external, internal)<br>Knee: flexion, extension<br>Ankle: plantar- and dorsiflexion | |
| Muscle Strength | | |
| UE | Shoulder: flexors, abductors<br>Elbow: flexors, extensors<br>Wrist: flexors, extensors<br>Fingers: flexors | MRC scale: grades 0–5, with 0 = no muscle activity, 5 = normal muscle strength |
| LE | Hip: flexors, extensors<br>Knee: flexors, extensors<br>Ankle: plantar- and dorsiflexors | |
| Disability | | |
| PEDI | Functional skills<br>Caregiver assistance<br>3 domains: self-care, mobility, social function | Healthy children > 7.5yr old = 100 |
| Ambulation | Nonwalking<br>Walking: therapy, household, neighborhood, community walking with or without crutches | Bleck's classification: 1–9, with 1 = nonwalker and 9 = community walker without use of crutches |
| Perceived Competence | | |
| Quality of Life | | |
| SPPC | Subscales<br>Age 8–12yr: scholastic competence, social acceptance, athletic performance, physical appearance behavioral conduct, global self-worth<br>Age 12–16yr: all of the above, plus romance, friendship | 1 = Strongly negative<br>2 = Fairly negative<br>3 = Fairly positive<br>4 = Strongly positive |

Abbreviations: MCP, metacarpophalangeal; MRC, Medical Research Council; PEDI, Pediatric Evaluation of Disability Inventory; SPPC; Self-Perception Profile for Children.

**Table 2: Patient Characteristics Related to the Different Types of OI**

| | Type I ($n$ = 17) | Type III ($n$ = 11) | Type IV ($n$ = 12) |
|---|---|---|---|
| Age (yr mean ± SD) | 12.0 ± 3.3 | 11.6 ± 2.4 | 14.5 ± 3.1 |
| Joint ROM | | | |
| Arms* | 0.5 (.06–1.5) | 1.75 (1.25–2.8) | 1.25 (1.25–1.75) |
| Legs* | 0 (0–0.7) | 3.3 (2.7–3.7) | 1.7 (0.5–3.0) |
| Muscle strength | | | |
| Arms† | 4.5 (4.2–4.5) | 3.5 (3.0–3.6) | 4.5 (3.6–4.5) |
| Legs† | 4.8 (4.0–4.9) | 3.6 (2.6–3.7) | 3.8 (3.5–4.0) |
| PEDI | | | |
| Mobility‡ | 91.7 (65.6–100) | 54.8 (44.3–60) | 67.4 (61.2–75.5) |
| Self-care‡ | 100 (100–100) | 81.4 (66–100) | 100 (93.0–100) |
| Social function‡ | 100 (100–100) | 100 (96.3–100) | 100 (100–100) |
| Caregiver assistance | | | |
| Mobility‡ | 100 (75.2–100) | 58.8 (49.8–66.7) | 100 (72.7–100) |
| Self-care‡ | 100 (80.4–100) | 76.7 (62.2–89.7) | 100 (79.5–100) |
| Social function‡ | 100 (100–100) | 100 (100–100) | 100 (100–100) |
| Level of ambulation¶ | 9 (4–9) | 2 (2–5) | 5 (2–6) |

NOTE. Values except age presented as median and P25–P75.
* Joint ROM arm and legs: score 0 to 4 (score 0 = normal; score 4 = maximally decreased).
† Muscle strength arm and legs: score 0 to 5 (score 0 = no contractions; score 5 = contraction against max resistance).
‡ PEDI: scaled score in healthy children > 7.5 years of age = 100.
¶ Ambulation level: score 1 to 9 (score 1 = nonwalker; score 9 = community walker without the use of crutches).

differences between the left and right extremities were found, the data of the left extremities were presented.

Functional skills (functional limitation, disability domain) were measured using the Pediatric Evaluation of Disability Inventory[16] (PEDI) which is a validated and reliable questionnaire used to measure functional skills and caregiver assistance in 3 domains (self-care, mobility, social function). Ambulation was scored according to Bleck[17] and classified as nonwalking or walking (table 1).

Perceived competence (QOL domain) was measured using the Harter Self-Perception Profile for Children,[18] which has been cross-culturally validated for Dutch children (table 1).[19]

### Data Analysis

Data regarding joint ROM, muscle strength, functional ability, and perceived competence are presented as the median and the 25th and 75th percentiles (P25, P75). Differences in perceived competence between the types of OI were analyzed using the Mann-Whitney $U$ test. Correlation analysis between the joint ROM, muscle strength, functional ability, caregiver assistance, walking capability, and perceived competence was performed using Spearman's correlation coefficients; $p$ values less than .05 were considered statistically significant. For statistical analysis we used the SPSS package,[b] version 6.01.

## RESULTS

### Impairment, Functional Limitation, and Disability

Patient characteristics, regarding joint ROM, and muscle strength of the arms and legs, functional ability, and the amount of caregiver assistance, as well as the level of ambulation are presented in table 2. In children with type I, joint ROM and muscle strength were almost comparable to the healthy population. In most of these children, the level of ambulation was normal and the functional skills, except for the mobility items, were also comparable with the healthy population. In children with type III, a severe decrease in joint ROM was measured, especially in the LEs, and muscle strength was severely decreased in both UEs and LEs. The median level of ambulation was exercise walking with the use of crutches, and the functional skills, especially in the self-care and mobility domain, were severely decreased. In children with type IV, joint ROM and muscle strength decreased, especially in the LEs.

**Table 3: Median (P25–P75) Perceived Competence and Comparison Between the Different Types of OI***

| Items SPPC | Type I ($n$ = 17) P50 (P25–P75)† | Type III ($n$ = 11) P50 (P25–P75)† | Type IV ($n$ = 12) P50 (P25–P75)† | I vs III ($p$ value) | I vs IV ($p$ value) | III vs IV ($p$ value) |
|---|---|---|---|---|---|---|
| Scholastic competence | 3.1 (2.8–3.5) | 3.3 (2.6–3.5) | 3.0 (2.5–3.4) | 1.0 | 0.6 | .02‡ |
| Social acceptance | 3.1 (2.6–3.8) | 3.6 (2.7–3.8) | 3.1 (2.3–3.4) | 0.8 | 0.2 | 0.4 |
| Athletic competence | 2.2 (1.3–2.8) | 2.6 (2.0–3.8) | 2.6 (2.1–2.8) | 0.4 | 0.5 | 0.9 |
| Physical appearance | 3.0 (2.5–3.8) | 3.2 (2.8–3.6) | 2.7 (2.3–3.1) | 0.6 | 0.3 | .04‡ |
| Behavioral conduct | 3.2 (2.5–3.6) | 3.0 (2.6–3.3) | 2.8 (2.6–3.0) | 0.7 | 0.5 | 0.6 |
| General self-worth | 3.5 (3.0–4.0) | 3.8 (3.2–4.0) | 3.4 (2.9–3.8) | 0.4 | 0.5 | 0.2 |
| Romance | 2.8 (2.5–3.1) | 2.4 (1.3–2.6) | 3.0 (2.3–3.2) | .04‡ | 0.7 | 0.1 |
| Friendship | 3.5 (3.2–3.7) | 3.2 (2.5–3.4) | 3.8 (3.3–3.9) | 0.2 | 0.2 | .03‡ |

* Score 1 = strongly negative perceived competence; score 2 = fairly negative perceived competence; score 3 = fairly positive perceived competence; score 4 = strongly positive perceived competence.
† P50 (P25–P75): 50th percentile (25th percentile/75th percentile).
‡ $p < .05$.

**Table 4: Correlations Between Impairment, Functional Limitations, Disability, and Perceived Competence**

| | Impairment: Level of Ambulation, Joint ROM, Muscle Strength | | | | FS and Disability: Functional Skills | | QOL: Perceived Competence | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | Ambulation | ROM arms | ROM legs | Muscle strength arms | Muscle strength legs | FS self-care | FS mobility | Scholastic competence | Social acceptance | Athletic performance | Physical appearance | Behavioral conduct | Global self-worth | Romance | Friendship |
| Ambulation | | 0.6 (<.001) | 0.8 (<.001) | −0.6 (<.001) | −0.8 (<.001) | .3 (NS) | 0.9 (<.001) | 0.3 (NS) | −0.2 (NS) | −.04 (NS) | 0.0 (NS) | 2 (NS) | 0.0 (NS) | 0.1 (NS) | 0.1 (NS) |
| ROM arms | | | 0.6 (<.001) | −0.5 (.001) | −0.5 (.002) | −0.2 (NS) | −0.5 (<.001) | 0.0 (NS) | 0.3 (NS) | −0.1 (NS) | 0.0 (NS) | −0.2 (NS) | 0.1 (NS) | −.02 (NS) | 0.1 (NS) |
| ROM legs | | | | −0.6 (<.001) | −0.7 (<.001) | −0.3 (NS) | −0.7 (<.001) | −0.2 (NS) | 0.1 (NS) | 0.1 (NS) | 0.0 (NS) | −0.3 (NS) | 0.1 (NS) | −0.4 (NS) | −0.3 (NS) |
| Muscle strength arms | | | | | 0.7 (.001) | 0.5 (<.001) | 0.8 (<.001) | 0.2 (NS) | −0.1 (NS) | 0.2 (NS) | −0.1 (NS) | 0.1 (NS) | 0.0 (NS) | −0.4 (NS) | 0.4 (NS) |
| Muscle strength legs | | | | | | 0.3 (.03) | 0.8 (<.001) | 0.1 (NS) | −0.2 (NS) | −0.2 (NS) | −0.1 (NS) | 0.2 (NS) | −0.1 (NS) | −0.2 (NS) | 0.0 (NS) |
| FS self-care | | | | | | | 0.5 (.001) | 0.0 (NS) | 0.0 (NS) | −0.2 (NS) | −0.2 (NS) | −0.1 (NS) | −0.2 (NS) | 0.1 (NS) | 0.1 (NS) |
| FS mobility | | | | | | | | 0.2 (NS) | −0.2 (NS) | 0.0 (NS) | 0.0 (NS) | −0.1 (NS) | −0.1 (NS) | 0.2 (NS) | 0.3 (NS) |
| Scholastic competence | | | | | | | | | 0.5 (.02) | 0.3 (NS) | 0.4 (.02) | 0.4 (.007) | 0.4 (NS) | 0.1 (NS) | 0.1 (NS) |
| Social acceptance | | | | | | | | | | 0.4 (NS) | 0.4 (.006) | 0.0 (NS) | 0.4 (NS) | 0.1 (NS) | 0.4 (NS) |
| Athletic performance | | | | | | | | | | | 0.3 (.03) | −0.2 (NS) | 0.3 (.04) | 0.7 (.001) | 0.4 (NS) |
| Physical appearance | | | | | | | | | | | | 0.3 (NS) | 0.6 (<.01) | 0.4 (NS) | 0.3 (NS) |
| Behavioral conduct | | | | | | | | | | | | | 0.4 (.01) | 0.1 (NS) | 0.0 (NS) |
| Global self-worth | | | | | | | | | | | | | | 0.3 (NS) | 0.2 (NS) |
| Romance | | | | | | | | | | | | | | | 0.7 (.001) |

NOTE. *p* values are presented in parentheses.
Abbreviation: FS, functional skills; NS, not significant.

Only in the mobility domain of the PEDI was a decrease measured, in comparison with healthy children.

### Perceived Competence

In patients with OI type I, fairly negative perceived competence was measured in the subscale athletic performance, whereas in OI type III, fairly negative perceived competence was measured in the romance subscale (table 3). In the other subscales of types III and IV and all subscales of type IV, fairly to strongly positive perceived competence were measured. Scholastic competence in type III increased significantly compared with type IV ($p = .02$). Physical appearance in type III increased significantly compared with type IV ($p = .04$). Children with OI type I perceived significantly more romance than type III ($p = .04$), whereas in type IV friendship was significantly more perceived compared with type III ($p = .03$). In all other subscales no significant differences between the different types were observed.

### Correlations Among Impairment, Functional Limitations, Disability, and Perceived Competence

The level of ambulation correlated highly with joint ROM and muscle strength (ambulation/arms: $r = -0.6$, $p < .001$, $r = 0.8$, $p < .001$, respectively; ambulation/legs: $r = -0.6$, $p < .001$, $r = 0.8$, $p < .001$, respectively) (table 4).

Functional skills regarding self-care correlated significantly with strength of the arm muscles ($r = 0.5$, $p < .001$). Functional skills regarding mobility correlated table 4 significantly with the level of ambulation ($r = 0.9$, $p < .001$); ROM of arms and legs ($r = -0.5$, $p < .001$, $r = -0.7$, $p < .001$ respectively), and muscle strength of both arms and legs (both $r = 0.8$, $p < .001$) (table 4).

Regarding the perceived competence, scholastic competence corelated moderately with social acceptance, physical appearance, and behavioral conduct. Social acceptance correlated moderately with physical appearance, and athletic performance correlated highly significantly with romance. Physical appearance correlated highly significantly with global self-worth, and romance correlated highly significantly with friendship. No correlations were found (1) between joint ROM and athletic performance and physical appearance, (2) between muscle strength and athletic performance or physical appearance, or (3) between either of the functional skills, and the subscales of the perceived competence.

## DISCUSSION

The observations in the impairment and functional limitation and disability domains were comparable with current literature. Joint ROM and muscle strength differed significantly between the different types of OI. Functional skills also differed significantly between the types, especially regarding mobility and self-care.[10,20] In children older than 7.5 years of age, a moderate to good correlation has been found between joint ROM, muscle strength, and functional skills.[10]

A prospective study with a follow-up of 1 year showed that almost all impairment parameters did not change significantly over time, whereas some disability parameters seemed to improve significantly.[11] In addition, a prospective study in which 49 children with different types of OI were followed for 4 years showed that almost all impairment parameters did not change significantly over time, whereas some disability parameters improved significantly. Progression in the level of ambulation in children with OI occurred in 35%, especially in younger children: in 17%, regression in level of ambulation was found, especially in girls who reach puberty, and in 48%, stabilization

in the level of ambulation was found. So, in 65% of the these children, a plateau phase in level of ambulation was observed.[21]

In our study, we found that overall perceived competence in children with OI was fairly to strongly positive, without significant differences between the different types of OI. The perceived athletic competence in type I was below average, whereas in types III and IV it was slightly above average. A possible explanation might be that children with OI types III and IV use (electric) wheelchairs for sport activities, whereas children with type I seldom use wheelchairs in sports. Another explanation may be that children with OI type I compare themselves with unaffected children, whom they cannot emulate and, therefore, have different expectations. In the most severe types of OI, skeletal disproportions are present and muscle strength and joint ROM as well as functional skills are decreased, when compared with healthy children. Although improvement in level of ambulation is only possible in a small group of children, mostly in the less severe types, children with types III and IV have a fairly to strongly positive perceived competence.

Other studies observed in children with chronic diseases showed no correlation between impairment and disability parameters on 1 side and the QOL parameter on the other. Gulmans et al[22] compared 2 groups of children with cystic fibrosis at the level of impairments and QOL. No significant differences were measured in perceived competence between an age-matched group of children, 1 group with severe obstructive pulmonary problems, and 1 group with mild pulmonary problems. This finding is in agreement with current literature on perceived competence and chronicity.[23-26]

The common understanding of a good QOL implies being in good health and experiencing subjective well-being and life satisfaction.[27] Conversely, one can argue that if people have disabilities, they cannot be considered to be in good health nor to possess a high level of life satisfaction. People with disabilities are assumed to be limited in function and role performance and quite possibly stigmatized and underpriviliged.[28] In practice, however, the patient's perceptions of personal health, well-being, and life satisfaction are often discordant with their objective health status and disability.[29]

As outlined by Berg et al,[29] patients tend to rate cognitive, emotional, and social items more highly than items related to physical health. This may indicate that perceived competence only measures an aspect of QOL, an aspect that is apparently not heavily influenced by disability.

Another important issue in the discussion of the failure of disability to correlate with perceived competence is adressed by Aasland and Diseth,[30] who conclude that the Harter Self-Perception Profile for Adolescents has limited ability to identify chronically diseased adolescents with adverse psychosocial outcome.

## CONCLUSION

Overall QOL may be less reflective of health, whereas health-related QOL reflects personal patient values, and health status measures provide information about specific areas of health. Therefore, we suggest measuring all these areas in children with OI when evaluating or predicting future health-related and patient-perceived outcomes.

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**Suppliers**

a. Mathys Medical Nederland BV, Huis ter Heideweg 28, 3705 LZ, Zeist, The Netherlands.
b. SPSS Inc, 233 S Wacker Dr, 11th F1, Chicago, IL 60606.