Health status according to the IFC in people with short stature due to skeletal dysplasia

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BACKGROUND Height of a person with skeletal dysplasia differs significantly from the normal values and is below 140 to 150 cm in adulthood. According to medical grounds, there are approximately 1,000 people defined as having a short stature in Finland. The most common diagnoses include diastrophic dysplasia, achondroplasia and cartilage-hair hypoplasia. These bone dysplasias are the largest group of diseases causing short stature in Finland and the target of this study. **METHODS** The comprehensive musculoskeletal postacute ICF core set: 72 categories of functioning operationalized to questions from:

THE PURPOSE OF THIS STUDY was to gain new information concerning the functioning and disability of people with short stature as well as their challenges concerning accessibility and equality. In addition, we tested the feasibility of the operationalized questions of the comprehensive musculoskeletal post-acute ICF core set, and evaluated associations between functioning and accessibility in individuals with short stature.

- the Patient-Reported Outcomes Measurement Information System (PROMIS®)
- the Finnish Regional Health and Well-being Study
- formulated from the ICF category by the research group.

The draft questionnaire was completed according to the iterative process with short stature people and feed-back based on their interviews. The final inquiry was sent to 384 adults with skeletal dysplasia due to the selected diagnosis who were identified from the patient register data of Children's Hospital, Helsinki University Hospital. In addition, social media and patient organizations were used to inform about the study. Age, gender and place of residence matched controls were also recruited.

RESULTS 80 short statured adults were analyzed and compared with 55 age, gender and place of residence matched controls.

In short stature people with skeletal dysplasia, 30 % had some difficulties and 50 % had much difficulties picking up products on a grocery store or could not do the task. Also 23 % had some difficulties and 44 % had much difficulty using ticket, bank or other automats.



	Skeletal dysplasia (n = 80)	Control (n = 55)	P-value
Women, n (%) / men, n (%)	/ (81) / 15 (19)	46 (84) / 9 (16)	0,722
Age, years, mean (SD)	43 (15)	46 (15)	0,261
Height, cm, mean (SD)	127 (12)	166 (8)	<0,001
Cordage, cm, mean (SD)	107 (18)	163 (27)	<0,001
Weight, kg, mean (SD)	51 (13)	73 (15)	<0,001

Mobility and daily living activities were significantly more difficult for short stature people with skeletal dysplasia compared to control subjects (p<0,001). Most limitations were in vigorous activities, such as:

- running
- lifting heavy objects
- participating in strenuous sports
- walking more than a mile (1.6 km)
- doing two hours of physical labor
- in lifting or carrying groceries
 in bending, kneeling, or stooping.

There was no significant differences in psychological well-being between the two groups. However, pain was moderately, severely or very severely present in 58 % of short stature people with skeletal dysplasia, whereas the number was 16 % in control subjects (p<0,001).

There was no significant difference in items in the field of social environment, except more dissatisfaction in participating in family activities (p=0,020).

Subjects in both groups were comparable in their satisfaction to work (p=0,193), with a lot or quite a lot of satisfaction in 64 % of people with skeletal dysplasia and in 78 % of control subjects. Short stature people with skeletal dysplasia were more dissatisfied in their ability to participate in activities in a club, organization, association, hobby group or spiritual community (p<0,001). They had also more difficulties in doing usual work (p=0,006) or regular leisure activities with others (p=0,023) compared with control subjects.

CONCLUSIONS Short stature restricts func-

tioning extensively within the different sectors of functional capacity but accessibility of the built environment and availability of equal services could promote an equal life for short-statured people and for people with other functional difficulties.

- They also had difficulties in mobility tasks such as:
- using public transportation with or without luggage
- standing unsupported for 30 minutes
- in walking usual stairs.









