Quality of Life in Patients with Congenital Short Femur

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Purpose. To assess the quality of life in persons with a congenital short femur.
Materials and methods. This was a cross-sectional study. The study population was 92 patients diagnosed since 1969 with congenital short femur. Inclusion criteria were: patients should be over 15 years of age and possess a known address or telephone number. Quality of life was determined from May to October 2003 by means of telephone interviews, carried out after obtaining informed consent, using Health Questionnaire SF-12. The results were compared to those obtained in a reference population.
Results. Only 39 patients participated in the study, of whom 48.7% were male and 51.3% were female. Mean age: 26.2 years (men: 24.8 years; women: 27.6 years). No differences were detected between men and women, nor across different age groups, with reference to the mean physical (PCS) and mental (MCS) variables. One hundred percent of the population had a normal MCS and 20.5% had an abnormal PCS.
Conclusion. Quality of life based on PCS is abnormal in younger patients. It is surprising that MCS should be normal in all patients. This could be due to the fact that this condition is present from birth and therefore does not have such a strong psychosocial impact as other conditions.

Key words: *quality of life, congenital anomalies, disability assessment.*

Calidad de vida en pacientes con fémur corto congénito

Objetivo. Valorar la calidad de vida en personas con defecto congénito de fémur.
Material y método. Se trata de un estudio transversal. La población está formada por 92 pacientes diagnosticados de fémur corto congénito desde 1969. Los criterios de inclusión fueron tener 15 años de edad o más y tener datos de domiciliación o de teléfono de contacto. La calidad de vida se midió con el cuestionario de salud SF-12 aplicado de mayo a octubre de 2003 telefónicamente, previa petición de consentimiento informado. Los resultados fueron comparados con una población de referencia.
Resultados. Sólo 39 pacientes se incluyeron en el estudio. La media de edad fue de 26,2 años (hombres 24,8 años; mujeres 27,6 años). El 48,7% eran hombres y el 51,3% mujeres. No se detectaron diferencias en las medias de los componentes físico (PCS) y mental (MCS) entre hombres y mujeres, ni entre los distintos estratos de edad. El 100% obtuvo una puntuación MCS considerada como normal y el 20,5% obtuvo una puntuación PCS anormal.
Conclusión. La calidad de vida de estos pacientes para el componente físico es anormal en los pacientes más jóvenes. Sorprende que el componente mental sea normal en todos. Podría deberse al hecho de ser un trastorno que se padece desde el mismo momento del nacimiento, y no tener un impacto psicosocial tan fuerte como con otro tipo de problemas.

Palabras clave: *calidad de vida, anomalías congénitas, evaluación de la discapacidad.*

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Received: June 2005.
Accepted: November 2005.

It is currently widely accepted that the impact of disease on patients cannot be fully described by means of objective health measures, such as a biopsy. Consideration of other factors that could be called subjective, such as pain, functional capacity or emotional well-being are considered indispensable when making a global assessment of an individual's health[1]. This understanding has led to the development of a field of research known as «health-related quality

of life» (HRQL)[2]. At the same time studies that evaluate the effectiveness of medical procedures increasingly use HRQL scores as a variable[3].

When evaluating an orthopedic technique one must not only take into account the clinical functional result obtained but also the patients' expectations as well as any changes in health status perceived by them[4]. In this connection, studies have been published that measure quality of life by means of the SF-12 or similar questionnaires, especially in elderly patients with a degenerative disease[5-7]. Nevertheless, no study has been carried out that assessed the quality of life of persons with congenital pathologies like the congenital short femur[8].

The term «congenital short femur» was adopted at a symposium held in Dallas in 1998 at the *Shriners Hospital for Crippled Children*, and has been used by several authors[9,10,11]. The term covers all known forms of congenital defect, which includes different femoral conditions ranging from a minimal shortening to complete absence, as well as modifications in the proximal, diaphyseal and distal parts of the bone. Moreover, defects are not limited to the femur, but can also affect other bones in the limb involved and even in other regions of the skeleton.

This paper sets out to look into the perceived health status or quality of life of patients that suffer from a congenital anomaly such as the congenital short femur. In these pathologies the ultimate goal of treatment is to afford patients a quality of life as close to normal as possible from childhood.

MATERIALS AND METHODS

This was a cross-sectional study. The population was made up of 92 patients diagnosed with a congenital short femur, treated at the department of pediatric orthopedics of two hospitals in the Madrid region (La Paz University Hospital and Ramón y Cajal Hospital) from 1969 onwards. The inclusion criteria were to be 15 or over and the provision of an address or contact telephone number. The study was carried out between May and October 2003. Patients were contacted by telephone. Before administering the questionnaire, patients were asked to give their informed consent to participating in the study. Interviews were given by a single interviewer who administered version 1 of the SF-12 questionnaire to evaluate the patients' quality of life.

For processing and analyzing data, we used version 11.0.0 of the SPSS statistics software. To calculate quality of life on the basis of the SF-12 questionnaire, two scores were used: the physical health score (PCS-12), which assesses the patient's physical role, bodily pain and general health status, and the mental health score (MCS-12), which evaluates vitality, social function, emotional role and mental health. The calculation algorithm, as well as the questionnaire itself, were supplied by the Municipal Institute for Medical Research of Barcelona[12].

Scores for the 8 dimensions of version 1 of the SF-12 cannot be interpreted in themselves since they have no meaning as they lack an absolute value; they must be transformed into uniform significance scales that are common to all dimensions so that they can be compared to reference population values or norms. To do that it was necessary to carry out non-lineal transformations, such as percentile calculations. Percentiles make it possible to place individual scores in the distribution of the formative (reference) group, which allows us to define that value's < normality > or < abnormality > by means of a cut-off point (P5) (lower ends of the distribution) of the subjects in a group. This means that scores above the cut-off point (P5) of the reference group will be normal[13]. For comparisons across different groups on the basis on gender, age and summation averages, the Mann-Whitney and Kruskal-Wallis non-parametric U tests were used.

RESULTS

Of the 92 clinical records reviewed only 39 fulfilled the inclusion criteria. Therefore, the final amount of patients included in the group was 39. The 39 patients agreed, by means o fan informed consent form, to answer the 12 questions in the SF-12 questionnaire.

The mean age of the respondents was 26.2 ± 6.6 years (range: 15-40). 48.7% (19) were men and 51.3% (20) women. Mean age of men was 24.8 ± 6.6 years and that of women 27.6 ± 6.4 years. No statistically significant differences were detected with respect to age between men and women ($p = 0.178$).

The mean PCS score for women was 48,8 (confidence interval [CI]: 95%) (range: 45.0-52.6) and the mean MCS score was 53.1 (CI 95%) (range: 50.4-55.7). The mean PCS score for men was 48.3 (CI: 95%) (range: 45.1-51.6) and the mean MCS score was 55.1 (CI 95%) (range: 53.3-56.9).

No statistically significant differences were detected between the mean PCS ($p = 0.869$) and MCS ($p = 0.252$) summarized scores; nor were differences found when comparing different age groups (Table 1).

Tables 2 and 3 show the percentages of the scores considered to lie in the normality range, as established by comparison with the reference population, for the physical and mental components. One-hundred percent of respondents obtained a MCS score higher than the P5 that defines normality. As regards the PCS score, 20,5% (8) of respondents (3 females and 5 males) obtained a score below the cut-off point (P5), which means that they obtained an abnormal score. Of the 8 patients with an abnormal score, 5 belong to the younger age group (15 to 24 years) and 3 to the intermediate age group (25 to 34 years).

Table 1. Differences in physical and mental scores according to age

Summarized score	Age (years)	n	Mean (SD)	P
MPS	15 to 24	19	48.0 (7.4)	
	25 to 34	14	49.2 (6.3)	
	35 to 44	6	49.7 (9.0)	
Total		39		< 0.05
MMS	15 to 24	19	56.2 (3.6)	
	25 to 34	14	51.9 (6.7)	
	35 t 44	6	52.1 (6.7)	
Total		39		< 0.05

MPS: mean physical score; MMS: mean mental score; SD: standard deviation.

Table 2. Percentage of scores considered in the range of normality established with the reference population for the MPS component

MPS	Gender	Age	Reference Normality*	Normality % Study
	Males	15 to 24	> 44.1	81,1% (9 out of 11 patients)
		25 to 34	> 44.3	50% (3 out of 6 patients)
		35 to 44	> 35.5	100% (2 out of 2 patients)
	Females	15 to 24	> 41.4	62.5% (5 out of 8 patients)
		25 to 34	> 39.5	100% (8 out of 8 patients)
		35 to 44	> 33.4	100% (4 out of 4 patients)

*«Reference normality»: percentile 5 (P5). MPS: mean physical score.

Tabl3 3. Percentage of scores considered in the range of normality established with the reference population for the MMS component

MMS	Gender	Age	Reference Normality*	Normality % Study
	Males	15 to 24	> 35.2	100% (11out of 11 patients)
		25 to 34	> 37.0	100% (6 out of 6 patients)
		35 to 44	> 37.7	100% (2 out of 2 patients)
	Females	15 to 24	> 27.8	100% (8 out of 8 patients)
		25 to 34	> 27.9	100% (8 out of 8 patients)
		35 to 44	> 28.4	100% (4 out of 4 patients)

*«Reference normality»: percentile 5 (P5). MMS: mean mental score.

DISCUSSION

As was to be expected, the quality of life of these patients as regards the physical component was not normal for all of them. However, we were surprised to observe that the mental score was normal in all cases. This means that the mental score was better than the physical score for these patients, which could be attributed to the fact that this is a disorder that patients are born with and its psycho-social impact for them is not as hard as that of other types of pathologies or accidents[14].

The reference population values stratify age starting at 18. In this study, patients under 18 (15 and 16 years respectively) were included in the 18-to-24 age group since the potential distortion in the results would be practically negligible.

We chose version 1 of the SF-12 questionnaire because of the multiple advantages it offered, given the great number of available studies[15-17] that compare it with its predecessor (SF-36) that show its reproducibility and validity. These are the result of its multidimensional nature (to assess an individual's general health status), its frequent use in studies dealing with chronic disorders, its widespread use in many countries, its availability in Spanish[11] and its conciseness and ease of use (it can be administered in approximately 2 minutes).

Another very important advantage is that several reference population values can be used to compare the results obtained. This is especially interesting for a study like this one where only one measurement was taken over a period of time.

We should however mention that version 1 of the SF-12 does not allow results to be broken down into different dimensions; we were only able to present global added-up results of the functional and mental components. Another disadvantage are losses to follow-up (57,6%); even if our initial population was 92 clinical records, only 39 could be tracked. These losses were due to changes of address and telephone number.

To date no quality of life studies have been published for congenital malformations, which means that we cannot compare results obtained in this study with any others. The reason why we carried out a cross-sectional study is that this type of study is the most appropriate one when knowledge on a certain subject, in this case quality of life of patients with a congenital abnormality, is scarce. With this paper our only aim was to shed some light on an increasingly popular subject such as health-related quality of life.

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Conflict of interests: We, the authors, have not received any economic support to carry out this study. Nor have we signed any agreement with any commercial firm to receive benefits or fees. On the other hand, no commercial firm has provided nor will provide economic support to non-profit foundations, educational institutions or any of the other non-profit organizations that we are members of.