Instrumentos de avaliação da qualidade de vida em pacientes com Distrofia Muscular de Duchenne: uma revisão sistemática

Evaluation tools for quality life improvements for patients with Duchenne Muscular Dystrophy: a systematic review

Instrumentos de evaluación de la calidad de vida en pacientes con Distrofia Muscular de Duchenne: una revisión sistemática

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Resumo

Objetivo: identificar os instrumentos de avaliação utilizados para mensurar a qualidade de vida em pacientes com Distrofia Muscular de Duchenne. **Metodologia**: foi realizada uma revisão sistemática dos artigos publicados de 2007 a 2017 sobre os instrumentos de avaliação de qualidade de vida em pacientes com Distrofia Muscular de Duchenne, pesquisados em três bases de dados: SciELO, PubMED e LILACS. **Resultados**: seis artigos atenderam aos critérios de inclusão e os instrumentos referidos foram: Life Satisfaction Index for Adolescents; Quality of Life Evaluation Scale; Medical Outcomes Study 36; World Health Organization Quality of Life Instrument; Health Related Quality of Life Questionnaire for Children and Young People and their Parents e Pediatric Quality of Life Inventory. **Conclusões**: os instrumentos para avaliação da qualidade de vida em pacientes com Distrofia Muscular de Duchenne são essenciais para determinar e apresentar uma efetividade no

tratamento voltado para suas principais dificuldades. No entanto, a falta de uma escala específica para esse diagnóstico interfere nos escores reais da qualidade de vida desses pacientes.

Palavras-chave: Distrofia Muscular de Duchenne; Qualidade de vida; Questionários; Instrumentos.

Abstract

Introduction: Duchenne Muscular Dystrophy (DMD) is characterized by membrane dissociation, resulting in the breakdown of the musculoskeletal fiber. **Objective:** to identify the assessment tools used to measure the quality of life in patients with DMD. **Methodology**: A systematic review of articles published from 2007 to 2017 on QOL assessment tools in patients with DMD was conducted in the SciELO, PubMED and LILACS databases. **Results:** 6 articles met the inclusion criteria, using the QOL assessment tools; Life Satisfaction Index for Adolescents; Quality of Life Evaluation Scale; Medical Outcomes Study 36; World Health Organization Quality of Life Instrument; Health Related Quality of Life Questionnaire for Children and Young People and their Parents e Pediatric Quality of Life Inventory. **Conclusions:** the tools for the evaluation of quality of life in patients with Duchenne Muscular Dystrophy (DMD) are essentials to determinate and to present an effective treatment focused on patient's priorities and their main difficulties. However the lack of a validated scale specifically focused on this diagnostic interferes in the real score of those patients quality of life.

Keywords: Duchenne muscular dystrophy; Quality of life; Questionnaire; Instruments.

Resumen

Objetivo: identificar los instrumentos de evaluación utilizados para medir la calidad de vida en pacientes con distrofia muscular de Duchenne. **Metodología**: se realizó una revisión sistemática de artículos publicados entre 2007 y 2017 sobre los instrumentos para evaluar la calidad de vida en pacientes con distrofia muscular de Duchenne, investigados en tres bases de datos: SciELO, PubMED y LILACS. **Resultados**: seis artículos cumplieron con los criterios de inclusión, utilizando los siguientes instrumentos de evaluación: Índice de Satisfacción de Vida para Adolescentes; Escala de evaluación de calidad de vida; Estudio de resultados médicos 36; Instrumento de calidad de vida de la Organización Mundial de la Salud; Cuestionario de calidad de vida relacionado con la salud para niños y jóvenes y sus padres e inventario de calidad de vida pediátrica. **Conclusiones**: los instrumentos para evaluar

la calidad de vida en pacientes con distrofia muscular de Duchenne son esenciales para determinar y presentar una efectividad en el tratamiento dirigido a sus principales dificultades. Sin embargo, la falta de una escala específica para este diagnóstico interfiere con las puntuaciones reales de calidad de vida de estos pacientes.

Palabras-clave: Distrofía Muscular de Duchenne; Calidad de vida; Cuestionarios; Instrumentos.

1. Introduction

The Duchenne Muscular Dystrophy (DMD), is a neuromuscular myopathy that causes progressive loss and muscle weakness, caused by a mutation on gen X, that leads to the absence, or lack of dystrophin protein and the continuous degeneration from the muscles fibers, as the extracellular matrix (Mah, 2016; Jacques et al., 2019).

The DMD affects ~0,1 a 1,8 out of 10.000 male individuals all around the world (Romitti et al., 2015; Zamani et al., 2016). The clinical manifestations are generally observed from 1 to 3 years old, with balance deficit, delayed march, difficulty in climbing stairs and progressive weakness in the inferior members, with the muscles around the calf, pelvis, and thigh, visibly more voluminous than the usual (Moraes et al., 2011; Ryder et al., 2017). Consequently, the individual develops a global muscular weakness with ambling loss typically around 12 years old (Jacques et al., 2019). The progressive muscular strength loss, the limitation in performing activities of daily living (ADLs) and the increase in the body mass index (BMI) affects the psychological well-being in high weight individuals, changing their (QOL) in a negative way (Fujino et al., 2016).

QOL can be understood by a set of situations and sensations that the individuals subjectively describe, from personal and collective values, cultural perspectives, acquired knowledge, and experiences (Burckhardt & Anderson, 2003). The World Health Organization (WHO) defines QOL as a perception that de individuals include their achievements, standard expectations and worries (Organization, 1995). For a greater analysis, the tools to assess QOL have been utilized to complement clinical standards besides being useful to evaluate the service's quality, health care's needs and the interventions' effectiveness (Souza et al., 2014).

The assessment questionnaires can work as a tool to collect objective and subjective information. For such, these tools need to be easily manageable, having low operational expenses, being standardized, easy to reproduce, reliable, and susceptible in detecting alterations promoted by the disease or treatment (Booth et al., 2002).

Those measures create a perspective from the patients about the need of healthcare and their choices for treatments (Carr & Higginson, 2002). However, the challenge in QOL's assessment depends on its singularity for the individuals that many times are considered pre-selected domains, and because of that, they are evaluated in general health status and not by QOL (Gierlaszyńska et al., 2016). In this context, the study's objective was to identify the evaluation tools used to measure the quality of life in patients with DMD.

2. Methodology

It is based on a systematic review, quantitative study (Pereira et al., 2018) guided by the following question: "what are the quality of life tools used in individuals diagnosed with Duchenne Muscular Dystrophy (DMD)?". In this review, three databases were used: PubMed, SciELO, Lilacs to for the biographic search, using the Health Science Descriptors (DeCS) and Medical Subject Headings (MeSH), combined in the following way: Duchenne Muscular Dystrophy *and* Qualidade de vida; Distrofia Muscular de Duchenne *and* Calidad de Vida; Muscular Dystrophy, Duchenne *and* Quality of Life; Muscular Dystrophy, Duchenne *and* Life Quality; Muscular Dystrophy, Duchenne *and* Health-Related Quality of Life.

The inclusion criteria for the studies selection were: searches that used (1) questionnaires and/or scales to assess QOL in patients diagnosed with DMD, (2) published articles from 2007 to 2017, (3) articles in Portuguese, English and Spanish. The definition in the last ten years occurred for being a huge and current period that contains the last studies related to the topic. The pre-established exclusion criteria were: (1) duplicated articles, (2) editorial, (3) letters, (4) comments, (5) dissertations or thesis and (6) articles that didn't have as the main outcome the evaluation in QOL patients with DMD diagnoses without the use of the specific tools.

The articles were submitted to the Relevance Test I (RTI), through the reading of titles and abstracts. Posteriorly, the articles were submitted to the Relevancy Test II (RTII), all the articles being in the line-up (Table 1).

Table 1. Application form for Relevance Tests I and II.

| Application form for Relevance Test I		
Inclusion criteria	Yes	No
Does the study approach the quality of life of patients with Duchenne muscular dystrophy using the specific questionnaires?		
Was the article published from 2007 to 2017?		
Is the article in English, Portuguese or Spanish?		
Exclusion criteria		
Is it editorial, letter, comment, review, isolated case report, dissertation, or thesis?		
Application form for Relevance Test II		
Inclusion criteria		
Did the study specify the quality of life assessment tool for Duchenne Muscular Dystrophy patients?		
Did the article use quality of life assessment tools?		
Exclusion criteria		
Does the article have as its main objective the analysis of Duchenne Muscular Dystrophy patient's quality of life?		

Source: Own creation.

The risk of bias for the selected works were classified as low, uncertain or high based on the criteria established by the *Cochrane Colaboration*'s tool, through RevMan software (version 5.3, *The Nordic Cochrane Centre, The Cochrane Collaboration*, Copenhagen, Denmark, 2014). In this study, two independent researchers selected two studies following the eligibility's criteria. The divergences were solved in consensus with a third one. The methodology quality results were presented in graphs.

3. Results

In the three searched databases, 605 articles were identified. From those, 588 were discarded by their titles and abstracts due to duplicity and/or being related to other treatments and pathologies. From the 17 selected publications, 1 was excluded for being a review article, 2 articles weren't available for complete reading and 8 were excluded for not presenting as the main outcome the QOL evaluation in patients diagnosed with DMD by the specific tools (Figure 1).

Figure 1. Flowchart of the followed steps to select the article.
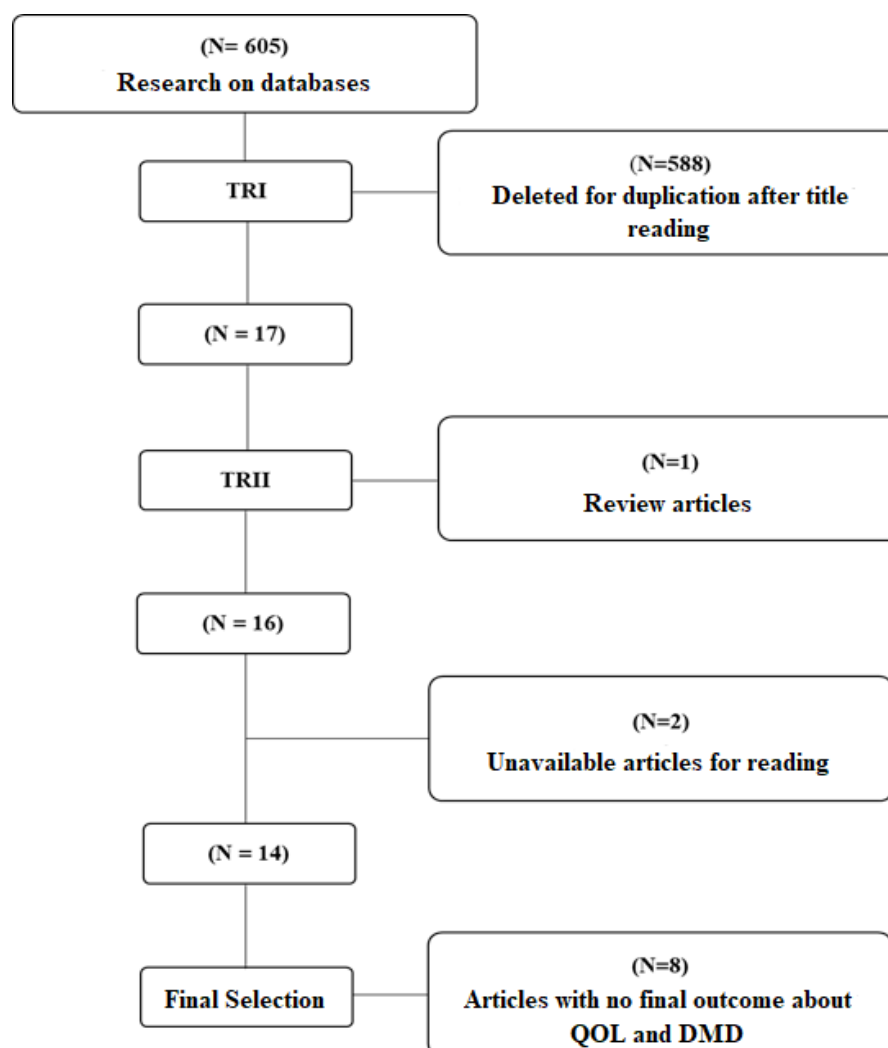
Source: Own creation.

Regarding the conjugate descriptors that were used for this search, "Dystrophy Muscular, Duchenne; Quality of Life", "Distrofia Muscular de Duchenne; Qualidade de Vida" and "Distrofia Muscular de Duchenne and Calidad de Vida" were the most found, respectively, corresponding to 33,05% (Table 2).

Table 2. Prevalence of the combined descriptors searched in the SciELO, PubMed and LILACS databases.

Descriptors	Found articles		Discarded articles		Utilized articles	
	N	%	N	%	N	%
Distrofia Muscular de Duchenne; Qualidade de Vida	3	0,49	2	0,34	1	16,6
Distrofia Muscular de Duchenne; Calidad de Vida	2	0,33	1	0,001	1	16,6
Dystrophy Muscular, Duchenne; Quality of Life	200	33,05	196	33,3	4	66,6
Dystrophy Muscular, Duchenne; Life Quality	200	33,05	200	34,01	0	0
Dystrophy Muscular, Duchenne; Health-Related Quality of Life	200	33,05	200	34,01	0	0
TOTAL	605	100	588	100	6	100

Source: Own creation.

The sample of this systematic review was composed by 6 studies that used the QOL evaluation tools in patients with DMD: (1) Life Satisfaction Index for Adolescents (LSI), (2) Quality of Life Evaluation Scale (AUQEI) qualitative and quantitative, (3) Medical Outcomes Study 36 (SF-36), (4) World Health Organization Quality of Life Instrument (WHOQOL-BREF), (5) Health Related Quality of Life Questionnaire for Children and (6) Young People and their Parents (KIDSCREEN-27) e Pediatric Quality of Life Inventory (PedQL).

Table 3 presents the studies' descriptions selected referring to the authors, title, year of publication, place of study, journal, language, objectives, sample profile, collecting data's tools and mainly results referenced to the QOL tools from patients with DMD.

Table 3. Selected articles referring to QOL assessment tools used in patients diagnosed with DMD.

AUTHOR/TITLE/YEAR/COUNTRY/LANGUAGE/JOURNAL	TOOL NAME	TARGET-AGE STUDY	NUMBER OF QUESTIONS	INTERVIEWED	INCLUDED DOMAINS
-Simon et al. -*Duchenne muscular dystrophy: quality of life among 95 patients evaluated using the Life Satisfaction Index for Adolescents* -2011 -Brasil -Inglês -Arquivos de Neuropsiquiatria	Life Satisfaction Index for Adolescents (LSI)	5 and 17 years	45	-Patients	- General well-being, interpersonal relationships, personal development, personal satisfaction and leisure and recreation
-Melo et al. -*Evaluation of the quality of life of children with Duchenne's*	- Autoquestionnaire Qualité de Vie Enfant Imagé	- average age group from 8-11 years old	26	-Patients	-Autonomy, leisure, function and family

progressive muscular dystrophy -2009 -Brasil -Inglês -Revista Brasileira de Promoção à Saúde	(AUQEI)				
-Lue et al. -*Quality of life of patients with Duchenne muscular dystrophy: from adolescence to young men* -2016 -Taiwan -Inglês -*Disability and Rehabilitation*	-Medical Outcomes Study 36 (SF-36) -World Health Organization Quality of Life (WHOQOL-BREF)	-Average age group of 15 years old	-SF-36: 36 itens -WHOQOL: 26 itens	-Patients	-SF-36: functional capacity, limitation due to physical aspects, pain, general status, vitality, social aspects, emotional aspects, and mental health -WHOQOL: physical, psychological, social relationships, and environment

-Zamani et al. -*The quality of life in boys with Duchenne muscular dystrophy* -2016 -Irã -Inglês -*Neuromuscular Disorders*	-KIDSCREEN-27	-Average group age of 12 years old	-27	-Patients with their caregivers	-Physical well-being, psychological, Family relationships and autonomy, social relationships and peers, school environment, and learning
-Landfeldt et al. -*Health-related quality of life in patients with Duchenne muscular dystrophy: a multinational, cross-sectional study* -2015	-*Pediatric Quality of Life Inventory* (PedsQL)	-Minimum group age of 5 years old	-34	-Patients with their caregivers	-General fatigue, disease aspects, treatment problems, friends and family interaction, worries, physical appearance, and

|				
-Alemanha, Itália, Reino Unido e Estados Unidos -Inglês				communication
-Uzarq et al. -*Health-Related Quality of Life in Children and Adolescents With Duchenne Muscular Dystrophy* -2012 -Texas -Inglês -*Pediatrics*	-*Pediatric Quality of Life Inventory* (PedsQL)	-Average group age of 10 years old	-34	-Patients with their caregivers	- General fatigue, disease aspects, treatment problems, friends and family interaction, worries, physical appearance, and communication

Source: Own creation.

Among the studies relating to DMD and the assessment tools for QOL, two of them (33,3%) were developed in Brazil, two (33,3%) in the United States and the others in Taiwan, Iran, Germany, Italy and United Kingdom, all published in English in different periods of time. In three studies (50%), the questionnaires were answered by the own patients and the others with the caregivers' help.

Figure 2 shows the risks of bias from the selected studies. All studies do not bring enough information to evaluate the bias existence in some items, in this way, it is observed that the methodology quality was considered moderate in the 4 evaluated domains.

Figure 2. Analysis graph of the risk of bias and applicability of the selected studies presented as percentages in all included studies.
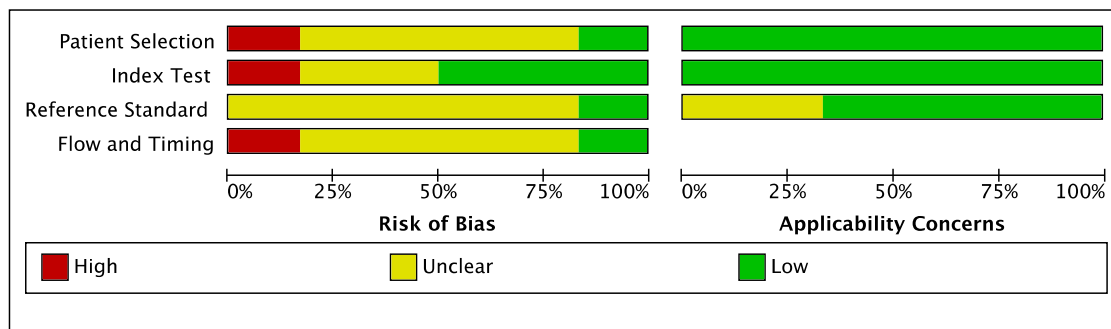
Source: Own creation.

3. Discussion

This systematic review emphasizes that in general, from the published studies that use questionnaires to assess QOL in DMD, just six of them answered the inclusion criteria. The QOL is associated to a complete physical, mental, and social well-being sensation. Although, to patients with DMD the QOL is harmed, making the patients learn to adapt to the limitations that are imposed by the disease's progression (Landfeldt et al., 2016).

The neuromuscular diseases cause physical restrictions that lead to adaptation needs, besides the psychological process on dealing with the pathology (Gierlaszyńska et al., 2016). Although there is some progress in diagnosis methods and treatments, attention to the psychological and environmental aspects in patients with DMD have been neglected, thus, the spread knowledge on the neuromuscular disease, as well as the social and economic factors, contribute to the interest in searches over QOL in DMD.

In this context, the questionnaires are precise and standardized searching tools, easy to apply, with objective and subjective questions, reliable reproduction in similar conditions, validity, sensibility, and reproducibility (Gierlaszyńska et al., 2016). According to Souza et al. (2014), the QOL indicators include generic tools for the QOL's global assessment and specific and personalized tools that quantify different life aspects for the individuals affected by some health condition or a specific disease.

LSI is a specific tool composed by 45 questions, used to evaluate patients with neuromuscular disturbances and it encompasses five domains: general well-being, interpersonal relationship, development, personal satisfaction, leisure and recreation (Simon et al., 2011). This questionnaire assesses the perception in QOL in teenagers between 12 and 19 years old with DMD (Reid & Renwick, 1994). According to Simon et al., (2011) when assessing QOL in 95 patients with DMD with ages between 5 and 17 years old, it was observed that there was not a QOL loss, even amid the older and are more affected children by DMD, probably for not excessively valorizing clinical aspects.

As for AUQEI, it consists of a questionnaire with 26 questions, that allows obtaining a children's satisfaction profile towards multiple factors (family relationship, social, activities, health, body functions and separation). This tool is based on self-evaluation with the support of 4 (four) images, associated to the referred factors and that the child can answer herself, with every question receiving a punctuation that ranges from 0 to 3 and being able to obtain a total score from 0 to 78 points (Assumpção et al., 2000). Longo et al. (2009) when assessing QOL's perception and the desires expressed by children between 8 and 11 years old, and their parents, observed a similarity in the answers with the predominance in the material goods and activities categories. Being an instrument capable of approaching family relationships, social, activities, health, body functions and separation.

SF-36 questionnaire is already structured with 36 questions arranged in 8 domains with the objective to assess: physical condition, physical pain, general health status, vitality, social relationships, emotional condition, and mental health (Lins & Carvalho, 2016). The domains are arranged in two summary scales that describe the physical and mental components. In Lue et al. (2016)'s study, the objectives were to use SF-36 and WHOQOLBREF tools to evaluate the health-related quality of life (HRQOL) and global QOL from teenagers and young men with DMD and to explore the associations between QOL and functional state in patients with DMD. The results presented here show that patients with

DMD have low QOL, this fact is related to physical condition and social activities generally affected by some problems related to pain.

WHOQOL-BREF, also applied in the (Lue et al., 2016)'s study, contains subjectivity aspects, multidimensionality, and individual characteristics, either positives or negatives. It is composed by 26 questions, characterized by great psychometric properties; it assesses four QOL domains: physical health, psychological health, social relationships, and environment. However, there is no cutoff point to demonstrate either better or worse QOL, and that makes the satisfaction analysis with the individuals' health even more difficult to assess (Silva et al., 2014).

KIDSCREEN questionnaire has the objective to analyze the QOL's perception and might present a longer version, KIDSCREEN-52, which contains 52 items, distributed in 10 domains that indicate good reliability and validity (Guedes & Guedes, 2011); the short version, KIDSCREEN-27, contains 27 items distributed in five domains: (1) health and physical well-being; (2) psychological well-being; (3) autonomy and the relationship with the parents; (4) social support and peer group; (5) school environment (Farias Júnior et al., 2017). The KIDSCREEN-27 considers in the evaluation the QOL related to health, beyond the indicators of lifestyle, which are influenced by demographic, social, cultural, environmental, and economic factors (Ravens-Sieberer et al., 2007). Zamani et al. (2016) applied the KIDSCREEN-27 composed by 27 questions to 85 young people, with ages between 8-18, the results indicated a satisfactory QOL, and that the young people with DMD have positive attitudes related to life, despite the disease's progressive nature and the limitations in physical activities and contact with friends.

The most used questionnaire in this study was the Pediatric Quality of Life Inventory (PedsQL ™), a promising tool for QOL evaluation and available with appropriate versions for kids age and parallel forms for children and parents (Reinfjell et al., 2006). In Landfeldt et al. (2016)'s study the PedsQL covered three domains: (1) about the neuromuscular disease in the youth (the disease process and associate symptomatology); (2) communication (patient's ability to communicate with health-care professionals and other people about their condition); and (3) family's resources (financial and social support from the family). The questions in PedsQL are transformed into a scale that varies from 0 to 100, which the higher punctuation indicates high HRQOL. It was observed that children and young people with DMD are classified as happy and with great health by their caregivers, regardless of the current

ambulatory category, indicating that some QOL domains remain unharmed with the DMD progression.

Uzark et al. (2012) while assessing 200 parents and 117 boys with DMD, observed that young people with DMD are more likely to have a significantly compromised psychosocial QOL. A better QOL overview can improve the impact of interventions, thus, facilitating the pediatric health-care professional's clinic decisions, optimizing treatment strategies for children and adolescents with DMD.

4. Conclusions

This study examined the importance of these tools regarding QOL's assessment in individuals with DMD. Such tools are essential for the evaluator's multidimensional view in terms of subjective questions that cannot be observed during clinical analysis. Furthermore, it is necessary to notice the sample's homogeneity so that the result will not be influenced. The evaluation must be done by a single evaluator, in order to reduce the risks of results with different interpretations over the questions and answers. As for the interviewed, it is necessary an evaluation concerning the socioeconomic profiles, the age group and the DMD stage so that the given answers do not suffer any influence regarding their own heath perception, QOL, and familiar relationship, reinforcing the need for more scientific studies that lead to the validation of QOL instruments specifically aimed at Muscular Dystrophy.

References

Assumpção, F. B. et al. (2000). Escala de Avaliação de Qualidade de Vida. *Arquivos de neuro-psiquiatria*, 58 (1), 119–11927.

Burckhardt, C. S. & Anderson, K. L. (2003). The Quality of Life Scale (QOLS): Reliability, Validity, and Utilization. *Health and Quality of Life Outcomes,* 1 (60), 1–7.

Booth, M. L.; Okely, A. D.; Chey, T. N. & Bauman, A. (2002). The reliability and validity of the Adolescent Physical Activity Recall Questionnaire. *Medicine & Science in Sports & Exercise,* 34 (12), 1986-1995.

Carr, A. J. & Higginson, I. J. (2002) Are quality of life measures patient centred? *BMJ (Clinical Research),* 322 (7298), 1357–1360.

Farias Júnior, J. C.; Loch, M. R.; Lima Neto, A. J. & Ferreira, F. E. L. L. (2017). Reprodutibilidade, consistência interna e validade de construto do KIDSCREEN-27 em adolescentes brasileiros. *Cadernos de Saúde Pública*, 33 (9), 1–14.

Fujino, H.; Iwata, Y.; Saito, T.; Matsumura, T.; Fujimura, H. & Imura, O. (2016). The experiences of patients with Duchenne muscular dystrophy in facing and learning about their clinical conditions. *International Journal of Qualitative Studies on Health and Well-being*, 11 (32045), 1-9.

Gierlaszynska, K.; Pudlo, R.; Jaworska, I. & Byrczek-Godula, K. (2016). Tools for assessing quality of life in cardiology and cardiac surgery. *Polish Journal of Cardio-thoracic surgery*, 13 (1), 78–82.

Guedes, D. P. & Guedes, J. E. R. P. (2011). Tradução, adaptação transcultural e propriedades psicométricas do KIDSCREEN-52 para a população brasileira. *Revista Paulista de Pediatria*, 29(3), 364-71.

Jacques et al. (2019). Quality of life in adults with muscular dystrophy. *Health and Quality of Life Outcomes,* 17 (121), 1-10.

Landfeldt, E. et al. (2016). Health-related quality of life in patients with Duchenne muscular dystrophy: A multinational, cross-sectional study. *Developmental Medicine and Child Neurology*, 58 (5), 508–515.

Lins, L. & Carvalho, F. M. (2016). SF-36 total score as a single measure of health-related quality of life: Scoping review. *SAGE Open Medicine*, 4 (1), 1-12.

Longo, E.; Moreno-Valdés, M. T.; Silva, C. E. P. & Badia, M. (2009). Quality of life and wishes in Duchenne Muscular Dystrophy: the perception of children and their parents. *Revista Brasileira em Promoção da saúde,* 22 (4), 233-239.

Lue, Y. J.; Chen, S. S. & Lu, Y. M. (2017). Quality of life of patients with Duchenne muscular dystrophy: from adolescence to young men. *Disability and Rehabilitation*, 39, (14), 1408–1413.

MAH, J. K. (2016). Current and emerging treatment strategies for Duchenne muscular dystrophy. *Neuropsychiatric Disease and treatment*, 12 (3), 1795–1807.

Moraes, F. M.; Fernandes, R. C. S. & Medina-Acosta, E. (2011). Distrofia Muscular de Duchenne: relato de caso. *Revista Cientifica da FMC*, 6 (2), 11–15.

Organization, T. W. H. (1995). The World Health Organization quality of life assessment (WHOQOL): position paper from the World Health Organization. *Social Science and Medicine,* 41 (10), 1403-1409.

Pereira, A. S., Shitsuka, D. M., Parreira, F. J., Shitsuka, R. *Methodology of cientific research.* [*e-Book*]. Santa Maria City. UAB / NTE / UFSM Editors. Available at: https://repositorio.ufsm.br/bitstream/handle/1/15824/Lic_Computacao_Metodologia-Pesquisa-Cientifica.pdf?sequence=1.

Ravens-Sieberer, U. et al. (2007). The KIDSCREEN-27 quality of life measure for children and adolescents: psychometric results from a cross-cultural survey in 13 European countries. *Quality of Life Research*; 16(8), 1347-56.

Reid, D. T. & Renwick, R. M. (1994). Preliminary validation of a new instrument to measure life satisfacion in adolecents with neuromuscular disorders. *International Journal of Rehabilitation Research*, 17 (1), 184–188.

Reinfjell, T.; Diseth, T. H.; Veenstra, M. & Vikan, A. (2006). Measuring health-related quality of life in young adolescents: Reliability and validity in the Norwegian version of the Pediatric Quality of Life Inventory^{TM} 4.0 (PedsQL) generic core scales. *Health and Quality of Life Outcomes*, 4 (1), 1–9.

Romitti et al. (2015). Prevalence of Duchenne and Becker Muscular Dystrophies in the United States. *HHS Public Access,* 135 (3), 513-521.

Ryder, S. et al. (2017). The burden, epidemiology, costs and treatment for Duchenne muscular dystrophy: an evidente review. *Orphanet Journal of Rare Diseases*, 26 (1), 1-21.

Silva, P. A. B.; Soares, S. M.; Santos, J. F. G. & Silva, L. B. (2014). Cut-off point for WHOQOL-bref as a measure of quality of life of older adults. *Revista de Saúde Publica*, 48 (3), 390–397.

Simon, V. A.; Resende, M. A. D.; Simon, M. A. V. P.; Zanoteli, E. & Reed, U. C. (2011). Duchenne muscular dystrophy: quality of life among 95 patients evaluated using the Life Satisfaction Index for Adolescents. *Arquivos de Neuro-Psiquiatria*, 69 (1), 19–22.

Souza, J. G. S.; Pamponet, M. A.; Souza, T. C.; Pereira, A. R.; Souza, A. G. S. & Martins, A. M. E. B. (2014). Instrumentos utilizados na avaliação da qualidade de vida de crianças Brasileiras. *Revista Paulista de Pediatria*, 32 (2), 272–278.

Uzark, K.; King, E.; Cripe, L. & Spicer, R. (2012). Health-Related Quality of Life in Children and Adolescents With Duchenne Muscular Dystrophy. *Pediatrics*, 130 (6), 1559–1566.

Zamani, G. et al. (2016). The quality of life in boys with Duchenne muscular dystrophy. *Neuromuscular Disorders,* 26 (7), 423–427.

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Lorenna Emília Sena Lopes – 8,5%

Stefane dos Santos – 8,5%

Luiz Eduardo Oliveira de Almeida – 8,5%

Carlos Roberto Xavier Santos Filho – 8,5%

Drielly Catarinny dos Santos Meneses – 8,5%

Sheilla da Silva Barroso – 8,5%

Ana Maria Gomes – 8,5%

Reinaldo Viana Belo Neto – 8,5%