

# Brazilian consensus on Duchenne muscular dystrophy. Part 2: rehabilitation and systemic care

## Consenso brasileiro para distrofia muscular de Duchenne. Parte 2: reabilitação e cuidados sistêmicos

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### ABSTRACT

Significant advances in the understanding and management of Duchenne muscular dystrophy (DMD) have occurred since the publication of international guidelines for DMD care in 2010. Our objective was to provide an evidence-based national consensus statement for multidisciplinary care of DMD in Brazil. A combination of the Delphi technique with a systematic review of studies from 2010 to 2016 was employed to classify evidence levels and grade of recommendations for the guideline. Our recommendations were divided in two parts. Guideline methodology and overall disease concept descriptions are found in Part 1. Here we present Part 2, where we provide the results and recommendations on rehabilitation and systemic care for DMD.

**Keywords:** muscular dystrophy, Duchenne; consensus; physical therapy modalities; exercise; rehabilitation; noninvasive ventilation; cognition; quality of life; orthotic devices; muscle stretching exercises; tracheostomy; vital capacity; respiratory function tests; cardiomyopathies; heart failure; nutrition disorders; nutritional support; intellectual disability.

### RESUMO

Avanços significativos na compreensão e no manejo da Distrofia Muscular de Duchenne (DMD) ocorreram desde a publicação de diretrizes internacionais para o cuidado destes pacientes em 2010. Nosso objetivo foi elaborar um consenso nacional baseado em evidências para o cuidado multidisciplinar dos pacientes com DMD no Brasil. Utilizamos uma combinação da técnica de Delphi com uma revisão sistemática da literatura de 2010 a 2016 para classificarmos os níveis de evidência e graus de recomendação para o consenso. Nossas recomendações foram divididas em duas partes. A metodologia utilizada na elaboração do consenso e conceitos gerais da doença encontram-se na parte 1. Neste artigo fornecemos os resultados e recomendações sobre reabilitação e cuidados sistêmicos para DMD.

**Palavras-chave:** distrofia muscular de Duchenne; consenso; modalidades de fisioterapia; exercício; reabilitação; ventilação não invasiva; cognição; qualidade de vida; aparelhos ortopédicos; exercícios de alongamento muscular; traqueostomia; capacidade vital; testes de função respiratória; cardiomiopatias; insuficiência cardíaca; transtornos nutricionais; apoio nutricional; deficiência intelectual.

Duchenne muscular dystrophy (DMD), the most common childhood muscular dystrophy, leads to severe disability and early death in the late teenage years if untreated. An X-linked degenerative disease, DMD affects approximately 1

in 3,500 to 5,000 live male births<sup>1</sup>. The condition is characterized by progressive loss of muscle strength, with some boys presenting with delayed motor milestones with or without intellectual disability. Diagnosis is generally suspected by the

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age of five years when physical ability divergent from their peers becomes evident. Females are usually asymptomatic, but some female carriers present with milder forms of the disease, generally associated with chromosomal rearrangements<sup>2</sup>. Duchenne muscular dystrophy occurs as a result of mutations in *DMD* (locus Xp21.2), which codes for the protein dystrophin<sup>3</sup>. Mutations that lead to dystrophin absence result in irreversible degeneration of the muscle tissue, accounting for the DMD phenotype<sup>1,3</sup>. Other mutations that lead to partial dystrophin expression are less severe, resulting in milder dystrophinopathy phenotypes, known as Becker muscular dystrophy<sup>4</sup>.

International guidelines for DMD care were published in 2010, with recommendations for DMD management, assessment and intervention<sup>4,5</sup>. These guidelines were generated by an international group of experts, mainly from Europe and the United States of America, based on literature reviews and expert opinion. They divided their work into the following topics: diagnosis, rehabilitation, orthopedic, psychosocial, cardiac, pulmonary, gastrointestinal/nutritional and steroid management<sup>4,5</sup>. However, because of significant advances in the understanding and management of DMD since then, an update review of previous guidelines has become of paramount importance. Improvements in general care, steroid treatment, noninvasive ventilatory support, cardiomyopathy and scoliosis management may significantly change the course of DMD. Therefore, a review of previous guidelines is necessary, as well as highlighting some new specific guidelines that are underway or have recently been published<sup>6,7</sup>.

Evidence-based practice has been heralded as the most appropriate way of ensuring that patients receive the most effective care possible.

Evidence-based practice involves much more than locating, analyzing, and appraising the best evidence available on the effectiveness of an intervention. Levels of evidence are based on study design and the methodological quality of individual studies. It is also important to make a judgment about the relevance and applicability of the evidence to the targeted patient group for the guideline, the consistency of the evidence, and the likelihood of clinical impact with the intervention. Finally, a link has to be made between the strength of the available evidence and the grade of the recommendation<sup>8</sup>.

The need to review guidelines published in 2010<sup>4,5</sup> in the light of the more recent publications, with a methodology that minimizes expert opinion, and with a focus on regional feasibility, was the motivation for the present work. Our objective was to produce an evidence-based consensus statement on the main management issues in DMD to be used as a guide for health practitioners following these patients.

Part 2 deals with rehabilitation and systemic care. For each DMD stage, priorities are presented. Measures for prevention of complications setting in early are considered priorities. Should a patient begin being followed at a later

point in the disease, with some of the interventions not yet having been started, these might be included in a later stage.

## METHODS

For detailed methodology, see Part 1 in the August 2017 issue of *Arquivos de Neuro-Psiquiatria*<sup>9</sup>.

The working groups began with a literature review from 2010 to 2016. A combination of evidence based level and recommendation (Table 1) and the Delphi technique<sup>10</sup> were used to produce this consensus.

## RESULTS

In Part 2, we focus on rehabilitation and systemic care.

As a result of the open question to the members of the working groups (based on the literature review and experience following DMD patients, and assessments and interventions) a list of items was generated (Table 2).

Each of those items was then subjected to careful evaluation according to the published material evidence and its feasibility in Brazil. Further discussion, in a joint meeting of all members of divergent opinions, resulted in the following consensus, organized here according to the chronological DMD stages.

### Stage 1: Presymptomatic (from birth up to three years of age)

The main goal at this stage is to help maintain as normal growth and development as possible.

#### Rehabilitation

Motor assessment and intervention starts with the suspicion of the diagnosis and should occur regularly. Original new articles on the topic of motor intervention have been few since the publication by Bushby et al.<sup>5</sup>.

#### Assessment

The assessment tools and recommendation levels are found in Table 3. Some of those are for routine use and others are for use in research. The aim of testing is to monitor disease

**Table 1.** Level of evidence and corresponding recommendation grade.

Study type	Level of evidence	Recommendation
Randomized clinical trials/ systematic review	1	A
Cohort studies	2	B
Case control studies	3	B
Case series	4	C
Expert opinion	5	D

progression, predict functional losses or identify the need for further interventions. Literature is scarce in indicating the frequency required for these assessments; nevertheless, it is reasonable to do so once or twice a year in routine follow-up.

A great number of different motor functional scales have been published, but many are applied in research rather than in regular clinical practice, either because they are time consuming or require specific equipment.

Monitor neurodevelopmental skills according to age.

### Interventions

Disease information for family members is crucial from the moment of the diagnosis, and emotional support should be considered.

The prevention of some respiratory infections with active immunization (Table 4) should not be forgotten in routine care of DMD patients (Level of evidence: 5D, Class of recommendation: D)<sup>30,31</sup>.

### Systemic care

Monitor height and weight.

Monitor vitamin D levels: prophylaxis is recommended from diagnosis if insufficient, supplement vitamin D as

needed (Table 4). Follow national recommendation for ferrous sulfate prophylaxis. (Level of evidence: 5D, Class of recommendation: D)<sup>32,33,34</sup>.

### Stage 2: Early phase of disease symptoms (from two to seven years of age)

From here on, measures to prevent rapid progression of motor function loss are prioritized.

### Rehabilitation

#### Assessment

From this stage on, joint mobility, muscle strength and timed function tests should be measured at each follow-up. Additional motor function evaluation with either the Motor Function Measure or North Star Ambulatory Assessment. (Level of evidence: 2B, Class of recommendation: B)<sup>11,12,13,15,6,17</sup> (Table 3).

There is an increased risk of autism (3.1%), attention deficit hyperactivity disorder (11.7%), obsessive-compulsive disorder and intellectual disability (34.8%) in DMD boys<sup>35,36</sup>. Different scales are used to assess cognition and neurodevelopment, such as the Wechsler Intelligence Scale for Children (WISC)-IV, Stanford-Binet, Raven's Matrices,

**Table 2.** List of items received from each working group after the first round of the Delphi Technique.

Topic	List
	Motor
	Assessment: joint mobility, muscle strength, functional ability, timed tests, independence, scales;
	Intervention: reduce physical inactivity, preserve flexibility, maintain functional abilities, active and assistive resisted training with light weights.
	Respiratory
	Assessment: vital forced capacity, pulse oximetry, cough peak flow, nocturnal oximetry, capnography, inspiratory and expiratory maximal pressure;
	Intervention: air stacking, manual or mechanical assisted cough, nocturnal/continuous noninvasive/invasive ventilation, anti-pneumococcal and anti-influenza immunization.
Rehabilitation	Orthopedic
	Assessment: clinical, radiological, Ca, P, alkaline phosphatase and 25-hydroxy vitamin D blood level, urinary Ca, Na and creatinine;
	Intervention: vitamin D, calcium, bisphosphonate, serial casting, surgical procedures.
	Psychosocial
	Assessment: clinical, neurodevelopmental scales, cognitive scales, neuropsychological tests, language and social skills, and autism-oriented evaluation;
	Intervention: information about the disease for family members and school, training social, cognitive and language skills, adaptations for accessibility, independence, sport and leisure, school intervention, in-house care and palliative care, psychotherapy, psychopharmacology, speech therapy, occupational therapy.
	Cardiac
	Assessment: clinical, electrocardiogram, Doppler echocardiography, resonance imaging, holter;
Systemic care	Intervention: prophylaxis, corticosteroids, angiotensin enzyme converting inhibitor, beta blocker, diuretics, eplerenone, angiotensin receptor inhibitor, poloxamer 188, idebenone, genetic therapy, stem cell.
	Cardiac care for carriers: assessment with electrocardiography, Doppler echocardiography, resonance imaging.
	Nutritional care: assess weight and stature, caloric intake, micronutrients and vitamins, swallowing ability.

Kaufman, Brunet-Lezine, and Vineland scales, all of which have internal validation and are currently used in Brazil, especially by neuropsychologists (Table 3). In current medical assessment, cognitive performance is based on general clinical evaluation by traditional mental function examination (questions about naming, reasoning, and attention) and the need for special education (Level of evidence: 2B, Class of recommendation: B)<sup>29</sup>.

### Intervention

Stretching should be done to maintain joint mobility. Active, active assisted, and/or passive, of the ankle/knee/hip, four to six days a week<sup>37</sup>, from this stage on (Level of evidence: 5D, Class of recommendation: D).

Short orthosis<sup>38</sup> for daytime use is recommended to prevent ankle deformity and prolong gait ability (Level of evidence: 4C, Class of recommendation: C).

**Table 3.** Assessment tools for DMD follow-up.

Feature	Tool -limitation	Reference (Recommendation level)
<b>Motor</b>		
Joint mobility	Goniometry – goniometer (range of motion)	11 (B)
Muscle strength	Manual: Medical Research Council	11 (B)
	Equipment – myometer (muscle strength)	
Functional ability	Motor Function Measure for ambulant/non-ambulant patients equipment: mat, chair and adjustable height table, stopwatch, tennis ball, CD; space to walk 6m;	12 (B)
	North Star Ambulatory Assessment for ambulant patients only equipment: mat, 15cm high box step, chair, space to walk 10 m.	13 (B)
	Performance of the upper limb scale for ambulant/non-ambulant patients equipment: chair and adjustable height table, metric weights (5g,10g,50g,100g, 200g, 500g and 1000g); 6 coins; plastic cup; Ziploc container; Osram dot-it push light button; 5 empty (light) and filled (heavy) cans; plastic tablecloth marked with circles).	14 (B)
	Time to get up from the floor	15,16,17 (B)
Timed tests	Time to walk 10 meters	
	6 Minute walking test - space requirement of 25 m	
	Time for steps up and down equipment: 4 standard stairs with handrails.	
Independence scales	Barthel index	18 (B)
	Egen Klassifikation Scale	19 (B)
Quality of life	Pediatric Quality of Life Inventory	20 (B)
	International classification of functioning disability and health: children and youth version	17 (B)
<b>Respiratory</b>		
Hypoventilation (inspiratory muscle weakness)	Sniff nasal inspiratory pressure	32 (B)
	Forced vital capacity	21 (C)
	Maximal Inspiratory/Expiratory Pressure	22 (C)
	Night oximetry	
Cough (expiratory muscle weakness)	Capnography	23 (C)
	Peak flow	
Orthopedic	Imaging – panoramic X-ray, Dual energy x-ray absorptiometry scan	24 (D)
<b>Psychosocial</b>		
Cognitive neuropsychological assessment	Wechsler tests (WISC-III/WISC-IV, WPPSI-III or WPPSI-IV; and Kaufman)	25,26,27,28 (B)
	Stanford-Binet	
	Raven's Matrices	
Medical cognitive assessment	Brunet-Lezine and Vineland-Doll battery tests	29 (B)
	Mental function examination, necessity for special education	
	Mini-Mental State Examination	

**Table 4. Medications, supplements and vaccines for DMD patients.**

Medications, supplements and vaccines	Dose
Vitamin D	Prophylactic
	< 1 year: 400 IU/day
	> 1 year: 600 IU/day
	higher maintenance (1,000 IU/day) is necessary for DMD boys <sup>62</sup>
	Therapeutic (deficiency of 25-OH-VitD: blood level < 12 ng/ml)
Calcium Carbonate (1g = 400 mg elemental calcium)	< 1 year: 2,000 IU/day (12 weeks)
	1-12 years: 3,000-6,000 IU/day (12 weeks)
Ferrous sulfate	> 12 years: 6,000 IU/day (12 weeks)
	Vitamin D deficiency
Biphosphonates	40-80 mg/kg/day elemental calcium, oral, 8/8h (4 weeks)
	Prophylactic
	Full term breastfeeding infants: 1 mg/kg/day elemental ferrous from 6 until 24 months
Pneumococcal Conjugate Vaccine (Pn10 or Pn13)	Alendronate 0.08mg/kg/day or 70 mg (oral) every 2 weeks if prophylactic; every week if treatment
	Pamidronate 6-9mg/kg/year IV 3 to 4 months
	Zoledronic acid 0.1mg/kg/year IV 3 to 4 months
Pneumococcal Polysaccharide Vaccine (Pn23)	2, 4 and 12 months
Anti-Influenza Vaccine	1 dose after the age of 2 years
Yellow Fever Vaccine	1 dose each year from 6 months of age on
Angiotensin-converting enzyme inhibitors	Contraindicated if the patient is using prednisone • 20mg/day (or equivalent)
	Captopril
	6 years to adolescence 12.5 mg/dose, 1 to 2 times a day
Beta blocker	adults 25 to 50 mg, 2 to 3 times a day
	Enalapril 0.1 mg/Kg 1 to 2 times a day (maximum of 0.5 mg/Kg/day)
	Propranolol 2 to 4 mg/Kg/day, 2 to 4 times a day
	Atenolol 1 to 2 mg/Kg/day, once a day
	Carvedilol 0.1 to 1 mg/Kg/day, 1 to 2 times a day
	Metoprolol 20 mg/day

Physical conditioning is important<sup>39</sup> (Level of evidence: 4C, Class of recommendation: C). Low intensity aerobic daily exercise<sup>40</sup> (Level of evidence: 3B, Class of recommendation: B) may prevent disuse comorbidity<sup>41</sup> (Level of evidence: 4C, Class of recommendation: C). This may be either by the use of an arm ergometer under supervision, which preserves and improves the functional level<sup>42</sup> (Level of evidence: 3B, Class of recommendation: B) or assisted bicycle training, for arms and legs, 15 minutes/day, five times a week, with the positive effect on slowing disease progression for the ambulant or wheelchair stage of DMD<sup>40</sup> (Level of evidence: 3B, Class of recommendation: B). Avoid eccentric muscle loading like jumping on a trampoline.

According to the language, intellectual and behavioral assessment, schooling options, reinforcement classes and other therapies, such as speech therapy and psychopedagogical stimulation, might be added (Level of evidence: 5D; Class of recommendation: D)<sup>29</sup>.

### Systemic care

#### Assessment

The mobility limitation, added to the use of steroids, are factors related to bone morbidity. Follow-up with blood

measures of calcium, phosphorus, magnesium, phosphatase, 25-OH vitamin D and parathyroid hormone is important<sup>24</sup> (Level of evidence: 5D; Class of recommendation: D).

Cardiological assessment should be implemented, since the involvement of the heart is usually silent or with nonspecific symptoms such as sleep disturbance, loss of appetite or nausea. Therefore, electrocardiography and echocardiography should be carried out on a yearly basis<sup>43</sup>. If available, cardiac resonance imaging can be used as well<sup>44</sup> (Level of evidence: 5D; Class of recommendation: D).

#### Intervention

Supplementation of vitamin D should be prescribed according to the blood tests. If available, a dual energy x-ray absorptiometry scan can help to monitor bone over time<sup>24</sup>.

Attention to excessive weight gain and advice on healthy nutrition, with control of caloric intake if needed, plays an important role<sup>43</sup>.

Preservation of heart function can be achieved by the use of steroids (see Part 1<sup>9</sup>). Prevention of cardiac insufficiency might include the use of beta blockers and angiotensin converting enzyme inhibitors (Table 4). As fibrosis may be seen on cardiac imaging, for any decrease in cardiac function from

baseline or for heart failure (shortening fraction < 28%, ejection fraction <55%), the use of cardioprotection should be discussed<sup>44,45</sup> (Level of evidence: 5D; Class of recommendation: D).

### Stage 3: Transitional phase

Rapid progression of loss of lower limb function. At this point, loss of function will evolve rapidly, and the age limits are very variable. Getting up from the floor, or using stairs will become troublesome, and focus must be on planning to maintain social and educational activities. Some walking aids could be considered.

### Rehabilitation

#### Assessment

The same as mentioned for Stage 2. In addition, if not done previously, respiratory assessment begins, with regular lung function tests that require specific equipment. Longitudinal studies<sup>46</sup> indicate that respiratory function can improve in the first decade, but after that a decline is expected. Sniff nasal inspiratory pressure is the first measure to show decline in young DMD patients<sup>46</sup> (Level of evidence: 2B, Class of recommendation: B).

#### Interventions

Stretching continues. Long orthosis, for joint positioning while standing allows for longer periods of stretching<sup>47</sup> (Level of evidence: 3B, Class of recommendation: B)<sup>48</sup> (Level of evidence: 4C, Class of recommendation: C). Attention to the degree of muscle weakness and fatigue.

Serial casting might be an option for a nonsurgical procedure handling ankle joint deformity<sup>49</sup>. (Level of evidence: 2B, Class of recommendation: B)<sup>50</sup> (Level of evidence: 4C, Class of recommendation: C).

The pulmonary interventions aim for the maintenance of ventilatory and cough capacities<sup>51</sup> (Level of evidence: 2B, Class of recommendation: B). Air stacking is a technique to promote pulmonary expansion, alveolar recruitment and improve cough efficacy. It can be achieved with glottic respiration, or with the help of a manual resuscitation bag, as well as with a ventilator. The aim is to increase the inspired volume. It should be initiated as a daily intervention as soon as the forced vital capacity falls below 80% of the predictive value<sup>52</sup> (Level of evidence: 4C, Class of recommendation: C).

Interventions for developmental delays include physiotherapy, speech/language, and occupational therapy, and should be targeted toward improving specific skills. Speech/language therapists are necessary to treat disorders in phonological awareness/processing (dyslexia). Occupational therapy improves fine motor abilities. Symptoms of depression and anxiety may respond to psychotherapy. Behavior modification therapy and cognitive-behavioral therapy have been shown to be effective in treating

attention deficit hyperactivity disorder and oppositional and obsessive-compulsive behaviors<sup>53</sup> (Level of evidence: 5, Class of recommendation: D).

### Systemic care

#### Assessment

In addition to regular cardiologic follow-up, a holter is sometimes ordered for the correct diagnosis of arrhythmias<sup>44</sup>.

### Stage 4: Initial stage of ambulation loss

From this point of the DMD timeline, the upper limbs deserve attention, to promote as much independence as possible, and respiratory care to limit morbidity and mortality.

### Rehabilitation

#### Assessment

Timed function tests and the North Star Ambulatory Assessment are no longer possible. The Motor Function Measure and Performance of the Upper Limb scale can be used to assess the functional ability. Joint mobility and muscle strength continue to be assessed.

Ventilatory parameters help to indicate the interventions. Low peak flow values (below 270 L/min) increase the risk of complications, secretion aspiration with consequent pneumonia and atelectasia<sup>23</sup>.

#### Intervention

Stretching should continue at this point in upper as well as lower limbs.

Hydrotherapy shows no benefit for muscle strength or reduction of body mass index but this modality can improve agility<sup>54</sup> (Level of evidence: 4C, Class of recommendation: C) and, through facilitation, can help the movement already lost outside the water in weaker muscles<sup>55</sup> (Level of evidence: 5D, Class of recommendation: D).

A manual wheel chair becomes part of daily living for DMD boys, and should be correctly adapted to slow scoliosis progression, keep a symmetrical posture when sitting on the wheelchair<sup>56</sup> (Level of evidence: 4C, Class of recommendation: C), and prevent skin ulcers<sup>38</sup> (Level of evidence: 4C, Class of recommendation: C).

Ventilatory support has a precise indication. Reduction in predicted forced vital capacity (20-25% of predicted) confirms ventilatory insufficiency, and the need to increase intervention. If it is less than 40% of the predicted value, the moment to start ventilatory support is approaching<sup>57</sup> (Level of evidence: 1A, Class of recommendation: A).

Assisted cough, either with a manual or mechanical maneuver, is indicated during respiratory infections, for those with a peak flow below 270 L/min, and to clear airways on a regular basis<sup>51,58</sup> (Level of evidence: 3C, Class of recommendation C).

Psychosocial intervention continues as in Stage 3.

## Systemic care

### Assessment

Once wheelchair bound, attention to spine deformity should be included. Monitor scoliosis on regular imaging and provide adaptation of the wheelchair. Eventually, in some, surgical spinal fusion may be needed<sup>59</sup> (Level of evidence: 4C, Class of recommendation: C). Bone fractures occur due to osteopenia and osteoporosis. Dual energy x-ray absorptiometry can be used to monitor the bones<sup>24,43</sup> (Level of evidence: 5D, Class of recommendation: D).

### Intervention

For the decision on surgical intervention, both the spine deformity itself (Cobb angle on imaging) and the respiratory status contribute. There are no randomized controlled trials of surgery for scoliosis in patients with DMD. The procedure results in a better sitting position and overall quality of life<sup>59</sup> (Level of evidence: 4C, Class of recommendation: C). Biphosphonates may be of help for bone health and are usually used when fractures occur<sup>24,43</sup> (Level of evidence: 5D, Class of recommendation: D).

Constipation is related to the permanent sitting position as well as being associated with hypohydration.

Aldosterone inhibitors can be prescribed for cardiac insufficiency<sup>44</sup> (Level of evidence: 5D, Class of recommendation: D).

## Stage 5: Later stage of disease (late adolescence to adult)

Attention to maintenance of comfort and independence.

### Rehabilitation

At this stage, a motorized wheelchair is needed, and arm support should also be provided<sup>37</sup> (Level of evidence: 5D, Class of recommendation: D).

Noninvasive ventilatory support has increased DMD survival in the last decades. Noninvasive ventilation is preferred to tracheostomy because it is less invasive, more comfortable and maintains speech capacity. Forced vital capacity below 40% (or 50% for some authors), or the presence of nocturnal hypoxemia or hypercapnia, with the symptoms of fatigue, early morning headache, somnolence, concentration difficulties, loss of appetite, and depression, are indications for the use of bilevel noninvasive ventilation. It should be started with low expiratory positive airway

pressure (0–4 cmH<sub>2</sub>O) and an inspiratory positive airway pressure 10 cmH<sub>2</sub>O higher<sup>60</sup> (Level of evidence: 2C, Class of recommendation: C). Oxygen supplementation must be used cautiously because of the high risk of hypercapnia narcosis and apnea.

Access to computer technology, and finding and/or keeping an activity day by day, helps the patient lead a meaningful life.

## Systemic care

In this stage of the disease, undernutrition may occur and dysphagia may play a role<sup>43</sup>; therefore attention should be directed toward these complications. According to the assessments, diet modifications might be applied.

Emotional support, and identifying and managing depression are also of great importance at this stage.

## CONCLUSIONS

There are priority recommendations regarding the follow-up and care of DMD patients at each stage of the disease, that have been described. Anticipating the known complications, and focusing on factors that lead to them, more rapidly help to modify the natural history of the disease.

Care standards recommend preventive measures to minimize contractures, starting at the presymptomatic stage. This has been achieved in some countries<sup>61</sup> and could be spread globally. Of course, early diagnosis is the main starting point for this achievement, but also the knowledge that non-drug therapies may have a great impact on the speed of the development of functional impairments.

Some protective measures are, unfortunately, not yet current practice: cardiac and respiratory care are examples of areas where a more strict adherence to recommendations should be taken, particularly from Stage 3 onwards. Mortality in DMD is related to cardiac or respiratory complications, and good management in this regard has modified the life expectancy of these patients<sup>62</sup>.

The literature review for the present consensus comprised the period from 2010 to 2016. The DMD Care Considerations Working Group recently published in 3 parts an update<sup>63,64,65</sup> for their previous two-part publication on DMD diagnosis and care<sup>45</sup>. With a different methodology of that we used, a very similar final approach is obtained, the priority for the preventable care of DMD and organized by disease stage.

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# Erratum

Araújo APQC, Nardes F, Fortes CPDD, Pereira JA, Rebel MF, Dias CM, et al. Brazilian consensus on Duchenne muscular dystrophy. Part 2: rehabilitation and systemic care. *Arq Neuropsiquiatr*. 2018 Jul;76(7):481-489.

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**In *Rehabilitation*, where it is written:**

Noninvasive respiratory support has increased DMD survival in the last two decades. A tracheostomy is preferred because it is less invasive, more comfortable and maintains speech capacity.

**Should be:**

Noninvasive ventilatory support has increased DMD survival in the last decades. Noninvasive ventilation is preferred to tracheostomy because it is less invasive, more comfortable and maintains speech capacity.