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Research Article

PHYSICAL THERAPY IN PATIENTS WITH DUCHENNE MUSCULAR DYSTROPHY: DYNAMICS OF THE COURSE OF THE DISEASE

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BACKGROUND: Duchenne muscular dystrophy is the most severe and common form among childhood muscular dystrophies, characterized by a progressive course. One of the main measures to slow down the rate of progression of the disease is physiotherapy, but now there are no recommendations to identify the optimal motor regimen, which is effective and safe for patients with Duchenne muscular dystrophy.

AIM: The aim of the study was to evaluate the effectiveness of regular dynamic aerobic exercise in patients with Duchenne muscular dystrophy who are capable of independent movement.

MATERIALS AND METHODS: We examined 15 patients with genetically confirmed Duchenne muscular dystrophy aged 4,9 to 9,0 years (mean age 6,9 years) who did not participate in rehabilitation programs with exercise therapy for >6 months prior to inclusion in the study. All patients underwent a course of physical therapy, the duration of the course was 4 months, the course was divided into 2 stages: preparatory stage (individual cardiac functional reserve 51–60% with the number of repetitions of each exercise 6–8 times) and training stage (individual cardiac functional reserve 61–70% with the number of repetitions each exercise 10–12 times) The duration of the training was 60 minutes. At the baseline and during dynamic observation after 2 and 4 months, the following were evaluated: 6-minute walk test, timed function test (time to stand from supine, running 10 m, time to climb and descent 4 stairs).

RESULTS: Statistically significant positive dynamics was revealed: the average values of the distance of the 6-minute walk at the baseline were 478,2 ± 10,1 m, then 489,5 ± 11,4 m ($p < 0.05$) after 2 months and 502,6 ± 10,7 m ($p < 0.005$) after 4 months. The average values of the time to stand from supine at the baseline was 3,7 ± 0,2 sec., after 2 months — 3,5 ± 0,2 sec. ($p < 0.08$), after 4 months — 3,5 ± 0,2 sec. ($p < 0.05$). Mean values of time to run 10 m at baseline 4,3 ± 0,1 sec., after 2 months — 4,1 ± 0,2 sec. ($p < 0,05$), and after 4 months — 4,1 ± 0,1 sec. ($p < 0.005$).

CONCLUSIONS: Thus, the regular performance of aerobic physical therapy exercises without weights in combination with training on an exercise bike can increase endurance and speed in ambulant patients with Duchenne muscular dystrophy.

Keywords: Duchenne muscular dystrophy; rehabilitation; physical therapy.

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Научная статья

ЛЕЧЕБНАЯ ФИЗКУЛЬТУРА У ПАЦИЕНТОВ С МЫШЕЧНОЙ ДИСТРОФИЕЙ ДЮШЕННА: ДИНАМИКА ТЕЧЕНИЯ ЗАБОЛЕВАНИЯ

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Актуальность. Мышечная дистрофия Дюшенна — наиболее тяжелая и распространенная форма среди мышечных дистрофий детского возраста, характеризующаяся прогрессирующим течением. Одной из основных мер замедления скорости прогрессирования заболевания считается лечебная физкультура, однако в настоящее время отсутствуют рекомендации, позволяющие выделить оптимальный двигательный режим, являющийся эффективным и безопасным для пациентов с мышечной дистрофией Дюшенна.

Цель — оценка эффективности регулярных динамических аэробных упражнений у пациентов с мышечной дистрофией Дюшенна, способных к самостоятельному передвижению.

Материалы и методы. Обследовано 15 пациентов с генетически подтвержденной мышечной дистрофией Дюшенна в возрасте от 4,9 до 9,0 года (средний возраст 6,9 года), не принимавших участие в реабилитационных программах с лечебной физкультурой более 6 мес. до включения в исследование. Все пациенты проходили курс лечебной физкультуры длительностью 4 мес. Курс был разделен на два этапа: подготовительный (51–60 % индивидуального функционального резерва сердца с повторением каждого упражнения 6–8 раз) и тренирующий (61–70 % индивидуального функционального резерва сердца с повторением каждого упражнения 10–12 раз). Длительность тренировки составляла 60 мин. На исходном уровне и при динамическом наблюдении через 2 и 4 мес. оценивали: дистанцию 6-минутного теста ходьбы, тесты на время (подъем с пола, бег на дистанцию 10 м, подъем и спуск по лестнице).

Результаты. Была выявлена статистически достоверная положительная динамика: средние значения дистанции 6-минутной ходьбы на исходном уровне 478,2 ± 10,1, 489,5 ± 11,4 м ($p < 0,05$) через 2 мес. и 502,6 ± 10,7 м ($p < 0,005$) через 4 мес. Средние значения скорости подъема с пола на исходном уровне 3,7 ± 0,2 с, через 2 мес. — 3,5 ± 0,2 с ($p < 0,08$), через 4 мес. — 3,5 ± 0,2 с ($p < 0,05$). Средние значения скорости бега на дистанцию 10 м на исходном уровне 4,3 ± 0,1 с, через 2 мес. — 4,1 ± 0,2 с ($p < 0,05$), через 4 мес. — 4,1 ± 0,1 с ($p < 0,005$).

Выводы. Таким образом, регулярное выполнение аэробных упражнений лечебной физкультуры без отягощения в сочетании с тренировками на велотренажере позволяет повысить показатели выносливости и скорости у пациентов с мышечной дистрофией Дюшенна на амбулаторных стадиях заболевания.

Ключевые слова: мышечная дистрофия Дюшенна; лечебная физкультура; реабилитация.

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BACKGROUND

Duchenne muscular dystrophy (DMD) is the most severe and common form of childhood muscular dystrophy. The prevalence is 3.3 per 10 thousand newborn boys. DMD is characterized by a steady progressive course, weakness, and atrophy with a predominant lesion in the skeletal muscles of the pelvic girdle, thighs, and lower legs, which leads to early disability and loss of the ability to move independently at the age of approximately 10–12 years [1, 2, 6, 22]. As the disease progresses, it is characterized by the involvement of the axial and proximal muscles of the upper extremities, as well as the development of cardiac, respiratory, and orthopedic complications. Exercise therapy (ET) is considered one of the main measures to prevent disease progression and concomitant orthopedic disorders [12, 21]. International guidelines for the treatment of patients with neuromuscular diseases recommended a light (submaximal) level of physical activity [6]; however, current data are limited, and only single studies have examined the efficiency of various training programs, most of which were conducted with adult patients. There are no recommendations to identifying the optimal motor regimen that can be effective and safe for patients with DMD and other neuromuscular diseases [24]. In the case of an incorrectly selected mode of motor activity, disease progression can be accelerated. Patients with an insufficient level of physical activity do not achieve clinically significant improvement, whereas excessive exercise may be accompanied by adverse effects, such as increased pain in the muscles, myoglobinuria, and ultimately deterioration in motor capabilities due to excessive damage to muscle fibers and increased nonspecific inflammatory activities [8, 9]. According to the World Health Organization recommendations, children should maintain moderate or vigorous physical activities for at least 60 min daily. Studies have shown that boys with DMD do not meet these criteria, and their activity decreases as they grow older, which exacerbates the disease course [5]. Naturally, after 12 months, patients with DMD demonstrate negative dynamics in the 6-min walk distance, with the data varying from -10.9 ± 69.2 to -25.8 ± 74.3 m [7, 11, 19]. At the age of 8.8 ± 2.0 years, patients with DMD have deteriorated results in the tests for rising from the floor, running for a distance of 10 m, and climbing stairs from –0.07 to –0.33 s during the follow-up period of 12 months or remained stable at the age of 5–6.9 years when in the "plateau" phase [4, 6, 17]. The control group performs a 10-m run test in an average of 3.0 s and rising from the floor test and climbing on four steps in 1–2 s.

Moreover, dynamic gymnastics (isotonic) involves movements accompanied by alternating muscle tension and relaxation, which provides for an aerobic way of energy supply. This occurs because of an increase in the activity of the sympathetic nervous system and a pronounced homeostatic effect, which is based on the improvement of hemocirculation and metabolic processes and contributes to an increase in the functional state of the body [14, 25]. The basis of the physiological reactions in dynamic gymnastics is a combination of the rate of motor reactions and the heart rate [14]. An adequate response to physical activity is accompanied by recovery and improvement of hemodynamics and, accordingly, optimization of the cardiovascular system. Such training significantly increases the functionality of the circulatory system [18]. Moreover, the disrupted balance between excitation and inhibition in the nervous system, and tension and relaxation of muscles is restored. Consequently, the functions of all organs and systems are optimized [15, 23].

Aspects of dynamic gymnastics in the norm are as follows: (1) Training of speed endurance of the muscle, first of all, depends on the speed of movements and not on the load duration. Speed training at an early age (up to 7 years) helps increase the speed of innate motor reactions. (2) The muscle mass increases due to an increase in the number of myofibrils. Muscle fibers are more often located parallel to the axis. There is an elongation of the muscle part and shortening of the tendon part of the muscle fiber. The muscle volume increases more than with isometric gymnastics. (3) An increase in dynamic load promotes the stimulation and lengthening of tubular bones [10, 18, 25]. Based on these aspects, dynamic exercises were developed to maximize the preservation of the affected muscles and improve hemodynamics and neuroregulation of both the muscles and the child's body.

The study aimed to evaluate the efficiency of regular dynamic aerobic exercise in patients with DMD who can move independently.

MATERIALS AND METHODS

The study was conducted at the St. Petersburg State Pediatric Medical University. Fifteen patients with genetically confirmed DMD at the age of 4.9–9.0 (mean, age 6.9) years were examined. Of these patients, 13 took glucocorticosteroid therapy daily at standard recommended dosages for >6 months.

Inclusion criteria for inclusion were as follows: (1) male sex, (2) genetically confirmed DMD, (3) ability to move independently (early and late outpatient stage), (4) recent participation in rehabilitation ET programs more than 6 months before the inclusion in the

study, and (5) opportunity for the parents/guardians and patients to visit the clinic regularly for rehabilitation courses throughout the study.

At the initial level and during case follow-up after 2 and 4 months, the 6-min walk test and time tests (rising from floor, 10-m run test, and climbing and descending the stairs) were performed. The functional class of the performance tests for time was assessed on a six-point scale (Tables 1–3).

A neurologist who had specialized training in these tests performed the assessments. For all patients, the tests were performed under equal comfortable conditions, taking into account the time of day, with the patient in comfortable light clothing and shoes. If the patient showed signs of physical or emotional exhaustion, the test was postponed to another day.

All patients performed a set of exercises, including the following:

(1) Dynamic aerobic exercises with an emphasis on the axial muscles, pelvic girdle, thighs, and lower legs

2) Exercises on an exercise bicycle on a model with a wide seat, a stable back, and adjustment for patients of different ages

3) Sitting and standing balance training

4) Exercises for shock absorption of the feet

5) Strengthening the stabilizers of the trunk (musculus transversus abdominis, abdominal muscles, and deep rotators of the spinal column) and lower extremities

6) Correction of the static stereotype (development of the skill of correct posture while sitting and standing)

7) Breathing exercises to increase the range of motion in the intercostal spaces and improvement of mobility in the costal–vertebral and costal–sternal joints.

The duration of the rehabilitation course was 4 months, which was divided into two preparatory and training. The trainings were performed under the supervision of an ET specialist. The intensity was selected for each patient individually, depending on age, and heart rate according to the equation of the individual functional reserve of the heart (IFRH) = 190–age (years). The intensity of the preparatory stage was 51%–60% IFRH with 6–8 repetitions of each exercise. The intensity for the training stage was 61%–70% IFRH with 10–12 repetitions of each exercise. The training session lasted for 60 min with a frequency of three times a week. Patent application was filed*.

Statistical analysis was performed using IBM SPSS Statistics version 23.0 (IBM Corp., Armonk, NY, USA). Mean values and confidence intervals were calculated at $p = 0.05$; paired Student's t-test was performed for dependent aggregates.

RESULTS

The average distance in the 6-min walk test was 478.2 ± 10.1 m. During repeated examinations after an ET course, a statistically significant improvement was noted, with the average distance of 489.5 ± 11.4 m ($p < 0.05$) after 2 months and 502.6 ± 10.7 m ($p < 0.005$) after 4 months (Fig. 1).

The average values of the speed of rising from the floor at the initial level were 3.7 ± 0.2 s. During case follow-up, improvement was noted, with the average time of 3.5 ± 0.2 s ($p < 0.08$) after 2 months and 3.5 ± 0.2 s ($p < 0.05$) after 4 months (Fig. 2).

* Priority reference No. 2021135761 dated 03.12.2021. Suslov V.M., Lieberman L.N., Suslova G.A. "Method of conducting therapeutic physical culture for patients with neuromuscular diseases".

Table 1 / Таблица 1

Description of the functional class when performing a timed functional test rising from the floor
Описание функционального класса при выполнении теста на скорость при подъеме с пола

Score / Баллы	Functional class of motor abilities of the rise from floor test / Функциональный класс при выполнении теста на подъем с пола
1	Unable to stand from supine, even with use of a chair / Не может встать из положения лежа, даже с опорой на стул
2	Requires chair to stand up from supine / Может встать с опорой на стул
3	Rolls over, stands up with both hands "climbing up" the legs / Переворачивается на живот, встает с опорой на ноги обеими руками
4	Rolls over, stands up with 1 hand support on leg/ Переворачивается на живот, встает с опорой на ногу одной рукой
5	Rolls to the side and stands up with one or both hands on the floor to start to rise but does not touch legs / Переворачивается на бок и встает с опорой на пол одной/двумя руками, не прикасаясь к ноге
6	Stands up without rolling over or using hands on legs / Может встать без переворота и опоры на ноги

Table 2 / Таблица 2

Description of the functional class when performing a timed functional test run 10 meters distance
Описание функционального класса при выполнении теста на скорость при беге на дистанцию 10 м

Score / Баллы	Functional class of motor abilities of the 10 meters run test / Функциональный класс при выполнении теста на бег 10 м
1	Unable to walk / Не может ходить
2	Unable to walk independently but can walk with knee-ankle-foot orthosis or support from a person / Может ходить, но использует коленно-голеностопные ортезы или постороннюю помощь
3	Cannot increase walking speed, highly adapted, wide-based lordotic gait / Не может ускорить ходьбу, выраженная адаптационная походка с широко расставленными ногами с поясничным гиперлордозом
4	Can pick up speed but cannot run, moderately adapted gait / Может ускорить ходьбу, не может бежать, умеренная адаптационная походка
5	Nearly running, but cannot achieve both feet off the ground simultaneously (Duchenne jog)/ «Бег» без отрыва обеих стоп от пола (Duchenne jog)
6	Runs and gets both feet off the ground / Нормальный бег с отрывом обеих стоп от пола

Table 3 / Таблица 3

Description of the functional class when performing a 4-step climbs and descend
Описание функционального класса при выполнении теста на скорость при подъеме и спуске на 4 ступени

Score / Баллы	Functional class of motor abilities of climb and descend on 4 steps / Функциональный класс при выполнении теста на подъем и спуск на 4 ступени
1	Unable to climb/descend 4 stairs / Не может подняться/спуститься на 4 ступени
2	Climbs/descend 4 standard stairs “marking time” (climbs one foot at a time, with both feet on a step before moving to next step), uses both arms on one or both handrails or uses 1 handrail and the other arm pushes on the leg or body / Поднимается/спускается приставными шагами, опираясь обеими руками на один или два перила или опираясь одной рукой на перила, а второй на ногу
3	Climbs/descend 4 standard stairs “marking time”(climbs one foot at a time, with both feet on a step before moving to next step), using one arm on one handrail or one hand pushing on leg or body / Поднимается/спускается приставными шагами, опираясь одной рукой на перила или ногу
4	Climbs/descend 4 standard stairs “marking time” (climbs one foot at a time, with both feet on a step before moving to next step), not needing handrail and not using hands to push on leg / Поднимается/спускается приставными шагами без помощи перил и опоры на руки
5	Climbs/descend 4 standard stairs alternating feet, needs handrail/s for support or uses arms to push on the leg or body / Поднимается/спускается через ступеньку, с опорой руками на перила или ноги
6	Climbs/descend 4 standard stairs alternating feet, not needing handrail support or using arm to push on the leg / Поднимается/спускается через ступеньку без перил и помощи рук

The mean values of the functional test of rising from floor were 4.4 ± 0.2 points at the initial level, 4.8 ± 0.1 points after 2 months, and 4.8 ± 0.1 points after 4 months.

The average running speed for a distance of 10 m at the initial level was 4.3 ± 0.1 s. With the follow-up, the mean time was 4.1 ± 0.2 s after 2 months ($p < 0.05$) and 4.1 ± 0.1 s after 4 months ($p < 0.005$) (Fig. 3).

The mean values of the functional test of rising from floor were 4.4 ± 0.2 points at the initial level, 4.8 ± 0.1 points after 2 months, and 4.8 ± 0.1 after 4 months.

The average timing of rising on four steps at the initial level, after 2 and 4 months, were 2.9 ± 0.1 s (Fig. 4). The average values of the functional test of rising on four steps were 4.9 ± 0.2 points at the initial level, 5.2 ± 0.2 points after 2 months, and 5.2 ± 0.2 points after 4 months.

The average values of the speed of descent by four steps at the initial level were 3.4 ± 0.2 s. With the follow-up, the mean time was 2.8 ± 0.1 s after

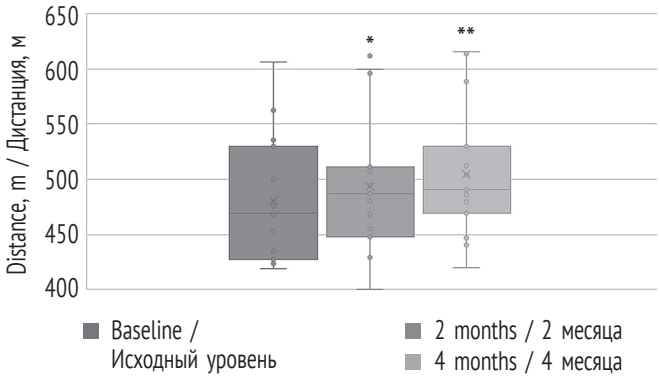

Fig. 1. Dynamic of the 6 minute walking test. * $p < 0.05$, ** $p < 0.005$

Рис. 1. Динамика дистанции 6-минутного теста ходьбы. * $p < 0,05$; ** $p < 0,005$

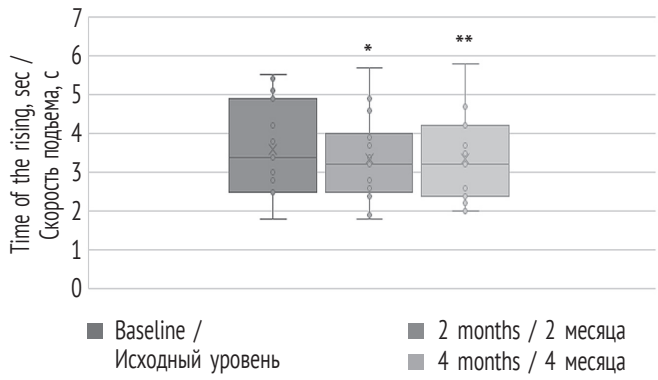

Fig. 2. The dynamics of the change in the time of the rising from the floor. * $p < 0.05$, ** $p < 0.005$

Рис. 2. Динамика изменения скорости подъема с пола. * $p < 0,05$; ** $p < 0,005$

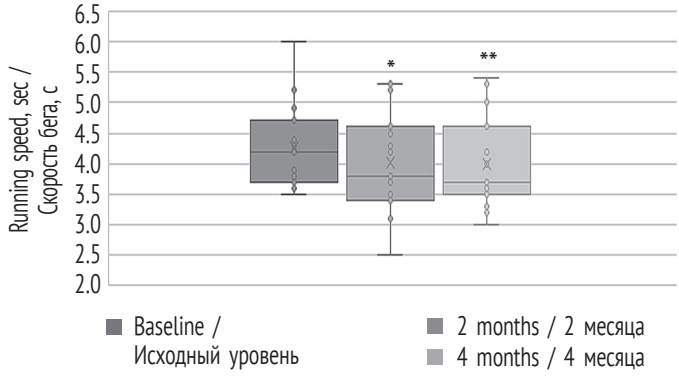

Fig. 3. Dynamics in the time of the running 10 meters distance. * $p < 0.05$, ** $p < 0.005$

Рис. 3. Динамика изменения скорости бега на дистанцию 10 м. * $p < 0,05$; ** $p < 0,005$

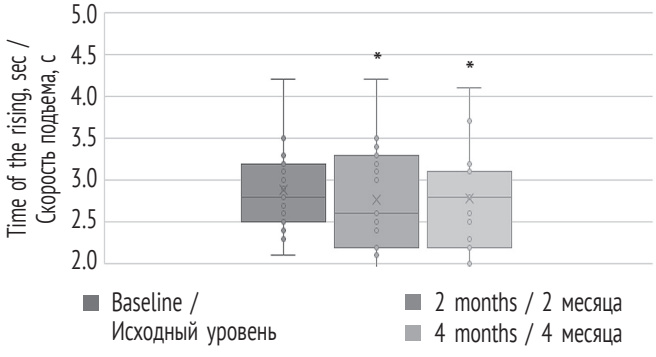

Fig. 4. Dynamics of the time of the 4 steps climbing. * $p < 0.05$

Рис. 4. Динамика изменения скорости подъема на 4 ступени. * $p < 0,05$

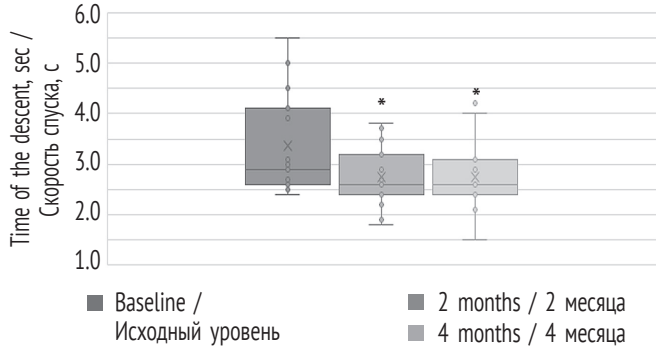

Fig. 5. Dynamics of the time of the 4 steps descend. * $p < 0.05$

Рис. 5. Динамика изменения скорости спуска на 4 ступени. * $p < 0,05$

2 months ($p < 0.005$) and 2.8 ± 0.1 s after 4 months ($p < 0.005$) (Fig. 5).

The average values of the functional test of descent by four steps were 4.1 ± 0.3 points at the initial level, 4.8 ± 0.3 after 2 months, and 4.9 ± 0.1 after 4 months.

DISCUSSION

This study assessed the motor abilities of patients who regularly performed aerobic exercises and exercised on an exercise bicycle for 4 months. Despite the average age of the patients, which is characterized by

the plateau phase or progressive symptoms, improvements in the distance of the 6-min walk test and time tests were noted in the study group. Improvement was also noted in performing aerobic exercises using a hand and foot bicycle ergometer, and an improvement was registered in the number of pedal revolutions on an exercise bicycle in 30 patients with DMD after a course of aerobic training for 24 weeks. An improvement in motor functions of the hands in patients unable to walk independently was also found [13].

During the 4-month training in patients with DMD, a statistically significant improvement in the performance of tests for time (rising from floor, 10-m running test, and descending on four steps) was registered. The time of climbing on four steps remained without statistically significant changes and amounted to 2.9 ± 0.1 s. However, in all time tests, including the four-step climb, an insignificant increase in the functional class of the task performance was found because of an improvement in the movement pattern and a decrease in the severity or absence of compensatory movements during the test. Improvement was a characteristic of the functional class at the initial level of 5 points for rising from floor and running 10 m and 4–5 points for climbing and descending the stairs, whereas an initial score of less than 3–4 points for all time tests was characterized by more persistent disorders during case follow-up, which was associated with a more severe clinical presentation and pronounced morphological changes in skeletal muscles.

Chronic DMD is characterized by steady deterioration, despite the rehabilitation measures [3, 11, 16]; however, regular exercise can slow down the progression of muscle weakness, improve cardiorespiratory function, reduce the severity of articular contractures, and improve the quality of life of the patients and their families [20].

CONCLUSION

Thus, regular performance of aerobic exercise without weights in combination with training on an exercise bicycle can improve endurance and speed in patients with DMD at the outpatient stages of the disease.

ADDITIONAL INFORMATION

Author contribution. Thereby, all authors made a substantial contribution to the conception of the work, acquisition, analysis, interpretation of data for the work, drafting and revising the work, final approval of the version to be published and agree to be accountable for all aspects of the work.

Competing interests. The authors declare that they have no competing interests.

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