Russian Journal of Biomechanics, Vol. 4, № 3: 74-79, 2000

THEORETICAL PREMISES AND PRACTICAL RESULTS OF LOWER LIMB LENGTH INCREASE IN PATIENTS WITH ACHONDROPLASIA

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Abstract: Achondroplasia is a congenital disease with infringement of osteogenesis causing an evident congenital dwarf proportions (short limbs and normal trunk). Increase of the patients’ height may be performed at the expense of lower limb elongation according to the Ilizarov technique. While determining proper duration of step period the limb length was related with the length of simple pendulum. After treatment relative step length of patients decreased, step period did not change practically and approached the calculated value, walking velocity increased by 15%, daily locomotory activity increased by 51%. Nevertheless, it was rather difficult to achieve the indices of locomotory activity, typical of normal subjects of the same age, probably due to attendant lesion of locomotor organs.

Key words: achondroplasia, Ilizarov technique, limbs, biomechanical analysis, osteogenesis

Introduction

Definitive size of limbs in patients with achondroplasia is almost half as much, than in normal subjects, because of their limb longitudinal growth lag behind [1]. In such cases increase of the patients’ height is performed at the expense of lower limb elongation according to the Ilizarov technique. Complete correction of growth lag behind is possible technically, but in case of significant decrease of muscular contractility. That is why improvement of the locomotor system function might be the main criterion of the required and sufficient amount of increment [2].

Step rate and step length in the natural locomotor act determine movement velocity and are related with each other. There is no rigorous connection between the step rate and step length only in children below 2 years of age [3]. In the process of further ontogenesis the larger step length conforms to the faster step rate, thereby ensuring the problem solution to increase the linear velocity of walking [4]. The increment of step length is considered to be related with children’s age increase, though really the effect of limb longitudinal growth should be taken into consideration. According to the data of some authors [5, 6], decrease of step rate to 125 steps per minute is accompanied in normal subjects by locomotion velocity increment due to the fact, that limb growth has relatively greater effect on movement velocity index (Fig. 1).

Baskakova, Vitenzon [7] have demonstrated that linear character of step length and step number relationship is maintained in case of rate increase up to 120 steps per minute. Break of this relation in patients with limitations of walking rate or limb length should lead to increase of the level of locomotion energy support if movement velocity increase is required. Changing walking rate or step length of the observed normal subjects, the authors have made the conclusion, that walking with different rate and step length presents two independent motor problems. In case of average-rate walking muscular work, determining step rate, arranges to the frequency of lower limb free vibrations [8].

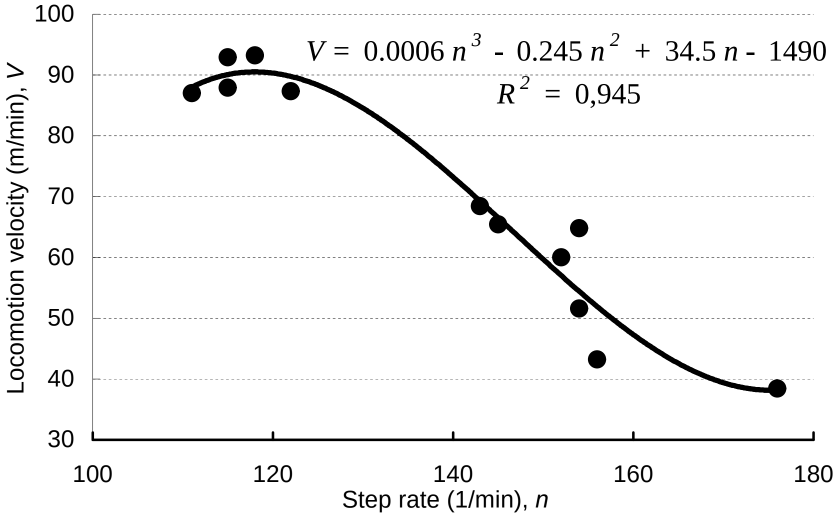

Fig. 1. Dependence of locomotion velocity in normal subjects on step rate (the curve is plotted according to the data of Muray, 1981, [5]; Sutherland et al., 1988, [6]).

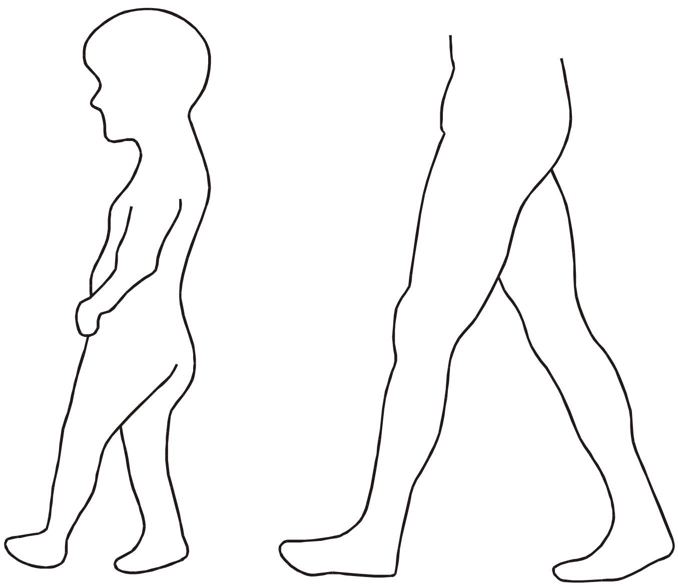

Fig. 2. A diagram, making clear difference of limb length and step in patients with achondroplasia before treatment and in normal subjects of the same age.

We have a unique possibility to test the above-mentioned principles concerning the effect of limb longitudinal growth on walking biomechanical characteristics, while studying patients with achondroplasia before and after their elongation according to the Ilizarov technique. Their selective retardation of longitudinal growth of tubular bones was almost two-fold.

Material and Methods

50 normal children and adolescents were studied as well as 48 patients with achondroplasia before treatment and in the long-term periods after surgical elongation of both lower limbs according to the Ilizarov technique at Russian Ilizarov Scientific Centre “Restorative Traumatology and Orthopaedics” (RISC “RTO”) at the expense of femoral and leg length increase. Lengths of lower limbs and body were measured, step length was

calculated (as the quotient obtained when the length of 100 m way part, covered with a usual rate, is divided by the number of steps), walking velocity and increment of heart rate by walking were determined. Moreover, daily motor activity (the length of daily covered way in km) was evaluated, using stepmeters (in view of way decrement in the control way part and step length).

Results and Discussion

Length of lower limbs in patients with achondroplasia is 58% of that in normal subjects of the same age, and length of their step is shorter (Fig. 2). It was established, that step length also became larger in normal children and adolescents as far as length of their lower limbs increased with age (Fig. 3). In patients with achondroplasia step length increased as well as far as limbs grew, however, the slope of linear regression equation was twice less, probably due to comparatively larger steady component. Evidently, patients were to increase step length. After surgical elongation of limbs with growth lag behind the revealed character of index relationship did not change. However, despite the increase of absolute values, the step of the patients after treatment was shorter in comparison with normal subjects of the same age.

The range of angular movements in the hip during walking decreased from 70° to 50° in the studied normal subjects as far as lower limb length increased with age. Not only step length, but duration of the step period as well is associated with limb length.

While determining proper duration of step period, using a known law, connecting simple pendulum string length (L) and its oscillation period (T=2η√L/g), it was revealed, that the length of conventional pendulum should be 45-55% of limb length for studied normal subjects.

Really obtained indices of locomotor function in patients with achondroplasia were compared with results of the mathematical calculations, based on the account of the effect of the length of the conventional pendulum on the studied indices.

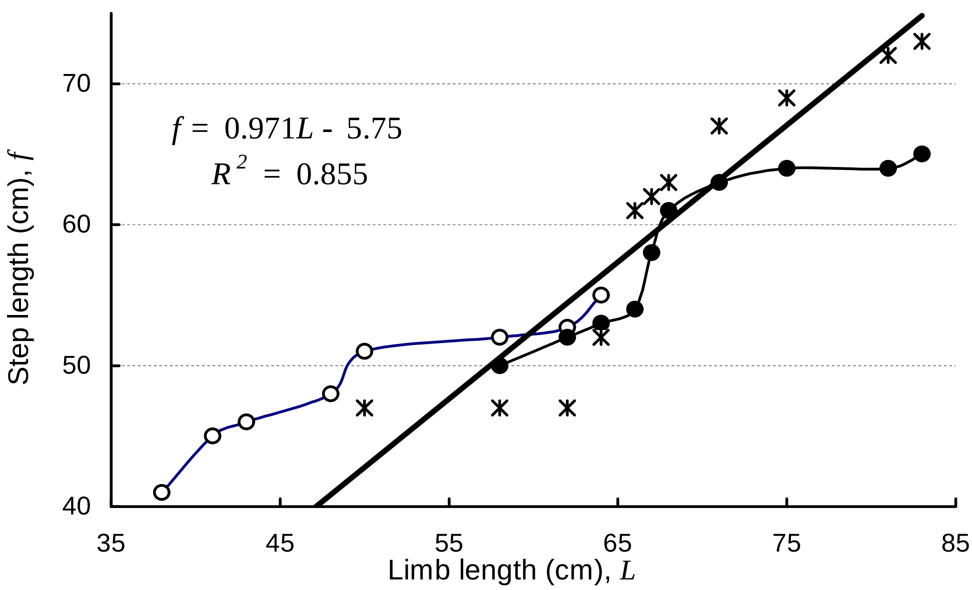

Fig. 3. Limb length - step length relation in patients before treatment (rings), after limb elongation (circles) and in normal children (solid straight line).

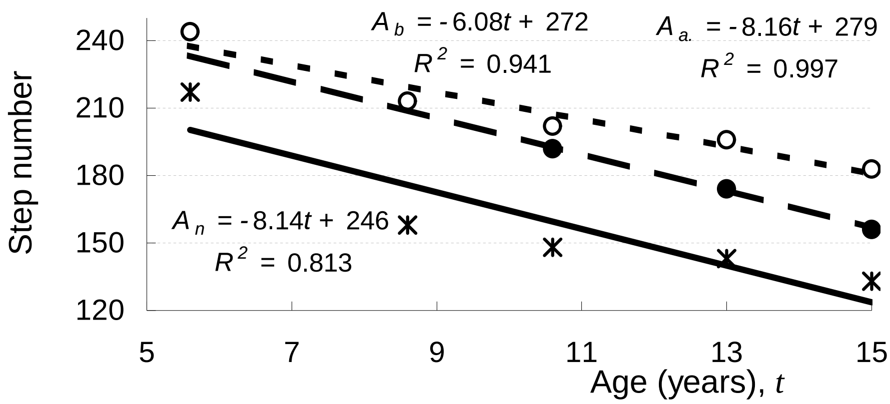

Fig. 4. Age dynamics of step number in the control way part in normal children, in patients before treatment (dotted line) and after limb elongation (dash line).

While studying locomotory activity it was revealed, that number of steps, required to cover the control way part, decreased with age in patients both before treatment and after it as far as limb length increased (Fig. 4). Mean locomotion velocity increased by 30% with age (5-15 years). For children it took 134 sec. to cover the control way part and for adolescents – 92 sec. The index of daily locomotor activity in patients with achondroplasia increased with age, coming to 1.81±0.39 km for children of 6-7 years and to 6.10±0.68 km for adolescents. Step length in patients before treatment was 47.5 cm on the average.

Lower limb length in patients before treatment was 49 cm on the average, number of steps for the control way part was equal to 110. The length of the conventional pendulum turned out to be significantly more than the calculated one and coincided with the limb length. Taking mass of limb tissues and their distribution into consideration, one can state that before treatment movement of limbs during walking does not coincide with the frequency of their free oscillations. Hence, it’s easy to understand why energy value of locomotions in patients increases and daily locomotor activity decreases.

After treatment the length of the conventional pendulum in patients was 54% of limb length, corresponding to the calculated normal length. After complete two-stage elongation the average length of lower limbs reached 71.3 cm. In view of the length of the conventional pendulum, the period of oscillations was equal to 1.25 sec., corresponding to 96 steps per minute, while the true value was 97 sec. After 45% lengthening (on the average) of limbs the step length increased by 24% and reached 59 cm.

Pulse rate in patients after covering the control way part increased by 17% and reached 98±2 beats per minute. After treatment energy value of walking, estimated by the pulse rate increment, decreased. After covering the control way part increment of heart rate decreased to 10.8±0.7 beats per minute.

Number of steps in the control way part was 169±5, that is 16% less, than that before treatment (see Fig. 4). Locomotion velocity increased by 15%, index of daily average locomotor activity came to 7.10±0.89 km, that is more by 51%, than before treatment.

Thus, both marked lower limb longitudinal growth lag behind with respect to the level of normal subjects of the same age and breaches of proportions of body parts have a significant effect on locomotor activity condition in patients with achondroplasia before treatment, who had relatively short steps, low indices of walking velocity and daily locomotor activity. Complete two-stage correction of height allowed to improve the main indices of locomotor activity in patients after treatment. Relative step length of patients decreased, step period did not change practically and approached the calculated value, walking velocity

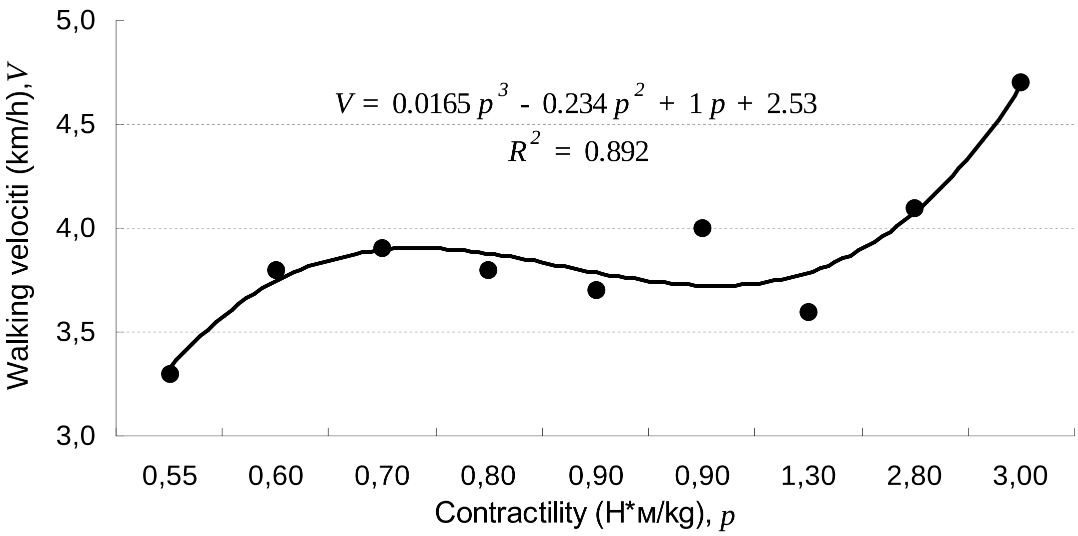

Fig. 5. Dependence of walking velocity in patients before and after treatment and in normal children on contractility of leg extensors.

increased by 15%, daily locomotory activity increased by 51%, period of covering the control way part decreased. Nevertheless, it was rather difficult to achieve the indices of locomotory activity, typical of normal subjects of the same age, probably due to attendant lesion of locomotor organs, anomaly of pelvic ring anatomical structure, specifically femoral neck shortening. The latter fact contributed to development of "waddling gait" in patients due to their desire for step length increase, due to mismatch of step rate and lower limbs' free oscillations frequency.

After limb elongation, indices of locomotor activity depended on the level of functional rehabilitation of joints and muscles. The amounts of limb segmental lengthening, found empirically on the basis of long clinical experience, probably, are close to optimal ones, because in case of their excess, particularly in femoral elongation, a delay is noted concerning restoration rate of muscular contractility, which limits increase of the level of the most important indices of locomotor activity in patients. During the analysis of the relationship of indices of walking velocity, daily locomotor activity and restoration level of contractility in different muscular groups, it was revealed, that these indices depended on the level of functional rehabilitation of leg extensors (Fig. 5).

Acknowledgment

We would like to thank N.G. Alfonsova for her help in translation of the manuscript.

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ТЕОРЕТИЧЕСКИЕ ПРЕДПОСЫЛКИ И ПРАКТИЧЕСКИЕ ПОСЛЕДСТВИЯ УВЕЛИЧЕНИЯ ДЛИНЫ НИЖНИХ КОНЕЧНОСТЕЙ У БОЛЬНЫХ С АХОНДРОПЛАЗИЕЙ

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Ахондроплазия является врожденным заболеванием нарушения остеогенеза, приводящим к очевидной при рождении карликовости с короткими конечностями, но нормальным туловищем. У здоровых детей и подростков (в группе из 50 человек) по мере возрастного увеличения длины нижних конечностей, а также после удлинения конечностей по методу Илизарова у больных ахондроплазией (в группе из 48 человек) увеличивается длина шага. У больных до лечения отмечались относительно короткий шаг, низкие показатели скорости ходьбы и суточной локомоторной активности. После оперативного увеличения длины конечностей период шага приблизился к должным расчетным показателям, скорость ходьбы увеличилась на 15%, суточная локомоторная активность - на 51%. Показатели локомоторной двигательной активности существенно зависели от уровня функциональной реабилитации суставов и мышц. Тем не менее, достигнуть показателей локомоторной активности, характерных для здоровых сверстников, не удается, по-видимому, из-за сопутствующих поражений органов движения, аномалии анатомического строения тазового кольца. Последнее обстоятельство способствовало развития у больных ”утиной походки” из-за стремления увеличивать длину шага, рассогласования частоты шагов с собственной частотой колебаний нижних конечностей. Библ. 8.

Ключевые слова: ахондроплазия, метод Илизарова, конечности, биомеханический анализ, остеогенез

Received 25 July 2000