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Factors influencing motor development in children with Down syndrome*

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Abstract

Psychomotor development in children with Down syndrome is affected in both the motor and the mental component. Motor development in a child with Down syndrome typically involves a delay in the attainment of the gross motor milestones achieved during the first year in non-impaired children, such as standing, sitting, crawling, reaching, rolling and walking. Furthermore, alterations may appear in fine motor development, visual motor control, speed, muscle strength, and static and dynamic balance.

Motor development is hindered to a large extent by hypotonia and ligament laxity and by constitutional problems such as shortness of the upper and lower limbs in relation to the trunk. Another reason for delayed acquisition of motor items can be DS-related medical problems, such as heart, stomach, intestine and respiratory problems, and ear canal infections.

Frequently, people with Down syndrome show alterations in their locomotor system due to an association of more or less pronounced muscle hypotonia and joint laxity. Joint instability increases because joint soft tissues are less functional. As a result, joints which undergo a more continuous load (hips, knees, feet) or are

subject to great mobility (atlantoaxial joint) tend to be more affected.

Keywords: Development. Down syndrome. Hypotonia. Joint instability. Psychomotor disorders.

1. Introduction

Motor development, one of the most easily observable areas of human development within just a few days of birth, requires structural, medical, cognitive, educational, emotional and other aspects to work in harmony. Good motor performance is not only sound functioning but also entails certain appropriate movement patterns.

Unlike other disabilities, when working with people who have Down syndrome (DS) we must aim for them not only to acquire functional motor activity, but also for this to be as good as possible, since many such individuals display compensatory patterns such as picking an object up off the floor with their knees locked in extension, or needing to lean on something to walk up or down stairs. Most people with DS typically share a lack of dexterity due to some degree of hypotonia, joint laxity and poor coordination and balance. As a result of this, they move differently.

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Motor development is neither a static process with a beginning and an end nor something that occurs at a specific age, though it is often associated with infancy and childhood. For Escribá (2002), development comprises all activities that are newly learnt and that allow people to perform actions they did not perform previously; therefore, it can also occur much later in life (1).

Motor development is not only contingent upon biological factors. Theory suggests that a person's motor behavior is determined by a set of systems that interact dynamically to produce movement. This set of systems, which includes the central nervous system, the musculoskeletal system, motivation, alertness, bodily growth, muscle strength, perception, cognition, the environment and so forth, works in self-organization to produce movement and motor development in the infant or child (2).

It is also important to consider the features of motor learning in children with DS which are common to all children's learning, with some subtle differences:

- Children with DS learn when they are motivated and eager to learn (playing, interest, etc.). Without motivation, movements are not integrated as easily as when they are made on the child's own initiative. The reward for the child must be greater than the effort involved in movement.

- Learning is better and quicker when positive reinforcement is given for the things the child can do.

- Children with DS learn to strive if the people around them provide support and the necessary resources to facilitate motor activity.

- They learn when difficulty increases gradually, apace with their learning. They tend to shy away from new challenges or those which involve learning beyond their level.

- They learn more easily in situations that are meaningful for them (at home, in the park, at nursery, etc.).

- They learn at their own individual pace, which is slower than the rest of the population's. Learning rate can even vary from one stage to the next in the same child.

- Learning is enhanced through imitation (visual information) and in situations involving social relationships (siblings, nursery classmates, parents, etc.). They prefer social interaction to handling objects.

- They learn when they are given enough time to react (they need more time than other children).

- They learn by a process of trial-error-correction-repetition, among others. Space and time should be provided for these children to experience their learning on their own and spontaneously shape and integrate it.

- They learn when communication with their environment is adequate. They need to understand what is being asked of them. If the task is unsuited to their level of understanding they will often freeze up or display rejection behavior towards all proposals; any attempt to provide guidance will be interpreted as an invasion.

It is also important to try to understand the child and respect his or her decisions. Children with DS often begin to speak long after they begin to understand what is said to them. Wanting to express themselves and being misinterpreted can lead to a feeling of powerlessness. Being understood makes them feel better about themselves and results in more appropriate behavior. In motor play, this is revealed when they frequently say 'no more', 'something else', 'yes', 'help me' and so forth.

2. Motor development conditioned by the features of Down syndrome

Gross motor development in infants and children with DS is also influenced by structural factors that are typical of the syndrome: brain features, musculoskeletal abnormalities and medical issues.

2.1 Brain alterations

The genetic information supplied by an extra chromosome 21 (or part of it) tends to cause pathological expression in the brain function and structure of people with DS. However, this information is also regulated by the individual's other genes, which gives rise to considerable variability amongst people with DS. Moreover, the brain is not a fixed or unchanging structure but is highly plastic, so environment and intervention can have a decisive influence on individual development.

Reduced brain differentiation from the central nervous system and delayed brain maturation associated with mild cortical dysplasia result in fewer neurons, abnormal synaptogenesis and delayed brain development (3).

The excess gene load causes a diffuse and widespread imbalance in the brains of people with DS. The following alterations are manifest:

- Lower brain weight and volume, usually coupled with smaller head circumference.
- Reduced neuron density (cerebral cortex, hippocampus, cerebellum, brainstem).
- Alterations in the structure and number of dendritic spines.
- Lower synaptic density, with characteristic morphology and a reduction in the number of neurotransmitters.
- Delayed myelination.

All of these initial abnormalities alter information-transmitting capacity, which has a direct effect on the subsequent assembly of neural circuits and connections, even in areas that have not been directly affected by the genetic alteration per se (4).

Because of this unique brain configuration, people with Down syndrome are slower to uptake, process, interpret and work out information, which is one of the causes behind the slower motor and cognitive development documented by numerous authors (1). They struggle to organize attention, memory, abstraction and deduction.

Motor characteristics in children with DS are determined by abnormalities in various brain structures, including the midbrain and the cerebellum, which determine muscle tone, balance and coordination.

Midbrain alterations in infancy reduce the child's alertness. Hypotonia, poor reaction to stimuli, difficulties in fixing the gaze on visual stimuli and interacting with the gaze of others, clumsiness and poor motor response, and a lack of initiative for seeking behavior may all be caused by a weaker involvement of neural systems associated with the midbrain. All of this has an influence on the all-important information input during a baby's first months.

The cerebellum is involved in performing precise, accurate body movements. It uses both proprioceptive information and kinesthetic sensations to execute movements well. It influences the tone that the various muscle groups must develop, contributes to holding balance and helps to perform movement patterns, particularly the more rapid, consecutive and simultaneous these movements are. As such, it appears to be necessary for learning and remembering pre-programmed motor sequences.

The prefrontal cortex is involved in decision making and initiating actions, processes which in turn require information to be integrated and processed. The hippocampus plays a role in memory, specifically in storing and integrating information in long-term memory.

The central nervous system in people with DS undergoes early ageing, adding new brain alterations to those already present. Head circumference growth slows down in early childhood, leading to microcephaly (5).

2.2. Musculoskeletal system alterations.

2.2.1. Hypotonia and ligament laxity

Hypotonia in people with DS is central in origin. The outcome of this hypotonia is that the muscles do not exert enough force on the joint structures. Added to the laxity in the ligament tissues and the joint capsule, this leads to increased joint motility in most joints, which very often makes them unstable (Figure 1). As a result, it is more difficult to achieve good balance and good movement coordination.

In cases where there is a serious heart defect, hypotonia is greater and motor development in these children is therefore further delayed compared to children with DS who do not have heart disease (5).

2.2.2. Muscle strength

Both muscle hypotonia and lack of exercise (sedentary habits, passivity) lie behind muscle fatigue which can be interpreted as a lack of strength, though this is highly variable.

2.2.3 Body size

Since the proportions of the long bones are diminished, children with DS have a significantly lower average height than their age group. This is because their legs tend to be shorter; sitting height is normal because this height deficit occurs largely before puberty.

DS-specific body proportions must be taken into account for their possible negative effects on strength, posture, locomotion and object handling (6).

2.3. Medical aspects.

People with DS tend to have medical problems that disrupt their development and therefore lack the optimum capacities needed to be alert and to react positively to their environment. If this situation is compounded by a low baseline level of activity, particularly at an early age, long periods of reduced motor practice will ensue and motor skill acquisitions will appear to be at a standstill.

Some of the most common medical conditions

2.3.2 Respiratory problems

Respiratory problems in DS are commonly related to cardiac, immunological or neurological complications, and are often secondary to a dysfunction of more than one system.

Hypotonia and the anatomical structure of the upper respiratory tract predispose people with DS to airway patency issues. These escalate in early childhood when children begin school, as they come into contact with germs more regularly and because their lung and immune system structures are not yet fully mature. These are often the primary causes of trips to the emergency department and hospital admissions among very young children. Middle ear infections and accumulated secretion are also frequent.

Another dysfunction that may appear is obstructive sleep apnea.

The smallness of the oral cavity together, macroglossia where present, and mucus in the upper airways usually result in a habit of mouth breathing which is extremely difficult to re-educate and may increase a propensity to develop problems in the lower respiratory tract.

All of these disorders (bronchitis, bronchiolitis, pneumonia and so on) lead children to become rather inactive, and in many cases full bed rest is prescribed.

2.3.3 Visual and auditory problems

Hearing and sight problems are very common in people with DS. If input structures are not in optimum condition, the intake of environmental stimuli is distorted and responses less than ideal, with the consequence that development is disrupted. Early, regular sight and hearing checks are therefore important.

The most frequent eye problems in DS are refraction errors (65-70%), strabismus, nystagmus, blepharoconjunctivitis, cataracts, keratoconus, optic nerve hypoplasia and retinal disorders.

Hearing capacity is usually below normal, with a high percentage of hearing loss. Hearing difficulties can affect the capacity for space-time structuring (1).

2.3.4 Gastrointestinal problems and hormonal imbalances

Disorders and malformations of the digestive tract or intestinal obstructions requiring surgery in the first few days after birth are present in 10 to 12% of babies with DS. Gastrointestinal problems present at birth, with symptoms such as poor

Figure 1. Ten-year-old girl in lateral hip rotation and abduction displaying major joint hypermobility.

include heart disease, respiratory problems, visual and auditory disabilities, digestive and hormonal problems, and epilepsy.

2.3.1 Cardiac problems.

Cardiac deformities affect 40% of people with DS. These must be detected as early as possible and appropriate action taken through either medical or surgical treatment, which considerably enhance quality of life.

Heart problems limit or determine motor performance in people with DS. They bring on premature exhaustion, which makes physical effort highly limited or contraindicated.

In the first few months, when many babies with DS are still awaiting surgery or a stabilizing drug regimen, motor efforts must be very gentle, with frequent breaks to avoid causing a state of crisis; but they must be kept up. Typical issues in the infant with heart disease are feeding difficulties, stunted growth, dyspnea, cyanosis, easy tiring and sweating.

Should the problem persist as the child ages, exhaustion will continue to arise easily. It leads to difficulty in performing activities that require considerable cardiovascular effort.

feeding, vomiting, swollen abdomen or absence of stool.

With regard to hormones, thyroid problems are common, particularly hypothyroidism, which presents with low energy levels, delayed physical and mental development, thickening of the skin, constipation, and drowsiness.

Infants with DS are born with an average weight and maintain this throughout childhood. Weight gain is normal after this stage, with a certain tendency to become overweight and even obese. A sedentary lifestyle and poor nutrition are the most common predisposing causes of obesity, apart from medical conditions (heart disease, hypothyroidism, muscular hypotonia and so forth) (Figure 2).

2.3.5. Epilepsy

The rate of epilepsy in children with DS is 2-15%, which is higher than that for the general population but lower than rates for other syndromes involving mental retardation. A strikingly high number of children experience onset in their first few years, 82% before they are five. The causes of this epilepsy are altered brain development, strokes secondary to heart disease, and perinatal asphyxia (5).

3. Delayed motor development

Children with DS do attain the same motor milestones as normal children, albeit at a later stage. Broadly speaking, the milestones that are most delayed are those which require a high level of coordination and which involve shifting weight (belly crawling, independent walking and so on).

Tables I, II and III display data from various sources in the literature (1, 7, 8) on psychomotor development in the early developmental stages of 'normal' infants as well as infants with DS. The periods in which motor milestones are acquired are very broad and variability is high, which makes it difficult to provide developmental prognoses. Figure 3 compares the data on the average age of main milestone acquisition in children with DS gathered by the three researchers from the tables above. Data should be gathered for a wide population sample using uniform criteria.

Of all the motor milestones, learning to walk is one of the most significant. Delays in walking affect other aspects such as the child's cognitive and social abilities, since being able to get around by walking independently is more than just deambulation. It allows children to develop

psychomotor capacities like gauging distance and depth, locating objects in space, body scheme and so forth. Walking is a major milestone in a person's self-sufficiency and achieving it therefore means less anxiety for the family (8).

Motor development may also be delayed by the medical, structural and neurological problems discussed above. Of these, hypotonia is the greatest hindrance to the child with DS. The initial hypotonia seen in babies with DS will continue throughout life although it may improve with growth. Its distribution pattern depends on each individual child and will chiefly affect functions in the area where it is most prevalent. If it predominantly affects the upper extremities, the infant will have trouble belly crawling and getting to stand up; if it predominantly affects the torso muscles, there will be more difficulty crawling and learning balance when standing. Children will tend to avoid doing the things they find hardest.

In terms of body structure, limbs that are short in relation to torso length will hinder the ability to use arms for support when sitting or climbing up stairs, or onto a sofa, and so forth (11) (Figure 4).

All of these factors must be borne in mind by both the family and the professionals who treat the child. Familiarity with these issues will enable

Figure 2. Overweight teenager doing supervised physical exercise

Table I.

Psychomotor milestone acquisition age according to
Escribà. 2001 (1)

Milestone	Down Syndrome (months)		General population (months)	
	Average	Range	Average	Range
Smiling	2	1-3	1	1-3
Rolling	6	2-12	5	2-10
Sitting	9	6-18	7	5-9
Belly crawling	11	7-21	8	6-11
Crawling	13	8-25	10	7-13
Standing	10	10-32	11	8-16
Caminar	20	12-45	13	8-18

Table II.

Psychomotor skill milestone acquisition age according
to Candel. 2003 (7)

Milestone	Average acquisition age in DS (months)	Range in DS
Head control when prone	2.7	1-9
Head control when held vertical	4.4	3-14
Rolling	8.0	4-13
Lateral support reaction	8.2	6-12
Sitting unsupported	9.7	7-17
Standing unsupported	13.3	8-24
Belly crawling	13.6	7-24
Crawling	17.7	9-36
Walking independently	24.1	16-39

Table III.

Psychomotor milestone acquisition age in DS according to
Winders, P. 1997.

Milestone	Average in months
Rolling from back to stomach	7
Rolling from stomach to back	6
Sitting	11
Pivoting 360° while prone	10
Standing on all fours	14
Sits up by moving to the side	17
Belly crawling	14
Crawling	17
Walking independently	26

carers to make more accurate, reasoned choices about how to help the child perform movements. Movement allows children to explore and relate to the world, a fundamental factor in their psychological development since physical and psychological development are very closely entwined.

Figure 3. Data comparison on motor milestone acquisition.**Figure 4.** Sitting with lower limbs in abduction. Short upper limbs.

4. References

1. Escribà A. Síndrome de Down, Propuestas de intervención. Madrid: Gimno; 2002.
2. Macías L. Desarrollo motor y aprendizaje del movimiento, Conceptos contemporáneos. En: Macías L, Fagoaga J. Fisioterapia en Pediatría. Madrid: McGraw-Hill Interamericana; 2003. p. 1-29.

3. Wisniewski K, Kida E y Brown WT. Repercusión de las Anormalidades Genéticas del Síndrome de Down sobre la Estructura y Función Cerebral. En: Rondal J, Perera J, Nadel L, Comblain A. Síndrome de Down: perspectivas psicológica, psicobiológica y socio-educacional. Madrid: Ministerio de Trabajo y Asuntos Sociales; 1997.
4. Flórez J, Troncoso MV. Síndrome de Down y Educación. Santander: Fundación Síndrome de Down de Cantabria; 1991.
5. Gassió R. Trastornos neurológicos y el niño con síndrome de Down. En: Corretger JM, Serés A, Casaldàliga J, Trias K, (eds). Síndrome de Down. Aspectos médicos actuales. Barcelona: Masson y Fundació Catalana Síndrome de Down; 2005. p. 70-82.
6. Burns Y, Gunn P. El síndrome de Down, Estimulación y actividad motora. Barcelona: Herder; 1995.
7. Candel I. Atención Temprana, niños con síndrome de Down y otros problemas del desarrollo. Madrid: FEIDS; 2003.
8. Winders P. Gross Motor Skills in children with Down Syndrome. Bethesda U.S.A.: Woodbine House; 1997.
9. Pueschel SM, Pueschel JK. Síndrome de Down. Problemática biomédica. Barcelona, Masson-Salvat Medicina, 1994.
10. Póo P, Gassió R, Desarrollo motor en niños con síndrome de Down, SD-DS Revista Médica Internacional sobre el síndrome de Down, 2000; 4: 34-40.
11. Ulrich DA, Ulrich BD, Angulo-Kinzler R, Yun J. Treadmill training of infants with Down syndrome: evidence-based developmental outcomes. Pediatrics 2001; 108: e84.