Letters to the editor

'Developmental impairment that is not immaturity'

SIR—Beckung et al. should be commended for their thorough study. Their results provide a comprehensive picture of gross motor development in Angelman syndrome that takes into account the heterogeneity encountered in this condition. They also confirm the importance of clinical diagnosis. Their questioning of whether the severity of neurological abnormalities seen in Angelman syndrome justifies its inclusion within the framework of cerebral palsy echoes a recent analytic study of this concept. Nevertheless, their conclusion that 'motor problems in individuals with Angelman syndrome mainly represent immature uncoordinated movement patterns, similar to what is seen in the early stages of motor development' needs clarifying. This conclusion implies that they organize their motor behaviour in the same way as individuals without disability who have no experience of the considered motor tasks. It appears to be contingent to the authors' quantification of motor competence as 'motor age', which fails to evaluate a dimension that links motor impairment to function, namely motor control. We studied the kinematics of intersegmental coordination in locomotion in toddlers without disability from their first steps through walking development and in children with Angelman syndrome. In normal 'mature' walking, this coordination is characterized by highly stereotyped coupling of elevation angles of the thigh, shank, and foot during the gait cycle, which has been termed the planar covariation. The planar covariation reflects the dynamic integration of postural stability with respect to gravity and forward progression; it correlates reliably with mechanical energy expenditure. For their very first steps, normal toddlers did not show planar covariation of the elevation angles of the thigh, shank, and foot during the gait cycle (Fig. 1Aii). In contrast, in children with Angelman syndrome the covariation of the elevation angles of the lower limb segments was constrained on a plane (Fig. 1Bii) whose spatial orientation significantly differed from normal controls. In addition, they did not show physiological in-phase forward rotation of the shank and foot at the end of the swing phase or in-phase backward rotation of these segments after the heel strike while the thigh elevation angle remained constant. We also studied the squatting movement in a group of 10 children aged 7 to 13 with Angelman syndrome and in four children without disability but with minimal experience of unsupported standing (3 to 9 weeks following the onset of walk-

Figure 1: (A) Child without disability (age 11 mo). (B) Child with Angelman syndrome. (Ai) and (Bi) are Sagittal kinograms: superimposed segments representing a model of body defined by opto-electronic recording of markers placed on anatomic landmarks (nose, ear, acromion, antero-superior iliac spine, trochanter, knee, malleolus, 5th metatarsal). Two consecutive segments = 20 ms. (Aii, iii), (Bii, iii) show covariation of elevation angles at thigh, shank, and foot during two successive steps at onset of unsupported walking. Sampling rate = 100 Hz. Mean value of each angular coordinate has been subtracted. Data are represented with respect to cubic frame of angular coordinates and best fitting plane (grid) in two different perspectives. Gait cycle paths progress in time in counter-clockwise direction, ground contact and toe-off phases corresponding roughly to the top and bottom of loops, respectively.
ing). The latter performed the movement using motor strategies that were different from that seen in Angelman syndrome. Toddlers without disability either kept their trunk erect and performed a low magnitude knee flexion (Fig. 2a) or they showed marked trunk and lower limb joint flexion (Fig. 2b), including ankle plantar flexion, in contrast to the pattern shown by children with Angelman syndrome (Fig. 2c). Both patterns (Fig. 2a, b) could be observed in one child, but no intermediate pattern was seen. The trunk-erect pattern was successfully recorded four times, the global flexor pattern nine times. Moreover, greater intra-individual kinematic variability was found in both tasks in toddlers without disability than in children with Angelman syndrome, suggesting adaptability based on diversity, competition, and choice in the former and predominance of a stereotyped ‘fail-safe’ mechanisms in the latter.

Furthermore, angular orientation of the head was more stable in toddlers than in those with Angelman syndrome, suggesting a mode of control relying on a head-centred referential – as further refined in older normal children but not in Angelman syndrome. Finally, in a study of upper limb and lower limb isometric postural tasks, we consistently found hypersynchronous rhythmic bursts of electromyographic activity in 14 children with Angelman syndrome that was not observed in infants, toddlers, or children without disability.

The question of whether patients with developmental impairment organize their motor behaviour in an immature fashion has long been a matter of debate with regard to pathophysiology, diagnosis, and management. These paradigms demonstrate differences between motor patterns observed in Angelman syndrome and 'immature' children without disability, that could not be identified with gross motor function measurement.

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Bernard Dan MD PhD,a,b
Guy Chéron PhDb
aDepartment of Neurology, Hôpital Universitaire des Enfants Reine Fabiola, Free University of Brussels
bLaboratory of Movement Biomechanics, ISEP, Free University of Brussels, Brussels, Belgium

Correspondence to: bernard.dan@ulb.ac.be

References

Figure 2: Sagittal kinograms of children squatting from standing position. (a) Toddler without disability demonstrating trunk-erect pattern. (b) Toddler without disability demonstrating global flexor pattern. (c) Child with Angelman syndrome. Sampling rate = 12.5 Hz.