Temperament and Physical Performance in Children With Osteogenesis Imperfecta

Stacy J. Suskauer, MD*; Holly L. Cintas, PT, PhD‡; Joan C. Marini, MD, PhD§; and Lynn H. Gerber, MD‡

ABSTRACT. *Objective.* **Children with osteogenesis imperfecta (OI) must participate in therapy to achieve motor performance objectives. Their behavioral style may influence motor performance. For this reason, the temperament of children with types III or IV OI was assessed prospectively to 1) compare their temperament with that of nondisabled children, 2) investigate the relationship between temperament and gross motor performance, and 3) examine relationships among temperament, parental overprotection and coping, physical activity, muscle strength, and motor performance.**

Methods. **Age-appropriate Carey Temperament Scales, Brief Assessment of Motor Function (BAMF), and the Vulnerable Child/Overprotecting Parents Scale were completed for 35 children 1 to 12 years old. Additional measures included the Childhood Health Assessment Questionnaire, Parent Daily Hassles Scale, manual muscle testing, Pediatric Activity Record, and a Summed Severity Score. Spearman correlations and multiple regression were used to identify and predict significant relationships.**

Results. **Temperament of children with OI differed from age-based norms in only 1 domain: activity. Motor performance (BAMF) correlated significantly with 3 domains of temperament: persistence ($r = -.48$), approach ($r = -.34$), and activity ($r = .40$). Activity was also related to the ratio of head circumference to body length ($r = -.45$) and the number of fractures in the preceding year ($r = -.35$). Parents' reports of their daily hassles significantly correlated with several domains of the child's temperament. No significant relationships were identified between parental overprotection and temperament or motor performance.**

Conclusions. **The temperament of children with types III and IV OI does not differ from that of their nondisabled peers, with the exception of lower activity scores. Although it is considered a biological attribute, the expression of temperament, specifically activity, may be influenced by learned behaviors. Because gross motor performance is related to activity, persistence, and approach/avoidance, knowledge of an individual's temperament may enhance the child's ability to benefit from interventions to improve motor skill and activity levels.** *Pediatrics* **2003;111:e153–e161. URL: http://www. pediatrics.org/cgi/content/full/111/2/e153;** *osteogenesis imperfecta, temperament, motor performance.*

ABBREVIATIONS. OI, osteogenesis imperfecta; VCOPS, Vulnerable Child/Overprotecting Parents Scale; CHAQ, Childhood Health Assessment Questionnaire; PDH, Parent Daily Hassles Scale; BAMF, Brief Assessment of Motor Function; PAR, Pediatric Activity Record.

C hildren with osteogenesis imperfecta (OI) are an engaging group of youngsters who interact well with other children as well as with adults. Despite relatively severe physical impairments and body segmental disproportion, they seem to be socially adept from a young age. It is uncertain whether this is attributable to advanced social skill or because the small stature typical of children with OI makes the child appear more capable socially than is actually the case. We felt that studying the temperament of children with OI would provide insight into the style of interaction that many of these children display, and could help us develop more effective rehabilitation strategies. We decided to begin by determining whether the temperamental attributes of children with OI vary from those of children their age without disability.

Participation in structured intervention programs can be effective for improving the gross motor performance of children with OI and their ability to function more independently. However, the ability of the child to focus on therapeutic tasks and form a working partnership with family and members of the health care team may depend heavily on dimensions of the child's temperament such as adaptability, activity, distractibility, and approach/withdrawal. We wished to investigate the relationship between domains of temperament and physical performance to examine pathways for achieving optimal gains within a rehabilitation plan.

Many parents are closely involved in most aspects of their young children's lives, and children with OI often require additional assistance from parents to complete everyday tasks of mobility, grooming, and therapy interventions. Moreover, the child's fragility and parental attitudes toward fractures or treatment

interventions may influence their interactions such that a child's movement experiences may be limited or enhanced by these interactions. Because parenting factors may play a role in the physical performance of children, we wished to determine if parental overprotection and parent coping are related to the motor performance of their children with OI.

This study was designed to assess 9 dimensions of temperament for children with type III or IV OI for the following purposes: 1) to compare the temperament of children with OI with that of nondisabled children; 2) to evaluate the relationship of temperament to physical performance; and 3) to determine if temperament and/or physical performance are linked to additional factors such as parental overprotection and coping, and the child's level of independence, physical activity, strength, and severity of the disorder.

OI

OI is a heritable disorder of collagen synthesis resulting from a mutation in the *COL1A1* or *COL1A2* genes. In the mildest form of OI, Sillence type I, the collagen gene mutation causes premature termination of synthesis of $\alpha 1(I)$ chains, resulting in a quantitative decrease in collagen molecules.[1] The mutations are qualitative in the remaining Sillence types, II, III, and IV. The majority are point mutations in which the replacement of a glycine residue with a different amino acid subsequently hinders the folding of collagen molecules.[2] Single exon splicing defects, large deletions, insertions, and retained introns have also been described in types II, III, and IV,[3] resulting in qualitatively abnormal collagen molecules that may also be associated with a decrease in the amount of secreted collagen.[4]

Wide phenotypic variation exists. Individuals with OI can manifest the following impairments at varied levels of severity: osteopenia, fracture and/or deformation of bone, weak muscles, joint laxity, segmental asymmetry and disproportion, and short stature. Although some children need no assistive gait devices,
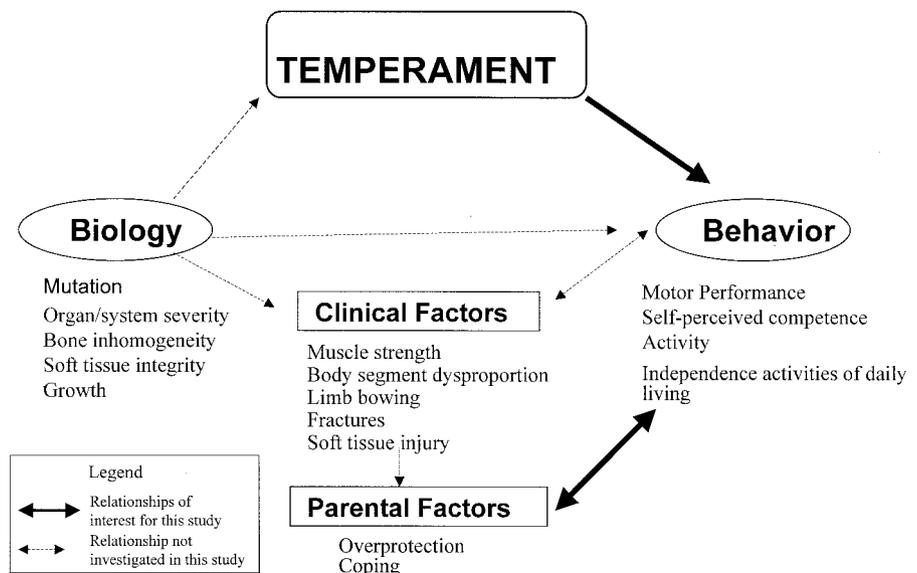
limitations associated with ambulation are common, and powered mobility may be required to achieve independent locomotion. Genotypic variation is an important contributor to disease severity, because heterogeneity in functional outcomes (such as ambulatory status) is apparent within subgroups of children with OI. However, some children with severe disease perform much better than would be expected on the basis of their structural limitations, and others with mild disease appear to perform well below their potential capability. It is likely that multiple factors, including the child's temperament, parental influences, and biological severity, may interact to promote or constrain observable motor performance. We chose to examine a subset of these relationships described in Fig 1.

Temperament

In the early 1950s, psychiatrists Thomas and Chess[5] proposed that intrinsic characteristics of a child's behavior influence the manner in which that child interacts with the environment. They chose the term temperament to represent behavioral style, or the manner of action, as opposed to the content or motivation of an action. Nine categories of temperament were identified: activity level (referring to general motion), first reaction (approach versus withdrawal), adaptability, threshold (of response to environmental stimuli), intensity of reaction, quality of mood, distractibility, persistence, and rhythmicity (regularity of bodily functions such as sleep, hunger, and bowel movements, and predictability of social behavior).[6] Twin studies support a genetic basis of temperament,[7,8] and longitudinal studies have demonstrated consistency in temperament from birth through early adulthood, especially after 2 years of age.[9–14]

Thomas and Chess originally interviewed parents as a means of assessing child temperament; other researchers have observed children in a laboratory or home setting to characterize behavioral style. Most studies, however, use parent-report questionnaires

Fig 1. Temperament and OI model illustrates relationships of interest.

TEMPERAMENT

Biology

Behavior

Mutation
Organ/system severity
Bone inhomogeneity
Soft tissue integrity
Growth

Clinical Factors

Muscle strength
Body segment dysproportion
Limb bowing
Fractures
Soft tissue injury

Motor Performance
Self-perceived competence
Activity

Independence activities of daily living

Parental Factors

Overprotection
Coping

Legend

Relationships of interest for this study

Relationship not investigated in this study

to measure temperament. A great benefit of parent-report measures is that they allow for responses based on a wealth of interaction in what is likely to be the child's most natural environment. Several investigators have examined the relationship between observer and maternal reports of temperament in infants between the ages of 3 and 24 months, and all report significant correlations.[15–18]

A child's behavioral style, apparently present at birth and consistent throughout childhood, influences interaction with friends, family, and health care providers. For that reason, it has been suggested that temperament plays a major role in the adjustment of children with disabilities.[19,20] Initially studies focused on the relationship between temperament and the psychological and social function of children with disabilities. Many investigators also chose to examine parent or family characteristics as they relate to a child's maladaptive behavior. For example, Lavigne and colleagues[21] studied the relationship of child temperament and coping ability, as well as family cohesiveness and organization, to psychological problems in children with myelomeningocele. They reported that "difficult temperament" and distractibility were associated with nearly half of the variance in internalizing behavior problems (such as phobias, somatic complaints, and obsessive/compulsive symptoms). Family cohesiveness was also correlated with behavior problems.

More recent research has been directed to understanding temperament as a factor in outcomes other than problem behaviors. Such studies, which are limited in number, have focused on temperament and other child characteristics, with less emphasis on parent and family variables. Two studies correlated temperament and glycemic control in children with type I diabetes. In 1988, using the Behavioral Style Questionnaire[22] and Middle Childhood Temperament Questionnaire,[23] Rovet and Ehrlich[24] reported that activity, rhythmicity, intensity of reaction, distractibility, and quality of mood accounted for 42% of the variance in glycosylated hemoglobin in children 6 to 13 years of age. Garrison and colleagues[25] employed the Dimensions of Temperament Scale[26] and reported that a child's activity, rhythmicity, and attention span were related to a subjective measure of the child's compliance with the diabetes regimen, while maternal factors predicted 90% of the variance in glycosylated hemoglobin. More recently Arnold et al[27] investigated the relationship of temperament and frontal lobe, motor, and cognitive function to phenylalanine levels in children with phenylketonuria. Using the Toddler Temperament Scale[28] and Behavioral Style Questionnaire,[22] they reported that difficult behavior is associated with higher current and average phenylalanine levels. Johnson and colleagues[29] used the Early Infancy Temperament Questionnaire[30] and the Infant Temperament Questionnaire[31] to measure temperament in infants with bronchopulmonary dysplasia. Medical treatments, dietary intake, and feeding problems were also assessed for the purpose of identifying risk factors for growth failure. They reported that difficult temperament was not significantly correlated with growth failure after the infants were discharged from the hospital.

Parental Overprotection and Vulnerable Children

In 1964 Green and Solnit[32] described the "vulnerable" child as one whose parents expect the child to die prematurely and who later demonstrates behavioral or developmental problems. They concluded that maternal resentment, guilt, and fear may lead to a sense of doom, failure, and a reluctance to face new challenges. Such emotions may promote avoidance of new situations and developmental delay. Furthermore, Green and Solnit proposed that the child may sense the parents' concern and subsequently develop a distorted self-image. The concept of vulnerability has since been expanded to include a broader grouping of children including those with chronic disease, premature birth, or gestational complications.[33–35]

Wright and colleagues[36] also describe a "vulnerable child/overprotective parents" syndrome. They explain that children who are perceived as physically vulnerable by their parents receive less than adequate stimulation and encouragement to attain their maximal physical and emotional capabilities. Thomasgard[37] defines parent overprotection as "an excessive and developmentally inappropriate level of parental protectiveness" and suggests that overprotection may influence competence in activities independent of parental involvement (such as academic performance), self-esteem, and social acceptance. Parental overprotection and perceived vulnerability have been shown to be significantly, though weakly, correlated.[34,35]

Although most assessments for perceived vulnerability and parental overprotection have been designed for use with parents of children who are not actually at increased risk for illness or injury, the Vulnerable Child/Overprotecting Parent Scale (VCOPS) was developed specifically for the purpose of quantifying overprotection by parents of children who are physically vulnerable.[36] Children with OI are appropriately classified as physically vulnerable due to their increased risk of bone fracture. Because parental overprotection might result in restricted opportunity for physical activity and developmental progress, we decided to examine the relationship between parental overprotection and physical performance in children with OI.

Thus, this study was designed to address these three questions: 1) How does the temperament of children with OI and disability compare to that of their nondisabled peers? 2) For children with OI, is there a relationship between temperament and measures of motor performance? 3) For children with OI, are there relationships among parental overprotection, parent-reported hassles, clinical severity, temperament, and physical performance?

METHODS

Participants

A convenience sample of 35 children (20 girls, 15 boys) ages 1 to 12 years (mean = 7.44, median = 8.21) with OI type III ($n = 18$) or IV ($n = 17$) was recruited from an established cohort of OI patients enrolled in an ongoing protocol at the National Institutes

of Health. Children are seen at 3-, 4-, 6-, or 12-month intervals depending on their age, and in which arm of the protocol they are enrolled. One infant (20 months old) has idiopathic thrombocytopenic purpura in addition to OI. There were no other significant comorbidities. Mothers of children were asked to complete the age-appropriate Carey Temperament Scale and the VCOPS parent report measures. Among the 34 mothers (1 mother had 2 children in the study), 1 is a stepmother who had lived with the child and her father for 2 years (biological concurrent mother deceased), and 1 has legally adopted her child (involved with child's care since the first year of life). Questionnaires were administered by the first author, and completed by mothers during a visit to the National Institutes of Health, through the mail, or by phone. In each case the Carey Temperament Scale was completed first, before the mother was presented with the VCOPS. The temperament and parenting questionnaires were completed in conjunction with a clinic visit in most cases, and clinical data were collected at the same time. In some cases, data were used from assessments completed on the previous visit (independence in self-care—Childhood Health Assessment Questionnaire [CHAQ]) and Parent Daily Hassles Scale [PDH]—are completed once each year).

Measures

Carey Temperament Scales (n = 35)

The Carey Temperament Scales are a series of 5 parent-report questionnaires used to assess the 9 dimensions of temperament in infants and children up to age 12 years.[22,23,28–30] The questionnaires have been shown to exhibit test-retest reliability and internal consistency, and normative data exist for ages 1 month to 12 years:

a. Early Infant Temperament Questionnaire (1–4 months)
b. Revised Infant Temperament Questionnaire (4–8 months)
c. Toddler Temperament Scale (1–2 years)
d. Behavioral Style Questionnaire (3–7 years)
e. Middle Childhood Temperament Questionnaire (8–12 years)

The 3 questionnaires spanning ages 1 through 12 years were used for the present study. Each questionnaire includes ~100 statements. The parent is asked to rate, on a 6-point scale, how frequently the child behaves in the manner described. For scoring, the items are separated into the 9 domains of temperament. Within each domain, each item is multiplied by a factor that depends on the parent's response. The sum of the products is totalled within each domain and then divided by the number of questions answered within the subscale. This average then becomes the category score for the temperament domain. Every child receives a category score for each of the 9 domains.

Brief Assessment of Motor Function (BAMF) (n = 35)

The BAMF was used to assess gross motor function.[38,39] This hierarchical scale is independent of age and designed for rapid description of the observable motor behaviors leading to independent bipedal locomotion. The Gross Motor Scale ranges from 0 (lifts head <45 degrees) to 10 (runs). Interrater reliability is intraclass correlation $(1, 1) = .996$, intrarater reliability is intraclass correlation $(3, 1) = 1.00$. Concurrent validity exists with gait parameters $(r = .68$-$.71; P < .0001)$, lower extremity manual muscle testing $(r = .74; P < .0001)$ and the Gross Motor Scale of the Peabody Developmental Motor Scales $(r = .95; P < .0001)$.

VCOPS (n = 35)

The VCOPS is a 28-item questionnaire on which parents rate, using a 6-point scale, the validity of statements concerning the parent's interaction with their child.[36] VCOPS items were selected from a pool of 301 items by physicians, psychologists, and child development specialists based on parent responses and stepwise multiple regression. Twenty-eight questions were found to provide maximum predictive efficiency of the parents' developmental stimulation tendencies. Test-retest reliability coefficients (with 2 different groups of subjects) were .74 and .77. Cronbach's α was .84, and the validity coefficient (based on health professional and day care work ratings) was .97.

CHAQ (n = 27)

The CHAQ is completed by a parent who evaluates the child's level of independence in self-care activities.[40] A higher CHAQ score indicates the child requires more assistance in self-care.

PDH (n = 27)

The PDH, a parent-report assessment of coping, measures the frequency and intensity of hassles experienced by parents in all aspects of their role as parents (not exclusively in relation to a child with special needs).[41] Higher scores indicate that the parents perceive that stresses occur more often (frequency scale) or are more intense (intensity scale).

Biological Severity and Clinical Mediators (n = 27)

Physicians involved in this study have extensive experience treating children with OI, and collaborated to enumerate those biological factors which appear to be linked to the severity of the disease. These measures of severity are: history of pulmonary infection requiring hospitalization and/or intravenous antibiotics, history of congestive heart failure, substantial radiographic inhomogenity in the epiphyseal bone, radiographic inhomogenity in the metaphyseal bone, and tubular bones on radiograph. Each factor was rated in a dichotomous fashion (1 = present, 0 = absent) for each child for whom data were available for all factors. The sum of the factors was used to create a total score for biological severity for each child.

As with biological severity, clinical conditions that could influence physical performance, and whose presence was likely related to activity, chance events, and other exposure, were identified. The mediators selected were number of lower extremity long bones bowed >30° on radiograph, number of documented fractures within the past 12 months, and number of surgeries in the past 12 months. These 3 clinical mediators were scored in the same dichotomous fashion described above, then summed with the biological severity score to produce a Summed Severity Score for those children for whom data were available on all factors described above.

Relative Body Proportions (n = 25)

Measurements of head circumference and chest circumference relative to total body length were included as potential influences on physical performance outcomes. Ratios of head and chest circumference to body length (using longer leg) were used to obtain relative measures.

Manual Muscle Testing (Kendall[42] 10-Point System; n = 20)

Manual muscle testing was used for evaluation of selected muscle groups for all children seen in the clinic who did not have a lower extremity fracture at the time of the visit. The score used for our analysis is the sum of the right and left hip abductors and extensors (maximum score = 40).

Pediatric Activity Record (PAR) (n = 16)

The PAR is a record of the amount of upright activity that the child performs at home during typical days (2 weekdays and 2 weekend days).[43] Completed by a caregiver or an adolescent patient every 6 months, the PAR includes the types of activity and the body position in which they occur, recorded daily. For this study, only the hours between wake-up and bedtime were included, and the score was calculated as the time spent upright in standing or walking as a fraction of the total time reported. A higher PAR score represents a greater percentage of time spent upright in standing or walking. The most recent PAR data within the past year were used for each child.

Data Analyses

For comparison of the temperament of children with OI with their nondisabled peers, scores for each temperament domain for each child were converted to z scores (using published norms for each age group) and analyzed using a 1-sample normal test. This was done to determine if the mean of the OI group was significantly different from zero. Bonferroni corrections were then used for multiple comparisons because we considered 9 separate, but closely related, domains.

Spearman correlations were used to identify the presence or absence of statistically significant relationships among the variables in relation to the domains of temperament or motor performance. Once significant relationships were identified, forward selection multiple regression was performed to determine the relative weights of factors contributing to motor performance, and

to identify predictors for criterion variables activity associated with temperament, and motor performance.

RESULTS

Children with OI were comparable to their non-disabled peers in all domains of temperament except one: activity. They were significantly less active than their peers without OI (Bonferroni adjusted $P = .001$). Descriptive statistics for the temperament domains and comparison measures are summarized in Table 1 and illustrated in Fig 2.

Three domains of temperament correlated significantly with gross motor performance (BAMF): persistence ($r = -.48$; $P = .003$), activity ($r = .40$; $P = .02$), and approach ($r = -.34$; $P = .05$). Thus, higher gross motor function linked to greater persistence, higher activity, and to approach rather than avoidant behaviors. Motor performance also correlated significantly with the ratio of chest circumference to body length ($r = -.78$; $P = .0001$), the ratio of head circumference to length ($r = -.73$; $P = .0001$), daily upright activities (PAR) ($r = .76$; $P = .0007$), and the Summed Severity Score ($r = -.60$; $P = .001$). Muscle strength values were also significantly related to BAMF scores ($r = .76$; $P = .0001$), but were not included in the stepwise regression model as data for the 2 muscle groups selected, hip abductors and extensors, were available for only 20 of the participants. Using these several parameters, individually significantly correlated with BAMF, in a forward selection multiple regression model resulted in 2 parameters included in the model: chest circumference to body length ratio ($R^2 = .59$; $F = 31.8$; $P = .0001$) and Summed Severity Score (incremental $R^2 = .07$; $F = 4.65$; $P = .04$) predicted 66% of the variance for BAMF.

The activity domain correlated significantly with 3 other domains of temperament: mood ($r = .47$; $P = .004$), adaptability ($r = .46$; $P = .005$), and intensity ($r = .40$; $P = .02$) (Table 2). The format of the Carey Scales is such that higher activity levels correlate with more negative mood, less adaptability, and more intensity. Following forward selection multiple regression, mood was the only predictor of the temperament activity domain ($R^2 = .20$; $F = 6.67$; $P = .016$). Significant negative correlations were identified between activity and the ratio of head circumference to body length ($r = -.45$; $P = .02$) and number of fractures in the preceding 12 months ($r = -.35$; $P = .05$).
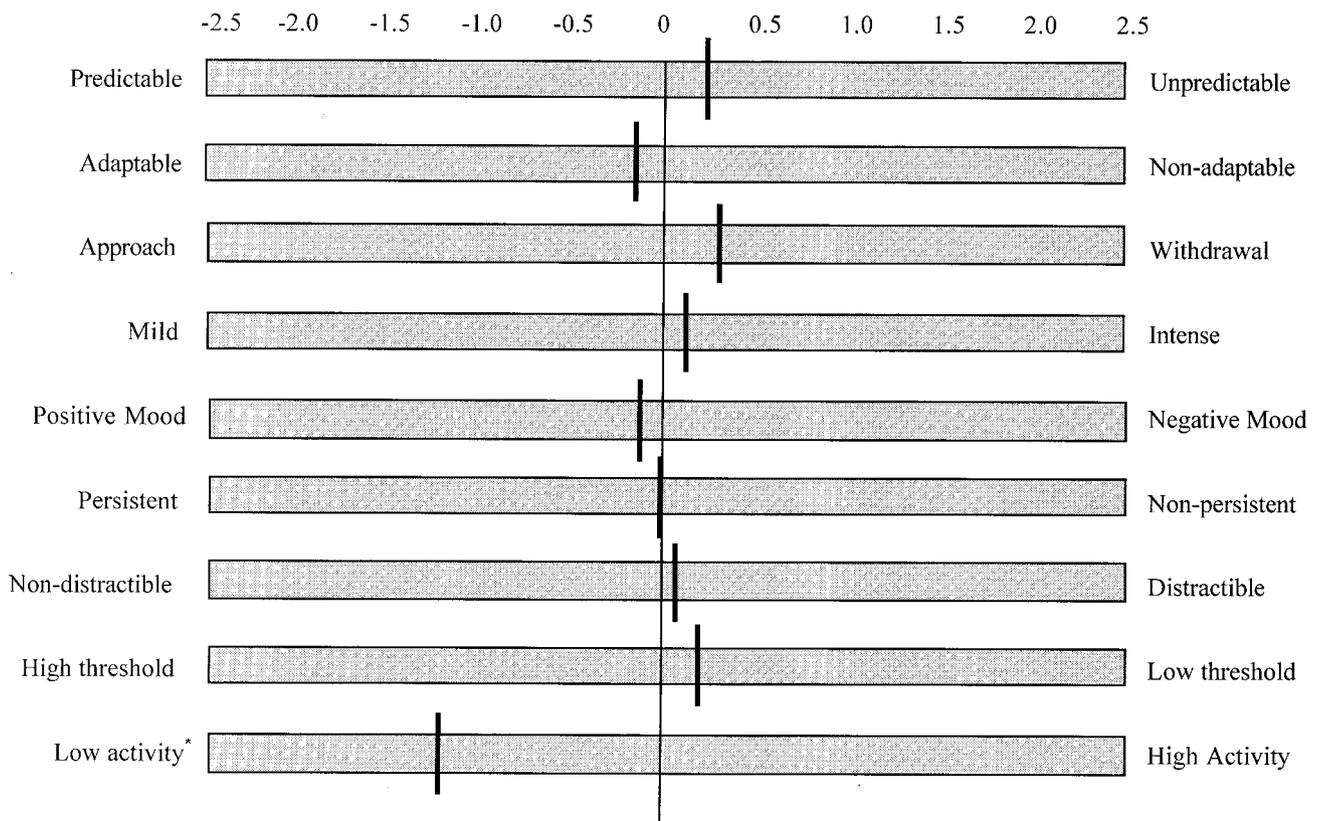
Independence in self-care (CHAQ) was inversely related to motor performance (BAMF) ($r = -.84$; $P = .0001$) and the temperament domain of activity ($r = -.45$; $P = .02$). Other variables significantly correlated with the CHAQ were persistence ($r = .40$; $P = .04$), head-to-length ratio ($r = .82$; $P = .0001$), chest-to-length ratio ($r = .81$; $P = .0001$), strength of hip abductors and extensors ($r = -.60$; $P = .005$), and the Summed Severity Score ($r = .59$; $P = .002$).

Parent coping assessed with the PDH correlated significantly with several domains of temperament. Frequency of parent daily hassles linked with children's mood ($r = .56$; $P = .003$), persistence ($r = .54$; $P = .004$), adaptability ($r = .47$; $P = .01$), and predictability ($r = .44$; $P = .02$). Intensity of parent daily hassles was associated with mood ($r = .48$; $P = .01$), persistence ($r = .47$; $P = .01$), and predictability ($r = .44$; $P = .02$). Higher frequency and intensity of hassles reported by parents were associated with child traits of negative mood, more intensity, less adaptability, and less predictability. Following forward selection multiple regression, mood predicted both the frequency ($R^2 = .38$; $F = 15.23$; $P = .0006$) and the intensity ($R^2 = .24$; $F = 8.08$; $P = .009$) of parents' daily hassles.

No significant relationships were identified between parental overprotection/child vulnerability (VCOPS) and any domains of temperament or physical performance.

TABLE 1. Descriptive Statistics for Variables

Measure	n	Mean	Median	Standard Deviation	Minimum	Maximum
Temperament						
Activity	35	−1.22	−1.34	0.93	−2.90	1.42
Adaptability	35	−0.15	−0.23	0.87	−1.79	2.00
Approach	35	0.29	0.11	0.98	−1.40	2.86
Mood	35	−0.13	−0.08	1.13	−2.54	2.06
Intensity	35	0.06	0.06	1.03	−1.43	2.15
Distractibility	35	−0.02	−0.02	0.98	−2.19	1.71
Persistence	35	−0.01	0.07	0.82	−1.24	1.76
Threshold	35	0.18	0.18	0.76	−1.24	1.85
Predictability	35	0.40	0.40	0.87	−1.49	1.84
Gross motor function (BAMF)	35	5.97	6	2.84	0	10
Parental overprotection (VCOPS)	35	−0.50	−0.52	1.07	−2.46	1.97
Independence in self-care (CHAQ)	27	1.91	2.25	0.97	0.25	3.00
Parent hassles—frequency (PDH frequency)	27	52.67	53	9.72	29	67
Parent hassles—intensity (PDH intensity)	27	43.33	41	11.3	24	65
Severity score	27	4.19	4	2.82	0	12
Manual muscle test (10-pt)	20	20.80	19	10.1	6	40
Upright activity (PAR)	16	0.17	0.15	0.13	0	0.5

	-2.5	-2.0	-1.5	-1.0	-0.5	0	0.5	1.0	1.5	2.0	2.5	
Predictable												Unpredictable
Adaptable												Non-adaptable
Approach												Withdrawal
Mild												Intense
Positive Mood												Negative Mood
Persistent												Non-persistent
Non-distractible												Distractible
High threshold												Low threshold
Low activity*												High Activity

*Children with OI differed significantly only in the Activity Domain
(Bonferroni adjusted P=.001)

Fig 2. Domains of temperament: children with OI compared with nondisabled peers using *z* scores (mean = 0).

TABLE 2. Significant Relationships for Activity (Temperament), Physical Performance, and Parent Coping

Physical Performance BAMF (n = 35)			Parent Coping PDH (n = 27)						Temperament Activity Subscale (n = 35)		
					Hassle Frequency		Hassle Intensity				
Activity	r = .40	P = .02	Adaptability	r = .47	P = .01				BAMF	r = .40	P = .02
Persistence	r = −.48	P = .004	Persistence	r = .54	P = .004	r = .47	P = .01		Adaptability	r = .47	P = .005
Approach/withdrawal	r = −.34	P = .05	Mood	r = .56	P = .003	r = .48	P = .01		Mood	r = .47	P = .004
Head:length ratio	r = −.73	P = .0001	Predictability	r = .44	P = .02	r = .44	P = .02		Intensity	r = .40	P = .02
Chest:length ratio	r = −.78	P = .0001							Head:		
									length ratio (n = 25)	r = −.45	P = .02
Clinical/biological severity	r = −.60	P = .001							Fractures (12 mo) (n = 27)	r = −.35	P = .05
Fractures (12 mo)	r = −.38	P = .05									

DISCUSSION

Our data suggest that, with the exception of hypo-activity, the temperament of children with types III and IV OI does not differ from that of their nondisabled peers. Gross motor performance (BAMF) was found to be related to 3 domains of temperament: persistence, activity, and approach (versus avoidance); however, the ratio of chest circumference to body length and the Summed Severity Score predict 66% of the variance for criterion variable BAMF. No relationships could be identified between parental overprotection, children's temperament, and motor performance. In contrast, parental coping ability, described by parents in terms of both frequency and intensity of daily hassles, correlated significantly with children's mood, persistence, and predictability.

The temperament of children with types III and IV OI differs from age-based norms only in the activity domain. It is not surprising to find that this cohort of children with OI, a musculoskeletal disorder, displays less activity when compared with nondisabled peers. The critical question is which factors contribute to hypoactivity, especially because motor skill

performance (BAMF) is significantly related to temperamental activity ($r = .40$; $P = .02$). It is important to emphasize that the activity associated with temperament and gross motor skill are not the same. Although some of the Carey questionnaire items that rate activity inquire about motor skills that children may have achieved, most questions are targeted to the intensity or vigor of nonpurposeful physical motion. Thus, the hypoactivity cannot be attributed solely to a delay in motor performance.

One would not expect physical impairments in children with OI to interfere with the expression of overall motor restlessness, however, in this population, activity may be influenced by biological determinants of disease, and/or learned behaviors. Studies of nondisabled children have shown that temperamental attributes tend to stabilize after the first 2 years of life. It is possible that early life experiences influence the child's expression of the activity domain of temperament. Perhaps children with OI learn very early that physical activity can lead to fractures and pain, and, as a consequence, they minimize activity. Temperamental activity does correlate significantly with the number of fractures in the previous 12 months ($r = -35$; $P = .05$). Multiple experiences in plaster after fractures and surgical procedures, which reinforce static behavior and limit spontaneous activity for long periods, may also contribute to generally lower activity levels.

The ratio of children's head circumference to body length was inversely associated with activity ($r = -.45$; $P = .02$) as well as motor performance ($r = -.73$; $P = .001$). This suggests that a disproportionately large head may contribute to a decrease in generalized activity, and also limit opportunities for motor skill practice and improvement. Infants with moderate to severe OI typically have heads that are dysproportional to their total length, and it is possible that relatively large head size in the early years is one of the factors that contributes to hypoactivity. Interestingly, chest circumference relative to body length does not significantly correlate with hypoactivity, although it is inversely related to motor performance ($r = -78$; $P = .001$). Perhaps chest propor-

tions in infants and young children with OI are not as atypical as they are in older children, and relative chest size, compared with head size, is less of a barrier to movement in infants and younger children.

Three among 9 domains of temperament were significantly and positively associated with motor performance (BAMF): persistence, approach, and activity. It is easily understood that persistence could engender improved motor function. This may be especially relevant for individuals with OI who deliberately work to achieve, then repeatedly reachieve, motor capabilities in a cyclic manner following fracture or surgery. Also, it is not surprising that children who display approach behaviors toward others, as opposed to avoidance, would be more successful interacting with health care professionals who can help them improve their motor function. Children with higher activity levels were also those found to have better motor skills. Possibly fidgety infants or children with high energy levels are more likely to interact with their surroundings, and, while doing so, expand and improve their motor capabilities. However, although activity and motor performance are positively associated, each, as noted above, has negative associations with biological and clinical factors (Table 3).

We identified significant inverse relationships between motor performance and the sum of clinical and biological severity factors. Impairments such as muscle weakness and decreased long bone growth, which are intrinsic to the disorder, clearly influence physical performance and may be so restricting that, even with therapeutic intervention, the child cannot surmount the physical obstacles to higher motor performance without compensatory devices. Manifestations of disease may affect the level of gross motor function that can be achieved by a child with OI, and the limit on performance increases with more severe disease. Notably, clinical and biological severity factors did not correlate significantly with any of the domains of temperament, suggesting that these factors may have a stronger influence on motor activities requiring strength and alignment, although both

TABLE 3. Significant Relationships Between Temperament Domains and Other Factors

	Temperament Activity	BAMF	VCOPS	CHAQ	PDH		PAR
					(Frequency)	(Intensity)	
Activity		.40**			.47*		
Adaptability	.47***						
Approach		−.34*					
Mood	.47***				.56***	.48*	
Intensity	.40*						
Distractibility							
Persistence		−.48***			.54***	.47*	
Threshold							
Predictability					.44*	.44*	
BAMF	.40**			−.84****			.76***
VCOPS							
Head:height	−.45*	−.73****		.82****			−.50*
Chest:height		−.78****	−.41*	.81****			−.71***
Manual muscle testing		.76****		−.63***			
Fractures (12 mo)	−.35*	−.38*					
Severity		−.60****	−.43*	.59***			

* $P = .05$; ** $P = .02$; *** $P = .01$; **** $P = .001$.

activity and motor skill were negatively associated with head to body length ratios.

Children's independence in their daily activities (CHAQ) correlated with higher motor performance (Table 3), but demonstrated no significant relationship to temperament. Temperamental activity did not correlate with our measures of upright activity (PAR), biological severity factors, clinical mediators, maternal impression of activity, or the ratio of chest circumference to body length. However, the BAMF, CHAQ, and PAR were strongly related to each other, and inversely related to ratios of head and chest to body length, and severity factors and clinical mediators. Motor skill is clearly essential for upright positions supporting some self-care activities, and physical constraints appear more influential in the attainment of independence in these activities than temperamental factors.

Parents' reports of both the frequency and intensity of daily hassles were significantly related to several temperament domains: mood, activity, persistence and predictability (Table 3). In contrast, we found no significant relationships between parents' reported hassles and measures of physical performance, or disease severity. It was not unexpected to find that certain qualities of a child's behavior, such as generally negative mood and high intensity of expression, would influence the parents' perception of hassles. It is surprising, however, that the severity of the child's disease and motor performance were not significantly related to this measure of parental coping. It appears that the child's behavioral style has a greater influence on parents' perception of stresses than do disease-related variables.

As temperament has been shown to correlate with physical performance outcomes as well as parents' reports of daily hassles, it is important to consider the influence a child's temperament may have on interaction with others. Children with OI often need help from parents and therapists to complete daily activities and expand their motor capabilities. Because specific temperamental attributes, such as mood and intensity, are associated with increased parental hassles, it is not unreasonable that this may generalize to health care providers. Positive or negative aspects of the child's temperament may shape the provider's assessment of the child and, in turn, influence the energy or enthusiasm that individual brings to the relationship. Thus, it is important for the provider to recognize the child's behavioral style, and its implications for the child's interactions not only with parents, but also with members of the health care team.

CONCLUSION

Our objectives were to determine whether children with OI differ from their nondisabled peers with respect to temperament and if specific domains of temperament are related to motor performance. The data suggest that children with OI differ only to the extent that they are relatively hypoactive, and the temperament domains of activity, persistence, and first reaction (approach/withdrawal) may be particularly important for promoting motor achievement.

Logically, a child who is persistent and displays approach behaviors toward unfamiliar persons (such as physicians and therapists) might more easily accept interactions and benefit from therapeutic interventions. To allow all children the opportunity for optimal benefit, particular attention should be paid to maximizing these traits. This study supports the recommendation that considering all aspects of a child's temperamental style in the development of a rehabilitation plan may enhance the child's ability to benefit from interventions to improve motor skill and functional independence.

REFERENCES

1. Willing MC, Deschenes SP, Slayton RL, Roberts EJ. Premature chain termination is a unifying mechanism for *COL1A1* null alleles in osteogenesis imperfecta type I cell strains. *Am J Hum Genet.* 1996;59:799–809
2. Kuivaniemi H, Tromp G, Prockop DJ. Mutations in collagen genes: causes of rare and some common diseases in humans. *FASEB J.* 1991; 5:2052–2060
3. Marini JC, Gerber NL. Osteogenesis imperfecta: rehabilitation and prospects for gene therapy. *JAMA.* 1997;277:746–750
4. Engel J, Prockop DJ. The zipper-like folding of the collagen triple helices and the effects of mutations that disrupt the zipper. *Annu Rev Biophy Biophy Chem.* 1991;20:137–152
5. Thomas A, Chess S. *Temperament and Development.* New York, NY: Brunner/Mazel; 1977
6. Thomas A, Chess S, Birch HG, Hertzig ME, Korn S. *Behavioral Individuality in Early Childhood.* New York, NY: New York Press; 1963
7. Wilson RS, Matheny AP Jr. Behavioral-genetics research in infant temperament: the Louisville Twin Study. In: Plomin R, Dunn J, eds. *The Study of Temperament: Changes, Continuities, and Challenges.* Hillsdale, NJ: Lawrence Erlbaum Associates; 1986:81–97
8. Robinson JL, Kagan J, Reznick JS, Corley R. The heritability of inhibited and uninhibited behavior: a twin study. *Dev Psychol.* 1992;28:1030–1037
9. McNeil TF, Persson-Blennow I. Stability of temperament characteristics in childhood. *Am J Orthopsychiatry.* 1988;58:622–626
10. Pedlow R, Sanson A, Prior M, Oberklaid F. Stability of maternally reported temperament from infancy to 8 years. *Dev Psychol.* 1993;29: 998–1007
11. Maziade M, Côté R, Bernier H, Boutin P, Thivierge J. Significance of extreme temperament in infancy for clinical status in pre-school years II: patterns of temperament change and implications for the appearance of disorders. *Br J Psychiatry.* 1989;154:544–551
12. Guerin DW, Gottfried AW. Developmental stability and change in parent reports of temperament: a ten-year longitudinal investigation from infancy through preadolescence. *Merrill Palmer Q.* 1994;40:334–55
13. Thomas A, Chess S. The New York Longitudinal Study: from infancy to adult life. In: Plomin R, Dunn J, eds. *The Study of Temperament: Changes, Continuities, and Challenges.* Hillsdale, NJ: Lawrence Erlbaum Associates; 1986:39–52
14. Mufson L, Fendrich M, Warner V. The stability of temperament by child and mother reports over two years. *J Am Acad Child Adolesc Psychiatry.* 1990;29:386–391
15. Bridges LJ, Palmer SA, Morales M, Hurtado M, Tsai D. Agreement between affectively based observational and parent-report measures of temperament at infant age 6 months. *Infant Behav Dev.* 1993;16:501–506
16. Matheny AP, Wilson RS, Nuss SM. Toddler temperament: stability across settings and over ages. *Child Dev.* 1984;55:1200–1211
17. Wilson RS, Matheny AP. Assessment of temperament in infant twins. *Dev Psychol.* 1983;19:172–183
18. Wolk S, Zeanah CH, Garcia Coll CT, Carr S. Factors affecting parents' perceptions of temperament in early infancy. *Am J Orthopsychiatry.* 1992;62:71–82
19. Schraeder BD. Children with disabilities. *J Pediatr Nurs.* 1995;10:166–172
20. Carey WB, McDevitt SC. *Coping With Children's Temperament: A Guide for Professionals.* New York, NY: BasicBooks; 1995
21. Lavigne JV, Nolan D, McLone DG. Temperament, coping, and psychological adjustment in young children with myelomeningocele. *J Pediatr Psychol.* 1988;13:363–378

22. McDevitt SC, Carey WB. *Behavioral Style Questionnaire.* Scottsdale, AZ: Behavioral-Developmental Initiatives; 1995

23. Hegvik RL, McDevitt SC, Carey WB. *Middle Childhood Temperament Questionnaire.* Scottsdale, AZ: Behavioral-Developmental Initiatives; 1995

24. Rovet JF, Ehrlich RM. Effect of temperament on metabolic control in children with diabetes mellitus. *Diabetes Care.* 1988;11:77–82

25. Garrison WT, Biggs D, Williams K. Temperament characteristics and clinical outcomes in young children with diabetes mellitus. *J Child Psychol Psychiatry.* 1990;31:1079–1088

26. Lerner RM, Palermo M, Spiro A III, Nesselroade JR. Assessing the dimensions of temperamental individuality across the life-span: the Dimensions of Temperament Survey (DOTS). *Child Dev.* 1982;53:149–59

27. Arnold GL, Kramer BM, Kirby RS, et al. Factors affecting cognitive, motor, behavioral and executive functioning in children with phenylketonuria. *Acta Paediatr.* 1988;87:565–570

28. Fullard W, McDevitt SC, Carey WB. *Toddler Temperament Scale.* Scottsdale, AZ: Behavioral-Developmental Initiatives; 1995

29. Johnson DB, Cheney C Monsen ER. Nutrition and feeding in infants with bronchopulmonary dysplasia after initial hospital discharge: risk factors for growth failure. *J Am Diet Assoc.* 1998;98:649–656

30. Medoff-Cooper B, Carey WB, McDevitt SC. *Early Infancy Temperament Questionnaire.* Scottsdale, AZ: Behavioral-Developmental Initiatives; 1995

31. Carey WB, McDevitt SC. *Revised Infant Temperament Questionnaire.* Scottsdale, AZ: Behavioral-Developmental Initiatives; 1995

32. Green M, Solnit AJ. Reactions to the threatened loss of a child: a vulnerable child syndrome. *Pediatrics.* 1964;34:58–66

33. Perrin EC, West PD, Culley BS. Is my child normal yet? Correlates of vulnerability. *Pediatrics.* 1989;83:355–363

34. Thomasgard M, Shonkoff JP, Metz WP, Edelbrock C. Parent-child relationship disorders. Part II. The vulnerable child syndrome and its relation to parental overprotection. *J Dev Behav Pediatr.* 1995;16:251–256

35. Thomasgard M. Parental perceptions of child vulnerability, overprotection, and parental psychological characteristics. *Child Psychiatry Human Dev.* 1998;28:223–240

36. Wright L, Mullen T, West K, Wyatt P. The VCOP Scale: a measure of overprotection in parents of physically vulnerable children. *J Clin Psychol.* 1993;49:790–798

37. Thomasgard M, Metz WP, Edelbrock C, Shonkoff JP. Parent-child relationship disorders. Part I. Parental overprotection and the development of the Parent Protection Scale. *J Dev Behav Pediatr.* 1995;16:244–250

38. Cintas H, Gerber L, Danoff J, Strickland D. Perception of competence in walking and nonwalking children with osteogenesis imperfecta. *Phys Ther.* 1998;10:180–181

39. Cintas HL, Siegel KL, Furst GP, Gerber LH. Brief assessment of motor function: reliability and validity of the Gross Motor Scale. *Am J Phys Med Rehabil.* 2003;82:1–9

40. Singh G, Athreya BH, Fries JF, Goldsmith DP. Measurement of health-status in children with juvenile rheumatoid arthritis. *Arthritis Rheum.* 1994;1761–1769

41. Crnic KA, Greenberg MT. Minor parenting stresses with young children. *Child Dev.* 1990;61:1628–1637

42. Kendall FP, McCreary EK, Provance PG. *Muscles: Testing and Function.* Baltimore, MD: Williams & Wilkins; 1993

43. Gerber LH, Cintas HL, Furst GP. *Pediatric Activity Record.* Developmental Version 1.2. Bethesda, MD: Rehabilitation Medicine Department

Temperament and Physical Performance in Children With Osteogenesis Imperfecta

Stacy J. Suskauer, Holly L. Cintas, Joan C. Marini and Lynn H. Gerber

Updated Information & Services	including high resolution figures, can be found at: http://pediatrics.aappublications.org/content/111/2/e153
References	This article cites 30 articles, 4 of which you can access for free at: http://pediatrics.aappublications.org/content/111/2/e153#BIBL
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): **Rheumatology/Musculoskeletal Disorders** http://www.aappublications.org/cgi/collection/rheumatology:musculo skeletal_disorders_sub
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.aappublications.org/site/misc/Permissions.xhtml
Reprints	Information about ordering reprints can be found online: http://www.aappublications.org/site/misc/reprints.xhtml

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN®

PEDIATRICS®

Temperament and Physical Performance in Children With Osteogenesis Imperfecta

Stacy J. Suskauer, Holly L. Cintas, Joan C. Marini and Lynn H. Gerber

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://pediatrics.aappublications.org/content/111/2/e153

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN®