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Psychogenetics of Turner syndrome: an investigation of 28 subjects and respective controls using the Bender test and Piagetian scales

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ABSTRACT. Piagetian scales and the Bender visual motor gestalt test (BT) were applied to 28 subjects with universal 45,X Turner syndrome (TS), and their respective controls, in order to investigate their cognitive performance. Dermatoglyphics were also analyzed to obtain clues concerning embryological changes that may have appeared during development of the nervous system and could be associated with cognitive performance of TS patients. Dermatoglyphic pattern distribution was similar to that reported in previous studies of TS individuals: ulnar loops in the digital patterns and finger ridge, a-b, and A'-d counts were more frequent, while arch and whorl patterns were less frequent compared to controls. However, we did not find higher frequencies of hypothenar pattern, maximum atd angle, and ulnarity index in our TS subjects, unlike other investigations. Furthermore, we found significant differences between TS and control T line index values. The BT scores were also lower in probands, as has been previously reported, revealing a neurocognitive deficit of visual motor perception in TS individuals, which could be due to an absence of, or deficiency in, cerebral hemispheric lateralization. However, TS subjects seemed to improve their performance on BT with age. Cognitive performance of the TS subjects was not significantly different from that of controls, confirming a previous study in which TS performance was found to be similar to that of the normal Brazilian population. There were significant correlations between BT scores and Piagetian scale levels with dermatoglyphic parameters. This association could be explained by changes in the common ectodermal origin of the epidermis and the central nervous system. TS subjects seem to succeed in compensating their spatial impairments in adapting their cognitive and social contacts. We concluded that genetic counseling should consider cognitive and psychosocial difficulties presented by TS subjects, providing appropriate treatment and orientation for them and their families.

Key words: Turner syndrome cognition; Bender test; Piagetian scales; Dermatoglyphics

INTRODUCTION

Turner syndrome (TS) is phenotypically characterized by short stature, gonadal dysgenesis and a consequent pubertal development delay, along with webbed neck, lymphedema, and other physical anomalies (shortened ulna, shield chest, multiple pigmented naevi, widely spaced nipples, and dysplastic nails), as well as cardiovascular, renal and liver abnormalities. This pathognomonic constellation seems to be specifically associated with X monosomy, an abnormal X as a second sex chromosome, various types of correlated X chromosomal aberrations and their corresponding mosaicisms.

Turner syndrome affects 1 in 2500 live female births (Elsheikh et al., 2002) and presents a neurocognitive profile of normal global intelligence, except in the case of an X gene dosage compensation disturbance (e.g., X ring chromosome). Nevertheless, TS individuals presenting mathematical, attention and motor coordination difficulties have been reported. Spatial impairment in TS subjects has been widely described, although the exact nature of such a deficit and its consistency and extent are still not completely understood.

In order to investigate TS psychogenetics, we used the Bender visual motor gestalt test (BT) and Piagetian scales (PS), which evaluate the construction of reflective abstraction and representation space. The latter apparently have not been applied to TS subjects before.

Piagetian scales make use of the clinical method (Vinh-Bang, 1970), which guarantees an affective-emotional optimization in order to promote the best child intellectual performance and to more accurately evaluate their individual cognitive potential.

MATERIAL AND METHODS

Finger and palm prints were taken from both hands according to the technique described by Saldanha (1968), and PS and BT were administered to 28 individuals with TS (45,X) and their respective controls, matched for age, race and socioeconomic status. Three age groups were defined: age group 1 (5 to 7), age group 2 (8 to 11) and age group 3 (12 to 15). Qualitative and quantitative dermatoglyphic variables were analyzed according to Saldanha (1968), Otto (1975), and Schaumann and Alter (1976).

The BT was administered according to Clawson (1980) (Figure 1). The examiner registered each subject's visual motor performance, which was further analyzed with the Koppitz method by psychologists specialized in BT. Piagetian scales consisted of the construction of reflective abstraction (Piaget, 1995) and of representation space (Piaget and Inhelder, 1993) tasks. The cognitive performance diagnoses were determined by two professionals with Ph.D. in education, specialists in developmental psychology supervision.

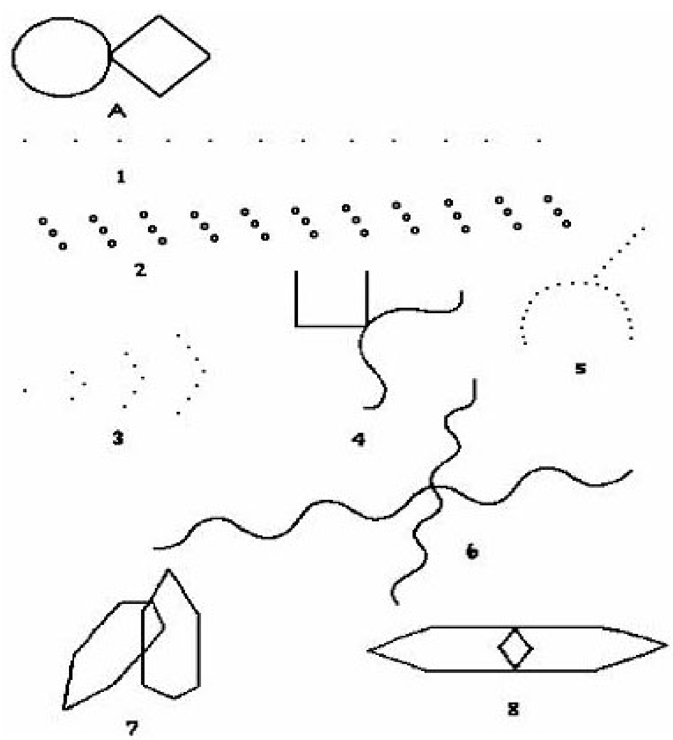

Figure 1. Stimulus-models from the Bender visual motor gestalt test used on the present investigation according to Clawson (1980).

Dermatoglyphic digital data of subjects and controls were compared using the Fisher exact test, and quantitative variables were compared by means of *t*- and paired *t*-tests. The Mann-Whitney test was used to compare BT and PS results of patient and control age groups.

Finally, absolute or transformed values of different variables were submitted to a stepwise multiple regression analysis for both BT and PS data as dependent and independent (intervening factors) variables to determine how they were associated with psychomotor development (Tables 8 and 9). The alpha level for significance was 5% (Tables 5 to 9).

RESULTS

Bender test

Only one in 28 patients and five of 28 controls presented either good visual motor ability or good perception maturity in their BT performance (Table 1). The remaining individuals displayed visual motor perception levels below their chronological age. TS girls had a mean of 11.6 brain lesion indicators, while controls had only 5.6. Figure 2 shows rotation and perseveration data from patient BT performances.

Piagetian scales

Reflective abstraction tasks - *Correlate formation task*

Data referring to performance by subjects and controls on the correlate formation and other Piagetian tasks are given in Tables 2 and 3, respectively. Three patients (N = 28) and four controls (N = 28) displayed Pre-Operatory Stage 1 performance at the initial diagnosis. Thus, for correlate formation task, those subjects (all belonging to the age group 1) did not succeed in organizing the matrixes; they could not establish the corresponding correlate of each object represented in the figures and could not maintain stable the pairs that they had previously organized. Remarkably, however, a greater number of control girls presented Pre-Operatory Stage 1 performance for that task when compared to TS girls.

Neither experimental nor control subjects presented transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level performances. Four of 28 propositi and three of 28 controls did not succeed in establishing correlates and could not maintain stable pairs, although they had built them regularly, based on the "it is for" idea, which corresponds to Pre-Operatory Equilibrium Level performance in this task (Tables 2 and 3).

It is noteworthy that a TS subject (No. 15), whose correlate formation task diagnosis was Pre-Operatory Equilibrium Level, revealed a better performance in correlate formation tasks than the respective control (Pre-Operatory Stage 1).

Transition between the performance of pre-operatory and concrete operatory periods was shown by six controls and 11 patients, mostly belonging to age groups 2 and 3. Thus, the latter TS subjects and their respective controls maintained stable pairs, although without organizing matrixes properly, which means they could not establish the corresponding correlate.

Table 1. Performances of subjects and controls on Bender test and respective ages (A) and significative (+) or not significative (-) number of brain lesion (BL) indicators.

Subjects	Age	Score	Visual motor perception level	BL	BL#	BL##	Controls	Age	Score	Visual motor perception level	BL	BL#	BL##
1	(10;11)	6	6 ½ and 6 years and 11 months	9	+	-	1	(10;11)	7	6 ½ and 6 years and 11 months	8	+	-
2	(14;02)	10	5 ½ and 5 years and 11 months	10	+	+	2	(14;02)	10	5 ½ and 5 years and 11 months	10	+	+
3	(08;11)	9	6 and 6 years and 5 months	7	-	-	3	(08;08)	8	6 and 6 years and 5 months	9	+	-
4	(11;05)	8	6 and 6 years and 5 months	7	-	-	4	(11;07)	3	8 ½ and 8 years 11 months	4	-	-
5	(13;01)	12	5 and 5 years and 5 months	13	+	+	5	(12;10)	0	This person has got enough perception conditions for a good visual motor ability	0	-	-
6	(08;06)	10	5 ½ and 5 years and 11 months	11	+	+	6	(08;07)	7	6 ½ and 6 years and 11 months	7	-	-
7	(11;05)	11	5 ½ and 5 years and 11 months	11	+	+	7	(11;00)	9	6 and 6 years and 5 months	9	+	-
8	(12;03)	6	6 ½ and 6 years and 11 months	7	-	-	8	(12;08)	6	6 ½ and 6 years and 11 months	7	-	-
9	(15;03)	9	6 and 6 years and 5 months	9	+	-	9	(15;03)	0	Good perception maturity	1	-	-
10	(12;01)	16	Below 5 years	17	+	+	10	(12;07)	5	7 and 7 years and 5 months	5	-	-
11	(07;08)	19	Below 5 years	18	+	+	11	(08;01)	13	5 and 5 years and 5 months	9	+	-
12	(13;06)	14	Below 5 years	17	+	+	12	(13;07)	4	8 and 8 years 5 months	4	-	-
13	(10;01)	13	5 and 5 years and 5 months	14	+	+	13	(10;04)	1	Good perception maturity	1	-	-
14	(14;02)	1	Good perception maturity	0	-	-	14	(13;10)	3	8 and 8 years 5 months	4	-	-
15	(06;07)	18	Below 5 years	14	+	+	15	(06;11)	11	5 ½ and 5 years and 11 months	10	+	+
16	(14;00)	4	8 and 8 years 5 months	4	-	-	16	(14;03)	3	8 ½ and 8 years and 11 months	4	-	-
17	(14;03)	8	6 and 6 years and 5 months	10	+	+	17	(14;07)	4	8 and 8 years 5 months	4	-	-
18	(06;05)	16	Below 5 years	12	+	+	18	(06;11)	11	5 ½ and 5 years and 11 months	5	-	-
19	(15;03)	4	8 and 8 years 5 months	8	+	-	19	(15;00)	2	Good perception maturity	2	-	-
20	(08;09)	17	Below 5 years	14	+	+	20	(08;09)	6	6 ½ and 6 years and 11 months	6	-	-
21	(06;05)	20	Below 5 years	19	+	+	21	(06;10)	6	6 ½ and 6 years and 11 months	5	-	-
22	(08;06)	11	5 ½ and 5 years and 11 months	12	+	+	22	(08;11)	3	Good perception maturity	4	-	-
23	(09;09)	12	5 and 5 years and 5 months	14	+	+	23	(09;04)	5	7 and 7 years and 5 months	6	-	-
24	(06;01)	22	Below 5 years	17	+	+	24	(05;08)	18	Below 5 years	11	+	+
25	(06;03)	18	Below 5 years	13	+	+	25	(06;01)	16	Below 5 years	12	+	+
26	(10;06)	8	6 and 6 years and 5 months	9	+	-	26	(10;10)	6	6 ½ and 6 years and 11 months	7	-	-
27	(06;04)	21	Below 5 years	19	+	+	27	(06;02)	10	5 ½ and 5 years and 11 months	5	-	-
28	(08;10)	9	6 and 6 years and 5 months	9	+	-	28	(09;01)	6	6 ½ and 6 years and 11 months	6	-	-

Age = years; months; #UnG (Guarulhos University) standard: $n \geq 8$; ##APEP (Psychology and Psychotherapy Studies Association) standard: $n \geq 10$.

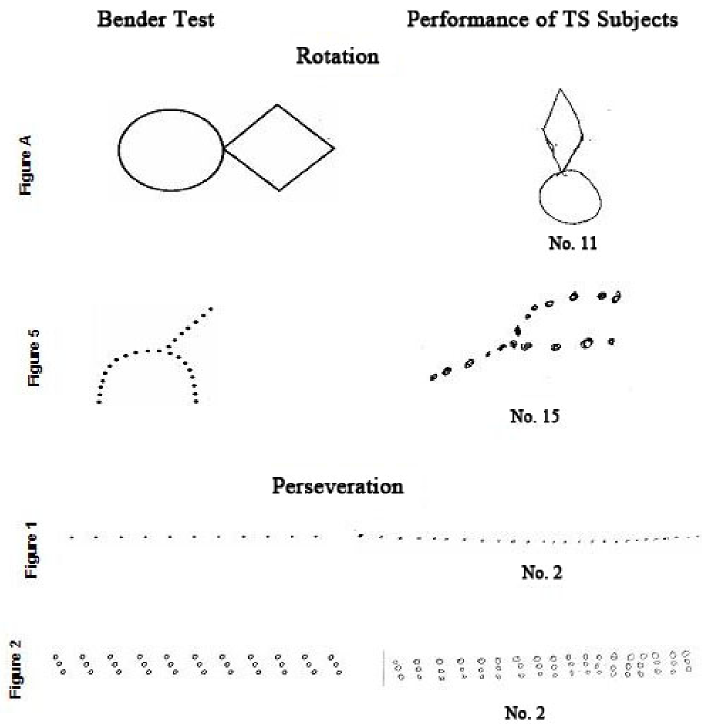

Figure 2. Graphic results of Bender visual motor gestalt test performances from some Turner syndrome (TS) subjects presenting rotation and perseveration examples.

Three of the TS probands and three times as many controls, all belonging to age groups 2 and 3, displayed performance corresponding to the beginning of the concrete operatory period in the correlate formation task; as these subjects succeeded in organizing the matrixes, building the correlates and maintaining stable pairs, however, without excluding counterexamples.

Transition between the beginning of the concrete operatory period and the equilibrium of the same cognitive period was observed in the performance of four TS subjects, all belonging to the age group 3 and of three subjects of the control group (two, age group 2; one, age group 3). Thus, all subjects succeeded in building the matrixes, establishing each respective correlate and maintaining stable pairs; though the counterexamples were excluded, these results did not justify a transition from the beginning to equilibrium of the concrete operatory period behavior.

Typical performance in the concrete operatory period in equilibrium (concrete operatory equilibrium level) was presented by three (two, age group 2; one, age group 3) of the experimental subjects, and by two (age groups 2 and 3) of the controls. Thus, the former as well as the latter subjects succeeded in building the matrixes, since they organized the correlates, maintaining stable pairs and justifying exclusion of the counterexamples.

Moreover, no subject from either experimental or control groups presented performance corresponding to transition between concrete operatory and formal operatory periods. Among controls, only one subject exhibited performance equivalent to the beginning stage of the formal period. Such a situation indicated that this subject (age group 3) had already fully built the necessary mental structures in order to have success in the correlate formation task. This means that she established the correlates systematically and perceived their respective analogies with proportions (Tables 2 and 3).

Relationships between surfaces and perimeters of rectangles task

Seven subjects from the experimental group (N = 28) and also seven (N = 28) controls presented global correspondence between surfaces and perimeters, since they had admitted either simultaneous increasing or diminishing of both, or their invariable permanence. Thus, those subjects (TS or not; mainly from the age group 1) showed a Pre-Operatory Stage 1 performance in the relationships between surfaces and perimeters of rectangles task (Tables 2 and 3).

Also two (age groups 1 and 2) of the TS subjects exhibited performance corresponding to transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level, because they presented solutions referring to those of Pre-Operatory Equilibrium Level. This result was not observed for any of the controls.

On the other hand, six of 28 control subjects and eight of 28 experimental subjects, aged between six and 15 years, showed performance equivalent to Pre-Operatory Equilibrium Level; they had hesitated between increases and decreases of surfaces and perimeters and also had not admitted the conservation of both measures, as with test A (boards with nails and string) and B (small wooden rectangles; Piaget, 1995). Moreover, when comparisons were requested, they asserted that those dimensions varied in the same way for one test situation as with the other, both varied in opposite directions; for instance, surface and perimeter could increase together after each transformation for test A and diminish simultaneously for test B.

Five patients and four controls belonging to age groups 2 and 3 presented performances diagnosable as transition between pre-operatory and concrete operatory periods (Tables 2 and 3); they admitted conservation of perimeter on test A and of surface on test B, after counter-argumentation. However, they changed their responses by hesitating between increases and decreases of the stimuli. This occurred mainly when summing the transformations observed in each situation. When comparing both tests, they also established compensations between the perceived dimensions, although in an inconsistent manner.

Only one subject from the experimental group (age group 2) presented behavior corresponding to the beginning of the concrete operatory period. On the other hand, five controls from age groups 2 and 3 presented this same performance (Tables 2 and 3). In this way, these subjects established relationships of compensation between perceived dimensions and were able to produce the correct summaries for both tests (A and B) but did not succeed in comparing them properly. Based on these coordinated responses, they formulated a commutability (or commutative) law, according to which what is summed up to a point (in this case, length) corresponds, quantitatively, to what is subtracted from another (in this case, width); this means that these subjects admitted compensation that was in fact inexistent, between dimensions, concerning relationships between surfaces and perimeters of the rectangles task.

No experimental subjects presented behavior corresponding to transition between the beginning and the equilibrium of the concrete operatory period. On the other hand, three controls (age groups 2 and 3) established the same relationships of compensation that characterize the previous period, but also accomplished correct summaries for A and B and compared both games properly (Tables 2 and 3).

Table 2. Performances of Turner syndrome subjects classified by Piagetian scales (PS) and respective ages (A), age groups (AG) and final diagnoses (FD).

Subjects	A (years; months)	AG	CF		SP		OPA		PSL		3M		Total	FD
			Diagnosis	Points	Diagnosis	Points	Diagnosis	Points	Diagnosis	Points	Diagnosis	Points		
1	(10;11)	2	CO2	4.0	CO2	4.0	CO1	3.0	CO1	3.0	tCO1/2	3.5	17.5	tCO1/2
2	(14;02)	3	tCO1/2	3.5	PO2	2.0	tPO/CO	2.5	CO1	3.0	CO1	3.0	14.0	tPO/CO
3	(08;11)	2	tPO/CO	2.5	tPO1/2	1.5	tPO/CO	2.5	CO1	3.0	tPO/CO	2.5	12.0	tPO/CO
4	(11;05)	2	tPO/CO	2.5	tPO/CO	2.5	CO1	3.0	tPO/CO	2.5	CO1	3.0	13.5	tPO/CO
5	(13;01)	3	tPO/CO	2.5	tPO/CO	2.5	tPO/CO	2.5	tPO/CO	2.5	CO1	3.0	13.0	tPO/CO
6	(08;06)	2	tPO/CO	2.5	CO1	3.0	tPO/CO	2.5	tPO/CO	2.5	CO1	3.0	13.5	tPO/CO
7	(11;05)	2	tPO/CO	2.5	tPO/CO	2.5	PO2	2.0	CO1	3.0	tCO1/2	3.5	13.5	tPO/CO
8	(12;03)	3	tCO1/2	3.5	CO2	4.0	tPO/CO	2.5	CO1	3.0	CO1	3.0	16.0	tCO1/2
9	(15;03)	3	tCO1/2	3.5	CO2	4.0	tPO/CO	2.5	CO1	3.0	tCO1/2	3.5	16.5	tCO1/2
10	(12;01)	3	CO1	3.0	PO1	1.0	tPO/CO	2.5	CO1	3.0	tPO/CO	2.5	12.0	tPO/CO
11	(07;08)	1	PO2	2.0	tPO1/2	1.5	PO1	1.0	PO1	1.0	tPO/CO	2.5	8.0	tPO1/2
12	(13;06)	3	tPO/CO	2.5	PO2	2.0	tPO/CO	2.5	CO1	3.0	tPO/CO	2.5	12.5	tPO/CO
13	(10;01)	2	tPO/CO	2.5	PO1	1.0	PO2	2.0	tPO/CO	2.5	CO1	3.0	11.0	tPO/CO
14	(14;02)	3	CO2	4.0	tCO/FO	4.5	tCO1/2	3.5	tPO/CO	2.5	tCO1/2	3.5	18.0	tCO1/2
15	(06;07)	1	PO2	2.0	PO2	2.0	PO2	2.0	CO1	3.0	tPO/CO	2.5	11.5	tPO/CO
16	(14;00)	3	tCO1/2	3.5	CO2	4.0	tCO1/2	3.5	CO1	3.0	CO2	4.0	18.0	tCO1/2
17	(14;03)	3	PO2	2.0	PO2	2.0	tPO/CO	2.5	CO1	3.0	CO1	3.0	12.5	tPO/CO
18	(06;05)	1	PO1	1.0	PO1	1.0	PO1	1.0	tPO/CO	2.5	CO1	3.0	8.5	tPO1/2
19	(15;03)	3	tPO/CO	2.5	tPO/CO	2.5	CO2	4.0	CO1	3.0	CO1	3.0	15.0	CO1
20	(08;09)	2	CO2	4.0	PO2	2.0	tPO/CO	2.5	tPO/CO	2.5	tPO/CO	2.5	13.5	tPO/CO
21	(06;05)	1	PO2	2.0	PO1	1.0	PO1	1.0	PO2	2.0	CO1	3.0	9.0	tPO1/2
22	(08;06)	2	tPO/CO	2.5	PO2	2.0	PO2	2.0	tPO/CO	2.5	CO1	3.0	12.0	tPO/CO
23	(09;09)	2	tPO/CO	2.5	tPO/CO	2.5	tPO/CO	2.5	CO1	3.0	tCO1/2	3.5	14.0	tPO/CO
24	(06;01)	1	PO1	1.0	PO1	1.0	PO1	1.0	PO1	1.0	PO1	1.0	5.0	PO1
25	(06;03)	1	tPO/CO	2.5	PO1	1.0	PO1	1.0	PO2	2.0	CO1	3.0	9.5	tPO1/2
26	(10;06)	2	CO1	3.0	PO2	2.0	tPO/CO	2.5	tPO/CO	2.5	tCO1/2	3.5	13.5	tPO/CO
27	(06;04)	1	PO1	1.0	PO1	1.0	PO1	1.0	PO1	1.0	PO1	1.0	5.0	PO1
28	(08;10)	2	CO1	3.0	PO2	2.0	tPO/CO	2.5	PO2	2.0	CO1	3.0	12.5	tPO/CO

PS tasks: CF = correlate formation; SP = surfaces and perimeters; OPA = order of practical actions; PSL = projective straight line; 3M = "three mountains". For abbreviations of diagnosis, see legend to Table 4.

Table 3. Performances of controls classified by Piagetian scales (PS) and respective ages (A), age groups (AG) and final diagnoses (FD).

Controls	A (years; months)	AG	CF		SP		OPA		PSL		3M		Total	FD
			Diagnosis	Points	Diagnosis	Points	Diagnosis	Points	Diagnosis	Points	Diagnosis	Points		
1	(10;11)	2	CO1	3.0	CO1	3.0	CO1	3.0	CO1	3.0	CO1	3.0	15.0	CO1
2	(14;02)	3	tPO/CO	2.5	tCO1/2	3.5	tPO/CO	2.5	CO1	3.0	CO1	3.0	14.5	tPO/CO
3	(08;08)	2	tPO/CO	2.5	CO1	3.0	tPO/CO	2.5	CO1	3.0	tCO1/2	3.5	14.5	tPO/CO
4	(11;07)	2	tCO1/2	3.5	CO2	4.0	CO1	3.0	CO1	3.0	OC2	4.0	17.5	tCO1/2
5	(12;10)	3	tCO1/2	3.5	CO2	4.0	CO2	4.0	CO1	3.0	OC2	4.0	18.5	tCO1/2
6	(08;07)	2	tCO1/2	3.5	PO2	2.0	tPO/CO	2.5	CO1	3.0	CO1	3.0	14.0	tPO/CO
7	(11;00)	2	PO2	2.0	tPO/CO	2.5	tPO/CO	2.5	CO1	3.0	tCO1/2	3.5	13.5	tPO/CO
8	(12;08)	3	CO1	3.0	PO2	2.0	CO1	3.0	CO1	3.0	tCO1/2	3.5	14.5	tPO/CO
9	(15;03)	3	FO1	5.0	FO1	5.0	FO1	5.0	CO1	3.0	CO2	4.0	22.0	FO1
10	(12;07)	3	PO2	2.0	PO2	2.0	CO1	3.0	CO1	3.0	tCO1/2	3.5	13.5	tPO/CO
11	(08;01)	1	PO2	2.0	PO1	1.0	tPO/CO	2.5	CO1	3.0	CO1	3.0	11.5	tPO/CO
12	(13;07)	3	CO2	4.0	tPO/CO	2.5	CO2	4.0	CO1	3.0	tPO/CO	2.5	16.0	tCO1/2
13	(10;04)	2	CO2	4.0	tCO1/2	3.5	CO1	3.0	tPO/CO	2.5	tCO1/2	3.5	16.5	tCO1/2
14	(13;10)	3	CO1	3.0	CO1	3.0	CO1	3.0	CO1	3.0	tCO1/2	3.5	15.5	tCO1/2
15	(06;11)	1	PO1	1.0	PO1	1.0	PO1	1.0	PO1	1.0	tPO/CO	2.5	6.5	tPO1/2
16	(14;03)	3	CO1	3.0	tCO1/2	3.5	CO2	4.0	CO1	3.0	CO1	3.0	16.5	tCO1/2
17	(14;07)	3	CO1	3.0	CO1	3.0	tPO/CO	2.5	CO1	3.0	tCO1/2	3.5	15.0	CO1
18	(06;11)	1	tPO/CO	2.5	PO1	1.0	PO1	1.0	PO2	2.0	tCO1/2	3.5	10.0	PO2
19	(15;00)	3	tPO/CO	2.5	tPO/CO	2.5	CO1	3.0	CO1	3.0	CO1	3.0	14.0	tPO/CO
20	(08;09)	2	tPO/CO	2.5	PO1	1.0	PO1	1.0	tPO1/2	1.5	CO1	3.0	9.0	tPO1/2
21	(06;10)	1	CO1	3.0	PO2	2.0	tPO1/2	1.5	tPO/CO	2.5	CO1	3.0	12.0	tPO/CO
22	(08;11)	2	CO1	3.0	tPO/CO	2.5	tPO/CO	2.5	CO1	3.0	CO1	3.0	14.0	tPO/CO
23	(09;04)	2	tPO/CO	2.5	PO2	2.0	tPO/CO	2.5	tPO/CO	2.5	tPO/CO	2.5	12.0	tPO/CO
24	(05;08)	1	PO1	1.0	PO1	1.0	PO1	1.0	tPO1/2	1.5	PO2	2.0	6.5	tPO1/2
25	(06;01)	1	PO1	1.0	PO1	1.0	tPO1/2	1.5	tPO1/2	1.5	tPO/CO	2.5	7.5	tPO1/2
26	(10;10)	2	CO1	3.0	PO2	2.0	tPO/CO	2.5	CO1	3.0	CO1	3.0	13.5	tPO/CO
27	(06;02)	1	PO1	1.0	PO1	1.0	PO2	2.0	CO1	3.0	tPO/CO	2.5	9.5	tPO1/2
28	(09;01)	2	CO1	3.0	CO1	3.0	tPO/CO	2.5	CO1	3.0	CO1	3.0	14.5	tPO/CO

PS tasks: CF = correlation formation; SP = surfaces and perimeters; OPA = order of practical actions; PSL = projective straight line; 3M = "three mountains". For abbreviations of diagnosis, see legend to Table 4.

Typical performance on concrete operatory equilibrium level was observed for four of the 28 probands (mainly from the age group 3) and two subjects (age groups 2 and 3) from the control group (Tables 2 and 3). Thus, TS subjects and their respective controls presented double conservation, both of perimeter and surface, by explaining the generalized conservation after each transformation by using compensations between diminishing width and increasing the length of geometric figures, except for the rectangle after the third transformation (R_3) on A. These subjects showed correct (or mostly correct) summaries for A and B, but presented clear identity tendency in the transformations, when making comparisons.

Only one subject (age group 3; Table 2) presented solutions typical of transition between concrete operatory and formal operatory periods, because she gave exact answers, showing conservation either of perimeter or of surface, and correct summaries and comparisons, although without understanding the incompatibility between conservations.

On the other hand, the same control who showed the best possible performance in the correlate formation task also presented solutions compatible with the formal operatory period (the beginning stage of the formal period) in the current task, taking into account the impossibility of perimeter and surface to conserve simultaneously during transformations of figures, by giving correct summaries and by establishing exact comparisons (Table 3).

Order of practical actions task

Six (age group 1) patients and four (age groups 1 and 2) controls presented performance equivalent to Pre-Operatory Stage 1 (Tables 2 and 3). Therefore, they did not dissociate form from content; they described the fulfilled actions in the proper order, but neither of them perceived, from that activity, a common form nor generalized it for other contents.

Performances typical of transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level were observed for two (age group 1) controls (Table 3). These subjects admitted, at least once, some similarity between actions performed with the doll and with the tower. No TS subject presented transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level behaviors.

Four (age groups 1 and 2) TS subjects and one (age group 1) control dissociated the common form from the variable contents in all situations; this situation indicated that there were likelihoods in the actions and also that they described actions such as doing-undoing and doing-redoing, but did not admit similarity among different contents. Consequently, their performance was characterized as typical of Pre-Operatory Equilibrium Level (Tables 2 and 3).

Performance equivalent to transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level was the most frequent as well among patients (13; age groups 2 and 3) as among controls (10; including all age groups; Tables 2 and 3). Thus, most subjects in our study described actions performed with objects, even when comparing actions, but not their execution order. They presented *réversibilité*[1] provided that they showed that direct-inverse connection to each pair of activities had priority over direct-direct or inverse-inverse relationships between each other pair. They admitted likelihoods but justified improperly or inverted the order of the relationships.

[1]Returning to the starting point, term to term, from each of the intermediary steps but without intentional inversion. It all about a physical or concrete action.

Typical behavior corresponding to the beginning of the concrete operatory period was observed in two (age group 2) experimental subjects and seven (age groups 2 and 3) controls. These subjects presented reversibility (*réversibilité*[2]) term to term and dissociated form from content. In addition, they showed that, in each pair of activities, direct-inverse connection did not have priority over direct-direct or inverse-inverse relationships between each other pair, for this task (Tables 2 and 3).

Furthermore, another two TS subjects (age group 3) showed typical performance as transition between the beginning and the equilibrium of the concrete operatory period (Table 2) provided that, although they did not explicitly refer to the order and to the comparison of order of actions, it seems they presented *réversibilité*, which characterizes the beginning of the concrete operatory period, besides a greater penetration of arguments, which delineates the concrete operatory equilibrium level. No control showed such a performance.

Also, only one proband (age group 3) presented solutions that could be classified as concrete operatory equilibrium level (Table 2), since she distinguished the kinds of orders and integrated them in only one common form of order by revealing greater penetration of arguments.

Finally, it is also remarkable that this same control individual conducted, in the order of practical actions, a corresponding best possible performance (the beginning stage of the formal period), as she did for other reflective abstraction tasks (correlate formation task and relationships between surfaces and perimeters of the rectangles task; Table 3).

Representation space tasks

Projective straight line construction

Moreover, three experimental subjects (age group 1) and one control subject exhibited performance equivalent to Pre-Operatory Stage 1 for the projective straight line construction task (Tables 2 and 3). Although they recognized a straight line and distinguished it from a curved one, they did not succeed in building a straight line parallel to one side of the table, since they built it along the model or did so with the elements kept too close together. This situation indicates the primacy of the topological straight line.

Typical behaviors of transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level had not been observed in probands; after fumbling, three (age groups 1 and 2) controls achieved the proper solution on at least one of the three steps of the task. Thus, they presented at least one of the typical aspects of the following cognitive level (Pre-Operatory Equilibrium Level), indicating a transition situation (Table 3).

Three experimental subjects and only one control reached the Pre-Operatory Equilibrium Level (Tables 2 and 3). After fumbling, these subjects achieved the proper solution by building a straight line parallel to one side of the table (perceptive straight line). However, they could not do without the table edge to build a straight line in a non-parallel position or in the displayed circle, a situation indicating the beginning of the representative straight line.

[2]The subject infers that there is symmetry between direct and indirect orientations and returns to the starting point by inverting intentionally, term to term, each of the intermediary steps and moreover by being able to describe them, even without performing the action but as a mental representation.

Nine experimental subjects, belonging to all age groups and three (age groups 1 and 2) controls presented performance corresponding to transition between pre-operatory and concrete operatory periods. Thus, these 12 subjects progressively discriminated the adequate viewpoints regarding the straight lines marked on the parallel of the table edge; afterwards, they gradually became free from the perceptive configuration for building the other positions of the straight line. These subjects aligned objects along a sight direction, and they perceived the most adequate viewpoint to be checked; however, they were not able to accurately sight during the task.

In the following level of performance, stimulus objects were aligned one to another relative to one viewpoint, so that the first stimulus hid the following ones; this situation means that subjects could accurately sight. As a result, most of the controls (20 of 28) belonging to all age groups and 13 of 28 probands belonging mainly to age groups 2 and 3 demonstrated performance typical of the beginning of the concrete operatory period (Tables 2 and 3). It is noteworthy that this cognitive level corresponds to the best possible one for this task.

View points or perspectives coordination task (“three mountains” task)

Two propositi did not understand the inquiry, so that the “three mountains” task application was not possible. Consequently, a typical performance of Pre-Operatory Stage 1 was characterized (Table 2). In addition, no control subject presented Pre-Operatory Stage 1 or transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level behaviors. Moreover, no typical performance at the latter level was observed among TS subjects.

Similarly, no experimental subject presented the Pre-Operatory Equilibrium Level typical behavior, which was observed in only one control (Table 3), who did not react to the inquiry and, by performing technique 1, she could not even reproduce what she could see from her own viewpoint.

Characteristic performance of transition between pre-operatory and concrete operatory periods was revealed by six all-age probands and by five (mainly age groups 1 and 2) controls (Tables 2 and 3). These subjects did not understand that distinct observers could see the mountains as a whole from different viewpoints. Furthermore, they reproduced what they could see from their own viewpoint on technique 1, and on technique 2, they chose preferentially the picture that represented their own viewpoint or any other picture. Finally, in technique 3, they placed the doll in their own perspective or in the middle of the mountains.

The beginning of the concrete operatory period was the most frequent performance by TS subjects (13 including all age groups) and by controls (11 including all age groups) (Tables 2 and 3). These subjects could understand the relativity of some inquiries but they indicated them as “false absolutes”. Otherwise, on technique 1, they would have turned each mountain around, without considering relationships between the first and the second planes. In addition, in technique 2, they did not consider left-right and front-back relationships, but they insisted on the presence of the indispensable elements. Also, with technique 3, they sought for another position, but failed by not considering the relationships of the elements to one another.

Another very frequent performance was transition between the beginning and the equilibrium of the concrete operatory period observed among TS individuals (six; age groups 2 and 3) and among controls (eight; mainly from age groups 2 and 3; Tables 2 and 3). These subjects presented true although incomplete relativity, provided that they started to understand which changes in the observer position would cause transformations in the relationships among the mountains. They also discovered left-right, front-back and upward and downward relationships but they did not ever orient correctly from another viewpoint. Moreover, in technique 1, predominance of their own viewpoint would have occurred, especially in left-right relationships; otherwise, in technique 2 they found exactly one relationship and neglected the others; also, in technique 3, they departed from one relationship among the three mountains and between these and the doll, but they centered only one mountain and neglected the others.

Only one proband (age group 3) and three controls (age groups 2 and 3) presented the best possible performance in the "three mountains" task, which is the concrete operatory equilibrium level (Tables 2 and 3). Thus, they presented complete relativity of the perspectives by coordinating all viewpoints. In the first technique, they reconstituted the perspective with the three cards through immediate or progressive coordination by establishing multiplicative relationships. In techniques 2 and 3, these subjects performed a foreseeable scheme and the logical multiplication of the pertinent relationships.

Final operatory diagnosis

Reference values for final operatory diagnoses of experimental and control subjects (Table 4) were obtained by summing individual performance points on each Piagetian task.

Table 4. Reference values for the final operatory diagnosis corresponding to the sum of points of individual performance on each Piagetian task.

Total points	Final diagnosis
1.0 to 5.0	PO1
5.5 to 9.5	tPO1/2
10.0	PO2
10.5 to 14.5	tPO/CO
15.0	CO1
15.5 to 18.5	tCO1/2
19.0	CO2
19.5 to 20.5	tCO/FO
21.0 to 22.0	FO1

PO = pre-operatory period; PO1 = the beginning stage of the pre-operatory period; tPO1/2 = transition between Pre-Operatory Stage 1 and the Pre-Operatory Equilibrium Level; PO2 = Pre-Operatory Equilibrium Level; tPO/CO = transition between pre-operatory and concrete operatory periods; CO = concrete operatory period; CO1 = the beginning stage of the concrete operatory period; tCO1/2 = transition between the beginning stage and the stage in equilibrium of the concrete operatory period; CO2 = concrete operatory equilibrium level; tCO/FO = transition between the concrete operatory and the formal operatory periods; FO1 = the beginning stage of the formal period; FO = formal operatory period.

Final diagnosis in Pre-Operatory Stage 1 was established for two (age group 1) TS subjects and for no controls. The final diagnosis was age compatible for TS with the age pattern of the Brazilian population.

Four (age group 1) experimental subjects and five (belonging mainly to the age group 1) controls were in a transition between Pre-Operatory Stage 1 and Pre-Operatory Equilibrium Level, taking into account their performance on all tasks. This diagnosis is likely compatible with their respective ages, even for the age group 2, taking as a reference normal Brazilian children.

Pre-Operatory Equilibrium Level performance was reached by only one control, a likewise age-compatible final diagnosis. Most TS subjects (16; mainly age groups 2 and 3) and similarly most controls (13; all age groups) obtained a final transition between pre-operatory and concrete operatory periods of the diagnosis.

Amongst subjects classified as transition between pre-operatory and concrete operatory periods in their final diagnosis, seven experimental subjects and four controls exhibited a performance delay. Only one TS subject (age group 3) and two controls (age groups 2 and 3), achieved a performance corresponding to the beginning of the concrete operatory period. Therefore, a cognitive delay was identified in the operatory behavior of one TS subject and one control.

Five probands (mostly age group 3) and six controls (age groups 2 and 3) varied from 15.5 to 18.5 in the sum of points in their performance on all Piagetian tasks. Hence, they achieved transition between the beginning and the equilibrium of the concrete operatory period in their final diagnosis. Moreover, three experimental subjects and one control presented delay in their development, considering the age profile of a similar Brazilian population. Delay of cognition development among patients and controls was characterized as follows: a) subjects from 9 years old exhibiting a final diagnosis of pre-operatory period or lower level; b) subjects from 11 years old displaying the beginning of the concrete operatory period or lower level as a final diagnosis; c) subjects from 14 years old revealing a concrete operatory equilibrium level or a lower level as a final diagnosis (Mantovani de Assis, 1976; Grossman, 1988; Campos, 1989; Zaia, 1996).

In conclusion, control subject number 9 was the only one achieving more than 20.5 in the sum of individual performance points on each Piagetian task. It is remarkable that her behavior on the PS was age-compatible with that of normal young Brazilian people.

Psychodiagnosis: comparison between subjects and controls

Medians and respective statistical significance were compared by age, results of psychodiagnosis tests and tasks in the total, and by age groups 1, 2 and 3, between subjects and controls, using the Mann-Whitney test (Table 5).

In all samples, comparison between patients and controls showed significant differences in BT scores, specifically in distortion, integration and perseveration forms; highly significant in the total score of BT, and extremely significant in rotation and brain lesion indicators.

Regarding age group 1, a significant difference between TS and controls was observed concerning distortion and integration forms, and highly significant referring to rotation and the total score of BT.

Finally, in relation to the age group 3, comparison between TS and controls revealed significant differences in total score and brain lesion indicators, and highly significant only for rotation, also in BT.

Summing up, except for partial or total BT results, the differences in age and psychodiagnosis between patients and controls were not significant. Concerning age, this lack of a significant difference may confirm proper age-sampling methodology. On the other hand, the difference in rotation (age group 1) scores between TS carriers and control individuals gave a borderline value of statistical significance (0.051; Table 5).

Table 5. Median comparisons and respective significance values related to age (A, in months) and to results of psychodiagnosis tests and tasks, considering total individuals and age groups (AG) 1, 2 and 3, between Turner syndrome (TS) subjects and controls (CON), through the Mann-Whitney non-parametric test.

Group (N)	A (months)	Bender test (BT)						Piagetian scales (PS)					
		F	R	I	Pe	Total	BL	CF	SP	OPA	PSL	3M	Total
Total (28)													
Median													
TS	123.50	4.50	5.0	2.50	0.0	11.0	11.50	2.50	2.0	2.50	2.50	3.0	12.75
CON	127.0	3.0	2.0	1.0	0.0	6.0	5.50	3.0	2.50	2.50	3.0	3.0	14.0
Significance	U = 789.50	U = 922.0	U = 1.048.50	U = 939.50	U = 885.50	U = 1.016.0	U = 1.073.50	U = 747.50	U = 738.0	U = 714.50	U = 713.50	U = 729.0	U = 716.0
P	0.896^{ns}	0.041*	<0.0001****	0.018*	0.047*	0.0004***	<0.0001****	0.401^{ns}	0.320^{ns}	0.225^{ns}	0.120^{ns}	0.236^{ns}	0.254^{ns}
AG1 (7)													
Median													
TS	77.0	7.0	7.0	5.0	1.0	19.0	17.0	2.0	1.0	1.0	2.0	2.50	8.50
CON	82.0	5.0	4.0	1.0	0.0	11.0	9.0	1.0	1.0	1.50	2.0	2.50	9.50
Significance	U = 50.50	U = 69.50	U = 68.0	U = 68.50	U = 59.0	U = 74.50	U = 76.50	U = 53.0	U = 55.50	U = 42.50	U = 47.0	U = 48.0	U = 46.0
P	0.847^{ns}	0.026*	0.051^{ns}	0.044*	0.408^{ns}	0.006**	0.002**	1.0^{ns}	0.656^{ns}	0.156^{ns}	0.513^{ns}	0.592^{ns}	0.441^{ns}
AG2 (11)													
Median													
TS	117.0	3.0	4.0	3.0	θ	10.0	11.0	2.50	2.0	2.50	2.50	3.0	13.50
CON	112.0	3.0	2.0	1.0	θ	6.0	6.0	3.0	2.50	2.50	3.0	3.0	14.0
Significance	U = 122.50	U = 148.50	U = 177.50	U = 157.0	θ	U = 177.50	U = 176.0	U = 116.50	U = 110.50	U = 115.0	U = 107.0	U = 121.0	U = 102.50
P	0.818^{ns}	0.149^{ns}	<0.001***	0.044*	θ	<0.001***	0.001**	0.507^{ns}	0.296^{ns}	0.414^{ns}	0.161^{ns}	0.720^{ns}	0.116^{ns}
AG3 (10)													
Median													
TS	169.0	3.0	3.50	1.0	θ	8.50	9.50	3.25	2.50	2.50	θ	3.0	14.50
CON	168.0	2.50	0.0	0.50	θ	3.50	4.0	3.0	3.0	3.0	θ	3.50	15.25
Significance	U = 102.50	U = 121.50	U = 142.0	U = 113.50	θ	U = 135.0	U = 136.0	U = 105.0	U = 98.50	U = 81.50	θ	U = 89.50	U = 91.0
P	0.879^{ns}	0.216^{ns}	0.004**	0.525^{ns}	θ	0.025*	0.020*	1.0^{ns}	0.645^{ns}	0.066^{ns}	θ	0.233^{ns}	0.306^{ns}

[θ]Unfeasible calculation due to equality of values referring to controls. BT tasks: F = form distortion on BT; R = rotation on BT; I = integration on BT; Pe = perseveration on BT; BL = brain lesion indicators. PS tasks: CF = correlate formation task; SP = relationships between surfaces and perimeters of rectangles task; OPA = order of practical actions task; PSL = projective straight line construction; 3M = points of view or perspectives coordination task “three mountains”. ns = nonsignificant.

Dermatoglyphics

Among the main fingerprint patterns and measurements of TS and their controls, loops, especially ulnar ones, were more frequent in both groups, as much in the left as in the right hand. Ulnar loop was the most frequent pattern in TS subjects in the right hand, left hand and both hands (Table 6). However, radial

Table 6. Frequency in percent of digital patterns, mean ± standard error of the quantitative digito-palmar dermatoglyphic measurements observed in Turner syndrome individuals (TS) and controls (CON) in the left hand (L), in the right hand (R) and in both hands (L+R).

	N	TS	N	CON		P
Patterns						
A						
L	28	2.9	28	9.4		
R	28	2.9	28	7.2		
L+R	28	2.9	28	8.3	$L^{\#}$	0.017*
L^r						
L	28	5.8	28	4.3		
R	28	1.4	28	2.2		
L+R	28	3.6	28	3.2		
L^u						
L	28	65.0	28	52.2	$R^{\#}$	0.002**
R	28	75.7	28	57.5		
L+R	28	70.4	28	54.9		
W						
L	28	26.3	28	34.1		
R	28	20.0	28	33.1	$L+R^{\#}$	0.0001***
L+R	28	23.1	28	33.6		
RC						
L	24	16.83 ± 0.98	26	12.04 ± 1.10	t = 3.23	0.002*
R	25	18.27 ± 0.99	26	12.73 ± 1.00	t = 3.85	0.0003***
L+R	23	35.04 ± 2.00	26	24.80 ± 2.10	t = 3.56	0.001**
Measurements						
a-b						
L	26	44.70 ± 2.10	28	43.46 ± 0.92	t = 0.51	0.610^{ns}
R	27	47.19 ± 1.10	27	42.52 ± 1.00	t = 3.01	0.004**
L+R	26	92.20 ± 2.70	27	85.85 ± 1.80	t = 1.94	0.059^{ns}
A'-d						
L	27	70.60 ± 3.70	28	61.90 ± 2.70	t = 1.92	0.061^{ns}
R	28	61.40 ± 3.60	28	49.10 ± 3.00	t = 2.63	0.011*
L+R	27	132.50 ± 7.00	28	111.00 ± 5.30	t = 2.45	0.018*
atd						
L	25	52.00 ± 1.70	26	47.81 ± 1.70	t = 1.70	0.096^{ns}
R	27	51.30 ± 2.30	26	47.31 ± 1.30	t = 1.51	0.140^{ns}
WI						
L	25	31.71 ± 1.70	27	29.62 ± 1.30	t = 0.98	0.330^{ns}
R	27	29.77 ± 1.70	26	27.98 ± 1.50	t = 0.78	0.440^{ns}
UI						
L	25	59.72 ± 1.20	26	60.84 ± 0.86	t = -0.76	0.450^{ns}
R	27	60.21 ± 1.70	26	62.76 ± 1.40	t = -1.16	0.250^{ns}
TI						
L	18	17.60 ± 0.99	26	22.52 ± 0.93	t = -3.62	0.001**
R	23	20.58 ± 1.20	26	23.40 ± 1.10	t = -1.71	0.094^{ns}
DI						
L	27	51.50 ± 2.30	27	51.90 ± 2.10	t = -0.12	0.900^{ns}
R	28	48.10 ± 1.90	28	48.51 ± 1.70	t = -0.18	0.860^{ns}
MLI						
L	27	7.63 ± 0.57	28	7.36 ± 0.44	t = 0.38	0.710^{ns}
R	28	7.93 ± 0.49	28	8.89 ± 0.44	t = -1.47	0.150^{ns}
L+R	27	15.59 ± 0.91	28	16.25 ± 0.66	t = -0.58	0.560^{ns}

A = arch; L^r = radial loop; L^u = ulnar loop; W = whorl; RC = ridge count; a-b = subdigital triradii "a" and "b" ridge count; A'-d = point A' and subdigital triradius "d" ridge count; atd = maximal "atd" angle; WI = Walker index; UI = Ulnarity index; TI = T line index; DI = D line index; MLI = Main-line index; $^{\#}$Fisher exact test (A + W *vs* $L^r + L^u$); ns = nonsignificant.

loop pattern was more frequent in the left hand for both TS patients and controls. Conversely, this pattern was the least frequent among controls in the left hand, right hand and both hands (Table 6).

Whorls were more frequent in control than in TS girls, although these patterns were the most frequent ones in the left hand. In the right hand, a greater difference was observed between controls and TS individuals. Consequently, frequencies in both hands presented lower variation between groups.

In addition, arch pattern was more frequent in controls than in the TS group: more than 3-fold in the left hand, more than twice in the right hand and approximately 3-fold in both hands. Yet, this was the least frequent digital pattern in the 137 fingers analyzed of the left hand and of 277 fingers of TS subjects. The highest frequency of this pattern was observed in the left hand of controls (Table 6).

Significant differences in fingerprint patterns were found in the ratio between the sum of arches and whorls over loops; all comparisons were significant (Table 6). Differences of ridge count in the left hand and also of A'-d count in the right hand and both hands, between TS individuals and controls, were significant. Differences in ridge count in both hands, a-b ridge count in the right hand and T line index in the left hand were highly significant; differences in ridge count in the right hand were also highly significant. All other quantitative dermatoglyphic parameters gave non-significant differences between subjects and controls.

Mean differences between subjects and controls in relation to left hand, right hand and both hands, and respective standard errors and significance levels were obtained for quantitative digito-palmar dermatoglyphic measurements (Table 7). Palmar ridge counts exhibited significant differences, as follows: a-b count in the right hand, A'-d count in both hands, as well as T line index in the right hand. Similarly, there were differences between patients and controls in relation to ridge count in the left hand, in the right hand and in both hands and also regarding A'-d count in the right hand, as for the T line index in the left hand. The other quantitative dermatoglyphic variables showed non-significant differences when analyzed with the paired Student *t*-test.

DISCUSSION

Bender test

Nearly all propositi (26/28) showed visual motor perception under their chronological age, which corroborates the conclusions of several studies about TS carriers, investigated with BT (Alexander et al., 1966; Brostein and Armendares, 1976; Ricardi, 1996), and other studies using various nonverbal IQ tests (Alexander et al., 1966 apud Dellantonio et al., 1984; Delooz et al., 1993; Ross et al., 2002).

Conversely, most (23/28) of the controls also presented visual motor perception below their corresponding age, which suggests insufficient social-stimulation of the subjects. In fact, Santucci and Galifret-Granjon (1968) admitted that such a lack of stimulation in the development of perceptual organization and in the sense of space might be revealed by BT. However, the difference between patients and controls was not significant (Fisher exact test, P = 0.101).

A higher frequency of rotation and perseveration of patients in BT performances in comparison to controls also confirms that the cognitive difference in TS is a function of a specific aspect of spatial perception. A similar result was observed in a previous investigation, though only in rotation performance (Ricardi, 1996).

Table 7. Mean differences between left hands (L), right hands (R) and both hands (L+R), respective standard errors (SE) and significance values, referring to quantitative digito-palmar dermatoglyphic measurements of the subject-control pairs.

	Pr	Mean ± SE	Significance#	P
RC				
L	23	4.93 ± 1.60	t = 3.08	0.005**
R	23	5.18 ± 1.71	t = 3.03	0.006**
L+R	22	10.43 ± 3.37	t = 3.09	0.005**
a-b				
L	26	1.00 ± 2.47	t = 0.40	0.690ns
R	26	4.77 ± 1.78	t = 2.68	0.013*
L+R	25	6.32 ± 3.59	t = 1.76	0.091ns
A'-d				
L	27	8.85 ± 4.95	t = 1.79	0.085ns
R	28	12.29 ± 4.33	t = 2.84	0.008**
L+R	27	22.15 ± 8.83	t = 2.51	0.019*
atd				
L	23	3.39 ± 2.67	t = 1.27	0.220ns
R	25	4.12 ± 2.54	t = 1.62	0.120ns
WI				
L	24	2.06 ± 2.24	t = 0.92	0.370ns
R	25	1.99 ± 2.30	t = 0.87	0.400ns
UI				
L	23	-1.05 ± 1.44	t = -0.73	0.470ns
R	25	-2.21 ± 2.39	t = -0.92	0.370ns
TI				
L	16	-5.96 ± 1.42	t = -4.20	0.001**
R	21	-3.42 ± 1.40	t = -2.44	0.024*
DI				
L	26	-0.01 ± 3.44	t = -0.00	1.00ns
R	28	-0.45 ± 2.86	t = -0.16	0.880ns
MLI				
L	27	0.18 ± 0.73	t = 0.25	0.80ns
R	28	-0.96 ± 0.64	t = -1.49	0.150ns
L+R	27	-0.89 ± 1.13	t = -0.79	0.440ns

Pr = number of pairs; RC = ridge count; a-b = subdigital triradii "a" and "b" ridge count; A'-d = point A' and subdigital triradius "d" ridge count; atd = maximal "atd" angle; WI = Walker index; UI = Ulnarity index; TI = T line index; DI = D line index; MLI = Main-line index; #*t*-paired test; ns = nonsignificant.

The above results as well as a high frequency of TS subjects with significant brain lesion indicators are likely to be related to a lower brain hemispheric lateralization in TS carriers, when compared to controls. However, this hemispheric specialization has often been established between males and females (Dellantonio et al., 1984). Thus, the establishment of occurrence and nature of such an asymmetry in TS carriers will require further evidence (Nijhuis-Van der Sanden et al., 2003).

Piagetian scales

This investigation continues a previous investigation (Ricardi, 1996) of the first evaluation of TS subjects through PS using clinical methods (Vinh-Bang, 1970) and is the first study employing reflective abstraction and construction of the representation space of Piagetian tasks. Thus, in our investigation, Piagetian scales have been identified as a proper alternative to solve methodological problems found in cognitive evaluations of TS that are not considered in previous investigations (Silbert et al., 1977). This methodological tool, missing in meaningful studies (Ross et al., 2002), is expected to have an important role in preventive psychopedagogical procedures.

Confirming the findings in an initial study (Ricardi, 1996) involving PS operatory performance of TS subjects and controls, we found no significant differences between TS carriers and controls. The data obtained through specific PS (RA and RS) indicated significant differences based on non-parametric tests for both total sample and separate age groups. The exact Fisher test also revealed that there were no significant differences (P = 0.391) between TS and controls in the delay of cognitive performance.

In psychopedagogical evaluation and socialization ability, PS could be used for monitoring biases occurring in usual IQ tests, such as WISC and WAIS, widely employed in neurocognitive analyses, which underestimate cognitive development of TS subjects because of their deficits of spatial perception. However, the PS data could reveal a cognitive compensation in TS performance, which enables them to achieve socio-cultural adaptation. Nevertheless, these hypotheses need confirmation through further investigation.

Dermatoglyphics

In our investigation, the significant difference in arch plus whorl in relation to loop frequencies between TS propositi and controls agrees with previous reports (Otto, 1975; Farman et al., 1979; Otto and Otto, 1980; Ricardi, 1996). There were increased loop frequencies and inversely reduced frequencies of arches or whorls when compared to women from the general population (Penhalber et al., 1994).

In addition, the significant differences in quantitative dermatoglyphics between subjects and controls confirmed conclusions from previous studies (Otto, 1975; Otto and Otto, 1980; Rignell, 1987; Ricardi, 1996). The most remarkable differences were found in digital, a-b and A'-d ridge counts, indicating different patterns of epigenetical organization between groups. Along this line, we found a significant difference in T line index average values between TS and control subjects that was not found in a previous study (Otto, 1975).

Association between Piagetian scales and Bender scores: a cognitive evolution in TS

The set analysis of partial data in this investigation (Ricardi, 2004), and those from a previous study (Ricardi, 1996), revealed a tendency of 45,X TS subjects to improve their BT performance according to age; a result likely due to late maturation for compensating the visual motor deficit (Ricardi et al., 2002).

This tendency was confirmed by illative analysis of our BT results; similarly, there was a significant difference between older TS and control subjects (age group 3) with a lower number of variables of the gestaltic visual motor test (three), in comparison with the performance of the youngest subjects (age groups 1 and 2, four variables; Table 5).

Romans et al. (1998) observed cognitive improvement with age in TS children and teenagers in tasks of specific aspects of spatial ability evaluation, such as judgment of line orientation (JLO) and gestalt closure. They suggested that late maturation, as well as other hypotheses, could explain such cognitive recuperation.

Kesler et al. (2004) submitted 13 TS carriers and their respective age controls to fMRI during their JLO task performance, in two versions with different difficulty degrees. Both groups showed activation of parieto-occipital regions involved in spatial orientation, during their performance in each of the tasks. However, this activation was significantly lower among TS subjects. Controls responded to the most difficult step by releasing executive frontal brain areas; this did not occur in TS individuals. The TS patients presented activation deficit in these areas as well as in parieto-occipital regions during JLO testing. Consequently, Kesler et al. (2004) suggested that activation and possibly deactivation deficits in those brain areas are responsible for visual motor difficulties found among TS carriers.

We found that TS subjects and their controls had no significant difference in Piagetian tasks, including RA and RS tasks in all age groups. These results indicate that TS subjects could have developed a cognitive compensation as they became older, a finding confirming the conclusions in the previous investigation (Ricardi, 1996).

Otherwise, PS-delayed performance exhibited by subjects 2, 17 and 19 could be due to a lack of socio-cultural environment solicitation, a similar performance shown by their respective controls.

Association between dermatoglyphic and cognitive variables: a stepwise multiple regression analysis

Significant differences in the results of a stepwise multiple regression analysis of Piagetian scales and Bender test values (dependent variables) over age and dermatoglyphic parameters (independent variables) were found in TS subjects compared to controls (Tables 8 and 9). Significant regressions of the BT scores of TS individuals were obtained for the following variables: A'-d count, T line index and atd angle (only in the right hand).

Table 8. Stepwise multiple regression analysis of Piagetian scale (PS) values and Bender test (BT) scores over significant independent variables of Turner syndrome subjects.

Hand	Dependent variable	Independent variable	a	$b \pm s_b$	t_o	P	r^2
Left	R	Age	9.742	-0.042 ± 1.440	-3.300	**	45.640
	I		5.830	-0.028 ± 1.220	-2.590	*	33.990
	Total BT		24.370	-0.105 ± 3.670	-3.240	**	44.720
	BL		19.610	-0.065 ± 3.370	-2.180	*	26.810
	CF	atd	6.856	-0.055 ± 0.519	-3.580	**	40.030
		DI		-0.024 ± 0.445	-2.380	*	59.230
	OPA	Age	0.385	0.017 ± 0.496	3.790	**	52.480
	Total PS		7.019	0.050 ± 2.030	2.780	*	37.260
Right	I	Age	6.918	-0.023 ± 1.110	-2.910	*	28.220
		A'-d		-0.029 ± 1.000	-2.160	*	45.280
	Pe	TI	-1.769	0.111 ± 0.668	4.040	***	50.530
	Total BT	atd	1.903	0.192 ± 3.680	2.350	*	25.680
	BL	A'-d	8.705	-0.154 ± 3.060	-4.410	***	27.760
	CF	DI	5.926	-0.038 ± 0.549	-2.710	*	20.650
		atd		-0.025 ± 0.492	-2.220	*	40.240
	OPA	Age	1.021	0.011 ± 0.497	2.840	*	33.490
	PSL		1.619	0.008 ± 0.462	2.250	*	23.980
	Total PS		8.167	0.039 ± 1.770	2.760	*	32.330

R = rotation on BT; I = integration on BT; Pe = perseveration on BT; BL = brain lesion indicators; CF = correlate formation task; OPA = order of practical actions task; PSL = projective straight line construction; A'-d = point A' and subdigital triradius "d" ridge count; atd = maximal "atd" angle; DI = D line index; TI = T line index; a = y intercept; $b \pm s_b$ = regression coefficient ± standard error; t_o = Student test; P = probability; r^2 = determination coefficient; *≤0.05; **≤0.01; ***≤0.001.

Table 9. Stepwise multiple regression analysis of Piagetian scale (PS) values and Bender test (BT) scores over significant independent variables of controls.

Hand	Dependent variable	Independent variable	a	$b \pm s_b$	t_o	P	r^2
Left	F	Age	6.687	-0.030 ± 1.310	-3.940	***	41.350
		Age		-0.035 ± 1.590	-4.080	***	25.080
	R	A'-d	12.445	-0.062 ± 1.480	-2.870	**	37.630
		MLI		-0.310 ± 1.360	-2.240	*	50.180
	I	atd	5.880	-0.060 ± 1.220	-2.290	*	17.400
	Pe	Age	3.439	-0.007 ± 0.412	-3.310	**	29.830
		UI		-0.038 ± 0.384	-2.090	*	41.960
	Total BT	Age	16.000	-0.077 ± 3.020	-4.330	***	45.970
	BL		10.740	-0.040 ± 2.690	-2.530	*	22.560
	CF	Age	-0.591	0.016 ± 0.778	4.530	***	27.600
		DI		-0.042 ± 0.638	-3.630	**	53.520
	SP	Age	-0.318	0.022 ± 0.780	4.810	***	51.240
	OPA	Age	-1.758	0.020 ± 0.635	6.330	***	56.550
		atd		0.039 ± 0.536	3.140	**	70.460
		Age		0.004 ± 0.402	2.910	**	28.820
	PSL	a-b	-0.612	0.068 ± 0.352	6.020	***	47.730
		TI		0.059 ± 0.321	5.060	***	58.780
		UI		-0.033 ± 0.233	-2.720	*	80.450
	Total PS	Age	-4.099	0.067 ± 2.180	5.810	***	55.710
		a-b		0.215 ± 1.950	2.530	*	66.070
Right	F	Age	1.761	-0.020 ± 1.380	-2.510	*	33.380
		DI		0.072 ± 1.240	2.430	*	48.570
	R	Age	4.964	-0.024 ± 1.580	-2.570	*	23.880
	Pe		1.026	-0.007 ± 0.435	-2.640	*	24.940
	Total BT	Age	2.443	-0.082 ± 3.160	-4.800	***	41.370
		UI		0.223 ± 2.790	2.630	*	56.460
		DI		0.115 ± 2.510	2.340	*	31.400
	BL	UI	-7.409	0.190 ± 2.320	3.030	**	44.300
		Age		-0.035 ± 2.030	-2.640	*	59.280
	CF	Age	4.674	0.017 ± 0.753	4.820	***	24.750
		UI		-0.051 ± 0.553	-2.960	**	63.270
	SP	Age	0.402	0.023 ± 0.749	5.980	***	48.790
	OPA	Age	-2.640	0.021 ± 0.606	6.220	***	55.230
		a-b		0.060 ± 0.541	2.520	*	66.010
	PSL	Age	-1.060	0.012 ± 0.510	4.200	***	29.990
		a-b		0.054 ± 0.444	2.780	*	49.480
	Total PS	Age	13.232	0.079 ± 2.230	9.630	***	52.510
		UI		-0.134 ± 1.500	-3.280	**	80.540
		TRC		0.140 ± 1.310	2.640	*	85.980

F = form distortion on BT; R = rotation on BT; I = integration on BT; Pe = perseveration on BT; BL = brain lesion indicators; CF = correlate formation task; SP = relations between surfaces and perimeters of rectangles task; OPA = order of practical actions task; PSL = projective straight line construction; a-b = subdigital triradii "a" and "b" ridge count; A'-d = point A' and subdigital triradius "d" ridge count; atd = maximal "atd" angle; DI = D line index; MLI = main-line index; TI = T line index; UI = Ulnarity index; TRC = total ridge count; a = y intercept; $b \pm s_b$ = regression coefficient ± standard error; t_o = Student test; P = probability; r^2 = determination coefficient; *≤0.05; **≤0.01; ***≤0.001.

The BT scores of controls were significantly associated with A'-d count, Main-line index and atd angle (only in the left hand), D line index in the right hand and Ulnarity index in both hands.

Piagetian scales were significantly associated with atd and D line index in TS propositi (both hands) as well as in controls (left hand). Similarly, PS was associated with T line index and a-b (left hand), total ridge count (only in the right hand) and Ulnarity index (both hands) among controls.

The significant association between several dermatoglyphic parameters and cognitive performance data shown by PS and BT could reveal common epigenetic changes occurring in embryological stages of ectodermic development into central nervous system and dermopapillary ridges. Likewise, Bogle et al. (1994) pointed out a significant association between dermatoglyphic intra-pair difference and behavior discordance in monozygotic twins. As previously discussed by Saldanha (1986), in a comparative study of penta-X syndrome, the "embryonic ectoderm is a parallel system in which developmental instability may be amplified, thus affecting psychomotor development and causing dermopapillary distortions".

There were also significant regressions of psychodiagnotic test and task values over a larger number of dermatoglyphic variables among controls compared to TS subjects. These findings could be due to an inability of the TS subjects to achieve organic balance caused by the absence of an X chromosome. Consequently, such results involve an absence or reduced frequency of more severe psychiatric disturbances, psychopathological deviations and social conflicts in adult carriers of this syndrome (Pelz et al., 1991).

Nevertheless, other research has suggested neuropsychological impairments in TS patients, such as greater psychiatric disturbances, intolerance to stress and low self-esteem (Romans et al., 1998), as well as frequent occurrence of psychosocial inabilities, school problems and hyperactivity (McCauley et al., 2001). Interpersonal relationships could be impaired in TS subjects because of difficulties in recognizing nonverbal communication, such as facial expressions, which could again be due to the TS-specific neurocognitive profile (McCauley et al., 1987). Such a situation requires specific psychopedagogical and psychological preventive assistance and adaptive nursing in TS carriers.

The significant regressions of psychodiagnosis values (especially brain lesion indicators) that we found in TS subjects, upon a greater number of dermatoglyphic variables in the right hand, could be associated with anatomic-functional changes found among the Turner syndrome individuals, mainly in the right-brain hemisphere (Reske-Nielson et al., 1982 apud Nijhuis-van der Sanden et al., 2003) or bilaterally (Brown et al., 2002), by impairing bilateral brain differentiation. Similarly, some studies about other syndromes or psychiatric disorders, such as congenital hemidysplasia with ichthyosiform nevus and limb defects (CHILD syndrome, MIM 308050; Konig et al., 2002), child brain paralysis (Abramova et al., 2000) and schizophrenia (Reilly et al., 2001), would indicate an association between brain degenerations or dysfunctions and lateralized or bilateral differences in epidermis-related traits (nevus or dermatoglyphics), which would have together a pathognomonic meaning in CHILD cases.

Cognitive deficits in psychopedagogical orientation and in genetic counseling

It is well known (Ricardi, 1996) that adaptation of most TS subjects to social environments, including specific cognitive evaluations in school and professional performances, is within average (Nielsen and Stradiot, 1987) or above average (Nielsen and Stradiot, 1987) mean values of the general population. This situation could reveal a unique kind of brain structure and mental performance (Dellantonio et al., 1984; Kesler et al., 2004; Rae et al., 2004), characterized by identification with their social environment and susceptibility to enrich social cohesion, bringing particular abilities, capabilities and trends of thinking with their peer group, which would help make a lasting social network (Levy, 1977).

However, TS individuals usually display cognitive and psychosocial difficulties (Delooz et al., 1993; Romans et al., 1998; McCauley et al., 2001; Ross et al., 2002) and need appropriate treatment and orientation (Nielsen, 1989; Delooz et al., 1993; McCauley et al., 1987, 2001). Case advice and support groups for those TS carriers should guarantee available information to parents and health and education professionals, avoiding parental super-protection risks (Nielsen and Stradiot, 1987; Pelz et al., 1991; Batch, 2002).

In addition, we advocate environmental solicitation as a psycho-pedagogical approach for supporting educative principles coherent with Piagetian theory (Mantovani de Assis, 1976; Zaia, 1996) for the improvement of cognitive development of TS subjects. This process could speed up the optimization of cognitive compensation, previously revealed in girls and teenagers by the Bender test. Accordingly, genetic and/or psychological counseling centered on genetic risk should be supported by a multi- and interdisciplinary staff (Saldanha, 1983). This approach in genetic counseling includes recruitment of pertinent family members for a proper perception of their resources and limitations and how to deal adequately with their real economic, social, cultural, and educational circumstances (Saldanha, 1978).

Finally, the psychodynamic process of interpersonal communication centered on genetic risks constitutes a modality of psychological counseling that must also consider the clinical risks associated with severity, prognosis, morbidity, and lethality degrees of the hereditary condition, as well as the psychological risks, generated by the emotional-affective correlates revealed by family members, in order to deal with these specific risks for obtaining necessary emotional readiness and providing lucid decisions. Genetic counseling, under such a perspective, transits along directive information (based only upon genetic risk) to achieve a non-directive (centered as a whole on the client and the family) approach (Saldanha, 1973, 1978, 1983).

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