

Title:

Executive Function in Preschool and Early School Aged Children with Sex Chromosome Trisomy

Authors:

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Introduction:

Sex chromosome trisomies (SCTs) are among the most common chromosomal variations. Children with these genetic differences are at increased risk for deficits in neurocognition such as executive functioning (EF). Despite growing awareness, there remains limited understanding of how these conditions affect EF in early childhood. This knowledge gap hinders the development of targeted interventions and support strategies.

Methods:

This is a longitudinal analysis from the eXtraordinary Babies Study of children with prenatally identified SCT. Behavior Rating Inventory of Executive Function (BRIEF) Preschool Version surveys were completed by parents at 2, 3, 4, and 5 year visits and BRIEF-2 surveys were administered at 6 and 8 year visits; to be included in this analysis, participants had to have at least one BRIEF-P and one BRIEF-2 survey completed. T-scores from the overlapping domains of the BRIEF-P and BRIEF-2 surveys (Inhibition, Shifting, Emotional Control, Working Memory, Planning/Organization, and the Global Executive Composite) were compared. Counts and percentages for elevated scores (t-score >60) were calculated. Logistic regressions adjusted for SCT condition were modeled for each domain to determine the odds of an elevated BRIEF-2, given an elevated BRIEF-P. Analyses were performed in R, version 4.4.0.

Results:

A total of 55 participants had both BRIEF-P and BRIEF-2 assessments (38 47,XXY, 10 47,XXX, and 7 47,XYY). Table 1 presents a summary of elevated BRIEF scores by domain and BRIEF version. The proportion that were elevated on the BRIEF-P ranged from 21.8% (Shifting) to 40.0% (Planning/Organization), while BRIEF-2 elevations ranged from 20.0% (Planning/Organization) to 32.7% (Emotional Control). For all domains, participants who were elevated on the BRIEF-P had significantly higher odds of being elevated on the corresponding BRIEF-2 domain. The odds ratios ranged from 4.9 for Working Memory (95% CI: [1.5, 18.2]; $p = 0.012$) to 12.4 for Inhibition (95% CI: [2.7, 73.8]; $p = 0.002$). Specifically, 12.7–20.0% of participants were elevated on both instruments across domains (Figure 1).

Conclusions:

In children with prenatally identified SCT, vulnerabilities in some EF domains can be appreciated in early childhood. Elevated scores in preschool-age children strongly predict later EF concerns in early school age.

Table 1. Elevation Counts by Domain and BRIEF version

	BRIEF-P	BRIEF-2	Elevated on Both BRIEF-P and BRIEF-2	Odds Ratio (95% CI) ¹ Elevated BRIEF-2 Given Elevated BRIEF-P
Inhibition	13 (23.6%)	12 (21.8%)	7 (12.7%)	12.4 [2.7, 73.8]; p = 0.002
Shifting	12 (21.8%)	15 (27.3%)	8 (14.5%)	9.9 [2.2, 55.6]; p = 0.004
Emotional Control	16 (29.1%)	18 (32.7%)	11 (20.0%)	11.1 [2.9, 50.2]; p < 0.001
Working Memory	21 (38.2%)	17 (30.9%)	11 (20.0%)	4.9 [1.5, 18.2]; p = 0.012
Planning/Organization	22 (40.0%)	11 (20.0%)	8 (14.5%)	5.4 [1.3, 28.4]; p = 0.027
Global Executive Composite	15 (27.3%)	16 (29.1%)	10 (18.2%)	10.9 [2.9, 47.9]; p < 0.001

¹Adjusted for SCT diagnosis

Figure 1. Maximum BRIEF T-Scores Trajectories between BRIEF Versions

