Verbal Perseveration in Boys with Fragile X and Down syndrome

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Abstract
Perseveration and “loss-of-set” on a verbal fluency test were examined in boys with fragile X syndrome with and without autism spectrum disorder (FXS-ASD, FXS-O), boys with Down syndrome (DS), and typically developing (TD) boys. Compared with TD boys, boys with FXS-O and FXS-ASD produced significantly more recurrent perseveration and boys with FXS-ASD produced significantly more loss-of-set responses after controlling for nonverbal mental age. However, significant differences did not remain after also controlling for expressive vocabulary skills. Clinical implications will be discussed.

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Background
- FXS – most common inherited cause of intellectual disability (ID) and leading single-gene disorder associated with autism
- DS – most common genetic cause of ID
- Perseveration and noncontingent language during conversation commonly reported for males with FXS
- Purpose of study – compare perseveration and “loss-of-set” responses of boys with FXS-O, FXS-ASD, DS, and TD on verbal word fluency test

Table 1.
Study Participants

<table>
<thead>
<tr>
<th></th>
<th>FXS-O</th>
<th>FXS-ASD</th>
<th>DS</th>
<th>TD</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>N</strong></td>
<td>30</td>
<td>24</td>
<td>28</td>
<td>25</td>
</tr>
<tr>
<td><strong>Mean Age (SD in years)</strong></td>
<td>11.8(2.54)</td>
<td>11.1(2.71)</td>
<td>10.8(3.07)</td>
<td>5.4(.80)</td>
</tr>
<tr>
<td><strong>Age Range</strong></td>
<td>6.4-15.8</td>
<td>7.4-15.5</td>
<td>6.3-17.0</td>
<td>3.9-7.3</td>
</tr>
<tr>
<td><strong>Mean Leiter-R Mental Age</strong></td>
<td>5.5(.65)</td>
<td>5.2(.43)</td>
<td>5.3(.86)</td>
<td>5.5(1.07)</td>
</tr>
<tr>
<td>(SD)</td>
<td>5.2(1.32)</td>
<td>5.1(1.32)</td>
<td>4.9(1.30)</td>
<td>6.0(1.31)</td>
</tr>
<tr>
<td>-------------</td>
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</tr>
<tr>
<td>Mean EVT Age Equivalent (SD)</td>
<td></td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

### Procedure
- Verbal Fluency subtest of *NEPSY* (Korkman, Kirk, & Kemp, 1998) – child generated words in two semantic categories (Animals & Food/Drink)
- Coded responses for:
  - Perseveration (Sandson & Albert, 1984)
    - Stuck-in-set (continuation of category)
    - Recurrent (repetition of previous response after intervening response)
    - Continuous (consecutive repetition of previous response)
  - Loss-of-set
    - Proactive interference
    - Tangential
    - Unrelated

### Results
- Conducted multivariate general linear model and ANCOVA
- Groups differed significantly on perseveration and loss-of-set with Leiter-R Mental Age included as a covariate (Table 2)
- Boys with FXS-O and FXS-ASD produced significantly more recurrent perseveration than TD boys
- Boys with FXS-ASD produced significantly more loss-of-set responses overall than TD boys
- Significant differences did not remain when second covariate, age equivalent score on the Expressive Vocabulary Test (Williams, 1997), was added to model

Table 2.
Adjusted Mean Proportions (adjusted for Mental Age)

<table>
<thead>
<tr>
<th>Outcome</th>
<th>FXS-O M (SE)</th>
<th>FXS-ASD M (SE)</th>
<th>DS M (SE)</th>
<th>TD M (SE)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stuck-in-set Perseveration</td>
<td>.003(.003)a</td>
<td>.004(.003)a</td>
<td>.002(.003)a</td>
<td>.000(.003)a</td>
</tr>
<tr>
<td>Recurrent Perseveration</td>
<td>.210(.028)a</td>
<td>.226 (.031)a</td>
<td>.187(.029)a,b</td>
<td>.100(.030)b</td>
</tr>
<tr>
<td>Continuous Perseveration</td>
<td>.051(.010)a</td>
<td>.047(.011)a</td>
<td>.022 (.011)a</td>
<td>.019(.011)a</td>
</tr>
<tr>
<td>Loss-of-set</td>
<td>.088(.018)a,b</td>
<td>.120(.020)a</td>
<td>.080(.019)a,b</td>
<td>.027(.020)b</td>
</tr>
</tbody>
</table>
Conclusions

• Findings
  o Boys with FXS, regardless of autism status, produced more recurrent perseveration than TD boys
  o Boys with FXS-ASD produced more loss-of-set responses than TD boys
  o Significant differences not found after controlling for EVT scores, suggesting an association between perseveration and loss-of-set and expressive vocabulary skills

• Clinical Implications
  o Perseveration and loss-of-set should be assessed in boys with FXS
  o Reduction of perseveration and loss-of-set in boys with FXS likely to be important intervention goals

• Future Research
  o Compare perseveration and loss-of-set of boys with FXS-ASD and boys with ASD only
  o Examine associations between perseveration and loss-of-set on verbal fluency test and perseveration and noncontingent language during connected speech
  o Examine perseveration on other structured tasks