Articulation in schoolchildren and adults with neurofibromatosis type 1
Marjan Cosyns*, Geert Mortier, Sandra Janssens, Famke Bogaert, Stephanie D’Hondt, and John Van Borsel

Introduction
Several authors mentioned the occurrence of articulation problems in neurofibromatosis type 1 (NF1) patients, variously referred to as “mild articulation difficulties”, “difficulties in speech articulation”, “imprecise consonants”, “mispronunciations”, “incorrect sequencing of sounds”, “lispings articulations”, “dysarticulation”, “difficulties pronouncing speech sounds”, “misarticulation”, and “articulation delay”. However, few studies have undertaken a detailed articulation analysis. Therefore, the aim of the present study was to document the articulation skills of Flemish schoolchildren and adults with NF1 both phonetically and phonologically, based on a standardized speech sample of 135 mono- and polysyllabic Flemish words containing all Flemish single speech sounds and most clusters in all their permissible syllable positions.

Methods
Participants
The study group consisted of 43 NF1 patients: 14 children and 29 adults. All fulfilled the diagnostic criteria for NF1, and none was diagnosed with oral, pharyngeal, or laryngeal neurofibromas. Demographic details are presented in Table 1.

Data collection
Speech samples (Figure 1) were recorded using a Sony HDR-SR1E camcorder, transcribed phonetically, and then analyzed. Phonetic transcriptions were made by two Master students in Logopaedic Sciences (F.B. and S.D.) and checked by a certified speech-language pathologist (M.C.). Analysis concentrated on consonants only and included the following:

1. A phonetic inventory analysis, which encompasses the determination of the Flemish consonants that a person is capable of producing without making reference to the target sounds.
2. A phonetic analysis, in which the consonants produced are compared with the target productions and errors at the segmental level are identified.
3. A phonological analysis, in which the consonants produced are compared with the target productions and errors between the segmental level are identified.
4. The calculation of the Percentage of Consonants Correct (PCC; Shriberg & Kwiatkowski, 1982). The PCC, a severity measure, is obtained by dividing the number of correct consonant productions by the total number of consonants and multiplying by 100.

Results
Phonetic inventory analysis
Incomplete inventories were found in 2 children (14.29%) and 5 adults (17.24%), all males. All had difficulties producing the sibilants /ʃ/ and /ʒ/ correctly. The /ʃ/ was substituted by either /s/ or /c/ and /ʒ/ by either /z/ or /s/.

Phonetic analysis
A two-way ANOVA (Figure 2) revealed that children made significantly more errors than adults (p = .002), and, regarding error type, distortions occurred significantly more than substitutions, omissions, and additions (Scheffé tests, p < .001). The most common distortions were sigmatismus stridens (i.e., a whistling /s/) and multiple ad- or interdentity (i.e., lisp). In children, rhotacismus non vibrans (i.e., an untrilled production of /r/) also occurred frequently.

Phonological analysis
Again, it was observed (Figure 3) that children displayed significantly more errors than adults (p = .015). Further, it was demonstrated that substitution errors occurred significantly more than assimilation errors (Scheffé tests, p = .045). Syllable structure errors did not differ significantly from substitution errors nor from assimilation errors. Devoicing and cluster simplification were the most common phonological errors observed. In addition, all children occasionally deleted the final consonant of a word.

PCC
In children, PCC ranged from 77.91% to 99.30% (M = 90.52, SD = 6.39) and differed significantly from that of adults (range = 86.87 – 99.77%, M = 96.07; SD = 2.83).

Conclusion
The speech of NF1 patients is characterized by mild articulation deviations. Sigmatismus stridens, multiple ad- or interdentity, devoicing, and cluster simplification seem to be the predominant articulation errors. The type of errors observed and the fact that articulation difficulties do not resolve completely with advancing age, leads us to designate the articulation errors observed as “residual phonological errors” (Ruscello, 2003).

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Table 1. Demographic details of the study group

<table>
<thead>
<tr>
<th></th>
<th>NF1 children</th>
<th>NF1 adults</th>
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<tbody>
<tr>
<td># Males/females</td>
<td>7/7</td>
<td>15/14</td>
</tr>
<tr>
<td>Mean (age)</td>
<td>10.8</td>
<td>34.11</td>
</tr>
<tr>
<td>SD (age)</td>
<td>2.9</td>
<td>9.9</td>
</tr>
<tr>
<td>Min (age)</td>
<td>7.5</td>
<td>17.11</td>
</tr>
<tr>
<td>Max (age)</td>
<td>16.0</td>
<td>53.6</td>
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</tbody>
</table>

Note. Age is expressed in years; months.

Figure 1. Articulation test for children. Adults were given either a description of the target word or a phrase that had to be completed with the target word.

Figure 2. Frequency of phonetic errors by error type for both NF1 children and adults

Figure 3. Frequency of phonological errors by error type for both NF1 children and adults