# Articulation in Beckwith-Wiedemann Syndrome: Two Case Studies

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Beckwith-Wiedemann syndrome (BWS) is a genetic disorder with abdominal wall defects, gigantism, and macroglossia as its main characteristics. A number of investigators have reported the presence of articulation errors in individuals with BWS due to macroglossia. However, few data are available on the exact nature of the articulation problems of subjects with BWS. This paper presents two case studies that highlight the articulatory characteristics associated with BWS. Subjects were a boy aged 5 years 9 months and a girl aged 3 years 6 months. A phonetic analysis was conducted in which it was found that primarily consonants with an anterior place of articulation were affected. The error patterns appeared to be related to inappropriate tongue and lip postures. An observer experiment in which naive and expert observers rated speech samples from three modes of presentation (auditory-only, visual-only, and audiovisual) showed that the subjects' speech was more disturbed visually than auditorily.

**Key Words:** Beckwith-Wiedemann syndrome, articulation, macroglossia

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Adler (1976) suggested that specific genetic syndromes may cause specific speech and language disorders. As such, clinicians should be aware of this possibility. Unfortunately, the role of speech-language pathologists in the differential diagnosis of congenital malformation syndromes is limited, a situation according to Sphrintzen (1997) that is untenable. If speech-language pathologists are to become more active team members in the identification of various genetic syndromes, knowledge of the phenotypic spectrum of genetic syndromes with respect to communication disorders urgently needs to be expanded. Moreover, knowledge of the phenotypic spectrum along with recognition of the natural history and prognosis of a disorder is critical to proper patient care (Sphrintzen, 1997), since such knowledge helps the clinician to focus diagnostic efforts and to direct plans for treatment. The present study represents a characterization of the phenotypic spectrum of Beckwith-Wiedemann syndrome (BWS).

BWS is a congenital multiple anomaly disorder first described by Beckwith (1963) and Wiedemann (1964). The condition has an estimated incidence of one in 13,700 births and affects males and females with equal frequency (Engström, Lindham, & Schofield, 1988). The majority of cases are sporadic occurrences. In about 15% of persons with BWS, there is an autosomal dominant inheritance (Pettenati et al., 1986). In a limited number of patients (e.g., 3 out of 76 in a study by Elliott, Bayly, Cole, Temple, & Maher, 1994), karyotypic abnormalities have been found at chromosome 11p15.5 (paternally inherited chromosome 11p15.5 duplication or additional chromosomal material of unidentified origin). The syndrome is characterized by three major features: abdominal wall defects, gigantism, and macroglossia (abnormal enlargement of the tongue). Other typical features include excessive creasing of the earlobes, facial nevus flammeus (discolored skin, so-called port wine stains), organomegaly (enlargement of the liver or spleen), nephromegaly (enlargement of the kidneys), and neonatal bursts of hypoglycemia (decreased levels of glucose in the blood). More rarely, patients show a hemihypertrophy (overgrowth of tissue on one side of the body), moderate to severe mental retardation, congenital heart defects, polydactyly (extra fingers or toes), neoplasia (development of a tumor), and cleft palate (Elliott et al., 1994).

Macroglossia appears to be the most constant anomaly in BWS, occurring in up to 82% of the patients with the syndrome (Elliott et al., 1994; McManamny & Barnett 1985; Pettenati et al., 1986). The macroglossia of patients with BWS is considered to be a true macroglossia due to muscle fiber hyperplasia, as opposed to relative macroglossia, in which no specific cause for tongue enlargement can be found (Vogel, Mulliken, & Kaban, 1986).

It is not uncommon for patients with BWS to undergo partial glossectomy for reducing the size of the tongue. In a cohort of 76 patients studied by Elliott et al. (1994), surgical tongue reduction had been performed in 28 cases (37%). And in 29 patients with BWS with macroglossia admitted to the Royal Children's Hospital in Melbourne, Australia, between 1973 and 1983, 16 (55%) underwent a glossectomy (McManamny & Barnett, 1985). In infancy and early childhood, partial glossectomy is usually performed because of respiratory difficulties (obstructive apnea) and/or feeding difficulties (Weng, Mortier, & Graham, 1995). Indications for surgical tongue correction during later childhood appear to be cosmesis, orthodontic abnormalities (especially an anterior open bite deformity and mandibular prognathism, i.e., marked protrusion of the mandible), and articulation difficulties (McManamny & Barnett, 1985; Menard, Delaire, & Schendel, 1995; Patterson, Ramasastry, & Davis, 1988; Rimell, Shapiro, Shoemaker, & Kenna, 1995; Siddiqui & Pensler, 1990; Smith, Mihm, & Flynn, 1982). It has also been suggested that surgical reduction of the tongue in early life may safeguard against dental deformity (Kveim, Fisher, Jones, & Gruer, 1985) and prevent speech defects (Shafer, 1968).

Little information is available, however, on the prevalence and nature of speech defects in subjects with BWS. According to McManamny and Barnett (1985), consonant production is affected in BWS, especially labiodental and linguodental sounds and alveolar fricatives. Production of linguodental sounds and alveolar fricatives was difficult in a 7-year-old patient reported by Mixter, Ewanowski, and Carson (1993). The patient had "difficulty elevating the tongue tip to produce the /t/, /d/, and /l/ sounds as well as positioning the tongue blade for the /s/, /z/, sh and zh sounds" (p. 1160). In another patient, 5 years old, Mixter and colleagues observed speech that was "marked by oral distortions on a number of different sounds, mostly consonants, as well as on several vowels" (p. 1161). In a sample survey (questionnaires filled out by parents) by Van Borsel, Van Snick, and Leroy (1999), 29 of the 40 Dutch-speaking BWS patients studied reportedly had articulation problems. The articulation errors persisted in some patients after corrective surgery. Of the Dutch singleton consonants, most problems occurred with alveolar sounds, especially /s/. The subjects also had problems, but to a lesser degree, with labiodental sounds /f/ and /v/, and with /r/. Some other sounds were problematic in only one subject (/p/, /b/, /ʃ/, /x/, /z/, /ɣ /, /n/, /ŋ/, /j /, /h/), whereas some sounds were never reported to be defective (/k/, /ʒ/, /m/, /w/). Of the initial consonant clusters, /dr/, /sx/, /str/, /kr/, and /sl/ were reported to be defective; of the final consonant clusters, /rst/, /rs/, and /st/ were misarticulated. None of the respondents noted any problem vowels.

As far as we could ascertain, there does not seem to exist any information on the severity of the articulation problems in individuals with BWS, either in terms of actual deviation or in terms of effects on the listener. There is some suggestion from data on the persons with Down syndrome that in cases of macroglossia, there may be a dissociation between the actual deviation and the severity of the problem as rated in terms of aesthetic appearance (i.e., visual acceptability of a person while speaking). Klaiman, Witzel, Margar-Bacal, and Munro (1988) had listeners rate preoperative and postoperative videotaped samples of eight patients with Down syndrome who underwent partial glossectomy for speech intelligibility and aesthetic appearance. Analysis revealed that after surgery, speech was significantly more acceptable aesthetically, but was not different in terms of speech intelligibility. In individuals with BWS, the effect of a macroglossia on the appearance of speech as compared to the actual deviance of articulation is an aspect that has not yet been studied.

Apart from the fact that the prevalence, nature, and severity of speech defects in individuals with BWS are poorly documented, it is not clear how the speech defects originate. The occurrence of articulation disorders is usually believed to be associated with the macroglossia that typifies the syndrome. Some investigators even assume that the articulation disorders derive directly from the macroglossia. McManamny & Barnett (1985), for instance, point out that the overly large tongue significantly alters the relationship between teeth and tongue. In spite of the fact that compensation for the dental deviation would be possible, the altered relationship would affect consonant production, especially labiodental and linguadental sounds and the alveolar fricatives. Mixter et al. (1993) mentioned that the tongue tip cannot be elevated to produce the /t/, /d/, and /l/ and that the tongue blade cannot be positioned as required for the /s/, /z/, /ʃ/, and /ʒ/ because of its size. Other investigators suggested that dentoskeletal abnormalities that can develop in response to the enlarged tongue may contribute toward speech disorders. Among these anomalies are mandibular prognatism, bimaxillary protrusion, anteriorly inclined mandibular incisors, and apertognathia (Patterson et al., 1988). However, exactly how these anomalies relate to tongue size is unclear. Another hypothesis to be considered is that, at least to some degree, the articulatory patterns observed in persons with BWS are due to compensatory movements that result from the size of the tongue. In individuals who underwent a tongue reduction it has been suggested that some articulation defects are due to alteration of lingual morphology (Siddiqui & Pensler, 1990) or to residual scarring (Mixter et al., 1993).

In summary, data on articulation abilities in BWS are scarce. Therefore, the purpose of the present study was to further delineate and document the speech problems associated with macroglossia in BWS patients. To this end, we report on a perceptual analysis of speech characteristics in two children with BWS. The investigation included two parts, the first of which (Study 1) was a phonetic analysis of the children's speech. In the second part (Study 2), speech of the two BWS subjects was perceptually rated for severity using a variety of modalities (auditory only, visual only, and audiovisual).

## Study 1

### Subjects

The subjects for this study were two children with BWS, a boy (Subject 1) and a girl (Subject 2), aged 5;9 (years;months) and 3;6, respectively.

#### *Subject 1 (S1)*

S1 was born after a normal pregnancy of 36 weeks. The delivery was normal and his birth weight was 3.1 kg (6.8 lbs) and his length was 48 cm (18.9 in.). The diagnosis of BWS was made clinically in the neonatal period. Macroglossia, hypoglycemia, hypocalcemia, and hyperthermia were noted and his record also mentioned hyperinsulinemia, episodes of jittery limb movements, cyanosis, and convulsions. According to his mother, postneonatal development was normal. He is the only child in his family and there is no evidence of features reminiscent of BWS in any other member in his pedigree.

When first seen at the Ghent University hospital at age 1;8, the diagnosis of BWS was confirmed. Clinical examination showed an obese but healthy boy with open mouth, relatively large tongue, and earlobe creases. Weight and head circumferences were above the 90th percentile, and height was between the 25th and 50th percentiles. The physical examination revealed no abnormalities except for a prominent abdomen with no abnormal masses, and a small but unmistakable omphalocele (navel rupture). S1's limbs were of normal proportion but looked somewhat squat. Neurological examination was normal although there was mild generalized hypotonia. S1 was alert with age-appropriate behavior. As for language development, he had reached the first word stage.

At age 3;2, S1 underwent an adenoidectomy and tonsillectomy because of frequent episodes of otitis media. One year later, the adenoidectomy was repeated and ear tubes were placed. In the same period, cryptorchidism (undescended testes) and the omphalocele were surgically corrected following complaints of abdominal pain.

At the age of 4;3, when S1 was in the second year of kindergarten, he was tested in a speech and hearing rehabilitation center because of delayed motor, speech, and language development. Psychological testing (Dutch version of the McCarthy Scales of Children's Abilities; Van Der Meulen & Smrkovsky, 1985) showed normal intelligence (general cognitive index: 103) but a heterogeneous profile with low scores on memory and language subtests. S1 was characterized as restless, nervous, distractable, and having poor concentration. On a test for neuromotor development, the Körperkoordinationstest für Kinder (Kiphard & Schilling, 1974), S1 obtained a motor quotient of 95 (32nd percentile). It was noted that S1 scored more poorly on all items involving tonus control and postural stability. Hearing (pure-tone audiometry) tested normal. Concerning speech, records indicated that on a standardized Dutch articulation test (Utrechts Artikulatie Onderzoek; Peddemors-Boon, Van Der Meulen, & De Vries, 1977), S1 showed interdental articulation of lingua-alveolar sounds, omissions, substitutions, and cluster reductions without detailing which sounds and clusters were involved. It was mentioned, however, that alveolar sounds were produced by the tongue making contact with the upper lip rather than with the alveolar ridge. And it was noted that the latter way of articulating resulted in intelligible speech.

A CT scan of the abdomen at age 4;9 revealed nephromegaly (kidney enlargement), especially on the left side, and a large but still normal-sized pancreas and spleen. On a follow-up clinical examination at the age of 5;0, S1 remained a relatively large (length: 90th percentile) and obese (weight: 75th–90th percentile) child with the mouth still open and the tongue still wide and especially thick. In comparison with the findings at 1;8 years, however, the macroglossia had decreased and the tongue was more mobile.

Data for the present study were collected at age 5;9. No speech treatment had been given until that time, in spite of the findings of speech tests at age 4;3.

#### *Subject 2 (S2)*

S2 was born at term as the third child of a second marriage after a complicated twin pregnancy. From 2 months of gestation, premature contractions made bed rest and treatment with B-mimetics necessary. After 31 weeks of gestation, the twin brother of S2 was stillborn. Postnatal hypoxia in S2 was resolved effectively by administration of oxygen and 3 days of incubation treatment. Her weight at birth was 2.970 kg (6.5 lbs), her length was 44 cm (17.4 in.), and her head circumference was 33 cm (13 in.). Clinical examination at approximately 3 months of age showed an alert child with a remarkably large tongue, ear creases on both earlobes, and a small exomphalos (umbilical hernia). This prompted the clinical diagnosis of BWS. The macroglossia did not cause respiratory problems or feeding problems. Neurological examination was normal. Her weight was 6.200 kg (13.7 lbs; 50th–90th percentile), her length was 57.5 cm (22.6 in.; 10th–50th percentile), and her head circumference was 40.6 cm (16 in.; 50th–90th percentile). A cytogenetic analysis at age 4 months revealed a normal female karyotype without any structural or numerical defects in the region 11p15.5. Subsequent follow-up examinations at age 6 months, 1 year, and 1;8 years confirmed the impression of normal development without any complications. S1 sat up unaided at 6 months, walked at age 1, and spoke her first recognizable words at age 1. At age 1;8, splenomegaly (enlarged spleen) and a slight nephromegaly (enlarged kidney) were observed ultrasonographically. The macroglossia, however, spontaneously regressed over a period of months and the tongue progressively fit better within the oral cavity. Data for the present study were gathered at age 3;6. Like S1, no speech treatment had ever been initiated for S2.

## Method

### *Data Collection*

In both subjects, a speech sample was collected that served as the database for this study. The procedure used was similar to that of Van Borsel (1996) in a study of articulation in Down syndrome adolescents and adults. Subjects were shown a randomized series of 135 black-and-white drawings of common objects and actions, which they were required to name. The pictures were chosen so that, when named correctly, they elicited instances of all Dutch single consonants in all permissible positions, most consonant clusters, and all vowels and diphthongs. (See Appendix for information on Dutch phonology.) The entire session took place in a sound-protected room at the university hospital and was videotaped using a Sony CCD-V900E recorder.

### *Phonetic Transcription*

Subsequently, two of the authors (JVB, KVS) made a consensus phonetic transcription of the recorded speech samples. First, we simultaneously, but independently, transcribed the sample and then compared the transcriptions. In the case of disagreements, we replayed the tape and discussed transcriptions until a consensus was reached. In transcribing the speech samples, the symbols and diacritics of the International Phonetic Alphabet (International Phonetic Association, 1949—the most recent version available to the authors at the time of the investigation) were used, supplemented with ad hoc signs and verbal descriptions when deemed necessary. Both auditory and visual aspects of the subjects' articulation were transcribed as narrowly as possible. In S1, only spontaneous productions identifying the pictures without any help were considered for transcription and further analysis. S2, the younger subject, was often unable to spontaneously name a stimulus picture. In her case, both spontaneous responses and responses elicited through imitation of the examiner or the child's mother were retained. In addition, incorrect identifications of a stimulus picture of which the target was unmistakably clear were also included. On the other hand, for both subjects, a small number of words could not be transcribed reliably and repeated listening to the tape did not yield a consensus transcription; these words were omitted from the analysis. In S1, the database sample consisted of 142 different words totalling 436 consonants and 241 vowels and diphthongs. In S2, a speech sample of 136 different words totalling 405 consonants and 215 vowels and diphthongs was obtained.

### *Analysis*

The transcribed speech samples were then subjected to several analyses. These included a phonetic inventory analysis; a phonotactic analysis; an assessment of error rate; an analysis of sounds affected; an error analysis in terms of the traditional categories of omission, substitution, and distortion; and an analysis of error patterns.

## Results

### *Phonetic Inventory Analysis*

A phonetic inventory analysis was conducted to assess the vowels, diphthongs, and consonants of the mother tongue the subjects were capable of producing. The inventory was conducted without making reference to the intended target sounds. A sound was considered to be present in the inventory when at least two instances of totally correct realizations of the sound were found. A totally correct realization was defined as a production that is consistent with the standard realization or with the local dialect. However, in deciding that a sound was absent from a subject's inventory, it was taken into account that there should have been a sufficient number of opportunities for the sound to occur. Recognizing the possibility of phonetic instability in realizing sounds, a criterion of at least four opportunities was adopted. Results of the phonetic inventory analysis are shown in Tables 1 and 2. As seen in Table 1, the vowel and diphthong inventory was nearly complete in both subjects. The only sound that was definitely lacking in S2 was the /u/. In addition, neither subject had two correct instances of the vowel /œ/. However, both subjects produced only two target words containing this vowel, and they were apparently capable of producing the /œ/ as the

**TABLE 1. Results of the vowel and diphthong inventory analysis for S1 and S2.**

| Sounds | i | y | e | ɛ | ø | ɪ | ə | œ | a | ɑ | u | o | ɔ | ɛi | œy | ɔu |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Subject S1 | + | + | + | + | + | + | + | x | + | + | + | + | + | + | + | + |
| Subject S1 | + | + | + | + | + | + | + | x | + | + | – | + | + | + | + | + |

*Note.* + = sound is present in the inventory; – = sound is absent from the inventory; x = fewer than four opportunities for the sound to occur.

**TABLE 2. Results of the consonant inventory analysis for S1 and S2.**

| Sounds | p | b | m | w | f | v | t | d | n | s | z | l | ʃ | ʒ | j | k | ɣ | x | ŋ | ʀ |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Subject S1 | + | + | – | + | + | + | + | + | + | + | + | + | – | x | + | + | + | + | + | + |
| Subject S1 | + | – | + | – | – | – | + | – | + | + | x | + | x | x | + | + | + | + | + | + |

*Note.* + = sound is present in the inventory; – = sound is absent from the inventory; x = fewer than four opportunities for the sound to occur.

first element of the diphthong /œy/. Results for consonants are shown in Table 2. In the consonant inventory of S1, only the /m/ and /ʃ/ were lacking. In the inventory of S2, the /b/, /w/, /f/, /v/, and /d/ were absent.

Consonants that have two permissible positions (syllable initial and syllable final) in Dutch were examined further for possible distributional constraints in the subjects' inventories. Since some sounds did not have several opportunities to occur in each position, we settled for a criterion of one occurrence of a sound to be considered present in this analysis. It should be stressed that this low criterion, as well as the limited number of possible occurrences in some cases, compels one to interpret the results with much caution. Of all the consonants that were present in the inventory of S2 and have two permissible positions, examples were found in syllable initial as well as syllable final position. S1, on the other hand, had no example of a correct /j/ in syllable final position, but there had been only one possibility for /j/ to occur in this position.

### *Phonotactic Analysis*

In the phonotactic analysis, target words and actual productions of the subjects were compared for number of syllables and for syllable shapes. The latter was done irrespective of whether target consonants and vowels were correctly produced.

It was found that, with one exception, both subjects always preserved the number of syllables of a target word. The speech sample of S1 contained 66 one-syllable words, 53 two-syllable words, 20 three-syllable words, and 3 four-syllable words. In all instances, the number of syllables of the words produced matched the number of syllables of the target words. The speech sample of S2 contained 71 one-syllable words, 45 two-syllable words, 17 three-syllable words, and 3 four-syllable words. Only once did the number of syllables produced not match the number of syllables of a target word. In a four-syllable word, S2 deleted an unstressed syllable.

Table 3 lists all the different syllable shapes that occurred in the subjects' speech samples. Comparison of the actual productions with the target words showed that the target syllable shapes had been almost always retained. In S1, syllable shape was changed in only 2.8% (4/142) of the words produced. Three times a nasal /n/ was added to words of the structure CV/CVC ending on /l/ (C = consonant, V = vowel, / = syllable boundary). In one three-syllable word, the onset of the first and the second syllable was rendered identical by addition of a sound in the second syllable. In S2, syllable shape was changed in only 6.6% (9/136) of the words produced. As previously mentioned, S2 deleted an entire unstressed syllable in a four-syllable word. In two words, there was an addition of a consonant (once to the initial vowel of a V/CV/CVCC word, and once after the initial consonant of the second syllable of a CCVC/CV/CVC word). In six words, an omission of a consonant occurred in either the initial consonant of a consonant cluster (three words) or in a syllable initial or syllable final singleton consonant (three words).

**TABLE 3. Syllable shapes produced by S1 and S2.**

| One-Syllable Words | Two-Syllable Words | Three-Syllable Words | Four-Syllable Words |
|---|---|---|---|
| VC | V/CV | V/CV/CV | CV/CV/CV/CV |
| VCC | V/CVC | V/CV/CVC[b] | CV/CV/CVC/CCVC[b] |
| CVC | V/CVCC[b] | V/CV/CVCC | CV/CV/CV/CCVC[a] |
| CVCC | VC/CVC[a] | CV/CV/CV | CVC/CVC/CV/CVC[b] |
| CVCCC[a] | VC/CVCC | CV/CV/CVC | CCVC/V/CV/CV[a] |
| CCV | CV/CV | CV/CV/CCVC | |
| CCVC | CV/CVC | CVC/CV/CVC | |
| CCVCC | CV/CCV | CVC/CVC/CCVC[a] | |
| | CVC/CV[b] | CVCC/CV/CVC[b] | |
| | CVC/CVC | CCV/CVC/CVC[b] | |
| | CVC/CVCC[b] | CCVC/CV/CV | |
| | CVC/CCVC | CCVC/CV/CVC | |
| | CVCC/CVC[b] | CCCVC/V/CVC | |
| | CCV/CV[a] | | |
| | CCV/CVC | | |
| | CCVC/CVC | | |
| | CCVC/CCVC[a] | | |
| | CCCV/CVC | | |

*Note.* V = vowel, C = consonant, / = syllable boundary.
[a]Not produced by S1. [b]Not produced by S2.

### *Error Rate*

Error rate was calculated on the basis of a sound-by-sound comparison of the children's actual productions with the target words. Any production of a sound that was not consistent with the standard realization or with the local dialect, or any nonproduction of a sound, was considered an error. In S1, an overall error rate of 44.2% (299/677) was found. Consonants were more often affected than vowels and diphthongs, with error rates of 62.6% (273/436) and 10.8% (26/241) respectively. In S2, the overall error rate was 45.2% (280/620). Once again, consonants were affected more often (57.8%; 234/405) than vowels and diphthongs (21.4%; 46/215).

### *Sounds Affected*

Next, the speech samples were examined for the specific Dutch consonants, vowels, and diphthongs affected most often in each subject. Table 4 shows the results for consonant sounds. Mispronunciations occurred with all consonants in S1 and with nearly all consonants in S2. The only exception was the /j/, which S2 produced correctly in all instances. No correct productions were found for /m/ and /ʒ/ in S1 and for /b/, /w/, /f/, /v/, /d/, and /ʃ/ in S2. Data seen in Table 4 suggests that the error rate was related to place of articulation of the target sounds attempted. In general, error percentages tended to be higher in consonants with a more forward place of articulation than in consonants with place of articulation farther back in the oral cavity. In order to further test this finding, error percentages were calculated for the different

**TABLE 4. Error rate for consonant sounds of S1 and S2.**

| | S1 | | S2 | |
|---|---|---|---|---|
| Sound | % Errors | # Errors/ Trials | % Errors | # Errors/ Trials |
| p | 45 | 10/22 | 25 | 7/28 |
| b | 58 | 11/19 | 100 | 8/8 |
| m | 100 | 16/16 | 68 | 13/19 |
| w | 67 | 8/12 | 100 | 12/12 |
| f | 73 | 8/11 | 100 | 12/12 |
| v | 75 | 9/12 | 100 | 7/7 |
| t | 70 | 44/63 | 73 | 43/59 |
| d | 43 | 6/14 | 100 | 9/9 |
| n | 76 | 37/49 | 53 | 16/30 |
| s | 80 | 33/41 | 83 | 49/59 |
| z | 78 | 7/9 | 50 | 1/2 |
| l | 49 | 23/47 | 46 | 22/48 |
| ʃ | 75 | 3/4 | 100 | 1/1 |
| ʒ | 100 | 1/1 | * | * |
| j | 40 | 2/5 | 0 | 0/4 |
| k | 35 | 11/31 | 17 | 5/29 |
| ɣ | 45 | 5/11 | 50 | 2/4 |
| x | 33 | 3/9 | 20 | 4/20 |
| ŋ | 25 | 1/4 | 50 | 2/4 |
| ʀ | 62 | 35/56 | 42 | 21/50 |

*S2 had no target words containing ʒ.

sound classes according to place of articulation.

As the results in Figure 1 show, error percentages indeed varied according to place of articulation, with two groups of sounds emerging. Definitely lower error percentages were seen in palatal, velar, and uvular sounds than in bilabial, labiodental, alveolar, and prepalatal sounds. The only exception was with the uvular sounds (actually the sound /ʀ/) in S1, for which an error percentage similar to that of bilabial sounds was found. In S1, error percentages for palatal, velar, and uvular sounds were 40%, 36%, and 62.5% respectively, and error percentages for bilabial, labiodental, alveolar, and prepalatal sounds were 65.2%, 73.9%, 67.3%, and 75% respectively. In S2, error percentages for palatal, velar, and uvular sounds were 0%, 22.8%, and 42% respectively and for bilabial, labiodental, alveolar, and prepalatal sounds were 59.7%, 100%, 67.6%, and 66.7% respectively. A statistical analysis (chi-square) comparing error rate of the group of palatal, velar, and uvular sounds (in S1: 57/116 or 49.1%; in S2: 34/111 or 30.6%) versus error rate of the group of bilabial, labiodental, alveolar, and prepalatal sounds (in S1: 216/320 or 67.5%; in S2: 200/294 or 68%) revealed a significant difference between these two groups in both subjects [S1: $\chi^2(1, N = 436) = 12.26$, $p < .0005$; S2: $\chi^2(1, N = 405) = 46.19$, $p < .00001$].

Table 5 shows the error percentages for vowels and diphthongs. Diphthongs were generally produced correctly in both subjects. For vowels, the error pattern appeared to be opposite to that found for consonants. That is, both subjects showed a tendency to produce more errors in vowels with a backward place of articulation (in S1, 16/48 or 33.3%; in S2, 34/48 or 70.8% error rate). The error rates were considerably lower for front and central vowels (in S1: front vowels, 3/65 or 4.6%, and central vowels, 6/110 or 5.5%; in S2: front vowels, 6/59 or 10.2%, and central vowels, 6/93 or 6.5%). Statistical analysis (chi square) confirmed the existence of a significant difference between the three types of vowels in either subject [S1: $\chi^2(2, N = 223) = 30.10$, $p < .00001$; S2: $\chi^2(2, N = 200) = 81.88$, $p < .001$]. One feature that the back vowels in Dutch also have in common, in addition to their place of articulation, is that they are all rounded. It could be that the higher error percentages in these vowels were related to the feature of

**FIGURE 1. Error percentages in S1 and S2 for the different classes of consonant sounds according to place of articulation (S2 produced no errors in palatal sounds).**

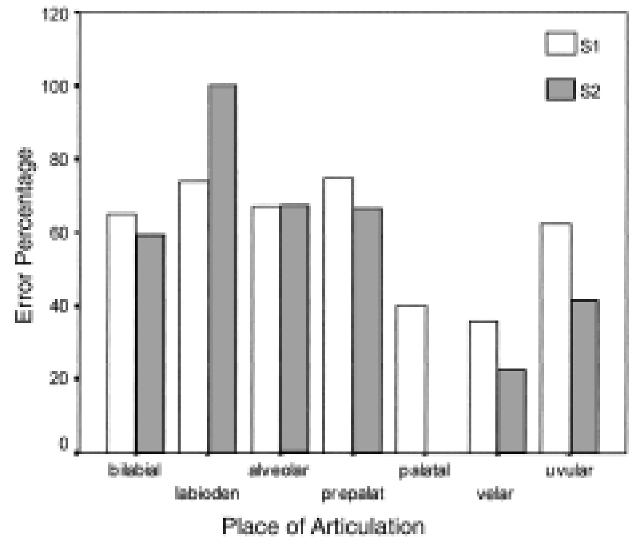


**TABLE 5. Error rate for vowels and diphthongs of S1 and S2.**

| | S1 | | S2 | |
|---|---|---|---|---|
| Sound | % Errors | # Errors/ Trials | % Errors | # Errors/ Trials |
| i | 5 | 1/20 | 12 | 2/17 |
| y | 0 | 0/3 | 33 | 1/3 |
| e | 0 | 0/18 | 7 | 1/15 |
| ɛ | 0 | 0/14 | 13 | 2/15 |
| ø | 33 | 1/3 | 0 | 0/3 |
| ɪ | 14 | 1/7 | 0 | 0/6 |
| ə | 4 | 2/54 | 10 | 4/39 |
| œ | 50 | 1/2 | 50 | 1/2 |
| a | 14 | 2/14 | 0 | 0/16 |
| ɑ | 2.5 | 1/40 | 3 | 1/36 |
| u | 33 | 3/9 | 89 | 8/9 |
| o | 33 | 6/18 | 71 | 12/17 |
| ɔ | 33 | 7/21 | 64 | 14/22 |
| ɛi | 0 | 0/8 | 0 | 0/6 |
| œy | 14 | 1/7 | 0 | 0/7 |
| ɔu | 0 | 0/3 | 0 | 0/12 |

rounding rather than to place of articulation. The finding that the rounded front vowels /y/ (in S2) and /ø/ (in S1) and the rounded central vowel /œ/ (in both subjects) were often in error supports this interpretation. In any event, the error rate for the group of rounded vowels (/y/, /ø/, /œ/, /u/, /o/, and /ɔ/) was higher than that for the group of unrounded vowels (/i/, /e/, /ɛ/, /ɪ/, /ə/, /a/, and /ɑ/) in S1 (viz., 18/56 or 32.1% vs. 7/167 or 4.2%) as well as in S2 (viz., 36/56 or 64.3% vs. 10/144 or 6.9%). The difference proved significant [chi square, S1: $\chi^2(1, N = 223) = 32.91$, $p < .00001$; S2: $\chi^2(1, N = 200) = 74.86$, $p < .0001$]. Additional support for the view that rounding might have been the determining feature came from the analysis into the error patterns to be presented in another section.

### *Omissions, Substitutions, and Distortions*

In a subsequent analysis, the errors against target consonant sounds were classified according to the traditional categories of omission, substitution, and distortion. An omission was defined as nonproduction of a target sound, a substitution was a deviant (i.e., nonstandard) production of a target sound crossing phoneme boundaries, and a distortion was a deviant production of a target sound not crossing phoneme boundaries. Instances of substitution in combination with distortion were classified along with substitutions. It was found that in both subjects, distortions were the predominant error type. In S1, distortions accounted for 97.8% (267/273) of consonant errors, substitutions constituted only 2.2% (6/273) of consonant errors, and omissions did not occur. In S2, distortions accounted for 86.3% (202/234) of consonant errors, substitutions for 10.7% (25/234), and omissions for 3% (7/234).

### *Error Patterns*

*Consonants.* A further analysis into the nature of the subjects' articulation errors showed that a majority of their erroneous consonant productions fell into distinct patterns. A first pattern that was most evident in bilabial consonants (/b/, /p/, /m/, /w/) was that of linguolabialization. In this pattern, a closure was made between the tongue and both lips rather than between both lips. A similar visible protrusion of the tongue between the lips was also observed a number of times in the production of alveolar, prepalatal, and velar sounds. A second pattern occurred with labiodental and alveolar sounds and consisted of linguodentalization. Instead of contact between the lower lip and upper incisors, or between the tongue tip and the alveolar ridge, contact was made between the tongue and the upper incisors. A third pattern emerged for the velar and uvular sounds. These sounds were often produced with the tongue resting on the lower lip. A pattern affecting bilabial sounds only was inversion of the lips in which a bilabial closure was made with an inward movement of both upper and lower lip. In the consonants /w/ and /ʒ/, which are both produced with lip rounding in Dutch, a tendency to unround was observed in some instances. Together, the patterns of linguolabialization, linguodentalization, the production of velars and uvulars with the tongue resting on the lower lip, inversion in bilabials, and unrounding in /w/ and /ʒ/ accounted for 90.5% of all consonant errors in S1 and for 72.6% of errors in S2.

Other error types were seen only in one subject and not in the other and/or occurred only occasionally in a limited number of instances. An error type observed with some frequency in S1 but not in S2 was production of a consonant with visible tongue thrusting against the inner wall of the lower lip. Sounds in which this erroneous pronunciation was seen were the alveolars /t/, /n/, /z/, and /l/, the palatal /j/, and the uvular /ʀ/. In S2, but not in S1, there was a tendency to produce some consonants with protrusion of the mandible including the /t/, /n/, /s/, /l/, /ʃ/ and /ʀ/. Among the rare error types encountered were weak articulation of certain sounds (both subjects), labiodentalization of a bilabial or alveolar sound (S1), devoicing of voiced consonants (both subjects), slight backing of some alveolars (S2), retroflex articulation of /t/ (S2), unreleased production of stops (S2), and derhotacizing (loss of trill) of the /ʀ/ (S2).

*Vowels.* In vowels, the predominant error pattern was unrounding, which accounted for 52% of all vowel errors in S1 and 61% in S2. Apart from a few instances of tongue thrusting against the inner wall of the lower lip in S1 and a few examples of weak articulation in S2, other error types occurred only once or twice.

## Discussion

A number of investigators have pointed out the existence of articulation problems in individuals with BWS, presumably due to macroglossia. In particular, labiodental, linguodental, and alveolar consonant sounds have been reported to be problematic in BWS (McManamny & Barnett, 1985; Mixter et al., 1993; Van Borsel et al., 1999). In one study (Mixter et al., 1993), vowel distortions were observed as well. Both subjects in the present study also displayed articulation errors. In S1, the older subject (age 5;9), 62.6% of consonants and 10.8% of vowels and diphthongs were affected. In S2, the younger subject (age 3;6), error rates of 57.8% for consonants and 21.4% for vowels and diphthongs were found. Occurrence of such a high number of errors at age 5;9 is certainly not normal. In the case of S2, it could be argued that the presence of some misarticulations is not uncommon at that age. However, it should be noted that the frequency of errors is fairly high. A study of speech acquisition in Dutch shows that 75% of children master all vowels and diphthongs by age 3 and 75% master singleton consonants by age 3;6 (Stes, 1977). Moreover, the analyses of the sounds affected and the nature of the misarticulations make it very difficult to dispose of the errors of S2 as purely common developmental ones. Analysis of the sounds involved revealed that, although errors occurred with all consonants in S1 and nearly all consonants in S2, certain consonants were more prone to misarticulation than others. In line with previous studies discussing articulation in BWS, it was found that consonants with a more forward place of articulation (bilabial, labiodental, alveolar, and palatal sounds) were most likely to be affected. In S1, the velar /ʀ/ also had a high error percentage. As the analysis into error patterns showed, the /ʀ/ was often produced with the tongue resting on the lower lip. The fact that no tongue activity is required for the production of /ʀ/ and that the tongue can rest on the lower lip may explain the difference in error rate between the /ʀ/ and the other back consonants (palatal and velar sounds). Analysis of vowels and diphthongs showed that particularly rounded vowels were subject to mispronunciation.

Relative to the nature of the articulation errors, it was found that over 90% of consonant errors of S1 and more than 72% of consonant errors of S2 fell into one of five error patterns of inappropriate tongue or lip posture: linguolabialization, linguodentalization, lip inversion, production of a sound with the tongue resting on the lower lip, and unrounding. The latter pattern also accounted for more than half of the subjects' vowel errors (52% in S1, 61.2% in S2).

The nature of the articulation patterns further suggested that, to a large extent, the misarticulations were the direct result of the presenting macroglossia. The overly large tongue apparently precludes contact between two articulators resulting in linguolabialization in bilabial sounds and in linguodentalization in labiodental sounds. The tongue in our subjects was also too large to allow contact with the appropriate articulator, so linguodentalization occurred with alveolar sounds. The oversized tongue was also too large for the subjects to keep in the mouth during the production of velar and uvular sounds, so instead the tongue rested on the lower lip. Also, the failure to appropriately round the lips in both consonants and vowels can be considered the direct result of the macroglossia. In case of inversion of the lips, as seen sometimes in bilabial sounds, this pattern might be explained as an attempt to close the lips in spite of the overly large tongue.

Results of the phonetic inventory analysis showed that, despite the high overall error rate, both subjects were apparently capable of producing most of the sounds of their mother tongue. The vowel and diphthong inventory was nearly complete in both subjects. In S1, the consonant inventory was relatively complete with only the /m/ and /ʃ/ being absent. In S2, the consonant inventory was less complete, lacking the /b/, /w/, /f/, /v/, and /d/. It is not quite clear how the difference in completeness of the consonant inventory in S1 and S2 can be explained. It could reflect intersubject variability, but it could also be ascribed to the fact that S2 was still younger. It may be that further spontaneous regression of the macroglossia will lead to better accommodation of the tongue in the oral cavity as the child grows older. The results of the inventory analyses are noteworthy and seem to indicate that the subjects were capable of articulating better than they usually did. The finding that there were no distributional constraints in the subjects' consonant inventories, along with the finding that the number of syllables and the syllable shapes of target words were nearly always retained and that the predominant error type was distortion of sounds, suggest that the disorder emanates from a phonetic rather than a phonological problem. As such, these results are in line with the hypothesis that the misarticulations are the result of the presenting macroglossia.

# Study 2

An interesting observation noted while phonetically transcribing the subjects' speech samples in Study 1 was that, in spite of numerous speech errors, both subjects had speech that was still highly intelligible. As well, the impression arose that their articulation was far better auditorily than visually in terms of acceptability. To test this hypothesis, an observer experiment was designed in which a panel of judges was asked to rate a speech fragment of each subject from three different modes of presentation: auditory-only presentation (only hearing a subject's speech), visual-only presentation (only seeing a subject's speech) and audiovisual presentation (hearing and seeing a subject's speech). Since we had the impression that the subjects' articulation was more acceptable auditorily than visually, it was hypothesized that articulation would be judged better in the auditory-only mode than in the audiovisual mode and better in the audiovisual mode than in the visual-only mode.

## Method

### *Panel*

Participants in the experiment were a panel of 34 individuals, 24 of whom were naive observers without any training or experience in speech-language pathology and 10 of whom were certified speech-language pathologists. Age of the participants ranged from 22 to 33 years. Twenty-two of them were female and 12 were male.

### *Speech Samples*

The samples used in the experiment were selected from the recorded data of Study 1. For each subject (S1 and S2), a sample was selected of 30 consecutive words uttered in response to the stimulus pictures they were shown. In each case, the first 30 words were chosen that the subject uttered readily without necessary interference from the examiner. In both subjects, the sample contained polysyllabic words as well as monosyllabic words with singleton consonants and consonant clusters. For presentation to the panel of judges, the samples of both subjects were dubbed from the original videotape onto a master videotape. As the stimulus pictures had been randomized (see data collection in Study 1), no further randomization was made of the 30 words selected.

**TABLE 6. Design of the observer experiment.**

| Panel | S1 | S2 |
|---|---|---|
| Group 1 | Auditory-only mode<br>Audiovisual mode | Visual-only mode<br>Audiovisual mode |
| Group 2 | Visual-only mode<br>Audiovisual mode | Auditory-only mode<br>Audiovisual mode |

## Design

The experiment was designed to control for possible order or sequence effects. The 34 participants were randomly divided into two groups, each consisting of 12 naive observers and 5 speech-language pathologists. Each group rated the samples of both subjects in two modes (see Table 6). Group 1 rated articulation of S1 from the auditory-only mode and the audiovisual mode and rated articulation of S2 from the visual-only mode and the audiovisual mode (in that order). Group 2 rated articulation of S2 from the auditory-only mode and the audiovisual mode and rated articulation of S1 from the visual-only mode and the audiovisual mode (in that order). For presentation of the samples, an ordinary TV set was used. Judges were told that they were going to see speech samples, which they would have to look at and listen to carefully, then score for articulation. They were required to rate the speech samples on a 10-point equal-interval scale with 0 as unacceptable, severely disordered articulation, and 10 as perfect articulation. Each sample was presented only once in each presentation mode and was scored immediately after presentation, with a separate scoring sheet for each presentation. During the entire experiment, judges had a list of the target words of each subject's sample at their disposal. Before the experiment started, judges were given the opportunity to request further clarification of the task required. During the experiment, the instruction was repeated again before each presentation. Judges were not told, however, that the purpose was to compare the rating from the different modes of presentation.

**TABLE 7. Comparison of average scores for the modes of presentation.**

| Mode of Presentation | S1 | S2 |
|---|---|---|
| Auditory-only | 7.3 | 6.4 |
| Audiovisual | 5.9 | 4.4 |
| Audiovisual | 6.8 | 4.5 |
| Visual-only | 5.1 | 3.6 |

**TABLE 8. Comparison of the mean ratings of the naive observers vs. speech-language pathologists.**

| Group | Mode of Presentation | Naive Observers | Speech-Language Pathologist Observers | *p* value |
|---|---|---|---|---|
| 1 | Auditory-only S1 | 7.3 | 7.4 | .7366 |
| | Audiovisual S1 | 6.1 | 5.6 | .5145 |
| | Visual-only S2 | 3.6 | 3.6 | 1.0000 |
| | Audiovisual S2 | 4.4 | 4.8 | .3838 |
| 2 | Auditory-only S2 | 5.8 | 7.8 | .0278 |
| | Audiovisual S2 | 4.4 | 4.6 | .5827 |
| | Visual-only S1 | 5.2 | 5.0 | .5886 |
| | Audiovisual S1 | 6.7 | 7.0 | .7886 |

*Note.* The rating scale was 0–10 (low–high).

## Results

Average scores for each mode of presentation are shown in Table 7. In both subjects, ratings from the auditory-only presentation were significantly higher than ratings from the audiovisual presentation (Wilcoxon matched-pairs signed-ranks test; S1: $Z = -3.26$, $p = .0011$; S2: $Z = -3.43$, $p = .0006$). And in both subjects, ratings from the audiovisual presentation were significantly higher than ratings from the visual-only presentation (Wilcoxon matched-pairs signed-ranks test; S1: $Z = -3.27$, $p = .0011$; S2: $Z = -2.00$, $p = .0452$).

Results of a comparison of the ratings for each subject by the naive observers versus ratings by the speech-language pathologists and for each mode of presentation are summarized in Table 8. A significant difference (Mann-Whitney U–Wilcoxon rank sum W test; $U = 9.5$, $p = .0278$) was found for the auditory-only presentation of S2 only, with ratings of the speech-language pathologists higher than ratings of the naive judges.

## Discussion

In a sample survey study (questionnaires filled out by parents) involving 40 patients with BWS, Van Borsel et al. (1999) found that the number of adverse clinical effects of macroglossia did not differ between individuals in whom macroglossia had been corrected surgically and individuals who had no tongue surgery. Also, the number of children who had been receiving speech treatment in the group with uncorrected macroglossia was not significantly larger than that in the group who had undergone a partial tongue reduction. Similar findings were reported by Weng, Moeschler, and Graham (1995) in a follow-up study of 15 children with BWS. Four of these children had speech treatment, but the need for treatment did not correspond to whether or not they had macroglossia reduction surgery. It is not clear whether such findings mean that surgical correction of macroglossia in BWS is ineffective for preventing or correcting speech defect. It is possible that the majority of BWS subjects with a surgically corrected macroglossia represent the patients in whom the macroglossia was originally severe, and that a lack of surgical intervention would have resulted in more severe articulation problems. It cannot be ruled out that, in some cases, surgical intervention itself represents, in part, the iatrogenic origin of persistent speech problems. After a glossectomy, tongue movements may have become more difficult due to scarring and ankylosis.

The results from the present observer experiment showed that, notwithstanding the above findings, a partial glossectomy may be considered a worthwhile option for improving articulation in BWS. In both subjects, neither of whom had undergone partial tongue reduction for correcting macroglossia, lay observers as well as experts judged articulation to be more disturbed visually than auditorily. In both subjects, ratings from the auditory-only presentation of a speech sample were significantly higher than ratings from the audiovisual presentation, and these in turn were significantly higher than ratings from the visual-only mode. Moreover, ratings from the lay observers and those from experts did not significantly differ. Only in the auditory-only mode in S2 was there significantly higher scores given by the speech-language pathologists than given by the naive judges. This may be related to the higher exposure of speech-language pathologists to more severe articulation disorders than lay persons. Because a partial glossectomy can substantially improve visual appearance of speech, such surgical intervention may be indicated. In this respect, we should point out that 40% of the respondents in the study of Van Borsel et al. (1999) indicated their children had been perceived by others as mentally retarded. As well, perception of mental retardation is one of the clinical manifestations listed by Vogel et al. (1986) as typical of macroglossia. In addition, when parents in the study by Weng, Moeschler, et al. (1995) were questioned about their child's peer interactions, 38% reported that their child had been teased because of their macroglossia and 63% reported difficulties with either adult or peer interactions.

## Conclusions

The combined results of Study 1 and Study 2 tempt one to conclude that the macroglossia in BWS leads to a distinct articulation pattern. This pattern is characterized by inappropriate tongue and lip postures that predominantly affect consonants with an anterior place of articulation and vowels and diphthongs with rounding and seems to disturb acceptability of articulation visually more than auditorily.

Since this work included only two subjects, the results of this study must be interpreted with caution. Replication of the study with a larger group of subjects is needed to confirm the existence of a distinct articulation pattern associated with BWS. In this respect, it would be extremely helpful to gather cross-linguistic information. Perhaps patterns similar to the ones outlined here may be found in children with different linguistic backgrounds. If so, this would confirm the hypothesis that the problems are caused by the physical mechanism of macroglossia. Further insight into the nature and origin of the articulation patterns in BWS may also be gained from comparisons between subjects who underwent tongue reduction surgery and subjects in whom the macroglossia remained untreated. Also, a comparison of articulation pre- and postoperatively may yield interesting findings regarding the nature and origin of the articulation patterns in BWS, as well as the study of longitudinal data from untreated subjects.

Nonetheless, one can draw from this study some practical conclusions. The finding that the visual aspect of speech in both subjects was judged to be more detrimental to communication than the actual production stresses the importance of not limiting the assessment of speech in disordered children to an auditory-only evaluation. Particularly in children with craniofacial anomalies, such as macroglossia in BWS, it is at least as important to assess the visual aspect of speech. Analogously, treatment should not be directed exclusively at improving speech in its auditory characteristics. Finally, the finding that both subjects of the present study, although untreated for articulation, were apparently capable of correctly forming most of the sounds of their mother tongue suggests that treatment efforts aimed at improving articulation in individuals with BWS should be attempted and that hope for treatment success is warranted.

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## Appendix

The Phonology of Dutch

### Dutch Vowel Inventory

| | Front Vowels | | Central Vowels | | Back Vowels | |
|---|---|---|---|---|---|---|
| | Unrounded | Rounded | Unrounded | Rounded | Unrounded | Rounded |
| Closed | i | y | | | | u |
| Half closed | e, ɪ | ø | | | | o |
| Half open | ɛ | | ə | œ | | ɔ |
| Open | | | a, ɑ | | | |

### Dutch Diphthong Inventory

ɛi, œy, ɔu

### Dutch Consonant Inventory

| | bilabial | | labiodental | | alveolar | | prepalatal | | palatal | | velar | | uvular | | glottal | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | –V | +V | –V | +V | –V | +V | –V | +V | –V | +V | –V | +V | –V | +V | –V | +V |
| plosive | p | b | | | t | d | | | | | k | | | | | |
| fricative | | | f | v | s | z | ʃ | ʒ | | | x | ɣ | | | | h |
| nasal | | m | | | | n | | | | | | ŋ | | | | |
| liquid | | | | | l | r | | | | | | | | ʀ | | |
| glide | | w | | | | | | | | j | | | | | | |

*Note.* The phoneme /r/ in Dutch is subject to regional and individual variation and may either be realized as an alveolar roll [r] or as a uvular roll [ʀ]. Both subjects of the present study used the uvular variant.
The /h/ in Dutch is a voiced consonant articulated with glottal frication and was not considered further in the analysis. The shape of the vocal tract with this sound is actually determined by the shape of the following vowel.

### Distribution of Dutch Consonants

| Syllable Position | p | b | t | d | k | f | v | s | z | ʃ | ʒ | x | ɣ | m | n | ŋ | l | ʀ/r | j | w |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Initial | + | + | + | + | + | + | + | + | + | + | + | + | + | + | + | – | + | + | + | + |
| Final | + | – | + | – | + | + | – | + | – | – | – | + | – | + | + | + | + | + | + | + |

*Note.* Based on Blancquaert (1969), Booij (1995), and Van Den Berg (1978). +V = voiced, –V = voiceless; + indicates that the sound occurs in that position, – indicates that the sound does not occur in that position.