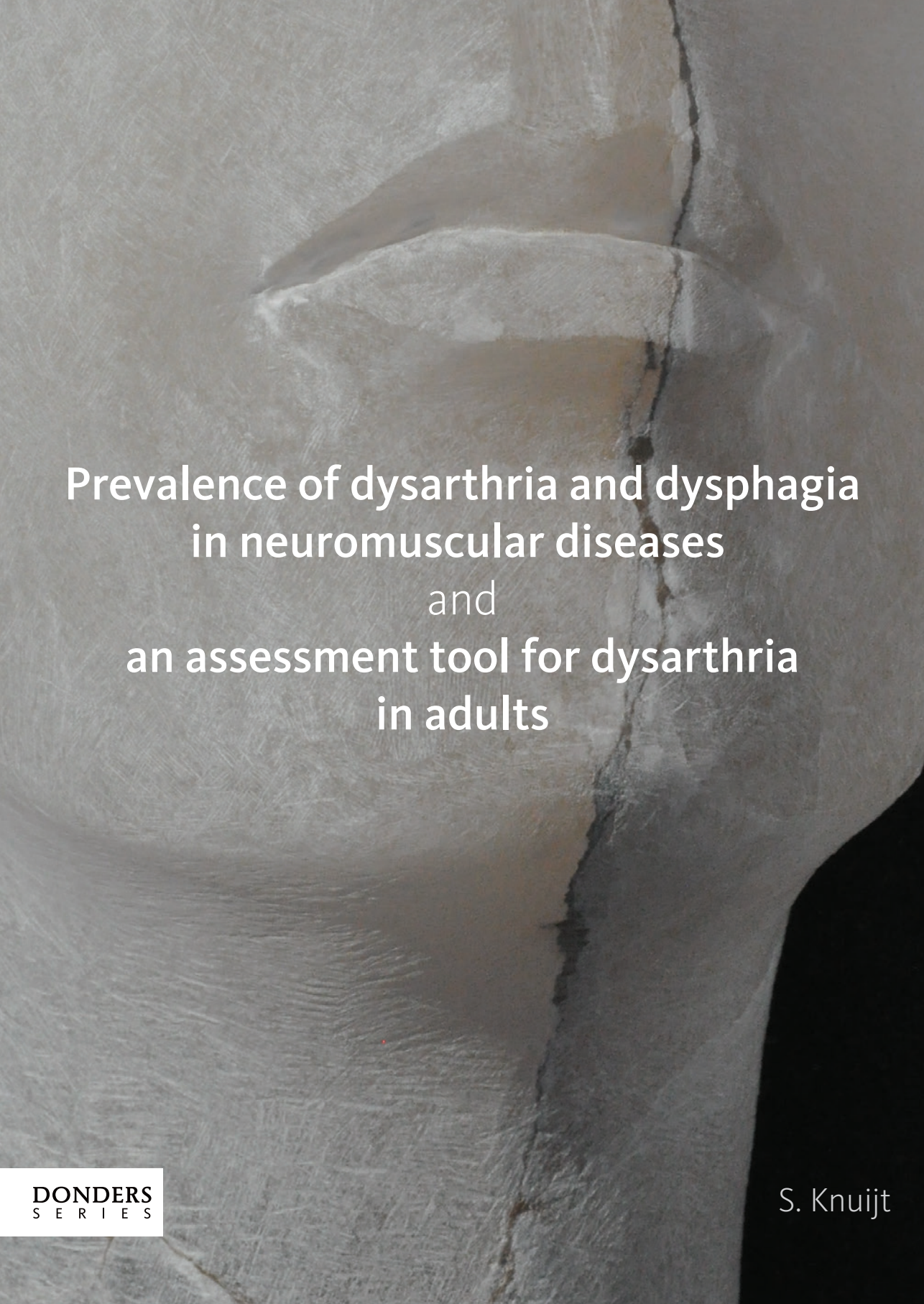

Prevalence of dysarthria and dysphagia in neuromuscular diseases and an assessment tool for dysarthria in adults

DONDERS SERIES

S. Knuijt

Prevalence of dysarthria and dysphagia in neuromuscular diseases
and
an assessment tool for dysarthria in adults

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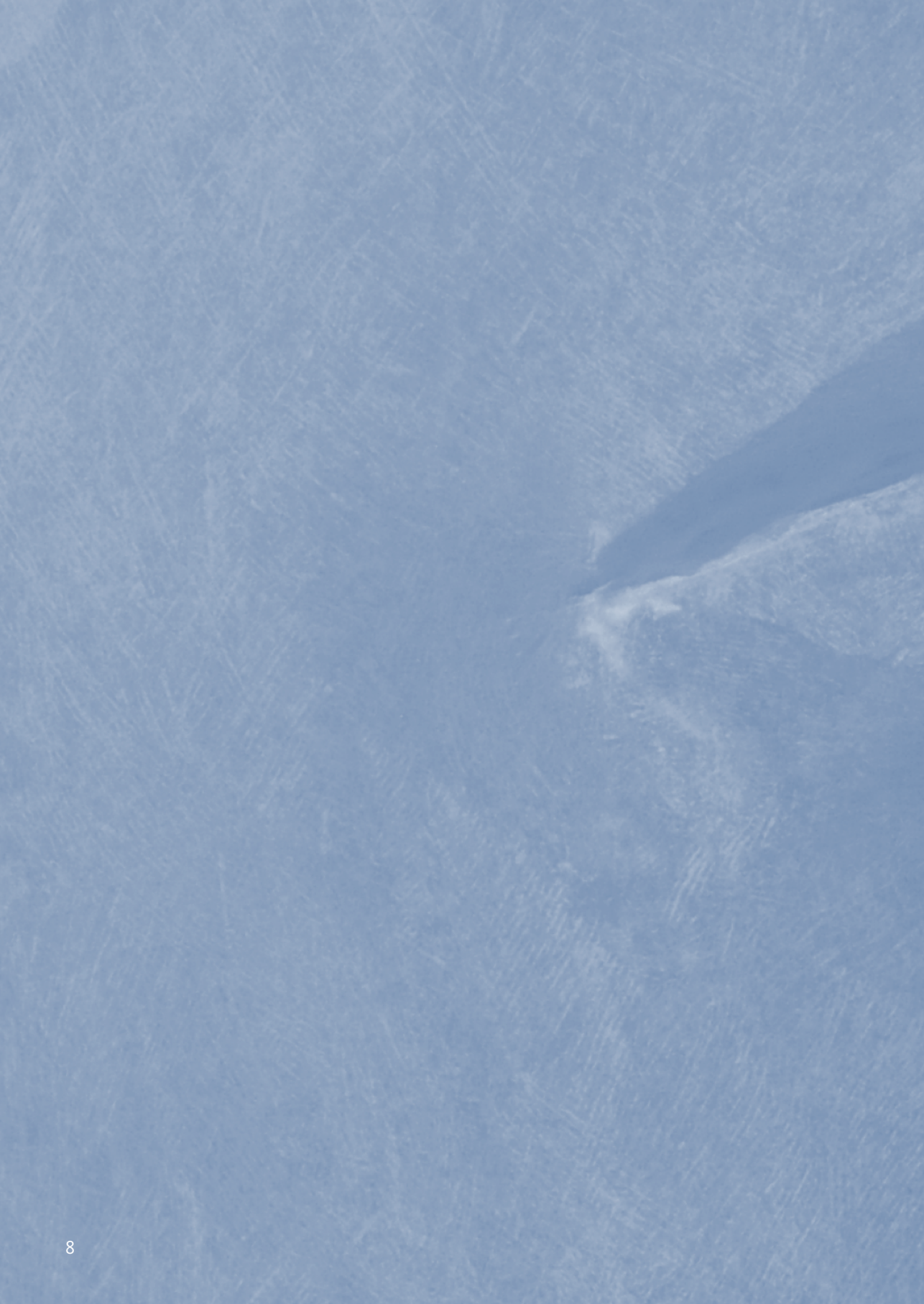


1 General introduction and outline of the thesis

SPEECH AND DYSARTHRIA

Speech is unique for humans and, despite the rise of social media, daily communication is highly dependent on spoken language. Speech is important for any kind of communication: from a casual chat to business-like conversations, and it is part of every social role one plays, e.g. being a friend, a partner or a colleague. Therefore, difficulties with speech can directly limit daily activities and social participation, which has a negative impact on quality of life.[1-5] An illustration of the impact of a speech disorder is given in Box 1.1.

When a neurological disease affects speech, this is called dysarthria. The word 'dysarthria' is derived from Latin, and means 'problematic' (*dys*) – 'articulation' (*arthron*). Duffy defined dysarthria as "a collective name for a group of neurologic speech disorders resulting from abnormalities in the strength, speed, range, steadiness, tone, or accuracy of movements required for control of the respiratory, phonatory, resonatory, articulatory, and prosodic aspects of speech production".[6]

Dysarthria can be caused by (sub-)acute central neurological, central neurodegenerative or peripheral neuromuscular diseases. The evolution of dysarthria depends on its etiology. (Sub-)acute central neurological diseases like stroke are generally associated with some level of recovery, whereas neurodegenerative and neuromuscular diseases are associated with more or less progressive dysarthria. Some well-known progressive dysarthrias are associated with, for instance, Parkinson's disease and amyotrophic lateral sclerosis.[7,8] Dysarthria may also result from neuromuscular diseases (NMD), which are relatively rare progressive diseases of the peripheral nervous system. More than 600 NMD have been identified, each with a low incidence. Table 1.1 provides an overview of neurological diseases in each category that can cause dysarthria.

In 1969, Darley, Aronson and Brown[9] were the first who listed 38 speech characteristics within the five aspects of speech production (i.e., articulation, resonance, phonation, respiration, and prosody), that can lead to 6 different types of dysarthria: the so-called DAB-classification – named after the authors. They showed that there was an association between the site of the lesion and the type of dysarthria. The types were named after their main speech characteristic: 'flaccid', 'spastic', 'ataxic', 'hypokinetic', 'hyperkinetic', and 'mixed'. This classifica-

Box 1.1. Illustration of the impact of a speech disorder on daily life

A 69-year-old man, suffers from dysarthria due to spinocerebellar ataxia. The dysarthria was the first symptom of his cerebellar disorder. He noticed that his speech rate slowed down and that he tripped over words more often. He had retired since a couple of years, but he was always busy doing voluntary work as a guide in the surroundings of his hometown. Even when his dysarthria was still mild, he had to stop almost all his voluntary work, because he was often unintelligible for strangers, especially during environmental noise. Gradually, staying intelligible in a noisy environment became a problem also during social events like birthday parties. He withdrew himself in silence while being in a group of people. He did not only struggle in the presence of strangers and when being in groups. Also at home there were growing irritations. His wife felt that he did not try hard enough to speak well, but speaking intelligible was very hard for him. Especially at home he did not want to have to pretend he was better than he really was, something he already had to do amongst others.

Table 1.1. Neurological diseases, categorized by localization, causing different types of dysarthria.

Localization	Disease	Type of dysarthria
(Sub-)acute central neurological		
Unilateral	Stroke, brain trauma, brain tumor, meningitis, encephalitis	Unilateral upper motor neuron
Bilateral	Stroke, brain trauma, brain tumor, meningitis, encephalitis	Spastic
Central neurodegenerative		
Extrapyramidal	Parkinson's disease Huntington's disease	Hypokinetic Hyperkinetic
Cerebellar	Cerebellar ataxias (e.g., spinocerebellar ataxia or autosomal dominant cerebellar ataxia)	Ataxic
Multiple localizations	Progressive supranuclear palsy, multiple system atrophy, multiple sclerosis, amyotrophic lateral sclerosis	Mixed (types dependent on site of lesion)
Peripheral neuromuscular		
Peripheral motor neuron	Spinal muscular atrophy	Flaccid
Peripheral nerve	Guillain-Barré, peripheral facial paralysis, hypoglossal paresis	Flaccid
Motor endplate/ neuromuscular junction	Myasthenia gravis, Lambert-Eaton myasthenic syndrome	Flaccid
Muscle	Facioscapulohumeral muscular dystrophy, myotonic dystrophy, Duchenne muscular dystrophy, polymyositis, mitochondrial myopathy	Flaccid

tion is still used in clinical practice. Later, Duffy added the 'unilateral upper motor neuron' (UUMN) dysarthria to this classification, because he argued that the UUMN dysarthria was a distinct type which could not be placed in the DAB-classification.[6] In Table 1.1, the different types of dysarthria that are associated with the different neurological diseases are summarized.

In the first decades after the introduction of the DAB-classification, speech-language therapists in the Netherlands were accustomed to use the lesion site to name the dysarthrias. 'Flaccid' was named 'bulbar', 'spastic' was named 'pseudobulbar', 'ataxic' was named 'cerebellar' and both 'hypokinetic' and 'hyperkinetic' dysarthrias were named 'extrapyramidal'. However, this anatomical classification has important limitations. For example, all flaccid dysarthrias were named 'bulbar', but a flaccid dysarthria due to a neuromuscular disorder is not caused by a bulbar lesion. And the hypo- and hyperkinetic dysarthrias, both being extrapyramidal disorders, are two totally different types of dysarthria. Therefore, about 15 years ago, our department decided to use the original DAB-classification consistently

Table 1.2. Different types of dysarthria with their most characteristic speech deficits, based on Darley, Aronson and Brown,[9] Duffy[6] and on clinical experience at the Radboud University Medical Center.

	Flaccid	Spastic	Ataxic	Hypokinetic	Hyperkinetic	Unilateral Upper Motor Neuron
Articulation	imprecise consonants due to weakness	slow, tensed	imprecise consonants due to coordination problems, distorted vowels	imprecise consonants due to small articulation movements, starting problems	irregular articulatory breakdowns, imprecise consonants, distorted vowels	imprecise articulation
Resonance	hypernasality	hypo- or hypernasality	variable	normal	hypernasality	
Phonation	hypotonic vocal use, breathy, reduced loudness	hypertonic vocal use, strained-strangled vocal quality	variable	hypertonic vocal use, breathy vocal quality, high pitch due to rigidity of the vocal folds	hypertonic vocal use, voice stoppages, strained-strangled vocal quality	breathy, harsh, reduced loudness
Respiration	short breath support, short breath groups	short breath groups	impaired breathing-voice coordination	shallow	short breath groups	
Prosody	monoloudness, monopitch	monoloudness, monopitch, slow rate	loudness variations, slow rate, monopitch	variable rate, monopitch, monoloudness	excess loudness variations, audible inspirations	slow rate

in our patient care, publications, and post-graduate dysarthria courses, in order to change the use of anatomical labels back to the DAB-classification.

Types of dysarthria

As mentioned earlier, the types of dysarthria are named after their main speech characteristic, with the exception of UUMN dysarthria that is actually named after the site of the lesion. An overview of the speech characteristics associated with the different types of dysarthria is depicted in Table 1.2. Information in this paragraph is based on worldwide accepted manuals by Darley, Aronson & Brown[9] and Duffy,[6] combined with the broad clinical experience of the speech therapists of our department.

In ***flaccid dysarthria***, weakness is the main characteristic. Weakness can be present in all aspects of speech production, leading to hypotonic articulation with hypernasality and hypotonic vocal use. Flaccid dysarthria is a characteristic feature of NMD, but it can also be part of mixed dysarthrias.

Spastic dysarthria results from bilateral damage to the upper motor neuron. Spasticity may also influence all aspects of speech production, but most of all speech will be slow with a hypertonic vocal use. Because only damage in both hemispheres causes spastic dysarthria, this dysarthria type is less frequent, but it can result from bilateral strokes, traumatic brain injury or cerebral palsy.

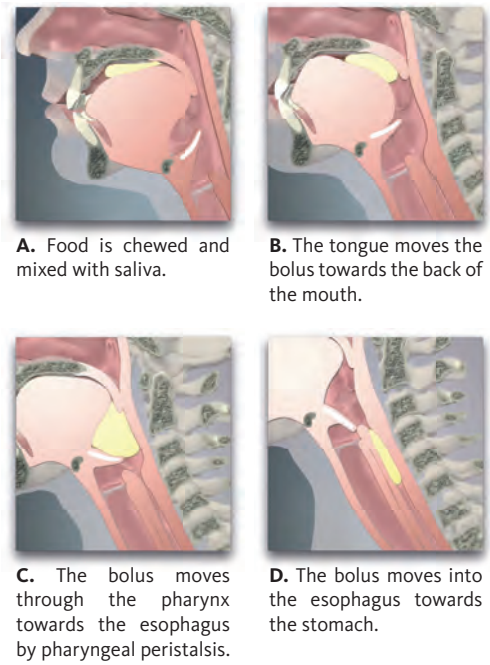
Ataxic dysarthria results from damage to the cerebellum, caused by for example degenerative diseases (cerebellar ataxias), demyelinating diseases (multiple sclerosis [MS]), stroke, and traumatic injury. The lack of coordination can be heard especially in articulation and prosody. Ataxic speech is often characterized as 'speaking like a drunk'.

Hypokinetic dysarthria is associated with lesions of the basal ganglia. The movement of the articulators and the vocal cords are small due to rigidity and slow due to bradykinesia, causing a soft voice with mumbling and monotonous speech. Especially phonation, articulation, and prosody will be affected in hypokinetic dysarthria, which is a typical feature of Parkinson's disease.

Hyperkinetic dysarthria is also associated with damage to the basal ganglia but, in contrast to hypokinetic dysarthria, involuntary movements occur. There are two types of hyperkinetic dysarthria: chorea (with unexpected involuntary movements) and dystonia (with increasing muscle tone during movements). In the case of chorea, the involuntary movements, leading to excessive loudness variations and sudden disruptions of articulation, will disrupt the speech. This type of dysarthria is generally associated with e.g. Huntington's disease, but can also be heard in people with Parkinson's disease and severe response fluctuations. In the case of dystonia (like in oromandibular dystonias), the increased muscle tone will lead to prolonged vowels, hypertonic vocal use, and almost normal articulation during periods in which muscle tone is not increased.

UUMN dysarthria results from unilateral damage to the upper motor neuron (UUMN), such as in unilateral stroke. In the Netherlands, the term UUMN dysarthria is not often used. In the acute phase of stroke, dysarthria will mostly be flaccid. In the chronic phase, ataxic or spastic components can be present.

Mixed dysarthrias are combinations of two or more different types of dysarthria. The underlying disease is characterized by damage to at least two parts of the nervous system. Some well-known diseases that may cause mixed dysarthria include amyotrophic lateral sclerosis (ALS), which



A. Food is chewed and mixed with saliva.

B. The tongue moves the bolus towards the back of the mouth.

C. The bolus moves through the pharynx towards the esophagus by pharyngeal peristalsis.

D. The bolus moves into the esophagus towards the stomach.

Figure 1.1. Swallowing process.

is caused by degeneration of both central and peripheral motor nerves and, therefore, will result in a combination of flaccid and spastic dysarthria. Second, MS caused by a demyelination of the central nervous system (i.e., spinal cord, brainstem, cerebellum, and cerebral hemispheres), most often causes ataxic dysarthria, combined with flaccid or spastic dysarthria. Finally, in multiple system atrophy (MSA), an atypical parkinsonism, damage to the basal ganglia and other systems (like brainstem and cerebellum) are often involved, resulting in hypokinetic dysarthria combined with ataxic, spastic or flaccid characteristics.[10] There are two subtypes: MSA-P with mainly parkinsonian features and MSA-C with mainly ataxic features.

Dysphagia

The term 'dysphagia' is derived from Greek and means 'problematic' (*dys*) – 'eating' (*phagein*). Swallowing is the process transporting (chewed) food and liquid from the mouth to the stomach. In Figure 1.1, the swallowing process is shown.

In this thesis, I will only focus on dysphagia due to NMD, because in NMD, dysarthria often co-occurs with dysphagia. Both disorders are caused by 'oromotor problems' within overlapping muscle groups.[7,11] Dysphagia in NMD is, similar to dysarthria, characterized by muscle weakness, which can influence the oral and pharyngeal stages of swallowing. Oral weakness can cause chewing problems and diminished bolus control or oral transport. Pharyngeal weakness can reduce pharyngeal constriction, resulting in residue of especially solid food.

Prevalence of dysarthria and dysphagia

Because a wide range of neurological diseases can cause dysarthria, it is probably one of the most prevalent neurological communication disorders, together with apraxia of speech and aphasia. For example, after stroke, dysarthria is present in approximately 40% of the patients.[12] Prevalence rates of other (sub-)acute central neurological diseases, like traumatic brain injury, are unknown. In neurodegenerative diseases, prevalence rates are higher, with 70% in Parkinson's disease[8,13] and approximately 44% in MS.[8] In the end stage of ALS, all patients will suffer from dysarthria or even anarthria,[7,14] and nearly all patients with Friedreich's ataxia will finally suffer from dysarthria.[15]

In neuromuscular diseases, prevalence rates of dysarthria are difficult to find, probably because NMD is a term that encompasses more than 600 different diseases, each with a low incidence (for an overview of NMD, see http://www.spierziekten.nl). In the literature, prevalence rates between 13% and 43% have been reported only for dysarthria in myasthenia gravis.[16,17] In summary, although the prevalence of dysarthria is

known in some well-known large disease groups, in many other diseases including NMD, prevalence rates are scarce.

Regarding dysphagia in NMD, there are some prevalence rates known. Willig *et al.* reported an overall prevalence rate of 35% in a group of 7 types of NMD (Duchenne muscular dystrophy, limb-girdle muscular dystrophy, facioscapulohumeral muscular dystrophy, spinal muscular atrophy, myotonic dystrophy, myasthenia gravis, and dermatomyositis/polymyositis).[11] In selected groups of patients with inclusion body myositis, a prevalence of 65% has been reported.[18] In myasthenia gravis, prevalence rates vary between 15% and 35%.[16,17]

Working at the Center of Expertise for Neuromuscular Diseases of the Radboud University Medical Centre, I felt that both dysarthria and dysphagia might occur in all kinds of NMD and co-occur more often than could be expected based on the literature. This discrepancy triggered to prospectively register all patients with NMD referred to a speech-language therapist and to review the literature for evidence regarding treatment options.

Assessment of dysarthria

Despite the widespread recognition of the different types of dysarthria, the actual assessment of dysarthria type is challenging for several reasons. First, the same type of dysarthria may sound differently across patients, due to inter-subject variability. Second, in a clinical setting, the type of dysarthria is always determined by (subjective) interpretation, which makes the judgment dependent on the level of training of the assessor (inter-rater variability). For dysarthria type classification, one should not only hear the deviant speech characteristics, but these have to be interpreted in order to acknowledge the underlying pathology, like weakness, coordination problems etc. Unfortunately, objective means to assess the type of dysarthria do not yet exist, but research in this field is ongoing.[19,20] Lastly, patients will often try to compensate for their speech deficit, which makes it difficult to differentiate deviant speech characteristics from compensatory mechanisms.

Looking at the International Classification of Functioning (ICF), speech is a bodily function, which can lead to problems at the levels of activity and participation (communication and conversation).[21] From a clinical perspective, assessment at the levels of activity and participation is most important, because of the impact of unintelligible speech on daily life. In the Netherlands, there are two intelligibility tests available at the level of activity: the 'Nederlandstalig Spraakverstaanbaarheidsonderzoek' on word level (NSVO)[22] and on sentence level (NSVO-Z).[23] For assessment at the level of speech functions (e.g. changes in vocal quality, speaking rate etc.), there was no validated assessment available in the Netherlands at the start of this PhD project. Assessment at the level of speech functions is important for several reasons. First, the best suitable exercises or speech therapy approach is determined by the specific speech disorder. For example, when weakness is the main problem, other exercises are indicated than when spasticity is the dominant feature. Furthermore, for treatment purposes, it is equally important to assess the severity of dysarthria. For example, when dysarthria is severe, treatment may be focused more on using communication aids than when dysarthria is mild. Severity assessment is also indispensable for the evaluation of treatment effects. Lastly, because speech characteristics in dysarthria reflect the site of the neurological lesion, determining the type of dysarthria can also support the process of disease diagnostics.

AIMS OF THIS THESIS

Two of the three aims of this thesis are focused on dysarthria and dysphagia in adult NMD. The third aim is focused on the assessment of dysarthria in general. The specific aims of this thesis are:

1. To examine prevalence rates of dysarthria and dysphagia in NMD;
2. To summarize the evidence for treatment of dysarthria and dysphagia in NMD;
3. To develop and evaluate a dysarthria assessment at the level of speech function.

The following research questions related to the above aims were formulated:

1a. What are the prevalence rates and severity scores of dysarthria and dysphagia in adult patients with NMD and how often do dysarthria and dysphagia co-occur?
1b. Is dysphagia present in patients with genetically proven myotonic dystrophy type 2?
2. Is there evidence for the efficacy of treatments directed at dysarthria and dysphagia administered by speech-language therapists in NMD patients?
3a. Is it possible to develop a valid and reliable dysarthria assessment for adult patients including maximum performance tests of speech production?
3b. What are the reference values for the maximum performance tasks of speech production in a large population of healthy Dutch adults, and are these related to relevant person characteristics?
3c. Does video training improve the correct identification of dysarthria type and severity by speech-language therapy students as well as by experienced speech-language therapists?

OUTLINE OF THE THESIS

This thesis is divided into two sections. Part I focuses on Aims 1 and 2 (i.e., prevalence rates of dysarthria and dysphagia and evidence for treatment in NMD) while Part II concerns Aim 3 (i.e., the development of a dysarthria assessment).

Part I: prevalence rates of dysarthria and dysphagia, and evidence for treatment in NMD

In ***chapter 2***, the prevalence rates of dysarthria and dysphagia in a large group of patients with a wide variation of NMD are presented (research question 1a). ***Chapter 3*** examines if dysphagia is present in myotonic dystrophy type 2 (research question 1b). In ***chapter 4***, evidence is identified for treatment of dysarthria and dysphagia in NMD (research question 2).

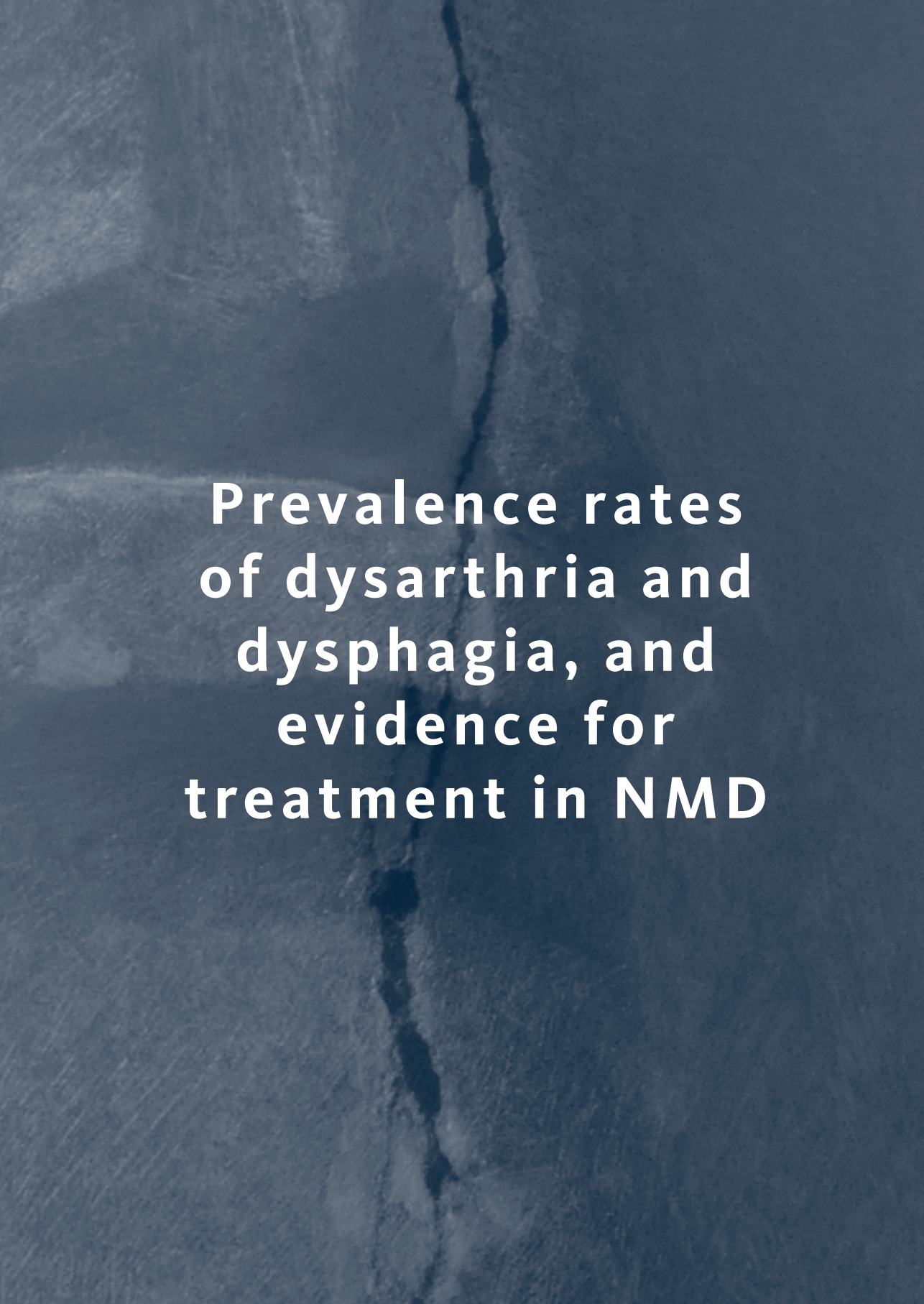
Part II: development of the Radboud Dysarthria Assessment (RDA)

Chapter 5 summarizes the development and validation of the Radboud Dysarthria Assessment (RDA) (research question 3a). In ***chapter 6***, reference values of a large population of healthy Dutch adults are presented in order to improve the interpretation of the performance of the dysarthric speakers (research question 3b). Finally, ***chapter 7*** explores whether video training using the RDA training videos is effective in labeling the type and severity of dysarthrias (research question 3c).

Finally, a summary and general discussion is provided in ***chapter 8***, followed by a summary in Dutch in ***chapter 9***.


1

Part I



Prevalence rates of dysarthria and dysphagia, and evidence for treatment in NMD

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Disability and Rehabilitation 2014; **36**(15): 1285–9

2 Dysarthria and dysphagia are highly prevalent among various types of neuromuscular diseases

ABSTRACT

Purpose

Patients with a neuromuscular disease (NMD) can present with dysarthria and/or dysphagia. Literature regarding prevalence rates of dysarthria and dysphagia is scarce. The purpose of this study was to determine prevalence rates, severity and co-presence of dysarthria and dysphagia in adult patients with NMD.

Methods

Two groups of adult patients with NMD were included: 102 consecutive outpatients (the 'unselected cohort') and 118 consecutive patients who were referred for multidisciplinary assessment (the 'selected cohort'). An experienced speech-language pathologist examined each patient in detail.

Results

The pooled prevalence of dysarthria was 46% (95% CI 36.5 – 55.9) and 62% (95% CI 53.3 – 70.8) in the unselected and selected cohorts, respectively. The pooled prevalence of dysphagia was 36% (95% CI 27.1 – 45.7) and 58% (95% CI 49.4 – 67.2) in the unselected and selected cohorts, respectively. There was a modest but significant association between the presence of dysarthria and dysphagia (r_s = 0.40; p < 0.01). Although the dysphagia was generally mild, dysarthria was moderate to severe in 15% of the dysarthric patients.

Conclusion

The prevalence rates of dysarthria and dysphagia among patients with various types of NMD are high. Physicians should therefore be aware of this prevalence and consider referring NMD patients to a speech-language pathologist.

INTRODUCTION

'Neuromuscular diseases' (NMD) is the generic term encompassing a group of approximately 600 rare diseases that are characterized by slowly progressive muscle weakness. NMDs can manifest at birth or later in life. NMDs can cause a wide range of disabilities, including oral motor disorders such as dysarthria and dysphagia. Dysarthria causes reduced speech intelligibility and can result in limitations in social interactions.[24] Dysphagia can also have a negative impact on daily functions and societal participation due to reduced food intake and/or restrictions in the consistency of food that can be ingested.[25] In addition, patients with severe dysphagia can experience life-threatening medical complications such as extreme weight loss or aspiration pneumonia.[26,27]

In the Netherlands, approximately 5% of patients with an NMD are referred to a speech-language pathologist;[28] however, the clinical prevalence rates of both dysarthria and dysphagia appear to be considerably higher. Although some studies have investigated the prevalence of dysphagia and dysarthria among NMD patients, these studies focused only on specific diseases such as myotonic dystrophy, myasthenia gravis, amyotrophic lateral sclerosis (ALS) and inclusion body myositis,[7,11,14,16-18,29,30] all of which are well-documented for causing dysarthria and/or dysphagia. However, little information is available regarding the prevalence rates of dysarthria and dysphagia in a wider range of NMD disorders. To address this question, we retrospectively investigated the prevalence, severity and co-presence of dysarthria and dysphagia in adult NMD patients.

METHODS

Patients

We retrospectively examined the clinical data obtained from two groups of adult patients with NMD who had been referred to our university hospital. Group 1 consisted of 102 consecutive outpatients who participated in a previous study to assess the policy for referring adult NMD patients in the Netherlands.[28,31,32] The inclusion criteria for this group were as follows: (i) probable or definite diagnosis of an NMD based on the medical records; (ii) 18 years of age or older at the time of the original study; and (iii) sufficient command of the Dutch language. Of 257 outpatients, 155 were excluded (80 did not meet the inclusion criteria, 14 did not respond, 53 refused to participate and 8 did not participate for another reason). The participants signed an informed consent form, and approval was obtained from the Medical Ethics Committee of the Radboud University Nijmegen Medical Center. Group 2 consisted of 133 patients who had been referred for a multidisciplinary assessment to our 'neuromuscular referral center' during a period of three and a half years. The patients were admitted if they had difficulty in multiple domains of daily functioning. Fifteen of the original 133 patients in Group 2 were excluded from the analysis; 14 patients could not be definitively diagnosed with an NMD, and speech and swallowing were not examined in one patient. The patients in Group 2 had not formally participated in any previous study. The clinical diagnoses were divided among the following four major disease categories: 'muscle diseases', 'disorders of the neuromuscular junction', 'neuropathies', and 'motor neuron diseases'. Based on the medical files, gender and age were recorded for all patients; information regarding the dura-

tion of the complaints associated with the NMD was available only for Group 1.

Dysarthria assessment

For both groups, a standardized history was obtained by an experienced speech-language pathologist in order to identify any speech and swallowing complaints; this history was followed by an examination of oral motor function, speech and swallowing. Dysarthria was diagnosed using the Nijmegen Dysarthria Assessment, which includes spontaneous speech, reading a standardised text, maximum repetition rate, maximum phonation duration and maximum phonation frequency range.[33,34] Based on this examination, the dysarthria was scored using the validated Dutch modified version of the dysarthria sub-scales of the Therapy Outcome Measures (TOM),[35] with a scale ranging from 5 to 0. We assigned the scores to the following three severity groups: absent (score 5), mild dysarthria (score 4 or 3), and moderate/severe dysarthria (score 2, 1 or 0).

Dysphagia assessment

Swallowing was clinically assessed using both quantitative and qualitative measures (instrumental investigation of swallowing like videofluoroscopy or endoscopy is not part of the standard protocol for neuromuscular patients in our center). For the quantitative assessment, swallowing tests were performed and included the swallowing speed and the dysphagia limit.[36,37] For the qualitative assessment, the patients were asked to drink water and eat a biscuit, and the rate of chewing, the frequency of swallowing, the presence or absence of compensatory behaviours (e.g. head movements) and signs of residue, penetration (the passage of food and/or liquid into the larynx) or aspiration (inhalation of food and/or liquid through the vocal cords) were observed and recorded.[38,39] The severity of the dysphagia was classified using the following scale: (1) no problems, (2) mild dysphagia, and (3) moderate/severe dysphagia. A patient who had no swallowing problems either in their history or during the clinical assessment scored a 1 ('no problems'). A patient who achieved full oral intake with some adaptations (e.g. in food consistency and/or drinking during the meal) and who exhibited minor problems during the qualitative assessment or performed just below normal on the swallowing tests scored a 2 ('mild'). Patients scored a 3 ('moderate/severe dysphagia') if they required considerable adaptations to achieve full oral intake, if a complete oral intake was not possible, if they exhibited many problems during the qualitative assessment, or if their performance was clearly abnormal or they were unable to perform the swallowing tests.

Statistical analysis

The prevalence rates of dysarthria and dysphagia were first calculated for each disease category. To obtain overall values, pooled prevalence rates were estimated per patient group by weighting each disease category according to sample size, as the patients were not distributed equally across the disease categories within the groups. The Chi-square test was used to analyze the between-group differences and the differences between the prevalence of dysarthria and dysphagia within each group. The Spearman correlation coefficient was calculated and used to determine the relationship between dysarthria and dysphagia as well as between the severity of impairment and complaint duration within each group. All calculations were performed using SPSS 20.0 (IBM SPSS Statistics, Chicago, IL), and differences with $p < 0.05$ were considered to be statistically significant.

RESULTS

Patients

Data were collected from 220 patients (Group 1 contained 102 patients, and Group 2 contained 118 patients) and analyzed. The groups did not differ significantly with respect to gender or age, as seen in Table 2.1. Because the category 'muscle diseases' encompassed a large, heterogeneous group of disorders, this category was sub-divided into 'muscular dystrophies', 'metabolic myopathies', 'inflammatory myopathies' and 'other myopathies'.

Table 2.1. Demographics and clinical characteristics of the two patient groups in this study.

Category/diagnosis*	Group 1	Group 2	Total
Men / women	50% / 50%	47% / 53%	-
Mean age, years (SD)	48 (13.3)	44 (13.8)	-
Mean complaint duration, years (SD)	12 (11)	-	-
Muscle diseases	**68**	**103**	**171**
Muscle dystrophies (myotonic dystrophy type I and II (MD), oculopharyngeal muscular dystrophy (OPMD), facioscapulohumeral muscular dystrophy (FSHD), limb-girdle muscular dystrophy, Becker muscular dystrophy, Duchenne muscular dystrophy)	39	55	94
Metabolic myopathies (mitochondrial myopathy)	11	30	41
Inflammatory myopathies (polymyositis (PM), dermatomyositis (DM), inclusion body myositis (IBM), ocular myositis)	8	6	14
Other myopathies (nemaline myopathy, Miyoshi myopathy)	10	12	22
Disorders of the neuromuscular junction (myasthenia gravis and Lambert Eaton myasthenic syndrome)	**7**	**0**	**7**
Neuropathies (chronic inflammatory demyelinating polyneuropathy (CIDP), Guillain-Barré syndrome, hereditary motor and sensory neuropathy (HMSN), chronic idiopathic axonal polyneuropathy (CIAP), multifocal motor neuropathy (MMN), polyneuropathy, neuralgic amyotrophy)	**19**	**9**	**28**
Motor neuron diseases (amyotrophic lateral sclerosis (ALS), spinal muscular atrophy (SMA))	**8**	**6**	**14**
Total	**102**	**118**	**220**

*Main categories are in bold.

Prevalence and severity of dysarthria

Overall, 123 of the 220 patients (56%) received a score of 4, 3, 2, 1 or 0 on the dysarthria severity scale, yielding a pooled prevalence of 46% (95% CI 36.5 – 55.9) in Group 1 and 62% (95% CI 53.3 – 70.8) in Group 2 (p = 0.006), as seen in Table 2.2. In Group 1, the highest prevalence rate was among patients with disorders of the neuromuscular junction (71%); in Group 2, the highest prevalence rate was in the muscle diseases category (68%), particularly within the muscular dystrophies (87%). The distribution of the severity of dysarthria is shown in Figure 2.1A. Moderate/severe dysarthria was present only in the categories 'neuropathies', 'disorders of the neuromuscular junction' and 'muscle diseases'; and the

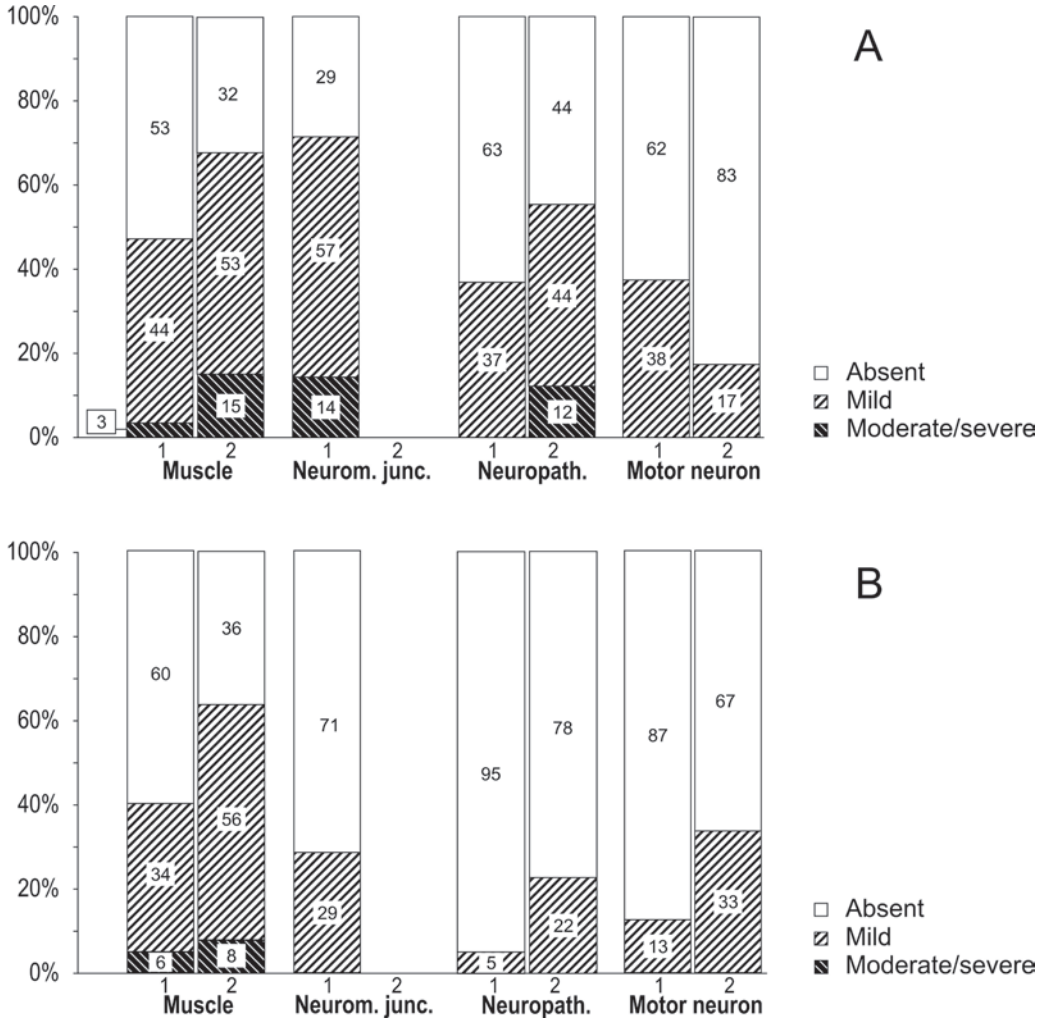


Figure 2.1. Relative distributions of the severity of dysarthria (A) and dysphagia (B) in Group 1 and Group 2 for the indicated NMD categories. Abbreviations used: Muscle, muscle diseases; Neurom. jun., disorders of the neuromuscular junction; Neuropath., neuropathies; Motor neuron, motor neuron diseases.

Table 2.2. Frequencies of dysarthria and dysphagia among the two groups.

Neuromuscular disorder	Dysarthria						Dysphagia					
	Group 1			Group 2			Group 1			Group 2		
	n	Present[a]	%	*n*	Present[a]	%	*n*	Present[b]	%	*n*	Present[b]	%
Muscle diseases (total)	68	32	47	103	70	68	68	27	40	103	66	64
Muscle dystrophies	39	20	51	55	48	87	39	16	40	55	42	76
Metabolic myopathies	11	5	46	30	16	53	11	6	55	30	16	53
Inflammatory myopathies	8	4	50	6	0	0	8	3	38	6	4	67
Other myopathies	10	3	30	12	6	50	10	2	20	12	4	33
Disorders of the neuromuscular junction	7	5	71	0	-	-	7	2	29	0	-	-
Neuropathies	19	7	37	9	5	56	19	1	5	9	3	33
Motor neuron diseases	8	3	38	6	1	17	8	1	13	6	2	33
Total (pooled prevalence)	102	47	46	118	76	62	102	31	36	118	71	58
			(36.5 – 55.9)[c]			(53.3 – 70.8)[c]			(27.1 – 45.7)[c]			(49.4 – 67.2)[c]

[a] Presence of dysarthria is defined as a score of 0, 1, 2, 3 or 4 (see 'Methods' section). [b] Presence of dysphagia is defined as a score of 2 or 3 (see 'Methods' section). [c] The 95% confidence interval is provided within parentheses.

majority of the dysarthric patients had mild dysarthria. There was no significant correlation between the severity of dysarthria and complaint duration (r_s = 0.107; p = 0.142).

Prevalence and severity of dysphagia

Overall, 103 of the 220 patients (47%) had mild to severe dysphagia, yielding a pooled prevalence of 36% (95% CI 27.1 – 45.7) in Group 1 and 58% (95% CI 49.4 – 67.2) in Group 2 (p <0.001), as seen in Table 2.2. The highest prevalence of dysphagia was in the category 'muscle diseases' for both Group 1 (40%) and Group 2 (64%). Within the muscle diseases category, the patients with metabolic myopathies (in Group 1; 55%) and the patients with muscular dystrophies (in Group 2; 76%) had the highest prevalence of dysphagia. The distribution of the severity of dysphagia is shown in Figure 2.1B. Overall, the majority of patients who had dysphagia presented with mild dysphagia; moderate/severe dysphagia was found only among patients in the muscle diseases category. There was no significant correlation between the severity of dysphagia and complaint duration (r_s = 0.105; p = 0.146).

Relationship between dysarthria and dysphagia

The analysis revealed modest but significant correlations between the occurrence of dysarthria and dysphagia in both Group 1 (r_s = 0.40, p <0.001) and Group 2 (r_s = 0.41, p <0.001). In total, 84 of the 220 patients in this study (38%) were diagnosed with both dysarthria and dysphagia; the majority of these 84 patients (76 patients, or 90%) had a muscle disease, two patients had a motor neuron disease, four patients had a neuropathy and two patients had a disorder of the neuromuscular junction. In both groups, the overall prevalence of dysarthria was higher than the overall prevalence of dysphagia; this difference reached significance in Group 2 (p = 0.013), but not in Group 1 (p = 0.078).

DISCUSSION AND CONCLUSION

The goal of this study was to investigate the frequency, severity and co-presence of dysarthria and dysphagia in various types of NMD in two relatively large patient cohorts who had previously visited our university hospital. The principal result is the high overall pooled prevalence rates of dysarthria (56%) and dysphagia (47%) in both groups. The prevalence of each disorder was consistently higher in the group of patients who had been referred for multidisciplinary assessment and advice (Group 2) than in the 'unselected' patient cohort (Group 1). This difference is likely due to selection bias, as the patients who had been referred for multidisciplinary assessment were referred by a specialist on the basis of their complaints and disabilities.

Interestingly, dysarthria and dysphagia were found among patients with NMD that are not typically associated with these disorders. For example, dysphagia has been mentioned in the context of inflammatory myopathies,[11,18] whereas dysarthria has not been described explicitly. However, we identified dysarthria in half of our patients. Another interesting finding was the prevalence of both dysarthria and dysphagia in half of the patients with metabolic and other myopathies; this finding has also not been described explicitly in any previous report. In the category 'neuropathies', only Guillain-Barré syndrome is known to cause dysphagia and dysarthria. However, we also found speech and swallowing impairments in two out of six patients with hereditary motor and sensory neuropathy, one out of two patients with chronic inflammatory demyelinating polyneuropathy, and one out

of four patients with multifocal motor neuropathy. On the other hand, it is well-documented that in ALS, both dysarthria and dysphagia can occur, and their prevalence increases during disease progression in nearly all patients;[7,40] in our patient cohort, all nine ALS patients were affected, albeit only mildly.

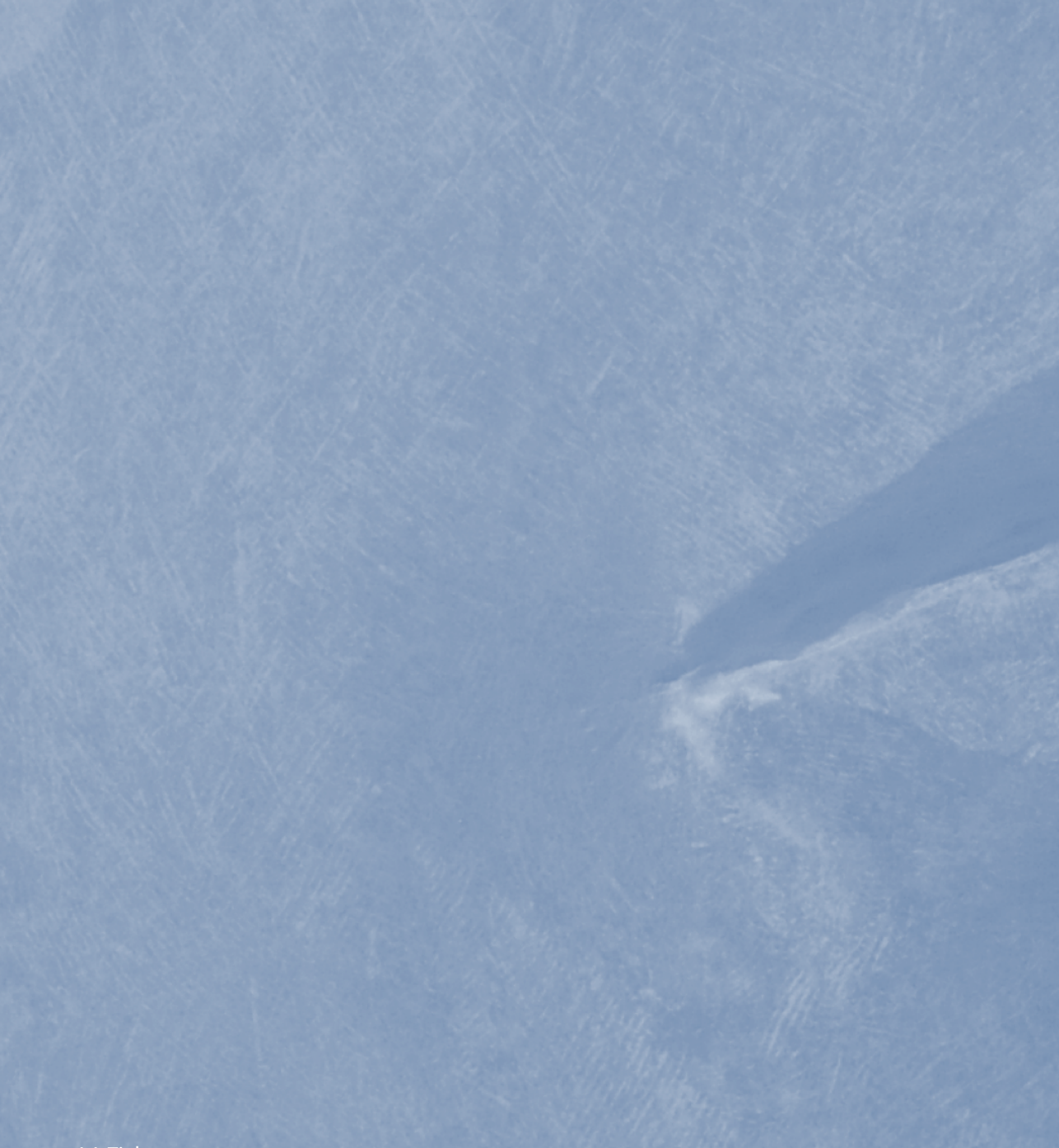
Overall, the prevalence rates of dysarthria and dysphagia in our study are higher than the rates reported by previous studies. This discrepancy might be due in part to the assessment methods. Most reported prevalence rates are based on interviews and/or questionnaires,[11,16,17,40] whereas our patients were assessed by an experienced speech-language pathologist who examined both speech and swallowing functions. Because a clinical examination is generally more sensitive than a self-reporting assessment by the patient,[41] an underreporting of complaints and disabilities can occur among patients with NMD, as many patients become accustomed to the relatively mild impairments in speech and swallowing during the progression of their disease.

Although the severity of symptoms generally increases with disease progression, we found no significant correlation between the duration of complaints and the severity of either dysarthria or dysphagia. This finding is likely due to the heterogeneity of the diagnoses included within our patient groups and the extremely slow progression of some of the individual diseases. In addition, recall bias may have played a role in affecting the patients' ability to accurately indicate the duration of their complaints.

The results of this study suggest that dysarthria is generally more prevalent than dysphagia among NMD patients. This finding is consistent with several other chronic neurological disorders such as Parkinson's disease and multiple sclerosis.[42,43] In our study, the observed correlation between dysarthria and dysphagia in each patient group was modest and can be explained physiologically. Although speech is generally considered to be a voluntary motor process, the pharyngeal and oesophageal stages of swallowing are characterized as a predominantly reflexive process. In addition, the lingual forces and jaw movements are quite different during the oral stages of swallowing than during vocalization; vocalization uses more rapid and differentiated movements than oral preparation and the transport of food.[44] Thus, because dysarthria need not necessarily present together with dysphagia, both disorders should be assessed.

The following limitations of our study should be mentioned. First, this was a retrospective cohort study in which the data were not originally collected for the purpose of defining the prevalence rates of dysarthria and dysphagia. Nevertheless, all of the patients were seen by a speech-language pathologist who provided us with complete data sets for all 220 patients. Second, the neurology and rehabilitation departments that were involved in this study specialize in the assessment and treatment of patients with NMD, and this may have caused an overrepresentation of some disease categories. Third, any comparison with previous studies that reported prevalence rates of dysarthria and dysphagia is hampered by the various assessment techniques that have been used. Therefore, future research should attempt to standardize the assessment methods so that symptomatic patients can be properly identified, characterized and referred. To this end, a short questionnaire was developed and evaluated for patients with NMD to facilitate their referral to a speech-language pathologist when warranted.[45]

Taken together, our results support the conclusion that physicians should be aware that speech and/or swallowing problems are highly prevalent among many NMD patient types, irrespective of the patient's disease stage. Even though some patients with a progressive disease may not benefit from treatment directed at improving their speech and/or swallowing efficacy, these patients may still benefit from learning individually tailored functional compensations delivered by an experienced speech-language pathologist.[6,28,46-48]



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3 Dysphagia is present but mild in Myotonic Dystrophy type 2

ABSTRACT

The phenotype of Myotonic Dystrophy type 2 (DM2) shows similarities as well as differences to that of Myotonic Dystrophy type 1 (DM1). Dysphagia, a predominant feature in DM1, has not yet been examined in DM2. In a recent nationwide questionnaire survey of gastrointestinal symptoms in DM2, 12 out of 29 DM2 patients reported to have difficulty in swallowing for solid food. The aim of the study was to investigate the presence of dysphagia in patients with genetically proven DM2 who reported difficulty in swallowing for solid food at the questionnaire survey. Swallowing function and fiberoptic endoscopic evaluation of swallowing (FEES) were examined by a speech therapist and otorhinolaryngologist, respectively. In DM2 patients who reported difficulty in swallowing the presence of dysphagia could be confirmed (clinically in 100%, by FEES in 88%). A correlation exists between Dysphagia Outcome and Severity Score (DOSS) and age ($p = 0.05$). None of the patients was underweight, and none of the patients had suffered aspiration pneumonia in the past. Dysphagia is present among DM2 patients and is more severe in older patients. However, dysphagia is generally mild, and does not lead to weight loss, or aspiration pneumonia.

INTRODUCTION

Myotonic Dystrophy type 2 (DM2, MIM 602668) is a dominantly inherited multisystem disorder characterized by progressive proximal muscle weakness, myotonia, cataracts, cardiac arrhythmia, and muscle pain. DM2, earlier known as Proximal Myotonic Myopathy (PROMM) is caused by a CCTG expansion in intron 1 of the ZNF9 gene on chromosome 3q21.[49] The clinical picture of DM2 shows similarities as well as differences to that of Myotonic Dystrophy type 1 (DM1). Shared core features are autosomal dominant inheritance, muscle weakness, myotonia, cataracts, and multi-organ involvement with cardiac conduction defects, insulin resistance, and gonadal atrophy. Notably absent is a congenital form of DM2, and evidence of anticipation is less striking than in DM1, while muscle pain is more prominent in DM2.[50]

Dysphagia, a predominant and potentially life-threatening feature in DM1, has not yet been examined in DM2. Dysphagia has been described in a few scattered PROMM and DM2 case reports.[51-53] We recently performed a prospective nationwide study to explore the presence and characteristics of gastrointestinal dysfunction in DM2 patients.[54] In that validated questionnaire survey, 12 out of 29 patients reported to have swallowing difficulties for solid food.

In this study, we set out to assess oropharyngeal swallowing in patients with genetically proven DM2 who reported difficulty in swallowing at the questionnaire survey in order to objectify this symptom.

PATIENTS AND METHODS

Patients

In a recently performed nationwide survey genetically proven DM2 patients filled out the Dutch Gastrointestinal Symptoms Questionnaire (GSQ).[54] This standardized questionnaire covers symptoms from all parts of the gastrointestinal tract and contains questions about the severity of gastrointestinal symptoms during the last 4 weeks.[55] All DM2 patients who reported swallowing difficulties at the GSQ were recruited for this study. Exclusion criterion for participating was pregnancy because of its increased risk of pulmonary aspiration. Patients were approached for additional information about their medical history and the body mass index (BMI) was calculated for all patients. The medical ethics committee approved the protocol and all patients gave written consent.

METHODS

All DM2 patients were clinically investigated by an experienced speech therapist in neuromuscular disorders. Eight DM2 patients gave additional informed consent to fiberoptic endoscopic evaluation of swallowing (FEES).

Clinical investigation

An orofacial examination was performed as previously described in literature.[56] In short, the clinical examination included:

- Assessment of the strength of the jaws, lips, and tongue; weakness was scored as mild, moderate or severe.
- Assessment of myotonia of the masseter muscles and tongue; myotonia was scored as absent or present.
- Observation while eating solid food (a bisquit).
- A timed test of swallowing.[37]

FEES

FEES is a valid and exceptionally safe technique for assessing pre-swallow anatomy and physiology in order to objectively evaluate patients with dysphagia.[57] Diagnosis of

oropharyngeal dysphagia was determined by premature spillage, pharyngeal residue, laryngeal penetration, and aspiration.

Dysphagia outcome and severity scale

All symptoms were scored with the Dysphagia Outcome and Severity Scale (DOSS), a reliable scale developed to rate the functional severity of dysphagia based on objective assessment.[58] Scores range from 1 (severe oropharyngeal dysphagia necessitating non-oral nutrition) to 7 (normal in all situations). Oropharyngeal dysphagia was diagnosed when a patient was scored on level 1 to 6.

Statistical analysis

We used the Pearson partial correlation coefficient for calculating correlations, and considered $p < 0.05$ significant.

RESULTS

Clinical investigation

Twelve DM2 patients complaining of swallowing difficulties were recruited. Ten (from eight DM2 families) of the 12 patients took part in the study, two patients refused to participate. Mean age was 58.0 years (SD 11.6) and mean age of disease onset was 26.7 years (SD 13.0). Mean BMI in DM2 patients was 25.1 (SD 4.0), none of the patients was underweight (BMI < 18.5). None of the patients had suffered aspiration pneumonia in the past. Seven patients showed signs of weakness of the jaws, lips, or tongue. None of the patients showed myotonia of the jaws or tongue.

Evaluation of eating showed slowness in all DM2 patients, and frequent swallowing in nine. Six patients demonstrated adapting head position upon swallowing. Coughing and wet phonation, indicating possible residue, penetration or aspiration, were observed in five patients.

A Timed Test of swallowing showed the swallowing speed (average volume per swallow) to be too low, that is to say lower than normal, in four patients (patient 3, 4, 8, and 9). Five other patients (patient 1, 5, 6, 7, and 10) drank more slowly than the mean swallowing speed of age and sex matched healthy controls from literature, but still fell within normal values (Table 3.1).[59]

FEES

One patient exhibited normal swallowing and seven (88%) exhibited dysphagia; residue of solid food in seven cases (88%), residue of milk in six cases (75%), and residue of saliva was observed in two cases (25%). Spillage of milk was revealed in two patients (25%), spillage of solid food was detected in one patient (13%). No penetration or aspiration of milk or solid food was detected (Table 3.1).

Dysphagia outcome and severity scale

DOSS was found to be abnormal in all DM2 patients, ranging in severity from 'mild-moderate' (score 4) to 'within functional limits' (score 6). DOSS score and age correlated in patients (*rho* = -0.66, $p = 0.05$), age at disease onset and swallowing speed correlated significantly (*rho* = -0.68, $p = 0.04$). There was no correlation between DOSS score and age at disease onset or disease duration.

DISCUSSION

In DM2 patients who reported difficulties in swallowing the presence of dysphagia could be confirmed (clinically 100%, by FEES 88%). Dysphagia was more pronounced in older DM2 patients. None of the 10 patients had suffered aspiration pneumonia in the past, and no patient was underweighted or had significantly lost weight. These findings

Table 3.1. Patient characteristics and results of clinical investigation (orofacial examination, timed test of swallowing and evaluation of eating), FEES and DOSS.

	Pt 1	Pt 2	Pt 3	Pt 4	Pt 5	Pt 6	Pt 7	Pt 8	Pt 9	Pt 10
Characteristics										
Sex	F	F	M	M	F	F	F	F	F	F
Age, y	68	55	37	64	71	72	55	62	46	48
Age at disease onset, y	43	18	27	52	22	12	10	28	26	29
Orofacial examination										
Weakness of jaws	–	–	–	–	–	±	–	–	–	–
Weakness of lips	–	–	–	±	–	±	–	–	±	–
Weakness of tongue	±	±	–	±	±	±	–	±	–	–
Timed test of swallowing										
Swallowing speed in mL/s	8	14	16	11	11	12	11	4	8	9
Evaluation of eating										
Slowness	++	+	+	+	+	+	++	++	+	±
Frequent swallowing	+	+	+	+	+	+	–	+	+	+
Adapting head position	+	+	+	–	–	+	–	+	–	+
Coughing, wet phonation	+	–	+	–	–	+	–	–	+	+
FEES	NA		NA							
Deviant anatomy		–		+	–	–	–	–	–	–
Residue of saliva		–		+	–	–	–	–	–	+
Spillage of milk		+		–	–	–	+	–	–	–
Spillage of biscuit		+		–	–	–	–	–	–	–
Residue of milk		+		+	+	+	–	+	–	+
Residue of biscuit		+		+	+	+	+	+	–	+
Penetration of milk		–		–	–	–	–	–	–	–
Penetration of biscuit		–		–	–	–	–	–	–	–
Aspiration of milk		–		–	–	–	–	–	–	–
Aspiration of biscuit		–		–	–	–	–	–	–	–
DOSS	4	4	5	4	4	4	5	4	6	4

Symbols used: (–) absent; (±) minimal signs; (+) clear signs; (++) severe; NA, not assessed. Abbreviations used: DOSS, Dysphagia Outcome and Severity Score (7 = normal, 6 = within functional limits, 5 = mild, 4 = mild-moderate, 3 = moderate, 2 = moderate-severe, 1 = severe dysphagia).[58]

are in correspondence with the results that dysphagia is generally mild.

These results give little or no indication of the underlying pathology of swallowing difficulties. Weakness of the oropharyngeal muscles as well as subclinical myotonia may play a role. In DM1 both myopathic weakness and myotonia encountered in oropharyngeal muscles play an important part in the oral and the pharyngeal phases of swallowing dysfunction.[60] In our group, minimal weakness of the orofacial muscles but no myotonia was observed clinically. Electrophysiological evaluation of swallowing is necessary to show the possible existence and frequency of subclinical electrophysiological abnormalities in oropharyngeal swallowing and may clarify the mechanisms of dysphagia.

Recognition of dysphagia may have implications for the management of DM2 patients and for recommendations regarding the prognosis. Firstly, DM2 patients with dysphagia may benefit from conservative interventions such as speech therapy and modification of food consistency. Secondly, since the major complication of dysphagia is aspiration, dysphagia may be a problem that can lead to pneumonia. We found, however, no evidence of severe late stage dysphagia and pneumonia due to aspiration in DM2 patients as is the case in adult-onset DM1 patients.

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Folia Phoniatrica et Logopaedica 2011; **63**(1): 15-20

4 Speech pathology interventions in patients with neuromuscular diseases: a systematic review

ABSTRACT

Purpose

A systematic review was conducted to summarize and evaluate the literature on the effectiveness of speech pathology interventions in adults with neuromuscular diseases.

Method

Databases searched included the Cochrane Database of Systematic Reviews, the Cochrane Central Register of Controlled Trials, MEDLINE, CINAHL, EMBASE, PsychINFO and Pubmed. A total of 1,772 articles were independently screened on title and abstract by two reviewers.

Results

No randomized controlled trials or clinical controlled trials were found. Four other designs were included. Only one study on oculopharyngeal muscle dystrophy (OPMD) appeared to have sufficient methodological quality. There is evidence indicating that correction of head position in patients with OPMD improves swallowing efficiency (level III evidence).

Conclusion

Despite 1,772 studies, there is only evidence of level III regarding the effectiveness of speech pathology interventions in patients with OPMD. Recommendations for future research are given.

INTRODUCTION

Neuromuscular diseases (NMD) represent a subfield of neurology, including heterogeneous groups of patients with approximately 600 diseases of the muscle, neuromuscular transmission, peripheral nerve and nerve root or motor neuron. NMD can lead to dysarthria and dysphagia, which are both diagnosed and treated by speech pathologists (SP).

Both dysarthria and dysphagia are not only relatively frequently prevalent, but also clinically relevant symptoms in NMD. The impact of dysarthria on quality of life is obvious, since communication is generally considered to be a critical determinant of quality of life. Dysphagia can lead to aspiration pneumonia and therefore to serious health problems. Dysphagia and concomitant pneumonia may even be life-threatening in otherwise weakened NMD patients.

In a sample of 102 NMD patients, we found that 45% of the patients had an indication for SP advice regarding dysarthria or dysphagia.[28] However, this judgment was based on the expertise of the participating speech pathologists. The aim of this article is to assess whether there is evidence for the efficacy of treatments administered by speech pathologists in patients with NMD.

METHODS

Search strategy

We searched the Cochrane Database of Systematic Reviews and the Cochrane Central Register of Controlled Trials (The Cochrane Library November 2007, Issue 3), MEDLINE (1966 through November 2007), CINAHL (1982 through November 2007), EMBASE (1980 through November 2007), PsycINFO (1806 through November 2007) and Pubmed (1950 through November 2007). The search strategy was built on different types of NMD and different types of SP interventions. For all search strategies, Medical Subject Headings (MeSH) or indexed terms were used as well as free text words. The full search strategy is shown in Table 4.1. Reference lists of reviews and

Table 4.1. Free text words and MeSH terms used in search strategy to find evidence for the effectiveness of speech pathology in NMD.

Free text words	
Speech pathology	speech pathology, speech therapy, speech and language therapy, speech disorder, communication disorder, articulation disorder, swallow disorder, deglutition disorder, dysphagia, dysarthria, dietary modification, swallow, intelligibility, alternative communication
NMD	neuromuscular disease, neuromuscular disorder, muscle disease, muscle disorder, neuromuscular junction disease, neuromuscular junction disorder, motor neuron disease, motor neuron disorder, motoneuron disease, motoneuron disorder, neuropathy, polyneuropathy, peripheral nervous system disease, peripheral nervous system disorder, neuralgia, neuritis, myopathy, dystrophy, myotony, myositis
MeSH terms	
Speech pathology	dysarthria, speech disorder(s), speech therapy, speech pathology, communication disorders, communication aids, speech intelligibility, facilitated communication, communication aid, alternative and augmentative communication, communication aids for the disabled, dysphagia, deglutition disorders
NMD	neuromuscular disease(s), neuromuscular disorders

selected articles were scanned for further potentially relevant articles.

Selection criteria

Inclusion was restricted to peer-reviewed articles published in the English, German, French or Dutch language. Randomized Controlled Trials (RCTs), Clinical Controlled Trials (CCTs) and other designs (ODs) were included. Single case studies were excluded. Participants included adults (>18 years) with various types of NMD (disorders of the muscle, neuromuscular transmission, peripheral nerve and nerve root or motor neuron).

SP interventions included information and advice or teaching compensatory strategies. Information and advice incorporated (1) dietary modification; (2) augmentative and alternative communication; or (3) instruction of the patient and relatives. Compensatory strategies included teaching swallowing maneuvers and/or strategies to improve intelligibility. Brain computer communication devices such as thought translation devices were excluded, as this is not considered a regular SP intervention. For the same reason, we also excluded medical interventions related to dysphagia, such as myotomy of the cricopharyngeal muscle.

The outcome measures included swallowing at the level of body functions and intelligibility, communication and eating and drinking at the level of activities and participation, according to the definitions of International Classification of Functioning, Disability and Health (ICF).[21]

Procedure for inclusion

Two reviewers (S.K. and E.H.C.) independently carried out a preliminary selection based on title and abstract of articles. Full texts of articles that seemed to fit the inclusion criteria were retrieved for further evaluation. A final selection was made independently by the two reviewers on the basis of full-texts. When different conclusions were drawn, consensus was reached through discussion.

Methodological quality

The methodological quality of the studies was independently rated using criteria recommended by Van Tulder et al.[61] The 2 reviewers independently assessed the methodological quality. Disagreements were resolved by discussion between the reviewers.

For RCTs and CCTs, the quality of reporting was scored on 6 criteria (eligibility criteria, comparability of groups, explicit description of interventions, adverse effects, short-term and long-term follow-up measurements). Methodological quality was scored on 9 criteria (randomization, concealed allocation, avoidance of co-interventions, compliance, blinding of outcome assessor, relevance of outcome measures, acceptable drop-out rate, comparable timing of outcome assessment and intention-to-treat analysis). Statistical quality was scored using 2 criteria (description of sample size and presentation of point estimates/measures of variability). One criterion regarding statistical quality (between-group statistical comparison) of the PEDro scale was added.[62]

Methodological quality of articles was considered sufficient when 3 out of 6 reporting criteria, 6 out of 9 methodological criteria, and 2 out of 3 statistical criteria were scored positively.[61] For ODs, the criteria of comparability of groups, randomization, allocation, and between-group comparison were not applicable. For these designs, 2 out of 5 reporting criteria, 4 out of 7 methodological criteria, and 1 of 2 statistical criteria had to be scored positively to be of sufficient quality.[63]

Best evidence synthesis

A best-evidence synthesis was performed according to the classification of the Dutch Institute for Health Care Quality Improvement.[64] Three levels of evidence and conclusions were formulated. Level I evidence refers to at least 2 RCTs of sufficient quality. Conclusions are formulated as "it has been shown that…". Level II evidence refers to 1 good quality RCT or at least 2 independent controlled studies (RCTs or CCTs) of less methodological quality. Conclusions are formulated as "it is likely that…". Level III evidence refers to an RCT or CCT of low methodological quality or at least 1 OD of sufficient methodological quality. Conclusions are formulated as "there are indications that…". When inconsistent findings were found in studies of similar design and methodological quality, conclusions are formulated as "there is insufficient evidence that…".

RESULTS

Selection

The search strategy identified 1,772 articles, duplicates excluded (Figure 4.1). After screening on title and abstract, 1,723 articles were excluded because they did not meet predefined criteria for study design, type of participants, type of intervention, type of outcome measures, or English, French, German or Dutch language. In 42 abstracts, there was discussion between the two reviewers, leading eventually to retrieval of 17 full-text articles that seemed to fit the inclusion criteria. In addition, 7 review articles were retrieved and screened for additional articles. Another 15 articles from reference lists were retrieved full text. Following evaluation of all full-text articles, 4 fulfilled all inclusion criteria.[46,65-67] These studies were subjected to assessment of methodological quality.

Methodological assessment

The 4 studies included were all ODs. The methodological quality of 3 studies was insufficient[65-67] and the methodological quality of 1 study was found sufficient (Table 4.2).[46] De Swart *et al.* found a negative influence of ptosis on swallowing function in patients with oculopharyngeal muscle dystrophy (OPMD).[46] The instruction 'head position slightly flexed', i.e. head not adapted to the ptosis, significantly increased swallowing efficiency. This was objectively evaluated with videofluoroscopy (with 20 ml. thin and thick liquid) and maximum swallowing volume.

Best-evidence synthesis

The study of De Swart *et al.* is an OD of sufficient methodological quality. Therefore, the evidence-based conclusion was that there are indications (level III evidence) that when head position is slightly flexed and not adapted to the ptosis, swallowing efficiency improves in OPMD-patients.

DISCUSSION

The main conclusion of this study is that the published evidence for effectiveness of SP in adults with NMD is very limited, as our search strategy did not identify RCTs or CCTs on this topic. There was only 1 OD (uncontrolled pre-post study) which was of sufficient methodological quality.

Regarding dysphagia, the only available evidence (level III) found was based on an uncontrolled study.[46] As we know from normal swallowing, extension of the neck leads to ineffective swallowing.[68-70] Therefore, basic instructions about the influence of posture of head and neck on swallowing seem valuable in NMD in general – par-

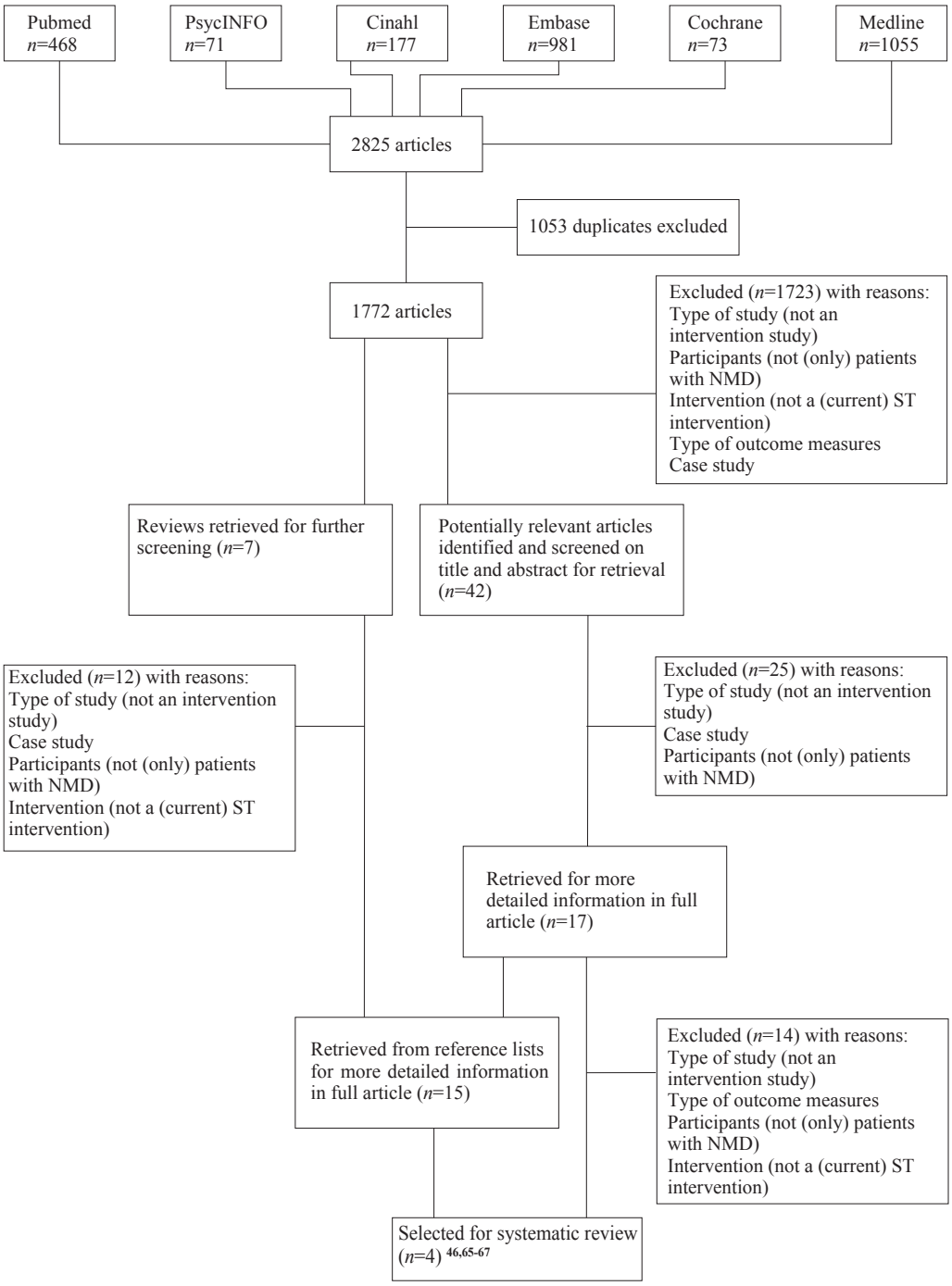


Figure 4.1. Selection strategy for the systematic review of the effectiveness of speech pathology interventions in adults with neuromuscular diseases.

ticularly when the disease leads to changes in posture or weakness in the head-neck-shoulder region or when the disease leads to weakness in muscles involved in speech or swallowing.

Regarding dysarthria, despite the extensive number of articles, no evidence was found for the effectiveness of SP in NMD. We only found two articles in which one-time manipulations of speaking rate led to a slower speaking rate, but this technique did not have a positive effect on intelligibility.[71,72] All 4 studies included a homogeneous patient population with regard to the medical diagnosis. This may limit the generalization of the results to other groups of patients

Table 4.2. Methodological quality of included studies showing 3 studies of insufficient methodological quality[65-67] and one study of sufficient methodological quality.[46]

Criteria	Silbergleit et al. 1991 [67]	Oh et al. 2007 [65]	Scott & Austin, 1994 [66]	De Swart et al. 2006 [46]
Reporting criteria				
Eligibility criteria specified?	Yes	Yes	Yes	Yes
Intervention explicitly described?	Yes	Yes	No	Yes
Adverse effects described?	No	Yes	Yes	No
Short-term follow-up measurement performed?	No	No	Unknown	Yes
Long-term measurement performed?	Yes	Yes	Yes	No
Total descriptive criteria (at least 2/5)	3/5	4/5	3/5	3/5
Methodological criteria				
Co-interventions avoided?	Unknown	No	No	Yes
The compliance acceptable?	Yes	Unknown	Yes	Yes
Outcome assessor not involved in the treatment?	Unknown	Unknown	No	N.A.
Outcome measures relevant?	Yes	Unknown	No	Yes
Withdrawal/drop-out rate described and acceptable?	Yes	No	Yes	Yes
Timing of the outcome assessment in all patients comparable?	No	No	Unknown	Yes
Analysis included an intention to treat analysis?	No	No	Unknown	No
Total internal validity (at least 4/7)	3/7	0/7	2/7	5/7
Statistical criteria				
Was the sample size of the group described?	Yes	Yes	Yes	Yes
Were point estimates and measures of variability presented for the primary outcome measures?	Yes	No	No	Yes
Total statistical criteria (at least 1/2)	2/2	1/2	1/2	2/2

with NMD. However, many NMD patients suffer from weakness in speech and swallowing muscles. Therefore, we hypothesize that the SP interventions are applicable to different groups of NMD patients with comparable impairments and disabilities. The comparison between studies is also difficult because of the different outcome measures used. Finally, considering the small number of patients, there might be a matter of a type II error in the included studies. This problem can be solved by including different groups of NMD patients with comparable impairments in one study.

Exclusion of studies

Our extensive search resulted in a large number of articles, most of which were excluded. Many studies did not evaluate the effectiveness of SP interventions but were descriptive studies, for instance on dysarthria or dysphagia in patients with NMD. The use of MeSH and free text words for different types of NMD resulted in many studies concerning other neurological conditions, such as Parkinson's disease or stroke, which did not fulfill our inclusion criteria.

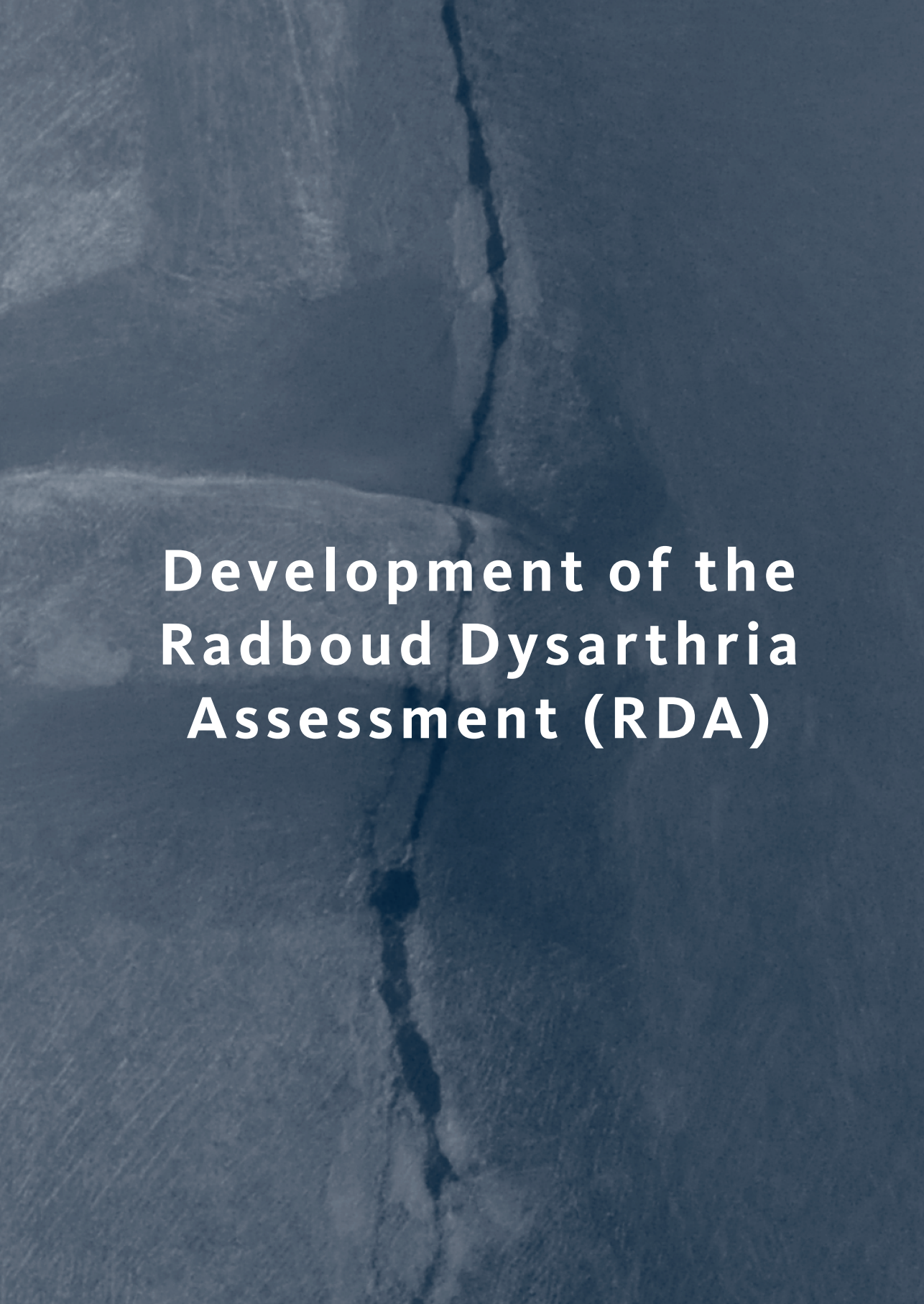
RECOMMENDATIONS FOR FUTURE RESEARCH

Despite their potential limiting effect on quality of life and general health condition, there are hardly any studies focusing on the effects of dysarthria and dysphagia. We would like to propose that both dysarthria and dysphagia are of utmost importance in NMD patients and treatment strategies with respect to these impairments should be the topic of well-designed RCTs. Preferably, these trials should be performed in homogenous patient samples, with comparable levels of impairments. Conclusions of those trials might be used in other groups of NMD patients, with similar impairments at the bulbar level. However, these remarks should not discourage researchers to perform less ambitious research, for example observational cohort studies or controlled trials. Results of such trials also add to the required level of evidence of SP.

As for the interventions, it is important that all patients are given standardized interventions and that SP interventions are being properly described. It should be noted that the feasibility of interventions depends on the type of intervention. Swallowing techniques are internationally uniform and well described in the literature.[73] However, there is less uniformity regarding techniques to improve intelligibility, although interventions like decreasing speaking rate or producing a louder voice can be objectively described in syllables per second or by measuring sound levels. Interventions regarding augmentative and alternative communication and instruction of the patient and relatives are tailor made to such an extent that they cannot be standardized. With regard to the type of outcome measures, we recommend using videofluoroscopy or flexible endoscopic evaluation of swallowing to objectively evaluate swallowing. Also, other quantitative measurements like the timed test[37] and the dysphagia limit[36] can be easily used as objective outcome measures. Secondary outcome measures like body weight, percentage of oral intake and quality of life can be valuable. For evaluating speech at the level of activities, we recommend using a speech intelligibility test.[22,74,75] At the level of participation, a scale of communicative participation is being developed.[76] Acoustic measurements are the most objective measurements to evaluate speech at the level of body functions. It should be kept in mind, however, that acoustic measurements have limited

predictive value with regard to communication effectiveness.[77-79] Therefore, we recommend using the acoustic measurements only in combination with measurements on the levels of activities or participation.
A consensus meeting with experts on speech pathology, neurology and methodology is recommended to further discuss and agree on the outlines of future trials. Previous meetings along these lines and covering various neuromuscular topics, such as on myotonic dystrophy, have been very successful.[80]

Part II



Development of the Radboud Dysarthria Assessment (RDA)

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5 The Radboud Dysarthria Assessment: development and clinimetric evaluation

ABSTRACT

Objective

In the absence of an adequate dysarthria assessment in the Netherlands, we developed the Radboud Dysarthria Assessment (RDA). This article describes its development and clinimetric evaluation.

Patients and methods

Forty-three patients were assessed with the RDA. The recording forms were subjected to exploratory factor analysis and estimation of internal consistency. The self-evaluation questionnaire was tested for internal consistency and the severity scale for intra- and inter-rater reliability. Construct validity of the severity scale and questionnaire was determined by relating them to the Speech Handicap Index (SHI), Dutch sentence intelligibility assessment (NSVO-Z), and category fluency task.

Results

Exploratory factor analysis extracted 4 factors (articulation, resonance, phonation, respiration/prosody) yielding an explained variance of 70.3%. Each factor showed good internal consistency (Cronbach's α = 0.89 – 0.91). The self-evaluation questionnaire showed excellent internal consistency (Cronbach's α = 0.90). Intra-class correlation coefficients of the severity scale (0.85 – 0.86) showed good reliability. The severity scores and self-evaluation questionnaire correlated substantially to strongly with the SHI (r_s = 0.40 and 0.80) and substantially with the NSVO-Z (r_s = -0.65 and -0.52).

Conclusions

The RDA is a valid and reliable tool, but further investigation is needed to demonstrate whether this instrument can successfully support speech-language therapists in correctly diagnosing the type of dysarthria.

INTRODUCTION

Dysarthria is a common feature of both central and peripheral neurological diseases, including stroke, brain trauma, neurodegenerative and neuromuscular disorders. Many speech-language therapists (SLTs), especially those working in acute care, nursing homes and rehabilitation centres, are regularly confronted with patients who are speaking effortful, who are poorly intelligible, or who sound abnormal. In these patients, it is important to identify whether the speech disorder can indeed be classified as dysarthria and, if so, what the type and severity of the dysarthria is. This involves insight in the neurological abnormalities regarding the 5 aspects of speech production, i.e., articulation, resonance, phonation, respiration and prosody[9] through proper assessment. In addition, diagnosing the type of dysarthria may support the medical diagnosis, because a speech disorder often reflects the localization of neurological dysfunction.[6,81] Lastly, a reliable judgment of the severity of dysarthria is needed for adequate monitoring and treatment evaluation.

Unlike aphasia tests, the clinical assessment of dysarthria is typically based on subjective judgments that are often less reliable than objective judgments. The subjective judgment of dysarthric speech is complex and notoriously difficult, because the 5 aspects of speech production interact with each other (e.g. the breathing pattern influences the vocal quality).[81] On top of that, speech can be influenced by other factors, like cognition, behavior, dentition, and emotion.[82,83] Based on our experience of providing post-graduate dysarthria courses for more than 10 years, we felt that many discussions about treatment are based on inadequate assessment of the dysarthria, resulting in misinterpretation of the dysarthric speech characteristics. We also learned that most SLTs use their own speech tasks and checklists to assess dysarthria. Hence, we felt there was a need for a proper and validated dysarthria assessment, which should be supported by video examples to facilitate the interpretation of the various speech characteristics.

As far as we could find in the international literature, the only published standardized diagnostic instrument for dysarthria in adults is the Frenchay Dysarthria Assessment (FDA-2).[84] The FDA-1 was translated and made available in the Netherlands between 1996 and 2010,[85] but we learned from our courses and our nationwide contacts that it never became widely used. When the Dutch version of the FDA-1 was no longer for purchase, we decided to improve and validate the Radboud Dysarthria Assessment (RDA) for adult patients that we made available online in 2007[34] and that we promoted during our post-graduate dysarthria courses. The original RDA was developed at the department of Rehabilitation of the Radboud University Medical Centre, based on the international literature and our clinical practice. It consisted of two components: a qualitative recording form and a severity scale, accompanied by a short manual with instructions how to perform and interpret the speech tasks (spontaneous speech, text reading, maximum repetition rate, maximum phonation time, maximum phonation volume and fundamental frequency range). Since its release in 2007, many SLTs in the Netherlands and Flanders have downloaded this freely available assessment.

We did not translate or promote the FDA-2 for of a couple of reasons. First, the FDA-2 combines observation of oral structures and nonverbal oral functions, assessment of speech characteristics, and measurement of intelligibility. Because SLTs in the Neth-

erlands have an orofacial examination at their disposal[86] as well as validated Dutch intelligibility tests on word and sentence levels,[22,23] there was no need for an instrument combining all these domains. Second, because the FDA-2 combines several domains, the assessment of speech characteristics is less extensive than we think is needed for a clinician to adequately assess dysarthria. Third, in the FDA-2, the dysarthria types are less specific than clinically required. For instance, hypokinetic and hyperkinetic dysarthrias are both categorized under 'extrapyramidal dysarthria', whereas these are clearly two distinctive disorders. Moreover, in the FDA-2, 'mixed dysarthria' is applied as a general term, although mixed flaccid/spastic dysarthria as in amyotrophic lateral sclerosis is clearly different from (and requires other a different therapeutic approach than) for example mixed hypokinetic/ataxic dysarthria as in multiple system atrophy.

Our 4-step approach to improve and validate the RDA was to: (1) seek national consensus for the tasks and the qualitative recording form, (2) add a self-evaluation questionnaire, (3) critically evaluate the clinimetric properties of the qualitative recording form, severity scale and self-evaluation questionnaire, and (4) add training videos of all types and severities of dysarthria including detailed interpretation of the assessment for self-training purposes.

This paper aims to describe the improvement of the original RDA for adult patients (step 1 and 2) and to report the dimensionality of the qualitative recording form (i.e., whether it is consistent with the five aspects of speech production), the internal consistency within each dimension and of the self-evaluation questionnaire, the construct validity of the severity scale and of the self-evaluation questionnaire, and the intra- and inter-rater reliability of the severity scale (step 3). The study that focused on the construction and usefulness of training videos (step 4) to improve the assessment of the type of dysarthria will be published in a separate paper.

METHODS

Approval was obtained from the Medical Ethics Committee of the Radboud University Medical Center and all patients signed an informed consent.

Improvement of the original RDA (step 1 and 2)

The process of improving the original RDA was led by an expert group of 7 SLTs working in four different hospitals in the Netherlands, 4 of them with more than 30 years of experience in motor speech disorders. The qualitative recording form of the freely available and unvalidated original RDA was thoroughly scrutinized by the expert group based on their experience of using it for a couple of years. A major adaptation was to structure the form according to the 5 aspects of speech production and to add information about posture and other speech-related aspects like dentition. Besides, speech characteristics were described more consistently regarding problems in strength, speed, range, tone, and precision of movements. Face validity of the recording form was established as follows. All items were described in detail in the test manual. The members of the expert group tested the recording form in clinical practice and their comments were collected and discussed until consensus about the recording form was reached. To create even broader support, a Delphi-method was used to ask for comments and feedback on the recording form and test manual from SLTs in the Netherlands and Belgium. 177 SLTs were contacted, and 49 (27.7%) agreed

to participate. These 49 colleagues worked in different settings (either in an [academic] hospital, rehabilitation center, nursing home, or private practice) and 58% of the respondents had more than 10 years of experience with neurological patients. Everyone readily agreed on the speech tasks, but much more time than we anticipated was needed to reach consensus on the terminology in Dutch. We named speech characteristics based on how they were referred to in the international literature,[6,84] but we experienced that many SLTs used their own terminology. After discussing all comments in the expert group and defining all terms used, the second versions of the recording form and test manual were constructed. In this process, we also used the feedback obtained after presentations of preliminary versions at international congresses.[87,88] Lastly, 3 independent experienced SLTs with a special interest in motor speech disorders thoroughly reviewed the second versions of the test manual and recording form. Based on their feedback, the final version of the RDA was constructed.

The RDA focuses on dysarthria, i.e. on the process of execution of speech. The aims of the RDA are to recognize the speech characteristics that lead to a particular type of (or mixed) dysarthria and to assess the severity of dysarthria. This requires the use of relevant speech tasks, of which spontaneous speech and reading are most functional and representative of daily life. There is a lively discussion about including maximum performance (speech-like) tasks in dysarthria assessment.[89] According to some authors speech and speech-like tasks are controlled by separate motor control systems,[42,90] whereas others believe that motor control of speech and speech-like tasks overlap or are, at least clinically, indistinguishable.[89,91] We decided to include maximum performance tasks primarily to facilitate the interpretation of specific characteristics observed in spontaneous speech and reading, such as signs of hypotonia, hypertonia, hypokinesia, hyperkinesia, and ataxia. Indeed, patients need to be challenged to examine the upper limits of especially phonation and articulation[33] to observe their voice and speech capacities without compensations. This is because compensatory strategies are harder to use in maximum performance tasks, thereby uncovering the true nature of the motor disorder. In addition, some types of dysarthria are characterized by a specific pattern in maximum performance tasks. For example, oral dysdiadochokinesis is a feature of ataxic dysarthria,[6,92] which manifests itself or becomes more prominent during a maximum repetition task. Another argument in favor of including speech-like tasks is that maximum performance tasks are effort-based that can help to identify therapeutic options. For instance, the often-normal maximum phonation volume in hypokinetic dysarthria reveals the voice capacity that is needed for successful training. Therefore, the following tasks were included in the RDA:

- ***Spontaneous speech by a semi-structured interview.*** Spontaneous speech is the task most representative of speaking in daily life and was, therefore, selected as the starting point of the assessment of speech characteristics.
- ***Text reading.*** When reading out loud, prosodic features like speaking rate and intonation become evident. Besides, fatigue can be provoked when the text is long enough, because there are no interruptions. A fictional reading text ('Niets' [93]), already used by many Dutch SLTs, was added after being adapted for acoustic measurements. In the Netherlands, clinicians generally

do not use acoustic measurements, but with this adaptation the text was made suitable for a more profound clinical examination as well as for research purposes. We paid particular attention to the correct position of plosives and vowels within words (in stressed or non-stressed syllables) and within sentences, taking into account the normal phonological processes.[94,95] Based on these criteria, sentences were rewritten and the final text now consists of 520 words.

- ***Maximum repetition rate (syllables/second).*** MRR of /pa/, /ta/, /ka/, and /pataka/ was included to assess the upper limits of the articulatory system.
- ***Maximum phonation time (seconds).*** MPT or sustained phonation on /a:/ was included to specifically assess the phonatory-respiratory systems. To distinguish the phonatory from the respiratory aspects of speech production, the sustained productions of /s/ and /z/ can be compared and the s/z ratio can be calculated, with a ratio of >1.4 indicating phonatory problems.[96] In the case of respiratory problems, both sustained /s/ and /z/ will be shorter than normal.
- ***Maximum phonation volume (dB).*** MPV was included to examine the dynamic range by asking the patient to produce "*Hallo!*" (Hello!) and "*Kom hier!*" (Come here!) as loud as possible. A decline in dynamic range may not be detected in spontaneous speech, because in normal conversation only a small part of the full range is used. Besides, if a loud voice is possible during MPV in a patient with hypokinetic dysarthria, it makes weakness unlikely and shows spare capacity that can be utilized for therapy.
- ***Fundamental frequency range (semitones).*** FFR was included to examine the melodic range by asking patients to produce an /a:/ from the lowest possible to the highest possible pitch and vice versa. Like in MPV testing, a decline of the FFR may not be detected in spontaneous speech when only a small part of the range is used.

All maximum performance tasks are performed 3 times and the best performance is rated.

In parallel with the improvement of the RDA, we collected maximum performance reference values of 224 men and women[97] to be able to compare the performance of each patient with the normal population. However, when clinicians do not assess the maximum performance tasks instrumentally (e.g. with PRAAT), they can score the performance of the patient subjectively on a 4-point Likert scale ranging from '0 = normal' to '3 = severe problems/impossible'. All other speech characteristics are rated on the recording form with this 4-point scale as well. When scoring ≥ 1, a qualitative judgement has to be given, which is printed on the qualitative recording form. In Appendix 5.1, the recording form is shown. It includes all relevant items to assess the 5 aspects of speech production. We translated the form from Dutch into English only for the purpose of this article and would like to emphasize that the wording is merely indicative of the Dutch original. In the Dutch manual of the RDA, all terms are explained in detail.

For the judgement of the severity of dysarthria, we included a 6-point scale inspired by the Therapy Outcome Measures[98] ranging from '0 = normal' and '5 = most severe'. The scores 1 to 5 were defined by applying the terminology of the qualitative recording form (Appendix 5.1). This severity score is an overall score that reflects the interpre-

tation of the performance on all different tasks and it is given after completion of the full assessment. If a patient scores mostly 0 or 1 on the qualitative recording form, the overall severity is considered to be mild. The overall severity score reflects the most severely affected aspect of speech production, so when, for example, the articulation is more severely affected than the phonation, the severity score will reflect the severity of the articulation.

Lastly, we included a short self-evaluation questionnaire to quantify the patient's speech complaints and experienced consequences in daily functioning, but also as a clinical guidance for the history taking. We adjusted the Radboud Oral Motor inventory for Parkinson's disease (ROMP-speech)[99] by replacing two items that were specific for hypokinetic dysarthria (vocal quality and starting problems) by items for all types of dysarthria (ability to speak for a longer period and ability to raise the voice). The self-evaluation questionnaire consists of seven questions with a 5-point response scale that cover the "function", "activity" and "participation" domains of the ICF 21, yielding a total score from 7 (no complaints) to 35 (most severe complaints).

Clinimetric evaluation of the RDA (step 3)

Table 5.1 provides an overview of the clinimetric evaluations that we used to validate the different parts of the RDA.

Patients

Forty-three adult patients with dysarthria were recruited from the 4 hospitals to which the members of the expert group were affiliated. No specific inclusion or exclusion criteria were applied, because the aim was to collect examples of all dysarthria types and severities.

Raters

Usually, the expert SLT who included the patient also made the video recordings of the dysarthria assessment, with a high definition camera and a separate microphone. After completion of all videos, the expert group discussed the type and severity of the dysarthria of each patient until full agreement was reached, while they were kept unaware for the clinical information (medical diagnosis etc.).

Table 5.1. Overview of the clinimetric evaluations.

Clinimetric characteristic	Based on judgement or scores by:
Qualitative recording form	
Face validity	Expert group
Dimensionality	Expert group
Internal consistency	Expert group
Severity scale	
Construct validity	Expert group
Reliability	SLTs
Self-evaluation questionnaire	
Construct validity	Patients
Internal consistency	Patients

Forty-six SLTs were approached by e-mail to ask for participation. This e-mail was sent to a study group of SLTs working in hospitals across the Netherlands and to a group of SLTs working with neurological patients in the region of Nijmegen. Twenty-two of them (48%), with on average 11 years of experience with dysarthria (range 2–24 years), agreed to participate.

Construct validity

To evaluate the construct validity of the severity scale (severity score given by the expert group) and the self-evaluation questionnaire, the scores were compared with (a) the Speech Handicap Index (SHI)[100], (b) the Dutch sentence intelligibility assessment (NSVO-Z)[23], and (c) a category fluency task[101]. The SHI and NVSO-Z were used to test convergent validity, whereas the category fluency task was used to test divergent validity as it assesses linguistic skills rather than speech. The NSVO-Z and the category fluency task were performed and videotaped after the assessment with the RDA by the expert SLT. Patients completed the self-evaluation questionnaire at home, maximally 1 week before the assessment. The expert SLT who performed the assessment of the RDA, NSVO-Z, and category fluency task was unaware of the patient's scores on the SHI and the self-evaluation questionnaire.

The SHI is a validated self-reported Dutch questionnaire for dysarthric patients containing 15 questions about dysarthria in the physical, functional and emotional domains. Patients answer the SHI questions on a 5-point-scale from 'never' to 'always' (0–4) generating a total score ranging from 0 to 60. The NSVO-Z contains 1200 semantically unpredictable sentences. For each patient, 18 sentences are randomly generated, which have to be read out loud. A member of the expert group, who was not responsible for videotaping the patient, transcribed all sentences spoken by the patient before discussing the type and severity of the dysarthria. The first author (S.K.) compared the transcription to the original sentences, resulting in a percentage of intelligibility for each patient. The category fluency task requires naming as many animals as possible in 1 min. The total number of animals is converted into a Z-score dependent on age and level of education.[101] When a slow speaking rate is responsible for a limited number of animals, correction is allowed. We hypothesized substantial to strong correlations ($r_s > 0.40$)[102] of the severity scale and the self-evaluation questionnaire with the SHI and the NSVO-Z, whereas weak correlations ($r_s < 0.40$) were expected with the category fluency task.

Reliability

Twenty-two SLTs agreed to score a set of videos at two separate times. Three SLTs scored 10 selected videos twice, with a 2-week interval, to assess the intra-rater reliability of the severity scale. The videos included minimally 1 example of each severity level and the SLTs scored the videos independently using a secured online connection. The other 19 SLTs scored 10 selected videos before and after an online video-training. Inter-rater reliability of the severity scale was determined by comparing their scores of the second assessment (after the online training).

Dimensionality and internal consistency

The qualitative recording forms filled in by the expert group during the consensus process were used to assess the dimensionality and internal consistency of the recording form. The self-evaluation questionnaires completed by the patients were used to calculate the internal consistency of the questionnaire.

Statistics

An exploratory factor analysis was performed to evaluate whether the items on the qualitative recording form loaded on the factors that corresponded with the a priori defined five aspects of speech production (articulation, resonance, phonation, respiration and prosody). Principal axis factoring was used as the extraction method together with varimax rotation. Extraction of the factors was based on the Kaiser's criterion for Eigenvalues being equal to or greater than 1.0. Cronbach's α was used to calculate the internal consistency of each factor found and to test the internal consistency of the self-evaluation questionnaire.

Construct validity was determined by calculating Spearman's correlation coefficients of the severity scale and the self-evaluation questionnaire with the SHI, NSVO-Z, and the category fluency task. We a priori considered (absolute) correlation coefficients < 0.40 as 'weak', 0.40 to 0.70 as 'substantial', and > 0.70 as 'strong'.[102]

The Intraclass Correlation Coefficient (ICC) for consistency (two-way random) was used to calculate the intra- and inter-rater reliability of the severity scale. A value of 0.70 was considered as a minimum standard for reliability.[103]

Calculations were performed with IBM SPSS Statistics 20 for Windows (IBM Corp., Armonk, NY) and SAS 9.2 for Windows (SAS Institute, Cary, NC).

RESULTS

We included 18 women and 25 men with a mean age of 61 years (range 14–79 years). Table 5.2 provides an overview of the type and severity of dysarthria in these 43 patients (including their medical diagnosis). Their mean SHI-score was 24 (range 6–49), the mean NSVO-Z score 86% (range 34–100) and the mean ROMP-score 18 (range 9–35).

Qualitative recording form

The exploratory factor analysis extracted four dimensions yielding an explained variance of 70.3% with all items loading on 4 components (Table 5.3). Those components could be identified as: phonation (1), articulation (2), respiration/prosody (3) and resonance (4). Cronbach's α was between 0.89 and 0.91 for the individual factors, indicating good internal consistency within each dimension.

Severity scale

The scores of the 19 SLTs were consistent with the expert group for 53.2%. The correlation of dysarthria severity with both the SHI (r_s = 0.40, p < 0.01) and the NSVO-Z (r_s = -0.65, p < 0.01) was substantial, whereas the correlation of dysarthria severity with the category fluency task was weak (r_s = -0.28, p = 0.05) (Table 5.4). Both the inter-rater and the intra-rater reliability of the severity scale were high (ICCs 0.85 or 0.86).

Self-evaluation questionnaire

The internal consistency of the self-evaluation questionnaire was high (Cronbach's α = 0.90). The correlation of the self-evaluation questionnaire with the SHI was strong (r_s = 0.80, p < 0.01), with the NSVO-Z substantial (r_s = -0.52, p < 0.01), and with category fluency weak (r_s = -0.27, p = 0.06) (Table 5.4).

DISCUSSION

In this paper, we present the RDA, a standardized set of common speech and maximum performance (speech-like) tasks for the perceptual analysis of speech, using a qualitative recording form, a severity scale

Table 5.2. Overview of the type and severity of the dysarthria and the medical diagnosis of the 43 patients.

Dysarthria severity*	Dysarthria type					
	Flaccid	**Spastic**	**Ataxic**	**Hypokinetic**	**Hyperkinetic**	**Mixed**
1					Dystonia (*n* = 1)	
2	Stroke (*n* = 2)	CP (*n* = 1)	Cerebellar ataxia (*n* = 2)		Dystonia (*n* = 1)	
	FSHD (*n* = 1)		MS (*n* = 1)		Huntington's disease (*n* = 1)	
	MD (*n* = 1)		MSA (*n* = 1)			
3	Stroke (*n* = 5)		Cerebellar ataxia (*n* = 3)	PD (*n* = 1)	Dystonia (*n* = 1)	TBI (*n* = 1)
	MG (*n* = 1)					MS (*n* = 1)
	PLS (*n* = 1)					Stroke (*n* = 2)
4	Stroke (*n* = 2)	CP (*n* = 1)		PD (*n* = 2)	Dystonia (*n* = 1)	Stroke (*n* = 2)
				MSA (*n* = 1)	Huntington's disease (*n* = 1)	PLS (*n* = 1)
				MSA (*n* = 1)		TBI (*n* = 1)
						PSP (*n* = 1)
5						Unknown (*n* = 1)

* 1 = minimal dysarthria; 2 = mild dysarthria; 3 = mild/severe dysarthria; 4 = severe dysarthria; 5 = very severe dysarthria. Abbreviations used: TBI: traumatic brain injury; CP: cerebral palsy; PD: Parkinson's disease; PLS: primary lateral sclerosis; MSA: multiple system atrophy; PSP: progressive supranuclear palsy; MS: multiple sclerosis; FSHD: facioscapulohumeral muscular dystrophy; MD: myotonic dystrophy; MG: myasthenia gravis.

Table 5.3. The factor loadings of the different items of the rating sheet on the four components.

Items	Component*			
	1	2	3	4
Lip movements	.55	**.60**		
Jaw movements	.51	**.43**		
Tongue movements		**.65**		
Vowels		**.73**		
Consonants		**.80**		
Clusters		**.81**		
Syllable structure		**.79**		
Oral diadochokinetics		**.72**		
Resonance				**.83**
Vocal quality	**.82**			
Vocal use	**.79**			
Loudness	**.66**			
Pitch	**.70**			
Fundamental frequency range	**.82**			
Maximum phonation volume	**.64**			
Maximum phonation duration	**.57**		.55	
Inhalation			**.59**	
Breathing pattern	.41		**.75**	
Breath groups	.44		**.68**	
Melodic accent	.52		**.53**	
Dynamic accent	.53		**.64**	
Temporal accent			**.67**	
Cronbach's α	*.89*	*.90*	*.91*	

* Factor loadings suppressed below 0.4. Bold numbers indicate the highest factor loading per item.

Table 5.4. The Spearman's correlation coefficients (p-value) between the severity of dysarthria, the self-evaluation questionnaire, the Speech Handicap Index (SHI),[100] intelligibility (NSVO-Z)[23] and the category fluency task in the 43 patients.

	Dysarthria severity	SHI	NSVO-Z	Fluency
Dysarthria severity	1.00 (n.a.)	0.40 ($p < 0.01$)	-0.65 ($p < 0.01$)	-0.28 ($p = 0.05$)
Self-evaluation questionnaire	0.53 ($p < 0.01$)	0.80 ($p < 0.01$)	-0.52 ($p < 0.01$)	-0.27 ($p = 0.06$)

Abbreviation used: n.a., not applicable.

and a self-evaluation questionnaire. We established its clinimetric properties in adult Dutch speaking persons, but the RDA itself is not language-specific and easy to construct in any other language.

Factor analysis indicated that the items on the recording form represent 4 domains of speech production: phonation, articulation, respiration/prosody and resonance. We had expected that prosody would be identified as a separate dimension, but our results demonstrated that it proved to be part of the respiration domain. This is conceivable as it is a supra-segmental speech aspect. Because the internal consistency within each dimension was good, it is safe to conclude that the items within each domain were adequate.

The severity scale showed good intra- and inter-rater reliability scores, indicating that it is sufficiently accurate to become used in clinical practice by SLTs. Regarding the validity, the fact that the correlation between dysarthria severity (which is clinician-rated) and the SHI (which is patient-rated) was substantial ($r_s \geq 0.40$) but not strong ($r_s \geq 0.70$) is plausible. Whether patients experience problems due to dysarthria as indicated by the SHI is highly personal and depends on their daily activities. This emphasizes that collecting clinician-rated scores as well as patient reports is important for setting shared treatment goals.[104]

The substantial ($r_s = -0.65$) (but not strong) correlation between dysarthria severity and the NSVO-Z is conceivable as well. Patients with severe dysarthria according to the RDA can still be intelligible according to the NSVO-Z, if they adapt their speaking technique.[6] In line with our hypothesis, dysarthria severity correlated only weakly with category fluency, because the latter is dependent on linguistic ability rather than speech capacity. Overall, the results support the construct validity and reliability of the severity scale.

The self-evaluation questionnaire showed good internal consistency, indicating that all questions measure the same construct. The strong correlation between the self-evaluation questionnaire and the SHI indicates that SLTs can choose either questionnaire to obtain information about the speech difficulties as perceived by the patient. The weak to substantial correlations with category fluency and the NSVO-Z further underline that the self-evaluation questionnaire measures a specific and patient-rated construct.

The improved RDA was based on national consensus and was, therefore, readily accepted as the preferred dysarthria assessment in the Netherlands according to sales figures and feedback during our post-graduate dysarthria courses. By striving to achieve consensus about the tasks,

the terminology of the recording form, and about the speech characteristics of the 43 patients, we learned at least two important lessons. We needed all tasks, including the maximum performance tasks, to reach consensus, in particular to distinguish primary deficits from compensatory mechanisms that patients used to optimize their speech. At the same time, we did not anticipate that reaching consensus with highly experienced clinicians would be such a time-consuming challenge. It seems that experienced clinicians all have their own internal acoustic anchors, which makes it even more important that we managed to reach consensus for the use of our instrument in the Netherlands and Flanders.

We primarily aimed at publishing a concise perceptual clinical assessment to be used by any dedicated SLT in daily practice. Nevertheless, by adding normative values for the maximum performance tasks[97] and a reading text adapted for acoustic measurements, the RDA is also suitable for scientific purposes, for instance for the objective registration and analysis of speech tasks. During our study, there was hardly any dysarthria assessment that we could use as a reference. Recently, the German instrument Bogenhausen Dysarthria Scales (BoDys) has been published,[105] which is also a fully perceptual assessment, but with 4 tasks to elicit connected speech only (conversational speech, sentence repetition, text reading, and picture story). Because this instrument is based on a different theoretical approach that strictly distinguishes speech motor control from the motor control of speech-like tasks,[89] a direct comparison between the RDA and BoDys is not possible.

Future perspectives

The RDA has been developed for assessing adult neurological patients, but a pediatric version is underway with speech tasks adapted to children aged from 4 to 18 years, including age-related reference values for the maximum performance tasks. Because the RDA includes internationally accepted speech tasks and terminology, the instrument can easily be translated into other languages, with two possible limitations: every language may need its own set of videos with all dysarthria types and severities and only future data gathering can demonstrate whether or not the reference values of the speech tasks are language-dependent.[106]

Acknowledgements

The authors would like to thank all patients and SLTs for their participation. In particular, Harry Goos, Judith Kocken, Lotte Kromhout, Marjo van Gerven and Puck Goossens are acknowledged for providing their expertise during the development of the RDA. We thank prof. dr. A. Rietveld and Roos Haasnoot for their participation in the adaptation of the reading text and dr. J.C.M. Hendriks for his statistical support.

APPENDIX

Radboud Dysarthria Assessment (RDA)

Name patient:	Date:
Date of birth:	SLT:

0 = normal 1 = minimal problems 2 = mild/clear problems 3 = severe problems/impossible

Articulation (spontaneous speech, reading, maximum repetition rate)

Lip movements:	0 - 1 - 2 - 3	O small O slow O asymmetric in disadvantage to left / right	O tense O flaccid	O uncontrolled O involuntary movements		
Jaw movements :	0 - 1 - 2 - 3	O small O slow	O tense O flaccid	O involuntary movements		
Tongue movements:	0 - 1 - 2 - 3	O small O slow O asymmetric in disadvantage to left / right	O tense O flaccid	O uncontrolled O involuntary movements		
Vowels:	0 - 1 - 2 - 3	O distorted	O prolonged	O shortened		
Consonants:	0 - 1 - 2 - 3	O flaccid	O tense	O imprecise	O prolonged	
Clusters:	0 - 1 - 2 - 3	O schwa- insertion		O reduced	O imprecise	
Syllable structure:	0 - 1 - 2 - 3	O phoneme-/syllable repetitions		O phoneme-/syllable omissions		

Reading better/worse than spontaneous speech: .....

Changed quality of articulation over time: improvement / deterioration .....

Maximum repetition rate	pa	0 - 1 - 2 - 3	O flaccid O tense	O slow O dysrhythmic	O problems increase
	ta	0 - 1 - 2 - 3	O flaccid O tense	O slow O dysrhythmic	O problems increase
	ka	0 - 1 - 2 - 3	O flaccid O tense	O slow O dysrhythmic	O problems increase
	pataka	0 - 1 - 2 - 3	O flaccid O tense	O slow O dysrhythmic	O disturbed pattern O problems increase

Observations .....

Resonance (spontaneous speech, reading)

Nasality:	0 - 1 - 2 - 3	O hyper	O hypo	O variable	O nasal emission

Reading better/worse than spontaneous speech: .....

Changed quality of nasal resonance over time: improvement / deterioration.....

Observations .....

Phonation (spontaneous speech, reading, fundamental frequency range, maximum phonation volume, maximum phonation duration)

Vocal quality:	0 - 1 - 2 - 3	O breathy O tremor O cracking	O harsh O staccato	O diplophonic O falsetto	O aphonic O unsteady
Vocal use:	0 - 1 - 2 - 3	O hypotonic	O hypertonic		
Loudness:	0 - 1 - 2 - 3	O increased	O reduced	O uncontrolled	
Pitch:	0 - 1 - 2 - 3	O high	O low	O uncontrolled	

Reading better/worse than spontaneous speech: .....

Radboud Dysarthria Assessment (RDA)

Changed quality of phonation over time: improvement / deterioration……

Fundamental freq range: 0 - 1 - 2 - 3 O reduced O uncontrolled

Max phonation volume: 0 - 1 - 2 - 3 O reduced O forced O strained

Max phonation time: 0 - 1 - 2 - 3 O impaired quality ……. O deviant duration: ……. sec.
s/z ratio: ……..

Observations ……

Respiration (during speech) (spontaneous speech, reading, maximum phonation volume, maximum phonation duration)

Place: O clavicular O costal O costo-abdominal O abdominal

Inhalation: 0 - 1 - 2 - 3 O irregular O stridor without phonation O stridor with phonation

Respiration during speech: 0 - 1 - 2 - 3 O short breath groups O short breath support O impaired coordination of breath and vocalisation

Pattern of breathing: 0 - 1 - 2 - 3 O too fast O forced O shallow O paradoxical

Reading better/worse than spontaneous speech: ……..

Observations: ……..

Prosody (spontaneous speech, reading)

Melodic accent: 0 - 1 - 2 - 3 O monopitch O unnatural O uncontrolled / excess

Dynamic accent: 0 - 1 - 2 - 3 O monoloudness O uncontrolled / excess

Temporal accent: 0 - 1 - 2 - 3 O slow O accelerations O variable O fast
O chanted O stop problems O start problems

Reading better/worse than spontaneous speech: ……..

Observations ……

Posture

Body: O normal O movements in extremities O movements in trunk
O flexion O extension O deviation to L / R O slumped

Head: O normal O rotation to L / R O movements with head
O flexion O extension O lateroflexion to L / R

Observations:……

Other

O apraxia of speech O dental problems, nl. ……
O physical capability ↓ O control of saliva ↓ O other. ……

Conclusion

Dysarthria:
__ no
__ yes* O spastic O ataxic O hypokinetic O hyperkinetic { O chorea/athetosis, O dystonia
O flaccid { O mononeuropathic, O myogene, O bulbar, O UUMN
O undetermined:……
Non-dysarthric components:……

* in case of mixed dysarthrias, tick two or more types

Radboud Dysarthria Assessment (RDA)

Severity scale:

0. No dysarthria
1. Minimal dysarthria: minimal problems with articulation, resonance, phonation or respiration.
2. Mild dysarthria: tonus, range of motion or speed of articulation movements are mildly affected, leading to mildly affected consonants and/or vowels. Mild problems with resonance, phonation or respiration.
3. Mild / severe dysarthria: tonus, range of motion or speed of articulation movements are clearly affected, leading to deviant consonants and/or vowels. Clearly affected resonance, phonation or respiration.
4. Severe dysarthria: tonus, range of motion or speed of articulation movements are severely affected, leading to severely affected consonants and/or vowels. Severely affected resonance, phonation or respiration. Very slow speech with only a few syllables per breath group.
5. Very severe dysarthria/anarthria: articulation movements are almost impossible, leading to mainly open vowels and very severely distorted consonants. Very severely affected resonance, phonation (aphonic) or respiration (almost no breath support).

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6 Reference values of maximum performance tests of speech production

ABSTRACT

Purpose

Maximum performance tests examine upper limits of speech motor performance, as used by speech-language pathologists in dysarthria assessment protocols. The Radboud Dysarthria Assessment includes maximum repetition rate, maximum phonation time, fundamental frequency range and maximum phonation volume to assist in detecting pathological performance. This study aims to obtain reference values for each of these tests.

Method

A group of 224 healthy Dutch adults aged 18–80 years performed the maximum performance tests. Age, sex, body height, smoking habit, and profession were registered. Using multivariable linear regression, a wide range of models was tested to examine the relationship between these person characteristics and speech performance. The Likelihood-Ratio was used to test the goodness of fit to the data.

Result

Above 60 years of age, maximum repetition rate, fundamental frequency range and maximum phonation volume were all negatively affected by age. Below 60 years, only women showed effects of age on fundamental frequency range (increase) and maximum phonation volume (decrease). Maximum phonation time was primarily related to body height (increase).

Conclusion

This study presents reference values of four maximum performance tests for comparing the performance of dysarthric patients with non-pathological performance. Age was identified as most important factor influencing maximum speech performance.

INTRODUCTION

Maximum performance tests of speech production examine the upper limits of speech motor performance[33] and are used in dysarthria assessment protocols by speech-language pathologists (SLPs) to investigate the articulatory and phonatory-respiratory systems more independently than in spontaneous speech.[6] Typically, in spontaneous speech all systems (articulatory, velopharyngal, phonatory and respiratory) are co-operating during highly variable speech patterns, while the maximum performance tests have a limited variability. In 2014, the Radboud Dysarthria Assessment (RDA) was published,[107] which includes four maximum performance tests of speech production: maximum repetition rate (MRR), maximum phonation time (MPT), fundamental frequency range (FFR) and maximum phonation volume (MPV). Despite the ongoing debate about motor control in speech versus non-speech tasks,[42,89,108,109] we think that maximum performance tasks are of utmost importance in clinical dysarthria assessment. The most important reason is that, compared to spontaneous speech, repetitive speech patterns are less variable and, thus, easier to judge. Other reasons to include maximum performance tests in the RDA are the following. First, in spontaneous speech, a person with dysarthria can compensate speech motor deficits, for example, by slowing down the speaking rate. During maximum performance tests, such compensatory strategies are much harder to use, resulting in a more realistic expression of the different capacities of the articulatory and phonatory-respiratory systems. Second, maximum performance tests can help SLPs with distinguishing different types of dysarthria. For example, a dysrhythmic MRR is a specific feature of ataxic dysarthria[6,92,110] and hypokinetic dysarthria is characterized by a normal MRR with reduced amplitude of the articulatory movements.[92] Finally, maximum performance tests can help to identify therapeutic options. For example, a high MPV in a patient with hypokinetic dysarthria reveals the voice capacity that is needed for successful training.

To distinguish pathological from non-pathological speech performance and to obtain an indication of the severity of pathological performance, reference values are needed. These data are partially available in the literature,[33,106,111] but reference values from a sizable population with a clinically relevant age span are lacking. In addition, maximum speech and voice capacity may be related to language or culture. Indeed, Icht and Ben-David[106] recently showed significant differences in MRRs between English, Portuguese, Farsi and Greek speaking persons. Therefore, the purpose of this study was to collect reference values for MRR, MPT, FFR and MPV in a large population of healthy Dutch adults and relate these to relevant person characteristics.

METHODS

Participants

We included 224 healthy native Dutch speakers (108 men and 116 women) aged 18–80 years. They were recruited by the investigators from the local community. Participants with a history of any swallowing, speech or voice problem were excluded. The participants were divided into age groups of one decade (18–29, 30–39, 40–49, 50–59, 60–69, 70–80) and we aimed to include a more or less equal number of participants per age group. We collected the following person characteristics that might influence maximum performanc-

es: age (years), sex (men/women), body height (cm), smoking habit (yes/no), and professional occupation. Age and sex were registered as basic person characteristics. In addition, body height was registered because of its effect on lung capacity[112] and smoking habit and professional occupation because of their known effect on the quality of the voice.[113,114] Based on profession, the level of vocal use was categorized by the classification of Koufman and Isaacson: (I) elite vocal performer (singers and actors), (II) professional voice user (teachers, receptionists), (III) non-vocal professionals (doctors, lawyers) and (IV) non-vocal non-professionals (students, laborers).[115]
All participants signed informed consent before participating in the study. We obtained approval from the Committee on Research Involving Human Subjects of Arnhem and Nijmegen.

Speech measurements

The participants performed all speech tasks three times, in similar order, while sitting upright. All performances were recorded with a linear PCM recorder (Tascam DR-05, Tokyo, Japan) and the best maximum performance was used in the statistical analysis. Five trained examiners recruited and instructed the participants and recorded all performances. The examiners worked in pairs, but the participant was assessed by just one examiner. The specific tasks were described as follows.

Maximum repetition rate
The participants were instructed to repeat the monosyllabic sequences /pa/, /ta/ and /ka/ and the trisyllabic sequence /pataka/ as fast as possible for at least 6 seconds. MRR was analyzed with Praat[116] and expressed in syllables per second. The count-by-time method was used during the first 5 seconds of the sequence.[117]

Maximum phonation time of /a:/
The participants were instructed to produce an /a:/ as long as possible after taking a maximal inhalation, at a comfortable pitch and at their habitual loudness. MPT was analyzed with Praat and expressed in seconds.

Fundamental frequency range
The participants were instructed to produce an /a:/ from the lowest possible to the highest possible pitch and vice versa. Producing a musical scale was also allowed. People who experienced difficulties while performing this test were stimulated to produce only their lowest and highest pitches. FFR was analyzed with Praat and expressed in Hz. FFR was converted from Hz to semitones using the formula: $ST = 39{,}87 * \log(F/50)$.[95]

Maximum phonation volume
The participants were instructed to produce "*Hallo!*" (Hello!) and "*Kom hier!*" (Come here!) as loud as possible. MPV was measured with a dB-meter (Voltcraft SL-100, Hirschau, Germany) at 30 cm distance from the mouth, which was standardized by using the A4 assessment form.

Statistical methods

First, we used univariate analysis to explore the association between each maximum performance task and each person characteristic using Pearson correlation coefficients (age and body height), Spearman correlation coefficients (profession), and independent-samples t-tests (sex and smoking habit) to identify possibly influential person characteristics. Regarding MRR (articulation), we only explored the association with sex and age, whereas for MPT, FFR and MPV (voice), we explored the association with all person characteristics. Characteristics with a p-value of < 0.05 were selected for multivariate analysis (determinants). Second, multivariate

linear regression was used to study the unique influence of the identified determinants (independent variables) on each maximum task performance (dependent variable) separately. We searched for the independent variable with the strongest influence to be able to construct reference lines. Therefore, we studied a wide range of models for each maximum performance test: first- to third-degree polynomials in age and body height, piece-wise regression in age and height, interaction terms with sex, and untransformed and logarithmic transformed values of the performance tests. The Likelihood-Ratio was used to test differences between the models for their goodness of fit to the data. With respect to MPT, the dependent variable was the logarithmic transformed value of the MPT. The antilog-transformed results were calculated. For all other maximum performance tests, the dependent variable was the original performance.

A paired-samples t-test was used to test differences between the four individual sequences of the MRR (α-level: $p = 0.05$). All statistical analyses were performed using SAS 9.2 for Windows (SAS Institute, Cary, NC).

Table 6.1. Characteristics of all participants (*n*=224).

	median	**(range)**		
Age (year)	43	(18 – 80)		
Body height (cm)	175	(155 – 201)		
Weight (kg)	73	(50 – 120)		
	n	**(%)**	**men (n)**	**(%)**
Total	224		108	(48.2)
18-29 y	76		30	(39.5)
30-39 y	28		14	(50.0)
40-49 y	27		15	(55.6)
50-59 y	37		22	(59.5)
60-69 y	30		15	(50.0)
70-80 y	26		12	(46.2)
Smokers	68	(30.4)		
Profession (level of vocal use)*				
level I	3	(1.3)		
level II	63	(28.1)		
level III	74	(33.0)		
level IV	84	(37.5)		

* Level I: elite vocal performer; level II: professional voice user; level III: nonvocal professionals; level IV: nonvocal nonprofessionals.

Table 6.2. The observed median and range of the maximum performance tests of speech production by age group.

age group	*n*	MRR				MPT (s)	FFR (semitones)	MPV (dB)
		/pa/ (syl/s)	/ta/ (syl/s)	/ka/ (syl/s)	/pataka/ (syl/s)			
		median (range)	*median (range)*	*median (range)*	*median (range)*	*median (range)*	*median (range)*	*median (range)*
18–29 y	76	6.8 (5.4–9.2)	6.6 (4.0–9.1)	6.0 (3.8–7.7)	7.0 (4.1–9.0)	18.4 (6.6–54.0)	24.9 (14.2–45.6)	100.6 (93.5–102.5)
30–39 y	28	7.0 (6.2–8.5)	6.8 (5.7–8.2)	6.4 (4.5–7.3)	7.1 (5.0–8.5)	20.5 (11.1–55.5)	34.4 (13.5–48.0)	100.2 (93.0–103.0)
40–49 y	27	6.9 (5.1–8.2)	6.9 (5.4–8.3)	6.2 (4.2–8.0)	7.0 (5.0–9.8)	21.0 (10.5–42.9)	31.7 (16.5–46.2)	100.6 (93.8–101.7)
50–59 y	37	6.9 (5.0–7.7)	6.6 (4.8–8.2)	6.0 (4.5–7.2)	6.7 (4.8–8.4)	19.6 (6.9–49.9)	29.5 (14.6–47.1)	100.5 (93.1–102.0)
60–69 y	30	6.8 (5.3–8.3)	6.6 (5.0–7.9)	6.1 (4.3–7.5)	7.0 (5.3–8.5)	21.8 (9.0–55.5)	32.1 (14.8–44.6)	99.4 (89.5–102.0)
70–80 y	26	6.4 (4.3–7.2)	6.1 (4.2–7.5)	5.7 (4.0–7.3)	6.2 (4.3–7.9)	18.0 (8.4–27.8)	26.8 (16.4–45.7)	98.5 (77.0–104.1)
18–80 y	224	6.8 (4.3–9.2)	6.5 (4.0–9.1)	6.0 (3.8–8.0)	6.9 (4.1–9.8)	19.4 (6.6–55.5)	28.9 (13.5–48.0)	100.3 (77.0–104.1)

Abbreviations used: syl: syllable; s: second; y: year; MRR: dB: decibel.

RESULTS

A total of 224 participants (108 men and 116 women) were included with a mean age of 43 years (standard deviation [SD] = 19.0, range 18–80) and a mean body height of 175.5 cm (SD = 9.6, range 155–201). Sixty-eight participants (30.4%) were smokers and 66 (29.4%) were vocal professionals (level I and II, see Table 6.1). The age group 18–29 years was the largest for two reasons. Initially, we started including participants from 20 years old, but we extended the youngest age group from 20–29 years to 18–29 years, as adulthood starts at 18 years and the pediatric version of the RDA (under construction) reaches up to 17 years. Second, this youngest group initially seemed to score lower than expected. By including more participants, we intended to obtain a better representation of this age group.
Overall, smoking habit and profession (vocal use) did not influence the maximum performance tests, leaving age, sex and body height as independent variables for the multivariate regression analyses. When testing the models for their goodness of fit, the best fit was the piece-wise regression model with a cut-off point chosen at 60 years of age. Regression coefficients with 95% confidence intervals (95% CI) of the final models are presented. The 5th, 25th, 50th, 75th and 95th percentile reference lines are visualized in a graph. Reference lines are presented by sex when relevant.

Maximum repetition rate

The median and range of each MRR sequence are shown by age group in Table 6.2. Across all age groups, /ka/ was by far the slowest sequence (with 6.0 syl/s) and differed significantly from /pa/ ($p < 0.01$), /ta/ ($p < 0.01$) and /pataka/ ($p < 0.01$). /Pataka/ was the fastest sequence (with 6.9 syl/s) and differed, in addition to /ka/, significantly from /pa/ ($p = 0.04$) and /ta/ ($p < 0.01$). Finally, /pa/ was a significantly faster sequence than /ta/ ($p < 0.01$).
MRR was only significantly related to age. In Table 6.3, the estimated mean decrease

Table 6.3. The estimated mean change per year in MRR, FFR, and MPV using a piece-wise linear regression model with cut-off point at 60 years.

	< 60 years		≥ 60 years	
	mean	**(95% CI)**	**mean**	**(95% CI)**
MRR				
/pa/ (syl/s)	-0.00	(-0.01– 0.00)	-0.03	(-0.06 – -0.01)
/ta/ (syl/s)	-0.00	(-0.01 – 0.01)	-0.06	(-0.09 – -0.02)
/ka/ (syl/s)	0.00	(-0.00 – 0.01)	-0.05	(-0.08 – -0.02)
/pataka/ (syl/s)	-0.00	(-0.01 – 0.01)	-0.04	(-0.08 – 0.01)
FFT (semitones)				
men	0.03	(-0.05 – 0.12)	-0.45	(-0.80 – -0.12)
women	0.24	(0.14 – 0.35)	-0.43	(-0.88 – 0.01)
MPV (dB)				
men	-0.00	(-0.02 – 0.02)	-0.11	(-0.20 – -0.02)
women	-0.05	(-0.08 – -0.01)	-0.19	(-0.34 – -0.05)

Abbreviations used: CI, confidence interval; syl, syllable; s, second.

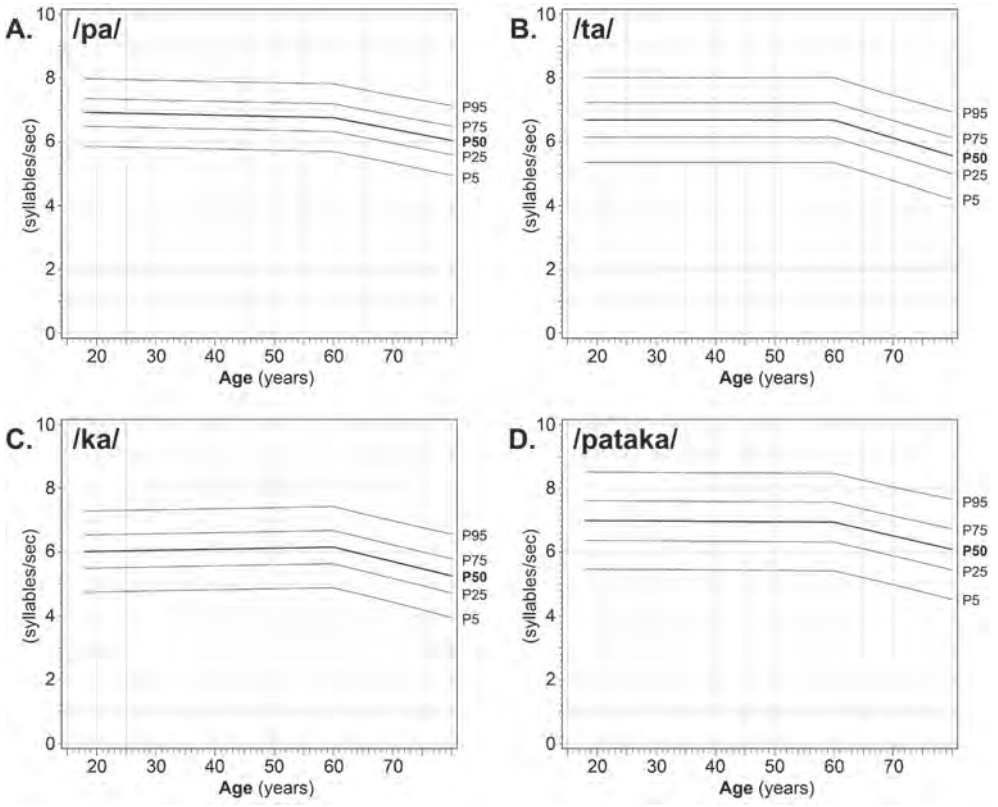

Figure 6.1. The percentile reference lines of the individual sequences of MRR against age, using a piece-wise linear regression with a cut-off point at 60 years of age.

in syllables per second per year is presented for each sequence, using a piece-wise linear regression model with a cut-off point at 60 years. After the age of 60 years, there was a significant decrease in the speed of performance (/pa/: r = -0.03 [95% CI -0.06 – -0.01]; /ta/: r = -0.06 [95% CI -0.09 – -0.02]; /ka/: r = -0.05 [95% CI -0.08 – -0.02]), whereas the age range between 18 and 60 years did not show a significant decline. The percentile reference lines are shown in Figure 6.1. Note that findings were nearly identical for men and women.

Maximum phonation time

In Table 6.2, the median and range of the MPT are shown by age group. Across all age groups, MPT was significantly related to body height ($p < 0.01$). In Table 6.4, the median and range are shown by category of body height. The percentile reference lines are shown in Figure 6.2. The mean difference between men and women was 4.9 s ($p < 0.01$), but the effect of body height was stronger.

Fundamental frequency range

The median and range of the FFR are shown by age group in Table 6.2. FFR was significantly related to age and sex. In Table 6.3, the estimated mean change per year is presented for both sexes using a piece-wise linear regression model with a cut-off point at 60 years. In men, there was a significant

Table 6.4. MPT by category of body height.

body height	n	MPT (s)	
		median	(range)
<159 cm	8	18.2	(11.2 – 24.0)
160-169 cm	53	17.1	(6.6 – 32.9)
170-179 cm	81	18.9	(7.7 – 47.7)
180-189 cm	65	26.8	(10.1 – 55.5)
>190 cm	17	21.1	(11.1 – 54.0)

decrease in FFR per year after the age of 60 (r = -0.45 [95% CI -0.80 – -0.12]), whereas in women this decrease was similar but did not reach significance (r = -0.43 [95% CI -0.88 – 0.01]). Yet for women, there was a significant increase in FFR per year in the age span of 18 to 60 years (r = 0.24 [95% CI 0.14 – 0.35]) (Table 6.3). The percentile reference lines are shown in Figure 6.3A and B.

Maximum phonation volume

In Table 6.2, the median and range of the MPV are shown by age group. MPV was significantly related to age and sex. In Table 6.3, the estimated mean decrease in MPV per year is presented using a piece-wise linear regression model with a cut-off point at 60 years. In men, there was a significant decrease above 60 years of age (r = -0.11 [95% CI -0.20 – -0.02]), which was also found in women (r = -0.19 [95% CI -0.34 – -0.05]). However, only for women, there was a significant decrease in MPV below the age of 60 years (r = -0.05 [95% CI -0.08 – -0.01]). MPV was influenced by body height as well, but the effect of age was stronger. Percentile reference lines are shown in Figure 6.3C and D.

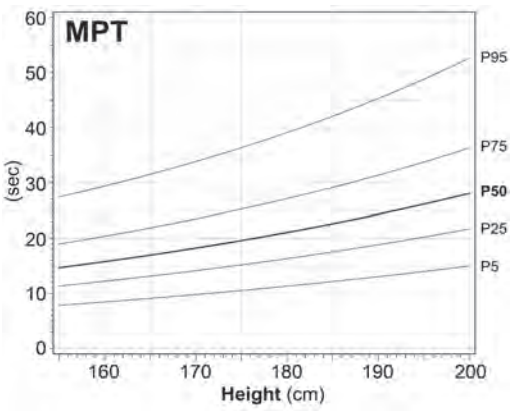

Figure 6.2. The percentile reference lines of the individual sequences of MRR against age, using a piece-wise linear regression with a cut-off point at 60 years of age.

DISCUSSION

This study presents reference values of four maximum performance tests of speech production from a sizeable healthy Dutch population. Overall, the data showed a fairly stable performance up to 60 years, but an age-related decline above the age of 60 years for MRR, FFR, and MPV, leaving the MPT relatively unaffected. Only in women fundamental frequency range showed a marked increase from 18 to 60 years. Smoking habit and profession (vocal use) had no influence on any of the performances.

Looking at MRR, the age effect we found for the monosyllabic sequences is consistent with other studies that found an age-related effect in people older than 65 years.[108,118,119] In contrast, Pierce *et al.* recently assessed healthy subjects older than 65 years and found no significant age effect between 65 and 86 years,[111] although the raw scores of the 75+ group were lower than of the 65+ group for all but one sequence. We found no age effect for the trisyllabic sequence, although the median speed of performance of the 70+ age group was by far the slowest. The absence of an age effect for the trisyllabic sequence under 60 years

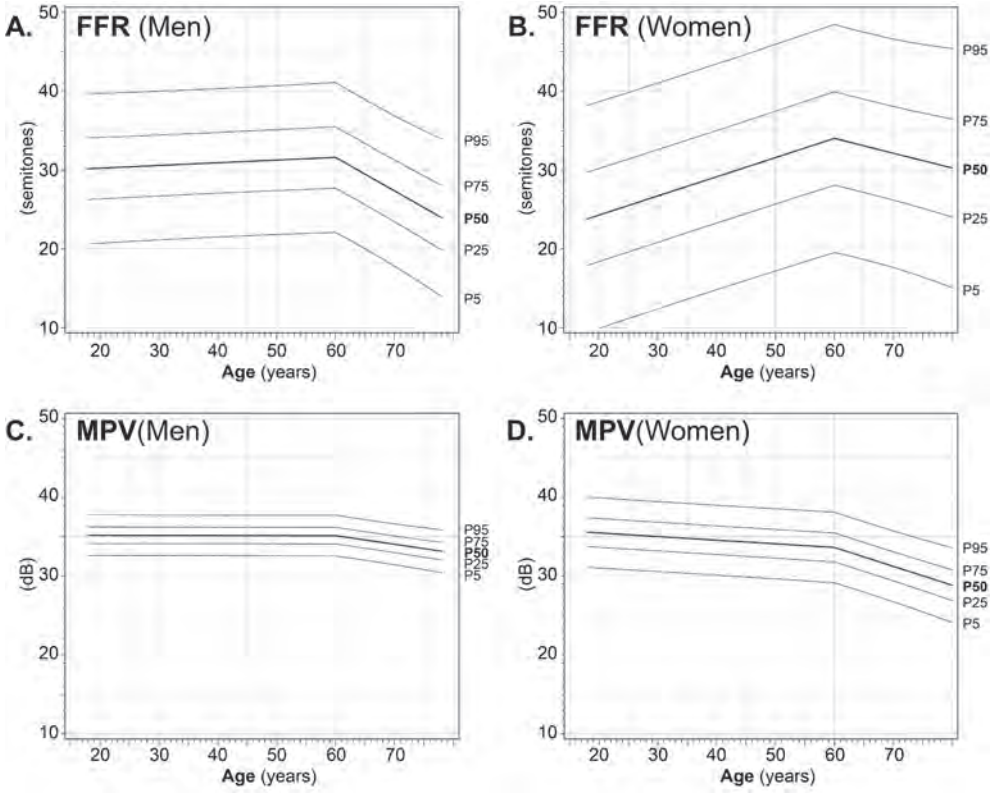

Figure 6.3. The percentile reference lines of FFR and MPV against age by sex, using a piece-wise linear regression model with a cut-off point at 60 years of age.

of age is in line with the study by Icht and Ben-David.[106] In all age groups, /ka/ was the slowest sequence, which is consistent with previous findings.[33,118] Pronouncing /ka/ requires moving the tongue dorsum, which requires movement of most of the mass of the tongue. Men and women performed equally for all MRR sequences, which is in line with the literature. Indeed, studies regarding speech production, speaking and articulation rates hardly ever revealed sex differences.[33,120,121]

Previously published norm data on the FFR are scarce. Only data on maximum pitch are available, as maximum pitch is one of the four parameters of the Dysphonia Severity Index.[122] In accordance with our study, the maximum pitch lowers in ageing men and women, although the causes of laryngeal changes are different between the sexes.[123-125] In ageing men, bowing and vocal fold atrophy are most often described, whereas in ageing women vocal fold edema is most frequent. Besides, the fundamental frequency (F_0) decreases in post-menopausal women and increases in elderly men.[126-128] To sum up, in men, the F_0 rises and the maximum pitch lowers, whereas in women both the F_0 and the maximum pitch lower, which may explain that the full range (FFR) decreases more in men than in women. MPT was not related to age, but only

to body height, most likely because of the relationship between body height and lung function.[112] Indeed, Awan reported a significant correlation between MPT and vital capacity.[129] The fact that we found higher rates for men compared to women is evident because of the interaction between sex and body height, and in line with the literature, revealing a longer MPT for men.[122-124] Unlike MPT, MPV was more dependent on age than on body height, although MPV depends on lung capacity as well, which is related to body height.[112] MPT may be more dependent on lung volume, whereas MPV may be more dependent on muscle strength. With ageing, a decreased muscle strength in combination with the above mentioned laryngeal changes may account for the larger influence on MPT of age than body height.

In three of the four tasks, age was the most important factor influencing maximum performance tests of speech production. Most of this effect was observed from the age of 60 years and older. Human functioning generally declines above the age of 60 years due to neurological, metabolic, and hormonal changes.[130] These changes can have a negative influence on speech, just as on the physical performance of a person.[131] Looking at speech, 'presbyphonia' is the term typically used for age-related vocal changes.[132] Yet, our study clearly shows that age-related changes are not confined to the voice, but reach out to the articulation domain as well, which could be termed 'presbyarthria'. Indeed, in 1974, Ryan and Burk suggested that speech of aged adults may fall at the mild end of a dysarthric continuum.[133] This conclusion was confirmed by Parnell and Amerman in a perceptual study, in which a mild dysarthric speaker was difficult to distinguish from healthy geriatric participants.[134] Other studies that confirm the age-related effects regarding articulation are those showing that speaking rate slows down with advanced age[131,135,136] and studies showing that the variability of acoustic and kinematic measures increases with older age.[44,137]

Hence, the question is justified which underlying mechanism is responsible for this decline of speech quality above the age of 60 years? As healthy persons typically use a small amount of their maximum tongue strength during speech,[138] normal age-related loss of orofacial muscle strength[139,140] can probably not account for loss of speech quality at older age. Recently, research has been conducted regarding non-muscular tissue stiffness. It was found that fibrosis (accumulations of excessive connective tissue), lipomatosis (accumulations of fatty cells), and amyloidosis (deposits of waxy proteins and polysaccharides) in tongue tissue increase progressively with age.[141,142] In line with these findings, Dietsch *et al.* found increased non-muscular tissue stiffness of the skin overlying the masseter, the cheek and lateral tongue with age.[143] Another example is the study by Mefferd and Corder,[144] who found that older adults (>65 years) were able to increase lower lip and jaw speed during an MRR test with /fa/, but that they had more difficulties with stiffness regulation and force production than younger adults (22–55 years). Consequently, it is plausible that non-muscular stiffness of oral structures is a relevant cause for the decline in speech quality above the age of 60–65 years. In addition, the above mentioned insidious neurological changes may have a negative effect on speech as well. Indeed, if the central and peripheral nervous systems gradually decline, there will be slowing of movements, loss of coordination, and an increase in speech variability,[145] although people can adapt to these changes by using compensatory strat-

egies (e.g., slowing down their speaking rate to ensure movement accuracy).[146]

Typically, for all maximum performance tests in this study, the range of non-pathological performance was large. Although a large range of normality has been found for other maximum performance tests such as maximum inspiratory pressure[147] or the 6-min walk test,[148] a large normal range may complicate the interpretation of the performance of individual dysarthric speakers. Yet, the reference lines provide the patient's performance with a percentile score. Nevertheless, qualitative characteristics of maximum performance tasks are equally important to identify underlying pathology (weakness, rigidity, coordination deficits) and, thus, to contribute to the assessment of the type and severity of dysarthria.

Strengths and limitations

Our participants formed a fair representation of the general Dutch population,[149] as we included various age groups between 18 and 80 years with a mean body height of 175.5 cm (SD = 9.6, range 155–201) and a variation in professional voice use. However, we assessed only participants who had Dutch as their first language. It is, therefore, questionable whether our reference values are also applicable to people with other first languages or to people with Dutch as a second language. Icht and Ben-David suggested that their across-language differences in the trisyllabic MRR sequence could be explained by different tongue settings, influencing the /t/ and /k/.[106] In addition, the English /p/, /t/ and /k/ are aspirated, whereas these syllables in Dutch are not. Therefore, it seems valuable to extend our population with participants speaking Dutch as a second language and to compare our data with equally sized groups with other first languages using the same assessment protocol.

Generalizability is related to age range as well. We included participants from 18 years old, because normal values of children up to 17 years are being collected in preparation of the pediatric RDA. Because we took 80 years as the upper age limit, the normal values are not applicable to dysarthric patients older than 80 years.

Another limitation is that we used several examiners to collect the data. Although they were all trained by the first author (S.K.), we cannot rule out subtle differences in examination approach due to inter-observer variability. We did not control for test-retest variability either, but all participants performed each task three times and we used their best performance for analysis.

CONCLUSION

This study provides reference values of four maximum performance tests of speech production to compare the performance of dysarthric patients with non-pathological speech performance. Age was identified as the most important factor influencing MRR, FFR, and MPV (> 60 years), whereas MPT was primarily influenced by body height. Only women showed effects of age on FFR (increase) and MPV (decrease) < 60 years. Interestingly, age-related changes were not confined to the voice, but reached out to the articulation domain as well, which could be referred to as 'presbyarthria'.

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ACH Geurts

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Submitted

7 Dysarthria assessment: a first step to reach better levels of accuracy by video training

ABSTRACT

Background

Diagnosing dysarthrias is notoriously difficult and requires extensive training. For this purpose, the Radboud Dysarthria Assessment was provided with 43 online training videos of all dysarthria types and severity levels.

Aims

This pilot study was conducted to evaluate whether the use of training videos improves the correct identification of dysarthria type and severity by both experienced speech-language therapists (SLTs) and SLT students.

Methods and procedures

Twenty-six SLT students received a one-day training including 10 videos and 19 SLTs were given access to 20 online videos for self-study. Before and after the training, both groups identified the type and severity of dysarthria in 10 new test videos. Their judgments were compared to a reference diagnosis, which was based on consensus by experts.

Outcomes and results

On average, the SLT students improved their agreement with the reference diagnosis on the type of dysarthria from 29.2% to 37.2%, and from 47.9% to 52.5% regarding the severity of dysarthria. Experienced SLTs improved their agreement regarding the type of dysarthria from 42.8% to 46.9%, and from 53.2% to 54.7% regarding the severity of dysarthria.

Conclusions and implications

Our results show that improvement in the correct identification of dysarthria type and severity is possible by means of a short training program with either 10 guided training (SLT students) or 20 self-study (experienced SLTs) videos, but that both training programs did not yet lead to acceptable levels of agreement with a reference diagnosis. To obtain better levels of diagnostic accuracy, more intensive training programs are needed.

INTRODUCTION

In 2014, we developed the Radboud Dysarthria Assessment (RDA), a diagnostic tool for Dutch and Flemish speech-language therapists (SLTs) to assess both the type and severity of dysarthria.[107] In the RDA, the speech characteristics are assessed using spontaneous speech, a reading text, and four maximum performance tasks of speech production (maximum repetition rate, maximum phonation duration, maximum phonation volume and fundamental frequency range). The development of the RDA and the construct validity and reliability of the severity scale have been described in a previous article.[150] In this study, we evaluate the correct assessment of the type of dysarthria, which has been proven to be very difficult, even for experienced SLTs.[151,152] Thus, in an attempt to stimulate SLTs to improve their diagnostic skills, the RDA has been provided with 43 training videos of all dysarthria types and severity levels to be used for self-study or education. We applied the Mayo Clinic classification system[9] to categorize dysarthria as either 'flaccid', 'spastic', 'ataxic', 'hypokinetic', 'hyperkinetic' or 'mixed'. This classification is commonly used by Dutch SLTs as part of their initial SLT education. The purpose of this pilot study was to evaluate our aim that adding training videos to the RDA would improve the correct identification of dysarthria type and severity by SLT students as well as by experienced SLTs.

METHODS

Participants

Twenty-six SLT students who participated in a minor 'Neurorehabilitation' at the HAN University of Applied Sciences agreed to participate. All participants had received basic education about dysarthria as part of their bachelor SLT education.

To recruit experienced SLTs, requests were sent by e-mail to 46 SLTs who were either part of a network of SLTs working in hospitals across the Netherlands or of a network of SLTs working with neurological patients in the region of Nijmegen; 22 of them (48%) responded positively. Because 3 SLTs were asked to score the selected videos twice to determine the intra-rater reliability of the severity scale,[150] 19 SLTs were included in this training study. Twelve SLTs worked in a general hospital, three in a university hospital, two in a nursing home, and one in a private practice. One SLT worked both in a hospital and in a nursing home. On average, the 19 SLTs had worked 12 years with dysarthric patients (range 4–26 years).

Videos

For the training videos, we videotaped 25 men (mean age 55, range 26–77 years) and 18 women (mean age 58, range 14–79 years) while they were assessed with the RDA. These 43 patients suffered from a variety of acute neurological, neuromuscular or neurodegenerative disorders. All video recordings were made with a high definition camera and a separate microphone. The type and severity of the dysarthria of each patient was discussed in an expert group of seven SLTs who developed the RDA, until consensus was reached.[150] The outcome of this consensus process is referred to as the 'reference diagnosis'. The 43 available videos showed all dysarthria types: 'flaccid' (n = 12), 'spastic' (n = 2), 'ataxic' (n = 8), 'hypokinetic' (n = 5), 'hyperkinetic' (n = 5), and 'mixed' (n = 11). We constructed two sets of 10 test videos, whereas 27 training videos were created by adding detailed patient information and diagnostic descriptions. We matched the test videos for type and severity of dysarthria with the

training videos (i.e., if the test video included an ataxic dysarthria, severity level 3, at least one of the training videos included a similar type and severity of dysarthria). In addition, one non-dysarthric person was videotaped, which we included in the set of 10 test videos for the experienced SLTs in order to be able to identify false positive judgments. The reason for this was that the experiment with the SLT students made us realize that before deciding on the type of dysarthria, judging whether speech is normal or pathological can already be difficult. All patients signed informed consent. Approval was obtained from the Medical Ethics Committee of the Radboud University Medical Center.

Procedure

The 26 SLT students received a one-day training by a lecturer specialized in motor speech disorders. Their training was focused on the correct assessment of the type and severity of dysarthria in 10 selected (based on the matching) training videos. The number of training videos was restricted to 10, because the training was limited to one day. One week before and ten days after the training, the students were asked to score the type and severity of dysarthria for the same set of 10 test videos. The 19 experienced SLTs were given access to 20 online training videos via a secured online web service. They also got access to the part of the test manual containing detailed diagnostic descriptions of all dysarthria types. These SLTs had 3 weeks to complete the online training, during which they noted the number of training hours. Shortly before and after the training they scored the same set of 10 online test videos, including the video of the non-dysarthric person, but they were kept unaware of the neurological diagnosis of the patients shown on the videos. All raters scored the dysarthria type and severity using a Microsoft Office Access form. All ratings were compared with the reference diagnosis.

Statistical analysis

The number of test videos (10) in relation to the number of dysarthria types (6) did not allow calculation of representative Kappa values, so instead we calculated the percentage of absolute agreement with the reference diagnosis per dysarthria type and severity before and after the training. The overall percentage of agreement was calculated per group for all 10 test videos. The following rules were applied to handle the 'mixed' dysarthrias. First, if a single dysarthria was incorrectly identified as 'mixed', the individual dysarthria type that was correctly identified was counted for *1/(number of dysarthria types identified)*. Second, if a 'mixed' dysarthria was scored as a single dysarthria type, this added to the agreement on the 'mixed' type for 50% if the single dysarthria type was part of the mixed reference diagnosis. Third, if a 'mixed' dysarthria type was scored partially correct, the individual dysarthria types that were correctly identified were counted, each for *1/(number of dysarthria types identified)*. Therefore, the column of the mixed dysarthria in Tables 7.1 and 7.3 does not always sum up to 100%.

Chi-square tests were used to assess whether the years of dysarthria experience and the number of training hours affected the level of agreement with the reference diagnosis (IBM SPSS Statistics 22 for Windows; IBM Corp., Armonk, NY).

RESULTS

SLT students

Before the training, the type of dysarthria was correctly identified by 29.2% of the 26 students (Table 7.1). After the training,

this number improved to 37.2% (27% relative improvement), while the error pattern stayed almost the same. Both 'flaccid' and 'hypokinetic' dysarthrias were correctly identified by the majority of the students, albeit with a higher percentage after the training. 'Spastic' and 'ataxic' dysarthrias were most frequently misclassified as 'flaccid' dysarthria, and 'hyperkinetic' as 'spastic' dysarthria. None of the students was able to correctly identify 'mixed' dysarthria, neither before nor after the training. In 32% of the cases before and in 21.2% after the training, 'mixed' dysarthria was scored

Table 7.1. Percentage absolute agreement of SLT students with the reference diagnosis (type of dysarthria) before and after the training.

	Reference diagnosis (type of dysarthria)											
	Flaccid (*n* = 3)		Spastic (*n* = 1)		Ataxic (*n* = 2)		Hypokinetic (*n* = 2)		Hyperkinetic (*n* = 1)		Mixed* (*n* = 1)	
	before	after	before	after	before	after	before	after	before	after	before	after
Flaccid	**64.6**	**71.0**	**34.0**	**38.5**	**58.3**	**57.7**	22.8	22.1	4.2	-	8.0	**28.8**
Spastic	1.3	6.3	19.6	15.4	2.0	-	11.7	10.6	**57.6**	**40.4**	**	**
Ataxic	12.6	15.7	25.3	36.5	12.2	30.8	11.3	3.8	26.4	36.5	**	**
Hypo	16.9	0.6	12.3	-	21.6	11.6	**35.0**	**65.4**	-	3.8	2.0	23.1
Hyper	4.7	6.1	8.7	9.6	6.0	-	19.3	-	11.8	19.2	**32.0**	3.8
Mixed	N. A.	N. A.	N. A.	N. A.	N. A.	N. A.	N. A.	N. A.	N. A.	N. A.	**32.0**	21.2

Bold: highest agreement. * mixed spastic-ataxic. This column does not amount to 100% (see section on statistical analysis). ** included in the mixed dysarthria for 50% per type. Abbreviations used: N. A., not applicable (they are included in the individual types for 50%).

Table 7.2. Percentage absolute agreement of SLT students with the reference diagnosis (severity of dysarthria) before and after the training.

	Reference diagnosis (severity of dysarthria*)											
	0		1		2 (*n* = 3)		3 (*n* = 4)		4 (*n* = 2)		5	
	before	after	before	after	before	after	before	after	before	after	before	after
0					2.5	0		3.1				
1					44.1	41.0	12.9	4.0				
2					**44.3**	**42.2**	29.6	38.5	1.9	3.9		
3					9.0	15.4	**40.6**	**44.1**	7.7	9.6		
4						1.3	16.0	8.7	**58.7**	**71.2**		
5							0.8	1.6	31.7	15.4		

Bold: highest agreement. * Severity rating: 0, no dysarthria; 1, minimal dysarthria; 2, mild dysarthria; 3, mild-moderate dysarthria; 4, severe dysarthria; 5, very severe dysarthria/anarthria.

partially correctly: i.e. only one of the dysarthria types contributing to 'mixed' dysarthria was scored correctly.

The severity of dysarthria was correctly identified by 47.9% of the students before the training (Table 7.2). After the training, this number improved to 52.5% (10% relative improvement).

Experienced SLTs

Before the training, the SLTs agreed with the reference diagnosis in 42.8% of the cases (Table 7.3). After the training, they

Table 7.3. Percentage absolute agreement of experienced SLTs with the reference diagnosis (type of dysarthria) before and after the training.

	Reference diagnosis (type of dysarthria)											
	Flaccid (*n* = 2)		Spastic (*n* = 1)		Ataxic (*n* = 2)		Hypokinetic (*n* = 1)		Hyperkinetic (*n* = 2)		Mixed* (*n* = 1)	
	before	after	before	after	before	after	before	after	before	after	before	after
Flaccid	**52.8**	**54.0**	10.5	10.5	30.3	36.8	28.9	26.3	13.9	18.4	**	**
Spastic	16.7	14.5	**55.3**	**34.2**	2.7	-	15.8	18.4	13.5	9.3	23.7	25.4
Ataxic	4.2	14.5	18.4	**34.2**	**36.9**	**44.7**	2.6	15.8	23.1	13.2	**	**
Hypo	19.5	17.1	2.6	-	22.4	13.2	**42.1**	**39.5**	8.4	9.2	9.6	5.3
Hyper	7.0	-	13.2	21.1	7.9	5.3	10.5	-	**41.3**	**50**	25.4	10.5
Mixed	N.A.	N.A.	N.A.	N.A.	N.A.	N.A.	N.A.	N.A.	N.A.	N.A.	**28.1**	**58.7**

Bold: highest agreement. * mixed flaccid-ataxic. This column does not amount to 100% (see section on statistical analysis). **included in the mixed diagnosis for 50%. N. A.: not applicable (they are included in the individual types for 50%).

Table 7.4. Percentage absolute agreement of experienced SLTs with the reference diagnosis (severity of dysarthria) before and after the training.

	Reference diagnosis (severity of dysarthria*)											
	0 (*n* = 1)		1		2 (*n* = 4)		3 (*n* = 3)		4 (*n* = 2)		5	
	before	after	before	after	before	after	before	after	before	after	before	after
0	**52.6**	36.8			2.6		1.8					
1	42.1	**57.9**			35.5	10.5	1.8					
2	5.3	5.3			**38.2**	**51.3**	22.8	8.8				
3					23.7	38.2	**50.9**	**59.7**	21.1	18.4		
4							22.8	28.2	**71.1**	**71.1**		
5									7.9	10.6		

Bold: highest agreement. * Severity rating: 0, no dysarthria; 1, minimal dysarthria; 2, mild dysarthria; 3, mild-moderate dysarthria; 4, severe dysarthria; 5, very severe dysarthria/anarthria.

agreed in 46.9% of all cases (10% relative improvement). Before the training, only one SLT was able to correctly identify 'mixed' dysarthria versus two SLTs after the training. Correct identification of one of the dysarthria types contributing to 'mixed' dysarthria was improved after the training from 28.1% to 58.7%. The highest agreement per dysarthria type was consistently in line with the reference diagnosis.

The non-dysarthric speaker was judged as being dysarthric nine times before the training (47.4% false positives) versus 12 times after the training (63.2% false positives). None of the dysarthric patients were judged as non-dysarthric (0% false negatives).

The severity of dysarthria was correctly identified by 53.2% of the SLTs before the training (Table 7.4). After the training, this number improved to 54.7% (3% relative improvement).

The SLTs used on average 5.5 hours (range 1.5–19 hours) for the online training. Eleven SLTs (57.9%) trained less than 5 hours, five SLTs (26.3%) 5 to 10 hours, and three SLTs (15.8%) 10 hours or more. Chi-square tests revealed no significant correlations between the level of agreement with the reference diagnosis and the number of training hours ($p = .541$), nor between level of agreement and years of experience ($p = .678$).

DISCUSSION

This pilot study showed that correct classification of dysarthria type and severity can be improved by a short training using 10 or 20 videos, but also that percentages of correct dysarthria type identification remained below 50%. Percentages of correct judgment of dysarthria severity were slightly higher, but did not exceed 55%. For SLT students, the initial percentages and the relative improvement can be considered acceptable because of their inexperience and the fact that they are still in a learning process. For the SLTs experienced with dysarthric patients, however, the overall agreement before and after training was unsatisfactory.

In both study groups, the training results showed only small improvements, which was much less than we expected. In fact, if we had not allowed the mixed dysarthrias to be partially correct, the overall agreement would have been even lower. We know from previous studies that absolute agreement rates regarding the type of dysarthria do not exceed 40% when the medical diagnosis is unknown, even in experienced SLTs, although this may differ for different types of dysarthria.[151-153] In these studies, however, the speech tasks presented were limited. We anticipated that providing all speech tasks of the RDA might result in higher levels of agreement, as critical speech characteristics leading to a particular dysarthria type may be more prominent in specific speech tasks. In addition, in both study groups, we explicitly highlighted the specific speech characteristics per dysarthria type in the different speech tasks. For example, mild coordination problems in ataxic dysarthria may not be identified in spontaneous speech if a patient slows down his speaking rate. But when performing maximum repetition rate, the sequence will likely be dysrhythmic or the articulation more slurred. However, because we did not explicitly compare the difference between scoring dysarthria type based on spontaneous speech versus using all speech tasks, we cannot draw any conclusions from this study about the added effect of providing all speech tasks.

In both study groups, agreement on the severity of dysarthria was higher than on the type of dysarthria. This is not surprising given the fact that the inter-rater reliability

of the severity scale showed good agreement (ICC 0.85) previously.[150] Remarkably, experienced SLTs showed a relatively high number of false positives judgments of the non-dysarthric patient, which may be explained by two factors. First, the non-dysarthric person spoke very articulated, which might have caused suspicion of this person using a compensation strategy for some type of dysarthria. Second, the SLTs were focused on classifying dysarthria and were kept unaware that a non-dysarthric speech sample was included.

Differences between the performance of SLT students and SLTs.

The observation that SLT students showed a lower level of agreement than experienced SLTs before and after the training is understandable, because the students lacked clinical experience. In fact, the majority of the SLT students had never seen a patient with dysarthria in real life yet. In contrast, we can assume that the experienced SLTs had developed internal perceptual representations of several types of dysarthria by assessing and treating these patients over the years. The lack of experience of SLT students may also explain why their relative improvement after training was larger than for the experienced SLTs. The fact that the experienced SLTs improved on average only 10% by watching the training videos may be explained by two factors. First, more than half of the SLTs trained less than 5 hours. As each video takes approximately 15 minutes to watch, the majority did not study all 20 videos in detail. Second, established internal perceptive representations may be difficult to change by only watching videos without interaction or feedback.

Remarkably, the SLT students were able to identify flaccid and hypokinetic dysarthria types better than spastic, ataxic and hyperkinetic types. The latter three probably represent more variable speech deficits, which are difficult to interpret for inexperienced SLT students. In accordance with the study of Fonville *et al.*,[152] our study also showed that mixed dysarthria was the most difficult type to identify.

Factors that may influence dysarthria type classification

Table 7.3 shows that the highest level of agreement per dysarthria type in experienced SLTs was always in line with the reference diagnosis, which supports the fact that the reference diagnoses were set correctly by the experts. Still, the percentage of agreement was around 50% or lower suggesting that, in general, SLTs wrongly classify dysarthric patients in half of the cases. There are several explanations for this result. One important reason is the misinterpretation of compensatory speech behavior. During the expert meetings in which the reference diagnoses were set, we learned that the use of compensatory speech strategies by dysarthric patients was the most problematic factor for reaching agreement. For example, some patients with a weak vocal quality compensated with hypertonic vocal use. The hypertonic vocal use was not consistently present during all speech tasks but, on occasion, it was erroneously classified as a spastic speech characteristic, which is incorrect because a dysarthric characteristic is supposed to be consistently present.[6] In line with this explanation, some speech characteristics may have been overrated, because the SLT students and experienced SLTs tried very hard to identify every deviant characteristic. Another explanation for the difficult identification of dysarthria type may be that we provided our participants only with the patients' speech performances. In clinical practice, however, SLTs usually perform an oral motor examination for a better interpretation

of the speech characteristics. In addition, the medical history of the patient is often known, which facilitates the recognition of typical speech characteristics.[153] Finally, we assessed and videotaped dysarthric patients without selecting 'typical' patients as they are reported in textbooks, which supports the clinical validity of our test and training videos, but also revealed that many different patients samples and many hours of training are needed to correctly diagnose motor speech disorders.

Finally, the relevance of a correct or incorrect dysarthria diagnosis may be questioned from a clinical perspective. In a medical diagnostic process correct identification is obviously of crucial importance, but what are the consequences of a misdiagnosis of dysarthria for its treatment? In acute neurological disorders such as stroke, SLTs make use of rehabilitation techniques that are different for predominant weakness than for predominant coordination problems.[6] In progressive neurological disorders, speech therapy is primarily aimed at learning compensatory speaking techniques, which are also (partly) dependent of dysarthria type. For example, in hypokinetic dysarthria, the Pitch Limiting Voice Treatment[154] or Lee Silverman Voice Treatment[155] are recommended to overcome the hypokinetic speech, whereas in amyotrophic lateral sclerosis speech therapy is more focused on communicative effectiveness and augmentative communication.[7] Thus, a correct diagnosis of dysarthria type seems to matter when deciding on the type of treatment.

Limitations of the current study

SLT students were trained as part of their initial education, whereas experienced SLTs were expected to be able to use the RDA after self-study of the videos and descriptions. Hence, because of the different experimental set-ups, the comparison between SLT students and experienced SLTs can only be descriptive. The experienced SLTs did not have an interactive training, where the interaction and discussions with the SLT students may have led to a relatively large improvement. We did not monitor the self-study of the experienced SLTs, nor did we ask them if they felt that they had been struggling or if they were surprised about the correct outcomes. As a consequence, we are unaware of which videos they studied, how they studied these videos, and how they valued the diagnostic tests. A final study limitation is that not all dysarthria types were equally available in our video material, which restricted the matching of the test videos with the training videos.

CONCLUSION AND FUTURE PERSPECTIVES

A short training program with 10 or 20 videos improves correct identification of dysarthria type and severity, but agreement rates are still clinical unacceptable. To improve and re-evaluate clinicians' skills to correctly diagnose dysarthria types, we aim to complete our set of video examples with all different dysarthria types and severity levels. Because this study suggests that access to exemplary videos alone is not very effective in itself, the next step will be to design attractive e-learning modules with explicit training tasks and questions or to develop a combined program of e-learning and an interactive course (blended learning). Although reports on the clinical consequences of misdiagnosed dysarthrias for the efficacy of treatment are lacking, we believe that with more effective learning programs agreement rates of 80% or higher may be attainable with a potentially im-

portant impact on the efficacy of dysarthria treatment.

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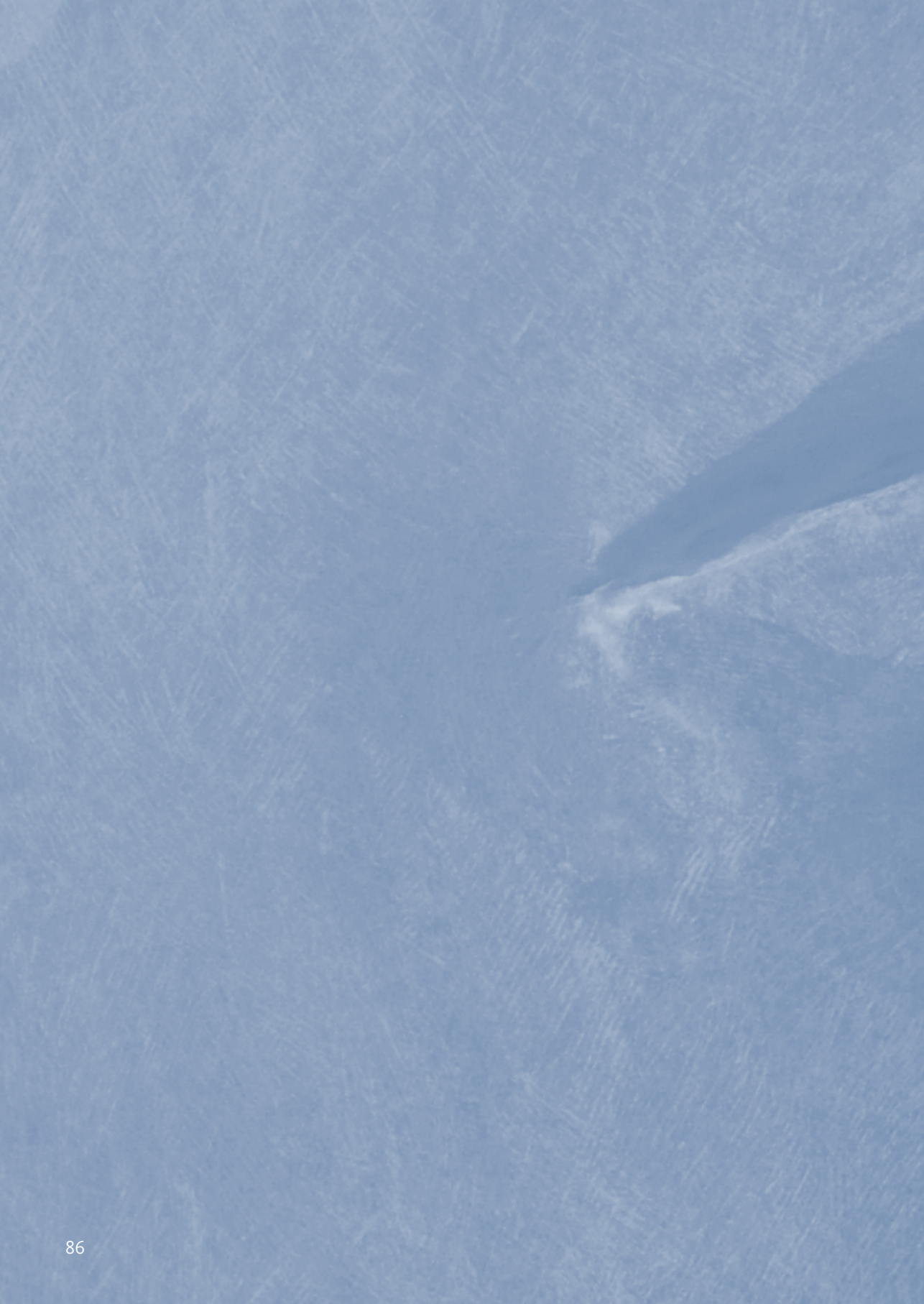


8 Summary and general discussion

SUMMARY

Speech is unique for humans and, despite the rise of social media, daily communication is highly dependent on spoken language. When a neurological disease affects speech, this is called dysarthria. Dysarthria can be caused by (sub-)acute central neurological, central neurodegenerative or peripheral neuromuscular diseases (NMD). The first part of this thesis focuses on the prevalence rates and the evidence for treatment of dysarthria and dysphagia in patients with NMD. Dysphagia is added, because dysarthria often co-occurs with dysphagia, as both disorders are caused by oromotor problems within overlapping muscle groups. The second part of this thesis focuses on the development of the Radboud Dysarthria Assessment (RDA).

This thesis aims to 1) examine prevalence rates of dysarthria and dysphagia in NMD, 2) summarize the evidence for treatment of dysarthria and dysphagia in NMD, and 3) develop and evaluate a dysarthria assessment at the level of speech function.

In ***chapter 1***, a general introduction to dysarthria and dysphagia is provided by presenting an overview of neurological diseases that can cause dysarthria and/or dysphagia. In addition, a theoretical approach to the different types of dysarthria is presented. Finally, aims and outline of this thesis are described.

Part I

In part one of this thesis, prevalence rates of dysarthria and dysphagia in NMD are presented, and evidence for treatment of dysarthria and dysphagia in NMD is identified (aims 1 and 2).

In ***chapter 2***, high prevalence rates of dysarthria and dysphagia in a large group (n = 220) of patients with various types of NMD are demonstrated. Pooled prevalence rates were estimated per patient group by weighting each disease category according to sample size, as the patients were not distributed equally across the disease categories within the groups. The pooled prevalence of dysarthria was 46% in an unselected cohort of outpatients and 62% in a selected cohort. The pooled prevalence of dysphagia was 36% in an unselected cohort and 58% in a selected cohort. There was a modest but significant association between the presence of dysarthria and dysphagia (r_s = 0.40; p <0.01). Although dysphagia was generally mild, dysarthria was moderate to severe in 15% of the dysarthric patients. Interestingly, dysarthria and dysphagia were found among patients with NMD that are not typically associated with these disorders. The most remarkable results were the high prevalence rates of both dysarthria and dysphagia in patients with metabolic and other myopathies. The fact that dysarthria and dysphagia were both highly prevalent in (most of the time) slowly progressive diseases emphasizes the need that physicians should evaluate both dysarthria and dysphagia independent of the type of NMD. Even though not every patient with a progressive disease will benefit from treatment directed at improving their speech and/or swallowing efficacy, these patients may still profit from learning individually tailored functional compensations delivered by an experienced speech-language pathologist.

Chapter 3 describes the presence of dysphagia in Myotonic Dystrophy type 2 (DM2). Dysphagia is a predominant and potentially life-threatening feature of Myotonic Dystrophy type 1, but had not been examined in DM2. DM2 patients who reported swallowing difficulties based on the Gastrointestinal Symptoms Questionnaire were recruited for this study (n = 10). They were clinically investigated by an

experienced speech therapist in neuromuscular disorders. Eight DM2 patients gave additional informed consent to fiberoptic endoscopic evaluation of swallowing (FEES). Dysphagia could be confirmed in all patients (clinically in 100%, with FEES in 88%), and was more pronounced in older DM2 patients. The severity of dysphagia was generally mild and none of the patients had suffered aspiration pneumonia in the past or had significant weight loss.

Chapter 4 identifies the evidence for the effects of interventions by speech-language pathologists in adult patients with NMD. In a systematic review, we summarized and evaluated the literature on the effectiveness of interventions for dysarthria and dysphagia. Inclusion was restricted to peer-reviewed articles published in the English, German, French or Dutch language. Inclusion criteria were 1) information and advice regarding dietary modification, augmentative and alternative communication and instruction of the patient and relatives, and 2) compensatory strategies regarding teaching of swallowing maneuvers and/or strategies to improve intelligibility. Single case studies were excluded. The search strategy identified 1.772 articles, but only 4 fulfilled all inclusion criteria. Those 4 studies were all ranked as 'other designs'. The methodological quality of the studies was found sufficient for only one uncontrolled pre-post study. This study assessed the influence of ptosis on swallowing in patients with oculopharyngeal muscle dystrophy (OPMD).[46] When the head position was slightly flexed and, thus, not adapted to the ptosis, there were indications that swallowing efficiency improved in patients with OPMD (level III evidence). The main conclusion from this review was that the published evidence for the effectiveness of interventions by speech-language pathologists in adults with NMD is still very limited.

Part II

In the second part of this thesis, the development of the Radboud Dysarthria Assessment (RDA) is described, including the provision of reference values for maximum performance tests of speech production (aim 3).

Chapter 5 summarizes the development and validation of the RDA. A first version of the RDA (freely available and unvalidated) was made available online in 2007, but we felt the need to improve and validate the instrument. First, national consensus was sought for the tasks and the qualitative recording form. Second, we added a self-evaluation questionnaire. Third, we clinimetrically evaluated the recording form, severity scale, and the self-evaluation questionnaire. Finally, training videos were added, demonstrating all types and severities of dysarthria including a detailed interpretation of the assessment for self-training purposes. Exploratory factor analysis of the recording form extracted four factors (articulation, resonance, phonation, respiration/prosody) that yielded an explained variance of 70.3%. Each factor showed good internal consistency. The self-evaluation questionnaire showed excellent internal consistency and intra-class correlation coefficients of the severity scale showed good reliability. The severity scores and self-evaluation questionnaire correlated substantially to strongly with the Speech Handicap Index (SHI) and substantially with the Dutch sentence intelligibility assessment (NSVO-Z). Based on these results, we concluded that the RDA is a valid and reliable tool for diagnosing the severity of dysarthria. Further investigation is needed to demonstrate whether this instrument can successfully support speech-language

therapists in correctly diagnosing also the type of dysarthria.

Chapter 6 describes a study assessing multiple maximum performance tests of speech production in a large population (n = 224) of healthy Dutch adults between 18 and 80 years with the aim to collect reference values. All subjects performed the following maximum performance tests: maximum repetition rate (MRR), maximum phonation time (MPT), fundamental frequency range (FFR), and maximum phonation volume (MPV). Initially, univariate analysis was used to explore the association between each maximum performance test and personal characteristics (age, sex, body height, profession, and smoking habit). Then, characteristics with a p-value of < 0.05 were included in a multivariate linear regression to study their unique influences. We used a wide range of models on each maximum performance test of speech production. The Likelihood-Ratio test was used to test the differences between the models for their goodness of fit to the data. MRR, FFR and MPV were statistically significant related to age, whereas MPT was related to body height. Thus, in three of the four tasks, age was the most influential factor, especially from the age of 60 years and older. It was not clear which underlying mechanism was responsible for the observed decline with age, but we argued that — in addition to neurological, metabolic, hormonal and physical changes — non-muscular tissue stiffness might play a role. The range of non-pathological performance was large, which may complicate the interpretation of the performance of individual dysarthric speakers.

In ***chapter 7***, we evaluate whether the use of the training videos of the RDA improves the correct identification of dysarthria type and severity by both experienced speech-language therapists (SLTs) and SLT students. The SLT students received a one-day training focused on the correct assessment of the type and severity of dysarthria in 10 selected training videos. One week before and ten days after the training, the students were asked to score the type and severity of dysarthria for the same set of 10 test videos. The experienced SLTs were given access to 20 online training videos via a secured online web service. They also got access to the part of the test manual containing detailed diagnostic descriptions of the different dysarthria types. The SLTs had 3 weeks to complete the online training, during which they noted the number of training hours. Shortly before and after the training they scored the same set of 10 online test videos. All judgments were compared to a reference diagnosis, which was based on consensus by multiple experts. Regarding the type of dysarthria, SLT students improved from 29.2% to 37.2% and SLTs from 42.8% to 46.9%. Regarding the severity of dysarthria, SLT students improved from 47.9% to 52.5% and SLTs from 53.2% to 54.7%. In SLTs, there were no significant correlations between the level of agreement with the reference diagnosis and the number of training hours, nor between the level of agreement and years of experience. Both training programs did not yet lead to acceptable levels of agreement with the reference diagnosis. To obtain better levels of diagnostic accuracy, it appears that more intensive, or adapted training programs are needed.

GENERAL DISCUSSION

The general discussion of this thesis is divided in three parts. Part one of this general discussion is aimed at the scope of international dysarthria assessments and the rationale behind some of the choices we made during the process of developing the RDA.

In addition, the importance of determining the type and severity of dysarthria are discussed. Subsequently, the importance of treating dysarthria (Part two of this general discussion) and dysphagia (Part three) in NMD will be discussed, as well as our thoughts regarding treatment options and future perspectives.

GENERAL DISCUSSION PART ONE

Dysarthria assessments worldwide

The first attempt to clinically assess dysarthria was made in 1969 by Darley, Aronson and Brown who listed 38 auditory parameters to rate on a 7-point scale: the Mayo Clinic rating system.[9] They concluded that specific combinations of speech characteristics could differentiate six types of dysarthria, i.e. flaccid, spastic, ataxic, hypokinetic, hyperkinetic, and mixed. However, the rating system never became a standardized assessment tool, probably because of its extensive number of parameters and limited inter-rater reliability.[151,156] Despite the fact that the Mayo Clinic rating system was not clinically used, a hand full of other dysarthria assessments have been developed over the last decades.

In 1980, the well-known and later on in German and Dutch translated Frenchay Dysarthria Assessment (FDA) was developed in the UK.[157] The FDA was revised in 2008 (FDA-2).[84] In the UK, the FDA-2 is still the standard assessment tool, although international contacts told us that especially experienced SLTs have developed their own scoring system. The FDA-2 combines observations of oral structures and nonverbal oral functions, assessment of speech characteristics, and measurement of intelligibility in eight sections and is, therefore, aimed at all domains of the International Classification of Functioning (ICF), i.e. function (motor speech and oral motor), activity and participation. After the assessment, the scores lead to a 'profile' which should match with a specific dysarthria type. One could argue that the FDA-2 has too much focus on the observation of oral structures, whereas the assessment of speech characteristics is relatively limited. Besides, intelligibility is tested with 12 randomly chosen words out of 50 words, leading to familiarity with the words by the SLT after only a couple of patients. Familiarity with the words influences the intelligibility scores, making the test less reliable.

In 1982, the Robertson Dysarthria Profile[158] was published in the UK, but it never became equally known as the FDA. The Robertson Dysarthria Profile is largely focused on facial musculature and speech-like tasks to examine the respiration and phonation, while articulatory characteristics are only scarcely assessed and described. The Italian version of the Robertson Dysarthria Profile is the most frequently used dysarthria assessment in Italy.

In 1993, the 'Dysartritest' was developed in Sweden, which was revised in 2015.[159] This dysarthria assessment is the standard dysarthria assessment in Sweden. It includes 14 oromotor tasks, but is also focused on speech functions and intelligibility. The 'Dysartritest' is not aimed at classifying dysarthria types, but only at severity and a description of dysarthric characteristics (e.g. slow speaking rate, monoloudness).

Recently, in 2017, the Bogenhausener Dysarthrieskalen (BoDys) was developed in Germany.[105] The BoDys is the only instrument that focuses purely on speech (conversational speech, sentence repetition, text reading, and picture story) because, based on the authors opinion, the motor system controlled for speech is independent of the motor control system for speech-like (e.g. maximum repetition rate) and non-speech

tasks (i.e. oromotor movements).[90,105,160] Like the 'Dysartritest', the BoDys focuses only on the severity of dysarthria, using a 4-point scale from most severe (0) to no impairment (4) for the following nine scales: respiration, voice level, voice quality, voice stability, articulation, nasal resonance, articulation rate, fluency, and prosodic modulation. The BoDys has a good inter-rater reliability and a high discriminant and convergent validity.[105] Italian SLTs are currently translating the BoDys with the aim to replace the Robertson Dysarthria Profile.

Parallel to the process of developing the BoDys, we developed the RDA to provide Dutch and Flemish SLTs with a standardized and valid dysarthria assessment (***Chapter 5***). In the Netherlands, the RDA is called 'Nederlandstalig Dysartrieonderzoek – volwassenen' (Dutch Dysarthria Assessment – adults). In international publications, the word 'Dutch' was changed into 'Radboud' – the institution where the instrument was developed. The addition 'Dutch' would suggest that the assessment is language-specific, which is not the case, as the assessment can easily be translated into another language. When developing the RDA, we explicitly chose to focus on both the type and severity of dysarthria, similar to the FDA-2. However, in contrast to the FDA-2, we aimed at an instrument to assess only speech function, using two speech (spontaneous speech and reading) and four speech-like tasks (maximum repetition rate, maximum phonation time, maximum phonation volume and fundamental frequency range). A description of these speech-like maximum performance tests can be found in Chapter 5. In the next paragraph, the choices we made for the use of four maximum performance tasks in our clinical dysarthria assessment will be discussed. Thereafter, the focus will be on the importance of both dysarthria severity and type classification.

The importance of using maximum performance tests

There is a lot of discussion about the importance of maximum performance tests in clinical dysarthria assessment. There are authors who call those tasks 'non-speech', because in maximum performance tests an acoustic signal is produced without a communicative goal.[42] Kent defined speech as 'movements or movement plans that produce as their end result acoustic patterns that accord with the phonetic structure of a language'.[109] In for example /pataka/, a phonetic-acoustic pattern is produced, however, the fact that such patterns are repetitive and have no meaning makes them 'speech-like', or 'quasi-speech', a term proposed by Weismer.[161]

There are two theories about motor control of speech and speech-like or non-speech tasks. The first theory adheres to the 'task-dependent model'[42,162] and proposes a specialized, distinct motor control system dedicated to speech production, independent of the control system for non-speech. Recently, the research group that proposed the task-dependent model, which is the same group that developed the BoDys, found dissociations between movement rates obtained from speech, maximum repetition rate, and oral motor tasks in 130 patients with a variety of neurogenic movement disorders, which strengthened their belief in the task-dependent model.[90] Therefore, the BoDys only includes speech tasks.

The second theory adheres to an 'integrative model'[91] and proposes that speech production involves a particular, unique combination of skills and properties, some of which are shared with other motor behaviors, such as those assessed in maximum

performance tests. The RDA is based on the integrative model, although the basis of the RDA is spontaneous speech. There is no discussion about the fact that spontaneous speech is the most representative task of daily communication. However, in order to interpret speech characteristics for the differential diagnostic goal of dysarthria type classification, maximum performance tests may be very helpful. For example, a vocal tremor can be heard better during the maximum phonation duration task than during spontaneous speech and patients who tend to slow down their speaking rate during spontaneous speech can exhibit some difficulties (e.g. slow maximum rate or a dysrhythmic sequence) during maximum repetition rate.

The importance of dysarthria severity classification

All currently used dysarthria assessments include severity classification. Scoring the severity of dysarthria is important for a couple of reasons. First, a severity score is helpful to monitor the dysarthria over time, for instance, to evaluate progression, recovery or treatment effects. Although the severity scale of the RDA showed high reliability scores, we did not assess its sensitivity to change. Second, a severity score is important for treatment focus. Generally, in the case of a minimal dysarthria, patients can be helped satisfactorily with subtle adjustments of their speech habits. In mild to moderate dysarthria, the therapy usually will be more focused on practicing speaking techniques, whereas in severe dysarthria speech functions are more compensated with augmentative and alternative communication strategies (see Part two of this general discussion).

The importance of dysarthria type classification

Categorizing the different types of dysarthria is a clinical challenge, not only because it is difficult, even for experienced SLTs, but also because it remains unsure whether defining the type of dysarthria is essential for diagnostic and/or treatment purposes. These two aspects will be discussed in more detail.

Why is categorizing the type of dysarthria difficult? There are a couple of reasons: 1) speech symptoms within a specific dysarthria type may vary in their severity, 2) there is considerable overlap in speech symptoms among the dysarthria types (e.g., imprecise consonants can be present in every type of dysarthria), 3) not all speakers with the same medical diagnosis exhibit exactly the same speech symptoms, and 4) patients use compensatory strategies for improving their intelligibility, which interferes with the dysarthric characteristics.

If categorizing the type of dysarthria is that difficult, is it essential for diagnostic and/or treatment purposes? Labeling the type of dysarthria by an expert SLT can be helpful for the medical diagnosis, because specific speech characteristics may point towards the site of the lesion or the underlying disease. For example, hypokinesia is a key characteristic of Parkinson's disease (PD). However, when besides the hypokinetic speech e.g. coordination problems are heard, the diagnosis idiopathic PD must be reconsidered, because a mixed dysarthria is more likely a feature of atypical parkinsonism. For this reason, at the Radboud University Medical Center, SLTs are frequently consulted to assist in medical diagnosing.

For treatment purposes, there are also important reasons to know the type of dysarthria, because some treatments are developed for a specific type of dysarthria. This

will be illustrated more in detail in Part two of this discussion. On the other hand, some treatment approaches are applicable to different types of dysarthria, especially compensatory speaking techniques. For example, both in ataxic and flaccid dysarthria, slowing down the speaking rate can be very effective.[48] And although the Lee Silverman Voice Treatment (LSVT) was developed for hypokinetic dysarthria in PD, studies show that this treatment may also be a successful approach for patients with an ataxic, spastic and unilateral upper motor neuron dysarthria.[163-165] Besides, it must be acknowledged that several studies have shown that dysarthria type is not a significant predictor of the efficacy of specific speech therapies.[48,166] So, if the treatment is purely focused on learning an adequate compensatory speaking technique, it could be argued that labeling the type of dysarthria is not crucial.

The alternative for characterizing the type of dysarthria is to describe dysarthria in terms of deviant speech characteristics. This is especially emerging in childhood dysarthria, because children tend to exhibit different speech characteristics than adults, probably due to the fact that 1) dysarthria in adults is generally caused by an acquired disease, whereas in children it is commonly due to an early acquired or inherited disease, 2) in children, the motor control system is still developing, whereas in adults it is fully developed, and 3) the adaptability of a child's brain is larger than the adaptability of the adult brain.[167] Because of these differences between adults and children and the fact that speech tasks may differ between adults and children (e.g. the reading text is not suitable for children), the pediatric RDA is being developed.

One example of describing dysarthria in terms of deviant speech characteristics is the BoDys. However, importantly, if we only identify speech characteristics, we still don't know why they occur. We need at least a correct interpretation of the speech characteristics. For example, 'imprecise consonants' can be due to weakness, spasticity or coordination problems. Or hypernasality can occur due to weakness or spasticity, etc. For those underlying deficits, different treatments will be needed.

How to train dysarthria type classification?

If we want to improve the diagnostic skills of SLTs (including SLT students) or other clinicians, the next question that arises is how to train them. We observed in our study that SLT students who had never seen patients in real life improved by 27% after a one-day course, leading to 37% agreement with the reference diagnosis (***Chapter 7***). This improvement can probably be enlarged, because learning modules for bachelor SLT students concerning a specific disorder like dysarthria are usually not restricted to one day. With many more video examples and perhaps diagnosing real-life patients more improvement is feasible. SLTs who watched videos with detailed descriptions, but without additional training, testing and monitoring their understanding, did not improve to acceptable levels of agreement. Skill training requires consistent feedback, and this can be offered by making the video training part of dedicated e-learning modules or by using blended learning as an educational method (a combination of e-learning and classroom methods). Blended learning appears to be more effective than non-blended instructions for knowledge acquisition in health professions.[168] In the case of dysarthria, in which perceptual judgments are given, it probably has an added value when the subjective perceptions can be discussed in a classroom situation instead of only being

told which speech characteristics should be heard.

The value of objective measurements for diagnosing dysarthrias

In the RDA, we explicitly chose to include maximum performance tests of speech production. When including those tests, reference values are indispensable to compare the performance of the patient to the healthy population (***Chapter 6***). However, if we want to use the reference values, we need to 'measure' speech-like performance (i.e. maximum repetition rate, maximum phonation time, maximum phonation volume and fundamental frequency range). The maximum phonation time is quite easy to analyze with a stopwatch and maximum phonation volume with a dB meter. Fundamental frequency range and maximum repetition rate are more elaborate to analyze. Software package 'Praat'[116] can be used for acoustic analysis, but this is not commonly used in the average SLT practice. The question is: do we need acoustic measurements to better diagnose dysarthrias? Experienced SLTs should be able to recognize non-pathological from pathological performance and they should be able to at least differentiate between some types of dysarthria. In 2015, an SLT-student from the Radboud University acoustically analyzed the dysarthric speech of the patients on the training videos of the RDA.[169] She analyzed speech and articulation rate, maximum repetition rate and its temporal stability and coefficient of variation, fundamental frequency range, maximum phonation time, and maximum phonation volume. Unfortunately, based on those measurements, she was not able to classify dysarthria types, but there were significant effects of dysarthria severity regarding speech rate, articulation rate, and maximum repetition rate. Recently, new attempts were made to acoustically qualify different types of dysarthria. One study with hypokinetic (n = 23) and ataxic (n = 9) dysarthrias showed that in ataxic dysarthria an increased spatial variability of sound pressure level occurs in comparison to hypokinetic dysarthria.[20] In addition, Boutsen *et al.* used an acoustic multidimensional prosody index (AMPI) for classification.[170] Spastic and ataxic dysarthrias showed longer vowel durations, whereas hypokinetic and flaccid dysarthrias showed a higher pitch. In short, it seems that we cannot rely on acoustic measurements alone to qualify dysarthria types, but some easy measurements may be helpful to distinguish at least pathological from non-pathological performances and to evaluate progression, recovery, or treatment effects.

In conclusion, with the development and publication of the RDA (NDO-V), Dutch and Flemish SLTs have the opportunity to use a standardized and validated dysarthria assessment, which is supported by 43 video examples. The RDA can be used to classify the severity and type of dysarthria, but determining the correct type of dysarthria is still challenging. For this purpose, we need better learning modules. Maybe in the future computerized techniques based on large databases of dysarthric speech can help the SLTs by making the correct diagnosis.

GENERAL DISCUSSION PART TWO

Relevance of dysarthria treatment in NMD

In ***chapter 2*** of this thesis, an overall prevalence rate of 56% for dysarthria in NMD patients was found, i.e. more than half of the patients with NMD will experience speech difficulties during the time course of their disease. Because communication is an essential need in life, dysarthria can direct-

ly affect communicative effectiveness[5,171] and participation,[5,104] as well as quality of life.[2,3,104,172] In the study of Hartelius *et al.*,[104] several very illustrative quotes of patients have been published about the burden of dysarthria on their lives, perfectly reflecting the difficulties patients deal with: "I often need to repeat what I've said, because people don't understand me"; "it's difficult to talk in a group of people that I don't know"; "my difficulties in communicating affect my possibilities to actively take part in work and studies"; "my difficulties in communicating affect my possibilities to express my personality in the way I would"; "my speech difficulties negatively affect my self-image".

Thus, dysarthria may not only directly limit communication, activities and participation, but also self-esteem and behavior. This huge and multi-facetted impact of dysarthria on a person's quality of life, in combination with the relatively high prevalence of dysarthria in NMD, underscores the importance of speech therapy interventions in this patient category. In the absence of evidence about treatment options in NMD, we will discuss our ideas according to current knowledge and personal experience.

Treatment options in NMD

Dysarthria therapy in NMD is usually not aimed at intensive motor training with the aim to recover speech functions, such as in stroke. This is because NMDs are characterized by progressive muscle weakness and gradual worsening of functions over time, which makes intensive motor training less feasible and unlikely to be effective. In NMD, therapy should be aimed at 1) education and awareness, 2) correction of inadequate compensations, and 3) learning to apply adequate compensations, either speaking techniques or use of augmentative and alternative communication (AAC) strategies. The combination of dysarthria severity and disease progression will direct the choice for a specific therapy. In slowly progressive or minimal to mild dysarthria, education, awareness, and corrections or small adjustments can be sufficient. In moderate dysarthria, therapy will be more focused on using compensatory speaking techniques, whereas in fast progressive or severe dysarthria the therapy is more focused on compensating speech functions with AAC strategies.

Education and awareness

In the case of dysarthria, education is aimed at giving insight in the process of speech production, the influence of the current disease on speech, and the effect of the dysarthria on communication. When the patient and his caregivers are aware of the consequences of the dysarthria and understand the problems at hand, this may elicit goals and solutions by the patient — instead of the speech therapist formulating solutions — that will help to increase the patient's treatment compliance. Besides, early education can prevent the occurrence of inadequate compensations.

Patients are also educated about conditions that may optimize speech. For example, the influence of posture on speech is explained, because a good posture can facilitate speech, for instance retroflexion of the head has a negative influence on the vocal quality by increasing laryngeal tension, while a flexed posture reduces the breath volume. In the case of myotonic dystrophy, myotonia may impede starting articulation, reduce speech rate or evoke disfluencies. Warming-up strategies can decrease those problems.[47] And in the case of myasthenia gravis, speaking can lead to exhaustion, which results in an increased flaccid articulation and hypernasality. Therefore, patients have to be educated about how to

prevent unnecessary muscle weakness due to relative overuse. If needed, attention for these aspects is continued during therapy.

Correction of inadequate compensations in NMD

When body functions change, compensation often occurs unconsciously. This is also true for speech: dysarthric patients often use compensatory strategies to optimize their speech. These compensatory strategies are sometimes adequate (e.g. slow articulation rate), but can also be inadequate (e.g. a hypertonic vocal use to compensate for a poor breath support). During dysarthria assessment, compensatory mechanisms get clear, most of the time because of speech characteristics that are not expected to be part of the flaccid dysarthria in NMD (i.e. hypertonic vocal use), but also because characteristics are not consistently heard. For example, the hypertonic vocal use is present during spontaneous speech, but not during maximum phonation duration. It is important to identify and to correct or stop inadequate compensations, as they may negatively interfere with the dysarthria. The already mentioned hypertonic vocal use can lead to voice problems. Patients who want to speak louder may demonstrate a quick and shallow breathing pattern, causing a chanted performance and sometimes even hyperventilation and patients with weak neck musculature may compensate by resting their head in their hands, thereby limiting the articulatory movements.

In slowly progressive NMD with minimal dysarthria, a single therapy session of one hour can be sufficient to perform dysarthria assessment, education and correction of inadequate compensations. However, it should be noted that, although patients may understand the problem and react positively to the corrections, it is generally difficult to maintain corrected speech in daily communicative situations after a single therapy session. For these patients, some kind of follow-up therapy may be needed, which will be discussed in the next section. In Box 8.1, an example of the effect of a single therapy session is given.

Learning to apply adequate compensations

When dysarthria severity is mild to moderate and disease progression is slow, therapy will be aimed at applying compensatory speaking techniques to improve intelligibility. Using the RDA for dysarthria assessment (as described in ***Chapter 5***) cannot only clarify the most impaired speech characteristics, but also the remaining capacities. Both these aspects can direct treatment focus.

For NMD, there are no specific therapy programs, like for other types of dysarthria. However, the techniques used in other therapy programs can be applicable in NMD, but this requires detailed assessment of the characteristics of the dysarthria,

Box 8.1. Example of the effect of a single therapeutic session.

A 28-year-old woman was diagnosed with a mitochondrial myopathy. During a single therapeutic session, she was made aware of her use of a slightly hypertonic voice to compensate beginning vocal weakness. In this session, she was educated about the effect of her mitochondrial disease on her voice and, after correction, she experienced that the release of the tension lowered her voice and resulted in a better vocal quality. She was immediately able to apply this modification in daily life and, therefore, a follow-up session was not deemed necessary. She was instructed, however, to contact the SLT in the case of questions or when her voice would deteriorate.

knowledge about neurological and physical aspects of the specific disease, and also skills to determine and deal with cognitive, behavioral, motivational, energetic and emotional characteristics of the patient. As a consequence, treatment of dysarthria in NMD patients is typically tailor-made and sometimes even based on trial and error. The large number of NMD and the diversity of the characteristics of the diseases in combination with patient-related speech performance and personality makes that treatments will be practice-based or context-based, rather than evidence-based. For most NMD, an SLT will have to find, together with the patient, a balance between residual capacities and participation in communicative daily situations (social, occupational or digital). In patients with severe dysarthria or anarthria, AAC strategies may be necessary to ensure such participation.

Techniques used in existing therapy programs

First, two well-known training programs for hypokinetic dysarthria in PD are the Pitch Limiting Voice Treatment (PLVT)[154] and the Lee Silverman Voice Treatment (LSVT).[155] Both programs focus on producing a loud voice, while the PLVT additionally aims at producing a low voice. In NMD, a soft voice can be a problem as well, however, the PLVT and LSVT are both intensive (3–4 times a week, 30 to 60 minutes, for 4–6 weeks) training programs, which are not direct applicable to many patients with NMD, especially not when respiratory-phonatory impairments are present. At some stage, NMD patients can be taught to save their energy and produce a loud voice when necessary, however, most of the time it is too intensive for them to persist speaking with a loud voice during all conversations.

Second, the intensive therapy program 'Be Clear' was recently published for patients with a non-progressive dysarthria due to stroke or traumatic brain injury.[173] This training program is based on the principles of motor learning and is aimed at intensive training of (compensatory) speaking techniques (i.e. rate reduction, over-articulation and a higher intensity) to improve intelligibility. Because speech therapy in NMD should not be aimed at intensive training to recover functions, Be Clear is generally not suitable for NMD. Still, its individual speaking techniques may be applicable.

As said before, speaking with a higher intensity is most of the time too intensive for NMD patients.

Over-articulation (i.e. speaking with intensified speech movements) is a technique that can be used to compensate vocal weakness when the articulation is relatively preserved. In the case of flaccid articulation, over-articulation may cost too much energy.

Speech rate reduction, however, may be easily applicable to NMD. Indeed, studies about the value of reducing speech rate showed that a wide range of patients might benefit.[48,174,175] Practically, there are different ways to reduce the speaking rate, e.g. speaking slower on demand or hand tapping on each word or syllable.

In NMD, speaking slower on demand is probably the most frequently given advise by SLTs, however, for many patients this technique is not easy to maintain during a regular conversation. While other ways to reduce the speaking rate may be easier to maintain, the fact that they are visible (hand tapping) may discourage their use. If speech rate reduction has been demonstrated to have a positive effect on intelligibility, the technique should be automated to be effective in all communicative situations. One way to practice and automate a slower

speaking rate is to use the (Dutch) 'Prosodietrainer' (Figure 8.1).[176] An example of this approach is given in Box 8.2.

In severe dysarthria, when patients are not able to reach sufficient intelligibility with compensatory speaking techniques and effective communication becomes a problem, AAC strategies should be introduced. Generally, there are not much NMD patients who depend on AAC strategies, except patients with ALS. AAC includes a wide range of strategies, from low-tech approaches, such as software on a tablet (Figure 8.2), to high-tech approaches, like an eye-tracking system. An SLT should guide the clinical decisions and implementation regarding AAC to ensure that patients can use these strategies in a functional way during daily communication. An example of the use a voice amplifier (Figure 8.3) is given in Box 8.3.

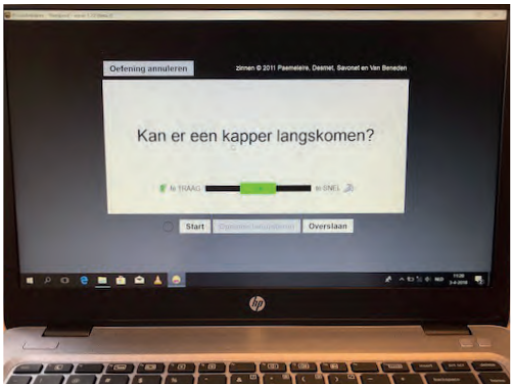


Figure 8.1. The "*Prosodietrainer*"

Principles of motor learning

Optimization and automatization of speaking techniques requires substantial practice.[177] As speech is a complex motor skill, principles of motor learning can be applied to achieve speaking skills. Motor learning is defined as 'a set of processes associated with practice or experience leading to relatively permanent changes in the capability for movement'.[178] Existing therapy programs

Box 8.2. Example of speech rate reduction.

A 39-year-old man with myotonic dystrophy type 1, who lived with his mother, complained about frequent unintelligibility, which led to irritations — even between him and his mother. Based on a thorough clinical dysarthria assessment, a moderately severe flaccid dysarthria was diagnosed with flaccid articulation, hypernasality, and fast speaking rate as key characteristics. Speech rate reduction was found to be the most effective compensatory speaking technique, which was confirmed by his mother who joined him during his visits at our outpatient clinic. He was motivated to improve his intelligibility and, also because computers were his hobby, he started practicing with the '*Prosodietrainer*'. Due to some mild cognitive problems, a diary was used to schedule his homework. The first month, he visited our outpatient clinic weekly and he practiced every day for at least 20 minutes with the '*Prosodietrainer*'. In this stage, the therapy was purely focused on learning a slow speaking rate, especially by reading sentences of increasing lengths. The next six weeks, he visited our outpatient clinic bi-weekly, while still practicing at home every day for at least 20 minutes. This stage of therapy was focused more and more on using the slow speech in conversations. It appeared, however, difficult for him to implement this technique in daily life. For this reason, a concrete implementation plan (e.g. choose the situations for using the technique, and how often it should be applied) was made, using his mother as a co-therapist. Yet, after four months, the use of slow speech was still not fully automated, perhaps due to the mild cognitive dysfunctions. However, he was able to better correct himself and to apply the technique more easily, with less misunderstandings and irritations. In addition, he accepted his mother as a co-therapist.

Box 8.3. Example of the use of a voice amplifier.

A 39-year-old man suffered from Duchenne muscular dystrophy, used mouthpiece ventilation due to a restricted pulmonary function. Therefore, he could not increase the loudness of his voice. He loved going to social events and festivals with his friends, but speaking with environmental noise was almost impossible. Also during transport in his van — while he was sitting in the back — the driver had difficulties to understand him. During a diagnostic therapy session, a voice amplifier was shown and tested. After he, his parents, and the speech-language therapist agreed on the effectiveness of the voice amplifier, a system was installed on his wheelchair. Everybody was very pleased with the final result.

like the PLVT and 'Be Clear', but also the 'Prosodietrainer', are based on those principles. However, when speech therapy is not based on a standardized program, the use of the principles of motor learning may be less explicit or consistent.

Traditionally, speech therapy exercises are based on tasks in a blocked schedule (e.g. practice the same consonant) based on constant practice variability (e.g. practice the consonant in the same word position). However, there is evidence regarding motor speech disorders that variable practice (e.g. practice of a consonant in different word positions), random practice (e.g. practice different consonants in one session), and complex targets (difficult sounds and sound sequences) can increase the effectiveness of motor speech therapy.[177] As for feedback, traditionally, the focus is on the kinematics ('knowledge of performance feedback'), e.g. placement of articulators. There is, however, evidence that feedback focused on 'knowledge of result' (whether the sound is correct) produces greater transfer of learned skills. Besides, low frequency feedback (after a couple of attempts) seems to be more effective than high frequency feedback, as is feedback provided with a delay compared to immediate feedback. Finally, there should be an increase in task complexity (reading, picture description, conversational speech) during the training period.[177]

An important prerequisite for learning speaking techniques, or for motor learning

Figure 8.2. AAC software.

Figure 8.3. A voice amplifier.

in general, is that patients have to be motivated and be able to learn. Motivational interviewing techniques can be very valuable to formulate learning goals and to elicit behavioral change.[179]

GENERAL DISCUSSION PART THREE

Relevance of treatment of dysphagia in NMD

In ***Chapter 2***, we demonstrated a high prevalence rate of dysphagia in a heterogeneous population of NMD patients. Dysphagia can have a wide range of consequences. First, dysphagia can lead to reduced oral intake of food and liquid causing malnutrition and weight loss, which may jeopardize physical health. Second, limitations in oral intake may directly affect quality of life as eating and drinking are important social activities.[180-182] Third, aspiration of liquid or food due to dysphagia combined with a reduced ability to cough may cause aspiration pneumonia and even death in the end stage of some NMD, for example myotonic dystrophy,[26,183] inclusion body myositis (IBM),[65,184] mitochondrial disease,[185] and Duchenne muscular dystrophy.[186] Finally, although nowadays rather rare, the most life threatening medical emergency is choking, i.e. the blockage of air passage into the lungs by food.[187] All these aspects underscore the relevance of dysphagia treatment in NMD, and with that the important role of an SLT in an interdisciplinary team treating these patients.

Treatment options in NMD

Like dysarthria therapy, dysphagia therapy in NMD is not focused on intensive motor training, but rather on education and awareness, correction of inadequate compensations, and learning to compensate more effectively. Treatment approaches are disease-specific, requiring knowledge of neurological and other physical aspects of the particular disease. In addition, cognitive, behavioral, and emotional characteristics of the patient have to be taken into account.

Education and awareness

In dysphagia therapy, education is aimed at explaining the normal swallowing process and the influence of a specific disease on this process to help the patient and caregivers to properly understand the problem, and to generate awareness of the swallowing function and problems that may occur.

Correction of inadequate compensations

When swallowing is difficult, inadequate compensations may become apparent, for instance when patients compensate pharyngeal transport problems with head

Box 8.4. Example of a single therapy session in dysphagia therapy.

A 54-year-old woman with facioscapulohumeral muscular dystrophy (FSHD) experienced the tendency of bread getting stuck in her throat. During a single therapy session, it was obvious that her body and head position were negatively influenced by weakness of the neck-shoulder region. She was educated about the effect of her head and body position on swallowing and these were both corrected. Despite the weakness, maintaining the head position was possible for the duration of a normal meal. During the session, she experienced that her altered position had a positive effect on the pharyngeal transport. After two weeks, a telephone call was made to evaluate the effect of the learned adaptation. She was able to maintain a good head position during eating and experienced no complaints anymore. It was agreed upon that she would contact the speech-language therapist whenever she would experience any progression of her swallowing complaints.

movements like pushing the head forwards or by tilting the head backwards (retroflexion of the head). In normal swallowing, it has been shown that the head position is an important factor and that retroflexion of the head may compromise effective swallowing.[68-70] In NMD, weak musculature of the trunk or neck-shoulder region is frequently present, which may lead to an adaptation of head and body position. This adaptation can have a negative influence on swallowing, in particular, laryngeal elevation. In addition, ptosis may be present in NMD, which can have a negative effect on head position, and thereby on swallowing as well (***Chapter 4***).[46] In minimal dysphagia, it is possible that a single therapy session of one hour, aimed at education, awareness and correction of inadequate compensations is sufficient, which is illustrated in Box 8.4.

Learning adequate compensations

Dysphagia in NMD is predominantly determined by muscle weakness (***Chapter 1***), causing deterioration of chewing and swallowing: orofacial weakness causes poor bolus formation and/or weak mastication, and pharyngeal weakness can result in pharyngeal residue. In order to reduce pharyngeal residue, swallowing techniques can be applied that focus on effortful swallowing.[73] In an effortful swallow, the posterior tongue base movement is increased to facilitate bolus clearance. The patient is instructed to swallow hard and squeeze the bolus down his throat. In NMD, this technique can be too intensive, but an effective swallow with good attention to the voluntary oral phase (bolus control and bolus transport) is possible. Other behavior techniques, e.g. consistently swallowing a bolus twice (double-swallow) or drinking during or after a meal, may reduce pharyngeal residue as well.

In addition, a few disease-specific remarks can be made. First, the Mendelsohn maneuver, which is focused on a voluntary prolongation of hyolaryngeal elevation at the peak of swallowing, has proven to be effective in a population of patients with IBM.[65] This is a clinically relevant approach, as patients maintained their oral intake without weight loss during a follow-up period of one to five years. Second, not all NMD patients tolerate the continuous use of swallowing techniques that require high levels of muscle strength. For example, myasthenia gravis is characterized by excessive exercise intolerance and effortful training causes fatigue also in mitochondrial myopathies.[188] In the case of exercise intolerance, patients should avoid to continuously use maximal muscle strength, but they can still be taught to make an effective swallow.[189] If patients are advised to use a specific swallowing technique, it is important to evaluate if the technique is effective and adequately applied in daily life. Adequate follow-up is always important to prevent non-compliance.[190] An example of a short rehabilitation process is given in Box 8.5.

When swallowing techniques are not effective anymore, adjustment of food consistencies becomes necessary in NMD. This is particularly relevant when pharyngeal residues exist. In fact, food getting stuck in the throat is the most frequently mentioned complaint by patients with NMD,[11,18,191] whereas most of them do not experience problems with fluids. This can be explained by the fact that swallowing fluids takes less effort than swallowing solid food, as muscle activity required to structurally displace the hyoid bone, tongue base and pharyngeal wall in every swallow depends on bolus size and consistency.[192] In the case of pharyngeal weakness, the solidity of the food is associated with more pharyngeal residue. Thus, softening the food is an effective way

Box 8.5. Example of a short rehabilitation process.

A 56-year-old man with auto-immune myositis suffered from recurrent pneumonias. The pulmonologist wondered if there was a relationship between his pneumonias and aspiration. During the diagnostic process by the speech-language therapist, the patient initially said that he had no swallowing problems, but after several swallowing tests he became more aware of the fact that he indeed experienced swallowing problems sometimes. He was educated about the effect of the myositis on his swallowing and went home with the homework to observe his swallowing problems in daily life. One week later, he returned to the outpatient clinic and listed the food that was difficult for him and the problems he experienced with solid food due to pharyngeal residue. While eating bread, the speech-language therapist found out that an effortful swallow almost immediately resulted in a more effective swallow. In the case of pharyngeal residue, drinking water was effective to clear the pharynx. Thus, the patient was advised to use these swallowing techniques at home. One week later, he returned and had practiced intensively with very good experiences, which motivated him to maintain using the techniques. If nevertheless he forgot to do so, in the case of food getting stuck in his throat, he was immediately able to correct himself.

to reduce pharyngeal residue[193] and can help to increase the ease of a full oral intake without exhaustion.[194,195] Besides the effect on pharyngeal residue, adjustment of food consistencies can also have a positive effect in the case of orofacial weakness, because soft food is easier to chew.

When swallowing consistently leads to aspiration or when a full oral intake cannot be maintained (e.g. because it is too intensive or it takes too long to eat enough), tube feeding is warranted. In particular, PEG (i.e. percutaneous endoscopic gastrostomy) tube feeding can reduce respiratory infections and result in weight gain.[196,197] Tube feeding does not automatically imply that oral intake is prohibited. If some consistencies are still safe to swallow, patients can maintain such oral intake. This can be very important for the quality of life.

FINAL REMARKS

Interdisciplinary treatment

Preferably, dysarthria and dysphagia treatment are part of an interdisciplinary rehabilitation approach, since the SLT is rarely the only professional involved. The interdisciplinary team will vary dependent on the problems of the individual patient. Because rehabilitation teams can be very large, we will highlight the caregivers that are most commonly involved in the rehabilitation of patients with NMD. A physician is involved for coordination and to medically optimize the physical health of the patient, which is a prerequisite for speech therapy. For example, some diseases, like myasthenia gravis and dermatomyositis, may respond positively to pharmacological therapy. Another medical intervention is dilatation or myotomy of the upper esophageal sphincter (UES) when videofluoroscopy demonstrates cricopharyngeal dysfunctioning in, for example, patients with OPMD and IBM.[25,65,198-200] This procedure requires the selection of appropriate patients, as not all NMD patients will benefit from a myotomy, because pharyngeal constriction and hyolaryngeal excursion is required to open the UES in order to get the food from the pharynx into the esophagus. If there is no propulsion of the bolus in the pharyngeal stage, the bolus might get stuck in the pharynx, even when the cricopharyngeal muscle is surgically 'opened'.

Working together with a dietician is important, in particular when food consistencies have to be adjusted. Patients should not lose weight and their awareness of nutritious food is important. A physical therapist can assist in finding an adequate posture that can be maintained during meals or to optimize physical endurance to facilitate training. An occupational therapist can train patients in applying energy conservation strategies to alleviate fatigue, which can create optimal conditions for speech therapy. Besides, an occupational therapist can supply adjusted cutlery. If necessary, a psychologist is involved in the case of acceptance problems.

The influence of external and personal characteristics

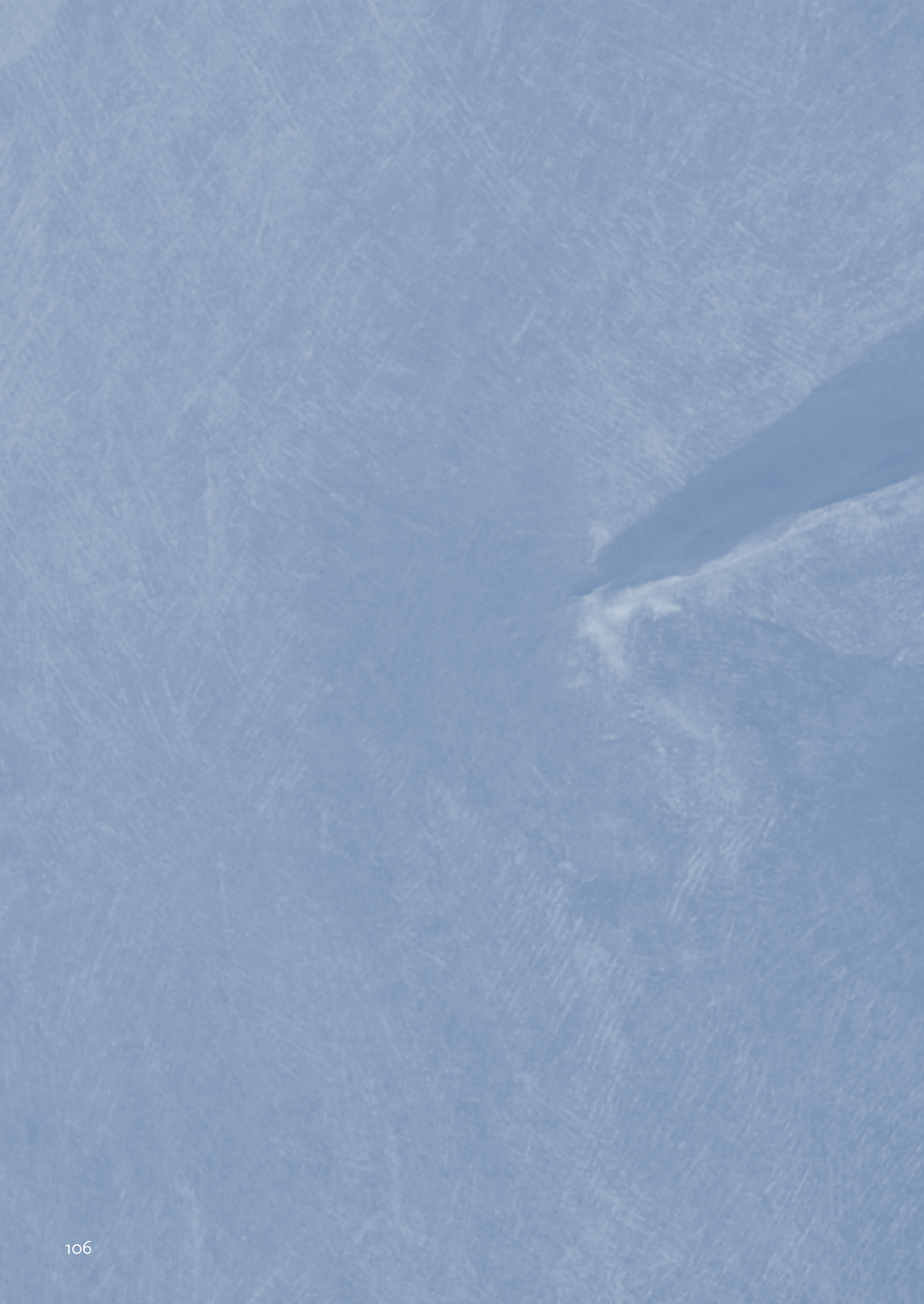
Until now, we discussed speech and swallowing therapy with the ultimate aim to improve the individual level of activity and participation.[21] Importantly, the influence of environmental and personal characteristics on the effect of speech-language therapy is indispensable: how large is the patient's need to communicate or eat; is he motivated to change his behavior; is he physically able to practice; are there cognitive impairments that need to be taken into account; is a caregiver available as a co-therapist, etc? It is important to realize that the level considered to be acceptable may differ between patients. This will be particularly dependent on personal and socio-economical aspects, and requires individual evaluation together with the patient and his caregivers. For example, a salesman will probably suffer more from a mild dysarthria than a technician. The lack of a strong relationship between the observed level of functioning by the SLT and patient burden (see ***Chapter 5***) emphasizes the value of a personalized rehabilitation approach adapted to the patient's needs. Overall, treatment of dysarthria and dysphagia in patients with NMD should be focused on education and awareness, correction of inadequate compensations, and learning adequate compensation techniques. Interventions should be personalized, taking multiple (disease-related, external and personal) characteristics into account.

Future perspectives

In our outpatient clinic, a lot of NMD patients undergo a single diagnostic session of one hour during which assessment and education are important goals, which is sometimes accompanied by learning effective compensations. We know from a previous study[28] and from patient-related outcome measures (PROMs) that patients highly appreciate these single sessions, but we do not know exactly what their effects are in individual patients. Which patients are able to change their behavior after one session and what is an optimal follow-up period?

The shortage of studies on interventions to alleviate or compensate dysarthria and dysphagia in NMD is undoubtedly due to the heterogeneity of these disorders, each with a low incidence, and due to differences in therapeutic approaches. Our review showed that about half of the patients with NMD have problems with speech and swallowing, so we presented several treatment options, but mainly based on clinical experience. In order to study the value of these approaches, there first needs to be a certain level of (inter-)national consensus among experienced SLTs, which requires both qualitative research and longitudinal studies. Only when there is general agreement about treatment options and the best timing of delivery, clinical trials become an obvious next step, using multiple centers to be able to include an adequate sample

size. However, it should be noted that even generally accepted treatment approaches in speech and swallow therapy call for personalized healthcare: interventions have to be tailor-made, based on the patient's predicted response, and taking into account individual characteristics, like cognitive and emotional functions.



9 Nederlandse samenvatting

(Summary in Dutch)

NEDERLANDSE SAMENVATTING (SUMMARY IN DUTCH)

Spreken is uniek voor de mens. Ondanks de huidige rol van sociale media, blijft de dagelijkse communicatie sterk afhankelijk van gesproken taal, oftewel spraak. Een spraakprobleem kan daarom een enorme invloed hebben op het dagelijks leven. Gestoorde spraak door een neurologische aandoening wordt een dysartrie genoemd. Een dysartrie kan veroorzaakt worden door (sub-)acute aandoeningen van het centrale zenuwstelsel (bijvoorbeeld een herseninfarct), chronische neurodegeneratieve ziekten van de hersenen (bijvoorbeeld de ziekte van Parkinson) of neuromusculaire aandoeningen (bijvoorbeeld spierziekten). In Deel I van dit proefschrift wordt, naast dysartrie, ook aandacht geschonken aan dysfagie, dat wil zeggen slikstoornissen, omdat deze vaak samen voorkomen. Beide aandoeningen worden tenslotte veroorzaakt door oraalmotorische problemen in overlappende spiergroepen, alhoewel de aansturing verschillend is (slikken is grotendeels een reflexmatig proces, terwijl spraak doelbewust is).

Zoals in ***hoofdstuk 1*** — de inleiding — is weergegeven, zijn voor dit proefschrift drie doelstellingen gedefinieerd: 1) het onderzoeken van de prevalentie van dysartrie en dysfagie in patiënten met neuromusculaire aandoeningen (NMA), 2) het zoeken naar evidentie voor de behandeling van dysartrie en dysfagie in NMA, en 3) het ontwikkelen en klinimetrisch evalueren van een gestandaardiseerde beoordeling van dysartrie bij volwassenen.

Deel I

In Deel I van dit proefschrift worden de prevalenties van dysartrie en dysfagie in NMA gepresenteerd, evenals de evidentie zoals gerapporteerd in de medische literatuur voor de behandeling van dysartrie en dysfagie in NMA (doelen 1 en 2).

Hoofdstuk 2 beschrijft het frequent vóórkomen van dysartrie en dysfagie in een grote, heterogene groep (n = 220) patiënten met een NMA. Per patiëntengroep zijn gepoolde prevalenties berekend door een weegfactor toe te kennen aan de ziektecategorie (spierziekten, aandoeningen van de neuromusculaire overgang, neuropathieën en voorhoornaandoeningen) op basis van het aantal patiënten in die categorie, aangezien de patiëntengroepen niet gelijk verdeeld waren. De prevalenties werden bepaald in zowel een ongeselecteerd cohort van poliklinische patiënten, dat wil zeggen een serie opeenvolgende patiënten die een poliklinisch logopedisch onderzoek kreeg, en een geselecteerd cohort, namelijk patiënten die specifiek het neuromusculair expertise en consultatiecentrum van het Radboudumc bezochten. De gepoolde prevalentie van dysartrie is 46% in het ongeselecteerd cohort en 62% in het geselecteerde cohort, terwijl voor dysfagie de getallen 36% en 58% bedragen. Er blijkt een matig, maar significant, verband tussen de aanwezigheid van een dysartrie en een dysfagie (r_s = 0,40; p <0,01). Over het algemeen is de ernst van de dysfagie in deze populatie licht, de dysartrie varieert meer en is matig ernstig tot ernstig in 15% van de gevallen. De meest opvallende uitkomst is dat dysartrie en dysfagie kunnen voorkomen in NMA waarbij je deze niet direct verwacht, zoals metabole myopathieën en de NMA die in de ziektecategorie 'overige myopathieën' zijn ondergebracht (bijvoorbeeld nemaline myopathieën). Het feit dat dysartrie en dysfagie veel voorkomen in patiënten met verschillende typen NMA, geeft aan dat het noodzakelijk is dat artsen kennis hebben van — en aandacht beste-

den aan — deze stoornissen, ongeacht het type NMA. Ondanks dat logopedie niet bij iedere patiënt met een progressieve ziekte zal kunnen leiden tot een betere verstaanbaarheid of slikfunctie, kunnen individueel bepaalde functionele compensaties vaak toch een positief effect hebben.

In ***hoofdstuk 3*** wordt de prevalentie van dysfagie onderzocht in een specifieke neurologische aandoening, namelijk in myotone dystrofie type 2 (DM2). Dysfagie is een bekend kenmerk van myotone dystrofie type 1, waarbij het zelfs levensbedreigend kan zijn. Het is echter nog nooit onderzocht in welke mate dysfagie voorkomt in DM2. DM2 patiënten die geselecteerd werden omdat zij slikproblemen hadden gerapporteerd op een vragenlijst naar gastrointestinale problemen, zijn gevraagd te participeren in deze studie (n = 10). Deze proefpersonen zijn klinisch onderzocht door een logopedist gespecialiseerd in NMA. Daarnaast hebben 8 patiënten toestemming gegeven voor een flexibele endoscopische evaluatie van het slikken (FEES). Een dysfagie werd bevestigd in alle patiënten (klinisch in 100%, 88% met FEES) en blijkt meer uitgesproken aanwezig in oudere DM2 patiënten. De ernst van de dysfagie is over het algemeen licht en er zijn geen patiënten die een aspiratiepneumonie hebben doorgemaakt of waarbij er sprake is van ernstig gewichtsverlies.

In ***hoofdstuk 4*** is het wetenschappelijk bewijs gezocht voor het effect van logopedische interventies in volwassen NMA-patiënten door middel van een systematische review. Hierbij zijn uitsluitend zgn. '*peer reviewed*' artikelen geïncludeerd die geschreven zijn in Engels, Duits, Frans of Nederlands. De uitkomstmaten moesten betrekking hebben op: 1) informatie en advies gericht op voedingsaanpassingen, ondersteunde communicatie en educatie gericht op de patiënt of zijn naaste, en 2) compensatiestrategieën gericht op slikken of spraakverstaanbaarheid. Casestudies werden geëxcludeerd. De zoekopdracht in de literatuur resulteerde in 1772 artikelen, maar slechts vier voldeden aan de kwaliteiten zoals omschreven in de inclusiecriteria. De methodologische kwaliteit van slechts één ongecontroleerde pre-post studie bleek voldoende. Deze studie onderzocht het effect van een ptosis op het slikken in patiënten met oculofaryngeale spierdystrofie (OPMD).[46] Deze studie liet zien dat wanneer het hoofd licht voorover gebogen was, en dus niet aangepast aan de ptosis, er een positief effect werd gevonden op de slikefficiëntie (niveau III evidentie). De conclusie van dit review is dat er slechts zeer beperkte gerapporteerde evidentie is voor het effect van logopedische interventies in volwassen NMA-patiënten.

Deel II

Deel II van dit proefschrift is gericht op de ontwikkeling van het Nederlandstalig Dysartrieonderzoek – volwassenen (Radboud Dysarthria Assessment [RDA] in het Engels), inclusief referentiewaarden voor de maximale prestatietaken (doel 3).

Hoofdstuk 5 beschrijft de ontwikkeling en validatie van het RDA. In 2007 werd een eerste versie van het (ongevalideerde) RDA gratis online beschikbaar gesteld, maar de behoefte bestond om het onderzoek te verbeteren en te valideren. De eerste stap was het verkrijgen van nationale consensus over de taken en het onderzoeksformulier. Als tweede stap werd een vragenlijst over de spraak toegevoegd voor de patiënt. In de derde stap werden het onderzoeksformulier, de ernstschaal en de vragenlijst klinimetrisch geëvalueerd. En tot slot werden er trainingsvideo's toegevoegd, waarop alle

typen dysartrie in verschillende ernstmaten gedemonstreerd werden. Bij de trainingsvideo's is een ingevuld onderzoeksformulier toegevoegd om zelfscholing mogelijk te maken. Een exploratieve factoranalyse van het onderzoeksformulier identificeerde vier factoren (articulatie, resonans, fonatie en respiratie/prosodie) die samen een verklaarde variantie van 70,3% hadden. Elke individuele factor had een goede interne consistentie. De vragenlijst bleek een zeer goede interne consistentie te hebben en de intra-class correlatiecoëfficiënten van de ernstschaal toonden een goede betrouwbaarheid van de schaal. De ernstscore van de dysartrie en de score op de vragenlijst correleerden aanzienlijk tot sterk met de Spraak Handicap Index (SHI) en aanzienlijk met het Nederlandstalig spraakverstaanbaarheidsonderzoek op zinsniveau (NSVO-Z). Gebaseerd op deze resultaten kan gesteld worden dat het RDA een valide en betrouwbaar onderzoeksinstrument is voor het diagnosticeren van de ernst van de dysartrie. Nader onderzoek moet aantonen of het gebruik van het RDA ook het diagnosticeren van het type dysartrie in de dagelijkse praktijk kan vergemakkelijken.

Hoofdstuk 6 beschrijft een studie naar de referentiewaarden van verschillende maximale prestatietaken van de spraak. Een grote groep (n = 224) gezonde Nederlandstalige volwassenen tussen 18 en 80 jaar hebben de volgende maximale prestatietaken uitgevoerd: maximale repetitiesnelheid (MRS), maximale fonatieduur (MFD), maximaal melodisch bereik (MMB) en maximaal dynamisch bereik (MDB). In eerste instantie is een univariate analyse gebruikt om te onderzoeken of er verbanden zijn tussen iedere maximale prestatietaak afzonderlijk en de persoonsgegevens (leeftijd, geslacht, lengte, beroep en rookgedrag). Vervolgens is met de data waarbij persoonsgegevens een significante invloed hadden op de betreffende taak (p < 0,05), een multivariate lineaire regressieanalyse uitgevoerd om de bijdrage van de individuele invloeden te bestuderen. Verschillende statistische modellen zijn toegepast op elke individuele maximale prestatietaak, en met behulp van de Likelihood Ratio is getest welk model het best passend model is. MRS, MMB en MDB zijn statistisch significant gerelateerd aan leeftijd. MFD is gerelateerd aan lengte. Kortom, leeftijd is in drie van de vier taken de meest bepalende factor, in het bijzonder in personen ouder dan 60 jaar. Het is onduidelijk welk onderliggend mechanisme verantwoordelijk is voor het verval na 60-jarige leeftijd jaar. Naast zowel neurologische, metabole, hormonale en fysieke veranderingen zou ook stijfheid van bindweefsel een rol kunnen spelen. Er moet worden opgemerkt dat de prestaties binnen de gezonde populatie ver uiteenlopen, iets wat de interpretatie van de individuele dysartrische spreker moeilijk maakt.

In ***hoofdstuk 7*** is onderzocht of het gebruik van de trainingsvideo's van het RDA het diagnosticeren van het type en de ernst van de dysartrie kan verbeteren in zowel ervaren logopedisten als logopediestudenten. De studenten kregen een eendaagse training gericht op het diagnosticeren van het type en de ernst van de dysartrie, waarbij gebruik werd gemaakt van de trainingsvideo's. Eén week voorafgaande aan de training en 10 dagen na de training werden de studenten gevraagd om van dezelfde 10 testvideo's het type en de ernst van de dysartrie te scoren. De logopedisten kregen toegang tot 20 online beschikbaar gestelde trainingsvideo's en tot achtergrondinformatie over de verschillende typen dysartrie. De logopedisten kregen drie weken tijd om de onlinetraining te volgen, waarbij het aantal uren wat zij eraan besteedden, registreerden. Vlak

voor en na de training beoordeelden zij eveneens 10 testvideo's. Alle scores van de logopedisten en studenten zijn vergeleken met een 'referentiediagnose' die gesteld was op basis van de consensus van zeven ervaren experts. Studenten verbeteren door de training van 29,2% naar 37,2% overeenkomst met de consensus over het type dysartrie, terwijl logopedisten van 42,8 naar 46,9% verbeterden. Met betrekking tot de ernst van de dysartrie, verbeteren studenten van 47,9% naar 52,5% en logopedisten van 53,2% naar 54,7%. Er is geen significante correlatie tussen het percentage overeenstemming met de referentiediagnose en het aantal uren zelfstudie in logopedisten, evenmin als tussen de overeenstemming en het aantal jaren ervaring. Beide trainingsprogramma's leiden niet tot een acceptabel niveau van overeenstemming met de referentiediagnose. Om dit te bereiken, zijn wellicht meer intensieve of andere vormen van training noodzakelijk.

Appendices

Appendix 1

Lekensamenvatting

APPENDIX 1 LEKENSAMENVATTING

Spraak is onmisbaar in het dagelijks leven, of je nu gezellig praat met vrienden, iemand telefonisch te woord moet staan of moet onderhandelen over de aankoop van een auto. Voor mensen met een dysartrie is dit allemaal niet zo vanzelfsprekend. Een dysartrie is een stoornis van de spraak (dat is iets anders dan een taalstoornis) ten gevolge van een neurologisch letsel — bijvoorbeeld een beroerte, de ziekte van Parkinson of een spierziekte. Afhankelijk van de plaats van het neurologisch letsel (bijvoorbeeld de kleine hersenen of de hersenzenuwen), kan een dysartrie op een bepaalde manier klinken. Er zijn zeven typen dysartrieën te onderscheiden, elk met een specifiek hoofdkenmerk: zwakte (slappe dysartrie), teveel spierspanning (spastische dysartrie), een coördinatiestoornis (atactische dysartrie), te weinig bewegingen (hypokinetische dysartrie), teveel bewegingen (hyperkinetische dysartrie), mengbeelden en een dysartrie na een beroerte (unilateraal upper motor neuron dysartrie). Omdat spraak zo belangrijk is in het dagelijks leven, heeft een dysartrie een grote invloed op deelname aan de samenleving en de kwaliteit van leven. Patiënten maken dit duidelijk, door opmerkingen zoals "ik moet vaak herhalen wat ik heb gezegd, dat is vervelend", "ik vind het moeilijk om in een groep te praten met mensen die ik niet ken" of "mijn dysartrie beïnvloedt mijn zelfbeeld negatief."

De eerste aanzet tot het onderzoek dat in dit proefschrift wordt beschreven was de gewaarwording dat bij patiënten met een spierziekte die het Neuromusculair Expertise en Consultatiecentrum van het Radboudumc bezochten een dysartrie veel vaker voor bleek te komen dan op basis van de literatuur verwacht mocht worden

Om dit beter in kaart te brengen, zijn we gaan registreren hoe vaak een dysartrie voorkwam bij patiënten, bij welk soort spierziekte dit dan was en hoe ernstig de dysartrie was. Dit wordt beschreven ***hoofdstuk 2*** van dit proefschrift. We vonden in een groep van 220 patiënten dat er in ruim de helft van de gevallen (56%) sprake was van een dysartrie. Opvallend was dat een dysartrie ook voorkwam bij spierziekten waarbij het niet direct verwacht werd of waarbij het nog niet eerder was beschreven in de literatuur. Er werd ook gekeken naar het voorkomen van een slikstoornis (dysfagie). Een dysartrie komt regelmatig samen voor met een dysfagie, omdat het dezelfde spiergroepen betreft. Een groot verschil is echter dat slikken grotendeels reflexmatig is, terwijl spraak bewust wordt aangestuurd. Bij de 220 patiënten kwam in 47% van de gevallen een dysfagie voor. In 38% van de patiënten kwamen een dysartrie en dysfagie samen voor.

In ***hoofdstuk 3*** hebben we het voorkomen van dysfagie in kaart gebracht bij patiënten met een specifieke spierziekte, myotone dystrofie type 2 (MD2). Een dysfagie is een bekend symptoom van myotone dystrofie type 1 (MD1), maar het was nog nooit onderzocht of het ook voorkwam in MD2. Het verschil tussen MD1 en MD2 is dat de klachten bij MD2 over het algemeen wat milder zijn dan bij MD1 en dat bij MD2 vooral sprake is van klachten rondom de nek, schouders en heupen, in tegenstelling tot de onderarmen en onderbenen bij MD1. In een groep van 29 MD2-patiënten gaf 41% aan moeite te hebben met het doorslikken van vaste voeding. Deze patiënten werden nader onderzocht door een logopedist en een KNO-arts die een FEES (flexibele endoscopische evaluatie van het slikken) uitvoerde. De slikklachten konden in 100% worden bevestigd door de logopedist en in 88% door de FEES. De slikproblemen waren relatief licht en kwamen vooral bij de oudere patiënten voor.

Omdat een dysartrie en dysfagie een grote impact op het dagelijks leven kunnen hebben, hebben we in ***hoofdstuk 4*** het wetenschappelijk bewijs in de medische literatuur samengevat hoe je een dysartrie en dysfagie logopedisch zou moeten behandelen. Dit bewijs blijkt er echter nagenoeg niet te zijn, omdat er weinig onderzoek is gedaan of onderzoeken van onvoldoende kwaliteit zijn. Er was slechts één studie van voldoende kwaliteit, die aantoonde dat mensen met oculofaryngeale spierdystrofie (OPMD) geleerd moeten worden om te slikken met de kin naar de borst, terwijl deze mensen juist de neiging hebben om het hoofd naar achteren te kantelen om onder hun hangende oogleden (een belangrijk symptoom van de ziekte) door te kijken.

Om een dysfagie te kunnen vaststellen bij een patiënt, zijn naast het luisteren naar de klacht (anamnese) en het klinisch logopedisch onderzoek enkele instrumentele onderzoeken mogelijk, bijvoorbeeld een FEES of een röntgenslikvideo. Voor het diagnosticeren van een dysartrie zijn er echter voor de klinische praktijk geen andere mogelijkheden dan gebruikmaken van het menselijk gehoor. De logopedist neem bepaalde spraakkenmerken waar, die vervolgens geïnterpreteerd moet worden. Omdat er in Nederland geen goed gestandaardiseerd dysartrieonderzoek beschikbaar was, hebben we er één ontwikkeld: het Nederlandstalig Dysartrieonderzoek — volwassenen (NDO-V), zoals beschreven in ***hoofdstuk 5*** van dit proefschrift. Een dergelijk dysartrieonderzoek is niet alleen belangrijk om te kunnen beoordelen van welk type dysartrie er sprake is en hoe ernstig deze dysartrie is, maar ook als aanknopingspunt voor een passende behandeling.

Bij de ontwikkeling van het NDO-V werden in meerdere discussieronden de meningen gevraagd van een groep logopedisten in Nederland en Vlaanderen over de bruikbaarheid van verschillende spreektaken en over de termen die gebruikt worden om afwijkende spraakkenmerken te beschrijven. Het uiteindelijke instrument bestaat uit een handleiding, een beoordelingsformulier, een ernstschaal, een korte vragenlijst voor de patiënt, een standaard leestekst, 43 video's van patiënten met verschillende typen dysartrie en 5 spreektaken die uitgevoerd moeten worden (spontaan spreken; hardop lezen; zo snel mogelijk lettergrepen herhalen zoals bijvoorbeeld pa, ta en ka; zo hard mogelijk roepen; zo hoog en zo laag mogelijk stemgeven en zo lang mogelijk een 'aa' aanhouden). De verschillende onderdelen zijn getoetst en het blijkt dat het NDO-V een geschikt onderzoek is, waarvan de verschillende onderdelen meten wat ze horen te meten (oftewel valide zijn), de vragenlijst en het beoordelingsformulier goed zijn samengesteld en er op een betrouwbare manier een ernstscore aan de dysartrie kan worden gegeven. Het bepalen van het type dysartrie blijkt echter niet eenvoudig.

Om de prestatie van patiënten met een dysartrie te kunnen vergelijken met gezonde personen, zijn de spreektaken ook afgenomen bij 224 gezonde personen. Op deze manier werden normaalwaarden voor de taken bepaald, zoals beschreven in ***hoofdstuk 6***.

Tot slot is gekeken of studenten van de opleiding logopedie en ervaren logopedisten het type dysartrie beter kunnen beoordelen wanneer zij geschoold zijn met de video's die horen bij het NDO-V, zoals beschreven in ***hoofdstuk 7***. Bij de studenten die een dag lang werden geschoold met de video's was het leereffect groter dan bij logopedisten die aan zelfscholing deden met behulp van dezelfde video's. Omdat uiteindelijk de logopedisten maar in 50% van de gevallen tot het juiste type dysartrie kwamen, zal vervolgonderzoek zich (ook) moeten richten op alternatieve manieren van scholing.

Appendix 2

Dankwoord

APPENDIX 2 DANKWOORD

Persoonlijk sla ik een proefschrift altijd open bij het dankwoord, omdat je in het dankwoord de persoonlijkheid van de auteur iets beter leert kennen. De meeste mensen weten van mij dat ik op schrift altijd kort en krachtig ben. Dat is dan ook de stijl van dit dankwoord.

Alle patiënten die aan de totstandkoming van de afzonderlijke artikelen hebben bijgedragen, verdienen het als eerste enorm bedankt te worden. Bij deze!

Vervolgens komt veel dank toe aan de personen zonder wie ik dit proefschrift niet tot een goed einde had kunnen brengen.

Prof. Geurts, beste Sander, de gesprekken met jou waren altijd erg inspirerend, waardoor ik na elk gesprek weer zin had om met de nieuwe ideeën of inzichten aan de slag te gaan. Ik vond het erg waardevol dat we spraakmotoriek konden bediscussiëren in het kader van de algehele motoriek. Maar het meest heb ik geleerd van jouw schrijfstijl: daar waar ik vastliep in lastige zinnen, wist jij altijd met een jaloersmakende eenvoud de boodschap weer te geven.

Prof. van Engelen, beste Baziel, jij was vooral betrokken bij Deel I van dit proefschrift. Jij kunt als de beste associëren en filosoferen, waardoor er tijdens de overlegmomenten vaak nieuwe kanten belicht werden. Het mooie aan jou is dat je je altijd afvraagt 'wat levert het de patiënt of de zorgverlener op', waardoor we altijd dichtbij de klinische praktijk bleven.

Dr. de Swart, beste Bert, wij kennen elkaar al erg lang. Bij jou is (bijna) alles mogelijk en dat bleek ook tijdens dit proces. Ik waardeer het enorm dat jij destijds hebt geaccepteerd dat ik wachtte op een promotietraject waar mijn hart lag. Waarschijnlijk is juist hierdoor het proces zonder ernstige dieptepunten verlopen. (Of ben ik echt gewoon zo'n nuchtere Zeeuw?) Ik heb heel veel van je geleerd en hoop de komende jaren ook nog veel van je te leren.

Dr. Kalf, beste Hanneke, jij bent onmisbaar geweest bij de totstandkoming van dit boekje. Jij bent enorm gedreven, maar hebt het ook geaccepteerd dat ik dit binnen de mogelijkheden van het runnen van een jong gezin deed. Daarnaast ben je uitermate correct in schrijven, waarvan ik heel veel heb geleerd. Onder het mom 'het wordt er alleen maar beter van' zijn er heel wat versies heen en weer gemaild, maar nu zet ik er toch echt een punt achter!

Ik dank de manuscriptcommissie voor het kritisch doorlezen van het manuscript en goedkeuren ervan.

Alle medeauteurs van de afzonderlijke artikelen dank ik hartelijk, met in het bijzonder dr. Jan Hendriks voor de statistische ondersteuning.

Een goede werksfeer verhoogt het werkplezier! Al mijn geweldige collega's (naast Bert en Hanneke) hebben het proces van heel dichtbij kunnen aanschouwen (alfabetisch): Irma, Frieda, Anne, Puck, Emmelien, Rosemarie, Renée, Jacintha, Judith, Kim, Pauline en Janneke. Een hele hoop persoonlijkheden bij elkaar, maar wat kunnen we goed samenwerken! We slagen er al jaren in om de patiëntenzorg op een zeer hoog niveau te houden. Naast hard werken kunnen we gelukkig ook heel hard lachen samen. Jullie hebben mij zoveel mogelijk uit de wind gehouden wanneer dat nodig was. Ik kan me geen betere collega's voorstellen!

Alle collega's van 'de kinderkant': Marjo, Lenie, Leenke, Karen, Sandra, Marloes en Sanne: we hebben elkaar de laatste jaren

(tot mijn grote vreugde) steeds meer weten te vinden, omdat kleine kinderen nou eenmaal groot worden. De discussies over verschillen tussen kinderen en volwassenen waren zeer waardevol en ik weet dan ook zeker dat het NDO-K een prachtig product gaat worden.

Ook dank ik mijn lieve paranimfen Heidi en Janneke (sorry Allan en Edith ;-)).

Heidi, wij leerden elkaar kennen op wintersport toen we allebei nog (heel) jong waren. Wat je op vakantie oppikt aan vrienden of liefdes is niet altijd een lang leven beschoren, maar dat is bij ons gelukkig anders. Ik ben jaren geleden jouw getuige geweest, jij nu de mijne!

Janneke, jij was aanvankelijk mijn partner in crime, maar bent een iets andere weg ingeslagen. Je bent in staat je hart te volgen en dat bewonder ik enorm aan je. Je bent een heel fijn mens en ik vind het geweldig dat jij vandaag naast mij staat!

Ook een heel speciaal woord van dank voor de projectgroep van het NDO-V: Harry Goos, Lotte Kromhout, Judith Kocken, Marjo van Gerven, Hanneke Kalf en Bert de Swart. We hebben heel wat uren vergaderd met cake, koek, chips en chocola. De eerlijkheid en openheid die jullie altijd hebben gehad, heb ik uitermate gewaardeerd. Ook in dit rijtje dank ik Puck Goossens die wat later in het proces aansloot toen de koppelstructuur met de HAN ontstond, evenals alle studenten die door PO-opdrachten bij verschillende projecten betrokken waren.

Er zijn veel logopedisten in Nederland en Vlaanderen die de moeite hebben genomen om mee te denken bij de ontwikkeling van het NDO-V. Allen hartelijk bedankt.

Alle leden van het OZO die de afgelopen jaren zijn gepasseerd: ik ga jullie niet allemaal persoonlijk noemen, omdat ik bang ben dat ik iemand vergeet. Ik leek misschien de eeuwige promovendus tussen alle fulltime onderzoekers, maar hierbij heb ik het tegendeel bewezen! Dank dat jullie altijd bereid waren om mee te denken over logopedische dilemma's.

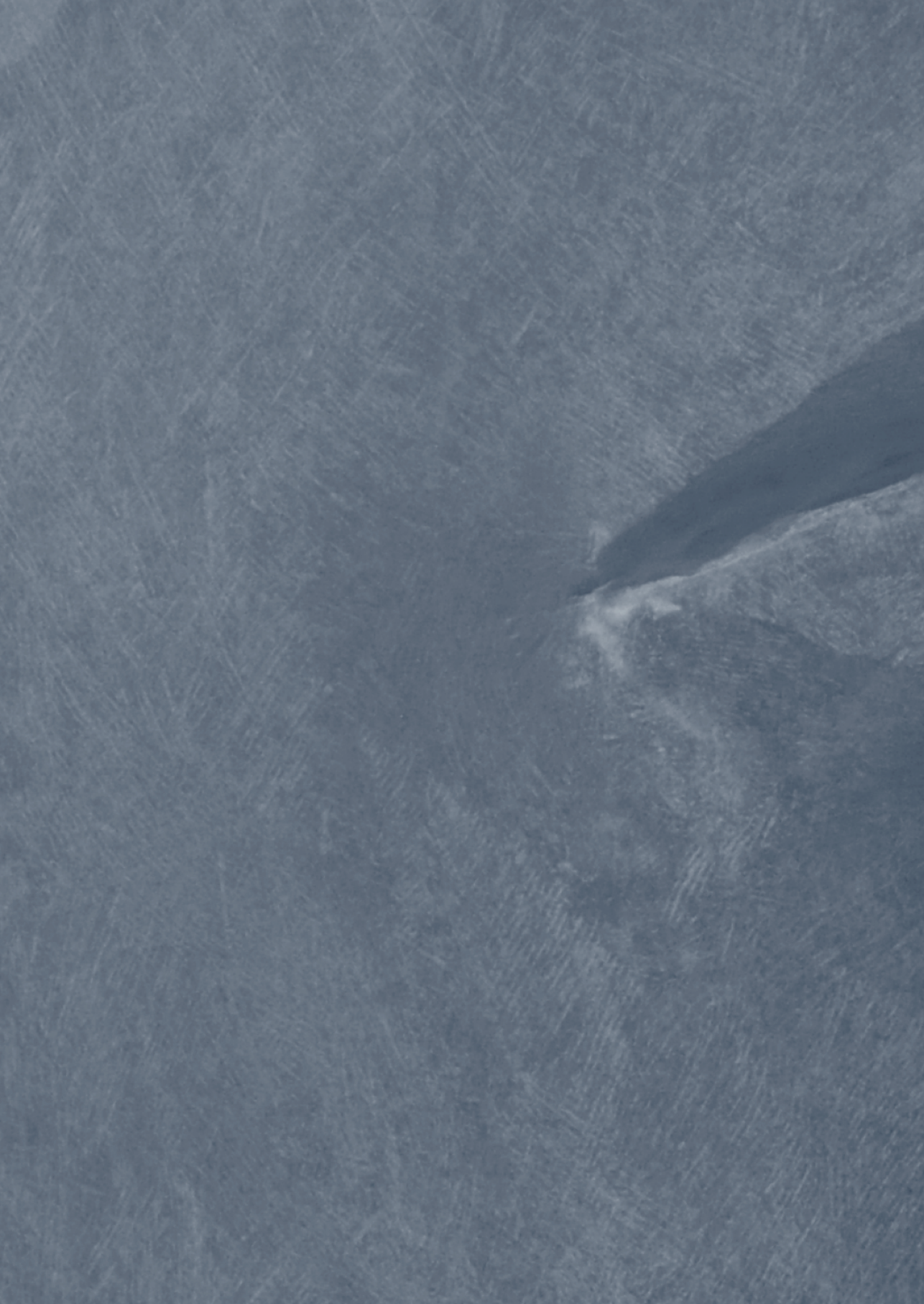
Lieve vrienden en vriendinnen, ik gooi jullie bij deze (bijna) allemaal op één hoop. Tijdens dit proces is er heel wat gegeten, geborreld, gelachen, gehuild, geshopt, gefeest, gereisd enz. enz. Ik ben blij met jullie allemaal! De enige vriendin die ik in het bijzonder wil noemen is Marieke Vloet. Jij liet je door het onderwerp inspireren om het beeld te maken voor op de omslag van dit proefschrift. Het is prachtig geworden en heeft het boekje extra bijzonder gemaakt!

Ik dank de Beachbabes en mijn tennismaatjes dat ik dit proces in een goede conditie heb kunnen voltooien.

Lieve familie en schoonfamilie, dank dat jullie altijd interesse hebben getoond en er altijd voor ons zijn geweest. Jullie wonen helaas allemaal niet zo dichtbij, maar dat maakt wel dat we er bij elk bezoek 'lekker tussenuit' waren – iets wat in een promotietraject onmisbaar is.

Lieve Luuk en Saar, jullie zijn mijn grootste schatten, bij wie ik (bijna) alle beslommeringen van het werk vergeet. Ik had natuurlijk heel wat uren zwembad, voetbalveld en dansschool anders kunnen benutten, maar ik heb geen moment spijt van het feit dat ik er (voor mijn gevoel) altijd voor jullie ben geweest.

Lieve Jasper, je hebt het als basaal onderzoeker goed uitgehouden met een klinisch onderzoeker in spe. Dat onderwijzersbloed in jou kon ik bij tijd en wijle wel drinken, maar ik moet toegeven dat het mij tegelijkertijd ook vaak heeft geholpen. En zonder jou was de lay-out van dit boekje een stuk minder fraai geworden. Dank voor alles!



Appendix 3

Curriculum vitae

APPENDIX 3 CURRICULUM VITAE

Simone Knuijt werd geboren in Middelburg op 17 maart 1973. Na het behalen van haar HAVO-diploma in 1990 (Christelijke Scholengemeenschap Walcheren, Middelburg) startte zij met de studie logopedie aan de Hogeschool van Arnhem en Nijmegen (HAN). Haar vierdejaars-stage liep Simone op de afdeling Logopedie van het Radboudumc (destijds Academisch Ziekenhuis Nijmegen). Hier werd haar enthousiasme gewekt voor neurologische spraak-, taal- en slikstoornissen.

Na haar afstuderen in 1994 kon ze parttime blijven werken in het Radboudumc, naast haar studie Spraak-taalpathologie aan de Radboud Universiteit Nijmegen. Deze studie rondde zij in 1997 af, waarna zij fulltime aan de slag ging als logopedist in het Radboudumc. De eerste kennismaking van Simone met wetenschappelijk onderzoek was het onderzoek naar de spraak-taalontwikkeling van ex-premature kinderen. Haar hart bleek echter te liggen bij volwassen patiënten met neuromusculaire aandoeningen.

Simone raakte betrokken bij onderzoek naar de indicatiestelling voor paramedische zorg bij neuromusculaire patiënten. Uit dit onderzoek kwamen de eerste artikelen van dit proefschrift tot stand. In 2011 werd, in samenwerking met het Lectoraat Neurorevalidatie van de HAN, een RAAK-subsidie verworven voor de ontwikkeling van het Nederlandstalig Dysartrieonderzoek – volwassenen (NDO-V), zoals beschreven in het tweede deel van dit proefschrift. Dit NDO-V wordt sinds 2014 uitgegeven door Bohn Stafleu van Loghum en werd in 2015 genomineerd voor zowel een RAAK-award als de Branco van Dantzigprijs. Simone werkt nog altijd met veel plezier als logopedist en onderzoeker in het Radboudumc.

Simone woont samen met Jasper Visser. Samen hebben zij twee kinderen, Luuk en Saar.

Appendix 4

List of publications

APPENDIX 4 LIST OF PUBLICATIONS

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Appendix 5

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APPENDIX 5 LIST OF REFERENCES

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