

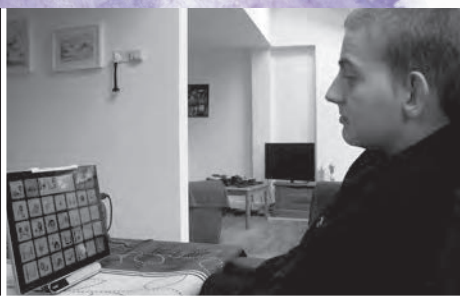

RETT SYNDROME
Communication Guidelines

Netherlands RETT EXPERTISE CENTRE
Maastricht University Medical Centre

Rettsyndrome.org
ACCELERATING RESEARCH. EMPOWERING FAMILIES.

RETT SYNDROME Communication Guidelines:

A handbook for therapists, educators, and families

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This UK and Ireland edition is printed with support from Rett UK

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This project was financed by a HeART Grant from Rettsyndrome.org.

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1st Edition: 2020

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Journal reference for the guidelines:
Townend, G.S., Bartolotta, T.E., Urbanowicz, A., Wandin, H., Curfs, L.M.G. (2020). Development of consensus-based guidelines for managing communication in individuals with Rett syndrome. *Alternative and Augmentative Communication, 36(2), 71-81.*

Citation for this book:
Townend, G.S., Bartolotta, T.E., Urbanowicz, A., Wandin, H., Curfs, L.M.G. (2020). Rett syndrome communication guidelines: a handbook for therapists, educators and families. Rett Expertise Centre Netherlands-GKC, Maastricht, NL, and Rettsyndrome.org, Cincinnati, OH.

Erratum: December 2020

Introduction, page 4: reference to "Appendix 6" was corrected to "Appendix 5".

Section 4, page 27: "decreasing" was deleted in the heading "Fatigue, alertness, sensory regulation, stress and anxiety" so that the format was consistent with other headings in this section.

Section 8, page 51: "making" was corrected to "making choices" in the Communicative Functions text box.

Section 8, page 62: reference to "Appendix 2" was corrected to "Appendix 5".

Appendix 1, pages 75, 79, 81-82, 84, 86-88: format was amended to aid clarity (line spacing and headings).

Appendix 3, pages 93-96: AAC Profile and ACETS were added; TASP and VB-MAPP were removed; and C-BiLLT entry was revised.

Acknowledgment from Authors

Many thanks to everyone who has contributed to the development of these guidelines. This project has been a joint effort by many people over a number of years and a truly international collaboration, an example of the teamwork that is such a key element in communication assessment and intervention.

Developing language and communication is a lifelong process, and we hope the guidelines shared in this handbook will support individuals with Rett syndrome, their families, and the professionals working with them as they make that journey together.

Gillian Townend
Theresa Bartolotta
Anna Urbanowicz
Helena Wandin
Leopold Curfs

February 2020

Acknowledgment from Rettsyndrome.org

As the International Rett Syndrome Foundation (known today as Rettsyndrome.org), we are proud to be a part of bringing this global research initiative to life for families affected by Rett syndrome and the community that is committed to helping them achieve more. The work of Gillian Townend, Theresa Bartolotta, Anna Urbanowicz, Helena Wandin, Leopold Curfs, and many other researchers and contributors is an excellent representation of the dedication and commitment to improving the quality of life for our children and loved ones living with Rett syndrome. It is our honour to support these distinguished researchers. We are grateful for their work.

We remain unwavering in our commitment to empowering you today while working to transform your tomorrow.

Table of Contents

All websites listed in this book can be accessed online at communication.rettsyndrome.org.

Foreword

When she was a year old, she spoke her first few words. When she was diagnosed with Rett syndrome 18 months later, she had completely lost her ability to speak. How was she to communicate her wants, needs, feelings, and opinions now?

For many families living with Rett syndrome, finding a way to communicate is one of their most challenging issues. Learning how to use augmentative and alternative communication (AAC) and learning how to facilitate their loved ones to maximise their potential takes belief in their abilities, a great deal of effort, and lots of support. Seeking knowledge and skills on how to adjust assessments and how to manage communication, parents (and professionals) find themselves in need of expert support, in need of a shared vision of how to approach these aspects. Yet, so far, little evidence-based guidance has been available to them.

To do justice to the specific challenges of Rett syndrome, such as apraxia, breathing disorder, and epilepsy, a broad consensus on assessment, intervention, and long-term management of communication is needed – for research as well as daily communication and education. The collaboration between parents, caregivers, professionals, and experts in the field of communication for Rett makes these international guidelines the valuable document that is presented here. These guidelines are an important step forward in enabling people with Rett syndrome to communicate more effectively.

Many girls, now teenagers and adults, have moved on from learning to use a few pictos to using a combination of low tech and robust vocabulary on eye-gaze devices. Young girls and boys are starting out with eye-gaze-controlled communication systems. They are learning to communicate, to tell us their wants, needs, feelings, and opinions. It is exciting to see how new possibilities are helping to change our perspective on their cognitive abilities.

Many thanks to the project team, to all the committed communication specialists and parents and caregivers who contributed to these guidelines. Our beautiful daughters and sons with Rett syndrome may be unable to use their own voices to speak, but surely these guidelines will help them move forward in communicating their needs and opinions and in speaking their minds.

On behalf of all Rett parents everywhere,

Mariëlle van den Berg
Chair, Rett Syndrome Association Netherlands (Nederlandse Rett Syndroom Vereniging)

Introduction

Background

Rett syndrome is a neurodevelopmental disorder affecting approximately 1 in 10,000 females. It occurs rarely in males. It is often associated with severe physical and communication disabilities. In most cases, the cause of Rett syndrome is linked to a mutation in the methyl-CPG-binding protein 2 (MECP2) gene.

Why we developed the communication guidelines

A severe disruption in communication skills is one of the characteristic features of Rett syndrome. This has a fundamental impact on the quality of life for individuals with Rett syndrome. Although the literature on best practices in communication assessment and intervention in Rett syndrome is growing, caregivers around the world continue to report their struggle to access appropriate, knowledgeable, timely, and ongoing assessments, interventions, technology, support, and advice tailored to the specific communication needs of the person with Rett syndrome. Communication professionals also report challenges in finding the information, training, and support they need to build up their knowledge and expertise in this area. Therefore, we developed the communication guidelines presented in this handbook to help caregivers, communication professionals, and others support the communication development of individuals with Rett syndrome.

How we developed the communication guidelines

The idea of creating guidelines came into being when we, a small group of motivated professionals working in the field of Rett syndrome and communication, joined together following the 3rd European Rett Syndrome Conference in Maastricht, The Netherlands, in October 2013. With funding from Rettsyndrome.org, we devised and led a project to develop the Rett Syndrome Communication Guidelines.

The project involved reviewing the literature and conducting surveys to identify best practices in the assessment, intervention, and longer-term management of communication in individuals with Rett syndrome around the world. Around 650 people from 43 countries participated in the project, with 490 caregivers and 120 communication professionals completing the surveys, and 36 professionals and parents forming our expert panel. As the project team, we developed draft statements and recommendations based on the findings of the literature reviews and the survey responses from caregivers and professionals. The draft statements and recommendations were then reviewed twice by our expert panel. The panel provided feedback to the project team, and we revised the statements and recommendations until consensus was reached. The final set of statements and recommendations that reached consensus became the Communication Guidelines. This means that these guidelines are firmly based on findings from the available literature combined with expert opinions from professionals and caregivers around the world.

The purpose of this handbook

This handbook was designed to share the recommended guidelines for the assessment, intervention, and longer-term management of communication in individuals with Rett syndrome and to provide relevant background information for caregivers and professionals. These guidelines are intended for all individuals with a diagnosis of typical or atypical Rett syndrome, both male and female, wherever they were born and wherever they live. The guidelines were created to be adaptable to different situations and circumstances. They do not offer step-by-step instructions that people in all countries and services must follow and apply in an identical way. They aim to provide basic information that families around the world can share with therapists and educators to enable any individual with Rett syndrome to meet their full communication potential.

How to use this handbook

The handbook is divided into eight sections, with information and recommendations relating to the following: (1) guiding principles; (2) professional practice; (3) features of Rett syndrome and coexisting conditions; (4) strategies to optimise engagement; (5) general communication assessment; (6) AAC assessment; (7) assessment of AAC system/device; and (8) intervention.

The handbook can be used in many different ways. It can serve as a guide for designing assessments that reflect best practices and enable teams to develop goals that are functional and meaningful. It can provide strategies for families and therapists to advance the communication skills of any individual with Rett syndrome. The handbook provides information on how to get started with communication as well as how to progress the skills of more experienced communicators. There is also key information that describes how the unique features of Rett syndrome may impact communication. Individuals looking for information on specific topics can consult the Table of Contents at the front of the handbook. Other readers who are new to Rett syndrome may find that the early chapters provide a foundation of information to help them as they begin their work with an individual. The handbook can serve as a tool for educating people about Rett syndrome and for advocating for the communication services and support they need.

Throughout the book, the guidelines are presented in several formats. Some are shown in boxes that stand out from the text; others are embedded within the text. All are equally important. The 'raw' guidelines (the complete, final set of statements and recommendations that reached consensus) are presented as tables in Appendix 1 at the back of the book.

Quotes from the caregivers' and professionals' surveys and from the expert panel are included throughout the handbook to highlight important information. Photographs of individuals with Rett syndrome illustrate a broad range of communication systems and settings, and short stories offer "A Parent's Perspective" on different stages of the communication journey. We are indebted to the many families who have graciously shared their photographs and their stories with us and are grateful to the even greater number of caregivers and professionals who took time to complete the surveys and engage with the project on multiple levels.

Links to a number of resources and useful websites and organisations are given in Appendix 5. This list is not intended to be exhaustive and will vary according to country and language. It is recommended, therefore, that each country produce its own list to supplement the Appendix. As far as possible, technical language is avoided in this handbook. Where this could not be avoided, the definition is usually given the first time a term is used. However, the Glossary of Terms on page 90 (Appendix 2) may also be helpful as you read through this book.

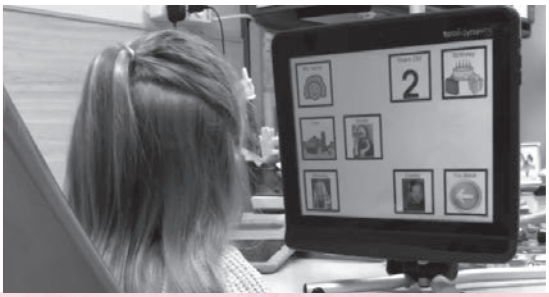
Language note:

1. In this handbook the term "individuals with Rett syndrome" is used. In some places this is shortened to "individuals" to make the sentences easier to read. Therefore, it is important to remember while reading the handbook that "individuals" always means "individuals with Rett syndrome."

2. In some places the gender neutral "they," "their," or "them" is used, but in other places "she" or "her" is used. This is not intended to exclude males with Rett syndrome, and everything in this handbook should always be read as applying to both males and females with Rett syndrome.

3. In this UK and Ireland edition spellings have been changed from the original American English to British English. The terms "speech-language pathologist" and "physical therapist" have also been changed to "speech and language therapist" and "physiotherapist" (except where these related to named people's job titles in Appendix 6) to reflect common usage in the UK.

ASSESSMENT OF AAC SYSTEM/DEVICE



"Trial periods are essential"

Assessment of AAC System/Device

Trial Periods as Assessment for an AAC System or Device

Trial periods are essential for assessing whether any AAC system or device is appropriate for an individual. This applies to both low- and high-tech systems or devices. Suitability of any AAC system or device cannot be judged adequately from a single session or a single point in time. Often, in the case of individuals with Rett syndrome, trial periods are used to assess the suitability of eye-gaze technology.

Trial Periods	During trial periods, the individual and their key communication partners should be well-supported by knowledgeable and experienced professionals who are familiar with/ trained to use the system/device on trial.

To a large extent, the success of a trial depends on how well the individual and their key communication partners are instructed and supported during the trial. This can place demands on one of the members of the team to support the other communication partners; to offer training, advice, and feedback; and to adjust the device settings and content according to individual needs. The professionals who offer support during a trial may include advisors who work for the communication aid company supplying the device and/or an SLT, OT, AAC/Assistive technology specialist advisor, and/or others who are experienced with the device/system on trial.

Trial Periods	During trial periods, the system/device should be used in multiple environments and with multiple communication partners.

The personal needs and preferences of the individual and their key communication partners may differ between systems and devices. Therefore, individuals should be able to trial more than one/multiple AAC systems and devices. They should also be allowed more than one trial of the same AAC system or device (e.g., if the first trial was judged unsuccessful due to the individual's poor health at that point in time).

Trial periods should be free/at no cost to the end user at point of use.

Trial Periods	Trial periods should be for a minimum of 8 weeks to ensure the following: • The individual with Rett syndrome gets a real chance to learn about and (attempt) to use the device/system. • The individual's primary/key communication partners get a real chance to learn about and to use the device/system.

Assessment of Device-Specific Features

During a trial period/device assessment, a number of device-specific features need to be considered.

Trial Periods	When assessing the suitability of a device for an individual, it is important to take the following features into consideration: • Portability • Options for mounting on a wheelchair, table, and wall • Robustness (durability) • Size • Weight • Battery life • Use in different environments (e.g., outside in sunshine and rain) • Ability to adjust response time/sensitivity of responses • Capacity/functions available within the device (including potential to access the internet and social media as well as control the environment)
	• Range of software available (including language programmes and symbol sets and any the individual is already familiar with) • Availability of pre-made page sets in the relevant language • Complexity of programming required to personalise/adapt the device to suit the end user • Level of technical skill/knowledge required by communication partners • Support from a wider community of families using the same device/software, including online communities (e.g., for sharing of page sets, and help with troubleshooting)
	• Support and training offered by supplier during a trial period • Long-term technical support and after-care offered by the supplier • Ease of repair in case of problems/breakdown • Obsolescence (age of model and remaining length of time that support, updates to software, parts, etc. will be available) • Cost to purchase • Cost of insurance • Availability of funding/approval for funding

Many of these aspects are brought together in Feature Matching.

Feature Matching is a model for best practice in AAC assessment that can be utilised for individuals with Rett syndrome. The aim of feature matching is to match an individual's needs and abilities with a suitable (aided) AAC system or device.

Feature Matching (Glennen & DeCoste, 1997)[1]

These system/device features should be considered:

- Type of symbols
- Language complexity, including options for expanding the language
- Type of synthetic voices and languages that are available
- Access methods
- Display and editing options (e.g., flexibility of grid sizes and settings as well as navigation)
- Portability and positioning options
- Operational and other features

[1] For further information, see: Feature Matching in: Glennen, S. & DeCoste, D. (1997). *The handbook of augmentative and alternative communication.* Singular. San Diego, CA.

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