


403

120





International guidelines for Management of Communication in Rett Syndrome

RETT SYNDROME IN NORDIC LIGHT

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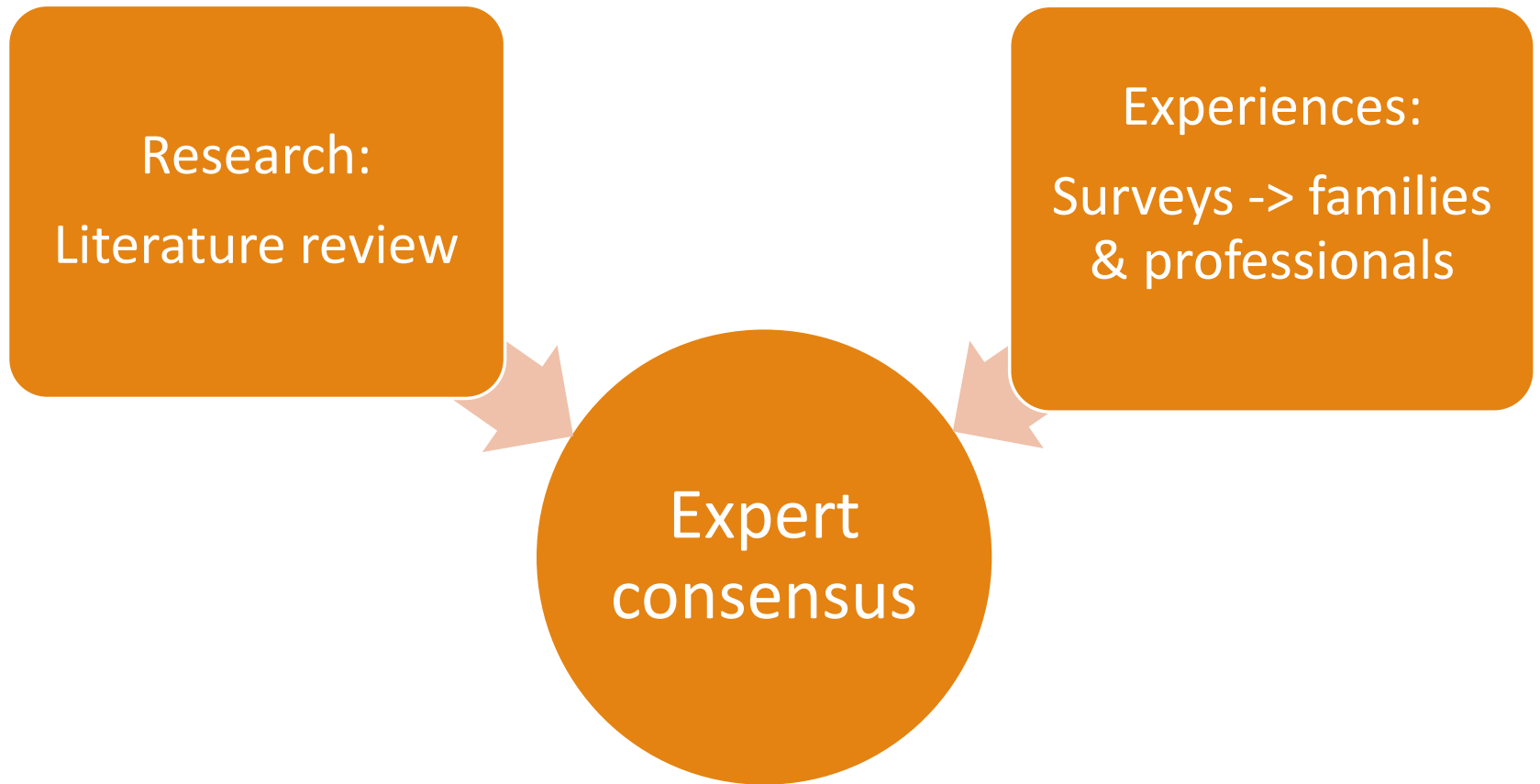


Key messages

- Everyone should believe that the individual with Rett syndrome is capable of communicating
- Value of combining research-based and clinical experience
- Team work and a shared team vision is fundamental.

How were the
guidelines developed?

Modified Delphi process





Disability and Rehabilitation

Taylor & Francis
Taylor & Francis Group

ISSN: 0963-8288 (Print) 1464-5165 (Online) Journal homepage: <http://www.tandfonline.com/loi/idre20>

Choice making in Rett syndrome: a descriptive study using video data

Anna Urbanowicz, Natalie Ciccone, Sonya Girdler, Helen Leonard & Jenny Downs

Gracie L. Cameron, Mandy Rippon, Rasmus Bang, Fabrizio Scuderi, Alessandro S. Curo

Surveys to families and professionals

Families

- Online survey
- 16 languages
- 403 responses,
- 39 countries

Professionals

- Online survey
- English
- N=120
- Countries=19
- Focus group 10

- Communication professionals with experience of working with at least one person with Rett syndrome

What did families tell us?

BELIEF IN INDIVIDUAL

She needs access
to AAC 100% of
the time

She has apraxia
and might take a
while to respond

She needs people to
believe she has the
capacity to understand
and can communicate

“She understands
more than she
can tell/show”

What did professionals tell us?

Our team shares a common vision... We ALL **assume competence** and agree on the importance of AAC. The parents are extremely involved and we all **work together**.

fun - fun - fun

Lots of **time**, LOADS of **modelling** on the communication system, a **motivated team** who is keen to learn.

A lot of **modelling** by communication partners, in different communication setting.

Expert consensus

- Panel: 36 persons
- Communication professionals working with >8 individuals with Rett for more than 2 years
- Parents – experience beyond their own child e.g. presidents of Rett associations

Delphi survey round one

- 279 statements, comments, open questions

1.2. Belief in the individual.

All communication partners should believe that the individual with Rett syndrome is capable of communicating.

The potential to communicate is frequently underestimated in individuals with Rett syndrome.

Strongly Agree

Agree

Neither agree nor disagree

Disagree

Strongly disagree

Comments:

“Presume competence” is a term that reflects a belief about individuals with Rett syndrome. Please tell us what “presume competence” means to you.

Delphi survey round one

7.7 Case scenarios.

To help us formulate pathways through communication intervention for individuals with Rett syndrome please comment on the following scenarios.

You are working with a young child newly diagnosed with Rett syndrome who is a beginning communicator with no speech and no AAC system. What goals do you identify first and what strategies do you use?

You are working with an individual who appears to have limited interest and engagement with any activities and high levels of agitation. It is difficult to identify motivators. What goals do you identify as priority and what strategies do you use?

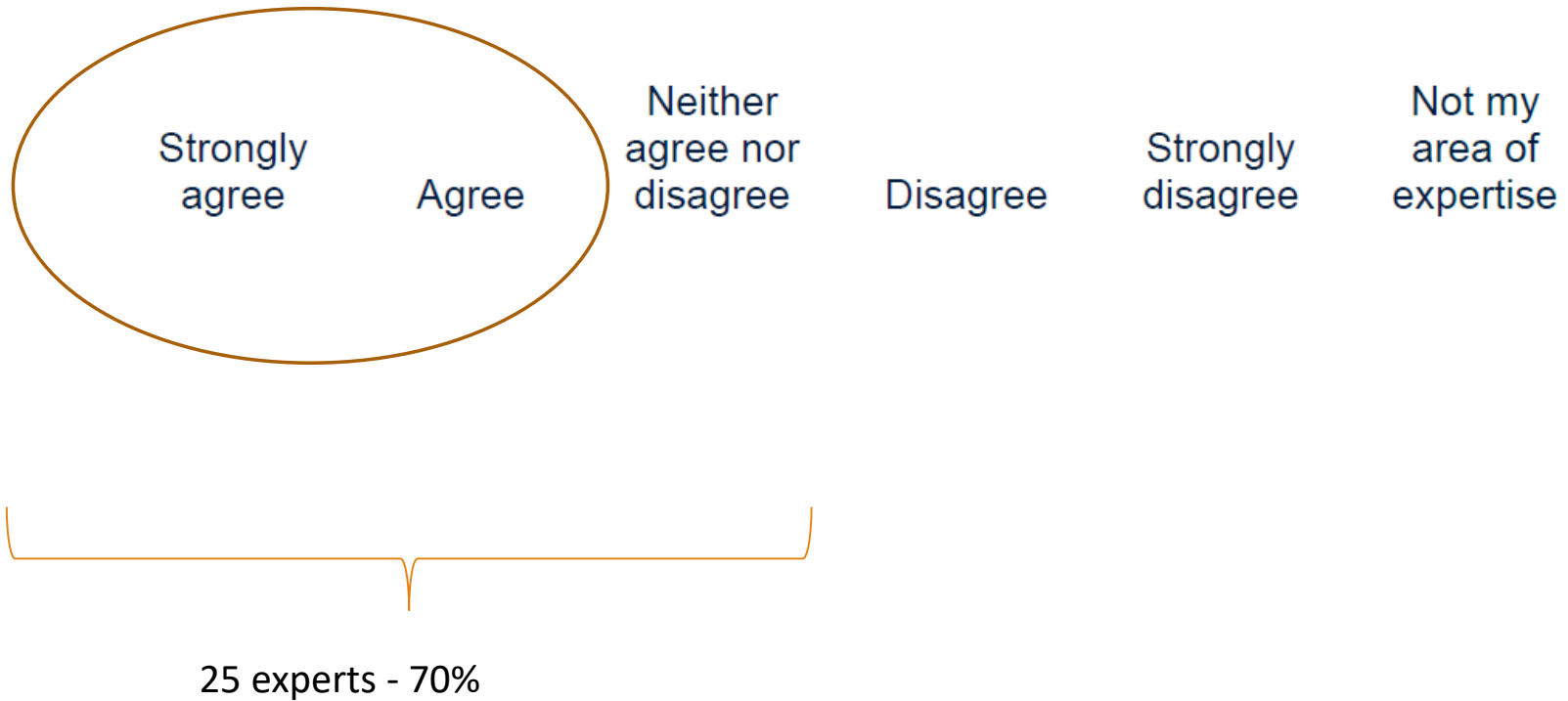
Delphi survey round one

7.9 Additional considerations for using eye gaze technology.

Please rate your agreement with the following issues and tell us what strategies you put in place to deal with them.

	Strongly agree	Agree	Neither agree nor disagree	Disagree	Strongly disagree	Not my area of expertise	What strategies do you put in place to deal with this?
A high tech device/SGD with eye gaze access cannot be used with an individual who is ambulant or has some level of mobility.	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="text"/>
A high tech device/SGD with eye gaze access cannot be used with an individual who wears glasses.	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="text"/>
A high tech device/SGD with eye gaze access cannot be used with an individual who has poor head	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="text"/>

How did the experts respond?



Delphi survey round two

129 statements:

- Consensus level not reached in Round One
- Rephrased
 - based on comments from Round One, or
 - for the sake of clarity, or to
 - To reduce reduplication/overlap
- Extra information from Round One

46	Goals to expand the range of communicative functions (e.g. requests, answers, protests, comments, questions, descriptions, greetings) should be included in intervention programs for individuals with Rett syndrome.	1	A	29/31 (93.5%)
47	Language intervention should begin with a small number of vocabulary items that can be expanded once the individual with Rett syndrome has demonstrated competence at this level.	3	A	18/32 (56.3%)
48	To expand an initial vocabulary, goals should include words that cover a range of ideas, interests and meanings including emotions, social and academic language.	1	A	29/32 (90.6%)
49	Language intervention should begin with exposure to a robust vocabulary (including core and fringe words) so that potential for communication is unlimited.	1	A	26/32 (81.3%)
50	Limiting the number of choices is a necessity for some individuals with Rett syndrome.	0	A	05/34 (14.7%)

Language intervention should begin with a small number of vocabulary items that can be expanded once the individual with Rett syndrome has demonstrated competence at this level.

The guidelines

Guidelines

- Download from webpages
- Statements
- Booklet
- Scientific article

Guiding principles

“All communication partners should believe that the individual with Rett syndrome is capable of communicating.”

Strongly agree 36/36 100%



Symptoms that impact

- Hand stereotypies. **Strongly agree 29/35 (82,9%)**
- Dyspraxia/apraxia **Strongly agree 33/34 (97,1%)**
- Epileptic seizures. **Strongly agree 33/33 100%**
- Vacant spells **Strongly agree 32/33 (97%)**
- Fatigue, reduced alertness **Strongly agree 33/35 (94,3%)**
- Heightened anxiety **Strongly agree 31/35 (94,3%)**



General strategies

“When communicating with individuals with Rett syndrome in any situation, it is important to address and talk directly to the individual with Rett syndrome rather than talking about them in their presence.”

Extremely important 34/35 (97,1%)



Assessment

“There are no prerequisite skills that must be demonstrated before aided AAC should be considered.”

Strongly agree 27/31 (87,1%)

“At the time of diagnosis individuals with Rett syndrome should be referred for AAC evaluation.”

Strongly agree 26/35 (74,3%)



Intervention

“Reading stories together is an activity that is appropriate for all individuals with Rett syndrome to develop language, communication and literacy skills.”

Strongly agree (31/34 91,2%)



Next step

- Finalize the guidelines
- Translation into multiple languages
- Implementation and training projects
- Publications in scientific journals



To discuss the guidelines further
and contribute your views/experiences

Speak to Helena at the conference or send an email to the project

manager: g.townend@maastrichtuniversity.nl

THANK YOU FOR YOUR ATTENTION