

Rett Syndrome: Health and Behavioral Analyses

July 2011

Dear MRSRA Family Member,

We are working to build a collaborative partnership among the MRSRA, Gillette Children's Specialty Healthcare, Gillette Lifetime, and the University of Minnesota. We are committed to using research to make a difference for girls and women with Rett Syndrome. To do so, we are developing a set of related projects that will begin moving this initiative forward. One of our goals is to successfully apply for federal research funding through the International Rett Syndrome Foundation (IRSF) and the National Institutes of Health (NIH). We think that we have a great opportunity to improve clinical and educational outcomes and meaningfully contribute to scientific knowledge about Rett Syndrome.

The specific projects we are planning are based directly on the survey we conducted approximately a year and a half ago and shared the initial results at last year's MRSRA's family meeting (2010) at Gillette. Specifically, we'd like to start addressing in much more detail many of the behavioral and health issues girls and women with Rett Syndrome live with. These studies will help us examine issues related to pain, language and communication, challenging behaviors, and autonomic (arousal) functioning. We are hoping for a number of girls and woman with Rett syndrome and their families to participate. This packet is designed to provide an overview of the different studies. Some of the studies can be accomplished in your home whereas for others we can make arrangements to meet with you during an already scheduled Gillette visit. If you have any questions, would like additional information, or would like your child/ward to participate in one or more of the studies, please contact me by phone at 612-626-8697, or email at symon007@umn.edu.

Thank you for your time,

Frank J Symons, Ph.D.
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Pain Assessments

Purpose – Why are we doing this?

We are interested in finding an accurate way of assessing pain in girls with Rett syndrome.

Procedures – What we would ask you and your daughter to do.

We will do a brief pain examination with your daughter consisting of range of motion movements of all four limbs and the head and neck. While this physical exam is being done the individual will be video recorded and the video will later be studied for signs of pain using an assessment scale. We will also ask you to record two 5 minute segments of each individual with Rett Syndrome. One of these videos will need to be recorded when you think your daughter is in pain and the other segment of video will need to be recorded when you are confident that your daughter is not in pain. We will measure the pain behaviors in each video using a well validated pain assessment scale to see if the scale does a good job differentiating between the pain and no pain videos that you provide.

Outcomes – What we hope to learn.

This study will help us determine whether scales that have been used with individuals with other disabilities are useful for assessing pain in girls with Rett syndrome. In the future, this may be helpful in assessing pain due to medical treatments or devices, as well as trauma. It may also help us understand if some individuals with Rett syndrome experience pain differently than others.

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Non-invasive measures of autonomic nervous system function

Purpose – Why are we doing this?

We are interested in testing some new technologies to see what they tell us about how your child's body is working.

Procedures – What we would ask you and your daughter to do.

A special camera would be used that senses the temperature of the objects photographed. It uses a special lens coating that allows it to record heat coming from anything it points at. This camera would be used to take pictures of your child/ward's hands and feet, and any other areas that you think might cause your child pain. We will look at the pictures to see if we can see changes as a result of injury, or altered blood flow.

We would also examine some chemicals in your child/ward's saliva that reflect how your child's body responds to stress. To collect the saliva, a salivette (like a toothbrush with an absorbent cotton dental roll, like when you go to the dentist) is swabbed inside your child/ward's mouth, and then squeezed into a plastic vial. We would show you how to collect the saliva. We would ask that you collect four samples a day at specified times (just after your child wakes, mid-morning, mid-afternoon, and before bed) for two days. It only takes moments to collect the saliva.

Outcomes – What we hope to learn.

To date, no one has used these technologies with individuals with Rett syndrome. The results of this study will help us determine whether thermal cameras are a useful, non-invasive tool for assessing factors related to pain and autonomic dysfunction in individuals with Rett syndrome. The saliva study would allow us to determine whether the daily patterns of cortisol activity in individuals with Rett syndrome are different from other individuals. This may, in turn, give us more information about how the bodies of individuals with Rett syndrome respond to stress.

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Language environment study

Purpose – Why are we doing this?

The primary goal of this study is to assess the frequency of verbal interaction that individuals with Rett syndrome experience in their day-to-day lives, as well as to identify activities or contexts that promote social interactions for these individuals.

Procedures – What we would ask you and your daughter to do.

We will provide a small audio recording device that is worn in a pocket in a special vest to be worn by your child. The device will be turned on in the morning when your child wakes up and worn throughout her daily activities. It will be turned off and removed before bed in the evening. The audio recording captured by the device will be analyzed by a special computer program that allows us to see how often individuals are interacting verbally with your child, as well as how often she vocalizes. By comparing these results to a log of your child's activities, we can see what types of activities are associated with the active periods of social interaction.

Outcomes – What we hope to learn.

We hope to learn more about how much language individuals with Rett syndrome are exposed to on a day-to-day basis, and whether there are patterns regarding the contexts or activities that promote interaction. This may have implications for helping parents, teachers, and other caregivers promote the most responsive and language-rich environment for individuals with Rett syndrome.

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Functional analysis of challenging or problem behavior study

Purpose – Why are we doing this?

There are many answers (genetic, biological, psychological, environmental) to the question of ‘why do people with developmental disabilities engage in challenging behavior’ but very few answers specific to girls with Rett syndrome. The main goal of this study is to better understand how the environmental piece contributes to repetitive, disruptive, destructive, self-injurious and aggressive behavior in Rett syndrome, looking carefully at the circumstances that may impact the onset of behavior, how long it lasts, and how often it happens. Understanding what environmental contexts and events are related to challenging behaviors can lead to effective ways of reducing them.

Procedures – What we would ask you and your daughter to do.

This project would involve 3 steps: an interview, an informal observation, and a formal functional analysis. The interview involves research staff talking with you and other caregivers about the circumstances involved when your child/ward engages in challenging behaviors (1-2 hours). We would then observe in your home during daily routines that you identify as difficult for you and your child/ward to look for patterns of challenging behavior in relation to things like task demands, changes in routine, or the presence or absence of people (2-3 hours; depending on where you live we may assist you in collecting video of these routines and sending them to us at the University instead of visiting you in your home). Finally, our research staff would use the information from the interview and observation to design an individualized functional analysis, which involves your child/ward experiencing 3-4 ‘test’ conditions over the course of approximately 90 minutes. The test conditions will allow us to more definitively determine what environmental circumstances ‘trigger’ your child/ward’s challenging behavior, and what ‘outcomes’ the behavior produces for them. The conditions will be designed individually for your child based on what research staff learn in the interview and informal observation. During this analysis, you or another primary caregiver (parent, teacher) will be coached to interact with and respond to your child/ward in certain ways such as giving task demands, limiting your attention, and responding to challenging behavior. This analysis will take place in your home, or on campus at the University of Minnesota, and may lead into (with your permission/interest) the Functional Communication Training intervention study described on the next page.

Outcomes – What we hope to learn.

We hope to identify specific circumstances in the environment that contribute to your child/ward’s challenging behaviors, and the extent to which the behaviors serve a communicative function for girls with Rett syndrome. This may have implications for decreasing the undesirable behaviors and improving communication. We will write up a report of our findings, review it with you, and give you a copy.

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Functional communication training study

Purpose – Why are we doing this?

If we identify environmental circumstances that contribute to your child/ward's challenging behavior, and determine that she reliably produces certain outcomes with these behaviors, we would evaluate an instructional procedure for teaching her an alternative way of producing those outcomes. A lot of research supports this procedure (called Functional Communication Training) in developmental disabilities generally, but very little of it has included girls with Rett syndrome.

Procedures – What we would ask you and your daughter to do.

Once we have identified a function for your child/ward's challenging behavior (see the Functional Analysis study, described above – common 'functions' include attention from others, access to toys or food, or escape from a task) we will identify a different behavior she can use to generate that same outcome. Depending on her existing skills, we might teach her to use spoken words, push a button that produces spoken words, point to a picture, or use a manual sign. Your child/ward will learn that her new communication behavior will get the desired outcome, which may reduce the need for (and frequency of) the challenging behavior.

Outcomes – What we hope to learn.

We hope to learn more about how well girls with Rett syndrome can learn to use an alternative method of communication to request a desired outcome, and whether this is successful in reducing challenging behaviors.

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Meal schedules study:

Purpose – Why are we doing this?

To determine whether smaller, more frequent meals reduce gastro-intestinal pain and challenging behaviors in individuals with Rett syndrome.

Procedures – What we would ask you and your daughter to do.

We would provide a calendar that would specify whether your child is to receive 3 standard meals, or 6 smaller meals on a given day. Your family would be asked to follow the schedule and fill out a simple food diary to track your child's food intake for each day of data collection. We would also provide a digital video camera with which you would record a 15-minute video of your child following the last meal of the day. Our research team will code these videos to determine whether there are changes in pain-related behaviors or challenging behaviors between 3-meal and 6-meal days.

Outcomes – What we hope to learn.

We hope to learn whether smaller, more frequent meals may reduce the intestinal discomfort that individuals with Rett syndrome often experience, and whether some challenging behaviors that they exhibit may be an expression of discomfort.

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Heart rate study

Purpose – Why are we doing this?

Other studies have shown that heart rate changes can change how we respond to our world. The specific purpose of this study is to examine how heart rate changes may be related to challenging behaviors among individuals with Rett syndrome.

Procedures – What we would ask you and your daughter to do.

If your child is participating in the Functional Analysis study, we would collect heart rate measures from your child during those sessions. When we collect heart rate from your child, we will ask you to raise your child's shirt and put an elastic belt around your child's chest. A small clip on the belt will record heart rate information onto a watch without using wires, so the belt will not get in the way of your child's movement. There is latex in the belt and it will not work if your child has a cardiac implant (like a pacemaker). Your child will wear the belt for up to 2 hours.

Outcomes – What we hope to learn.

We hope to learn whether we can observe patterns in heart rate related to certain behaviors such as self-injury and repetitive behaviors. This may give us information about whether these behaviors play a role in how your daughter regulates her levels of arousal, for example to calm herself when she is over-excited, or to provide stimulation when she is bored.