



“Behavior in CHARGE syndrome”

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AUTHORS AND THEIR CONNECTION TO THE CHARGE SYNDROME FOUNDATION:



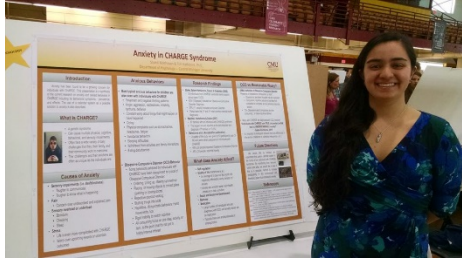
Tim Hartshorne, PhD, (on left, with several of the other authors) is a professor of psychology at Central Michigan University (CMU) in Mt. Pleasant, Michigan. He is the director of the CHARGE Syndrome Lab at CMU, where he mentors students and continues research focused on the behavioral and family issues in CHARGE syndrome (CS). He has published extensively on CS, has presented at countless conferences, and has received many awards and recognition for his CS work around the world. Tim has been involved with the CHARGE Syndrome Foundation since the beginning and currently serves as the Chair of the Clinical Advisory Board.



Kasee Stratton, PhD, is assistant professor of school psychology at Mississippi State University (MSS) in Starkville, Mississippi. Once a student of Tim Hartshorne, she is now a recognized CS expert in her own right. She has established a CS Lab at MSS where she mentors many students. Her research involves behavioral and quality of life issues in CS, including the areas of adolescence and sexuality. Kasee has presented at multiple CS conferences and is an active member of the CHARGE Syndrome Foundation.



David Brown is a deafblind educational specialist who has been working with children with CS since 1983. His observations and writings have contributed greatly to the understanding of behavior in CS and best practices for evaluation of children with CS. David is based in San Francisco, California, and is currently working freelance following his retirement from California Deafblind Services. David remains an active member of the CS community, presenting information on sensory deficits and CS around the world.



Shanti Madhavan-Brown is a third-year doctoral candidate in school psychology at Central Michigan University where she is working with Tim Hartshorne. Her research is focused on anxiety in children with CS. She has attended multiple CS conferences as part of the Hartshorne CHARGE Lab at CMU.



Megan Schmittel is a pre-doctoral intern at Monroe-Meyer Institute, based in Carney, Nebraska. Her doctoral research through the Hartshorne CHARGE Lab at Central Michigan University is focused on the development of social play in children with CS. She has presented her research at several CS conferences around the world.

SUMMARY OF THE PAPER:

Abstract: Specific unusual behaviors are often associated with specific genetic syndromes. This is sometimes described as a behavioral phenotype. In contrast to providing a psychiatric diagnosis, a behavioral phenotype describes what is unique to the behavior associated with specific syndromes. While behaviors in CHARGE are as complex and variable as other aspects of the syndrome, there are some commonalities that raise the possibility of common sources for these behaviors. This article addresses how pain, sensory issues, and anxiety may impact the behavior of individuals with CS, and how the development of self-regulation skills might help to mitigate some of these behaviors.

Additional summary: Behavior issues are extremely common in CS. In one study, nearly half of all individuals with CS were on psychotropic medications, with most of those on more than one medication. Individuals with CS often are given one or more labels to describe their behaviors, ranging from OCD or ASD and Tourette to schizophrenia. Table 1 is a description of a common CS behavioral phenotype. Although there may be some behaviors which are inevitably part of CS, perhaps most of the behaviors observed can be explained in large part as responses to the child's environment – both internal and external. The authors posit that many of the behaviors stem from difficulty with self-regulation. Self-regulation, in turn, is dramatically influenced by the behavioral triangle of pain, anxiety, and sensory issues (Figure 1). Understanding the origins and status of pain, anxiety, and sensory issues in a particular child may help in development of strategies to increase the ability to self-regulate and at the same time reduce the problem behaviors observed.

The authors describe each portion of the behavior triangle and the aspects of CS that contribute to each. For example, sources of pain can range from migraines to surgical pain to chronic sinusitis. Strategies for attempting to identify sources and level of pain are discussed. Sensory issues are not simply anatomic differences in vision, hearing, balance, etc., but also in the ability to integrate sensory information, which is often very impaired in CS. "Similar to autism spectrum disorders, poor integration of sensory systems may interfere with well-modulated responses to stimulations." Sensory integration is also related to the need for various types of self-stimulation. Deliberately teaching self-regulation strategies

is discussed as a strategy to help decrease problem behaviors. Behaviors also improve as communication improves.

The authors end with this statement (emphasis added): *“Challenging behavior is not inevitable in individuals with CS, but it is highly predictable given problems with pain, sensory issues, and anxiety. Pain should always be checked first, particularly if there has been a change in behavior. Helping children to compensate for multi-sensory impairment is essential. Anxiety can be reduced by increasing routine and predictability. Children need to be supported in the development of self-regulation skills.”*

WHAT DOES THIS MEAN TO FAMILY/PERSON WITH CHARGE?

This is a nice summary of what we understand about behavior in CS to date. Those of you who have heard Tim or David present or have read their previous publications will recognize much of this information. Behavior is complex, and it is helpful to read about many of the same issues again and again in slightly different formats. This paper brings together much of what the “behavior team” has been looking at for the last 30 or so years.

SHOULD I READ IT? SHOULD ONE OF MY DOCTORS READ IT?

Yes, you should read it, even if you have read other things by these authors before. It would also be a good paper to share with anyone working with your child. Anyone new to CS should also read some of the many resources on these topics available on the CHARGE Syndrome Foundation website (especially the [Educational Professional Packet](#)) and through [NCDB](#).

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