

ARTICLE

Genetic Counseling of Adults With Williams Syndrome: A First Study

KATRINA FARWIG, AMANDA G. HARMON, KRISTINA M. FONTANA, CAROLYN B. MERVIS, AND COLLEEN A. MORRIS*

We report on a study of genetic counseling to 43 adults with Williams syndrome (WS). Participants were initially asked what they knew about how WS occurs. Genetic counseling was provided with a focus on the basic genetics of WS, recurrence risk, and on participants' attitudes toward socio-cultural topics. Forty-nine percent indicated that they would be okay or happy if their baby had WS, 44% said they would be sad or upset, and 5% were unsure. The sad/upset group was significantly older than the okay/happy group and a significantly higher proportion of the former group indicated that they did not plan to have children. During the post-counseling session participants were questioned to determine if they recalled the facts previously presented. Eighty-one percent correctly gave the odds that their child would have WS. Fifty-three percent considered the 50–50 odds to be a high chance. After genetic counseling, 61% were able to state something that had been taught and 88% indicated that they would want to test their baby for WS before birth. Ninety-eight percent would recommend genetic counseling to others. Findings indicate that based on the type of genetic counseling provided in this study, the majority of individuals with WS—a genetic disorder associated with intellectual disability but with relative strengths in (concrete) language and in verbal rote memory—are able to learn simple facts about the genetics of WS and are eager to respond to socio-cultural questions regarding topics typically included in genetic counseling sessions. © 2010 Wiley-Liss, Inc.

KEY WORDS: genetic counseling; Williams syndrome; intellectual disability

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INTRODUCTION

Williams syndrome (WS) is a well-recognized condition consisting of distinctive facial features, delays in development, connective tissue abnormalities, a cognitive profile characterized by particular strengths and weaknesses, and specific personality characteristics. WS is caused by a sub-

microscopic deletion of chromosome 7q11.23 including 26 genes. Cardiovascular disease, (most commonly supra-valvar aortic stenosis) is the most significant cause of morbidity and mortality in this disorder. The WS cognitive profile is associated with relative strengths in (concrete) language and in verbal short-term memory and severe weakness in visuospatial construction

(including visual–motor integration) [Mervis et al., 2000; Mervis and Morris, 2007; Mervis and John, 2010]. The WS personality profile is associated with overfriendliness, high approach behavior (including indiscriminate approach to strangers), anxiety (including repetitive question-asking behavior), empathy, and visibility [Klein-Tasman and Mervis, 2003; Järvinen-Pasley et al.,

Additional supporting information may be found in the online version of this article.

Katrina Farwig, MS, is a senior genetic counselor and instructor in the Department of Pediatrics at the University of Nevada School of Medicine and is interested in genetic counseling in special populations.

Amanda G. Harmon, MS, is a doctoral student in the Department of Psychological and Brain Sciences at the University of Louisville. Her primary research focus is on the language, cognitive, physiological, and behavioral development of children with Williams syndrome, Down syndrome, and duplication of the Williams syndrome region.

Kristina M. Fontana, MA, has been providing genetic counseling services in southern Nevada for over 20 years and is a member of the Genetics Division of the Department of Pediatrics, University of Nevada School of Medicine.

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Colleen A. Morris, MD, is the Chief of the Genetics Division in the Department of Pediatrics at the University of Nevada School of Medicine. She has studied the natural history and genotype–phenotype correlations of Williams syndrome for 25 years and also has research interests in syndrome delineation and identification and treatment of children with Fetal Alcohol Spectrum Disorders.

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2008]. About 65% of children and adolescents with WS meet DSM-IV criteria for Attention Deficit Hyper- activity Disorder (ADHD) [Leyfer et al., 2006]. Adaptive behavior is typically at or below the level expected for intellectual ability (IQ), with relative strength in socialization skills and relative weakness in daily living skills (e.g., self-help skills) [Mervis et al., 2001; Mervis and John, 2010; Howlin et al., in press]. Very few adults with WS live independently, although with appropriate services greater independence is possible [Davies et al., 1997; Stinton et al., 2010].

WS occurs sporadically. An individual with WS has a 50% risk of having a child with WS. There are several reports in the literature of adults with WS who have had offspring with WS [Morris et al., 1993; Sadler et al., 1993; Ounap et al., 1998; Scherer et al., 2005]. However, no studies of genetic counseling of individuals with WS have been reported. In the present study, we provided limited genetic counseling to adults with WS. The goals of the study were to determine if individuals with WS would be able to acquire basic factual information about the genetics of WS, including the recurrence risk; to explore characteristics of individuals with WS that might influence whether or not they planned to have a child; and address participant satisfaction with the counseling session.

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his/her Genetic counseling involves both

own attitudes so that he/she can provision of factual information
and use these as a basis for decisions regarding psychosocial discussion. Two
approaches the issues addressed during genetic to modification of genetic counseling
intellectual counseling. Finucane argued that it is approaches for individuals with
described. important for the goals set by the genetic tual disabilities have been
teaching counselor to be limited, specific, con- Watkins et al. [1989] focused on
crete, and related to the reason for basic facts to the individual being
referral. counseled using simplified language
In the present study, we combined and repetition as needed. They
also the approaches advocated by Watkins recommend using yes/no type
questions et al. [1989] and Finucane [1998b, 2010] rather than open-ended
questions when for communicating genetic information trying to elicit factual
information from to individuals who have intellectual individuals with intellectual
disability. disability. As advocated by Finucane In contrast, Finucane [1998b, 2010]
[1998b, 2010], the genetic counseling emphasized the importance of a
psy-

more
feelings
the
provision of
traditional
facts, including a brief description of chromosomes, the fact that if a woman with WS (or the partner of a man with WS) is pregnant there is a 50–50 chance that the baby will have WS, and the fact that there is a prenatal test available to psychosocial approach in which determine if the baby has WS before it is a more conversational style born.

focused on feelings and attitudes takes precedence over the more

METHODS

traditional focus on provision

Participants of facts.

This study was reviewed and approved by the University of Nevada, Reno Institutional Review Board. Adults with genetic counseling approach relies on a genetically confirmed diagnosis of WS the ability of the individual attending a national Williams Syn- counseled to be able to reason about drome Association Conference were abstract concepts, to have solid invited to participate in the study. The of number and quantity, and to exclusion criteria were individuals under act in terms of not only his/her 18 years and those who were nonverbal. needs but also what is best for Informed consent was obtained from 44 members of the family. She argued that individuals, of whom 1 later withdrew in contrast, most individuals with intel- from the study. The remaining 43 lectual disability reason concretely (in participants (28 females and 15 males) Piagetian terms, are in the

preopera-
periods of
formal
understand
tend to
they
perspective of
Finucane,
is to
explore
dorm, 3 (9%) that they lived independ-

were 19–53 years old ($M=28.8$ years, $SD=8.6$ years). Demographic information regarding living arrangements and employment was determined based on numbers or quantity reliably and participants' self-report. Thirty participants (70%) indicated that they lived at home with family members, 6 (14%) that they lived in a group home, 3 (7%) that they lived in a supervised apartment or a dorm, 3 (9%) that they lived independ-

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ently with roommates, and 1 (2%) that she lived alone. Three individuals (7%) stated that they were still in school, 28 (65%) stated they were working, and 12 (28%) indicated that they were neither in school nor working. Of the 28 participants who stated that they were working 3 (11%) indicated that they were employed full time and 25 (89%) said they were employed part time. Types of employment reported by the participants included: food service (8), manufacturing (2), retail (6), housecleaning or maintenance (4), office work (6), child-care (1), and animal care (1).

Materials

Visual aids depicting chromosomes and the missing chromosome piece causing WS were used to enhance the overall effectiveness of the genetic counseling session (see supporting information Fig. 1 which may be found in the online version of this article).

Procedure

The procedure included two components: a genetic counseling session and

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a post-counseling interview. Participants were seen individually and sessions were audiotaped for later analysis. The questions asked during the study are listed in Table I.

Genetic counseling. The genetic counseling session was conducted by a board certified genetic counselor (K.F.). The interview began with a series of demographic questions to ascertain information regarding the participant's type of residence and school or employment status, followed by an open-ended question: (1) "What do you know about how Williams syndrome occurs?"

The genetic counseling session included both verbal and visual information interspersed with both open- and closed-ended questions. The counselor explained that a person's chromosomes come from his/her mother and father and that WS occurs because a tiny piece of chromosome 7 is missing. The participant was told that the piece was missing at the very beginning of his/her mother's pregnancy and that the missing piece could have come from either the mother or the father. The counselor

stressed the fact that the missing piece was no one's fault. The participant also was told that he/she had a 50–50 chance to pass the chromosome with the missing piece to each of his/her children. The counselor also told the participant that there was a test available that a pregnant woman could have to determine if the baby had WS before the baby was born. After the counselor explained the genetics of WS, the participant was asked the next three questions: (2) If you or your partner were pregnant, would you want to know if the baby had WS before the baby was born? (3) How would you feel if the doctor told you your baby had WS? (4) Do you plan to have children?

Post-counseling interview. The post-counseling interview was conducted by a board certified geneticist (C.A.M.) immediately after the genetic counseling session. The interviewer asked the participant: (5) Did you learn anything new? If the participant responded affirmatively, he/she was asked, (6) What did you learn? The interviewer also asked, (7) Would you recommend genetic counseling to other people with WS?

TABLE I. Coding Scheme for Questions Asked During the Counseling and Post-Counseling Interviews

Question Codes % Agreement

Counseling interview

1. What do you know about how WS occurs? WS is genetic/Don't know 95 2. If you or your partner were pregnant, would you want to know if your baby had WS before the baby was born?

Yes/No/Don't know 95

3. How would you feel if the doctor told you your baby had WS? Okay-happy/Scared-sad/Don't know 100 4. Do you plan to have children? Yes/No/Don't know 100

Post-counseling interview
5. Did you learn anything new? Yes/No 100 6. If so what? More about genetics of WS/Risks/WS is

not anyone's fault/No response

98

7. Would you recommend genetic counseling to other people with WS?

Yes/No/Don't know 100

8. If a person has WS, what is the chance they will have a child with WS?

50-50/Don't know 100

9. Is that a high or a low chance? High/Medium/Low/Don't know 100 10. Would you be interested in testing the baby for WS during a

pregnancy?

Yes/No/Don't know 100

11. Have you ever taken care of a baby or a child (like babysitting)? Yes/No 100

aNo one responded with an incorrect percentage. bCoded "yes" only if participant indicated that he/she had taken primary responsibility for a child multiple times.

and (8) If a person has WS, what is the chance he/she will have a child with WS? If the person answered correctly, he/she was asked, (9) Is that a high or a low chance? If the participant indicated that he/she did not know what the chance of having a child with WS was, the interviewer reminded the participant that the chance was “50–50” and then asked (9). The interviewer then asked, (10) Would you be interested in testing the baby during the pregnancy for WS? The final question was, (11) Have you ever taken care of a baby or a child, like babysitting?

Response coding

Responses to all 11 questions were coded independently by K.F. and C.A.M. The questions, the codes used to classify the responses, and the percentage of coded responses on which the coders agreed for each question are provided in Table I. The few coding discrepancies were resolved by discussion.

RESULTS

Statistical Analyses

Categorical data were analyzed using Fisher’s exact test, the McNemar test, or the binomial test. Continuous data were analyzed using the Mann–Whitney U- test or the Kruskal–Wallis test. Below we first report analyses for responses to the questions asked during the Genetic Counseling portion of the study and then present the findings for the Post-Counseling Interview questions. In the final section, we present results of analyses relating responses from the two components of the study.

Genetic Counseling Session

Early in the genetic counseling session, each participant was asked what he/she knew about how WS occurs. Twenty-five participants (58%) indicated that they did not know how WS occurs. The remaining 18 participants (42%) stated that WS was genetic. Two of these individuals also correctly stated the

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recurrence rate. Some of the comments included:

- (1) Female (21 years): Well, I heard it has to do with 7th chromosome and your gene. That’s all I really know. (2) Female (22 years): It has to do with your genes, missing elastin. (3) Female (28 years): I understand that it’s a gene missing on the number 7 chromosome, but that’s all I understand. I understand that I could have children but there is a 50% chance.

After K.F. completed the genetic counseling, she asked each participant, “If you (or your partner) were pregnant, would you want to know if your baby had Williams syndrome before the baby was born?” Most of the participants (34 of 43, or 79%) responded affirmatively. Seven participants (16%) said they would not want to know and two (5%) indicated they were unsure.

When asked how they thought they would feel if they found out the baby had WS, 21 of 43 participants (49%) said they would be okay or happy; 19 (44%) said they would be scared, sad, shocked, or upset; and 3 (7%) indicated they did not know how they would feel. Examples of participants’ comments included:

- (1) Male (32 years): I wouldn’t mind. It would be just like me. (2) Male (20 years): Personally, I like having Williams syndrome because it is different from everybody else. I’d feel perfect about it. (3) Female (43 years): I would feel very shocked at first. It would take me time, a long time to explain to my child. I wouldn’t know how, I just wouldn’t know how to explain that I just heard about this and when I heard I was very shocked myself. I would show the baby who she is, so she or he would really understand who they are first, to build their confidence, to let them know I am there for them and I would not walk away from them and I would stay there as long as they need me and I would teach them at home in a more—I would rather do that in a friendlier confines than put them in

a school where they don’t know what the heck is going on. They could be teased. They could have their self-esteem

damaged to a point where it would take me time to help them readjust. It took me forever to figure out how could this happen. (4) Female (37 years): I'd probably feel a little sad. I know it has been hard for me. I've been going through you know poking, prodding, having a barium, having tests all my life. It would be sad for my kid for a while. And I'd have to tell my kid people are going to call you retarded, call you mean names. It would be sad for my kid so I think that is why I decided not to have kids. I didn't want mine to go through that. So I understand that if I were to have one, that would or that could happen.

We hypothesized that older individuals with WS would be less likely than younger individuals to react egocentrically to the news that their child had WS. Therefore, we expected that the group of individuals who said they would be sad or upset if their child had WS to be significantly older than the group of individuals who said they would be okay or happy. To test this hypothesis, we computed a Mann–Whitney U-test with participant's age as the dependent variable. The three individuals who indicated that they did not know how they would feel were excluded. The mean age of the 21 participants who said they would be okay or happy was 25.48 years (SD=5.98 years). In contrast, the mean age of the 19 participants who said they would be sad or upset if they found out their child had WS was 33.00 years (SD=9.65 years). The group whose reaction would be to be scared or upset was significantly older than the group whose reaction would be to okay or happy ($U=7.71$, $P=0.02$).

Participants were also asked if they planned to have children. Eleven of the 36 individuals who had not been sterilized (31%) responded affirmatively, 18 (50%) stated that they did not plan to have children, and 7 (19%) indicated that they were uncertain. To determine if

there was an association between participant gender and intent to have children (Table II), a Fisher's exact test was computed. Results indicated that the relation between participant gender and intent to have children was not significant ($P=0.49$). Mean ages were 26.64 years ($SD=6.25$) for the group that planned to have children, 30.83 years ($SD=9.70$) for the group that did not plan to have children, and 22.43 years ($SD=2.76$) for the group that was undecided. The results of a Kruskal–Wallis test indicated that the groups did not differ significantly in age ($H=5.73$, $P=0.06$).

Post-Counseling Interview

At the start of the post-counseling interview, the participant was asked if he/she had learned anything new. Of the 43 participants, 8 (19%) stated they had not learned anything new and 35 (81%) responded affirmatively. However, only 27 of the 35 (63% of the total sample) were able to tell the interviewer at least one fact that had been taught. Examples of the responses of participants who were able to state something that had been taught included:

(1) Female (43 years): I learned about the chromosomes, which, which I could never understand what the chromosomes were until she showed me the picture of the missing gene. Now I've realized what, what this is all about now. (2) Female (37 years): But I did learn that, what I didn't know is, I thought with, like the two 7 genes, one whole one was missing and I learned a little tiny part was missing instead. And I learned that there's a 50–50 percent chance.

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TABLE II. Relation Between Gender and Procreation Plan

Gender

Procreation plan

Do not plan to have children Plan to have children Not sure

Female 12 5 5 Male 6 6 2

(3) Female (53 years): There's a test for parents for Williams syndrome which I didn't know about. (4) Male (32 years): Yeah I learned that the parents don't cause me to have Williams syndrome. It wasn't my mom or my dad, it was the genetic, it was the genes itself. Because of the chromosome, one of them showed that it was together, one of them showed that it was a piece missing. So that's how I got there. That's amazing. I never knew that; blew my mind whenever, when I heard that. Wow. It wasn't my parents fault. It was nobody's fault. It just happens. (5) Female (25 years): I learned that if you have a child that either you can have a whole chromosome 7 as a mom and a dad or the child can have the deletion of number 7 too. And I told her [K.F.] I don't wanna have that risk.

Participants were asked if they would recommend genetic counseling to other individuals with WS. Forty-two of 43 (98%) responded affirmatively and 1 (2%) responded negatively.

To determine if participants knew what the odds of their having a child with WS were, the interviewer asked, "If a person has WS, what is the chance they will have a child with WS?" Thirty-five out of 43 participants (81%) correctly stated "50 – 50" and 8 (19%) said they did not know. No one responded incorrectly. If the participant indicated that he/she did not know the odds, the interviewer reiterated that there was a 50–50 chance of having a child with WS. Participants were then asked if the 50–50 chance of their baby's having WS was a high or a low chance. Although not included as one of the alternatives provided by the interviewer, some participants stated that the chance was "medium" or "in the middle." Twenty-three of the 43 participants (53%) stated that the chance was high, 6 (14%) said the chance was medium, and 12 (28%) said the chance was low. The remaining two (5%) stated that they did not know if "50–50" was a high or low chance.

To determine if there was an association between age and participants' perception of the level of risk represented by "50–50," a Kruskal–Wallis test was computed. The two participants who said that they did not know if "50–50" was a high or low chance were excluded. Mean age was 29.8 years ($SD=9.1$ years) for the group who said the risk was low, 26.5 years ($SD=8.5$) for the group who said the risk was medium, and 29.1 years

(SD=8.9) for the group who said the risk was high. Perception of risk did not differ as a function of participants' age (U=0.967, P=0.62).

Participants also were asked if they would be interested in testing a baby during the pregnancy to find out if the baby had WS. Thirty-eight out of 43 (88%) responded affirmatively and 5 (12%) responded negatively.

Participants were asked if they had ever taken care of a baby or child (like babysitting). All of the participants who responded affirmatively provided specific examples. Participants were coded as having child care experience only if their responses indicated that they took care of a child regularly; participants were coded as not having child care experience if they responded that they had never taken care of a child or if their examples indicated that they only rarely took care of a child. Twenty-six out of 43 participants (61%) were coded as having childcare experience and 17 (39%) were coded as not having childcare experience.

To determine if there was a relation between having regular child care experience and planning to have children, a Fisher's exact test was performed (Table III). The seven participants who were sterilized were excluded. Results indicated a significant relation (P=0.035). A post hoc one-way Fisher's exact test examining the responses of the

group who did not have regular child care experience indicated that significantly more of these individuals indicated that they did not plan to have children than would have been expected by chance ($P=0.002$). A post hoc

A post hoc one-way Fisher's exact test examining the responses of the group who did not have regular child care experience indicated that significantly more of these individuals indicated that they did not plan to have children than would have been expected by chance ($P =0.002$).

one-way Fisher's exact test examining the responses of the group who did have regular child care experience indicated that their responses were approximately evenly divided between planning to have children, planning not to have children, and being unsure ($P=0.80$).

To determine if there was a significant relation between perception of

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TABLE III. Relation Between Procreation Plans and Child Care Experience

Procreation plan	No regular child care experience	Regular child care experience
	Do not plan to have children 11 7	Plan to have children 2 9

TABLE IV. Relation Between Perception of Risk of Having a Child With Williams Syndrome and Procreation Plans

Classification of risk

amount of risk that a baby would have WS and planning to have children (Table IV), a Fisher's exact test was conducted, excluding the seven individuals who were sterilized. The relation between these variables was not significant ($P=0.94$).

We hypothesized that within the group who indicated that they would be sad or upset if their baby had WS, more individuals would be likely to state that they did not plan to have a child than would have been expected by chance.

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In contrast, within the group of individuals who indicated that they would be okay or happy if their baby had WS, we did not expect to find a relation with

Procreation plan	Do not plan to have children	Plan to have children	Not sure
	High 8 5 4	Medium 2 3 1	Low 6 3 2

High 8 5 4 Medium 2 3 1 Low 6 3 2

stated plans for procreation. To address these possibilities, we conducted two one-way Fisher's exact tests (Table V). As predicted, more individuals in the first group stated that they did not plan to have children than would have been expected by chance ($P=0.008$). Individuals in the second group were approximately evenly divided among the three types of procreation plans ($P=0.78$).

Analyses Relating Responses From Genetic Counseling and Post-Counseling Sessions

Participants were asked twice if they would want to know if their baby had WS before the baby was born, once by the genetic counselor (K.F.) after she finished providing the counseling information and once almost immediately afterward by the interviewer in the post-counseling session (C.A.M.). Thirty-nine participants (91%) gave the same response both times, with 34 (79%) responding affirmatively and 5 (12%) responding negatively. Four participants (9%) changed their responses. All four indicated during the post-counseling interview that they would want to know if their baby had WS prior to the baby's birth. Two of the four had originally indicated that they would not want to know prior to the baby's birth and two had indicated that they were unsure.

At the beginning of the genetic counseling session, participants were asked what they knew about how WS occurs. During the post-counseling interview, participants were asked what they had learned during the counseling. Nine of the 43 (21%) participants were not able to state any fact about the genetics of WS on either occasion, 7 (16%) stated that WS was genetic at the beginning of the genetic counseling session but were not able to state anything they had learned during the post-counseling interview, 16 (37%) did not know that WS was genetic prior to the counseling session but were able to state a genetic fact that had been learned when asked during the post-counseling interview, and 11 (26%) stated facts indicating that they knew WS was

genetic on both occasions. We hypothesized that the number of participants who had not known that WS was genetic at the start of the counseling session but were able to state at least one fact indicating that WS was genetic after counseling would be significantly larger than the number of participants who had stated that WS was genetic prior to genetic counseling but who after counseling were not able to indicate at least one fact that they had learned. A one-tailed McNemar test indicated a significant difference in the direction predicted ($P = 0.047$), indicating that significantly more participants were able to state that WS was genetic after genetic counseling than before.

DISCUSSION

The present study had three goals: (1) to determine if individuals with WS would be able to acquire basic factual information about the genetics of WS, including the recurrence risk, after limited genetic counseling; (2) to provide a preliminary exploration of the characteristics of individuals with WS that may influence their procreation plans; and (3) to assess participant satisfaction with the genetic counseling provided. Below we discuss our findings relative to each of these goals and then consider the limitations of this study.

Our first goal was to determine if individuals with WS would succeed in acquiring basic factual information about WS after limited genetic counseling. The results of our study clearly indicate that as a group, individuals with WS were able to learn at least some of the basic facts about WS from a brief genetic counseling session designed to take into

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TABLE V. Relations Between Stated Response to Learning Their Baby Had Williams Syndrome and Procreation Plans

Procreation plan

Response to learning baby had WS

Okay or happy Sad or scared

Do not plan to have children 6 10 Plan to have children 8 3 Not sure 5 1

account the strategies suggested by Watkins et al. [1989] and Finucane [1998b, 2010] for working with individuals who have intellectual disability.

The results of our study clearly indicate that as a group, individuals with WS were able to learn at least some of the basic facts about WS from a brief genetic counseling session designed to take into account the strategies suggested by Watkins et al. and Finucane for working with individuals who have intellectual disability.

In particular, following genetic counseling 81% of the participants correctly answered a direct question concerning the chance that the child of a person with WS would also have WS. Furthermore, 63% of the participants were able to state at least one additional fact that was taught during the genetic counseling session. Importantly, significantly more individuals were able to state one or more facts indicating that WS was genetic after the counseling session than before.

The second goal was to begin to identify characteristics of individuals with WS that may have an influence on whether or not they plan to have children. We identified at least one such factor: the response that the individual said he/she would have to finding out

that his/her baby had WS. The range of sentiments expressed mirrored that expressed by individuals in the general population when told their child would have a genetic condition [Bennett et al., 2003]. Thus, across the group of participants a wide range of emotions was expressed, with only 7% indicating that they did not know how they would feel. A common theme expressed by the 44% who said that they would be shocked, sad, or upset if they

found out their baby would have WS was that the child would face stigmatization, as they themselves had. The themes expressed suggest that the participants in this group were able to look at the situation from the baby's point of view, rather than simply considering themselves. Significantly more

A common theme expressed by the 44% who said that they would be shocked, sad, or upset if they found out their baby would have WS was that the child would face stigmatization, as they themselves had. The themes expressed suggest that the participants in this group were able to look at the situation from the baby's point of view, rather than simply considering themselves.

individuals in this group indicated that they did not plan to have children than would have been expected by chance.

A common theme expressed by the 49% who said that they would be okay or happy if they found out the baby had WS was that the baby would be, "just like me." These individuals responded in the egocentric manner that Finucane [1998a,b, 2010] had found to be common among individuals with intellectual disability. Within this group, individuals were approximately equally divided

among those who planned to have a child, those who did not plan to have a child, and those who were uncertain.

The group of participants who indicated they would be sad or upset for their baby to have WS was significantly older than the group who indicated they would be okay or happy. This pattern suggests that the attitudes of individuals with WS toward finding out that their baby had WS may change with maturity.

Participants' responses to the question of whether they considered "50–50" to mean their baby had a high chance or a low chance to have WS were not related to their procreation plans. The pattern of responses for the group that considered 50–50 to correspond to a high chance of having a baby with WS was almost identical to that of the group that considered 50–50 to correspond to a low or medium chance. This finding may be due to the phenomenon of acculturation described by Finucane [1998a] in which having a child is viewed as "the great equalizer [p. 38]." In addition, it is possible that many of the participants did not understand the interviewer's question but were either unaware that they did not understand or unwilling to ask for clarification so simply responded at random or gave the response they thought the interviewer wanted. Even in adulthood, many individuals with WS have difficulty comprehending both conceptual/relational terms and analogies (or idioms or metaphors) [Mervis et al., 2003; Mervis and John, 2010]. In the question asked by the interviewer, "high–low chance" is an analogy expressed with conceptual/relational terms, making it especially difficult for many individuals with WS to comprehend. At the same time, comprehension monitoring (realizing when you do not understand what has been said to you and requesting clarification) is also limited for most individuals with WS [e.g., John et al., 2009]. In situations in which a question is not understood, it is very common for an individual with WS (or other syndromes associated with intellectual disability) to simply respond rather than to request clarification. [Abbeduto et al.,

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2008; John et al., 2009]. Thus, in many cases the participant's response may not have reflected a considered decision.

Our third goal was to assess participant satisfaction with the genetic counseling provided in this study. Studies of the responses of individuals in the general population to genetic counseling have considered a variety of outcomes to gauge patient satisfaction. Examples include reduction of patient anxiety or depression [Michie et al., 1997b], gaining information [MacLeod et al., 2002], comprehension of risk after counseling [Sorenson et al., 1981; Rona et al., 1994; Michie et al., 1997a], and general satisfaction with the genetic counseling encounter [Michie et al., 1997b; Veach et al., 1999; Davey et al., 2005; Aalfs et al., 2007]. As described earlier in this section, the data from the present study suggest that if participant satisfaction is judged based on the suggested criterion of gaining information, the counseling provided in this study was successful. If participant satisfaction is measured by comprehension of the risk, the genetic counseling provided also was reasonably successful. Although we cannot be confident that the participants specifically understood what is meant by the 50–50 chance that their child would have WS, most of the participants were able to state these odds. Furthermore, based on the participants' comments, it is likely that the majority at least understood that these odds meant that maybe their baby would have WS and maybe it would not. Finucane [2010] would consider this a satisfactory outcome when counseling individuals with an intellectual disability syndrome associated with autosomal-dominant inheritance, as indicated by her discussion of a case study of a woman with 22q11.2 deletion syndrome.

Rona et al. [1994] found that even patients who did not correctly recall the risk figure appropriate to them were often still pleased to have had genetic counseling. In their study, although only one-third of the couples correctly recalled their risk, 84% stated that they were pleased to have had genetic counseling. In keeping with their finding, we also addressed participant sat-

isfaction by asking the participants directly if they would recommend genetic counseling to other people with WS. The participants' responses made it clear that by this criterion, they were highly satisfied with their genetic counseling session: 98% responded affirmatively.

This study was the first in which genetic counseling was provided for individuals with WS. As such, it had

certain limitations. In order to include a large group of participants, the study was conducted at a national family support conference, the Biennial Meeting of the Williams Syndrome Association. At a minimum, attendance at this conference suggests that the participants' families were likely to be interested in learning more about WS. Although their family's socio-economic status was not assessed, it is likely that in most cases it was above average given that their families were financially able to attend a national conference. As IQ was not assessed as part of this study, we were not able to examine systematically possible relations between general IQ or abstract nonverbal reasoning ability and either measures of what participants learned during genetic counseling or their responses to the socio-cultural topics discussed. As long-term follow-up was not part of the study, we do not know if the information the participants learned during the study was retained or if reproductive decisions were impacted by the genetic counseling provided.

In summary, we have demonstrated that genetic counseling may successfully be provided to individuals with a specific syndrome that includes intellectual disability. The responses of most of the participants indicated that they had learned new information regarding the genetics of WS, including the chance that a person with WS would have a baby with WS. Limiting the amount of information presented and using visual aids was helpful in communicating the factual information. Adults with WS responded very well to the inclusion of socio-cultural topics in the counseling and were able to express their emotional responses to the possibility that their

baby would have WS. In keeping with Finucane's [1998b] statement that "women with mental retardation are more like than unlike other counselees in their emotional responses to genetic morbidity (p. 80, italics in the original)," the responses of the participants in our study reflected a wide range of emotional reactions similar to that found across individuals in the general population.

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