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Self concept in people with Williams syndrome and Prader–Willi syndrome

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Abstract

This study explored self concepts in matched groups of adolescents and adults with Williams syndrome (WS) and Prader–Willi syndrome (PWS), using Damon and Hart's [Self-understanding in Childhood and Adolescence, Cambridge University Press, New York, 1988] semi-structured interview. The main findings were that the WS participants were more productive in their responses to the interview, providing more self characteristics. The WS group also used more social and psychological categories in describing their self concept, and their responses were in general at a higher level than the responses for the PWS group. There were significant age-group differences, with the adolescents offering more self descriptions that were coded as physical and active, whereas the adults gave more social and psychological responses. Different themes were emphasized in the interviews from the two groups. These themes reflected the distinct phenotypes associated with these disorders, the participants life experiences, and their overall attitudes toward their syndrome.

Keywords: self concept; Williams syndrome; Prader–Willi syndrome

1. Introduction

Although the development of self understanding has captured the interest of psychologists for many years (Damon & Hart, 1982, 1988; Erickson, 1968; Harter, 1983, 1990; Kagan, 1982; Livesly & Bromley, 1973; Montemayor & Eisen, 1977; Secord & Peevers, 1974), only recently have researchers begun to

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examine how people with developmental disorders conceptualize themselves. The goal of this study was to explore self concepts in adolescents and adults with two genetically-based neurodevelopmental disorders, each with distinct cognitive and behavioral characteristics: Williams syndrome (WS) and Prader–Willi syndrome (PWS). Similarities and differences in how people with these syndromes reflect on their lives and view themselves, and changes in self concept in these populations between adolescence and adulthood were the focus of our investigation.

The notion of self concept represents a multifaceted construct comprising several dimensions that define a person's view of his or her 'personhood' or individuality. These typically include physical attributes, cognitive and psychological features, as well as interpersonal and social dimensions that ensure a connection to society, integration into a social-cultural world, and especially differentiation from others. Damon and Hart's model of self understanding (1982, 1988) provides a comprehensive framework for studying both the structural organization and developmental changes along multiple, interacting dimensions of the self concept. The basis of this model is the multidimensional self theory of James (1892), who first distinguished between the ''me''-aspect (i.e., the set of self attributes that one can objectively know, that are organized into a hierarchical structure of material, social, and spiritual constituents), and the ''I''-aspect (i.e., the person's subjective experience of individual identity reflected in an awareness of one's own distinctness, continuity over time, volition and reflective ability) of the self concept. Damon and Hart (1982, 1988) utilized this scheme as a theoretical starting point for devising their semi-structured self-understanding interview. Responses to interview questions are classified according to a coding scheme consisting of four self-as-object (the ''me'' in James' theory) *categories*—physical descriptors, activities, social and psychological aspects of self description—and three self-as-subject (the ''I'' in James' theory) *categories*—self continuity, distinctness, and agency. Categories are further classified into four *levels*, reflecting an increase in the complexity of self description, from defining the self in concrete terms of ''surface'', observable characteristics, to a concept of self as perceived in reference to the reactions of others and to one's life philosophy.

Besides its use in developmental studies of the self concepts of typical children and adolescents, this approach has also been proven sensitive to capturing the specificity of atypical developments in self understanding related to mental health difficulties, or neurodevelopmental disorders (e.g., studies of anorexic adolescent girls, see Schorin, 1985, adolescents with conduct disorder, see Melcher, 1986, children and adolescents with autism, see Lee & Hobson, 1998).

Investigations of self concepts among people with mental retardation or developmental disorders (Fine & Caldwell, 1967; Gowans & Hulbert, 1983; Shurr, Joiner, & Towne, 1970; Widaman, Macmillan, Hemsley, Little, & Balow, 1992; Zeitlin & Turner, 1988) have been sporadic and focused primarily on global self-worth or on academic self-perceptions. In many of these studies the nature of the participants' disabilities was often left unspecified, and it appears that

individuals with different syndromes or mental retardation etiologies were mixed into a single group and population differences were not explored. Our study used Damon and Hart's (1988) semi-structured interview to investigate in depth self concepts in two developmental disorders, WS and PWS, both of which have well-defined behavioral phenotypes.

1.1. Williams syndrome

WS is caused by a microdeletion on chromosome 7q11.23, a region that includes at least 20 genes, many of which have been mapped. It is characterized by a unique phenotype that typically includes dysmorphology, and an unusual combination of cognitive and behavioral features (Morris & Mervis, 1999). The majority of people with WS have mild to moderate levels of mental retardation, but their performance on standard IQ tests shows striking unevenness across subtests measuring different cognitive abilities (Bellugi, Mills, Jernigan, Hickok, & Galaburda, 1999): strengths in expressive language, auditory memory and face recognition skills (Bellugi, Wang, & Jernigan, 1994), but extremely low performance on tests of visual-spatial construction (Mervis et al., 2000). Musical abilities or intense interest in music and performing are a distinguishing feature of the phenotype.

People with WS are extremely sociable, extroverted, and highly empathic in their responses to other people (Gosch & Pankau, 1994; Jones et al., 2000; Udwin & Yule, 1991). However, this sociability does not translate into more advanced social cognitive abilities, such as performance on theory of mind tasks (Sullivan & Tager-Flusberg, 1999; Tager-Flusberg & Sullivan, 2000). This cognitive and behavioral phenotype clearly has the potential to influence the way others in their social environment interact with individuals with WS. It may lead to a sense of acceptance that would affect the way people with WS perceive their own success and failures, which, in turn, will modulate the valence of their self concept (Evans, 1998).

1.2. Prader–Willi syndrome

PWS is a genetic disorder caused by the absence of paternally expressed genes in the q11-q13 region of chromosome 15 either through deletion or maternal uniparental disomy (Butler, 1990; Thompson, Butler, MacLean, Joseph, & Delaney, 1999). Although they are not dysmorphic, people with PWS do have a characteristic facial appearance. They tend to be short in stature and have hypogonadism (Dykens & Cassidy, 1999). Similar to WS, the majority of individuals with PWS have borderline to moderate levels of mental retardation (Dykens, Hodapp, Walsh, & Nash, 1992). They do not have a distinctive cognitive profile, but visual-spatial processing is often a strength (Curfs, Wiegers, Sommers, Borghgraef, & Fryns, 1991).

The most striking feature of PWS is their excessive appetite, which, if left unchecked, results in extreme obesity that can be life-threatening. This hyperphagia colors much of the life of a person with PWS, placing restrictions on living

conditions, employment, and social activity. Obesity is a physical characteristic that is likely to have a significant impact on self concept. The extreme efforts made by parents to control the weight of their child with PWS may influence how much of the child's self conception relates to physical characteristics, or it may lead to a negative self conception rooted in this aspect of the disorder. PWS is also characterized by behavioral dysfunction, and a profile of negative maladaptive behaviors, including tantrums, obstinate and oppositional behavior, skin picking, impulsivity, aggression, obsessions and compulsions, such as hoarding (Dykens & Cassidy, 1995; Dykens & Kasari, 1997; State, Dykens, Rosner, Martin, & King, 1999). There is a relatively high rate of affective disorders, especially in adults with PWS (Beardsmore, Dorman, Cooper, & Webb, 1998). Their behavior is also sometimes described as inappropriate, especially in response to emotional situations (Sullivan & Tager-Flusberg, 1998), and they have poor coping skills on the Vineland socialization domain (Dykens et al., 1992). These social difficulties suggest that individuals with PWS may have a limited self understanding in terms of social and psychological constituents.

The present investigation explored the development of self concept in matched groups of adolescents and adults with WS or PWS, based on Damon and Hart's (1988) semi-structured interview. Individuals with PWS provide a good comparison group for the WS subjects due to the similar IQ distribution in the population and comparable language abilities, while their cognitive profile is not characterized by the discrepancies (peaks and valleys) associated with WS. Comparing groups well matched on general levels of cognitive functioning, but with distinct personality profiles and behavior, offers unique opportunities to explore if and how these characteristics would be reflected in their verbally expressed opinions about themselves.

2. Method

2.1. Participants

Twenty-eight individuals with WS (14 adolescents, 14 adults) and 28 individuals with PWS (equal numbers of adolescents and adults) participated in this study. All participants came from middle-class socio-economic backgrounds and were native English speakers.

All of the individuals with WS were diagnosed by a clinical geneticist as having the syndrome, and most had the diagnosis confirmed by genetic testing (Fluoresence in situ hybridization test). The majority (22) of these participants were interviewed at the 1998 Convention of the Williams Syndrome Association. The remaining 6 were referred by the New England Williams Syndrome Association and the National Williams Syndrome Association and were interviewed in their homes.

The participants with PWS were also all diagnosed by a clinical geneticist, and the majority had their diagnoses confirmed by genetic testing. Half the

participants with PWS were interviewed at the 1997 and 1998 National PWSA Conferences. Additional participants (9) were referred by the Prader–Willi Association of New England and were tested in their homes. Five adults with PWS were recruited through a residential rehabilitation facility in Brewster, MA, and were interviewed there.

Participants were administered Form M of the Peabody Picture Vocabulary Test-Revised (PPVT-R) (Dunn & Dunn, 1981), a receptive one-word vocabulary test, and the Kauffman Brief Intelligence Test (K-BIT) (Kauffman & Kauffman, 1990), a measure of general cognitive ability. The K-BIT yields verbal and non-verbal composite scores, as well as an overall composite score that is equivalent to a full-scale IQ score. The participants with WS were individually matched with the participants with PWS on chronological age, $t(54) = .2$, $p = .84$, PPVT-R standard score, $t(54) = .63$, $p = .53$, and full-scale IQ, $t(54) = .47$, $p = .64$. Details of the characteristics of the participants are summarized in Table 1.

2.2. Procedure

Damon and Hart's (1988) interview was individually administered to each participant by an investigator in a quiet room. The interviews, lasting between 30 and 60 min, were audio-taped and later transcribed.

The interview contains seven main items, shown in Table 2, that were presented in a fixed order. The wording of a question was slightly altered if the participants' initial response suggested that they did not fully understand the question. Additional questions, or probes, were asked to clarify the meaning and reasoning behind the initial responses. Probes were given until the participant repeated a response, said "I don't know", or showed frustration or signs of waning attention. The transcripts were coded by a rater blind to the participants'

Table 1
Participant characteristics

	WS adolescents	WS adults	PWS adolescents	PWS adults
	$N = 14$ (10 F; 4 M)	$N = 14$ (8 F; 6 M)	$N = 14$ (5 F; 9 M)	$N = 14$ (10 F; 4 M)
Chronological age				
M (SD)	13;4 (3;3)	27;7 (6;9)	13;4 (2;7)	29;1 (6;1)
Range	9;8–19;5[a]	18;4–41;7	8;10–18;1	20;6–39;0
PPVT-R standard score				
M (SD)	78 (14)	74 (18)	77 (16)	70 (15)
Range	56–107	44–100	51–101	32–89
Full-scale IQ score				
M (SD)	65 (13)	70 (14)	67 (13)	72 (15)
Range	45–92	40–93	48–97	40–96

[a] Because participants were individually matched on chronological age, PPVT-R standard scores and full scale IQ, one participant older than 18 had to be included in the group of adolescents.

Table 2
Items in the self understanding interview (from Damon & Hart, 1988)

Item 1: Self definition
- a. What are you like?
- b. What kind of a person are you? What does that say about you?
- c. What are you *not* like? What does that say about you?

Item 2: Self evaluation
- a. What are you especially proud of about yourself?
- b. What do you like most about yourself? What does that say about you? Why is that important?

Item 3: Self in past and present
- a. Do you think you'll be the same or different 5 years from now? What will be the same? What will be different? Why is that important?
- b. How about when you're an adult? Probe as in (a).
- c. How about 5 years ago? Probe as in (a).
- d. How about when you were a baby? Probe as in (a).

Item 4: Continuity
- a. If you change from year to year, how do you know it's still always you? Is that an important thing to say about you? Why? In what ways do you stay the same?

Item 5: Agency
- a. How did you get to be the way you are?

Item 6: Self interest
- a. If you could have 3 wishes, what would they be? Why would you wish for that?
- b. Are those things you hope for most in life? What else do you hope for in life? Why do you hope for that?
- c. What do you want to be like?
- d. What kind of a person do you want to be? Why do you want to be that way?
- e. What is good for you? Why is that good for you?

Item 7: Distinctness
- a. Do you think there is anyone else who is exactly like you?
- b. What makes you different from anyone you know? What difference does that make? Why is that important? In what other ways are you different? Are you completely different or just partly different? How do you know?

syndrome and age group, after having established reliability with a second rater on 15% of the protocols. The raters followed the coding scheme and procedures described below.

2.3. Coding scheme

Each transcript was first divided into units of scoring, called ''chunks''. A chunk is defined as a self-characteristic mentioned by the participant, together with responses to the related probe questions, as well as any further discussion of the specific characteristic that immediately followed the probed responses. Chunks were derived from responses throughout the interview, and were coded into four aspects of the self concept (*Categories of self concept*), then rated at one

of four levels (*Levels of description*), based on Damon and Hart's (1988) coding scheme and scoring criteria.

(a) *Categories of self concept*—Category assignments were made according to the following qualities of the **self-as-object** (hereafter, called 'self'): (1) *Physical*—Statements that refer to physical characteristics or material possessions; (2) *Active*—Statements that refer to one's abilities and typical activities; (3) *Social*—References to one's social relationships, group membership, or interpersonal interactions; (4) *Psychological*—Responses concerning one's thoughts, emotions, perceptions, beliefs, or knowledge.

(b) *Levels of description*—Level-ratings, which are orthogonal to the categories described above, were assigned in the following way: (1) A level 1 rating was assigned to responses that captured categorical descriptions of 'surface features', such as purely concrete, observable characteristics of the self or activity-based responses, with taxonomic value, but with no further underlying significance (e.g., *"I have curly hair (...) That's it"—for the physical self scheme; "I'm a happy person (...) because I'm not a sad person"*—for the psychological self scheme); (2) A level 2 rating was used for comparative assessments, in which information about the self was used to support actual or implied comparisons (e.g., *"I'm the best in class at running. I like running more than other kids"*—for the active self scheme); (3) A level 3 rating was given to responses concerned with the interpersonal implications of self characteristics (e.g., *"I'm very strong and in terrific shape, so everyone respects me for it"*—for the physical self scheme, or *"I'm a nice person (...) It is good if you want to have many friends"*—for the social self scheme); (4) A level 4 rating was given to responses that reflected a consciously systematic conception of self, as in statements referring to one's beliefs and life plans, goals, choices or personal life philosophy or outlook (e.g., *"Reading well, being a good reader helps you get into a good college and get a better chance of getting a good education; it makes me feel proud of myself and I also want to do something that requires a lot of education (...) be a writer or something"*—for the active self scheme).

As illustrated in the examples above, all *categories* can be defined in conjunction with all of the *levels*. For example, a physical statement would have a level 3 assignment if a physical feature was discussed in regard to how it impacted one's social appeal; or a psychological rating would have a level 1 assignment if the statement focused only on "moods, thoughts, feelings unrelated to permanent dispositions, abilities or beliefs" (Damon & Hart, 1988, p. 61).

(c) Content coding.

In addition to the coding system based on the method presented above, we also coded the presence or absence of recurring themes in participants' responses to three questions: (1) What are you most proud of?; (2) If you could have three wishes, what would they be?; (3) How did you get to be the way you are?

These questions were chosen because they provided opportunities for engaging participants in more in-depth, detailed discussions of those aspects of self concept that reflect specific choices, self evaluations, and reflection on their own agency and development. Finally, we looked across the entire interview to identify all references by the participants to their syndrome. These references were coded for whether they discussed their syndrome in positive, negative or neutral terms.

2.4. Coding procedures

Each transcript was randomly assigned a number so that the participant's diagnostic group membership (WS or PWS) and age group could not be identified. Two raters, of whom one was blind to the hypothesis and nature of the study, identified chunks in 30% (16) of the transcripts and compared the numbers obtained for reliability training for chunk parsing. After a high level of agreement (92%) regarding what constitutes a chunk was achieved, all chunks were then coded for categories and levels of the self concept scheme, according to Damon and Hart's (1988) scoring system.

Both raters scored 15% of the transcripts—which included 4 randomly selected WS transcripts (2 adolescents, 2 adults) and 4 PWS transcripts (2 adolescents, 2 adults)—to determine reliability for content and level coding. When chunks were codable at more than one level within the same category, only the highest-level score was assigned. If the chunk was codable in two or more categories at different levels, the final score indicated only the category with the highest level. The two raters achieved high proportions of agreement for *category* (Cohen's kappa = .85), and *level* (kappa = .83) ratings, as well as for *category by level* (kappa = .79).

3. Results

The results are presented in three sections. First, we compare verbal productivity measures related to the interview output in the two syndrome groups and two age groups. Next, we present analyses of the categories and levels of the self concept based on Damon and Hart's (1988) scoring criteria, focusing on significant differences as a function of syndrome and age group. The specific syndrome group comparisons of interest are those of proportions of social and psychological statements at higher levels of description provided by each group, while the specific age group comparisons of interest are related to differences in proportions of statements at higher versus lower levels of self-description across categories. Finally, we explore recurrent themes in the participants' self descriptions, using the content-based coding scheme that we developed to capture some of the particular concerns and aspects of self concept characteristic of the syndromic samples and age groups interviewed in this study.

We ran preliminary analyses comparing the responses of males and females on each of our coding schemes. However, we found no significant gender differences on any measures. To examine the potential role of IQ in relation to the measures of

self-concept used, we computed correlations between full-scale IQ scores and the productivity measure (total number of chunks), as well as *category* and *level* ratings. There were no significant correlations between full-scale IQ scores and the productivity measure and no systematic IQ effects for *category* and *level* ratings. Therefore, IQ scores were not included in subsequent statistical analyses.

3.1. Productivity

The participants with WS produced significantly more chunks overall than the participants with PWS (WS: $M = 17.9$, PWS: $M = 12.6$), $F(1, 54) = 14.44$, $p < .001$. There were no significant mean differences in the overall number of chunks as a function of age-group. Given the syndrome-group differences in productivity measures, all analyses were conducted using proportions for each variable. Specifically, we derived proportion scores by dividing the frequencies of each category at each level (physical levels 1–4, active levels 1–4; social levels 1–4; and psychological levels 1–4) by the total number of chunks provided by each participant within the self-as-object scheme. The 16 resulting scores for each participant reflect the proportion of responses rated in each category at each level within that participant's transcript, thus making scores comparable across transcripts, despite differences in the total number of chunks provided.

3.2. Categories and level analyses

The mean proportion of chunks by syndrome group, age group, category and level are presented in Table 3. Given the non-normal distributions of the dependent variables, nonparametric methods were used to examine differences in proportions of specific categories and level responses as a function of syndrome group or age group. We do not report all possible comparisons, but only those directly related to the expectation of syndrome group differences with respect to higher level social and psychological categories, and age group differences for proportions of lower versus higher level responses. However, in all analyses the alpha level was set conservatively at .01, given the fairly large number of comparisons conducted.

Consistent with our expectations, significant syndrome-group effects were found only for the social and psychological self-scheme categories. Mann–Whitney U tests indicated significant differences between the WS and the PWS groups for *social category* statements coded at level 3, $z = -2.85$, $p < .004$, and at level 4, $z = -2.79$, $p < .005$, and for *psychological category* statements at level 4, $z = -2.81$, $p < .005$, the WS group providing significantly more statements in these categories and levels than did the PWS group. Across categories, the WS group provided more level 4 responses overall than the PWS participants, $z = -3.36$, $p < .001$.

To examine developmental changes in self-descriptions, we conducted category and level comparisons for each syndrome group separately. In the WS group differences between adults and adolescents were significant for level 4 responses

Table 3
Mean proportions of chunks for the self-as-object scheme (categories by level), as a function of population and age group

Category	Williams syndrome		Prader–Willi syndrome	
	Adolescents	Adults	Adolescents	Adults
Physical				
Level 1	.283	.119	.320	.171
Level 2	.067	.075	.082	.080
Level 3	.024	.037	.005	.023
Level 4	.000	.056	.005	.012
Active				
Level 1	.133	.075	.220	.078
Level 2	.060	.040	.039	.076
Level 3	.031	.040	.046	.000
Level 4	.000	.044	.000	.007
Social				
Level 1	.083	.045	.099	.104
Level 2	.092	.081	.070	.143
Level 3	.027	.087	.003	.021
Level 4	.014	.072	.000	.007
Psychological				
Level 1	.068	.062	.038	.069
Level 2	.031	.072	.025	.113
Level 3	.060	.050	.042	.091
Level 4	.027	.045	.006	.000

overall, $z = -3.02$, $p < .003$. As expected, the adults provided more higher level responses in all categories than did the adolescents (see Table 3).

In the PWS group fewer significant differences by level were found for specific category responses, but the adolescents provided proportionally more level 1 responses than did the adults, $z = -2.44$, $p < .015$. As indicated in Table 3, level 3 and level 4 responses were very rare among the PWS participants across categories.

Overall, the participants with WS and the adults in both populations produced more 'sophisticated' responses than did the participants with PWS or the adolescents. This was reflected in the use of more social and psychological categories, as well as more categories rated at higher levels of description (levels 3 and 4), showing that they were able to take into account the interpersonal implications or the life-philosophy ramifications of the various self characteristics mentioned.

3.3. Content coding

3.3.1. Responses to three specific questions

To arrive at a richer picture of those aspects of self concept which may reveal personal concerns and features of self understanding unique to participants with

Table 4
Percentage of participants including content themes in responses

Content theme	WS adolescents	WS adults	PWS adolescents	PWS adults
(a) *Health*				
Weight	21.4	14.3	50.0	50.0
Medical conditions	7.1	28.6	7.1	.0
(b) *Possessions*				
Money	14.3	.0	35.7	21.4
Pets	21.4	.0	14.3	21.4
Trips, events	57.1	14.3	21.4	28.6
(c) *Religious faith*	42.9	57.1	.0	.0
(d) *Family and friends*	37.5	28.6	28.6	35.7
(e) *Humanitarian issues*	1.7	42.6	.0	.0
(f) *Plans for future*	14.3	85.7	35.1	42.6
(g) *Personal achievement*				
Grades, skills	71.4	50.0	50.0	64.3
Personal goals	7.1	50.0	7.1	7.1
(h) *Living place/autonomy*	14.3	21.4	14.3	21.4
(i) *Acceptance by others*	50.0	37.5	37.5	42.6

the developmental disorders interviewed, we analyzed a series of content categories from responses to specific questions. Table 4 presents the percentage of participants in each group mentioning each of the major themes identified. Only differences significant at a p-value less than .01 in proportion of participants mentioning each theme are reported. The content analysis revealed that even when both syndrome groups referred to the same or related themes, differences in their approach to common concerns emerged, as illustrated in some of the examples below. Two raters coded 20% of the transcripts for content categories, obtaining high reliability (kappa .82).

(a) *Health and weight issues.* Health-related hardships, issues regarding medication, surgeries, and illness were themes brought up by both the WS and the PWS groups. More adults with WS compared to the adults with PWS referred to aspects of their medical condition as part of their self description. However, they also explicitly mentioned wishes for good health and a long life, despite their current difficulties, while the adults with PWS never did so in the context of the interview. There were no significant differences related to these issues between adolescents' responses in the two groups.

A theme which clearly differentiated the two syndrome groups in the participants' self descriptions concerned food and weight references, brought up significantly more often by the participants with PWS, $\chi^2(1) = 6.45$, $p < .01$. For half of the PWS group, adolescents as well as adults, food and weight loss were often mentioned as salient issues around which the discussion of self concept tended to be organized. For example one adolescent male with PWS said: "*I am very proud about me losing weight. I lost over*

three hundred pounds. . . . I wish to get thin, so I'll get more friends . . . And they won't laugh and make fun of me."

(b) *Possessions/money, pets, material goods.* Personal possessions or wishes for such possessions were also an integral part of the participants' self concepts, especially among adolescents. Overall, more participants with PWS wished for money or to possess a house, car, or even their own business, compared to the WS group (see Table 4).

Adolescents in both syndrome groups mentioned having pets or a desire to work with animals in the future. Responses involving references to animals ranged from expressing the wish to own a cat or dog, to plans to work in a Zoo, or have a veterinary practice, or open a pet store with 'health-food' for animals. The following is a fairly typical response from an adolescent girl with WS: "*I have a cute dog . . . I love pets. . . . I want to be a veterinarian when I grow up*."

(c) *Faith, religion, church membership.* A striking difference between the two syndromes emerged from the spontaneous references to religion made by many of the participants with WS in contrast to the PWS group. Thus, half the WS group mentioned their faith in God, and Church membership as major sources of support in their lives, while none of the participants with PWS did so, $\chi^2(1) = 16.93$, $p < .0001$. For many individuals with WS, faith in God appeared to be part of their self-definition and a major source of strength in dealing with adversity. For example, one woman with WS said: "*I'm a person who believes in God . . . 'Cause if I didn't believe in God, I don't think I would have survived some of the stuff I've been through*."

(d) *Role of family and friends.* Participants in both syndrome groups mentioned their families and friends as playing an important role in self formation. About one-third of participants in each group mentioned the support and encouragement of their family, especially parents, in fostering their positive sense of self, in spite of their difficulties with the challenges of everyday life, including difficulties in social interactions with peers. An example illustrating this is taken from the interview of an adolescent female with WS: "*I was brought up in a home where they said to love someone and just love them for who they are and I was brought up and I was taught that way by my parents and other people around me*."

(e) *Altruistic and humanitarian issues.* Empathic responses were more prevalent among the adults with WS compared to the other groups. Significantly more adults with WS wished for improvement of social issues than the adults or adolescents with PWS, $\chi^2(1) = 7.64$, $p < .01$. Moreover, talking about their wishes, a substantial number of adults with WS referred to altruistic, socially relevant problems of general human significance as important to them, such as peace and freedom, eradicating homelessness, poverty, discrimination, or speaking out for the disabled, as illustrated in the following response from a woman with WS: "*I would wish that discrimination would stop . . . Because I don't like people getting teased over. I don't think it's right . . . I wish that everybody, that is out there, that is homeless and has a problem, has a family*

to go to. . . . Because I could trade up my home if I had a home and go live somewhere else so somebody could live in my house."

(f) *Plans for a mainstream future.* Both WS and PWS participants mentioned desires and hopes to live a 'normal life', described in cohort-appropriate activities and events, such as getting married, having children, getting a driver's license, going to college and even having a career. Proportionally however, more of the adults with WS (78.6%) mentioned marriage and starting a family as their main wishes, compared to adults with PWS (42.9%), or to adolescents with WS (14.3%), $\chi^2(1) = 11.63$, $p < .001$. The following example, taken from an adolescent girl with PWS, reflects such hopes and plans: "*I wish to have a normal life. To go to college and . . . receive a graduation diploma. . . . Go to college and then go to medical school. Then . . . become a doctor and a mother. . . . Because, again, I am the kind of person that I care about people . . . before I care about myself.*"

However, a common thread only among the participants with WS was planning for careers based on their special musical inclinations. Many of the adults (35.7%) and adolescents (42.8%) with WS considered themselves endowed with a special gift, and expressed their belief in the power of music to reach and transform people for the better, as illustrated in the following response from a woman with WS: "*I have always had this one dream in mind and that was to be a good musician and to perform for people and to do like benefit concerts and so that's what I want to do play my music and influence people and do it only for the good.*"

(g) *Personal achievement.* Doing well in school and obtaining good grades were common themes mentioned as achievement-related wishes. The WS group, however, mentioned having special talents more often than the PWS group did, although both groups expressed pride in their abilities and accomplishments in school, crafts, musical activities or sports. Two of the participants with WS expressed even more ambitious hopes for personal achievement. In fact, their dreams of careers were not restricted to the musical area, but reached into the larger entertainment world, as illustrated in the wishes of one of the adults with WS: "*I would like to be a movie star . . . I guess I could show the world what a Williams movie star could be.*"

(h) *Living place, control over the environment, social autonomy.* For all participants the desire and efforts to control their disability could be seen as part of their larger struggle for control over their environment and for social autonomy. In this respect, the issue of their living place becomes a salient aspect of self concept. Thus, those individuals living in residential placements expressed diverse, mixed feelings and attitudes about their situation, compared to participants living at home with their parents, or those living independently. Among the participants with PWS some (7.1%) strongly wished to be able to leave their group home and live independently in their own apartment, or return to living with their family, while others (10.7%) were very happy to be among people with the same syndrome, to have an organized schedule with opportunities for sports, exercise and

controlled diet, to benefit from the support of counselors and therapists, admitting that placement in the residential setting helped them tremendously in their struggle with disability. For the participants with WS it seemed that involvement in their Church or participation in Music Camps played a similar supportive role. Overall, many of the adults in both groups (21.4% vs. 14.3% adolescents) appeared to be concerned with issues of social autonomy (being able to live on their own), as well as social conformity (getting along with others and being liked by people).

(i) *Acceptance by others.* Social acceptance with dignity and respect appeared very important for both syndrome groups, as reflected in the words of one girl with WS: "*I want to be an independent, respectful [i.e., respected] person ... and, being respectful is hard for other people. But me, I know how important it is to respect people for who they are, and mostly for what they do ... If you don't respect someone, they aren't going to respect you, and it feels good to respect someone. Because you know they aren't going to hurt you, if you respect them.*"

Friendships are perceived as an important marker of social acceptance. Participants in both syndrome groups mentioned numerous times that friends were very important and often described themselves in terms of psychological qualities that help one 'make many friends' (e.g., being nice, helpful, kind, fun to be with). However, it appears that the actual friendships and social networks of individuals with WS or PWS are very limited and very few participants mentioned having one 'best friend'. As one girl with WS said: "*I don't really have any friends at all .... I hardly do, 'cause people make fun of me ... They hurt me.*"

Acceptance from others and empathy towards others appeared to be strongly interconnected in the self-conceptions of individuals with WS. Significantly more participants with WS (53.6%) expressed their interest in other people's feelings and problems compared to the PWS group (14.3%), $\chi^2(1) = 9.64$, $p < .002$. Moreover, the participants with WS in both age groups seemed to display a socially appropriate appreciation of the value of humor in human interaction, and often expressed pleasure and pride in their ability to make people laugh or to use humor in dealing with difficult situations.

3.3.2. References to syndrome

Participants in both the WS and PWS groups highlighted their syndrome as a salient aspect of their self-description. In fact, when asked the question "What are you like?" many participants responded by referring to their syndrome. Given the relevance of this theme for the self concepts of the participants in this study, references to syndrome were tallied throughout the interview. Table 5 presents the percentage of participants referring to their syndrome in positive, negative and neutral terms across all questions.

Overall, significantly more participants with WS (78.6%) mentioned their syndrome throughout the interview compared to the PWS group (39.3%), $\chi^2(1) = 7.29$, $p < .007$. For the participants with WS, syndrome appeared to

Table 5
Percentage of participants mentioning their syndrome

Content category	WS adolescents	WS adults	PWS adolescents	PWS adults
Positive reference	35.7	50.0	.0	14.3
Negative reference	7.1	21.4	14.3	35.7
Neutral reference	28.6	7.2	14.3	.0
Total	71.4	78.6	28.6	50.0

be so much part of their self definition that many interpreted the question "*Do you think there is anyone else exactly like you?*" in terms of references to their special condition, as illustrated by the response of an adolescent girl with WS: "*All of the Williams kids out there [are exactly like me]. Yep ... yeah we all look alike*".

Significantly more participants with WS (42.8%) in both age groups mentioned syndrome in positive light compared to the participants with PWS (7.1%), $\chi^2(1) = 9.52$, $p < .002$. Although some of the positive references to Williams syndrome were not followed by relevant justifications regarding the individual's sense of pride in having the syndrome, many responses included such justifications, based on comparison with other 'special conditions' and with 'normal people', as in the following example from a woman with WS: "*I'm proud that I have Williams syndrome. I'm proud that a lot of people care about the syndrome. Because it's different from Downs. And it's different from a normal person.*" In contrast, the participants with PWS often perceived their condition as a burden, a challenge they struggled with daily, an obstacle which interfered with their normal functioning in social contexts, and with their attempts to live independently (e.g., difficulties in keeping a job, struggling with anger, and efforts to control their behavior and their weight). Thus, the participants with PWS appeared to feel more marginalized by their condition, which involves both obesity and mental retardation.

However, a closer examination of the participants' references to their syndrome cautions against unmitigated generalizations about the two populations' attitudes toward their condition. Thus, many WS individuals, especially among the adults, revealed mixed attitudes and feelings regarding their syndrome. They acknowledged some of the social and medical difficulties related to "being Williams syndrome" (marginalization, lack of respect from others, difficulties in establishing long-term, emotionally involved relationships, including romantic relationships, need for medication and surgeries), and expressed their longing for 'normality'. In the words of one girl with WS: "*I wish to get rid of William's syndrome (...) Well, no matter how you are, it's not gonna be any different. People are still going to make fun of you.*" Such responses reflect a keen awareness of the well documented social and adaptive difficulties experienced by people with developmental disorders, awareness shared more by the adults than the adolescents in our sample, possibly as result of many years of encountering problems with social interactions in general.

4. Discussion

Our interviews with the participants in this study provided a rich portrait of how people with WS and PWS view themselves, understand their lives, and interpret their place in society. As in previous research, the self-understanding interview developed by Damon and Hart (1982, 1988), provided a unique tool for understanding the depth and limitations of the self concepts of adolescents and adults with these genetic syndromes.

Analyses of participants' responses revealed interesting parallels and differences between the two syndrome groups. One consistent finding across the interview was that the participants with WS offered more social and psychological responses that were rated at higher levels than the responses from the participants with PWS. They also provided more 'chunks' or units of information about their self concept overall. These differences between the groups, which denote developmentally more advanced self understanding in the participants with WS, cannot be interpreted in terms of differences in the cognitive or linguistic levels of the participants in the two groups because they were well matched on both IQ and language level.

One explanation for these findings is that they reflect the greater sociability of people with WS (e.g., Gosch & Pankau, 1994; Jones et al., 2000), a unique component of their phenotype. This suggests that they are more focused on social aspects of their environment, thus leading them to view themselves in the context of their relationships with other people. On this view, their social engagement leads to more advanced self understanding in people with WS, in comparison to other people with mental retardation. An alternative explanation is that the participants with PWS are constrained in their opportunities to develop greater awareness of themselves, thus limiting the developmental level that they achieve in self understanding. Primarily because of their hyperphagia, people with PWS live quite restricted lives, usually in more secluded environments, and they may therefore have fewer opportunities and more limited social contexts in which to develop a more reflective perspective on their self concept.

We also found interesting differences in the themes that emerged in the interviews in response to the three questions. Some of these reflect the striking and significant aspects of the phenotypes that are associated with each of these syndromes, which are clearly an important component of the self concept of individuals with WS or PWS. For example, participants with PWS were significantly more likely to discuss food and weight concerns, whereas the participants with WS, especially the adults, were more likely to discuss medical conditions and illnesses that are quite frequent in this population (Morris & Mervis, 1999). The participants with WS also discussed their anxiety, in terms of specific fears among the adolescents, or in a more generalized way among the adults (Gosch & Pankau, 1997). Interestingly, the participants with PWS did not mention any of their maladaptive behaviors, such as obsessive compulsive behaviors or skin picking, even though these are quite prevalent in this group (Dykens & Cassidy, 1995).

Some of the differences in the themes discussed by the groups were more surprising. One striking example is the significance of religion and the Church in the lives of people with WS, half of whom mentioned this in their interviews. For most of the participants with WS faith in God appeared to be an important source of support in their lives, especially in the context of belonging to a Church. Adults with WS were also more likely to express concern for broader humanitarian issues such as poverty and hunger. It is possible that people with WS may have been exposed to extensive discussion about these social issues when attending Church, so that these two themes are interrelated. Why is the Church, and faith in God so important to people with WS? One explanation for this is that the Church offers people with WS an opportunity to become a member of a community in a place where they will be accepted with their disability, thus meeting their needs for social interaction despite their difficulties making close friends. Church services are also appealing because they are filled with music, including singing by the whole congregation—clearly an attractive option for people who love music so much!

For both the participants with WS and PWS, one important component of their self concept is their syndrome, mentioned at least once during the interview by more than three-quarters of the participants with WS, and over one-third of the participants with PWS. In general, the participants with WS had more positive attitudes toward having WS, and saw it as less of a burden than did the participants with PWS. This is not surprising, given some of the negative characteristics of the PWS phenotype, especially the insatiable appetite that is a life-long struggle with potentially lethal consequences, for most individuals with this disorder. This focus on their syndrome, by both the WS and PWS groups, as an aspect of their self concept may also reflect how others respond to them. Other people tend to be charmed and react positively to the warm, friendly, musical, cute-looking people with WS. In contrast, many features of PWS, including the negative behavior, obesity, and skin-picking, may elicit more withdrawal, and disapproval from others. Thus, the self concept in these groups is shaped by their own perceptions of their physical appearance, talents and behavior, as well as the reactions they experience from others throughout their lives.

Perhaps the most poignant themes discussed by both groups in this study were their aspirations for the future, which they perceived in mainstream ways. The participants with WS and PWS discussed their plans for marriage, children, careers and social autonomy, even though few could realistically ever achieve such goals.

Another main question that we addressed in this study concerned developmental changes in self concept during adolescence and adulthood for people with mental retardation. In general, the developmental trajectory of self understanding for the WS and PWS groups is similar in sequence, but delayed, in comparison to what has been found for non-retarded individuals, providing support for the developmental approach to mental retardation (Hodapp, Burack, & Zigler, 1990). For both syndromes, participants' responses were rated at lower levels than expected for their respective age group in non-retarded individuals, and included

more physical and active self descriptors than other categories of self concept. This is the pattern of responses expected in younger non-retarded individuals, suggesting that self understanding is constrained by a person's cognitive and linguistic level, which mediate how people interpret their life experiences.

Nevertheless, among both the WS and PWS participants, several significant differences were found between the two age groups included in this study, as shown in Table 3. The adolescents generally provided responses that were coded in categories viewed as less advanced. focusing on physical and active aspects of the self. Even within these categories, the adolescents tended to provide responses that were coded at lower levels than did the adults, whose responses were more likely to be coded at level 3 or 4, reflecting an orientation toward the social qualities or the social implications of the self concept. These age-related changes in self description revealed that self concepts undergo a process of elaboration, suggesting development over time in the ways people with WS and PWS engage in self reflection.

Much of what we understand about people with these syndromes, or with other developmental disorders comes from the descriptions and reports from clinicians, researchers, or family members about their behavior and lives. Through the self-understanding interview that we administered we can begin to see how people with WS and PWS view themselves, using their own words to describe their self concept, and reflect on their lives. These interviews offer us a window on to their perceptions of themselves, their relationship with other people, the larger society in which they live, and their perspective on what their place is within that society.

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