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Genetic Syndromes of Mental Retardation

Should They Matter for the Early Interventionist?

**Robert M. Hodapp, PhD; Jean L. DesJardin, MS;
Leila A. Ricci, MA**

Over the past 20 years, increasing numbers of studies have examined the etiology-related behaviors in children with genetic mental retardation syndromes. In this article, we focus on the clinical aspects of such behaviors for interventionists working with young children. After reviewing the concept of *behavioral phenotype*, we discuss how etiology-related strengths and weaknesses gradually emerge with age and, potentially, interact with environmental input (emergentism). Second, given that children with different genetic disorders show diverse profiles of strengths and weaknesses, this article discusses how interventionists can best provide more focused and more effective interventions. **Key words:** *aptitude by treatment interactions (ATI), behavioral phenotypes, Down syndrome, emergentism, Williams syndrome*

IN THE COURSE of their day-to-day professional work, many early interventionists learn about different causes of mental retardation. These professionals work with families of a child with a particular condition, and have informally learned about the disorder from other professionals, parents, or parent-professional groups.

At the same time, however, knowledge of the child's etiology or cause of mental retardation is often considered unimportant by the formal field of early intervention. Historically, the research fields of psychology, early intervention, and special education have denied the importance of etiology-related behaviors (Hodapp & Dykens, 1994). In two widely used special education textbooks, for example, the authors argue that the focus of educational programs "varies according to the de-

gree of the student's retardation" (Hallahan & Kauffman, 2000, p. 138) and that "classification systems based on etiology or clinical types have little value in education." (Blackhurst & Berdine, 1993, p. 425) This disregard for the child's cause of mental retardation is common to most educational interventions for children of all ages.

Given this disjunction between interventionists themselves and their professional fields, to what extent should early interventionists care about the child's cause of mental retardation? Is it really the case that etiology has "little value" in early intervention work?

In this article, we argue that etiology does matter, that attention to the child's type of mental retardation may eventually lead to more precise, targeted forms of early intervention. Focusing on how different genetic disorders influence the behaviors of the young children who have them, we first provide some brief background, and then examples from several genetic mental retardation disorders. Only after defining and exemplifying this approach can we tackle issues related specifically to early intervention per se.

From the UCLA Graduate School of Education and Information Studies, Los Angeles, Calif.

Corresponding author: Robert M. Hodapp, UCLA Graduate School of Education and Information Studies, Los Angeles, Calif (e-mail: hodapp@gseis.ucla.edu).

HOW GENETIC DISORDERS INFLUENCE BEHAVIOR

Because of recent, remarkable advances in human genetics, we now know that mental retardation is caused by many different genetic disorders. At last count, approximately 750 genetic disorders were associated with mental retardation (Opitz, 1996). Many of these syndromes occur rarely, but also included are such commonly occurring disorders as Down syndrome and fragile X syndrome. Genetic disorders now account for a sizeable percentage of children with mental retardation, with estimates ranging from 1/3 to 1/2 of all such children (Rutter, Simonoff, & Plomin, 1996; Stromme & Hagberg, 2000).

In addition to physical-medical features, specific mental retardation syndromes also show etiology-related behaviors, or *behavioral phenotypes*. Etiology-related behaviors have increasingly been documented in Down syndrome, fragile X syndrome, Prader-Willi syndrome, and Williams syndrome, with some work also in 5 to 10 other genetic disorders. For many conditions, the numbers of behavioral research articles have more than doubled from the 1980s to the 1990s (Dykens & Hodapp, 2001).

But how, exactly, does a specific genetic disorder affect behavior? Although the exact mechanisms remain unknown, a few general principles seem clear. Such principles can be seen in the very definition of behavioral phenotype, which has been described as “the heightened probability or likelihood that people with a given syndrome will exhibit certain behavioral or developmental sequelae relative to those without the syndrome.” (Dykens, 1995, p. 523). Let us discuss 3 issues related to this definition.

Probabilistic nature of behavioral phenotypes

Focusing on *the heightened probability or likelihood* of certain behaviors highlights the probabilistic nature of genetic disorders’ behavioral phenotypes. That is, not every per-

son with a specific genetic disorder necessarily shows that disorder’s *characteristic* behavior or behaviors.

A good example concerns Down syndrome. As described below, children with Down syndrome generally show extreme difficulties in language (ie, difficulties beyond their overall mental ages). Yet even in the face of such usual deficits in language, there are exceptions. Consider Francoise, a 32-year-old woman with Down syndrome (trisomy 21), who has an overall IQ of 64. In an in-depth description, Rondal (1995) finds that Francoise’s language abilities are exceptionally strong. Francoise utters 10 to 15 word sentences that can also be complicated, with many indented or relative clauses. Although most individuals with Down syndrome show difficulties in language, exceptions do occur.

Unique versus shared behaviors

In summarizing the effects of different genetic disorders, Opitz (1985) concludes that “The causes are many, but the developmental outcomes are few.”^{10(p99)} As a result, a few genetic disorders display unique behavioral outcomes, whereas other outcomes are “shared” with one or more other disorders. For example, children with Prader-Willi syndrome show hyperphagia (extreme overeating) that is not found to such an extreme extent in any other genetic mental retardation disorder (Dykens & Cassidy, 1999). In the same way, children with Williams syndrome show a pattern of relatively high language, but exceptionally low visuospatial skills—again, such an extreme language-over-visuospatial profile has to date been found only in this single syndrome (Mervis, Morris, Bertrand, & Robinson, 1999). A few other genetic syndromes also show such 1:1 correspondences between 1 genetic disorder and 1 behavioral outcome.

More commonly, however, two or more genetic syndromes share a particular behavior or profile (Hodapp, 1997). To give one example, extremely high rates of hyperactivity are found both among boys with fragile X syndrome as well as among children with

5p- (cri-du-chat) syndrome. Similarly, 2 or more groups sometimes show an identical intellectual profile on a single task (even as they may differ on other tasks or behaviors). Such is the case in Prader-Willi syndrome and in fragile X syndrome. Although the 2 groups differ in many ways (eg, extreme overeating in Prader-Willi syndrome and extreme social anxiety in fragile X syndrome), both groups show higher abilities in *simultaneous* (ie, holistic, Gestalt) processing versus *sequential* (step-by-step, serial) processing on the Kaufman Assessment Battery for Children, or K-ABC (Kaufman & Kaufman, 1983; see Dykens, Hodapp, & Finucane, 2000, for reviews). Although a few genetic disorders are unique in their behaviors, more often several genetic disorders will "share" in some of their behavioral effects.

Possible intervention implications

Even given the above caveats, it now seems possible to attempt etiology-related interventions. Consider the case of Williams syndrome, a disorder caused by a microdeletion on chromosome 7 (Poher & Dykens, 1996). Children with this syndrome generally show a characteristic, "elfin-like" facial appearance, heart and other health problems, and hyperacusis, or a hypersensitivity to sound (Van Borsel, Curfs, & Fryns, 1997).

Behaviorally, children with Williams syndrome, as a group, show relatively strong abilities in language (Udwin & Yule, 1991) and are most often exceptionally—even indiscriminately—outgoing and friendly (Dykens & Rosner, 1999). In addition, many children with Williams syndrome are exceptionally fearful and anxious. Dykens (in press) recently found that more than half of children and adults with Williams syndrome reported being afraid of interpersonal situations (being teased, 92% of sample; getting punished, 85%; getting into arguments with others, 85%). Children with Williams syndrome also feared such physical events as shots-injections (90%), being in a fire or getting burned (82%), or getting stung by a bee

(79%), with still other fears related to these children's hyperacusis (loud noises-sirens, 87%; thunderstorms, 78%). Although not every child with Williams syndrome shows every fear, the vast majority seem overly fearful (and the numbers of such intense fears greatly surpassed fears in others with mental retardation).

How best to intervene with children with Williams syndrome? Dykens and Hodapp (1997) have proposed treatment-intervention guidelines related to these children's relatively high linguistic skills, their sociability, and their tendency toward fears and anxiety. Thus, children and adults with Williams syndrome might benefit from team or buddy systems at work or school and from working in people-oriented jobs (eg, receptionists). In contrast to many with mental retardation, these children and adults may also benefit from group or verbally oriented therapies. And, although sociable, children with Williams syndrome often need social skills training to enable them to make friends and to be wary of strangers. Each recommendation arises directly from etiology-related behaviors specific to Williams syndrome.

To date, such etiology-related interventions have mainly focused on clinical issues and mostly related to older persons, either children or adults. In addition, such guidelines remain suggestive; so far, the effectiveness of etiology-related treatments has not been evaluated. Still, given the recent advances in our knowledge of behavioral outcomes of many different genetic mental retardation syndromes, such recommendations make sense.

In extending our discussions to the early intervention field, we change our focus from maladaptive behaviors to profiles of strengths and weaknesses. Profiles can occur in such domains as language, cognition, motor, or adaptive skills. We also discuss how etiology-related strengths and weaknesses emerge over the first few years of life and how early intervention work might be facilitated by knowledge of such etiology-related developmental profiles.

GENETIC ETIOLOGY AND EARLY INTERVENTION

In considering the behavioral effects of different genetic disorders on young children, we focus on Down syndrome and Williams syndrome. Through these 2 syndromes, we begin to appreciate existing strengths—weaknesses and the possibilities for etiology-related strategies early intervention.

Examples from two genetic syndromes

Down syndrome

As the most common and most familiar genetic cause of mental retardation, Down syndrome is often thought by both professionals and nonprofessionals to be synonymous with mental retardation. But children with Down syndrome often differ in their behaviors from children with other types of mental retardation. Such etiology-related patterns appear in 2 areas: visual versus auditory short-term memory, and various aspects of language.

Children with Down syndrome display relative strengths in visual versus auditory short-term memory skills (Marcell, Armstrong, 1982; McDade & Adler, 1980). Specifically, children with Down syndrome show levels of visual short-term memory that are 1 or more years advanced over identical short-term memory tasks that are presented auditorily (see Hodapp, Evans, & Gray, 1999, for a review). Furthermore, babies and toddlers with Down syndrome show a tendency for remembering hand movements and other visual gestures (Harris, Bellugi, Bates, Jones, & Rossen, 1997). Indeed, toddlers with Down syndrome, compared to typically developing children at the same comprehension and production levels, had levels of gesture at the 77th and 80th percentiles, respectively.

In contrast, most children with Down syndrome display weak language skills in expressive language, grammar, and articulation. By the time overall mental ages reach 24 months, the majority show delays of 6 or more months in expressive versus receptive language abili-

ties (Miller, 1999). Most children with Down syndrome also have extreme difficulties in linguistic grammar (Fowler, 1990), with most showing expressive (and, to a lesser extent, receptive) grammatical levels below their levels of overall mental age (Chapman, Seung, Schwartz, & Kay-Raining Bird, 1998). In addition, mothers of almost 95% of children with Down syndrome report that their child sometimes or always has difficulty being understood by others (Kumin, 1994).

Williams syndrome

In contrast to children with Down syndrome, children with Williams syndrome show a relative strength in auditory short-term memory (Mervis et al., 1999) and in expressive language skills (Bellugi, Wang, & Jernigan, 1994). Although initial research suggested that children with Williams syndrome perform at their chronological age on a variety of linguistic tasks (Bellugi et al., 1994), such *chronological age-level performance* has now been found to occur in only about 5% of children with Williams syndrome (Bishop, 1999; Mervis et al., 1999). Nevertheless, language skills and auditory processing are considered relative strengths in children with Williams syndrome.

In contrast, many children with Williams syndrome perform poorly on tasks involving visuospatial skills (Udwin & Yule, 1991; Udwin, Yule & Martin, 1987). For example, children with Williams syndrome have trouble in putting together a jigsaw puzzle or in drawing (Bellugi et al., 1994; Dykens, Rosner, & Ly, 2001). These children appear to have difficulties in what have been called *constructive visuospatial skills*—visuospatial activities requiring one to mentally “put together” an object’s various pieces or parts. Table 1 summarizes the distinct strengths and weaknesses in both Down and Williams syndromes.

Strengths-weaknesses over age and the idea of emergentism

Complicating the picture still further is the idea of changing patterns of strengths and weaknesses with development. As a

Table 1. Summary of cognitive/linguistic profiles in children with Down syndrome and Williams syndrome

Syndrome	Strengths	Weaknesses	Everyday early learning activities/strategies
Down	Visual short-term memory Visual gestures	Auditory short-term memory Grammar Expressive language Articulation	<ul style="list-style-type: none"> • Visual cues • Picture/photo activity schedules • Color-coded picture stories • Sign language or gestures to supplement oral language • Use of real objects, materials, actual labels, and wrappers
Williams	Language skills Linguistic affect Auditory short-term memory Music	Visual-spatial construction Perceptual-motor Difficulty with loud sounds	<ul style="list-style-type: none"> • Auditory cues • Music activities, singing/rhyming songs, and toys with soft sounds • Teaching communication skills through daily routines • Use of language expansions and parallel talk

general rule, etiology-related strengths and weaknesses become more evident as children become older. For example, in children with Down syndrome, with increasing age their strength in visual memory tasks becomes more advanced compared to auditory memory tasks. Consider children's performances on 2 short-term memory tasks on the Stanford-Binet IV intelligence test. One measure—Bead Memory—involves visual short-term memory, whereas the other—Sentence Memory—involves auditory (and linguistic) short-term memory. Comparing groups of 5-10 year olds, 10-15 year olds, and 15-21 year olds, Hodapp and Ricci³⁴ have recently noted that the "Bead Memory over Sentence Memory" age-equivalent scores increasingly diverge with age. Among the 5-10 year olds with Down syndrome, the average difference was about 4 months (0.31 years), and only 36% of subjects showed a 6+ month advantage on the visual subtest. By 10 to 15 years, discrepancies averaged 8 months (0.66 years), with 60% of subjects showing 6+ month visual advantages. By 15 to 21 years, "visual over auditory" age-equivalent scores were, on average, 18 months (1.52 years) apart, and 75%

of subjects showed 6+ month discrepancies favoring visual short-term memory.

A similar finding has recently been noted in Williams syndrome. In this case, strengths involve levels of receptive vocabulary (on the Peabody Picture Vocabulary Test, or PPVT) and weaknesses involve drawing measures such as the Visual-Motor Integration (VMI) test (Bellugi, Mills, Jernigan, Hickok, & Galaburda, 1999; Jarrold, Baddeley, Hewes, & Phillips, 2001). As in Down syndrome, older compared to younger children showed more pronounced patterns of cognitive-linguistic strengths and weaknesses.

Why do such gaps widen as the child gets older? One theory—emergentism—holds that gaps widen because the child's learning is deeply and continuously embedded within the child's learning environment (Abbeduto, Evans, & Dolan, 2001). Thus, while many children with a particular genetic disorder show a propensity or disposition to display a particular strength or weakness even during the preschool years, parents, teachers, and others may then reinforce such propensities. Specifically, families may play to the child's strengths—encouraging what

the child is able to do more successfully—while avoiding behaviors or interactions that focus on the child's weaker areas. In the same way, children themselves may gravitate toward performing behaviors reflecting relative strengths as opposed to relative weaknesses.

PRACTICAL ISSUES

Playing to strengths or ameliorating weaknesses?

Given these etiology-related cognitive strengths and weaknesses—and the fact that the “strength over weakness” pattern becomes more pronounced with increasing age—early interventionists ultimately face the question of whether they should teach to the child's strengths, remediate weaknesses, or do both. Given that children with many syndromes gravitate toward their strengths—and avoid or circumvent their weaknesses—our general suggestion would be, whenever possible, to “play to the child's strengths.”

In making such recommendations, we are implicitly advocating what have been called *aptitude by treatment interactions*, or ATIs. According to the ATI approach, individuals with a particular aptitude respond better to one type of treatment, while those with a different aptitude benefit more from another treatment (Smith & Sechrest, 1991). In the case of children with genetic mental retardation syndromes, aptitudes consist of etiology-related cognitive and linguistic profiles.

Although aptitude X treatment interactions have not always been easy to document in clinical psychology (Smith & Sechrest, 1991), education (Braden & Kratochwill, 1997), or special education (Gresham & Witt, 1997), we believe this approach may nevertheless result in promising interventions for children with genetic mental retardation syndromes. Specifically, in considering children's cognitive-linguistic profiles, early interventionists should capitalize on children's strengths, thereby using strategies that are

most natural for the child to aid development.

One example might involve using visual modes as a way into language for young children with Down syndrome. Several studies have now examined children with Down syndrome by presenting them with interventions either using sign language or total communication (ie, both signs and spoken language). In all cases, children showed a sign advantage—with manual signs coming in before spoken words—which (as in typically developing children taught sign) was then followed shortly thereafter by spoken words (Abrahamsen, Cavallo, & McCluer, 1985; Kouri, 1989; Jago, Jago, & Hart, 1984). Similarly, at later ages Buckley (1999) reports success in teaching young children with Down syndrome to read. In comparing 2 groups of children with Down syndrome—those taught reading and those not taught reading—over a several year period, Laws, Buckley, Bird, MacDonald, and Broadley (1995) found that the reading group performed better on tasks of receptive vocabulary, receptive grammar, auditory memory, and visual memory.

In many ways, Down syndrome seems an ideal example of the strategy of playing to etiology-related strengths. Just as training in sign and in reading capitalize on visual strengths, both also allow entryways—and practice—in the weak domain of language. Similar recommendations are now needed in other disorders, and evaluations must determine whether *etiology-specific interventions* indeed work better than current approaches. In the years to come, we envision the development of interventions tailored to the cognitive-linguistic profiles of children with many different genetic disorders.

The role of parents and families

Families also play a key role in early intervention and in the future development of their children. Through collaborative interchanges between the family and early interventionist, professionals have opportunities to create mutual empowerment for families (Turnbull & Turnbull, 1997).

In the case of genetic mental retardation syndromes, early childhood interventionists need to value the knowledge and expertise that families already possess about their child and their child's syndrome. Granted, this information differs by syndrome. Compared to parents of children with Williams syndrome and other, lesser known syndromes, for example, parents of children with Down syndrome generally have more in-depth understandings of their child's etiology-related strengths and weaknesses (Fidler, Hodapp, & Dykens, in press). Even so, many parents of children with less common (or well-known) genetic syndromes become the "local expert" on the child's disorder, oftentimes being the sole person among the educational team who has any experience or knowledge of a particular syndrome (Fidler et al., in press; Hatton et al., 2000).

A further resource involves parent groups. Over the past few decades, parent-professional organizations have arisen to help families and professionals working with children with Down syndrome, Williams syndrome, and many other disorders. Groups are most often nationally based, sometimes with chapters in different regions or states. These groups provide literature, as well as annual or biennial conferences in which parents, researchers, and interventionists all interact. Information about such groups can be obtained through books (Dykens et al., 2000, pp. 307-308), Web sites (www.ndss.org for the National Down Syndrome Society, or www.williams-syndrome.org for Williams syndrome Association), or umbrella organizations (Alliance of Genetic Support Groups; the National Organization for Rare Genetic Disorders, NORD).

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TOWARD BETTER EARLY INTERVENTIONS FOR CHILDREN WITH GENETIC MENTAL RETARDATION DISORDERS

In considering how best to utilize information about etiology-related behaviors of children with genetic mental retardation disorders, we appreciate just how little we now know. Specifically, we know only the basics of cognitive, linguistic, and adaptive strengths and weaknesses, and this information exists only for a small number of the 750+ genetic mental retardation disorders. We also do not know if any etiology-based interventions are indeed more effective than current early intervention practices.

Nevertheless, recent advances lead us to hope for better interventions in the future. Over the past few years, we have indeed learned more—much more—about the behavioral phenotypes of children with several different genetic mental retardation disorders. Small, but growing, sets of American and European (mainly British) behavioral researchers now examine etiology-related behaviors, and knowledge is accumulating very rapidly.

As research information grows, the job ahead involves translating etiology-based behavioral information to help both parents and professionals, to develop etiology-based intervention programs, and to rigorously examine whether these programs work. Only then can we truly claim to have made the leap between research and practice, thereby helping the day-to-day practice of all interventionists as they work with parents and their children with various genetic forms of mental retardation.

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