The More You Know the Less You Know, But That’s OK: Developments in the Developmental Approach to Intellectual Disability

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Abstract

The adage “the more you know, the less you know” may best describe the contributions of the developmental approach to the study of persons with intellectual disability. Based on this premise, we trace the development of the approach from its origins in the 19th century, and highlight both the advances and difficulties of an increasingly precise science that involves differentiating among etiological groups and fine-tuned developmental concepts. We also provide an overview of the five sections of the volume.

Keywords: Intellectual disability, intellectual development, developmental approach, cognition, defect approach

The adage that “the more you know, the less you know” might best describe the contributions of the developmental approach to the study of persons with intellectual disability (ID). The tremendous growth of developmentally oriented research in the two decades since our (Hodapp, Burack, & Zigler, 1990) first volume on The Developmental Approach to Mental Retardation led to remarkable advances in understanding the developmental pathways of persons with ID across all areas of functioning in relation to specific etiologies and ages (e.g., see Burack, Hodapp, & Zigler, 1998). The broadening of the scope involves the implementation of the framework of the whole person, in which personality, the social and emotional characteristics of the person, and familial and larger contextual factors are considered in addition to cognitive functioning. Yet, the original adage also serves to highlight the vast task of precisely charting functioning in all the areas, sub-areas, and sub-sub-areas of functioning for each of the many different etiologies of ID at different ages in relation to familial, community, and environmental factors. Accordingly, the adoption of a developmental framework entails more fine-grained theory, more sophisticated experimental methodologies and, as a result, more knowledge about persons with ID at levels of both precision and breadth than could ever have been anticipated in prior conceptualizations.

The increased sophistication in science and specificity of knowledge highlight the futility of attempts to generalize findings across the heterogeneous group that we refer to as persons with ID and clearly renders meaningless the notion of a single grouping or field of research under the title of ID. Thus, the seemingly large advances in understanding only serve to set the stage for encounters with greater and increasingly fine-tuned, complex, and intertwined questions associated with etiology-specific conceptualizations within the context of developmental theory and considerations (for reviews, see Burack, Evans, Klaiman, & Iarocci, 2001; Burack, Root, & Shulman, 1996; Hodapp & Burack, 2006)—the more we know, the more we know how little we know.

Although facing the enormity of the developmental task may be daunting, it is preferable to the prior state of research on ID, in which the research
contribution might have best been described as a variation of our adage, or, "the more we think we know, the less we really know." In that context of monolithic pronouncements of the origins of ID regardless of etiology, in which persons with ID were considered a homogeneous group (for a discussion of predevelopmental conceptualizations of persons with intellectual disability, see Burack, 1990), big stories abounded, but none was scientifically sound and little was known about the nuances of each group or individual. Although each of more than 1,000 etiologies associated with ID differs from all the others in meaningful ways in virtually every aspect of functioning, persons with ID were grouped together as if they were a single entity, thereby obscuring important characteristics associated with the individual etiologies. Flawed research methodologies, including the obviously erroneous practice of matching persons with and without ID on chronological age (CA), led to false claims of defects among persons with ID in a variety of areas of functioning (for discussions of these shortcomings, see Burack et al., 2001; Iarocci & Burack, 1998). Thus, much was thought to be known about ID on a grand scale, but the reality was that the lack of theoretical and scientific precision left the field in the shambles of ignorance—"the more we knew, the less we knew."

Yet, this is not a call of despair. The goal of the developmental approach was to promote the vision of theoretically sophisticated and methodologically precise research that can be cobbled together to provide a more precise understanding of persons with ID. This approach has its origins in 19th-century writings of Langdon Down and William Wetherspoon Ireland, who pioneered etiologically-specific research, and in the 20th-century writings by developmental theorists, including Heinz Werner, Edward Zigler, Dante Cicchetti, and their colleagues, who promoted the interface of developmental psychology and the study of ID. This approach finds fruition in the 21st-century sophistication of experimental technology and empirical methodology in the study of genetics, brain functioning, behavior, social and interpersonal functioning, and emotional well-being, as well as in the study of the relations among them. Through this synergy, the key to understanding the heterogeneous group of persons who fall under the diagnostic heading of ID is a bottom-up process, with small but fine-tuned and precise empirical "stories" of smaller homogeneous groupings, rather than a top-down process with bigger and more general, but essentially flawed accounts. With the developmental approach, we know much more about persons with ID than we have in the past, but are also painfully aware of the extent to which we only tap the surface of all there is to know; thus, the more we know, the less we know, but that is OK, because—"the more we know the less we know, the more we (really) know."

Theoretical Shift from a Singular Defect to a More Fine-tuned Understanding of Development

The study of cognitive and neurocognitive functions or abilities may best highlight the contributions of the developmental approach to ID. These types of functions were the primary focus of scientific research on ID during the second half of the 20th century, when most empirical work in the field was characterized by a race to identify "the deficit" that was the primary cause or marker of reduced intellectual functioning. Those who undertook this frantic search emphasized broad constructs of cognition that were considered essential across all domains of functioning, including cognitive rigidity, memory processes, discrimination learning, and attention—retention capabilities, among many others (for a review of these approaches, see Burack, 1990). With the use of experimental paradigms that were sophisticated for the time, researchers presented compelling evidence of deficient performance in virtually all of these areas of functioning, and each specific defect was touted as the central cause of ID. Unfortunately, the evidence was fatally flawed as the researchers failed to consider essential and seemingly obvious conceptual and methodological issues, such as the inherent differences in developmental levels of functioning between persons with and without ID of the same CA, the multiplicity of etiologies associated with ID, the uniqueness of each etiology with regard to phenotypic expression, and social factors related to the life experiences of persons with ID (for reviews, see Burack et al., 2001; Hodapp & Zigler, 1986).

In critiquing and debunking the various claims of the defect theorists, Zigler and colleagues (e.g., Hodapp, Burack, & Zigler, 1990b, 1998; Zigler, 1967, 1969; Zigler & Balla, 1982; Zigler & Hodapp, 1986) highlighted these formerly ignored issues as the hallmarks of a nascent approach to ID that would be based on classic developmental theory. As the developmental approach evolved, the critiques led to the growth of more sophisticated conceptual and methodological frameworks that, in
turn, resulted in more nuanced and precise understandings of persons with ID. For example, in his classic early articulation of this approach, Zigler (1967) proposed the “two group approach to mental retardation” and argued that persons for whom the cause of ID was familial should be differentiated from those for whom ID could be classified as organic. In this framework, persons with familial ID are simply those at the lower end of the normal distribution of intelligence whose development should, therefore, be typical in every way except that it occurs at a slower rate and reaches a lower asymptote. In contrast, persons with ID associated with organic causes could not be expected to show typical patterns of development because of the lack of integrity of their physiological systems. Differences between these groups were not expected in the developmental sequence of the acquisition of skills within any given domain of functioning as these sequences are considered to be universal (for a review, see Hodapp, 1990), but rather in the profile of functioning across domains (Burack, 1990; Weiss, Weisz, & Bronfield, 1986; Weisz & Yeates, 1981; Zigler & Hodapp, 1986). As the two-group approach was extended, each of the more than 1,000 etiologies and subetiologies (e.g., trisomy 21, mosaicism, and translocation forms of Down syndrome [DS]) was associated with a unique pattern of strengths and weaknesses, and in many occasions, with a specific profile of developmental trajectory (see Burack, Hodapp, & Zigler, 1988; Dykens & Hodapp, 1999).

The impact of Zigler’s theory is reflected in several methodological considerations that are now considered fundamental to research on ID. The first methodological impact is reflected in the issue of developmental level and the use of mental age (MA) matching to compare the performance between groups of persons with ID in relation to other groups, most commonly typically developing persons, to determine whether specific aspects of functioning are commensurate with or impaired relative to the comparison group. When compared to typically developing persons, the use of MA matching provides an implicit measure of functioning in relation to the expected developmental level for that function.

The second advance in research is reflected in the study of profiles of cognition and social functioning that are etiology specific. As advances in the field of genetics led to the discovery of even more etiologies associated with ID, the need to further differentiate groups became apparent, and the quest to find the core deficit of ID was largely abandoned in favor of the characterization of strengths and weaknesses across etiologies.

Considering Developmental Level
The most obvious, and therefore the most troubling, of the essential flaws that were inherent to the defect approach is that the groups of persons with and without ID within a study were typically matched on CA (for a discussion of the implications of these problematic outcomes, see Burack et al., 2001). By definition, then, those persons with ID were lower functioning than those without ID and would be expected to perform worse on any task that was age-appropriate and sufficiently sensitive to differentiate between groups with considerably different levels of functioning. Yet, despite the inevitability of the findings of group differences, the defect theorists cited the impaired performance among the persons with ID as evidence of a core deficit.

In highlighting one example of the extent to which advocates of the defect approach misled the field, Burack et al. (2001) and Iarocci and Burack (1998) demonstrated that the notion of attention as the core (or at least a central) defect, which was perpetuated from the 1960s through the 1990s, was based on a series of articles in which matching was exclusively based on CA. Accordingly, Burack and colleagues argued that the findings of a deficit was inevitable for virtually every aspect of attention. In other words, the proponents of the attention defect theory had simply found that “lower functioning persons were functioning at lower levels than higher functioning persons.” This, of course, is not at all surprising, and not at all informative. In contrast, Iarocci and Burack highlighted in a review of the literature that when matching was based on MA, attentional functioning was generally developmentally appropriate with some exceptions in certain aspects of functioning among persons with specific syndromes.

One common argument among the proponents of CA matching is that this approach allows for the identification of areas of “spared of abilities.” The idea is that if persons with IDs perform similarly to a CA-matched group of typically developing persons in a specific area of functioning, then that area could be considered to be uniquely “spared.” Unfortunately, this reasoning is inconsistent with fundamental tenets of experimental research. The failure to find group differences can rarely be considered unqualified, or even strong, evidence that the groups perform similarly. Rather, this finding is
more likely the consequence of one or more methodological problems. In the case of research on persons with ID, the failure to find group differences is often due to the use of participant groups that are not functioning at the developmental levels at which differences in the specific area of functioning might optimally be identified. For example, if an area of functioning emerges at age X, testing groups considerably prior to age X would not elicit group differences since the area of functioning would not have yet developed for even the typically developing persons, and both typically developing children and children with ID would display poor levels of performance. Conversely, differences would be less likely to be elicited at ages that are significantly older than age X with persons with ID would have had ample opportunity to attain the requisite skills, even if their development was slower as compared to typically developing persons. Even in the case in which the participants are all at the ideal age for finding group differences, the failure to find these differences might be attributable to the lack of sufficient sensitivity of the task to differentiate level of performance among groups. Thus, without more evidence, the failure to find group differences tells us little about the relative abilities of the groups, and the case for the “spare abilities” is more likely a consequent of problematic methodology than of any meaningful characteristic of the specific group.

The obvious, and simplest, solution to the matching problem of comparing groups with different levels of ability is to utilize groups with similar levels of ability. In the study of cognitive and cognitive-related tasks among persons with and without intellectual disabilities, the relevant aspect of developmental level is intellectual functioning, or MA, and the task is to include groups of persons with and without ID that are similar on these measures. In this way, findings of deficits among persons with IDs cannot simply be attributed to the generally lower levels of functioning. Rather, the case might be made that the deficit in the area of interest among the group of persons with ID is evident even when general functioning is equated. This is, of course, a much stronger argument than when the level of functioning differs between the groups. Thus, the original call to match by MA, or general developmental level (for a review, see Burack et al., 2001), was a meaningful improvement over the CA-matching strategies of yesteryear since the apparent deficits of persons with ID could not be simply attributed to a priori levels of development and functioning.

Despite these advantages of MA as the primary matching variable, the increased precision that is associated with MA only serves to highlight that the process of matching is inherently flawed. It is used to equate two groups that are essentially different, so that they can be compared on some variable—thereby allowing researchers to conclude that any discrepancies in performance or scores can be considered a consequence of some essential difference, rather than of any a priori differences between the groups. However, the inherent difficulty with this type of task is that groups that are essentially different are, in fact, essentially different. For example, a group of children with DS can be matched to typically developing children on MA, but many crucial differences between the groups can effect performance on the relevant tasks, including differences in developmental rate, CA, a life time of experiences, physical and motor abilities, and interaction styles, among many others.

Even in the imaginary scenario in which a person with ID and a typically developing person have the exact same MA for a moment in time, they would differ in many different ways. By definition, the typically developing child would have attained the given MA at a faster rate and at a younger age, whereas the person with ID would have lived longer and had more life experiences. And, the nature of those past life experiences would likely have varied considerably, as typically developing children are likely to have experienced considerably more successes and positive reinforcement than their peers with ID. Their futures would also diverge. In the case of an “ideal” test with perfect sensitivity and reliability, the moment the MA of the two children are perfectly commensurate, the typically developing child would surge ahead.

Despite some conceptual concerns about the trajectory of MA, basic MA matching is particularly useful in the study of persons whose ID is familial, who, like typically developing persons, are thought to show relatively flat profiles across cognitive domains. However, with the advent of primary interest in the different etiological groups and in the differences in profile among them, general scores of MA were no longer sufficient to mitigate against claims of a priori differences between the groups.

Among others, Burack et al. (2004) highlight the need to utilize matching measures that are even more precise than MA, so that the matching is not by some general construct of developmental level but rather is linked to the development of abilities that are pertinent to the specific function or task.
This strategy minimizes the chances that differences in performance between the groups might be an artifact of a specific relative strength or weakness displayed by one of the groups in the area of functioning related to the experimental task.

The advances in matching by specific aspects of developmental level allow for considerably more precise assessments of the implications of various findings but, as in many other examples of development, progress is also associated with some reorganization. For example, the intricacies and difficulties in optimizing matching strategies even led to some recent calls to forego matching in favor of the use of regression models that allow researchers to chart developmental changes (e.g., Jarrold & Brock, 2004). Thus, the discussions are less about "the way" or even "best way" to match, but rather, more humbly, the "least bad way" for the specific study. These types of discussions both highlight the essential contribution of the consideration of developmental level and matching strategies to understanding the performance in specific aspects of functioning among persons with ID and point the way to the need for continued development of the thinking and research in this field.

The Developmental Approach as Represented in This Volume

This volume differs from many Handbooks as the contributors were not simply assigned specific topics to review, but rather were asked to provide original conceptual contributions in their general areas of research. Thus, they were able to uniquely frame their chapters in ways that could maximize their contribution to both the volume and the general literature. As a result, the chapters differ considerably in their orientation. In some chapters, specific topics are discussed in relation to different etiological groups, whereas the focus in others is on a specific etiological group in relation to a given topic. We imposed a structure of five general sections, with each focused on a domain of functioning or aspect of life that is inherent to an integrated, transactional perspective of development within the context of the unique manifestations evident across the specific etiological groups associated with ID. The focus of the first section is genetics, with examples of the expression of genes in various etiological groups. The next two sections are focused on the development of cognition and of language. The focus of the fourth section is socioemotional development and the fifth on the relationships among persons with ID, the members of their families, and the broader environment. In all cases, the emphasis is on delineating a more fine-tuned understanding of ID by focusing on developmental processes for one or more well-defined etiological groups.

In the opening section on genetics, intelligence, and behavior, Iarocci and Petrill focus on ID that is not associated with a specific disorder or condition, but rather is thought to be due to natural genetic variation. They review the history of the two-group approach and the need to differentiate between organic and nonpathological forms of ID, and highlight the concept of "polygenic inheritance" to discuss extreme variations in IQ, and in particular, the low IQ that is found in the nonorganic type of ID. In the next two chapters, the focus is the relationship of specific genetic syndromes in relation to behavioral phenotypes. Elsabbagh and Karmiloff-Smith discuss genetic, developmental, neuroanatomical, and behavioral characteristics of persons with Williams syndrome (WS) and depict how these characteristics are incorporated into theoretical models of gene–environment interactions. Cornish, Bertone, Kogan and Sceur focus specifically on fragile X syndrome (FXS), a particularly striking case of gene–behavior interaction, as the level of intellectual impairment of a person with FXS is inextricably linked to the number of repeats of a particular DNA sequence. With the greater the number of the repeats, the greater the severity of the disorder and the intellectual impairments associated with it.

The second section is on cognitive development. In the opening chapter, Landau highlights the manner in which a fine-tuned understanding of a specific etiological group can inform about general developmental processes—in this case, the organization and development of spatial representation are examined through the unique example of WS. Landau proposes that development involves the specialization of function, factors that constrain each specialized system, and the importance of timing in the emergence of brain and cognitive systems, and she applies these principles to the development of spatial representation among both persons with WS and typically developing persons. The focus of the next several chapters in this section is a specific aspect of cognitive development and the extent to which relevant performance is impaired in different etiological groups. Iarocci, Porporino, Enns, and Burack update the review of literature on attention from the first edition of the Handbook as they examine the finding on disparate aspects of attention among persons with various etiologies, including
DS, WS, and FXS. They apply a framework for understanding developmental change and stability in selective attention that is based on the fundamental dimensions of attentional selection. Specifically, they explore whether attentional selection occurs with or without awareness and whether the origin of the selective process is exogenous, and does not require learning, or is endogenous, and involves learning and prior experience. Vicari reports on specific profiles of memory capacities among persons with IDs of different etiologies, especially individuals with DS and WS. Consistent with a neuropsychological approach, distinct memory profiles among persons with genetic syndromes can be traced to the characteristics of brain development and architecture. Jarrold and Brock refer to Baddeley's (1986) model, in which working memory involves the combined functioning of a central control system (the central executive) and two peripheral short-term memory systems (the phonological loop and the visuo-spatial sketch pad) that are specialized for the maintenance of verbal and visuo-spatial information, respectively. Jarrold and Brock highlight the range of working memory impairments found across the genetic syndromes associated ID. Russo, Dawkins, Huizinga, and Burack examine various aspects of executive functioning among person with DS, FXS, Prader-Willi syndrome, and phenylketonuria. They structure their interpretation of findings to fit within the context of a developmental framework and consider the methodological issues inherently related to the study of executive function among persons with IDs. Bhatara, Quintin, and Levitin address the link between intelligence and musical ability among individuals with WS, DS, FXS, tuberous sclerosis complex, and Rett syndrome. Key and Thornton-Wells end the section with an introduction to event-related potential and magnetic resonance imaging technology for studying cognition among persons with ID, and they illustrate how these methods are used with persons with DS, Prader-Willi syndrome, WS, and FXS. Across the chapters in this section, the need to differentiate by specific etiology is highlighted as the groups are unique in so many ways, even as they share the common criterion for ID of a significantly lowered intellectual functioning.

In the section on language development, the focus is on the development of language within specific etiological groups, with two chapters on DS and one each on FXS and WS. Chapman and Kay-Raining Bird summarize the strengths and weaknesses of the emerging language profile among children, adolescents, and young adults with DS. Kay-Raining Bird and Chapman focus on literacy, and highlight the evidence that interventions can improve emergent literacy, word recognition and decoding skills, orthographic knowledge, and phonological awareness among persons with DS. Abbeduto, McDuffie, Brady, and Kover provide a comprehensive characterization of language problems typically associated with FXS. They describe the extent and profile of delays and impairments, the syndrome-specific features and within-syndrome variation of the linguistic profile, and how both are influenced by genetic and environmental factors. Mervis considers research on early language acquisition, and on the language abilities of school-aged children and adolescents with WS. She argues that, rather than providing evidence for the independence of language from cognition, the evidence from WS is consistent with their interdependence throughout development. As in the case of cognitive development, the chapters on language highlight the unique patterns of development seen across different etiologies, and highlight the need for a more fine-tuned understanding of the relevant developmental trajectories.

The fourth section is on social-emotional development, and the chapters are organized so that each addresses a different aspect of social-emotional development, one on social-emotional development in relation to brain activity, and one in relation to the diagnosis of autism and dual diagnoses. Kasari, Jahromi, and Gulsrud highlight essential developmental milestones in social-emotional development for typical child development and use this model as a framework for considering the extent to which children with developmental disabilities are delayed or different in their emotional development. They address several areas of emotional development, including emotion recognition and understanding, emotional expressions, emotional responsiveness, and emotion regulation among children with autism, DS, and FXS. Niccols, Thomas, and Schmidt synthesize the current state of research on the relationship among brain, behavior, and social-emotional development in children with the six most common genetic syndromes associated with an ID, including DS, FXS, 22q11.2, Williams, Prader-Willi, and Angelman syndromes. They conclude that the increased prevalence of some socioemotional difficulties in children with genetic syndromes may be partially explained by neurodevelopmental differences. Moss, Howlin, and Oliver provide an extensive review of the assessment and presentation-related behaviors associated with tuberous sclerosis, Angelman, and Prader-Willi syndromes. One is an etiologically heterogeneous condition that includes conditions that involve specific brain malformations, with the most frequent being focal malformations of the cerebellum, cerebellar vermis, and brainstem. The review emphasizes the importance of recognizing and addressing the cognitive, academic, and social-emotional needs of children with tuberous sclerosis.
and presentation of autism spectrum disorder and related behaviors among individuals with severe ID associated with several genetic syndromes, including tuberous sclerosis complex, FXS, DS, WS, and Angelman, Coffin-Lowry, Cohen, Rett, and Cornelia de Lange syndromes. The relatively frequent comorbidity between certain syndromes and autism spectrum disorder suggests possible links that might inform about the syndrome, autism spectrum disorders, and the relationship between them. The unique patterns of social-emotional development and prevalence of autism spectrum disorders across different etiologies highlight that etiological differences extend beyond the domains of functioning, cognition, and language most associated with ID to affect every aspect of individuals’ lives.

The final section of this volume is on relationships between persons with ID and the various members of their family across the lifespan. In delineating the issues faced by family members, especially mothers, some of the contributions include first-person accounts. Glidden focuses on the reactions of parents to raising a child with an ID. She addresses reports of both positive and negative reactions, as well as the intensity and duration of the reaction to having children with DS, FXS, and Smith Magenis, Prader-Willi, and other forms of ID. Hauser-Cram, Howell-Moneta, and Young discuss the physical and behavioral characteristics of children with DS and WS that influence their social interactions, and conclude that similarities and contrasts between profiles of children with each syndrome are informative for those who seek possible avenues of intervention for promoting optimal mother–child interactions when the child has a genetic etiology. Feninger-Schaal et al. provide a review of attachment theory, with an emphasis on factors relating to the parents’ representation of their children, as reflected both in their insightfulness into the experiences of their child and their reactions to their child’s diagnosis. Both factors impact caregiving behavior and the attachment that children form to their parents. Al-Yagon and Margalit examine parents’ perspectives on having children with DS in an attempt to characterize family sources of stress and coping. Fidler contrasts two frameworks used for understanding the effect on families of children with different etiologies including DS, FXS, WS, Smith Magenis syndrome, Prader-Willi syndrome, and Angelman syndrome. One is an etiology-specific framework rooted in Hodapp’s (1997) notion of direct and indirect effects in families of children with ID of different etiologies, and the other is a biocultural framework that provides a more distal account of child eliciting factors in children with disabilities and takes into account evolutionary influences on parent–child relationships. Esbensen, Selzler, and Krauss present a life course perspective on research on families of persons with specific genetic syndromes. They focus on patterns of individual developmental trajectories, with an emphasis on the effect of context on outcome. The editors conclude the volume with some reflections on its contributions to the theory, research, and understanding of persons with intellectual disability from a developmental perspective, and outline directions for future work in the field.

As developmental approaches to understanding persons with ID continue to emerge, the contributions to this volume provide conceptual foundations for examining the developmental trajectories across persons with any of the many different etiologies. More than 40 papers after Zigler’s (1967) initial call to arms for a developmental approach to ID, and 20 years after our initial volume (Hodapp, Burack, & Zigler, 1990)—the more we know, the more we need to know (even) more.

Acknowledgments

Work on this chapter was supported by funding from the Social Sciences and Humanities Research Council of Canada to Jacob A. (Jake) Burack.

References


